Third edition

Notes & Notes

For MRCP part 1 & 2

By

Dr. Yousif Abdallah Hamad

Ophthalmology

Updated

2022

Chapter 12 Ophthalmology

Acute angle closure glaucoma (AACG)

Definition: sudden and sharp **increase in intraocular pressure (IOP)** caused by an obstruction of aqueous outflow (most commonly as a result of an occlusion of the iridocorneal angle)

Pathophysiology: blockage of the trabecular meshwork → ↓ drainage of aqueous humor from the eye → ↑ IOP

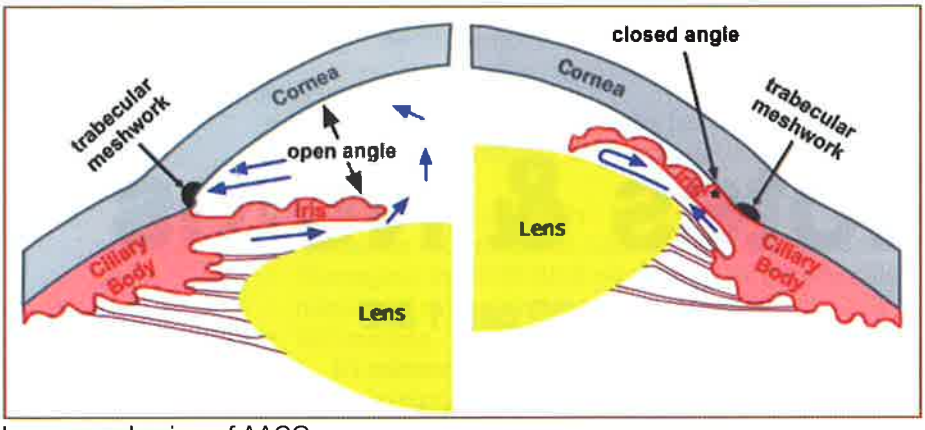


Image: mechanism of AACG

Risk factors

- hypermetropia (long-sightedness)
- lens growth associated with age
- neovascular glaucoma (new blood vessels grow into the angle of the eye and block the aqueous outflow). Vascular endothelial growth factor (VEGF) is a growth factor protein that works by stimulating angiogenesis. Hence, inhibiting VEGF (e.g. Bevacizumab) can stop the progression of neovascularization.
- **Drugs:** pupillary dilatation by mydriatic drops, **anticholinergics (e.g., atropine) and tricyclic antidepressants**

Features

- Sudden onset of symptoms
- Unilaterally inflamed, reddened, and severely painful eye (hard on palpation)
- Blurred vision and halos seen around light
- Mid-dilated, irregular, unresponsive pupil
- Frontal headaches, vomiting, nausea
- Complications: rapid permanent vision loss due to ischemia and atrophy of the optic nerve symptoms worse with mydriasis (e.g. watching TV in a dark room)

Diagnosis

- Tonometry: → Elevated IOP (> 21 mm Hg)
- **Gonioscopy: the gold-standard diagnostic test** → Narrowing/closure of the iridocorneal angle.

Management

- urgent referral to an ophthalmologist
- Acetazolamide intravenously, along with a topical beta-blocker and a topical alpha-agonist

- Reducing aqueous secretions with acetazolamide and inducing pupillary constriction with topical pilocarpine.
- **Pilocarpine should not be the initial treatment as it is ineffective at pressures above 40 mmHg.**
- Mannitol is typically reserved for refractory cases, not responding to the initial medical treatment.

Top Tips

Acute angle closure glaucoma is associated with hypermetropia, where as primary open-angle glaucoma is associated with myopia

Treatment of acute glaucoma - acetazolamide + pilocarpine

Do not use mydriatic drugs (e.g., atropine and epinephrine) during ophthalmologic examination in patients with acute angle-closure glaucoma! Moreover, do not cover the eye, since darkness induces mydriasis and worsens the condition

Primary open-angle glaucoma (POAG)

Epidemiology:

- **The most common type of glaucoma.** present in 2% of people older than 40 years.
- Second leading cause of blindness following age-related macular degeneration (AMD).

Pathophysiology

- Secondary clogging of the trabecular meshwork or reduced drainage → gradual ↑ in IOP → vascular compression → ischemia to the optic nerve → progressive visual impairment.

