Synopsis of
Ophthalmology

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Preface to the sixth edition

The many new developments in the field of ophthalmology since the last edition of the *Synopsis* have necessitated a complete revision of the text. The aim of the book is, however, unaltered – to provide a quick reference to the main aspects of ophthalmology in an orderly and easily reproducible manner. It is hoped that the book will be of particular value as a quick revision to those preparing for postgraduate examinations in the speciality.

Jack J. Kanski
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Chapter 1

The eyelids

CYSTIC LESIONS

External hordeolum (stye)

Cause

An acute staphylococcal infection of a lash follicle and its associated gland of Zeis or Moll.

Clinical features

This very common lesion presents at any age with an acute onset of a tender swelling which points through the skin.

Treatment

- Removal of the associated eyelash and application of an antibiotic ointment to the lid margin to prevent spread of the infection.
- Incision of a large abscess may be required.

Meibomian cyst (chalazion)

Cause

Meibomian gland dysfunction is characterized by:

- Duct obstruction and retention of sebum.
- Secondary chronic lipogranulomatous inflammation.

Clinical features

This very common lesion presents at any age with a gradual onset of a painless swelling in the tarsal plate.

Treatment

- Incision through the conjunctiva is the treatment of choice.
- Injection of triamcinolone into the cyst may also be effective.

Internal hordeolum

Cause

An acute staphylococcal infection of a meibomian gland.

Clinical features

This very common lesion presents in a similar manner to a stye but it is more painful and it may discharge anteriorly or posteriorly.

Treatment

This is similar to a stye but incision may be necessary if a hard nodule remains.

Miscellaneous

Cyst of Moll is a tiny translucent retention cyst of a modified sweat gland which contains clear fluid.

Cyst of Zeis is a retention cyst of a modified sebaceous gland which is less translucent than a cyst of Moll.

Sebaceous cyst is a cyst of an ordinary sebaceous gland which contains cheesy secretions.

BENIGN TUMOURS

Molluscum contagiosum

Cause

Virus infection.

Clinical features

Signs – presents usually in children with small pale umbilicated lid nodules.

Complications – ipsilateral chronic follicular conjunctivitis and epithelial keratitis.

Treatment

Expression or cauterization of the nodule.

Miscellaneous

Squamous papilloma may be sessile or pedunculated.

Verruca vulgaris characterized by filiform warts which may grow in crops.

Seborrhoeic keratosis which is greasy brown and friable.

Senile keratosis characterized by multiple, flat and scaly lesions which may occasionally undergo malignant change.

Xanthelasma are flat, yellow plaques.

Keratoacanthoma which starts as a red papule, then turns into a nodule with a crater and regresses within weeks. It may mimic a squamous cell carcinoma.

MALIGNANT TUMOURS

Basal cell carcinoma

Clinical features

This is the most common malignant eyelid tumour. It is locally invasive but does not metastasize. The tumour typically presents in old age in one of the following two ways.

Noduloulcerative (rodent ulcer) is the most common type. It consists of a well-defined nodule with telangiectatic blood vessels, central ulceration and a 'pearly' appearance due to associated hyperkeratosis.

Sclerosing (morphea type) is less common. The tumour may be multifocal and because it grows radially it has a flat, ill-defined, scar-like appearance.

Treatment

- *Local excision* is the treatment of choice.
- *Radiotherapy* for tumours unsuitable for exision but it should be avoided for the medial canthus because deep infiltration is common.
- *Cryotherapy* for small superficial tumours.
- *Mohs' micrographic technique (chemosurgery)* for diffusely growing tumours in which the lesion is excised in layers and examined by frozen section.
- *Exenteration* in the rare event of orbital invasion.

Squamous cell carcinoma

Clinical features

This very rare tumour presents in late adult life as an ulcer (most common), a nodule, a 'papilloma' or a cutaneous horn. The tumour grows faster than a rodent ulcer and it may metastasize.

Treatment

Wide excision.

Sebaceous gland carcinoma

Clinical features

This very rare tumour may be multifocal and it may mimic 'recurrent chalazion' or 'severe chronic blepharitis', hence the frequent delay in diagnosis and poor prognosis.

Treatment

Wide excision.

DISORDERS OF EYELASHES

Trichiasis

Definition

An inward misdirection of normal eyelashes.

Causes

- Primary (rare).
- Secondary to entropion (common).

Treatment

- *Epilation* is easy but recurrence occurs within 4–6 weeks.
- *Electrolysis* is tedious and recurrences are common.
- *Cryotherapy* is effective but it may cause skin depigmentation in Black patients.
- *Argon laser destruction* is effective but may need to be repeated.
- *Protective contact lenses* as a temporary measure.

Distichiasis

Definition

An extra row of lashes arising from meibomian gland orifices.

Causes

- Congenital and familial (very rare).
- Secondary to cicatrizing conjunctivitis (see Chapter 4).

Treatment

- Cryotherapy for the lower eyelid.
- Lid splitting and cryotherapy for the upper lid.

ENTROPION

Classification

- Involutional
- Cicatricial
- Congenital
- Acute spastic

Involutional entropion

Cause

A combination of the following age-related changes:

- Preseptal orbicularis overrides pretarsal orbicularis (factor a).
- Horizontal lid laxity (factor b).
- Weakness of lower lid retractors (factor c).

Treatment

- *Cautery* through the skin – corrects factor a.
- *Transverse lid eversion suture* – corrects factor a.
- *Wies procedure* (horizontal lid splitting and marginal rotation) – corrects factors a and c.
- *Horizontal lid shortening* – corrects factor b.
- *Inferior aponeurosis tucking* – corrects factors a and c.
- *Fox's procedure* (excision of base-down triangle of tarsus).

Cicatricial entropion

Cause

Scarring of the palpebral conjunctiva by:

- Chemical burns.
- Cicatrizing conjunctivitis (see Chapter 4).

Treatment

- Weis procedure in mild cases.
- Lamellar grafts in severe cases.

Congenital entropion

Cause

Hypertrophy of the skin and orbicularis muscle.

Treatment

Excision of an ellipse of skin and orbicularis.

Acute spastic entropion

Cause

Spasm of orbicularis muscle due to chronic ocular irritation.

Treatment

- Elimination of the cause if possible.
- Transverse lid-everting suture.

ECTROPION

Classification

- Involutional
- Cicatricial
- Congenital
- Paralytic

Involutional ectropion

Cause

A combination of the following age-related changes:

- Excessive horizontal lid length.

- Weakness of the pretarsal orbicularis muscle.
- Weakness of the medial and lateral canthal tendons.

Treatment

- *Ziegler cautery* 5 mm below the punctum for mild medial ectropion associated with punctal eversion.
- *Medial conjunctivoplasty* (excision of diamond-shaped block) for mild medial ectropion.
- *Bick's* procedure (excision of lateral wedge of eyelid) for mild to moderate cases.
- *Fox's* procedure (excision of base-up triangle of tarsus) for mild to moderate cases.
- *Kuhnt–Szymanowski* procedure (excision of pentagon of tarsus and triangle of lateral skin) for severe ectropion.

Cicatricial ectropion

Cause

Scarring of the eyelid.

Treatment

Excision of the scar.

Congenital ectropion

Cause

Shortage of skin which may be associated with the blepharophimosis syndrome (see under Ptosis).

Treatment

Skin grafts.

Paralytic ectropion

Cause

Facial nerve palsy.

Treatment

Of temporary cases

- Lubricants to prevent corneal drying.
- Strapping of the eyelids to prevent exposure keratopathy.

Of permanent cases

- Tarsorrhaphy.
- Canthoplasty.
- Silicone slings.

PTOSIS

Classification

- Neurogenic
- Aponeurotic
- Mechanical
- Myogenic

Neurogenic ptosis

Causes

Third nerve palsy – see Chapter 15.

Oculosympathetic palsy – see Chapter 15.

Synkinetic ptosis of two types:

- Marcus Gunn jaw-winking phenomenon in which jaw movements induce retraction of the ptotic eyelid.
- Misdirection of the third nerve in which eye movements induce bizarre movements of the upper eyelid.

Aponeurotic ptosis

Causes

- *Involutional* (senile) due to degeneration of the levator aponeurosis.

- *Postoperative* following cataract or retinal detachment surgery.
- *Blepharochalasis* due to thinning of the levator aponeurosis following recurrent episodes of idiopathic lid oedema.

Mechanical ptosis

Causes

Excess weight on eyelid by:

- Lid oedema.
- Tumours (e.g. neurofibroma).
- Dermatochalasis (excessive skin).
- Conjunctival scarring.

Myogenic ptosis

Causes

Congenital

Simple is the most common and is usually unilateral. It is due to a dystrophy of the levator muscle which causes both poor contraction and incomplete relaxation. It may be associated with weakness of the superior rectus muscle

Blepharophimosis syndrome is very rare and dominantly inherited. It is characterized by a wide intercanthal distance, epicanthus inversus and congenital ectropion.

Acquired

- Myasthenia gravis.
- Dystrophia myotonica.
- Ocular myopathy (see Chapter 15).

Operations for ptosis

Levator resection for congenital myogenic ptosis with some levator function.

Frontalis (brow) suspension if levator function is absent.

Tarsoconjunctival resection (Fasanella–Servat procedure) is useful for mild ptosis of 2 mm or less.

Aponeurosis strengthening.

BLEPHARITIS

Staphylococcal blepharitis

Cause

Chronic staphylococcal infection of bases of the lashes which is particularly common in patients with dry eyes and atopic eczema.

Clinical features

Presentation of this very common condition is with mild bilateral chronic ocular irritation.

Signs

- Anterior lid margin shows dilated blood vessels (rosettes).
- Hard scales around the base of the lashes (collarettes).
- Lashes show trichiasis, are few in number (madarosis) and white (poliosis).

Complications

- Recurrent styes (very common).
- Recurrent conjunctivitis (common).
- Mild inferior punctate epithelial erosions (PEEs) (common).
- Marginal keratitis (common).
- Inferior pannus (rare).

Seborrhoeic blepharitis

Cause

Abnormal glands of Zeis secrete excessive neutral lipids which are split by *Corynebacterium acnes* into irritating free fatty acids. It is particularly common in patients with seborrhoeic dermatitis.

Clinical features

Presentation is similar to staphylococcal blepharitis but the symptoms are usually less severe.

Signs

- Anterior lid margin is waxy due to excess lipids.
- Soft scales occur anywhere along the lid margin.
- Lashes are greasy and stuck together.

Complications

- Papillary conjunctivitis.
- Mid-zone PEEs.

Posterior blepharitis (meibomian gland dysfunction)

Cause

An abnormality of meibomian glands. It is particularly common in patients with acne rosacea and seborrhoeic dermatitis.

Clinical features

Presentation is usually in adults with bilateral ocular stinging and transient blurring of vision due to excess oil in the tear film.

Signs

- Oil globules are present at the meibomian gland orifices.
- Tarsal plates are distorted and contain dilated meibomian glands with solidified sebum.
- Foam in the tears (meibomian seborrhoea) is a characteristic feature.

Complications

- Recurrent meibomian cysts.
- Mild conjunctivitis.
- Mild PEEs.
- Tear film instability with a reduced tear film break-up time.

Treatment of blepharitis

Lid hygiene consisting of removal of crusts and excess lipids with a 50% solution of a baby shampoo.

Topical antibiotics (bacitracin or gentamicin) for staphylococcal blepharitis.

Systemic antibiotics (tetracycline or erythromycin) for severe cases particularly if associated with seborrhoea or acne rosacea.

Topical steroids as short term for secondary complications.

Artificial tears if the tear film break-up time is reduced.

Chapter 2

The orbit

THYROID OPHTHALMOPATHY

Classification

Eyelid retraction

Infiltrative ophthalmopathy

- Soft tissue involvement
- Proptosis
- Optic neuropathy
- Restrictive myopathy

Eyelid retraction

Cause

A combination of:

- Overaction of Müller's muscle due to sympathetic overstimulation.
- Overaction of the 'levator–superior rectus complex' secondary to restrictive myopathy of the inferior rectus muscle.
- Restrictive myopathy of the levator muscle.

Clinical features

- Lid retraction (Dalrymple's sign).
- Lid lag (von Graefe's sign).

- Staring appearance (Kocher's sign).
- Fine tremor on lid closure.
- Jerky movements on lid opening.
- Infrequent blinking.

Infiltrative ophthalmopathy

Cause

A combination of:

- Increase in mucopolysaccharides in extraocular muscles leading to enlargement, round cell infiltration, fibrosis and restrictive myopathy.
- Proliferation and round cell infiltration of orbital fat leading to fluid retention, increase in intraorbital pressure, proptosis and optic neuropathy.

Clinical features

Soft tissue involvement

- Lid oedema (common).
- Conjunctival injection and chemosis (common).
- Superior limbic keratoconjunctivitis (uncommon).

Proptosis is common and it may be unilateral, bilateral and asymmetrical. It is usually self-limiting and is not influenced by treatment of the hyperthyroidism.

Optic neuropathy is rare and it may occur without significant proptosis.

Restrictive myopathy is uncommon. The order of frequency of muscle involvement is:

1. Inferior rectus leading to defective elevation.
2. Medial rectus leading to defective abduction.
3. Superior rectus leading to defective depression.
4. Lateral rectus leading to defective adduction.

Treatment of thyroid ophthalmopathy

Non-specific

- *Head elevation* at night to reduce periorbital oedema.
- *Taping of eyelids* at night for exposure keratopathy.
- *Prismatic spectacles* for mild diplopia.

Topical

- *Lubricants* for ocular irritation.
- *Guanethidine 5% drops* for lid retraction (rarely used).

Systemic

- *Diuretics* for periorbital oedema (usually disappointing).
- *Steroids* for optic neuropathy, severe proptosis, chemosis and pain.
- *Cytotoxic agents* are of doubtful benefit.

Radiotherapy may help in steroid-resistant cases.

Surgery

- *Orbital decompression* for severe exposure keratopathy, optic neuropathy and cosmetically unacceptable proptosis.
- *Squint surgery* for diplopia, provided the deviation is stable for at least 6 months.
- *Tarsorrhaphy* for severe exposure keratopathy.
- *Levator weakening* for severe lid retraction.
- *Blepharoplasty* to reduce excessive periorbital fat and skin.

ORBITAL INFECTIONS

Preseptal cellulitis

Cause

Usually follows a severe lid infection, an insect bite or a skin laceration.

Clinical features

This *common* condition presents with an acute onset of unilateral periorbital swelling, erythema and tenderness.

Treatment

Systemic antibiotics (as out-patient).

Orbital cellulitis

Cause

This is caused by the spread of infection from the sinuses (usually ethmoids) or the nasopharynx.

Clinical features

Presentation of this *rare* condition is typically in children or young adults with acute unilateral lid oedema, chemosis, ophthalmoplegia and proptosis.

Complications

- Meningitis and brain abscess.
- Cavernous sinus thrombosis.
- Central retinal artery occlusion.

Treatment

Systemic antibiotics (as in-patient).

Indications for surgery:

- Resistance to antibiotics.
- Decreasing visual acuity.
- Subperiosteal or orbital abscess.

Mucormycosis

Cause

This is an opportunistic infection by fungi of the order of Mucorales which spreads from the palate, nose or sinuses. It typically affects severely debilitated patients.

Clinical features

Presentation of this *very rare* condition is with a subacute onset of unilateral chemosis, ophthalmoplegia, proptosis, visual loss and

facial pain. A black eschar is present due to ischaemic infarction and septic necrosis of the palate, turbinate, nasal septum and skin.

Complications include CNS involvement which has a high mortality.

Treatment

- Intravenous amphotericin B.
- Wide excision of necrotic tissue.

INFLAMMATORY ORBITAL DISEASE (PSEUDOTUMOUR)

Definition

A *rare*, idiopathic, space-occupying, periocular inflammatory lesion that may involve all or any of the soft tissue components of the orbit.

Classification

Typical

Variants

- Bilateral
- Orbital myositis
- Tolosa–Hunt syndrome

Typical pseudotumour

Clinical features

Presentation is during middle age with usually a subacute onset of pain, lid oedema, chemosis, ophthalmoplegia and proptosis.

Clinical course may be short and benign, intermittent, or severe and prolonged. In severe cases the end-result is a 'frozen orbit'.

Differential diagnosis

In bilateral cases the following should be considered:

- Thyroid ophthalmopathy.
- Lymphoma.
- Polyarteritis nodosa.
- Wegener's granulomatosis.
- Waldenström's macroglobulinaemia.

Treatment

- None in mild cases.
- Systemic steroids are effective in 75% of severe cases.
- Local radiotherapy may be effective in steroid-resistant cases.
- Cytotoxic agents may help in radiotherapy-resistant cases.

Orbital myositis

Clinical features

Acute myositis is characterized by an acute onset of pain on eye movement and a short course.

Chronic myositis causes a large muscle which may be mistaken for thyroid myopathy.

Treatment

The response to systemic steroids is good in acute myositis and poor in chronic myositis.

Tolosa–Hunt syndrome

Clinical features

Subacute onset of unilateral painful ophthalmoplegia, motor pupillary involvement, and sensory loss of the first and second divisions of the fifth nerve.

Treatment

Systemic steroids are very effective.

ORBITAL TUMOURS

Classification

Vascular

- Varix
- Capillary haemangioma
- Cavernous haemangioma
- Lymphangioma

Lacrimal gland

- Epithelial (mixed cell tumour, carcinoma)
- Non-epithelial (pseudotumour, lymphoma)

Lymphoproliferative

Rhabdomyosarcoma

Histiocytosis X

- Letterer–Siwe disease
- Hand–Schüller–Christian disease
- Eosinophilic granuloma

Cystic lesions

- Dermoid cyst
- Blood cyst
- Mucocele

Neural

- Optic nerve glioma
- Optic nerve sheath meningioma

Metastases

Orbital varices

Clinical features

This *uncommon* condition usually presents in childhood with intermittent non-pulsatile painless proptosis without a bruit. The Valsalva manoeuvre increases the amount of proptosis.

Treatment

Usually unnecessary.

Capillary haemangioma

Clinical features

This *rare* tumour presents in infancy with a unilateral anterior orbital swelling which increases in size when crying. 'Strawberry' naevi on the eyelids are a frequent associated finding. The haemangioma enlarges during the first year of life, then stabilizes and usually disappears by the age of 5 years.

Treatment

Indications

- Amblyopia.
- Optic nerve compression.
- Exposure keratopathy.

Methods

- Systemic steroids.
- Steroid injection into the lesion.
- Radiotherapy.
- Excision with a cutting cautery.

Cavernous haemangioma

Clinical features

This is the *most common benign orbital tumour in adults.* It presents in young adults with a gradual unilateral painless axial proptosis.

Treatment

Excision is easy because the tumour is well encapsulated.

Lymphangioma

Clinical features

This *very rare* tumour presents in childhood, usually with a gradual unilateral painless proptosis. Some cases resolve spontaneously.

Treatment

Usually unnecessary.

Mixed cell lacrimal gland tumour

Clinical features

This *rare* tumour presents during the fourth to fifth decade with a unilateral, painless, slow growing, smooth, firm and non-tender swelling in the lacrimal fossa. X-rays may show indentation without bony destruction.

Treatment

Excision through a lateral orbitotomy; biopsy is contraindicated.

Lacrimal gland carcinoma

Clinical features

This *very rare* tumour presents in adults with a unilateral, painful, fast growing swelling in the lacrimal fossa. X-rays in early cases may be normal and in late cases show bony destruction.

Treatment

Biopsy followed by exenteration with or without radiotherapy. The prognosis is very poor.

Lymphoproliferative disorders

Clinical features

This *uncommon* condition presents in the fifth to sixth decade with involvement of any part of the orbit or lacrimal gland. Occasionally both orbits are affected.

Treatment

Radiotherapy.

Rhabdomyosarcoma

Clinical features

This *very rare* tumour is the most common primary orbital malignancy in children. It presents around the age of 7 with a unilateral subacute onset of painful progressive proptosis and a swelling most commonly in the superonasal quadrant. It may also present with ptosis or a lid mass.

Treatment

Biopsy followed by chemotherapy and radiotherapy. The survival rate is 65–90%

Histiocytosis X

Definition

Very rare childhood disorder characterized by histiocytic proliferation and granuloma formation.

Main types

Letterer–Siwe disease rarely involves the orbit.

Hand–Schüller–Christian disease is characterized by proptosis, diabetes insipidus and bony skull defects.

Eosinophilic granuloma frequently involves the orbit.

Treatment

- Evacuation of single lesions.
- Radiotherapy for multiple lesions.
- Chemotherapy and systemic steroids for disseminated disease.

Simple dermoid cyst

Clinical features

This *uncommon* lesion presents in infancy with a unilateral, firm, painless, smooth swelling in the upper temporal or upper nasal quadrant of the anterior orbit. The cyst is *not* associated with bony defects.

Treatment

Excision.

Complicated dermoid cyst

Clinical features

This *rare* lesion presents in late childhood or adult life with gradual unilateral painless proptosis or an indistinct anterior orbital mass. The cyst *may be* associated with an underlying bony defect.

Treatment

Excision.

Blood cysts

Causes

- Blunt trauma.
- Lymphangioma.
- Cavernous haemangioma.
- Blood dyscrasias.

Clinical features

This uncommon condition presents with progressive unilateral proptosis.

Treatment

Aspiration.

Mucocele

Cause

Orbital extension of an ethmoidal or a frontal mucocele.

Clinical features

This *uncommon* condition presents usually in middle age with a gradual, unilateral, slowly progressive, painless proptosis with displacement of the globe either laterally or inferolaterally.

Treatment

Excision.

Orbital metastases

In children

- Neuroblastoma presents with frequently bilateral, acute painful proptosis and lid ecchymosis.
- Ewing's tumour presents with an acute unilateral haemorrhagic proptosis.
- Wilms' tumour.

In adults

- Bronchus.
- Breast.
- Prostate.
- Kidney.
- Gastrointestinal tract.

Neural tumours

See Chapter 15.

BLOW-OUT FRACTURE OF ORBITAL FLOOR

Mechanism

An object larger than 5 cm in diameter strikes the orbit and the increased intraorbital pressure fractures the orbital floor.

Clinical features

- Periocular ecchymosis and oedema.
- Enophthalmos may be present initially or it may develop after 10–14 days as the oedema subsides.
- Infraorbital nerve anaesthesia involving the lower eyelid, cheek, side of nose, upper lip and upper teeth.
- Diplopia is typically vertical in both up- and down-gaze (double diplopia). It is due to tethering of muscles in the fracture line.
- Positive forced duction test.
- Ocular damage is relatively rare.

Special investigations

- Plain X-rays (Waters' view).
- CT scanning (axial and coronal sections).

Treatment

- *Small cracks* without diplopia require no treatment.
- Fractures of *less than 50%* of the floor associated with *improving diplopia* require no treatment unless enophthalmos is more than 2 mm.
- Fractures of *over 50%* of the floor associated with *persistent* diplopia should be repaired within 2 weeks by freeing the entrapped tissue and covering the defect with a plastic plate.

Chapter 3

The lacrimal system

KERATOCONJUNCTIVITIS SICCA

Causes

Atrophy

- *Pure keratoconjunctivitis sicca* (KCS) in which only the lacrimal gland is damaged by infiltration with mononuclear cells.
- *Primary Sjögren's syndrome* (sicca complex) consisting of KCS and a dry mouth (xerostomia).
- *Secondary Sjögren's syndrome* consisting of the sicca complex and a connective tissue disease (usually rheumatoid arthritis).

Blockage of excretory ductules

This is usually caused by severe conjunctival scarring (see Chapter 4).

Destruction

- Granuloma.
- Tumour.
- Chronic inflammation.

Miscellaneous

- Surgical excision.
- Meibomian gland dysfunction.
- Neurogenic lesions.

Clinical features

Symptoms

Common

- Bilateral chronic gritty irritation.
- Foreign body sensation.
- Stringy mucous discharge.

Less common

- Burning.
- Itching.
- Heavy or tired eyelids.
- Photophobia.
- Resistant 'conjunctivitis'.

Signs

Precorneal tear film

- Contains excess mucus and debris.
- Marginal tear strip is thin or absent.

Cornea

- Inferior punctate epithelial erosions.
- Filaments.
- Mucous plaques.
- Dellen and thinning (rare).

Special tests

- *Tear film break-up time* (BUT) is less than 10 seconds (normal is over 10 seconds).
- *Rose bengal* stains the interpalpebral conjunctiva and filaments.
- *Schirmer's test* is less than 5 mm in 5 min (normal is 10–25 mm, borderline is 10–5 mm).

Treatment

Tear conservation

- Air humidifiers.

- Protective spectacles.
- Lateral tarsorrhaphy to reduce surface area for evaporation.

Topical therapy

- Tear substitutes (cellulose, polyvinyl alcohol and mucomimetics).
- Petrolatum mineral oil ointment.
- Acetylcysteine 5% four times a day for excess mucus.
- Hydroxypropylcellulose slow-release rods.
- Sodium hyaluronate.
- Gel tears.

Reduction of tear drainage

- *Temporary* with plugs or rods.
- *Permanent* with cautery to the proximal canaliculi in severe cases.

OBSTRUCTION OF LACRIMAL PASSAGES

Causes

- Obstruction of lacrimal drainage.
- Lacrimal pump failure.

Special investigations

- *Intubation dacryocystography* – contrast medium is injected into the canaliculi.
- *Radionucleotide testing* (scintillography) – tears are labelled with a γ-emitting substance.
- *Dye testing.*

Treatment

Punctal stenosis

- *One-snip ampullotomy* – vertical 2 mm snip in the posterior wall.
- *Two-snip* – vertical and a horizontal snip.
- *Three-snip* – excision of the posterior wall of the ampulla and a small portion of the canaliculus.
- *Argon laser punctoplasty* is useful for punctal overgrowth by conjunctiva.

Individual canalicular obstruction

- *At least 8 mm patent* – canaliculodacryocystorhinostomy (CDCR) and intubation.
- *Less than 8 mm patent* – Lester Jones tube.

Common canalicular obstruction

- *Medial end obstruction* – dacryocystorhinostomy (DCR) and intubation.
- *Lateral end obstruction* – CDCR or Lester Jones tube.

Nasolacrimal duct obstruction

- *Acquired* – DCR.
- *Congenital* – initially with hydrostatic massage over the duct (10 strokes four times a day) and by probing if the block is still present by the age of 12 months.

Chapter 4

The conjunctiva

EVALUATION OF CONJUNCTIVAL INFLAMMATION

Clinical features

Discharge

Watery

- Viral infections.
- Allergic disorders.
- Toxic reaction.

Mucoid

- Vernal disease.
- Keratoconjunctivitis sicca.

Purulent – severe bacterial infection.

Mucopurulent

- Mild bacterial infection.
- Chlamydial infection.

Reaction

Follicles

- Viral infections.

- Chlamydial infections.
- Toxic reaction.

Papillae

- Allergy disorders.
- Non-specific reaction.

Pseudomembranes

- Severe adenoviral infections.
- Gonococcal infection.
- Severe vernal disease.

Membranes – diphtheria.

Subconjunctival haemorrhages – viral infections.

Preauricular lymphadenopathy

- Viral infections.
- Chlamydial infections.
- Parinaud's syndrome.

