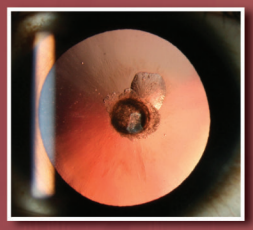
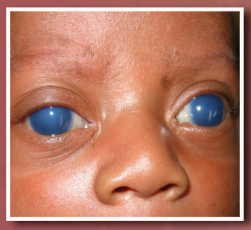
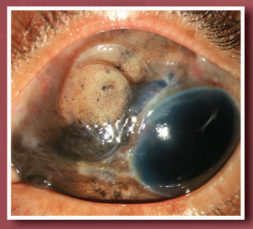
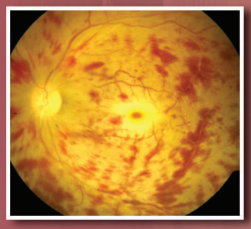
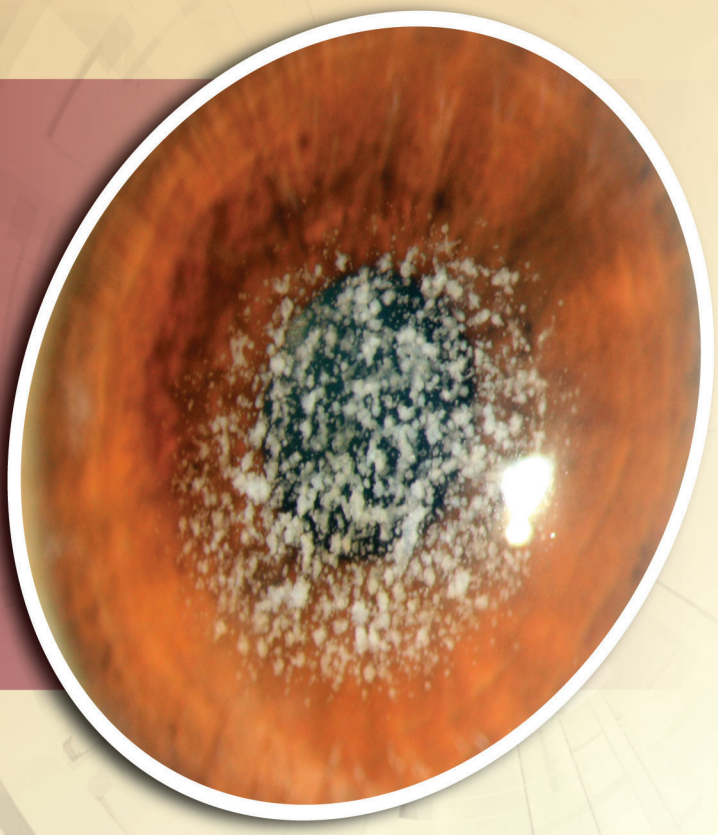
ATLAS OF CLINICAL OPHTHALMOLOGY

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- Baggy eyelids
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- Ankyloblepharon
- Nodular hemangiomas
- Tarsorrhaphy

Congenital Lid Conditions

Epicanthus

- Most common congenital lid condition
- Unilateral or bilateral
- May give rise to pseudo-convergent squint
- Semilunar vertical skin folds at the medial canthi running between two eyelids **(Fig. 1.1.1)**
- Four types:
  - *Epicanthus tarsalis:* Fold is most prominent in the upper lid **(Fig. 1.1.2)**
  - *Epicanthus palpebralis:* Skin fold is equally distributed is the upper and lower eyelids **(Fig. 1.1.3)**
  - *Epicanthus inversus:* Fold is most prominent in the lower eyelid **(Fig. 1.1.4)**
  - *Epicanthus supraciliaris:* Skinfold covering the medical canthus, extends high up to the eyebrow.
- *Treatment:* By plastic repair.

Fig. 1.1.1: Epicanthus

Fig. 1.1.2: Epicanthus tarsalis

Fig. 1.1.3: Epicanthus palpebralis

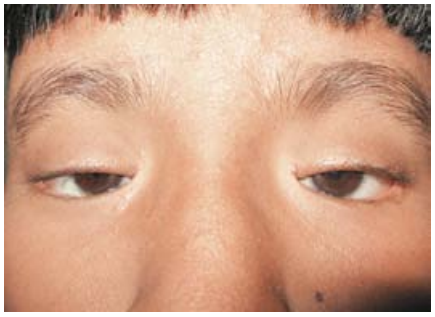


Fig. 1.1.4: Epicanthus inversus

Telecanthus

- Increased distance between medial canthi **(Figs 1.2.1 and 1.2.2)**
- Due to long abnormal medial canthal tendon
- Should not be confused with hypertelorism **(Fig. 1.2.3)** in which there is wide separation of bony orbits
- May also give rise to pseudo-convergent squint **(Figs 1.2.4A and B)**

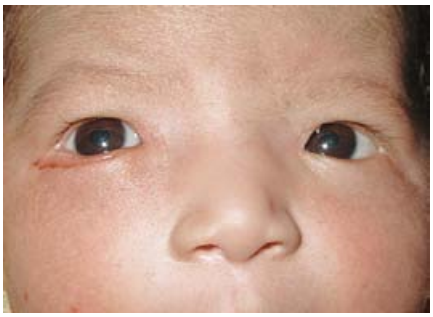


Fig. 1.2.1: Telecanthus

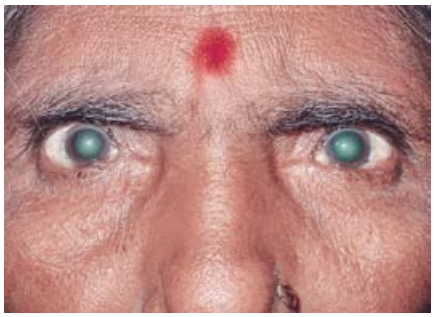


Fig. 1.2.2: Telecanthus

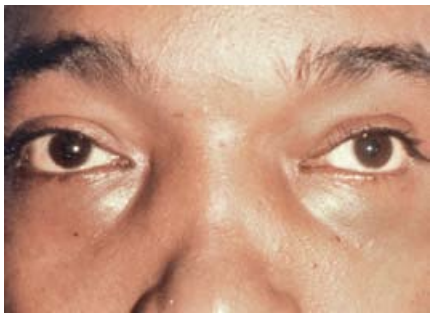


Fig. 1.2.3: Hypertelorism

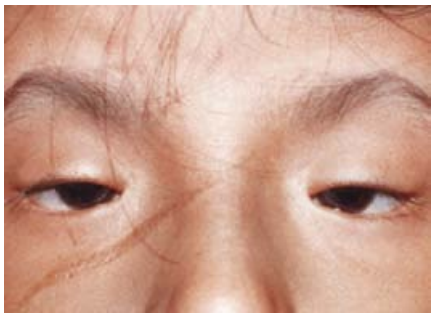


Fig. 1.2.4A: Telecanthus—pseudoconvergent squint

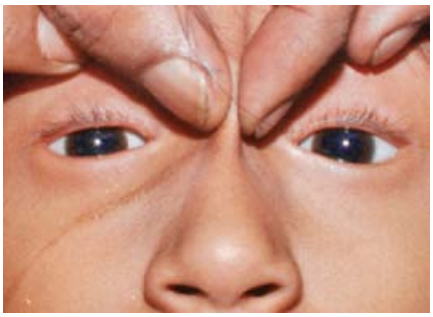


Fig. 1.2.4B: Telecathus—pseudoconvergent squint—correction after pinch test

Coloboma of the Eyelid

- A notch or defect of the lid margin
- Unilateral **(Fig. 1.3.1)** or bilateral **(Fig. 1.3.2)**; upper or lower
- Upper lid coloboma: At the junction of middle and inner thirds
- May be associated with Goldenhar syndrome **(Figs 1.3.3 and 1.3.4)**
- Lower lid coloboma: At middle and outer thirds junction **(Fig. 1.3.5)**
- Associated with Treacher Collins syndrome
- May be acquired in traumatic cases **(Figs 1.3.6 and 1.3.7)**
- *Treatment:* Urgent plastic repair at a very early age to prevent exposure keratitis and corneal ulcer.

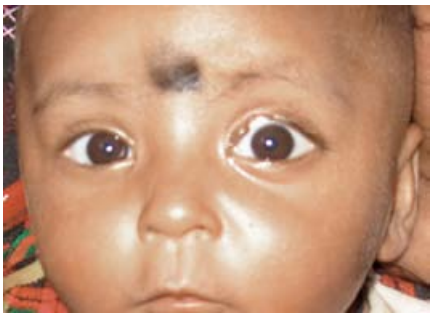


Fig. 1.3.1: Coloboma left upper eyelid

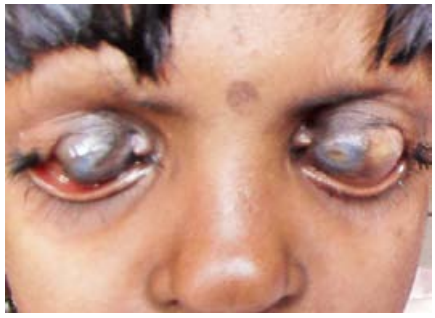


Fig. 1.3.2: Bilateral upper lid coloboma

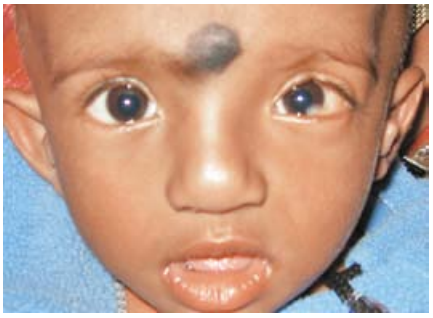


Fig. 1.3.3: Coloboma left upper lid, limbal dermoid and accessory auricle—Goldenhar syndrome

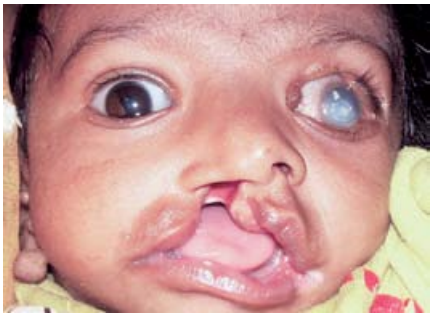


Fig. 1.3.4: Coloboma left upper lid with corneal ulcer and cleft palate in Goldenhar syndrome

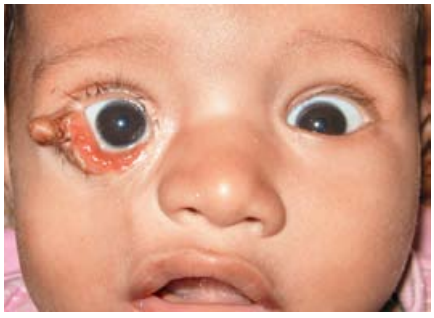


Fig. 1.3.5: Congenital coloboma—right lower lid

Fig. 1.3.6: Acquired coloboma of left upper lid

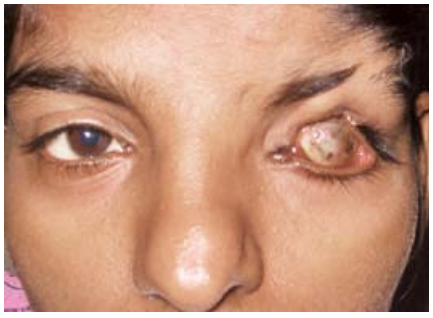


Fig. 1.3.7: Coloboma lid with symblepharon following trauma

Distichiasis

- Hereditary and congenital condition
- Extra posterior row of cilia, occasionally present in all four lids
- Partial **(Fig. 1.4.1)** or complete **(Fig. 1.4.2)**
- They occupy the position of meibomian gland orifices
- Eyelashes may irritate cause to corneal epithelial defects
- May be also seen in Stevens Johnson's syndrome—acquired distichiasis **(Fig. 1.4.3)**
- *Treatment:* By cryotherapy or excision with grafting.

Fig. 1.4.1: Distichiasis of both lids

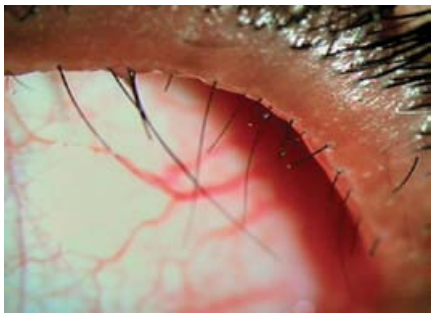


Fig. 1.4.2: Distichiasis of upper lid

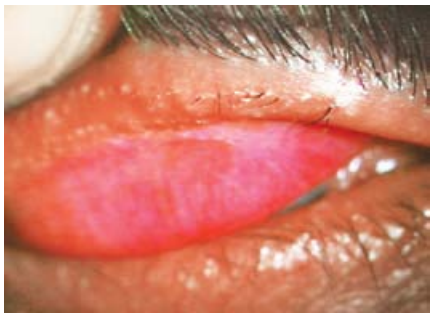


Fig. 1.4.3: Acquired distichiasis in Stevens Johnson syndrome

Blepharophimosis Syndrome

- Autosomal dominant **(Figs 1.5.1 and 1.5.2)**
- Syndrome consists of bilateral **(Fig. 1.5.3)**
  - Narrowing of vertical and horizontal palperbral apertures
  - Telecanthus
  - Inverse epicanthus folds
  - Lateral ectropion and moderate to severe ptosis
- May be asymmetrical **(Fig. 1.5.4)** and without epicanthic folds **(Fig. 1.5.5)**
- *Treatment:* Plastic reconstruction of lids, along with bilateral brow suspension for ptosis.

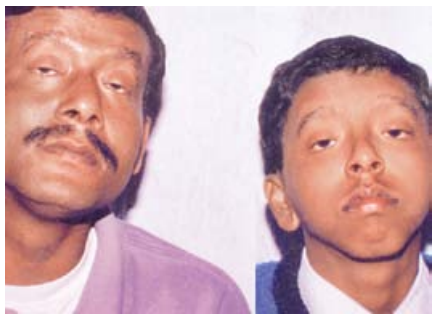


Fig. 1.5.1: Blepharophimosis syndrome—father and son

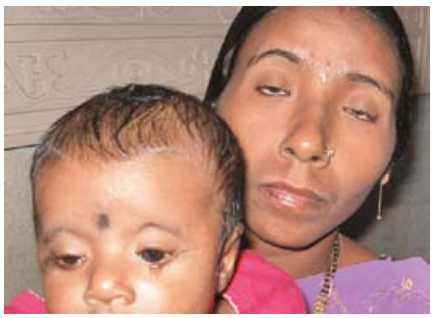


Fig. 1.5.2: Blepharophimosis syndrome—mother and daughter

Fig. 1.5.3: Blepharophimosis syndrome

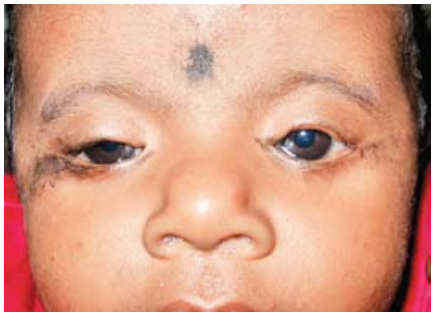


Fig. 1.5.4: Blepharophimosis syndrome—asymmetrical

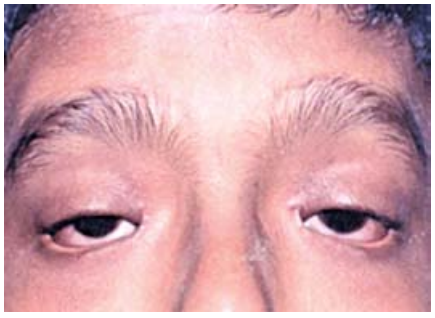


Fig. 1.5.5: Blepharophimosis syndrome—without epicanthic folds

Epiblepharon

- Extra fold of skin in the lower lid with inturning of eyelashes **(Fig. 1.6.1)**
- Nasal 1/3rd is most commonly affected
- *Treatment*: Plastic repair if necessary to prevent recurrent infection.

Fig. 1.6.1: Epiblepharon

Euryblepharon

Rare, congenital, bilateral, not so serious condition

- Palpebral apertures are larger than normal **(Fig. 1.6.2A)** and may be with epicanthus **(Fig. 1.6.2B)**
- Excessive watering may be a problem due to more exposure
- *Treatment*: No treatment for most of the cases; lateral tarsorrhaphy for symptomatic cases.

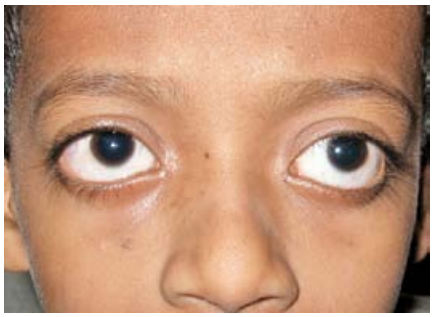


Fig. 1.6.2A: Euryblepharon

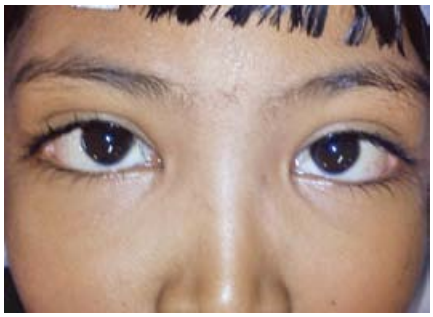


Fig. 1.6.2B: Euryblepharon with epicanthus

Eyelash Abnormalities

Trichiasis

- Inward misdirection of eyelash(es) which irritate the cornea and/or conjunctiva (**Figs 1.7.1 and 1.7.2)**
- It causes punctate epithelial erosion (PEE) (**Figs 1.7.3A and B**) or may cause frank corneal ulcer (**Fig. 1.7.4**)
- When associated with entropion—called *pseudo-trichiasis* (**Fig. 1.7.5**)
- *Treatment:* Temporarily by epilation; permanently by electrolysis, cryotherapy, or argon laser cilia ablation. *If more cilia are involved:* Operative procedure as entropion is most effective.

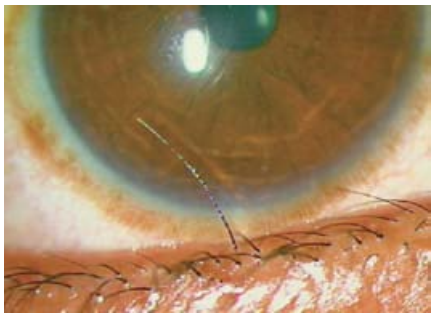


Fig. 1.7.1: Trichiasis

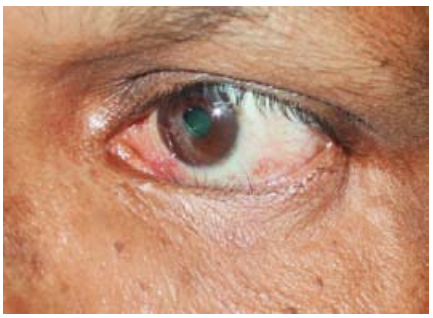


Fig. 1.7.2: Trichiasis—multiple lashes

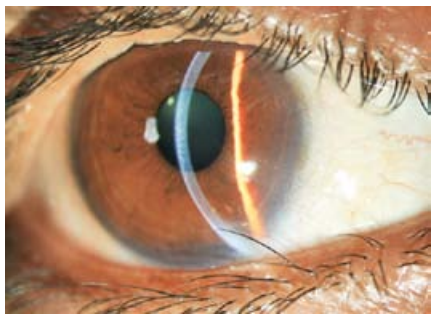


Fig. 1.7.3A: Trichiasis with PEE

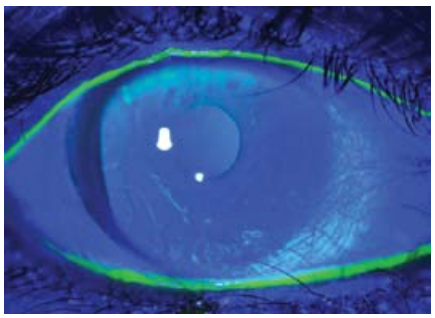


Fig. 1.7.3B: Trichiasis with PEE—after fluorescein stain

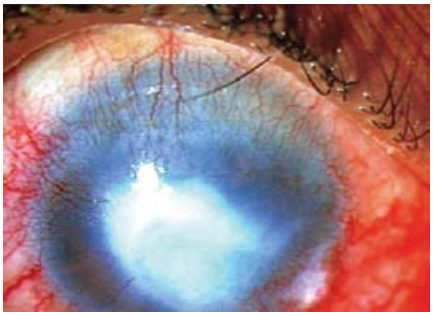


Fig. 1.7.4: Trichiasis with corneal ulcer

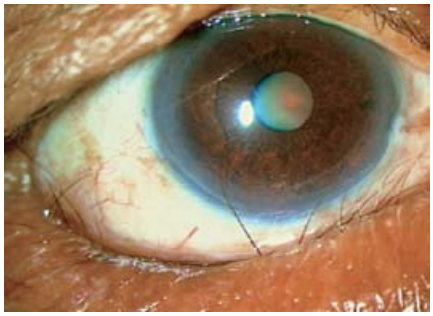


Fig. 1.7.5: Pseudotrichiasis in entropion

Eyelash Ptosis

- Downward drooping of upper lid eyelashes (**Fig. 1.8.1**)
- Congenital or may be seen after prolong use of latanoprost eye drop
- *It does not require any treatment.*

Fig. 1.8.1: Eyelash ptosis

Lash in the Punctum

- An uncommon phenomenon which may cause a pricking sensation on blinking
- Mostly seen in lower punctum (**Fig. 1.9.1**), may be in the upper punctum (**Fig. 1.9.2**)
- *Treatment:* Simple removal of the offending eyelash.

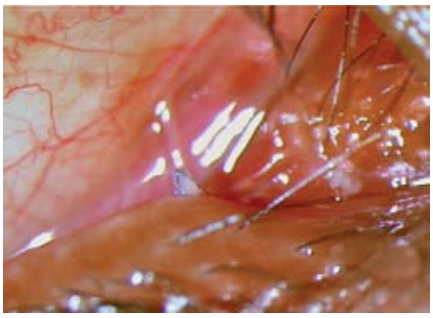


Fig. 1.9.1: Eyelash in lower punctum

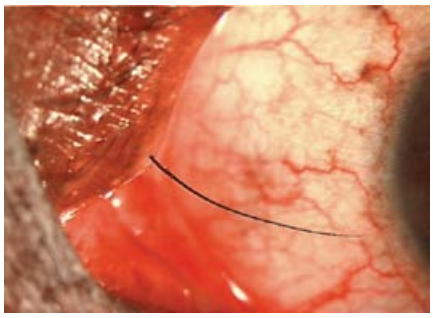


Fig. 1.9.2: Eyelash in upper punctum

Eyelash in the Anterior Chamber

- Rare occurrence after an open globe injury (**Fig. 1.10.1**)
- May cause iridocyclitis or implantation cyst.

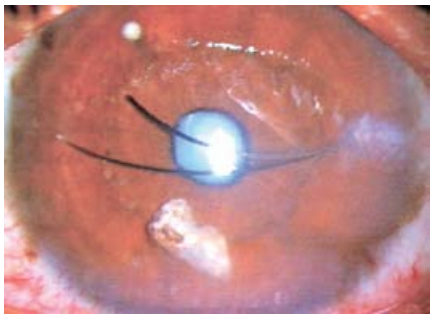


Fig. 1.10.1: Eyelash in anterior chamber

Metaplastic Eyelash(es)

- Not so rare situation which may cause redness and irritation (**Fig. 1.11.1**)
- May be seen in cicatricial lid condition like chemical burn, Stevens-Johnson (SJ) syndrome, ocular cicatricial pemphigoid or raditional injury
- Can also cause corneal ulcer (**Fig. 1.11.2**)
- *Treatment:* Just pull the eyelash to remove it.

Fig. 1.11.1: Metaplastic eyelash

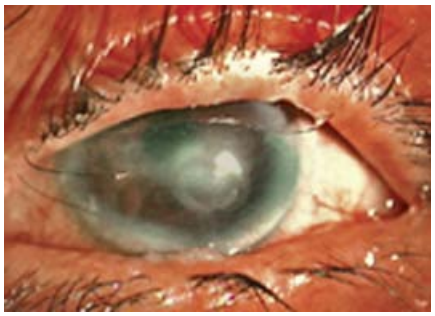


Fig. 1.11.2: Metaplastic eyelash with healed ulcer

Poliosis

- Whitening of eyelashes; partial or total, unilateral or bilateral
- *Causes:*
  - Aging (**Fig. 1.12.1**)
  - Albinism (**Fig. 1.12.2**)
  - Sympathetic ophthalmia
  - Vogt-Koyanagi-Harada (VKH) syndrome (**Figs 1.12.3A and B**)
  - Waardenburg syndrome
  - Idiopathic (**Figs 1.12.4A and B**)
- *Treatment:* No specific treatment.

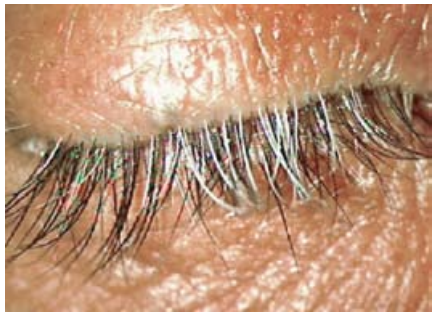


Fig. 1.12.1: Poliosis—aging

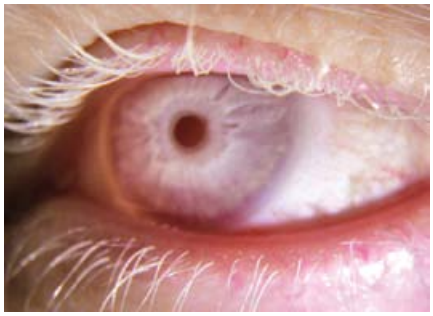


Fig. 1.12.2: Poliosis—albinism

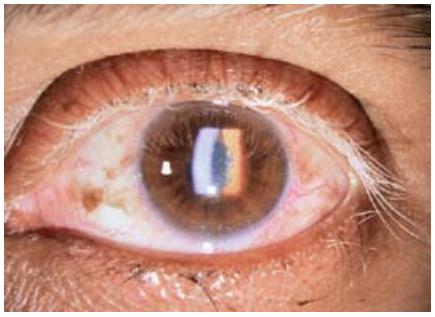


Fig. 1.12.3A: Poliosis—VKH syndrome

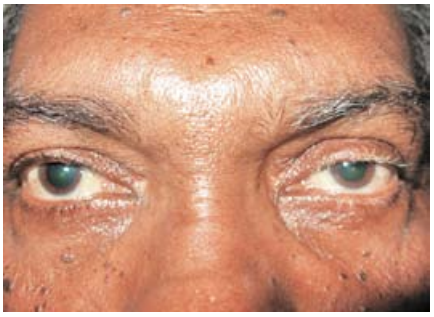


Fig. 1.12.3B: Poliosis—VKH syndrome without anterior uveitis

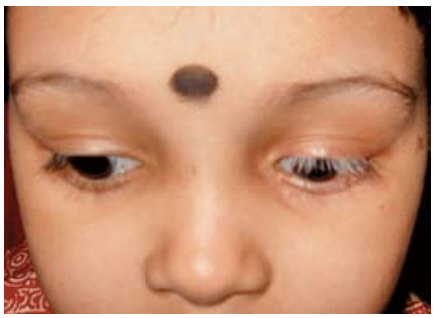


Fig. 1.12.4A: Poliosis—idiopathic unilateral

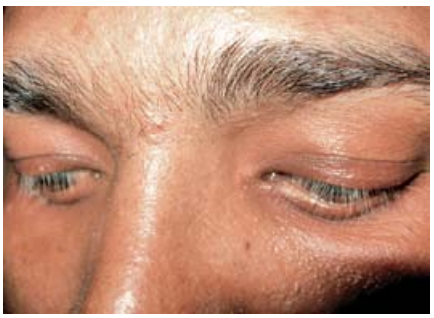


Fig. 1.12.4B: Poliosis—idiopathic bilateral

Madarosis

- Partial or complete loss of eyelashes
- *Causes:*
  - Chronic blepharitis (**Fig. 1.13.1**)
  - Burns
  - Post herpes zoster (HZO) (**Fig. 1.13.2**)
  - Leprosy (**Fig. 1.13.3**)
  - Trichotillomania
  - Generalized alopecia (**Figs 1.13.4A and B**)
  - Myxedema
- *Treatment* of the cause.

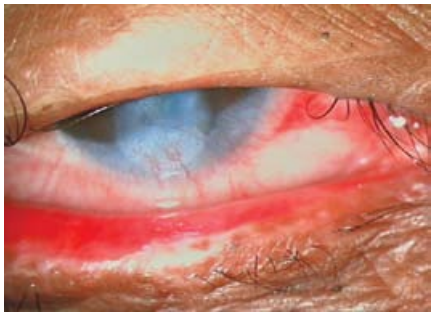


Fig. 1.13.1: Madarosis—blepharitis

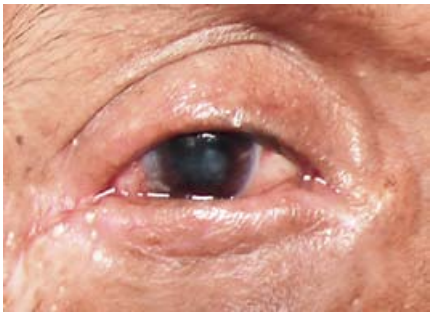


Fig. 1.13.2: Madarosis—HZO

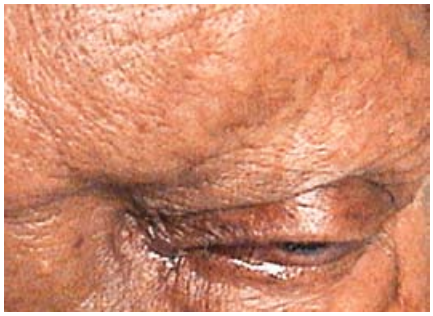


Fig. 1.13.3: Madarosis—leprosy

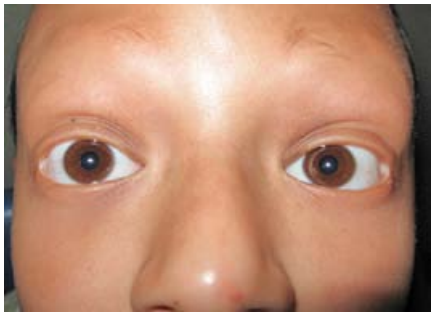


Fig. 1.13.4A: Madarosis—generalized alopecia

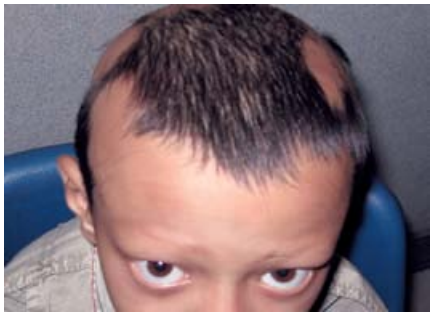


Fig. 1.13.4B: Madarosis—generalized alopecia

Brittle Eyelash

- Rare, bilateral condition
- Eyelashes break with simple rubbing or with slightest manipulation (**Figs 1.14.1 and 1.14.2**)
- Associated with congenital ectodermal dysplasia.

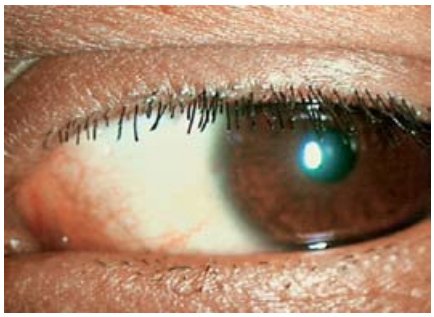


Fig. 1.14.1: Brittle eyelash

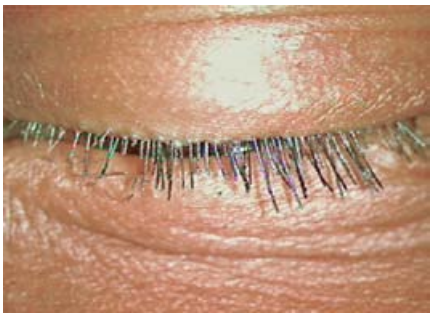


Fig. 1.14.2: Brittle eyelash

Trichomegaly

- Excessively long and dense luxuriant eyelashes (**Figs 1.15.1 and 1.15.2**)
- May be associated with newer antiglaucoma medication, like brimatoprost
- *No treatment* is required.

Fig. 1.15.1: Trichomegaly

Fig. 1.15.2: Trichomegaly

Matting of Eyelashes

- Few eyelashes are stuck together
- Mostly seen in upper eyelids
- *Causes:*
  - Acute conjunctivitis **(Fig. 1.16.1)**
  - Blepharitis **(Fig. 1.16.2)**
  - Bacterial corneal ulcer (**Fig. 1.16.3**) or panophthalmitis (**Fig. 1.16.4**)
  - Use of eye ointment
- *Treatment* of the cause.

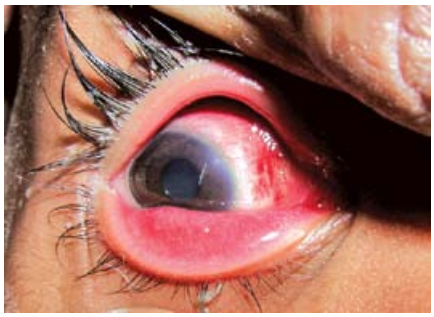


Fig. 1.16.1: Matting of lashes—conjunctivitis

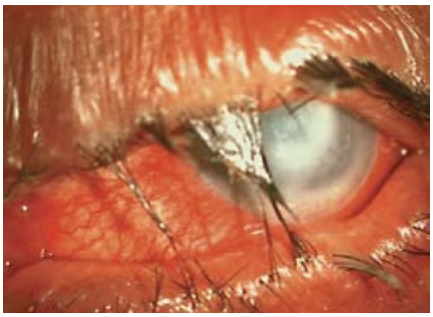


Fig. 1.16.2: Matting of lashes—blepharitis

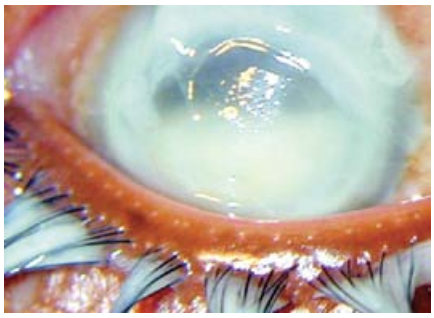


Fig. 1.16.3: Matting—bacterial corneal ulcer

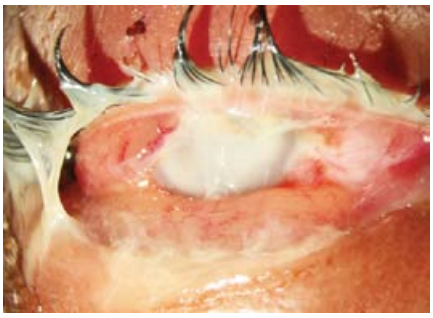


Fig. 1.16.4: Matting of eyelashes—panophthalmitis

Entropion

Inward turning of the eyelids towards the globe.

Congenital Entropion

- Rare, may be associated with microphthalmos/anophthalmos
- Medial 1/3rd is commonly involved (**Figs 1.17.1 and 1.17.2**)
- May be associated with epiblepharon (**Fig. 1.17.3**)
- *Treatment:* Excess skin may be removed with resection of tarsus.

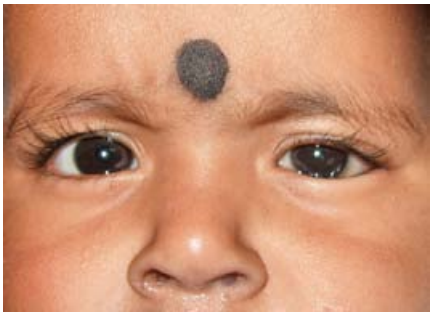


Fig. 1.17.1: Congenital entropion—left lower lid

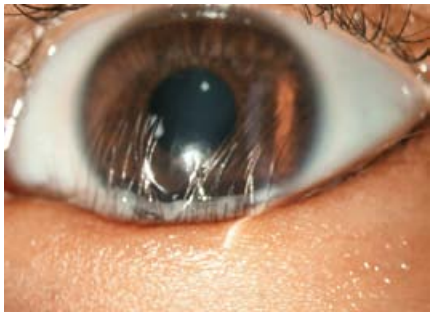


Fig. 1.17.2: Congenital entropion

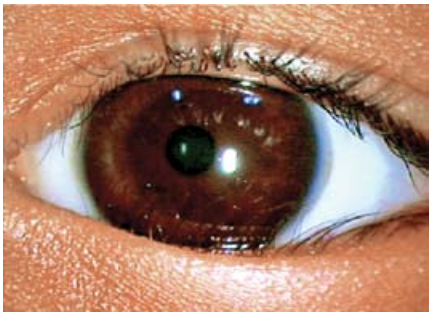


Fig. 1.17.3: Congenital entropion with epiblepharon

Involutional (Senile) Entropion

- Involutional or senile entropion is most common and affects the lower lid only (**Fig. 1.18.1**). Can be easily corrected by simple digital pressure
- May be unilateral or bilateral (**Figs 1.18.2A and B**)
- Very rearly in upper lid alone (**Figs 1.18.3A and B**) or in both eyelids (**Fig. 1.18.4**)
- It is caused by horizontal lid laxity and over-riding of preseptal part of orbicularis
- *Treatment:* Temporary—adhesive tape, cautery, transverse lid everting suture, etc.
  Permanent: Weis' procedure, horizontal lid shortening, tucking of inferior lid retractors, etc.

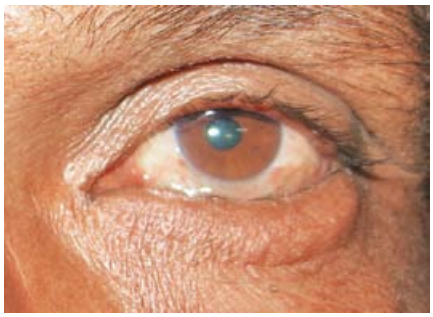


Fig. 1.18.1: Senile entropion

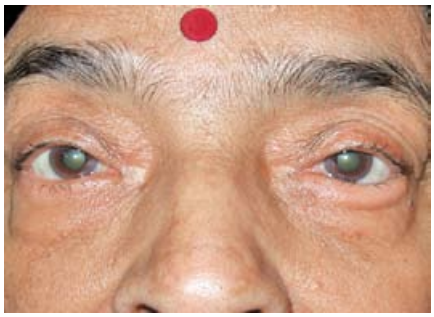


Fig. 1.18.2A: Senile entropion—left lower lid

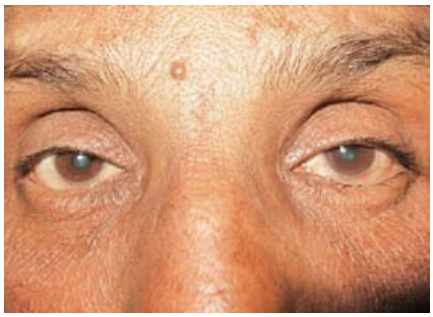


Fig. 1.18.2B: Senile entropion—both lower lids

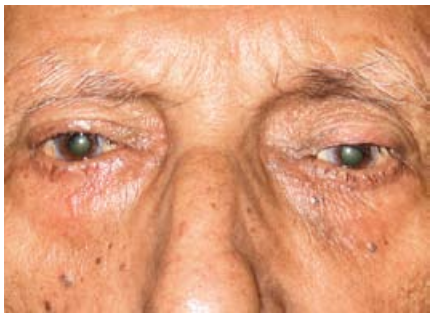


Fig. 1.18.3A: Senile entropion—left upper lid

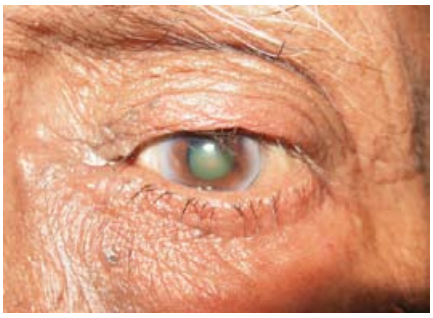


Fig. 1.18.3B: Senile entropion—left upper lid

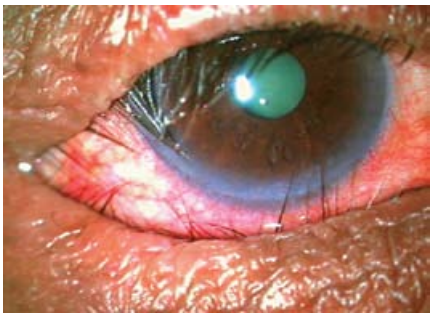


Fig. 1.18.4: Senile entropion—both lids—same eye

Cicatricial Entropion

- Due to scarring of the palpebral conjunctiva
- It usually affects the upper lid (**Fig. 1.19.1**) and cannot be corrected by digital manipulation
- *Causes:* Chemical burn (**Fig. 1.19.2**), trachoma, Stevens Johnson syndrome (**Fig. 1.19.3**), ocular pemphigoid, etc.
- *Treatment:* Tarsal wedge resection, tarsal fracture, etc.

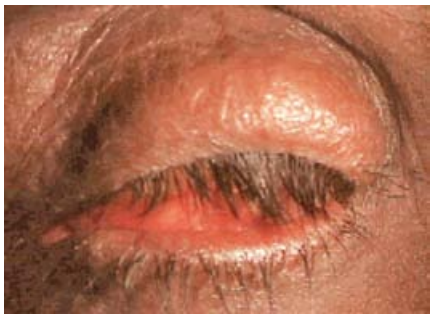


Fig. 1.19.1: Cicatricial entropion—trachoma

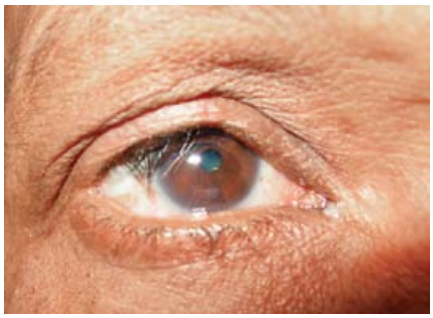


Fig. 1.19.2: Cicatricial entropion—upper lid—chemical injury

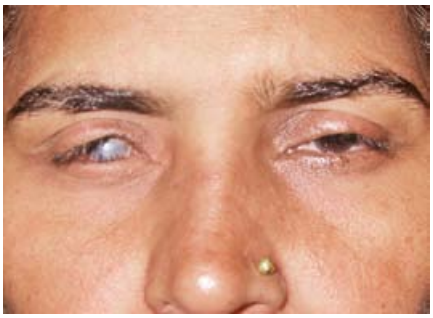


Fig. 1.19.3: Cicatricial entropion—both upper and right lower

Acute Spastic Entropion

- Associated with blepharospasm, mainly affects the lower lids (**Fig. 1.20.1**)
- *Causes:* Chronic conjunctivitis, keratitis, corneal abrasion (**Fig. 1.20.2**) and postoperative
- *Treatment*: Adhesive tape and removal of the cause.

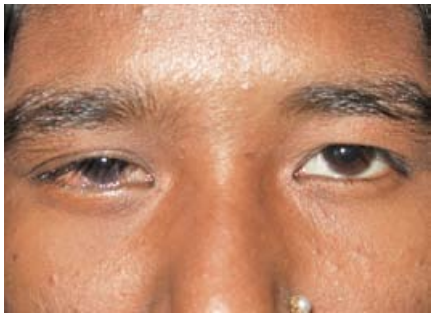


Fig. 1.20.1: Spastic entropion

Fig. 1.20.2: Spastic entropion—right lower lid after corneal abrasion

Ectropion

Outward turning of the eyelid away from the globe.

Involutional (Senile) Ectropion

- Age-related condition which affects the lower lid (**Fig. 1.21.1**)
- It is the most common form—may be unilateral (**Fig. 1.21.2**) or bilateral (**Fig. 1.21.3**)
- Due to excessive horizontal eyelid length with weakness of the preseptal part of orbicularis. Laxity of medial canthal tendon is marked
- *Treatment:* Medial conjunctivoplasty, Bick's procedure, horizontal lid shortening, etc.

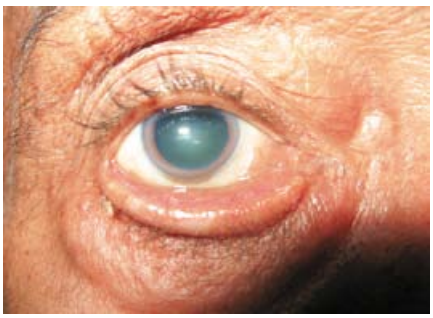


Fig. 1.21.1: Senile ectropion

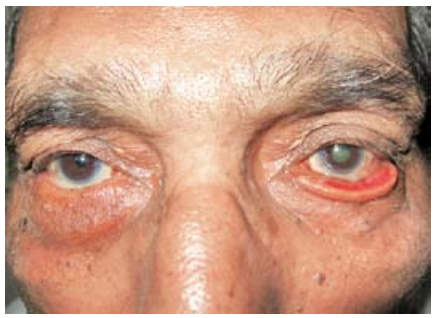


Fig. 1.21.2: Senile ectropion—unilateral

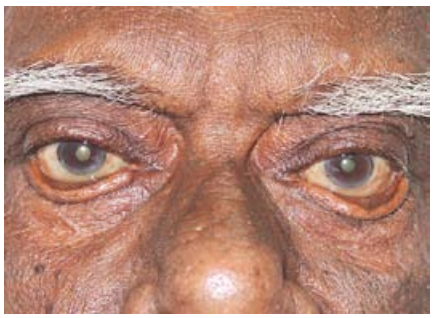


Fig. 1.21.3: Senile ectropion—bilateral

Cicatricial Ectropion

- Contracture of the skin and underlying tissues of the lower eyelids
- May be unilateral or bilateral (**Figs 1.22.1 to 1.22.3**)
- *Causes*: Chemical (**Fig. 1.22.4**) or thermal burn (**Fig. 1.22.5**), trauma/lacerated injury (**Fig. 1.22.6**) or skin disorders (**Fig. 1.22.7**)

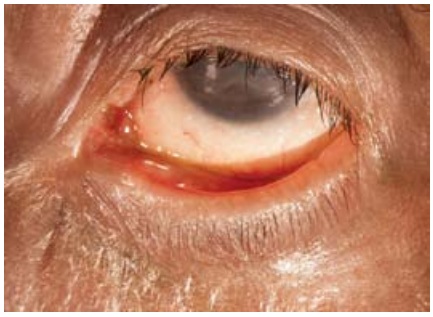


Fig. 1.22.1: Cicatricial ectropion—following contact dermatitis

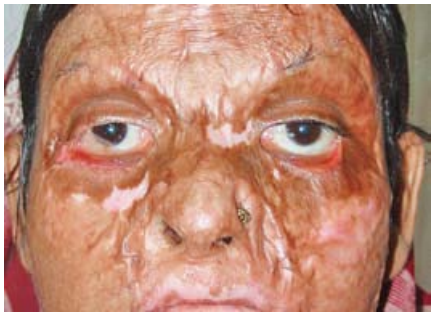


Fig. 1.22.2: Cicatricial ectropion both lower lids following acid burn

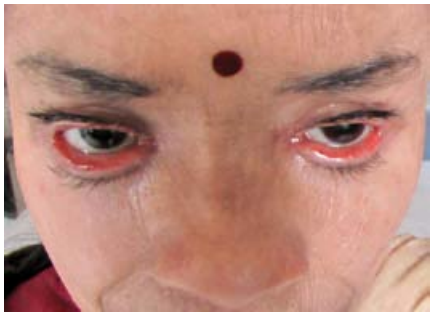


Fig. 1.22.3: Cicatricial ectropion of all four lids—scleroderma

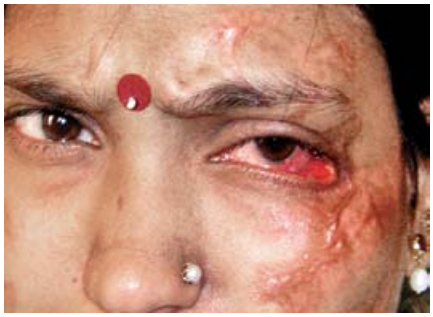


Fig. 1.22.4: Cicatricial ectropion—chemical injury

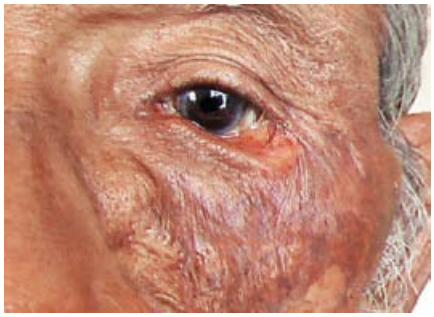


Fig. 1.22.5: Cicatricial ectropion—thermal injury

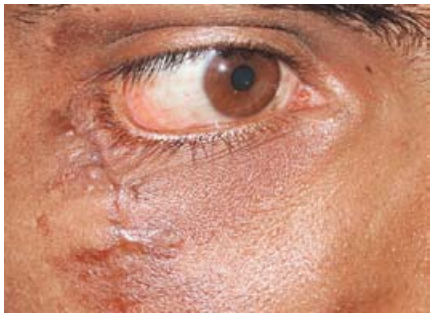


Fig. 1.22.6: Cicatricial ectropion—trauma

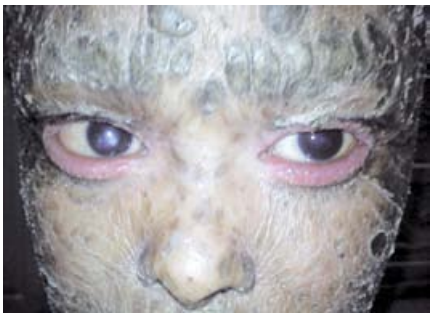


Fig. 1.22.7: Cicatricial ectropion—all lids in ichthyosis vulgaris

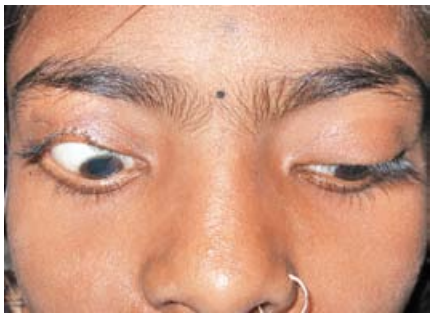


Fig. 1.22.8: Cicatricial ectropion—upper lid following trauma

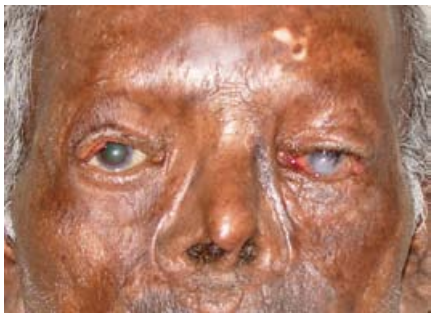


Fig. 1.22.9: Cicatricial ectropion—upper lid following acid burn

- Depending upon the nature of the contracture, upper eyelids may also be affected (**Figs 1.22.8 and 1.22.9**)
- *Treatment:*
  - Excision of the scar with a skin graft to the raw area (**Figs 1.22.10A and B**)
  - Lengthening of vertical shortening by Z-plasty.

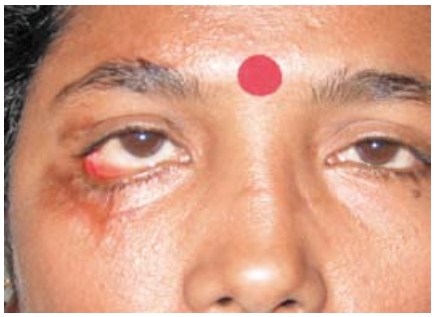


Fig. 1.22.10A: Cicatricial ectropion—before correction

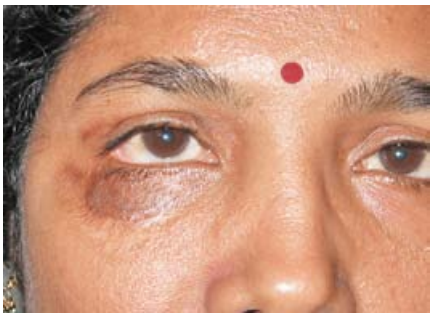


Fig. 1.22.10B: Cicatricial ectropion—after correction **(Fig. 1.22.9A)**

Paralytic Ectropion

- Caused by paralysis of orbicularis and associated with lagophthalmos (**Fig. 1.23.1**)
- Incomplete blinking and epiphora
- May cause corneal ulcer in severe cases (**Fig. 1.23.2**)
- *Treatment:*
  - *Mild cases:* Tear substitute to prevent corneal drying and antibiotic eye ointment at night
  - *Severe cases*: Lateral tarsorrhaphy, lateral canthoplasty.

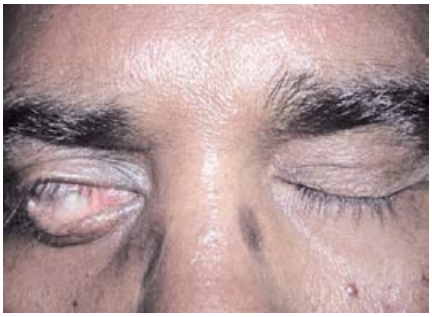


Fig. 1.23.1: Paralytic ectropion

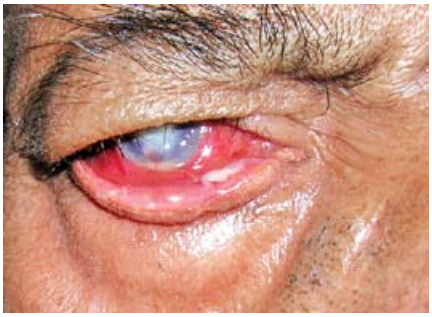


Fig. 1.23.2: Paralytic ectropion with corneal ulcer

Mechanical Ectropion

- It is just a sequel to a swelling of the lower eyelid, e.g. a tumor, lid edema, or a large chalazion (**Figs 1.24.1 and 1.24.2**)
- *Treatment:* Can be corrected by removing the lesion.

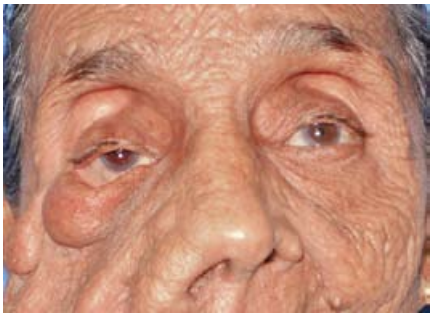


Fig. 1.24.1: Mechanical ectropion—plexiform neurofibroma lower lid

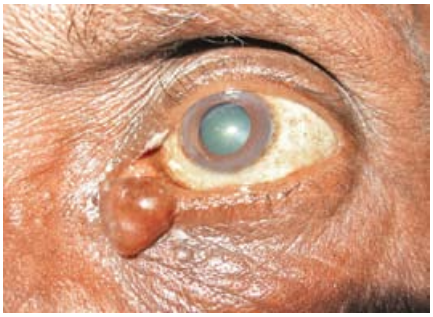


Fig. 1.24.2: Mechanical ectropion at medial third

Inflammatory Lid Conditions

Acute Contact Dermatitis

- Unilateral (**Figs 1.25.1 and 1.25.2**) or bilateral (**Fig. 1.25.3**) condition, caused by sensitivity to topical medication, hair dyes, cosmetics, etc.
- Edema, erythema, vesiculation and later on crusting (**Fig. 1.25.4**)
- *Treatment:* Withdrawal of the irritants, antihistaminics and/or corticosteroids.

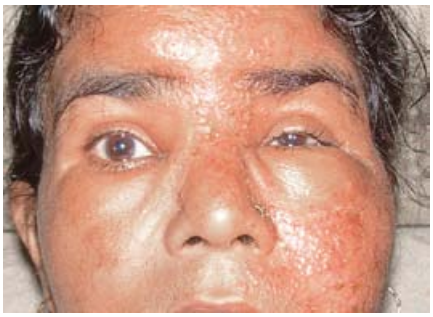


Fig. 1.25.1: Acute contact dermatitis—with neomycin

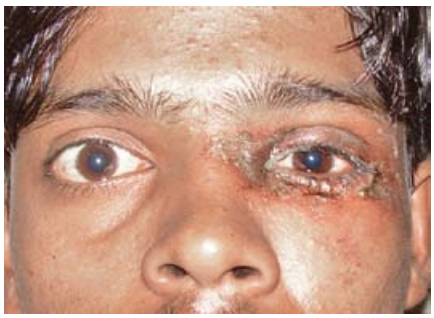


Fig. 1.25.2: Acute contact dermatitis following injury with color in Holi festival

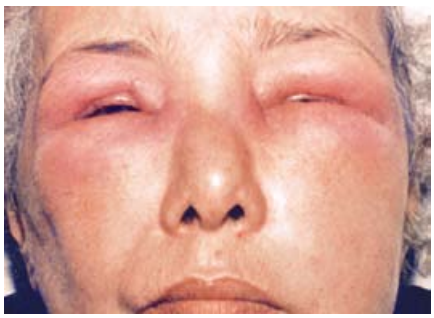


Fig. 1.25.3: Bilateral acute contact dermatitis following hair dye

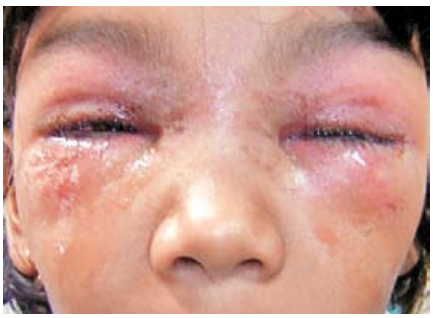


Fig. 1.25.4: Bilateral severe contact dermatitis with atropine

Chronic Contact Dermatitis

- Caused by chronic irritants, e.g. topical medication (**Fig. 1.26.1**), cosmetics, spectacles frame
- Thickening and crusting of the skin (**Fig. 1.26.2**)
- *Treatment:* Identification of the offending agent and rectify it, anti-biotic-steroids ointment.

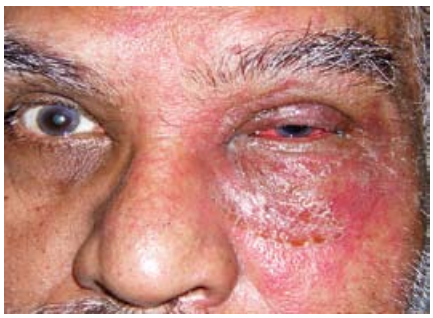


Fig. 1.26.1: Chronic contact dermatitis

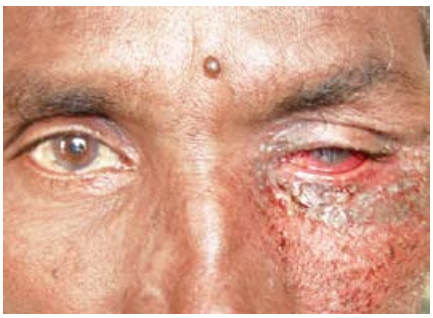


Fig. 1.26.2: Chronic contact dermatitis

Atopic Dermatitis of the Lid

- Uncommon and more generalized with skin condition
- Eyelids are thickened and fissured (**Figs 1.27.1A and B**)
- Associated with atopic kerato-conjunctivitis.

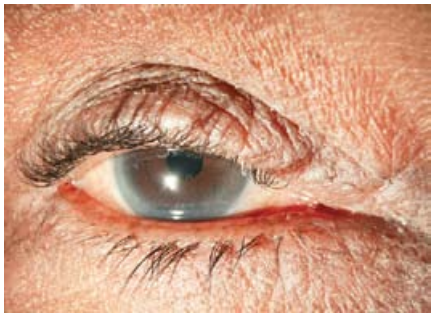


Fig. 1.27.1A: Atopic dermatitis—RE

Fig. 1.27.1B: Atopic dermatitis—LE

Angular Blepharoconjunctivitis

- Unilateral or bilateral infection, caused by *Moraxella*
- Frequently associated with conjunctivitis (**Fig. 1.28.1**)
- Fissuring, maceration, erythema and scaling of one or both canthi (**Fig. 1.28.2**)
- *Treatment:* Oxytetracycline eye ointment and zinc oxide.

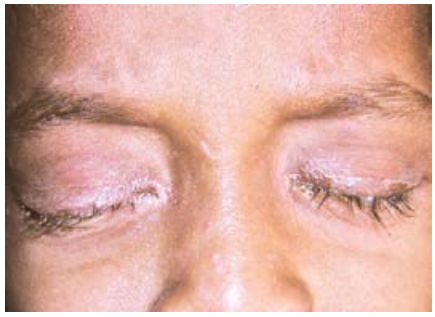


Fig. 1.28.1: Angular blepharoconjunctivitis

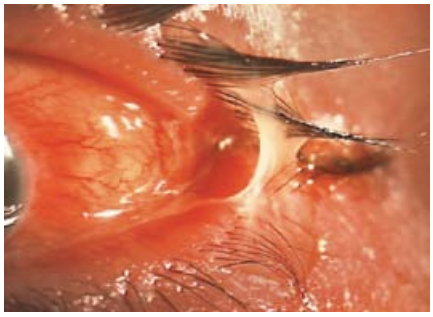


Fig. 1.28.2: Angular blepharoconjunctivitis

Primary Herpes Simplex of Lids

- Uncommon, unilateral condition, may be associated with immune deficiency states
- Crops of small vesicles, ruptures and crust formation, may be with secondary infection (**Figs 1.29.1 and 1.29.2**)
- Healing without scarring by seven days
- May be associated with acute follicular conjunctivitis and dendritic keratitis (**Fig. 1.29.3**)
- *Treatment:* Topical acyclovir ointment.

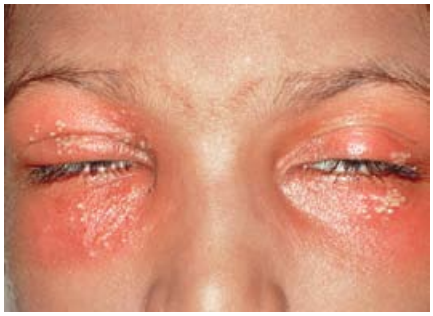


Fig. 1.29.1: Lid lesions in primary herpes simplex virus (HSV)—bilateral

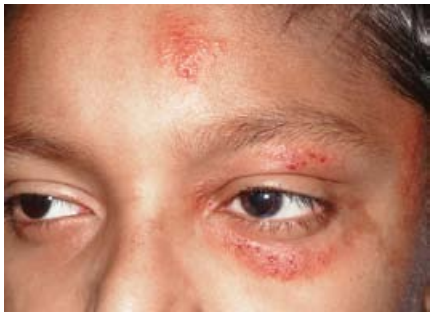


Fig. 1.29.2: Primary herpes simplex of lids

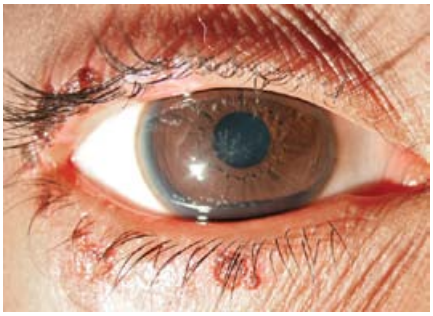


Fig. 1.29.3: Vesicular lid lesions with crust and dendritic keratitis

Herpes Zoster Ophthalmicus

- More common unilateral condition, may be severe in immunocompromised conditions
- Painful maculopapular rash involving the first division of trigeminal nerve with chemosis of lid (**Fig. 1.30.1**)
- Development of vesicles, pustules, and ulceration with crusting (**Fig. 1.30.2**)
- Periorbital edema, secondary infection may lead to bacterial cellulitis
- *Treatment:* High dose of oral acyclovir, investigation to find out the cause of immune deficiency.

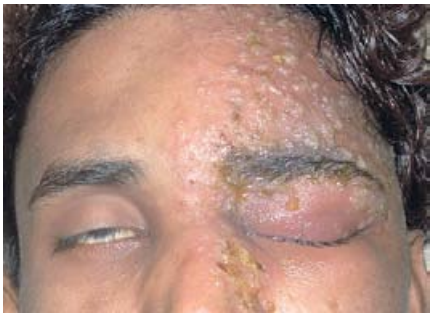


Fig. 1.30.1: Herpes zoster ophthalmicus—fresh lesions

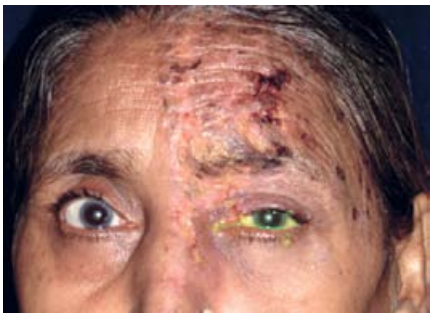


Fig. 1.30.2: Herpes zoster ophthalmicus—healed lesions

Blepharitis

- Sub-acute or chronic inflammation of the eyelids
- Mostly in children and usually bilateral
- Associated with seborrhea (dandruff) of the scalp
- *Squamous blepharitis:*
  - Hyperemia of lid margins
  - White dandruff-like scales on the lid margins (**Figs 1.31.1A and B**)
  - Falling of eyelashes (madarosis)
  - Thickening of the lid margins (tylosis)
- *Ulcerative blepharitis:*
  - Soreness of the lid margins (**Fig. 1.31.2**)
  - Loss of eyelashes
  - Yellow crust at the root of the eyelashes with matting (**Figs 1.31.3 and 1.31.4**)
  - Small ulcers at the base of the crust (**Fig. 1.31.5**)
  - Marginal keratitis is a common association
- *Traetment:* Lid hygiene (lid scrub), vertical lid massage, antibiotic-steroid ointment, systemic tetracycline/doxycycline, treatment of dandruffs, etc.

Fig. 1.31.1A: Squamous blepharitis—RE

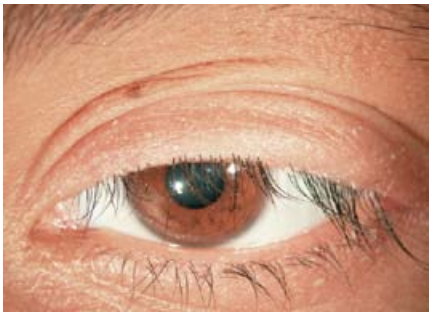


Fig. 1.31.1B: Squamous blepharitis—LE

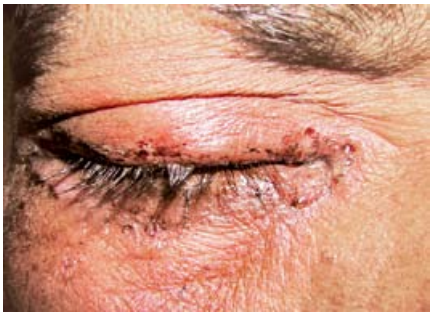


Fig. 1.31.2: Ulcerative blepharitis

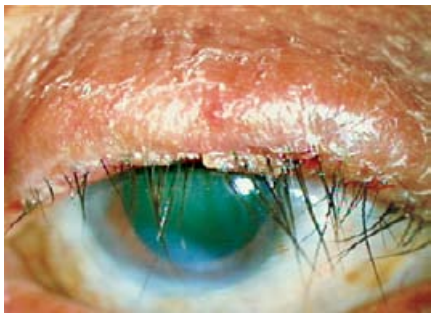


Fig. 1.31.3: Ulcerative blepharitis—madarosis

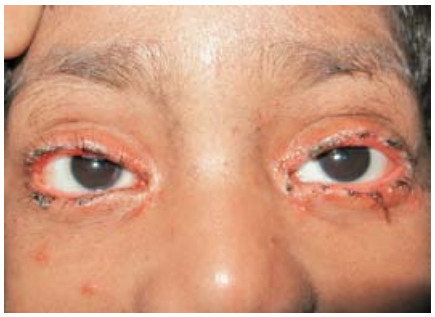


Fig. 1.31.4: Ulcerative blepharitis with crust and madarosis

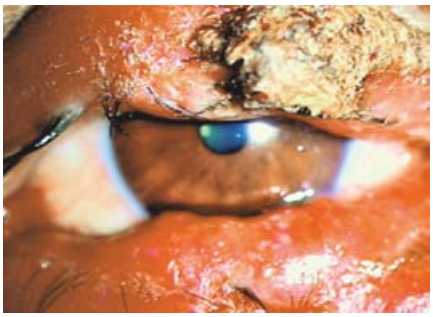


Fig. 1.31.5: Ulcerative blepharitis—crust formation

Meibomianitis

- Chronic infection of the meibomian glands
- Occurs in the middle age
- White, frothy secretion on the eyelid margins and at the outer canthus (seborrhea) (**Fig. 1.32.1**)
- Plugged duct opening (**Fig. 1.32.2A)** and vertical yellowish streaks shining through the conjunctiva (**Fig. 1.32.2B)**
- Blocked meibomian ducts (**Figs 1.32.3 and 1.32.4**)
- Thick secretion on expression ('toothpaste sign') (**Fig. 1.32.5**)
- Recurrent blepharoconjunctivitis (**Fig. 1.32.6**) and marginal keratitis (**Fig. 1.32.7**) may be a common association
- *Treatment:* Tarsal (vertical lid) massage, steroid-antibiotic ointment, systemic doxycycline, tears substitutes, etc.

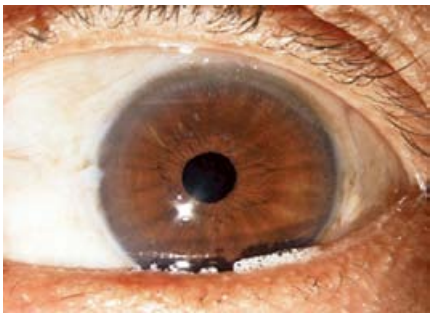


Fig. 1.32.1: Meibomian seborrhea

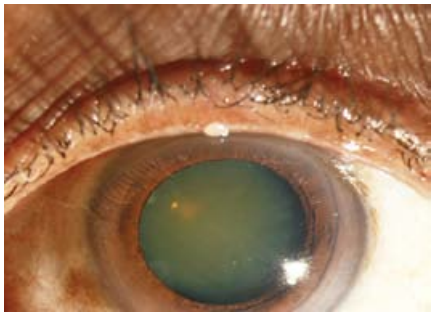


Fig. 1.32.2A: Meibomianitis—plugging of the meibomian duct opening

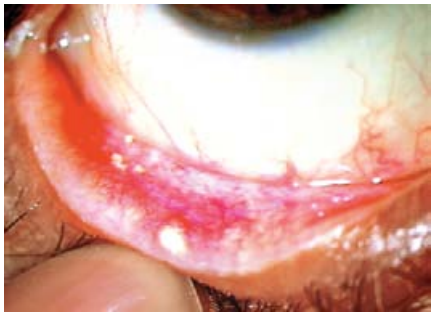


Fig. 1.32.2B: Meibomianitis—vertical streak

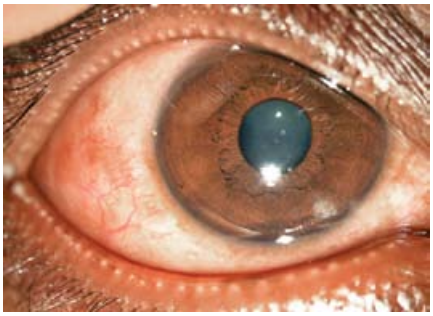


Fig. 1.32.3: Meibomianitis—blocked ducts

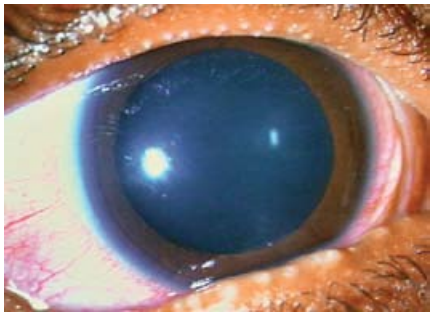


Fig. 1.32.4: Meibomianitis—blocked ducts

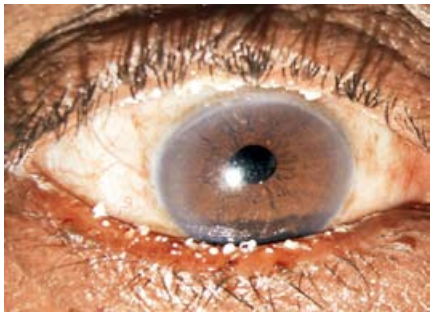


Fig. 1.32.5: Meibomianitis—toothpaste sign

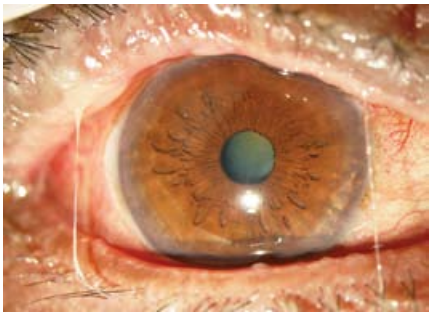


Fig. 1.32.6: Meibomianitis—blepharoconjunctivitis

Preseptal Cellulitis

- Unilateral erythema and edema with tenderness involving the upper eyelid (**Figs 1.33.1 and 1.33.2**)
- May lead to lid abscess (**Fig. 1.33.3**) and pyogenic granuloma formation (**Figs 1.33.4A and B**)
- *Differential diagnosis with orbital cellulitis* (**Fig. 1.33.5**)
  - No proptosis
  - Normal visual acuity, ocular movement and pupillary reactions
- *Treatment:* Systemic antibiotics, analgesics, hot compress and topical antibiotics.

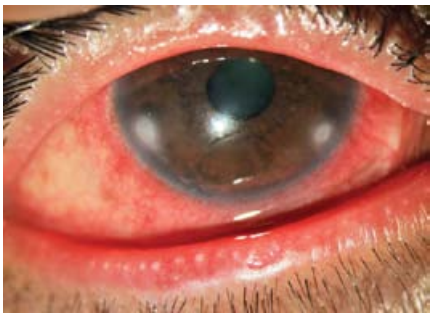


Fig. 1.32.7: Meibomianitis with marginal keratitis

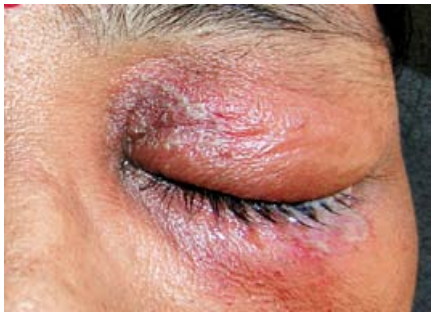


Fig. 1.33.1: Preseptal cellulitis

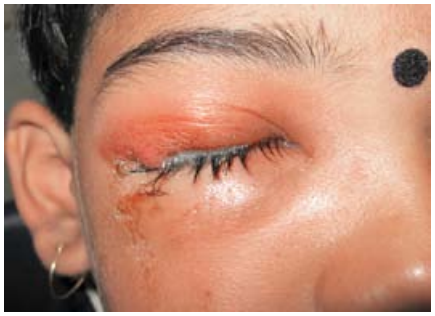


Fig. 1.33.2: Preseptal cellulitis

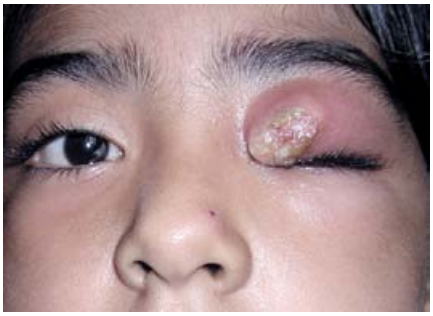


Fig. 1.33.3: Lid abscess

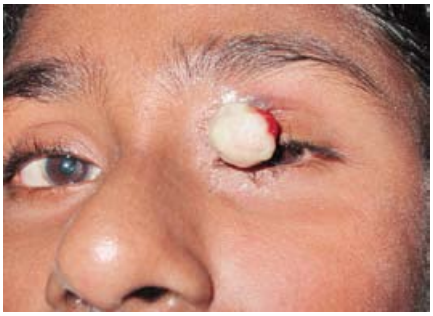


Fig. 1.33.4A: Lid abscess—pyogenic granuloma

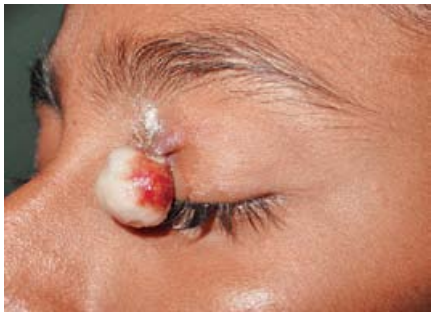


Fig. 1.33.4B: Lid abscess—pyogenic granuloma

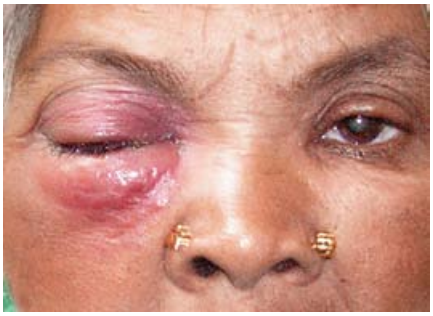


Fig. 1.33.5: Orbial cellulitis

External Hordeolum (Stye)

- Acute suppurative inflammation of the follicle of an eyelash
- A swollen area at the lid margin **(Fig. 1.34.1)**
- A whitish, round, raised pus point at eyelash root **(Fig. 1.34.2)**
- May be associated with preseptal cellulitis and conjunctival chemosis **(Fig. 1.34.3)**
- *Treatment:* Hot compress, systemic analgesics, topical antibiotics, epilation of the offending eyelash.

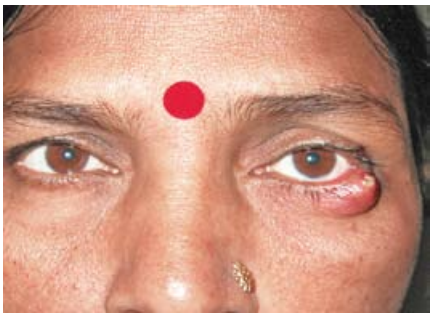


Fig. 1.34.1: External hordeolum—lower lid

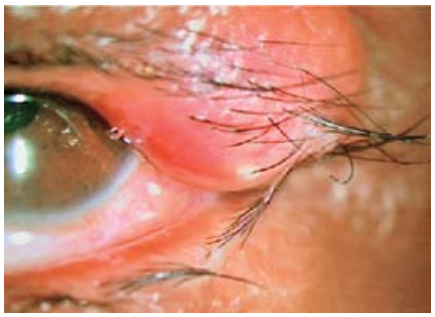


Fig. 1.34.2: External hordeolum—pus point at eyelash root

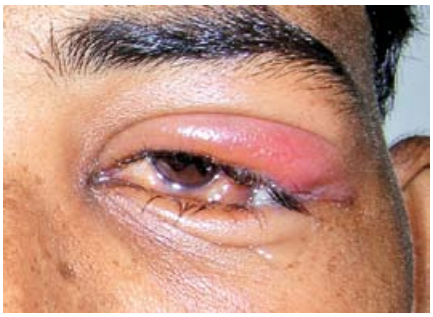


Fig. 1.34.3: External hordeolum with preseptal cellulitis

Internal Hordeolum (Inflamed Chalazion)

- Unilateral acute infection of the meibomian gland
- Tender, diffuse, inflamed swelling within the tarsal plate
- The swelling is away from the lid margin (**Fig. 1.35.1**)
- Pus-point away from the eyelash root (**Fig. 1.35.2**)
- May be associated with preseptal cellulitis
- *Treatment:* Treatment of acute infection followed by incision and curettage of the chalazion later on.

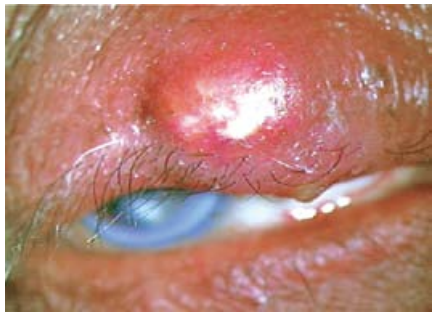


Fig. 1.35.1: Internal hordeolum—pus point at meibomian duct

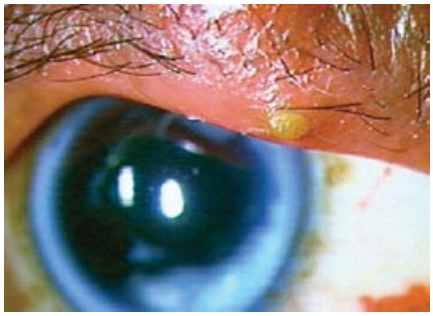


Fig. 1.35.2: Internal hordeolum—pus point away from eyelash

Chalazion

- A chronic nonspecific inflammatory granuloma of the meibomian gland
- Painless nodular swelling of the eyelid (**Fig. 1.36.1A**)
- Tarsal conjunctiva underneath the nodule is velvety red or purple and slightly elevated (**Fig. 1.36.1B**)
- May be single—medium (**Fig. 1.36.2**) or large/giant (**Fig. 1.36.3**) and multiple (**Fig. 1.36.4**)
- It may turn into 'marginal chalazion' (**Fig. 1.36.5**)
- *Treatment:* Steroid-antibiotic ointment for small chalazion and 'incision and curettage' for large one.

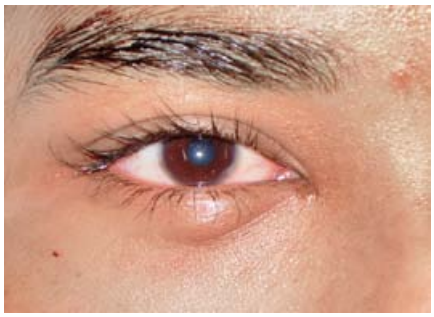


Fig. 1.36.1A: Chalazion RLL—skin surface

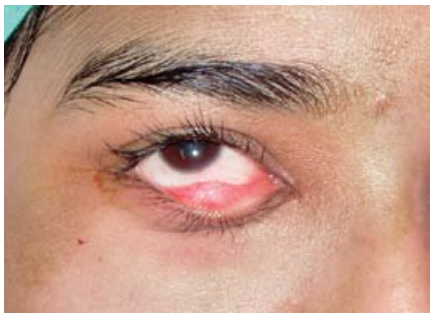


Fig. 1.36.1B: Chalazion RLL—conjunctival surface

Fig. 1.36.2: Chalazion—medium—right lower lid

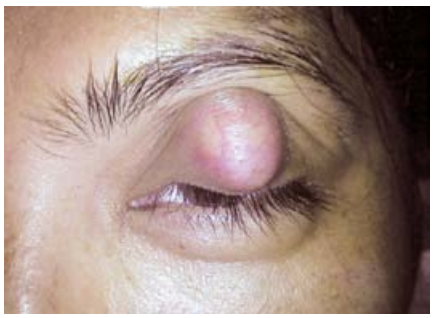


Fig. 1.36.3: Giant chalazion

Fig. 1.36.4: Multiple chalazion of all four lids

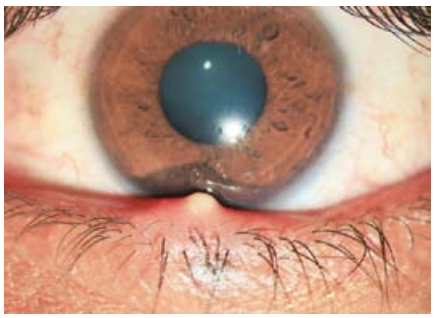


Fig. 1.36.5: Marginal chalazion—lower lid

Ptosis: Drooping of Upper Lid

Ptosis

- Drooping of the upper eyelid
- Unilateral (**Fig. 1.37.1**) or bilateral (**Fig. 1.37.2**), and partial or complete
  - *Mild ptosis:* 2 mm (**Fig. 1.37.3**)
  - *Moderate ptosis:* 3 mm (**Fig. 1.37.4**)
  - *Severe ptosis:* 4 mm or more (**Figs 1.37.5 and 1.37.6**).

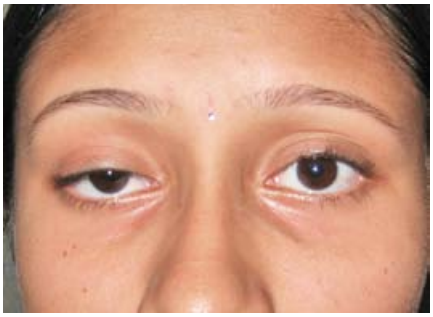


Fig. 1.37.1: Unilateral ptosis

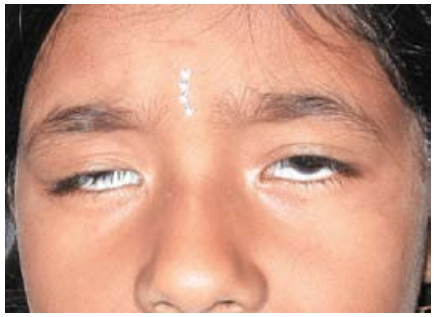


Fig. 1.37.2: Bilateral ptosis

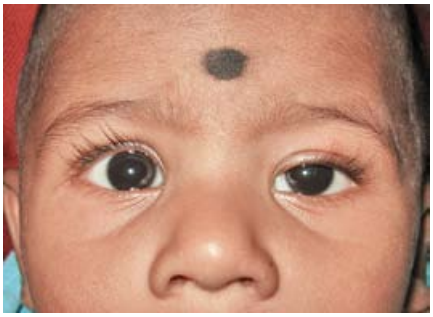


Fig. 1.37.3: Mild ptosis

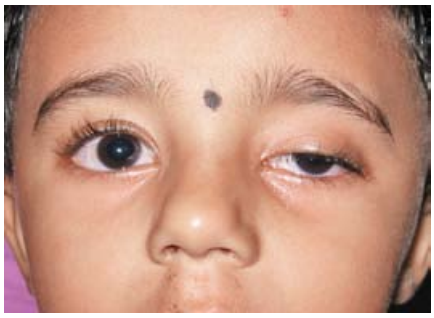


Fig. 1.37.4: Moderate ptosis

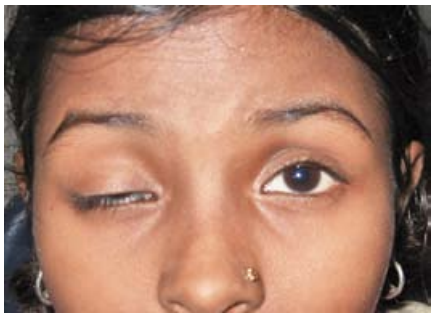


Fig. 1.37.5: Severe ptosis

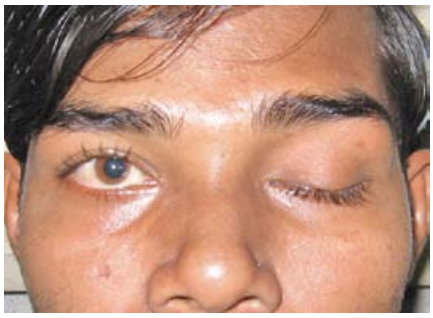


Fig. 1.37.6: Severe complete ptosis

Synkinetic Ptosis

- *Marcus-Gunn jaw winking phenomenon:* Retraction of the ptotic eyelid with ipsilateral jaw movement **(Figs 1.38.1A and B)**
- Sometimes, more ptosis occurs with jaw movement—*a reverse Marcus-Gunn phenomenon* **(Figs 1.38.2A and B)**
- *Misdirected third nerve:* Retraction or ptosis of upper lid with various ocular movement **(Figs 1.38.3A to C)**

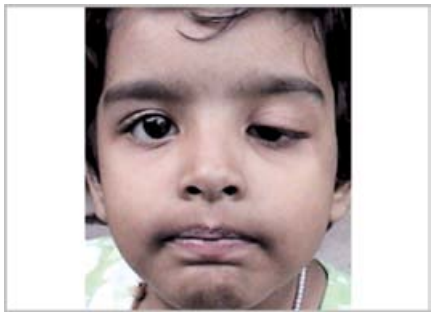


Fig. 1.38.1A: Marcus Gunn phenomenon—ptosis

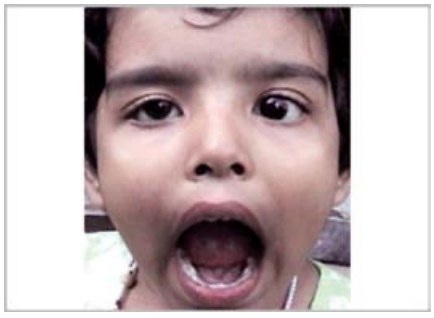


Fig. 1.38.1B: Marcus Gunn phenomenon—ptosis corrected on jaw movement

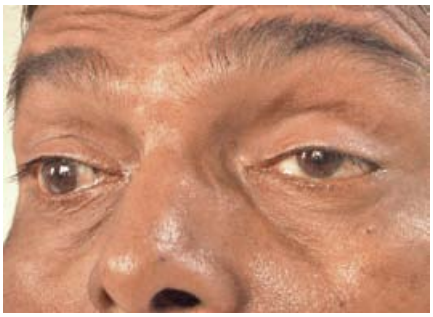


Fig. 1.38.2A: Reverse Marcus-Gunn phenomenon

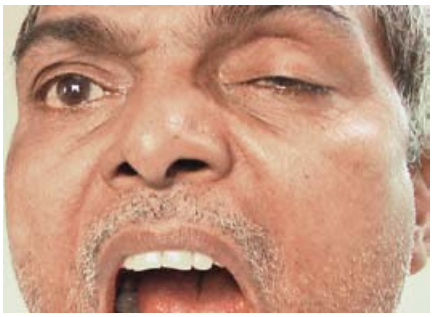


Fig. 1.38.2B: Reverse Marcus-Gunn phenomenon

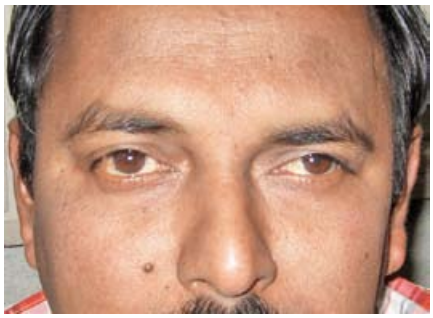


Fig. 1.38.3A: Ptosis—misdirected third nerve

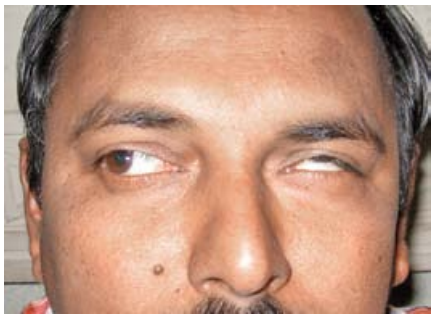


Fig. 1.38.3B: Ptosis—misdirected third nerve

Fig. 1.38.3C: Ptosis—misdirected third nerve

Pseudoptosis

- Anophthalmos **(Fig. 1.39.1)**, microphthalmos or phthisis bulbi **(Fig. 1.39.2)**
- Due to hypotropia **(Fig. 1.39.3)**
- Large conjunctival cyst at the upper limbus **(Figs 1.39.4A and B)**
- Dermatochalasis **(*see* Figs 1.47.2A and B).**

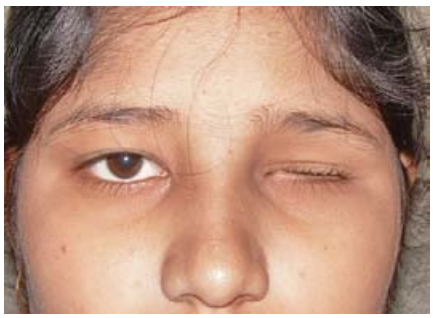


Fig. 1.39.1: Pseudoptosis—anophthalmic socket

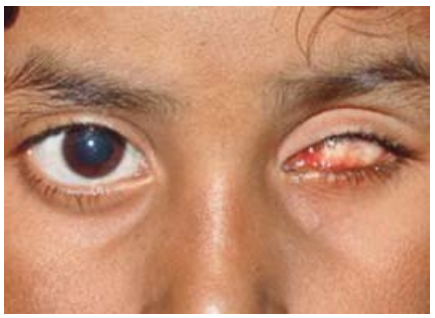


Fig. 1.39.2: Pseudoptosis—phthisis bulbi

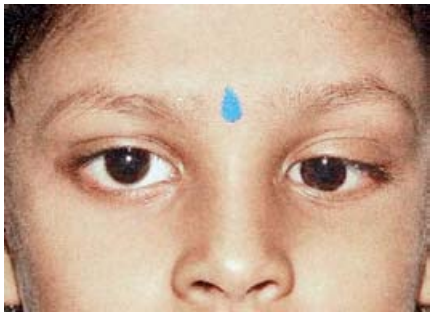


Fig. 1.39.3: Pseudoptosis—left hypotropia

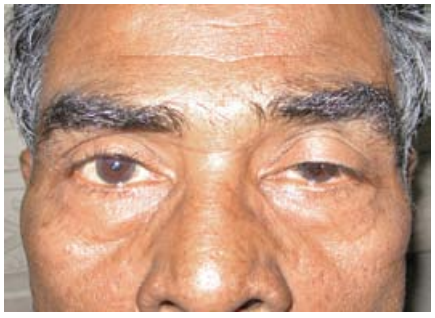


Fig. 1.39.4A: Pseudoptosis—large conjunctival cyst

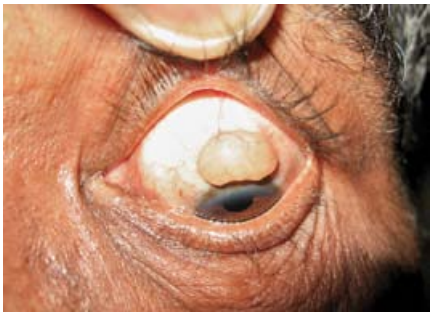


Fig. 1.39.4B: Pseudoptosis—large conjunctival cyst

Congenital Ptosis

- Unilateral or bilateral with varying severity—mild, moderate or severe **(Figs 1.40.1 to 1.40.3)**
- May be simple or with other anomalies like "jaw-winking phenomenon" **(*see* Figs 1.38.1A and B)**
- May be associated with blepharophimosis syndrome **(*see* Fig. 1.5.2)**
- *Treatment:* Depends on severity, early intervention is required in severe ptosis to prevent amblyopia.

Fig. 1.40.1: Congenital unilateral ptosis

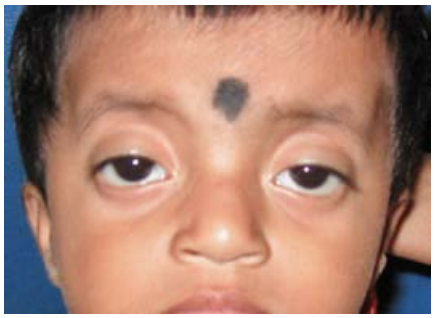


Fig. 1.40.2: Congenital bilateral ptosis

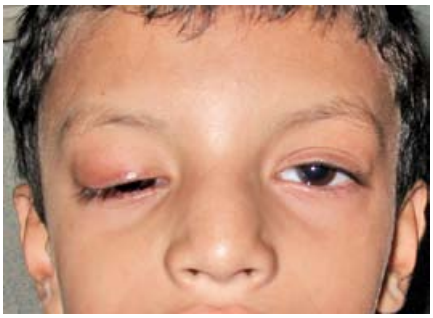


Fig. 1.40.3: Congenital bilateral ptosis—asymmetrical

Acquired Ptosis

- *Neurogenic:* Third nerve palsy **(Figs 1.41.1 and 1.41.2)** or Horner's syndrome **(Fig. 1.41.3)**
- *Myogenic*
  - *Myasthenia gravis:* Tensilon (edrophonium) or Prostigmin test (neostigmine)—a positive test means improvement of ptosis with intravenous injection **(Figs 1.41.4A and B)**
  - *Ocular myopathy* **(Fig. 1.41.5)**
  - *Senile ptosis*
- *Traumatic* **(Fig. 1.41.6)**
- *Mechanical* **(Figs 1.41.7 to 1.41.10).**

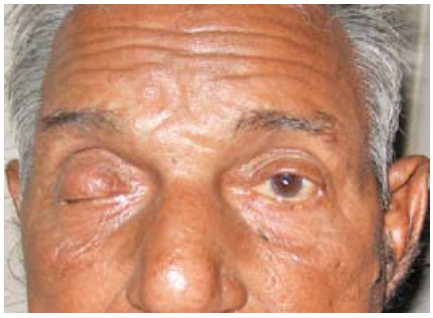


Fig. 1.41.1: Acquired ptosis—total 3rd nerve palsy

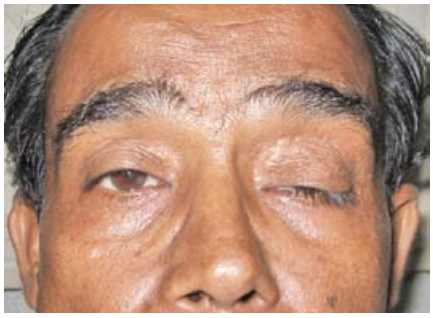


Fig. 1.41.2: Acquired ptosis—partial 3rd nerve palsy

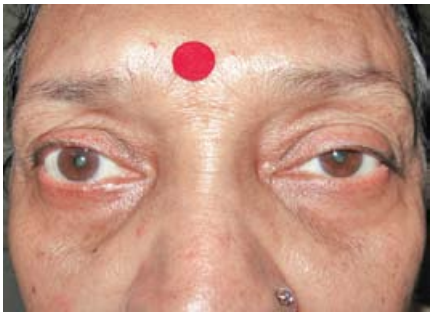


Fig. 1.41.3: Horner's syndrome—left side

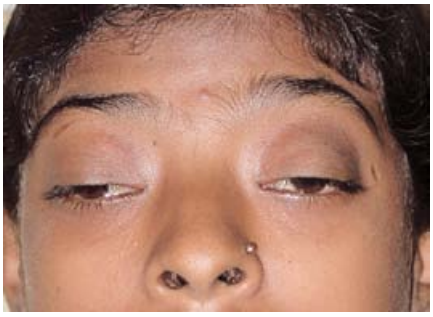


Fig. 1.41.4A: Myasthenia gravis (before Tensilon test)

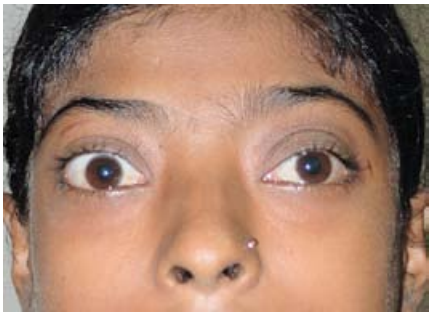


Fig. 1.41.4B: Myasthenia gravis (after Tensilon test)

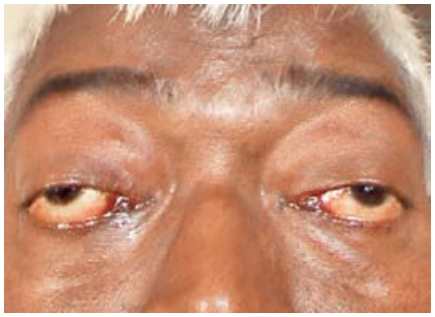


Fig. 1.41.5: Myogenic ptosis—ocular myopathy

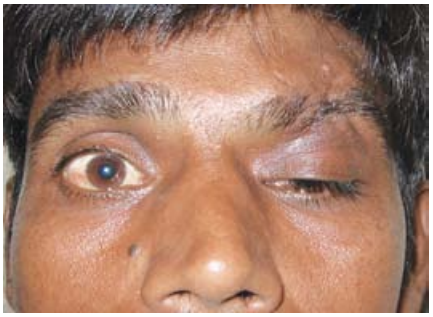


Fig. 1.41.6: Traumatic ptosis

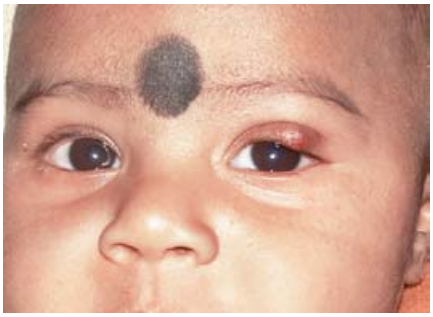


Fig. 1.41.7: Mechanical ptosis—upper lid mass

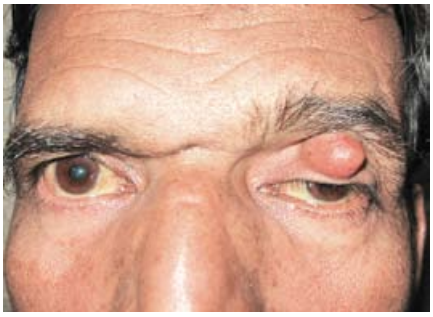


Fig. 1.41.8: Mechanical ptosis—upper lid mass

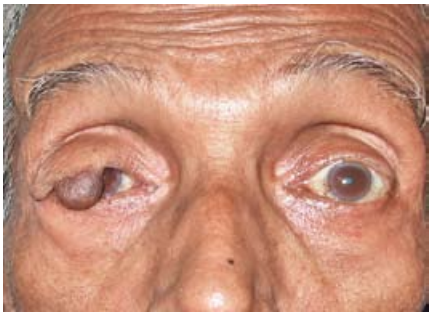


Fig. 1.41.9: Mechanical ptosis plexiform neurofibroma

Fig. 1.41.10: Mechanical ptosis—hemangioma upper lid

Senile (Aponeurotic) Ptosis

- Common unilateral or bilateral ptosis caused by defect in levator aponeurosis **(Fig. 1.42.1)**
- Good levator function
- Absent or high upper lid crease
- May be with baggy eyelids **(Fig. 1.42.2)**
- Thinning of upper lid above the tarsal plate
- Deep upper supratarsal sulcus (**Fig. 1.42.3**)
- *Treatment:* Surgical correction in severe cases.

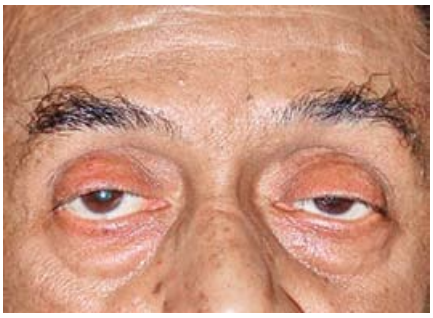


Fig. 1.42.1: Bilateral senile ptosis

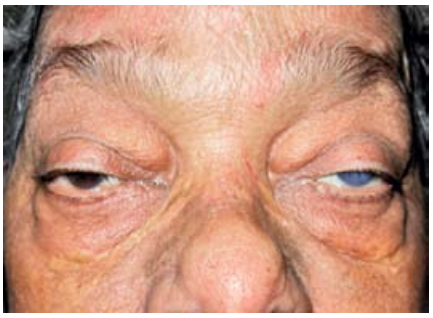


Fig. 1.42.2: Senile ptosis with baggy lids

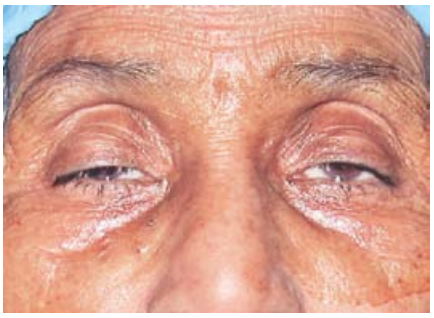


Fig. 1.42.3: Senile ptosis—deep supratarsal sulcus

Lid Retraction

- May be congenital **(Fig. 1.43.1)** or acquired
- Unilateral or bilateral retraction **(Fig. 1.43.2)** of the upper lid or sometimes both lids in the primary position
- *Causes:* Thyroid eye diseases **(Fig. 1.43.3)**, neurogenic **(Figs 1.43.4 and 1.43.5)**, surgical overcorrection **(Fig. 1.43.6)**, phenylephrine eye drops **(Fig. 1.43.7)**, hydrocephalus, etc.
- To be differentiated from pseudoretraction as seen in unilateral ptosis **(Fig. 1.43.8)**
- *Treatment* is directed towards the cause.

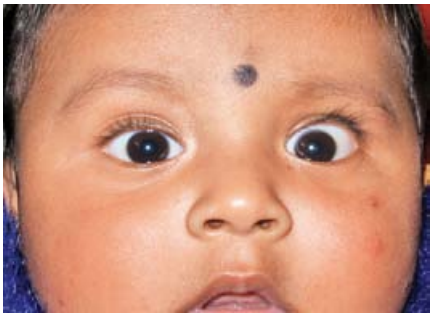


Fig. 1.43.1: Congenital lid retraction—LE

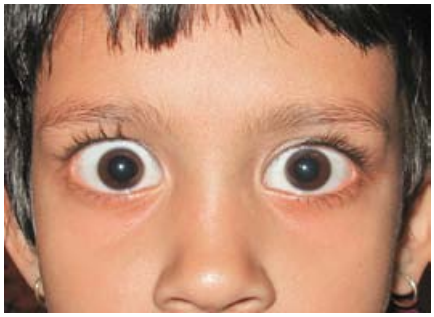


Fig. 1.43.2: Congenital lid retraction—both eyes

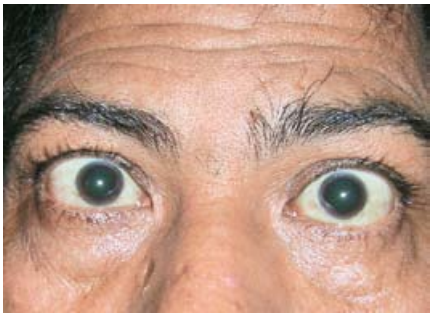


Fig. 1.43.3: Lid retraction—thyroid eye diseases

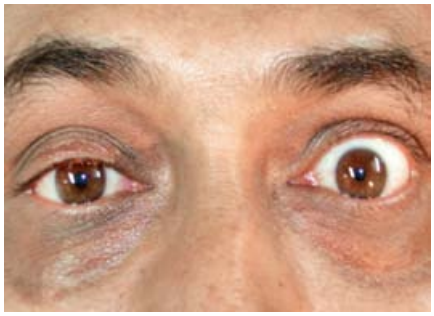


Fig. 1.43.4: Lid retraction—neurogenic

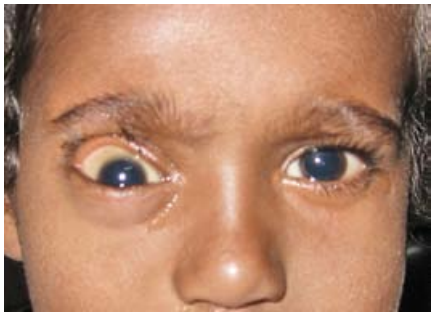


Fig. 1.43.5: Lid retraction with right hypotropia following brain abscess drainage—neurogenic

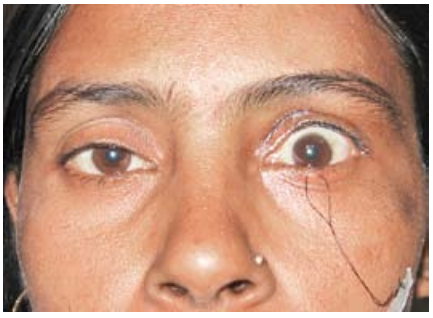


Fig. 1.43.6: Lid retraction—post-surgical

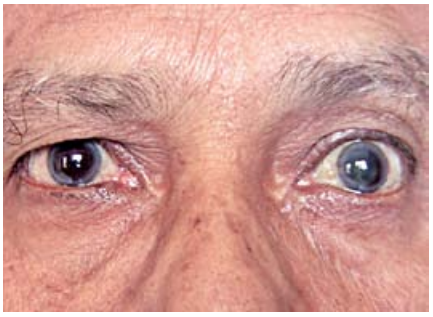


Fig. 1.43.7: Lid retraction—pharmacological in LE

Fig. 1.43.8: Pseudoretraction in LE with ptosis in RE

Other Lid Conditions

Chemosis or Lid Edema

- Diffuse edematous swelling of the eyelids
- May be associated with conjunctival chemosis (**Fig. 1.44.1**)
- *Causes:*
  - Blepharitis (**Fig. 1.44.2**)
  - Conjunctivitis (**Fig. 1.44.3**)
  - Blepharoconjunctivitis (**Fig. 1.44.4**)
  - Acute dacryocystitis (**Fig. 1.44.5**) and dacryoadenitis (S-shaped lid margin) (**Fig. 1.44.6**)
  - Simple allergy (insect bite or urticaria) (**Fig. 1.44.7**)
  - Postsurgical
  - Inflammatory orbital diseases (**Fig. 1.44.8**) or orbital cellulitis (**Fig. 1.44.9**)
- *Treatment* is directed towards the cause.

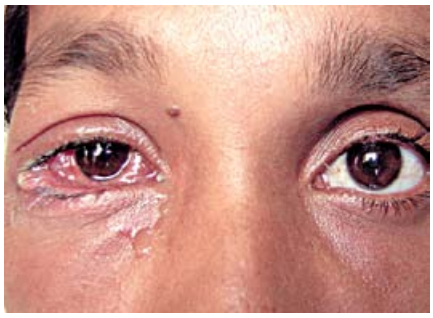


Fig. 1.44.1: Blepharoconjunctival chemosis

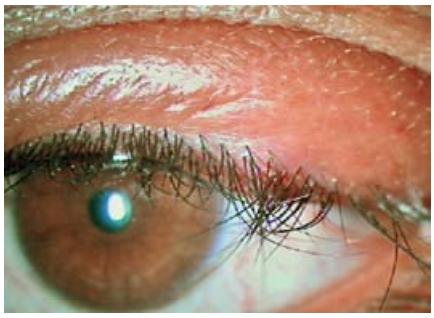


Fig. 1.44.2: Chemosis of lid—blepharitis

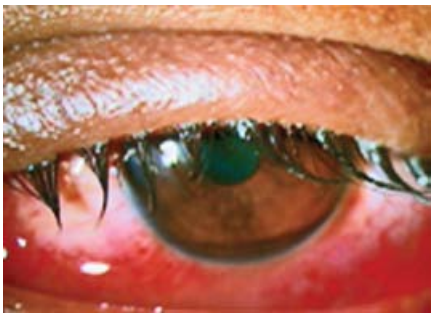


Fig. 1.44.3: Chemosis of lid—conjunctivitis

Fig. 1.44.4: Chemosis—blepharoconjunctivitis

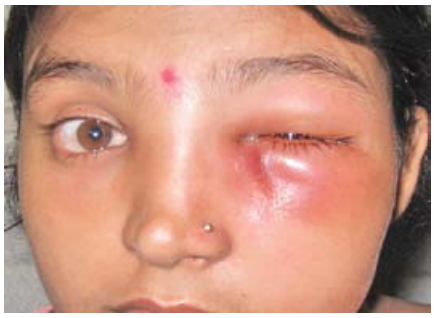


Fig. 1.44.5: Chemosis of lid—acute dacryocystitis

Fig. 1.44.6: Chemosis of lids—dacryoadenitis

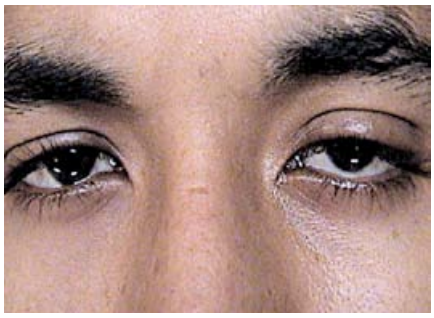


Fig. 1.44.7: Chemosis of lid—insect bite—left eye

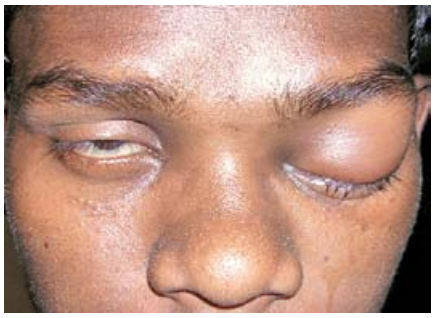


Fig. 1.44.8: Chemosis of lid—inflammatory orbital disease

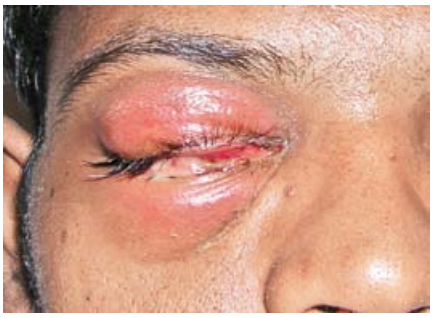


Fig. 1.44.9: Chemosis of lids—orbital cellulitis

Lagophthalmos

- Inadequate closure of the upper eyelid **(Fig. 1.45.1A)**
- Associated ectropion of lower eyelid
- Dryness of the lower part of the bulbar conjunctiva and cornea, and causing exposure keratitis **(Fig. 1.45.1B)**
- Rarely it may be bilateral with corneal ulceration **(Fig. 1.45.2A and B)**
- *Treatment:* Artificial tears, tarsorrhaphy and lid-load operation.

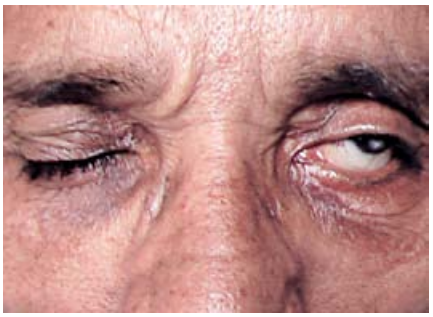


Fig. 1.45.1A: Lagophthalmos—incomplete closure

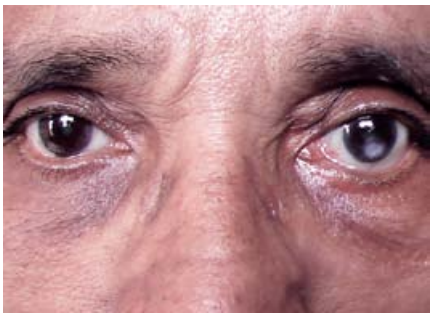


Fig. 1.45.1B: Lagophthalmos—healed exposure keratitis

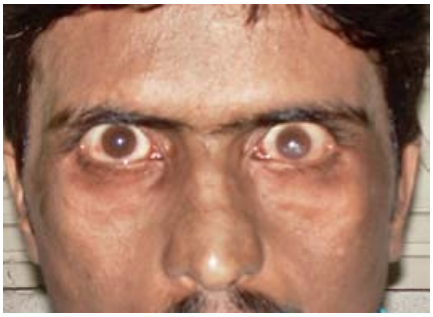


Fig. 1.45.2A: Bilateral lagophthalmos

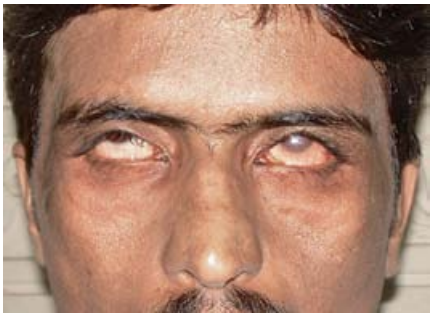


Fig. 1.45.2B: Bilateral lagophthalmos—exposure keratitis—LE

Blepharochalasis

- Younger individuals; may be unilateral or bilateral
- Starts at puberty with an intermittent angioneurotic edema **(Fig. 1.46.1)** and redness of the lid
- Skin hangs down over the upper eyelid **(Figs 1.46.2 and 1.46.3)**
- *Treatment:* Towards the allergic problems and surgical correction if required.

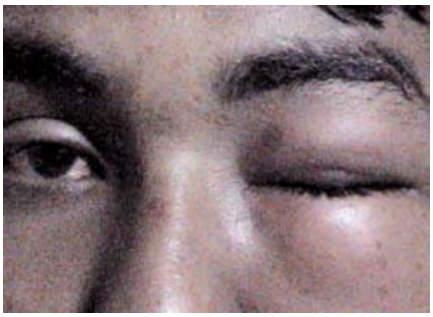


Fig. 1.46.1: Blepharochalasis—angioneurotic edema

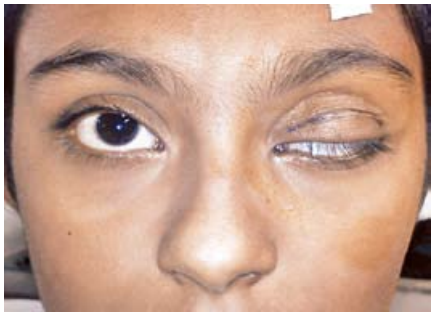


Fig. 1.46.2: Blepharochalasis—unilateral

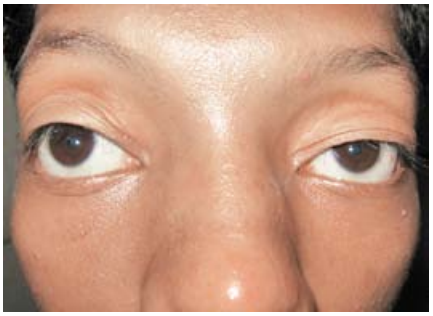


Fig. 1.46.3A: Blepharochalasis—bilateral

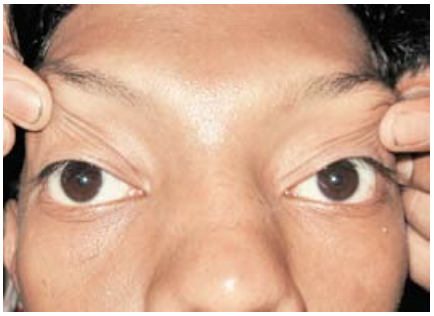


Fig. 1.46.3B: Blepharochalasis—loose skin

Dermatochalasis

- Older individual; usually bilateral
- Loss of skin elasticity of the lids
- Prolapse of the fat, mainly nasally in the upper lid **(Fig. 1.47.1)**
- Loose upper lid skin. Lower lid is also involved with bagginess **(Figs 1.47.2A and B)**
- *Treatment:* Cosmetic surgery is useful, but a recurrence is common.

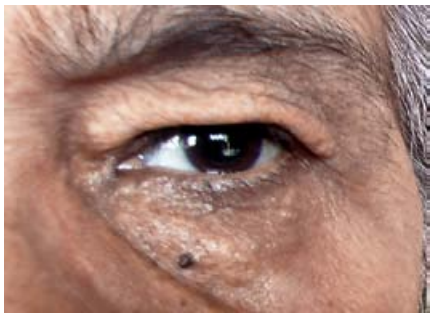


Fig. 1.47.1: Dermatochalasis

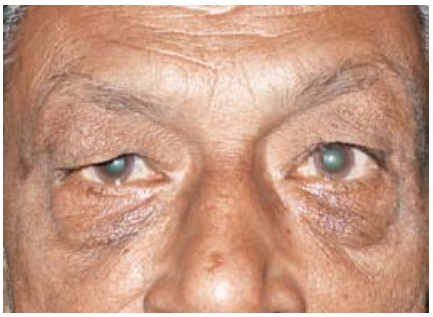


Fig. 1.47.2A: Dermatochalasis

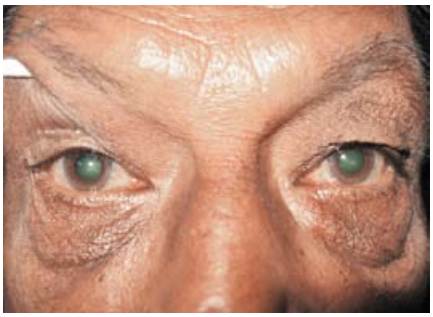


Fig. 1.47.2B: Dermatochalasis—lifting the loose skin

Floppy Eyelid Syndrome

- Floppy, easily eversible upper eyelid with papillary conjunctivitis **(Fig. 1.48.1)**
- Chronically red and irritable eye
- Typically obese and with sleep apnea syndrome
- *Treatment:* Artificial tears, antibiotic ointment at night, correct sleep posture and lid taping.

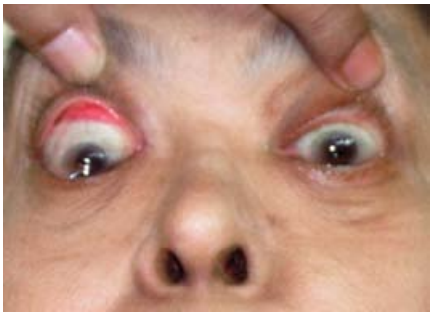


Fig. 1.48.1: Floppy eyelid syndrome

Symblepharon

- Adhesion of lid with the globe as a result of adhesion between bulbar and palpebral conjunctiva
- May be in the form of small band **(Fig. 1.49.1)** or frank broad adhesion **(Fig. 1.49.2)**
- *Causes:* Chemical burn **(Fig. 1.49.3)**, traumatic **(Fig. 1.49.4)**, thermal burn **(Fig. 1.49.5)**, ocular pemphigoid **(Figs 1.49.6A and B)**, Stevens Johnson's syndrome **(Figs 1.49.7A and B)**, trachoma, etc.
- May be anterior, posterior or total
- *Treatment:* Radical excision of scar tissue along with diseased conjunctiva and conjunctival autograft.
  Amniotic membrane transplantation may be helpful. *Prevention* by sweeping a glass rod, and symblepharon ring.

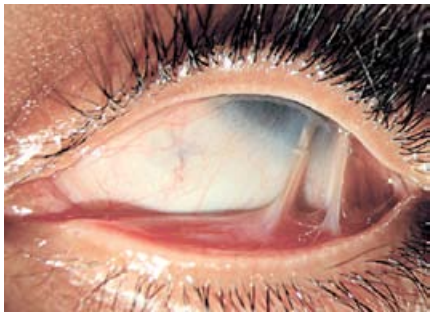


Fig. 1.49.1: Symblepharon—chemical burn

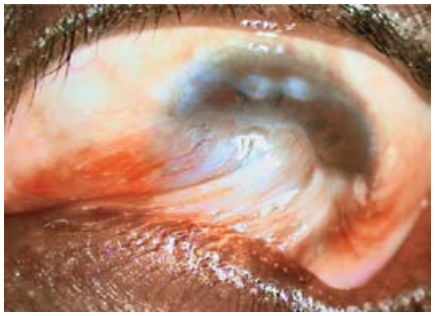


Fig. 1.49.2: Symblepharon—broad adhesion

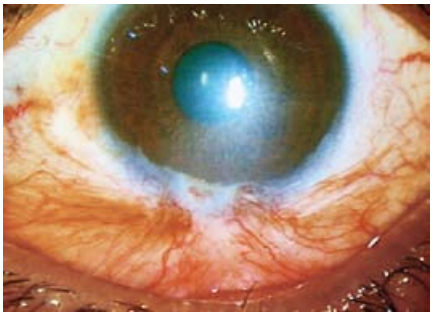


Fig. 1.49.3: Symblepharon—chemical burn

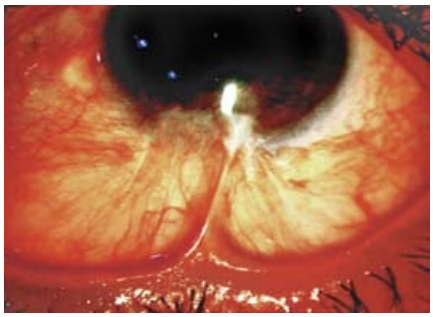


Fig. 1.49.4: Symblepharon—traumatic

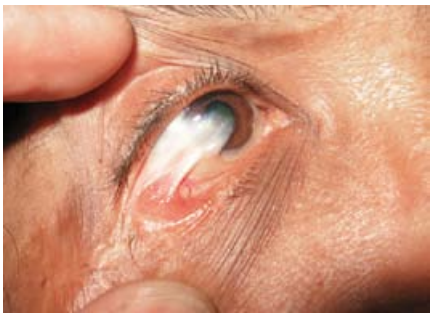


Fig. 1.49.5: Symblepharon and cicatricial ectropion following thermal burn

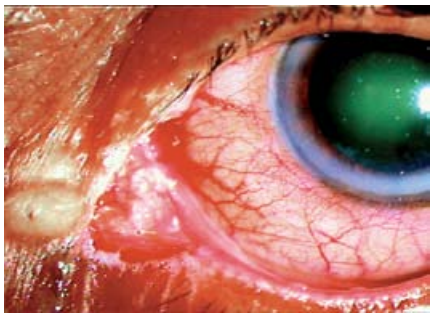


Fig. 1.49.6A: Symblepharon in ocular cicatricial pemphigoid

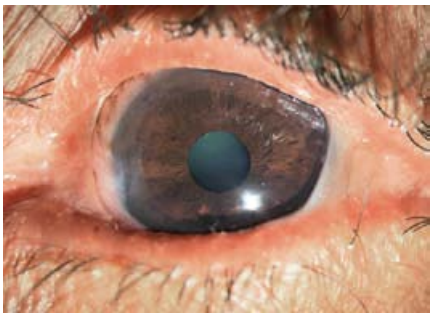


Fig. 1.49.6B: Symblepharon in ocular cicatricial pemphigoid

Essential Blepharospasm

- Spontaneous in older patient
- Involuntary tonic, spasmodic, bilateral contraction of orbicularis oculi (**Figs 1.50.1 and 1.50.2**)
- Other muscles of face may involve simultaneously
- *Treatment:* Botulinum toxin, alcohol injection
- ***Reflex blepharospasm*** may occur in superficial corneal problems (**Fig. 1.50.3**) and is abolished by topical anesthesia.

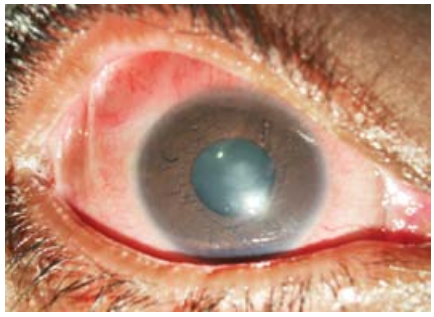


Fig. 1.49.7A: Symblepharon in Stevens-Johnson syndrome—RE

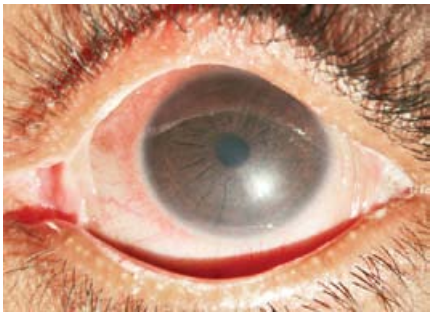


Fig. 1.49.7B: Symblepharon in Stevens-Johnson syndrome—LE

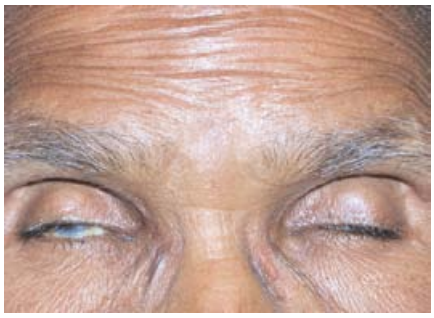


Fig. 1.50.1: Essential blepharospasm

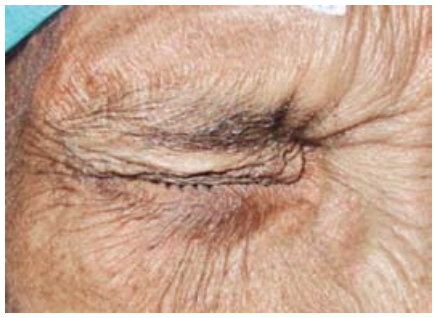


Fig. 1.50.2: Essential blepharospasm

Fig. 1.50.3: Reflex blepharospasm—left eye

Ecchymosis of the Eyelids

- Occurs after a blunt trauma **(Fig. 1.51.1)**
- Painful edema with variable degree of ecchymosis
- Called 'Panda bear' or 'Raccoon eyes' sign **(Fig. 1.51.2)** when both lids are involved as after a head injury or severe whooping cough
- *Treatment:* No active treatment is required.

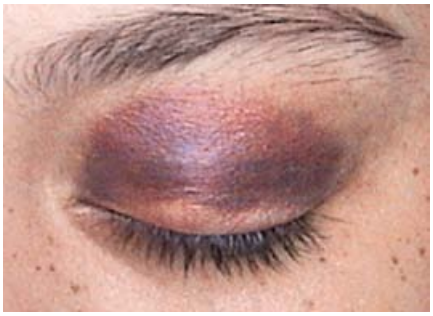


Fig. 1.51.1: Adult phthirus pubis

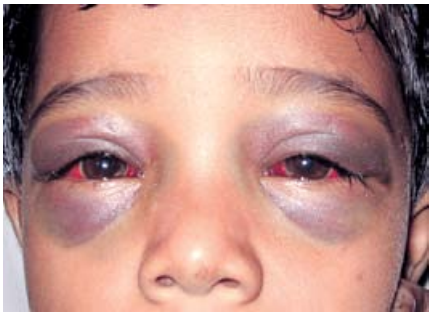


Fig. 1.51.2: Bilateral ecchymosis of lids—Panda bear (Raccoon eyes) sign

Phthiriasis Palpebrum

- Infestation of eyelashes with crab louse "*Phthirus pubis*" **(Fig. 1.52.1)** and its ova, called nits
- Typically affects children and young female
- The lice are adherent to the skin and nits stuck to the eyelashes **(Fig. 1.52.2)**
- *Treatment:* Cotton pellet soaked in pilocarpine eye drop applied over the eyelashes for few minutes then the lice can be easily removed
  - *Simultaneous treatment* for body louse infestation for both partners.

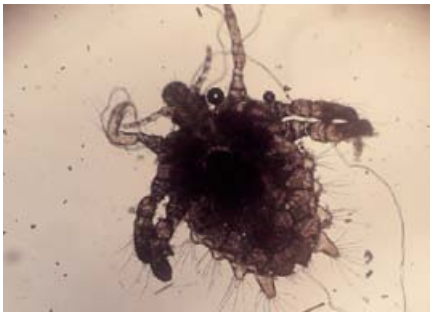


Fig. 1.52.1: Adult phthirus pubis

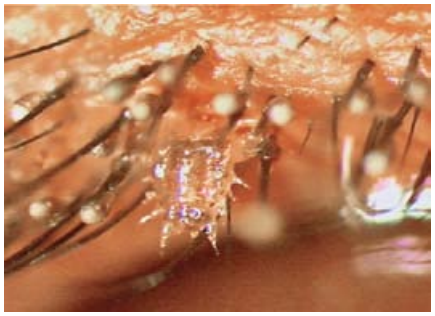


Fig. 1.52.2: Phthiriasis palpebrum with nits

Benign Lid Conditions

Xanthelasma

- Raised, yellow plaques, most commonly found at the inner portion of the upper eyelids **(Fig. 1.53.1)** rarely in lower lid **(Fig. 1.53.2)**
- Often symmetrical and progress slowly
- Eventually, it spreads towards all four lids **(Fig. 1.53.3)** and may be in circular fashion **(Fig. 1.53.4)** and like a periocular mask **(Fig. 1.53.5)**
- May be associated with familial hypercholesterolemia
- Produce only a cosmetic defect
- *Treatment:* Cosmetic surgery may be tried.

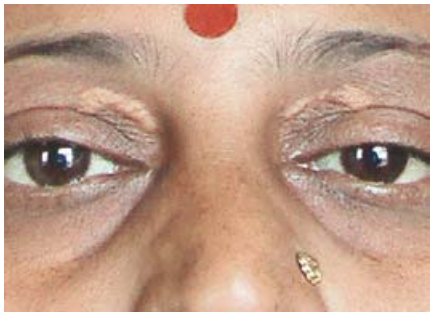


Fig. 1.53.1: Xanthelasma—upper lids

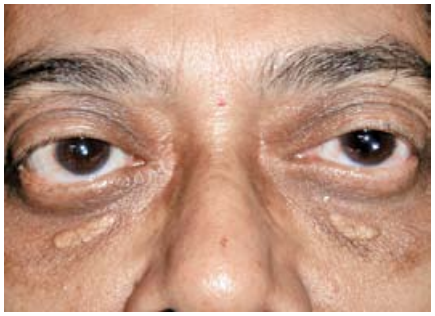


Fig. 1.53.2: Xanthelasma—lower lids

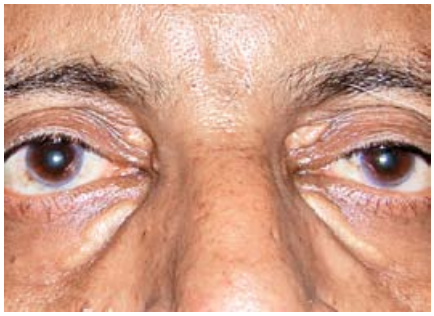


Fig. 1.53.3: Xanthelasma—all four lids

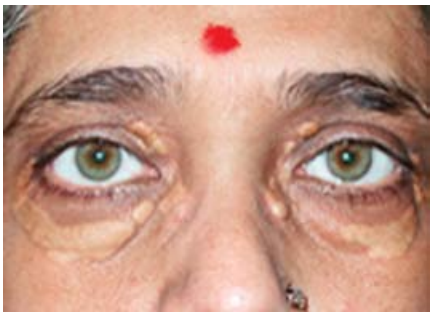


Fig. 1.53.4: Xanthelasma—circular pattern

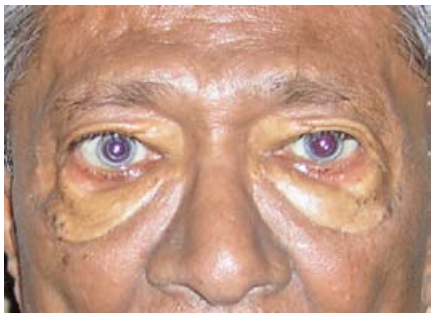


Fig. 1.53.5: Xanthelasma—extensive periocular mask

Capillary Hemangioma (Strawberry Nevus)

- Develop soon after birth, then grows for 6 months to 1 year
- Irregular, raised, bright red lesion (**Figs 1.54.1 and 1.54.2**)
- May involute spontaneously in some cases as an elevated solitary mass (**Fig. 1.54.3**)
- It blanches on pressure and may swell on crying
- Associated with similar skin lesions elsewhere
- *Treatment:* Hypertonic saline/corticosteroids injection and laser therapy.

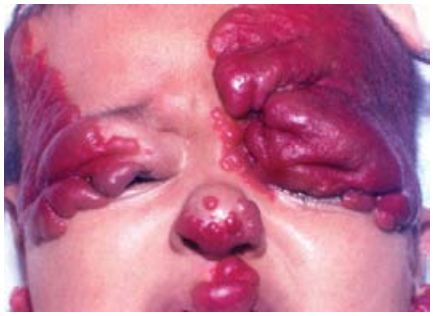


Fig. 1.54.1: Capillary (strawberry) hemangioma

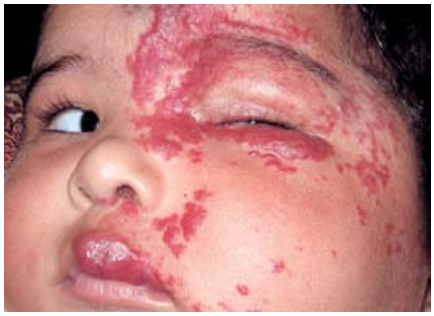


Fig. 1.54.2: Capillary (strawberry) hemangioma

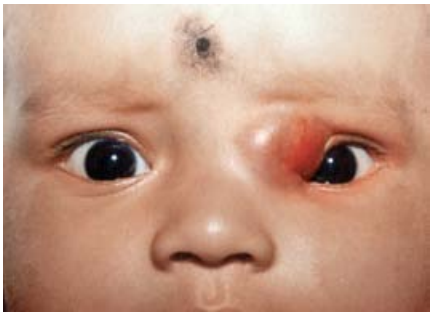


Fig. 1.54.3: Capillary hemangioma—solitary

Port-wine Stain (Nevus Flammeus)

- Congenital bilateral or unilateral lesion
- Sharply demarcated red to purple patch along the first and second divisions of the fifth nerve (**Fig. 1.55.1**)
- Lesion does not blanch on pressure
- Smaller lesion does not have any implication
- Larger lesion may be associated with Sturge-Weber syndrome *(choroidal hemangioma with secondary glaucoma and hemangioma of leptomeninges)* (**Fig. 1.55.2**)
- *Treatment:* Laser therapy and treatment of glaucoma if present.

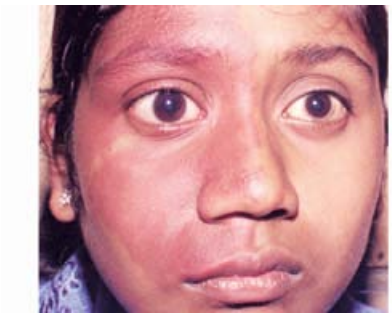


Fig. 1.55.1: Port-wine stain

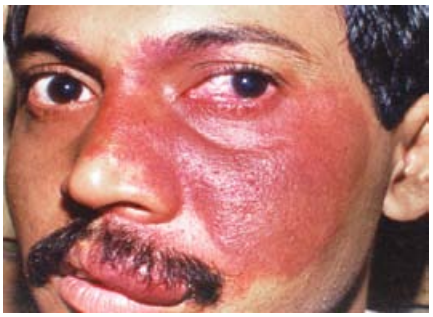


Fig. 1.55.2: Port-wine stain—Sturge-Weber syndrome

Molluscum Contagiosum

- Unilateral or bilateral disease with single or multiple lesions
- In immunodeficiency conditions, it may be more severe and confluent, often simultaneously with other parts of body
- Small, pale, yellowish-white umbilicated lesions (**Fig. 1.56.1**)
- May be a presenting feature in AIDS (**Fig. 1.56.2)** and ulceration seen in severe cases **(Fig. 1.56.3**)
- Associated with keratitis or follicular conjunctivitis (**Fig. 1.56.4**)
- *Treatment:* Investigations for immune defficiency states, chemical cautery in some cases.

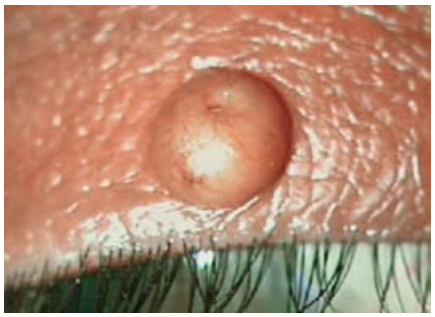


Fig. 1.56.1: Molluscum contagiosum—solitary lesion

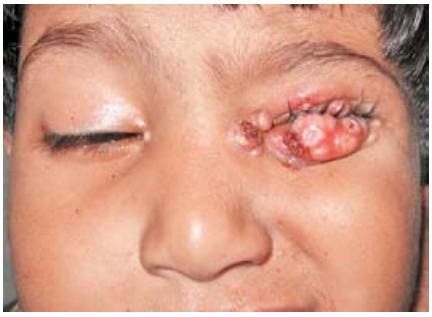


Fig. 1.56.2: Molluscum contagiosum in crops in HIV positive patient

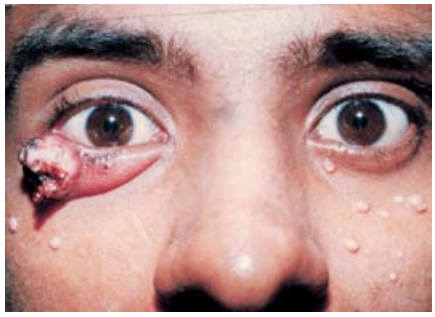


Fig. 1.56.3: Giant molluscum contagiosum with ulceration in AIDS

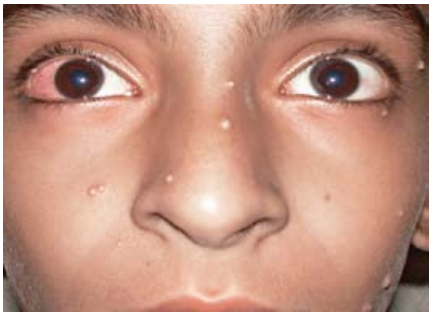


Fig. 1.56.4: Molluscum contagiosum—multiple with conjunctivitis

Sebaceous Cyst

- Bilateral and multiple cysts are more common (**Fig. 1.57.1**)
- They vary in shape and size
- Solitary cyst is more common near the canthus (**Fig. 1.57.2**)
- Yellowish-white color and cyst is filled up with inspissated sebaceous secretion, often large in size (**Figs 1.57.3 and 1.57.4**)
- *Treatment:* Excision for cosmetic reason or if there are symptoms for mechanical reasons

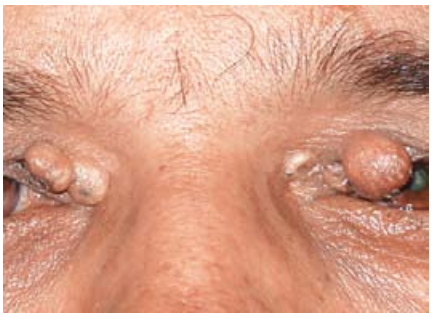


Fig. 1.57.1: Sebaceous cyst—bilateral and multiple

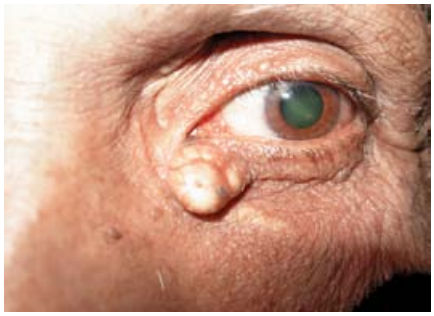


Fig. 1.57.2: Sebaceous cyst—solitary

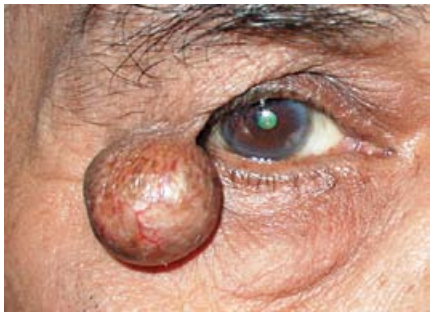


Fig. 1.57.3: Large sebaceous cyst

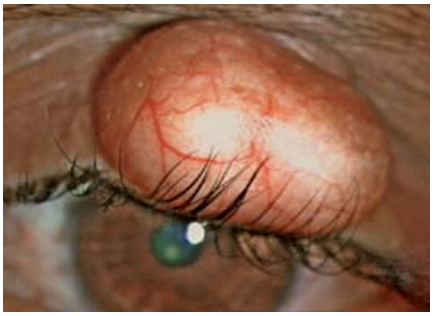


Fig. 1.57.4: Large sebaceous cyst

Keratoacanthoma

- Rare, fast-growing benign tumor
- Firm, pinkish nodule with rolled out edge and a keratin filled crater (**Figs 1.58.1 and 1.58.2**)
- *Treatment:* Excision in case of large lesion.

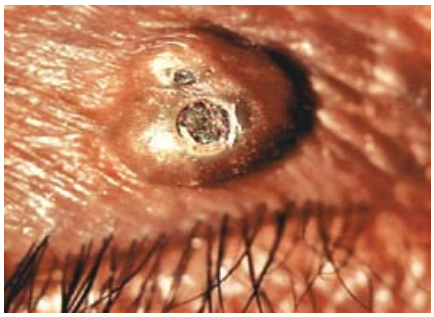


Fig. 1.58.1: Keratoacanthoma

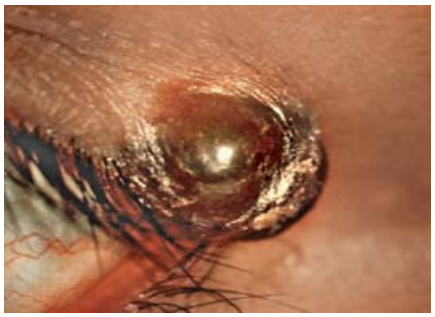


Fig. 1.58.2: Keratoacanthoma

Keratic Horn

- A rare benign condition
- Hyperkeratotic horn-like lesion which protrudes from skin surface **(Figs 1.59.1 and 1.59.2)**
- *Treatment:* Excision of the mass if necessary.

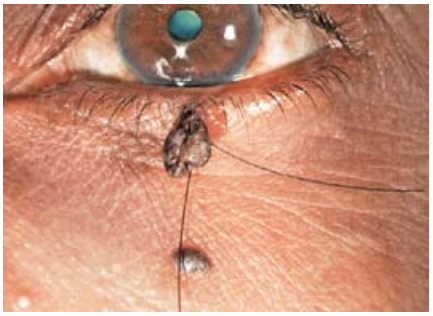


Fig. 1.59.1: Keratic horn—small

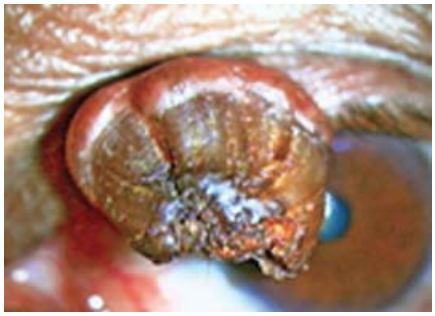


Fig. 1.59.2: Keratic horn—large

Squamous Cell Papilloma (Viral Wart)

- Most common benign tumor of the lid
- Pedunculated or sessile lesion with a characteristic irregular raspberry like surface **(Figs 1.60.1 and 1.60.2)**
- *Treatment:* Excision if necessary.

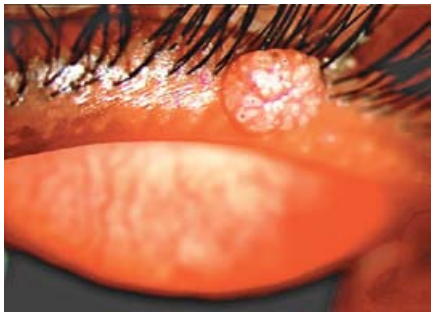


Fig. 1.60.1: Squamous cell papilloma

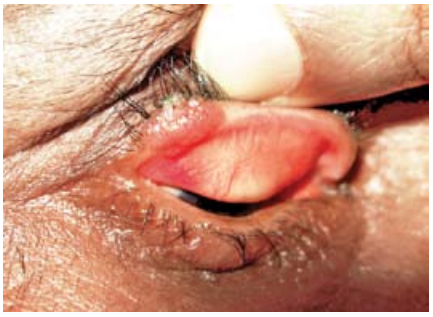


Fig. 1.60.2: Squamous cell papilloma

Basal Cell Papilloma (Seborrheic Keratosis)

- Common in middle-aged and elderly people
- Discrete, round, brownish or blackish lesion with variegated surface (**Fig. 1.61.1**)
- It may have papillomatous appearence (**Fig. 1.61.2**)
- *Treatment:* Excision if necssary in severe cases.

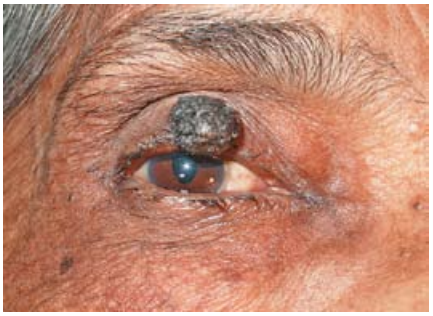


Fig. 1.61.1: Basal cell papilloma

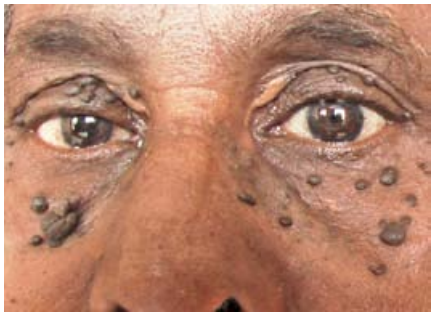


Fig. 1.61.2: Multiple basal cell papilloma

Oculodermal Melanocytosis (Nevus of Ota)

- Rare, congenital condition
- Bluish-gray discoloration of the skin affecting the 5th nerve **(Fig. 1.62.1)**
- Associated conjunctival melanosis
- Hyperpigmentation of the iris (heterochromia).

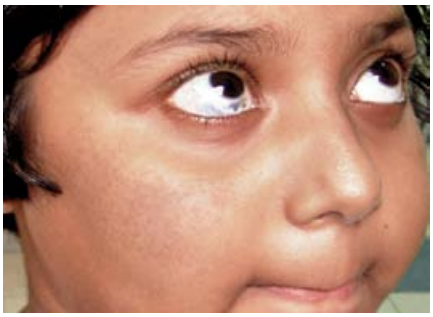


Fig. 1.62.1: Oculodermal melanocytosis (nevus of Ota)

Acquired Nevus

- *Three types:* Junctional **(Fig. 1.63.1)**, compound **(Fig. 1.63.2)** and intradermal **(Fig. 1.63.3)**
- Elevated or flat lesions with variable degree of brown to black pigmentation
- Intradermal type may be of large size to obscure the visual axis and lash may protrude through it (**Figs 1.63.4A and B)**
- May be of kissing type in any variety **(Fig. 1.63.5)**
- Junctional and compound nevi have low malignant potential whereas intradermal nevus is benign
- *Treatment:* Surgical excision for cosmetic reason and when there is obstruction of visual axis.

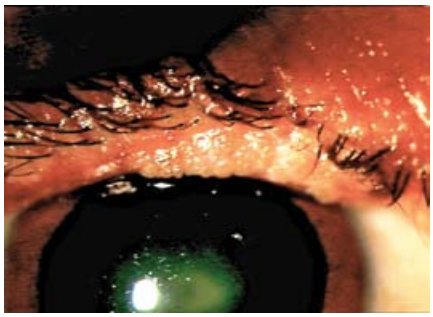


Fig. 1.63.1: Junctional nevus

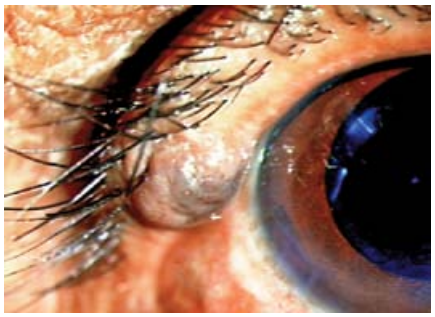


Fig. 1.63.2: Compound nevus

Fig. 1.63.3: Intradermal nevus

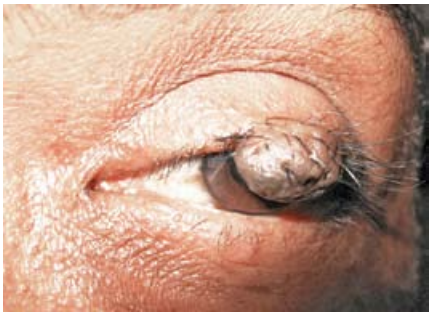


Fig. 1.63.4A: Intradermal nevus—obstructing the visual axis

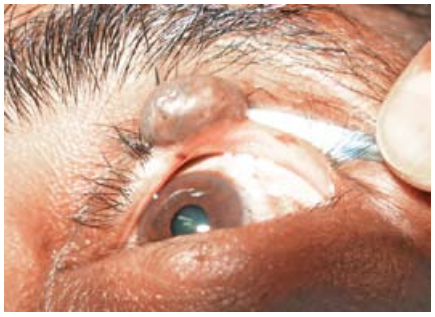


Fig. 1.63.4B: Intradermal nevus—obstructing the visual axis

Fig. 1.63.5: Intradermal nevus—kissing type

Milia and Comedones

- Small, multiple, round, superficial cysts **(Fig. 1.64.1)**
- It tends to occurs in crops and often bilateral
- *No treatment is required*
- ***Comedones:*** More common in acne vulgaris patients and consist of keratin and/or sebum filled multiple cysts. They may be black-headed or white headed depending upon presence of melanin **(Figs 1.64.2 to 1.64.4)**
- *Treatment:* For cosmetic reason.

Fig. 1.64.1: Milia

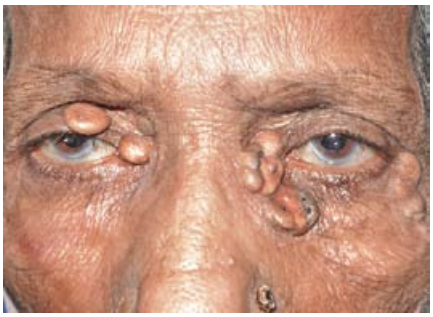


Fig. 1.64.2: Sebaceous cyst with white head comedones

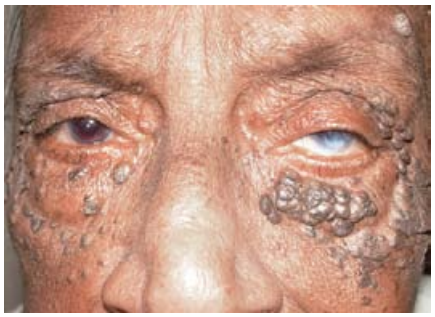


Fig. 1.64.3: Black head comedones

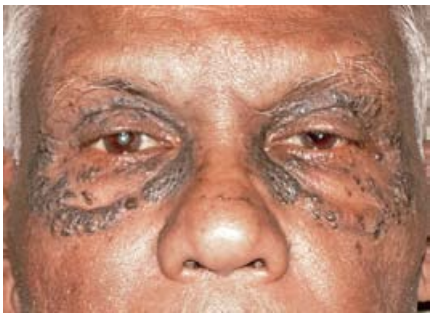


Fig. 1.64.4: Black head comedones

Cyst of Moll

- Very common, painless, chronic, transparent cystic nodule just on the lid margin containing serous secretion **(Figs 1.65.1A and B)**
- Cyst of Zeis:
  - Similar kind of cystic swelling on the external aspect of lid margin
  - It contains oily secretion **(Fig. 1.65.2)**
- ***Eccrine hidrocystoma:***
  - Less common and similar to cyst of Moll.
  - It occurs near medial or lateral angle and does not involve the lid magin **(Fig. 1.65.3)**

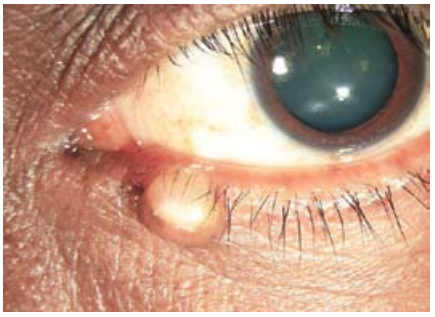


Fig. 1.65.1A: Cyst of Moll

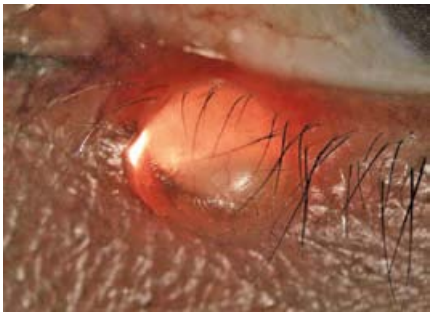


Fig. 1.65.1B: Cyst of Moll

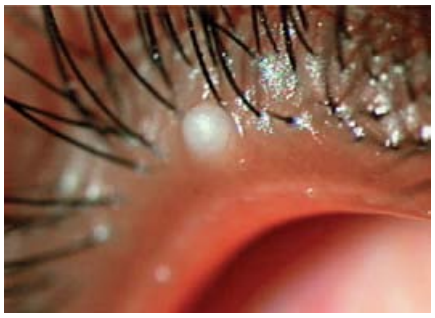


Fig. 1.65.2: Cyst of Zeis

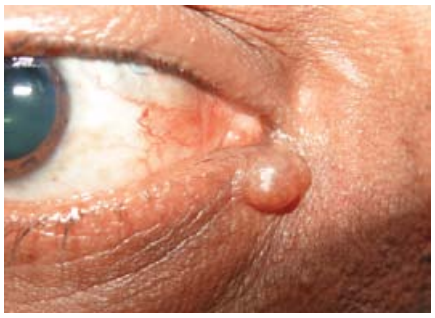


Fig. 1.65.3: Eccrine hidrocystoma

Epidermal Inclusion Cyst

- Rare condition, due to trauma or after surgery
- Solitary, firm slowly progressive subepithelial nodule **(Fig. 1.66.1)**
- Most commonly found in upper lid, may be mistaken as a chalazion
- *Treatment:* Removal of cyst from skin surface or marsupialization.

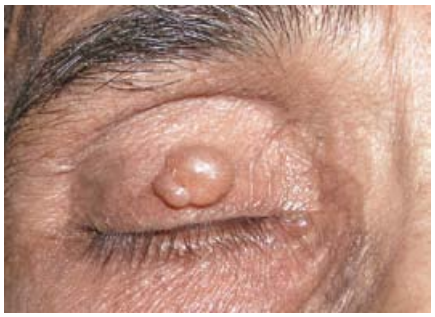


Fig. 1.66.1: Epidermal inclusion cyst

External Angular Dermoid

- Not so rare condition, present since early infancy
- Smooth, subcutaneous, firm, slow growing swelling most frequently located just below the lateral eyebrow **(Figs 1.67.1A and B)**
- May be associated with bony orbital defect
- It may occur at the inner canthus **(Fig. 1.67.2)** or near medial and anterior part of orbit **(Fig. 1.67.3)**
- *Treatment*: Excision of the mass, internal extension should be dissected carefully.

Fig. 1.67.1A: External angular dermoid

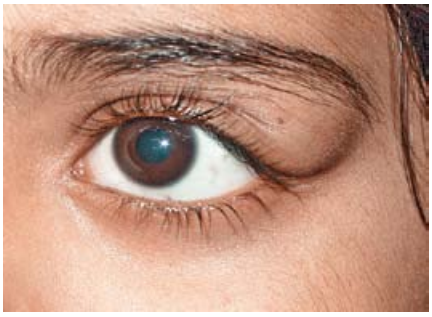


Fig. 1.67.1B: External angular dermoid

Fig. 1.67.2: Angular dermoid—medial canthus

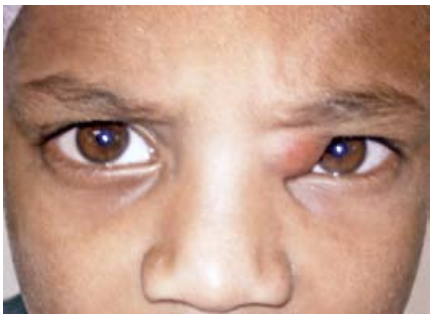


Fig. 1.67.3: Angular dermoid—internal

Malignant Lid Conditions

Basal Cell Carcinoma (Rodent Ulcer)

- Most common malignant tumor of the eyelid
- The lower lid is more commonly involved, especially near the inner canthus **(Fig. 1.68.1)**
- Initially, it starts as a small ulcer, then slowly progresses **(Figs 1.68.2 and 1.68.3)**
- The edges are raised (rolled out edges) and indurated **(Fig. 1.68.4)**

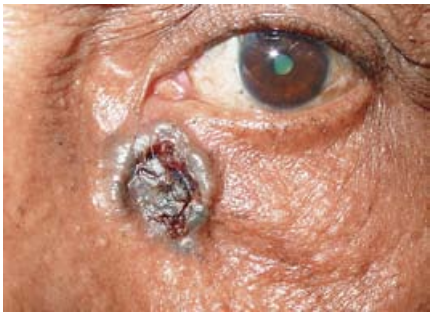


Fig. 1.68.1: Basal cell carcinoma—noduloulcerative

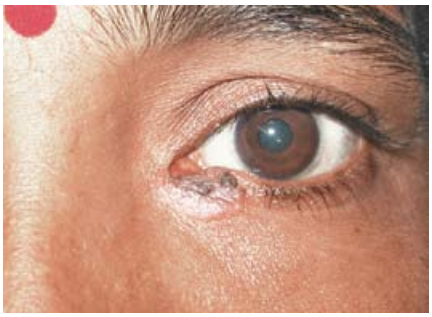


Fig. 1.68.2: Basal cell carcinoma—ulcerative type

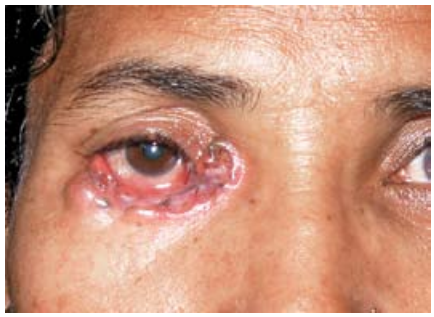


Fig. 1.68.3: Basal cell carcinoma—ulcerative type

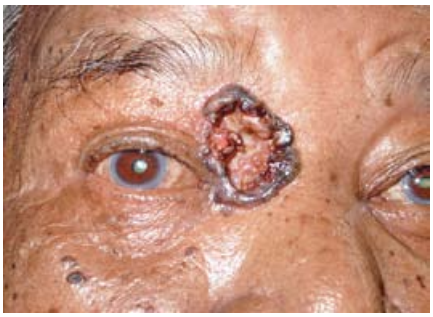


Fig. 1.68.4: Basal cell carcinoma—rolled out edges

- It may be sclerosing **(Fig. 1.68.5)**, ulcerative, nodular **(Fig. 1.68.6)** or noduloulcerative type **(Fig. 1.68.7)**
- Ulcer may erode the lid margin like a bridge **(Figs 1.68.8A and B)**
- With time, it may destroy both the lids **(Fig. 1.68.9)**
- It does not metastasize to the lymph nodes
- *Treatment:* Surgical excision, radiotherapy.

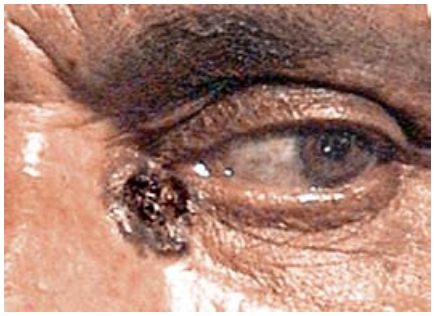


Fig. 1.68.5: Basal cell carcinoma—sclerosing

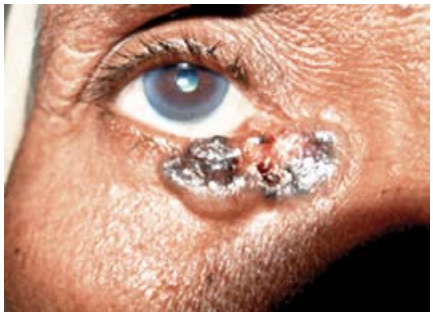


Fig. 1.68.6: Basal cell carcinoma—nodular type

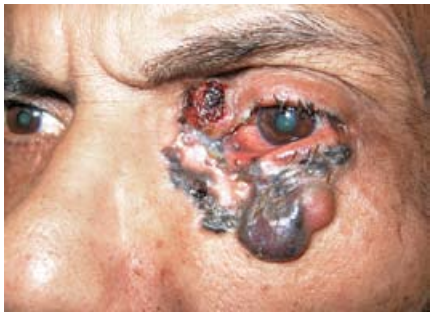


Fig. 1.68.7: Basal cell carcinoma—noduloulcerative—extensive

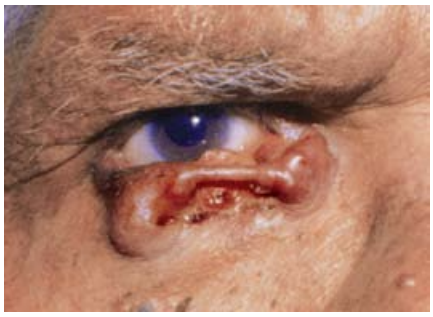


Fig. 1.68.8A: Basal cell carcinoma—bridge formation

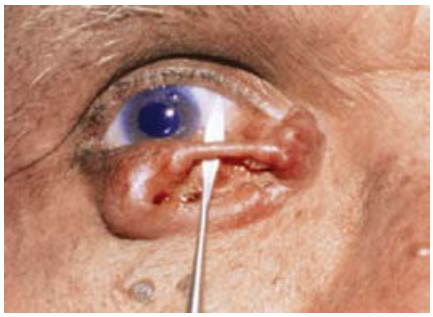


Fig. 1.68.8B: Basal cell carcinoma—bridge formation

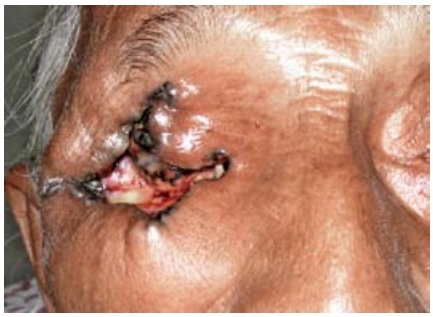


Fig. 1.68.9: Basal cell carcinoma—ulcerative with extensive extension

Squamous Cell Carcinoma

- Second most common malignancy of the eyelid
- Appears as a nodule (**Fig. 1.69.1**) or an ulcerative lesion **(Figs 1.69.2 and 1.69.3)**, or a papilloma (**Figs 1.69.4 and 1.69.5**)
- Sometimes the growth covers the whole eyeball (**Fig. 1.69.6**) or may cause ankyloblepharon (**Fig. 1.69.7**)

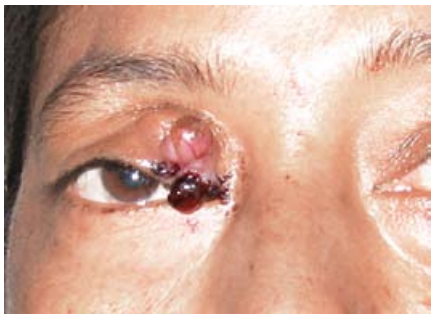


Fig. 1.69.1: Squamous cell carcinoma—nodular type

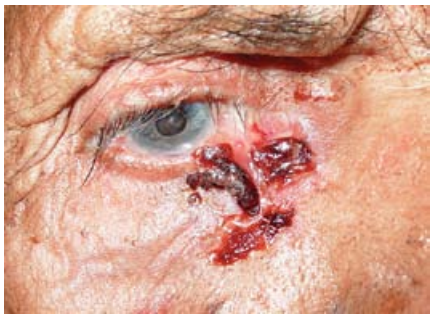


Fig. 1.69.2: Squamous cell carcinoma—ulcerative type

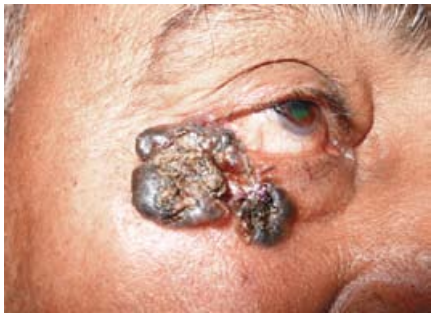


Fig. 1.69.3: Squamous cell carcinoma—noduloulcerative

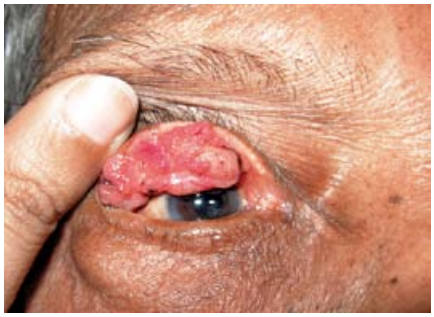


Fig. 1.69.4: Squamous cell carcinoma—papillomatous

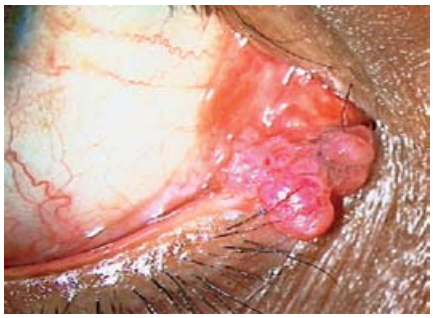


Fig. 1.69.5: Squamous cell carcinoma—papillomatous

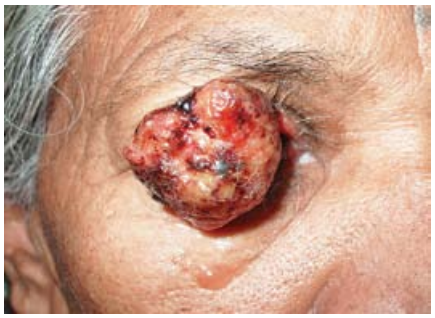


Fig. 1.69.6: Squamous cell carcinoma—large nodular

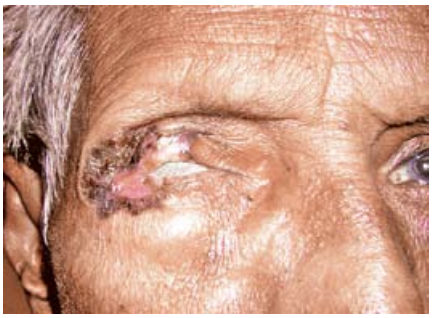


Fig. 1.69.7: Squamous cell carcinoma—ulcerative type with ankyloblepharon

- Growth rate is faster than basal cell carcinoma
- It metastasizes into the regional lymph nodes
- *Treatment*: Radical surgery with reconstruction.

Meibomian Gland Carcinoma

- Appears as a discrete, yellow, firm nodule which is sometimes incorrectly diagnosed as 'recurrent chalazion' **(Figs 1.70.1 and 1.70.2)**
- It may infiltrate along the lid margin **(Fig. 1.70.3)**
- Widespread metastasis is common in large tumor **(Figs 1.70.4 and 1.70.5)**
- Prognosis is always poor
- *Treatment*: Radical excision with reconstruction of the involved lid.

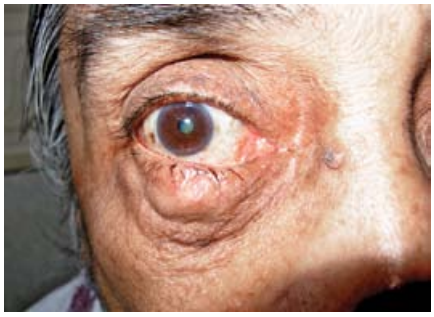


Fig. 1.70.1A: Meibomian gland carcinoma—lower lid

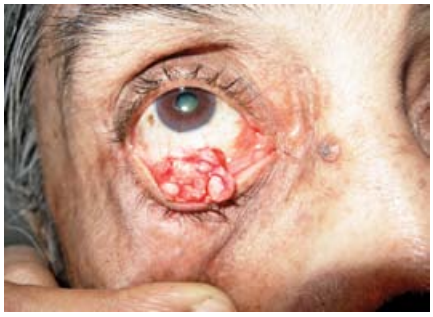


Fig. 1.70.1B: Meibomian gland carcinoma—lower lid

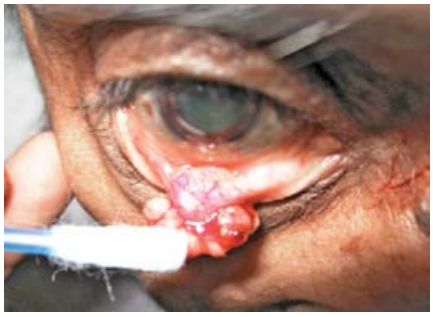


Fig. 1.70.1C: Sebaceous gland carcinoma—lower lid

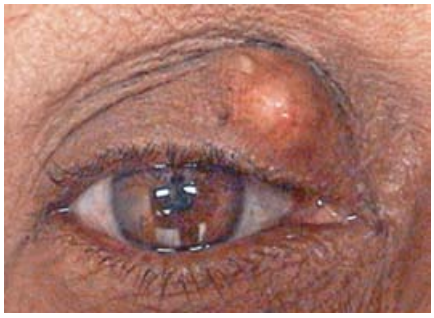


Fig. 1.70.2A: Meibomian gland carcinoma—mimicking chalazion upper lid

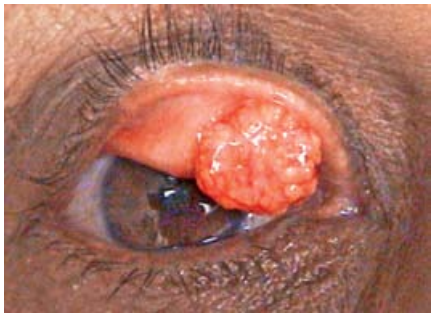


Fig. 1.70.2B: Meibomian gland carcinoma—mimicking chalazion upper lid

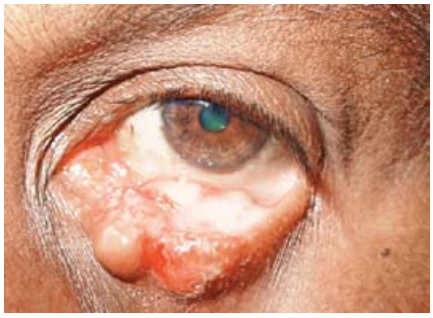


Fig. 1.70.3: Meibomian carcinoma—lower lid infiltrative type

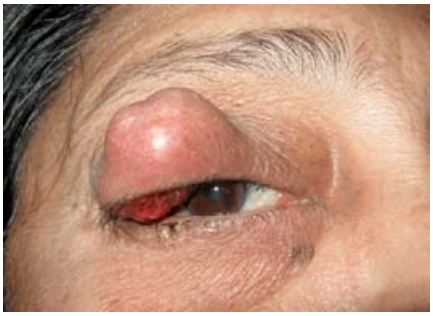


Fig. 1.70.4A: Meibomian gland carcinoma—large

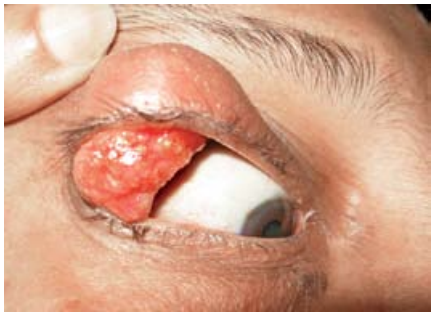


Fig. 1.70.4B: Meibomian gland carcinoma—large

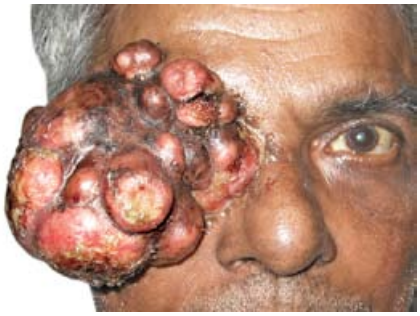


Fig. 1.70.5: Meibomian gland carcinoma—extensive orbital extension

Carcinoma of Gland of Zeis

- Affects elderly patients
- Discrete nodular slow growing tumor on the lid margin **(Fig. 1.71.1)**
- May be associated with loss of local eyelashes
- *Treatment*: Radical excision with reconstruction of the lid.

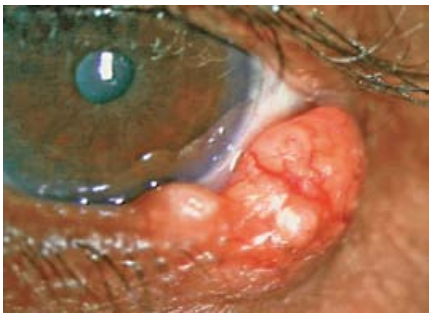


Fig. 1.71.1: Carcinoma of gland of Zeis

Other Malignant Lid Conditions

Lentigo Malignum

- Affects elderly patients
- Preinvasive stage of malignant melanoma
- Pigmented macular lesion may transform to nodular lesion **(Fig. 1.72.1)**

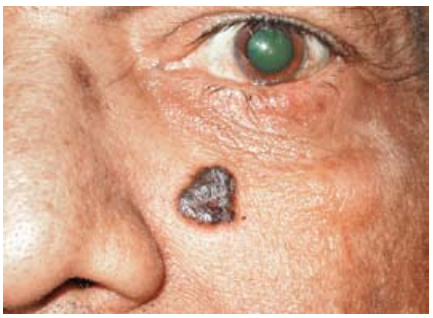


Fig. 1.72.1: Lentigo malignum

Nodular Malignant Melanoma

- May arise *de novo* or from precancerous melanoma **(Figs 1.73.1 and 1.73.2)**
- Large nodule with irregular pigmentation **(Fig. 1.73.3)**
- May show rapid growth with break down of epithelium
- *Treatment*: Radical excision or exenteration.

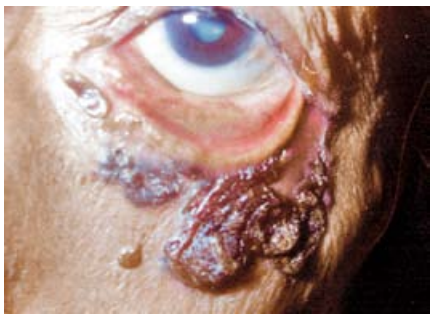


Fig. 1.73.1: Precancerous melanoma

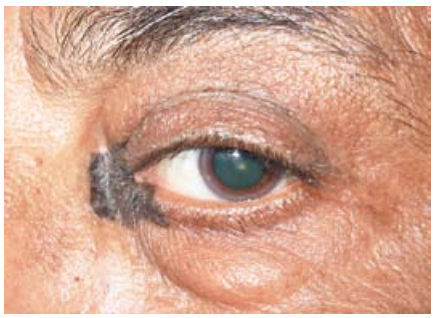


Fig. 1.73.2: Precancerous melanoma

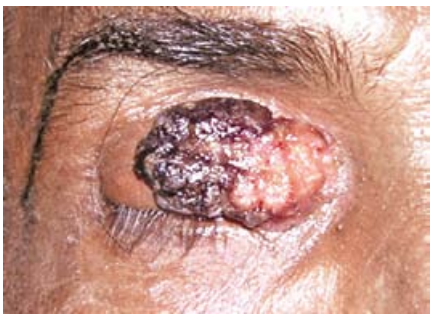


Fig. 1.73.3: Malignant melanoma

Miscellaneous Lid Conditions

Baggy Eyelids

- Usually bilateral, age related condition
- Due to orbital fat herniation through the weakened orbital septum **(Fig. 1.74.1)**
- Initially, fat-pockets herniate into the medial aspect of the upper lid, then lower lids **(Fig. 1.74.2)**
- *Treatment*: Cosmetic surgical correction, but recurrence is common

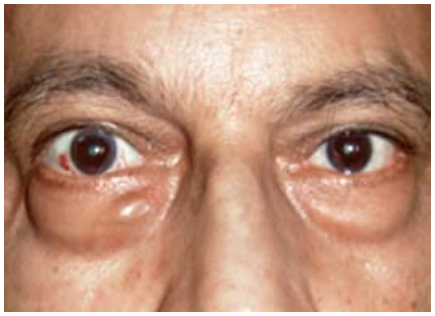


Fig. 1.74.1: Baggy eyelids

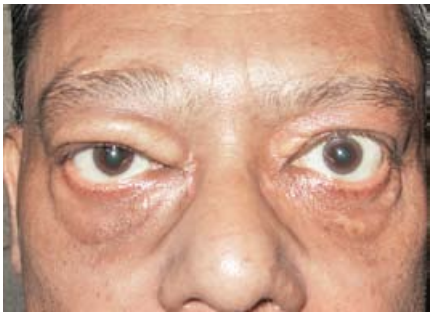


Fig. 1.74.2: Baggy eyelids with mechanical ptosis—right eye

Depigmentation of Periocular Skin

- Rare, unilateral condition
- *Cause*
  - VKH syndrome **(Fig. 1.75.1)**
  - Sympathetic ophthalmia
  - Drug induced; like topical sparfloxacin, trimethoprim drops, topical prostaglandin analogs, etc. **(Figs 1.75.2 to 1.75.4)**
- *Treatment:* Withdrawal of topical drops in selective cases.

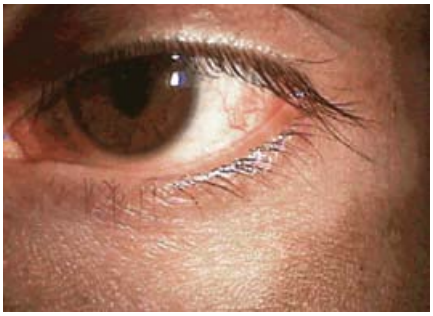


Fig. 1.75.1: Periocular depigmentation—VKH

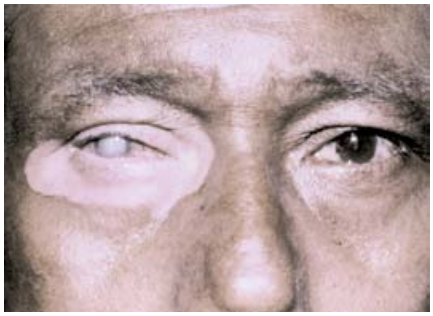


Fig. 1.75.2: Periocular depigmentation—drug induced

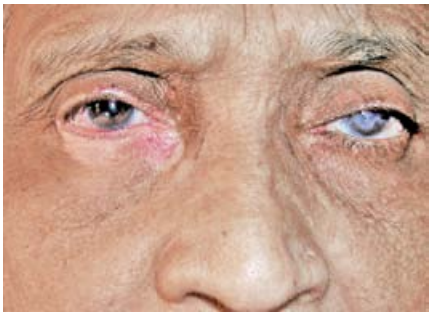


Fig. 1.75.3: Periocular depigmentation following eye drop

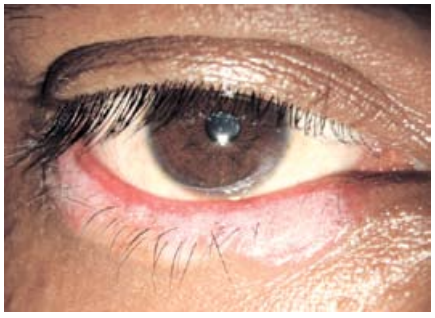


Fig. 1.75.4: Periocular depigmentation—following trimethoprim drop

Ankyloblepharon

- Adhesion between upper and lower eyelids
- May be partial or complete
- Causes
  - Congenital **(Fig. 1.76.1)**
  - Acquired: Due to chemical burn **(Figs 1.76.2 and 1.76.3)** or thermal burn **(Fig. 1.76.4)**
- Eyeball is usually disorganized
- *Treatment* is difficult and may be considered only in extreme situation.

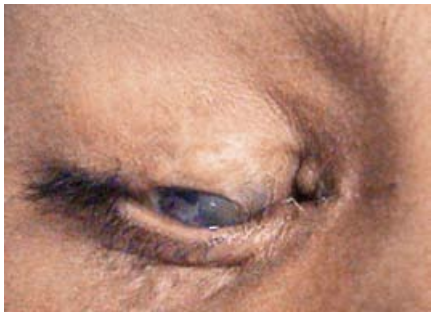


Fig. 1.76.1: Ankyloblepharon—congenital

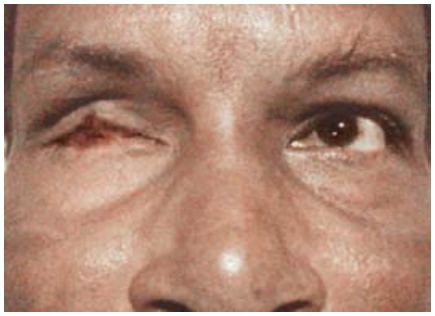


Fig. 1.76.2: Ankyloblepharon—acquired

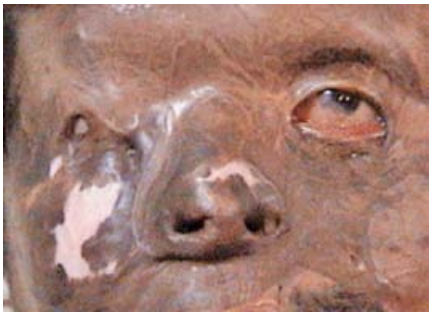


Fig. 1.76.3: Ankyloblepharon—severe acid burn

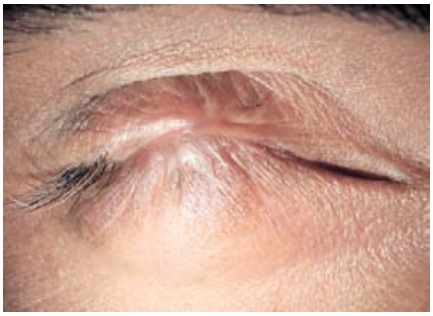


Fig. 1.76.4: Ankyloblepharon following malten metal burn

Nodular Hemangiomas

- Localized solitary nodular lesion just away from the lid margin **(Figs 1.77.1 to 1.77.4)**
- May be cavernous or capillary type
- Bright red in color
- *Treatment:* Excision of the mass and histopathological examination which confirms the diagnosis.

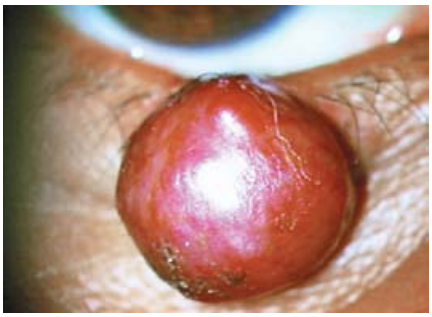


Fig. 1.77.1: Capillary hemangioma—isolated nodular

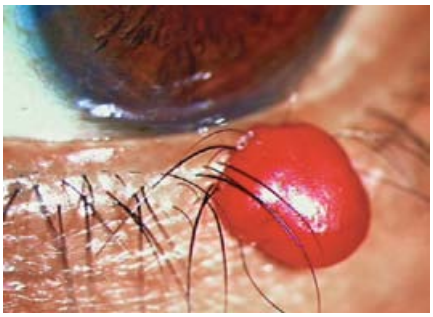


Fig. 1.77.2: Capillary hemangioma—isolated nodular

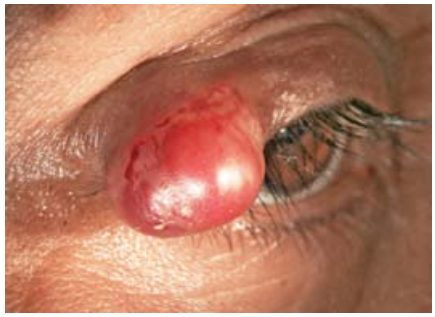


Fig. 1.77.3: Capillary hemangioma upper lid—solitary

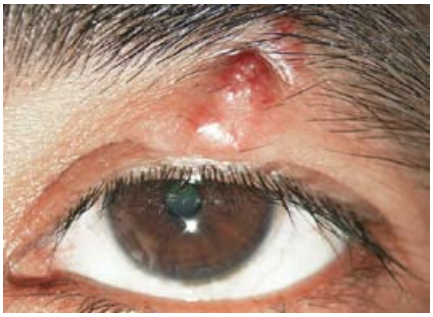


Fig. 1.77.4: Cavernous hemangioma—left upper lid

Tarsorrhaphy

- It is required to protect the integrity of the eyeball as in—impending corneal perforation, usually done for a temporary period—lateral **(Fig. 1.78.1)** or paramedian **(Fig. 1.78.2)**
- To prevent exposure keratitis, as in case of chronic lagophthalmos **(Figs 1.78.3 and 1.78.4)**, after cerebellopontine angle (CPA) tumor operation—usually permanent **(Fig. 1.78.5)** or in case of severe dry eye **(Figs 1.78.6A and B)**
- *In temporary cases*—the tarsorrhaphy is released after satisfactory recovery.

Fig. 1.78.1: Temporary lateral tarsorrhaphy

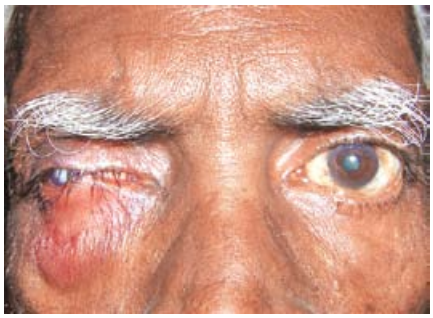


Fig. 1.78.2: Temporary median tarsorrhaphy

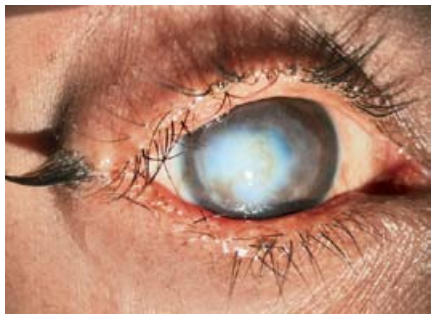


Fig. 1.78.3: Permanent lateral tarsorrhaphy—in lagophthalmos

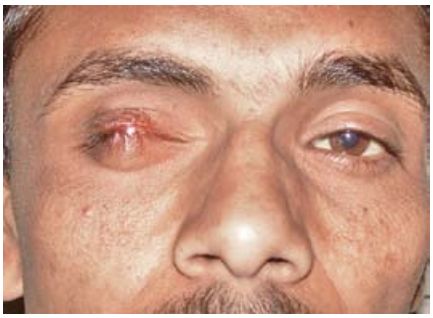


Fig. 1.78.4: Permanent tarsorrhaphy in lagophthalmos

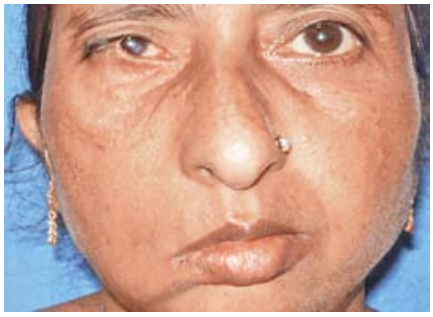


Fig. 1.78.5: Permanent tarsorrhaphy in operated CPA tumor

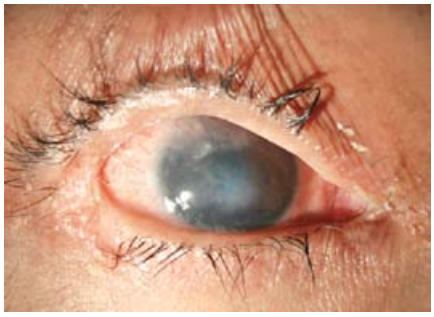


Fig. 1.78.6A: Permanent lateral tarsorrhaphy—RE (severe dry eye)

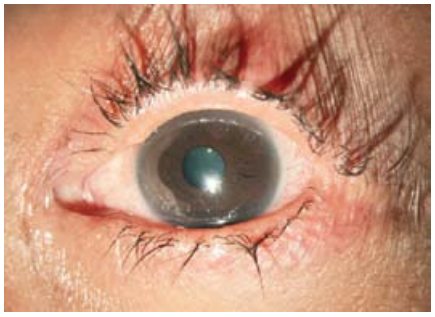


Fig. 1.78.6B: Permanent lateral tarsorrhaphy—RE (severe dry eye)

CHAPTER

2 Diseases of the Conjunctiva

CHAPTER OUTLINE

Signs of Conjunctival Diseases

- Hyperemia of the conjunctiva
- Congestion
- Subconjunctival hemorrhage
- Edema (chemosis)
- Follicles
- Papillae
- Membranes
- Preauricular lymphadenopathy

Acute Conjunctivitis

- Acute mucopurulent conjunctivitis
- Purulent conjunctivitis
- Ophthalmia neonatorum
- Membranous conjunctivitis
- Angular conjunctivitis
- Acute hemorrhagic conjunctivitis
- Acute follicular conjunctivitis

Chronic Conjunctivitis

- Simple chronic conjunctivitis
- Trachoma

Allergic Conjunctivitis

- Phlyctenular conjunctivitis
- Vernal conjunctivitis (keratoconjunctivitis) or VKC
- Atopic conjunctivitis
- Acute allergic conjunctivitis
- Giant papillary conjunctivitis (GPC)

Conjunctival Degenerations

- Concretion (lithiasis)
- Pinguecula
- Pterygium

Benign Lesions

- Conjunctival cysts
- Epibulbar limbal dermoid
- Dermolipoma (lipodermoid)
- Choristomas

Malignant Lesions

- Conjunctival intraepithelial neoplasia (CIN)
- Invasive squamous cell carcinoma

Pigmented Lesions

- Flat superficial pigmentation
- Benign epithelial melanosis
- Benign subepithelial melanocytosis
- Simple nevus
- Melanocytoma
- Precancerous melanosis
- Malignant melanoma

Other Conjunctival Conditions

- Subconjunctival hemorrhage
- Conjunctival xerosis
- Bitot's spot
- Keratoconjunctivitis sicca (KCG)
- Mucus fishing syndrome
- Lid imbrication syndrome
- Ocular cicatricial pemphigoid (OCP)
- Stevens-Johnson syndrome (SJS)
- Superior limbic keratoconjunctivitis (SLK)
- Vascular malformations
- Non-Hodgkin's lymphoma
- Amyloidosis of conjunctiva
- Conjunctivochalasis
- Conjunctival granulomas
- Limbal stem cell deficiency (LSCD)
- Xeroderma pigmentosa
- Metastatic tumors of the conjunctiva

Signs of Conjunctival Diseases

Hyperemia of the Conjunctiva

- Passive dilatation of the conjunctival blood vessels (**Fig. 2.1.1**)
- Without exudation or cellular infiltration
- Mainly occur due to irritation
- Only redness, no other symptoms (**Fig. 2.1.2**)
- Temporary blanching is noted with 1 in 1000 epinephrine solution.

Congestion

- ***Conjunctival congestion:***
  - Bright red, superficial vessels more intense at the fornices (**Fig. 2.2.1**)
  - Branched dichotomously (**Fig. 2.2.2**)
  - May be diffused or localized (**Figs 2.2.3A and B**)
  - Seen in different types of conjunctivitis.
- ***Ciliary congestion:***
  - Deeper vessels, dusky red in color and seen mostly at limbus
  - Arranged in radial fashion
  - Seen in keratitis (**Fig. 2.2.4**), iridocyclitis (**Fig. 2.2.5**) or angle closure glaucoma (**Fig. 2.2.6**).

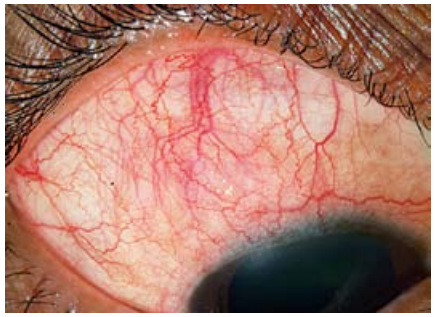


Fig. 2.1.1: Conjunctival hyperemia

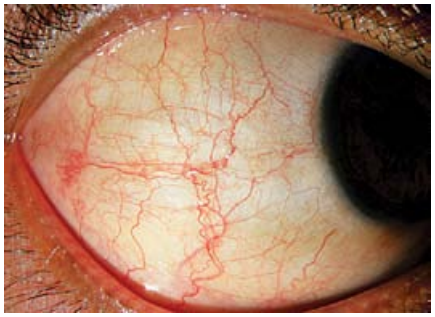


Fig. 2.1.2: Conjunctival hyperemia

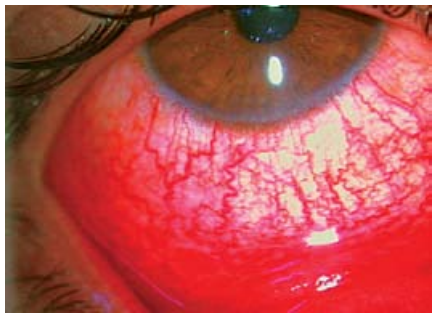


Fig. 2.2.1: Conjunctival congestion—more in fornix

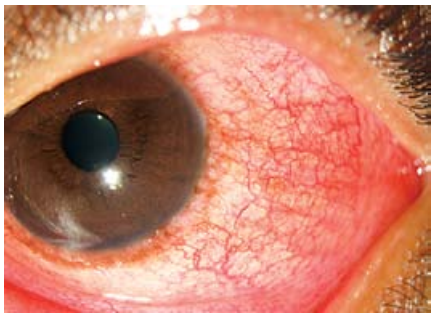


Fig. 2.2.2: Conjunctival congestion—bulbar

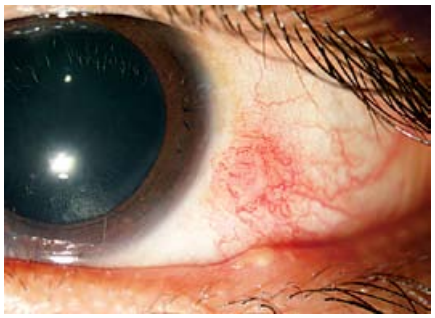


Fig. 2.2.3A: Localized conjunctival congestion—meibomianitis

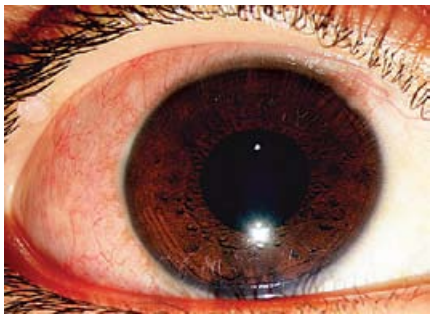


Fig. 2.2.3B: Localized conjunctival congestion—molluscum contagiosum

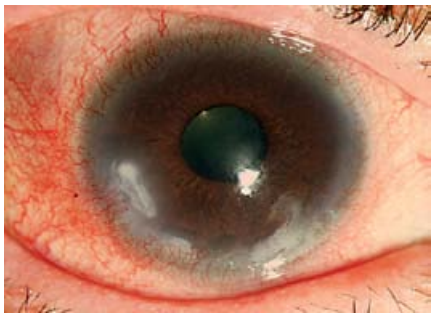


Fig. 2.2.4: Ciliary congestion—keratitis

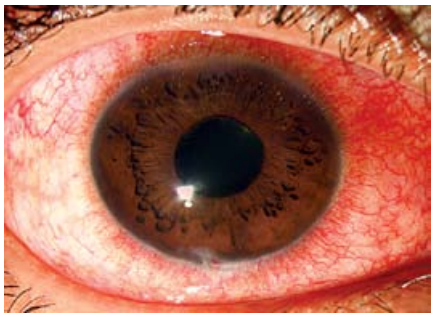


Fig. 2.2.5: Ciliary congestion—acute iridocyclitis

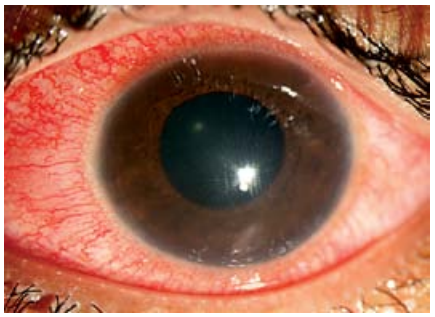


Fig. 2.2.6: Ciliary congestion—acute attack in angle closure glaucoma

Subconjunctival Hemorrhage

- Acute hemorrhagic conjunctivitis (*picorna* virus) (**Fig. 2.3.1**)
- Adenoviral conjunctivitis
- Bacterial conjunctivitis (*Pneumococcus* or *Haemophilus* spp.)
- Appears as petechial (**Fig. 2.3.2**) or frank subconjunctival hemorrhage.

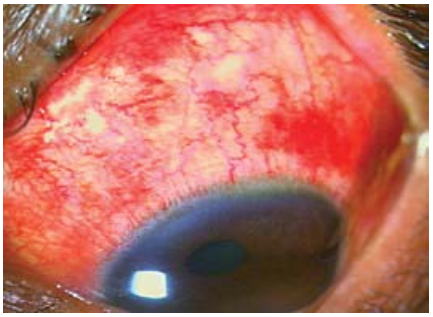


Fig. 2.3.1: Subconjunctival hemorrhage—conjunctivitis

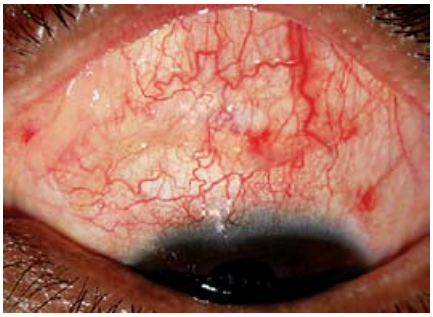


Fig. 2.3.2: Subconjunctival petechial hemorrhage—in conjunctivitis

Edema (Chemosis)

- Large quantity of the exudates causes ballooning of the bulbar conjunctiva **(Fig. 2.4.1)**
- Lids are often edematous
- Seen in severe bacterial conjunctivitis, orbital inflammation **(Fig. 2.4.2)** and acute allergic inflammation **(Fig. 2.4.3)**.

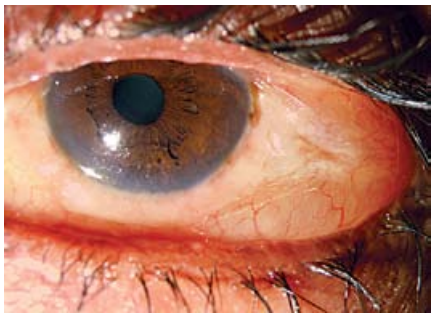


Fig. 2.4.1: Chemosis of conjunctiva

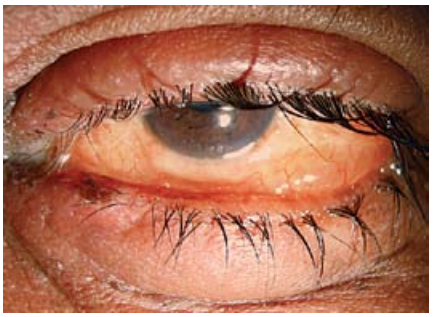


Fig. 2.4.2: Chemosis—with lid edema

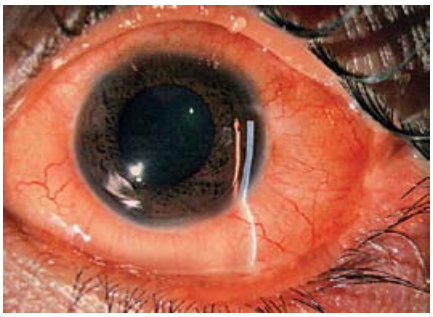


Fig. 2.4.3: Chemosis—allergic conjunctivitis

Follicles

- Round swellings (0.5-2 mm in diameter)
- Each follicle is encircled by tiny blood vessels
- Mostly seen in fornices and palpebral conjunctiva **(Figs 2.5.1 and 2.5.2)**
- Seen in follicular conjunctivitis, trachoma, toxin or drug-induced.

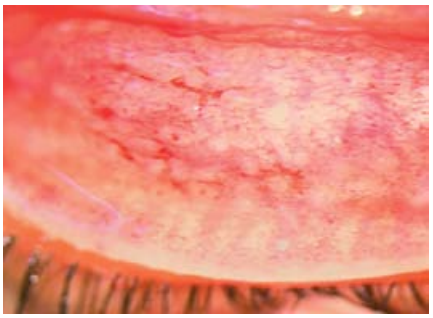


Fig. 2.5.1: Conjunctival follicles—fornicial

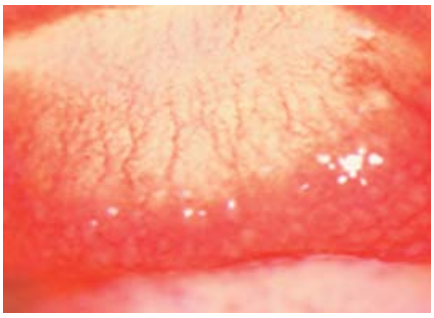


Fig. 2.5.2: Conjunctival follicles—upper palpebral

Papillae

- Vascular structures invaded by inflammatory cells
- Hyperplasia of the normal system of vascularization (**Fig. 2.6.1**)
- They are hyperemic flat-topped elevations on the tarsal conjunctiva
- It may vary from simple papillary hyperplasia to giant papillae
- Seen in allergic conjunctivitis, trachoma and giant papillary conjunctivitis
- Small papillae along the medial aspect of upper tarsal conjunctiva is a normal finding
- *Grades of papillae:*
  - *Grade 0* = Normal tarsal conjunctiva, no papilla (**Fig. 2.6.2**)
  - *Grade 1* = Multiple small papillae with smooth velvety appearance (**Fig. 2.6.3**)
  - *Grade 2* = Micropapillae, each with a diameter of 0.3–1.0 mm (**Fig. 2.6.4**)
  - *Grade 3* = Giant papillae, each with a diameter of >1.0 mm (**Fig. 2.6.5**)
  - *Grade 4* = Large enormous protruding papillae (**Fig. 2.6.6**).

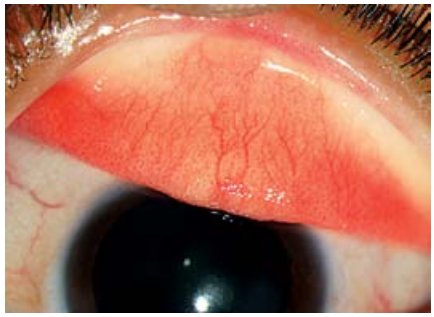


Fig. 2.6.1: Conjunctival papillae

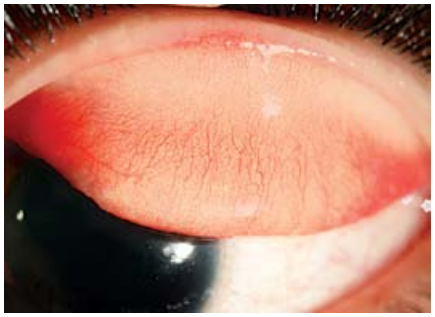


Fig. 2.6.2: Papillae—Grade 0

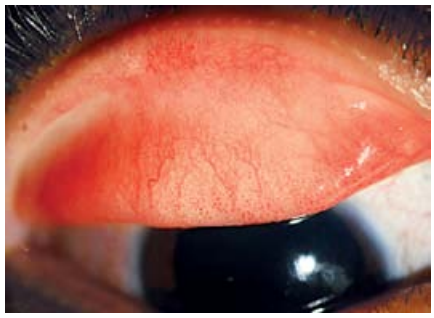


Fig. 2.6.3: Papillae—Grade 1

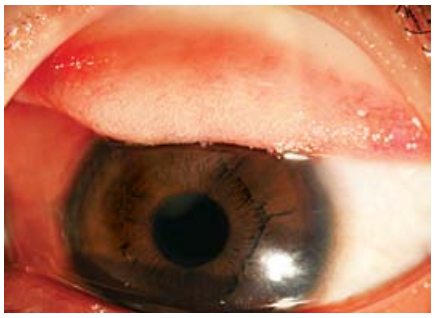


Fig. 2.6.4: Papillae—Grade 2

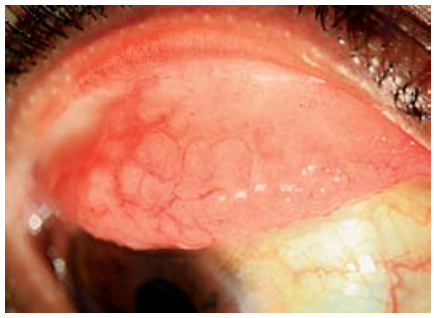


Fig. 2.6.5: Papillae—Grade 3

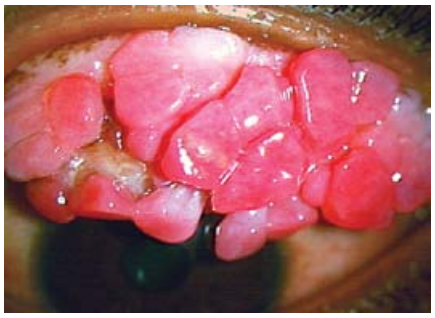


Fig. 2.6.6: Papillae—Grade 4

Membranes

- *True membranes* (**Fig. 2.7.1**):
  - Seen in certain bacterial conjunctivitis, especially in diphtheria
  - Attempt to remove the membrane may cause bleeding
- *Pseudomembranes* (**Fig. 2.7.2**):
  - Coagulated exudates loosely adherent to the inflamed conjunctiva (**Figs 2.7.3A and B**)
  - Can be easily peeled off without bleeding
  - Seen in adenoviral infection, vernal conjunctivitis and other inflammation.

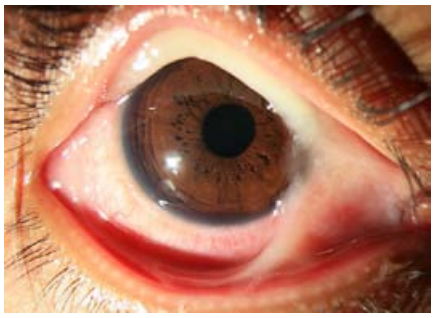


Fig. 2.7.1: True membrane

Fig. 2.7.2: Pseudomembrane

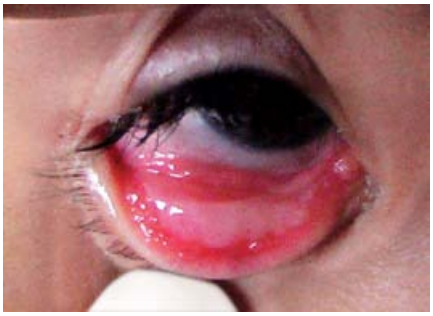


Fig. 2.7.3A: Pseudomembrane

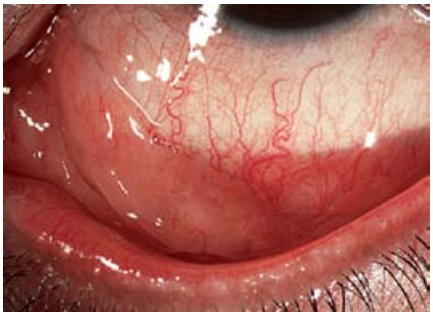


Fig. 2.7.3B: Pseudomembrane

Preauricular Lymphadenopathy

- Mostly seen in *Viral* and *Chlamydial* infections of conjunctiva
- Sometimes, it may be tender.

Acute Conjunctivitis

Acute Mucopurulent Conjunctivitis

- Marked hyperemia
- Lid edema, matting of the eyelashes **(Fig. 2.8.1)**
- Chemosis of the conjunctiva, petechial hemorrhage **(Fig. 2.8.2)**
- Mucopurulent discharge or flakes of mucopus **(Fig. 2.8.3)**
- May be associated with acute blepharitis **(Fig. 2.8.4)**
- *Treatment:*
  - Frequent eye wash with luke-warm water
  - Broad-spectrum antibiotic eye drops
  - Antibiotic ointment at night
  - Prevention of contamination and spread of disease.

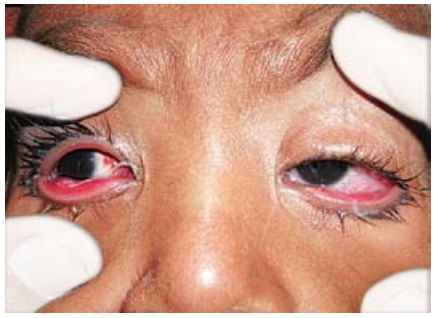


Fig. 2.8.1: Acute mucopurulent conjunctivitis

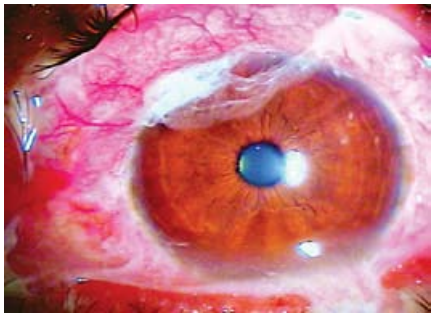


Fig. 2.8.2: Acute mucopurulent conjunctivitis—chemosis

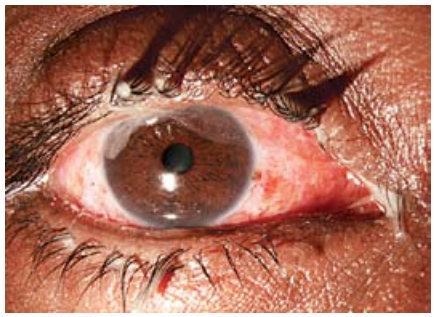


Fig. 2.8.3: Acute mucopurulent conjunctivitis—mucus flakes

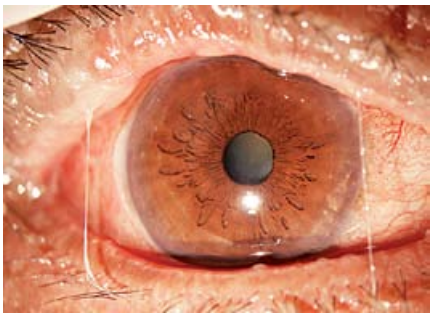


Fig. 2.8.4: Acute blepharoconjunctivitis

Purulent Conjunctivitis

- Copious purulent discharge (**Figs 2.9.1A and B**)
- Right eye is commonly involved in male patient (**Fig. 2.9.2**)
- Lid edema and crusting of eyelashes (**Fig. 2.9.3**)
- Conjunctival chemosis with or without membrane formation
- Cornea may be involved in severe cases (**Fig. 2.9.4**)

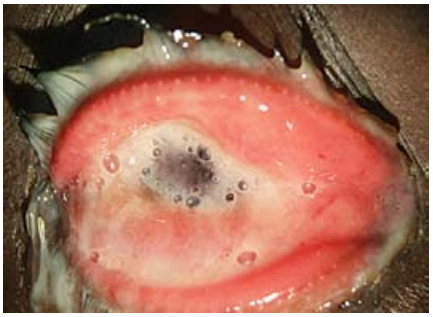


Fig. 2.9.1A: Purulent conjunctivitis—adult—RE

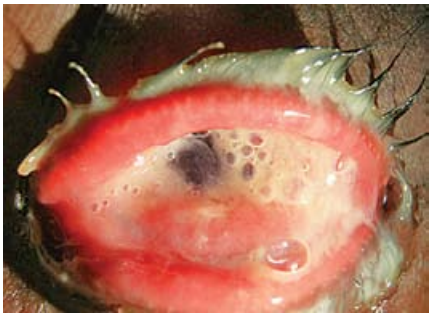


Fig. 2.9.1B: Purulent conjunctivitis—adult—LE

- *Treatment:*
  - Patient should be kept in isolation
  - Frequent irrigation of eyes with lukewarm normal saline
  - Injection ceftriaxone (IM)—1.0 gm/daily × 5 days
  - Crystalline penicillin (1 in 10,000) eye drop or ciprofloxacin eye drop hourly
  - Ciprofloxacin eye ointment at night.

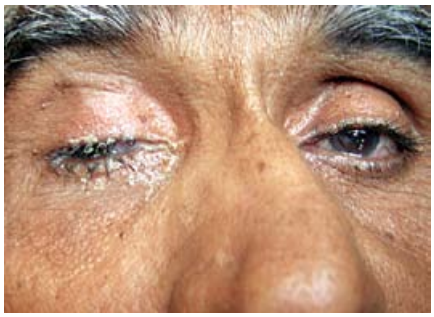


Fig. 2.9.2: Purulent conjunctivitis—affected—RE

Fig. 2.9.3: Purulent conjunctivitis

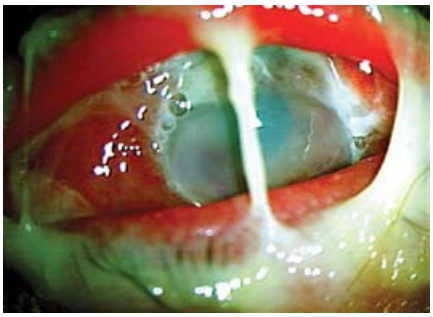


Fig. 2.9.4: Purulent conjunctivitis—corneal involvement

Ophthalmia Neonatorum

- In *Gonococcal* infection, hyperacute purulent conjunctivitis **(Figs 2.10.1A and B)**
- In other cases, it is a catarrhal, or mucopurulent conjunctivitis **(Fig. 2.10.2)**
- *Treatment:*
  - Ciprofloxacin or penicillin (1 in 5,000) eye drop hourly and ciprofloxacin ointment at night
  - Injection ceftriaxone (IM)—50 mg/kg/day × 5 days
  - For *Chlamydia*—sulfacetamide (10%) eye drop—4 times daily
  - *Prevention by:* Proper antenatal care, asepsis during delivery, antibiotic eye drop.

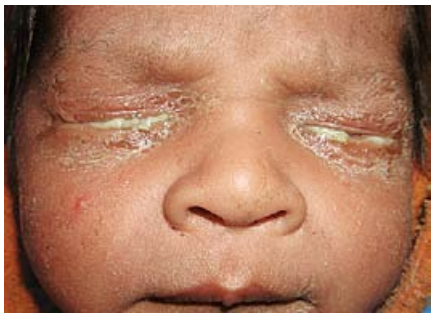


Fig. 2.10.1A: Neonatal gonococcal conjunctivitis

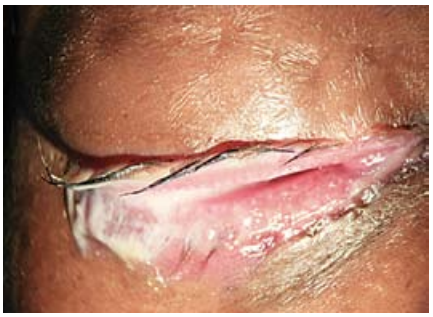


Fig. 2.10.1B: Neonatal gonococcal conjunctivitis

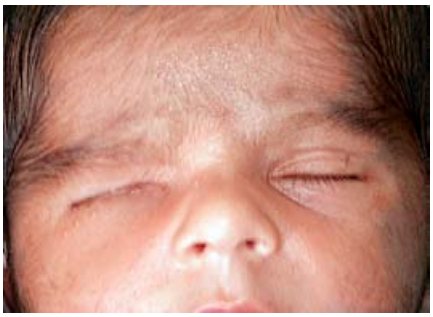


Fig. 2.10.2: Neonatal conjunctivitis—RE

Membranous Conjunctivitis

- Edema of the lids
- Mucopurulent or sanious discharge
- Thick white or grayish-yellow membrane on the palpebral conjunctiva (**Fig. 2.11.1**)
- Bleeding is very common on removal of the membrane
- Symblepharon may occur in late stage (**Fig. 2.11.2**)
- *Treatment:* Isolation, crystalline penicillin, antidiphtheric serum, erythromycin eye ointment.

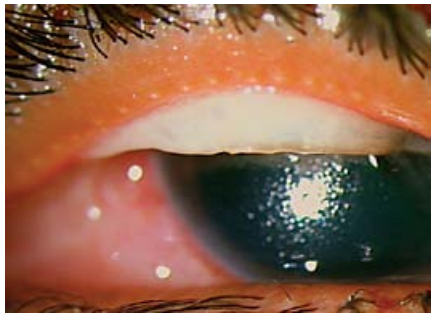


Fig. 2.11.1: Membranous conjunctivitis

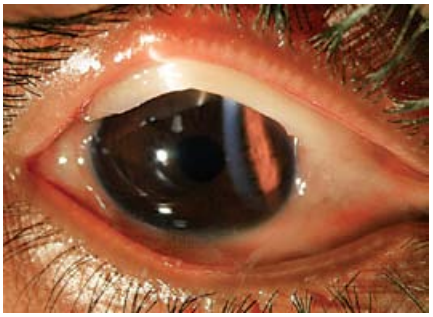


Fig. 2.11.2: Membranous conjunctivitis—symblepharon formation

Angular Conjunctivitis

- Conjunctival inflammation is limited to inner marginal strip, especially at the outer (**Fig. 2.12.1)** or inner canthi (**Fig. 2.12.2)**
- Excoriation of the skin at the outer or inner canthi
- Congestion of adjacent bulbar conjunctiva
- Caused by organism—*Moraxella axenfeld*
- *Treatment:* Zinc oxide containing eye drop and oxytetracycline (1%) eye ointment at night.

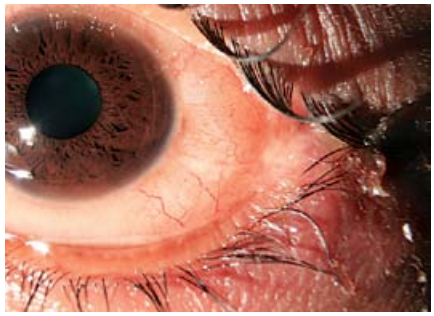


Fig. 2.12.1: Angular conjunctivitis—outer canthus

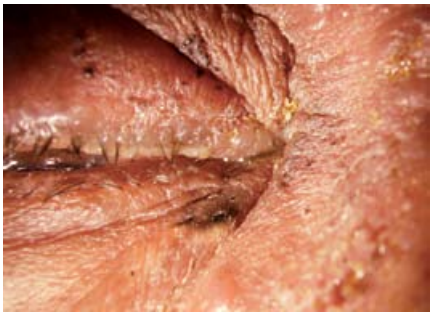


Fig. 2.12.2: Angular conjunctivitis—innner canthus

Acute Hemorrhagic Conjunctivitis

- Associated with subconjunctival hemorrhage and may be with frank serosanguinous or sanguinous discharge (**Figs 2.13.1 and 2.13.2)**
- Caused by *Picorna* virus, *Coxackie* and *Enterovirus – 70.*
- The symptoms and signs are similar to adenoviral infection
- *Treatment:* Similar to adenoviral infection.

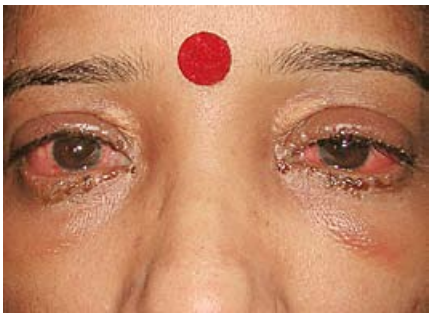


Fig. 2.13.1: Hemorrhagic conjunctivitis—serosanguinous discharge

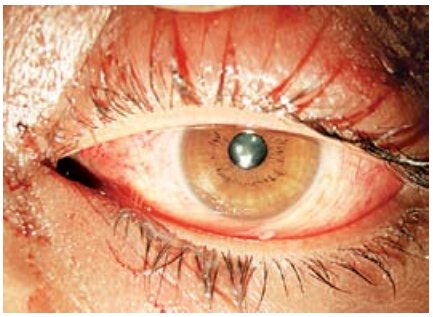


Fig. 2.13.2: Hemorrhagic conjunctivitis—sanguinous discharge

Acute Follicular Conjunctivitis

- Inflammation of the conjunctiva with appearance of follicles (**Figs 2.14.1 and 2.14.2)**
- Always a tendency of corneal involvement, the typical lesion is a keratoconjunctivitis **(Fig. 2.14.3)** and petechial hemorrhage
- *Inclusion conjunctivitis:* Associated with superficial punctate keratitis (SPKs) and pannus formation
- *Epidemic keratoconjunctivitis (EKC):* Almost always associated with subepithelial infiltrates and SPKs **(Figs 2.14.4 and 2.14.5)**
- *Pharyngoconjunctival fever:* With fever and pharyngitis, but SPKs are rare **(Fig. 2.14.6)**
- *Acute herpetic keratoconjunctivitis:* Follicles are usually large and SPKs with small dendritic lesion on the cornea
- *Treatment:* Astringents eye drop, sulfacetamide in inclusion conjunctivitis and acyclovir in herpetic infection; tear substitutes in corneal involvement.

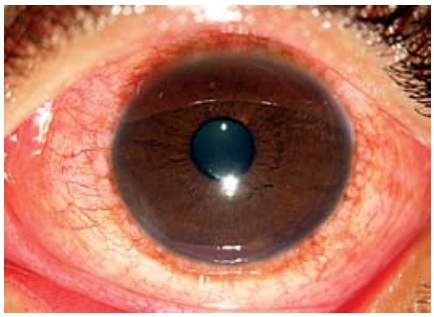


Fig. 2.14.1: Acute follicular conjunctivitis—limbal follicle

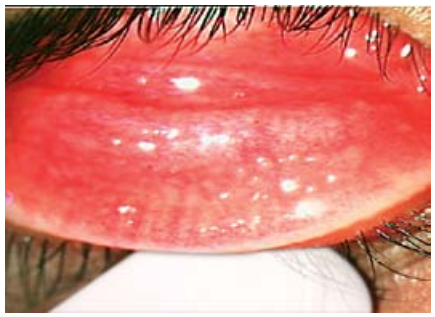


Fig. 2.14.2: Acute follicular conjunctivitis—palpebral follicle

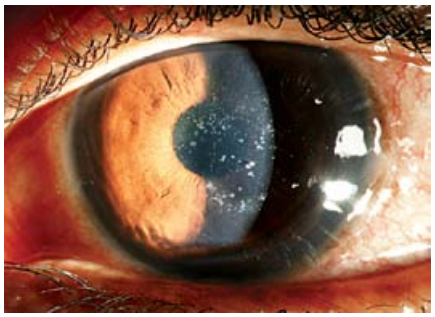


Fig. 2.14.3: Epidemic kerato-conjunctivitis—corneal involvement

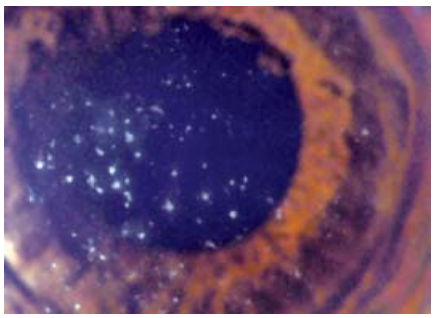


Fig. 2.14.4: Follicular conjunctivitis—corneal involvement

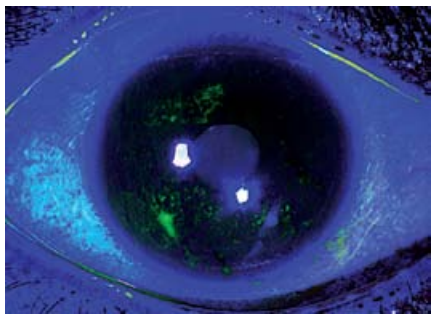


Fig. 2.14.5: Epidemic kerato-conjunctivitis—corneal involvement

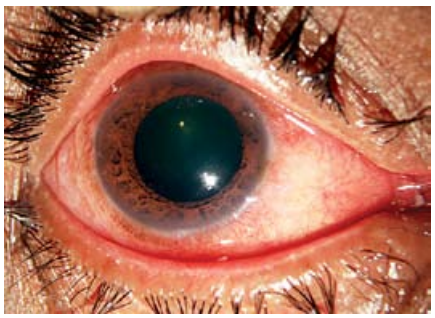


Fig. 2.14.6: Acute viral conjunctivitis—pharyngoconjunctival fever

Chronic Conjunctivitis

Simple Chronic Conjunctivitis

- Redness, more in the evening
- Lower fornix is congested
- Thick frothy discharge with palpebral congestion (**Figs 2.15.1 and 2.15.2)**
- Associated with meibomianitis
- *Treatment:* Antibiotic drop and ointment, treatment of meibomianitis.

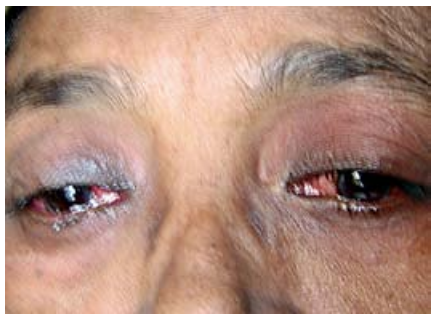


Fig. 2.15.1: Chronic conjunctivitis

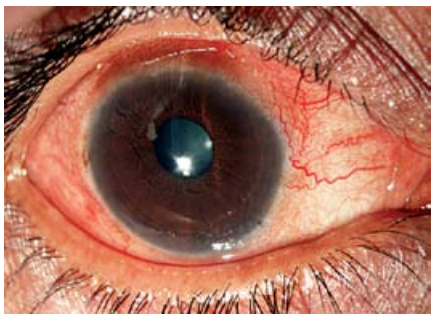


Fig. 2.15.2: Chronic conjunctivitis

Trachoma

Chronic inflammation of the conjunctiva and the cornea, characterized by the presence of follicles and papillary hypertrophy of the conjunctiva, with pannus formation over upper part of the cornea caused by *Chlamydia trachomatis*

The features are:

- Bulbar congestion
- Velvety papillary hypertrophy **(Fig. 2.16.1)**
- *Follicles*—mostly seen in upper tarsal conjunctiva **(Fig. 2.16.2)**
- *Pannus*—mainly seen at the upper limbus and upper part of cornea **(Figs 2.16.3 and 2.16.4)**
- *Herbert's pits* at the limbus—a pathognomonic sign **(Fig. 2.16.5)**
- *Scarring* (Arlt's line) of upper tarsal conjunctiva **(Fig. 2.16.6)**
- *A blinding condition* mainly due to trichiasis, entropion and subsequent corneal opacification
- It is included in *Vision-2020 program*
- *Treatment:* Sulfacetamide (20% or 30%)—4 times daily, tetracycline eye ointment at night; oral tetracycline or single dose of oral azithromycin
- *Prevention:* 'SAFE' strategy to prevent trachoma blindness:
  - S = Surgery for trichiasis and entropion
  - A = Antibiotics
  - F = Facial cleanliness
  - E = Environmental sanitation.

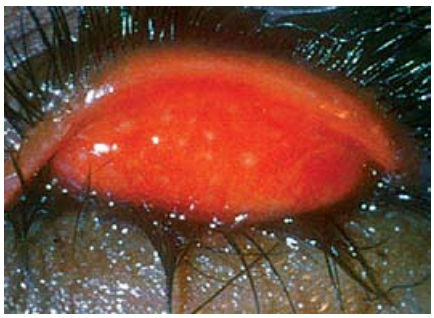


Fig. 2.16.1: Trachoma—papillary hypertrophy and follicle

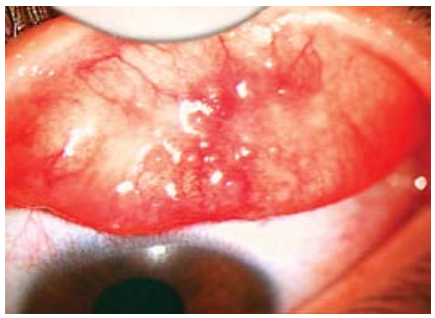


Fig. 2.16.2: Trachoma follicles

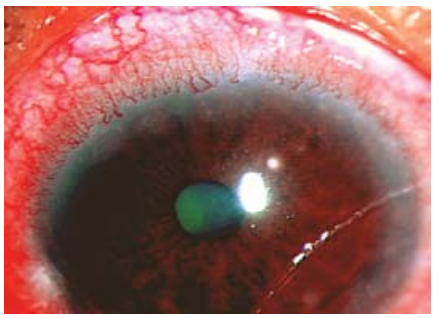


Fig. 2.16.3: Trachomatous pannus

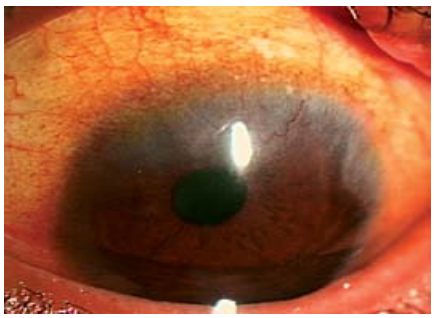


Fig. 2.16.4: Healed trachomatous pannus

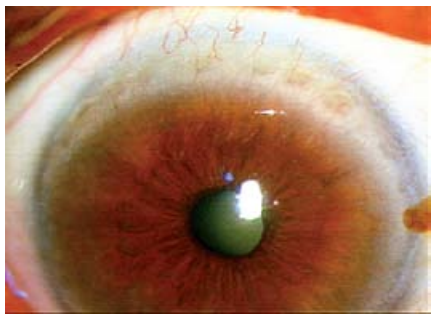


Fig. 2.16.5: Herbert's pit in upper limbus

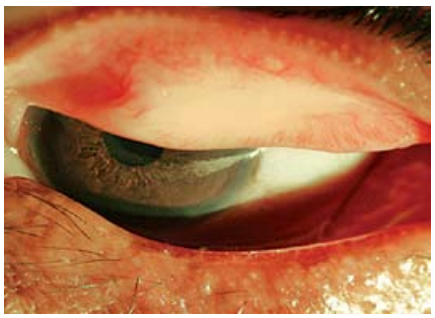


Fig. 2.16.6: Trachoma scar—Arlt's line

Allergic Conjunctivitis

Phlyctenular Conjunctivitis

- Allergic conjunctivitis caused by endogenous bacterial toxin
- Characterized by bleb or nodule formation near or at the limbus in young children (**Fig. 2.17.1**)
- Pinkish-white or gray in color, 1–3 mm in diameter
- Localized bulbar congestion surrounding the nodule (**Fig. 2.17.2**)
- Multiple, recurrent phlycten may be seen in tuberculous infection (**Fig. 2.17.3**)
- Bleb may rupture to form phlyctenular ulcer which may spread towards cornea leading to fascicular ulcer (**Figs 2.17.4 and 2.17.5**)
- It may be of *three types:*
  - *Phlyctenular conjunctivitis:* When the conjunctiva alone is involved and a large phlycten may cause localized corneal dellen **(Figs 2.17.6A and B)**
  - *Phlyctenular keratoconjunctivitis*: When at the limbus, and involves both the conjunctiva and cornea
  - *Phlyctenular keratitis:* When cornea alone is involved (rare)
- *Treatment:* Corticosteroid eye drops, investigation and general measures.

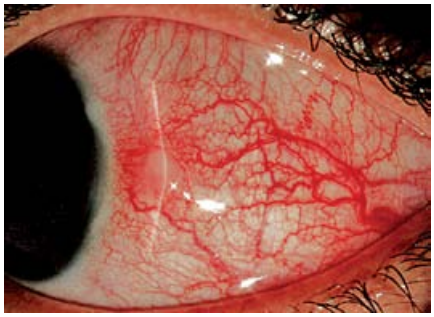


Fig. 2.17.1: Conjunctival phlycten

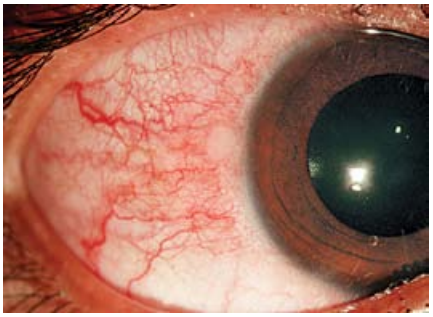


Fig. 2.17.2: Limbal phlycten

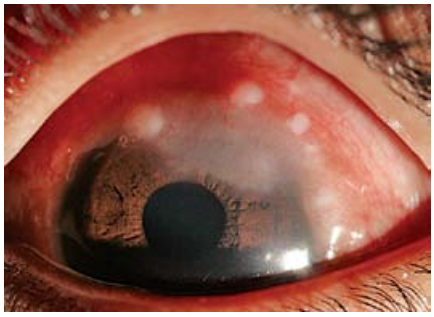


Fig. 2.17.3: Multiple phlyctens with pannus

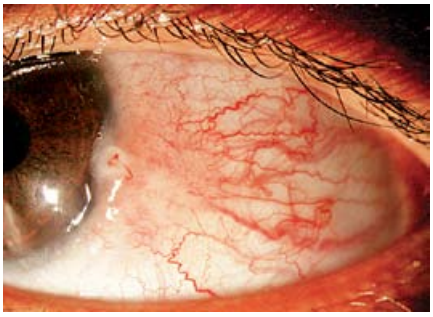


Fig. 2.17.4: Phlyctenular corneal ulcer

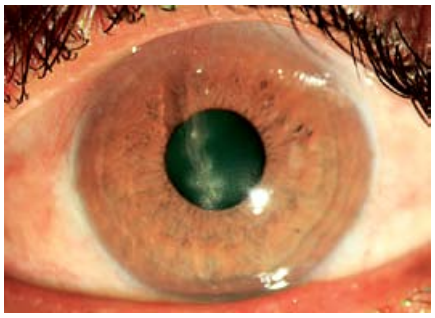


Fig. 2.17.5A: Fascicular ulcer

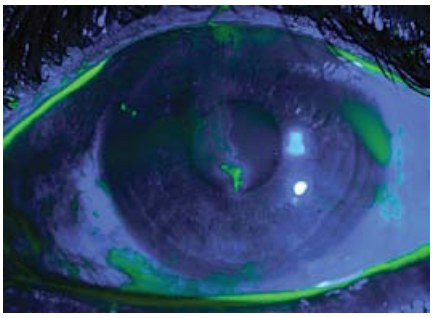


Fig. 2.17.5B: Fascicular ulcer—fluorescein stain

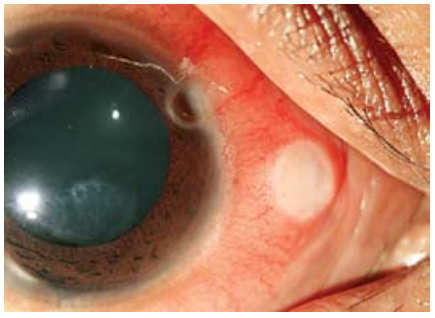


Fig. 2.17.6A: Conjunctival phlycten with corneal dellen

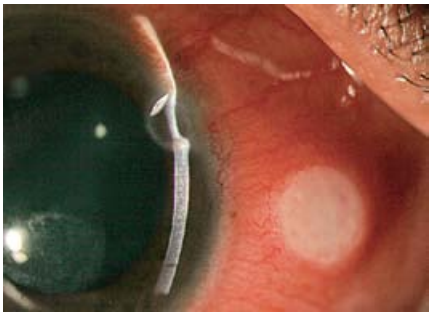


Fig. 2.17.6B: Conjunctival phlycten with corneal dellen

Vernal Conjunctivitis (Keratoconjunctivitis) or VKC

- Bilateral, recurrent, seasonal allergic conjunctivitis in children caused by exogenous allergens
- Presents in one of the *three forms*: palpebral, bulbar and mixed
- *Palpebral:*
  - *Cobble-stone papillae:* Papillary hypertrophy of upper palpebral conjunctiva **(Fig. 2.18.1)**
  - The bluish-white papillae are separated by white connective tissue septa
- *Bulbar:*
  - Multiple, small, nodule-like gelatinous thickening around the limbus, mostly at the upper part **(Fig. 2.18.2)**
  - May be associated with palpebral type, called *mixed type* **(Fig. 2.18.3)**
  - Typical conjunctival pigmentation **(Fig. 2.18.4)** more seen in dark pigmented races
  - Discrete superficial white spots, called Horner-Tranta's dots **(Fig. 2.18.5)** and micropannus **(Fig. 2.18.6)** around the upper limbus
- *Pseudogerontoxon*—resembles an arcus senilis with appearance of *'cupid's bow'* **(Fig. 2.18.7)**
- Epithelial microerosions—leading to corneal ulceration *(shield ulcer)* **(Figs 2.17.8A and B)**
- In severe cases, chronic limbitis may cause 360 degrees limbal cell deficiency **(Figs 2.18.9 and 2.18.10)**
- Patients with vernal catarrh also have a higher incidence of keratoconus **(Fig. 2.18.11)**
- *Treatment:* Topical corticosteroids, antihistaminics and mast-cell stabilizer eye drop
- Steroid-induced cataract and glaucoma are not so uncommon problem in severe cases **(Fig. 2.18.12)**.

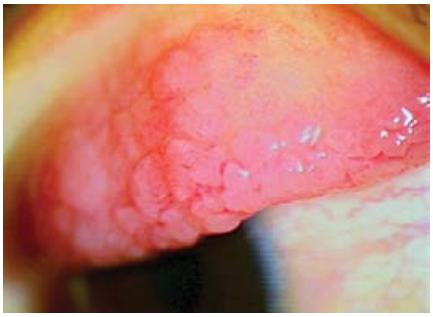


Fig. 2.18.1: VKC—palpebral type—cobble stone papillae

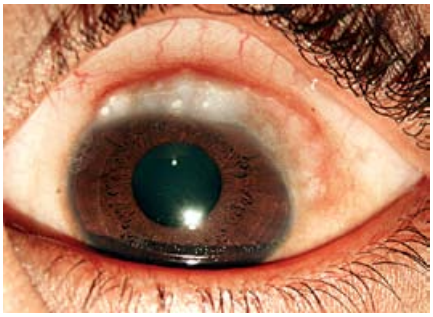


Fig. 2.18.2: Vernal conjunctivitis—limbal type

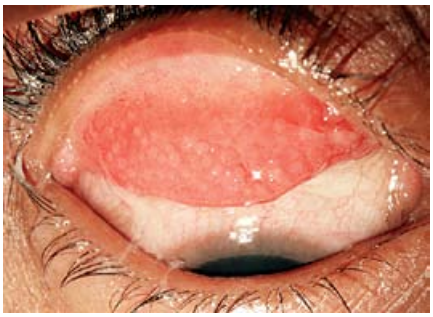


Fig. 2.18.3: Vernal conjunctivitis—mixed type

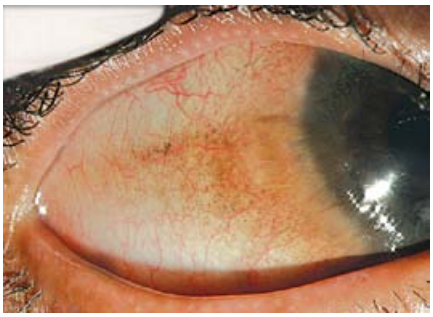


Fig. 2.18.4: VKC—conjunctival pigmentation

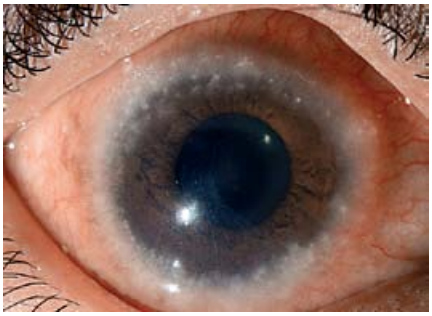


Fig. 2.18.5: VKC—Horner-Tranta's dots

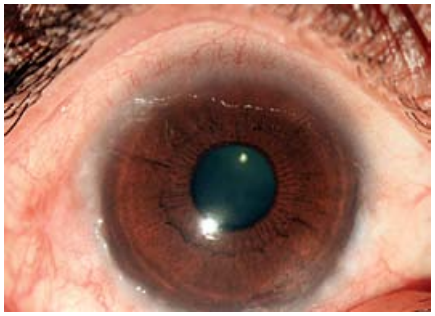


Fig. 2.18.6: VKC—Limbal micropannus

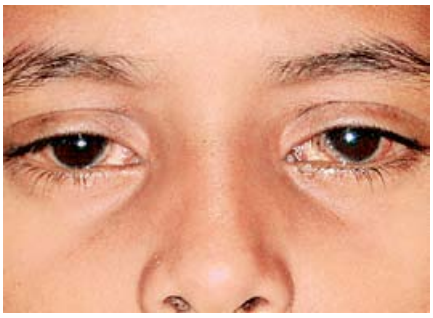


Fig. 2.18.7: VKC—pseudogerontoxon (cupid's bow)—BE

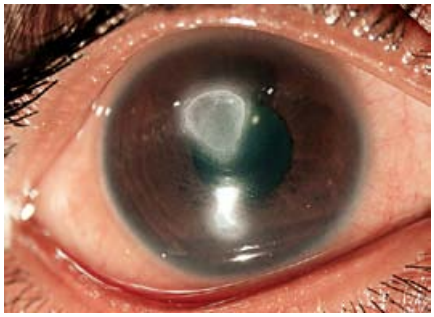


Fig. 2.18.8A: VKC—shield ulcer

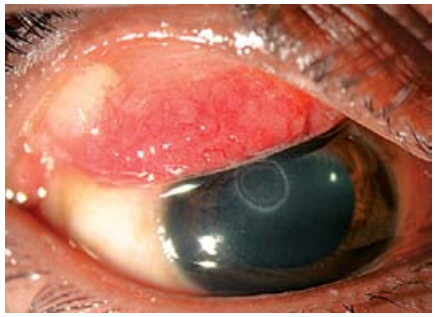


Fig. 2.18.8B: VKC—shield ulcer

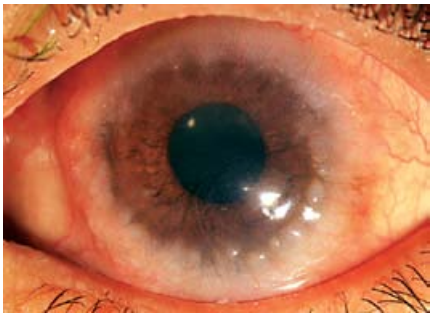


Fig. 2.18.9: VKC—severe limbal involvement with dry eye

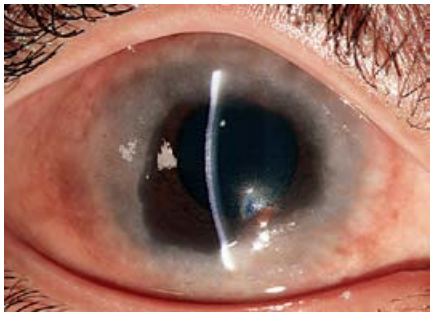


Fig. 2.18.10: VKC—severe limbal form with 360 degrees limbal cell deficiency

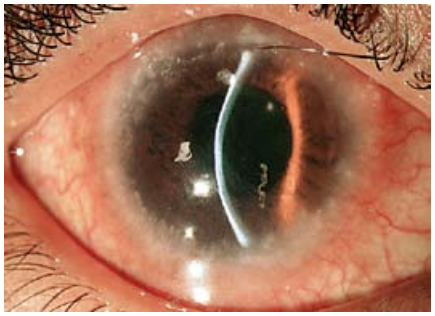


Fig. 2.18.11: Severe VKC with keratoconus

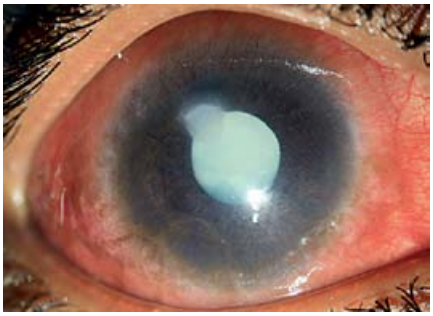


Fig. 2.18.12: Severe VKC with shield ulcer, dry eye and steroid-induced cataract

Atopic Conjunctivitis

It is seen in 30% of atopic dermatitis, especially with eczema. Perennial in character with some seasonal exacerbation.

- Severe itching, watering, swelling of lids and photophobia
- Thickened scaly dermatitis of the eyelids **(Figs 2.19.1A and B)**, conjunctival congestion, papillary reaction of the conjunctiva and superficial corneal vascularization. Hay fever, asthma and urticaria are often present
- *Treatment:* Tears substitutes, soft corticosteroid eye drops, tacrolimus (0.03%) ointment to treat lid condition.

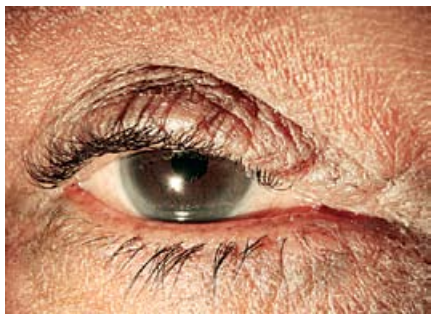


Fig. 2.19.1A: Atopic blepharo-conjunctivitis—RE

Fig. 2.19.1B: Atopic blepharo-conjunctivitis—LE

Acute Allergic Conjunctivitis

- Acute allergic reaction of the conjunctiva to dust, pollens, or other allergens
- Itching, watery discharge and a history of allergic diathesis
- Conjunctival chemosis, moderate to severe congestion, papillae and lid edema. No preauricular lymphadenopathy **(Figs 2.20.1A and B)**
- *Treatment:* Elimination of allergen; cold compress several times; frequent preservative-free tears substitutes; antihistaminic eye drops; olopatadine (0.1%), epinastine (0.05%) or ketotifen eye drops twice daily. In severe cases, dilute (1/10th dilution of dexamethasone) or soft steroid (fluoromethalone or loteprednol) eye drops for 1–2 weeks and oral antihistaminics.

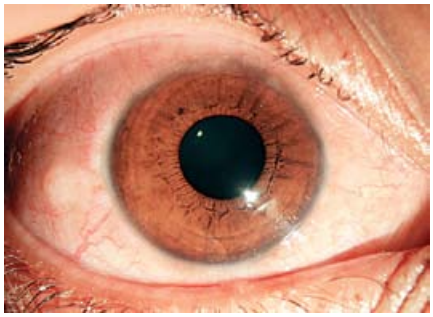


Fig. 2.20.1A: Seasonal allergic conjunctivitis—RE

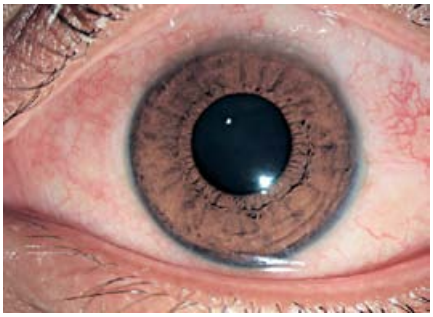


Fig. 2.20.1B: Seasonal allergic conjunctivitis—LE

Giant Papillary Conjunctivitis (GPC)

- Foreign body associated allergic conjunctivitis with characteristic giant papillae (>1 mm)
- Seen among soft contact lens wearer, artificial eye wearer and postoperative patient with protruding ends of monofilament nylon sutures
- Clinical picture is similar to palpebral type of vernal conjunctivitis with presence of giant papillae (**Fig. 2.21.1**)
- Dry eye is a common association, especially with contact lens wearer (**Figs 2.21.2A and B**)
- *Treatment:* Same as vernal conjunctivitis and avoidance of offending agents.

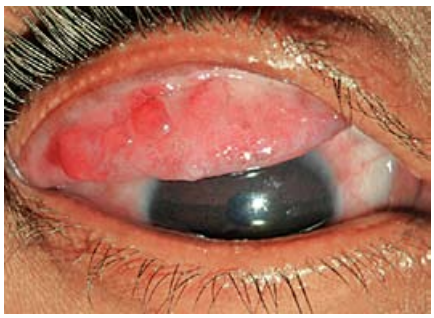


Fig. 2.21.1: Giant papillary conjunctivitis (GPC)

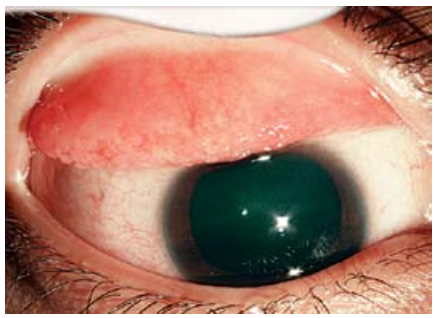


Fig. 2.21.2A: Giant papillary conjunctivitis with dry eye

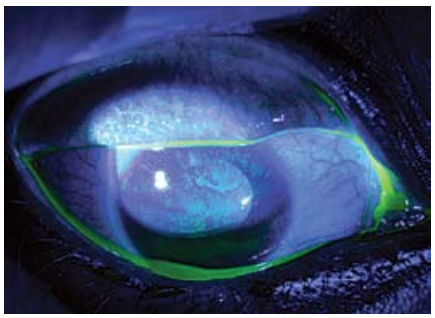


Fig. 2.21.2B: GPC with dry eye—fluorescein staining of cornea

Conjunctival Degenerations

Concretion (Lithiasis)

- Minute hard yellow/white spots usually in the upper palpebral conjunctiva (**Fig. 2.22.1**) and sometimes in lower tarsal conjunctiva (**Fig. 2.22.2**)
- They are not calcium deposits, but inspissated mucus and degenerated cells
- Sometimes, they may project and irritate the cornea
- *Treatment:* May be evacuated with a sharp needle.

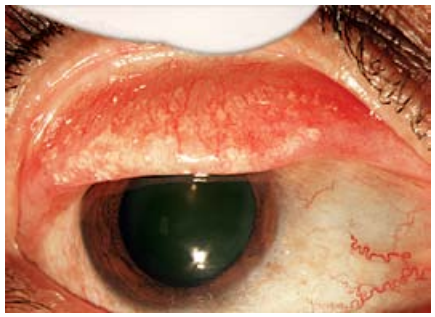


Fig. 2.22.1: Conjunctival concretions of upper palpebral conjunctiva

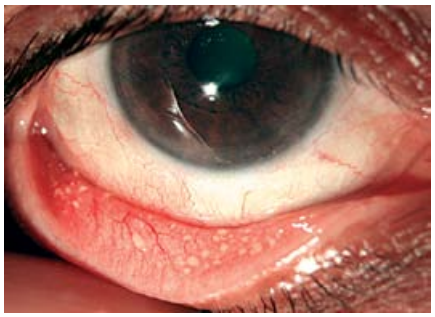


Fig. 2.22.2: Conjunctival concretions of lower palpebral conjunctiva

Pinguecula

- Yellowish, triangular deposit on the conjunctiva near the limbus at the palpebral aperture (**Fig. 2.23.1**)
- The apex is towards the cornea
- Affects the nasal side first, then the temporal
- Symptomless and does not require any treatment
- Sometimes, it may be inflamed (**Fig. 2.23.2**) and then treated with topical corticosteroids.

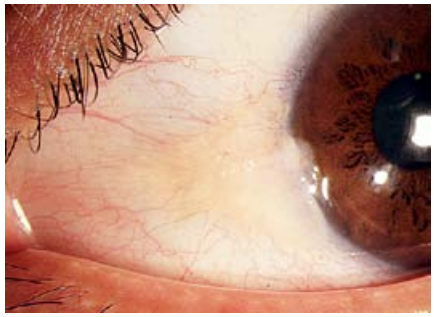


Fig. 2.23.1: Pinguecula

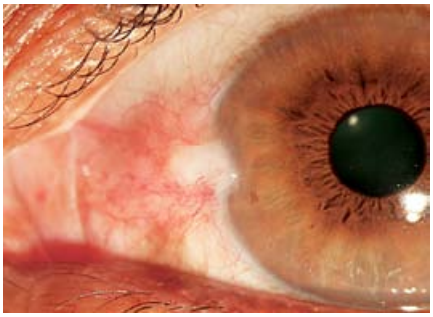


Fig. 2.23.2: Inflamed pinguecula

Pterygium

- Subconjunctival tissue proliferates as a triangular wing-shaped tissue-mass to invade the cornea, involving the Bowman's membrane and the superficial stroma
- Usually bilateral, may be asymmetrical, among elderly patients (**Fig. 2.24.1**)
- Usually involves the nasal limbus with variable extension onto the cornea (**Fig. 2.24.2**)
- Rarely it affects the temporal limbus alone (**Fig. 2.24.3**)
- It may be progressive (fleshy) and stationary (atrophic) types
- *Progressive (fleshy):*
  - Thick, fleshy with prominent vascularity
  - Increasing in size and encroaching towards the center of cornea (**Figs 2.24.4A and B**)

Fig. 2.24.1: Bilateral large pterygium

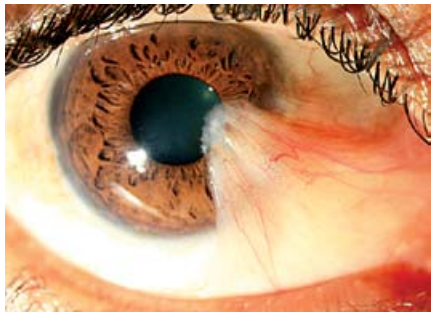


Fig. 2.24.2: Pterygium—nasal and progressive

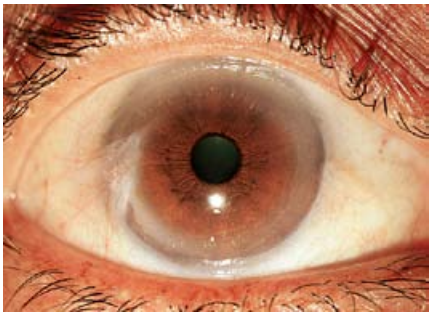


Fig. 2.24.3: Temporal pterygium

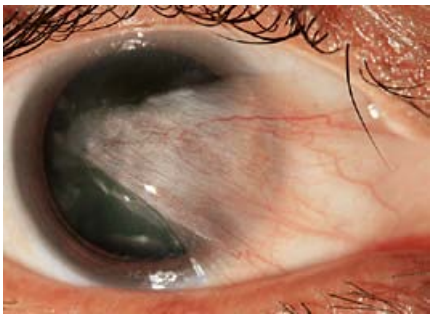


Fig. 2.24.4A: Pterygium—covering pupil—RE

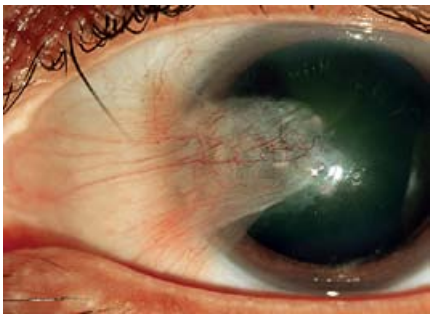


Fig. 2.24.4B: Pterygium—covering pupil—LE

- Opaque infiltrative spot is seen, known as the 'cap' (**Fig. 2.24.5**)
- *Stocker's line*—is seen on the corneal epithelium in front of the apex (**Fig. 2.24.6**)

- *Atrophic (stationary):*
  - Thin, attenuated with poor vascularity
  - No opaque spot (cap) is seen (**Fig. 2.24.7**)
- *Other types:*
  - *Primary double pterygium:* Both nasal and temporal limbus are involved (**Fig. 2.24.8)**
  - *Recurrent pterygium:* More scarring and sometimes much more wider (**Figs 2.24.9A to C**)
  - *Malignant pterygium:* Recurrent pterygium with symblepharon formation and may be associated with restriction of ocular movement on opposite side (**Figs 2.24.10A and B**)

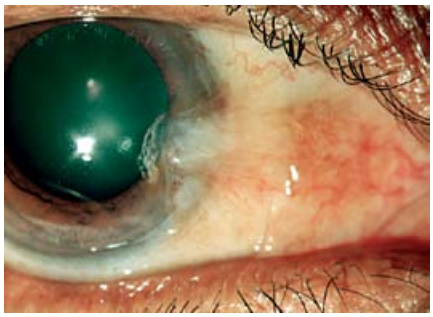


Fig. 2.24.5: Progressive pterygium—cap

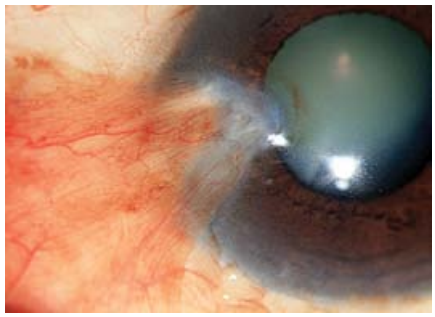


Fig. 2.24.6: Pterygium—Stocker's line

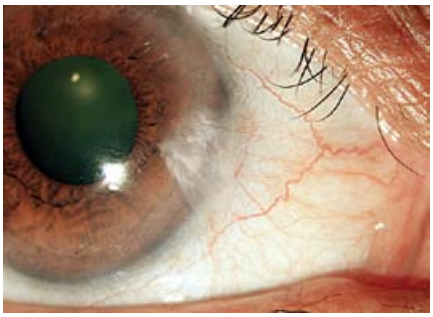


Fig. 2.24.7: Atrophic pterygium

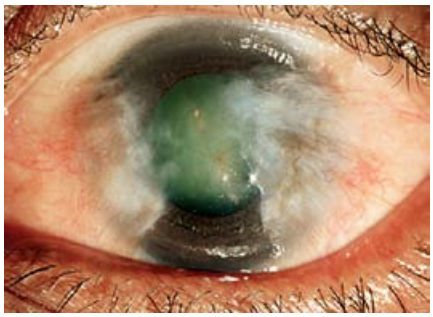


Fig. 2.24.8: Double headed pterygium with Stocker's lines

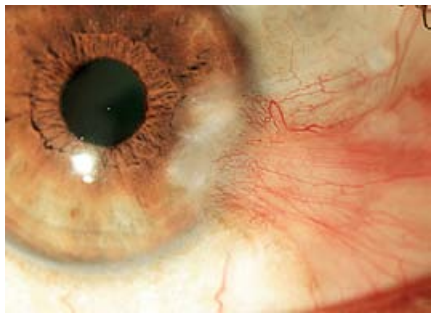


Fig. 2.24.9A: Recurrent pterygium—early

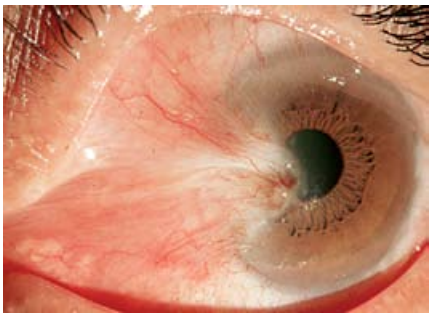


Fig. 2.24.9B: Recurrent pterygium—advanced with shortening of the fornix

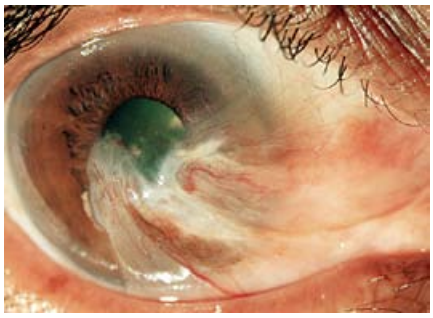


Fig. 2.24.9C: Recurrent pterygium—large

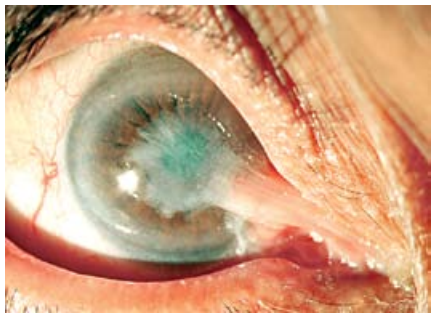


Fig. 2.24.10A: Recurrent pterygium with symblepharon

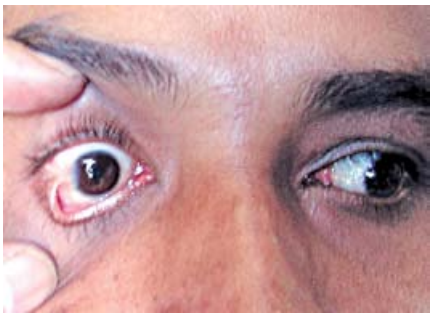


Fig. 2.24.10B: Recurrent pterygium—symblepharon with movement restriction

- Recurrence may sometimes be associated with pyogenic granuloma formation (**Fig. 2.24.11**)
- Primay pterygium may undergo cystic changes with or without corneal dellen (**Figs 2.24.12A and B**) and an eyelash may present underneath the pterygium (**Fig. 2.24.13**)

- *Treatment:* Excision of pterygial tissue (subconjunctival dissection) with conjunctival free/limbal autograft (CAG/CLAU) is the treatment of choice (**Figs 2.24.14A and B**). Treatment of the bare sclera by MMC and covering with amniotic membrane are the other options
- *Pseudopterygium*
  - Adhesion of a fold of conjunctiva to the peripheral cornea
  - Usually unilateral, stationary and at any meridian (**Figs 2.24.15 and 2.24.16**)
  - A probe can be passed easily beneath the neck of the pterygium *(Probe test)*
- *Treatment:* Simple excision.

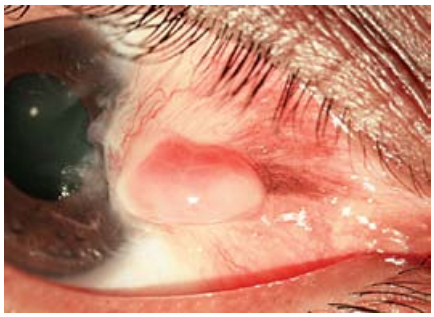


Fig. 2.24.11: Recurrent pterygium with pyogenic granuloma

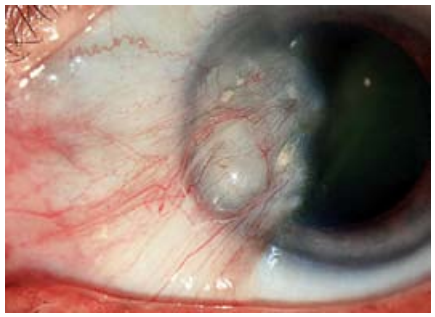


Fig. 2.24.12A: Pterygium with cystic changes

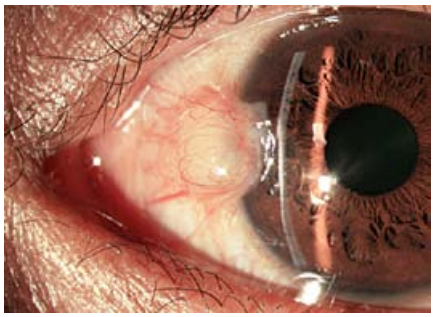


Fig. 2.24.12B: Pterygium—cystic changes with corneal dellen

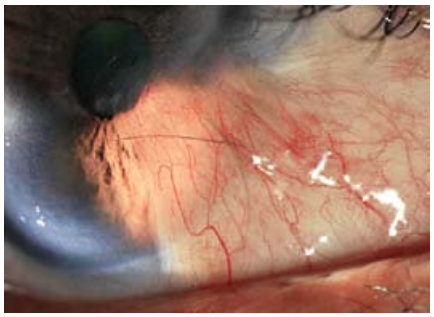


Fig. 2.24.13: Eyelash underneath the pterygium

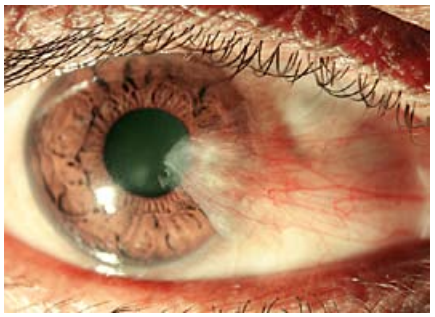


Fig. 2.24.14A: Progressive pterygium before surgery

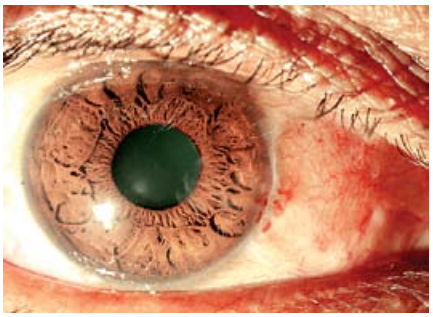


Fig. 2.24.14B: Pterygium resection with conjunctival autograft

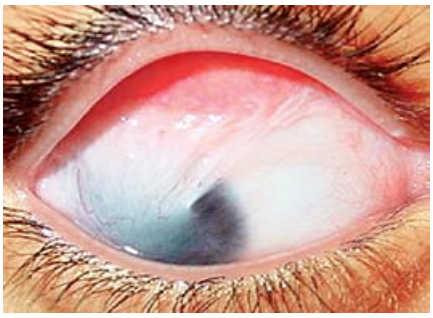


Fig. 2.24.15: Pseudopterygium

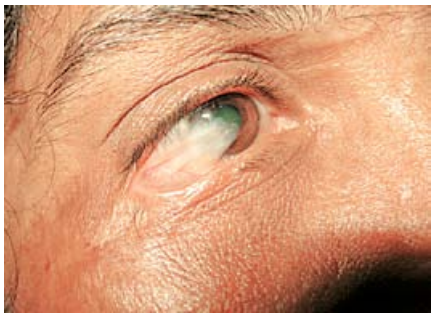


Fig. 2.24.16: Pseudopterygium with cicatricial ectropion following thermal burn

Benign Lesions

Conjunctival Cysts

- *Lymphangiectasia:* appears as dilatation of lymph vessels **(Figs 2.25.1 to 2.25.3)**
- *Retention cysts:* Occur due to the obstruction of the ducts of accessory lacrimal gland of Krause **(Figs 2.25.4 and 2.25.5)**
- *Implantation cysts:* Occur due to the implantation of conjunctival epithelial cells after an operation or trauma **(Figs 2.25.6 and 2.25.7)**

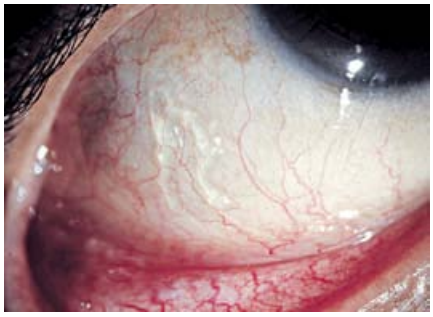


Fig. 2.25.1: Lymphangiectasia

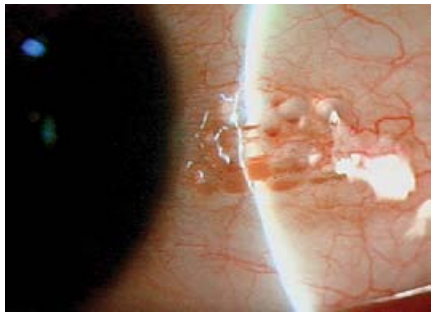


Fig. 2.25.2: Lymphangiectasia—multiple small cysts

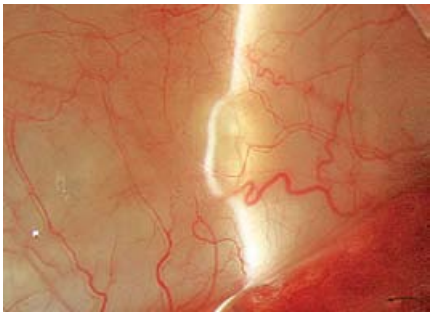


Fig. 2.25.3: Lymphatic cyst

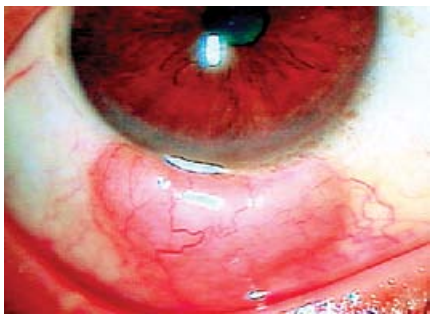


Fig. 2.25.4: Retention cyst

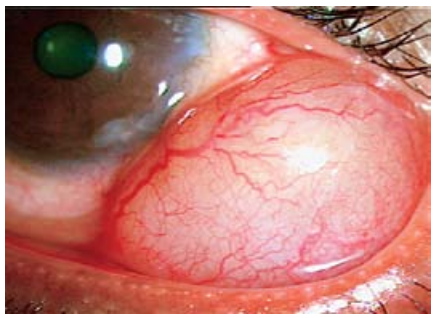


Fig. 2.25.5: Retention cyst—large

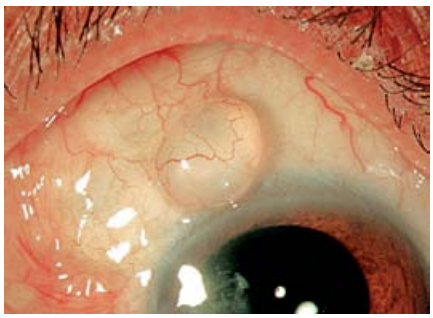


Fig. 2.25.6A: Implantation cyst—following cataract surgery

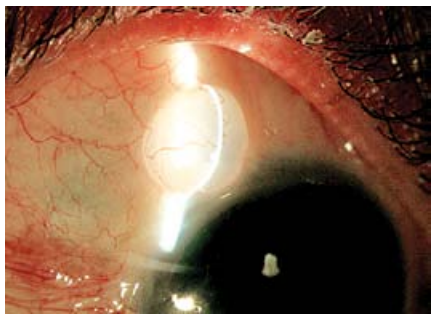


Fig. 2.25.6B: Implantation cyst—following cataract surgery

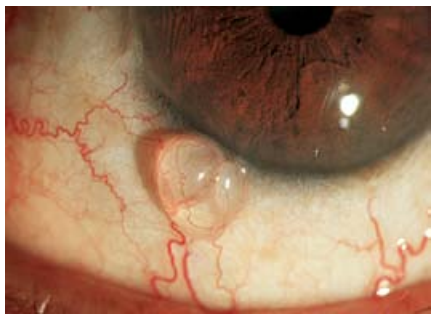


Fig. 2.25.7A: Implantation cyst—following trauma

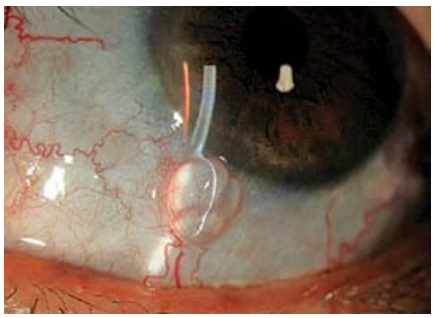


Fig. 2.25.7B: Conjunctival cyst—following trauma

- *Parasitic cysts:* Occur rarely due subconjunctival cysticercus or hydatid cyst **(Figs 2.25.8 and 2.25.9)**
- *Treatment:* Simple excision in most of the cases.

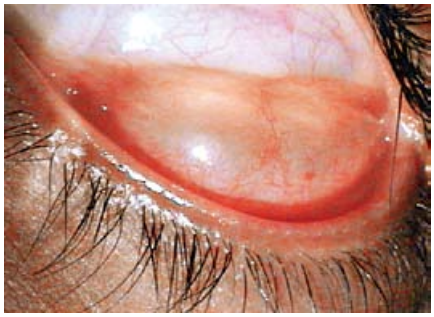


Fig. 2.25.8A: Parasitic cyst—cysticercosis

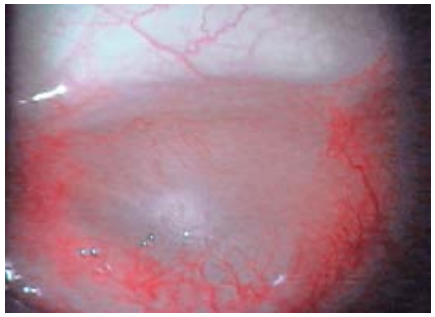


Fig. 2.25.8B: Subconjunctival cysticercus scolex

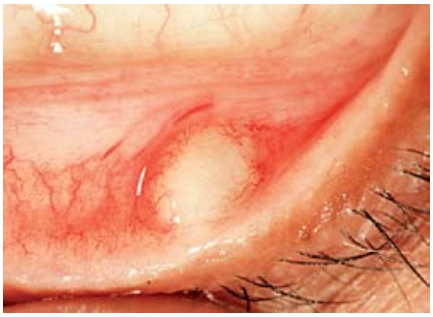


Fig. 2.25.9: Subconjunctival cysticercosis

Epibulbar Limbal Dermoid

- Congenital lesion, either isolated or with systemic association
- Solid, smooth, round white masses most frequently at the limbus, especially in inferotemporal quadrant (**Fig. 2.26.1**)
- May occur in any quadrant (**Figs 2.26.2A to C)** or may be away from the limbus (**Fig. 2.26.3**)
- Large lesion may also involve the cornea (**Fig. 2.26.4**)
- In some cases, they are associated with *Goldenhar's syndrome* (preauricular skin tags, vertebral anomalies and hemifacial hypoplasia) which may be unilateral or bilateral (**Figs 2.26.5 and 2.26.6**). There may be associated lid coloboma (**Figs 2.26.7A to C)** or microphthalmos (**Fig. 2.26.8**)
- Consist of skin with sebaceous glands and hair
- *Treatment:* Excision of the mass with or without lamellar keratoplasty.

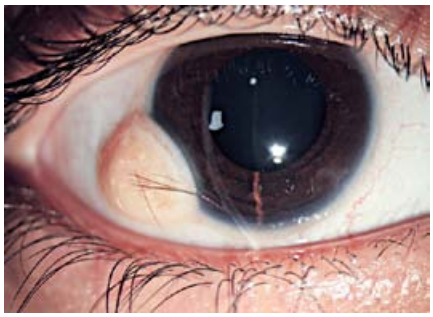


Fig. 2.26.1: Limbal dermoid—inferotemporal

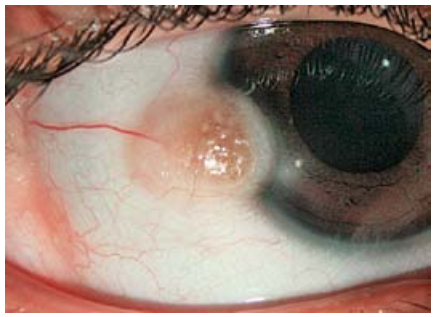


Fig. 2.26.2A: Limbal dermoid—temporal

Fig. 2.26.2B: Limbal dermoid—inferior

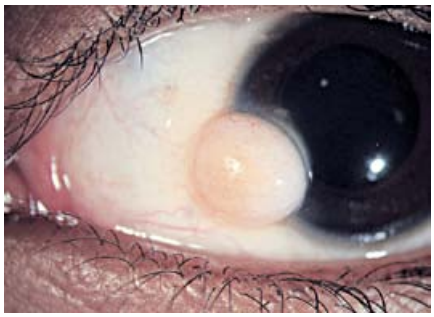


Fig. 2.26.2C: Limbal dermoid—nasal

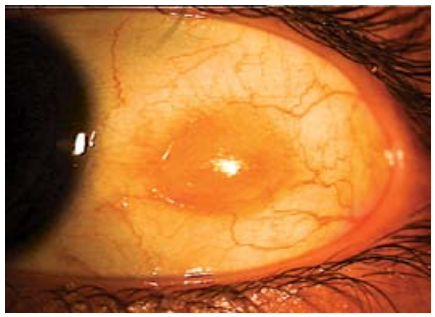


Fig. 2.26.3: Dermoid away from limbus

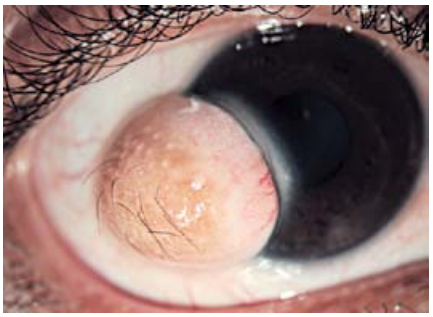


Fig. 2.26.4: Large limbal dermoid—corneal involvement

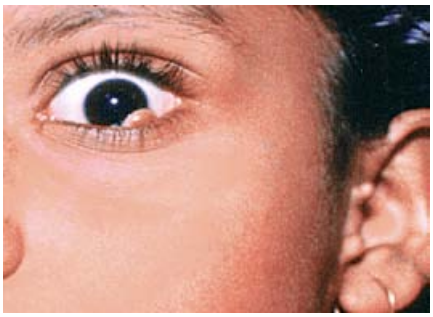


Fig. 2.26.5: Goldenhar's syndrome—unilateral

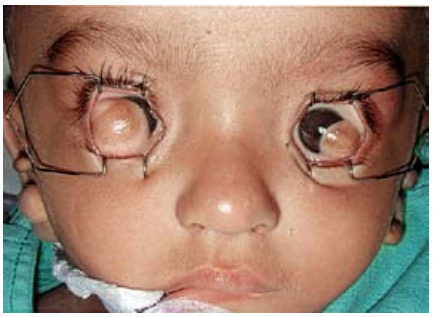


Fig. 2.26.6: Goldenhar's syndrome—bilateral

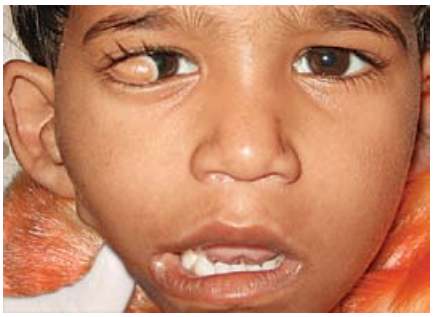


Fig. 2.26.7A: Goldenhar's syndrome—bilateral with facial asymmetry right side and coloboma upper lid left side

Fig. 2.26.7B: Goldenhar's syndrome—large epibulbar dermoid—RE

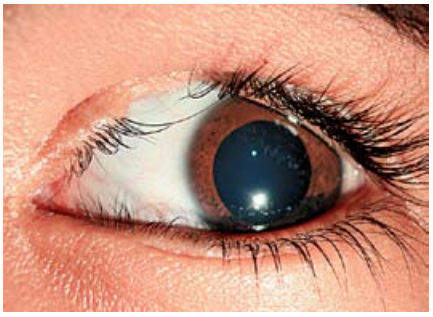


Fig. 2.26.7C: Bilateral Goldenhar's syndrome—coloboma left upper lid

Fig. 2.26.8: Goldenhar's syndrome—with lid coloboma and microphthalmos

Dermolipoma (Lipodermoid)

- Large, soft, yellow, movable subconjunctival masses, located at the outer canthus or at the limbus (**Figs 2.27.1 and 2.27.2**)
- The lesions extend beyond fornix and impossible to visualize the posterior limit (**Fig. 2.27.3**)
- Extra skin tag with hair may hang from the outer canthus (**Fig. 2.27.4**)
- Sometimes, it may be associated with Goldenhar's syndrome (**Fig. 2.27.5**)
- *Treatment:* Excision of the mass.

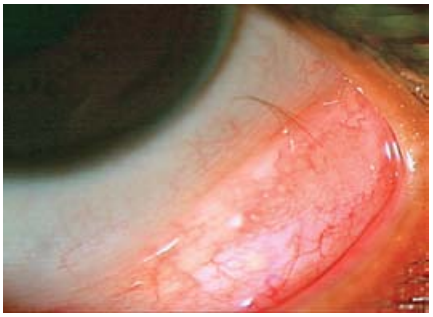


Fig. 2.27.1: Dermolipoma—lower fornix

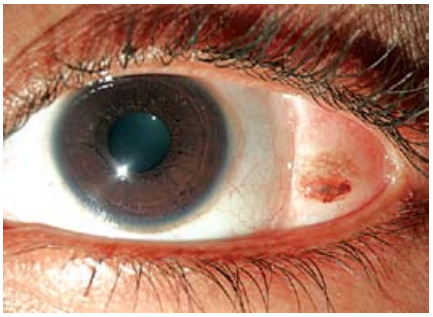


Fig. 2.27.2: Dermolipoma—outer canthus

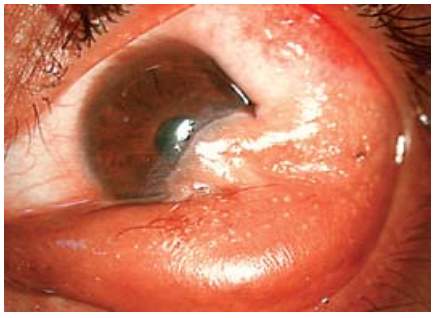


Fig. 2.27.3: Large dermolipoma

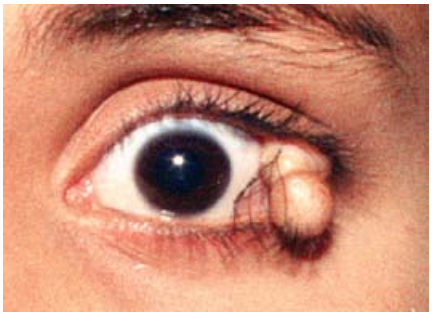


Fig. 2.27.4: Dermolipoma with skin tag

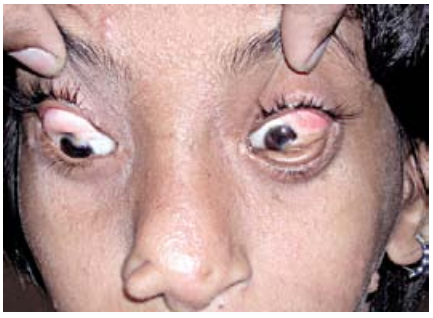


Fig. 2.27.5: Bilateral dermolipoma—Goldenhar's syndrome

Choristomas

- Similar to dermoid but has sharper edge and located just outside the limbus (**Fig. 2.28.1**)
- More fleshy and more vascularized (**Fig. 2.28.2**)
- It contains mature bone
- Very rarely other ectopic tissues may be seen, e.g. cartilage, lacrimal gland, fat, smooth muscles, etc.
- *Treatment:* Excision of the mass with or without lamellar keratoplasty.

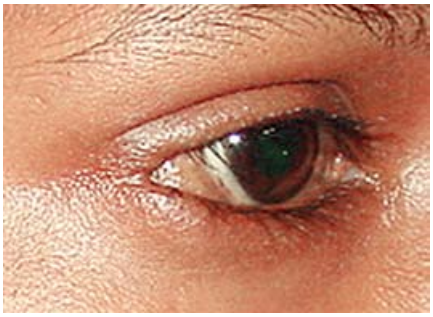


Fig. 2.28.1: Choristoma

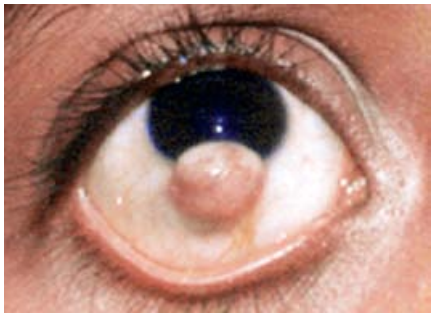


Fig. 2.28.2: Choristoma

Conjunctival Lipoma

- Similar to dermolipoma and located towards the fornix (**Fig. 2.29.1**)
- Yellowish in color and looks softer in consistency (**Fig. 2.29.2**). It contains only fatty tissue
- *Treatment:* Excision of the mass.

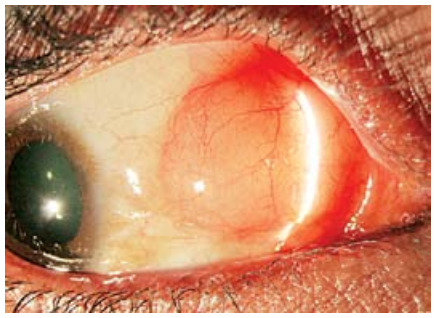


Fig. 2.29.1: Lipoma conjunctiva

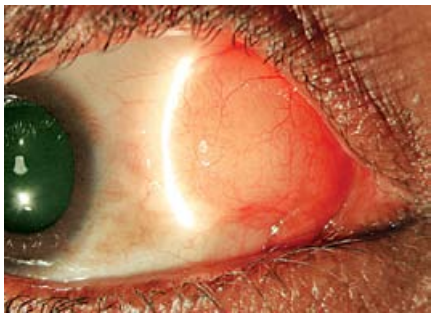


Fig. 2.29.2: Lipoma conjunctiva

Malignant Lesions

Conjunctival Intraepithelial Neoplasia (CIN)

- Rare unilateral premalignant condition *(carcinoma in situ)*
- Seen in elderly individuals with fair complexion
- Previously called—Bowen's disease, intraepithelial epithelioma or dyskeratosis
- Slightly elevated, fleshy mass with tuft of blood vessels at the limbus (**Fig. 2.30.1**)
- May involve the adjacent cornea (**Fig. 2.30.2**)
- It may be—*fleshy gelatinous* (**Fig. 2.30.3**), *leukoplakic type* (**Fig. 2.30.4**) *or papillary type* **(Fig. 2.30.5)**
- *Treatment:* Excision of the mass with double cryo-thawing of the dissected area and edge. It may recur with deeper involvement **(Fig. 2.30.6)**.