Risk factors: age, family history, black patients, myopia, hypertension, diabetes mellitus

Features:

- **bilateral, progressive visual field loss (from peripheral to central) (Loss of nasal visual field) progressing to 'tunnel vision'**
- Fundoscopy: cupping and pallor of optic disc

Management:

- Eye drops to lower intra-ocular pressure (IOP)
- Laser trabeculoplasty
  - ⇨ An alternative first-line treatment
  - ⇨ refractory to pharmacotherapy

Medication	Mode of action	Notes
Prostaglandin analogues (e.g. Latanoprost)	Increases uveoscleral outflow	Once daily administration **Preferred first-line therapy.** **should be used first-line in patients with a history of asthma.** **Adverse effects**: ⇨ brown pigmentation of the iris, ⇨ growth of eyelashes ⇨ Epithelial keratopathy ⇨ Systemic: paresthesia, hypokalemia, renal stones, acidosis, and aplastic anemia.
Beta-blockers (e.g. Timolol)	Reduces aqueous production	**Should be avoided in asthmatics and patients with heart block**
Sympathomimetics (e.g. brimonidine, an alpha2-adrenoceptor agonist)	Reduces aqueous production and increases outflow	Avoid if taking MAOI or tricyclic antidepressants Adverse effects include hyperaemia
Carbonic anhydrase inhibitors (e.g. acetazolamide)	Reduces aqueous production	Systemic absorption may cause sulphonamide-like reactions
Miotics (e.g. pilocarpine, a muscarinic receptor agonist)	Increases uveoscleral outflow	Adverse effects included a constricted pupil, headache and blurred vision

Age related macular degeneration (AMD)

Epidemiology

- The most common cause of blindness

Pathophysiology

- progressive degenerative changes in the **central part** of the retina (macula) → visual impairment

Risk factors:

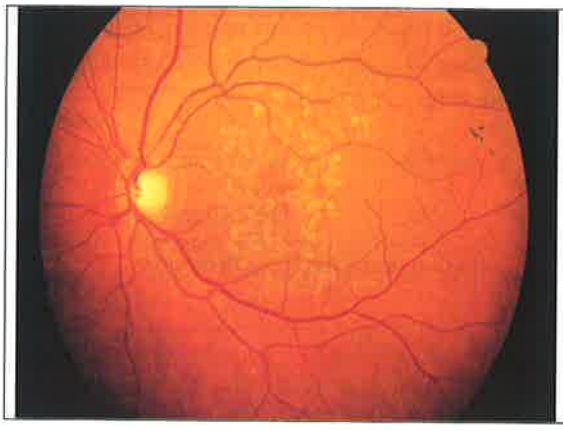
- Advanced age, smoking

Classification

	Dry AMD (nonexudative, atrophic)	Wet AMD (exudative , neovascular)
Prevalence	~ 90%	~ 10%
Pathophysiology	Deposition of drusen (yellow round spots) in the retinal pigment epithelium.	Choroidal neovascularization (between the retinal pigment epithelium and Bruch's membrane)
Onset	slow progressive visual impairment (**usually over decades**)	acute or insidious onset (over weeks to months)
Presentation	Bilateral	manifests in one eye
Fundoscopy	Drusen **(a small, yellowish, granular, subretinal deposits that are age related).**	Subretinal and intraretinal hemorrhage and/or exudate. **if neovascularisation is present fluorescein angiography is performed**
Treatment	Supportive: stop smoking Diet: high dose of beta-carotene, vitamins C and E, and zinc. **Supplements should be avoided in smokers due to an increased risk of lung cancer**	First-line: injection of VEGF inhibitors (ranibizumab, bevacizumab, pegaptanib) into the vitreous body.
Symptoms	Reduced visual acuity: 'blurred', 'distorted' vision, **central vision is affected first (central scotomas)**	

Differential diagnosis

Differential diagnosis of vision loss		
Condition	**Clinical features**	**Fundoscopy**
Age related macular degeneration	• May be insidious (dry AMD) or rapid (wet AMD) onset • Impairment of central vision only (vision loss is rare)	• Drusen • Macula depigmentation
Open-angle glaucoma	• Insidious onset • Peripheral vision loss (tunnel vision)	• Disc cupping with high intraocular pressure
Central Vessel occlusion (retinal artery)	• Acute or subacute onset • Complete vision loss	• Swollen disc • Retinal haemorrhages • Cotton wool spots
Retinal detachment	• Acute onset • Partial or complete vision loss (falling curtain)	• Detached or floating retina
Cataract	• Insidious onset • Blurred, dim vision, and a glare • Absent or opacified red-reflex	• Retina may not be visible (in advanced disease)



The fundus shows small pale dots over the macular area typical of drusen. This is macular degeneration and one of the commonest causes of blindness.