Laboratory investigations

Scrapings

Giemsa stain identifies the cytology of inflammatory cells:

- Neutrophils = bacteria.
- Mononuclears = viruses.
- Neutrophils + mononuclears = *Chlamydia*.
- Halberstaedter–Prowazek basophilic inclusions = *Chlamydia*.
- Eosinophils = allergy.

Gram stain differentiates Gram-positive from Gram-negative bacteria.

Cultures – see Table 1.

Table 1

Media	*Bacteria*	*Fungi*
Blood agar	++ (37°C)	++ (room temp.)
Chocolate agar	*Neisseria, Haemophilus* spp.	–
Thyoglycolate broth	Facultative anaerobes	
Sabouraud's medium	–	++
Brain–heart infusion	++ (37°C)	++

BACTERIAL CONJUNCTIVITIS

Simple bacterial conjunctivitis

Causative bacteria

Most frequent are *Staphylococcus epidermidis, Staphylococcus aureus, Haemophilus* sp. and streptococci.

Clinical features

Presentation of this *very common* condition is with a subacute onset of bilateral, but frequently asymmetrical, irritation and a mucopurulent discharge.

Signs

- Generalized conjunctival hyperaemia most marked in the fornices.
- Mild papillary reaction.

Treatment

- Chloramphenicol or gentamicin drops 2-hourly during the day and ointment at night for at least 10 days.
- Fusidic acid drops three times a day are a useful alternative.

Gonococcal conjunctivitis

Clinical features

Presentation of this *very rare* condition is with an acute onset of bilateral purulent discharge.

Signs

- Severe conjunctival injection and chemosis.
- Pseudomembranes may be present.

Complications are corneal ulceration and perforation.

Treatment

- Systemic aqueous procaine penicillin G 4.8 mega-units i.m. in two divided doses combined with probenecid 1 g by mouth.
- In penicillin-resistant cases spectinomycin 2 g i.m. in a single dose.
- Topical bacitracin ointment 2-hourly.

VIRAL CONJUNCTIVITIS

Adenoviral keratoconjunctivitis

Causative viruses

- Adenovirus types 3 and 7 in pharyngoconjunctival fever (PCF).
- Adenovirus types 8 and 19 in epidemic keratoconjunctivitis (EKC).

Clinical features

Presentation of this *common* condition is with an acute onset of bilateral discomfort and a watery discharge.

Signs

- Follicular conjunctivitis.
- Preauricular lymphadenopathy.
- Subconjunctival haemorrhages and pseudomembranes in severe cases.
- Keratitis which can be subdivided into three stages:
  stage 1: diffuse punctate epithelial keratitis which either resolves within 2 weeks or passes into stage 2;

  stage 2: focal subepithelial opacities which may pass into stage 3;

  stage 3: anterior stromal infiltrates which may persist for months.

Complications

- Mild anterior uveitis (common).
- Disciform keratitis (rare).

Treatment

- *Of conjunctivitis* is supportive because antivirals are ineffective.
- *Of severe keratitis* is with topical steroids.

Molluscum contagiosum

Cause

Toxic response to a nearby skin virus.

Clinical features

Presentation of this *uncommon* condition is during childhood with unilateral chronic ocular irritation.

Signs

- Small pale umbilicated lid nodules.
- Ipsilateral follicular conjunctivitis.
- Ipsilateral fine punctate keratitis.

Complications

- Superior micropannus (uncommon).
- Punctal occlusion (rare).

Treatment

Expression or cauterization of the lid nodules.

CHLAMYDIAL CONJUNCTIVITIS

Adult inclusion conjunctivitis

Cause

This *uncommon* infection with serotypes D–K of *Chlamydia trachomatis* is acquired venereally and may be associated with urethritis or cervicitis.

Clinical features

Presentation is typically in young adults with a subacute onset of unilateral or bilateral mucopurulent discharge. Unless treated the infection becomes chronic.

Signs

- Large follicles in the fornices and near the limbus.
- Preauricular lymphadenopathy.
- Superior epithelial keratitis and peripheral subepithelial infiltrates.

Complication – superior micropannus in chronic cases.

Management

Laboratory tests

- Necessary to confirm the diagnosis.
- Tests for syphilis and gonorrhoea should also be performed.

Treatment

- Topical tetracycline ointment four times a day for 6 weeks.
- Oral tetracycline 250 mg four times a day for 6 weeks (erythromycin in pregnancy).
- Doxycycline 300 mg weekly for 3 weeks or 100 mg daily for 1–2 weeks as an alternative to tetracycline.

Trachoma

Cause

Serotypes A, B, Ba and C of *Chlamydia trachomatis*.

Clinical features

- *Stage 1* (incipient trachoma) – immature follicles in upper tarsus.
- *Stage 2* (established trachoma) – mature follicles.
- *Stage 3* (cicatricial trachoma) – follicles and scarring of upper tarsus.
- *Stage 4* (healed trachoma) – conjunctival scarring without follicles.

Complications

Conjunctiva

- Stellate and linear scars (Arlt's line).
- Xerosis due to destruction of goblet cells.
- Keratoconjunctivitis sicca due to scarring of lacrimal ductules.

Cornea

- Herbert's pits at the limbus.
- Peripheral and central infiltrates.
- Pannus.

Eyelids

- Trichiasis.
- Cicatricial entropion of the upper eyelid.

Treatment

- Of active disease is similar to adult inclusion conjunctivitis.
- Surgery for corneal and eyelid complications.

NEONATAL CONJUNCTIVITIS (OPHTHALMIA NEONATORUM)

Definition

A conjunctival inflammation occurring during the first month of life.

Chemical

Caused by prophylactic antibiotics or silver nitrate.

Presentation of this *common* condition is within the first few hours of birth with mild and transient conjunctival hyperaemia.

Treatment is unnecessary.

Chlamydial

Caused by transmission of the infection from the mother's birth canal.

Presentation of this *common* infection is between the fifth and fourteenth day with a mucopurulent papillary (not follicular) conjunctivitis.

Complications are conjunctival scarring and superior corneal pannus. It may be associated with otitis, rhinitis and pneumonitis.

Treatment is with topical tetracycline and systemic erythromycin for 3 weeks.

Gonococcal

Caused by transmission of the infection from the mother's birth canal.

Presentation of this *very rare* infection is between the second and fourth day with an acute purulent conjunctivitis and chemosis which may be associated with pseudomembranes.

Complications are corneal ulceration and perforation.

Treatment is with topical and systemic penicillin.

Herpes simplex virus (HSV)

Caused by transmission of HSV type 2 virus from the mother's birth canal.

Presentation of this *rare* infection is between the fifth and seventh days with blepharoconjunctivitis which may be associated with a keratitis.

Treatment is with topical antivirals.

Bacterial

Caused by transmission of infection from the mother's birth canal or postnatally.

Presentation of this *common* condition is at any time during the first month of life.

Treatment is with topical antibiotics.

ALLERGIC CONJUNCTIVITIS

Hay fever (seasonal allergic) conjunctivitis

Cause

Allergic reaction occurring in patients with hay fever.

Clinical features

Presentation of this *very common* condition is usually during childhood or early in adult life with bilateral acute itching and watering.

Treatment

- Sodium cromoglycate four times a day.
- Systemic antihistamines.

Chronic 'allergic' conjunctivitis

Causes

Hypersensitivity or toxic reaction to:

- Preservatives.

- Prolonged use of antiviral agents.
- Antiglaucoma drugs.

Clinical features

Presentation of this *common* condition is at any age with a gradual onset of unilateral or bilateral non-specific chronic irritation and redness.

Signs are conjunctival hyperaemia with follicles or papillae.

Treatment

- Removal of the irritant if known.
- Sodium cromoglycate.

Vernal keratoconjunctivitis (spring catarrh)

Cause

IgE-mediated hypersensitivity.

Clinical features

Presentation of this *uncommon* condition is during childhood with a subacute onset of bilateral itching, lacrimation and photophobia associated with a mucoid discharge. Remissions and exacerbations are frequent.

Signs

- Palpebral conjunctivitis (common) – diffuse papillae in the upper tarsus which may later turn into cobblestones.
- Limbitis (rare) – thick oedematous limbal conjunctiva with mucoid nodules and Trantas' dots.

Complications

- Superior punctate epithelial erosions (very common).
- Macroerosions coated with exudate leading to plaques (uncommon).

- Subepithelial scarring (rare).
- 'Cupid bow' pseudogerontoxon which resembles arcus senilis (rare).

Treatment

- Topical steroids as initial short-term treatment and during severe exacerbations.
- Sodium cromoglycate 2% drops (Opticrom) four times a day for long-term therapy and prophylaxis.

Atopic keratoconjunctivitis

Cause

The adult equivalent of vernal disease which typically affects young adult atopic males.

Clinical features

Presentation of this *uncommon* condition is similar to vernal disease but in a young adult.

Signs

- Papillary conjunctivitis in early cases.
- Corneal pannus in severe long-standing cases.
- Eyelids are fissured from eczema.
- Associated staphylococcal blepharitis is common.

Associations

- Keratoconus.
- Posterior subcapsular cataracts.

Treatment

Topical steroids and sodium cromoglycate.

Giant papillary conjunctivitis

Causes

Allergic reaction to:

- Contact lens deposits.
- Preservatives in contact lens cleansing solutions.
- Artificial eyes.
- Protruding monofilament sutures following surgery.

Clinical features

Presentation of this *uncommon* condition is with chronic ocular irritation and a mucoid discharge.

Signs are mild superior papillary reaction later forming giant papillae.

Treatment

- Removal of the cause if possible (e.g. change type of contact lens).
- Short course of a topical steroid or sodium cromoglycate.

CICATRIZING CONJUNCTIVITIS

Ocular cicatricial pemphigoid

Cause

Autoimmunity.

Systemic features

- *Skin lesions* are recurrent, vesiculobullous, non-scarring eruptions and localized erythematous plaques.
- *Mucous membrane lesions* are submucous blisters which on rupturing lead to scarring and stricture formation.

Ocular features

Presentation of this *rare* disease is during late adult life with a subacute onset of bilateral but asymmetrical ocular irritation and mucoid discharge.

Signs

In chronological order:

- Papillary conjunctivitis.
- Subconjunctival bullae.
- Conjunctival ulceration due to burst bullae.
- Healing and chronic inflammation.
- Fibrosis and conjunctival shrinkage.

Complications

Conjunctival

- Symblepharon due to formation of adhesions between the palpebral and bulbar conjunctiva (very common).
- Dryness due to scarring of lacrimal ductules (common) and destruction of goblet cells (uncommon).

Corneal

- Scarring due to entropion, trichiasis, lagophthalmos and KCS (common).
- Bacterial keratitis due to epithelial defects (uncommon).
- Perforation (rare).

Eyelid

- Ankyloblepharon due to formation adhesions between the upper and the lower eyelids at the lateral canthi (common).
- Cicatricial entropion due to conjunctival scarring (uncommon).
- Lagophthalmos due to conjunctival scarring (uncommon).

Treatment

Medical

- Steroids, both systemic and topical, during the acute phase.
- Cytotoxic agents may be helpful during the chronic phase.
- Ocular lubricants.

Surgical

- For eyelid deformities.
- Keratoprostheses for severe corneal scarring.

Stevens–Johnson syndrome (erythema multiforme)

Cause

Hypersensitivity reaction to drugs and viruses giving rise to an acute vasculitis of the skin and conjunctiva.

Systemic features

- Malaise, arthralgia etc.
- Vesiculocutaneous eruption with haemorrhages into the vesicles (target lesions) followed by healing with or without scarring.

Ocular features

Presentation of this *uncommon* condition is at any age with a bilateral subacute onset of mucopurulent conjunctivitis.

Signs

In chronological order:

- Non-specific conjunctivitis.
- Focal conjunctival infarcts leading to membrane formation.
- Spontaneous resolution or complications in severe cases.

Complications

- Conjunctival scarring and keratinization.
- Corneal scarring secondary to conjunctival keratinization and lid deformities.
- Cicatricial entropion and acquired distichiasis.

Treatment

- Steroids, both topical and systemic, during the acute stage.
- Surgery for eyelid deformities.

MISCELLANEOUS SYNDROMES

Superior limbic keratoconjunctivitis of Theodore

Clinical features

Presentation of this *uncommon* condition is typically in middle-aged

women with thyroid dysfunction with bilateral, asymmetrical, chronic recurrent ocular irritation.

Signs

- Papillae on superior tarsus.
- Hyperaemia and keratinization of superior bulbar conjunctiva.
- Limbal papillae and thickening.
- Superior corneal punctate epithelial erosions.
- Superior corneal filaments.

Treatment

For symptomatic relief

- Adrenaline 1% drops.
- Silver nitrate 1% to the superior tarsus.
- Soft bandage contact lenses.
- Acetylcysteine 5% drops for severe corneal filaments.

For permanent cure

- Recession or resection of the superior bulbar conjunctiva.
- Thermocauterization of the superior bulbar conjunctiva.

Parinaud's oculoglandular conjunctivitis

Causes

- Cat scratch fever.
- Tularaemia.
- Sporotrichosis.
- Tuberculosis.
- Sarcoidosis.
- Syphilis.
- Lymphogranuloma venereum.

Clinical features

- Severe unilateral granulomatous conjunctivitis.
- Marked preauricular lymphadenopathy.

Treatment

Varies according to the cause.

PIGMENTED CONJUNCTIVAL LESIONS

Classification

Melanocytic

- Conjunctival epithelial melanosis
- Congenital melanosis oculi
- Naevus
- Precancerous melanosis
- Primary melanoma

Non-melanocytic

- Endogenous
- Exogenous

Conjunctival epithelial melanosis

Clinical features

Presentation of this *common* lesion is during childhood with unilateral or bilateral flat patches of brown pigmentation, most marked at the limbus.

Treatment

Unnecessary.

Congenital melanosis oculi

Clinical features

This *rare* condition presents at birth and is of two types.

Isolated

- Usually unilateral subepithelial bluish-black patches.
- No malignant potential.

Naevus of Ota

- Subepithelial melanosis.
- Skin hyperpigmentation in the distibution of the first and second divisions of the trigeminal nerve.
- Uveal hyperpigmentation (common).
- Subsequent glaucoma (rare).
- Malignant uveal melanomas (very rare).

Treatment

Unnecessary but watch for glaucoma and melanoma in the naevus of Ota.

Conjunctival naevus

Clinical features

Presentation of this *uncommon* lesion is during childhood as a single, unilateral, well-demarcated flat lesion near the limbus with variable pigmentation. Both the size and degree of pigmentation may increase at puberty or during pregnancy.

Treatment

Excision for comfort or cosmesis.

Precancerous melanosis

Clinical features

Presentation of this *very rare* condition is during old age. The two types are:

- *Superficial spreading melanoma* – one or more areas of slowly growing intraepithelial conjunctival pigmentation.
- *Lentigo maligna* (Hutchinson's freckle) – typically affects the conjunctiva and face of the very elderly.

Treatment

Excision if the lesion is thick and nodular.

Primary conjunctival melanoma

Clinical features

Presentation of this *very rare* tumour is in adults with a pigmented or non-pigmented nodule most commonly located near the limbus.

Treatment

- Local excision if deep invasion is absent.
- Enucleation if scleral invasion is present.
- Exenteration if the lids or orbit are invaded.

NON-PIGMENTED CONJUNCTIVAL TUMOURS

Papilloma

Clinical features

This *rare* benign tumour may be sessile or pedunculated.

Treatment

Excision but recurrences are common.

Intraepithelial epithelioma (carcinoma in situ)

Clinical features

This *very rare* tumour presents as a vascularized, elevated, fleshy mass located near the limbus.

Treatment

Excision.

Invasive squamous cell carcinoma

Clinical features

This *very rare* tumour is similar to an intraepithelial epithelioma but it may penetrate deeply.

Treatment

- Local excision of early lesions.
- Enucleation or exenteration of advanced tumours.

Choristoma

Definition

A congenital overgrowth of normal tissue in abnormal locations.

Clinical features

This *uncommon* lesion is of two types:

- *Dermoid* is a solid, white, limbal mass.
- *Lipodermoid* is a soft, yellow, movable subconjunctival mass at or near the limbus or outer canthus.

Associations

Goldenhar's syndrome, which consists of the following features, may be associated with choristomas:

- Preauricular skin tags.
- Hemifacial hypoplasia.
- Vertebral anomalies.

Treatment

Excision.

Chapter 5

The cornea and sclera

EVALUATION OF CORNEAL LESIONS

Punctate epithelial erosions (PEEs)

Appearance – depressed superficial grey–white epithelial spots.

Staining is good with fluorescein but poor with rose bengal.

Causes

- *Superior* – subtarsal foreign body, vernal disease, chlamydial infections and superior limbic keratoconjunctivitis.
- *Inferior* – trichiasis, entropion, staphylococcal blepharitis, exposure keratopathy, acne rosacea and drug toxicity.
- *Interpalpebral* – seborrhoeic blepharitis, keratoconjunctivitis sicca, neurotrophic keratopathy and exposure to ultraviolet light.

Epithelial filaments

Appearance – comma-shaped filaments of mucus and degenerate epithelial cells.

Staining is poor with fluorescein but good with rose bengal.

Causes

- Keratoconjunctivitis sicca.
- Superior limbic keratoconjunctivitis.
- Neurotrophic keratopathy.
- Herpes zoster keratitis.
- Recurrent corneal erosions.
- Prolonged patching.

Punctate epithelial keratitis (PEK)

Appearance – small granular opalescent epithelial or subepithelial spots

Staining is poor with fluorescein but good with rose bengal.

Causes

- Viral infections.
- Chlamydial infections.

Pannus

Appearance – ingrowth of fibrovascular tissue from the limbus.

Causes

- *Superior* – contact lens wear and chlamydial infections.
- *Inferior* – trichiasis, exposure keratopathy and acne rosacea.
- *Generalized* – atopic keratoconjunctivitis and cicatrizing conjunctivitis.

Pigmentation

See Table 2.

Table 2

Type of pigment	*Causes*	*Location*
Iron	Keratoconus (Fleischer's ring)	Epithelium
	Pterygium (Stocker's line)	Epithelium
	Filtering bleb (Ferry's line)	Epithelium
	Old age (Hudson–Stahli line)	Epithelium
	Siderosis	Stroma
Silver	Argyrosis	Stroma
Gold	Chrysiasis	Epithelium
Copper	Wilson's disease (Kayser–Fleischer ring)	Descemet's membrane
Melanin	Pigment dispersion (Krukenberg's spindle)	Endothelium

Vital staining

- *Fluorescein* remains extracellular, stains the tear film and shows up epithelial defects but does not stain mucus.
- *Rose bengal* stains damaged cells and mucus but not epithelial defects.

MICROBIAL KERATITIS

Bacterial keratitis

Predispositions

- Chronic adnexal infection.
- Underlying corneal disease.
- Keratoconjunctivitis sicca.
- Contact lens wear (particularly soft).
- Neurotrophic and exposure keratopathy.
- Long-term use of topical steroids.

Clinical features

Presentation of this very serious *uncommon* condition is with a subacute onset of unilateral pain, redness and blurred vision.

Signs

- Staphylococci and streptococci – oval, yellow–white, dense suppuration surrounded by relatively clear cornea.
- *Pseudomonas* sp. – irregular suppuration and a thick mucopurulent exudate.

Management

Identification of organism by scraping the base of the ulcer and performing cultures.

Preparation of fortified antibiotics

- *Gentamicin:* take 2 ml (40 mg) of parenteral preparation and add to 5 ml bottle of commercially available gentamicin eye drops.
- *Cephazolin:* (1) take 2 ml of sterile saline; (2) add to 500 mg of

cephazolin and dissolve powder; (3) remove 2 ml from a 15 ml bottle artificial tears; and (4) inject reconstituted cephazolin into bottle of artificial tears.

Treatment

- Topical gentamicin and cephazolin at half-hourly intervals.
- Subconjunctival gentamicin 40 mg and cephazolin 125 mg at 24-hourly intervals.

Fungal keratitis

Predispositions

- Filamentous (*Aspergillus* and *Fusarium* spp.) typically is preceded by ocular trauma with vegetable matter.
- Yeast (*Candida*) usually occurs in a compromised host.

Signs

- *Filamentous* – greyish-white lesion with feathery projections into the stroma with an intact overlying epithelium.
- *Yeast* – dense white–yellow suppuration similar to bacterial keratitis.

Treatment

- *Filamentous* with oral and topical ketoconazole.
- *Yeast* with oral and topical flucytosine.

Acanthamoeba keratitis

Predisposition

This *rare* condition typically affects wearers of soft contact lenses.

Signs

Recurrent breakdown of epithelium associated with chronic deep paracentral infiltrate or abscess.

Treatment

- Brolene ointment and drops.
- Keratoplasty in resistant cases.

Interstitial keratitis

Definition

An inflammation of the corneal stroma without primary involvement of the epithelium or endothelium.

Causes

- Late manifestation of congenital syphilis.
- Tuberculosis.
- Cogan's syndrome (interstitial keratitis + acute tinnitus + vertigo + deafness).

Treatment

- Luetic with systemic penicillin and topical steroids.
- Tuberculous with antituberculous drugs.
- Cogan's syndrome with topical and systemic steroids.

HERPES SIMPLEX KERATITIS

Classification

Primary

- Blepharoconjunctivitis
- Keratitis

Recurrent

- Epithelial
- Stromal

Trophic

Primary infection

Cause

Direct transmission of virus through infected secretions to a non-immune subject.

Clinical features

Presentation of this *rare* condition is between the age of 6 months and 5 years with unilateral ocular irritation and redness.

Signs

- Unilateral follicular conjunctivitis.
- Blepharitis.
- Keratitis in 50% of cases.

Treatment

Antivirals.

Recurrent epithelial keratitis

Cause

Reactivation of latent virus and invasion of the epithelium.

Clinical features

Presentation

This *common* condition presents with an acute onset of unilateral irritation, slight photophobia and variable blurring of vision.

Signs

Dendritic ulcer:

- Starts with coarse stellate PEK and develops into a branching ulcer.
- Fluorescein stains bed of ulcer and rose bengal stains its margins.
- Corneal sensation is reduced.

Geographical ulcer starts as a dendritic ulcer and enlarges to assume a geographical shape.

Treatment

Antiviral agents

Acyclovir 3% ointment five times a day:

- Efficacy ++++
- Toxicity +

Trifluorothymidine 1% drops 2-hourly:

- Efficacy ++++
- Toxicity +++

Adenine arabinoside 3% ointment, 0.1% drops:

- Efficacy +++
- Toxicity ++

Idoxuridine 0.5% ointment, 0.1% drops:

- Efficacy +++
- Toxicity +++

Indications for débridement

- Resistance to antivirals.
- Allergy to antivirals.
- Unavailability of antivirals.
- Non-compliance.

Stromal necrotic keratitis

Cause

Direct viral invasion and destruction of the stroma.

Clinical features

Presentation of this *uncommon* condition is with a gradual onset of unilateral pain, redness and severe visual impairment.

Signs are a cheesy and necrotic stroma similar to bacterial or fungal infection.

Complications

- Vascularization and scarring (common).
- Perforation (rare).

Treatment

- *Lubricants* and patching to heal an associated epithelial defect.

- *Acyclovir* may be useful as it penetrates the stroma.
- *Steroids* may be useful once the epithelium has healed but must be used with great caution.

Disciform keratitis

Cause

Hypersensitivity reaction to the virus.

Clinical features

Presentation of this *common* lesion is with a subacute onset of unilateral blurred vision which may be associated with haloes around lights.

Signs

- Central (occasionally eccentric) zone of stromal and epithelial oedema.
- Keratic precipitates underlying the involved area.
- Small infiltrates (Wessely's ring) may surround the lesion.

Treatment

- *Steroids* initially five times a day then reduced gradually over several weeks; weak dilutions of prednisolone (0.25–0.025%) are also useful.
- *Acyclovir* ointment twice daily as an 'umbrella' but beware of epithelial toxicity with long-term use.

Trophic keratitis

Cause

Persistent defects in basement membrane.

Treatment

See later.

HERPES ZOSTER OPHTHALMICUS

Classification

- Stage 1 – acute
- Stage 2 – chronic
- Stage 3 – recurrent

Stage 1 (acute)

Skin lesions

Signs in chronological order:

- Unilateral maculopapular rash.
- Pustules which burst.
- Crusting ulcers.
- Scarring.

Treatment

- Oral acyclovir 800 mg 5 times a day for 7 days.
- Topical antibiotic–steroid ointment applied three times a day.

Ocular lesions

Signs

Mucopurulent conjunctivitis (common and transient).

Episcleritis (common and transient).

Keratitis can be of the following types:

- PEK (very common).
- Filamentary (common).
- Epithelial microdendrites (common).
- Nummular deposits in the anterior stroma (very common).
- Disciform (uncommon).

Anterior uveitis in about 50% (see Chapter 6).

Scleritis (rare).

Treatment

Topical steroids for keratitis and uveitis.

Neurological complications

These are rare and include:

- Extraocular nerve palsies.
- Optic neuritis.
- Encephalitis.
- Contralateral hemiplegia.

Stage 2 (chronic)

Skin lesions

Punched-out scars.

Ocular lesions

- Mucus-secreting conjunctivitis.
- Scleritis.
- Keratitis.
- Chronic anterior uveitis.

Stage 3 (recurrent)

- Episcleritis.
- Scleritis.
- Keratitis.
- Anterior uveitis.

PERIPHERAL CORNEAL THINNING AND ULCERATION

Dellen

Cause

Localized tear film instability and stromal dehydration secondary to raised limbal lesions or chemosis.

Clinical features

This *common* condition is characterized by a unilateral, saucer-shaped thinning which is usually asymptomatic, transient and innocuous.

Treatment

Patching and lubricants to promote corneal rehydration.

Marginal keratitis (catarrhal ulcer)

Cause

Hypersensitivity reaction to staphylococcal exotoxins which may be associated with staphylococcal blepharitis.

Clinical features

Presentation of this *very common* condition is with a unilateral subacute onset of mild irritation and lacrimation.

Signs in chronological order:

- Whitish subepithelial limbal infiltrate.
- May spread circumferentially.
- Epithelial breakdown.
- Healing without scarring within a few days.

Treatment

Topical steroids for about 1 week are very effective but associated blepharitis should also be treated.

Rosacea keratitis

Clinical features

Presentation of this *common* condition, which affects about 5% of patients with acne rosacea, is similar to a marginal keratitis but it may be bilateral.

Signs in chronological order:

- Inferior PEEs.
- Peripheral infltration and vascularization.
- Wedge-shaped scarring.

Complication in severe cases is corneal thinning which may rarely lead to perforation.

Treatment

- Topical steroids as a short-term measure.
- Systemic oxytetracycline – 250 mg four times a day for 1 month then 250 mg daily for 6 months.

Phlyctenulosis

Cause

Delayed hypersensitivity to staphylococcal or other bacterial antigens.

Clinical features

Presentation of this *rare* disorder is in children with an acute onset of usually unilateral pain and severe photophobia.

Signs

- *A conjunctival phlycten* is a small pinkish nodule near the limbus which resolves spontaneously.
- *A corneal phlycten* sits astride the limbus and usually resolves spontaneously.

Complications of a corneal phlycten are occasional extension into the cornea and ulceration leaving a triangular scar.

Treatment

Topical steroids.

Terrien's marginal degeneration

Clinical features

Presentation of this *uncommon* condition is during adult life. Initially the condition is asymptomatic and later it causes blurred vision due to astigmatism.