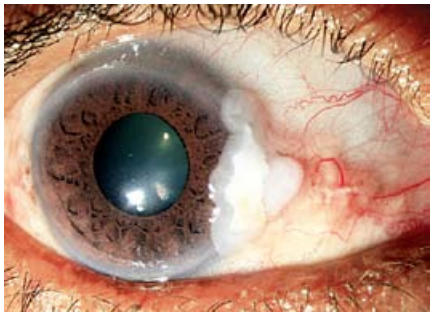


Fig. 2.30.1: Conjunctival intraepithelial neoplasia

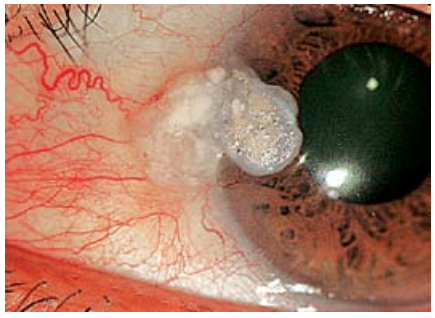


Fig. 2.30.2: Conjunctival intraepithelial neoplasia—corneal

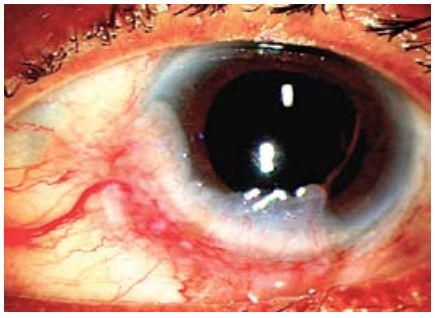


Fig. 2.30.3: CIN—gelatinous type

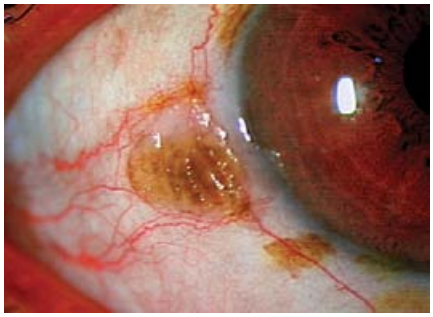


Fig. 2.30.4: CIN—leukoplakic type

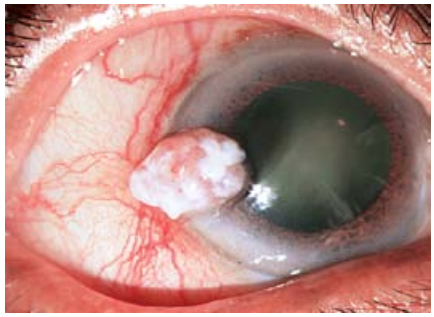


Fig. 2.30.5: CIN—papillary type

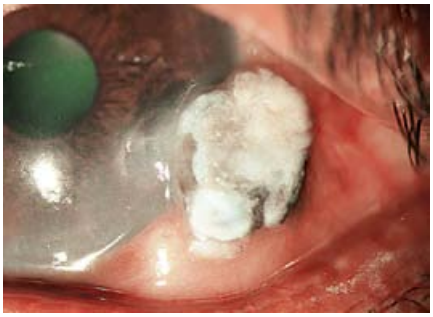


Fig. 2.30.6: Recurrent CIN with deeper involvement

Invasive Squamous Cell Carcinoma

- Slow growing, locally invasive tumor at the limbus (**Fig. 2.31.1**)
- It arises from papilloma or carcinoma *in situ* (**Fig. 2.31.2**)
- Reddish-gray fleshy mass with broad base, and characterized by deep invasion into the stroma with fixation to the underlying structure **(Fig. 2.31.3)**
- Frequently associated with large feeder vessels **(Fig. 2.31.4)**
- It may also involve the adjacent cornea **(Figs 2.31.5A and B)**
- Rarely it may arise from fornix **(Fig. 2.31.6)** or from caruncle **(Fig. 2.31.7)**
- It is usually sessile types. But papillary, ulcerative, nodular or pagetoid types are not uncommon **(Figs 2.31.8 to 2.31.11)**
- *Treatment:* Excision of the mass with double cryo-thawing of the dissected area with peroperative MMC. Recurrence at the site of lesion may occur in 5–10% of cases **(Figs 2.31.12A and B)**

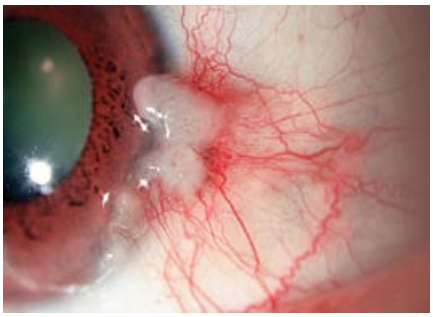


Fig. 2.31.1: Invasive squamous cell carcinoma

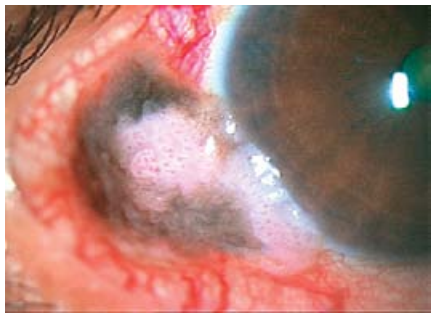


Fig. 2.31.2: CIN to squamous cell carcinoma

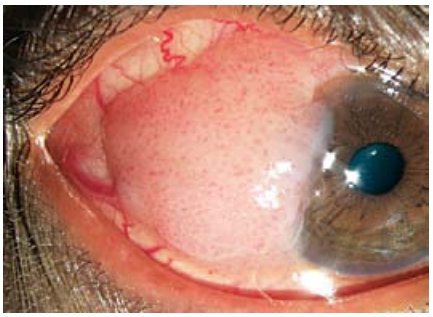


Fig. 2.31.3: Invasive squamous cell carcinoma

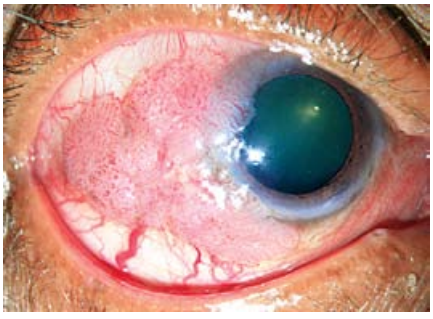


Fig. 2.31.4: Invasive squamous cell carcinoma—large feeder vessels

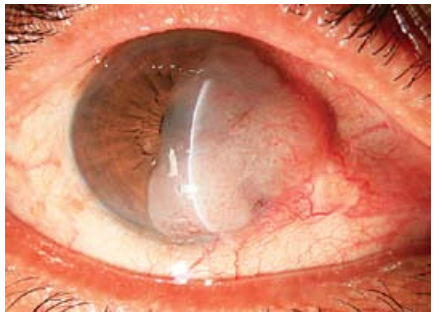


Fig. 2.31.5A: Invasive squamous cell carcinoma—corneal involvement

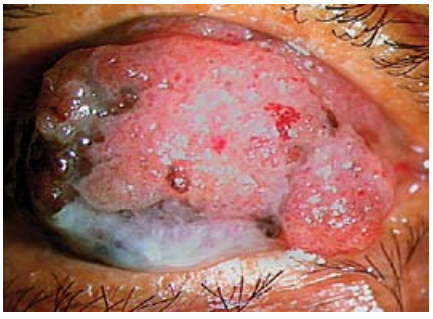


Fig. 2.31.5B: Severe corneal involvement

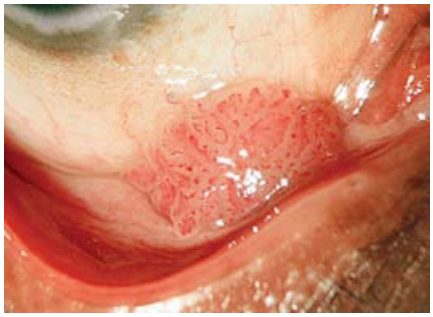


Fig. 2.31.6: Squamous cell carcinoma—arising from fornix

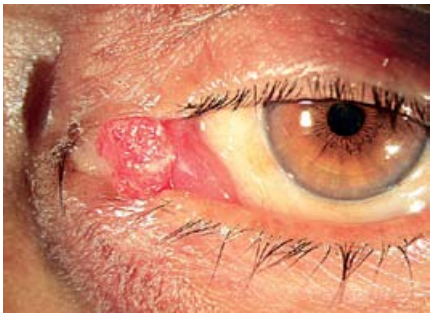


Fig. 2.31.7: Invasive squamous cell carcinoma—from caruncle

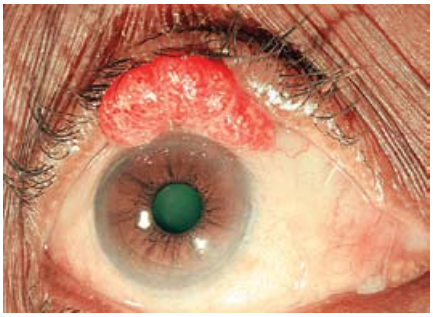


Fig. 2.31.8: Squamous cell carcinoma—papillary type

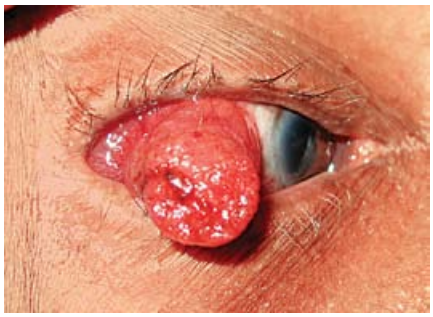


Fig. 2.31.9: Squamous cell carcinoma—ulcerative type

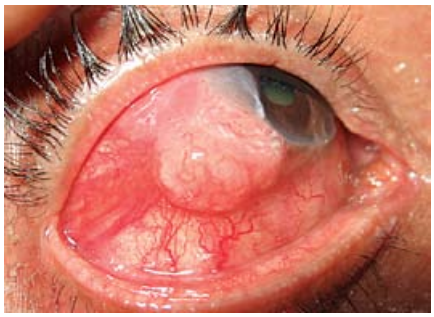


Fig. 2.31.10: Invasive squamous cell carcinoma—nodular type

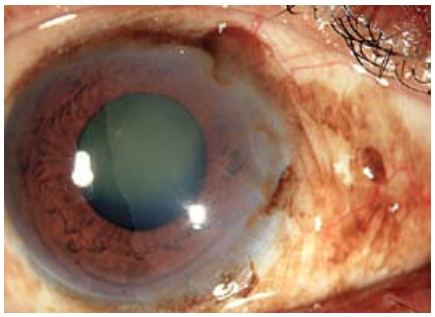


Fig. 2.31.11A: Invasive squamous cell carcinoma—pagetoid type

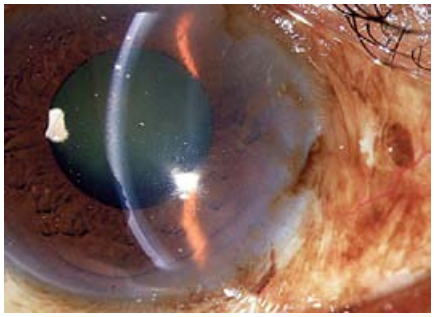


Fig. 2.31.11B: Invasive squamous cell carcinoma—pagetoid type

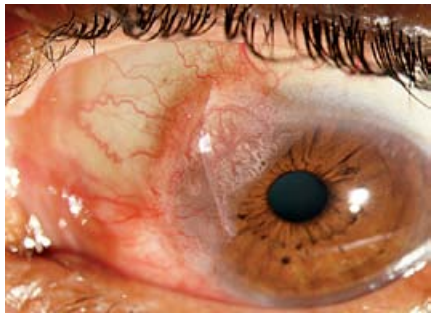


Fig. 2.31.12A: Recurrent squamous cell carcinoma

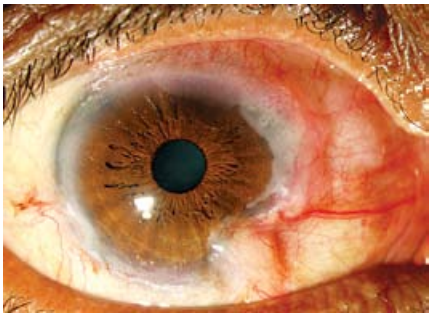


Fig. 2.31.12B: Recurrent squamous cell carcinoma

Pigmented Lesions

Flat Superficial Pigmentation

- These are common, congenital, small focal lesions
- The *causes are*: Conjunctival freckles, melanosis around Axenfeld loop (an intrascleral nerve loop 4 mm away from limbus) and melanosis around anterior ciliary artery **(Figs 2.32.1 and 2.32.2)**
- *No treatment is necessary.*

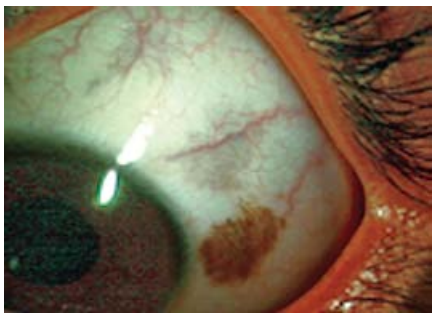


Fig. 2.32.1: Flat superficial pigmentation

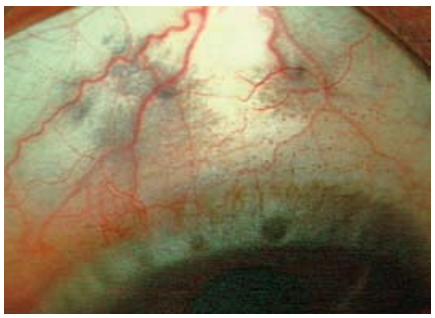


Fig. 2.32.2: Flat superficial pigmentation

Benign Epithelial Melanosis

- Bilateral, yellowish-brown or brownish-black patches most prominent at the limbus (**Fig. 2.33.1)**
- It is also present at the interpalpebral areas and gradually fades towards fornix
- Lesions can be easily moved on the globe
- *No treatment is necessary.*

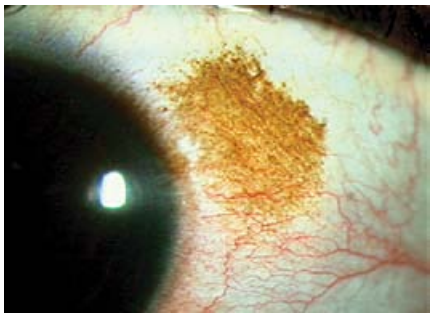


Fig. 2.33.1: Benign epithelial melanosis

Benign Subepithelial Melanocytosis

- Rare, unilateral congenital condition with a slate blue-gray discoloration (**Fig. 2.34.1)**
- It affects the episcleral and scleral tissue and cannot be moved
- It may affect the skin and mucous membrane in the distribution of fifth nerve
- There may be isolated *melanocytosis oculi*, or *Oculodermal melanocytosis* (nevus of Ota) which involves both the skin and globe (**Figs 2.34.2A and B)**
- *No treatment is necessary.*

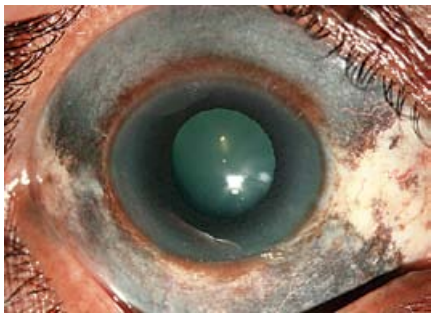


Fig. 2.34.1: Subepithelial melanocytosis—blue scleral appearance

Fig. 2.34.2A: Nevus of Ota

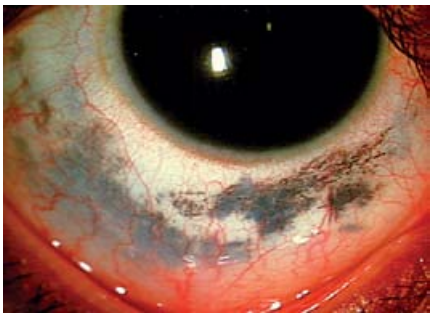


Fig. 2.34.2B: Subepithelial melanocytosis—nevus of Ota

Simple Nevus

- More common benign tumor
- Single, sharply demarcated, flat or slightly elevated lesion (**Fig. 2.35.1**)
- Has predilection for the limbus (**Fig. 2.35.2)** plica (**Fig. 2.35.3**), caruncle (**Fig. 2.35.4**) or lid margin

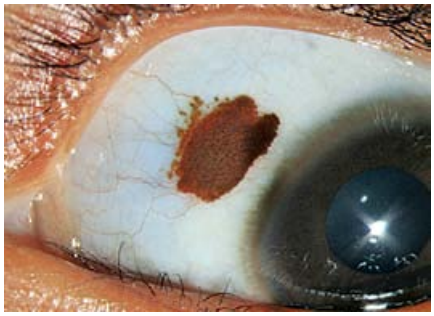


Fig. 2.35.1: Simple nevus

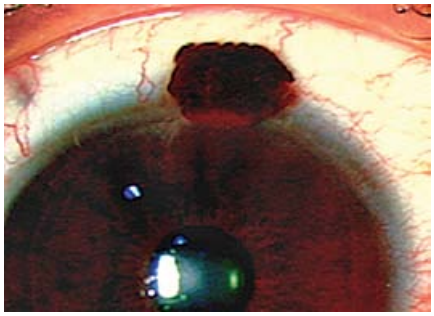


Fig. 2.35.2: Simple nevus—limbal

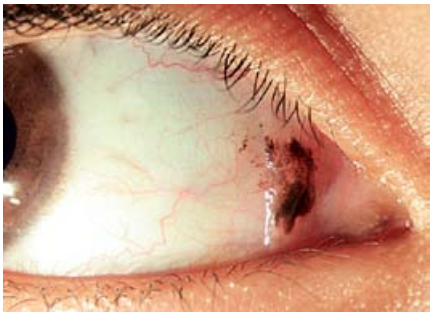


Fig. 2.35.3: Simple nevus—at plica

- Most nevi have a tan or brown color (**Fig. 2.35.5**), and 25% are less pigmented or nonpigmented (**Figs 2.35.6A and B**). Sometimes, a nevus may undergo cystic changes with appearance of microcysts within it **(Figs 2.35.7A and B)**
- Tends to enlarge or darken during puberty or pregnancy
- *Treatment:* Usually not necessary, but sometimes for cosmetic reason.

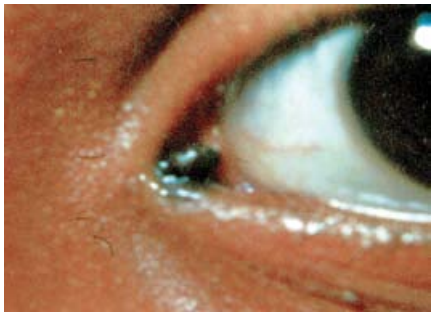


Fig. 2.35.4: Simple nevus—at caruncle

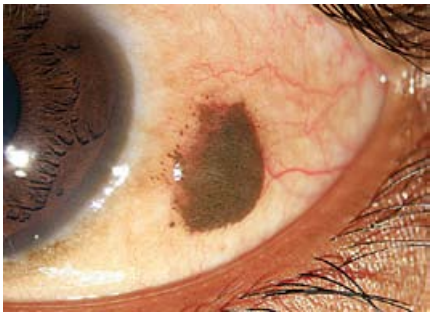


Fig. 2.35.5: Simple nevus—brown color

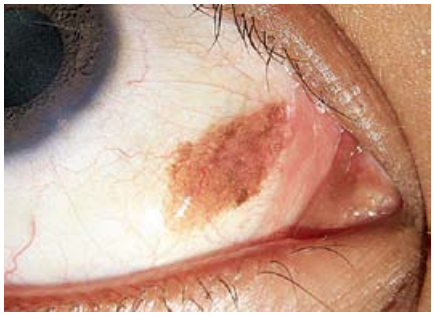


Fig. 2.35.6A: Simple nevus—less pigmented

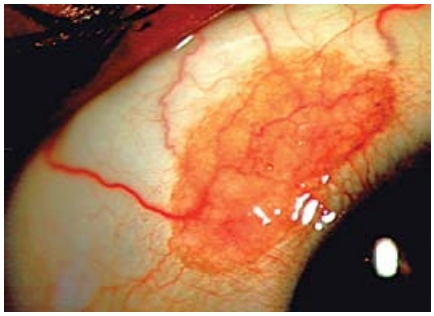


Fig. 2.35.6B: Simple nevus—nonpigmented

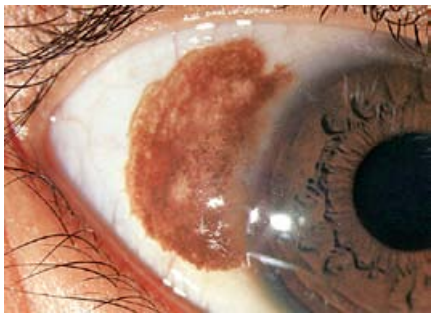


Fig. 2.35.7A: Conjunctival nevus with microcystic changes

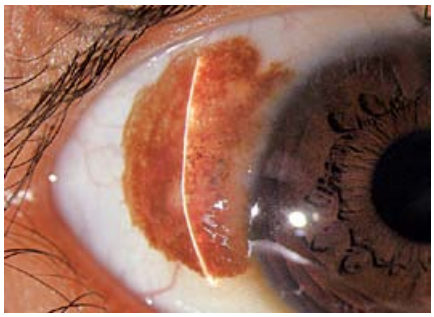


Fig. 2.35.7B: Conjunctival nevus with microcystic changes

Melanocytoma

- Very rare, congenital benign tumor
- Black, slowly growing mass which does not move freely over the globe **(Fig. 2.36.1)**
- Tends to enlarge during puberty
- *Treatment:* Not necessary.

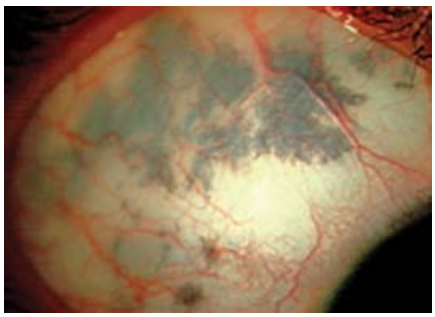


Fig. 2.36.1: Conjunctival melanocytoma

Precancerous Melanosis

- Small pigmented tumor which spread as a diffuse patch of pigmented lesion (**Figs 2.37.1A and B**)
- Mostly occur in elderly patients
- 20% cases it proceeds to frank malignancy (**Figs 2.37.2A and B**). Sudden color changes and the appearance of prominent feeder vessels are important (**Figs 2.37.3 and 2.37.4**)
- *Treatment:* Excision in suspected cases.

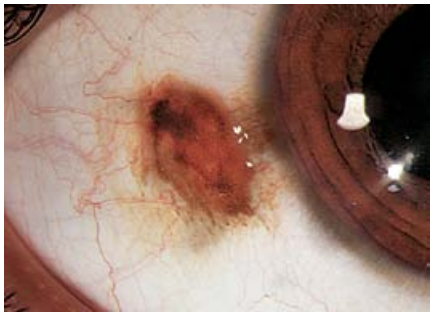


Fig. 2.37.1A: Precancerous melanosis

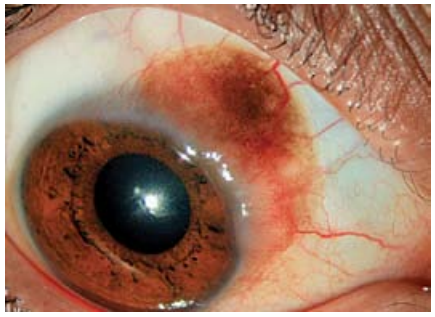


Fig. 2.37.1B: Precancerous melanosis

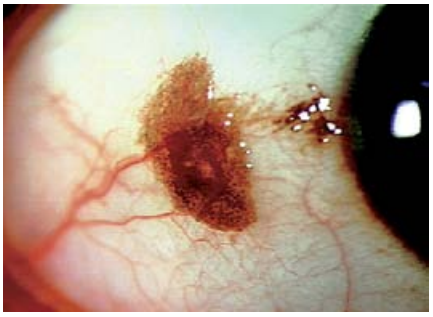


Fig. 2.37.2A: Precancerous melanosis—malignant change

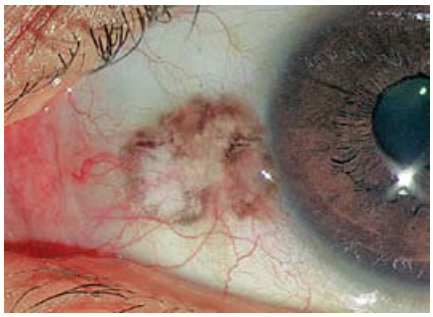


Fig. 2.37.2B: Precancerous melanosis—malignant changes

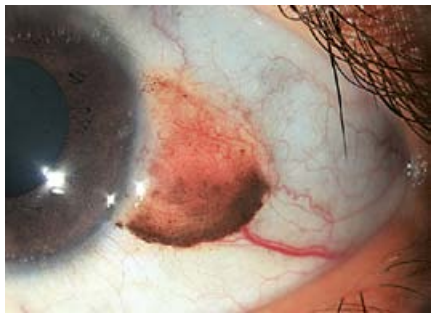


Fig. 2.37.3: Precancerous melanosis—color changes

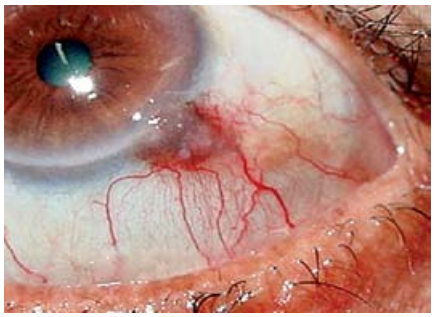


Fig. 2.37.4: Precancerous melanosis—prominent feeder vessels

Malignant Melanoma

- May be pigmented (**Fig. 2.38.1**) or nonpigmented lesion (**Figs 2.38.2A and B**) which affects elderly people
- Elevated lesion can occur in any part of conjunctiva **(Figs 2.38.3 and 2.38.4)** but has a predilection for limbus **(Fig. 2.38.5)**
- Larger lesion may involve cornea, adjacent part of eyelid and orbit **(Figs 2.38.6 and 2.38.7)**
- Metastasis is common
- *Treatment:* Excision of the mass.

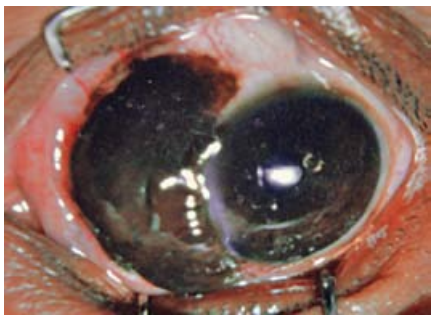


Fig. 2.38.1: Conjunctival melanoma

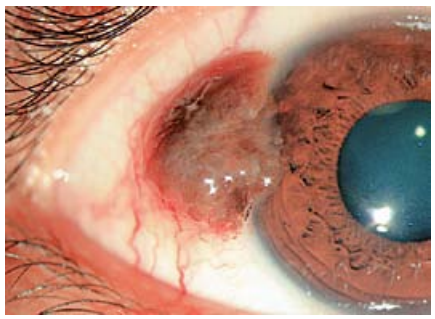


Fig. 2.38.2A: Amelanotic melanoma of the conjunctiva

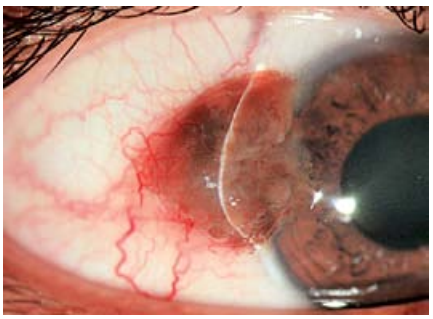


Fig. 2.38.2B: Amelanotic melanoma of the conjunctiva

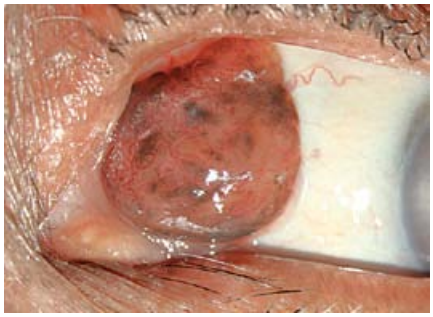


Fig. 2.38.3: Melanoma conjunctiva—near inner canthus

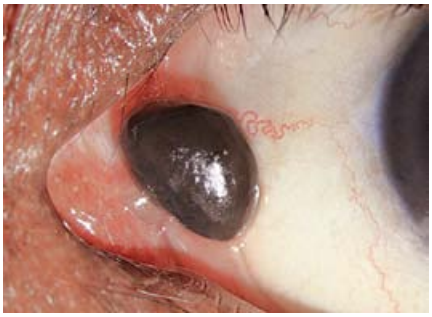


Fig. 2.38.4: Conjunctival melanoma—at the caruncle

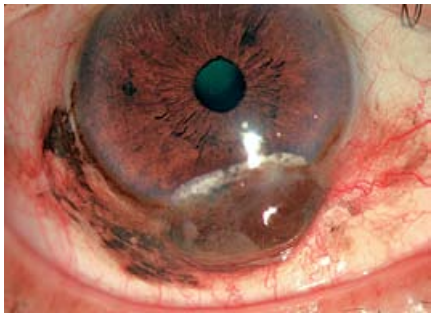


Fig. 2.38.5: Conjunctival melanoma—at limbus

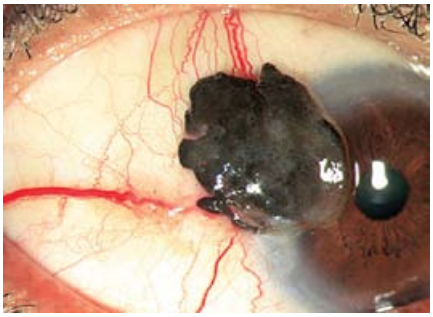


Fig. 2.38.6: Conjunctival melanoma—onto the cornea

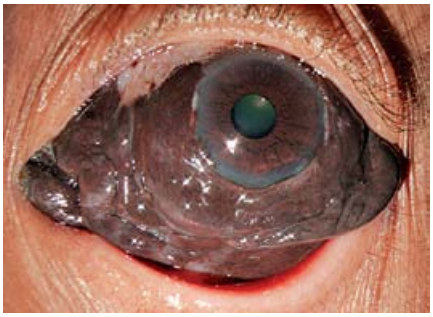


Fig. 2.38.7: Conjunctival melanoma—involving whole conjunctiva

Other Conjunctival Conditions

Subconjunctival Hemorrhage

- Rupture of a conjunctival blood vessel causes a bright red, sharply delineated area surrounded by the normal appearing conjunctiva
- Usually unilateral (**Fig. 2.39.1**), but it may be bilateral (**Fig. 2.39.2**) when precipitated by some straining factor like whooping cough. If associated with lid ecchymosis, called Panda bear (Raccoon eyes) sign (**Fig. 2.39.3**)
- May be associated with conjunctivitis (**Fig. 2.39.4**) or postsurgical trauma (**Fig. 2.39.5**)
- Traumatic hemorrhage may be associated with hyphema, chemosis or ecchymosis of lid (**Fig. 2.39.6**)
- When caused by head injury—the posterior limit is not visible (**Fig. 2.39.7**)
- *Treatment:* Only assurance and topical astringents.

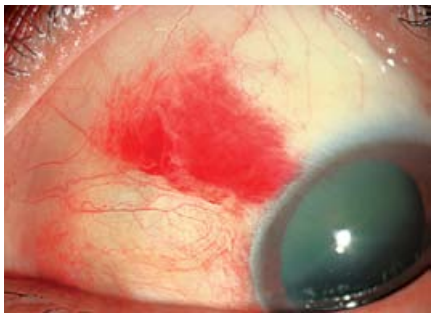


Fig. 2.39.1: Subconjunctival (SC) hemorrhage—small unilateral

Fig. 2.39.2: Bilateral subconjunctival hemorrhage—whooping cough

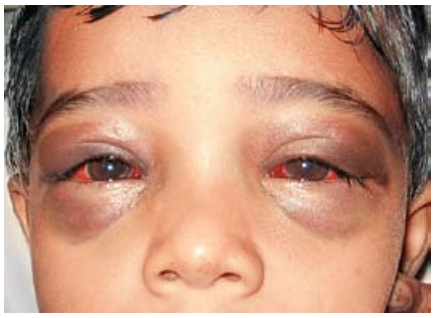


Fig. 2.39.3: Bilateral subconjunctival hemorrhage and ecchymosis

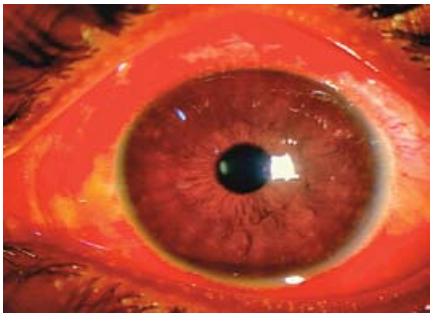


Fig. 2.39.4: SC hemorrhage—conjunctivitis

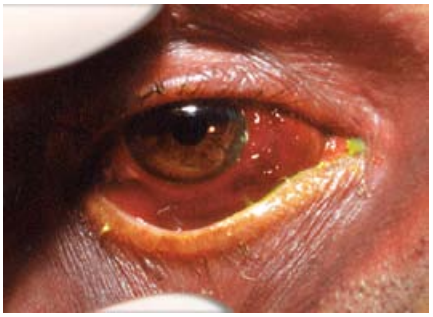


Fig. 2.39.5: SC hemorrhage—postsurgical

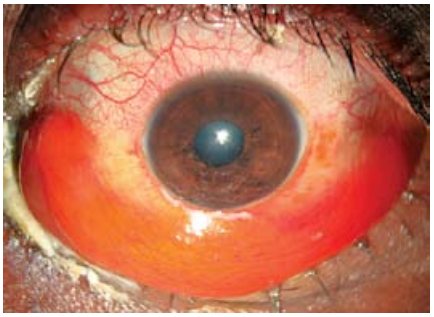


Fig. 2.39.6: SC hemorrhage—chemosis

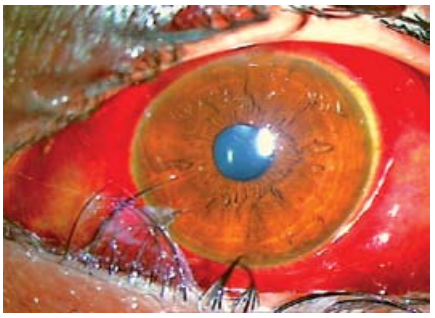


Fig. 2.39.7: SC hemorrhage—head injury

Conjunctival Xerosis

- Due to vitamin A deficiency with associated PEM (**Fig. 2.40.1)**
- Burns, pemphigoid, diphtheria, ß-blocker or following prolonged exposure, etc.
- Conjunctiva is pale, lustureless and with folds and variable pigmentation **(Figs 2.40.2 and 2.340.3)**
- *Treatment:* Active prompt vitamin A supplementation and tear substitutes.

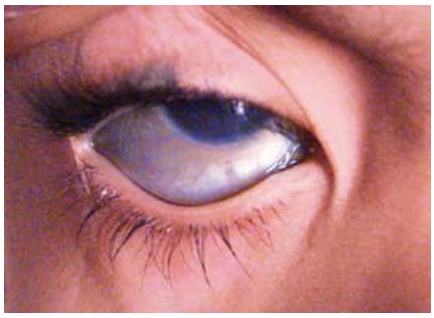


Fig. 2.40.1: Xerosis—vitamin A deficiency

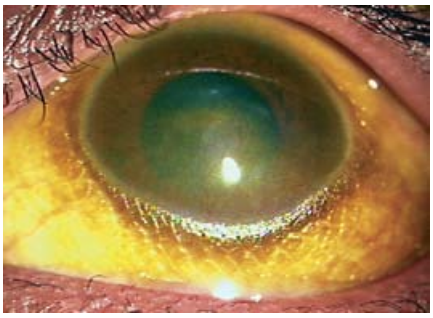


Fig. 2.40.2: Conjunctival xerosis

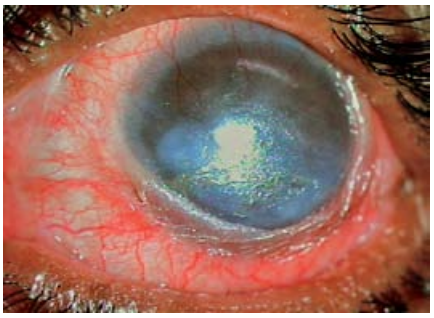


Fig. 2.40.3: Conjunctival xerosis

Bitot's Spot

- Bilateral lesion in young children, associated with vitamin A deficiency
- Appears as triangular patch, with base towards limbus and usually on the temporal side (**Fig. 2.41.1)**
- It may be cheesy or foamy in appearance (**Figs 2.41.2 and 2.41.3)**
- Associated with conjunctival xerosis and conjunctival folds
- Isolated Bitot's spot with wrinkling and pigmentary changes represents an area of old squamous metaplasia **(Figs 2.41.4A and B)**
- *Treatment:* Prompt vitamin A supplementation and correction of PEM.

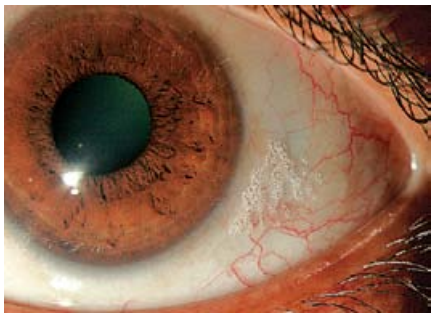


Fig. 2.41.1: Bitot's spot—fresh

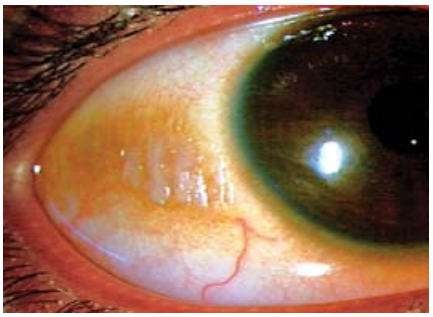


Fig. 2.41.2: Bitot's spot—cheesy appearance

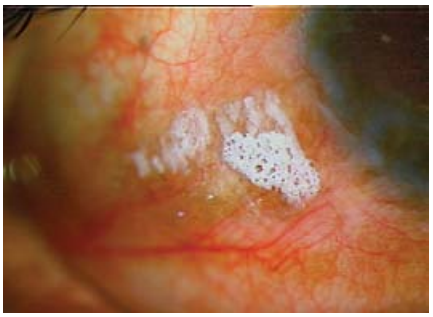


Fig. 2.41.3A: Bitot's spot—foamy appearance

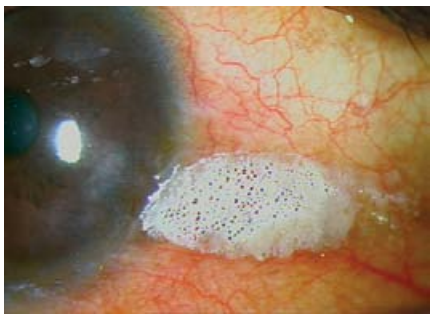


Fig. 2.41.3B: Bitot's spot—foamy appearance

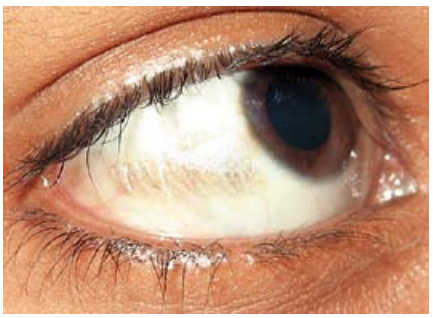


Fig. 2.41.4A: Old Bitot's spot with pigmentation—RE

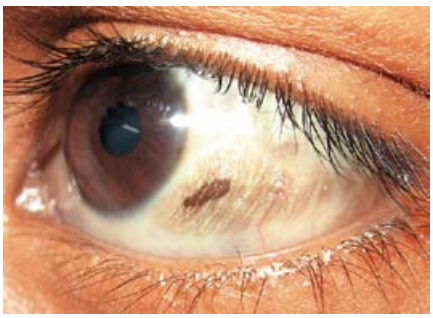


Fig. 2.41.4B: Old Bitot's spot with pigmentation—LE

Keratoconjunctivitis Sicca (KCS)

- Very common bilateral condition, leading to dry eye and ocular surface disorders
- Reduced or absent tear meniscus height (**Fig. 2.42.1**)
- Corneal filaments and mucus plaques
- Positive fluorescein **(Fig. 2.42.2)** or rose bengal staining of interpalpebral conjunctival area in a triangular fashion **(Fig. 2.42.3)**; more extensive stain in severe KCS with rose bengal **(Fig. 2.42.4)** or with lissamine green **(Fig. 2.42.5)**
- Corneal staining is also present associated with other corneal changes **(*See Chapter 3*)**
- *Treatment:* Tear substitutes, topical cyclosporine (0.05%) or 'soft steroids', oral omega 3 fatty acid, punctal occlusion, etc.

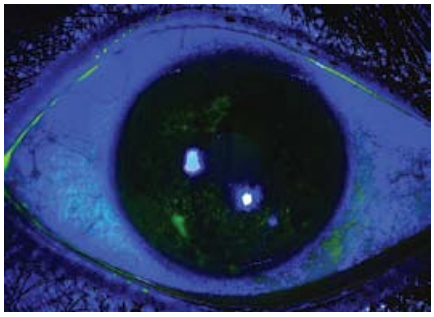


Fig. 2.42.1: Low tear meniscus height

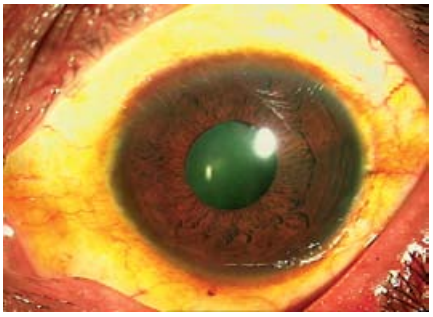


Fig. 2.42.2: KCS—fluorescein stain

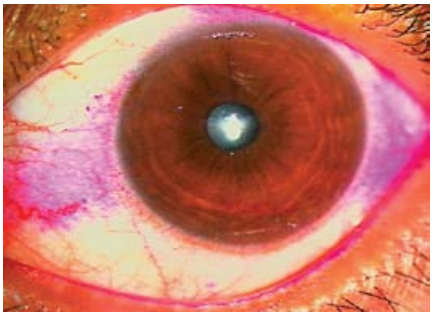


Fig. 2.42.3: KCS—rose bengal stain—classical

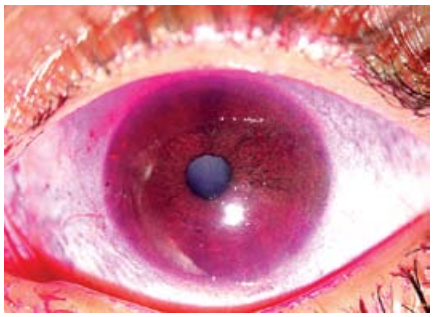


Fig. 2.42.4: KCS—rose bengal stain—more extensive

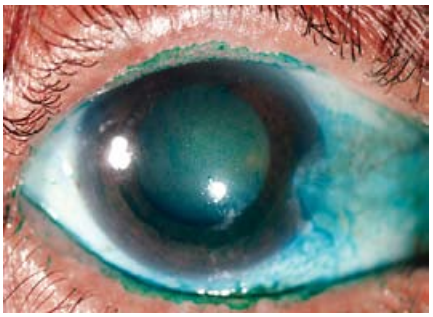


Fig. 2.42.5: KCS—lissamine green staining

Mucus Fishing Syndrome

- Rare unilateral or bilateral self trauma to the conjunctiva
- Excessive mucus and isolated conjunctival areas staining with rose bengal **(Fig. 2.43.1)**
- *Treatment:* Reassurance.

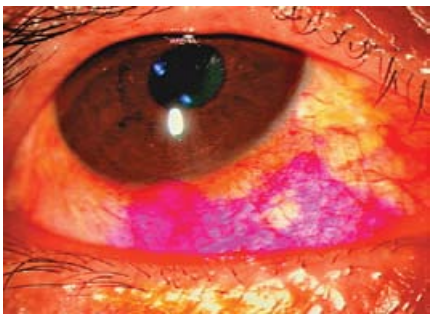


Fig. 2.43.1: Mucus fishing syndrome

Lid Imbrication Syndrome

- Rare condition caused by overriding of the lower lid by the upper lid **(Fig. 2.44.1)**
- Staining of upper tarsus with rose bengal.

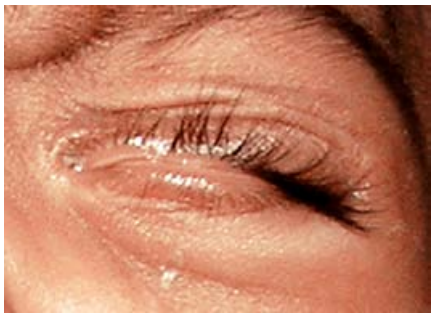


Fig. 2.44.1: Lid imbrication syndrome

Ocular Cicatricial Pemphigoid (OCP)

- Conjunctival involvement is seen in majority
- Associated oral mucosal lesions
- Conjunctival inflammation with fine subepithelial fibrosis
- Conjunctival shrinkage with shortening of inferior fornix **(Fig. 2.45.1)**
- Loss of plica semilunaris outline
- Medial symblepharon followed by total symblepharon formation **(Figs 2.45.2 and 2.45.3)** and eventually and total obliteration of fornices **(Fig. 2.45.4)**
- Trichiasis and metaplastic eyelashes, persistent epithelial defect and vascularization and keratinization of the cornea **(Figs 2.45.5 and 2.41.6)**
- Ultimately, total keratinization of the ocular surface **(Figs 2.45.7 and 2.41.8)**
- *Treatment:* Preservative free tears substitutes, removal of metaplastic eyelashes, systemic immunosuppressants, etc.

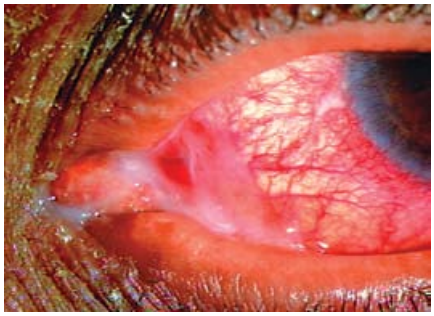


Fig. 2.45.1: Ocular cicatricial pemphigoid

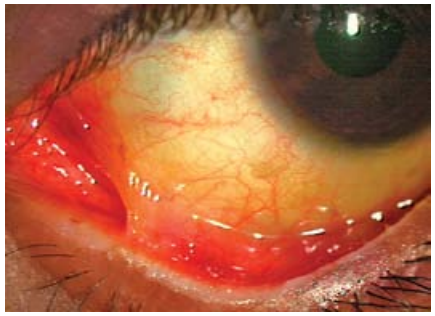


Fig. 2.45.2: OCP—medial symblepharon

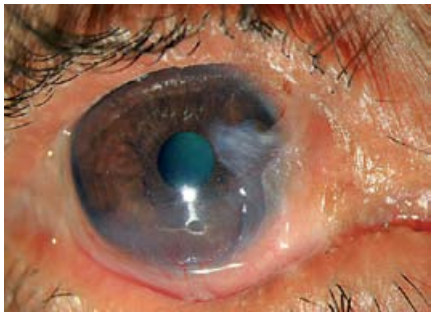


Fig. 2.45.3: OCP—increasing symblepharon

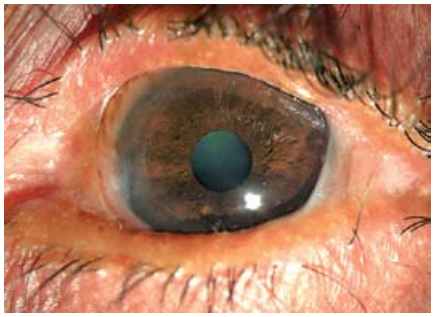


Fig. 2.45.4: OCP—obliteration of the fornices

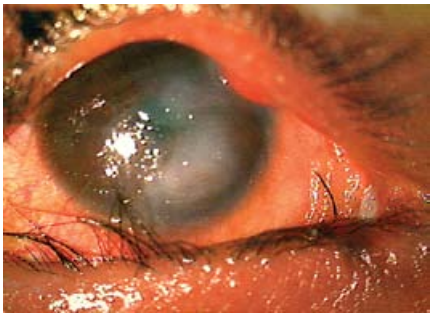


Fig. 2.45.5: OCP—trichiasis and epithelial defect

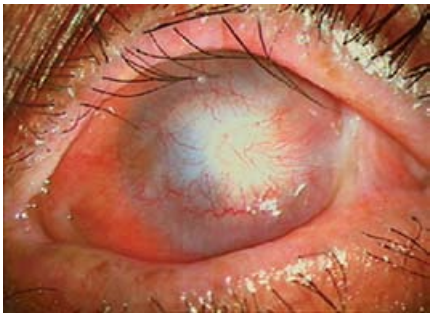


Fig. 2.45.6: OCP—vascularization

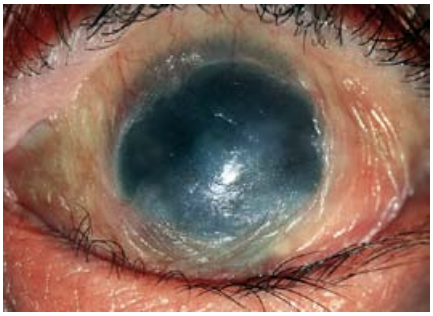


Fig. 2.45.7: OCP—subtotal keratinization of the ocular surface

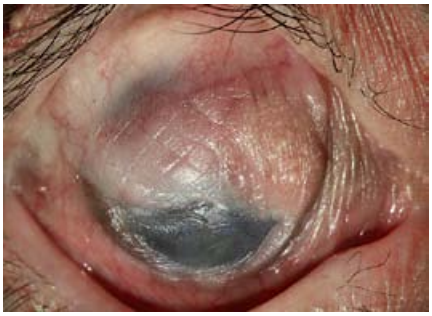


Fig. 2.45.8: OCP—total keratinization of the ocular surface

Stevens-Johnson Syndrome (SJS)

- Mucocutaneous vesicullobullous disease caused by a hypersensitivity reaction to certain drugs (**Fig. 2.46.1**). Toxic ectodermal necrolysis (TEN) is the most severe form (**Fig. 2.46.2)**
- Conjunctiva is involved in 50% of cases
- Mucopurulent conjunctivitis with membrane or pseudomembrane formation (**Fig. 2.46.3)**
- Oral mucous membrane lesions (**Figs 2.46.4 and 2.46.5)**
- Secondary scarring of the conjunctiva and lid margins—with trichiasis (acquired distichiasis), symblepharon (**Figs 2.46.6 and 2.46.7)** and obliteration of the fornices
- *Treatment:* Amniotic membrane transplantation in early stage, preservative free tears substitutes, periodic removal of metaplastic eyelashes, buccal mucous membrane graft of the lower tarsus, stepwise systemic immunosuppressants, etc.

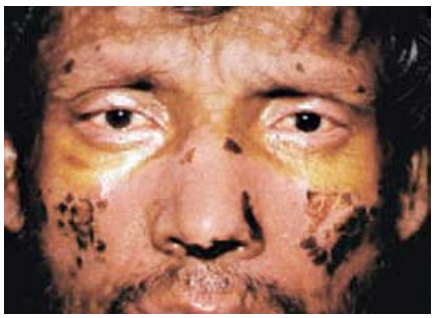


Fig. 2.46.1: Stevens-Johnson syndrome—skin involvement

Fig. 2.46.2: Toxic ectrodermal necrolysis (TEN)

Fig. 2.46.3: Stevens-Johnson syndrome—severe ocular lesion

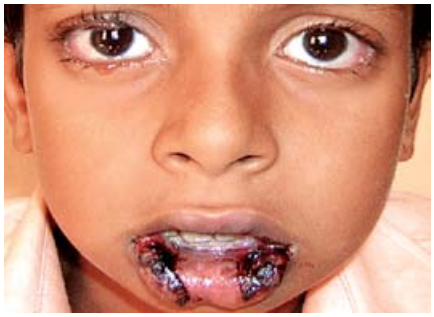


Fig. 2.46.4: SJ syndrome—oral and mild ocular lesion

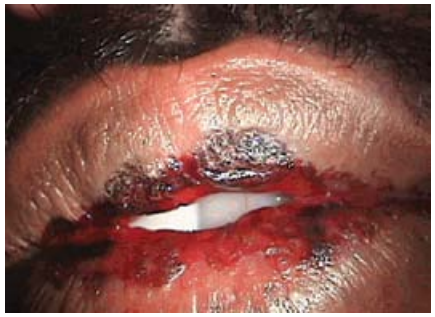


Fig. 2.46.5: SJ syndrome—oral lesion

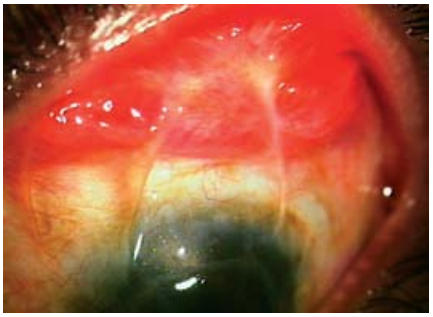


Fig. 2.46.6: SJ syndrome—symblepharon

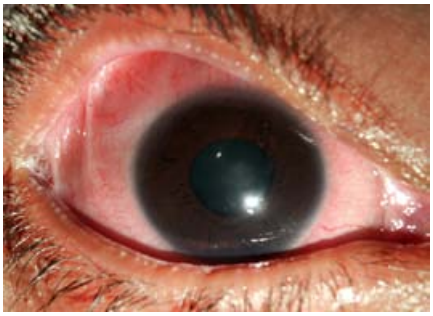


Fig. 2.46.7A: Symblepharon in Stevens-Johnson syndrome—RE

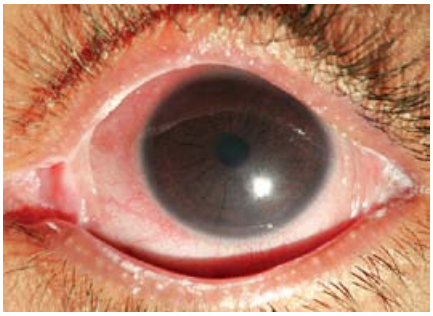


Fig. 2.46.7B: Symblepharon in Stevens-Johnson syndrome—LE

Superior Limbic Keratoconjunctivitis (SLK)

- Bilateral noninfectious keratoconjunctivitis, sometimes associated with thyroid dysfunction (**Fig. 2.47.1**)
- Papillary hypertrophy and thickening of superior tarsal conjunctiva (**Fig. 2.47.2**)
- Hyperemia and thickening of superior bulbar conjunctiva (**Figs 2.47.3A and B**)
- Positive rose bengal staining of upper part of the cornea
- Filaments in adjacent cornea
- *Treatment*: Tears substitutes, topical 0.05% cyclosporin A, excision of the upper bulbar conjunctiva.

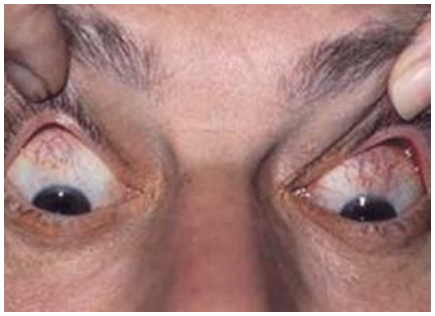


Fig. 2.47.1: Superior limbic keratoconjunctivitis

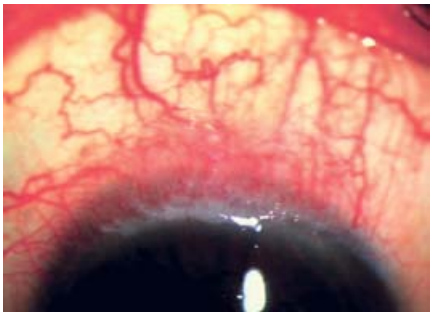


Fig. 2.47.2: SLK—thickening of superior bulbar conjunctiva

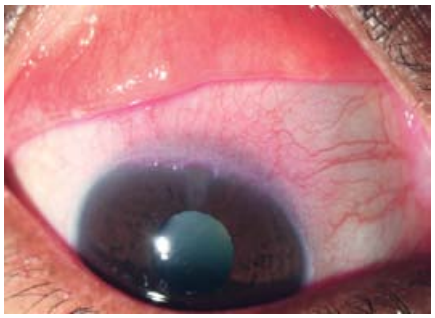


Fig. 2.47.3A: SLK—papillary hypertrophy and hyperemia of superior bulbar conjunctiva

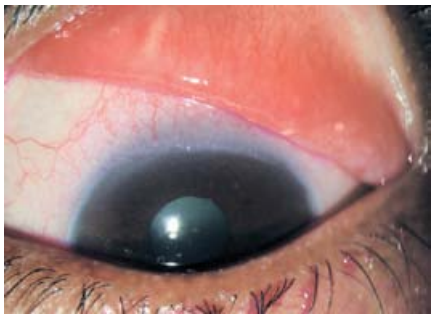


Fig. 2.47.4B: SLK—papillary hypertrophy and hyperemia of superior bulbar conjunctiva

Vascular Malformations

- *Capillary hemangioma*
  - Uncommon, and may be associated with hemangiomas of lid or orbit (**Figs 2.48.1 to 2.48.3)**
  - Bright red lesion of variable size, which blanches on pressure
  - May bleed following trivial trauma, or spontaneously
  - Responsible for bloody tears
- *Caput medusae*
  - Perilimbal dilated and tortuous blood vessels (**Fig. 2.48.4)**
  - Associated with cavernous sinus thrombosis or caroticocavernous fistula
- *Telangiectasias*
  - Dilated and tortuous blood vessels with loop formation on the bulbar conjunctiva (**Figs 2.48.5 to 2.48.7)**
  - *Causes:* Hematological disorder, metabolic disorders, Sturge-Weber's syndrome, etc.
  - May bleed spontaneously and responsible for bloody tears.

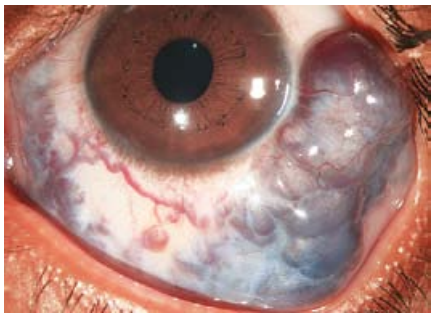


Fig. 2.48.1: Vascular malformation—hemangioma

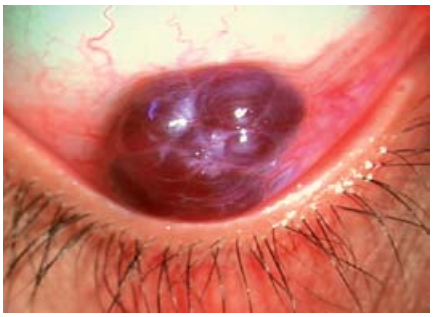


Fig. 2.48.2: Vascular malformation—conjunctival hemangioma

Fig. 2.48.3: Vascular malformation—conjunctival hemangioma

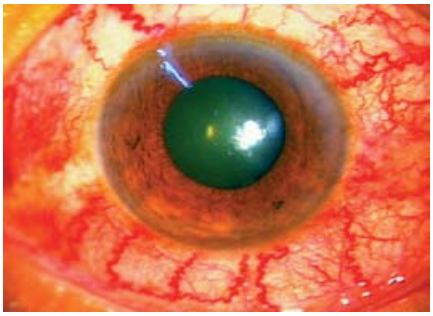


Fig. 2.48.4: Vascular malformation—caput madusae

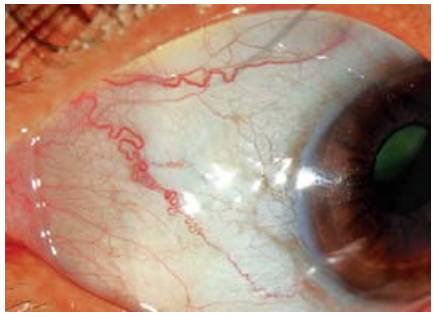


Fig. 2.48.5A: Vascular loop

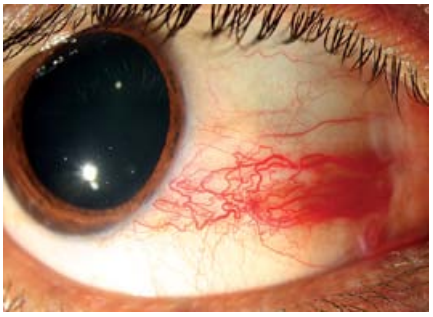


Fig. 2.48.5B: Vascular loops and prominence

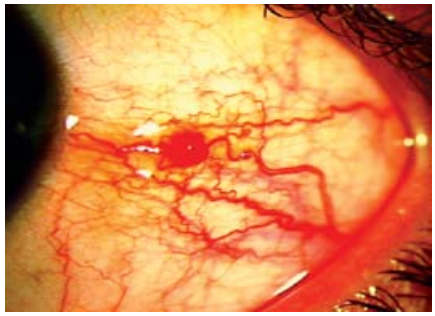


Fig. 2.48.6: Vascular malformation

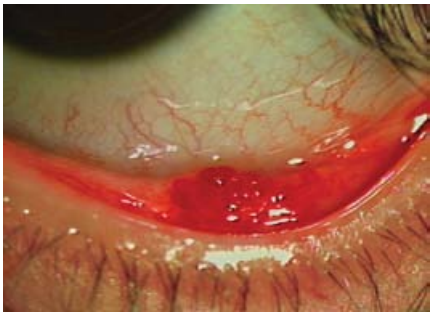


Fig. 2.48.7: Vascular malformation

Non-Hodgkin's Lymphoma

- Single or multiple lesion; bilateral in 20% cases
- Smooth fleshy subconjunctival infiltrate which may take the shape of a large mass
- Usually located in upper or lower fornices (**Figs 2.49.1 and 2.49.2)**
- To be differentiated histopathologically from *reactive benign lymphoid hyperplasia* (**Figs 2.49.3 and 2.49.4)** which is similar in appearance.

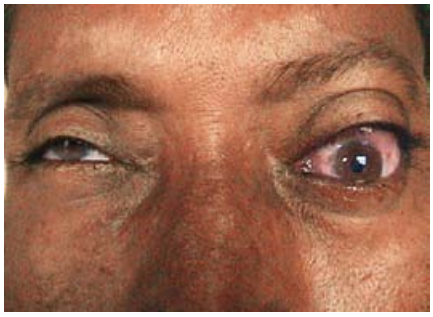


Fig. 2.49.1: Non-Hodgkin's lymphoma

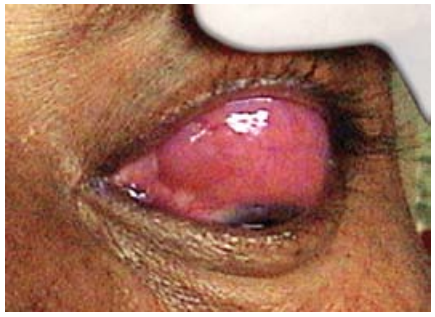


Fig. 2.49.2: Non-Hodgkin's lymphoma

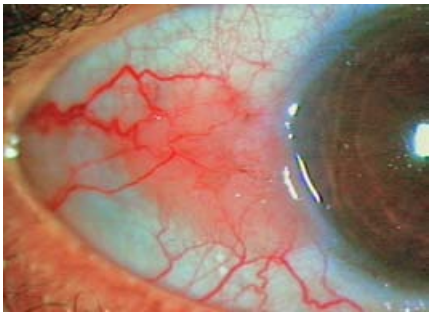


Fig. 2.49.3: Reactive benign lymphoid hyperplasia

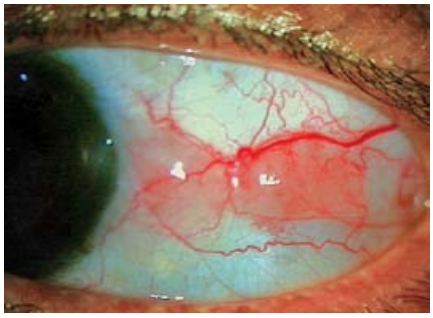


Fig. 2.49.4A: Reactive benign lymphoid hyperplasia

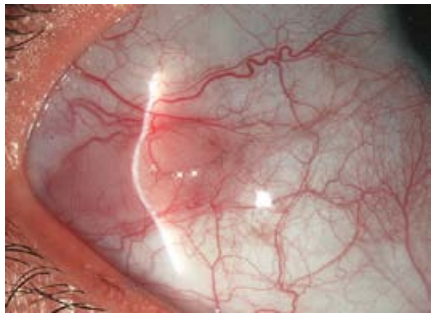


Fig. 2.49.4B: Reactive benign lymphoid hyperplasia

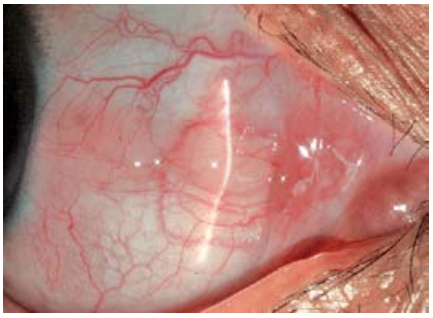


Fig. 2.49.4C: Reactive benign lymphoid hyperplasia

Amyloidosis of Conjunctiva

- Very rare condition, may be associated with systemic amyloidosis
- Yellowish-white lesion on the palpebral conjunctiva (**Figs 2.50.1 and 2.50.2)**
- Confirmation by excision and histochemical examination of the mass.

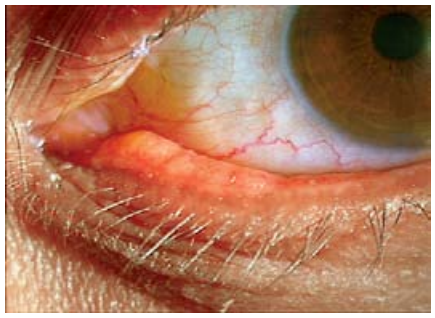


Fig. 2.50.1: Conjunctival amyloidosis

Fig. 2.50.2: Conjunctival amyloidosis

Conjunctivochalasis

- Loose conjunctiva in old age may appear as conjunctival folds mainly in the inferior fornix (**Fig. 2.51.1)**
- Fluorescein or rose bengal staining of the lower part of the cornea (**Figs 2.51.2A and B)**
- May be responsible for watering and recurrent conjunctivitis
- *Treatment:* Excision of the redundant conjunctiva and then suturing or fibrin glue.

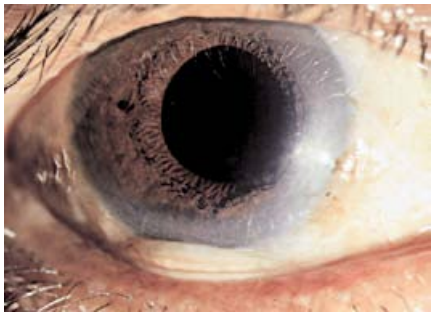


Fig. 2.51.1: Conjunctivochalasis

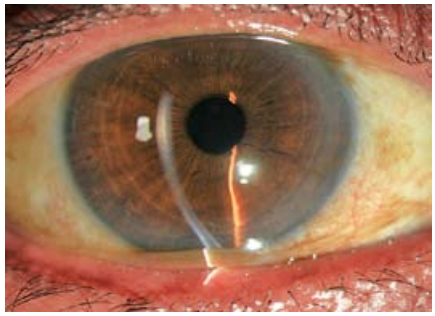


Fig. 2.51.2A: Conjunctivochalasis

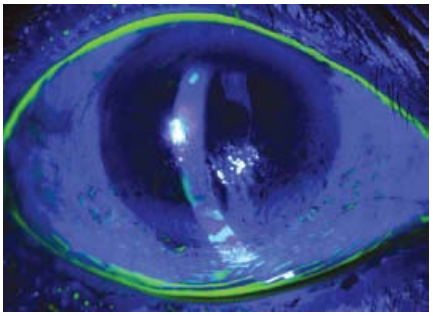


Fig. 2.51.2B: Conjunctivochalasis after fluorescein stain

Conjunctival Granulomas

- *Burst chalazion:* On tarsal conjunctiva, appears as a granulomatous mass (**Figs 2.52.1 and 2.52.2)**, a history of chalazion is important
- *Pyogenic granuloma*: Most commonly occurs after conjunctival surgery—like, pterygium (**Fig. 2.52.3)** or excision of symblepharon **(Fig. 2.52.4)** or after a trauma **(Fig. 2.52.5)**
- *Rhinosporidiosis*: Rare fungal infection usually after a foreign body in conjunctiva (**Figs 2.52.6 to 2.52.8)**.

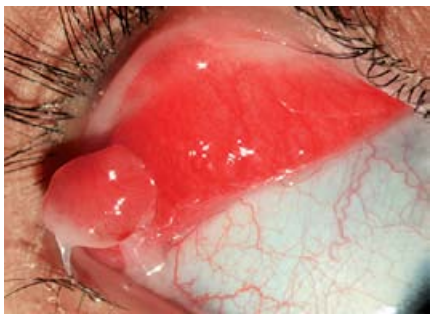


Fig. 2.52.1: Granuloma—brust chalazion—upper tarsus

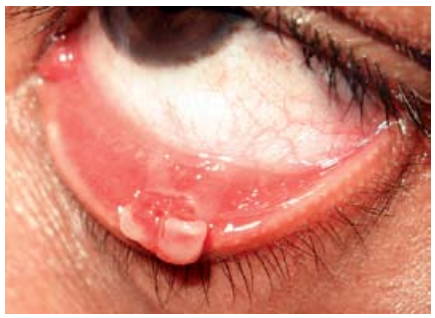


Fig. 2.52.2: Granuloma—brust chalazion—lower tarsus

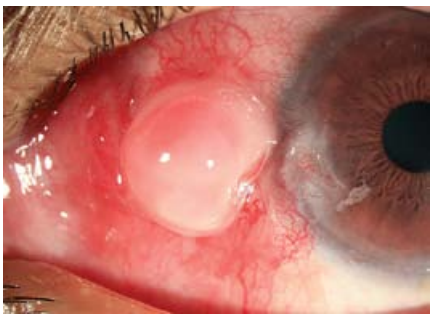


Fig. 2.52.3: Pyogenic granuloma—after pterygium operation

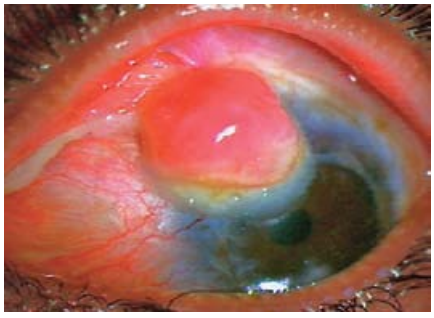


Fig. 2.52.4: Granuloma—after symblepharon operation

Fig. 2.52.5: Traumatic—pyogenic granuloma with canalicular tear—RE

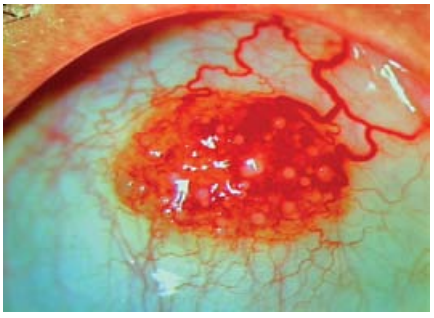


Fig. 2.52.6: Granuloma—rhinosporidiosis

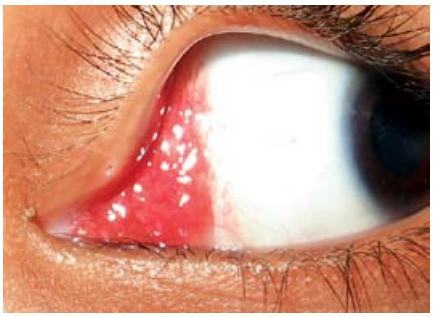


Fig. 2.52.7: Granuloma—rhinosporidiosis

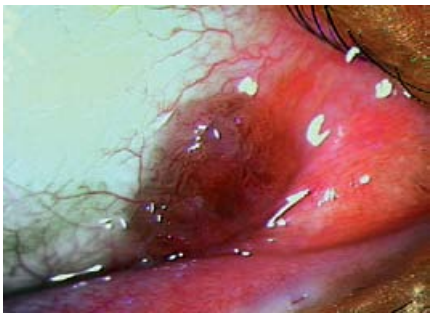


Fig. 2.52.8: Granuloma—rhinosporidiosis

Limbal Stem Cell Deficiency (LSCD)

- A disease caused by the loss or dysfunction of limbal stem cells. May be partial (**Fig. 2.53.1)** or total
- In LSCD, the corneal epithelium integrity and function cannot be maintained and consequently epithelial defects (**Fig. 2.53.2)** occur causing persistent pain and severe visual impairment
- By far the most common cause is from chemical burns. Other causes are: SJS, OCP, VKC, aniridia, etc. (**Figs 2.53.3 and 2.53.4)**
- *Treatment:* Limbal stem cell transplantation is the treatment of choice in LSCD:
  It may be autografts (CLAU) in unilateral cases; or from a living relative or cadaver (allografts in bilateral cases) (CLAL-lr or CLAL-c).
  In cases of allografts, systemic immunosuppression is required which is associated with enormous risk of toxicity whereas cultivated autologous limbal tissue transplantation eliminates the requirement of immunosuppression.

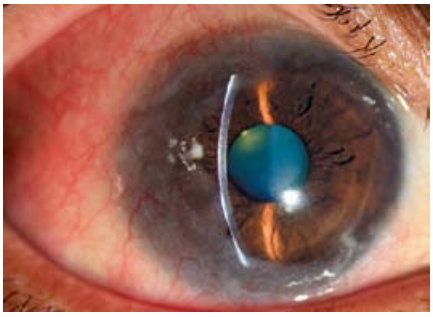


Fig. 2.53.1: Partial LSCD following chemical burn

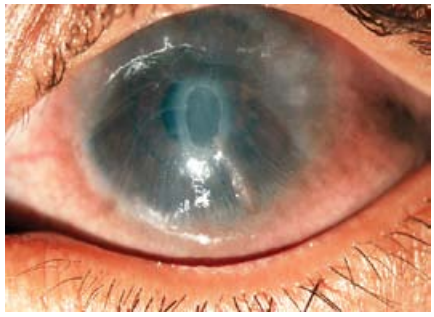


Fig. 2.53.2: Total LSCD—persistent epithelial defect

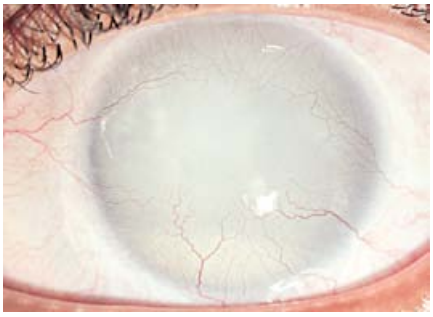


Fig. 2.53.3: Total limbal cell deficiency in aniridia

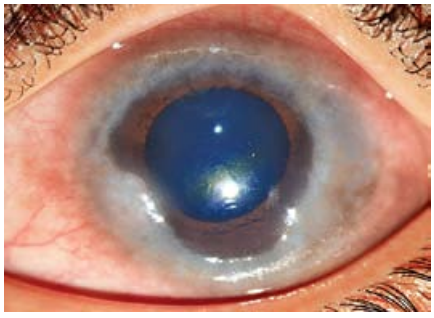


Fig. 2.53.4A: Total limbal stem cell deficiency in severe VKC

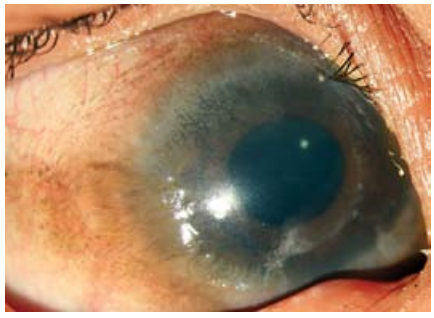


Fig. 2.53.4B: Total LSCD in severe VKC before treatment

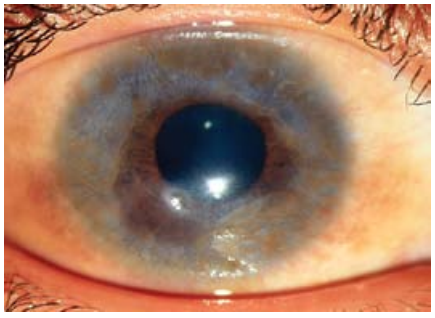


Fig. 2.53.4C: Total LSCD in severe VKC after treatment

Xeroderma Pigmentosa

- Rare, recessively inherited condition
- Photosensitive pigmented skin lesions **(Fig. 2.54.1)** which may turn into multiple cutaneous malignancies **(Fig. 2.54.2)**
- Recurrent conjunctivitis and scarring leading to conjunctival xerosis
- Corneal xerosis and clouding in the late stage **(Figs 2.54.3 and 2.54.4)**
- *Treatment:* Tears substitute and protection from ultraviolet light.

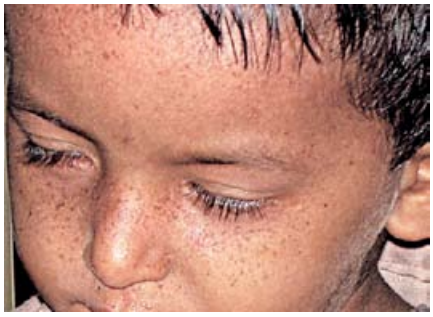


Fig. 2.54.1: Xeroderma pigmentosa—early with photophobia

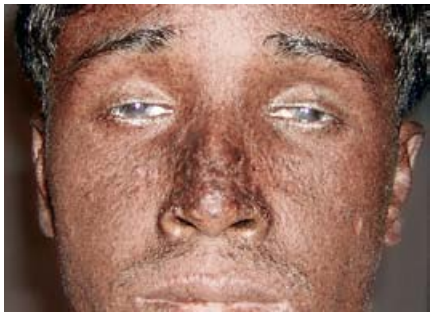


Fig. 2.54.2: Xeroderma pigmentosa—cutaneous malignancies

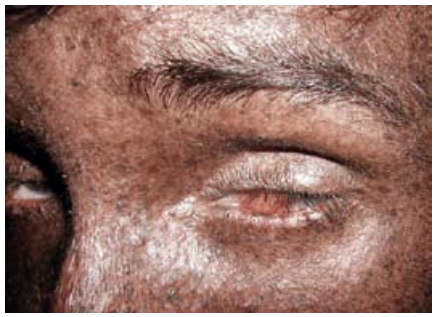


Fig. 2.54.3: Xeroderma pigmentosa—conjunctival xerosis

Fig. 2.54.4: Xeroderma pigmentosa—corneal xerosis

Metastatic Tumors of the Conjunctiva

- Metastatic tumors to the conjunctiva appear at an advanced stage of the systemic carcinoma when there are other ocular and organ metastases
- Leukemia and lymphoma can also occur in the conjunctiva
- Carcinoma breast is the most common malignancy to metastasize to the conjunctiva **(Figs 2.55.1A and B)**; may be also from bronchogenic carcinoma **(Fig. 2.55.2)**
- *Treatment:* Excisional biopsy to confirm, external beam radiotherapy or chemotherapy.

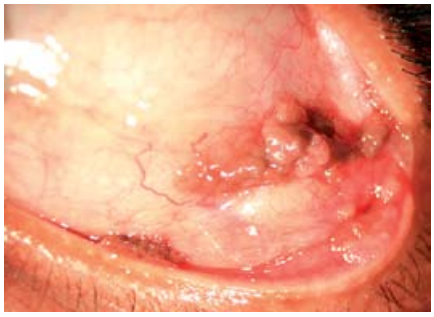


Fig. 2.55.1A: Metastatic carcinoma of conjunctiva—primary site breast

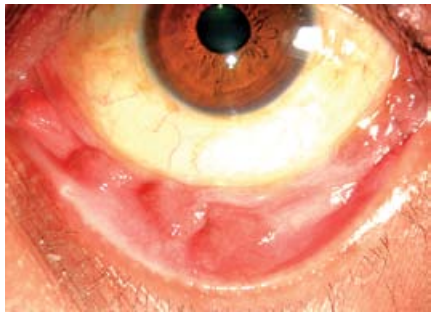


Fig. 2.55.1B: Metastatic carcinoma of conjunctiva—primary site breast

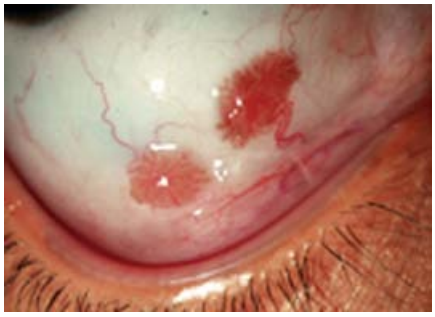


Fig. 2.55.2: Metastatic carcinoma of conjunctiva—primary site bronchus

CHAPTER

3 Diseases of the Cornea

CHAPTER OUTLINE

Cornea: Slit Lamp Examination

- Direct diffuse illumination
- Direct focal illumination
- Indirect illumination
- Retroillumination
- Specular reflection
- Sclerotic scatter
- Oscillatory illumination
- Special stains for epithelial lesions

Pigments Deposition in the Cornea

- Iron
- Copper
- Melanin
- Dye

Congenital Corneal Conditions

- Microphthalmos
- Nanophthalmos
- Megalocornea
- Buphthalmos
- Keratoglobus
- Posterior keratoconus
- Peter's anomaly
- Posterior embryotoxon
- Cornea plana
- Sclerocornea
- Keratectasia

Corneal Edema and Bullous Keratopathy

- Corneal edema
- Bullous keratopathy

Keratitis: Suppurative/Infectious

- Suppurative keratitis
- Bacterial keratitis/ulcer
- Fungal keratitis (corneal ulcer) or keratomycosis
- Acanthamoeba keratitis
- Perforated corneal ulcer
- Sequel of healded corneal ulcer

Viral Keratitis

- Herpes simplex virus (HSV) keratitis
- Primary HSV infection
- Recurrent HSV keratitis
- Dendritic HSV keratitis
- Geographical keratitis
- Stromal necrotic keratitis
- Metaherpetic keratitis (trophic ulcer)
- Disciform keratitis (HSV endotheliitis)
- Herpes zoster ophthalmicus (HZO)
- Acute ocular lesions
- Chronic ocular lesions

Other Infective Keratitis

- Atypical mycobacterial keratitis
- Nocardia keratitis
- Microsporidial keratitis
- Infectious crystalline keratitis

Keratitis (Other Types): Noninfectious and Peripheral

- Lagophthalmic (exposure) keratitis
- Neurotrophic keratitis (NTK)
- Nummular keratitis
- Marginal keratitis (catarrhal ulcer)
- Marginal ulcers with systemic collagen diseases or peripheral ulcerative keratitis (PUK)
- Phlyctenular keratitis
- Interstitial keratitis (IK)
- Punctate epithelial erosions (PEEs)
- Punctate epithelial keratitis (PEK)
- Superficial punctate keratitis (of Thygeson's)
- Superior limbic keratoconjunctivitis (SLK)
- Atheromatous ulcer
- Striate keratopathy (keratitis)
- Filamentary keratopathy (keratitis)
- Shield ulcer in vernal keratoconjunctivitis
- Keratitis medicamentosa
- Sclerosing keratitis

Corneal Degenerations

- Corneal degeneration: Major features
- Arcus senilis (gerontoxon)
- Terrien's marginal degeneration
- Pellucid marginal degeneration
- Mooren's ulcer
- Band-shaped keratopathy (BSK)
- Salzmann's nodular degeneration
- Spheroidal degeneration (climatic droplet keratopathy)
- Lipid degeneration (keratopathy)
- White limbal girdle of vogt

Corneal Dystrophies

Anterior Dystrophies (Epithelium and Bowman's Membrane)

- Meesmann's dystrophy
- Reis-Buckler's dystrophy

Stromal Dystrophies

- Granular dystrophy
- Macular dystrophy
- Lattice dystrophy
- Avellino (granular-lattice) dystrophy
- Central (Schnyder) crystalline dystrophy
- Central discoid dystrophy
- Congenital hereditary stromal dystrophy (CHSD)

Posterior Dystrophies

- Posterior polymorphous dystrophy
- Cornea guttata
- Fuchs' endothelial dystrophy
- Congenital hereditary endothelial dystrophy (CHED)
- Keratoconus

Miscellaneous Corneal Conditions

- Corneal abrasion
- Keratoconjunctivitis sicca (KCS)
- Vortex keratopathy
- Descemet's detachment
- Descemet's tear
- Descemet's folds and wrinkles
- Krukenberg's spindle
- Corneal dellen
- Crocodile shagreen
- Prominent corneal nerves
- Xerophthalmia (corneal signs in avitaminosis-A)
- Tunnel abscess
- Recurrent corneal erosion (RCE)
- Blood staining of the cornea
- Corneal tumors

Corneal Graft-Related Problems

Important Corneal Problems/Complications in PK

- Primary donor failure
- Persistent epithelial defects
- Loose sutures and broken sutures
- Early graft infection
- Recurrence of previous diseases in graft
- Induced high and irregular astigmatism
- Late PK—wound dehiscence
- Late graft infection
- Allograft rejection
- Endothelial rejection
- Late donor failure
- Donor dislocation in DSEK
- Double anterior chamber in DALK
- Late interface scarring in LK

- Epithelial ingrowth in DSEK
- Late secondary glaucoma

Refractive Surgery-Related Corneal Problems

- Late rupture of RK wound
- Post-PRK corneal haze
- Wrinkling of the flap
- Diffuse lamellar keratitis (DLK)
- Epithelial ingrowth
- LASIK flap displacement
- Post-LASIK ectasia
- Post-LASIK dry eye
- Corneal ulcer and/or interface infection
- Recurrence of original disease after PTK
- Late flap displacement or lost flap

Contact Lens-Related Corneal Problems

- Superficial punctate keratitis
- Contact lens—immune response keratitis
- Acute hypoxia of cornea
- Tight lens syndrome
- Corneal neovascularization
- Contact lens (toxic) keratopathy
- Infectious corneal infiltrates/ulcer
- Giant papillary conjunctivitis

Cornea: Slit Lamp Examination

Slit lamp biomicroscope offers a variety of illuminating and observing methods:

- Diffuse illumination
- Direct (focal) illumination
  - Broad beam (parallelopiped)
  - Narrow beam (optic section)
- Indirect-illumination
- Retroillumination
  - Direct
  - Indirect
- Specular reflection
- Sclerotic scatter
- Oscillatory illumination.

Direct Diffuse Illumination (Figs 3.1.1A to C)

- It is a good method of observing the eye and adnexa in general. Diffusers are generally ground glass plates that cover the light source. The slit should be opened wide and the magnification should be set as low as possible to enable a large field of view.

Direct Focal Illumination

- This is the most common method of viewing all tissues of the anterior eye, the focused slit is viewed directly by the observer through the microscope. The magnification can be increased quite markedly (10X to 40X or more) to view any areas of interest in greater detail
- Generally a very wide beam is used for surface study, whilst a very narrow one is used for sections.

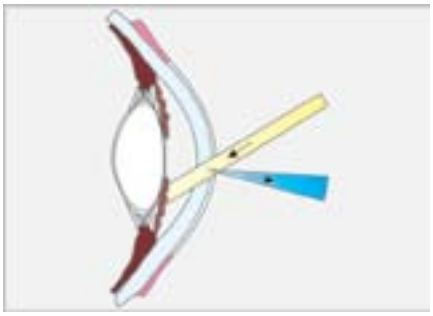


Fig. 3.1.1A: Direct diffuse illumination

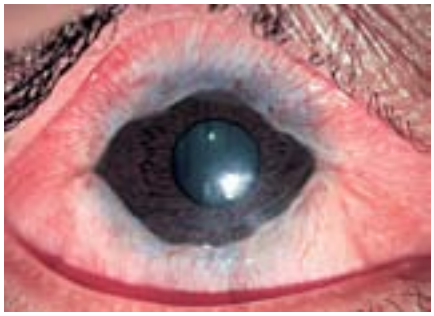


Fig. 3.1.1B: Direct diffuse illumination

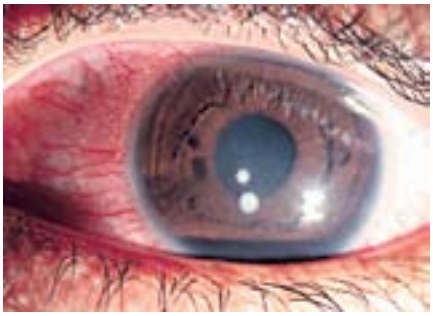


Fig. 3.1.1C: Direct diffuse illumination

Narrow Beam (Optical Section)
(Figs 3.1.2A and B)

- Once an abnormality has been found it is easier to determine the precise depth using an optical section. Generally the angle between the illuminating and observation systems should be set around 45 to 60 degrees. A good corneal section will allow at least 4 layers to be seen—tears

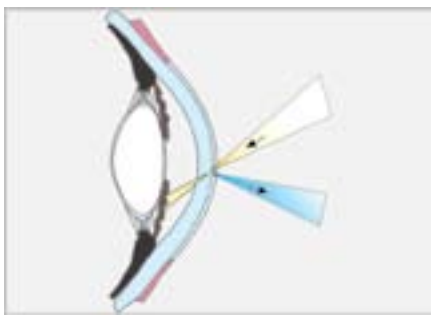


Fig. 3.1.2A: Direct focal (narrow beam)

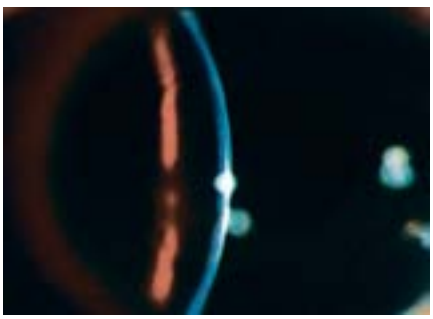


Fig. 3.1.2B: Direct focal (narrow beam)

(outer), epithelium (and Bowman's membrane), stroma seen as the central grey granular area and the fainter back line which is the endothelium (and Descemet's membrane).

Broad Beam (Parallelopiped)
(Figs 3.1.3A and B)

- A useful combination of the two is the parallelopiped section of the cornea, which uses a 2 mm slit width enabling corneal surface as well as stroma to be studied. This allows us to ascertain the depth of any interesting feature, e.g. foreign body, corneal abrasion. Direct illumination on the front surface of the crystalline lens reveals the 'orange peel' effect and on the iris allows observation of iris pattern.

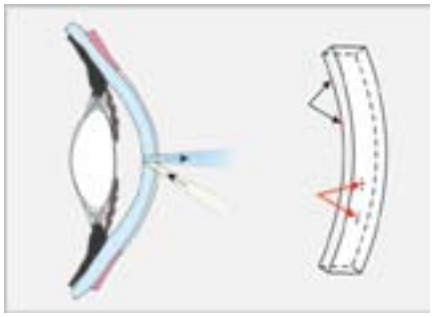


Fig. 3.1.3A: Direct focal (broad beam illumination)

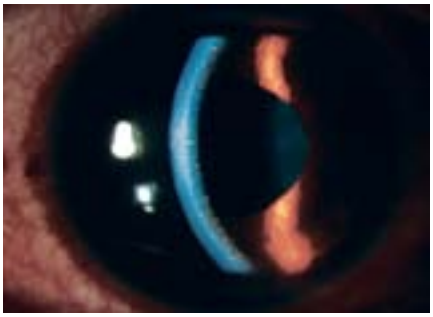


Fig. 3.1.3B: Direct focal (broad beam illumination)

Indirect Illumination (Figs 3.1.4A and B)

- Structures are often easier to see under indirect illumination as glare is reduced, e.g. opacities, corneal nerves and limbal vessels. When using the slit lamp direct and indirect illumination are viewed simultaneously, structures viewed in the illuminated field are seen under direct illumination, but as this does not fill the whole of the field of view, anything which reflects or scatters light from outside the illuminated area is being viewed by indirect
- To view certain features by indirect illumination, first locate it by direct illumination and keeping the viewing system unchanged swing the lamp to one side.

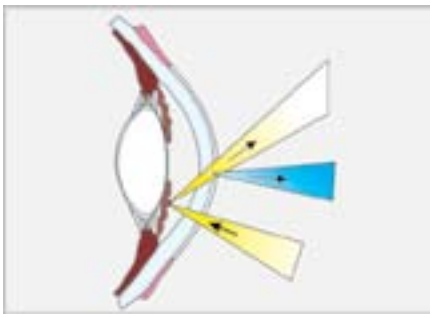


Fig. 3.1.4A: Indirect illumination

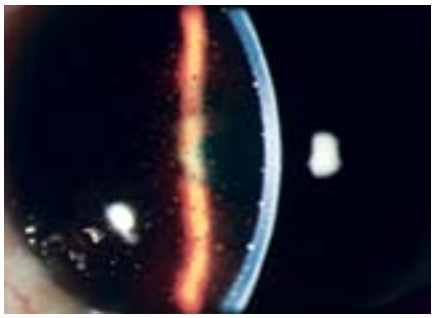


Fig. 3.1.4B: Indirect illumination

Retroillumination

- The light is reflected off the deeper structures, such as the iris or retina, while the microscope is focused to study the cornea in the reflected light. Features that are opaque to light appear dark against a light background (e.g. scars, pigment, and vessels containing blood).
- ***For direct retroillumination*** **(Figs 3.1.5A and B)*:*** The observed feature on the cornea is viewed in the direct pathway of reflected light. The angle between the microscope and the illuminating arm is about 60°.

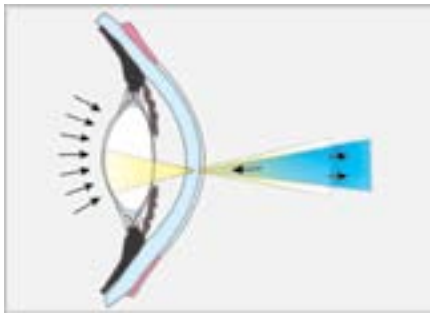


Fig. 3.1.5A: Direct retroillumination

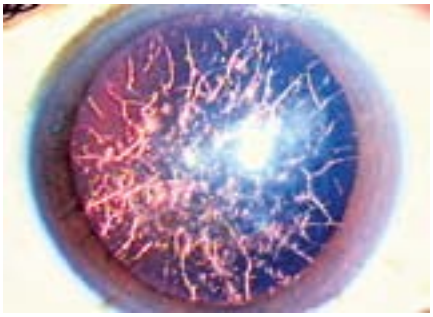


Fig. 3.1.5B: Direct retroillumination

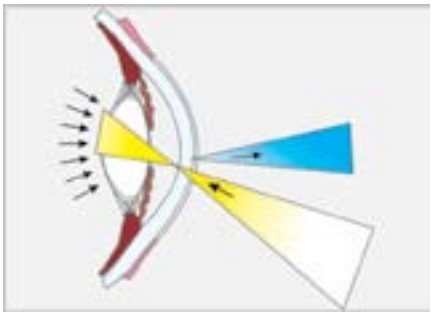


Fig. 3.1.6A: Indirect retroillumination

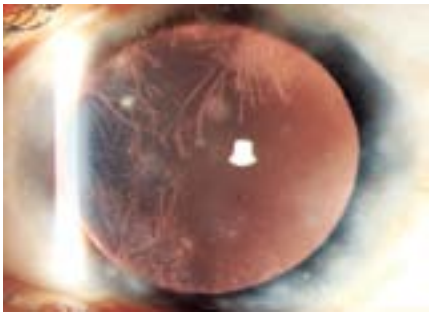


Fig. 3.1.6B: Indirect retroillumination

- ***For indirect retroillumination*** **(Figs 3.1.6A and B):** The angle between the microscope and slit lamp arms is greatly reduced or increased so that the feature on the cornea is viewed against a dark background.

Specular Reflection (Figs 3.1.7A and B)

- This type of viewing is achieved by positioning the beam of light and microscope such that the angle of incidence is equal to the angle of reflection. The light can be reflected from either the anterior (i.e. tears and epithelium) or posterior (i.e. endothelium) corneal surface
- Note that the reflected light should pass through only one eyepiece, and therefore this method is monocular.

Method for Viewing the Posterior Surface

- The angle between the light and microscope arms should be about 50°–60°
- A 2 mm wide parallelopiped and magnification of 20–25X is used
- Find the image of the illuminating bulb, then move the light beam until the image of the bulb is just behind the posterior surface of the parallelopiped (Incidence = reflection when the dazzle from the precorneal fluid is seen)
- Focus on the back of the parallelopiped. A mosaic of hexagonal endothelial cells will appear. The posterior endothelium and keratic precipitates may thus be studied
- This method of illumination is particularly useful to examine the endothelium layer of the cornea (e.g. blebs, polymegathism), although very high magnification is necessary, at least 40X is required and to see individual cells at 80X **(Fig. 3.1.7C)**.

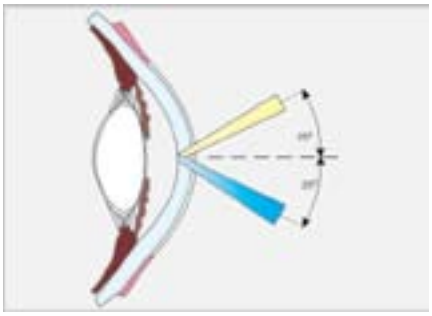


Fig. 3.1.7A: Specular reflection

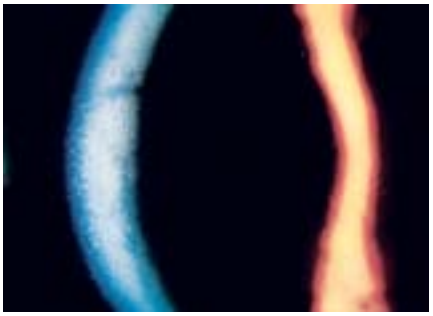


Fig. 3.1.7B: Specular reflection

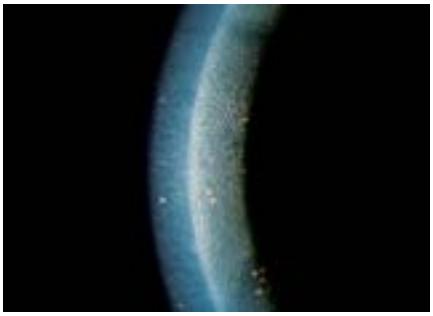


Fig. 3.1.7C: Specular reflection

Sclerotic Scatter (Figs 3.1.8A and B)

- This method uses the principle of total internal reflection. A narrow vertical slit (1–1.5 mm in width) is directed in line with the temporal (or nasal) limbus. A halo of light will be observed around the limbus as light is internally reflected within the cornea, but scattered by the sclera. Any corneal opacities, edema or foreign bodies will be made visible by the scattering light, appearing as bright patches against the dark background of the iris and pupil. It is important that the room illumination is as dark as possible.

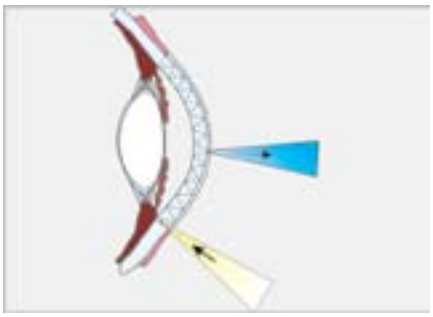


Fig. 3.1.8A: Sclerotic scatter

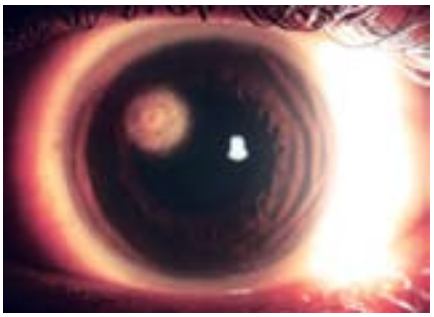


Fig. 3.1.8B: Sclerotic scatter

Oscillatory Illumination

- A beam of light is rocked back and forth by moving the illuminating arm or rotating the prism or mirror. Occasional aqueous floaters and glass foreign body in the anterior chamber are easier to observe.

Special Stains for Epithelial Lesions

- ***Fluorescein staining*** **(Fig. 3.1.9)**: To stain various corneal pathologies in the epithelium level, e.g. *corneal* abrasion, erosions, filaments, epithelial defects, dendrite in HSV keratitis, SPKs, for Seidel's test, tear film break up time (TBUT), etc.
- ***Rose bengal staining*** **(Fig. 3.1.10):** It is useful devitalized tissue, e.g. dendrite in HSV keratitis, PEE in dry eye and other lesions, conjunctival stain in dry eyes. It causes mild to moderate irritations of the eyes
- ***Lissamine green staining*** **(Fig. 3.1.11):** It is same as rose bengal, except it does not cause much irritation.

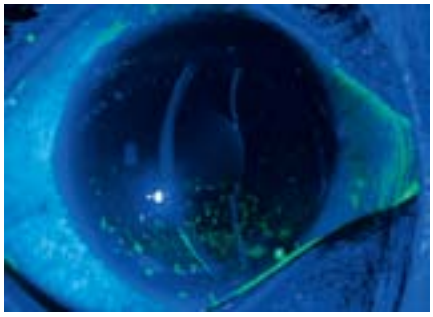


Fig. 3.1.9: Fluorescein staining of cornea

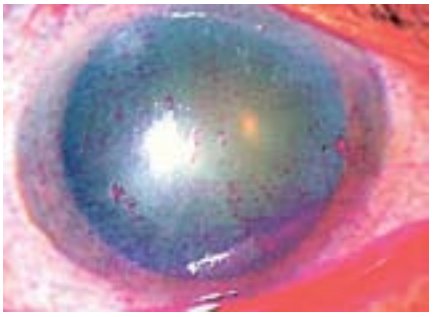


Fig. 3.1.10: Rose bengal staining of cornea

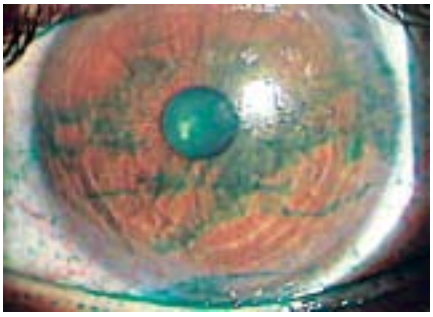


Fig. 3.1.11: Lissamine green staining of cornea

Pigments Deposition in the Cornea

Iron

Keratoconus *Fleischer's ring* **(Figs 3.2.1A and B)**	Epithelium
Old opacity *Hudson-Stahli line* **(Fig. 3.2.2)**	Epithelium
Pterygium *Stocker's line* **(Fig. 3.2.3)**	Epithelium
Filtering bleb *Ferry's line* **(Fig. 3.2.4)**	Epithelium
Siderosis **(Fig. 3.2.5)**	Stroma
Blood staining **(Fig. 3.2.6)**	Stroma

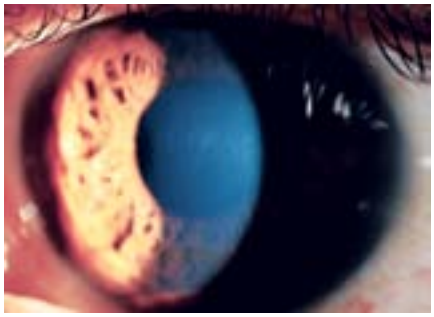


Fig. 3.2.1A: Fleischer's ring

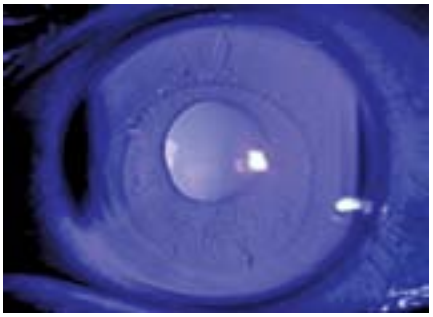


Fig. 3.2.1B: Fleischer's ring in cobalt blue filter

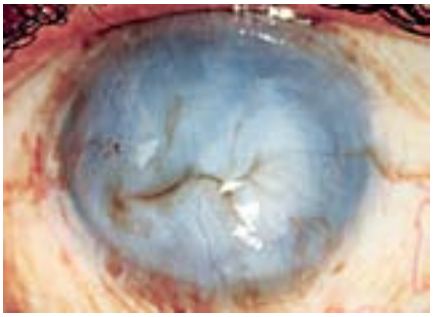


Fig. 3.2.2: Hudson-Stahli's line

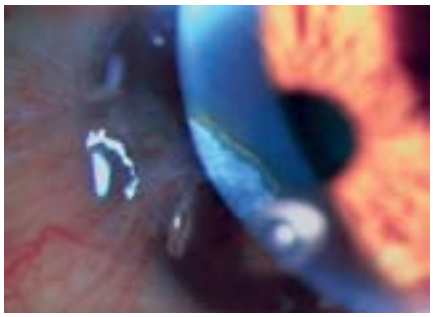


Fig. 3.2.3: Stocker's line

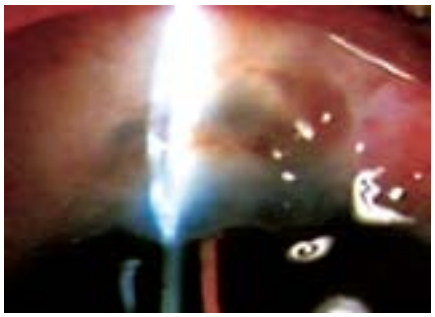


Fig. 3.2.4: Ferry's line

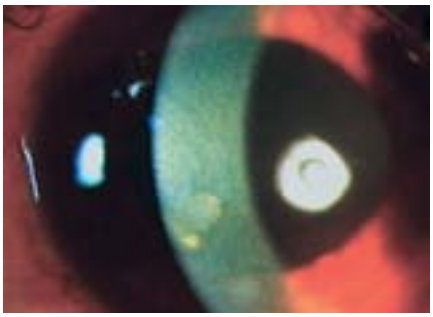


Fig. 3.2.5: Cornea in siderosis

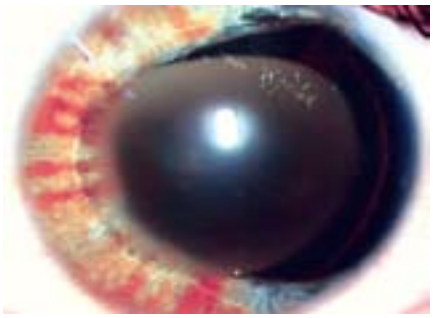


Fig. 3.2.6: Blood staining of cornea

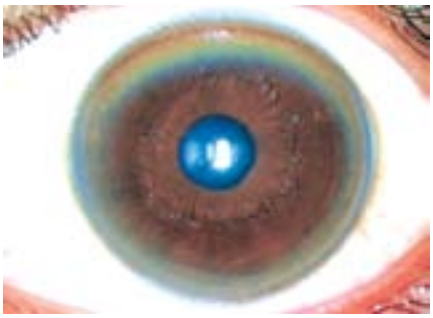


Fig. 3.2.7A: Kayser-Fleischer ring

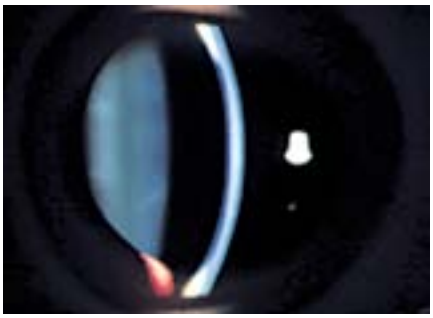


Fig. 3.2.7B: Kayser-Fleischer ring (slit section)

Copper

- Wilson's disease Descemet's level
  - *Kayser-Fleischer ring* **(Figs 3.2.7A and B)**

Melanin

- Pigment dispersion Endothelium **(Fig. 3.2.8)**
  - *Krukenberg's spindle*

Dye

- Tattooing of corneal opacity Subepithelial **(Fig. 3.2.9)**

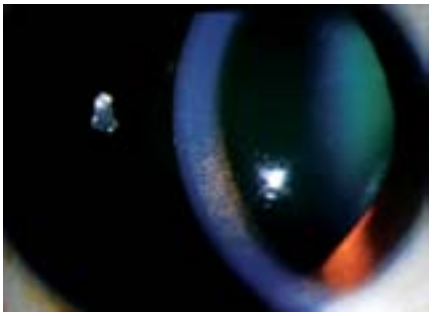


Fig. 3.2.8: Krukenberg's spindle—pigmentary glaucoma

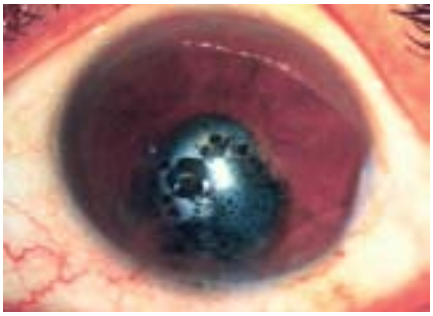


Fig. 3.2.9: Pigments in tattooing

Congenital Corneal Conditions

Microphthalmos (Fig. 3.3.1A)

- Unilateral (**Fig. 3.3.1A**) or bilateral congenital abnormality in which the axial length is reduced. Visual acuity is usually poor and depends upon the associated anomalies. Microphthalmos is commonly associated with coloboma (**Figs 3.3.1B to D**)
- *Treatment:* Not available, precautions to be taken during any intraocular surgery.

Fig. 3.3.1A: Microcornea with microphthalmos—RE

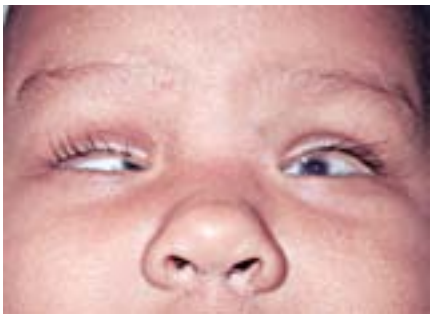


Fig. 3.3.1B: Severe microphthalmos—BE

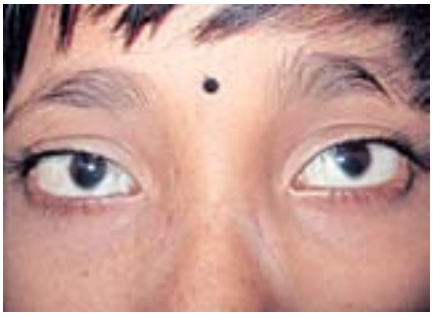


Fig. 3.3.1C: Microcornea with coloboma—BE

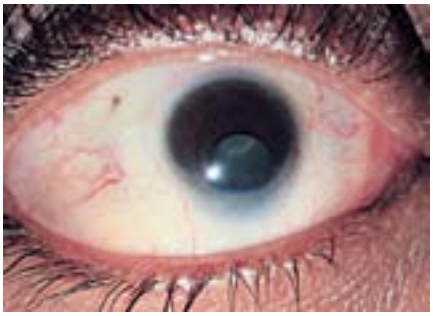


Fig. 3.3.1D: Microcornea, microphthalmos with coloboma

Nanophthalmos

- Uncommon, congenital, bilateral condition (**Figs 3.3.2A and B**), with small globe in all dimensions (*nano* means 'dwarf'). Anatomically, the eye is grossly normal. Very high hypermetropia, axial length is less than 20 mm. Fundus shows a crowded disk with vascular tortuosity and macular hypoplasia
- *Treatment:* High plus power spectacles correction, contact lens; and during cataract surgery in later life, precaution is to be taken to prevent complications, like choroidal effusion, expulsive hemorrhage, etc.

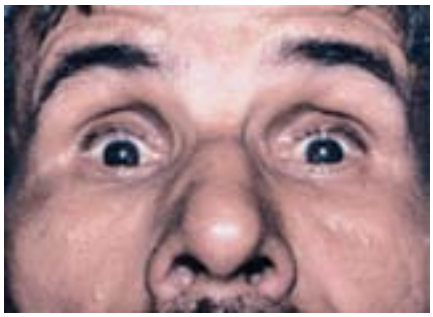


Fig. 3.3.2A: Nanophthalmos

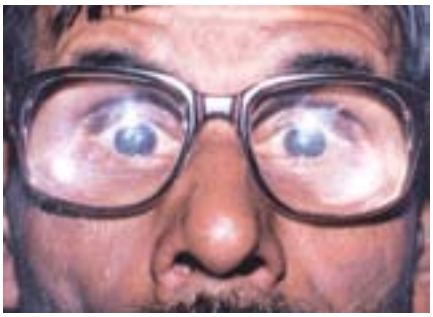


Fig. 3.3.2B: Nanophthalmos—high hypermetropia

Megalocornea

- Rare, congenital and bilateral condition. Corneal diameter is more than 13 mm (**Fig. 3.3.3**). Very deep anterior chamber. High myopia and astigmatism with good visual acuity. Normal intraocular pressure
- No active treatment is required.

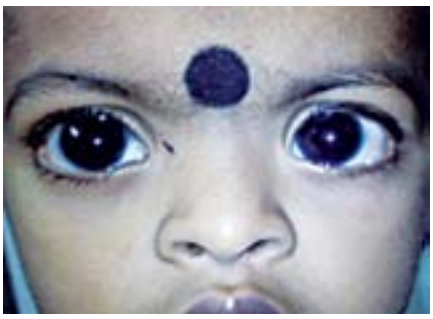


Fig. 3.3.3: Megalocornea

Buphthalmos

- Unilateral or bilateral condition in which the eyeball is large due to stretching as a result of increased intraocular pressure within first-three years of life **(Figs 3.3.4A and B)**
- Large cornea with variable scarring; horizontal tear of Descemet's membrane (Haab's striae); very deep anterior chamber; disk cupping
- *Treatment:* First reduce the IOP by different glaucoma surgical procedures applicable for congenital glaucoma. After that penetrating keratoplasty may be considered in selected cases.

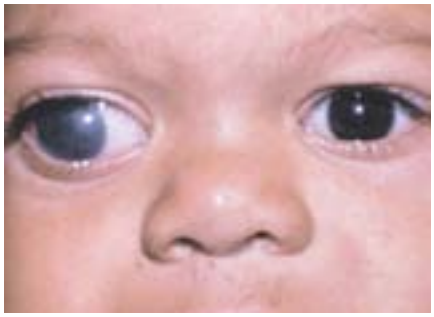


Fig. 3.3.4A: Buphthalmos—right eye

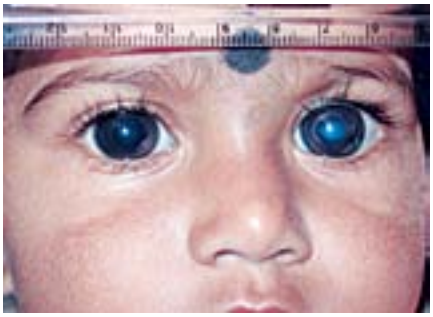


Fig. 3.3.4B: Buphthalmos—BE

Keratoglobus

- Congenital bilateral very rare condition. Mid-peripheral thinning resulting in protrusion or bulging of whole cornea, with an appearance of globular shape **(Figs 3.3.5A and B)**. Very deep anterior chamber
- Acute hydrops may occur in extreme cases **(Figs 3.3.5C and D)**
- *Treatment:* Contact lens and later penetrating keratoplasty under extreme precaution and with special technique.

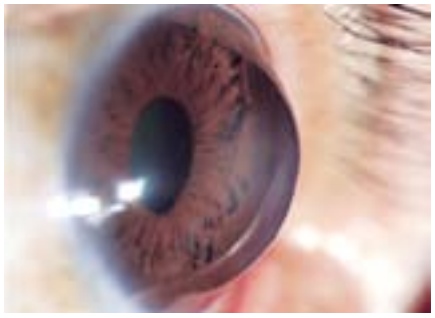


Fig. 3.3.5A: Keratoglobus

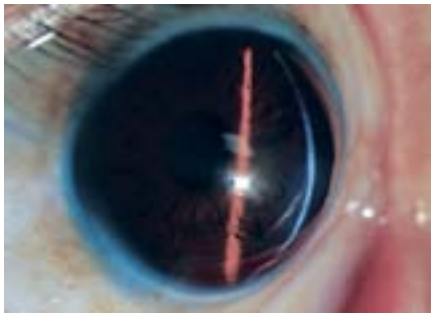


Fig. 3.3.5B: Keratoglobus

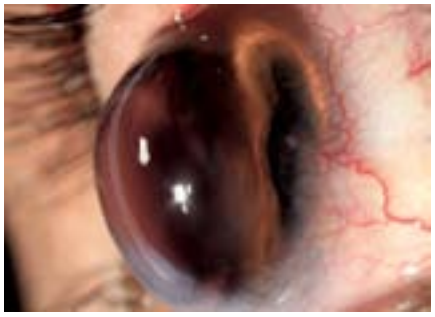


Fig. 3.3.5C: Keratoglobus—hydrops started in lower part

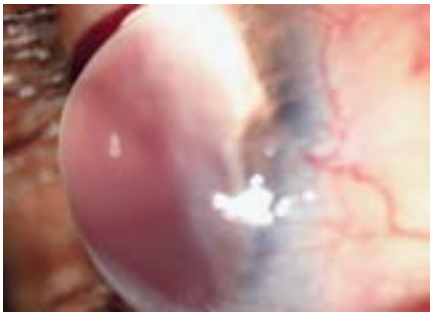


Fig. 3.3.5D: Keratoglobus—total hydrops after 7 days

Posterior Keratoconus

- Rare, unilateral or bilateral condition no progression
- The posterior cornea is having excavation and the anterior cornea does not protrude **(Figs 3.3.6A and B)**
- Common association with Peter's anomaly
- *Treatment:* Central lesion needs penetrating keratoplasty.

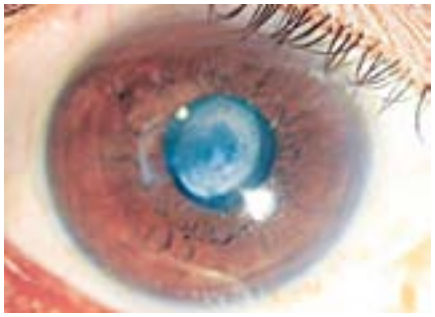


Fig. 3.3.6A: Posterior keratoconus

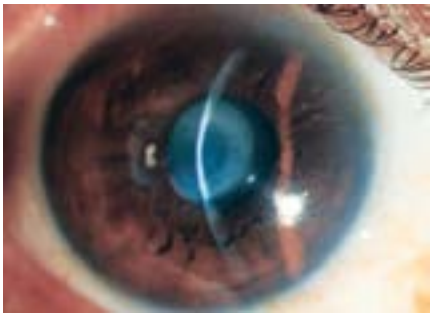


Fig. 3.3.6B: Posterior keratoconus

Peter's Anomaly (*See also* Figs 9.2.10 and 9.2.11)

- Central corneal opacity, due to defect in embryogenesis
- In milder form, it only causes posterior keratoconus
- But in most cases, associated with anterior polar cataract, iris adhesion, angle abnormalities, or secondary glaucoma **(Figs 3.3.7A and B)**
- *Treatment* is difficult and depends on clinical situation and visual potential of that eye.

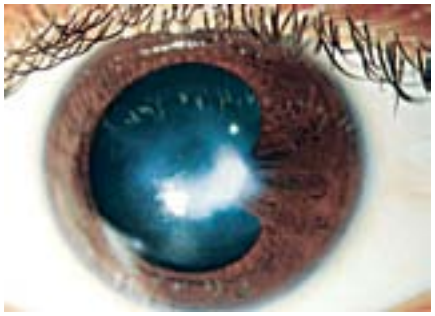


Fig. 3.3.7A: Peter's anomaly

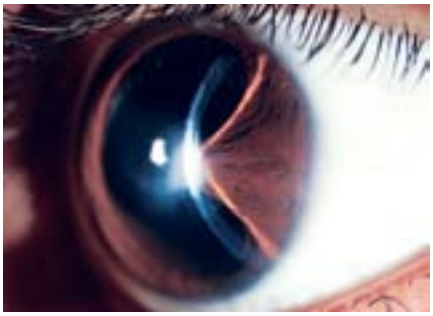


Fig. 3.3.7B: Peter's anomaly

Posterior Embryotoxon

- An unusual prominence of Schwalbe's line which is the peripheral termination of Descemet's membrane. It appears as ring opacity in the deeper layer of the cornea **(Figs 3.3.8A and B)**
- *Treatment* not required. Only IOP monitoring is required.

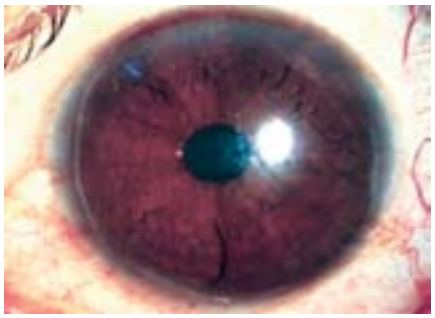


Fig. 3.3.8A: Posterior embryotoxon

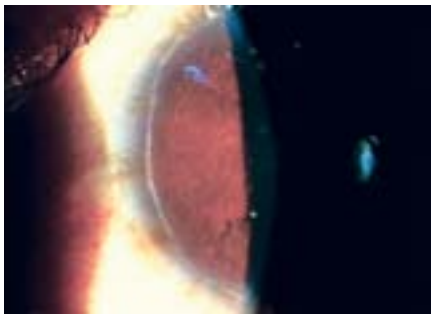


Fig. 3.3.8B: Posterior embryotoxon

Cornea Plana

- Rare, congenital, bilateral condition
- Severe decrease in corneal curvature, high hypermetropia and shallow anterior chamber **(Figs 3.3.9A and B)**
- But sometimes may be associated with high myopia
- May be associated with microcornea and partial sclerocornea **(Fig. 3.3.9C)**
- *Treatment:* Only refractive correction.

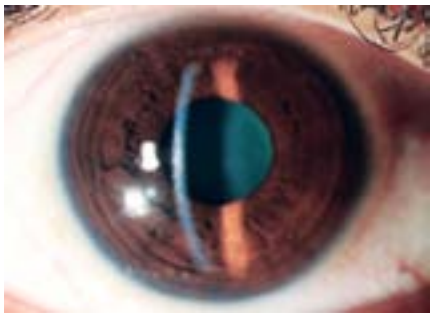


Fig. 3.3.9A: Cornea plana

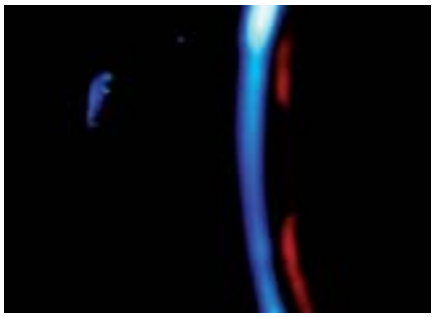


Fig. 3.3.9B: Cornea plana—slit section

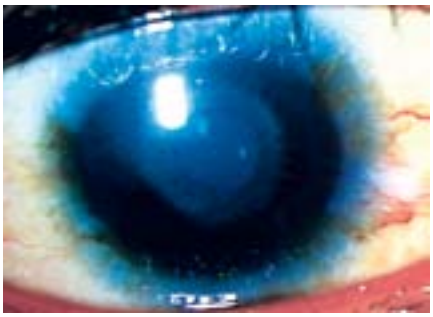


Fig. 3.3.9C: Cornea plana with sclerocornea

Sclerocornea

- Rare, congenital, usually bilateral condition; opacification and vascularization of peripheral or entire cornea **(Figs 3.3.10A and B)**. Cornea appears smaller if the scelerization is only peripheral **(Fig. 3.3.10C)**.
- May be associated with microphthalmos, blue sclera or cornea plana
- *Treatment:* Difficult. May be considered for penetrating keratoplasty.

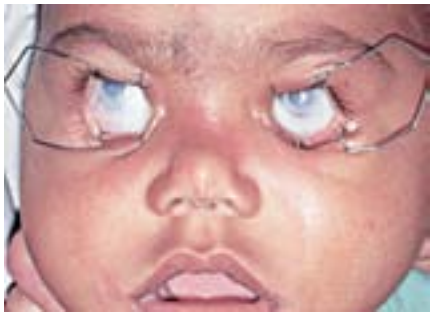


Fig. 3.3.10A: Sclerocornea—bilateral

Fig. 3.3.10B: Sclerocornea

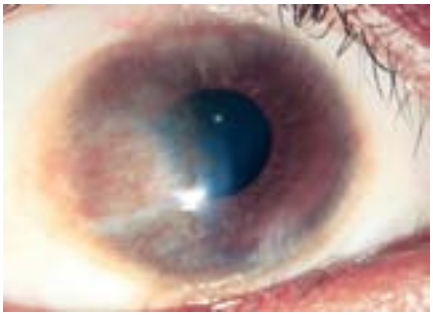


Fig. 3.3.10C: Sclerocornea—partial

Keratectasia

- Rare, usually unilateral condition, probably due to intrauterine keratitis or maternal vitamin A deficiency followed by perforation
- Severe corneal ectasia beyond the eyelid with opacification and vascularization **(Fig. 3.3.11)**
- *Treatment:* Not available. Only for cosmetic purpose, evisceration and followed by prosthesis.

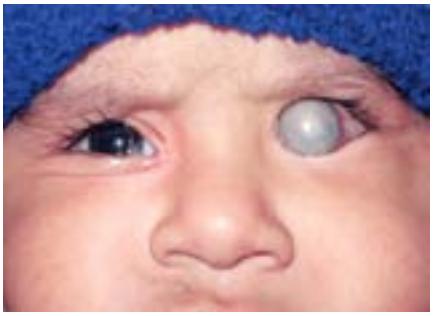


Fig. 3.3.11: Keratectasia

Corneal Edema and Bullous Keratopathy

Corneal Edema

- It is associated with increased in corneal thickness due to accumulation of fluid with variable degree of loss in corneal transparency
- Edema may be focal **(Figs 3.4.1A and B)** or generalized **(Figs 3.4.2A and B)**; may be with epithelial defect **(Fig. 3.4.3)** and may be severe **(Figs 3.4.4A and B)**.

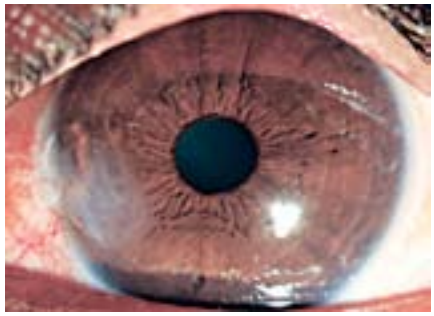


Fig. 3.4.1A: Corneal edema—focal

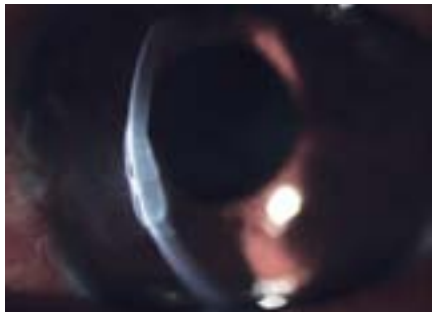


Fig. 3.4.1B: Corneal edema—focal on slit section

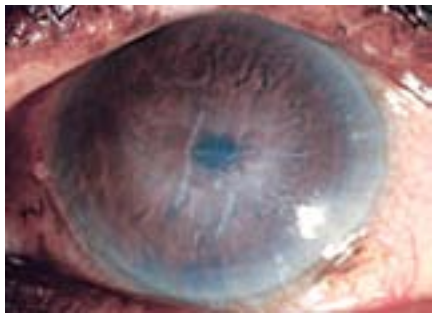


Fig. 3.4.2A: Corneal edema—generalized

Fig. 3.4.2B: Corneal edema—generalized on slit section

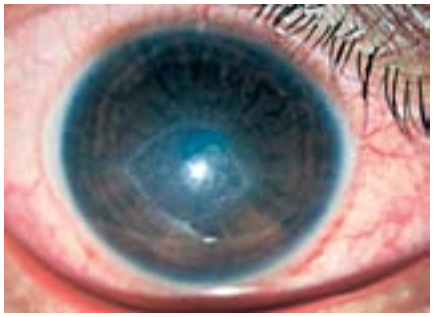


Fig. 3.4.3: Corneal edema with epithelial defect

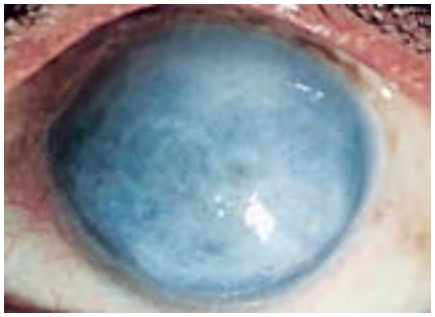


Fig. 3.4.4A: Severe corneal edema

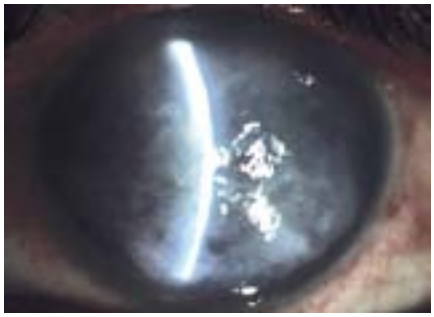


Fig. 3.4.4B: Severe corneal edema on slit section

Important Causes

- *Inflammatory*: Corneal ulcer, erosions, acute iridocyclitis, endotheliitis, etc. (due to endothelial damage) **(Figs 3.4.5 and 3.4.6)**
- *Traumatic:* Mechanical trauma and post-surgical—due to endothelial damage, especially, when vitreous remains adherent to it. Sometimes lens fragments in anterior chamber or haptic of IOL touching the endothelium may be responsible **(Figs 3.4.7 to 3.4.9)**
- *Increased IOP:* Acute edema in angle-closure glaucoma and epidemic dropsy glaucoma
- *Dystrophic or dysgenesis condition of the cornea:* Acute hydrops in keratoconus **(Fig. 3.4.10)**, Fuchs' endothelial dystrophy or ICE syndrome **(Figs 3.4.11A and B)**

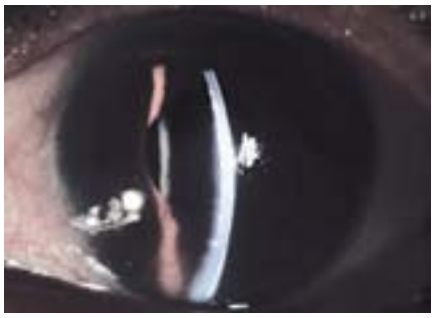


Fig. 3.4.5: Corneal edema in iridocyclitis

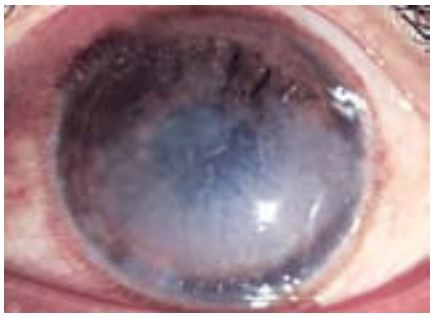


Fig. 3.4.6A: Corneal edema in disciform keratitis

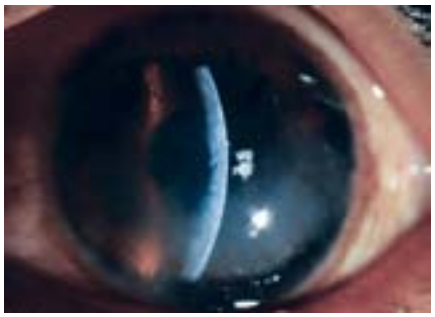


Fig. 3.4.6B: Corneal edema in disciform keratitis—slit section

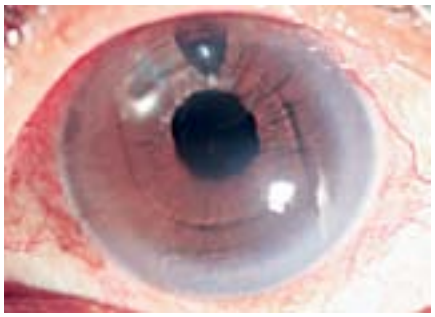


Fig. 3.4.7A: Corneal edema—AC IOL

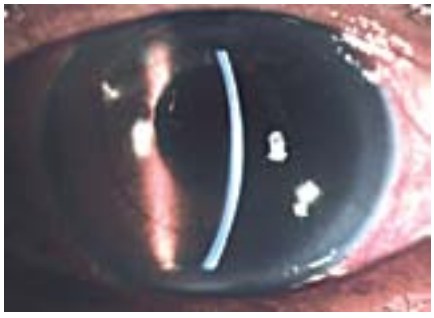


Fig. 3.4.7B: Corneal edema—AC IOL in slit section

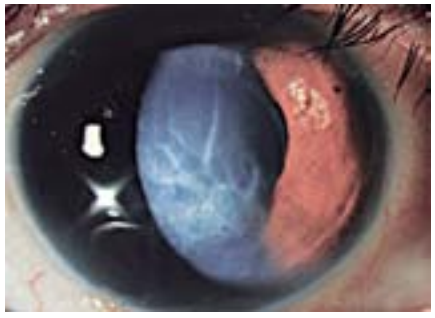


Fig. 3.4.8A: Corneal edema in PC IOL

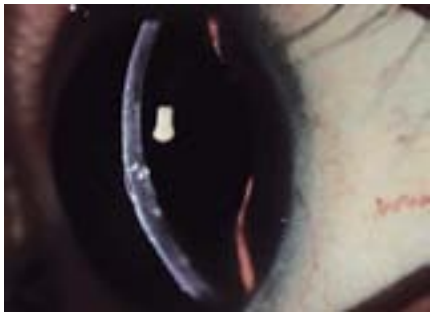


Fig. 3.4.8B: Corneal edema in PC IOL—slit section

Fig. 3.4.9: Corneal edema—vitreocorneal touch

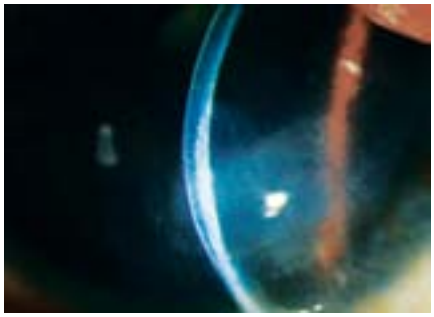


Fig. 3.4.10: Corneal edema in hydrops

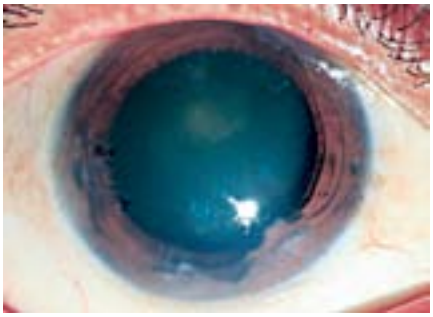


Fig. 3.4.11A: Corneal edema in ICE syndrome

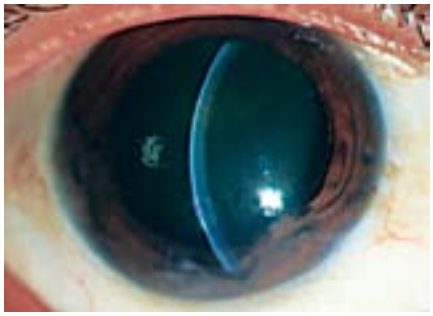


Fig. 3.4.11B: Corneal edema in ICE syndrome—slit section

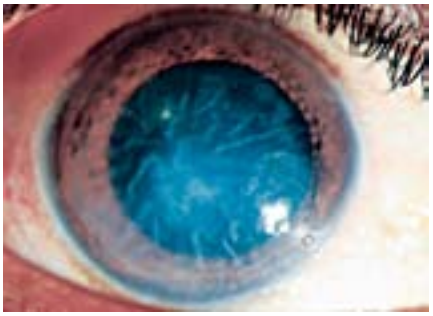


Fig. 3.4.12A: Corneal edema—calotropis latex

Fig. 3.4.12B: Corneal edema—calotropis latex in slit section

- Chronic edema in long standing cases—as in absolute glaucoma and buphthalmos
- Chemical/toxic injury to the cornea; as in alkali burn, toxic latex of plants, etc. **(Figs 3.4.12A and B)**
- *Hypoxia of the cornea:* As in contact lens wearer due to epithelial edema, as a result of prolonged deprivation of atmospheric oxygen.

Bullous Keratopathy

- In long standing cases, the epithelium tends to be raised into large vesicles or bullae, leading to *bullous keratopathy* **(Figs 3.4.13 to 3.4.18)**
- It may be phakic, aphakic or pseudophakic type. Vitreocorneal touch and AC IOL is an important factor in aphakic or pseudophakic bullous keratopathy. Phacoemulsification in compromised cornea or in suprahard cataracts is also responsible
- *Treatment:* Use of hot air (by hair-dryer); frequent instillation of concentrated sodium chloride (5%) solution, or ointment; bandage soft contact lenses, especially in case of ruptured bullae or multiple anterior stromal puncture with BCL; epithelium is stripped off, and is to be replaced with a thin conjunctival flap; Descemet's stripping endothelial keratoplasty (DSEK) **(Figs 3.4.19 and 3.4.20)** or PK to improve visual status.

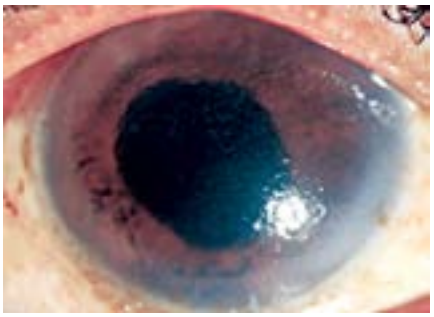


Fig. 3.4.13A: Corneal edema—epithelial bedewing

Fig. 3.4.13B: Corneal edema—epithelial bedewing in slit section

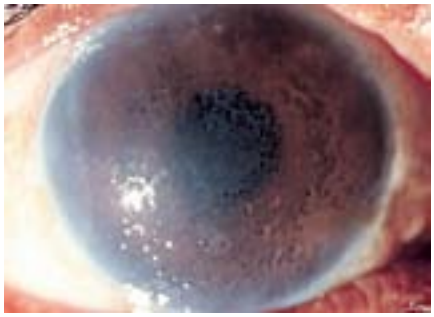


Fig. 3.4.14A: Corneal edema—microbullae

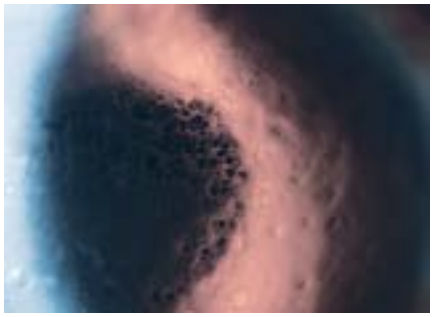


Fig. 3.4.14B: Corneal edema—microbullae

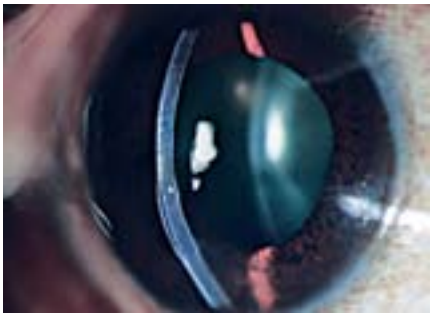


Fig. 3.4.15: Corneal edema—epithelial bulla

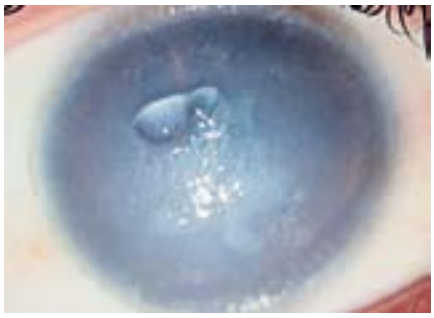


Fig. 3.4.16A: Bullous keratopathy in phakic eye

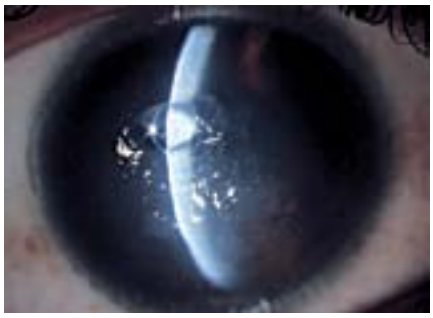


Fig. 3.4.16B: Bullous keratopathy in phakic eye—slit section

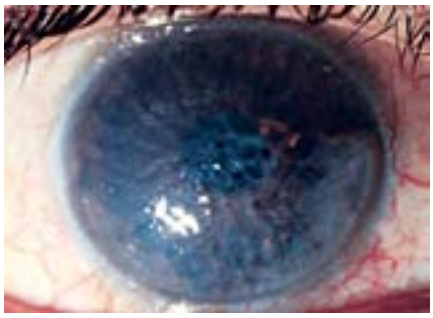


Fig. 3.4.17A: Bullous keratopathy—pseudophakic PC IOL

Fig. 3.4.17B: Bullous keratopathy—pseudophakic—slit section

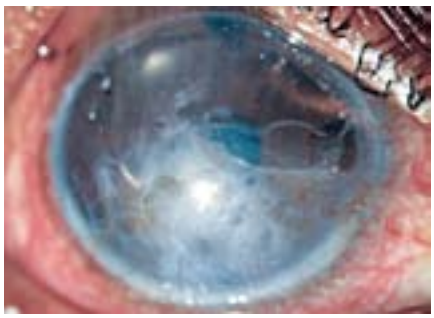


Fig. 3.4.18A: Pseudophakic bullous keratopathy—AC IOL

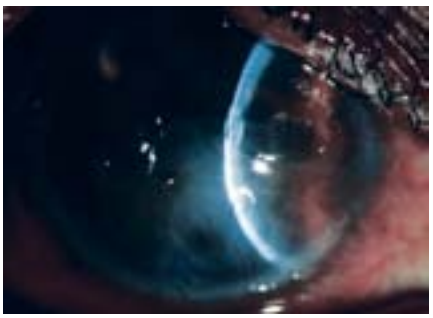


Fig. 3.4.18B: Pseudophakic bullous keratopathy—AC IOL

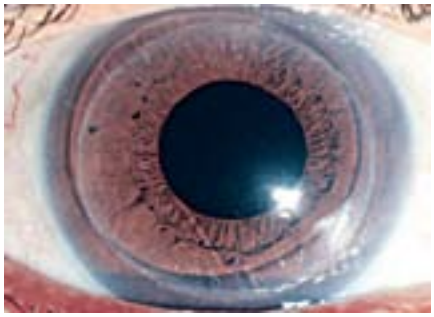


Fig. 3.4.19A: DSEK—in PBK of **Figure 3.4.17**

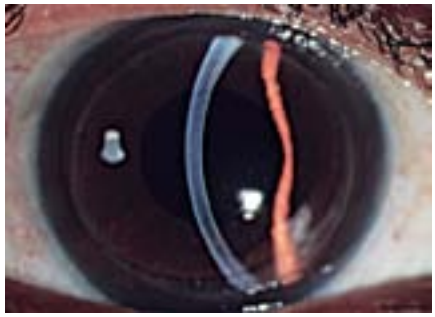


Fig. 3.4.19B: DSEK—in PBK of **Figure 3.4.17**

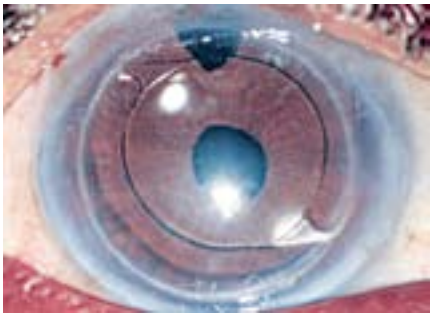


Fig. 3.4.20: DSEK in AC IOL of **Figure 3.4.18**

Keratitis: Suppurative/Infectious

- **Definition of keratitis:** *Keratitis is the inflammation of the cornea.*
- A corneal ulcer or suppurative keratitis is defined as a loss of corneal epithelium with underlying stromal infiltration and suppuration associated with signs of inflammation with or without hypopyon.

Suppurative Keratitis

- Exogenous infection is most common and almost always starts as a small infiltration with some kind of trauma, mechanical, with vegetable materials or foreign body **(Figs 3.5.1 and 3.5.2)**
- Sometimes associated with underlying other ocular factors, e.g. corneal erosion, bullous keratopathy, herpetic keratitis, lagophthalmos, dry eyes, chronic dacryocystitis, etc.

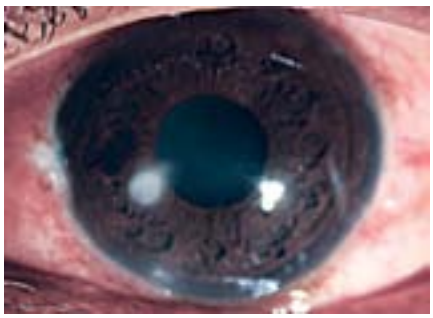


Fig. 3.5.1: Corneal infiltration

Fig. 3.5.2: Linear nail injury with corneal infiltration

- Other factors like, use of topical steroids, diabetes, immuno compromise states are also important
- It may be *bacterial*, *fungal*, or due to *Acanthamoeba.*

Bacterial Keratitis/Ulcer

- Most frequent causative agents are *Staph. aureus*, *Pseudomonas* and *Strept. pneumoniae.* Signs vary with the severity and on causative agents.
- Lid edema, marked ciliary congestion; epithelial breakdown followed by stromal suppuration; ulcer usually starts as a grayish-white circumscribed infiltration; edema of the surrounding tissue; margins are over hanging and the floor is covered by necrotic material **(Figs 3.5.3 and 3.5.4)**

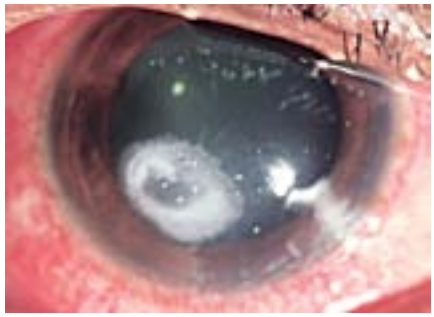


Fig. 3.5.3: Bacterial corneal ulcer—wet look

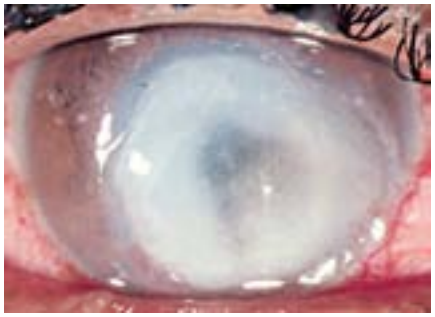


Fig. 3.5.4: Severe corneal ulcer with stromal melting

- Overall look of the ulcer is wet; hypopyon may be present in variable amount with flat border
- Secondary anterior uveitis and glaucoma in some cases.

Special Features of Specific Bacterial Keratitis

- ***Staphylococcal keratitis*** **(Figs 3.5.5A and B):** Well defined grayish-white or creamy stromal infiltrate which may progress to form dense stromal abscess. The surrounding cornea is relatively clear.
- ***Pneumococcal keratitis*** **(Fig. 3.5.6):** Spread superficially with a serpiginous leading edge and associated with severe anterior uveitis with hypopyon formation. The surrounding cornea is relatively clear.
- ***Pseudomonas keratitis*** **(Fig. 3.5.7):** Rapidly spreading, melting suppurative lesion associated with hypopyon and greenish mucopurulent discharge. 'Ground glass' appearance of the surrounding cornea.
- ***Enterobacteriaceae (E. coli, Proteus*** **or** ***Klebsiella)*** **(Figs 3.5.8A and B):** Shallow ulcer with pleomorphic grayish-white necrotic areas. Sometime they produce ring shaped corneal infiltrates.

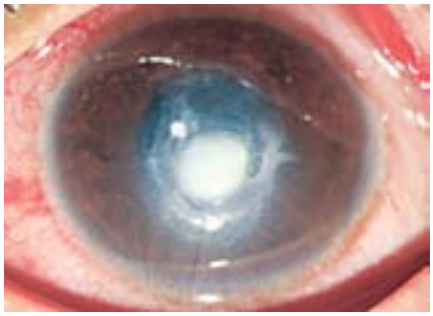


Fig. 3.5.5A: Bacterial ulcer—staphylococcal

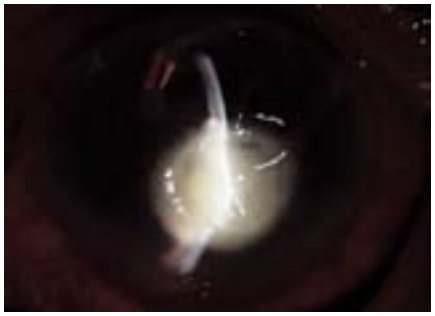


Fig. 3.5.5B: Bacterial ulcer—staphylococcal

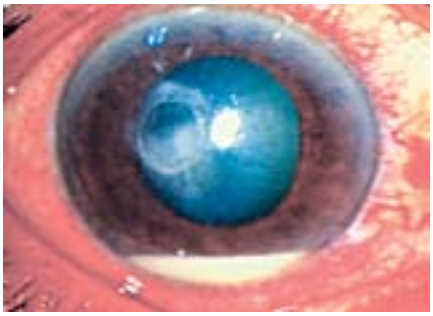


Fig. 3.5.6: Bacterial ulcer with hypopyon—pneumococcal

Fig. 3.5.7: Bacterial ulcer—*Pseudomonas*

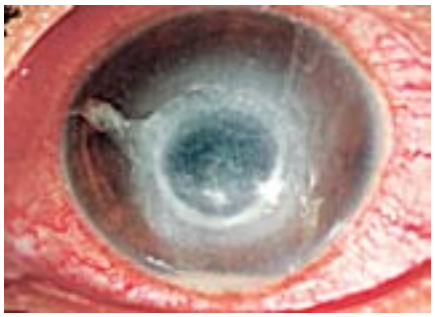


Fig. 3.5.8A: Bacterial ulcer—*Enterobacteriaceae*

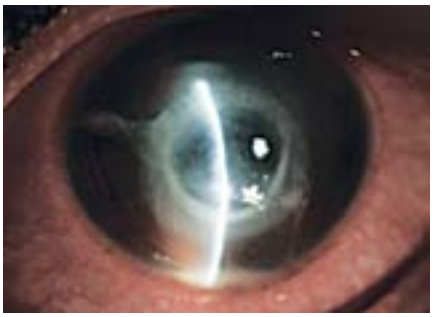


Fig. 3.5.8B: Bacterial ulcer—*Enterobacteriaceae*

- ***Corneal abscess* (Figs 3.5.9A and B):** Localized suppuration in the deeper stroma under intact corneal epithelium. Usually associated with staphylococcal infection
- *Treatment:* Scraping and smear preparation to identify the microorganism quickly and then prompt initiation of therapy with frequent (hourly) topical broad-spectrum antibiotics. *Other therapy* includes—atropine, treating secondary glaucoma. Therapeutic PK is required in extreme cases **(Figs 3.5.10A and B)**.

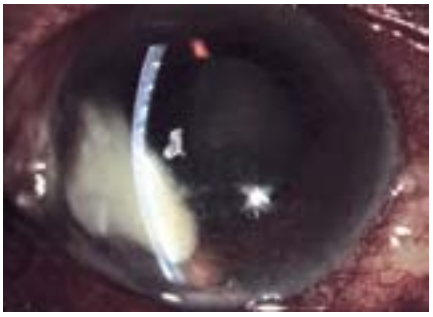


Fig. 3.5.9A: Corneal abscess

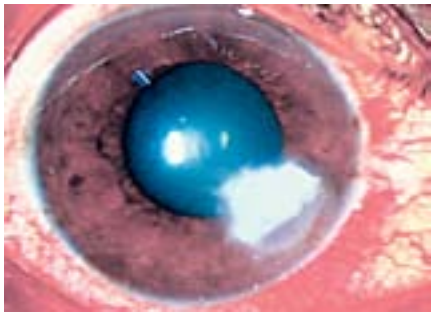


Fig. 3.5.9B: Corneal abscess

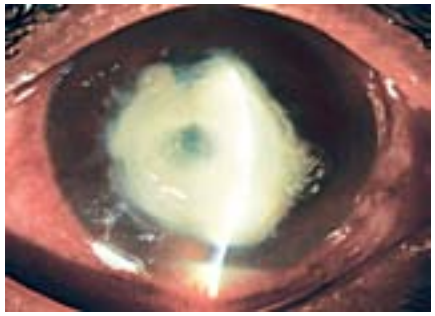


Fig. 3.5.10A: Bacterial ulcer—non-responding and impending perforation

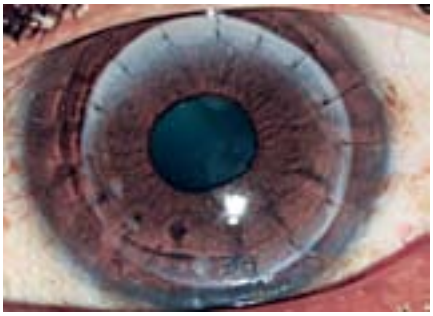


Fig. 3.5.10B: Same eye after therapeutic penetrating keratoplasty

Fungal Keratitis (Corneal Ulcer) or Keratomycosis

- Causative agent may be *Filamentous* fungi or *Candida.* Typically preceded by ocular trauma with agricultural and vegetable matters
- Relatively painless, dry looking, elevated yellowish-white lesion with indistinct margin. Delicate, feathery, finger-like projections into the adjacent stroma. Massive dense hypopyon which is immobile with convex upper border (**Figs 3.6.1A to D)**
- Slowly progressive stromal destruction may lead to corneal perforation with its sequelae.
- *Candida keratitis* **(Figs 3.6.2A and B):** Gray-white infiltrate (often as collar-button abscess) similar to bacterial ulcer. Also common in suture-related keratitis following PK (**Fig. 3.6.3)**
- *Filamentary keratitis* **(Figs 3.6.4 and 3.6.5):** Typical feathery appearance with finger-like projection and satellite lesion
- *Dematiaceous fungal keratitis* **(Figs 3.6.6A to C):** Often produce pigments on the surface of the ulcer.

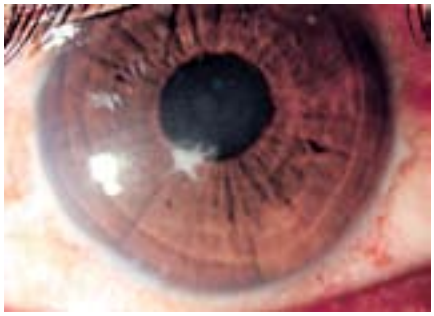


Fig. 3.6.1A: Fungal corneal infiltrates

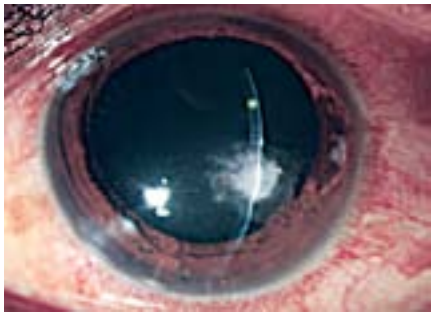


Fig. 3.6.1B: Fungal keratitis—small

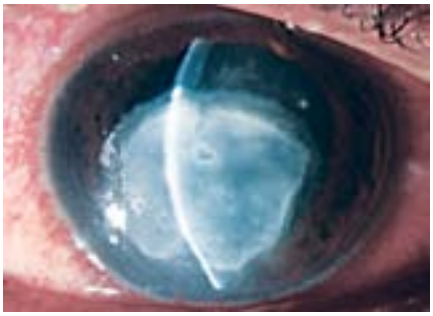


Fig. 3.6.1C: Fungal keratitis—dry look and elevated

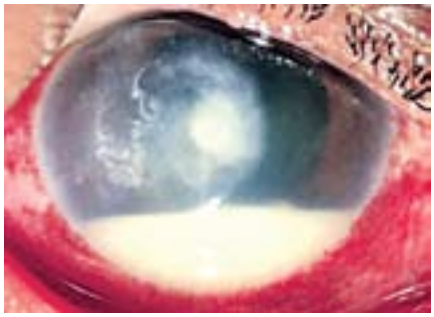


Fig. 3.6.1D: Fungal keratitis—convex hypopyon

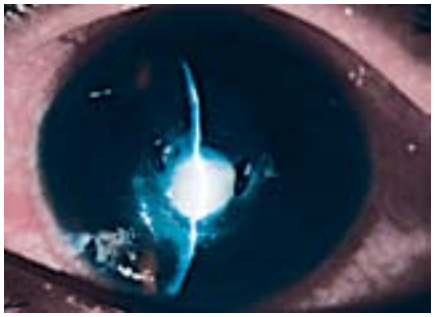


Fig. 3.6.2A: Fungal corneal ulcer—*Candida*

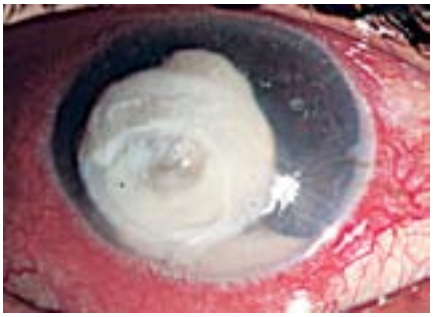


Fig. 3.6.2B: Fungal corneal ulcer—*Candida*

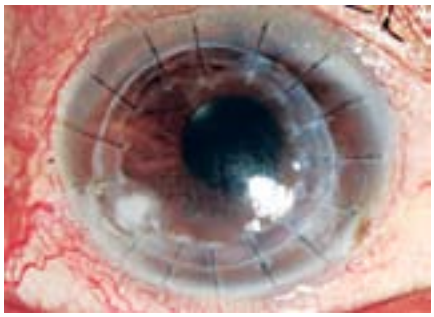


Fig. 3.6.3: Fungal corneal ulcer—*Candida*

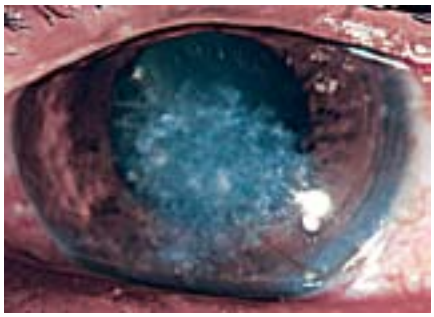


Fig. 3.6.4A: Fungal keratitis by filamentary fungus—feathery edge

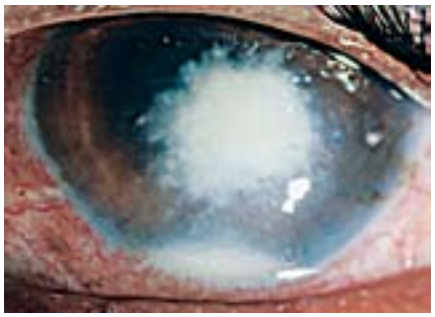


Fig. 3.6.4B: Fungal ulcer—ulcer progression in same eye of **Figure 3.6.5A**

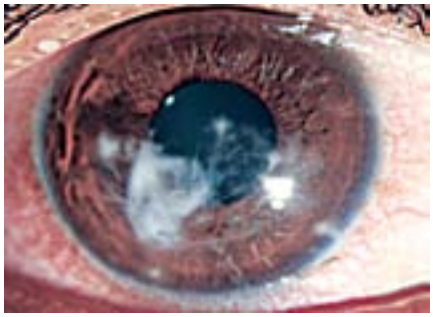


Fig. 3.6.5: Fungal ulcer with satellite lesions—filamentary

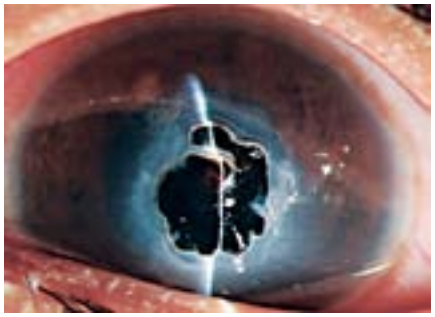


Fig. 3.6.6A: Dematiaceous fungal keratitis

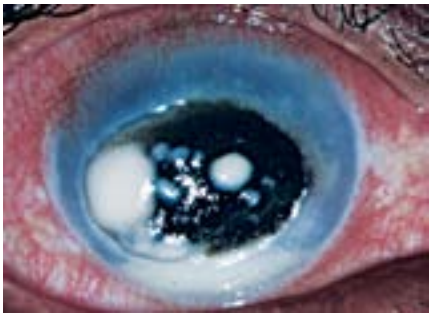


Fig. 3.6.6B: Dematiaceous fungal keratitis

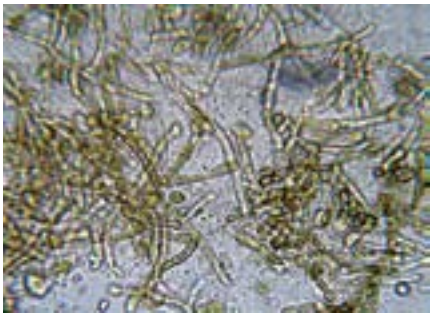


Fig. 3.6.6C: Pigment producing dematiaceous fungi in KOH mount preparation

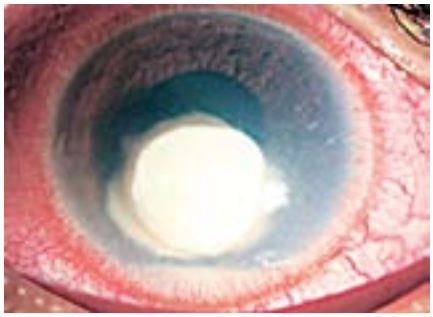


Fig. 3.6.7A: Fungal keratitis—*Candida*

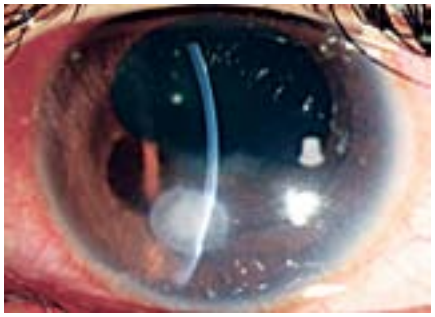


Fig. 3.6.7B: After intracameral amphotericin B injection—same eye in **Figure 3.6.7A**

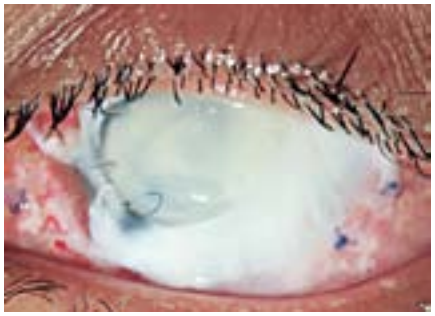


Fig. 3.6.8A: Fungal keratitis with melting following lamellar keratoplasty

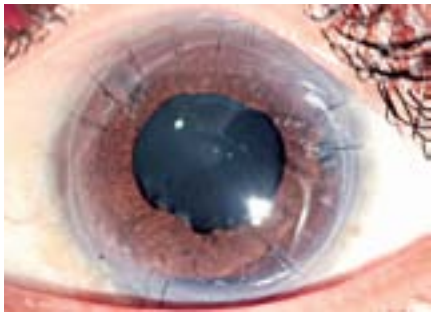


Fig. 3.6.8B: Therapeutic PK in same eye **(Fig. 3.6.8A)** after 6 weeks

- *Treatment:* Topical antifungal agents—natamycin (5%), amphotericin B (0.15%), or voriconazole—1–2 hourly and systemic antifungals in selected cases. Intracameral amphotericin B **(Figs 3.6.7A and B)** may be tried in deeper ulcers and a therapeutic PK may require in severe nonresponsive cases **(Figs 3.6.8A and B)**.

Acanthamoeba Keratitis

- Very rare unilateral keratitis that typically affects the soft contact lens wearer
- Pseudodendrite **(Fig. 3.7.1);** radial keratoneuritis **(Fig. 3.7.2)**; progressive chronic stromal keratitis with recurrent breakdown of corneal epithelium and stromal melting **(Figs 3.7.3A and B)**
- Paracentral ring-shaped ulcer **(Figs 3.7.4 and 3.7.5)** or abscess is the hallmark of advanced infection
- *Treatment:* PHMB, propamidine isethionate, chlorhexidine (0.02%) eyedrops, and neomycin eye ointment, etc. Therapeutic PK in resistance cases.

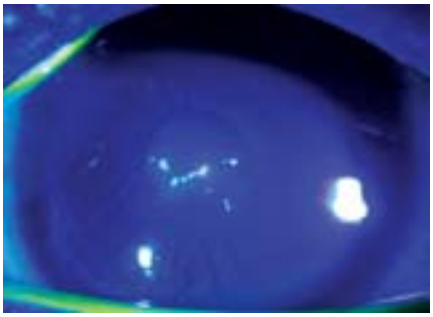


Fig. 3.7.1: *Acanthamoeba* keratitis—pseudodendrite

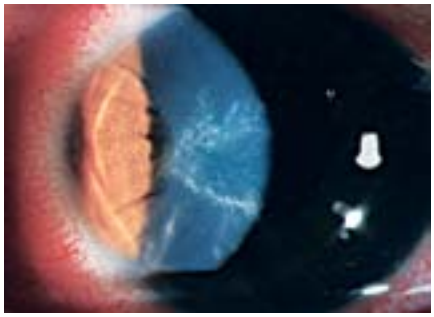


Fig. 3.7.2: *Acanthamoeba* keratitis—radial keratoneuritis

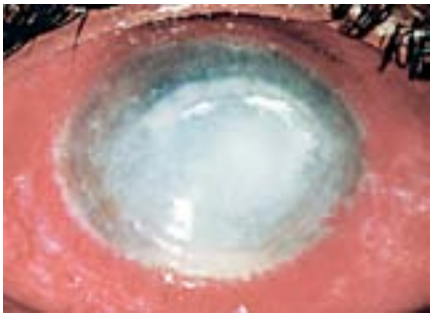


Fig. 3.7.3A: *Acanthamoeba* keratitis—stromal melting

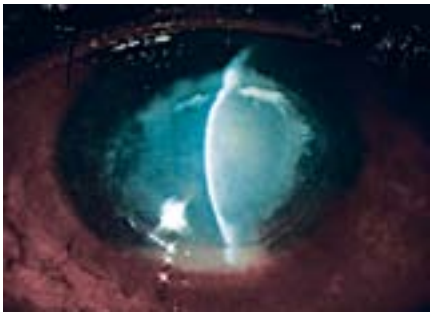


Fig. 3.7.3B: *Acanthamoeba* keratitis—stromal melting

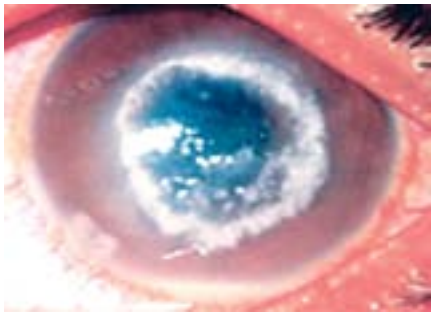


Fig. 3.7.4: *Acanthamoeba* keratitis—ring infiltrates

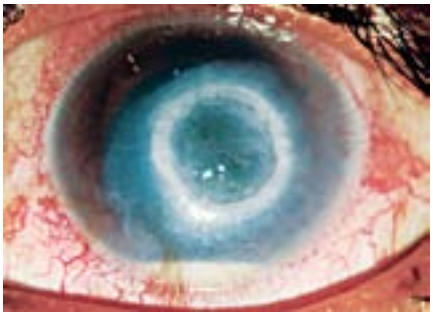


Fig. 3.7.5: *Acanthamoeba* keratitis—ring-shaped ulcer

Perforated Corneal Ulcer

- Corneal perforation may occur in any type of corneal ulcer in advanced cases
- Prior to that the patient may present with *Descemetocele*, i.e. herniation of the elastic Descemet's membrane as a transparent vesicle **(Figs 3.8.1A and B)**
- The other types of presentation are:
  - *Corneal fistula:* Seidel's test is important to determine it **(Figs 3.8.2A and B)**
  - *Cystoids cicatrix:* Seidel's test may also be positive **(Figs 3.8.3A and B)**
  - *Iris prolapse:* If the perforation is large peripheral or paracentral **(Figs 3.8.4A and B)**
  - *Pseudocornea formation* **(Figs 3.8.5A and B)**
  - *Ectatic cicatrix* **(Fig. 3.8.6):** Thinned, scarred cornea bulges even under normal IOP
  - *Anterior staphyloma:* Partial or total **(Figs 3.8.7A and B)**
  - *Phthisis bulbi* **(Fig. 3.8.8)**
  - *Endophthalmitis and Panophthalmitis* **(Figs 3.8.9 and 3.8.10)**.

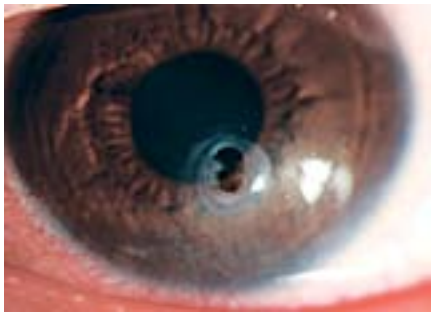


Fig. 3.8.1A: Descemetocele

Fig. 3.8.1B: Descemetocele—slit section

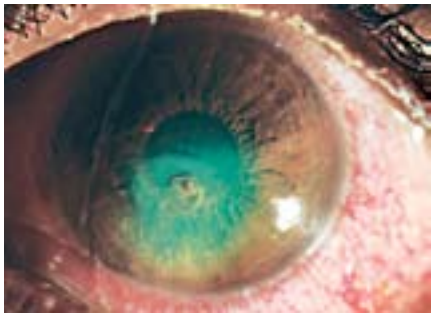


Fig. 3.8.2A: Corneal fistula

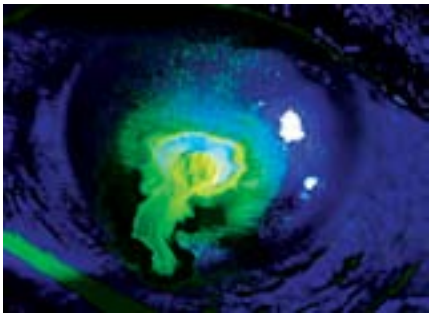


Fig. 3.8.2B: Corneal fistula—Seidel's test positive

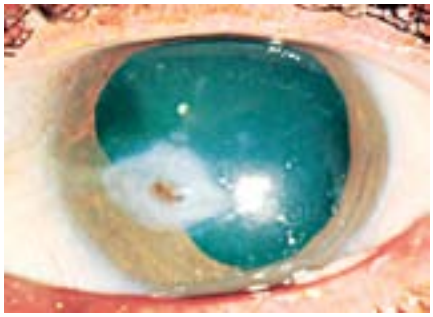


Fig. 3.8.3A: Cystoid cicatrix

Fig. 3.8.3B: Cystoid cicatrix—Seidel's test positive

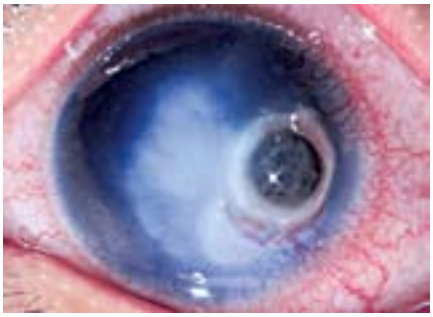


Fig. 3.8.4A: Perforated corneal ulcer with iris prolapse

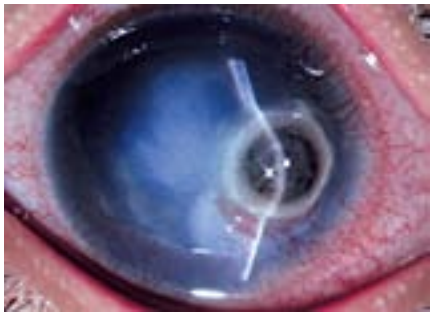


Fig. 3.8.4B: Corneal ulcer wirh perforation

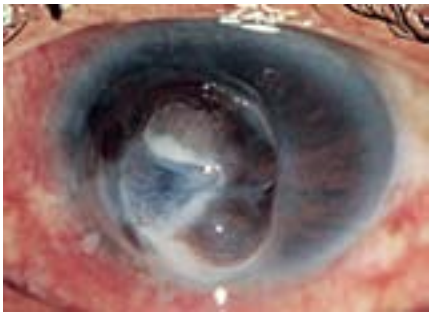


Fig. 3.8.5A: Pseudocornea formation

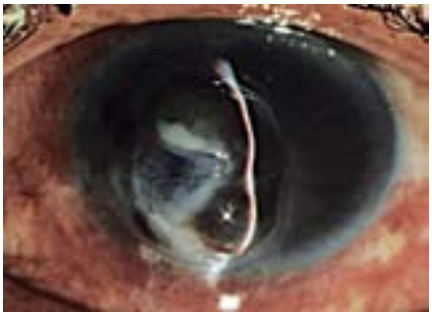


Fig. 3.8.5B: Pseudocornea formation—slit section

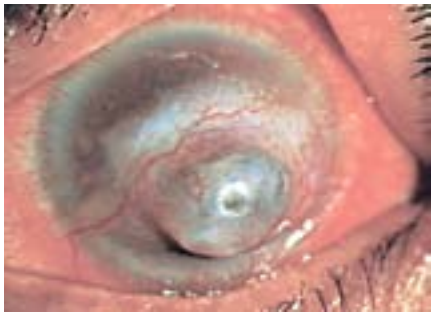


Fig. 3.8.6: Healing ulcer—ectatic cicatrix

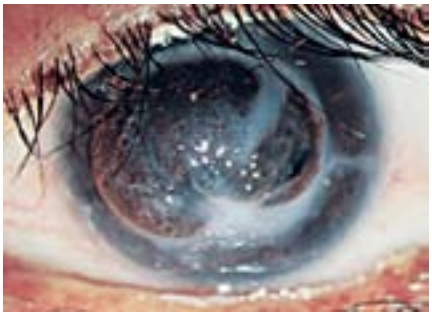


Fig. 3.8.7A: Anterior staphyloma—partial

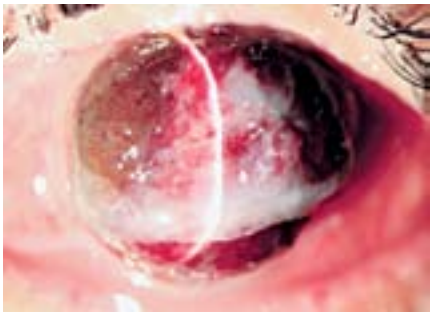


Fig. 3.8.7B: Anterior staphyloma—total—slit section

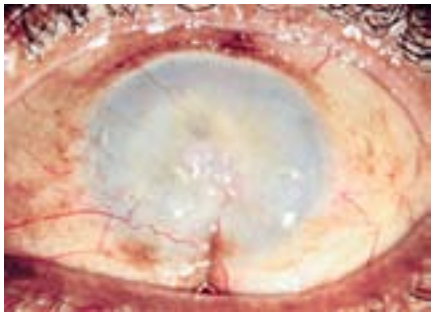


Fig. 3.8.8: Corneal ulcer—phthisis bulbi

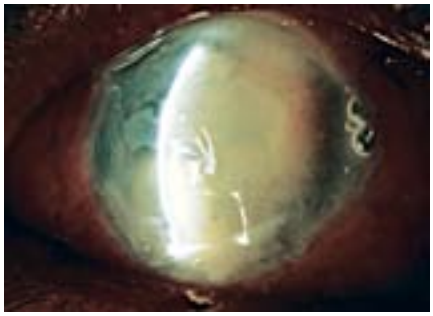


Fig. 3.8.9: Corneal ulcer—endophthalmitis

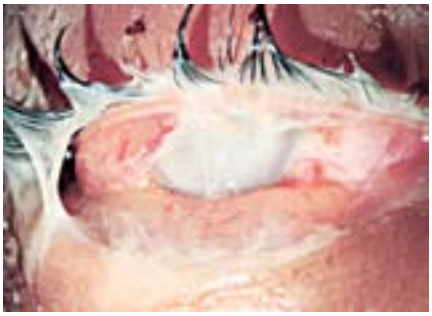


Fig. 3.8.10: Corneal ulcer—panophthalmitis

Sequel of Healded Corneal Ulcer

- Nebula **(Fig. 3.8.11)**, macula **(Figs 3.8.12A and B)**, leucoma **(Figs 3.8.13A and B)**, adherent leucoma **(Figs 3.8.14A and B)**, vascularization may occur in any form of opacity **(Figs 3.8.15 and 3.8.16)**
- Sudden perforation may result in extrusion of the contents of the globe **(Figs 3.8.17 and 3.8.18)**
- After healing it may lead to adherent leucoma, anterior staphyloma or phthisis bulbi, depending upon the size of perforation.

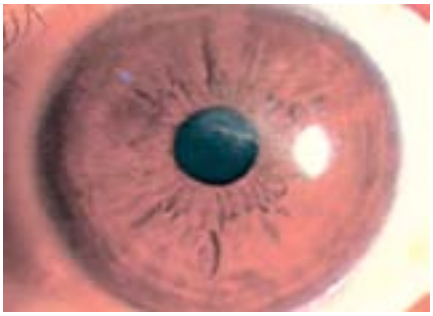


Fig. 3.8.11: Corneal opacity—faint opacity

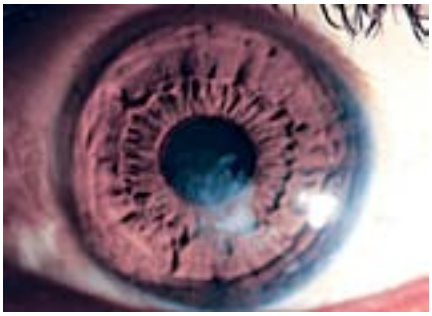


Fig. 3.8.12A: Corneal opacity—macula—partial thickness involvement

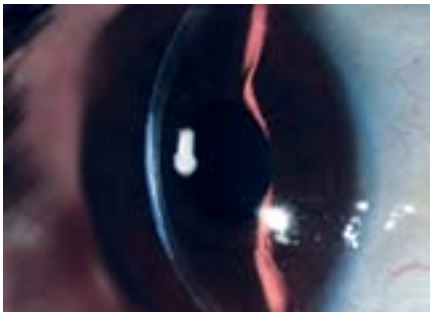


Fig. 3.8.12B: Corneal opacity—macula—partial thickness involvement

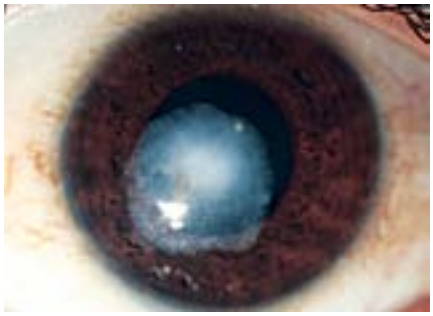


Fig. 3.8.13A: Corneal opacity—leucoma

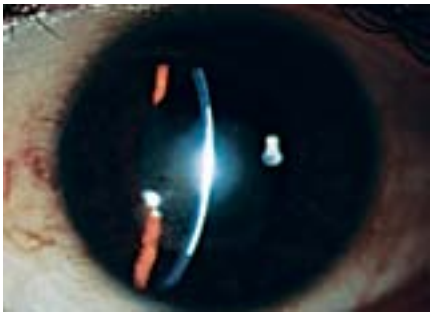


Fig. 3.8.13B: Corneal opacity—leucoma—full thickness involvement

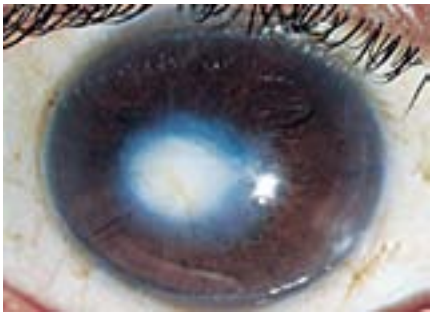


Fig. 3.8.14A: Corneal opacity—adherent leucoma

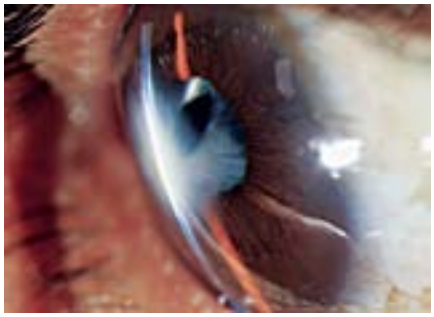


Fig. 3.8.14B: Corneal opacity—adherent leucoma

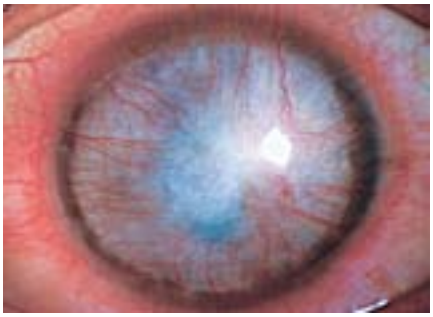


Fig. 3.8.15: Leucoma with severe vascularization

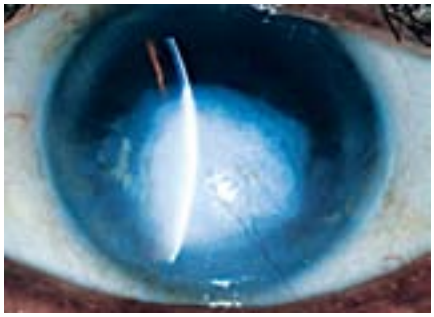


Fig. 3.8.16: Vascularized corneal opacity

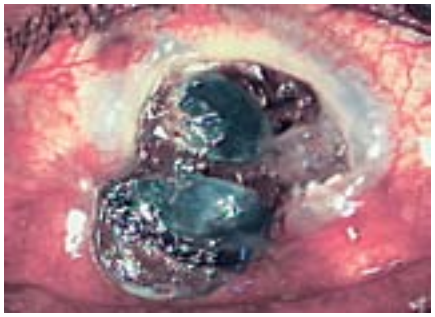


Fig. 3.8.17: Perforation—extrusion of contents

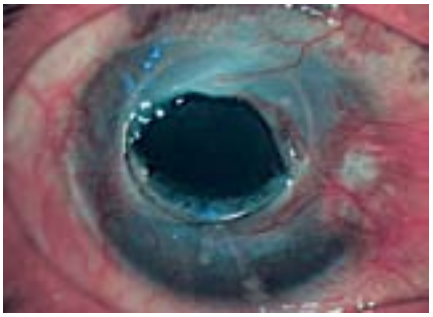


Fig. 3.8.18: Perforation—with exposed IOL

Viral Keratitis

Herpes Simplex Virus (HSV) Keratitis

- Infection with herpes simplex virus (HSV) is extremely common
- The majority of the patients present with recurrent lesion.

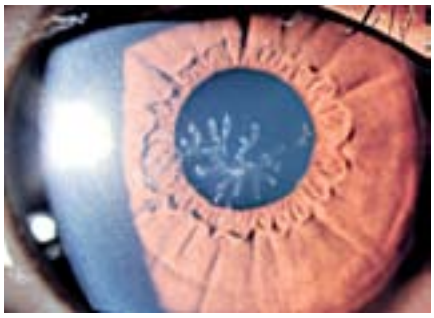


Fig. 3.9.1A: Primary HSV dendritic keratitis

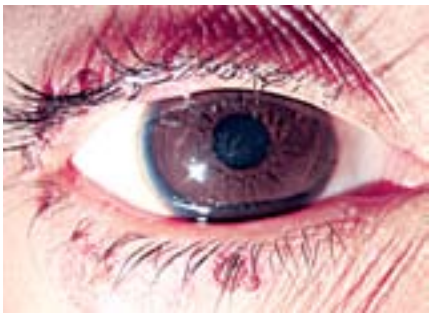


Fig. 3.9.1B: Primary HSV keratitis with lid lesions same eye as **Figure 3.9.1A**

Primary HSV Infection

- Usually subclinical or may present with mild ocular problem
- Typically, it occurs between 6 months and 5 years of age
- The main lesion is acute follicular conjunctivitis
- Fine epithelial keratitis may be present, which sometimes progresses into dendritic figure **(Fig. 3.9.1A)**
- Vesicular eruptions and edema of the lids **(Fig. 3.9.1B)**
- It seldom causes serious ocular problem
- *Treatment:* Acyclovir eye ointment—5 times daily for 7–10 days.

Recurrent HSV Keratitis

- The virus travels down along the sensory division of the fifth cranial nerve to affect the target tissues
- Associated with some predisposing factors.

Dendritic HSV Keratitis

- Initially starts as superficial punctuate erosions which coalesce **(Fig. 3.9.2A)**. They send out lateral branches with knobbed ends, to form *'dendritic'* or *'tree-like'* figure and this is *pathognomonic* **(Figs 3.9.2B and C)**

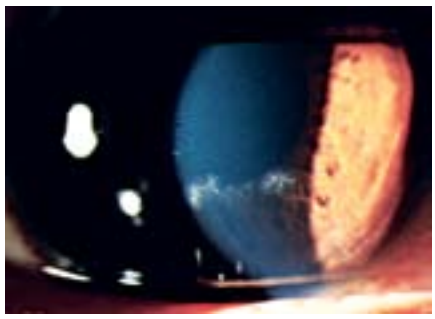


Fig. 3.9.2A: Early dendritic keratitis

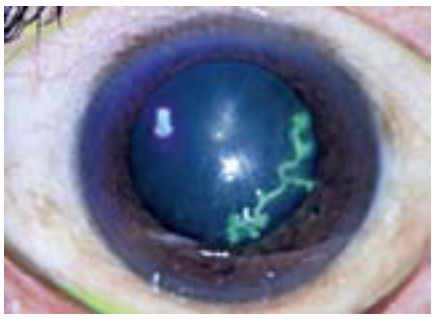


Fig. 3.9.2B: Dendritic keratitis

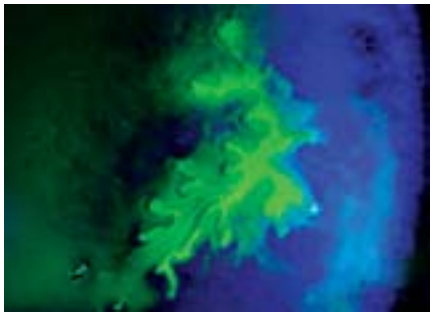


Fig. 3.9.2C: Dendritic keratitis

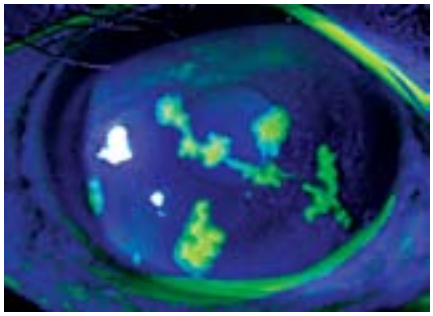


Fig. 3.9.2D: Dendritic keratitis—multiple dendrites

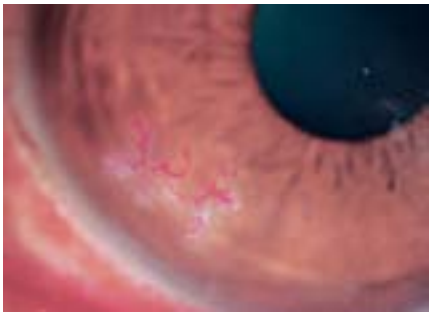


Fig. 3.9.2E: Dendritic keratitis—rose bengal stain

- Bed of the ulcer stains with fluorescein **(Fig. 3.9.2D)** and the swollen diseased cells at the margin take up rose bengal stain **(Fig. 3.9.2E)**
- *Treatment:* Debridement, topical antiviral (acyclovir), mild cycloplegic, oral acyclovir to prevent recurrence.

Geographical Keratitis

- Larger epithelial lesion—*'geographical'* or *'amoeboid'* configuration **(Figs 3.9.3A and B)**
- May occur as a continued enlargement of dendritic keratitis (dendrogeographic) **(Figs 3.9.3C and D)**
- Likely to occur following inadvertent use of topical steroids
- *Treatment:* Similar to dendritic keratitis.

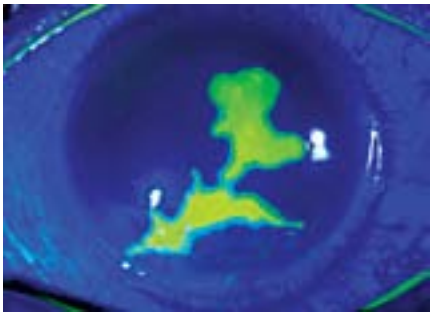


Fig. 3.9.3A: HSV—geographical keratitis

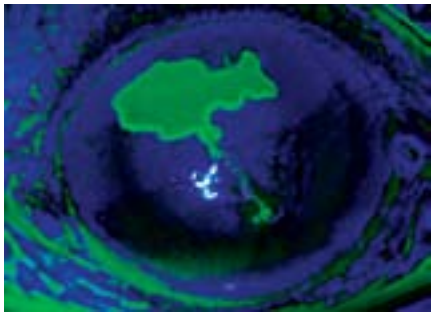


Fig. 3.9.3B: HSV—geographical keratitis

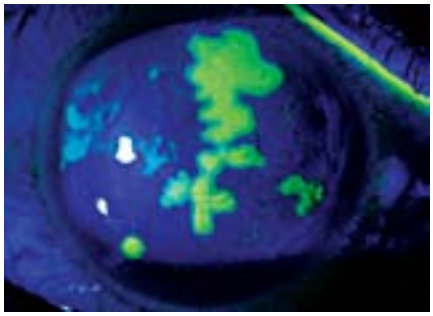


Fig. 3.9.3C: HSV—dendro-geographical keratitis

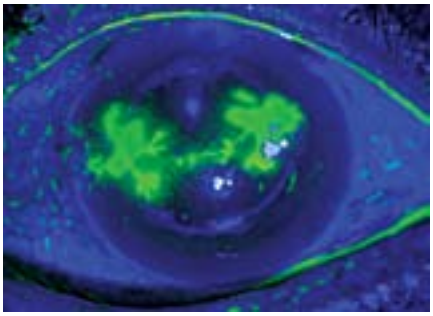


Fig. 3.9.3D: Dendro-geographical ulcer

Stromal Necrotic Keratitis

- May be associated with epithelial breakdown and anterior uveitis **(Figs 3.9.4A and B)**

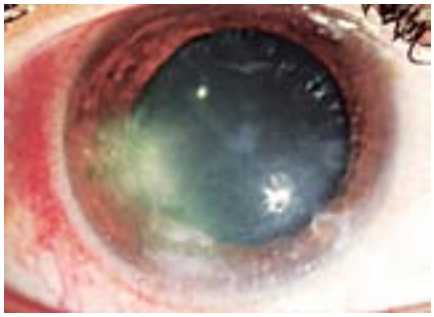


Fig. 3.9.4A: HSV—epithelial and stromal keratitis

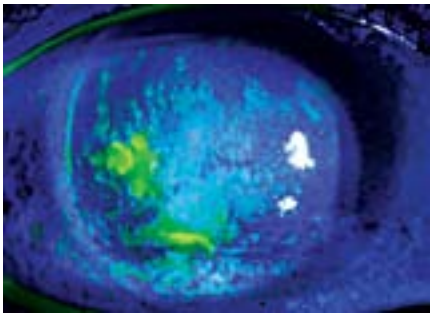


Fig. 3.9.4B: HSV—epithelial and stromal keratitis

- Cheesy and necrotic appearance of the stroma **(Figs 3.9.5A and B)**
- Vascularization, scarring and even perforation may occur
- *Treatment:* Oral acyclovir, topical antiviral, cycloplegic and judicious use of topical corticosteroids.

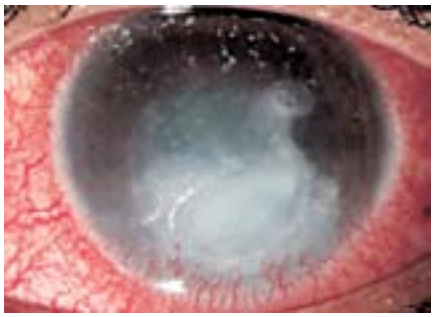


Fig. 3.9.5A: HSV—stromal necrotic keratitis

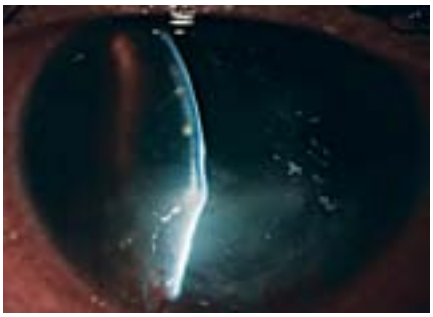


Fig. 3.9.5B: HSV—stromal necrotic keratitis

Metaherpetic Keratitis (Trophic Ulcer)

- Due to persistent defects in the basement membrane
- Margin is gray and thickened due to heaped-up epithelium **(Figs 3.9.6A and B)**. Not an active viral disease
- *Treatment:* Artificial tears and bandage contact lens (BCL).

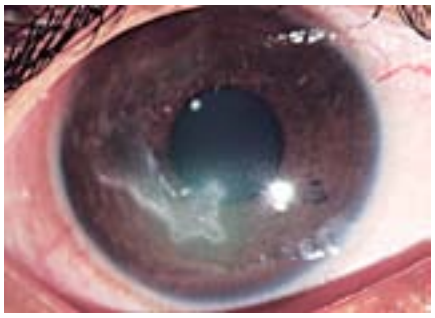


Fig. 3.9.6A: Metaherpetic keratitis

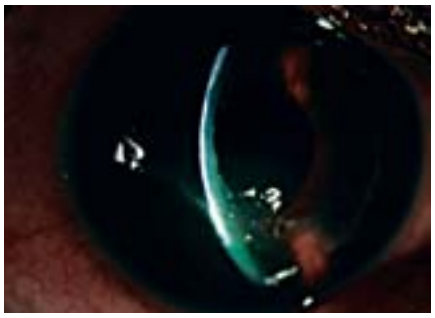


Fig. 3.9.6B: Metaherpetic keratitis

Disciform Keratitis (HSV Endotheliitis)

- Deep keratitis with disk-like edema—immunogenic reaction to HSV. Focal central stromal edema with fine KPs **(Figs 3.9.7A and B)**
- Presence of Descemet's folds and increased central corneal thickness in severe cases **(Figs 3.9.8A and B)**

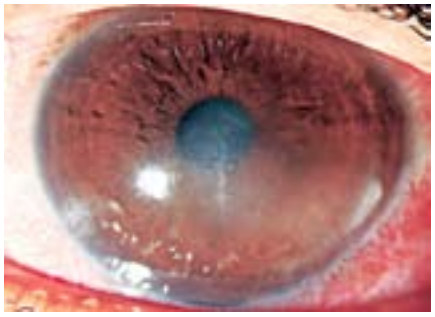


Fig. 3.9.7A: Disciform keratitis—endotheliitis

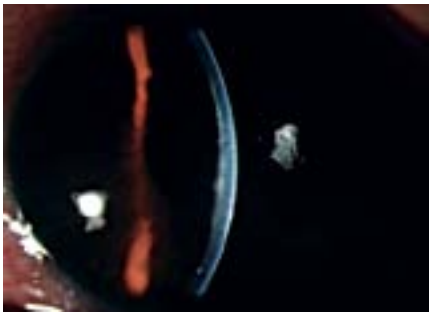


Fig. 3.9.7B: Disciform keratitis—endotheliitis

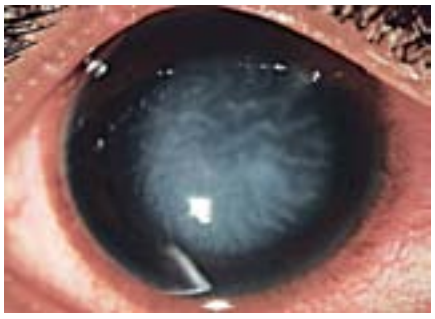


Fig. 3.9.8A: HSV—disciform keratitis

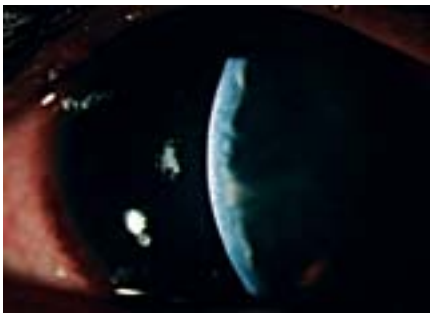


Fig. 3.9.8B: HSV—disciform keratitis—slit section

- *Wessley's immune ring* surrounding the edema in long standing cases **(Figs 3.9.9A and B)**
- *Treatment:* Topical corticosteroids (full strength or diluted) with acyclovir eye ointment in equal frequency for 2–4 weeks in tapering doses and cycloplegic. In severe cases, systemic steroids with oral antiviral coverage.

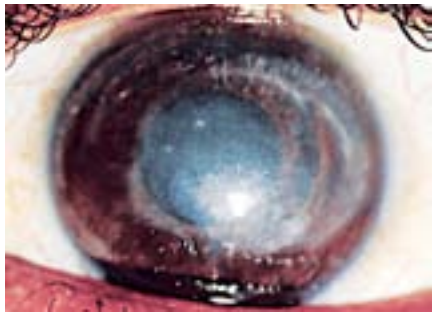


Fig. 3.9.9A: Disciform keratitis—Wessley's immune ring

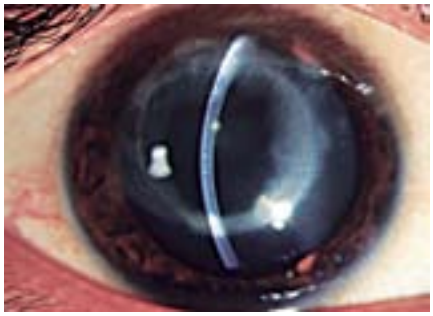


Fig. 3.9.9B: Wessley's immune ring

Herpes Zoster Ophthalmicus (HZO)

- Caused by vericella zoster virus affecting the elderly people
- More common in immunocompromised hosts
- Vesicular eruptions around the eye, forehead and scalp **(Fig. 3.10.1A)**
- Severe pain along the ophthalmic division of fifth cranial nerve
- *Hutchinson's rule/sign:* When the tip of the nose is involved, the eye will also be involved, since both are supplied by the nasociliary nerve **(Fig. 3.10.1B)**
- The ocular lesions may be *acute, chronic or recurrent.*

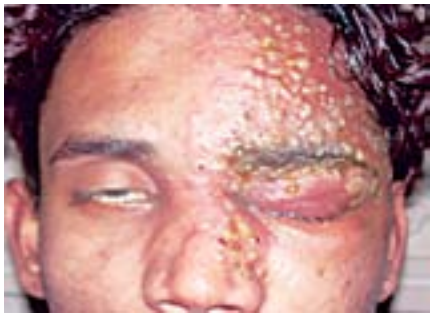


Fig. 3.10.1A: Herpes zoster ophthalmicus—fresh lesions

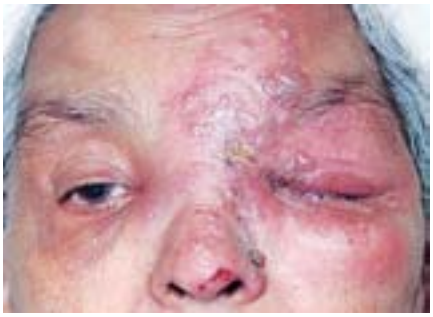


Fig. 3.10.1B: Herpes zoster ophthalmicus—Hutchinson's rule

Acute Ocular Lesions

Lids: Redness, edema and vesicular eruptions.

Cornea

- Punctuate epithelial keratitis
- *Microdendrites:* Small, fine, multiple dendritic or stellate lesions **(Fig. 3.10.2)**
- *Nummular keratitis:* Multiple granular lesions surrounded by a halo of stromal haze **(Fig. 3.10.3)**
- Sensation may be diminished
- Disciform keratitis
- *Iris:* Acute iridocyclitis with hyphema and patches of iris atrophy **(Fig. 3.10.4)**
- *Neuro-ophthalmological:* Optic neuritis and cranial nerve palsies—affecting the 3rd (most common), 4th and 6th nerves **(Fig. 3.10.5).**

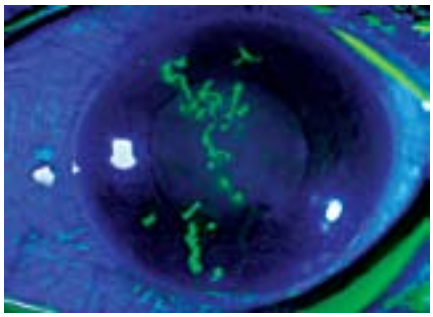


Fig. 3.10.2: HZV—microdendrites

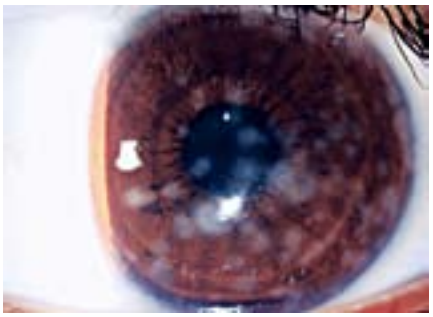


Fig. 3.10.3: Nummular keratitis

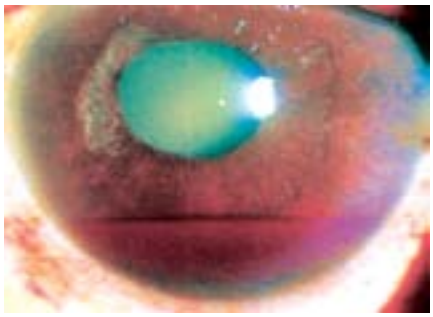


Fig. 3.10.4: HZO—uveitis with hyphema

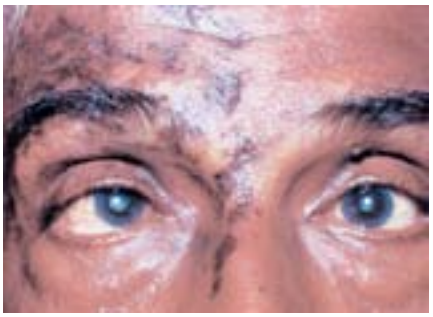


Fig. 3.10.5: HZO—lateral rectus palsy

Chronic Ocular Lesions

- Ptosis due to scarring of the lid
- Trichiasis, entropion and lid notching
- Scleritis, nummular keratitis, ocular surface instability
- *Recurrent lesions:* Mucus plaque keratitis, neurotrophic keratitis **(Fig. 3.10.6)** or secondary glaucoma
- *Treatment:* In acute cases, oral acyclovir (800 mg 5 times daily for 7 days), and topical steroids in presence of keratitis or iridocyclitis; systemic corticosteroids in neuro-ophthalmologic problems.

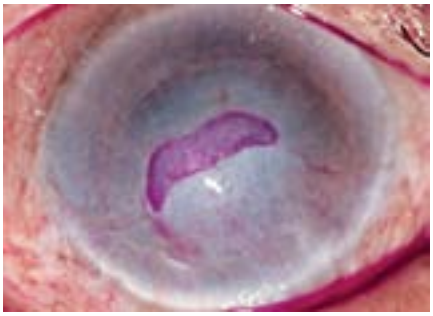


Fig. 3.10.6: HZO—neurotrophic keratitis

Other Infective Keratitis

Atypical Mycobacterial Keratitis

- Atypical or nontuberculous *Mycobacteria* (NTM) are known to cause infectious keratitis, most of them caused by two species: *M. fortuitum* and *M. chelonae.* Commonly, it occurs after either accidental or surgical trauma and more recently after LASIK and PRK **(Fig. 3.11.1)**
- Keratitis is usually characterized by indolent course (relatively slow rate of progress), and resistance to medical treatment. Clinical sign, which often develop within 2 to 8 weeks after LASIK reveals the formation of a sharply demarcated gray-white branching, round or needle-like opacities in the corneal interface. Ring infiltrates are common **(Fig. 3.11.2)** and may cause confusion with herpetic, fungal or *Acanthamoeba* keratitits. Intact overlying epithelium is sometimes observed
- *Treatment:* Intensive therapy with topical fourth generation fluoroquinolones. Topical amikacin 1.5% may also be effective. Oral clarithromycin—500 mg 2 times for 2 weeks simultaneously. Topical antibiotics are to be continued for months. A superficial keratectomy or flap amputation may be needed in post-LASIK cases.

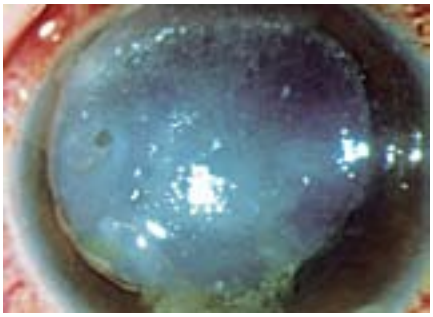


Fig. 3.11.1: Atypical mycobacteria keratitis—post-LASIK

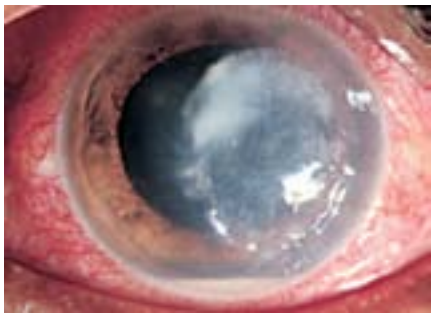


Fig. 3.11.2: Atypical mycobacteria keratitis

Nocardia Keratitis

- *Nocardia asteroids*, a Gram-positive, aerobic filamentous bacterium, affects the immunocompromised individuals, following minor trauma. It may be misdiagnosed clinically, as the picture may resemble mycotic keratitis **(Fig. 3.12.1)** or keratitis caused by atypical mycobacteria
- Multiple anterior stromal pin-head like infiltrations, typically arranged in the form of a ring, called "wreath" pattern **(Fig. 3.12.2)**. Patchy infiltrates which are predominantly anterior stromal with associated involvement of epithelium and subepithelial tissues, are pathognomonic of *Nocardia* keratitis
- *Treatment:* Fortified amikacin (2.0–2.5%) eyedrop is the treatment of choice. Gentamicin and 4th generation fluoroquinolones (moxifloxacin or gatifloxacin) are also effective.

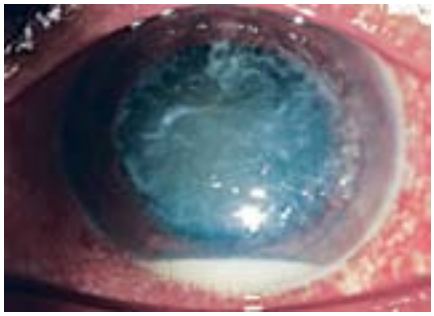


Fig. 3.12.1: Nocardia keratitis with hypopyon

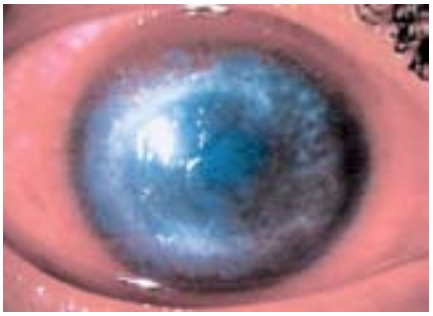


Fig. 3.12.2: Nocardia keratitis—typical wreath pattern

Microsporidial Keratitis

- Microsporidia are eukaryotic, spore forming obligate intracellular parasites, only two species—*Nosema* and *Encephalitozoon*, are known to cause ocular infections, mainly keratitis
- Common features were follicular papillary conjunctivitis and superficial coarse punctate epithelial lesions in three patterns—diffuse, peripheral, and paracentral—evolving into nummular keratitis before resolution **(Fig. 3.13.1)**
- The other type of presentation is—corneal stromal keratitis which begins insidiously and mimics a progressive herpetic disciform keratitis with corneal edema **(Figs 3.13.2A and B)**. There may be associated diffuse endotheliitis and limbitis. Diagnosis can be made on corneal biopsy
- *Treatment:* Topical propamidine isethionate (0.1%) six times daily, neosporin eye ointment—twice daily; oral albendazole 400 mg three times daily for a week, and then 400 mg daily for 4–6 weeks. Identifying microsporidia to species specific level could have important implications in the clinical management.

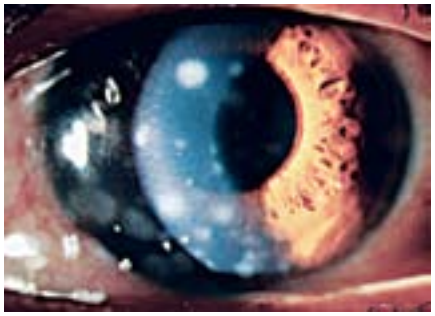


Fig. 3.13.1: Microsporidial keratitis—nummular appearance

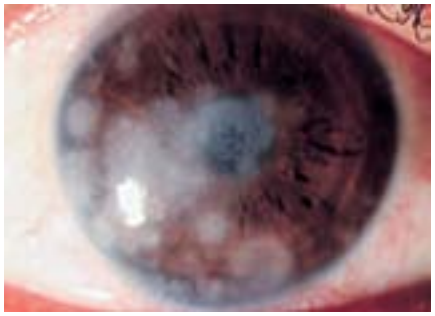


Fig. 3.13.2A: Microsporidial keratitis—nummular and disciform appearance

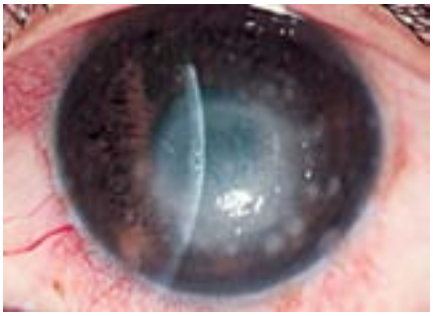


Fig. 3.13.2B: Microsporidial keratitis—disciform appearance

Infectious Crystalline Keratitis

- It is a rare indolent infection usually associated with long-term use of topical steroids as in PK. It is usually caused by *Streptococcus viridians*, though other organism may also be responsible **(Fig. 3.14.1)**
- It appears as slowly progressive grey-white branching opacities in the anterior or mid-stroma **(Figs 3.14.2A and B)**. There is minimum inflammation and overlying epithelium is intact
- *Treatment:* Intensive topical antibiotics (fluoroquinolones) for several weeks. Sudden withdrawal of steroids drops without topical antibiotic may aggravate this condition or even frank suppuration.

Fig. 3.14.1: Infectious crystalline keratitis

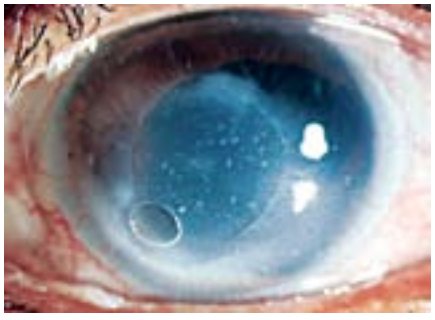


Fig. 3.14.2A: Infectious crystalline keratitis

Fig. 3.14.2B: Infectious crystalline keratitis

Keratitis (Other Types): Noninfectious and Peripheral

Lagophthalmic (Exposure) Keratitis

- Owing to dryness and desiccation, the lower third of epithelium is cast off and the raw area is invaded by microorganism. Typical lesions heals with scarring at the lower third of the cornea **(Figs 3.15.1A and B)**
- Seen in facial palsy **(Fig. 3.15.2)**, leprosy **(Figs 3.15.3A and B)**, proptosis, thyroid exophthalmos, comatose patient, etc.
- *Treatment:* Lid taping, tarsorrhaphy, lid-load operation and treatment of the cause.

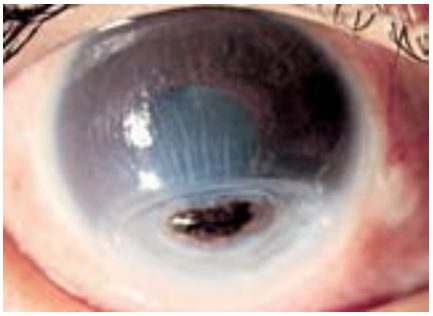


Fig. 3.15.1A: Lagophthalmic keratitis

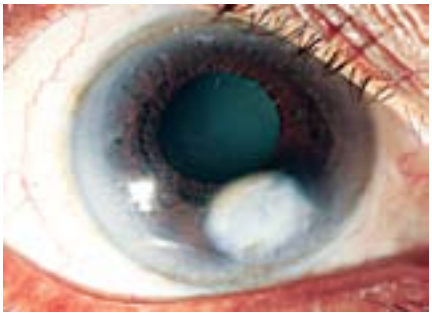


Fig. 3.15.1B: Lagophthalmic keratitis—healed

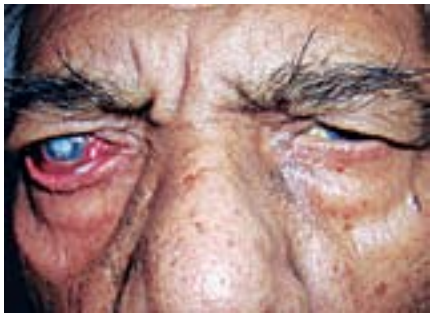


Fig. 3.15.2: Lagophthalmic keratitis

Fig. 3.15.3A: Lagophthalmic keratitis—leprosy

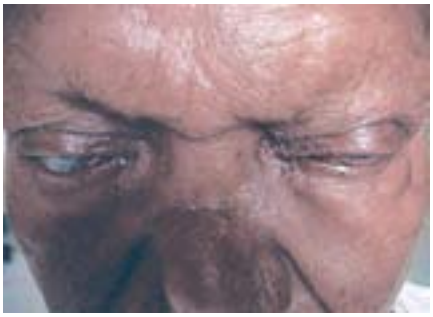


Fig. 3.15.3B: Lagophthalmic keratitis—leprosy

Neurotrophic Keratitis (NTK)

- Occurs in anesthetic cornea which alters the metabolism of the epithelium. Mostly seen in HSV and HZV keratitis. Punctate epithelial erosion involving the intrapalpebral area. Edema and exfoliation followed by epithelial ulceration **(Figs 3.16.1A and B)**
- *Treatment:* Ointment and patching, in severe cases amniotic membrane transplantation (AMT) or tarsorrhaphy.

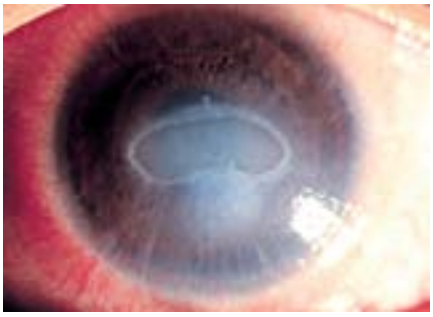


Fig. 3.16.1A: Neurotrophic keratitis

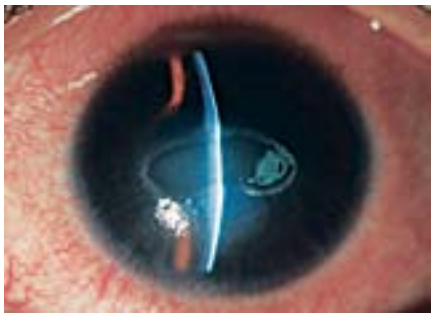


Fig. 3.16.1B: Neurotrophic keratitis

Nummular Keratitis

- Unilateral or bilateral subepithelial lesions mainly seen in vericella zoster virus infection
- Large, multiple, round or oval granular subepithelial deposits just beneath the Bowman's layer and surrounded by a halo of stromal haze **(Figs 3.17.1A and B)**
- *Treatment:* Topical dilute steroid drops in tapering doses and tear lubricants.

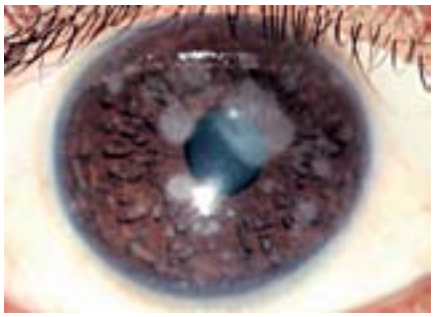


Fig. 3.17.1A: Nummular keratitis

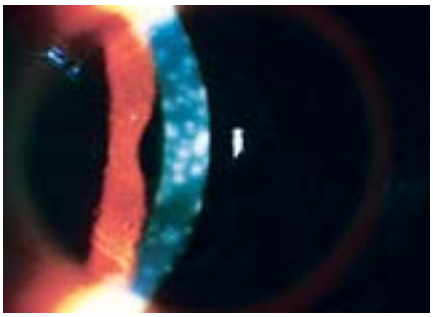


Fig. 3.17.1B: Nummular keratitis

Marginal Keratitis (Catarrhal Ulcer)

- Caused by hypersensitivity reaction to *Staphylococcal* exotoxin. Subepithelial infiltrates at the periphery, mostly at 4-8 o'clock position, or at 10-2 o'clock position **(Figs 3.18.1A and B)**
- *Treatment:* Topical corticosteroids, steroid-antibiotic ointment and simultaneous treatment of blepharitis.

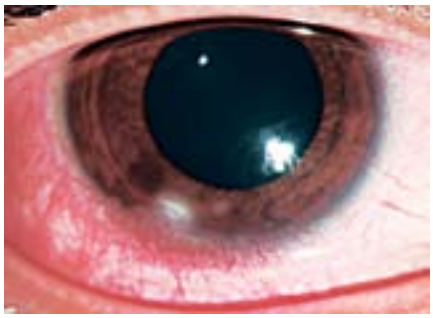


Fig. 3.18.1A: Marginal keratitis

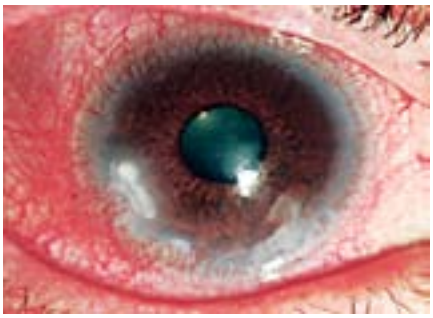


Fig. 3.18.1B: Marginal keratitis—multiple lesions

Marginal Ulcers with Systemic Collagen Diseases Or Peripheral Ulcerative Keratitis (PUK)

- Seen in rheumatoid arthritis, polyarteritis nodosa, SLE, or in Wegener's granulomatosis
- Ulceration and thinning **(Figs 3.19.1A and B)**; total peripheral keratolysis **(Fig. 3.19.2)** and gradual progression towards center **(Fig. 3.19.3)** and lastly appearance of *contact lens cornea* **(Fig. 3.19.4)**, etc.
- *Treatment:* Systemic and topical corticosteroids, tear substitutes, oral immunosuppressive agents (e.g. azathioprim), peritomy, glue and BCL, or peripheral tectonic (patch) graft.

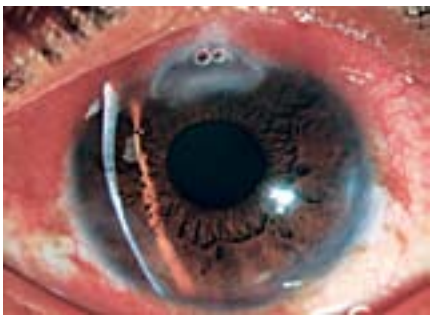


Fig. 3.19.1A: PUK—thinning and recurrent melts

Fig. 3.19.1B: PUK— extreme thinning

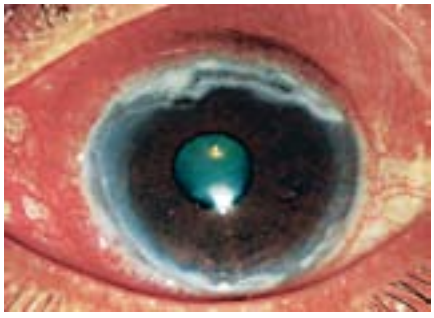


Fig. 3.19.2: PUK—peripheral melts

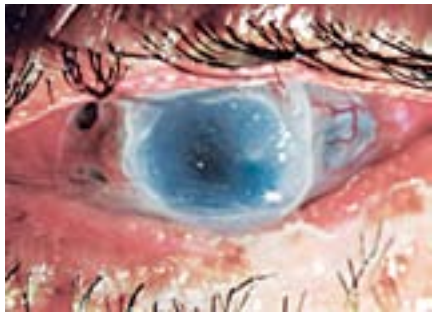


Fig. 3.19.3: PUK—progressive rheumatoid melts

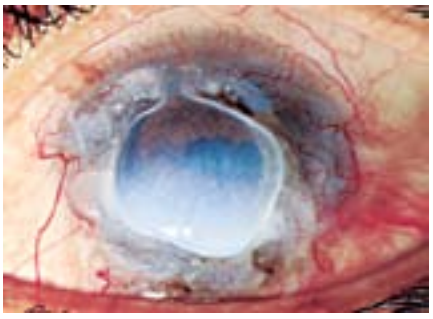


Fig. 3.19.4: Total peripheral keratolysis with contact lens cornea

Phlyctenular Keratitis

- Predominantly affects the children
- Corneal phlycten is a gray nodule, slightly raised above the surface, and a phlyctenular ulcer is yellow in color **(Fig. 3.20.1)**. It may resolves spontaneously or may extends towards the center of cornea
- ***Fascicular ulcer:*** Phlyctenular ulcer slowly migrates from the limbus towards the center of the cornea in a serpiginous way. It carries leash of blood vessels which lie in a shallow gutter formed by the ulcer
- Formation of corneal opacity which is densest at its apex **(Fig. 3.20.2)**
- *Treatment:* Topical corticosteroids, cycloplegic and topical antibiotic.

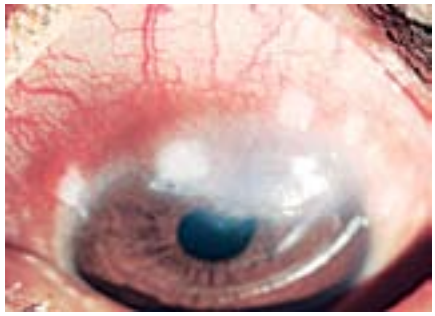


Fig. 3.20.1: Phlyctenular keratitis—multiple

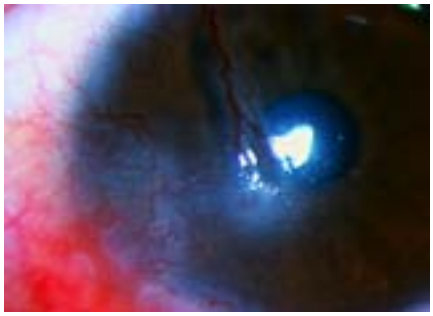


Fig. 3.20.2: Phlyctenular keratitis—fascicular ulcer

Interstitial Keratitis (IK)

- Inflammation of the corneal stroma without primary involvement of epithelium or endothelium. Rare bilateral condition, seen in syphilis, tuberculosis or Cogan's syndrome
- Vascularized, midstromal, non-suppurative inflammation, giving a ground glass appearance **(Figs 3.21.1A and B).** There may be intrastromal bleeding **(Fig. 3.21.2)**
- In inactive stage, there is variable stromal scarring with *ghost vessels* **(Fig. 3.21.3)**
- *Hutchinson's triad: IK, deafness and Hutchinson's teeth*—a part of congenital syphilis
- *Treatment:* Systemic penicillin, topical steroids, cycloplegic and AT drugs in selective cases.

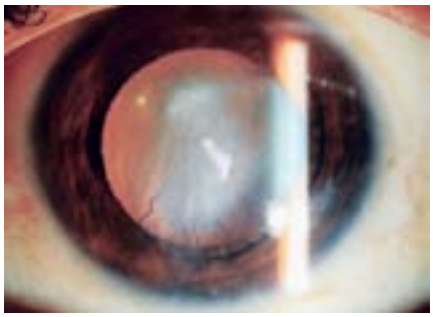


Fig. 3.21.1A: Interstitial keratitis

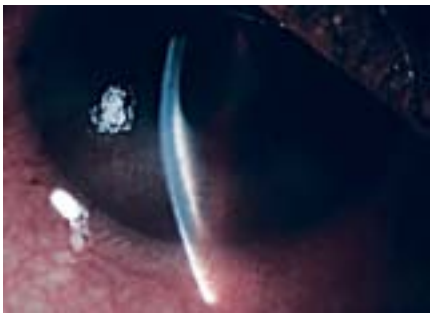


Fig. 3.21.1B: Interstitial keratitis—salmon patch

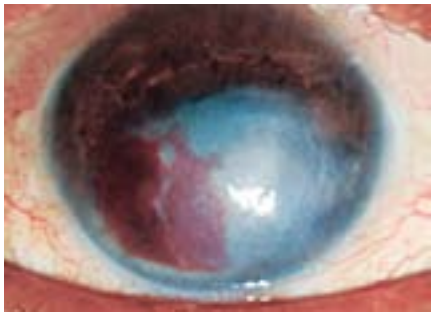


Fig. 3.21.2: Interstitial keratitis—intrastromal hemorrhage

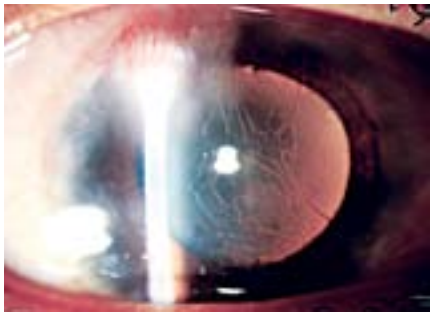


Fig. 3.21.3: Interstitial keratitis—ghost vessels

Punctate Epithelial Erosions (PEEs)

- Very common nonspecific corneal epithelial lesions seen in variety of corneal diseases
- The causes are: Keratoconjunctivitis sicca, meibomianitis, contact lens wearers, foreign body in subtarsal

sulcus, caterpillar hair, keratitis medicamentosa, photokeratitis, etc.

- Tiny, grayish-white, slightly depressed dots scattered in different fashion on the cornea **(Figs 3.22.1 and 3.22.2)**
- They represent the area of epithelial discontinuity and stain with fluorescein but not with rose bengal
- *Treatment:* Tear substitutes and others are directed towards the cause.

Punctate Epithelial Keratitis (PEK)

- Punctate epithelial lesions scattered all over the cornea
- Usually seen after acute follicular conjunctivitis of viral origin or after HZV infection
- The epithelial opacities appear as raised gray dots, scattered all over the cornea **(Figs 3.23.1A and B)**
- Sometimes, they extend into the Bowman's membrane and superficial stroma
- The lesions stain poorly with fluorescein, but turn bright red with rose-bengal
- *Treatment:* Tear substitutes and dilute topical steroids.

Superficial Punctate Keratitis (of Thygeson's)

- Uncommon, usually bilateral, idiopathic condition
- Round, oval or stellate conglomerations of grayish-white distinct dots which are intraepithelial, may be associated with mild subepithelial haze **(Figs 3.24.1A and B)**
- The conjunctiva is not involved
- *Treatment:* Tears substitutes and reassurance.

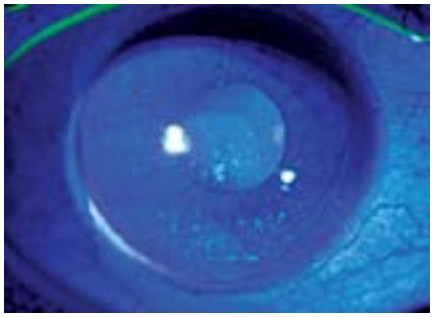


Fig. 3.22.1A: Punctate epithelial erosion (PEE)—CL wear

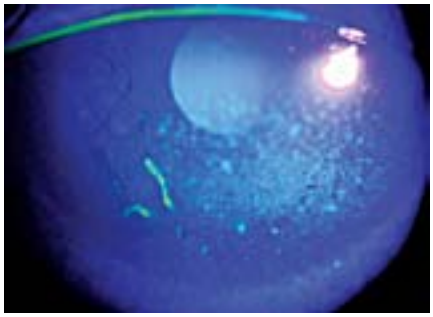


Fig. 3.22.1B: Punctate epithelial erosion —dry eye

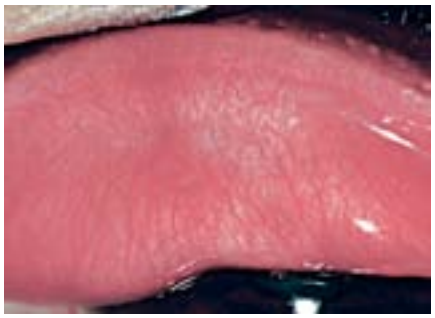


Fig. 3.22.2A: PEE—caterpillar hair

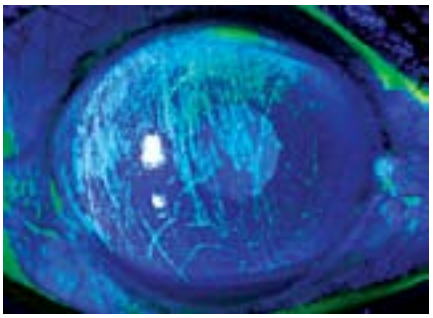


Fig. 3.22.2B: Linear PEE—caterpillar hairs

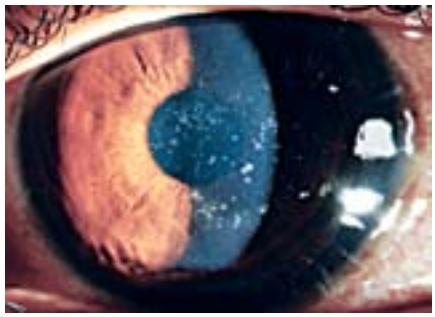


Fig. 3.23.1A: Punctate epithelial keratitis—adenoviral

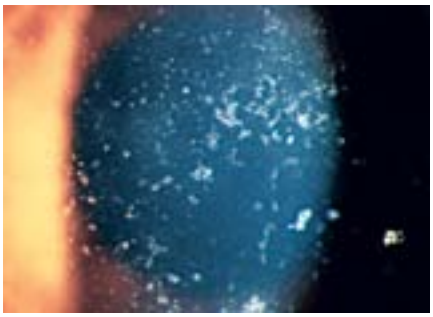
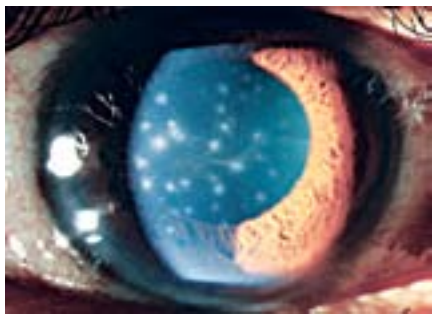


Fig. 3.23.1B: Punctate epithelial keratitis—adenoviral



3.24.1A: Superficial punctate keratitis (Thygeson's)—RE

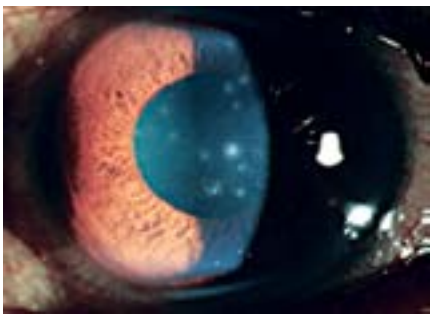


Fig. 3.24.1B: Superficial punctate keratitis (Thygeson's)—LE

Superior Limbic Keratoconjunctivitis (SLK)

- More in female and with thyroid dysfunction
- Bilateral chronic inflammation of the superior tarsal and bulbar conjunctiva
- Papillary hypertrophy of the superior tarsal conjunctiva
- Edema and thickening of the conjunctiva at the superior limbus **(*see* Figs 2.47.1 to 2.47.4)**
- Superior cornea shows punctate epithelial erosion and filaments which stains with fluorescein and rose-bengal **(Fig. 3.25.1)**
- *Treatment:* Artificial tears, bandage contact lens, block resection of superior bulbar conjunctiva or thermo-cauterization.

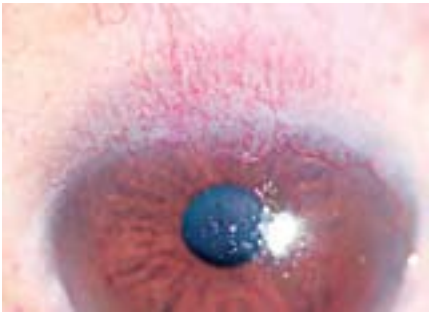


Fig. 3.25.1: Superior limbic kerato-conjunctivitis—rose bengal staining

Atheromatous Ulcer

- Develops over an old leucoma with degenerative changes
- The ulcer progresses rapidly with little tendency to heal **(Figs 3.26.1A and B)**
- Easily gets infected and perforation may occur
- *Treatment:* Ointment and patching, BCL and tears substitutes.

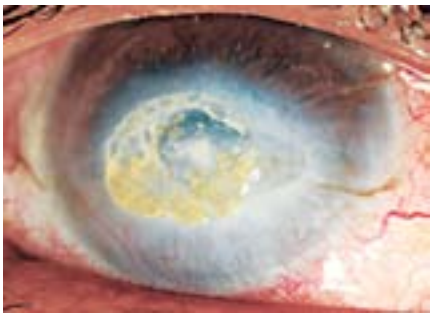


Fig. 3.26.1A: Atheromatous ulcer

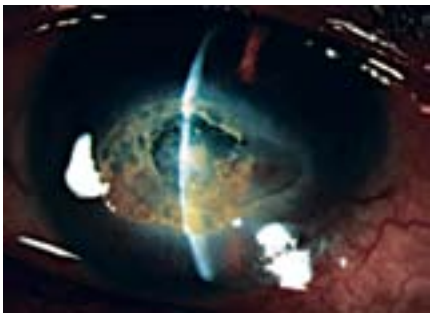


Fig. 3.26.1B: Atheromatous ulcer

Striate Keratopathy (Keratitis)

- Unilateral corneal edema with Descemet's folds; usually after a cataract surgery. It appears as delicate gray lines in deeper cornea **(Fig. 3.27.1)** Disappears spontaneously. Sometimes, they persist and end up with corneal decompensation
- *Treatment:* Like those of corneal edema.

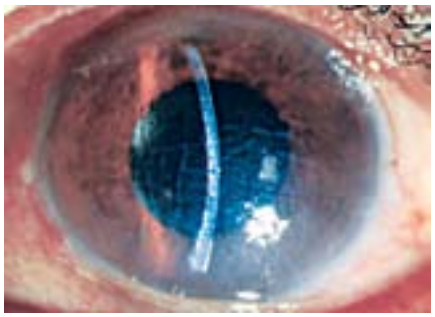


Fig. 3.27.1: Striate keratopathy

Filamentary Keratopathy (Keratitis)

- Mostly seen in severe dry eye. Formation of epithelial threads (filaments) on the cornea which adhere to the cornea by one end, while the other end moves freely **(Figs 3.28.1A and B)**. It is stained beautifully by fluorescein dye. May also be seen in other conditions **(Figs 3.28.2 and 3.28.3)**
- *Treatment:* Removal of filaments by scraping then BCL and frequent artificial tears.

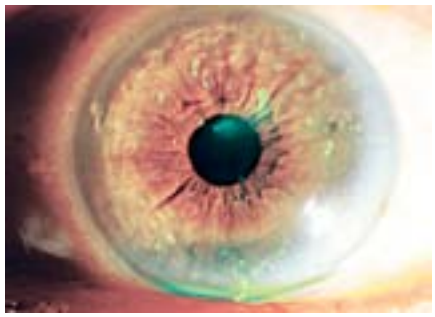


Fig. 3.28.1A: Filamentary keratitis

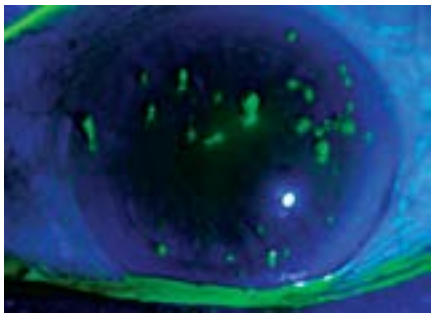


Fig. 3.28.1B: Filamentary keratitis

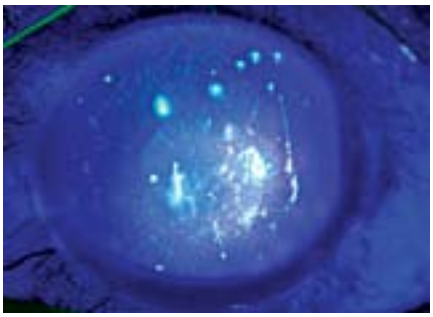


Fig. 3.28.2: Filamentary keratitis—microsporidiosis

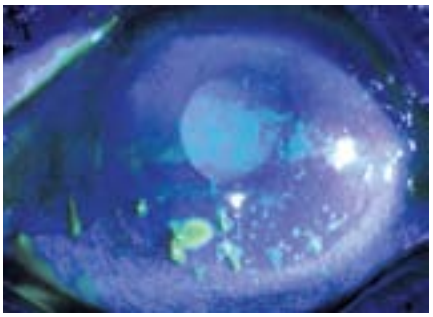


Fig. 3.28.3: Filamentary keratitis—tarsal foreign body

Shield Ulcer in Vernal Keratoconjunctivitis

- It occurs occasionally in patients with severe vernal catarrh
- Superior oval elevated lesion with grayish opacification of the bed
- Better to be designated as plaque **(Fig. 3.29.1)**
- Other signs of vernal conjunctivitis **(Fig. 3.29.2)**
- *Treatment:* Scraping or dissection of the plaque followed by BCL or amniotic membrane graft and supratarsal injection of triamcinolone acetonide.

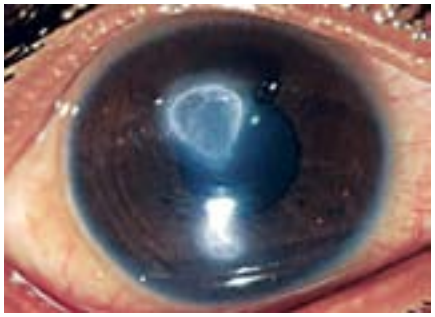


Fig. 3.29.1: Shield ulcer in VKC—raised plaque

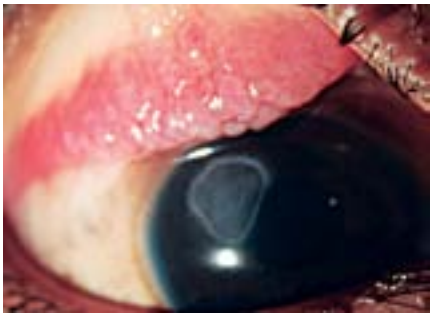


Fig. 3.29.2: Shield ulcer in VKC—tarsal papillae

Keratitis Medicamentosa

- Due to preservatives or drug itself. Initial presentation is superficial punctate keratitis, scattered all over the cornea **(Figs 3.30.1 to 3.30.3)**. Later, patient may present with dry eye with other signs
- *Treatment:* Withdrawal of specific drug and preservative-free artificial tears frequently.

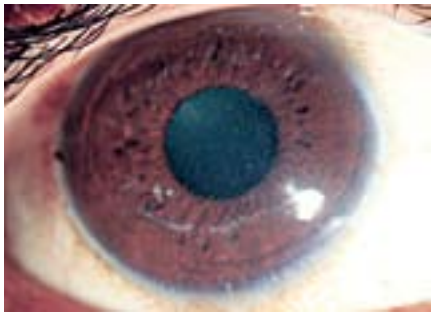


Fig. 3.30.1A: Keratitis medicamentosa

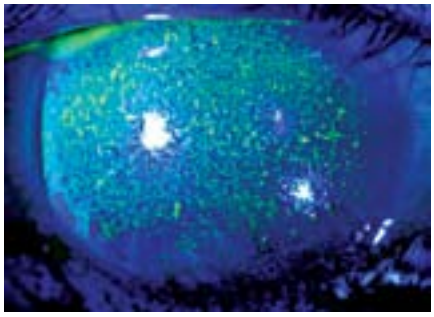


Fig. 3.30.1B: Keratitis medicamentosa

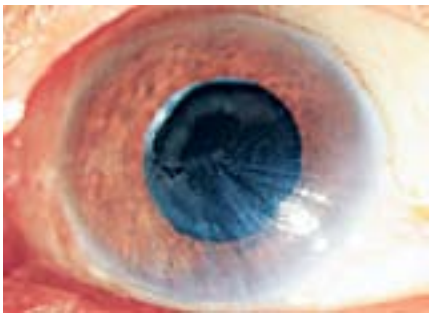


Fig. 3.30.2: Keratitis medicamentosa

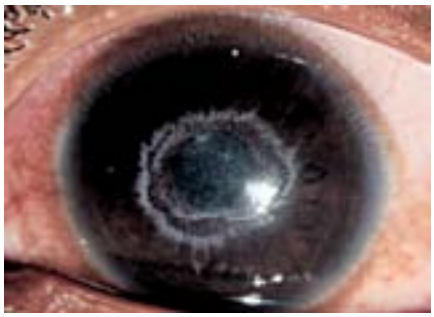


Fig. 3.30.3: Keratitis medicamentosa—ciprofloxacin deposits

Sclerosing Keratitis

- Rare condition may occur in isolation or with scleritis
- Gradual peripheral stromal thickening and opacification **(Figs 3.31.1 and 3.31.2)**
- Vascularization and lipid deposition may also occur
- *Treatment:* Systemic/topical steroids and systemic investigations.

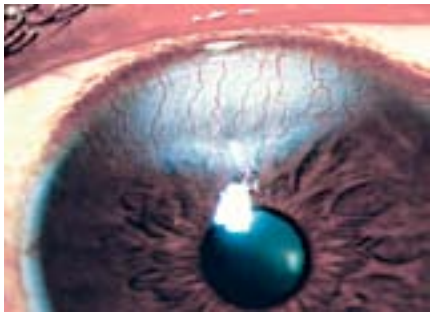


Fig. 3.31.1: Sclerosing keratitis

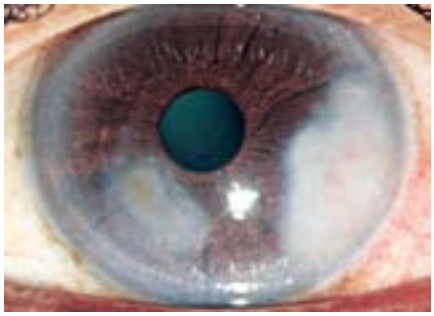


Fig. 3.31.2A: Sclerosing keratitis

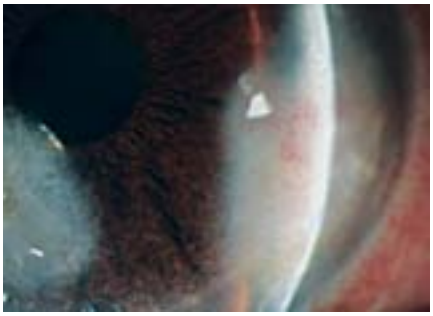


Fig. 3.31.2B: Sclerosing keratitis

Corneal Degenerations

Corneal Degeneration: Major Features

1. Usually unilateral and asymmetrical.
2. Located peripherally.
3. Accompanied by vascularization.
4. No inheritance pattern.
5. Onset in middle life or later.
6. Secondary to some compromising factors, e.g. aging, inflammation, chemicals, trauma or systemic diseases.

Arcus Senilis (Gerontoxon)

- Bilateral lipid degeneration of the peripheral cornea, affects most elderly persons
- Starts in the superior and inferior perilimbal cornea **(Fig. 3.32.1)**
- Then progresses circumferentially to form a white band 1 mm or more wide **(Figs 3.32.2A and B)**
- Peripheral sharp edge is separated from the limbus by a clear zone of cornea which may become thin to form senile furrow—called, peripheral furrow degeneration **(Fig. 3.32.3)**

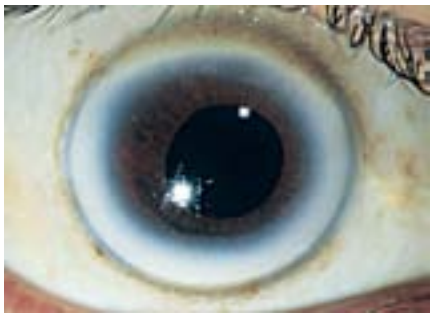


Fig. 3.32.1: Arcus senilis

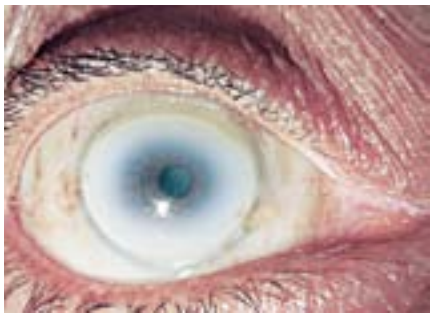


Fig. 3.32.2A: Dense arcus senilis—RE

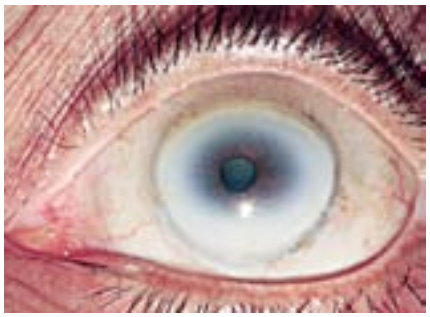


Fig. 3.32.2B: Dense arcus senilis—LE

- When it occurs below the age of 40 years, it is called ***arcus juvenilis*** (anterior embryotoxon), which may be associated with systemic hyperlipidemia **(Fig. 3.32.4)**
- ***Pseudogerontoxon (see Fig. 2.18.7):*** Most frequently seen in vernal conjunctivitis and takes cupid's bow configuration
- *Treatment:* No treatment is required.

Terrien's Marginal Degeneration

- Lesion starts as fine yellow-white punctate stromal opacities, usually at the upper part of the cornea. Sharp edge towards the center becomes demarcated by yellow-white lipid deposits **(Figs 3.33.1A and B)**. Eventually, thinning to form a peripheral gutter **(Figs 3.33.2A and B)**. Overlying epithelium is intact and vascularization is prominent. Rarely it may be inferior **(Fig. 3.33.3)**. There may be pseudopterygium formation **(Fig. 3.33.4)**
- *Treatment:* RGP contact lens, deep lamellar sectorial keratoplasty in severe cases or peripheral PK in perforation.

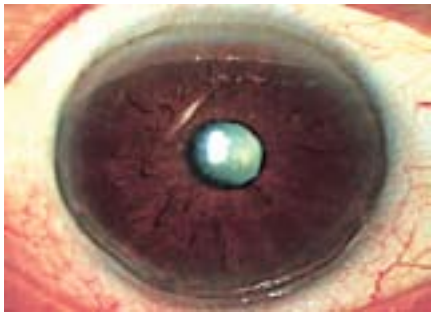


Fig. 3.32.3: Furrow degeneration

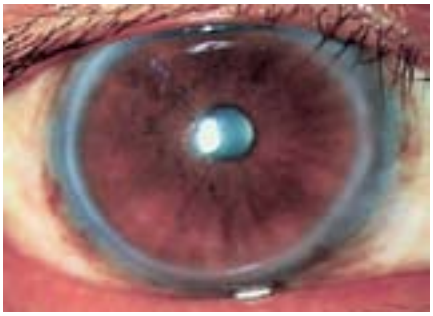


Fig. 3.32.4: Arcus juvenilis

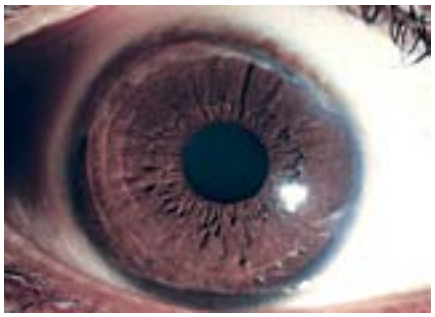


Fig. 3.33.1A: Terrien's marginal degeneration

Fig. 3.33.1B: Terrien's marginal degeneration

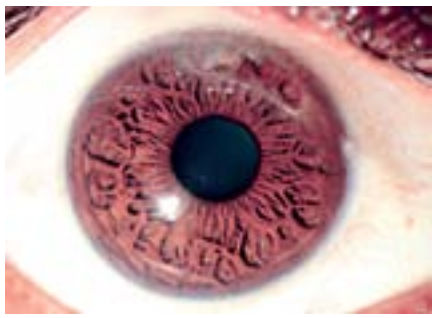


Fig. 3.33.2A: Terrien's marginal degeneration—thinning

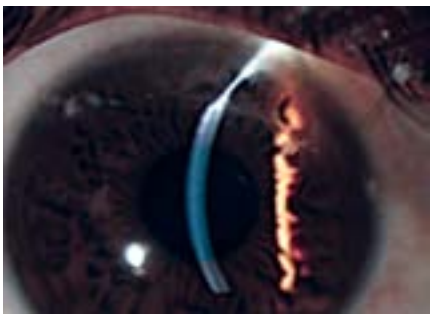


Fig. 3.33.2B: Terrien's marginal degeneration—marked thinning

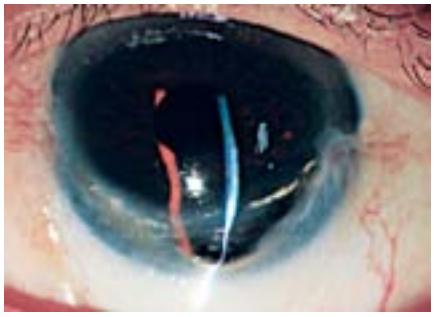


Fig. 3.33.3: Terrien's degeneration—inferior—Descemetocele with pseudopterygium

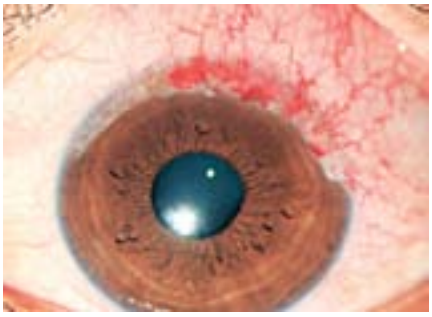


Fig. 3.33.4: Terrien's marginal degeneration—pseudopterygium

Pellucid Marginal Degeneration

- Thinning involves only the inferior cornea (between 4 o'clock to 8 o'clock) with ectasia just above the area of thinning, giving rise the appearance of keratoconus **(Fig. 3.34.1)**. No vascularization (as in Mooren's or Terrien's), or no lipid deposition as in Terrien's. Hydrops may occur as keratoconus **(Figs 3.34.2A and B)**. Rarely there may be associated keratoconus **(Figs 3.34.3A and B)**
- *Treatment:* Correction of astigmatism by RGP contact lens, tectonic patch graft.

Fig. 3.34.1: Pellucid marginal degeneration (PMD)

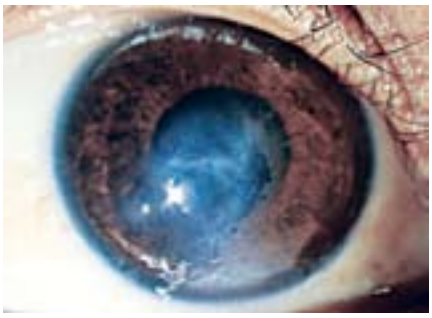


Fig. 3.34.2A: PMD—hydrops

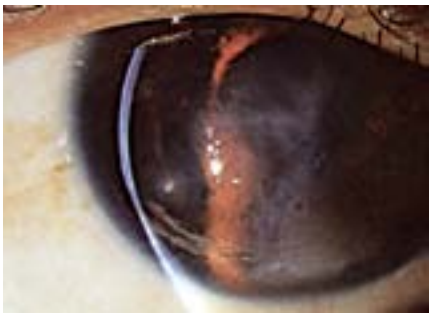


Fig. 3.34.2B: PMD—hydrops in slit section

Fig. 3.34.3A: PMD with keratoconus

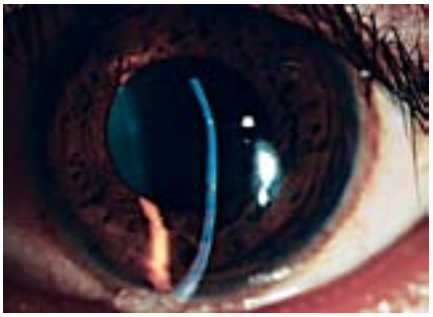


Fig. 3.34.3B: PMD with keratoconus

Mooren's Ulcer

- Chronic peripheral ulcer of unknown etiology due to ischemic necrosis from vasculitis of perilimbal vessels **(Figs 3.35.1A and B)**
- Peripheral ulcer with overhanging edge. Later involves the entire circumference and also spread towards the center **(Fig. 3.35.2)**. May be secondarily infected with hypopyon formation **(Fig. 3.35.3)**. Perforation may occur with minor trauma **(Fig. 3.35.4)**

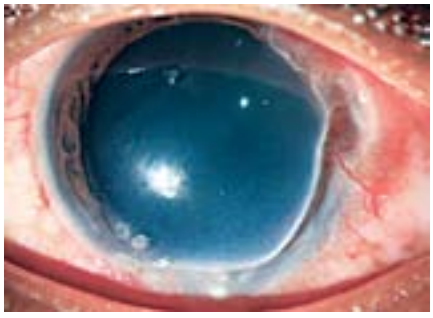


Fig. 3.35.1A: Mooren's ulcer

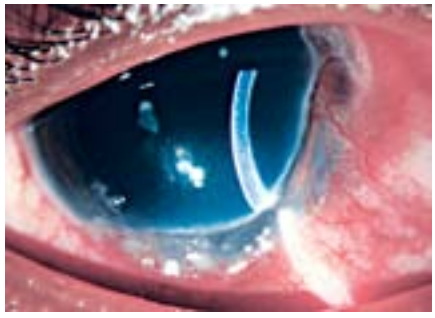


Fig. 3.35.1B: Mooren's ulcer—overhanging edge

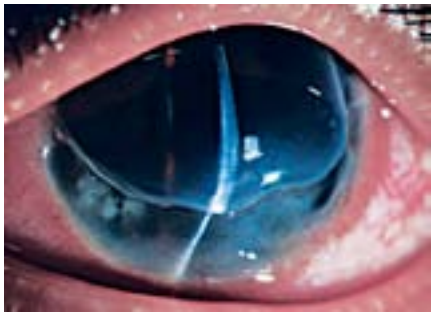


Fig. 3.35.2: Mooren's ulcer—extensive

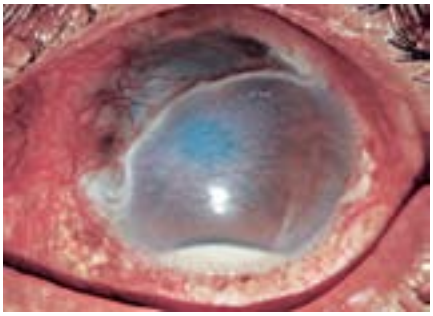


Fig. 3.35.3: Mooren's ulcer—secondary infection

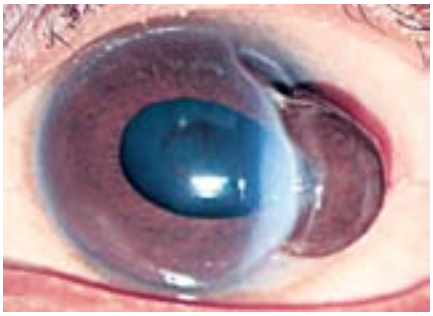


Fig. 3.35.4: Mooren's ulcer—perforation

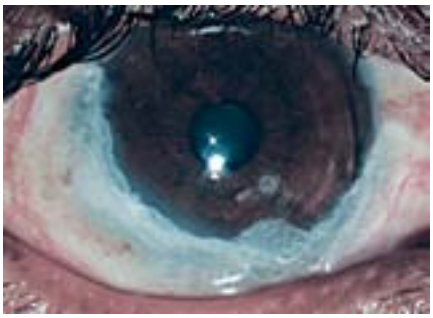


Fig. 3.35.5A: Mooren's ulcer—peritomy with glue and BCL

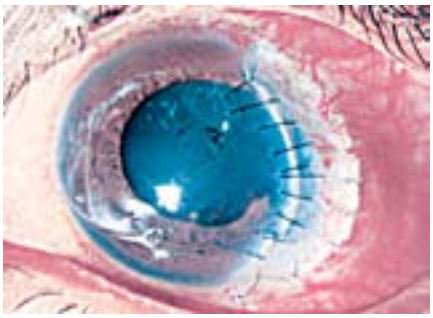


Fig. 3.35.5B: Mooren's ulcer—tectonic patch graft of **Figure 3.35.4**

- *Treatment:* Peritomy, Glue with BCL **(Fig. 3.35.5A)**, systemic immunosuppressants, lamellar banana-shaped tectonic graft **(Fig. 3.35.5B)**, etc.

Band-shaped Keratopathy (BSK)

- Calcific horizontal band, largely at the palpebral fissure, separated from limbus by a clear zone. Begins at periphery between 3 and 9 o'clock position, and then affects the central area **(Fig. 3.36.1)**. May be primary **(Fig. 3.36.2)** or secondary **(Fig. 3.36.3)** and associated with idiopathic juvenile arthritis, chronic uveitis **(Fig. 3.36.4)**, following VR surgery **(Fig. 3.36.5),** etc.
- *Treatment:* Scraping of the epithelium, treatment with chelating agent like, di-sodium EDTA, and lamellar keratoplasty in severe cases.

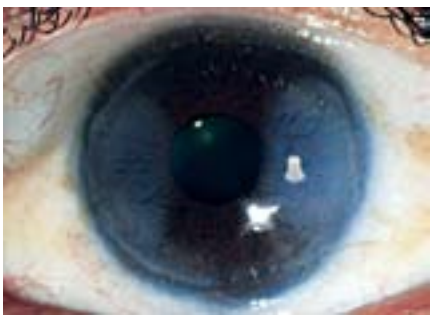


Fig. 3.36.1: Band-shaped keratopathy—primary

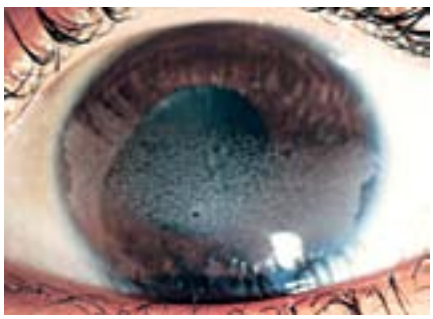


Fig. 3.36.2: Band-shaped keratopathy—primary

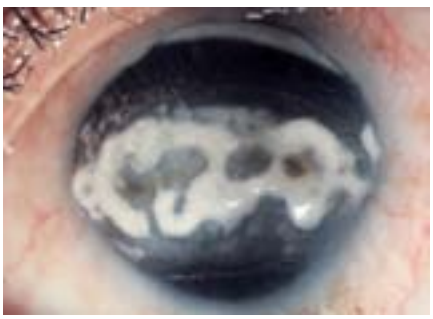


Fig. 3.36.3: Band-shaped keratopathy—lipid degeneration

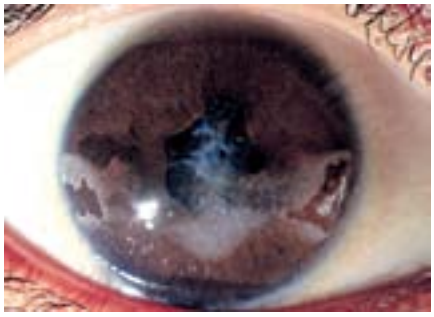


Fig. 3.36.4: Band-shaped keratopathy—idiopathic juvenile arthritis (JRA)

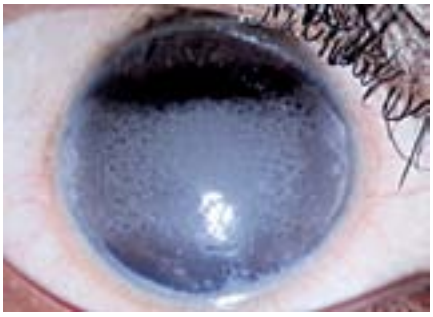


Fig. 3.36.5: Band-shaped keratopathy—following VR surgery

Salzmann's Nodular Degeneration

- Uncommon, and mostly unilateral. Elevated subepithelial bluish-grey nodules in a scarred cornea and surrounding cornea is clear **(Figs 3.37.1 and 3.37.2)**. Some cases are bilateral and may be with severe and extensive lesions **(Figs 3.37.3A and B)**.
- *Treatment:* Not necessary except regular use of tears substitute; phototherapeutic keratectomy (PTK) or epithelial debridement in case of central lesion. Anterior lamellar keratoplasty in severe and recurrent cases.

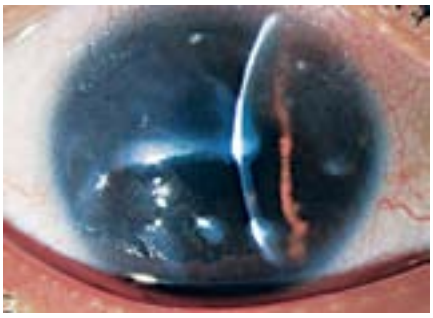


Fig. 3.37.1: Salzmann's nodular degeneration

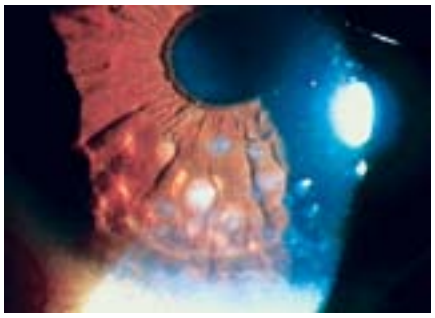


Fig. 3.37.2: Salzmann's nodular degeneration

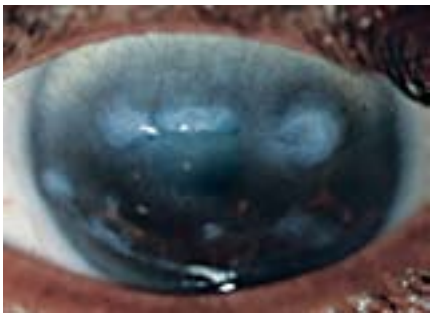


Fig. 3.37.3A: Salzmann's nodular degeneration—RE

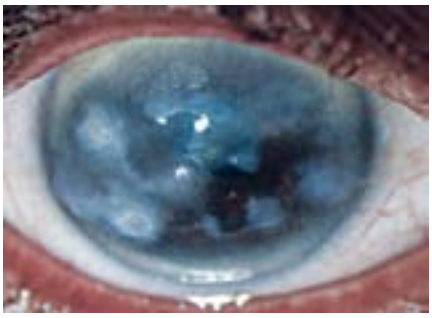


Fig. 3.37.3B: Salzmann's nodular degeneration—LE

Spheroidal Degeneration (Climatic Droplet Keratopathy)

- Rare, bilateral, small amber colored granules in the superficial stroma and conjunctiva, mainly in the interpalpebral area. Lesions then spread centrally and coalesce to become denser. It may be primary **(Figs 3.38.1A and B)** or secondary to other corneal diseases, like in BSK, old corneal opacity, lattice dystrophy, etc. **(Figs 3.38.2 to 3.38.4)**
- *Treatment:* Lamellar or penetrating keratoplasty, especially in secondary cases.

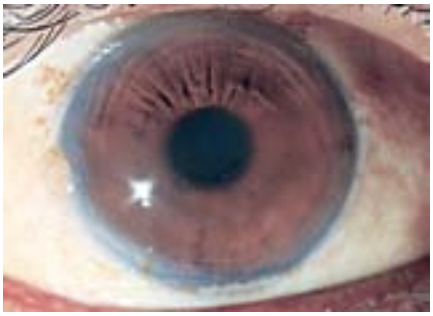


Fig. 3.38.1A: Spheroidal degeneration

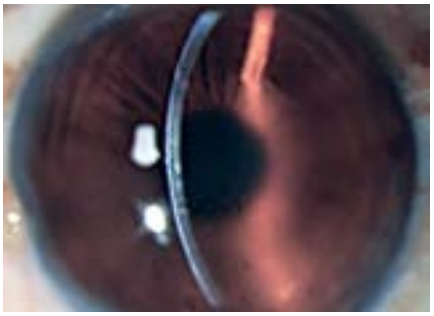


Fig. 3.38.1B: Spheroidal degeneration—slit section

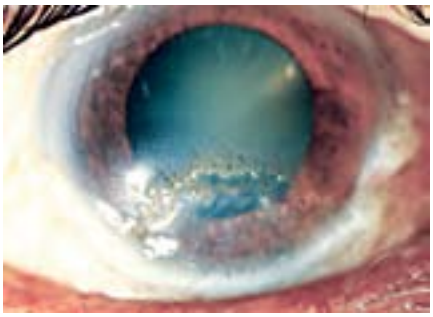


Fig. 3.38.2: Spheroidal degeneration—old corneal opacity

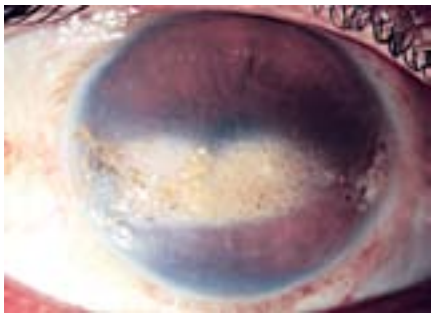


Fig. 3.38.3: Spheroidal degeneration—BSK

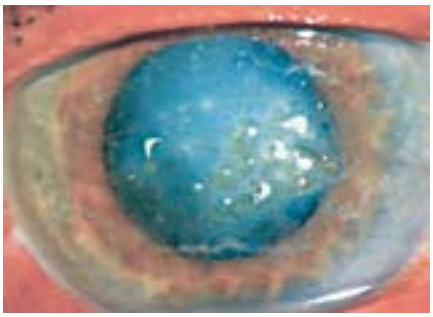


Fig. 3.38.4: Spheroidal degeneration—lattice dystrophy

Lipid Degeneration (Keratopathy)

Two types: Primary and secondary.

- *Primary:* It is very rare and occurs spontaneously. It is characterized by yellowish-white deposit within corneal stroma without any vascularization **(Fig. 3.39.1)**.
- *Secondary:* It is much more common and is associated with previous corneal disease or injury. Always associated with vascularization **(Figs 3.39.2 and 3.39.3)** and sometimes with intracorneal hemorrhage **(Fig. 3.39.4)**. Most commonly seen after herpetic (both HSV and HZO) keratitis.
- *Treatment:* Medical control of primary disease. Argon laser photocoagulation or needle cautery of blood vessels may be successful to reduce lipid deposition. Subconjunctival injection of Avastin (to the offending blood vessels) may reduce the lipid deposition. PK may be required in advanced cases.

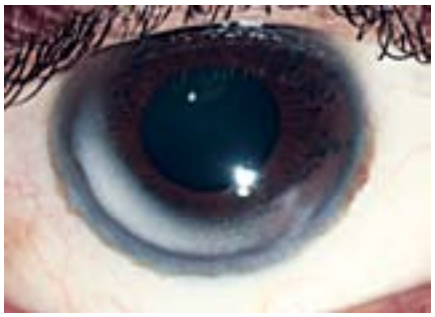


Fig. 3.39.1: Lipid keratopathy—primary

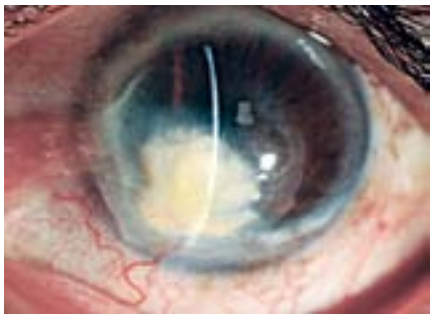


Fig. 3.39.2: Lipid keratopathy—secondary

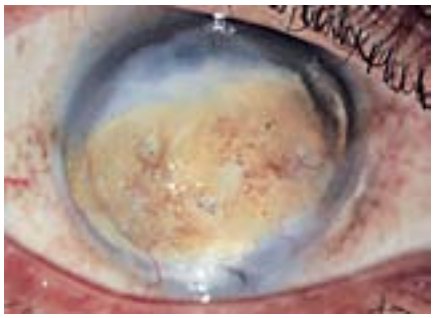


Fig. 3.39.3: Lipid keratopathy—secondary

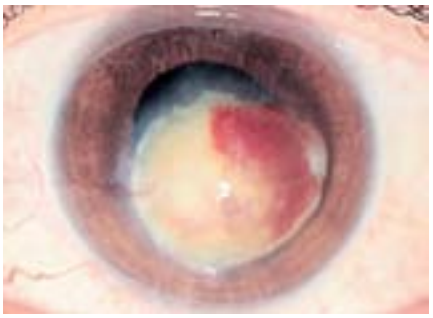


Fig. 3.39.4: Lipid keratopathy—intracorneal hemorrhage

White Limbal Girdle of Vogt

- Very common, bilateral, innocuous age-related condition
- Chalky white, crescentic linear opacities along the nasal and temporal limbus found at the interpalpebral area **(Figs 3.40.1 and 3.40.2)**
- *No treatment is required.*

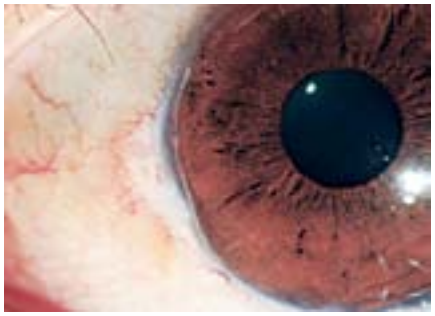


Fig. 3.40.1: Limbal girdle of Vogt

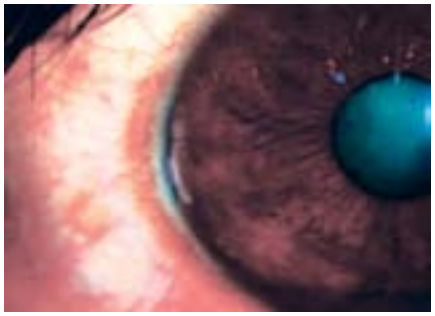


Fig. 3.40.2: White limbal girdle of Vogt

Corneal Dystrophies

Corneal Dystrophy: Major Features

1. Usually bilateral and symmetrical
2. Located centrally
3. No vascularization
4. Hereditary (usually autosomal dominant)
5. Early in onset
6. Unrelated to any systemic or local disease, or condition.

Anterior Dystrophies (Epithelium and Bowman's Membrane)

- Probably, the more common dystrophies, but frequently misdiagnosed due to variable presentation
- Most of the patients remain asymptomatic, but the others develop recurrent corneal erosions
- All are autosomal dominant inheritance.

The Common Varieties are:

- Map-dot-fingerprint dystrophy (Cogan's microcystic)
- Variety of microcysts, dots, fingerprint or map-like epithelial lesions **(Figs 3.41.1A and B)**

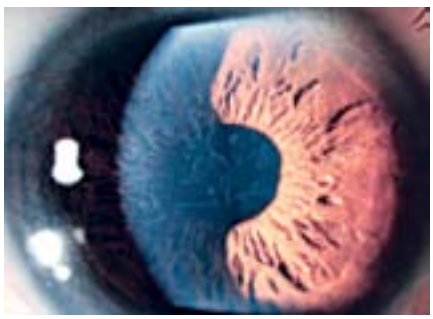


Fig. 3.41.1A: EBMD—Map-dot-fingerprint dystrophy

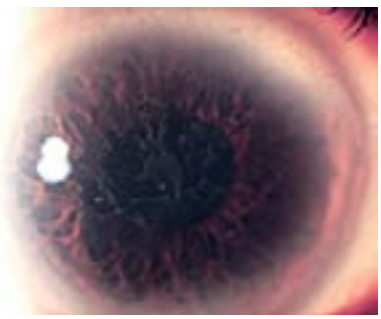


Fig. 3.41.1B: EBMD—Map-dot-fingerprint dystrophy in sclerotic scatter

- In some patients epithelial hypertrophy is the prominent feature **(Figs 3.41.2A and B)**
- May occur singly or in combination and best appreciated in oblique illumination or sclerotic scatter
- Patient may present with signs of bilateral recurrent corneal erosions **(Fig. 3.41.3)**
- *Treatment:* Epithelial debridement and BCL: PTK is also helpful in recurrent lesions.

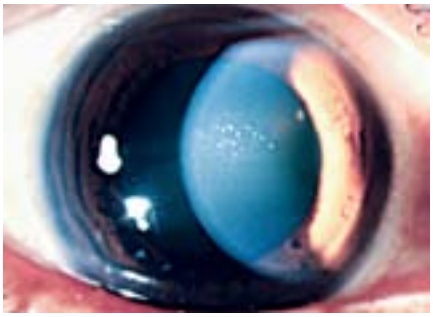


Fig. 3.41.2A: EBMD—epithelial hypertrophy

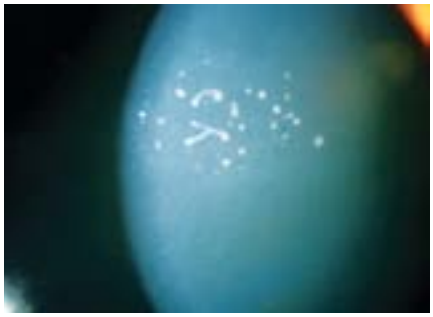


Fig. 3.41.2B: EBMD—epithelial hypertrophy

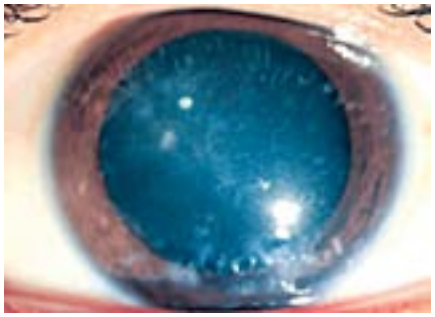


Fig. 3.41.3: EBMD—Map-dot-fingerprint dystrophy—with erosion

Meesmann's Dystrophy

- Very rare and innocuous condition
- Tiny epithelial cysts all over the cornea and more numerous in interpalpebral areas **(Figs 3.42.1A and B)**
- They are best visible by retroillumination or by sclerotic scatter, otherwise they appear gray **(Fig. 3.42.2)**
- *No treatment is necessary.*

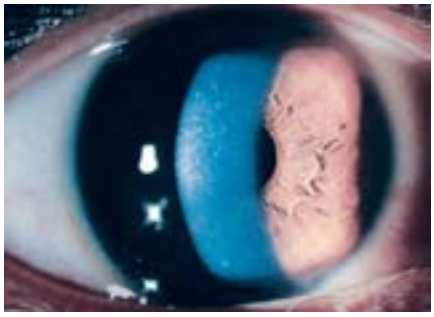


Fig. 3.42.1A: Meesmann's dystrophy

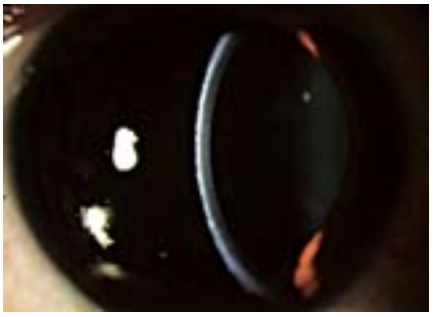


Fig. 3.42.1B: Meesmann's dystrophy in slit section

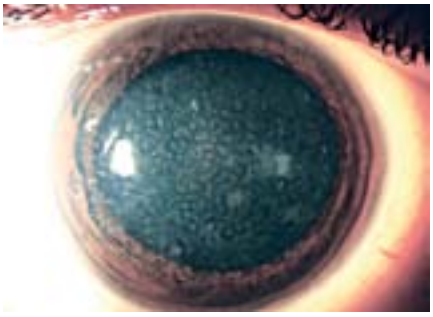


Fig. 3.42.2: Meesmann's dystrophy in sclerotic scatter

Reis-Buckler's Dystrophy

- Relatively common, bilateral condition with autosomal dominant inheritance
- Reduced visual acuity in second or third decade
- Ring-shaped subepithelial opacities giving 'honeycomb' appearance **(Figs 3.43.1A and B)**

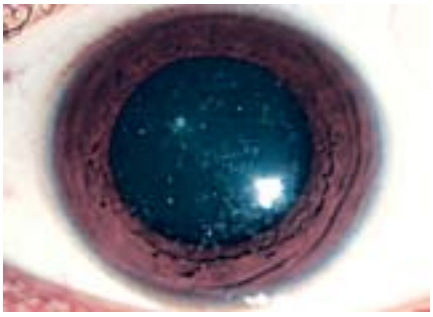


Fig. 3.43.1A: Reis-Buckler's dystrophy—early

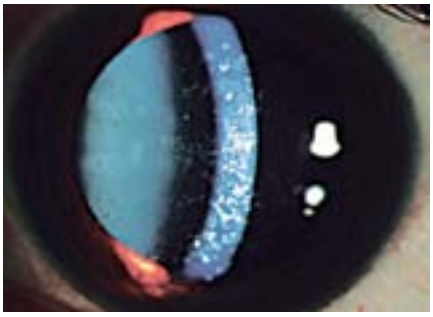


Fig. 3.43.1B: Reis-Buckler's dystrophy—early in slit section

- Gradually it increases **(Figs 3.43.2A and B)**
- Ultimately, the entire cornea is affected with more involvement of the central area **(Figs 3.43.3A and B)**
- *Treatment:* Tear lubricants, lamellar or deep anterior lamellar keratoplasty (DALK) in selective cases.

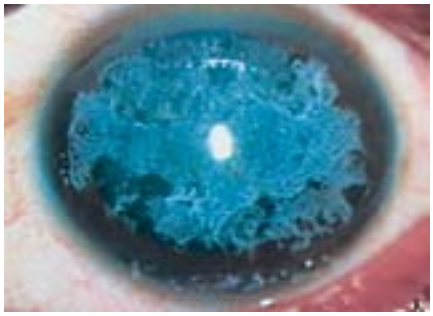


Fig. 3.43.2A: Reis-Buckler's dystrophy—severe

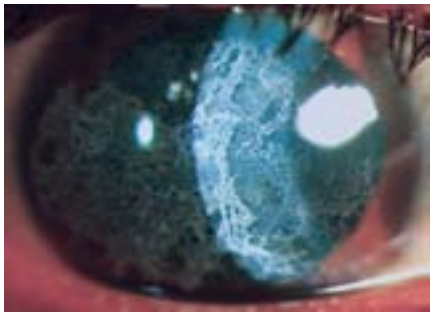


Fig. 3.43.2B: Reis-Buckler's dystrophy—severe

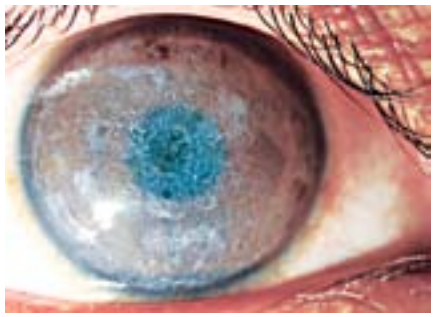


Fig. 3.43.3A: Reis-Buckler's dystrophy—advanced—RE

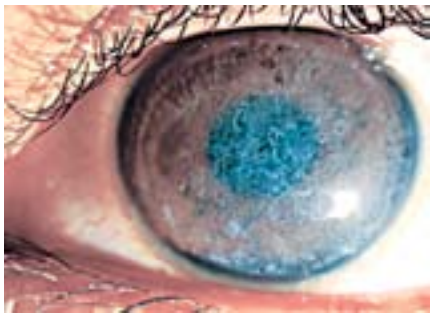


Fig. 3.43.3B: Reis-Buckler's dystrophy—advanced—LE

Stromal Dystrophies

Granular Dystrophy

- Relatively common, autosomal dominant, bilateral disease
- Starts around puberty as small multiple discrete white dots in the stroma **(Figs 3.44.1A and B)** and progress slowly
- The lesions appear as discrete, crumb-like white granules within the anterior stroma of the central cornea **(Fig. 3.44.2)**; some lesions may be linear **(Figs 3.44.3A and B)**; or stellate-shaped **(Figs 3.44.4A and B)**

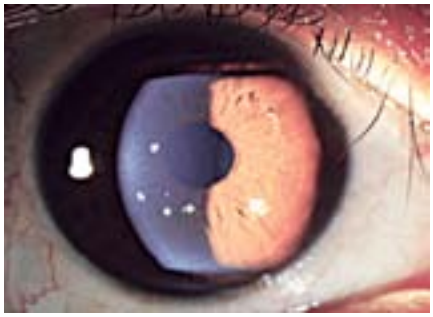


Fig. 3.44.1A: Granular dystrophy—early—RE

Fig. 3.44.1B: Granular dystrophy—early—LE

Fig. 3.44.2: Granular dystrophy—discrete lesions

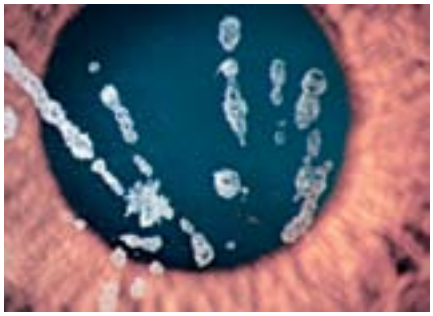


Fig. 3.44.3A: Granular dystrophy—linear form

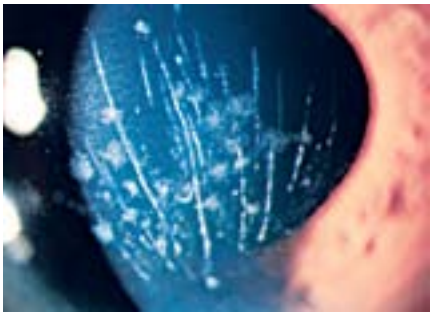


Fig. 3.44.3B: Granular dystrophy—linear pattern

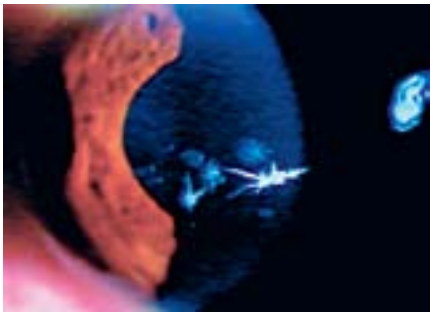


Fig. 3.44.4A: Granular dystrophy—stellate shaped

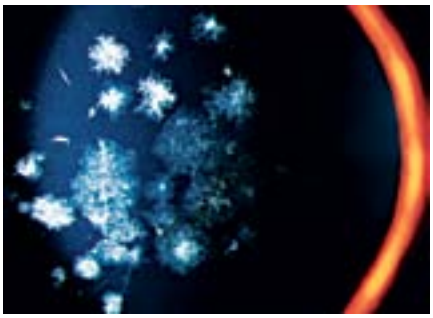


Fig. 3.44.4B: Granular dystrophy—star-shaped

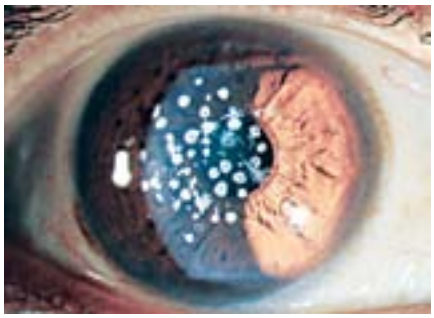


Fig. 3.44.5A: Granular dystrophy—discrete lesion in late stage

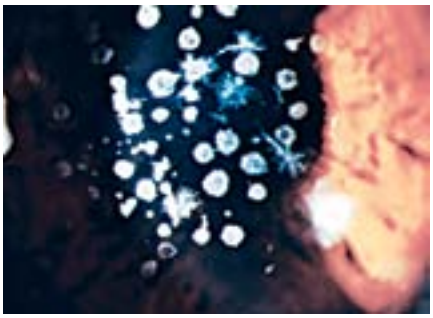


Fig. 3.44.5B: Granular dystrophy—round, stellate and linear

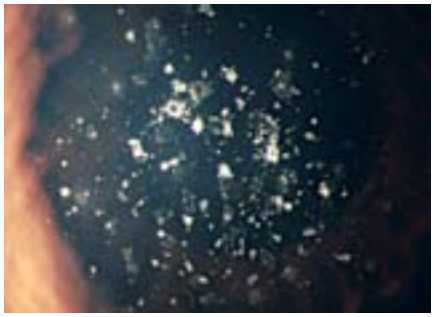


Fig. 3.44.6A: Granular dystrophy—clear space in between opacity

Fig. 3.44.6B: Granular dystrophy—clear space in between opacity in slit section

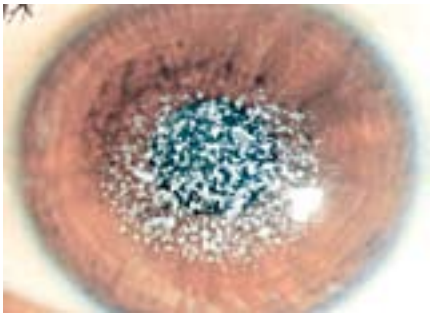


Fig. 3.44.7A: Granular dystrophy—most severe form

- With time the lesions become larger and more numerous and extend into deeper stroma **(Figs 3.44.5A and B)**
- The stroma in between the opacities and the peripheral cornea remains clear **(Figs 3.44.6A and B)**
- In most of the cases, visual acuity usually remains good, but in severe form it affects vision **(Figs 3.44.7A and B)**

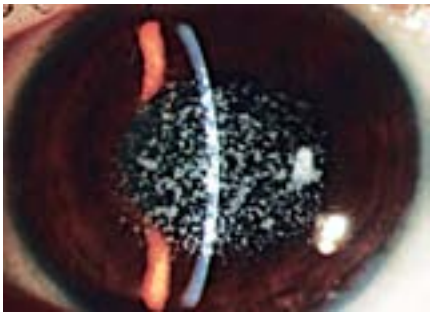


Fig. 3.44.7B: Granular dystrophy—most severe form

- *Treatment:* Deep anterior lamellar keratoplasty (DALK) or penetrating keratoplasty in severe cases. PTK may be tried, but there is high chance of recurrence **(Figs 3.44.8A and B)**.

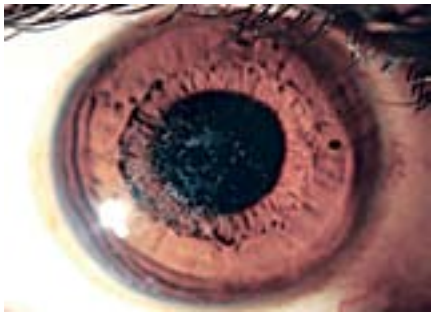


Fig. 3.44.8A: Recurrence of granular dystrophy after PTK

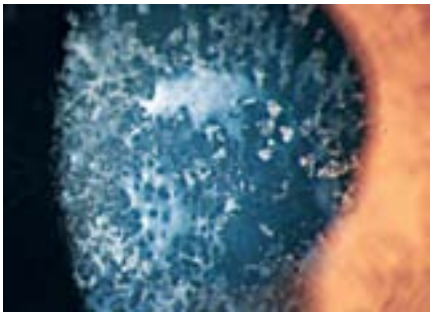


Fig. 3.44.8B: Recurrence of granular dystrophy after PTK

Macular Dystrophy

- Rare, bilateral disease with autosomal recessive inheritance
- Three types:
  - **Type I:** Presents in childhood with recurrent erosion **(Figs 3.45.1A and B)**
  - **Type II:** Presents in second decade with mild erosion **(Figs 3.45.2A to C)**
  - **Type III:** Presents in infancy with severe erosive attacks **(Figs 3.45.3A and B)**

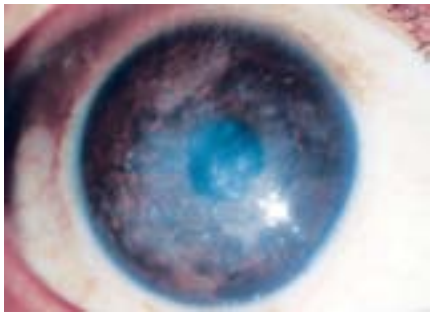


Fig. 3.45.1A: Macular dystrophy—Type I with corneal erosion—RE

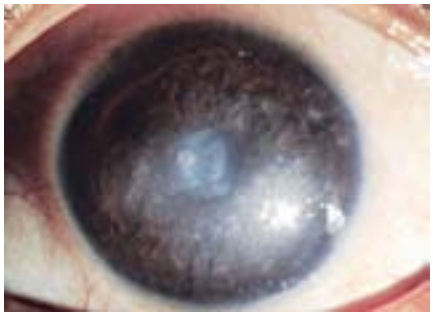


Fig. 3.45.1B: Macular dystrophy—Type I with corneal erosion—LE

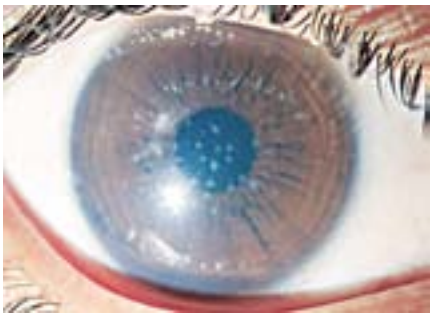


Fig. 3.45.2A: Macular dystrophy—early

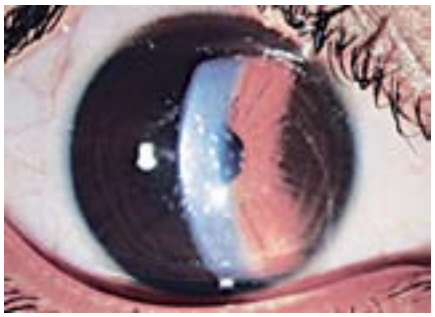


Fig. 3.45.2B: Macular dystrophy—Type II—in slit section

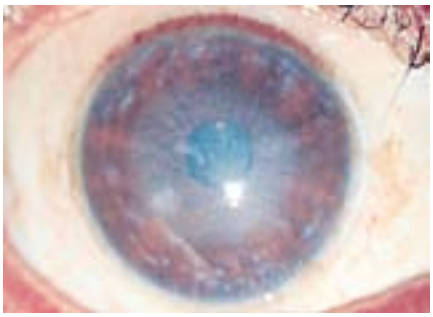


Fig. 3.45.2C: Macular dystrophy—moderate

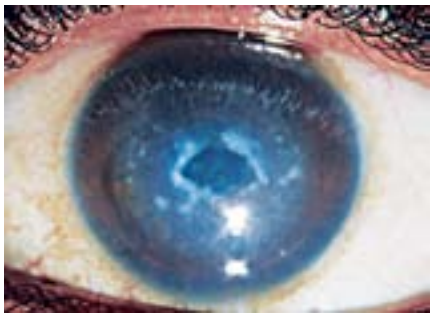


Fig. 3.45.3A: Macular dystrophy—Type III—with erosion

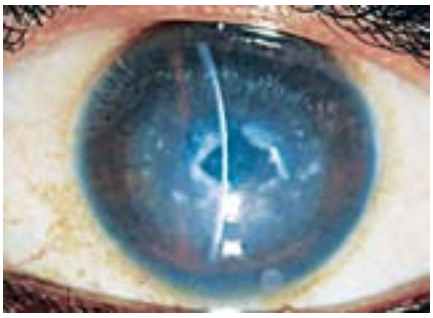


Fig. 3.45.3B: Macular dystrophy—Type III—with erosion

- Significant impairment of vision at an early stage
- Central, focal, grey-white poorly defined opacities in cloudy stroma
- The lesions involve the entire thickness of stroma and it also extends up to the limbus **(Figs 3.45.4A and B)**
- In some cases, here may be associated cornea guttata or frank endothelial dysfunctions **(Figs 3.45.5A and B)**
- *Treatment:* Penetrating keratoplasty; DALK may be possible in selected cases **(Fig. 3.45.6)**.

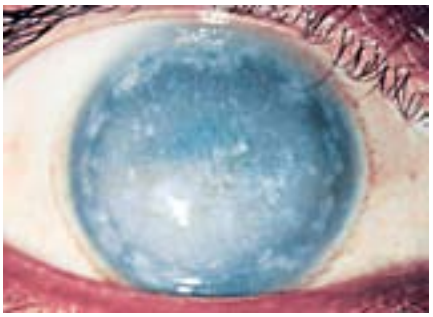


Fig. 3.45.4A: Macular dystrophy—Type III—severe form—RE

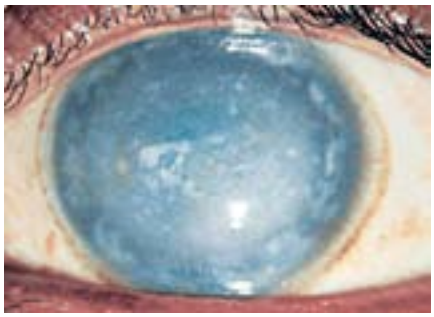


Fig. 3.45.4B: Macular dystrophy—Type III—severe form—LE

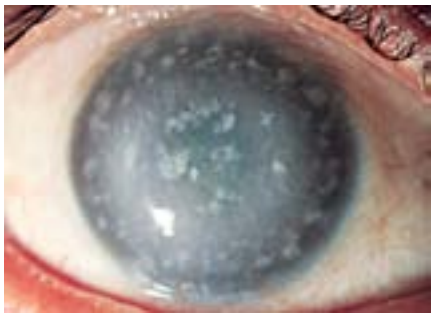


Fig. 3.45.5A: Macular dystrophy—Hassall-Henle bodies—RE

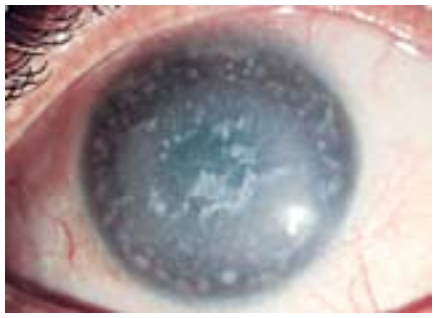


Fig. 3.45.5B: Macular dystrophy—severe with Hassall-Henle bodies—LE

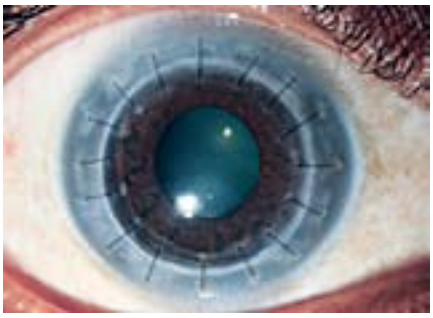


Fig. 3.45.6: Macular dystrophy after DALK of the eye in **Figure 3.45.2C**

Lattice Dystrophy

- Uncommon, bilateral condition with mixed inheritance
- Also *three types:*
  - **Type I:** Autosomal dominant; fine branching spider-like refractile lines which interlace and overlap at different levels within the stroma **(Figs 3.46.1A and B)**
  - **Type II:** Autosomal dominant associated with systemic amyloidosis; thicker lattice lines and less numerous **(Fig. 3.46.2)**

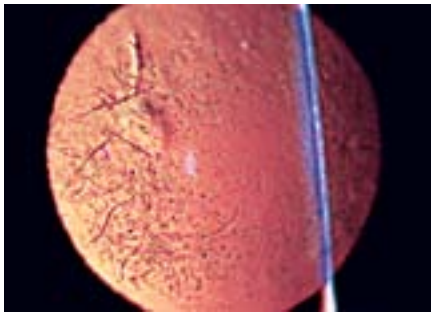


Fig. 3.46.1A: Lattice dystrophy—Type I

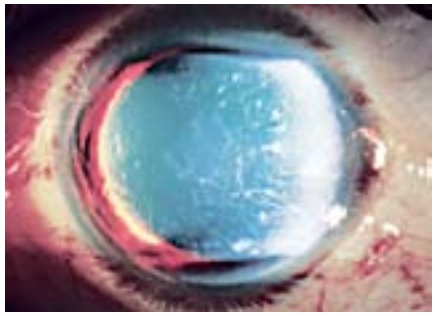


Fig. 3.46.1B: Lattice dystrophy—Type I

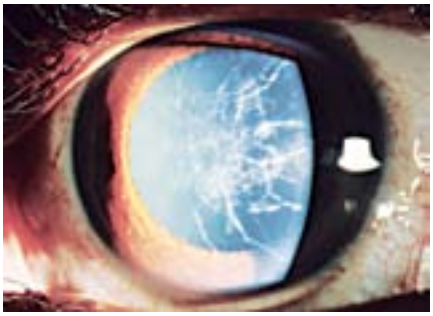


Fig. 3.46.2: Lattice dystrophy—Type II

  - **Type III:** Autosomal recessive; lattice lines coarser than type I and may extend up to the limbus **(Fig. 3.46.3)**
- With time, a diffuse corneal haze develops. Fine Descemet's excrescences is seen in many cases **(Fig. 3.46.4)**
- Visual acuity may be significantly impaired by 30–40 years of age
- Recurrent erosions **(Figs 3.46.5A and B)** and secondary spheroidal degeneration **(*see* Fig. 3.38.4)** are the common problems
- *Treatment:* Penetrating keratoplasty or DALK. Recurrence of the disease in the graft is common.

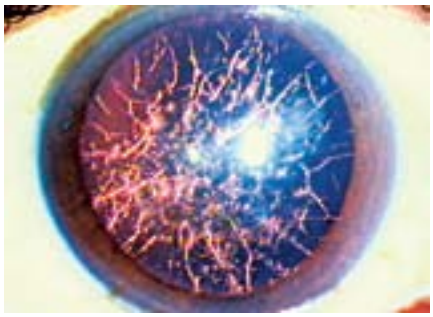


Fig. 3.46.3: Lattice dystrophy—Type III

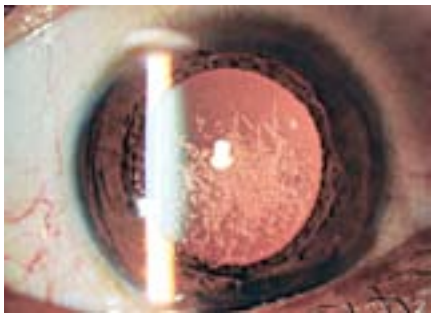


Fig. 3.46.4: Lattice dystrophy—Descemet's excresences

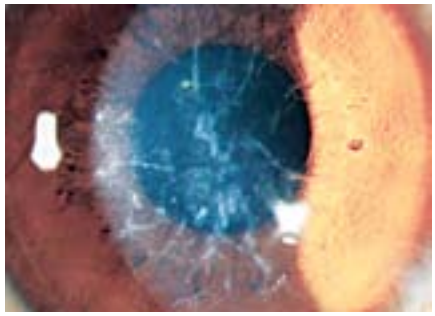


Fig. 3.46.5A: Lattice dystrophy with erosion

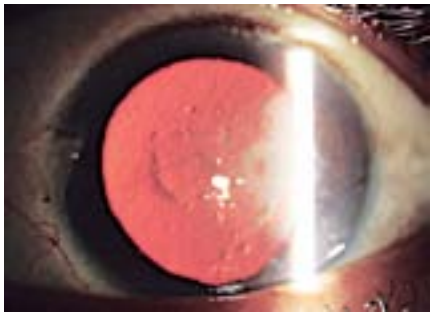


Fig. 3.46.5B: Lattice dystrophy with erosion

Avellino (Granular-Lattice) Dystrophy

- Very rare, autosomal dominant
- Geographically, more found in Avellino, Italy
- Anterior stromal dystrophy is granular and posterior stromal lesions suggestive of lattice **(Figs 3.46.6A and B)**
- Histochemically positive both for hyaline (granular) and amyloid (lattice)
- *Treatment:* Penetrating keratoplasty or DALK.