Top tips

Drusen = Dry macular degeneration

Macular degeneration - smoking is risk factor

Cataracts

Normal, clear lens

Lens clouded by cataract

A cataract is an opacity of the normally clear lens which may develop as a result of aging, metabolic disorders, trauma or heredity

Definition

- **opacification of the lens**

Causes

- **Majority**
  - ⇨ age related (Senile cataracts)
    - the most common cause
      - ❖ 17% of people older than 40 years
      - ❖ 50% of people older than 75 years
  - ⇨ UV light
- **Systemic**
  - ⇨ diabetes mellitus
  - ⇨ steroids
    - **Inhaled steroids can cause cataracts**
  - ⇨ infection (congenital rubella)
  - ⇨ metabolic:
    - diabetes
    - hypocalcaemia,
    - galactosaemia
      - ❖ but if the galactosaemia is treated, the cataract is reversible.
  - ⇨ myotonic dystrophy,
  - ⇨ Down's syndrome
- **Ocular**
  - ⇨ trauma
  - ⇨ uveitis
  - ⇨ high myopia
  - ⇨ topical steroids

Feature

- Symptoms
  - ⇨ painless, progressive, and slow vision loss
- Physical exam
  - ⇨ absent red reflex

Classification

- **Nuclear sclerosis**:
  - ⇨ **the most common type** of cataract,
  - ⇨ involves the central or 'nuclear' part of the lens.
  - ⇨ **common in old age**
  - ⇨ reduction of vision is the major symptom.

- ⇨ change lens refractive index,
  - often leads to an **increase in refractive power of the lens** causing **nearsightedness (problems with distance vision**).
- Polar: localized, commonly inherited, lie in the visual axis
- **Subcapsular:**
  - ⇨ glare is the major symptom
    - **Glare** is difficulty seeing in the presence of bright light such as direct or reflected sunlight or artificial light such as car headlamps at night.
  - ⇨ due to steroid use, just deep to the lens capsule, in the visual axis
  - ⇨ **Posterior subcapsular cataracts** are associated with:
    - **retinitis pigmentosa**
    - **chronic steroid** use.
  - ⇨ Anterior subcapsular cataracts are associated with:
    - idiopathic or
    - secondary to trauma and iotragenic causes.
- Dot opacities
  - ⇨ common in normal lenses,
  - ⇨ also seen in:
    - diabetes
    - myotonic dystrophy