Signs in chronological order:

- Bilateral fine, yellow–white, punctate, peripheral stromal opacities.
- Progression to circumferential guttering and lipid deposition.

Complications

- Pseudopterygia (common).
- Severe astigmatism (common).
- Perforation (rare).

Treatment

Lamellar keratoplasty in advanced cases.

Mooren's ulcer

Cause

Vasculitis of the limbal vessels which leads to an ischaemic necrosis.

Clinical features

Presentation of this *very rare* condition is in adult life with severe pain.

Two main types:

- *Limited* type is unilateral and affects the elderly.
- *Progressive* type is bilateral and affects younger individuals.

Signs in chronological order:

- Grey peripheral corneal infiltrate.
- Undermining of the epithelium and superficial stroma.
- Circumferential and occasional central spread.

Complications

- Perforation.
- Cataract.
- Secondary glaucoma.

Treatment

- Topical steroids.
- Conjunctival excision adjacent to the ulcer.
- Systemic immunosuppressive agents.

Peripheral ulceration in rheumatoid arthritis

Clinical features

- *Sclerosing keratitis* – stromal thickening, vascularization and lipid deposition.
- *Acute stromal keratitis* – stromal infiltration, vascularization ± thinning.
- *Chronic peripheral thinning* (contact lens cornea) is asymptomatic but it may perforate.
- *Keratolysis* – acute severe thinning which may perforate.

Treatment

This is similar to Mooren's ulcer.

MISCELLANEOUS KERATOPATHIES

Exposure keratopathy

Cause

Incomplete blinking leading to inadequate corneal wetting by tears due to:

- Facial nerve palsy.
- Severe proptosis.
- Lid scarring.

Clinical features

Initially inferior PEEs which may progress to ulceration and infection.

Treatment

- Eliminate cause if possible.
- Ocular lubricants.
- Tape eyelids at night.
- Protective soft contact lenses but beware of infection.
- Eyelid surgery.

Neurotrophic keratopathy

Cause

Corneal anaesthesia due to:

Acquired lesions *(common)*

- Section of the fifth nerve.
- Herpes simplex and zoster keratitis.
- Leprosy.

Congenital lesions *(very rare)*

- Familial dysautonomia (Riley–Day syndrome).
- Anhidrotic familial ectodermal dysplasia.
- Insensitivity to pain.

Clinical features

Initially interpalpebral PEEs which may progress to ulceration and infection.

Treatment

Similar to exposure keratopathy.

Thygeson's superficial punctate keratitis (SPK)

Clinical features

Presentation of this *uncommon* condition is in early adult life with an acute onset of bilateral irritation and photophobia. The keratitis persists for several years with remissions and recurrences.

Signs – small, stellate conglomerates of grey–white intraepithelial dots which may be associated with a mild subepithelial haze.

Treatment

- Topical steroids are effective but may prolong the condition.
- Ocular lubricants and soft contact lenses for symptomatic relief.
- Trifluorothymidine may also be effective.

CORNEAL DEGENERATIONS

Lipid keratopathy

Causes

- *Primary* occurring spontaneously in a normal cornea (very rare).
- *Secondary* degeneration in a vascularized cornea (common).

Clinical features

Yellow stromal deposits of cholesterol and fat.

Treatment

- Argon laser photocoagulation to the new vessels may induce resorption.
- Keratoplasty in severe cases.

Band keratopathy

Causes

- Chronic anterior uveitis, particularly in children (common).
- Phthisis bulbi (common).

- Idiopathic in the elderly (rare).
- Hypercalcaemia (very rare)

Clinical features

Subepithelial calcification involving the interpalpebral cornea with a clear space at the limbus.

Treatment

Overlying epithelium is scraped off and sodium versenate (0.01 M solution) is applied to the denuded area (chelation).

Salzmann's nodular degeneration

Causes

A secondary degeneration following chronic keratitis.

Clinical features

This *uncommon* degeneration is characterized by elevated subepithelial nodules in scarred cornea. The base of the nodule may be surrounded by epithelial iron deposits.

Treatment

Keratoplasty if severe.

CORNEAL DYSTROPHIES

Classification

Anterior dystrophies

- Cogan's microcystic
- Recurrent corneal erosions syndrome
- Reis–Bücklers
- Meesmann's

Stromal dystrophies

- Lattice
- Macular
- Granular

Posterior dystrophies

- Fuchs'
- Posterior polymorphous

Ectatic dystrophies

- Anterior keratoconus
- Posterior keratoconus
- Keratoglobus
- Pellucid marginal degeneration

Cogan's microcystic dystrophy

Other names

Map–dot–fingerprint, bleb-like dystrophy and epithelial basement membrane dystrophy.

Inheritance

None but more common in females.

Clinical features

Presentation of this *common* dystrophy is during the third decade with recurrent corneal erosions which may be bilateral.

Signs are bilateral dot-like, cystic or linear fingerprint-like epithelial lesions.

Treatment

Unnecessary in absence of recurrent erosions.

Recurrent corneal erosion syndrome

Causes

- Corneal dystrophies – Cogan's microcystic, Reis–Bückler's and lattice.
- Traumatic corneal abrasion (most common).

Clinical features

This *very common* condition usually presents on waking with acute pain, lacrimation and photophobia.

Treatment

- *Mild* cases with tear substitutes during the day and ointment prior to going to sleep at night.
- *Severe isolated* episodes by removal of the loose epithelium and padding until the epithelium regenerates.
- *Severe recurrent* with bandage contact lenses for 3 months.

Reis–Bücklers' dystrophy

Inheritance

Dominant.

Clinical features

Presentation of this *uncommon* dystrophy is during the first decade with recurrent erosions.

Signs are bilateral, superficial, ring-like opacities at the level of Bowman's layer.

Treatment

Lamellar keratoplasty by the second to third decade in some cases.

Meesmann's dystrophy

Inheritance

Dominant.

Clinical features

Presentation of this *very rare* dystrophy is between the first and second decades with mild discomfort.

Signs are bilateral tiny epithelial cysts which are most numerous in the interpalpebral cornea.

Treatment

Unnecessary.

Lattice dystrophy

Inheritance

Dominant.

Clinical features

Presentation of this *rare* dystrophy is during the first decade with recurrent erosions.

Signs are bilateral branching and interlacing spider-like deposits in the stroma.

Histological staining

Congo red and Masson trichrome.

Treatment

Penetrating keratoplasty by the third decade.

Granular dystrophy (Groenouw's type I)

Inheritance

Dominant.

Clinical features

Presentation of this *rare* dystrophy is during the first decade without symptoms.

Signs are bilateral, discrete, crumb-like, white hyaline granules in the anterior stroma without involvement of the peripheral cornea.

Histological staining

Masson trichrome.

Treatment

Usually unnecessary.

Macular dystrophy (Groenouw's type 2)

Inheritance

Recessive.

Clinical features

Presentation of this *rare* dystrophy is during the first decade with poor visual acuity and mild irritation.

Signs are bilateral grey–white poorly delineated opacities which eventually involve the entire stroma and the peripheral cornea.

Histological staining

Alcian blue.

Treatment

Penetrating keratoplasty by second to third decade.

Fuchs' dystrophy

Inheritance

Occasionally dominant and more common in females.

Clinical features

Presentation of this *uncommon* dystrophy is between the fifth and seventh decades with initially unilateral impaired visual acuity and discomfort.

Signs in chronological order:

- Asymptomatic bilateral corneal guttata (tiny excrescences on Descemet's membrane).
- Progressive but asymmetrical stromal oedema.
- Bullous keratopathy and pain.

Treatment

- Hypertonic agents (5% sodium chloride) to reduce epithelial oedema.
- Bandage contact lenses to protect exposed nerve endings and reduce pain.
- Penetrating keratoplasty in advanced cases (80% success).

Posterior polymorphous dystrophy

Inheritance

Dominant.

Clinical features

Presentation of this *very rare* dystrophy is during the first decade without symptoms.

Signs are usually bilateral, vesicular, geographical or band-like opacities on the posterior cornea which may be associated with iris anomalies and glaucoma.

Treatment

Usually unnecessary although some develop corneal oedema.

Anterior keratoconus

Inheritance

Usually nil.

Clinical features

Presentation of this *common* dystrophy is during the second to third

decade with impaired visual acuity due to irregular astigmatism; 85% of cases are bilateral but asymmetrical.

Signs in chronological order:

- Irregular astigmatism confirmed by keratometry or Placido's disc.
- Fine vertical folds in the deep stroma (Vogt's striae).
- Progressive central thinning with bulging of the lower lid on down-gaze (*Munson's sign*) and epithelial iron deposits at base of cone (*Fleischer's ring*).
- Ruptures in Descemet's membrane and leakage of fluid into the stroma and epithelium (acute hydrops).
- Scarring.

Treatment

- Contact lenses to correct astigmatism.
- Hypertonic saline, patching ± soft contact lenses for acute hydrops.
- Keratoplasty or epikeratophakia for scarring.

Associations

Systemic

- Syndromes: Down's, Ehlers–Danlos, Marfan's and Turner's.
- Atopic dermatitis
- Osteogenesis imperfecta.

Ocular

- Retinitis pigmentosa and Leber's amaurosis.
- Vernal disease.
- Aniridia.
- Ectopia lentis.

Posterior keratoconus

Inheritance

Nil.

Clinical features

A *very rare*, bilateral, non-progressive excavation in the posterior cornea.

Treatment

Unnecessary.

Keratoglobus

Inheritance

Mode of transmission is unknown.

Clinical features

A *very rare* bilateral thinning and protrusion of the entire cornea.

Treatment

Unnecessary.

Pellucid marginal degeneration

Inheritance

Nil.

Clinical features

A *very rare*, bilateral, progressive, inferior peripheral thinning with protrusion of the cornea above.

Treatment

Keratoplasty in some cases.

CORNEAL CHANGES IN TOXIC AND METABOLIC DISORDERS

Wilson's disease (hepatolenticular degeneration)

Cause

Deficiency in the α_2-globulin ceruloplasmin leading to the deposition of copper in the tissues.

Systemic presentations

- Infancy with a flapping tremor.
- Early childhood with jaundice and hepatosplenomegaly.
- Late childhood with cerebral degeneration.
- Early adult life with cirrhosis or mental changes.

Ocular features

- Kayser–Fleischer ring in the periphery of Descemet's membrane (common).
- Sunflower cataracts (uncommon).

Cornea verticillata (vortex keratopathy)

Causes

- *Chloroquine* – unrelated to either the dose or the duration of administration; reversible on cessation.
- *Amiodarone* – related to the dose and duration of administration; severe keratopathy occurs with doses of 400–1400 mg/day.
- *Fabry's disease* – deficiency in α-galactosidase A giving rise to telangiectatic skin lesions (angiokeratomas), cardiovascular and renal lesions, and pain of fingers and toes.

Clinical features

Bilateral greyish or golden epithelial deposits swirling outwards from a point below the centre of the cornea.

Crystalline deposits

Causes

- Gold (chrysiasis) – tiny epithelial particles.
- Cystinosis – stromal deposits.
- Monoclonal gammopathy – stromal deposits.

Mucopolysaccharidoses

Stromal corneal deposits occur in:

- Hurler (severe and present at birth).
- Scheie (severe and present at birth).
- Morquio (mild).
- Maroteaux–Lamy (mild).

CONTACT LENSES

Types of contact lenses

Hard lenses

Material is polymethylmethacrylate (PMMA).

Types

- Corneal with a diameter of 8.5–10 mm are most frequently used.
- Scleral (haptic) override the cornea and rest on the sclera, and are now rarely used.
- Hybrid have the same diameter as the cornea and are used to hide unsightly eye.

Advantages

- Durable.
- Visual acuity is excellent.
- Low incidence of serious complications.

Disadvantages

- Suitable only for daily wear as they are impermeable to oxygen.
- Require a prolonged adaptation period.

Soft (hydrophilic) lenses

Material is hydroxymethylmethacrylate (HEMA); the degree of hydration varies between 25% and 85%.

Advantages

- Suitable for long-term wear as they are permeable to oxygen.
- Very comfortable and stable.
- Do not require a prolonged adaptation period.

Disadvantages

- Delicate and less durable than hard lenses.
- Visual acuity may not be as crisp.
- High incidence of serious complications (see later).

Gas permeable lenses

Material is a mixture of hard and soft material.

Advantages

- Permeable to oxygen.
- Comfortable and stable.

Disadvantages

- More brittle than soft lenses.
- Easily scratched.

Medical indications for contact lense wear

Optical

- Unilateral aphakia with good vision in the fellow phakic eye to permit fusion.
- High myopia particularly with maculopathy because magnification is increased and visual acuity improved.
- Irregular astigmatism particularly in keratoconus.

Corneal disorders

- Corneal irregularities to replace an irregular surface with a smooth surface.
- Epithelial healing defects to promote healing.
- Severe recurrent corneal erosions.

- Bullous keratopathy to reduce pain.
- Wound leaks as a temporary measure to promote healing.
- Protection of a normal corneal epithelium in trichiasis and exposure.

Miscellaneous

- Ptosis if unsuitable for surgery because of the absence of Bell's phenomenon.
- Cosmetic to hide an unsightly eye.
- Occluders in amblyopia.
- Vehicle for drug delivery in dry eyes and glaucoma.
- Prevention of symblepharon in chemical burns and ocular cicatricial pemphigoid.

Complications of contact lens wear

- Chronic allergic conjunctivitis (see Chapter 4).
- Giant papillary conjunctivitis (see Chapter 4).
- Epithelial oedema due to hypoxia.
- Peripheral corneal vascularization is most common with prolonged wear lenses.
- Sterile corneal ulceration.
- Infection is most common with soft lenses and may be due to *Acanthamoeba* sp. (see Chapter 5).
- Corneal warping with extended wear lenses (rare).

CORNEAL SURGERY

Keratoplasty

Indications

- *Optical* to improve vision.
- *Tectonic* to preserve or restore corneal anatomy.
- *Therapeutic* to remove infected corneal tissue.
- *Cosmetic* to improve appearance.

Prognostic factors

Favourable

- Fresh donor tissue.
- Young patients because the endothelial cell count is denser.
- Localized avascular scar.
- Corneal dystrophies.

Adverse

- Severe stromal vascularization.
- Impaired corneal sensation.
- Active corneal inflammation.
- Uncontrolled glaucoma or uveitis.
- Uncontrolled and progressive conjunctival inflammation.
- Tear film dysfunction

Surgical steps

1. Determination of graft size (ideal size is 7.5 mm).
2. Excision of donor tissue.
3. Excision of host tissue.
4. Fixation of donor tissue with sutures.

Postoperative management

- Steroids four times a day for 2 weeks, then daily for 6 months and finally on alternate days for 6 months.
- Suture removal after 12 months.

Postoperative complications

Early

- Flat anterior chamber.
- Iris prolapse.
- Persistent epithelial defects.
- Infection.

Late

- Glaucoma.

- Astigmatism.
- Retrocorneal membrane formation.
- Wound separation.
- Cystoid macular oedema.
- Recurrence of the initial disease in the graft.

Causes of graft failure

- *Early* is due to endothelial dysfunction; the graft is oedematous on the first postoperative day.
- *Late* is due to allograft reaction and presents within 6 months with ciliary flush, graft oedema and keratic precipitates.

Refractive corneal surgery

Radial keratotomy

Mechanism is flattening of cornea by 16 deep radial corneal incisions.

Indications are adults with stable myopia between 2 D and 8 D.

Contraindications

- Over 8 D because results are unpredictable.
- Age less than 21 years because myopia is still unstable.
- Pre-existing corneal disease.

Results

- 60% are within 1 D of emmetropia.
- 30% are undercorrected by more than 1 D.
- 10% are overcorrected by more than 1 D.

Complications

- Loss of between 1 and 2 Snellen lines of visual acuity in 10%.
- Diurnal fluctuations of vision and glare with night driving.
- Intrastromal epithelial cysts.
- Inability to subsequently wear contact lenses.

Epikeratophakia

Mechanism – refraction is altered by sewing on a lenticle of donor cornea.

Indications

- Severe unilateral myopia.
- Unilateral childhood aphakia.
- Keratoconus.

EPISCLERITIS AND SCLERITIS

Episcleritis

Clinical features

Presentation of this *very common* condition is typically in healthy young women with unilateral redness and slight discomfort.

Signs

- Sectorial or diffuse redness (simple episcleritis).
- Nodule with surrounding injection (nodular episcleritis).

Treatment

- In mild cases is usually unnecessary.
- In severe and persistent cases is with topical steroids and systemic indomethacin 50 mg twice daily.

Scleritis

Classification

Anterior

- Non-necrotizing – diffuse or nodular
- Necrotizing – with or without inflammation

Posterior

Causes

- Idiopathic.
- Herpes zoster is the most common local cause.
- Connective tissue disorders, especially rheumatoid arthritis.
- Sarcoidosis and tuberculosis (both rare).

Anterior non-necrotizing scleritis

Clinical features

Presentation of this *uncommon* condition is with unilateral redness and discomfort.

Signs

- Localized nodule of oedematous sclera (*nodular scleritis*).
- Widespread inflammation (*diffuse scleritis*).

Treatment

Indomethacin 100 mg daily for 4 days then 75 mg daily until resolution.

Anterior necrotizing scleritis with inflammation

Clinical features

Presentation of this *very rare* condition is with severe pain.

Signs are localized scleral injection and necrosis.

Complications

- Keratitis.
- Cataract.
- Secondary glaucoma.

Treatment

Systemic steroids or immunosuppressives.

Anterior scleritis without inflammation (scleromalacia perforans)

Clinical features

Presentation of this *rare* condition is usually in a female with *seropositive rheumatoid arthritis* with an asymptomatic dark patch due to scleral atrophy.

Signs are large patches of scleral necrosis exposing the uvea.

Complications – scleral perforation may occur following ocular trauma but spontaneous perforation is rare.

Treatment

None effective.

Posterior scleritis

Clinical features

A combination of:

- Uveal effusion.
- Exudative retinal detachment.
- Macular and optic disc oedema.
- Proptosis.
- Defective ocular motility.

Treatment

Systemic steroids.

Chapter 6

Uveitis

CLASSIFICATION

Anatomical

- Anterior
- Intermediate
- Posterior
- Diffuse

Clinical

- Acute – sudden onset and duration of less than 6 weeks
- Chronic – insidious onset and lasts for months or years

Aetiological

- Associated with systemic disease
- Parasitic infestations
- Viral infections
- Fungal infections
- Idiopathic specific uveitis entities
- Idiopathic non-specific uveitis entities

Pathological

- Granulomatous
- Non-granulomatous

CLINICAL FEATURES

Anterior uveitis

Symptoms of acute anterior uveitis

- Photophobia is frequently the first symptom.
- Pain due to spasm of the pupil and ciliary muscle.
- Redness.
- Decreased visual acuity if severe.
- Lacrimation if very severe.

In chronic anterior uveitis symptoms may be initially absent.

Signs

- Circumcorneal ('ciliary') injection in acute anterior uveitis.
- Keratic precipitates (KPs) are cellular deposits on the endothelium.
- Aqueous cells are inflammatory white cells circulating in the aqueous humour – their number is graded from +1 to +4.
- Aqueous flare due to leakage of proteins from iris blood vessels – its density is graded from +1 to +4.
- Iris nodules occur in granulomatous inflammation (Koeppe at pupil and Busacca away from pupil).
- Iris atrophy occurs in Fuchs' uveitis syndrome, herpes zoster and herpes simplex.

Posterior uveitis

Symptoms

- Floaters due to vitritis.
- Impaired visual acuity due to macular involvement.

Signs

- Vitreous cells, opacities, flare and posterior detachment.
- Fundus shows chorioretinitis which may be *focal, multifocal* or *geographical.*
- Vasculitis in some cases.

UVEITIS AND ARTHRITIS

Ankylosing spondylitis

Systemic features

- This *common* condition typically affects HLA-B27-positive young men.
- Chronic seronegative (for rheumatoid factor) inflammatory arthritis predominantly affecting the axial skeleton.
- Associations in some cases are ulcerative colitis and Crohn's disease.

Ocular features

Recurrent, unilateral, non-granulomatous, acute anterior uveitis in 30%.

Differential diagnosis of arthritis and acute anterior uveitis.

- Reiter's syndrome.
- Psoriatic arthritis.
- Behçet's disease.
- Sarcoidosis (in young patients).

Reiter's syndrome

Systemic features

- This *uncommon* condition typically affects young men, 70% of whom are HLA-B27 positive.
- Triad of urethritis, conjunctivitis and arthritis.
- Keratoderma blenorrhagica.
- Circinate balanitis.
- Nail dystrophy.
- Painless mouth ulcers.
- Plantar fasciitis.
- Three types of presentation: postvenereal, postdysenteric and articular.

Ocular features

- Conjunctivitis (very common).
- Punctate epithelial or subepithelial keratitis (common).
- Acute anterior uveitis in 20%.

Psoriatic arthritis

Systemic features

- This *uncommon* condition has no sexual preferential but it is associated with an increased prevalence of HLA-B27 and HLA-B17.
- Asymmetrical erosive, inflammatory arthritis occurring in 5% of patients with psoriasis.
- Nail changes consist of pitting and onycholysis.

Ocular features

- Conjunctivitis in 20%
- Acute anterior uveitis (uncommon).
- Keratoconjunctivitis sicca (rare).

Juvenile chronic arthritis

Systemic features

This is an *uncommon* inflammatory arthritis which occurs prior to the age of 16 years.

Presentations based on the onset and the extent of joint involvement during the first 3 months are:

- Systemic onset (20%) with remittent fever, transient maculopapular rash, lymphadenopathy and hepatosplenomegaly. Uveitis is *very rare*.
- Polyarticular onset (20%) with involvement of five or more joints. Systemic features are mild or absent and uveitis is *uncommon*.
- Pauciarticular onset (60%) with involvement of four or fewer joints. Systemic features are absent but uveitis is *very common*.

Ocular features

- Chronic, frequently bilateral, non-granulomatous anterior uveitis.
- Patients positive for *antinuclear antibodies* are at increased risk of uveitis.

UVEITIS IN NON-INFECTIOUS SYSTEMIC DISEASES

Sarcoidosis

Systemic features

The two modes of presentation are:

- *Acute onset* with hilar adenopathy and erythema nodosum in *young* patients (good prognosis).
- *Insidious onset* with pulmonary fibrosis in *older* patients (guarded prognosis).

Diagnostic tests

- Chest X-rays are positive in 90%.
- Biopsy of lung, lacrimal gland, lymph nodes, tonsil, liver and conjunctiva.
- Bronchoalveolar lavage for T-lymphocytes.
- Kveim–Slitzbach skin test is positive in 80%.
- Angiotensin converting enzyme is increased in active disease.
- Calcium – hypercalciuria (common) and hypercalcaemia (rare).
- Gallium-67 scan is positive in active disease.

Ocular features

External

- Eyelids may show violaceous plaques (lupus pernio) and granulomata.
- Conjunctival granuloma may be suitable for biopsy.
- Lacrimal gland granuloma may cause dry eyes.
- Scleral granuloma (very rare).

Anterior uveitis

- Acute unilateral non-granulomatous in *young* patients with acute sarcoid.
- Chronic frequently bilateral granulomatous in *older* patients with chronic lung disease.

Posterior segment

- Vasculitis may be manifest as mild focal peripheral periphlebitis (most common), 'candlewax drippings', retinal branch vein occlusion and peripheral 'seafan' neovascularization.
- Retinal involvement consisting of small retinal and pre-retinal granulomata (Landers' sign) or rarely 'acute sarcoid retinopathy'.
- Choroid granulomata (rare).
- Optic nerve granulomata and disc neovascularization (rare).

Behçet's disease

Systemic features

The diagnostic requirements are at least three 'major' or two 'major' features and at least two 'minor' of the following features:

Four major features

1. Recurrent painful mouth ulcers.
2. Genital ulcers.
3. Skin lesions.
4. Uveitis.

Six minor features

1. Thrombophlebitis.
2. Arthritis.
3. Gastrointestinal lesions.
4. CNS lesions.
5. Cardiovascular lesions.
6. Positive family history

Ocular features

External (rare)

- Conjunctivitis.
- Episcleritis.
- Keratitis.

Anterior uveitis which is non-granulomatous, recurrent and which may be associated with a transient hypopyon.

Posterior segment

- Diffuse vascular leakage with retinal oedema and cystoid macular oedema.
- Periphlebitis which may cause venous occlusion and secondary neovascularization.
- Retinitis in the form of transient white necrotic infiltrates.
- Acute massive retinal exudation leading to necrosis and atrophy.

Vogt–Koyanagi–Harada syndrome

Systemic features

Cutaneous

- Vitiligo (patches of depigmentation).
- Alopecia.
- Poliosis (white eyelashes).

Neurological

- Headaches and neck stiffness.
- Encephalopathy.
- Auditory features (tinnitus, vertigo and deafness).
- CSF lymphocytosis.

Ocular features

- Chronic granulomatous anterior uveitis.
- Multifocal choroiditis.
- Exudative retinal detachment in Harada's disease.

UVEITIS IN CHRONIC SYSTEMIC INFECTIONS

Acquired syphilis

Systemic features

- *Primary* – chancre develops between 10 days and 10 weeks following sexual contact.
- *Secondary* – malaise, fever, lymphadenopathy, condylomata lata, mucous patches and meningitis (3–6 weeks after chancre).
- *Latent* – follows resolution of secondary syphilis.
- *Tertiary* – meningovasculitis, tabes dorsalis, generalized paralysis of the insane and gummata.

Diagnostic tests

- FTA-ABS is a specific test which remains positive throughout life.
- VDRL is a non-specific test which frequently becomes negative after treatment.
- MHA-TP.

Ocular features

- Acute anterior uveitis which may become chronic unless treated.
- Chorioretinitis (multifocal or diffuse).
- Neuroretinitis which may cause optic atrophy.

Management

- Lumbar puncture to rule out neurosyphilis.
- Systemic penicillin.

Tuberculosis

Systemic features

- Primary complex in the chest which consists of a Ghon focus and regional adenopathy.
- Postprimary due to re-infection.

Diagnostic tests

- Examination of sputum for bacilli.

- Chest X-ray.
- Tuberculin test.
- Isoniazid test for suspected ocular involvement (300 mg/day for 3 weeks).

Ocular features

- Granulomatous chronic anterior uveitis.
- Choroiditis.

Treatment

Systemic isoniazid, pyridoxine and rifampicin for 12 months.

UVEITIS IN PARASITIC INFESTATIONS

Toxoplasmosis

***Forms of* Toxoplasma gondii**

- *Oocyst* – spore excreted in cat faeces which may be ingested by children (pica).
- *Bradyzoite* – inactive form encysted in tissues which may be ingested by eating uncooked meat.
- *Tachyzoite* (trophozoite) – active form which causes tissue destruction. It may be transferred to the fetus via the placenta.

Clinical stages of toxoplasmosis

- *Acute* follows ingestion of oocytes which pass through the intestinal mucosa and become disseminated in tissues to form intracellular cysts.
- *Chronic* – inactive intracellular cysts (bradyzoites).
- *Recurrent* – when immunity is reduced the cysts rupture and cause tissue destruction.

Diagnostic tests

- Dye test (Sabin–Feldman).
- Indirect fluorescent antibody.
- Haemagglutination.
- Enzyme-linked immunosorbent assay (ELISA).