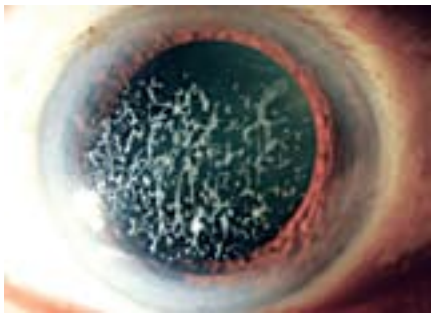


Fig. 3.46.6A: Avellino dystrophy

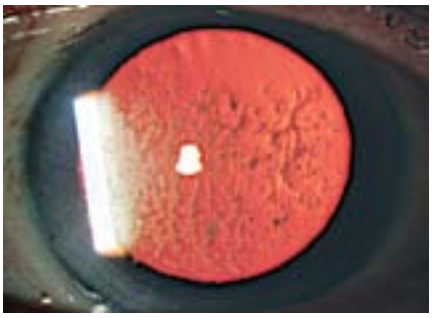


Fig. 3.46.6B: Avellino dystrophy in retroillumination

Central (Schnyder) Crystalline Dystrophy

- Very rare with autosomal dominant inheritance
- Needle-shaped crystalline lesions involving the central stroma **(Figs 3.47.1A and B)**
- Associated with diffuse central stromal haze **(Fig. 3.47.2)**
- *Treatment:* Required in severe cases.

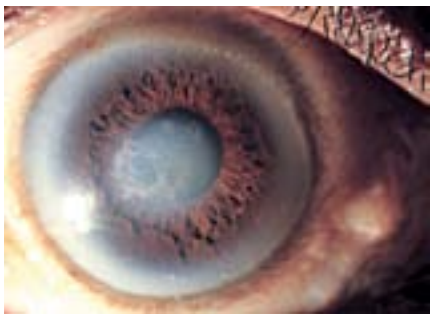


Fig. 3.47.1A: Central (Schnyder) crystalline dystrophy—RE

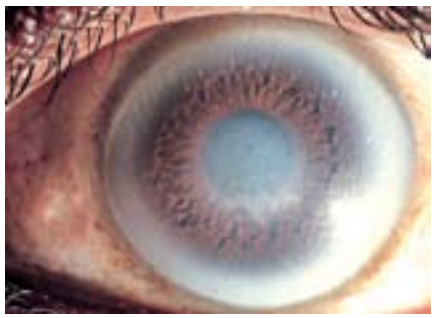


Fig. 3.47.1B: Central (Schnyder) crystalline dystrophy—LE

Fig. 3.47.2: Central (Schnyder) crystalline dystrophy in slit section

Central Discoid Dystrophy

- Very rare
- Central disk-shaped lesion, anterior stromal in nature **(Figs 3.47.3A and B)**
- *Treatment is not required.*

Fig. 3.47.3A: Discoid dystrophy—RE

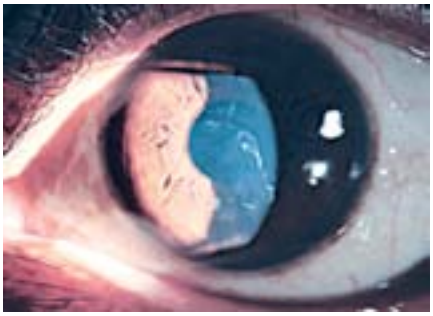


Fig. 3.47.3B: Discoid dystrophy—LE

Congenital Hereditary Stromal Dystrophy

- Very rare, bilateral, with autosomal dominant inheritance and usual manifestation during infancy
- Corneal clouding without edema **(Figs 3.48.1A and B)**
- Usually associated with nystagmus and squint **(Fig. 3.48.2)**
- It is to be differentiated from CHED and congenital glaucoma
- *Treatment:* Penetrating keratoplasty at the earliest. DALK may be considered in selective cases.

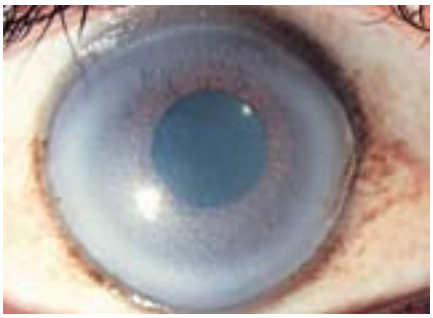


Fig. 3.48.1A: Congenital hereditary stromal dystrophy

Fig. 3.48.1B: Congenital hereditary stromal dystrophy—slit section

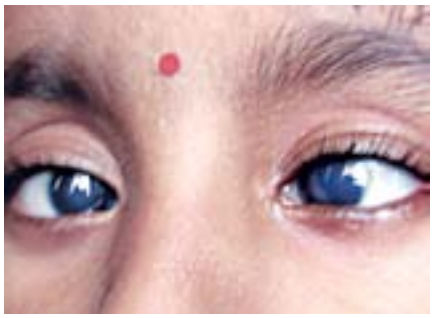


Fig. 3.48.2: Congenital hereditary stromal dystrophy

Posterior Dystrophies

Posterior Polymorphous Dystrophy

- Uncommon, bilateral condition
- Vesicular, band-like, or geographical lesions on the posterior surface of the cornea **(Figs 3.49.1 to 3.49.3)**

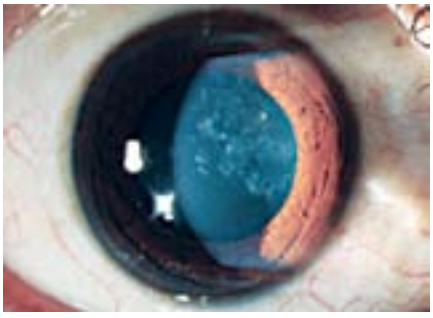


Fig. 3.49.1: Posterior polymorphous dystrophy—vesicular type

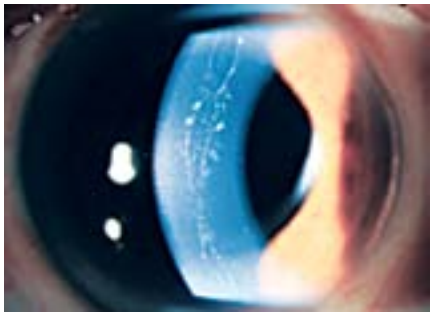


Fig. 3.49.2A: Posterior polymorphous dystrophy—band like

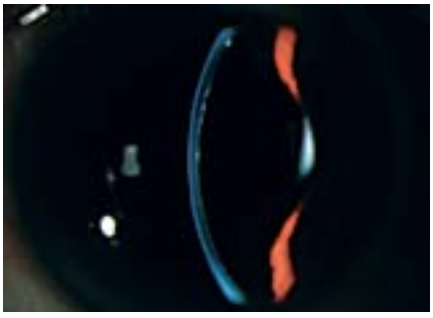


Fig. 3.49.2B: Posterior polymorphous dystrophy—band like

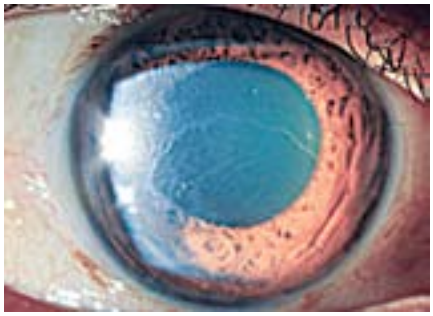


Fig. 3.49.3A: Posterior polymorphous dystrophy—geographical type

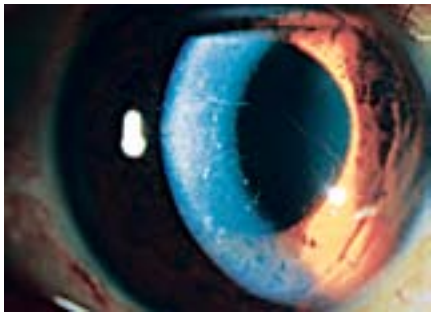


Fig. 3.49.3B: Posterior polymorphous dystrophy—geographical type

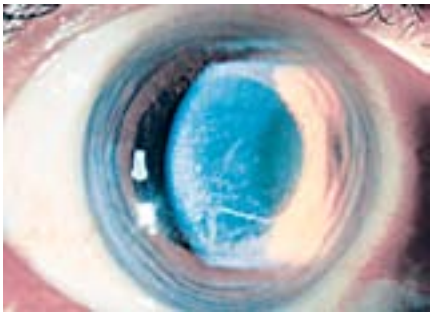


Fig. 3.49.4: Posterior polymorphous dystrophy with corneal edema

- The lesion may be easily overlooked
- In severe cases, there may be corneal edema **(Fig. 3.49.4)**
- *Treatment:* Descemet's stripping endothelial keratoplasty (DSEK or DSAEK) may be considered in selective cases.

Cornea Guttata

- A common aging process resulting in focal accumulation of excrescences on the posterior surface of the Descemet's membrane
- They disrupt the normal endothelial mosaic
- With confluent lesions, they appear as dark spots or beaten metal appearance **(Figs 3.50.1A and B)**
- They may be seen in early stage of Fuchs' dystrophy
- *Treatment:* Close observation with serial specular microscopy to see the progress **(Figs 3.50.2A and B)**. Special precaution to be taken protect endothelium during cataract surgery.

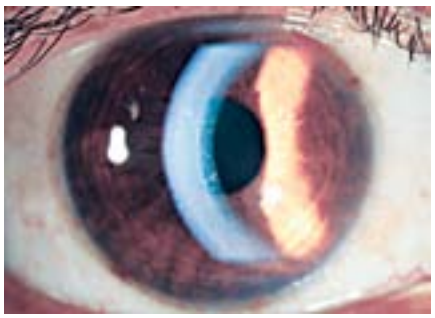


Fig. 3.50.1A: Cornea guttata

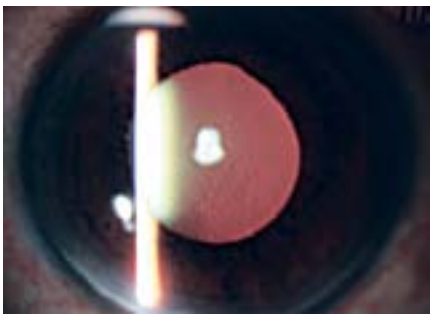


Fig. 3.50.1B: Cornea guttata—retroillumination

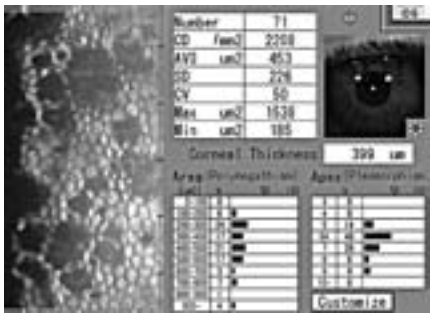


Fig. 3.50.2A: Specular microscopy in cornea guttata

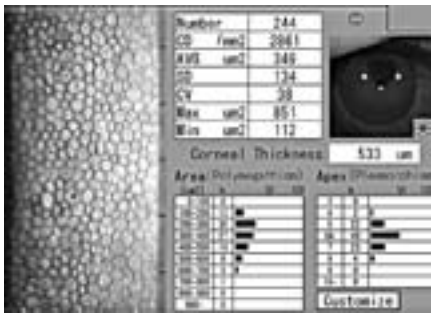


Fig. 3.50.2B: Specular microscopy in normal cornea

Fuchs' Endothelial Dystrophy

- Relatively common, bilateral condition with autosomal dominant inheritance
- Slowly progressive, and more common in elderly female
- Central corneal guttata without any symptom which gradually spreads towards periphery **(Fig. 3.51.1)**
- Stroma becomes edematous with endothelial decompensation **(Figs 3.51.2A and B)**

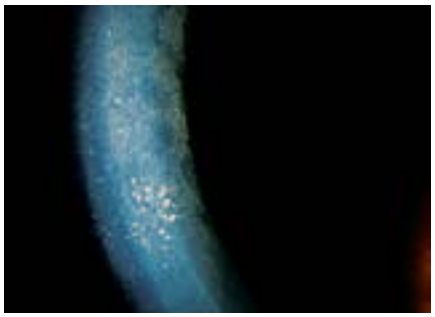


Fig. 3.51.1: Fuchs' dystrophy—guttate changes in specular reflection

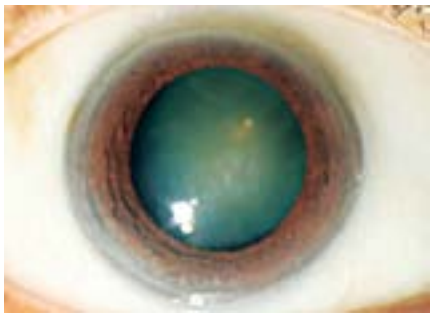


Fig. 3.51.2A: Fuchs' dystrophy—central corneal edema

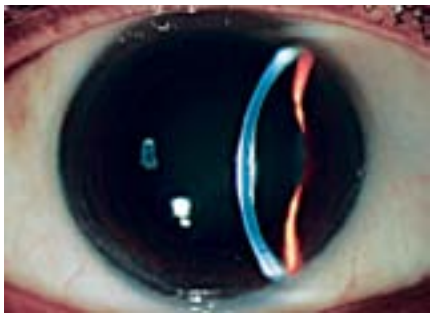


Fig. 3.51.2B: Fuchs' dystrophy—central corneal edema

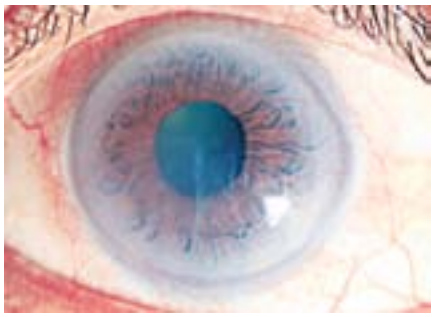


Fig. 3.51.3: Fuchs' dystrophy—central corneal edema—endothelial folds

- Epithelial edema gradually develops with impairment of vision **(Fig. 3.51.3)**
- Ultimately, bullous keratopathy develops with severe symptoms **(Figs 3.51.4 to 3.51.6)**
- Gradually, scarring occurs with vascularization **(Fig. 3.51.7)**
- *Treatment:* Hypertonic saline, bandage contact lens and ultimately DSEK or rarely PK **(Figs 3.51.8A and B)**.

Fig. 3.51.4: Fuchs' dystrophy—epithelial edema with bullous changes

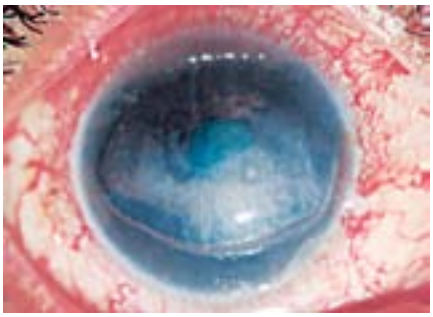


Fig. 3.51.5: Fuchs' dystrophy—epithelial edema with severe bullous changes

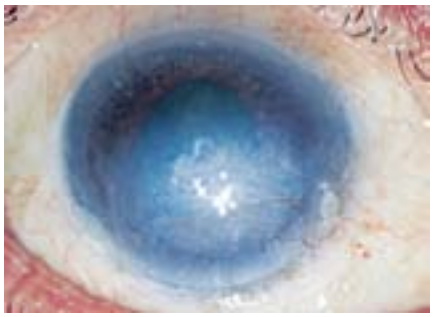


Fig. 3.51.6A: Fuchs' dystrophy—central corneal edema—vascularization

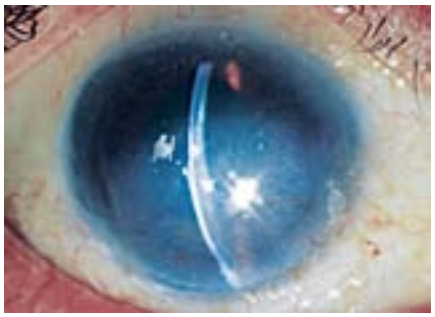


Fig. 3.51.6B: Fuchs' dystrophy—central corneal edema—vascularization

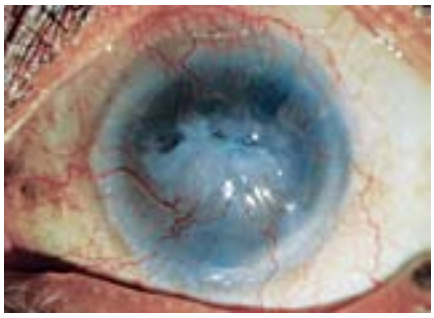


Fig. 3.51.7: Fuchs' dystrophy—advanced with severe vascularization

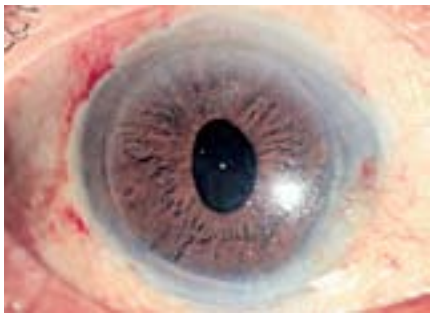


Fig. 3.51.8A: Fuchs' dystrophy—DSEK same patient **Figures 3.51.6A and B**

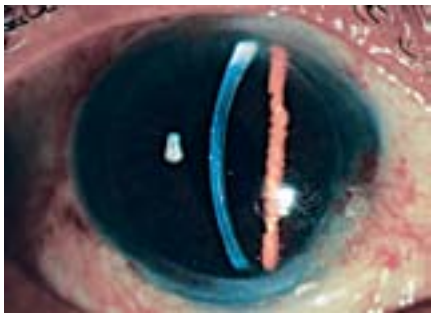


Fig. 3.51.8B: Fuchs' dystrophy—DSEK same patient **Figures 3.51.6A and B**

Congenital Hereditary Endothelial Dystrophy (CHED)

- Two types: CHED1 and CHED2. Both are bilateral and rare.
  - *CHED1* is autosomal dominant and less severe; whereas,
  - *CHED2* is autosomal recessive and more severe with total absence of endothelium.
- They present as bilateral corneal edema any time during first decade of life. In milder form, cornea is ground-glass appearance **(Figs 3.52.1A and B)**; gradually it becomes more cloudy **(Figs 3.52.2 and 3.52.3)**
- And ultimately in most severe form, it is totally opaque **(Figs 3.52.4A and B)**

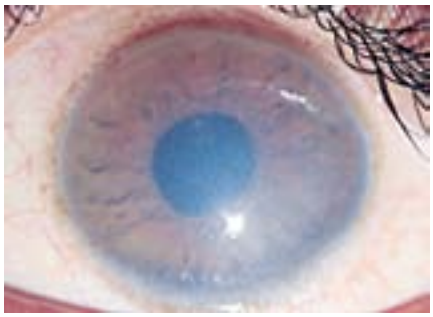


Fig. 3.52.1A: Congenital hereditary endothelial dystrophy

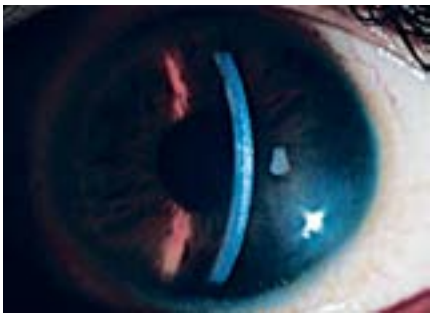


Fig. 3.52.1B: Congenital hereditary endothelial dystrophy—corneal edema

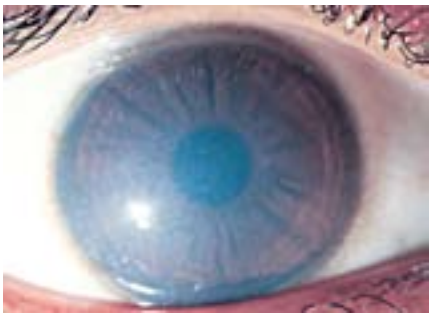


Fig. 3.52.2A: Congenital hereditary endothelial dystrophy—more edema

Fig. 3.52.2B: Congenital hereditary endothelial dystrophy—more edema

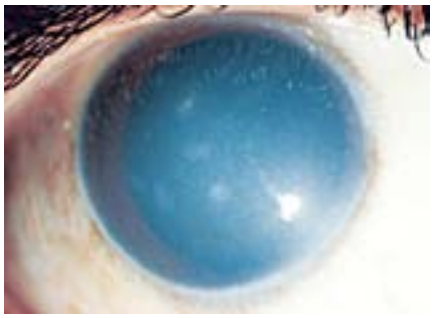


Fig. 3.52.3A: Congenital hereditary endothelial dystrophy—severe edema

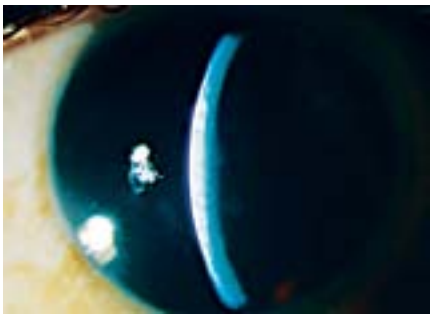


Fig. 3.52.3B: Congenital hereditary endothelial dystrophy—severe edema

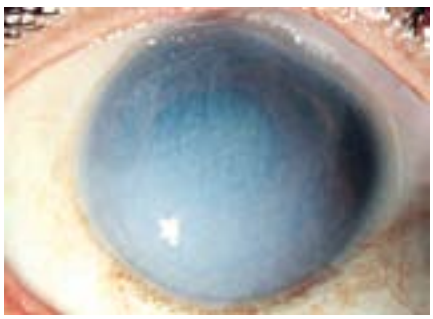


Fig. 3.52.4A: Congenital hereditary endothelial dystrophy—most severe form

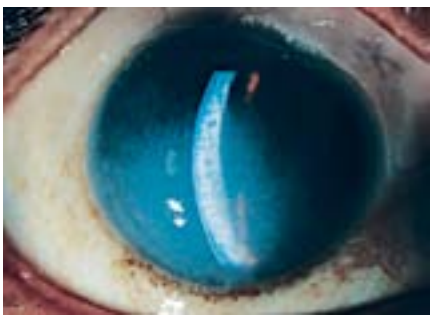


Fig. 3.52.4B: Congenital hereditary endothelial dystrophy—most severe form

- Infantile form should be differentiated from congenital glaucoma **(Figs 3.52.5A and B)**
- *Treatment:* Penetrating keratoplasty at the earliest. DSEK can also be performed with favorable results.

Fig. 3.52.5A: Congenital hereditary endothelial dystrophy

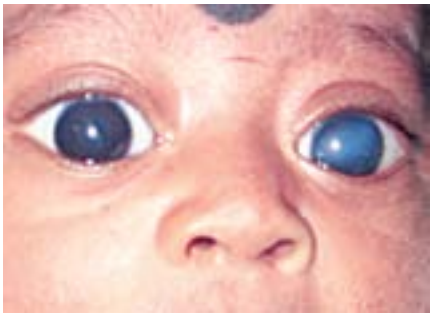


Fig. 3.52.5B: Bilateral congenital glaucoma—LE more severe

Keratoconus

- Bilateral conical protrusion of the central part of the cornea with thinning
- Starts around puberty and slowly progressive
- May be associated with vernal conjunctivitis, atopic conjunctivitis, *ectopia lentis* or Down's syndrome
- Irregular retinoscopic reflex and high irregular myopic astigmatism
- Abnormal *oil-droplet* red reflex
- Thinning of the central cornea with protrusion just below and nasal to the center **(Figs 3.53.1 and 3.53.2)**
- *Munson's sign:* Bulging of lower lids in down gaze **(Fig. 3.53.3)**
- *Vogt's lines (striae):* Vertical folds at the level of deep stroma and Descemet's membrane **(Fig. 3.53.4A)**
- *Fleischer's ring:* Epithelial iron line at the base of the cone **(Figs 3.53.4B and C)**

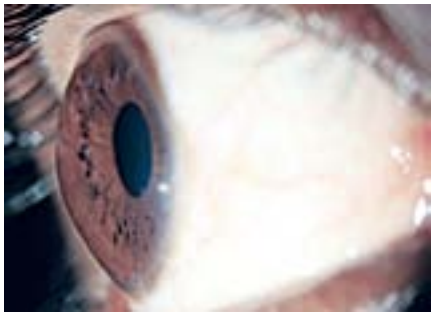


Fig. 3.53.1A: Keratoconus—conical protrusion with thinning

Fig. 3.53.1B: Keratoconus—conical protrusion with thinning in slit section

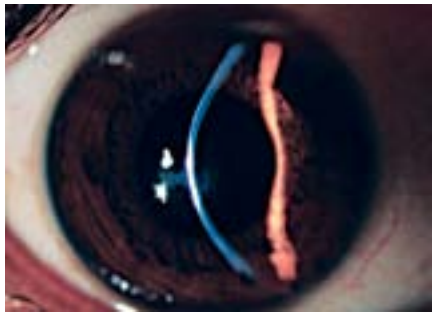


Fig. 3.53.2: Keratoconus advanced—thinning at center

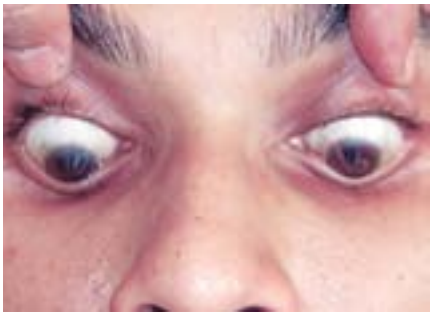


Fig. 3.53.3: Keratoconus—Munson's sign

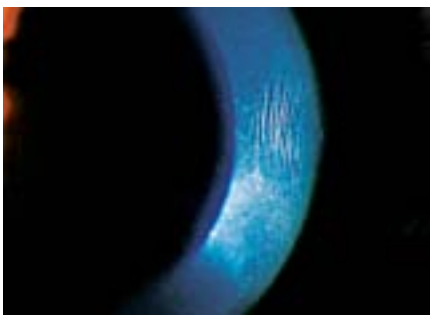


Fig. 3.53.4A: Keratoconus—Vogt's striae

Fig. 3.53.4B: Fleischer's ring under cobalt blue light

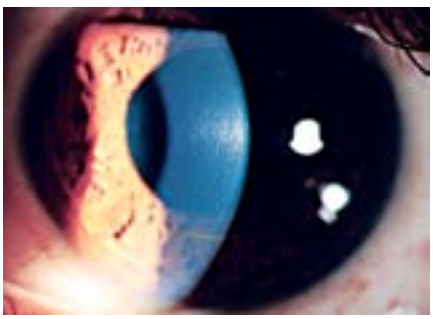


Fig. 3.53.4C: Keratoconus—Vogt's striae and Fleischer's ring

- *Rizutti's sign:* Corneal reflection on the nasal limbus when light is thrown from the temporal limbus **(Figs 3.53.5A and B)**
- Prominent corneal nerves **(Figs 3.53.6A and B)**
- ***Acute hydrops:*** Sudden corneal edema due to acute seepage of the aqueous into the corneal stroma and epithelium resulting from rupture of Descemet's membrane **(Figs 3.53.7A and B)**
- Variable degree of apical corneal scarring **(Figs 3.53.8A and B)** and the extent of rupture can be visible **(Fig. 3.53.9)**

Fig. 3.53.5A: Keratoconus—Rizutti's sign

Fig. 3.53.5B: Keratoconus—Rizzuti's sign

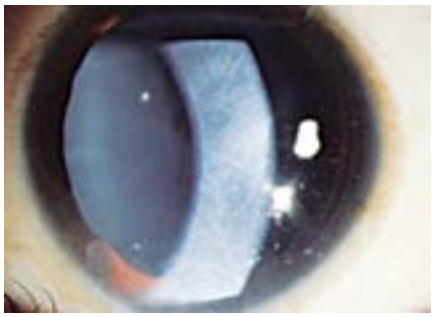


Fig. 3.53.6A: Keratoconus—prominent corneal nerves

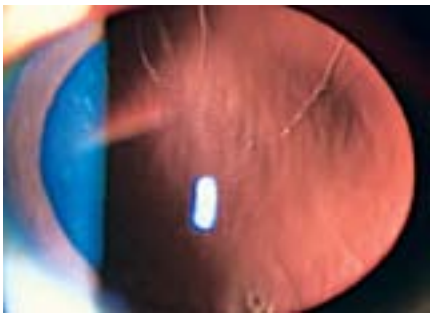


Fig. 3.53.6B: Keratoconus—prominent corneal nerve

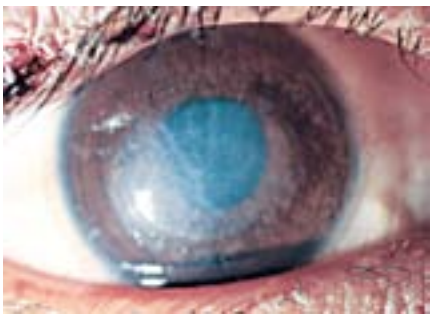


Fig. 3.53.7A: Keratoconus—acute hydrops

Fig. 3.53.7B: Keratoconus—acute hydrops

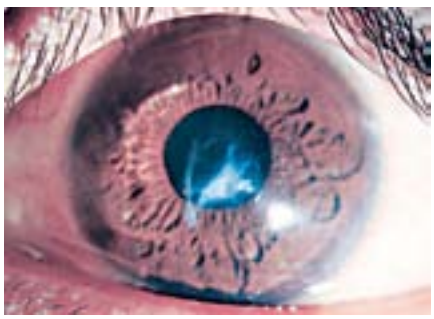


Fig. 3.53.8A: Keratoconus—apical scarring

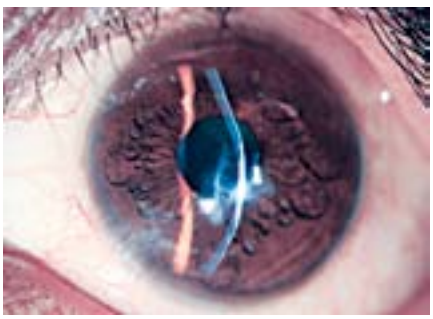


Fig. 3.53.8B: Keratoconus—apical scarring

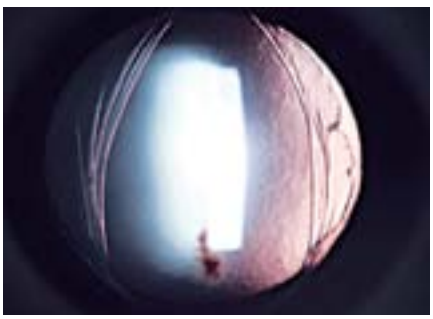


Fig. 3.53.9: Keratoconus—after a large rupture of Descemet's membrane

- *Treatment:* Spectacles, corneal collagen cross-linking with riboflavin (C3R), RGP contact lenses, Rose K contact lens, INTACS, deep anterior lamellar keratoplasty (DALK) **(Figs 3.53.10A to C)** or penetrating keratoplasty.

Fig. 3.53.10A: DALK in keratoconus—preoperative

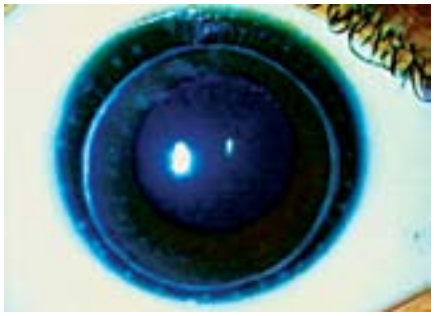


Fig. 3.53.10B: DALK in keratoconus—postoperative

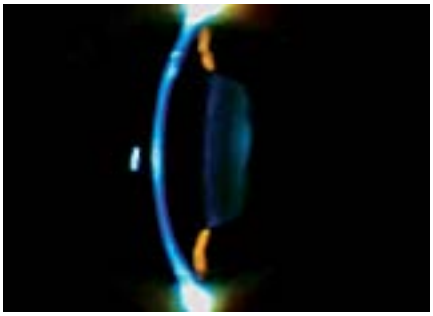


Fig. 3.53.10C: DALK in keratoconus—postoperative

Miscellaneous Corneal Conditions

Corneal Abrasion

- Common, unilateral condition usually associated with trauma. Epithelial defect can be easily diagnosed by diffuse illumination and after fluorescein staining **(Figs 3.54.1 and 3.54.2)**
- *Treatment:* Antibiotic ointment and patching for 24 hours usually sufficient for epithelial healing. It is important to examine the patient on the next day.

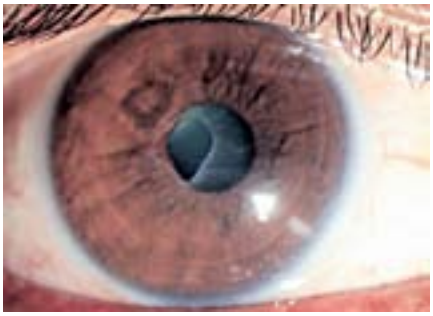


Fig. 3.54.1A: Corneal abrasion

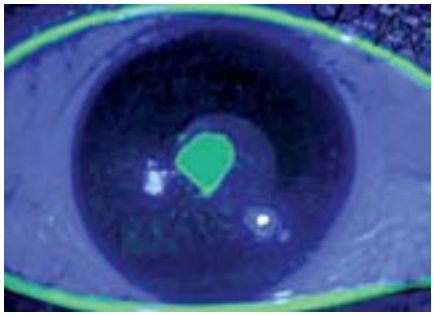


Fig. 3.54.1B: Corneal abrasion after fluorescein staining

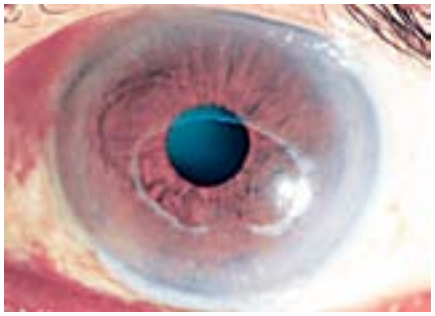


Fig. 3.54.2: Corneal abrasion—large

Keratoconjunctivitis Sicca (KCS)

- Very common bilateral condition mainly occurs in postmenopausal women which may occur in isolation or in associated with some systemic diseases, like rheumatoid arthritis
- Mucus debris or plaque **(Fig. 3.55.1)**
- Decreased tear meniscus height **(Fig. 3.55.2)**

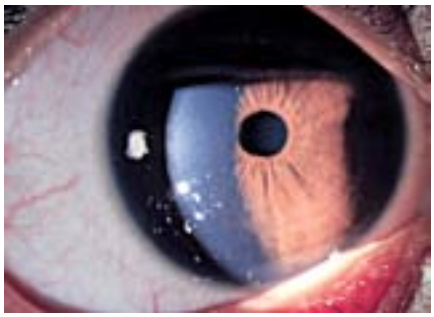


Fig. 3.55.1: Keratoconjunctivitis sicca (KCS)—mucus debris

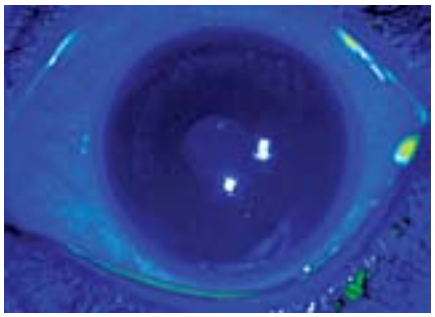


Fig. 3.55.2: KCS—reduced tear meniscus height

- Reduced tear film break-up-time (<10 second is clinically significant)
- Staining of interpalpebral area with rose bengal in triangular fashion **(Figs 3.55.3A and B and *see* Fig. 2.42.3)**, later on staining of the cornea **(Fig. 3.55.4)**. Lissamine green can be used to stain instead of rose bengal and it is less **(Fig. 3.55.5)**
- Lustreless cornea **(Fig. 3.55.6)**
- Superficial punctate keratitis stained with rose bengal or fluorescein **(Figs 3.55.7A and B)**
- Filamentary keratopathy stained with fluorescein **(Fig. 3.55.8)**
- Dellen and corneal thinning leading to Descemetocele formation **(Figs 3.55.9 and 3.55.10)**

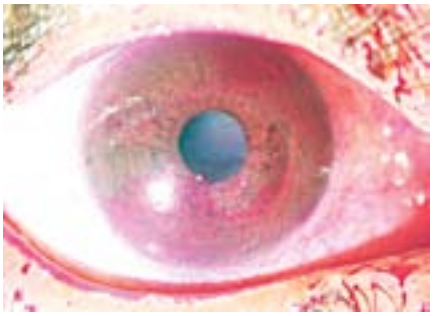


Fig. 3.55.3A: KCS—rose bengal staining

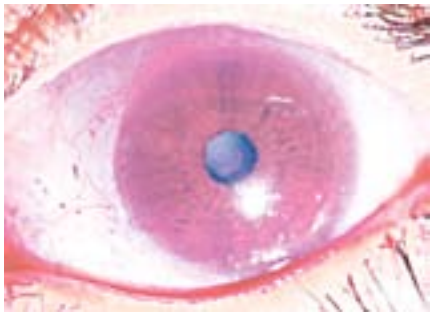


Fig. 3.55.3B: KCS—rose bengal staining—moderate

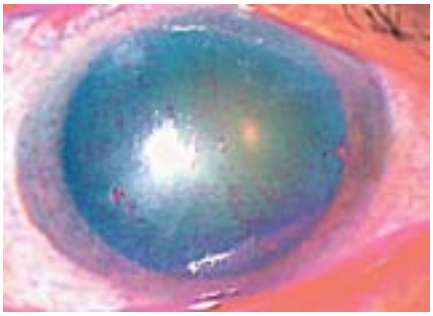


Fig. 3.55.4: KCS—rose bengal staining—extensive

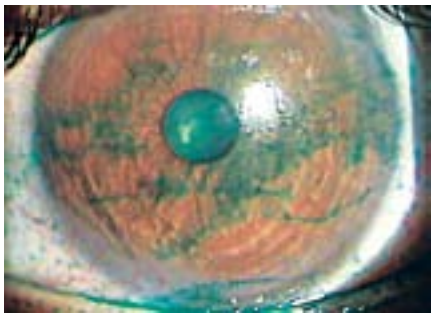


Fig. 3.55.5: KCS—Lissamine green stain

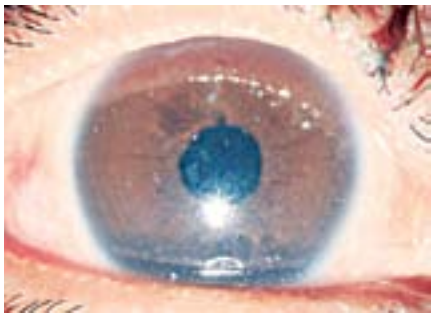


Fig. 3.55.6: KCS—lustreless cornea

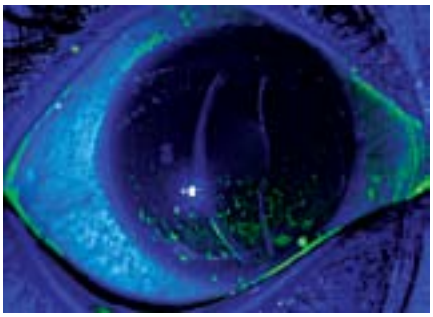


Fig. 3.55.7A: KCS—fluorescein stain—SPK

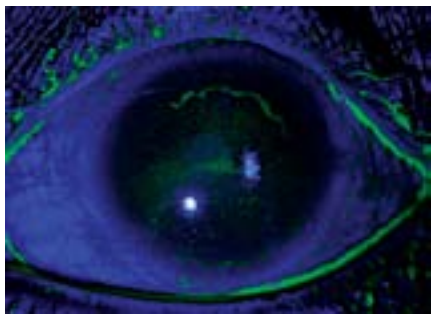


Fig. 3.55.7B: KCS—fluorescein staining—extensive

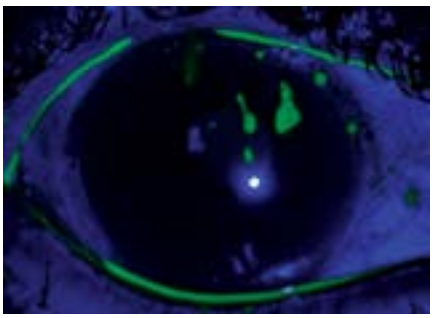


Fig. 3.55.8: KCS—filaments

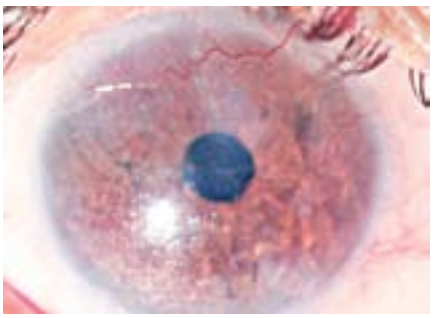


Fig. 3.55.9: Dry eye—dellen formation and vascularization

- Vascularization and keratinization in severe cases **(Figs 3.55.11 and 3.55.12)**
- *Treatment:* Preservative-free tear substitute 4 times to 1 hourly; topical cyclosporine (0.05%) twice daily for at least 6 months; soft steroids like loteprednol in tapering doses; oral omega 3 fatty acid; punctal occlusion, autologous serum drops (20%), partial tarsorrhaphy, etc.

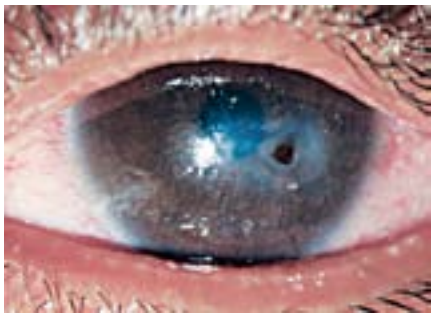


Fig. 3.55.10: Dry eye—thinning and descemetocele

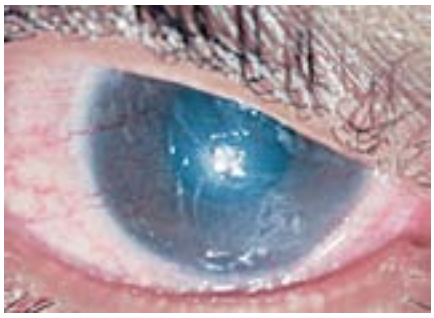


Fig. 3.55.11: Dry eye—vascularization

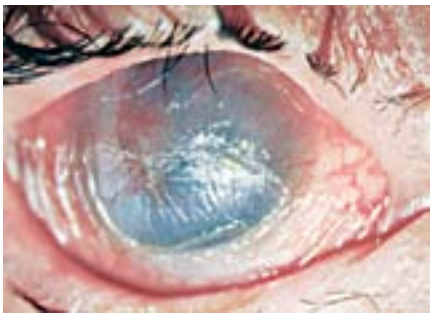


Fig. 3.55.12: Dry eye—keratinization

Vortex Keratopathy

- Bilateral condition, more commonly caused by a variety of oral medicine and also in Fabry's disease
- The drugs are: Hydroxychloroquine, amiodarone, indomethacin, tamoxifen, chlorpromazine, etc. **(Figs 3.56.1A and B)**
- Appear as minute grayish or golden epithelial deposits arranged in a vortex fashion
- They start at a point below the pupil and swirling outwards without involving the limbus **(Figs 3.56.2A and B)**
- *Treatment:* Withdrawal of the specific drug and frequent tears substitutes.

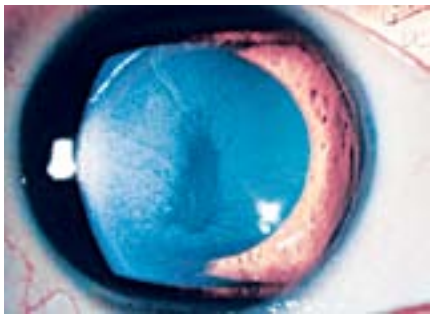


Fig. 3.56.1A: Vortex keratopathy—tamoxifen induced

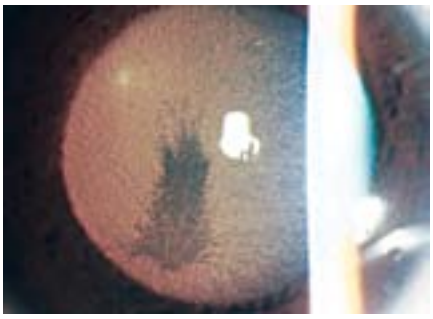


Fig. 3.56.1B: Vortex keratopathy in retroillumination

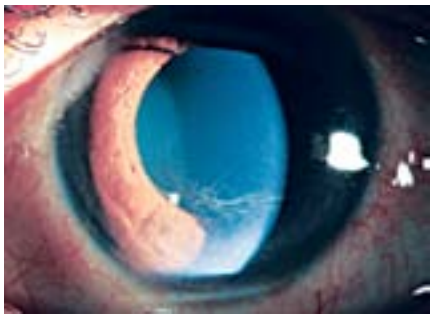


Fig. 3.56.2A: Vortex keratopathy

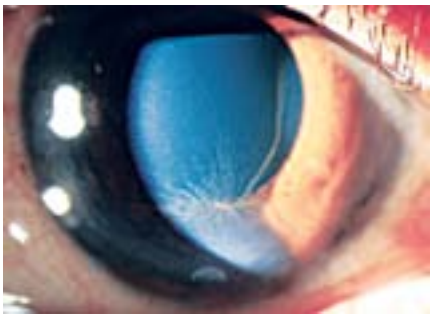


Fig. 3.56.2B: Vortex keratopathy

Descemet's Detachment

- Descemet's membrane detachment is mainly caused by surgical trauma especially during cataract surgery. It may be partial or total and associated with diffuse or localized corneal edema **(Figs 3.57.1 to 3.57.5)**

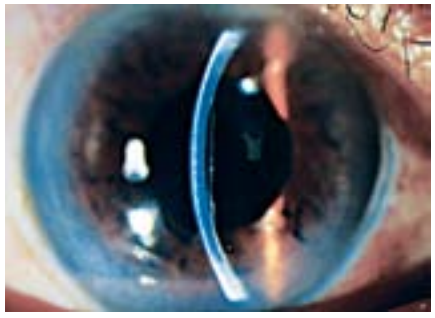


Fig. 3.57.1: Descemet's detachment

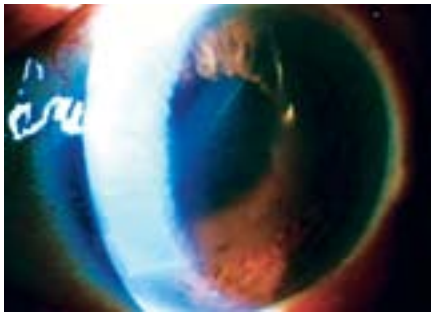


Fig. 3.57.2: Descemet's detachment—curled

- *Treatment:* Surgery with tamponade by air or C_3F_8 gas to reattach the membrane. In total detachment—if this fails, DSEK may be considered.

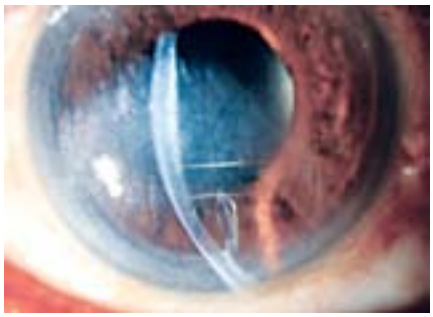


Fig. 3.57.3: Descemet's tear—roll—post-surgical with corneal edema

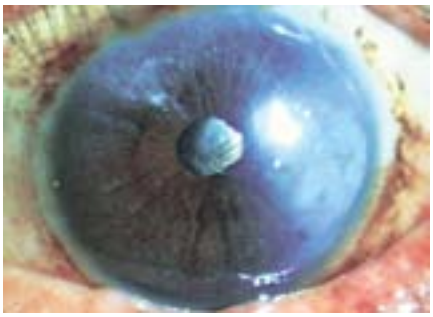


Fig. 3.57.4: Descemet's detachment—partial with localized edema

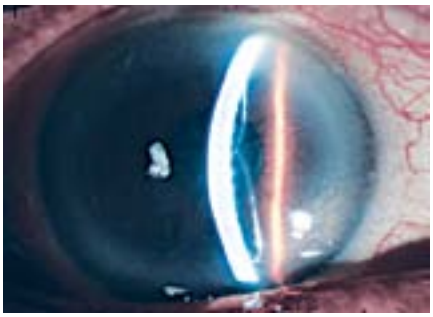


Fig. 3.57.5: Descemet's detachment—total corneal edema

Descemet's Tear

- *Causes*: Vertical/oblique tear in birth trauma **(Figs 3.58.1A and B)**; horizontal tear in congenital glaucoma (Haab's striae) **(Fig. 3.58.2)**; elliptical tear in keratoconus/keratoglobus **(Figs 3.58.3A and B)**; and in any direction in surgical trauma
- *Treatment:* Observation; no active treatment is required.

Fig. 3.58.1A: Descemet's tear—birth trauma

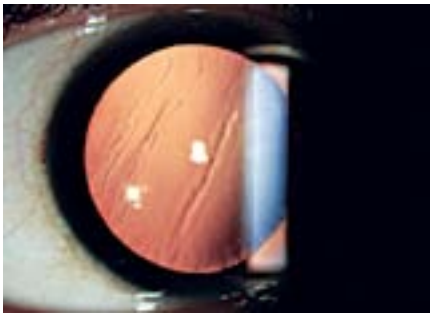


Fig. 3.58.1B: Descemet's tear—birth trauma

Fig. 3.58.2: Haab's striae—congenital glaucoma

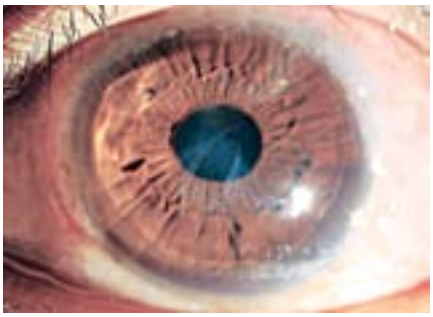


Fig. 3.58.3A: Descemet's tear—keratoconus

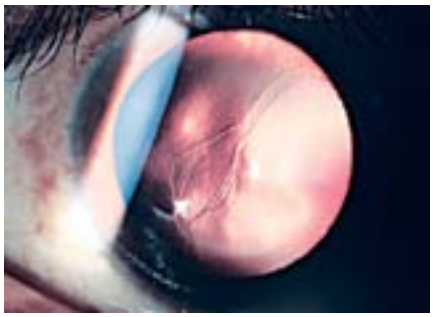


Fig. 3.58.3B: Descemet's tear—keratoconus

Descemet's Folds and Wrinkles

- Associated with a variety of ocular conditions and best visible in slit lamp under retroillumination
- It may be seen following surgical trauma **(Figs 3.59.1A and B)** as *striate keratopathy*, retained lens

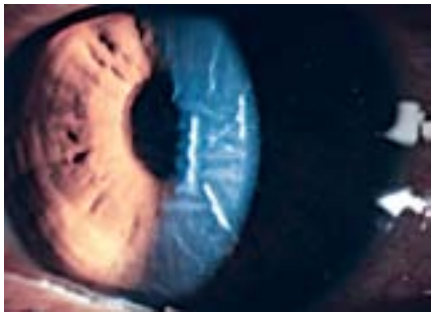


Fig. 3.59.1A: Descemet's fold following phacoemulsification

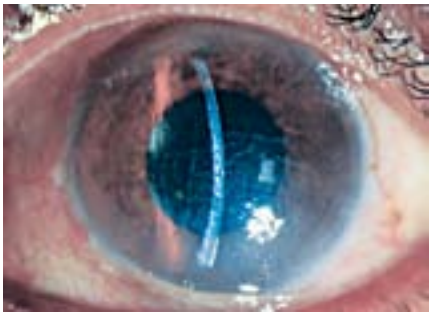


Fig. 3.59.1B: Descemet's folds following manual SICS

fragment in AC **(Fig. 3.59.2)**, tight suturing, chemical burn/toxic keratopathy **(Figs 3.59.3A and B)**, ocular hypotony, disciform keratitis **(Fig. 3.59.4)**, congenital syphilis, blunt trauma **(Fig. 3.59.5)**, birth trauma **(Fig. 3.59.6)**, etc.

- *Treatment:* According to the cause. In most cases, it disappears after treatment. Rarely, when it is with severe endothelial decompensation, DSEK surgery may be required.

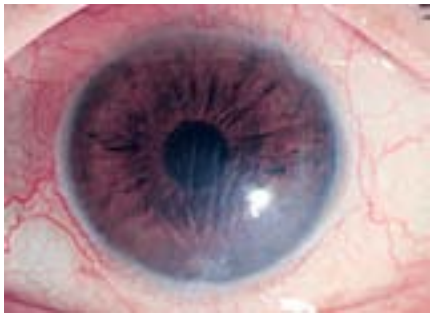


Fig. 3.59.2: Descemet's folds—retained lens frament after phacoemulsification

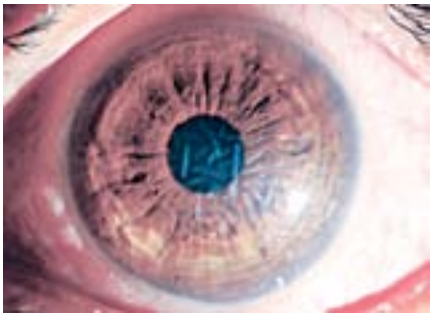


Fig. 3.59.3A: Descemet's fold following toxic keratopathy (calotropis latex)

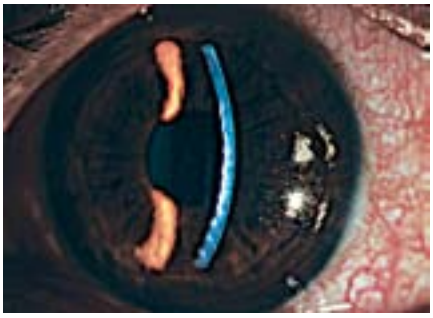


Fig. 3.59.3B: Descemet's fold following toxic keratopathy (calotropis latex)

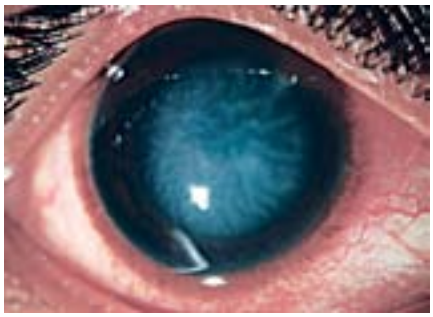


Fig. 3.59.4: Descemet's folds—disciform keratitis

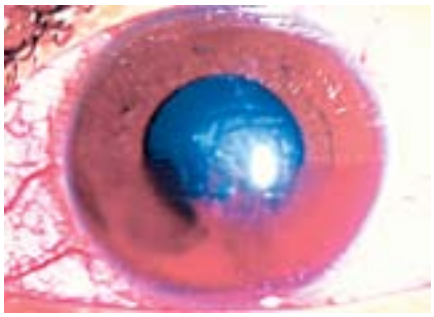


Fig. 3.59.5: Descemet's fold—blunt trauma

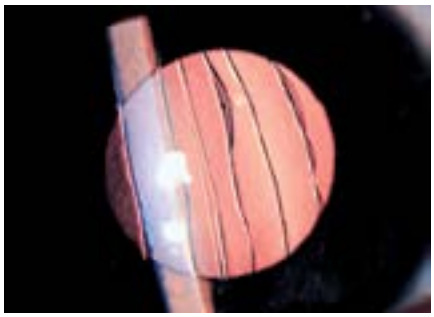


Fig. 3.59.6: Extensive Descemet's folds following birth trauma

Krukenberg's Spindle

- Spindle-shaped deposition of the pigments on the back of the cornea. It is usually arranged vertically on the endothelial surface **(Figs 3.60.1A and B)**
- May be associated with pigment dispersion syndrome with glaucoma
- *Treatment:* Total glaucoma work up and investigations followed by management of glaucoma if present.

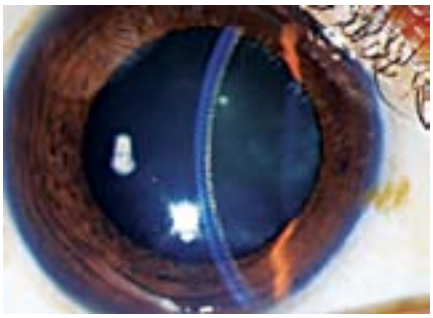


Fig. 3.60.1A: Krukenberg's spindle

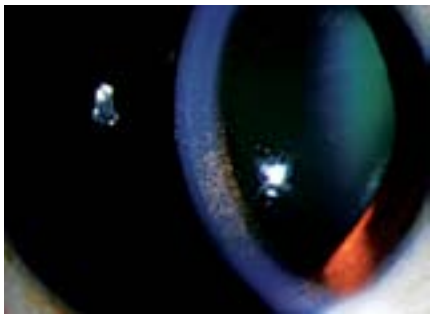


Fig. 3.60.1B: Krukenberg's spindle in pigmentary glaucoma

Corneal Dellen

- It may be seen in *keratoconjunctivitis sicca* (KCS), adjacent to a pterygium or pinguecule and other nodular limbal lesion. Epithelium is intact and does not stain with fluorescein, but pooling of the dye may be present. It may be small **(Figs 3.61.1A and B)** or large **(Figs 3.61.2A and B)**
- *Treatment:* Frequent tears substitutes and treatment of the cause.

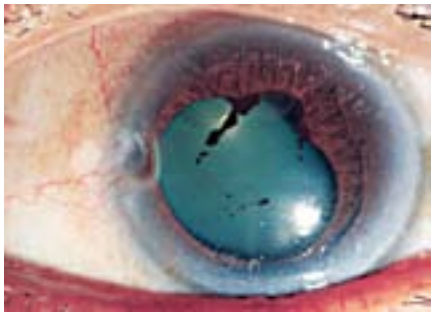


Fig. 3.61.1A: Corneal dellen—small adjacent to a phlycten

Fig. 3.61.1B: Corneal dellen—small

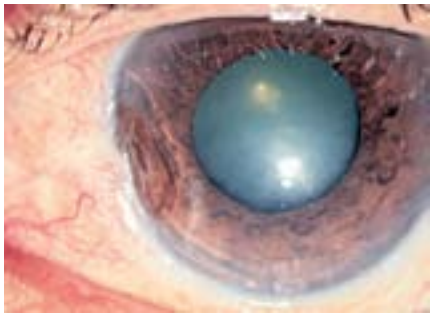


Fig. 3.61.2A: Corneal dellen—large

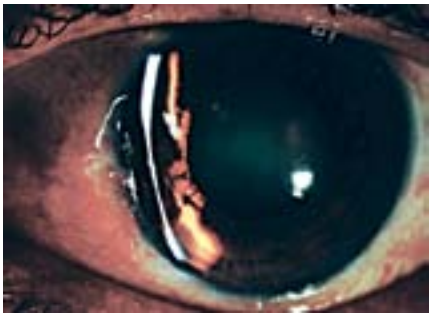


Fig. 3.61.2B: Corneal dellen—large

Crocodile Shagreen

- Uncommon, bilateral, innocuous condition. Grayish-white polygonal opacities separated by clear spaces
- *Anterior crocodile shagreen*—involve anterior two-third of stroma and usually more peripheral **(Fig. 3.62.1)**. It is more frequent than posterior type which is usually central in location **(Figs 3.62.2A and B)**
- *Treatment:* Not required.

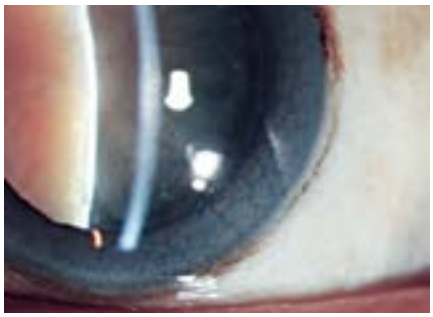


Fig. 3.62.1: Crocodile shagreen—anterior and peripheral

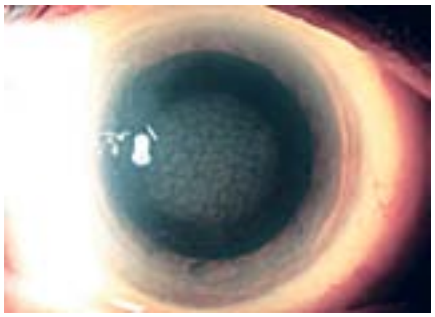


Fig. 3.62.2A: Crocodile shagreen—posterior and central

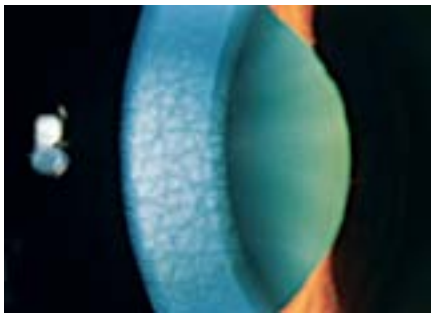


Fig. 3.62.2B: Crocodile shagreen—posterior and central

Prominent Corneal Nerves

- They are associated with a variety of ocular and systemic conditions. *The causes are:* neurofibromatosis, leprosy, primary amyloidosis, keratoconus, *Acanthamoeba* keratitis, failed graft, etc. **(Figs 3.63.1A and B and *see* Figs 3.53.6A and B)**
- *No treatment is required.*

Fig. 3.63.1A: Prominent corneal nerves

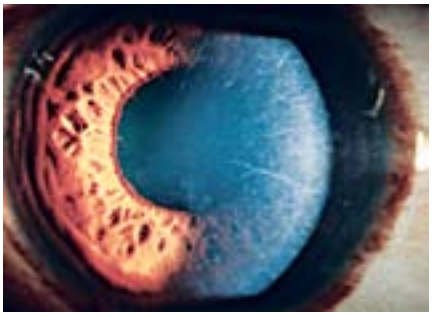


Fig. 3.63.1B: Prominent corneal nerves

Xerophthalmia (Corneal Signs in Avitaminosis-A)

- *Corneal xerosis (X2):* Hazy lusterless, dry cornea, mainly in the inferior part **(Fig. 3.64.1)**
- *Keratomalacia (X3A, 3B):* Round, oval, punched out defects, surrounded by xerotic cornea; perforation may occur within 24 hours with pseudocornea and anterior staphyloma formation **(Fig. 3.64.2)**
  - X3A: when <1/3rd cornea is involved
  - X3B: when >1/3rd cornea is involved
- *Xerophthalmic scar (Xs):* Healed sequelae of X3A or X3B, typically inferior in location **(Fig. 3.64.3)**. It includes nebula, macula, leucoma, adherent leucoma, etc.
- *Treatment:* It is a medical emergency; massive dose of vitamin A (orally or parenterally); treating malnutrition and underlying systemic illness; prophylactic vitamin A therapy up to the age of 6 years.

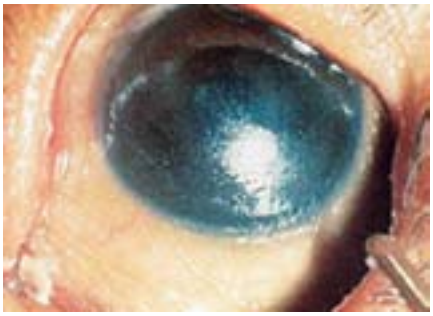


Fig. 3.64.1: Corneal xerosis—X2

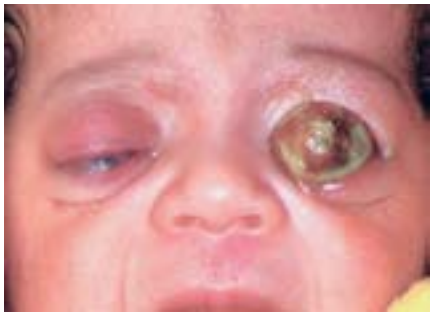


Fig. 3.64.2: Keratomalacia—RE and with anterior staphyloma—LE—X3B

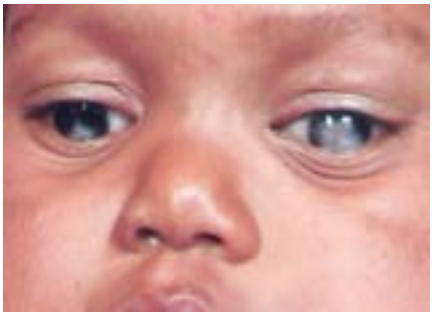


Fig. 3.64.3: Xerophthalmic scar—Xs

Tunnel Abscess

- Associated with infiltrations within the tunnel after phaco or SICS **(Figs 3.65.1 and 3.64.2)**. Variable degree of anterior chamber reactions and eventually it may progress into frank endophthalmitis **(Fig. 3.65.3)**. Sometime it may perforate with iris prolapse **(Fig. 3.65.4)**
- *Treatment:* Exploration and scraping the materials for culture and sensitivity, and tunnel wash with vancomycin or amphotericin-B. Prognosis is usually poor. In difficult cases, a tectonic sclera-corneal patch graft often save the eyeball **(Figs 3.65.5 and 3.65.6)**.

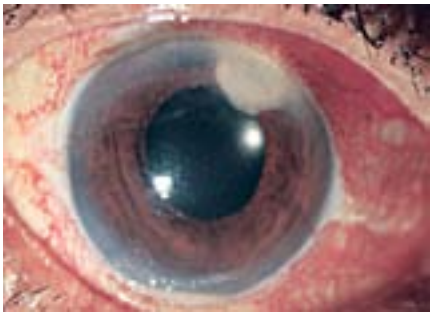


Fig. 3.65.1: Tunnel abscess after phacoemulsification—side port

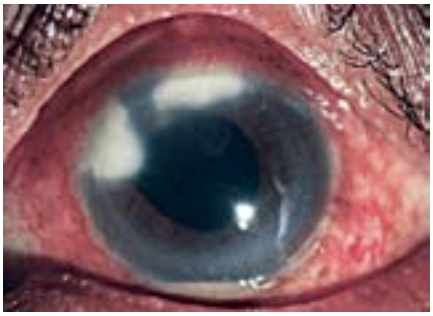


Fig. 3.65.2: Tunnel abscess after phacoemulsification—main port and side port

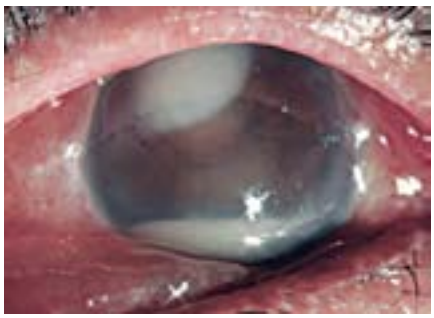


Fig. 3.65.3: Tunnel abscess with endophthalmitis

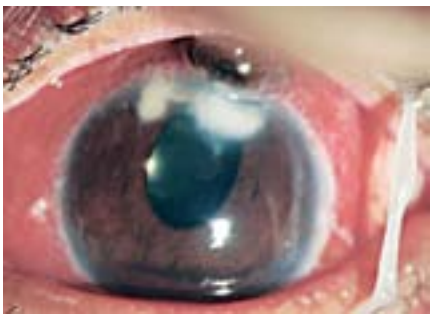


Fig. 3.65.4: Tunnel abscess with iris prolapse

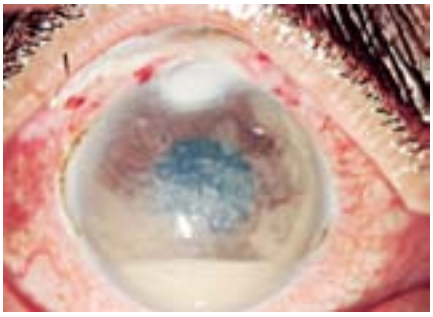


Fig. 3.65.5: Tunnel abscess with gross hypopyon—before tectonic patch graft

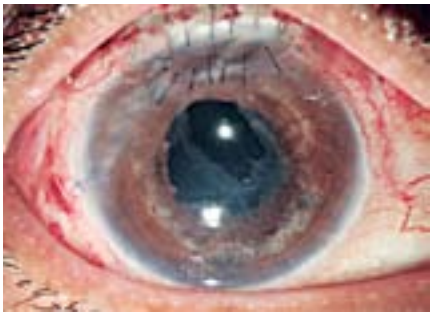


Fig. 3.65.6: Tunnel abscess treated with tectonic graft (same of **Fig. 3.65.5**)

Recurrent Corneal Erosion (RCE)

- It is a condition in which there is disturbance at the level of corneal epithelial basement membrane, resulting in defective adhesions and recurrent breakdowns of the epithelium
- It may occur after minor trauma (finger nail injury) or spontaneously (as in diabetes or dystrophy). Painful blurring of vision, foreign body sensation, photophobia, epithelial defects stained with fluorescein **(Figs 3.66.1A and B)**
- *Treatment:* Topical artificial tears, bandage contact lens (BCL); debridement of epithelium and basement membrane followed by BCL; anterior stromal micropuncture; excimer laser PTK in difficult cases.

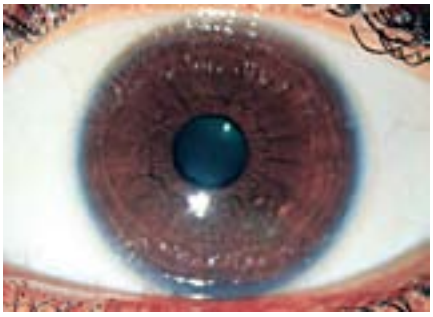


Fig. 3.66.1A: Recurrent corneal erosion

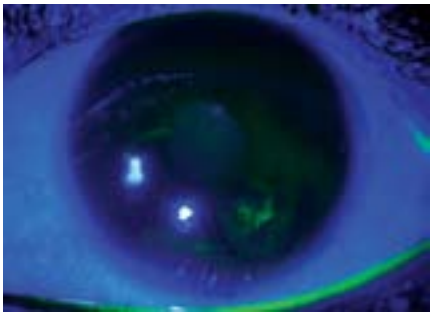


Fig. 3.66.1B: Recurrent corneal erosion—fluorescein stain

Blood Staining of the Cornea

- Occurs after a traumatic hyphema with raised intraocular pressure and if treatment is delayed whole cornea is stained with greenish-brown appearance **(Figs 3.67.1A and B)**. With treatment, cornea slowly clears up from the periphery towards the center and usually takes around two years or more **(Figs 3.67.2A and B)**.
- *Treatment:* May require PK in some cases. Prognosis is always poor.

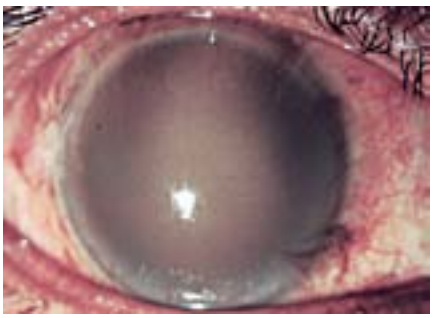


Fig. 3.67.1A: Blood staining of the cornea

Fig. 3.67.1B: Blood staining of the cornea

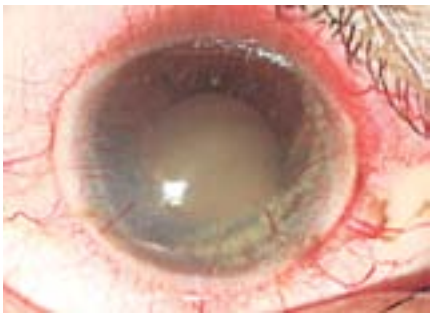


Fig. 3.67.2A: Blood staining of the cornea—clearing from periphery

Fig. 3.67.2B: Blood staining of the cornea—clearing from periphery

Corneal Tumors

- Primary corneal tumors are rare. Mainly they are the extension of the conjunctival or limbal tumors—like, limbal dermoids, dermolipoma, *carcinoma in situ*, invasive squamous cell carcinoma of the conjunctiva, pigmented lesions of the conjunctiva, etc. **(Figs 3.68.1 to 3.68.7)**
- *Treatment:* Excision of the mass with 5 mm healthy conjunctival resection. In neoplastic cases, a double cryo-thaw is required. Additional application of intraoperative mitomycin C (0.02%) for two minutes in recurrent cases; or postoperatively as eye drops (0.04%) 4 days for four weeks. In most of the cases, the prognosis is good. Sectorial lamellar keratoplasty may be required in case of limbal dermoid or dermolipoma.

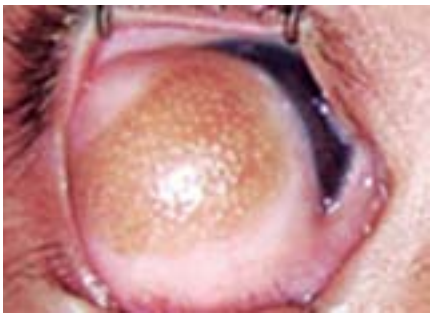


Fig. 3.68.1: Central corneal dermolipoma

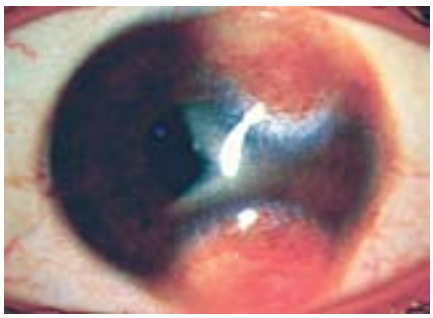


Fig. 3.68.2: Multiple dermolipoma on the cornea

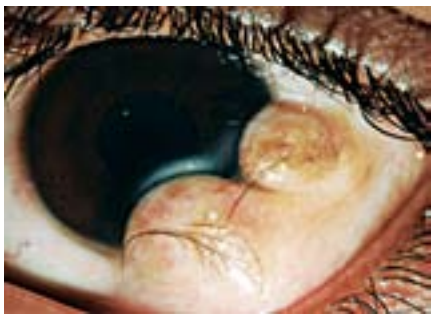


Fig. 3.68.3: Multiple limbal dermoid on the cornea

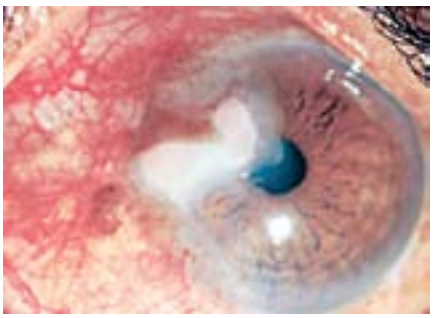


Fig. 3.68.4: Ocular surface squamous neoplasia with corneal involvement

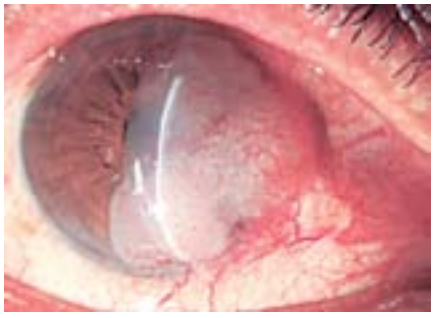


Fig. 3.68.5: Ocular surface squamous neoplasia invoving half of the cornea

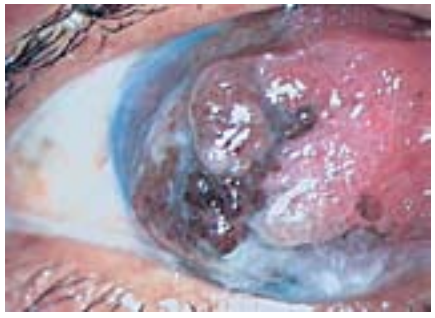


Fig. 3.68.6: Extension of conjunctival squamous cell carcinoma onto the cornea

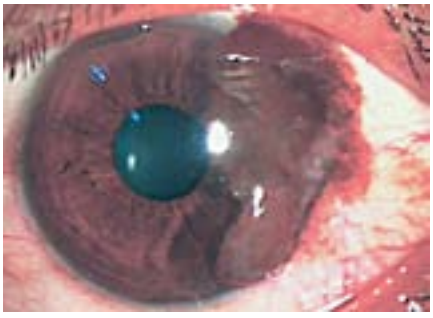


Fig. 3.68.7: Conjunctival melanoma encroaching onto the cornea

Corneal Graft-Related Problems

- A corneal transplantation may be full thickness (penetrating keratoplasty or PK) **(Figs 3.69.1A and B)** or partial thickness (lamellar keratoplasty or LK). In spite of its gold-standard, full thickness keratoplasty has more complications than the others
- A lamellar keratoplasty may be anterior lamellar, deep anterior lamellar keratoplasty (DALK) **(Figs 3.69.2A and B)** or posterior lamellar keratoplasty (PLK)
- Descemet's stripping (automated) endothelial keratoplasty or DSEK/DSAEK (most popular type of PLK) is now the choice of surgery in endothelial dysfunctions **(Fig. 3.69.3)**
- A therapeutic PK is performed in an emergency basis to save the integrity of the globe. The typical example is non-healing corneal ulcer, corneal ulcer with threatened perforation, etc. **(Figs 3.69.4A and B)**
- Tectonic graft is required to support cornea as in corneal thinning, peripheral ulcerative keratitis or Descemetocele **(Figs 3.69.5A and B)**

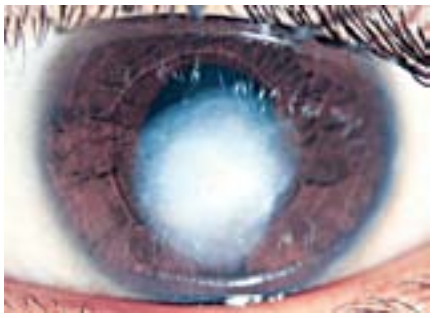


Fig. 3.69.1A: Penetrating keratoplasty—before

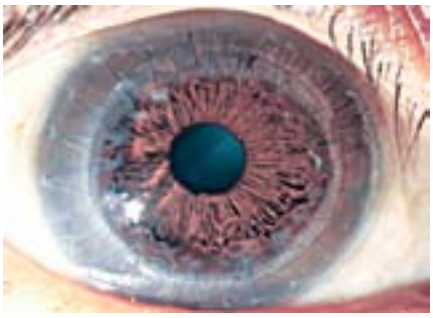


Fig. 3.69.1B: Penetrating keratoplasty—after

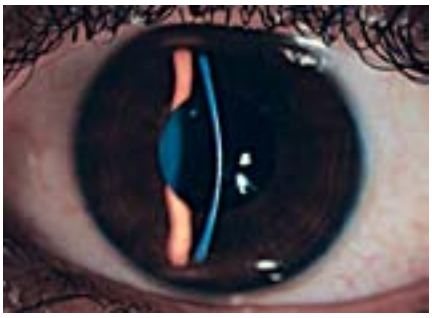


Fig. 3.69.2A: DALK—before in keratoconus

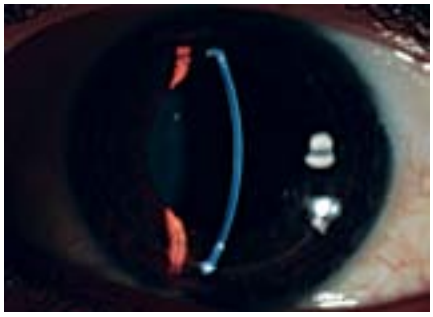


Fig. 3.69.2B: DALK—after in keratoconus

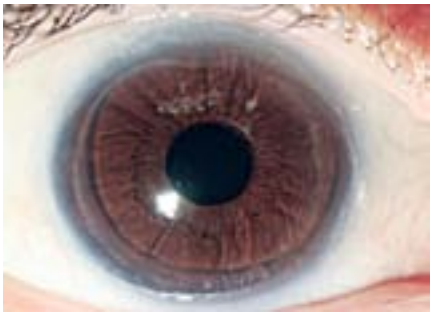


Fig. 3.69.3: Descemet stripping endothelial keratoplasty

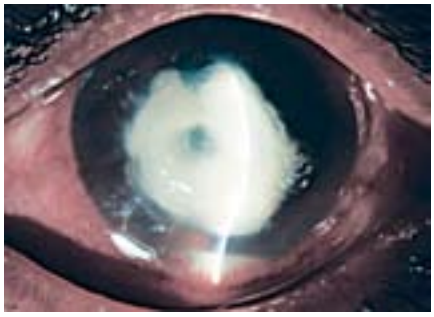


Fig. 3.69.4A: Corneal ulcer—impending perforation

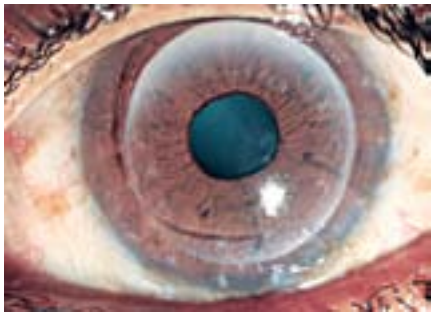


Fig. 3.69.4B: Therapeutic keratoplasty in same patient **(Fig. 3.69.4A)**

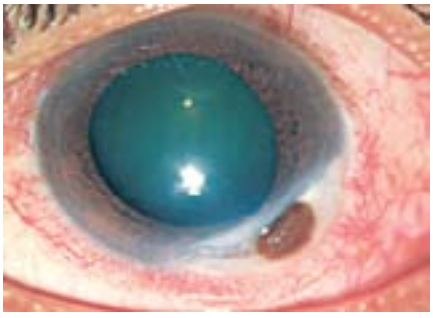


Fig. 3.69.5A: Mooren's ulcer—perforation

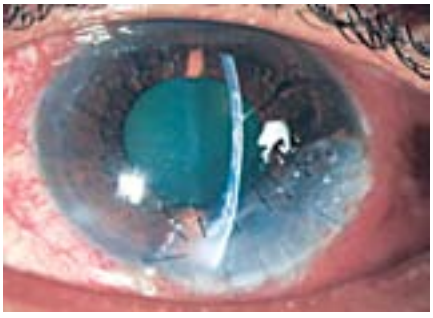


Fig. 3.69.5B: Mooren's ulcer—tectonic graft of same **(Fig. 3.69.5A)**

- Keratoprosthesis is the artificial corneal implants, used in extreme cases where the chance of graft survival is very poor after conventional keratoplasty **(Figs 3.69.6A and B)**. There are many types of keratoprosthesis, like—Boston's keratoprosthesis (KPRO); Osteo-odonto-keratoprosthesis (OOKP) or Pintuchi's keratoprosthesis. Each type has its own indication
- *All types of corneal transplantation surgery including KPRO, have problems which can be easily solved. But, in neglected cases, they may have serious consequences. Here, the follow-ups are much more important than the surgery itself.*

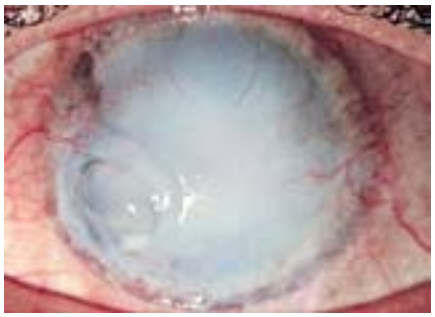


Fig. 3.69.6A: Boston keratoprosthesis—preoperative

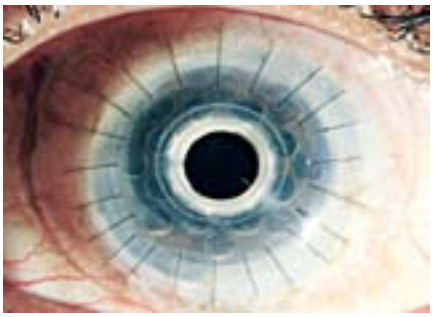


Fig. 3.69.6B: Boston keratoprosthesis—postoperative

Important Corneal Problems/Complications in PK

Primary Donor Failure

- The graft does not clear within 8 weeks (in early postoperative days). It is most commonly occurred in DSEK **(Figs 3.70.1A and B)**. It is edematous and with Descemet folds. It is due to poor donor quality and excessive mishandling the donor tissue during operation
- *Treatment:* Once it is recognized, it should be immediately replaced with a fresh healthy donor corneal button.

Persistent Epithelial Defects

- When epithelial defects do not heal within the first 10 to 14 days with conventional treatment they are called persistent epithelial defects (PEDs) and they are considered one of the most common early postoperative complications after PKP **(Fig. 3.71.1)**. Nonresponsive epithelial defects may also result from herpes simplex virus (HSV) infection, even in patients without a clinical history of HSV **(Figs 3.71.2A and B)**
- *Treatment:* Frequent preservative-free artificial tears and a bandage contact lens (BCL) for 4–6 weeks usually solve the problem. It may require a temporary tarsorrhaphy.

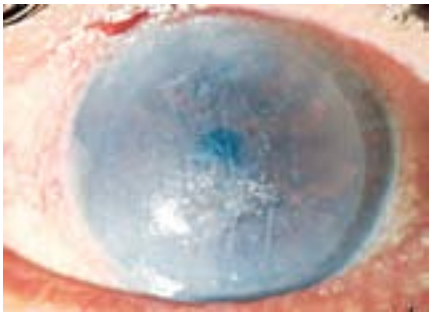


Fig. 3.70.1A: Primary donor failure in DSEK

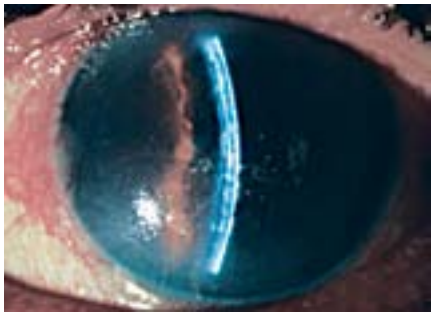


Fig. 3.70.1B: Primary donor failure in DSEK in slit section

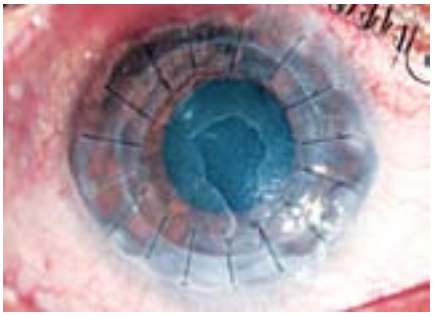


Fig. 3.71.1: Persistent epithelial defect

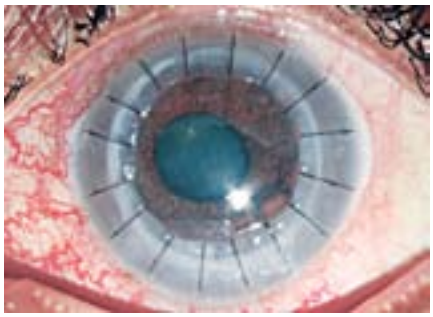


Fig. 3.71.2A: Persistent epithelial defect

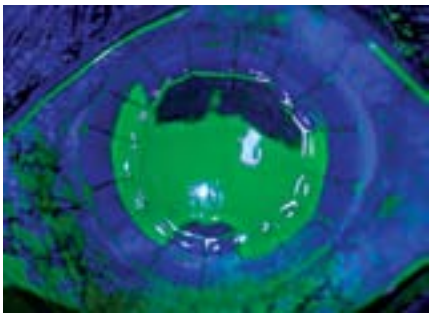


Fig. 3.71.2B: Persistent epithelial defect—fluorescein staining

Loose Sutures and Broken Sutures

- Protruding suture ends which may irritate the eye and sometimes with giant papillary conjunctivitis (GPC); vascularization along the sutures; loose sutures **(Figs 3.72.1 to 3.72.3)**; broken sutures and infiltration at the suture ends **(Fig. 3.72.4)**, suture abscess **(Figs 3.72.5 and 3.72.6)** or sometimes with frank infection which may be bacterial or fungal with or without hypopyon **(Fig. 3.72.7)**
- Broken suture may present with wind shield wiper phenomenon **(Figs 3.72.8A and B)**
- *Treatment:* All the offending sutures are to be removed under sterile precautions. In noninfective cases, the steroid drops frequency is to be increased for a short time and antibiotics drops 4 times for few days.
- In infective cases, remove the suture by pulling through the shortest possible route. Smear preparation to detect bacteria or fungus; and culture of the sutures and treat it intensively with topical fluoroquinolones/fortified antibiotics or antifungal as given in microbial keratitis.

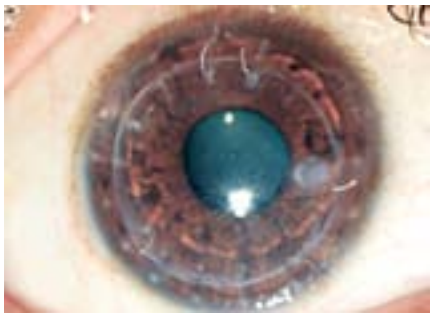


Fig. 3.72.1: Loose sutures

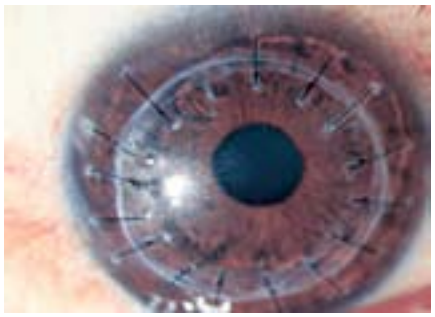


Fig. 3.72.2: Loose sutures with infiltration at 10-0 o'clock position

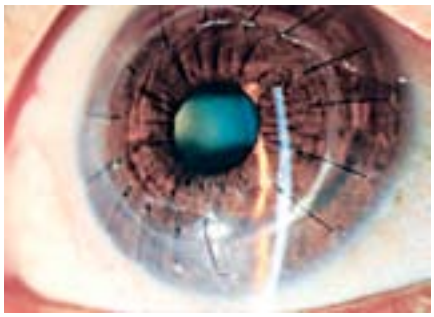


Fig. 3.72.3: Loose suture with vascularization at 6-0 o'clock position

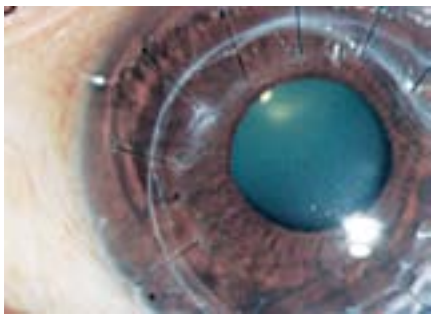


Fig. 3.72.4: Broken suture with small infiltration at 10-0 o'clock position

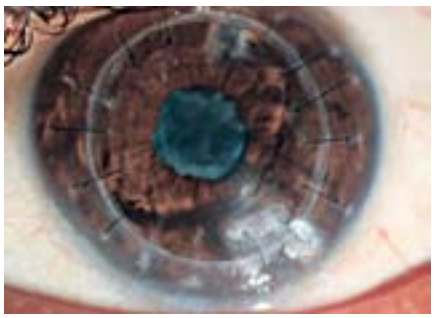


Fig. 3.72.5: Loose suture with fungal infiltration at 5-0 o'clock position

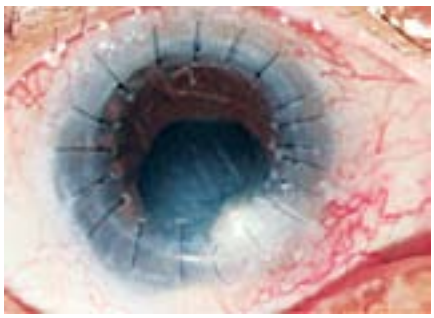


Fig. 3.72.6: Loose suture with deep suture abscess with hypopyon

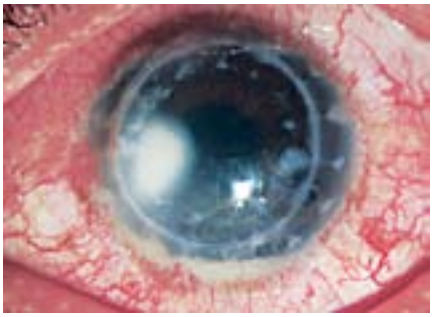


Fig. 3.72.7: Broken suture—graft infection with hypopyon

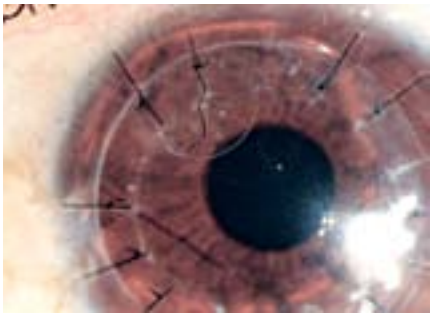


Fig. 3.72.8A: Broken suture with windshield wiper phenomenon

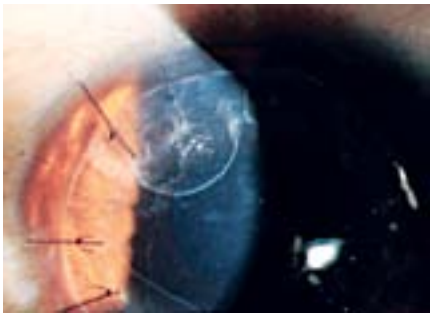


Fig. 3.72.8B: Broken suture with windshield wiper phenomenon

Early Graft Infection

- It may be bacterial or fungal and seen within 1–3 weeks of transplantation. It is sometimes associated with frank endophthalmitis **(Figs 3.73.1 and 3.73.2)**. Primarily occurs due to contaminated donor tissue. The picture is similar to that of bacterial or fungal keratitis. It is most common in therapeutic graft. Early interface infection may occur after DALK **(Figs 3.73.3A and B)** or DSEK procedures **(Figs 3.73.4A and B)**
- *Treatment:* Aggressive antimicrobial therapy. Donor-button culture reports are helpful to determine the type of infection and initiation of therapy. Urgent regrafting is required in many cases with a fresh donor button.

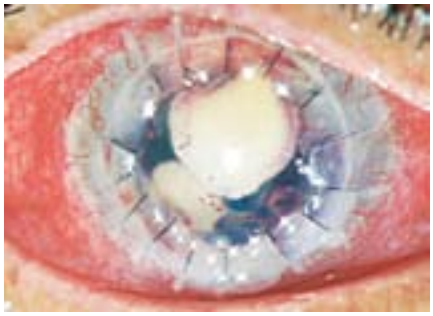


Fig. 3.73.1: Early postoperative graft infection (fungal) after PK

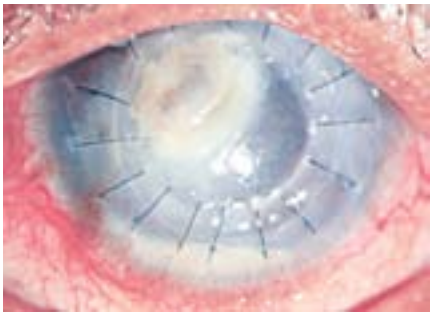


Fig. 3.73.2: Early graft infection—fungal—after therapeutic PK

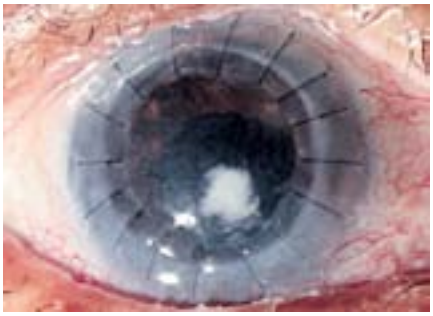


Fig. 3.73.3A: Interface infection in DALK

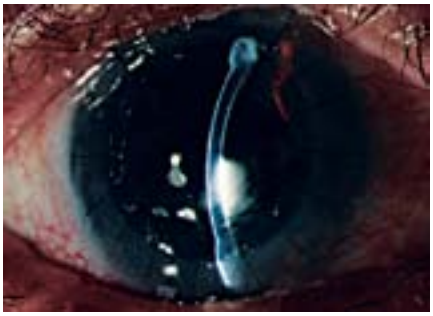


Fig. 3.73.3B: Interface infection in DALK

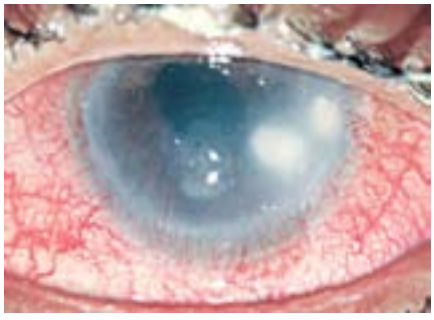


Fig. 3.73.4A: Interface infection in DSEK

Fig. 3.73.4B: Interface infection in DSEK

Recurrence of Previous Diseases in Graft

- *The important diseases are:* HSV keratitis, lattice dystrophy, granular dystrophy, Reis-Buckler's dystrophy, etc. **(Figs 3.74.1 and 3.74.2)**
- *Treatment* may be required in some cases. HSV keratitis is to be treated medically as per protocol. Tab acyclovir (400 mg) twice daily for 6 months to 1 year may reduce recurrence rate.

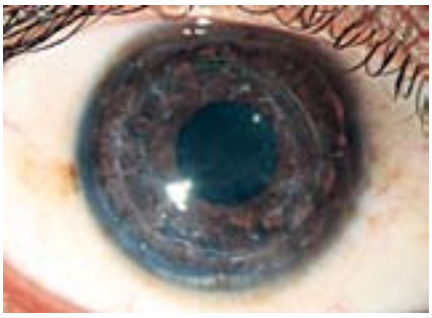


Fig. 3.74.1: Recurrence of primary disease—Reis-Buckler's dystrophy after DALK

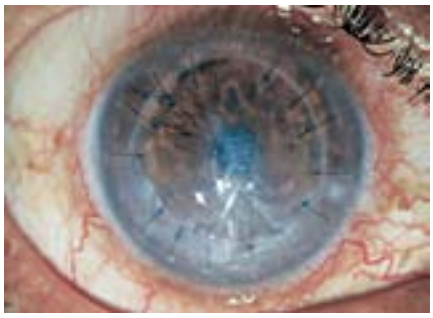


Fig. 3.74.2A: Recurrence of previous diseases in graft—post-HSV keratitis

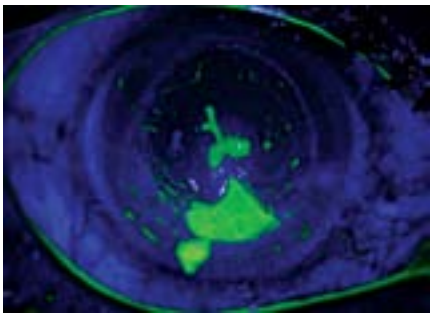


Fig. 3.74.2B: Recurrence of previous diseases in graft—post-HSV keratitis

Induced High and Irregular Astigmatism

- In spite of crystal clear graft, some patients may not gain good vision due to high and irregular astigmatism. It is due to suturing problem **(Figs 3.75.1A and B)**. Refraction (abnormal shadow), keratometry and corneal topography can detect this condition
- *Treatment:* Topography-guided selective suture removal, RGP contact lens can improve the vision. Post-PK LASIK is required in some cases to reduce the astigmatism.

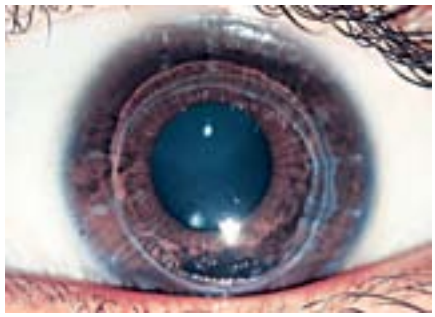


Fig. 3.75.1A: Induced high and irregular astigmatism

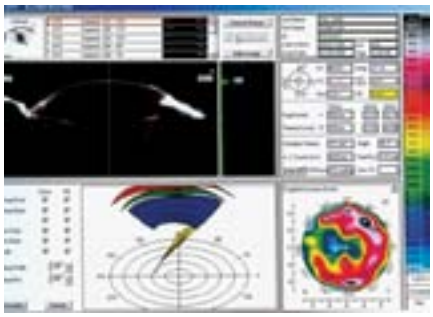


Fig. 3.75.1B: Induced high and irregular astigmatism—corneal topography

Late PK—Wound Dehiscence

- Keratoplasty wound never heals. Late dehiscence of wound may occur after a minor trauma in the eye or sometimes even after a forcible slap on cheek several years after the surgery **(Figs 3.76.1 to 3.76.3)**. In full-thickness graft, the stroma never attaches completely with the host. Attachments only occur at

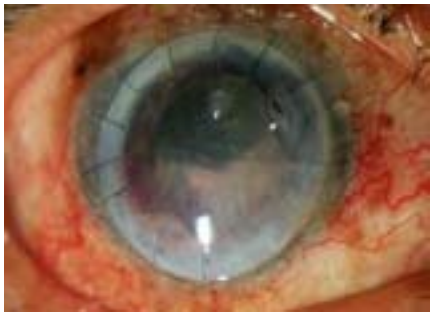


Fig. 3.76.1: Late wound dehiscence in PK after 1 year

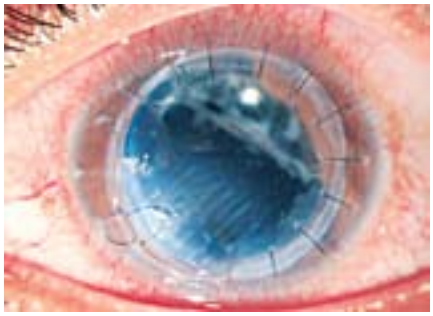


Fig. 3.76.2: Late wound dehiscence in PK after 3 years

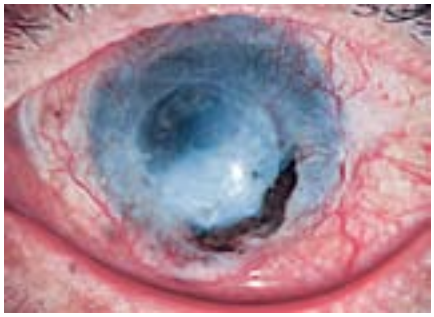


Fig. 3.76.3: Late wound dehiscence in PK even after 8 years

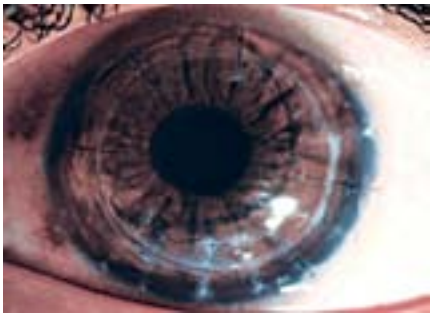


Fig. 3.76.4A: Late wound dehiscence in DALK after 1 year

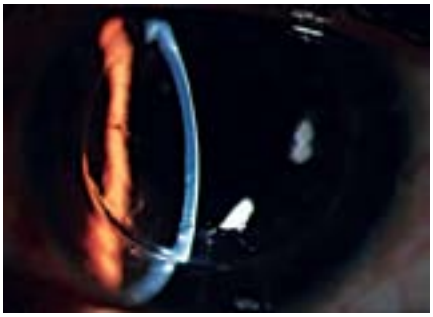


Fig. 3.76.4B: Late wound dehiscence in DALK after 1 year

the epithelium by proliferation and endothelial layer by sliding. Wound dehiscence also occurs in DALK with minor trauma **(Figs 3.76.4A and B)**

- *Treatment:* Emergency resuturing of the wound as far as possible.

Late Graft Infection

- It may occur lately due to surface or suture-related problem. The keratitis may be due to bacterial or fungal. In fungal infection, *Candida* is the most common organism **(Fig. 3.77.1 to 3.77.4)**
- *Treatment:* Intensive antimicrobial therapy after scraping and smear preparation. Culture reports are important to change the antimicrobial agent. In severe cases, a regrafting procedure is required with a fresh donor cornea.

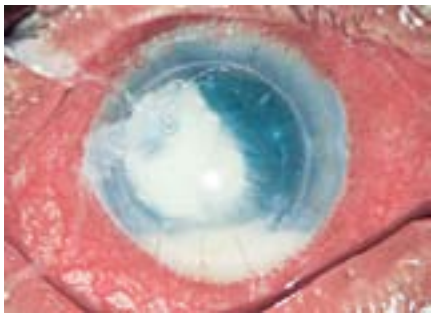


Fig. 3.77.1: Late graft infection in PK—bacterial

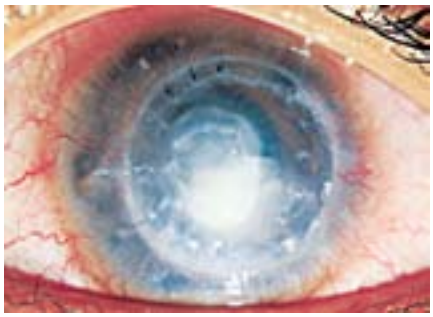


Fig. 3.77.2: Late graft infection in PK—bacterial

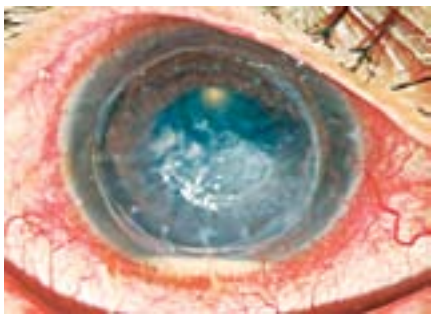


Fig. 3.77.3: Late graft infection in PK—fungal

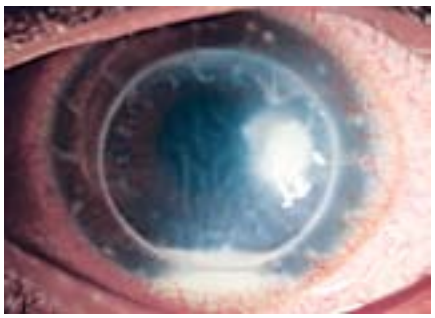


Fig. 3.77.4A: Late graft infection in DALK—double hypopyon

Fig. 3.77.4B: Late graft infection in DALK—double hypopyon

Allograft Rejection

- Corneal allograft rejection of any layer can occur following PK and less frequently in LK procedures. Endothelial rejection is most common and most serious as it may lead to severe endothelial cell loss followed by decompensation. Epithelial and stromal rejections are less frequent and less serious. They are often asymptomatic and diagnosed on routine follow-up examination.

- ***Epithelial rejection:*** Circumciliary congestion accompanied by irregular elevated line of abnormal epithelium, often called epithelial rejection line.
- ***Stromal rejection:*** Localized stromal neovascularization, subepithelial infiltrates, surrounding stromal edema, multiple white spots in anterior stroma (Krachmer's spot) in the donor cornea **(Figs 3.78.1A and B)**
- *Treatment:* Increase frequency of topical steroids and topical cycloplegic are sufficient to control both stromal and epithelial rejections.

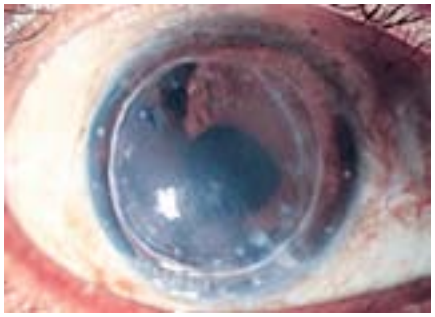


Fig. 3.78.1A: Graft rejection—stromal

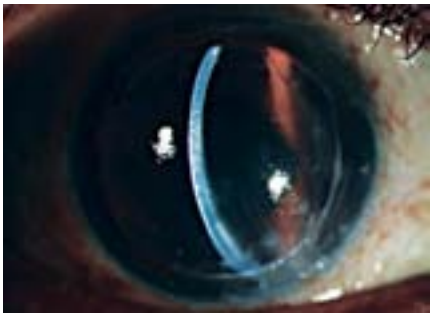


Fig. 3.78.1B: Graft rejection—stromal

Endothelial Rejection

- Photophobia and watering is present, but no discharge. Ciliary congestion, anterior chamber reaction, new keratic precipitates (KPs), often these KPs deposit in a characteristic linear pattern on the corneal endothelium, called Khodadoust's line (endothelial rejection line) **(Figs 3.78.2 and 3.78.3)**. There are associated areas of inflammation at the graft margin. Associated stromal edema is an indication of graft failure. Endothelial rejection also occur in DSEK **(Figs 3.78.4A and B)**
- *Treatment:* It is an emergency situation and intensive treatment may reverse the rejection episode. Single dose intravenous injection methyl prednisolone (500–1000 mg) is given, followed by oral prednisolone 1 mg/kg/day in tapering doses; topical prednisolone acetate 1 hourly for 24 hours, then in tapering doses; topical cycloplegics and control IOP **(Figs 3.78.5 and 3.78.6)**

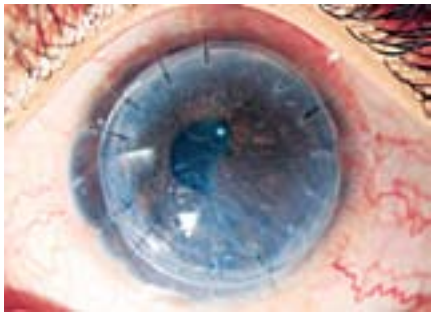


Fig. 3.78.2A: Graft rejection—endothelial

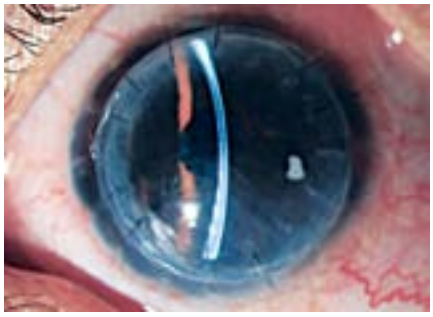


Fig. 3.78.2B: Graft rejection—endothelial

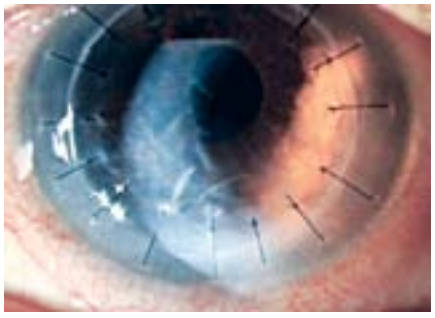


Fig. 3.78.3: Graft rejection—endothelial—Khodadoust's line

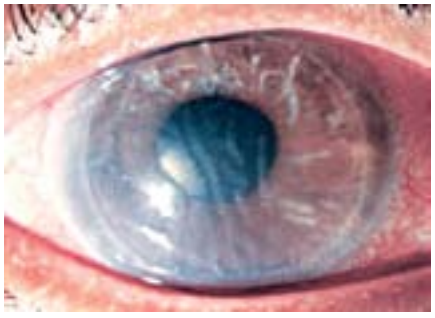


Fig. 3.78.4A: Endothelial graft rejection in DSEK

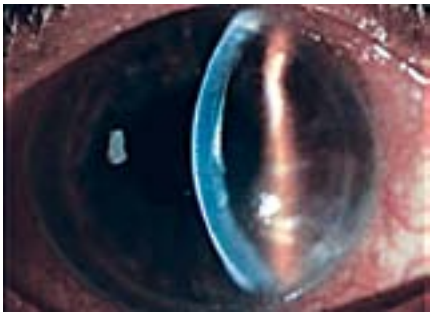


Fig. 3.78.4B: Endothelial graft rejection in DSEK

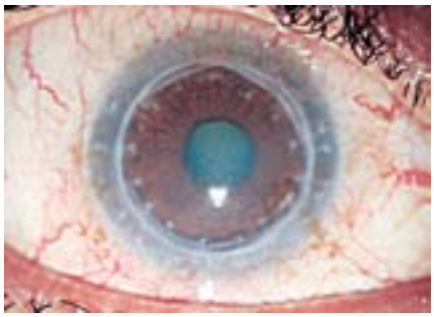


Fig. 3.78.5A: Graft rejection in PK—before treatment

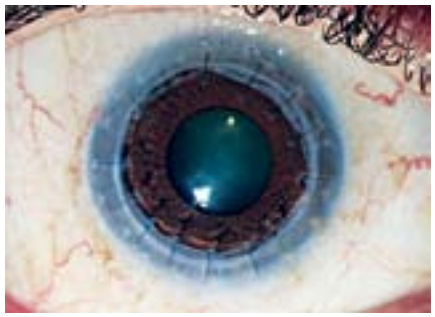


Fig. 3.78.5B: Endothelial graft rejection in PK after medical treatment

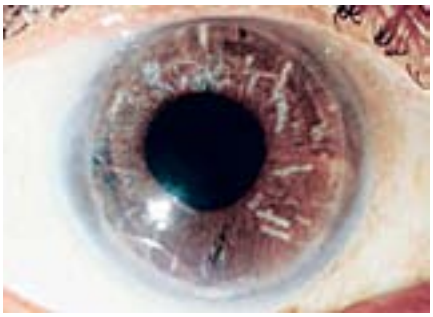


Fig. 3.78.6: Endothelial graft rejection in DSEK after medical treatment (same as **Fig. 3.78.4A**)

- Topical cyclosporine 1 to 2%—twice to four times daily may be helpful in the treatment or prevention of graft rejection in high-risk cases.

Late Donor Failure

- This occurs after many months or years due to gradual decompensation of donor corneal endothelium and without any previous complication which may cause endothelial insult. The endothelial cell loss in PK is 2% per year as compared to normal endothelium which is only 0.5% per year **(Fig. 3.79.1)**. Critical endothelial cell count is around 400-500 cells/sq mm, below which cornea starts decompensating
- In long standing cases, the graft becomes opaque and severe vascularization **(Figs 3.79.2 to 3.79.4)** may occur
- *Treatment:* Regrafting with a healthy donor cornea. In many cases, DSEK is also possible with better results. For vascularized failed graft, it is better not to regraft. But, for one eyed cases, Boston keratoprosthesis—Type 1 may be an answer.

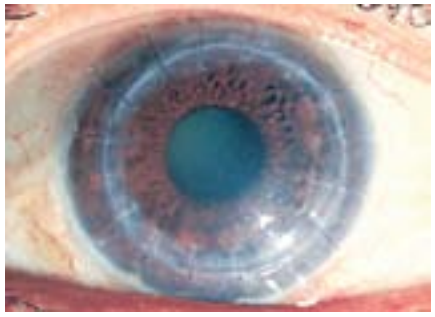


Fig. 3.79.1: Late donor failure in PK

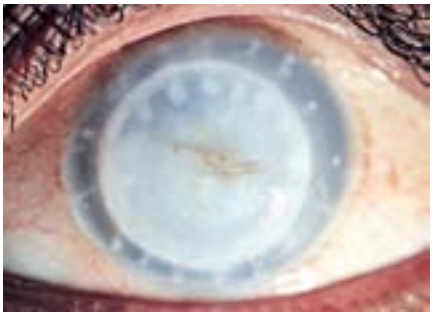


Fig. 3.79.2: Failed opaque graft in PK

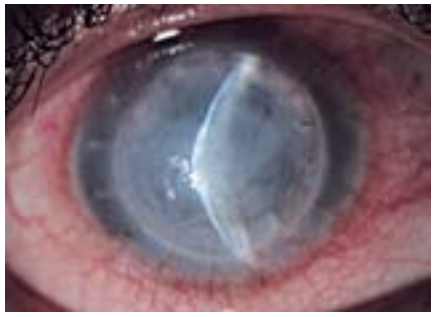


Fig. 3.79.3: Vascularized failed graft in PK

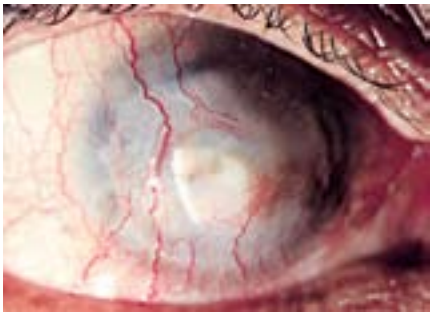


Fig. 3.79.4: Failed opaque graft with severe vascularization

Donor Dislocation in DSEK

- It is a unique complication of DSEK/DSAEK, where the donor button is dislocated within the anterior chamber. It typically happens within 1-7 days following DSEK. Both the host cornea and donor corneal button appears edematous **(Figs 3.80.1A and B)**. It is more common with ACIOL, aphakia or Post-PK cases
- *Treatment:* Urgent rebubbling with air is necessary to reattach the donor button against recipient corneal stroma **(Fig. 3.80.2)**. If it fails, consider for Re-DSEK or PK.

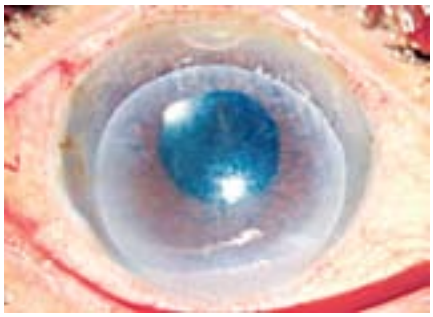


Fig. 3.80.1A: Donor dislocation in DSEK

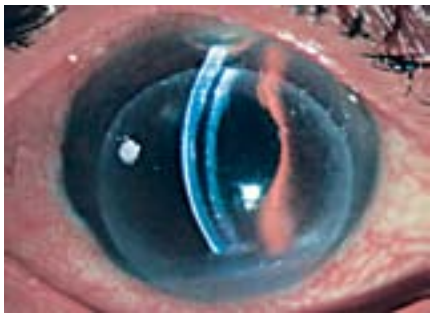


Fig. 3.80.1B: Donor dislocation in DSEK in silt section

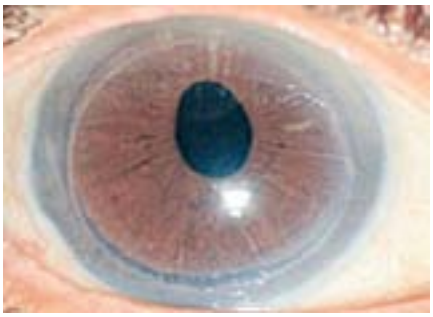


Fig. 3.80.2: Reattachment in donor button after rebubbling (same eye)

Double Anterior Chamber in DALK

- This is also a unique complication of deep anterior lamellar keratoplasty (DALK) where there is non-attachment of recipient Descemet's membrane with donor cornea. This usually occurs within 1–3 days following DALK. Clinically the anterior chamber appears double, first one is in between donor posterior surface and recipient Descemet's membrane, and the second one is between recipient endothelium and iris-lens diaphragm **(Fig. 3.81.1)**
- *Treatment:* Air bubble tamponade or C_3F_8 gas and try to keep that for few days. IOP is to be checked during the follow-up period.

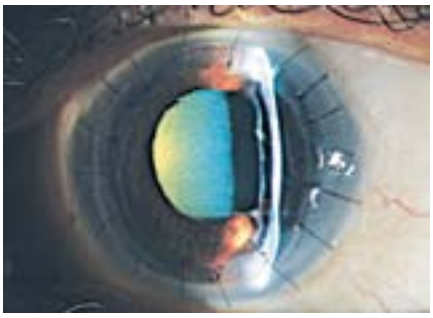


Fig. 3.81.1: Double anterior chamber in DALK

Late Interface Scarring in LK

- This may happen as a late complication in ALK **(Fig. 3.82.1)** and DSEK **(Fig. 3.82.2A and B)**, but very rarely in DALK. This is due to scarring at the graft-host interface; and it may reduce some amount of vision. Interface haze is seen under slit lamp in focal oblique (parallelopiped) illumination
- *Treatment:* If the vision is within acceptable limit, no intervention is required. Otherwise a PK is the treatment of choice.

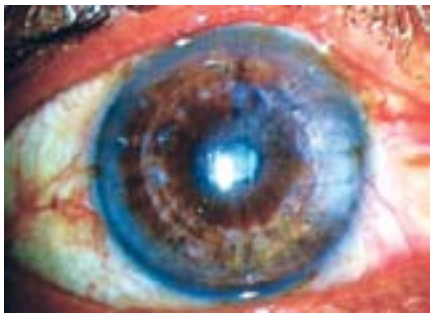


Fig. 3.82.1: Late interface scarring in ALK

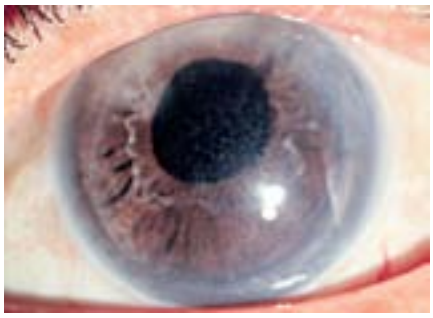


Fig. 3.82.2A: Late interface scarring in DSEK

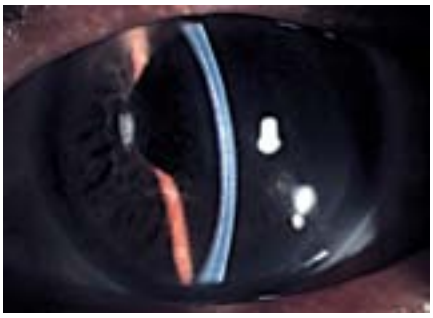


Fig. 3.82.2B: Late interface scarring in DSEK

Epithelial Ingrowth in DSEK

- This is a rare complication of DSEK. Usually mid peripheral where venting (stab) incision is given during surgery to drain fluid from the interface **(Figs 3.82.3A to C)**
- *Treatment:* To scoop out the epithelial nests from a sideport after negotiating through the interface.

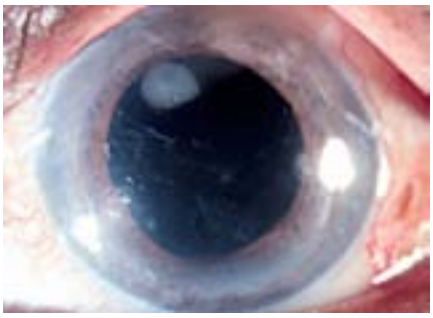


Fig. 3.82.3A: Epithelial ingrowth following DSEK

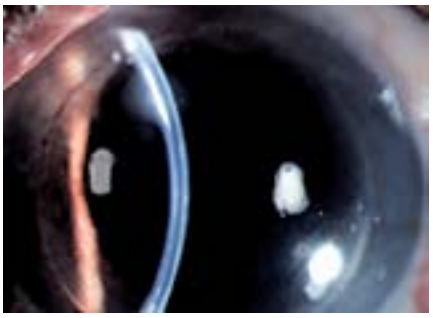


Fig. 3.82.3B: Epithelial ingrowth following DSEK

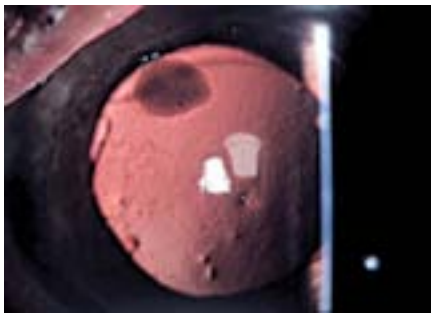


Fig. 3.82.3C: Epithelial ingrowth following DSEK in retroillumination

Late Secondary Glaucoma

- Late secondary glaucoma may occur in any type of corneal grafting. Primarily due to steroid induced glaucoma due in poor follow-up. Other causes include—peripheral anterior synechiae, suture induced, vitreous in anterior chamber, ICE syndrome, etc. Patients often presents late with decreased vision, graft edema and with epithelial bedewing **(Figs 3.83.1A and B)**.
- *Treatment:* Careful monitoring of IOP is a must for all cases of keratoplasty, either PK, DALK or DSEK. Timely shift to soft steroids is important. Antiglaucoma medication is used routinely. In severe cases, trabeculectomy with MMC is required or in recalcitrant cases glaucoma valve operation may be done.

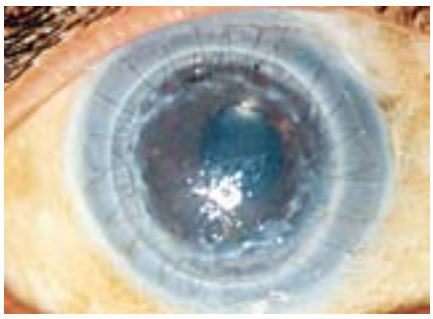


Fig. 3.83.1A: Late secondary glaucoma with graft edema

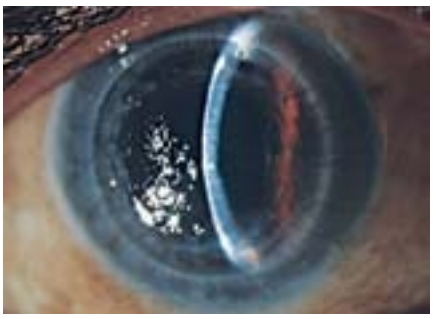


Fig. 3.83.1B: Late secondary glaucoma—epithelial bedewing

Refractive Surgery-Related Corneal Problems

Late Rupture of RK Wound

- Old radial keratotomy wound may rupture easily even with minor trauma. Multiple linear rupture of the cornea along the radial cuts and it gives multiple wedge-shaped corneal parts attached to the limbus. It may be associated with iris prolapse **(Figs 3.84.1A and B)**

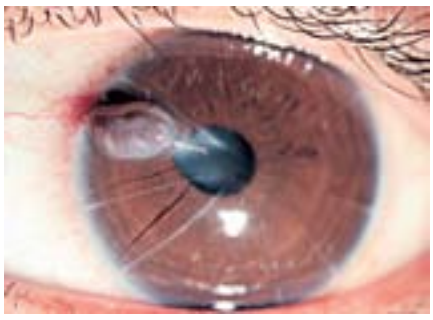


Fig. 3.84.1A: RK rupture with iris prolapse

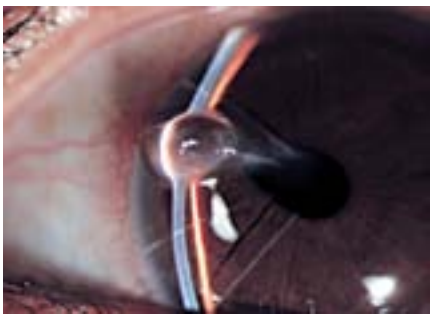


Fig. 3.84.1B: RK rupture with iris prolapse and shallow AC

- Sometimes, it may be associated with sight-threatening corneal abscess and endophthalmitis **(Fig. 3.84.2)**
- *Treatment:* Urgent repair with 10-0 nylon **(Fig. 3.84.3)**. But it is always very difficult.

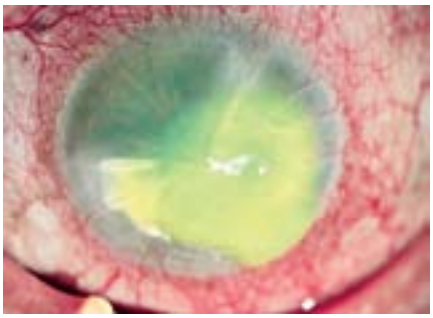


Fig. 3.84.2: RK rupture with endophthalmitis

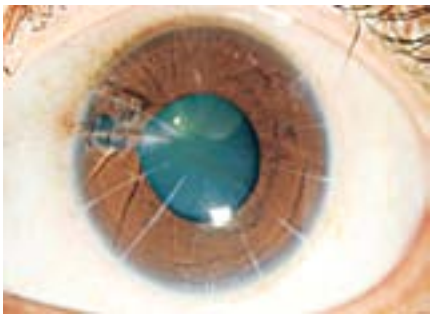


Fig. 3.84.3: RK rupture—repair of the same eye of **Figure 3.84.1**

Post-PRK Corneal Haze

- Corneal scarring (haze) may be seen in some cases after 2 weeks to several months **(Figs 3.85.1A and B)**
- It is associated with irregular astigmatism. Late post-PRK subepithelial scarring is also a problem in same cases **(Figs 3.85.2A and B)**
- Raised IOP may be a concern
- Late central corneal haze is also a problem with old-fashioned extensive RK **(Figs 3.86.1A and B)**
- *Treatment:* Increase frequency of steroid drops. Severe haze may require PTK with or without mitomycin C.

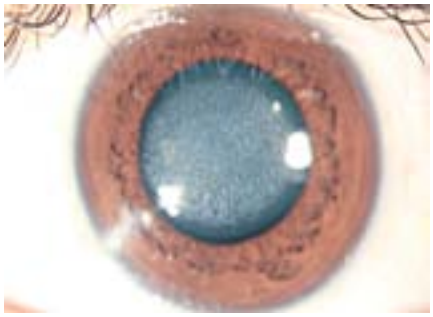


Fig. 3.85.1A: Post-PRK haze

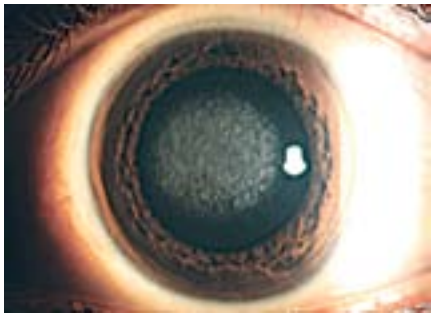


Fig. 3.85.1B: Post-PRK haze

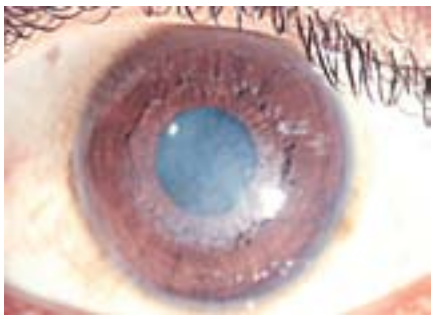


Fig. 3.85.2A: Late PRK induced fibrosis

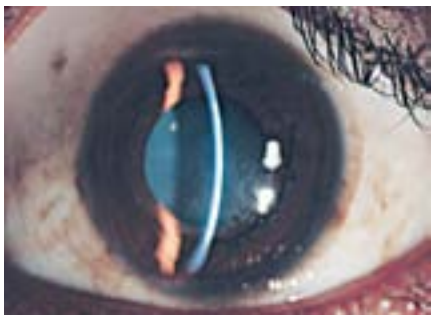


Fig. 3.85.2B: Late PRK induced fibrosis

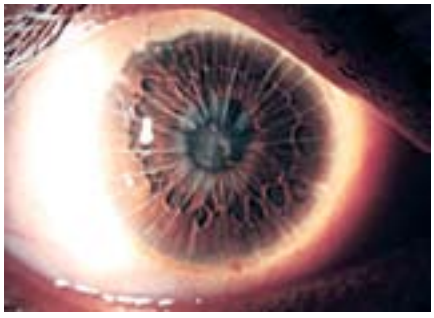


Fig. 3.86.1A: RK marks—extensive with central scarring

Fig. 3.86.1B: RK marks—extensive with central scarring

Wrinkling of the Flap

- It may be seen in early weeks. Multiple flap folds are seen in a concentric manner **(Figs 3.87.1A and B)**. Visual acuity may reduce
- *Treatment:* Immediately lift the flap and refloat within 24 hours. Sometimes suturing may be required.

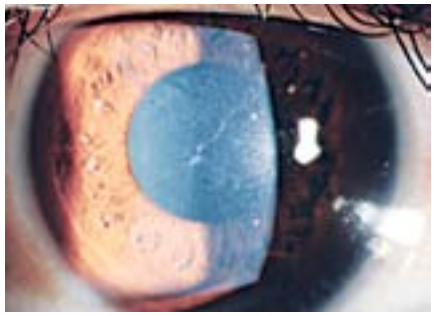


Fig. 3.87.1A: Wrinkled LASIK flap

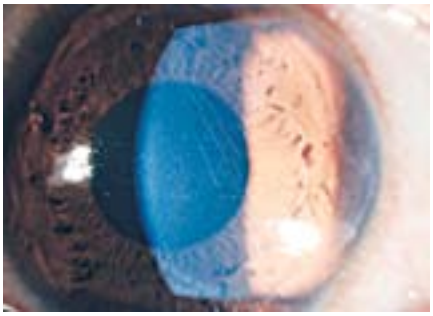


Fig. 3.87.1B: Wrinkled LASIK flap

Diffuse Lamellar Keratitis (DLK)

- It is also known as "sands of Sahara" because of its appearance. It may develop within 3–7 days following LASIK and appears as multiple fine granular inflammatory infiltrates in the flap interface. Best visible by sclerotic scatter method **(Figs 3.88.1A and B)**
- *Treatment:* Intensive treatment with frequent topical steroids with topical antibiotic cover. In severe cases, the interface may be irrigated with BSS after lifting the flap.

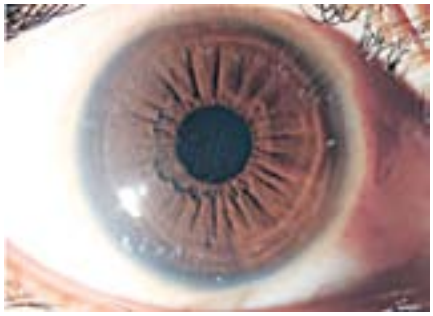


Fig. 3.88.1A: DLK after LASIK—sands of Sahara

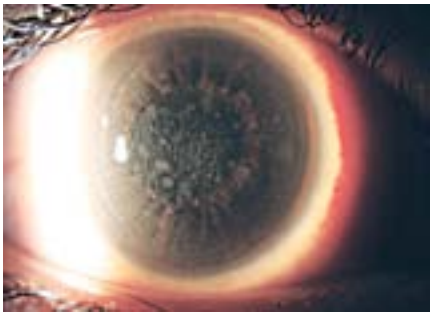


Fig. 3.88.1B: DLK after LASIK—sands of Sahara in sclerotic scatter

Epithelial Ingrowth

- Epithelial ingrowth may occur after an epithelial defect **(Figs 3.89.1A and B)**. Vision may be affected in case of central lesion.
- *Treatment:* Observation, especially if the lesion is peripheral and not affecting the vision. Surgical debridement is required if it is dense and involving the visual axis.

Fig. 3.89.1A: LASIK complication—epithelial ingrowth

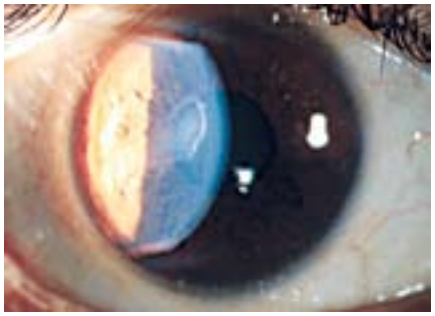


Fig. 3.89.1B: LASIK complication—epithelial ingrowth

LASIK Flap Displacement

- Another important complication in early postoperative period within first week and the patient may present with dimness of vision

- The flap gets displaced on at one side depending upon the hinge **(Figs 3.90.1A and B)**. There may be associated wrinkling
- *Treatment:* Similar to wrinkling of flap—immediately lift the flap and refloat within 24 hours. Sometimes suturing may be required.

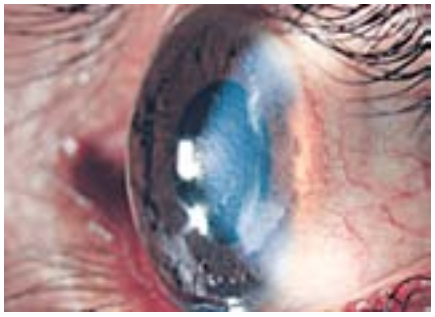


Fig. 3.90.1A: LASIK complication—flap displacement

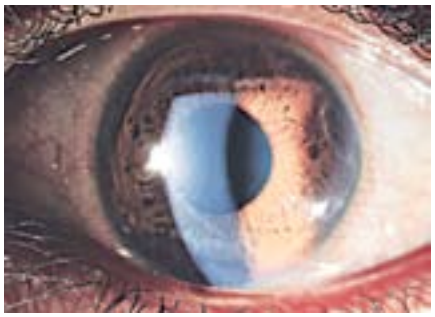


Fig. 3.90.1B: LASIK complication—flap displacement

Post-LASIK Ectasia

- Another very important complication and the patient presents with secondary keratoconus
- It may occur any time after 3 months of LASIK surgery. The thinning is more and affects a wider area **(Figs 3.91.1A and B)**. Sometime, the thinning and ectasia may be very severe **(Fig. 3.91.2)**
- *Treatment:* Collagen cross-linking with riboflavin may not be possible in most of the cases because of more thinning than keratoconus. Deep anterior lamellar keratoplasty (DALK) may be considered first, otherwise penetrating keratoplasty.

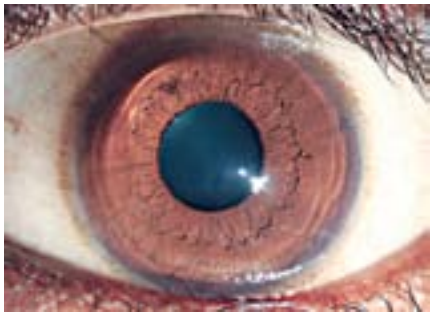


Fig. 3.91.1A: Post-LASIK ectasia

Fig. 3.91.1B: Post-LASIK ectasia

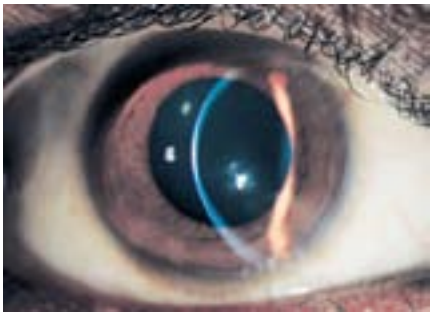


Fig. 3.91.2: Post-LASIK ectasia—severe

Post-LASIK Dry Eye

- It is seen after 6 weeks to 6 months and it is almost universal because of neurotropic elements. A fluorescein and rose bengal stain may be positive in many cases **(Figs 3.92.1 and 3.92.2)**
- *Treatment:* Preservative-free tear lubricants for a long time. Cyclosporine eye drop (0.05%) twice daily is required in moderate to severe cases for at least 6 months to 1 year.

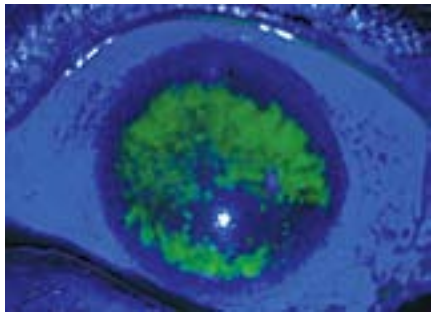


Fig. 3.92.1: Post-LASIK dry eye—fluorescein staining

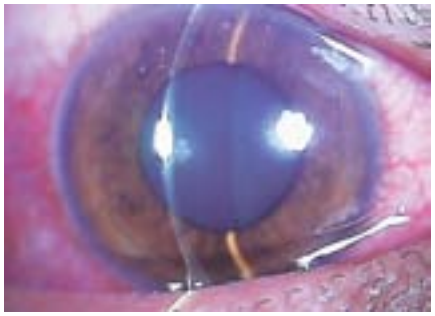


Fig. 3.92.2: Post-LASIK dry eye—rose bengal staining

Corneal Ulcer and/or Interface Infection

- Usually bacterial, may be bilateral in some cases. The picture is similar as bacterial keratitis **(Fig. 3.93.1)**. The common microorganisms responsible are—atypical *Mycobacteria*, *Nocardia*, etc.
- *Treatment:* Urgent lifting of the flap and irrigation with vancomycin and amikacin. In severe cases the flap may be amputed followed by intensive antibiotics drop after microbiological evaluation. 4th generation fluoroquinolones are more potent for atypical mycobacteria.

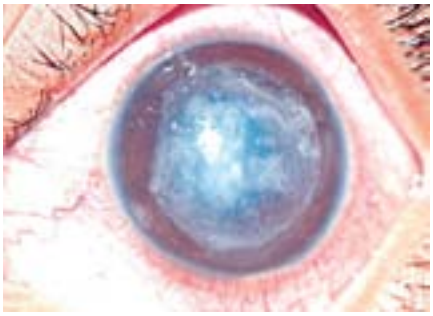


Fig. 3.93.1: Post-LASIK infectious keratitis—*Nocardia*

Recurrence of Original Disease after PTK

- For superficial lesions, like BSK, granular dystrophy, Avellino dystrophy, PTK is indicated by some surgeons. But, in some cases, the original disease may recur **(*see* Figs 3.44.8A and B)**.
- *Treatment:* Deep anterior lamellar keratoplasty (DALK) is indicated in such cases.

Late Flap Displacement or Lost Flap

- Lasik flap never heals. There may be detachment or loss of LASIK flap with minor trauma or rubbing of the eyes several years after LASIK procedure. This situation may similarly occur during VR procedures if it is required later on.
- *Treatment:* Urgent anterior lamellar therapeutic keratoplasty.

Contact Lens-Related Corneal Problems

- Any contact lens wearer with photophobia, pain, redness, discharge or watering should immediately discontinue the use of lens and have a through eye examination as early as possible. ***The important problems are:***

Superficial Punctate Keratitis

- Most common complication
- Fluorescein staining pattern may be an important clue for its detection **(Figs 3.94.1A and B)**
- *Treatment:* Frequent tears substitute; temporary withdrawal of CL or reduce lens wearing time; may require change of CL.

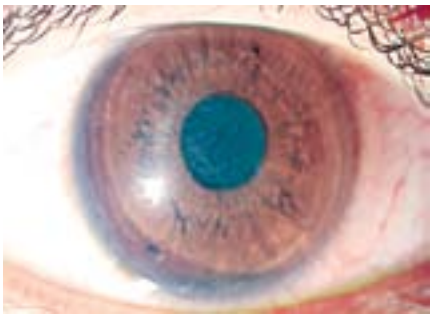


Fig. 3.94.1A: Contact lens induced SPK

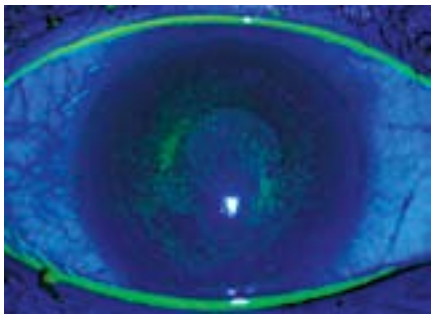


Fig. 3.94.1B: Contact lens induced SPK

Contact Lens—Immune Response Keratitis

- CLARE or contact lens related acute red eye may be associated with marginal infiltrates on the cornea. May be the result of sensitivity reaction to bacterial toxins and similar to marginal keratitis associated with blepharitis **(Figs 3.95.1 and 3.95.2)**
- *Treatment:* Discontinue CL wear; antibiotics and steroids eye drops and lens hygiene.

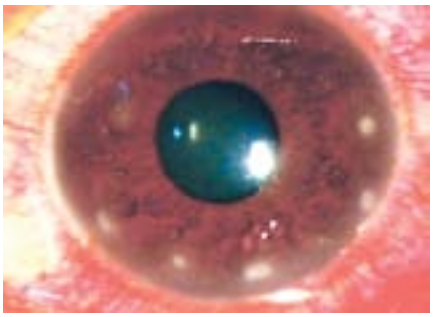


Fig. 3.95.1: Contact lens—immune response keratitis

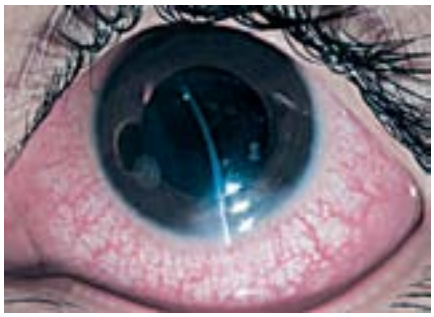


Fig. 3.95.2: Contact lens—immune response keratitis

Acute Hypoxia of Cornea

- Epithelial microcysts formation with necrosis and small endothelial blebs **(Figs 3.96.1A and B)**
- Microerosions can be demonstrated by fluorescein staining. CL overwear may be the cause
- *Treatment:* Reduce wearing time drastically, higher DK-value CL and frequent tear substitutes.

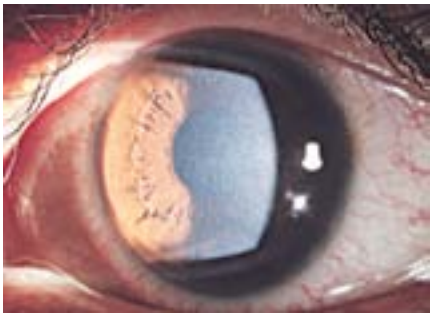


Fig. 3.96.1A: Acute hypoxia of cornea

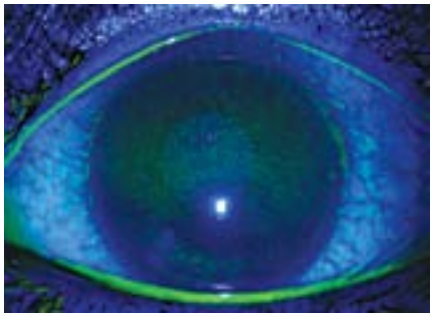


Fig. 3.96.1B: Acute hypoxia of cornea

Tight Lens Syndrome

- Usually develops within 1–2 days of use of CL usually with a soft lens. No lens movement with blinking and CL appears 'sucked-on' to the cornea
- Staining of the conjunctival epithelium as a ring around cornea **(Fig. 3.97.1)**; may be associated with central corneal edema, SPKs and anterior chamber reaction
- *Treatment:* Discontinue CL wear; cycloplegic; refit the patient with flatter lens.

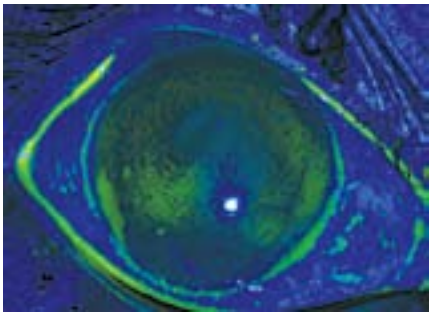


Fig. 3.97.1: Tight lens syndrome

Corneal Neovascularization

- Superficial corneal vascularization more than 1 mm inside limbus **(Figs 3.98.1A and B)**
- Chronic hypoxia is the main cause due to overuse or may be seen in extended wear lenses
- *Treatment:* Discontinue wearing CL temporarily; soft steroids, like loteprednol 0.5% 4 times daily; refit with higher oxygen permeable CL.

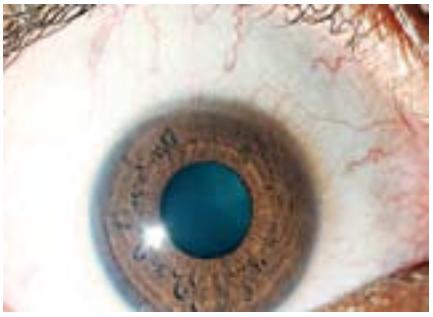


Fig. 3.98.1A: Corneal neovascularization

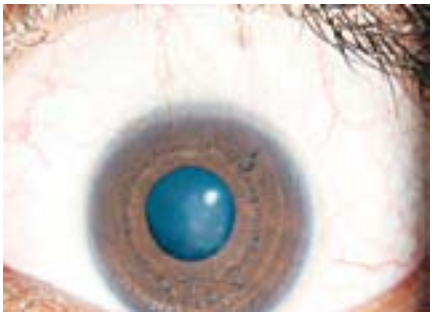


Fig. 3.98.1B: Corneal neovascularization

Contact Lens (Toxic) Keratopathy

- Also called pseudosuperior limbic keratoconjunctivitis; redness and fluorescein staining of superior bulbar conjunctiva at the limbus; SPKs and subepithelial haze **(Figs 3.99.1A and B)**
- May represent chronic toxicity of preservatives in contact lens solution (especially with thiomersal) resulting in apparent limbal stem cell failure

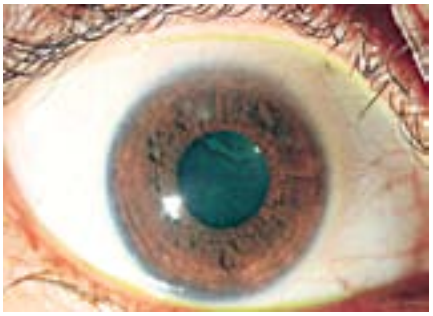


Fig. 3.99.1A: Contact lens (toxic) keratopathy

Fig. 3.99.1B: Contact lens (toxic) keratopathy

- *Treatment:* Preservative-free tears substitute; loteprednol 0.5% 4 times daily; concomitant antibiotics drop; thiomersol-free contact lens solution for cleaning; shift to daily disposable CL.

Infectious Corneal Infiltrates/ Ulcer

- Caused by bacterial (*Pseudomonas* being the most common), fungal and *Acanthamoebal*. (See Chapter: 4) **(Figs 3.100.1 to 3.100.4)**
- *Treatment:* Immediate scraping, smears and culture. Contact lens and solution should also be cultured separately. Start intensive antimicrobial agents every 30 minutes for two hours and then hourly, atropine/homatropine eye drop 2–3 times.

For further management see Chapter 4.

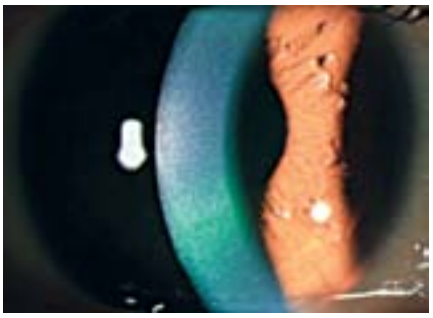


Fig. 3.100.1: Contact lens induced early keratitis

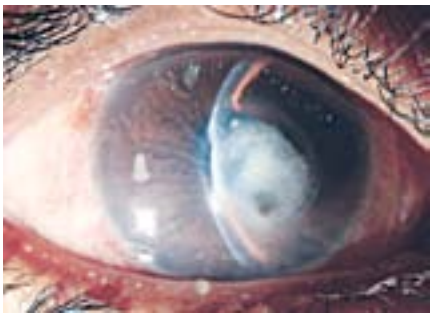


Fig. 3.100.2: Contact lens induced keratitis—note extended wear CL

Fig. 3.100.3: Contact lens induced keratitis—note BCL

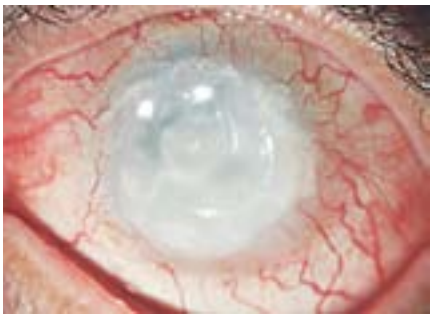


Fig. 3.100.4: Contact lens induced keratitis—total melting of cornea

Giant Papillary Conjunctivitis

- Severe itching; mucoid discharge and large/giant (>1 mm) superior tarsal conjunctival papillae **(Fig. 3.101.1)**; lens intolerance. Upper eyelid eversion should be a part of routine examination in any patient with contact lens
- Sometimes, it is associated with contact lens-induced dry eye **(Fig. 3.101.2)** with significant fluorescein and rose bengal staining
- *Treatment:* Discontinue CL; topical antihistaminics and potent mast cell stabilizer; short-term pulse therapy of soft steroids (loteprednol 0.5% eye drop); contact lens hygiene; replace and refit with a new CL after 2–4 months when the symptoms and signs clear.

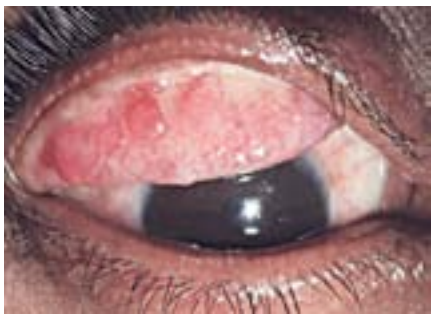


Fig. 3.101.1: Gaint papillary conjunctivitis

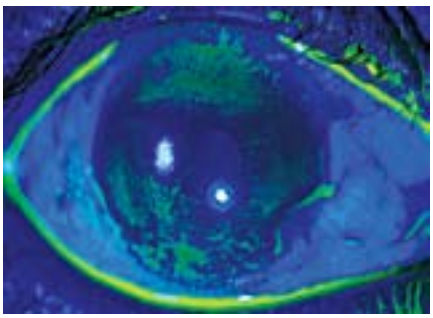


Fig. 3.101.2: Contact lens-induced GPC and dry eye

CHAPTER

4 Diseases of the Sclera

CHAPTER OUTLINE

Inflammation of the Sclera

Blue Sclera

- Sclera appears more blue than white
- Due to increased visibility of the underlying uveal pigment through the thinned sclera **(Fig. 4.1.1)** or may be due to pigment deposition on the normal sclera **(Fig. 4.1.2)**
- *Causes:* Buphthalmos **(Fig. 4.1.3)**, osteogenesis imperfecta, keratoglobus, congenital high myopia, following diffuse scleritis **(Fig. 4.1.4),** ciliary staphyloma, **(Fig. 4.1.5)** Marfan's and Ehlers-Danlos syndrome **(Fig. 4.1.6)**, oculodermal melanocytosis **(Fig. 4.1.7)**.

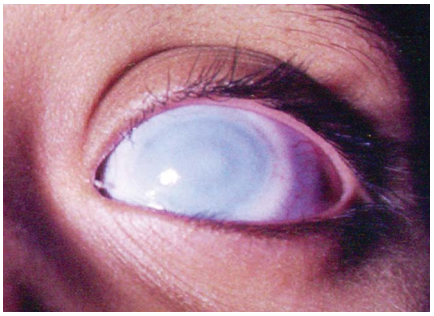


Fig. 4.1.1: Blue sclera—buphthalmos

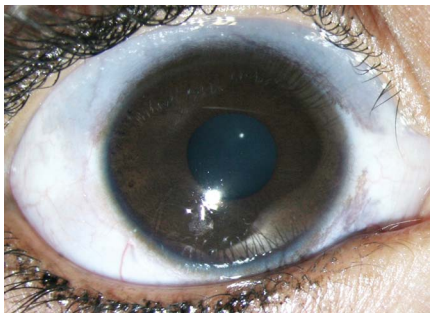


Fig. 4.1.2: Blue sclera—pigment deposition on a normal sclera

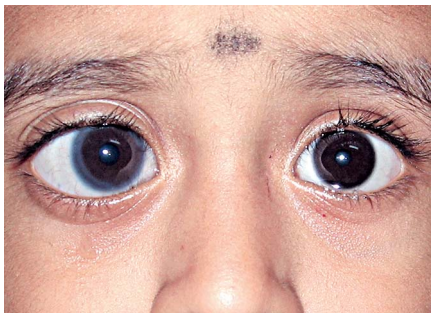


Fig. 4.1.3: Blue sclera—right side in congenital glaucoma

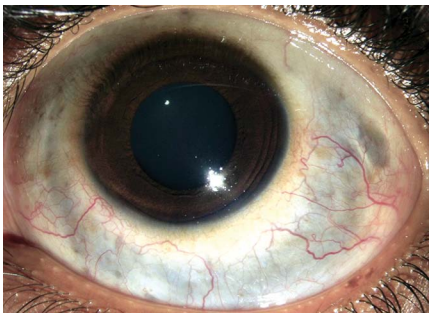


Fig. 4.1.4: Blue sclera following healed scleritis

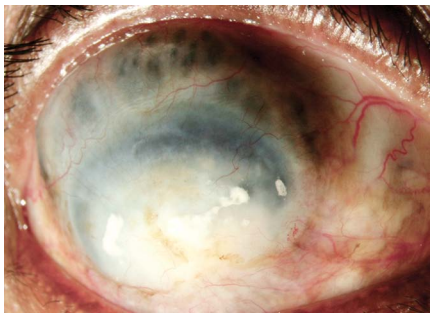


Fig. 4.1.5: Blue sclera—ciliary staphyloma

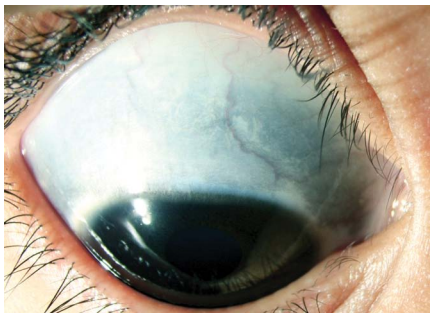


Fig. 4.1.6: Blue sclera—Ehlers-Danlos syndrome

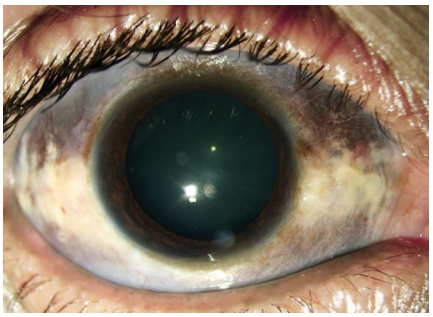


Fig. 4.1.7: Blue sclera—oculodermal melanocytosis

Focal Discoloration of Sclera

- Localized discoloration (blue or brown-black) of the sclera seen in variety of conditions
- *Causes:* Healed focal scleritis **(Fig. 4.2.1)**, equatorial staphyloma **(Fig. 4.2.2)**, long-standing metallic foreign body, alkaptonuria (pigmentation at the insertion of horizontal recti) **(Fig. 4.2.3)** or in extreme old age **(Figs 4.2.4A and B)**.

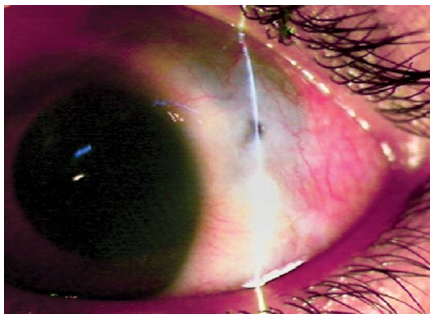


Fig. 4.2.1: Focal discoloration—healed scleritis

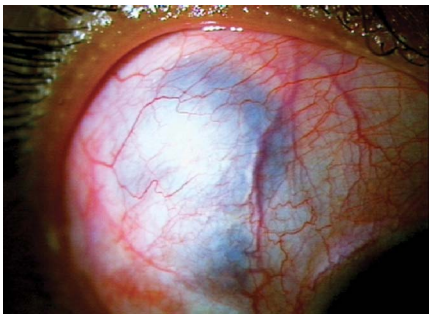


Fig. 4.2.2: Focal blue discoloration—equatorial staphyloma

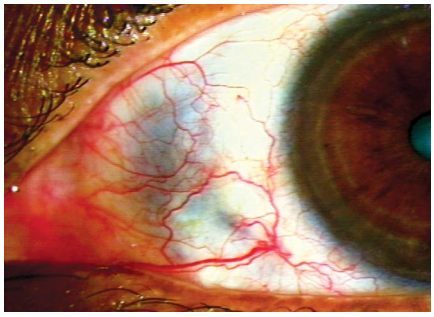


Fig. 4.2.3: Focal discoloration—alkaptonuria

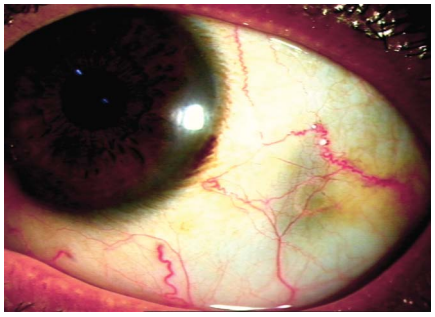


Fig. 4.2.4A: Focal discoloration—old age

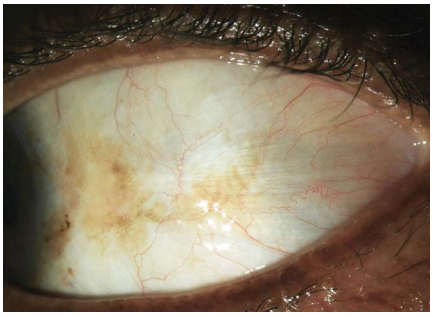


Fig. 4.2.4B: Focal discoloration with prominent lateral rectus

Inflammation of the Sclera

Episcleritis

- Benign inflammation of the episcleral tissue, not so serious

May be *simple* or *nodular*

Simple Episcleritis

- Sectorial or diffuse redness involving the middle episcleral vessels **(Figs 4.3.1A and B)**
- May be bilateral **(Figs 4.3.2A and B)**
- Blanching effect on instillation of phenylepinephrine.

Nodular Episcleritis

- Purple solitary nodule with surrounding injection which can be moved over the sclera
- Situated 1-2 mm away from the limbus **(Figs 4.3.3A and B)**.
- *Treatment:* Oral anti-inflammatory agents, dilute topical corticosteroids, or nonsteroidal anti-inflammatory eye drops.

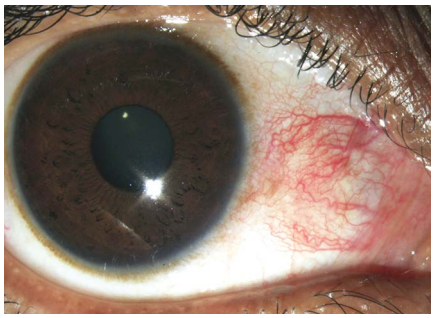


Fig. 4.3.1A: Simple episcleritis

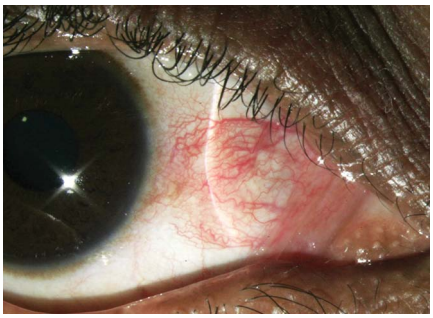


Fig. 4.3.1B: Simple episcleritis

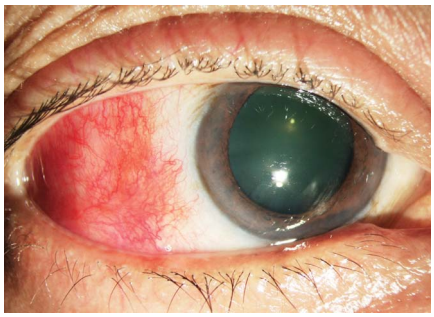


Fig. 4.3.2A: Episcleritis—bilateral—RE

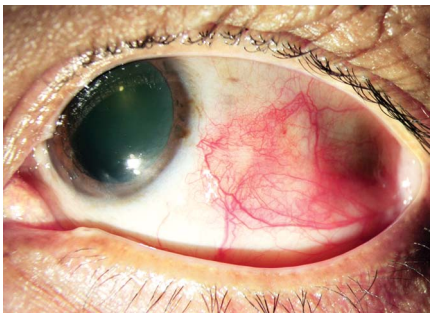


Fig. 4.3.2B: Episcleritis—bilateral—LE

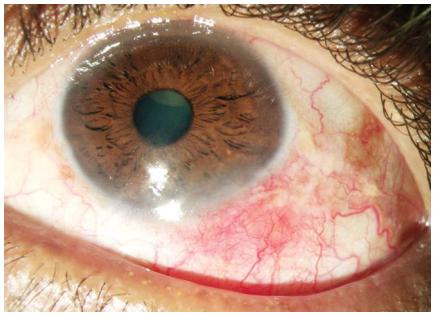


Fig. 4.3.3A: Nodular episcleritis

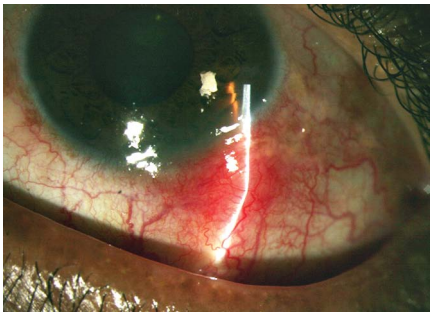


Fig. 4.3.3B: Nodular episcleritis

Scleritis

Anterior Scleritis

- Inflammation of the sclera which is more serious than episcleritis
- Often bilateral and frequently in women
- Associated with systemic collagen disorders in 50% of cases, e.g. rheumatoid arthritis, polyarteritis nodosa, systemic lupus erythematosus, Wegener's granulomatosis, relapsing polychondritis, etc.
- No blanching on phenylepinephrine
- It may be *diffuse*, *nodular* or *necrotizing*

Diffuse Anterior Scleritis

- Involves either a segment **(Fig. 4.4.1)** or the entire anterior sclera **(Figs 4.4.2A and B)**
- Diffuse redness and distortion of pattern of deep episcleral vascular plexus
- Variable episcleral and conjunctival congestion **(Fig. 4.4.3)**
- Entire anterior sclera may be involved with intense deep seated vascularization, called *brawny scleritis* **(Fig. 4.4.4)** which may be associated with anterior uveitis.

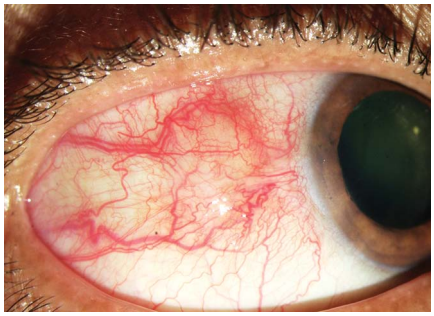


Fig. 4.4.1: Anterior scleritis—segmental

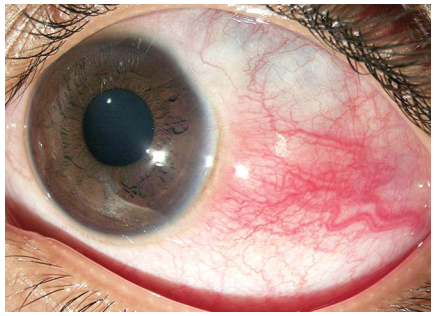


Fig. 4.4.2A: Diffuse anterior scleritis

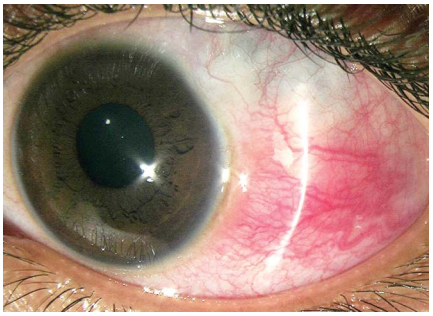


Fig. 4.4.2B: Diffuse anterior scleritis

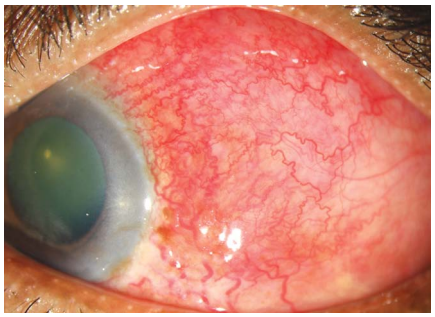


Fig. 4.4.3: Diffuse anterior scleritis

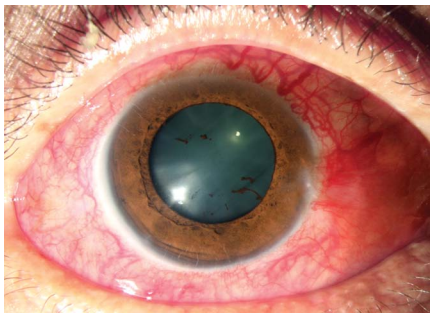


Fig. 4.4.4: Diffuse scleritis—brawny scleritis

Nodular Scleritis (Non-necrotizing)

- Extremely tender, usually solitary **(Figs 4.5.1A and B)** or multiple **(Fig. 4.5.2)** firm immobile nodule separated from the overlying congested episcleral tissue
- Some nodule may be large **(Figs 4.5.3A and B)**
- In collagen diseases, the lesion may recur in a new site **(Fig. 4.5.4)**.

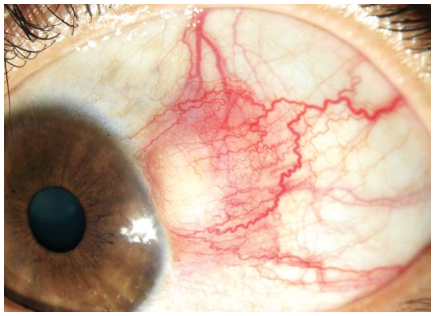


Fig. 4.5.1A: Nodular scleritis

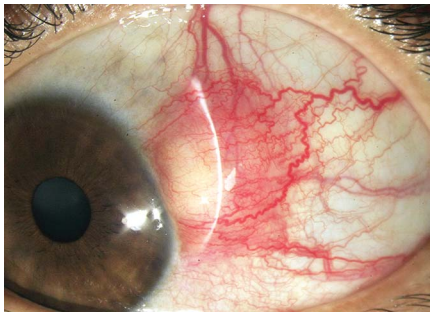


Fig. 4.5.1B: Nodular scleritis

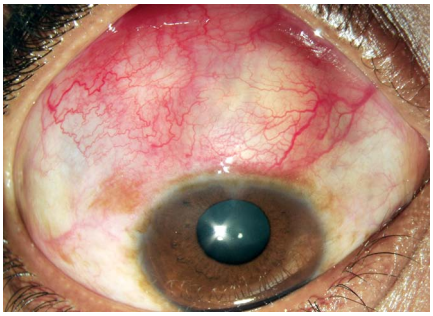


Fig. 4.5.2: Anterior scleritis—multinodular

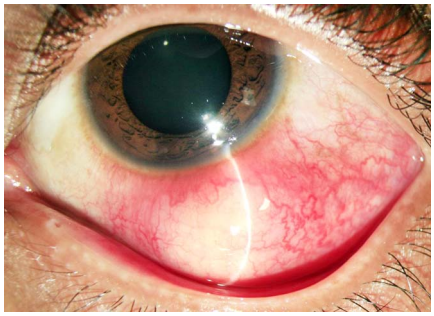


Fig. 4.5.3A: Nodular scleritis—large nodule

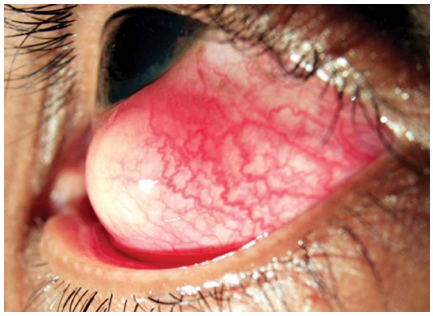


Fig. 4.5.3B: Nodular scleritis—large nodule

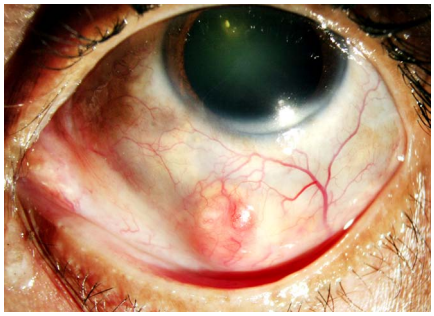


Fig. 4.5.4: Nodular scleritis—recurrent lesion

Anterior Necrotizing Scleritis with Inflammation

- Avascular patches appearing in the episcleral tissue with scleral necrosis and melting **(Fig. 4.6.1)**
- Marked thinning of the sclera with increased visibility of underlying uvea **(Fig. 4.6.2)**
- Associated anterior uveitis and severe keratitis in some cases **(Figs 4.6.3 and 4.6.4)**.

Anterior Necrotizing Scleritis without Inflammation (Scleromalacia Perforans)

- Classically seen in patient with long-standing rheumatoid arthritis

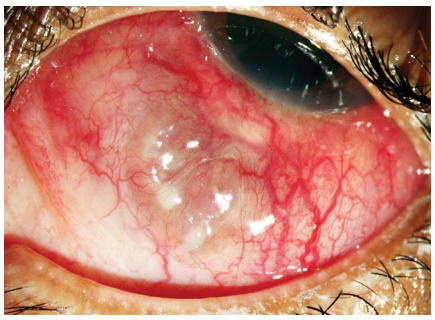


Fig. 4.6.1: Anterior necrotizing scleritis with inflammation

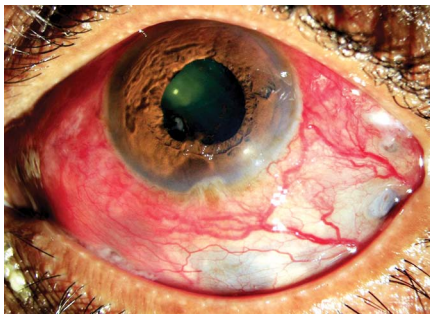


Fig. 4.6.2: Anterior necrotizing scleritis with inflammation

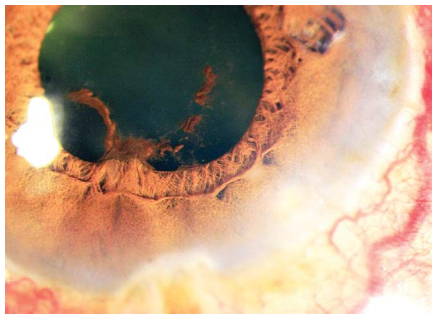


Fig. 4.6.3: Anterior necrotizing scleritis with inflammation with uveitis

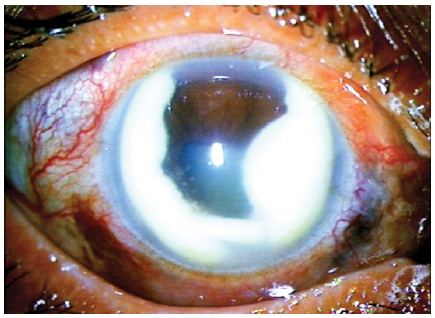


Fig. 4.6.4: Necrotizing scleritis—associated keratitis and anterior uveitis

- Painless, and starts as a white necrotic patch in the normal sclera **(Fig. 4.7.1)**
- Eventually, extreme scleral thinning with exposure and bulging of underlying uvea **(Figs 4.7.2A and B)**
- *Treatment:* Systemic and topical corticosteroids, nonsteroidal anti-inflammatory agents, immunosuppressive (antimetabolites) in severe and unresponsive cases; and investigations for systemic collagen disorders.

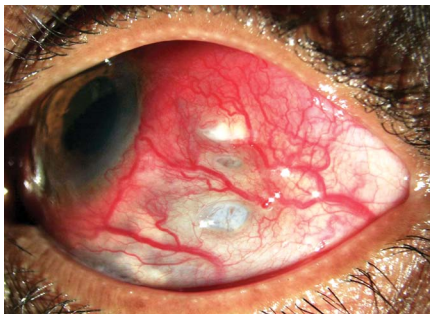


Fig. 4.7.1: Scleromalacia perforans

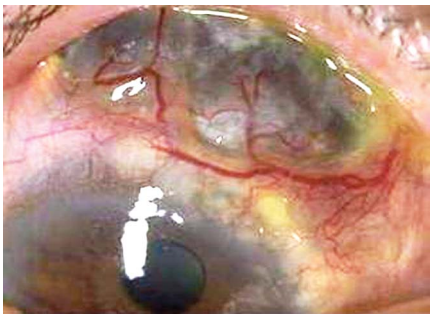


Fig. 4.7.2A: Scleromalacia perforans

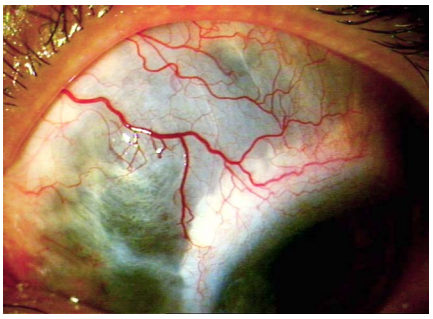


Fig. 4.7.2B: Scleromalacia perforans

Posterior Scleritis

- The signs depend upon site of maximum involvement primarily
- May be associated with anterior scleritis
- Inward extension gives rise to 'uveal effusion syndrome'—choroiditis, choroidal effusion, exudative retinal detachment, macular edema **(Fig. 4.8.1)**
- Outward extension into the orbit gives rise to proptosis with extraocular muscle involvement
- *Treatment:* Systemic and topical corticosteroids, nonsteroidal anti-inflammatory agents; immunosuppressive in severe and unresponsive cases.

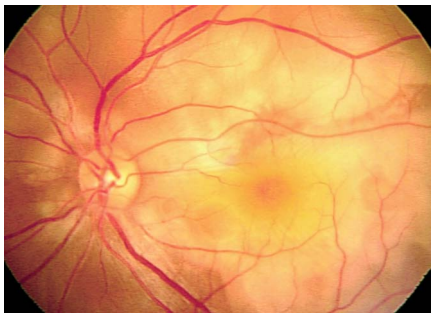


Fig. 4.8.1: Posterior scleritis

Scleral Abscess

- Rare, unilateral condition
- Nodular yellowish-white lesion, with mild to moderate pain **(Fig. 4.9.1)** and localized congestion
- Usually traumatic, with impacted foreign body **(Figs 4.9.2 and 4.9.3A and B)**

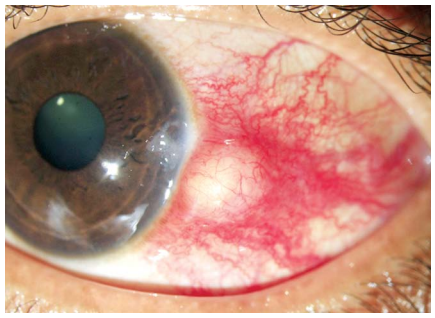


Fig. 4.9.1: Scleral abscess

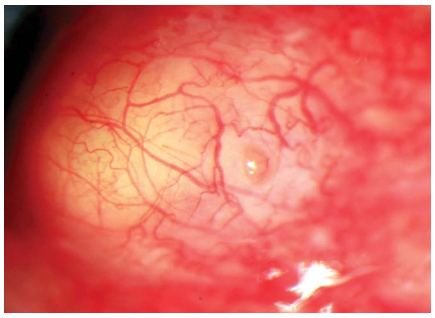


Fig. 4.9.2: Scleral abscess—caterpillar hair

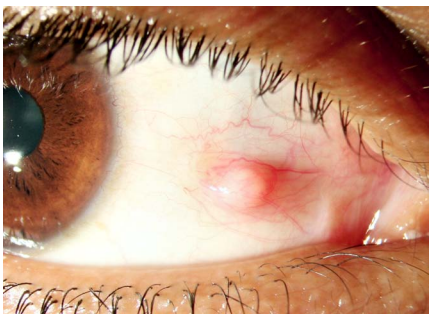


Fig. 4.9.3A: Scleral abscess—thorn prick

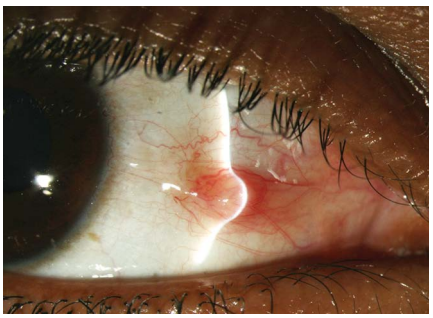


Fig. 4.9.3B: Scleral abscess—thorn prick

- May occur following tuberculous infection with or without associated anterior uveitis **(Figs 4.9.4 and 4.9.5)**
- *Treatment:* Topical 4th generation fluoroquinolones; incision and drainage of abscess; culture sensitivity and appropriate antibiotics; in tuberculosis, full course with antitubercular drugs.

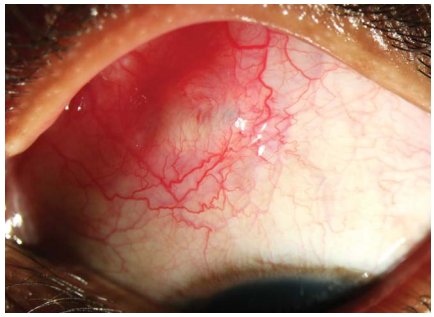


Fig. 4.9.4A: Scleral abscess—tubercular

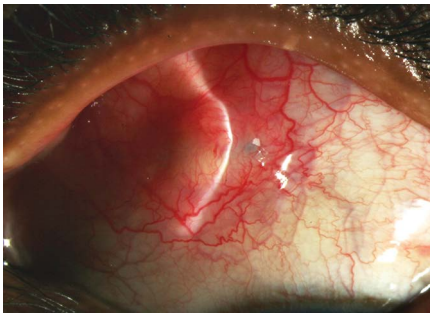


Fig. 4.9.4B: Scleral abscess—tubercular

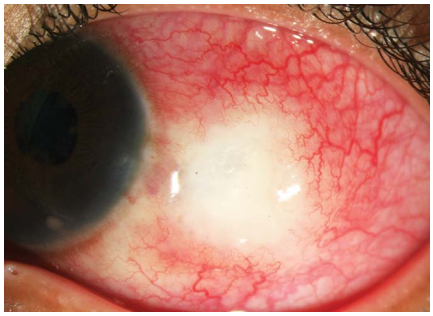


Fig. 4.9.5A: Scleral abscess—tubercular

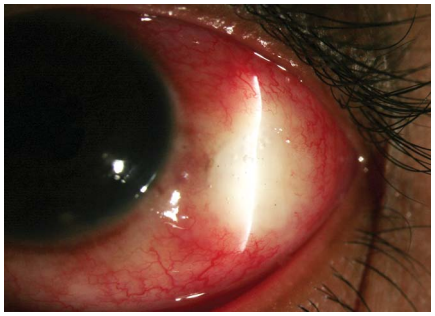


Fig. 4.9.5B: Scleral abscess—tubercular

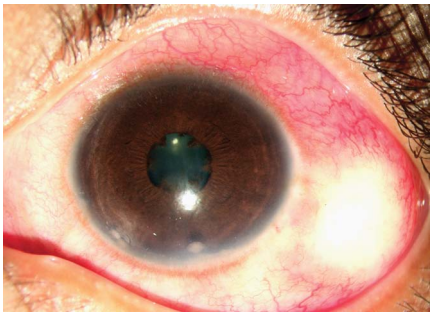


Fig. 4.9.5C: Scleral abscess—tubercular with anterior uveitis

Scleral Cyst

- Very rare unilateral condition, and usually traumatic
- Translucent-white cystic lesion at the limbus with mild to moderate pain and minimum localized congestion; in cases, it may extend in the cornea **(Figs 4.10.1A and B)**
- The fluid may be clear or turbid
- Diagnosis is confirmed by UBM
- *Treatment:* De-roofing of the cyst with scleral or sclerocorneal lamellar patch graft.

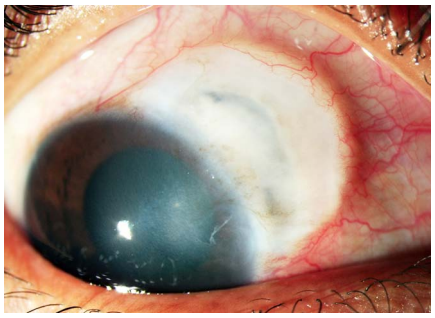


Fig. 4.10.1A: Scleral cyst

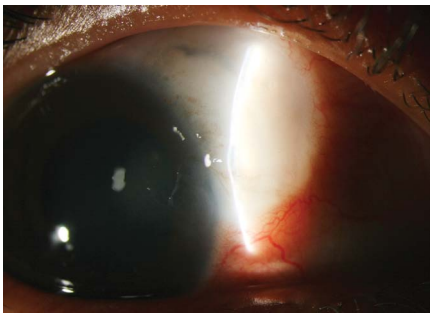


Fig. 4.10.1B: Scleral cyst—scleromalacia

Surgically Induced Necrosis of Sclera (SINS)

- Rare postoperative immune-mediated necrosis of sclera
- May be triggered by excessive cautery, use of topical antimetabolities (like, MMC or 5-FU) during surgery, etc.
- *May be noticed following:*
  - Cataract surgery **(Figs 4.11.1 and 4.11.2)**

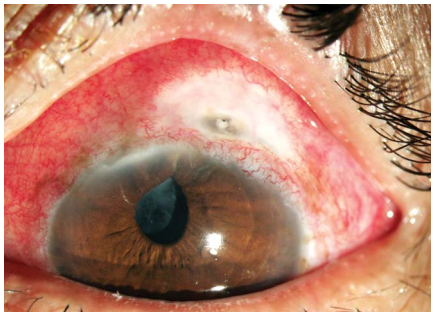


Fig. 4.11.1A: SINS—post-cataract surgery

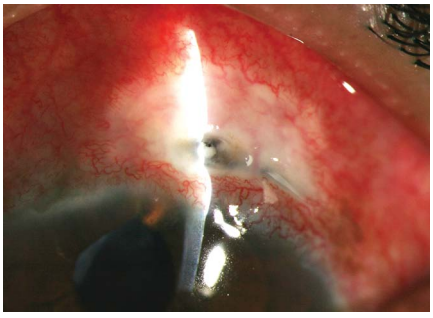


Fig. 4.11.1B: Surgically induced necrosis of sclera

- Pterygium surgery **(Figs 4.11.3A to F)**
- There may be associated frank infection of the sclera bed in pterygium surgery **(Figs 4.11.4A and B)**
- Vitreoretinal surgery **(Figs 4.11.5 and 4.11.6)**

- *Treatment:* Topical and systemic steroids, systemic immune-suppressants and scleral patch graft extreme situation.

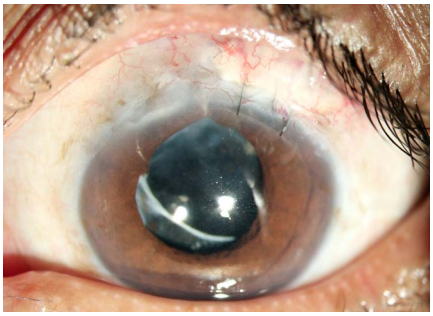


Fig. 4.11.2: Scleral patch graft—same eye **(Fig. 4.11.1)**

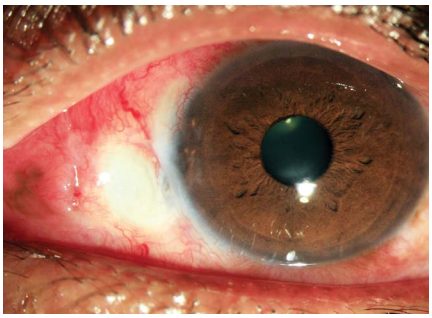


Fig. 4.11.3A: SINS—following pterygium surgery

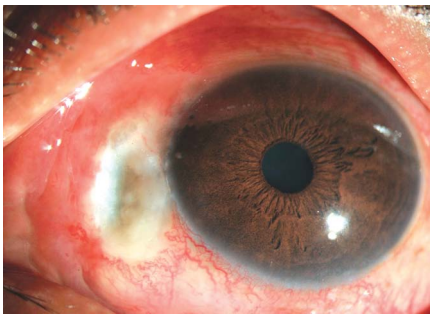


Fig. 4.11.3B: SINS—following pterygium surgery

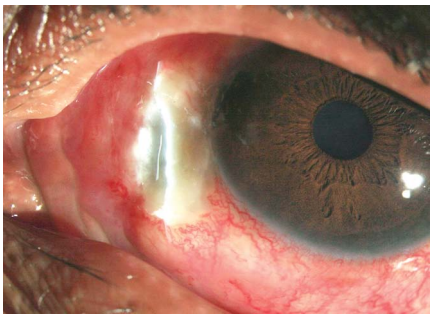


Fig. 4.11.3C: SINS—following pterygium surgery

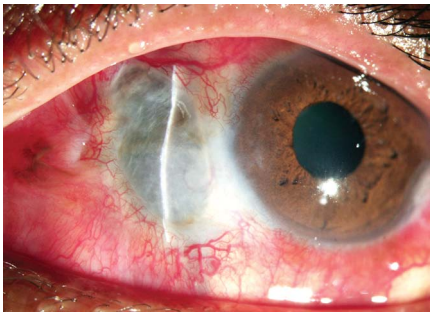


Fig. 4.11.3D: SINS—following pterygium surgery

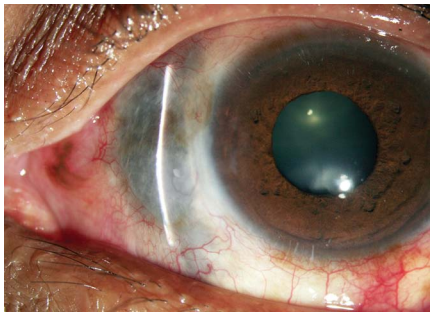


Fig. 4.11.3E: SINS—following pterygium surgery

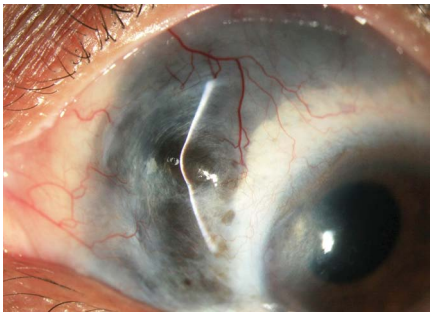


Fig. 4.11.3F: SINS—after pterygium surgery—same patients after 2 years

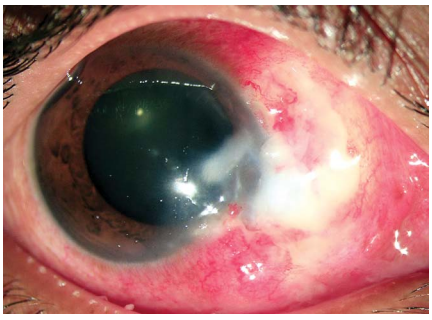


Fig. 4.11.4A: SINS after pterygium surgery—frank infection of the scleral bed

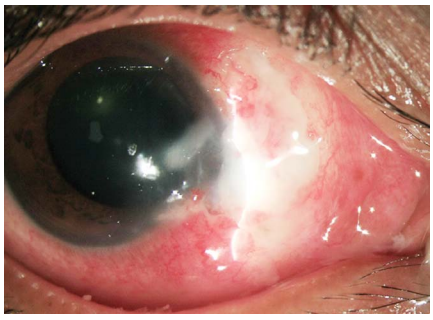


Fig. 4.11.4B: SINS after pterygium surgery—frank infection with thinning

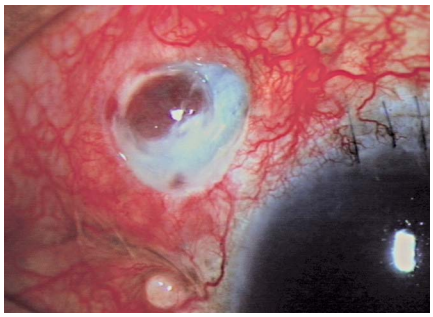


Fig. 4.11.5: SINS—post-VR surgery

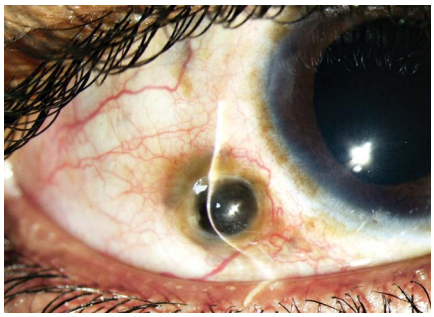


Fig. 4.11.6: SINS following VR surgery

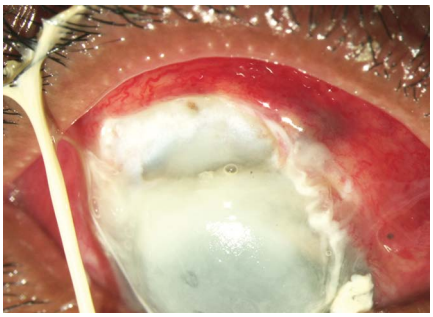


Fig. 4.12.1: Scleral tunnel abscess—following manual SICS

Scleral Tunnel Abscess

- Associated with infiltrations within the sclerocorneal tunnel after Phaco or SICS
- Variable degree of anterior chamber reactions and eventually it may progress into frank endophthalmitis **(Fig. 4.12.1)**
- ***More in Chapter 3 (Page No:.138-139: Figs 3.65.1 to 3.65.6)***.

CHAPTER

5 Abnormalities in Anterior Chamber

CHAPTER OUTLINE

Abnormal Depth of the Anterior Chamber

- Shallow anterior chamber
- Deep anterior chamber
- Irregular anterior chamber

Abnormal Contents of the Anterior Chamber

- Blood (hyphema)
- Pus (hypopyon)
- Inverse hypopyon and pseudohypopyon
- Albuminous materials
- Other abnormal contents in anterior chamber

Anterior Chamber

- Space between the cornea and the iris
- Contains crystal clear aqueous humor
- Central depth is approximately 2.5 mm, and uniformly, and gradually becoming shallower towards the periphery **(Fig. 5.1.1)**
- Depth varies in normal condition and also in diseases **(Figs 5.1.2 to 5.1.5)**.

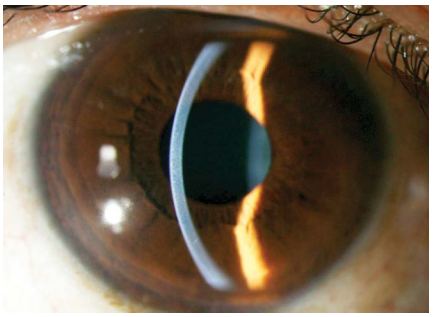


Fig. 5.1.1: Normal depth of anterior chamber (AC)

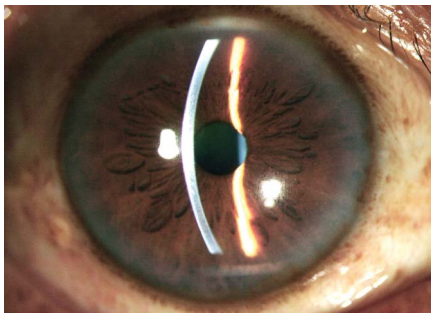


Fig. 5.1.2: Mild shallow AC

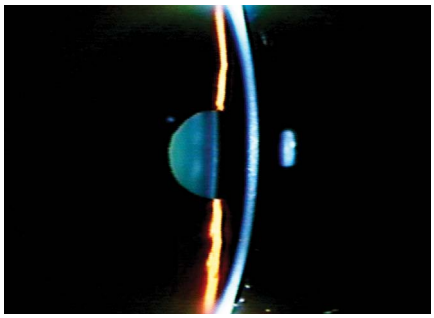


Fig. 5.1.3: Moderately shallow AC

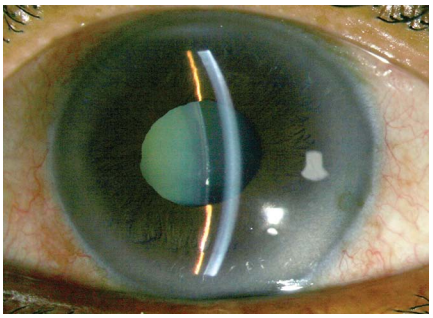


Fig. 5.1.4: Very shallow AC

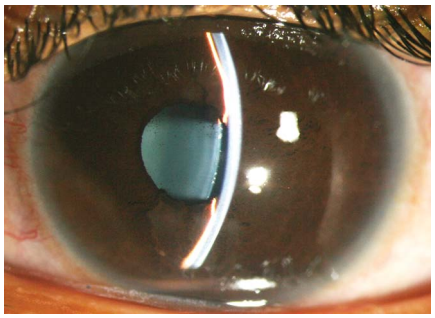


Fig. 5.1.5: Extremely (Flat) AC

Abnormal Depth

Shallow Anterior Chamber

- *Causes:*
  - Cornea plana **(Fig. 5.2.1)**
  - Hypermetropia **(Fig. 5.2.2)**
  - Intumescent and hypermature cataract **(Figs 5.2.3A and B)**
  - Pupillary block or narrow angle glaucoma **(Figs 5.2.4 and 5.2.5)**
  - Over filtration of glaucoma filtering bleb **(Fig. 5.2.6)** or choroidal detachment following trabeculectomy **(Fig. 5.2.7)**
  - Neovascular glaucoma **(Fig. 5.2.8)**
  - Wound leak after intraocular surgery
  - Penetrating injury **(Fig. 5.2.9)** or following perforation of a cornea ulcer **(Fig. 5.2.10)**
  - Malignant glaucoma following trabeculectomy (extremely shallow or flat AC) **(Fig. 5.2.11)**.

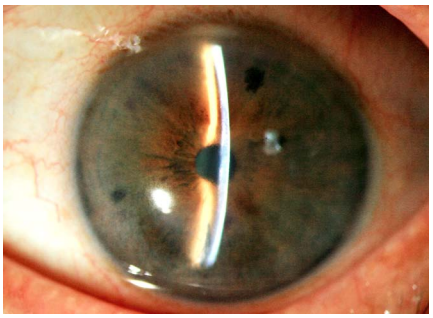


Fig. 5.2.1: Shallow AC in cornea plana

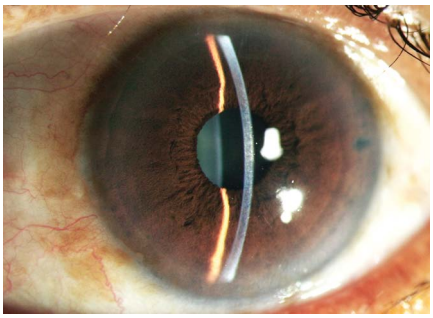


Fig. 5.2.2: Shallow AC—hypermetropia

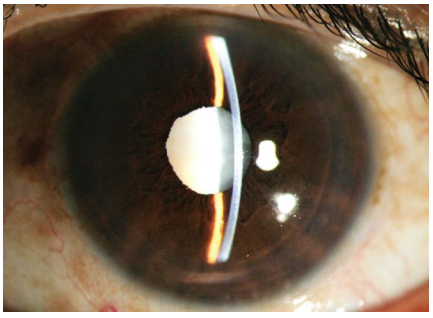


Fig. 5.2.3A: Shallow AC—hypermature and intumescent cataract

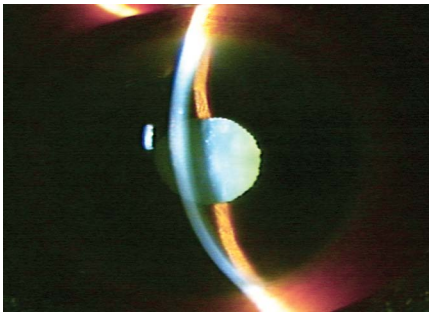


Fig. 5.2.3B: Shallow AC—intumescent cataract

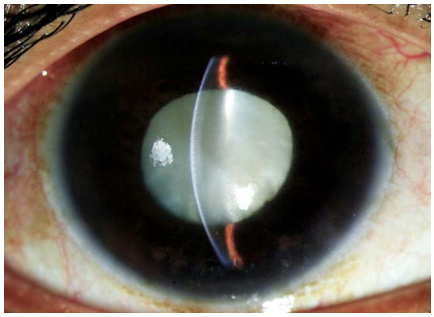


Fig. 5.2.4: Shallow AC—lens induced secondary angle closure

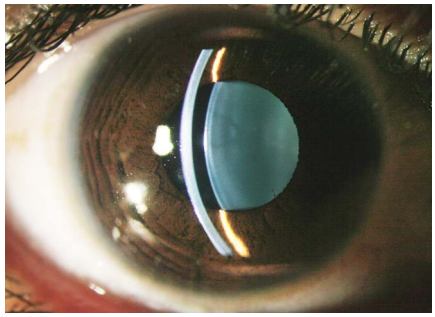


Fig. 5.2.5: Shallow AC in narrow angle glaucoma

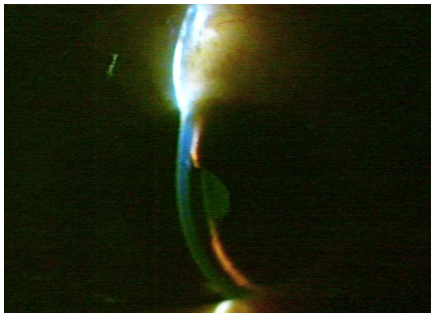


Fig. 5.2.6: Shallow AC—excessive bleb filtration

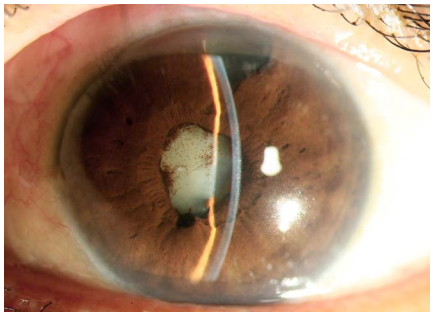


Fig. 5.2.7: Shallow AC—choroidal detachment after trabeculectomy

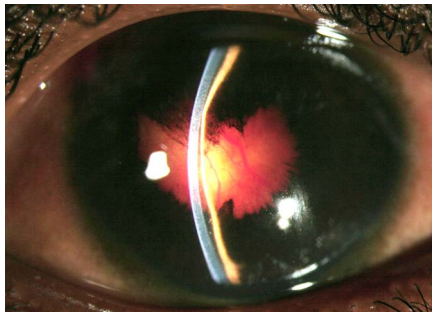


Fig. 5.2.8: Shallow AC—neovascular glaucoma

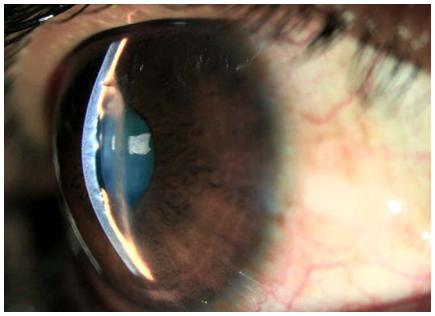


Fig. 5.2.9: Shallow AC—following penetrating injury

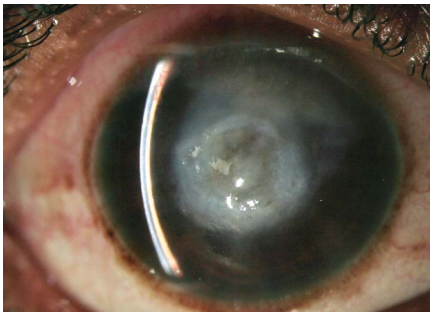


Fig. 5.2.10: Very shallow (Flat) AC—healed perforted cornea

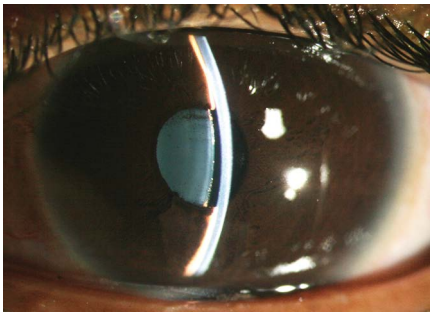


Fig. 5.2.11: Extremely shallow (Flat) AC—malignant glaucoma

Deep Anterior Chamber

- *Causes:*
  - Myopia **(Fig. 5.3.1)**
  - Aphakia and pseudophakia **(Figs 5.3.2A and B)**
  - Megalocornea **(Fig. 5.3.3)**
  - Keratoglobus **(Fig. 5.3.4)**, keratoconus **(Fig. 5.3.5)** and pellucid marginal degeneration **(Fig. 5.3.6)**
  - Buphthalmos **(Fig. 5.3.7)**
  - Posterior dislocation of the lens.

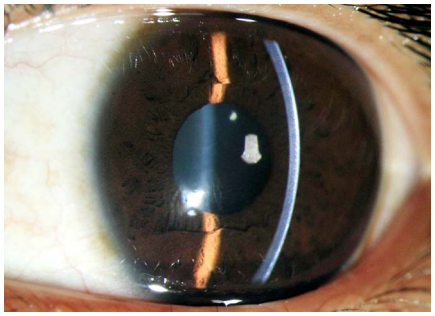


Fig. 5.3.1: Deep AC—myopia

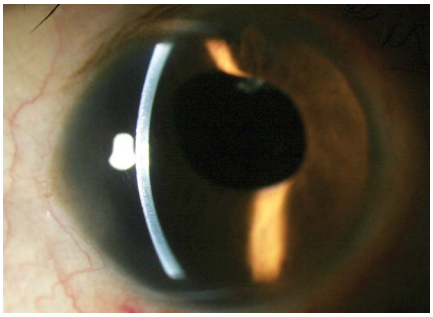


Fig. 5.3.2A: Deep AC—aphakia

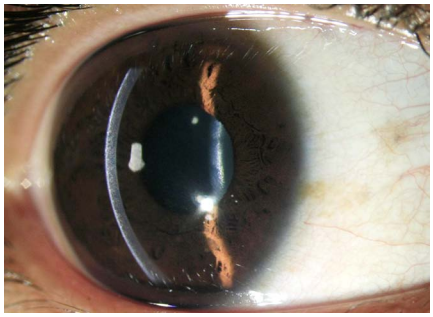


Fig. 5.3.2B: Deep AC—pseudophakia

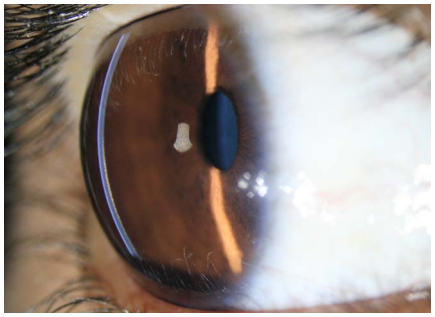


Fig. 5.3.3: Deep AC—megalocornea

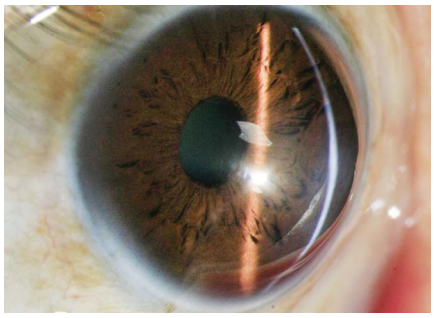


Fig. 5.3.4: Deep AC—keratoglobus

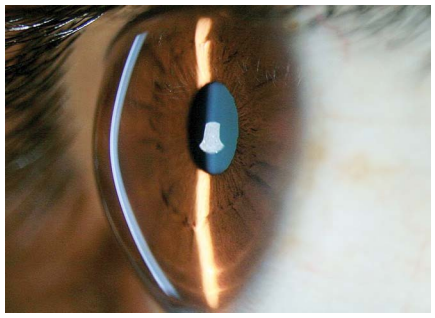


Fig. 5.3.5: Deep AC—keratoconus

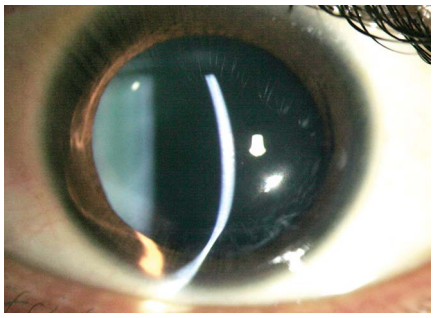


Fig. 5.3.6: Deep AC—pellucid marginal degeneration

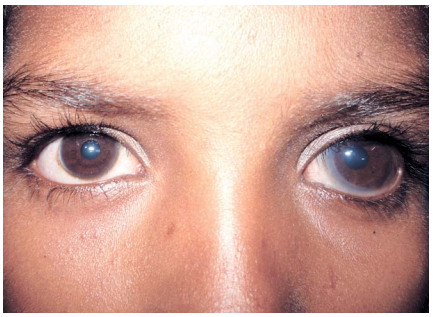


Fig. 5.3.7: Deep AC—Juvenile glaucoma—left side

Irregular Anterior Chamber

- *Causes:*
  - Ectopia lentis **(Fig. 5.4.1)** or traumatic subluxation of crystalline lens **(Fig. 5.4.2)**
  - Iris bombe: funnel shaped **(Figs 5.4.3 and 5.4.4)**
  - Iris cyst or tumor **(Fig. 5.4.5)**
  - Adherent corneal leukoma **(Fig. 5.4.6)**
  - Penetrating injury or corneal ulcer with iris prolapse **(Fig. 5.4.7)**
  - Angle recession after blunt trauma.

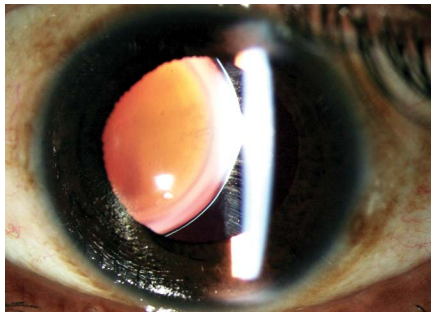


Fig. 5.4.1: Irregular AC—ectopia lentis

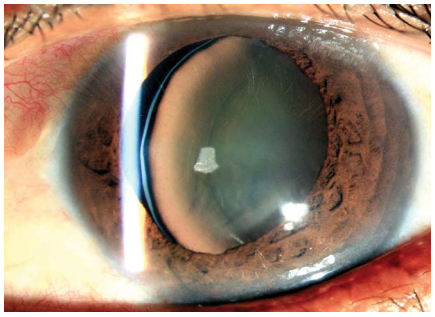


Fig. 5.4.2: Irregular AC—traumatic subluxation of crystalline lens

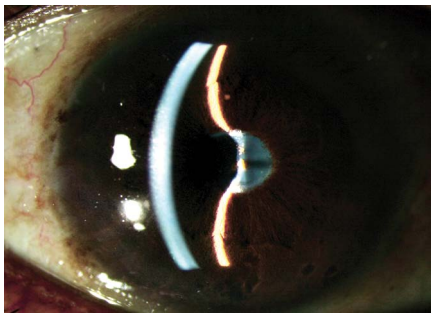


Fig. 5.4.3: Funnel shaped AC—iris bombe

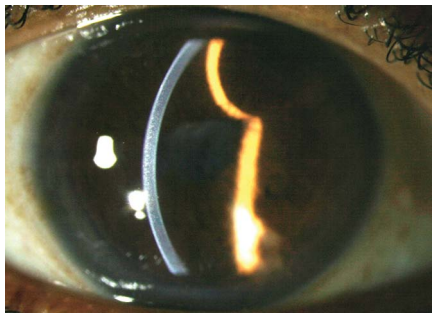


Fig. 5.4.4: Funnel shaped AC—iris bombe

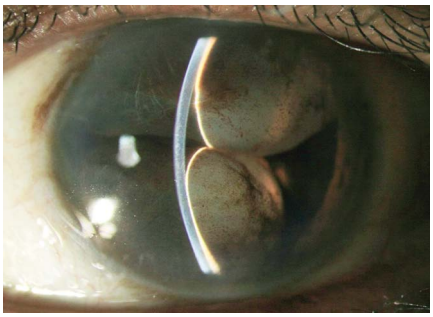


Fig. 5.4.5: Irregular AC—iris cyst

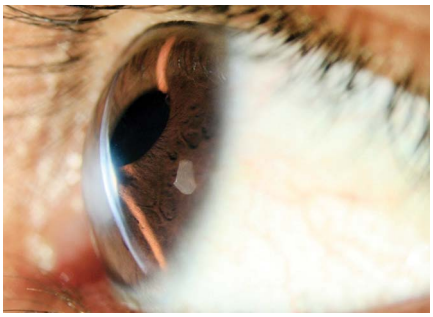


Fig. 5.4.6: Irregular AC—adherent leukoma

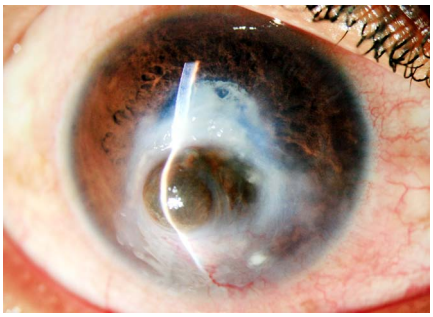


Fig. 5.4.7: Irregular AC—iris prolapse

Abnormal Contents

Blood (Hyphema)

- *Traumatic:* It may be fresh **(Fig. 5.5.1)**, partially absorbed or organized **(Fig. 5.5.2)**, and clotted—8-ball or black ball hyphema **(Figs 5.5.3A and B)**
- *Postoperative:* Mainly in cataract surgery—bleeding from tunnel or iridodialysis **(Figs 5.5.4A and B)**
- Herpetic iridocyclitis
- Neovascularization of iris (NVI) **(Figs 5.5.5A and B)**
- *Spontaneous:* As in leukemia or other blood dyscrasias **(Fig. 5.5.6)**.

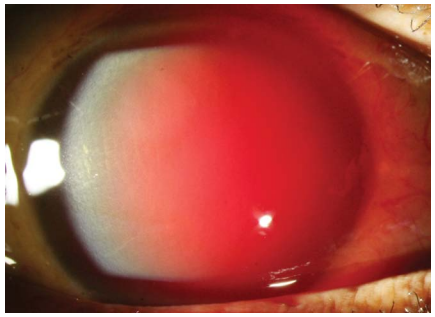


Fig. 5.5.1: Blood in AC—traumatic hyphema—fresh

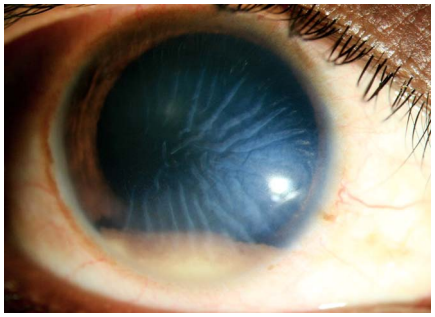


Fig. 5.5.2: Traumatic hyphema—organized blood

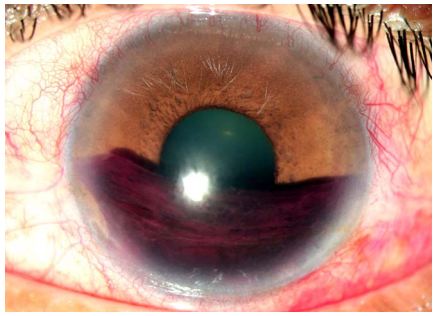


Fig. 5.5.3A: Cotted blood—8-ball hyphema

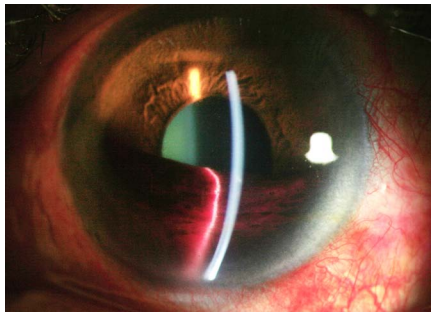


Fig. 5.5.3B: Cotted blood—8-ball hyphema

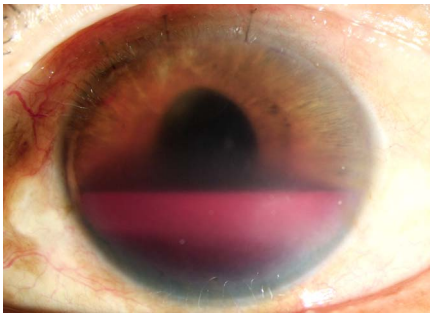


Fig. 5.5.4A: Postoperative hyphema

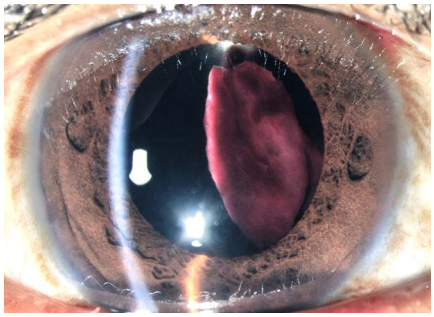


Fig. 5.5.4B: Postoperative hyphema—blood clot in pupillary area

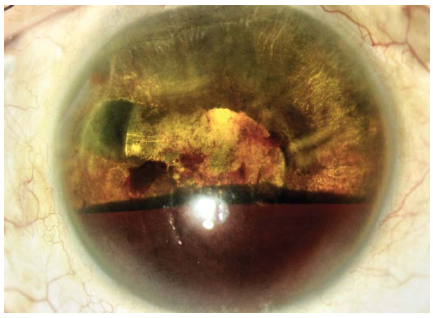


Fig. 5.5.5A: Hyphema in neovascularization of iris

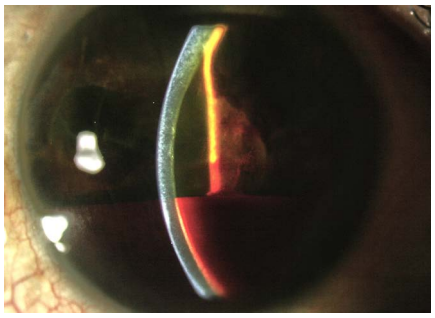


Fig. 5.5.5B: Hyphema in neovascularization of iris—note blood level

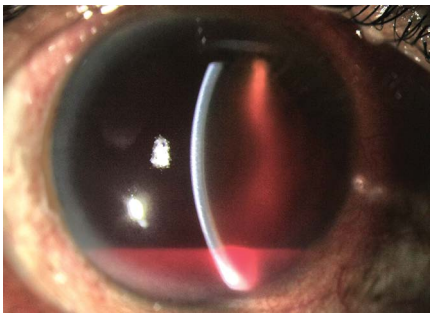


Fig. 5.5.6: Hyphema—spontaneous in blood dyscrasia and partially settled

Pus (Hypopyon)

It may be mild, moderate, or may be total or full chamber hypopyon:

- Corneal ulcer **(Figs 5.6.1A to C)**
- Acute iridocyclitis **(Fig. 5.6.2)**
- *Endophthalmitis:* May be postoperative **(Figs 5.6.3 and 5.6.4)**, post-traumatic **(Fig. 5.6.5)** or endogenous **(Figs 5.6.6 and 5.6.7)**
- Panophthalmitis **(Fig. 5.6.8)**.

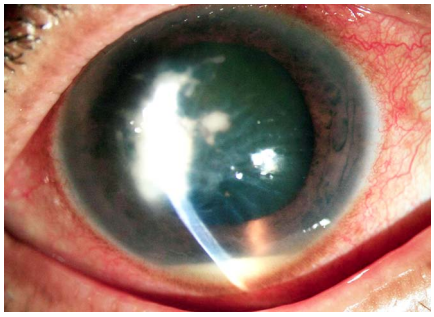


Fig. 5.6.1A: Pus in AC—mild hypopyon—corneal ulcer

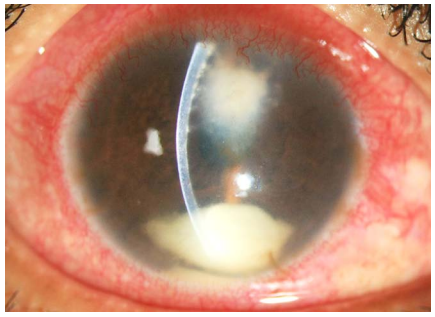


Fig. 5.6.1B: Moderate hypopyon—corneal ulcer

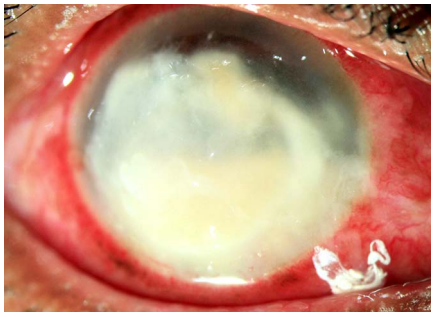


Fig. 5.6.1C: Severe hypopyon—corneal ulcer

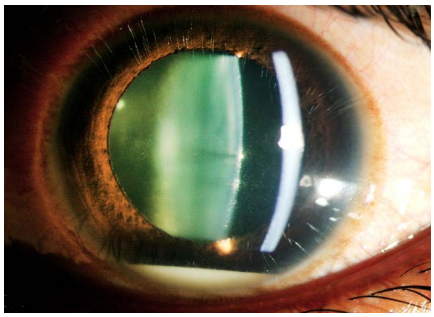


Fig. 5.6.2: Pus in AC—mild hypopyon—acute iridocyclitis

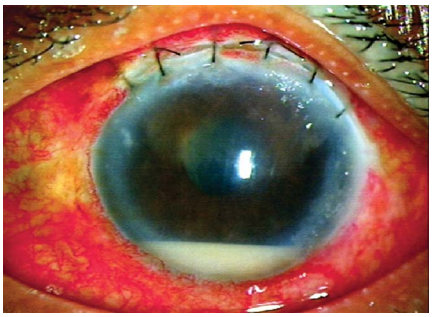


Fig. 5.6.3A: Hypopyon—postoperative (ECCE) endophthalmitis

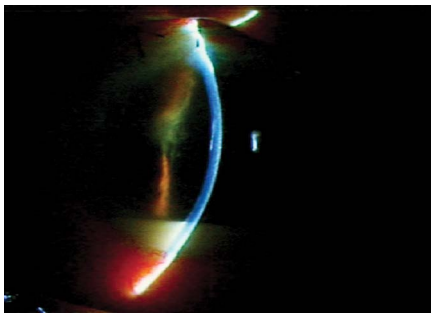


Fig. 5.6.3B: Hypopyon—postoperative (ECCE) endophthalmitis

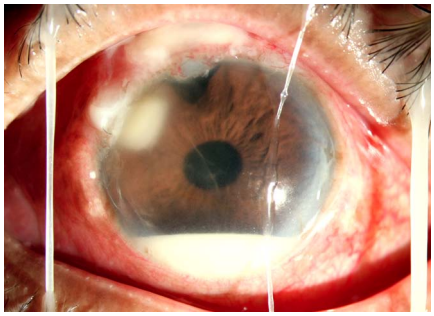


Fig. 5.6.4: Hypopyon—postoperative (phaco) endophthalmitis

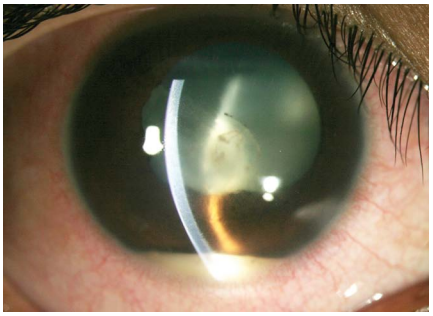


Fig. 5.6.5: Hypopyon—post-traumatic endophthalmitis

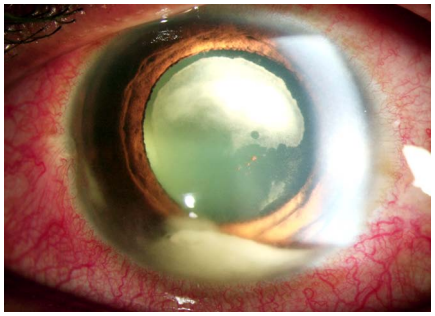


Fig. 5.6.6: Hypopyon—endogenous endophthalmitis

Fig. 5.6.7: Hypopyon—endogenous fungal endophthalmitis

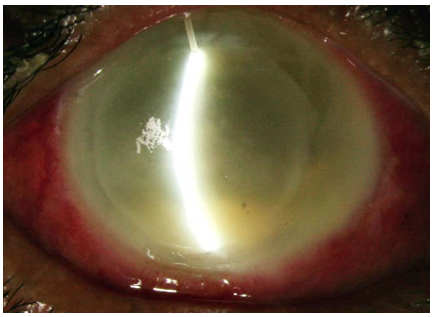


Fig. 5.6.8: Hypopyon—full chamber in panophthalmitis

Inverse Hypopyon and Pseudohypopyon

- ***Inverse hypopyon (silicone oil):*** It is seen after vitreoretinal surgery with silicone oil **(Figs 5.7.1A to D)**; a gonioscopy is helpful to detect small amount of silicone oil in the anterior chamber in the upper part **(Fig. 5.7.2)**. In long standing cases, it may appear as double hypopyon **(Fig. 5.7.3)**
- ***Pseudohypopyon:*** In the lower part same as hypopyon
- Malignant cells, as in retinoblastoma **(Figs 5.8.1 and 5.8.2)**
- Liquefied milky cortex in hypermature Morgagnian cataract **(Figs 5.8.3 and 5.8.4)**
- Cholesterolosis bulbi **(Fig. 5.8.5)**
- Rarely in sclerocorneal cyst **(Figs 5.8.6A and B)**.

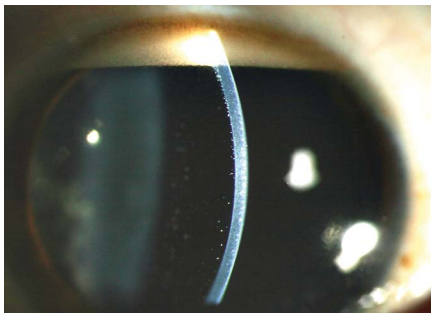


Fig. 5.7.1A: Inverse hypopyon—emulsified silicone oil in phakic eye

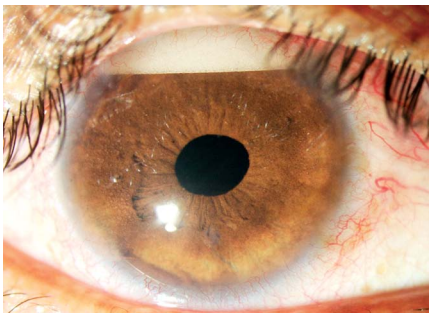


Fig. 5.7.1B: Inverse hypopyon in aphakic eye—emulsified silicone oil

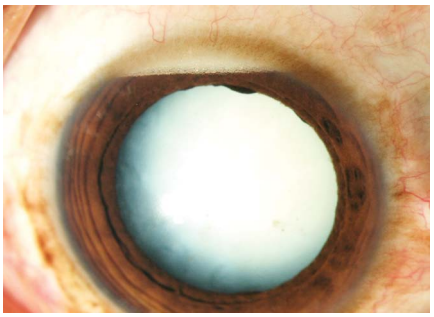


Fig. 5.7.1C: Inverse hypopyon—with mature cataract

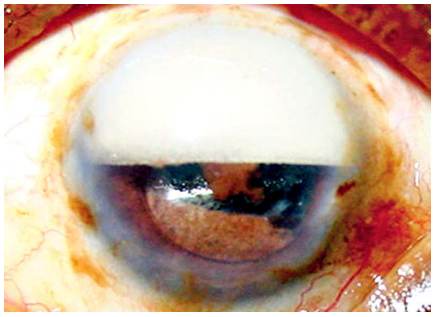


Fig. 5.7.1D: Inverse hypopyon—emulsified silicone oil at angle

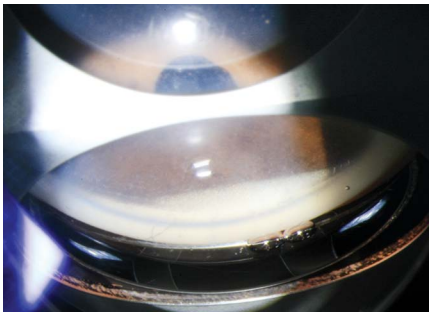


Fig. 5.7.2: Inverse hypopyon—emulsified silicone oil at angle

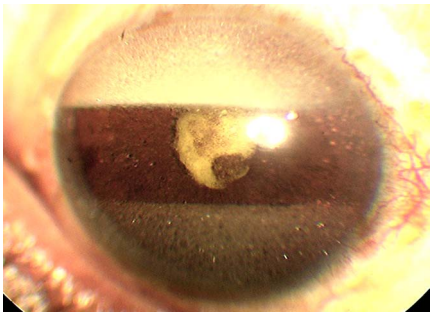


Fig. 5.7.3: Double hypopyon—emulsified silicone oil

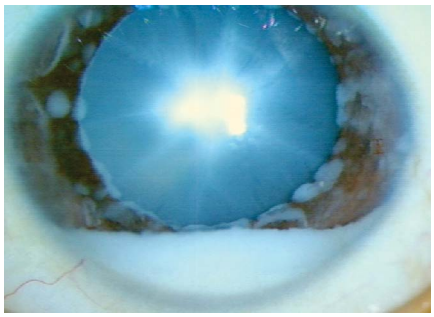


Fig. 5.8.1: Pseudohypopyon—retinoblastoma

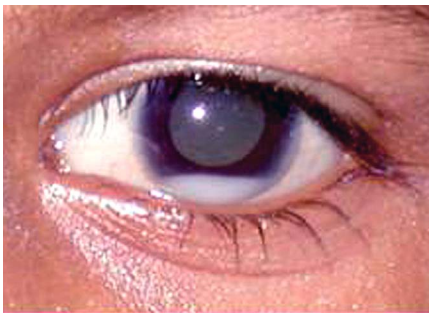


Fig. 5.8.2: Leukocoria—retinoblastoma—pseudohypopyon

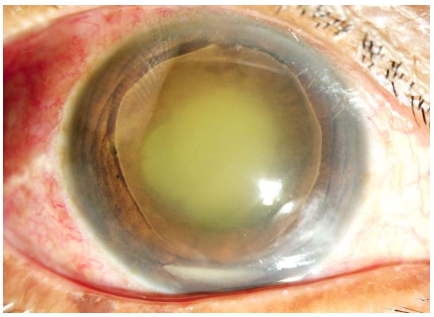


Fig. 5.8.3: Crystalline lens nucleus in AC with pseudohypopyon

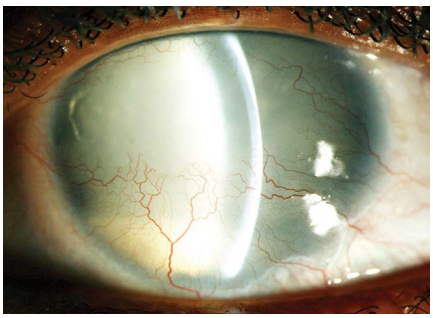


Fig. 5.8.4: Pseudohypopyon—liquefied lens cortex in phacolytic

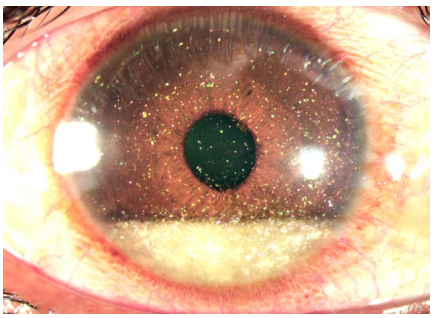


Fig. 5.8.5: Pseudohypopyon—cholesterolosis bulbi

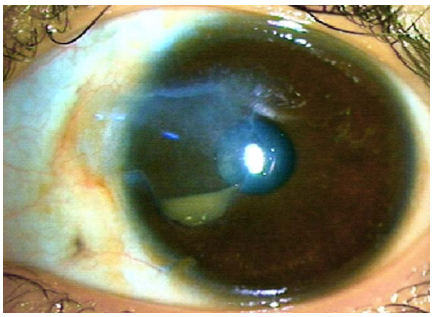


Fig. 5.8.6A: Sclerocorneal cyst—pseudohypopyon

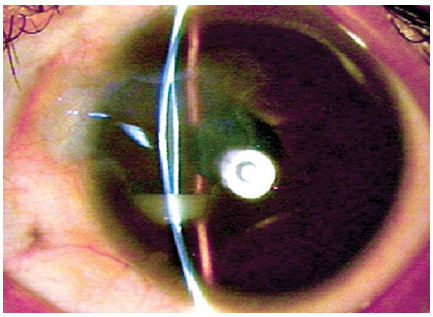


Fig. 5.8.6B: Sclerocorneal cyst—intracorneal pseudohypopyon

Albuminous Materials

- Aqueous flare in iritis **(Figs 5.9.1 and 5.9.2)**
- Aqueous cells in iritis **(Figs 5.9.3 and 5.9.4)**
- Fibrinous exudates in iritis **(Fig. 5.9.5)**
- Dense pyogenic exudates in endophthalmitis **(Figs 5.9.6 to 5.9.8)**.

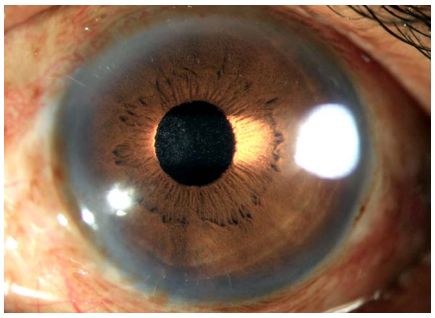


Fig. 5.9.1: Aqueous flare—postoperative iritis

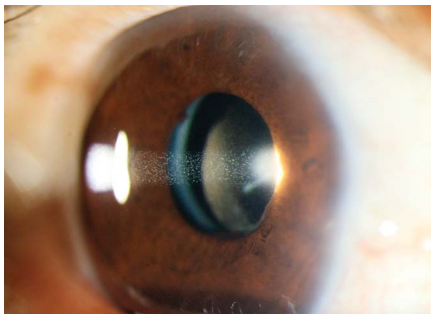


Fig. 5.9.2A: Aqueous flare in acute iridocyclitis

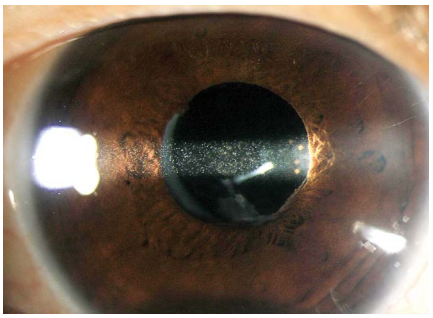


Fig. 5.9.2B: Aqueous flare—postoperative iritis

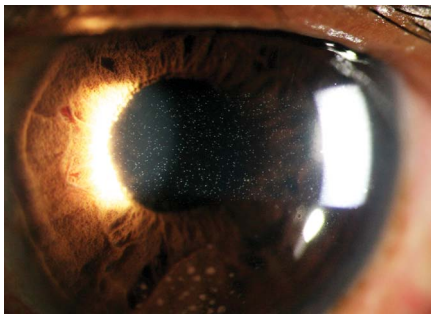


Fig. 5.9.3: Cells in AC—acute iritis

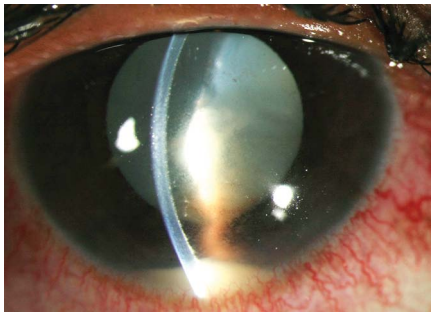


Fig. 5.9.4: Dense cells in AC—endophthalmitis

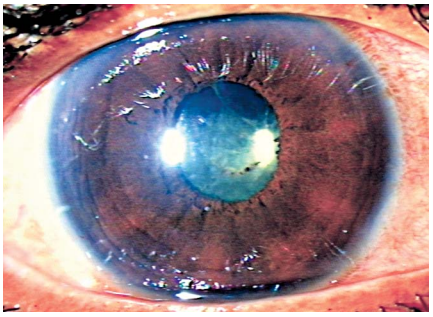


Fig. 5.9.5: Fibrinous exudate—iritis

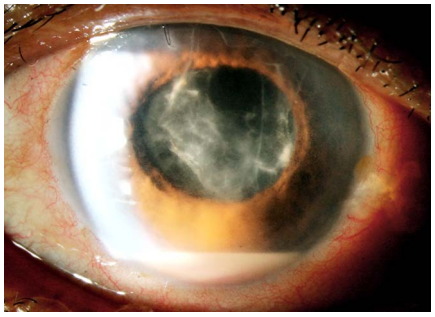


Fig. 5.9.6: AC exudates in postoperative endophthalmitis

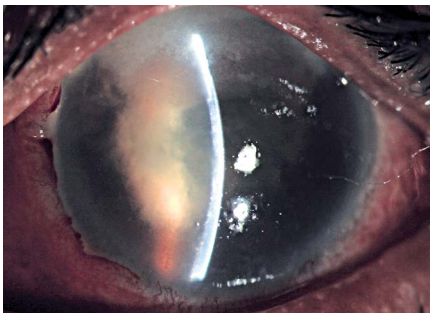


Fig. 5.9.7: Frank exudate in AC—endophthalmitis

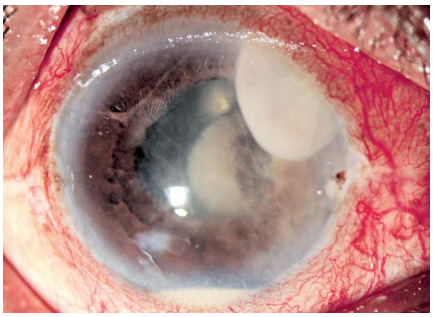


Fig. 5.9.8: Frank exudate in AC—endophthalmitis

Other Abnormal Contents in the Anterior Chamber

- Crystalline lens in AC **(Figs 5.10.1A and B)**
- Anterior chamber IOL **(Figs 5.10.2A to C)**
- Posterior chamber IOL in AC **(Fig. 5.10.3)**
- Lens cortical matter in AC **(Fig. 5.10.4)**
- Lens fragment in AC **(Fig. 5.10.5)**

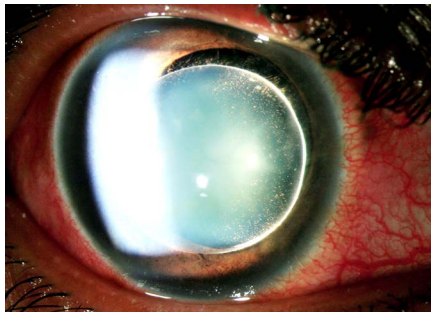


Fig. 5.10.1A: Crystalline lens in AC—microspherophakia

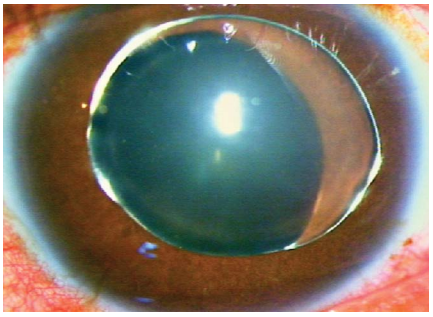


Fig. 5.10.1B: Crystalline lens in AC—traumatic

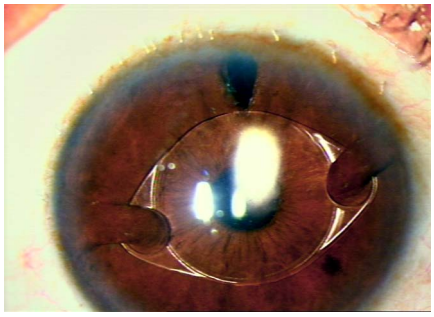


Fig. 5.10.2A: Anterior chamber IOL—Worst-Singh

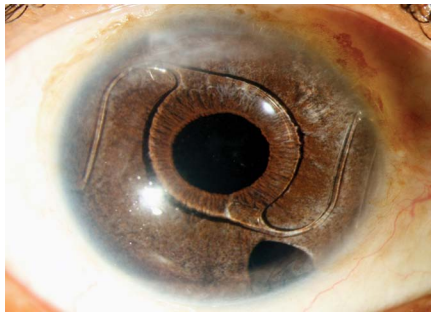


Fig. 5.10.2B: Anterior chamber IOL—Kelman multiflex

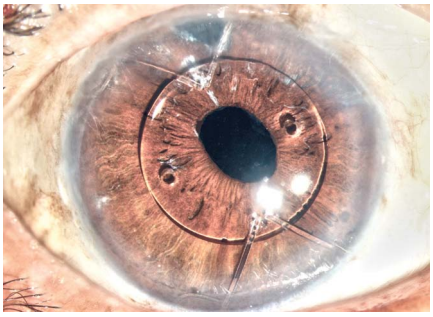


Fig. 5.10.2C: Anterior chamber—Shepard IOL

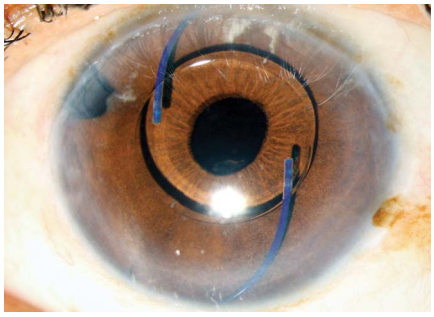


Fig. 5.10.3: PC IOL in AC

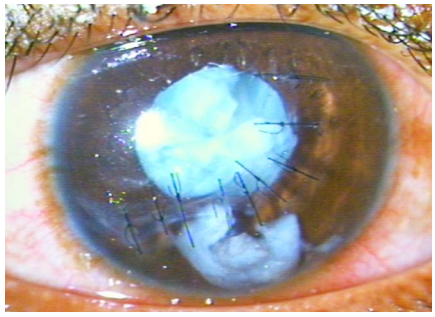


Fig. 5.10.4: Lens cortex

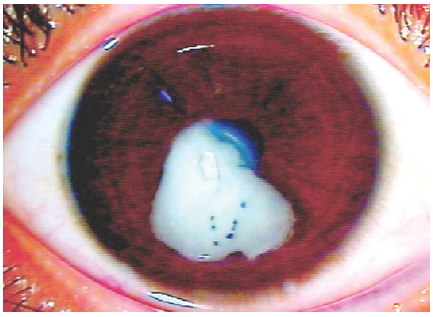


Fig. 5.10.5: Lens fragment

- Vitreous in AC **(Figs 5.10.6A to C)**
- Air bubble in AC **(Figs 5.10.7 and 5.10.8)**
- Silicone oil in AC **(Fig. 5.10.9 and 5.10.10)**
- Foreign body in AC **(Figs 5.10.11 to 5.10.13)**
- Eyelash in AC **(Fig. 5.10.14)**
- Fungal granuloma in AC **(Figs 5.10.15A and B)**
- Parasite dead or live: filaria **(Figs 5.10.16 to 5.10.19)**; cysticercosis **(Figs 5.10.20A and B)**
- Cholesterolosis bulbi **(Figs 5.10.21A and B)**
- Milky liquefied lens cortex **(Fig. 5.10.22)**
- Pseudoexfoliative material **(Fig. 5.10.23)**
- Caterpillar hair **(Figs 5.10.24A and B)**
- Cotton fibers in AC **(Figs 5.10.25 to 5.10.27)**
- End of tube of Ahmed glaucoma valve (AGV) **(Fig. 5.10.28)**
- Epithelial cyst (pearl) in AC **(Fig. 5.10.29)**.

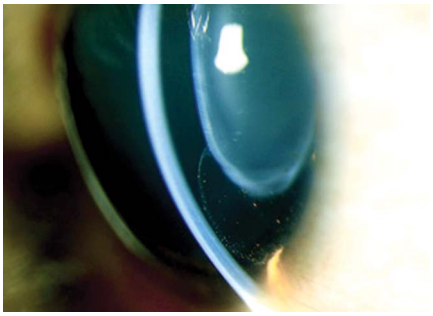


Fig. 5.10.6A: Vitreous in subluxation of lens

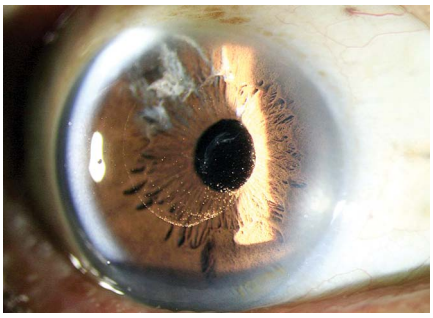


Fig. 5.10.6B: Vitreous harniation in AC in aphakia

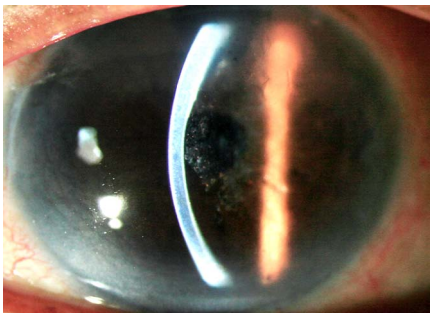


Fig. 5.10.6C: Vitreous in AC touching cornea in aphakia

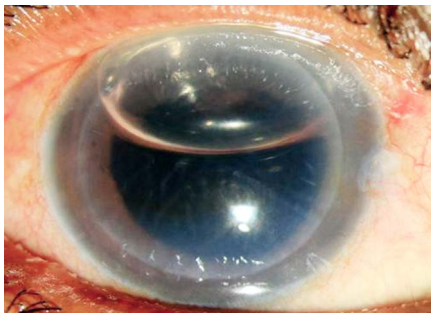


Fig. 5.10.7: Air bubble—postoperative in DSEK

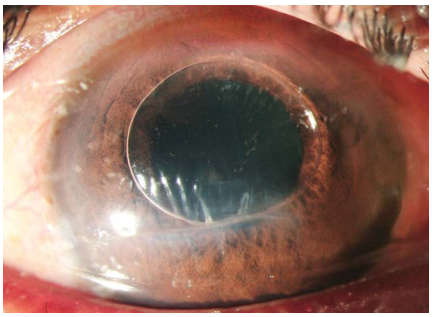


Fig. 5.10.8A: Air bubble—after open globe injury

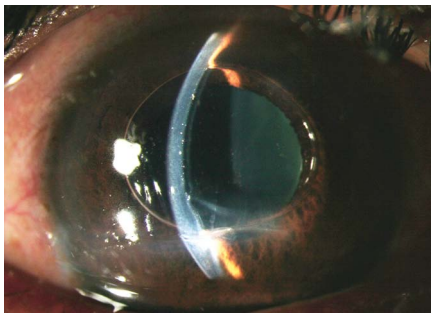


Fig. 5.10.8B: Air bubble—after open globe injury

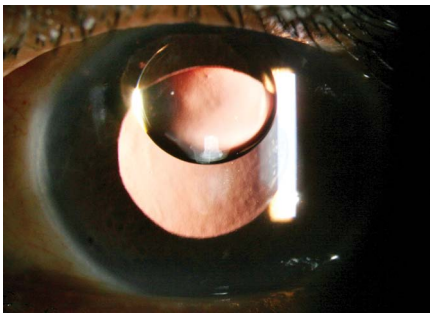


Fig. 5.10.9: Silicone oil—single bubble

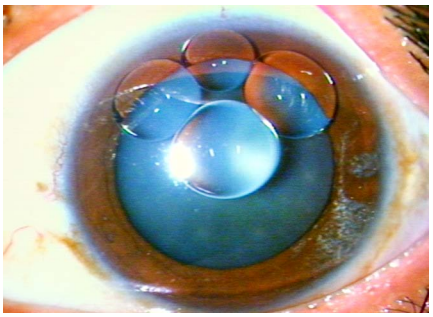


Fig. 5.10.10A: Silicone oil—multiple bubble

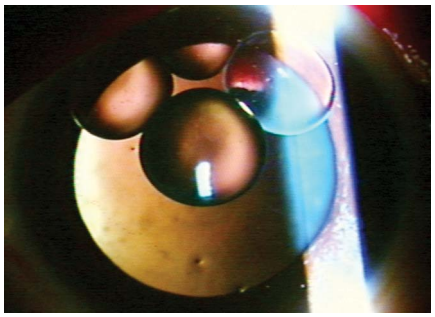


Fig. 5.10.10B: Silicone oil—multiple bubble

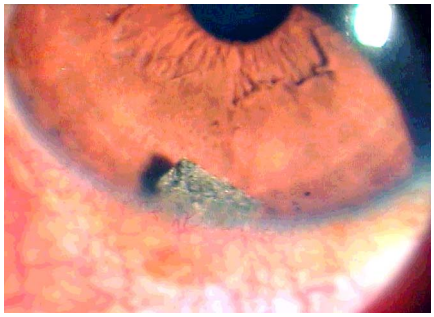


Fig. 5.10.11: Foreign body

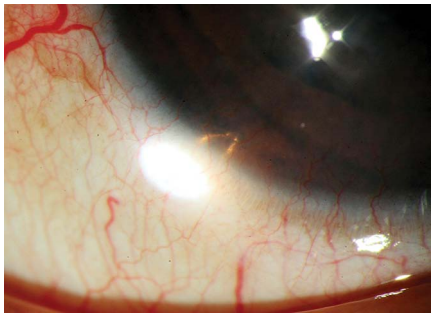


Fig. 5.10.12A: Gold foreign body in AC

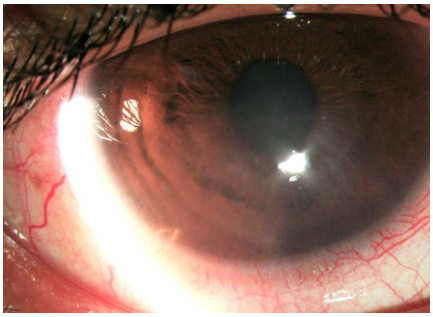


Fig. 5.10.12B: Gold foreign body in AC

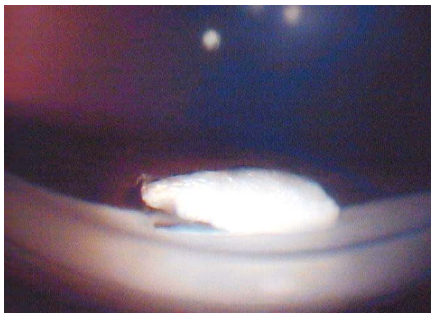


Fig. 5.10.13: Foreign body in angle of AC

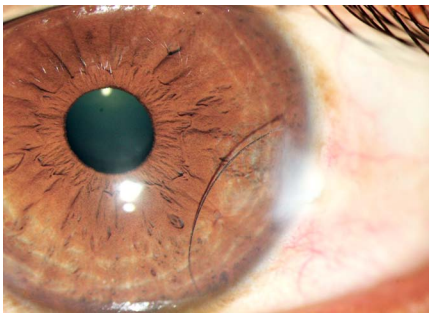


Fig. 5.10.14: Eyelash in AC—after open globe injury

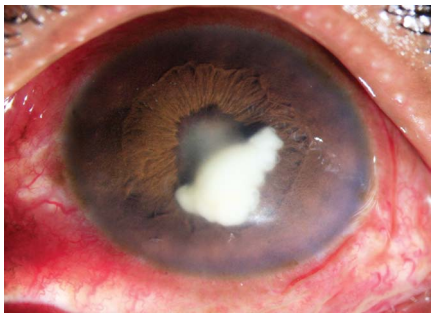


Fig. 5.10.15A: Fungal granuloma

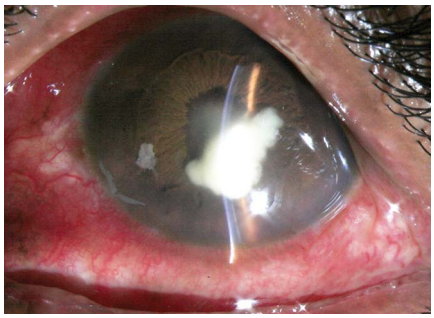


Fig. 5.10.15B: Fungal granuloma

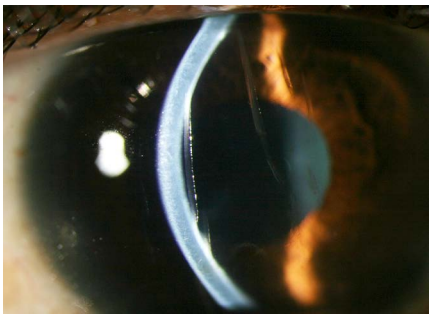


Fig. 5.10.16A: Dead filarial fossil in AC

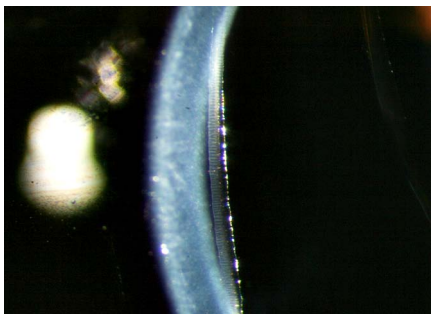


Fig. 5.10.16B: Dead filarial fossil in AC

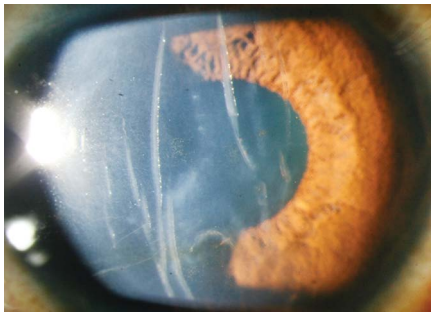


Fig. 5.10.17: Dead filarial fossil in AC

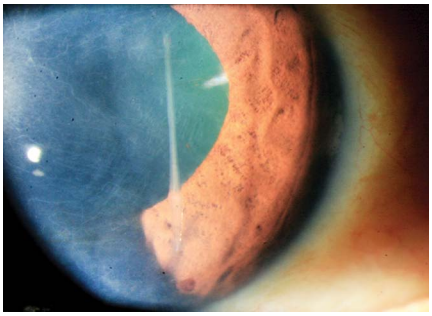


Fig. 5.10.18: Dead filarial fossil in AC

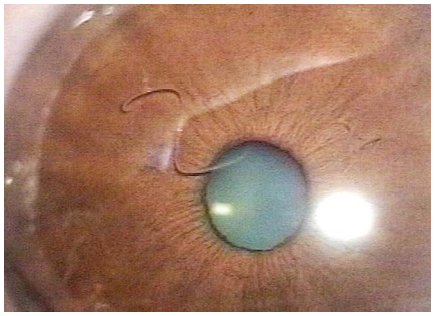


Fig. 5.10.19: Live microfilaria

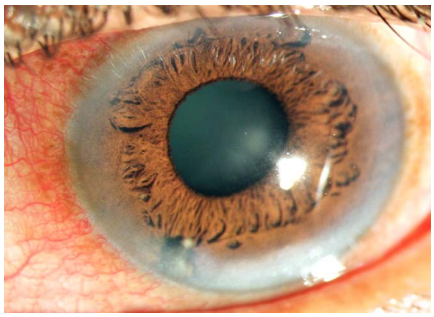


Fig. 5.10.20A: Live cysticercosis

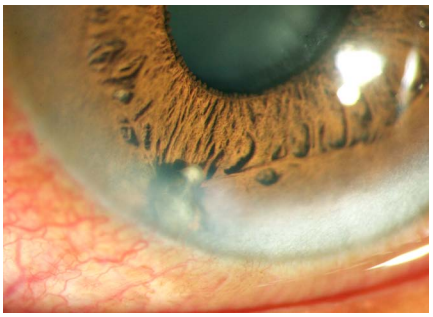


Fig. 5.10.20B: Live cysticercosis

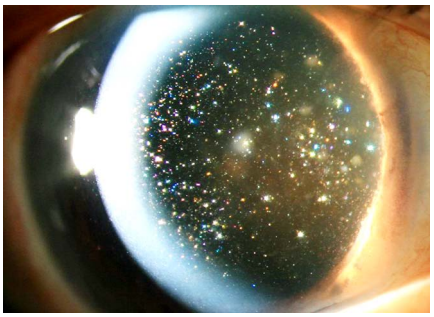


Fig. 5.10.21A: Cholesterolosis bulbi

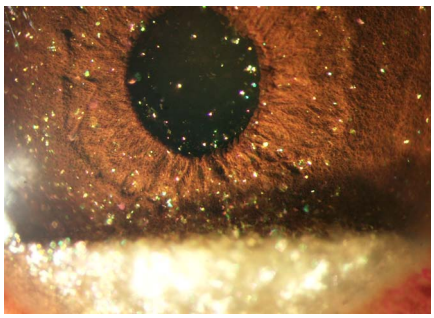


Fig. 5.10.21B: Cholesterolosis bulbi

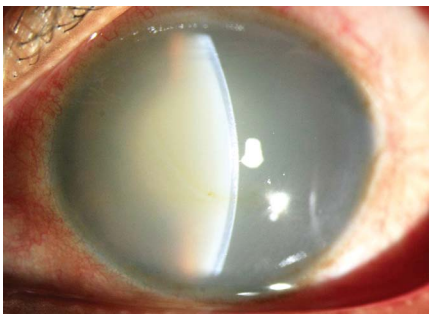


Fig. 5.10.22: Milky liquefied lens cortex in AC

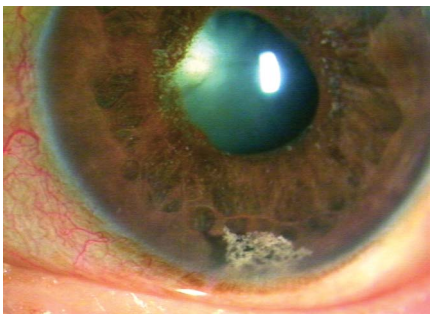


Fig. 5.10.23: Pseudoexfoliative materials

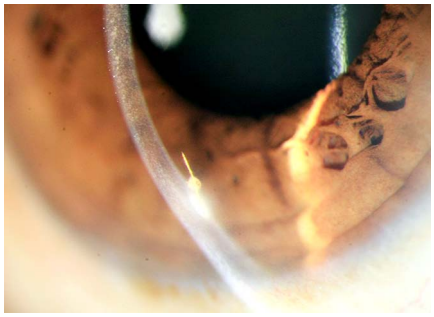


Fig. 5.10.24A: Caterpiller hair

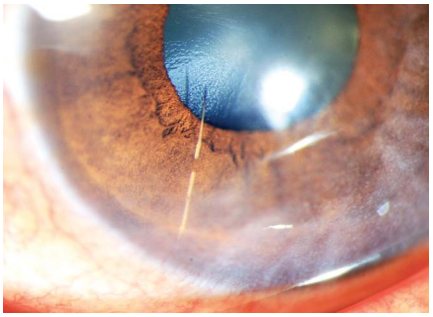


Fig. 5.10.24B: Caterpiller hair in AC piercing the iris

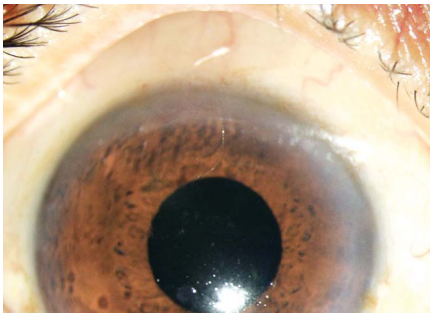


Fig. 5.10.25: Cotton fiber in AC and tunnel—note predonisolone acetate smeared end superiorly

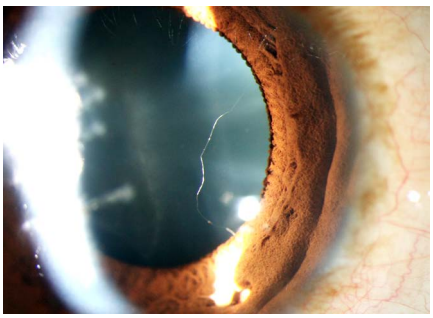


Fig. 5.10.26: Single cotton fiber in AC

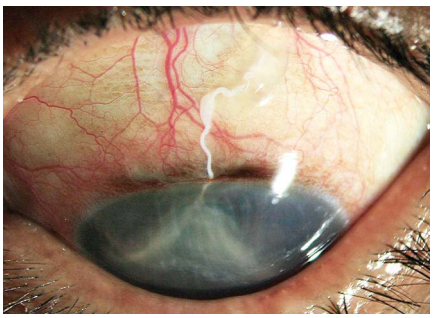


Fig. 5.10.27: Cotton fibers in AC—a large one

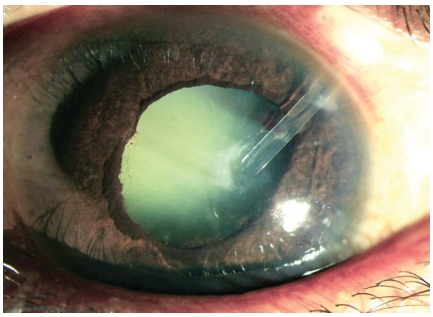


Fig. 5.10.28: End of tube of AGV

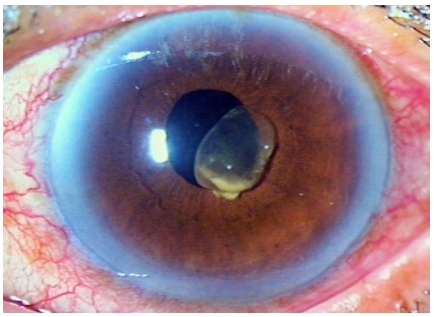


Fig. 5.10.29: Epithelial cyst

CHAPTER

6 Diseases of the Uvea

CHAPTER OUTLINE

Congenital Conditions

- Aniridia
- Coloboma
- Albinism
- Heterochromia of the iris
- Ectropion uveae

Anterior Uveitis (Iridocyclitis)

- Aqueous flare
- Anterior chamber
- Iris nodules
- Heterochromia (Cyclitis of Fuchs')
- Posterior synechiae

Choroiditis

- Toxoplasmosis
- Toxocariasis
- Vogt-Koyanagi-Harada (VKH) syndrome
- Sympathetic ophthalmitis
- Birdshot retinochoroiditis
- Endophthalmitis
- Panophthalmitis

Iridocorneal Endothelial (ICE) Syndromes

- Progressive essential iris atrophy
- Iris nevus syndrome (Cogan-Reese)
- Chandler's syndrome

Other Uveal Diseases

- Iridoschisis
- Iris atrophy
- Rubeosis iridis
- Primary choroidal sclerosis
- Iris cysts
- Gyrate atrophy of the choroid
- Choroideremia
- Angioid streaks
- Acute posterior multifocal placoid pigment epitheliopathy (APMPPE)
- Serpiginous (geographical) choroidopathy
- Choroidal detachment

Benign Uveal Lesions/Nodules/Mass

- Iris floccules
- Iris freckles
- Lisch nodules (spots)
- Brushfield spots
- Iris pearls
- Iris mammillations
- Iris nevus
- Choroidal hemangioma
- Benign melanocytoma (nevus)
- Choroidal osteoma

Malignant Uveal Tract Lesions

- Metastatic deposits of retinoblastoma
- Malignant melanoma of the iris
- Medulloepithelioma (diktyoma)
- Malignant melanoma of the ciliary body
- Malignant melanoma of the choroid
- Metastatic carcinoma of the uveal tract

Congenital Conditions

Aniridia

- Whole of the iris is appeared to be missing on external examination **(Figs 6.1.1A and B)**
- Ciliary processes and suspensory ligaments of the lens are visible **(Figs 6.1.2A and B)**
- In some cases, there is associated limbal stem cell deficiency (LSCD) **(Figs 6.1.3A and B)**
- Subluxation of lens and secondary glaucoma in 25% of the cases **(Fig. 6.1.4)**
- In *partial aniridia*, iris is missing in a sector with visibility of ciliary process and suspensory ligaments **(Figs 6.1.5 and 6.1.6)**
- Aniridia and Wilms' tumor are associated with deletion of the short arm of chromosome-11 (Miller's syndrome)
- *Treatment:* In total aniridia, iris-painted contact lens; and care of the secondary glaucoma and subluxation.

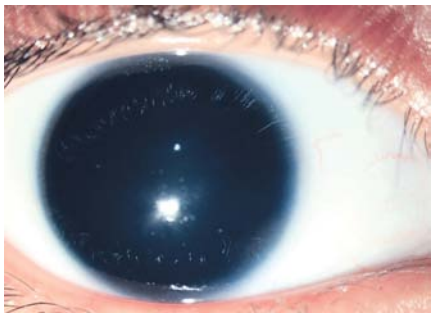


Fig. 6.1.1A: Total aniridia—RE

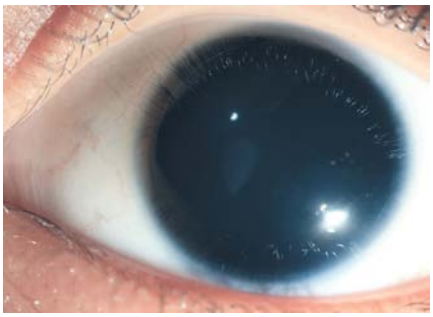


Fig. 6.1.1B: Total aniridia—LE

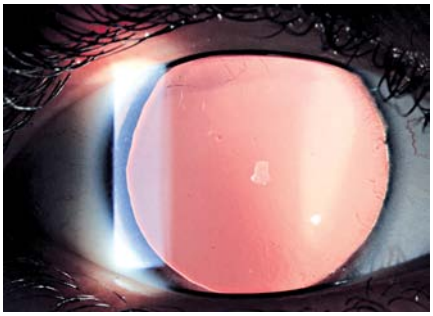


Fig. 6.1.2A: Total aniridia—RE in retroillumination

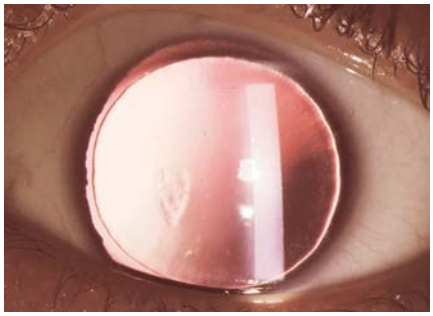


Fig. 6.1.2B: Total aniridia—LE in retroillumination

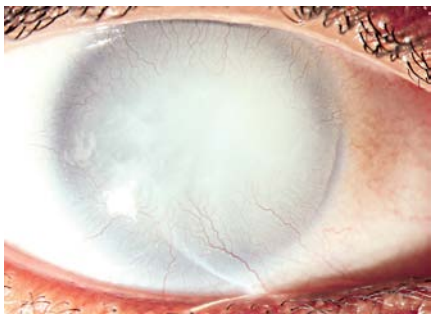


Fig. 6.1.3A: Total aniridia with mature cataract with total LSCD—RE

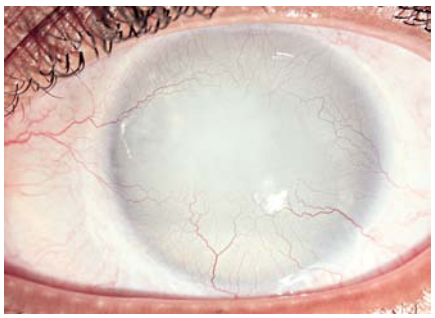


Fig. 6.1.3B: Total aniridia with hypermature Morgagnian cataract with total LSCD—LE

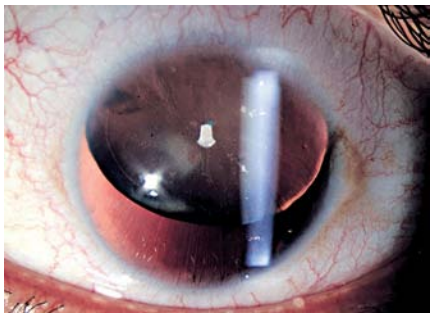


Fig. 6.1.4: Aniridia with ectopia lentis—LE

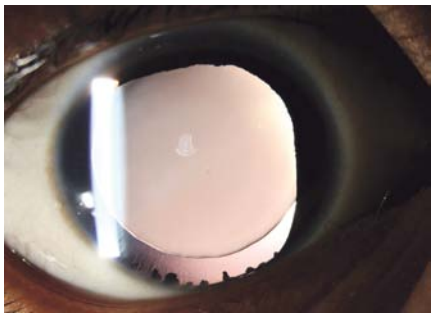


Fig. 6.1.5A: Aniridia partial—RE

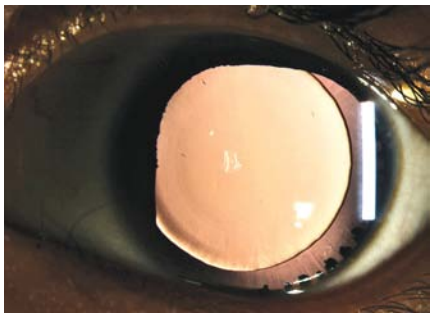


Fig. 6.1.5B: Aniridia partial—LE

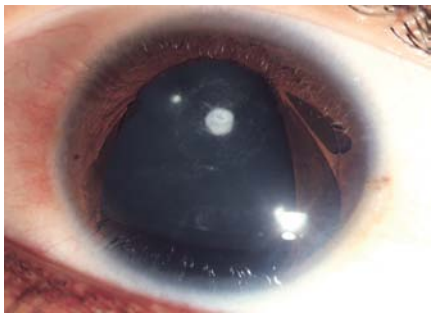


Fig. 6.1.6A: Partial aniridia with anterior polar cataract

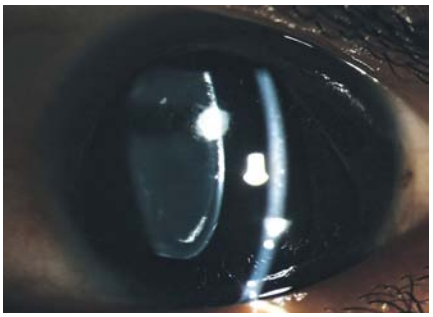


Fig. 6.1.6B: Partial aniridia with anterior polar cataract

Coloboma

- Development imperfect closure of the fetal fissure
- Lower nasal sector of the eye
- Coloboma may be typical or atypical
- Extends from the pupil to the optic nerve **(Figs 6.2.1A and B)**
- May stop short of the optic nerve, iris or else or even partitioned by islands of normal tissue **(Figs 6.2.2A to C)**

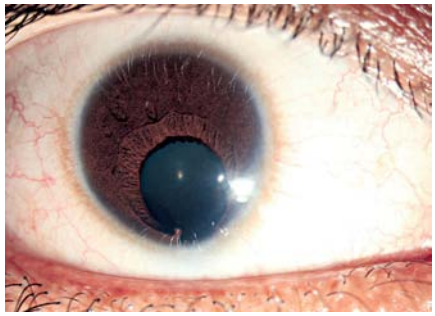


Fig. 6.2.1A: Typical coloboma—RE

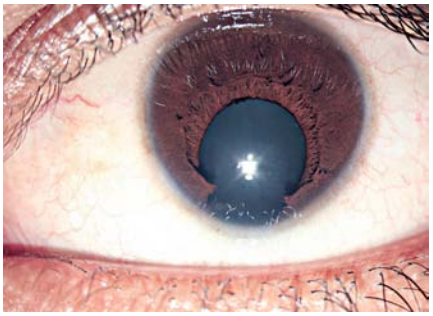


Fig. 6.2.1B: Typical coloboma—LE

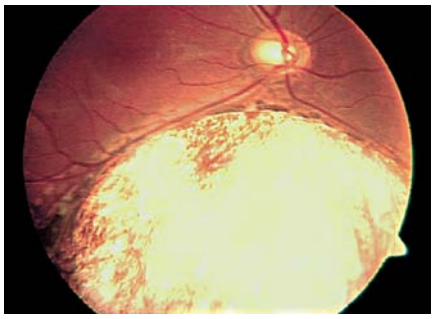


Fig. 6.2.2A: Choroidal coloboma

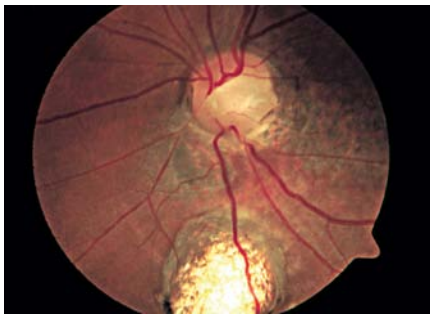


Fig. 6.2.2B: Isolated choroidal coloboma

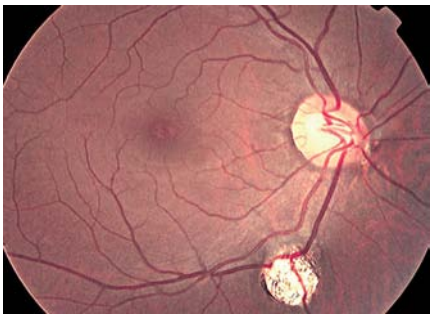


Fig. 6.2.2C: Isolated choroidal coloboma

- *Typical coloboma of the iris*
  - Pear shaped with broad base towards the pupillary margin
  - It may be *complete*; and usually associated with microcornea or severe microphthalmos **(Figs 6.2.3A and B)** or with normal cornea **(Fig. 6.2.4)**
  - Or it may be *incomplete* **(Figs 6.2.5A and B)**
- *Atypical coloboma of the iris*
  - Isolated coloboma with or without lens coloboma which occurs at any meridian **(Fig. 6.2.6)**
  - May be congenital or iatrogenic **(Figs 6.2.7A and B)**
- *Treatment:* Hypermetropic correction; cataract surgery is difficult, because of small cornea and shallow AC and less space. Hard cataract is rather common in this group of eyes. A CTR in-the-bag during IOL implantation is always a better option.

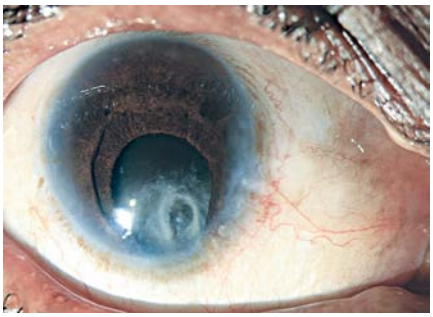


Fig. 6.2.3A: Typical colobomatous microphthalmos—RE

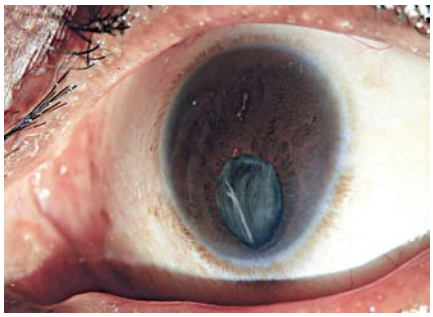


Fig. 6.2.3B: Typical colobomatous microphthalmos—LE

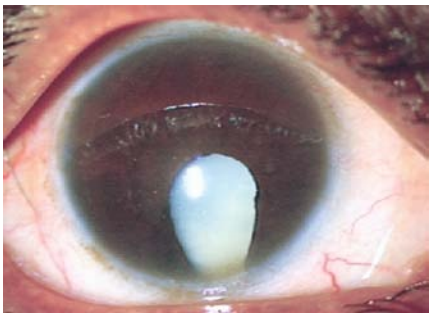


Fig. 6.2.4: Typical iris coloboma with normal cornea

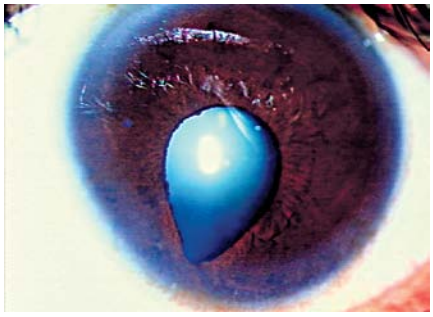


Fig. 6.2.5A: Incomplete iris coloboma

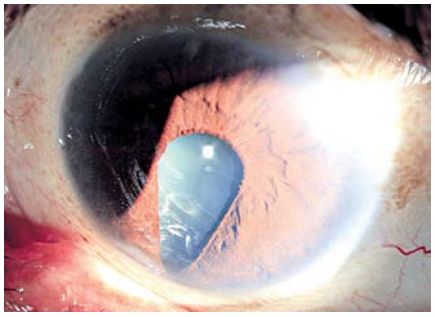


Fig. 6.2.5B: Incomplete coloboma

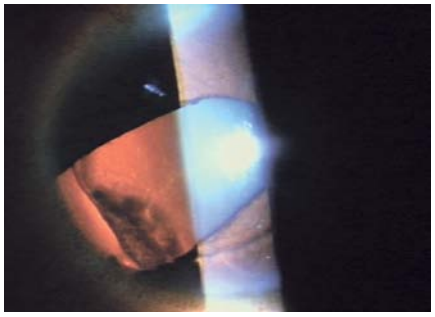


Fig. 6.2.6: Atypical iris coloboma—congenital

Albinism

- Hereditary disorder in which there is absence or reduction of melanin pigmentation throughout the body **(Figs 6.3.1A and B)**
- *Two main types*: Oculocutaneous and ocular
- Poliosis is common; iris looks pink and totally translucent, owing to lack of pigment **(Figs 6.3.2 and 6.3.3)**

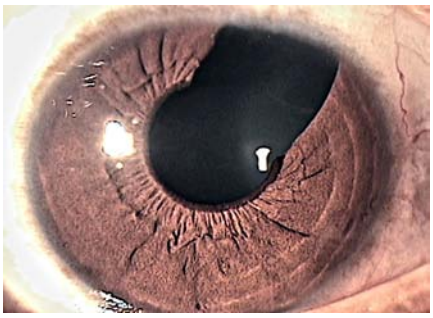


Fig. 6.2.7A: Atypical iris coloboma—surgical

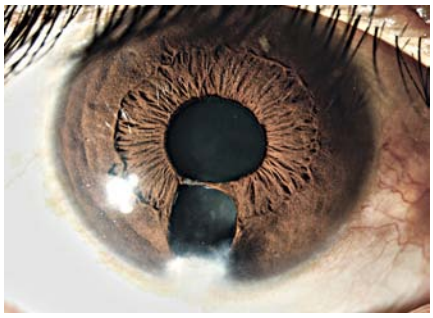


Fig. 6.2.7B: Atypical bridge iris coloboma—surgical

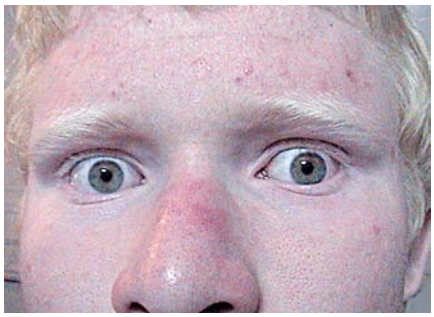


Fig. 6.3.1A: Oculocutaneous albinism—iris spread

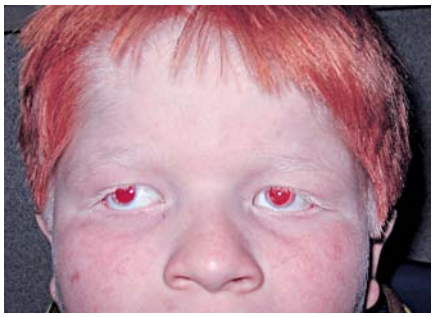


Fig. 6.3.1B: Oculocutaneous albinism—orange-pink fundal glow

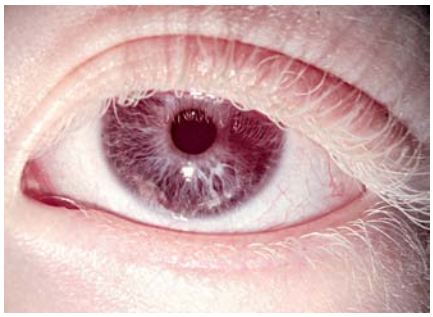


Fig. 6.3.2: Oculocutaneous albinism—poliosis and pink iris

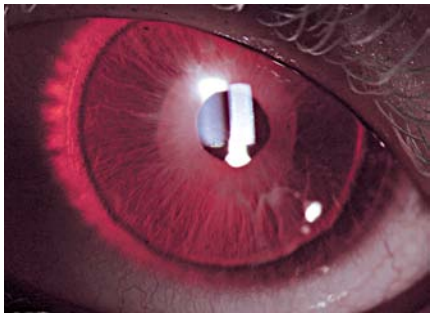


Fig. 6.3.3A: Oculocutaneous albinism—iris transillumination

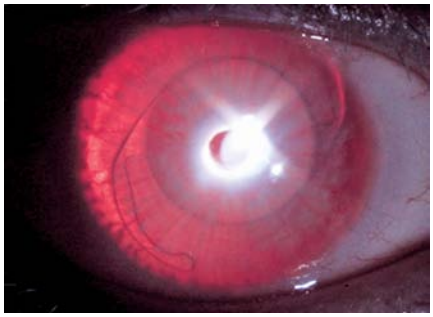


Fig. 6.3.3B: Single-piece PC IOL seen easily through transilluminated iris

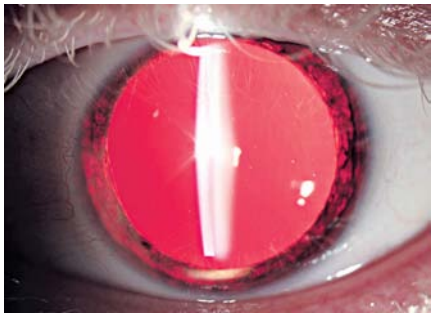


Fig. 6.3.4: Oculocutaneous albinism—orange-pink fundal glow

- Fundus appears orange-pink in color **(Fig. 6.3.4)**. The retinal and choroidal vessels are difficult to differentiate **(Fig. 6.3.5)**
- *Partial albinism* **(Fig. 6.3.6)**
  - Absence of pigment is limited to the choroids and retina
  - Iris is brown or blue colored.

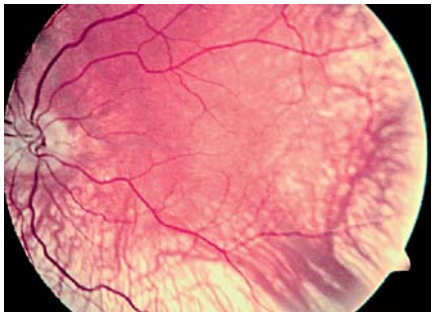


Fig. 6.3.5: Oculocutaneous albinism—fundus appearance

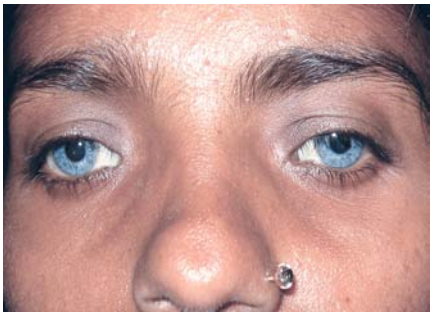


Fig. 6.3.6: Partial albinism—blue iris with pink fundal glow

Heterochromia of Iris

- Two irides show a significant difference in color **(Figs 6.4.1 to 6.4.3)**
- *Heterochromia iridis*: Usually a sector, shows difference in color from the remainder **(Fig. 6.4.4)**

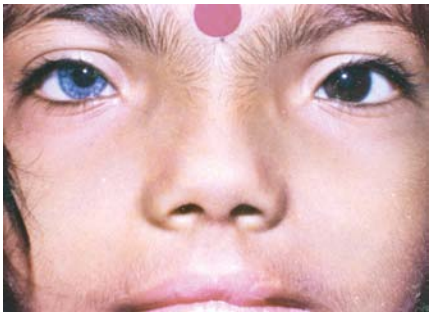


Fig. 6.4.1: Heterochromia—Waardenburg syndrome

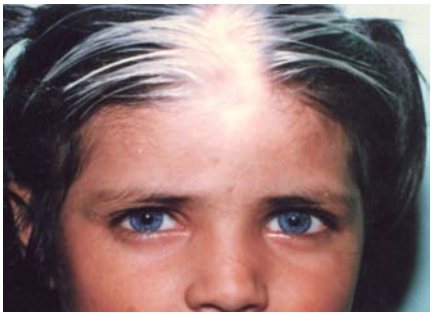


Fig. 6.4.2: Heterochromia—Waardenburg syndrome (sister of previous Figure)

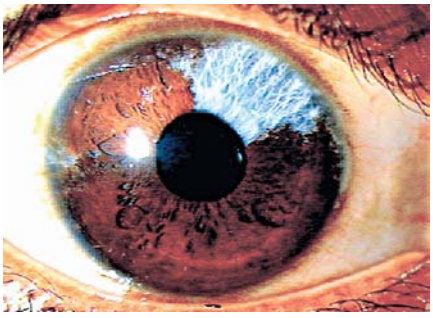


Fig. 6.4.3A: Heterochromic heterochromia—RE

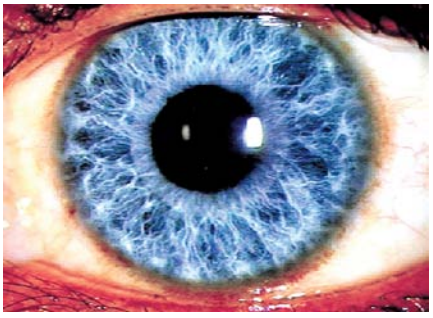


Fig. 6.4.3B: Hypochromic heterochromia—LE

- *Hypochromic* **(Figs 6.4.5 and 6.4.6)**
  - Eye with lighter-colored iris is abnormal
- *Hyperchromic* **(Figs 6.4.7A and B)**
  - Iris on the side of the disease is darker than its fellow.

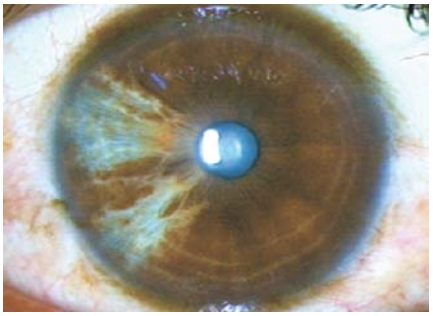


Fig. 6.4.4: Heterochromia iridis

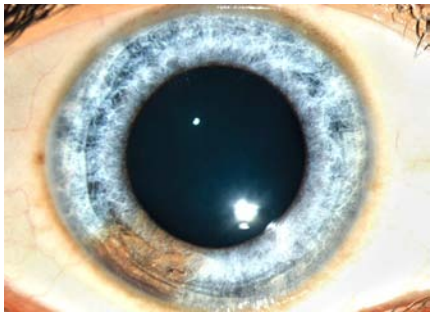


Fig. 6.4.5: Hypochromic heterochromia

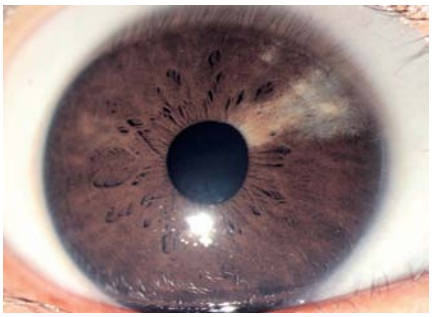


Fig. 6.4.6: Hypochromic heterochromia

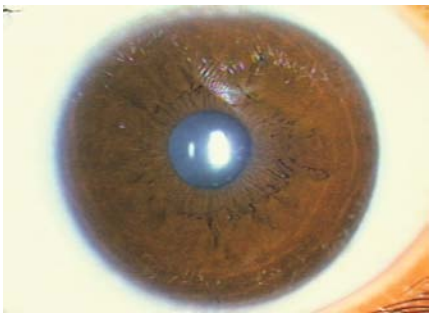


Fig. 6.4.7A: Hyperchromic heterochromia—normal right eye

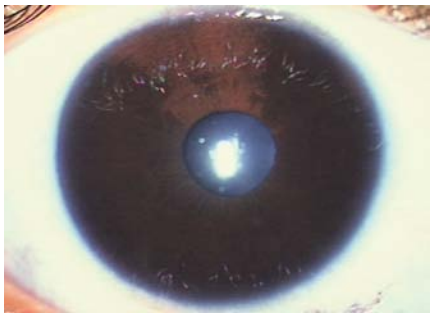


Fig. 6.4.7B: Hyperchromic heterochromia—left eye

Ectropion Uveae

- Pigmented iris surface appears anteriorly, especially along the pupillary border
- *Causes:*
  - Isolated congenital condition: nonprogressive **(Fig. 6.5.1)**
  - Axenfeld-Rieger's anomaly **(Fig. 6.5.2)**
  - Neurofibromatosis **(Fig. 6.5.3)**
  - Rubeosis iridis **(Figs 6.5.4A and B)**

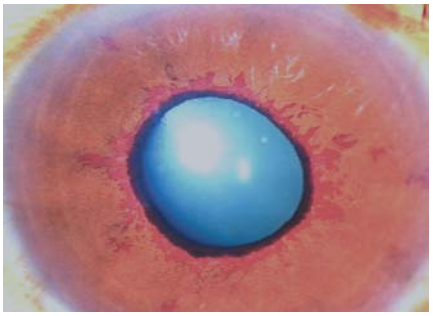


Fig. 6.5.1: Ectropion uveae—congenital

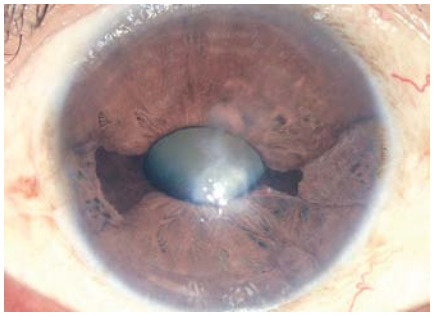


Fig. 6.5.2: Ectropion uveae—Axelfeld-Rieger's anomaly

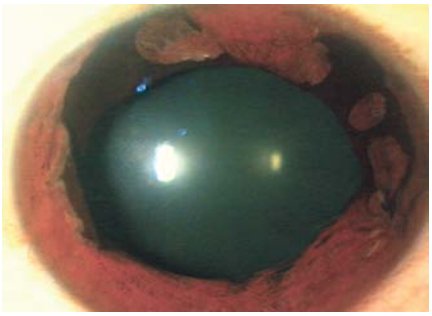


Fig. 6.5.3: Ectropion uveae—neurofibromatosis

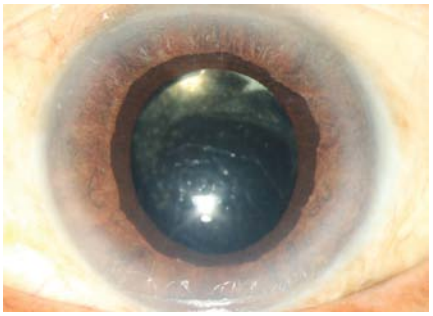


Fig. 6.5.4A: Ectropion uveae—rubeosis iridis

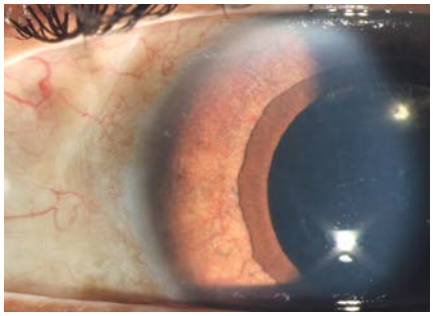


Fig. 6.5.4B: Ectropion uveae—rubeosis iridis

  - Iris coloboma **(Fig. 6.5.5)**
  - Siderosis bulbi **(Fig. 6.5.6)**
- Patients should be investigated for glaucoma.

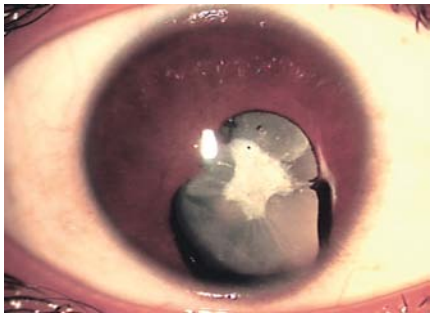


Fig. 6.5.5: Ectropion uveae—coloboma iris

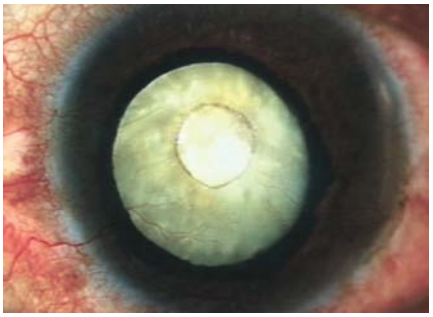


Fig. 6.5.6: Ectropion uveae—siderosis

Anterior Uveitis (Iridocyclitis)

- May be acute nongranulomatous **(Fig. 6.6.1)** or chronic granulomatous **(Figs 6.6.2A and B)** type
- Circumcorneal (ciliary) congestion **(Fig. 6.6.3)**
- *Keratic precipitates (KPs):* They are cellular deposits on the corneal endothelium
  - *Distribution*: They arrange in a base-down triangular area at the lower part of the cornea—called, Arlt's triangle **(Figs 6.6.4A and B)**
  - In Fuchs' cyclitis, they are scattered throughout the endothelium **(Fig. 6.6.5)**

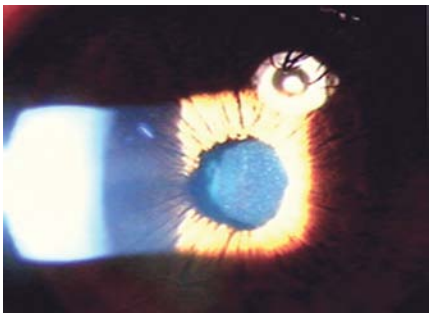


Fig. 6.6.1: Iridocyclitis—acute nongranulomatous type

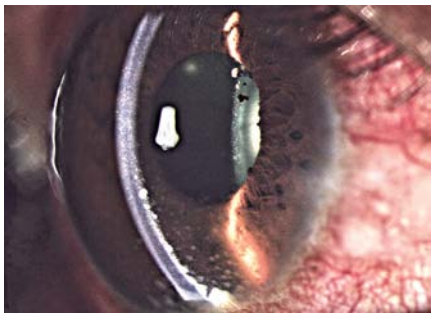


Fig. 6.6.2A: Iridocyclitis—chronic granulomatous

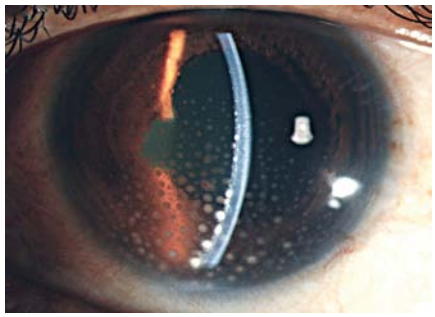


Fig. 6.6.2B: Chronic granulomatous uveitis

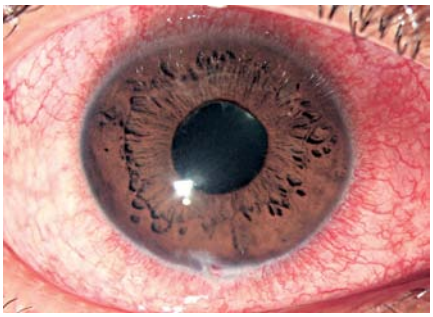


Fig. 6.6.3: Iridocyclitis—ciliary congestion

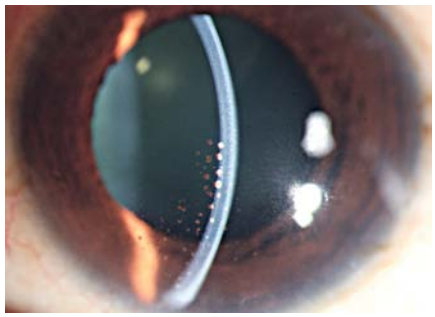


Fig. 6.6.4A: KPs—Arlt's triangle

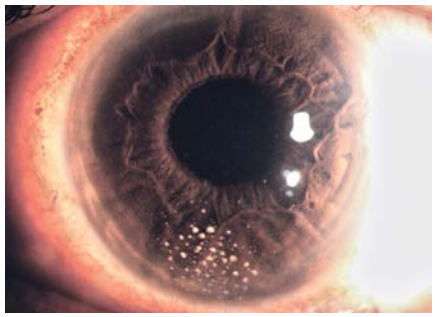


Fig. 6.6.4B: KPs—Arlt's triangle

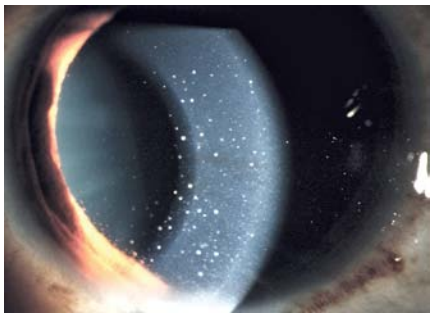


Fig. 6.6.5: Fine KPs—scattered all over in Fuchs' cyclitis

- *Size:* KPs may be small **(Fig. 6.6.6)**, medium **(Figs 6.6.7A and B)**, large **(Fig. 6.6.8)** and 'mutton fat' **(Fig. 6.6.9)** types
- Small and medium size KPs are due to deposition of lymphocytes and plasma cells and they are seen in *non-granulomatous* or acute uveitis **(Fig. 6.6.10)**
- Large KPs are due to deposition of macrophage and epithelioid cells, and they are seen in *granulomatous* uveitis, as in tuberculosis or sarcoidosis **(Fig. 6.6.11)**
- *Age:* KPs may be fresh or old **(Fig. 6.6.12)**.