Diabetic retinopathy See endocrinology

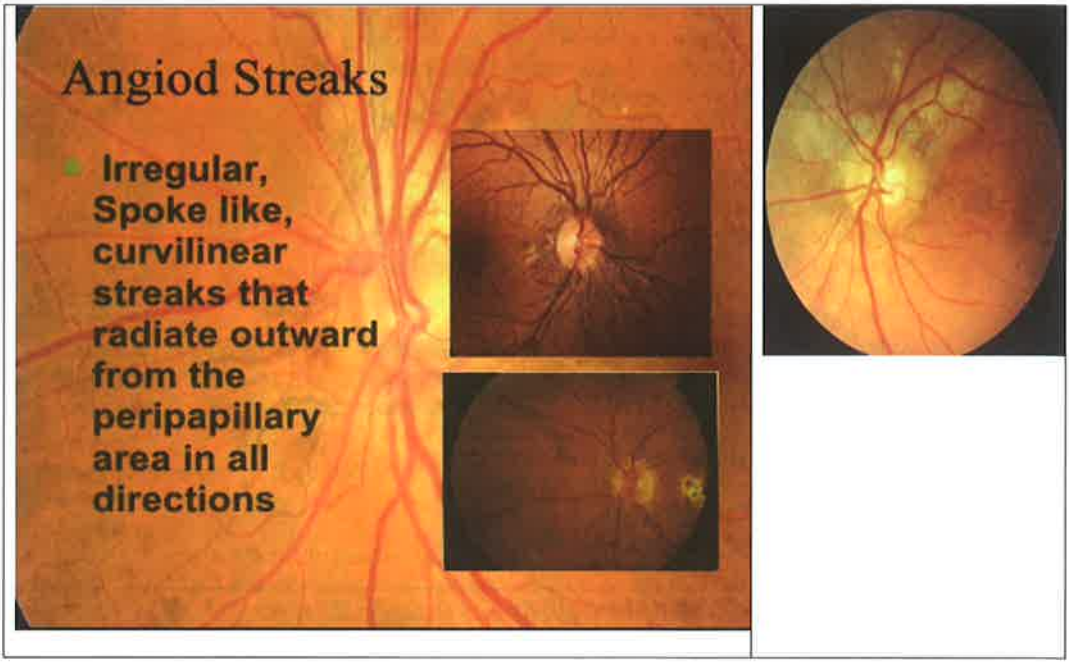
Angioid retinal streaks

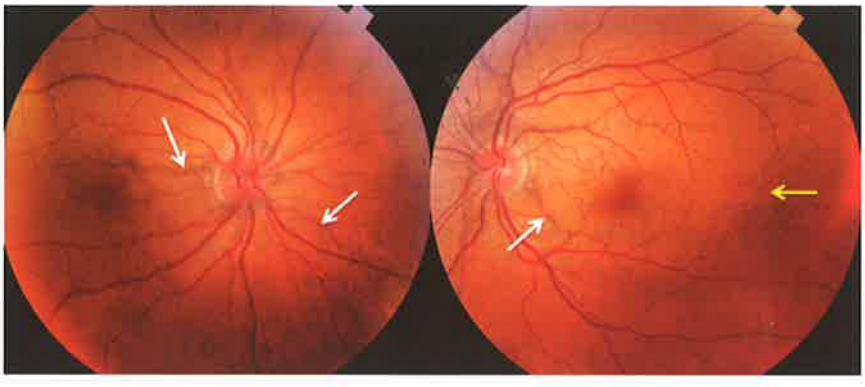
- Angioid retinal streaks are seen on fundoscopy as irregular dark red streaks radiating from the optic nerve head. They are caused by degeneration, calcification and breaks in Bruch's membrane .

Causes

A useful mnemonic for angioid retinal streak is **SLAPPERS**:

- S - **Sickle-cell anaemia**
- L - Lead poisoning
- A - Abetalipoproteinaemia/acromegaly
- P - **Paget's disease** /phacomatoses (tuberous, sclerosis, neurofibromatosis, Sturge-Weber)
- P - **Pseudoxanthoma elasticum**
- E - Ehlers-Danlos syndrome
- R - Raised calcium or phosphate
- S - Short people (dwarfism).





Mydriasis

Causes of mydriasis (large pupil)

- third nerve palsy
- Holmes-Adie pupil
- traumatic iridoplegia
- phaeochromocytoma
- congenital
- **Drug causes of mydriasis**
  - ⇨ topical mydriatics: tropicamide, atropine
  - ⇨ sympathomimetic drugs: amphetamines, pseudoephedrine, amphetamines and cocaine,
  - ⇨ anticholinergic drugs: eg antihistamines, atropine and tricyclic antidepressants
  - ⇨ Poisons (atropine, CO, ethylene glycol).

Miosis

Causes of small pupils include:

- Horner's syndrome
- Old age
- **Pontine haemorrhage**
- Argyll Robertson pupil
- Drugs, and
- Poisons (opiates, organophosphates).

Holmes-Adie pupil

Holmes ADIe = DIlated pupil, females, absent leg reflexes

Abnormally dilated pupil (mydriasis) which does not constrict in response to light, loss of deep tendon reflexes, and abnormalities of sweating.

Holmes-Adie pupil is a benign condition most commonly seen in women. It is one of the differentials of a dilated pupil.

Overview

- unilateral in 80% of cases
- dilated pupil (**tonically dilated pupil**)
- slowly reactive to accommodation but very poorly (if at all) to light
- once the pupil has constricted it remains small for an abnormally long time
- associated with absent ankle/knee reflexes and impaired sweating
  - The cause of the associated arreflexia is unknown.

Pathophysiology

- Viral or bacterial infection causes → damage to neurons in the **ciliary ganglion, located in the posterior orbit**, that provides parasympathetic control of eye constriction.
- damage to the **dorsal root ganglia of the spinal cord** → problems with autonomic control of the body.

Diagnosis

- **testing with low dose (1/8%) pilocarpine may constrict the tonic pupil due to cholinergic denervation super-sensitivity. A normal pupil will not constrict with the dilute dose of pilocarpine.**

Argyll-Robertson pupil

- the prostitute's pupil - accommodates but doesn't react.
- Another mnemonic used for the Argyll-Robertson Pupil (ARP) is Accommodation Reflex Present (ARP) but Pupillary Reflex Absent (PRA)

Features

- small, irregular pupils
- no response to light but there is a response to accommodate

Causes

- diabetes mellitus
- syphilis (neurosyphilis)

Anisocoria

- is a condition characterized by an unequal size of the eyes' pupils.
- Affecting 20% of the population,
- it can be an entirely harmless condition or a symptom of more serious medical problems
- **The history of anisocoria, with headaches and diplopia should ring alarm bells, in that a life-threatening posterior communicating artery aneurysm/berry aneurysm needs to be excluded urgently.**