Systemic toxoplasmosis

Acute acquired

- Subclinical (most common).
- Febrile lymphadenitis.
- Influenza-like illness.
- Meningoencephalitis.
- Exanthematous.

Congenital

- Active at birth may cause death, severe damage, miscarriage and convulsions.
- Inactive at birth but may cause bilateral macular scars.

Recurrent toxoplasmic retinochoroiditis

Most common cause of a focal retinochoroiditis.

Signs

- Anterior segment is normal or it may show an anterior uveitis.
- Vitritis is frequently severe.
- Fundus usually shows a focal necrotizing retinitis adjacent to old inactive scar (satellite lesion) which heals within 1–4 months and leaves an atrophic scar surrounded by pigment.

Complications

- Direct involvement of the fovea, papillomacular bundle or optic nerve head.
- Indirect involvement of the fovea by cystoid macular oedema or macular pucker.
- Tractional retinal detachment (rare).

Indications for treatment

- Lesion threatening the macula or the optic nerve head.
- Very severe vitritis which might cause a tractional retinal detachment.

Drugs

- *Clindamycin* 300 mg four times a day for 3 weeks.
- *Sulphonamides* 2 g loading dose and then 1 g four times a day for 3–4 weeks.
- *Pyrimethamine* 100 mg loading dose and then 25 mg/day for 1 week.
- *Steroids* – systemic or posterior sub-Tenon's for very severe vision-threatening lesions.

Toxocariasis

Mode of human infestation

The ova of a roundworm of dogs (*Toxocara canis*) are shed in puppies faeces and accidentally ingested by a young child.

Systemic features (visceral larval migrans)

Occurs at about the age of 2 years with fever, hepatosplenomegaly, pneumonitis and convulsions but *no* eye involvement.

Ocular toxocariasis

Signs are invariably unilateral:

- *Chronic endophthalmitis* which presents between 2 and 9 years.
- *Posterior pole granuloma* which presents between 6 and 14 years.
- *Peripheral granuloma* which presents between 6 and 40 years.

Treatment by vitrectomy may be beneficial for endophthalmitis, cyclitic membranes and tractional retinal detachment.

UVEITIS IN VIRAL INFECTIONS

Herpes zoster anterior uveitis

Clinical features

Incidence – occurs in 50% of patients with herpes zoster ophthalmicus.

Signs – unilateral non-granulomatous acute anterior uveitis which becomes chronic unless treated.

Complications

- Large segmental iris atrophy in 20%.
- Secondary glaucoma due to trabeculitis in 10%.

Treatment

Topical steroids.

Acquired cytomegalovirus retinitis

Predispositions

- AIDS.
- Cytotoxic therapy for malignant disease.
- Long-term immunosuppressive therapy following organ transplantation.

Clinical features

Signs in chronological order:

- Cotton-wool spots.
- Peripheral, geographical, yellow–white, granular areas.
- Central haemorrhagic areas along vascular arcades.

Complications

- Retinal and optic atrophy.
- Retinal detachment.

Treatment

Intravenous antiviral agents.

UVEITIS IN FUNGAL INFECTIONS

Presumed ocular histoplasmosis syndrome

Mode of infection

Inhalation of *Histoplasma capsulatum.*

Ocular features

Signs are usually bilateral:

- Vitreous is *always clear*.
- Atrophic 'histo spots' in the mid-retinal periphery and the posterior pole.
- Peripapillary atrophy.
- Linear streaks of chorioretinal atrophy.
- Neovascular maculopathy may develop between the ages of 20 and 40 years.

Complications are disciform scarring secondary to neovascular maculopathy.

Treatment

Early subretinal neovascular membranes outside the foveal avascular zone can be treated by argon laser photocoagulation.

Candidiasis

Predispositions

- Drug addicts using infected needles.
- Patients with long-term indwelling catheters.
- Patients with impaired immunity (e.g. AIDS).

Clinical features

Signs are multifocal retinitis followed by extension into the vitreous to form 'cotton balls'.

Complications

- Vitreoretinal abscess.
- Retinal necrosis.
- Retinal detachment.

Treatment

- Oral 5-flucytosine and ketoconazole.
- Pars plana vitrectomy and intravitreal injection of amphotericin B in advanced cases.

IDIOPATHIC SPECIFIC UVEITIS ENTITIES

Fuchs' uveitis syndrome

Clinical features

Presentation of this *common* condition is usually between the second and fourth decades with unilateral cataract.

Signs are unilateral:

- Keratic precipitates (KPs) are small, round or stellate and scattered *throughout* the cornea.
- Anterior chamber shows a mild chronic anterior uveitis.
- Iris initially shows loss of crypts. Later diffuse atrophy leads to heterochromia. Koeppe nodules and mild rubeosis may be present but posterior synechiae are *always absent.*
- Anterior vitreous contains cells.
- Angle may show fine neovascularization with bleeding on paracentesis (Amsler's sign).

Complications

- Cataract (common).
- Secondary glaucoma (uncommon).

Treatment

None because steroids are ineffective, but watch for glaucoma.

Intermediate uveitis

Clinical features

Presentation of this *common* condition is usually between the second and third decades with either floaters or impaired vision due to cystoid macular oedema.

Signs are bilateral in 80% but frequently asymmetrical:

- KPs are small and few in number.
- Anterior chamber shows a mild chronic anterior uveitis.
- Vitreous shows cells, cotton balls and posterior detachment.
- Retina may show a mild peripheral vasculitis and 'snowbanking' of the inferior pars plana.

Complications

- Cystoid macular oedema (common).
- Cataract (uncommon).
- Cyclitic membrane formation (rare).
- Tractional retinal detachment (rare).

Treatment

- Posterior sub-Tenon's injection of triamcinolone when visual acuity is less than 6/9 due to cystoid macular oedema.
- Acetazolamide may also be helpful for cystoid macular oedema.
- Cyclocryotherapy of the inferior pars plana is of doubtful benefit.

Acute anterior uveitis in young adults

Clinical features

Presentation of this *common* condition is typically in young men who are HLA-B27 positive.

Signs are recurrent, unilateral, non-granulomatous, acute anterior uveitis.

Complications

- Pigment on anterior lens capsule.
- Posterior synechiae.

Treatment

Topical or anterior sub-Tenon's steroids.

Sympathetic uveitis (ophthalmitis)

Clinical features

Presentation of this *very rare* condition is a few weeks after penetrating ocular trauma to one eye which frequently involves the uvea.

Signs are bilateral chronic granulomatous panuveitis.

Complications

- Cataract (common).
- Phthisis bulbi (common).

Treatment

- Prophylaxis – uveitis can usually be prevented by enucleating the 'exciting' traumatized eye within 2 weeks of the injury.
- Cytotoxic agents may be required in severe steroid-resistant cases.

Acute posterior multifocal placoid pigment epitheliopathy

Clinical features

Presentation of this *uncommon* condition is usually in young adults with a subacute loss of vision in one eye and then a few days later in the other eye.

Signs are multiple, deep, placoid, cream-coloured lesions at the equatorial region and posterior pole.

Complications are residual changes in the retinal pigment epithelium but usually good vision.

Treatment

None effective.

Serpiginous choroidopathy

Clinical features

Presentation of this *rare* condition is between the fifth and sixth

decades with a gradual loss of vision first in one eye and then frequently in the other eye after several months.

Signs are cream-coloured choroidal opacities which spread outwards from the disc leaving behind permanent atrophic 'punched-out' areas.

Complications are permanent impairment of visual acuity due to macular involvement.

Treatment

None effective.

Birdshot retinochoroidopathy

Clinical features

Presentation of this *very rare* condition is between the fourth and sixth decades with bilateral floaters or reduced visual acuity due to cystoid macular oedema.

Signs are scattered cream–yellow hypopigmented spots in the fundus.

Complications

- Cataract.
- Cystoid macular oedema.

Treatment

The response to steroids is usually poor.

Acute retinal necrosis

Clinical features

Presentation of this *very rare* condition is at any age with periorbital pain and visual loss.

Signs (bilateral in 30–50%) in chronological order:

- Multifocal yellow–white patches with arteriolar sheathing.
- Coalescence of patches.

- Necrotizing retinitis.
- Retinal hole formation.

Complications are retinal detachment which is very difficult to treat.

Treatment

Prophylactic laser photocoagulation in early cases.

TREATMENT OF UVEITIS

Mydriatics

Indications

- To give comfort.
- To prevent posterior synechiae.
- To break down posterior synechiae.

Preparations

See Table 3.

Table 3

Drug (concn)	*Mydriasis – maximal recovery*		*Cycloplegia – maximal recovery*	
	(min)	*(days)*	*(hours)*	*(days)*
Atropine (1%)	40	10+	6	14
Hyoscine (0.25–0.5%)	30	7	1	7
Homatropine (1–5%)	60	3	1	3
Cyclopentolate (0.5–1%)	60	1	1	1
Tropicamide (0.5–1%)	40	0.25	0.5	0.25
Phenylephrine (10%)	20	0.25	Nil	–

Topical steroids

Indications

Anterior uveitis.

Preparations

See Table 4.

Table 4

Drug	*Concentration*	*Drops*	*Ointment*	*'Potency' in uveitis*
Prednisolone acetate	1%	+		+++++
Dexamethasone	0.1%	+		++++
Betamethasone	0.1%	+	+	+++
Prednisolone phosphate	0.5%	+		+++
Fluorometholone	0.1%	+		++
Clobetasone	0.1%	+		+

Complications

- Glaucoma in 'steroid' reactors (see Chapter 7).
- Cataract with long-term use (common).
- Enhancement of herpes simplex infection (uncommon).
- Corneal melting (rare).
- Systemic absorption, usually of little clinical significance.

Periocular steroid injections

Indications

Anterior sub-Tenon

- Very severe anterior uveitis.
- Anterior uveitis resistant to drops.
- Poor patient compliance.
- Following intraocular surgery.

Posterior sub-Tenon

- Intermediate uveitis.
- Some cases of posterior uveitis.

Preparations

See Table 5.

Table 5

Short-acting (1 day)	*Long-acting (several weeks)*
Betamethasone 4 mg/ml	Methylprednisolone acetate (Depo-medrone) 40 mg/ml
Dexamethasone 4 mg/ml	Triamcinolone acetonide (Kenalog) 40 mg/ml

Systemic steroids

Indications

Uveitis resistant to periocular steroid injections.

Complications

Short-term therapy

- Peptic ulceration (common).
- Mental changes (uncommon).
- Aseptic necrosis of head of femur (rare).
- Hyperosmolar, hyperglycaemic, non-ketotic coma (very rare)

Long-term therapy

- Cataract (common).
- Cushingoid state (common).
- Limitation of growth in children (common).
- Reactivation of tuberculosis (now rare).

Cytotoxic agents

Indications

- Potentially blinding (usually bilateral), steroid-resistant uveitis.
- Intolerable side effects from systemic steroid therapy.

Preparations

- Chlorambucil.
- Azathioprine.
- Cyclophosphamide.

Complications

- Bone marrow depression.
- Gastrointestinal ulceration.
- Stomatitis.
- Liver damage.
- Sterility.
- Alopecia.
- Neoplasia.
- Haemorrhagic cystitis.
- Nausea and vomiting.
- Genetic damage.

Chapter 7

The glaucomas

CLASSIFICATION

According to state of drainage angle

- Open angle
- Angle closure

According to presence of associated factors

- Primary
- Secondary

According to age of onset

- Congential
- Infantile
- Juvenile
- Adult

Primary glaucomas

- Open angle
- Angle closure
- Congenital (developmental)

Secondary glaucomas

- Open angle
- Angle closure

OPEN-ANGLE GLAUCOMAS IN ADULTS

Primary open-angle glaucoma (POAG)

Definition

A chronic, usually bilateral disease with an insidious onset characterized by an intraocular pressure (IOP) greater than 21 mmHg, normal angles and glaucomatous damage.

Pathogenesis

Increased resistance to aqueous outflow in the drainage channels.

Prevalence

- *Most prevalent* of all glaucomas affecting 1 in 200 of the general population over the age of 40 years.
- Affects both sexes equally but is more common in Blacks than in Whites.

Inheritance

First-degree relatives are at increased risk. In *siblings* the risk is about 10% and in *offspring* it is about 4%.

Steroid responsiveness

Prevalence

Based on the rise in IOP induced by 6-week course of betamethasone drops the population can be divided into three groups:

1. *High responders* show a marked IOP increase to over 30 mmHg (5% of general population).
2. *Moderate responders* show a moderate IOP increase of 22–30 mmHg (35% of general population).
3. *Non-responders* show no change in IOP (60% of normal population).

Patients with POAG, high myopia and diabetes have an increased incidence of steroid responsiveness.

Strong steroids such as dexamethasone, betamethasone and prednisolone are equipotent in ability to elevate IOP.

Weak steroids such as fluorometholone and clobetasone have less propensity to elevate IOP.

Ocular associations of POAG

- High myopia.
- Central retinal vein occlusion.
- Rhegmatogenous retinal detachment.
- Fuchs' endothelial dystrophy.
- Retinitis pigmentosa.

Systemic associations of POAG

- Diabetes.
- Low levels of protein-bound iodine.
- Untreated hypertension.

Clinical features

Symptoms – initially asymptomatic until it has caused a significant loss of visual field.

Progression of glaucomatous cupping

- *Early* cupping takes the form of either round concentric or vertical enlargement of the optic cup.
- *Advanced* cupping is characterized by total loss of inferior and superior disc tissue with nasal displacement of the central blood vessels.
- *Total* cupping is characterized by atrophy of both the temporal and the nasal rim.

Progression of visual field defects

- *Early* defects are scotomata in Bjerrum's area between 10° and 20° of fixation or isolated paracentral nasal scotomata.
- *Bjerrum scotomata* then elongate circumferentially.
- *Nasal step* or a *temporal wedge* is usually associated with other defects.
- *Late* defects are arcuate due to coalescence of scotomata in Bjerrum's area.

- *Peripheral breakthrough* then occurs followed by a double arcuate (ring) scotoma due to joining together in opposite halves of the visual field.
- *Terminal* defects consist of a residual central island due to peripheral and central spread.

Principles of treatment

Basic rules of drug treatment:

- Initial therapy is usually medical.
- Use the lowest concentration of the drug as infrequently as possible.
- Choose the drug with the fewest side effects.

Initial medical therapy is usually with either a β-blocker or a sympathomimetic.

Subsequent medical therapy if response is unsatisfactory:

- Increasing the strength is unlikely to have a profound effect.
- Stop the initial drug and substitute another.
- Add another drug: sympathomimetic to a β-blocker is usually ineffective; β-blocker to a sympathomimetic may be effective; pilocarpine to a sympathomimetic and/or a β-blocker is usually effective; carbonic anhydrase inhibitors only as short-term therapy.

Indications for argon laser trabeculoplasty (ALT)

- Uncontrolled IOP despite maximal tolerated medical therapy.
- Young patients (<55 years) uncontrolled with sympathomimetics or β-blockers because miotics are poorly tolerated.
- Poor compliance.

Indications for filtration surgery

- Failed ALT.
- Unsuitable for ALT.
- Advanced disease requiring a very low IOP.

Low tension glaucoma

Clinical features

These are similar to POAG except that the IOP is consistently under 22 mmHg.

Pathogenesis

Low vascular perfusion pressure which makes the optic disc more susceptible to damage.

Treatment

- *Non-progressive type* may require no treatment.
- *Progressive type* – reduce the IOP to less than 12 mmHg to prevent further damage. This is best achieved by filtration surgery.

Ocular hypertension

Clinical features

These are similar to POAG except that damage is absent.

Treatment

This depends on the level of IOP and the presence of associated risk factors as follows:

- IOP >30 mmHg – treat.
- IOP <30 mmHg: (1) if risk factors *present* – treat; (2) if risk factors *absent* – do not treat but follow carefully.
- Rising IOP – carefully monitor discs and fields.
- POAG in fellow eye – treat even if IOP <30 mmHg.
- Family history of POAG – treat even if IOP <30 mmHg.
- High myopia or systemic vascular disease – probably treat.

Pseudoexfoliative glaucoma

Pathogenesis

Trabecular open-angle glaucoma due to blockage of the trabeculum by pseudoexfoliative material.

Clinical features

Symptoms

This *common* condition affects the *elderly* and has the same symptoms as POAG.

Signs at presentation are bilateral in 30% of cases:

- Lens shows a central disc, a peripheral band and a clear zone in between.
- Iris shows pseudoexfoliative material on the pupillary border, defects in the pupillary ruff, pigment granules on the iris surface and sphincter atrophy giving rise to 'moth-eaten' transillumination defects.
- Cornea shows deposits of pseudoexfoliative material and pigment.
- Gonioscopy shows hyperpigmentation of the trabeculum most pronounced inferiorly; scalloped pigment band running onto or anterior to Schwalbe's line (*Sampaolesi's line*); and 'dandruff-like' deposits of pseudoexfoliative material inferiorly.

Treatment

The same as for POAG but the prognosis is less good.

Pigmentary glaucoma

Pathogenesis

Trabecular open-angle glaucoma due to blockage of the trabeculum by pigment derived from the posterior iris surface.

Clinical features

Symptoms

This *uncommon* condition which typically affects *young myopic males* has the same symptoms as POAG.

Signs are always bilateral:

- Corneal endothelium shows a vertical spindle-shaped area of fine pigment granules (*Krukenberg's spindle*).
- Anterior chamber is excessively deep in the midperiphery.

- Iris shows fine pigment granules on its surface and midperipheral slit-like transillumination defects.
- Gonioscopy shows hyperpigmentation most marked over the posterior trabeculum.

Treatment

The same as for POAG.

PRIMARY ANGLE-CLOSURE GLAUCOMA

Definition

Primary angle-closure glaucoma (PACG) is a condition in which aqueous outflow is obstructed solely by closure of the angle by the peripheral iris.

Prevalence

- Affects 1 in 1000 of the general population over the age of 40 years.
- Females are more commonly affected by a ratio of 4:1.

Inheritance

The anatomical predisposition is inherited.

Classification

- Latent
- Intermittent (subacute)
- Acute
- Chronic
- Absolute

Latent PACG

Clinical features

Symptoms are absent.

Signs

- IOP is normal.
- Anterior chamber is shallow.
- Iris–lens diaphragm has a convex shape.
- Iris is in close proximity to the peripheral cornea.
- Angle is narrow and capable of closure.

Treatment

Laser iridotomy if the fellow eye has had acute or intermittent PACG.

Intermittent PACG

Mechanism

Rapid partial closure and re-opening of the angle precipitated by mydriasis.

Clinical features

Symptoms are transient blurring and haloes (blue end of the spectrum nearest the light source) due to corneal epithelial oedema associated with eye ache or frontal headache.

Signs are epithelial oedema and a semidilated pupil.

Treatment

- Initially 2% pilocarpine every 5 min to the *affected eye* and prophylactic 1% pilocarpine four times a day to the *fellow eye*.
- Laser iridotomy to *both* eyes once IOP is normal.

Acute congestive PACG

Mechanism

A sudden and total closure of the angle.

Clinical features

Symptoms are an acute onset of unilateral visual loss and pain.

Signs

- IOP is between 50 and 80 mmHg.
- Ciliary flush
- Epithelial oedema with vesicles.
- Shallow anterior chamber with peripheral iridocorneal contact.
- Aqueous cells and flare.
- Vertically oval and unreactive pupil.
- Dilated and congested iris blood vessels.
- Completely closed angle.

Differential diagnosis

- Secondary angle closure by an intumescent (swollen) lens.
- Phacolytic glaucoma.
- Neovascular glaucoma.
- Galucomatocyclitic crisis.

Treatment

Medical

- Acetazolamide 500 mg intravenously and then 250 mg four times a day orally.
- Timolol 0.5% twice a day and dexamethasone four times a day.
- Pilocarpine 2% four times a day when IOP is reduced and to the fellow eye.

Laser iridotomy when the congestion has subsided and the cornea is clear and also to the fellow eye prophylactically.

Filtration surgery is required subsequently in about 25% of cases.

Chronic PACG

Mechanisms

- *Group 1* is due to a gradual progressive (creeping) angle closure.
- *Group 2* follows a subacute attack of PACG.
- *Group 3* is due to a combination of POAG with a narrow angle and the long-term use of miotics.

Clinical features

These are similar to POAG with a variable amount of angle closure.

Treatment

- Group 1 with iridectomy and topical therapy. Filtration surgery may be required in some cases.
- *Group 2* with medical therapy (iridectomy has already been performed).
- *Group 3* with iridectomy and medical therapy.

Absolute PACG

This is the end-stage of PACG in which the eye is completely blind.

NEOVASCULAR GLAUCOMA

Causes

Retinal hypoxia secondary to the following.

Ischaemic central retinal vein occlusion (common) (see Chapter 10).

Diabetes mellitus – particularly following vitrectomy.

Miscellaneous (rare):

- Carotid obstructive disease.
- Carotid–cavernous fistula.
- Intraocular tumours.
- Chronic intraocular inflammation.
- Retinopathy of prematurity.

Classification

- Secondary open-angle glaucoma
- Secondary synechial angle-closure glaucoma

Secondary open-angle glaucoma

Mechanism

Pre-trabecular glaucoma due to obstruction of the angle by a fibrovascular membrane.

Clinical features

Gradual progressive rise of IOP associated with rubeosis iridis.

Treatment

- Medical as for POAG.
- Combined with panretinal argon laser photocoagulation (PRP).

Secondary synechial angle-closure glaucoma

Mechanism

Angle closure by contracting fibrovascular tissue.

Clinical features

Symptoms are pain and severe visual impairment.

Signs

- Very high IOP.
- Congestion and redness.
- Corneal epithelial oedema.
- Aqueous flare.
- Severe rubeosis iridis and ectropion uveae.

Treatment

- *PRP* if the media are clear but the success rate is very low.
- *Peripheral retinal cryotherapy* if the media are hazy.
- *Medical* with atropine and steroid drops to reduce inflammation and pain. β-Blockers and carbonic anhydrase inhibitors are of limited benefit.
- *Surgery* with artificial filtering shunts.

- *Cyclodestruction* by cyclocryotherapy or trans-scleral YAG (yttrium–aluminium–garnet) laser.
- *Retrobulbar alcohol* injection.
- *Enucleation* as a last resort.

INFLAMMATORY GLAUCOMAS

Classification

- Pupil block with secondary angle closure
- Secondary synechial angle closure without pupil block
- Secondary open-angle glaucoma

Pupil block with secondary angle closure

Mechanism

Pupil block by seclusio pupillae (360° iridolenticular adhesions).

Clinical features

- Seclusio pupillae and iris bombé.
- Shallow anterior chamber.
- Gonioscopy shows variable angle closure by iridotrabecular contact.

Treatment

This is initially medical followed by a peripheral iridectomy.

Secondary synechial angle closure without pupil block

Mechanism

Contraction of inflammatory debris in the angle.

Clinical features

- Gradual, frequently asymptomatic rise in IOP.
- Gonioscopy shows variable angle closure by peripheral anterior synechiae.

Treatment

- Initially medical
- Surgery may be required in resistant cases.

Secondary open-angle glaucoma

Mechanism

Trabecular block either by proteins, inflammatory cells and debris or by trabecular oedema and scarring.

Clinical features

Elevated IOP associated with an open angle.

Treatment

- In *acute* anterior uveitis this is medical until the inflammation subsides.
- In *chronic* anterior uveitis this is the same as for secondary synechial angle-closure glaucoma.

UNCOMMON ACQUIRED SECONDARY GLAUCOMAS

Phacolytic (lens protein) glaucoma

Mechanism

Trabecular open-angle glaucoma due to blockage of the trabeculum by macrophages containing phagocytosed soluble proteins that have leaked through an intact capsule of a hypermature cataract.

Clinical features

An acute unilateral rise in IOP in an eye with a hypermature cataract and a deep anterior chamber.

Treatment

This is initially medical followed by cataract extraction.

Pupil block glaucoma by an intumescent lens

Clinical features

These are similar to acute PACG.

Treatment

This is the same as for acute PACG followed by lens extraction.

Pupil block glaucoma by lens dislocation into the anterior chamber

Causes

- Blunt ocular trauma.
- Small lens (microspherophakia).
- Weak zonules.

Clinical features

These are an acute severe unilateral rise of IOP.

Treatment

- Hyperosmotic agents to reduce IOP.
- Followed either by an attempt to reposition the lens in the posterior chamber by dilating the pupil and placing the patient in the supine position, or by lens extraction.

Angle-recession (cleavage) glaucoma

Mechanism

Trabecular scarring following blunt ocular trauma that has caused a laceration of the anterior face of the ciliary body.

Clinical features

- Gradual rise in IOP usually several years after the initial injury.
- Gonioscopy shows a scarred angle with pigment within its recess.

Treatment

This is the same as for POAG.

Iridocorneal endothelial syndromes

Mechanism

Secondary synechial angle closure by contraction of abnormal corneal endothelial cells.

Clinical features

The condition typically affects one eye of a young or middle-aged woman. The three types are the following.

Essential iris atrophy

- Displacement of the pupil towards an area of peripheral anterior synechiae.
- Ectropion uveae.
- Stromal atrophy with iris holes.

Iris naevus (Cogan–Reese) syndrome

- Diffuse naevus covering the anterior iris.
- Iris nodules may be present.

Chandler's syndrome is intermediate between essential iris atrophy and the iris naevus syndrome.

Treatment

This is very difficult and surgery is usually required.

PRIMARY CONGENITAL GLAUCOMA

Definition

A condition in which elevation of IOP is caused by a congenital maldevelopment of the trabeculum and the iridotrabecular junction (isolated trabeculodysgenesis) which is not associated with other major ocular anomalies.

Prevalence

- This *very rare* condition affects 1 in 10 000 births.
- Boys are more commonly affected than girls by a ratio of 3:2.

Inheritance

Recessive with incomplete penetrance.

Clinical features

These depend on the age of onset. Both eyes are affected in 75% of cases but the severity of involvement is frequently asymmetrical.

Age of onset

- *True congenital glaucoma* (40%) presents at birth with large eyes (buphthalmos).
- *Infantile glaucoma* (55%) presents prior to the age of 2 years.
- *Juvenile glaucoma* (5%) presents between the ages of 2 and 16 years.

Signs

- Epithelial corneal oedema giving rise to lacrimation, photophobia and blepharospasm.
- Buphthalmos if the IOP becomes raised prior to the age of 3 years.
- Breaks in Descemet's membrane which may lead to permanent corneal scarring; *Haab's striae* are healed breaks in Descemet's membrane.
- Glaucomatous cupping which may regress when the IOP is normalized.
- Gonioscopy shows absence of the angle recess with the iris inserted directly into the trabeculum.

Differential diagnosis

Cloudy cornea at birth

- Trauma.
- Intrauterine rubella.
- Peter's anomaly (see later).
- Metabolic disorders (mucopolysaccharidoses, lipidosis and cystinosis).

Large cornea at birth

- Megalocornea.
- Very high myopia.

Secondary glaucomas

- Tumours (retinoblastoma and juvenile xanthogranuloma).
- Persistent hyperplastic primary vitreous.
- Advanced retinopathy of prematurity.

Treatment

- Goniotomy is the procedure of choice with an 80% success rate.
- Trabeculotomy is useful if the cornea is cloudy or if goniotomy fails.
- Treat refractive errors and amblyopia.