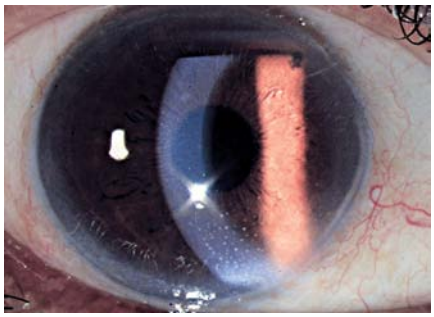


Fig. 6.6.6: Small KPs

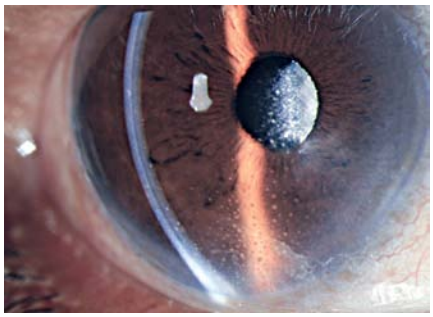


Fig. 6.6.7A: Medium KPs

Fig. 6.6.7B: Medium KPs

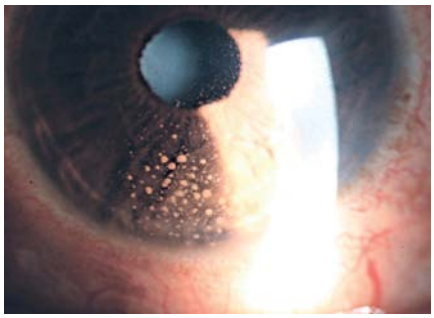


Fig. 6.6.8: Large KPs

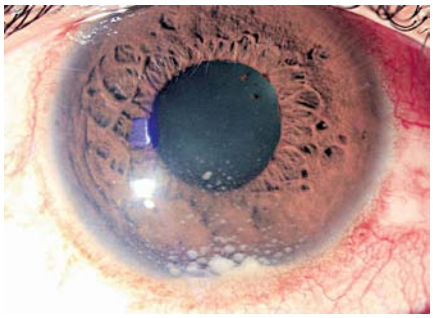


Fig. 6.6.9: Chronic iridocyclitis—mutton fat KPs

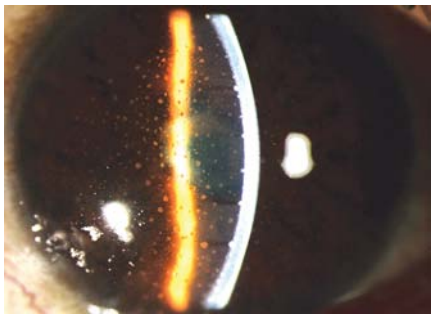


Fig. 6.6.10: Small to medium KPs

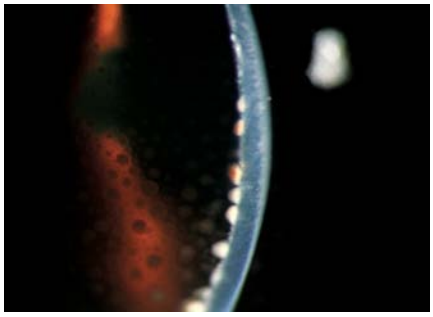


Fig. 6.6.11: Mutton fat KPs—tubercular

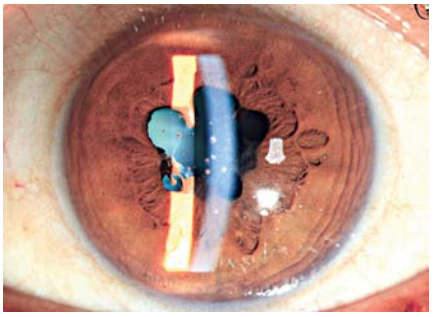


Fig. 6.6.12: Old KPs

Aqueous Flare

- Proteins leaks through the damaged capillaries, causing a Tyndall effect **(Figs 6.7.1 and 6.7.2).**

Anterior Chamber

- *Aqueous cells* **(Figs 6.8.1 and 6.8.2)**
- *Hypopyon:* Classically seen in Behcet's syndrome, infectious cases (herpetic uveitis) or in severe cases of acute iridocyclitis **(Figs 6.8.3A to C)**
- *Hyphema in herpetic uveitis* **(Figs 6.8.4A and B)**

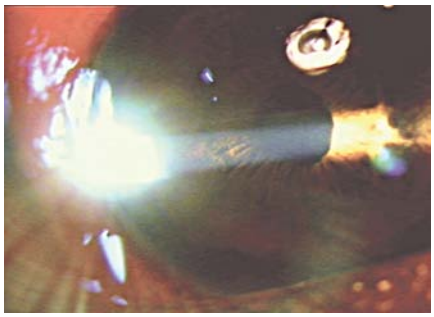


Fig. 6.7.1: Aqueous flare

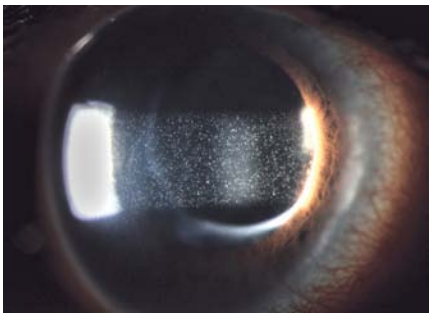


Fig. 6.7.2: Aqueous flare and cells

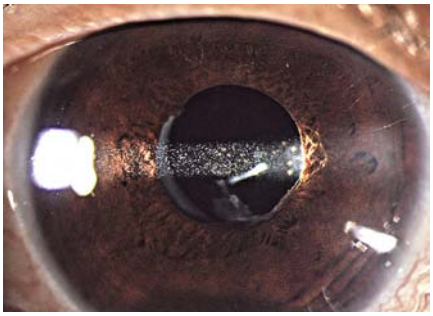


Fig. 6.8.1: Aqueous cells and flare

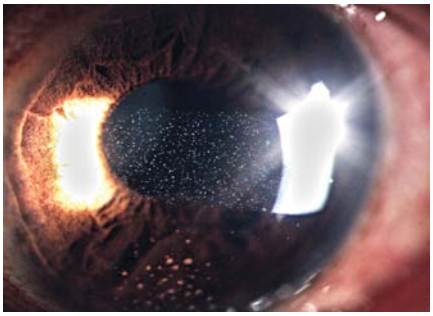


Fig. 6.8.2: Aqueous cells

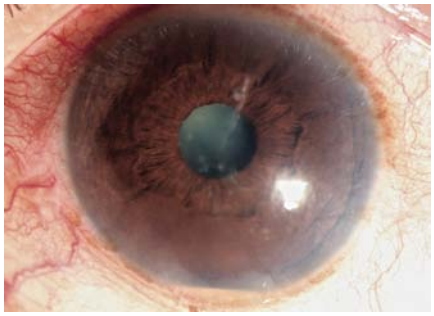


Fig. 6.8.3A: Acute iridocyclitis—hypopyon

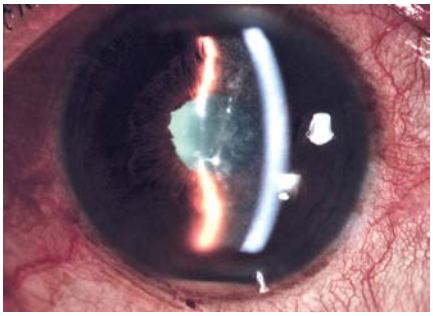


Fig. 6.8.3B: Aqueous cells—hypopyon

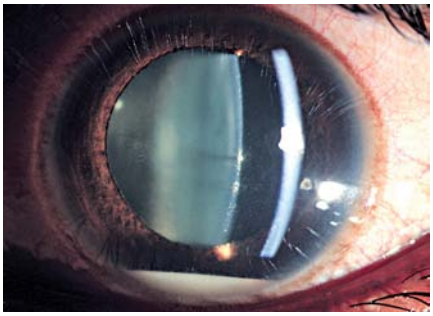


Fig. 6.8.3C: Acute iridocyclitis—hypopyon

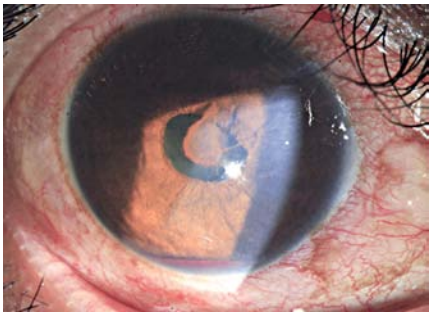


Fig. 6.8.4A: Herpetic iridocyclitis—hyphema

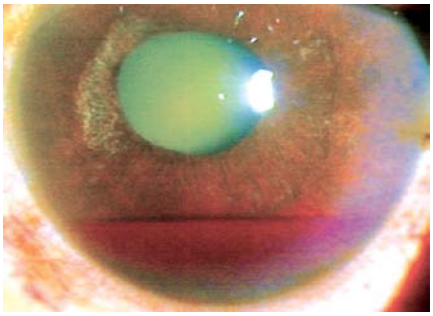


Fig. 6.8.4B: Herpetic iridocyclitis—hyphema

- *Deep and irregular:* In posterior synechiae **(Figs 6.8.5A to B)**
- *Funnel-shaped:* In iris bombe **(Figs 6.8.6A and B)**.

Iris Nodules

- *Koeppe's nodules*: At the pupillary border and smaller in size **(Fig. 6.9.1)**
- *Busacca nodules*: On the surface of the iris, away from the pupil **(Figs 6.9.2A and B)**.

Heterochromia (Cyclitis of Fuchs')

- Usually unilateral in young patients between 20 to 40 years of age
- A low-grade iridocyclitis without much symptoms
- Associated with fine diffuse KPs, heterochromia of iris (hypochromic) and cataract formation **(Figs 6.9.3A and B)**; without any posterior synechiae
- Prognosis for cataract surgery is usually good.

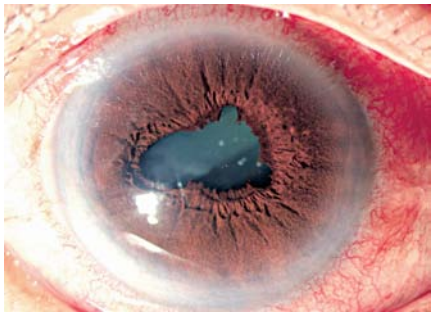


Fig. 6.8.5A: Acute iridocyclitis—partial posterior synechiae

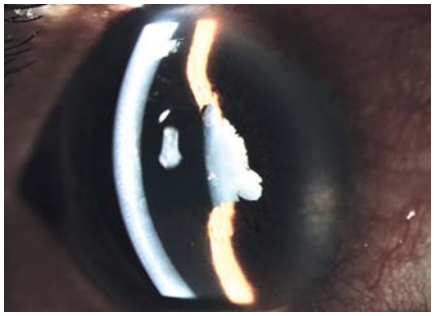


Fig. 6.8.5B: Acute iridocyclitis—partial posterior synechiae

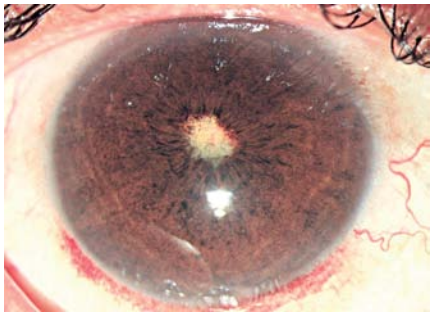


Fig. 6.8.6A: Acute iridocyclitis—posterior ring synechiae and iris bombe

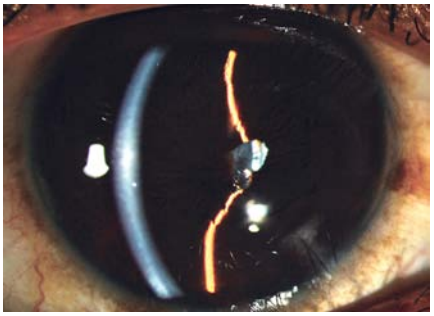


Fig. 6.8.6B: Acute iridocyclitis—iris bombe

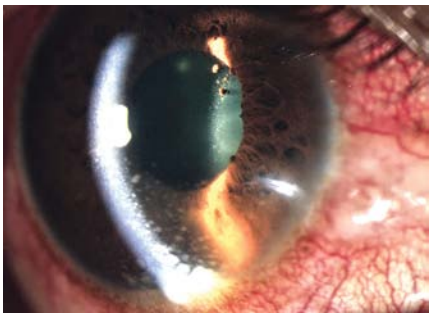


Fig. 6.9.1: Koeppe's nodules

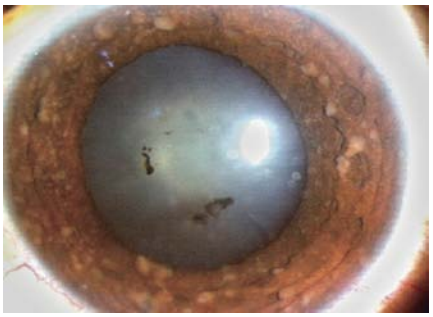


Fig. 6.9.2A: Busacca nodules

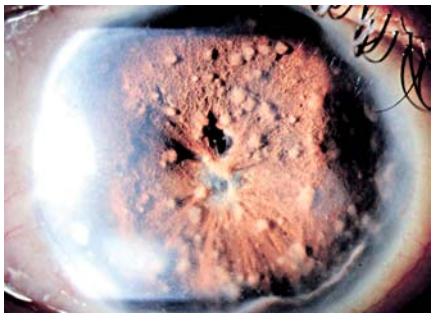


Fig. 6.9.2B: Busacca and koeppe's nodules—extensive

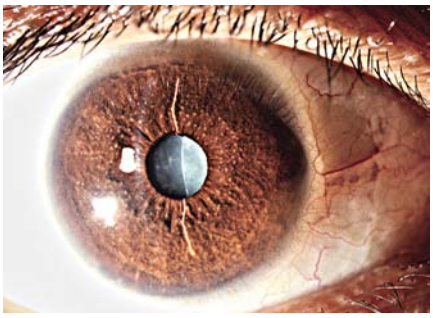


Fig. 6.9.3A: Heterochromic cyclitis of Fuchs' with cataract—RE

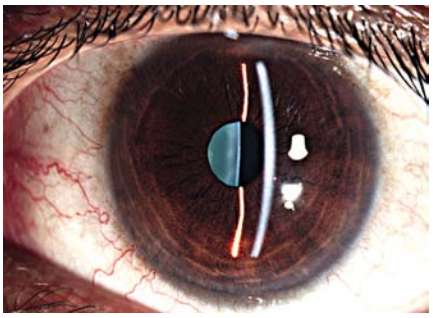


Fig. 6.9.3B: Heterochromic cyclitis of Fuchs' normal eye—LE

Posterior Synechiae

- Ring (annular) synechiae iris bombe **(Fig. 6.10.1 and** *see* **Figs 6.8.6A and B)**
- Irregular or 'festooned pupil' **(Figs 6.10.2 and 6.10.3)**
- *Occlusio pupillae* **(Figs 6.10.4A and B)**
- Complicated cataract **(Figs 6.10.5 and 6.10.6)**
- Band-shaped keratopathy (BSK) **(Figs 6.10.7 and 6.10.8)**; more common in children with juvenile rheumatoid arthritis, and chronic iridocyclitis
- Membrane formation **(Figs 6.10.9 and 6.10.10)**
- Pigments on anterior lens surface **(Figs 6.10.11 and 6.10.12).**

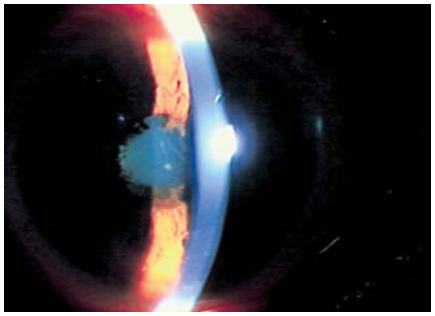


Fig. 6.10.1: Ring synechiae—iris bombe

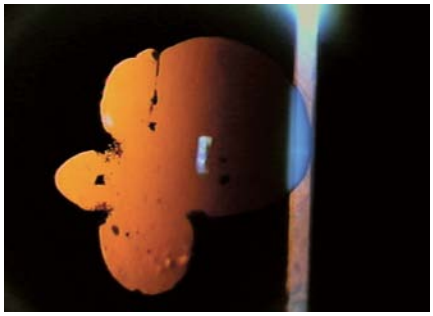


Fig. 6.10.2: Irregular synechiae—festooned pupil

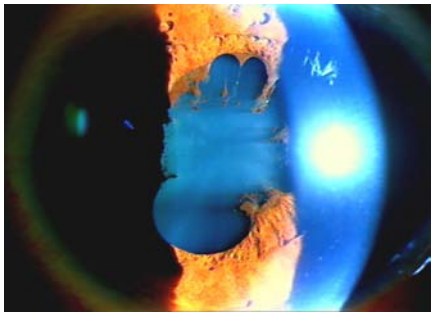


Fig. 6.10.3: Festooned pupil

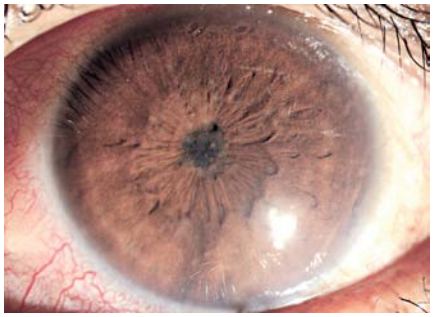


Fig. 6.10.4A: Occlusio pupillae

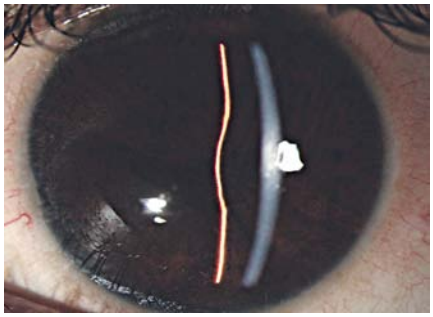


Fig. 6.10.4B: Occlusio pupillae

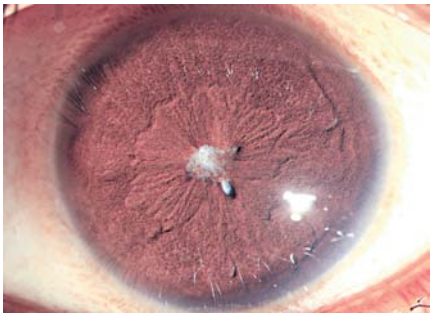


Fig. 6.10.5: Healed iritis—complicated cataract

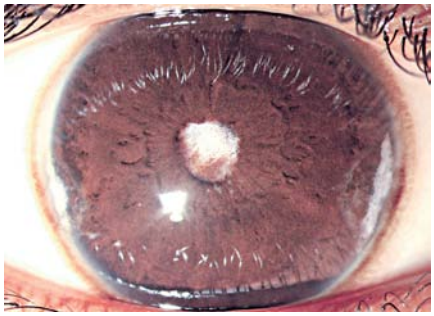


Fig. 6.10.6: Chronic iridocyclitis—complicated cataract, posterior

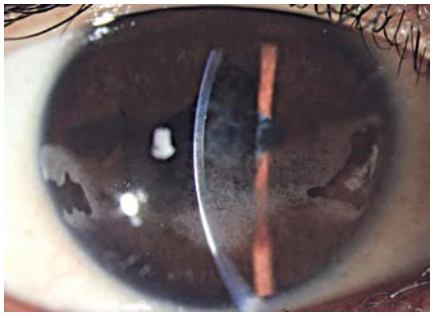


Fig. 6.10.7A: Chronic uveitis in idiopathic rheumatoid arthritis

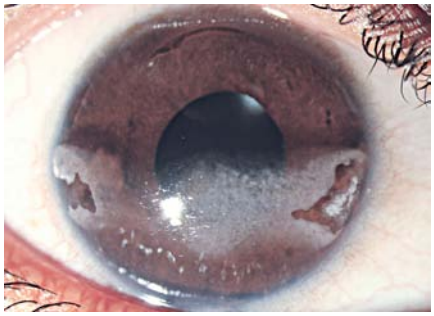


Fig. 6.10.7B: Chronic uveitis in idiopathic rheumatoid arthritis

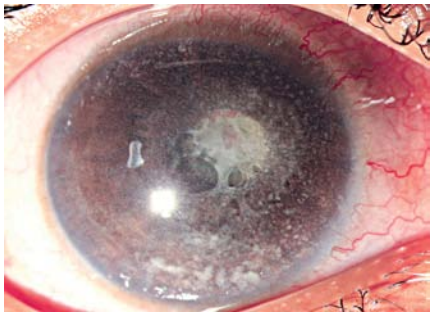


Fig. 6.10.8A: BSK and complicated cataract in chronic iridocyclitis

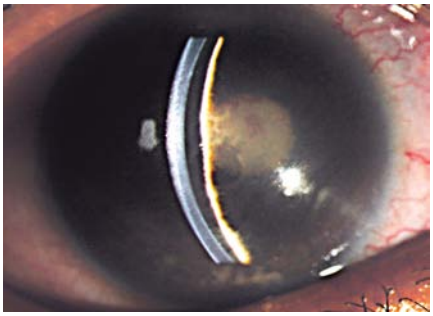


Fig. 6.10.8B: BSK and complicated cataract in chronic iridocyclitis

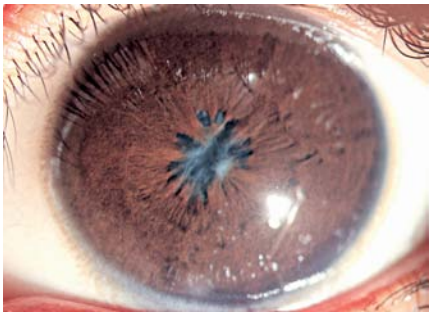


Fig. 6.10.9: Healed uveitis—membrane formation

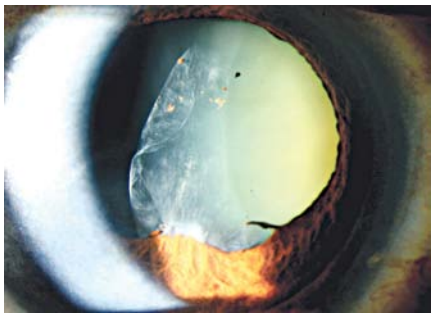


Fig. 6.10.10: Healed uveitis—membrane formation

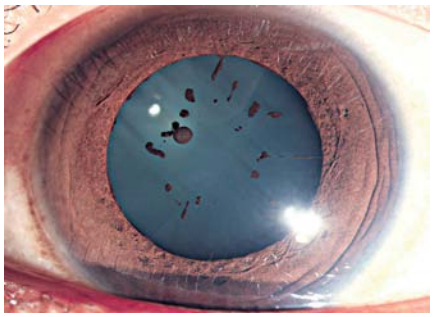


Fig. 6.10.11A: Healed uveitis—pigments on anterior lens surface

Fig. 6.10.11B: Healed uveitis—pigments on anterior lens surface

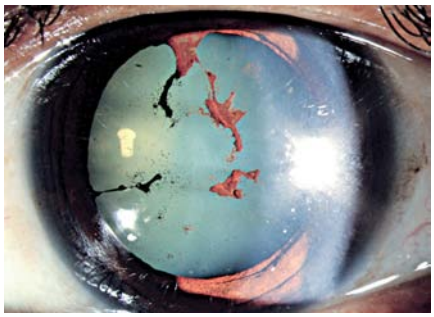


Fig. 6.10.12: Healed uveitis—pigments on anterior lens surface—broken posterior synechiae

Choroiditis

- *Active lesions:* Well defined yellowish-white patches of chorioretinal inflammation **(Figs 6.11.1A and B)**

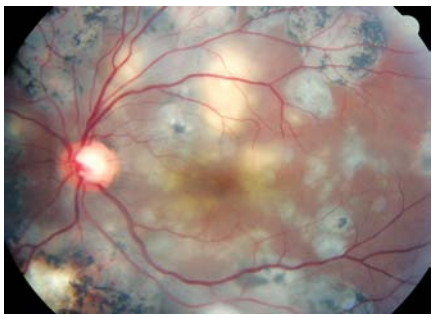


Fig. 6.11.1A: Active choroiditis

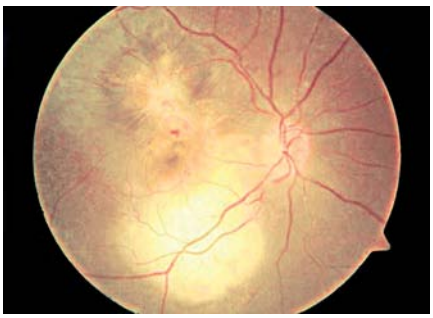


Fig. 6.11.1B: Active tubercular choroiditis

- It may be disseminated **(Figs 6.11.2A and B)**, central **(Figs 6.11.3A and B)** or juxtapapillary **(Fig. 6.11.4)**
- *Inactive lesions:* Well defined white patches of chorioretinal atrophy with pigmented margins **(Figs 6.11.5 and 6.11.6)**
- *Perivasculitis:* Periphlebitis is more common and it may be seen as 'candle-wax dripping' in sarcoidosis **(Figs 6.11.7A and B).**

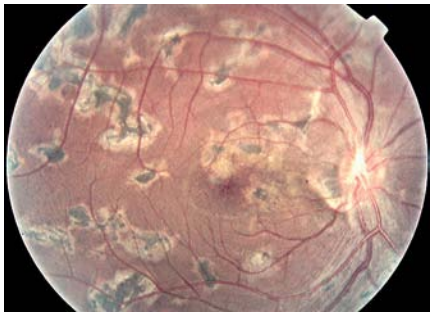


Fig. 6.11.2A: Active choroiditis

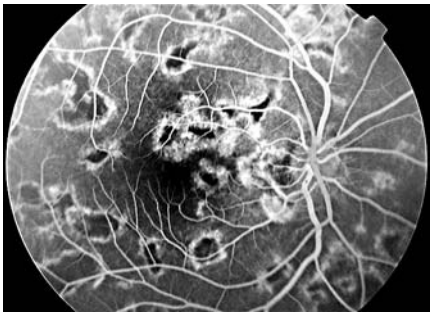


Fig. 6.11.2B: Active choroiditis—FFA findings

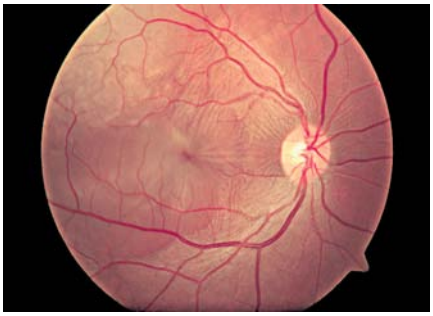


Fig. 6.11.3A: Active choroiditis—central

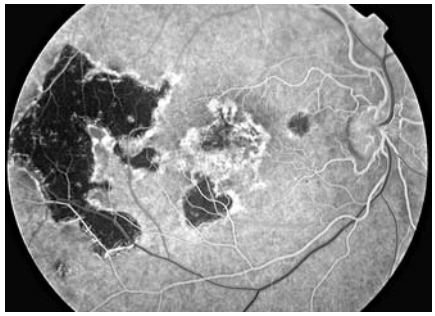


Fig. 6.11.3B: Active choroiditis—central

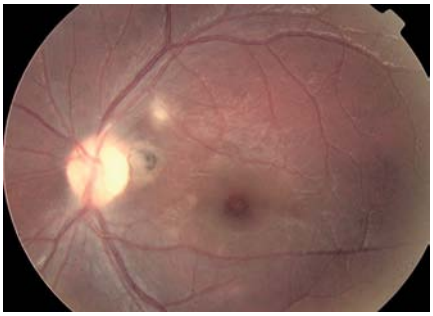


Fig. 6.11.4: Active choroiditis—juxtapapillary in toxoplasma

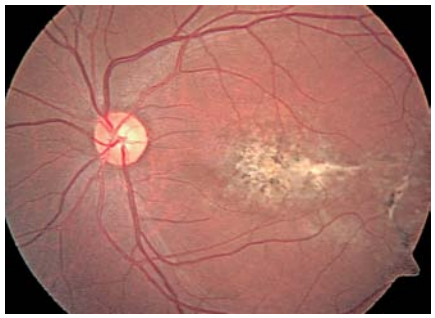


Fig. 6.11.5: Healed central choroiditis

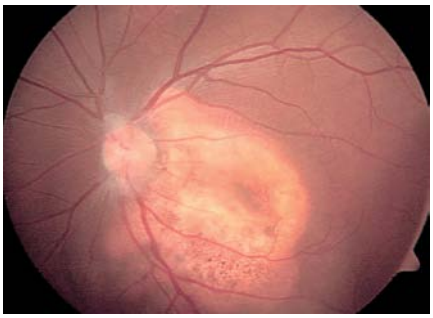


Fig. 6.11.6: Healed choroiditis—juxtapapillary

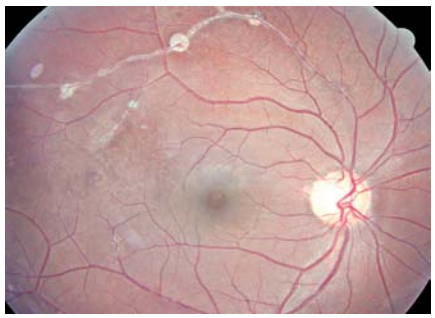


Fig. 6.11.7A: Perivasculitis—candle wax dripping in sarcoidosis

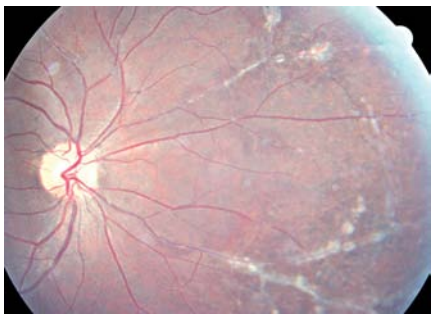


Fig. 6.11.7B: Perivasculitis—candle wax dripping in sarcoidosis

Toxoplasmosis

- Bilateral healed chorioretinal scars **(Figs 6.12.1A and B)**, may be unilateral **(Fig. 6.12.2)**
- *Focal necrotizing retinitis* is a 'satellite lesion' adjacent to the edge of the old scar **(Fig. 6.12.3)**
- *In reactivation*, white or yellowish-white lesions with fluffy indistinct edges visible at the post-equatorial fundus—"head light in fog" appearance **(Figs 6.12.4 and 6.12.5)**.

Toxocariasis

- Chronic endophthalmitis, vitreous clouding and severe cyclitic membrane formation **(*see* also Figs 7.9.3A and B)**
- ***See in Chapter 7***.

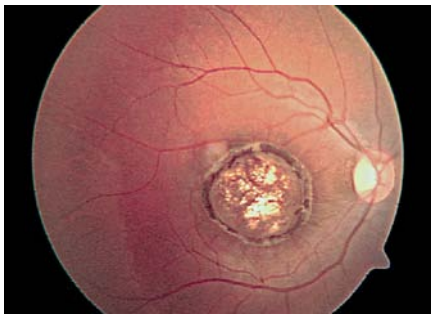


Fig. 6.12.1A: Toxoplasma choroiditis scar—bilateral—RE

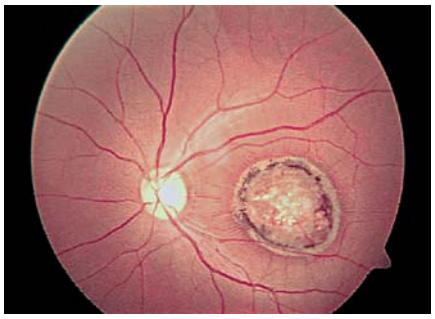


Fig. 6.12.1B: Toxoplasma choroiditis scar—bilateral—LE

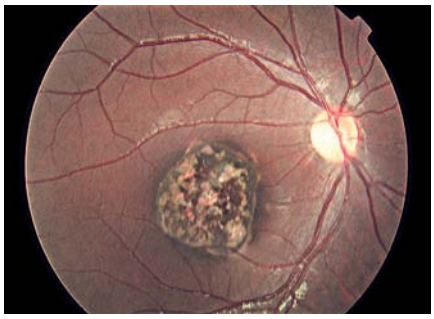


Fig. 6.12.2: Toxoplasma choroiditis scar—unilateral

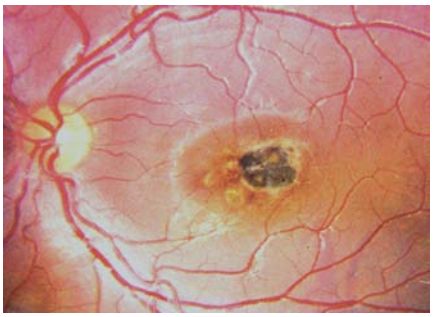


Fig. 6.12.3: Toxoplasma scar—satellite lesion

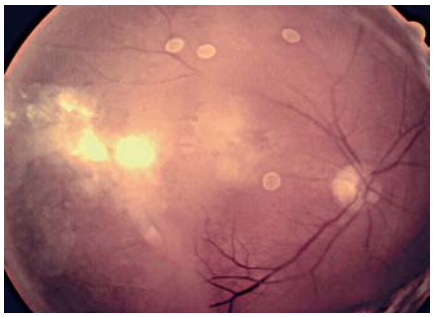


Fig. 6.12.4: Toxoplasma—reactivation—'head light in fog' appearance

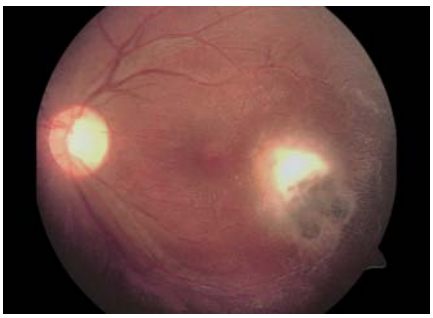


Fig. 6.12.5A: Toxoplasma—reactivation—'head light in fog' appearance

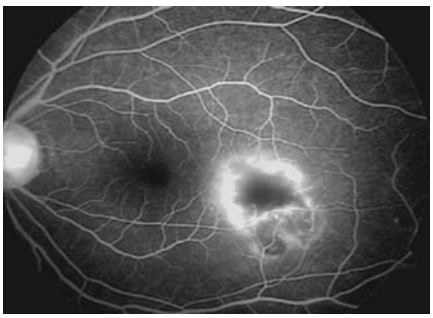


Fig. 6.12.5B: Toxoplasma—reactivation—FFA appearance

Vogt-Koyanagi-Harada (VKH) Syndrome

- Rare, idiopathic condition affects Asians, especially the Japanese
- Alopecia, vitiligo and poliosis are the hallmarks
- May be unilateral **(Fig. 6.13.1)** or bilateral **(Fig. 6.13.2)**
- Chronic granulomatous anterior uveitis **(Fig. 6.13.3)** with multifocal choroiditis **(Figs 6.13.4A to C)**
- Multiple sensory retinal detachments may be with exudation (Harada's) **(Fig. 6.13.5)**
- Prognosis is fairly good in unilateral cases, but poor in bilateral cases.

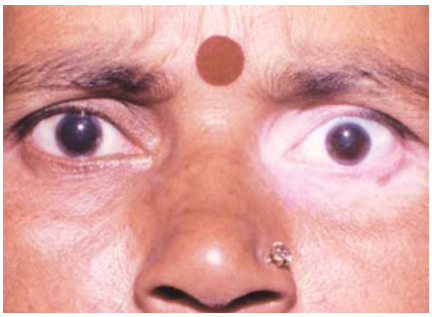


Fig. 6.13.1: Vogt-Koyanagi-Harada syndrome—unilateral—LE

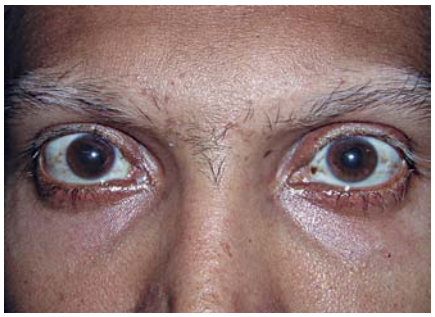


Fig. 6.13.2: Vogt-Koyanagi-Harada syndrome—bilateral

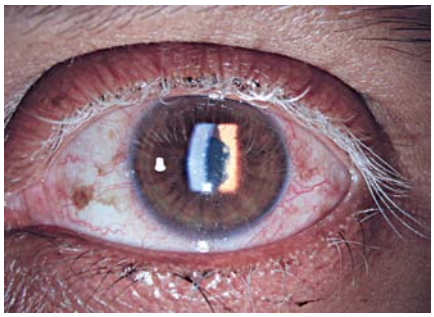


Fig. 6.13.3: VKH syndrome—chronic granulomatous anterior uveitis

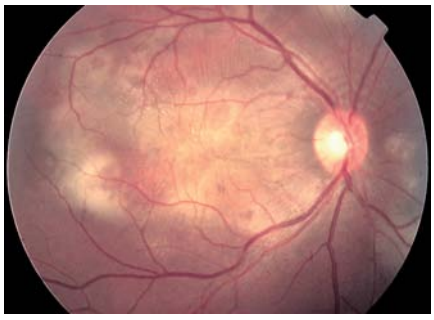


Fig. 6.13.4A: VKH syndrome—retinal findings

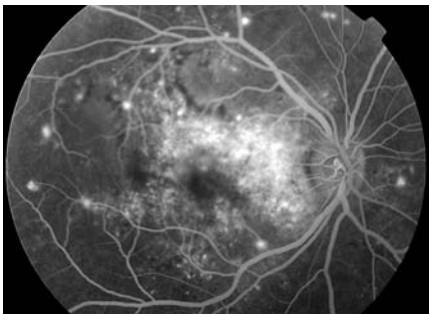


Fig. 6.13.4B: VKH syndrome—retinal findings

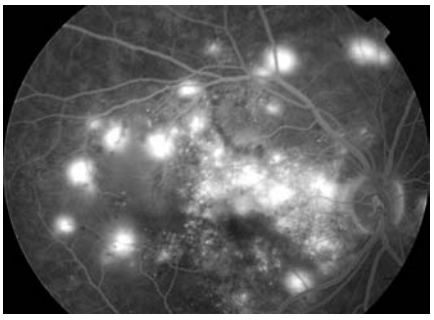


Fig. 6.13.4C: VKH syndrome—retinal findings

Sympathetic Ophthalmitis

- In the 'exciting eye'—there is ciliary congestion and evidence of initial insult
- In the 'sympathizing eye'—typical granulomatous uveitis with mutton-fat KPs, iris nodules
- Dalen-Fuchs' nodules scattered throughout posterior pole **(Fig. 6.14.1)**.

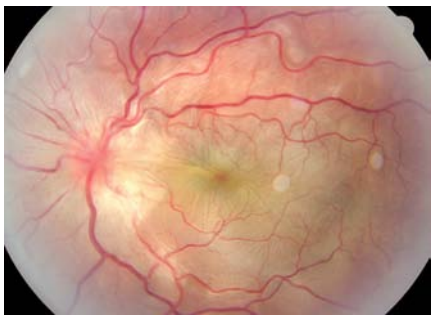


Fig. 6.13.5: VKH syndrome—exudative retinal detachment

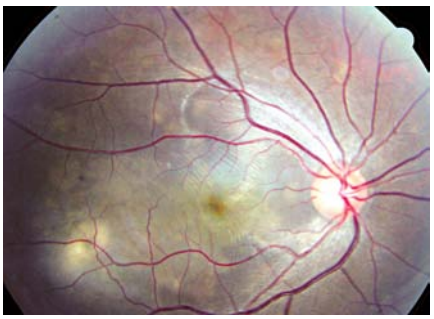


Fig. 6.14.1: Sympathetic ophthalmia—Dalen-Fuchs nodules

Birdshot Retinochoroiditis

- Uncommon, usually a bilateral condition
- Creamy-yellow, deep ovoid spots of moderate size with indistinct edges, radiate from the optic disk towards the equator **(Figs 6.15.1A and B)**
- Prognosis is often guarded.

Endophthalmitis

- *Exogenous*
  - Following open globe injury **(Figs 6.16.1A and B)**
  - Following intraocular surgery **(Figs 6.16.2A and B)** exudate may confine within the bag **(Figs 6.16.3A and B)**

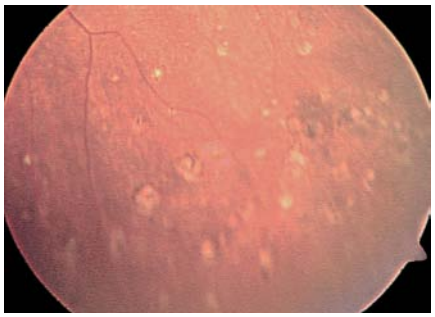


Fig. 6.15.1A: Birdshot retinochoroiditis

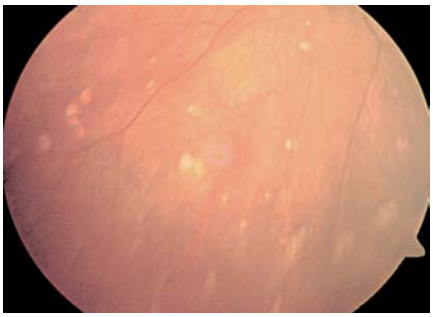


Fig. 6.15.1B: Birdshot retinochoroiditis

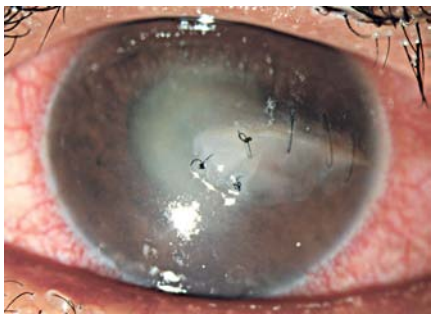


Fig. 6.16.1A: Endophthalmitis—penetrating injury

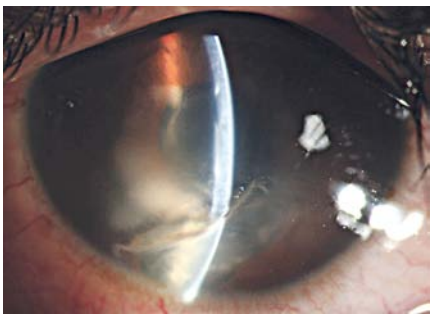


Fig. 6.16.1B: Endophthalmitis—penetrating injury

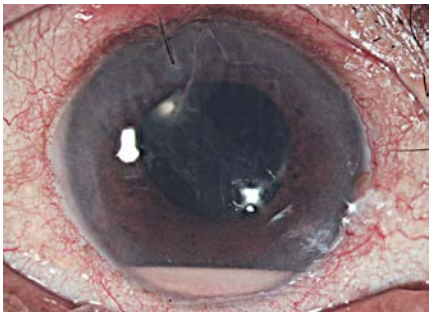


Fig. 6.16.2A: Endophthalmitis—cataract surgery

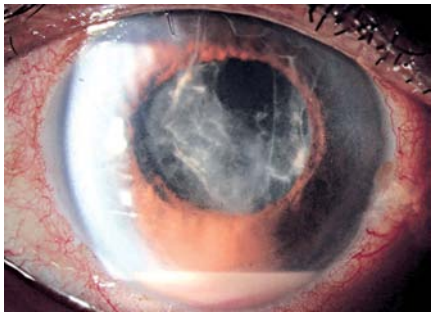


Fig. 6.16.2B: Endophthalmitis—cataract surgery

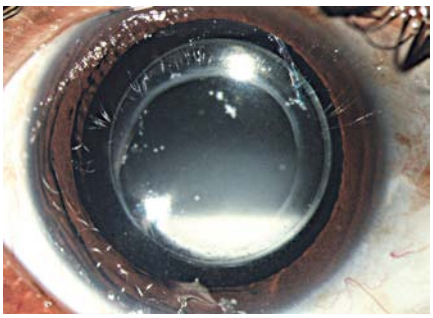


Fig. 6.16.3A: Endophthalmitis—cataract surgery—exudates in the bag

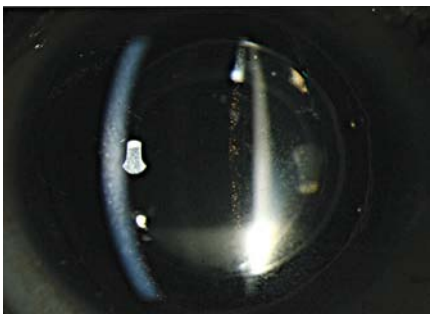


Fig. 6.16.3B: Endophthalmitis—cataract surgery—exudates in the bag

  - Following wound leak or bleb infection (after glaucoma surgery) **(Figs 6.16.4A and B)**
- *Endogenous*
  - Septic emboli (metastatic endophthalmitis) **(Figs 6.16.5A and B)**
  - *Toxic (sterile)*
  - Toxic reaction to the chemicals used during surgery—as in toxic anterior segment syndrome (TASS) **(Figs 6.16.6A and B)**.

Panophthalmitis

- Marked lid edema
- Purulent discharge
- Conjunctival chemosis and congestions
- Anterior chamber is full of pus
- Following corneal ulcer **(Figs 6.17.1A and B)**, open globe injury or post-surgical **(Fig. 6.17.2)**.

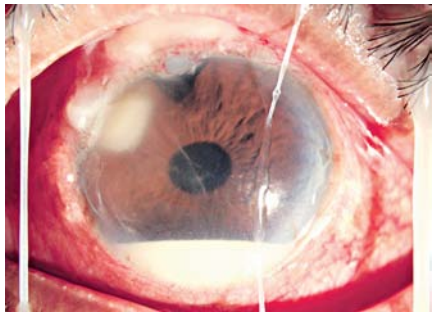


Fig. 6.16.4A: Blebitis—endophthalmitis after phacotrabeculectomy

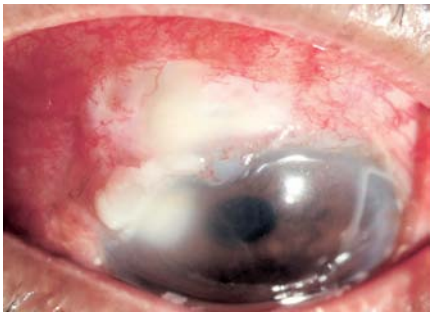


Fig. 6.16.4B: Blebitis—endophthalmitis

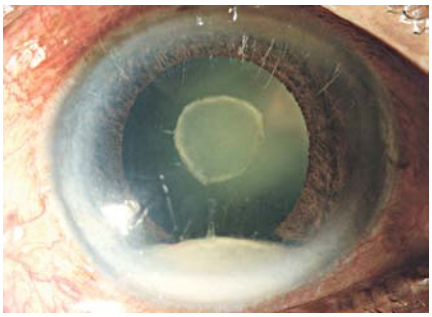


Fig. 6.16.5A: Metastatic endophthalmitis

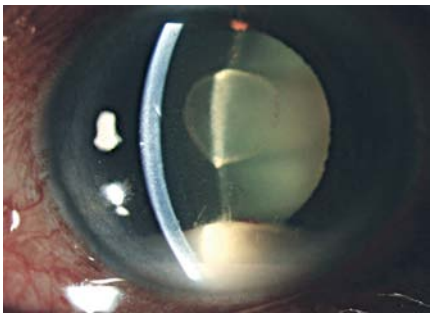


Fig. 6.16.5B: Metastatic endophthalmitis

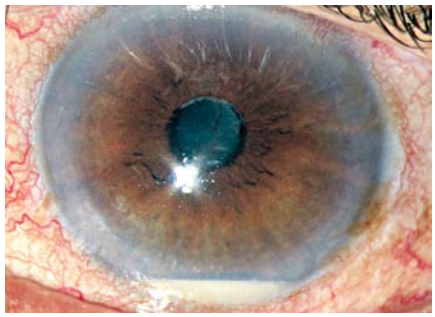


Fig. 6.16.6A: Toxic endophthalmitis—cataract surgery (TASS)

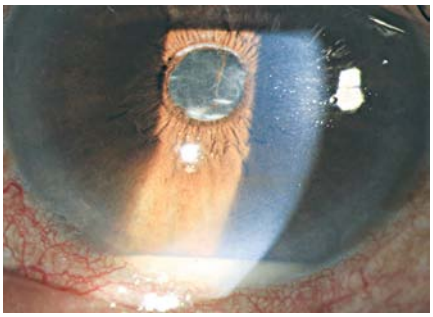


Fig. 6.16.6B: Toxic endophthalmitis—cataract surgery (TASS)

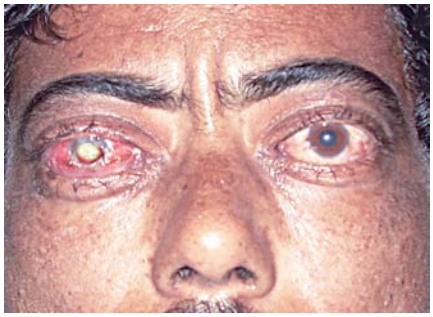


Fig. 6.17.1A: Panophthalmitis—corneal ulcer

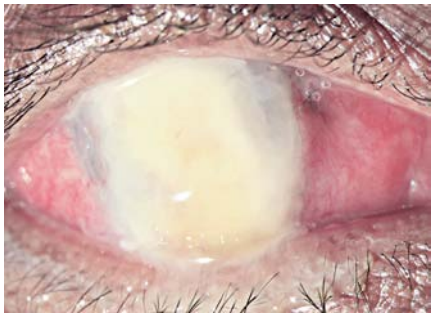


Fig. 6.17.1B: Panophthalmitis—corneal ulcer

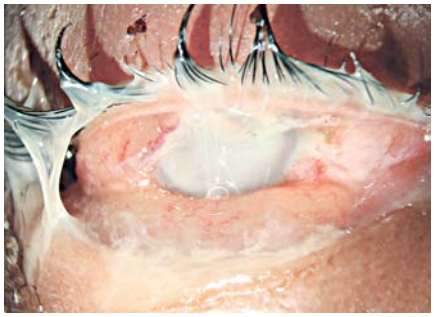


Fig. 6.17.2: Panophthalmitis following open globe injury

Iridocorneal Endothelial (ICE) Syndromes

- Rare, unilateral condition, typically affects women and associated with secondary intractable glaucoma
- After metaplasia, Descemet's membrane-like material covers the anterior iris surface and angle of the anterior chamber, causing iris anomalies, secondary glaucoma and corneal edema
- *It is of three types:*

Progressive Essential Iris Atrophy

- Patchy iris atrophy with partial or complete hole formation **(Fig. 6.18.1A)**
- Corectopia
- Pseudopolycoria, and gradual enlargement of the iris holes **(Fig. 6.18.1B)**
- Broad peripheral anterior synechiae and other eye absolutely normal **(Figs 6.18.2A and B)**.

Iris Nevus Syndrome (Cogan-Reese)

- Dark-brown pigmented nodules in the iris stroma as small woolen-spherules **(Fig. 6.19.1)**
- Peripheral anterior synechiae and secondary glaucoma may also occur **(Fig. 6.19.2)**
- Endothelial decompensation appears later.

Chandler's Syndrome

- Corneal endothelium appears 'beaten-silver' appearance
- With gradual endothelial disturbances, corneal edema develops **(Figs 6.20.1A and B)**
- Iris atrophy is minimal as compared to previous two

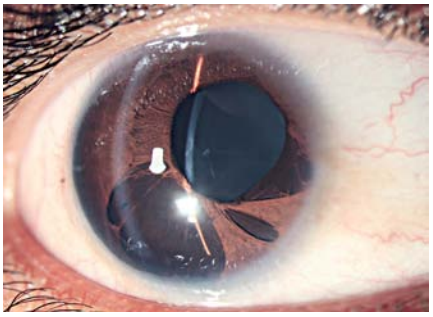


Fig. 6.18.1A: Essential iris atrophy

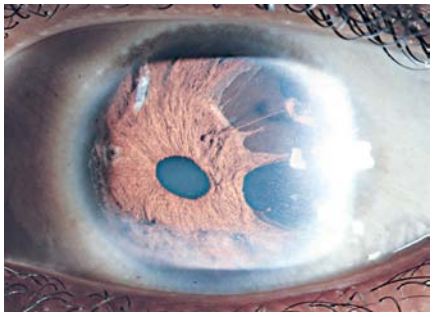


Fig. 6.18.1B: Essential iris atrophy—polycoria and PAS

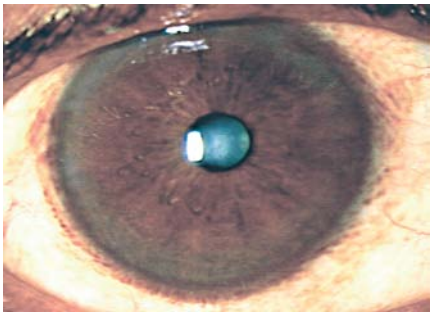


Fig. 6.18.2A: Essential iris atrophy—normal unaffected eye—RE

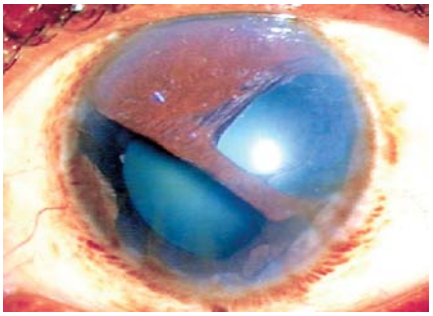


Fig. 6.18.2B: Essential iris atrophy—polycoria and PAS in affected eye—LE

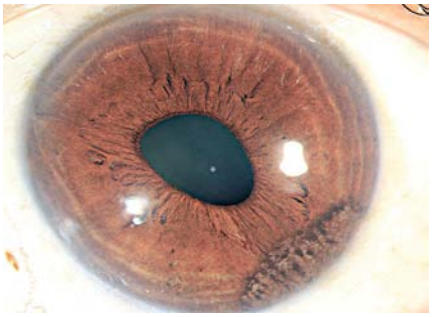


Fig. 6.19.1: Iris nevus syndrome (Cogan-Reese)

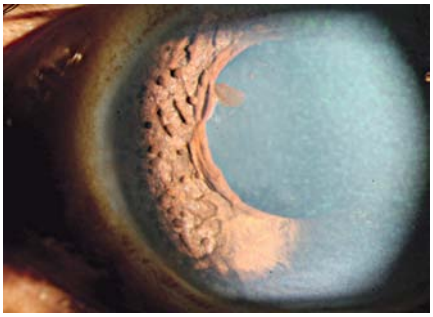


Fig. 6.19.2: Iris nevus syndrome (Cogan-Reese)

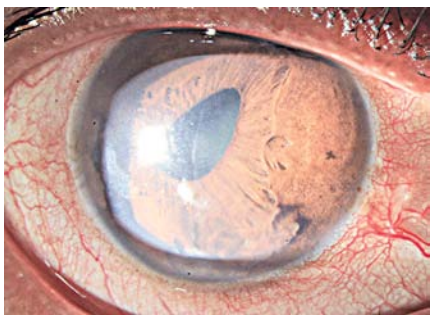


Fig. 6.20.1A: Chandler's syndrome with corneal edema

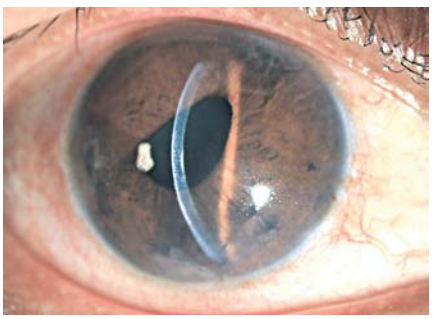


Fig. 6.20.1B: Chandler's syndrome with corneal edema

- Like previous two, the other eye is normal **(Figs 6.20.2A and B)**
- *Treatment* of ICE syndrome is necessary to treat secondary glaucoma. Medical treatment in early stages. Surgical treatment in late cases. But it often fails due to continuous nature of disease.
- In early cases of corneal edema, DSEK is the choice of surgery.

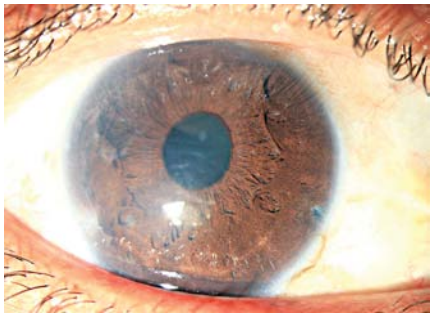


Fig. 6.20.2A: Chandler's syndrome—PAS and corneal edema—RE

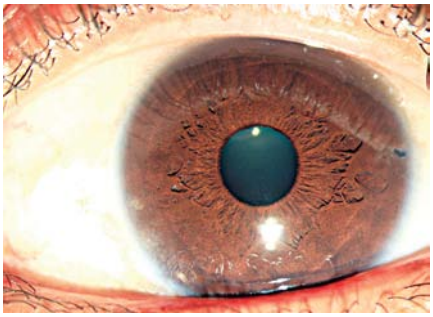


Fig. 6.20.2B: Chandler's syndrome—normal LE

Other Uveal Diseases

Iridoschisis

- Senile degenerative condition of the iris
- Large dehiscences appear on the anterior mesodermal layer of the iris **(Fig. 6.21.1)** and strands of tissue may float into the anterior chamber **(Fig. 6.21.2)**
- *Treatment* is not necessary.

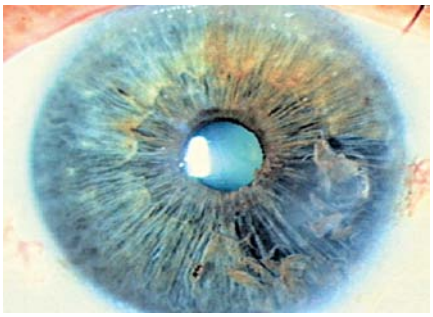


Fig. 6.21.1: Iridoschisis

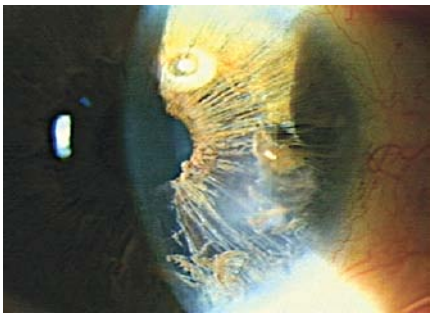


Fig. 6.21.2: Iridoschisis—floating strands in the anterior chamber

Iris Atrophy

- May be diffuse or sectorial
- *Diffuse type* occurs in old age **(Figs 6.22.1A and B)**, following iridocyclitis **(Figs 6.22.2 and 6.22.3)** post cataract surgery or other anterior segment surgeries **(Fig. 6.22.4)**
- *Sectorial type* found in—herpetic iritis **(Fig. 6.22.5)**, angle closure glaucoma **(Fig. 6.22.6)**, after surgery **(Fig. 6.22.7)** or trauma **(Figs 6.22.8A and B)**.

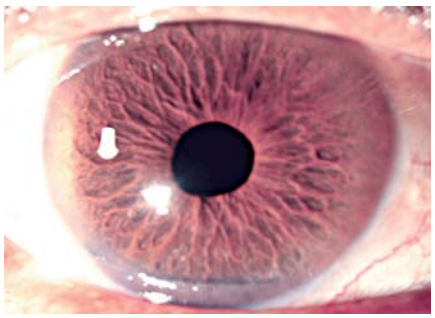


Fig. 6.22.1A: Iris atrophy in old age

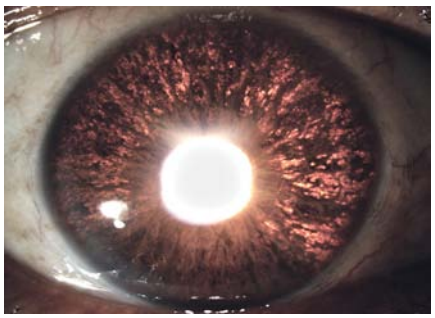


Fig. 6.22.1B: Same eye in **Figure 6.22.1A**—iris transillumination positive

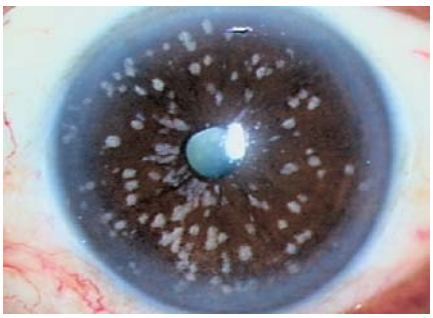


Fig. 6.22.2: Iris atrophy—diffuse moth-eaten iris

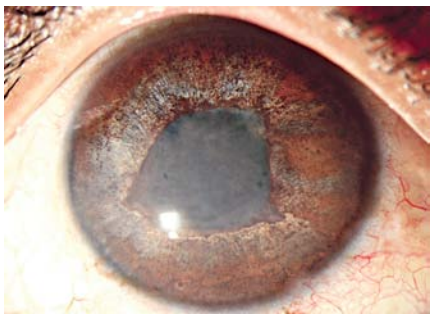


Fig. 6.22.3: Iris atrophy—diffuse

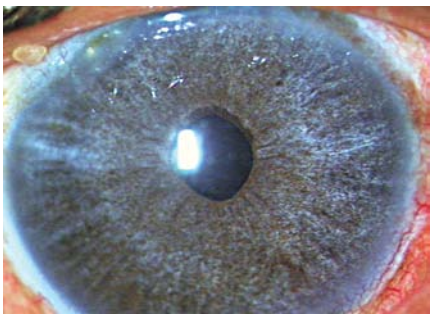


Fig. 6.22.4: Iris atrophy following cataract surgery—diffuse

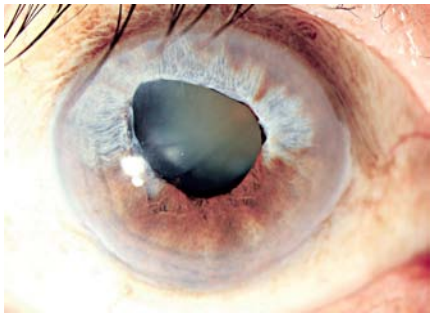


Fig. 6.22.5: Iris atrophy—sectorial—post-herpetic

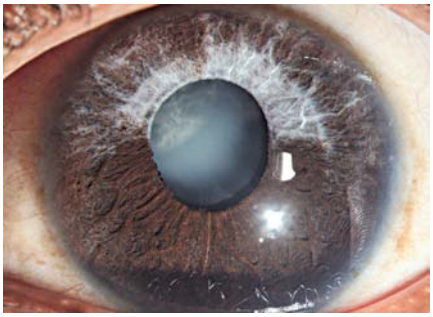


Fig. 6.22.6: Iris atrophy—sectorial—post-herpetic

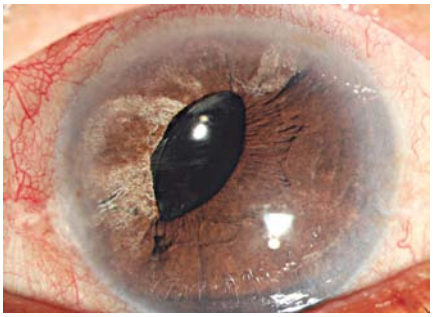


Fig. 6.22.7: Iris atrophy—post-surgical

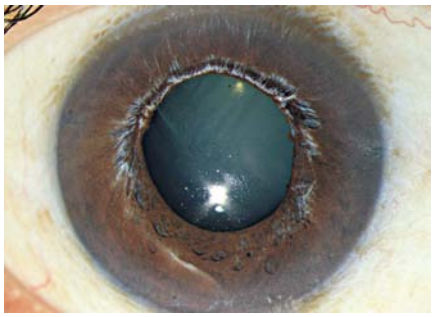


Fig. 6.22.8A: Iris sphincteric atrophy—post-traumatic

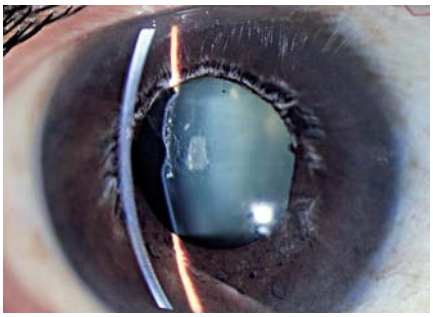


Fig. 6.22.8B: Iris sphincteric atrophy—post-traumatic with subluxation

Rubeosis Iridis

- Start as tiny dilated capillaries at the pupillary border
- Radial iris neovascularization **(Fig. 6.23.1)**
- Followed by irregularly distributed network of new vessels on the iris surface and the stroma **(Fig. 6.23.2)**
- New blood vessels and associated fibrous tissue may cover angle and the trabecular meshwork
- Cause peripheral anterior synechiae and intractable neovascular glaucoma **(Fig. 6.23.3)** ***(see also in Chapter 9)***

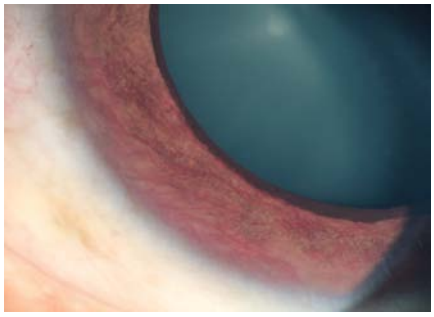


Fig. 6.23.1: Rubeosis iridis

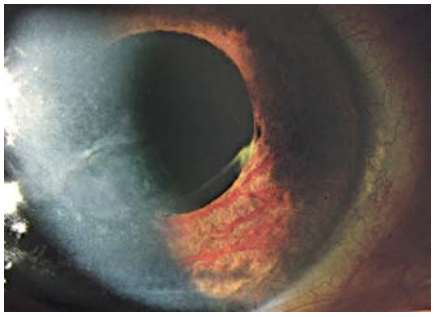


Fig. 6.23.2: Rubeosis iridis

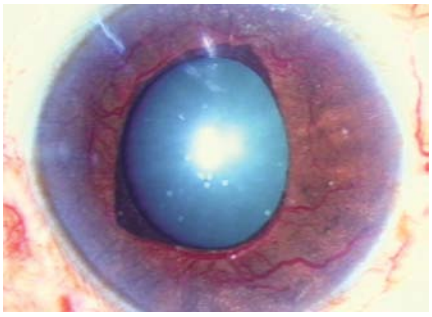


Fig. 6.23.3: Rubeosis iridis—PAS formation

- Ectropion uveae is common in late stage **(Fig. 6.23.4)** and new vessels may spread over the anterior surface of the lens **(Fig. 6.23.5)**
- *Treatment:* Panretinal photocoagulation; panretinal cryocoagulation in presence of opaque media.

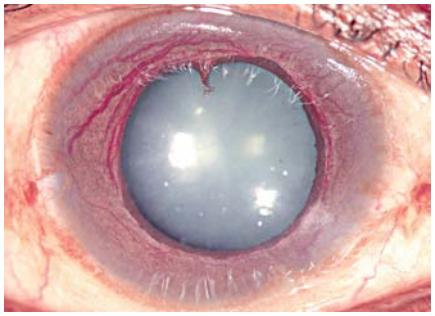


Fig. 6.23.4: Rubeosis iridis—ectropion uveae

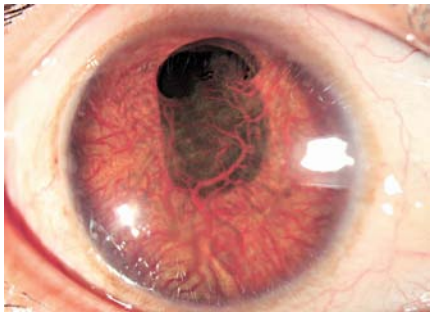


Fig. 6.23.5: Rubeosis iridis—new vessels on lens surface

Primary Choroidal Sclerosis

- Diffuse atrophy of the RPE and choriocapillaris
- 'Tessellated' appearance of the fundus
- It occurs in *two forms:*
  - Diffuse or generalized sclerosis **(Fig. 6.24.1)**
  - *Localized sclerosis:* Affects the central or circumpapillary region **(Figs 6.24.2 and 6.24.3)**.

Iris Cysts

- Iris cysts are not so uncommon, and they may be primary or secondary
- ***Primary iris cysts:*** Are stromal and occur in young children. It is filled with clear fluid and may often touch the cornea with endothelial decompensation **(Figs 6.25.1 and 6.25.2)**

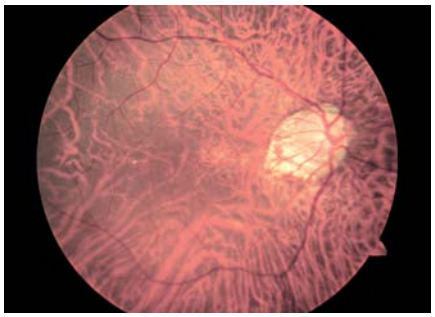


Fig. 6.24.1: Choroidal sclerosis—diffuse

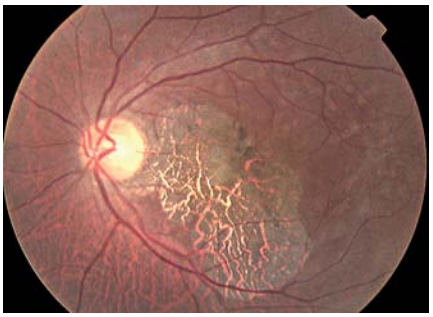


Fig. 6.24.2: Choroidal sclerosis—localized

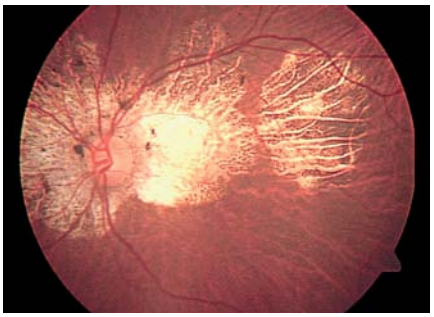


Fig. 6.24.3: Choroidal sclerosis—localized and peripapillary

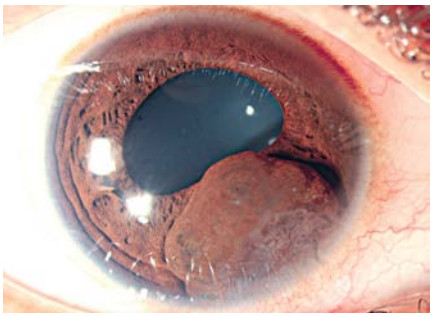


Fig. 6.25.1A: Iris cyst—primary

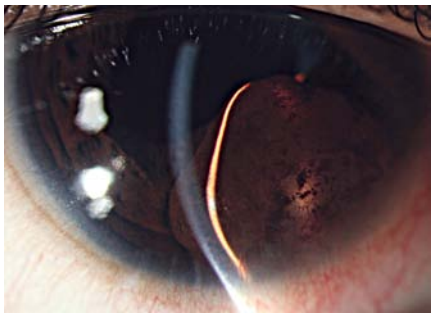


Fig. 6.25.1B: Iris cyst—primary

- ***Secondary iris cysts:*** May be post-surgical or post-traumatic following blunt or open globe injury:
  - *Post-surgical iris cysts:* A form of epithelial down growth and occur following cataract surgery **(Fig. 6.25.3A)**. Cysts lie on the anterior surface of the iris, and grayish-white in color and filled with clear fluid with anterior clear wall **(Fig. 6.25.3B)**
  - *Post-traumatic iris cysts:* Grayish-white or white in color and filled with turbid fluid with anterior clear wall **(Figs 6.25.4 and 6.25.5)**
- *Treatment*: Surgical excision or sometimes YAG laser deroofing of the cyst. Corneal endothelial problem can be managed seperately later on.

Gyrate Atrophy of the Choroid

- Deficiency of ornithine ketoacid aminotransferase enzyme, resulting in hyperornithinemia
- Scalloped to circular patches of chorioretinal atrophy in the far and midretinal periphery **(Figs 6.26.1 and 6.26.2)**.

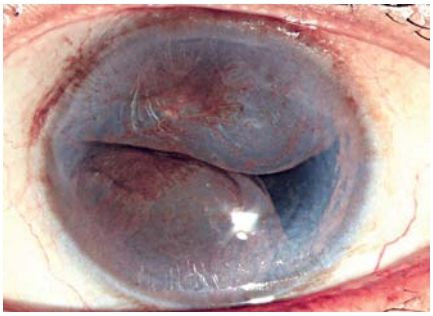


Fig. 6.25.2A: Iris cyst—primary

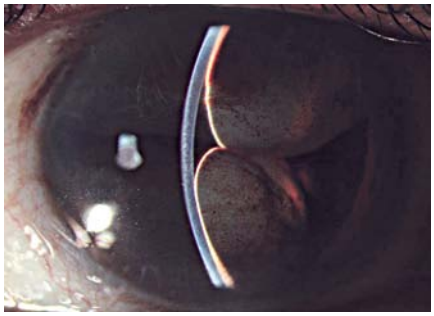


Fig. 6.25.2B: Iris cyst—primary

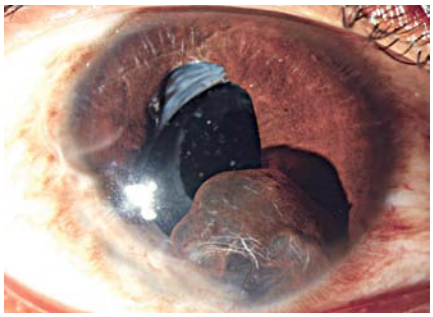


Fig. 6.25.3A: Iris cyst—post cataract surgery

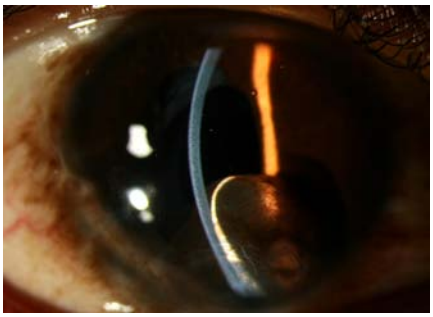


Fig. 6.25.3B: Iris cyst—post-surgical with clear fluid

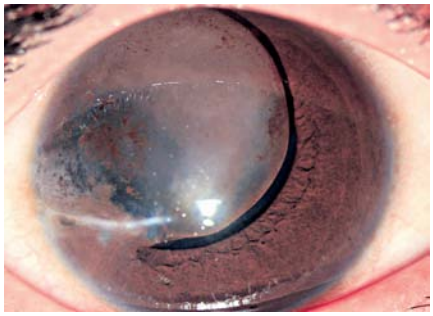


Fig. 6.25.4A: Iris cyst with turbid fluid—post-traumatic

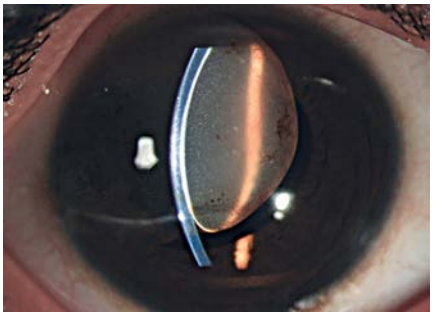


Fig. 6.25.4B: Iris cyst with turbid fluid—post-traumatic

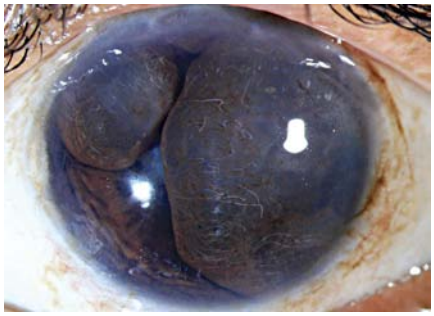


Fig. 6.25.5: Iris cyst with cataract—post-traumatic

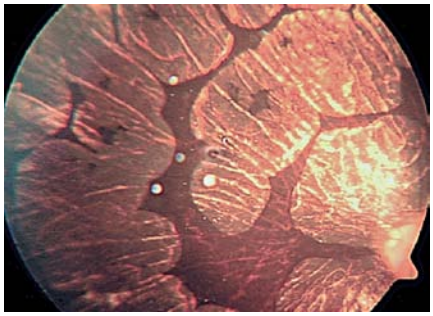


Fig. 6.26.1: Gyrate atrophy of the choroid

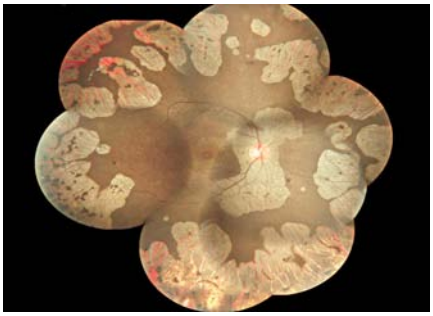


Fig. 6.26.2: Gyrate atrophy of the choroid—montage image

Choroideremia

- Rare, X-linked recessive inheritance
- Diffuse mottled depigmentation of the RPE **(Figs 6.27.1A to C)**
- In late stage, extensive RPE and choroidal atrophy with sparing of the fovea.

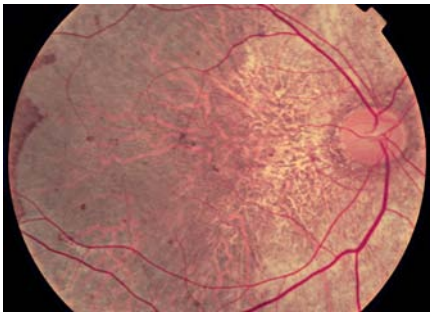


Fig. 6.27.1A: Choroideremia

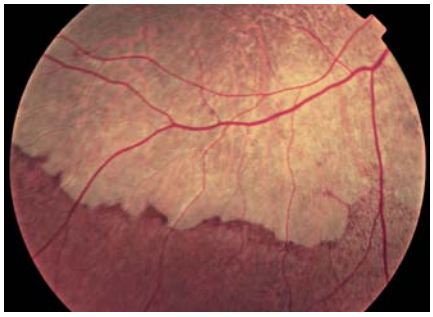


Fig. 6.27.1B: Choroideremia

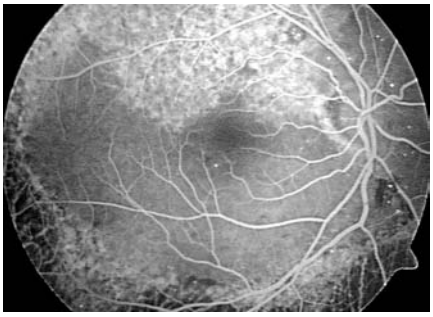


Fig. 6.27.1C: Choroideremia on FFA

Angioid Streaks

- Angioid streaks are irregular and jagged network of red to brown lines, mainly seen in the central fundus **(Fig. 6.28.1)**
- Lesions are approximately the width of a retinal vessel, which may thus resemble ('angioid')
- But, the streaks are darker, have an irregular contour with serrated edges, and tend to terminate abruptly **(Fig. 6.28.2)**.

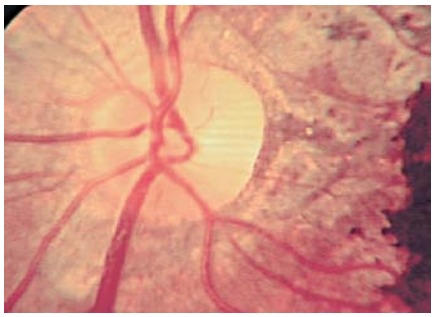


Fig. 6.28.1: Angioid streak

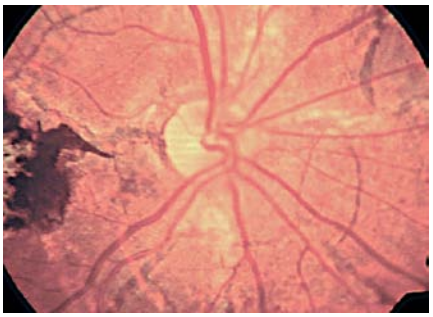


Fig. 6.28.2: Angioid streak

Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE)

- Typical deep, cream-colored placoid lesions, involving the posterior pole within the equatorial region **(Figs 6.29.1A to C)**.

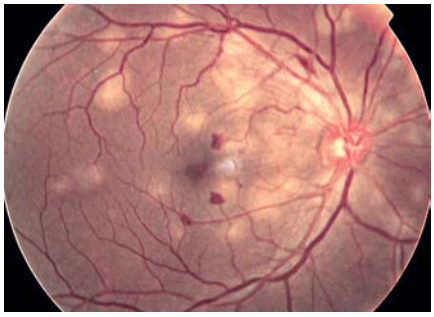


Fig. 6.29.1A: APMPPE

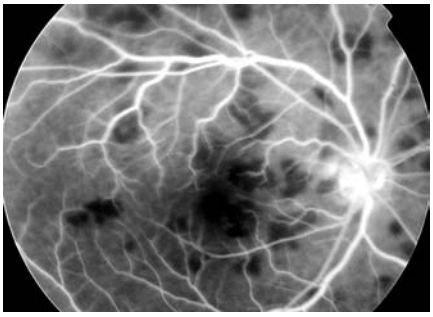


Fig. 6.29.1B: APMPPE on FFA

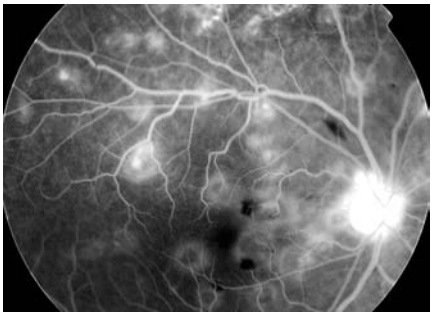


Fig. 6.29.1C: APMPPE on FFA

Serpiginous (Geographical) Choroidopathy

- Lesion usually starts around the optic disk, and spreads outwards in all directions
- Cream-colored opacities with hazy border without any inflammatory cells in the vitreous **(Figs 6.30.1A and B)**
- Fresh acute lesions usually arise as extensions from old scars, and successive attacks result in serpiginous extension of the destruction process peripherally from the peripapillary area **(Fig. 6.30.2)**.

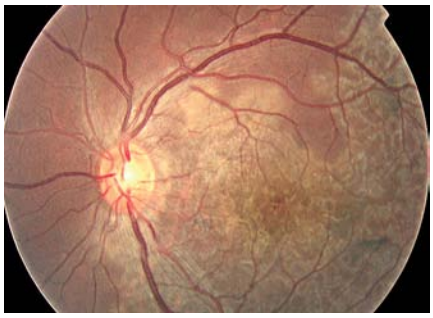


Fig. 6.30.1A: Serpiginous geographical choroidopathy

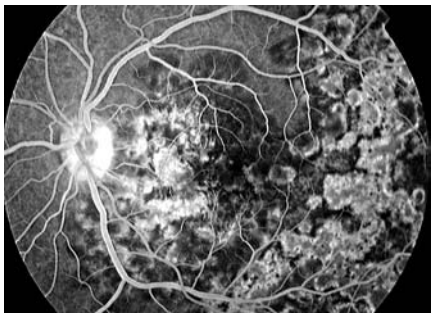


Fig. 6.30.1B: Serpiginous geographical choroidopathy

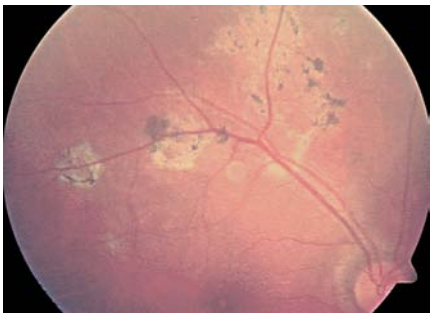


Fig. 6.30.2: Serpiginous geographical choroidopathy at periphery

Choroidal Detachment

- Means separation of the choroids from the sclera
- Shallow anterior chamber
- Shows smooth, elevated, dark-brown bullous lesions, which are more prominent on the temporal and nasal sides **(Figs 6.31.1 and 6.31.2)**
- Sometimes, the lesions appear on both temporal and nasal sides, called kissing choroidals **(Fig. 6.31.3)**
- May be associated with retinal detachment **(Fig. 6.31.4)**.

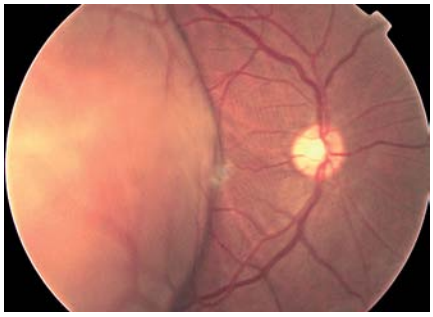


Fig. 6.31.1: Choroidal detachment—temporal

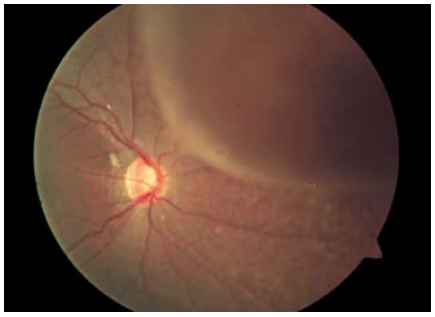


Fig. 6.31.2: Choroidal detachment—superonasal

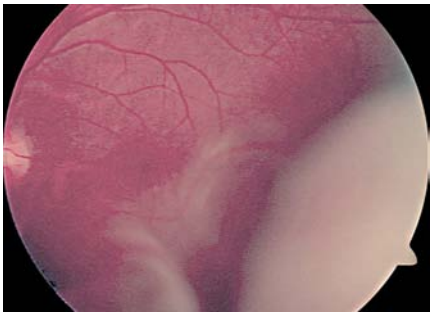


Fig. 6.31.3: Choroidal detachments—kissing choroidals

Benign Uveal Lesions/ Nodules/Mass

Iris Floccules

- Bilateral, uncommon condition
- Pupillary tags or tiny solid pigmentary nodules around the pupil **(Fig. 6.32.1)**.

Iris Freckles

- Bilateral tiny nevi on the iris surface
- Mainly seen in adults, rarely in children
- Multiple, discrete colorful spots on the anterior iris surface **(Figs 6.33.1A and B)**.

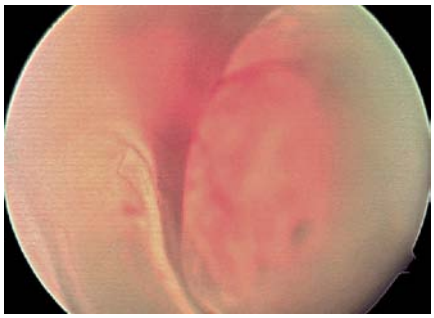


Fig. 6.31.4: Choroidal detachment with retinal detachment

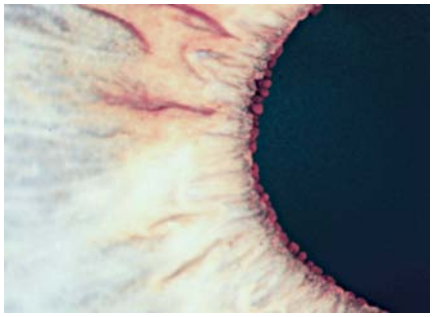


Fig. 6.32.1: Iris floccules

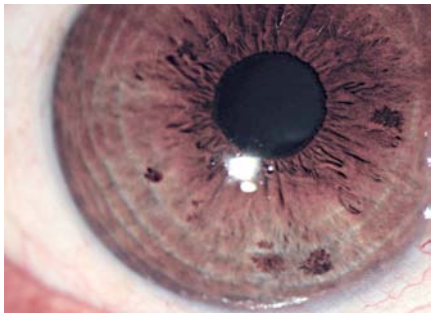


Fig. 6.33.1A: Iris freckles

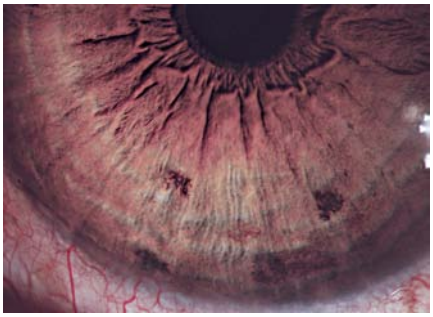


Fig. 6.33.1B: Iris freckles

Lisch Nodules/Spots

- Bilateral melanocytic hamartomas found in patients with neurofibromatosis after puberty **(Figs 6.34.1A and B)**
- Small brown or yellowish-brown dome-shaped nodules **(Figs 6.34.2A and B)** or spots **(Fig. 6.34.3)** on the anterior surface, called Lisch nodules/spots.

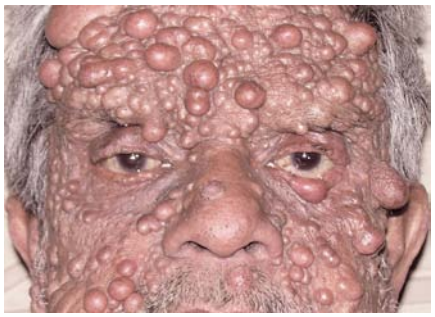


Fig. 6.34.1A: Neurofibromatosis

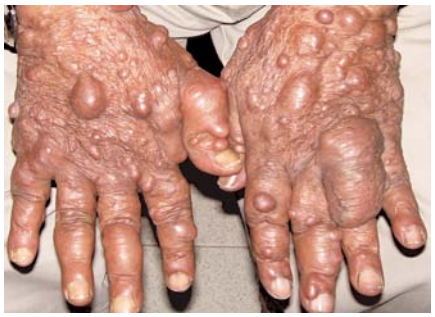


Fig. 6.34.1B: Neurofibromatosis

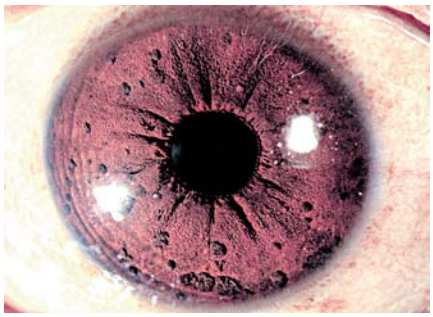


Fig. 6.34.2A: Lisch nodules—RE—neurofibromatosis—same patient **(Fig. 6.34.1A)**

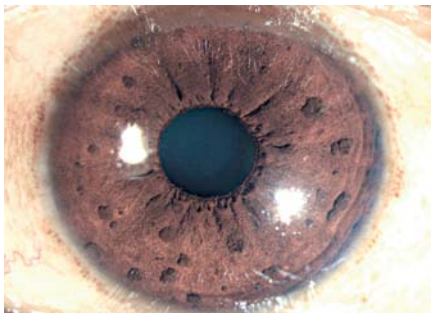


Fig. 6.34.2B: Lisch nodules—LE—neurofibromatosis—same patient **(Fig. 6.34.1)**

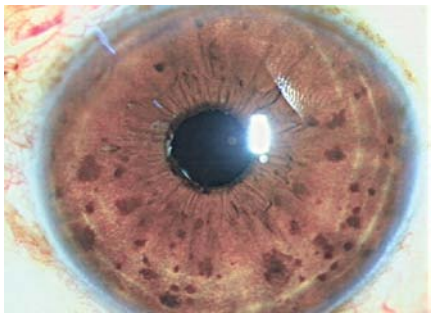


Fig. 6.34.3: Lisch spots

Brushfield Spots

- Bilateral, and are usually found in Down syndrome, may be seen in general population
- Tiny yellowish or white spots arranged in a ring at the junction of middle and outer third of iris surface **(Figs 6.35.1A and B)**.

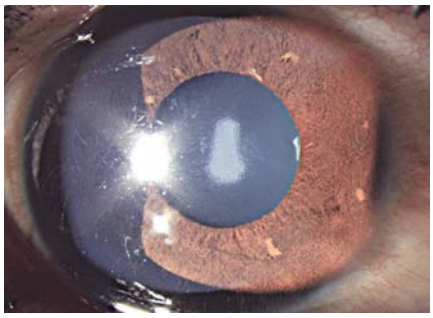


Fig. 6.35.1A: Brushfield spot in Down syndrome—RE

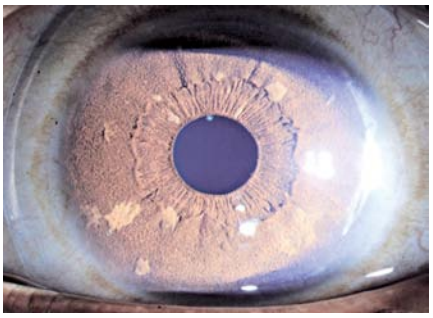


Fig. 6.35.1B: Brushfield spot in Down syndrome—LE

Iris Pearls

- Bilateral small or large, pearl-like clear nodules on the iris surface and pupillary margin **(Fig. 6.36.1)**
- May be seen in lepromatous leprosy
- May drop into AC and eventually disappear.

Iris Mammillations

- Unilateral or bilateral rare condition
- Tiny, smooth, villiform lesions which are regularly spaced **(Fig. 6.37.1)**
- May found in normal people or with some syndromes.

Iris Nevus

- Common, usually unilateral
- Elevated or flat, localized, discrete, small pigmented mass **(Fig. 6.38.1)**
- Usually does not distort the pupil
- May cause heterochromia iridis **(Fig. 6.38.2)**.

Choroidal Hemangioma

- In 50% cases associated with skin angioma, as in Sturge-Weber syndrome **(Fig. 6.39.1A)**
- Appears as a dome-shaped or diffuse, reddish-orange lesion mostly at the posterior pole **(Fig. 6.39.1B)**
- In 50% of the cases, it appears as an isolated entity **(Figs 6.39.2A and B)**.

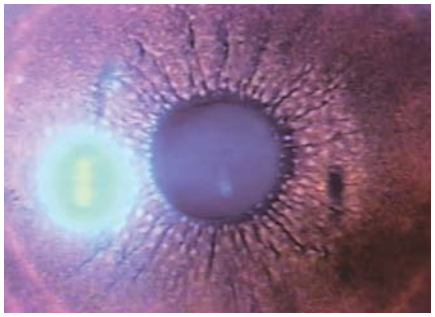


Fig. 6.36.1: Iris pearls in leprosy

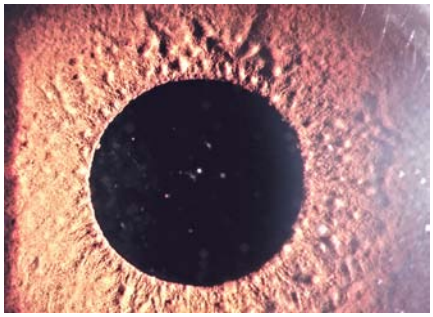


Fig. 6.37.1: Iris mammillations

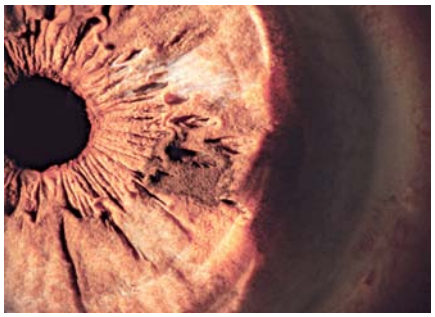


Fig. 6.38.1: Iris nevus

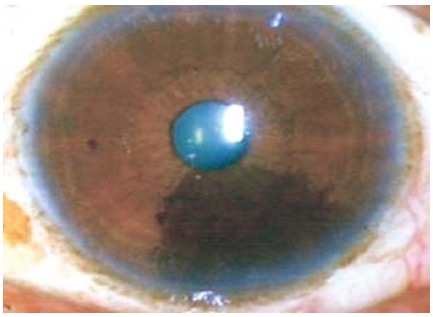


Fig. 6.38.2: Iris nevus

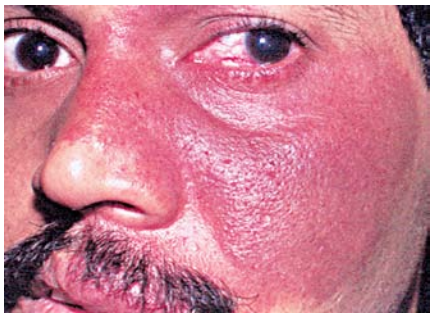


Fig. 6.39.1A: Port-wine stain—Sturge-Weber syndrome

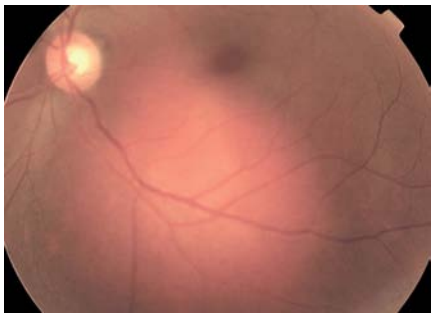


Fig. 6.39.1B: Sturge-Weber syndrome—choroidal hemangioma

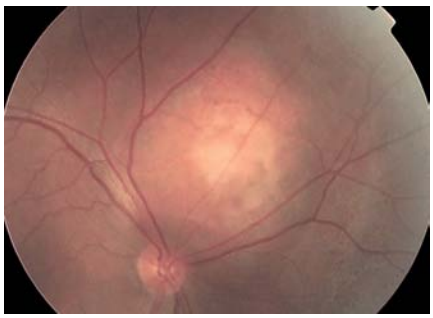


Fig. 6.39.2A: Choroidal hemangioma—isolated

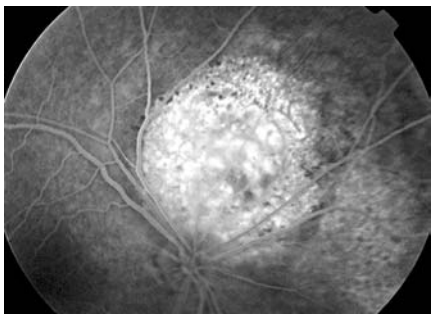


Fig. 6.39.2B: Choroidal hemangioma on FFA

Benign Melanocytoma (Nevus)

- Typical benign melanomas of the choroid are flat or slightly elevated, oval or circular state-gray lesions **(Fig. 6.40.1)**
- Occur most frequently at the posterior half of the fundus **(Fig. 6.40.2)**
- May be also central lesion, in which case, patient complaints of central scotoma **(Fig. 6.40.3)**
- *Treatment:* Not indicated, except the patient should be followed up regularly with serial photographs.

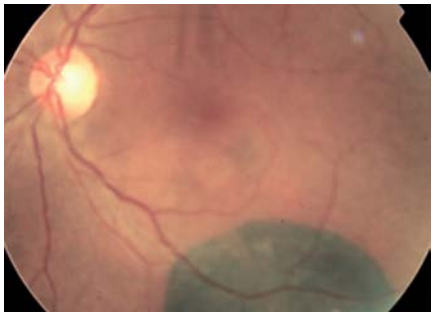


Fig. 6.40.1: Benign melanocytoma of the choroid—peripheral

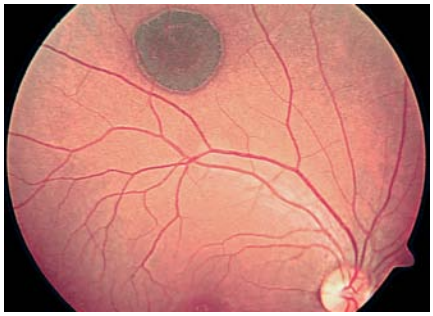


Fig. 6.40.2: Benign melanocytoma (nevus) of the choroid

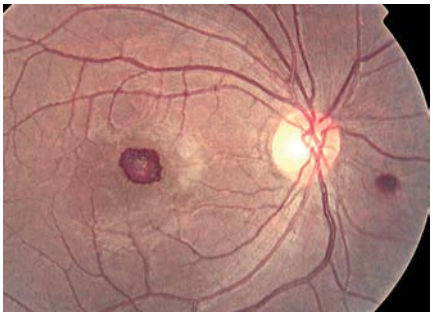


Fig. 6.40.3: Benign melanocytoma of the choroid—central lesion

Choroidal Osteoma

- Very rare, juxtapapillary lesion, that typically affects the young females
- Yellowish-orange lesion with scalloped borders adjacent to the optic disk **(Figs 6.41.1 and 6.41.2)**
- *No treatment is required.*

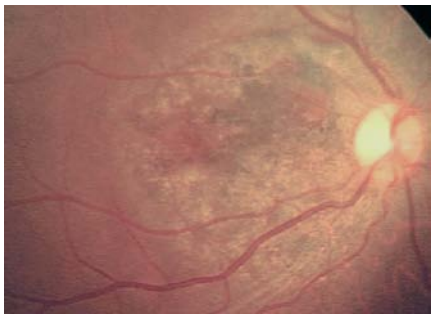


Fig. 6.41.1: Choroidal osteoma

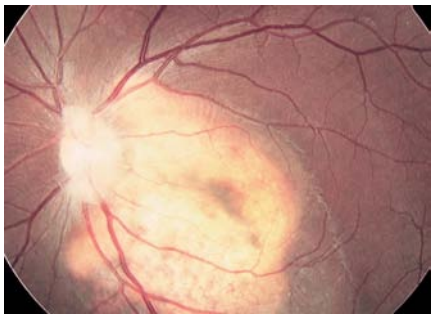


Fig. 6.41.2: Choroidal osteoma

Malignant Uveal Tract Lesions

Metastatic Deposits of Retinoblastoma

- Rare presentation of retinoblastoma
- Pale-white multiple iris nodules, often associated with pseudo-hypopyon **(Fig. 6.42.1)**.

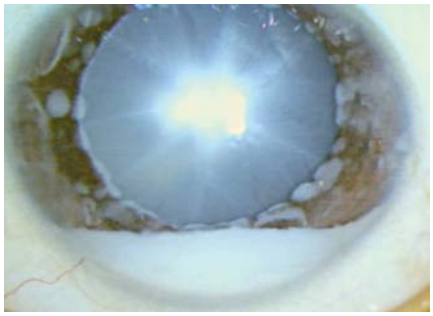


Fig. 6.42.1: Retinoblastoma—metastatic deposits

Malignant Melanoma of the Iris

- Very rare, slow-growing tumor with relative low malignant potential
- Noticed as a brown or nonpigmented mass on the iris surface, usually located in the inferior half of the iris **(Fig. 6.43.1)**

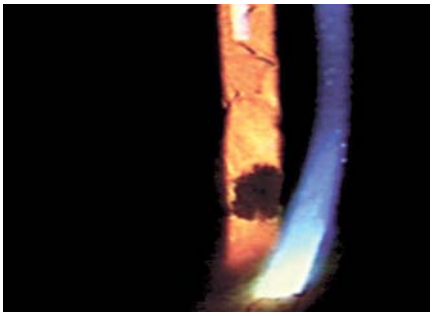


Fig. 6.43.1: Malignant melanoma of the iris—a small lesion

- May start at the angle of AC **(Fig. 6.43.2)** and then gradually spread towards center **(Fig. 6.43.3)**
- Pupil is distorted; ectropion uveae and secondary lens opacities are seen
- May extend into the anterior chamber angle giving rise to secondary glaucoma **(Fig. 6.43.4)**
- *Forward extension of the ciliary body melanoma*
  - May not be confined to inferior half **(Fig. 6.43.5)**
  - Examination after dilatation confirms the diagnosis **(Fig. 6.43.6)**.

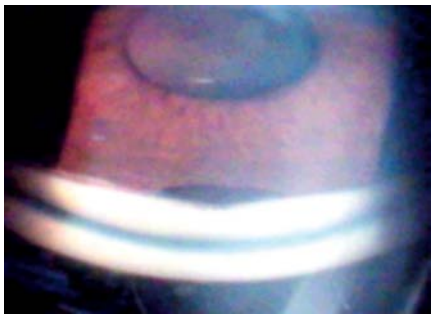


Fig. 6.43.2: Iris melanoma at the angle—visible gonioscopically

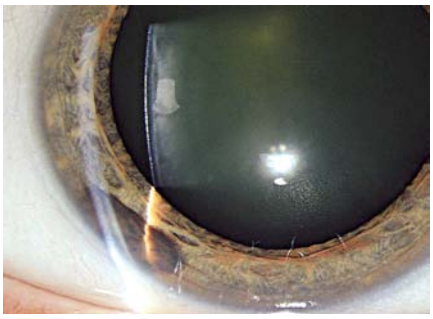


Fig. 6.43.3: Iris melanoma—near angle

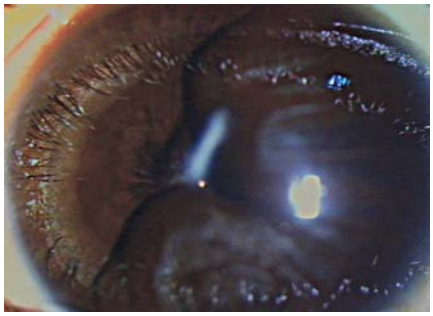


Fig. 6.43.4: Malignant melanoma of the iris—large lesion

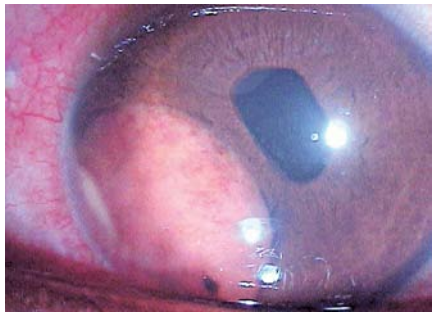


Fig. 6.43.5: Ciliary body melanoma—forward extension

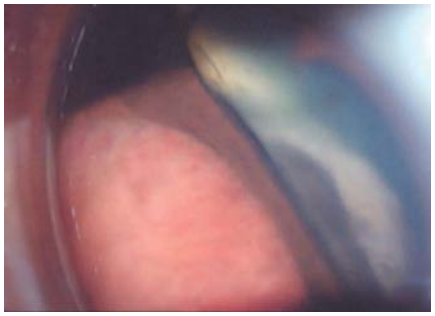


Fig. 6.43.6: Ciliary body melanoma—forward extension

Medulloepithelioma (Diktyoma)

- Extremely rare, arises from the non-pigmented epithelium of the ciliary body
- Presents with a white pupil (leukocoria), secondary cataract and secondary glaucoma.

Malignant Melanoma of the Ciliary Body

- Ciliary body melanoma is more common than iris melanoma
- *May present:*
  - As subluxation of the lens with secondary glaucoma
  - As dilated episcleral blood vessels ('sentinel vessels')
  - As a diffuse mass around the ciliary body **(Figs 6.44.1A and B)**

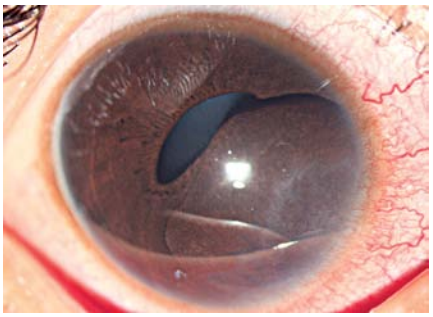


Fig. 6.44.1A: Ciliary body melanoma

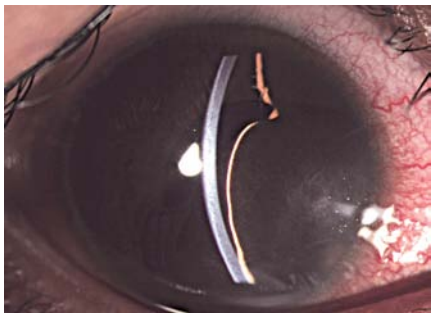


Fig. 6.44.1B: Ciliary body melanoma

- *Treatment:* By enucleation (for large tumors) and local resection (for small tumors).

Malignant Melanoma of the Choroid

- Chief symptoms result from the exudative retinal detachment with secondary glaucoma
- Typically appears as a pigmented and elevated oval mass **(Fig. 6.45.1)**
- As the tumor grows, a brown exudative detachment results **(Figs 6.45.2 and 6.45.3)**
- *Treatment*: Small melanoma can be treated by laser **(Fig. 6.45.4)**, for large melanoma—enucleation.

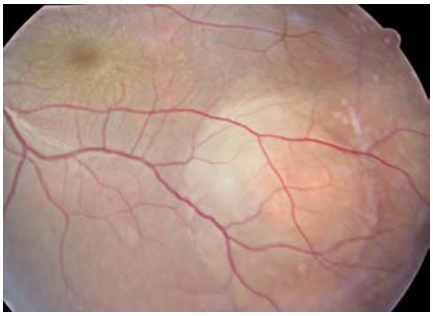


Fig. 6.45.1: Malignant melanoma of the choroid

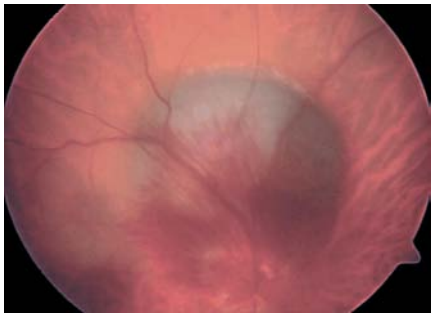


Fig. 6.45.2A: Choroidal melanoma

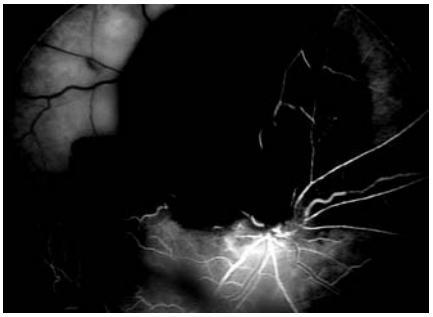


Fig. 6.45.2B: Choroidal melanoma—FFA

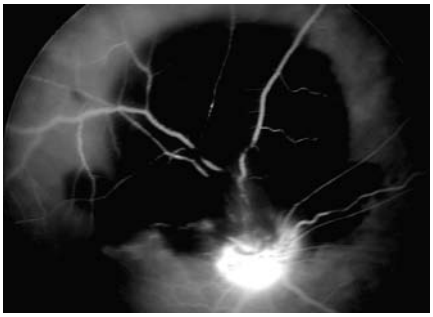


Fig. 6.45.2C: Choroidal melanoma—FFA

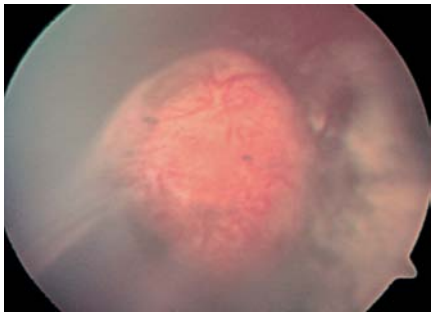


Fig. 6.45.3: Choroidal melanoma with exudative RD

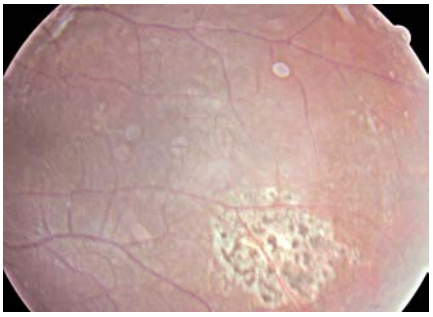


Fig. 6.45.4: Choroidal melanoma—post laser

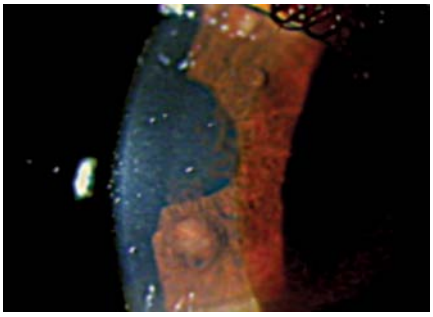


Fig. 6.46.1: Metastatic carcinoma of the iris—from bronchogenic carcinoma

Metastatic Carcinoma of the Uveal Tract

- *Most frequent primary sites:* Bronchus in males, and breast in females
- May deposit on iris **(Figs 6.46.1 and 6.46.2)**, ciliary body or choroid
- Typically, appear as solitary **(Fig. 6.46.3)** or multiple **(Fig. 6.46.4)**, creamy-white, placoid or oval lesions which infiltrate laterally
- Careful examination of the other eye is important as bilateral metastases are common
- *Treatment:* Must be directed to the primary disease. Enucleation is contraindicated unless the eye is painful and blind.

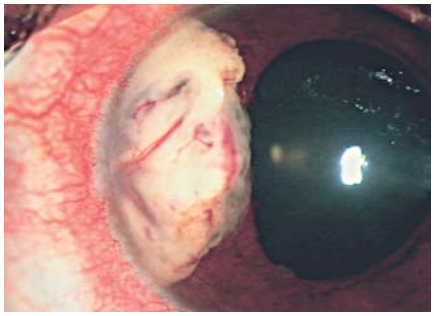


Fig. 6.46.2: Metastatic carcinoma of the iris—from breast carcinoma

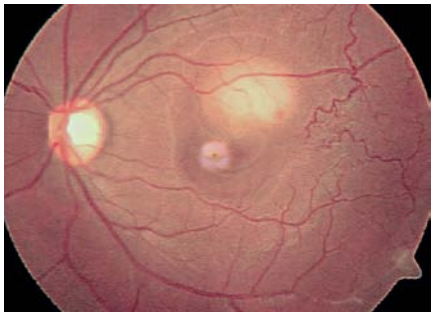


Fig. 6.46.3: Metastatic carcinoma of the choroid—from breast carcinoma

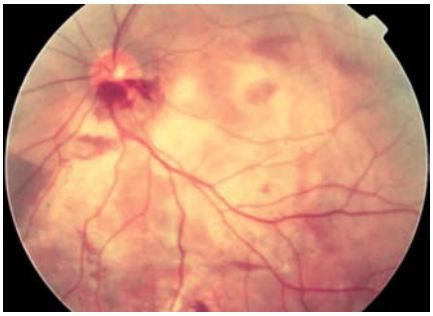


Fig. 6.46.4: Metastatic carcinoma of the choroid—from renal carcinoma

CHAPTER

7 Pupil and its Abnormalities

CHAPTER OUTLINE

Abnormal Pupil

- Acoria
- Corectopia
- Polycoria/pseudopolycoria
- Persistent pupillary membrane
- Small (miotic) pupil
- Large (mydriatic) pupil
- Abnormalities in the shape of pupil
- Anisocoria

Leukocoria or White Pupillary (Amaurotic Cat's Eye) Reflex

- Congenital cataract
- Retinoblastoma
- Retinopathy of prematurity
- Toxocara endophthalmitis
- Persistent hyperplastic primary vitreous (PHPV)
- Retinal detachment
- Coat's disease
- Choroidal coloboma

Abnormal Pupil

Acoria

- Means absence of pupil
- *Congenital*: Very rare, bilateral or unilateral condition; an extreme form of persistent pupillary membrane **(Fig. 7.1.1)**
- Anterior chamber remained formed by aqueous drained via micro-openings in the pupillary area
- *Acquired*: Also very rare and unilateral **(Fig. 7.1.2)**
- Patient ultimately develops hypotony and atrophic bulbi
- *Treatment:* Nd:YAG laser pupilloplasty may be tried **(Fig. 7.1.3)**.

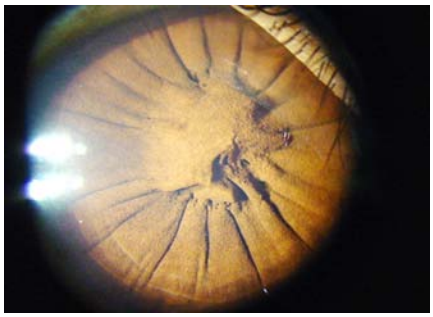


Fig. 7.1.1: Acoria—persistent pupillary membrane—congenital

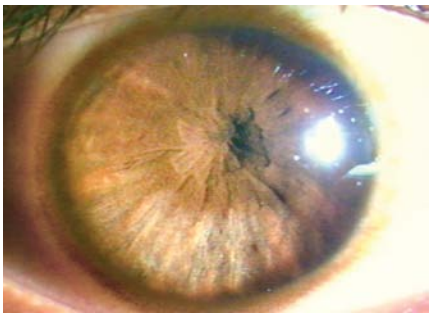


Fig. 7.1.2: Acoria—occlusio pupillae—acquired

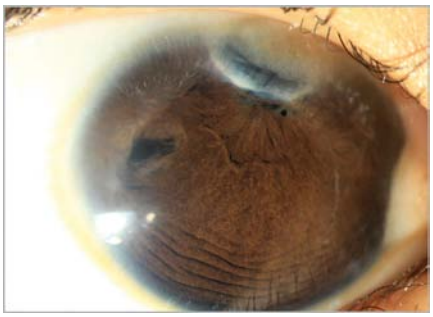


Fig. 7.1.3: YAG laser pupilloplasty

Corectopia

- Displacement of the pupil from its normal position, usually more towards the nasal side **(Fig. 7.2.1)**
- May be associated with ectopia lentis **(Figs 7.2.2A and B)**
- *The other conditions are:* Coloboma iris **(Fig. 7.2.3)**, Axenfeld anomaly **(Fig. 7.2.4)**; iridocorneal endothelial (ICE) syndrome **(Figs 7.2.5A and B)**, after trauma **(Fig. 7.2.6)** or after intraocular surgery.

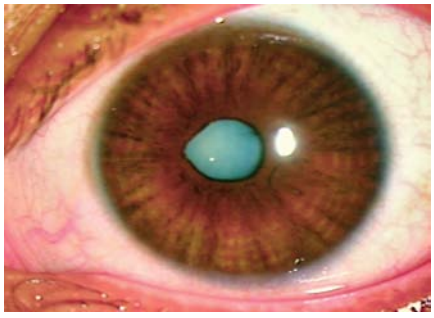


Fig. 7.2.1: Corectopia

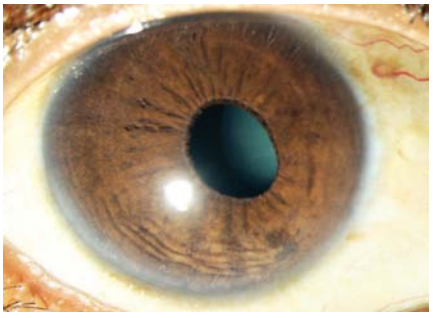


Fig. 7.2.2A: Corectopia—ectopia lentis *et pupillae*

Fig. 7.2.2B: Corectopia—ectopia lentis *et pupillae* after dilatation

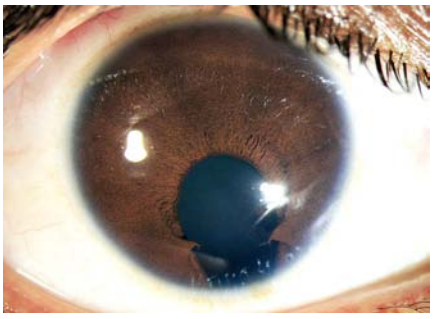


Fig. 7.2.3: Corectopia—iris coloboma

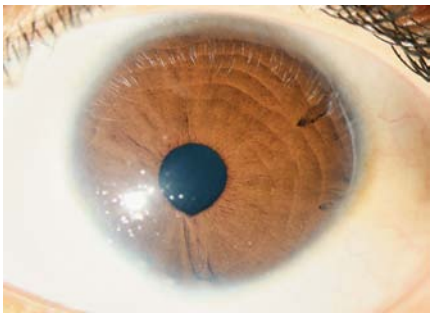


Fig. 7.2.4: Corectopia—Axenfeld anomaly

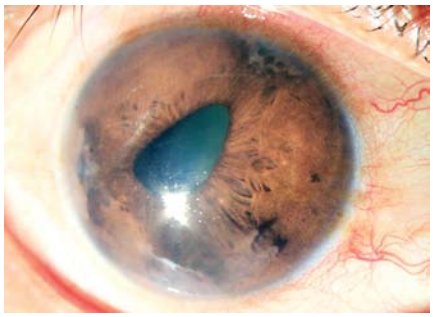


Fig. 7.2.5A: Corectopia—ICE syndrome

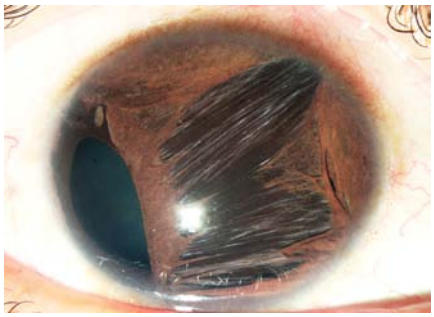


Fig. 7.2.5B: Corectopia—ICE syndrome

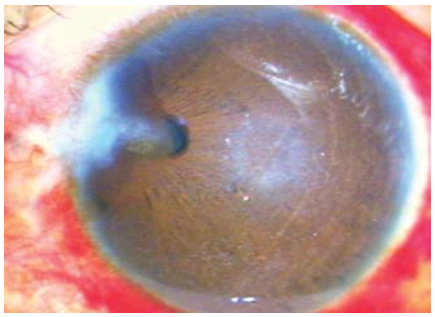


Fig. 7.2.6: Corectopia—post-traumatic

Polycoria/Pseudopolycoria

- Multiple pupils
- *True polycoria:* Extremely rare, multiple pupils, each having a sphincter muscle
- *Pseudopolycoria:* Not uncommon, found in ICE syndrome **(Figs 7.3.1 to 7.3.4)**, trauma and after surgery **(Figs 7.3.5 and 7.3.6)**.

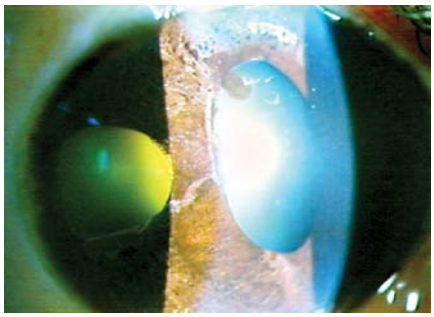


Fig. 7.3.1: Pseudopolycoria—ICE syndrome

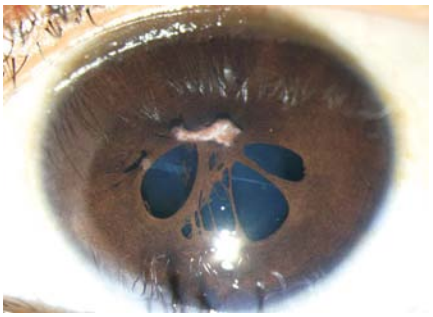


Fig. 7.3.2: Pseudopolycoria—ICE syndrome

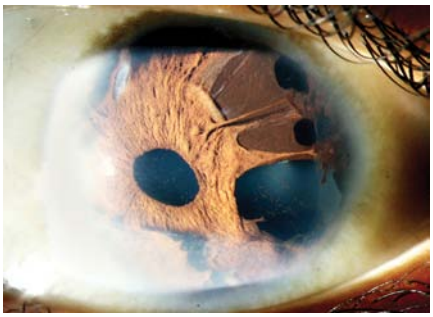


Fig. 7.3.3: Pseudopolycoria—ICE syndrome

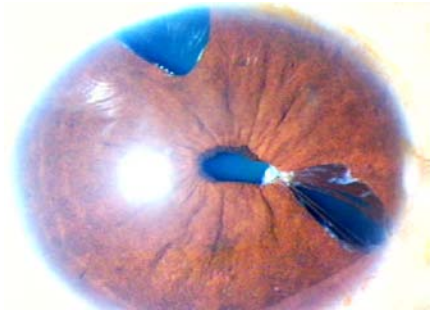


Fig. 7.3.4: Pseudopolycoria—ICE syndrome and post-surgical

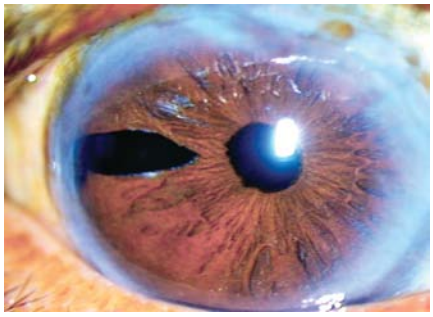


Fig. 7.3.5: Pseudopolycoria—post-surgical

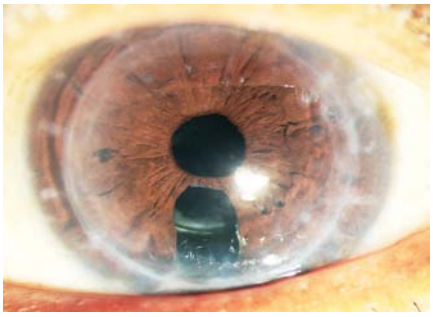


Fig. 7.3.6: Pseudopolycoria—post-surgical

Persistent Pupillary Membrane

- It is the continued existence of the part of anterior vascular sheath of the lens, a fetal structure, which normally disappears shortly before birth
- Appears as fine strands of membrane, running inwards from the collarette inserting into the anterior lens capsule **(Figs 7.4.1A and B)**
- Usually, it does not interfere with vision
- Rarely, it may block the pupil
- Associated with anterior polar cataract and in those cases, the membrane is fixed with the opacity **(Fig. 7.4.2)**
- In some cases, part of the pupil is involved **(Figs 7.4.3 and 7.4.4)**
- *Treatment:* Surgical intervention is only necessary if it obstructs the visual axis.

Small (Miotic) Pupil

- Normally it varies between 2-4 mm, but depends on level of illumination
- Pupil size less than 2 mm
- *Causes:*
  - Extreme of ages **(Fig. 7.5.1)**
  - In bright light
  - Opium/morphine addict **(Fig. 7.5.2)**
  - Pontine hemorrhage
  - Healed iridocyclitis **(Figs 7.5.3 and 7.5.4)**
  - Use of miotics, e.g. pilocarpine as in angle closure glaucoma **(Fig. 7.5.5)** or in open angle glaucoma **(Fig. 7.5.6)**
  - Pseudoexfoliation **(Fig. 7.5.7)**
  - During sleep.

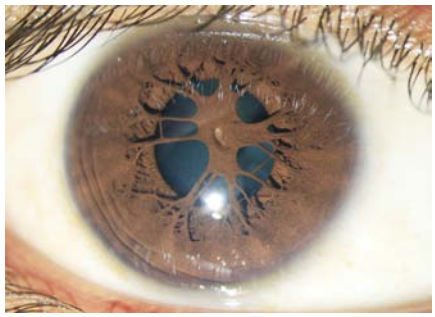


Fig. 7.4.1A: Persistent pupillary membrane—RE

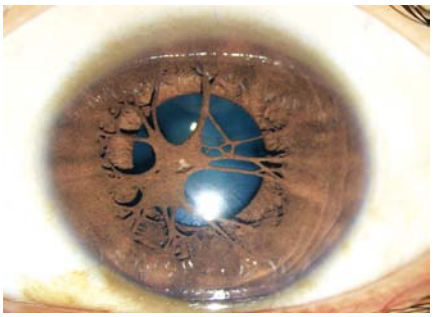


Fig. 7.4.1B: Persistent pupillary membrane—LE

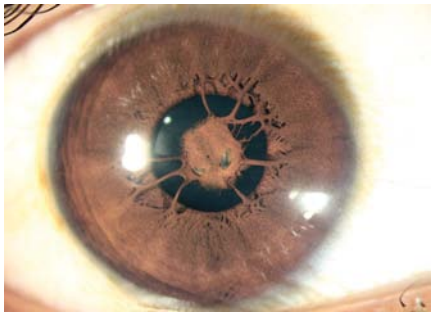


Fig. 7.4.2: Persistent pupillary membrane—central part adherence

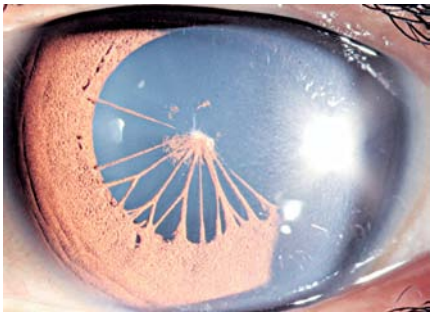


Fig. 7.4.3: Persistent pupillary membrane partial

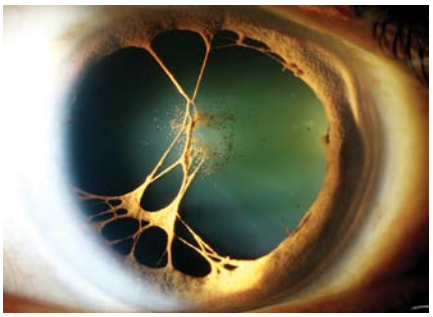


Fig. 7.4.4: Persistent pupillary membrane with epicapsular star

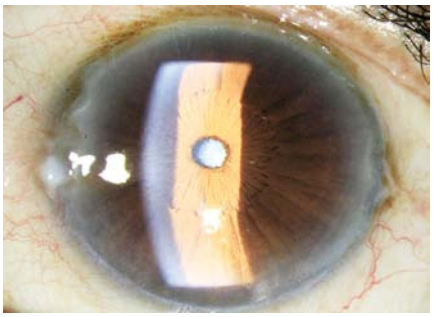


Fig. 7.5.1: Miotic pupil—aging

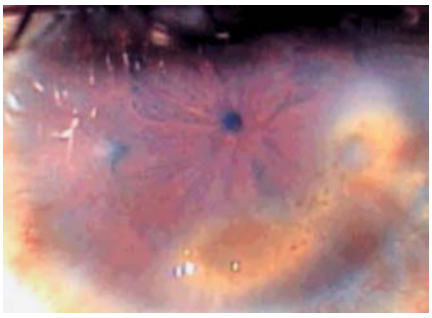


Fig. 7.5.2: Extreme miotic pupil—opium addict

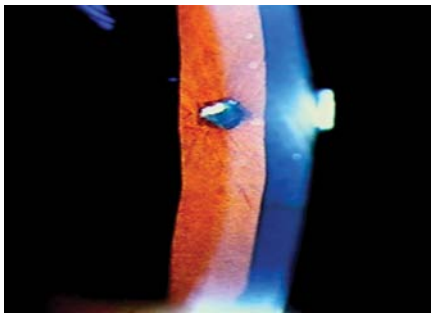


Fig. 7.5.3: Miotic pupil—healed iritis

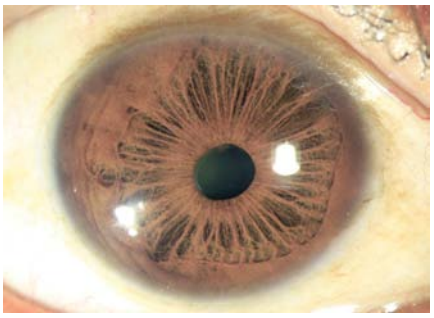


Fig. 7.5.4: Miosis—healed iritis

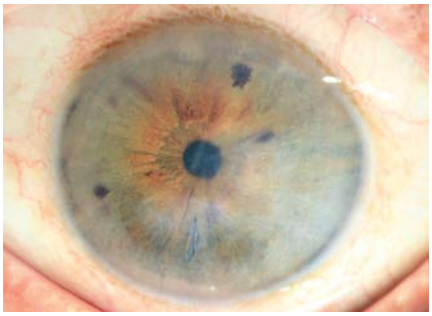


Fig. 7.5.5: Miosis—pilocarpine in narrow angle glaucoma

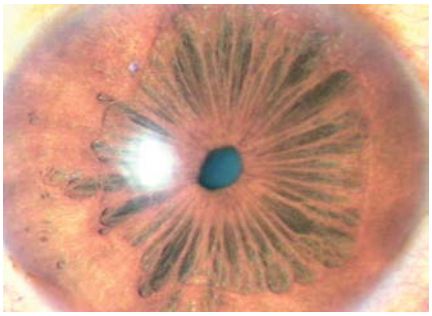


Fig. 7.5.6: Miosis—pilocarpine in POAG

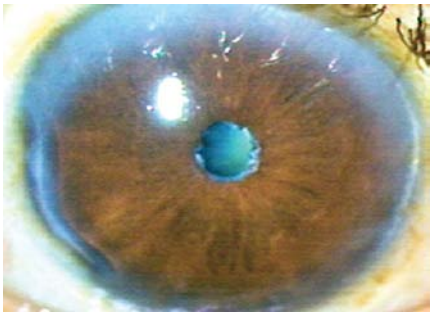


Fig. 7.5.7: Miosis—pseudoexfoliation syndrome

Large (Mydriatic) Pupil

- Pupil size is 6 mm or more
- *Causes:*
  - Congenital anomaly of sphincter pupillae **(Figs 7.6.1A and B)**
  - In dark
  - Optic atrophy **(Figs 7.6.2A to D)**
  - Absolute glaucoma
  - Comatose patient/head injury
  - Third nerve palsy
  - Post-traumatic **(Figs 7.6.3 to 7.6.5)**
  - Use of mydriatics **(Figs 7.6.6A and B)**.

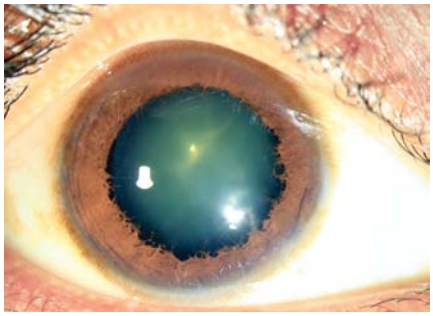


Fig. 7.6.1A: Mydriasis—congenital anomaly of sphincter pupil RE

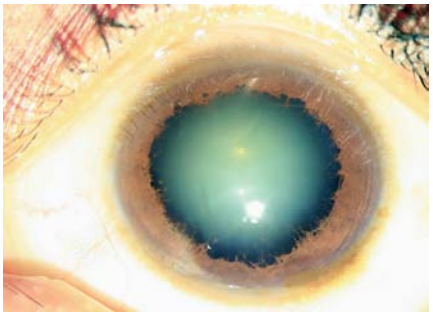


Fig. 7.6.1B: Mydriasis—congenital anomaly of sphincter pupil LE

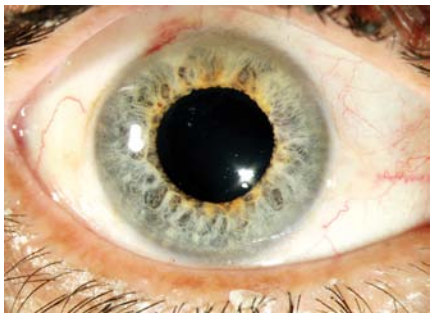


Fig. 7.6.2A: Mydriasis—optic atrophy—RE

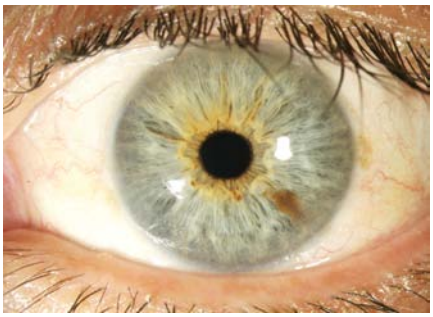


Fig. 7.6.2B: Normal pupil—LE

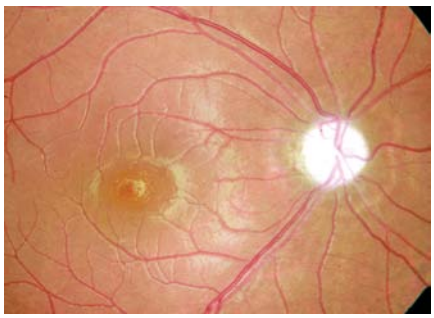


Fig. 7.6.2C: Mydriasis—optic atrophy same eye in **Figure 7.6.2A**

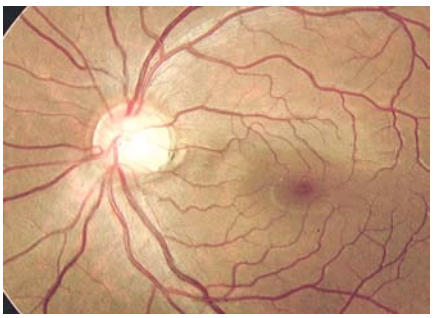


Fig. 7.6.2D: Normal optic disk same eye in **Figure 7.6.2B**

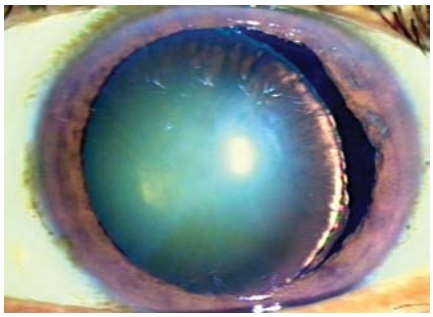


Fig. 7.6.3: Mydriasis—post-traumatic with subluxation

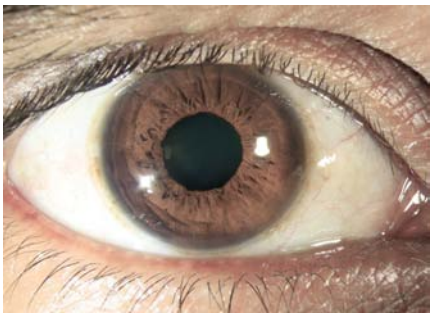


Fig. 7.6.4A: Mydriasis—post-traumatic with subluxation—RE

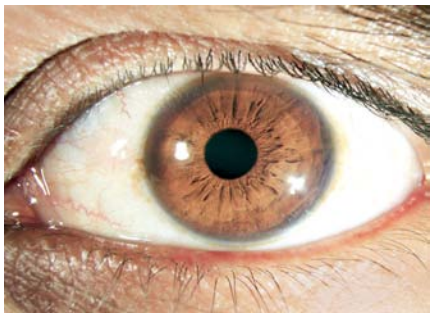


Fig. 7.6.4B: Normal pupil—LE

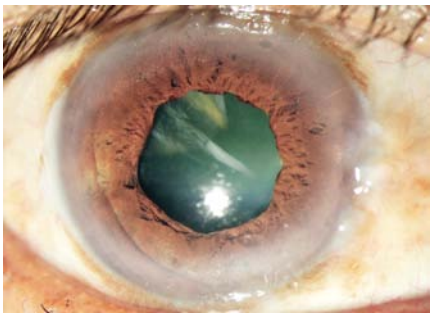


Fig. 7.6.5: Mydriasis—post-traumatic sphincteric tear

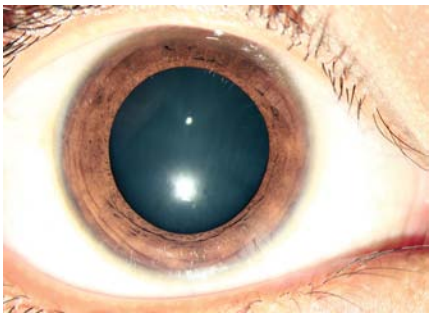


Fig. 7.6.6A: Mydriasis—pharmacological—RE

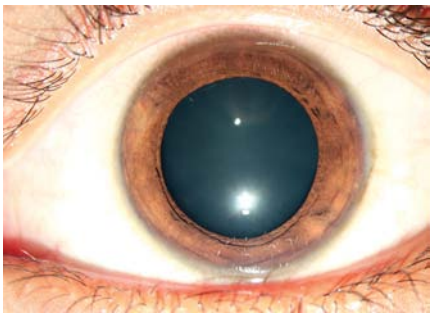


Fig. 7.6.6B: Mydriasis—pharmacological—LE

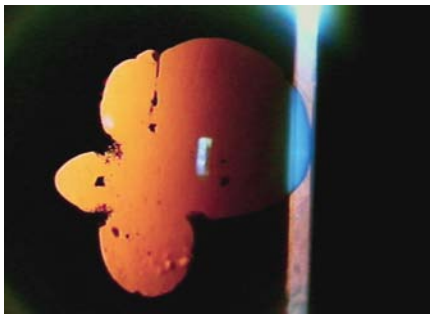


Fig. 7.7.1A: Irregular pupil—iritis

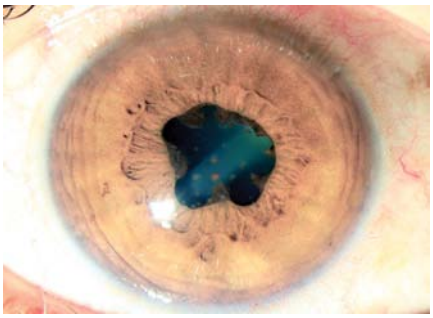


Fig. 7.7.1B: Irregular pupil—iritis

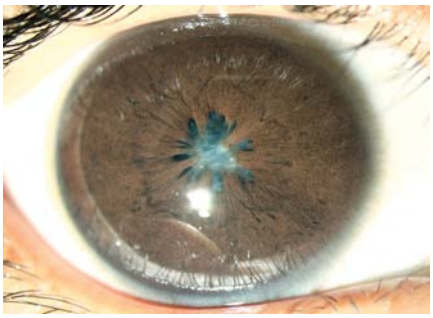


Fig. 7.7.1C: Irregular pupil—iritis

Abnormalities in the Shape of Pupil

- *Irregular:* Iritis **(Figs 7.7.1A to C)** or post-traumatic, surgical sphincterotomy **(Fig. 7.7.2)**
- *D-shaped:* Iridodialysis **(Fig. 7.7.3)**
- *Pear-shaped:* Incarceration of iris with the wound post-traumatic **(Fig. 7.7.4A)**; after surgery **(Fig. 7.7.4B)** or Axenfeld anomaly **(Fig. 7.7.4C)**
- *Festooned:* Iridocyclitis **(Figs 7.7.5A and B)**

Fig. 7.7.2: Irregular pupil—sphincterotomy

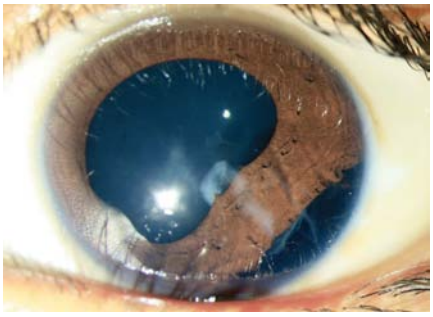


Fig. 7.7.3: D-shaped pupil in iridodialysis

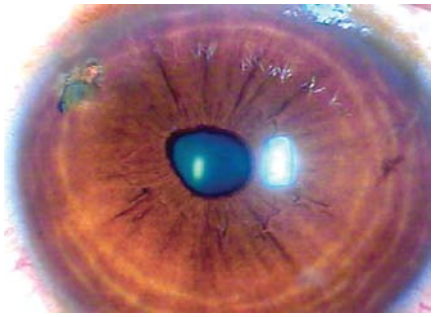


Fig. 7.7.4A: Pear-shaped pupil—traumatic

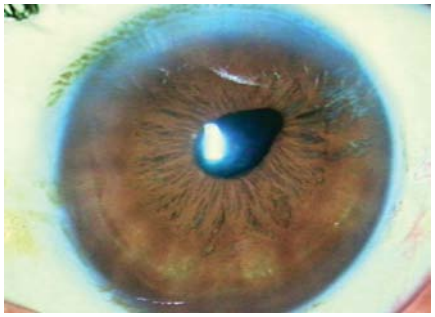


Fig. 7.7.4B: Pear-shaped pupil—post-surgical

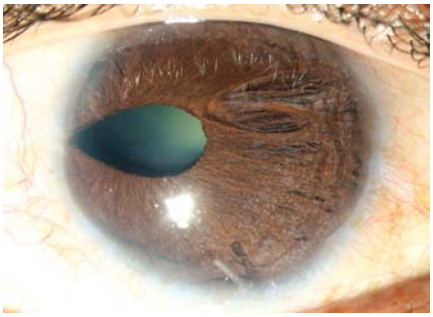


Fig. 7.7.4C: Pear-shaped—Axenfeld anomaly

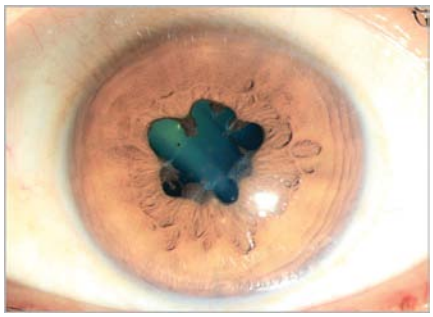


Fig. 7.7.5A: Festooned pupil—iridocyclitis

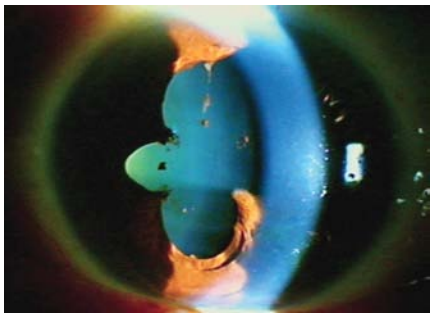


Fig. 7.7.5B: Festooned pupil—iridocyclitis

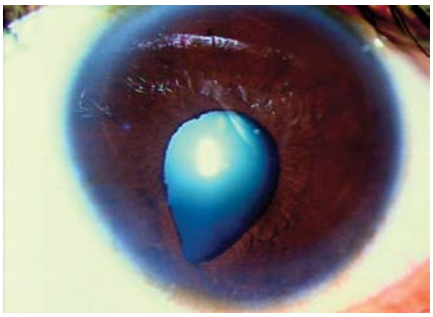


Fig. 7.7.6: Inverted pear-shaped—incomplete coloboma

- *Inverted pear-shaped:* Coloboma *(inferonasal)* of the iris **(Fig. 7.7.6)**, penetrating injury with iris prolapse **(Fig. 7.7.7)**
- *Mid-dilated and oval:* Acute attack of angle closure glaucoma **(Fig. 7.7.8)**
- *Boat- or Hammock-shaped:* Vitreous loss in cataract surgery and remaining in anterior chamber; also in ICE syndrome **(Fig. 7.7.9)**
- *Elliptical, triangular or square*: IOL optic capture **(Figs 7.7.10 to 7.7.12)**
- *Slit-like pupil*: Axenfeld-Rieger's syndrome **(Fig. 7.7.13)** and also in IOL optic capture **(Fig. 7.7.14)**
- *Updrawn pupil:* After cataract surgery **(Figs 7.7.15A and B)**
- *Large semicircular and up:* Complete iridectomy in cataract surgery **(Fig. 7.7.16)**.

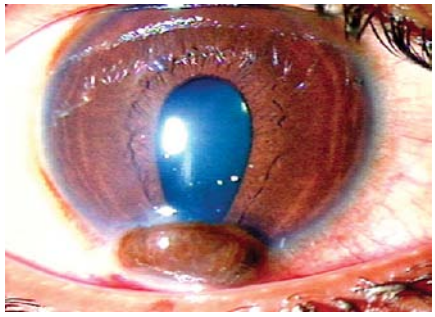


Fig. 7.7.7: Inverted pear shaped—perforating injury

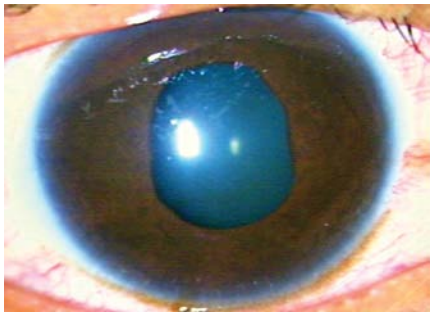


Fig. 7.7.8: Oval pupil—acute attack ACG

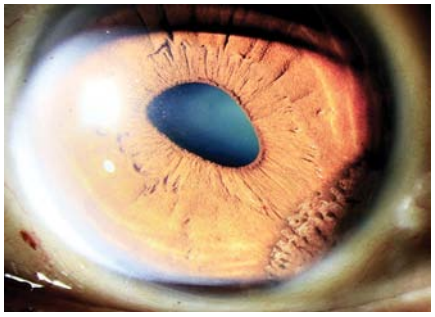


Fig. 7.7.9: Boat-shaped pupil—Cogan-Reese syndrome

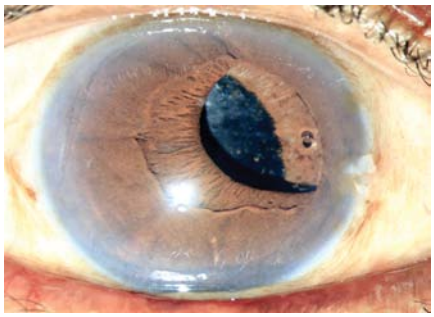


Fig. 7.7.10A: Elliptical pupil—IOL optic capture

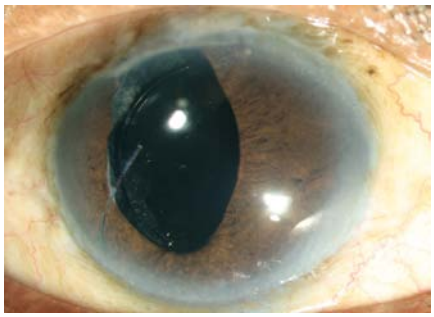


Fig. 7.7.10B: Elliptical pupil—haptic catch in AC

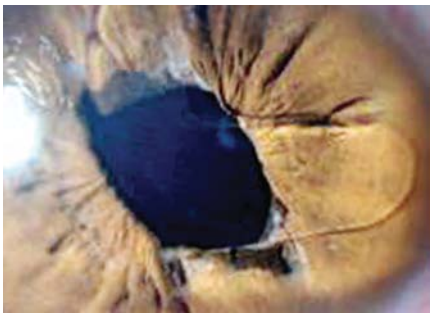


Fig. 7.7.11: Square pupil—IOL optic capture

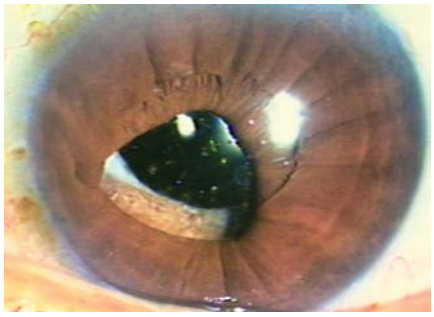


Fig. 7.7.12: Triangular pupil—iris capture

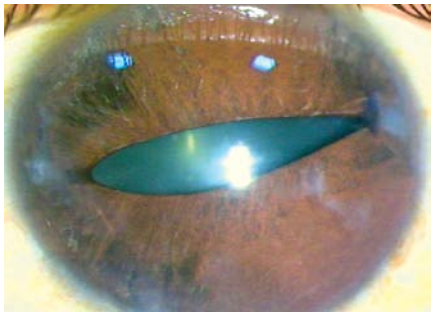


Fig. 7.7.13: Slit-like pupil—Axenfeld Rieger's syndrome

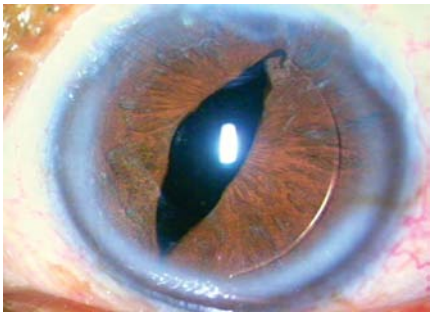


Fig. 7.7.14: Slit-like pupil—IOL optic capture

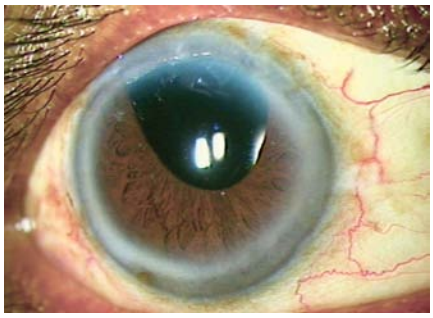


Fig. 7.7.15A: Updrawn pupil following cataract surgery

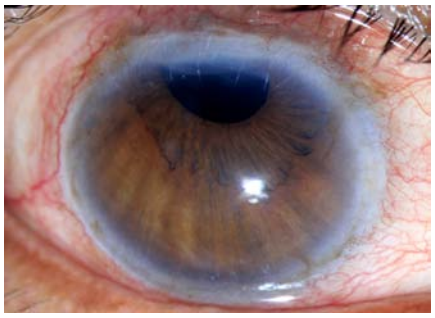


Fig. 7.7.15B: Updrawn pupil following cataract surgery

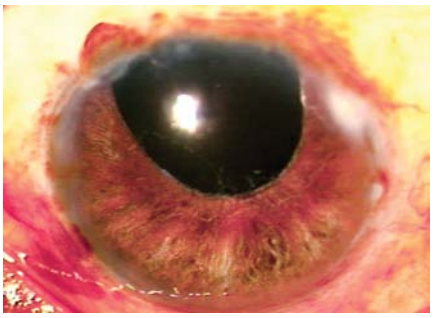


Fig. 7.7.16: Pupil after complete iridectomy in aphakia

Anisocoria

- Unequal size of the pupil between two eyes
- Pupil of one eye is normal, and the other eye is either miotic or mydriatics
- *Causes:*
  - Horner's syndrome
  - 3rd nerve palsy **(Fig. 7.8.1)**
  - Use of miotic or mydriatic in one eye **(Figs 7.8.2A and B)**
  - Optic atrophy in one eye **(Fig. 7.8.3)**
  - Adie's pupil **(Figs 7.8.4A and B)**
  - Springing pupil **(Fig. 7.8.5).**

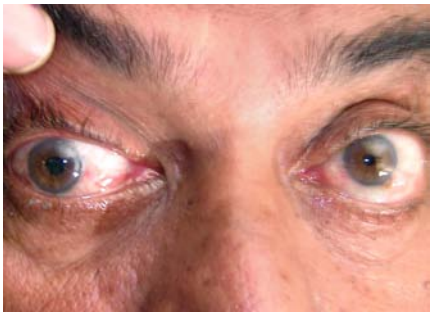


Fig. 7.8.1: Anisocoria—third nerve palsy

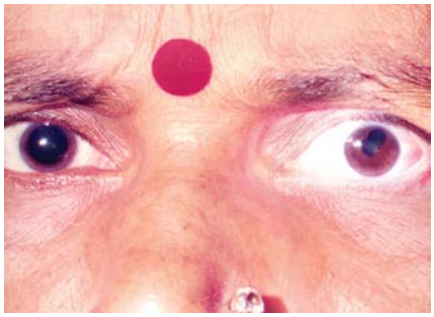


Fig. 7.8.2A: Anisocoria—iritis—use of mydriatics

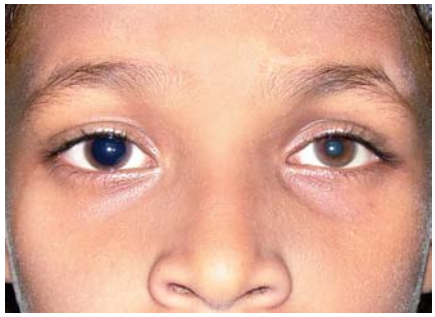


Fig. 7.8.2B: Anisocoria—mydriatic use

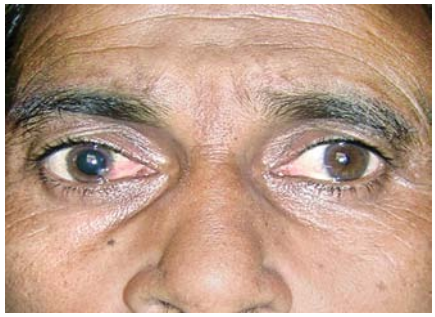


Fig. 7.8.3: Anisocoria—optic atrophy

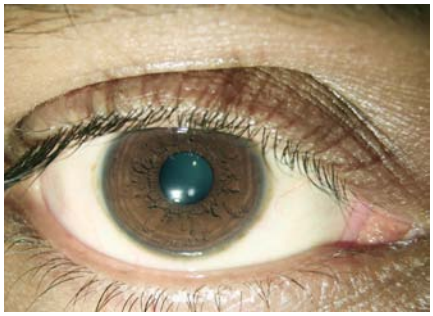


Fig. 7.8.4A: Anisocoria—normal pupil—RE

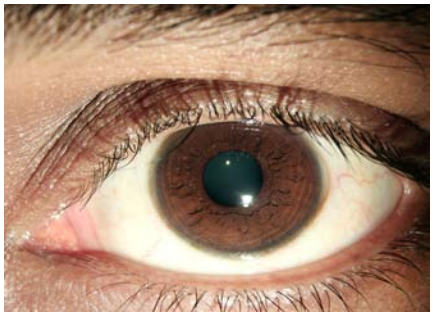


Fig. 7.8.4B: Anisocoria—Adie's pupil—LE

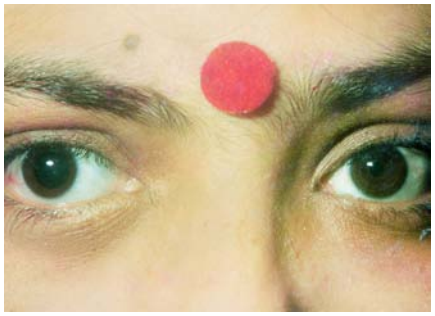


Fig. 7.8.5: Anisocoria—springing pupil

Leukocoria or White Pupillary (Amaurotic Cat's Eye) Reflex

- Relatively common condition
- May be unilateral or bilateral
- *Common causes:* Congenital cataract, retinoblastoma, retinopathy of prematurity, toxocara endophthalmitis, persistent hyperplastic primary vitreous, retinal dysplasia, Coat's disease, choroidal coloboma, etc.
- *Congenital cataract*
  - Unilateral or bilateral **(Figs 7.9.1 and 7.9.2)**
  - Opacity in the lens clearly indicates the presence of cataract
  - Discussed in *Chapter 8*
- *Retinoblastoma*
  - Unilateral, progressive, malignant condition
  - Usual age at diagnosis is 18 months **(Figs 7.9.3 and 7.9.4)**

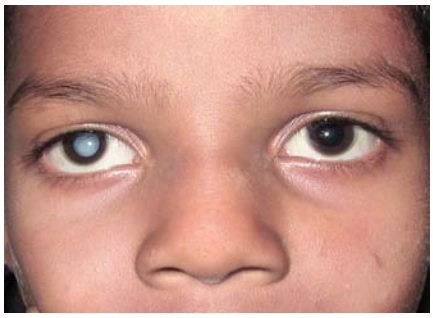


Fig. 7.9.1: Leukocoria—congenital cataract—unilateral

Fig. 7.9.2: Leukocoria—congenital cataract—bilateral

Fig. 7.9.3: Leukocoria—retinoblastoma—LE

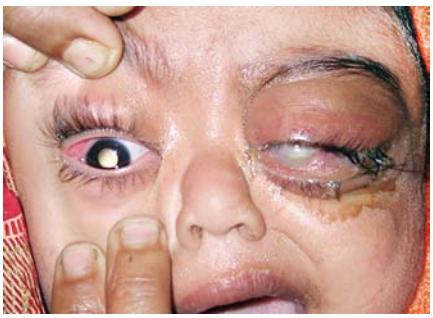


Fig. 7.9.4: Leukocoria—retinoblastoma—bilateral

- No inflammatory sign in anterior segment
- Ophthalmoscopy shows, a pearly-white mass with presence of secondary calcification **(Fig. 7.9.5)**
- Lens is usually transparent
- Intraocular pressure is high.

- *Retinopathy of prematurity*
  - Prematurity and low birth weight with history of prolonged exposure to oxygen
  - Bilateral in 100% of cases **(Fig. 7.9.6)**
  - First noted in neonatal period
  - Presence of tractional retinal detachment
  - Intraocular pressure is normal.
- *Toxocara endophthalmitis*
  - Unilateral with history of contact with pet cat or dog
  - Presentation is between 2 to 9 years of age
  - Signs of inflammation in anterior segment and vitreous **(Fig. 7.9.7)**
  - Retinal detachment, low intraocular pressure and eventually, the eye may be phthisical **(Fig. 7.9.8)**.
- *Persistent hyperplastic primary vitreous (PHPV)*
  - Usually unilateral and first noted in neonatal period
  - Associated with microphthalmos and cataract **(Fig. 7.9.9)**
  - Elongated ciliary processes are visible through the dilated pupil
  - Intraocular pressure may be high.
- *Retinal detachment*
  - Unilateral usually present at birth
  - Pink or white retrolental mass **(Figs 7.9.10A and B)**

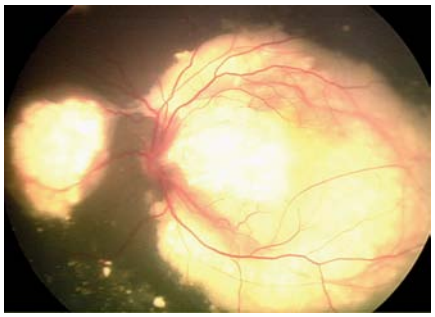


Fig. 7.9.5: Leukocoria—retinoblastoma—endophytic lesion

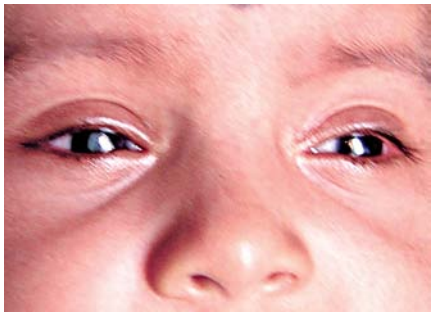


Fig. 7.9.6: Leukocoria—retinopathy of prematurity

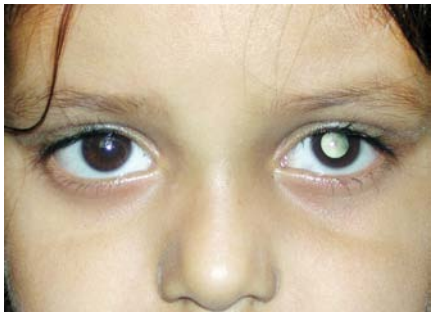


Fig. 7.9.7: Leukocoria—toxocara endophthalmitis

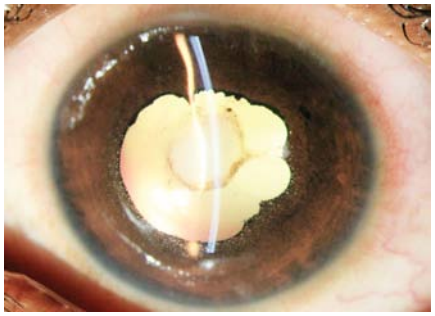


Fig. 7.9.8: Leukocoria—toxocara endophthalmitis

Fig. 7.9.9: Leukocoria—PHPV

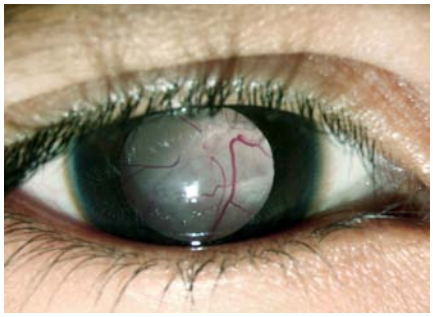


Fig. 7.9.10A: Leukocoria—retinal detachment

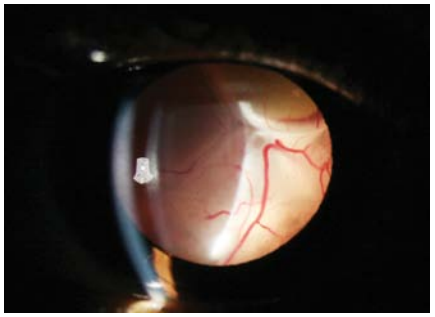


Fig. 7.9.10B: Leukocoria—retinal detachment

- Microphthalmic eye with shallow anterior chamber and elongated ciliary processes
- Associated with severe systemic abnormalities.

- *Coat's disease*
  - Unilateral, occurs primarily in older boys
  - large areas of retinal or subretinal exudates with cholesterol crystals
  - Dilated and tortuous retinal blood vessels at the posterior pole
  - Exudative detachment as a retrolental mass occurs eventually **(Figs 7.9.11A and B)**
- *Choroidal coloboma*
  - Unilateral or bilateral, present at birth
  - Leukocoria is only with a large choroidal coloboma with microphthalmos
  - Inferonasal in location
  - Fundus shows, the defect is at the embryonic fissure, with shiny-white sclera **(Fig. 7.9.12) (*see* Fig. 6.2.2)**.

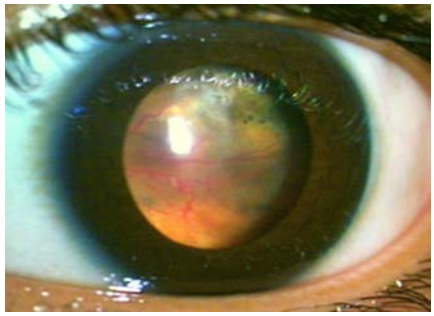


Fig. 7.9.11A: Leukocoria—Coat's disease

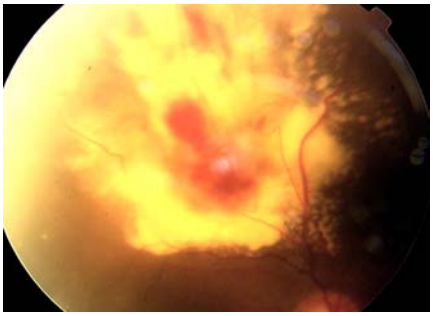


Fig. 7.9.11B: Leukocoria—Coat's disease

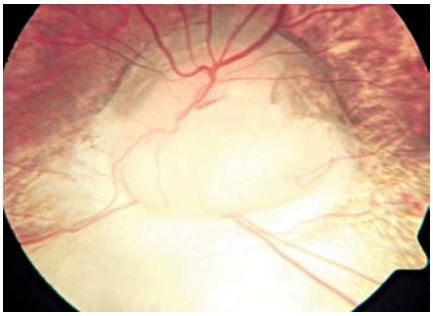


Fig. 7.9.12: Choroidal coloboma and shiny sclera

CHAPTER

8 Diseases of the Lens

CHAPTER OUTLINE

Congenital Conditions of Lens

- Coloboma of the lens
- Anterior lenticonus
- Posterior lenticonus
- Spherophakia (microspherophakia)
- Ectopia lentis
- Marfan's syndrome
- Homocystinuria
- Weill-Marchesani syndrome

Lens-Induced Ocular Diseases

- Phacolytic glaucoma
- Phacogenic (phacomorphic) glaucoma
- Phacotoxic uveitis
- Phaco-anaphylactic uveitis

Cataract

- Lental opacity

Congenital or Developmental Cataract

- Anterior polar cataract
- Posterior polar cataract
- Coronary cataract
- Zonular (lamellar) cataract
- Central pulverulent cataract
- Rubella cataract
- Sutural cataract
- Blue dot cataract
- Other types of congenital/developmental cataract

Senile (Adult) Cataract

- Cortical cataract
- Stages of cortical cataract
  - Immature stage
  - Intumescent stage
  - Mature stage
  - Hypermature stage
- Cupuliform cataract (Posterior subcapsular cataract)
- Nuclear cataract
- Other types of adult cataract
- Surgical techniques in senile cataracts

Specific Cataract Entities

- Diabetic cataract
- Complicated cataract
- Galactose cataract
- Traumatic cataract
- Glaukomflecken

Opacities in Aphakia or Pseudophakia

- Anterior capsular opacification/fibrosis
- Posterior capsular opacification or 'after cataract'
- Soemmering's ring
- Cortical matter behind the IOL
- Capsular inflammatory deposits/plaques

- Posterior capsular folds
- Pseudophakic deposits/opacities/defects

Displacement of the Crystalline Lens and IOL

- Subluxation of the crystalline lens
- Dislocation of the crystalline lens
- Displacement of the IOL
- Miscellaneous pseudophakic conditions

Aphakia

Congenital Conditions of Lens

Coloboma of the Lens

- Rare unilateral or bilateral condition, may occur with other typical colobomatous defects of the uveal tract **(Fig. 8.1.1)**; or in isolation **(Figs 8.1.2A to C)** or in atypical position, called atypical coloboma
- Notching of the lens at the inferior equator or other area with absence of zonules **(Figs 8.1.3A and B)**.

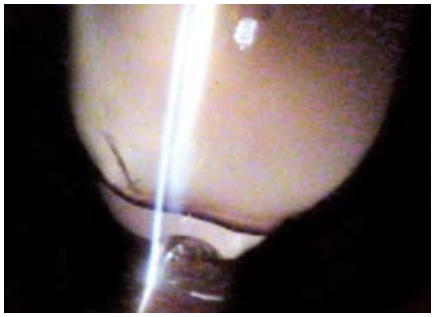


Fig. 8.1.1: Typical coloboma of the lens

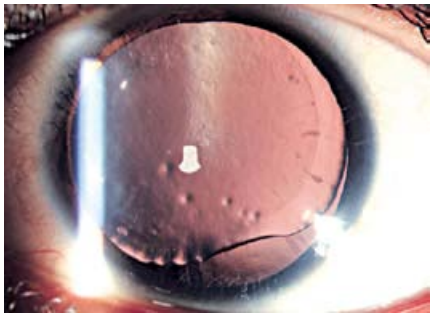


Fig. 8.1.2A: Isolated lens coloboma—typical position

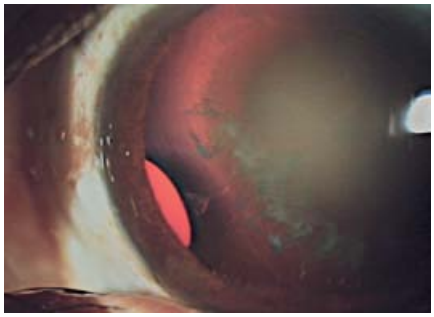


Fig. 8.1.2B: Coloboma of the lens—atypical

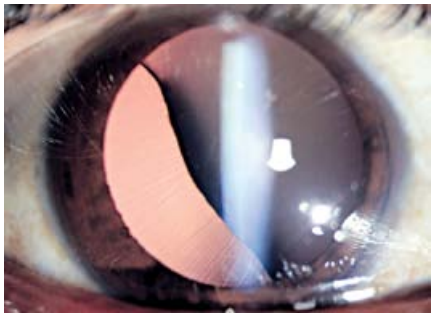


Fig. 8.1.2C: Coloboma of the lens—atypical

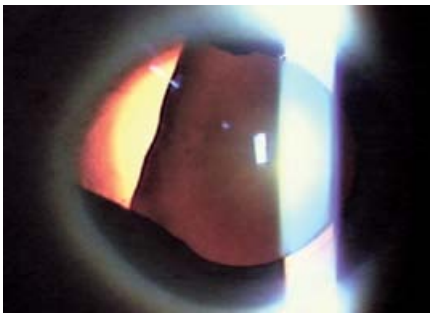


Fig. 8.1.3A: Atypical lens coloboma with atypical iris coloboma

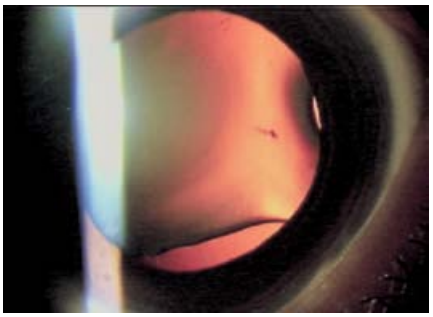


Fig. 8.1.3B: Double coloboma of the lens—typical and atypical

Anterior Lenticonus

- Rare bilateral condition, often associated with Alport's syndrome
- Anterior conical projection at the center of the lens **(Figs 8.2.1A and B)**
- Oil-droplet sign on distant direct ophthalmoscopy
- High lenticular myopia
- May be associated with lenticular opacification.

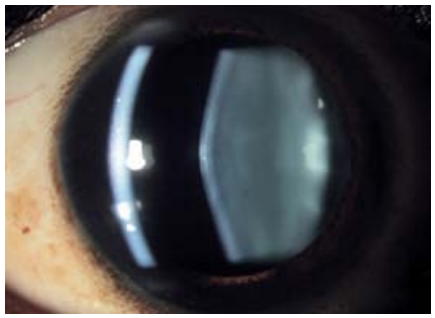


Fig. 8.2.1A: Anterior lenticonus—RE

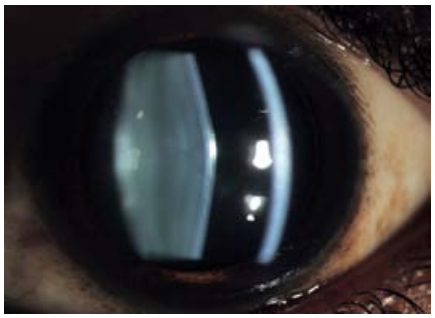


Fig. 8.2.1B: Anterior lenticonus—LE

Posterior Lenticonus

- Rare bilateral condition, may be associated with Lowe's syndrome
- Posterior conical **(Fig. 8.3.1)** or globular, called lentiglobus **(Fig. 8.3.2)** bulge in the axial zone of the lens
- May be associated with posterior subcapsular or capsular opacification of the lens
- *Treatment:* With visual impairment, go for phacoemulsification with IOL. Adequate measures are to be taken to tackle posterior capsular rent.

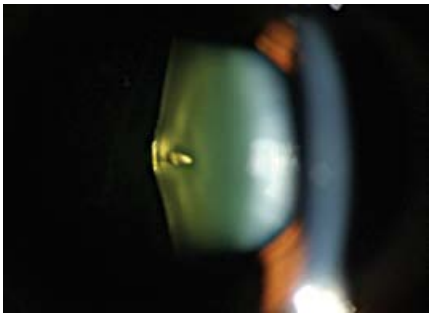


Fig. 8.3.1: Posterior lenticonus with posterior subcapsular cataract

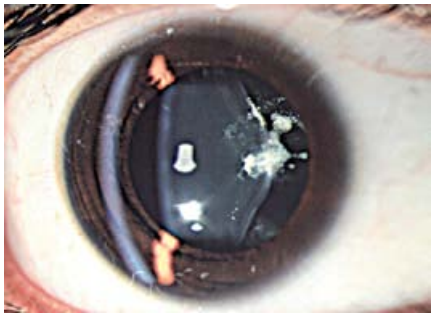


Fig. 8.3.2: Posterior lentiglobus with PPC

Spherophakia (Microspherophakia)

- Smaller diameter lens with spherical shape **(Fig. 8.4.1)**. Zonules can be seen 360 degrees beyond the lens equator **(Figs 8.4.2A to D)**
- Subluxation or anterior dislocation of the lens is common with corneal touch **(Figs 8.4.3A and B)**
- Induced lenticular myopia
- Pupillary block glaucoma may occur and this glaucoma is aggravated by miotics and relieved by mydriatics (called *inverse glaucoma*)
- Associated with Weill-Marchesani syndrome
- *Treatment:* It is difficult in most cases. One can go for phacoemulsification and IOL with adequate capsular support. Or intracapsular cataract operation with scleral fixation IOL.

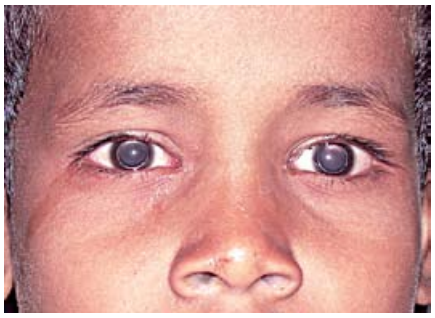


Fig. 8.4.1: Spherophakia—bilateral

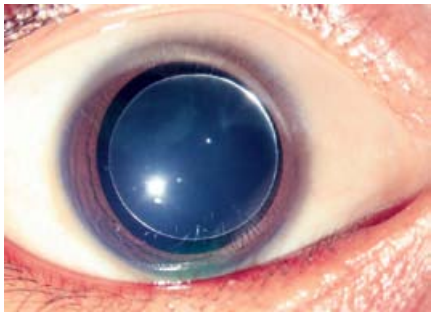


Fig. 8.4.2A: Spherophakia—RE

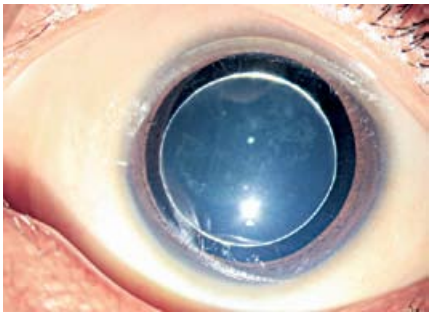


Fig. 8.4.2B: Spherophakia—LE

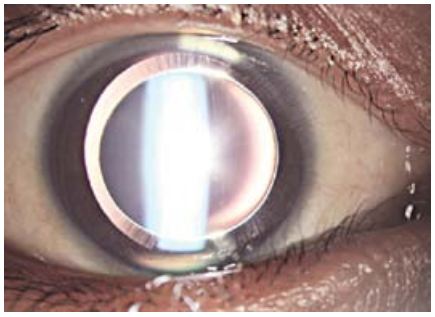


Fig. 8.4.2C: Spherophakia in retroillumination—RE

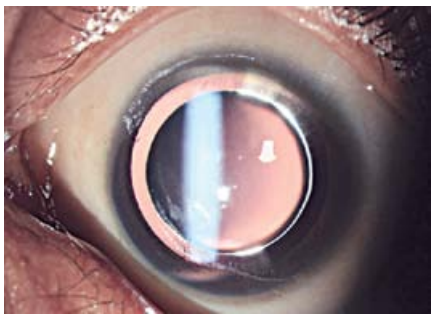


Fig. 8.4.2D: Spherophakia in retroillumination—LE

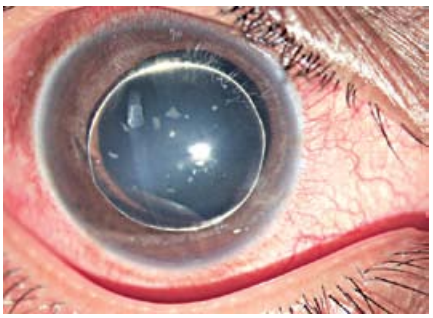


Fig. 8.4.3A: Spherophakia BL anterior dislocation of the lens

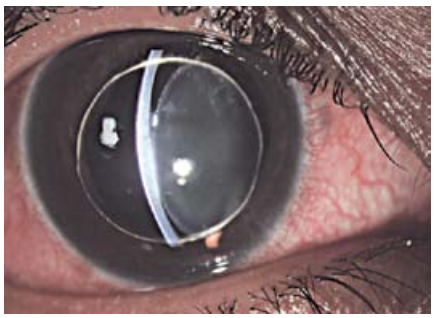


Fig. 8.4.3B: Spherophakia lenticulo-corneal touch—RE (slit section)

Ectopia Lentis

- Ectopia lentis is a congenital or developmental bilateral subluxation or dislocation of the lens **(Figs 8.5.1A and B)**
- Diagnosis should be confirmed after full dilatation of the pupil, otherwise the surgeon can miss the diagnosis **(Figs 8.5.2A and B)**

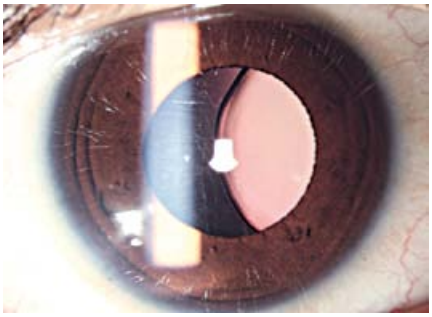


Fig. 8.5.1A: Ectopia lentis

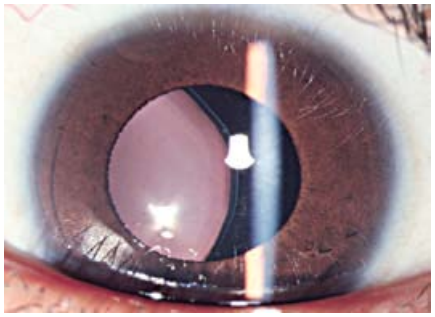


Fig. 8.5.1B: Ectopia lentis

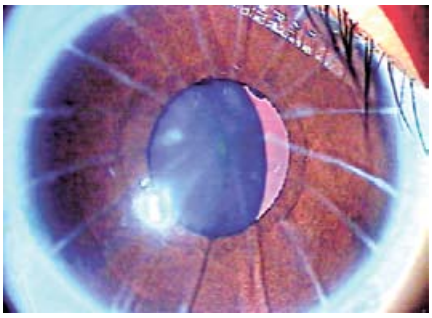


Fig. 8.5.2A: Ectopia lentis—detected after dilatation

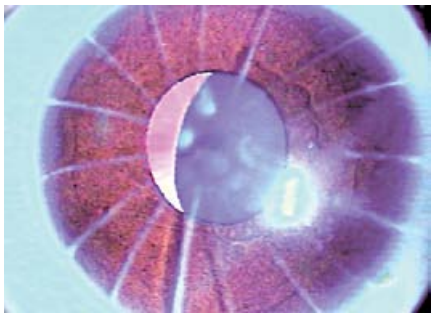


Fig. 8.5.2B: Ectopia lentis—detected after dilatation

- Edge of the displaced lens appears in the pupillary area as a dark or golden crescent **(Fig. 8.5.3)**
- Displaced lens may touch the cornea **(Figs 8.5.4A and B)**
- The pupillary area may be aphakic in some cases **(Fig. 8.5.5)**
- It may be associated with aniridia **(Fig. 8.5.6)**
- They are mainly hereditary in nature
- *Causes of ectopia lentis:*
  - *Ectopia lentis et pupillae:* Pupil and lens are displaced in opposite or same direction **(Figs 8.5.7A and B)**
  - Marfan's syndrome
  - Homocystinuria
  - Weill-Marchesani syndrome
  - Ehlers-Danlos syndrome
  - Hyperlysinemia
  - Familial ectopia lentis
  - Sulfite oxidase deficiency.

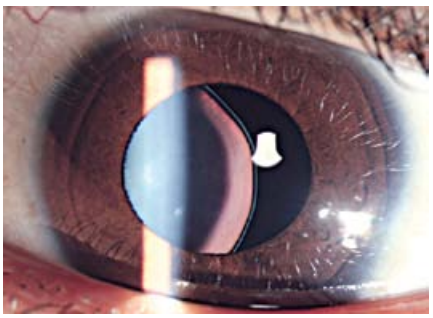


Fig. 8.5.3: Ectopia lentis—golden crescent

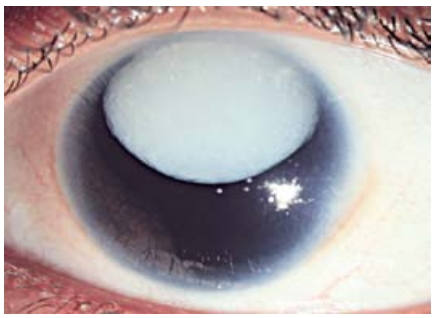


Fig. 8.5.4A: Ectopia lentis with aniridia

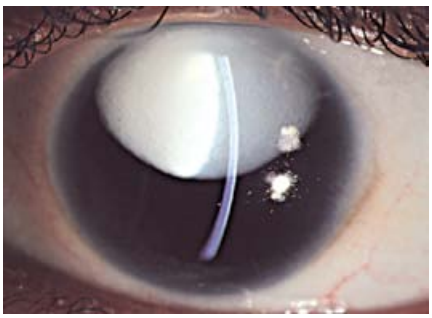


Fig. 8.5.4B: Ectopia lentis with aniridia—corneal touch

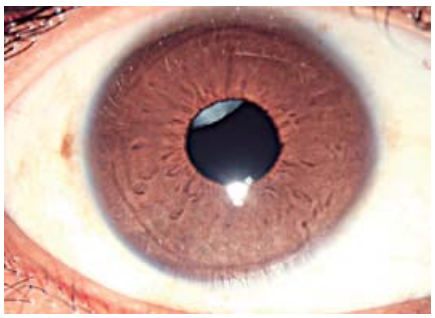


Fig. 8.5.5: Ectopia lentis—mostly aphakic

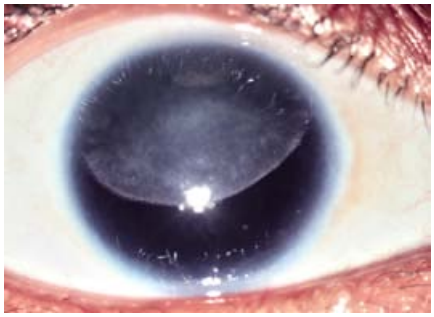


Fig. 8.5.6: Ectopia lentis with aniridia

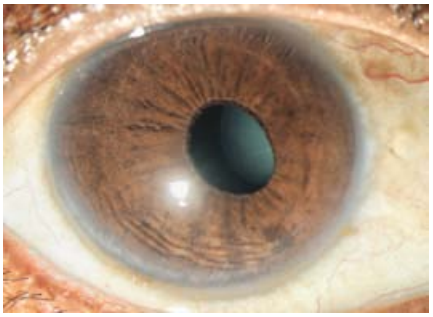


Fig. 8.5.7A: Ectopia lentis et pupillae

Fig. 8.5.7B: Ectopia lentis et pupillae

Marfan's Syndrome

- Autosomal dominant, a multisystem mesodermal dysplasia
- Lens is typically subluxated in upward and inward direction **(Figs 8.6.1A and B)**
- May be subluxated in up and out **(Fig. 8.6.2)** or any quadrant (**Fig. 8.6.3**)
- Iris hypoplasia causes poor pupillary dilatation (**Fig. 8.6.4**)
- *Systemic features:*
  - Arachnodactyly (spider fingers) (**Fig. 8.6.5**)

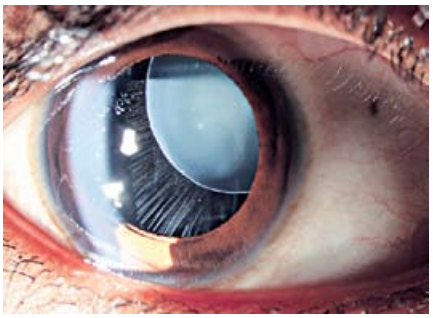


Fig. 8.6.1A: Marfan's syndrome—RE—displacement up and in

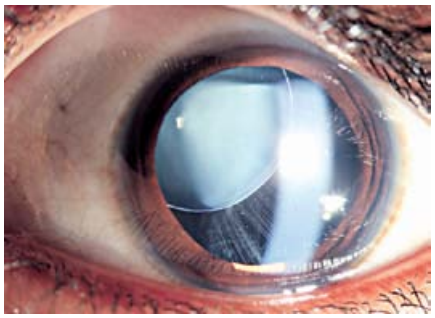


Fig. 8.6.1B: Marfan's syndrome—LE—displacement up and in

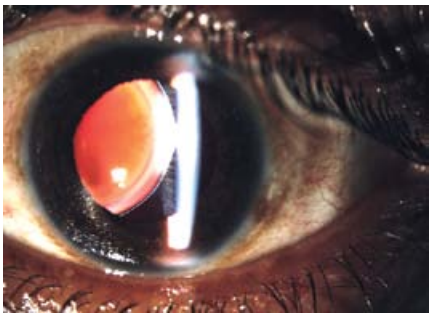


Fig. 8.6.2: Marfan's syndrome—RE—displacement up and out

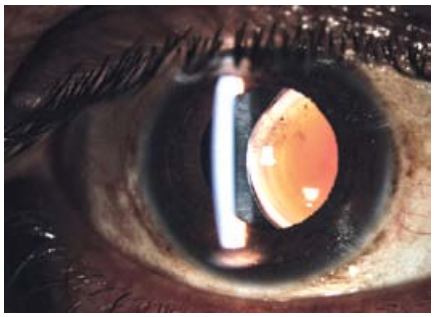


Fig. 8.6.3: Marfan's syndrome—LE—displacement outwards

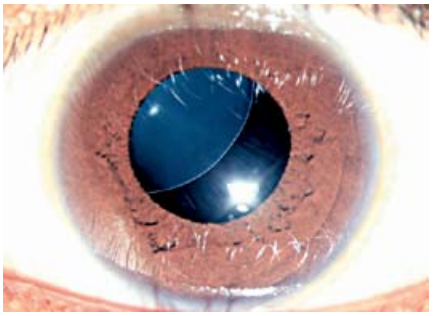


Fig. 8.6.4: Poor pupillary dilatation

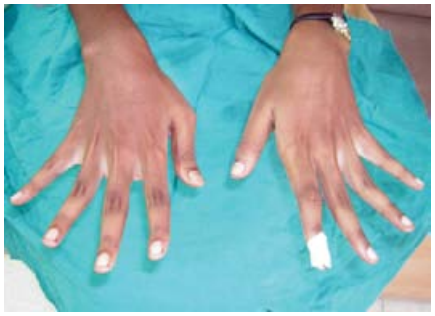


Fig. 8.6.5: Marfan's syndrome—arachnodactyly

- Long extremities (**Fig. 8.6.6**)
- Hyperextensibility of the joints
- Cardiovascular anomalies.

Fig. 8.6.6: Marfan's syndrome—long extremities

Homocystinuria

- Autosomal recessive, an inborn error of metabolism
- Inability of convert methionine to cystine
- Lens displacement is typically downward and outward **(Figs 8.7.1A and B)** or it may be downwards **(Figs 8.7.2A and B)**
- Diagnosis is confirmed by urine test with sodium nitroprusside
- Anesthetic hazards during operation.

Weill-Marchesani Syndrome

- *Autosomal recessive:* A mesodermal dysplasia
- Short stature, stubby fingers **(Fig. 8.8.1)**
- Microspherophakia, which may be subluxated, usually downward **(Figs 8.8.2A and B)**.

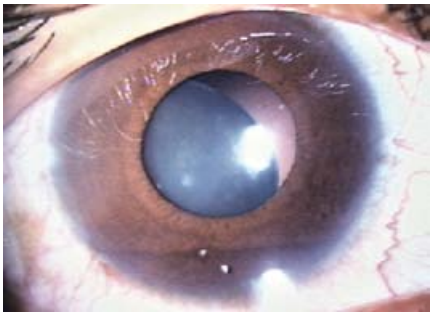


Fig. 8.7.1A: Ectopia lentis—homocystinuria—displacement downwards

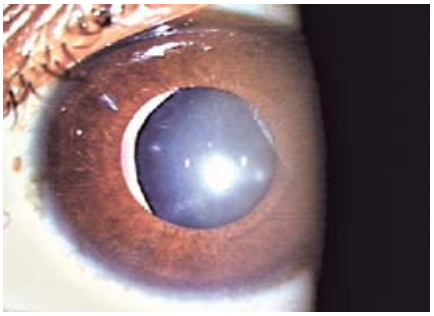


Fig. 8.7.1B: Ectopia lentis—homocystinuria—displacement downwards

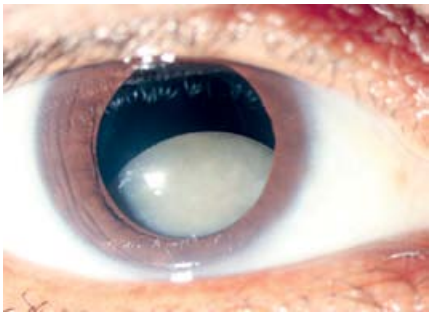


Fig. 8.7.2A: Ectopia lentis—homocystinuria displacement downwards—RE

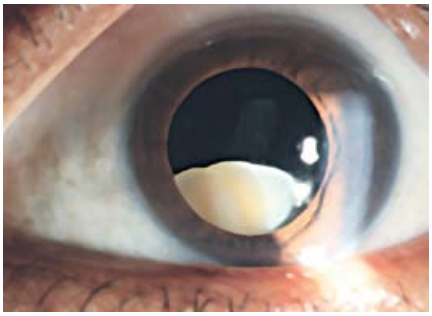


Fig. 8.7.2B: Ectopia lentis—homocystinuria displacement downwards—LE

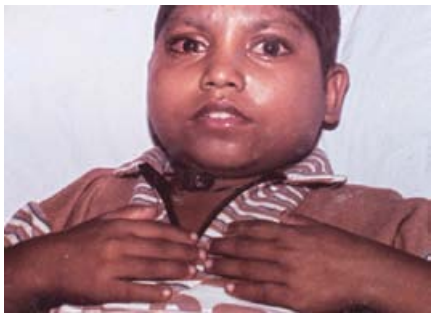


Fig. 8.8.1: Weill-Marchesani syndrome

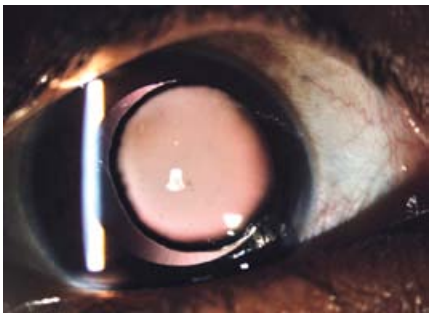


Fig. 8.8.2A: Spherophakia—ectopia lentis

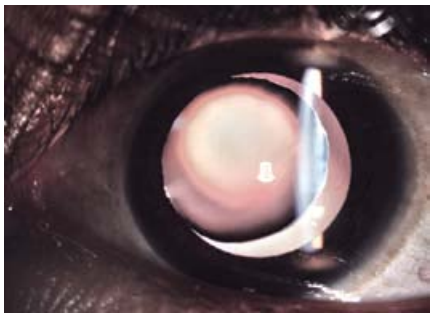


Fig. 8.8.2B: Spherophakia—ectopia lentis

- *Treatment of the ectopia lentis*
  - Spectacles or contact lenses are used to correct the optical defects through the phakic part
  - Pars plana lensectomy with vitrectomy with scleral fixation IOL is better than other surgical means
  - In milder degree of subluxation—Phaco with PCIOL implantation after placing capsular tension rings, either 360-degree and/or segmental ring (CTR) **(Fig. 8.9.1)**.

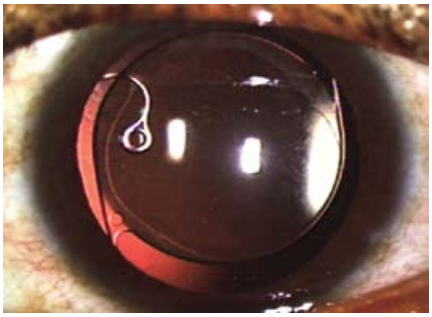


Fig. 8.9.1: Ectopia lentis—PC IOL with CTR

Lens-Induced Ocular Diseases

Phacolytic Glaucoma

- In some hypermature cataract the capsule leaks, and large phagocytes filled with lens material obstruct the trabecular meshwork **(Figs 8.10.1A and B)**
- One of the important causes of secondary open-angle glaucoma
- Anterior chamber is always deep
- With treatment edema gets cleared and milky white fluid absorbed within 2-3 days **(Figs 8.10.2A and B)**
- *Treatment:* Glaucoma must be controlled medically first and then the lens has to be extracted by ECCE/manual SICS with PC IOL.

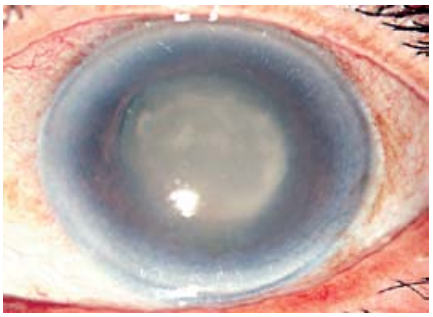


Fig. 8.10.1A: Phacolytic glaucoma

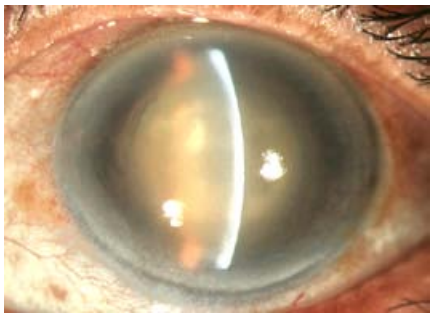


Fig. 8.10.1B: Phacolytic glaucoma

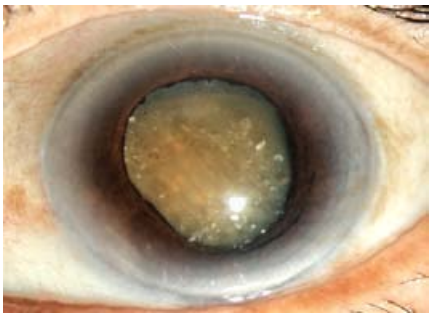


Fig. 8.10.2A: Phacolytic glaucoma—same eye as in **Figure 8.10.1**

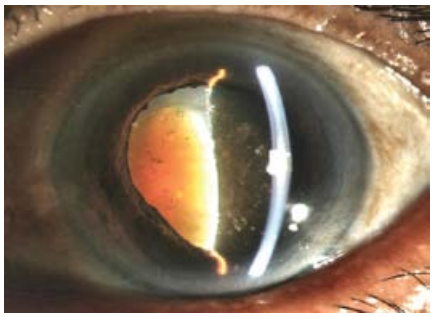


Fig. 8.10.2B: Phacolytic glaucoma—same eye as in **Figure 8.10.1**

Phacogenic (Phacomorphic) Glaucoma

- Rapid swelling of the lens in the intumescent stage **(Fig. 8.11.1A)**
- Anterior chamber becomes very shallow **(Fig. 8.11.1B)**
- May cause a secondary angle-closure glaucoma
- *Treatment:* Urgent control of glaucoma and then followed extraction of lens with PC IOL.

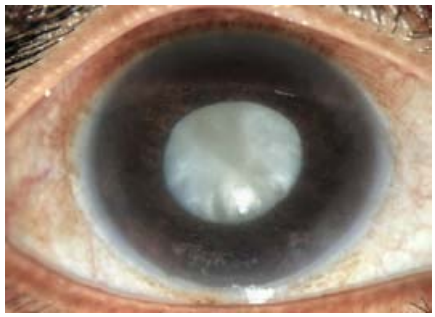


Fig. 8.11.1A: Phacomorphic glaucoma—late presentation with corneal edema

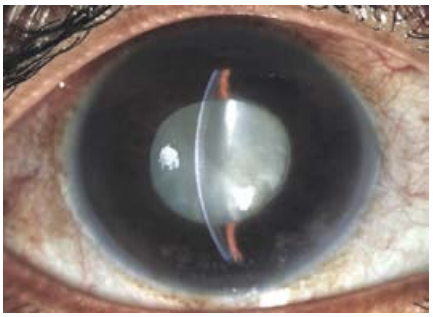


Fig. 8.11.1B: Phacomorphic glaucoma—late presentation with corneal edema

Phacotoxic Uveitis

- Lens proteins are relatively poor antigens
- Sometimes, a granulomatous uveitis may develop and may be with secondary glaucoma **(Fig. 8.12.1)**
- *Treatment:* High dose of systemic corticosteroids and cycloplegic; followed by cataract extraction with IOL implantation
- *Phacoanaphylactic uveitis*
  - Dislocation of lens nucleus or nuclear fragments in the vitreous cavity during an extracapsular extraction or phacoemulsification
  - Causes severe granulomatous uveitis.

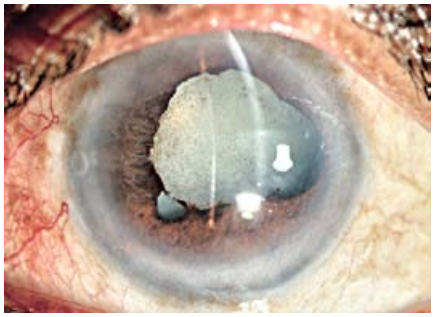


Fig. 8.12.1: Phacotoxic uveitis

Cataract

Any opacity of the lens or its capsule, causing visual impairment, is called cataract (**Figs 8.13.1 to 8.13.4**).

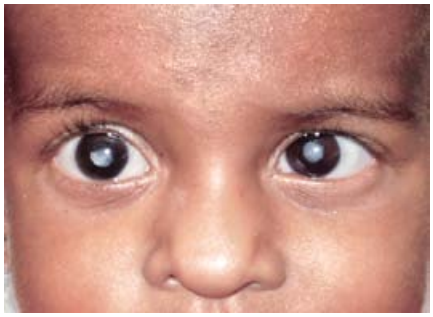


Fig. 8.13.1: Congenital total cataract

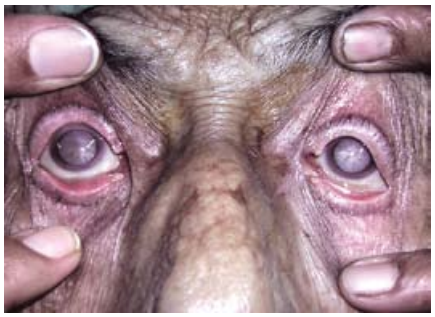


Fig. 8.13.2: Bilateral mature cataract

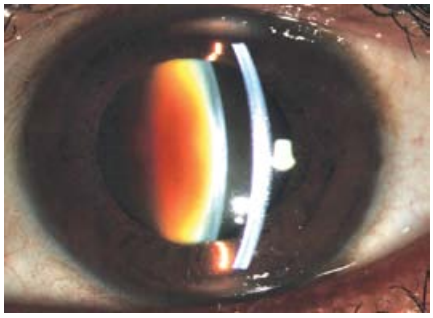


Fig. 8.13.3: Nuclear brown cataract

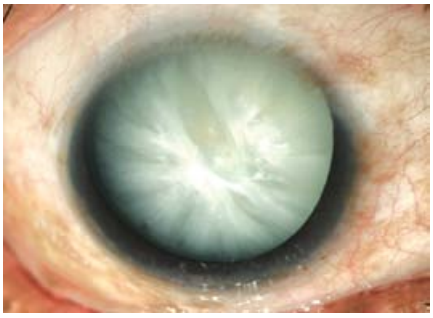


Fig. 8.13.4: White mature cataract

Lental Opacity

- Many congenital small lenticular opacities are stationary **(Figs 8.14.1A and B)**
- Even in senile cataract the opacities may remain localized for many years

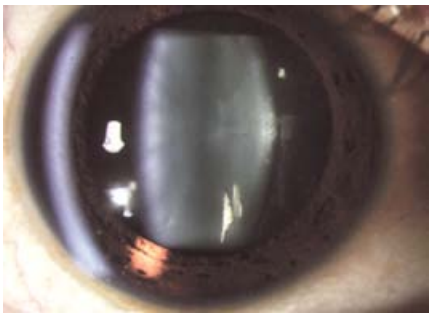


Fig. 8.14.1A: Isolated lental opacity

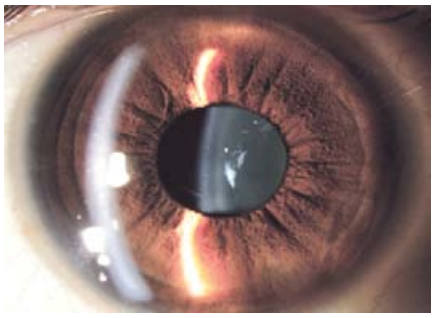


Fig. 8.14.1B: Lental opacity—central

- The peripheral lental opacities are not associated with visual impairment **(Figs 8.14.2A and B)**
- So, it is better to term them as *lental opacity.*

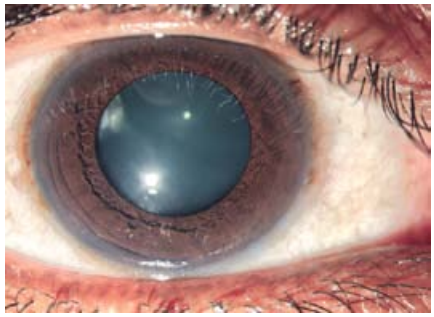


Fig. 8.14.2A: Lental opacity—bilateral—RE

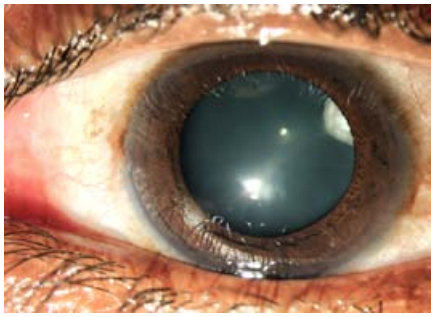


Fig. 8.14.2B: Lental opacity—bilateral—LE

Congenital or Developmental Cataract

- Presents at birth **(Fig. 8.15.1A)**, or develops within first few years after the birth **(Fig. 8.15.1B)**
- Some congenital opacities escape detection at birth
- 1/3rd hereditary, 1/3rd idiopathic and 1/3rd are associated with some systemic problems
- Idiopathic variety may be unilateral (**Fig. 8.15.1C**) or bilateral **(Fig. 8.15.1D)** whereas others are usually bilateral
- One type of opacity may co-exist with other type
- Half of the eyes with congenital cataract have some other ocular anomalies
- *Treatment:* Depends upon laterality, type of cataract and age of presentation. After 2 years of age, phacoemulsification with single-piece hydrophobic IOL implantation. As the chance of PCO is very high, a posterior continuous curvilinear capsulorhexis (PCCC) is necessary to keep the visual axis clear in long-term **(Figs 8.15.2A and B)**.

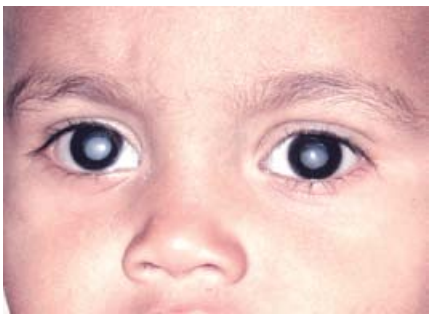


Fig. 8.15.1A: Congenital cataract

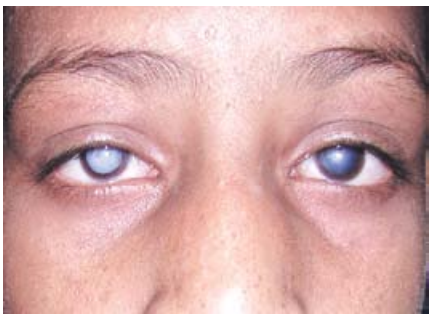


Fig. 8.15.1B: Developmental cataract

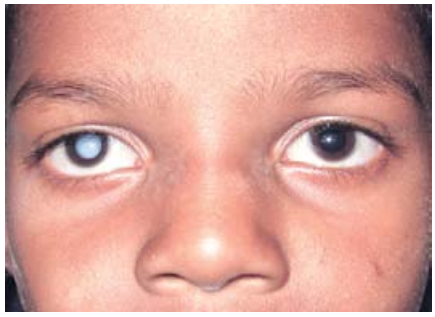


Fig. 8.15.1C: Congenital cataract—unilateral

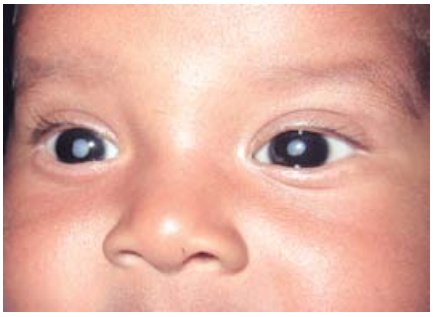


Fig. 8.15.1D: Congenital cataract—bilateral

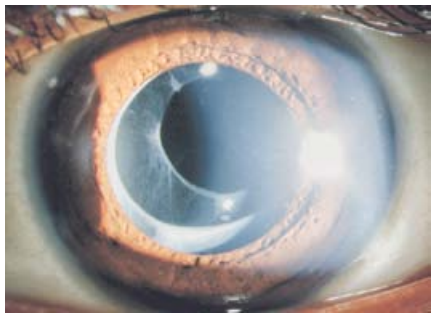


Fig. 8.15.2A: IOL in-the-bag with PCCC

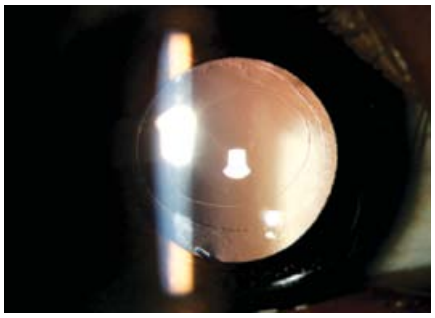


Fig. 8.15.2B: IOL in-the-bag with PCCC

Anterior Polar Cataract

- Rare, usually bilateral and sometimes hereditary
- The opacity involves the capsule or both capsule and anterior cortex **(Fig. 8.16.1)**
- May be pyramidal and may project into the anterior chamber *(pyramidal cataract)* **(Figs 8.16.2A to C)**
- Usually associated with Peter's anomaly **(Fig. 8.16.3)**, posterior keratoconus or persistent pupillary membrane **(Fig. 8.16.4)**
- *Treatment:* Most of the cases do not require any treatment, except when it is obstructing the visual axis.

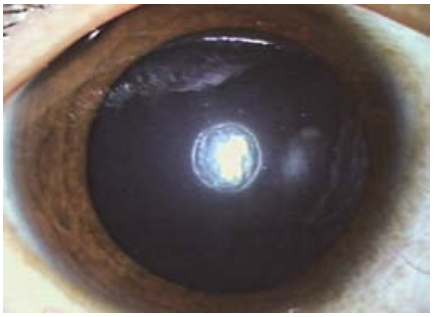


Fig. 8.16.1: Anterior polar cataract

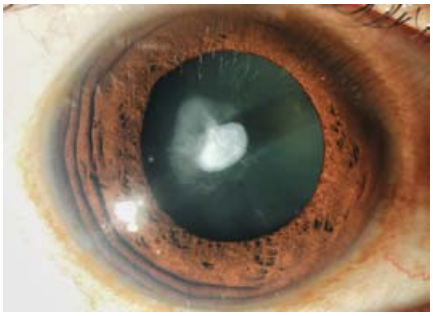


Fig. 8.16.2A: Anterior polar cataract with reduplicating cataract

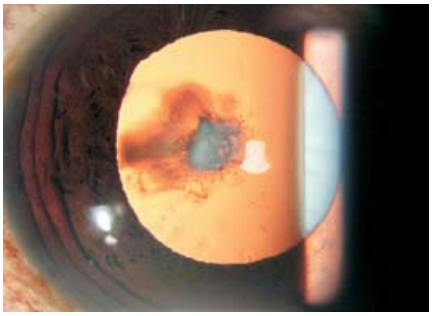


Fig. 8.16.2B: Anterior polar cataract with reduplicating cataract

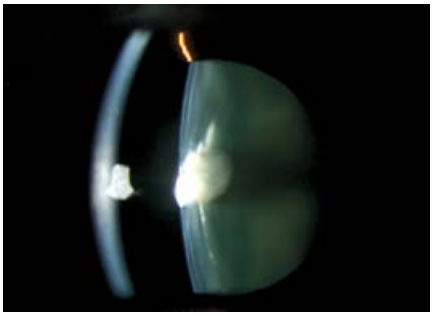


Fig. 8.16.2C: Anterior polar cataract with reduplicating cataract

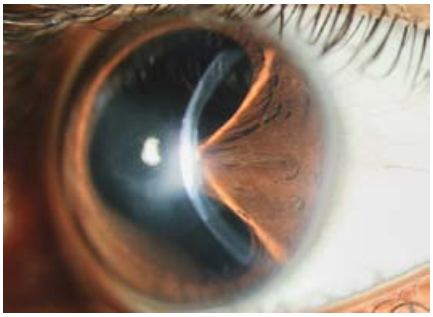


Fig. 8.16.3: Anterior polar cataract—Peter's anomaly

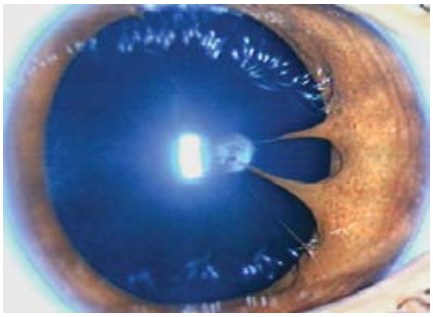


Fig. 8.16.4: Anterior polar cataract—persistent pupillary membrane

Posterior Polar Cataract

- Opacity may involve only the posterior capsule **(Figs 8.17.1A to C)**

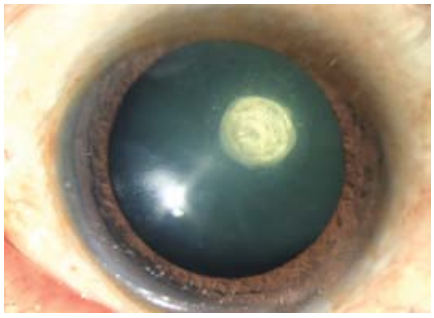


Fig. 8.17.1A: Posterior polar cataract

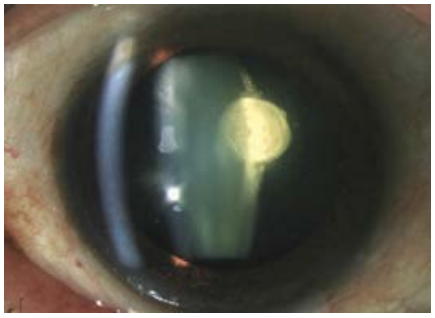


Fig. 8.17.1B: Posterior polar cataract in slit section

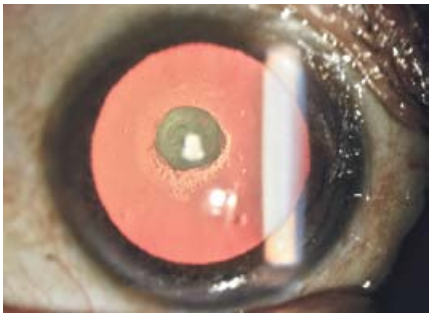


Fig. 8.17.1C: Posterior polar cataract—interoillumination

- Sometimes, it forms a plaque on the posterior cortex with onion ring appearance **(Figs 8.17.2A and B)**
- Few dense dots are seen within the opacity and at the outer ring **(Figs 8.17.3A and B)**
- Posterior capsule may rupture spontaneously or with minor trauma **(Figs 8.17.4 and 8.17.5)**
- May be associated with residue of the attachment of hyaloid artery on the posterior lens capsule as small dots—*Mittendorf's dot* **(Figs 8.17.6A to C)**
- May be with anterior type of persistent hyperplastic primary vitreous (PHPV) **(Figs 8.17.7A to C)**
- *Treatment:* It is difficult. As the incidence of posterior capsular rent is high, the surgeon should take extra precaution during the surgery to prevent this complication.

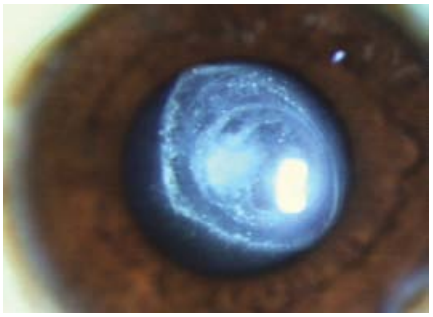


Fig. 8.17.2A: PPC—multiple onion ring appearance

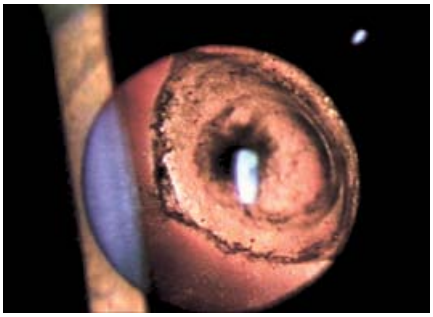


Fig. 8.17.2B: PPC—multiple onion ring appearance

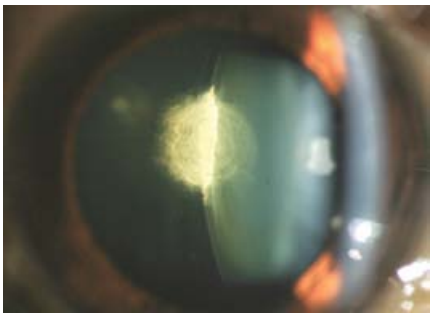


Fig. 8.17.3A: PPC—white dots at the outer ring

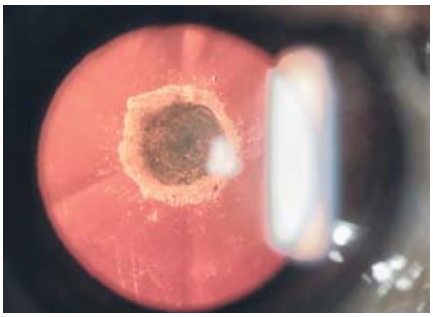


Fig. 8.17.3B: PPC—white dots at the outer ring

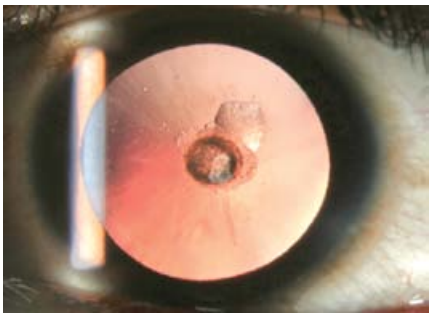


Fig. 8.17.4: Posterior polar cataract partial spontaneous break

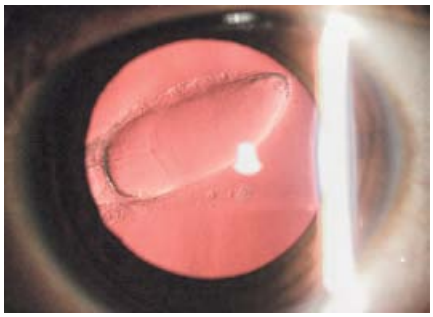


Fig. 8.17.5: PPC—spontaneous large posterior capsular tear

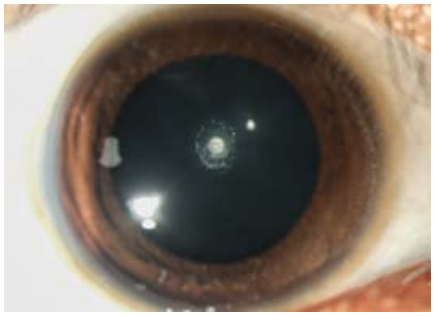


Fig. 8.17.6A: Mittendorf's dot

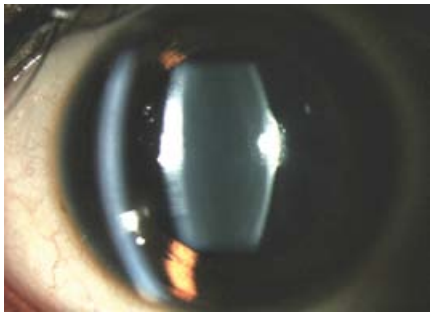


Fig. 8.17.6B: Mittendorf's dot in slit section

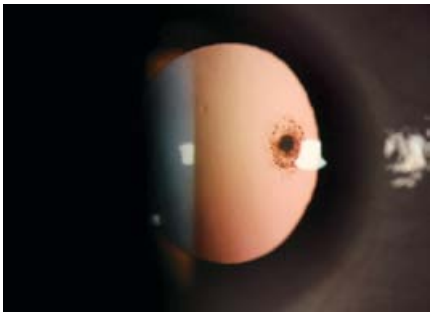


Fig. 8.17.6C: Mittendorf's dot in retroillumination

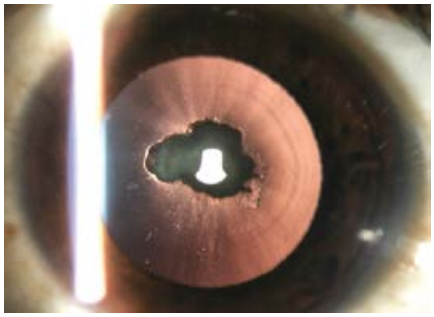


Fig. 8.17.7A: PPC with anterior type of PHPV

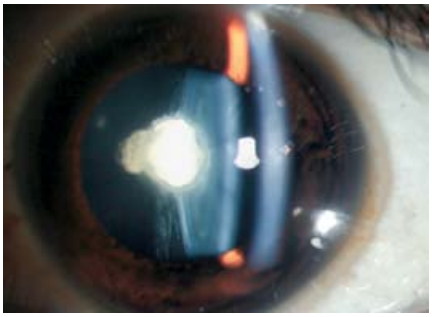


Fig. 8.17.7B: PPC with anterior type of PHPV

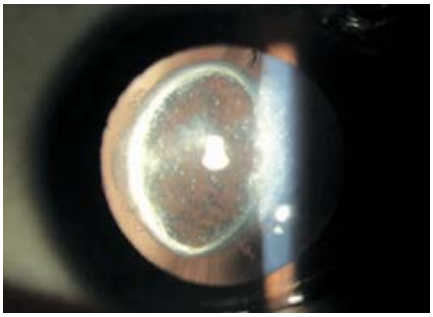


Fig. 8.17.7C: PPC with large gap and PHPV

Coronary Cataract

- Usually sporadic developmental cataract, occurring at puberty
- Appears as a 'corona' or club-shaped opacities, near the periphery of the lens cortex **(Figs 8.18.1A and B)**
- They are hidden by the iris, full dilatation of the pupil is essential for is diagnosis
- Does not interfere with vision
- May co-exist with other types of congenital or developmental lens opacity
- *Treatment:* The opacity remains stationary and treatment is not necessary in most of the cases.

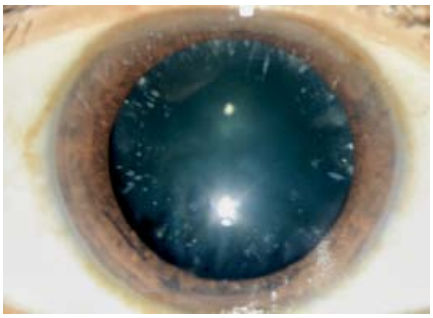


Fig. 8.18.1A: Coronary cataract

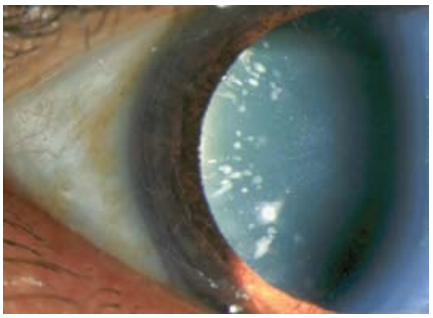


Fig. 8.18.1B: Coronary cataract near the equator

Zonular (Lamellar) Cataract

- Most common type of developmental cataract presenting with visual impairment
- Usually dominant, but may be recessive
- Consists of concentric, sharply demarcated zone (lamellae) of opacities surrounding a clear nuclear core, and enveloped by the clear cortex externally **(Figs 8.19.1 and 8.19.2)**
- Linear opacities, like spokes of a wheel (called *riders*) **(Fig. 8.19.3)** that extend outwards toward the equator—and it is pathognomonic.

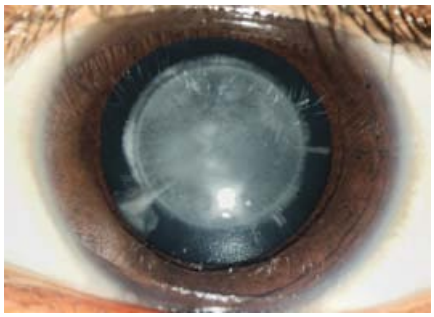


Fig. 8.19.1A: Zonular cataract

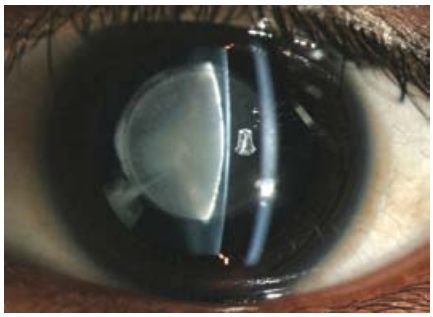


Fig. 8.19.1B: Zonular cataract

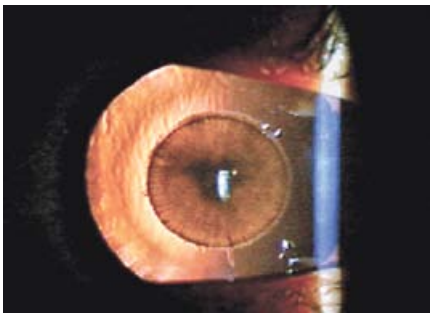


Fig. 8.19.2: Zonular cataract in retroillumination

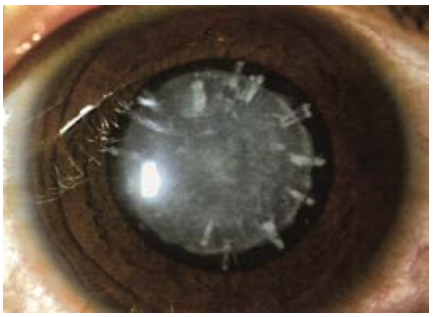


Fig. 8.19.3: Zonular cataract—riders

Central Pulverulent Cataract

- Rare, nonprogressive cataract with dominant inheritance
- Spheroidal powdery opacity, 2-4 mm in diameter within the lens nucleus, with a relatively clear center **(Figs 8.19.4A and B)**
- It should be differentiated from zonular cataract by absence of riders
- Not so important clinically.

Fig. 8.19.4A: Pulverulent cataract

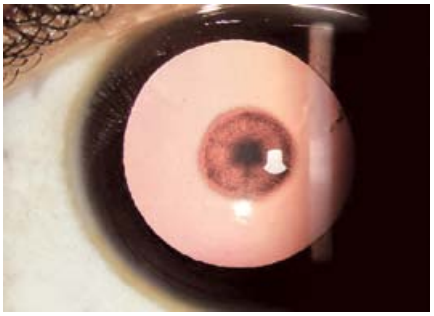


Fig. 8.19.4B: Pulverulent cataract in retroillumination

Rubella Cataract

- Cataract is originally nuclear and progresses to become total—pearly white in color **(Fig. 8.20.1)**
- Associated with microphthalmos **(Fig. 8.20.2)**, nystagmus, strabismus, glaucoma **(see Fig. 9.2.2)** and pigmentary retinopathy
- Maternal history is very important
- Removal of such cataract frequently provokes an intense uveitis or endophthalmitis.

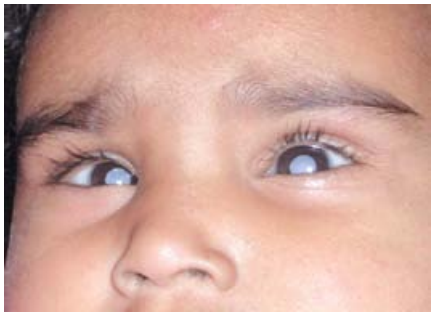


Fig. 8.20.1: Congenital cataract—rubella—pearly white

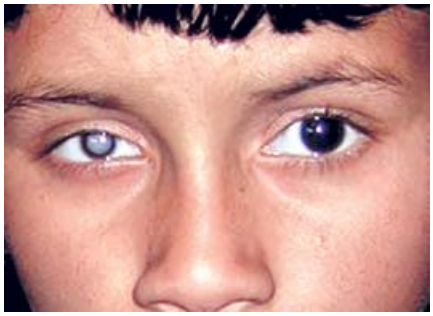


Fig. 8.20.2: Congenital cataract—rubella—microphthalmos

Sutural Cataract

- Rare, bilateral cataract, usually does not interfere with vision
- X-linked recessive inheritance, with males has significant opacities than females
- Opacities follow the course and shape of anteroposterior Y-sutures of the lens (**Figs 8.21.1A and B**)
- May be associate with other types of congenital cataract **(Figs 8.21.2 and 8.22.2)**
- Usually no treatment is necessary.

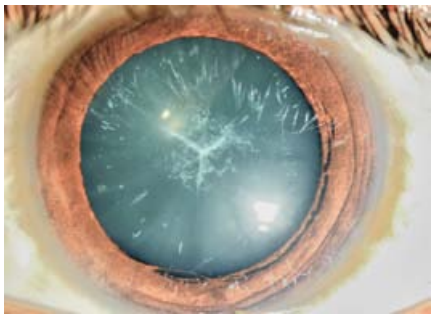


Fig. 8.21.1A: Sutural cataract

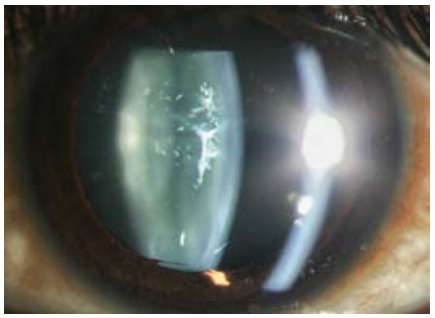


Fig. 8.21.1B: Sutural cataract

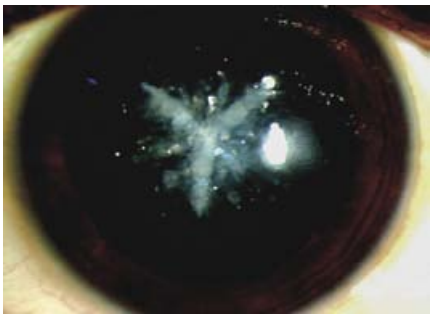


Fig. 8.21.2: Sutural cataract

Blue Dot Cataract

- Bilateral, rather common, and innocuous and does not cause visual problem **(Fig. 8.22.1)**
- Usual detection is during routine ophthalmological examination
- No treatment is necessary
- May co-exist with other type of congenital cataract **(Figs 8.22.2 and 8.22.3)** and then it may require treatment.

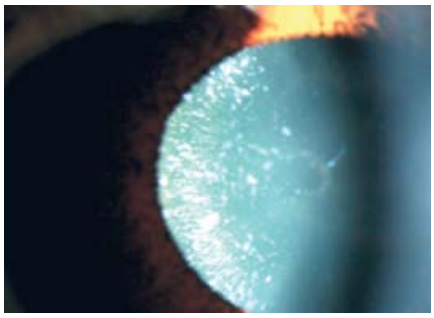


Fig. 8.22.1: Blue dot cataract

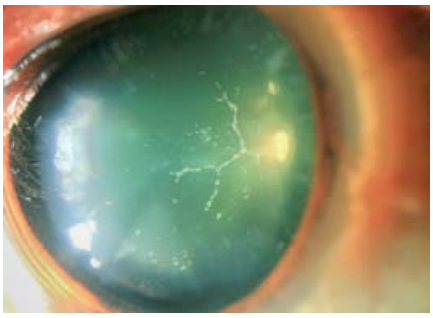


Fig. 8.22.2: Blue dot and sutural cataracts

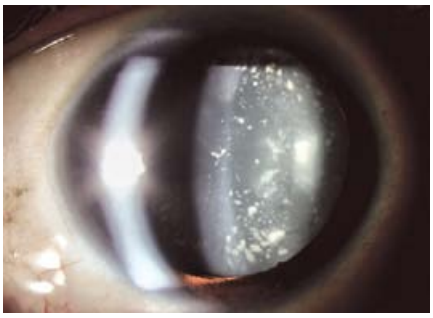


Fig. 8.22.3: Blue dot and coronary cataracts

Other Types of Congenital/ Developmental Cataract

- Coralliform cataract **(Figs 8.23.1A and B)**
- Floriform cataract **(Figs 8.23.2A to D)**

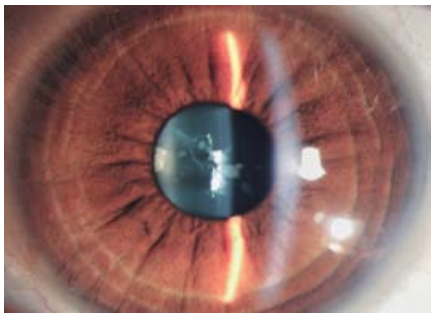


Fig. 8.23.1A: Coralliform cataract

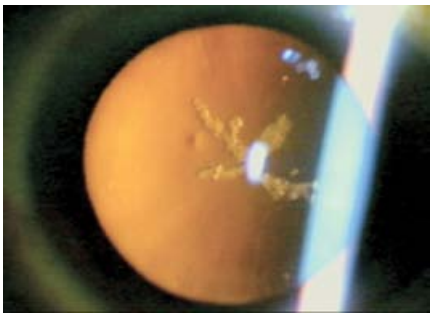


Fig. 8.23.1B: Coralliform cataract

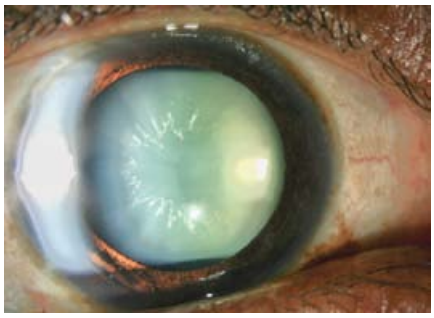


Fig. 8.23.2A: Floriform cataract—RE

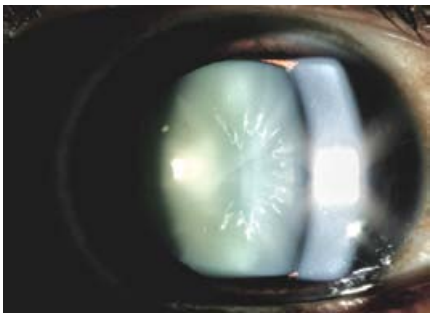


Fig. 8.23.2B: Floriform cataract—LE

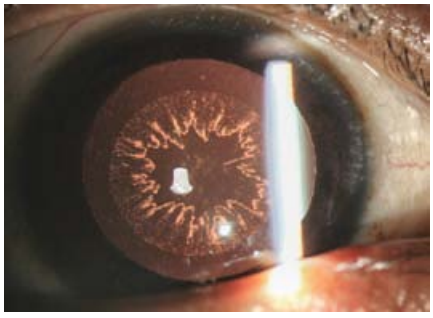


Fig. 8.23.2C: Floriform cataract—RE in retroillumination

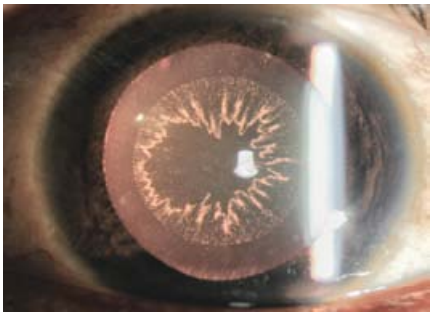


Fig. 8.23.2D: Floriform cataract—LE in retroillumination

- Spoke-like opacities **(Figs 8.23.3A and B)**
- Star-like opacities **(Fig. 8.23.4)**
- Epicapsular pigment stars **(Figs 8.23.5A and B)**.

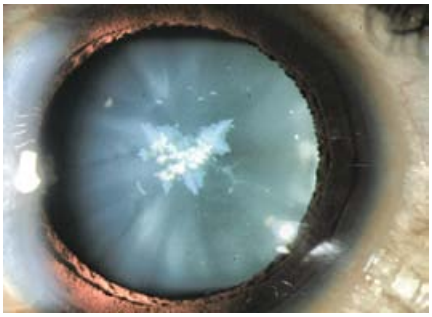


Fig. 8.23.3A: Spoke-like cataract

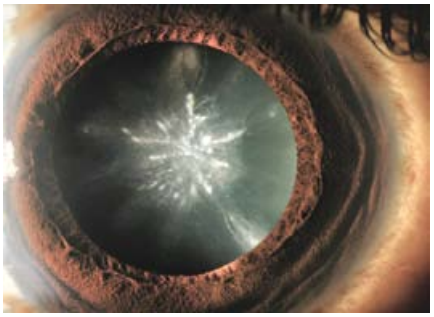


Fig. 8.23.3B: Spoke-like cataract

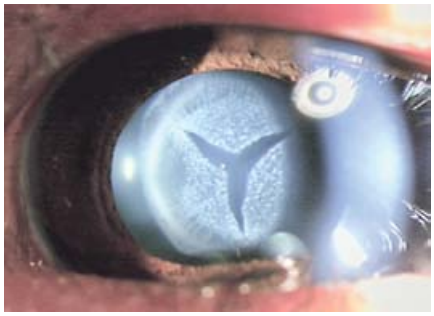


Fig. 8.23.4: Star-like cataract

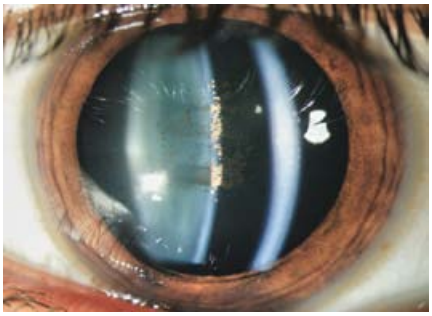


Fig. 8.23.5A: Epicapsular pigment stars

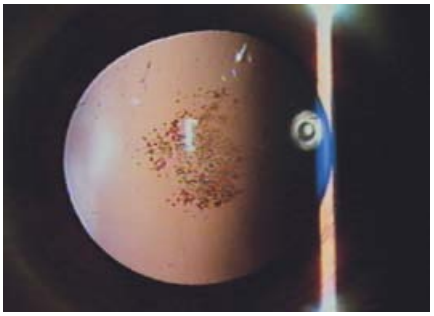


Fig. 8.23.5B: Epicapsular pigment stars

Senile (Adult) Cataract

- Cortical or soft cataract
- Nuclear or hard cataract
- Corticonuclear or mixed cataract.

Cortical Cataract

- *Cuneiform:* Nuclear or hard cataract and gradually encroaches towards center **(Figs 8.24.1 and 8.24.2)**
- *Cupuliform cataract or posterior subcapsular cataract:* Opacity appears in the posterior cortex just beneath the capsule and gradually it forms a dense opacity **(Figs 8.24.3A and B)**.

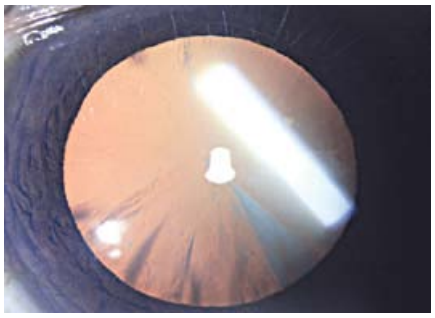


Fig. 8.24.1: Cortical cataract—early

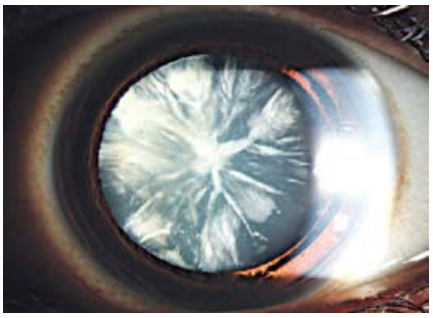


Fig. 8.24.2A: Cortical cataract—cuneiform cataract

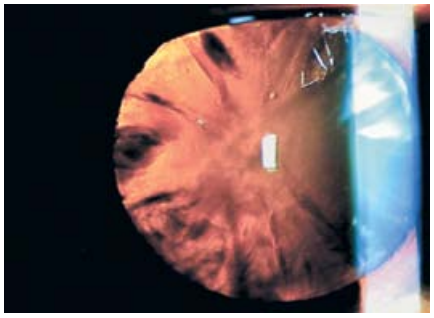


Fig. 8.24.2B: Cortical cataract—cuneiform cataract

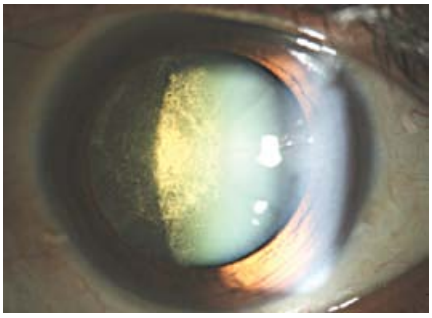


Fig. 8.24.3A: Cortical cataract—posterior subcapsular type

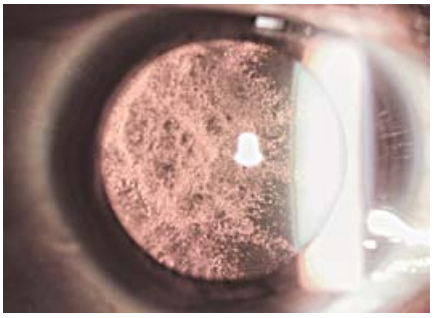


Fig. 8.24.3B: Cortical cataract—posterior subcapsular type

Stages of Cortical Cataract

Immature Stage

- Lens is grayish or grayish white in color
- Iris shadow is present
- May start as corticonuclear opacity **(Figs 8.25.1A and B)**.

Intumescent Stage

- Lens becomes swollen due to progressive hydration of the cortical fibers (**Fig. 8.25.2**)
- There is gradual change towards white color (**Fig. 8.25.3**)
- Anterior chamber becomes shallower (**Figs 8.25.4A and B**)
- Chance of secondary angle-closure glaucoma.

Mature Stage

- Lens is white or pearly white in color **(Fig. 8.25.5)**
- No iris shadow
- In many developing countries, patient may present with bilateral mature cataract **(see Figs 8.13.2 and 8.25.3)**.

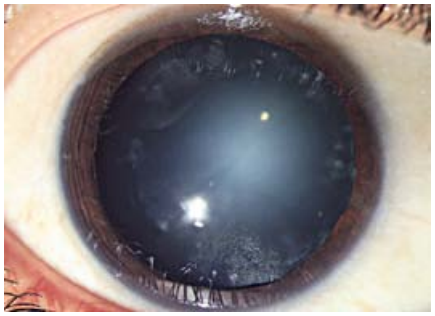


Fig. 8.25.1A: Immature cortical cataract with NS II

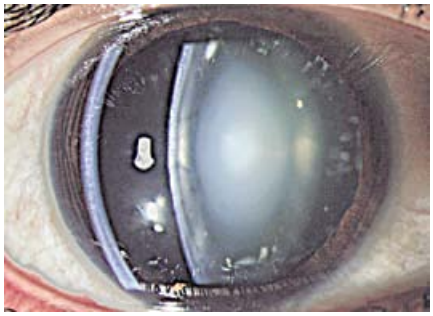


Fig. 8.25.1B: Immature cortical cataract with NS II

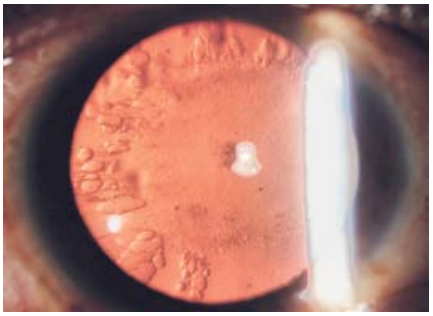


Fig. 8.25.2: Fluid vacuolation in the lens

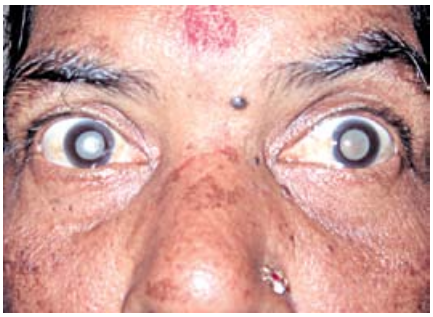


Fig. 8.25.3: Bilateral white mature cataract

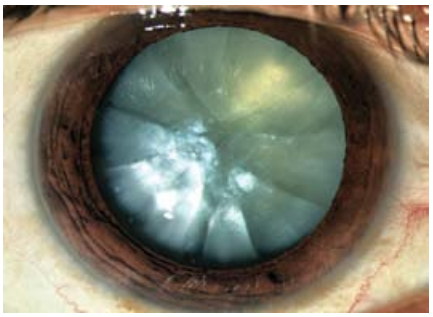


Fig. 8.25.4A: Intumescent cataract—fluid clefts

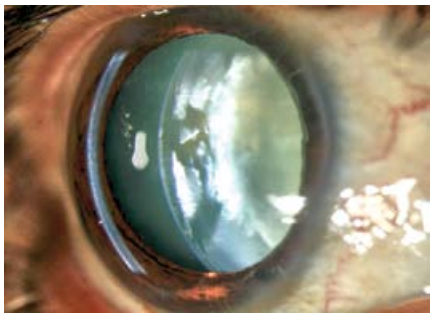


Fig. 8.25.4B: Intumescent cataract—fluid clefts

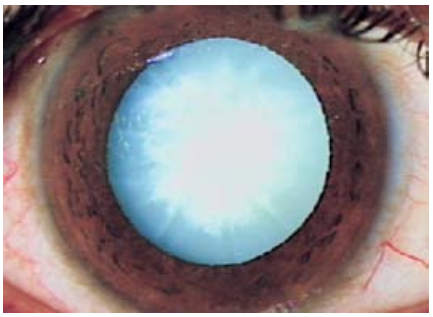


Fig. 8.25.5: Mature cataract

Hypermature Stage

- Cortex becomes disintegrated and then liquefied, or transformed into a pultaceous mass
- *Morgagnian cataract*
  - Cortex becomes fluid and the brown nucleus may sink at the bottom within the lens capsule **(Fig. 8.25.6A)**
  - Fluid is milky-white in appearance
  - A semicircular line above the nucleus, which may change its position **(Fig. 8.25.6B)**
  - Capsular deposits may occur in some cases **(Fig. 8.25.6C)**
- *Sclerotic cataract*
  - More and more inspissated, and shrunken in appearance, due to loss of fluid **(Figs 8.25.7A and B)**
  - Lens is more flat and yellowish-white in appearance
  - Calcific deposits in some part of the capsule.

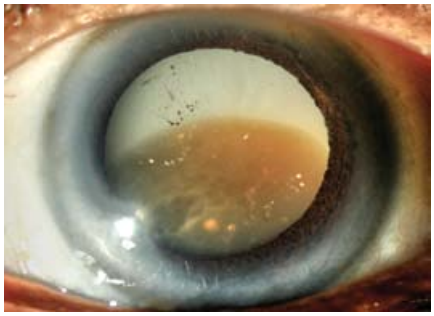


Fig. 8.25.6A: Hypermature cataract—Morgagnian

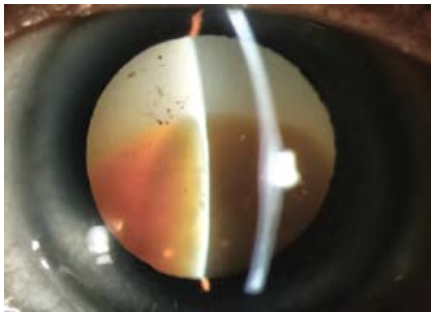


Fig. 8.25.6B: Hypermature cataract—Morgagnian

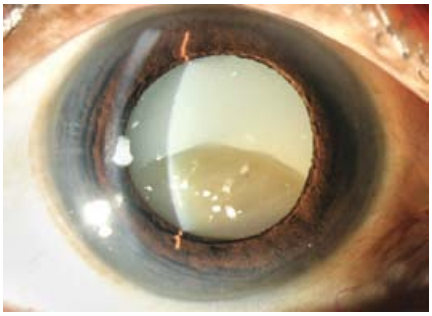


Fig. 8.25.6C: Hypermature cataract—Morgagnian—capsular deposits

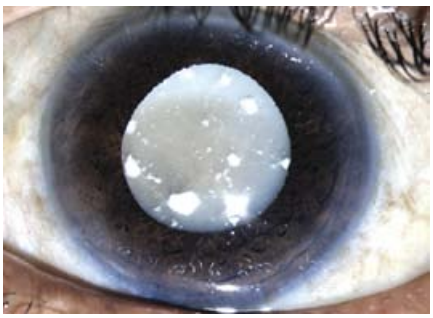


Fig. 8.25.7A: Hypermature cataract—sclerotic type

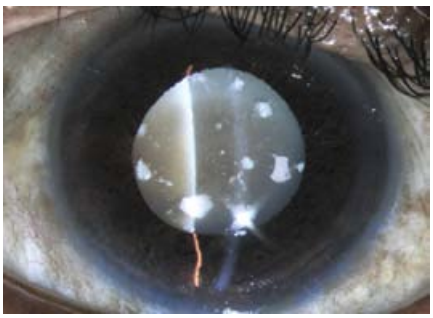


Fig. 8.25.7B: Hypermature sclerotic cataract—note deep AC

Cupuliform Cataract (Posterior Subcapsular Cataract)

- Starts in the axial region of the posterior cortex **(Figs 8.26.1A and B)**
- Slowly progresses to involve the entire posterior cortex **(Figs 8.26.2A and B)**
- Marked diminution of vision, as the opacity is near the nodal point of the eye
- This dimness of vision happens in bright sunlight of infront of bright headlight at night
- Patients with posterior cortical cataract always see better in dim illumination (dawn or dusk)
- Opacity is best judged by a slit-lamp, and with dilated pupil
- Appears as dirty yellowish-white layer in the posterior cortex and sometimes with formation of dense plaque **(Figs 8.26.3A and B)**
- Early cataract surgery is required inspite of better vision.

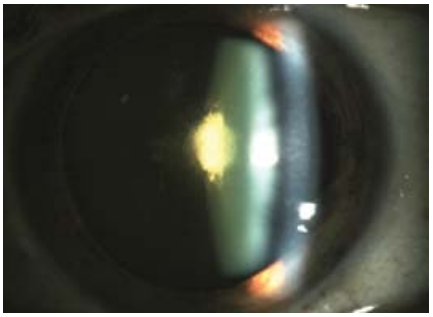


Fig. 8.26.1A: Early posterior subcapsular cataract

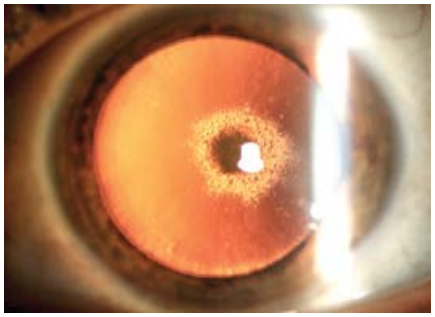


Fig. 8.26.1B: Early posterior subcapsular cataract

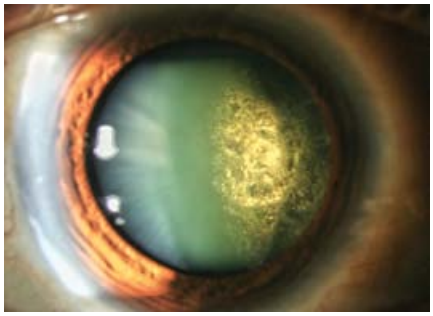


Fig. 8.26.2A: Posterior subcapsular cataract

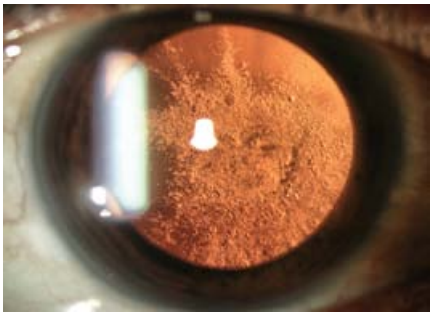


Fig. 8.26.2B: Posterior subcapsular cataract

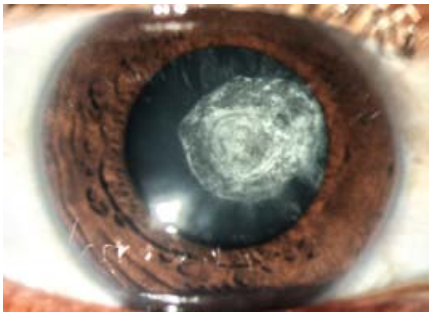


Fig. 8.26.3A: Dense posterior subcapsular cataract with plaque

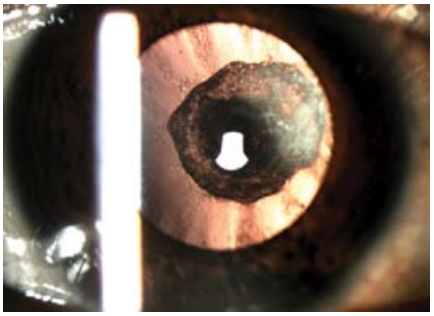


Fig. 8.26.3B: Dense posterior subcapsular cataract with plaque

Nuclear Cataract

- Tends to occur earlier than cortical variety
- More commonly seen in degenerative myopia, post-vitrectomy or in diabetes
- Varies in density and color which gradually spreads towards the cortex
- With time the lens becomes yellow **(NS II: Figs 8.27.1A and B)**, amber **(NS III: Figs 8.27.2A and B)**, brown *(cataracta brunescens)* **(NS IV: Figs 8.27.3 and NS V: Figs 8.27.4)**, or black *(cataracta nigra)* **(NS VI: Figs 8.27.5A and B)**
- May be associated with cortical type and called, corticonuclear cataract **(Figs 8.28.1 and 8.28.2)**
- Opacity is always better appreciated in dilated pupil with slit section and retroillumination.

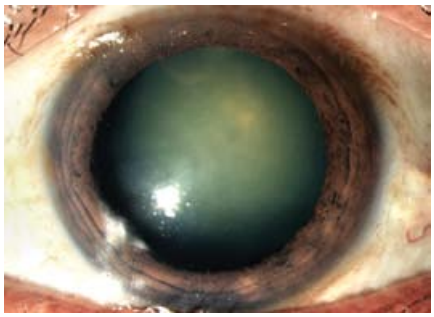


Fig. 8.27.1A: Nuclear cataract NS II

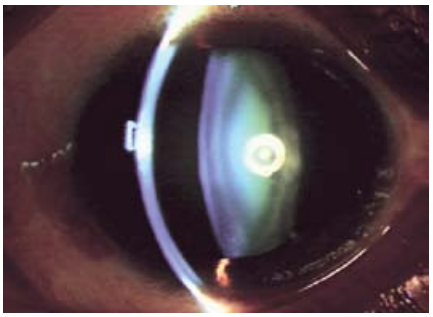


Fig. 8.27.1B: Nuclear cataract NS II and PSC

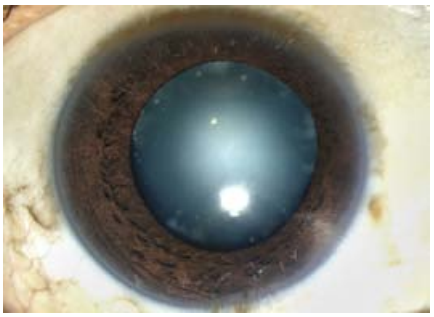


Fig. 8.27.2A: Nuclear cataract NS III

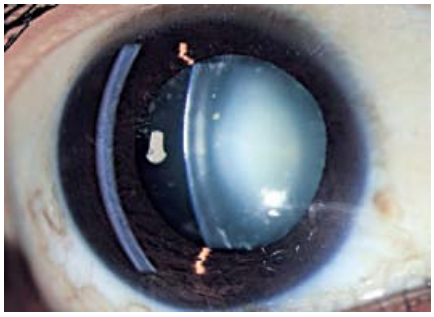


Fig. 8.27.2B: Nuclear cataract NS III

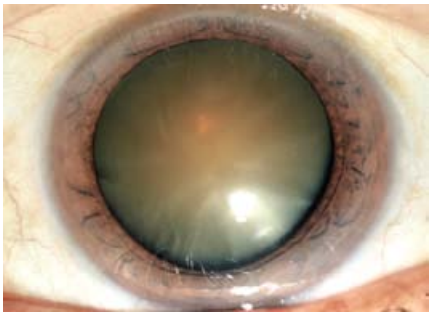


Fig. 8.27.3A: Nuclear cataract NS IV—brown cataract

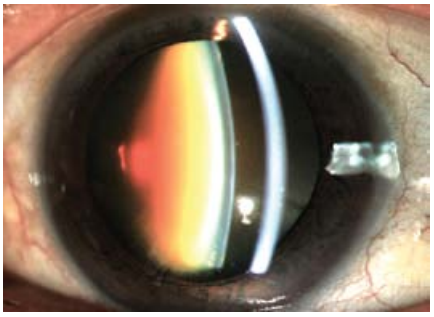


Fig. 8.27.3B: Nuclear cataract IV—brown cataract

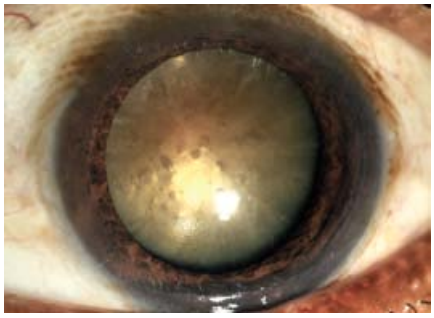


Fig. 8.27.4A: Nuclear cataract NS V—brown cataract

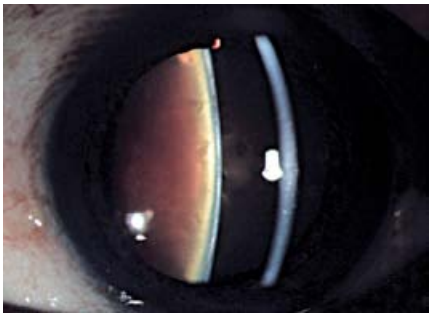


Fig. 8.27.4B: Nuclear cataract NS V—brown cataract

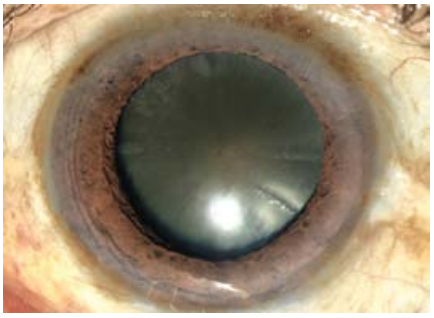


Fig. 8.27.5A: Nuclear cataract NS VI—black cataract

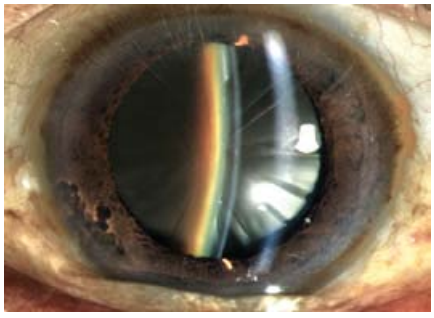


Fig. 8.27.5B: Nuclear cataract NS VI—black cataract

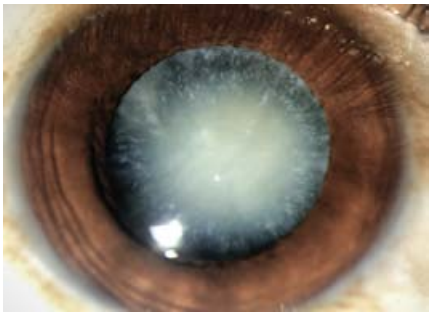


Fig. 8.28.1A: Corticonuclear cataract

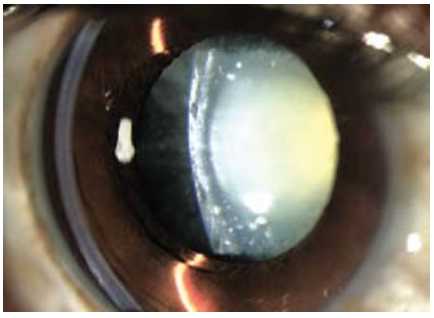


Fig. 8.28.1B: Corticonuclear cataract

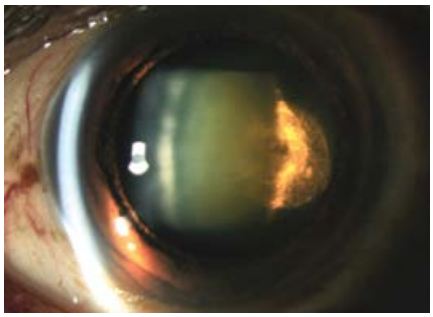


Fig. 8.28.2A: Corticonuclear cataract

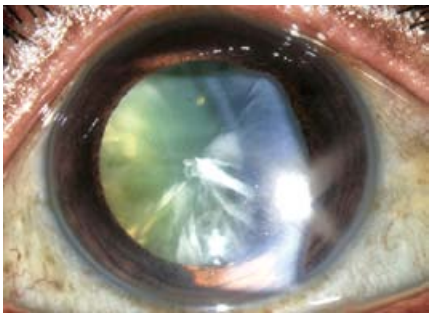


Fig. 8.28.2B: Corticonuclear cataract

Other Types of Adult Cataract

- *Christmas tree cataract:*
  - Uncommon age-related cataract
  - Polychromatic needle like opacities in the deeper cortex and nucleus **(Fig. 8.29.1)**
  - May be present with other opacities **(Fig. 8.29.2)**
  - It is present in myotonic dystrophy.
- *Anterior subcapsular deposits:*
  - Bilateral, fine granular opacity in the anterior cortex **(Figs 8.30.1A and B)**
  - Usually associated with certain drugs like, chlorpromazine
- *Steroid-induced cataract:*
  - Mainly occurs after prolonged use of topical or systemic steroids. The common problems are in VKC, after cornea grafting or after anterior uveitis or in renal transplant patients
  - The opacity first starts at the posterior subcapsular region **(Figs 8.30.2A and B)**, then it also deposits in the anterior capsule (**Figs 8.30.3A and B)** and eventually it may be total (**Figs 8.30.4A and B)**.

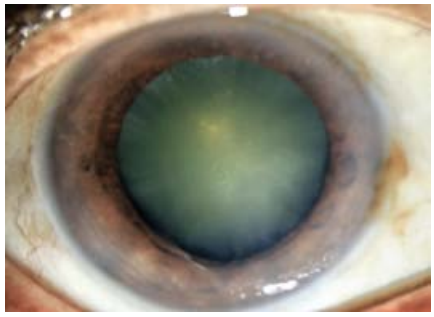


Fig. 8.29.1: Christmas tree cataract in myotonic dystrophy

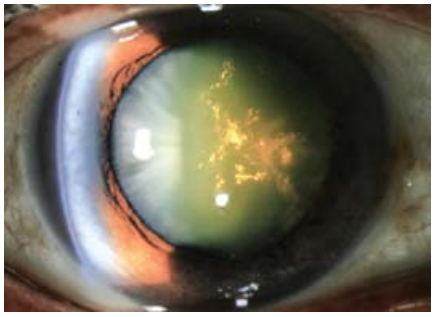


Fig. 8.29.2: Christmas tree cataract in myotonic dystrophy

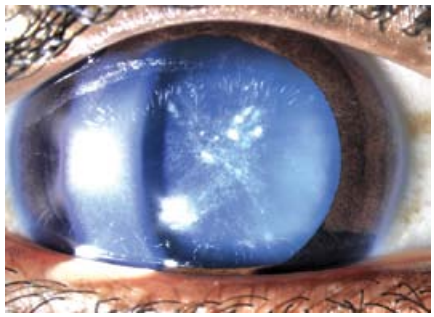


Fig. 8.30.1A: Drug-induced cataract—anterior subcapsular deposits

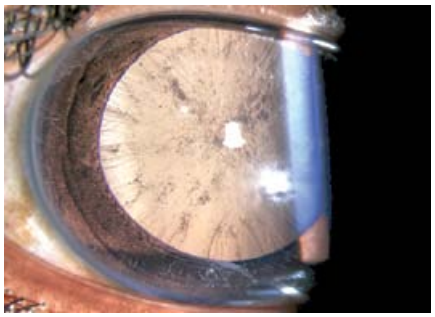


Fig. 8.30.1B: Drug-induced cataract—anterior subcapsular deposits

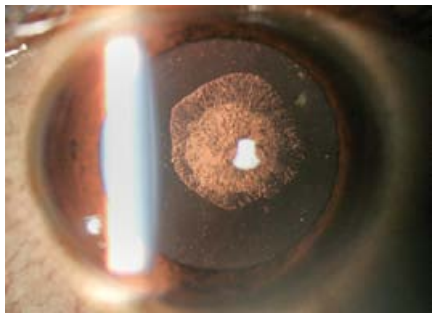


Fig. 8.30.2A: Steroidn-induced cataract in renal transplant patient

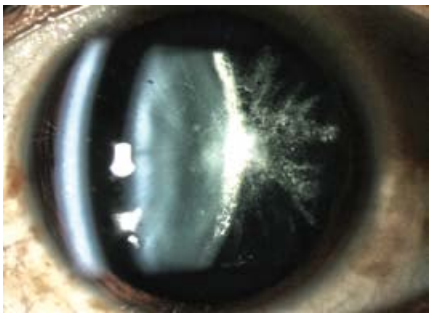


Fig. 8.30.2B: Steroid-induced cataract in renal transplant patient

Surgical Techniques in Senile Cataracts

- *Intracapsular cataract extraction (ICCE):* Obsolete nowadays
- *Extracapsular cataract extraction (ECCE)*
- *Manual (stitchless) small incision cataract surgery (MSICS):* Popular in most develop countries because of its cost-effectiveness
- *Phacoemulsification*: Preferred method with implantation of foldable intraocular lens (IOL) in-the-bag.

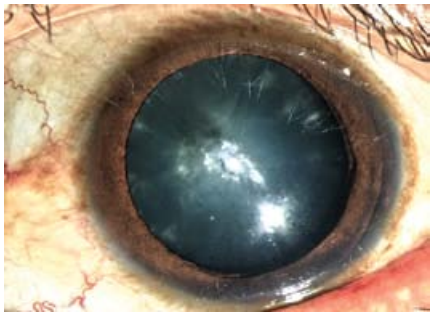


Fig. 8.30.3A: Steroid-induced cataract—both anterior and posterior

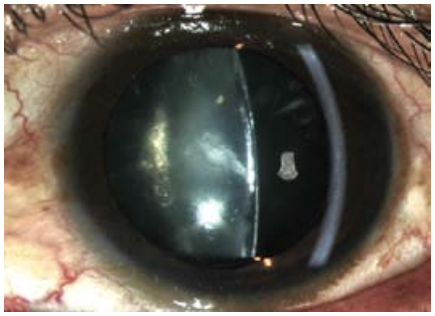


Fig. 8.30.3B: Steroid-induced cataract—both anterior and posterior

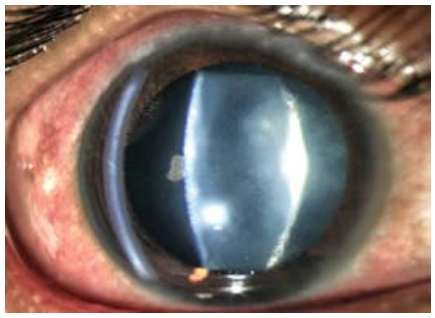


Fig. 8.30.4A: Steroid-induced cataract in VKC—note limbal lesions—RE

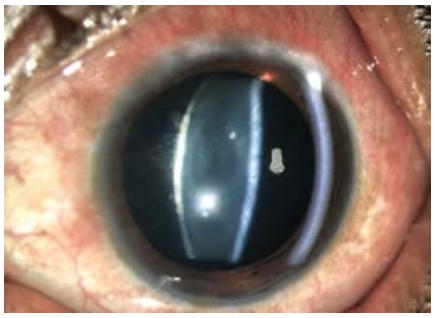


Fig. 8.30.4B: Steroid-induced cataract in VKC—note limbal lesions—LE

Specific Cataract Entities

Diabetic Cataract

- Early onset of nuclear cataract
- Posterior and anterior subcapsular opacities of varying degree
- *True diabetic cataract:*
  - More common in uncontrolled juvenile diabetics
  - Bilateral cortical cataract
  - Consists of minute white dots of varying size like 'snowflakes' **(Figs 8.31.1A and B)** and are usually called 'snow-storm cataract' **(Fig. 8.31.2)**
- *Treatment:* As in adult cataract, but associated retinopathy often reduces the visual outcome.

Fig. 8.31.1A: Diabetic cataract—snow-flakes

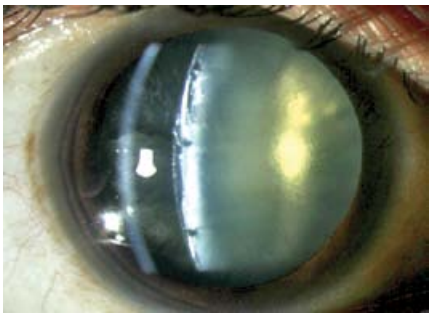


Fig. 8.31.1B: Diabetic cataract—snow-flakes

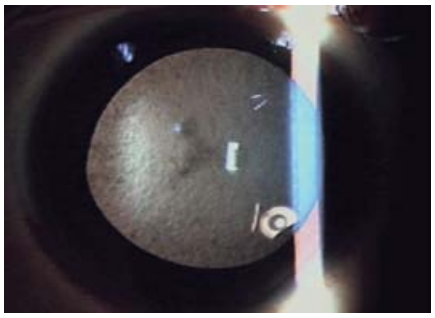


Fig. 8.31.2: Diabetic cataract—snow-strom cataract

Complicated Cataract

- Results from disturbances in lens metabolism in inflammatory or degenerative diseases
- *Causes:* Most common—iridocyclitis; the others are—degenerative myopia, retinitis pigmentosa, retinal detachment, post-vitrectomy cases, etc.
- Opacity usually commences in the axial region of posterior cortex *(posterior cortical cataract)* **(Figs 8.32.1A and B)**
- Appears as grayish-white opacity with irregular border extending towards the equator and in oblique illumination it gives breadcrumbs appearance **(Figs 8.32.2A to C)**
- Shows a characteristic rainbow display of colors, the polychromatic lustre **(Fig. 8.32.3)**
- Vision is much impaired as the opacity is near the nodal point of the eye
- Gradually the opacity may be total, as it happens quickly in cyclitis of Fuchs' **(Fig. 8.32.4)**
- It may be associated with BSK, especially in juvenile idiopathic arthritis **(Figs 8.32.5A and B)**

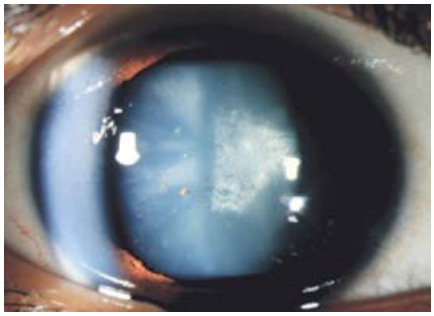


Fig. 8.32.1A: Complicated cataract in anterior uveitis—posterior

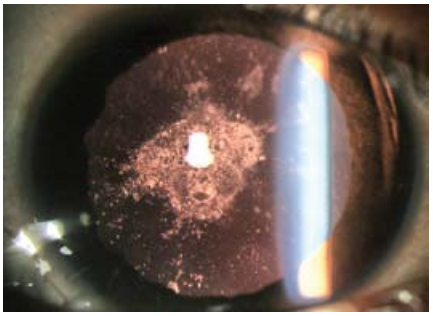


Fig. 8.32.1B: Complicated cataract in anterior uveitis—posterior

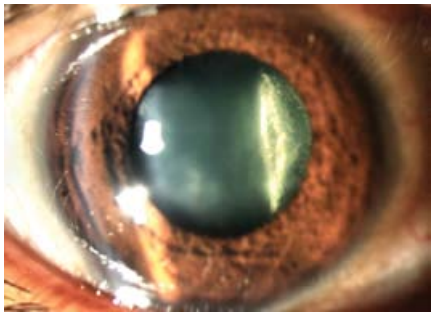


Fig. 8.32.2A: Breadcrumbs appearance

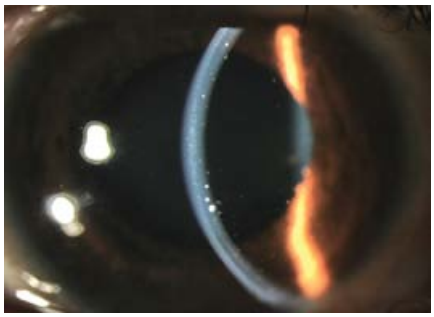


Fig. 8.32.2B: Breadcrumbs appearance—old KPs

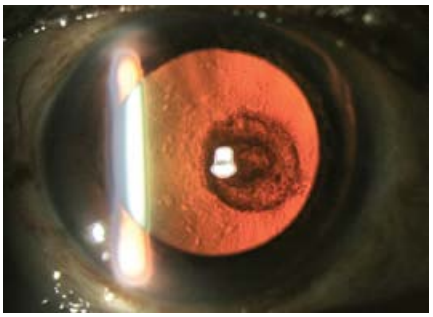


Fig. 8.32.2C: Breadcrumbs appearance

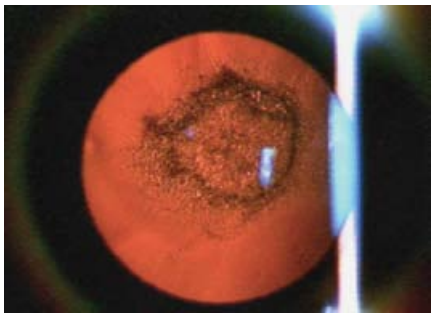


Fig. 8.32.3: Polychromatic lustre

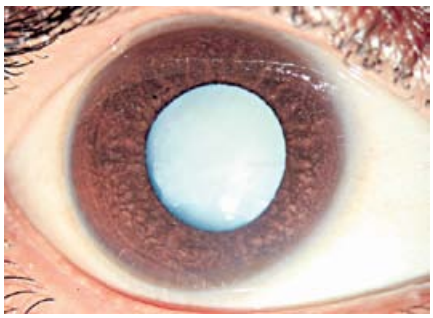


Fig. 8.32.4: Complicated cataract—in heterochromic cyclitis of Fuchs'

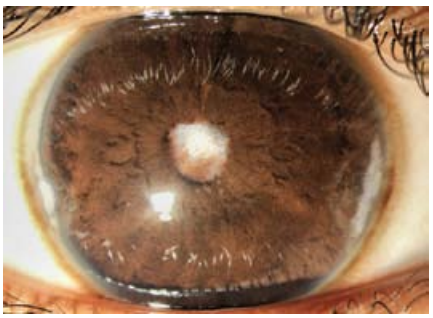


Fig. 8.32.5A: Complicated total cataract with posterior synechiae

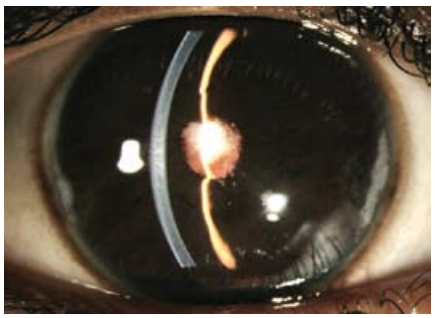


Fig. 8.32.5B: Complicated total cataract with posterior synechiae

- In post-vitrectomy cases there may be associated subcapsular dense plaque with posterior capsular weakness **(Figs 8.32.6A and B)**
- Complicated cataract may also happen after different types of chemical injuries **(Fig. 8.32.7)**
- *Treatment:* It is difficult in most of the cases except in cyclitis of Fuchs' where the prognosis is best.

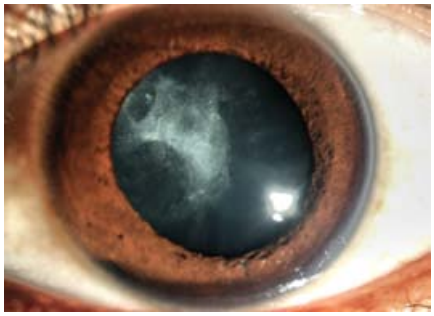


Fig. 8.32.6A: Complicated cataract—posterior cortical with dense plaque—post PPV

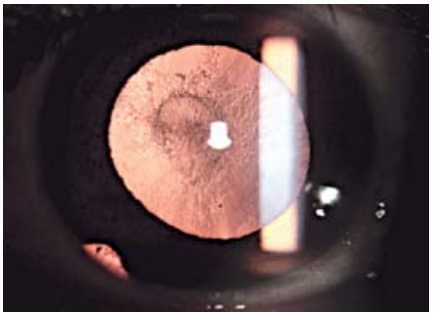


Fig. 8.32.6B: Complicated cataract—posterior cortical with dense plaque—post PPV

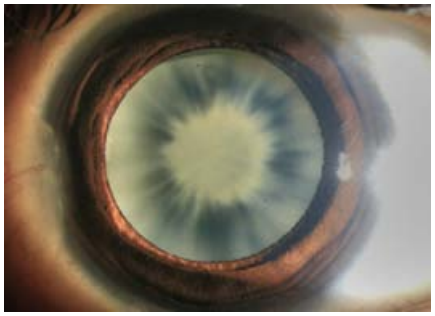


Fig. 8.32.7: Complicated cataract—chemical injury (*calotropis procera* latex)

Galactose Cataract

- Rare, bilateral, recessively inherited condition
- *Classical galactosemia:* About 75% of the sick infants with classical galactosemia develop bilateral 'oil-droplet' lental opacities which may progress to maturity within a few months
- *Galactokinase deficiency:* Milder type and only associated with cataract in healthy child.

Traumatic Cataract

- Due to concussion, penetrating or other type of injuries to the eyes
- *Heat cataract:*
  - Produced by prolonged exposure to infrared rays, and occurs in industry
  - Among glass blower's in glass factories *('glass blower's cataract')* **(Fig. 8.33.1A)**
  - Associated 'exfoliation' *(true exfoliation)* of the lens capsule
  - Lamella of the capsule may be curled up in the pupillary area as large sheets **(Fig. 8.33.1B)**
  - Must be differentiated from cataract with pseudoexfoliation **(Figs 8.33.2A and B)**.
- *Concussion cataract:*
  - After a blunt trauma
  - Initially early rosette **(Fig. 8.34.1)** then late rosette (**Fig. 8.34.2**) cataract and later on total cataract
  - Sometime seen as double rosette cataract (**Fig. 8.34.3**)
  - May be with subluxation (**Fig. 8.34.4**)
  - With partial absorbed (**Fig. 8.34.5**)

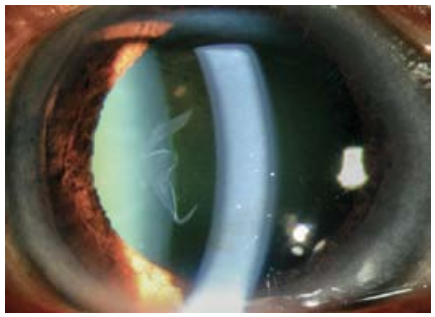


Fig. 8.33.1A: Heat cataract—glass blower's cataract

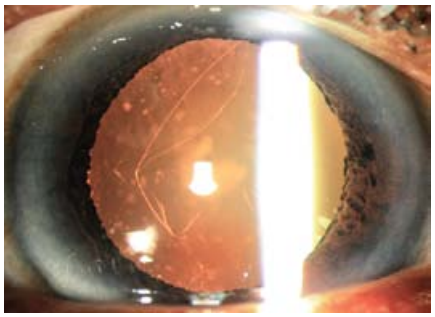


Fig. 8.33.1B: True exfoliation—glass blower's cataract

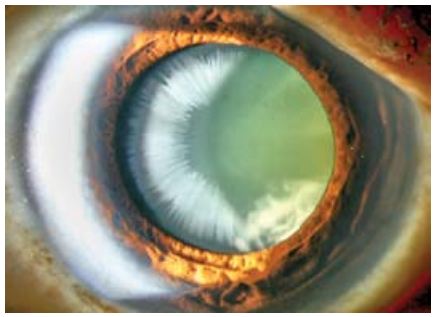


Fig. 8.33.2A: Pseudoexfoliation of the lens capsule

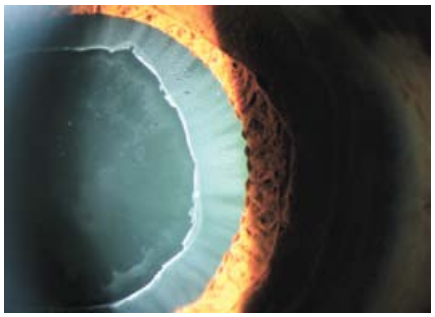


Fig. 8.33.2B: Pseudoexfoliation of lens capsule

- *Penetrating injury:*
  - After a penetrating injury
  - May be associated with capsular rupture **(Figs 8.34.6A and B)**
  - Fluffy white cortical matter in the anterior chamber **(Figs 8.34.7A and B)**
  - White cataract in most cases
  - With or without intraocular foreign body **(Figs 8.34.8A and B)**
  - May be associated with siderosis bulbi **(Figs 8.34.9A and B)**
  - Rarely trauma by the tube of Ahmed glaucoma valve (AGV) **(Figs 8.34.10A and B)**
- *Radiation cataract:* Associated with exposure to radiation therapy **(Figs 8.34.11A and B)**
- *Electric cataract:* Associated with high voltage current or hit by lightening **(Figs 8.34.12A and B)**.

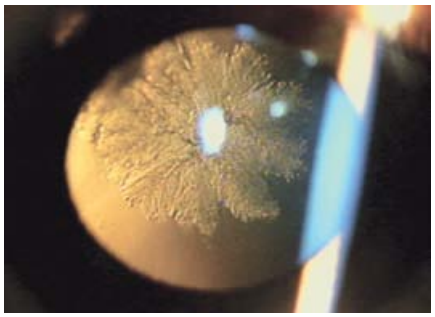


Fig. 8.34.1: Early rosette cataract

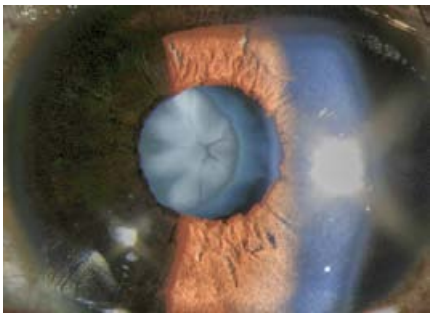


Fig. 8.34.2: Late rosette cataract

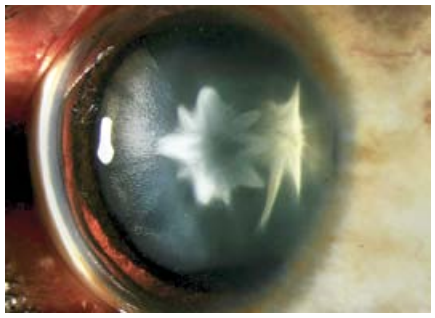


Fig. 8.34.3: Double rosette cataract

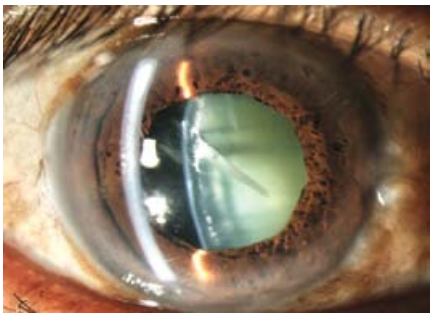


Fig. 8.34.4: Traumatic cataract—subluxation following blunt

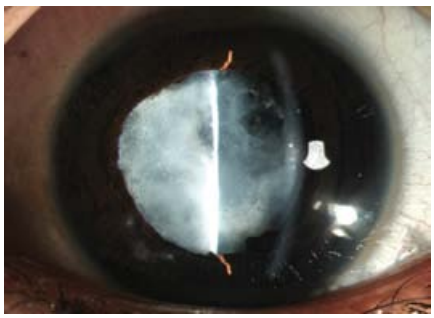


Fig. 8.34.5: Traumatic cataract—partially absorbed following

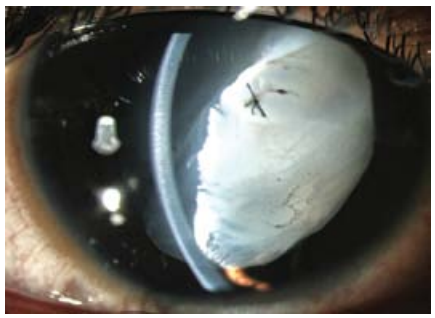


Fig. 8.34.6A: Traumatic cataract—subluxation following blunt

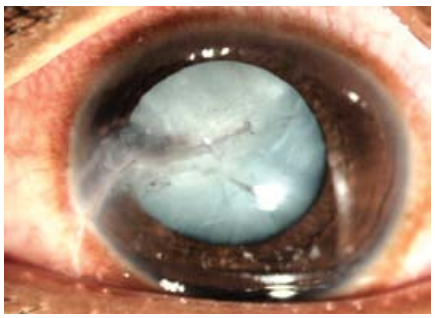


Fig. 8.34.6B: Traumatic cataract after penetrating injury

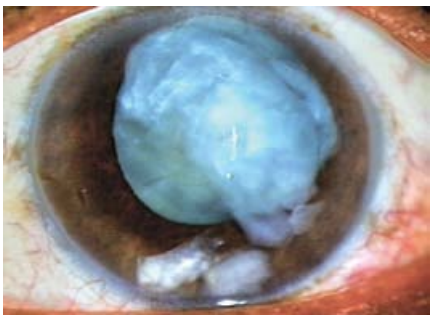


Fig. 8.34.7A: Traumatic cataract with fluffy cortex

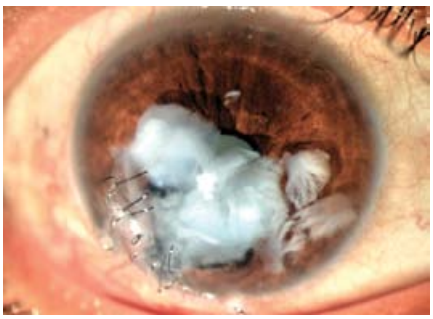


Fig. 8.34.7B: Traumatic cataract—fluffy cortex in AC following

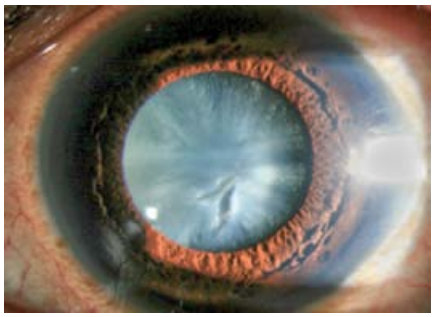


Fig. 8.34.8A: Traumatic cataract with lenticular FB

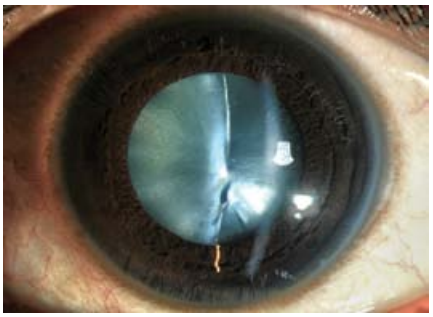


Fig. 8.34.8B: Traumatic cataract with lenticular FB

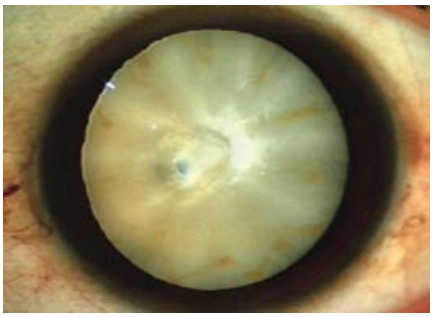


Fig. 8.34.9A: Traumatic cataract intralenticular iron FB with siderosis

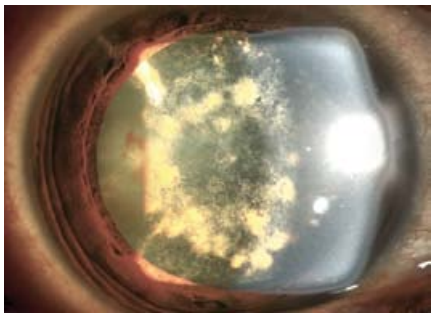


Fig. 8.34.9B: Traumatic cataract with siderosis

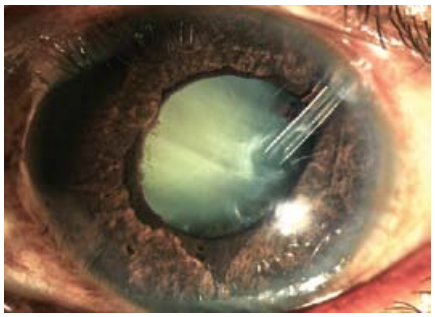


Fig. 8.34.10A: AGV-induced—end of tube touching lens

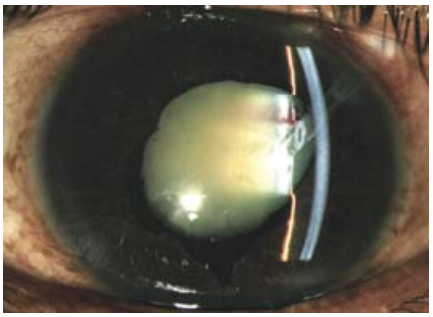


Fig. 8.34.10B: AGV-induced traumatic cataract

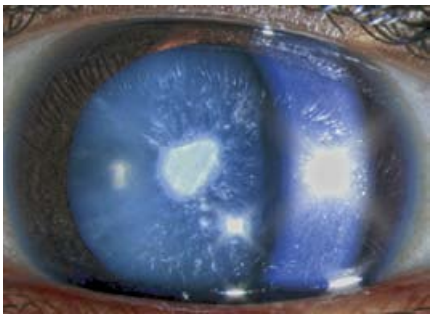


Fig. 8.34.11A: Radiation cataract—anterior capsular fibrosis

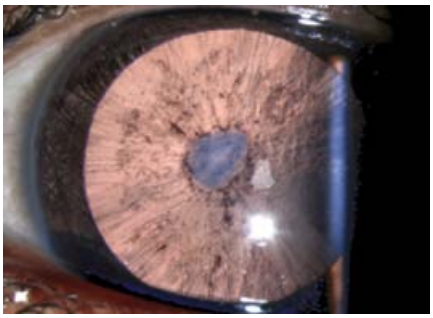


Fig. 8.34.11B: Radiation cataract—anterior capsular fibrosis

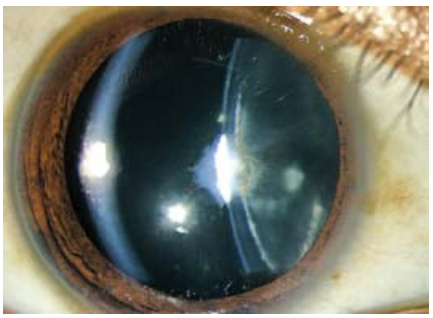


Fig. 8.34.12A: Electric cataract—anterior capsular fibrosis

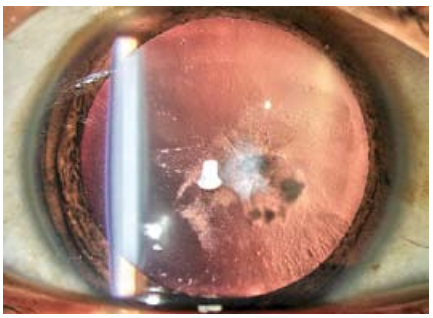


Fig. 8.34.12B: Electric cataract—anterior capsular fibrosis

Glaukomflecken

- Not so uncommon, usually unilateral condition
- Follows after an acute attack of angle closure glaucoma **(Fig. 8.35.1)**
- Grayish-white opacity in the pupillary zone which resembles 'spilled milk' **(Fig. 8.35.2)**
- May be associated with iris-sphincter atrophy.