GLAUCOMA AND IRIDOCORNEALDYSGENESIS

Definition

A group of congenital overlapping disorders involving the cornea and the iris, some of which are associated with glaucoma.

Classification

- Posterior embryotoxon
- Rieger's anomaly and Rieger's syndrome
- Peters' anomaly
- Aniridia

Posterior embryotoxon

This *innocuous* anomaly which consists of a prominent anteriorly displaced Schwalbe's line is present in 10% of normals.

Axenfeld's anomaly

This *rare* condition which is very occasionally associated with glaucoma consists of strands of peripheral iris tissue attached to a posterior embryotoxon.

Rieger's anomaly and Rieger's syndrome

Inheritance

Dominant.

Ocular features of Rieger's anomaly

This *very rare* bilateral condition is characterized by:

- Posterior embryotoxon and occasional detachment of Schwalbe's line.
- Iris shows stromal hypoplasia, ectropion uveae, full-thickness holes (pseudopolycoria) and a displaced pupil (corectopia).
- Gonioscopy in mild cases is similar to Axenfeld's anomaly. In severe cases leaves of iris stroma extend anteriorly to Schwalbe's line.
- *Glaucoma* during early childhood develops in 50% of cases.

Systemic features of Rieger's syndrome.

- *Dental anomalies* are small teeth (microdontia) and few teeth (hypodontia).
- *Facial anomalies* are hypoplasia of the maxilla, a broad flat nasal bridge, lateral displacement of the medial canthi (telecanthus) and an increased interorbital distance (hypertelorism).

Peters' anomaly

Inheritance

Dominant.

Clinical features

This *very rare* anomaly which is bilateral in 80% of cases is characterized by:

- Corneal opacity associated with posterior stromal defects, absence of Descemet's membrane and iris adhesions.
- *Glaucoma* due to an associated angle anomaly occurs in 50% of cases.

Aniridia

Inheritance

This is dominant in two-thirds of cases and in the remaining third sporadic.

Ocular features

This *rare* bilateral condition is characterized by:

- Variable amount of aniridia.
- Corneal pannus and epibulbar dermoids.
- Cataract and lens subluxation.
- Foveal and optic nerve hypoplasia.
- *Glaucoma* due to secondary angle closure by rudimentary iris tissue develops during late childhood in 50% of cases.

Systemic associations

- Wilms' tumour develops in 20% of sporadic cases prior to age 2 years.
- Miscellaneous include mental handicap, digital anomalies and anomalies of the genitourinary system.

GLAUCOMA IN THE PHACOMATOSES

Sturge–Weber syndrome

Inheritance

Not inherited.

Systemic features

This *rare* condition is characterized by:

- Facial skin capillary haemangioma (naevus flammeus) which is present at birth and involves the area of distribution of the first and second divisions of the fifth nerve. It may be associated with hypertrophy of the involved area.
- Ipsilateral meningeal haemangioma frequently involving the parieto-occipital region which may cause epilepsy, hemiparesis, hemianopia and mental deficiency.

Ocular features

- Diffuse unilateral choroidal haemangioma.
- Haemangiomas of the episclera, iris and ciliary body.
- *Glaucoma* due to either an angle anomaly or raised episcleral venous pressure occurs in 30% of cases. In 60% of cases the rise in IOP develops within the first 2 years of life and causes buphthalmos.

Neurofibromatosis (von Recklinghausen's disease)

Inheritance

Dominant.

Systemic features

This *uncommon* condition is characterized by:

- CNS tumours of the brain, spinal cord, meninges, cranial nerves, peripheral nerves, sympathetic nerves and acoustic neuromas.
- Phaeochromocytoma in a few cases.
- Congenital bony defects involving the greater wing of the sphenoid bone which may cause a pulsating proptosis, and occasional defects in the vertebrae and long bones.
- Skin lesions are café-au-lait spots, fibromata mollusca and plexiform neurofibromata.
- Facial hemihypertrophy

Ocular features

- Bilateral iris hamartomas (Lisch nodules).
- Optic nerve glioma.
- Congenital ectropion uvea.
- Choroidal naevi.
- Proptosis due to a defect in the sphenoid bone, optic nerve glioma or an orbital neurofibroma.
- *Glaucoma* is uncommon. It is usually present at birth and is associated with an ipsilateral lid neurofibroma.

ANTI-GLAUCOMA DRUGS

β-Blockers

Mode of action is suppression of aqueous secretion.

Clinical uses

- POAG – first choice in absence of systemic contraindications.
- Secondary glaucomas – useful in most.

Systemic side effects

- Bradycardia due to β_1-adrenergic blockade.
- Brochospasm due to β_2-adrenergic blockade.
- Miscellaneous – fatigue, depression, headache, nausea, dizziness, decreased libido, skin rashes and aggravation of myasthenia gravis.

Local side effects

- Stinging is usually mild.
- Blepharoconjunctivitis occurs occasionally with long-term use.
- Punctate keratitis is rare and innocuous.
- Dry eyes are rare and mild.
- Corneal anaesthesia is very rare and mild.
- Mydriasis occurs when combined with sympathomimetics.

Timolol (Timoptol, Timoptic)

- Concentrations: 0.25%, 0.5%.
- Pharmacology: non-selective β_1- and β_2-blocker.
- Action: 30 min (onset), 2 hours (peak), up to 24 hours (duration).
- Administration: twice a day.
- Efficacy: very good.

Betaxolol (Betoptic)

- Concentration: 0.5%.
- Pharmacology: relative cardioselective β_1-blocker.
- Action: 30 min (onset), 2 hours (peak), up to 12 hours (duration).

- Administration: twice a day.
- Efficacy: slightly less than timolol.

Levobunolol (Betagan)

- Concentration: 0.5%.
- Pharmacology: same as timolol.
- Action: 1 hour (onset), 2–6 hours (peak), up to 24 hours (duration).
- Administration: once daily use may suffice.
- Efficacy: same as timolol.

Carteolol (Teoptic)

- Concentrations: 1%, 2%.
- Pharmacology: non-selective β-blocker with intrinsic sympathomimetic activity. It has more selective action on the eye than on the cardiopulmonary system and may induce less bradycardia than timolol.
- Action: same as timolol.
- Administration: twice a day.
- Efficacy: same as timolol.

Metipranolol (Glauline)

- Concentrations: 0.1%, 0.3%, 0.6%.
- Properties: same as timolol.

Sympathomimetics

Mode of action is by a combination of reduced aqueous secretion and improved aqueous outflow.

Clinical uses

- POAG – first choice in the presence of systemic contraindications to β-blockers.
- Secondary glaucomas – useful in most.

Systemic side effects

- Headaches are occasionally troublesome.
- Palpitations due to β_1-agonist action.
- Hypertension is not a problem.
- Bronchodilatation may be advantageous in asthmatics.
- Nervousness is very rare.

Local side effects

- Stinging is common.
- Rebound conjunctival injection is common.
- Allergic blepharoconjunctivitis is uncommon.
- Adrenochrome conjunctival deposits are common with very long-term use.
- Mydriasis may be dangerous in eyes with narrow angles.
- Aphakic cystoid macular oedema may occur with the 2% concentration.

Adrenaline (Eppy, Simplene, Epifrin, Isopto Epinal)

- Concentrations: 0.5% 1%, 2%.
- Action: 1 hour (onset), 12–24 hours (duration).
- Administration: twice a day.
- Efficacy: less than pilocarpine and β-blockers

Dipivefrin (Propine)

- Concentration: 0.1%.
- Pharmacology: pro-drug converted to adrenaline after absorption.
- Action: same as adrenaline.
- Administration: twice a day.
- Efficacy: same as 1% adrenaline.
- Local side effects: less than adrenaline.

Miotics

Mode of action

- In POAG increase outflow by widening intertrabecular pores.

- In PACG pull iris away from trabeculum by inducing miosis.

Clinical uses

- POAG – second choice.
- PACG – first choice.
- Secondary glaucomas – may be counterproductive in some.

Systemic side effects are seldom of clinical significance:

- Bradycardia.
- Increased sweating.
- Diarrhoea.
- Increased salivation.
- Anxiety.
- Scoline apnoea is the most serious complication.

Local side effects

- Reduction of night vision and visual field.
- Reduction of visual acuity in the presence of axial lens opacities.
- Pupil block in eyes with narrow angles.
- Myopia may be a problem in young patients.
- Frontal headache may occur at the start of therapy.
- Retinal detachment is very rare.
- Anterior subcapsular cataract may develop with long-term use of long-acting cholinesterase inhibitors.
- Iris cysts may develop with long-acting cholinesterase inhibitors. They can be prevented by the simultaneous use of 2.5% phenylephrine or 1% adrenaline twice a day.

Pilocarpine (Isopto Carpine, Sno Pilo)

- Concentrations: 1%, 2%, 3%, 4%.
- Pharmacology: parasympathomimetic muscarinic agonist.
- Action: 20 min (onset), 2 hours (peak), up to 4 hours (duration).
- Administration: four times a day when used alone, twice a day may suffice when combined with timolol.
- Efficacy: same as timolol, greater than adrenaline.

Carbachol (Isopto Carbachol)

- Concentrations: 0.75%, 3%.
- Pharmacology: parasympathomimetic muscarinic agonist and a weak cholinesterase inhibitor.
- Action: 40 min (onset), up to 12 hours (duration).
- Administration: three time a day.
- Efficacy: same as pilocarpine.

Ecothiopate iodide (Phospholine Iodide)

- Concentrations: 0.03%, 0.06%, 0.125%, 0.25%.
- Pharmacology: long-acting cholinesterase inhibitor.
- Action: 2 hours (onset), up to 24 hours (duration).
- Administration: once or twice a day.
- Efficacy: excellent in POAG.

Demecarium bromide (Humorosol, Tosmilen)

- Concentrations: 0.125%, 0.25%.
- Properties: same as ecothiopate iodide.

Carbonic anhydrase inhibitors

Mode of action is suppression of aqueous secretion.

Clinical uses

- Short term in acute PACG.
- Long term in other glaucomas with a high risk of visual loss.

Systemic side effects

- Paraesthesia.
- Malaise.
- Fatigue.
- Depression.
- Gastric irritation.
- Diarrhoea.
- Nausea.

- Stevens–Johnson syndrome.
- Renal stones.
- Bone marrow depression.

Acetazolamide (Diamox) tablets

- Tablets: 250 mg.
- Dose: 250–1000 mg daily in divided doses.
- Action: 1 hour (onset), 4 hours (peak), 6–12 hours (duration).

Acetazolamide slow release capsules (Diamox Sustets)

- Capsules: 500 mg
- Dose: 500 mg daily or twice a day.
- Action: up to 24 hours.

Acetazolamide parenteral

- Powder: 500 mg vials.
- Dose: 500 mg intramuscular or intravenous.
- Action: immediate (onset), 30 min (peak), 4 hours (duration).

Dichlorphenamide (Daranide)

- Tablets: 50 mg
- Dose: 50–100 mg two or three times a day.
- Action: 1 hour (onset), 3 hours (peak), 6–12 hours (duration).

Methazolamide (Neptazane)

- Tablets: 50 mg and 25 mg.
- Dose: 50–100 mg twice a day.
- Action: 3 hours (onset), 6 hours (peak), 10–18 hours (duration).

Osmotic agents

Mode of action is by withdrawing water from the vitreous due to an increase in the osmotic gradient between blood and vitreous.

Clinical uses are for an acute and very severe rise in IOP which is unresponsive to other measures.

Side effects

- Cardiovascular overload.
- Urinary retention.
- Headache and backache.
- Nausea.
- Mental confusion.

Oral agents

Glycerol has a sweet and sickening taste and must therefore be mixed with pure lemon juice. It is metabolized to glucose.

Isosorbide has a minty taste. Because it is metabolically inert it can safely be given to diabetics:

- Dose: 1–2 g/kg body weight or 2–4 ml/kg body weight of 50% solution.
- Action: 1 hour (peak), 3 hours (duration).

Intravenous agents

Mannitol is the most frequently used:

- Dose: 1–2 g/kg body weight or 5–10 ml/kg body weight of 20% solution in water.
- Administration: over 20–30 min via a drip with not more than 60 drops/min.
- Action: 30 min (peak), 6 hours (duration).

LASER THERAPY OF GLAUCOMA

Argon laser trabeculoplasty

Mode of action of argon laser trabeculoplasty (ALT) is increased aqueous outflow due to widening of the intertrabecular spaces.

Indications

- Adult open-angle glaucomas uncontrolled despite maximal tolerated medical therapy.
- Occasionally as primary therapy in patients with poor compliance to medical therapy.

Contraindications

- Poor view of trabeculum.
- When it is known to be of no value.

Technique

- *Laser settings* are spot size of 50 μm, duration of 0.1 second, and initial power of 700 mW.
- *Number of burns* is 100 for 360° in one or two sessions.
- *Location of burns* is at the junction of the pigmented with the non-pigmented trabeculum.

Complications

During treatment

- Peripheral anterior synechiae if applied too posteriorly.
- Small haemorrhages if a blood vessel is hit.

After treatment

- Increase in IOP.
- Iritis.

Results

In POAG

- *Early* – 75% have an average fall of IOP of 8 mmHg.
- *Late* – incidence of subsequent failure is 25% after 1 year and 10% per year thereafter.

In other glaucomas

- Pseudoexfoliative glaucoma is similar to POAG.
- Pigmentary glaucoma is similar to POAG in older patients but worse in young patients.
- Secondary glaucoma is very poor.
- Paediatric glaucoma is extremely poor.

Nd:YAG laser iridotomy

Indications

- PACG – acute, intermittent and chronic.
- Fellow eyes with acute PACG in the opposite eye.
- Secondary angle closure with pupil block.
- POAG with narrow angles.

Contraindications

- Corneal clouding that impairs accurate focusing.
- Peripheral iris is very close to the cornea because of danger of endothelial damage.

Technique

- *Laser settings* vary according to the machine.
- *Location of burns* is as peripheral as possible to avoid lens damage.

Complications

- Bleeding which is usually mild and transient.
- Corneal damage is usually innocuous.
- Increase in IOP.
- Iritis.
- Localized lens opacities.

GLAUCOMA SURGERY

Peripheral iridectomy

Indications are prevention or relief of pupil block glaucoma if a laser is either unavailable or laser iridotomy is inappropriate.

Surgical steps

1. *Incision* – make a vertical groove in the cornea 1 mm from limbus and deepen it until the anterior chamber is entered.

2. *Prolapsing of iris* – exert pressure on posterior lip of the incision with an iris repositor.
3. *Iridectomy* – grasp the iris with Colibri forceps and excise apex of the tented up iris with one cut.
4. *Iris reposition* – stroke the peripheral cornea radially with an iris repositor.

Complications

- Hyphaema due to excessive pulling on the iris.
- Cataract caused by instruments introduced into the anterior chamber.
- Pupil block caused either by an incomplete iridectomy or by closure of the iridectomy.
- Uniocular diplopia because the iridectomy is not covered by the upper eyelid.
- Malignant glaucoma (very rare).
- Endophthalmitis (very rare).

Trabeculectomy

Indications are controlled adult glaucomas despite maximal tolerated therapy and ALT.

Surgical steps

1. *Conjunctival flap* based at the limbus or fornix.
2. *Scleral flap* 3 mm × 4 mm dissected with a Bard Parker knife.
3. *Sclerectomy* – excision of a deep rectangular block (1.5 mm × 3 mm) containing anterior scleral spur, Schlemm's canal, trabeculum and peripheral cornea.
4. *Peripheral iridectomy* to prevent blockage of internal opening by the peripheral iris.
5. *Suturing of the flap.*

Complications

Shallow anterior chamber may be due to:

- Wound leak (common).

- Excessive filtration (common).
- Ciliary shutdown and choroidal detachment (uncommon).
- Ciliary block malignant glaucoma (very rare).

Miscellaneous

- Cataract (uncommon).
- Endophthalmitis (very rare).
- Total loss of visual field may rarely occur in eyes with advanced visual field loss.

Artificial filtering shunts

Definition – plastic devices that communicate between the anterior chamber and sub-Tenon's space.

Indications are intractable secondary glaucomas that have either failed or are likely to fail with conventional filtering procedures.

Main types are White pump shunt, Krupin Denver valve, Molteno tube and the Schocket tube and gutter.

Complications

- Drainage failure by blockage of the tube by blood or fibrovascular tissue.
- Overdrainage which may cause hypotony, choroidal detachment and choroidal haemorrhage.
- Implant erosion.
- Endothelial decompensation due to corneal touch by the end of the tube.

Goniotomy

Definition – a procedure that establishes communication between the anterior chamber and Schlemm's canal by removing congenitally abnormal tissue in the angle.

Indications

- Primary congenital glaucoma under the age of 3 years.
- Selected cases of Sturge–Weber syndrome and aniridia.

Surgical steps

1. Introduction of the goniotomy knife into the anterior chamber through the limbus and advancement into the angle.
2. Incision at the midpoint of the trabeculum.
3. Extension of the incision circumferentially right and then left.

Complications

- Hyphaema.
- Damage to the lens, iris and ciliary body.

Trabeculotomy

Definition – a procedure that establishes communication between the anterior chamber and Schlemm's canal by removing a part of the trabeculum.

Indications are congenital glaucomas in which the angle cannot be visualized.

Surgical steps

1. Triangular three-quarter thickness scleral flap is made.
2. Radial incision is made within the scleral bed over Schlemm's canal.
3. Lower arm of the trabeculotome is introduced into Schlemm's canal.
4. Arm of the trabeculotome is rotated into the anterior chamber.

Complications are similar to goniotomy.

Cyclodestructive procedures

Definition – procedures that lower IOP by destroying a part of the secretory ciliary epithelium.

Indications are intractable glaucomas that have failed with filtration procedures.

Types

- *Trans-scleral* are cyclocryotherapy, Nd:YAG laser cycloablation and high intensity ultrasound.
- *Direct* argon laser via the pars plana or pupil in aphakic eyes.

Chapter 8

Disorders of the lens

CATARACT

Classification

Senile

- Subcapsular
- Nuclear
- Cortical

Traumatic

- Penetrating injury
- Concussion
- Infrared irradiation
- Electric shock
- Ionizing radiation

Metabolic

- Diabetes
- Galactosaemia
- Mannosidosis
- Fabry's disease
- Lowe's (oculocerebrorenal) syndrome
- Hypocalcaemia

Toxic

- Steroids
- Chlorpromazine
- Miotics
- Busulphan (Myleran)
- Amiodarone
- Gold

Secondary (complicated) cataract

- Chronic anterior uveitis
- Hereditary retinal dystrophies
- High myopia
- Glaukomflecken (following acute glaucoma)

Maternal infections

- Rubella
- Toxoplasmosis
- Cytomegalic inclusion disease

Syndromes associated with cataract

- Down's
- Werner's
- Rothmund's

Hereditary

Cataract surgery

Indications for extraction

Visual improvement is by far the most frequent.

Medical

- Phacolytic glaucoma.
- Secondary angle-closure glaucoma due to an intumescent lens.
- Treatable retinal disease hampered by the presence of cataract.

Cosmetic to obtain a black pupil is a rare indication.

Intracapsular cataract extraction (ICCE)

Disadvantages

- Unsuitable for persons under 35 years of age because of the high risk of vitreous loss due to the intact capsulohyaloid ligament.
- Increased risk of pupil block, vitreous touch, vitreous wick, retinal detachment and cystoid macular oedema.

Surgical steps

1. *Incision* – a vertical groove is made through two-thirds of peripheral clear cornea, the anterior chamber is entered and the incision is completed with scissors.
2. *Peripheral iridectomy* is made at 12 o'clock to prevent postoperative pupil block.
3. *Zonulysin* (α-chymotrypsin) is injected into the posterior chamber to dissolve the zonules.
4. *Lens is extracted* with a cryoprobe.
5. *Lens implantation* – the pupil is constricted with acetylcholine (Miochol); hyaluronate (Healonid) is injected into the anterior chamber and an anterior chamber intraocular lens (IOL) is implanted.
6. *Completion* – the incision is sutured with 10–0 nylon and a mixture of betamethasone and gentamicin is injected subconjunctivally.

Extracapsular cataract extraction (ECCE)

Disadvantages

- Opacification of posterior capsule occurs in up to 50% of cases.
- Cannot be performed if the zonules are disrupted.

Surgical steps

1. *Incision* is the same as for ICCE but slightly smaller.
2. *Anterior capsulotomy* – many small radial cuts are made in the anterior capsule for 360°.
3. *Nucleus is expressed* by exerting counterpressure on the sclera at 6 o'clock and 12 o'clock.
4. *Cortical clean-up* is performed with either a manual or an automated infusion–aspiration device.

5. *Lens implantation* – Healonid is injected into the anterior chamber and into the capsular bag and a posterior chamber IOL is implanted.
6. *Completion* is the same as for ICCE.

Phacoemulsification

Advantages

- Small incision.
- Minimal astigmatism.
- Fast healing.

Disadvantages

- Difficult technique to master.
- Expensive instrumentation.

Surgical steps

1. *Anterior capsulotomy* as for ECCE but larger.
2. *Sculpting* of the anterior lens.
3. *Prolapse* of the nucleus.
4. *Emulsification* of the nucleus.
5. *Cortical clean-up* as for ECCE.
6. *Lens implantation* – either a small-incision IOL is implanted or the incision is enlarged sufficiently to allow the insertion of a conventional IOL.
7. *Completion* is the same as for ECCE.

Intraocular lenses

Relative contraindications

- Young children.
- Active intraocular inflammation.
- High myopia.

Designs

- *Anterior chamber,* angle-supported IOLs are now mainly used as 'back-up' lenses in the event of accidental rupture of the posterior capsule during ECCE or for secondary implantation following ICCE.
- *Posterior chamber* IOLs are implanted either into the ciliary sulcus or preferably into the capsular bag.

Complications of cataract extraction

Operative complications

- *Rupture of posterior capsule* predisposes to vitreous loss.
- *Vitreous loss* predisposes to postoperative updrawn pupil, iris prolapse, vitreous touch, vitreous wick syndrome, uveitis, retinal detachment and cystoid macular oedema.
- *Expulsive haemorrhage* (very rare).

Early postoperative complications

- *Iris prolapse* predisposes to defective wound healing, astigmatism, uveitis, epithelial ingrowth, cystoid macular oedema and bacterial endophthalmitis.
- *Hyphaema* is usually transient and innocuous.
- *Striate keratopathy* due to endothelial trauma.
- *Wound leak* due to inadequate suturing.
- *Pupil block* may occur following ICCE with anterior chamber IOL.
- *Bacterial endophthalmitis* (very rare).

Late postoperative complications

- *Cystoid macular oedema* is more common following ICCE (for management see Chapter 12).
- *Opacification of the posterior capsule* occurs in up to 50% after ECCE.
- *Retinal detachment* occurs in 2% after ICCE and 0.5% after ECCE.
- *Bullous keratopathy* due to endothelial decompensation in eyes with anterior chamber IOLs (uncommon).
- *Epithelial ingrowth* may cause intractable glaucoma (very rare).
- *Vitreous touch syndrome* due to vitreo-endothelial contact following following prolapse of vitreous gel into the anterior chamber (rare).
- *Vitreous wick syndrome* due to prolapse of vitreous gel through a defect in the incision (rare).
- *'UGH' syndrome* (uveitis, glaucoma and hyphaema) in eyes with anterior chamber IOLs (rare).
- *'Sunset' syndrome* in which a posterior chamber IOL dislocates inferiorly (very rare).

Management of infantile cataract

Unilateral vs bilateral

The prognosis for dense unilateral cataract is usually very poor because of intractable stimulus deprivation amblyopia.

Density of cataract

- *Very dense* – no view is obtained of the fundus with either the indirect or the direct ophthalmoscope.
- *Moderately dense* – poor view with direct but good with indirect.
- *Mild* – good view with both.

Visual function

- *Nystagmus* in early infancy is a sign of poor vision until proved otherwise.
- *Pupillary reactions* are considered poor if the fundus can be seen with an indirect ophthalmoscope through an undilated pupil. A poor pupillary reaction in an eye with cataract implies either an associated optic nerve lesion (e.g. severe hypoplasia) or extensive retinal disease (e.g. Leber's amaurosis).
- *Fixation reflexes* during feeding and looking at the mother's face.
- *Induced nystagmus* by rotating the child. After cessation of the rotation in a sighted infant the nystagmus will stop before the third beat or within 5 seconds, whereas in a blind infant the nystagmus will continue for longer.
- *Preferential looking* is an accurate but time-consuming test.
- *Catford drum* is a gross test.
- *Visually evoked response.*

Laboratory investigations

- *Rubella* – serology for IgM antibody, virus cultures, platelet count and maternal rubella titres.
- *Galactosaemia* – urinalysis for reducing substance, and red cell galactose-1-phosphate uridylyltransferase (GPUT) and galactokinase (GK) activity.
- *Hypoglycaemia* – blood glucose.
- *Lowe's syndrome* – urine chromatography for amino acids.
- *Hypocalcaemia* – serum calcium and phosphorus, and skull X-rays for calcification of the basal ganglia.

Indications for surgery

- Bilateral advanced cataract – surgery within a few weeks of birth.
- Bilateral moderate – postpone surgery if near vision is good.
- Bilateral mild – surgery is unnecessary.
- Unilateral advanced – the results of surgery are very poor because of dense stimulus deprivation amblyopia.

Techniques

- Lensectomy in which the entire lens is excised with a vitreous cutter.
- Aspiration of the lens with an infusion–aspiration device, but opacification of the posterior capsule is very common.

ECTOPIA LENTIS

Causes

Marfan's syndrome

Inheritance is dominant.

Cause is a widespread connective tissue abnormality.

Systemic features

- *Cardiac anomalies* – mitral incompetence with a floppy valve, aortic incompetence, dilated aortic root and dissecting aortic aneurysm.
- *Skeletal anomalies* – limbs are inappropriately long as compared with the trunk, long spider-like fingers (arachnodactyly), funnel chest, unstable joints, flat feet, scoliosis and a high arched palate.
- *Miscellaneous* – muscular underdevelopment and spontaneous pneumothorax.

Ocular features

- Lens subluxation which is upward, bilateral and non-progressive in 80% of cases.
- Microspherophakia (uncommon).
- Glaucoma may occur either due to lens subluxation or an associated congenital angle anomaly (uncommon).

- Miscellaneous – hypoplasia of dilator pupillae, flat cornea, axial myopia and retinal detachment.

Weill–Marchesani syndrome

Inheritance is recessive.

Systemic features

- Short stature.
- Short stubby fingers (bradydactyly).
- Mental handicap.

Ocular features

- Microspherophakia which may be associated with anterior lens dislocation.
- Glaucoma may occur either due to pupil block by the lens or an associated congenital angle anomaly.

Homocystinuria

Inheritance is recessive.

Cause is a deficiency of cystathione synthase.

Systemic features

- *Vascular anomalies* – increased platelet stickiness which may predispose to thrombosis particularly after general anaesthesia.
- *Skeletal anomalies* – arachnodactyly, osteoporosis and fractures.
- *Miscellaneous* – mental handicap, fine hair and malar flush.

Ocular features

- Lens subluxation is usually downwards with complete disruption of the zonules.
- Glaucoma may occur either due to pupil block or dislocation of the lens into the anterior chamber.

Other causes of ectopia lentis

Hereditary

- *Hyperlysinaemia* – a deficiency in lysine dehydrogenase characterized by motor and growth retardation, and mental handicap.
- *Ehlers–Danlos syndrome* characterized by joint hyperextensibility, skin hyperelasticity, blue sclera, angioid streaks, keratoconus and retinal detachment.
- *Familial ectopia lentis* unassociated with any systemic defect.
- *Aniridia* (see Chapter 7).

Acquired

- Trauma.
- Large eye (buphthalmos, high myopia).
- Anterior uveal tumours.
- Chronic cyclitis.
- Hypermature cataract.
- Syphilis.

ABNORMALITIES OF SHAPE

Coloboma is usually inferior and may be associated with colobomas of the iris and choroid as well an an increased risk of giant retinal tears.

Anterior lenticonus consits of an anterior axial bulge. It may occur in *Alport's syndrome* (hereditary progressive haematuric nephritis and sensorineural deafness).

Posterior lenticonus consists of a posterior axial bulge.

Lentiglobus is a generalized hemispherical deformity of the lens.

Microphakia is a small lens which occurs in Lowe's syndrome.

Microspherophakia is a small spherical lens. It may be an *isolated* finding or it may occur in *Marfan's syndrome, Weill–Marchesani syndrome* and *hyperlysinaemia.*

Chapter 9

Retinal detachment

Definition

A retinal detachment (RD) is a deparation of the sensory retina from the retinal pigment epithelium (RPE) by subretinal fluid (SRF).

Classification

Rhegmatogenous

- Spontaneous
- Traumatic

Non-rhegmatogenous

- Tractional
- Exudative

PATHOGENESIS

Rhegmatogenous retinal detachment

Due to an interplay between vitreoretinal traction and a weakness in the peripheral retina (most frequently lattice degeneration).

Vitreoretinal traction in chronological order:

- Degenerative liquefaction of the vitreous gel (synchysis senilis).
- Passage of fluid through a hole in the posterior hyaloid.

- This induces an acute posterior vitreous detachment (PVD) with collapse.
- Residual attachments of vitreous to the peripheral retina exert traction and cause a retinal tear which subsequently leads to RD.

Lattice degeneration

- Bilateral, sharply demarcated, circumferentially orientated, spindle-shaped lesions.
- Location – between the equator and the posterior border of the vitreous base and most common superotemporally.
- Associated features are white lines, snowflakes, RPE hyperplasia, small holes and overlying vitreous synchysis with exaggerated attachments to the retina.

Tractional retinal detachment

Due to progressive contraction of fibrous or fibrovascular membranes over large areas of vitreoretinal adhesion.

Causes

- Proliferative retinopathies.
- Penetrating ocular trauma.

Exudative retinal detachment

Due to subretinal disorders that damage the RPE and allow the passage of fluid from the choroid into the subretinal space.

Causes

- Intraocular tumours.
- Intraocular inflammation.
- Panretinal photocoagulation.
- Retinal surgery.
- Toxaemia of pregnancy.

CLINICAL FEATURES

Rhegmatogenous retinal detachment

Prevalence – affects 1 in 10 000 of the population each year.

Symptoms in chronological order:

- Photopsia caused by vitreoretinal traction.
- Vitreous floaters of two types: a large (Weiss) ring due to the detached annular attachment to the optic disc or a sudden shower of many tiny spots due to vitreous haemorrhage.
- Visual field defect due to spread of SRF posterior to the equator.
- Loss of central vision due to macular detachment.

Signs

- *Fresh RD* – retinal breaks are present, the configuration of the RD is convex, mobility is good and shifting fluid is absent.
- *Old RD* – the retina is thin and secondary intraretinal cysts and subretinal demarcation lines (high-water marks) may be present.
- *Proliferative vitreoretinopathy (PVR):*

  grade A (minimal) – diffuse vitreous haze;
  grade B (moderate) – breaks with rolled edges and surface wrinkling of the retina;
  grade C (marked) – full-thickness retinal folds involving one to three quadrants;
  grade D (massive) – full-thickness retinal folds involving four quadrants and a funnel-shaped RD.

Tractional retinal detachment

Prevalence – common.

Symptoms – photopsia and floaters are usually absent.

Signs

- Fibrous or fibrovascular proliferation is present.
- Configuration of the RD is concave.
- Mobility is severely restricted.

- *Absent* – retinal breaks, secondary intraretinal cysts, high water marks and shifting fluid.

Exudative retinal detachment

Prevalence – rare.

Symptoms – photopsia is absent and floaters are usually absent.

Signs

- Shifting fluid is presenet.
- Configuration of the RD is convex.
- Retinal mobility is marked.
- *Absent* – PVR, retinal breaks, secondary intraretinal cysts and high water marks.

DIFFERENTIAL DIAGNOSIS

Acquired retinoschisis

Definition – a splitting of the neuroretina into two layers.

Prevalence – very common.

Symptoms

- Photopsia and floaters are absent.
- A visual field defect is seldom present because spread posterior to the equator is rare. If it does occur the field defect is absolute.

Signs

- Retinal breaks may be absent or present in one or both layers.
- Configuration is convex.
- Mobility is severely restricted.
- Absent – PVR, secondary intraretinal cysts and shifting fluid.
- Associated features include snowflakes and sheathed vessels in the inner layer and a 'beaten metal' appearance of the outer layer; the schisis cavity may contain rows of torn grey–white neural tissue.

Choroidal detachment

Definition – an effusion of fluid into the suprachoroidal space.

Prevalence – common.

Symptoms

- Photopsia and floaters are absent.
- A visual field defect is seldom present because spread posterior to the equator is rare.

Signs – the elevation is convex, brown in colour and immobile.

PROPHYLAXIS OF RHEGMATOGENOUS RETINAL DETACHMENT

Retinal breaks

The following factors should be considered when contemplating prophylaxis.

Type – a tear is more dangerous than a hole.

Size – a large tear is more dangerous than a small tear.

Symptoms – symptomatic breaks associated with PVD are more dangerous than asymptomatic breaks and all should be treated prophylactically.

Location – superior breaks are more dangerous than inferior breaks, and temporal breaks are more dangerous than nasal because, in the event of RD, the macula is threatened.

Pigmentation around the break is indicative of long duration and a low risk of RD.

Miscellaneous – any break should be taken seriously in the presence of:

- Aphakia.
- High myopia.
- Only eye.
- Positive family history of RD.
- Marfan's syndrome, Stickler's syndrome and Ehlers–Danlos syndrome because of poor prognosis of RD.

Lattice degeneration

Indications for prophylactic treatment are:

- RD in the fellow eye.
- Extensive lattice in aphakia and high myopia.
- Strong family history of RD.
- Marfan's syndrome, Stickler's syndrome and Ehlers–Danlos syndrome.

RETINAL SURGERY

Scleral buckling

Purposes

- To close breaks by apposing the RPE to the sensory retina.
- To release vitreoretinal traction.

Configuration of buckle

Radial for:

- Large 'U'-shaped tears.
- Posterior breaks.

Segmental circumferential for:

- Multiple breaks.
- Anterior breaks.
- Wide breaks such as dialyses and giant tears.

Encircling for:

- Breaks in three or four quadrants.
- Extensive lattice degeneration.
- Extensive RD without detectable breaks.
- PVR grade C.

Surgical steps

1. *Peritomy* – the conjunctiva is incised near the limbus.
2. *Bridle sutures* are placed under the rectus muscles.
3. *Localization* of retinal breaks in relation to the sclera.
4. *Cryotherapy* to breaks and predisposing lesions.
5. *Scleral buckling* either local or encircling.
6. *Drainage of SRF,* if necessary.
7. *Completion* – sutures tightened over the buckle and subconjunctival injection of betamethasone and gentamicin.

Indications for drainage of subretinal fluid

- Difficulty in localizing breaks in a bullous RD.
- Immobile retina – in order to appose the sensory retina to the RPE.
- Long-standing – RD because the SRF is viscous and takes a long time to absorb.
- Inferior break – because gravity tends to keep the break open.
- Fear of elevating the IOP in eyes with advanced glaucoma, thin sclera and recent cataract extraction.

Indications for air injection

- Hypotony after drainage of SRF.
- Fishmouthing of the tear.
- To flatten severe radial retinal folds.
- Posterior breaks, macular holes and certain giant tears.

Causes of failure

Early

- Missed retinal break.
- Buckle failure due to inadequate size, incorrect position or inadequate height.

Late

- Most frequently due to PVR.
- Reopening of break due to inadequate chorioretinal reaction.
- New retinal break.
- Late buckle failure due to slipping of an encircling band and extrusion or removal of the explant.

Postoperative complications

Early

- Acute orbital cellulitis.
- Choroidal detachment.
- Bacterial endophthalmitis.

Late

- Exposure or infection of the explant.
- Maculopathy.
- Diplopia.

Pars plana vitrectomy

Indications

- Tractional RD involving or threatening the macula.
- PVR grade D.
- Certain RD due to giant retinal tears.

Surgical steps

1. *Excision* of vitreous gel.
2. *Mobilization* of the retina by the segmentation of epiretinal membranes.
3. *Flattening* of the retina by internal drainage of SRF or internal silicone/fluid exchange.
4. *Endophotocoagulation* in active proliferative diabetic retinopathy.

Postoperative complications

These vary according to the indications for surgery and include:

- Recurrence of vitreous haemorrhage.
- Cataract.
- Neovascular glaucoma, particularly in eyes with proliferative diabetic retinopathy and persistent RD.

Chapter 10

Retinal vascular disorders

DIABETIC RETINOPATHY (DR)

Classification

Background

Maculopathy

- Focal
- Diffuse
- Ischaemic
- Mixed

Pre-proliferative

Proliferative

Advanced diabetic eye disease

Background diabetic retinopathy (BDR)

Clinical features

Incidence is 30% after 10 years and 90% after 30 years.

Signs

- Microaneurysms – small red dots in inner retinal layers.
- Dot and blot haemorrhages located in the deeper retinal layers.

- Hard exudates are frequently located in a circinate pattern peripheral to areas of chronic leakage.
- Retinal thickening due to diffuse retinal oedema.

Management

- Good metabolic control.
- Control hypertension.
- Control anaemia.
- Annual fundus examination.

Focal diabetic maculopathy

Clinical features

Signs

- BDR with hard exudates.
- Visual acuity is reduced by mild macular oedema or rings of hard exudates spilling over into the fovea.

Fluorescein angiography shows focal leakage but good perfusion.

Treatment

Technique – focal argon laser photocoagulation to microaneurysms and centres of hard exudate rings (spot size 50–200 μm, duration 0.1 second).

Results are reasonably good with frequent stabilization but seldom improvement of visual acuity.

Diffuse diabetic maculopathy

Clinical features

Signs

- BDR with few if any hard exudates.
- Visual acuity is reduced by diffuse retinal oedema involving the fovea.
- Cystoid macular oedema and lamellar holes in long-standing cases.

Fluorescein angiography shows diffuse leakage but good macular perfusion.

Treatment

Technique – grid argon laser photocoagulation with 150–200 low intensity burns to the macula but not the fovea (spot size 50–200 μm, duration 0.05–0.1 second).

Results are frequently disappointing.

Ischaemic diabetic maculopathy

Clinical features

Signs are similar to diffuse maculopathy.

Fluorescein angiography shows poor macular perfusion.

Treatment

There is no treatment but the patient should be watched carefully because of increased risk of proliferative DR.

Pre-proliferative diabetic retinopathy

Clinical features

Signs

- Cotton-wool spots.
- Intraretinal microvascular abnormalities (IRMAs) thought to represent intraretinal new vessels.
- Venous changes consisting of dilatation, beading, looping and sausage-like segmentation.
- Arteriolar narrowing which may resemble a branch retinal artery occlusion.
- Large dark blot haemorrhages which represent haemorrhagic infarcts.

Fluorescein angiography shows large areas of capillary non-perfusion.

Treatment

Usually is unnecessary but the patient should be watched carefully for the development of proliferative DR.

Proliferative diabetic retinopathy (PDR)

Clinical features

Incidence – affects 5% of diabetics.

Signs in chronological order:

- Neovacularization on the disc (NVD) or elsewhere (NVE).
- Posterior vitreous detachment which is usually gradual and incomplete.
- Elevation of the new vessels.
- Vitreous haemorrhage.
- Tractional RD.

Prognosis without treatment

- Severe NVD + vitreous haemorrhage – 40% blind within 2 years.
- Moderate NVD – 25% blind within 2 years.
- Mild NVD – 10% blind within 2 years.
- Severe NVE + vitreous haemorrhage – 30% blind within 2 years.
- Modertate NVE – 15% blind within 2 years.
- Mild NVE – 7% blind within 2 years.

Treatment

Technique – panretinal argon laser photocoagulation with 2000–3000 burns (spot size – 500 μm, duration – 0.2 second) in one or two sessions.

Results are good provided the condition is not too advanced.

Advanced diabetic eye disease

Clinical features

Incidence – rare following adequate treatment of proliferative DR.

Signs

- Persistent vitreous haemorrhage.
- Tractional RD.
- Opaque membranes on the posterior surface of the detached hyaloid between the superior and inferior arcades.

- Neovascular glaucoma is common following unsuccessful vitrectomy with persistent RD.

Treatment

Indications

- Persistent dense vitreous haemorrhage.
- Tractional RD involving the fovea.

Technique is pars plana vitrectomy.

Results are reasonably good with improvement of visual acuity in about 70% of cases.

RETINAL VEIN OCCLUSION

Predisposing factors

- Systemic hypertension increases the risk of both branch (BRVO) and central vein occlusion (CRVO)
- Raised IOP increases the risk of CRVO.
- Hypermetropia increases risk of BRVO by an unknown mechanism.
- Blood hyperviscosity
- Inflammatory periphlebitis

Classification

Branch retinal vein occlusion (BRVO)

- Main vein (hemisphere) occlusion at the disc ± macular oedema
- Major vein occlusion away from the disc ± macular oedema
- Macular vein occlusion
- Peripheral vein occlusion

Central retinal vein occlusion (CRVO)

- Ischaemic.
- Non-ischaemic

Branch retinal vein occlusion

Clinical features

Presentation of this *very common* condition is usually with a unilateral subacute onset of a visual field defct.

Early signs

- Venous tortuous and dilated.
- Flame-shaped and dot and blot haemorrhages.
- Cotton-wool spots and retinal oedema.

Late signs

- Venous sheathing.
- Cystoid macular oedema.
- Microaneurysms and hard exudates.
- Collaterals and shunts.

Prognosis depends on the extent of macular venous drainage involved by the occlusion and the integrity of perifoveal capillary network. In general 50% of eyes have a visual acuity of 6/12 or better after 6 months.

Complications

- Chronic macular oedema is the most common cause for poor visual acuity.
- Neovascularization occurs in 30% of eyes with major BRVO.

Treatment

- Focal argon laser photocoagulation in eyes with visual acuity less than 6/12 due to chronic macular oedema.
- PRP for neovascularization.

Non-ischaemic central retinal vein occlusion

Clinical features

Presentation of this *common* condition is with a unilateral subacute onset of generalized mild to moderate blurring of vision.

Signs

- Visual acuity is moderately reduced.
- Relative pupillary conduction defect is absent or mild.
- Venous tortuosity and dilatation are mild or moderate.
- Haemorrhages are most numerous in the periphery.
- Cotton-wool spots are few or absent.
- Optic disc swelling and macular oedema are mild.

Fluorescein angiography shows venous stasis but poor perfusion.

Prognosis – 50% resolve with improvement of visual acuity and 50% have persistently poor visual acuity due to macular oedema.

Treatment

Usually is unnecessary in absence of neovascularization.

Ischaemic central retinal vein occlusion

Clinical features

Presentation of this *uncommon condition* is similar to the non-ischaemic type but visual impairment is more severe.

Signs are all more severe than in the non-ischaemic type.

Fluorescein angiography initially shows masking by blood and later it shows extensive capillary non-perfusion.

Prognosis – 50% incidence of neovascular glaucoma within 3 months and some patients develop new retinal vessels and vitreous haemorrhage.

Treatment

PRP to prevent neovascular glaucoma.

RETINAL ARTERY OCCLUSION

Causes

Embolization from the heart:

- Calcific emboli from aortic valves.
- Thrombus due to myocardial infarction, mitral stenosis and prolapsed mitral valve.

- Vegetations in subacute bacterial endocarditis.
- Myxomatous material from an atrial myxoma.

Embolization from stenosed carotid arteries in the neck:

- Cholesterol emboli (Hollenhorst's plaques) are usually asymptomatic.
- Fibrinoplatelet emboli may cause amaurosis fugax.
- Calcific emboli may cause permanent occlusion.

Vaso-obliteration

- Atheroma.
- Arteritis associated with collagen vascular disorders.

Acute, very severe rise in IOP:

- Acute angle-closure glaucoma.
- Accidental pressure on globe.
- During RD surgery.

Central retinal artery occlusion (CRAO)

Clinical features

Presentation of this *rare* condition is with a unilateral, acute, severe generalized loss of vision.

Signs

- Visual acuity is severely impaired.
- Afferent pupillary conduction defect is marked.
- Retina is white and oedmatous especially at the posterior pole where a 'cherry-red spot' at the fovea is evident.
- Arterioles are very narrow and irregular in calibre.
- Blood column in the veins and arterioles is sludgy and segmented (cattletrucking).

Prognosis is very poor.

Branch retinal artery occlusion

Clinical features

Presentation of this *uncommon* condition is with a unilateral acute visual field defect.

Signs are similar to CRAO but confined to the area supplied by the obstructed artery.

Prognosis is very poor except in transient obstruction by platelet emboli.

Management of retinal artery occlusion

Emergency treatment

- Lie patient flat to help maintain the circulation.
- Apply intermittent firm ocular massage for 15 min to lower the IOP, improve blood flow and dislodge emboli.
- Give intravenous acetazolamide (Diamox) 500 mg to lower the IOP.
- Other measures include inhalation of mixture of 5% carbon dioxide and 95% oxygen, and anterior chamber paracentesis.
- ESR to exclude giant cell arteritis.

Cardiovascular investigations

- *Physical examination* involving palpation and auscultation of the external carotids to detect stenosis.
- *Non-invasive investigations* are ophthalmodynamometry, carotid artery imaging by B-scan ultrasound or Doppler.
- *Invasive investigations* are carotid angiography and digital subtraction angiography.

Treatment of carotid disease

- Endarteriectomy.
- Antiplatelet therapy or anticoagulants if surgery is inappropriate.

HYPERTENSIVE RETINOPATHY

Hypertensive features

- *Grade 1* – mild generalized arteriolar constriction.
- *Grade 2* – severe grade 1 + focal constriction.
- *Grade 3* – grade 2 + haemorrhages, cotton-wool spots and hard exudates.
- *Grade 4* – grade 3 + disc swelling.

Arteriosclerotic features

- *Grade 1* – broad arteriolar reflex + simple vein concealment.
- *Grade 2* – grade 1 + deflection of veins at arteriovenous crossings (Salus's sign).
- *Grade 3* – grade 2 + 'copper wire' arterioles + marked changes at arteriovenous crossings.
- *Grade 4* – grade 3 + 'silver wire' arterioles.

SICKLE-CELL RETINOPATHY

Proliferative

Clinical features

- *Stage 1* – peripheral arteriolar occlusion.
- *Stage 2* – peripheral arteriovenous anastomoses.
- *Stage 3* – peripheral 'seafan' neovascularization from anastomoses.
- *Stage 4* – vitreous haemorrhage.
- *Stage 5* – tractional RD.

Treatment

- Peripheral retinal photocoagulation.
- Pars plana vitrectomy for advanced disease.

Non-proliferative

Clinical features

Asymptomatic lesions

- Venous tortuosity.
- Arteriolar silver-wiring.
- Chorioretinal atrophy ('black sunbursts').
- Pink superficial 'salmon patch' haemorrhages.
- Refractile haemosiderin deposits.
- Retinal breaks.
- Angioid streaks.

Symptomatic lesions are occlusions of veins, arteries and choroidal vessels.

RETINOPATHY OF PREMATURITY

Active

Risk factors

- Premature infants exposed to a high ambient oxygen concentration.
- Birth weight less than 1300 grams.

Clinical features

Signs

- *Stage 1* – demarcation line consisting of a thin tortuous white line parallel with the ora serrata most prominent in the temporal periphery, which separates the avascular immature retina from vascularized retina.
- *Stage 2* – line develops into a ridge containing isolated neovascular tufts.
- *Stage 3* – ridge is associated with extraretinal fibrovascular proliferation which may cause a vitreous haemorrhage.
- *Stage 4* – subtotal tractional RD typically occurs at about 10 weeks.
- *Stage 5* – total tractional RD.

Prognosis – spontaneous regression occurs in 80% of cases, even from stage 3.

Management

Screening – most useful time is between the seventh and ninth weeks of life of infants who have received supplemental oxygen and who were either born at less than 36 weeks' gestation or who weighed less than 2000 grams.

Treatment

- Cryotherapy to ablate peripheral avascular immature retina.
- Pars plana vitrectomy for tractional RD.
- Vitamin E is of uncertain value.

Cicatricial

Clinical features

In increasing order of severity:

- Peripheral retinal pigmentation and haze at the vitreous base.
- Myopia.
- Peripheral retinal breaks and lattice degeneration.
- Fibrous band in temporal retinal periphery which may pull on the vascular arcades and cause heterotopia of the macula.
- Retrolental cyclitic membrane.
- Funnel-shaped total tractional RD.
- Secondary angle-closure glaucoma.

Treatment

- Prophylactic laser for breaks and degenerations.
- Pars plana vitrectomy for tractional RD.
- Lensectomy and anterior vitrectomy for secondary angle-closure glaucoma.

RETINAL TELANGIECTASIAS

Idiopathic juxtafoveolar retinal telangiectasia

Clinical features

Presentation of this *rare* condition is in adults with usually unilateral, gradual impairment of visual acuity.

Signs are *unilateral* or *bilateral* dilated microvascular channels and capillary non-perfusion near fovea.

Complications are reduced visual acuity due to intraretinal oedema or hard exudates.

Treatment

Focal argon laser photocoagulation may be helpful.

Leber's miliary aneurysms

Clinical features

Presentation of this *rare* condition is in young adults with usually unilateral gradual visual impairment.

Signs are *unilateral* or *bilateral* fusiform, saccular dilatation of veins and arteries in the temporal retinal periphery.

Complications are reduced visual acuity due to macular hard exudates.

Treatment

Peripheral photocoagulation or cryotherapy.

Coats' disease

Clinical features

Presentation of this *uncommon* condition is usually in young boys with unilateral, gradual visual impairment or a white pupil (leukocoria).

Signs are *unilateral* hard exudate at the posterior pole and periphery.

Complications are severe visual loss due to exudative RD, retrolental mass, rubeosis iridis, cataract, uveitis, glaucoma and phthisis.

Treatment

Photocoagulation or cryotherapy in early cases may prevent progression.

Chapter 11

Acquired maculopathies

AGE-RELATED (SENILE) MACULAR DEGENERATION

Drusen

This *very common* lesion may be the earliest clinical manifestations of age-related macular degeneration (AMD).

Histopathological features

- Thickening of Bruch's membrane.
- Atrophy of retinal pigment epithelium (RPE).
- Degeneration of retinal receptors.

Clinical features

Signs

Bilateral, yellow–white, slightly elevated spots at the macula which are of four main types:

- Small round and discrete (hard).
- Large with ill-defined borders (soft or diffuse).
- Mixed hard and soft.
- Calcified glistening.

Prognosis in the vast majority of patients is excellent although a few subsequently develop visual loss.

Non-exudative (dry) AMD

Clinical features

Presentation of this *very common* condition is during the seventh to eighth decades with usually initially unilateral and gradual impairment of visual acuity.

Signs are sharply demarcated circular areas of atrophy of RPE and choriocapillaris (geographical or areolar atrophy).

Treatment

None apart from low vision aids.

Exudative (neovascular) AMD

Clinical features

In chronological order:

Detachment of RPE

- *Symptoms* are subacute onset of metamorphopsia and mild to moderate impairment of visul acuity.
- *Signs* are sharply circumscribed dome-shaped elevation at macula with clear or turbid sub-RPE fluid.
- *Subsequent course* is either spontaneous resolution leaving an atrophic RPE or detachment of the sensory retina.

Choroidal subretinal neovascular membranes (SRNVMs)

- *Symptoms* are a subacute onset of metamorphopsia.
- *Signs* – the lesion may be clinically invisible or it may appear as a grey–green or pinkish-yellow sub-RPE lesion.

Subsequent course of SRNVMs

- Rupture of blood vessel within SRNVMs.
- Haemorrhagic detachment of RPE appearing as a black elevated mound.
- Haemorrhagic detachment of the sensory retina.
- Organization of subretinal blood.

- Formation of disciform scarring (common).
- Profuse leakage from disciform scar (uncommon).
- Exudative retinal detachment (rare).

Treatment of SRNVM

Indications are extrafoveal SRNVMs 200 μm or more from centre of the fovea.

Contraindications

- SRNVMs closer than 200 μm from the fovea with associated detachment of the RPE and/or subfoveal blood.
- Visual acuity of less than 6/24.

Technique – SRNVMs are covered with 200 μm (0.5 s) confluent intense white burns.

Results – severe visual loss is initially reduced by 50% but late recurrences are frequent.

CENTRAL SEROUS RETINOPATHY

Clinical features

Presentation of this common condition is typically in men during the third to fourth decades with a unilateral subacute onset of metamorphopsia, a positive relative scotoma and mild impairment of visual acuity.

Signs

- Visual acuity is mildly reduced (6/9–6/12) and may be improved to 6/6 with a weak plus lens.
- Fundoscopy shows a round or oval elevation of the sensory retina at the posterior pole outlined by a glistening reflex (halo).

Fluorescein angiography shows either a smoke-stack or an ink-blot appearance.

Prognosis

- 90% resolve within 1–6 months with return of visual acuity to normal although mild symptoms may remain longer.
- In 10% visual acuity remains impaired due to RPE changes.
- Recurrence rate is 40%

Treatment

Relative indications for argon laser photocoagulation are:

- Recurrences that have caused loss of visual acuity.
- Impaired visual acuity in the fellow eye from central serous retinopathy.
- Duration longer than 4 months.

CYSTOID MACULAR OEDEMA

Causes

Retinal vascular disease

- Background diabetic retinopathy.
- Retinal vein occlusion.

Inflammation

- Post-cataract (Irvine–Gass syndrome).
- Intermediate uveitis.
- Posterior uveitis.

Miscellaneous

- Topical 2% adrenaline in aphakia.
- Retinitis pigmentosa.
- Secondary to other maculopathies.
- Peripheral retinal photocoagulation.

Clinical features

Presentation of this *very common* condition is usually with unilateral variable gradual impairment of visual acuity and a relative central scotoma.

Signs are loss of foveal depression due to retinal thickening with multiple cystoid spaced at the fovea.

Fluorescein angiography shows the typical 'flower-petal' pattern of leakage.

Treatment

- *Medical* with steroids and/or acetazolamide may help in some cases of retinitis pigmentosa, intermediate uveitis and Irvine–Gass syndrome.
- *Surgical* by pars plana vitrectomy in Irvine–Gass syndrome associated with anterior segment vitreous complications

MACULAR HOLE

Causes

- Idiopathic in elderly patients – 10% are eventually bilateral.
- High myopia – may cause a retinal detachment.
- Trauma.
- Solar retinopathy may cause a small lamellar hole or a cyst.