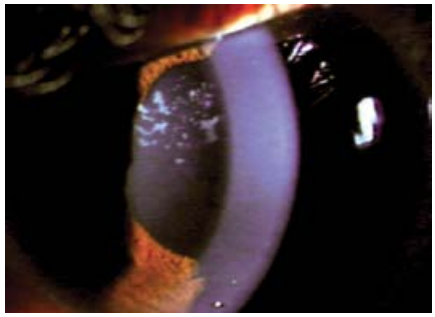


Fig. 8.35.1: Glaukamflecken following acute attack of PACG

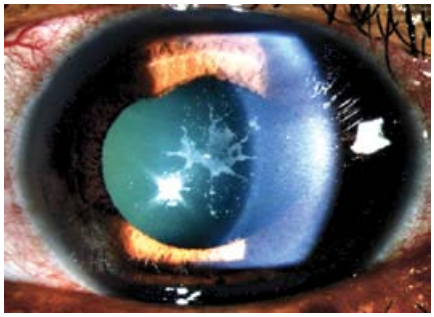


Fig. 8.35.2: Glaukamflecken—spilled milk appearance

Opacities in Aphakia or Pseudophakia

Anterior Capsular Opacification/Fibrosis

- Usually appears within 3 to 6 months after the surgery **(Figs 8.36.1A and B)** and gradually increases
- Fibrosis may be associated with anterior capsular phimosis **(Figs 8.36.2 and 8.36.3)**
- *Capsular phimosis:* In some cases, it gradually increases **(Figs 8.36.4 and 8.36.5)** and ultimately the capsulorhexis opening may close completely **(Figs 8.36.6A and B)**
- It may be associated with decentration of the IOL **(Fig. 8.36.7)** and especially it happens with silicone IOL **(Figs 8.36.8A and B)**

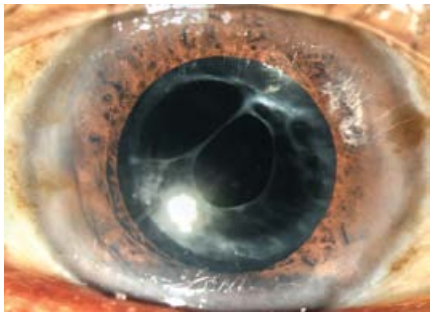


Fig. 8.36.1A: Anterior capsular opacification

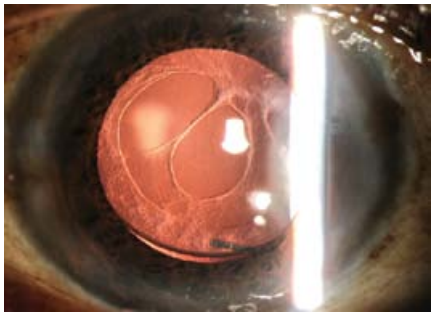


Fig. 8.36.1B: Anterior capsular opacification

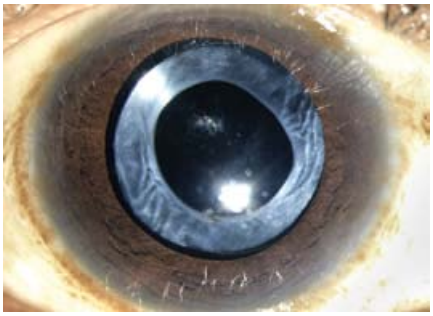


Fig. 8.36.2: Anterior capsular opacification (ACO)

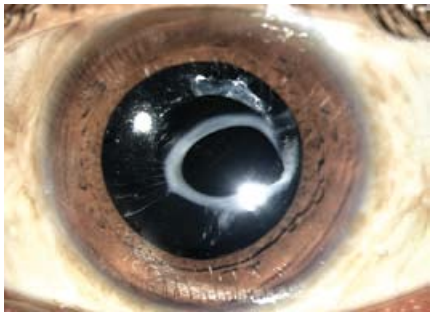


Fig. 8.36.3A: Anterior capsular phimosis

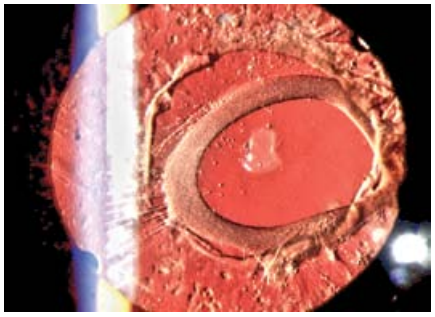


Fig. 8.36.3B: ACO with capsular phimosis

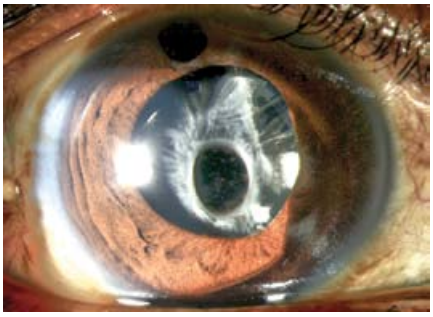


Fig. 8.36.4: ACO with more capsular phimosis

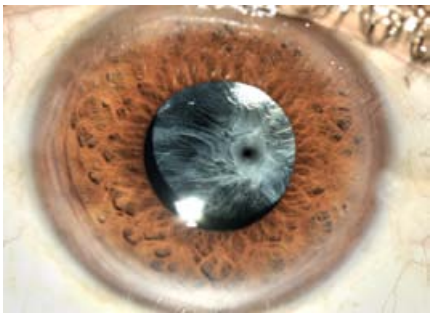


Fig. 8.36.5: Anterior capsular phimosis—near closure

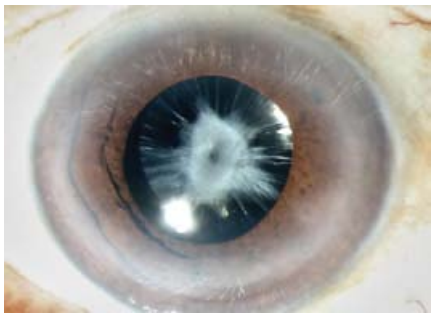


Fig. 8.36.6A: Anterior capsular phimosis—complete closure

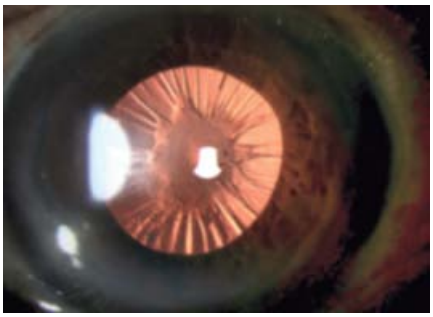


Fig. 8.36.6B: Anterior capsular phimosis—complete closure

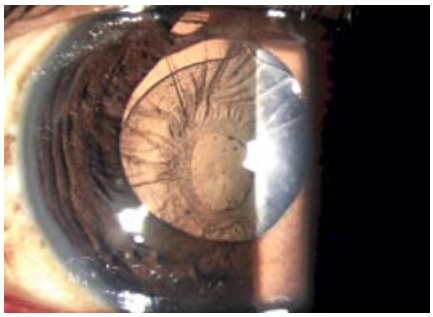


Fig. 8.36.7: ACO with phimosis with decentration of IOL

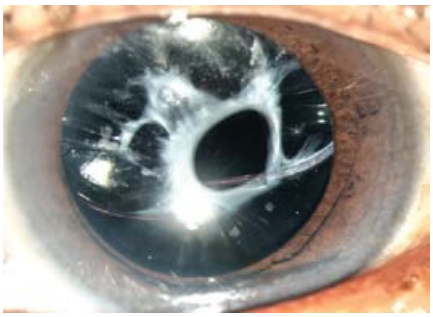


Fig. 8.36.8A: Anterior capsular phimosis—decentration of IOL

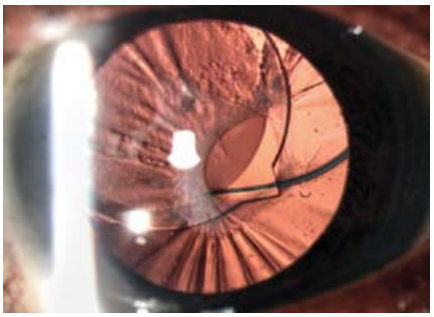


Fig. 8.36.8B: Capsular phimosis—decentration of silicone IOL

- Capsulorhexis may be small or presence of signs of iridocyclitis
- May be associated with posterior capsular opacification
- *Treatment:* In extreme cases YAG laser anterior capsulotomy may be required **(Fig. 8.36.9)**.

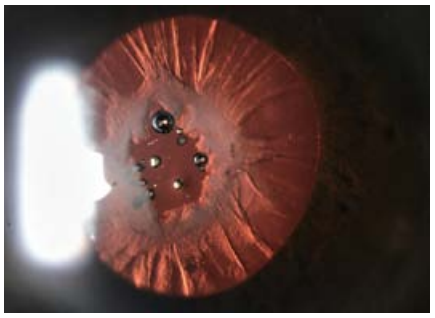


Fig. 8.36.9: After YAG anterior capsulotomy—same eye **(Fig. 8.36.6B)**

Posterior Capsular Opacification (PCO) or 'After Cataract'

- In adults, between 1 to 65% cases depending upon the type, IOL and quality of the cataract surgery
- In pediatric cataract, may be up to 100% cases
- Membranous white irregular opacity (fibrotic type) with or without Elschnig's pearls formation
- Significantly reduces the vision
- After extracapsular cataract extraction
- *Posterior capsular fibrosis:*
  - Membranous, white capsular fibrosis formed by the remnants of anterior and posterior capsules of the lens **(Figs 8.37.1 and 8.37.2)**
  - Causes less disturbances in vision
- *Elschnig's pearls:*
  - Subcapsular cubical cells proliferate and instead of forming lens fibers, they develop into large balloon-like cells which fill the papillary aperture **(Figs 8.37.3 to 8.37.6)**
  - This balloon lens-cell looks like pearl, and is known as Elschnig's pearls **(Fig. 8.37.7)**

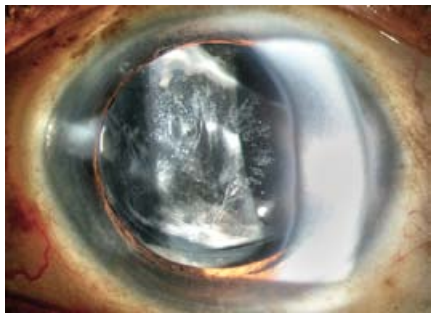


Fig. 8.37.1A: Early fibrous PCO

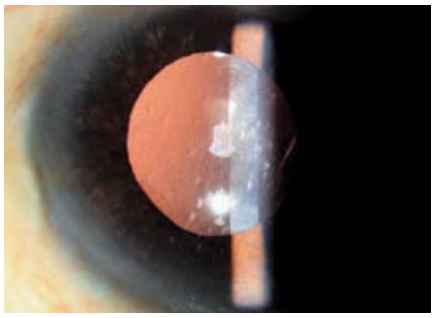


Fig. 8.37.1B: Early fibrous PCO

- More common in younger patients
- Appear after several months or years
- Causes more disturbances in vision

- *Late capsular bag distention syndrome:* It is more common with hydrophilic acrylic IOLs. Usually happens 4-5 years after the surgery. The distended capsular bag contains milky turbid fluid **(Figs 8.37.8A and B)** which is responsible for dimness of vision
- Treatment:
  - *YAG-laser capsulotomy* **(Fig. 8.37.9)** for all three conditions
  - In case of children and young adults, even after YAG-laser capsulotomy the Elschnig's pearls continue to grow and appear as a *pearl necklace* around the capsulotomy opening **(Figs 8.37.10A to D)** and may continue to grow and eventually close the capsulotomy opening **(Figs 8.37.11A and B)**.

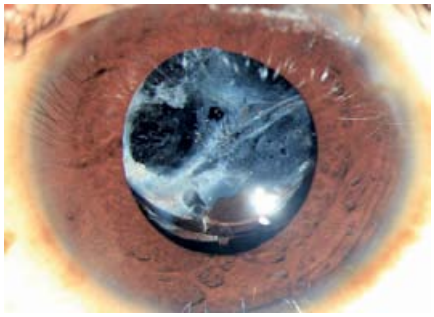


Fig. 8.37.2A: Dense fibrous PCO—difficult for YAG capsulotomy

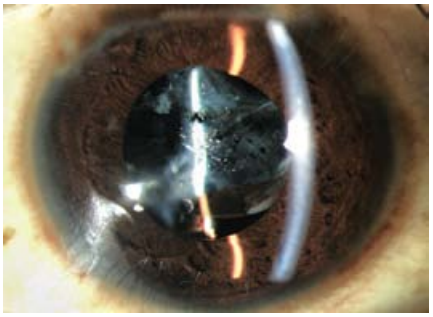


Fig. 8.37.2B: Dense fibrous PCO—difficult for YAG capsulotomy

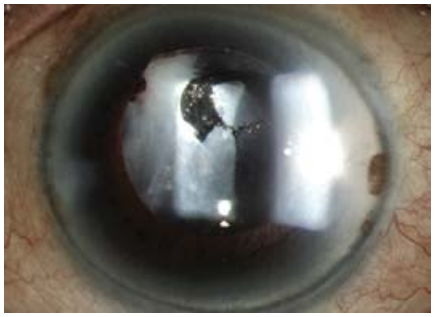


Fig. 8.37.2C: Fibrous PCO—after YAG capsulotomy

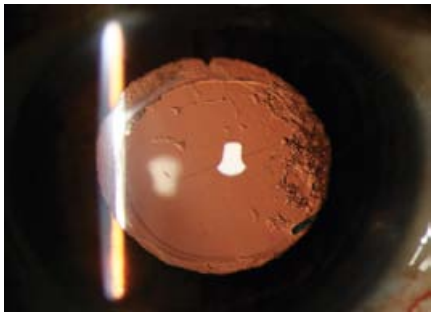


Fig. 8.37.3A: Mild PCO—Elschnig's pearls visual axis clear

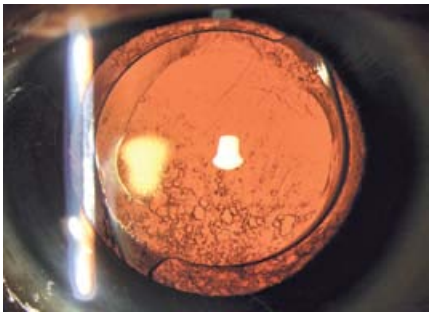


Fig. 8.37.3B: Mild PCO—Elschnig's pearls

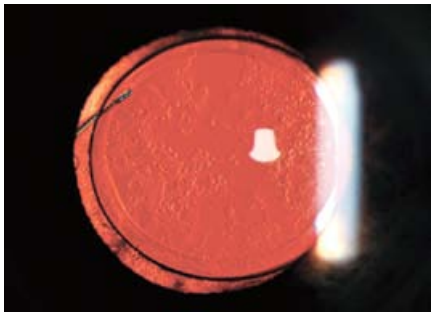


Fig. 8.37.4: Moderate PCO—Elschnig's pearls

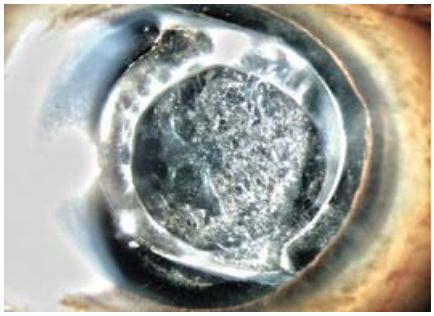


Fig. 8.37.5A: Severe PCO—Elschnig's pearls

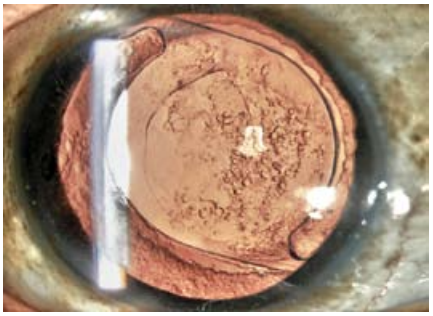


Fig. 8.37.5B: Severe PCO—Elschnig's pearls

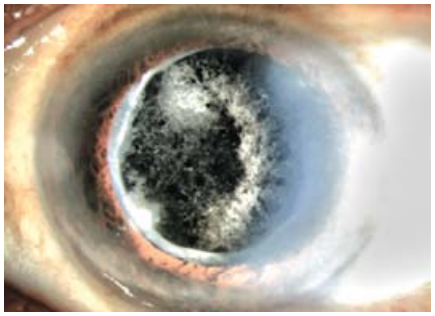


Fig. 8.37.6A: Extensive PCO—Elschnig's pearls

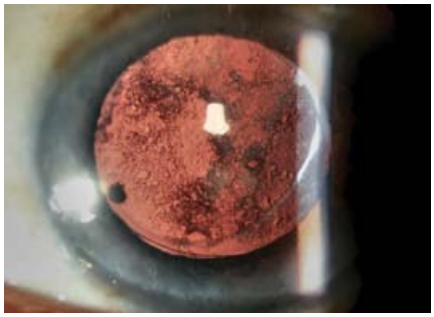


Fig. 8.37.6B: Extensive PCO—Elschnig's pearls

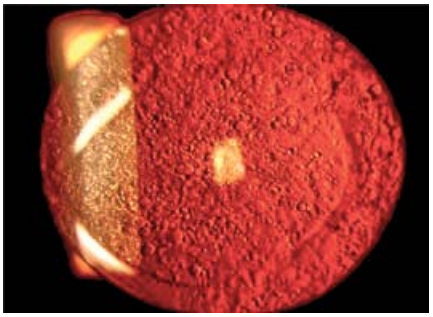


Fig. 8.37.6C: Extensive PCO hydrophilic foldable IOL

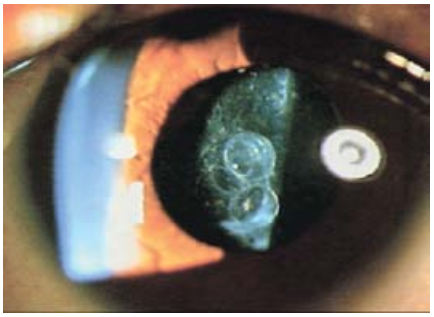


Fig. 8.37.7: PCO—large Elschnig's pearls

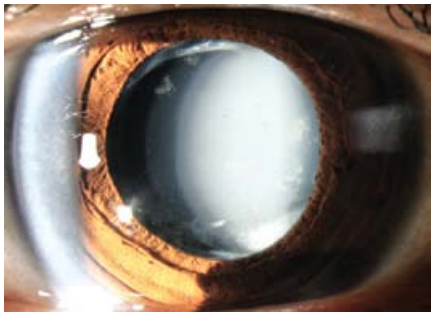


Fig. 8.37.8A: Late posterior capsular distention syndrome (PCDS)

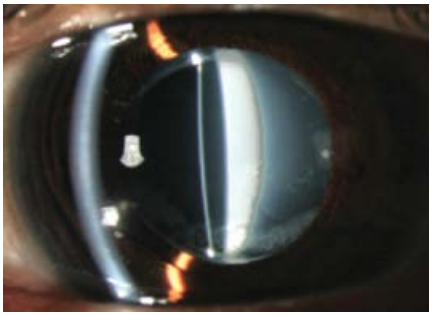


Fig. 8.37.8B: Late PCDS with milky turbid fluid within the bag

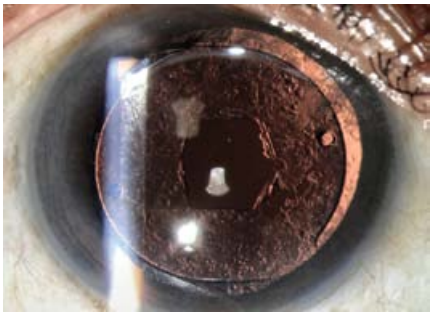


Fig. 8.37.9: YAG capsulotomy in PCO

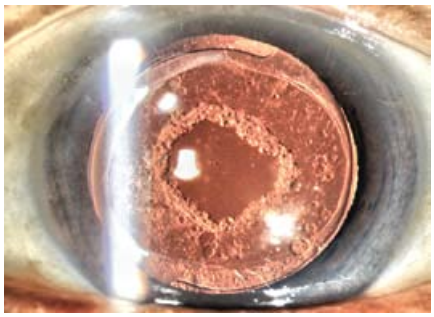


Fig. 8.37.10A: YAG capsulotomy—appearance of Elschnig's pearls

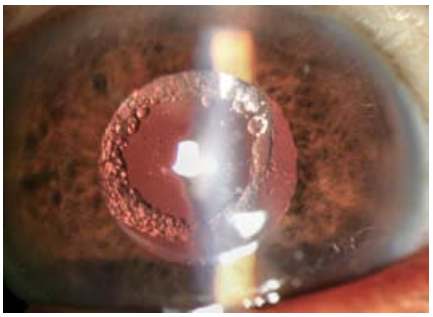


Fig. 8.37.10B: Pearl necklace after YAG capsulotomy

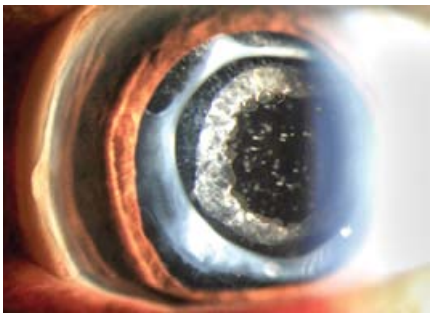


Fig. 8.37.10C: Pearl necklace after YAG capsulotomy

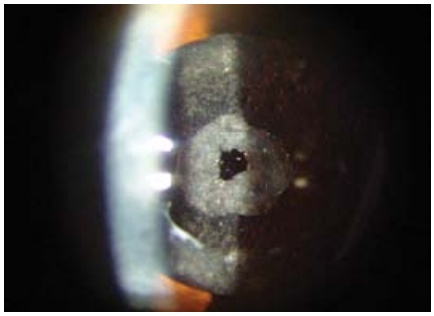


Fig. 8.37.10D: Pearl necklace after YAG capsulotomy near closure

Fig. 8.37.11A: PCO—closed YAG capsulotomy opening

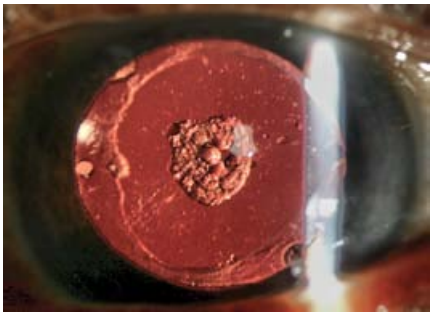


Fig. 8.37.11B: PCO—closed YAG capsulotomy opening

Soemmering's Ring

- Doughnut-like white ring behind the iris and seen mostly in pediatric cases **(Fig. 8.38.1)** and also in old needling operation cases **(Figs 8.38.2A and B)**
- Formed by the lens fibers enclosed between the two layers of lens capsule at the equatorial region
- Found in some cases where the peripheral cortex not cleaned properly **(Fig. 8.38.3)**
- This ring may dislocate in the pupillary area causing sudden visual loss **(Fig. 8.38.4)**
- *Treatment:* In case of pseudophakia, careful cleaning of the equatorial area. In aphakic cases—pars plana lensectomy with vitrectomy is the choice.

Fig. 8.38.1: Soemmering's ring BE, after PC IOL implantation

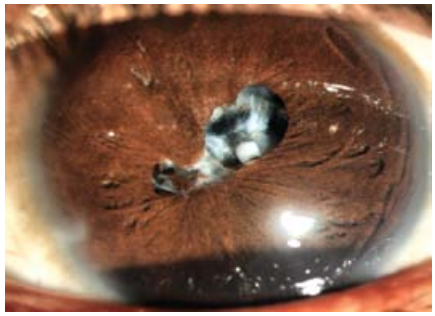


Fig. 8.38.2A: PCO and Soemmering's ring after needling operation

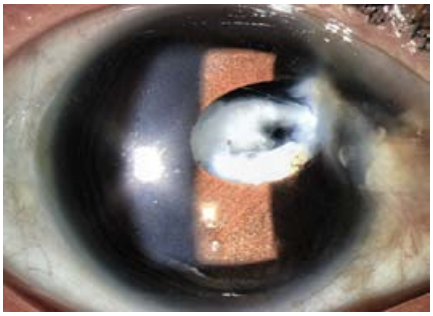


Fig. 8.38.2B: Soemmering's ring—dislocation into AC

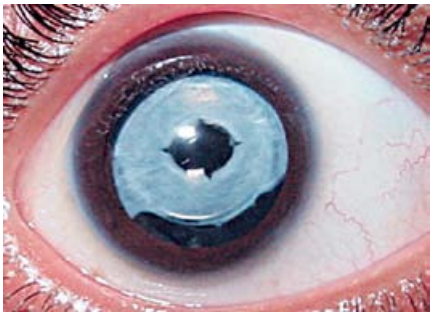


Fig. 8.38.3: Soemmering's ring—inadequate cortical clearing

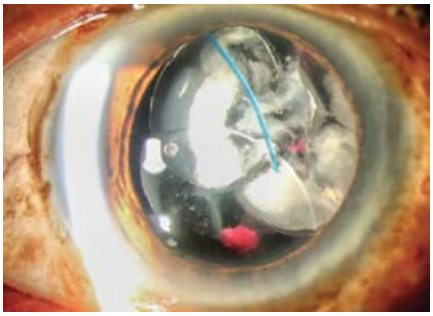


Fig. 8.38.4: Dislodgement of Soemmering's ring and IOL decentration

Cortical Matter Behind the IOL

- Associated with improper cleaning of posterior sheet of cortex during surgery **(Fig. 8.39.1)**—as it happens in case of posterior capsular rent or in small pupil cases
- Associated with uveal inflammatory reaction **(Fig. 8.39.2)**
- Presence of an AC IOL is not uncommon **(Fig. 8.39.3)**
- *Treatment*: To be removed surgically as early as possible with or without anterior vitrectomy.

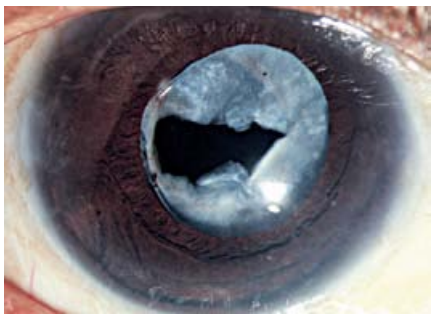


Fig. 8.39.1: Cortical matter behind PC IOL

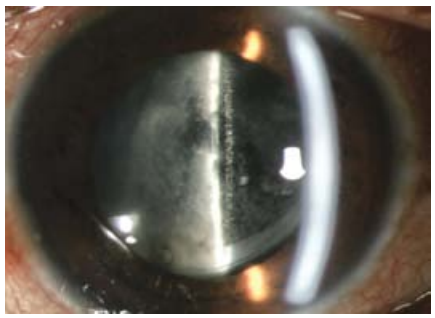


Fig. 8.39.2: Cortical matter behind IOL with inflammatory reaction

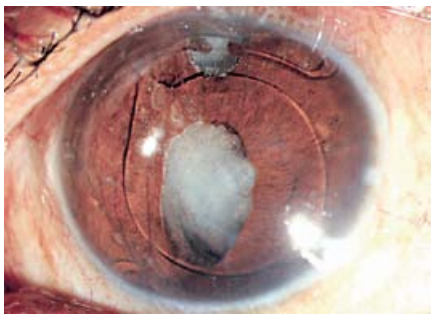


Fig. 8.39.3: Cortical matter behind AC IOL

Capsular Inflammatory Deposits/Plaques

- Associated with chronic iridocyclitis in some cases of aphakia **(Fig. 8.40.1)** or pseudophakia **(Fig. 8.40.2)**
- Localized fluffy white thickening of the capsule which are often pigmented **(Fig. 8.40.3)** and sometimes in association with dense PCO **(Fig. 8.40.4)**
- *Treatment:* YAG laser capsulotomy steroids or nonsteroidal anti-inflammatory drops.

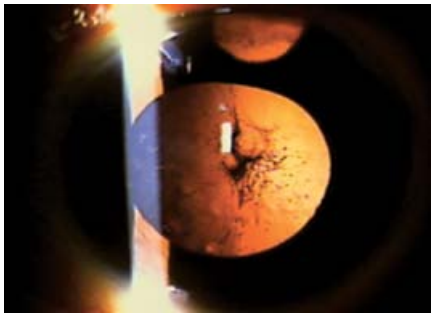


Fig. 8.40.1: Pigmentary PCO in aphakia

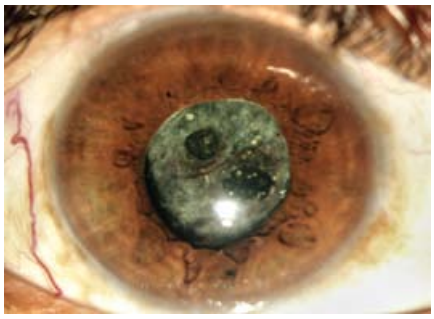


Fig. 8.40.2: Pigmentary PCO in pseudophakia

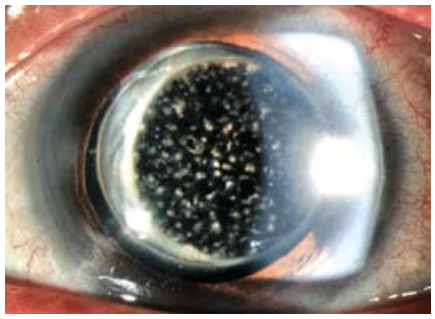


Fig. 8.40.3: Inflammatory deposits in pseudophakia

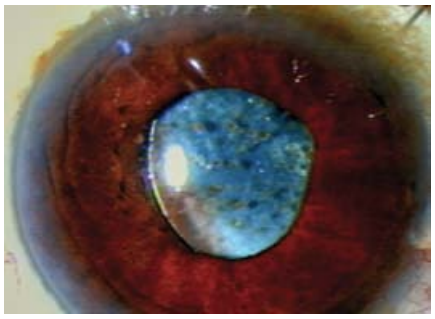


Fig. 8.40.4: Dense PCO—inflammatory

Posterior Capsular Folds

- Seen in some cases of pseudophakia, due to abnormality in IOL—size placed in-the-bag
- Capsule is stretched in one direction (parallel to the haptics) with capsular folds **(Fig. 8.41.1)**
- May be seen after a single-piece IOL and may give rise to Maddox rod effects **(Figs 8.41.2A and B)**
- Sometimes, may create star folds **(Fig. 8.41.3)**.

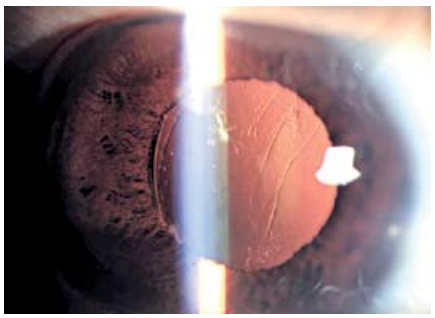


Fig. 8.41.1: Posterior capsular folds

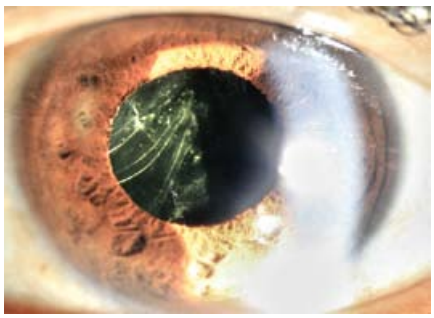


Fig. 8.41.2A: Posterior capsular folds—Maddox rod effect

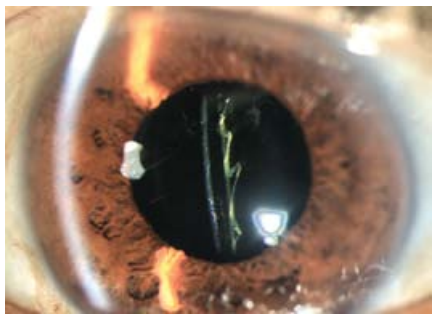


Fig. 8.41.2B: Posterior capsular folds—Maddox rod effect

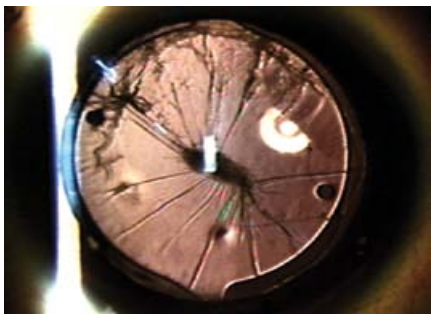


Fig. 8.41.3: Posterior capsular star folds

Pseudophakic Deposits/ Opacities/Defects

- *Inflammatory precipitates:* Associated with postoperative uveitis **(Figs 8.42.1A to D)**
- *Pigments deposition*: Associated with iritis **(Fig. 8.42.2)**
- *IOL glistening:* It is a very common problem with hydrophobic acrylic IOL **(Figs 8.42.3A to C)**. It may be seen in other types of IOL. Normally, it does not interfere with vision except in severe cases where there may be 1-2 lines drop in vision
- *Pitting:* Seen after inadvertent injury to the IOL optic by YAG laser **(Figs 8.42.4 and 8.42.5)**
- *Opacification of IOL:* Seen in some cases of hydrophilic foldable IOL **(Figs 8.42.6A and B)** hydrophobic IOL **(Figs 8.42.7A to C)** and silicone IOL **(Fig. 8.42.8)**
- *Blue discoloration:* Seen in hydrophilic foldable IOL after trypan blue capsulorhexis
- *Cracking of IOL optic:* May be seen in any type of IOL **(Figs 8.42.9A to D)**
- *Yellow coloration:* With yellow blue-blocker IOLs
- *Multiple rings:* Normal seen with multifocal IOL **(Figs 8.42.10A and B)** and may be with some design of monofocal IOL **(Fig. 8.42.11)**.

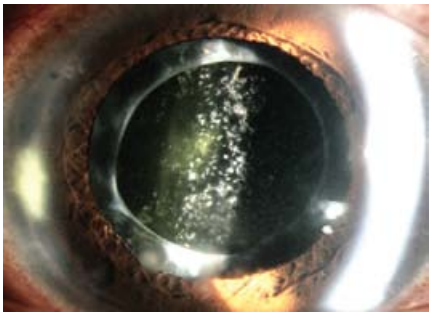


Fig. 8.42.1A: Inflammatory deposits on hydrophobic acrylic IOL

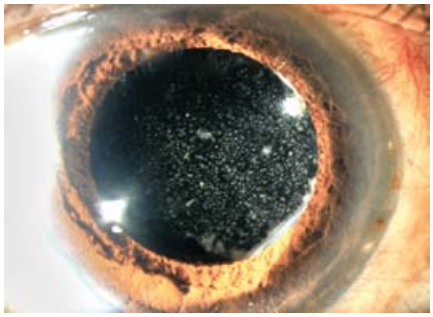


Fig. 8.42.1B: Inflammatory deposits on hydrophilic IOL surface

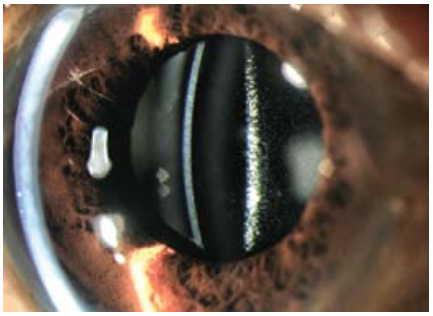


Fig. 8.42.1C: Inflammatory deposits on hydrophilic IOL surface

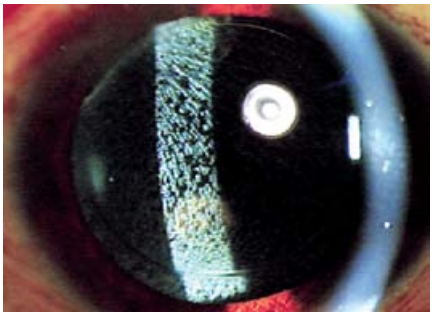


Fig. 8.42.1D: Inflammatory deposits on PMMA IOL surface

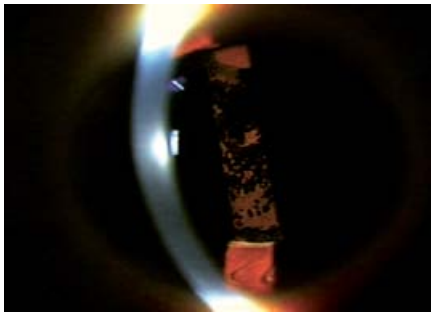


Fig. 8.42.2: Pigment deposition on IOL

Fig. 8.42.3A: Glistening of hydrophilic acrylic IOL

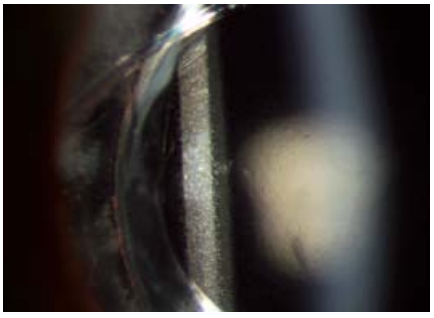


Fig. 8.42.3B: Glistening of hydrophobic acrylic IOL

Fig. 8.42.3C: Glistening of hydrophobic acrylic IOL

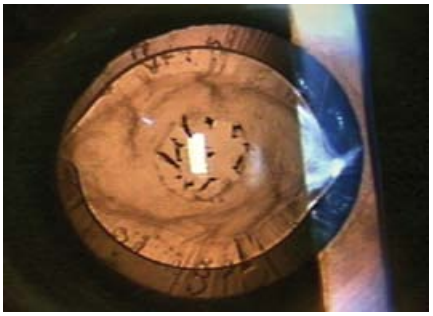


Fig. 8.42.4A: IOL pitting after YAG capsulotomy

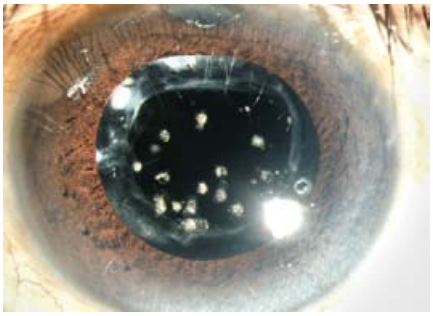


Fig. 8.42.4B: IOL pitting after YAG capsulotomy

Fig. 8.42.4C: IOL pitting after YAG capsulotomy

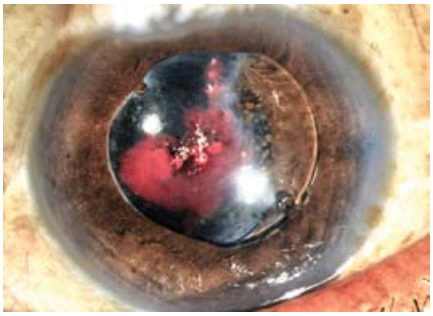


Fig. 8.42.5: IOL pitting with precapsular hemorrhage after YAG

Fig. 8.42.6A: IOL total opacification—hydrophilic materials

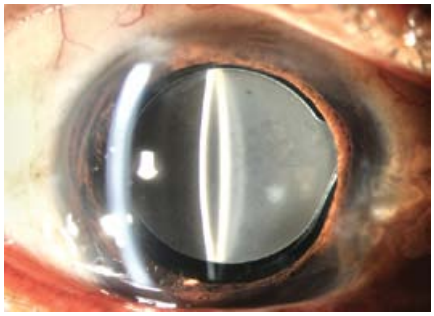


Fig. 8.42.6B: IOL total opacification—hydrophilic materials

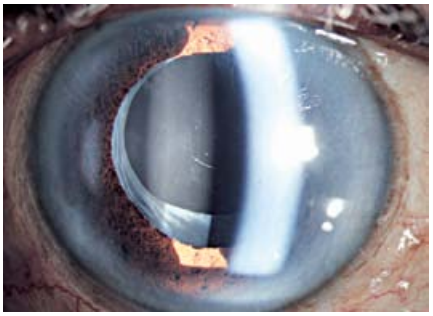


Fig. 8.42.7A: IOL partial opacification—hydrophobic material

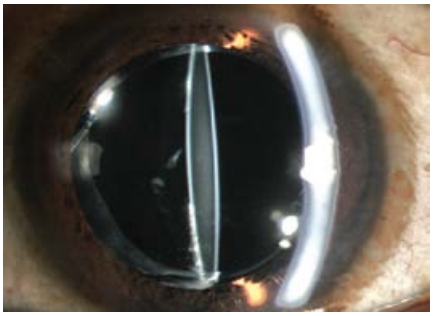


Fig. 8.42.7B: IOL partial opacification—hydrophobic material

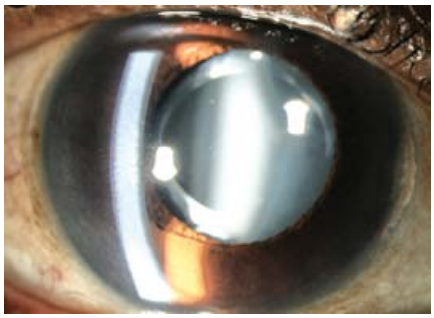


Fig. 8.42.7C: IOL partial opacification—hydrophobic material

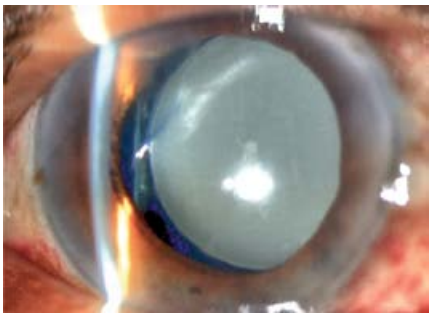


Fig. 8.42.8: IOL total opacification—silicone material

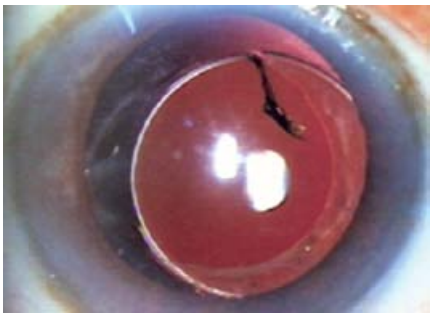


Fig. 8.42.9A: IOL—optic crack

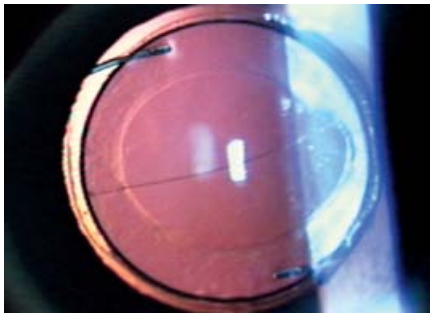


Fig. 8.42.9B: IOL—optic crack

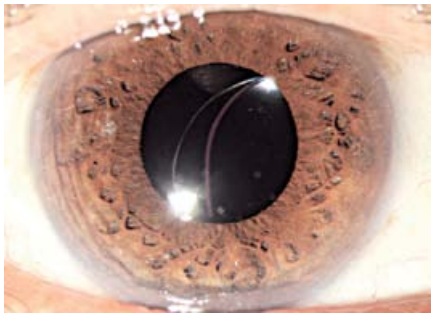


Fig. 8.42.9C: Decentered IOL—broken haptic

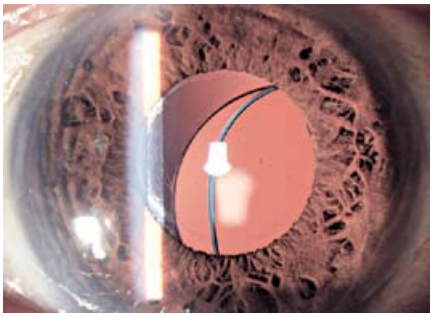


Fig. 8.42.9D: Decentered IOL—broken haptic

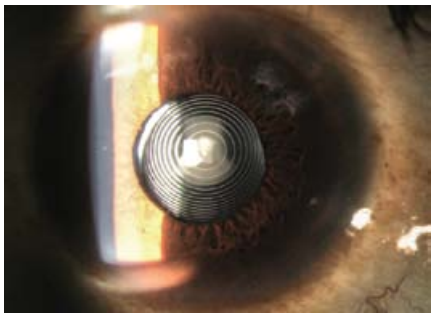


Fig. 8.42.10A: Multiple rings—multifocal—IOL optic

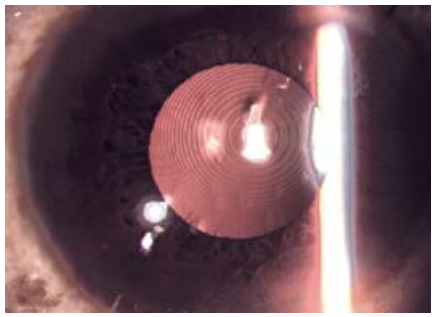


Fig. 8.42.10B: Multiple rings—multifocal—IOL optic

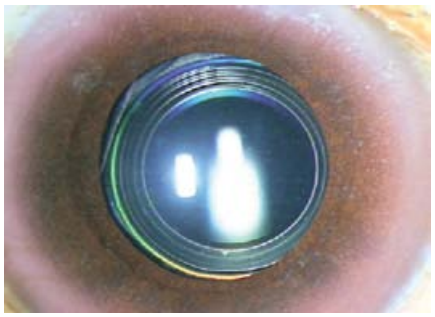


Fig. 8.42.11: Multiple rings—monofocal IOL optic

Displacement of the Crystalline Lens and IOLs

- There may be subluxation or dislocation of the crystalline lens and IOLs
- *Causes:*
  - *Congenital ectopia lentis* (already discussed)
  - *Acquired:* Traumatic, degenerative, syphilis, hypermaturity, anterior uveal tumor, etc.
  - *For IOL:* Capsulozonular problems during opeartion, trauma or spontaneous.

Subluxation of the Crystalline Lens

- A portion of the supporting zonules is absent, and the lens lacks support in that quadrant
- Diagnosis should be confirmed after full dilatation of the pupil **(Figs 8.43.1A and B)**
- Edge of the lens is visible as a golden crescentic line in oblique illumination **(Fig. 8.43.2)**
- May be associated with vitreous prolapse in anterior chamber **(Figs 8.43.3A and B)**.

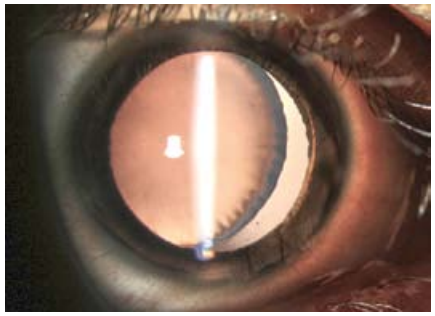


Fig. 8.43.1A: Subluxation of the lens—traumatic

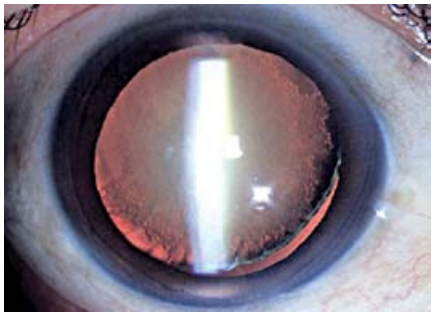


Fig. 8.43.1B: Subluxation of lens in high myopia

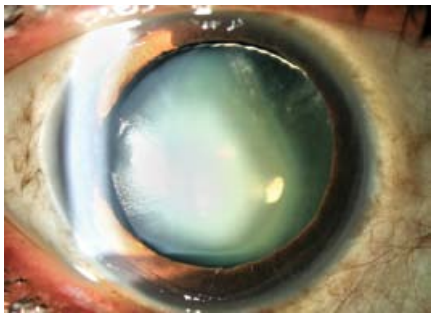


Fig. 8.43.2: Subluxation of the lens—golden crescent—trauma

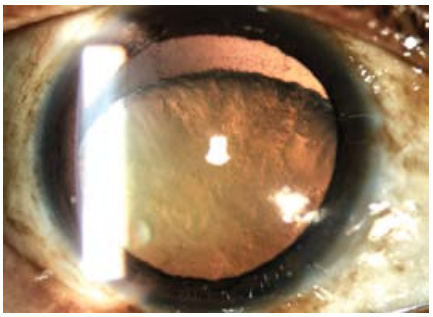


Fig. 8.43.3A: Subluxation of the lens with vitreous herniation

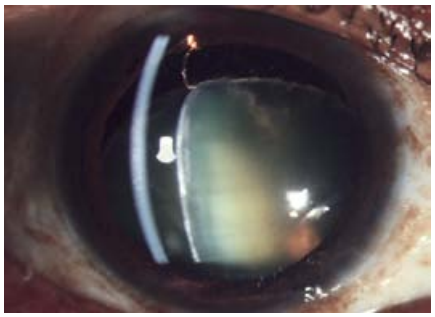


Fig. 8.43.3B: Subluxation of the lens with vitreous herniation

Dislocation of the Crystalline Lens

- Crystalline lens is completely unsupported by the zonular fibers
- Displaced from the pupillary area
- Presence of signs of aphakia
- Associated with other signs in traumatic cases
- Lens dislocation may be anterior **(Figs 8.44.1A and B)**, posterior **(Fig. 8.44.2)** or rarely subconjunctival, called phacocele **(Figs 8.44.3A and B)**.

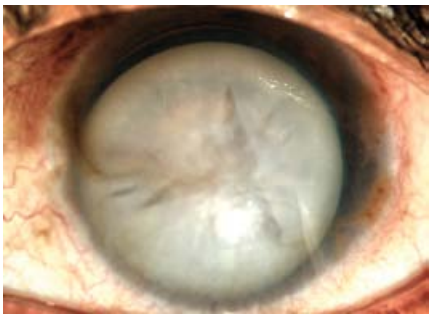


Fig. 8.44.1A: Anterior dislocation of mature cataract

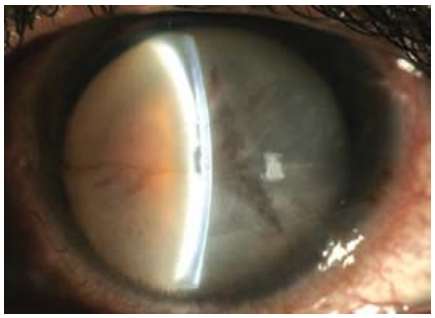


Fig. 8.44.1B: Anterior dislocation of mature cataract

- *Anterior dislocation:*
  - Lens is dislocated into the bottom of the anterior chamber
  - Appears as an 'oil-globule' due to total internal reflection **(Figs 8.44.4A and B)**
  - Cataractous lens may also dislocate anteriorly **(Figs 8.44.5A and B)**.
- *Posterior dislocation:*
  - Lens can be seen as translucent or opaque mass in the vitreous cavity **(Figs 8.44.6A and B)**
  - May be fixed or mobile (wandering) in the vitreous
- *Treatment: In anterior dislocation,* gentle removal of the crystalline lens via manual small incision, anterior vitrectomy and then either AC IOL or scleral fixation IOL implantation

  In posterior dislocation: Pars plana lensectomy with vitrectomy with AC IOL or scleral fixation IOL implantation.

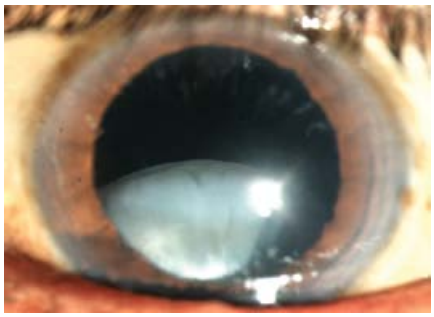


Fig. 8.44.2: Posterior dislocation of the lens into the vitreous

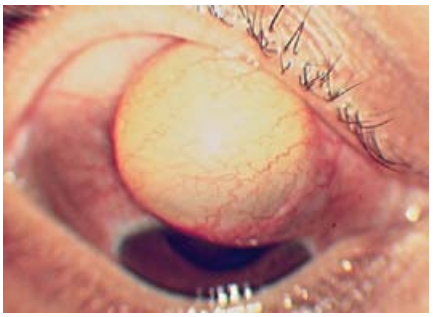


Fig. 8.44.3A: Subconjunctival dislocation of the lens after cow-horn trauma phacocele

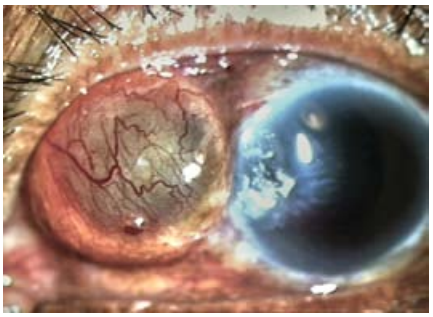


Fig. 8.44.3B: Subconjunctival dislocation of the lens after trauma

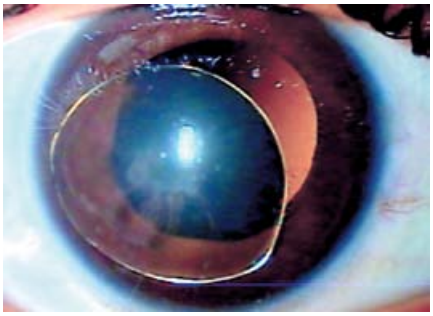


Fig. 8.44.4A: Anterior dislocation of the lens—oil globule appearance

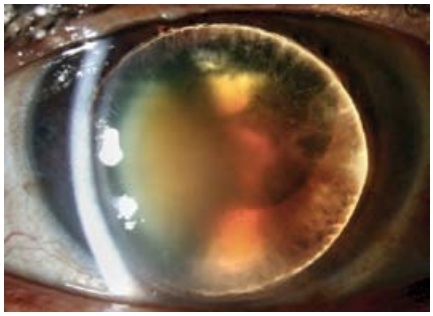


Fig. 8.44.4B: Anterior dislocation of the lens—oil globule appearance

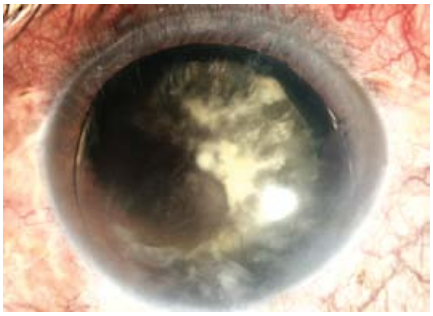


Fig. 8.44.5A: Anterior dislocation of suprahard cataract

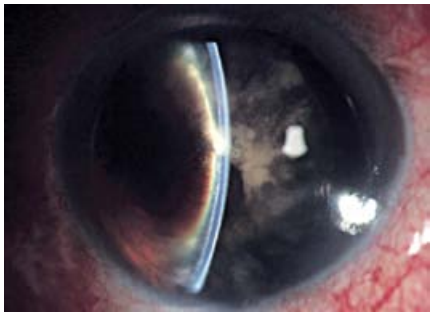


Fig. 8.44.5B: Anterior dislocation of suprahard cataract

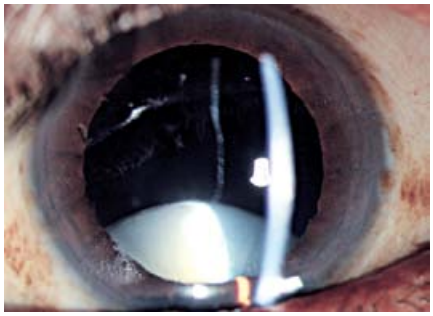


Fig. 8.44.6A: Posterior dislocation of the lens with intact vitreous face

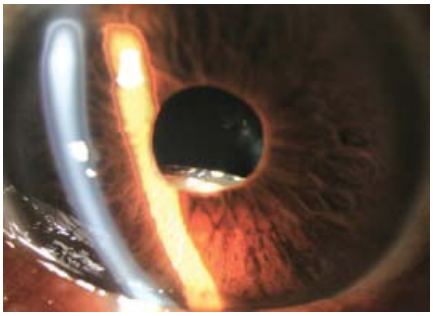


Fig. 8.44.6B: Posterior dislocation of the lens into the vitreous

Displacement of the IOL

- Ideally, it should be well centered and preferable in-the-bag **(Figs 8.45.1A to D)** and in case of AC IOL again it should be well centered. It is usually angle-fixated as in Kelman multiflex **(Figs 8.45.2A and B)** or it may be old Shepherd's AC IOL **(Fig. 8.45.3)** or iris fixated AC IOL **(Figs 8.45.4A and B)**. In case of phakic posterior chamber IOL, again it should be well centered with good vaulting **(Figs 8.45.5A and B)**
- Displacement usually associated with posterior capsular rent of variable degree; with pseudoexfoliation and with preoperative diagnosis of subluxation
- A PC IOL may be displaced downwards—called 'sunset sign' **(Figs 8.45.6 and 8.45.7)**, upwards **(Figs 8.45.8A and B)** or sideways **(Fig. 8.45.9)**
- Diagnosis is usually obvious after full dilatation of the pupil

- In case of AC IOL, there may be various kind of displacements with or without corneal endothelial decompensation **(Figs 8.45.10 and 8.45.11)**
- Sometimes, in traumatic cases, the PC IOL can dislocate into the AC **(Fig. 8.45.12)** and with intact capsular bag **(Figs 8.45.13A and B)**.
- Sometime, only the part of the haptic is visible in the pupillary area **(Fig. 8.45.14)**
- An IOL may be dislocated in the vitreous and the pupillary aperture remains clear
- Rarely may dislocate in the subconjunctival space after a trauma, called *pseudophacocele* **(Figs 8.45.15 and 8.45.16)**.

Fig. 8.45.1A: PMMA PC IOL in-the-bag

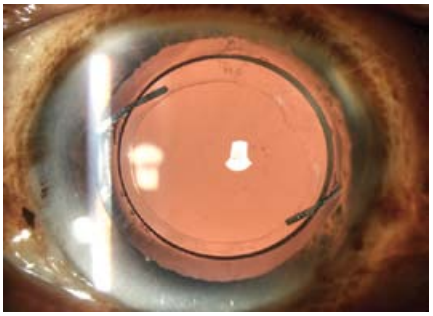


Fig. 8.45.1B: Multipiece foldable PC IOL in-the-bag

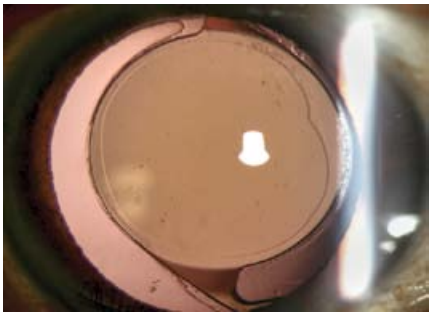


Fig. 8.45.1C: Single-piece foldable PC IOL in-the-bag

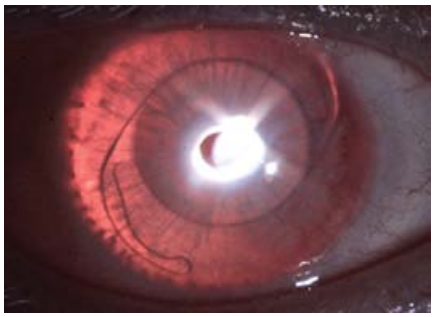


Fig. 8.45.1D: Single-piece IOL in-the-bag in albinism—note iris transillumination

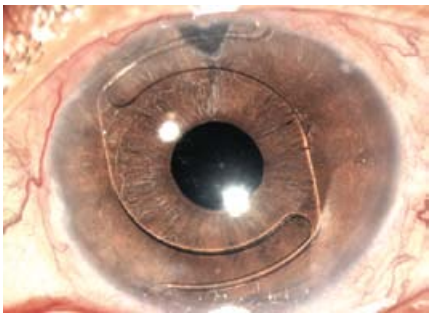


Fig. 8.45.2A: Angle fixated AC IOL—Kelman multiflex—good centration

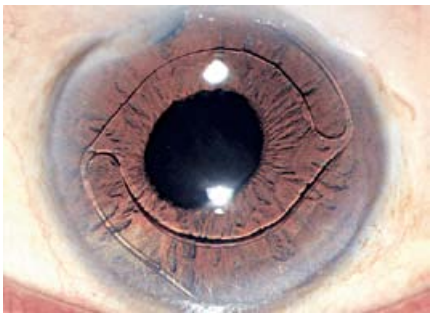


Fig. 8.45.2B: Kelmen multiflex AC IOL—mild decentration with oval pupil

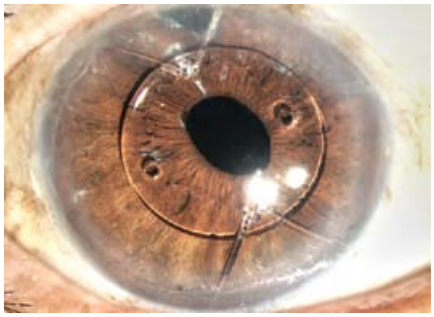


Fig. 8.45.3: Angle fixated AC IOL—Shepherd's IOL

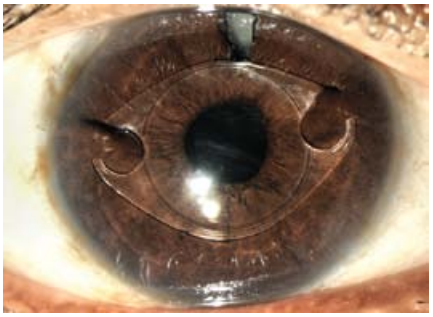


Fig. 8.45.4A: Iris fixated AC IOL—Worst-Singh IOL

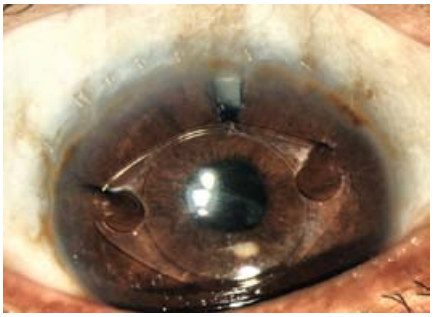


Fig. 8.45.4B: Iris fixated AC IOL—Worst-Singh—with steel sutures at limbus

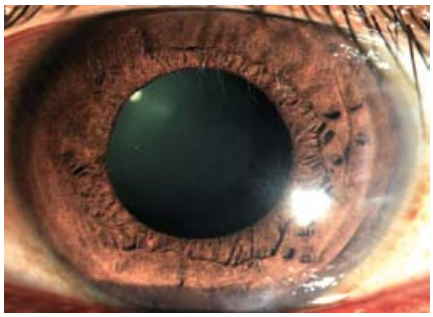


Fig. 8.45.5A: Phakic IOL in high myopia—posterior chamber

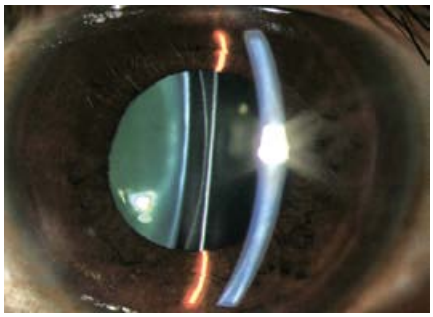


Fig. 8.45.5B: Phakic IOL in high myopia—posterior chamber

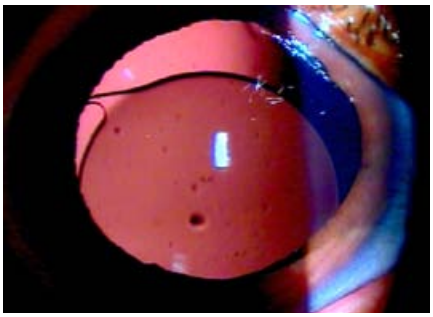


Fig. 8.45.6A: Decentration of PC IOL

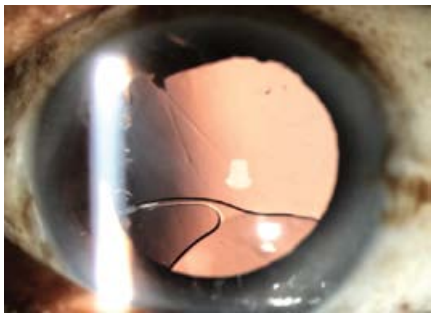


Fig. 8.45.6B: PMMA IOL—downward displacement—sunset sign

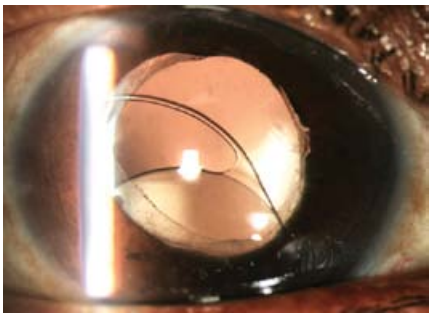


Fig. 8.45.6C: PMMA IOL—downward displacement—sunset sign

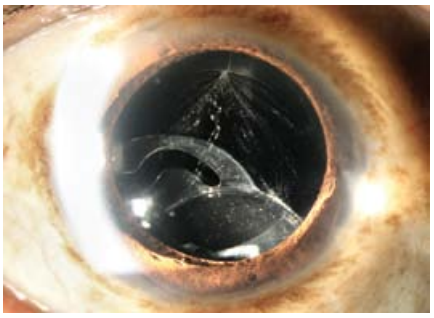


Fig. 8.45.7A: Foldable IOL—downward displacement—sunset sign

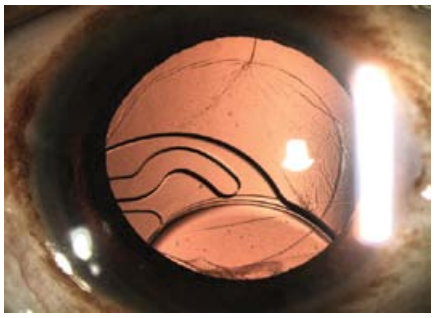


Fig. 8.45.7B: Foldable IOL—downward displacement—sunset sign

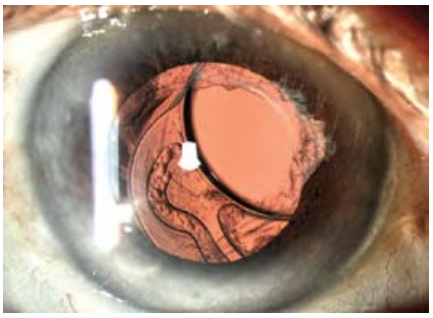


Fig. 8.45.8A: IOL—upward displacement

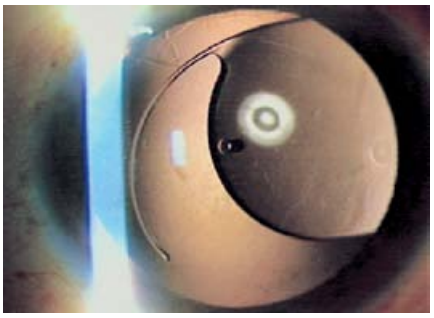


Fig. 8.45.8B: IOL—sideway displacement

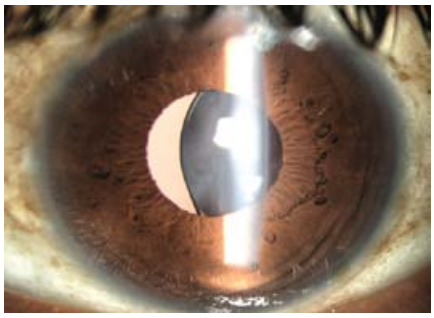


Fig. 8.45.9: Decentration of PC IOL—sideways

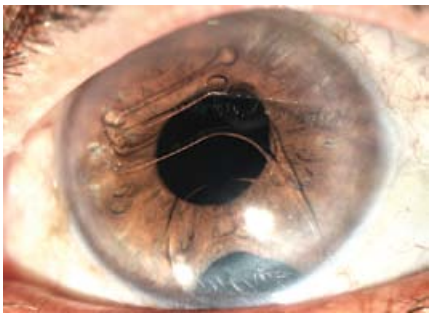


Fig. 8.45.10A: Decentration of AC IOL

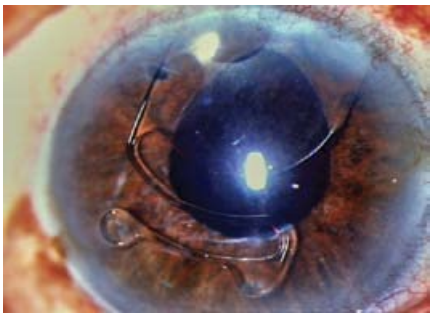


Fig. 8.45.10B: IOL decentration—upward displacement of AC IOL

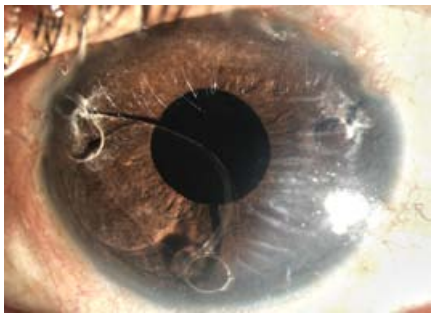


Fig. 8.45.11A: Decentration of IOL—Worst-Singh IOL

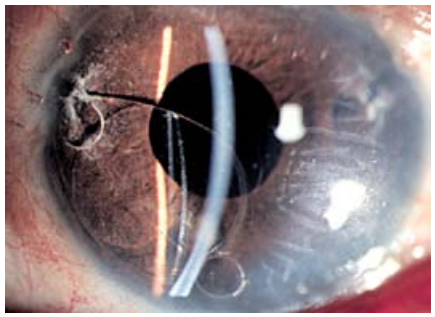


Fig. 8.45.11B: Decentration of IOL—Worst-Singh IOL

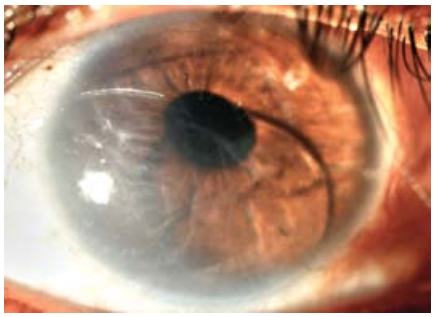


Fig. 8.45.12: Anterior dislocation of PC IOL—post-traumatic

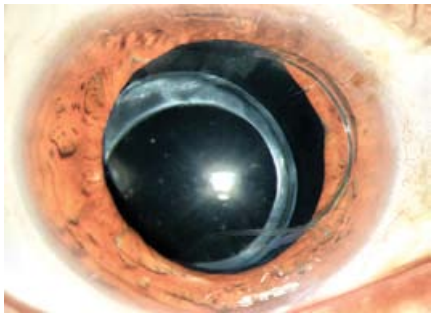


Fig. 8.45.13A: Dislocation of PC IOL with the capsular bag—posterior

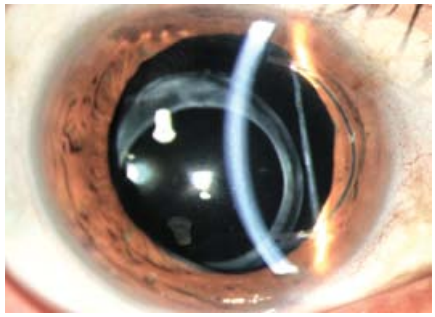


Fig. 8.45.13B: Dislocation of PC IOL with the capsular bag—posterior

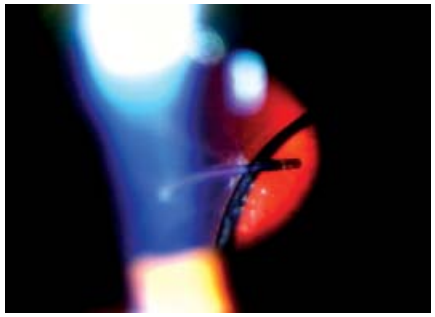


Fig. 8.45.14: Posterior dislocation of PC IOL—post-traumatic

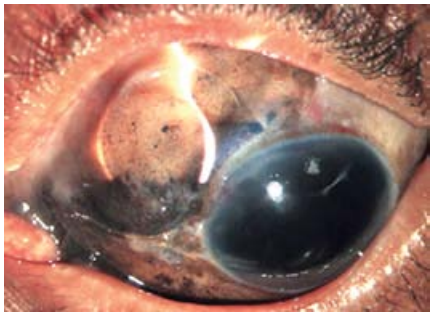


Fig. 8.45.15: Subconjunctival dislocation of IOL—pseudophacocele trauma by cow-horn

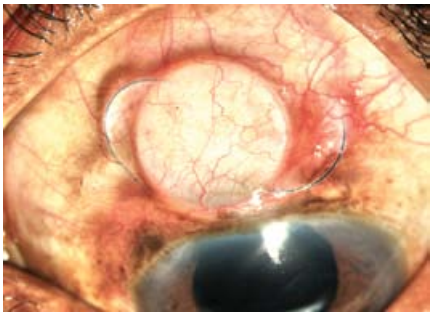


Fig. 8.45.16: Subconjunctival dislocation of IOL—pseudophacocele trauma by plastic ball

Miscellaneous Pseudophakic Conditions

- *Iris capture:* It is usually associated with posterior chamber IOL and more common with PMMA material **(Figs 8.46.1 and 8.46.2)**
- *PC IOL in AC:* It is a bad and very wrong surgical practice! Ultimately, it gives rise to pseudophakic bullous keratopathy **(Figs 8.46.3 and 8.46.4)**
- *Haptic in tunnel or out of the wound:* In AC IOL **(Figs 8.46.5A and B)**, PC IOL **(Fig. 8.46.5C)** or scleral fixation IOL
- *One haptic of PC IOL in the anterior chamber* **(Figs 8.46.6A to C)**
- *Cotton fiber behind the IOL* **(Fig. 8.46.7)** or *plastic tags behind the IOL* as it happens with hydrophobic IOL with injector syetem. It is the Teflon coating of the cartridge **(Fig. 8.46.8)**

- *Unfolded haptics of foldable IOL:* It is more common with both single-piece hydrophilic **(Figs 8.46.9A to D)** and hydrophobic IOL **(Figs 8.46.9E and F)**. Also occur in multipiece foldable IOL especially with injector system **(Figs 8.46.10A and B)**
- The PC IOL may be sulcus fixated **(Fig. 8.46.11A)** or in-the-bag, in presence of posterior capsular rent **(Fig. 8.46.11B)**
- Silicone oil adherence with IOL, especially it occurs with silicone IOL **(Fig. 8.46.12).**

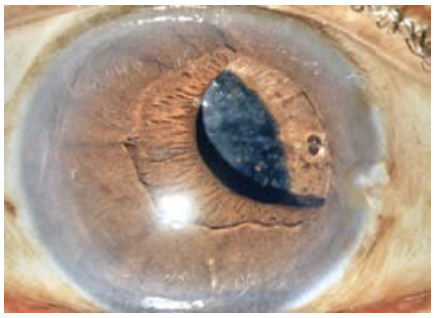


Fig. 8.46.1A: Iris capture of IOL

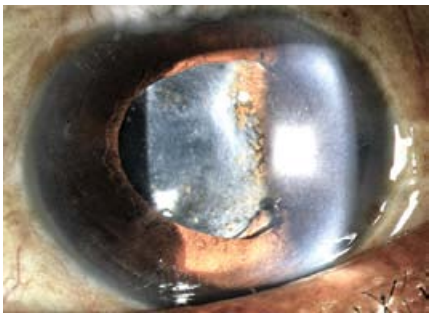


Fig. 8.46.1B: Iris capture of IOL with fibrous PCO

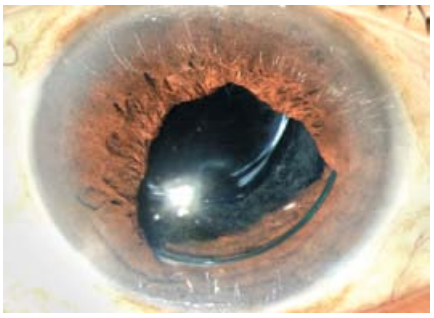


Fig. 8.46.2A: Iris capture of haptic of an IOL

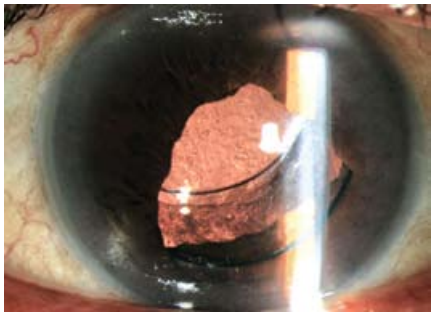


Fig. 8.46.2B: Iris capture of haptic of an IOL

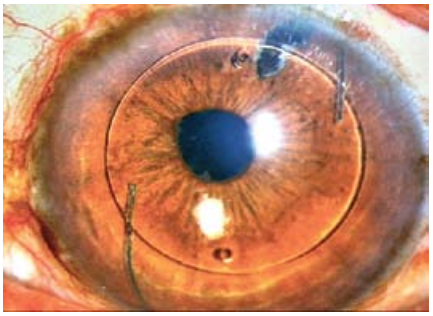


Fig. 8.46.3A: PC IOL in AC !

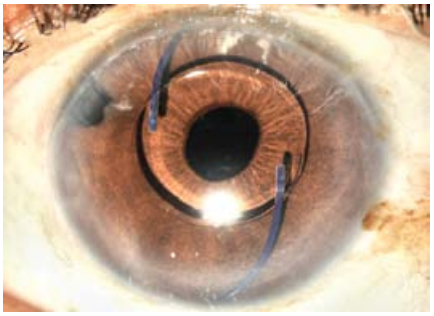


Fig. 8.46.3B: PC IOL in AC !

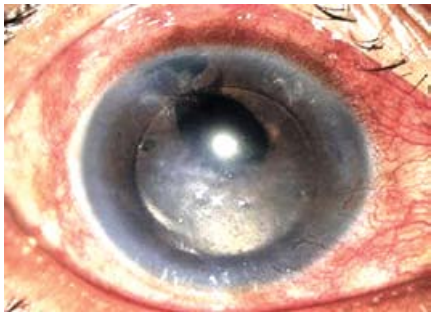


Fig. 8.46.4: PC IOL in AC ! with bullous keratopathy

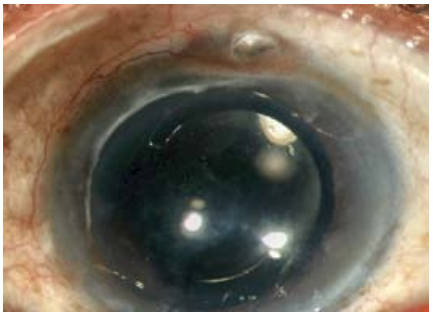


Fig. 8.46.5A: Haptic of AC IOL outside the wound

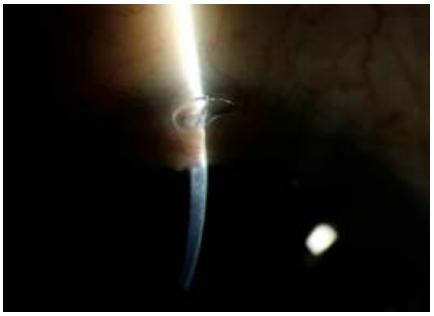


Fig. 8.46.5B: Haptic of AC IOL outside the wound

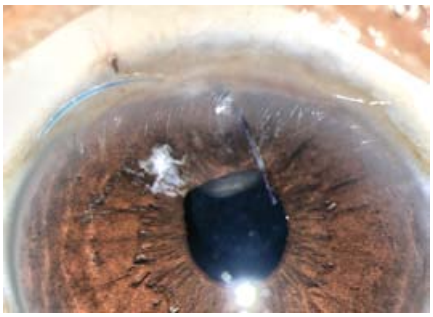


Fig. 8.46.5C: Haptic of PC IOL outside the wound

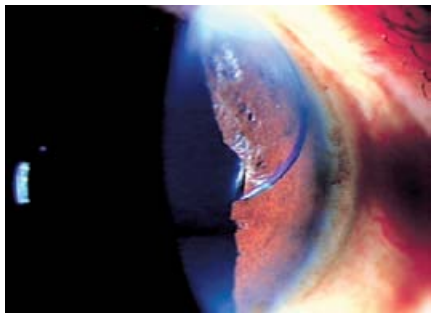


Fig. 8.46.6A: One haptic of PC IOL in AC

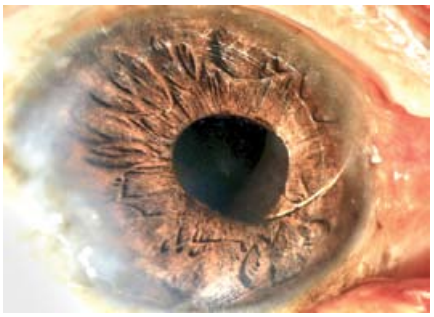


Fig. 8.46.6B: One haptic of PC IOL in AC

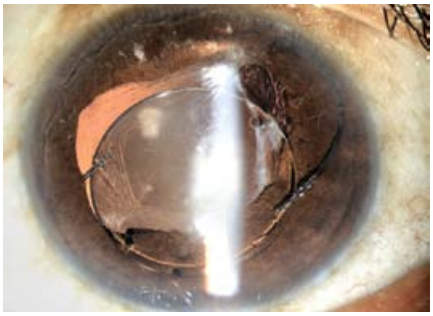


Fig. 8.46.6C: One haptic of PC IOL in AC with optic in AC

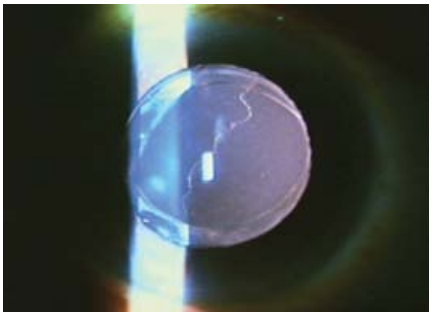


Fig. 8.46.7: Cotton fiber behind the PC IOL

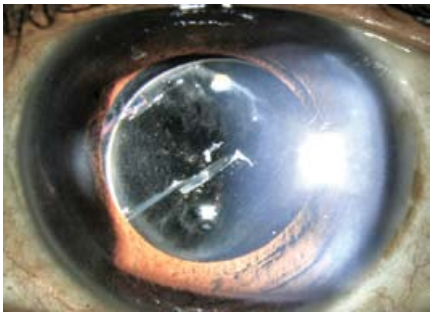


Fig. 8.46.8: Plastic tags (from teflon coating of IOL cartridge)

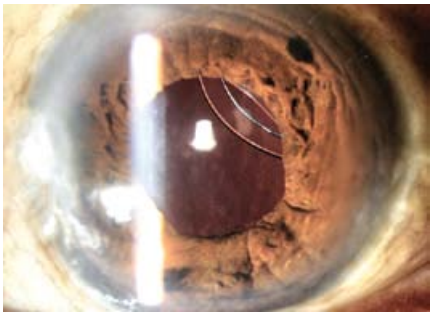


Fig. 8.46.9A: Folded haptic—one haptic

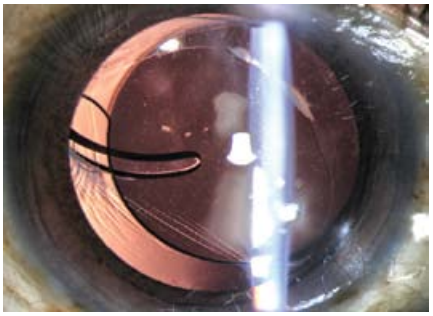


Fig. 8.46.9B: Folded haptic—one haptic—tail of an IOL!

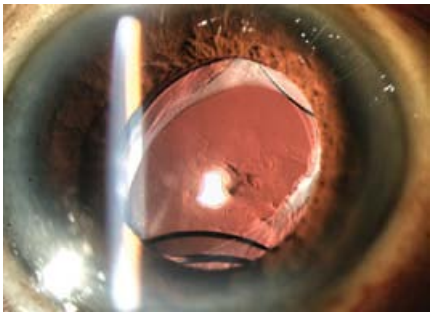


Fig. 8.46.9C: Folded haptic—hydrophilic IOL—both haptics

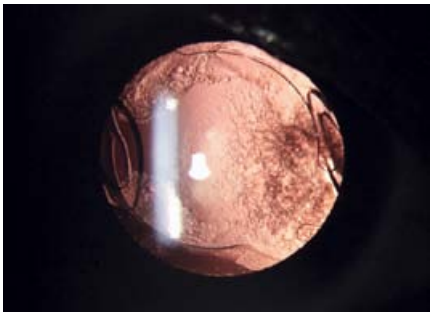


Fig. 8.46.9D: Folded bath haptics with PCO—hydrophilic IOL

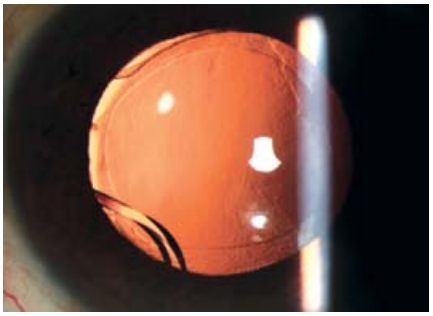


Fig. 8.46.9E: Folded haptic—hydrophobic IOL—one haptic

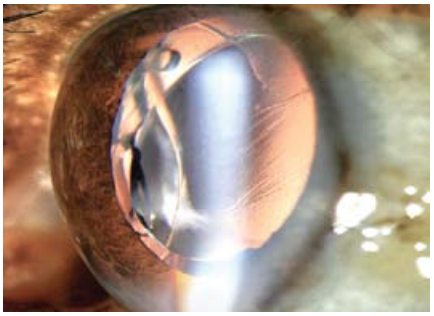


Fig. 8.46.9F: Haptic fold in hydrophobic IOL—one haptic

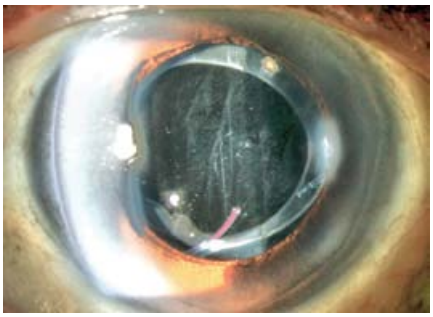


Fig. 8.46.10A: Deformed haptic in pupillary area—multipiece

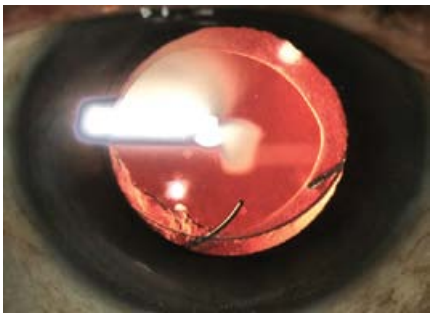


Fig. 8.46.10B: Deformed haptic in pupillary area—multipiece

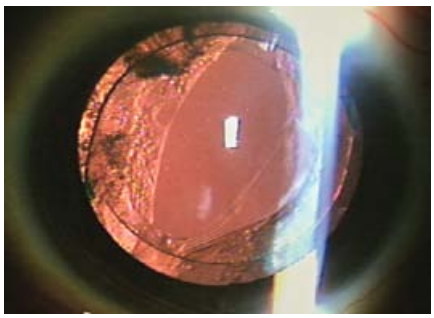


Fig. 8.46.11A: Sulcus fixation PC IOL—PMMA IOL

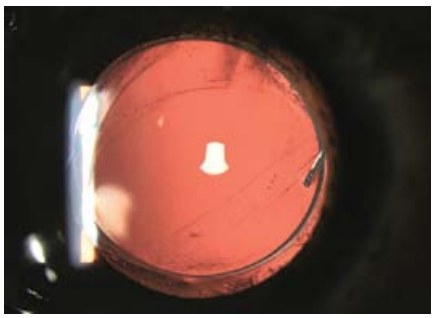


Fig. 8.46.11B: PC rent and well centered PC IOL in-the-bag

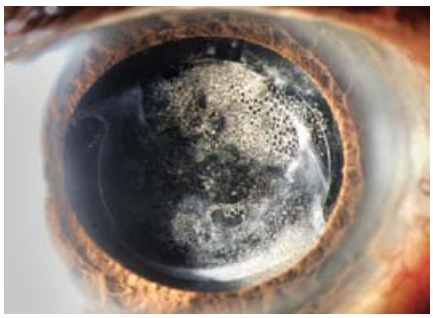


Fig. 8.46.12: Silicone oil adherence with IOL

Aphakia

- Most common cause—surgical **(Fig. 8.47.1)**, and then traumatic **(Fig. 8.47.2)**, though sometimes spontaneous dislocation and absorption occurs
- Deep anterior chamber
- Jet black pupil
- Tremulousness of iris
- Associated peripheral iridectomy
- Vitreous herniation is common **(Figs 8.47.3A and B)**
- *Treatment:* Aphakic glasses, contact lens or secondary anterior chamber or scleral fixation IOL.

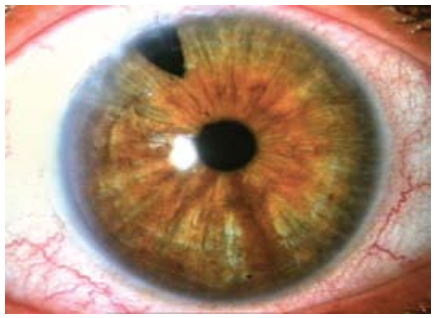


Fig. 8.47.1: Good surgical aphakia

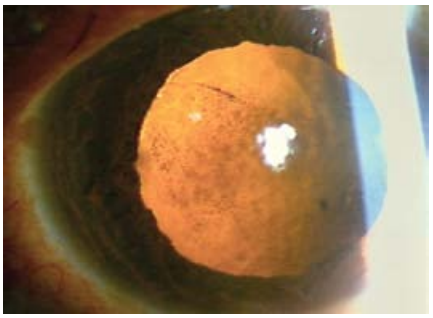


Fig. 8.47.2: Traumatic aphakia with mydriasis

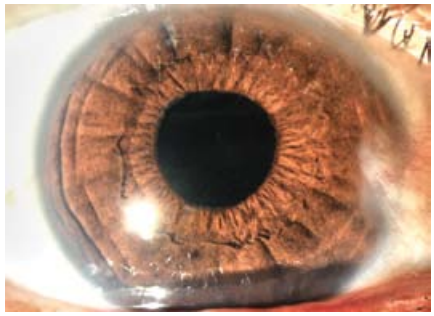


Fig. 8.47.3A: Surgical aphakia with vitreous herniation

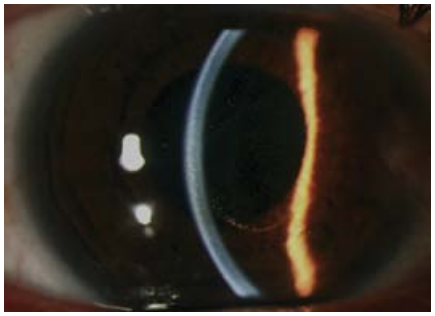


Fig. 8.47.3B: Surgical aphakia with vitreous herniation

CHAPTER

9 Glaucomas

CHAPTER OUTLINE

Congenital or Infantile Glaucoma (Buphthalmos)

- Primary congenital glaucoma
- Secondary developmental glaucoma

Primary Angle-Closure Glaucoma

- Acute congestive attack
- Chronic congestive attack
- Stage of absolute glaucoma
- Slit lamp grading of angle (van Herick)

Primary Open Angle Glaucoma

- Cupping of the optic disk

Secondary Glaucomas

- Pseudoexfoliation glaucoma
- Phacomorphic glaucoma
- Phacolytic glaucoma
- Lens-particle glaucoma
- Associated with ectopia lentis/subluxation or dislocation of lens
- Pigmentary glaucoma
- Inflammatory secondary glaucoma
- Glaucoma associated with trauma
- Glaucoma following intraocular surgery
- Malignant (ciliary block) glaucoma
- Neovascular glaucoma
- Iridocorneal endothelial (ICE) syndrome
- Other causes of secondary glaucoma

Filtering Bleb Abnormalities

- Normal functioning bleb
- Failed/flat filtering bleb
- Large cystic filtering bleb
- Multilocular filtering bleb
- Overhanging filtering bleb
- Other morphological types of bleb
- Overfiltering bleb
- Leaking filtering bleb
- Blebitis (bleb infection)

Other Complications

- Bleb hemorrhage
- Wind-shield wiper sign
- Pseudobleb

Congenital or Infantile Glaucoma (Buphthalmos)

Primary Congenital Glaucoma

- Congenital or infantile glaucoma is due to simple outflow obstruction
- Rare, unilateral **(Fig. 9.1.1A)** or bilateral **(Fig. 9.1.1B)** condition, may be asymmetrical at the time of presentation **(Fig. 9.1.2)**
- Autosomal recessive inheritance
- 40% cases are true congenital and 50% are infantile **(Fig. 9.1.3)**
- Boys are more affected than girls
- Eyeball becomes enlarged, if the IOP becomes elevated prior to age of three years
- Cornea is enlarged, globular and steamy **(Fig. 9.1.4)** and with photophobia and blepharospasm **(Fig. 9.1.5)**
- Horizontal curvilinear lines are seen on the back of the cornea, known as Haab's striae **(Fig. 9.1.6)**
- Blue discoloration of sclera **(Figs 9.1.7A to C)**
- Deep anterior chamber
- Cupping of the disk
- It is not uncommon to see unilateral cases with late presentation **(Figs 9.1.8A and B)**
- Unilateral cases must be differentiated from unilateral high myopia **(Fig. 9.1.9)**
- *Treatment:* Examination under anesthesia; goniotomy, trabeculotomy or trabeculectomy and trabeculotomy, and visual rehabilitation.

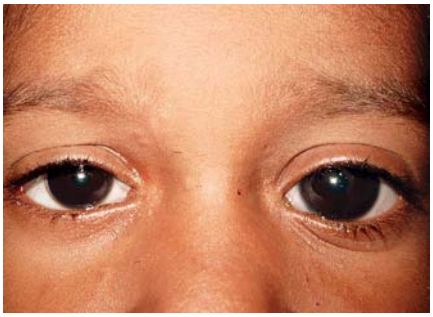


Fig. 9.1.1A: Primary congenital glaucoma—unilateral

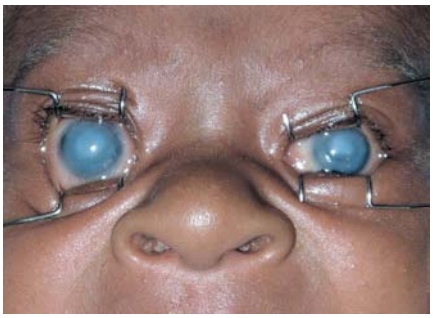


Fig. 9.1.1B: Primary congenital glaucoma—bilateral

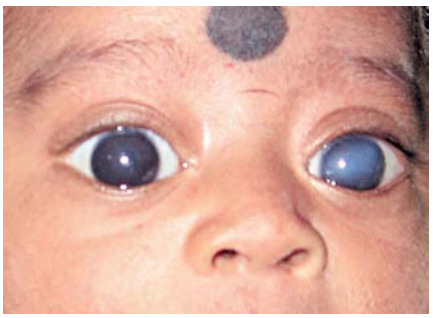


Fig. 9.1.2: Primary congenital glaucoma—bilateral asymmetrical

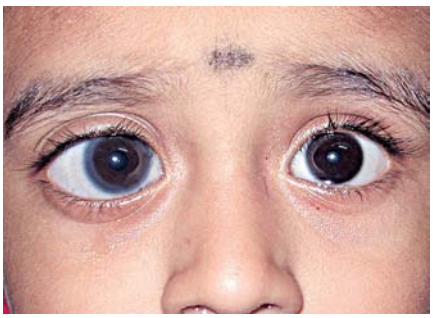


Fig. 9.1.3: Primary infantile glaucoma—RE

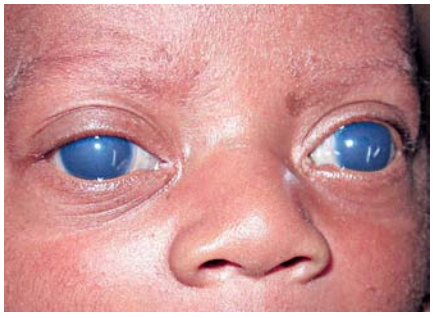


Fig. 9.1.4: Primary congenital glaucoma—cornea globular and steamy

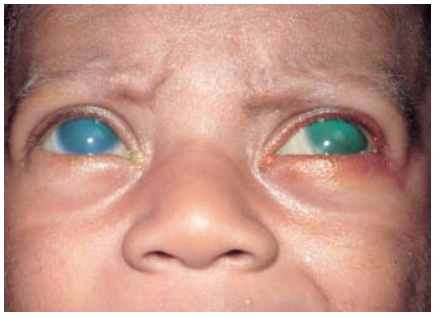


Fig. 9.1.5: Congenital glaucoma—blepharospasm and photophobia

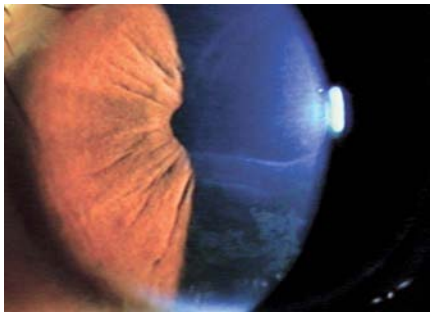


Fig. 9.1.6: Congenital glaucoma—Haab's striae

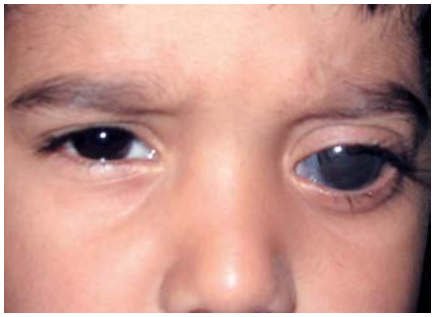


Fig. 9.1.7A: Congenital glaucoma—blue sclera—LE

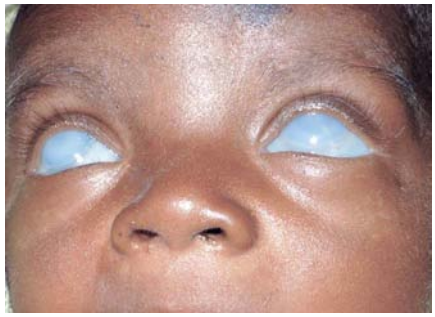


Fig. 9.1.7B: Congenital glaucoma—blue sclera

Fig. 9.1.7C: Infantile glaucoma—buphthalmos with blue sclera

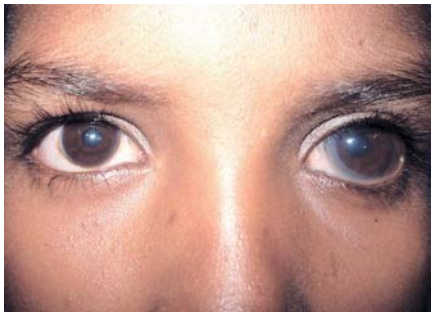


Fig. 9.1.8A: Juvenile glaucoma

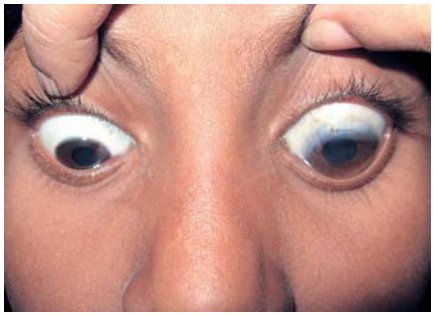


Fig. 9.1.8B: Juvenile glaucoma—LE

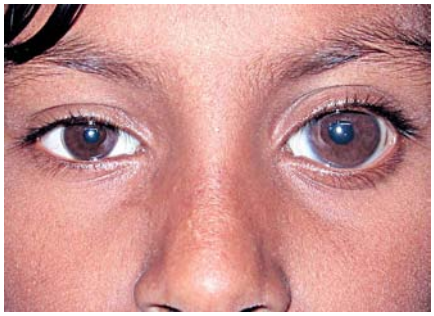


Fig. 9.1.9: Buphthalmos like appearance in unilateral very high myopia

Secondary Developmental Glaucoma

May be associated with other ocular disorders or systemic diseases (secondary form). They are:

- Aniridia **(Figs 9.2.1A and B)**
- Rubella syndrome **(Fig. 9.2.2)**
- Sturge-Weber syndrome **(Fig. 9.2.3)**
- Neurofibromatosis
- Mesodermal dysgenesis:
  - *Posterior embyrotoxon* **(Figs 9.2.4A and B):** Starts as prominent anteriorly displaced Schwalbe's line. Sometimes, a normal varient.
  - *Axenfeld anomaly* **(Figs 9.2.5A and B):** Posterior embryotoxon associated with peripheral iris strands that blocks the angle. 50-60% chance of developing glaucoma due to extensive PAS formation **(Figs 9.2.6A and B)**

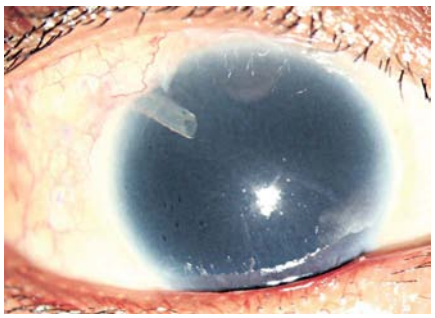


Fig. 9.2.1A: Total aniridia—operated glaucoma drainage device—RE

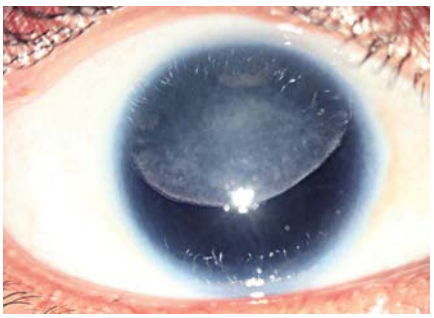


Fig. 9.2.1B: Total aniridia and ectopia lentis with secondary glaucoma—LE

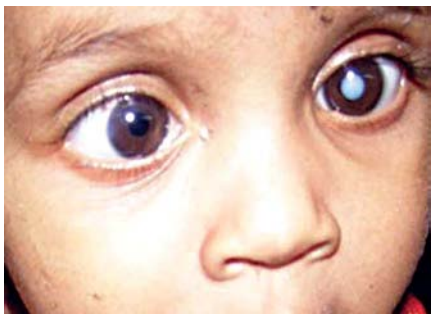


Fig. 9.2.2: Rubella syndrome—buphthalmos with total cataract

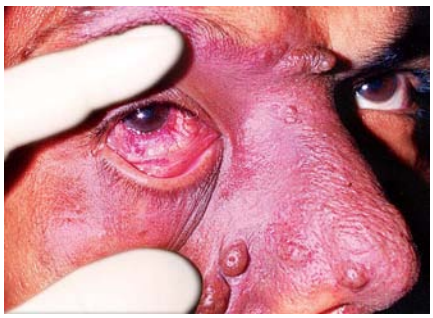


Fig. 9.2.3: Sturge-Weber syndrome

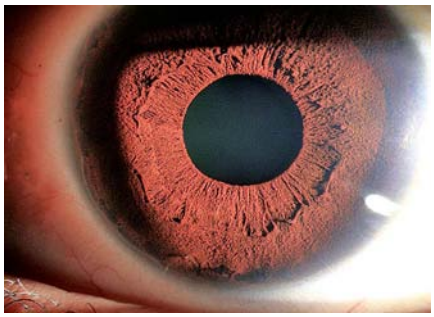


Fig. 9.2.4A: Posterior embryotoxon—as the begining of Axenfeld anomaly

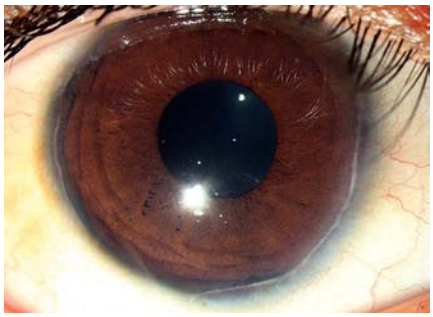


Fig. 9.2.4B: Posterior embryotoxon—narmal variation

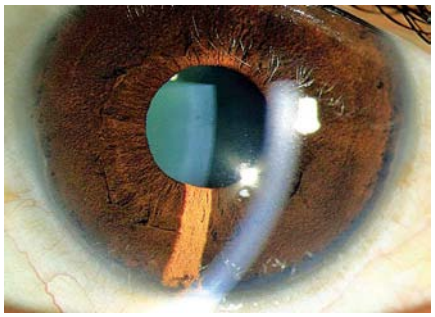


Fig. 9.2.5A: Axenfeld anomaly

Fig. 9.2.5B: Axenfeld anomaly—peripheral iris strands

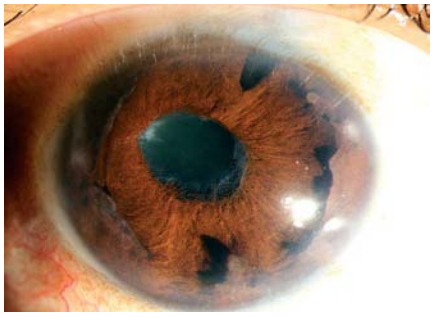


Fig. 9.2.6A: Axenfeld-Rieger's syndrome with failed trabeculectomy

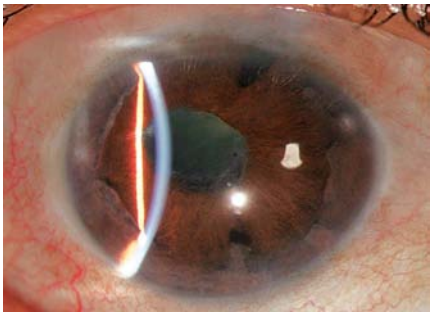


Fig. 9.2.6B: Axenfeld-Rieger's syndrome with failed trabeculectomy

  - *Rieger's anomaly:* When it is further associated with iris thinning and pupillary distortion **(Figs 9.2.7A and B)**
  - *Axenfeld-Rieger's syndrome:* When either of them is associated with dental, craniofacial and skeletal abnormalities **(Figs 9.2.8 and 9.2.9)**
  - *Peter's anomaly:* As described earlier, it is associated with iris strands from corneal defect to lens anterior surface **(Figs 9.2.10A and B)**. In severe form, it may present at birth **(Fig. 9.2.11)**
- Manifestation occurs in early childhood or later
- Presence of other systemic signs
- *Treatment:* Same as primary type **(Fig. 9.2.12)**.

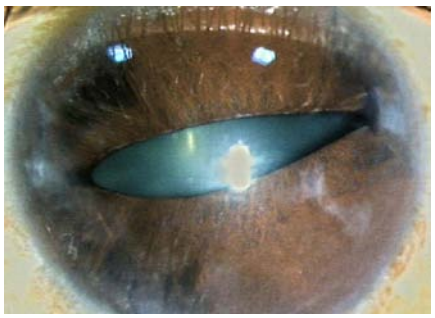


Fig. 9.2.7A: Rieger's anomaly

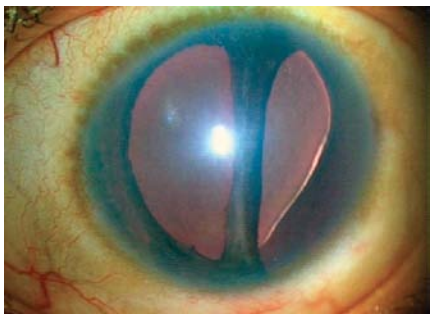


Fig. 9.2.7B: Rieger's anomaly

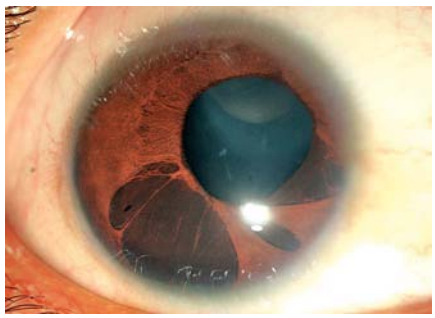


Fig. 9.2.8A: Axenfeld-Rieger's syndrome—RE

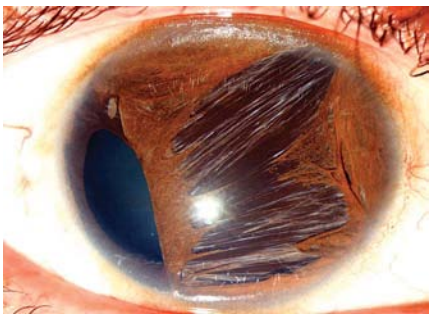


Fig. 9.2.8B: Axenfeld-Rieger's syndrome—LE

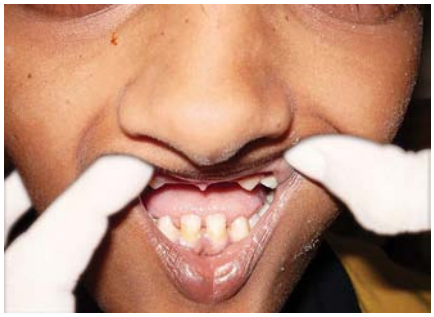


Fig. 9.2.9: Axenfeld-Rieger's syndrome—dental abnormalities

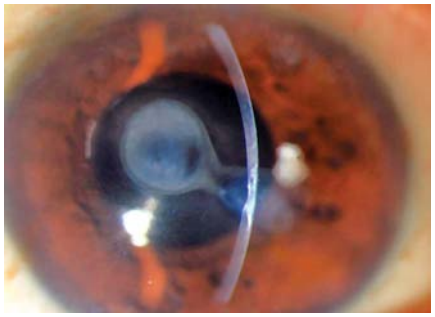


Fig. 9.2.10A: Peter's anomaly—only posterior keratoconus

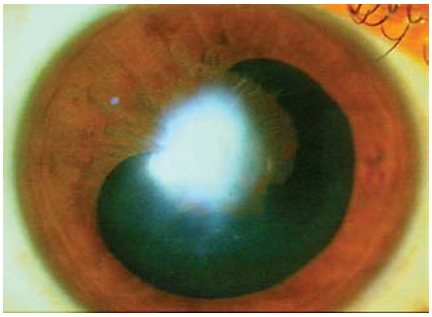


Fig. 9.2.10B: Peter's anomaly—iridocorneo-lental adhesion

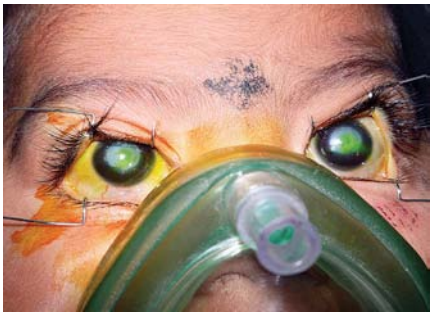


Fig. 9.2.11: Peter's anomaly—bilateral presented at birth

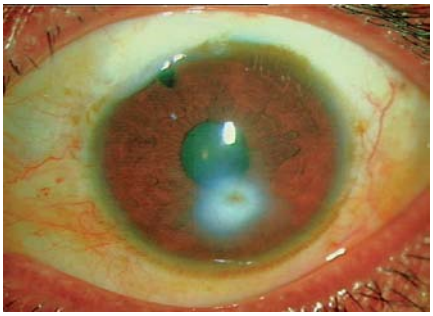


Fig. 9.2.12: Peter's anomaly—operated trabeculectomy

Primary Angle-Closure Glaucoma

Primary angle-closure glaucoma (PACG) is an acute, subacute or chronic glaucoma due to obstruction of the aqueous outflow, solely caused by closure of the angle by the peripheral iris

- Typically, the eye is hypermetropic, with shallow anterior chamber **(Figs 9.3.1A and B)** and narrow angle, even that can be detected by van Herick's method **(Fig. 9.3.1C)**

Acute Congestive Attack

- Lid edema with circumcorneal ciliary congestion **(Fig. 9.3.2)**
- Steamy and insensitive cornea
- Shallow anterior chamber
- Pupil is mid-dilated and oval **(Fig. 9.3.3A)**
- Iris shows atrophic changes adjacent to the sphincter muscle **(Fig. 9.3.3B)**

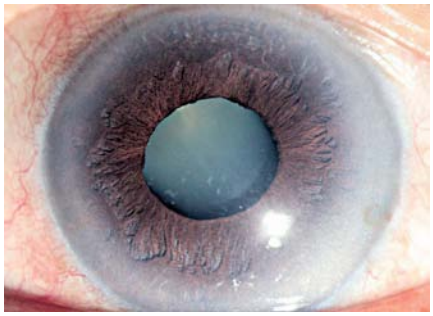


Fig. 9.3.1A: Hypermetropic eye—shallow AC

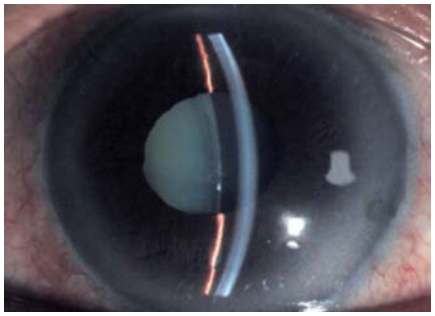


Fig. 9.3.1B: Hypermetropic eye—shallow AC

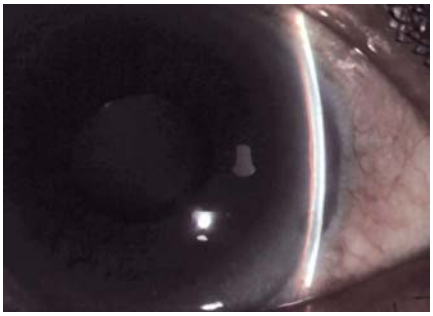


Fig. 9.3.1C: Extremely narrow angle detected by van Herick's method

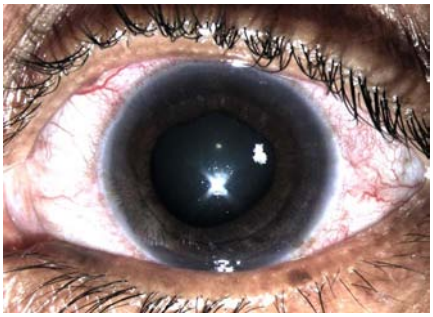


Fig. 9.3.2: PACG - acute attack—lid edema and ciliary congestion

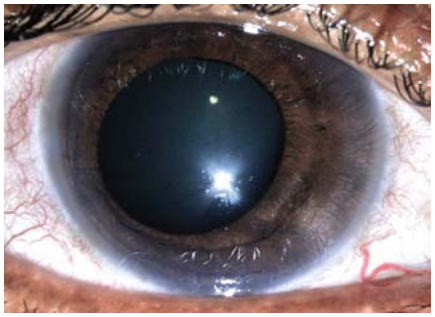


Fig. 9.3.3A: PACG—acute attack—mid-dilated and oval pupil

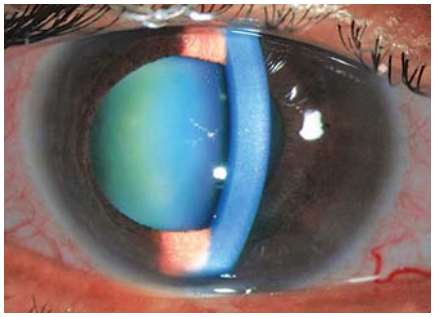


Fig. 9.3.3B: PACG—acute attack—sphinteric atrophy at 12 o'clock

- *Glaucomflecken* **(Fig. 9.3.4)** are small grayish-white anterior subcapsular opacities occur in the pupillary zone—diagnostic of previous attack of angle-closure glaucoma
- Peripheral anterior synechiae develop **(Fig. 9.3.5)**, mostly in the upper part of the angle, but gradually spread around the whole circumference.

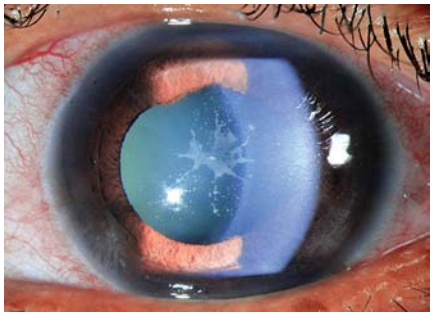


Fig. 9.3.4: PACG—acute attack—glaucomflecken

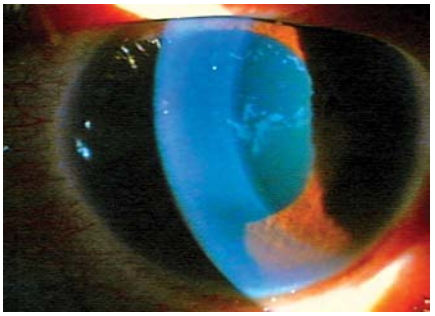


Fig. 9.3.5: PACG—PAS formation

Chronic Congestive Attack

- Angle becomes slowly and progressively closed
- Creeping angle closure **(Fig. 9.3.6)**
- Variable degree of cupping of the disk.

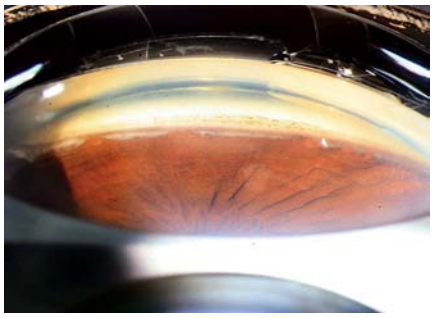


Fig. 9.3.6: PACG—creeping angle closure

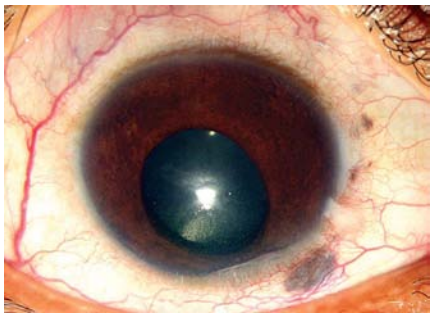


Fig. 9.3.7: Absolute glaucoma

Stage of Absolute Glaucoma

- Reddish-blue zone surrounding the limbus, due to dilated anterior ciliary vein
- Cloudy cornea, may be bullous changes
- Very shallow anterior chamber
- Dilated and grayish pupil **(Fig. 9.3.7)**
- Iris atrophy with ectropion uveae **(Fig. 9.3.8)**

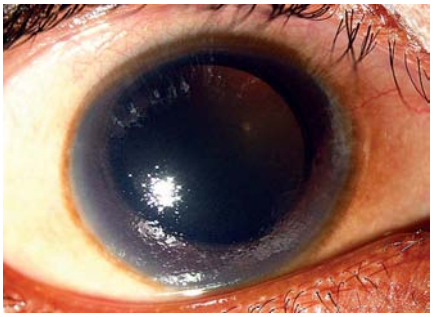


Fig. 9.3.8: Absolute glaucoma—ectropion uveae and corneal edema

- Treatment of ACG is always surgical
  - Pilocarpine (2%) eye drop, oral acetazolamide, I/V mannitol, etc. initially,
  - Followed by YAG laser peripheral iridotomy **(Figs 9.3.9A and B)** or surgical PI in initial stage
  - If the PAS is more—trabeculectomy is the treatment of choice
  - Of fellow eye YAG laser PI.

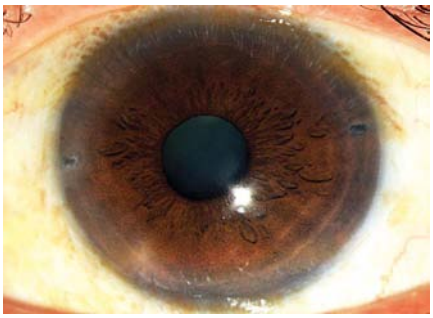


Fig. 9.3.9A: YAG PI openings at 9.00 and 2.30 o'clock

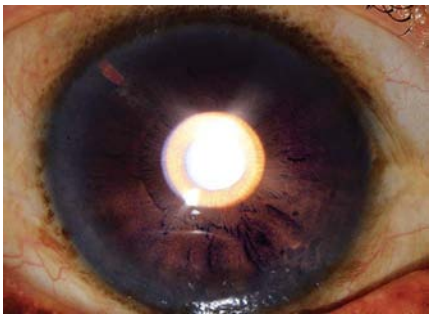


Fig. 9.3.9B: YAG PI opening at 10 o'clock position

Slit Lamp Grading of Angle (van Herick)

- Used with a fair accuracy
- Useful when a gonioscopy is difficult to perform
- Depth of the 'peripheral anterior chamber' (PAC) is estimated by comparing it to the adjacent 'corneal thickness' (CT) 1 mm inside the limbus
- *Four grades:*
  - *Grade 4*: PAC = 1CT = wide open angle **(Fig. 9.4.1)**
  - *Grade 3*: PAC = 1/4th to ½ CT = open angle **(Fig. 9.4.2)**
  - *Grade 2*: PAC = 1/4th CT = moderately narrow **(Fig. 9.4.3)**
  - *Grade 1*: PAC < 1/4th CT = extremely narrow **(Fig. 9.4.4)**.

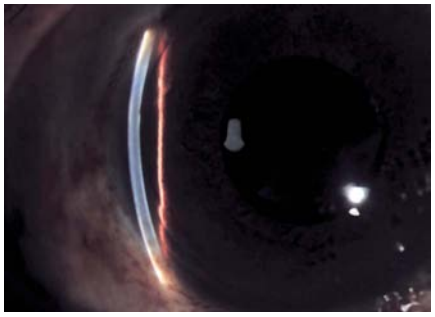


Fig. 9.4.1: van Herick grading—Grade 4

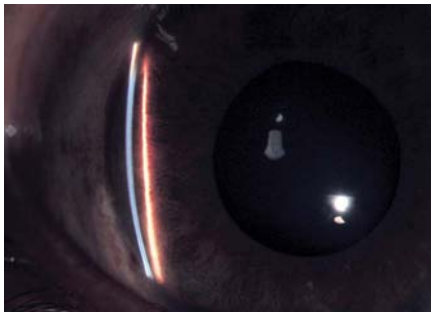


Fig. 9.4.2: van Herick grading—Grade 3

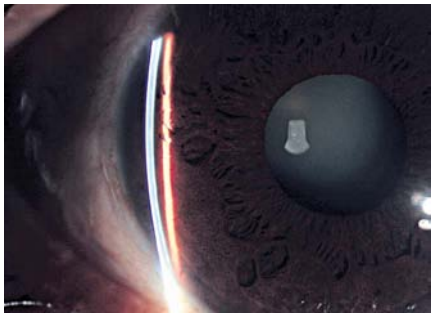


Fig. 9.4.3: van Herick grading—Grade 2

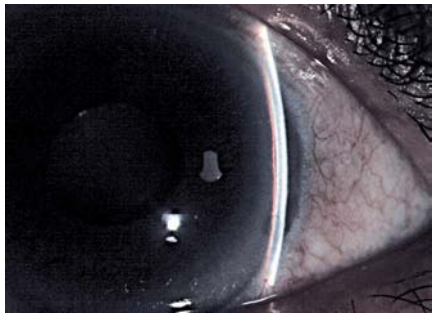


Fig. 9.4.4: van Herick grading—Grade 1

Primary Open Angle Glaucoma

- Chronic, bilateral slowly progressive glaucoma with typical cupping of the optic disk and characteristic field changes
- Usually older people, 40 years and above
- Positive family history in 15–20% cases
- Some people are corticosteroids responder
- Eyeball otherwise looks normal
- *Classical triad:* Raised IOP, cupping of the disk **(Figs 9.5.1A and B)** and classical field defects

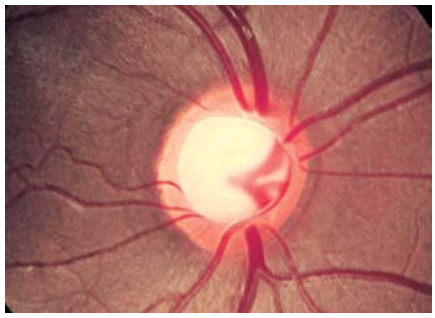


Fig. 9.5.1A: Glaucoma cupping—RE

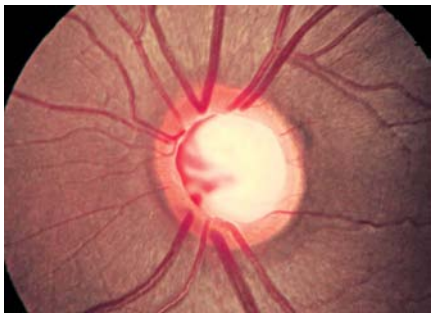


Fig. 9.5.1B: Glaucoma cupping—LE

- *Treatment: Primarily medical*
  - *Medical:* Pilocarpine, beta blockers, carbonic anhydrase inhibitor, brimonidine, bimatoprost, latanoprost, travoprost, etc. used alone or in combination
  - *Laser:* Argon laser trabeculoplasty, diode-cyclophotocoagulation
  - *Surgical:* Trabeculectomy with or without wound modulators (like, MMC or 5 FU), nonpenetrating deep trabeculoplasty, etc.
  - *Combination therapy.*

Cupping of the Optic Disk

- *Asymmetry of the cupping* (C:D ratio difference more than 0.2) **(Figs 9.6.1A and B)**
- Cupping starts as focal enlargement *(notching)* at the inferotemporal quadrant **(Fig. 9.6.2)**. It may be superior notching or both **(Figs 9.6.3 and 9.6.4).** Inferior notching presents as upper arcuate scotoma and superior notching as inferior arcuate scotoma. When the notchings are present both upper and lower—there will be double arcuate scotoma
- *Bayoneting sign*: Double angulations of the blood vessels, pass sharply backwards and then turn along the steep wall of the excavation before angling again onto the floor of the cup **(Fig. 9.6.5)**
- *Thinning* of the neuroretinal rim **(Figs 9.6.6 and 9.6.7)**
- *Nasal shifting* of retinal blood vessels at the disk **(Figs 9.6.8A and B)**
- *Over passing phenomena* of the disk blood vessels **(Fig. 9.6.9)**
- *Splinter hemorrhages* at the disk margin **(Fig. 9.6.10)**
- *Baring of circumlinear blood vessels* may be seen at the disk margin **(Fig. 9.6.11)**
- *Near total cupping* **(Figs 9.6.12A and B)**

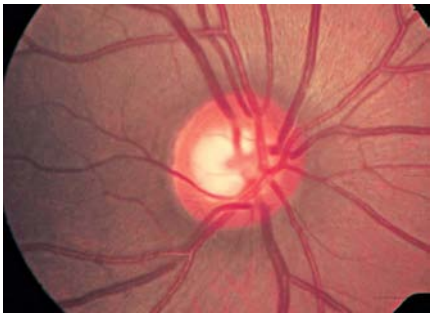


Fig. 9.6.1A: Glaucoma cupping—asymmetry

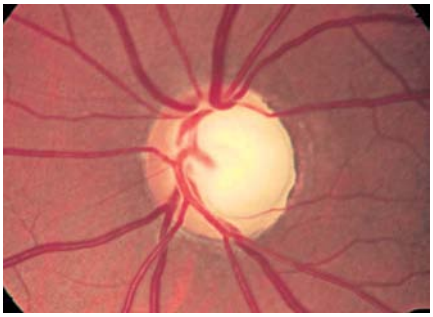


Fig. 9.6.1B: Glaucoma cupping—asymmetry—LE

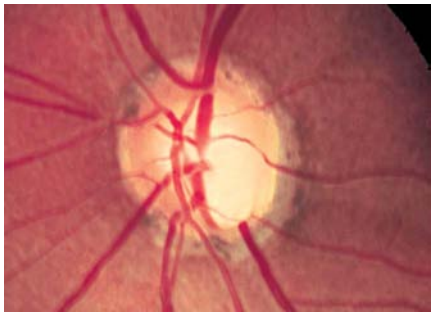


Fig. 9.6.2: Glaucoma cupping—inferior notching

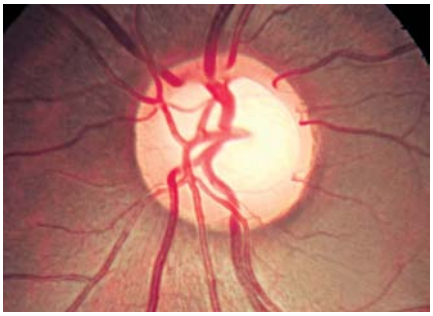


Fig. 9.6.3: Glaucoma cupping—superior notching

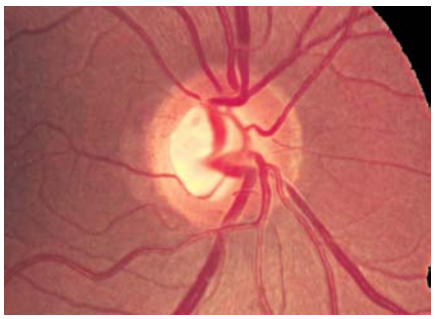


Fig. 9.6.4A: Glaucoma cupping—superior and inferior notching

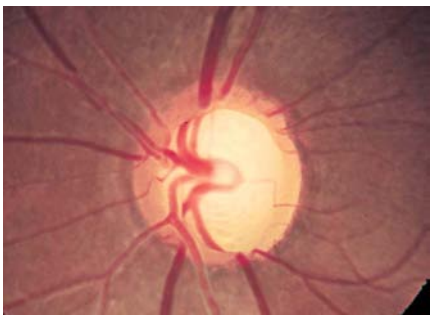


Fig. 9.6.4B: Glaucoma cupping—superior and inferior notching

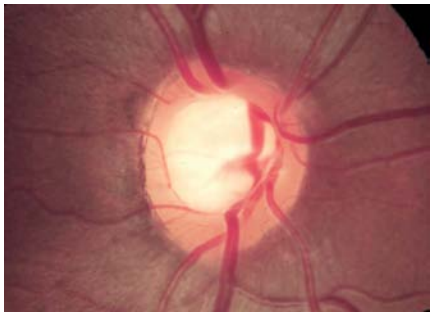


Fig. 9.6.5: Cupping—bayoneting sign

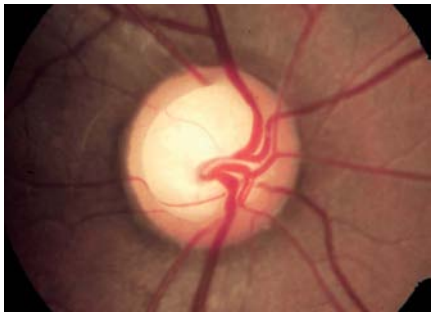


Fig. 9.6.6: Glaucoma cupping—neuro-retinal rim thinning

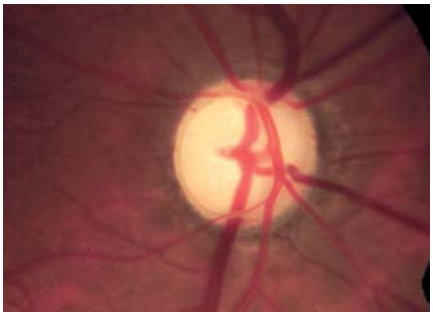


Fig. 9.6.7: Glaucoma cupping—neuro-retinal rim thinning

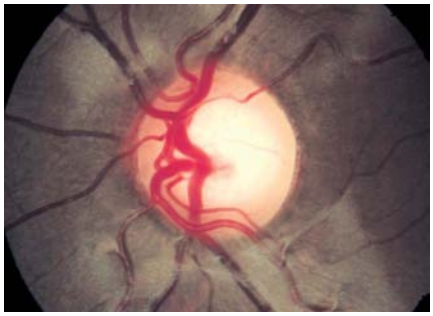


Fig. 9.6.8A: Glaucoma cupping—nasal shifting of blood vessel

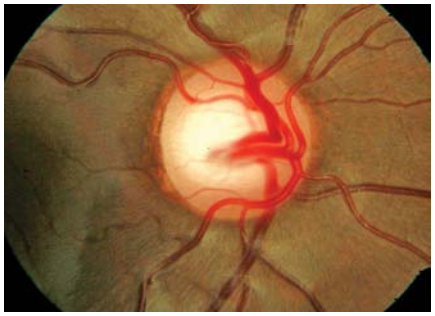


Fig. 9.6.8B: Glaucoma cupping—nasal shifting of blood vessel

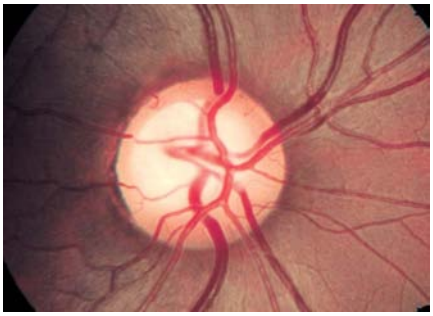


Fig. 9.6.9: Glaucoma cupping—overpassing phenomena

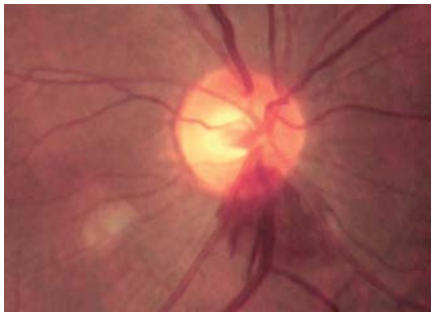


Fig. 9.6.10: Glaucoma cupping—splinter hemorrhage

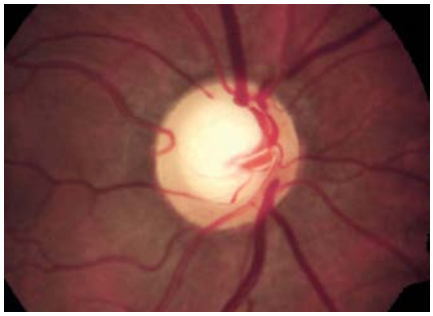


Fig. 9.6.11: Glaucoma cupping—baring of blood vessels

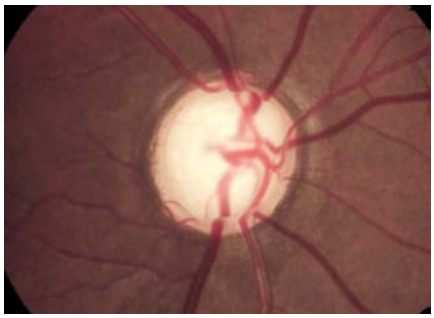


Fig. 9.6.12A: Glaucoma cupping—near total cupping—RE

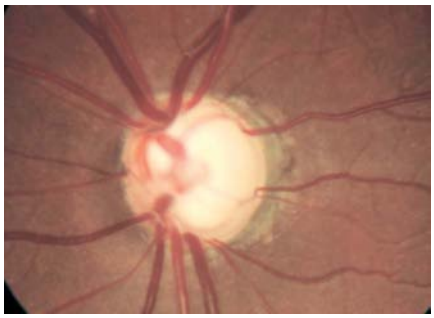


Fig. 9.6.12B: Glaucoma cupping—near total cupping—LE

- *Total cupping:* It appears as a white disk with loss of all neural rim, and bending of all retinal vessels at the margin of the disk—called *'bean-pot cupping'* **(Figs 9.6.13A and B)**
- *Total pallor of the disk*, as there is glaucomatous optic atrophy **(Figs 9.6.14 and 9.6.15)**
- Visible up to the margin of the disk *'laminar dot sign'* **(Fig. 9.6.16)**.

Absolute Glaucoma in POAG

- In untreated cases, the eye eventually becomes an absolute eye with no perception of light
- Reddish-blue zone surrounding the limbus, due to dilated anterior ciliary vein
- Cloudy cornea, may be bullous changes **(Fig. 9.6.17)**
- Normal anterior chamber depth
- Dilated and grayish pupil
- Fundus is not visible at this stage; an USG B-scan may detect deep glaucoma cupping
- *Treatment:* Antiglaucoma medication followed by cyclocryopexy or cyclophotocoagulation.

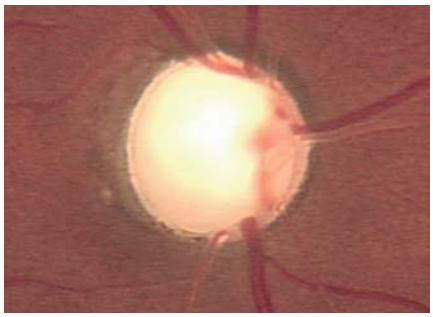


Fig. 9.6.13A: Glaucoma total cupping—bean-pot cupping—RE

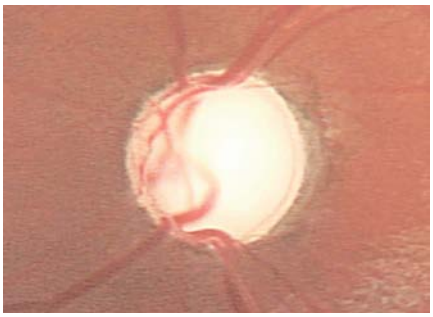


Fig. 9.6.13B: Glaucoma total cupping—bean-pot cupping—LE

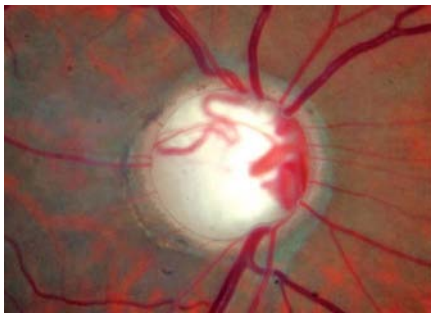


Fig. 9.6.14: Glaucoma cupping—total pallor

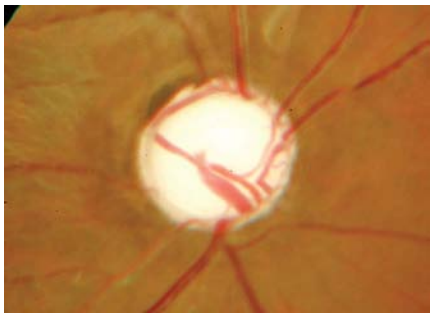


Fig. 9.6.15: Glaucoma total cupping—optic atrophy

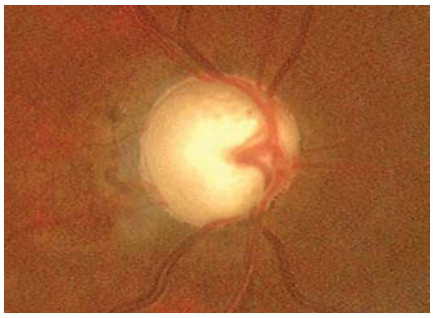


Fig. 9.6.16: Glaucoma total cupping—optic atrophy with laminar dot sign

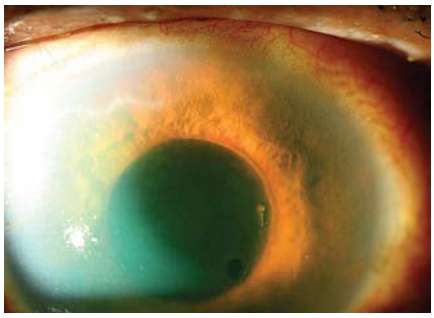


Fig. 9.6.17: Absolute glaucoma in POAG

Secondary Glaucomas

Pseudoexfoliation Glaucoma

- Secondary glaucoma with pseudoexfoliation syndrome
- Deposition of fibrillar basement membrane like material blocking the trabecular meshwork
- White exfoliative materials on the anterior lens capsule **(Figs 9.7.1A and B)**, pupillary margin and angle of the anterior chamber **(Fig. 9.7.2)**
- *Treatment:* Almost same as PAOG.

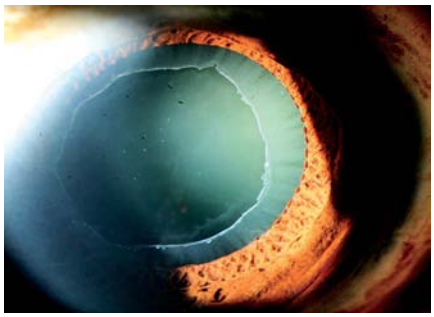


Fig. 9.7.1A: Pseudoexfoliation of lens capsule

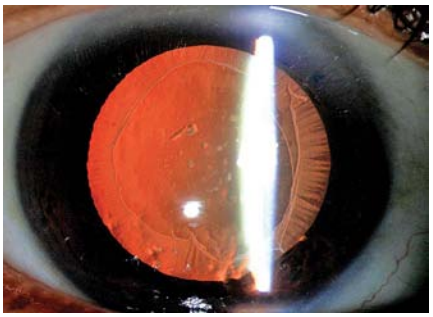


Fig. 9.7.1B: Pseudoexfoliation of lens capsule

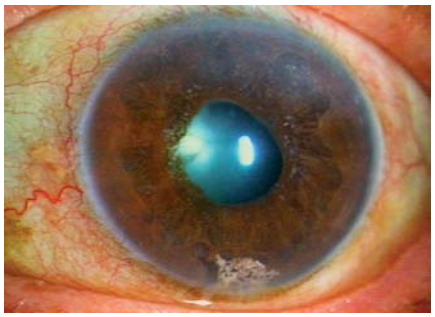


Fig. 9.7.2: Pseudoexfoliative glaucoma

Phacomorphic Glaucoma

- A swollen intumescent cataract **(Figs 9.8.1A and B)** may cause secondary angle-closure glaucoma
- Shallow anterior chamber and sometime with frank corneal edema **(Fig. 9.8.2)**
- Clinical picture is similar to angle-closure glaucoma
- In case of blunt trauma, there may be swollen lens which may obstruct the angle **(Figs 9.8.3A and B)**
- *Treatment:* Urgent reduction of IOP followed by cataract surgery with or without trabeculectomy.

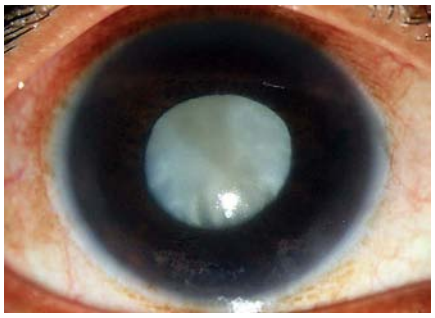


Fig. 9.8.1A: Phacomorphic glaucoma—with corneal edema

Fig. 9.8.1B: Phacomorphic glaucoma—with corneal edema

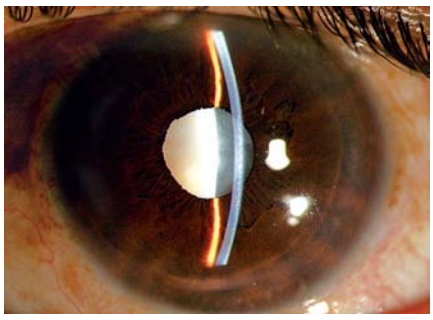


Fig. 9.8.2: Phacomorphic changes—intumescent cataract with angle closure

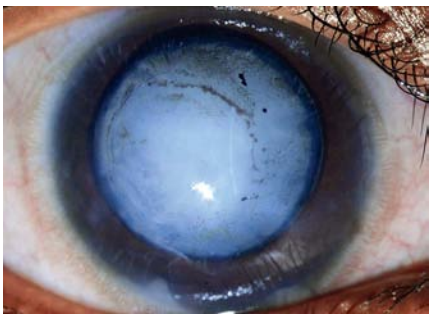


Fig. 9.8.3A: Lens-induced glaucoma—swollen lens after a blunt trauma

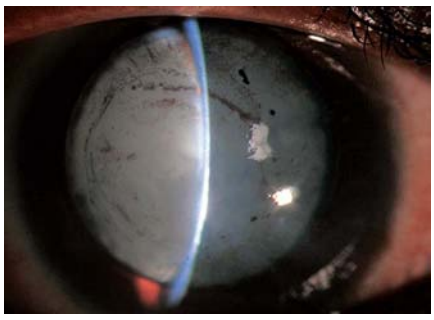


Fig. 9.8.3B: Lens-induced glaucoma following trauma with flat AC

Phacolytic Glaucoma

- Secondary open-angle glaucoma due to micro-leak of lens capsule with leakage of turbid milky fluid cortical matter in a hypermature Morgagnian cataract
- IOP-rise is due to obstruction of trabecular meshwork by macrophages which ingest the lens protein
- Deep or normal anterior chamber with turbid aqueous **(Figs 9.9.1A and B)**
- *Treatment:* Urgent reduction of IOP medically followed by cataract surgery with or without trabeculectomy.

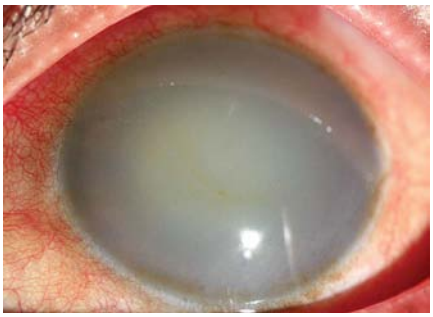


Fig. 9.9.1A: Phacolytic glaucoma—milky cortex in AC

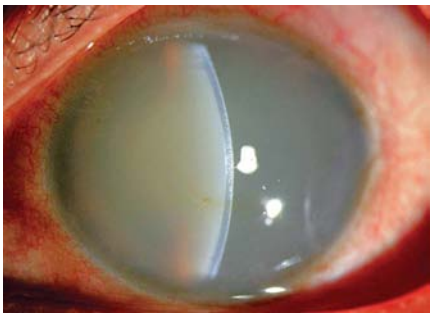


Fig. 9.9.1B: Phacolytic glaucoma—deep AC with milky cortex

Lens-Particle Glaucoma

- Typically following ECCE, or after penetrating injury of the lens capsule **(Fig. 9.10.1)**
- Due to direct obstruction of trabecular meshwork by the lens particles or fluffy lens cortex and associated trabeculitis
- *Treatment*: Surgical removal of lens materials after controlling the IOP medically.

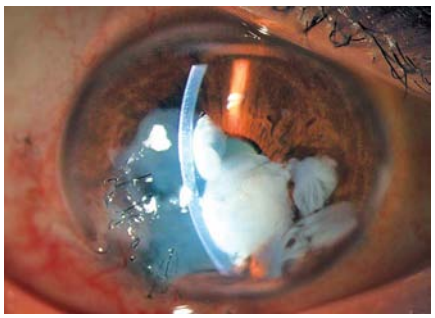


Fig. 9.10.1: Lens particle glaucoma following penetrating injury

Associated with Ectopia Lentis/Subluxation or Dislocation of Lens

- Secondary glaucoma may develop in ectopia lentis or with subluxated or dislocated lens **(Figs 9.11.1 and 9.11.2)**
- Caused by pupillary block or by developing PAS
- *Inverse pupillary block glaucoma* may occur in microspherophakia and a mydriatic is essential to break the pupillary block **(Figs 9.11.3 and 9.11.4)**

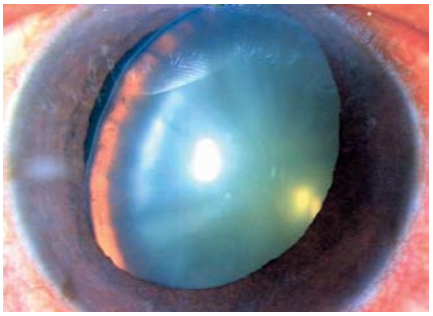


Fig. 9.11.1A: Glaucoma with subluxation

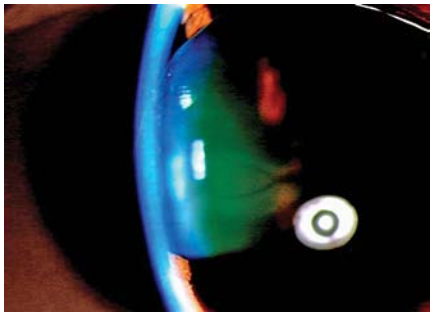


Fig. 9.11.1B: Glaucoma with subluxation—pupillary block

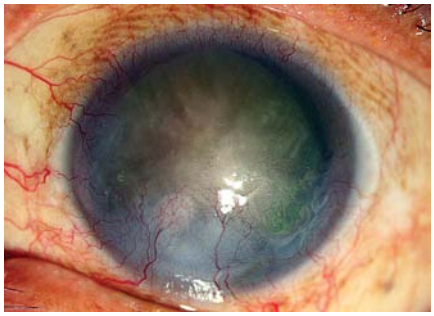


Fig. 9.11.2: Lens-induced glaucoma—anterior dislocation of hard cataract—absolute glaucome

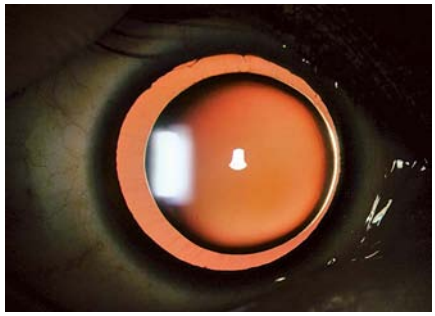


Fig. 9.11.3A: Microspherophakia—probability of inverse glaucoma

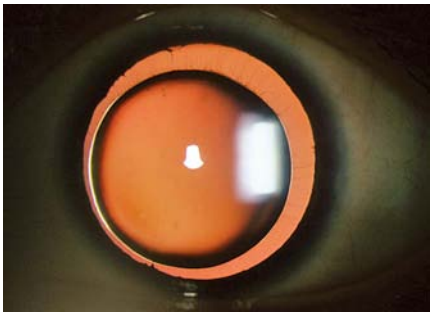


Fig. 9.11.3B: Microspherophakia—probability of inverse glaucoma

- *Treatment:* Extraction of lens with anterior vitrectomy with peripheral iridectomy or trabeculectomy.

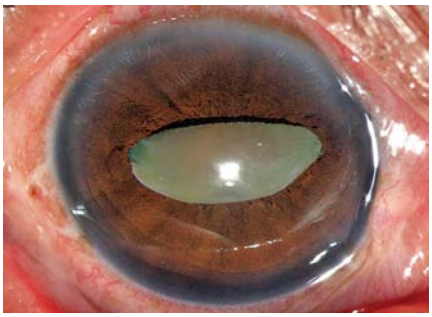


Fig. 9.11.4A: Spherophakia—inverse glaucoma

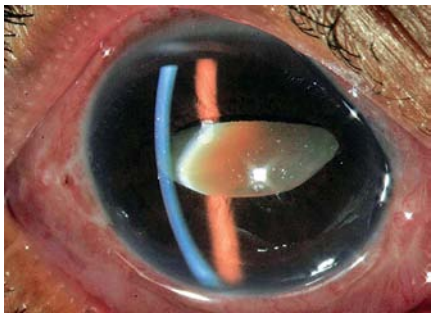


Fig. 9.11.4B: Spherophakia—inverse glaucoma

Pigmentary Glaucoma

- Pigment dispersion occurs throughout the anterior segment **(Fig. 9.12.1)**
- Deposition of pigment on the corneal endothelium in a vertical line called *Krunkenberg's spindle* **(Figs 9.12.2A and B)**
- Loss of pigments from the iris—with positive iris transillumination **(Fig. 9.12.3)**
- *Zentmayer's line:* Seen near the equator of the crystalline lens **(Fig. 9.12.4)**
- Accumulation of pigment along the Schwalbe's line, especially inferiorly, as a dark line—*Sampaolesi's line* **(Fig. 9.12.5)**
- *Treatment:* Same as POAG.

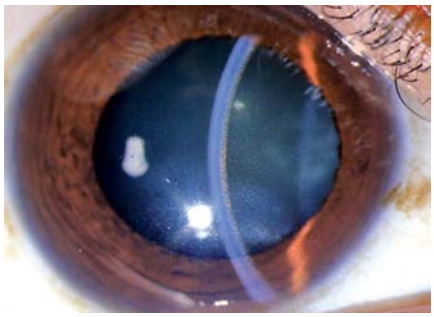


Fig. 9.12.1: Pigmentary glaucoma—Krukenberg's spindle pigment

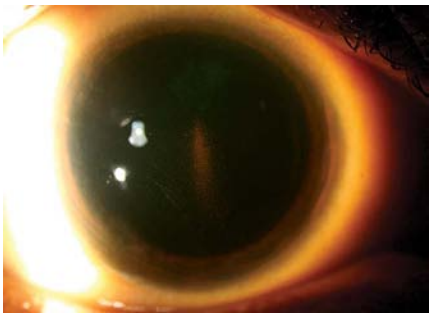


Fig. 9.12.2A: Pigmentary glaucoma—Krukenberg's spindle—RE

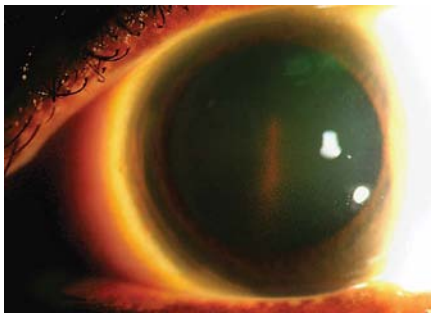


Fig. 9.12.2B: Pigmentary glaucoma—Krukenberg's spindle—LE

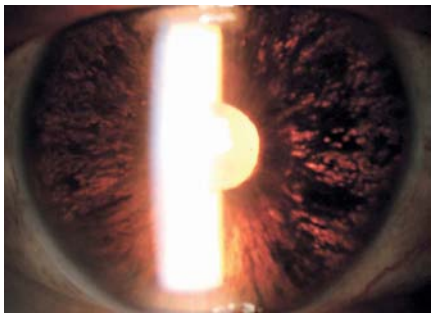


Fig. 9.12.3: Pigmentary glaucoma—iris transillumination

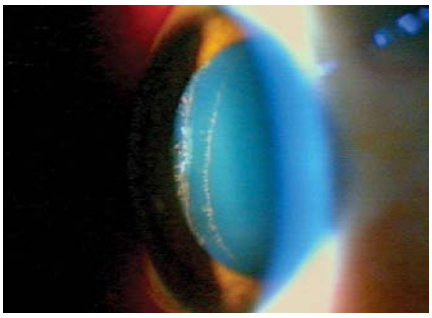


Fig. 9.12.4: Zentmayer's line

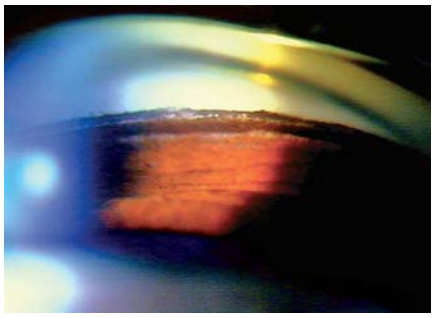


Fig. 9.12.5: Pigmentary glaucoma—Sampaolesi's line

Inflammatory Secondary Glaucoma

- Associated with inflammation of other structures
- *Causes:*
  - Iridocyclitis **(Figs 9.13.1 and 9.13.2)**
  - Posner-Schlossmann syndrome (Glaucomatocyclitic crisis) **(Fig. 9.13.3)**
  - Corneal ulcer with hypopyon
  - Adherent leucoma
- *Treatment:* Medical and if necessary surgical and treatment of the cause.

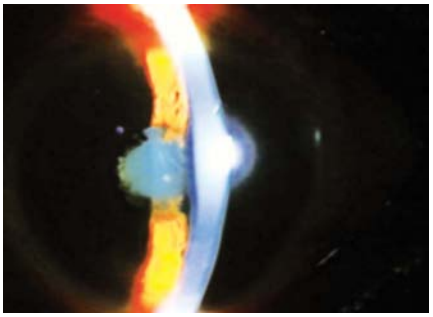


Fig. 9.13.1: Glaucoma with iridocyclitis—iris bombe

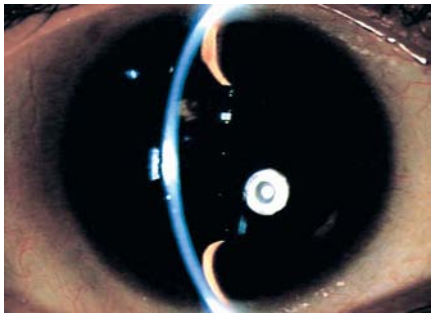


Fig. 9.13.2A: Glaucoma with iridocyclitis—iris bombe

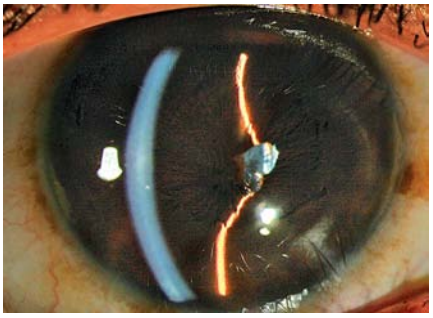


Fig. 9.13.2B: Pupillary block by 360-degree synechia in iridocyclitis

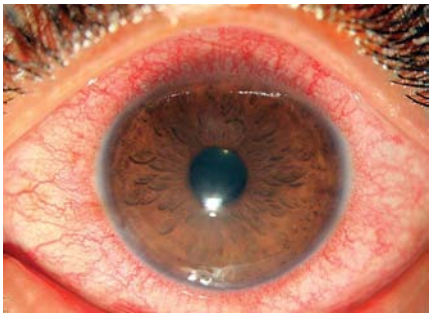


Fig. 9.13.3: Glaucoma in iridocyclitis—Posner-Schlossmann syndrome

Glaucoma Associated with Trauma

- *Blunt injury:* More than one mechanism are involved:
  - *Hyphema* **(Figs 9.14.1A and B)**; in long-standing cases, there may be blood staining of the cornea **(Fig. 9.14.2)**
  - Subluxation or dislocation of lens
  - Angle recession glaucoma **(Fig. 9.14.3A and B)**
- Penetrating injury
- *Chemical injury:* Both by acid or alkali burn
- *Treatment:* Antiglaucoma medication, but no miotics.

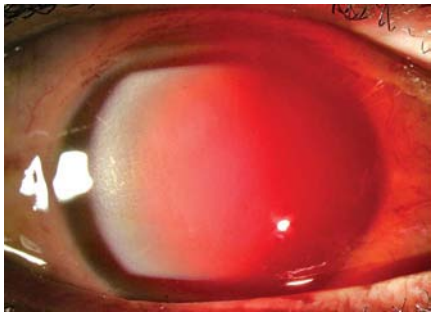


Fig. 9.14.1A: Traumatic hyphema—glaucoma

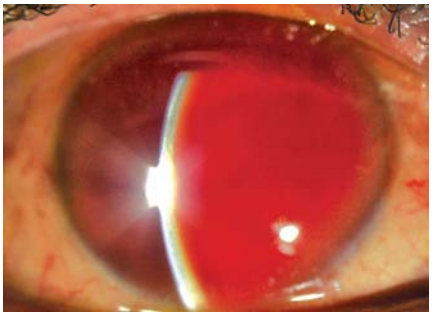


Fig. 9.14.1B: Traumatic hyphema—glaucoma

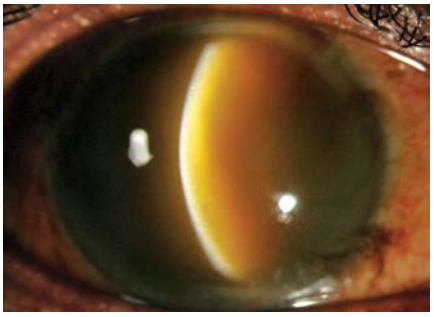


Fig. 9.14.2: Traumatic hyphema—glaucoma—blood staining of the cornea

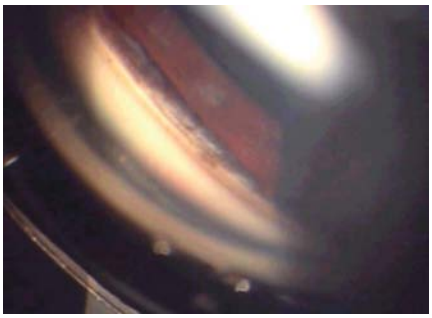


Fig. 9.14.3A: Angle recession glaucoma

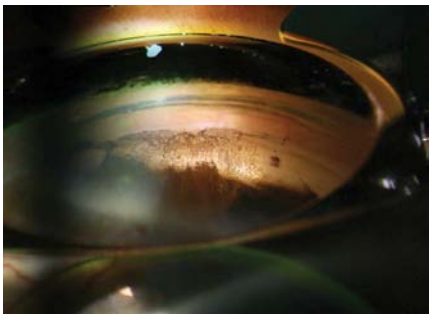


Fig. 9.14.3B: Angle recession glaucoma

Glaucoma Following Intraocular Surgery

- *Pseudophakic:* Hyphema, iridocyclitis, pupillary block or steroid induced
- *Aphakic:* Pupillary block, hyphema, vitreous in AC, iris cyst **(Fig. 9.15.1)**, etc.
- *Post-keratoplasty:* Tight suturing **(Fig. 9.15.2)**, steroid-induced glaucoma and PAS
- *After Descemet stripping endothelial keratoplasty (DSEK):* By air-induced pupillary block **(Fig. 9.15.3)**; by PAS **(Fig. 9.15.4)**; after TASS **(Figs 9.15.5A and B);** and also steroid-induced
- Postvitreoretinal surgery
- *Treatment:* Antiglaucoma medication and treatment of the cause.

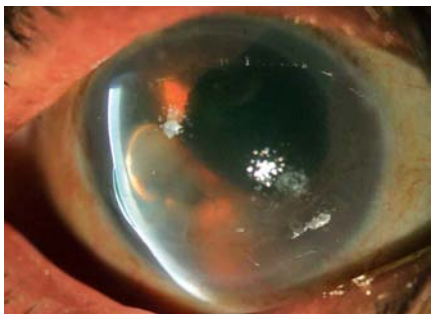


Fig. 9.15.1: Glaucoma—iris cyst

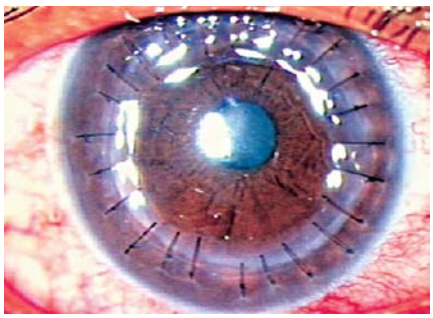


Fig. 9.15.2: Secondary glaucoma—tight and extensive suturing in PK

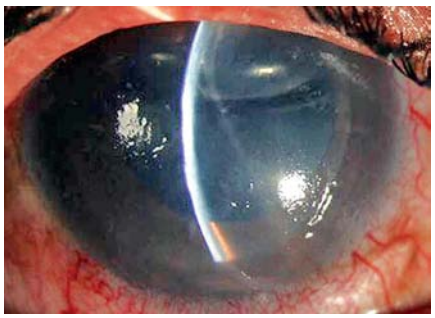


Fig. 9.15.3: Pupillary block glaucoma by air following DSEK

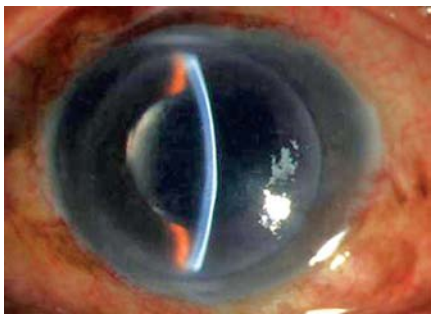


Fig. 9.15.4: Secondary angle closure following DSEK

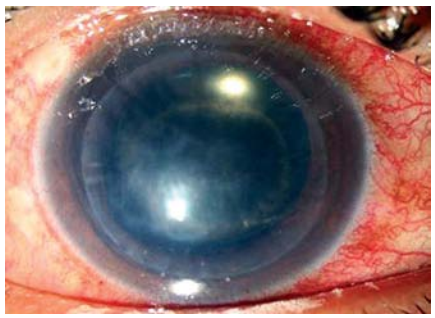


Fig. 9.15.5A: Secondary glaucoma due to TASS

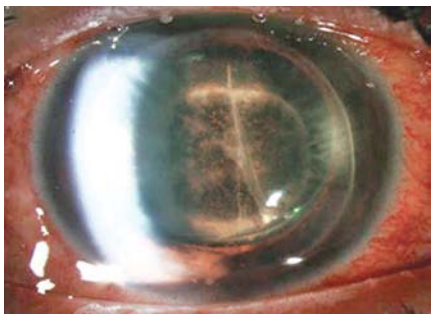


Fig. 9.15.5B: Secondary glaucoma due to TASS

Malignant (Ciliary Block) Glaucoma

- Total shallowing (both central and peripheral) of anterior chamber **(Fig. 9.16.1)**
- Poor response to conventional antiglaucoma medication
- *Two types:*
  - Cilio-lenticular block: After trabeculectomy **(Figs 9.16.2A and B)**
  - Cilio-vitreal block: After cataract operation (mainly after ICCE)
- Treatment:
  - 4% atropine eye drop
  - USG-localization of vitreous pockets
  - Immediate lens extraction in phakic cases
  - YAG laser hyaloidotomy in pseudophakic cases
  - Vitrectomy in aphakic cases.

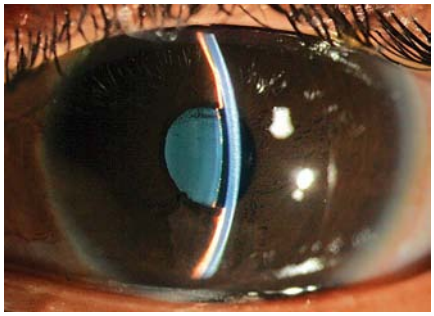


Fig. 9.16.1: Malignant glaucoma—very shallow (flat) AC following trabeculectomy

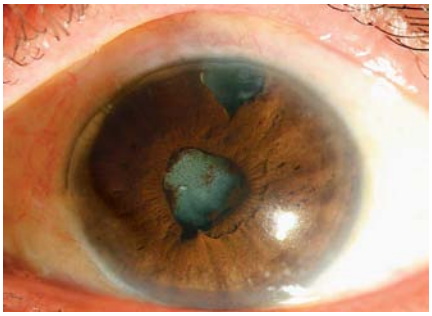


Fig. 9.16.2A: Malignant glaucoma—following tabeculectony with 360-degree and cataract synechiae

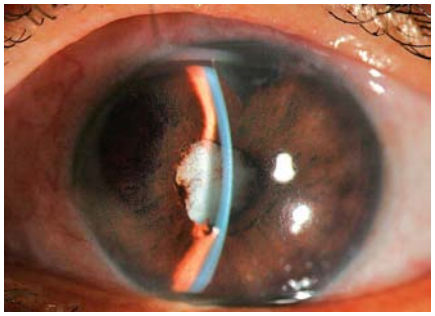


Fig. 9.16.2B: Malignant glaucoma—following trabeculectomy with 360-degree and cataract synechiae

Neovascular Glaucoma

- Secondary glaucoma due to rubeosis iridis and neovascularization of the angle
- *Three stages:*
  - *Preglaucomatous:* Rubeosis iridis **(Fig. 9.17.1)**
  - *Open angle glaucoma:* Due to intense neovascularizaton at the angle
  - *Angle closure glaucoma:* Due to goniosynechia **(Fig. 9.17.2)** and PAS formation **(Figs 9.17.3A and B)**

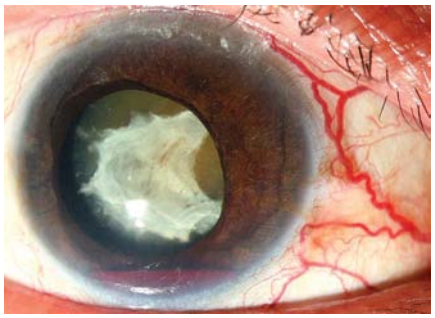


Fig. 9.17.1: Glaucoma—rubeosis iridis

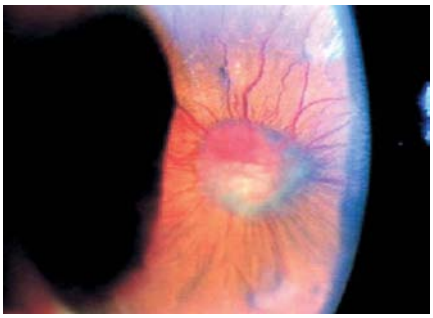


Fig. 9.17.2: Neovascular glaucoma—total posterior synechiae

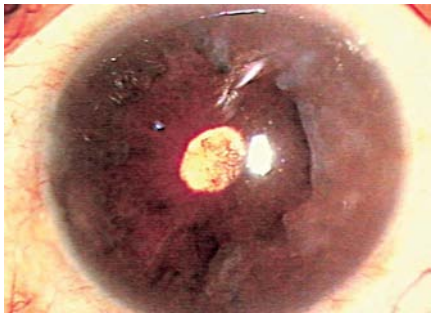


Fig. 9.17.3A: Neovascular glaucoma—total PAS

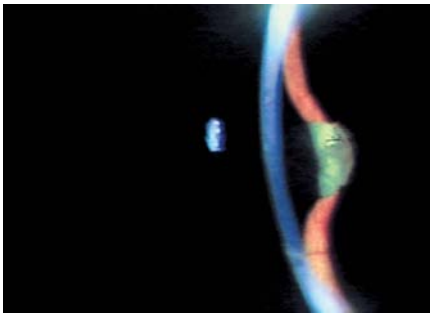


Fig. 9.17.3B: Neovascular glaucoma—total PAS—iris bombe

– Ultimately, total collapse of the anterior chamber with flat AC **(Fig. 9.17.4)**

- *Treatment:* Prophylactic panretinal photocoagulation, anterior retinal cryopexy and cyclophotocoagulation.

Fig. 9.17.4: Glaucoma—rubeosis iridis—total flat AC

Iridocorneal Endothelial (ICE) Syndrome

- Angle is blocked by Descemet's membrane like material and it continues to progress
- Fortunately, only one eye is involved and other eye absolutely remains normal **(Figs 9.18.1A and B)**
- Peripheral angle synechia leading to secondary angle-closure glaucoma
- *Essential iris atrophy:* Iris atrophy, pseudopolycoria, PAS formation glaucoma is most common **(Fig. 9.18.2)**
- *Cogan-Reese syndrome:* Small iris spherules with PAS formation; glaucoma is not so common **(Figs 9.18.3 and 9.18.4)**

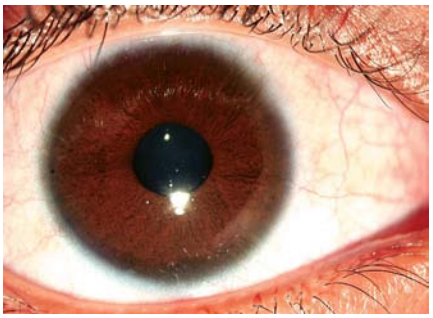


Fig. 9.18.1A: ICE syndrome—essential iris atrophy—normal RE

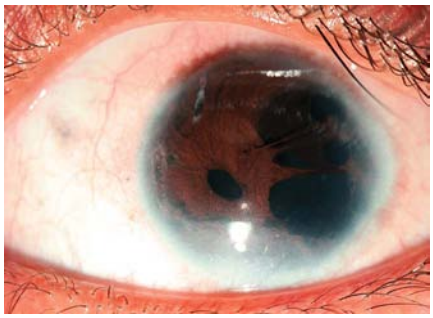


Fig. 9.18.1B: ICE syndrome—essential iris atrophy—affected LE

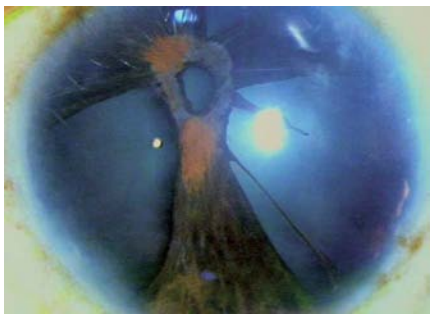


Fig. 9.18.2: ICE syndrome—essential iris atrophy

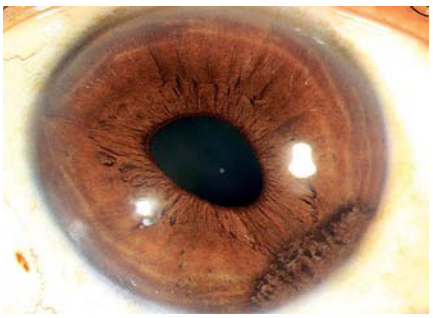


Fig. 9.18.3A: ICE syndrome—Cogan-Reese syndrome

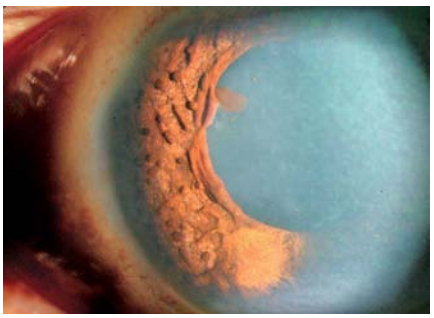


Fig. 9.18.3B: ICE syndrome—Cogan-Reese syndrome

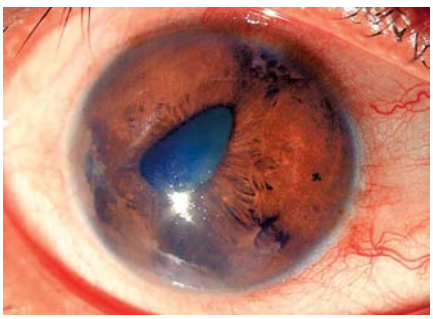


Fig. 9.18.4A: ICE—Cogan-Reese syndrome with corneal decompensation

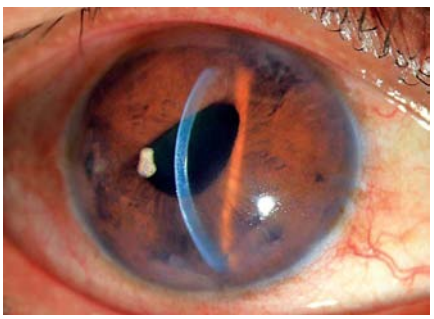


Fig. 9.18.4B: ICE—Cogan-Reese syndrome with corneal decompensation

- *Chandler's syndrome:* Corneal decompensation is rather common; but the glaucoma is least common **(Figs 9.18.5 and 9.18.6)**
- *Treatment:* Trabeculectomy with MMC, but it often fails. In corneal decompensation DSEK is the choice of surgery.

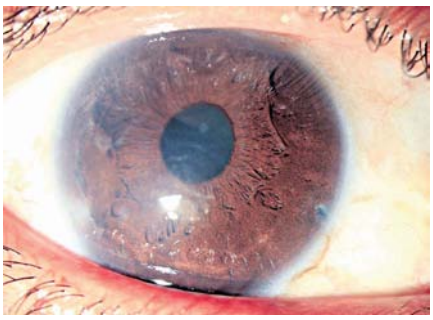


Fig. 9.18.5A: Glaucoma—ICE syndrome—Chandler's syndrome—affected—RE

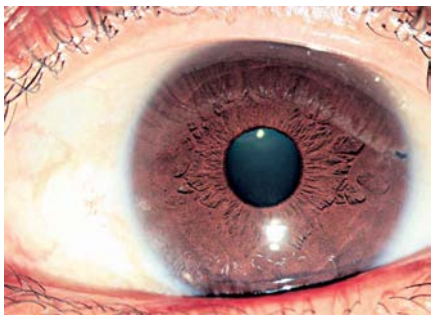


Fig. 9.18.5B: ICE syndrome—normal unaffected—LE

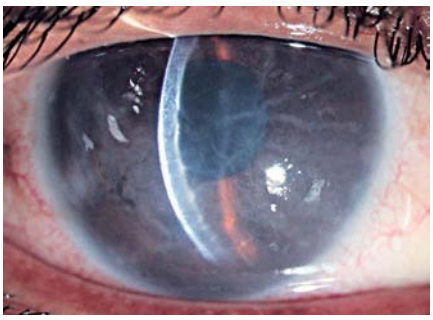


Fig. 9.18.6: Chandler's syndrome—same RE in **Figure 9.18.5A** after 1 year

Other Causes of Secondary Glaucoma

- *Intraocular tumor:* Retinoblastoma, malignant melanoma of the iris or ciliary body tumor; may be both angle-closure and open-angle type
- *Steroid-induced glaucoma:* Open angle type of secondary glaucoma; mostly happens with long-term use of topical corticosteroids; raised IOP and optic disk cupping are prominent findings
- *Hemorrhagic glaucoma:* Mosltly secondary open angle glaucoma
- *Raised episcleral venous pressure:* Superior vena caval syndrome, orbital varices **(Figs 9.19.1 and 9.19.2)** or caroticocavernous thrombosis **(Fig. 9.19.3)**
- *Epidemic dropsy glaucoma:* By consuming adulterated mustard oil; by *mixing Argemone mexicana seeds containing sanguinarine.*

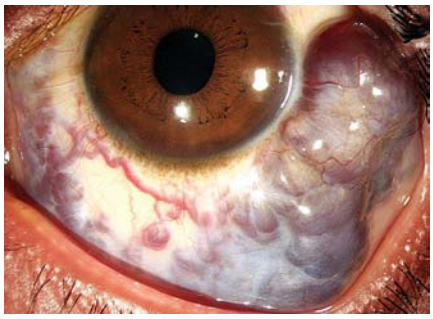


Fig. 9.19.1: Glaucoma in raised episcleral venous pressure

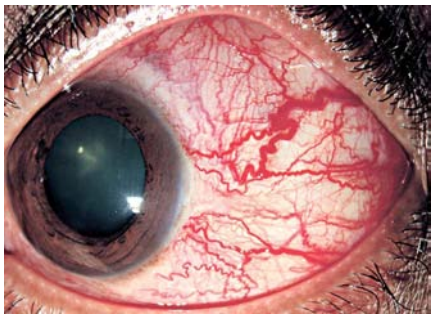


Fig. 9.19.2: Glaucoma in raised venous pressure—orbital varix

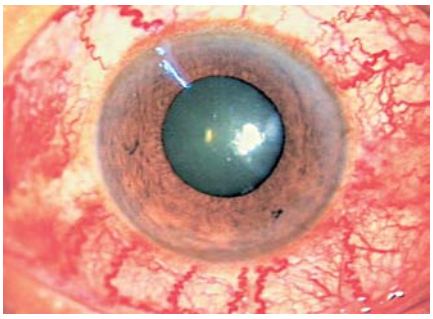


Fig. 9.19.3: Glaucoma in raised episcleral venous pressure—CCF

Filtering Bleb Abnormalities

Normal Functioning Bleb

- Thin-walled, diffuse bleb, relatively avascular and with transconjunctival flow of fluid
- Microcystic spaces seen under slit-lamp is a good sign of bleb function **(Figs 9.20.1A to C)**
- Blebs achieved with the limbus-based conjunctival flap are usually avascular, highly elevated, translucent, thin walled, and cystic
- In contrast, blebs with the fornix-based flap are more vascular, moderately elevated, large in surface area, without well demarcated margins.

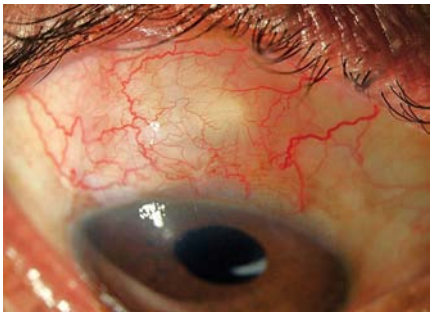


Fig. 9.20.1A: Normal good functioning bleb

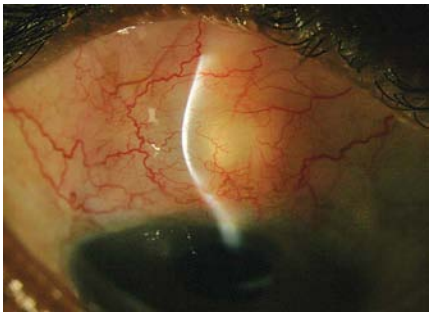


Fig. 9.20.1B: Normal good functioning bleb

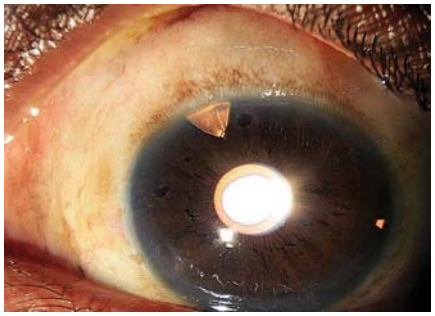


Fig. 9.20.1C: Normal good functioning bleb with PI

Failed/Flat Filtering Bleb

- A flattened bleb with little or no function, in which the scarred conjunctiva firmly adheres to the underlying sclera **(Figs 9.20.2A and B)**
- May be seen in early postoperative periods **(Figs 9.20.2C and D)**.

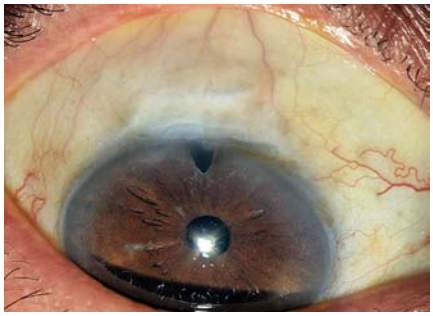


Fig. 9.20.2A: Failed filtering bleb

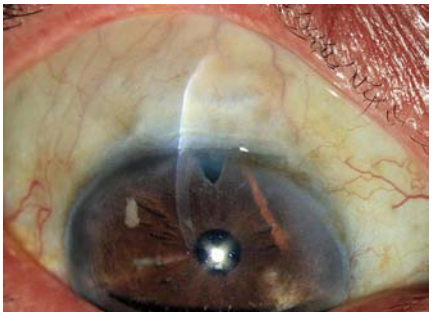


Fig. 9.20.2B: Failed filtering bleb

Large Cystic Filtering Bleb

- Very large well functioning, thin-walled bleb **(Figs 9.20.3A and B)**
- Tendency to have complications, like—rupture or leak
- May be a cause of pseudoptosis **(Figs 9.20.3C and D)**.

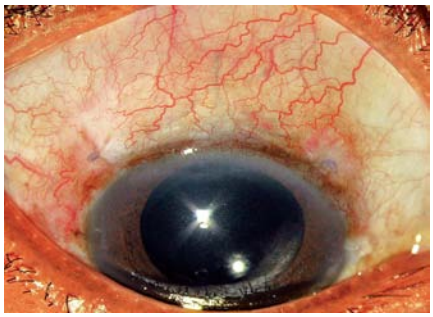


Fig. 9.20.2C: Failed filtering bleb

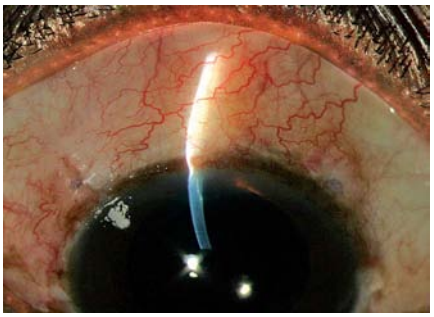


Fig. 9.20.2D: Failed filtering bleb

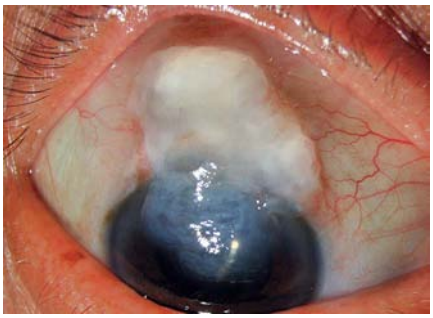


Fig. 9.20.3A: Cystic filtering bleb

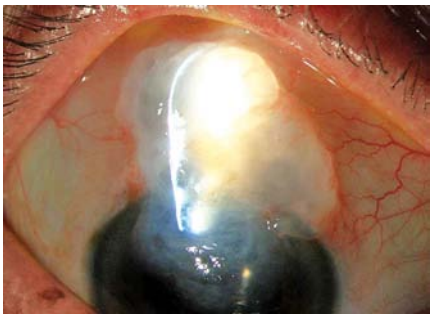


Fig. 9.20.3B: Cystic filtering bleb

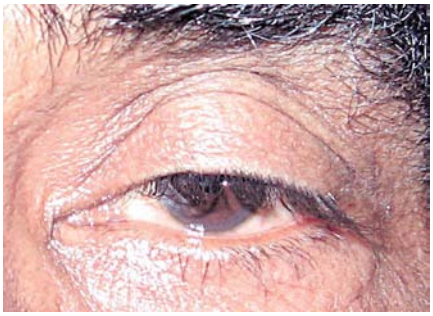


Fig. 9.20.3C: Large cystic and overhanging filtering bleb—pseudoptosis

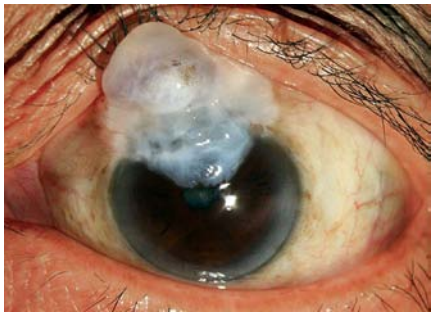


Fig. 9.20.3D: Large cystic and overhanging filtering bleb—pseudoptosis

Multilocular Filtering Bleb

- Multiple large cystic spaces visible by slit-lamp examination **(Figs 9.20.4A and B)**
- Some of these spaces are functioning as filtering blebs to some extent.

Overhanging Filtering Bleb

- Large filtering bleb that overhangs onto the cornea **(Figs 9.20.5A and B)**
- Can be bothersome to the patient, especially when it causes discomfort or even visual impairment
- It may be very thin and cystic **(Figs 9.20.5C and D)**.

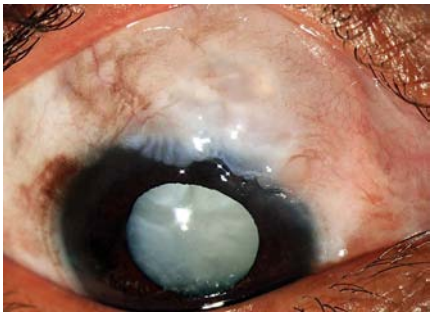


Fig. 9.20.4A: Multilocular filtering bleb

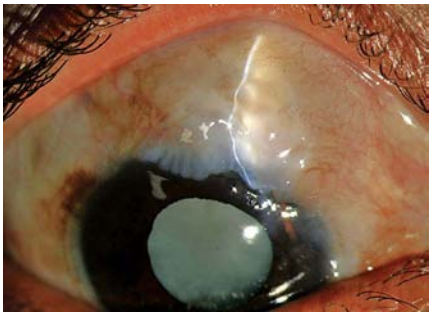


Fig. 9.20.4B: Multilocular filtering bleb

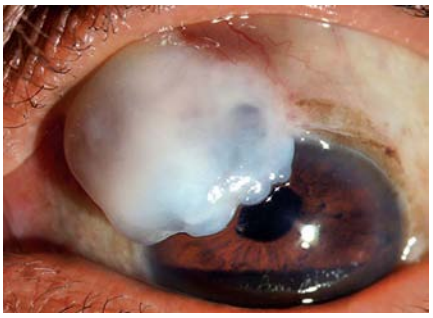


Fig. 9.20.5A: Overhanging filtering bleb—large

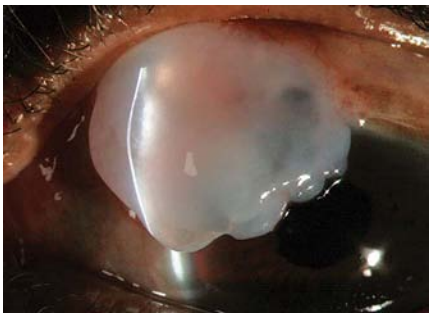


Fig. 9.20.5B: Overhanging filtering bleb—large

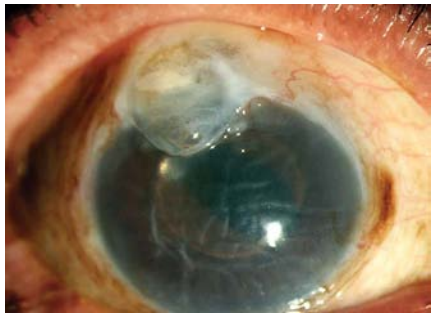


Fig. 9.20.5C: Overhanging cystic filtering bleb

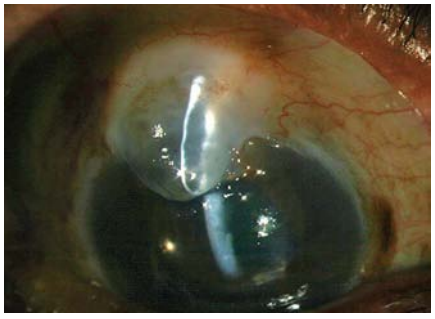


Fig. 9.20.5D: Overhanging very thin cystic filtering bleb

Other Morphological Types of Bleb

- *Avascular filtering bleb:* **(Figs 9.20.6A and B)**
- *Encapsulated filtering bleb:*
  - Also called *Tenon's capsule cyst;* may be small **(Figs 9.20.7A and B)** or may be large **(Figs 9.20.7C and D)**
  - Tense, elevated, smooth dome with a thick wall aqueous-filled cyst **(Figs 9.20.7E and F)**
  - Absence of conjunctival microcysts
- *Thin bleb with large ostium:* **(Fig. 9.20.8)**.

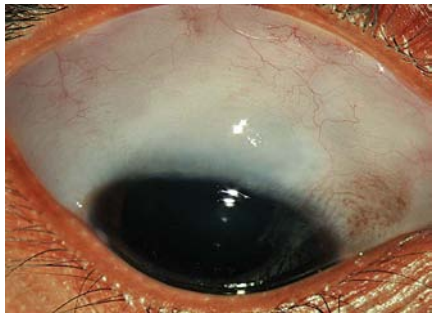


Fig. 9.20.6A: Avascular filtering bleb

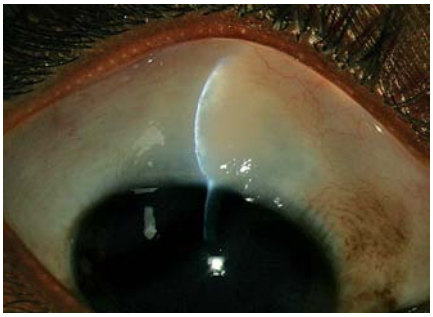


Fig. 9.20.6B: Avascular filtering bleb

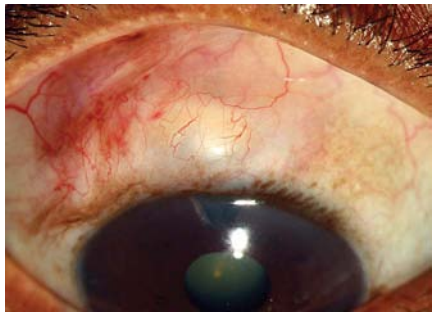


Fig. 9.20.7A: Encysted bleb—small

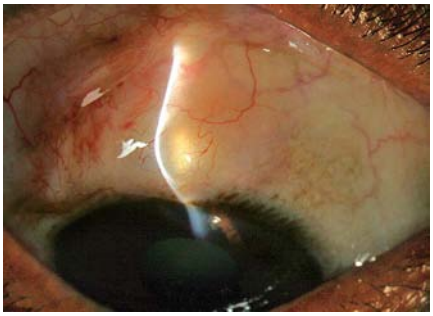


Fig. 9.20.7B: Encysted bleb—small

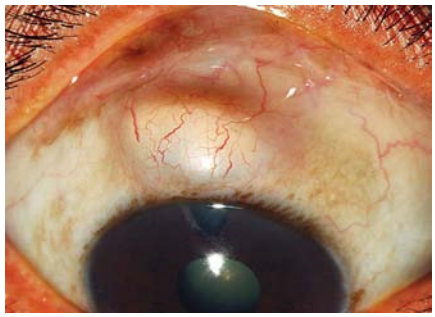


Fig. 9.20.7C: Bleb—encysted nonfunctioning

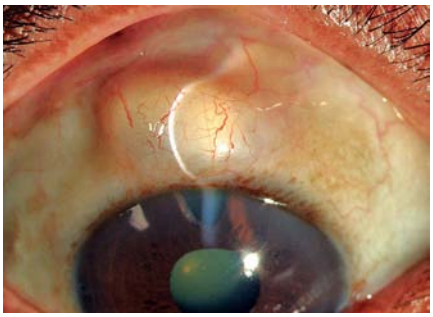


Fig. 9.20.7D: Bleb—encysted nonfunctioning

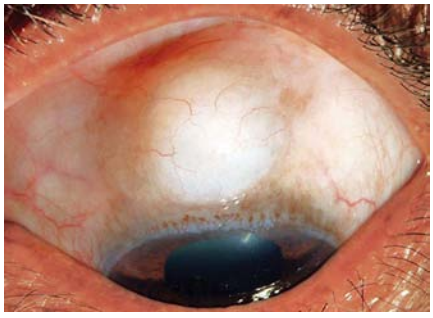


Fig. 9.20.7E: Filtering bleb—Tenon's capsule cyst

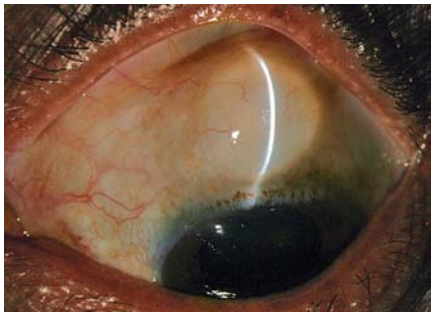


Fig. 9.20.7F: Filtering bleb—Tenon's capsule cyst

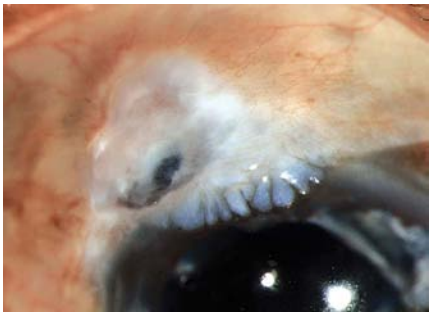


Fig. 9.20.8: Thin bleb with large ostium

Overfiltering Filtering Bleb

- Shallow anterior chamber **(Figs 9.20.9A and B)**
- Corneal folds due to hypotony **(Fig. 9.20.9C)**
- Choroidal folds and hypotonic maculopathy **(Fig. 9.20.9D)**
- *Treatment:* Repair of the bleb to reduce overfiltration.

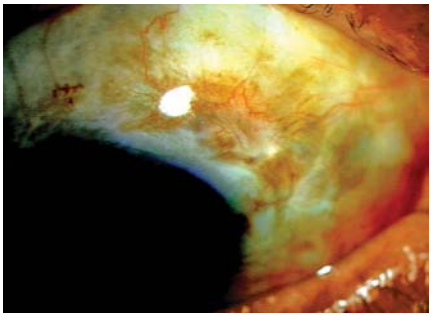


Fig. 9.20.9A: Overfiltering bleb

Fig. 9.20.9B: Overfiltering bleb—shallow AC

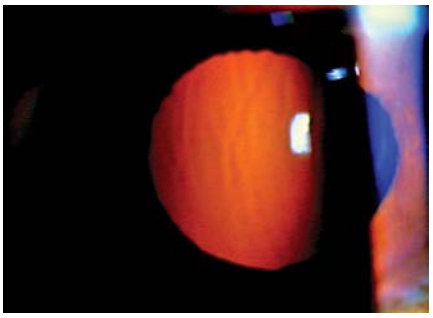


Fig. 9.20.9C: Overfiltering bleb—hypotony—corneal folds

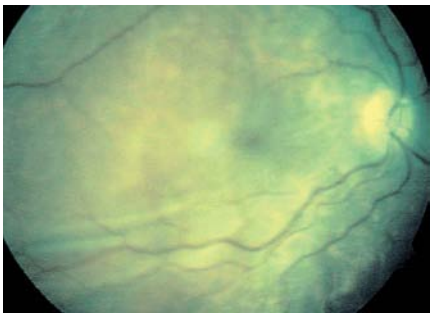


Fig. 9.20.9D: Overfiltering bleb—hypotony—choroidal folds

Leaking Filtering Bleb

- Shallow anterior chamber **(Fig. 9.20.10A)**
- Corneal and fundal changes may be present
- Positive Seidel's test **(Fig. 9.20.10B)**
- *Treatment:* Repair of the bleb with amniotic membrane **(Fig. 9.20.10C)**.

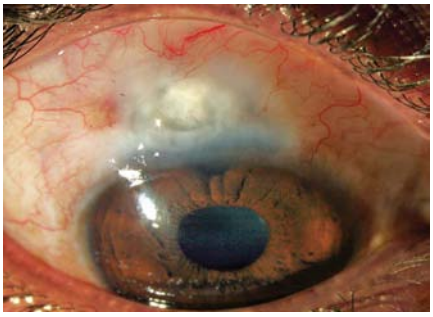


Fig. 9.20.10A: Leaking bleb

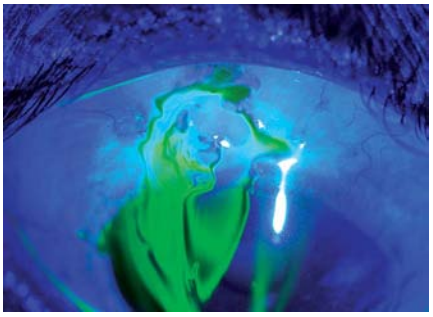


Fig. 9.20.10B: Leaking bleb—positive Seidel's test

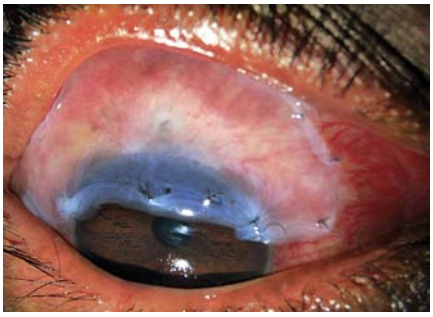


Fig. 9.20.10C: Leaking bleb—repair by AMT (same eye)

Blebitis (Bleb Infection)

- Suppurative infection of bleb mainly by bacteria
- Bleb appears yellowish-white in color with surrounding congestion **(Fig. 9.20.11A)**
- Anterior chamber reaction and hypopyon **(Fig. 9.20.11B)**
- May be associated with frank endophthalmitis **(Figs 9.20.12A and B)**
- *Treatment:* Systemic and topical antibiotics with surgical revision of bleb.

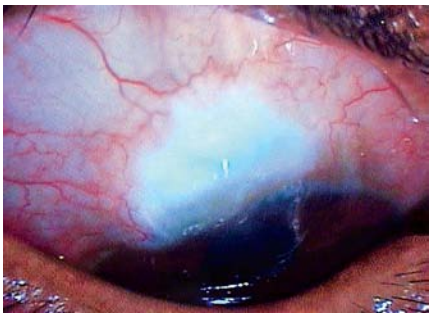


Fig. 9.20.11A: Bleb infection—blebitis

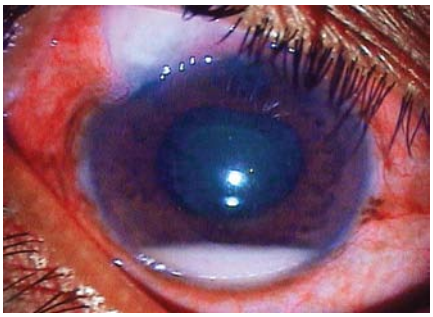


Fig. 9.20.11B: Bleb infection—blebitis

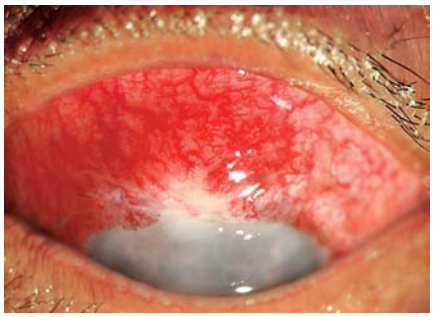


Fig. 9.20.12A: Bleb infection—blebitis with endophthalmitis

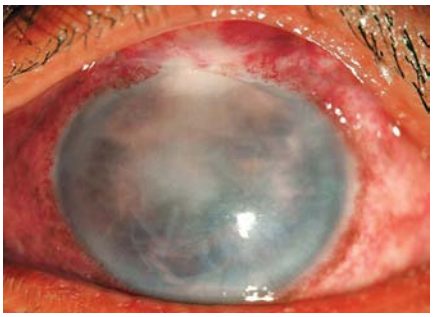


Fig. 9.20.12B: Bleb infection—blebitis with endophthalmitis

Other Complications

- Hemorrhage within filtering bleb **(Figs 9.20.13A and B)**
- Iris incarceration (peaking) **(Fig. 9.20.14)**
- Wind-shield wiper sign with a releasable suture **(Figs 9.20.15A and B)**.

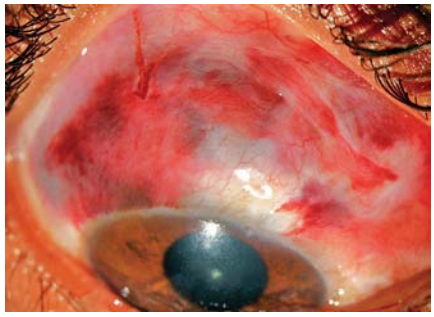


Fig. 9.20.13A: Filtering bleb with hemorrhage

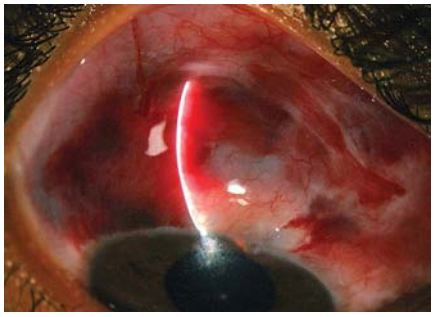


Fig. 9.20.13B: Filtering bleb with hemorrhage

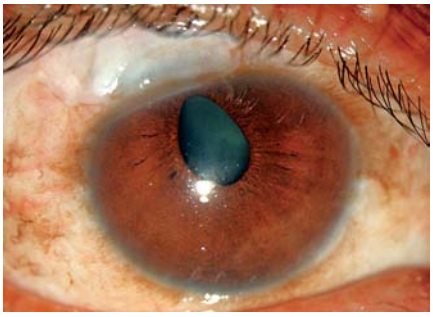


Fig. 9.20.14: Bleb with iris peaking

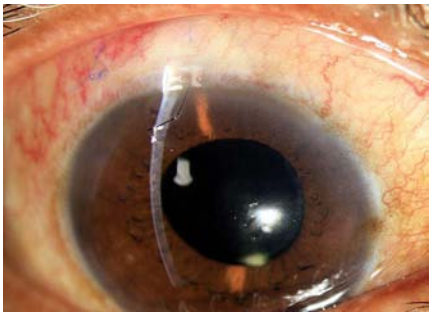


Fig. 9.20.15A: Filtering surgery with releasable suture

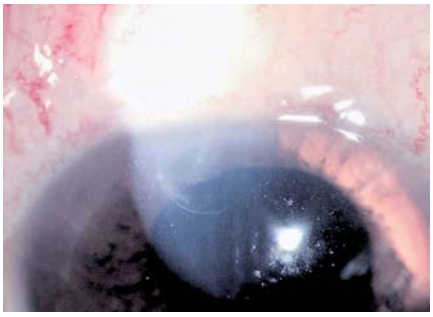


Fig. 9.20.15B: Filtering surgery with releasable suture—windshield wiper sign

Pseudobleb

- May occur without doing a filtering surgery
- Usually seen following cataract surgery by conventional ECCE **(Figs 9.20.16A and B)**
- No treatment is required.

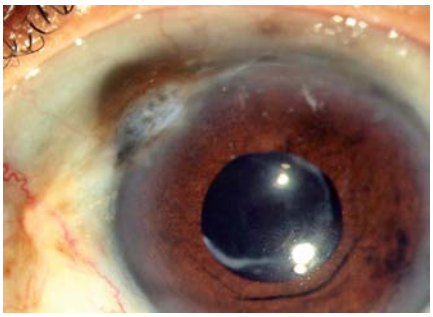


Fig. 9.20.16A: Pseudobleb following ECCE

Fig. 9.20.16B: Pseudobleb following ECCE

CHAPTER

10 Diseases of the Vitreous

CHAPTER OUTLINE

Vitreous Opacities

- Muscae volitantes
- Asteroid hyalosis (Benson's disease)
- Synchysis scintillans
- Persistent hyperplastic primary vitreous (PHPV)
- Vitreous cells
- Vitreous exudates
- Vitreous hemorrhage

Miscellaneous Vitreous Opacities

- Pigment cells ('tobacco dust' sign)
- Cotton ball exudates
- Parasite in the vitreous cavity
- Foreign bodies in the vitreous
- Vitreous prolapse

Vitreous Opacities

- Normally, under slit-lamp it appears optically clear
- Many natural or foreign substances may be suspended in the vitreous cavity **(Figs 10.1.1A to D)**
- *Endogenous*: Vascular remnants, leukocytes, RBCs, tumor cells, pigments, calcium or cholesterol crystals, etc.
- *Exogenous*: Parasite, foreign particles.

Muscae Volitantes

- Unilateral or bilateral minute fly-like or cobweb opacities best perceived by the patient against white or bright light background
- Represent physiologic remnants of primitive hyaloid vascular system
- Reassurance is the only treatment.

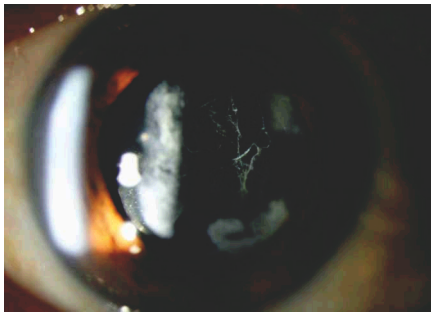


Fig. 10.1.1A: Vitreous opacity—fibrils

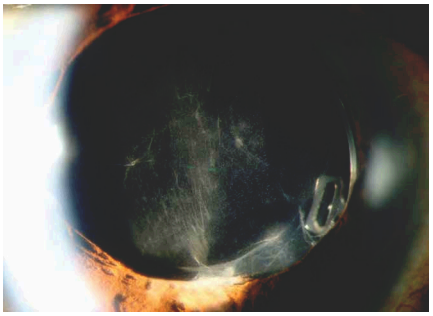


Fig. 10.1.1B: Vitreous opacity—cells and fibrils

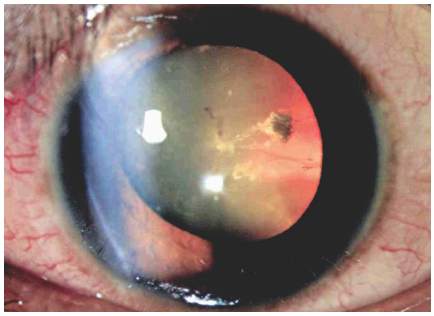


Fig. 10.1.1C: Vitreous opacity—suspended small blood clot

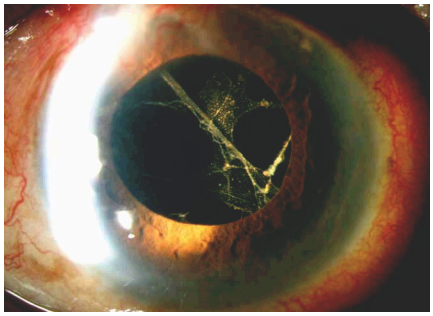


Fig. 10.1.1D: Vitreous opacity in aphakia

Asteroid Hyalosis (Benson's Disease)

- An involutional, usually bilateral condition that affects aged patients
- Appear as numerous, white, round or discoid bodies suspended throughout, or in a portion of the solid vitreous **(Fig. 10.2.1)**
- Represent calcium soap crystals from degeneration of vitreous fibrils **(Fig. 10.2.2)**
- Symptom less and do not require treatment.

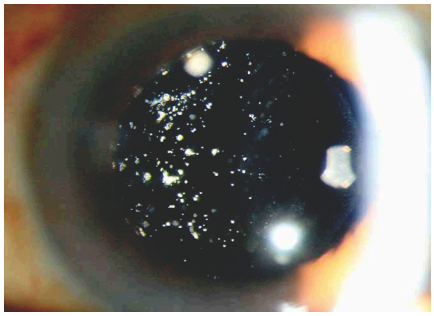


Fig. 10.2.1: Asteroid hyalosis

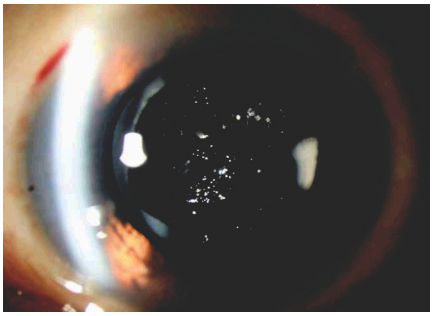


Fig. 10.2.2: Asteroid hyalosis

Synchysis Scintillans

- Unilateral condition, with previous history of vitreous hemorrhage or inflammation
- Crystals appear as golden or multi-colored glittering particles which settle at the bottom of the vitreous cavity **(Fig. 10.3.1)**
- Can be thrown upwards by the ocular movements, to form a 'golden shower' **(Fig. 10.3.2)**
- After ICCE or in traumatic cases, the crystals may also be seen in the anterior chamber **(Fig. 10.3.3)**
- No effective treatment.

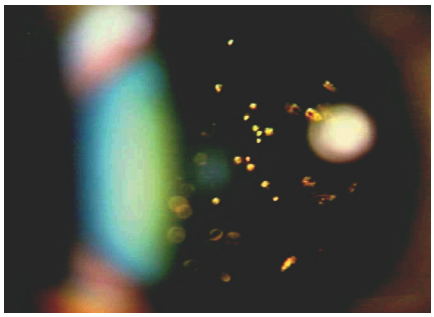


Fig. 10.3.1: Synchysis scintillans—anterior vitreous

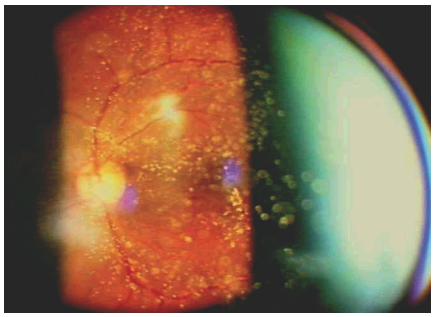


Fig. 10.3.2: Synchysis scintillans—posterior vitreous

Fig. 10.3.3: Synchysis scintillans—anterior chamber

Persistent Hyperplastic Primary Vitreous (PHPV)

- Unilateral condition, and the affected eye is smaller than the fellow eye due to faulty development
- White pupillary reflex is noticed in the full-term infant, shortly after birth
- Under slit-lamp, a yellowish-white mass may be visible in the anterior vitreous
- Complete PHPV is usually associated with microphthalmos, cataract, or long ciliary processes
- It may be *anterior or posterior* type.
- *Anterior type* may be with posterior polar cataract and small stump of blood vessels attached to it **(Figs 10.4.1A and B)**
- *Posterior type* is associated with Bergmeister's papilla and a tuft of blood vessels **(Figs 10.4.2 and 10.4.3)**

Fig. 10.4.1A: Persistent hyperplastic primary vitreous—anterior

Fig. 10.4.1B: Persistent hyperplastic primary vitreous—anterior

Fig. 10.4.2A: Persistent hyperplastic primary vitreous—Bergmeister's papilla

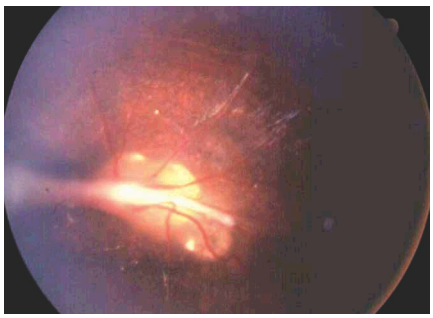


Fig. 10.4.2B: PHPV—Bergmeister's papillae

- Must be differentiated from other causes of white pupillary reflex or leukocoria *(See Chapter 7)*
- *Treatment:* Lensectomy and vitrectomy as early as possible depending upon the type of PHPV.

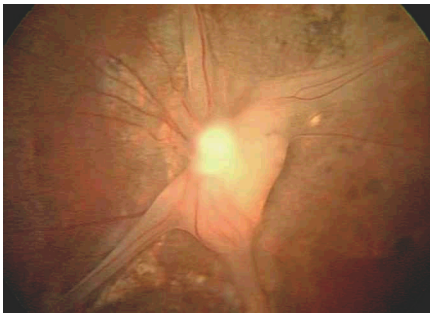


Fig. 10.4.3: Persistent hyperplastic primary vitreous

Vitreous Cells

- *Iridocyclitis*: Retrolental cells along with aqueous cells, especially when the cyclitis is predominant **(Figs 10.5.1A and B)**
- *Intermediate uveitis*: Different degree of vitreous cells and inferior peripheral vitreous snow-banking
- *Panuveitis:* Aqueous cells, vitreous cells and KPs will be noticed **(Fig. 10.5.2)**
- *Infective endophthalmitis*: Cells or frank exudates in the vitreous. May be acute in case of penetrating injury with or without retained IOFB **(Figs 10.5.3 and 10.5.4)** and post-surgery **(Figs 10.5.5A and B)**
- Endogenous enophthalmitis **(Fig. 10.5.6)**
- *Posterior uveitis*: Cells are most dense adjacent to the lesions.

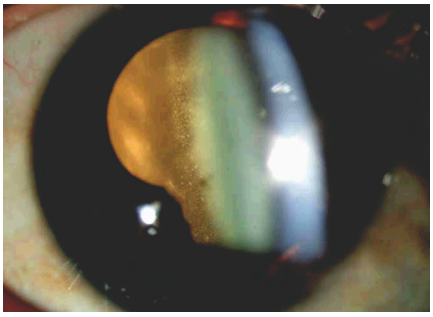


Fig. 10.5.1A: Vitreous cells in panuveitis

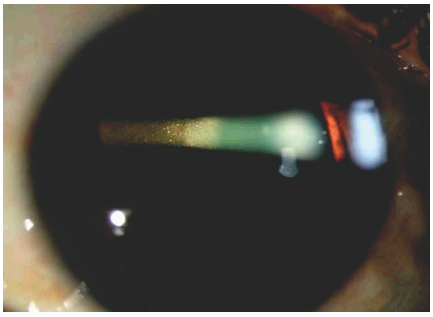


Fig. 10.5.1B: Vitreous cells in iridocyclitis

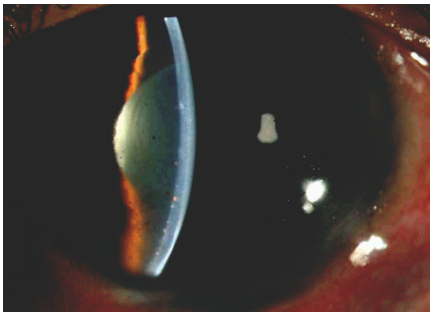


Fig. 10.5.2: Vitreous exudates in panuveitis

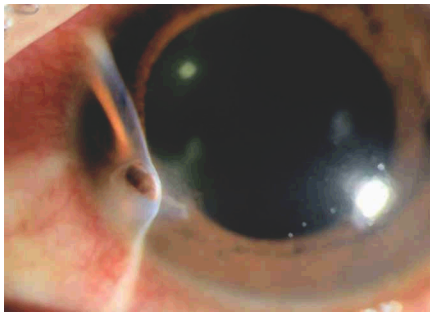


Fig. 10.5.3A: Vitreous changes in penetrating injury

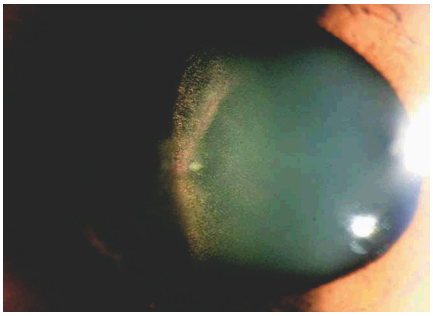


Fig. 10.5.3B: Dense vitreous cells—same eye as in **Figure 10.5.3A**

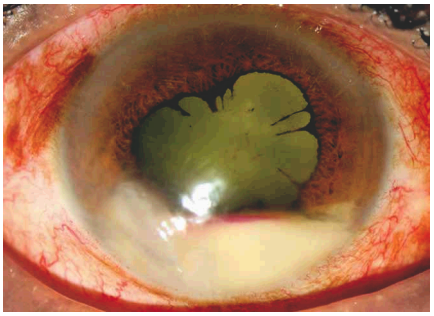


Fig. 10.5.4: Vitreous exudates in intraocular foreign body

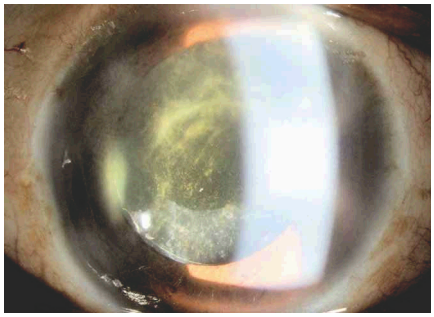


Fig. 10.5.5A: Vitreous exudates after cataract surgery

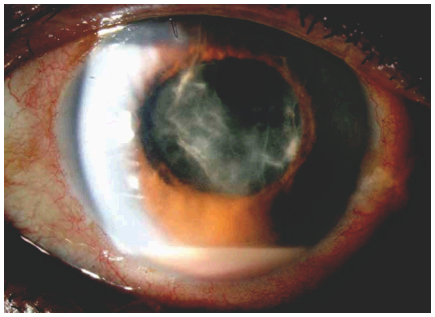


Fig. 10.5.5B: Postoperative endophthalmitis following cataract surgery

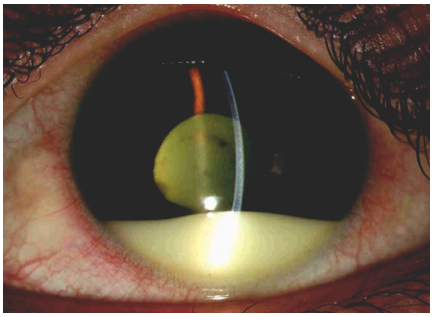


Fig. 10.5.6: Vitreous exudates in endogenous endophthalmitis

Vitreous Hemorrhage

- May occur as an intravitreal or a preretinal phenomenon **(Figs 10.6.1 and 10.6.2)**
  - *Retinal break:* Traumatic or by vitreous traction
  - *Proliferative retinopathies*: Rupture of newly formed blood vessels **(Figs 10.6.3 to 10.6.5)**
  - Acute posterior vitreous detachment (PVD)
  - Bleeding disorders
  - Periphlebitis or Eales' disease **(Figs 10.6.6A and B)**
  - Avulsion of the optic nerve
- May be seen as minute red colored or dark spots, as red mass, or with no fundal view
- Organized vitreous hemorrhage may appear as clotted blood **(Figs 10.6.7A to C)** and ultimately with membrane formation **(Figs 10.6.8A and B)**
- *Treatment:* Investigation, medical and/or pars plana vitrectomy with endolaser photocoagulation.

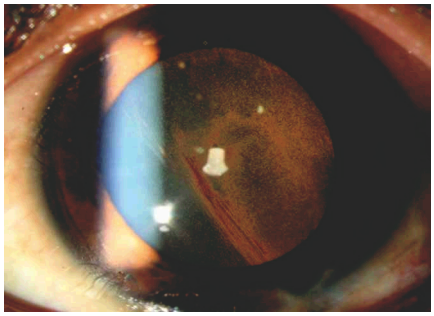


Fig. 10.6.1A: Vitreous hemorrhage—mild—after a blunt trauma

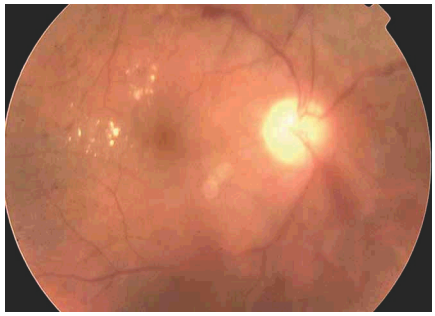


Fig. 10.6.1B: Vitreous hemorrhage—mild

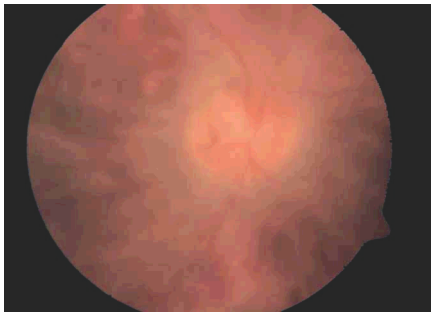


Fig. 10.6.1C: Vitreous hemorrhage—dense

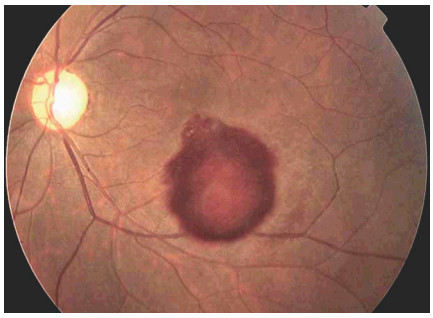


Fig. 10.6.2A: Preretinal (subhyaloid hemorrhage)

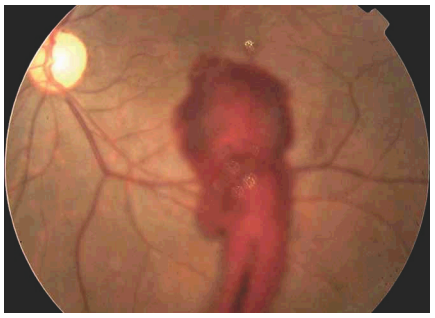


Fig. 10.6.2B: Vitreous hemorrhage—same eye after YAG laser hyaloidotomy

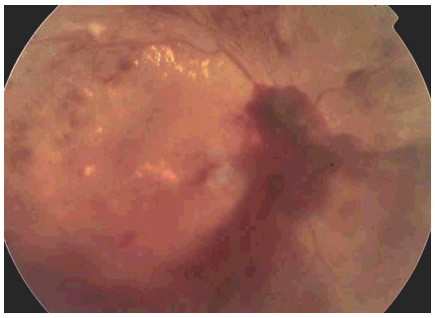


Fig. 10.6.3: Vitreous hemorrhage in PDR

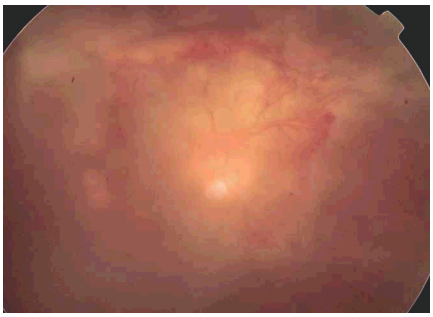


Fig. 10.6.4: Vitreous hemorrhage from NVD

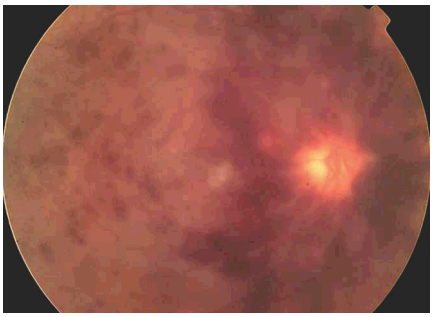


Fig. 10.6.5: Vitreous hemorrhage in CRVO

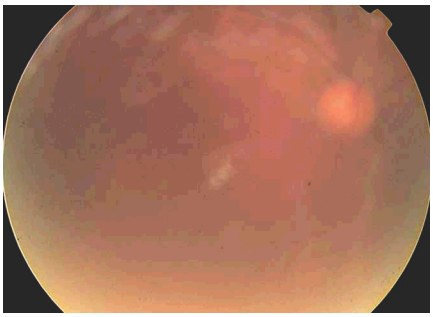


Fig. 10.6.6A: Vitreous hemorrhage—Eales' disease

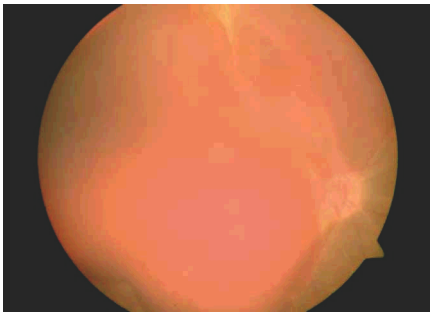


Fig. 10.6.6B: Vitreous hemorrhage—Eales' disease

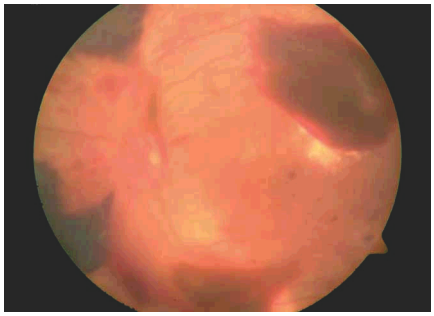


Fig. 10.6.7A: Organized vitreous hemorrhage—clot formation

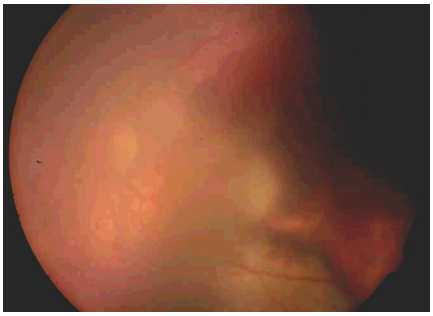


Fig. 10.6.7B: Organized vitreous hemorrhage—dense clot formation

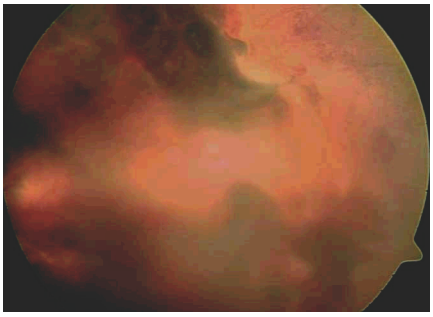


Fig. 10.6.7C: Organized vitreous hemorrhage—clot formation

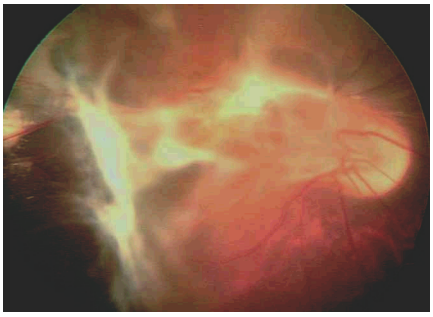


Fig. 10.6.8A: Vitreous hemorrhage—membrane formation

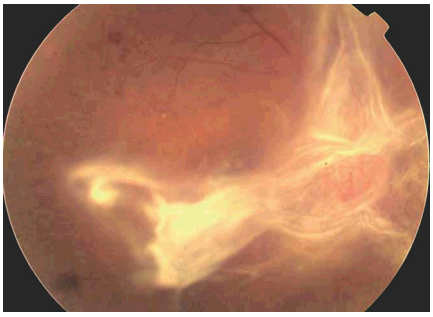


Fig. 10.6.8B: Vitreous hemorrhage—membrane formation

Miscellaneous Vitreous Opacities

Pigment Cells ('Tobacco Dust' Sign)

- Consist of macrophages containing retinal pigment epithelial cells
- Mainly visible in the anterior vitreous **(Figs 10.9.1 and 10.9.2)**
- *Causes:* Retinal tears with PVD, rhegmatogenous retinal detachment, trauma, excessive retinal cryopexy.

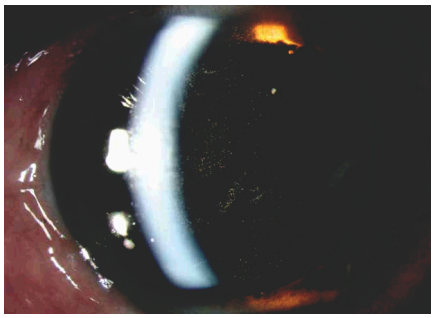


Fig. 10.9.1: Vitreous pigments—tobacco dust sign

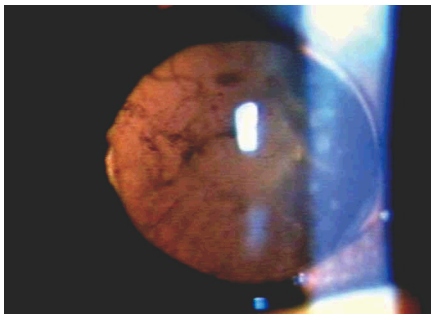


Fig. 10.9.2: Vitreous membrane with tobacco dust sign

Cotton Ball Exudates

- Seen in intermediate uveitis
- Sarcoidosis
- Candidiasis **(Figs 10.10.1 to 10.10.4)**: mainly seen in case of metastatic endophthalmitis.

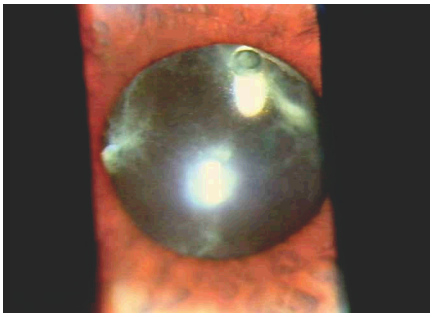


Fig. 10.10.1: Fungal cotton ball exudates in endophthalmitis

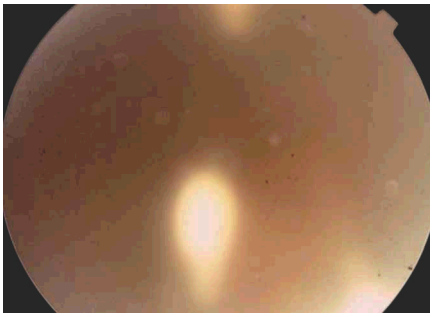


Fig. 10.10.2: Candidiasis—cotton ball opacity in vitreous

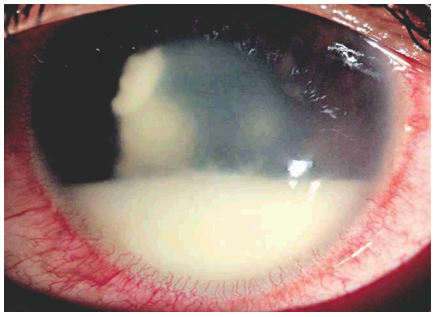


Fig. 10.10.3: Candidiasis—cotton ball exudates in vitreous

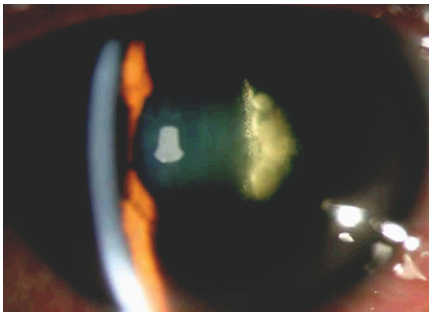


Fig. 10.10.4: Candidiasis—cotton ball exudates in vitreous

Parasite in the Vitreous Cavity

- May be live or dead parasite
  - Cysticercosis **(Figs 10.11.1A and B)**
  - Gnathostomiasis **(Figs 10.11.2A and B)**.

Fig. 10.11.1A: Cysticercus in vitreous

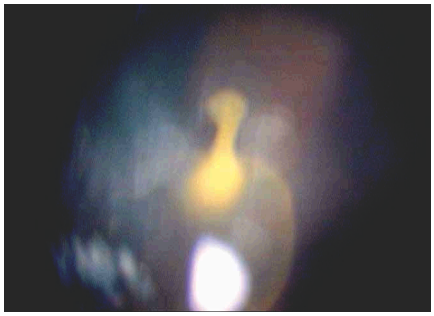


Fig. 10.11.1B: Cysticercus in vitreous

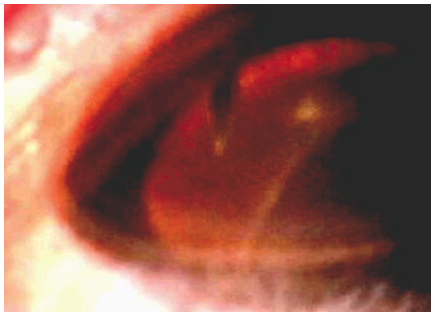


Fig. 10.11.2A: Gnathostoma in vitreous

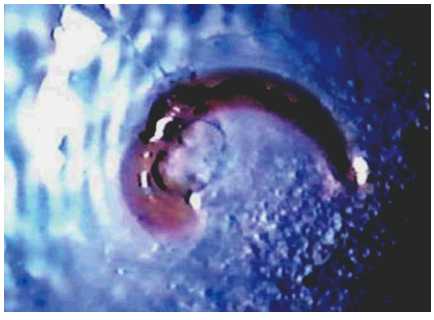


Fig. 10.11.2B: Live Gnathostoma outside—from the same eye

Foreign Bodies in the Vitreous

- Metallic **(Fig. 10.12.1)**
- Intravitreal triamcinolone injection **(Fig. 10.12.2)**
- *Silicone oil:* After VR surgery
- Air bubble and other gasses **(Figs 10.12.3 and 10.12.4)** after vitreoretinal surgery.

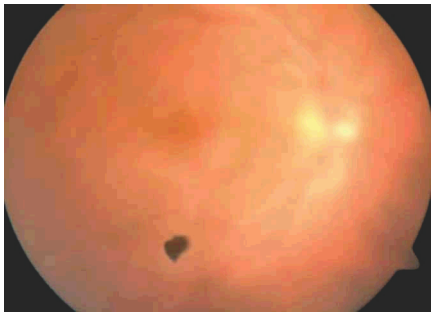


Fig. 10.12.1: Metallic foreign body in vitreous

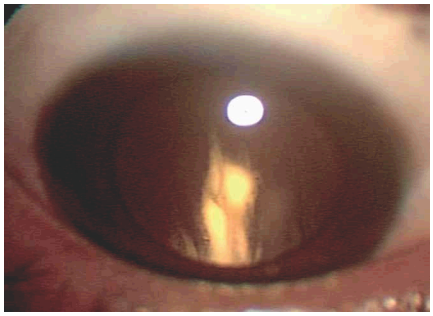


Fig. 10.12.2: Intravitreal triamcinolone

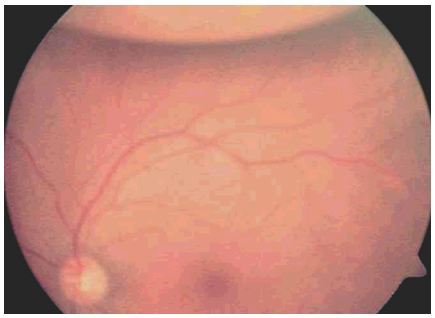


Fig. 10.12.3: Gas bubble in vitreous

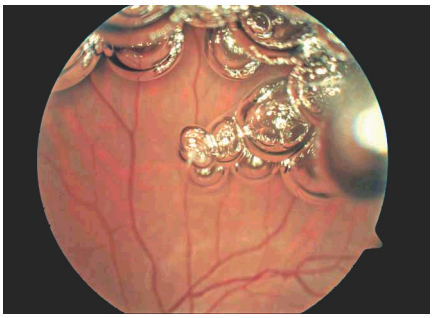


Fig. 10.12.4: Gas bubbles in vitreous

Vitreous Prolapse

- After rupture of the posterior lens capsule (in ECCE), the anterior vitreous may herniate into the anterior chamber to fill it completely **(Figs 10.13.1 and 10.13.2)**
- Vitreous also herniates into the anterior chamber in subluxation or dislocation of lens **(Fig. 10.13.3 and 10.13.4)**
- Direct contact of the vitreous with other structures, such as the cornea or the angle. Long standing vitreocorneal touch may cause corneal endothelial decompensation and eventually corneal edema **(Figs 10.14.1A and B)**
- Incarceration of the vitreous into the operative wound
- *Treatment:* Anterior vitrectomy before final procedure.

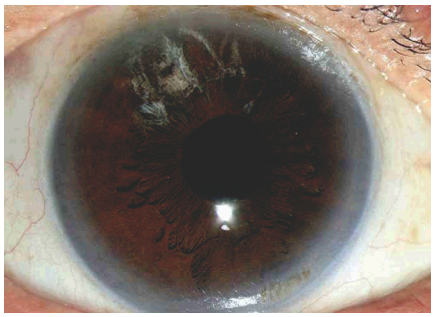


Fig. 10.13.1A: Vitreous prolapse in AC—aphakia

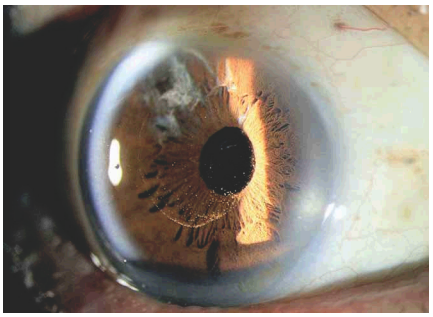


Fig. 10.13.1B: Vitreous knuckle in AC—Aphakia

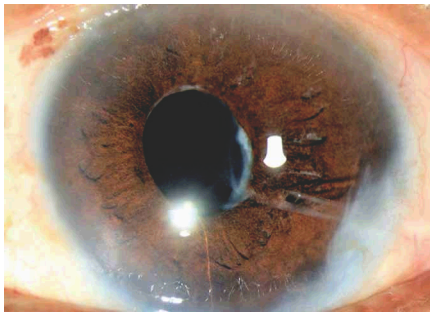


Fig. 10.13.2A: Vitreous herniation in AC in aphakia

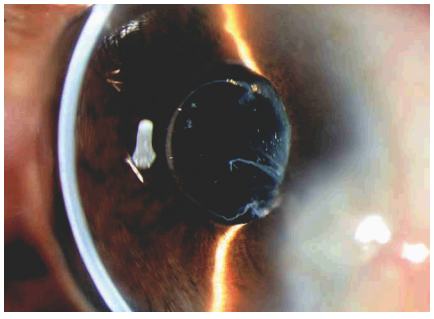


Fig. 10.13.2B: Vitreous herniation in AC—after PC rent

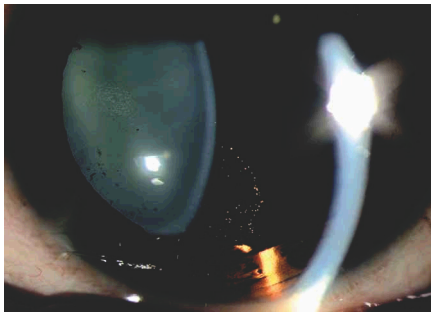


Fig. 10.13.3A: Vitreous prolapse in AC—subluxated lens

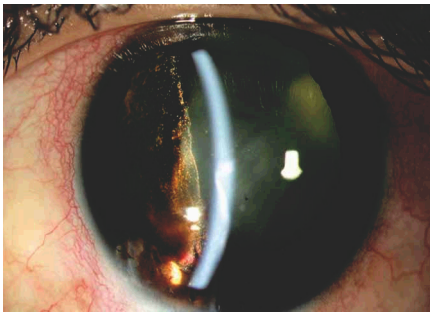


Fig. 10.13.3B: Vitreous prolapse in AC—subluxated lens

Fig. 10.13.4A: Vitreous in AC—posterior dislocation of lens

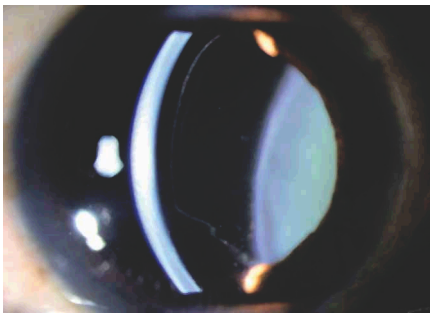


Fig. 10.13.4B: Vitreous in AC—traumatic dislocation of lens

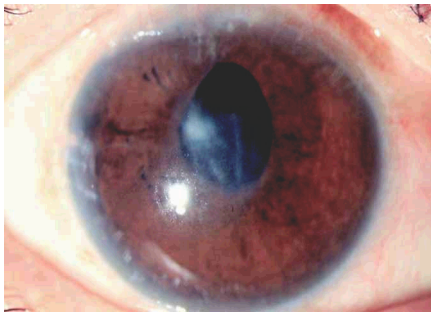


Fig. 10.14.1A: Vitreous prolapse in AC—vitreocorneal touch

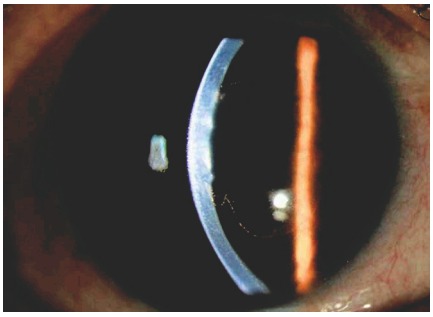


Fig. 10.14.1B: Vitreocorneal touch in aphakia with corneal edema

CHAPTER

11 Diseases of the Optic Nerve

CHAPTER OUTLINE

Congenital Abnormalities of Optic Disk

- Myelinated nerve fibers
- Persistent hyaloid artery
- Optic disk drusen
- Optic disk conus
- Congenital optic disk pit
- Coloboma of the optic disk
- 'Morning glory' syndrome
- Small disk: High hypermetropia
- Hypoplastic optic disk
- Tilted disk
- Large disk: Myopia
- Large disk (megalopapilla)
- Disk with large cup
- Glaucomatous cupping

Vascular Anomalies at the Disk

- Disk collaterals (veno-venular shunt)
- New vessels on the disk (NVD)
- Opticociliary shunts
- Disk hemorrhage
- Dragged vessels of the disk

Optic Neuritis

- Papillitis
- Retrobulbar neuritis
- Neuroretinitis
- Anterior ischemic optic neuropathy (AION)

Other Causes of Unilateral Disk Edema

- Central retinal venous occlusion
- Central vasculitis (papillophlebitis)
- Long standing hypotony
- Posterior scleritis

Papilledema

- Early papilledema
- Established (acute) papilledema
- Chronic papilledema
- Atrophic papilledema

Other Causes of Bilateral Disk Edema

- Malignant hypertension

Optic Atrophy

- Primary optic atrophy
- Secondary optic atrophy
- Consecutive optic atrophy
- Glaucomatous optic atrophy
- Temporal pallor

Primary Optic Disk Tumors

- Optic disk capillary hemangioma
- Optic disk cavernous hemangioma
- Arteriovenous malformation of optic disk
- Optic disk melanocytoma
- Astrocytoma of the optic disk
- Optic nerve glioma and meningioma

Infiltrative Disk Lesions

Congenital Abnormalities of Optic Disk

Myelinated Nerve Fibers

- Myelin sheaths of the optic nerve fibers cease normally at the lamina cribrosa
- Congenital condition, present 1% of normal population
- Appear as white patches with radial striations (feathery) at peripheral edges **(Fig. 11.1.1)**
- Usually unilateral and bilateral in 20% cases **(Figs 11.1.2A and B)**
- Usually peripapillary; and sometimes peripheral and isolated **(Figs 11.1.3A and B)**
- Myelin sheaths disappear in case of optic atrophy
- *No treatment* is required.

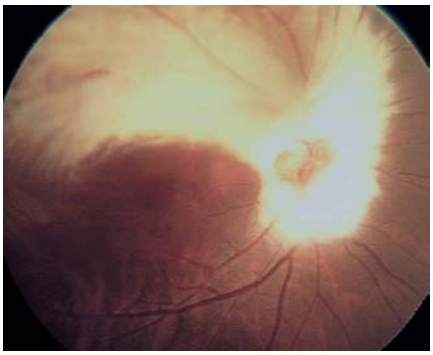


Fig. 11.1.1: Myelinated nerve fibers

Persistent Hyaloid Artery

- A short stub of this vessel projects into the vitreous cavity from the center of the optic disk **(Fig. 11.2.1)**, and is surrounded by a small mass of glial tissue—called *Bergmeister's papilla* **(Fig. 11.2.2)**
- May project into the vitreous cavity as persistent hyaloid vessels—with glial proliferation **(Fig. 11.2.3)**

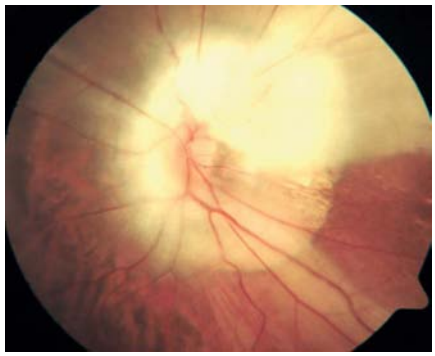


Fig. 11.1.2A: Myelinated nerve fibers—bilateral—RE

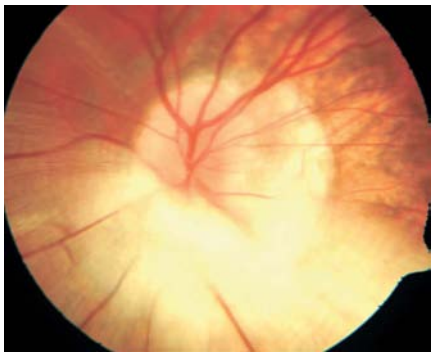


Fig. 11.1.2B: Myelinated nerve fibers—bilateral—LE

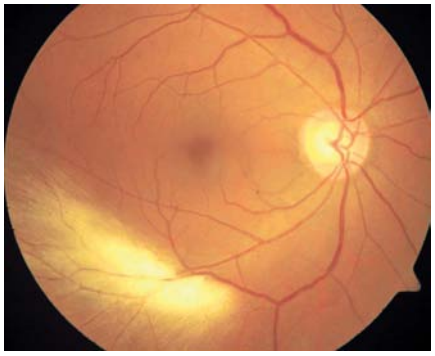


Fig. 11.1.3A: Myelinated nerve fibers—peripheral and bilateral—RE

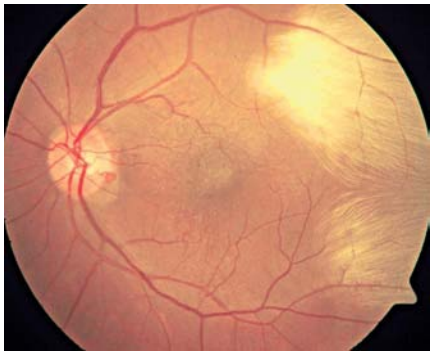


Fig. 11.1.3B: Myelinated nerve fibers—peripheral and bilateral—LE

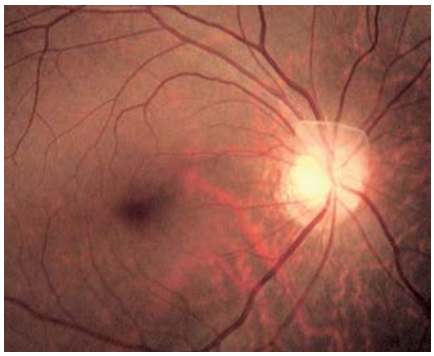


Fig. 11.2.1: Bergmeister's papilla

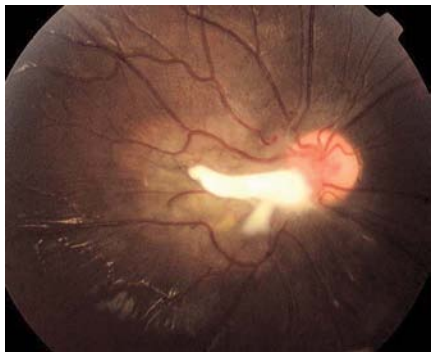


Fig. 11.2.2: Persistent hyaloid vessels—Bergmeister's papilla

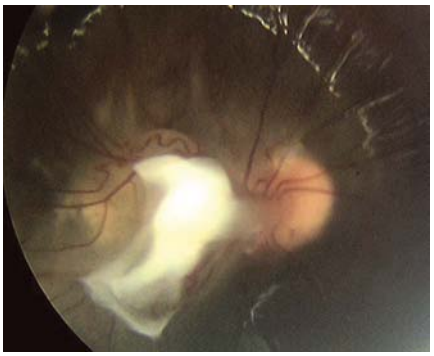


Fig. 11.2.3: Persistent hyaloid vessels with glial proliferation

- Sometimes, it attaches to the posterior capsule of the lens *(Mittendorf's dot)*
- Failure of the hyaloid artery of regress cause persistent primary vitreous, which may proliferate to PHPV as a retrolental white mass **(Figs 11.2.4 and 11.2.5)**
- *Treatment* is required in extreme cases.

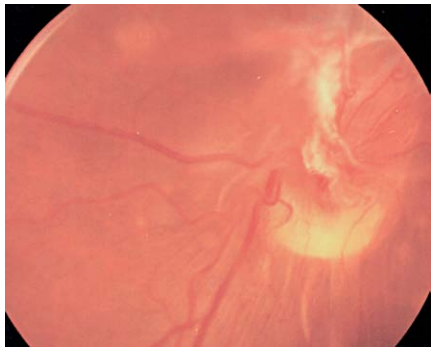


Fig. 11.2.4: Persistent hyperplastic primary vitreous PHPV—posterior type

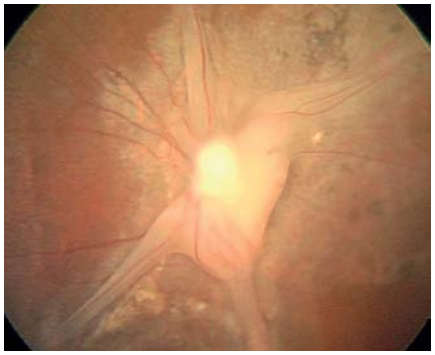


Fig. 11.2.5: Posterior PHPV

Optic Disk Drusen

- *Common drusens* are laminated and nodular
- It may be single nodule **(Fig. 11.3.1)** or multiple
- The multiple may give the disk margins a blurred appearance (pseudopapilledema) **(Fig. 11.3.2)** and type rarely causes visual disturbances
- Second type is *giant drusens*, which are astrocytic hamartomas that occur in patient with tuberous sclerosis **(Fig. 11.3.3)**.

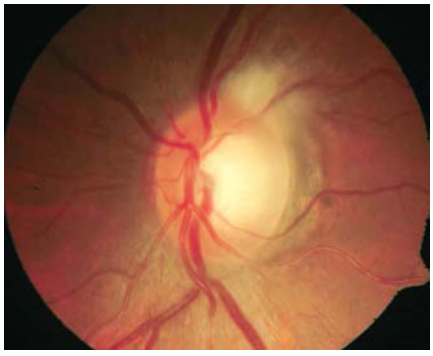


Fig. 11.3.1: Optic nerve dursen—single

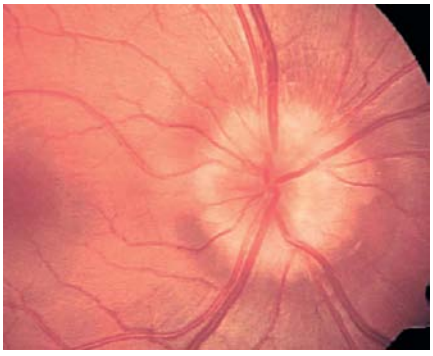


Fig. 11.3.2: Optic nerve drusen—multiple

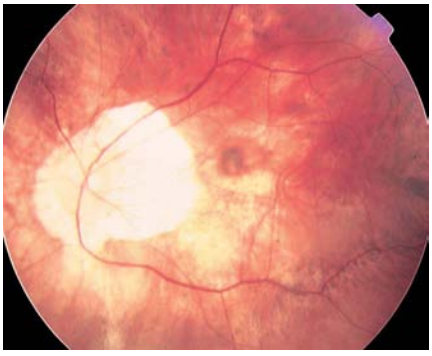


Fig. 11.3.3: Optic disk astrocytoma

Optic Disk Conus

- Congenital optic disk crescent
- A large, white, semilunar area of sclera, seen adjacent to the disk, in the region of primitive retinal fissure (inferonasal) **(Figs 11.4.1 and 11.4.2)**
- Often associated with visual field defect

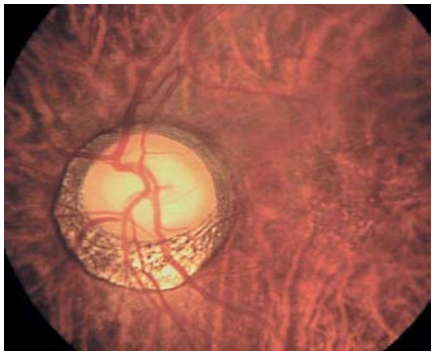


Fig. 11.4.1: Optic disk conus

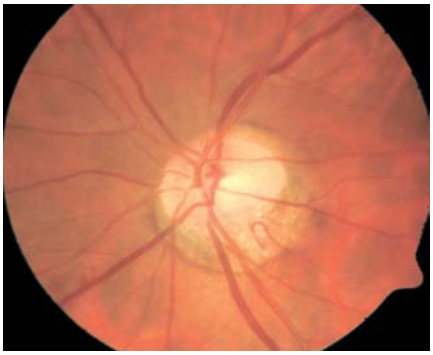


Fig. 11.4.2: Optic disk conus

- A myopic crescent has a similar appearance, and is located temporal to the disk **(Figs 11.4.3A and B)**.

It is not present at birth, and is associated with high myopia.

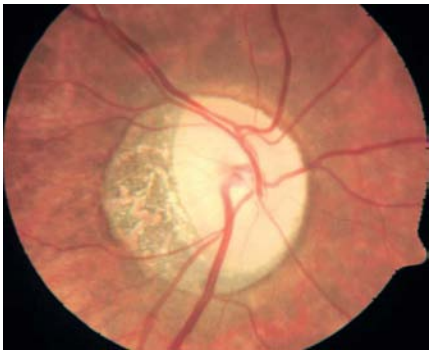


Fig. 11.4.3A: Myopic temporal crescent

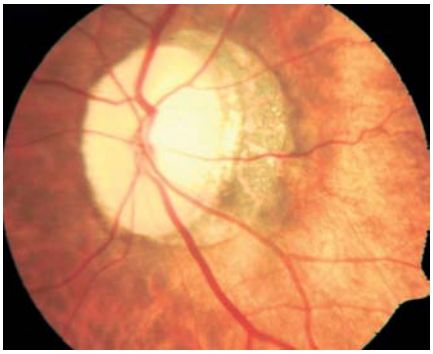


Fig. 11.4.3B: Myopic temporal crescent

Congenital Optic Disk Pit

- Isolated congenital unilateral condition
- Round or oval depression, most frequently involve the temporal margin and usually round or oval **(Figs 11.5.1 and 11.5.2)**
- A central scotoma is often present, and in 30% of the cases, a central serous retinopathy (CSR) develops **(Figs 11.5.3 and 11.5.4)**
- A central pit is less common **(Fig. 11.5.5)**.

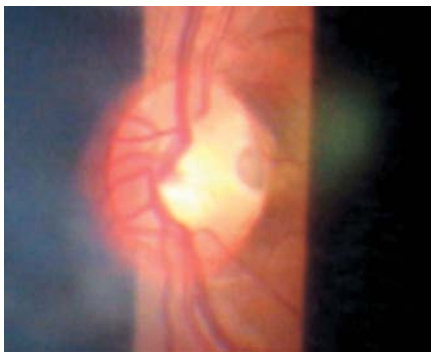


Fig. 11.5.1: Congenital optic disk pit

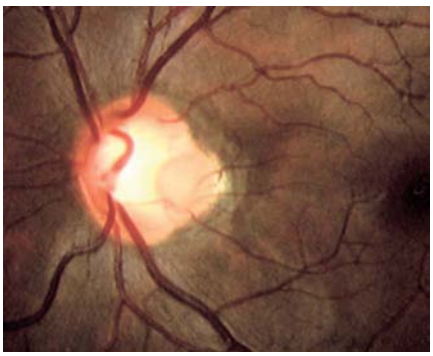


Fig. 11.5.2: Congenital optic disk pit

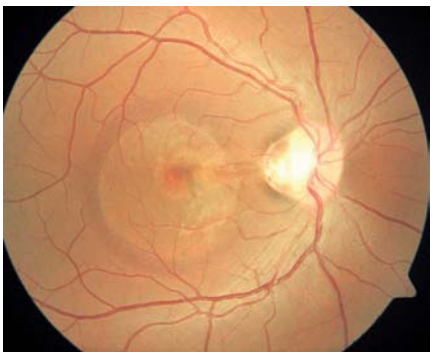


Fig. 11.5.3: Congenital optic disk pit with CSR

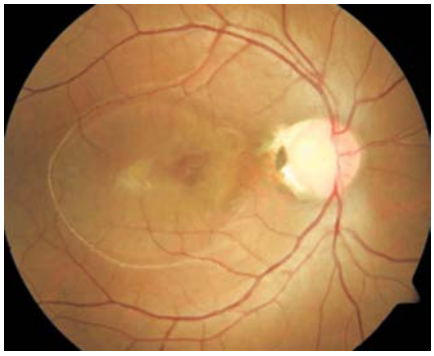


Fig. 11.5.4A: Congenital optic disk pit with CSR

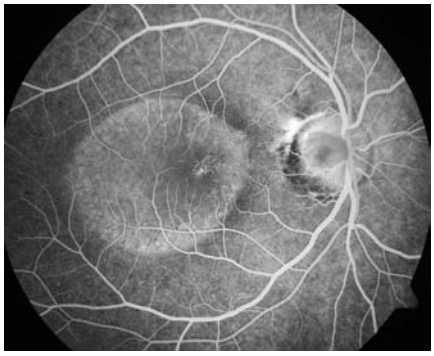


Fig. 11.5.4B: Congenital optic disk pit with CSR on FFA

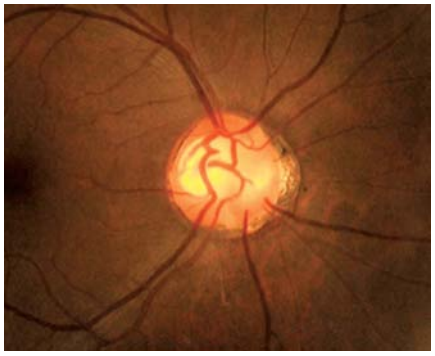


Fig. 11.5.5: Congenital central optic disk pit

Coloboma of the Optic Disk

- Rare and is usually unilateral **(Fig. 11.6.1)**; but bilateral optic disk colobomata occur as an autosomal dominant hereditary defect **(Figs 11.6.2A and B)**
- Typically, large disk with inferior excavation with glistening appearance of underlying sclera **(Fig. 11.6.3)**
- May be an extension with typical choroidal coloboma of other parts **(Fig. 11.6.4)**
- May be associated with isolated choroidal coloboma of other parts giving rise to "double disk" **(Fig. 11.6.5)** or triple disk appearance **(Figs 11.6.6 and 11.6.7)**.

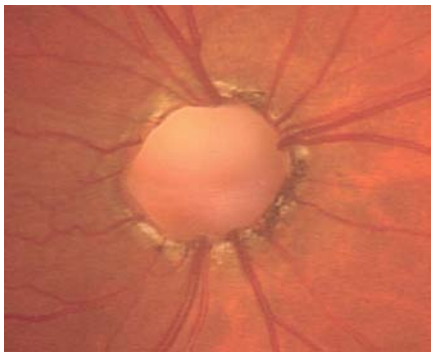


Fig. 11.6.1: Optic disk coloboma

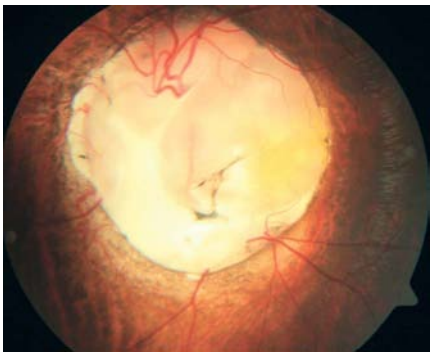


Fig. 11.6.2A: Optic disk coloboma—bilateral—RE

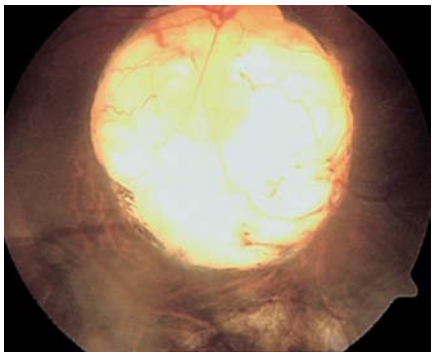


Fig. 11.6.2B: Optic disk coloboma—bilateral—LE

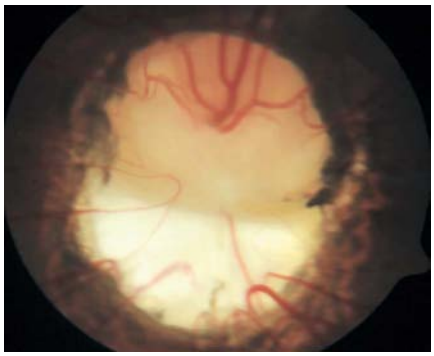


Fig. 11.6.3: Optic disk coloboma

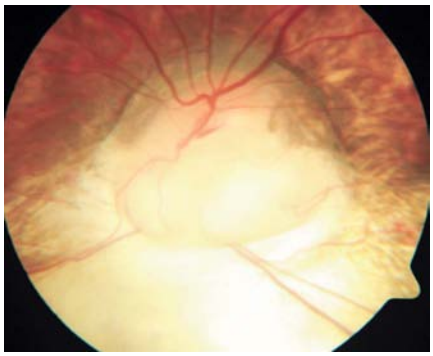


Fig. 11.6.4: Retinochoroidal coloboma including optic disk coloboma

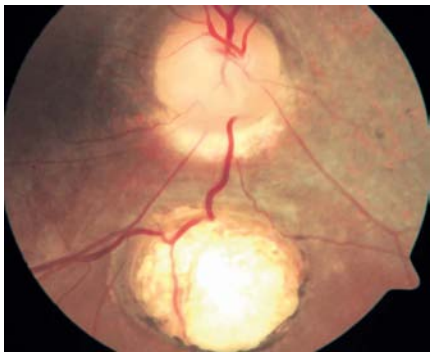


Fig. 11.6.5: Coloboma isolated—double disk appearance

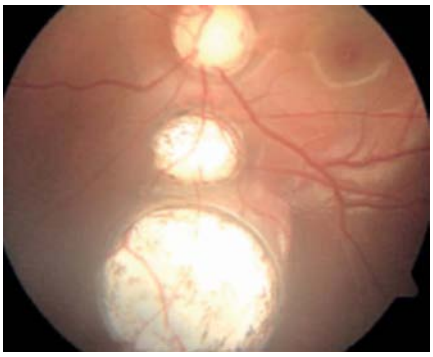


Fig. 11.6.6: Isolated colobomas—'triple disk' appearance

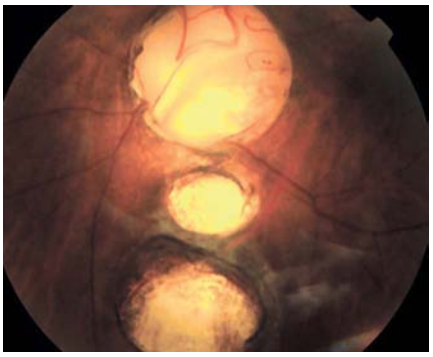


Fig. 11.6.7: Disk coloboma and isolated choroidal coloboma—triple disk appearance

'Morning Glory' Syndrome

- Usually unilateral, congenital and with poor visual acuity
- Disk looks large with core of white tissue **(Fig. 11.7.1)**
- Vessels have an abnormal distribution emerging only around the edges in a spoke-like fashion **(Fig. 11.7.2)**
- Surrounded by an annular choroidal ring **(Fig. 11.7.3)**
- May be associated with other ocular abnormalities.

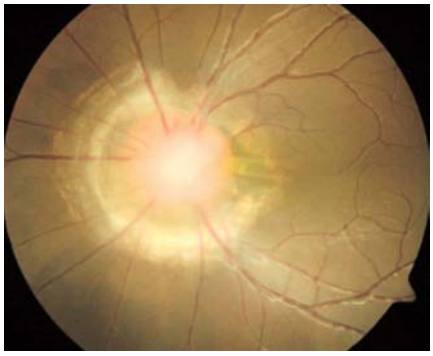


Fig. 11.7.1: Morning glory syndrome—core of white tissue

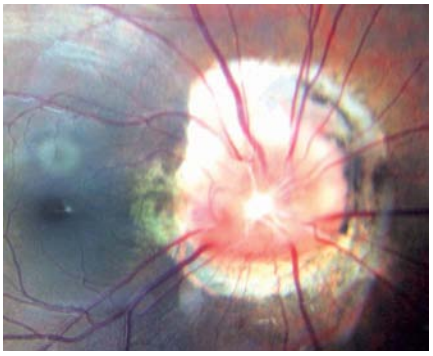


Fig. 11.7.2: Morning glory syndrome—spoke-like blood vessels

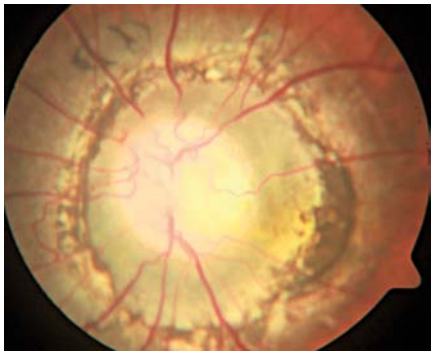


Fig. 11.7.3: Morning glory syndrome—annular choroidal ring

Small Disk: High Hypermetropia

- Disk is more pink and slightly elevated with small or absent cup **(Figs 11.8.1A and B)**
- Crowding of normal retinal vessels
- Should be differentiated from papilledema as the disk margin is blurred.

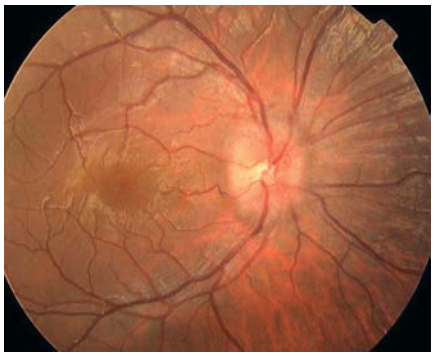


Fig. 11.8.1A: Small disk—high hypermetropia—RE

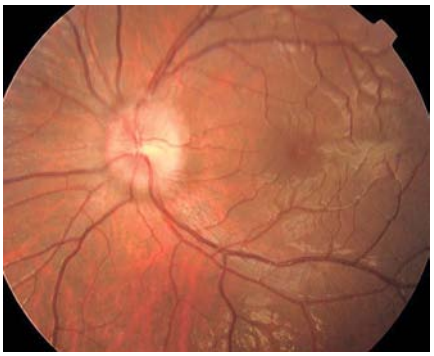


Fig. 11.8.1B: Small disk—high hypermetropia—LE

Hypoplastic Optic Disk

- Failure of the axons of the ganglionic cells to develop or to reach the disk causes a small hypoplastic optic disk **(Fig. 11.9.1)**
- Unilateral or bilateral congenital condition
- Typical appearance consists of a small gray optic disk surrounded by a yellowish peripapillary halo of hypopigmentation (*double-ring sign*) **(Figs 11.9.2A and B)**
- Outer ring corresponds to the margin of normal disk
- Blood vessels are of normal caliber.

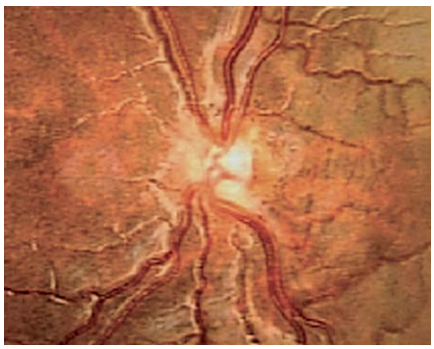


Fig. 11.9.1: Hypoplastic optic disk

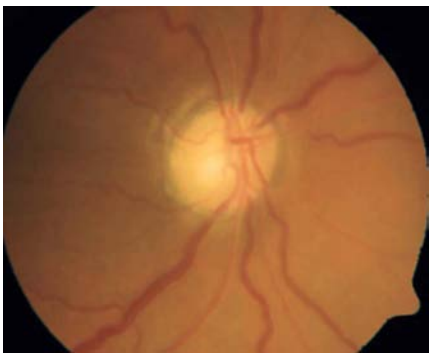


Fig. 11.9.2A: Hypoplastic optic disk—double ring sign

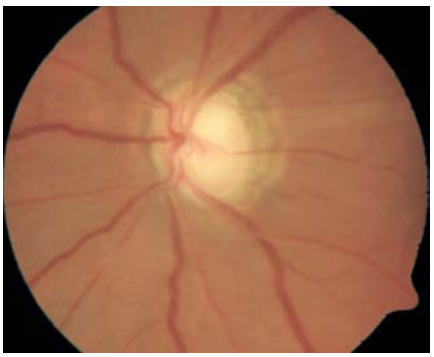


Fig. 11.9.2B: Hypoplastic optic disk—double ring sign

Tilted Disk

- It is due to an oblique entrance of the optic nerve into the globe
- Congenital and usually bilateral
- Appearance of the disk is extremely oval or 'D-shaped', with the vertical axis directed obliquely **(Fig. 11.10.1)**
- Hypopigmentation of inferonasal fundus and peripapillary atrophy **(Fig. 11.10.2)**
- *Situs inversus*: Temporal blood vessels deviate nasally before turning temporally **(Figs 11.10.3A and B)**
- May be associated with myopia, and sometimes with temporal field defect.

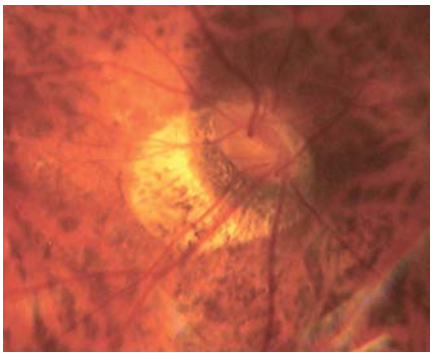


Fig. 11.10.1: Tilted optic disk

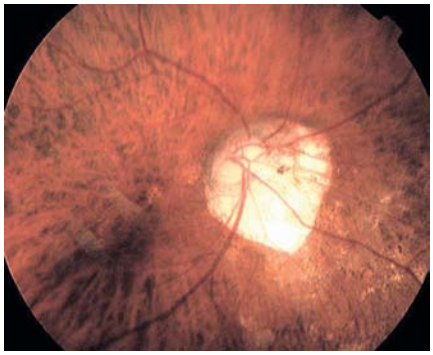


Fig. 11.10.2: Tilted optic disk

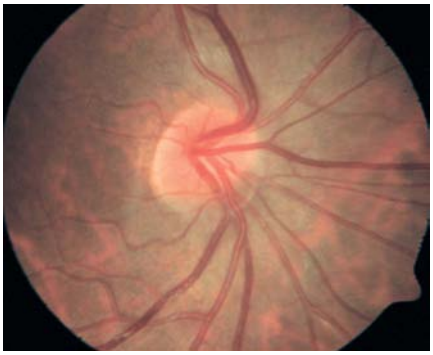


Fig. 11.10.3A: Tilted disk—situs inversus optica—RE

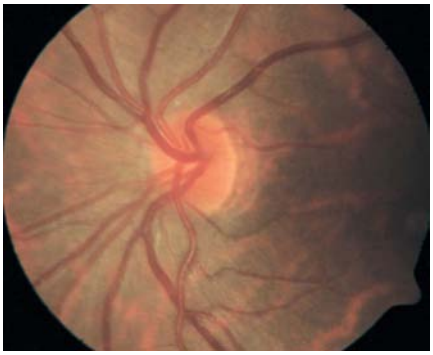


Fig. 11.10.3B: Tilted disk—situs inversus optica—LE

Large Disk: Myopia

- Disk is larger than normal, with normal or large cup **(Fig. 11.11.1)**
- Should be differentiated from glaucomatous cupping
- Temporal **(Figs 11.11.2A and B)** or annular crescent **(Figs 11.11.3A and B)** and peripapillary atrophy **(Fig. 11.11.4)**
- May be associated more degenerative changes with posterior staphyloma in very high myopia **(Figs 11.11.5A and B)**.

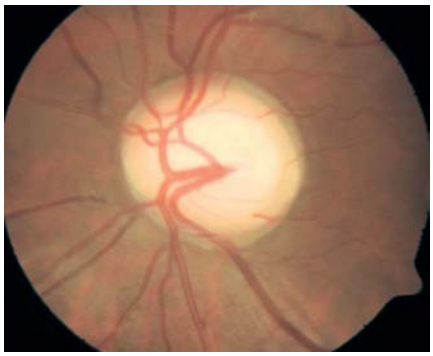


Fig. 11.11.1: Myopic disk with large cup

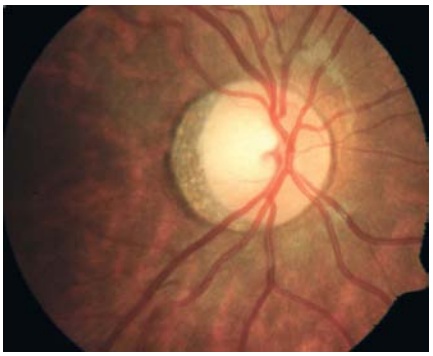


Fig. 11.11.2A: Myopic disk—temporal crescent—RE

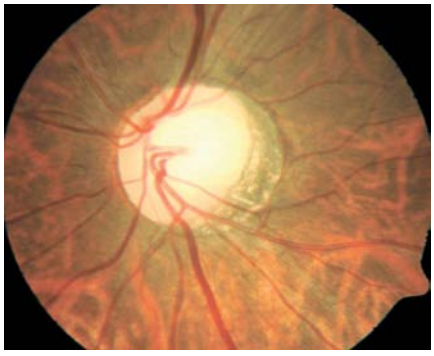


Fig. 11.11.2B: Myopic disk—temporal crescent—LE

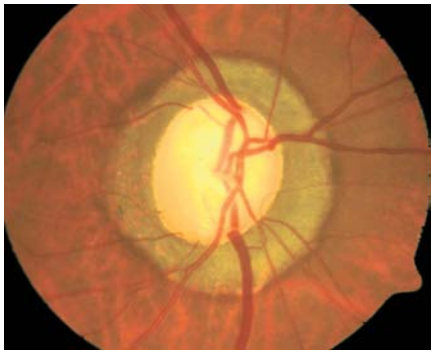


Fig. 11.11.3A: Myopic disk—annular crescent—RE

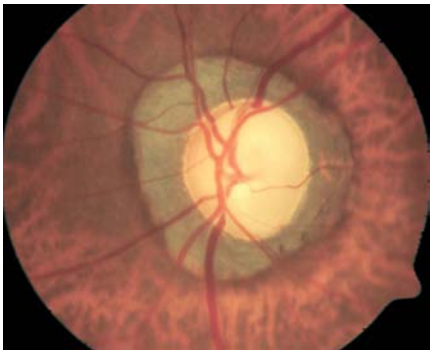


Fig. 11.11.3B: Myopic disk—annular crescent—LE

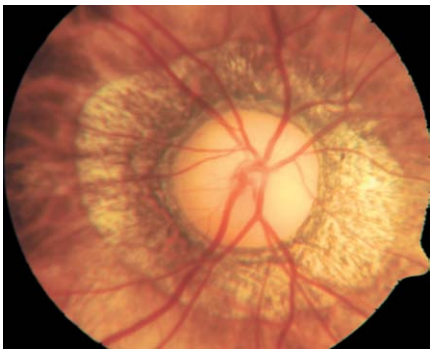


Fig. 11.11.4: Myopic disk—annular crescent with peripapillary atrophy

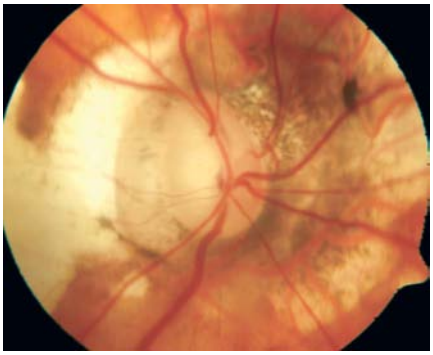


Fig. 11.11.5A: Degenerative myopia—posterior staphyloma

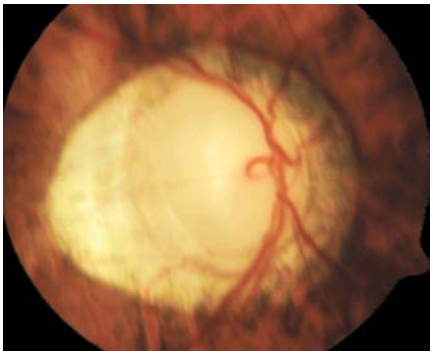


Fig. 11.11.5B: Degenerative myopia—posterior staphyloma

Large Disk (Megalopapilla)

- Very rare, congenital unilateral condition
- Disk diameter +2.0 mm or more **(Figs 11.12.1A and B)**
- Reduced distance between foveola and temporal edge of the disk
- Blood vessels appear normal.

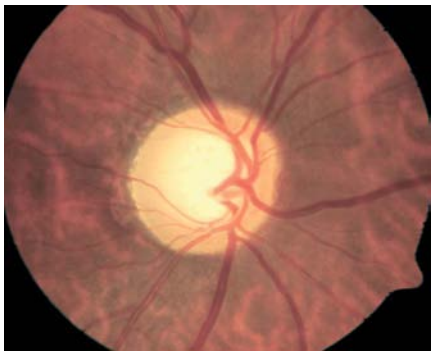


Fig. 11.12.1A: Normal disk—RE

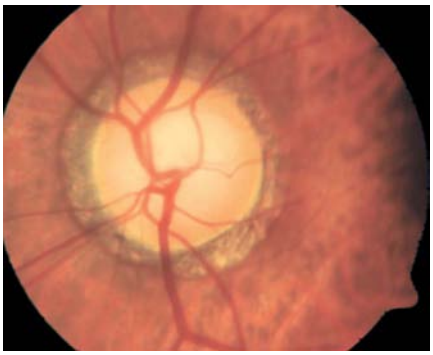


Fig. 11.12.1B: Megalopapilla—same patient—LE

Disk with Large Cup

- Normal cups are of three types:
  - Small dimple cup
  - Punched out cup
  - Cup with temporal slopping
- Normal cup : disk ratio is 0.3 or less **(Figs 11.13.1A and B)**
- A cup : disk ratio > 0.4 is present in 2–5% of normal population **(Figs 11.13.2A and B)**

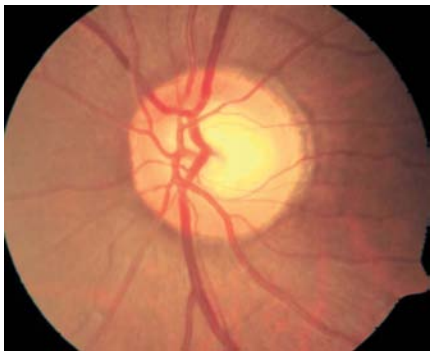


Fig. 11.13.1A: Normal cupping of the disk

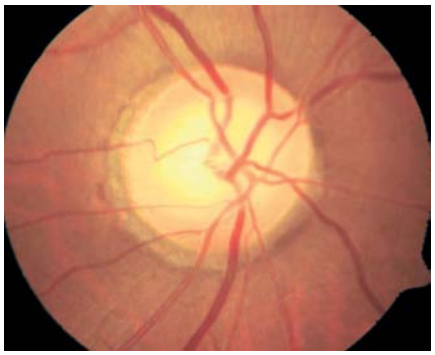


Fig. 11.13.1B: Normal cupping of the disk

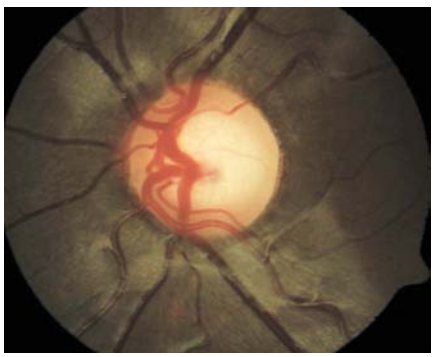


Fig. 11.13.2A: Large cup in emmetropia

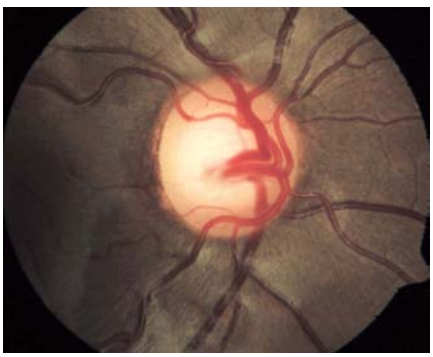


Fig. 11.13.2B: Large cup in emmetropia

- Bilateral and symmetrical **(Figs 11.13.3A and B)**
- Absent of notching
- Peripapillary striation of nerve fiber layer can be seen in most cases.

Glaucomatous Cupping

See Chapter 9.

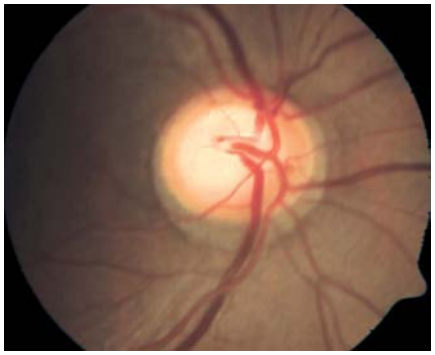


Fig. 11.13.3A: Large cup in myopia—RE

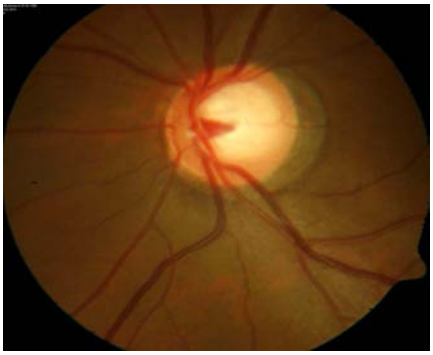


Fig. 11.13.3B: Large cup in myopia—LE

Vascular Anomalies at the Disk

Disk Collaterals (Veno-venular Shunt)

- Common shunts that develop within the exiting vascular system
- They are distended flat vessels that start and end on the disk surface **(Figs 11.14.1 and 11.14.2)**
- *Most common cause*: Central retinal venous occlusion.

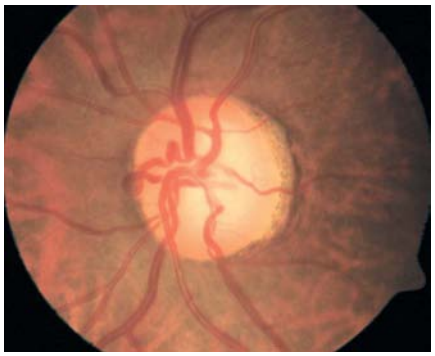


Fig. 11.14.1: Disk collaterals

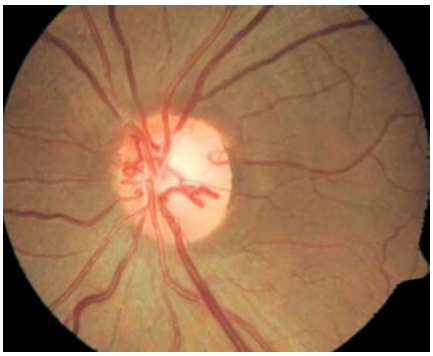


Fig. 11.14.2: Disk collaterals

Opticociliary Shunts

- Rare, unilateral anastomosis between the retinal and choroidal blood vessels
- Vessels hook from the center of the cup to the disk—retina junction **(Fig. 11.14.3)**
- *Causes*: Optic nerve sheath meningioma (25% cases), rarely in optic nerve glioma and papilledema.

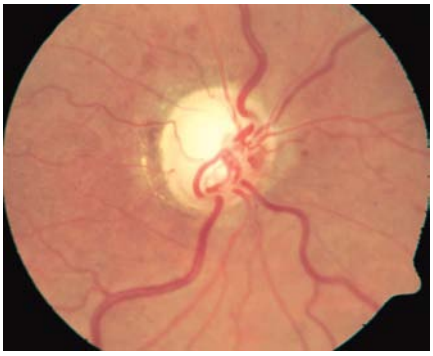


Fig. 11.14.3: Opticociliary shunt vessels

New Vessels on the Disk (NVD)

- Very common and associated with areas of capillary drop out
- Unilateral or bilateral, depending upon the cause
- Lace-like fine vessels, may be flat or elevated **(Figs 11.15.1 and 11.15.2)**

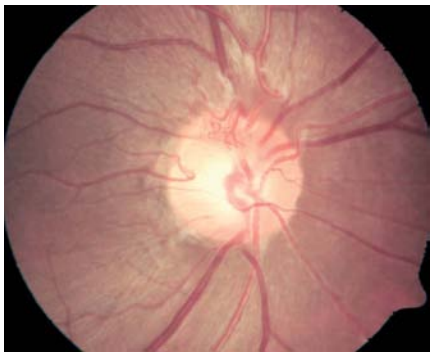


Fig. 11.15.1: Neovascularization of disk

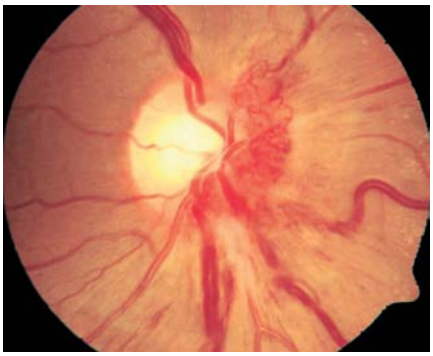


Fig. 11.15.2: Neovascularization of disk

- Vessels may extend beyond the peripapillary region **(Fig. 11.15.3)** and may be associated with vitreous hemorrhage **(Fig. 11.15.4)**
- Gliosis may be present in variable amount
- A fundus fluorescein angiography often important to confirm the diagnosis **(Figs 11.15.5A and B)**
- *Causes*: Proliferative diabetic retinopathy, retinal venous occlusion (CRVO/BRVO), central retinal vasculitis, retinal ischemia, etc.

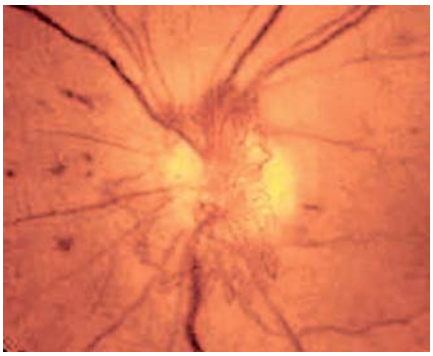


Fig. 11.15.3: Neovascularization of disk

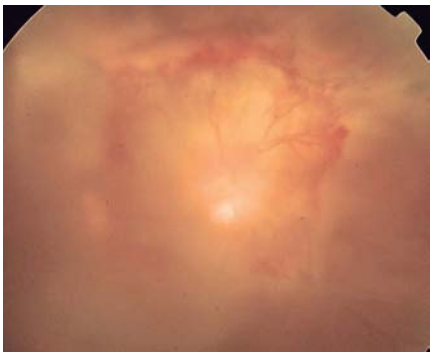


Fig. 11.15.4: Massive NVD with vitreous hemorrhage

Disk Hemorrhage

- Seen as splinter hemorrhage over the disk and at disk—retina junction **(Figs 11.16.1 to 11.16.5)**
- *Causes*: Papilledema, anterior ischemic optic neuropathy (AION), optic neuritis, open angle glaucoma, diabetic papillopathy, acute PVD, etc.

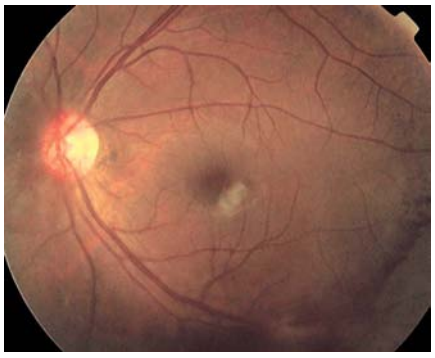


Fig. 11.15.5A: Neovascularization of the disk

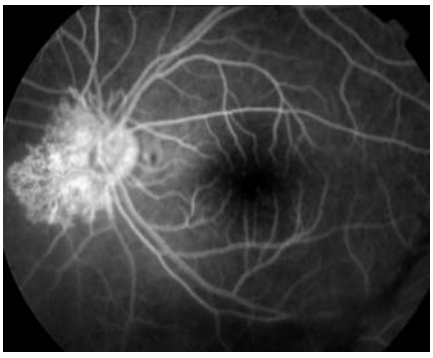


Fig. 11.15.5B: Neovascularization of the disk on FFA

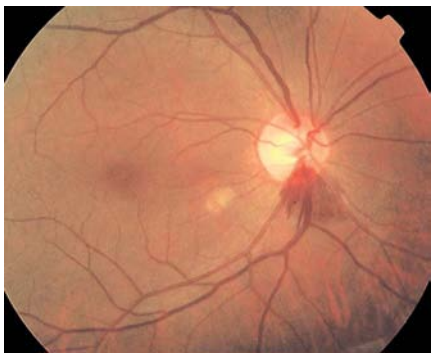


Fig. 11.16.1: Disk hemorrhage

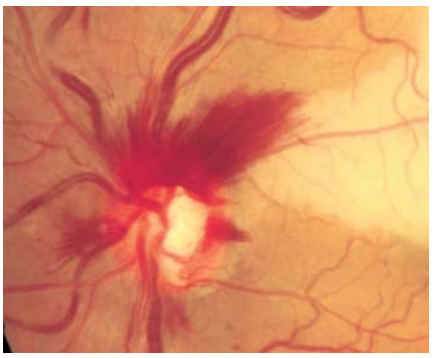


Fig. 11.16.2: Disk hemorrhage

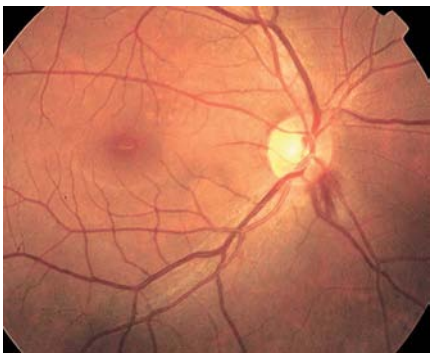


Fig. 11.16.3: Disk hemorrhage

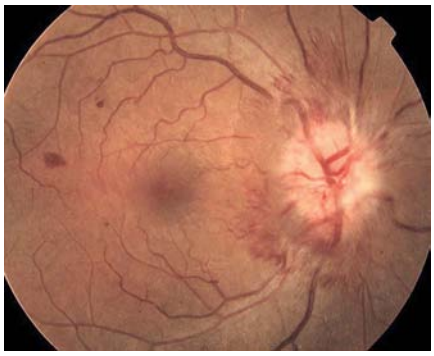


Fig. 11.16.4A: Disk hemorrhage—bilateral—RE

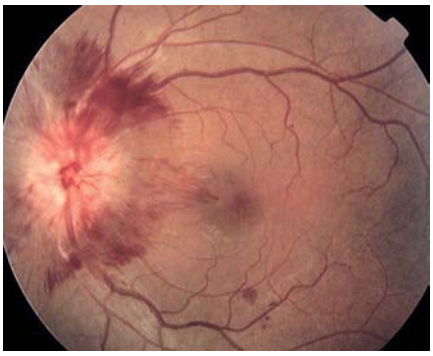


Fig. 11.16.4B: Disk hemorrhage—bilateral—LE

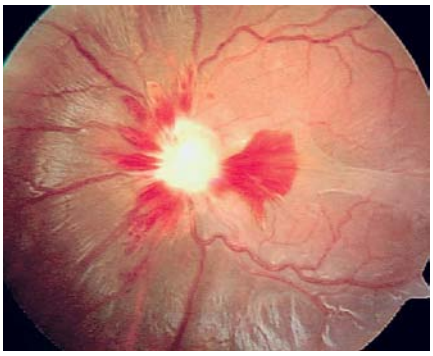


Fig. 11.16.5: Disk hemorrhage—post-traumatic

Dragged Vessels of the Disk

- Seen in proliferative retinopathies, e.g. PDR, sickle cell retinopathy, ROP, toxocara granuloma and certain hamartomas
- Dragging may be of any direction, but temporal dragging is common **(Figs 11.17.1 to 11.17.3)**.

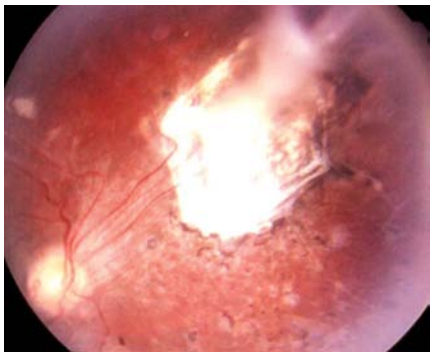


Fig. 11.17.1: Dragged vessels

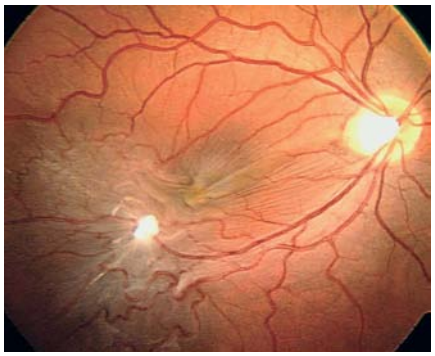


Fig. 11.17.2: Dragged vessels

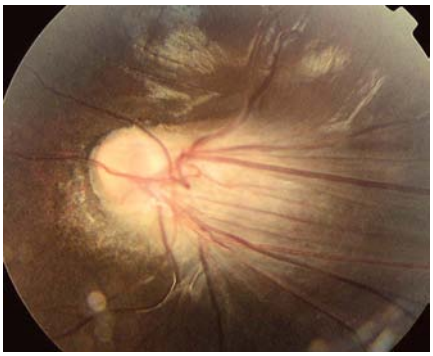


Fig. 11.17.3: Dragged vessels FEVR

Optic Neuritis

- Optic neuritis is an inflammatory or demyelinating disorder of the optic nerve (from the optic disk to the lateral geniculate body)
- Idiopathic type, or from demyelination, typically affects the patients between 20-40 years of age; but post-viral type typically occurs in children
- It may be *papillitis* (optic neuritis proper), *retrobulbar neuritis* and *neuroretinitis*
- *Signs of optic neuritis*:
  - Sudden visual loss and pupillary signs
  - Disk edema with obliteration of the physiological cup
  - Hyperemia and blurring of the disk margin **(Figs 11.18.1 and 11.18.2)**
  - Hemorrhages on the disk **(Fig. 11.18.3)**
  - Slit-like defect in the retinal nerve fiber layer **(Fig. 11.18.4)**
  - Inflammatory cells into the adjacent posterior vitreous

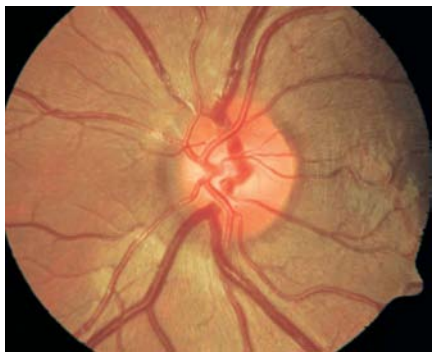


Fig. 11.18.1: Optic neuritis—hyperemia

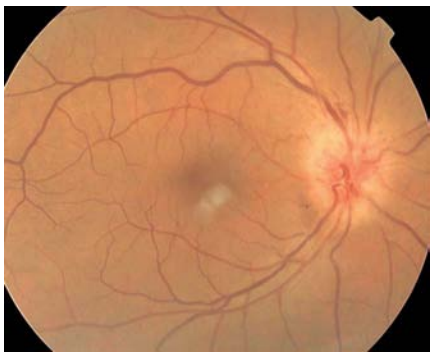


Fig. 11.18.2: Optic neuritis—disk edema

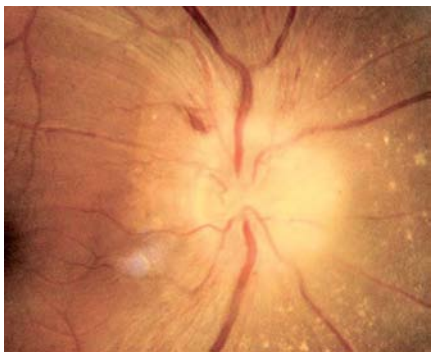


Fig. 11.18.3: Optic neuritis—disk hemorrhage

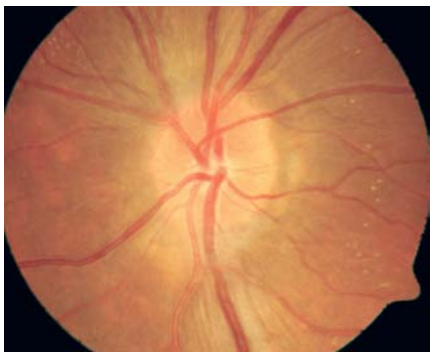


Fig. 11.18.4: Optic neuritis—slit-like defect in nerve fiber layer

- Glial tissue proliferation **(Fig. 11.18.5)**.