Clinical features of a senile hole

Presentation of this *common* condition is with unilateral gradual impairment of visual acuity.

Signs are a round 'punched-out' area at the fovea with yellow pigment in its base surrounded by a grey halo of retinal elevation.

Treatment

None possible.

MYOPIC MACULOPATHY

Clinical features

Presentation of this *common* condition is with usually initially unilateral metamorphopsia or impaired visual acuity.

Signs

- *Early* are atrophy of the RPE and choriocapillaris with unmasking of the large choroidal vessels and sclera.
- *Late* are proliferation of the RPE at the fovea (Fuchs' spot) or large breaks in Bruch's membrane (lacquer cracks) and secondary neovascular maculopathy.

Treatment

None possible.

MACULAR PUCKER

Causes

- Idiopathic in the elderly – 20% are eventually bilateral.
- Retinal vascular disorders.
- Iatrogenic following retinal surgery, retinal photocoagulation or cryotherapy.

Clinical features

Presentation of this *common* condition is with unilateral metamorphopsia and variable impairment of visual acuity.

Signs

- *Grade 1* (cellophane maculopathy) is characterized by a translucent epiretinal membrane with an irregular light reflex and mild retinal wrinkling and vascular tortuosity.
- *Grade 2* (macular pucker) is characterized by a dense epiretinal membrane with marked retinal wrinkling and vascular tortuosity.

Treatment

In a few highly selected cases the membrane can be surgically peeled off the retina.

CHOROIDAL FOLDS

Causes

- Idiopathic in hypermetropic eyes.
- Retrobulbar mass.
- Thyroid ophthalmopathy.
- Choroidal tumour.
- Ocular hypotony

Clinical features

Presentation of this *rare* condition is with a gradual onset of metamorphopsia and variable impairment of visual acuity.

Signs are fine parallel striae at the posterior pole with the elevated crescent being brighter than the trough.

Treatment

Treatment is that of the cause.

ANGIOID STREAKS

Clinical features

Presentation of this rare condition is with initially unilateral metamorphopsia and variable impairment of visual acuity through involvement of the fovea by a streak or neovascular maculopathy.

Signs

- Linear streaks with irregular edges radiating from the disc.
- RPE mottling (peau d'orange).
- Peripapillary chorioretinal atrophy.
- Optic disc drusen are a frequent finding.

Systemic associations

- None in 50%.

- Pseudoxanthoma elasticum ('Grönblad–Strandberg' syndrome).
- Paget's disease.
- Ehlers–Danlos syndrome.
- Sickle-cell disease (rare).

CHLOROQUINE MACULOPATHY

Incidence

Rare with a total dose of less than 300 g (i.e. 250 mg daily for 3 years); risk increases thereafter.

Clinical features

Premaculopathy

- Visual acuity is normal.
- Scotoma to a red target between 4° and 9° of fixation.
- Disappears if drug is stopped.

Established maculopathy

- Visual acuity is slightly reduced.
- Loss of the foveolar reflex and RPE stippling.
- Usually non-progressive if drug is stopped.

Bull's eye maculopathy

- Visual acuity is moderately reduced.
- Central foveolar hyperpigmentation surrounded by a depigmented zone and encircled by a hyperpigmented ring.
- May progress even if drug is stopped.

Advanced retinopathy

- Visual acuity is severely reduced.
- Arterioles are constricted and the peripheral retina shows pigmentary changes.
- Irreversible.

Chapter 12

Hereditary disorders of the retina and choroid

RETINITIS PIGMENTOSA

Typical retinitis pigmentosa

Inheritance

- *Autosomal recessive* is the most common and the most severe.
- *Autosomal dominant* is the next most common and is relatively benign.
- *X-linked recessive* is least common and is relatively severe.

Clinical features

Presentation of this *uncommon* condition is usually during the second decade with night blindness.

Signs

- Bilateral peripheral retina shows perivascular 'bone-spicule' pigmentation with unmasking of the larger choroidal blood vessels and arteriolar attenuation.
- Optic disc is initially normal but later becomes waxy pale.
- Maculopathy may be cystoid, atrophic or cellophane.

Miscellaneous associations

- Open-angle glaucoma in 3%.
- Posterior subcapsular cataracts occur early in recessive cases.
- Myopia is common.
- Keratoconus is rare.

Atypical retinitis pigmentosa

Sine pigmento with minimal or absent pigmentary changes.

Retinitis punctata albescens in which scattered white dots may precede typical retinitis pigmentosa.

Sector in which one quadrant (usually nasal) or one half (usually inferior) is involved.

Pericentric in whch the pigmentary changes are confined to the pericentral retina.

Systemic associations of retinitis pigmentosa

Bassen–Kornzweig syndrome (abetalipoproteinaemia)

- *Inheritance* is recessive; typically affects Ashkenazi Jews.
- *Systemic features* are fat malabsorption, spinocerebellar ataxia, acanthocytosis, abetalipoproteinaemia and ultimate death.
- *Treatment* is with vitamin A.

Refsum's syndrome (phytanic acid storage disease)

- *Inheritance* is recessive.
- *Systemic features* are peripheral neuropathy and cerebellar ataxia; CSF protein is increased in the absence of pleocytosis, deafness and ichthyosis.
- *Treatment* is with phytanic acid-free diet and plasma exchange.

Usher's syndrome

- *Inheritance* is recessive.
- *Systemic features* are congenital non-progressive sensorineural deafness.

Cockayne's syndrome

- *Inheritance* is recessive.
- *Systemic features* are dwarfism, progressive mental handicap, deafness, nystagmus, ataxia, and premature ageing and death.

Kearns–Sayre syndrome

- *Inheritance* – nil.
- *Systemic features* are ocular myopathy, heart block which may cause sudden death, short stature, delayed puberty, mental handicap, cerebellar ataxia and deafness.

Laurence–Moon–Biedl syndrome

- *Inheritance* is recessive.
- *Systemic features* are mental handicap, obesity, polydactyly and hypogonadism.

Friedreich's ataxia

- *Inheritance* is recessive.
- *Systemic features* are posterior column disease, ataxia and nystagmus.

LEBER'S AMAUROSIS

Inheritance

Usually recessive.

Ocular features

Presentation of this *very rare* condition is either at birth or within a few years of birth with blindness associated with nystagmus, strabismus photophobia and the oculodigital syndrome.

Signs

- *Early* – fundus may be normal but the electroretinogram (ERG) is extinguished.
- *Late* – arteriolar attenuation, optic atrophy, 'salt and pepper' changes, diffuse white spots and bone-spicule pigmentation.

Associations are keratoconus, keratoglobus and cataract.

Systemic features

- Psychomotor retardation and mental handicap.
- Deafness.
- Epilepsy.
- Renal anomalies.

CONE DYSTROPHY

Inheritance

Usually dominant.

Clinical features

Presentation of this *very rare* condition is between the first and third decades with impaired visual acuity, decreased vision in good illumination (day blindness), defective colour vision and nystagmus.

Signs

- *Early* – a 'bull's eye' macular lesion.
- *Late* – vascular attenuation, bone-spicule pigmentation and optic atrophy.

BEST'S VITELLIFORM MACULAR DYSTROPHY

Inheritance

Dominant.

Clinical features

This *rare* condition can be divided into five stages.

Stage 1 (previtelliform)

- Normal visual acuity.
- Normal fundus.
- Electro-oculogram (EOG) is reduced.

Stage 2 (vitelliform)

- Visual acuity is normal or slightly reduced.
- Fundus initially shows yellow (lipofuscin) spots at level of RPE, later followed by an egg-yolk macular lesion.

Stage 3 (pseudohypopyon)

- Visual acuity is slightly reduced.
- Part of vitelliform lesion becomes absorbed.

Stage 4 (vitelliruptive)

- Visual acuity is moderately reduced.
- Egg-yolk breaks up into a 'scrambled egg'.

Stage 5 (end-stage)

- Visual acuity is severely reduced.
- Fundus shows either a hypertrophic or an atrophic macular lesion.

STARGARDT'S DISEASE AND FUNDUS FLAVIMACULATUS

Inheritance

Usually recessive.

Clinical features of Stargardt's macular dystrophy

Presentation of this *uncommon* condition is between the first and second decades with gradual asymmetrical impairment of visual acuity.

Signs

- *Early* – non-specific macular mottling.
- *Late* – a 'snail-slime' or 'beaten-bronze' oval macular lesion which may be surrounded by yellow-white flecks.

Clinical features of fundus flavimaculatus

Presentation is later than in Stargardt's disease.

Signs

Prominent yellow flecks of various shapes throughout the posterior poles in one of the following patterns:

- Maculopathy without flecks.
- Maculopathy with perifoveal flecks.
- Maculopathy with diffuse flecks.
- Diffuse flecks without maculopathy.

ERG and *EOG* are abnormal in advanced cases.

ALBINISM

Complete oculocutaneous albinism

Inheritance

Usually recessive.

Cause

Inability to synthesize melanin (tyrosinase negative).

Clinical features

Complexion is pale skin and blond hair.

Eyes show lack of pigment which gives rise to:

- Photophobia due to blue irides which transilluminate completely.
- Reduced visual acuity and nystagmus due to foveal hypoplasia and hypopigmentation of the fundus.

Incomplete oculocutaneous albinism

Inheritance

Usually recessive.

Cause

Incomplete ability to synthesize melanin (tyrosinase positive).

Clinical features

- *Complexion* is variable.
- *Eyes* contain a variable amount of pigment.

Ocular albinism

Inheritance

Usually X-linked and occasionally recessive.

Clinical features

- *Complexion* is normal.
- *Eyes* are similar to oculocutaneous albinism.
- *Asymptomatic female carriers* may show iris transillumination and peripheral retinal granularity and depigmentation.

VITREORETINAL DEGENERATIONS

Wagner's syndrome

Inheritance

Dominant.

Clinical features

Signs

- *Vitreous* is optically empty and contains equatorial, translucent, circumferential membranes.
- *Retina* shows lattice-like radial perivascular pigmentation with foci of chorioretinal atrophy.
- *Associations* are myopia, cataract and strabismus.
- *Complications* are retinal detachment in Jansen's variant of Wagner's syndrome.

Stickler's syndrome

Inheritance

Dominant.

Ocular features

Signs

- *Vitreous* and *retina* are similar to Wagner's syndrome.
- *Associations* are myopia, cataract and open-angle glaucoma.
- *Complications* are frequent retinal detachment.

Systemic features

- *Orofacial* are midfacial flattening, cleft palate, micrognathia and glossoptosis.
- *Skeletal* are hyperextensibility and enlargement of joints.

Congenital retinoschisis

Inheritance

X-linked.

Clinical features

Signs

- *Maculopathy* is present in all cases and initially consists of a 'bicycle-wheel' pattern of radial striae centred on the fovea. It progresses to an atrophic lesion with reduced visual acuity.
- *Retinoschisis* (vitreous veils) are present in 50–70% of cases. The inner leaf is very thin and contains large oval defects.
- *Complications* are retinal detachment in 5%.

Exudative vitreoretinopathy (Criswick–Schepens syndrome)

Inheritance

Dominant.

Clinical features

Signs

- *Vitreous* shows membranes, fibrillary strands and vitreous base condensations.
- *Retina* is similar to retinopathy of prematurity with neovascular fibroproliferation with temporal dragging of the macula.
- *Complications* are occasional retinal detachment.

CHOROIDAL DYSTROPHIES

Choroideremia

Inheritance

X-linked recessive.

Clinical features

Presentation of this *very rare* condition is during the first decade with night blindness.

Signs

- *Early* – diffuse mottled depigmentation of the RPE.
- *Late* – enlarging midretinal patches of atrophy of choroid and RPE.
- Legal blindness between the second and fourth decades.
- *Female carriers* show peripheral retinal atrophy and mottling of the RPE.

Gyrate atrophy

Inheritance

Recessive.

Cause

An inborn error of *ornithine ketoacid aminotransferase* activity with increased levels of ornithine in the plasma, urine, CSF and aqueous.

Clinical features

Presentation of this *very rare* condition is during the first decade with night blindness.

Signs

- *Early* – midretinal circular patches of chorioretinal atrophy.
- *Late* – large areas of atrophy due to enlargement and coalescence of the patches.

- Legal blindness by the fifth decade.

Associations are myopia, vitreous degeneration and cataract.

Treatment

Pyridoxine (vitamin B_6) and a diet low in proteins and arginine.

Central areolar choroidal dystrophy

Inheritance

Recessive or dominant.

Clinical features

Presentation of this *rare* condition is during the fifth to sixth decades with impairment of visual acuity.

Signs

- Circumscribed atrophic macular lesions with prominent choroidal vessels.
- Legal blindness by the seventh decade.

Chapter 13

Intraocular tumours

TUMOURS OF THE UVEA

Choroidal melanoma

Clinical features

Incidence

- Most frequent tumour of the uvea.
- Extremely rare in Blacks.
- Occurs with increased frequency in congenital ocular melanosis (*naevus of Ota*).

Presentation of this *uncommon* tumour is usually during the sixth decade with unilateral visual impairment.

Signs

- Unilateral brown, black or amelanotic oval-shaped mass.
- *Associations* are orange lipofuscin pigment in retinal pigment epithelium (RPE), choroidal folds and hard yellow exudation.
- *Complications* are secondary exudative retinal detachment (very common), uveitis and haemorrhage (subretinal or vitreous).

Differential diagnosis

- Retinal detachment.
- Choroidal detachment.
- Metastatic tumour to the choroid.

- Localized choroidal haemangioma.
- Large choroidal naevus.
- Haemorrhagic detachment of the RPE.

Diagnostic tests

General medical evaluation

- To exclude a metastatic tumour to the choroid from the bronchus, breast, kidney and gastrointestinal tract.
- To exclude metastases from the choroidal tumour to the liver and lungs (liver function tests and chest X-ray).

Ultrasonography

- *A-scan* shows a high internal spike and low–medium internal reflectivity. It is useful in documenting increasing tumour thickness.
- *B-scan* shows acoustic hollowness, choroidal excavation and orbital shadowing. It is particularly useful in eyes with opaque media.

Fluorescein angiography

- May detect a 'double' circulation in small tumours.
- Useful in differentiating a tumour from a haemorrhagic RPE detachment.

^{32}P uptake

- *Positive* in malignant melanoma and metastases.
- *Negative* in benign tumours (haemangioma).

Transillumination – pigmented tumours and dense haemorrhage do not transilluminate.

Treatment

- *Enucleation* for large tumours with loss of vision.
- *Radioactive* plaques for small to medium-sized tumours.
- *Heavy ion irradiation* is an alternative to enucleation for large and medium-sized tumours.
- *Xenon arc photocoagulation* for selected small tumours away from fovea.

- *Local resection* (partial choroidectomy) for certain peripheral tumours.
- *Exenteration* if extrascleral extension is present.
- *Palliation* for metastatic disease.
- *Observation* of a small tumour if the diagnosis is uncertain or of a slow growing tumour in an only seeing eye.

Histological classification

- *Spindle cell A* – slender spindle-shaped cells with a flattened nucleus devoid of a nucleolus. The 5-year mortality is 5%.
- *Spindle cell B* – larger than a spindle A cell with a round or oval nucleus and a prominent nucleolus. The 5-year mortality is 15%.
- *Fascicular* consists of pallisading spindle A or spindle B cells. The 5-year mortality is 15%.
- *Epithelioid* – oval or round cells of variable size with a round nucleus and a prominent nucleolus. The cytoplasm is eosinophilic, the mitotic figures are many, and the degree of pigmentation is variable. The 5-year mortality is 70%.
- *Mixed cell* consists of both spindle and epithelioid cells. The 5-year mortality is 50%.
- *Necrotic* in which the cell type cannot be recognized. The 5-year mortality is 50%.

Prognostic factors

- *Cell type* – best for spindle A and worst for epithelioid.
- *Tumour size* – good for small and bad for large.
- *Intactness of Bruch's membrane* – bad if ruptured.
- *Age of patient* – worse if over 65 years.
- *Pigmentation* – worse if highly pigmented.
- *Diffuse growth* – bad because it consists of epithelioid cells.
- *Extrascleral extension* is very bad.

Ciliary body melanoma

Clinical features

Incidence – 15–20% of all uveal melanomas.

Presentation of this *rare* tumour is usually in the sixth to seventh decades in one of the following ways:

- Pressure on the lens giving rise to astigmatism, subluxation and a localized cataract.
- External features consisting of dilated episcleral (sentinel) blood vessels or an epibulbar mass.
- Anterior extension through the iris root.
- Posterior extension giving rise to secondary exudative retinal detachment.
- Anterior uveitis due to necrosis.
- Diffuse circumferential growth.

Differential diagnosis

- Congenital cyst.
- Medulloepithelioma (diktyoma).

Diagnostic tests

- *Slit-lamp biomicroscopy* with a contact lens to detect forward erosion into the iris root.
- *Transillumination* to differentiate a tumour from a cyst.
- *Ultrasonography* is useful for eyes with opaque media and in assessing the dimensions and extent of posterior extension.
- ^{32}P *uptake* to differentiate a benign from a malignant tumour.

Treatment

- Enucleation for a large tumour.
- Local resection (iridocyclectomy) for a small or medium-sized tumour.
- Radiotherapy with plaques or heavy ions in selected cases.

Iris melanoma

Clinical features

Incidence – 5–10% of uveal melanomas.

Presentation of this *rare tumour* is usually during the fifth decade.

Signs

- *Solitary tumour* (common) consists of a pigmented or non-pigmented inferior iris nodule.
- Associations are ectropion uveae, pupillary distortion and iris neovascularization.
- Complications are localized cataract and secondary glaucoma if extensive angle involvement is present.
- *Diffuse tumour* (rare) may cause a hyperchromic heterochromia.

Fluorescein angiography of a melanoma will show a separate blood supply and leakage.

Differential diagnosis

- Cyst.
- Large naevus.
- Granuloma.
- Leiomyoma.
- Metastasis.

Treatment

- Local resection for a localized tumour.
- Enucleation for a large or diffuse tumour.

Choroidal naevus

Clinical features

Presentation – this *common* tumour is usually discovered by chance in the eye of an adult.

Signs

- Usually asymptomatic flat or minimally elevated, oval or round, slate-grey lesion.
- Usually less than 3 mm in diameter.
- Associations are overlying drusen.
- Complications are rarely choroidal neovascularization.

Fluorescein angiography usually shows blockage of background choroidal fluorescence.

Differential diagnosis

- Small malignant melanoma.
- Congenital hypertrophy of RPE.

Treatment

Unnecessary, although suspicious large lesions should be photographed and watched.

Localized choroidal haemangioma

Clinical features

Presentation of this *very rare* tumour is usually in adult life with unilateral impairment of vision.

Signs

- Dome-shaped or placoid orange–red lesion at the posterior pole.
- Pressure on globe induces blanching of the tumour.
- Associations are secondary retinal cystoid degeneration and pigment mottling on the tumour surface.
- Complications – exudative retinal detachment (common).

Differential diagnosis

- Amelanotic malignant melanoma
- Metastasis.

Diagnostic tests

- *A-scan* ultrasound shows high initial spike and high internal reflectivity.
- *B-scan* shows a sharp anterior border with neither choroidal excavation nor orbital shadowing.
- *Fluorescein angiography* shows up large choroidal vessels with late staining.
- ^{32}P is negative.

Treatment

Usually unnecessary although photocoagulation may be required in the event of macular detachment.

Metastatic choroidal carcinoma

Primary sites

Most frequent are the bronchus and breast; occasional are the kidney, gastrointestinal tract and testis.

Clinical features

Presentation of this *common* tumour is with unilateral or bilateral impairment of vision.

Signs

- Solitary or multiple, unilateral or bilateral, creamy-white placoid oval lesions with ill-defined borders most commonly at the posterior pole.
- Associations are mottled pigment clumping.
- Complications – exudative retinal detachment.

Diagnostic tests

- Ultrasonography shows diffuse choroidal thickening.
- Fluorescein angiography has a variable appearance.
- ^{32}P is positive.

Treatment

Palliation with chemotherapy and external irradiation.

Osseous choristoma (choroidal osteoma)

Clinical features

Presentation of this *very rare* tumour is typically in young women with unilateral visual impairment although it may be discovered by chance.

Signs

- Usually unilateral, slightly elevated, orange–yellow lesion at the posterior pole with geographical and well-demarcated borders.
- Associations are diffuse mottling of the RPE and a fine vascular network.

Diagnostic tests

- Fluorescein angiography shows a diffuse mottled hyperfluorescence.
- ^{32}P is positive because the phosphorus is taken up by bone.
- Plain X-rays may show up the tumour.

Treatment

Unnecessary.

TUMOURS OF THE RETINA

Retinoblastoma

Inheritance

- *Familial* – 6% have a positive family history; inheritance is dominant with high but incomplete (70%) penetrance.
- *Sporadic* – 94%; of these 25% are *germinal* mutations which may pass on the tumour to the offspring, and 75% are *somatic* mutations which will not pass on the tumour.

Clinical features

Incidence – this *very rare* tumour affects 1 in 20 000 live births. It is the most common primary ocular malignancy in children.

Presentation is typically at about the age of 18 months in one of the following ways:

- Leukocoria in 70%.
- Strabismus in 20%.
- Secondary glaucoma and buphthalmos (rare).
- Proptosis which may mimic orbital cellulitis (rare).

- Anterior chamber inflammation which may mimic anterior uveitis (rare).
- On routine examination of a patient at risk (rare).

Signs (30% bilateral)

- *Endophytic tumour* grows into the vitreous. It is white or pearly pink and it may contain fine blood vessels on its surface.
- *Exophytic tumour* grows in the subretinal space and causes a retinal detachment.

Differential diagnosis of leukocoria

- Cataract.
- Persistent hyperplastic primary vitreous.
- Advanced retinopathy of prematurity.
- Coats' disease.
- Toxocariasis.
- Retinal dysplasia.
- Retinal astrocytoma.

Diagnostic tests

Indirect ophthalmoscopy is usully sufficient in most cases.

For secondary calcification – plain X-rays, B-scan ultrasonography and CT scan.

Miscellaneous

- Aqueous humour assay for lactic dehydrogenase.
- Assays for carcinoembryonic antigen.
- Lumbar puncture and bone marrow aspiration for metastases.

Treatment

- *Enucleation* is the method of choice for a large tumours in the first eye.
- *Radiotherapy* by external beam for large tumours and cobalt plaques for small tumours.

- *Xenon arc photocoagulation* for small tumours not invading either the macula or the optic nerve.
- *Cryotherapy* for small peripheral tumours.
- *Systemic chemotherapy* for metastatic disease which may involve the skull, orbit, long bones, viscera, spinal cord and lymph nodes.

Prognostic factors

Overall mortality is 15–20%.

Optic nerve involvement

- If tumour is present beyond the transection – 65% mortality.
- If the lamina cribrosa is involved – 15% mortality.
- If the optic nerve is uninvolved – 8% mortality.

Size and location – prognosis is best for small posterior tumours.

Cellular differentiation

- Well differentiated – 8% mortality.
- Undifferentiated – 40% mortality.

Retinal astrocytoma

Ocular features

Presentation – this *very rare* tumour is asymptomatic and is usually discovered on fundus examination in a patient suspected of having *Bourneville's disease* (tuberous sclerosis).

Signs (15% bilateral and may be multiple)

- *Early* – semitranslucent roundish lesions of retina or the optic disc.
- *Late* – tumour becomes calcified and mulberry-like.

Treatment is unnecessary.

Systemic features of Bourneville's disease

Incidence – 50% of patients with Bourneville's disease have retinal astrocytomas.

Skin lesions

- Nodular fibroangiomas ('adenoma sebaceum').
- Achromic naevi.
- Café-au-lait spots.
- Shagreen patches.

Central nervous system

- Astrocytic hamartomas in the brain in 80% (may cause epilepsy).
- Mental handicap in 60%.

Visceral hamartomas of the heart, kidneys, subungual etc.

Retinal capillary haemangioma

Inheritance

Dominant.

Ocular features

Presentation of this *very rare* tumour is in early adult life with initially unilateral visual impairment.

Signs (50% bilateral)

- *Early* – single or multiple microaneurysm between arteriole and venule in the retina or the optic disc.
- *Late* – small red nodule becomes larger, round, orange–red tumour with dilatation of the supplying artery and the draining vein.
- *Complications* are hard exudates at macula, haemorrhage and retinal detachment.

Treatment is with photocoagulation, cryotherapy or diathermy.

Systemic features

Incidence – 25% of patients with haemangiomas will have associated von Hippel–Lindau disease.

Central nervous system haemangioblastomas of the cerebellum, medulla, pons and spinal cord.

Visceral lesions

- Cysts of the kidneys, pancreas, liver, epididymis, ovary and lungs.
- Hypernephroma and phaeochromocytoma.

Polycythaemia.

Retinal cavernous haemangioma

Ocular features

Presentation – this *very rare* tumour is usually asymptomatic and is discovered by chance.

Signs

- Unilateral, congenital, thin-walled, saccular aneurysms similar to a cluster of grapes of the retina or optic nerve head.
- May have a meniscus between plasma and red cells.

Treatment is unnecessary.

Systemic features

Some patients have similar lesions of the skin and CNS.

Retinal racemose haemangioma

Ocular features

Presentation of this *very rare* tumour is with unilateral visual impairment.

Signs

- Unilateral congenital arteriovenous malformation of the retina or the optic disc with grossly enlarged and tortuous vessels.
- Complications are occasional exudation and haemorrhage.

Treatment is unnecessary.

Systemic features

Some patients have similar lesions in the CNS, maxilla, mandible, orbit, and the facial skin (Wyburn–Mason syndrome).

Chapter 14

Strabismus

EXAMINATION TECHNIQUES

Corneal reflection tests

- *Hirschberg* is a rough test without the use of prisms.
- *Krimsky* prisms are used until the corneal reflexes are symmetrical.

Cover tests

- *Cover–uncover* for a manifest deviation (heterotropia).
- *Alternate cover* for a latent deviation (heterophoria).
- *Prism and alternate cover* to measure the total deviation.

Dissimilar image tests

- *Maddox wing* for heterophoria at near.
- *Maddox rod* for heterophoria at distance.
- *Hess test* for a paretic deviation.

Tests for sensory anomalies

- *Worth's four dot* for suppression and anomalous retinal correspondence (ARC).
- *After-image* to demonstrate the visual direction of the two foveae.
- *Bagolini striated glassses* for ARC.
- *Synoptophore* for grades of binocular vision, suppression, amblyopia and ARC.

Tests for stereopsis

- *Titmus* – booklet with a large (Wirt's) fly, circles and animals viewed with polaroid glasses.
- *TNO* – seven plates with various shapes viewed with polaroid glasses.

CHILDHOOD ESOTROPIAS

Classification

Infantile

Accommodative

- Refractive
- Non-refractive
- Mixed

Non-accommodative

- Stress induced
- Sensory deprivation
- Divergence insufficiency
- Spasm of the near reflex
- Consecutive
- Sixth nerve palsy

Infantile esotropia

Clinical features

Presentation of this *common* condition is within the first 6 months of life.

Signs

- Angle is large with crossed fixation on side gaze.
- Nystagmus is frequent.
- Refraction is normal for the patient's age (+1.50 D).

Management

Initial

- Refraction and fundus examination.
- Treatment of amblyopia if present.

Subsequent – surgery between the age of 12 and 18 months (bilateral medial rectus recessions).

Late complications

- Inferior oblique overaction.
- Dissociated vertical deviation (DVD).
- Amblyopia.

Accommodative esotropia

Clinical features

Presentation of this *very common* condition is typically at 2.5 years (range 6 months–7 years).

Signs of refractive esotropia

- Angle is virtually the same for near and distance.
- Refraction shows high hypermetropia (+4 to +7 D).
- Accommodative-convergence to accommodation (AC/A) ratio is normal.

Signs of non-refractive esotropia

- Angle is marked for near; nil or small for distance.
- Refraction usually shows emmetropia.
- AC/A ratio is high.

Signs of mixed esotropia

- Angle is more for near than for distance.
- Refraction shows high hypermetropia.
- AC/A ratio is high.

Management

Refraction

Cyclopentolate usually is adequate in most children:

- 0.5% for patients under 3 months and 1% thereafter.
- Two drops 5 min apart and refract after 30 min.

Atropine in high hypermetropia or very pigmented irides:

- 0.5% for patients under 3 months and 1% thereafter.
- Three times a day for 3 days.

Prescription of spectacles

- Under 6 years give the full cycloplegic refraction.
- Over 6 years give the maximal 'plus' tolerated by the patient.
- Bifocals for high AC/A ratio.

Fundus examination to exclude organic disease.

Miotics as short term for patients with a high AC/A ratio.

Treatment of amblyopia

- Occlusion.
- Atropine.
- Manipulation of glasses.
- Pleoptics.
- Stripe (CAM) therapy.

Surgery if the deviation is not controlled by spectacles.

Non-accommodative esotropia

- *Stress induced* due to a breakdown of previously effective fusion usually requires surgery.
- *Sensory deprivation* due to monocular organic lesions may require cosmetic surgery.
- *Divergence excess* in which the angle is larger for distance than for near may require surgery.
- *Spasm of near reflex* occurring in hysterical patients is treated with cycloplegics.

CHILDHOOD EXOTROPIAS

Classification

Intermittent

Constant

- Congenital
- Decompensated intermittent
- Sensory deprivation
- Consecutive

Intermittent exotropia

Clinical features

Presentation of this *common* condition is at about the age of 2 years.

Signs

- Deviation fluctuates between phoria and tropia.
- Refraction shows emmetropia or myopia.
- Amblyopia is rare.

Management

Non-surgical

- Correct myopia if present.
- Orthoptic therapy with antisuppression, diplopia awareness and to improve fusional convergence.

Surgery is frequently required by the age of 5 years.

Congenital exotropia

Clinical features

Presentation of this *uncommon* condition is at birth.

Signs

- Angle is large and constant.
- Left eye may be used in left gaze and the right eye in right gaze.
- Refraction is normal for age.
- Amblyopia is uncommon.

Treatment

Surgery is performed at about the age of 18 months.

Miscellaneous constant exotropias

- *Decompensated intermittent* is usually treated surgically.
- *Sensory deprivation* due to disruption of binocular reflexes by lesions acquired after the age of 5 years may require cosmetic surgery.
- *Consecutive* follows overcorrection of an esotropia and usually requires further surgery.

SPECIAL OCULAR MOTILITY DEFECTS

Duane's retraction syndrome

Clinical features

Presentation of this *uncommon* condition is at birth.

Signs (bilateral in 15%)

- *Type 1* – abduction is limited but adduction is normal.
- *Type 2* – abduction is normal but adduction is limited.
- *Type 3* – both abduction and adduction are limited.
- On attempted adduction there is retraction of the globe and narrowing of the palpebral fissure.
- On attempted abduction the globe assumes its normal position and the palpebral fissure opens.

Treatment

Usually unnecessary because the eyes are straight in the primary position and amblyopia is rare.

Brown's superior oblique tendon sheath syndrome

Causes

- Congenital.
- Acquired due to trauma.

Clinical features

Presentation of this *rare* condition is at birth except in the case of acquired traumatic cases.

Signs (usually unilateral)

- Elevation in adduction is limited.
- Elevation in abduction is normal.
- Overaction of the superior oblique is absent.
- In the primary position the eyes are straight.

Treatment

For congenital cases this is usually unnecessary although acquired cases may require surgery.

SURGICAL PROCEDURES

Weakening procedures

- Recession.
- Marginal myotomy to further weaken a recessed muscle.
- Myectomy, usually of an overactive inferior oblique.

Strengthening procedures

- Resection.
- Tucking, usually of the superior oblique in a fourth nerve palsy.
- Advancement to enhance the action of a recessed muscle.

To change direction of muscle action

- Vertical transposition of horizontal recti for 'A' and 'V' patterns.
- Posterior fixation suture (Faden) for dissociated vertical deviation.
- Hummelsheim procedure for sixth nerve palsy.
- Jensen's procedure for sixth nerve palsy.

Adjustable sutures are used mainly for paretic deviations in adults.

Chapter 15

Neuro-ophthalmology

ACQUIRED OPTIC NERVE DISORDERS

Classification

Optic neuritis

- Retrobulbar
- Optic papillitis
- Neuroretinitis

Anterior ischaemic optic neuropathy

- Arteritic
- Non-arteritic

Toxic

- Toxic amblyopia
- Drug induced

Leber's optic neuropathy

Disc oedema

Tumours

- Glioma
- Meningioma

Optic neuritis

Causes

Idiopathic – most frequent.

Demyelination

- *Incidence* – optic neuritis is the presenting feature in 25% of patients with multiple sclerosis (MS) and occurs in 70% of established cases.
- *Risk factors* of subsequent MS are recurrent attacks, winter onset, HLA-DR2 positive and Uhtoff's phenomenon (vision worsens when body temperature rises or during physical exercise).

Postviral is the most common type in children and is usually bilateral.

Granulomatous due to sarcoid, tuberculosis or syphilis is rare.

Spread of infection from adjacent structures.

Clinical features of idiopathic optic neuritis

Presentation of this *common* condition is usually during the third and fourth decades with a unilateral acute onset of monocular visual loss, frequently associated with periocular discomfort made worse on ocular movement.

Clinical course – visual impairment is progressive and maximal at the end of the second week and usually recovers after 4–6 weeks.

Signs

- Visual acuity is variably impaired.
- Relative afferent pupillary conduction defect is present.
- Colour vision is impaired even in mild cases.
- Light brightness appreciation is decreased.
- Fundus – *retrobulbar neuritis* (normal), *papillitis* (disc oedema and vitreous cells), *neuroretinitis* (disc oedema and macular star).

Investigations

- *In typical* cases are usually unnecessary.
- *In atypical* cases – skull X-rays, CT scanning, magnetic resonance imaging (MRI) and visual evoked response (VER).

Treatment

Usually unnecessary although steroids speed up the recovery of vision but do not affect the final visual outcome.

Non-arteritic anterior ischaemic optic neuropathy

Causes

- Atherosclerosis frequently associated with hypertension.
- Small or absent optic cups are predisposing factors.

Clinical features

Presentation of this *uncommon* condition is during the sixth and seventh decades with a unilateral, acute onset of altitudinal visual loss unassociated with premonitary symptoms.

Clinical course – maximal visual loss occurs at the onset and any subsequent improvement is rare. The fellow eye becomes involved within months or years in 30% of cases.

Signs

- Visual acuity – mild to moderate impairment.
- Fundus shows diffuse or sectorial disc oedema with either pallor or hyperaemia and peripapillary splinter-shaped haemorrhages.
- Visual fields typically show an inferior altitudinal defect.

Treatment

None effective but hypertension should be treated and the possibility of arteritis excluded.

Arteritic anterior ischaemic optic neuropathy

Causes

- Giant cell arteritis is by far the most common.
- Collagen vascular disorders (systemic lupus erythematosus and polyarteritis nodosa).

Ocular features

Presentation of this *uncommon* condition is during the seventh and eighth decades with a usually unilateral, acute onset of very severe loss of vision which may be preceded by transient obscurations.

Clinical course – maximal visual loss occurs at the onset and any subsequent improvement is extremely uncommon. Without treatment the fellow eye becomes involved within weeks in 65% of cases.

Signs

- Visual acuity is severely impaired (counting fingers and hand movements).
- Fundus shows diffuse disc oedema and pallor with peripapillary splinter-shaped haemorrhages.

Systemic features

- Headache and scalp tenderness.
- Jaw claudication.
- Polymyalgia rheumatica.
- Malaise, weight loss, night sweats and fever.

Investigations

- Erythrocyte sedimentation rate (ESR) is frequently very high.
- C-reactive protein is invariably raised.
- Temporal artery biopsy to confirm the diagnosis.

Treatment

- Hydrocortisone 250 mg intravenously.
- Oral prednisone as follows: 80 mg for 3 days; 60 mg for 3 days; 40 mg for 4 days, then reduce by 5 mg weekly for 6 weeks; 10 mg for 3 months, then gradually reduce according to symptoms and level of ESR.

Toxic amblyopia

Cause

Heavy drinking and pipe smoking with a protein-deficient diet.

Clinical features

Presentation of this *rare* condition is during adult life with a gradual, bilateral but asymmetrical impairment of colour vision and visual acuity.

Signs

- Visual acuity is variably impaired.
- Fundus initially is normal and later may show optic atrophy.

- Visual fields show centrocaecal scotomata which are larger for red than white targets.

Treatment

Hydroxycobalamin 1000 units weekly for 10 weeks.

Drug-induced optic neuropathies

- Ethambutol.
- Chloramphenicol.
- Isoniazid.
- Streptomycin.

Leber's optic neuropathy

Inheritance

Exact mechanism is unknown but 85% of patients are male.

Clinical features

Presentation of this *very rare* condition is during the third decade with unilateral acute visual loss.

Clinical course is progressive and visual loss is permanant. The fellow eye becomes involved within weeks or months.

Signs

- Fundus shows mild disc oedema and hyperaemia with irregular dilatation of the pre- and peripapillary capillaries and glistening peripapillary retinal nerve fibres.
- Visual fields show a centrocaecal scotoma.

Treatment

None effective.

Papilloedema

Causes

Raised intracranial pressure by:

- Blockage of the ventricular system by congenital or acquired lesions.

- Obstruction of CSF absorption by the arachnoid villi.
- Space-occupying lesions.
- Hypersecretion of CSF by a choroid plexus tumour (rare).

Clinical features

Early

- Symptoms – absent.
- Visual acuity – normal.
- Fundus – superior and inferior disc margins are indistinct and hyperaemic with blurring of the peripapillary nerve fibre layer and loss of previous spontaneous venous pulsation.
- Blind spot is normal.

Established

- Symptoms – transient obscurations may be present.
- Visual acuity may be reduced by macular exudates.
- Fundus – disc surface is elevated, cup is obliterated and the traversing blood vessels are obscured. The disc is surrounded by flame-shaped haemorrhages and cotton-wool spots, and the veins are engorged. An incomplete macular star may be present.
- Blind spot is enlarged.

Vintage

- Visual acuity – variable.
- Fundus – optic disc looks like a champagne cork, but haemorrhages and exudates are absent.

Secondary optic atrophy

- Visual acuity – severely impaired.
- Fundus – optic disc is white and has indistinct margins.

Other causes of disc swelling

Vascular

- Central retinal vein occlusion.
- Malignant hypertension.
- Anterior ischaemic optic neuropathy.

Inflammatory

- Optic papillitis.
- Uveitis.

Compression

- Optic nerve tumours.
- Raised intraorbital pressure.

Infiltration

- Granulomata.
- Tumours.

Ocular hypotony.

Glioma of the optic nerve

Clinical features

Presentation of this *very rare* tumour is between the ages of 4 and 8 years with unilateral gradual visual loss and proptosis.

Signs

- Visual acuity is impaired.
- Fundus shows disc swelling or atrophy.
- Variable proptosis is present.
- Neurofibromatosis is present in 55% of cases.

Investigations

- Plain X-rays show uniform enlargement of the optic foramen.
- CT and ultrasound show enlargement of the optic nerve.

Treatment

- Surgery – local resection with preservation of the globe.
- Radiotherapy if the tumour is beyond surgical excision.

Optic nerve sheath meningioma

Clinical features

Presentation of this *very rare* tumour is typically in middle-aged women with unilateral gradual visual loss.

Signs

- Visual acuity is impaired.
- Fundus typically shows opticociliary shunt vessels on the optic disc.
- Restriction of ocular motility due to splinting of the optic nerve may be present.
- Proptosis is a feature of advanced tumours.

Investigations

- Plain X-rays show increased bone density.
- CT and ultrasound show enlargement of the optic nerve.

Treatment

Surgery or radiotherapy.

CONGENITAL OPTIC DISC ANOMALIES

Optic disc drusen (hyaline bodies)

Clinical features

Prevalence – uncommon (1:300 patients) and familial.

Signs (bilateral)

Buried drusen (in children) may mimic early bilateral disc swelling. The distinguishing features are:

- Optic cup is absent.
- Disc colour is pink or yellow.
- Margins are lumpy and indistinct.
- Anomalous branching of blood vessels.

- Veins are not dilated.
- Spontaneous venous pulsation is present in 80%.
- Peripapillary striations are normal.

Exposed drusen (during early teens) appear as pearly irregularities on the disc surface.

Optic disc pit

Clinical features

Prevalence – uncommon.

Signs (unilateral).

- Visual acuity is usually normal in absence of macular detachment.
- Fundus shows a dark round or oval inferotemporal pit in a large disc.
- Visual fields are normal.
- Complications are serous detachment of the macula in 50% of cases.

Optic disc coloboma

Clinical features

Prevalence – rare.

Signs (usually unilateral)

- Visual acuity is always reduced.
- Fundus shows a large inferior disc excavation which may mimic glaucomatous cupping.
- Visual fields show a superior defect.

Tilted disc

Clinical features

Prevalence – common.

Signs (frequently bilateral)

- Visual acuity is normal.
- Fundus shows an oval-shaped disc with an inferior crescent and situs inversus. Inferonasal retinal hypopigmentation is common.
- Myopia is very common.
- Visual fields frequently show a superotemporal defect.

Optic nerve hypoplasia

Clinical features

Prevalence – rare.

Signs (60% bilateral)

- Visual acuity is variably reduced.
- Fundus shows a small and grey disc surrounded by a yellow halo (double-ring sign) with blood vessels of normal calibre.
- Visual fields show variable defects.

Associations

- Aniridia.
- Nystagmus.
- Absent foveal reflex.
- Small optic canals.
- Midfacial anomalies.
- CNS anomalies – septo-optic dysplasia (de Morsier's syndrome), hydrancephaly and anencephaly.
- Maternal – diabetes or drugs during pregnancy.

Morning glory syndrome

Clinical features

Prevalence – very rare.

Signs (unilateral)

- Visual acuity is severely reduced.
- Fundus shows enlargement and excavation of the optic nerve head with persistent hyaloid remnants within its base and a radial pattern of emerging blood vessels.
- Retinal detachment is common.

CHIASMAL DISORDERS

Pituitary adenoma

Clinical features

Presentation of this *uncommon* tumour is in adults with non-specific headache and blurred vision.

Signs

- Visual fields – chiasmal compression from *below* initially involves the crossing inferonasal fibres giving rise to upper temporal defects which spread anticlockwise in the left eye and clockwise in the right eye.
- Diplopia from lateral extension into the cavernous sinus may occur.
- See-saw nystagmus of Maddox (rare).

Radiological features

The destruction starts posteriorly and spreads anteriorly as follows:

- Erosion of the dorsum sellae giving rise to the 'double floor' sign.
- Erosion of the anterior clinoids.
- Complete destruction of all anatomical landmarks.

Treatment

- Surgery – transfrontal approach for large tumours and trans-sphenoidal for small tumours.
- Bromocriptine shrinks a prolactin-secreting tumour.
- Radiotherapy.

Craniopharyngioma

Clinical features

Presentation

- In children this *uncommon* tumour interferes with hypothalamic function causing dwarfism, delayed sexual development and obesity.
- In adults the tumour presents with defects in visual fields.

Visual fields

Chiasmal compression from *above* and *behind* initially involves the upper nasal fibres giving rise to inferotemporal defects which spread clockwise in the left eye and anticlockwise in the right eye.

Radiological features

- Initially spreading apart of the anterior and posterior clinoids and then erosion of the dorsum sellae.
- Suprasellar calcification is present in some cases.

Treatment

Surgery but recurrences are common.

Meningioma

Clinical features

Presentation of this *uncommon* tumour is typically in a middle-aged female with headache and defects in visual field.

Visual field defects depend on the location of the tumour:

- From tuberculum sellae – compression of the junction of the chiasm with the optic nerve giving rise to an ipsilateral central scotoma and a contralateral upper temporal defect (junctional scotoma).
- From sphenoidal ridge – optic nerve compression.
- From olfactory groove – optic nerve compression associated with impaired sense of smell.

Radiological features

Hyperostosis is frequently seen on plain X-rays.

Treatment

Surgery.

THIRD, FOURTH AND SIXTH NERVE DISEASE

Third nerve palsy

Clinical features

Benedikt's syndrome

- *Location* of the lesion is the dorsal part of fasciculus as it passes through the red nucleus.
- *Signs* are an ipsilateral third nerve palsy associated with contralateral ataxia and flapping tremor.

Weber's syndrome

- *Location* of the lesion is the ventral part of fasciculus as it passes through the cerebral peduncle.
- *Signs* are an ipsilateral third nerve palsy associated with a contralateral hemiplegia.

Isolated complete third nerve palsy

- Weakness of the levator causes ptosis.
- Unopposed action of the lateral rectus causes abduction.
- Because the superior oblique is intact there is intorsion on attempted down-gaze.
- Weakness of the medial rectus causes limited adduction.
- Weakness of the superior rectus causes limited elevation.
- Weakness of the inferior rectus causes limited depression.
- Parasympathetic palsy causes motor involvement of pupil and defective accommodation.

Important causes of an isolated third nerve palsy

- Vascular (pupil is usually spared), hypertension, diabetes and atherosclerosis.
- Trauma which may result in aberrent regeneration.
- Aneurysm at the junction of the posterior communicating with the internal carotid artery (pupil is usually involved).
- Tumours (pupil is usually involved).

Fourth nerve palsy

Important anatomical facts

- *Longest* and *slenderest* of all cranial nerves.
- Only cranial nerve to emerge *dorsally* from the brain.
- Only completely *crossed* cranial nerve.

Clinical features of a complete fourth nerve palsy

- Hyperdeviation due to paralysis of the superior oblique muscle which is more marked with the head tilted to same shoulder (Bielschowsky's head tilt test) and which is compensated for by depression of the chin.
- Excyclotorsion which is compensatory by a head tilt to the opposite shoulder
- Limited depression in adduction.

Important causes of an isolated fourth nerve palsy

- Trauma usually causes bilateral palsies.
- Vascular.

Sixth nerve palsy

Clinical features

Foville's syndrome

- *Location* of the lesion is the dorsal pons as the fasciculus passes through the pontine paramedial reticular formation.
- *Signs* are an ipsilateral sixth nerve palsy combined with a gaze palsy, ipsilateral facial weakness and analgesia, Horner's syndrome and deafness.

Millard–Gubler syndrome

- *Location* of the lesion is ventral involving the pyramidal tract.
- *Signs* are an ipsilateral sixth nerve palsy and a contralateral hemiplegia.

Isolated complete sixth nerve palsy

- Weakness of the lateral rectus with no abduction beyond the midline.
- Unopposed action of the medial rectus causes an esotropia in the primary position.
- Compensatory face turn to the same side.

Important causes of an isolated sixth nerve palsy

- Vascular.
- Trauma.
- Aneurysm.
- Tumours.

CAROTID–CAVERNOUS FISTULA

Direct fistula

Causes

- Basal skull fracture tearing the artery.
- Spontaneous rupture of an aneurysm or an atherosclerotic artery.

Clinical features

Presentation of this *uncommon* condition is usually in elderly patients with sudden onset of headache, a noise in the head and diplopia.

Signs

- Engorged episcleral blood vessels causing chemosis, redness and raised IOP.

- Proptosis which is pulsatile and associated with both a bruit and a thrill which are abolished by ipsilateral carotid compression.
- Ophthalmoplegia involving the sixth nerve ± third and fourth nerves.
- Anterior segment ischaemia in severe cases.
- Fundus shows vascular engorgement and haemorrhages.

Treatment

Surgery if spontaneous closure does not occur.

Indirect fistula (dural shunt)

Causes

- Congenital.
- Spontaneous after minor trauma or straining.

Clinical features

These are similar to but much less severe than in a direct fistula.

Treatment

Usually is unnecessary.

ABNORMAL PUPILLARY REACTIONS

Total afferent conduction defect (amaurotic pupil)

Cause

A complete optic nerve lesion.

Clinical features

- Unilateral.
- Size – both pupils are equal.

- Light reflex: when the affected eye is stimulated neither pupil reacts; when the normal eye is stimulated both pupils react normally.
- Near reflex is normal in both eyes.

Relative afferent conduction defect (Marcus Gunn pupil)

Cause

An incomplete optic nerve lesion or severe retinal disease.

Clinical features

- Similar to a total defect but more subtle.
- Difference between the pupillary reactions is enhanced by the *swinging-flashlight test* in which the abnormal pupil dilates instead of constricting when stimulated.

Argyll Robertson pupil

Cause

Neurosyphilis.

Clinical features

- Usually bilateral but asymmetrical.
- Size – pupils are small and irregular.
- Light reflex is absent or very sluggish.
- Near reflex is normal (light–near dissociation).
- Very difficult to dilate.

Holmes–Adie (tonic) pupil

Cause

Denervation of the postganglionic supply to the musculus sphincter pupillae and the ciliary muscle which may follow a viral illness.

Clinical features

- Unilateral in 80%.
- May be associated with diminished tendon reflexes.
- Size – large and regular.

- Light reflex is absent or very slow.
- Near reflex – constriction is very slow and tonic and is associated with vermiform movements of the iris. Re-dilatation is also very slow.
- Accommodation is slow.

Pharmacological test

Mecholyl 2.5% or pilocarpine 0.125% is instilled into both eyes:

- Normal pupil will not constrict.
- Abnormal pupil will constrict.

Oculosympathetic palsy (Horner's syndrome)

Cause

Disruption of the sympathetic pathways.

Clinical features

- Usually unilateral.
- Mild ptosis due to weakness of Müller's muscle.
- Slight elevation of inferior eyelid due to weakness of the inferior tarsal muscle.
- Miosis due to unopposed action of the sphincter pupillae.
- Pupillary reactions are normal to light and near.
- Reduced ipsilateral sweating, but only if the lesion is *below* the superior cervical ganglion.
- Amplitude of accommodation is reduced.
- Heterochromia is occasionally present if the lesion is *congenital.*

Pharmacological test

To confirm Horner's syndrome instil 4% cocaine into both eyes:

- Normal pupil will dilate.
- Horner's pupil will not dilate.

Preganglionic vs postganglionic lesion – instil 1% hydroxyamphetamine into both eyes:

- In a *preganglionic* lesion both pupils will dilate.
- In a *postganglionic* lesion the Horner's pupil will not dilate.

NYSTAGMUS

Classification

Pendular – equal velocity in each direction:

- Horizontal
- Vertical
- Oblique
- Rotary

Jerky – has a fast and a slow phase with the direction of the nystagmus designated by the direction of the fast component as follows:

- Right
- Left
- Up
- Down
- Rotary

Mixed – pendular nystagmus in the primary position and jerky nystagmus on lateral gaze

Clinical types

Physiological

- End point.
- Optokinetic.
- Vestibular.

Sensory deprivation (ocular)

It is always present if vision is lost prior to the age of 2 years and is usually pendular and horizontal.

Motor imbalance

Congenital

- Inheritance is X-linked recessive or autosomal dominant.
- Type is usually jerky and horizontal.

- Present at birth.
- Duration – persists throughout life.

Spasmus nutans

- Type – asymmetrical, pendular, fine, rapid and usually horizontal.
- Associated abnormal head posture and head nodding.
- Presents between fourth and twelfth months.
- Duration is up to the age of 3 years.

Latent

- Type – jerky bilateral nystagmus when one eye is covered. It may occur in infantile esotropia.
- Presents in early childhood.

Ataxic occurs in the abducting eye of a patient with internuclear ophthalmoplegia.

Downbeat

- Type – fast phase is downwards.
- Causes are lesions of cervicomedullary junction at the foramen magnum.

Upbeat

- Type – fast phase is upwards.
- Causes are drugs and lesions of the posterior fossa.

Convergence–retraction

- Type – jerky, fast phase causes convergence and retraction of the globe.
- Causes are lesions of the pretectal area (Parinaud's syndrome).

See-saw nystagmus of Maddox

- Type – one eye rises and intorts whilst the other falls and extorts.
- Cause is bitemporal hemianopia usually from a chiasmal lesion.

Periodic alternating

- Type – jerk with rhythmic changes in amplitude and direction.
- Causes are brainstem lesions.

MYOPATHIES OF EXTRAOCULAR MUSCLES

Myasthenia gravis

Classification

- Progressive
- Remittent
- Ocular

Clinical features

Presentation

- This *uncommon* condition typically presents in females during third and fourth decades with excessive fatiguability of skeletal muscles.
- Ocular involvement is present in 90% of cases and is the presenting feature in 60%.

Signs

- Ptosis is bilateral but asymmetrical and worse towards the end of the day or when tired.
- Diplopia is frequently vertical although all or part of the extraocular muscle may be affected.

Special investigations

Tensilon test is performed as follows:

1. Measure amount of ptosis or motility defect (Hess test).
2. Inject atropine 0.3 mg i.v.
3. Inject test dose of 0.2 ml edrophonium (Tensilon) (2 mg) i.v.
4. Inject the remaining 0.8 ml (8 mg) after 60 seconds provided there is no hypersensitivity.
5. Take measurement quickly because the effect of edrophonium lasts only 5 minutes.

Other tests

- Electromyography may be combined with Tensilon test.
- Antibodies to anti-acetylcholine receptors are present in 90% of cases.
- Chest X-ray may show a thymoma.

Treatment

- Long-acting anticholinesterase drugs (pyridostigmine).
- Systemic steroids.
- Cytotoxic agents (azathioprine, cyclophosphamide).
- Plasmapheresis to remove antibodies.
- Thymectomy.

Chronic progressive external ophthalmoplegia (ocular myopathy)

Clinical features

Presentation of this *very rare* condition is in adults with an insidious onset of slowly progressive ptosis and ophthalmoplegia.

Signs

- Ptosis.
- Ophthalmoplegia but *no diplopia* because muscle involvement is symmetrical.

Associated features

- Primary type has no other features.
- Kearns–Sayre syndrome (see Chapter 12).
- Oculopharyngeal dystrophy with involvement of the pharyngeal muscles and wasting of the temporalis.

Dystrophia myotonica

Systemic features

This *rare* condition is characterized by:

- Excessive contractility and difficult relaxation of skeletal muscles.

- Hypogonadism.
- Baldness.
- Cardiac anomalies.

Ocular features

- Ptosis and weakness of facial muscles giving rise to a mournful expression.
- Presenile Christmas tree cataracts.
- Pigmentary changes at the macula or retinal periphery.
- Light–near dissociation of the pupil.

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