- *Sign of retrobulbar neuritis*
  - Here, the optic disk, and retinal nerve fiber layer are normal, as the site of involvement is behind the globe
  - Visual loss and pupillary sign (RAPD) are important.
- *Neuroretinitis*
  - Never associated with demyelinating diseases
  - Signs of optic neuritis
  - Macular star in addition **(Fig. 11.18.6)**
  - Juxtapapillary exudates may be seen.

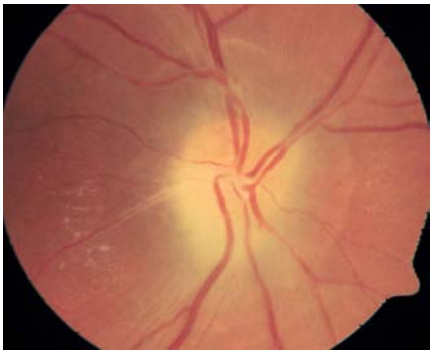


Fig. 11.18.5: Optic neuritis—glial tissue formation

Fig. 11.18.6: Neuroretinitis with macular star

Anterior Ischemic Optic Neuropathy (AION)

- *Two types*: Nonarteritic AION and arteritic AION
- *Nonarteritic AION*:
  - Nonarteritic AION is a segmental or generalized infarction of the anterior part of the optic nerve, caused by the occlusion of short posterior ciliary arteries
  - Usually unilateral, aged patient between 60-65 years
  - Pale, sectorial (usually upper part) disk edema which may be surrounded by splinter hemorrhages **(Figs 11.19.1 and 11.19.2)**
  - Associated with sudden altitudinal hemianopia (usually lower field).
- *Arteritic AION:*
  - Caused by giant cell arteritis, more elderly people
  - Unilateral severe loss of vision
  - A swollen, usually diffuse white or pale disk, with splinter hemorrhages around **(Figs 11.19.3 and 11.19.4)**
  - With time, the entire optic disk becomes pale **(Fig. 11.19.5)**
  - The fellow eye is frequently affected
  - Tender nodular temporal arteries and in severe cases there may be scalp necrosis.

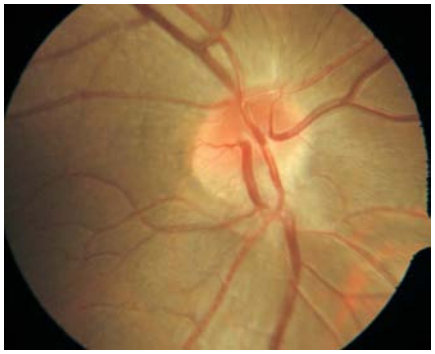


Fig. 11.19.1: Anterior ischemic optic neuropathy

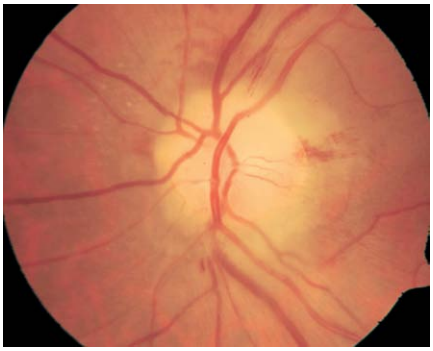


Fig. 11.19.2A: AION—splinter hemorrhage

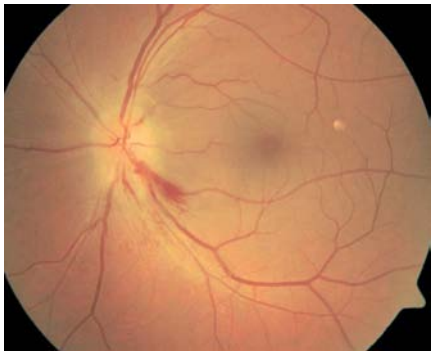


Fig. 11.19.2B: Nonarteritic AION—splinter hemorrhage

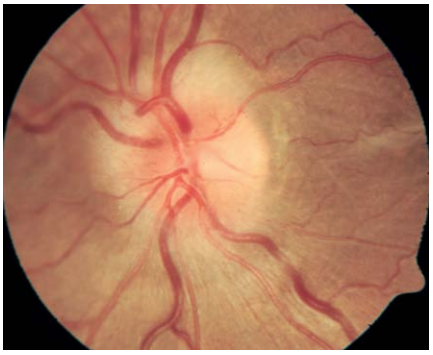


Fig. 11.19.3: Arteritic AION

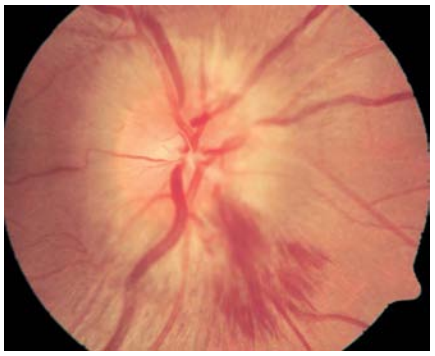


Fig. 11.19.4: Arteritic AION—splinter hemorrhage

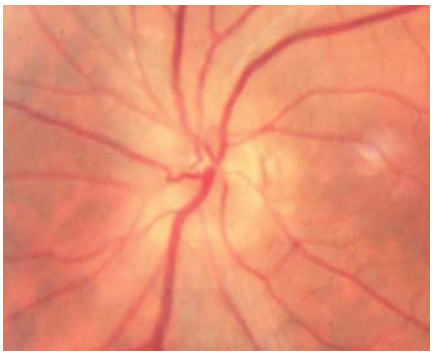


Fig. 11.19.5: Anterior ischemic optic neuropathy—pale disk

Other Causes of Unilateral Disk Edema

Central Retinal Venous Occlusion

- Unilateral condition, with moderate to severe visual loss in elderly patients
- Disk edema with splinter hemorrhages on the disk and peripapillary area **(Fig. 11.20.1)**
- Venous engorgement and widespread retinal hemorrhages **(Fig. 11.20.2)**
- Soft exudates may be present.

Central Vasculitis (Papillophlebitis)

- Similar to CRVO, but it affects in younger, otherwise healthy individuals **(Fig. 11.21.1)**
- Variable disk edema, venous engorgement, peripapillary and retinal hemorrhages **(Figs 11.21.2 to 11.21.4)**
- Prognosis is better than CRVO.

Long Standing Hypotony

- Seen after glaucoma filtration surgery, prolonged wound leak, chronic anterior uveitis, choroidal detachments etc. **(Figs 11.22.1A and B)**.

Posterior Scleritis

- Unilateral or bilateral condition with some systemic disease
- Disk edema without hemorrhage **(Fig. 11.23.1)**.

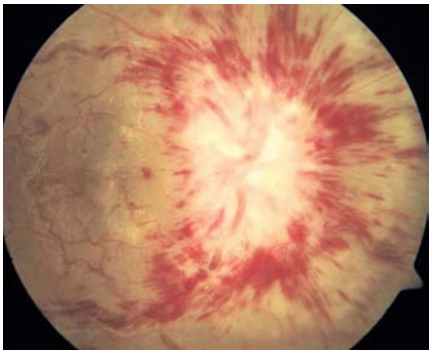


Fig. 11.20.1: Disk edema in central retinal venous thrombosis

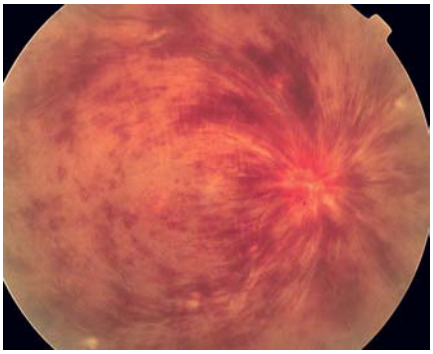


Fig. 11.20.2: Disk edema in central retinal venous thrombosis

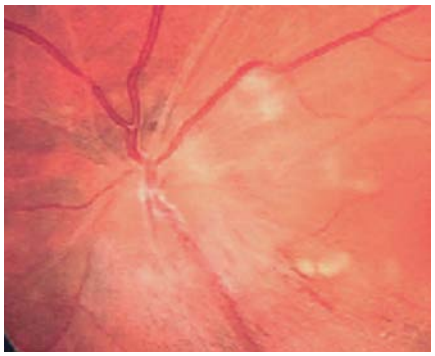


Fig. 11.21.1: Disk edema in central vasculitis

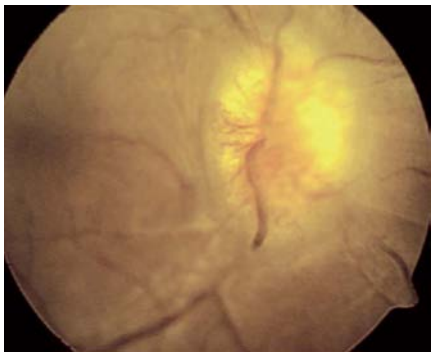


Fig. 11.21.2: Disk edema in central periphlebitis (red free)

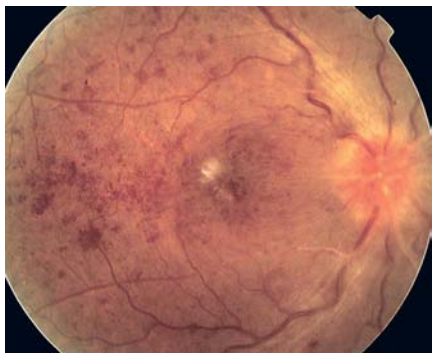


Fig. 11.21.3: Disk edema in central periphlebitis

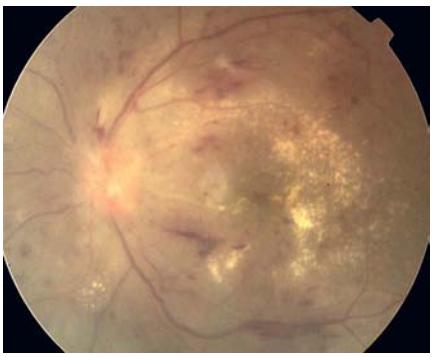


Fig. 11.21.4: Disk edema in central vasculitis

Fig. 11.22.1A: Disk edema—long standing hypotony due to overfiltering bleb

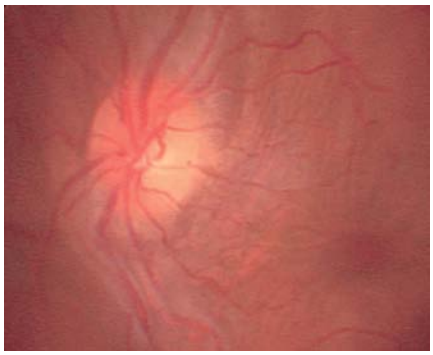


Fig. 11.22.1B: Disk edema—long standing hypotony same eye

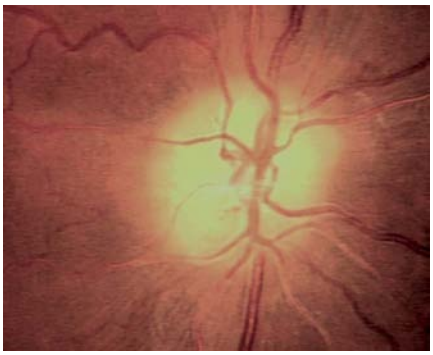


Fig. 11.23.1: Disk edema in posterior scleritis

Papilledema

- Papilledema is the bilaterial, non-inflammatory passive swelling of the optic disk, produced by raised intracranial tension (ICT)
- Unilateral papilledema with optic atrophy on the other side suggests a frontal lobe tumor or olfactory meningioma of the opposite side—the Foster-Kennedy syndrome **(Figs 11.24.1A and B)**
- The signs of papilledema depend on duration and its severity
- *Four stages*: Early, established, chronic and atrophic.

Early Papilledema

- Disk hyperemia, elevation and preservation of optic cup **(Figs 11.24.2A and B)**
- Blurring of the nasal sector of disk margin first, then the superior and inferior margins **(Figs 11.24.3A and B)**
- Splinter hemorrhages at or just off the disk—margin **(Fig. 11.24.4)**
- Absent spontaneous venous pulsation (also absent in 20% of general population).

Established (Acute) Papilledema

- Disk elevation is marked
- Entire disk margins become indistinct and central cup is obliterated **(Figs 11.24.5A and B)**

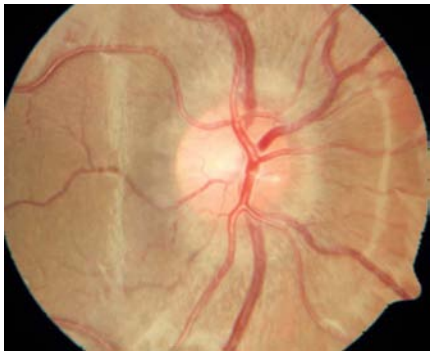


Fig. 11.24.1A: Foster-Kennedy syndrome—papilledema—RE

Fig. 11.24.1B: Foster-Kennedy syndrome—optic atrophy—LE

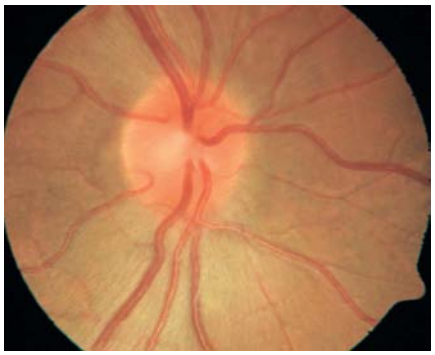


Fig. 11.24.2A: Early papilledema—blurring of nasal disk margin—RE

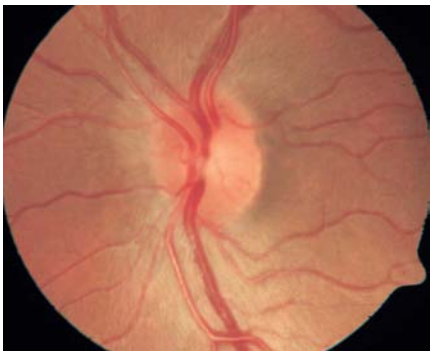


Fig. 11.24.2B: Early papilledema—blurring of nasal disk margin—LE

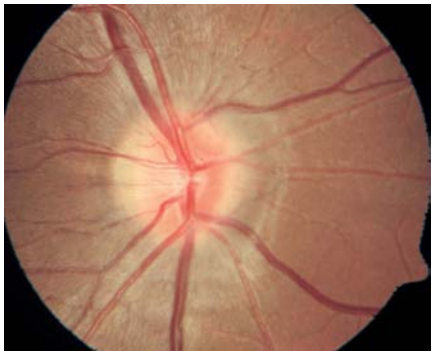


Fig. 11.24.3A: Early papilledema—more blurring—RE

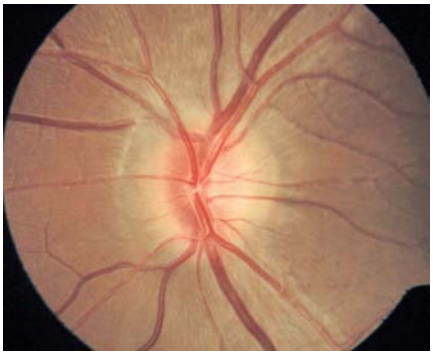


Fig. 11.24.3B: Early papilledema—more blurring—LE

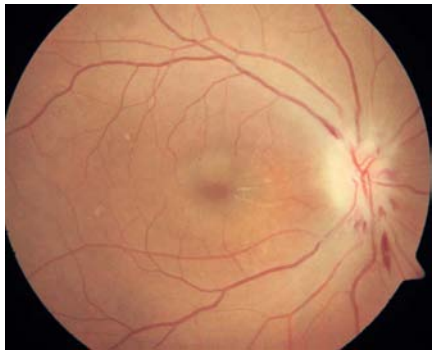


Fig. 11.24.4: Early papilledema—flame shaped hemorrhage

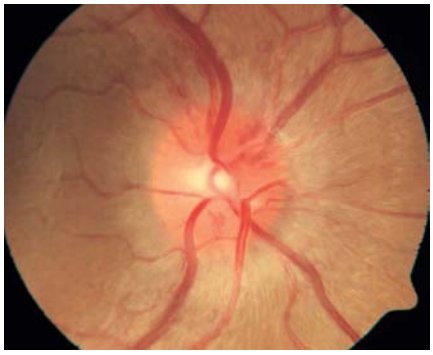


Fig. 11.24.5A: Established papilledema—RE

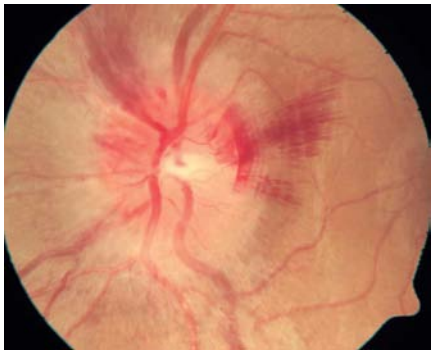


Fig. 11.24.5B: Established papilledema—flame shaped hemorrhage—LE

- Venous engorgement, flame-shaped hemorrhage and peripapillary edema with macular star **(Fig. 11.24.6)**
- *Patton's line*—peripapillary and perifoveal circumferential retinal folds due to accumulation of fluid **(Figs 11.24.7A and B)**

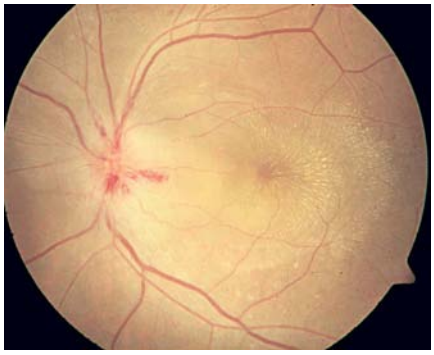


Fig. 11.24.6: Established papilledema—macular star

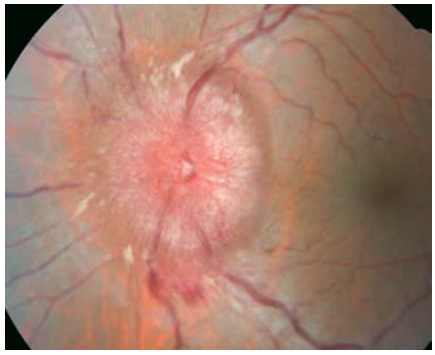


Fig. 11.24.7A: Established papilledema—disk hemorrhage and Patton's line

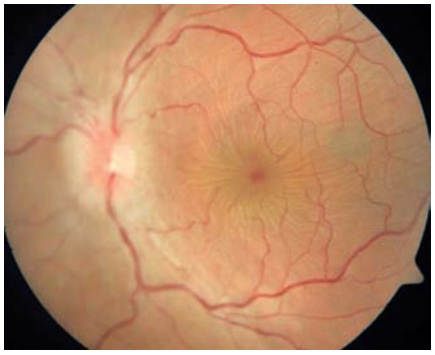


Fig. 11.24.7B: Established papilledema—perifoveal radial folds

Chronic Papilledema

- The hemorrhagic and exudative components gradually resolve **(Figs 11.24.8A and B)**
- Optic disk appears as a '*champagne cork*' like elevation **(Figs 11.24.9A and B)**
- Opticociliary shunt may be visible
- A macular star may be present.

Atrophic Papilledema

- Dirty-white appearance of the optic disk, due to reactive gliosis—leading to secondary optic atrophy **(Figs 11.24.10A and B)**

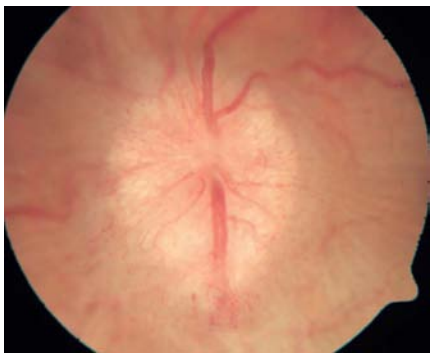


Fig. 11.24.8A: Chronic papilledema—RE

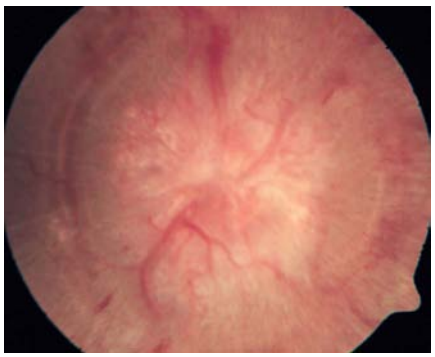


Fig. 11.24.8B: Chronic papilledema—LE

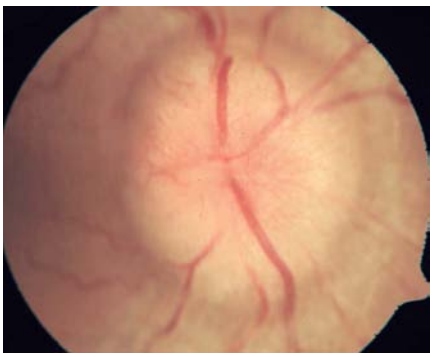


Fig. 11.24.9A: Chronic papilledema—champagne cork appearance—RE

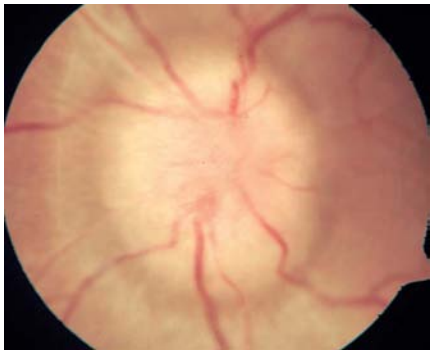


Fig. 11.24.9B: Chronic papilledema—champagne cork appearance—LE

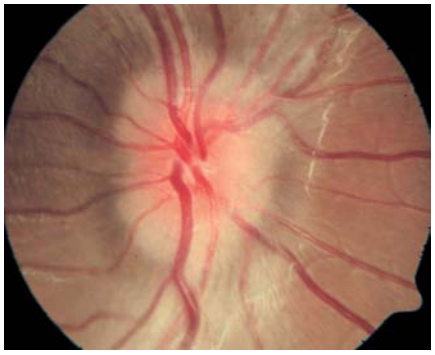


Fig. 11.24.10A: Atrophic papilledema—RE

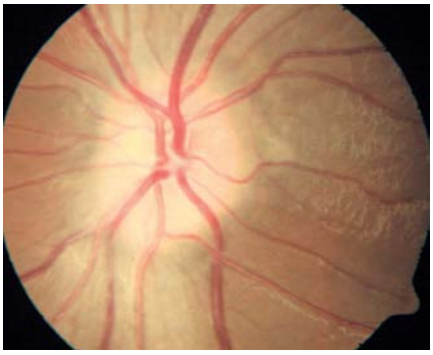


Fig. 11.24.10B: Atrophic papilledema—LE

- Retinal vessels are attenuated with perivascular sheathing **(Figs 11.24.11A and B)**
- Peripapillary pigmentary changes.

Other Causes of Bilateral Disk Edema

Malignant Hypertension

- Presence of severe rise in blood pressure
- More in secondary hypertension
- Bilateral disk edema, flame-shaped hemorrhage, soft exudates and a macular star **(Figs 11.25.1A and B)**
- Variable amount of hypertensive retinal changes
- Peripheral pale areas of choroidal infarcts.

Other Causes

- Compressive thyroid optic neuropathy
- Acute methyl alcohol toxicity
- Bilateral optic neuritis.

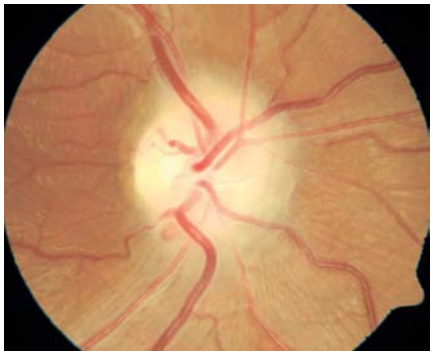


Fig. 11.24.11A: Atrophic papilledema—reactive gliosis—perivascular sheathing

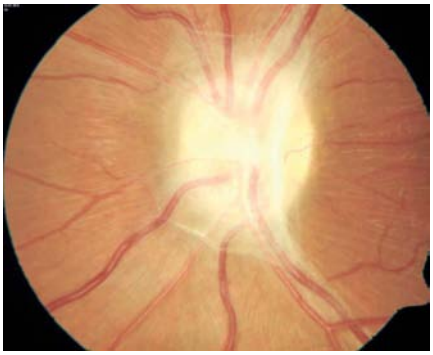


Fig. 11.24.11B: Atrophic papilledema—reactive gliosis—perivascular sheathing

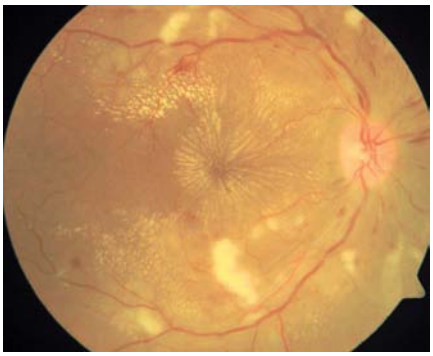


Fig. 11.25.1A: Malignant hypertensive retinopathy—RE

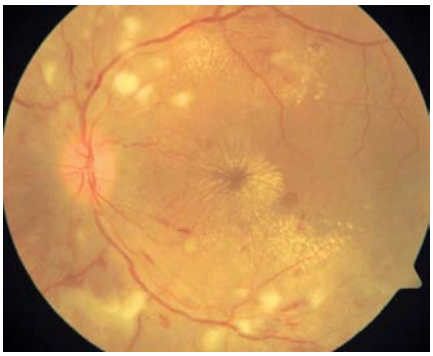


Fig. 11.25.1B: Malignant hypertensive retinopathy—LE

Optic Atrophy

- Degeneration of optic nerve fibers with loss of their myelin sheaths, and characterized by the pallor of the optic disk
- 'Pallor' of the disk is not due to atrophy of the nerve fibers, but due to loss of vascularity owing to obliteration of the disk capillaries
- *Three types:* Primary, secondary and consecutive.

Primary Optic Atrophy

- Usually bilateral, pallor of the disk, with white or gray color, without significant gliosis
- Disk is flat, margin is sharply defined, and peripapillary retina is normal looking **(Figs 11.26.1A and B)**
- Reduction in number of small vessels crossing the disk **(Fig. 11.26.2)**
- Thinning of nerve fiber layer.

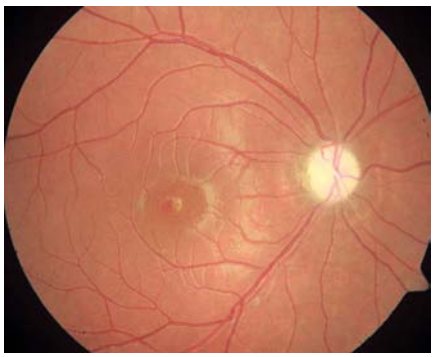


Fig. 11.26.1A: Primary optic atrophy—bilateral—RE

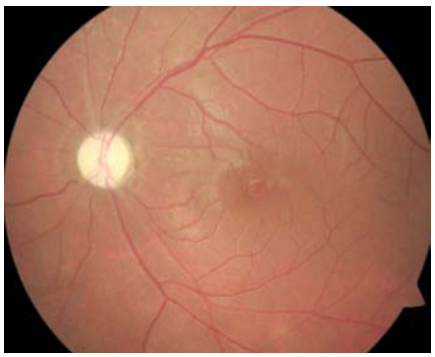


Fig. 11.26.1B: Primary optic atrophy—bilateral—LE

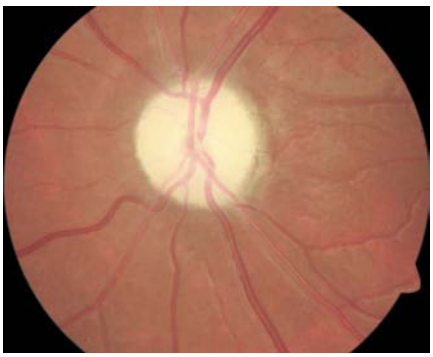


Fig. 11.26.2: Primary optic atrophy—reduction in number of small vessels on the disk

Secondary Optic Atrophy

- Preceded by edema of the optic nerve head
- Dirty-gray color with blurred margins
- Cup is full, and lamina cribrosa is obscured **(Figs 11.27.1A and B)**
- Narrowing of the blood vessels with sheathing **(Fig. 11.27.2)**
- Typically seen in after papilledema, optic neuritis or AION.

Consecutive Optic Atrophy

- Yellowish-waxy pallor of the disk **(Fig. 11.28.1)**
- Disk margins are less sharply defined
- Marked narrowing or even obliteration of arterioles **(Fig. 11.28.2)**
- Typically seen in retinitis pigmentosa **(Figs 11.28.2 and 11.28.3)**

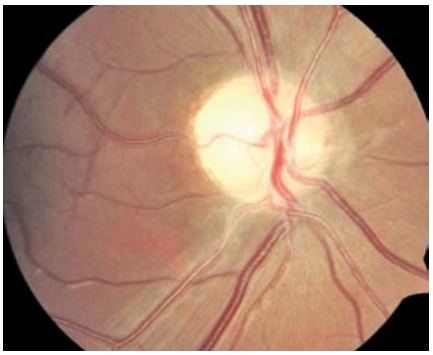


Fig. 11.27.1A: Secondary (postpapilledemic) optic atrophy

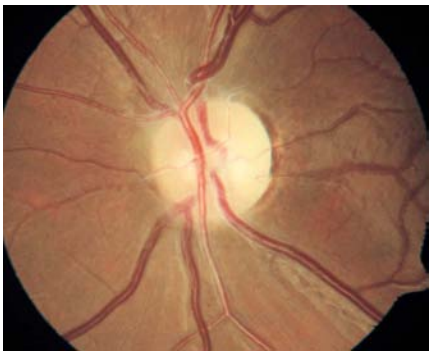


Fig. 11.27.1B: Secondary (postpapilledemic) optic atrophy

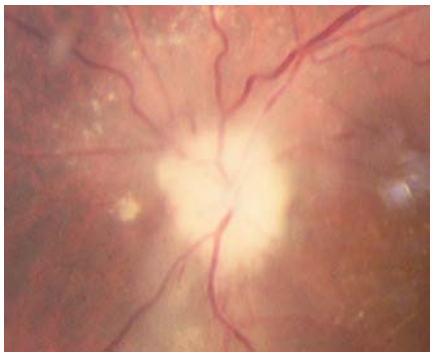


Fig. 11.27.2: Secondary (postneuritic) optic atrophy

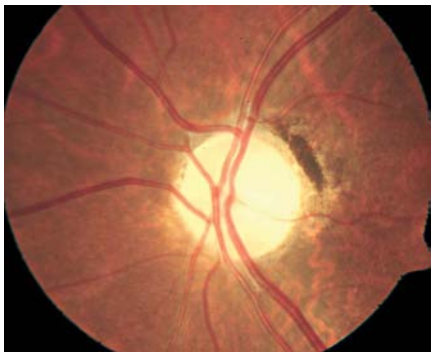


Fig. 11.28.1: Consecutive optic atrophy—waxy pallor

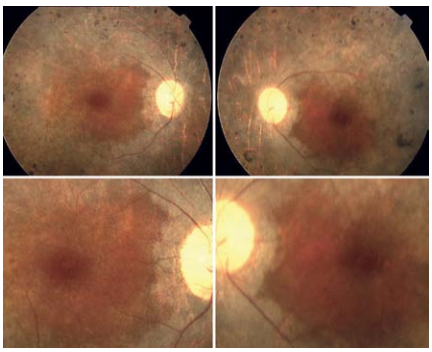


Fig. 11.28.2: Consecutive optic atrophy

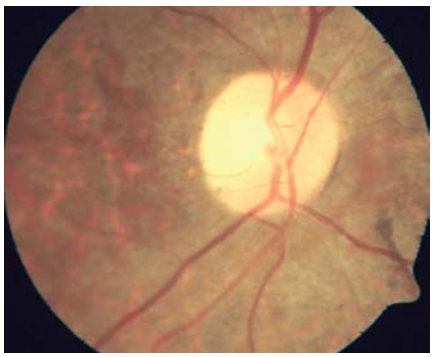


Fig. 11.28.3A: Consecutive optic atrophy—retinitis pigmentosa—RE

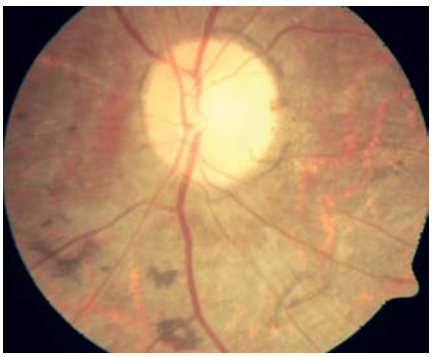


Fig. 11.28.3B: Consecutive optic atrophy—retinitis pigmentosa—LE

Glaucomatous Optic Atrophy (Figs 11.29.1 and 11.29.2; see Figs 9.6.15 and 9.6.16)

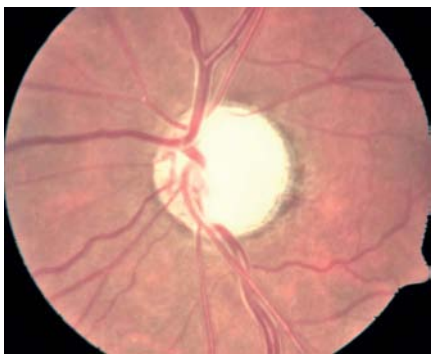


Fig. 11.29.1: Glaucomatous optic atrophy

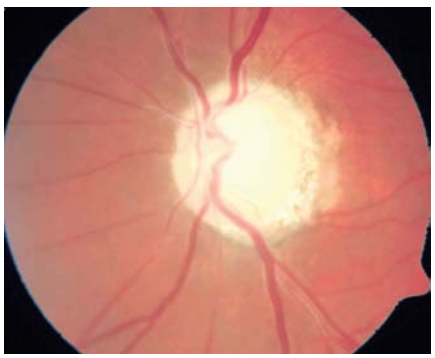


Fig. 11.29.2: Glaucomatous optic atrophy

Temporal Pallor

- Form of partial optic atrophy, involve a loss of temporal fibers including the papillomacular bundles results in *temporal pallor* **(Figs 11.30.1A and B)**
- Temporal side is normally relatively pale, because the retinal vessels emerge from the nasal side and the temporal side is normally less vascular.

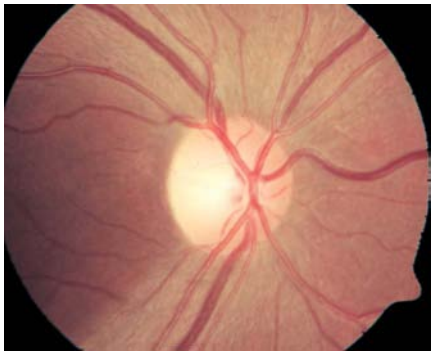


Fig. 11.30.1A: Temporal pallor—RE

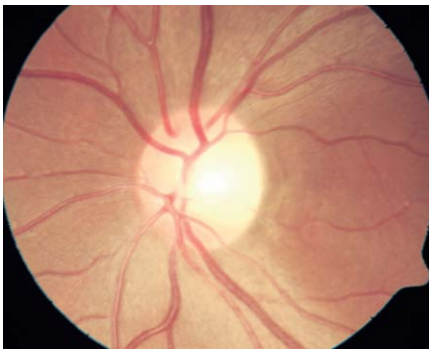


Fig. 11.30.1B: Temporal pallor—LE

Primary Optic Disk Tumors

Optic Disk Capillary Hemangioma

- Rare, unilateral, may also involve the retina
- 25% patients have von Hippel-Lindau syndrome
- Orange-red lesion of the optic disk with macular hard exudates **(Figs 11.31.1A to C)**
- Peripheral retinal angiomas or grossly dilated blood vessels.

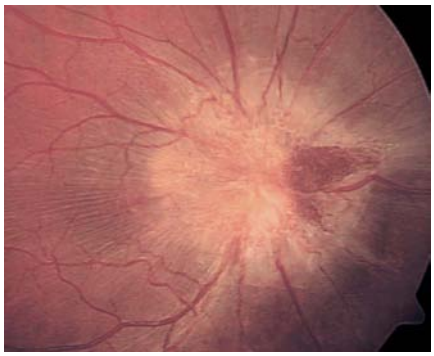


Fig. 11.31.1A: Optic disk hemangioma

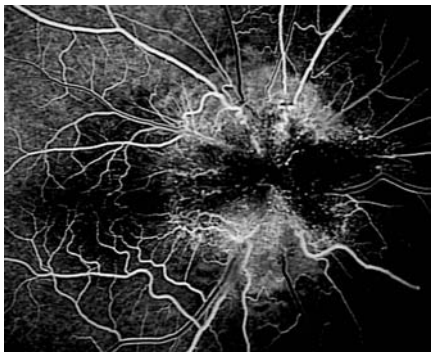


Fig. 11.31.1B: Optic disk hemangioma on FFA in early phase

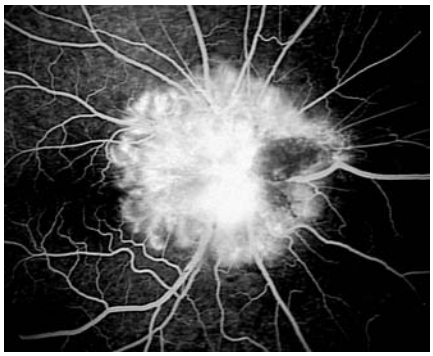


Fig. 11.31.1C: Optic disk hemangioma on FFA in late phase

Optic Disk Cavernous Hemangioma

- Easily recognized by its characteristic appearance of saccular "grape-like" lesions **(Fig. 11.32.1)**
- It is a vascular hamartoma composed of clusters of thin-walled saccular aneurysms partially filled with dark venous blood
- It can grow on the disk and can cause vitreous hemorrhage severe enough to require vitrectomy.

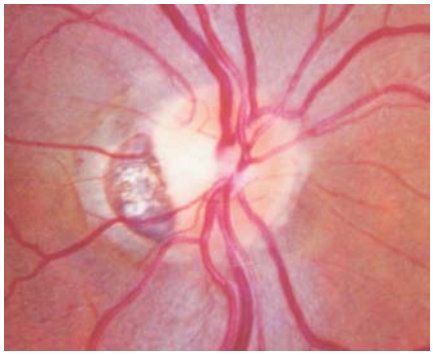


Fig. 11.32.1: Optic disk cavernous hemangioma

Arteriovenous Malformation on Optic Disk

- Tortuosity of the blood vessels may be a normal finding **(Figs 11.33.1A and B)**. May be seen in high hypermetropia, venous stasis retinopathy, papilledema, etc.
- Abnormally dilated retinal vessels accompanied by an abnormal or absent capillary system **(Figs 11.33.2A and B)**
- When arteriovenous communications are large, visual acuity may be severely affected **(Figs 11.33.3A and B)**
- Patients with more severe retinal involvement are more likely to have cerebral or periorbital arteriovenous abnormalities, which comprises the Wyburn-Mason syndrome.

Optic Disk Melanocytoma

- Rare unilateral, benign tumor in pigmented races
- Pigmented disk lesion frequently located eccentrically **(Figs 11.34.1 and 11.34.2)**
- FFA confirms the diagnosis **(Figs 11.34.3A and B)**
- May induce disk swelling in some cases **(Fig. 11.34.4)**
- May interfere with vision.

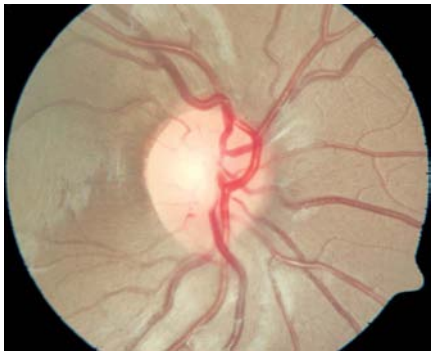


Fig. 11.33.1A: Abnormal tortuosity of the disk and retinal blood vessels—RE

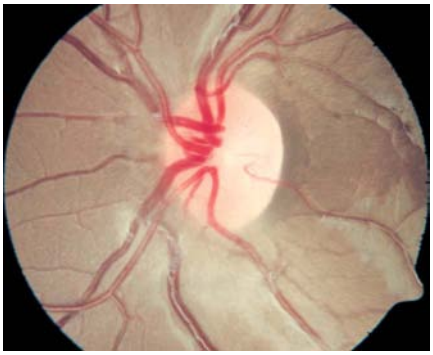


Fig. 11.33.1B: Abnormal tortuosity of the disk and retinal blood vessels—LE

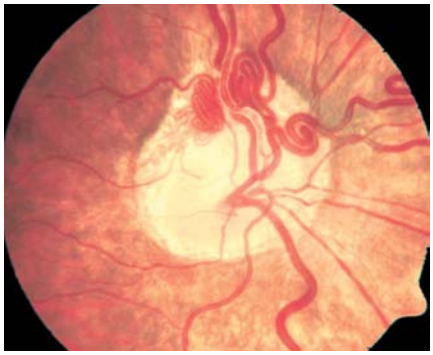


Fig. 11.33.2A: Arteriovenous malformation on the optic disk—RE

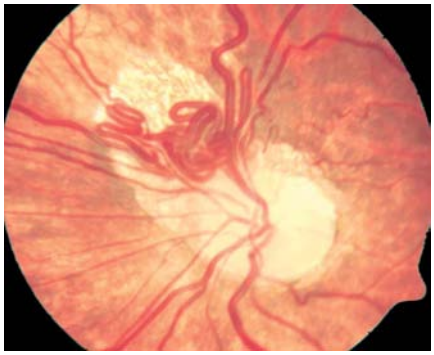


Fig. 11.33.2B: Arteriovenous malformation on the optic disk—LE

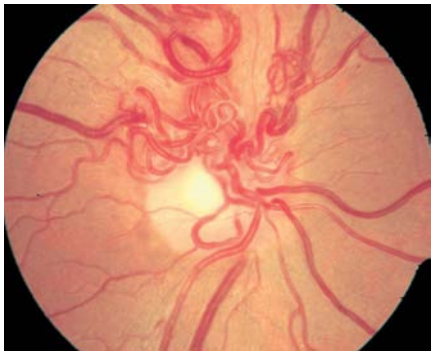


Fig. 11.33.3A: Arteriovenous malformation on the optic disk—more involvement—RE

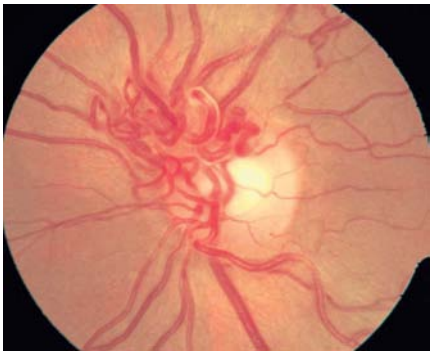


Fig. 11.33.3B: Arteriovenous malformation on the optic disk—LE

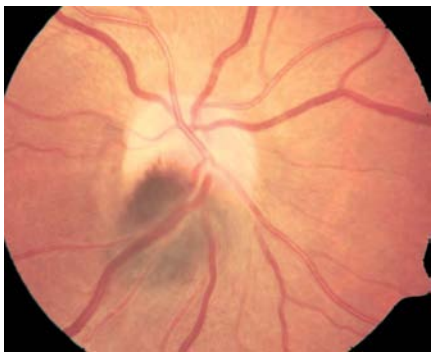


Fig. 11.34.1: Optic disk melanocytoma

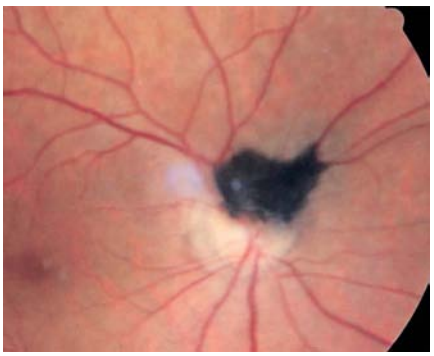


Fig. 11.34.2: Optic disk melanocytoma

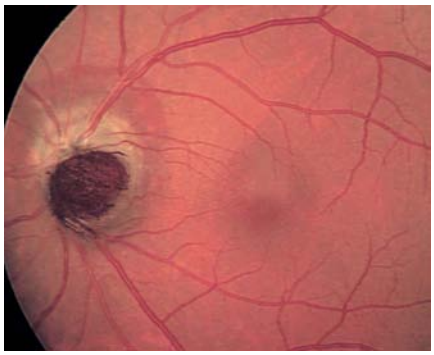


Fig. 11.34.3A: Optic disk melanocytoma

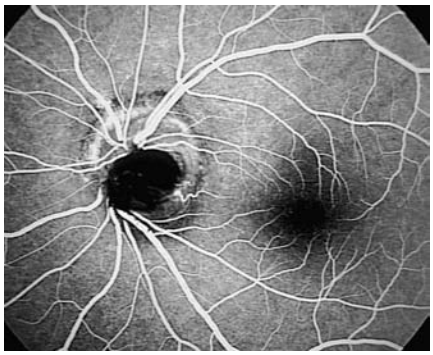


Fig. 11.34.3B: Optic disk melanocytoma—FFA

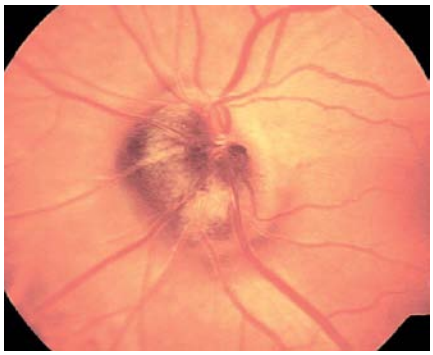


Fig. 11.34.4: Optic disk melanocytoma with disk swelling

Astrocytoma of the Optic Disk

- Unilateral giant drusen, a benign condition
- Frequently associated with adenoma sebaceum of the face in tuberous sclerosis or *epiloia* **(Fig. 11.35.1A)**
- Semitransparent, mulberry-like lesion that displays autofluorescence **(Fig. 11.35.1B)**.

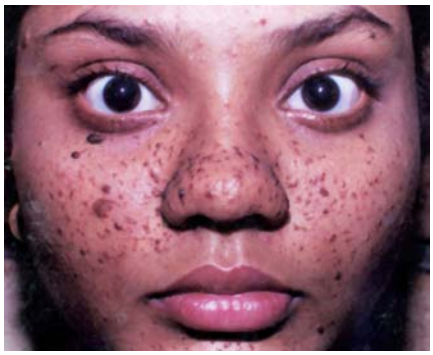


Fig. 11.35.1A: Tuberous sclerosis—adenoma sebaceum

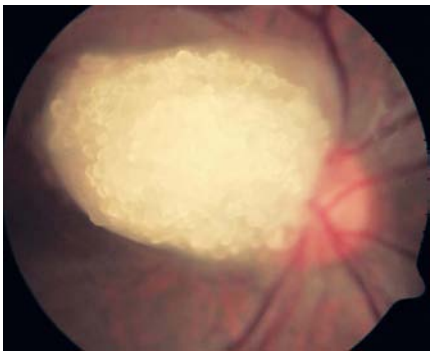


Fig. 11.35.1B: Tuberous sclerosis—mulberry-like astrocytoma

Optic Nerve Glioma and Meningioma

- They present as proptosis and ***see Chapter 13***
- In late stage there is pallor of the disk and primary optic atrophy
- Long-standing visual impairment, a pale swollen disk, and opticociliary shunt vessels is pathognomonic of optic nerve sheath meningioma.

Infiltrative Lesions

Retinoblastoma, choroidal melanoma, leukemia, metastatic carcinoma **(Fig. 11.36.1)**, toxocara infection **(Fig. 11.36.2)**, cytomegalovirus (CMV) infection **(Fig. 11.36.3)**, etc.

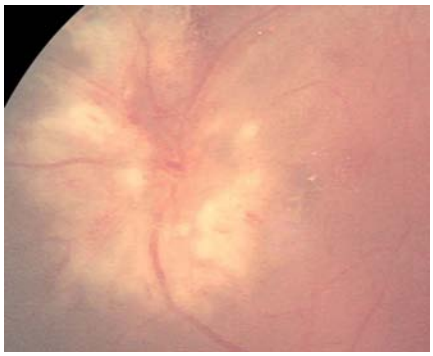


Fig. 11.36.1: Optic disk infiltration—metastatic deposits

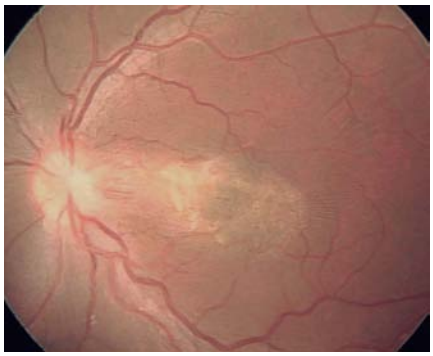


Fig. 11.36.2: Optic disk infiltration—toxocara

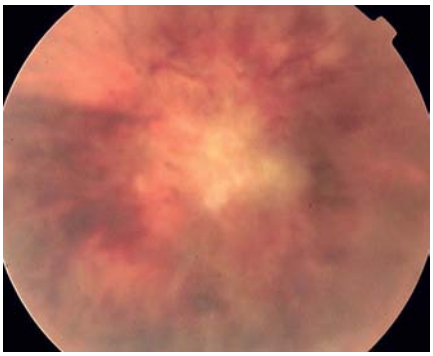


Fig. 11.36.3: Optic disk infiltration—CMV retinitis

CHAPTER

12 Diseases of the Retina

CHAPTER OUTLINE

Congenital Retinal Disorders

- Myelinated nerve fibers
- Phakomatosis
- Albinism
- Retinochoroidal coloboma

Retinal Vascular Disorders

- Central retinal artery occlusion (CRAO)
- Central retinal venous occlusion (CRVO)
  - Nonischemic CRVO
  - Ischemic CRVO
  - Venous stasis retinopathy
- Branch retinal venous occlusion (BRVO)
- Preretinal hemorrhage
- Roth's spot
- Subretinal hemorrhage
- Sub-RPE hemorrhage
- Choroidal hemorrhage
- Neovascularization
- Collaterals
- Retinal angiomas
- Retinal vasculitis
- Frosted branch angiitis

Retinal Exudates

- Hard exudates
- Macular stars
- Soft exudates or cotton wool spots
- Subretinal exudates
- Coats' diseases

Inflammatory Retinal Lesions

- Multiple evanescent white dot syndrome (MEWDS)
- Punctate inner choroidopathy
- Birdshot retinochoroidopathy
- Acute posterior multifocal placoid pigment epitheliopathy
- Multifocal choroiditis with panuveitis
- Disseminated choroiditis
- Serpiginous choroidopathy
- Focal toxoplasmosis
- Toxocariasis
- Candidiasis
- Cytomegalovirus (CMV) retinitis
- Acute retinal necrosis (ARN)
- Progressive outer retinal necrosis (PORN)
- Vogt-Koyanagi-Harada syndrome
- Sympathetic ophthalmia
- Tuberculous choroiditis

Macular Lesions (Maculopathies)

Hereditary Maculopathies
- Stargardt's heredomacular dystrophy
- Central areolar choroidal dystrophy
- Best's vitelliform macular dystrophy
- Hereditary cone dystrophy (Bull's eye maculopathy)
- Myopic maculopathies
- Dry age-related macular degenerations

Exudative Maculopathies
- Cystoid macular edema (CME)
- Central serous retinopathy (CSR)
- RPE detachment
- Wet age-related macular degeneration
- Macular holes

Other Maculopathies
- Cellophane maculopathy
- Commotio retinae (Berlin's edema)
- Crystalline maculopathy

Multiple Flecked Retinal Lesions

- Familial dominant drusen
- Doyne's honeycomb dystrophy
- Fundus flavimaculatus
- Fundus albipunctatus
- Benign flecked retina syndrome
- Pattern macular dystrophy
- Hard drusens
- Soft dursens
- Cuticular (basal laminar) drusen

Pigmentary Retinopathy

- Typical retinitis pigmentosa
- Atypical retinitis pigmentosa

Retinal Detachment

- Rhegmatogenous retinal detachment (RRD)
- Associated predisposing factors
- Tractional retinal detachment
- Exudative retinal detachment

Hypertensive Retinopathies

Diabetic Retinopathy

- Nonproliferative diabetic retinopathy (NPDR)
- Diabetic maculopathy
- Proliferative diabetic retinopathy (PDR)
- Advanced proliferative diabetic retinopathy

Retinopathy of Prematurity

Retinoblastoma

Miscellaneous Retinal Conditions

- Idiopathic polypoidal choroidal vasculopathy
- Familial exudative vitreoretinopathy
- Stickler's diseases
- Idiopathic juxtafoveal telangiectasis
- Goldmann-Favre syndrome
- Congenital hypertrophy of retinal pigment epithelium (CHRPE)
- Retinoschisis
- Purtscher's retinopathy
- Retinal cysts

Congenital Retinal Disorders

Myelinated Nerve Fibers

- Myelinated (opaque) nerve fibers is a congenital condition
- Present in one percent of normal population
- Appear as white patches with radial striations at peripheral edges—feathery appearance ***(See more in Chapter 11)***
- Usually peripapillary **(Figs 12.1.1A and B),** or sometimes peripheral and isolated **(Fig. 12.1.2)**
- Rarely, it is extensive and distributed along the retinal nerve fiber layer **(Figs 12.1.3 and 12.1.4)**
- *No treatment is required.*

Phakomatosis

- A group of conditions (hamartomas) in which there are congenital, disseminated, usually benign tumors of the blood vessels or neural tissues
- Often ocular, cutaneous, and intracranial in location
- *Neurofibromatosis*
- (von Recklinghausen's disease)
  - Often hereditary in nature **(Fig. 12.2.1A)**
  - Most common type, with typical subcutaneous nodules **(see Figs 6.34.1A and B)** and café-au-lait spots
  - Iris nodule—Lisch's nodules **(see Figs 6.34.2A and B)** and Lisch spots **(*see* Fig. 6.34.3)**
  - Plexiform tumors of lids with ptosis of the upper lid, ectropion of the lower lid thickened corneal nerves **(Figs 12.2.1B and C).**

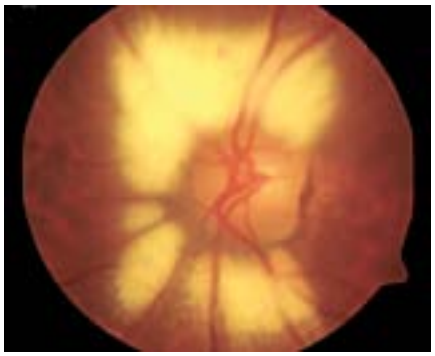


Fig. 12.1.1A: Myelinated nerve fibers

Fig. 12.1.1B: Myelinated nerve fibers

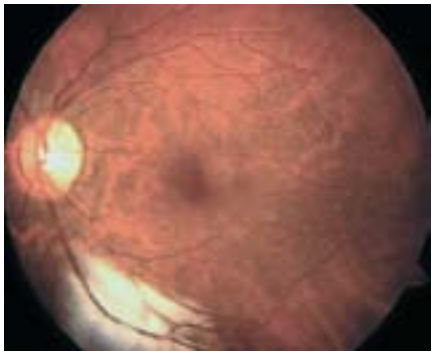


Fig. 12.1.2: Myelinated nerve fibers—isolated in the periphery

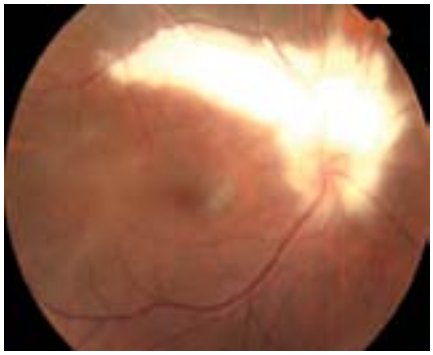


Fig. 12.1.3: Myelinated nerve fibers—along the nerve fiber

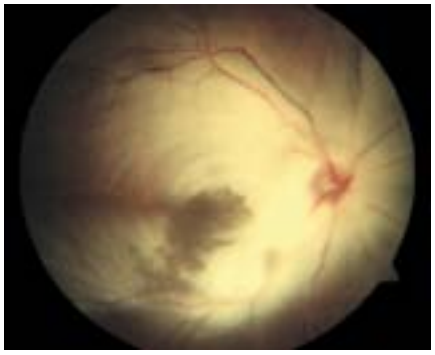


Fig. 12.1.4: Myelinated nerve fibers—extensive along the RNFL

Fig. 12.2.1A: Neurofibromatosis—father (nodular type) and son (plexiform type)

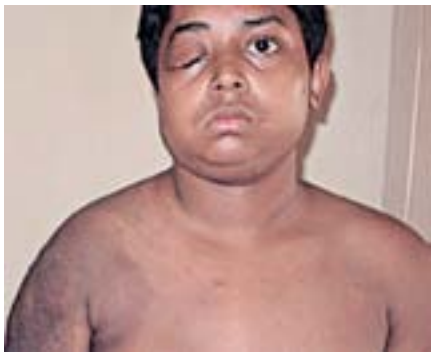


Fig. 12.2.1B: Plexiform neurofibroma with ptosis—RE

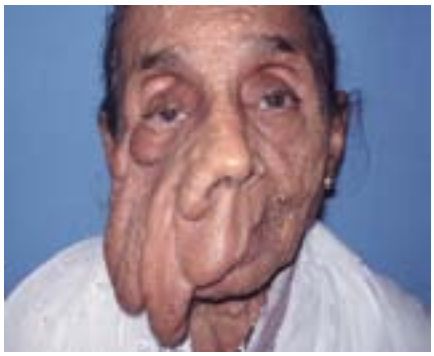


Fig. 12.2.1C: Plexiform neurofibromatosis

- *Tuberous sclerosis*
- (Bourneville's disease)
  - Diagnostic triad are epilepsy, mental retardation and adenoma sebaceum **(Fig. 12.2.2A)**
  - Called *'epiloia'*: *epi* (epilepsy), *loi* (low IQ), and *a* (adenoma sebaceum)
  - *Ocular lesion:* Optic disk astrocytoma **(Fig. 12.2.2B)** and also of the retina **(Figs 12.2.3 and 12.2.4)**
  - *See more in Chapter 11.*
- *Angiomatosis retinae* (von Hippel-Lindau disease)
  - A reddish, slightly elevated tumor is seen in the retina which is nourished by dilated large retinal artery and vein **(Fig. 12.2.5)**, and with surrounding hard exudates **(Fig. 12.2.6)**.

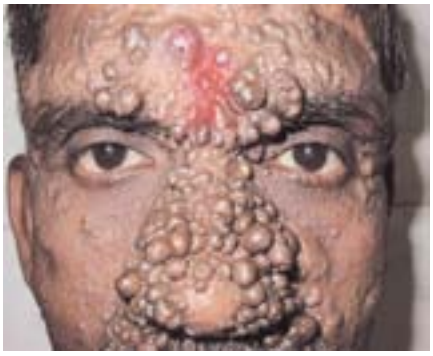


Fig. 12.2.2A: Tuberous sclerosis—adenoma sebaceum

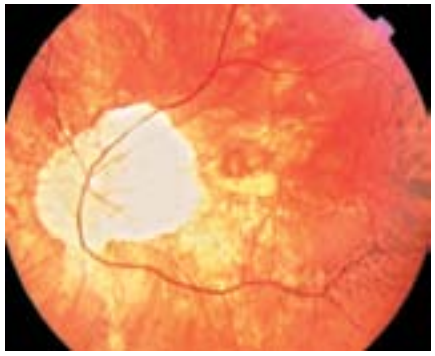


Fig. 12.2.2B: Optic disk astrocytoma

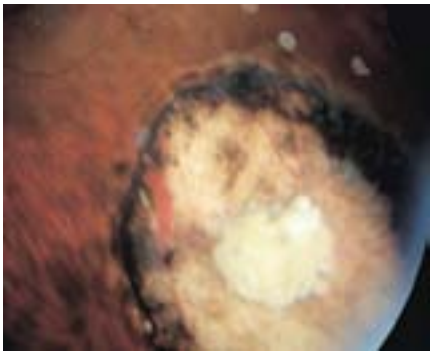


Fig. 12.2.3: Retinal astrocytoma

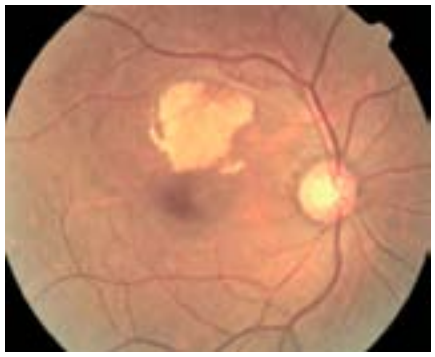


Fig. 12.2.4: Retinal astrocytoma

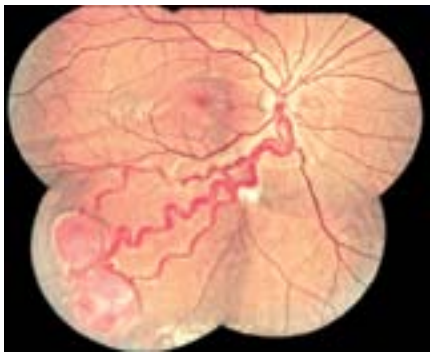


Fig. 12.2.5: Angiomatosis retinae—von Hippel-Lindau disease

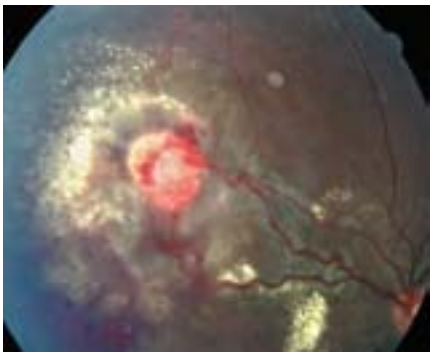


Fig. 12.2.6: Angiomatosis retinae—von Hippel-Lindau disease

- *Encephalotrigeminal angiomatosis* (Sturge-Weber syndrome)
  - Port-wine stain along the distribution of the trigeminal nerve **(Figs 12.2.7 and see 1.55.1 and 9.2.3)**
  - Choroidal hemangioma which may result in a congenital glaucoma **(see Figs 6.39.2A and B)**
  - *See more in Chapter 6.*

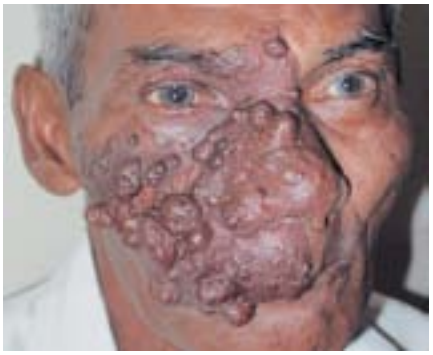


Fig. 12.2.7: Sturge-Weber syndrome

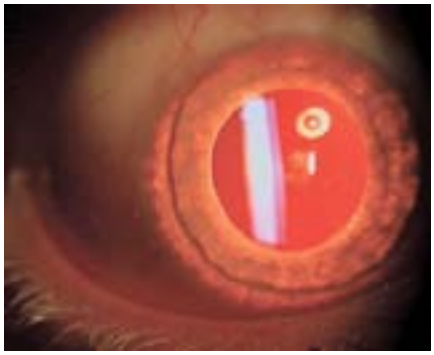


Fig. 12.3.1: Albinism—pink fundal glow

Albinism

- Pink fundal glow **(Fig. 12.3.1)**
- Generalized orangish color of the fundus with visible choroidal vasculature **(Figs 12.3.2A and B)**
- Positive iris transillumination
- *See more in Chapter 6* ***(Figs 6.3.1 to 6.3.5)***.

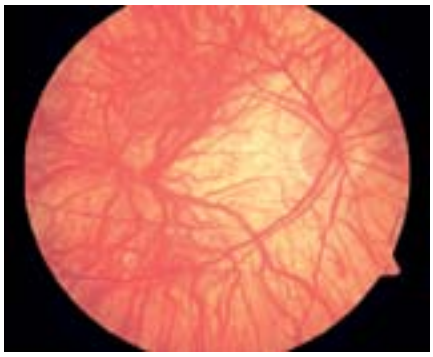


Fig. 12.3.2A: Albino fundus—orangish color with visible choroidal vasculature—RE

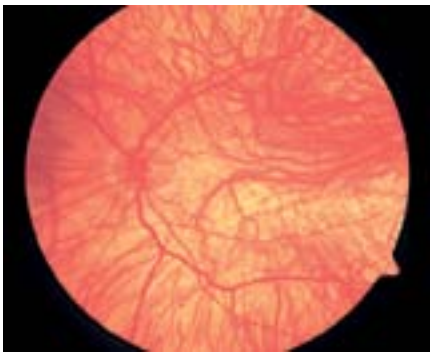


Fig. 12.3.2B: Albino fundus—orangish color with visible choroidal vasculature—LE

Retinochoroidal Coloboma

- Large oval, or semicircular sharply-defined white area inferior to the optic disk **(Figs 12.4.1A and B)**
- Sometimes, it may include the disk **(see Fig. 11.6.4)**
- May appear as isolated coloboma **(Fig. 12.4.2)** with a bridge of normal retinochoroidal tissue in between, giving rise to "double-disk" **(Fig. 12.4.3)** or "triple-disk" **(Fig. 12.4.4)** appearance
- May be associated with total coloboma with microphthalmos.

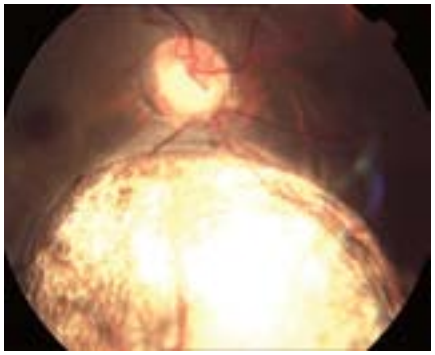


Fig. 12.4.1A: Retinochoroidal coloboma

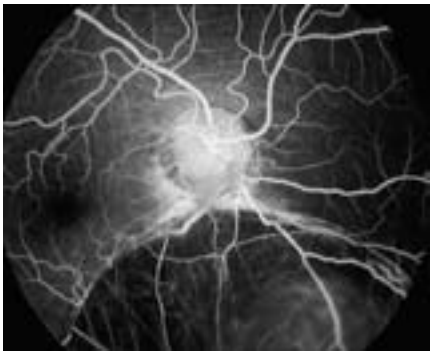


Fig. 12.4.1B: Retinochoroidal coloboma

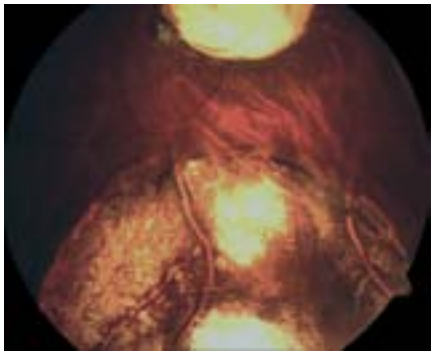


Fig. 12.4.2: Retinochoroidal isolated bridge coloboma

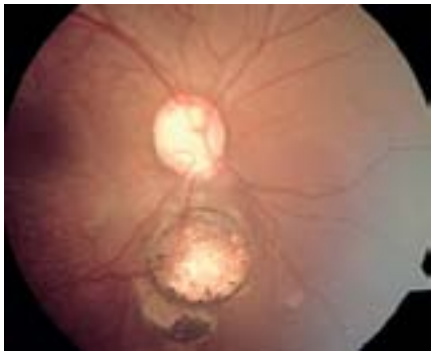


Fig. 12.4.3: Retinochoroidal isolated coloboma—double-disk appearance

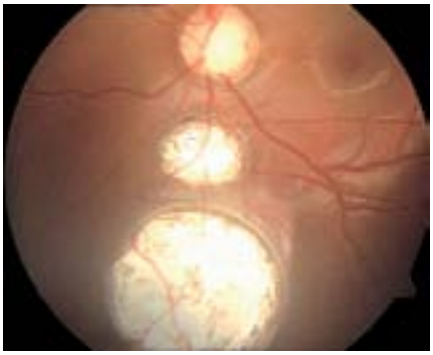


Fig. 12.4.4: Retinochoroidal isolated bridge colobomas—triple-disk appearance

Retinal Vascular Disorders

Central Retinal Artery Occlusion (CRAO)

- Painless, sudden loss of vision
- Occlusion may affect the central retinal artery occlusion (CRAO) itself
- A peripheral branch (arteriole), when the effect is localized
- Retina loses its transparency, and becoming opaque and milky-white, especially around the posterior pole **(Figs 12.5.1A and B)**

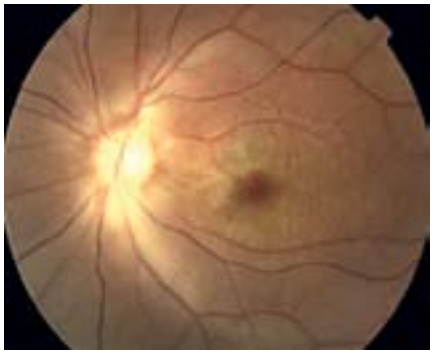


Fig. 12.5.1A: Central retinal artery occlusion

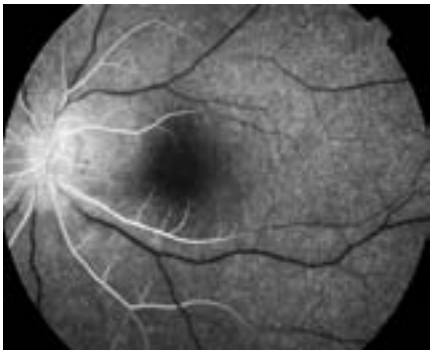


Fig. 12.5.1B: Central retinal artery occlusion—on FFA

- A cherry-red spot appears at the fovea with surrounding edema **(Figs 12.5.2A and B)**
- 'Cattle-track' appearance of the retinal arterioles in incomplete obstruction **(Figs 12.5.3A and B)**
- Branch occlusion occurs at a bifurcation and usually by an embolus **(Figs 12.5.4A and B)**
- Partial occlusion may be limited to the cilioretinal artery only in some cases **(Figs 12.5.5A and B)**

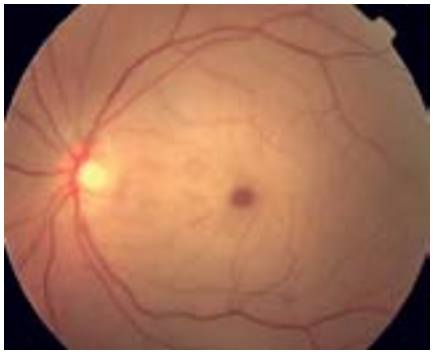


Fig. 12.5.2A: Central retinal artery occlusion—cherry-red spot

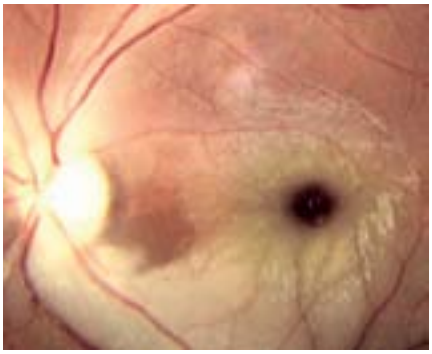


Fig. 12.5.2B: Cherry-red spot

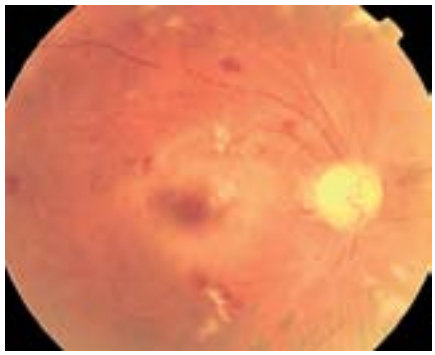


Fig. 12.5.3A: Incomplete CRAO—cattle tract appearance

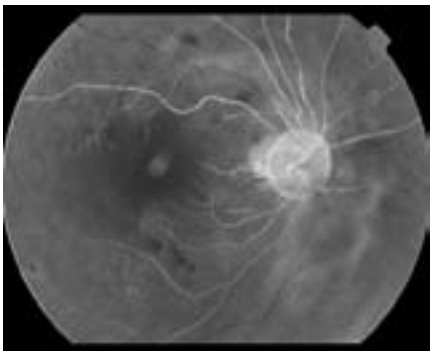


Fig. 12.5.3B: Incomplete CRAO—cattle tract appearance on FFA

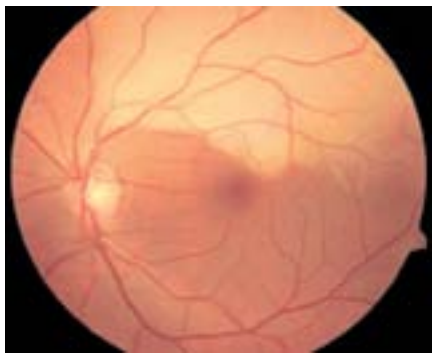


Fig. 12.5.4A: Branch retinal artery occlusion—upper temporal

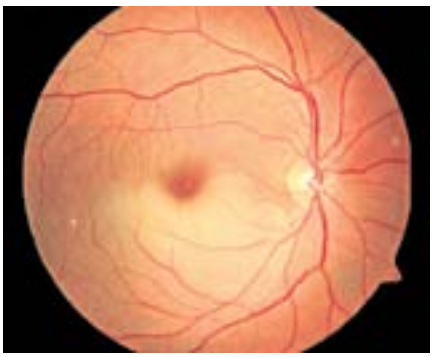


Fig. 12.5.4B: Branch retinal artery occlusion—lower temporal

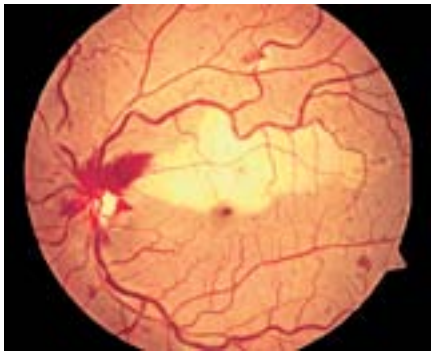


Fig. 12.5.5A: Cilioretinal artery occlusion

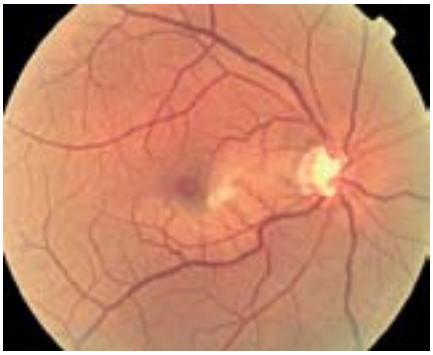


Fig. 12.5.5B: Cilioretinal artery occlusion

- An atheromatous embolus may be visible as a refractile body (Hollenhorst plaques) within the artery **(Figs 12.5.6A to C)**
- In presence cilioretinal artery (25% cases), macula may be often spared **(Figs 12.5.7A and B)**
- After a week or so, retina appears normal with optic atrophy with pale disk **(Figs 12.5.8A and B)**
- *Treatment:* Emergency vigorous digital massage, paracentesis, IV acetazolamide, CO_2 inhalation, etc.
- Prognosis is invariable poor, if the obstruction is more than 6 hours.

Retinal Vein Occlusion

- Obstruction of the central retinal vein, with sudden loss of vision
- Associated with systemic hypertension, diabetes, raised IOP, hypercellularity/viscosity of blood, or central periphlebitis
- *Two types: Nonischemic CRVO and ischemic CRVO*

Nonischemic CRVO

- Mild tortuosity and dilatation of all branches of the central retinal vein **(Figs 12.6.1A and B)**

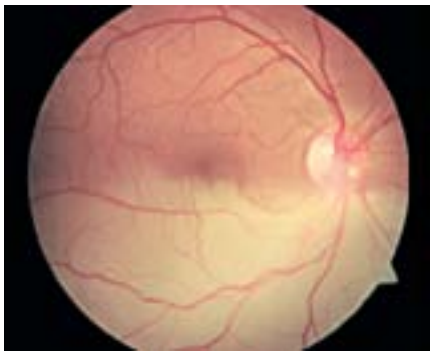


Fig. 12.5.6A: Hemiretinal artery occlusion—Hollenhorst plaques at the lower bifurcation

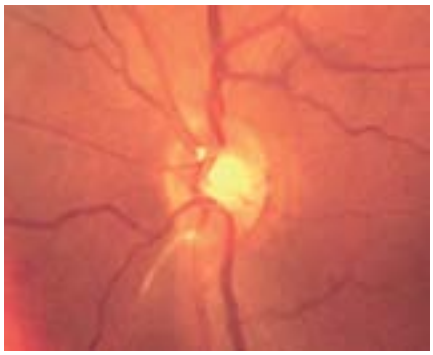


Fig. 12.5.6B: Hollenhorst plaques at the upper and lower bifurcations

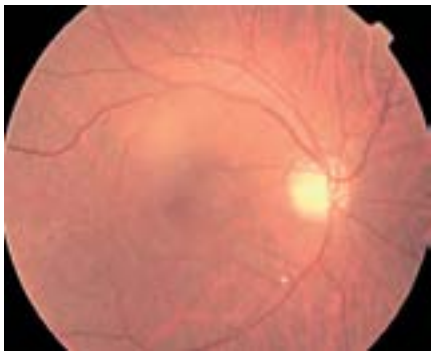


Fig. 12.5.6C: Hollenhorst plaques at the branch arteriole

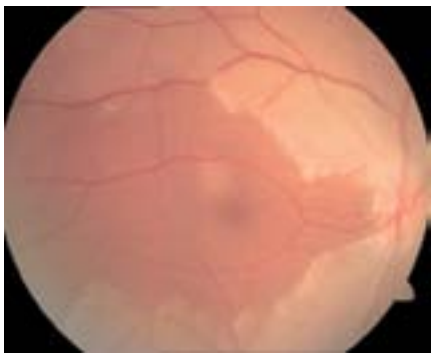


Fig. 12.5.7A: CRAO—patent cilioretinal artery

Fig. 12.5.7B: CRAO—patent cilioretinal

Fig. 12.5.8A: CRAO—optic disk pallor

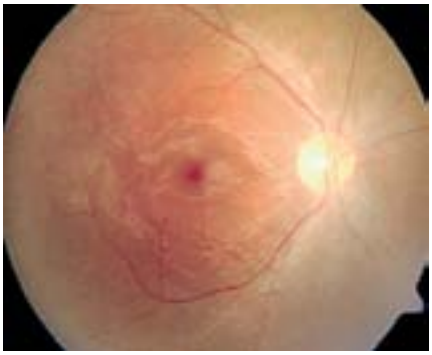


Fig. 12.5.8B: CRAO—optic atrophy

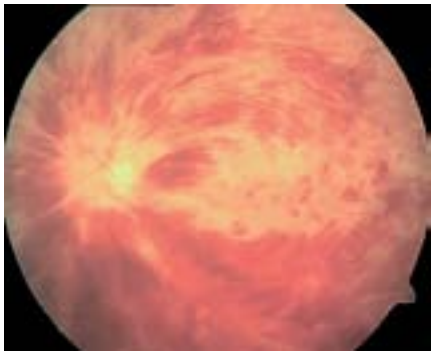


Fig. 12.6.1A: Nonischemic CRVO

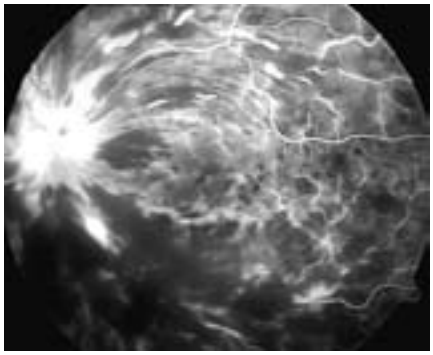


Fig. 12.6.1B: Nonischemic CRVO

- 'Dot' and 'blot'; and 'flame' shaped hemorrhages are seen throughout all four quadrants of the retina **(Figs 12.6.2A and B)**
- Sometimes, called 'Blood and Thunder' fundus **(Fig. 12.6.3)** and it takes long time to resolve completely **(Fig. 12.6.4)**
- Sclerosed veins may be seen in old CRVO **(Fig. 12.6.5)**
- *Treatment:* Treatment of the predisposing factors.

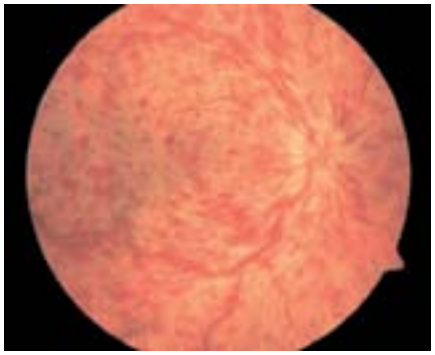


Fig. 12.6.2A: Nonischemic CRVO

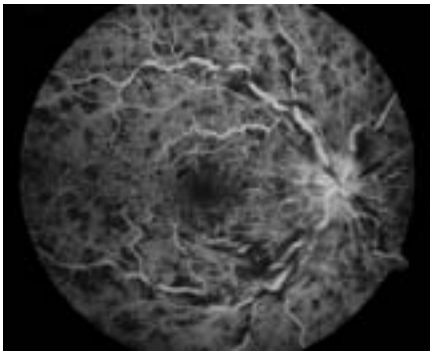


Fig. 12.6.2B: Nonischemic CRVO

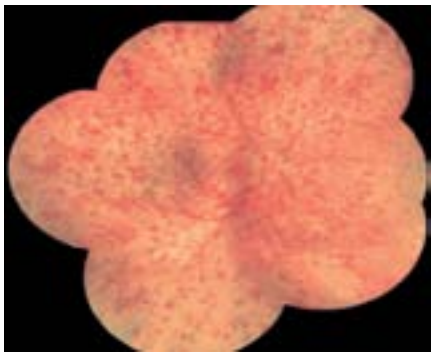


Fig. 12.6.3: CRVO—blood and thunder fundus

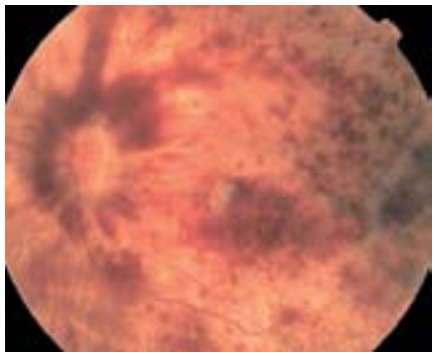


Fig. 12.6.4: Resolving nonischemic CRVO

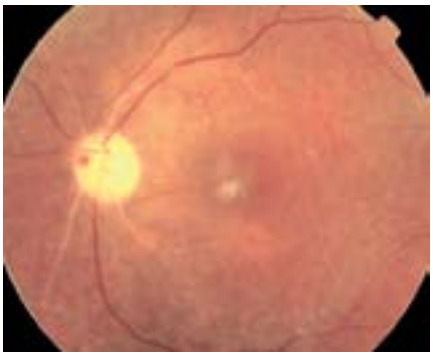


Fig. 12.6.5: Old CRVO—sclerosed veins

Ischemic CRVO

- Marked tortuosity and engorgement of the retinal veins
- Massive superficial flame-shaped and deep blotchy hemorrhages throughout the fundus **(Figs 12.7.1A and B)**
- *Cotton-wool exudates* are most prominent in ischemic CRVO **(Figs 12.7.2A and B)**

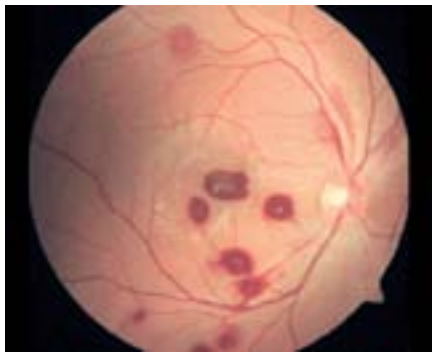


Fig. 12.7.1A: Ischemic CRVO—RE

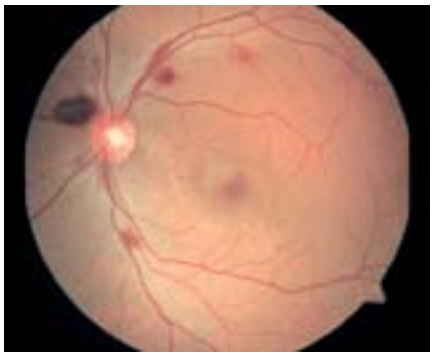


Fig. 12.7.1B: Ischemic CRVO—LE

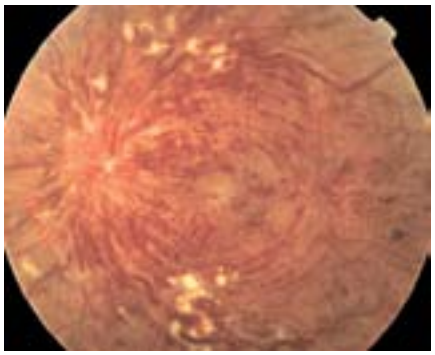


Fig. 12.7.2A: Ischemic CRVO

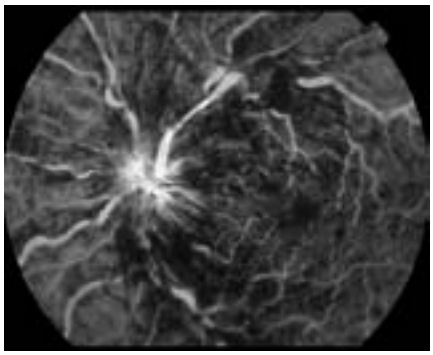


Fig. 12.7.2B: Ischemic CRVO

- May be associated with preretinal hemorrhage **(Figs 12.7.3A and B)**
- Optic disk is swollen and hyperemic **(Figs 12.7.4A and B)**
- Macular edema and hemorrhages
- Very rarely, a CRVO may present simultaneously with CRAO **(Fig. 12.7.5)**
- *Treatment:* Patient should be followed up closely to prevent rubeosis iridis and neovascular glaucoma.

Venous stasis retinopathy

- Milder form of nonischemic CRVO
- Venous engorgement and tortuosity is a prominent feature **(Fig. 12.7.6)**
- Associated with multiple small 'dot and blot' preretinal and flame-shaped hemorrhage
- It is seen in hyperviscosity syndrome, like, polycythemia vera, acute hypoxia in high altitude **(Figs 12.7.7A and B)**, multiple myeloma, macroglobulinemia, etc.

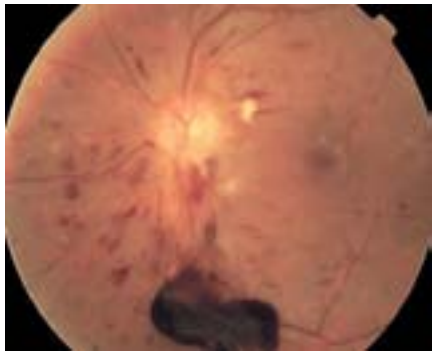


Fig. 12.7.3A: Ischemic CRVO—preretinal hemorrhage

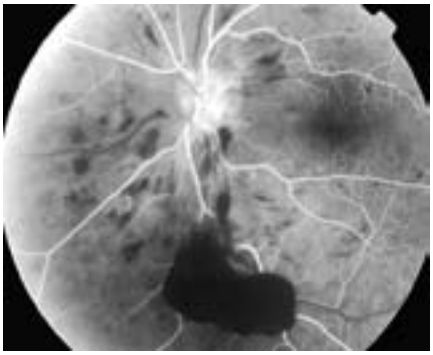


Fig. 12.7.3B: Ischemic CRVO—preretinal hemorrhage

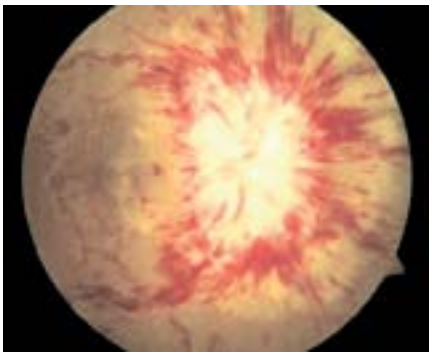


Fig. 12.7.4A: CRVO—disk edema—RE

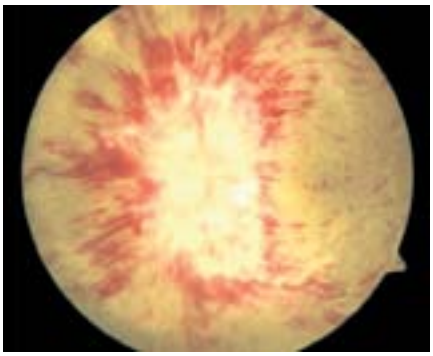


Fig. 12.7.4B: CRVO—disk edema—LE

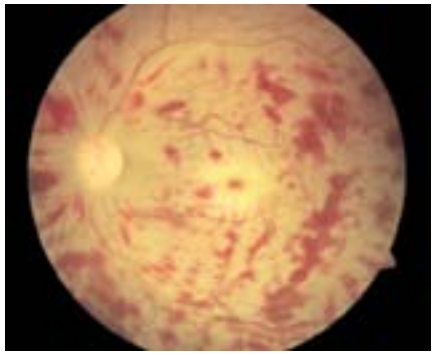


Fig. 12.7.5: CRVO and CRAO—simultaneous presentation

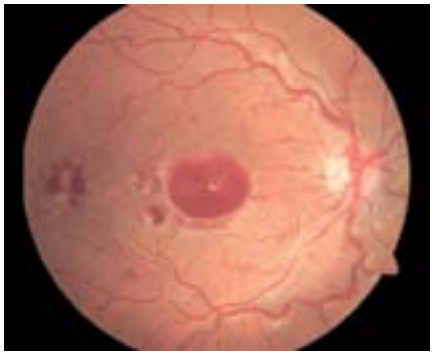


Fig. 12.7.6: Venous stasis retinopathy—RE

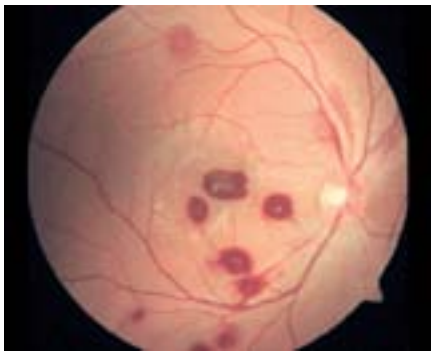


Fig. 12.7.7A: Venous stasis retinopathy—RE

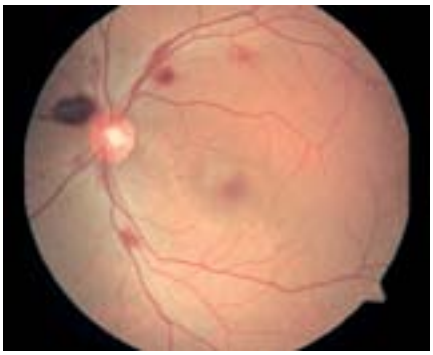


Fig. 12.7.7B: Venous stasis retinopathy—LE

Branch Retinal Venous Occlusion (BRVO)

- BRVO may occur near the optic disk and involves a major quadrant of the retina **(Figs 12.8.1A and B)**
- It also occurs at a peripheral crossing with an artery **(Figs 12.8.2A and B)**
- *Ophthalmoscopically,* the affected part of the retina, drained by the obstructed vein shows:
  - Dilated and tortuous veins
  - Flame-shaped, and 'dot' and 'blot' hemorrhage in the affected quadrant
  - Edema and cotton-wool spots
- Blockage of the superotemporal (ST) vein frequently involves the macula **(Figs 12.8.3A and B)** but sometimes, macula is spared **(Fig. 12.8.4)**
- Even with inferotemporal (IT) vein, macula may also involved **(Fig. 12.8.5)** which may be bilateral in rare situation **(Figs 12.8.6A and B)**

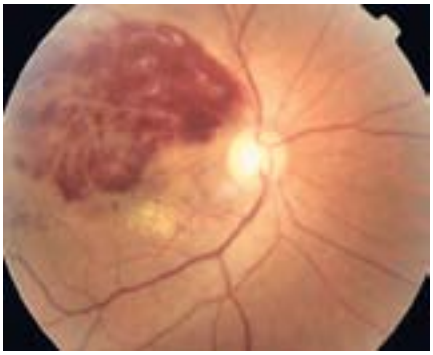


Fig. 12.8.1A: BRVO—at the disk margin

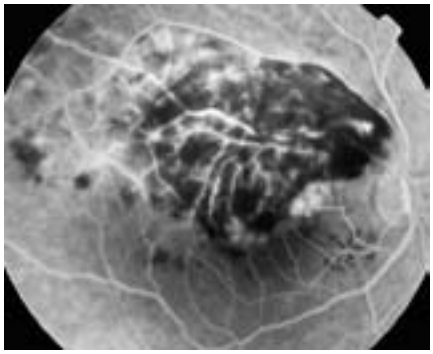


Fig. 12.8.1B: BRVO—at the disk margin

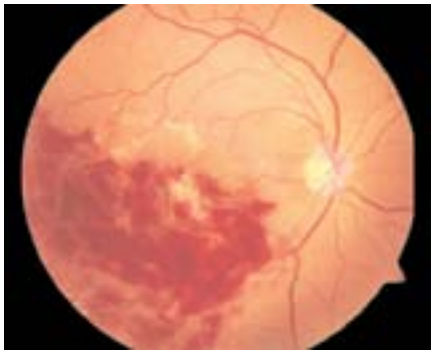


Fig. 12.8.2A: BRVO—at the AV crossing

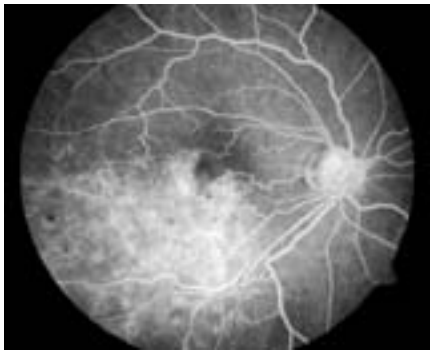


Fig. 12.8.2B: BRVO—at the AV crossing

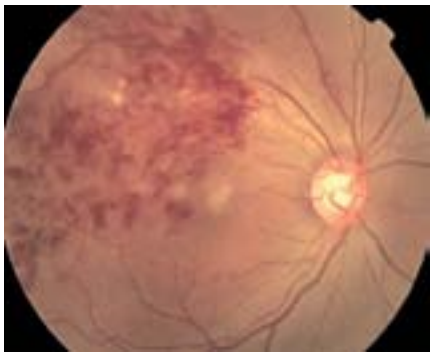


Fig. 12.8.3A: BRVO—superotemporal with macular involvement

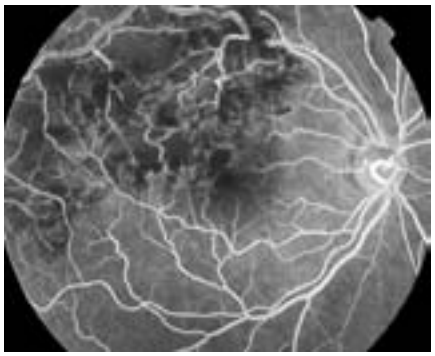


Fig. 12.8.3B: BRVO—superotemporal with macular involvement

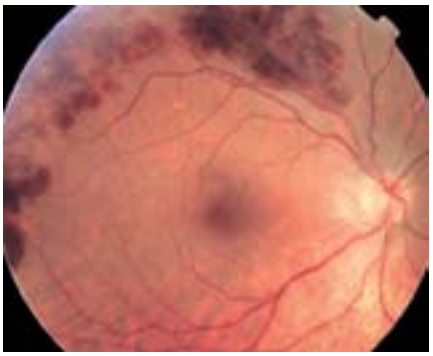


Fig. 12.8.4: BRVO—superotemporal with macular sparing

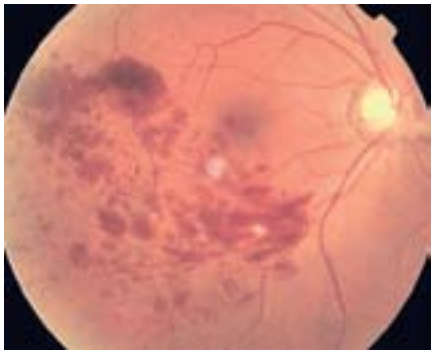


Fig. 12.8.5: Inferotemporal (IT) BRVO—macular involvement

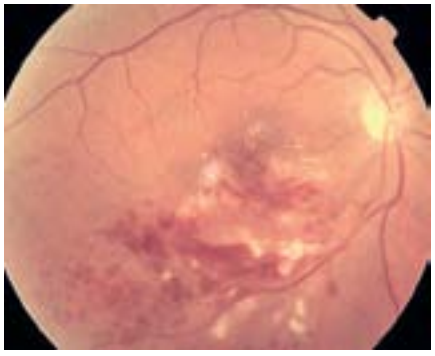


Fig. 12.8.6A: Bilateral inferotemporal BRVO—RE

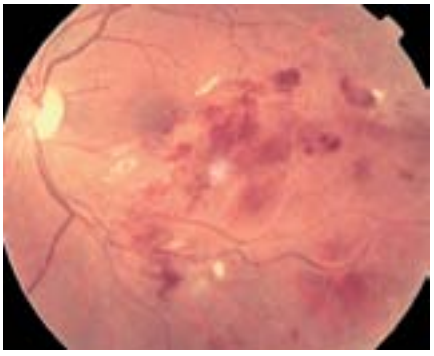


Fig. 12.8.6B: Bilateral inferotemporal BRVO—LE

- Sometimes there is *hemiretinal venous occlusion*, involving the upper half or lower half of the retina **(Figs 12.8.7A and B)**
- There may be *tributary venous occlusion* with or without macular involvement **(Figs 12.8.8 to 12.8.10)**
- In case of old BRVO their may be sclerosis of blood vessels **(Fig. 12.8.11)** with formation of collaterals **(Figs 12.8.12A and B)** and neovascularization

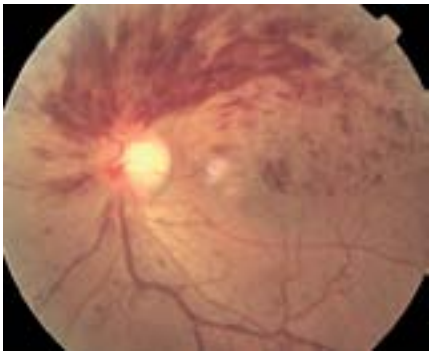


Fig. 12.8.7A: Hemiretinal venous occlusion

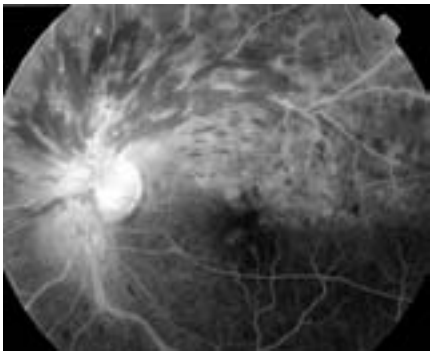


Fig. 12.8.7B: Hemiretinal venous occlusion

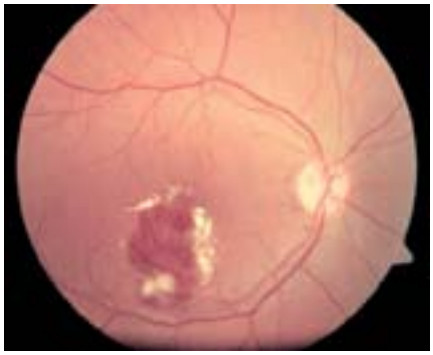


Fig. 12.8.8: BRVO—inferior tributary macular BRVO

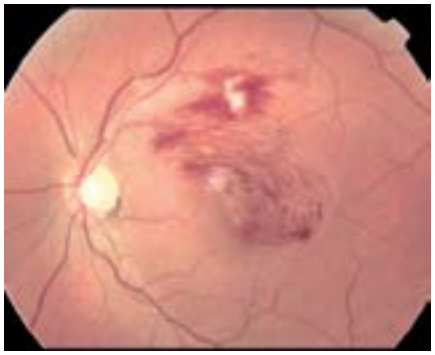


Fig. 12.8.9A: Superotemporal (ST) BRVO—tributary

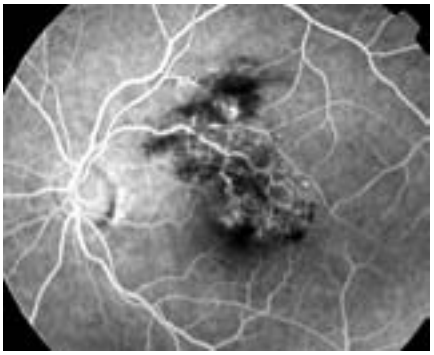


Fig. 12.8.9B: ST BRVO—tributary occlusion with macular involvement

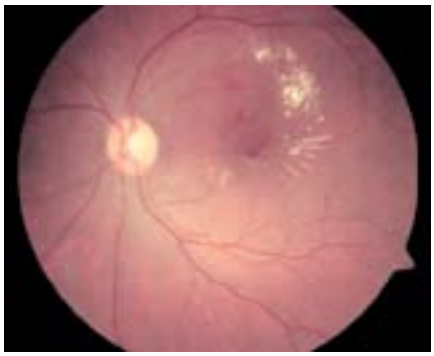


Fig. 12.8.10A: BRVO—tributary occlusion with macular edema

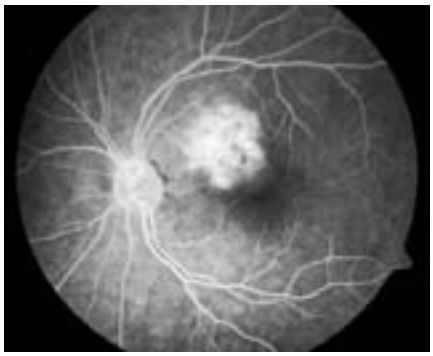


Fig. 12.8.10B: BRVO—tributary occlusion with macular edema

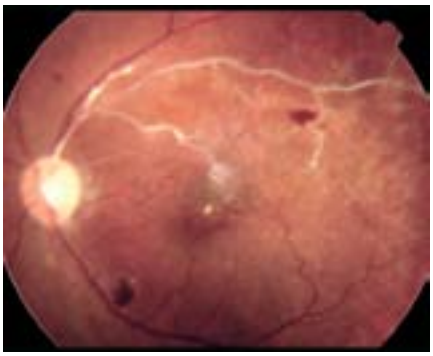


Fig. 12.8.11: ST BRVO—sclerosed vessels

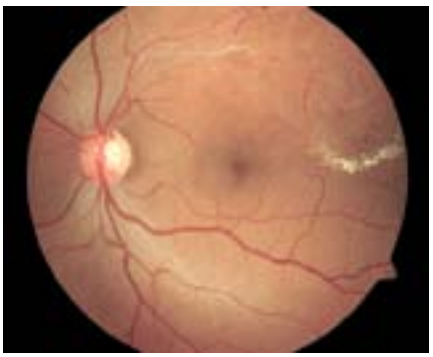


Fig. 12.8.12A: Old ST BRVO—sclerosed vessels

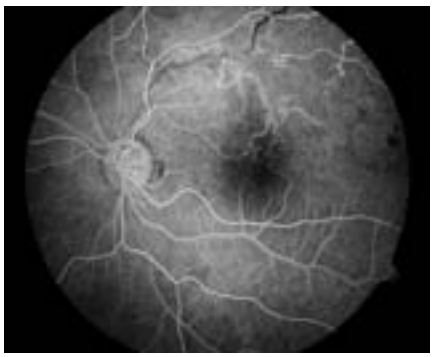


Fig. 12.8.12B: Old ST BRVO—sclerosed vessels with collaterals

(Figs 12.8.13A and B) and sometimes with epiretinal fibrovascular membrane formation **(Figs 12.8.14 and 12.8.15)**

- *Treatment:* Fundus fluorescein angiography (FFA) to know the macular perfusion status and later on macular grid photocoagulation to treat neovascularization and anti-VGEF injection to treat CME.

Preretinal (Subhyaloid) Hemorrhage

- Usually solitary and located at the posterior pole obscuring the visualization of underlying retinal vessels **(Figs 12.9.1A and B)**
- Initially round with or without a fluid level **(Figs 12.9.2 to 12.9.4)**, but later turn into *boat-shaped hemorrhage* due to settling by the gravity **(Figs 12.9.5A and B)**

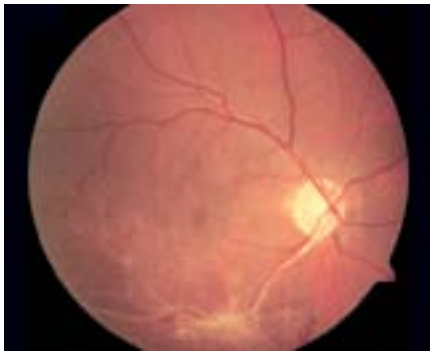


Fig. 12.8.13A: Old BRVO—with NVE

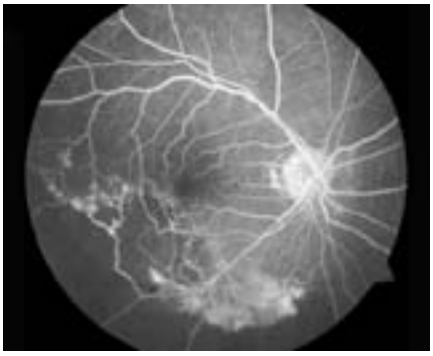


Fig. 12.8.13B: Old BRVO—with NVE with capillary drop out area

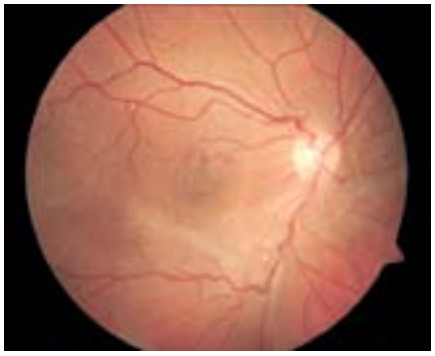


Fig. 12.8.14: Old BRVO—with ERM

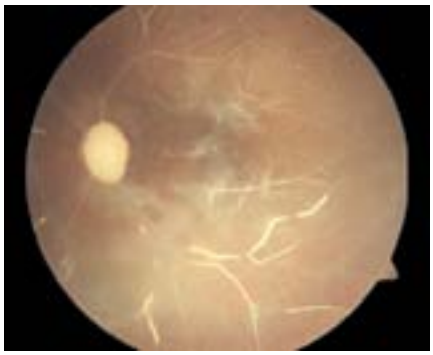


Fig. 12.8.15: Old BRVO—sclerosed vessels with glaucomatous cupping and ERM

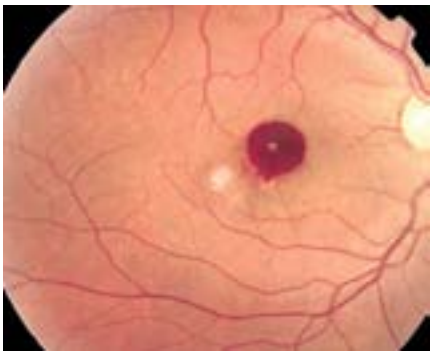


Fig. 12.9.1A: Preretinal (subhyaloid) hemorrhage

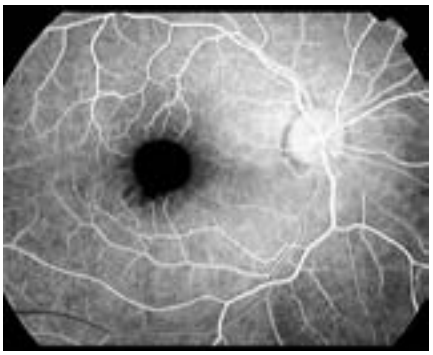


Fig. 12.9.1B: Preretinal (subhyaloid) hemorrhage

Fig. 12.9.2: Preretinal (subhyaloid) hemorrhage—small macular and with fluid level

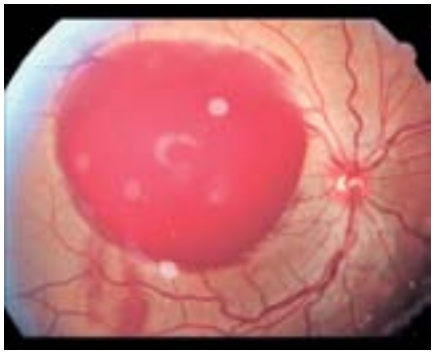


Fig. 12.9.3: Preretinal (subhyaloid) hemorrhage

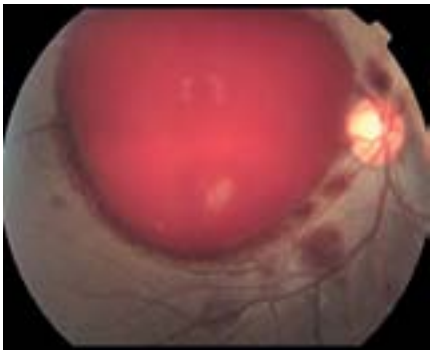


Fig. 12.9.4: Preretinal (subhyaloid) hemorrhage—large

- Large hemorrhage may break into the vitreous cavity spontaneously
- Absorption occurs from the top with yellow-white discoloration **(Figs 12.9.6 and 12.9.7)**. With time all blood gets absorbed leaving behind impression corresponds to the affected area **(Fig. 12.9.8)**
- Fundus fluorescein angiogram (FFA) reveals blocking of retinal fluorescence **(*see* Figs 12.9.1B, 12.9.7B and 12.9.9A and B)**
- *Causes:* Trauma, Valsalva retinopathy, proliferative retinopathies, Terson's syndrome (with subarachnoid hemorrhage)
- *Treatment:* Nd:YAG laser hyaloidotomy to drain the blood into the vitreous cavity **(Figs 12.9.10 and 12.9.11)**. This drained blood in the vitreous cavity gets absorbed quickly.

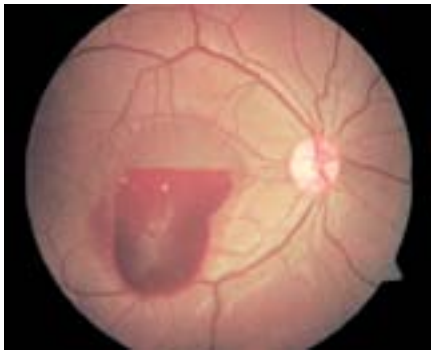


Fig. 12.9.5A: Preretinal (subhyaloid) hemorrhage with fluid level

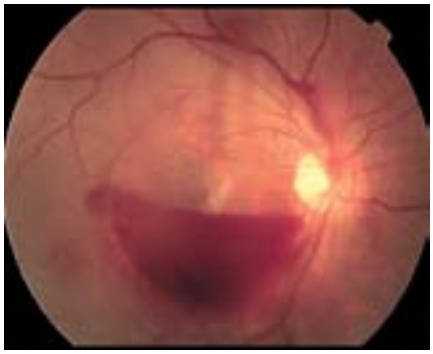


Fig. 12.9.5B: Preretinal (subhyaloid) hemorrhage—boat-shaped

Fig. 12.9.6: Preretinal (subhyaloid) hemorrhage

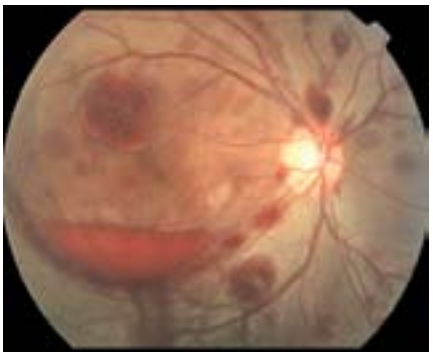


Fig. 12.9.7A: Subhyaloid hemorrhage

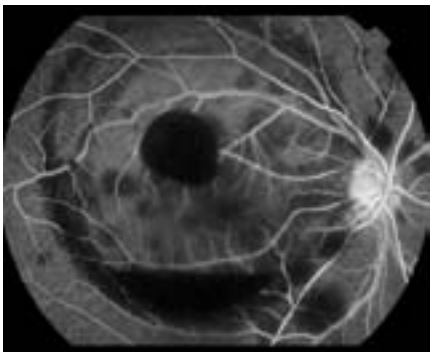


Fig. 12.9.7B: Subhyaloid hemorrhage

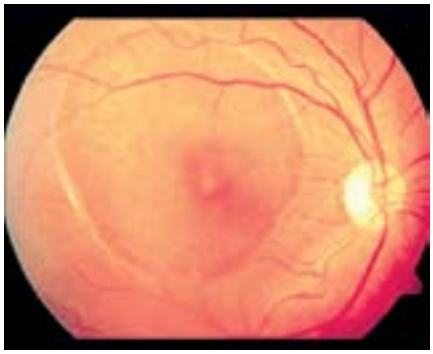


Fig. 12.9.8: Preretinal (subhyaloid) hemorrhage—absorbed

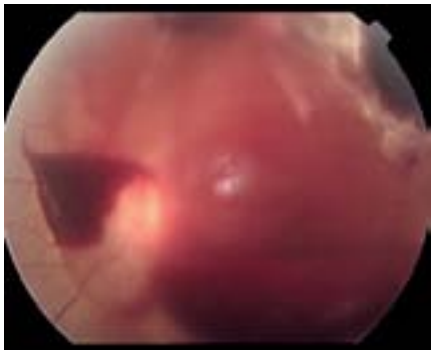


Fig. 12.9.9A: Preretinal hemorrhage—large

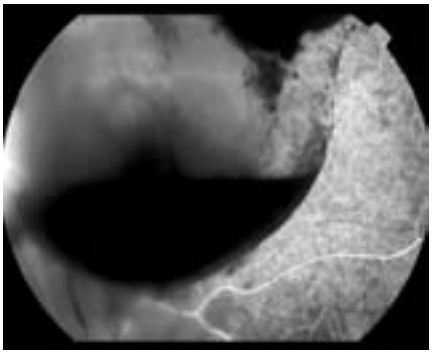


Fig. 12.9.9B: Preretinal hemorrhage—large on FFA

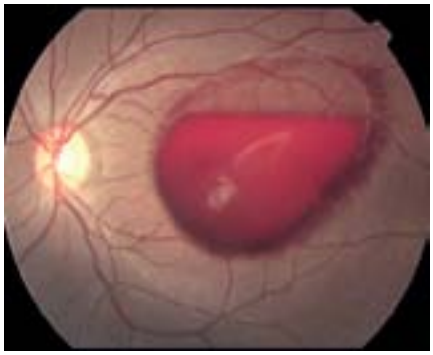


Fig. 12.9.10A: Preretinal (subhyaloid) hemorrhage

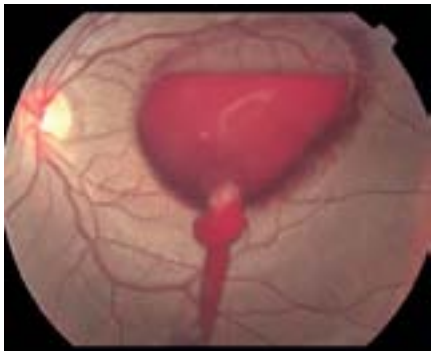


Fig. 12.9.10B: Preretinal (subhyaloid) hemorrhage—YAG hyaloidotomy

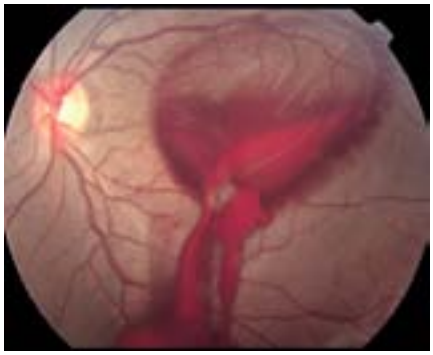


Fig. 12.9.10C: Preretinal (subhyaloid) hemorrhage—YAG hyaloidotomy

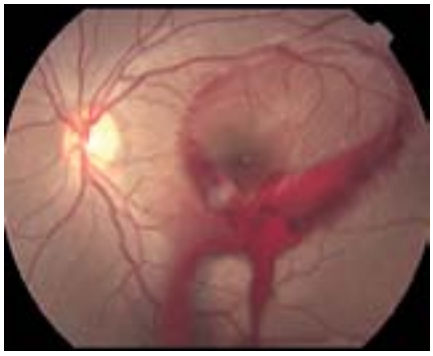


Fig. 12.9.10D: Preretinal (subhyaloid) hemorrhage—YAG hyaloidotomy

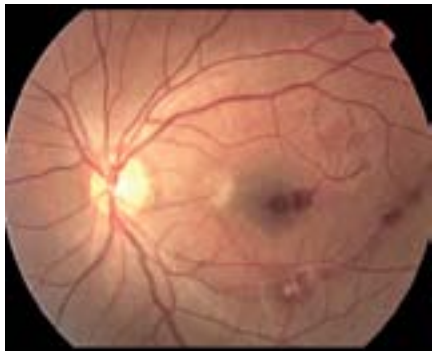


Fig. 12.9.10E: Preretinal (subhyaloid) hemorrhage—YAG hyaloidotomy

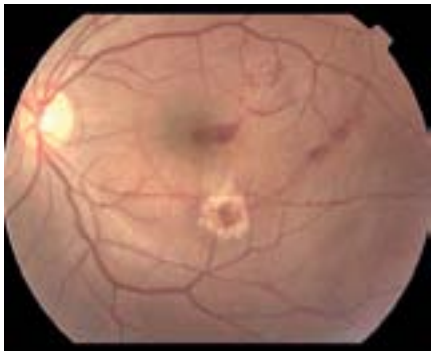


Fig. 12.9.10F: Preretinal (subhyaloid) hemorrhage—focal laser done after complete drainage of blood

Roth's Spot

- Retinal hemorrhage with white center **(Figs 12.10.1 and 12.10.2)**
- *Causes:* Leukemia, dysproteinemias, severe anemia, dysproteinemias, subacute bacterial endocarditis, HIV retinopathy
- A complete hemogram is necessary.

Fig. 12.9.11: Subhyaloid hemorrhage—after YAG laser hyaloidotomy

Fig. 12.10.1A: Roth's spot—severe anemia

Fig. 12.10.1B: Roth's spot—severe anemia

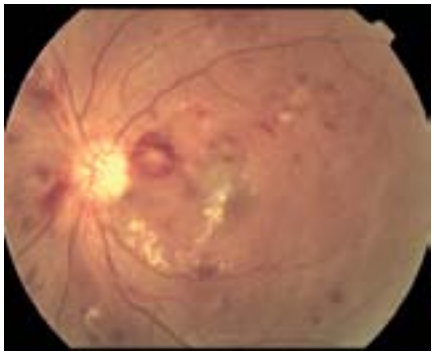


Fig. 12.10.2: Roth's spot with retinal hemorrhage—leukemia

Subretinal Hemorrhage

- Blood is localized between the photoreceptors layer and retinal pigment epithelium (RPE) layer
- Large, bright red area with indistinct outline **(Figs 12.11.1 and 12.11.2)**
- May be associated with sub-RPE hemorrhage in some cases and may cause large subretinal hematoma **(Figs 12.11.3A and B)**
- Overlying retina is slightly elevated with visible retinal blood vessels
- *Causes:* Blunt trauma, choroidal neovascularization, ruptured retinal macroaneurysm, etc.

Sub-RPE Hemorrhage

- Blood from choroid seeps into the space between the Bruch's membrane and RPE
- Solitary, usually very dark-red with well defined outline **(Figs 12.12.1 and 12.12.2)**

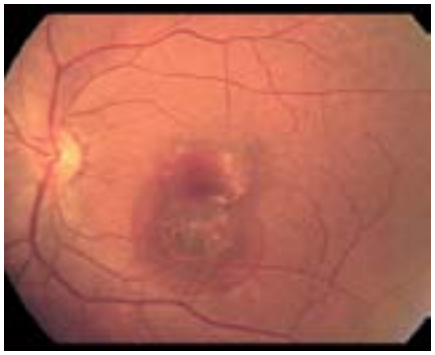


Fig. 12.11.1A: Subretinal hemorrhage

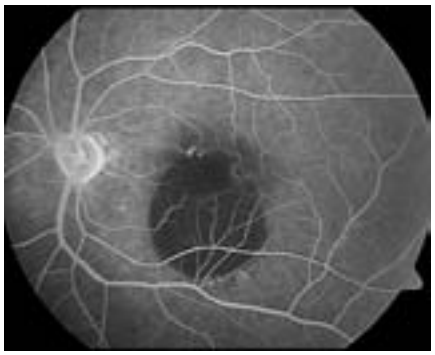


Fig. 12.11.1B: Subretinal hemorrhage on FFA

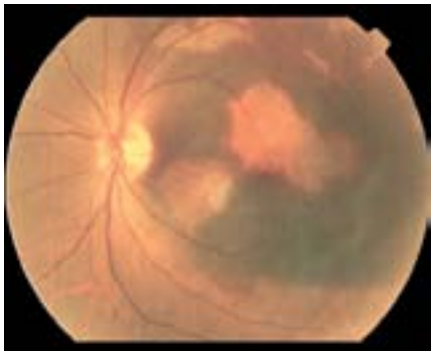


Fig. 12.11.2A: Subretinal hemorrhage—post-traumatic

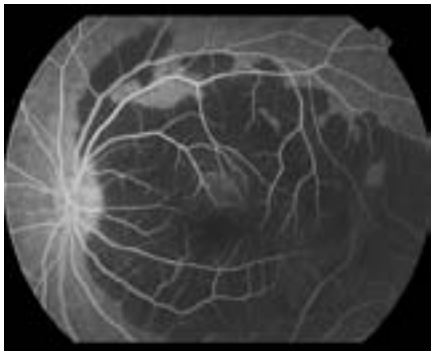


Fig. 12.11.2B: Subretinal hemorrhage—post-traumatic

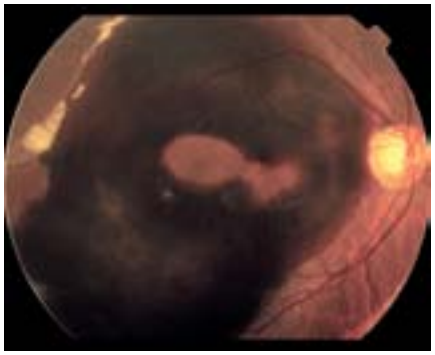


Fig. 12.11.3A: Subretinal hematoma following CNVMs

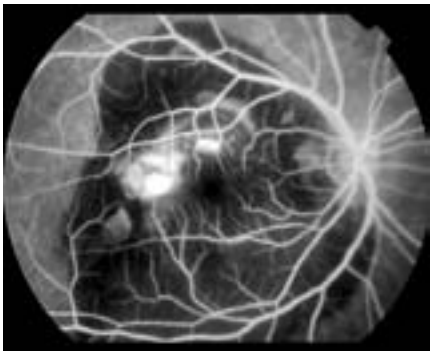


Fig. 12.11.3B: Subretinal hematoma following CNVMs

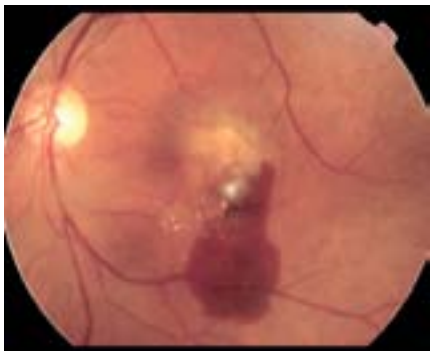


Fig. 12.12.1A: Sub-RPE hemorrhage

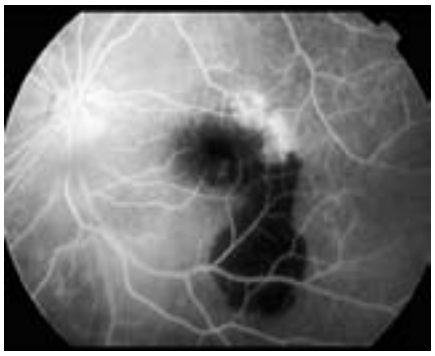


Fig. 12.12.1B: Sub-RPE hemorrhage

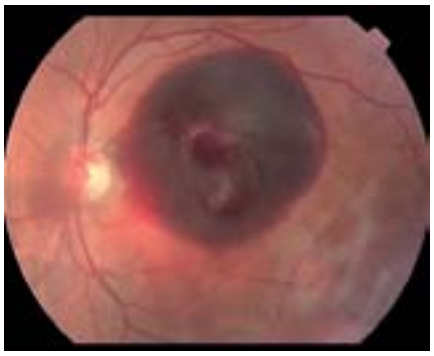


Fig. 12.12.2: Sub-RPE hemorrhage

- May be multiple in some cases **(Figs 12.12.3A and B)**
- FFA shows blocked background choroidal fluorescence
- *Most common cause:* Subretinal choroidal neovascular membranes (SRNVMs).

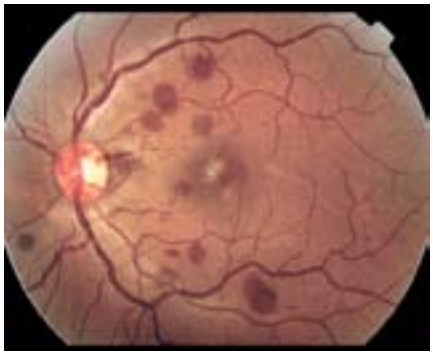


Fig. 12.12.3A: Sub-RPE hemorrhage—multiple

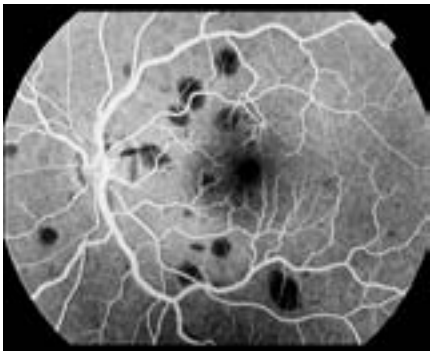


Fig. 12.12.3B: Sub-RPE hemorrhage—multiple on FFA

Choroidal Hemorrhage

- Dark-red, almost black, with well defined outline **(Fig. 12.13.1)** may be associated with exudative RD **(Fig. 12.13.2)**
- *Causes:* Blunt trauma, often associated with choroidal tear; during drainage of subretinal fluid
- In case of severe trauma, there may be combination of sub-RPE and choroidal hemorrhage **(Fig. 12.13.3)**.

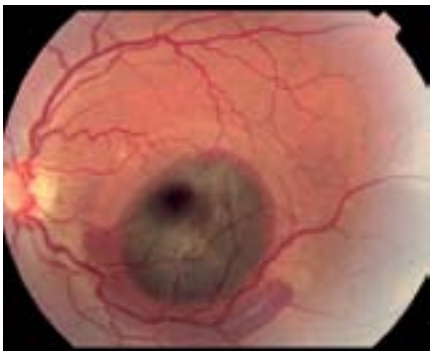


Fig. 12.13.1: Choroidal hemorrhage

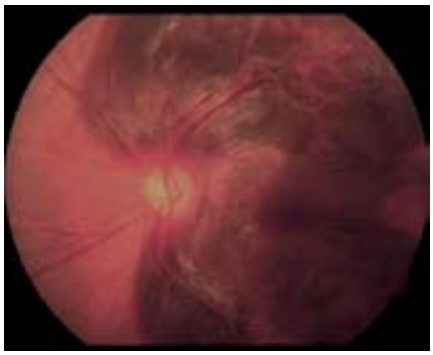


Fig. 12.13.2: Choroidal hemorrhage—with exudative RD

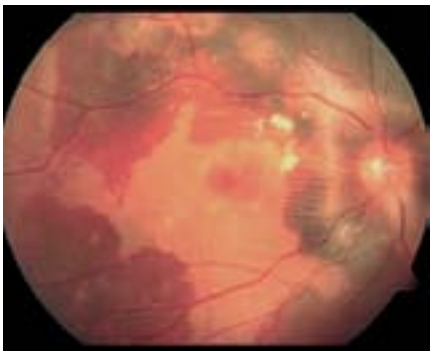


Fig. 12.13.3: Extensive subretinal, sub-RPE and choroidal hemorrhage in trauma

Neovascularization

- May involve the optic nerve head, called neovascularization of the disk (NVD) **(Figs 12.14.1 and 12.14.2)** and when it involves central or peripheral retina, it is called neovascularization elsewhere (NVE) **(Fig. 12.14.3)**

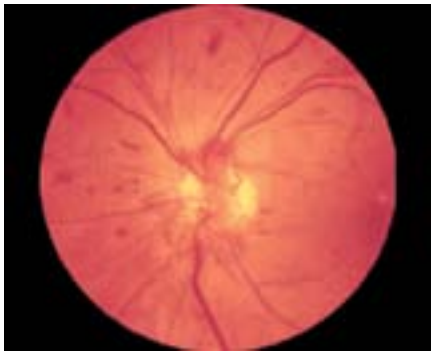


Fig. 12.14.1: Neovascularization of the disk (NVD)

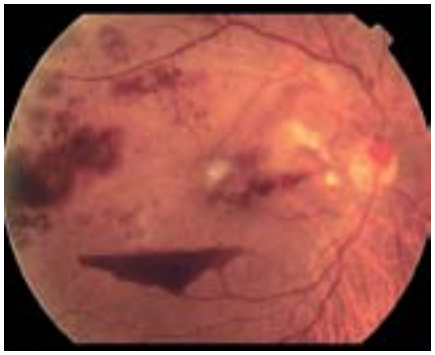


Fig. 12.14.2A: NVD with preretinal hemorrhage

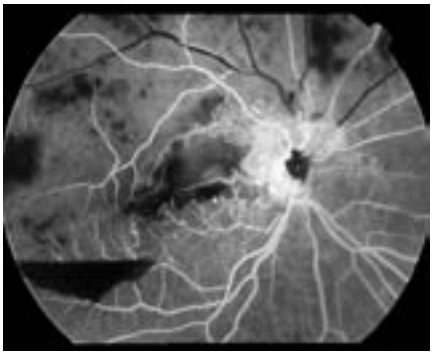


Fig. 12.14.2B: NVD with preretinal hemorrhage

- Both NVD and NVE exist together especially in case of proliferative diabetic retinopathy **(Figs 12.14.4 and 12.14.5)**
- May involve peripheral retina and takes the configuration of "sea fan" which can be demonstrated beautifully on FFA (**Figs 12.14.6A to C)**

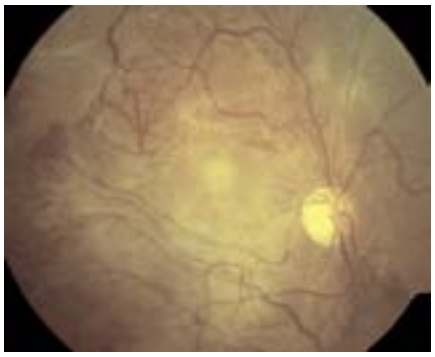


Fig. 12.14.3: Extensive neovascularization elsewhere

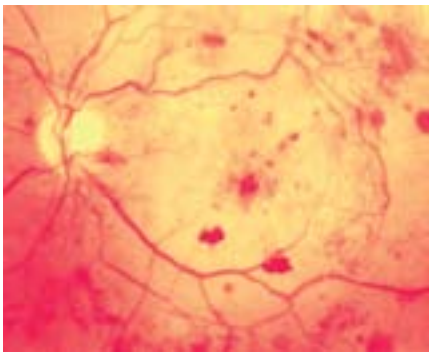


Fig. 12.14.4: Both NVD and NVE in PDR

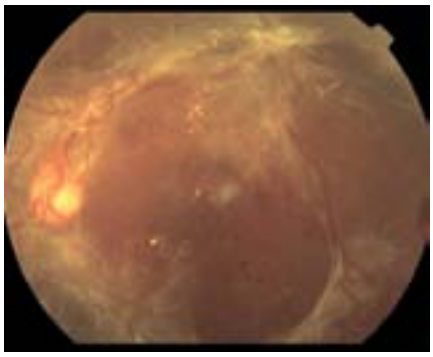


Fig. 12.14.5A: Both NVD and NVE in extensive PDR

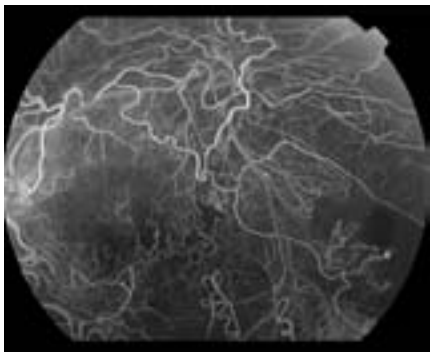


Fig. 12.14.5B: Both NVD and NVE in extensive PDR on FFA

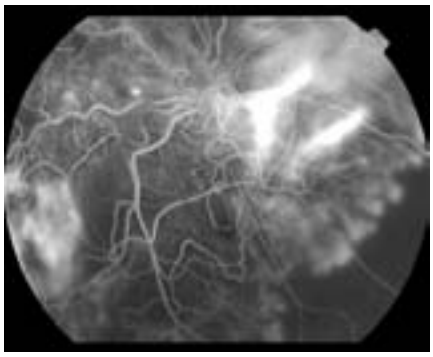


Fig. 12.14.5C: Both NVD and NVE in extensive PDR on FFA late phase

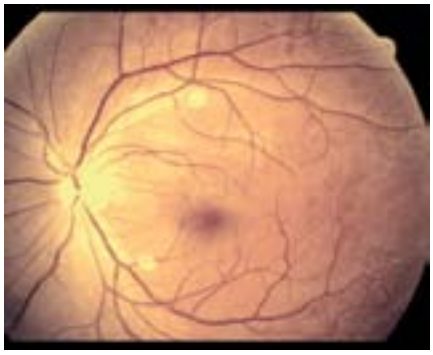


Fig. 12.14.6A: NVE in peripheral retina—sea fan neovascularization

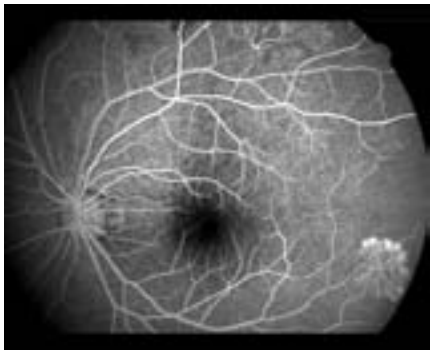


Fig. 12.14.6B: NVE in peripheral retina—sea fan neovascularization on FFA

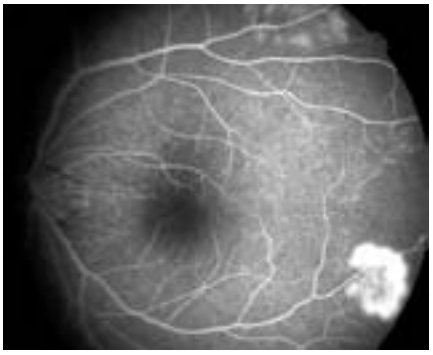


Fig. 12.14.6C: NVE in peripheral retina—sea fan neovascularization on FFA late phase

- New vessels often bleed, resulting in intraretinal or vitreous hemorrhage **(Figs 12.14.7 and 12.14.8)**
- New vessels may be flat, elevated or mixed and associated with variable degree of fibrovascular proliferation **(Fig. 12.14.9)**
- When this fibrovascular tissue contracts, it may result in tractional retinal detachment **(Figs 12.14.10A and B)**
- *Causes:* PDR, old CRVO or BRVO, retinal vasculitis, Eales' disease, ROP, sickle cell retinopathy, etc.

Fig. 12.14.7: NVD with vitreous hemorrhage

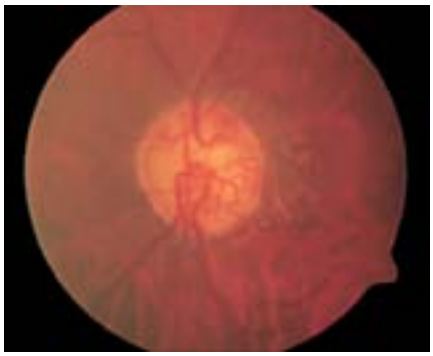


Fig. 12.14.8: NVD with vitreous hemorrhage

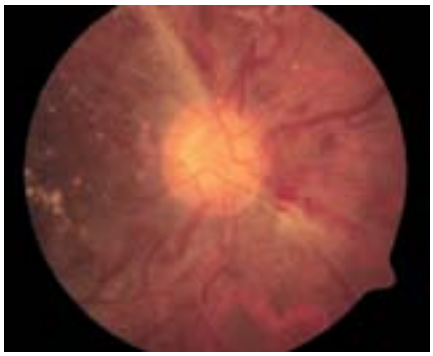


Fig. 12.14.9: NVD with fibrovascular proliferation

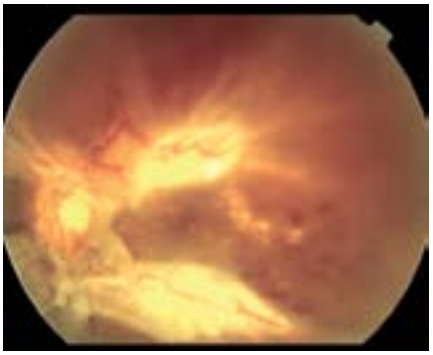


Fig. 12.14.10A: Neovascularization with fibrovascular proliferation—tractional RD

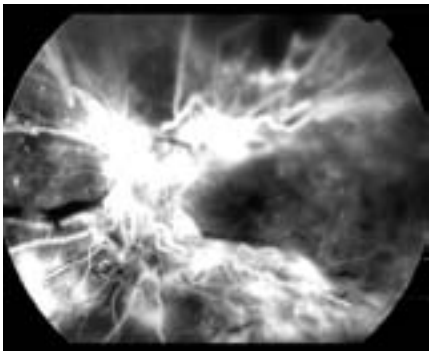


Fig. 12.14.10B: Neovascularization with fibrovascular proliferation—tractional RD

Collaterals

- Are the pre-existing capillaries which occur in response to any retinal vascular insult **(Figs 12.15.1A and B)**
- These shunts are acquired communication in between:
  - *Artery and vein* as in diabetic retinopathy

Fig. 12.15.1A: Retinal collaterals

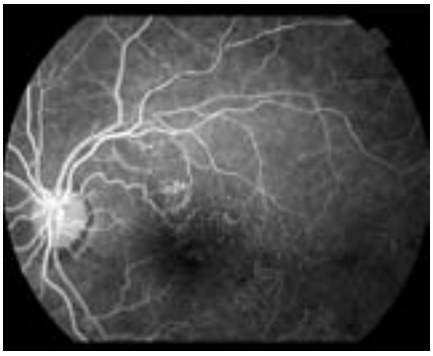


Fig. 12.15.1B: Retinal collaterals

  - *Artery and artery* as in BRVO
  - *Vein and vein* as in BRVO **(Figs 12.15.2 and 12.15.3)** or in CRVO
- Larger in caliber than that of neovascularization and never crosses the major retinal vessels
- Can occur on the disk **(Figs 12.15.4 and 12.15.5)**
- Collaterals should not be lasered as they are more physiological
- Collaterals and neovascularization of the disk or elsewhere may exist together **(Figs 12.15.6A to C)**.

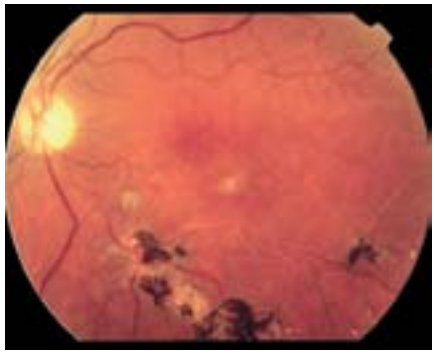


Fig. 12.15.2A: Retinal collaterals with NVE

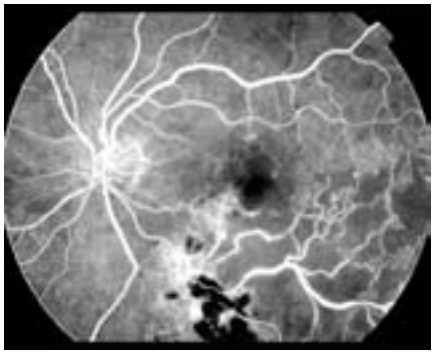


Fig. 12.15.2B: Retinal collaterals with NVE

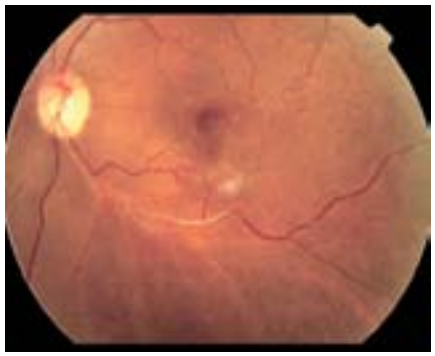


Fig. 12.15.3A: Retinal collaterals—BRVO

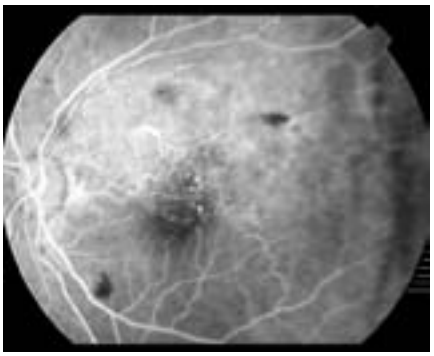


Fig. 12.15.3B: Retinal collaterals—BRVO

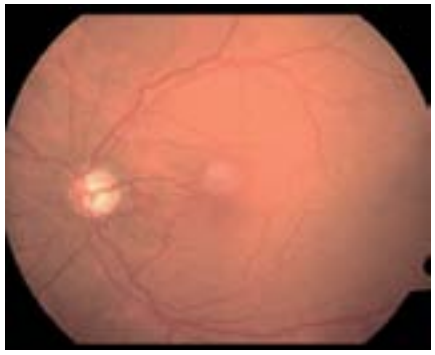


Fig. 12.15.4A: Collaterals—optic disk

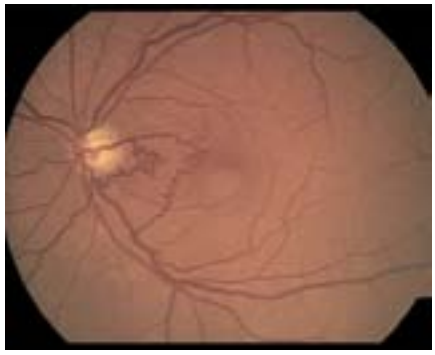


Fig. 12.15.4B: Collaterals—optic disk (in red free light)

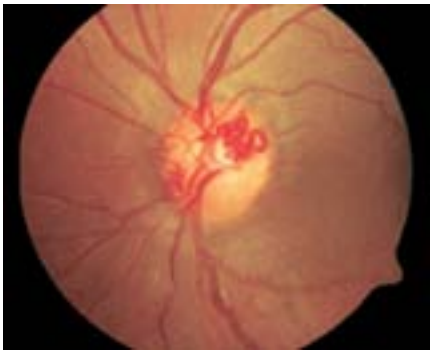


Fig. 12.15.5: Optic disk collaterals

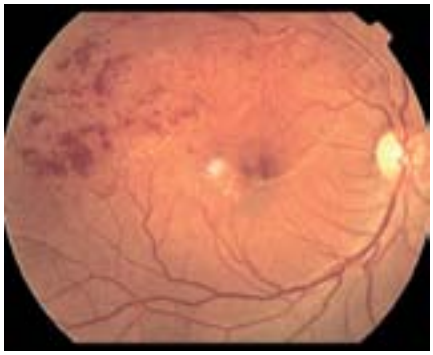


Fig. 12.15.6A: Collaterals and neovascularization

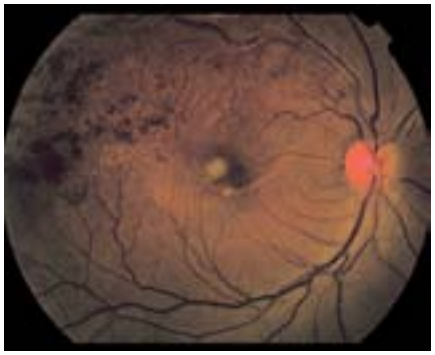


Fig. 12.15.6B: Collaterals and neovascularization (red free)

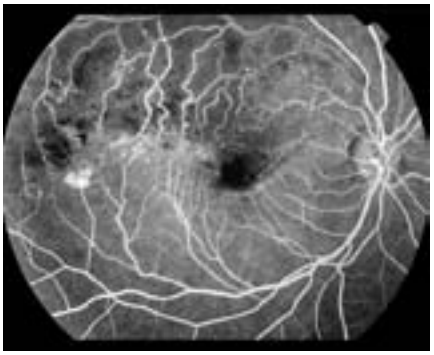


Fig. 12.15.6C: Collaterals and neovascularization

Retinal Hemangiomas

- Uncommon, often multiple, benign lesion
- May be associated with von Hippel-Lindau syndrome **(*see* Fig. 12.2.5)**
- Small to large orangish-red nodular lesion associated with dilated and tortuous blood vessels **(Fig. 12.16.1)**
- Often associated with profuse hard exudates **(Figs 12.16.2A and B)**.

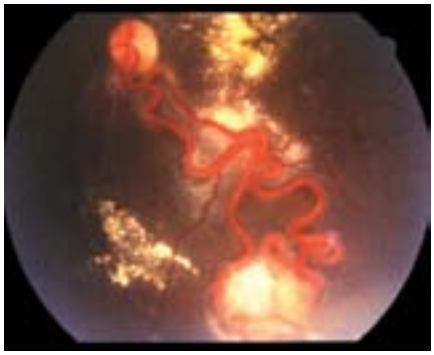


Fig. 12.16.1: Angiomatosis retinae

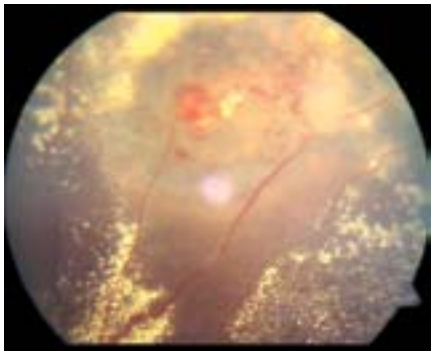


Fig. 12.16.2A: Retinal angioma—angiomatosis retinae with hard exudates

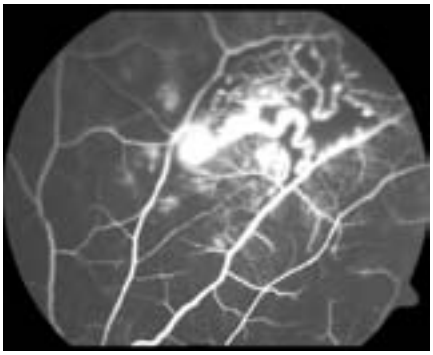


Fig. 12.16.2B: Retinal angioma—angiomatosis retinae with hard exudates

Retinal Vasculitis

- More commonly involves retinal vein (periphlebitis), rarely the arterioles (periarteritis)
- Active vasculitis appears as fluffy white perivascular cuffing better visible on FFA **(Figs 12.17.1A and B)**
- Severe periphlebitis may give rise to BRVO **(Figs 12.17.2A to C)** or CRVO **(Figs 12.17.3A and B)** like pictures

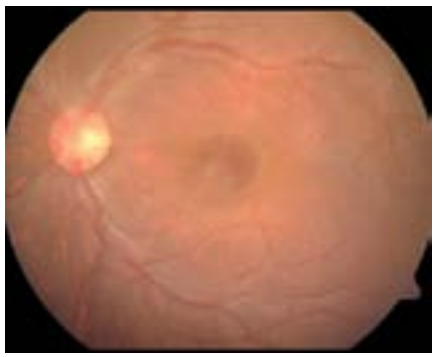


Fig. 12.17.1A: Vasculitis—periphlebitis

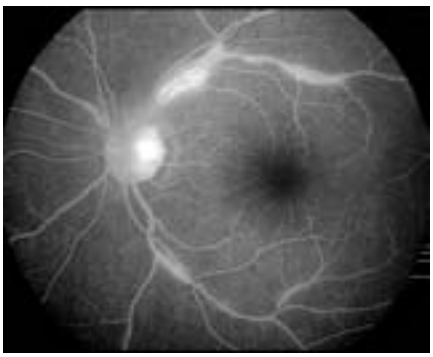


Fig. 12.17.1B: Vasculitis—periphlebitis

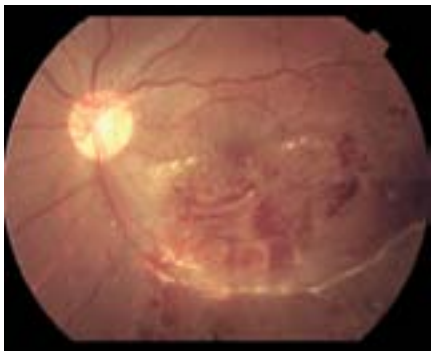


Fig. 12.17.2A: Vasculitis in inferotemporal BRVO

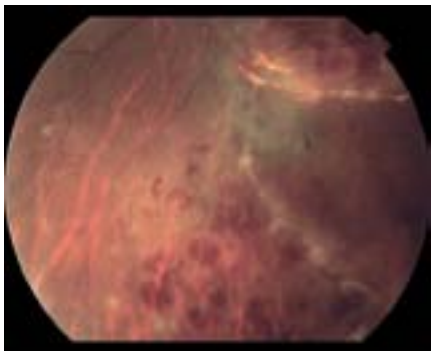


Fig. 12.17.2B: Vasculitis in inferotemporal BRVO

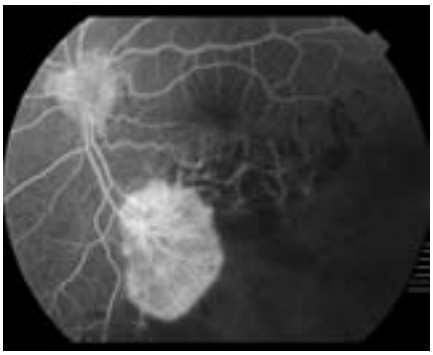


Fig. 12.17.2C: Vasculitis inferotemporal BRVO on FFA

- Periphlebitis in sarcoidosis may appear as 'candle wax dripping' around the veins **(Figs 12.17.4A and B)**
- Periarteritis may block a branch arteriole
- *Causes:*
  - *Periphlebitis:* Sarcoidosis, Eales' disease, intermediate uveitis, CMV retinitis, frosted branch angiitis, etc.
  - *Periarteritis:* Collagen disorders like, SLE, polyarteritis nodosa, etc.

Frosted Branch Angiitis

- Uncommon, unilateral or bilateral acute inflammation of the retinal vessels
- Sheathing of the blood vessels with appearance of frosted branches of a tree **(Figs 12.18.1A and B)**
- FFA findings confirm the diagnosis **(Figs 12.18.2A to C)**
- May be associated with CMV retinitis **(Fig. 12.18.3)**
- *Causes:* CMV retinitis; rubella; in patient with renal transplantation on immunosuppressants, etc.

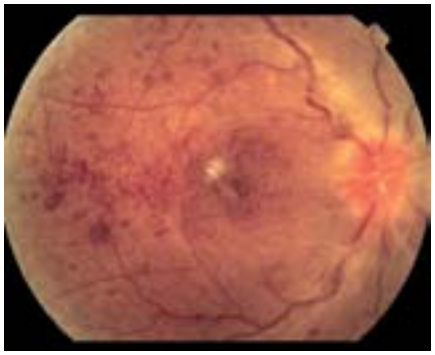


Fig. 12.17.3A: Vasculitis—central periphlebitis result in CRVO

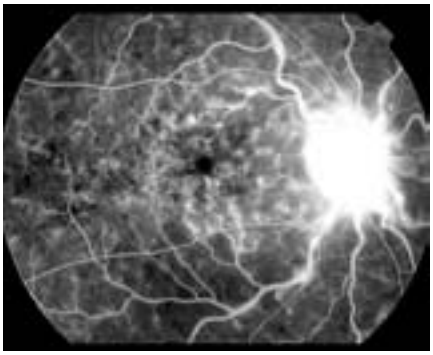


Fig. 12.17.3B: Vasculitis—central periphlebitis result in CRVO

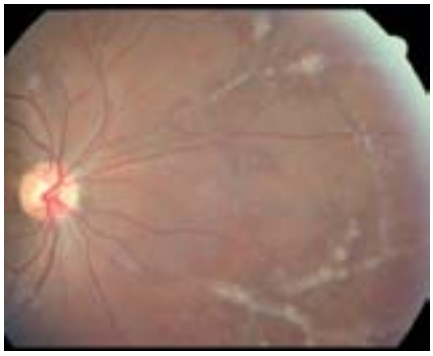


Fig. 12.17.4A: Vasculitis—periphlebitis—candle wax dripping

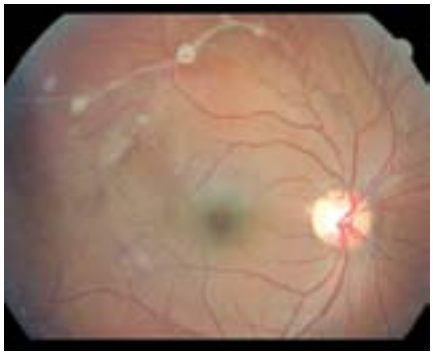


Fig. 12.17.4B: Vasculitis—periphlebitis—candle wax dripping

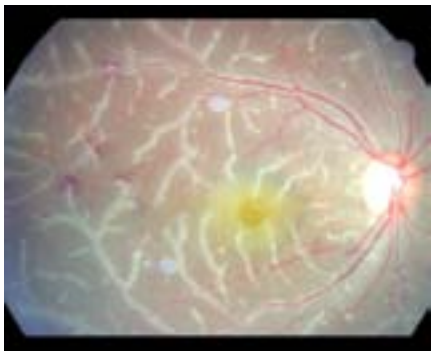


Fig. 12.18.1A: Frosted branch angiitis

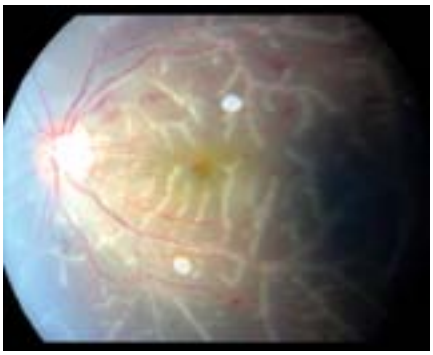


Fig. 12.18.1B: Frosted branch angiitis

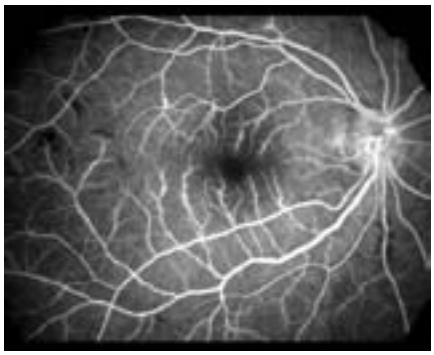


Fig. 12.18.2A: Frosted branch angiitis—FFA

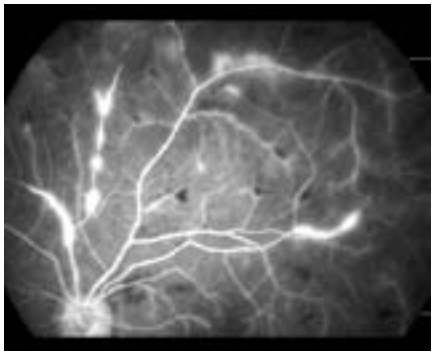


Fig. 12.18.2B: Frosted branch angiitis—FFA

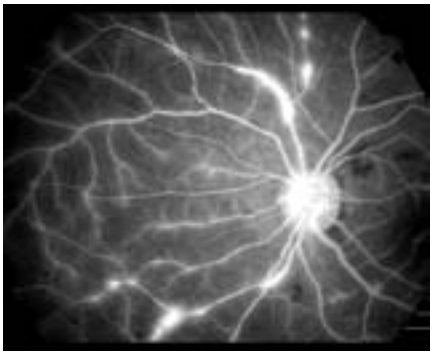


Fig. 12.18.2C: Frosted branch angiitis—FFA

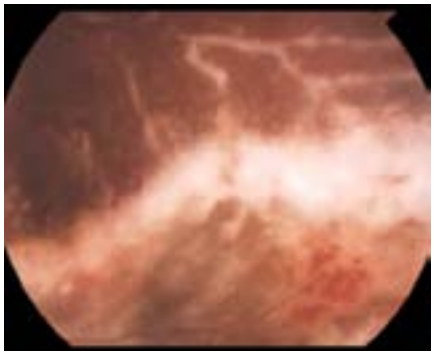


Fig. 12.18.3: Frosted branch angiitis—CMV retinitis

Retinal Exudates

Hard Exudates

- Yellow-waxy deposits or plaques with fairly distinct margins
- Most frequently seen in the posterior pole
- Vary in size and configuration
  - *Very few* **(Figs 12.19.1 and 12.19.2)**
  - *Isolated dense clumps* **(Figs 12.19.3 and 12.19.4)**
  - *Ring shaped* **(Figs 12.19.5 and 12.19.6)**
  - *Circinate pattern* around a leaking capillary **(Figs 12.19.6 to 12.19.8)**
  - May be with *dense subretinal exudates* **(Fig. 12.19.9)**
- *Causes:* Diabetic retinopathy, old BRVO, retinal macroaneurysm or hemangiomas.

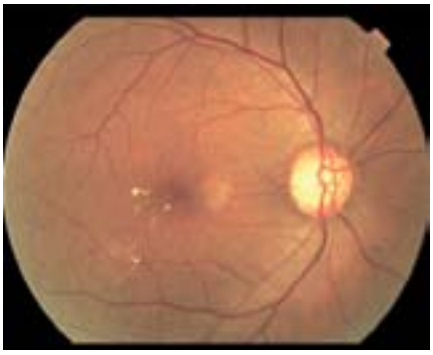


Fig. 12.19.1: Hard exudates—very few

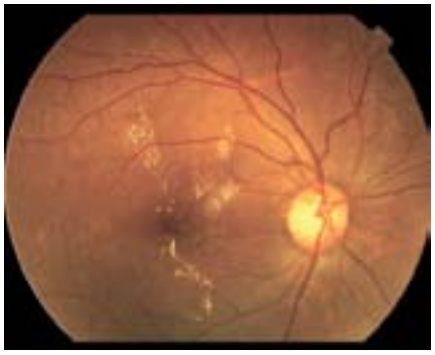


Fig. 12.19.2: Hard exudates—very few

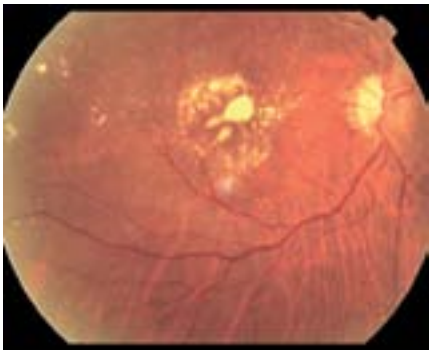


Fig. 12.19.3: Hard exudates—isolated clumps

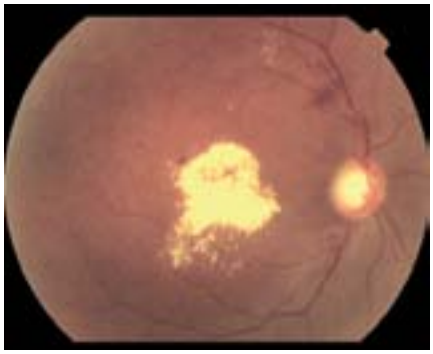


Fig. 12.19.4: Hard exudates—isolated dense clumps

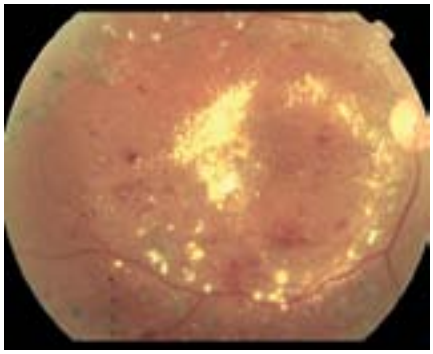


Fig. 12.19.5: Hard exudates—ring shaped

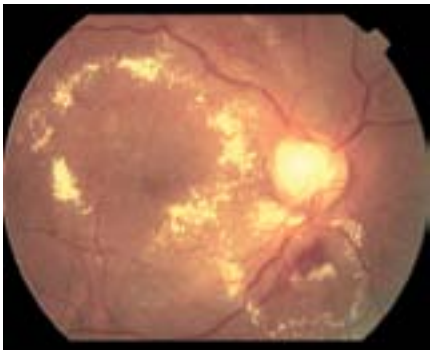


Fig. 12.19.6: Hard exudate—ring shaped and circinate pattern

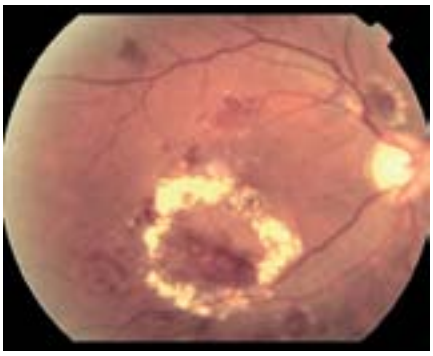


Fig. 12.19.7: Hard exudate—circinate pattern

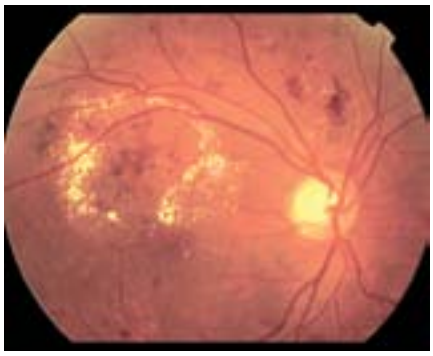


Fig. 12.19.8: Hard exudates—circinate pattern

Fig. 12.19.9: Hard exudates with dense subretinal exudates

Macular Stars

- Hard exudates around the macula often take the shape of a macular star **(Fig. 12.20.1)**
- May be incomplete **(Figs 12.20.2 and 12.20.3)** or complete **(Fig. 12.20.4)**
- Sometimes, associated with optic disk swelling as in neuroretinitis **(Fig. 12.20.5)** or papilledema
- *Causes:* Papilledema, malignant hypertension **(Fig. 12.20.6)**, neuroretinitis, peripapillary capillary angiomas, choroidal hemangioma **(Fig. 12.20.7)**.

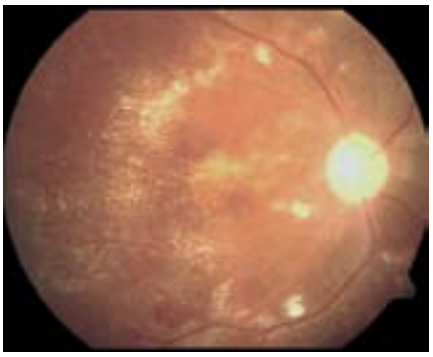


Fig. 12.20.1: Macular star

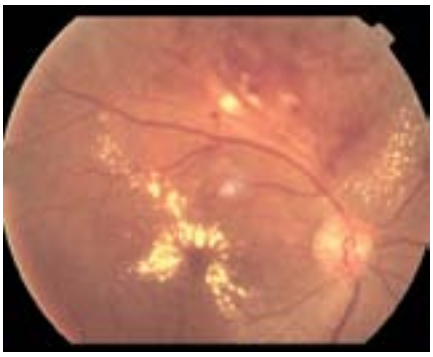


Fig. 12.20.2: Macular star—incomplete

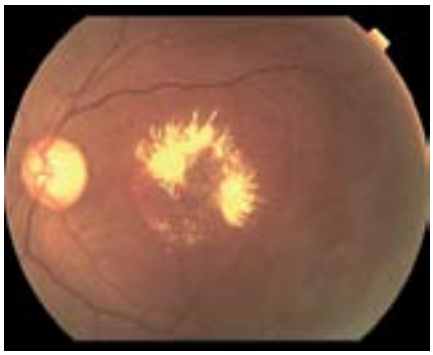


Fig. 12.20.3A: Macular star—incomplete

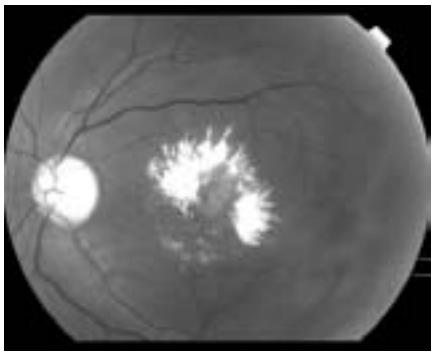


Fig. 12.20.3B: Macular star—incomplete

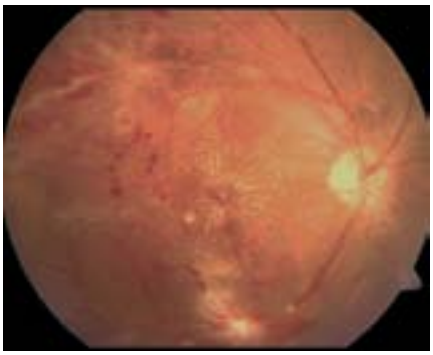


Fig. 12.20.4: Macular star—complete in BRVO

Fig. 12.20.5: Macular star in neuroretinitis

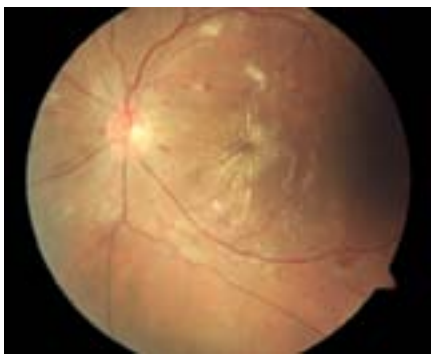


Fig. 12.20.6: Macular star in hypertensive retinopathy

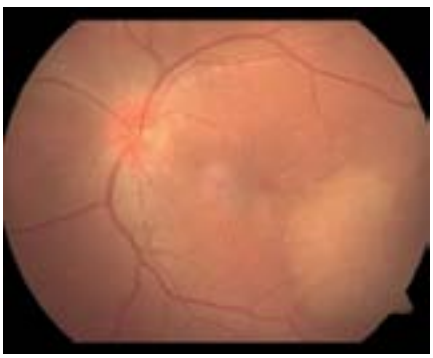


Fig. 12.20.7: Macular star in choroidal hemangioma

Soft Exudates or Cotton Wool Spots

- White, cotton-wool like spots with indistinct margins **(Figs 12.21.1 and 12.21.2)**
- May obscure the underlying small blood vessels **(Fig. 12.21.3)**
- Are the localized accumulation of axoplasmic debris in the nerve fiber layer as a result of ischemia
- May be associated with hard exudates also **(Fig. 12.21.4)**
- *Causes:* CRVO or BRVO, hypertensive retinopathy, preproliferative diabetic retinopathy, HIV microangiopathy, severe anemia, etc.

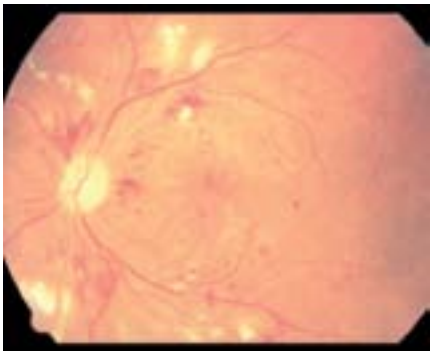


Fig. 12.21.1: Soft exudate—cotton wool spots

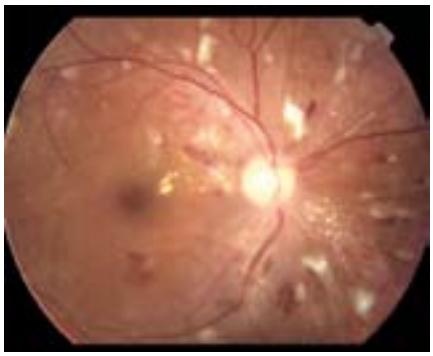


Fig. 12.21.2: Soft exudates—cotton wool spots in venous stasis retinopathy

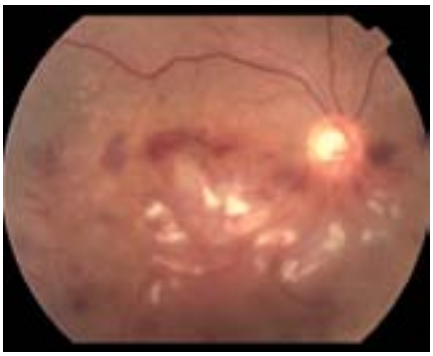


Fig. 12.21.3: Soft exudates in BRVO

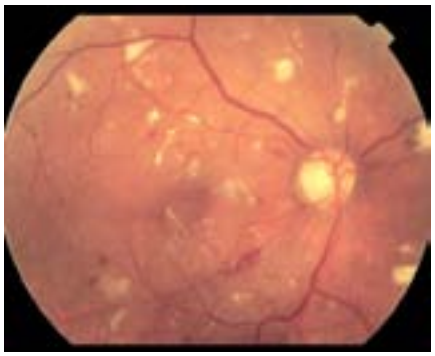


Fig. 12.21.4: Soft and hard exudates in diabetic retinopathy

Subretinal Exudates

- Yellowish-white, exudative lesions with indistinct margin in the central or peripheral retina **(Figs 12.22.1 and 12.22.2)** with blocked fluorescein
- Frequently associated with elevation of overlying retina **(Figs 12.22.3 and 12.22.4)**
- *Causes:* Chronic leakage from SRNVMs, Coats' diseases, toxocariasis.

Fig. 12.22.1: Subretinal exudates

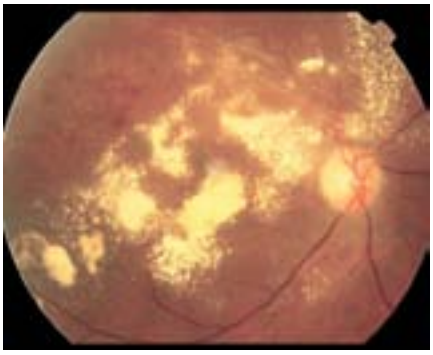


Fig. 12.22.2A: Subretinal exudates

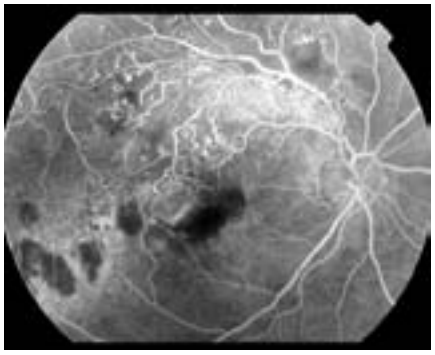


Fig. 12.22.2B: Subretinal exudates—blocked fluorescein

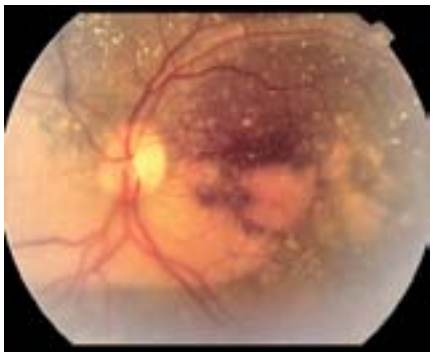


Fig. 12.22.3A: Subretinal exudates—in Coat's disease

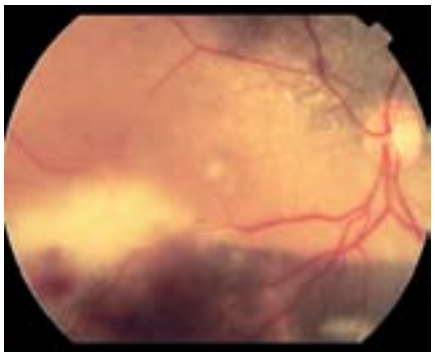


Fig. 12.22.3B: Subretinal exudates with elevated retina

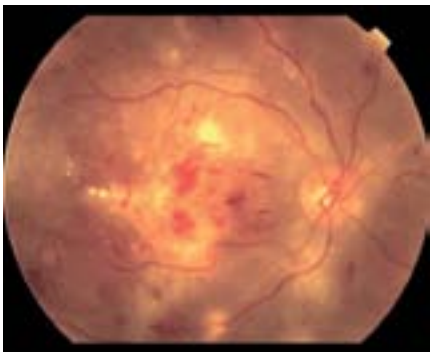


Fig. 12.22.4: Subretinal exudates with elevated retina in SRNVMs

Coats' Disease

- It usually affects boys between 18 months to 18 years of age.
- Main symptoms are decreased visual acuity and a 'white pupillary reflex'
- Yellowish-white exudative patches are seen behind the retinal blood vessels **(Figs 12.23.1 and 12.23.2)**
- Blood vessels have a tortuous course with aneurysms, fusiform dilations, and loop formations **(Figs 12.23.3 and 12.23.4)**.

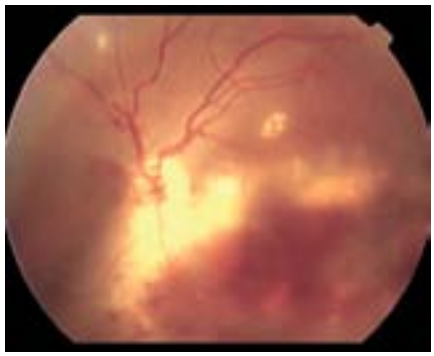


Fig. 12.23.1: Coats' disease

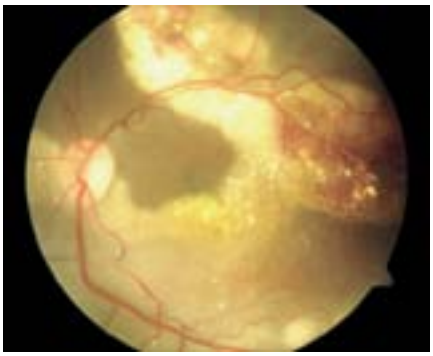


Fig. 12.23.2: Coats' disease

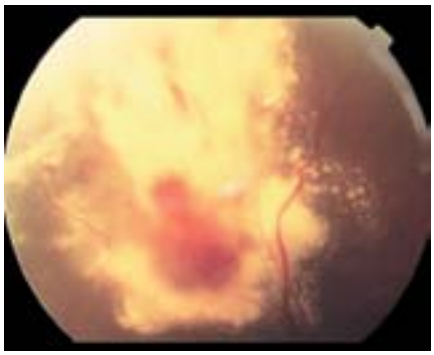


Fig. 12.23.3: Coats' disease

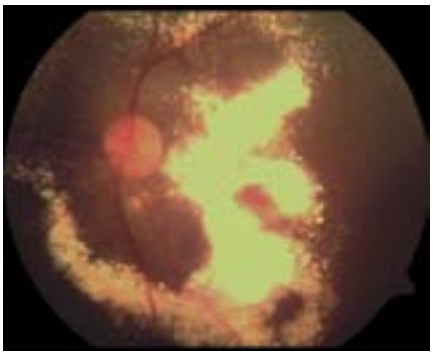


Fig. 12.23.4: Coats' disease

Inflammatory Retinal Lesions

Multiple Evanescent White Dot Syndrome (MEWDS)

- Rare, unilateral condition affects the young women
- Mild vitritis
- Multiple, very small white dot like lesion at the posterior pole and mid-periphery **(Figs 12.24.1A and B)**
- They are at the level of RPE and clearly visible on FFA
- Prognosis is always good.

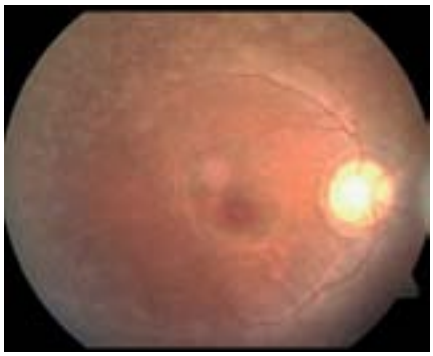


Fig. 12.24.1A: Multiple evanescent white dot syndrome (MEWDS)

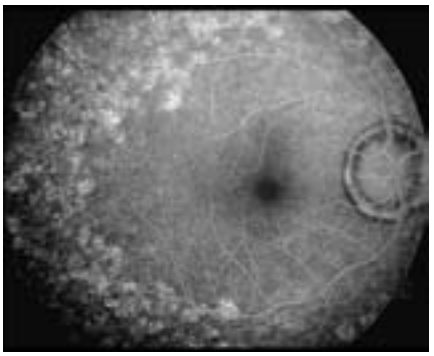


Fig. 12.24.1B: MEWDS on FFA

Punctate Inner Choroidopathy (PIC)

- Uncommon, bilateral condition affects young ladies with myopia
- No sign of vitritis
- Small yellowish-white indistinct choroidal lesions at the posterior pole **(Figs 12.25.1A to C)**
- May be associated with a serous retinal detachment
- Prognosis is always guarded.

Birdshot Retinochoroidopathy

- *See Chapter 6.*

Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE)

- Large cream-colored, placoid lesions at the posterior pole and midperiphery **(Fig. 12.26.1A)**
- Located at the level of RPE, appreciated by FFA: initial hypofluorescence followed by late diffuse staining **(Figs 12.26.1B and C)**
- *See also Chapter 6.*

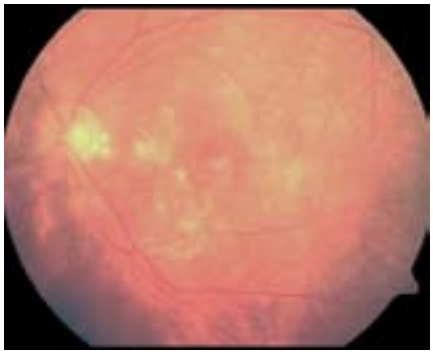


Fig. 12.25.1A: Punctate inner choroidopathy

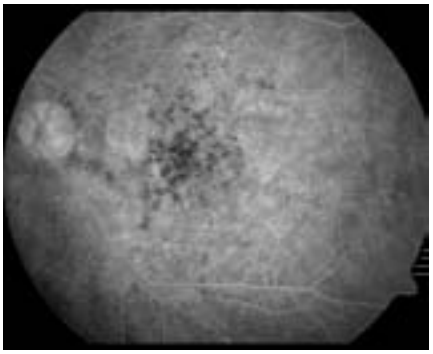


Fig. 12.25.1B: Punctate inner choroidopathy on FFA

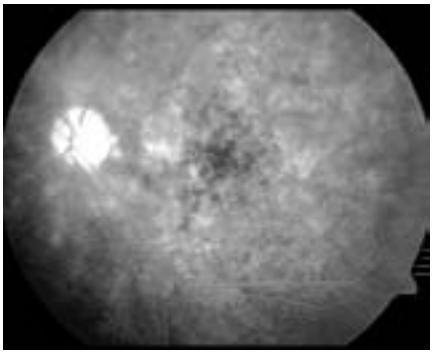


Fig. 12.25.1C: Punctate inner choroidopathy on FFA

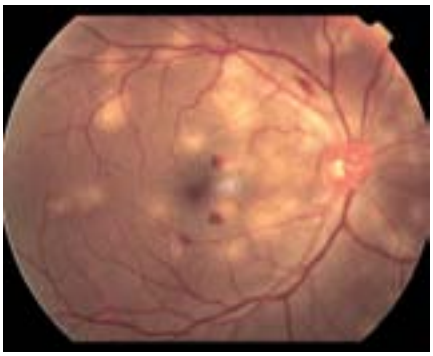


Fig. 12.26.1A: APMPPE

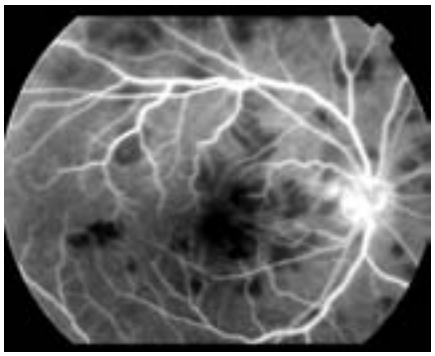


Fig. 12.26.1B: APMPPE on FFA

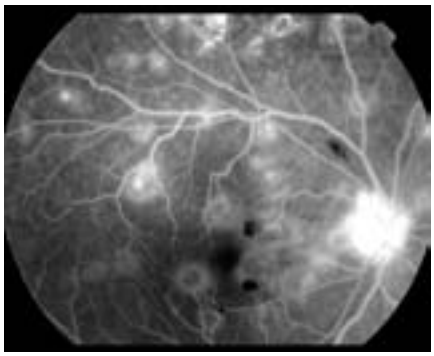


Fig. 12.26.1C: APMPPE on FFA

Multifocal Choroiditis with Panuveitis

- Unilateral or bilateral uncommon condition, affects middle-aged women
- Often recurrent in nature
- Associated anterior uveitis
- Small, discrete, fresh and old lesions mixed at mid-peripheral retina and posterior pole **(Fig. 12.27.1)**
- Variable areas of chorioretinal atrophy
- Lesions are more accurately visible on FFA **(Figs 12.27.2 and 12.27.3)**.

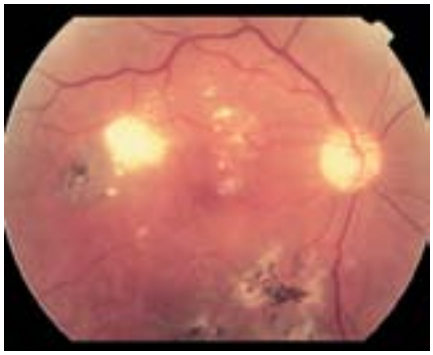


Fig. 12.27.1: Multifocal choroiditis

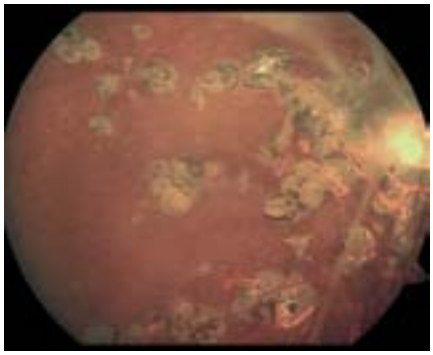


Fig. 12.27.2A: Active multifocal choroiditis—bilateral

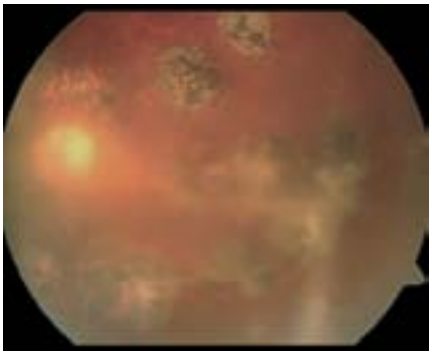


Fig. 12.27.2B: Active multifocal choroiditis with vitritis—bilateral

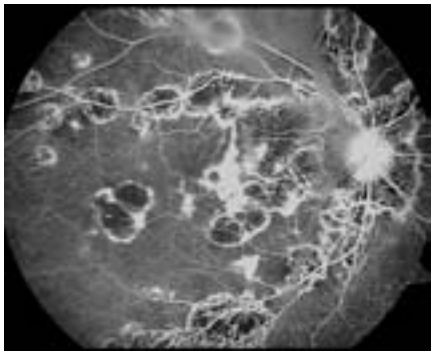


Fig. 12.27.2C: Active multifocal choroiditis—bilateral on FFA

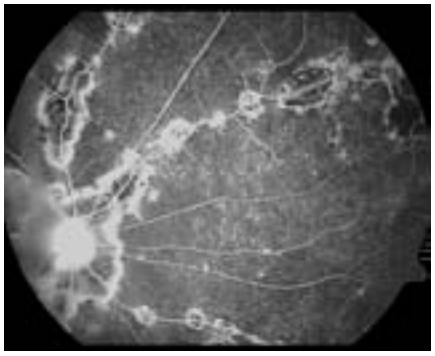


Fig. 12.27.2D: Active multifocal choroiditis—bilateral on FFA

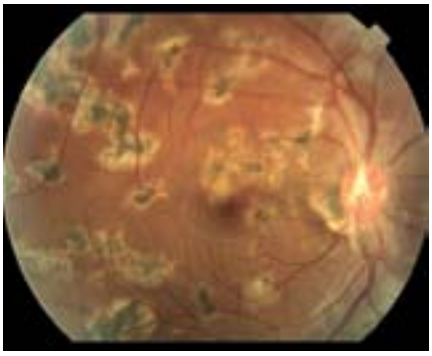


Fig. 12.27.3A: Active multifocal choroiditis—bilateral

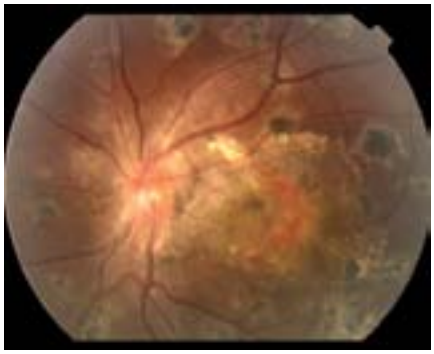


Fig. 12.27.3B: Active multifocal choroiditis—bilateral

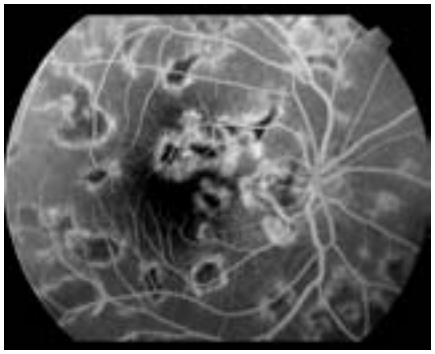


Fig. 12.27.3C: Active multifocal choroiditis—bilateral

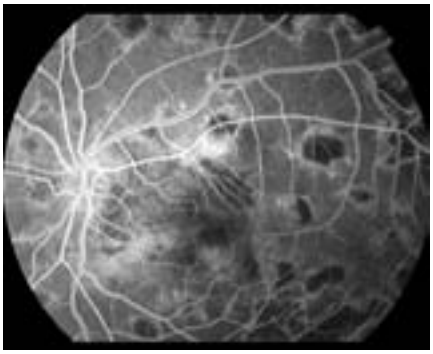


Fig. 12.27.3D: Active multifocal choroiditis—bilateral

Disseminated Choroiditis

- Unilateral or bilateral lesions, may occur at any age
- Multiple chorioretinal yellowish-white lesions scattered all over the fundus **(Figs 12.28.1A and B)**
- Associated with mild vitritis
- FFA shows typical lesions of initial hypofluorescence followed by hyperfluorescence **(Figs 12.28.1C and D)**
- Prognosis is fair.

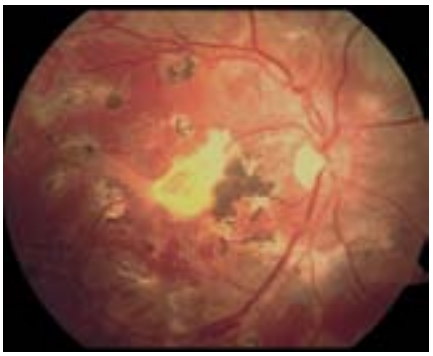


Fig. 12.28.1A: Disseminated choroiditis

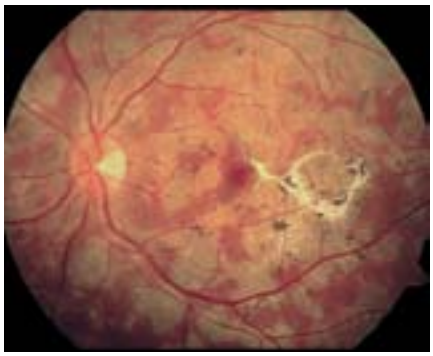


Fig. 12.28.1B: Disseminated choroiditis

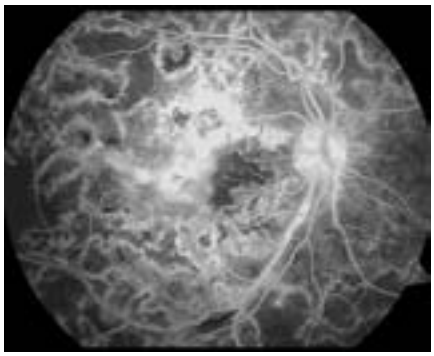


Fig. 12.28.1C: Disseminated choroiditis on FFA

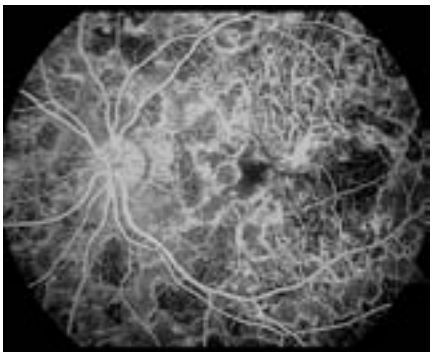


Fig. 12.28.1D: Disseminated choroiditis on FFA

Serpiginous Choroidopathy

- *See Chapter 6.*

Focal Toxoplasmosis

- Unilateral or bilateral protozoan infection caused by *Toxoplasma gondii* **(Fig. 12.29.1)**
- Fresh lesions, typically affects young individual between 15 and 30 years of age
- Moderate to severe vitritis
- Focal retinitis adjacent to the edge of old lesion **(Figs 12.29.2A and B)**
- May be with cystoid macular edema
- In acquired toxoplasmosis, it may not be associated with old scar

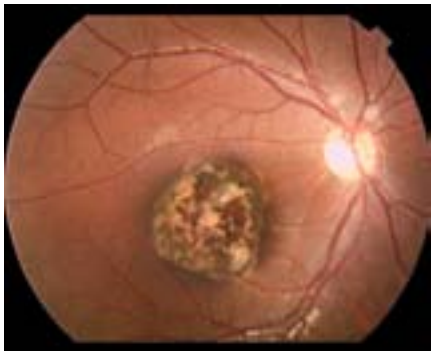


Fig. 12.29.1: Congenital toxoplasmosis—old scar

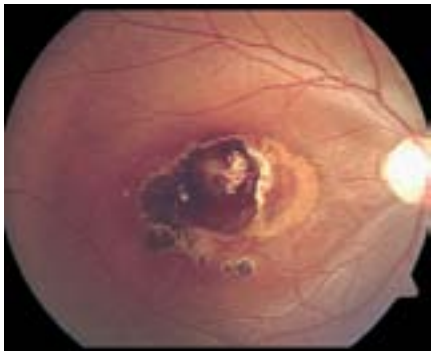


Fig. 12.29.2A: Focal lesion adjacent to the old *Toxoplasma* scar—RE

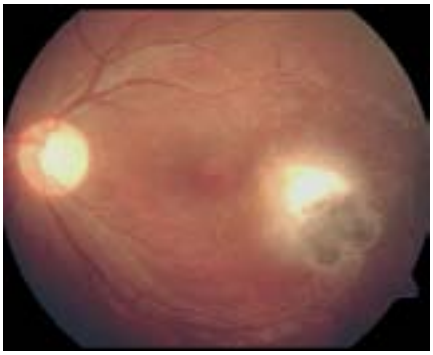


Fig. 12.29.2B: Toxoplasmosis—reactivation in other eye of same patient—LE

- Extensive late leakage on FFA **(Figs 12.29.2C and D)**
- May be associated with anterior granulomatous uveitis
- *See also in Chapter 6.*

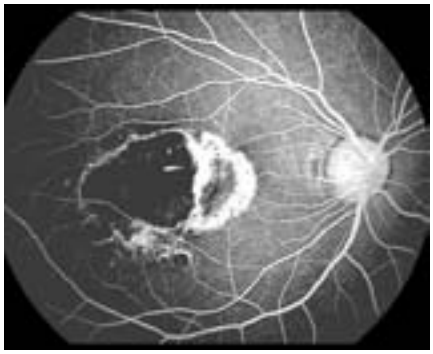


Fig. 12.29.2C: Focal lesion adjacent to the old *Toxoplasma* scar—RE

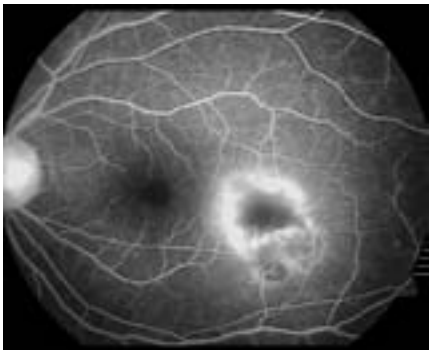


Fig. 12.29.2D: Toxoplasmosis—reactivation in other eye of same patient—LE

Toxocariasis

- *See also in Chapter 11.*

Candidiasis

- Rare, unilateral or bilateral fungal infection, typically affects drug addicts, severe diabetics or patients on immunosuppressants
- Solitary or multiple deep retinal infiltrates **(Fig. 12.30.1)**
- Cotton ball opacity in vitreous with vitritis **(Figs 12.30.2 and 12.30.3)**
- Prognosis is usually poor.

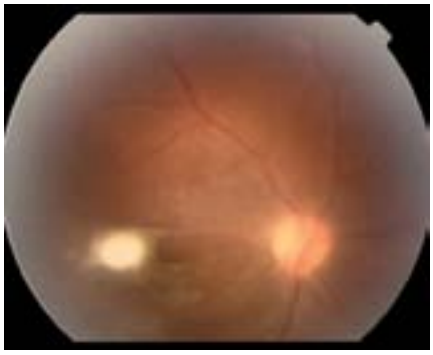


Fig. 12.30.1: Candidiasis—retinal lesion

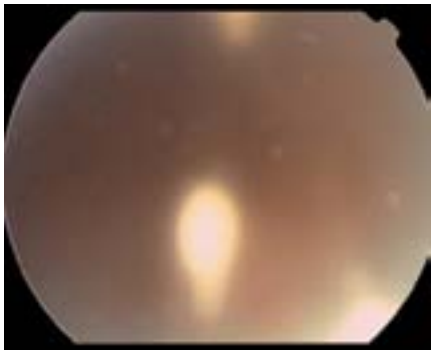


Fig. 12.30.2: Candidiasis—cotton ball exudate in vitreous

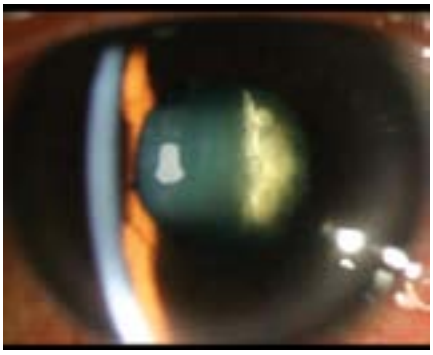


Fig. 12.30.3: Candidiasis—cotton ball exudates in the vitreous

Cytomegalovirus (CMV) Retinitis

- 25% of patients of AIDS suffer from CMV retinitis and bilaterality in 50% cases
- Yellowish-white areas of retinal infiltration with advancing brush-like border **(Figs 12.31.1 and 12.31.2)**

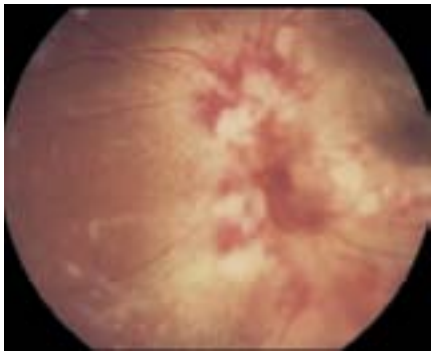


Fig. 12.31.1: CMV retinitis—disk and peripapillary involvement

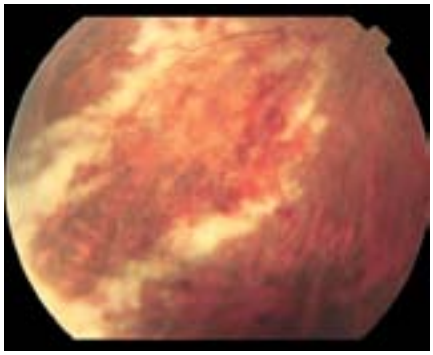


Fig. 12.31.2: CMV retinitis

- Slowly progressive and typically start at the posterior pole and spread along the vascular arcades **(Figs 12.31.3A and B)**
- Hemorrhages in the midst of retinitis
- Diffuse atrophy in late stage
- *Other fundal changes in AIDS patients:*
  - *Retinal microvasculopathy* as evident by cotton wool spots, superficial and deep hemorrhage **(Fig. 12.31.4)**
  - Immunocomplex deposits in the precapillary arterioles
  - Central retinal venous occlusion like picture **(Figs 12.31.5A and B)**
  - Immune-recovery uveitis **(Fig. 12.31.6)**
- *Other causes of CMV retinitis:* Patient on cytotoxic chemotherapy or renal transplant patient.

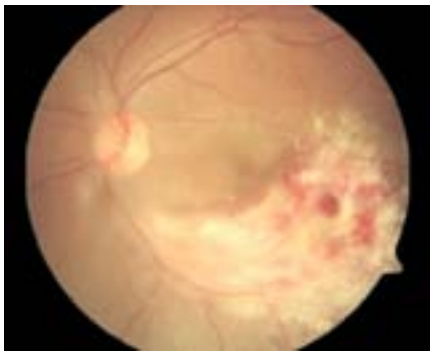


Fig. 12.31.3A: CMV retinitis—BRVO

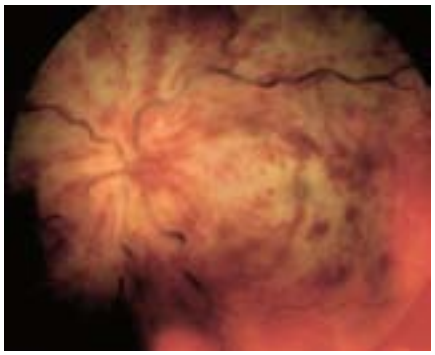


Fig. 12.31.3B: CMV retinitis—CRVO

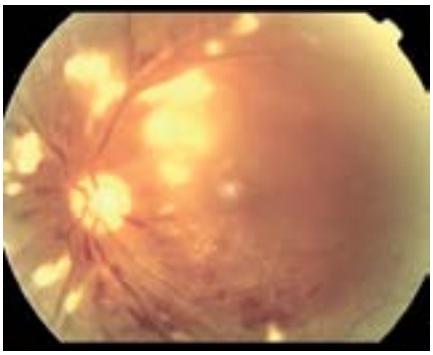


Fig. 12.31.4: Retinal microvasculopathy—HIV

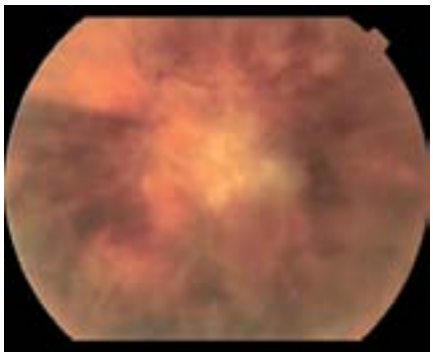


Fig. 12.31.5A: CMV retinitis—disk involvement with CRVO

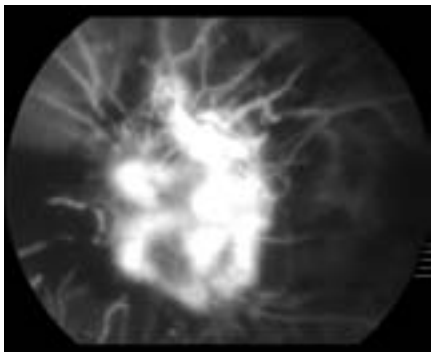


Fig. 12.31.5B: CMV retinitis—disk involvement with CRVO

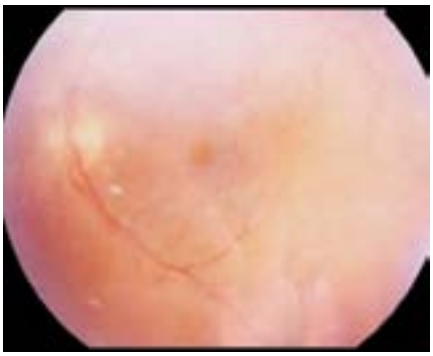


Fig. 12.31.6: Immune-recovery uveitis

Acute Retinal Necrosis (ARN)

- Extremely rare, devastating necrotizing retinitis affects otherwise healthy subjects
- Anterior uveitis and vitritis
- Confluent yellowish-white patches with sheathing and occlusion of blood vessels **(Figs 12.32.1 and 12.32.2)**
- Most eyes develop multiple retinal holes with rhegmatogenous retinal detachment
- Progression is faster and macula often gets spared.

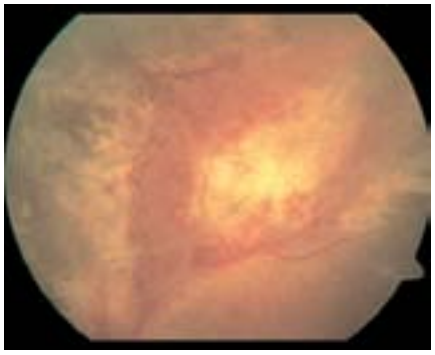


Fig. 12.32.1: Acute retinal necrosis—CMV retinitis

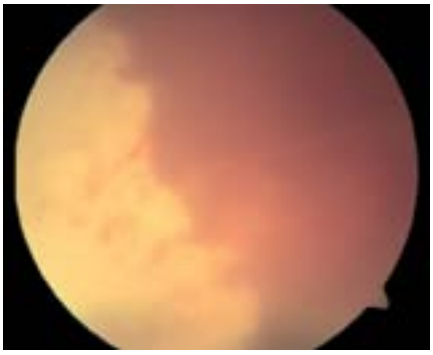


Fig. 12.32.2: Acute retinal necrosis—CMV retinitis

Progressive Outer Retinal Necrosis (PORN)

- A severe variant of acute retinal necrosis in patients with AIDS
- May be bilateral in 70% cases
- Deep multifocal lesions giving rise to peripheral necrotizing retinitis **(Fig. 12.33.1)**
- No occlusive vasculitis
- Very rapid progression with involvement of optic disc and macula

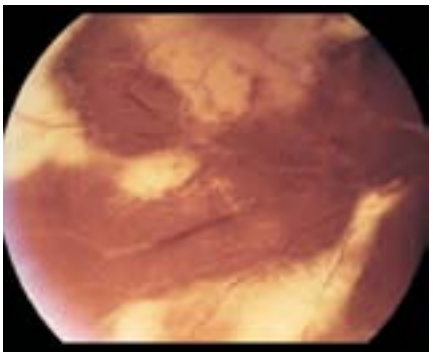


Fig. 12.33.1: Progressive outer retinal necrosis

Vogt-Koyanagi-Harada Syndrome

- *See Chapter 6*

Sympathetic Ophthalmia

- *See Chapter 6*

Tuberculous Choroiditis

- Not so uncommon cause of uveitis specially in developing countries
- *May appear in many ways:*
  - Chronic granulomatous anterior uveitis
  - Active central multifocal choroiditis: unilateral **(Figs 12.34.1A and B)** or bilateral in some cases **(Figs 12.34.2A and B)**
  - Vasculitis or periphlebitis
  - Occasionally as unilateral solitary choroidal tubercular granuloma **(Figs 12.34.3 and 12.34.4)**

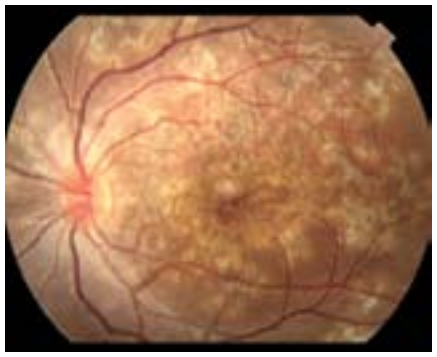


Fig. 12.34.1A: Tubercular active central multifocal choroiditis

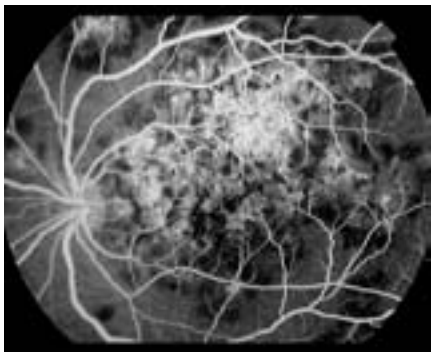


Fig. 12.34.1B: Tubercular active central multifocal choroiditis on FFA

Fig. 12.34.2A: Tubercular granuloma - bilateral—RE

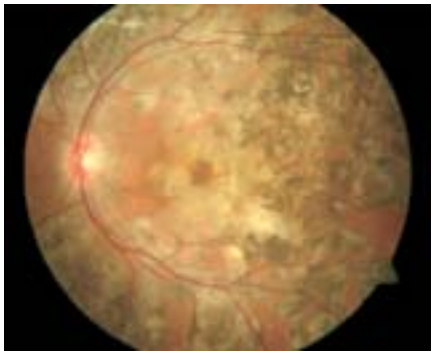


Fig. 12.34.2B: Tubercular granuloma - bilateral—LE

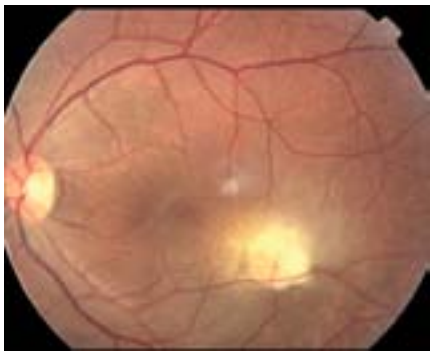


Fig. 12.34.3: Active tubercular granuloma

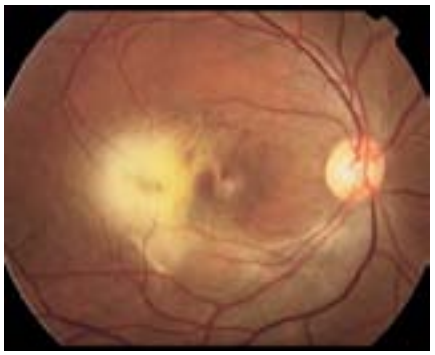


Fig. 12.34.4A: Tubercular granuloma

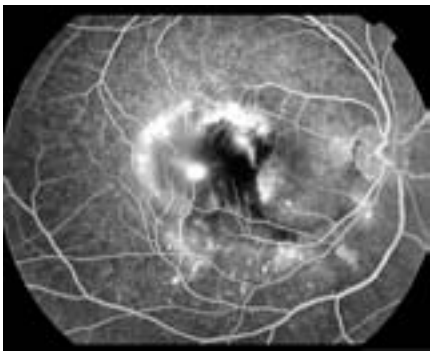


Fig. 12.34.4B: Tubercular granuloma on FFA

Macular Lesions (Maculopathies)

Hereditary Maculopathies

Stargardt's Macular Dystrophy

- Rare, bilateral, recessive condition starts around 14 to 16 years
- May results in severe visual loss within five years
- *Macula may appear as:*
  - Isolated atrophic maculopathy **(Figs 12.35.1A and B)**
  - Atrophic maculopathy with fish tail-shaped flecks **(Figs 12.35.2A to D)**
  - With diffuse flecks around the posterior pole **(Figs 12.35.3A to D)**
  - Beaten metal at the posterior pole
- Prognosis is always poor.

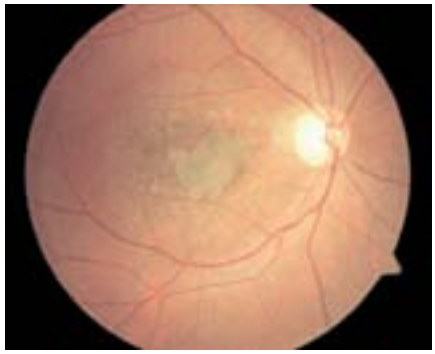


Fig. 12.35.1A: Stargardt's—atrophic maculopathy

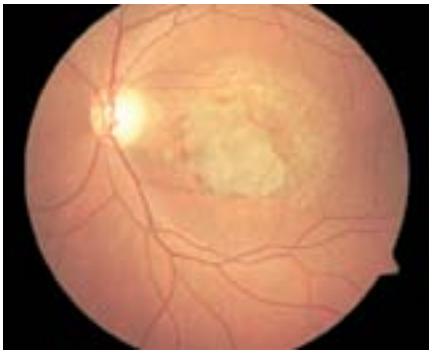


Fig. 12.35.1B: Stargardt's—atrophic maculopathy

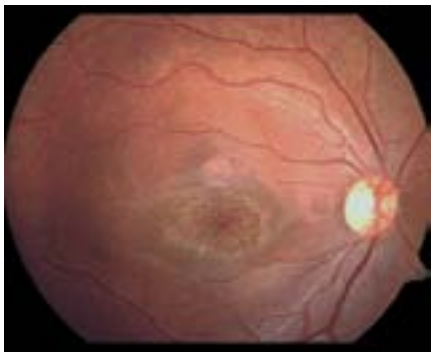


Fig. 12.35.2A: Stargardt's atrophic maculopathy with fish tail-shaped flecks

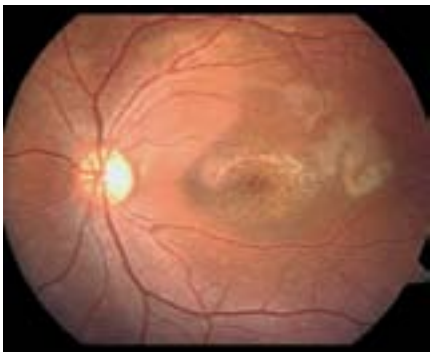


Fig. 12.35.2B: Stargardt's atrophic maculopathy with fish tail-shaped flecks

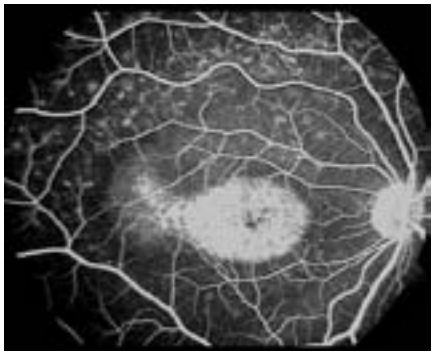


Fig. 12.35.2C: Stargardt's atrophic maculopathy with fish tail-shaped flecks—FFA

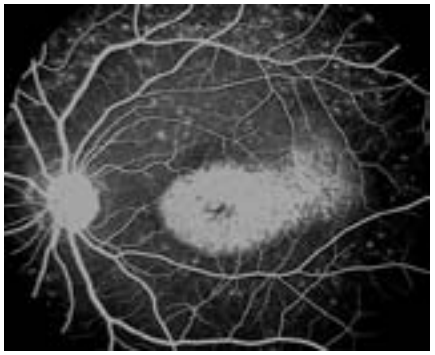


Fig. 12.35.2D: Stargardt's atrophic maculopathy with fish tail-shaped flecks—FFA

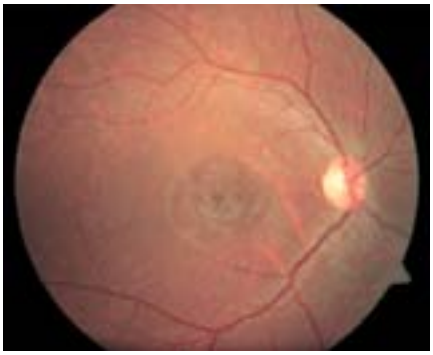


Fig. 12.35.3A: Stargardt's—maculopathy with flecks

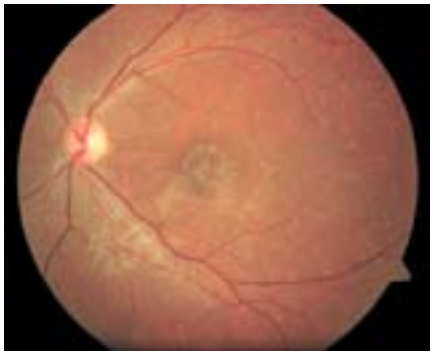


Fig. 12.35.3B: Stargardt's—maculopathy with flecks

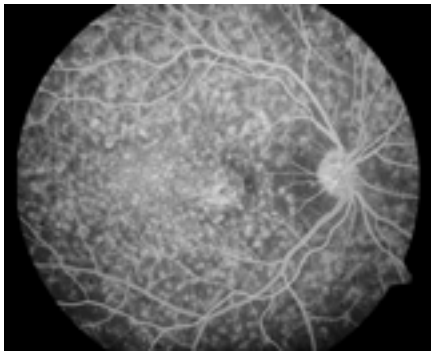


Fig. 12.35.3C: Stargardt's—maculopathy with flecks

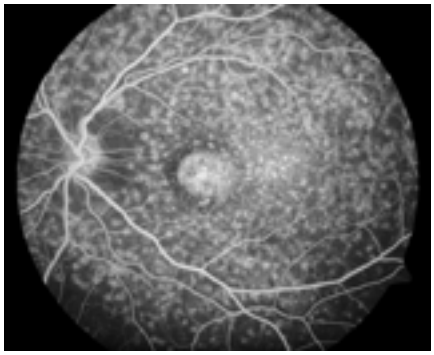


Fig. 12.35.3D: Stargardt's—maculopathy with flecks

Central Areolar Choroidal Dystrophy

- Rare, bilateral condition with dominant inheritance
- Loss of central vision at around 50 to 60 years
- Well circumscribed, bilateral atrophic macular lesion of 1 to 3 disk diameter **(Figs 12.36.1A to D)**
- Pigmented spots may be seen within the lesions **(Figs 12.36.2A and B)**
- Not associated with drusen.

Best's Vitelliform Macular Dystrophy

- Very rare, dominant hereditary condition starts in childhood
- Usually bilateral, but often asymmetrical
- Clinically, it has the *following stages*:
  - *Vitelliform stage:* Egg yolk-like macular lesion **(Fig. 12.37.1)**; also called "sunny side up" macular lesion
  - *Pseudohypopyon stage:* With partial absorption **(Figs 12.37.2A and B)**

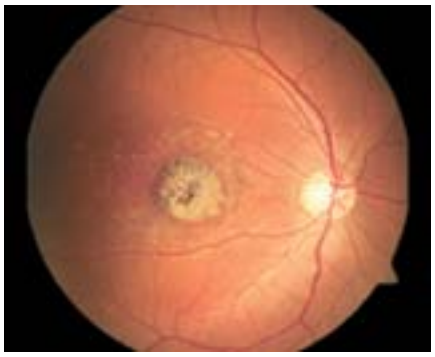


Fig. 12.36.1A: Central areolar choroidal dystrophy—RE

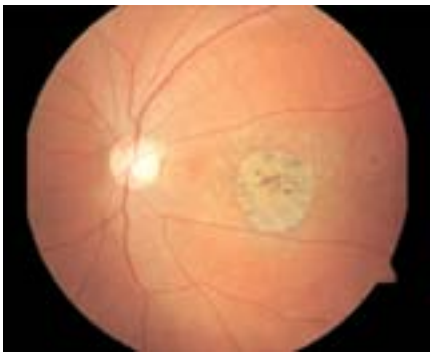


Fig. 12.36.1B: Central areolar choroidal dystrophy—LE

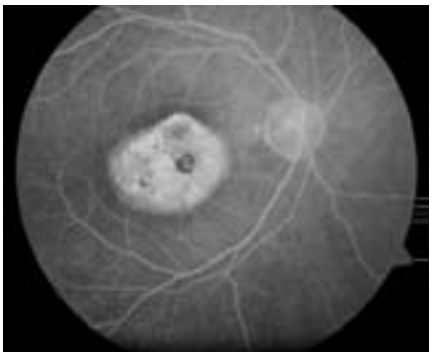


Fig. 12.36.1C: Central areolar choroidal dystrophy

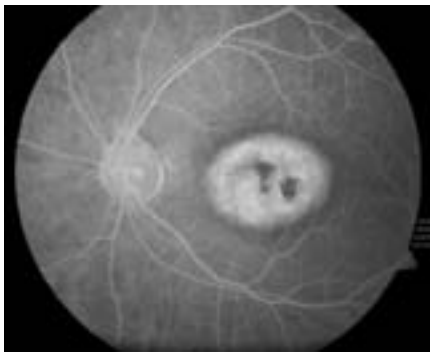


Fig. 12.36.1D: Central areolar choroidal dystrophy

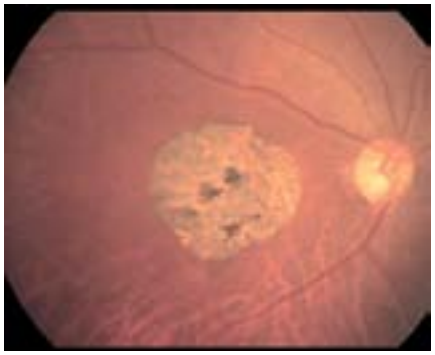


Fig. 12.36.2A: Central areolar choroidal dystrophy

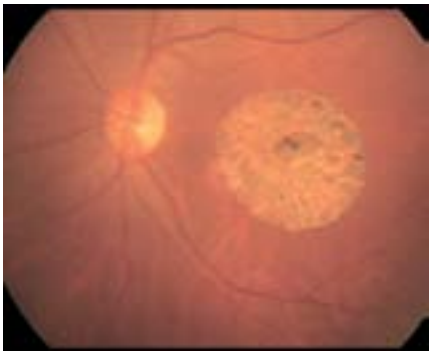


Fig. 12.36.2B: Central areolar choroidal dystrophy

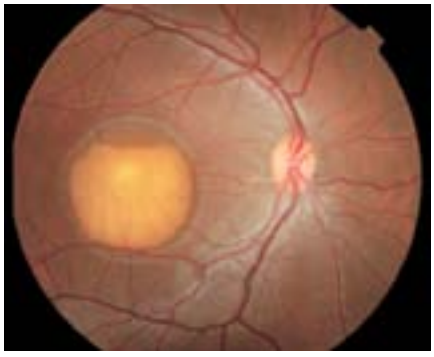


Fig. 12.37.1: Vitelliform dystrophy—egg yolk lesion (sunny-side up)

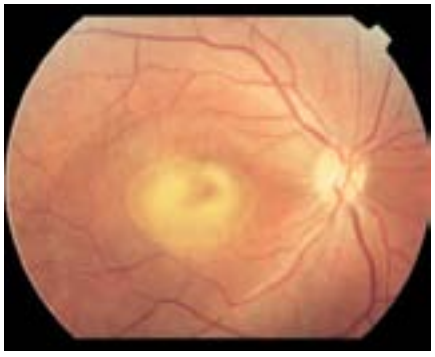


Fig. 12.37.2A: Vitelliform dystrophy—pseudohypopyon stage

Fig. 12.37.2B: Vitelliform dystrophy—pseudohypopyon stage

  - *Vitelliruptive stage:* Scrambled egg appearance macular lesion **(Fig. 12.37.3)**
  - *End stage:* With atrophic maculopathy and disciform scarring **(Fig. 12.37.4)**
- Family member may be diagnosed early in *pre-vitelliform stage* by electro-oculogram (EOG)
- Prognosis is always poor.

Hereditary Cone Dystrophy (Bull's Eye Maculopathy)

- Very rare bilateral macular dystrophy which presents as Bull's eye maculopathy
- Autosomal dominant or X-linked recessive
- Presents between first to third decade
- *Bull's eye macular lesions*, with golden reflex **(Figs 12.38.1A to D)**
- Vascular attenuation and temporal pallor may be present
- *Other causes of Bull's eye maculopathy:*
  - *Chloroquine toxicity:* Dose-dependent and occurs after prolonged use, especially if used for rheumatoid arthritis; associated retinal vascular attenuation and may not be reversible in some cases **(Figs 12.38.2A to D)**
  - *Late stage of Stargardt's disease* **(Figs 12.38.3A to D)**
  - *Inverse retinitis pigmentosa.*

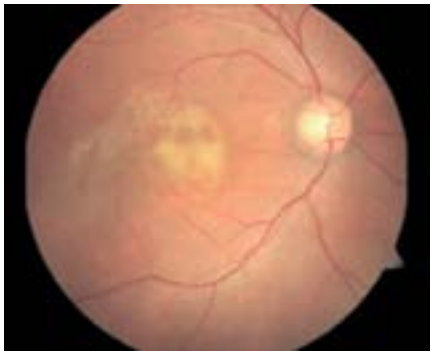


Fig. 12.37.3: Vitelliform dystrophy—scrambled egg appearance

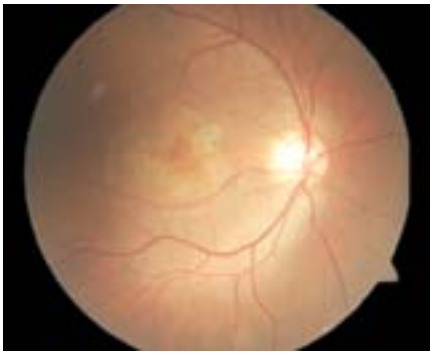


Fig. 12.37.4: Vitelliform dystrophy—end stage with disciform scarring

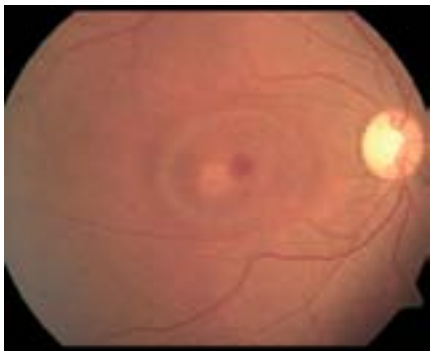


Fig. 12.38.1A: Hereditary cone dystrophy—Bull's eye maculopathy—RE

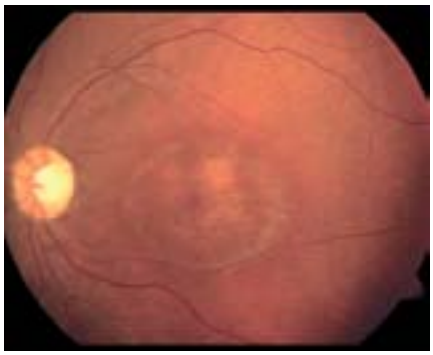


Fig. 12.38.1B: Hereditary cone dystrophy—Bull's eye maculopathy—LE

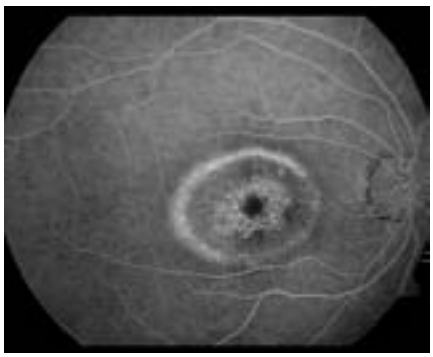


Fig. 12.38.1C: Hereditary cone dystrophy—Bull's eye maculopathy—RE

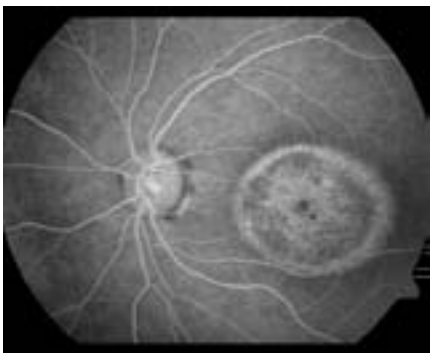


Fig. 12.38.1D: Hereditary cone dystrophy—Bull's eye maculopathy—LE

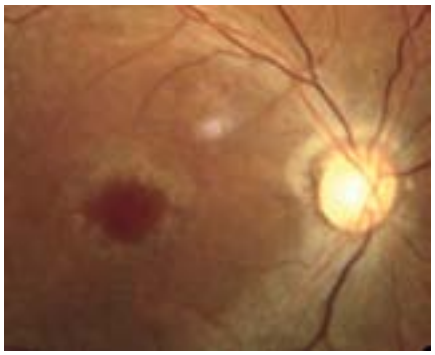


Fig. 12.38.2A: Bull's eye maculopathy—chloroquine toxicity—RE

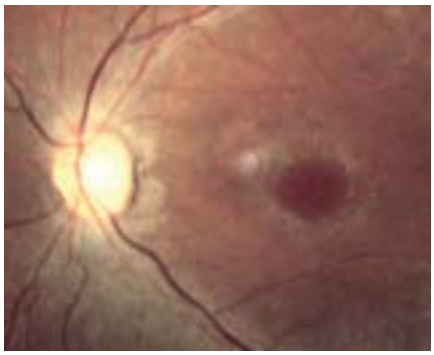


Fig. 12.38.2B: Bull's eye maculopathy—chloroquine toxicity—LE

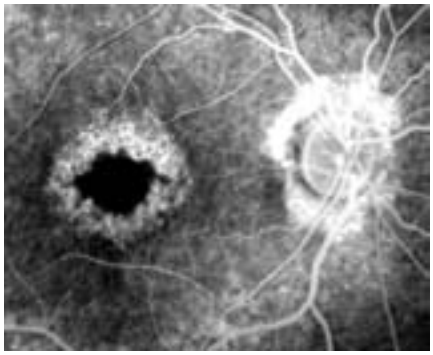


Fig. 12.38.2C: Bull's eye maculopathy—chloroquine toxicity—RE

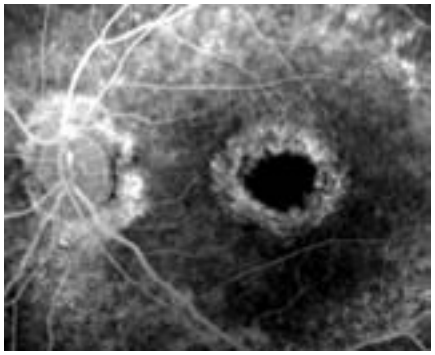


Fig. 12.38.2D: Bull's eye maculopathy—chloroquine toxicity—LE

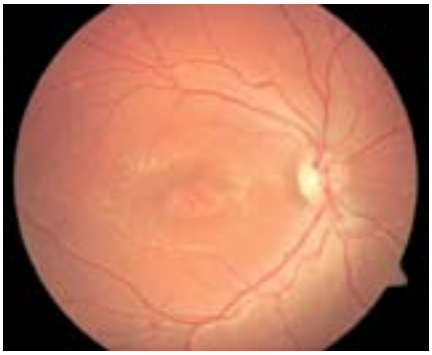


Fig. 12.38.3A: Bull's eye maculopathy—Stargardt's disease—RE

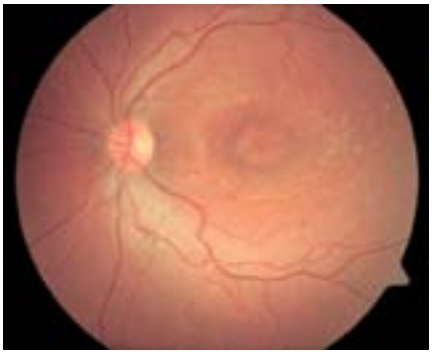


Fig. 12.38.3B: Bull's eye maculopathy—Stargardt's disease—LE

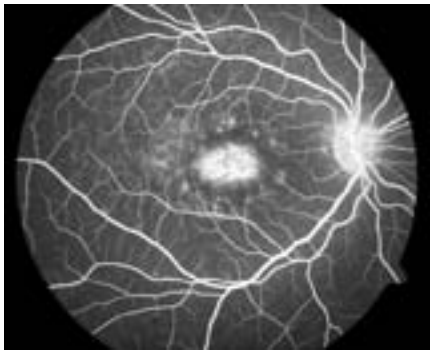


Fig. 12.38.3C: Bull's eye maculopathy—Stargardt's disease—RE

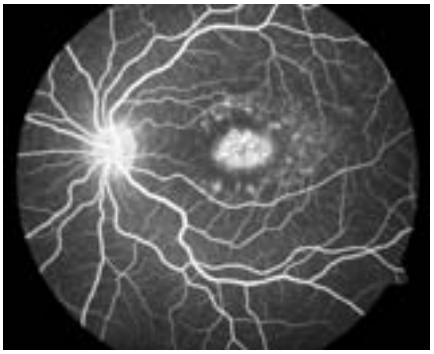


Fig. 12.38.3D: Bull's eye maculopathy—Stargardt's disease—LE

Myopic Maculopathies

- Common, bilateral, often asymmetrical macular lesions in adults with high and pathological myopia
- Lesions are usually progressive
- *Changes are:*
  - Lacquer cracks at posterior pole **(Figs 12.39.1A and B)**
  - Atrophic maculopathy **(Figs 12.39.2 and 12.39.3A)**

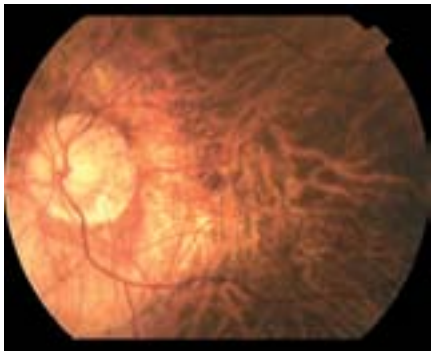


Fig. 12.39.1A: Myopic maculopathy—lacquer cracks

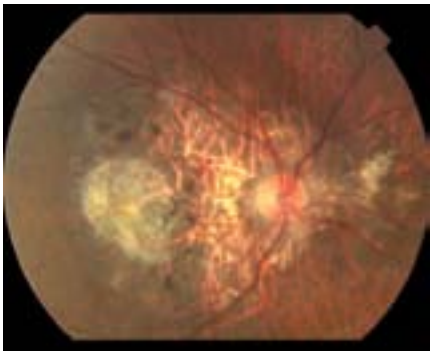


Fig. 12.39.1B: Myopic maculopathy—lacquer cracks and Fuchs' spot

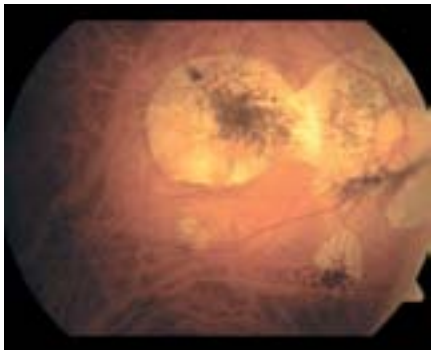


Fig. 12.39.2: Myopic atrophic maculopathy

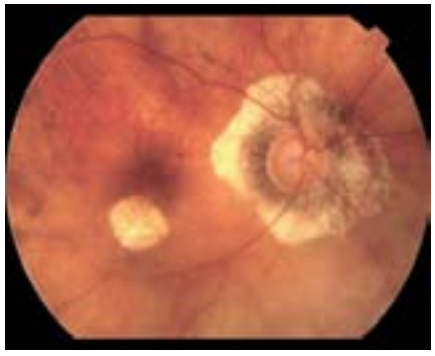


Fig. 12.39.3A: Myopic maculopathy—atrophic maculopathy—RE

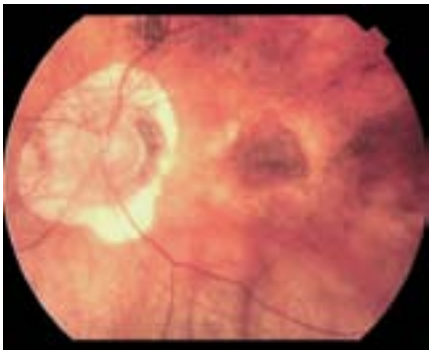


Fig. 12.39.3B: Myopic maculopathy—CNVM—LE

- Macular hemorrhage with choroidal neovascularization **(Figs 12.39.3B to 12.39.5)**
- Macular hole with or without retinal detachment **(Fig. 12.39.6)**
- Foster-Fuchs' pigmented spot at fovea **(Figs 12.39.7A and B)**

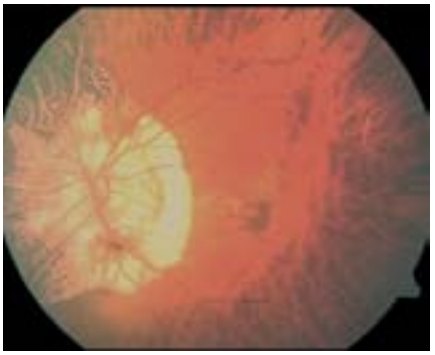


Fig. 12.39.4A: Myopic maculopathy—hemorrhage from CNV

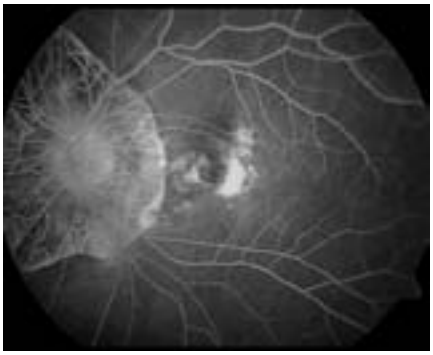


Fig. 12.39.4B: Myopic maculopathy—hemorrhage from CNV

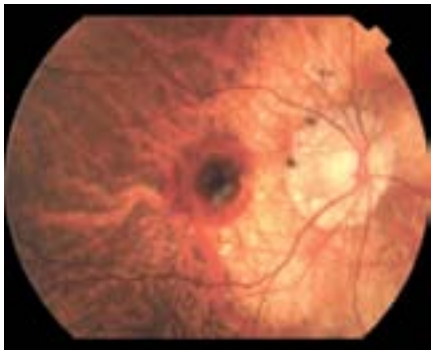


Fig. 12.39.5A: Myopic maculopathy—coin shaped hemorrhage from CNVM

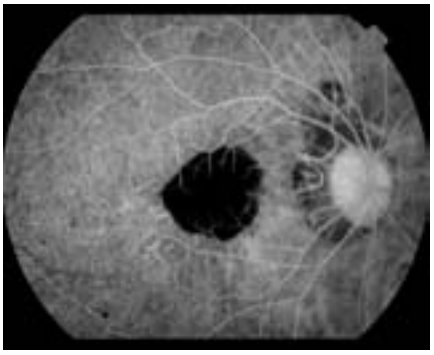


Fig. 12.39.5B: Myopic maculopathy—coin shaped hemorrhage from CNVM

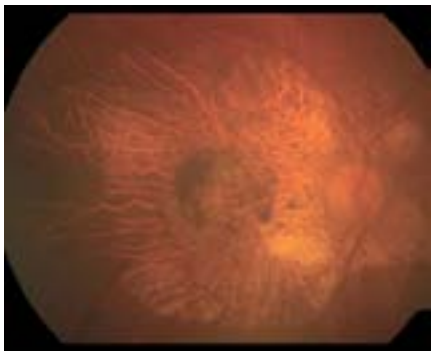


Fig. 12.39.6: Myopic macular hole

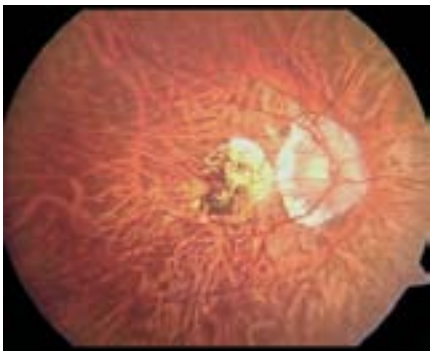


Fig. 12.39.7A: Myopic maculopathy—Foster-Fuchs' spots—RE

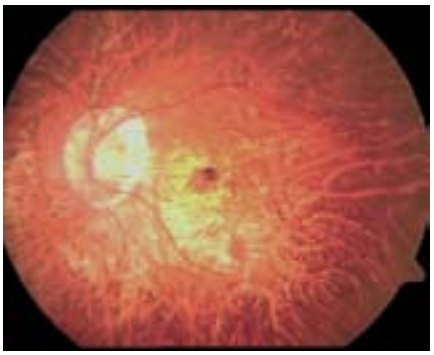


Fig. 12.39.7B: Myopic maculopathy—Foster-Fuchs' spots—LE

- Posterior staphyloma **(Figs 12.39.8 and 12.39.9)**
- Overall gross tessellation of the entire fundus including the macular area **(Fig. 12.39.10)**

- Peripheral retinal degenerative lesions are also common in myopia.

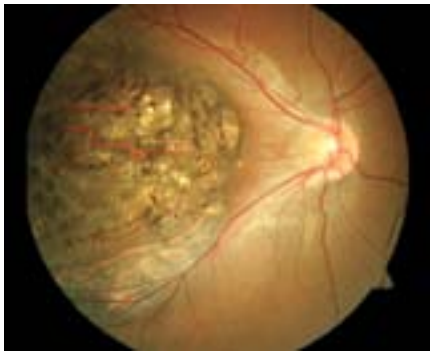


Fig. 12.39.8A: Myopic maculopathy—posterior staphyloma

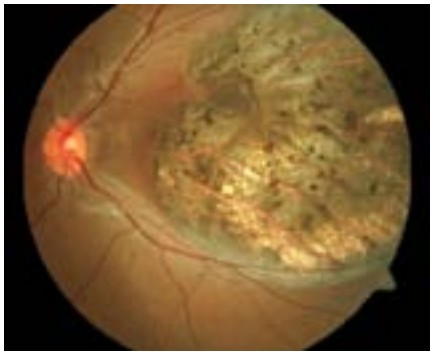


Fig. 12.39.8B: Myopic maculopathy—posterior staphyloma

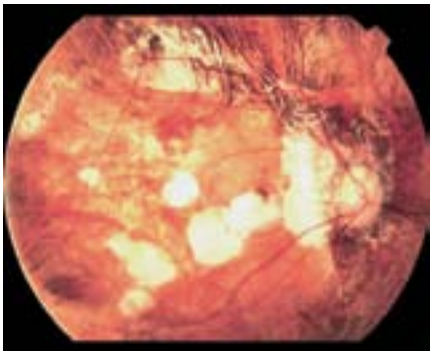


Fig. 12.39.9A: Myopic maculopathy—posterior staphyloma

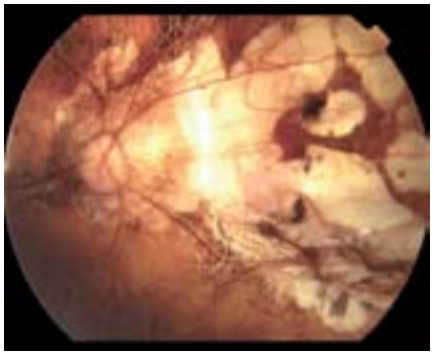


Fig. 12.39.9B: Myopic maculopathy—posterior staphyloma

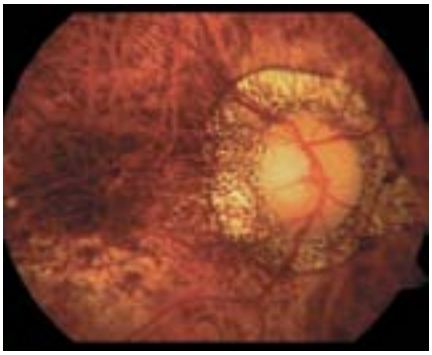


Fig. 12.39.10: Myopic maculopathy—tessellated fundus

Dry Age-Related Macular Degenerations

- Dry AMD is very common, bilateral condition, often bilateral
- Most common form of macular degeneration
- Affecting the patient above sixty years of age
- Sharply demarcated areas of macular hypopigmentation or depigmentation **(Figs 12.40.1A and B)**
- Associated with hard drusen **(Figs 12.40.2A and B)**
- Visible underlying choroidal vessels (geographical atrophy)
- FFA shows widespread window defect with late staining **(Figs 12.40.2C and D)**.

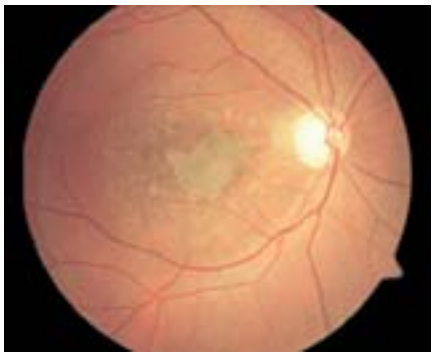


Fig. 12.40.1A: Dry AMD—geographical atrophy—RE

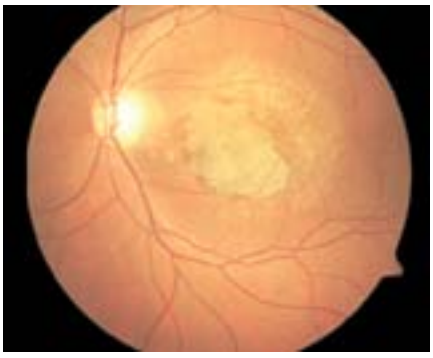


Fig. 12.40.1B: Dry AMD—geographical atrophy—LE

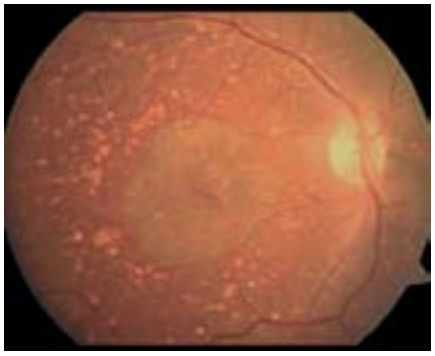


Fig. 12.40.2A: Dry AMD—with hard drusen—RE

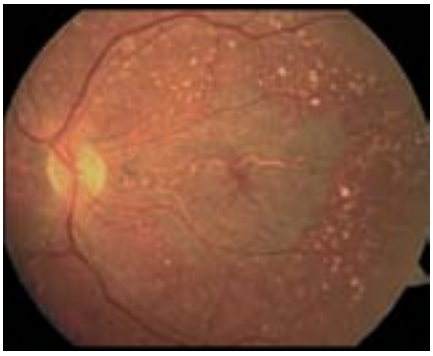


Fig. 12.40.2B: Dry AMD—with hard drusen—LE

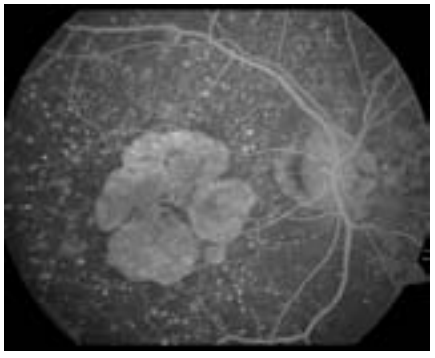


Fig. 12.40.2C: Dry AMD—geographical atrophy on FFA—RE

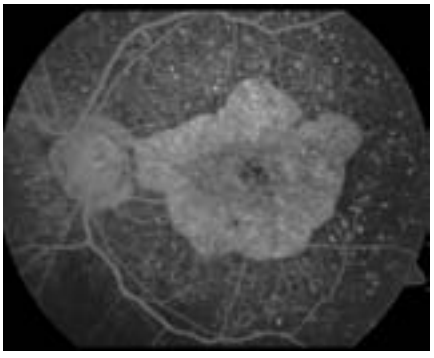


Fig. 12.40.2D: Dry AMD—geographical atrophy on FFA—LE

Exudative Maculopathies

Cystoid Macular Edema (CME)

- Cystoid macular edema (CME) is an accumulation of fluid in the outer plexiform (Henle's) layer and inner nuclear layer of the retina, in the macular region
- Shows an irregularity and blurring of the foveal reflex **(Fig. 12.41.1A)**
- Macula is wrinkled, edematous, and may show multiple small cystic changes
- FFA shows typical 'flower-petal' or 'spoke' pattern of leakage at the fovea during the late phase **(Figs 12.41.1B and C)**
- In long standing cases (chronic CME) **(Figs 12.41.2A and B)**, there is formation of epiretinal membrane **(Fig. 12.41.3)** and eventually macular hole formation
- *Causes:* Cataract surgery (Irvine-Gass syndrome), chronic iridocyclitis, intermediate uveitis, YAG capsulotomy, retinitis pigmentosa, diabetes, etc.

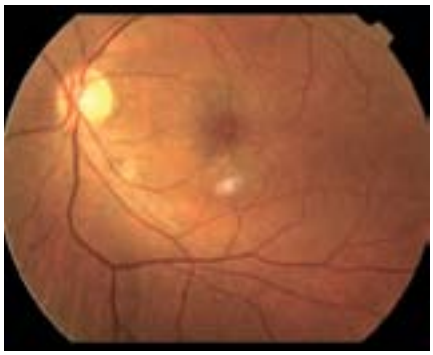


Fig. 12.41.1A: Cystoid macular edema

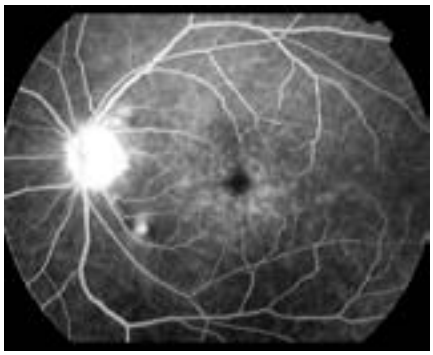


Fig. 12.41.1B: Cystoid macular edema—flower-petal appearance on FFA

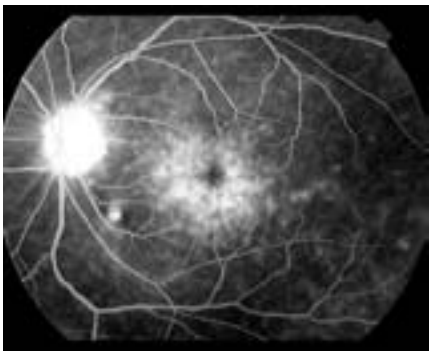


Fig. 12.41.1C: Cystoid macular edema—flower-petal appearance on FFA

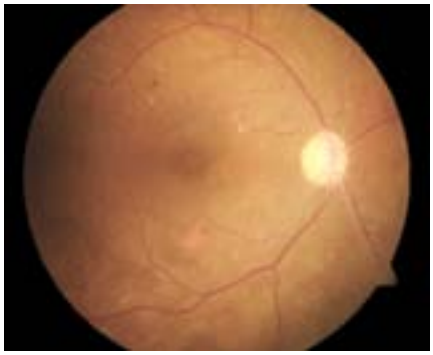


Fig. 12.41.2A: Cystoid macular edema—chronic

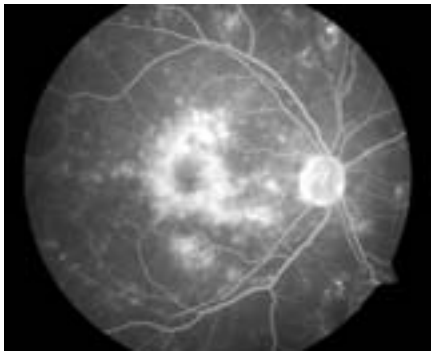


Fig. 12.41.2B: Cystoid macular edema—chronic

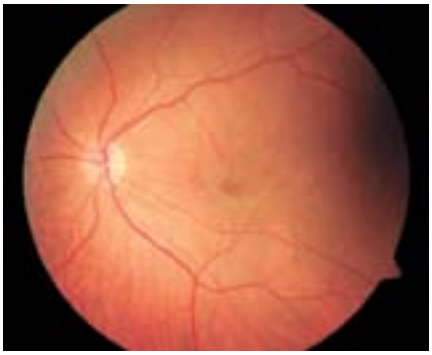


Fig. 12.41.3: Chronic CME with epiretinal membrane and lamellar hole

Central Serous Retinopathy (CSR)

- CSR is actually the detachment of the neurosensory retina from the RPE by serous fluid
- Characterized by sudden shallow elevation of the neuroretina in the macular area with indistinct margin
- Macula appears as an oval or circular dark swelling, about the size of the optic disk, often with glistening ring-reflex **(Fig. 12.42.1)**

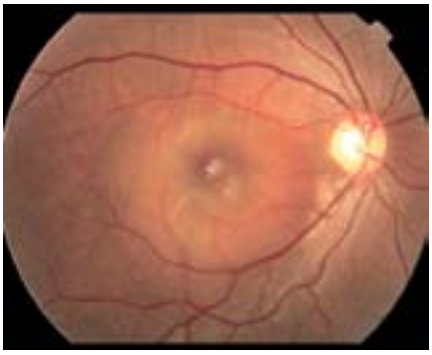


Fig. 12.42.1: Central serous retinopathy

- Sometimes, tiny subretinal precipitates may be visible **(Figs 12.42.3A and 12.42.4A)**
- FFA shows characteristic 'smoke-stack'—pattern leak **(Figs 12.42.2A to D)** or sometimes gradual 'ink-blot'—pattern leak **(Figs 12.42.3A to D)** under the neurosensory retina
- Occasionally, there may be atypical and extrafoveal CSR **(Figs 12.42.4A to C)** or CSR with multiple leak **(Figs 12.42.5A to C)**

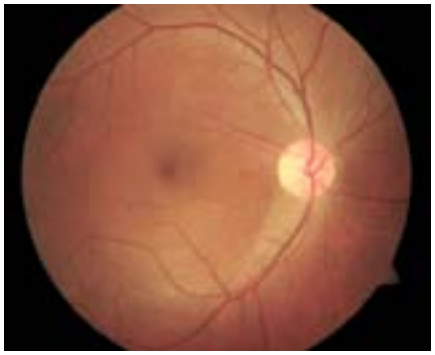


Fig. 12.42.2A: Central serous retinopathy

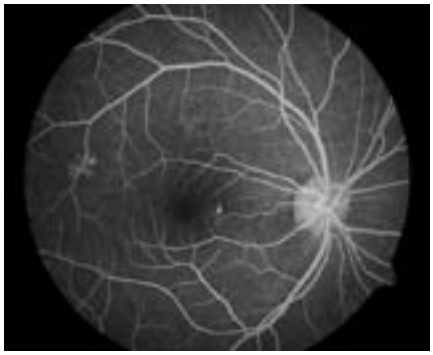


Fig. 12.42.2B: Central serous retinopathy

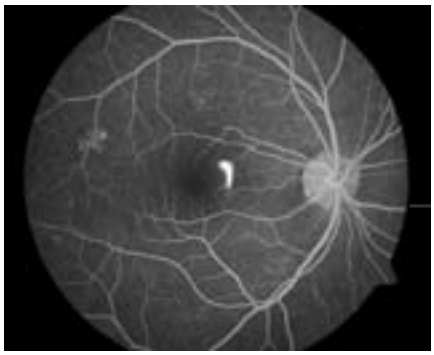


Fig. 12.42.2C: Central serous retinopathy

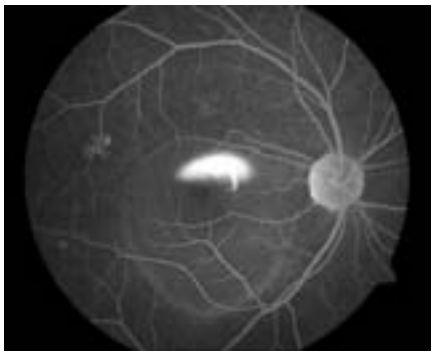


Fig. 12.42.2D: Central serous retinopathy

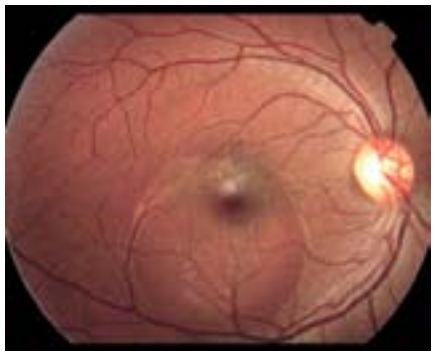


Fig. 12.42.3A: Central serous retinopathy

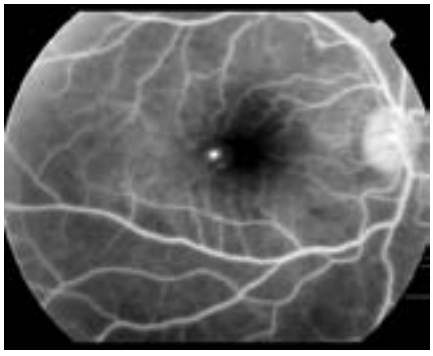


Fig. 12.42.3B: CSR—ink-blot pattern on FFA

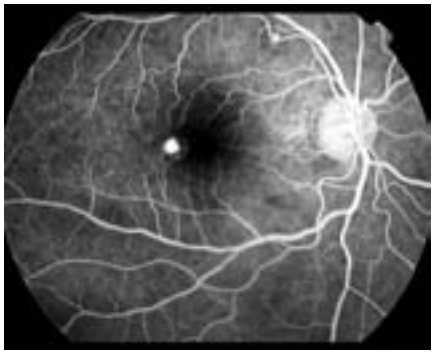


Fig. 12.42.3C: CSR—ink-blot pattern on FFA

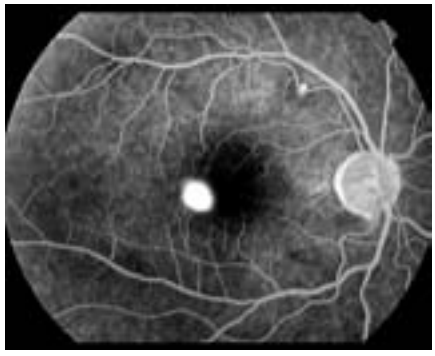


Fig. 12.42.3D: CSR—ink-blot pattern on FFA

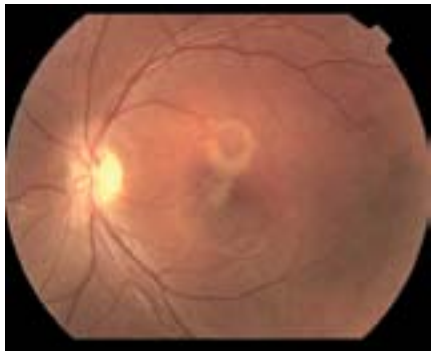


Fig. 12.42.4A: CSR—extrafoveal

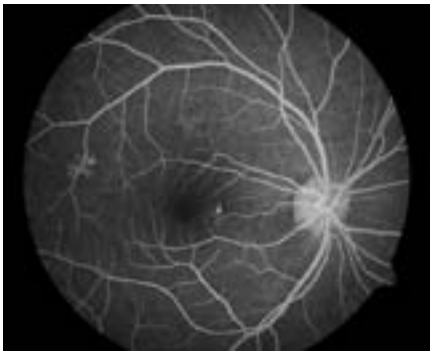


Fig. 12.42.4B: CSR—extrafoveal

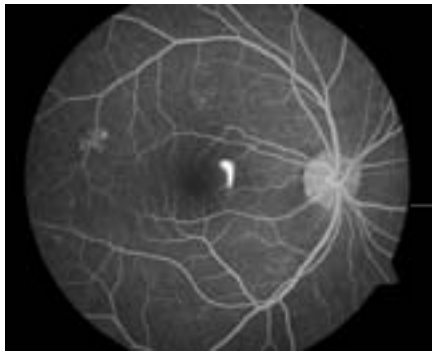


Fig. 12.42.4C: CSR—extrafoveal

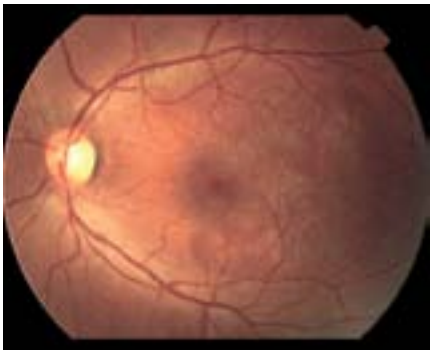


Fig. 12.42.5A: CSR—extrafoveal with multiple leaks

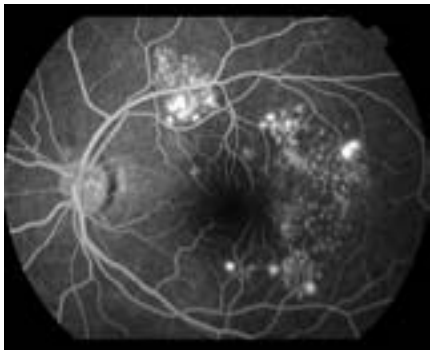


Fig. 12.42.5B: CSR—extrafoveal with multiple leaks on FFA

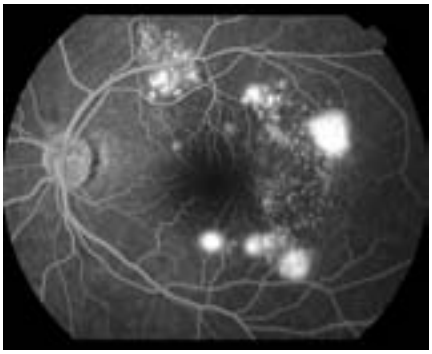


Fig. 12.42.5C: CSR—extrafoveal with multiple leaks on FFA

- Chronic CSR may leaks profusely in a different way **(Figs 12.42.6A and B)**
- Resolving CSR may have sharp margin with more tiny crystals as precipitates **(Figs 12.42.7A and B)**.

RPE Detachment

- Separation of retinal pigment epithelium (RPE) from the Bruch's membrane
- Unilateral or bilateral sharply demarcated dome shaped lesions of varying size at the posterior pole **(Figs 12.43.1A and B)**

Fig. 12.42.6A: Chronic CSR

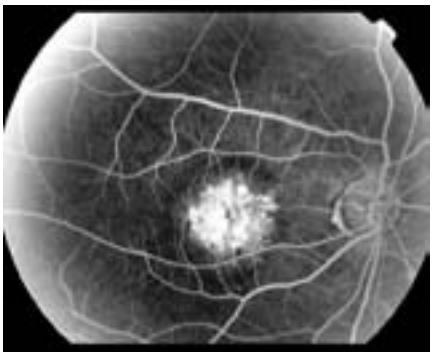


Fig. 12.42.6B: Chronic CSR on FFA

Fig. 12.42.7A: Resolving CSR with tiny precipitates—RE

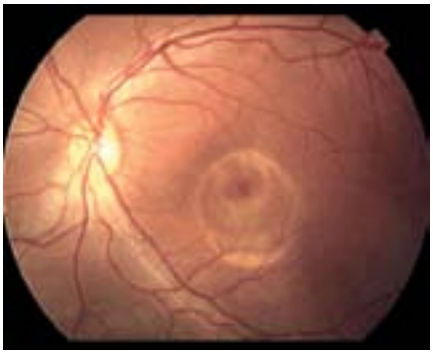


Fig. 12.42.7B: Resolving CSR with tiny precipitates—LE

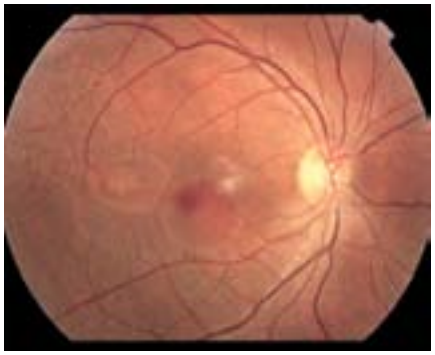


Fig. 12.43.1A: RPE detachment—bilateral—RE

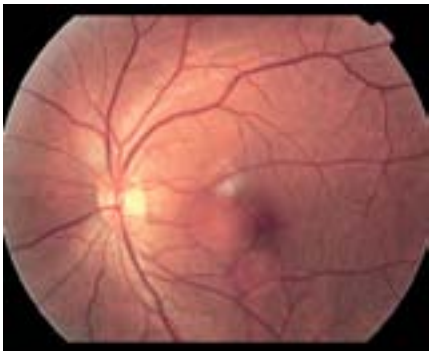


Fig. 12.43.1B: RPE detachment—LE

- FFA shows area of hyperfluorescence which increases in intensity but not in size in late phase **(Figs 12.43.1C to F)**
- PED may coexist with CSR **(Figs 12.43.2A to C)**
- There may be hemorrhage in the detached area—called hemorrhagic PED **(Fig. 12.43.3)**

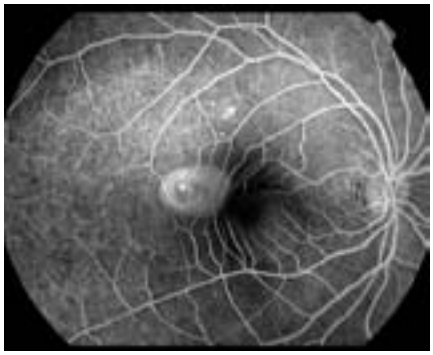


Fig. 12.43.1C: PED—FFA finding—RE

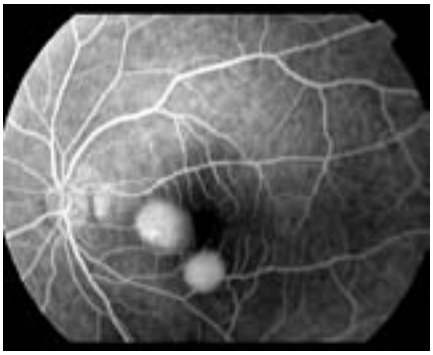


Fig. 12.43.1D: PED on FFA—LE

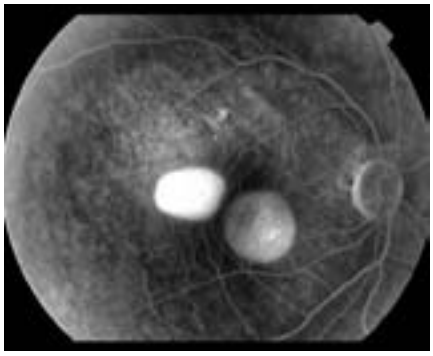


Fig. 12.43.1E: PED—FFA finding—RE

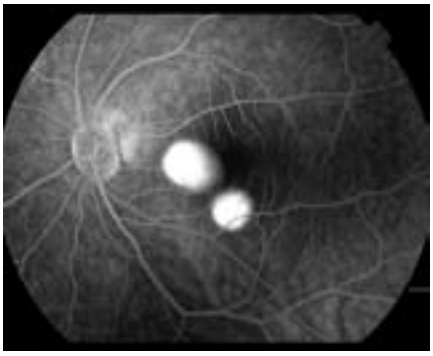


Fig. 12.43.1F: PED on FFA—LE

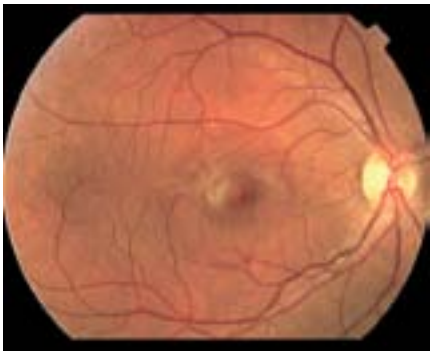


Fig. 12.43.2A: PED with CSR

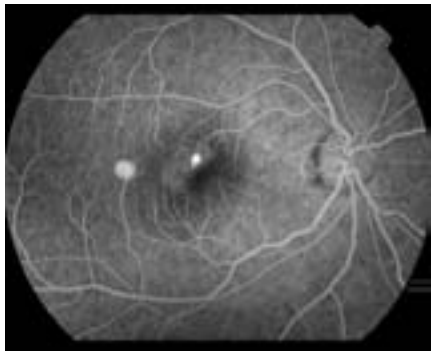


Fig. 12.43.2B: PED with CSR

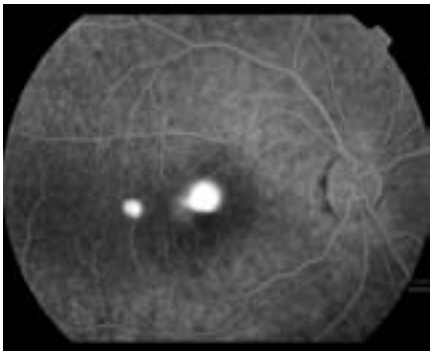


Fig. 12.43.2C: PED with CSR

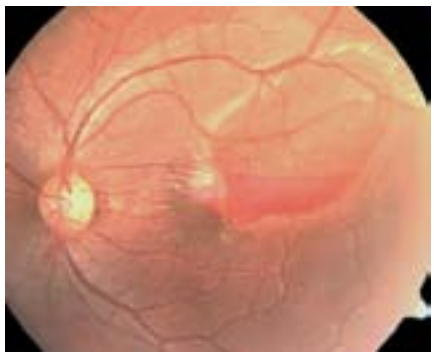


Fig. 12.43.3: Hemorrhagic PED

Wet Age-Related Macular Degeneration

- Wet AMD is a common bilateral disease affecting the elderly people and one of the important cause of blindness
- Characterized by formation of subretinal neovascular membranes (SRNVMs) at the macula
- SRNVMs consists of proliferations of fibrovascular tissue from the choriocapillaris
- Clinically appears as pinkish-yellow slightly elevated subretinal lesion of variable size **(Fig. 12.44.1)** and often with bleeding **(Figs 12.44.2 and 12.44.3)**
- FFA of SRNVMs shows characteristic lacy pattern during the early phase, followed by hyperfluorescence and late leakage **(Figs 12.44.4 and 12.44.5)**

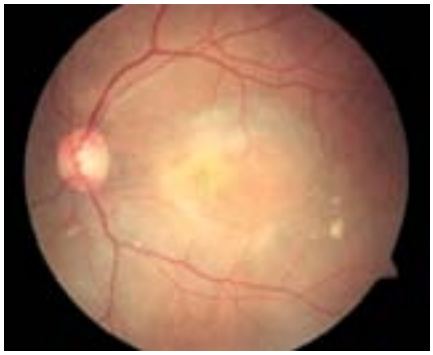


Fig. 12.44.1: Wet AMD—classical lesion

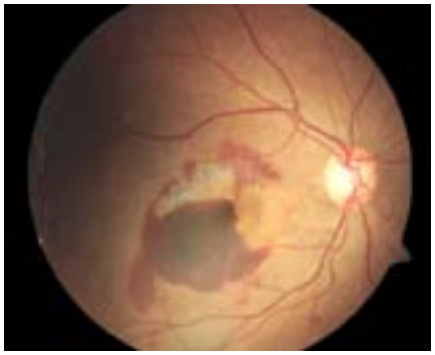


Fig. 12.44.2: Wet AMD—bleeding SRNVMs

Fig. 12.44.3A: Wet AMD—bleeding SRNVMs

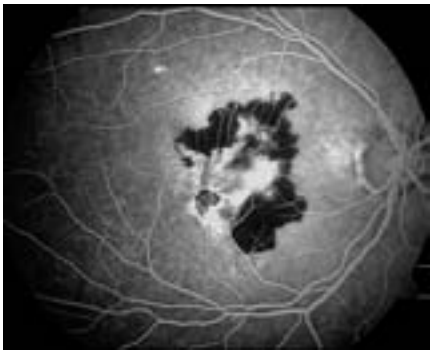


Fig. 12.44.3B: Wet AMD—bleeding SRNVMs on FFA

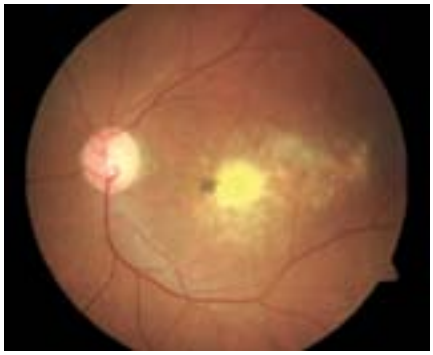


Fig. 12.44.4A: Wet AMD—classical SRNVMs

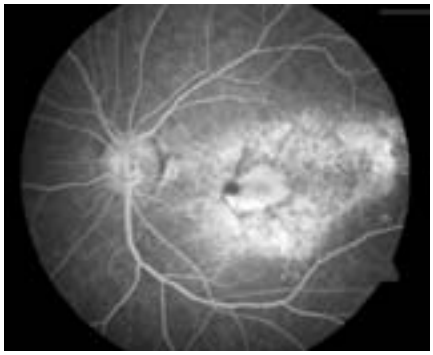


Fig. 12.44.4B: Wet AMD—classical SRNVMs on FFA

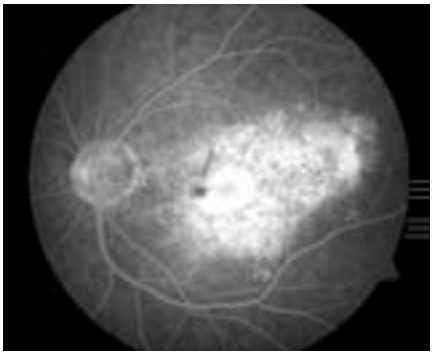


Fig. 12.44.4C: Wet AMD—classical SRNVMs on FFA

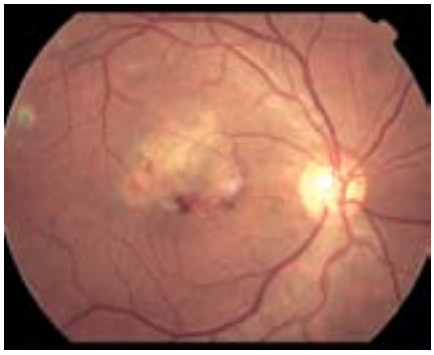


Fig. 12.44.5A: Wet AMD—SRNVMs

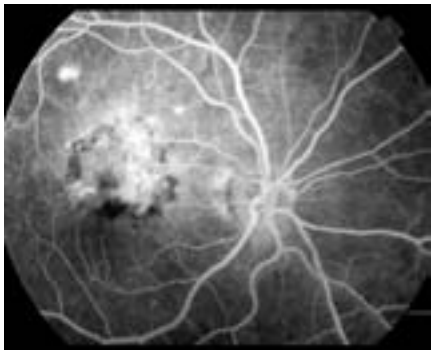


Fig. 12.44.5B: Wet AMD—SRNVMs on FFA

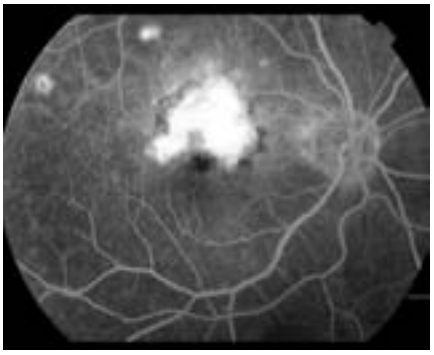


Fig. 12.44.5C: Wet AMD—SRNVMs on FFA

- Extensive subretinal macular fibrosis occurs in end stage with disciform scarring of macula **(Figs 12.44.6A and B)**.

Macular Holes

- May be full thickness or lamellar **(Figs 12.45.1 and 12.45.2)**
- *Idiopathic type:*
  - A common age-related condition with bilateral affection in 15% cases

Fig. 12.44.6A: Wet AMD—scarring and disciform change

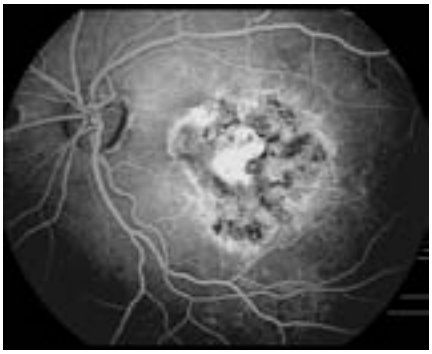


Fig. 12.44.6B: Wet AMD—scarring and disciform change

Fig. 12.45.1A: Macular hole

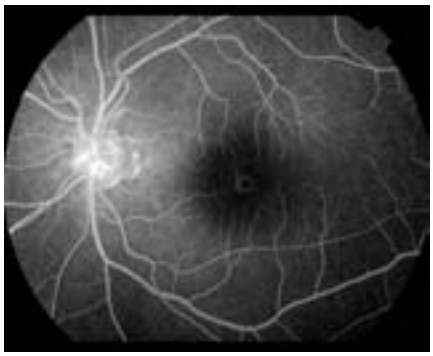


Fig. 12.45.1B: Macular hole

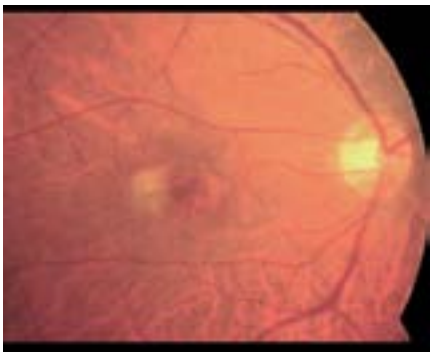


Fig. 12.45.2: Macular hole—lamellar

  - *Stage I:* Yellow foveolar spot with loss of depression
  - *Stage II:* Central round foveal defect with an elevated retinal rim **(Fig. 12.45.3)**
  - *Stage III:* Central round foveal defect with a central smaller operculum **(Fig. 12.45.4)**
  - *Stage IV:* Central round foveal defect with complete separation of operculum **(Figs 12.45.5A and B)**

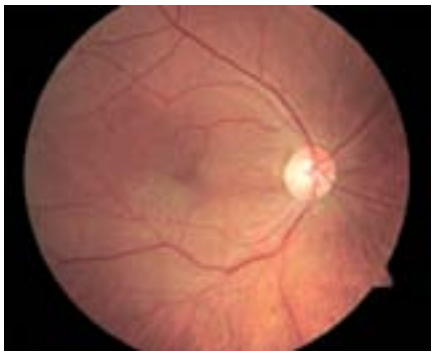


Fig. 12.45.3: Macular hole—stage II

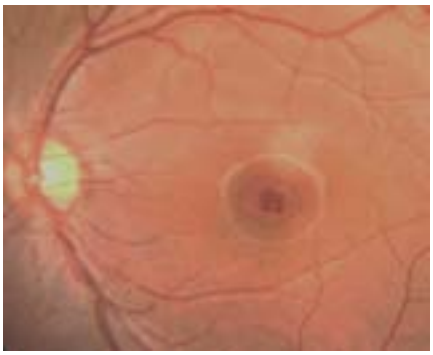


Fig. 12.45.4: Macular hole—stage III

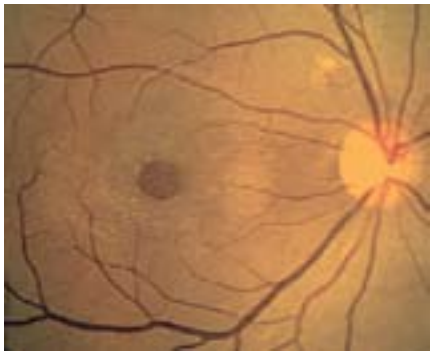


Fig. 12.45.5A: Macular hole—stage IV

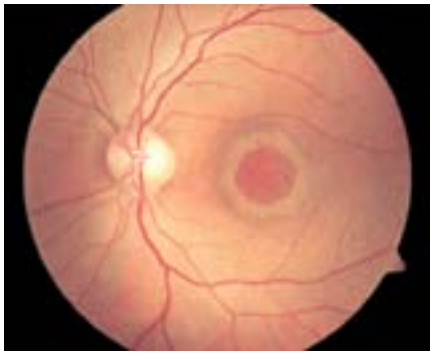


Fig. 12.45.5B: Macular hole—stage IV

  - Multiple yellow deposits at the level of RPE within the hole **(Fig. 12.45.6)**
  - FFA shows a corresponding circular zone of hypofluorescence **(Figs 12.45.7A and B)**
- *Other types:*
  - Macular hole in myopia
  - Traumatic macular hole **(*see* Fig. 16.10.3)**
  - Lamellar hole after prolonged CME
- *Pseudohole:* It is the discontinuity of an epiretinal membrane over the macula **(Figs 12.45.8A and B)**.

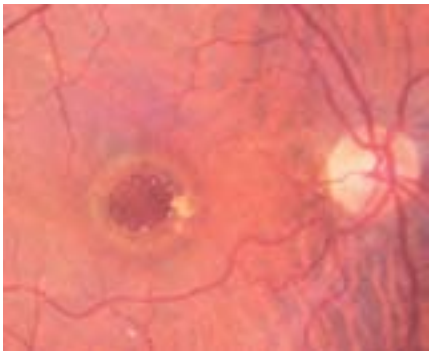


Fig. 12.45.6: Macular hole—yellow deposits

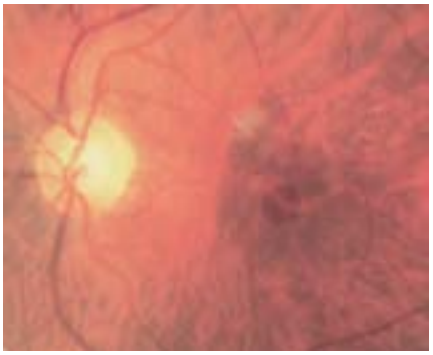


Fig. 12.45.7A: Lamellar macular hole

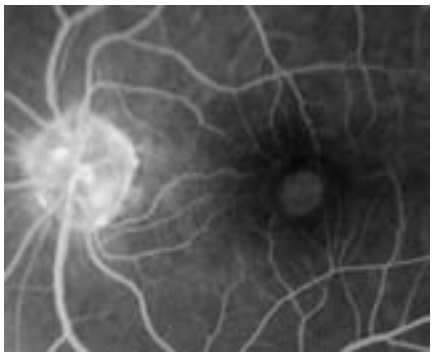


Fig. 12.45.7B: Lamellar macular hole on FFA

Fig. 12.45.8A: Epiretinal membrane—with macular pseudohole

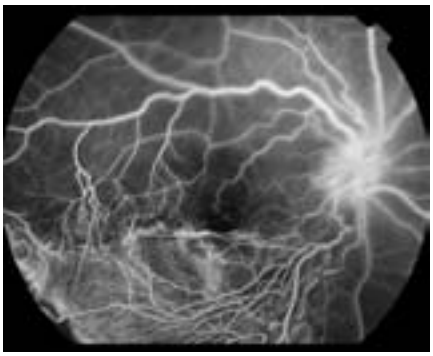


Fig. 12.45.8B: Epiretinal membrane—with macular pseudohole on FFA

Other Maculopathies

Cellophane Maculopathy

- Preretinal macular fibrosis caused by contraction of epiretinal membrane over the macula
- Appears as a transparent membrane with fine retinal striation **(Fig. 12.46.1)**
- Associated vascular tortuosity is best appreciated by FFA **(*see* Figs 12.45.8 and 12.46.2)**

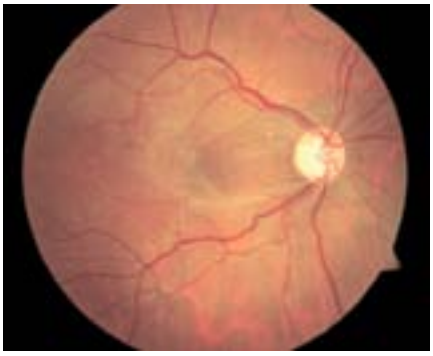


Fig. 12.46.1: Cellophane maculopathy—epiretinal membrane

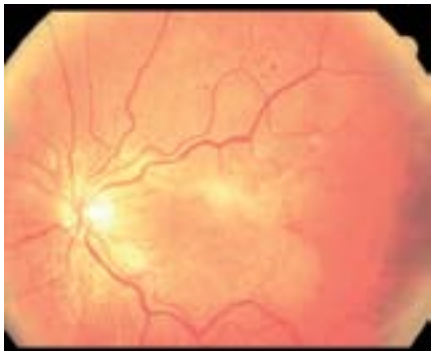


Fig. 12.46.2A: Cellophane maculopathy—epiretinal membrane

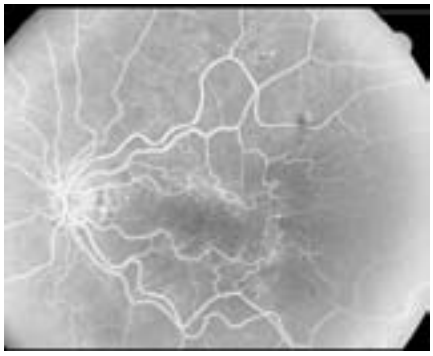


Fig. 12.46.2B: Epiretinal membrane on FFA

- Later the membrane becomes more opaque with more wrinkling **(Figs 12.46.3 and 14.46.4)**.

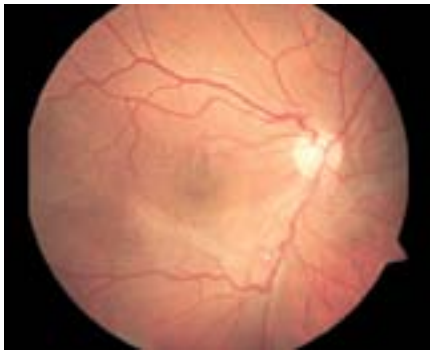


Fig. 12.46.3: Epiretinal membrane—dense

Fig. 12.46.4: Epiretinal membrane—more dense

Commotio Retinae (Berlin's Edema)

- Localized macular edema, often with cherry red spot **(Fig. 12.47.1)**
- Caused by blunt trauma
- May be associated with disk hemorrhage or retinal hemorrhage **(Fig. 12.47.2)**.

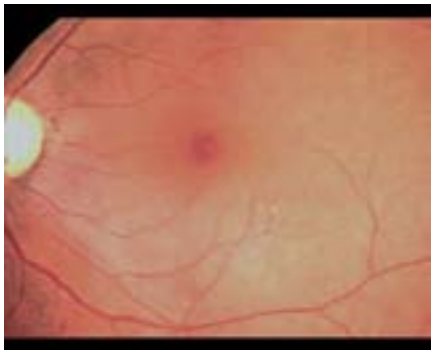


Fig. 12.47.1: Commotio retinae—Berlin's edema

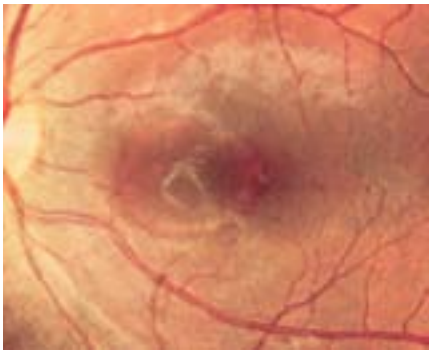


Fig. 12.47.2: Commotio retinae—Berlin's edema

Crystalline Maculopathy

- Multiple tiny glistening crystals at the posterior pole and parafoveal region **(Figs 12.48.1A and B)**
- May be drug-induced like, tamoxifen, canthaxanthin, or talc **(Figs 12.48.2A and B)**
- May also seen in metabolic disorder like, cystinosis.

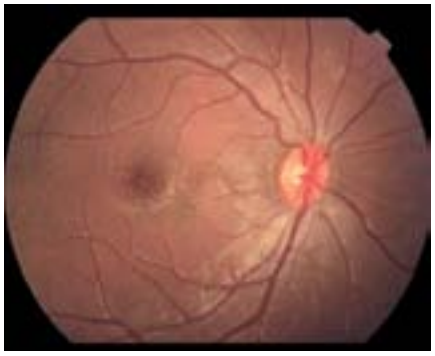


Fig. 12.48.1A: Crystalline maculopathy—RE

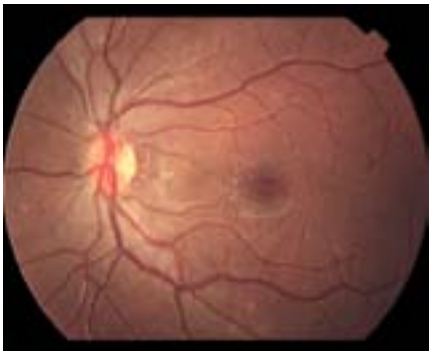


Fig. 12.48.1B: Crystalline maculopathy—LE

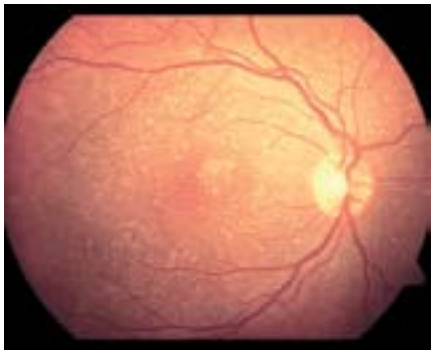


Fig. 12.48.2A: Crystalline maculopathy—tamoxifen induced—RE

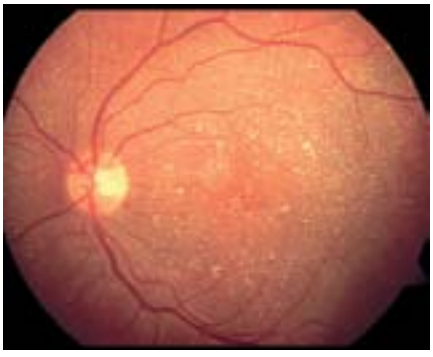


Fig. 12.48.2B: Crystalline maculopathy—tamoxifen induced—LE

Multiple Flecked Retinal Lesions

Familial Dominant Drusen (Doyne's Honeycomb Dystrophy)

- Rare condition with dominant inheritance, affects the younger individuals
- Relatively large, discrete nodular drusen mainly affects the area either temporal to the fovea or nasal to the disk **(Figs 12.49.1A and B)**
- Often in a symmetrical distribution
- With time they merge together and become confluent with an appearance of honeycomb
- In late stage pigmentary changes occur with loss of central vision.

Fundus Flavimaculatus

- Rare bilateral condition with recessive inheritance
- Yellowish-white fish-tail like flecks at the posterior pole and midperipheral retina **(Figs 12.50.1A to D)**
- Flecks may occur in isolation or with maculopathy, as in Stargardt's disease **(Figs 12.50.2A and B)**

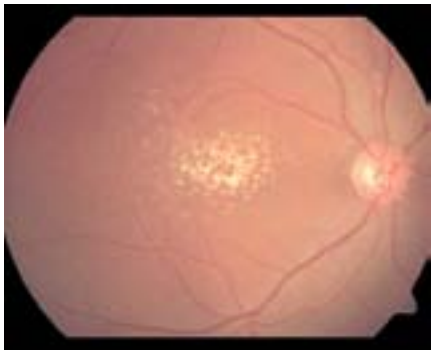


Fig. 12.49.1A: Familial drusen—RE

Fig. 12.49.1B: Familial drusen—LE

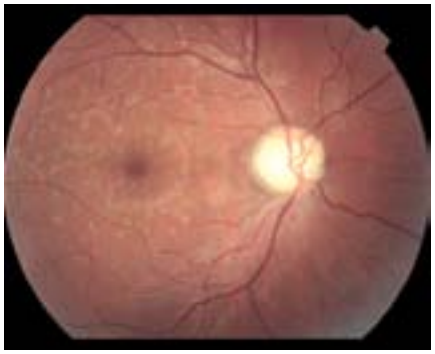


Fig. 12.50.1A: Fundus flavimaculatus—RE

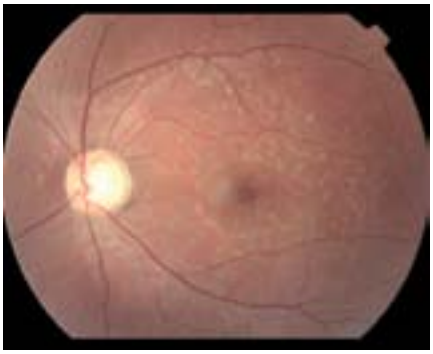


Fig. 12.50.1B: Fundus flavimaculatus—LE

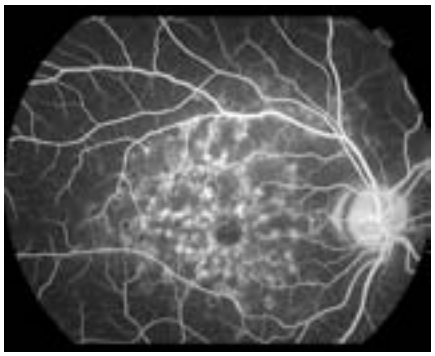


Fig. 12.50.1C: Fundus flavimaculatus—RE

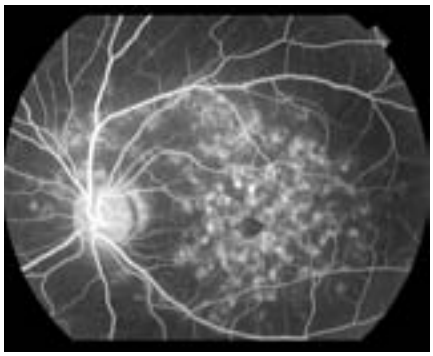


Fig. 12.50.1D: Fundus flavimaculatus—LE

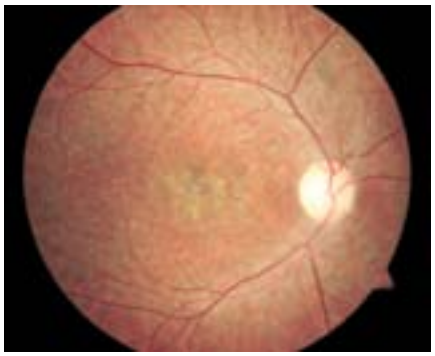


Fig. 12.50.2A: Fundus flavimaculatus—RE

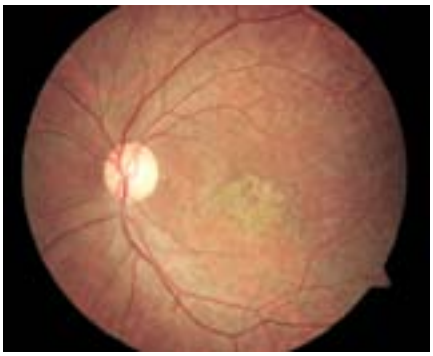


Fig. 12.50.2B: Fundus flavimaculatus—LE

- FFA shows hyperfluorescence due to atrophic changes **(Figs 12.50.2C and D)**.

Fundus Albipunctatus

- Rare, bilateral recessively inherited condition with congenital stationary night blindness
- Multiple tiny yellowish-white dots from posterior pole to the periphery **(Figs 12.51.1A and B)**
- Macula is spared and visual acuity remains normal **(Figs 12.51.2A and B)**
- Prognosis is relatively better.

Benign Flecked Retina Syndrome

- Very rare, bilateral condition
- Multiple tiny yellowish-white flecks all over the fundus **(Figs 12.52.1A and B)**
- Similar to fundus flavimaculatus, but the macula is always spared **(Figs 12.52.1C and D)**
- Prognosis is excellent with normal vision.

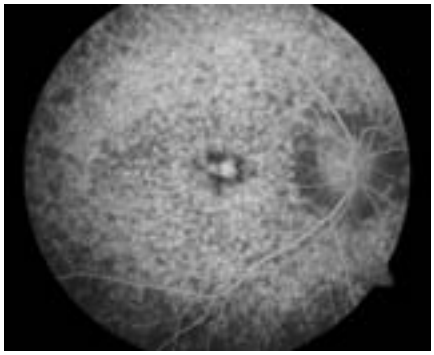


Fig. 12.50.2C: Fundus flavimaculatus—RE on FFA

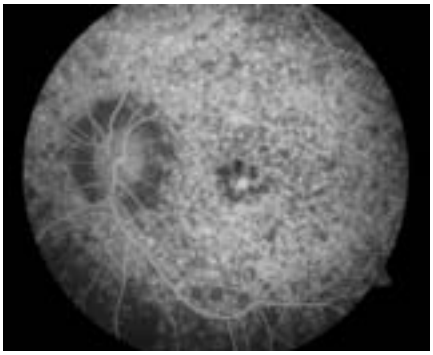


Fig. 12.50.2D: Fundus flavimaculatus—LE on FFA

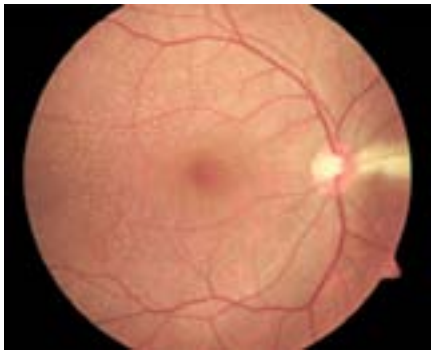


Fig. 12.51.1A: Fundus albipunctatus—congenital stationary night blindness (CSNB)—RE

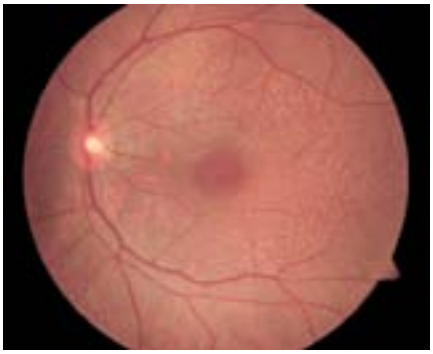


Fig. 12.51.1B: Fundus albipunctatus—congenital stationary night blindness (CSNB)—LE

Fig. 12.51.2A: Fundus albipunctatus—RE

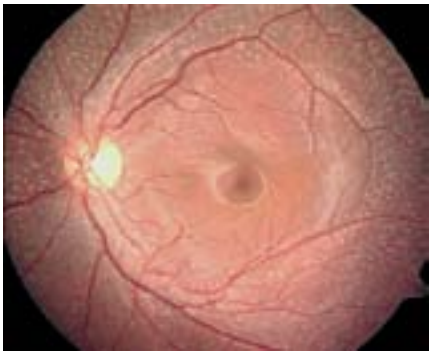


Fig. 12.51.2B: Fundus albipunctatus—LE

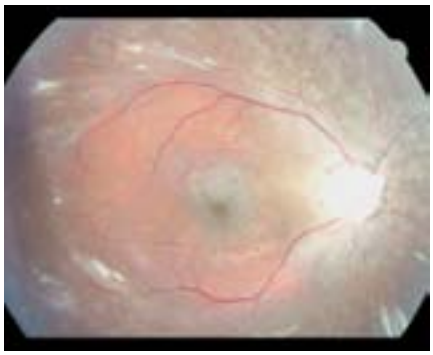


Fig. 12.52.1A: Benign flecked retina syndrome—RE

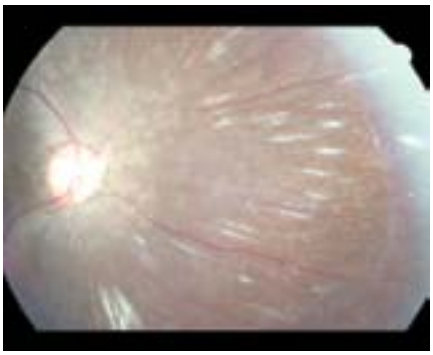


Fig. 12.52.1B: Benign flecked retina syndrome—LE

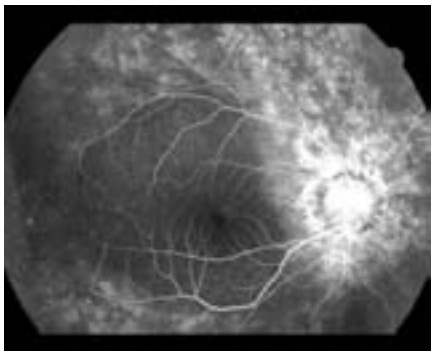


Fig. 12.52.1C: Benign flecked retina syndrome—macula spared—RE

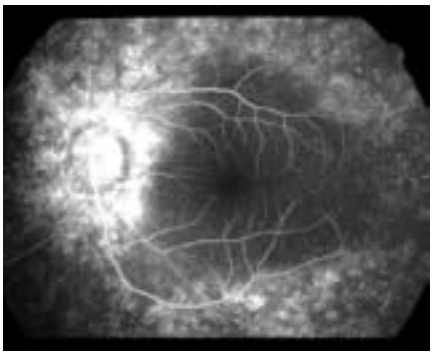


Fig. 12.52.1D: Benign flecked retina syndrome—macula spared—LE

Pattern Dystrophy

- Very rare, bilateral condition with dominant inheritance
- Slowly progressive symmetrical lesions with various geometrical configurations **(Figs 12.53.1A to D)**
- FFA shows distinct symmetrical different pattern of the posterior pole **(Figs 12.53.2A to D)**
- Prognosis is relatively good.

Fig. 12.53.1A: Pattern macular dystrophy—RE

Fig. 12.53.1B: Pattern macular dystrophy—LE

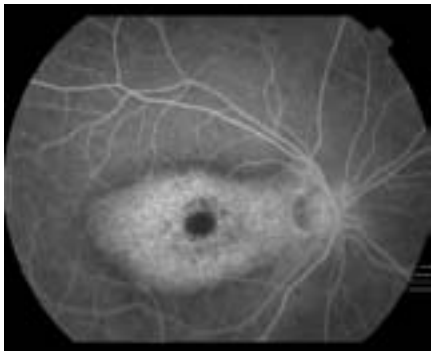


Fig. 12.53.1C: Pattern macular dystrophy—RE

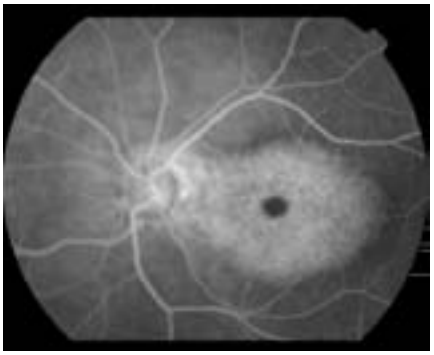


Fig. 12.53.1D: Pattern macular dystrophy—LE

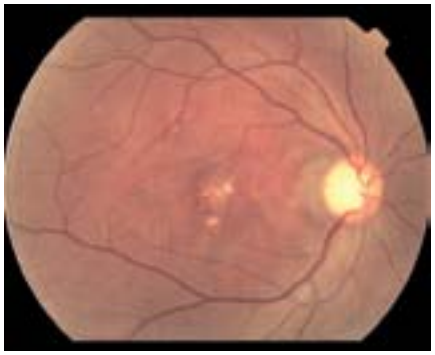


Fig. 12.53.2A: Pattern macular dystrophy—RE

Fig. 12.53.2B: Pattern macular dystrophy—LE

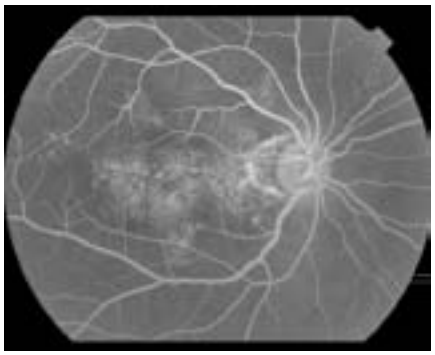


Fig. 12.53.2C: Pattern macular dystrophy—RE

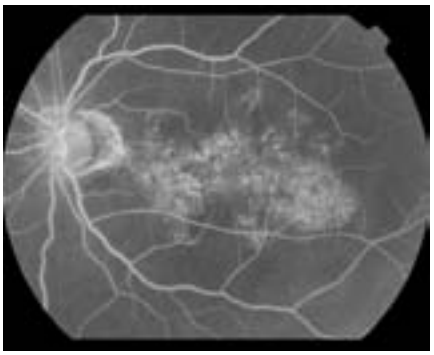


Fig. 12.53.2D: Pattern macular dystrophy—LE

Hard Drusens

- Very common age-related condition
- Usually not associated with subsequent development of macular degeneration
- Multiple, bilaterally symmetrical, small, discrete, yellow-white, slightly elevated lesions at the posterior poles of both fundi **(Figs 12.54.1 and 12.54.2)**
- *Calcified drusen:* Secondary calcification in long standing cases gives them glistening-white appearance with conspicuous margin **(Figs 12.54.3A to D)**

Fig. 12.54.1: Central hard drusen

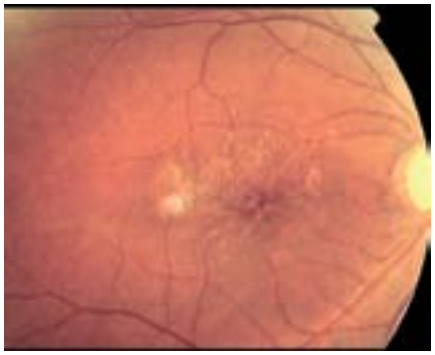


Fig. 12.54.2A: Hard drusen—RE

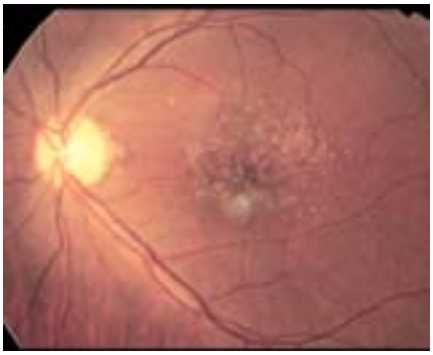


Fig. 12.54.2B: Hard drusen—LE

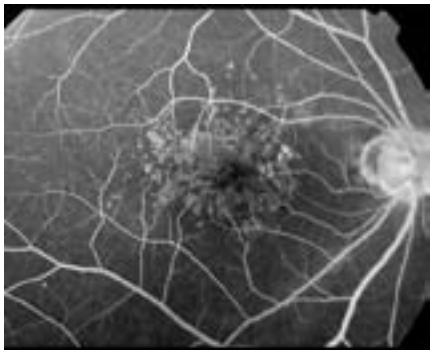


Fig. 12.54.2C: Hard drusen on FFA—RE

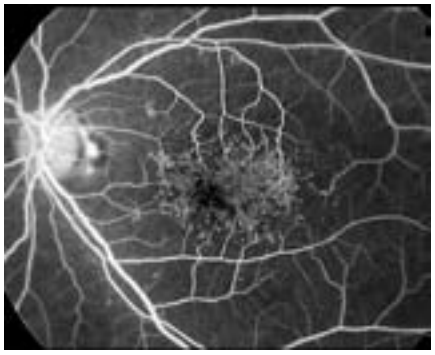


Fig. 12.54.2D: Hard drusen on FFA—LE

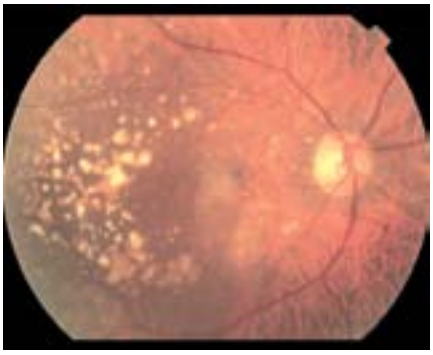


Fig. 12.54.3A: Calcified drusen—RE

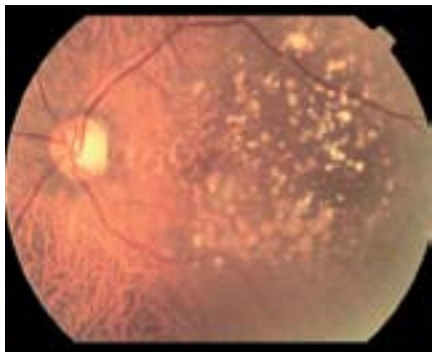


Fig. 12.54.3B: Calcified drusen—LE

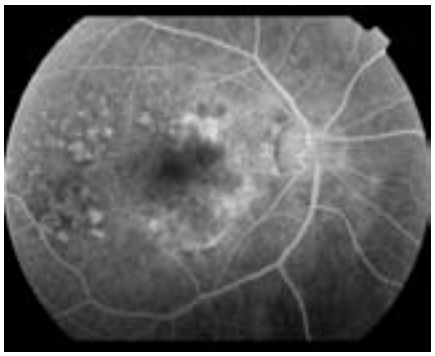


Fig. 12.54.3C: Calcified drusen on FFA—RE

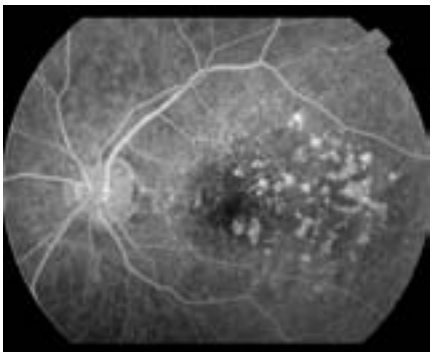


Fig. 12.54.3D: Calcified drusen on FFA—LE

- Hard and calcified drusens may exist together **(Figs 12.54.4 and 12.54.5)**
- FFA shows multiple RPE-window defects with hyperfluorescence spots, and late staining of the drusen.

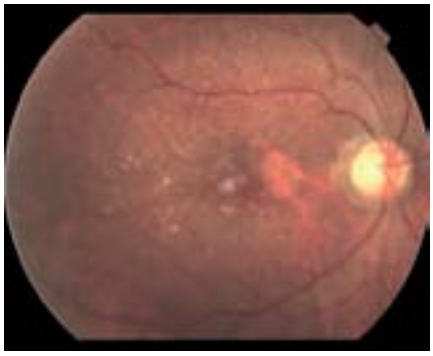


Fig. 12.54.4A: Hard and calcified dursen—RE

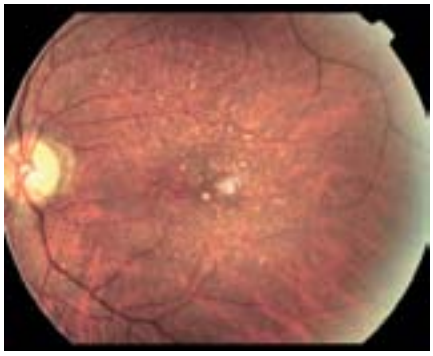


Fig. 12.54.4B: Hard and calcified dursen—LE

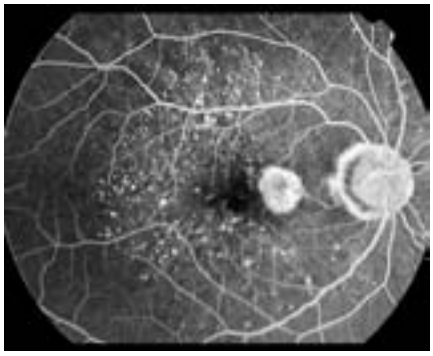


Fig. 12.54.4C: Hard and calcified dursen on FFA—RE

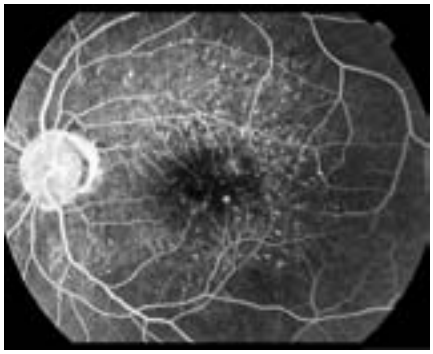


Fig. 12.54.4D: Hard and calcified dursen on FFA—LE

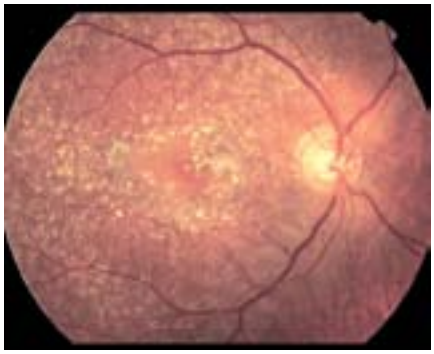


Fig. 12.54.5A: Hard and calcified drusen—RE

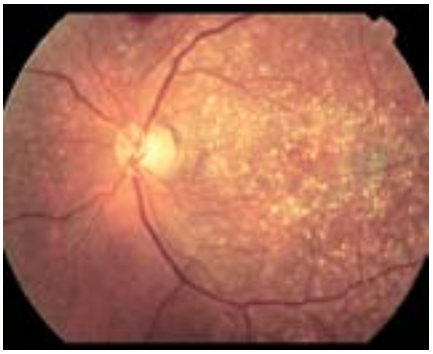


Fig. 12.54.5B: Hard and calcified drusen—LE

Soft Drusens

- Common age-related lesions associated with increased risk of wet ARMD
- Are larger than hard drusen with indistinct edges **(Figs 12.55.1A to C)**

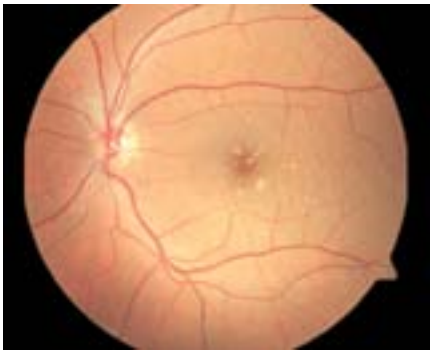


Fig. 12.55.1A: Soft drusen—very few

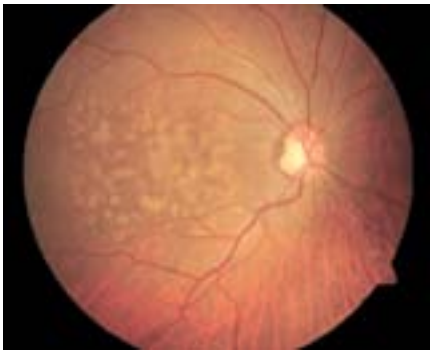


Fig. 12.55.1B: Soft drusen—RE

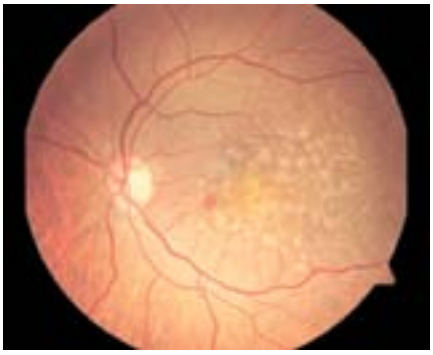


Fig. 12.55.1C: Soft drusen—LE

- Become confluent with time **(Figs 12.55.2A and B)**
- Lesions are often asymmetrical and secondary RPE changes are quite common
- FFA shows hyperfluorescence **(Figs 12.55.2C and D)**
- Sometimes, they may be found along with hard drusen.

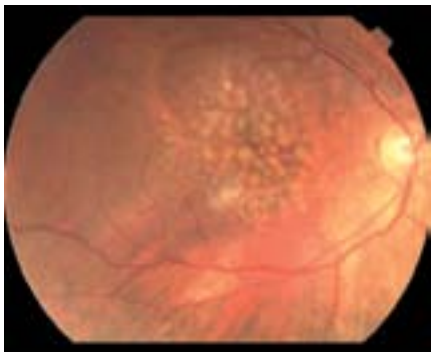


Fig. 12.55.2A: Soft drusen—confluent—RE

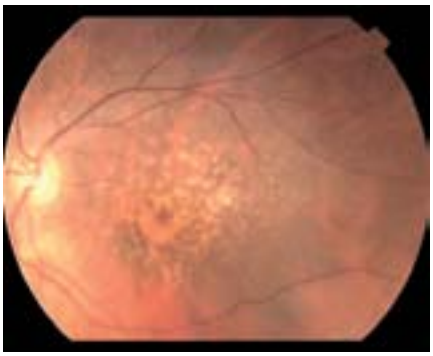


Fig. 12.55.2B: Soft drusen—confluent—LE

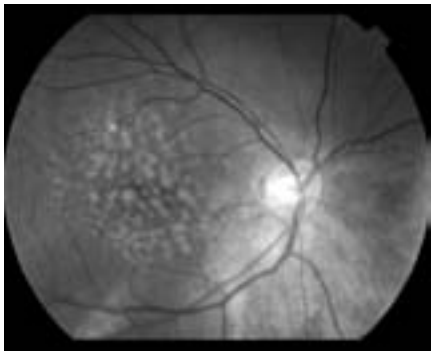


Fig. 12.55.2C: Soft drusen on FFA—RE

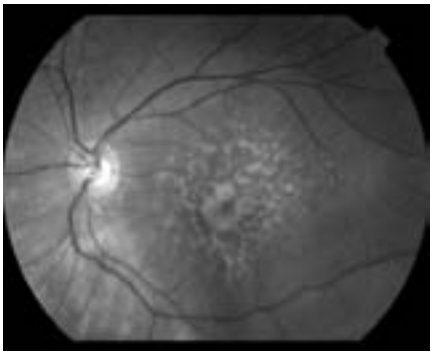


Fig. 12.55.2D: Soft drusen on FFA—LE

Cuticular (Basal-Laminar) Drusens

- Rare kind of drusens affects the middle aged person
- Bilaterally symmetrical, innumerable, small uniform, discrete round slightly elevated yellowish subretinal lesions all over the posterior pole **(Figs 12.56.1A and B)**
- 'Starry night' or 'Milky way' appearance of the fundus which can be appreciated better on FFA
- In late stage, there are pigmentary changes in the macula **(Figs 12.56.2A and B)**.

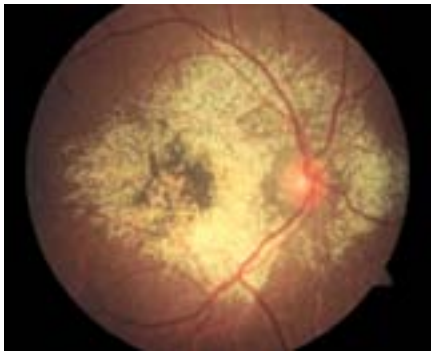


Fig. 12.56.1A: Cuticular drusen

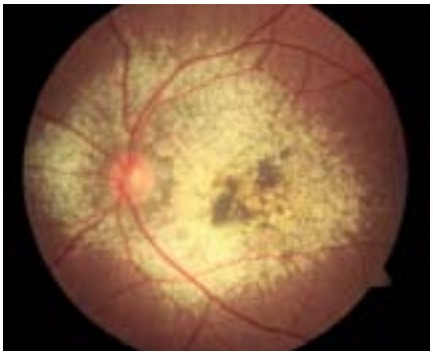


Fig. 12.56.1B: Cuticular drusen

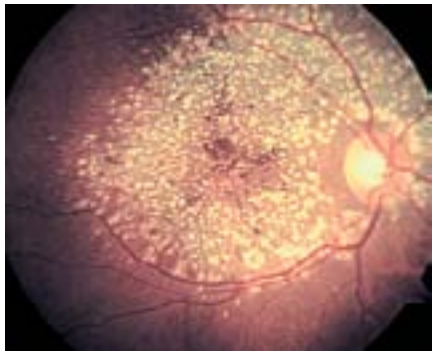


Fig. 12.56.2A: Cuticular drusen—macular pigments—RE

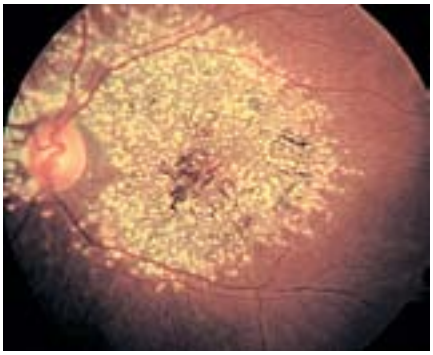


Fig. 12.56.2B: Cuticular drusen—macular pigments—LE

Pigmentary Retinopathy

Typical Retinitis Pigmentosa

- Typical RP is a bilateral, symmetrical, progressive diffuse pigmentary retinal dystrophy which predominantly affecting the rods **(Figs 12.57.1A and B)**
- Patients usually present in the second decade of life and ultimately have severe visual loss in later life
- *Classical triad of RP:*
  - Bony-spicule pigmentation **(Fig. 12.57.2)**
  - Arteriolar attenuation
  - Waxy-pallor of the optic disk
- Pigmentary changes are typically perivascular, and have a bone-spicule appearance, which are observed mostly at the equatorial region of the retina **(Fig. 12.57.3)**
- In late stages of the disease, the unmasking of the larger choroidal vessels gives the fundus a tessellated appearance **(Figs 12.57.4 and 12.57.5)**
- Cystoid *macular edema* **(Fig. 12.57.6)**, *atrophic* or *cellophane maculopathy* may be present up to 70% of the patients with RP **(Figs 12.57.7 and 12.57.8)**.

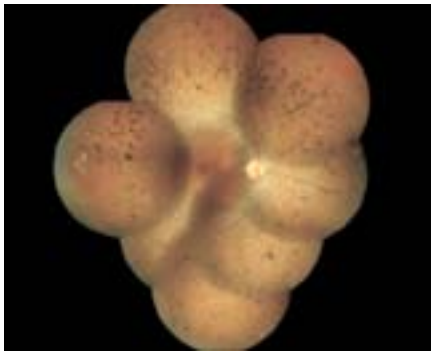


Fig. 12.57.1A: Retinitis pigmentosa—RE

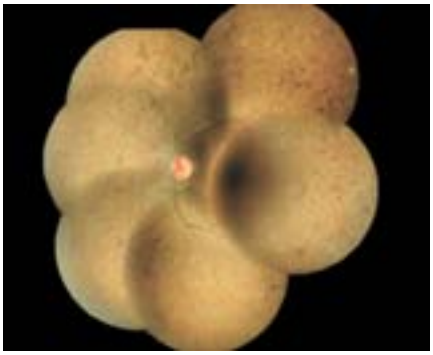


Fig. 12.57.1B: Retinitis pigmentosa—LE

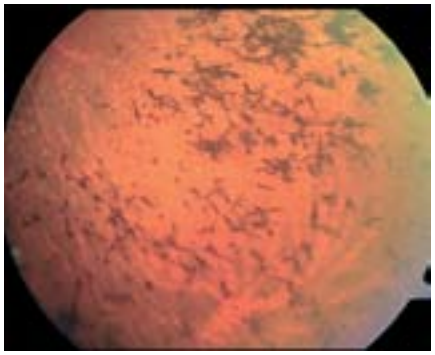


Fig. 12.57.2: Retinitis pigmentosa—typical bony spicules pigmentation

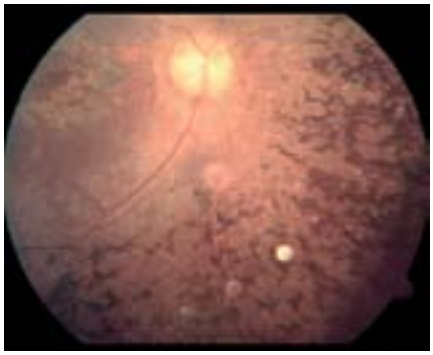


Fig. 12.57.3: Retinitis pigmentosa—periequatorial distribution

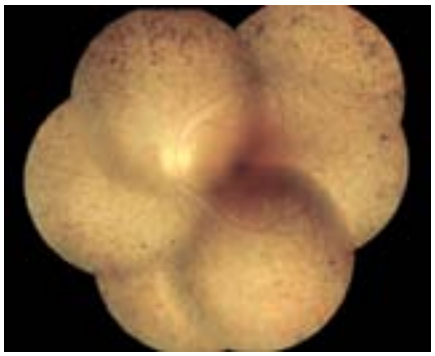


Fig. 12.57.4: Retinitis pigmentosa—tessellated fundus with waxy pallor of the disk

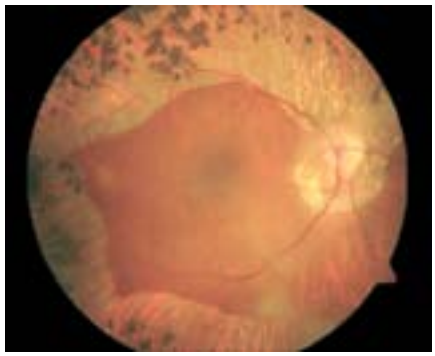


Fig. 12.57.5: Retinitis pigmentosa—tessellated fundus

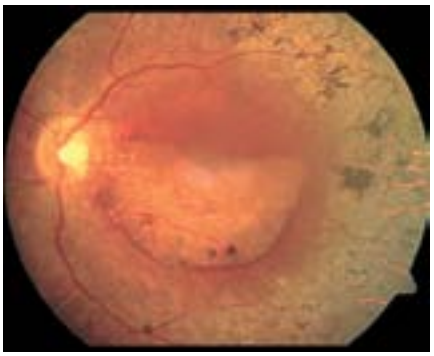


Fig. 12.57.6: Retinitis pigmentosa—CME

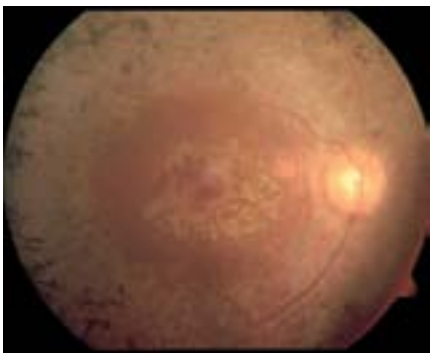


Fig. 12.57.7A: Retinitis pigmentosa—atrophic maculopathy

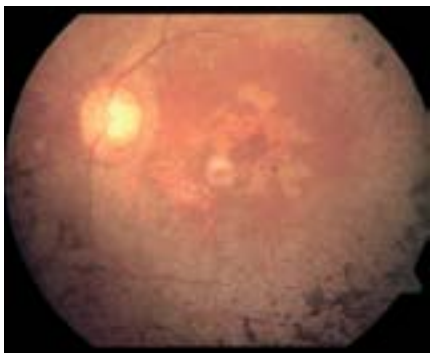


Fig. 12.57.7B: Retinitis pigmentosa—atrophic maculopathy

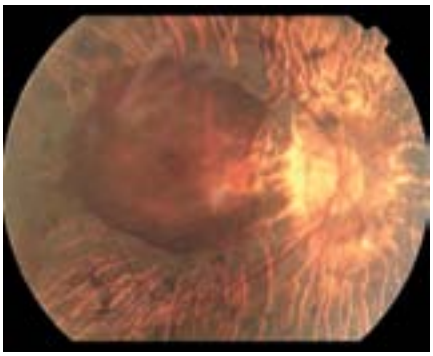


Fig. 12.57.8A: RP with epiretinal membrane

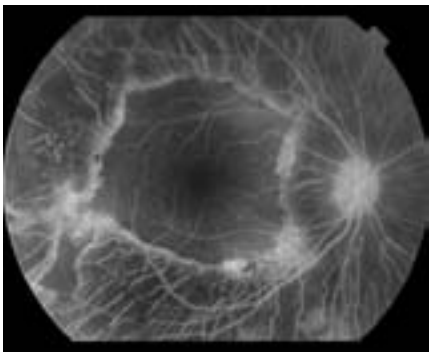


Fig. 12.57.8B: RP with epiretinal membrane on FFA

Atypical Retinitis Pigmentosa

- *Retinitis punctata albescens:* characterized by multiple scattered white dots, mostly between the posterior pole and the equator **(Fig. 12.58.1)**. Other findings are similar to typical RP, like CME **(Fig. 12.58.2)**
- *Retinitis pigmentosa sine pigmento:* Presence of arteriolar attenuation and waxy-pallor of the disk with subnormal ERG
- *Unilateral retinitis pigmentosa* **(Figs 12.58.3A and B)**
- *Sectorial retinitis pigmentosa:* One quadrant or one half (usually inferior) of the fundus **(Figs 12.58.4A and B)**

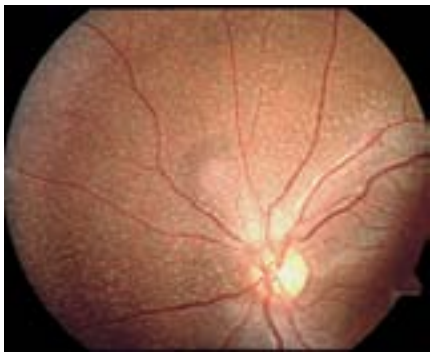


Fig. 12.58.1: Retinitis punctata albescens

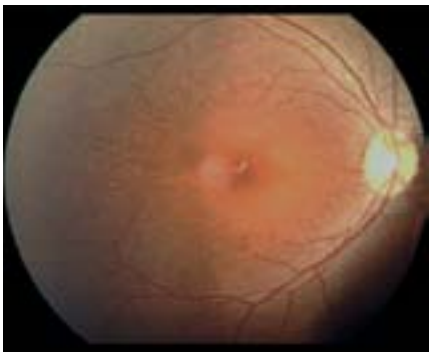


Fig. 12.58.2: Retinitis punctata albescens—CME

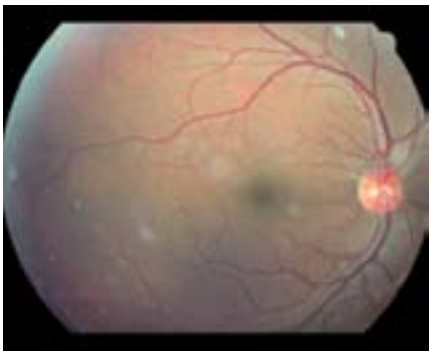


Fig. 12.58.3A: Retinitis pigmentosa—normal eye—RE

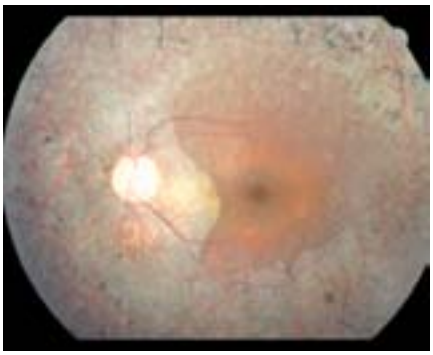


Fig. 12.58.3B: Retinitis pigmentosa—unilateral—LE

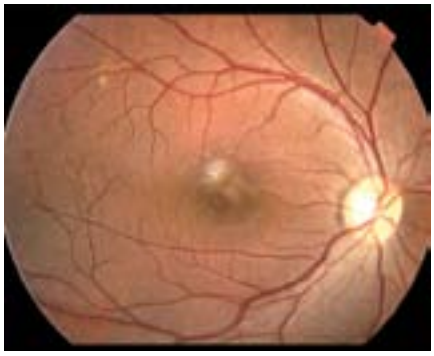


Fig. 12.58.4A: Retinitis pigmentosa—normal eye—RE

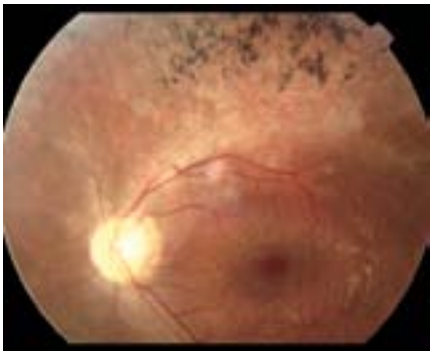


Fig. 12.58.4B: Retinitis pigmentosa—unilateral sectorial—LE

- *Pericentric or central retinitis pigmentosa:* RP, except that the pigmentary changes are confined to central or pericentral area, sparing the periphery **(Figs 12.58.5A and B)**
- *With systemic association*
  - *Laurence-Moon-Biedl syndrome:* RP, mental retardation, obesity, polydactyly and hypogonadism **(Fig. 12.58.6)**
  - *Refsum's disease:* RP, peripheral neuropathy, deafness, ataxia and ecchymosis
  - *Bassen-Kornzweig syndrome:* RP, ataxia, acanthocytosis, fat malabsorption

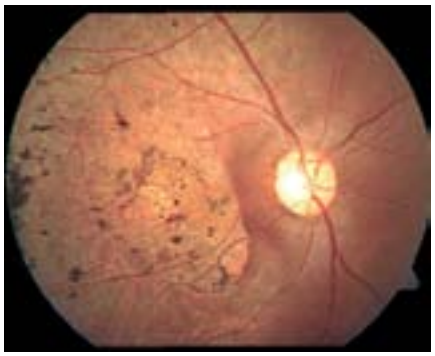


Fig. 12.58.5A: Retinitis pigmentosa—central

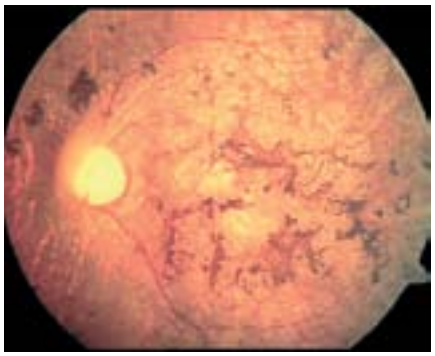


Fig. 12.58.5B: Retinitis pigmentosa—central

Fig. 12.58.6: Retinitis pigmentosa in both siblings—LMB syndrome—note polydactyly

  - *Usher syndrome:* RP and sensory-neural deafness
  - *Cockayne's syndrome:* RP, dwarfism, bird-like face, ataxia, mental retardation and premature aging
  - *Kearn-Sayre syndrome:* RP, ocular myopathy (CPEO) and heart block
  - *NARP syndrome:* Neuropathy, ataxia, and RP (NARP) is a rare hereditary condition. It is related to changes in mitochondrial DNA (mutations in the MT-ATP6 gene).

Retinal Detachment

Rhegmatogenous Retinal Detachment

- Detached retina is slightly opaque, convex and corrugated in appearance with loss of underlying choroidal pattern **(Figs 12.59.1 and 12.59.2)**
- Blood vessels appear darker than normal attached retina with macula on both for inferior and superior RD **(Figs 12.59.3 and 12.59.4)**
- Detached retina undulates freely with ocular movements
- Vitreous shows 'tobacco dust' (Shaffer's sign) in the anterior vitreous, with posterior vitreous detachment

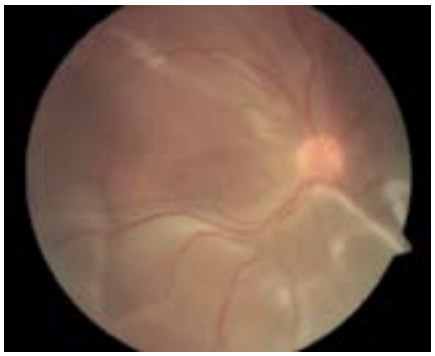


Fig. 12.59.1: Rhegmatogenous retinal detachment

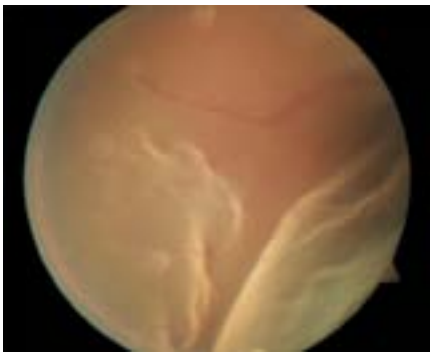


Fig. 12.59.2: Rhegmatogenous retinal detachment

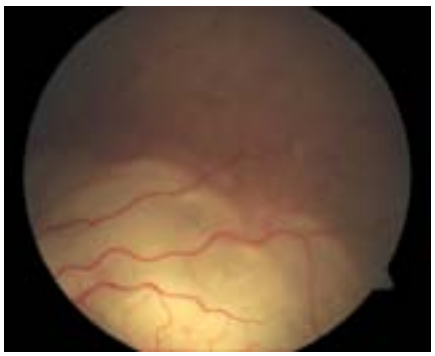


Fig. 12.59.3: Rhegmatogenous retinal detachment—inferior RD—macula on

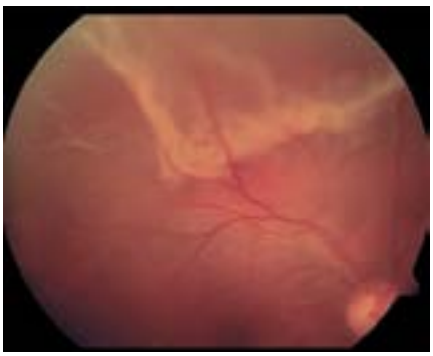


Fig. 12.59.4: Rhegmatogenous retinal detachment—superior RD—macula on

- Breaks appear as red areas (holes or tears) of discontinuity mainly in the peripheral retina **(Figs 12.59.5 and 12.59.6)**
- Sometimes, associated with choroidal detachment **(Figs 12.59.7A and B)**
- *In long standing cases:*
  - Retinal thinning with demarcation lines **(Fig. 12.59.8)**
  - Secondary intraretinal cyst and fibrosis **(Fig. 12.59.9)**
  - Proliferative vitreoretinopathy (PVR) develops depending upon the duration of the detachment **(Figs 12.59.10A to C)**
- If untreated, RDs become total and eventually gives rise to complicated cataract, chronic uveitis, hypotony and phthisis bulbi.

Associated Predisposing Factors

- *Lattice degeneration:* Peripheral, sharply demarcated, spindle shaped arborizing white lines arranged in a lattice pattern, often with variable pigmentation
- *Snail-track degeneration:* Peripheral, sharply demarcated tightly packed white snow flecks bands which appears frosted

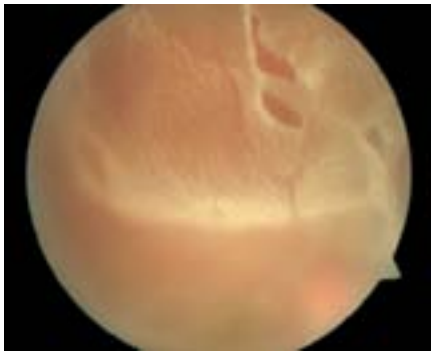


Fig. 12.59.5: Rhegmatogenous retinal detachment—multiple tears

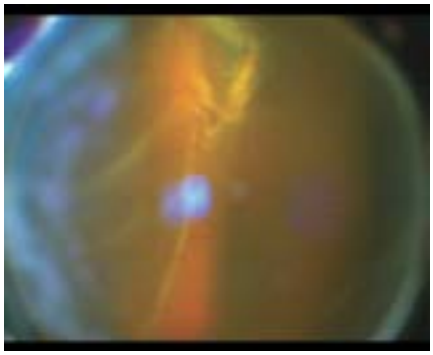


Fig. 12.59.6: Rhegmatogenous RD—horse-shoe tear

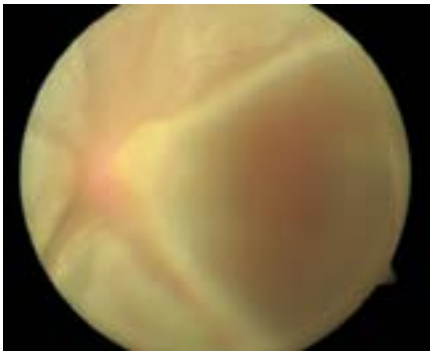


Fig. 12.59.7A: Rhegmatogenous RD—choroidal detachment

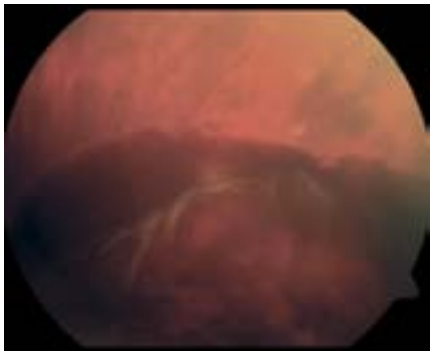


Fig. 12.59.7B: Rhegmatogenous RD—choroidal detachment

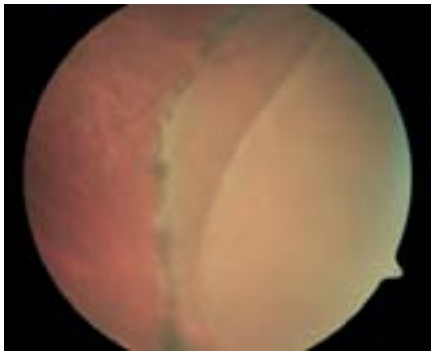


Fig. 12.59.8: Rhegmatogenous RD—demarcation line

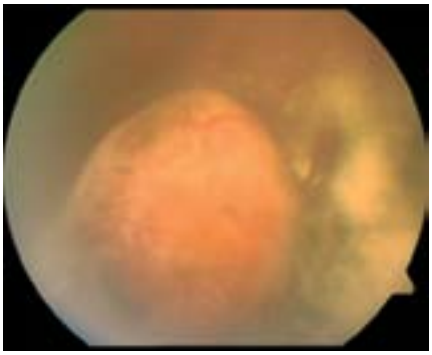


Fig. 12.59.9: Rhegmatogenous RD—intraretinal cyst

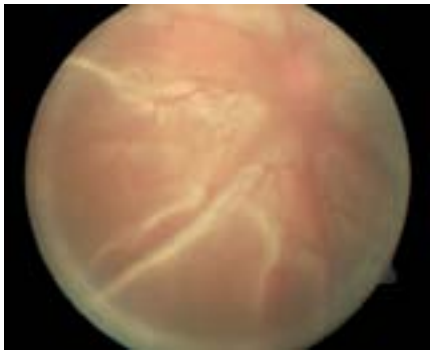


Fig. 12.59.10A: Rhegmatogenous RD—early PVR changes

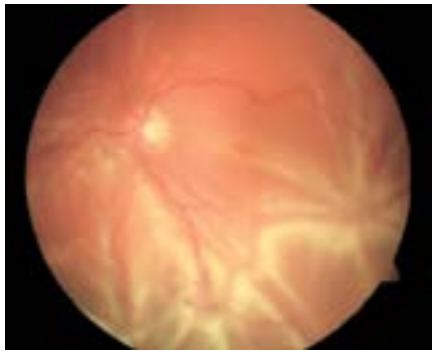


Fig. 12.59.10B: Rhegmatogenous RD—intermediate PVR changes

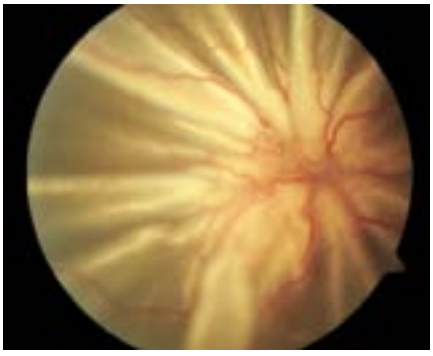


Fig. 12.59.10C: Rhegmatogenous RD—late PVR changes

- *Retinal holes:* A small break, usually circular, without any vitreous traction **(Fig. 12.60.1)**
- *Retinal tear:* A retinal break, usually horse-shoe shaped, with vitreous tractional element. A tear is more dangerous than a hole **(Fig. 12.60.2A)**. A tear may be of large size more than 1 clock hour—called *giant retinal tear* **(Fig. 12.60.2B)**
- *Retinal dialysis* **(Figs 12.60.3A and B)**.

Tractional Retinal Detachment

- Configuration of detached retina is concave
- Shallow detachment which seldom extends beyond equator **(Fig. 12.61.1)**
- Highest elevation of retina occurs at the site of vitreoretinal traction **(Fig. 12.61.2)**
- Mobility of detached retina is severely reduced
- Retinal breaks are usually absent
- *Causes:* Proliferative retinopathies **(Figs 12.61.3A and B)**, penetrating injuries **(Figs 12.61.4A and B)** and severe intraocular inflammation **(Fig. 12.61.5).**

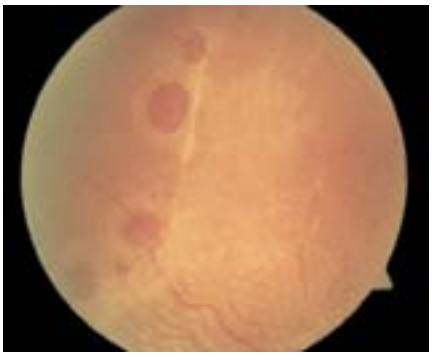


Fig. 12.60.1: Rhegmatogenous RD—multiple retinal holes

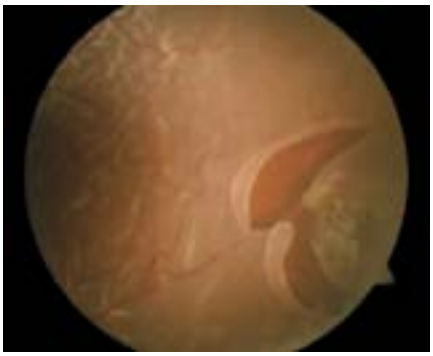


Fig. 12.60.2A: Rhegmatogenous RD—large horse-shoe tear

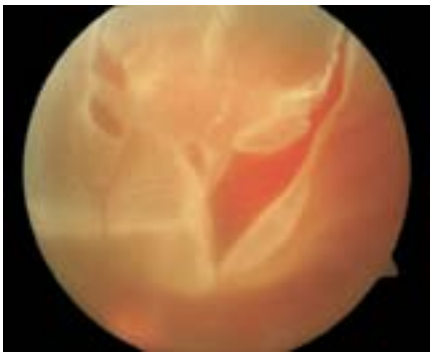


Fig. 12.60.2B: RRD—giant retinal tears

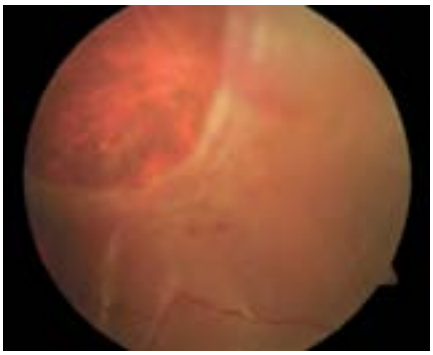


Fig. 12.60.3A: Rhegmatogenous RD with retinal retinal dialysis

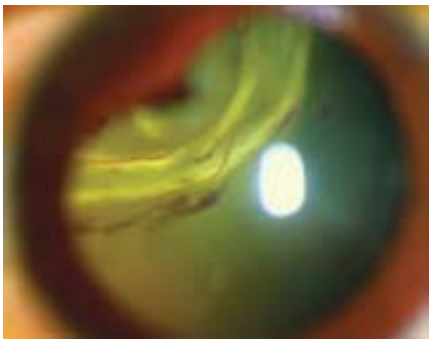


Fig. 12.60.3B: Rhegmatogenous RD with retinal dialysis

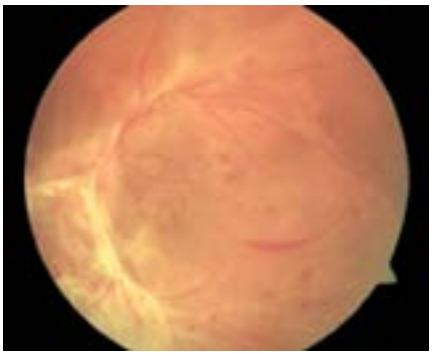


Fig. 12.61.1: Tractional RD—traumatic

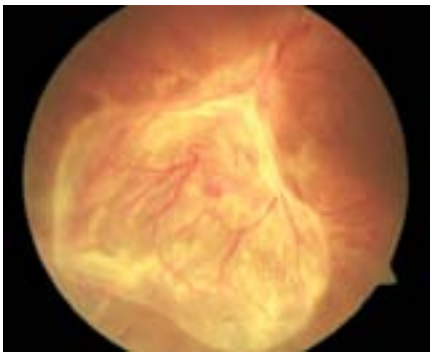


Fig. 12.61.2: Tractional RD

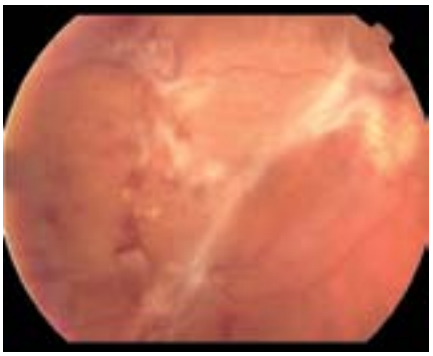


Fig. 12.61.3A: Tractional RD—PDR

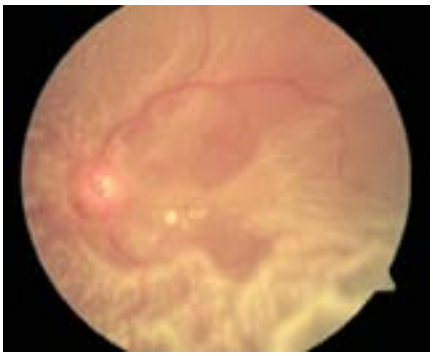


Fig. 12.61.3B: Tractional RD—PDR

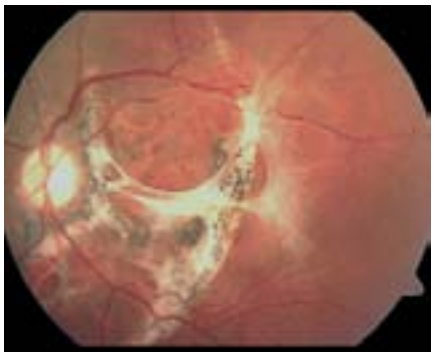


Fig. 12.61.4A: Tractional RD—traumatic

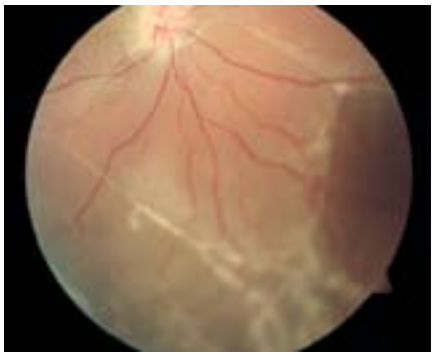


Fig. 12.61.4B: Tractional RD—traumatic

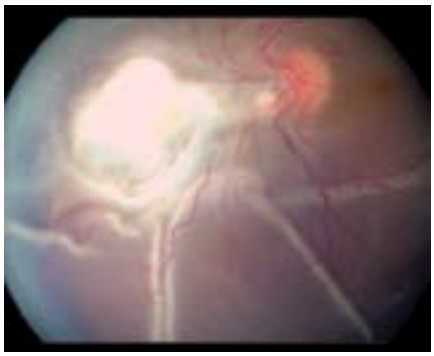


Fig. 12.61.5: Tractional RD—inflammatory—toxocara infection

Exudative Retinal Detachment

- RD configuration is very much convex
- Detached retina is smooth, non-corrugated and bullous **(Fig. 12.62.1)**
- May even touch the back of the lens in severe cases
- Marked retinal mobility
- Shifting of fluid is the hallmark of exudative detachment due to force of gravity **(Figs 12.62.2A and B)**
- Retinal breaks are absent
- Obvious ocular pathology is frequently evident and may associated with choroidal detachment **(Fig. 12.62.3)**
- *Causes:* Choroidal tumors, VKH syndrome, posterior scleritis, hypertension, toxemia of pregnancy, uveal effusion syndrome, etc. **(Fig. 12.62.4)**.

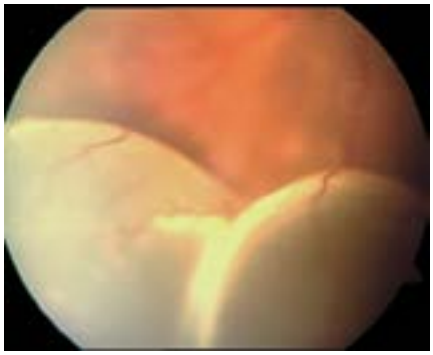


Fig. 12.62.1: Exudative RD—bullous detachment

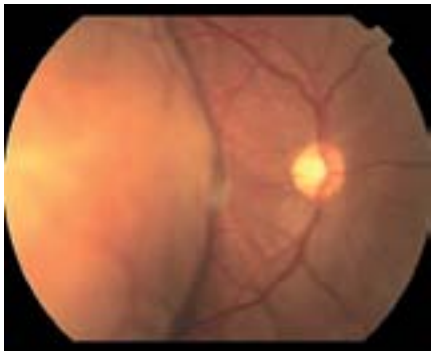


Fig. 12.62.2A: Exudative RD—shifting of fluid

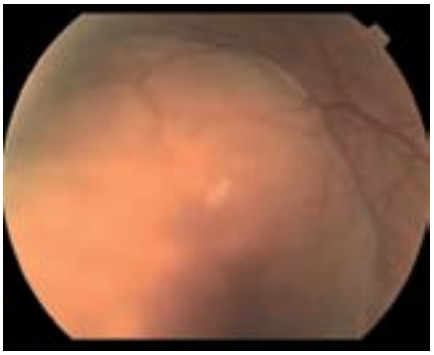


Fig. 12.62.2B: Exudative RD—shifting of fluid

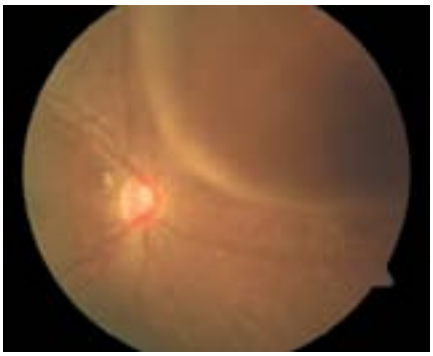


Fig. 12.62.3: Exudative RD with choroidal detachment

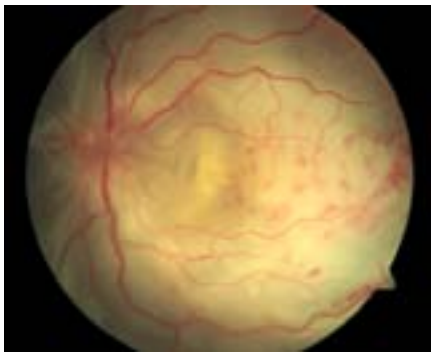


Fig. 12.62.4: Exudative retinal detachment in VKH syndrome

Hypertensive Retinopathy

- Focal retinal edema, superficial retinal hemorrhage and hard exudates in chronic hypertension **(Figs 12.63.1A to D)**
- Hard exudates often deposit around the macula as macular star
- Cotton-wool patches in acute or severe hypertension **(Figs 12.63.2A and B)**
- Papilledema including the neuroretinal edema may occur in malignant hypertension **(Figs 12.63.3A and B)**
- Various types of arteriovenous crossing changes
- Associated choroidal changes like, RPE changes, pigment deposition along the blocked choroidal vessels, Elschnig's spot, etc.
- Exudative bullous detachment in severe cases.

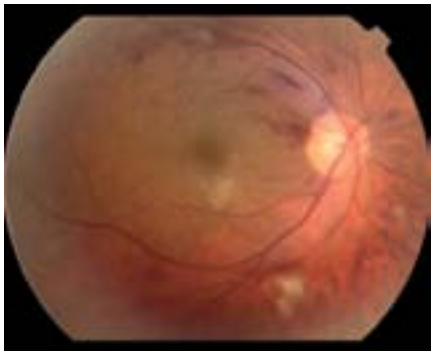


Fig. 12.63.1A: Hypertensive retinopathy—superficial flame shaped hemorrhage—RE

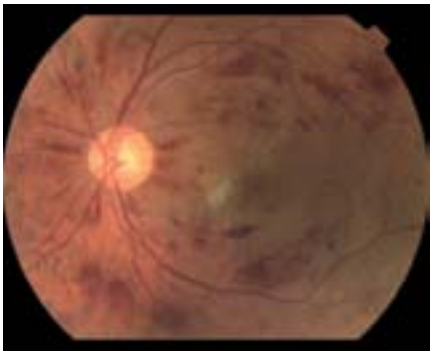


Fig. 12.63.1B: Hypertensive retinopathy—superficial flame shaped hemorrhage—LE

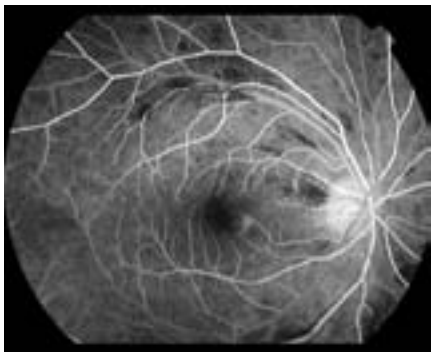


Fig. 12.63.1C: Hypertensive retinopathy—superficial flame shaped hemorrhage—RE

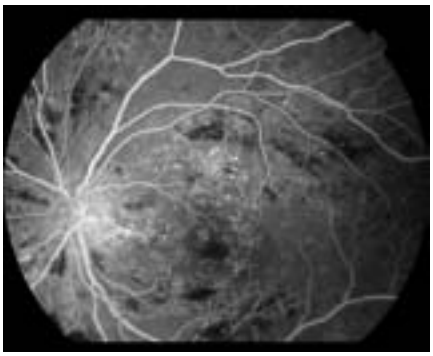


Fig. 12.63.1D: Hypertensive retinopathy—superficial flame shaped hemorrhage—LE

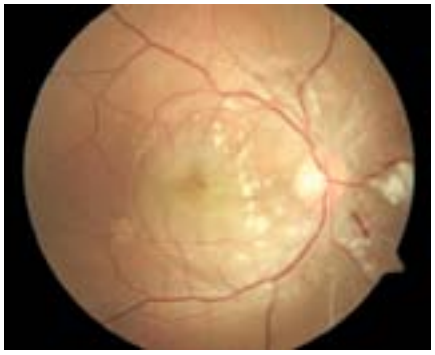


Fig. 12.63.2A: Hypertensive retinopathy—soft exudates—RE

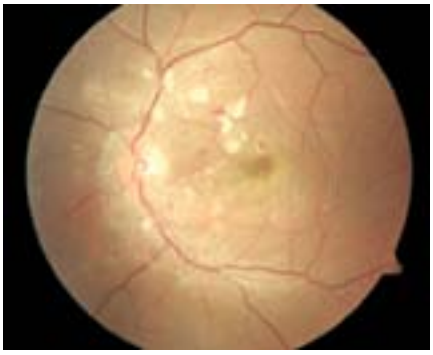


Fig. 12.63.2B: Hypertensive retinopathy—soft exudates—LE

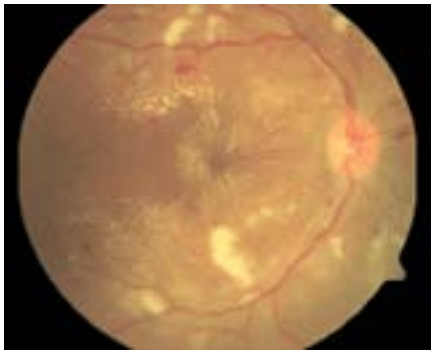


Fig. 12.63.3A: Malignant hypertensive retinopathy—RE

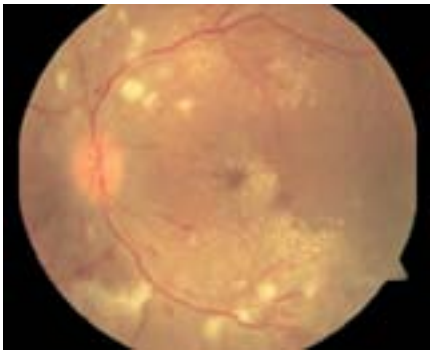


Fig. 12.63.3B: Malignant hypertensive retinopathy—LE

Diabetic Retinopathy

Nonproliferative Diabetic Retinopathy (NPDR)

- Microaneurysm, small 'dot' and 'blot' hemorrhage **(Figs 12.64.1A and B)**
- Vascular changes, like—looping, beading, etc. **(Figs 12.64.2A and B)**
- Intraretinal microvascular abnormalities (IRMA)
- Hard exudates **(Figs 12.64.3A and B)**
- Further classified as mild, moderate, severe and very severe NPDR.

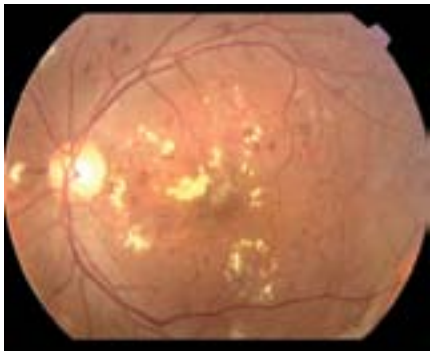


Fig. 12.64.1A: NPDR—dot and blot hemorrhage and hard exudates

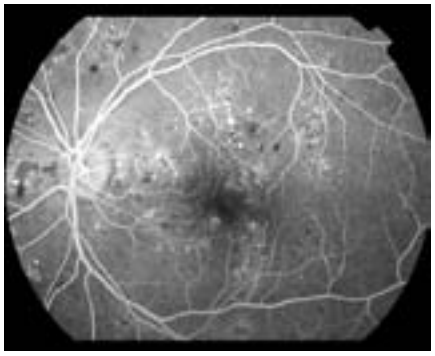


Fig. 12.64.1B: NPDR—dot and blot hemorrhage and hard exudates—FFA

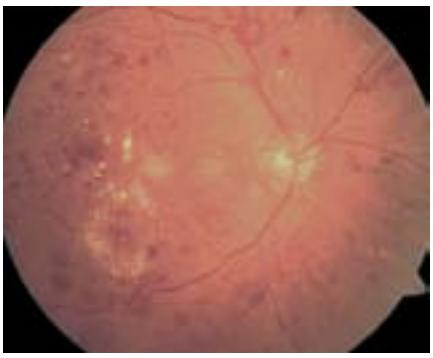


Fig. 12.64.2A: NPDR—looping, dot and blot hemorrhage

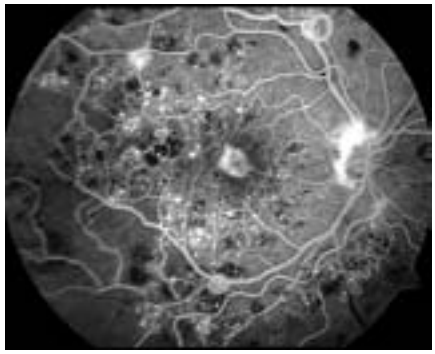


Fig. 12.64.2B: NPDR—vascular looping, dot and blot hemorrhage

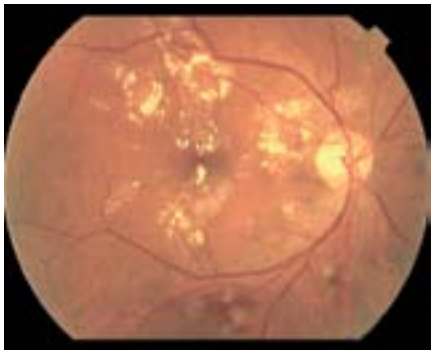


Fig. 12.64.3A: NPDR—hard exudates—RE

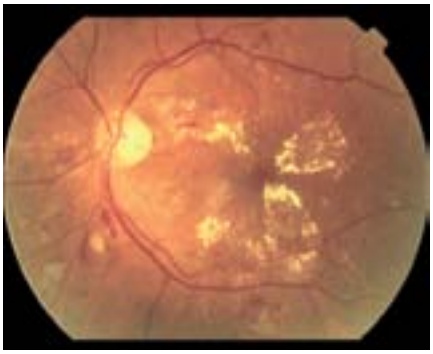


Fig. 12.64.3B: NPDR—hard exudates—LE

Diabetic Maculopathy

- Clinically significant macular edema (CSME) is defined as:
  - Retinal edema or thickening at or within 500 micron of the center of fovea
  - Hard exudates at or within 500 micron of the center of fovea **(Figs 12.65.1 to 12.65.3).**

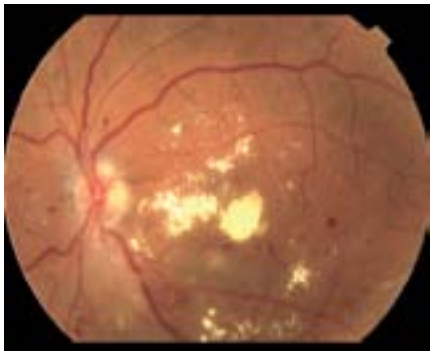


Fig. 12.65.1: NPDR with CSME

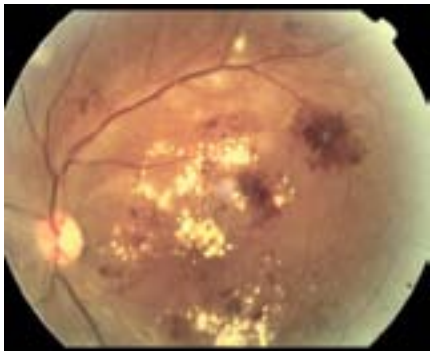


Fig. 12.65.2: NPDR—CSME with large dot and blot hemorrhage

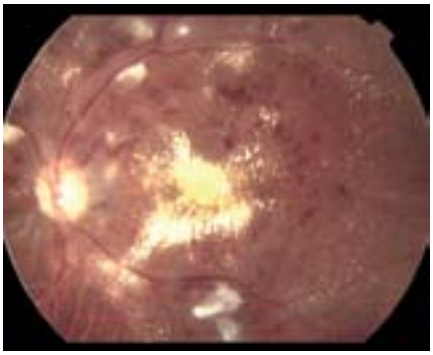


Fig. 12.65.3: NPDR with CSME

– Area of retinal thickening of 1 dd size (1500 micron) or at least part of which within 1 dd of the center of fovea.

Proliferative Diabetic Retinopathy (PDR)

- Affects 5% of diabetic population and more with insulin dependent diabetes mellitus (IDDM)
- Neovascularization is the hallmark of PDR **(Figs 12.66.1 and 12.66.2)**
- It may be asymmetrical—one eye may be of NPDR changes and other eye may have PDR **(Figs 12.66.3A to D)**

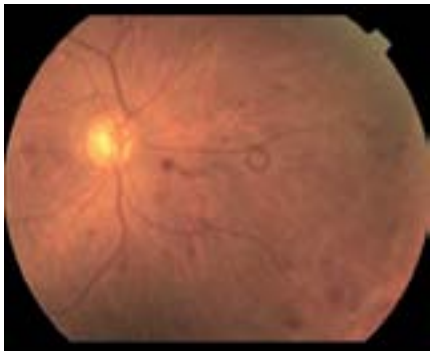


Fig. 12.66.1A: Early PDR with vascular looping

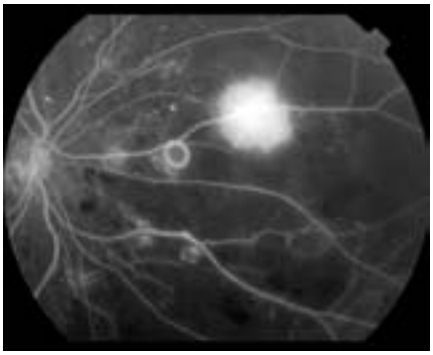


Fig. 12.66.1B: Early PDR with vascular looping and NVE on FFA

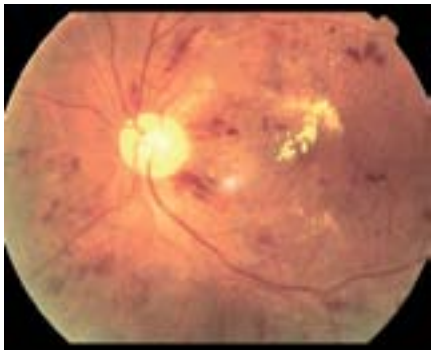


Fig. 12.66.2A: Early PDR—NVE at lower temporal arcade

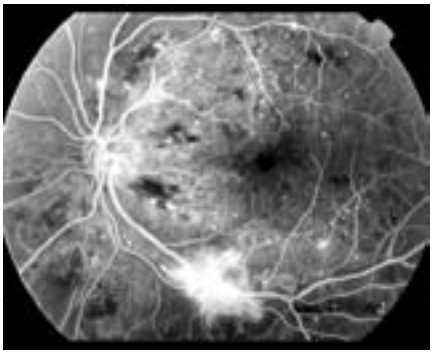


Fig. 12.66.2B: Early PDR—NVE at lower temporal arcade on FFA

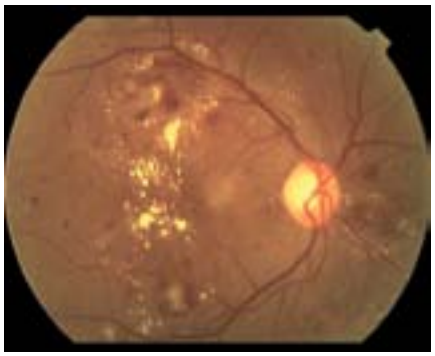


Fig. 12.66.3A: NPDR—RE

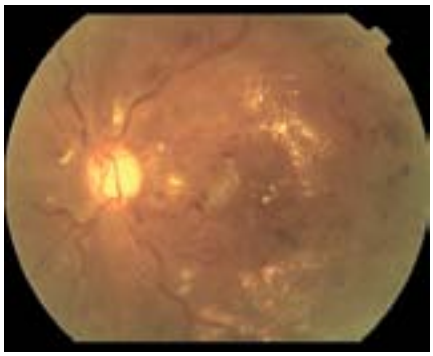


Fig. 12.66.3B: Early PDR with NVD—LE

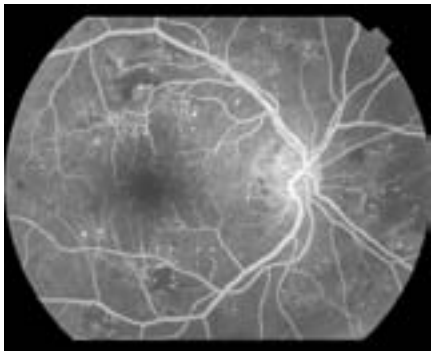


Fig. 12.66.3C: NPDR on FFA—RE

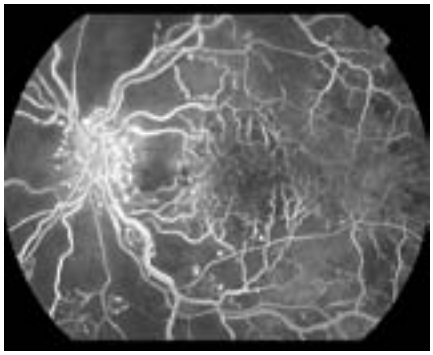


Fig. 12.66.3D: Early PDR with NVD and capillary drop out areas—LE

- New vessels at the disk (NVD), or along the course of major blood vessels, called NVE—'new vessels elsewhere'**(Figs 12.66.4 to 12.66.6)**
- Posterior vitreous detachment
- Hemorrhage may occur in the form of intraretinal, preretinal and vitreous hemorrhage
- Laser panretinal photocoagulation (PRP) is a part of treatment in PDR **(Fig. 12.66.7)**.

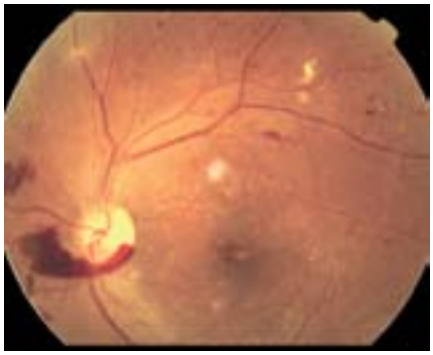


Fig. 12.66.4A: Late PDR with NVD and NVE

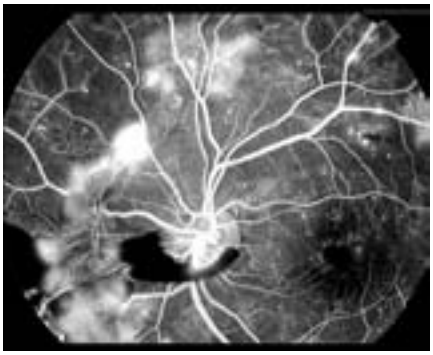


Fig. 12.66.4B: Late PDR with NVD and NVE on FFA

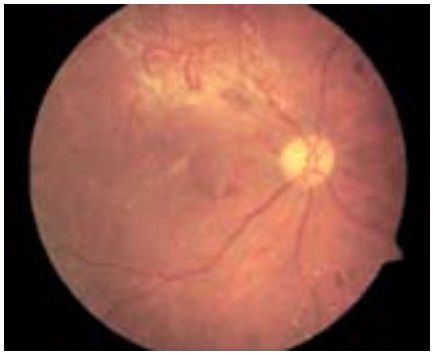


Fig. 12.66.5A: Late PDR with NVE

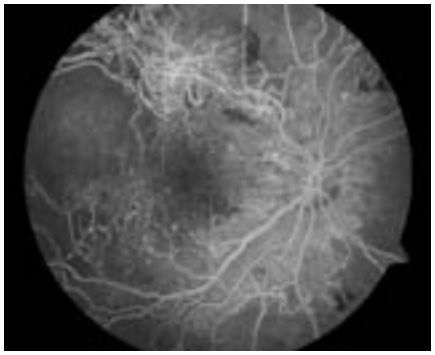


Fig. 12.66.5B: Late PDR showing NVE and capillary nonperfusion area on FFA

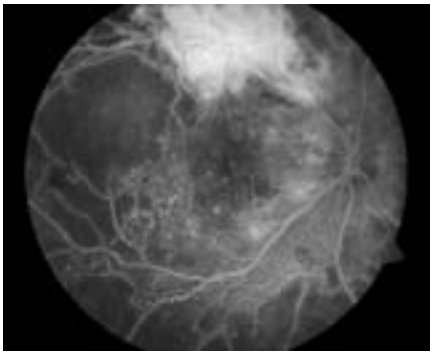


Fig. 12.66.5C: Late PDR showing NVE and capillary nonperfusion area on FFA

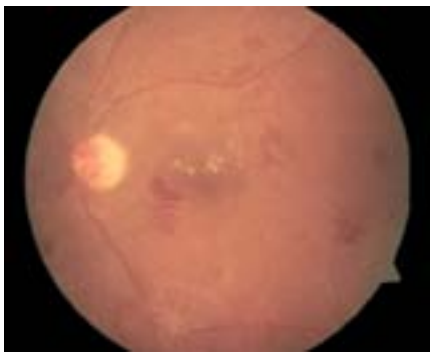


Fig. 12.66.6A: Late PDR with NVD and NVE

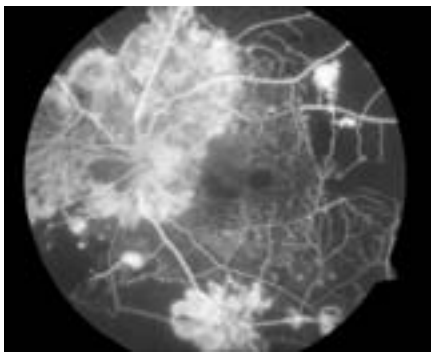


Fig. 12.66.6B: Late PDR with NVD and NVE showing macular ischemia on FFA

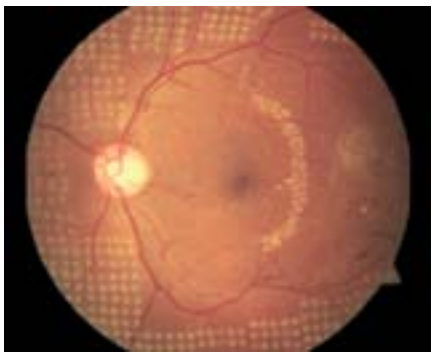


Fig. 12.66.7: Laser panretinal and macular grid photocoagulation

Advanced Proliferative Diabetic Retinopathy (Advanced PDR)

- Central or branch retinal venous occlusion **(Figs 12.67.1A and B)**

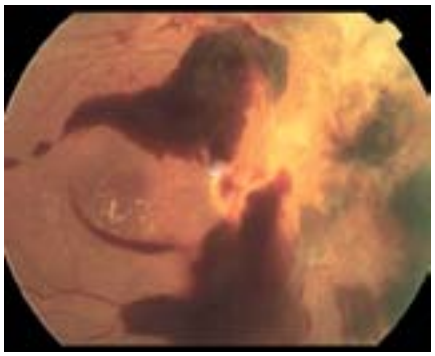


Fig. 12.67.1A: Advanced PDR—CRVO with preretinal hemorrhage

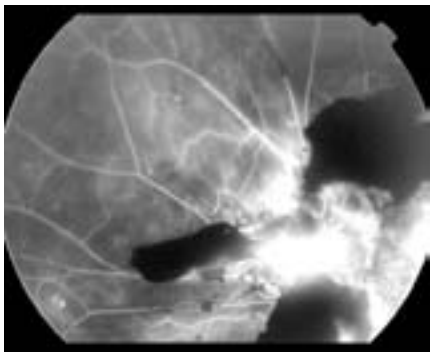


Fig. 12.67.1B: Advanced PDR—CRVO with preretinal hemorrhage

- Recurrent vitreous hemorrhage **(Figs 12.67.2 and 12.67.3)**
- Tractional retinal detachment **(see Figs 12.61.3A and B)**
- Brunt out proliferative diabetic retinopathy and ultimately featureless retina **(Figs 12.67.4A to D)**
- Neovascular glaucoma with or without disk cupping.

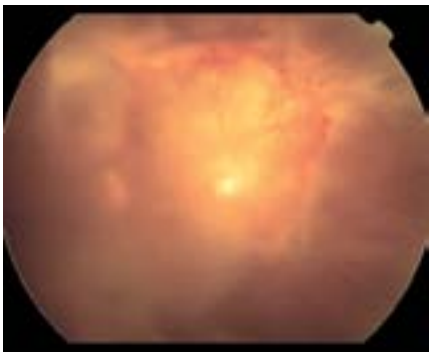


Fig. 12.67.2A: Advanced PDR—vitreous hemorrhage from NVD

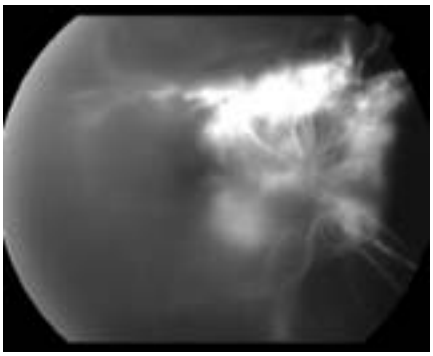


Fig. 12.67.2B: Advanced PDR—vitreous hemorrhage from NVD

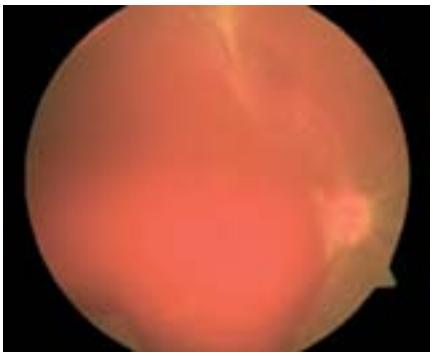


Fig. 12.67.3A: Advanced PDR—vitreous hemorrhage and preretinal hemorrhage

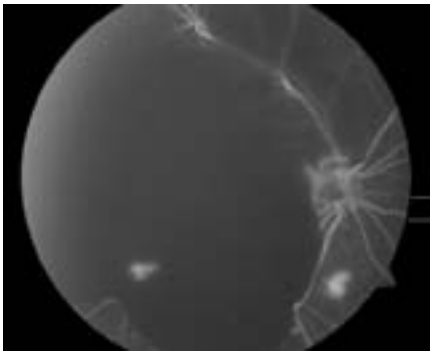


Fig. 12.67.3B: Advanced PDR—vitreous hemorrhage and preretinal hemorrhage

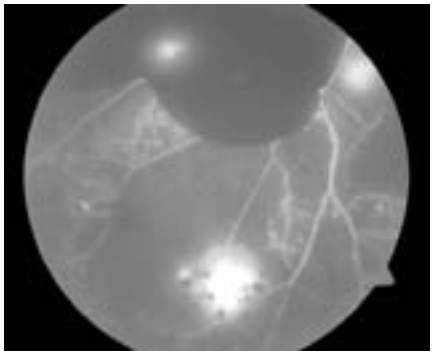


Fig. 12.67.3C: Advanced PDR—vitreous hemorrhage and preretinal hemorrhage

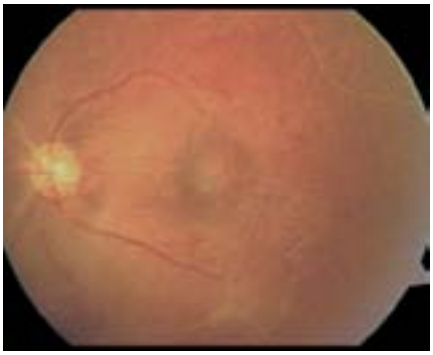


Fig. 12.67.4A: Advanced PDR—featureless retina

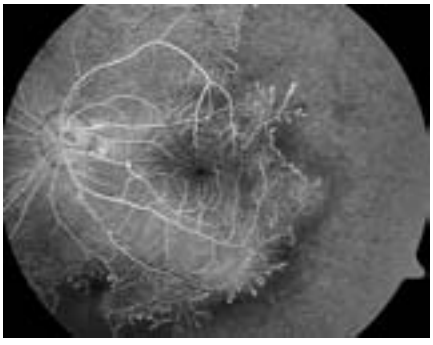


Fig. 12.67.4B: Advanced PDR—featureless retina

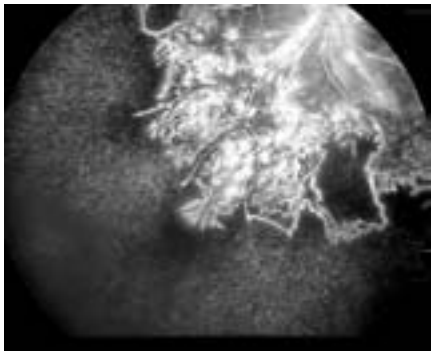


Fig. 12.67.4C: Advanced PDR—featureless retina

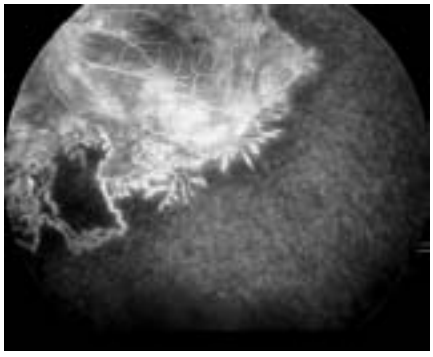


Fig. 12.67.4D: Advanced PDR—featureless retina

Retinopathy of Prematurity (ROP)

- Occurs some weeks after birth, in premature infants (less than 1500 gm birth weight), who have been given high concentration of oxygen during the first 10 days of life
- Appearance of hazy white patches in the temporal peripheral retina **(Figs 12.68.1A and B)**

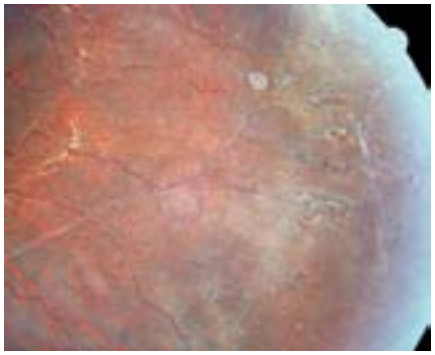


Fig. 12.68.1A: Retinopathy of prematurity

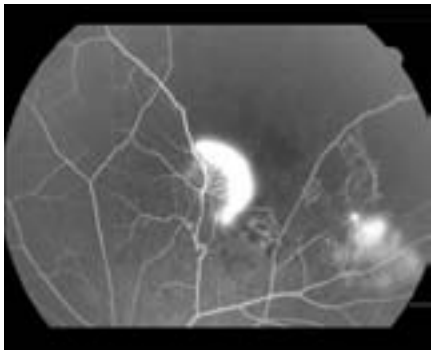


Fig. 12.68.1B: Retinopathy of prematurity

- Milder forms result in sea fan neovascularization **(Figs 12.68.2A and B)** and ultimately tractional bands at upper temporal quadrant producing an ectopic macula or dragged optic disk **(Fig. 12.68.3)**
- In severe cases, fibrovascular tissue proliferates to form a mass behind the lens—causing *pseudoglioma and leukocoria (See Chapter 11)*
- Eventually tractional retinal detachment occurs and the vision is usually lost.

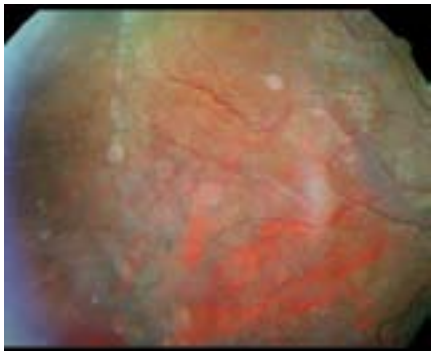


Fig. 12.68.2A: ROP—sea fan neovascularization

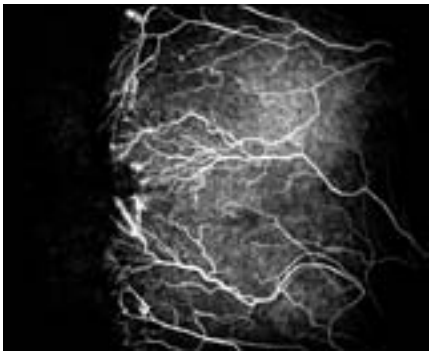


Fig. 12.68.2B: ROP—sea fan neovascularization

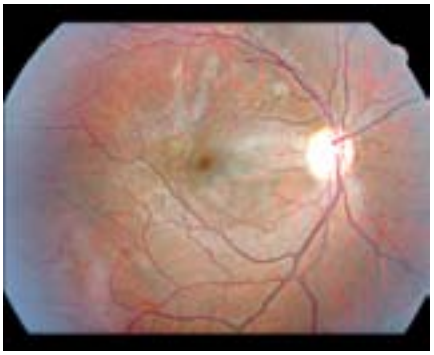


Fig. 12.68.3: Retinopathy of prematurity—ectopic macula

Retinoblastoma

- Malignant intraocular tumor originating in the outer nuclear layer
- Average age of presentation is 18 months and 20% cases are bilateral
- Leukocoria or 'amaurotic cat's eye reflex'—the most common mode of presentation in 60% of cases **(Fig. 12.69.1)**
- Squinting of the eye—the second most common mode of presentation (20%)
- Secondary glaucoma which may be associated with buphthalmos **(Fig. 12.69.2)**

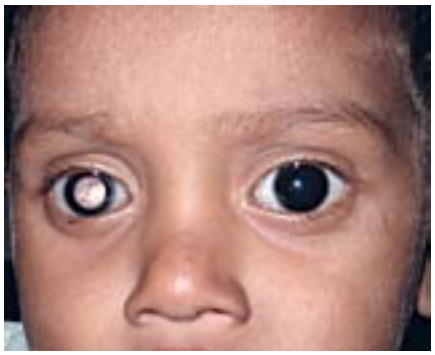


Fig. 12.69.1: Leukocoria—retinoblastoma—RE

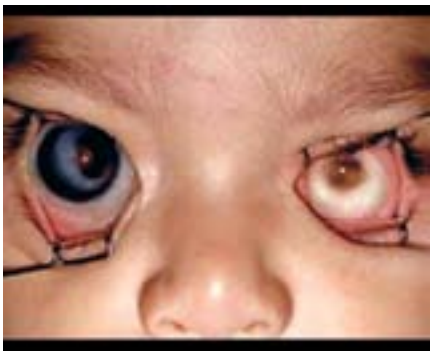


Fig. 12.69.2: Retinoblastoma—buphthalmos—RE and phthisis bulbi—LE

- In neglected cases, there is orbital involvement, leading to proptosis *(See Chapter 14)*
- Endophthalmitis or anterior uveitis may also be a presenting feature which may lead to atrophy of the globe **(Fig. 12.69.2)**
- *Endophytic type* is most common:
  - White or pearly-pink colored mass with sharp margin projects into the vitreous cavity **(Figs 12.69.3 to 12.69.5)**
  - Presence of calcium deposits giving an appearance 'cottage cheese' of the fundus **(Fig. 12.69.6)**
  - Multiple seedling may occur in vitreous cavity
- *Exophytic type* usually gives rise to exudative retinal detachment and the tumor itself is difficult to visualize
- Should be differentiated from other causes of leukocoria *(See Chapter 7)*
- *Treatment:* Enucleation, radiotherapy, chemotherapy, photocoagulation or cryotherapy and in extreme cases exenteration.

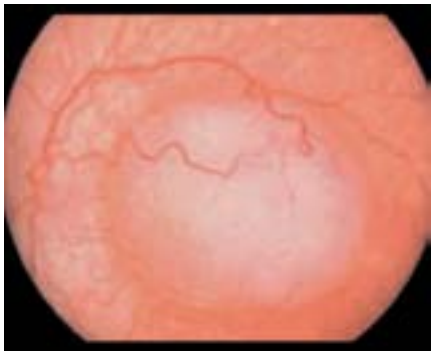


Fig. 12.69.3: Large endophytic RB

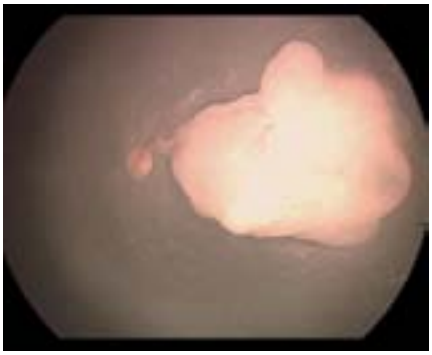


Fig. 12.69.4: Large endophytic RB

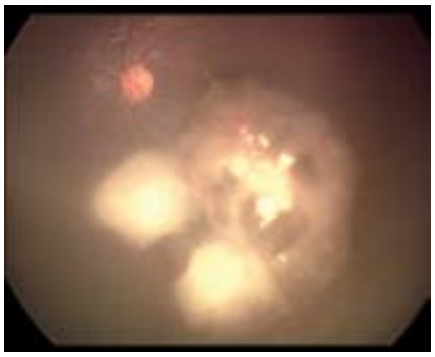


Fig. 12.69.5: Large endophytic retinoblastoma

Fig. 12.69.6: Retinoblastoma—cottage cheese appearance

Miscellaneous Retinal Conditions

Idiopathic Polypoidal Choroidal Vasculopathy

- Rare, unilateral condition which affects the darker women
- IPCV consists of dialated inner choroidal vessels at the macula or at the juxtapapillary region which causes repeated episodes of subretinal or sub-RPE hemorrhage
- Clinically orange-red subretinal nodules are seen in the posterior pole which represent polyps **(Figs 12.70.1 and 12.70.2A)**
- Multiple area of hemorrhagic PED surrounded by hard exudates
- FFA shows stippled hyperfluorescence in the region of polyp which become fuzzy in the late stage **(Figs 12.70.2B and 12.70.3)**

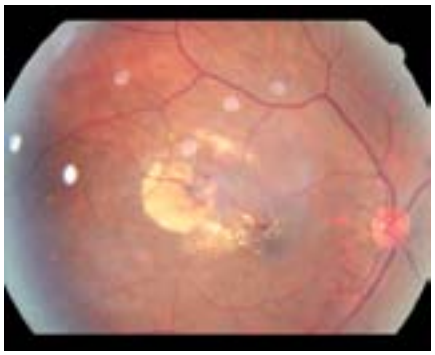


Fig. 12.70.1: Idiopathic polypoidal choroidal vasculopathy

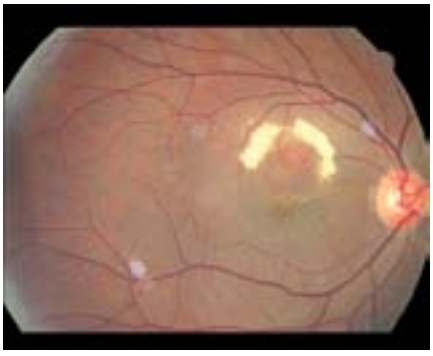


Fig. 12.70.2A: Idiopathic polypoidal choroidal vasculopathy

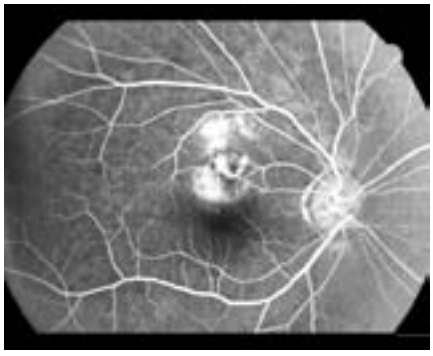


Fig. 12.70.2B: Idiopathic polypoidal choroidal vasculopathy—FFA

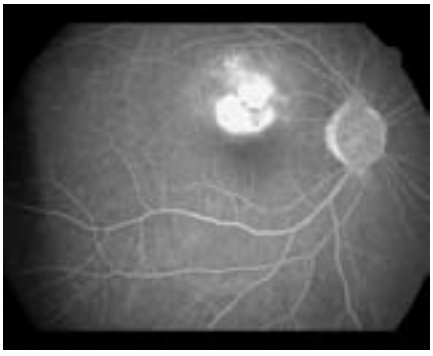


Fig. 12.70.2C: Idiopathic polypoidal choroidal vasculopathy—FFA

- An ICG angiography is more diagnostic
- Prognosis is usually good.

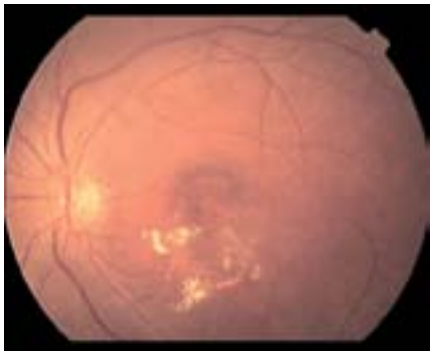


Fig. 12.70.3A: Idiopathic polypoidal choroidal vasculopathy

Fig. 12.70.3B: Idiopathic polypoidal choroidal vasculopathy

Familial Exudative Vitreoretinopathy

- Rare, dominant inherited condition, with a clinical picture similar to retinopathy of prematurity
- The condition is not associated with prematurity or low birth weight babies
- Dragged retinal vessels from the optic disk with an abrupt termination and retina at the equatorial area has a scalloped edge **(Figs 12.71.1A to C)**

Fig. 12.71.1A: Familial exudative vitreoretinopathy—dragged retina

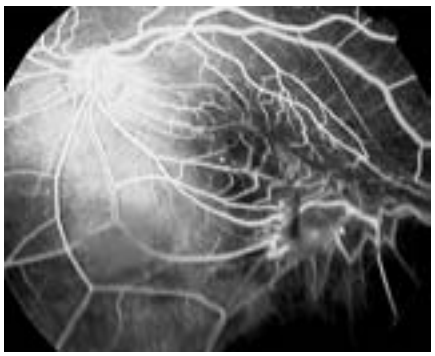


Fig. 12.71.1B: Familial exudative vitreoretinopathy on FFA

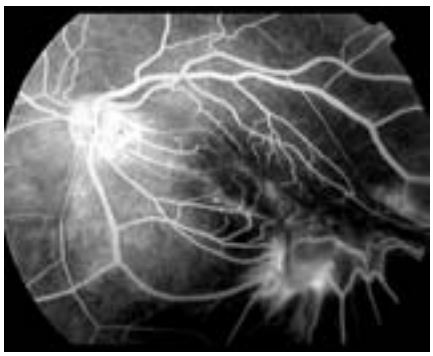


Fig. 12.71.1C: Familial exudative vitreoretinopathy on FFA

Stickler's Disease

- Autosomal dominant, with Marfanoid habitus
- Lattice-like radial perivascular pigmentation with overlying chorioretinal atrophy **(Figs 12.72.1 and 12.72.2)**
- Associated vitreous degeneration with liquefaction, myopia and cataract
- General orofacial and skeletal anomalies
- Increased risk of retinal detachment
- Periodic checkup is necessary to treat retinal breaks early.

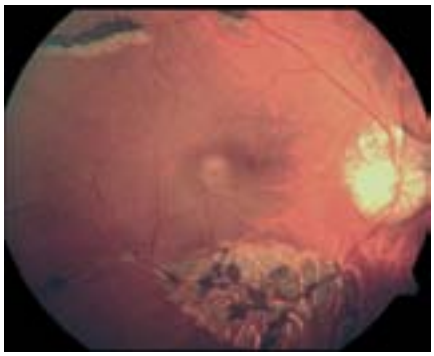


Fig. 12.72.1A: Stickler's disease

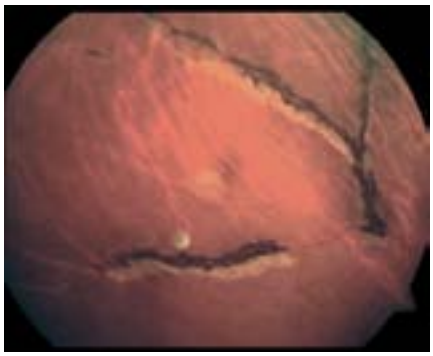


Fig. 12.72.1B: Stickler's disease

Idiopathic Juxtafoveal Telangiectasia

- Rare, unilateral or bilateral; may be congenital or acquired
- Presence of focal microaneurysm or saccular dilatation of some pattern, of the juxtafoveal or parafoveal capillary network

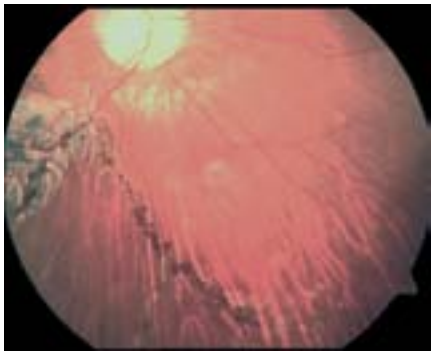


Fig. 12.72.2A: Stickler's disease

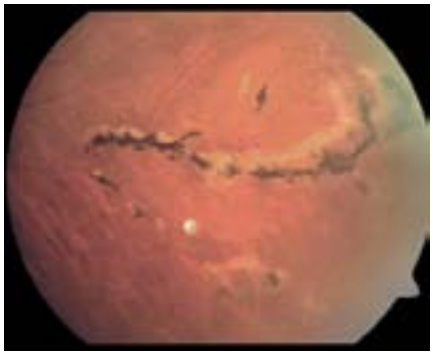


Fig. 12.72.2B: Stickler's disease

- It also involves the area temporal to the fovea with formation of SRNVMs **(Figs 12.73.1A and B)**
- Entire network may involve in late stage and in some cases progressive obliteration of parafoveolar capillary networks
- FFA shows pathology clearly with late staining **(Figs 12.73.2A to D)**.

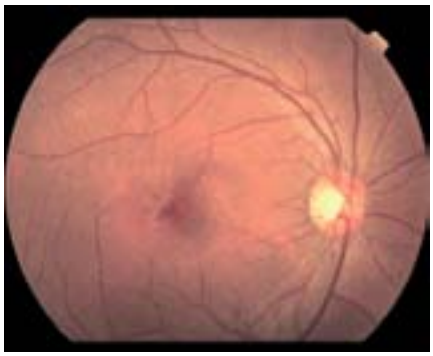


Fig. 12.73.1A: Idiopathic juxtafoveal telangiectasia—unilateral

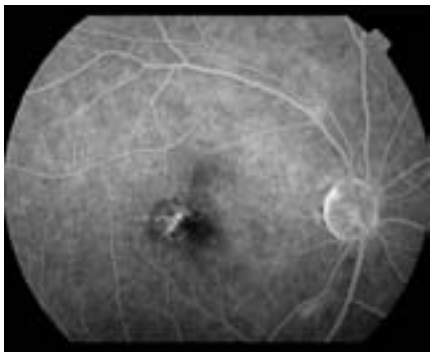


Fig. 12.73.1B: Idiopathic juxtafoveal telangiectasia—unilateral on FFA

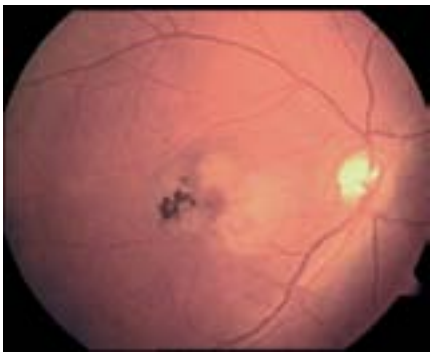


Fig. 12.73.2A: Idiopathic juxtafoveal telangiectasia—RE

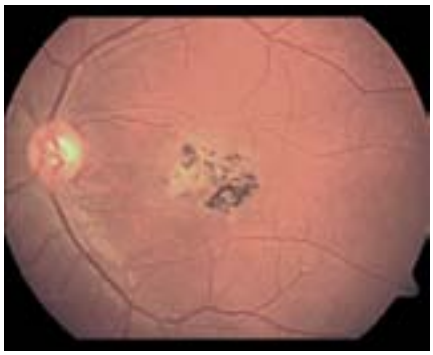


Fig. 12.73.2B: Idiopathic juxtafoveal telangiectasia—LE

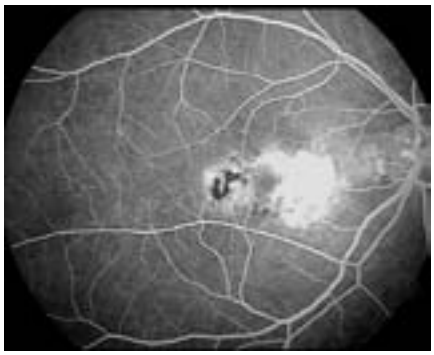


Fig. 12.73.2C: Idiopathic juxtafoveal telangiectasia—RE on FFA

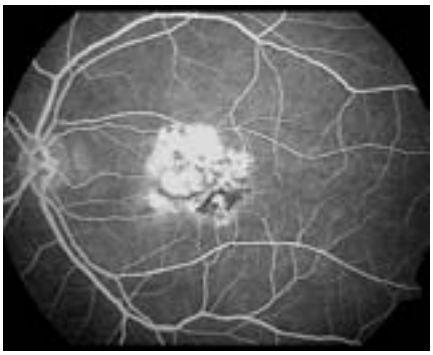


Fig. 12.73.2D: Idiopathic juxtafoveal telangiectasia—LE on FFA

Goldmann-Favre Syndrome

- It is an autosomal recessive disorder.
- This is also called "enhanced S-cone" or "blue cone" syndrome
- Patients with Goldmann-Favre syndrome typically experience retinoschisis or edema of the macula, pigmentary degeneration of the retina **(Figs 12.74.1A and B)**, hemeralopia, liquefied vitreous body and extinguished electroretinogram. Cataract is also a common feature
- *Treatment:* No effective treatment for the disorder, but visual function can be improved with low vision aids. Cataract surgery may be beneficial.

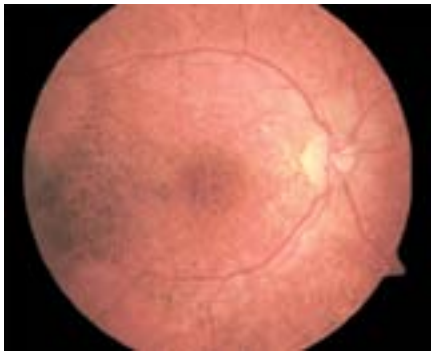


Fig. 12.74.1A: Goldmann-Favre syndrome

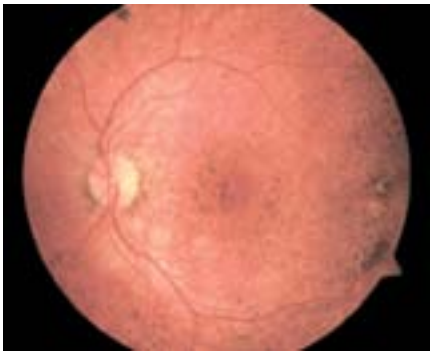


Fig. 12.74.1B: Goldmann-Favre syndrome

Congenital Hypertrophy of the Retinal Pigment Epithelium (CHRPE)

- Benign pigmented fundus lesions that commonly discovered during routine eye examination
- Usually asymptomatic
- Well-demarcated, round, solitary or multiple gray-brown or black lesions which have flat or scalloped margins
- Depigmented or hypopigmented punched-out lacunae or fenestration lesions may be evident within larger lesions

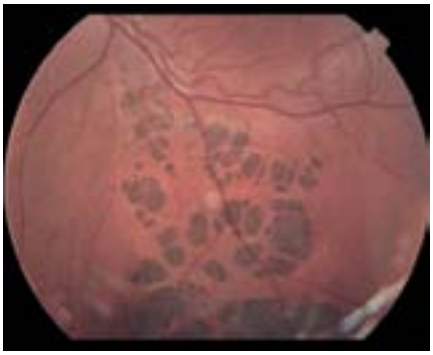


Fig. 12.75.1: Congenital hypertrophy of the retinal pigment epithelium (CHRPE)

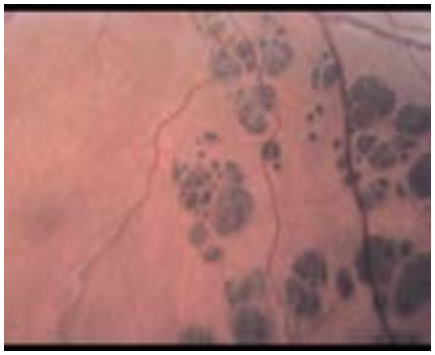


Fig. 12.75.2: CHRPE—bear tracks

- Multiple areas of grouped CHRPE simulating the animal foot-print are also called *'bear tracks'* **(Figs 12.75.1 and 12.75.2)**
- Generally located in the peripheral but may occasionally in the peripapillary region
- FFA demonstrates blocked choroidal fluorescence by the hypertrophied RPE and no leakage of dye.

Retinoschisis

- ***Acquired Retinoschisis:***
  - Characterized by the abnormal splitting of the retina's neurosensory layers, usually in the outer plexiform layer
  - Usually asymptomatic and nonprogressive
  - Ophthalmoscopy reveals a smooth, stationary, bullous elevation of either the inferotemporal or superotemporal retina with multiple holes **(Fig. 12.76.1)**
  - Blood vessels traverse the dome and cast shadows on the underlying structures
- *Treatment:* Risk of detachment occurring in a retinoschisis (even with double layer holes) is exceedingly low, so there is no treatment beyond routine monitoring every 6 –12 months.
- ***X-linked Juvenile Retinoschisis:***
  - Slowly progressive with decreased vision
  - Foveal schisis with fine radiating folds from fovea; spoke-like foveal cysts and microcystic foveal elevation **(Figs 12.76.2A and B)**
  - Vitreous hemorrhage often the cause of decreased vision
  - A macular OCT which shows macular cysts in fovea schisis is useful to confirm the diagnosis **(Figs 12.76.2C and D)**
- *Treatment:* Children with juvenile retinoschisis should avoid even in minor trauma; periodic check up is essential.

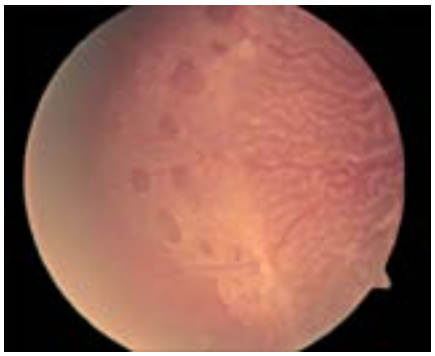


Fig. 12.76.1: Retinoschisis—multiple small holes

Fig. 12.76.2A: X-linked juvenile retinoschisis—RE

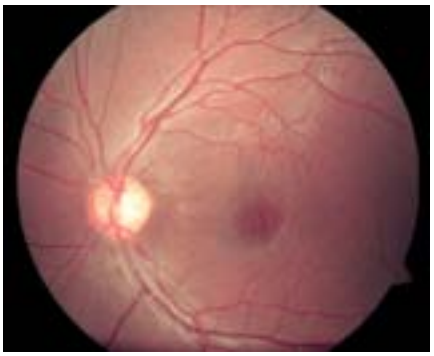


Fig. 12.76.2B: X-linked juvenile retinoschisis—LE

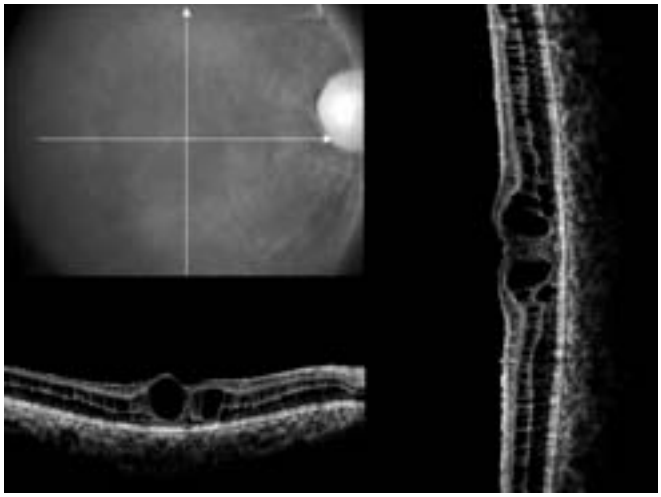


Fig. 12.76.2C: X-linked juvenile retinoschisis—RE

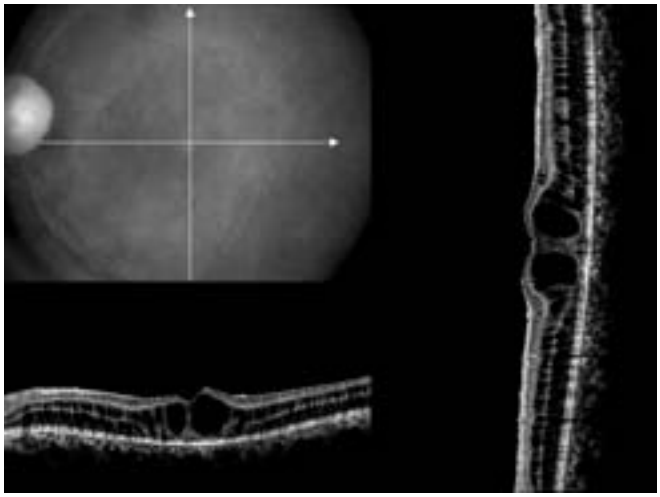


Fig. 12.76.2D: X-linked juvenile retinoschisis—LE

Purtscher's Retinopathy

- Bilateral, multiple patches of retinal whitening, cotton wool exudates, and hemorrhages that surround the optic disk **(Figs 12.77.1A and B)**. Happens after severe compressive injury of the chest, multiple long bone fractures with fat emboli or head injury
- May have disk edema and a RAPD
- Similar type of retinopathy may be seen in pancreatitis, leukemias, amniotic fluid embolism
- *Treatment:* Usually resolves within weeks or months.

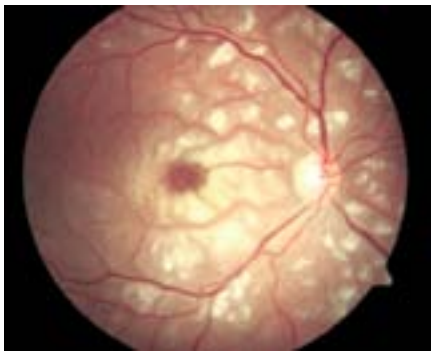


Fig. 12.77.1A: Purtscher's retinopathy—RE

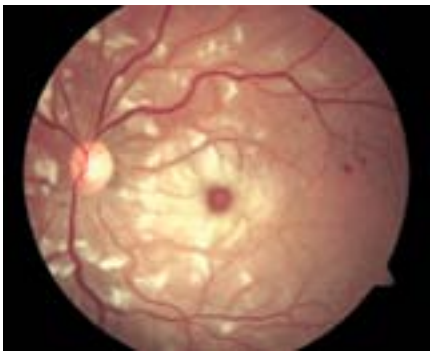


Fig. 12.77.1B: Purtscher's retinopathy—LE

Retinal Cysts

- May be subretinal or intraretinal cyst
- Intraretinal cysts form in case of old retinal detachment
- Subretinal cysts are caused by parasite like, subretinal *cysticercosis* **(Figs 12.78.1 and 12.78.2)**
- May cause subretinal fibrosis and secondary pigmentary changes **(Fig. 12.78.3)**.

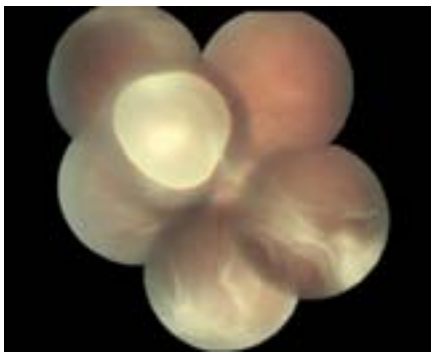


Fig. 12.78.1: Subretinal cysticercosis

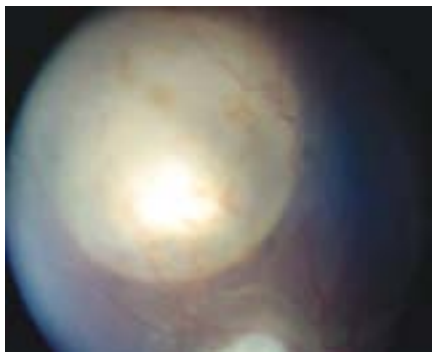


Fig. 12.78.2: Subretinal cysticercosis

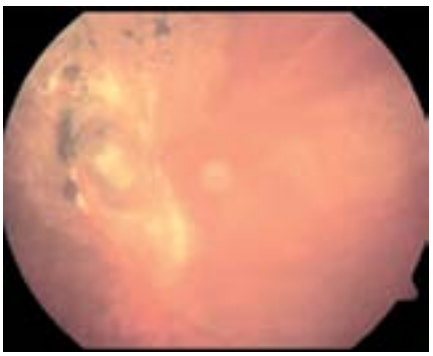


Fig. 12.78.3: Subretinal cysticercosis—fibrosis and TRD

CHAPTER

13 Diseases of the Lacrimal Apparatus

CHAPTER OUTLINE

Lacrimal Gland Diseases

- Acute dacryoadenitis
- Chronic dacryoadenitis

Epiphora

- Mechanical obstruction
- Lacrimal pump failure
- Chronic canaliculitis
- Occlusion of puncta or canaliculus

Dacryocystitis

- Acute (on chronic) dacryocystitis
- Chronic dacryocystitis
- Tumors of the lacrimal sac

Other Lacrimal Conditions

- Lacrimal fistula
- Accessory lacrimal punctum
- Accessory lobe of lacrimal gland
- Pyogenic granuloma of the lacrimal puncta
- Punctal burn following laser DCR
- Canalicular tear
- Hemangioma of the lacrimal punctum
- Centurion syndrome

Lacrimal Gland Diseases

Acute Dacryoadenitis

- Associated with systemic diseases like mumps, influenza or glandular fevers
- Characterized by unilateral or bilateral acute local pain, swelling and tenderness **(Figs 13.1.1 and 13.1.2)**
- Drooping of the outer part of upper lid with an S-shaped curve
- *Treatment:* Usually self-limiting, but systemic antibiotics and local hot compress are required in some cases.

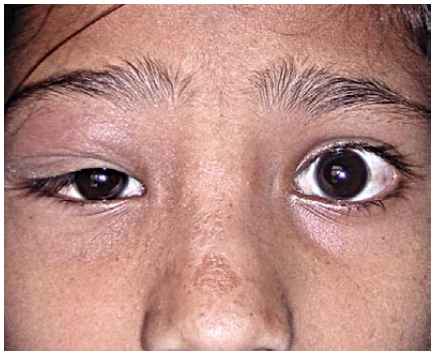


Fig. 13.1.1: Acute dacryoadenitis—RE

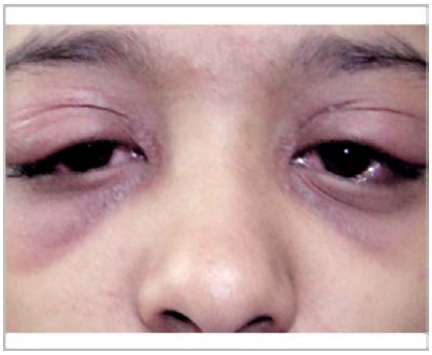


Fig. 13.1.2: Bilateral acute dacryoadenitis

Chronic Dacryoadenitis

- *Mikulicz syndrome:* Characterized by a symmetrical swelling of the lacrimal and salivary glands of both sides
  - May occur as a part of sarcoidosis, Hodgkin's lymphoma or one of the leukemias

Fig. 13.2.1A: Dacryops

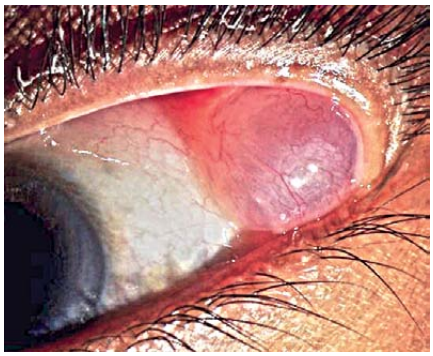


Fig. 13.2.1B: Dacryops

- *Dacryops:* Cystic swelling in the upper fornix due to retention of lacrimal gland secretion owing to blockage of one of the lacrimal ducts **(Figs 13.2.1A and B)**. Transillumination test is positive **(Fig. 13.2.2)**
- *Lacrimal glands tumors:*
  - Relatively rare and show a marked resemblance to the parotid gland tumors
  - Appears as hard, nodular and slightly mobile lumps
  - Proptosis is downwards and medially **(Figs 13.2.3A and B)**
  - *Benign mixed tumors* are the majority and have excellent prognosis **(Fig. 13.2.4)**
  - *Lacrimal gland carcinoma* has rapid onset with proptosis and involvement of lymph nodes **(Figs 13.2.5A to C)**. It needs radical surgery after needle biopsy or exenteration of the orbit.

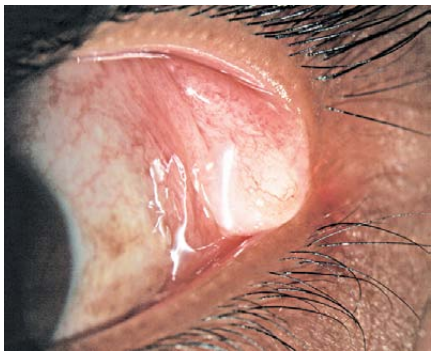


Fig. 13.2.2: Dacryops—positive transillumination test

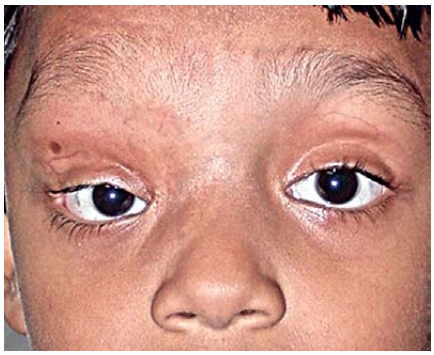


Fig. 13.2.3A: Lacrimal gland tumor—RE

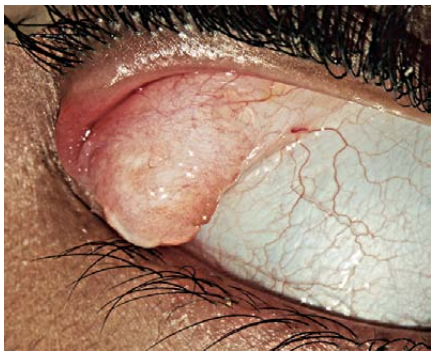


Fig. 13.2.3B: Lacrimal gland tumor visible after lid pulling

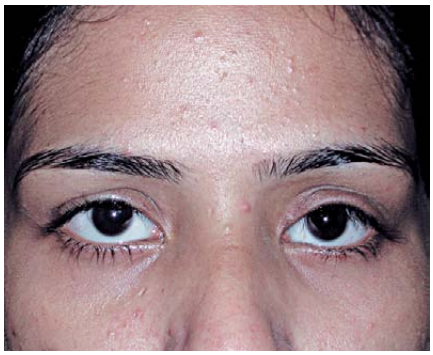


Fig. 13.2.4: Bilateral lacrimal gland tumor

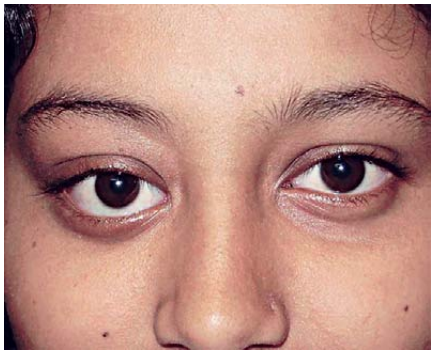


Fig. 13.2.5A: Adenocystic carcinoma of the lacrimal gland—right side

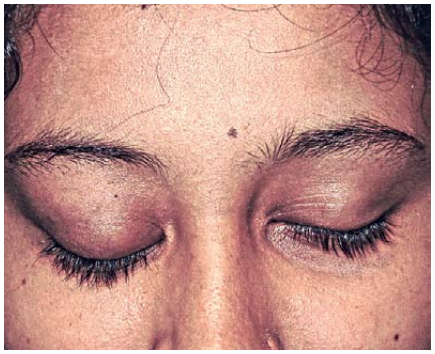


Fig. 13.2.5B: Adenocystic carcinoma of the lacrimal gland—right side

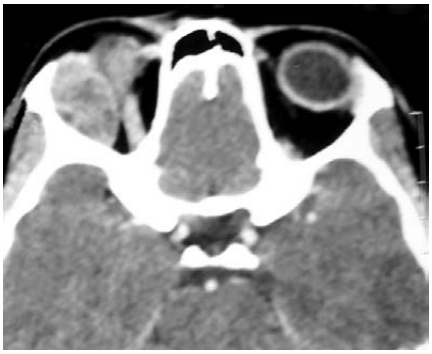


Fig. 13.2.5C: Adenocystic carcinoma of the lacrimal gland right side on CT scan

Epiphora

- Watering is associated with blurring of vision and continuous discomfort caused by tears running down the cheek
- In all cases it is necessary to differentiate it from *lacrimation* which is due to excessive secretion of tears
- *Epiphora* is due to defective drainage of tears.

Mechanical Obstruction

- Congenital absence or occlusion of puncta
- Punctal atresia or stenosis **(Figs 13.3.1A and B)**
- Foreign body in the puncta **(Fig. 13.3.2)**
- Malposition of puncta **(Figs 13.3.3 to 13.3.5)**

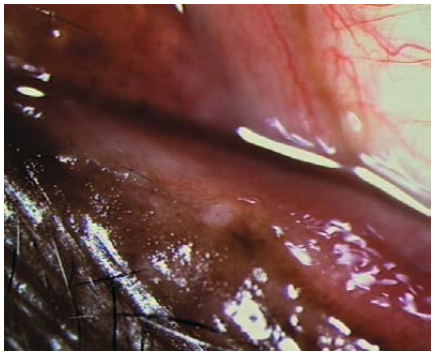


Fig. 13.3.1A: Punctal atresia

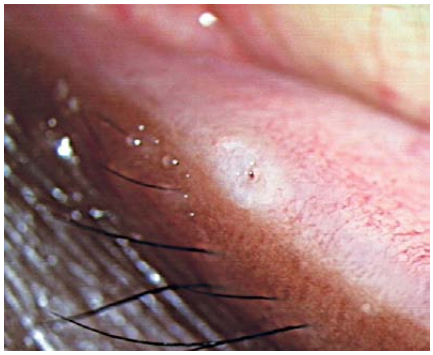


Fig. 13.3.1B: Punctal stenosis—IDU toxicity

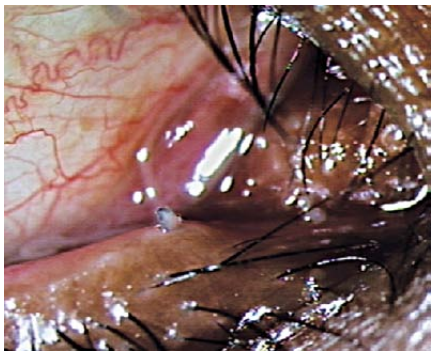


Fig 13.3.2: Eyelash in lower punctum

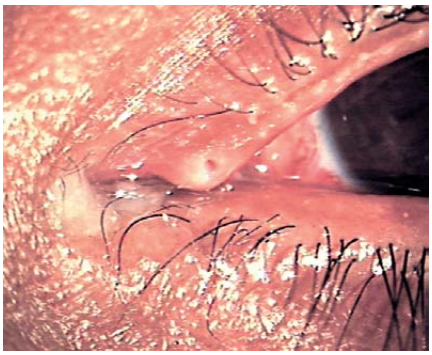


Fig. 13.3.3: Lacrimal punctal malposition —upper

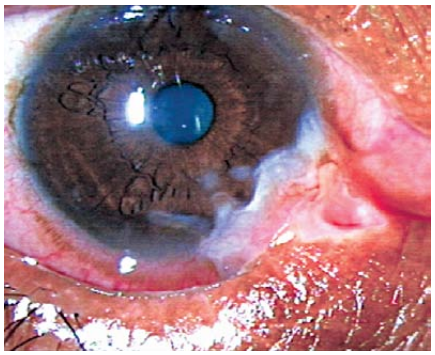


Fig. 13.3.4: Lacrimal punctal malposition—lower

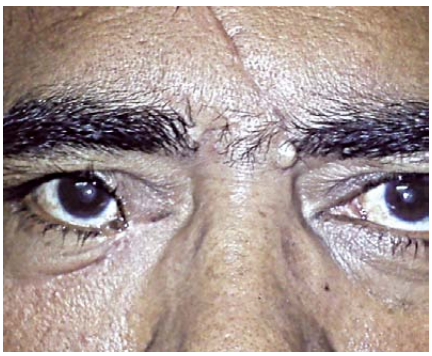


Fig. 13.3.5: Lacrimal punctal malposition—right lower lid

- Malapposition of puncta **(Figs 13.3.6A and B)**
- Congenital dacryocystitis
- Chronic dacryocystitis
- Neoplasm of the lacrimal sac
- Nasal pathology—like, polyps, tumors of inferior meatus of the nose.

Lacrimal Pump Failure

- Lower lid laxity
- Weakness of orbicularis, as in Bell's palsy **(Figs 13.4.1A and B)**
- Associated with lower lid ectropion and lagophthalmos.

Chronic Canaliculitis

- Commonly caused by *Actinomyces israelii* (streptothrix) **(Fig. 13.5.1)**
- Chronic infection causes—a localized swelling at the canalicular site **(Fig. 13.5.2)**, a pouting punctum with frank discharge and concretions within the canaliculi **(Figs 13.5.3 and 13.5.4)**

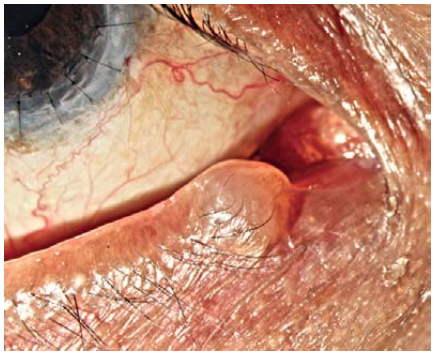


Fig. 13.3.6A: Lacrimal punctal malapposition—lower

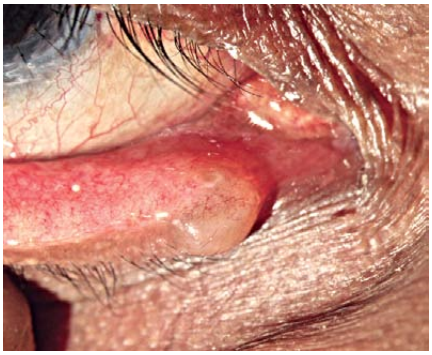


Fig. 13.3.6B: Lacrimal punctal malapposition—lower

Fig. 13.4.1A: Lacrimal pump failure—Bell's palsy—right side

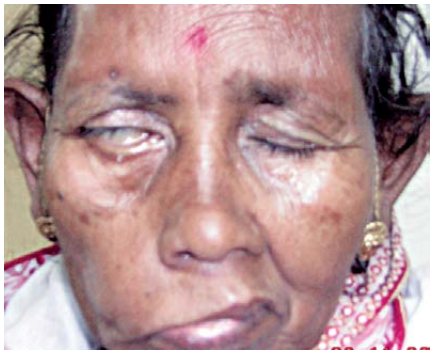


Fig. 13.4.1B: Lacrimal pump failure—Bell's palsy

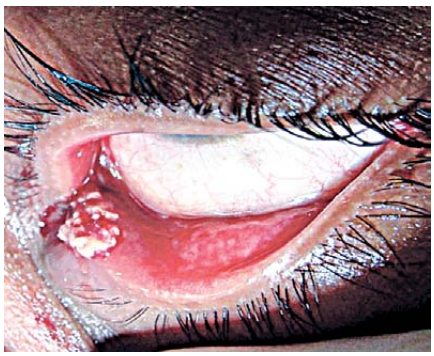


Fig. 13.5.1: Chronic canaliculitis—streptothrix

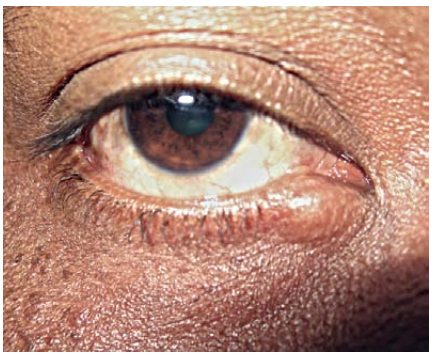


Fig. 13.5.2: Chronic canaliculitis

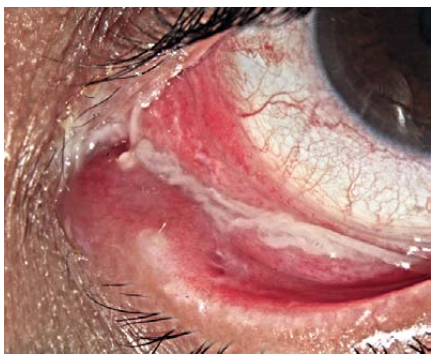


Fig. 13.5.3A: Acute canaliculitis—note a small foreign body

Fig. 13.5.3B: Acute canaliculitis—drainage of pus by pressure

- Chronic infection also causes canalicular abscess formation **(Fig. 13.5.5)**
- Sometimes with stone formation, called dacryolith
- *Treatment:* Linear incision to remove the dacryolith.

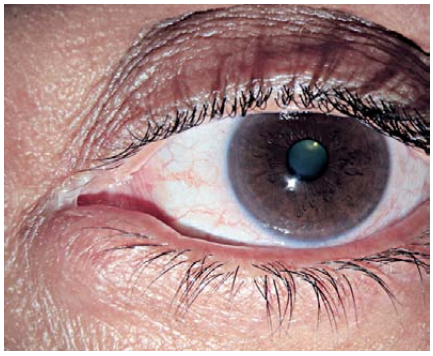


Fig. 13.5.4A: Chronic canaliculitis—small cystic swelling

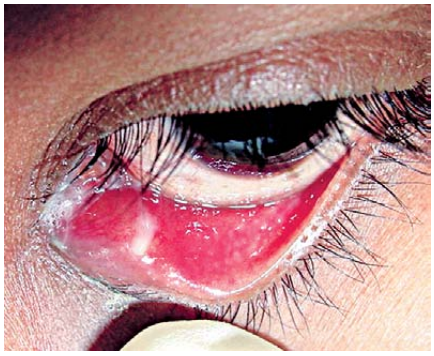


Fig. 13.5.4B: Chronic canaliculitis

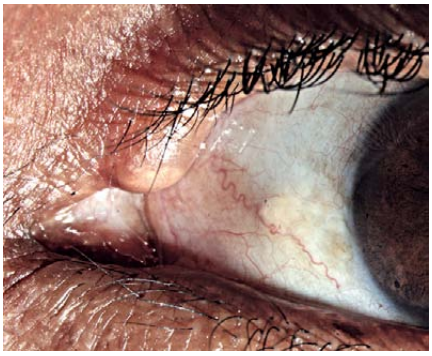


Fig. 13.5.5: Canalicular abscess formation—upper punctum

Occlusion of Puncta or Canaliculus

- Punctal stenosis or atresia as mentioned earlier **(*see* Figs 13.3.1A and B)**
- Foreign body in the punctum (eyelash —most common) **(Figs 13.6.1 and see Fig. 13.3.2)** or concretion
- Scarring of lower punctum **(Fig. 13.6.2)**
- *Drug induced:* Like prolonged use of IDU eye drops as mentioned earlier
- *Iatrogenic:* By punctal plugs for treating dry eye conditions **(Fig. 13.6.3)**.

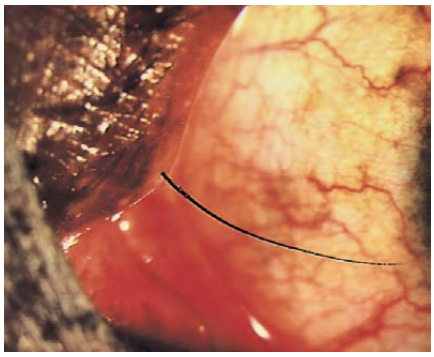


Fig. 13.6.1: Eyelash in upper punctum

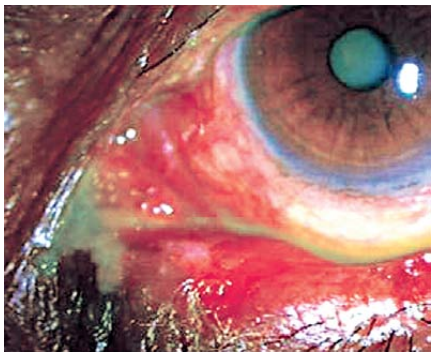


Fig. 13.6.2: Scarring of the lower punctum

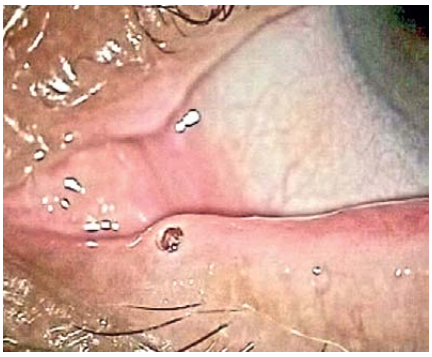


Fig. 13.6.3: Punctal plug—for dry eye treatment

Dacryocystitis

Acute (On Chronic) Dacryocystitis

- Suppurative inflammation of the lacrimal sac, with an associated cellulitis of the overlying tissues **(Figs 13.7.1 and 13.7.2)**

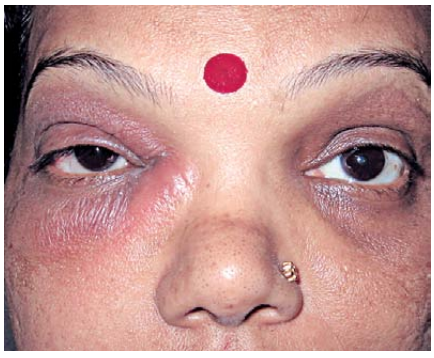


Fig. 13.7.1: Acute dacryocystitis in right side

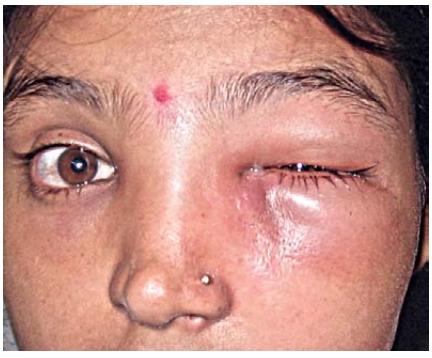


Fig. 13.7.2: Acute dacryocystitis with surrounding cellulitis on left side

- *Lacrimal abscess* **(Figs 13.7.3A and B)** which may often rupture spontaneouly with drainage of pus and scar formation **(Figs 13.7.4A and B)**
- May often with perforation of the skin just below medial palpebral ligament, leading to formation of *lacrimal fistula* **(Fig. 13.7.5)**
- Acute dacryocystitis is a very rare situation in case of congenital dacryocystitis **(Fig. 13.7.6)**
- *Treatment:* Hot compress, systemic antibiotics, drainage of abscess if necessary; and when the acute phase subsides, dacryocystorhinostomy (DCR) or dacryocystectomy (DCT) operation.

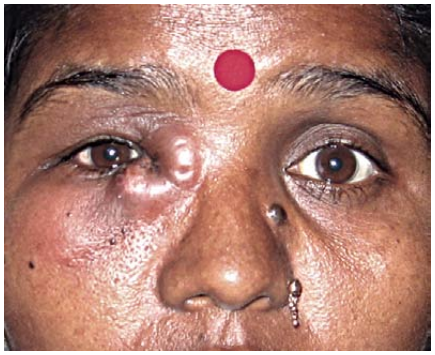


Fig. 13.7.3A: Lacrimal abscess

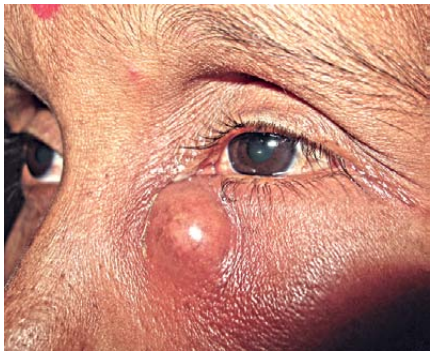


Fig. 13.7.3B: Lacrimal abscess

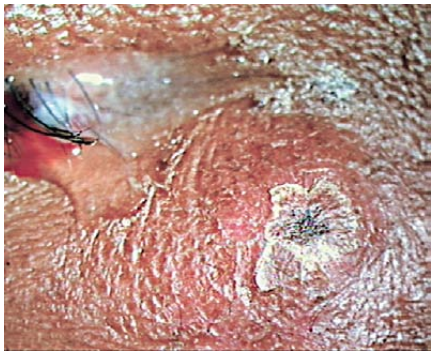


Fig. 13.7.4A: Lacrimal abscess

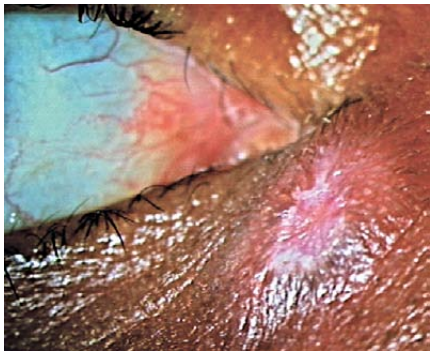


Fig. 13.7.4B: Lacrimal abscess—scar formation

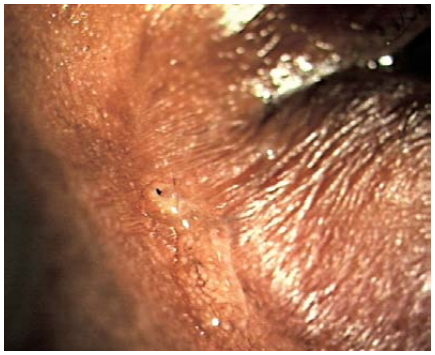


Fig. 13.7.5: Lacrimal fistula

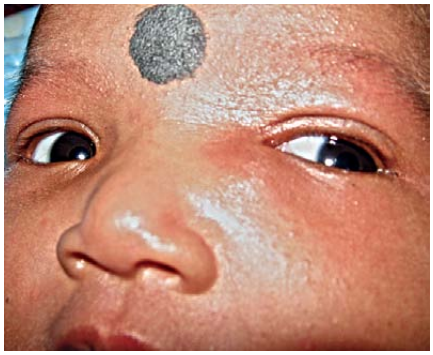


Fig. 13.7.6: Congenital dacryocystitis—acute

Chronic Dacryocystitis

- Most frequently in newborn infants and middle-aged women
- Chronic mucopurulent discharge at inner angle of the eye **(Fig. 13.8.1)**
- Regurgitation of the pus or mucopus through the puncta, on pressure over the sac region

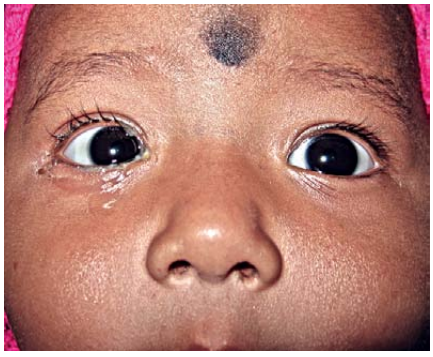


Fig. 13.8.1: Congenital dacryocystitis—right side

- In long-standing cases, there is an extraordinary dilatation and thinning of the lacrimal sac, called *mucocele* **(Figs 13.8.2 and 13.8.3)**, or *pyocele* **(Fig. 13.8.4)** of the lacrimal sac
- *Hypopyon corneal ulcer:* Mostly due to *pneumococci* **(Figs 13.8.5 and 13.8.6)**
- *Treatment:*

  Congenital cases
  - Hydrostatic sac massage and antibiotic eye drops
  - Probing and syringing usually at 10–12 months
  - If sac massage fails
  - If probing and syringing fails, then planned DCR operation under GA.

  Adult cases
  - Dacryocystorhinostomy (DCR) is the choice of operation; otherwise a dacryocystectomy (DCT) operation can be performed
  - *In case of corneal ulcer:* An emergency DCR or temporary punctal cautery or plugging is required.

Tumors of the Lacrimal Sac

- Tumors of the lacrimal sac are extremely rare and represent a potentially life-threatening condition.
- *Triad of lacrimal sac malignancy are:*
  - A mass below the medial palpebral ligament **(Fig. 13.9.1)**
  - A chronic dacryocystitis that irrigates freely
  - Regurgitation of bloody mucopus
- *Treatment:* In early cases—dacryocystectomy followed by radiation.

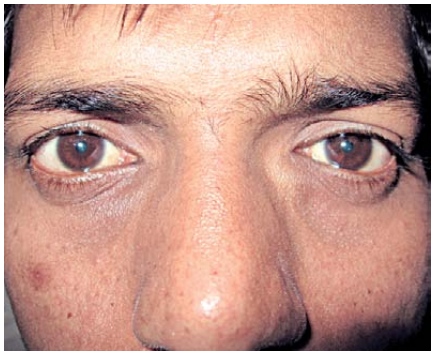


Fig. 13.8.2: Mucocele of lacrimal sac

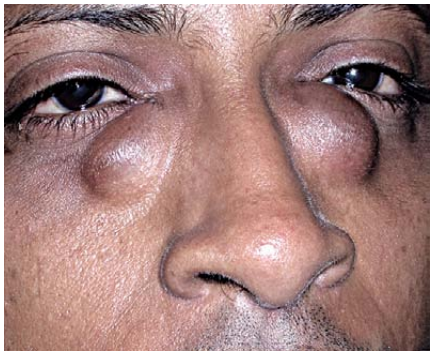


Fig. 13.8.3: Bilateral large mucocele of the lacrimal sac

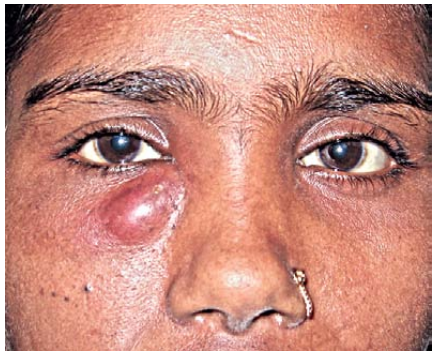


Fig. 13.8.4: Pyocele of the lacrimal sac

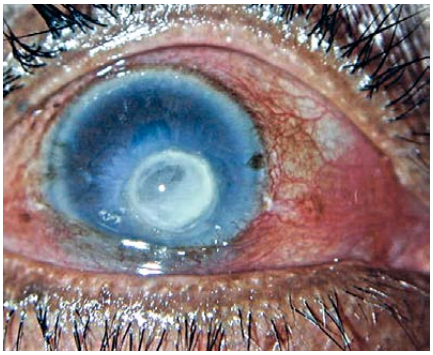


Fig. 13.8.5: Hypopyon corneal ulcer—the same eye in **Figure 3.7.4A**

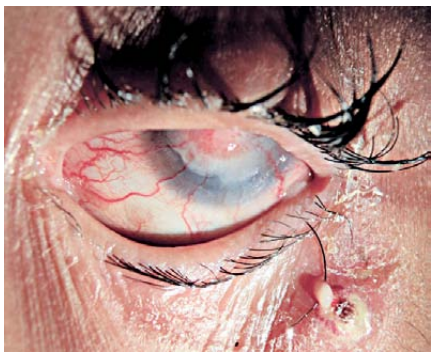


Fig. 13.8.6: Hypopyon corneal ulcer—note the lacrimal abscess

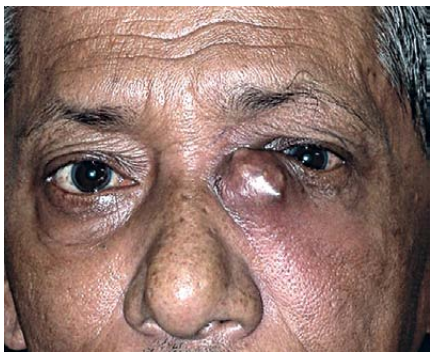


Fig. 13.9.1: Lacrimal sac carcinoma

Other Lacrimal Conditions

Lacrimal Fistula

- Rare, unilateral or bilateral condition
- May occur in congenital **(Figs 13.10.1 and 13.10.2)** or acquired **(Fig. 13.10.3)** form
- Causing watering from a separate opening on the skin just below the medial canthal ligament
- *Congenital form* usually does not cause problem, but rarely it may be associated with chronic dacryocystitis **(Fig. 13.10.4)**
- *Acquired from* had a history of acute dacryocystitis in the past
- *In congenital form:* Usually no treatment is required; rarely a DCR is required
- *In adult form:* Fistulectomy along with DCR or DCT.

Fig. 13.10.1: Congenital lacrimal fistula

Fig. 13.10.2: Congenital lacrimal fistula

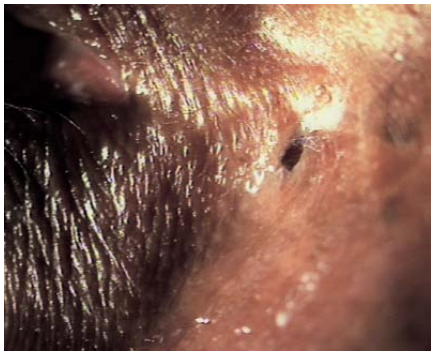


Fig. 13.10.3: Acquired lacrimal fistula

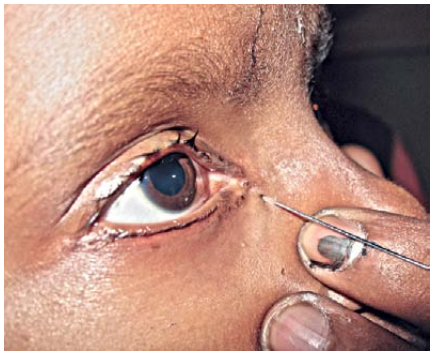


Fig. 13.10.4: Congenital lacrimal fistula with chronic dacryocystitis—note the discharge

Accessory Lacrimal Punctum

- Rare, congenital **(Fig. 13.11.1)** or acquired **(Fig. 13.11.2)** condition
- An accessory lacrimal punctum is seen medial to the original punctum of lower lid
- In acquired cases it may be due to trauma
- No treatment is required.

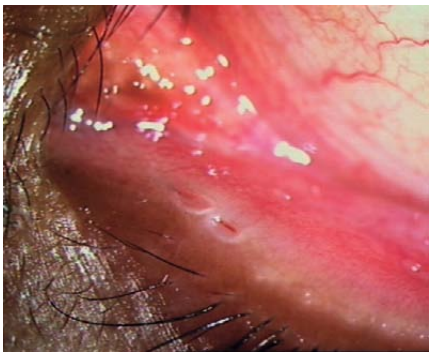


Fig. 13.11.1: Accessory lacrimal punctum—congenital

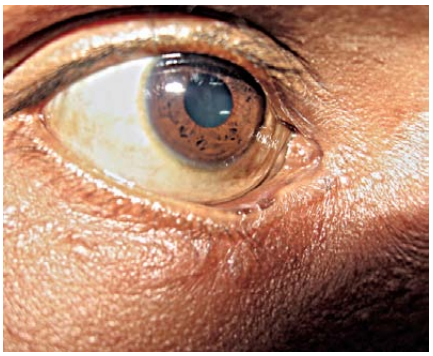


Fig. 13.11.2: Accessory lacrimal punctum—acquired

Accessory Lobe of Lacrimal Gland

- Visible sometimes at the superolateral fornix **(Figs 13.12.1 and 13.12.2)**
- To be differentiated from dermolipoma **(Fig. 13.12.3)** or orbital fat prolapse **(Figs 13.12.4A and B)**
- Does not require any treatment.

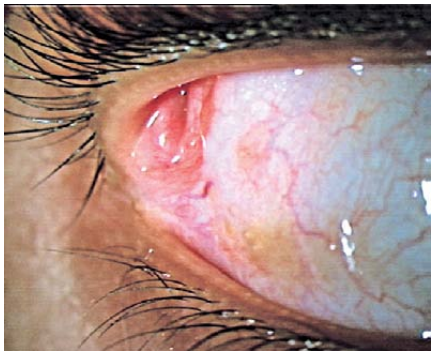


Fig. 13.12.1: Accessory lacrimal gland lobe

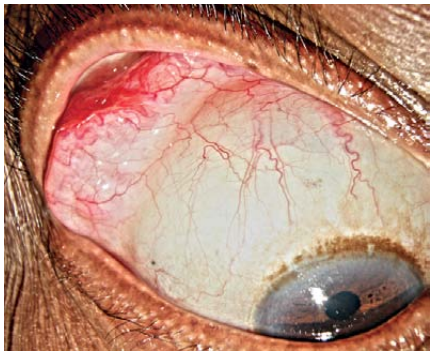


Fig. 13.12.2: Accessory lacrimal gland lobe

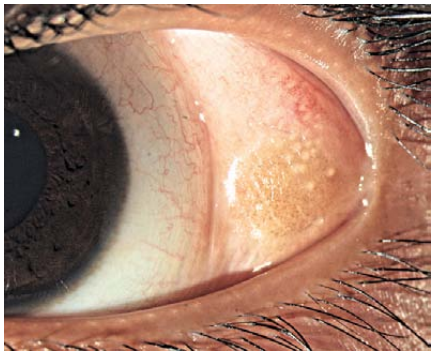


Fig. 13.12.3: Dermolipoma

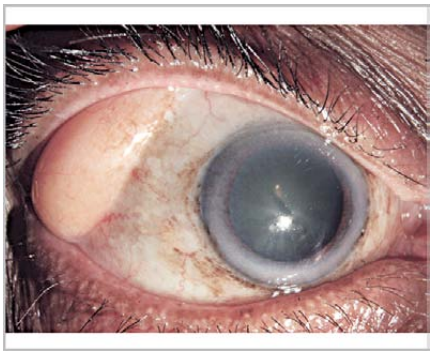


Fig. 13.12.4A: Fat prolapse—RE

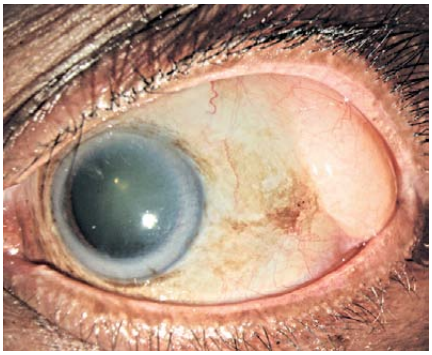


Fig. 13.12.4B: Fat prolapse—LE

Pyogenic Granuloma of the Lacrimal Puncta

- Usually happens as a complication of punctal plugging; after silicone plug or smart plug **(Fig. 13.13.1)**
- Very rarely, after an attack of acute dacryocystitis, canaliculitis or transcanalicular laser DCR operation **(Fig. 13.13.2)**
- *Treatment:* Antibiotics; excision of the granuloma and further management depending upon the situation.

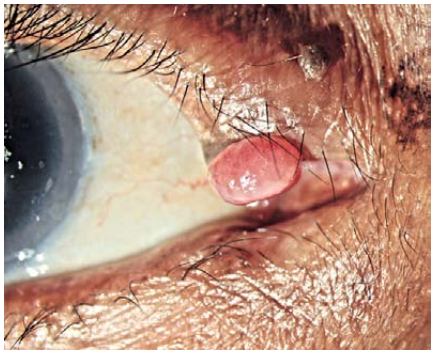


Fig. 13.13.1: Pyogenic granuloma—arising from upper puncta

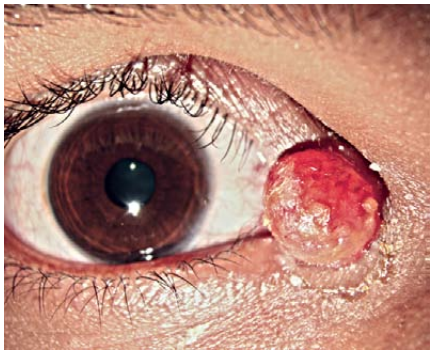


Fig. 13.13.2: Pyogenic granuloma—arising from lower puncta

Punctal Burn Following Laser DCR

- One of the complications of transcanalicular laser DCR
- May affect lower punctum or both punctum **(Figs 13.14.1 and 13.14.2)**
- Scarring occurs in late stage.

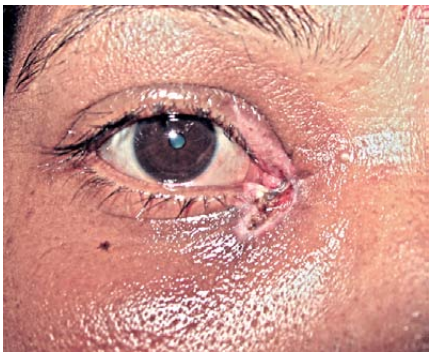


Fig. 13.14.1: Punctal burn following laser DCR

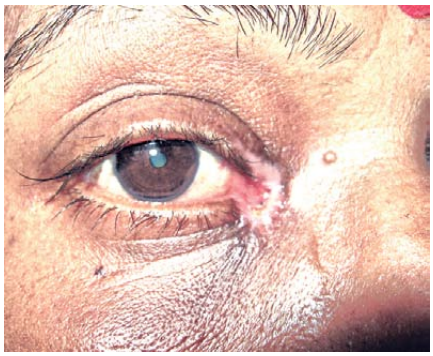


Fig. 13.14.2: Punctal burn following laser DCR

Canalicular Tear

- Rare, usually lower canaliculus is affected **(Fig. 13.15.1)**
- Needs urgent and meticulous surgical repair to preserve the canalicular patency.

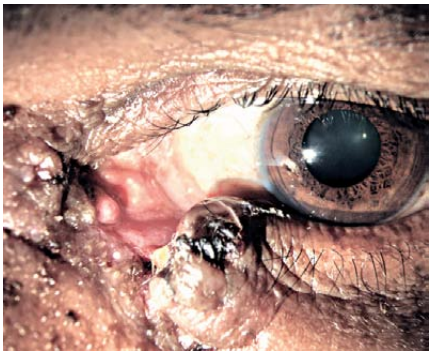


Fig. 13.15.1: Canalicular tear—lower

Hemangioma of the Lacrimal Punctum

- Rare, may cause epiphora **(Fig. 13.16.1)**
- Needs excision followed by biopsy.

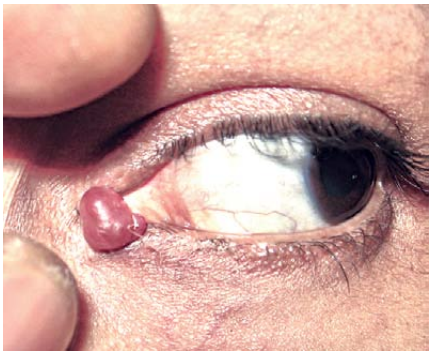


Fig. 13.16.1: Hemangioma lower punctum

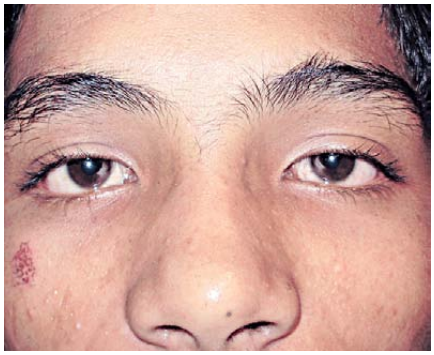


Fig. 13.17.1: Centurion syndrome

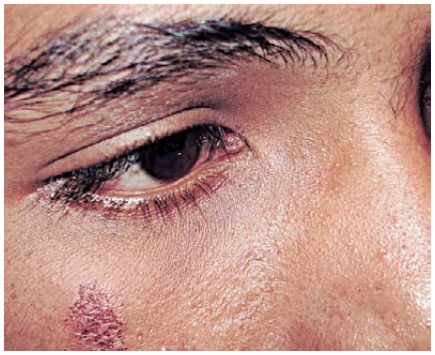


Fig. 13.17.2A: Centurion syndrome—RE

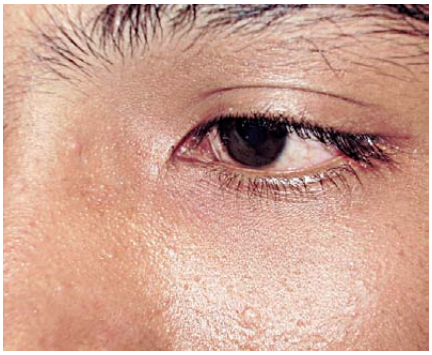


Fig. 13.17.2B: Centurion syndrome—LE

Centurion Syndrome

- Centurion syndrome is a unique clinical entity that may manifest as epiphora during puberty
- *The anomaly consists of:*
  - An anterior displacement of the anterior limb of the medial canthal tendon
  - A prominent nasal bridge **(Fig. 13.17.1)**
  - With lid malposition away from the globe
  - Displacement of the lacrimal puncta out of the tear lake **(Figs 13.17.2A and B)**
- Syringing shows a patent NLD with delayed dye disappearance time
- *Treatment:* Surgery to restore lid globe apposition medially by release of the anterior limb of the medial canthal tendon with or without conjunctivoplasty or lower lid retractor plication.

CHAPTER

14 Diseases of the Orbit

CHAPTER OUTLINE

Congenital/Developmental Bony Anomalies

- Craniosynostosis
- Orbital dysplasia
- Anophthalmos or extreme microphthalmos
- Cryptophthalmos
- Orbitopalpebral (colobomatous) cyst

Proptosis

- Pseudoproptosis
- Enophthalmos

Proptosis in Children

- Craniosynostosis (shallow orbit)
- Orbitopalpebral (colobomatous) cyst
- Capillary hemangioma
- Preseptal and orbital cellulitis
- Lymphangioma
- Glioma of the optic nerve
- Rhabdomyosarcoma
- Orbital extension of retinoblastoma
- Acute leukemia

Other Orbital Tumors in Children

Proptosis in Adults

- Inflammatory orbital diseases (orbital pseudotumor)
- Orbital cellulitis in adults
- Thyroid associated ophthalmopathy
- Cavernous hemangioma
- Orbital varix
- Meningioma of the optic nerve
- Carotid cavernous fistula
- Cavernous sinus thrombosis
- Other causes of proptosis in adults

Miscellaneous Orbital Lesions

- Orbital rim lesions
- Luxatio bulbi
- Orbital fat prolapse
- Contracted socket

Congenital/Developmental Bony Anomalies

Craniosynostosis

- Follows premature closure of one or more cranial sutures
- Thereby, a complete arrest of bone growth perpendicular to the closed suture
- Compensatory growth of the cranium in other diameters which causes the typical shape of the skull
- *Common features are*:
  - Bilateral proptosis due to shallow orbit
  - Esotropia or exotropia
  - Chemosis of the conjunctiva, corneal exposure
  - Papilledema due to increased ICT
  - Optic atrophy
- *Treatment:* By craniotomy or orbital decompression to reduce CSF pressure and papilledema.
- ***Craniofacial dysostosis (Crouzon)***
  Brachycephaly (clover-leaf skull): Premature closure of all sutures
  - Combined with hypoplasia of the maxilla **(Figs 14.1.1A and B)**
  - Often hereditary in nature **(Fig. 14.1.2).**

  Ophthalmic features **(Fig. 14.1.3)**
  - Widely separated eyeballs (hypertelorism)
  - Shallow orbits with proptosis
  - Conjunctival chemosis
  - Corneal problems due to exposure
  - Divergent squint
  - Optic atrophy
- ***Mandibulofacial dysostosis (Treacher-Collin)***
  - Hypoplasia of the zygoma and mandible
  - Indistinct inferior orbital margin
  - Coloboma (notching) of the lower lid **(Fig. 14.1.4)**
  - Antimongoloid slanting
- ***Median facial-cleft syndrome***
  - Hypertelorism with telecanthus
  - Divergent squint
  - Cleft nose, lip and palate **(Figs 14.1.5 to 14.1.7)**
  - Coloboma of the lids
  - V-shaped frontal hairline (widow's peak)
  - Often with hypoplasia of maxilla **(Fig. 14.1.8)**

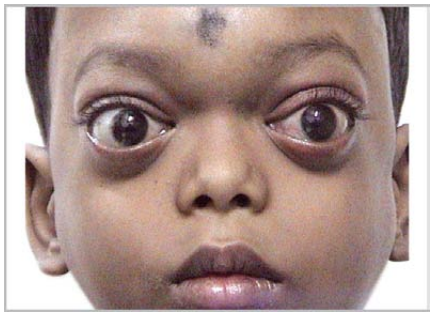


Fig. 14.1.1A: Crouzon's syndrome

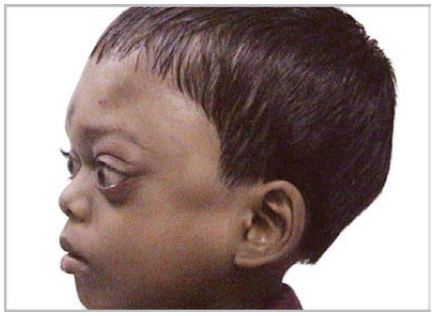


Fig. 14.1.1B: Crouzon's syndrome—on profile view

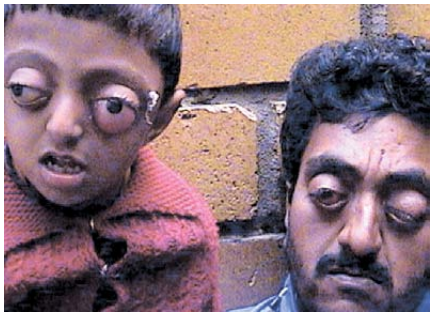


Fig. 14.1.2: Crouzon's syndrome—father and son

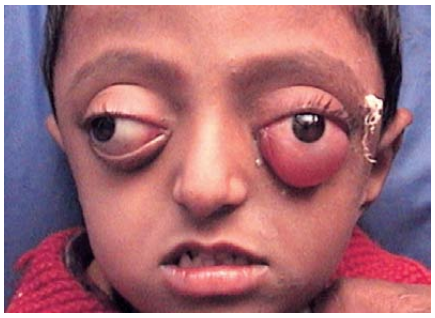


Fig. 14.1.3: Crouzon's syndrome—proptosis and chemosis of the conjunctiva

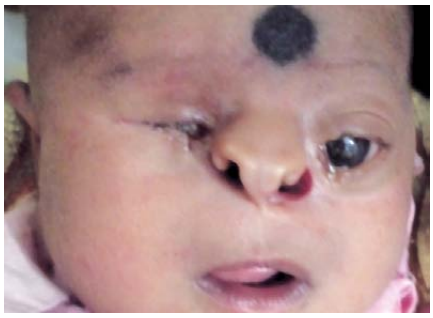


Fig. 14.1.4: Treacher-Collin syndrome

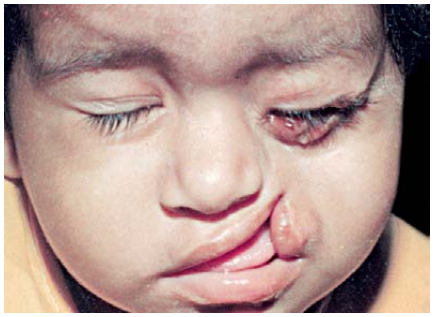


Fig. 14.1.5: Median facial cleft syndrome

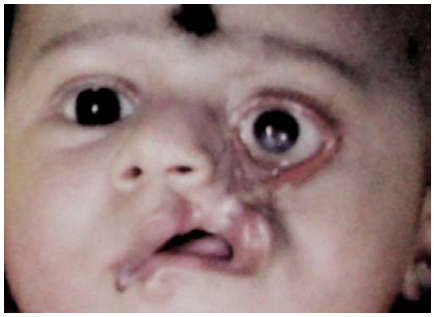


Fig. 14.1.6: Median facial cleft syndrome—coloboma of lid

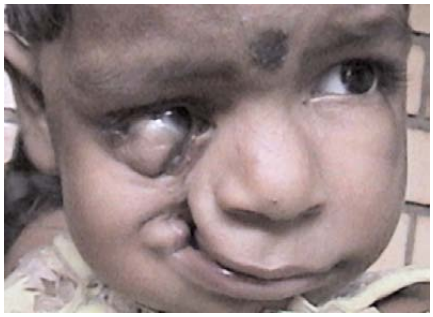


Fig. 14.1.7: Median facial cleft syndrome—coloboma of lid

- ***Oxycephaly-syndactyle (Apert)***
  - Tower skull with flat occiput
  - Hypertelorism, shallow orbits and proptosis **(Fig. 14.1.9)**
  - Syndactyle
  - Mental retardation
- ***Hypertelorism***
  - Increased separation of the eyes
  - Separated orbits, and broad nasal bridge **(Fig. 14.1.10)**
  - Divergent squint, telecanthus and antimongoloid slanting
  - Should be differentiated from *true telecanthus* **(Fig. 14.1.11)**.

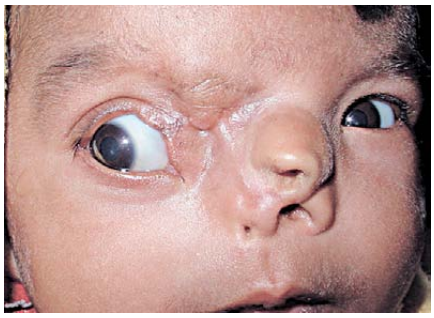


Fig. 14.1.8: Median facial cleft syndrome—maxillary hypoplasia

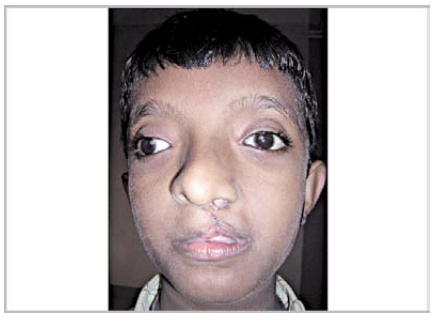


Fig. 14.1.9: Oxycephaly—hypertelorism, repaired cleft lip

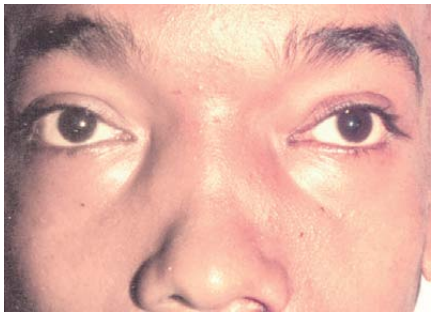


Fig. 14.1.10: Hypertelorism

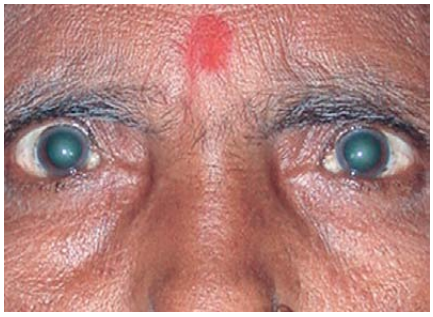


Fig. 14.1.11: Telecanthus

Orbital Dysplasia

- Asymmetry of levels of orbital margins, especially the inferior **(Figs 14.2.1 and 14.2.2)**
- May be associated with hypertelorism, hemifacial microsomia, Goldenhar's syndrome **(Fig. 14.2.3)**, etc.

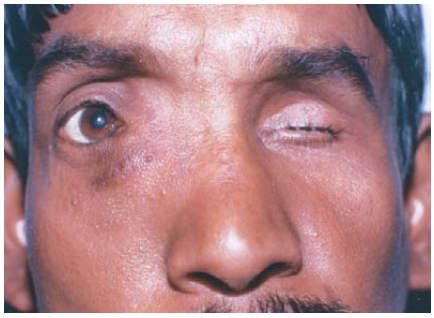


Fig. 14.2.1: Orbital dysplasia

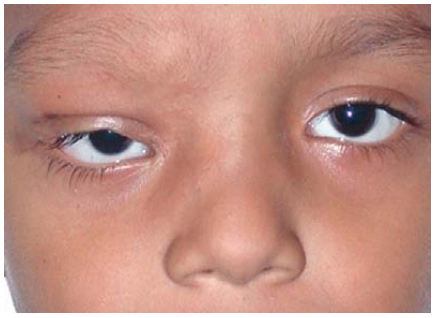


Fig. 14.2.2: Orbital dysplasia

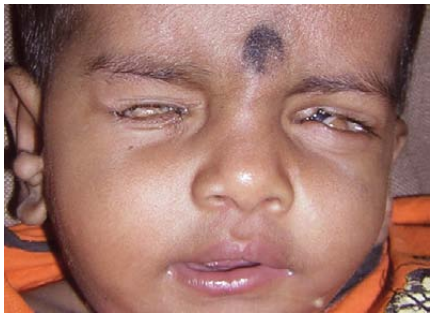


Fig. 14.2.3: Orbital dysplasia—Goldenhar's syndrome

Anophthalmos or Extreme Microphthalmos

- Under development of bony orbit due to extreme microphthalmos **(Fig. 14.3.1)** or anophthalmos **(Fig. 14.3.2)**
- Facial asymmetry is common
- Features of anophthalmic socket
- *Oculodigital sign* (a blind child with eye-poking sign) may be present **(Fig. 14.3.3)**
- To be differentiated from acquired anophthalmos **(Fig. 14.3.4)**.

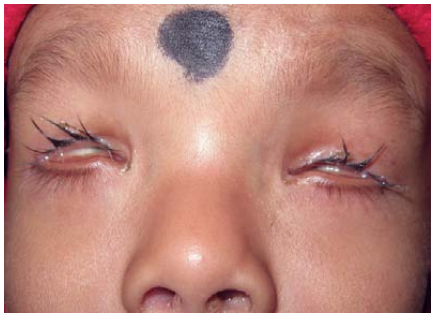


Fig. 14.3.1: Clinical anophthalmic socket

Fig. 14.3.2: Anophthalmic socket—clinical anophthalmos

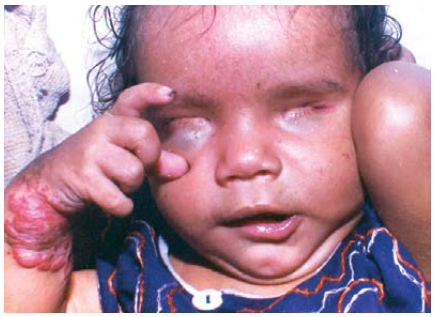


Fig. 14.3.3: Anophthalmic socket—clinical anophthalmos—oculodigital sign

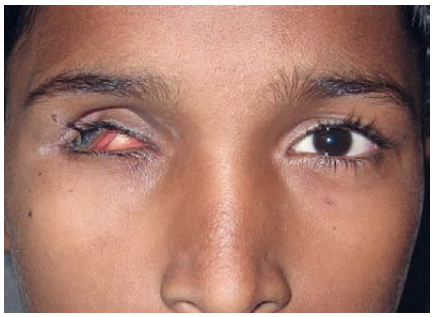


Fig. 14.3.4: Anophthalmic socket—surgical

Cryptophthalmos

- Microphthalmos sometimes associated with an extremely rare condition called cryptophthalmos
- Hidden eyeball behind the non-formed eyelids **(Fig. 14.3.5)**
- No conjunctival sac
- Malformed globe or orbit
- Facial asymmetry with or withour cleft lips and cleft palate **(Fig. 14.3.6)**.

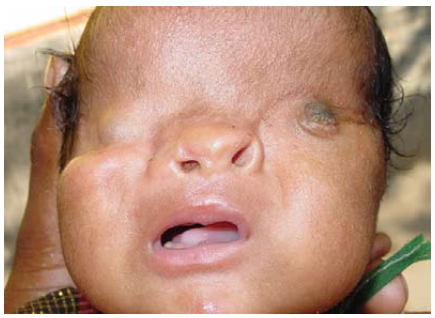


Fig. 14.3.5: Cryptophthalmos

Fig. 14.3.6: Cryptophthalmos with median cleft syndrome

Orbitopalpebral (Colobomatous) Cyst

- Usually associated with microphthalmos with coloboma **(Figs 14.4.1A and B)**

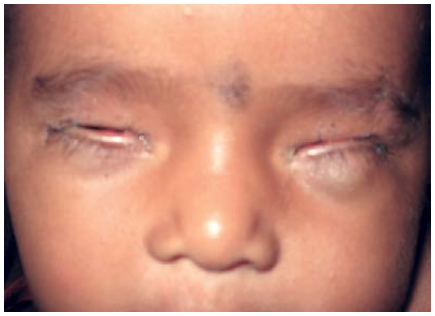


Fig. 14.4.1A: Orbitopalpebral cyst—left side

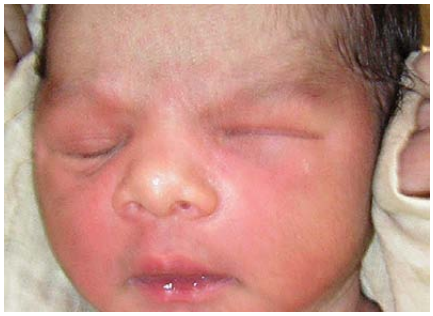


Fig. 14.4.1B: Orbitopalpebral cyst—left side

- May be with normal but deformed globe **(Fig. 14.4.2)**
- Classical site of the cyst is inferonasal, which corresponds to fetal fissure
- Bluish in color with upward displacement of eyeball **(Fig. 14.4.3)**
- *Congenital cystic eyeball* is another variation of congenital developmental defect of the whole eyeball **(Fig. 14.4.4)**.

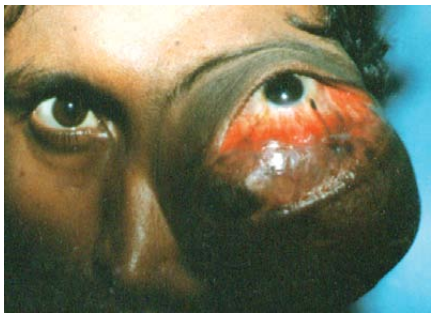


Fig. 14.4.2: Orbitopalpebral cyst—large

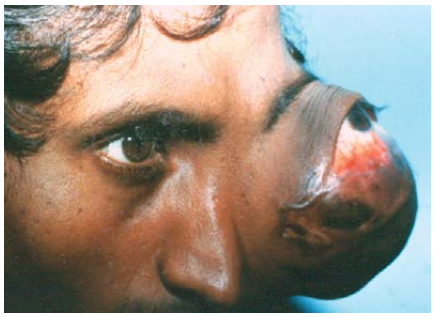


Fig. 14.4.3: Orbitopalpebral cyst—large

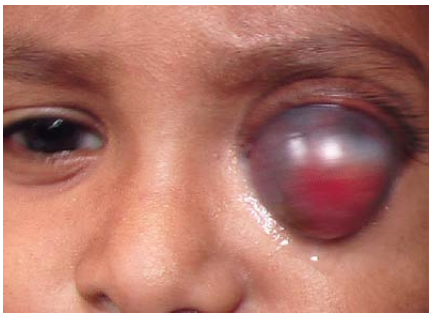


Fig. 14.4.4: Congenital cystic eyeball

Proptosis

- Forward protrusion of the eyeball much beyond the orbital margin **(Figs 14.5.1A to D)**
- It may be dynamic, called *exophthalmos* (as in thyroid ophthalmopathy) or passive, called *proptosis* by pushing by a retro-orbital mass
- Most readily assessed clinically by standing behind and viewing the eyes from above **(Figs 14.5.2A and B)**
- Usually measured by Hertel's exophthalmometer or by a plastic ruler
  - *Normal value:* Less than 20 mm (average = 16 mm)
  - *In proptosis:* 21 mm or more or a difference >2 mm between two eyes
- Rule out the possibilities of *pseudoproptosis* and *enophthalmos*.

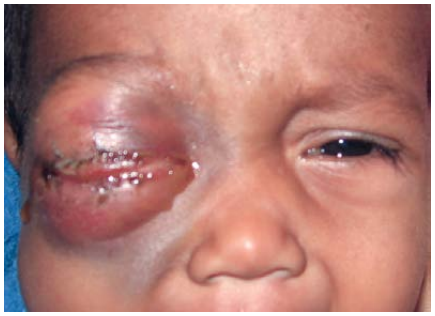


Fig. 14.5.1A: Proptosis in a child

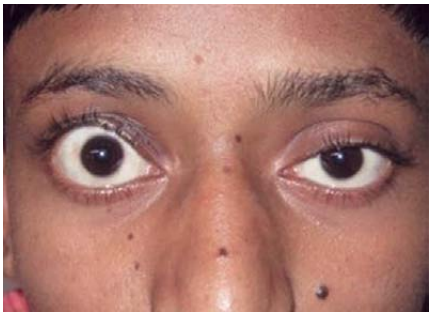


Fig. 14.5.1B: Proptosis in young adult

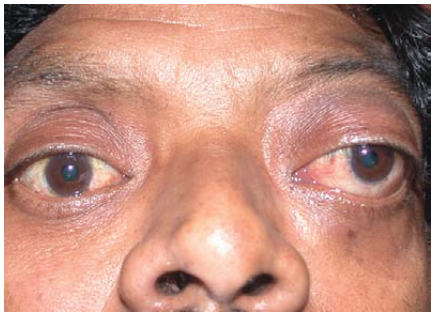


Fig. 14.5.1C: Proptosis in adult

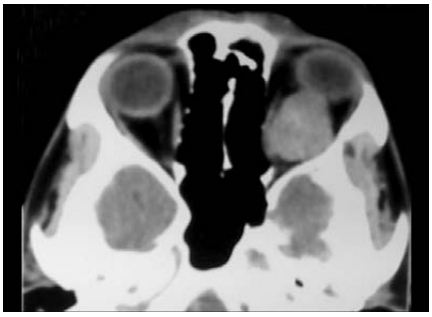


Fig. 14.5.1D: Proptosis—on CT scan

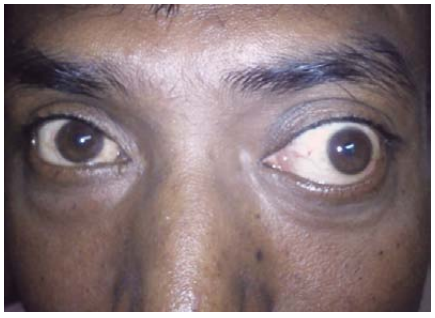


Fig. 14.5.2A: Proptosis—LE

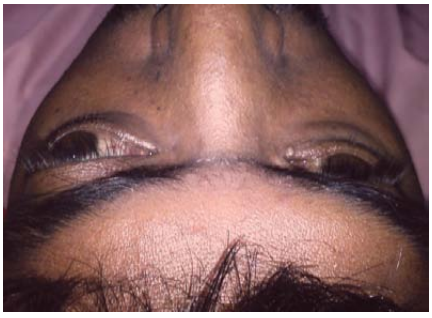


Fig. 14.5.2B: Proptosis—viewing from above—LE

Pseudoproptosis

- One eye may be larger than the fellow eye
- *Causes:*
  - Unilateral high axial myopia **(Fig. 14.6.1)**
  - Unilateral buphthalmos **(Fig. 14.6.2)**
  - Pseudocornea or anterior staphyloma **(Figs 14.6.3 to 14.6.5)**
  - Retraction of the eyelid of one eye **(Fig. 14.6.6)**
  - Enophthalmos of the opposite eye **(Figs 14.6.7A and B)**.

Fig. 14.6.1: Pseudoproptosis—high myopia (LE)—note the CL

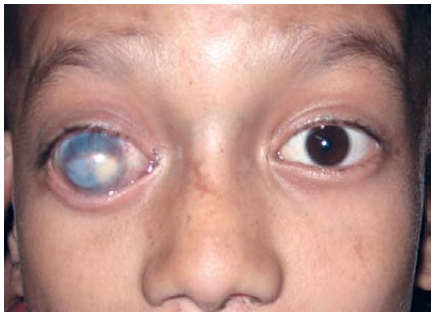


Fig. 14.6.2: Pseudoproptosis—buphthalmos—LE

Fig. 14.6.3: Pseudoproptosis—pseudocornea

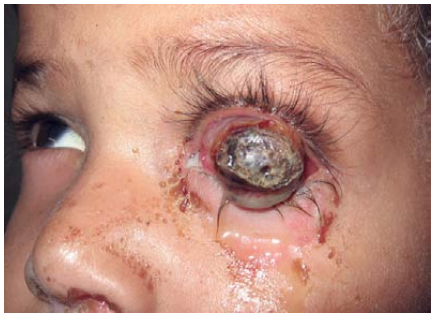


Fig. 14.6.4A: Pseudoproptosis—total corneal perforation—autoevisceration

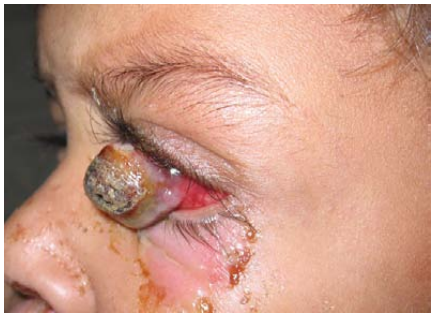


Fig. 14.6.4B: Pseudoproptosis—total corneal perforation—autoevisceration

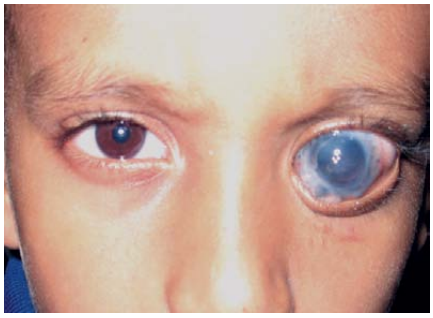


Fig. 14.6.5: Pseudoproptosis—anterior staphyloma

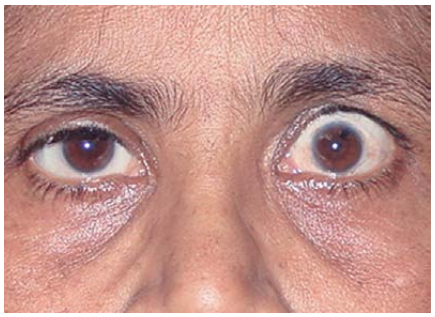


Fig. 14.6.6: Pseudoproptosis—lid retraction—LE

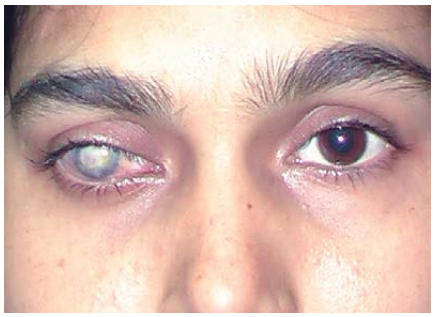


Fig. 14.6.7A: Pseudoproptosis—enophthalmos—RE

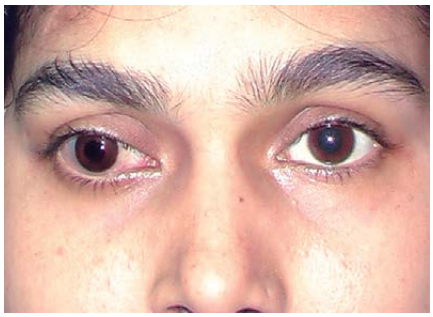


Fig. 14.6.7B: Enophthalmos—after cosmetic CL

Enophthalmos

- A condition in which eyeball is recessed within the orbit
- About 25% of patients with enophthalmos are initially misdiagnosed as having contralateral proptosis or ipsilateral ptosis
- *Causes:*
  - Microphthalmos or anophthalmos **(Fig. 14.7.1)**
  - Phthisis bulbi, unilateral or bilateral **(Figs 14.7.2 to 14.7.4)**
  - Blowout fracture ***(see* Fig. 16.12.1)**
  - Horner's syndrome.

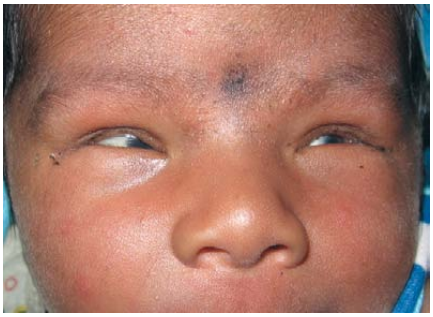


Fig. 14.7.1: Enophthalmos—microphthalmos

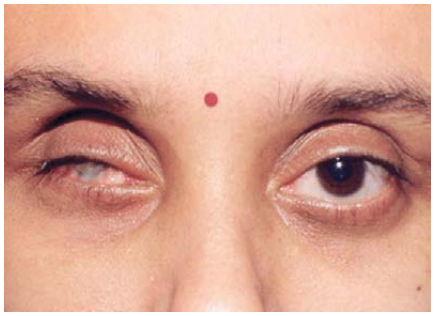


Fig. 14.7.2A: Enophthalmos—phthisis bulbi

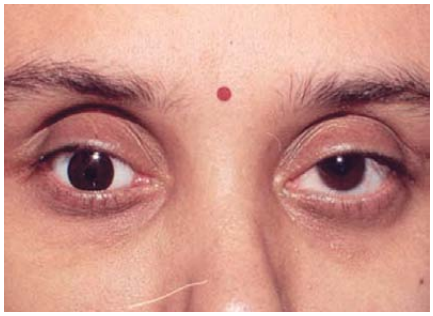


Fig. 14.7.2B: Enophthalmos—phthisis bulbi—after cosmetic scleral shell in RE

Fig. 14.7.3: Bilateral enophthalmos—phthisis bulbi

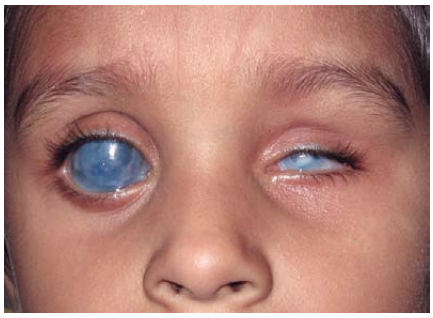


Fig. 14.7.4: Pseudoproptosis (RE) and enophthalmos (LE)—vitamin A deficiency

Proptosis in Children

Craniosynostosis (Shallow Orbit)

- *Already discussed earlier.*

Orbitopalpebral (Colobomatous) Cyst

- *Already discussed earlier.*

Capillary Hemangioma

- Present at birth or early infancy **(Figs 14.8.1 and 14.8.2)**

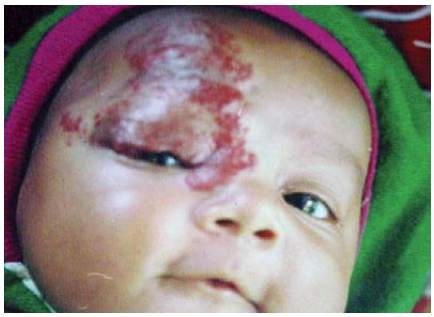


Fig. 14.8.1: Capillary hemangioma

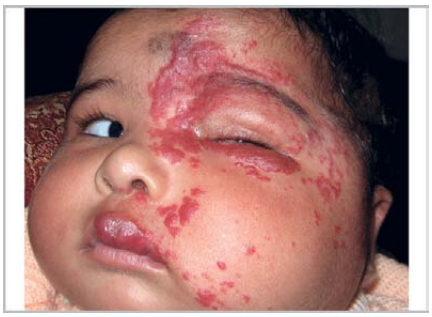


Fig. 14.8.2: Capillary hemangioma

- Slowly growing unilateral proptosis due to hemangiomatous mass in the upper or lower anterior orbit **(Figs 14.8.3 and 14.8.4)**
- Becomes engorged, and may enlarge when the baby cries
- May have similar lesion in eyelids or elsewhere.

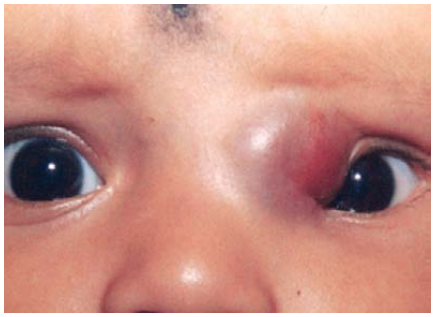


Fig. 14.8.3: Cavernous hemangioma

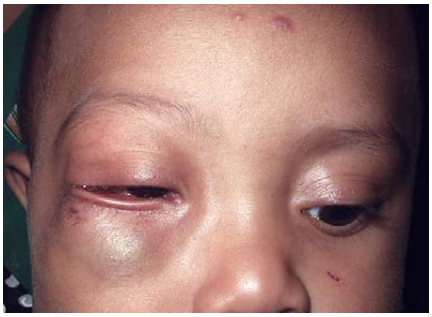


Fig. 14.8.4: Cavernous hemangioma

Preseptal and Orbital Cellulitis

- Usually child or young adult
- Ethmoidal sinusitis—is the most common cause
- ***Preseptal cellulitis:***
  - Acute periorbital swelling and redness **(Fig. 14.9.1)**
  - Conjunctival chemosis
  - Fluctuating mass signifies abscess formation **(Fig. 14.9.2)**
- *Treatment:* Oral antibiotics and analgesics, hot compress, and topical antibiotics; drainage of lid abscess if necessary.
- ***Orbital cellulitis:***
  - Very sick child with rise in temperature **(Fig. 14.9.3)**
  - Sudden onset, rapidly growing unilateral proptosis with severe pain and chemosis of lids **(Fig. 14.9.4)**
  - Severe chemosis and congestion of conjunctiva
  - Eyeball is displaced laterally and downwards **(Figs 14.9.5A and B)**
  - Diplopia due to limitation of ocular movements
- *Treatment:* Systemic intravenous antibiotics; analgesics, hot compress, topical antibiotics and drainage of orbital abscess if necessary.

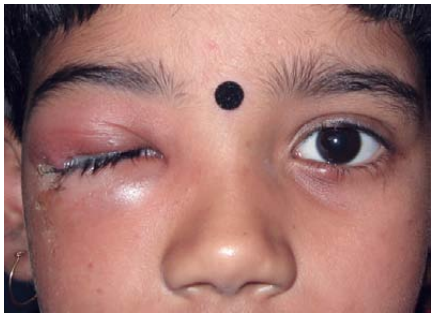


Fig. 14.9.1: Preseptal cellulitis

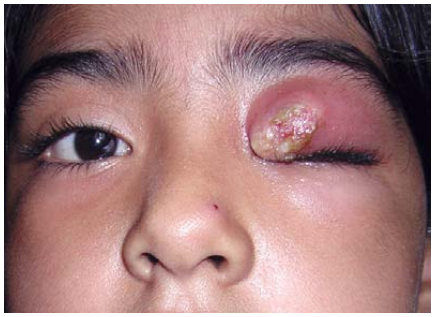


Fig. 14.9.2: Preseptal cellulitis

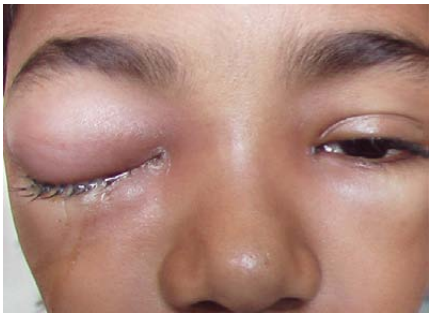


Fig. 14.9.3: Orbital cellulitis—sick child

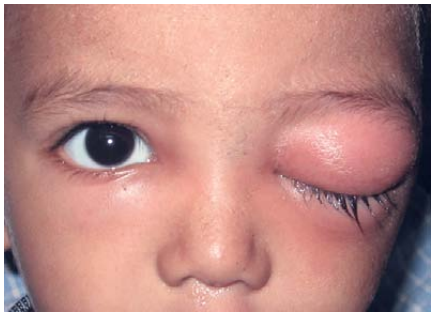


Fig. 14.9.4: Orbital cellulitis in a child

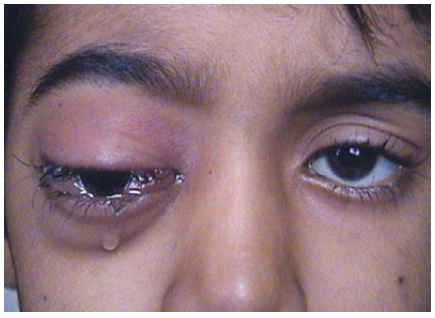


Fig. 14.9.5A: Orbital cellulitis—eyeball displaced downwards

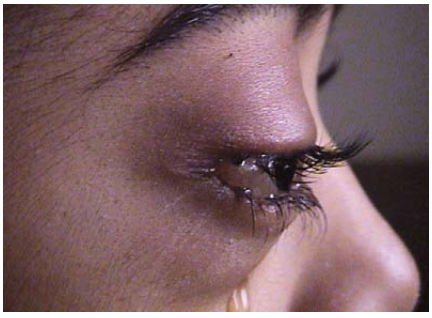


Fig. 14.9.5B: Orbital cellulitis—chemosis

Lymphangioma

- Benign tumor between 1 to 15 years
- May occur in isolation or be combined with lid or conjunctival lesions
- Soft bluish mass mainly in the superior orbit, which may remain stationary **(Fig. 14.10.1)**
- May also involve the sinuses or oropharynx **(Fig. 14.10.2)**
- Periodic sudden enlargement with upper respiratory tract infection or with spontaneous bleeding (chocolate cyst)
- Visual and motility disturbances are common
- May be simultaneously associated with lid or orbital hemangioma—lymphohemangioma **(Figs 14.10.3A and B)**.

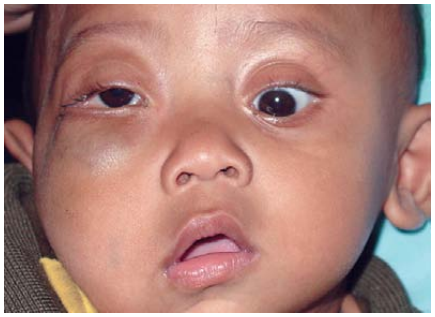


Fig. 14.10.1: Lymphangioma

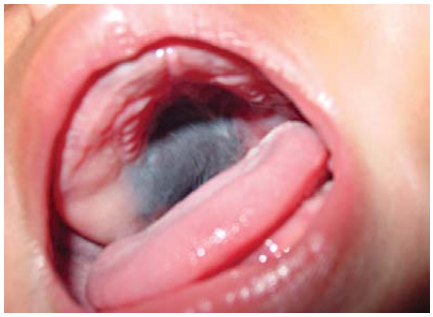


Fig. 14.10.2: Lymphangioma—oropharynx

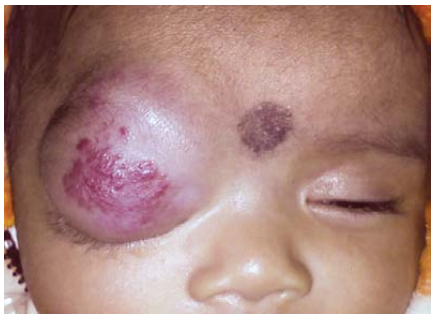


Fig. 14.10.3A: Lymphohemangioma

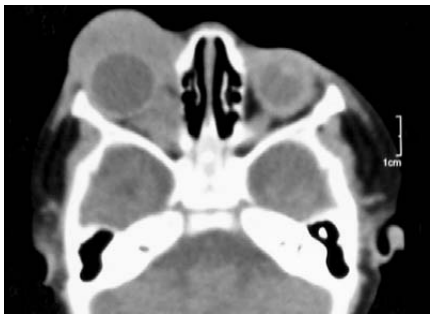


Fig. 14.10.3B: Lymphohemangioma

Glioma of the Optic Nerve

- Ectodermal tumor of the optic nerve
- Most prevalent in childhood between 2 to 8 years
- 50% cases associated with neurofibromatosis
- Presents as unilateral proptosis with visual impairment **(Figs 14.11.1A and B)**
- Slow growing unilateral proptosis which is axial **(Fig. 14.11.2)**
- Marcus-Gunn pupillary reaction and optic atrophy
- Enlargement of optic foramen in X-ray is seen in 90% cases
- *Treatment:* Lateral orbitotomy in most cases.

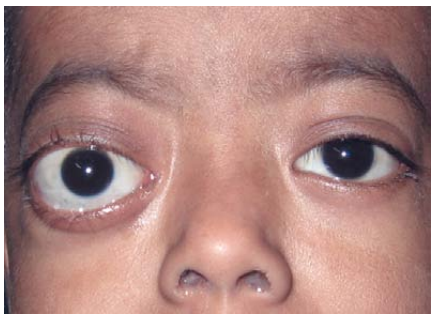


Fig. 14.11.1A: Axial proptosis—optic nerve glioma

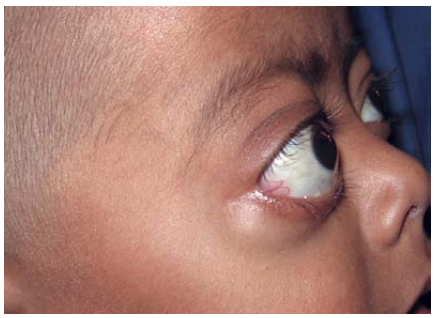


Fig. 14.11.1B: Axial proptosis—optic nerve glioma on left lateral view

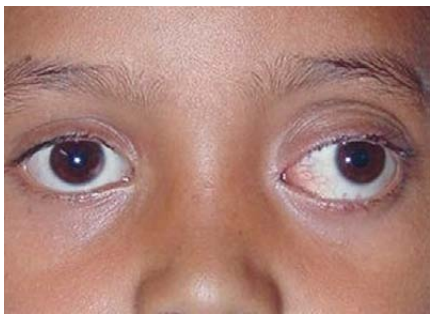


Fig. 14.11.2: Optic nerve glioma—dilated pupil with divergent left eye

Rhabdomyosarcoma

- Most common primary malignant orbital tumor in children
- Occurs at about 7 years of age and more in boys
- Tumor arises from striated muscles
- Rapidly growing proptosis with chemosis of conjunctiva and lids **(Fig. 14.12.1)**
- Typically, a mass is visible and palpable in the superonasal quadrant or superior orbit **(Figs 14.12.2A and B)**
- In late stage, it may extend into the cranium
- *Treatment:* Combination of radiotherapy and chemotherapy; in unresponsive cases, exenteration of the orbit may be required. A paramedian tarsorrhaphy is required to prevent exposure **(Fig. 14.12.3)**.

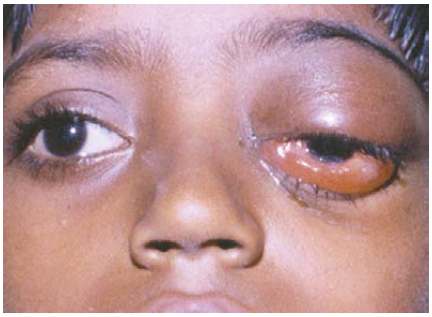


Fig. 14.12.1: Rhabdomyosarcoma

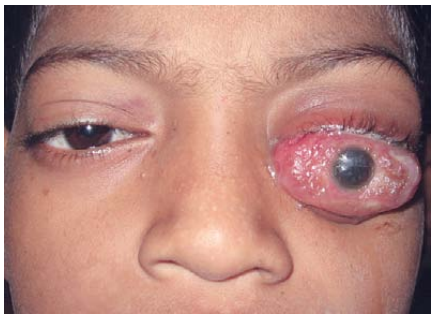


Fig. 14.12.2A: Rhabdomyosarcoma—exposure and keratinization

Fig. 14.12.2B: Rhabdomyosarcoma—exposure and keratinization

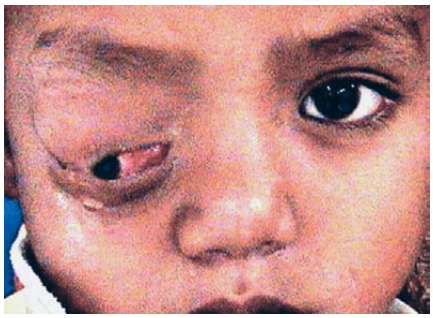


Fig. 14.12.3: Rhabdomyosarcoma—late stage

Orbital Extension of Retinoblastoma

- In neglected cases, retinoblastoma may present with proptosis due to direct orbital extension **(Figs 14.13.1 to 14.13.3)**
- The child looks toxic at this stage **(Fig. 14.13.4)**
- *Treatment:* Exenteration followed by radiotherapy
- Prognosis is extremely poor.

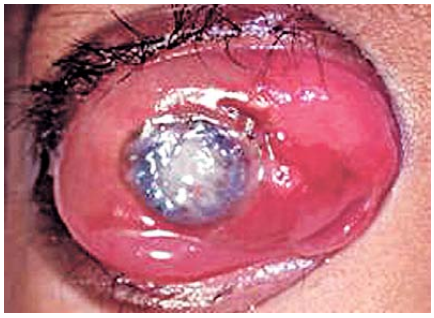


Fig. 14.13.1: Orbital extension of retinoblastoma

Fig. 14.13.2: Orbital extension of retinoblastoma

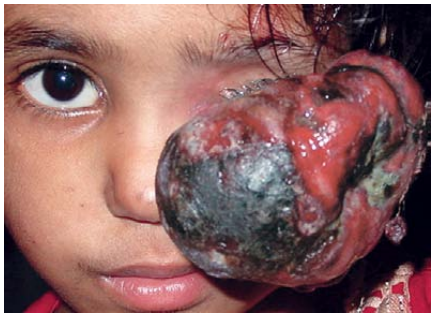


Fig. 14.13.3: Orbital extension of retinoblastoma—massive

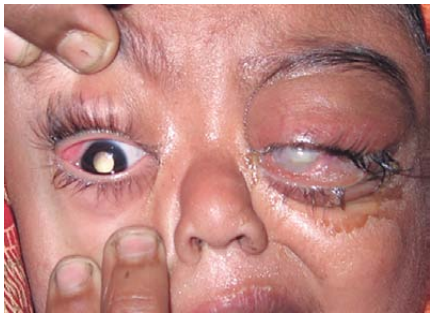


Fig. 14.13.4: Bilateral retinoblastoma with orbital extension in LE

Acute Leukemia

- Most frequently occurs between 5 to 8 years of age
- Orbital soft tissue involvement in *chloroma* is not associated with abnormal peripheral blood smear which occurs more in African and Asian children
- Unilateral or bilateral rapidly progressive proptosis **(Fig. 14.14.1)**
- May be associated with ecchymosis and chemosis of lids **(Fig. 14.14.2)**
- Prognosis is poor.

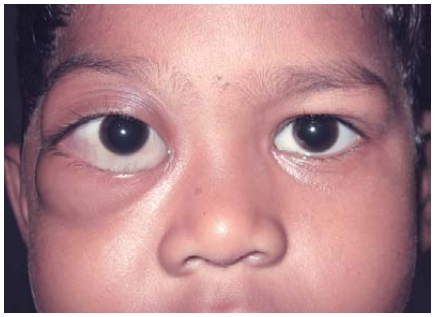


Fig. 14.14.1: Leukemic deposits in right orbit

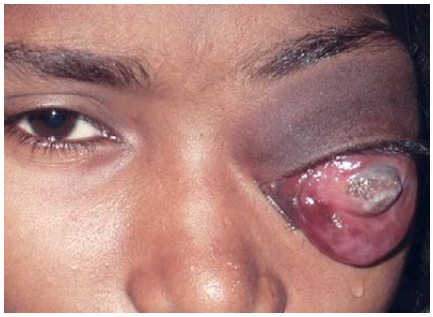


Fig. 14.14.2: Acute leukemia—unilateral acute proptosis

Other Orbital Tumors in Children

- *Metastatic neuroblastoma:* Sudden, rapidly growing proptosis with superolateral orbital mass **(Figs 14.15.1 and 14.15.2)**
- *Metastatic Ewing's sarcoma:* Rapidly growing proptosis with ecchymosis and chemosis
- *Metastatic Wilms' tumor:* Associated with aniridia
- *Fibrosarcoma:* History of previous radiotherapy to orbit for retinoblastoma **(Fig. 14.15.3)**
- *Sinus tumors involving the orbit:* Osteogenic sarcoma, Burkitt's lymphoma, etc.
- *Enchondromatosis:* Very rare, islands of cartilage in metacarpals and phalanges; hemangioma and enchondromas of the orbit gives rise to proptosis **(Figs 14.15.4A and B)**

Fig. 14.15.1A: Proptosis (RE)—metastatic neuroblastoma

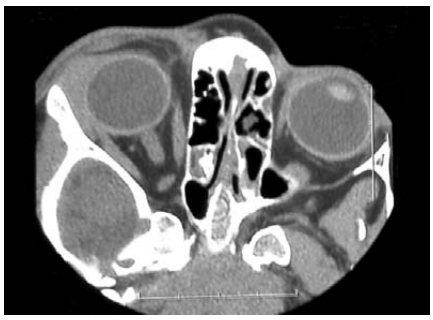


Fig. 14.15.1B: Proptosis (RE)—metastatic neuroblastoma—CT scan

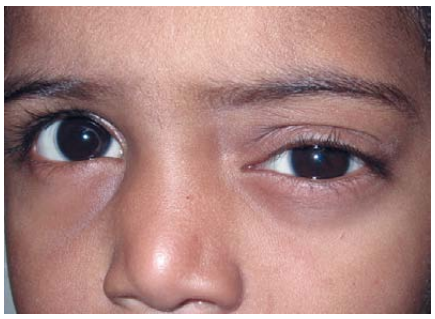


Fig. 14.15.2: Metastatic neuroblastoma—proptosis—LE

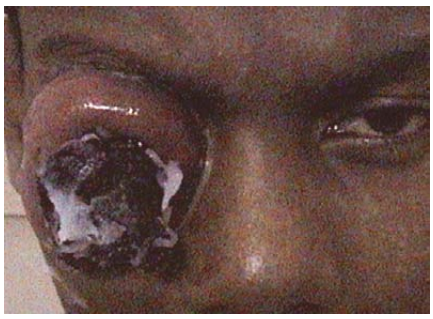


Fig. 14.15.3: Metastatic fibrosarcoma

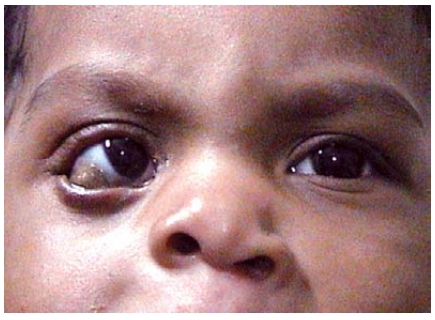


Fig. 14.15.4A: Proptosis—RE—enchondromatosis

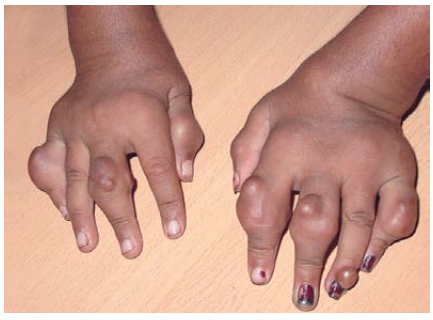


Fig. 14.15.4B: Enchondromatosis—metacarpals and phalanges

- *Dermoid cyst of the orbit:* May be anterior or posterior **(Figs 14.15.5 to 14.15.7)**
- *Orbital teratoma:* Again very rare, congenital benign orbital tumor may present with proptosis; rarely turn into malignant germ cell tumor even after resection **(Figs 14.15.8 and 14.15.9)**
- *Ethmoidal sinus mucocele:* **(Figs 14.15.10A and B)**
- *AV malformation with or without bony deformity of the orbit:* **(Figs 14.15.11 and 14.15.12)**
- *Idiopathic orbital inflammatory mass:* **(Figs 14.15.13A and B)**

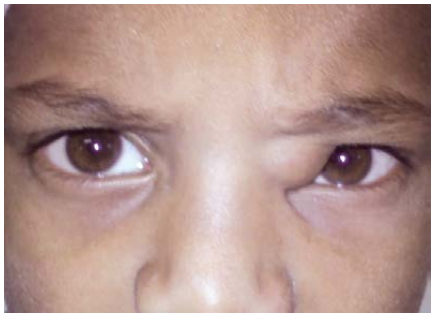


Fig. 14.15.5: Anterior orbital dermoid

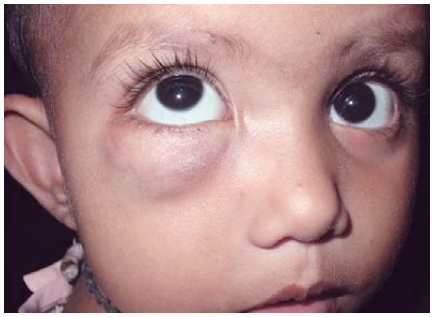


Fig. 14.15.6: Proptosis—dermoid cyst in lower orbit

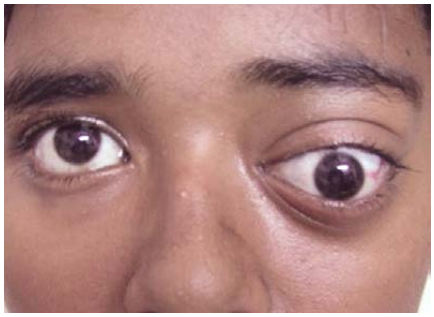


Fig. 14.15.7A: Proptosis—dermoid cyst in upper orbit

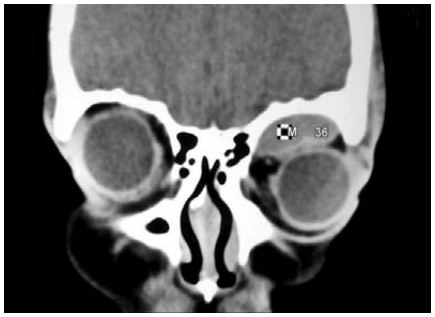


Fig. 14.15.7B: Proptosis—dermoid cyst in upper orbit

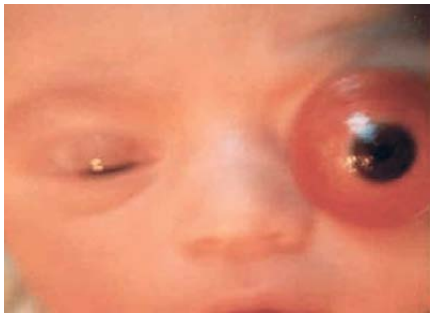


Fig. 14.15.8: Ethmoidal sinus mucocele—LE

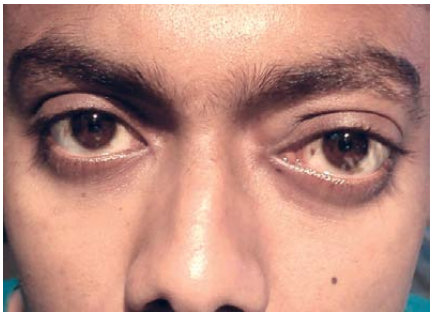


Fig. 14.15.9: Ethmoidal sinus mucocele—on CT scan

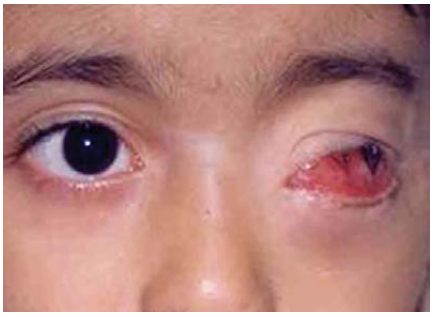


Fig. 14.15.10A: Orbital teratoma—4 days old child

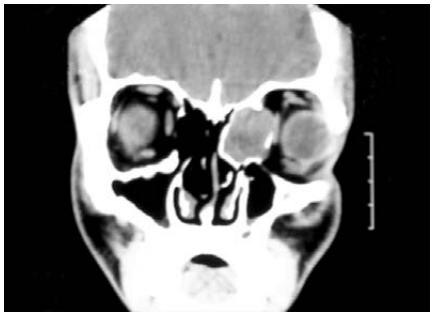


Fig. 14.15.10B: Orbital teratoma—recurrence after 5 years of resection

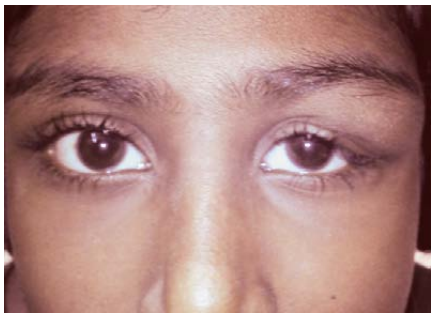


Fig. 14.15.11A: AV malformations of blood vessels within the orbit

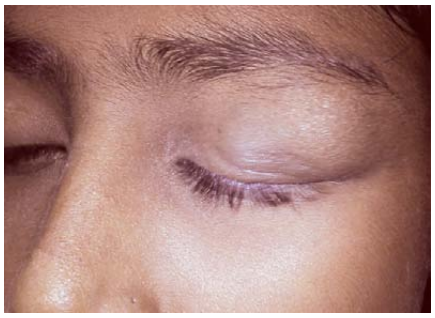


Fig. 14.15.11B: AV malformations of blood vessels within the orbit

Fig. 14.15.11C: AV malformations of blood vessels within the orbit

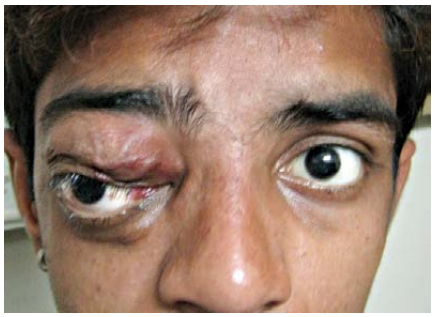


Fig. 14.15.12: AV malformations with bony deformity

Fig. 14.15.13A: Idiopathic orbital inflammatory mass

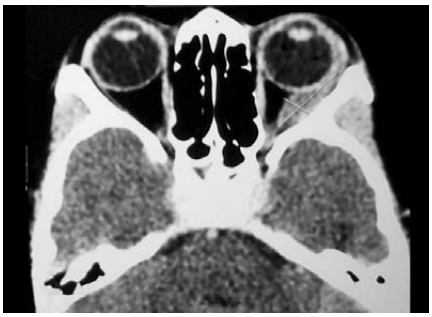


Fig. 14.15.13B: Idiopathic orbital inflammatory mass

- *Juvenile xanthogranuloma:*
  - Very rare, benign infantile condition
  - Unilateral proptosis, often associated with strabismus
  - Cutaneous lesions, heterochromia of the iris and spontaneous orbital hemorrhage **(Figs 14.15.14A and B)** or hyphema may be present.

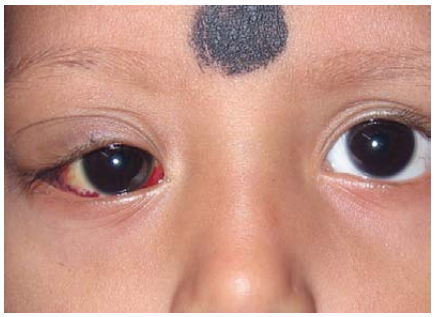


Fig. 14.15.14A: Juvenile xanthogranuloma of right orbit

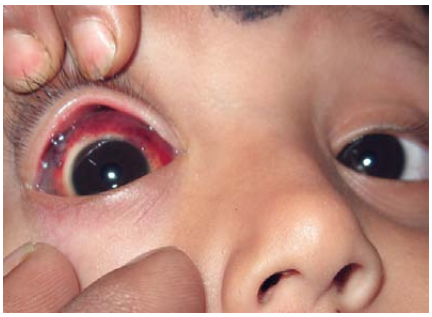


Fig. 14.15.14B: Juvenile xanthogranuloma of right orbit

Proptosis in Adults

Inflammatory Orbital Diseases (Orbital Pseudotumor)

- Idiopathic, non-neoplastic, non-microbial space-occupying periocular lesion, which may simulate an orbital neoplasm (pseudotumor)
- Typically affects the males between 20 to 50 years
- Unilateral axial proptosis with variable degree of chemosis of the lids **(Figs 14.16.1A and B)**
- Conjunctival chemosis and congestion
- Limitation of extraocular movements and diplopia
- To be differentiated from thyroid ophthalmopathy and orbital cellulitis **(Fig. 14.16.2)**
- USG B scan orbit and CT scan are important tools for diagnosis
- *Treatment:* Observation, systemic corticosteroids, and cytotoxic agents in resistance cases.

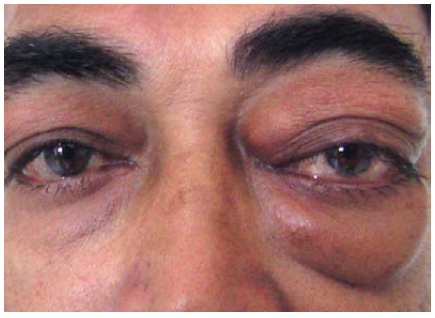


Fig. 14.16.1A: Inflammatory orbital diseases—left side

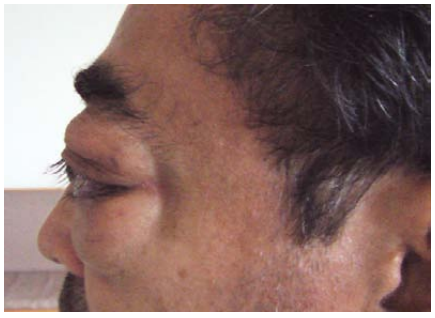


Fig. 14.16.1B: Inflammatory orbital diseases on left lateral view

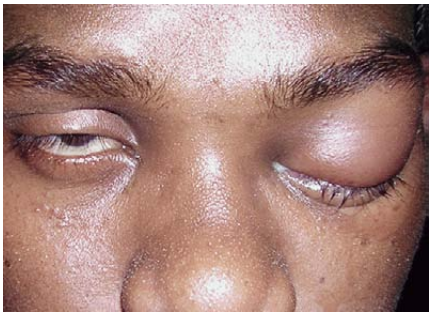


Fig. 14.16.2: Inflammatory orbital diseases—left side

Orbital Cellulitis in Adults

- More often from infection of adjacent structures like, acute dacryocystitis, internal hordeolum, dental infection, etc.
- May be post-traumatic or post-surgical
- Painful, unilateral rapid proptosis with severe lid edema **(Fig. 14.17.1)**
- Like children, this is to be differentiated from preseptal cellulitis and lid abscess in adults **(Figs 14.17.2 and 14.17.3)**
- Limitation of extraocular movements
- *Treatment:* High dose of broad-spectrum parenteral antibiotics, systemic analgesics, drainage of abscess if required.

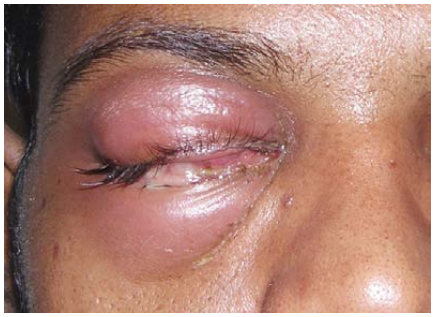


Fig. 14.17.1: Orbital cellulitis in adult

Fig. 14.17.2: Preseptal cellulitis with lid abscess in adult

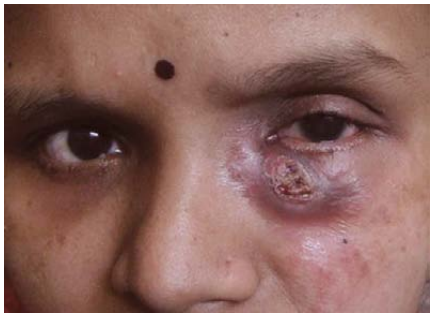


Fig. 14.17.3: Preseptal cellulitis following acute dacryocystitis

Thyroid Associated Ophthalmopathy

- Most common cause of proptosis in adults
- Unilateral **(Figs 14.18.1 and 14.18.2)** or bilateral chronic slow growing axial proptosis **(Figs 14.18.3 and 14.18.4)**
- Retraction of the eyelid and lid lag **(Fig. 14.18.5A)**
- Hyperemia of conjunctiva near horizontal rectus muscles **(Fig. 14.18.5B)**

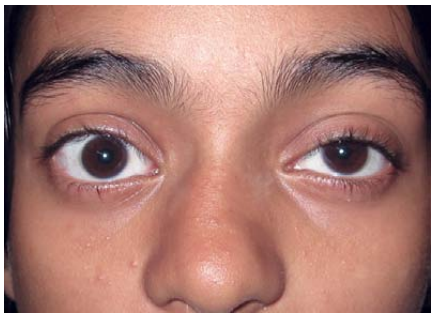


Fig. 14.18.1: Thyroid exophthalmos—unilateral

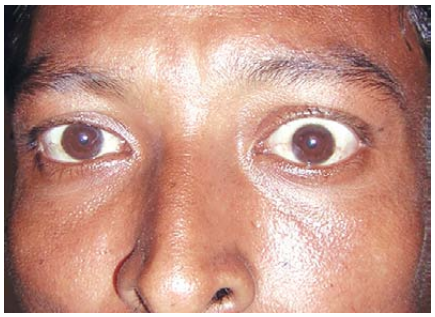


Fig. 14.18.2: Thyroid exophthalmos—unilateral

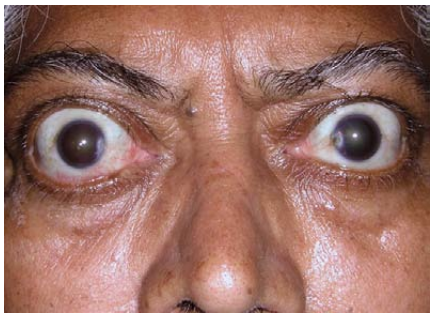


Fig. 14.18.3: Thyroid exophthalmos—bilateral

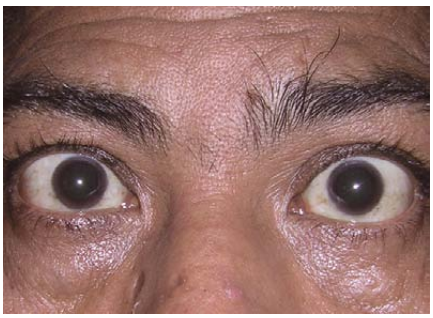


Fig. 14.18.4: Thyroid exophthalmos—bilateral

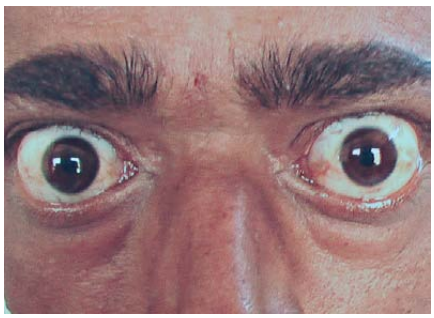


Fig. 14.18.5A: Thyroid exophthalmos—bilateral

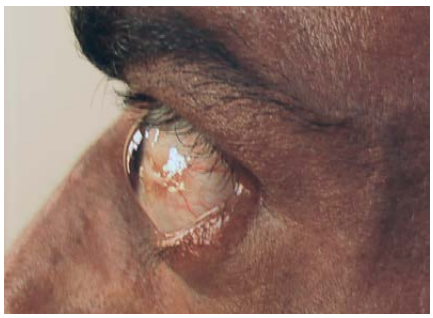


Fig. 14.18.5B: Thyroid exophthalmos—profile view

- Thyroid swelling may be obvious in some cases **(Fig. 14.18.5C)**
- Extraocular muscle involvement (most common—inferior rectus, then medial rectus, least common—lateral rectus) with restriction of movements
- Conjunctival chemosis and severe congestion with inflammation in advance cases **(Fig. 14.18.6A)**
- Superior limbic keratoconjunctivitis
- Corneal involvement like, exposure keratopathy or dry eye in late stage **(Fig. 14.18.6B)**
- Compressive optic neuropathy, disk edema and chorioretinal folds in late stage **(Fig. 14.18.7)**
- **WARNER's classification of ocular changes**
  (*Mnemonic* = **"NO SPECS"**)
  Class: Ocular changes
  0. No symptom and sign.
  1. Only sign (lid retraction, lid lag or proptosis).
  2. Soft tissue changes
     Symptoms—*photophobia, lacrimation, foreign body* sensation
     Signs—hyperemia and chemosis of conjunctiva, resistance to retroplacement, palpable lacrimal gland.
  3. Proptosis.
  4. Extraocular muscles involvement (*most common*—inferior rectus, then—medial rectus, least common—lateral rectus).
  5. Corneal involvement (exposure keratopathy or dry eye).
  6. Sight loss due to optic neuropathy and corneal involvement.
- *Treatment:* Antithyroid drugs, frequent artificial tears, antibiotic ointment at bed time, systemic corticosteroids or cytotoxic agents, lateral tarsorrhaphy and orbital decompression in severe cases.

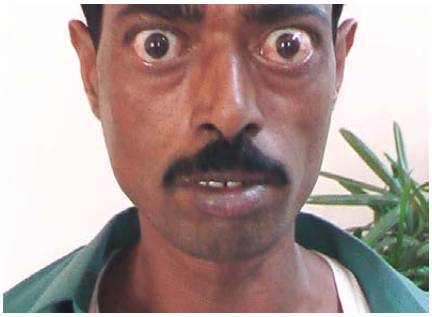


Fig. 14.18.5C: Thyroid exophthalmos—thyroid swelling

Fig. 14.18.6A: Thyroid exophthalmos with severe chemosis and congestion

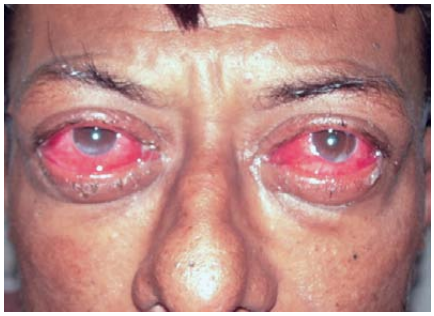


Fig. 14.18.6B: Thyroid exophthalmos—bilateral—with exposure keratitis

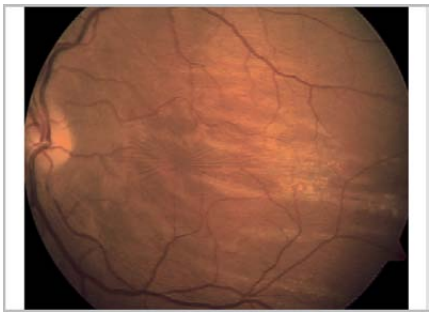


Fig. 14.18.7: Thyroid ophthalmopathy—choroidal folds

Cavernous Hemangioma

- Most common benign tumor in adults
- Presents between 30 to 50 years of age
- Unilateral, slowly progressive proptosis which is usually axial **(Figs 14.19.1 and 14.19.2)**
- In severe cases, it may be associated with chemosis and conjunctival congestion **(Fig. 14.19.3)**

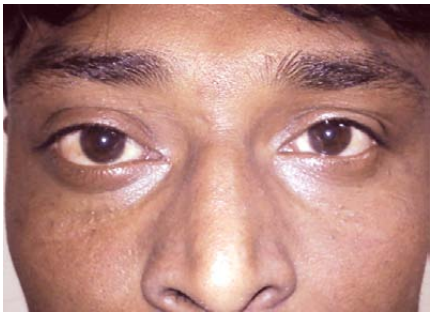


Fig. 14.19.1A: Cavernous hemangioma—right orbit

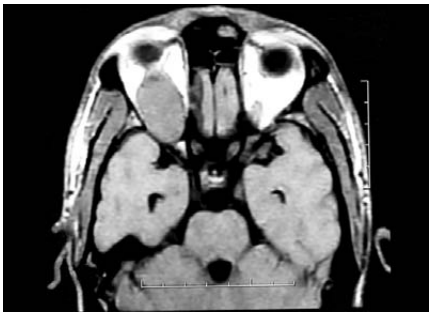


Fig. 14.19.1B: Cavernous hemangioma on MRI

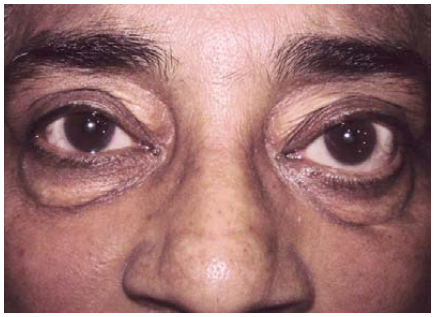


Fig. 14.19.2A: Cavernous hemangioma—left orbit

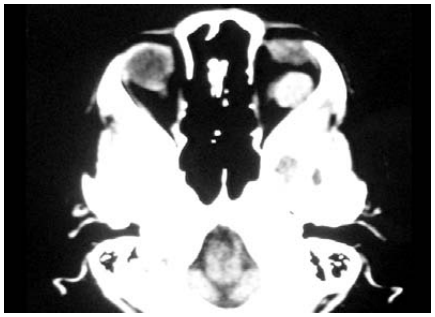


Fig. 14.19.2B: Cavernous hemangioma

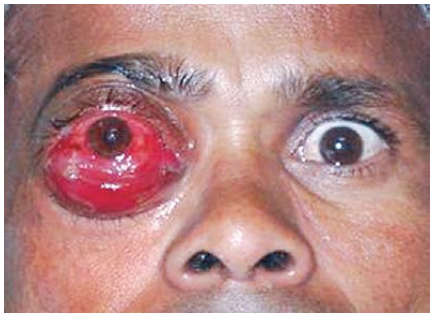


Fig. 14.19.3: Cavernous hemangioma with conjunctival chemosis

- Usually not associated with optic nerve compression unless it is at the orbital apex
- *Treatment:* Surgical excision of the mass by lateral orbitotomy.

Orbital Varix

- Usually unilateral, vascular malformation (mainly venous origin), can occur at any age
- Nonpulsatile, intermittent axial proptosis, not associated with a bruit and with prominent conjunctival vessels **(Fig. 14.20.1)**
- Proptosis may be accentuated or precipitated by dependent head posture or by performing Valsalva maneuver **(Figs 14.20.2A to C)**
- *Vascular malformations:* Associated vascular lesion of the eyelids or conjunctiva and it can originate from both arteries or veins **(Figs 14.20.3A and B)**.

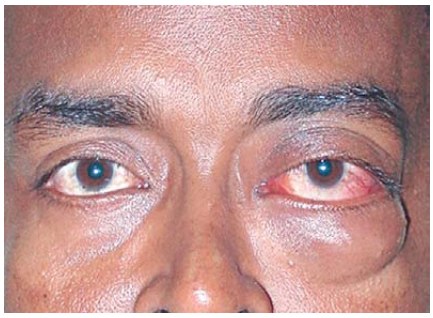


Fig. 14.20.1: Orbital varix

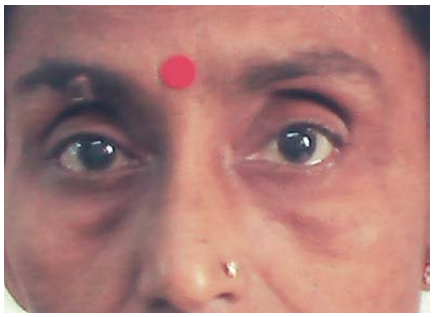


Fig. 14.20.2A: Orbital varix—normal looking on primary gaze

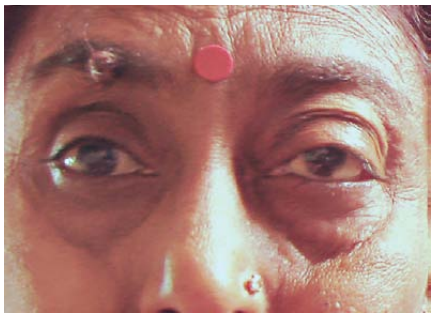


Fig. 14.20.2B: Proptosis in LE—just after dependent head posture

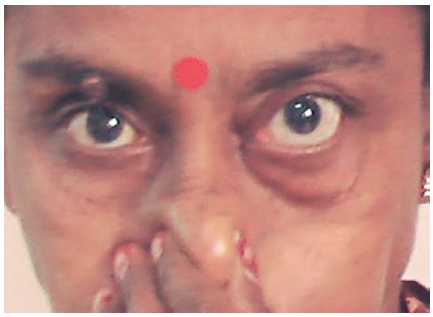


Fig. 14.20.2C: Orbital varix—proptosis precipitated by Valsalva maneuver

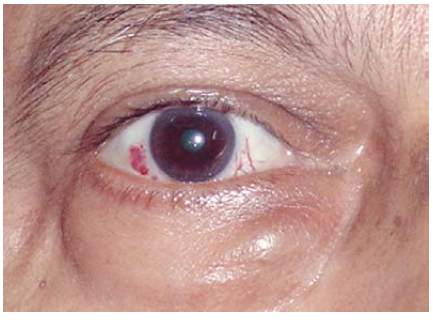


Fig. 14.20.3A: Orbital varix—vascular malformation of lid and conjunctiva

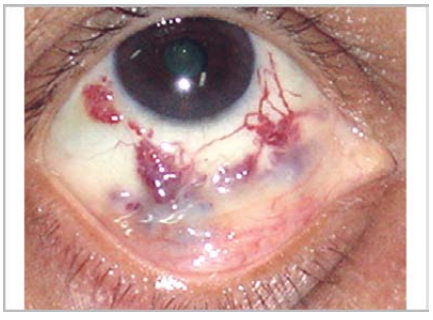


Fig. 14.20.3B: Orbital varix—vascular malformation of lid and conjunctiva

Meningioma of the Optic Nerve

- Occurs predominantly in the middle-aged women
- Ocular features are related to the site of primary involvement by the tumor
- *Tuberculum sellae meningioma:* Compress the optic chiasma
- *Sphenoidal-ridge meningioma:*
  - Slowly growing, painless down and out proptosis **(Fig. 14.21.1)**
  - Fullness of the temporal fossa of the affected side **(Fig. 14.21.2)**
  - Visual impairment due to optic nerve compression
- *Optic nerve-sheath meningioma:* Slowly-growing unilateral axial proptosis **(Figs 14.21.3A and B)**
  - The *triad* of long-standing visual impairment, a pale swollen optic disk, and opticociliary shunt vessels is virtually pathognomonic of optic nerve-sheath meningioma

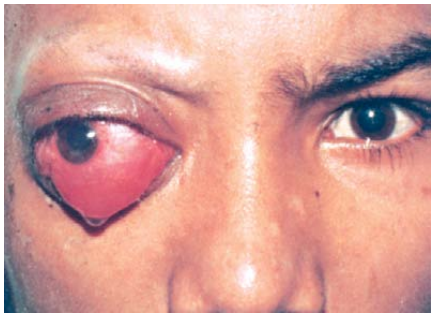


Fig. 14.21.1: Sphenoidal ridge meningioma

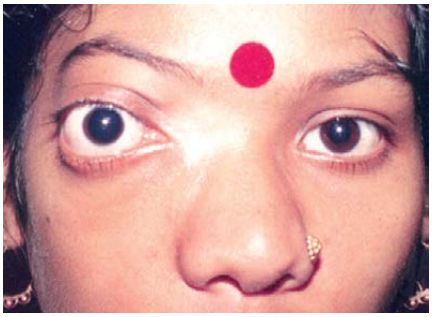


Fig. 14.21.2: Sphenoidal ridge meningioma—temporal fossa fullness

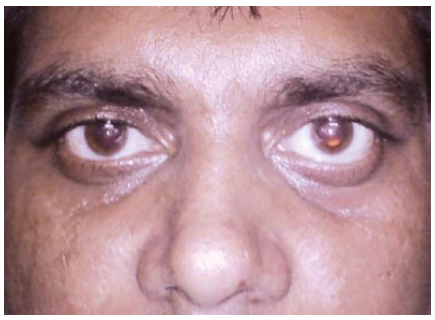


Fig. 14.21.3A: Optic nerve sheath meningioma—axial proptosis

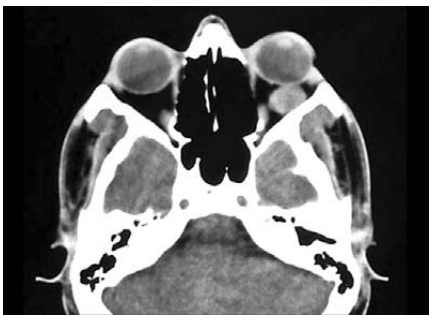


Fig. 14.21.3B: Optic nerve sheath meningioma—axial proptosis

- *Treatment:* Surgical excision of the tumor by lateral orbitotomy.

Carotid Cavernous Fistula

- Results from an abnormal communication between the cavernous sinus and the internal carotid artery, giving the classical picture of a pulsating exophthalmos **(Figs 14.22.1A and B)**
- May be traumatic or spontaneous
- Marked unilateral, sudden pulsatile proptosis with severe chemosis
- Redness and prominent dilatation of the episcleral blood vessels in radial pattern giving *caput medusae* appearance **(Figs 14.22.2A and B)**
- Ophthalmoplegia due to involvement of the 3rd, 4th and 5th nerve
- Retinal venous congestion with hemorrhage—a picture similar to CRVO **(Fig. 14.22.2C)**
- Raised IOP, due to elevated episcleral venous pressure
- Anterior segment ischemia may develop in some patients
- *Treatment:* Intracavernous surgery and balloon catheter embolization.

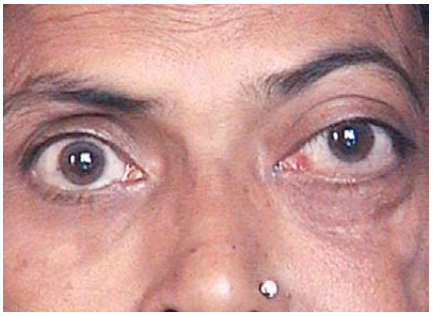


Fig. 14.22.1A: Caroticocavernous fistula

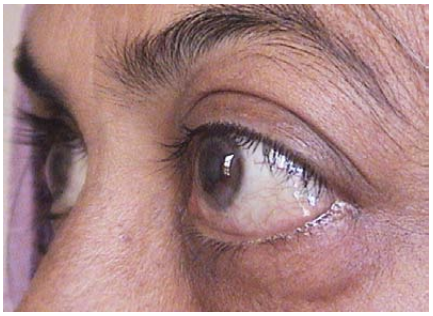


Fig. 14.22.1B: Caroticocavernous fistula

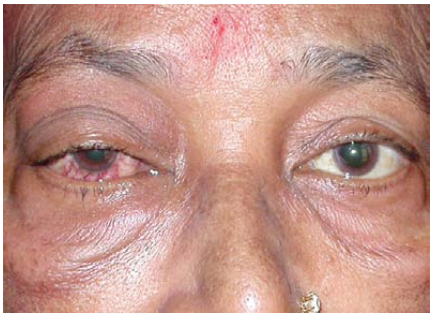


Fig. 14.22.2A: Caroticocavernous fistula—note the conjunctival vessels in RE

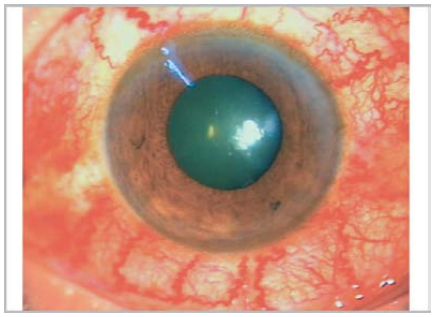


Fig. 14.22.2B: Caput medusae of same patient (**Fig. 14.22.2A**)

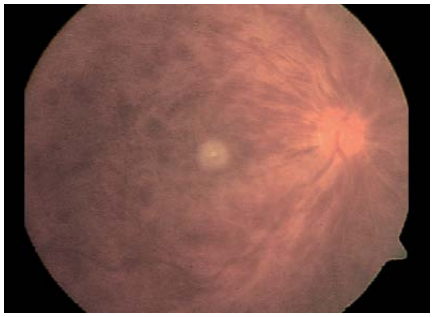


Fig. 14.22.2C: CRVO of same patient (**Fig. 14.22.2A**)

Cavernous Sinus Thrombosis

- An acute thrombophlebitis with violent onset
- Features are almost similar to caroticocavernous fistula **(Fig. 14.22.3)**
- Edema of the mastoid region indicating back-pressure in the mastoid emissary vein—a *pathognomonic sign*
- *Treatment:* Intensive intravenous antibiotics and anticoagulants.

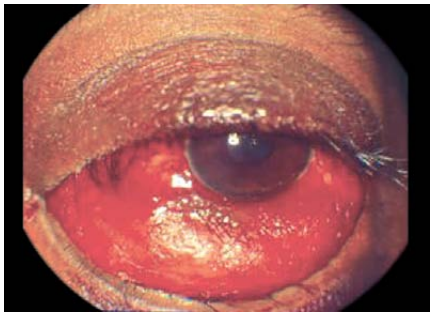


Fig. 14.22.3: Cavernous sinus thrombosis—conjunctival chemosis and hemorrhage

Other Causes of Proptosis in Adults

- **Lymphoid tumors**
  - Usually in old age
  - *Benign lymphoma:* Unilateral **(Figs 14.23.1A and B)** or bilateral **(Figs 14.23.2A and B)** with orbital puffiness
  - Rubbery consistency on palpation

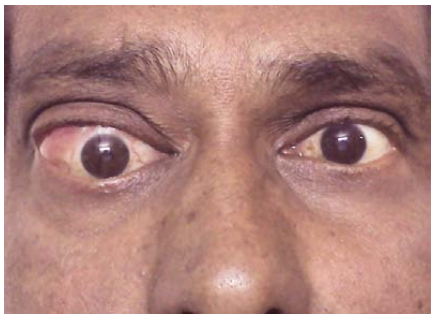


Fig. 14.23.1A: Orbital benign lymphoma—unilateral—RE

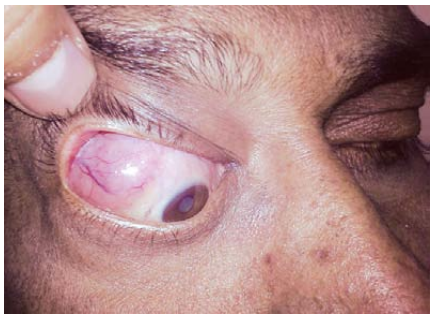


Fig. 14.23.1B: Orbital benign lymphoma—unilateral

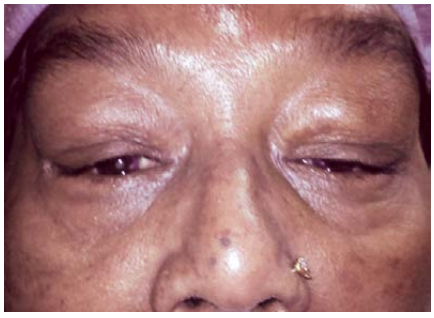


Fig. 14.23.2A: Orbital benign lymphoma—bilateral

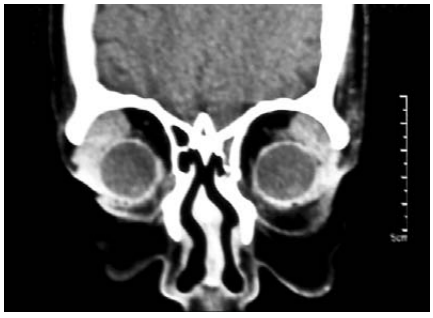


Fig. 14.23.2B: Orbital benign lymphoma—bilateral

- Associated conjunctival extension
- *Non-Hodgkin's lymphoma:* **(Figs 14.23.3A and B)**

- **Optic nerve glioma:**
  - As in children
  - Slow growing axial proptosis with visual loss **(Figs 14.23.4A to C)**
  - RAPD on affected side

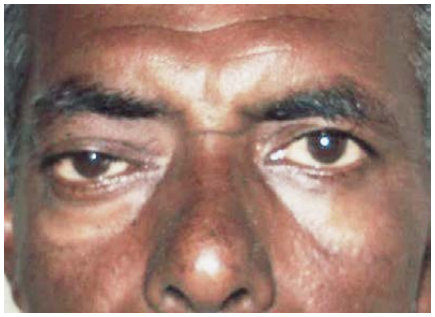


Fig. 14.23.3A: Non-Hodgkin's lymphoma—left side

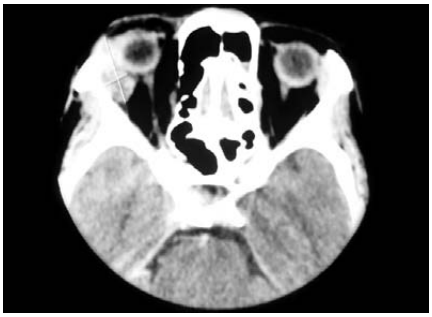


Fig. 14.23.3B: Non-Hodgkin's lymphoma—left side

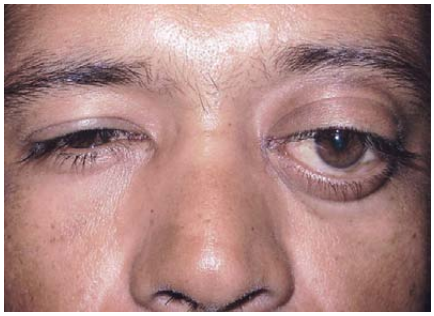


Fig. 14.23.4A: Proptosis in adult

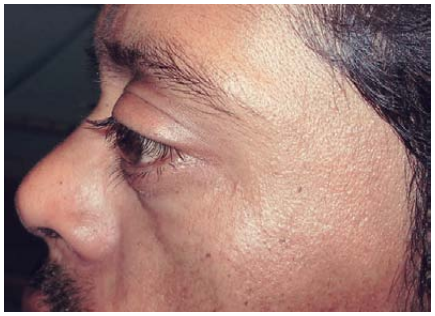


Fig. 14.23.4B: Glioma optic nerve—adult

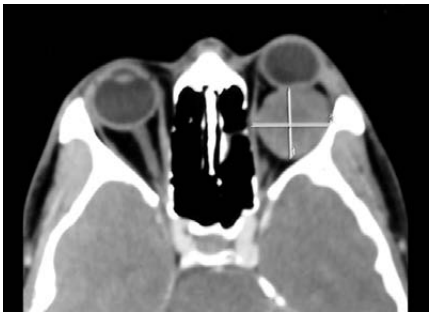


Fig. 14.23.4C: Glioma optic nerve—adult—CT scan

- **Intraorbital dermoid:**
  - Slow growing unilateral axial or nonaxial proptosis **(Figs 14.23.5 and 14.23.6)**
- **Primary tumors of the adnexa or adjacent structures:**
  - *Lacrimal gland tumors:* Pleomorphic adenoma **(Figs 14.23.6 and 14.23.7)** and adenocystic carcinoma **(*see* Figs 13.2.5A to C)**
  - *Sebaceous gland tumor* **(Figs 14.23.8A to C)**
  - *Maxillary sinus carcinoma* **(Fig. 14.23.9)**
  - *Ethmoidal sinus carcinoma* **(Figs 14.23.10A and B)**
- **Metastatic tumors:**
  - Relatively rapid onset proptosis
  - Often with pain and diplopia
  - Severe chemosis and conjunctival congestion
  - *Primary site:* Bronchus, prostate, breast **(Fig. 14.23.11)** or gastrointestinal tract
  - Leukemic deposits also occur in adults **(Figs 14.23.12 and 14.23.13)**

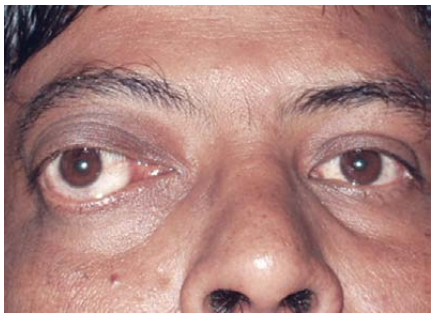


Fig. 14.23.5A: Intraorbital dermoid—axial proptosis

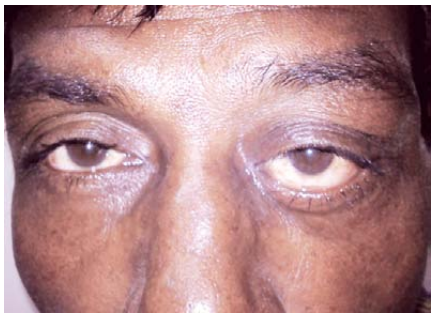


Fig. 14.23.5B: Intraorbital dermoid—axial proptosis

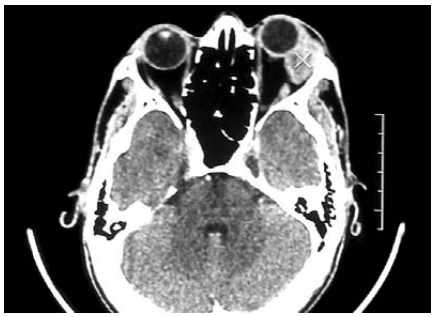


Fig. 14.23.5C: Intraorbital dermoid—axial proptosis on CT scan

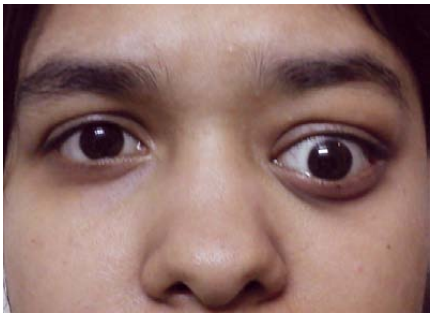


Fig. 14.23.6: Pleomorphic adenoma of lacrimal gland

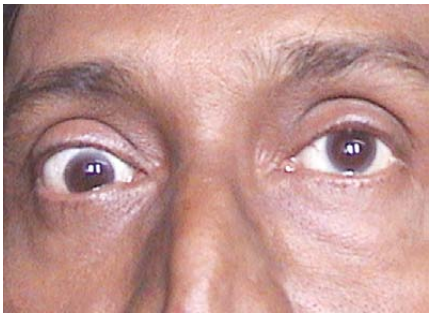


Fig. 14.23.7: Proptosis (RE)—pleomorphic adenoma of lacrimal gland

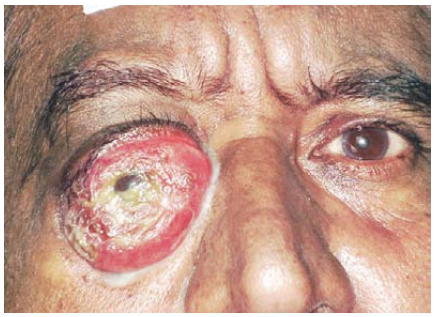


Fig. 14.23.8A: Proptosis (RE)—primary sebaceous carcinoma of lacrimal gland

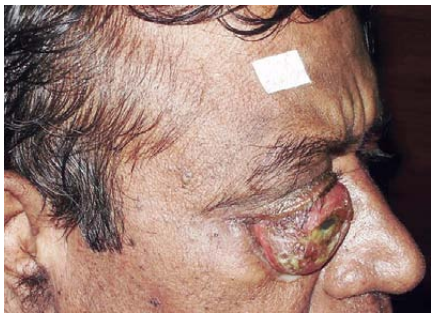


Fig. 14.23.8B: Proptosis (RE)—primary sebaceous carcinoma of lacrimal gland

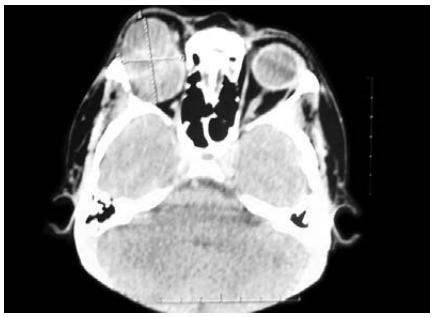


Fig. 14.23.8C: Proptosis (RE)—primary sebaceous carcinoma of lacrimal gland on CT scan

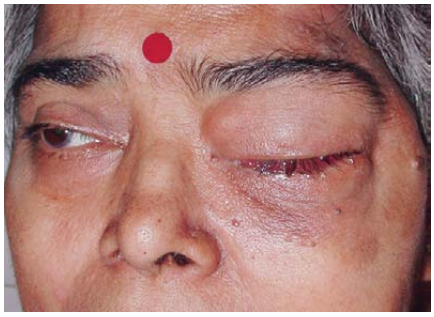


Fig. 14.23.9: Proptosis—from maxillary sinus carcinoma

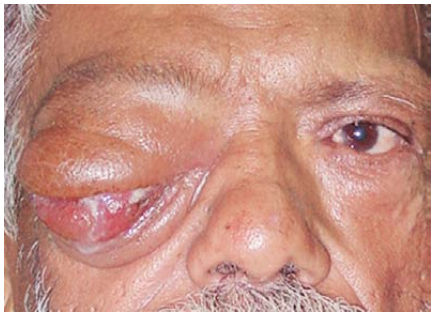


Fig. 14.23.10A: Proptosis—from ethmoidal sinus carcinoma

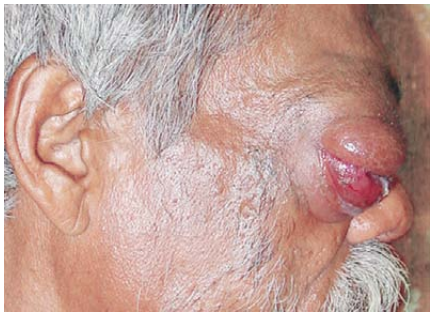


Fig. 14.23.10B: Proptosis—from ethmoidal sinus carcinoma

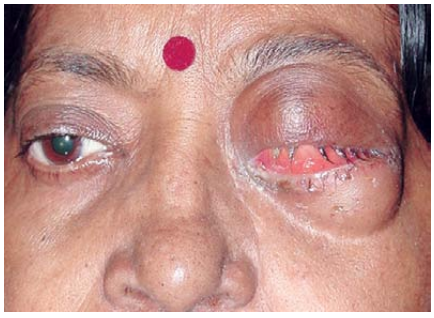


Fig. 14.23.11: Metastatic deposits—breast carcinoma

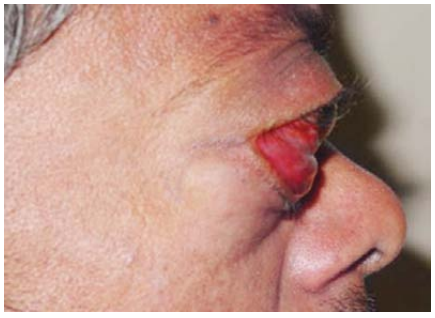


Fig. 14.23.12: Metastatic deposits—leukemia

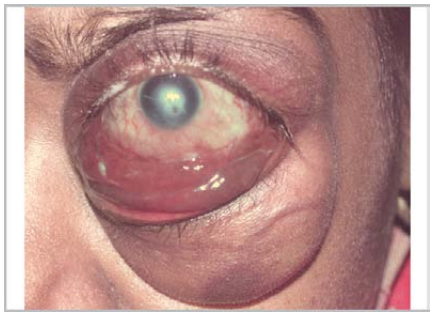


Fig. 14.23.13: Metastatic deposits—acute leukemia

- *Granulocytic sarcoma (chloroma):* Represents a localized form of acute myeloblastic leukemia, called a chloroma because the myeloperoxidase within the tumor imparts a green due to its surface **(Figs 14.23.14A and B)**
- *Rhabdomyosarcoma in adults:* **(Figs 14.23.15A to C)**

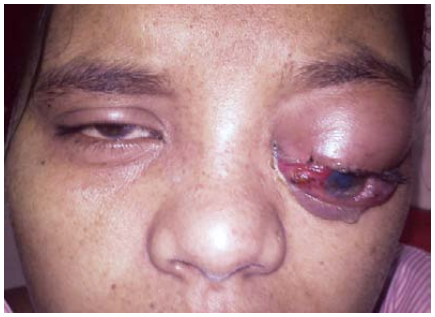


Fig. 14.23.14A: Metastatic deposits—chloroma

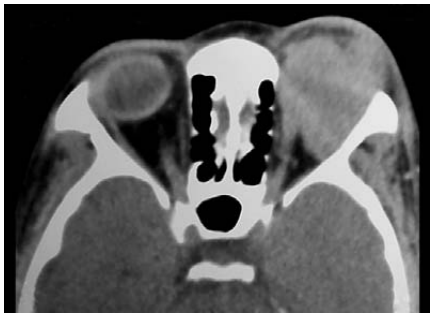


Fig. 14.23.14B: Metastatic deposits—chloroma

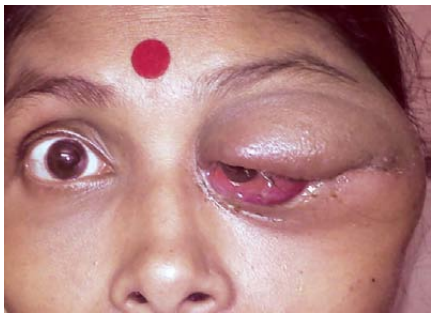


Fig. 14.23.15A: Rhabdomyosarcoma in adult

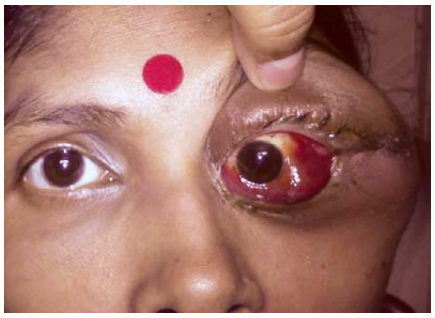


Fig. 14.23.15B: Rhabdomyosarcoma in adult

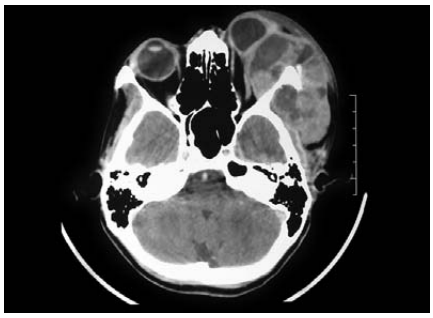


Fig. 14.23.15C: Rhabdomyosarcoma in adult on CT scan

- **Cystic lesions within the orbit:**
  - *Nonspecific inflammatory cyst* **(Figs 14.23.16A to C)**
  - *Orbital cysticercosis* **(Figs 14.23.17A to C)**
  - *Orbitopalpebral cyst* **(Figs 14.23.18A and B)**.

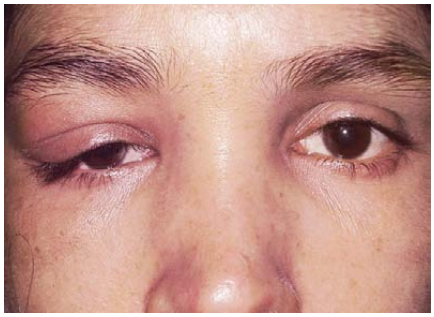


Fig. 14.23.16A: Nonspecific inflammatory cyst—RE

Fig. 14.23.16B: Nonspecific inflammatory cyst—RE

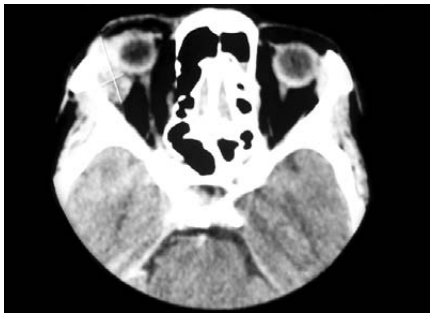


Fig. 14.23.16C: Nonspecific inflammatory cyst on CT scan

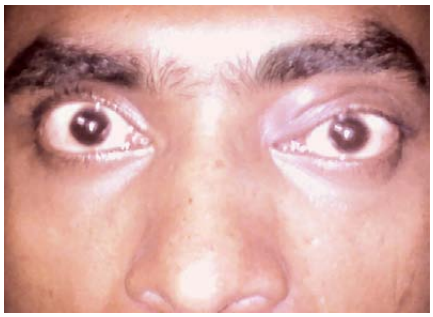


Fig. 14.23.17A: Orbital cysticercosis—superonasal area—LE

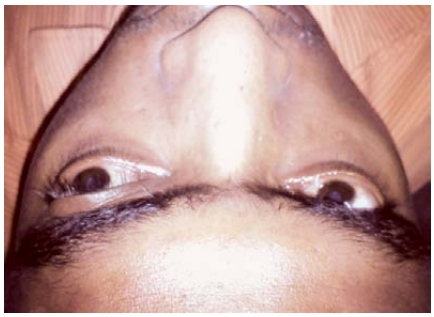


Fig. 14.23.17B: Orbital cysticercosis—LE

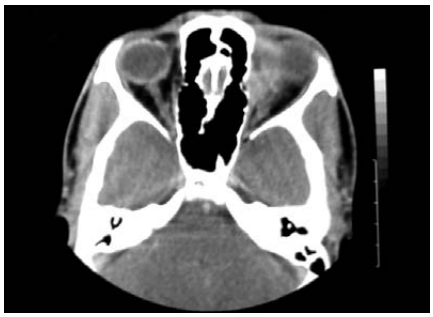


Fig. 14.23.17C: Orbital cysticercosis—LE on CT scan

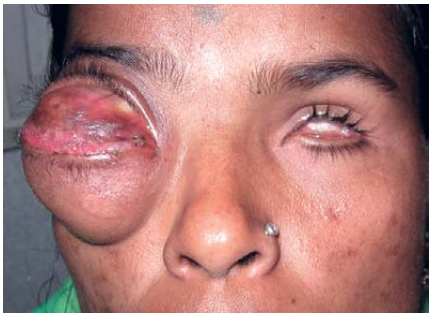


Fig. 14.23.18A: Orbitopalpebral cyst—RE and anophthalmic socket—LE

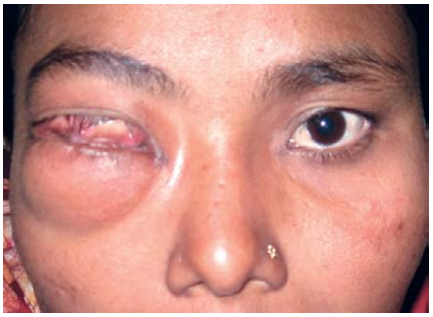


Fig 14.23.18B: Orbitopalpebral cyst—RE and normal—LE

Miscellaneous Orbital Lesions

Orbital Rim Lesions

- Almost always unilateral and may be anywhere
- Associated with bony defects
- *Causes:*
  - *External angular dermoid*: Mostly at upper and outer quadrant **(Figs 14.24.1A and B)**
  - Subperiosteal lesions
  - *Cavernous hemangioma* **(Fig. 14.24.2)**
  - Fracture orbital margin/wall **(*see* Figs 15.24.1 and 15.24.2)**
  - Plexiform neurofibroma **(Fig. 14.24.3)**
  - Orbital mucocele **(Figs 14.24.4A and B)**.

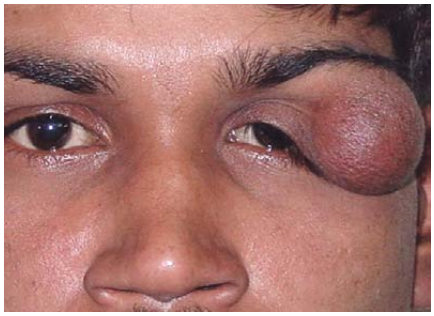


Fig. 14.24.1A: External angular dermoid—superolateral orbital angle

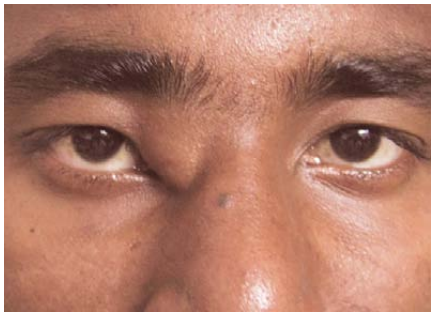


Fig. 14.24.1B: External angular dermoid—superomedial orbital angle

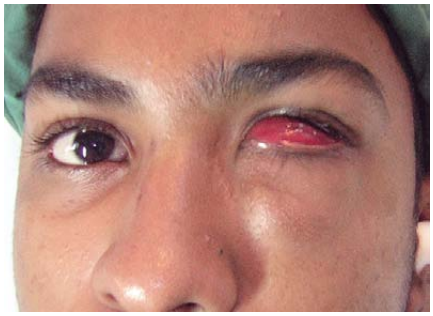


Fig. 14.24.2: Cavernous hemangioma

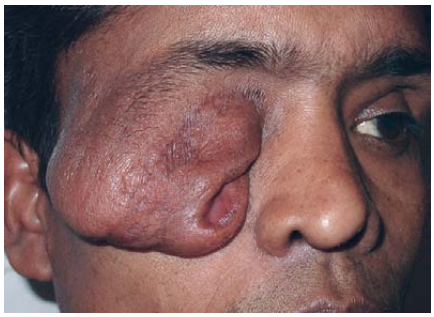


Fig. 14.24.3: Plexiform neurofibroma

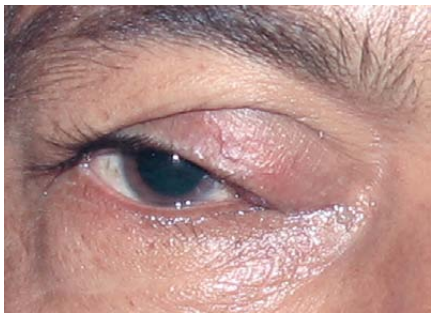


Fig. 14.24.4A: Orbital mucocele

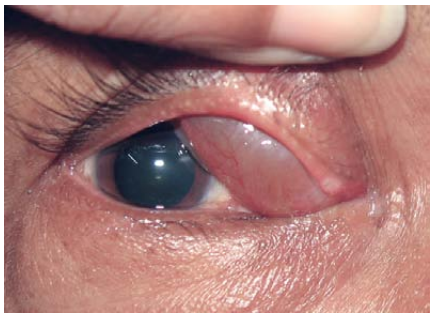


Fig. 14.24.4B: Orbital mucocele

Luxatio Bulbi

- Extremely rare, usually unilateral
- Whole globe is luxated out of the orbital cavity
- Severe chemosis of the conjunctiva, sometimes with anterior segment ischemia
- In case of delay, there is exposure keratitis and eventual corneal ulcer **(Figs 14.25.1 and 14.25.2)**
- *Cause:* Crouzon's syndrome, blunt trauma, malignant thyroid ophthalmopathy, psychiatric problem *(oedipism)*, etc.
- *Treatment:* Required urgently to save the globe.

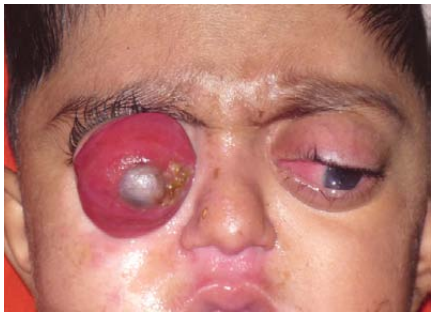


Fig. 14.25.1: Luxatio bulbi—severe chemosis

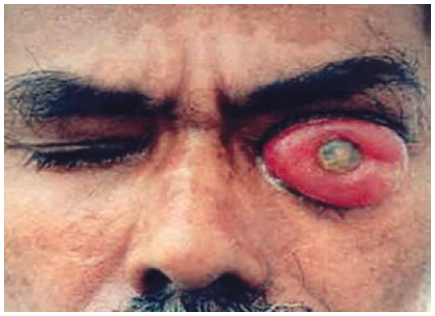


Fig. 14.25.2A: Luxatio bulbi

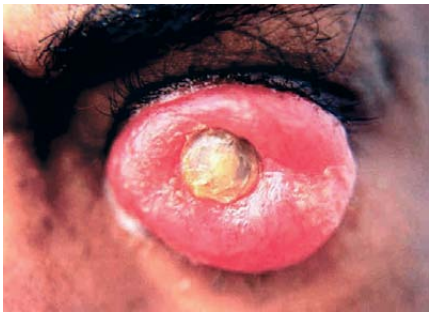


Fig. 14.25.2B: Luxatio bulbi—severe chemosis

Orbital Fat Prolapse

- Not so uncommon, occurs in old age
- Unilateral or bilateral, often mistaken as lymphoid hyperplasia
- Associated with defects in orbital septum and baggy eyelids **(Figs 14.26.1 and 14.26.2)**.

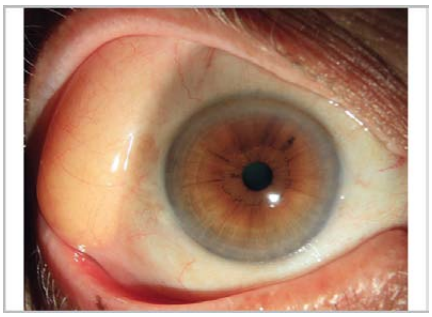


Fig. 14.26.1A: Orbital fat prolapse—RE

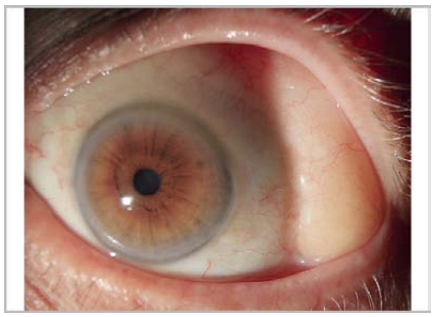


Fig. 14.26.1B: Orbital fat prolapse—LE

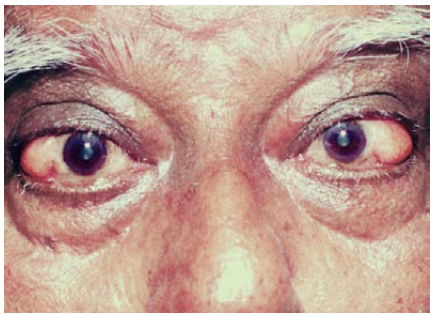


Fig. 14.26.2: Prolapsed orbital fat—baggy lids

Contracted Socket

- A good anophthalmic socket has adequate superior and inferior fornices **(Fig. 14.27.1)**, without any symblepharon and discharge and it should fit the artificial eye with good cosmesis **(Figs 14.27.2A to C)**
- *Contracted socket* occurs due to chronic inflammation, trauma, prior ocular surgeries, cicatricial ocular pemphigoid, etc. **(Fig. 14.27.3)**
- It results in conjunctival shortage, leading to shallowing of the fornices and inability to wear the artificial shell
- *According to severity it may be of following types:*
  - *Grade I:* Shallowing of inferior fornix **(Figs 14.27.4A and B)**

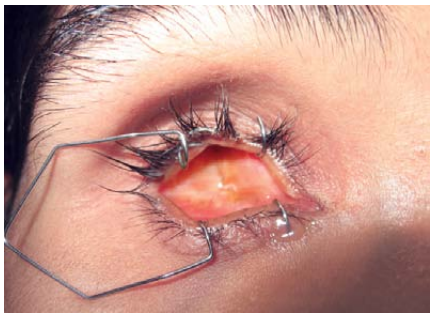


Fig. 14.27.1: Good anophthalmic socket

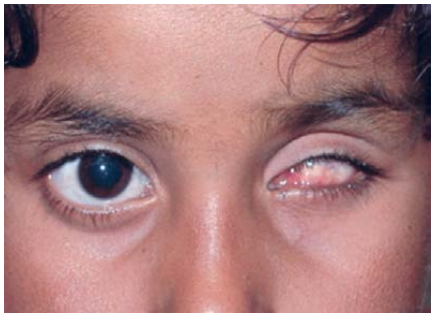


Fig. 14.27.2A: Good anophthalmic socket

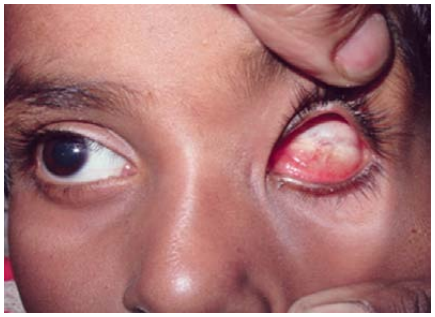


Fig. 14.27.2B: Good anophthalmic socket

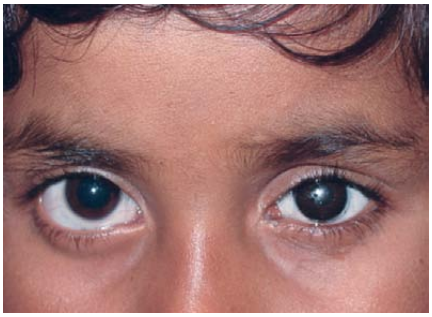


Fig. 14.27.2C: Good anophthalmic socket—good cosmesis

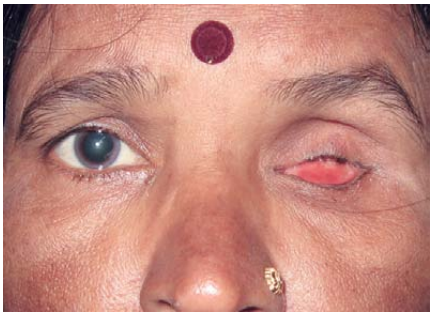


Fig. 14.27.3: Contracted socket

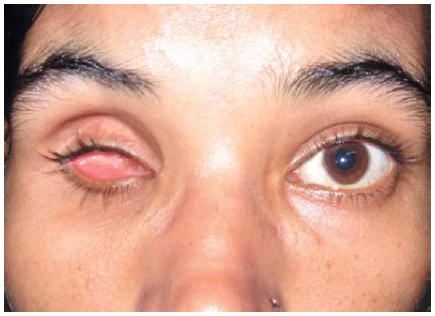


Fig. 14.27.4A: Contracted socket—grade I

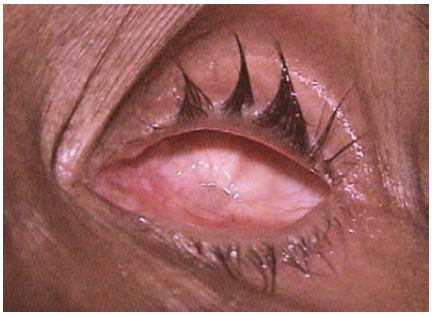


Fig. 14.27.4B: Contracted socket—grade I

- *Grade II:* Shallowing of both inferior and superior fornices **(Fig. 14.27.5)**
- *Grade III:* Formation of symblepharon bands **(Fig. 14.27.6)**
- *Grade IV:* Gross contracture (especially, after radiotherapy) of whole socket **(Figs 14.27.7 and 14.27.8)**
- *Grade V:* Wet/discharging socket **(Figs 14.27.9A and B)**

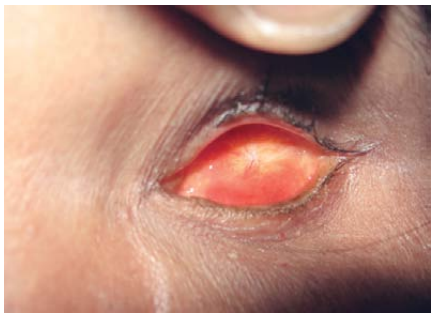


Fig. 14.27.5: Contracted socket—grade II

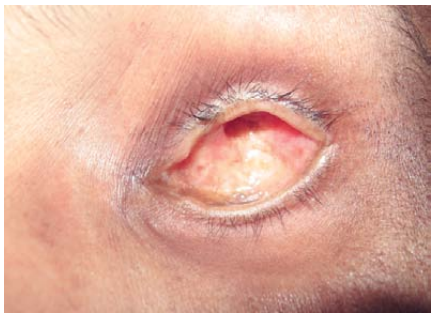


Fig. 14.27.6: Contracted socket—grade III

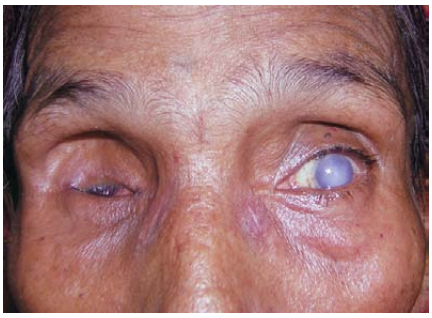


Fig. 14.27.7: Anophthalmic socket—grade IV

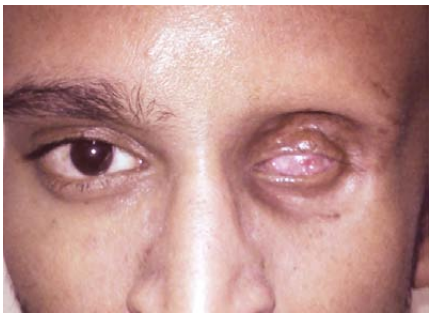


Fig. 14.27.8A: Contracted socket—grade IV—after radiotherapy

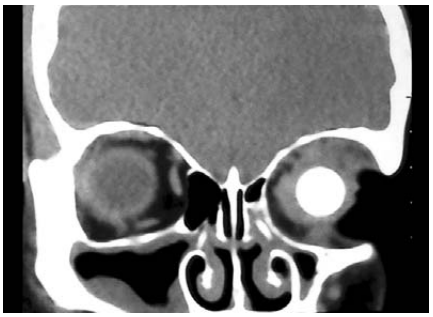


Fig. 14.27.8B: Contracted socket—grade IV—after radiotherapy

- *Treatment:* Reconstructive procedures like—fornix forming sutures; buccal mucous membrane graft; dermis fat graft, etc. **(Figs 14.27.10A to C)**.
- *Exposed orbital implants:* Not so uncommon, especially with acrylic ball (sphere) due to recurrent orbital infection **(Figs 14.27.11 and 14.27.12)**.

Fig. 14.27.9A: Anophthamic socket—grade V—discharging socket

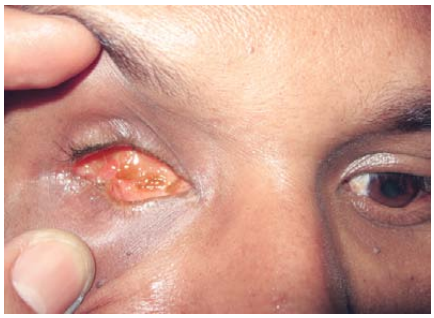


Fig 14.27.9B: Anophthamic socket - grade V

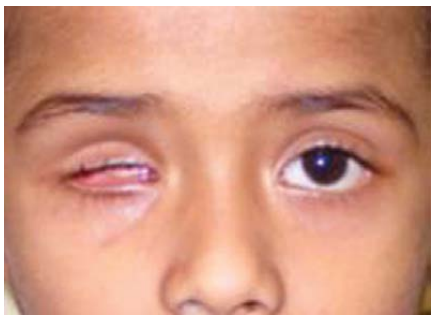


Fig. 14.27.10A: Contracted socket—preoperative

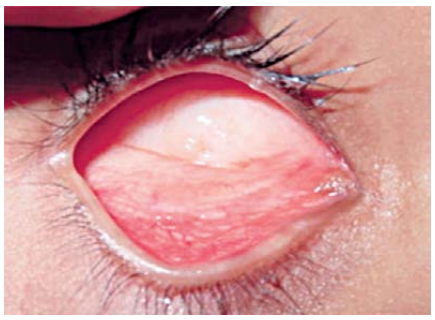


Fig. 14.27.10B: Contracted socket after reconstruction surgery—dermis fat graft

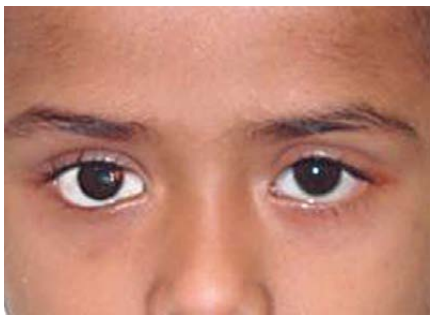


Fig. 14.27.10C: Contracted socket—good cosmesis after reconstruction

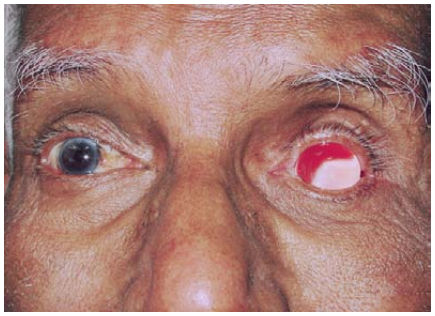


Fig. 14.27.11: Exposure of orbital implant

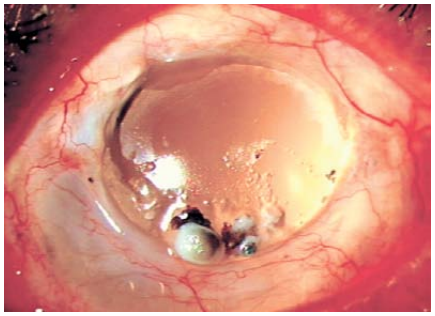


Fig. 14.27.12: Exposure of orbital implant—fungal infection

CHAPTER

15 Ocular Motility Disturbances and Squint

CHAPTER OUTLINE

Pseudostrabismus (Apparent Squint)

Concomitant Strabismus

- Convergent strabismus (esotropia)
- Divergent strabismus (exotropia)
- Alternating strabismus
- Vertical strabismus
- Inferior oblique overaction
- Dissociated vertical deviation

Paralytic Strabismus

- Total ophthalmoplegia
- External ophthalmoplegia
- Third (oculomotor) nerve palsy
- Fourth (trochlear) nerve palsy
- Sixth (abducens) nerve palsy

Miscellaneous Ocular Movement Disorders

- Duane's retraction syndrome
- Superior oblique tendon sheath syndrome (Brown's syndrome)
- Double elevator palsy
- Restrictive strabismus
- Congenital fibrosis of extraocular muscles (CFEOM)
- Orbital fractures
- Strabismus fixus

Pseudostrabismus (Apparent Squint)

- Here, the visual axes are in fact parallel, but the eyes seem to have squint.
- *Causes:*
  - *Prominent epicanthic folds:* May simulate a convergent squint **(Fig. 15.1.1)**
  - *Telecanthus:* Also simulates a convergent squint **(Figs 15.1.2A and B)**
  - *Hypertelorism:* May simulate a divergent squint **(Fig. 15.1.3 and *see* Fig. 14.1.6)**
  - *Hypermetropia:* An apparent divergent squint due to large angle kappa
  - *Myopia:* An apparent convergent squint due to smaller or even negative angle kappa
- Squint in these conditions may be easily excluded by checking the relative position of corneal light reflections and also by other means.

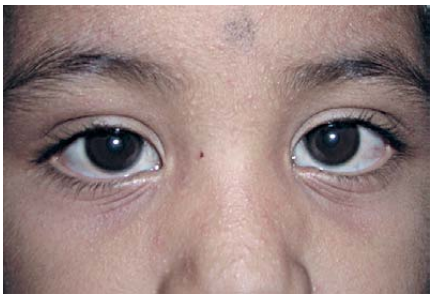


Fig. 15.1.1: Epicanthic folds—pseudo-convergent squint

Fig. 15.1.2A: Telecanthus—pseudo-convergent squint

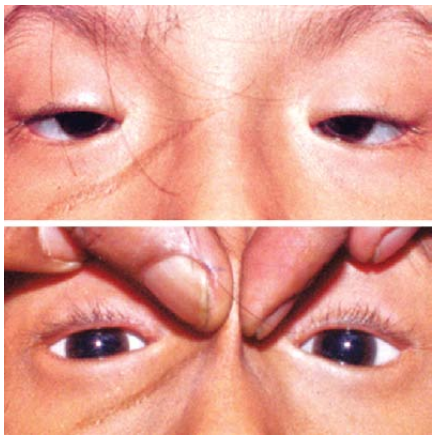


Fig. 15.1.2B: Telecanthus—pseudo-convergent squint

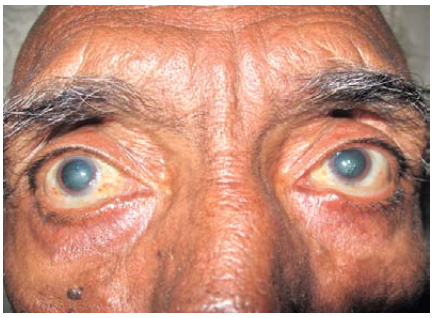


Fig. 15.1.3: Hypertelorism—pseudo-divergent squint

Concomitant Strabismus

- Angle of deviation remains the same in all directions of gaze, irrespective of eyes of fixation
- May be *monocular* or *alternating*
- In 85% cases the squint is *monocular,* in the sense one eye habitually takes up fixation and the other eye is squinting eye.

Convergent Strabismus (Esotropia)

- One eye always deviated inwards
- Typically develops in early childhood before the binocular reflexes are firmly established **(Fig. 15.2.1)**
- Amblyopia is a frequent association

Types of Concomitant Esotropia

- ***Infantile esotropia:***
  - Develops before the age of 6 months **(Fig. 15.2.2)**
  - Large and stable angle
  - May have alternate or cross-fixation **(Figs 15.2.3 and 15.2.4)**
  - Normal refraction for age
  - Poor potential for binocular vision
  - Inferior oblique overaction may be present initially or develop later **(Fig. 15.2.5)**.

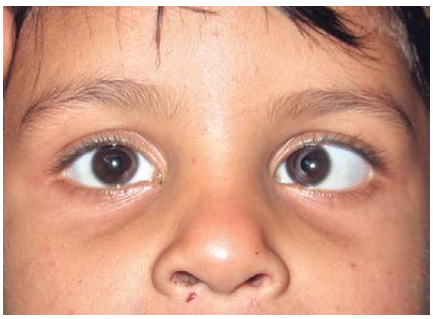


Fig. 15.2.1: Esotropia

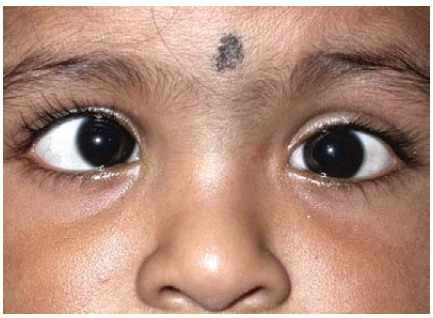


Fig. 15.2.2: Infantile esotropia

Fig. 15.2.3: Infantile esotropia—alternate fixation

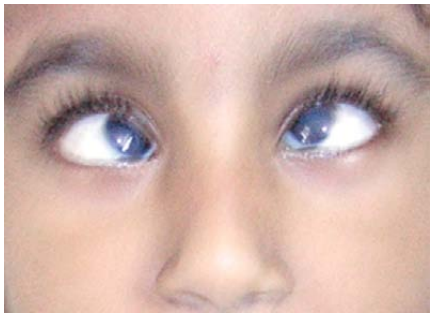


Fig. 15.2.4: Infantile esotropia—cross-fixation

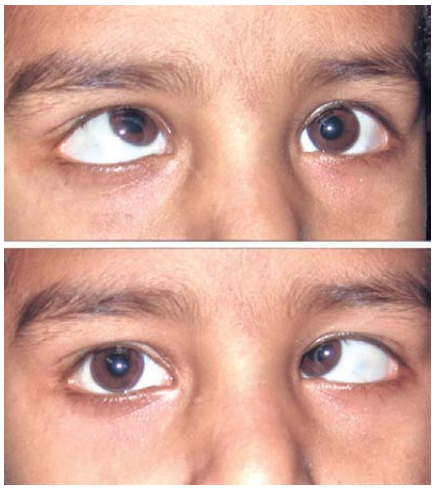


Fig. 15.2.5: Infantile esotropia—with inferior oblique overaction

- ***Accommodative esotropia:***
  - *Fully accommodative:* Spectacles fully correct the deviation **(Figs 15.3.1A and B)**
  - *Partially accommodative:* Spectacles partly correct the deviation **(Figs 15.3.2A and B)**
  - Onset is between 2 and 3 years, rarely earlier
  - Hypermetropia is a common finding
  - Atropine cycloplegic refraction is a must.
- ***Nonaccommodative esotropia:***
  - Onset during childhood, but after 6 months of age
  - Insignificant refractive error
  - No excess accommodative element
  - Angle of deviation same for distance and near fixation **(Fig. 15.3.3)**.

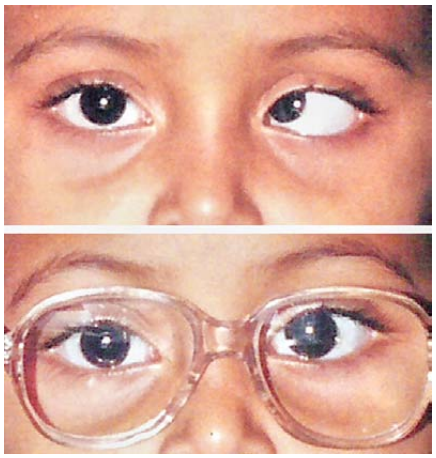


Fig. 15.3.1A: Fully accommodative esotropia—fully corrected by spectacles

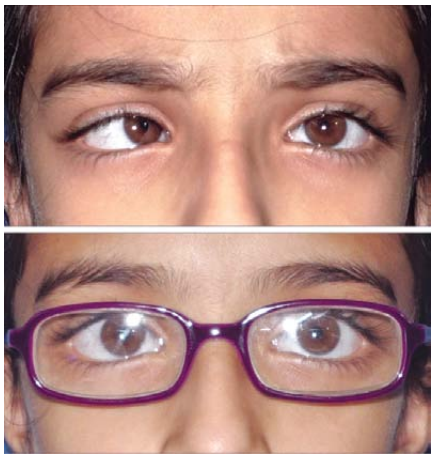


Fig. 15.3.1B: Fully accommodative esotropia—fully corrected by spectacles

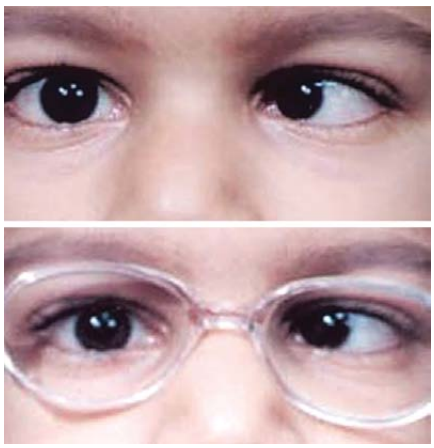


Fig. 15.3.2A: Partially accommodative esotropia

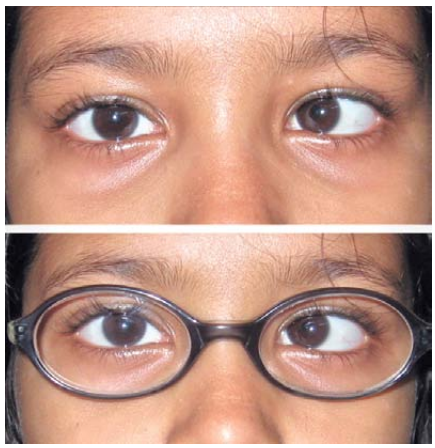


Fig. 15.3.2B: Partially accommodative esotropia

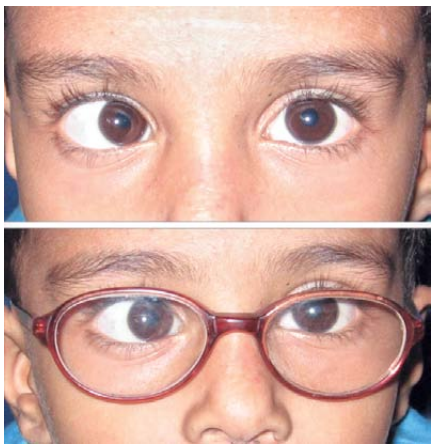


Fig. 15.3.3: Non-accommodative esotropia

- ***Nonrefractive accommodative esotropia:***
  - Onset between 6 months and 3 years
  - With high AC/A ratio, with increased accommodative convergence
  - Normal near point of accommodation
  - No significant refractive error
  - Straight eyes for distance, but esotropia for near.
- ***Sensory esotropia:***
  - Caused by unilateral reduction in visual acuity at an early age (before 6 years) which interferes or abolishes fusion
  - Other eye remains normal
  - *Causes:* Congenital cataract, corneal opacity, coloboma, microphthalmos, etc. **(Figs 15.4.1 and 15.4.2).**

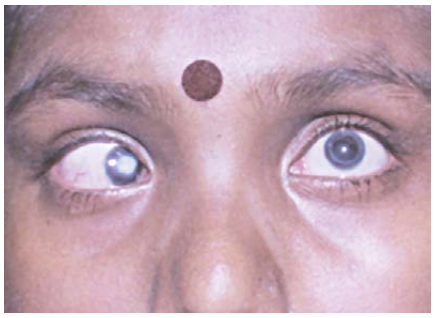


Fig. 15.4.1A: Sensory esotropia—corneal scar

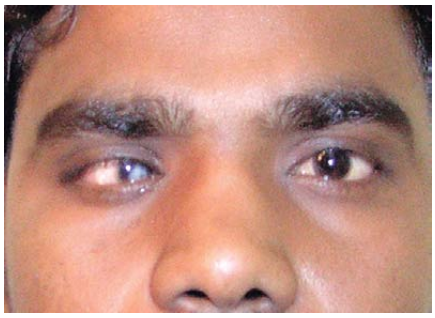


Fig. 15.4.1B: Sensory esotropia—corneal scar

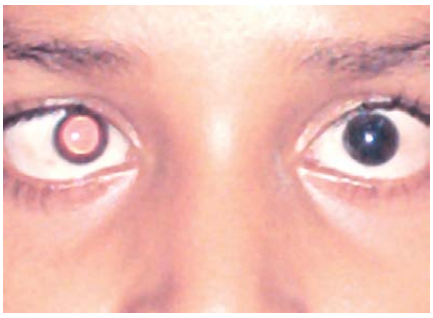


Fig. 15.4.2: Sensory esotropia—microphthalmos

- ***Consecutive esotropia:***
  - Occurs after surgical overcorrection of an exotropia or slippage of lateral rectus muscle during surgery **(Figs 15.5.1 and 15.5.2).**

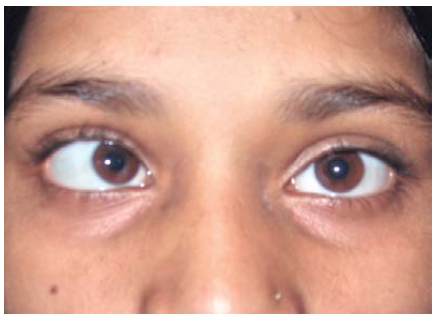


Fig. 15.5.1: Consecutive esotropia following overcorrection of exotropia

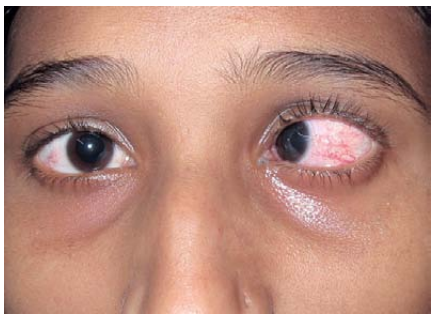


Fig. 15.5.2: Consecutive esotropia following lateral rectus slip during surgery

- ***Concomitant esotropia may be:***
  - *Small angle* **(Figs 15.6.1A and B)**
  - *Medium angle* **(Fig. 15.6.2)**
  - *Large angle* **(Fig. 15.6.3)**
  - *Extra large angle* **(Fig. 15.6.4).**

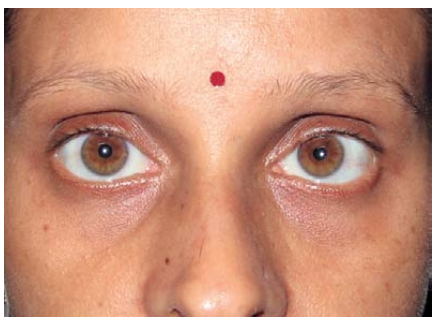


Fig. 15.6.1A: Esotropia (LE)—small angle

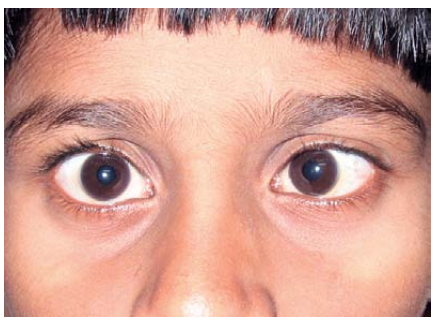


Fig. 15.6.1B: Esotropia (LE)—small angle

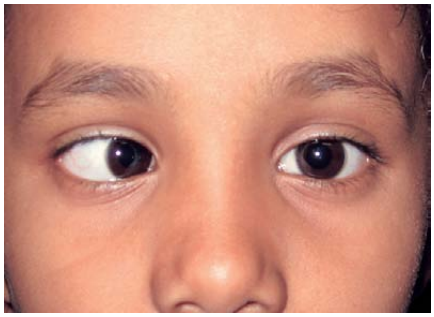


Fig. 15.6.2: Esotropia (RE)—medium angle

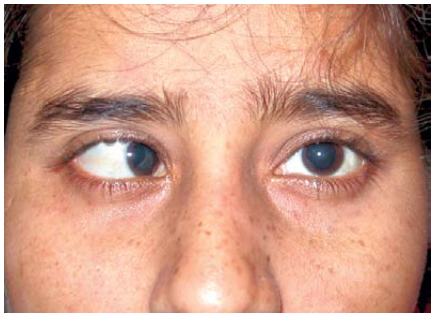


Fig. 15.6.3: Esotropia (RE)—large angle

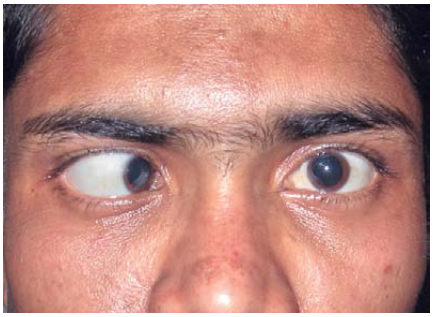


Fig. 15.6.4: Esotropia (RE)—extra large angle esotropia

Divergent Strabismus (Exotropia)

- One eye always deviates outwards
- More likely to be latent or intermittent than esotropia
- Constant exotropia is usually seen in older patients
- Amblyopia is less common

Types

- ***Congenital exotropia***
  - Much less common than congenital esotropia
  - Commonly seen in craniofacial anomalies like, hypertelorism **(Fig. 15.7.1)**, Crouzon's syndrome **(Fig. 15.7.2).**
- ***Basic exotropia:***
  - Initially intermittent **(Figs 15.8.1A and B)** then becomes constant **(Fig. 15.8.2)**
  - According to the degree of deviation, it may be—small angle **(Figs 15.8.3A and B)**, medium

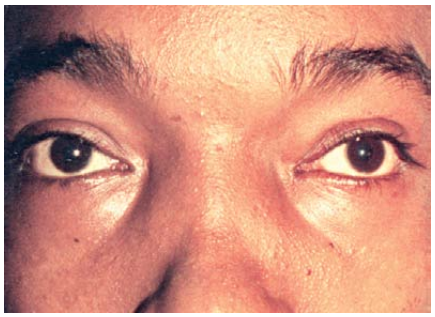


Fig. 15.7.1: Hypertelorism—exotropia

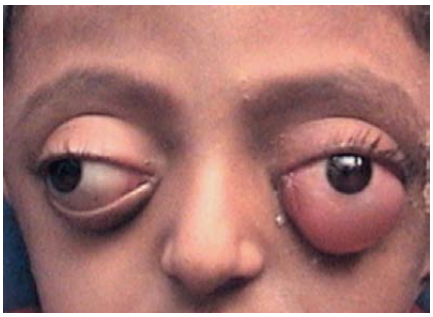


Fig. 15.7.2: Crouzon's syndrome—exotropia

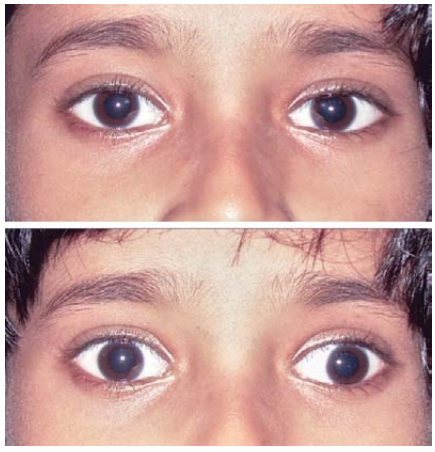


Fig. 15.8.1A: Intermittent exotropia

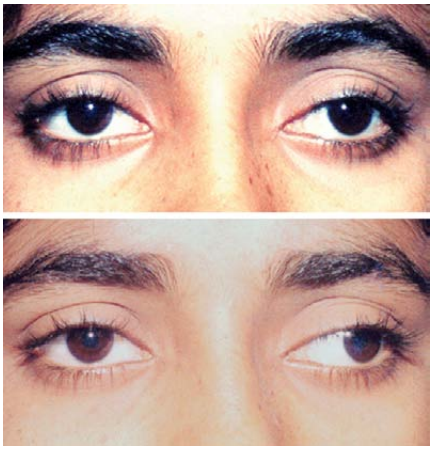


Fig. 15.8.1B: Intermittent exotropia for distant

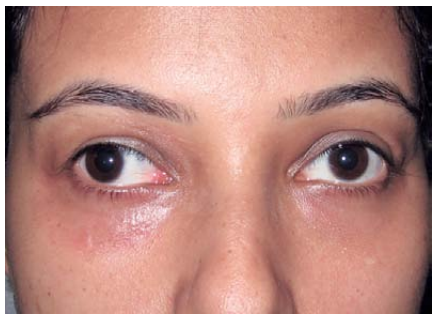


Fig. 15.8.2: Basic exotropia—constant

Fig. 15.8.3A: Basic exotropia—small angle

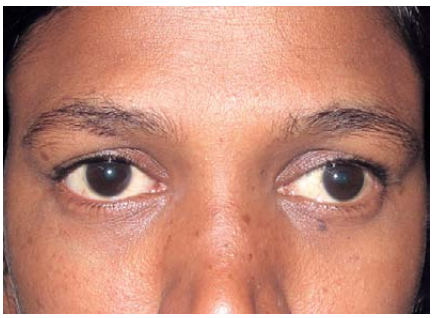


Fig. 15.8.3B: Basic exotropia—small angle

angle **(Fig. 15.8.4)**, large angle **(Fig. 15.8.5)** or extra large angle **(Figs 15.8.6A and B)** exotropia
- Angle of deviation is more for distant fixation than near
- Myopia may be a common association.

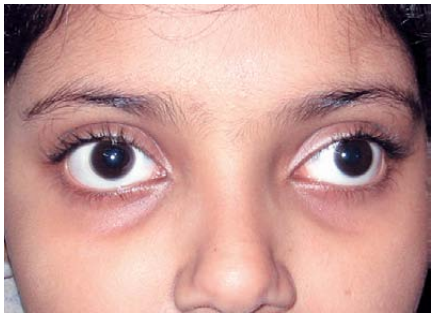


Fig. 15.8.4: Basic exotropia—medium angle

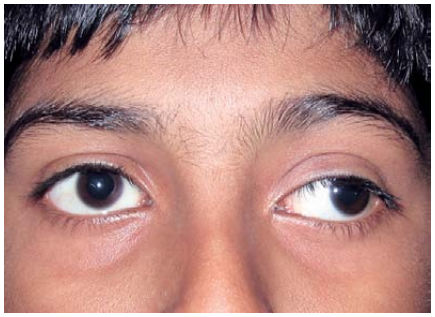


Fig. 15.8.5: Basic exotropia—large angle

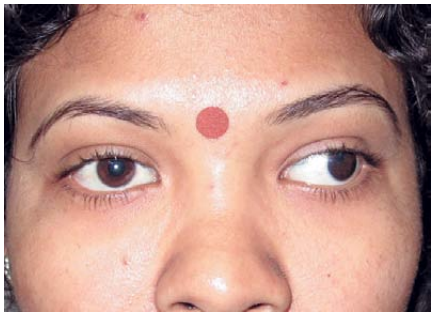


Fig. 15.8.6A: Basic exotropia—extra large angle

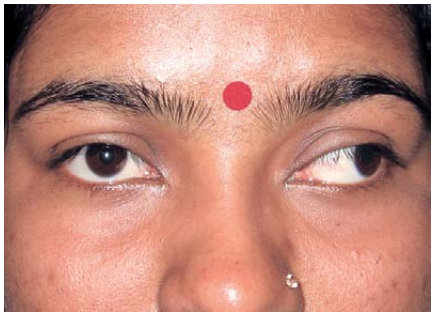


Fig. 15.8.6B: Basic exotropia—extra large angle

- ***Sensory exotropia:***
  - Caused by uniocular sensory visual deprivation usually after 6 years of age
  - Most commonly by cataract, aphakia, optic atrophy, corneal opacity, after injury, etc. **(Figs 15.9.1 to 15.9.5).**
- ***Consecutive exotropia:***
  - Following overcorrection of a convergent squint.

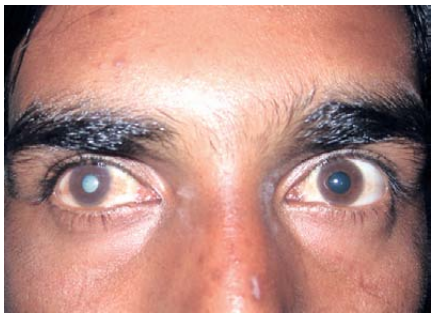


Fig. 15.9.1: Sensory exotropia—cataract—RE

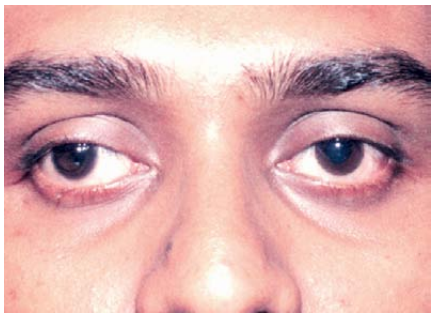


Fig. 15.9.2: Sensory exotropia—aphakia—note RGP contact lens in RE

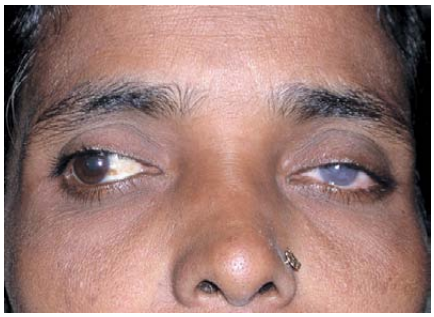


Fig. 15.9.3: Sensory exotropia—optic atrophy—RE

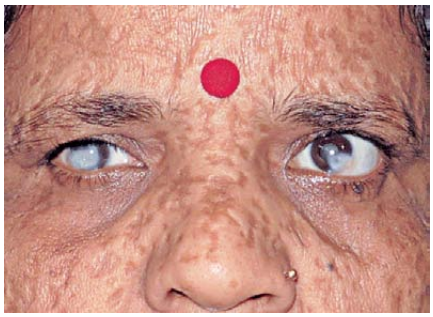


Fig. 15.9.4: Sensory exotropia—corneal opacity—RE

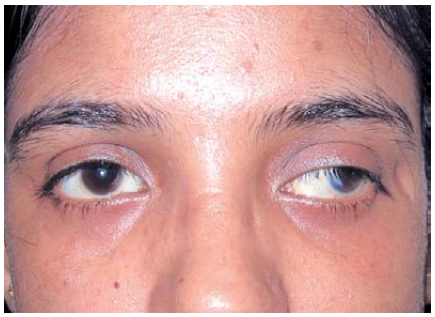


Fig. 15.9.5: Sensory exotropia—traumatic aphakia—LE

Alternating Strabismus

- Alternating squint means when one eye fixes, the other eye deviates
- Either of the eyes can adopt fixation alternately and freely
- Visual acuity remains normal or near normal in each eye
- Chance of amblyopia is least
- Alternating squint may be convergent **(Figs 15.10.1 and 15.10.2)** or divergent type **(Figs 15.10.3 and 15.10.4)**.

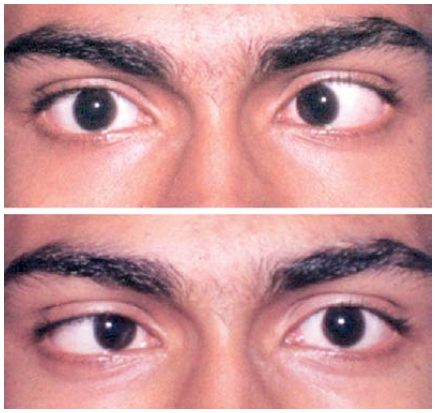


Fig. 15.10.1: Alternate esotropia—small angle

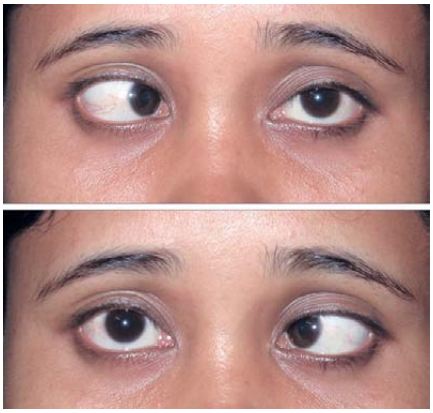


Fig. 15.10.2: Alternate esotropia—large angle

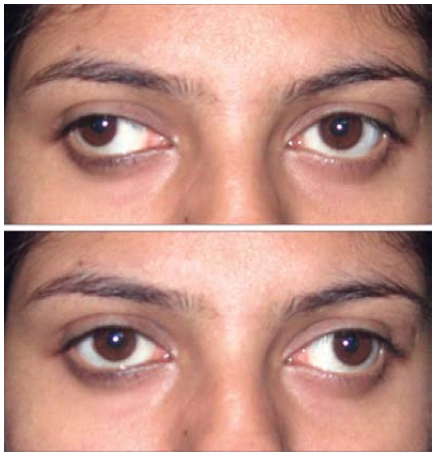


Fig. 15.10.3: Alternate exotropia—small angle

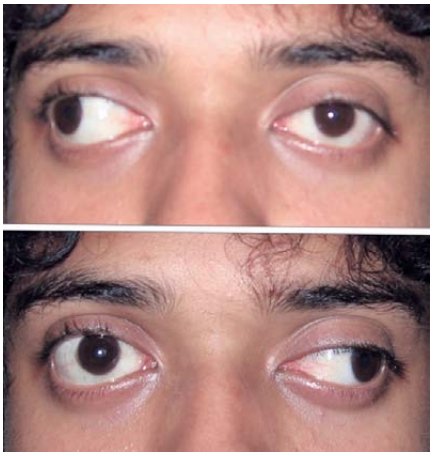


Fig. 15.10.4: Alternate exotropia—large angle

Vertical Strabismus

- ***Hypertropia***
  - Hyperdeviation of the nonfixing eye in primary position **(Fig. 15.11.1)**
  - May be primarily caused by compromised ipsilateral depressors **(Fig. 15.11.2)**
  - Or secondary to compromised contralateral elevators with fixing paretic eye or congenital fibrosis of extraocular muscles **(Figs 15.11.3 and 15.11.4)**.

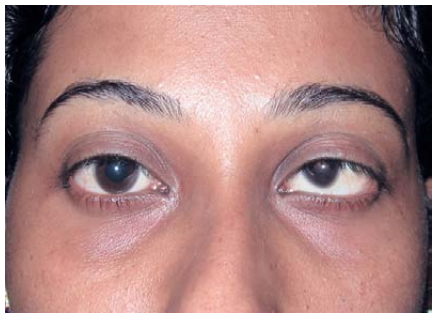


Fig. 15.11.1: Left hypotropia—RE fixing—LE with uveal coloboma

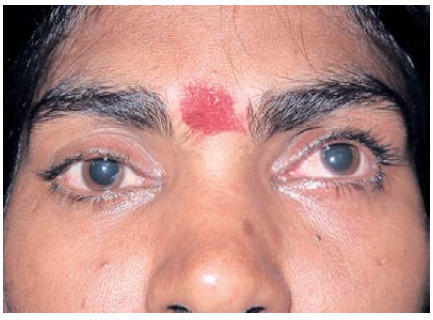


Fig. 15.11.2: Left hypertropia—right eye fixing

- ***Hypotropia***
  - Hypodeviation of nonfixing eye in primary gaze
  - May be primary or secondary, depending upon wheather non-paretic or paretic eye is fixing **(Figs 15.11.5 to 15.11.7).**

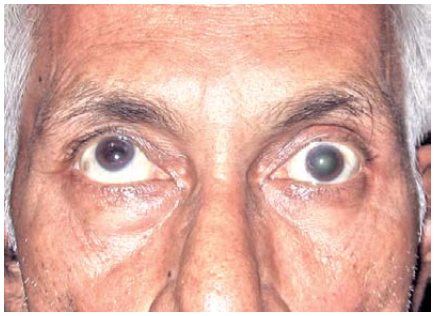


Fig. 15.11.3: Right hypertropia—left eye fixing

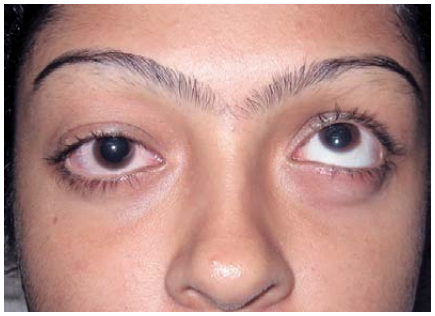


Fig. 15.11.4: Left hypertropia—congenital fibrosis of extraocular muscle

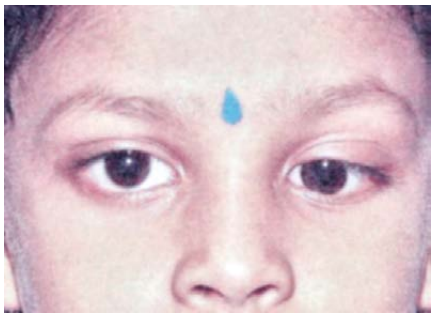


Fig. 15.11.5: Left hypotropia—RE fixing

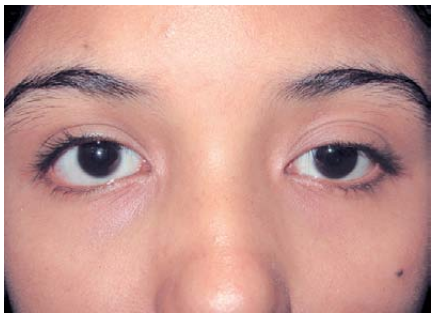


Fig. 15.11.6: Left hypotropia—RE fixing

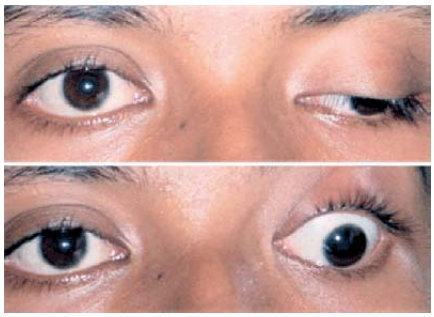


Fig. 15.11.7: Left hypotropia—RE fixing

Inferior Oblique Overaction

- Inferior oblique overaction may be unilateral or bilateral **(Fig. 15.12.1)**
- Primary overaction are frequently bilateral and cause upshooting in adduction **(Fig. 15.12.2)**

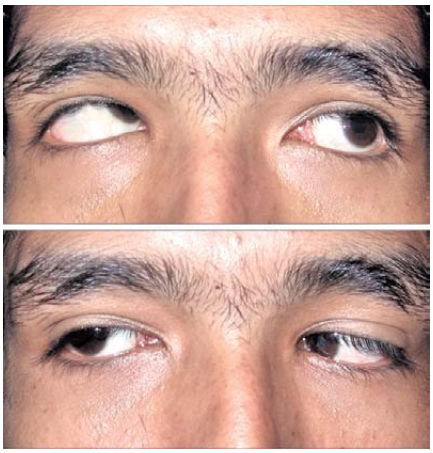


Fig. 15.12.1: Unilateral inferior oblique (IO) overaction—RE

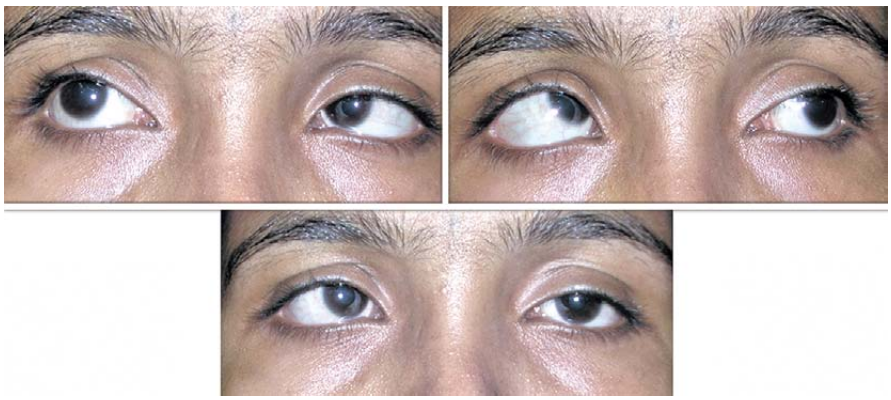


Fig. 15.12.2: Bilateral inferior oblique overaction

- Common with esodeviations **(Fig. 15.12.3)**
- Associated with inferior oblique overaction with V-phenomenon in esodeviation **(Fig. 15.12.4)**.

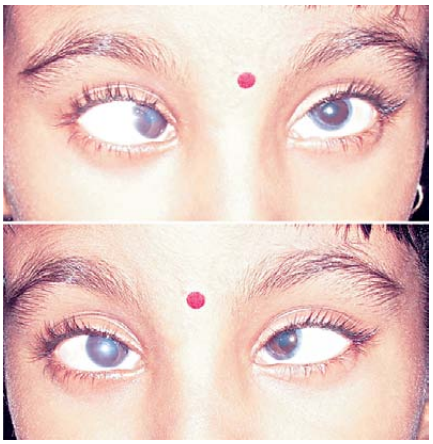


Fig. 15.12.3: Infantile esotropia—bilateral inferior oblique overaction

Dissociated Vertical Deviation

- It is a slow upward movement of one eye, with cortical suppression of the eye while it is deviated. On returning downward to take up fixation, the DVD slow movement is reversed **(Fig. 15.12.5)**
- Affects both eyes **(Fig. 15.12.6)**, but can occur unilaterally or asymmetrically and appreciated better during cover test
- It is often associated with latent or manifest nystagmus and, as well as occurring with infantile esotropia
- DVD is often mistaken for overaction of the inferior oblique extraocular muscles.

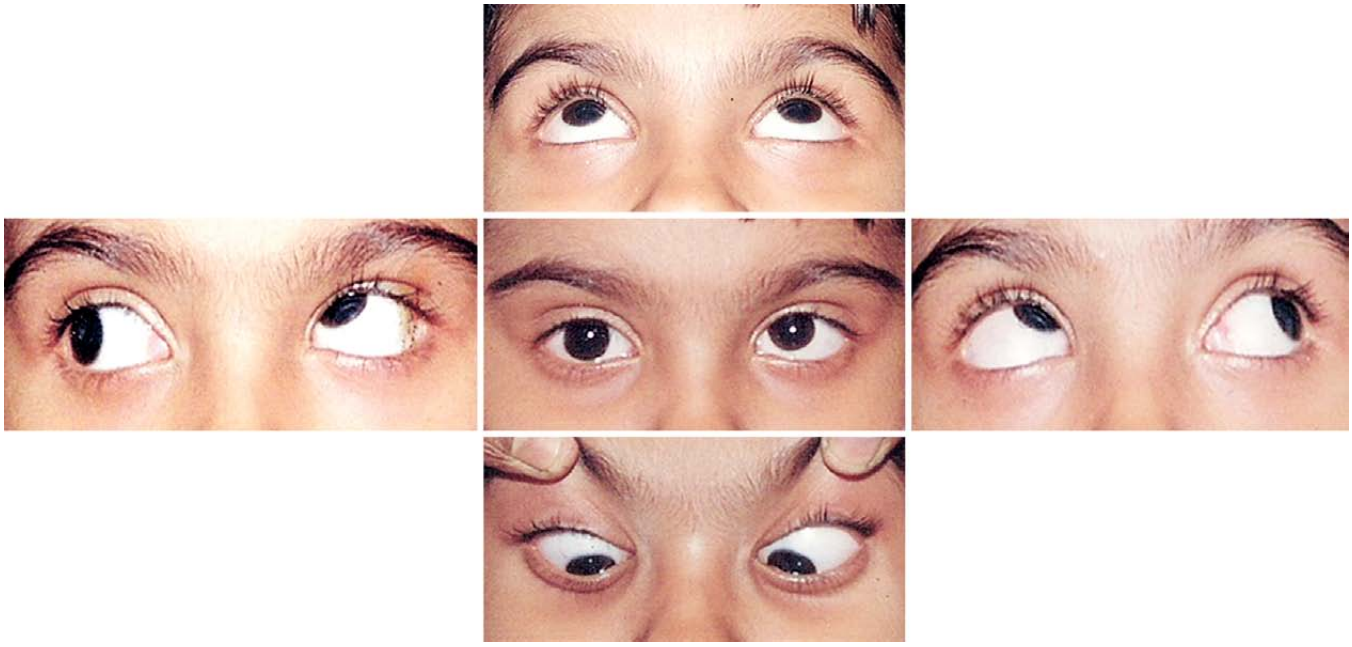


Fig. 15.12.4: Bilateral inferior oblique overaction—V-phenomenon

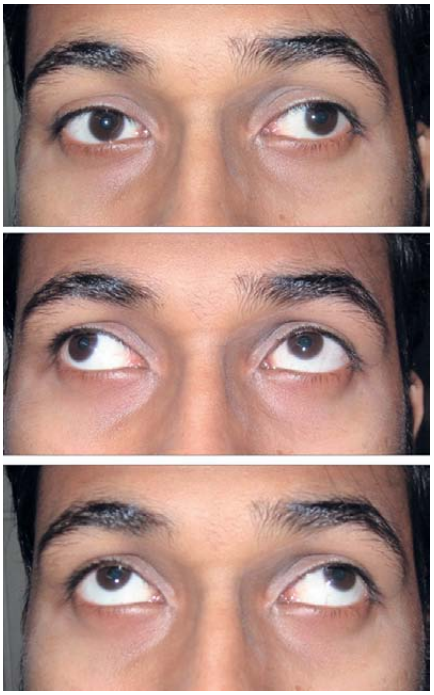


Fig. 15.12.5: Dissociated vertical deviation

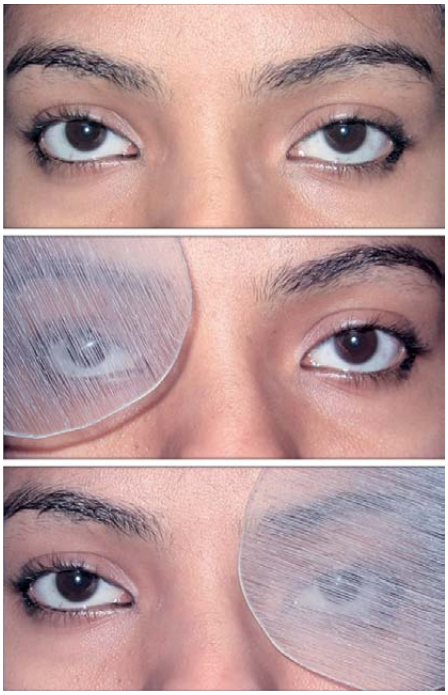


Fig. 15.12.6: Dissociated vertical deviation–cover test

Paralytic Strabismus

- Misalignment of the visual axes as a result of paresis, or paralysis of one or more extraocular muscles **(Figs 15.13.1 and 15.13.2)**
- Impaired movement in the field of action of muscles
- Angle of deviation varies in different direction of gaze and with fixation of eyes
- Secondary angle of deviation is more than primary deviation
- *Compensatory head posture:* In paralytic squint, to neutralize diplopia the chin may be elevated or depressed, the face is turned to right or left side, and the head may be tilted to the right or left shoulder *(ocular torticollis)* **(Figs 15.13.3 and 15.13.4)**
  - In paralysis of horizontal rectus muscle, the face is turned to field of action of the paralyzed muscle
  - In case of cyclovertical muscle palsy, it is more complicated and less valuable diagnostically

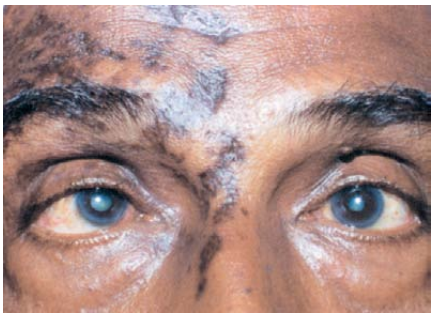


Fig. 15.13.1: Paralytic squint following HZV ophthalmicus

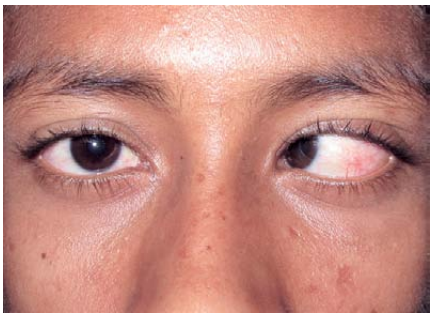


Fig. 15.13.2: Paralytic squint following left lateral rectus palsy

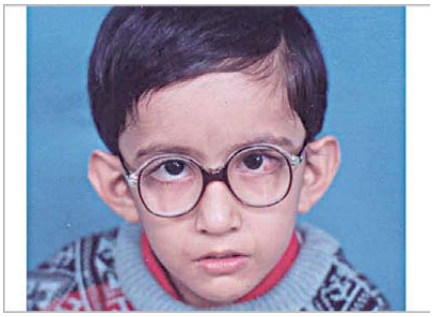


Fig. 15.13.3: Compensatory head posture

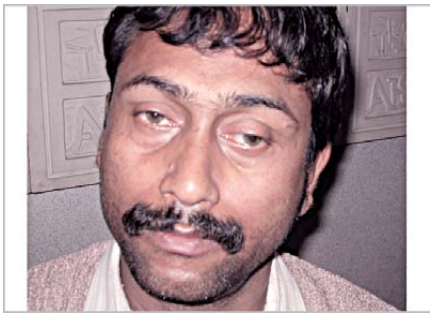


Fig. 15.13.4: Compensatory head posture in cyclovertical muscle palsy

- Visual acuity remains normal in both eyes, without amblyopia
- *Treatment:* To give relief from diplopia; investigations to find out the cause and observation.

Total Ophthalmoplegia

- Means involvement of both extrinsic and intrinsic muscles
- Lesion is in the cavernous sinus or in the superior orbital fissure and in bilateral cases; the lesion is widespread in the brainstem due to vascular or inflammatory cause
- *Clinical signs* **(Fig. 15.14.1)**:
  - Ptosis
  - Eyeball is slightly proptosed and divergent
  - No movement of the eyeball
  - Fixed dilated pupil
  - Total loss of accommodation.

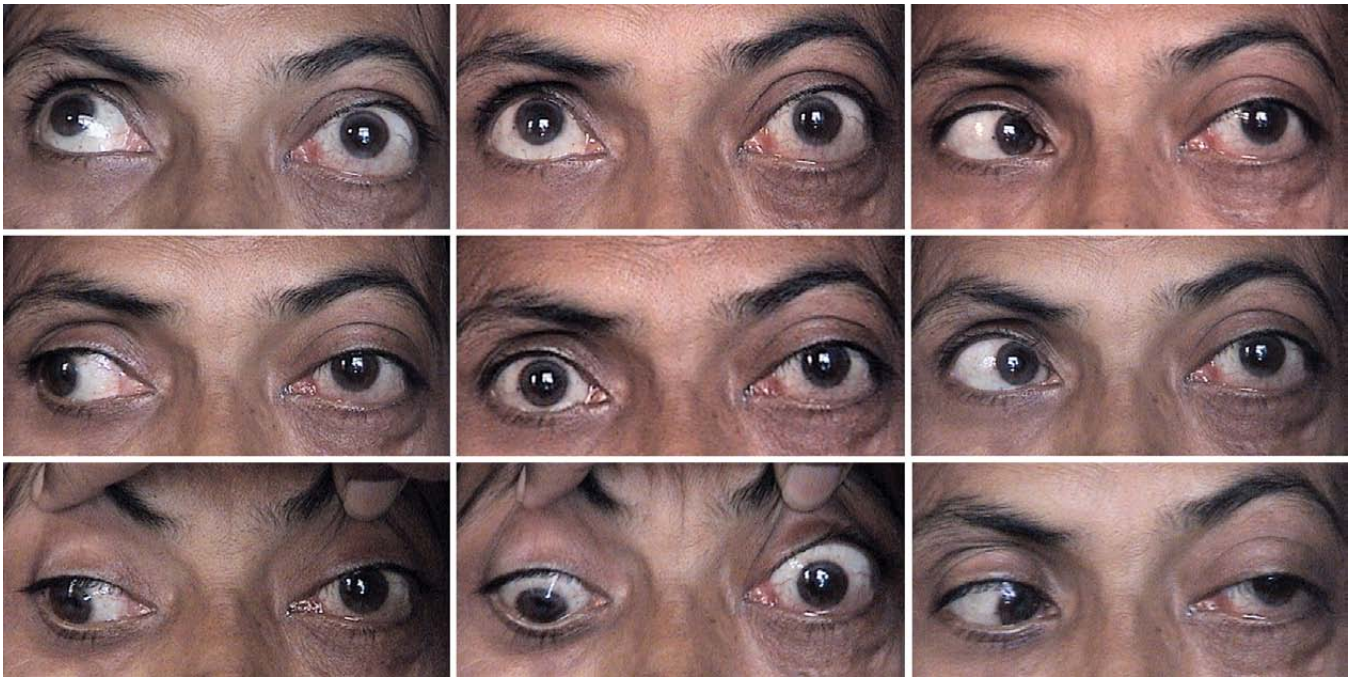


Fig. 15.14.1: Total ophthalmoplegia

External Ophthalmoplegia

- Extrinsic muscles along with levator palpebrae superioris
- Lesion without affecting the Edinger-Westphal nucleus which supplies the intrinsic muscles **(Figs 15.15.1 and 15.15.2)**
- *Other causes:* Chronic progressive external ophthalmoplegia (CPEO) **(Figs 15.15.3 and 15.15.4)**, myasthenia gravis **(Figs 15.15.5A and B)**
- Pupillary reactions and accommodation are normal.

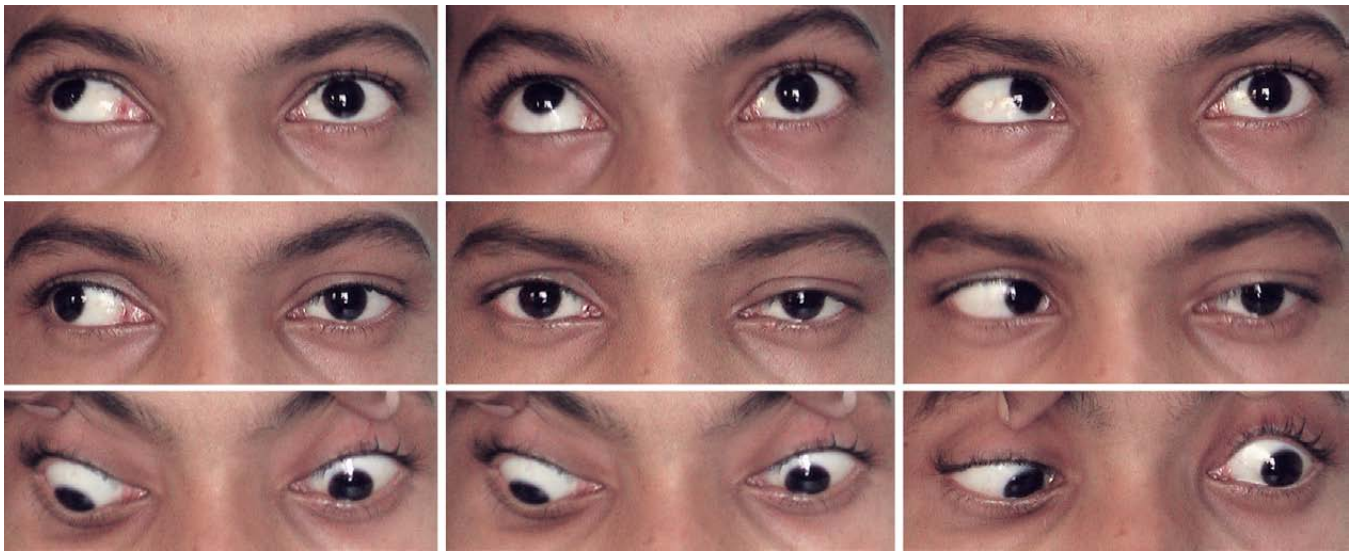


Fig. 15.15.1: External ophthalmoplegia

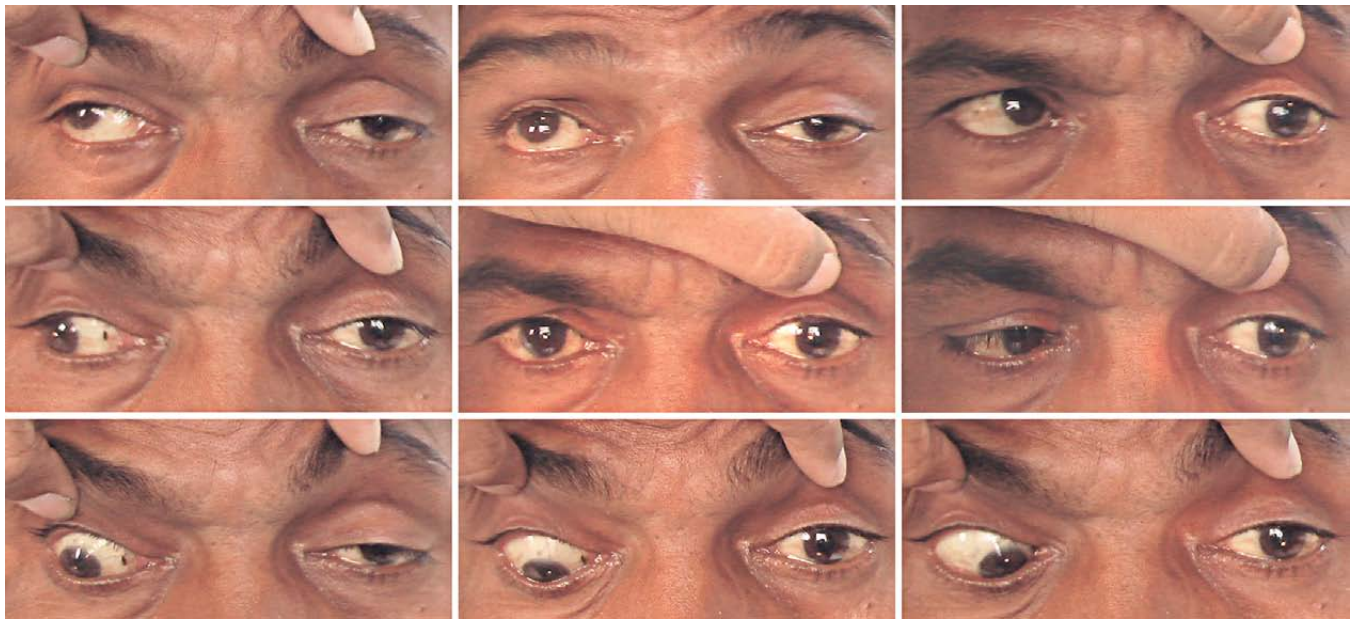


Fig. 15.15.2: External ophthalmoplegia

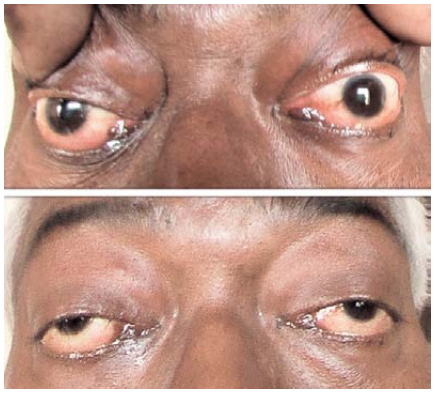


Fig. 15.15.3: External ophthalmoplegia—CPEO

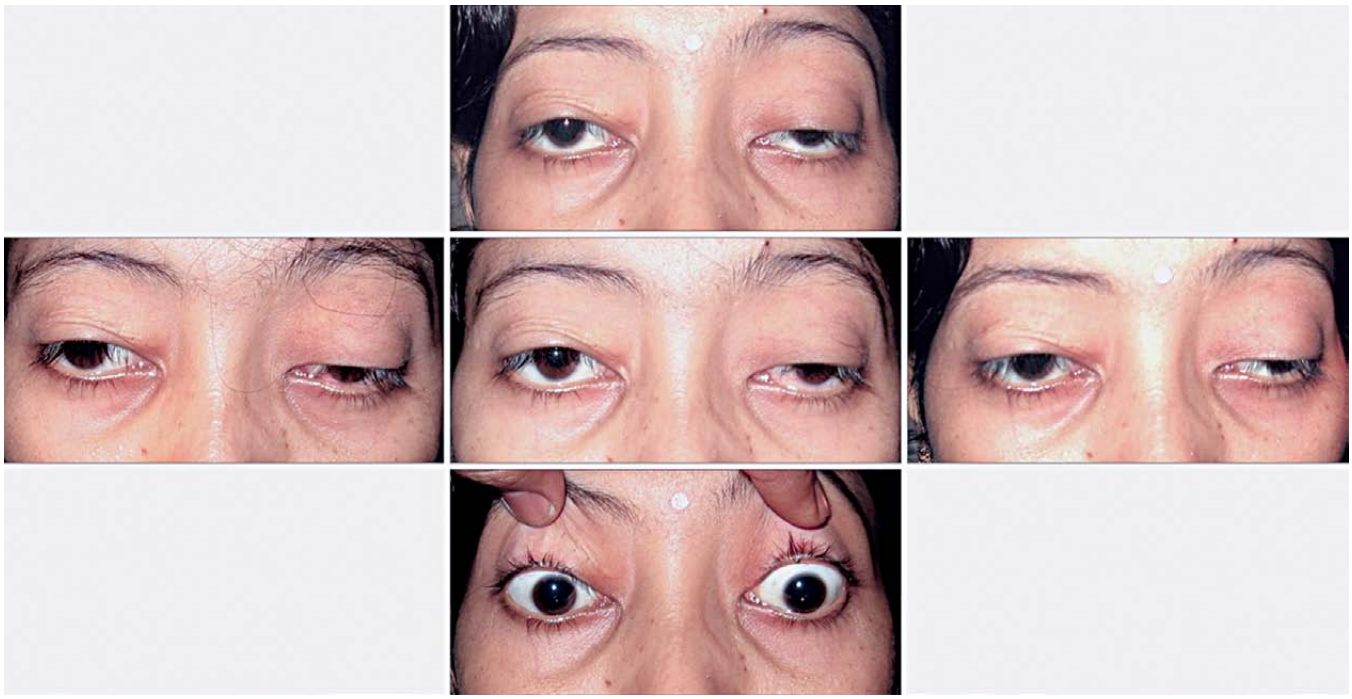


Fig. 15.15.4: External ophthalmoplegia—CPEO

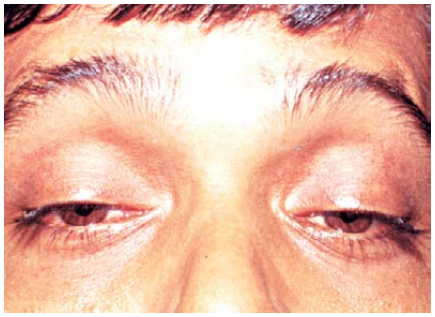


Fig. 15.15.5A: External ophthalmoplegia—myasthenia gravis—before injection

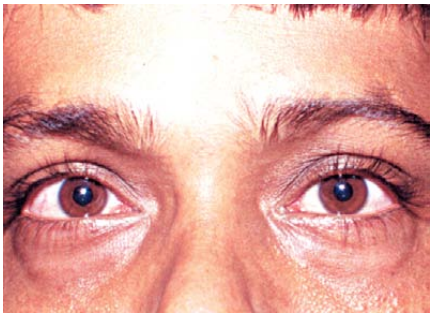


Fig. 15.15.5B: External ophthalmoplegia—myasthenia gravis after injection of edrophonium (Tensilon)

Third (Oculomotor) Nerve Palsy

- Ptosis
- Eyeball rotates outwards (divergent) and slightly downwards **(Fig. 15.16.1)**
- Intorsion of the eyeball on attempted downgaze
- Ocular movements are restricted in all gaze except laterally
- Pupil is dilated and fixed.

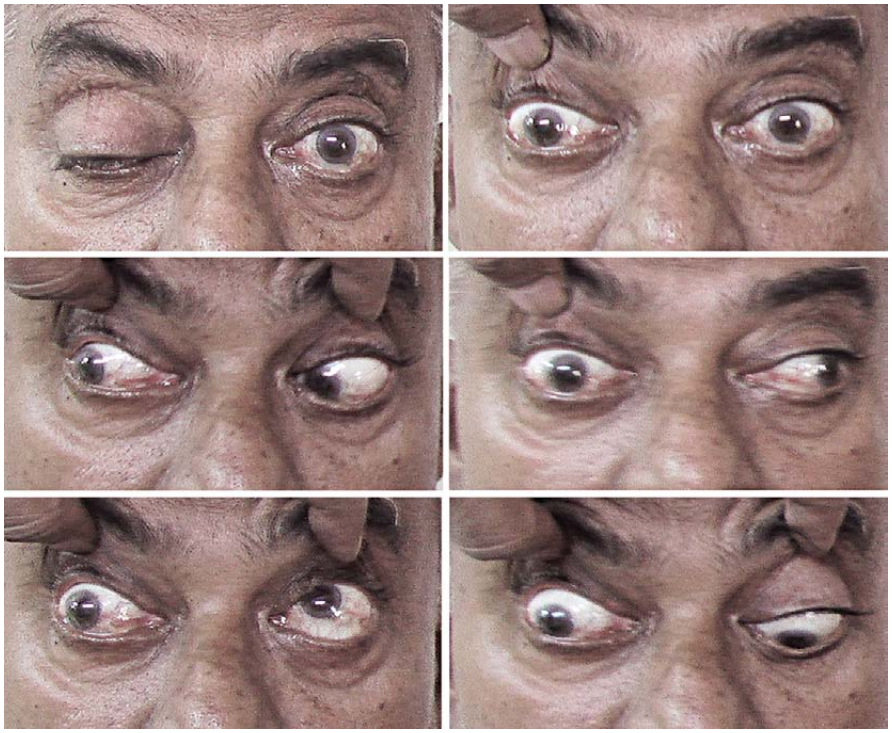


Fig. 15.16.1: Right third nerve palsy—complete

Fourth (Trochlear) Nerve Palsy

- Trauma is the most common cause of isolated fourth nerve palsy
- Abnormal head posture
- Eyeball deviated upwards and inwards (ipsilateral hypertropia)
- Extorsion of eyeball
- Restriction of the ocular movements on downwards and inwards
- *Bielschowsky's head tilt* test is useful to diagnose a fourth nerve palsy **(Figs 15.17.1 and 15.17.2).**

Fig. 15.17.1: Left fourth nerve (superior oblique) palsy

Fig. 15.17.2: Left fourth nerve (superior oblique) palsy

Sixth (Abducens) Nerve Palsy

- This is the most common type, and commonly occurs in raised intracranial tension
- The six nerve palsy is a false localizing sign
- The eyeball is rotated inwards (convergent squint) **(Fig. 15.18.1)**
- In paresis there is little movement of lateral rectus muscle **(Fig. 15.18.2)**
- Defective abduction of the eye
- Face turns towards the field of action of paralyzed muscle.

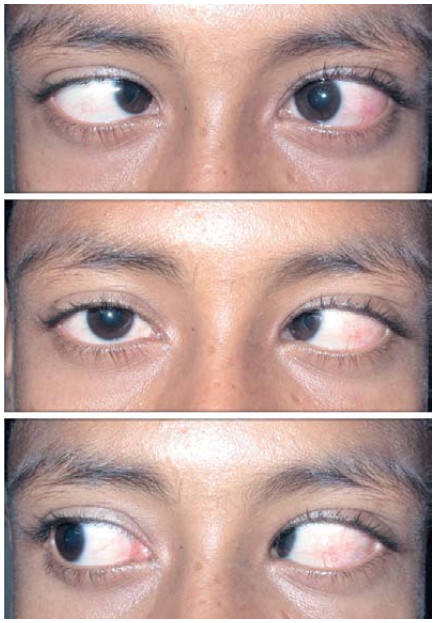


Fig. 15.18.1: Left sixth nerve (lateral rectus) palsy

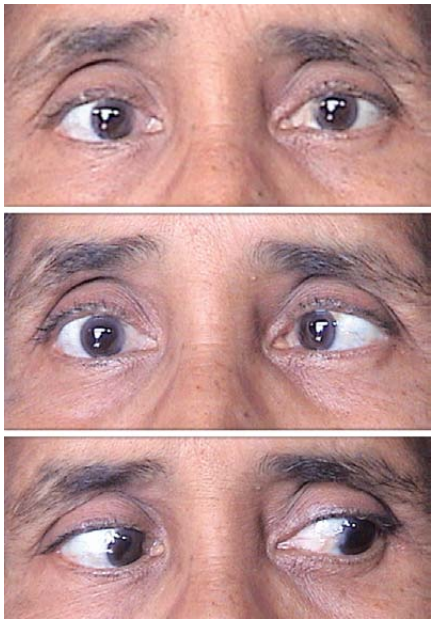


Fig. 15.18.2: Right sixth nerve (lateral rectus) paresis

Miscellaneous Ocular Movement Disorders

Duane's Retraction Syndrome

- This type of non-comitant squint, occurs because of aberrant innervation which causes contraction of lateral rectus muscle instead of relaxation
- Marked restriction
- Absence of abduction
- Slight restriction of the adduction
- Retraction of the globe on attempted adduction
- Palpebral aperture normal or narrowing on adduction, and widening on attempted abduction
- 'Up-shoot' or 'down-shoot' of the eyeball on adduction
- This is Type I (most common type)
  - It may be unilateral **(Fig. 15.19.1)** or bilateral **(Fig. 15.19.2)**
- The other types:
  - *Type II:* Limitation of adduction with relatively normal abduction
  - *Type III:* Limitation of the both abduction and adduction
- *Treatment:* Majority remain asymptomatic and no treatment is required
- Amblyopia is uncommon and when present, rarely dense
- This can be treated with occlusion, and any refractive error can also be corrected.

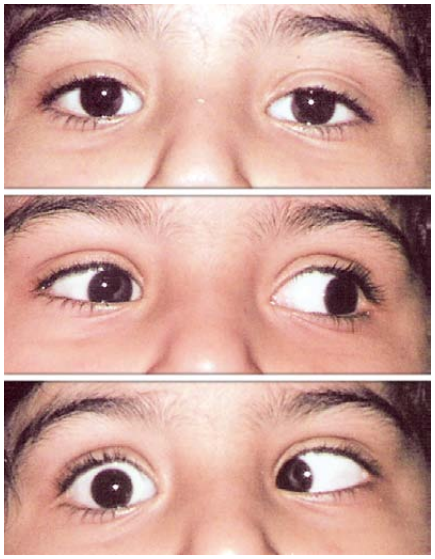


Fig. 15.19.1: Duane's retraction syndrome—unilateral

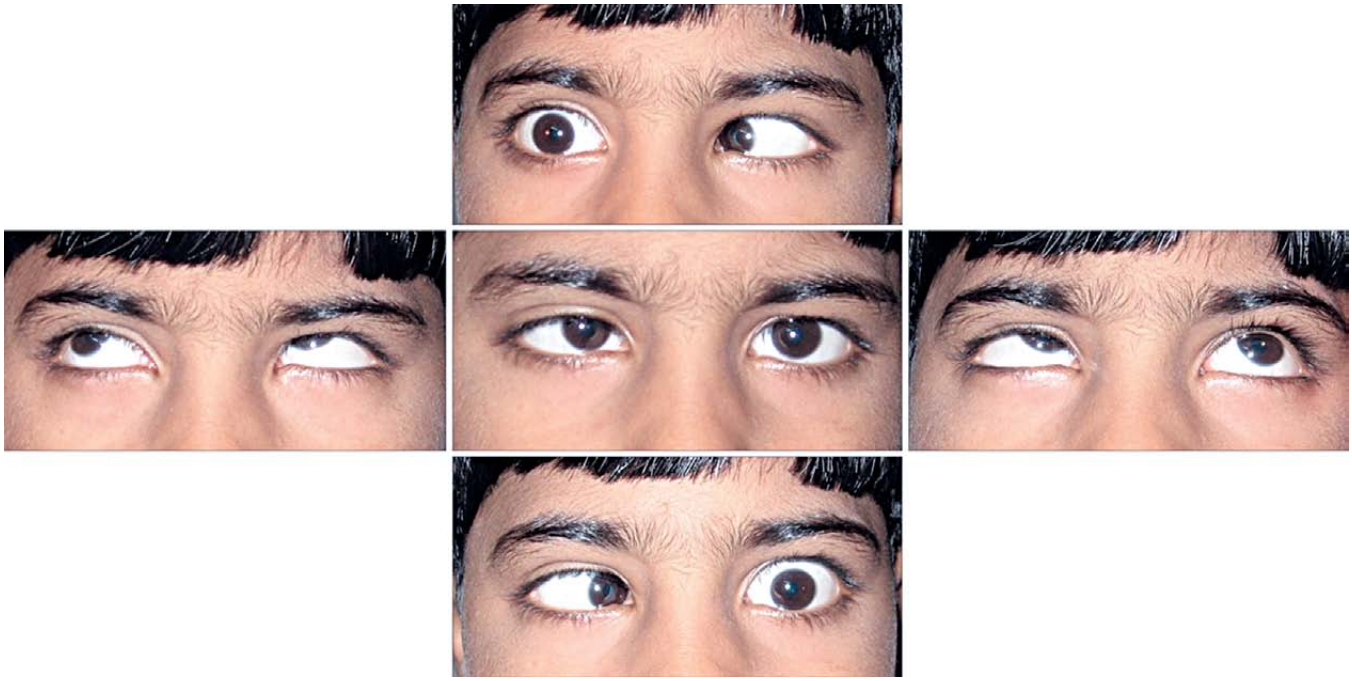


Fig. 15.19.2: Duane's retraction syndrome—bilateral

Superior Oblique Tendon Sheath Syndrome (Brown's Syndrome)

- Superior oblique tendon-trochlea apparatus
- Limitation of elevation of the eye on adduction, simulating inferior oblique muscle palsy
- Normal elevation on abduction **(Fig. 15.20.1)**
- Eyes are straight on primary position
- There is no overaction of superior oblique muscle
- *No treatment is usually required.*

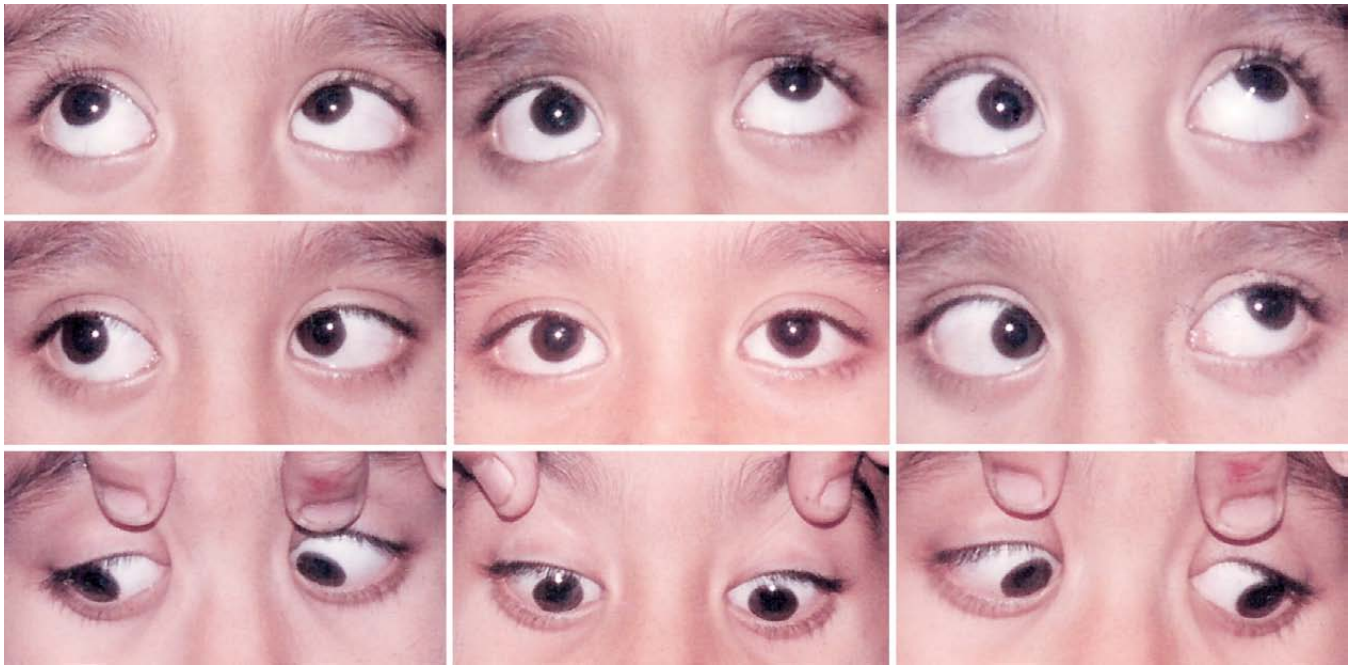


Fig. 15.20.1: Brown's (superior oblique tendon sheath) syndrome

Double Elevator Palsy

- May be congenital **(Fig. 15.21.1)** or acquired **(Fig. 15.21.2)**
- Eyeballs are straight in primary position
- Mild degree of ptosis may be present in some cases
- Limitation of elevation in all position of upgaze
- Normal adduction and abduction movements
- Normal depression in all downgaze positions
- *Surgical correction is required in selective cases.*

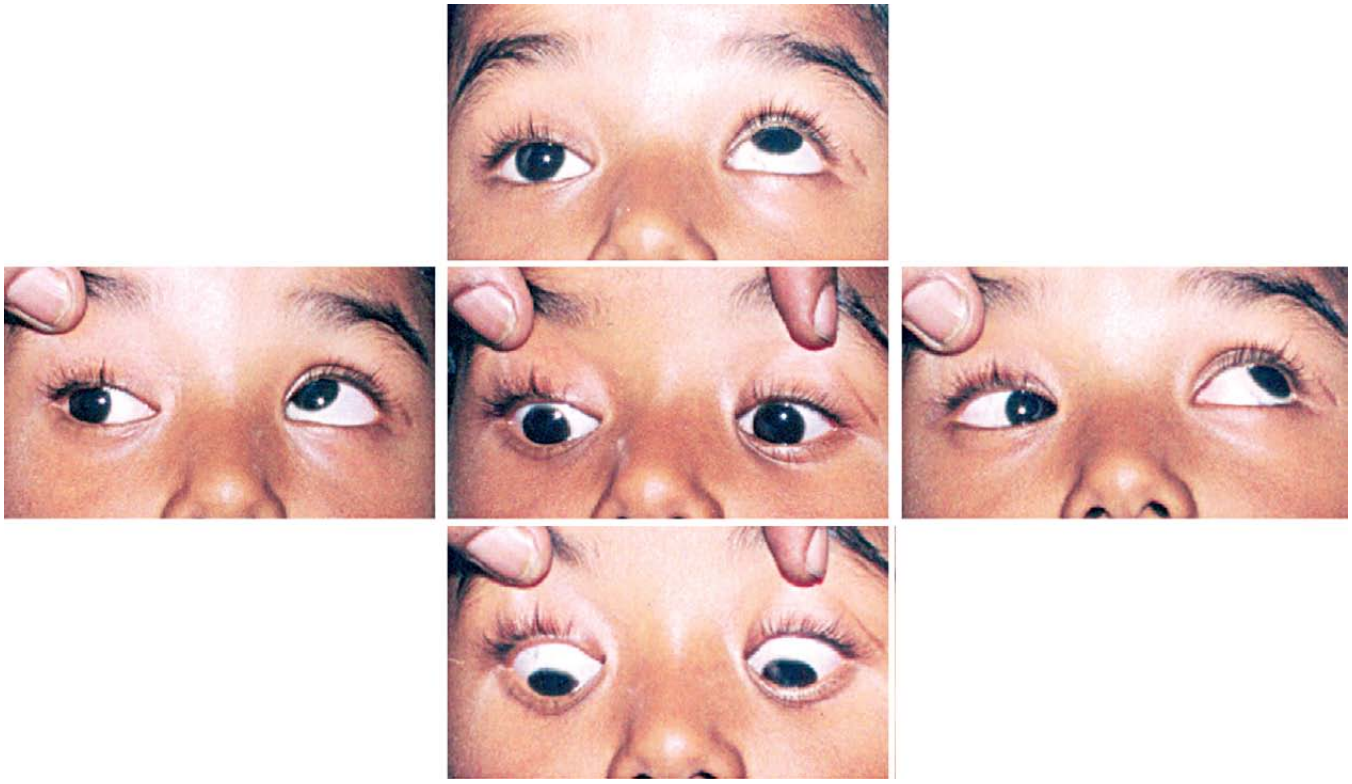


Fig. 15.21.1: Double elevator palsy—congenital—right side

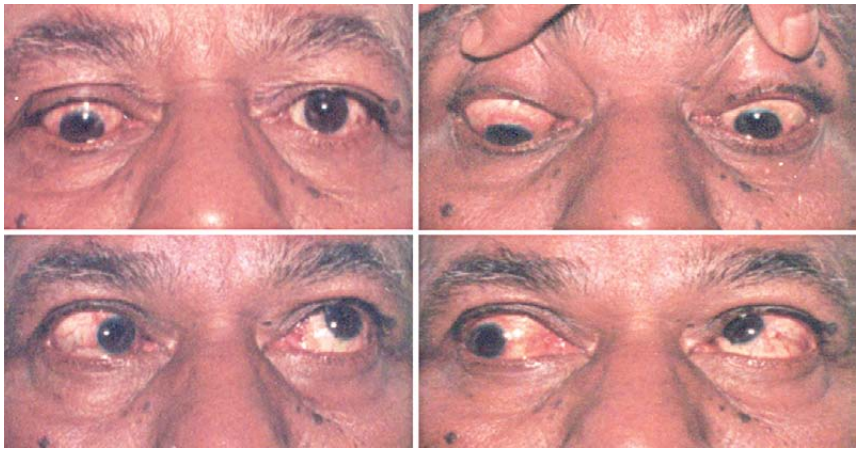


Fig. 15.21.2: Double elevator palsy—acquired—right side

Restrictive Strabismus

- Not so uncommon
- Conditions that causes tethering of extraocular muscles **(Fig. 15.22.1)**
- Restriction of ocular movement in the direction of action of the affeced muscle (incomitant strabismus)
- Positive forced duction test (FDT)
- *Common causes:* Congenital fibrosis of extraocular muscles; thyroid ophthalmopathy, following orbital tumor surgery, orbital fractures (floor or medial wall); strabismus fixus.

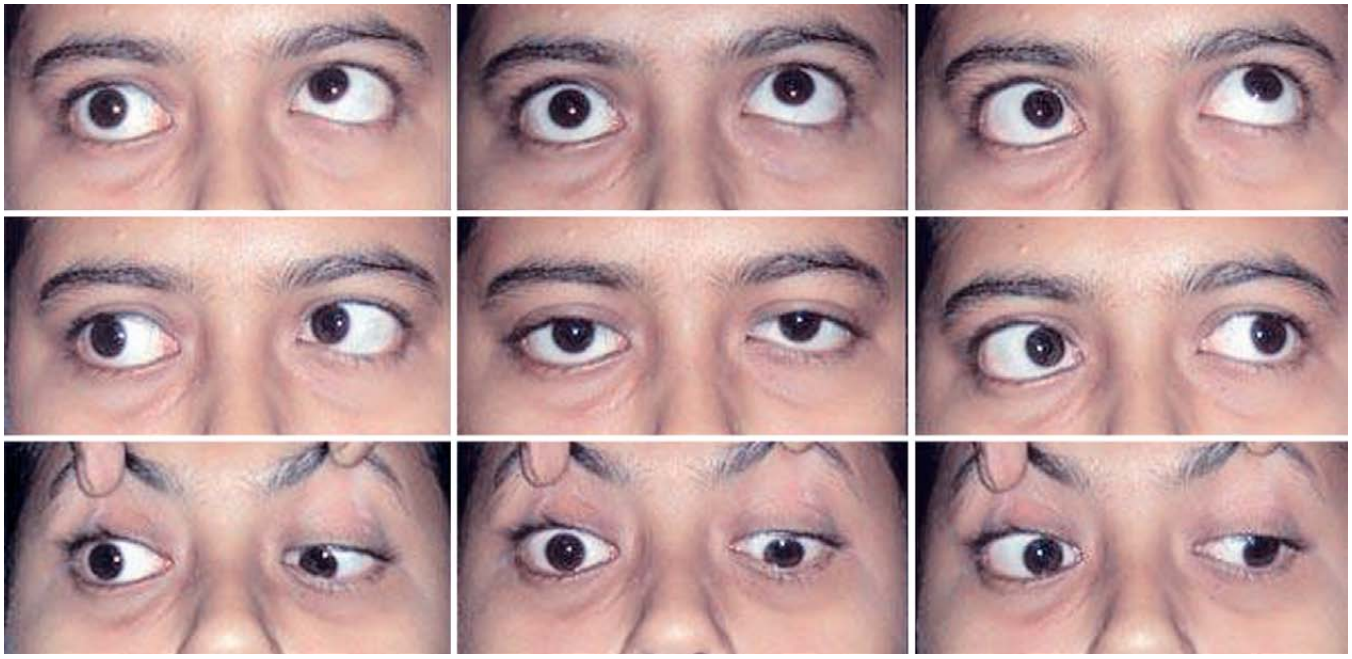


Fig. 15.22.1: Restrictive strabismus following orbital tumor surgery

Congenital Fibrosis of Extraocular Muscles (CFEOM)

- Rare genetic disorders affecting one or more extraocular muscles
- Individuals with CFEOM have varying degrees of ophthalmoplegia and ptosis **(Fig. 15.23.1)**
- It is present from birth and nonprogressive and runs in families
- Isolated superior rectus or inferior rectus muscle is commonly affected **(Figs 15.23.2A to C)**
- *No treatment is required.*

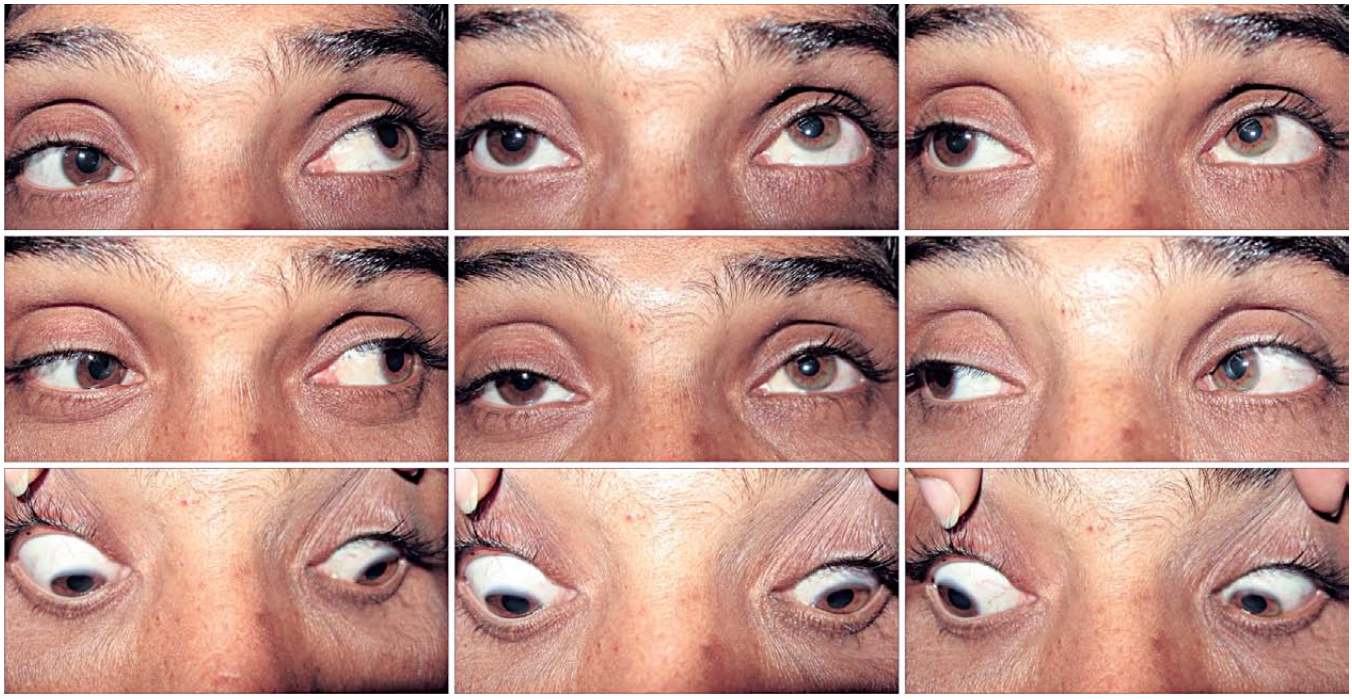


Fig. 15.23.1: CFEOM—ptosis and restriction in upgaze

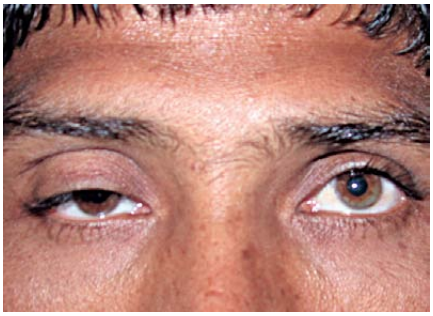


Fig. 15.23.2A: CFEOM—ptosis with superior rectus muscle involvement—RE

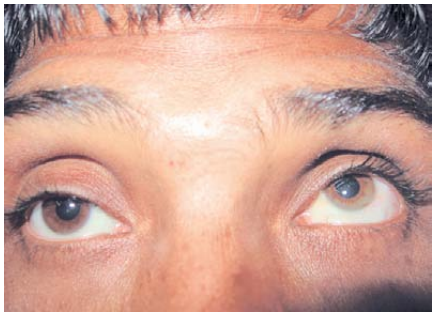


Fig. 15.23.2B: CFEOM—ptosis with superior rectus muscle involvement—RE

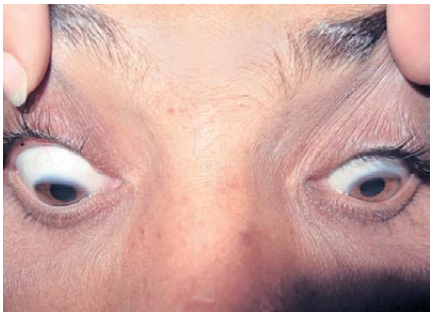


Fig. 15.23.2C: CFEOM—ptosis with superior rectus muscle involvement—RE

Orbital Fractures

- ***Orbital floor fracture: (see also Chapter 16)***
  - Diplopia in upgaze, enophthalmos and infraorbital nerve hypoesthesia
  - This is due to prolapse and entrapment of orbital content in the maxillary sinus
  - Limitation of movement in upgaze on affected side **(Fig. 15.24.1A)**
  - CT scan demonstrates classical "tear drop sign" **(Fig. 15.24.1B)**
- *Treatment:* Surgical repair of the orbital floor after 1 week of injury (to allow orbital swelling to subside), except in pediatric cases where urgent repar is needed.
- ***Medial wall (nasoethmoidal) fracture:***
  - This involves lacrimal and ethmoidal bones
  - Nasolacrimal duct injury and emphysema of lid and orbit
  - Medial rectus entrapment is there. Causing limitation of movement of lateral gaze **(Fig. 15.25.1A)**
  - CT scan confirms the diagnosis **(Fig. 15.25.1B)**

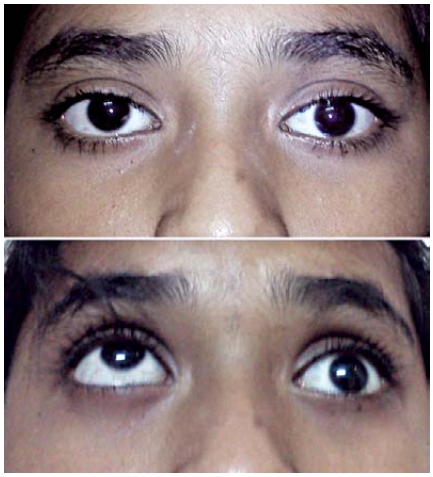


Fig. 15.24.1A: Orbital floor fracture—restriction in upgaze—LE

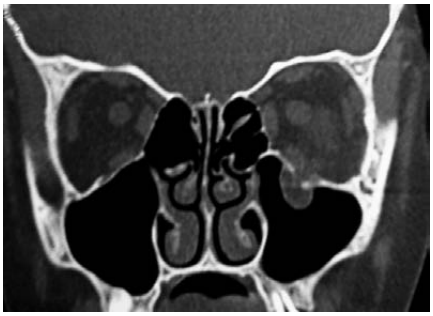


Fig. 15.24.1B: Orbital floor fracture—tear drop sign on left side maxilla

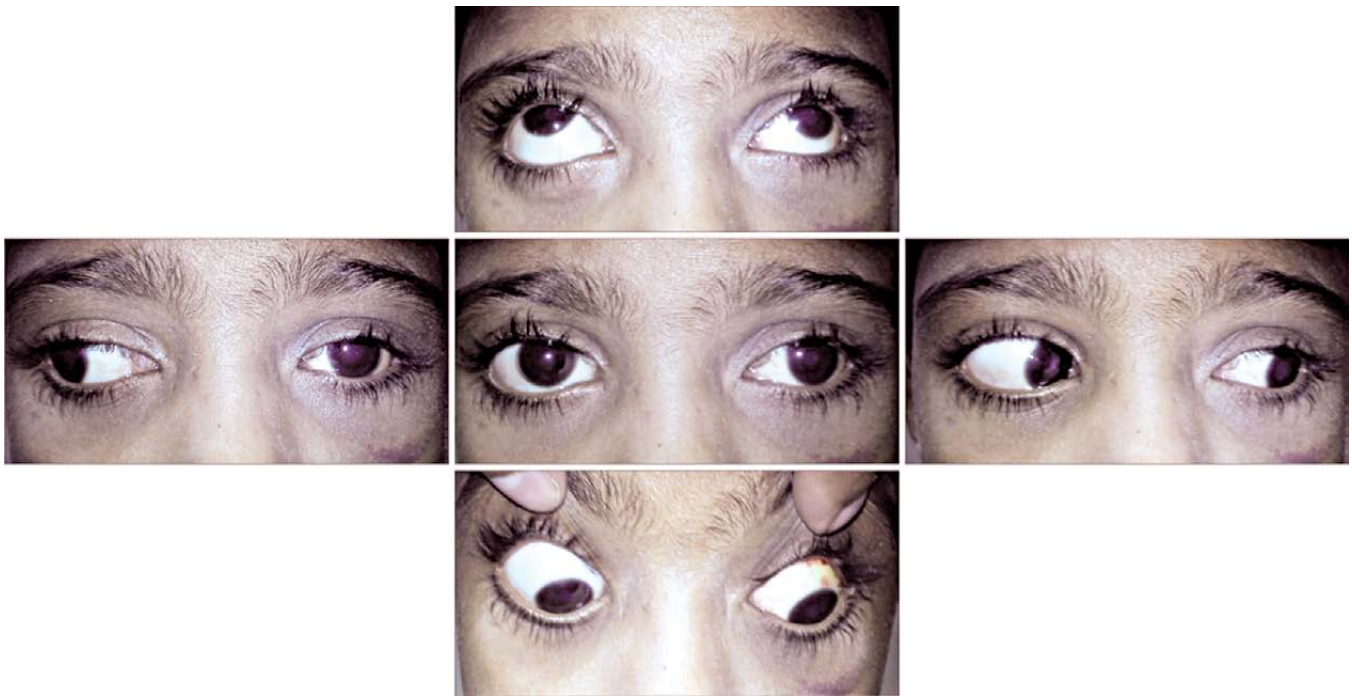


Fig. 15.25.1A: Orbit fracture—medial wall and floor

– *Treatment:* ENT consultation is required. Repair and release of medial rectus muscle. NLD is to be repaired if necessary.

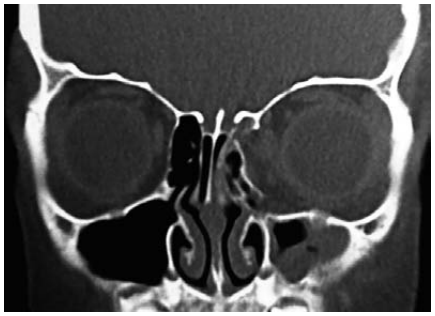


Fig. 15.25.1B: Orbit fracture—medial wall and floor

Strabismus Fixus

- Very rare unilateral or bilateral squinting condition
- Horizontal muscle affected in most cases
- Medial rectus is more affected than the lateral rectus muscle **(Fig. 15.26.1)**
- Affected eye is fixed in either convergent or divergent position due to fibrous contracture of medial or lateral rectus muscle
- Forced duction test (FDT) is positive.

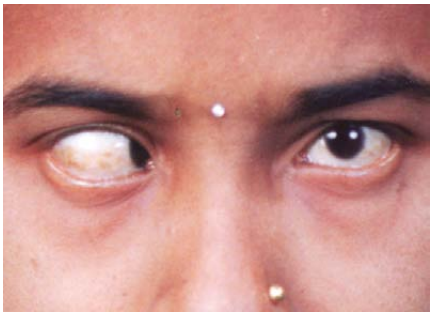


Fig. 15.26.1: Strabismus fixus

CHAPTER

16 Ocular Injuries

CHAPTER OUTLINE

Mechanical Injuries—Types

- Closed globe injury
- Open globe injury

Contusions (Blunt Injuries)

Various Effects Resulting from Contusions

- Eyelids
- Conjunctiva
- Cornea
- Sclera
- Anterior chamber
- Iris
- Ciliary body
- Crystalline lens
- Vitreous
- Choroid
- Retina
- Optic nerve
- Orbit

Penetrating (Perforating) Injuries

- Immediate effect of trauma
- Signs of globe perforation
- Introduction of infection
- Sympathetic ophthalmia

Foreign Bodies in the Eye

- Extraocular foreign bodies
- Conjunctival foreign body
- Corneal foreign body
- Intraocular foreign body (IOFB)
- Siderosis bulbi
- Chalcosis bulbi
- Miscellaneous organic materials

Chemical Injuries (Burns)

- Other chemical injuries
- Thermal burns

Miscellaneous Injuries

- Blast injuries
- Radiational injuries

Mechanical Injuries—Types

Closed Globe Injury

- Contusion
- Lamellar laceration
- Superficial foreign body
- Mixed
  - *Zone I:* Conjunctiva, cornea and sclera involved
  - *Zone II:* AC and lens is involved
  - *Zone III:* Posterior part is involved.

Open Globe Injury

- Rupture
- Penetrating
- IOFB
- Perforating
- Mixed
  - *Zone I:* Only cornea is involved
  - *Zone II:* Cornea and sclera up to 5 mm
  - *Zone III:* Wound extends > 5 mm of sclera.

Contusions (Blunt Injuries)

- Ocular injuries by various blunt instruments vary in severity depending upon the nature of impact
- It may be from simple subconjunctival hemorrhage to rupture of the globe
- Moreover some effects are progressive or may be delayed
- So during treatment, a guarded prognosis should be given to such injuries

Various Effects Resulting from Contusions

Eyelids

- Lid lacerations **(Fig. 16.1.1)**
- Swelling and ecchymosis (black eye) of the lids **(Fig. 16.1.2)** also responsible for *'Panda bear'* sign **(Fig. 16.1.3)**
- Emphysema of the eyelids due to fracture of the orbital wall with communication with PNS **(Fig. 16.1.4)**

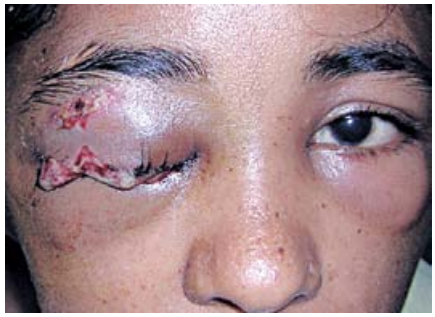


Fig. 16.1.1: Lid laceration following blunt trauma

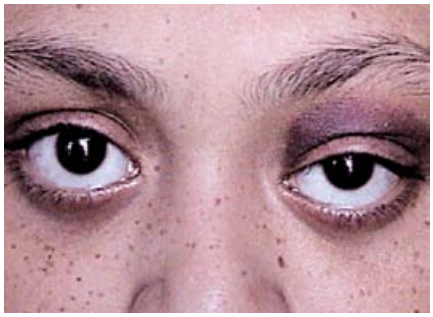


Fig. 16.1.2: Ecchymosis of eyelid—LE

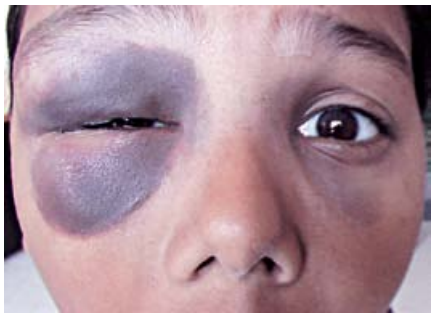


Fig. 16.1.3: Black eye—Panda bear sign—RE

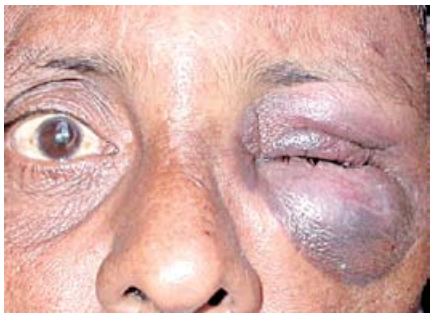


Fig. 16.1.4: Ecchymosis with emphysema of the lid

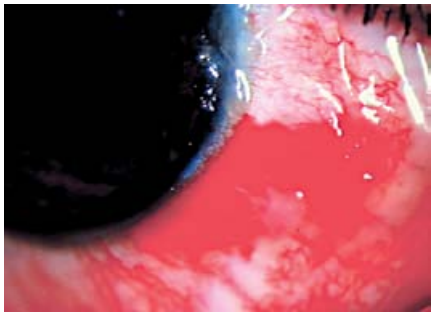


Fig. 16.2.1: Conjunctival laceration

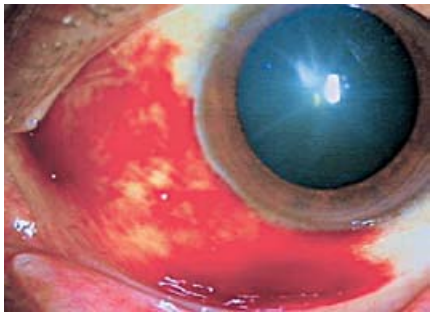


Fig. 16.2.2: Subconjunctival hemorrhage

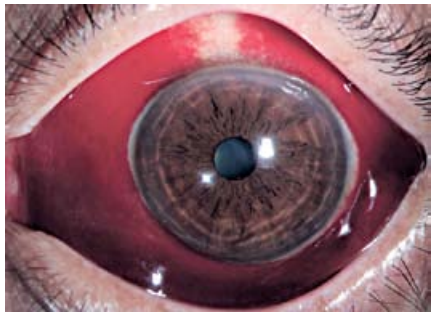


Fig. 16.2.3: Subconjunctival hemorrhage

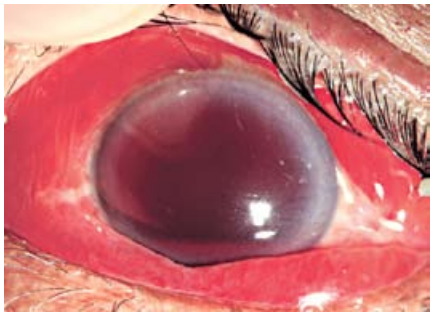


Fig. 16.2.4: Subconjunctival hemorrhage with chemosis of the conjunctiva

- *Treatment:* As such no active treatment is required. X-ray or CT scan of the orbit is required for suspected orbital wall fracture.

Conjunctiva

- Conjunctival lacerations **(Fig. 16.2.1)**
- Subconjunctival hemorrhage which may be small or large **(Figs 16.2.2 and 16.2.3)**

- May be associated with conjunctival chemosis **(Fig. 16.2.4)**
- *Treatment:* Large laceration may require repair. In chemosis, one should exclude the possibility of scleral rupture.

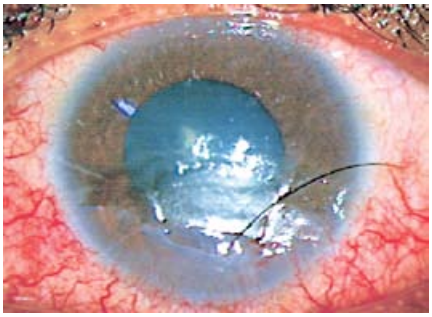


Fig. 16.3.1: Corneal abrasion

Cornea

- Simple abrasions **(Figs 16.3.1 and 16.3.2)**
- Rupture of the Descemet's membrane with folds and corneal edema **(Fig. 16.3.3)**
- *Blood staining of the cornea*
  - Traumatic hyphema with raised IOP
  - Endothelial damage and subsequently whole cornea is reddish-brown or greenish stained **(Figs 16.3.4 and 16.3.5)**
  - Cornea gradually and slowly clears from the periphery **(Figs 16.3.6 and 16.3.7)**
- *Treatment:* Good control of IOP followed by urgent paracentesis is required to prevent it.

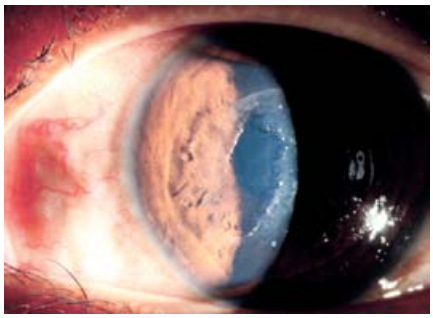


Fig. 16.3.2A: Corneal abrasion

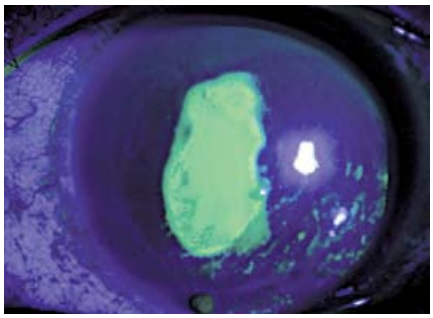


Fig. 16.3.2B: Corneal abrasion with fluorescein staining—same eye

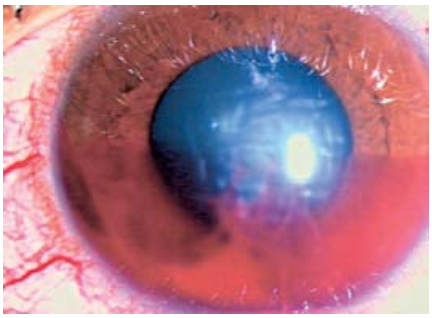


Fig. 16.3.3: Descemet's folds and corneal edema with hyphema

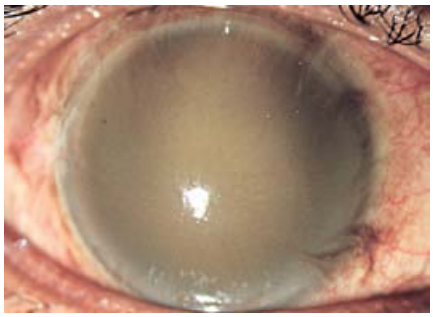


Fig. 16.3.4A: Blood staining of cornea—early

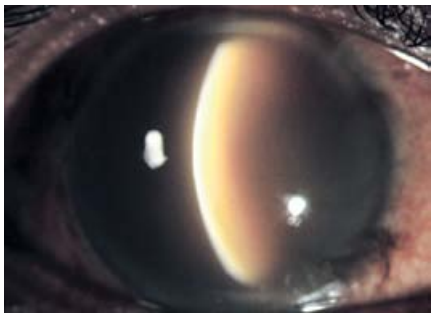


Fig. 16.3.4B: Blood staining of cornea—early

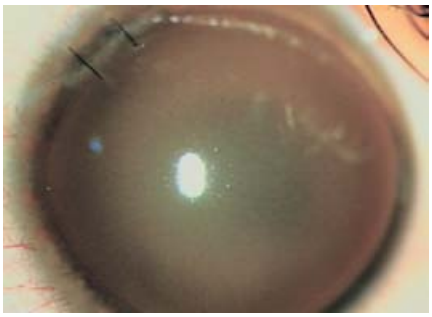


Fig. 16.3.5A: Blood staining of cornea—persisted even after paracentesis

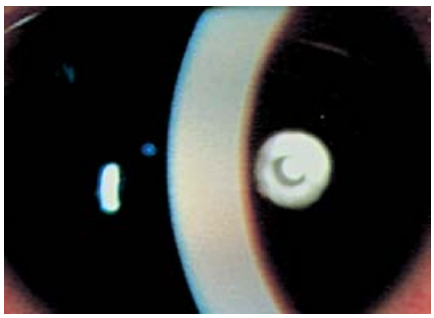


Fig. 16.3.5B: Blood staining of cornea—persisted even after paracentesis

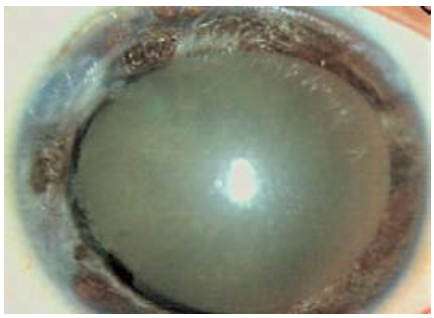


Fig. 16.3.6: Blood staining of cornea—started clearing from the periphery

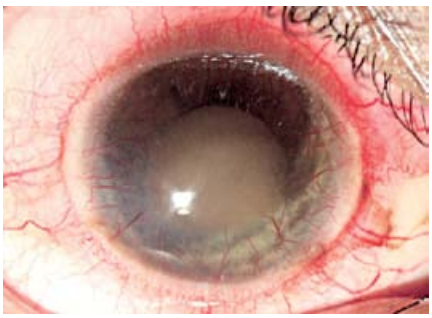


Fig. 16.3.7: Blood staining of cornea—absorbing from the periphery

Sclera

- *Scleral rupture (rupture of the globe)*
  - Usually near canal of Schlemm (weakest point) and runs concentrically with the limbus **(Figs 16.4.1A and B)**
  - Conjunctiva is often intact
  - Associated with uveal prolapse **(Fig. 16.4.2)** or subconjunctival dislocation of crystalline lens **(Fig. 16.4.3)**
  - In pseudophakic eyes, the rupture occurs at the limbal or scleral tunnel with uveal prolapse **(Figs 16.4.4 and 16.4.5)** or with dislocation of IOL in the subconjunctival space **(Figs 16.4.6 to 16.4.8)**
  - Eyeball usually collapses with total loss of vision
  - Prognosis is often very poor
- *Treatment:* Surgical repair with abscission of the prolapsed iris if any **(Figs 16.4.9A and B)**

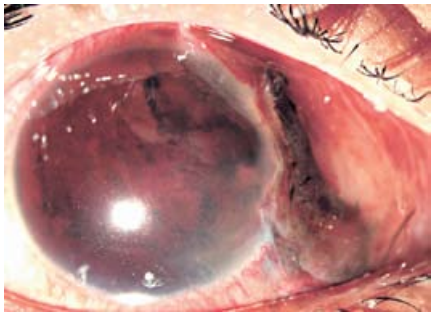


Fig. 16.4.1A: Scleral rupture at the limbus with uveal prolapse

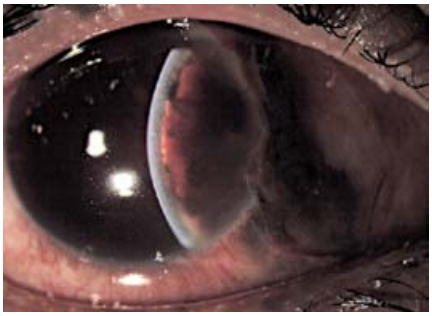


Fig. 16.4.1B: Scleral rupture at the limbus with uveal prolapse

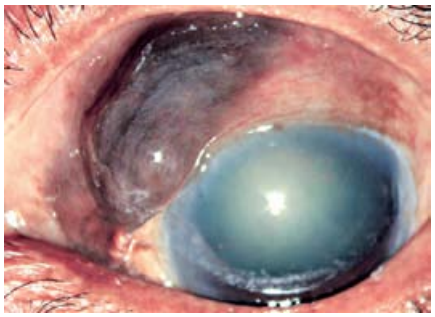


Fig. 16.4.2: Scleral rupture with uveal prolapse

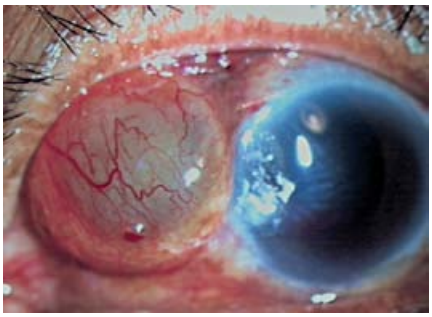


Fig. 16.4.3: Scleral rupture—subconjunctival dislocation of the lens after trauma—phacocele

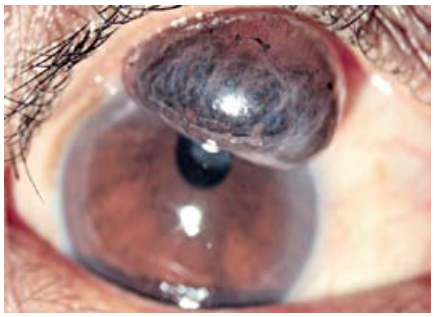


Fig. 16.4.4A: Limbal rupture with iris prolapse with cystic changes

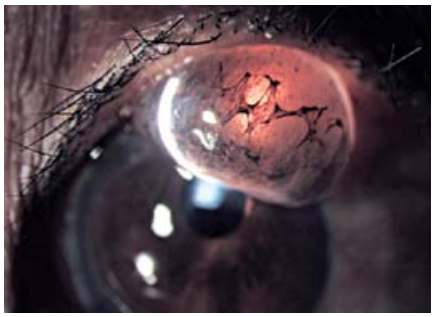


Fig. 16.4.4B: Same eye with cystic changes—transillumination positive

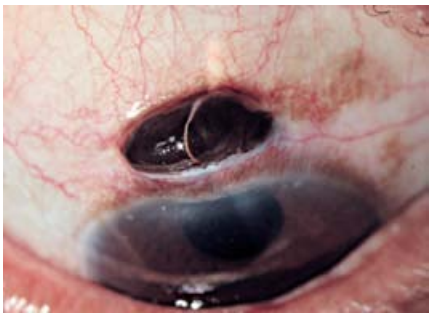


Fig. 16.4.5: Trauma—iris prolapse through phaco tunnel

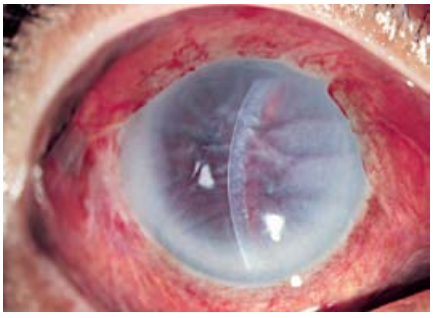


Fig. 16.4.6A: Scleral rupture—uveal prolapse and IOL extrusion

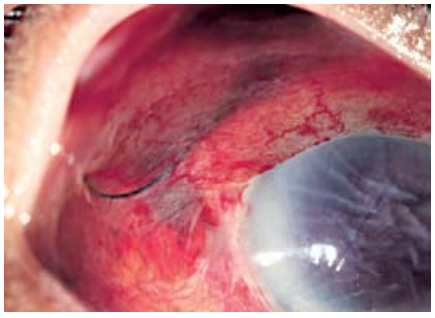


Fig. 16.4.6B: Scleral rupture—uveal prolapse and IOL extrusion

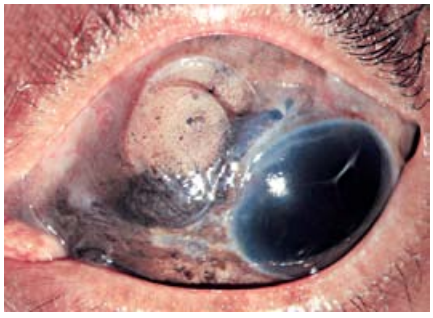


Fig. 16.4.7: Scleral rupture with subconjunctival dislocation of IOL

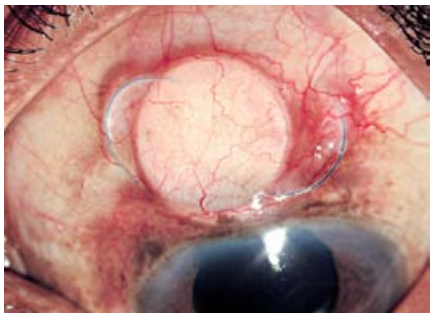


Fig. 16.4.8: Subconjunctival dislocation of IOL—pseudophacocele—trauma by plastic ball

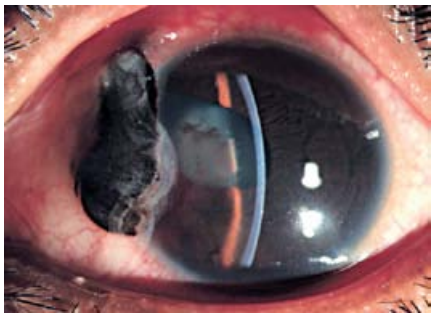


Fig. 16.4.9A: Scleral rupture at the limbus with iris prolapse

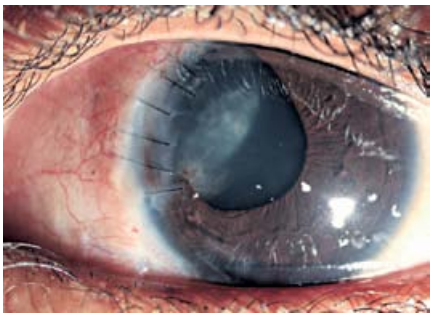


Fig. 16.4.9B: Same eye after surgical repair with abscission of iris

- *Mutilated globe rupture:* A severe blow or by direct blunt injury by heavy object can cause mutilated globe injury with extrusion of all the contents of the eyeball **(Fig. 16.4.10).** *Enucleation is advised in this cases.*

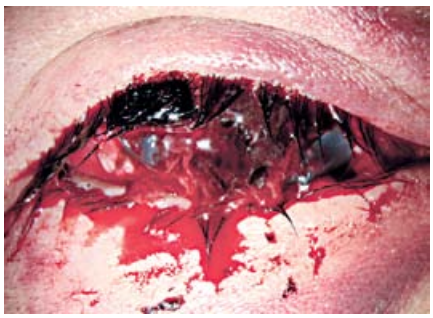


Fig. 16.4.10: Mutilated globe rupture following cricket ball injury

Anterior Chamber

- *Hyphema* (blood in the anterior chamber)
  - Fresh blood usually does not get clotted **(Fig. 16.5.1)**
  - Recurrent and more severe bleeding may occur within 24–72 hours
  - With rest blood gets settled with fluid level **(Fig. 16.5.2)**, but in some cases it gets partially clotted **(Fig. 16.5.3)**
  - With clotted blood, the hyphema appears as black ball **(Figs 16.5.4A and B)**, called "8 ball hyphema" (like No. '8' ball in billiards game)
  - Associated with secondary glaucoma

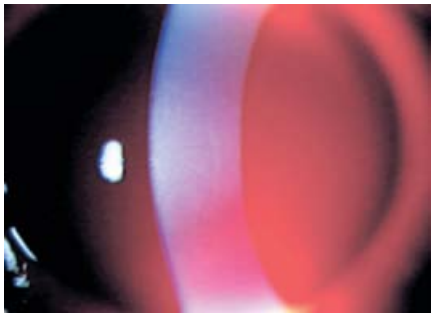


Fig. 16.5.1: Traumatic hyphema—fresh blood

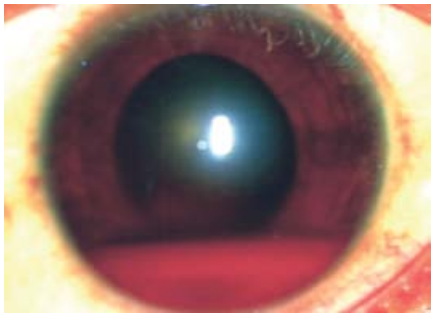


Fig. 16.5.2: Traumatic hyphema—settled with blood level

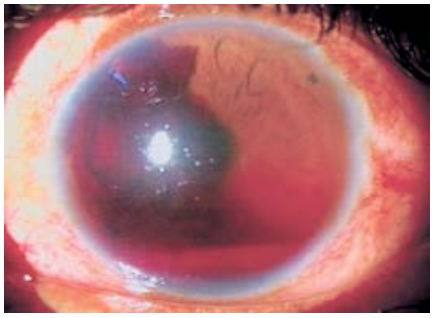


Fig. 16.5.3: Blunt trauma—hyphema—partially clotted blood

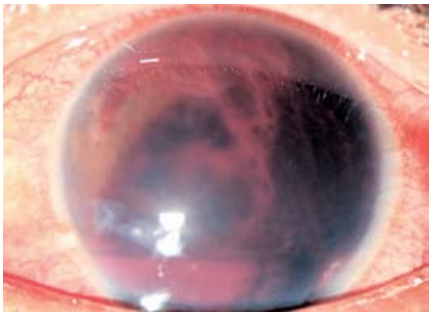


Fig. 16.5.4A: Blunt trauma—8-ball (black ball) hyphema

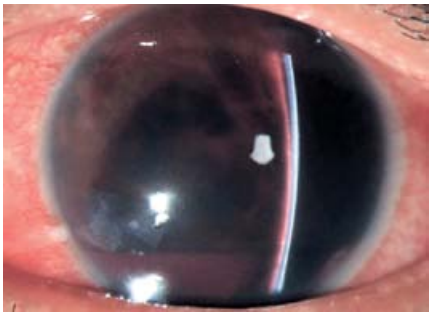


Fig. 16.5.4B: Blunt trauma—8-ball (black ball) hyphema

- *Treatment:* Conservative with systemic and topical steroids, and antiglaucoma medication; followed by paracentesis if necessary if the IOP does not control within 5–7 days.

Iris

- *Iridodialysis:*
  - Iris is partially torn away from its ciliary attachment
  - Biconvex area is seen at the periphery, causing a D-shaped pupil **(Figs 16.6.1A and B)**
  - A fundal glow is obtained through the peripheral gap **(Figs 16.6.2A and B)**
  - It may be associated with sphincteric tears **(Figs 16.6.3A to C)** or subluxation of the crystalline lens **(Fig. 16.6.4)**
- *Sphincteric tear:* Isolated sphincteric tears may occur with multiple small notches at the pupillary margin, associated with hyphema **(Figs 16.6.5A and B)**

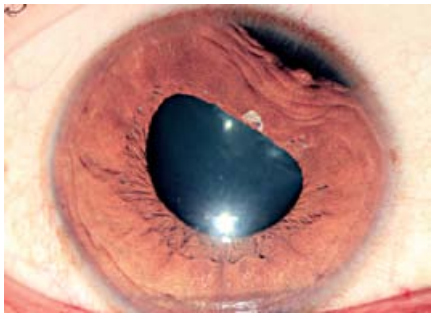


Fig. 16.6.1A: Iridodialysis—D-shaped pupil

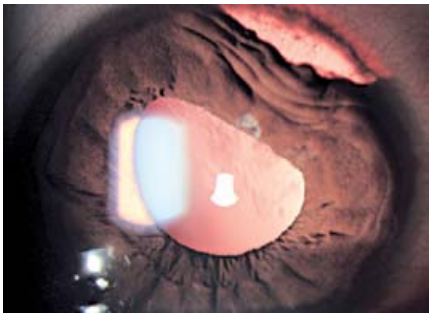


Fig. 16.6.1B: Iridodialysis—D-shaped pupil

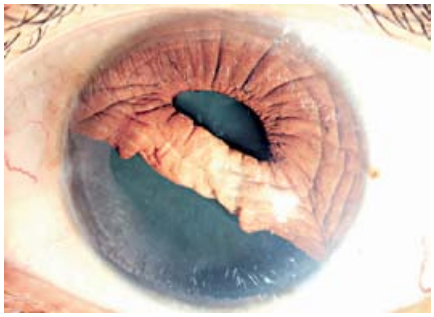


Fig. 16.6.2A: Iridodialysis—large—D-shaped pupil

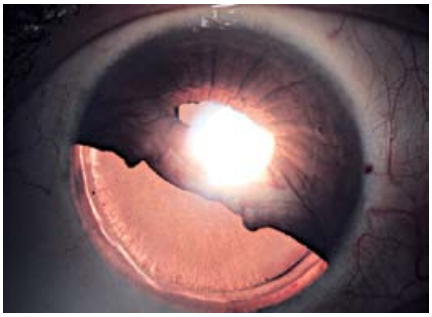


Fig. 16.6.2B: Iridodialysis—D-shaped pupil

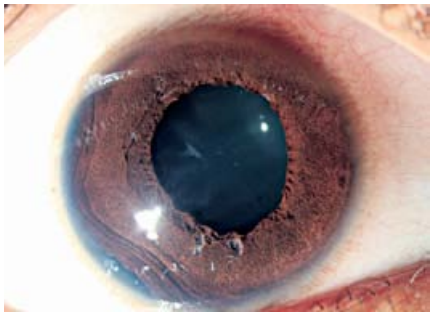


Fig. 16.6.3A: Iridodialysis with sphincteric tears—not clearly visible

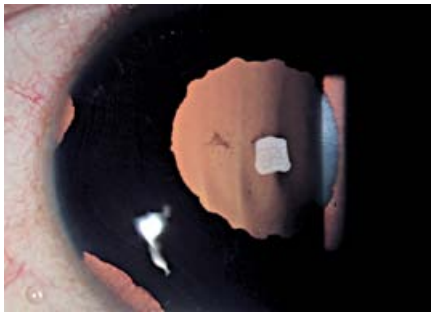


Fig. 16.6.3B: Iridodialysis (double) with sphincteric tears

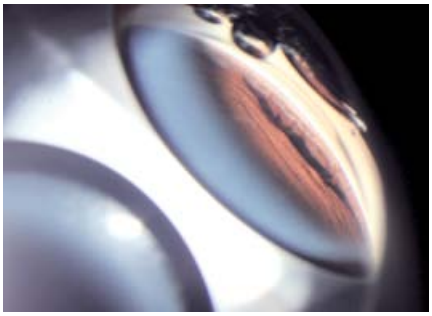


Fig. 16.6.3C: Iridodialysis in gonioscopy

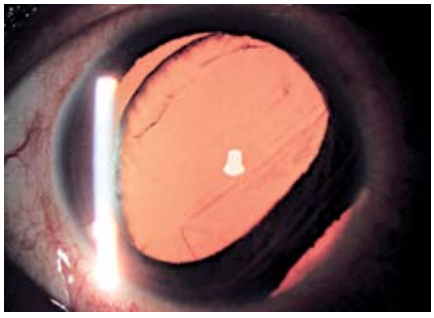


Fig. 16.6.4: Iridodialysis with subluxation of lens

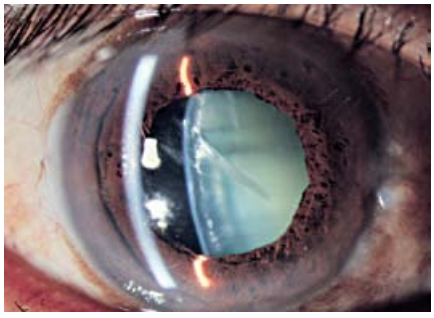


Fig. 16.6.5A: Sphincteric tears—small and multiple

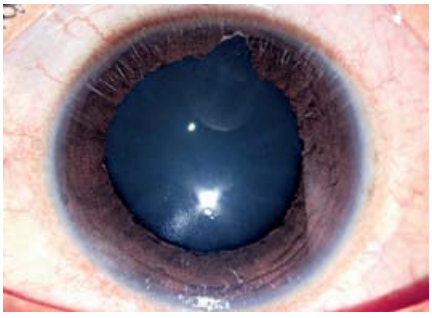


Fig. 16.6.5B: Sphincteric tear—large

- *Traumatic aniridia:* Partial **(Fig. 16.6.6)** or total. In total aniridia, iris may appear as a ball in the anterior chamber or whole detached iris may be displaced subconjunctivally through a scleral rupture **(Figs 16.6.7A and B)**
- *Anteflexion and retroflexion* of the iris **(Figs 16.6.8 and 16.6.9)**
- *Traumatic iridocyclitis*—usually mild **(Fig. 16.6.10)**
- *Traumatic mydriasis* **(Figs 16.6.11 and 16.6.12)**
- *Post-traumatic iris atrophy* **(Figs 16.6.13 and 16.6.12).**

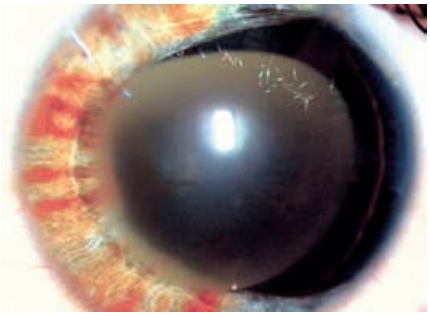


Fig. 16.6.6: Traumatic aniridia—partial

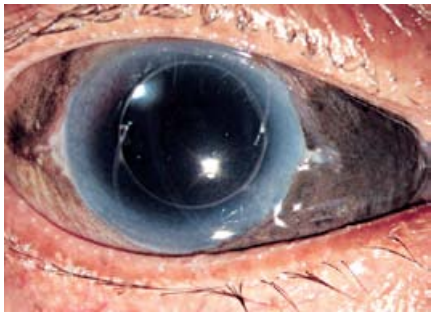


Fig. 16.6.7A: Traumatic aniridia—total iris in subconjunctival space

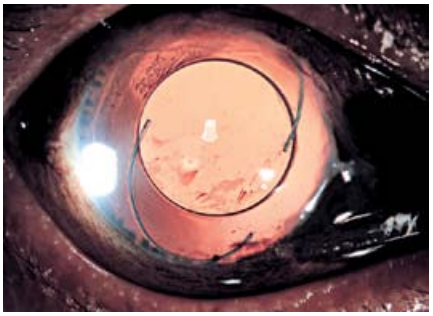


Fig. 16.6.7B: Traumatic aniridia—total iris in subconjunctival space

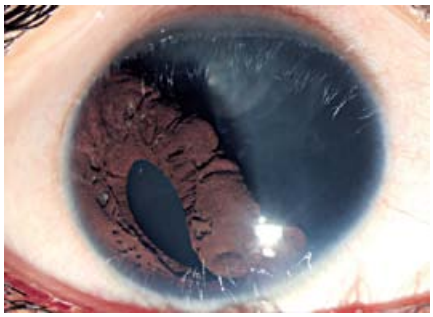


Fig. 16.6.8A: Iridodialysis—antiflexion of the iris

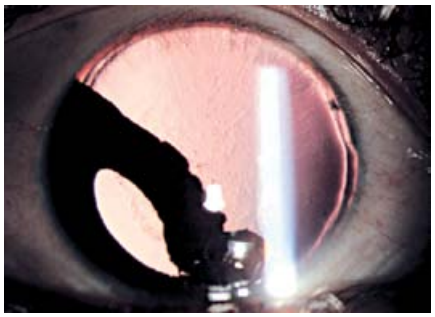


Fig. 16.6.8B: Iridodialysis—antiflexion of the iris

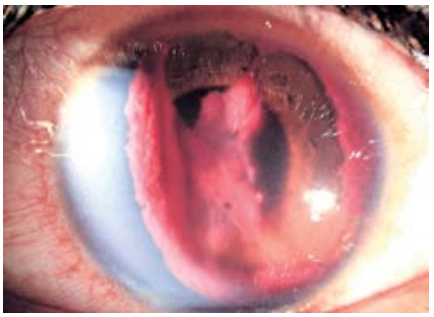


Fig. 16.6.9: Retroflexion of the iris with hyphema

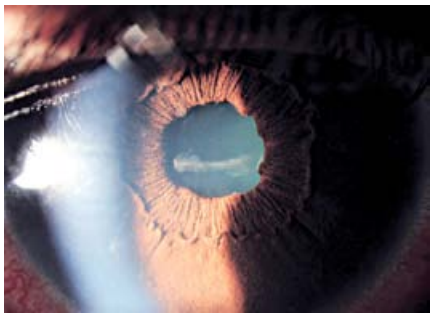


Fig. 16.6.10: Traumatic iridocyclitis with sphincteric tears

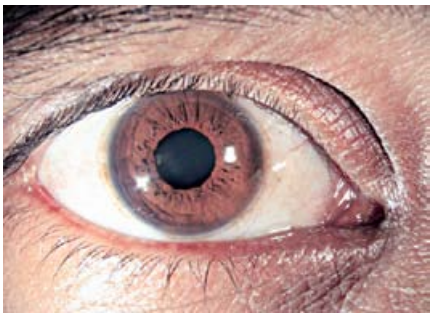


Fig. 16.6.11A: Traumatic mydriasis—affected—RE

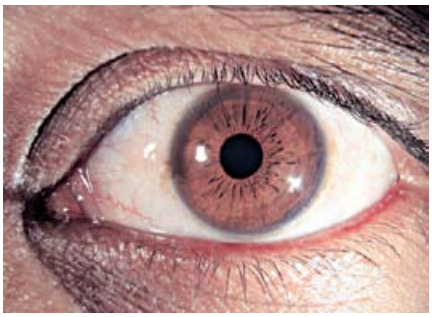


Fig. 16.6.11B: Normal unaffected—LE

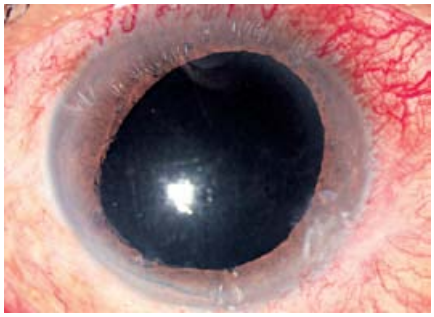


Fig. 16.6.12: Traumatic mydriasis with lens dislocation

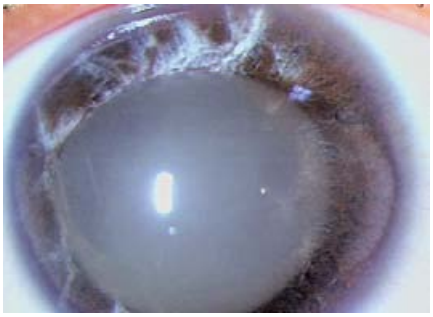


Fig. 16.6.13: Post-traumatic iris atrophy

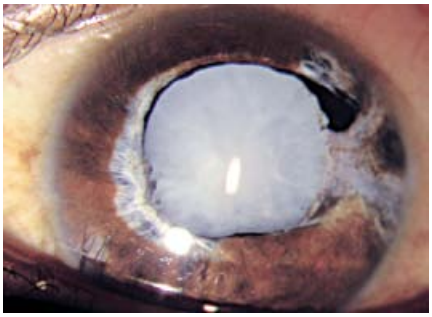


Fig. 16.6.14: Post-traumatic iris atrophy with total cataract

Ciliary Body

- *Angle recession:*
  - Longitudinal tear in the face of the ciliary body
  - Splitting of the circular fibers from the longitudinal fibers **(Fig. 16.7.1)**
  - Deepening of the anterior chamber
  - Widening of the ciliary band on gonioscopy **(Fig. 16.7.2)** with late glaucoma.

Fig. 16.7.1: Angle recession

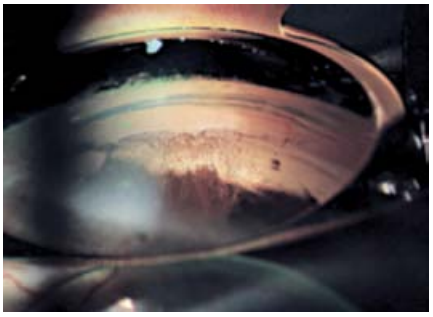


Fig. 16.7.2: Angle recession glaucoma

Crystalline Lens

- *Vossius's ring:* A circular pigmented imprint of the moised pupil on the anterior lens surface **(Fig. 16.8.1)**
- *Concussion cataract:* Imbibitions of aqueous through the damaged capsule, and partly due to direct mechanical effect on the lens fibers:
  - *Localized cataract* often behind the iris **(Fig. 16.8.2)**
  - *Posterior subcapsular cataract* **(Figs 16.8.3 and 16.8.4)**

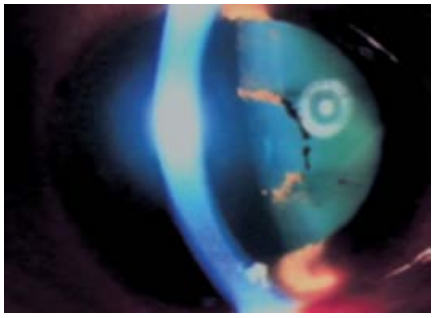


Fig. 16.8.1: Vossius's ring

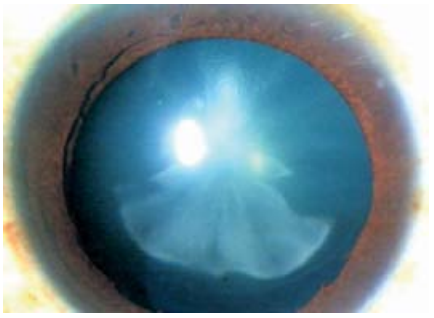


Fig. 16.8.2: Traumatic partial cataract

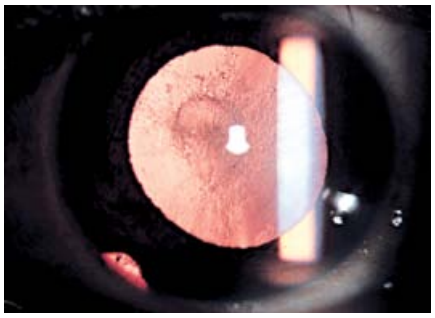


Fig. 16.8.3: Traumatic cataract—posterior subcapsular—note small iridodialysis at 7 o'clock

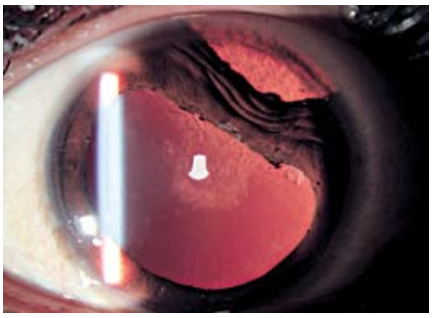


Fig. 16.8.4: Traumatic posterior subcapsular cataract with iridodialysis

- *Total cataract* with or without capsular tear **(Figs 16.8.5 and 16.8.6)**
- *Partially absorbed cataract* **(Figs 16.8.7 and 16.8.8)**

- *Early rosette cataract:* Star-shaped cataract, usually in the posterior cortex
  - The leaves of the feathery opacities are formed by the suture acting as a vein from which the opacities radiate **(Figs 16.8.9 and 16.8.10)**
  - Remains stationary, and sometimes, it may progress into a total cataract.
- *Late rosette cataract:*
  - Usually develops 1–2 years after a concussion
  - Smaller and more compact with short sutural extension
  - Feathery opacities lie in the angle between two adjacent sutures **(Fig. 16.8.11)**

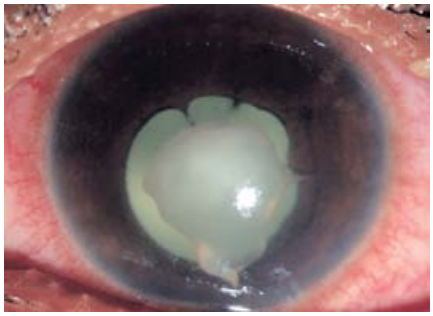


Fig. 16.8.5A: Blunt trauma—cataract with capsular rupture

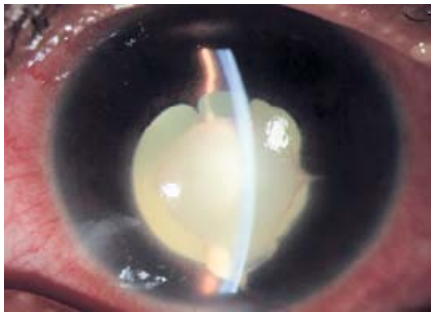


Fig. 16.8.5B: Blunt trauma—cataract with capsular rupture

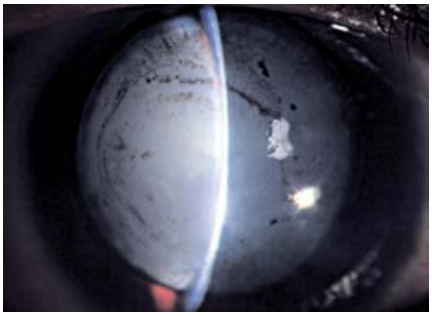


Fig. 16.8.6: Blunt trauma—total cataract with capsular rupture

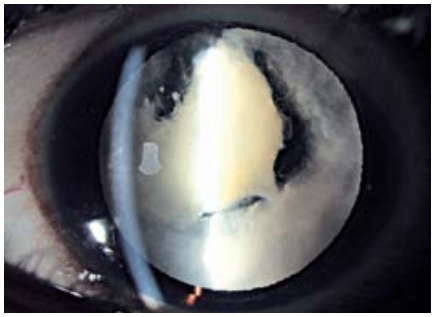


Fig. 16.8.7: Blunt trauma—capsular rupture with partial absorption of cataractous lens

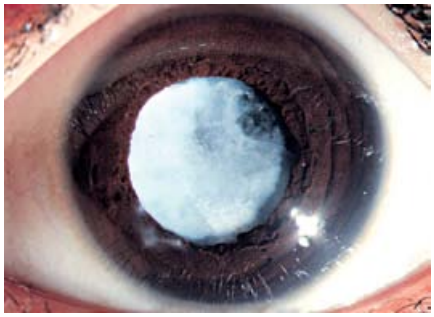


Fig. 16.8.8A: Partially absorbed cataract following blunt trauma

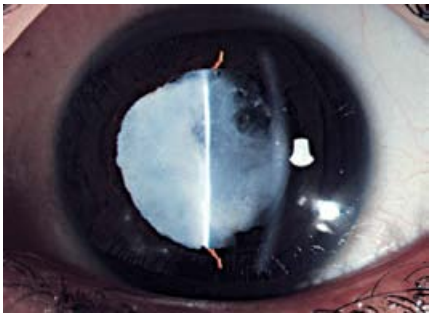


Fig. 16.8.8B: Partially absorbed cataract following blunt trauma

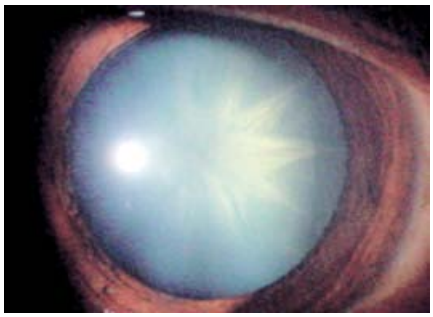


Fig. 16.8.9: Early rosette cataract

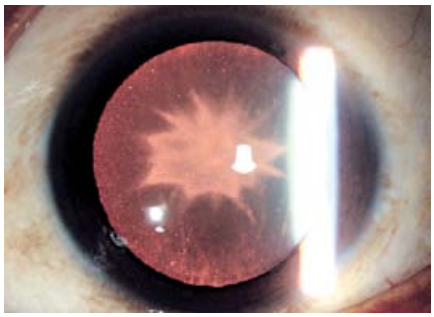


Fig. 16.8.10: Early rosette cataract

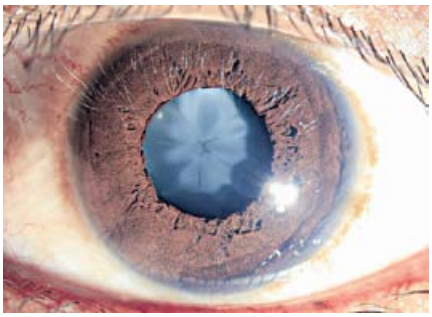


Fig. 16.8.11: Late rosette cataract

  - Best appreciated by retroillumination **(Figs 16.8.12A and B)**
- *Subluxation* of the crystalline lens or IOL **(Figs 16.8.13 and 16.8.14)**. Vitreous in AC or corneal touch is also seen in some cases of subluxation **(Figs 16.8.15 to 16.8.17)**
- *Dislocation* of the lens or IOL: in the anterior chamber **(Figs 16.8.18 and 16.8.19)** or in the vitreous cavity **(Figs 16.8.20 to 16.8.22)**
- *Traumatic aphakia* **(Figs 16.8.23 and 16.8.24)**.

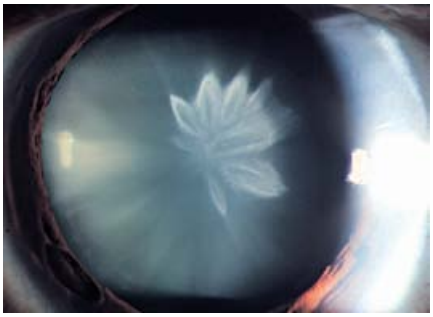


Fig. 16.8.12A: Late rosette cataract

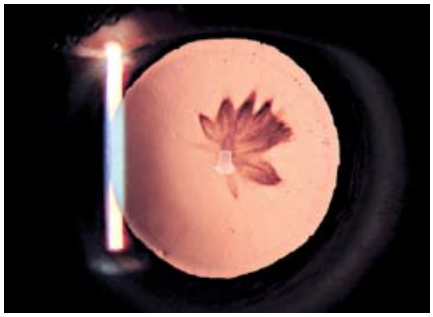


Fig. 16.8.12B: Late rosette cataract in retroillumination

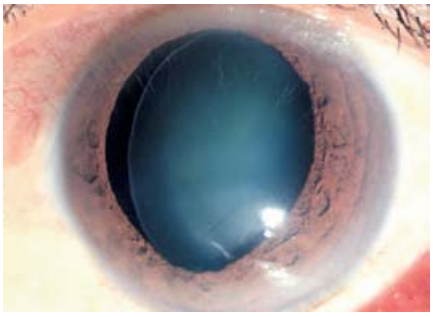


Fig. 16.8.13A: Subluxation of crystalline lens

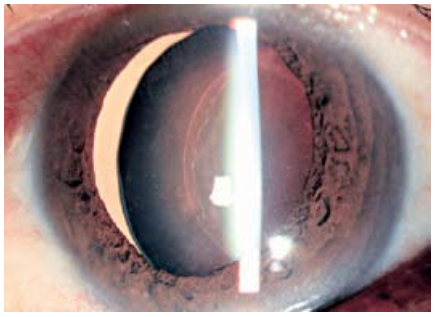


Fig. 16.8.13B: Subluxation of crystalline lens

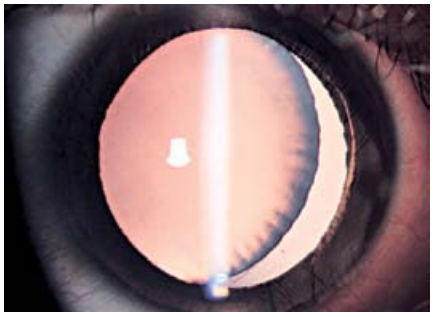


Fig. 16.8.14A: Subluxation of crystalline lens

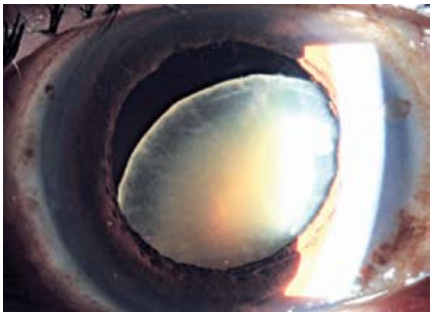


Fig. 16.8.14B: Subluxation of crystalline lens

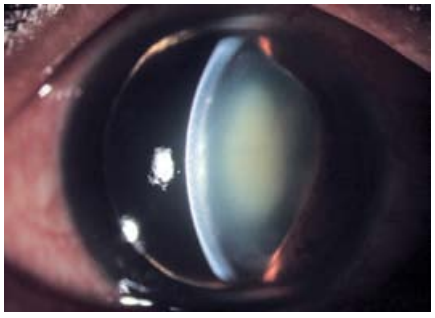


Fig. 16.8.15: Trauma subluxated lens touching the cornea with edema

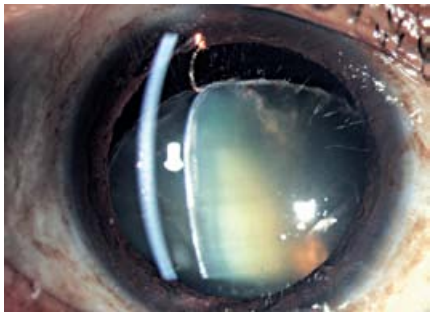


Fig. 16.8.16: Subluxation of crystalline lens—vitreous in AC

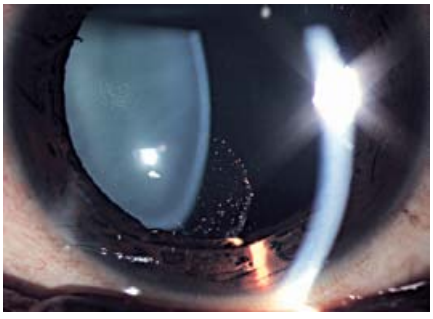


Fig. 16.8.17: Subluxation of crystalline lens—vitreous in AC

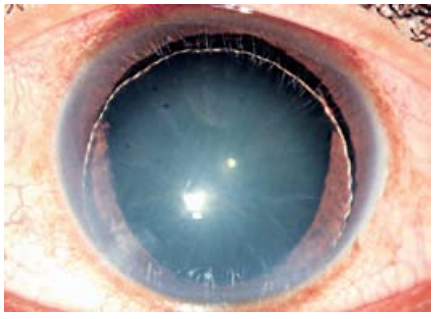


Fig. 16.8.18A: Dislocation of crystalline lens—in the anterior chamber

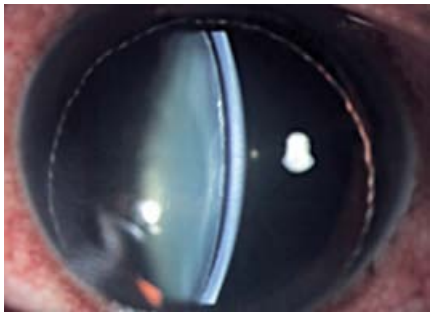


Fig. 16.8.18B: Dislocation of crystalline lens—in the anterior chamber

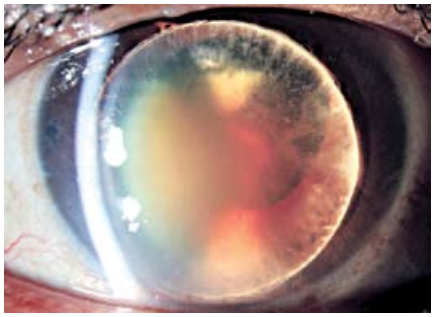


Fig. 16.8.19: Dislocation of the lens—anterior chamber

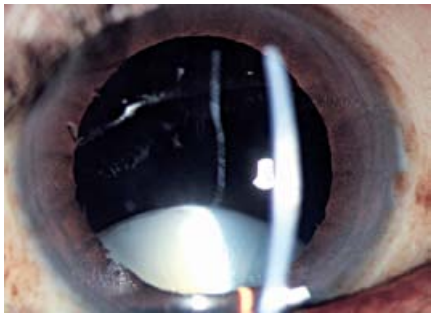


Fig. 16.8.20: Posterior dislocation of lens—in the vitreous

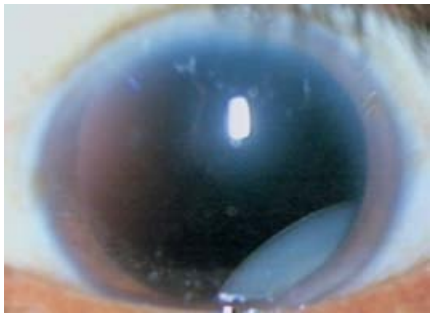


Fig. 16.8.21: Dislocation of lens—vitreous

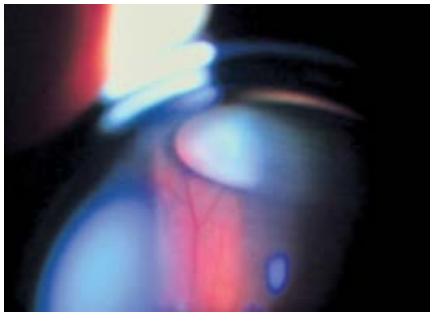


Fig. 16.8.22: Dislocation of lens—vitreous

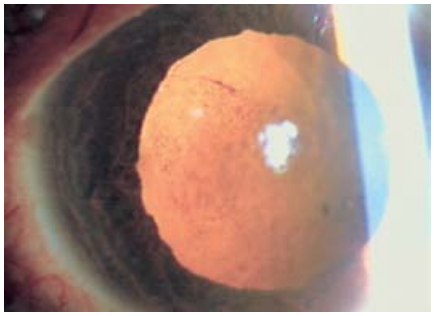


Fig. 16.8.23: Dislocation of crystalline lens—total

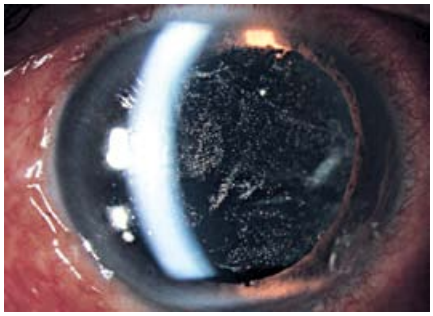


Fig. 16.8.24: Dislocation of crystalline lens—aphakia

Vitreous *(See also in Chapter 10)*

- Liquefaction of the vitreous
- Posterior vitreous detachment
- Vitreous hemorrhage with its sequelae **(Fig. 16.9.1)**
- Synchysis scintillans.

Choroid

- *Choroidal rupture:*
  - Rupture is seen as one or two curved white lines **(Figs 16.9.2 and 16.9.3)**
  - Early rupture may be associated with Berlin's edema **(Fig. 16.9.4)**
  - Concentric with the disk margin and on its temporal side

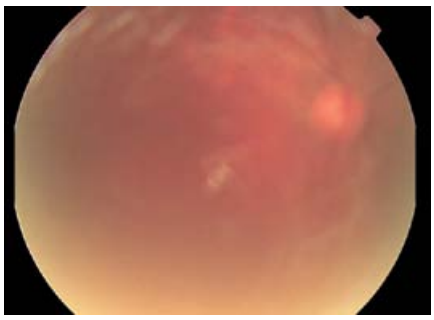


Fig. 16.9.1: Vitreous hemorrhage—blunt trauma

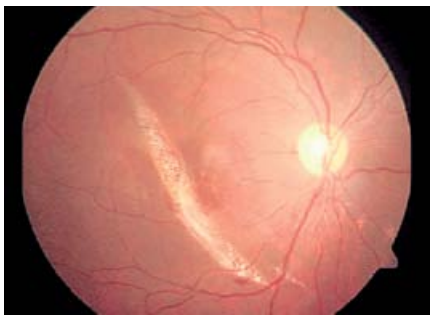


Fig. 16.9.2: Choroidal rupture—late

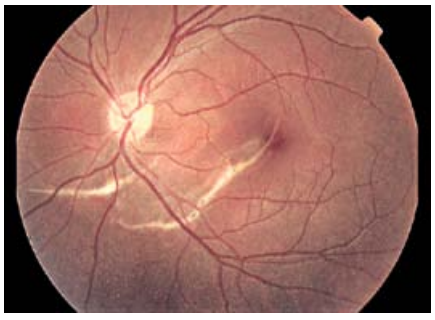


Fig. 16.9.3: Choroidal rupture—two ruptured lines and one through fovea

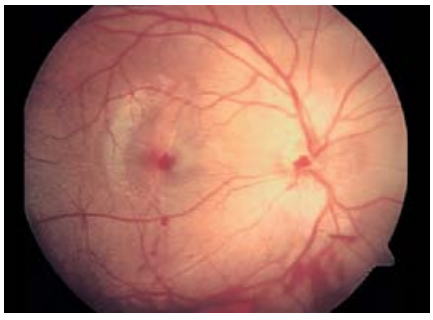


Fig. 16.9.4: Choroidal rupture—early with Berlin's edema

- Edges of this line are often pigmented and also with tractional RD **(Fig. 16.9.5)**

- *Traumatic sub-RPE hemorrhage*, may also be associated with choroidal rupture **(Figs 16.9.6A and B)**
- *Choroidal hemorrhage or hematoma:* **(Fig. 16.9.7)**

Retina

- *Commotio retinae* (Berlin's edema):
  - Milky white cloudiness at the macular area due to edema
  - Often with a cherry-red spot **(Fig. 16.10.1)**
  - Later there may be pigmentary deposits at the macula
- *Traumatic macular hemorrhage* **(Fig. 16.10.2)** with the formation of a macular cyst and subsequently, a *macular hole* **(Fig. 16.10.3)**
- Preretinal and multiple retinal and subretinal hemorrhages **(Figs 16.10.4 and 16.10.5)**

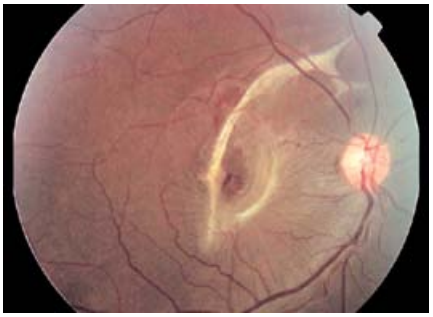


Fig. 16.9.5: Choroidal rupture through the fovea with small tractional RD

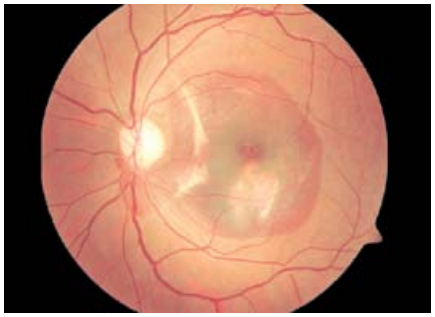


Fig. 16.9.6A: Sub-RPE hemorrhage with choroidal tear

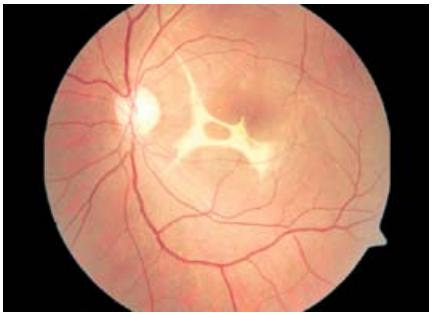


Fig. 16.9.6B: Sub-RPE hemorrhage with choroidal tear with scar formation

Fig. 16.9.7: Choroidal hemorrhage

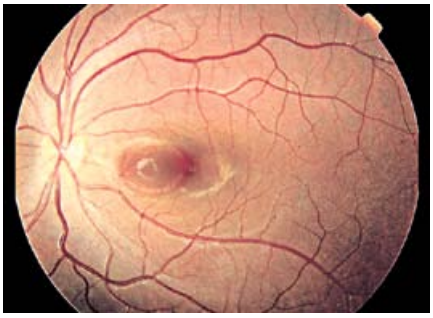


Fig. 16.10.1: Berlin's edema—cherry-red spot

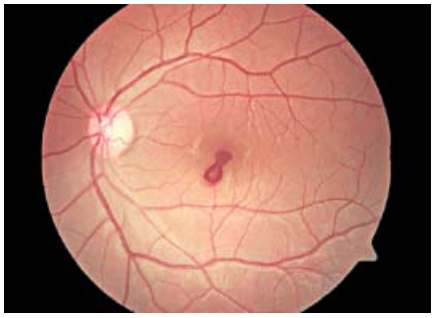


Fig. 16.10.2: Traumatic macular hemorrhage

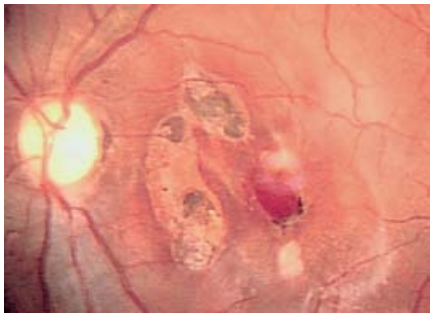


Fig. 16.10.3: Traumatic macular hole

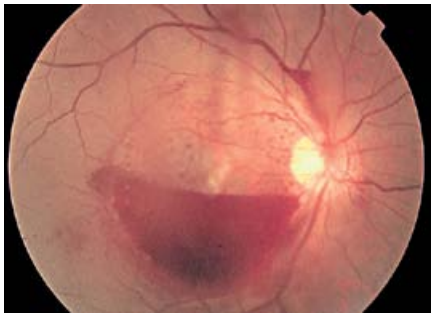


Fig. 16.10.4: Traumatic preretinal hemorrhage

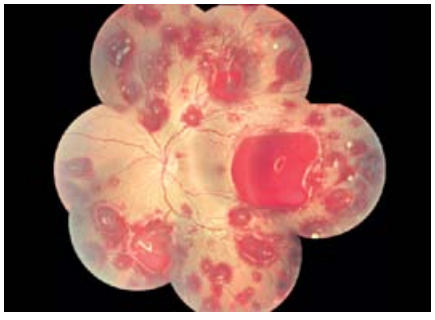


Fig. 16.10.5A: Traumatic preretinal, subretinal and multiple retinal hemorrhages

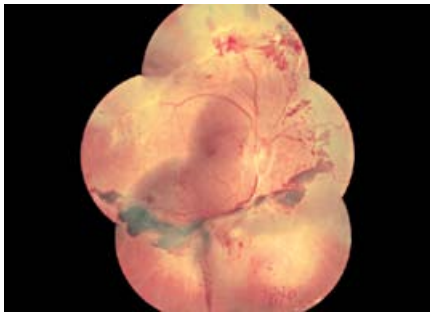


Fig. 16.10.5B: Traumatic retinal and subretinal hemorrhages

- Retinal tears at periphery with rhegmatogenous retinal detachment, especially in myopic subjects **(Figs 16.10.6A and B)**
- Tractional retinal detachment **(Fig. 16.10.7)**
- Traumatic proliferative vitreoretinopathy usually secondary to vitreous hemorrhage **(Figs 16.10.8 and 16.10.9).**

Optic Nerve

- Optic nerve sheath hemorrhage **(Fig. 16.11.1)**
- Traumatic peripapillary hemorrhage **(Fig. 16.11.2)**
- Avulsion of the optic nerve leading to optic atrophy, which may be partial or total **(Fig. 16.11.3).**

Orbit

- *Blow-out fracture:* Pure blow-out fracture is typically caused by a sudden rise in intraorbital pressure by a striking object which is greater than 5 cm in diameter, like tennis ball or blow by a first **(Fig. 16.12.1)**

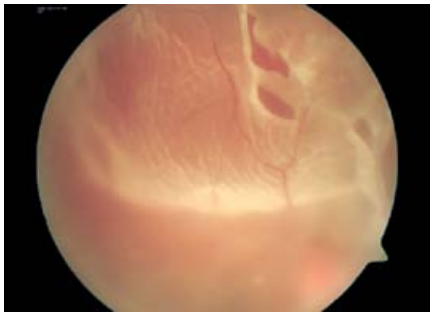


Fig. 16.10.6A: Traumatic rhegmatogenous retinal detachment

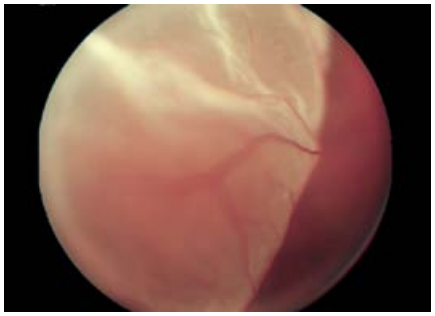


Fig. 16.10.6B: Traumatic rhegmatogenous retinal detachment

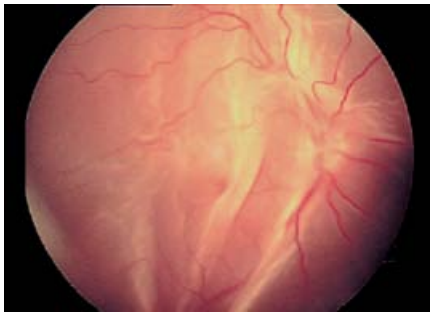


Fig. 16.10.7: Traumatic tractional detachment

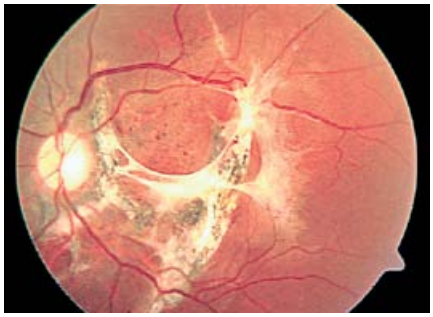


Fig. 16.10.8: Traumatic proliferative vitreoretinopathy

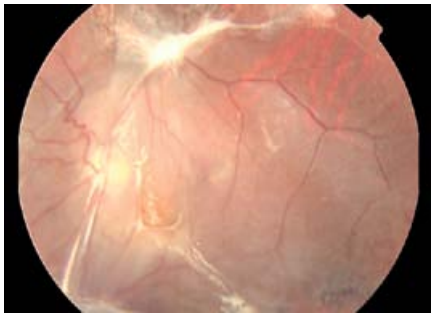


Fig. 16.10.9: Traumatic proliferative vitreoretinopathy

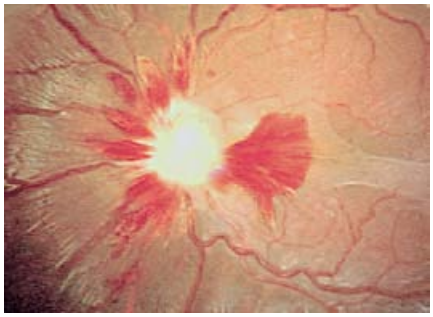


Fig. 16.11.1: Optic nerve sheath hemorrhage

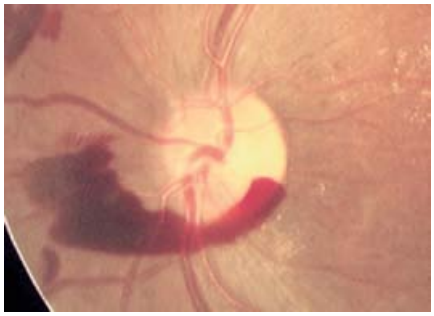


Fig. 16.11.2: Peripapillary hemorrhage

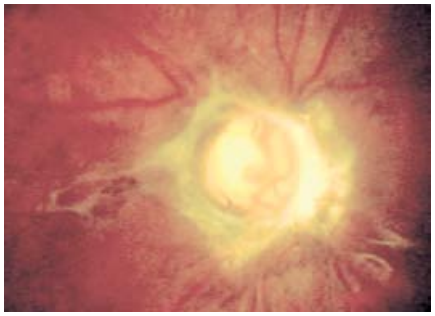


Fig. 16.11.3: Avulsion of optic nerve with atrophy

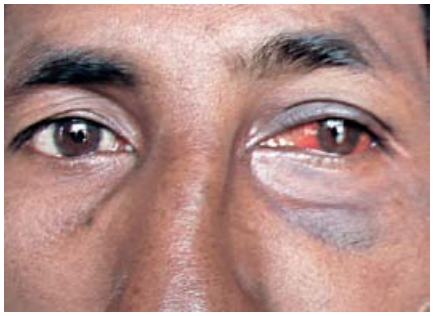


Fig. 16.12.1: Blow-out fracture (LE)—enophthalmos

- Periocular ecchymosis and edema
- Enophthalmos with pseudoptosis usually appears after 10–14 days
- Infraorbital nerve anesthesia
- Nasal bleeding and subconjunctival hemorrhage
- Restriction of ocular movement in upgaze **(Figs 16.12.2A and B)**
- Forced duction test (FDT) is positive
- X-ray and CT scan (typical teardrop sign within maxillary sinus) is diagnostic **(Fig. 16.12.2C)**

- Fractures of the orbital bones
- Retrobulbar hemorrhage **(Fig. 16.12.3)**
- Orbital emphysema and proptosis.

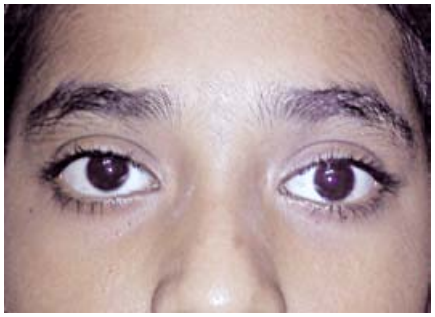


Fig. 16.12.2A: Blow-out (orbit floor) fracture

Fig. 16.12.2B: Blow-out (orbit floor) fracture—restriction in upgaze

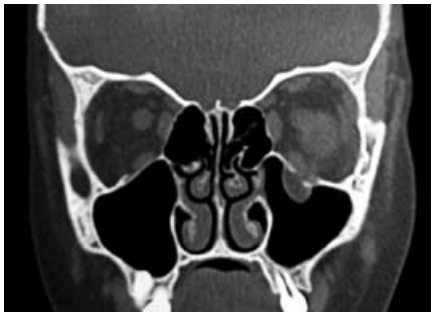


Fig. 16.12.2C: Blow-out fracture—teardrop sign on left side within maxillary antrum

Fig. 16.12.3: Retrobulbar hemorrhage

Penetrating (Perforating) Injuries

- They are caused by sharp objects or projectile foreign bodies
- All perforating injuries are potentially serious
- So, the patients should be urgently admitted and treated promptly
- ***Penetrating injuries*** by definition penetrate into the eye but not through and through there is no exit wound
- ***Perforating injuries*** have both entry and exit wounds (a through and through injury)
- S*eriousness arises from*:
  - Immediate effect of trauma
  - Introduction of infection
  - Chance of sympathetic ophthalmia.

Immediate Effect of Trauma

- Wounds of the lid
  - Upper lid injury **(Figs 16.13.1A to C)**
  - Lower lid and canalicular injury **(Fig. 16.13.2)**

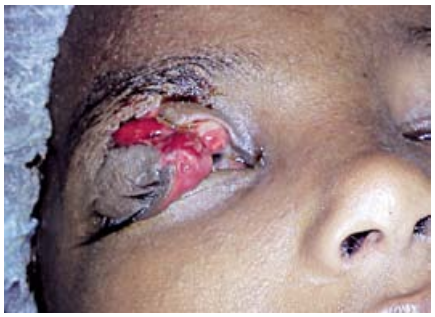


Fig. 16.13.1A: Cut injury of lid

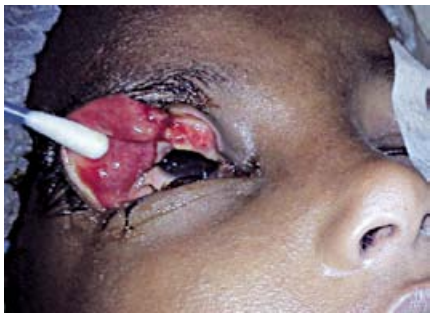


Fig. 16.13.1B: Cut injury of lid

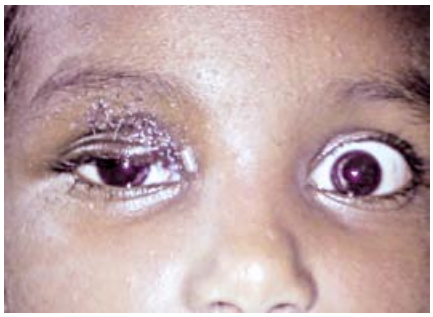


Fig. 16.13.1C: Cut injury of lid after repair—same patient

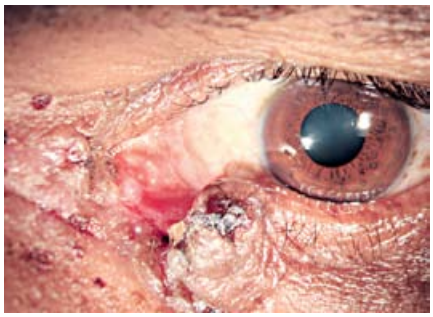


Fig. 16.13.2: Lid injury—lower canaliculus

  - Faulty repair may give rise to lid notching **(Fig. 16.13.3)**
  - Cut injury of the lid may be of partial thickness **(Fig. 16.13.4)**
- Conjunctival tear by sharp object **(Figs 16.13.5A and B)**
- Traumatic pyogenic granuloma may occur if the reapir is not done on time **(Fig. 16.13.6)**

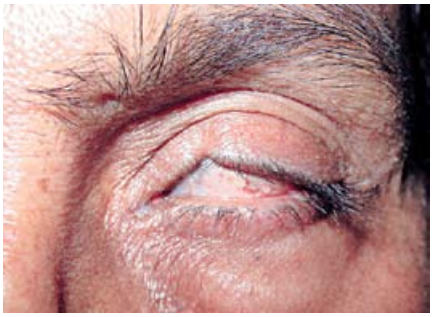


Fig. 16.13.3: Lid notching following lid repair

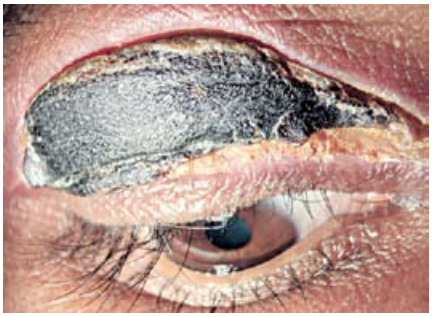


Fig. 16.13.4: Lid injury—partial thickness and old

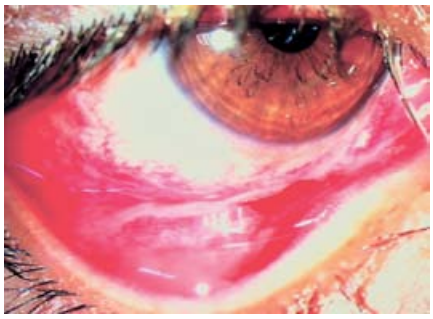


Fig. 16.13.5A: Conjunctival tear

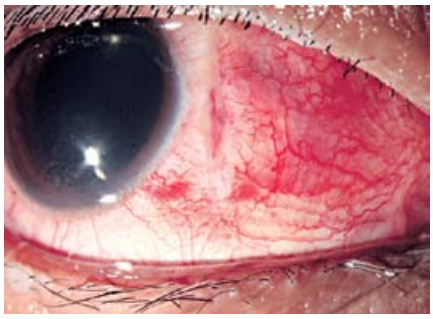


Fig. 16.13.5B: Conjunctival tear

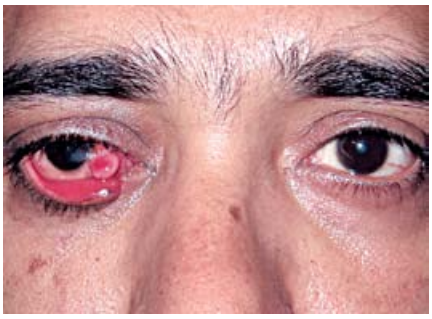


Fig. 16.13.6: Traumatic pyogenic granuloma with canalicular tear—RE

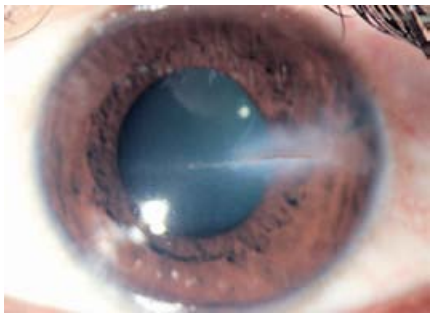


Fig. 16.13.7A: Corneal rupture—linear

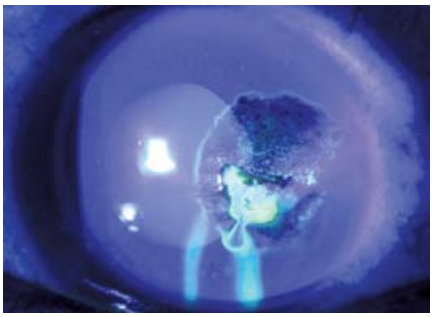


Fig. 16.13.7B: Corneal rupture—Seidel's test positive

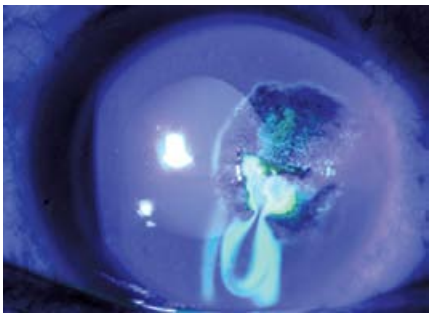


Fig. 16.13.7C: Corneal rupture—Seidel's test positive

- *Wounds of the cornea:*
  - May be linear **(Figs 16.13.7A to C)**, curved **(Figs 16.13.8A and B)**, zigzag **(Figs 16.13.9A to C),** triradiate **(Figs 16.13.10A and B)** and lacerated **(Figs 16.13.11 and 16.13.12)**

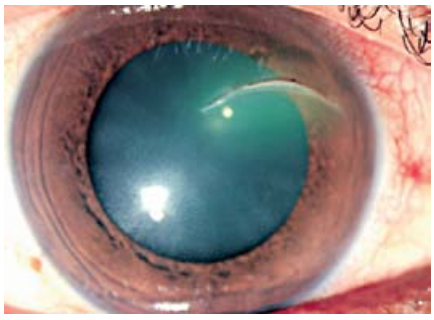


Fig. 16.13.8A: Corneal rupture—curved

Fig. 16.13.8B: Corneal rupture—curved

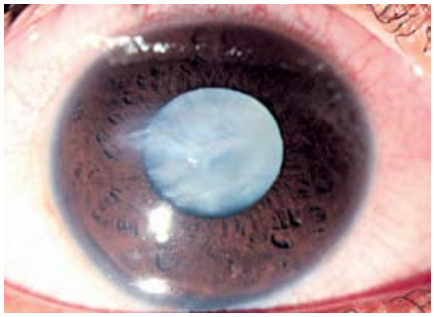


Fig. 16.13.9A: Corneal rupture—zigzag

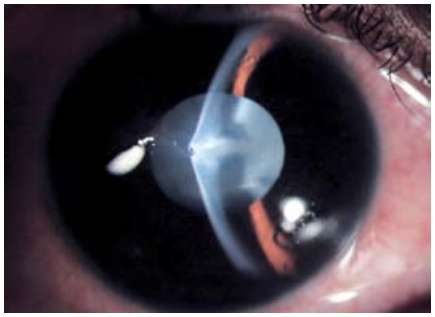


Fig. 16.13.9B: Corneal rupture—zigzag

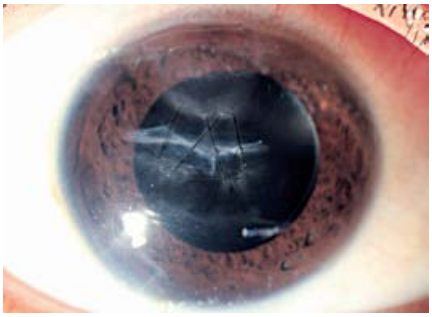


Fig. 16.13.9C: Corneal rupture—zigzag—after repair of same eye

- May be small **(Figs 16.13.13A and B)**, large or corneoscleral **(Figs 16.13.14 and 16.13.15)** with or without iris prolapse
- May be at the limbus
- May be associated with iris prolapse or frank iris incarceration **(Figs 16.13.16A and B)**
- May be lamellar **(Figs 16.13.17 and 16.13.18)**
- Margins soon swell-up after the injury, and become cloudy

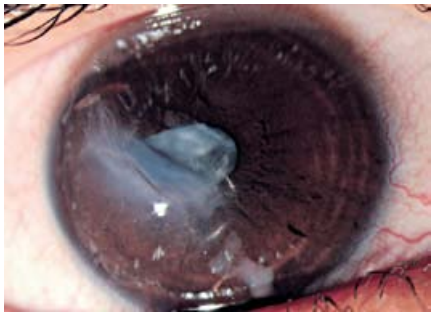


Fig. 16.13.10A: Corneal rupture—triradiate

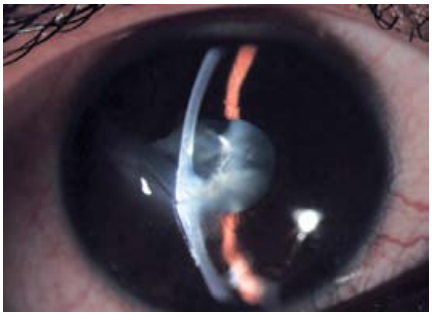


Fig. 16.13.10B: Corneal rupture—triradiate

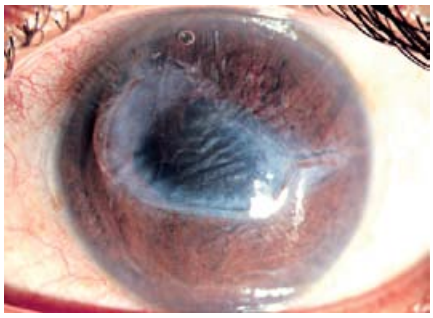


Fig. 16.13.11A: Corneal rupture—curved and lacerated

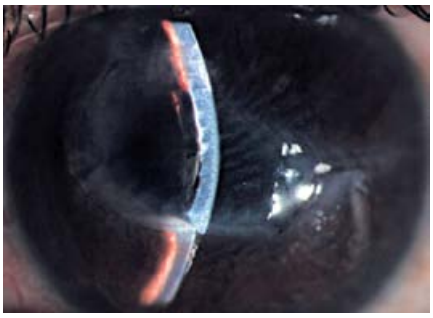


Fig. 16.13.11B: Corneal rupture—curved and lacerated

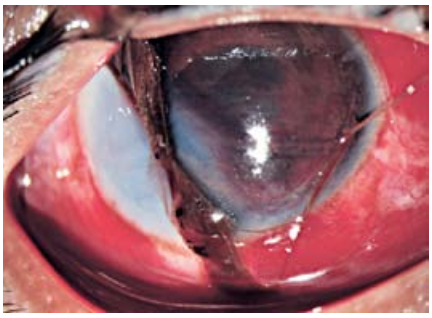


Fig. 16.13.12: Corneal rupture—large and lacerated

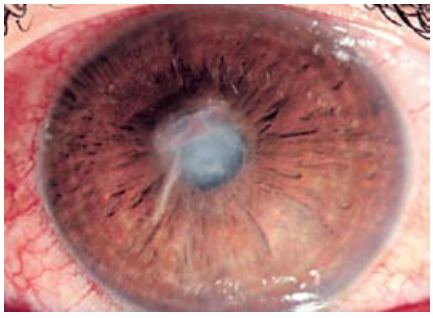


Fig. 16.13.13A: Corneal rupture—small lacerated

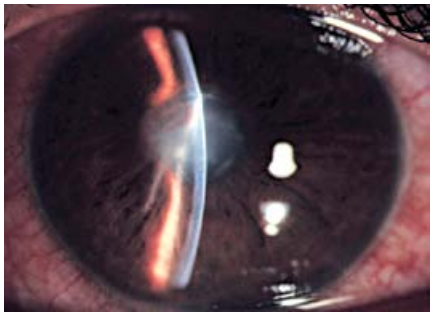


Fig. 16.13.13B: Corneal rupture—small lacerated

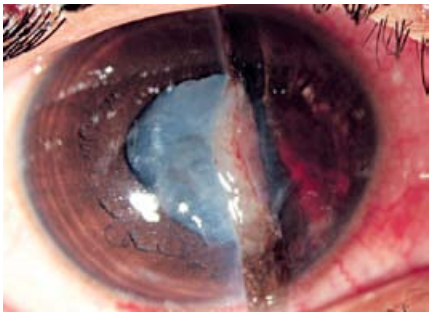


Fig. 16.13.14: Corneoscleral rupture

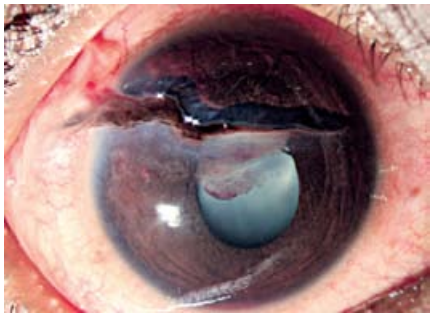


Fig. 16.13.15A: Corneoscleral rupture

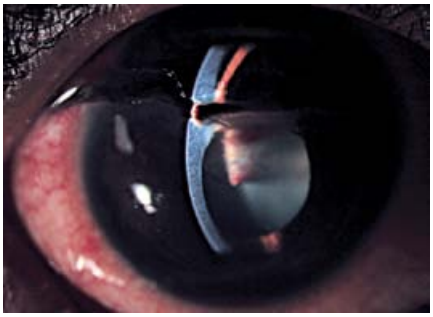


Fig. 16.13.15B: Corneoscleral rupture

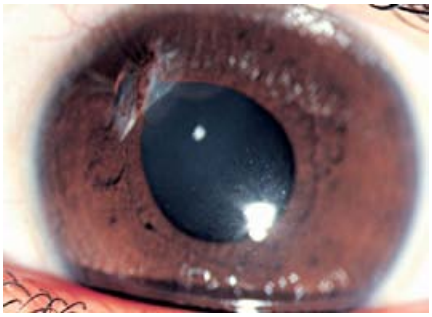


Fig. 16.13.16A: Corneal perforation with iris incarceration

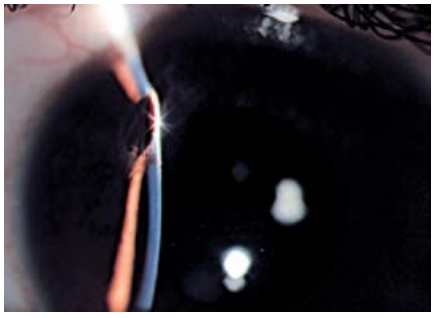


Fig. 16.13.16B: Corneal perforation with iris incarceration

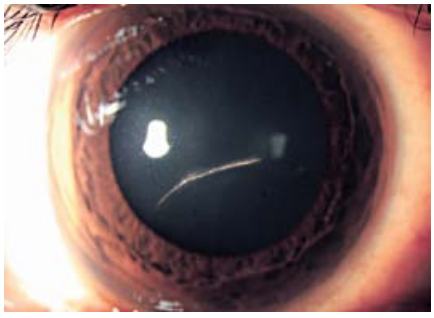


Fig. 16.13.17: Lamellar injury of cornea

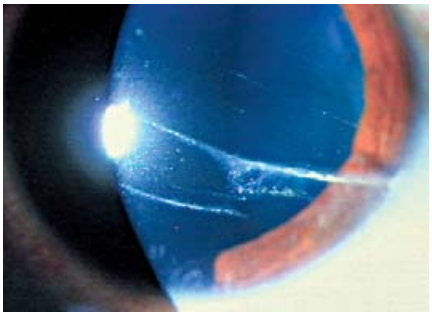


Fig. 16.13.18: Lamellar injury of cornea

- With round pointed object there may be tissue loss **(Figs 16.13.19A and B)** with a positive Seidel's test
- There may be total rupture of the globe

- *Treatment:* Urgent repair of the wound. In case of round perforation cyanoacrylate glue or patch graft is required **(Figs 16.13.20A to D)**
  - Phthisis bulbi may occur even after repair **(Figs 16.13.21A and B)**

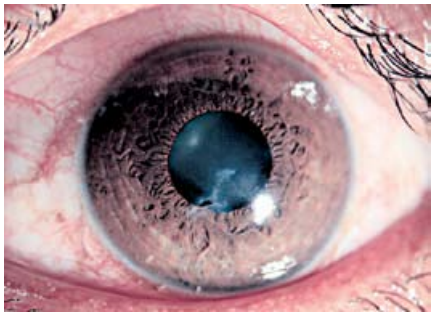


Fig. 16.13.19A: Small corneal perforation—small tissue loss

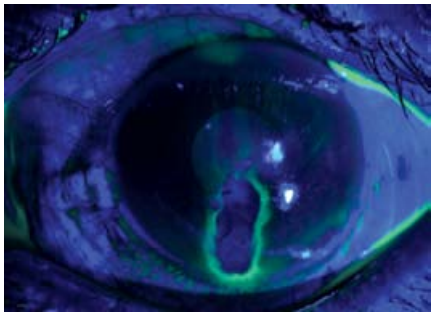


Fig. 16.13.19B: Small corneal perforation—Seidel's test positive

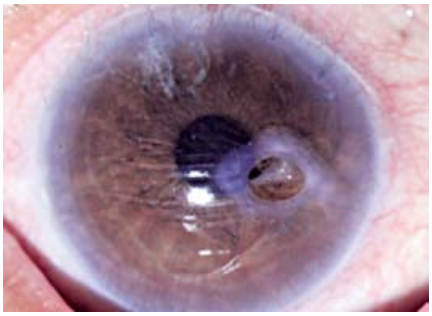


Fig. 16.13.20A: Corneal injury with tissue loss

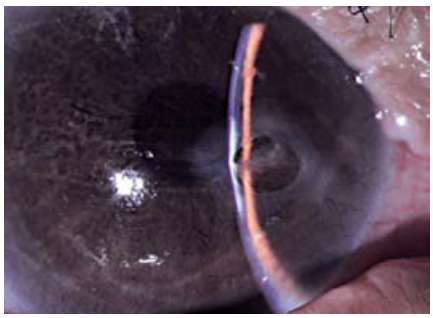


Fig. 16.13.20B: Corneal injury with tissue loss

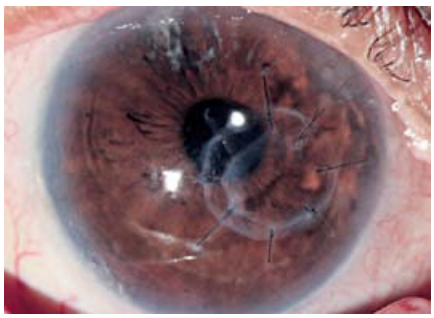


Fig. 16.13.20C: Corneal injury with tissue loss

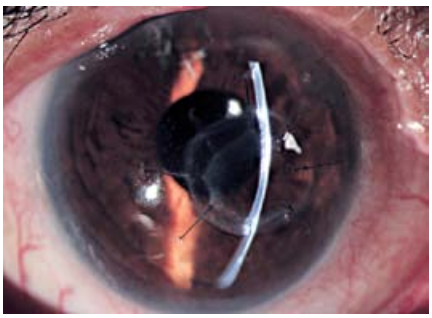


Fig. 16.13.20D: Corneal injury with tissue loss

- *Wounds of the sclera*
  - Recognized by the uveal incarceration or prolapse **(Figs 16.13.22 and 16.13.23)**
  - May be associated with vitreous prolapse **(Figs 16.13.24A and B)** if the lesion is more posterior
- *Wounds of the lens and its capsule*
  - Localized lens opacity **(Figs 16.13.25A to C)**
  - May be with early rosette cataract **(Fig. 16.13.26)**
  - Total lens opacity with or without capsular tear **(Figs 16.13.27 to 16.13.29)**
  - In case of large capsular wound, the flocculent white cortical matters protrude through the capsular opening **(Figs 16.13.30 and 16.13.31)**
  - In some cases, anterior chamber is full of white flocculi **(Figs 16.13.32 and 16.13.33)**
- Injuries in other parts of the eye in cases of severe impact

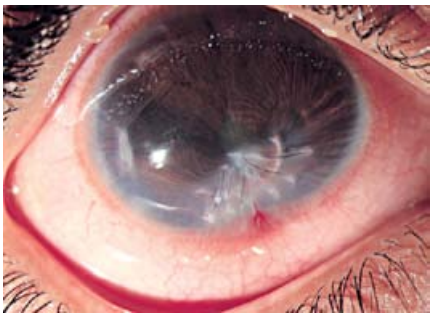


Fig. 16.13.21A: Corneal injury repaired with phthisis bulbi

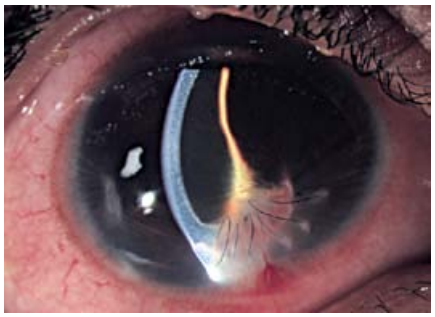


Fig. 16.13.21B: Corneal injury repaired with phthisis bulbi

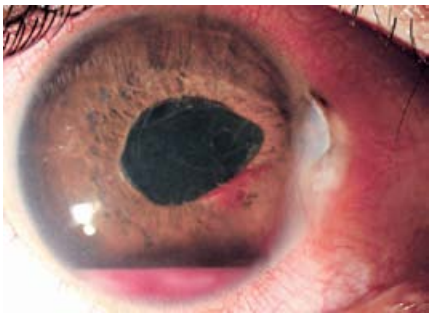


Fig. 16.13.22: Scleral perforation—uveal incarceration

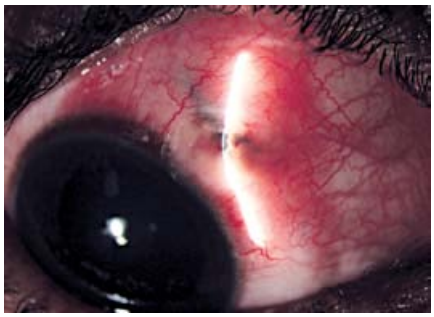


Fig. 16.13.23: Scleral perforation—uveal prolapse

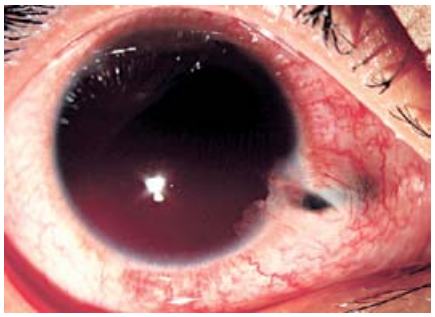


Fig. 16.13.24A: Scleral perforation—uveal and vitreous prolapse

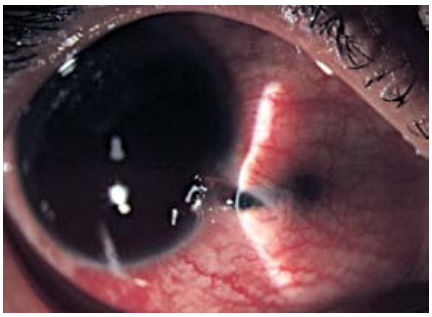


Fig. 16.13.24B: Scleral perforation—uveal and vitreous prolapse

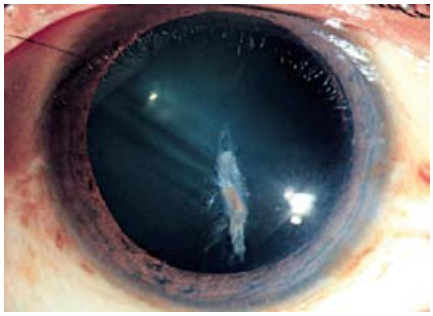


Fig. 16.13.25A: Local cataract—sealed capsular tear

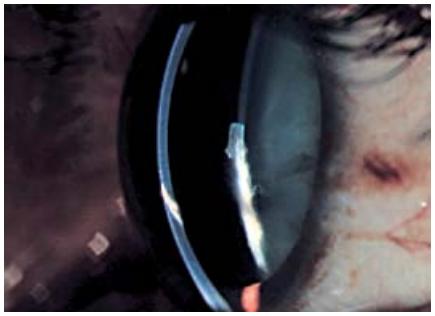


Fig. 16.13.25B: Local cataract—note corneal wound of entry

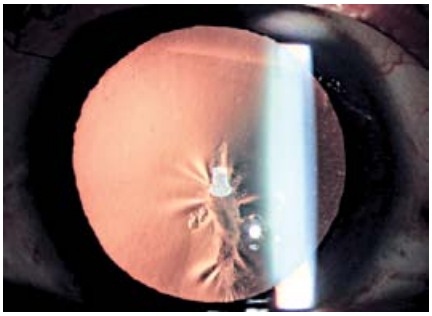


Fig. 16.13.25C: Local cataract—sealed capsular tear

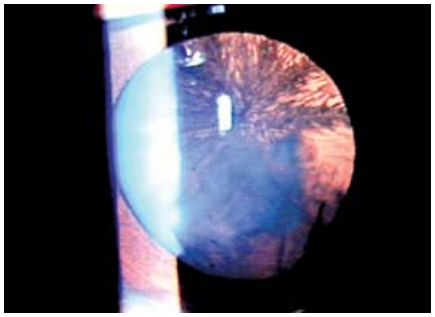


Fig. 16.13.26: Early rosette cataract following corneal perforation

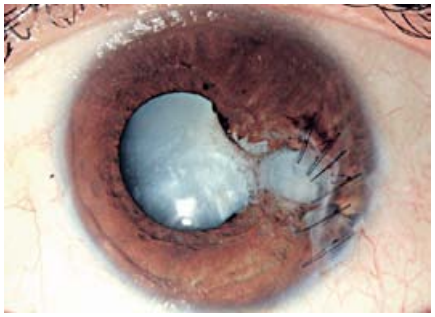


Fig. 16.13.27A: Total cataract—capsular tear

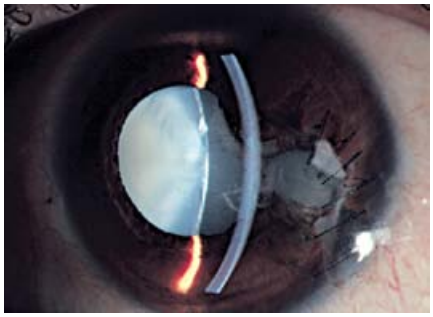


Fig. 16.13.27B: Total cataract

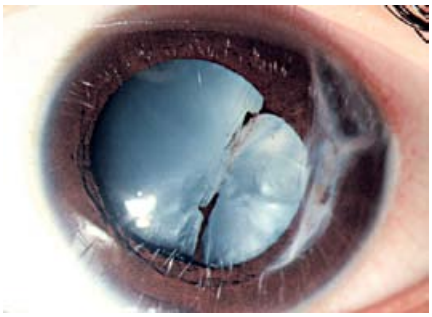


Fig. 16.13.28: Total cataract—capsular tear

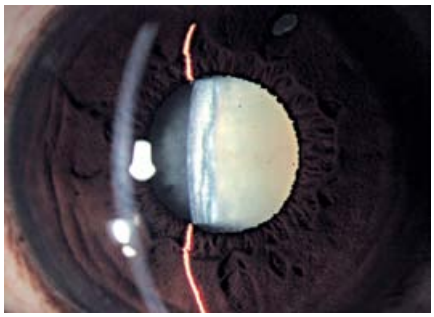


Fig. 16.13.29: Total cataract

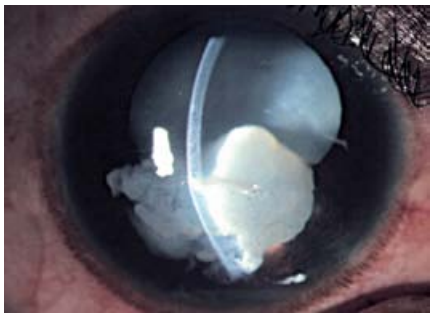


Fig. 16.13.30: Lens matter in AC

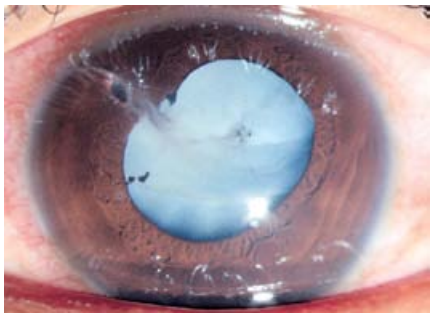


Fig. 16.13.31: Lens matter in AC

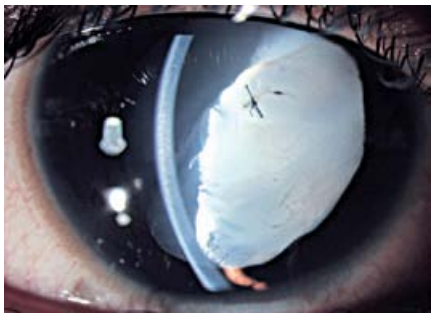


Fig. 16.13.32: Lens matter in AC touching the cornea

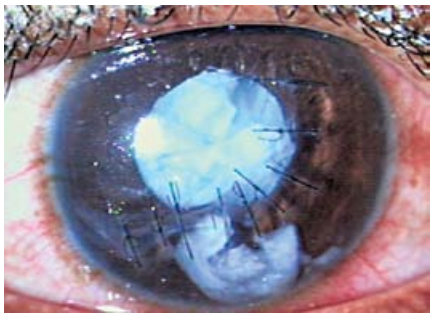


Fig. 16.13.33: Lens matter in AC

- *Iatrogenic globe perforation by needle:* May happen during peribulbar or retrobulbar block **(Fig. 16.13.34).** It is an emergency situation.

Signs of Globe Perforation

- Conjunctival chemosis
- Shallow or flat anterior chamber
- Alteration of papillary size, shape and location
- Focal iris tear or hole
- Injury tract in the cornea, sclera, lens or vitreous
- Wound leak (a positive Seidel's test)
- Hypotony in case of larger perforation.

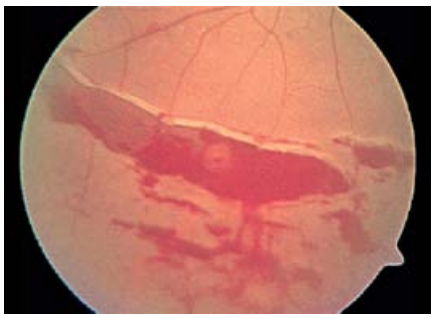


Fig. 16.13.34: Globe puncture during peribulbar injection

Introduction of Infection

- Suppurative keratitis **(Figs 16.14.1A and B)**
- Purulent iridocyclitis with hypopyon **(Fig. 16.14.2)**
- Endophthalmitis **(Figs 16.14.3 and 16.14.5)**
- Panophthalmitis in extreme situation
- *Lental abscess:*
  - After a sharp penetrating injury with direct inoculation of organism inside the lens
  - Yellowish exudate visible within the lens **(Figs 16.14.6 and 16.14.7).**

Sympathetic Ophthalmia

Most dreadful complication of a penetrating injury—discussed in **Chapter 6.**

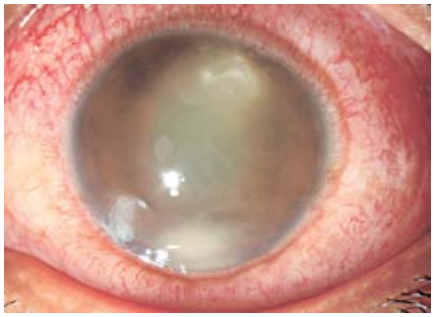


Fig. 16.14.1A: Penetrating injury—keratitis, purulent iritis and hypopyon

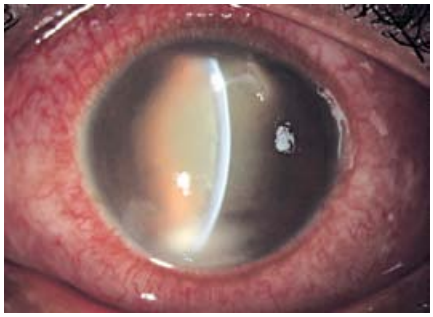


Fig. 16.14.1B: Penetrating injury—keratitis, purulent iritis and hypopyon

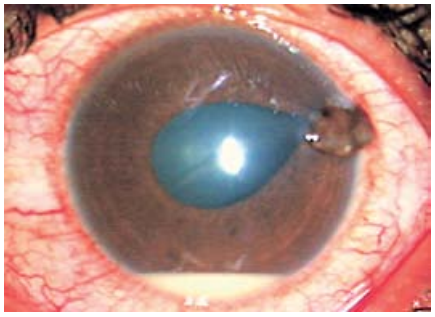


Fig. 16.14.2: Penetrating injury—purulent iritis with hypopyon

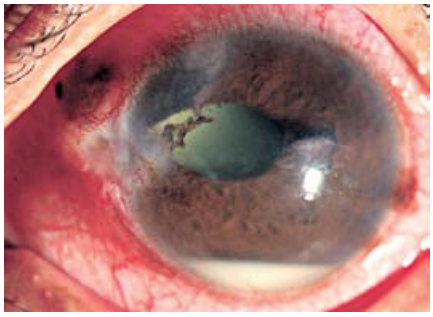


Fig. 16.14.3: Penetrating injury—endophthalmitis

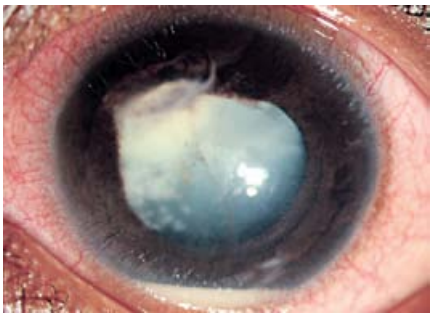


Fig. 16.14.4A: Penetrating injury—endophthalmitis

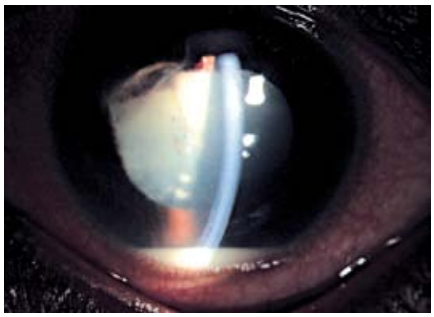


Fig. 16.14.4B: Penetrating injury—endophthalmitis

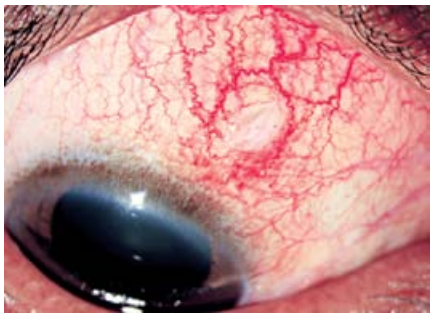


Fig. 16.14.5A: Penetrating injury through sclera—endophthalmitis

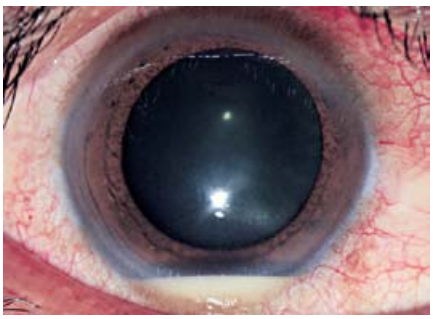


Fig. 16.14.5B: Penetrating injury through sclera—same eye

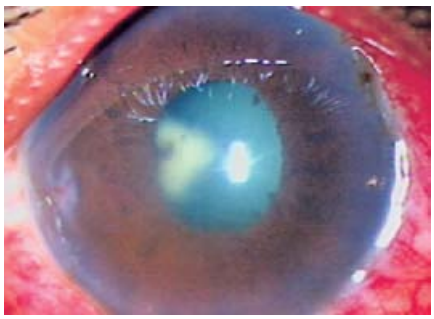


Fig. 16.14.6: Penetrating injury—lental abscess

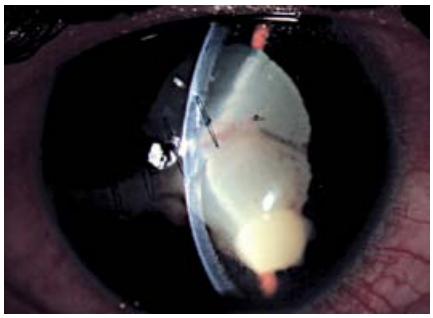


Fig. 16.14.7: Penetrating injury—lental abscess

Foreign Bodies in the Eye

Extraocular Foreign Bodies

- Small foreign bodies—coal, dust, sand, iron particles, eyelash, wood-piece, husks of seeds, wings of insects, etc. may pitch upon the conjunctiva, cornea or the limbus.

Conjunctival Foreign Body

- *Commonest site:* At the middle of the upper subtarsal sulcus **(Figs 16.15.1A and B)**
- May be embedded in bulbar conjunctiva, fornix **(Figs 16.15.2 and 16.15.3)** or at the limbus **(Figs 16.15.4 and 16.15.5)**
- Lid eversion is a must for all cases if there is corneal abrasion or scratch marks on the cornea **(Figs 16.15.6A to C)**
- *Treatment:* Easy removal with a sterile cotton bud in most of the cases.

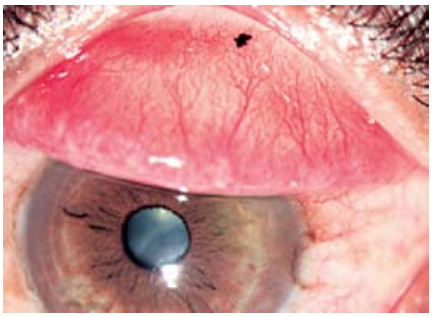


Fig. 16.15.1A: Foreign body—supratarsal sulcus

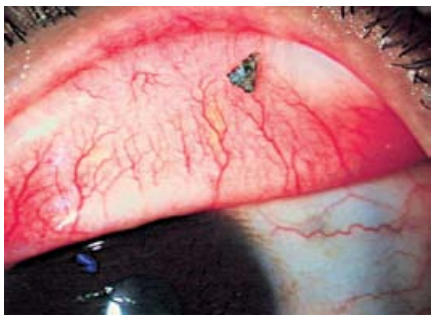


Fig. 16.15.1B: Foreign body—supratarsal sulcus

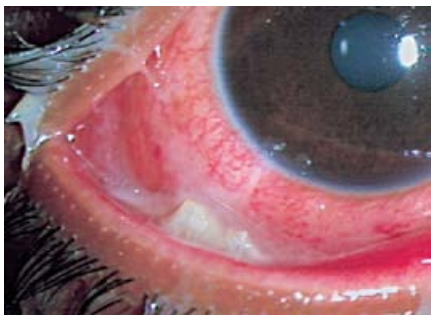


Fig. 16.15.2: Foreign body at the fornix

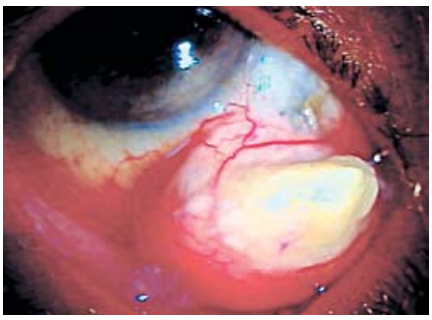


Fig. 16.15.3: Large foreign body at the fornix

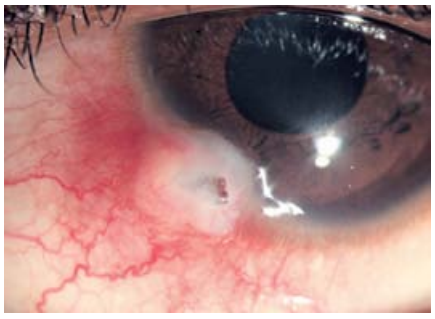


Fig. 16.15.4A: Foreign body conjunctiva with granuloma—at the limbus

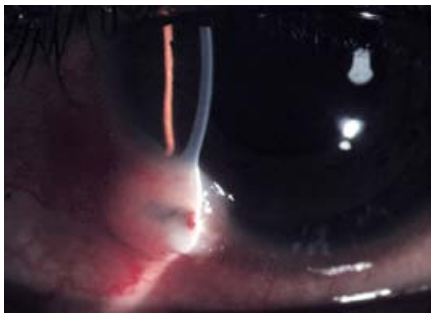


Fig. 16.15.4B: Foreign body conjunctiva with granuloma—at the limbus

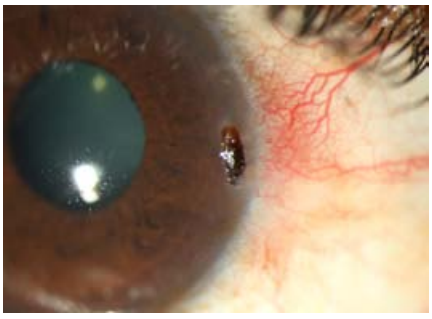


Fig. 16.15.5: Foreign body just inside limbus—insect wing

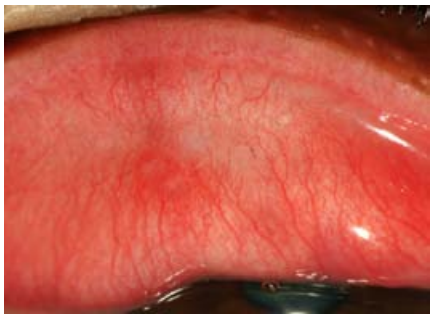


Fig. 16.15.6A: Foreign body—CP hair in tarsal conjunctiva

Fig. 16.15.6B: Foreign body—CP hair in tarsal conjunctiva

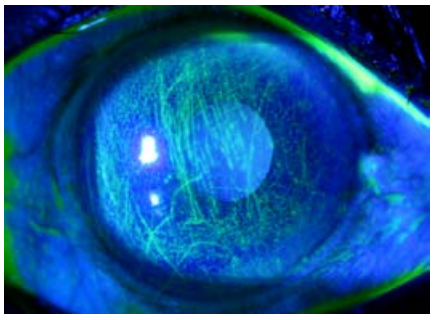


Fig. 16.15.6C: CP hairs in tarsal conjunctiva—linear scratch marks on cornea

Corneal Foreign Body

- Foreign body sensation
- Marked photophobia and redness
- Ciliary congestion
- Particle may be:
  - Superficial **(Figs 16.16.1 and 16.16.2)**,
  - Anterior stromal **(Figs 16.16.3 and 16.16.4)**,
  - Deep posterior stromal **(Figs 16.16.5 and 16.16.6)**, or

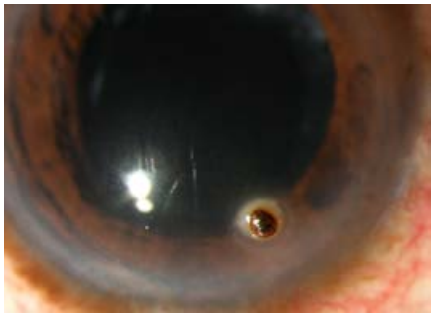


Fig. 16.16.1A: Foreign body cornea—superficial

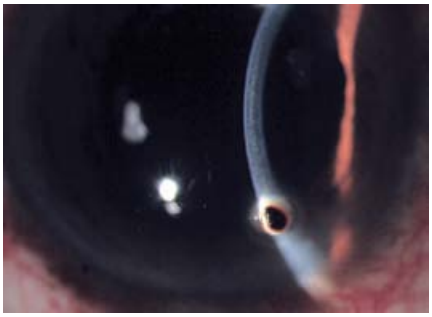


Fig. 16.16.1B: Foreign body cornea—superficial

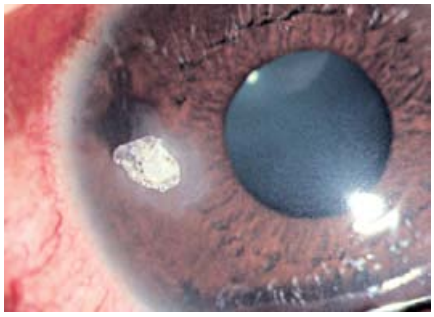


Fig. 16.16.2A: Foreign body cornea—superficial

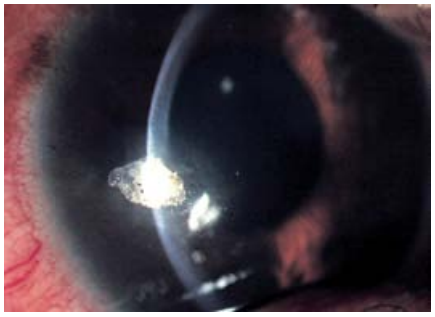


Fig. 16.16.2B: Foreign body cornea—superficial

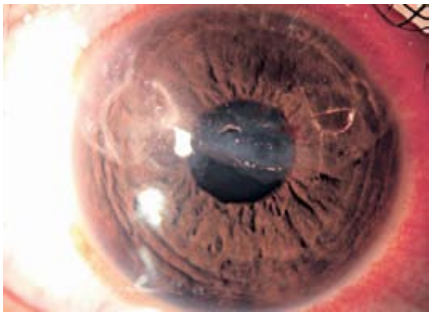


Fig. 16.16.3A: Foreign body cornea—glass—anterior stromal

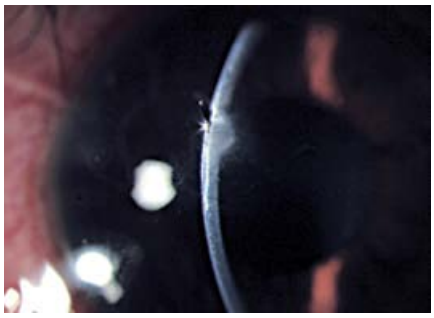


Fig. 16.16.3B: Foreign body cornea—glass—anterior stromal

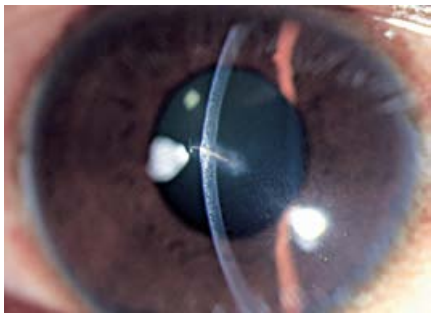


Fig. 16.16.4A: Foreign body cornea—vegetable matter—anterior stromal

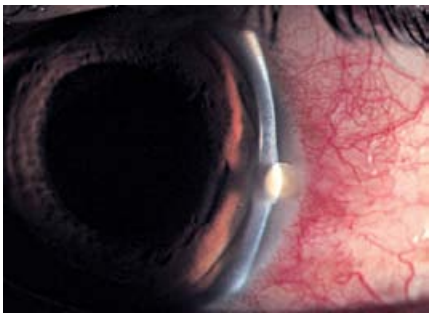


Fig. 16.16.4B: Foreign body cornea—anterior stromal

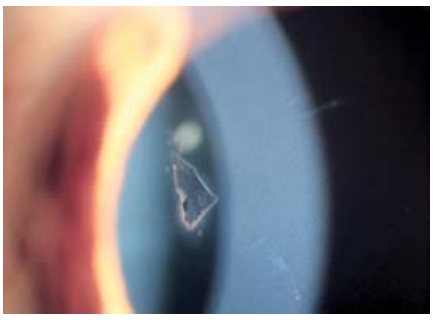


Fig. 16.16.5A: Foreign body cornea—glass—deep stromal

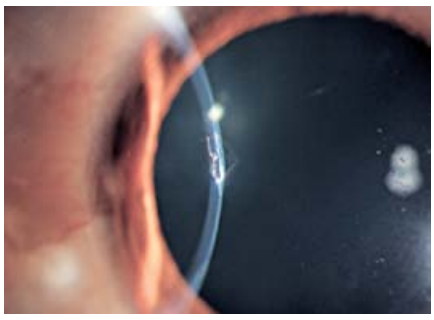


Fig. 16.16.5B: Foreign body cornea—glass—deep stromal

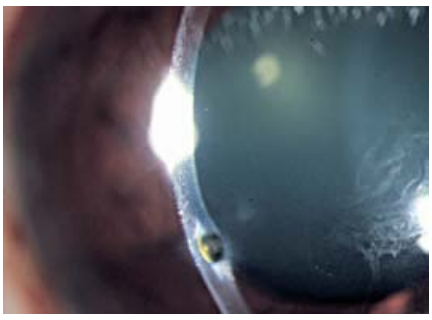


Fig. 16.16.6: Foreign body cornea—metal—deep posterior stromal

  - Partly in the anterior chamber and partly within the cornea **(Figs 16.16.7A and B)**
  - Partly in the anterior chamber and partly outside the cornea **(Fig. 16.16.8)**
- In old cases of iron foreign body, there may be associated iron-rust ring **(Figs 16.16.9 and 16.16.10)**
- In addition, there may be surrounding infiltration **(Fig. 16.16.11 and see Figs 16.16.1A and B)** or frank corneal ulcer
- Check for tarsal conjunctiva for foreign body and chronic dacryocystitis in all cases of corneal foreign bodies
- *Treatment:* Removal of foreign body by a disposable needle under strict aseptic condition and broad spectrum antibiotic drops and ointment for few days.

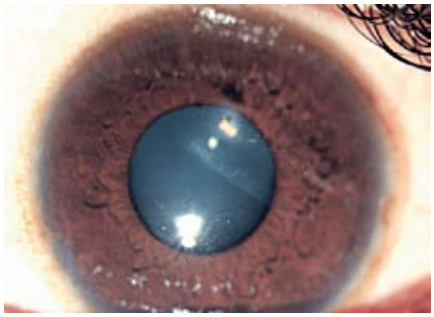


Fig. 16.16.7A: Foreign body—vegetable matter—partly in AC

Fig. 16.16.7B: Foreign body—vegetable matter—partly in AC

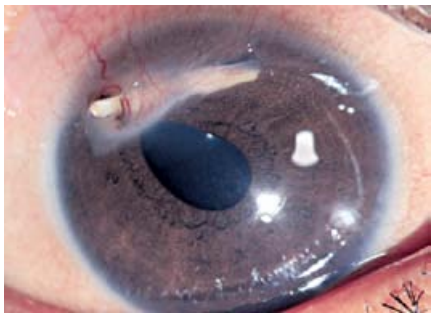


Fig. 16.16.8: Penetrating injury—vegetable matter—partly in AC and partly outside

Fig. 16.16.9: Foreign body cornea—rust ring

Intraocular Foreign Body (IOFB)

- While chipping the stone, with an iron-chisel and a hammer, it is a chip of the chisel (from cutting edge, or from its mushroomed head) which enters the eyes, but not the chip of the stone
- May pass directly through the conjunctiva, cornea **(Fig. 16.17.1)** or sclera **(Figs 16.17.2A and B)**, or sometimes through the lid and then sclera **(Figs 16.17.3A to C)**

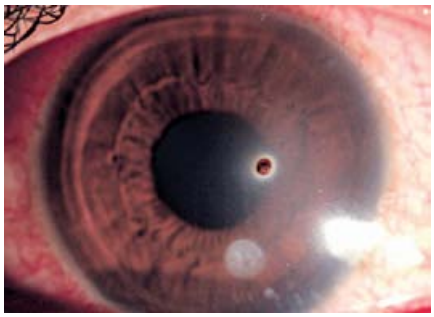


Fig. 16.16.10: Foreign body cornea—rusty deposits

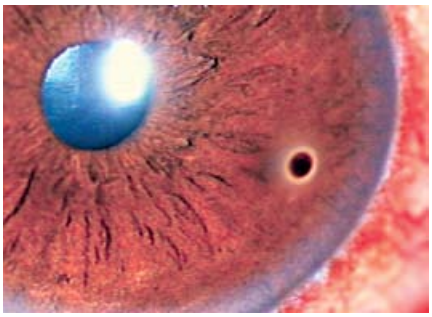


Fig. 16.16.11: Foreign body cornea with ring infiltration around the foreign body

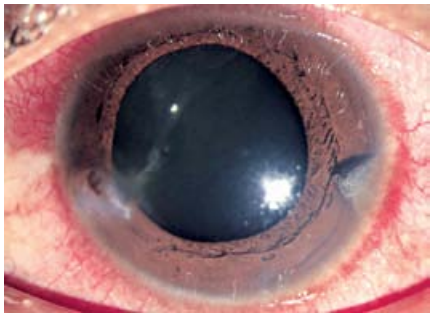


Fig. 16.17.1: Retained IOFB at 3 o'clock position—wound of entry—cornea at opposite side

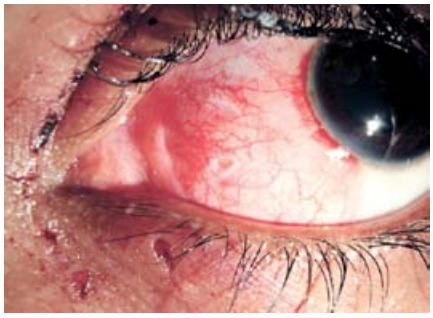


Fig. 16.17.2A: Retained IOFB—wound of entry—sclera

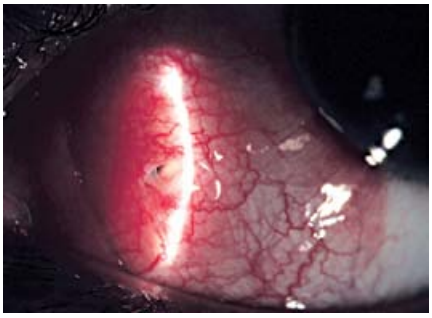


Fig. 16.17.2B: Retained IOFB—wound of entry—sclera

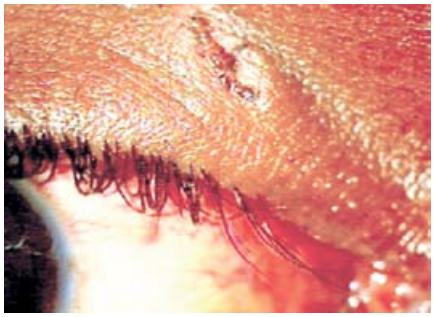


Fig. 16.17.3A: Retained IOFB—wound of entry—through lid and then sclera

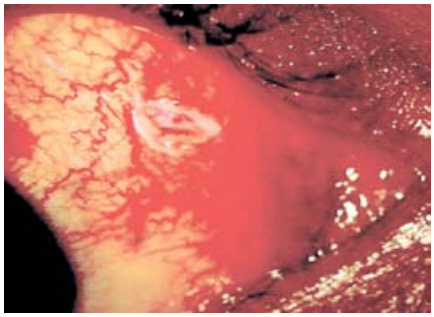


Fig. 16.17.3B: Retained IOFB—wound of entry—through lid and then sclera

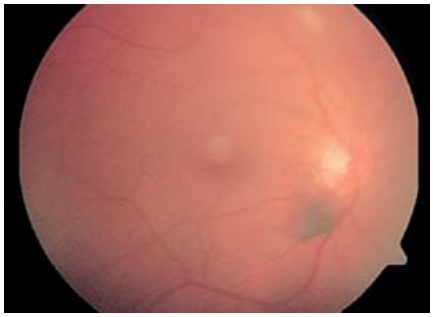


Fig. 16.17.3C: Retained IOFB—on the retina same eye

- IOFB may retain in the iris **(Figs 16.17.4 and 16.17.5)** or anterior chamber angle **(Figs 16.17.6 to 16.17.8)**
- May lodge within the crystalline lens **(Figs 16.17.9 and 16.17.10)** causing cataract or siderotic cataract **(Figs 16.17.11A and B)**
- Passes through the iris causing iris hole **(Figs 16.17.12 and 16.17.13)** or through the pupil, and causes a traumatic localized cataract or total cataract **(Fig. 16.17.14)**

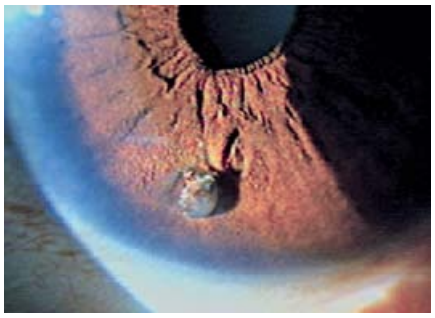


Fig. 16.17.4: Retained IOFB—on the iris surface

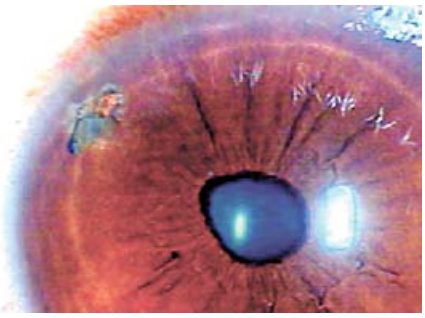


Fig. 16.17.5: Retained IOFB—in the iris substance

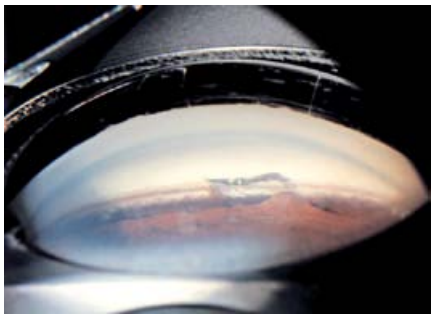


Fig. 16.17.6: Retained IOFB—angle of AC seen gonioscopically

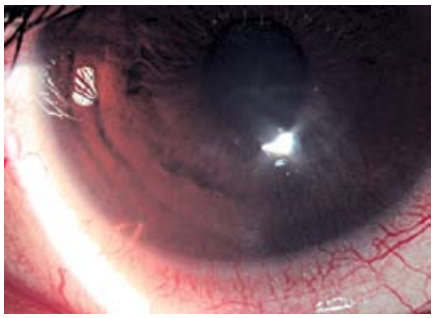


Fig. 16.17.7: Retained IOFB—Gold FB in AC

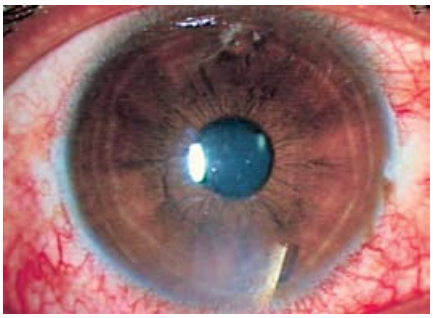


Fig. 16.17.8: Retained IOFB—angle of AC

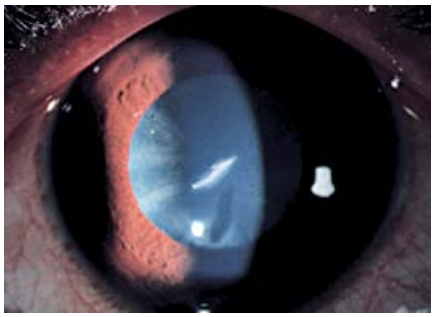


Fig. 16.17.9: Retained intralenticular FB—with cataract

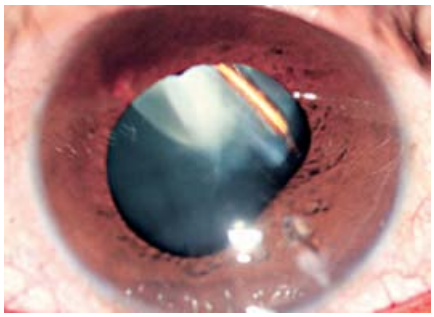


Fig. 16.17.10A: Retained IOFB—intralenticular Gold

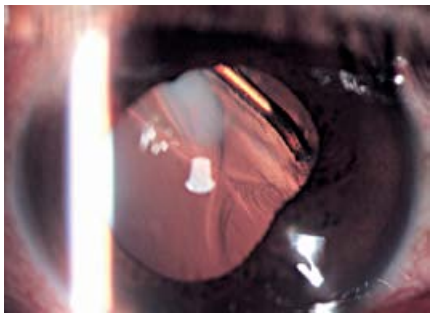


Fig. 16.17.10B: Retained IOFB—intralenticular Gold

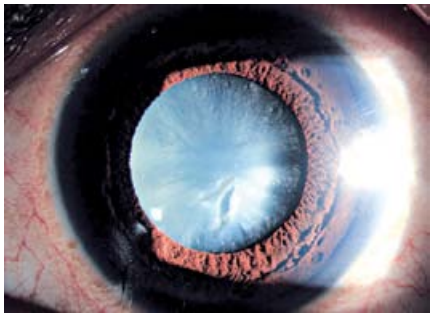


Fig. 16.17.11A: Retained intralenticular FB—cataract same eye of **Figure 16.17.9**

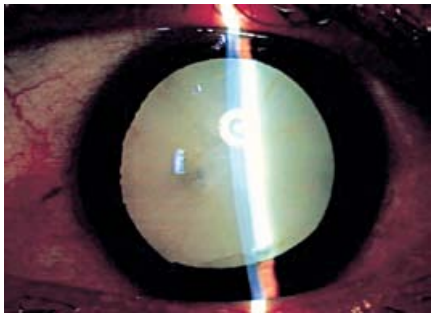


Fig. 16.17.11B: Retained IOFB within the lens—siderosis

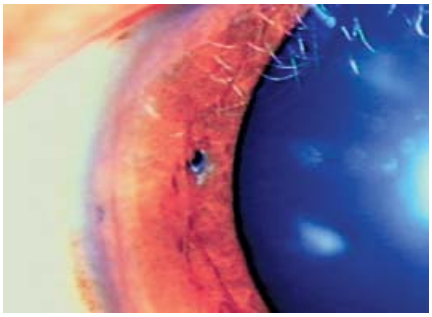


Fig. 16.17.12: Retained IOFB—iris hole

- May retain in the vitreous **(Fig. 16.17.15)** and also cause vitreous hemorrhage **(Fig. 16.17.16)**
- May rest on to the retina **(Figs 16.17.17A to C)** and may be with retinal or preretinal hemorrhage **(Fig. 16.17.18)**

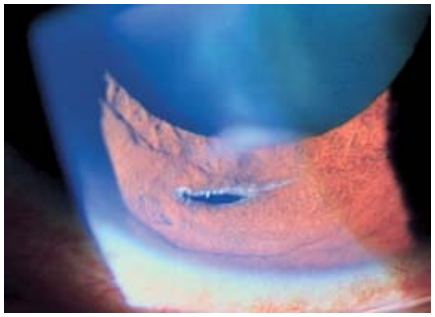


Fig. 16.17.13: Retained IOFB—iris hole

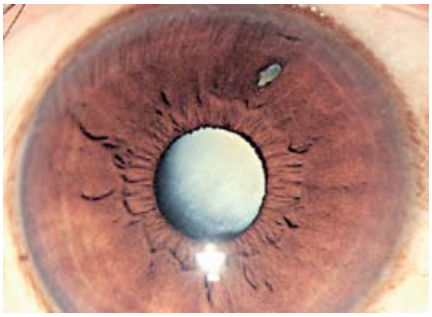


Fig. 16.17.14: Retained IOFB—iris hole

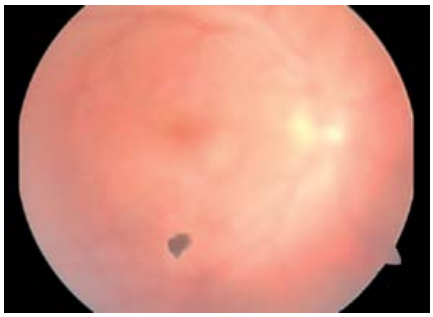


Fig. 16.17.15: Retained IOFB—in the vitreous

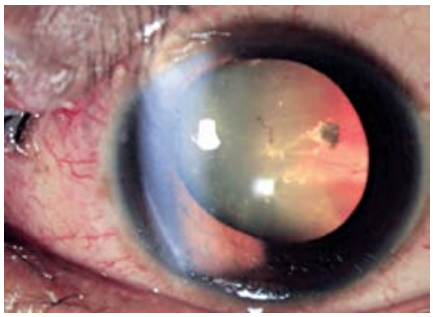


Fig. 16.17.16: IOFB—vitreous hemorrhage—wound of entry on nasal side

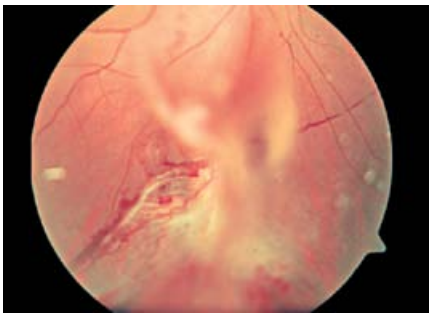


Fig. 16.17.17A: Retained IOFB—on retina

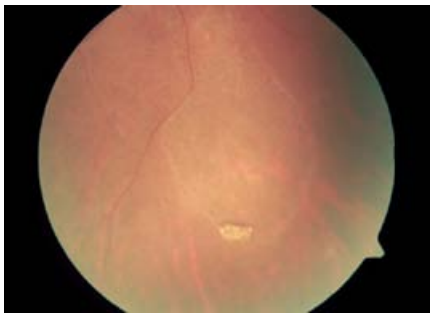


Fig. 16.17.17B: Retained IOFB—on retina

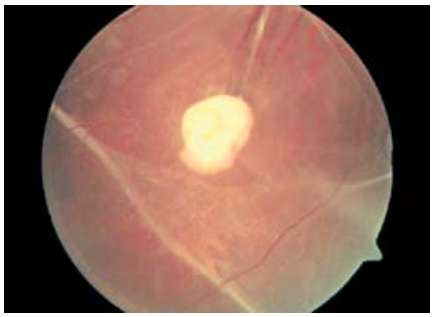


Fig. 16.17.17C: Retained IOFB—on retina

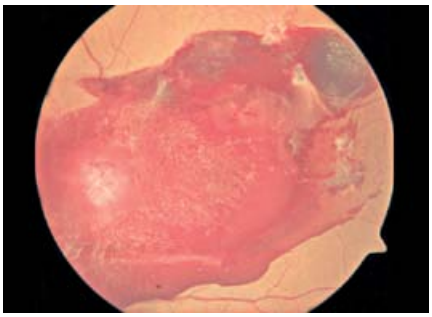


Fig. 16.17.18: Retained IOFB—on the retina with preretinal hemorrhage

- Rarely pierce the opposite wall and rest within the orbit (double perforation *or* penetrating injury) **(Figs 16.17.19 and 16.17.20)**
- May be associated with retinal hole, degeneration, or retinal detachment.

Siderosis Bulbi

- Chronic irreversible degenerative changes of the ocular tissues, caused by retained intraocular iron (and also steel, in proportion of its ferrous content) foreign body
- Rusty deposition on the anterior lens surface (earliest clinical sign) **(Fig. 16.18.1)**
- Frank siderotic cataract of variable degree **(Figs 16.18.2 to 16.18.4)**
- Heterochromia of the iris **(Figs 16.18.5 and 16.18.6)** and rusty discoloration of the cornea **(Figs 16.18.7A to C)**
- Retinal pigmentary changes with attenuation of blood vessels
- Prognosis is always poor.

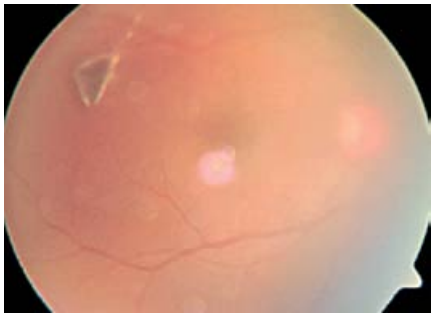


Fig. 16.17.19: Double perforation

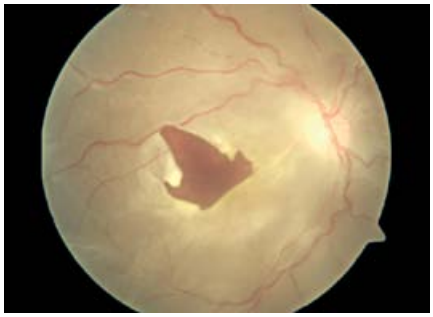


Fig. 16.17.20: Double perforation

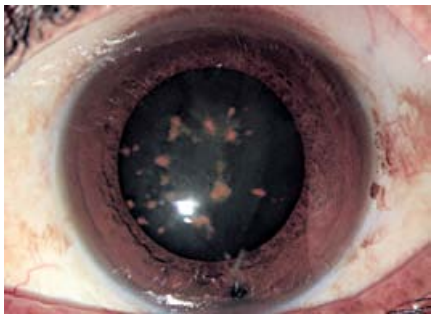


Fig. 16.18.1: Siderosis bulbi—rusty deposits—note the iris hole at 6 o'clock position

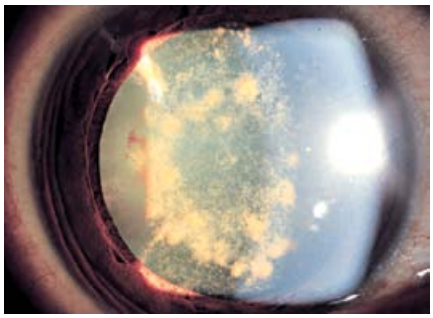


Fig. 16.18.2: Siderosis bulbi—rusty deposits and early cataract

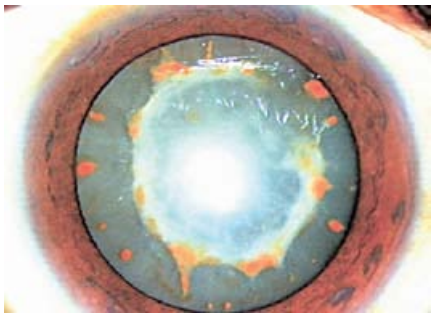


Fig. 16.18.3: Siderosis bulbi—siderotic cataract

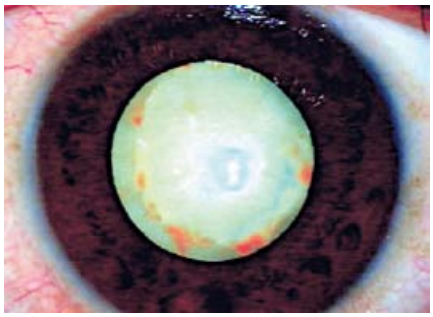


Fig. 16.18.4: Siderosis bulbi—siderotic total cataract

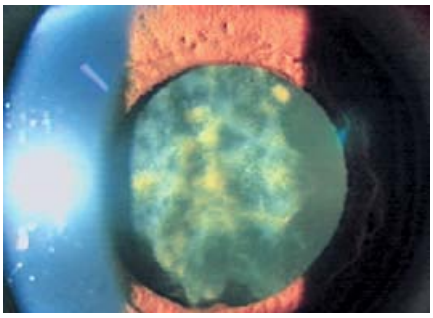


Fig. 16.18.5: Siderosis bulbi—iris discoloration

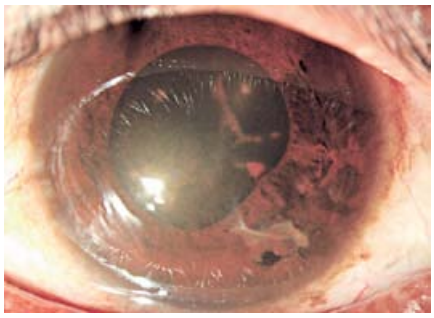


Fig. 16.18.6A: Siderosis bulbi—iris discoloration

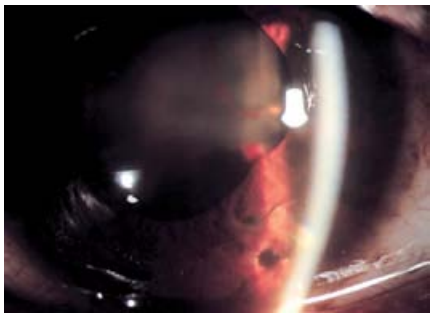


Fig. 16.18.6B: Siderosis bulbi—iris discoloration—note the iris hole

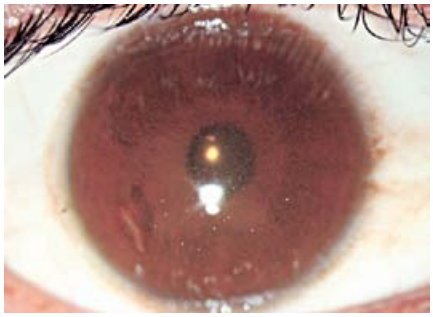


Fig. 16.18.7A: Siderosis bulbi—corneal deposits

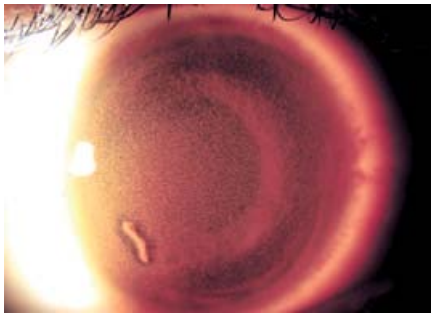


Fig. 16.18.7B: Siderosis bulbi—corneal deposits

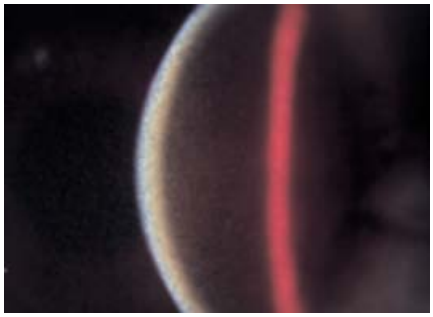


Fig. 16.18.7C: Siderosis bulbi—corneal deposits

Chalcosis Bulbi

- Kayser-Fleischer's (K-F) ring: A golden-brown ring at the level of Descemet's membrane of the cornea
- *Sun-flower cataract*: A brilliant golden-green sheen in the form of petals of sun-flower **(Figs 16.19.1 and 16.19.2)**
- Associated retinal changes with golden plaques
- Prognosis is always favorable.

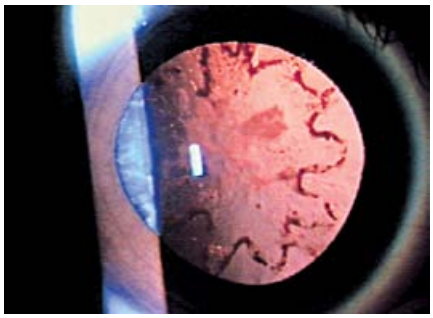


Fig. 16.19.1: Chalcosis bulbi—early sunflower cataract

Miscellaneous Organic Materials

- *Intraocular eyelashes:* Cause proliferation of the hair root epithelium leading to the formation of intraocular inflammation **(Figs 16.20.1 to 16.20.3)** or cyst formation

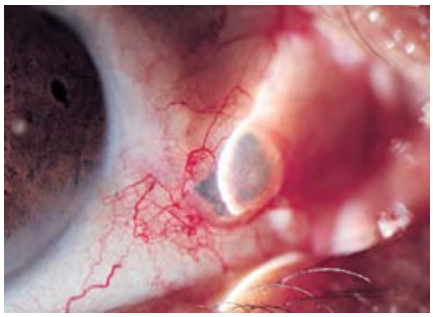


Fig. 16.19.2A: Chalcosis bulbi—copper foreign body

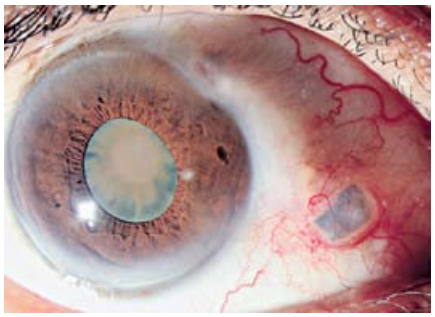


Fig. 16.19.2B: Chalcosis bulbi—sunflower cataract

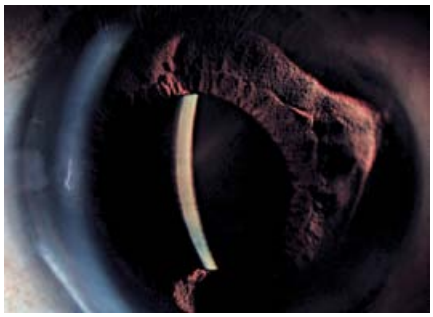


Fig. 16.19.2C: Chalcosis bulbi—sunflower cataract—golden green color—same eye

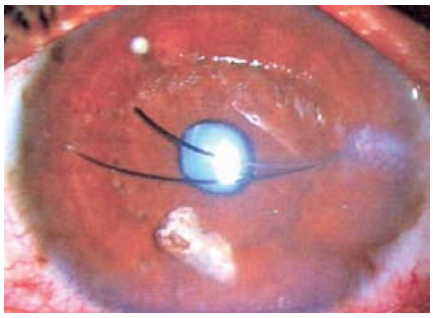


Fig. 16.20.1: Intraocular eyelashes

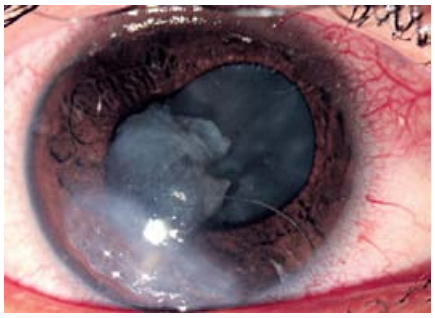


Fig. 16.20.2: Intraocular eyelashes

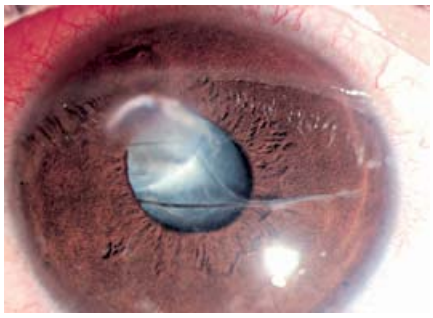


Fig. 16.20.3: Intraocular eyelashes

- *Caterpillar (CP) hairs*—they may lodge in various parts of the eyeball. They may be intracorneal with or without granuloma formation **(Figs 16.20.4 and 16.20.5)**. May lodge into the anterior chamber **(Fig. 16.20.6)**, pierce the iris **(Figs 16.20.7 and 16.20.8)** or on the lens surface causing localized cataract **(Fig. 16.20.9).** It also excites a chronic iridocyclitis with granulomatous nodules called *ophthalmia nodosa* **(Figs 16.20.10A and B)**
- Wood, stone or vegetable materials: Produce severe proliferative granulomatous reactions.

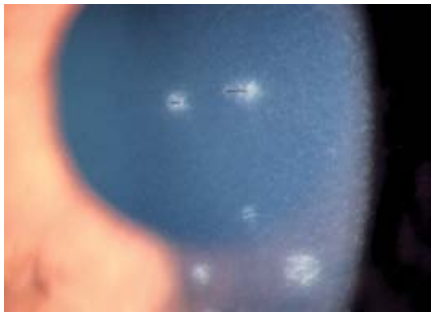


Fig. 16.20.4: Caterpillar hair—intracorneal

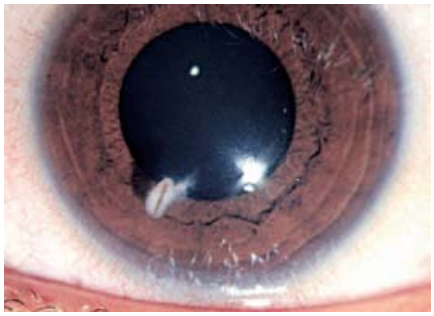


Fig. 16.20.5A: Retained CP hair—endothelial surface with granuloma formation

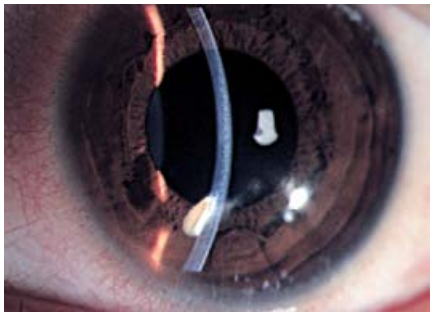


Fig. 16.20.5B: Retained CP hair—endothelial surface with granuloma formation

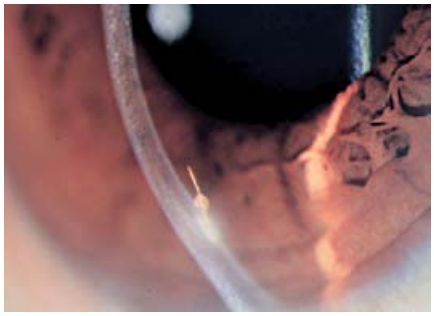


Fig. 16.20.6: Caterpillar hair—in AC

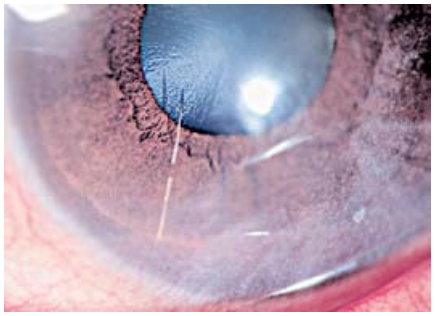


Fig. 16.20.7: Caterpillar hair—within the iris tissue

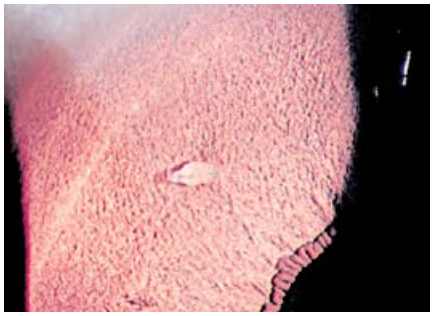


Fig. 16.20.8: Caterpillar hair—ophthalmia nodosa

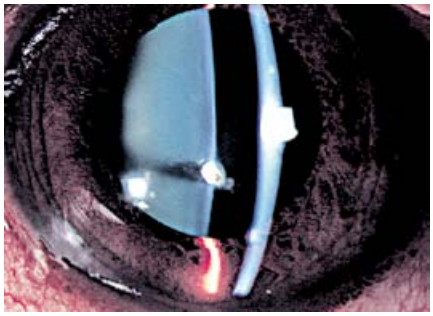


Fig. 16.20.9: Caterpillar hair—within the lens with cataract formation

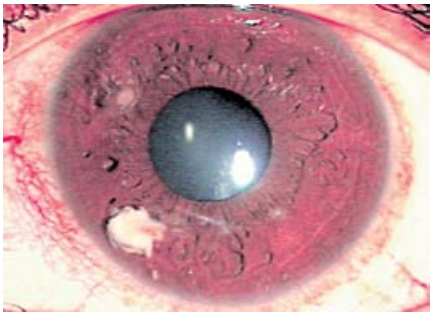


Fig. 16.20.10A: Caterpillar hair—ophthalmia nodosa

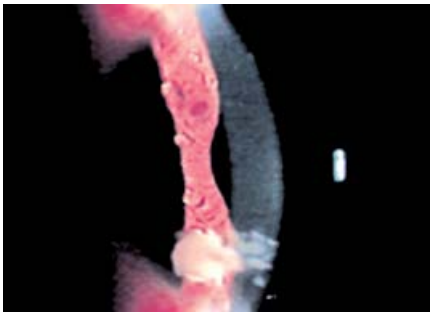


Fig. 16.20.10B: Caterpillar hair—ophthalmia nodosa

Chemical Injuries (Burns)

- Alkali burns are more dangerous than acid burns
- Severity depends on type of chemical, its quantity, pH of the solution, concentration of chemical agents, duration of exposure and the time of presentation **(Figs 16.21.1 and 16.21.2)**

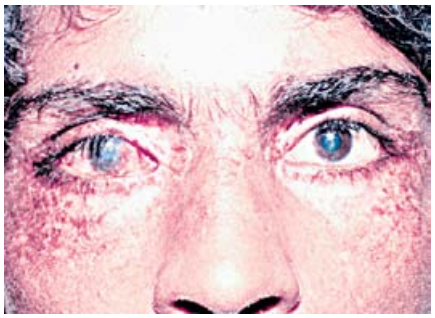


Fig. 16.21.1: Acid burn—moderate

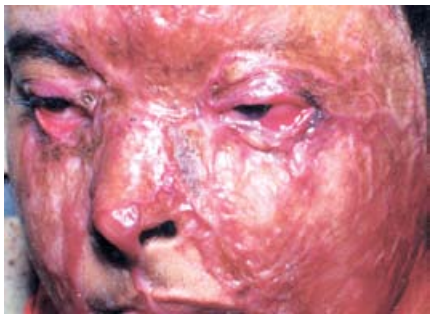


Fig. 16.21.2: Severe acid burn

- *Acute phase* (up to 1 week)
  - Chemosis of conjunctival and eyelids
  - Congestion and discharge **(Figs 16.21.3A and B)**
  - Corneal epithelial defects **(Fig. 16.21.4A)**
  - Perilimbal ischemia of variable degree and stromal clouding **(Fig. 16.21.4B)**
  - Increased IOP
- *Early reparative phase* (1–3 weeks):
  - Cornea and conjunctival regeneration
  - Persistent stromal edema and corneal epithelial defects **(Figs 16.21.5 to 16.21.7)**

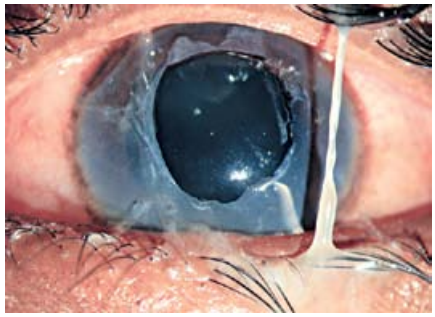


Fig. 16.21.3A: Acid burn in acute phase—moderate

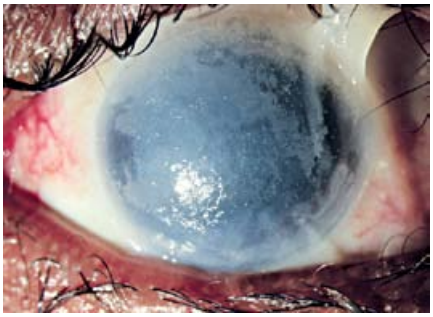


Fig. 16.21.3B: Alkali burn—acute phase

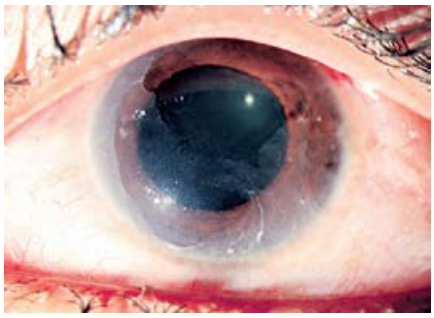


Fig. 16.21.4A: Acid burn in acute phase—epithelial defect

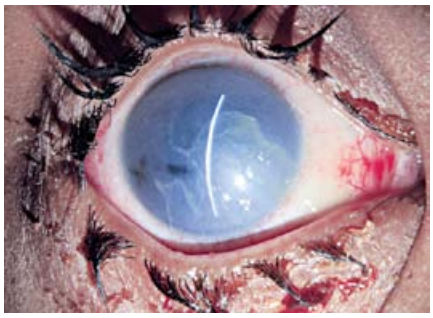


Fig. 16.21.4B: Alkali burn in acute phase—perilimbal ischemia and stromal clouding

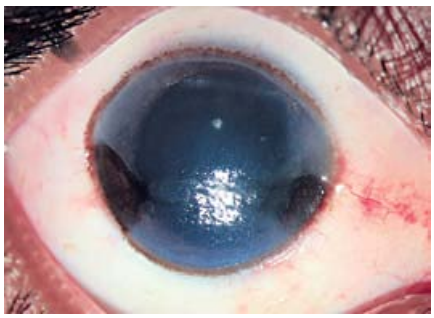


Fig. 16.21.5A: Alkali burn—stromal clouding

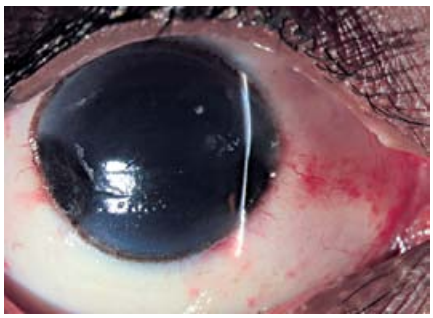


Fig. 16.21.5B: Alkali burn—epithelial defect

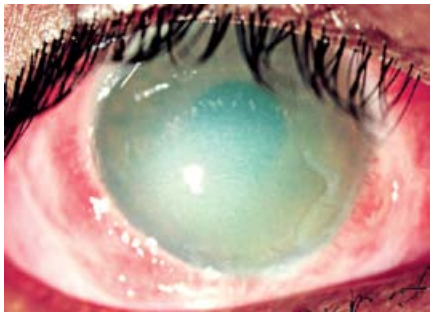


Fig. 16.21.6A: Alkali burn—intermediate with persistent epithelial defect

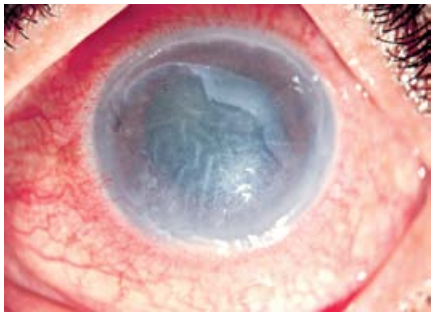


Fig. 16.21.6B: Alkali burn—intermediate with persistent epithelial defect

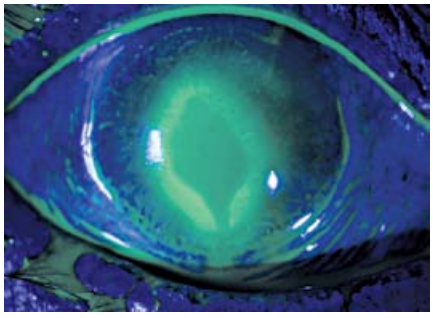


Fig. 16.21.7: Alkali burn—intermediate with persistent epithelial defect

- Corneal opacity of variable degree **(Fig. 16.21.8A)**
- Descemet's fold and iridocyclitis **(Fig. 16.21.8B)**

- *Late reparative phase* (3 weeks to several months):
  - irregular scarring of the cornea with partial limbal stem cell deficiency **(Figs 16.21.9A and B)**
  - corneal thinning and Descemetocele formation
  - Dry eye, due to scarring of the ducts of lacrimal glands and goblet cells
  - Hypotony due to ciliary shock
  - Development of symblepharon, entropion and trichiasis **(Fig. 16.21.10)**
  - Ultimately in some cases, with severe burn, there is total 360-degree limbal stem cell deficiency **(Figs 16.21.11 to 16.21.15)**

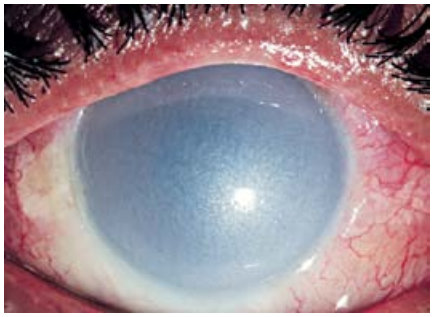


Fig. 16.21.8A: Alkali burn—stromal opacity

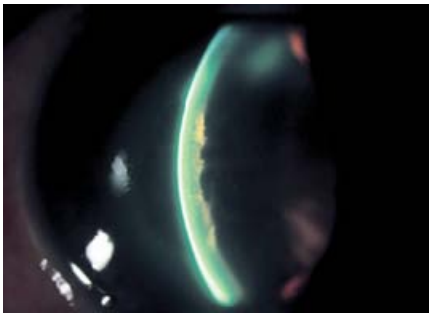


Fig. 16.21.8B: Alkali burn—Descemet's fold and iridocyclitis

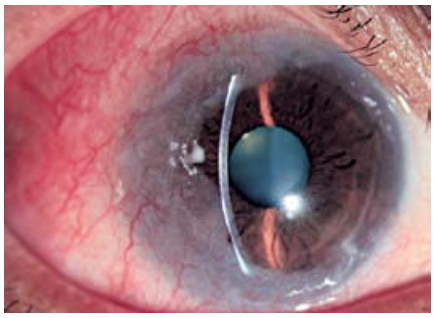


Fig. 16.21.9A: Partial limbal stem cell deficiency following chemical burn

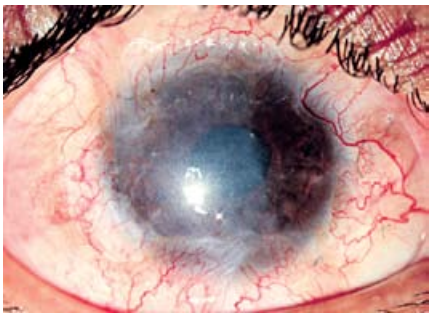


Fig. 16.21.9B: Chemical burn—corneal opacity with partial limbal stem cell deficiency

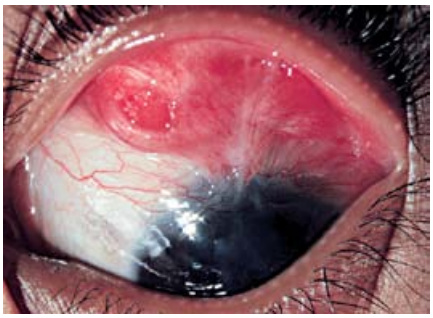


Fig. 16.21.10: Chemical burn—corneal opacity with partial limbal stem cell deficiency with symble

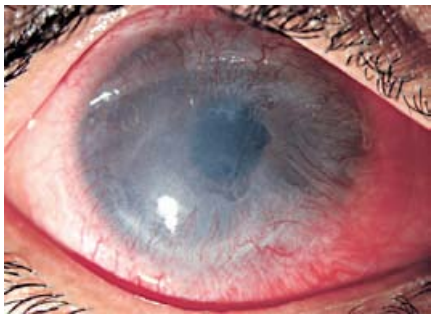


Fig. 16.21.11: Alkali burn—late phase with persistent epithelial defect

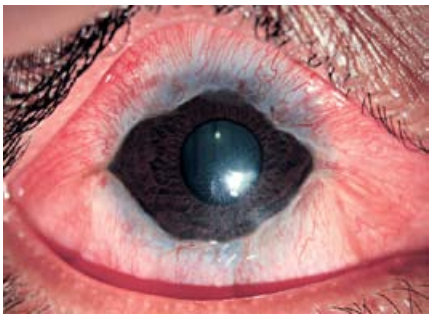


Fig. 16.21.12: Alkali burn—late phase with persistent epithelial defect

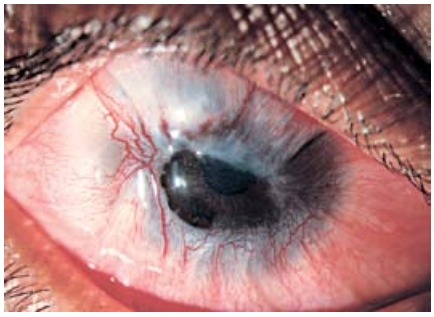


Fig. 16.21.13: Alkali burn with severe limbal stem cell deficiency

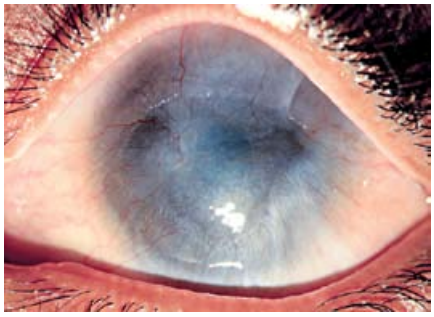


Fig. 16.21.14: Chemical burn—corneal opacity with limbal stem cell deficiency

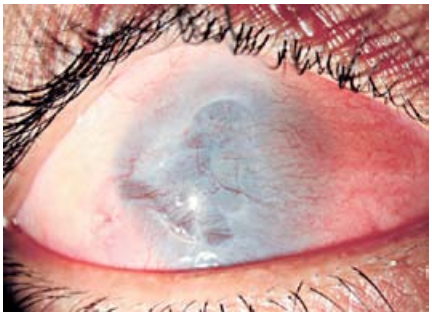


Fig. 16.21.15: Chemical burn—corneal opacity with severe limbal stem cell deficiency

- *Treatment:* Good wash, topical cortcosteroids, tab vitamin C, cycloplegics, immediate amniotic membrane transplantation **(Figs 16.21.16A and B)** and limbal stem cell transplantation in late stage.

Other Chemical Injuries

- *Latex of Calotropis procera injury:*
  - The picture is like that of mild alkali burn
  - Epithelium remains intact
  - Severe Descemet's fold with corneal edema **(Figs 16.21.17A and B)**
  - Usually subsides with topical corticosteroids.
- *Holi color injury:*
  - Usually due to green color crystals (Malachite green)
  - Corneal epithelial abrasion of various degree **(Figs 16.21.18 to 16.21.20)**
  - In severe cases, there may be LSCD
- *Formocresol injury:*

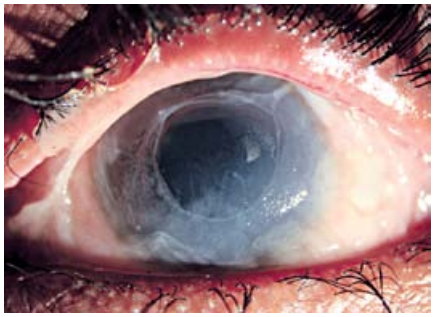


Fig. 16.21.16A: Chemical burn—acute phase—preoperative

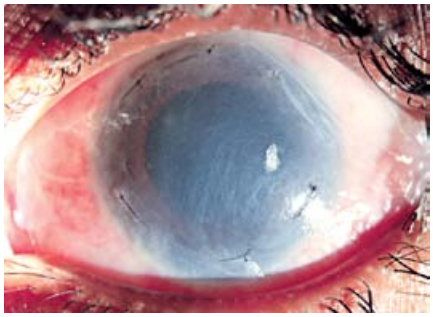


Fig. 16.21.16B: Chemical burn—acute phase—immediate amniotic membrane transplantation

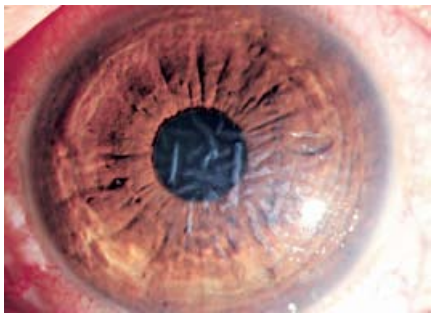


Fig. 16.21.17A: Corneal edema with DM folds after *Calotropis procera* latex injury

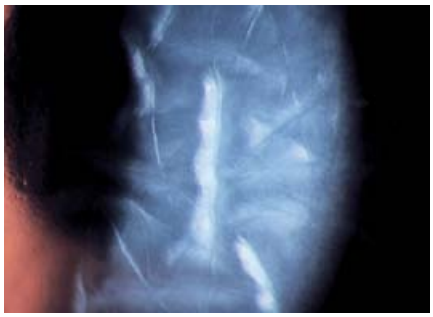


Fig. 16.21.17B: Corneal edema with DM folds after *Calotropis procera* latex injury

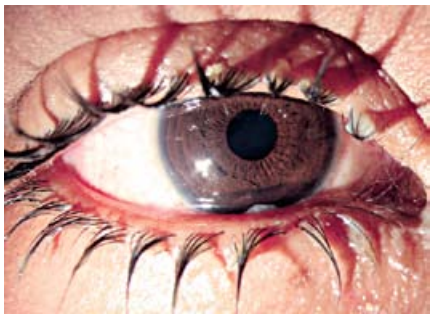


Fig. 16.21.18A: Holi color injury—RE

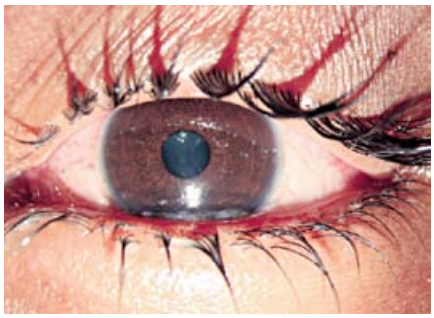


Fig. 16.21.18B: Holi color injury—LE

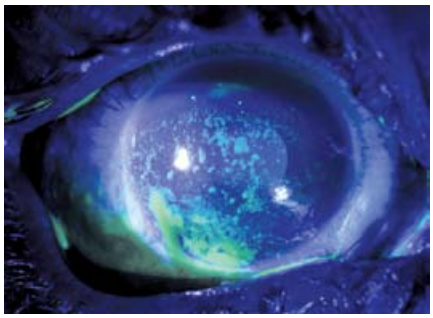


Fig. 16.21.18C: Holi color injury—RE

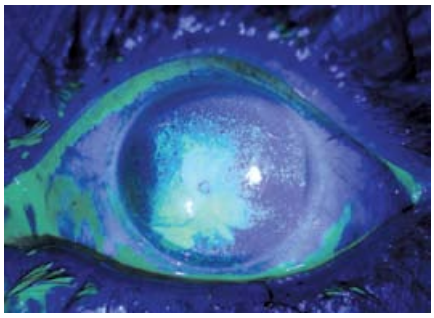


Fig. 16.21.18D: Holi color injury—LE

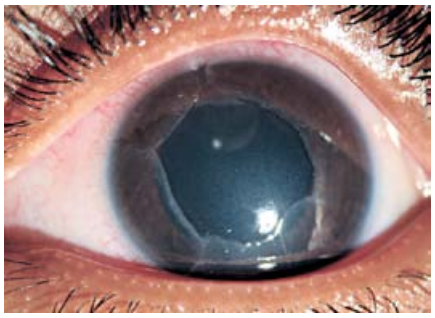


Fig. 16.21.19A: Holi color injury—large abrasion

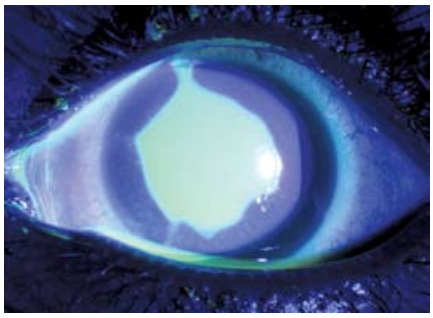


Fig. 16.21.19B: Holi color injury—large abrasion

– Formocresol solution used for dental filling is used mistakenly as eye drop
– Corneal abrasion is the main finding **(Figs 16.21.21A to C).**

Thermal Burns

- Usually do not involve the eyeball proper **(Fig. 16.22.1)**
- May be due to direct burn which may be mild **(Fig. 16.22.2)** or may be severe **(Figs 16.22.3A and B)**; by boiling hot cooking oil **(Figs 16.22.4A and B)** or by different types molten metal in factories **(Figs 16.22.5 to 16.22.6)**
- Thermal burns due to molten metal are always serious and cause limbal stem cell deficiency **(Figs 16.22.7A to C)**, symblepharon **(Fig. 16.22.8),** even ankyloblepharon formation **(Figs 16.22.9A to C)**

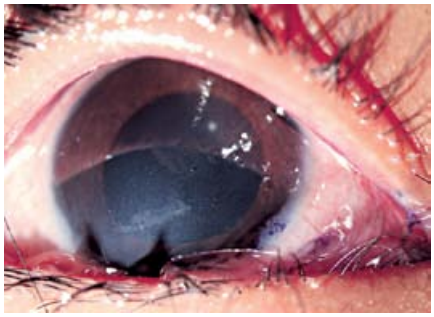


Fig. 16.21.20A: Holi color injury—large abrasion—RE

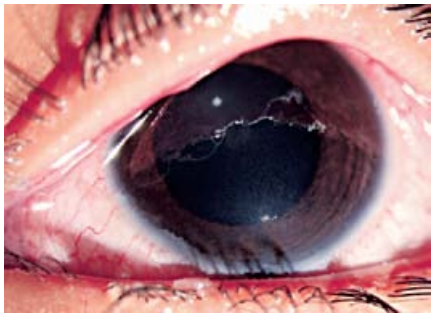


Fig. 16.21.20B: Holi color injury—large abrasion—LE

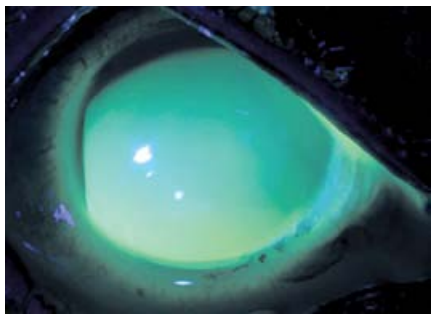


Fig. 16.21.20C: Holi color injury—large abrasion—RE

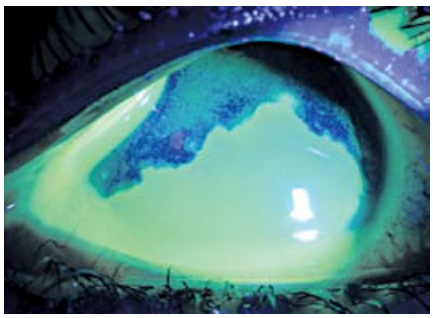


Fig. 16.21.20D: Holi color injury—large abrasion—LE

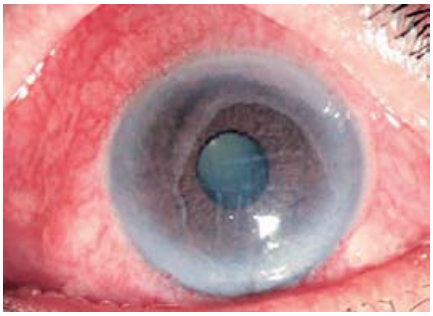


Fig. 16.21.21A: Formocresol drop injury—large abrasion

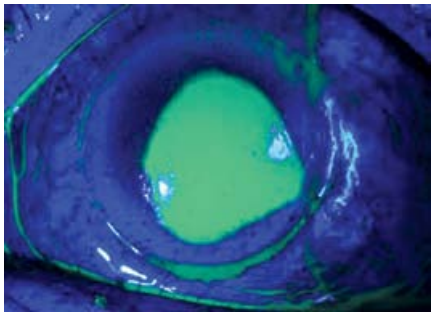


Fig. 16.21.21B: Same eye after fluorescein staining

Fig. 16.21.21C: Formocresol bottle and ciprofloxacin eye drop bottle

Fig. 16.22.1: Thermal injury—singing of eyelashes

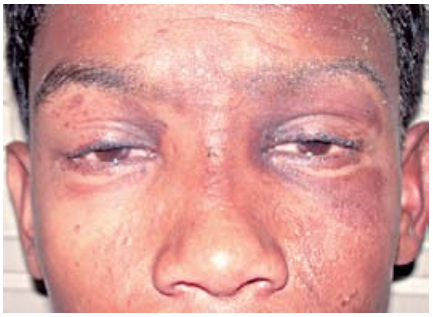


Fig. 16.22.2: Thermal burn—lids and face by gun powder

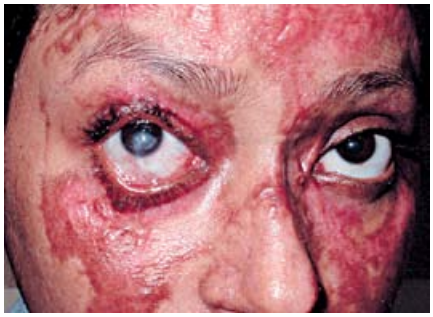


Fig. 16.22.3A: Thermal burn—facial burn including eyelids

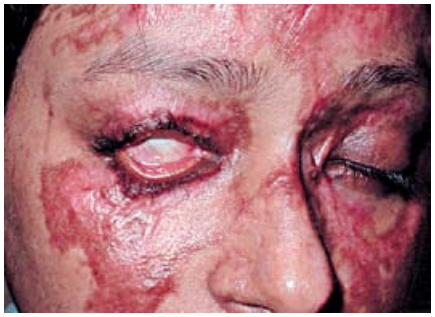


Fig. 16.22.3B: Thermal burn—facial burn including eyelids

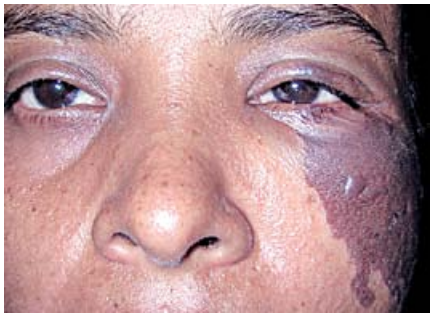


Fig. 16.22.4A: Thermal burn—by boiling hot cooking oil

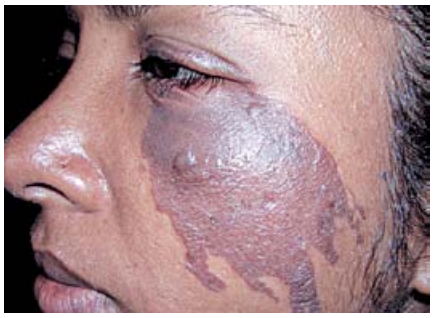


Fig. 16.22.4B: Thermal burn—by boiling hot cooking oil

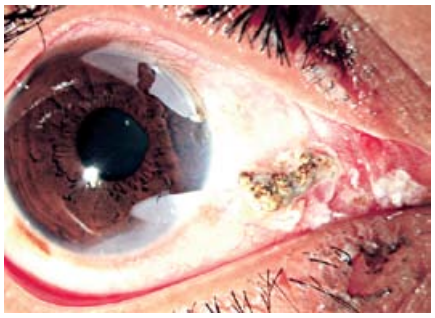


Fig. 16.22.5A: Molten Gold at fornix—conjunctival and corneal burn

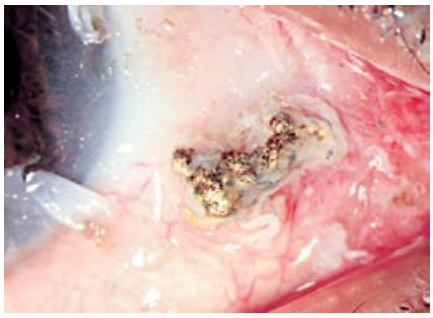


Fig. 16.22.5B: Molten Gold at fornix—conjunctival and corneal burn

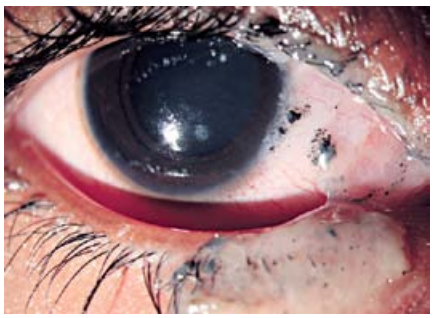


Fig. 16.22.6A: Molten metal injury—lid, conjunctiva and corneal burn

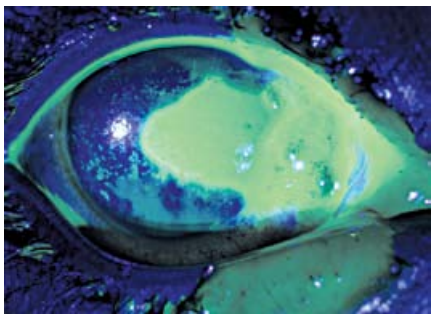


Fig. 16.22.6B: Molten metal injury—lid, conjunctiva and corneal burn

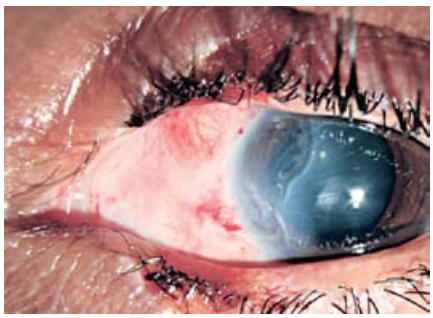


Fig. 16.22.7A: Molten metal injury

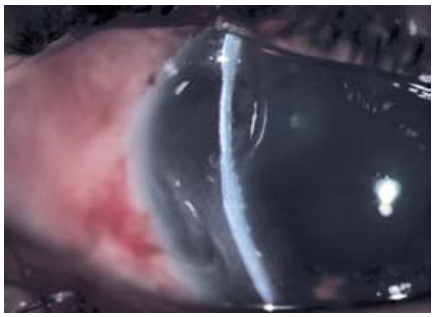


Fig. 16.22.7B: Molten metal injury—corneal burn with limbal ischemia

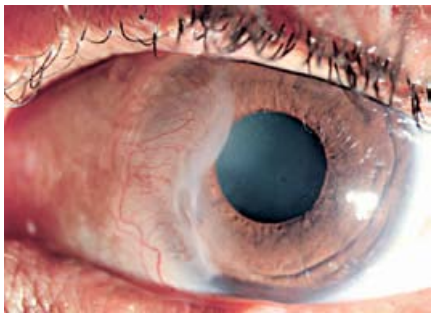


Fig. 16.22.7C: Molten metal injury—corneal burn with limbal stem cell deficiency after healing

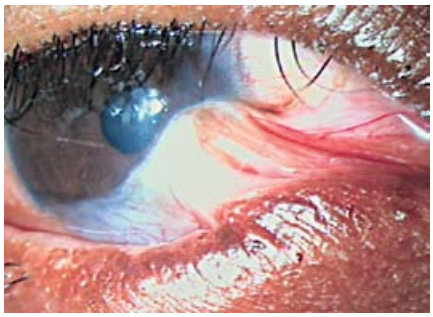


Fig. 16.22.8: Thermal burn—molten metal—keratoblepharon

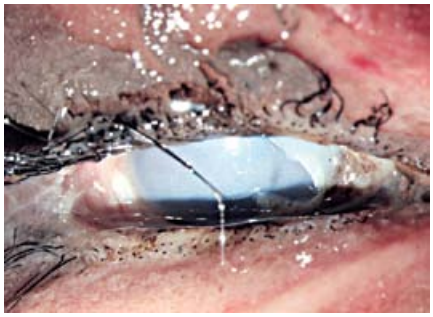


Fig. 16.22.9A: Molten metal injury—total lid and corneal burn

Fig. 16.22.9B: Molten metal injury—total lid and corneal burn

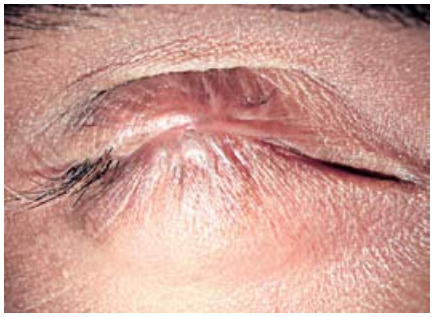


Fig. 16.22.9C: Molten metal injury—ankyloblepharon same eye

- Thermal burns of the eyelids require prompt care to prevent corneal ulcer **(Figs 16.22.10A and B)** and ectropion formation
- Severe dry eye with keratinization and formation of symblepharon **(Figs 16.22.11A and B)**
- Early skin grafting may be required in some cases
- Associated other features may be due to asphyxia and carbon monoxide poisoning.

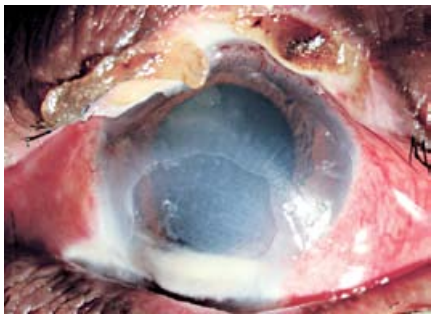


Fig. 16.22.10A: Molten metal injury—lid and corneal burn with hypopyon

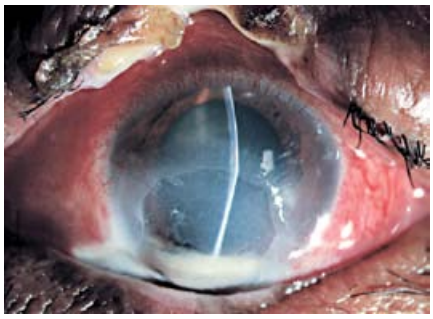


Fig. 16.22.10B: Molten metal injury—lid and corneal burn with hypopyon

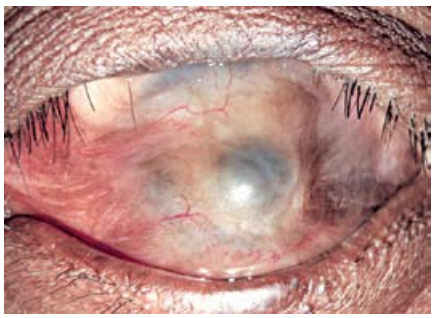


Fig. 16.22.11A: Molten metal—dry eye, keratectasia and symblepharon

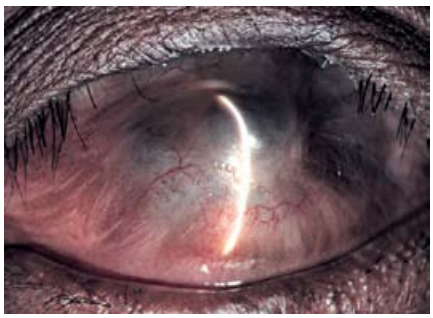


Fig. 16.22.11B: Molten metal—dry eye, keratectasia and symblepharon

Miscellaneous Injuries

Blast Injuries

- More common nowadays by fireworks, explosives or bomb blast injury
- Fire crackers injury is usually thermal and associated with facial burn or lid injuries **(Figs 16.23.1 and 16.23.2)**

Fig. 16.23.1: Thermal burn—eyelids by firework

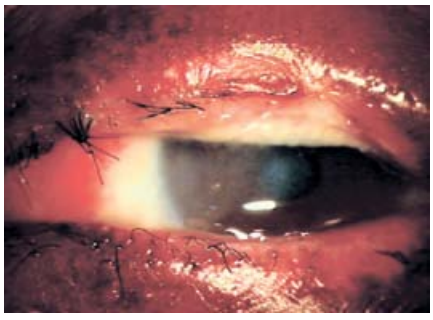


Fig. 16.23.2: Thermal burn—eyelids by firecracker injury

- May also be accidental, due to gas cylinder burst, car-tyre burst, etc. Here both thermal and concussion injuries happen with dusty particles scattered all over the face **(Figs 16.23.3 and 16.23.4)** and ocular surface **(Figs 16.23.5 and 16.23.6)**
- *Bomb blast* injuries are associated with multiple sulfur particles scattered all over the anterior surface of the eye **(Figs 16.23.7 and 16.23.8)** and in some cases they may be intracameral on the anterior surface of the iris or the lens **(Figs 16.23.9 and 16.23.10)**

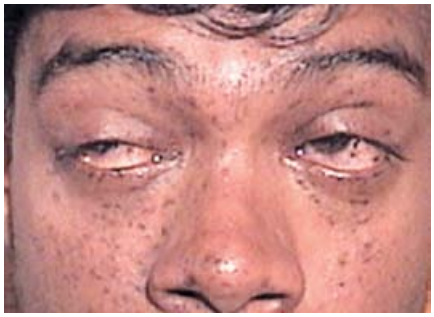


Fig. 16.23.3: Blast injury—gas cylinder

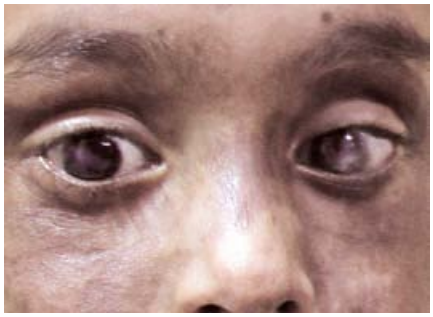


Fig. 16.23.4: Blast injury—gas cylinder—associated facial burn

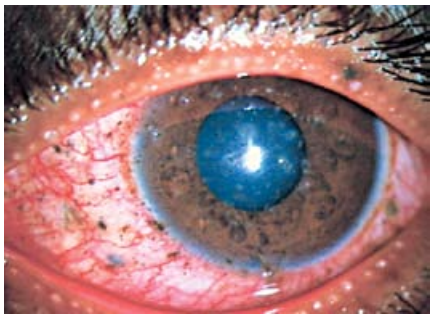


Fig. 16.23.5A: Blast injury—gas cylinder

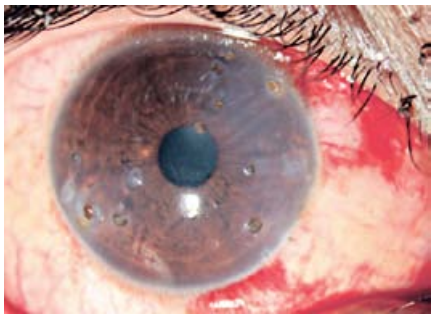


Fig. 16.23.5B: Blast injury—exploded car tyre

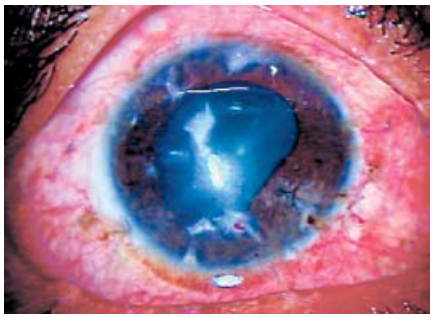


Fig. 16.23.6: Blast injury—gas cylinder—after healing

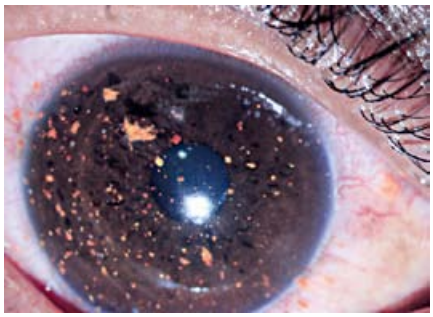


Fig. 16.23.7A: Bomb blast injury—sulphur particles—RE

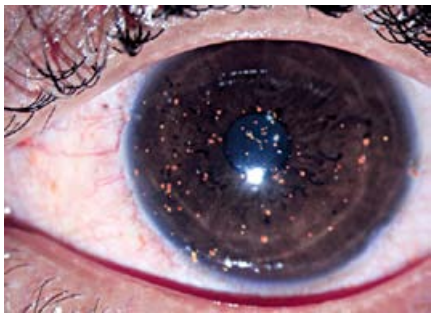


Fig. 16.23.7B: Bomb blast injury—sulphur particles—LE

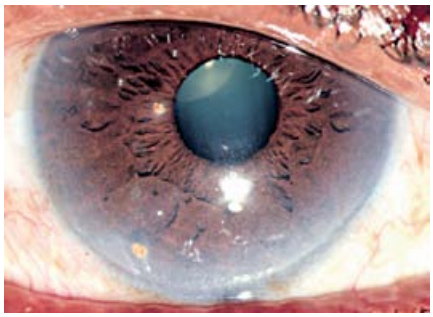


Fig. 16.23.8A: Bomb blast injury—sulphur particles—RE

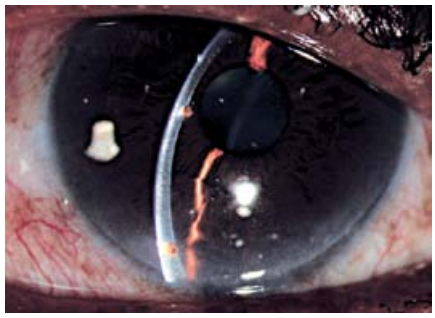


Fig. 16.23.8B: Bomb blast injury—sulphur particles—RE

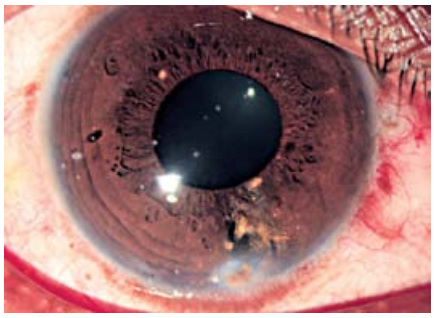


Fig. 16.23.9A: Bomb blast injury—sulphur particles on the iris surface

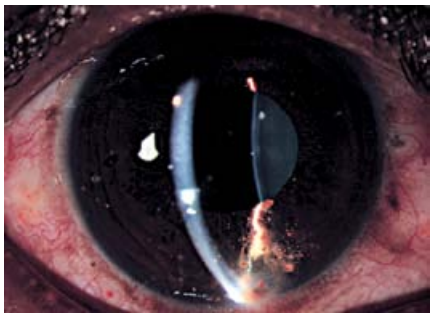


Fig. 16.23.9B: Bomb blast injury—sulphur particles on the iris surface

- Also responsible for traumatic cataract with capsular tear **(Fig. 16.23.11)**, corneal opacity with vascularization and partial or total limbal stem cell deficiency **(Figs 16.23.12 to 16.23.14)**
- Severe injury of any kind may cause open globe injury with iris prolapse **(Figs 16.23.15A and B)** or total rupture of the globe **(Fig. 16.23.16)**
- *Treatment:* Difficult, initially scraping and BCL, and later a lamellar keratoplasty may be helpful in some cases.

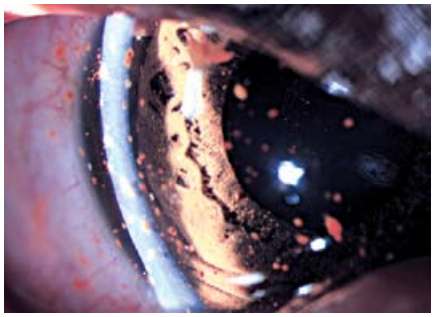


Fig. 16.23.10A: Bomb blast injury—sulfur particles on the iris surface

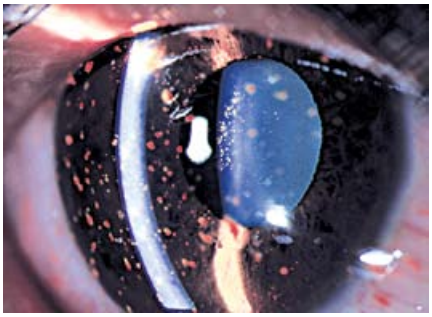


Fig. 16.23.10B: Bomb blast injury—sulfur particles over the lens surface

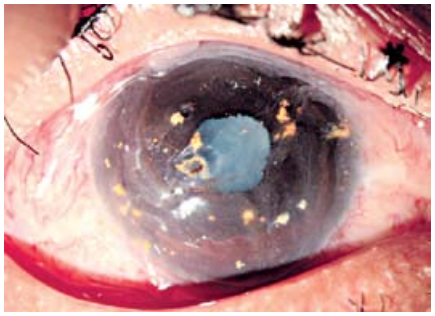


Fig. 16.23.11: Bomb blast injury—severe—intracameral sulfur particles with cataract

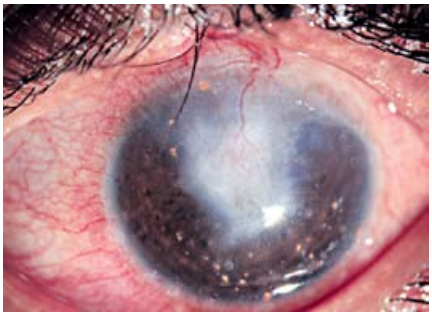


Fig. 16.23.12: Bomb injury—sulfur particles with partial LSCD

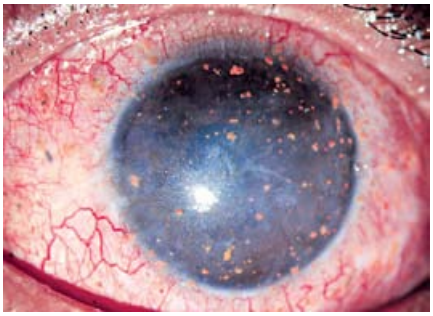


Fig. 16.23.13: Bomb blast injury—severe—360 degrees LSCD

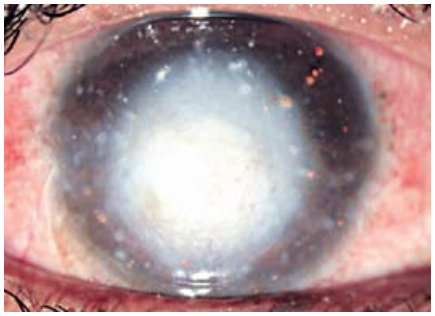


Fig. 16.23.14A: Bomb blast injury—severe—central corneal opacity with vascularization—RE

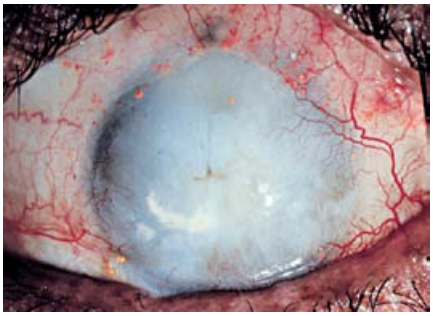


Fig. 16.23.14B: Bomb blast injury—severe—corneal opacity with vascularization—LE

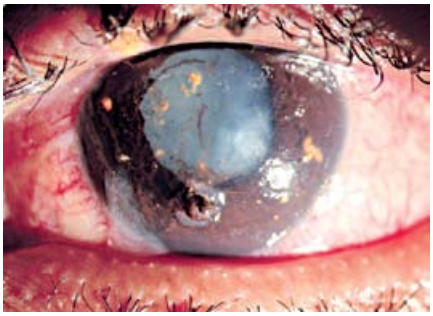


Fig. 16.23.15A: Bomb blast injury—severe-—intracameral sulfur particles with cataract and iris

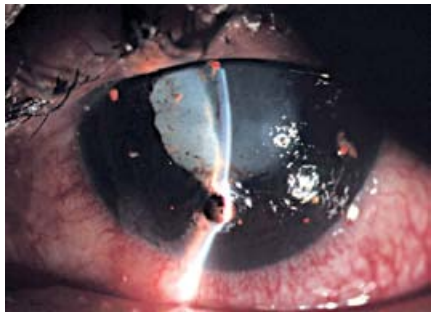


Fig. 16.23.15B: Bomb blast injury—severe—intracameral sulfur particles with cataract and iris

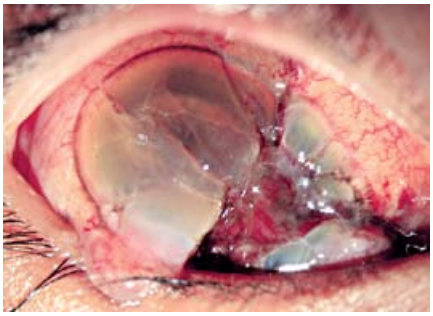


Fig. 16.23.16: Severe blast injury—transformer blast with mutilated globe

Radiational Injuries

- *Ultraviolet (UV-rays):* The cornea transmits the longer UV-rays (300–400 nm), but its epithelium absorbs the UV-rays with wavelengths below 300 nm. This results *photokeratitis* (UV keratitis) or snow blindness, or photophthalmia due to diffuse punctate epithelial erosions **(Fig. 16.24.1)**
- *Infrared rays (above 700 mm):* They are absorbed by the iris, and the resultant heat is transmitted to the lens, which becomes cataractous *(glass-blower's cataract)* **(Fig. 16.24.2)**
  - Solar eclipse with the naked eye, causes solar retinopathy
  - Causing an acute focal macular burn (eclipse burn or eclipse blindness) **(Fig. 16.24.3)**
  - May also be caused by observing sun for a long period in the morning as a part of worship of Sun God **(Figs 16.24.4A and B)**

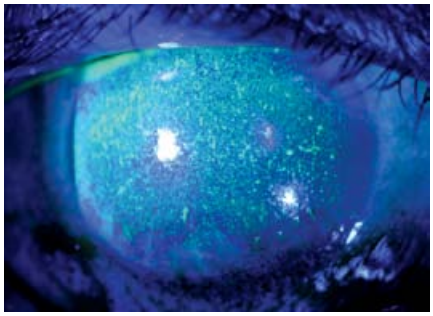


Fig. 16.24.1: Photophthalmitis after welding (UV) light exposure

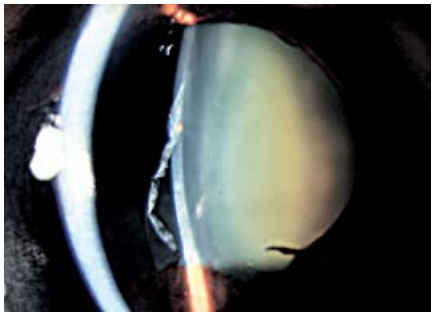


Fig. 16.24.2: Glass blower's cataract

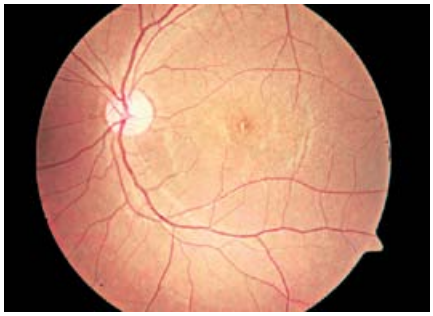


Fig. 16.24.3: Solar retinopathy—eclipse burn

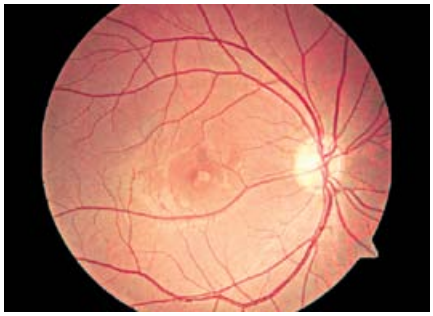


Fig. 16.24.4A: Solar maculopathy—bilateral—RE

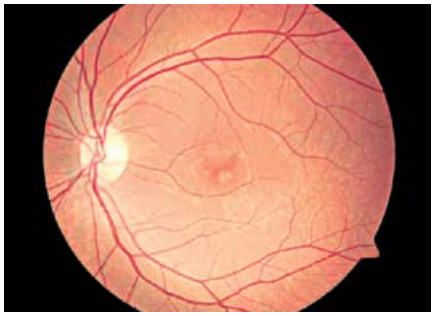


Fig. 16.24.4B: Solar maculopathy—bilateral—LE

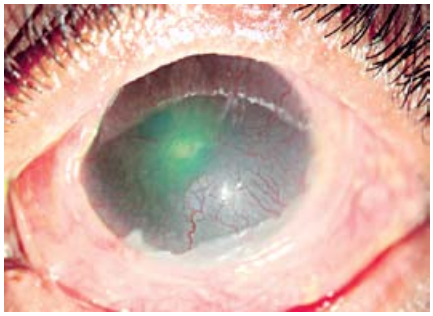


Fig. 16.24.5A: Radiation keratopathy—SJS like picture

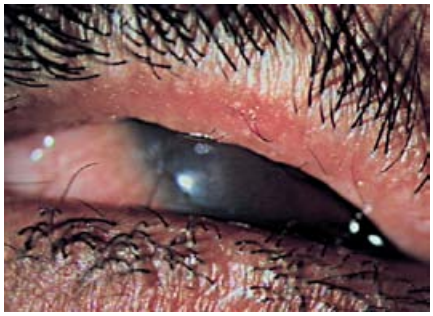


Fig. 16.24.5B: Radiation keratopathy—dry eye

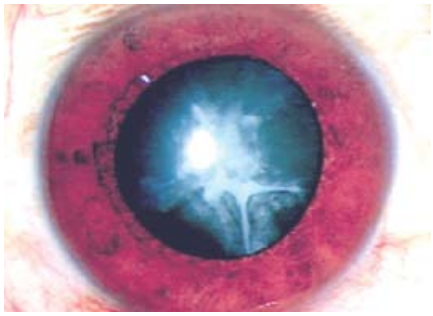


Fig. 16.24.6A: Radiation cataract—anterior capsular fibrosis

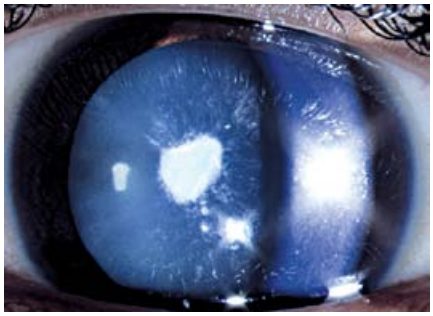


Fig. 16.24.6B: Radiation cataract with anterior capsular fibrosis

- *Electromagnetic energy* of short wavelengths (X-rays or gamma rays)
  - Any part of the eye is affected, e.g. blepharoconjunctivitis (Stevens-Johnson syndrome-like picture), keratitis **(Figs 16.24.5A and B)**, radiation cataract with anterior capsular fibrosis **(Figs 16.24.6A and B)** and radiation retinopathy
- *Electric injury or injury after elctrocaution:*
  - May cause electric cataract in some cases
  - Other organ may also be affected
  - Retinal damage in various extent may be associated with this injuries
- *Treatment:* Adequate prophylactic measures are to be taken to prevent radiational injuries. Different types of protective glasses are available, which can prevent lesions caused by ultraviolet and infrared rays
- Adequate and necessary protection is also to be taken during watching a solar eclipse.