Optic atrophy

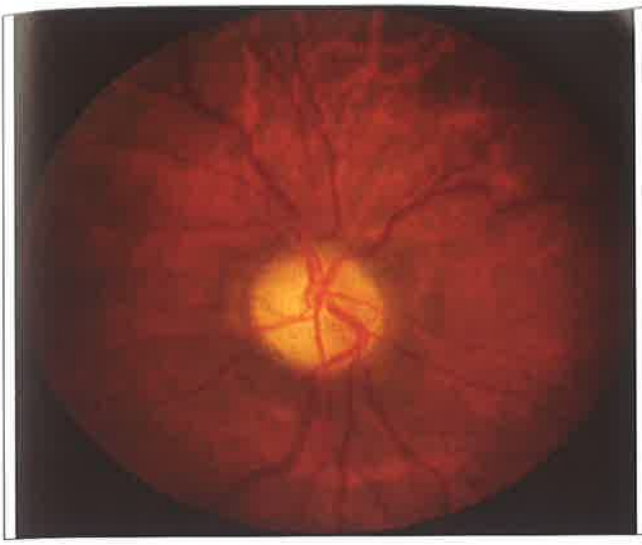
- Optic atrophy is a descriptive term, it is the optic neuropathy that results in visual loss
- Usually bilateral and causes a gradual loss of vision.
- On fundoscopy optic atrophy is seen as pale, well demarcated disc.
- Causes may be acquired or congenital

Acquired causes

- multiple sclerosis
- papilloedema (longstanding)
- raised intraocular pressure (e.g. glaucoma, tumour)
- retinal damage (e.g. choroiditis, retinitis pigmentosa)
- ischaemia
- toxins: tobacco amblyopia, quinine, methanol, arsenic, lead
- nutritional: vitamin B1, B2, B6 and B12 deficiency

Congenital causes

- Friedreich's ataxia
- **mitochondrial disorders e.g. Leber's optic atrophy**
  - ⇨ usually affects young men.
  - ⇨ It causes sequential optic neuropathies in days to weeks.
  - ⇨ It is typically painless and severe.
  - ⇨ Visual acuity fails to improve.
  - ⇨ Mutations in the MT-ND1, MT-ND4, MT-ND4L, and MT-ND6 genes
    - These genes are contained in mitochondrial DNA.
  - ⇨ Specifically, more than 50% of males with a mutation and more than 85% of females with a mutation never experience vision loss or related medical problems.
- DIDMOAD - the association of cranial Diabetes Insipidus, Diabetes Mellitus, Optic Atrophy and Deafness (also known as Wolfram's syndrome)



This patient has **optic atrophy** as revealed by a particularly **pale disc**. Causes include:

- **Glaucoma**
- External compression of the optic nerves, for example, pituitary tumour, and
- Multiple sclerosis.

Optic neuritis

The patient sees nothing and the doctor sees nothing

- Optic neuritis is a broad term which can be used to describe inflammation, degeneration or demyelination of the optic nerve.
- Optic neuritis is very rare in people over the age of 50.
- It encompasses a number of conditions, including:
  - ⇨ Papillitis (anterior optic neuritis) - the intraocular portion of the nerve is affected, and the **optic disc is swollen**
    - It is important to note that the disc changes in papilloedema may closely resemble those of papillitis but visual acuity is markedly reduced in papillitis and not papilloedema.
  - ⇨ Retrobular neuritis - the distal portion of the optic nerve is affected, and the disc is therefore not swollen
  - ⇨ Neuroretinitis - optic disc and adjacent temporal retina are affected.

Causes

- multiple sclerosis
- diabetes
- syphilis

Features

- unilateral decrease in visual acuity over hours or days
  - ⇨ Visual loss typically **occurs over days rather than hours.** Sudden visual loss due to optic neuritis is very unusual.
  - ⇨ **Optic neuritis presents with a particular type of central visual loss - a central scotoma.**
- poor discrimination of colours, 'red desaturation' - ie when red looks paler to one eye than the other -
- **The retrobulbar neuritis seen with ethambutol may be unilateral or bilateral; as such unilateral symptoms do not preclude the diagnosis.**

- pain worse on eye movement
- relative afferent pupillary defect during the 'swinging flashlight test'.
- central scotoma
- **Most cases of optic neuritis are retrobulbar and hence there are no abnormalities on fundoscopy.**
  - ⇨ **the most likely finding on fundoscopy → Normal optic disc**

Diagnosis

- **MRI with gadolinium of the brain** will likely show → **enhancement of the optic nerve**
- **Abnormal visual evoked potentials (VEP)**

Management

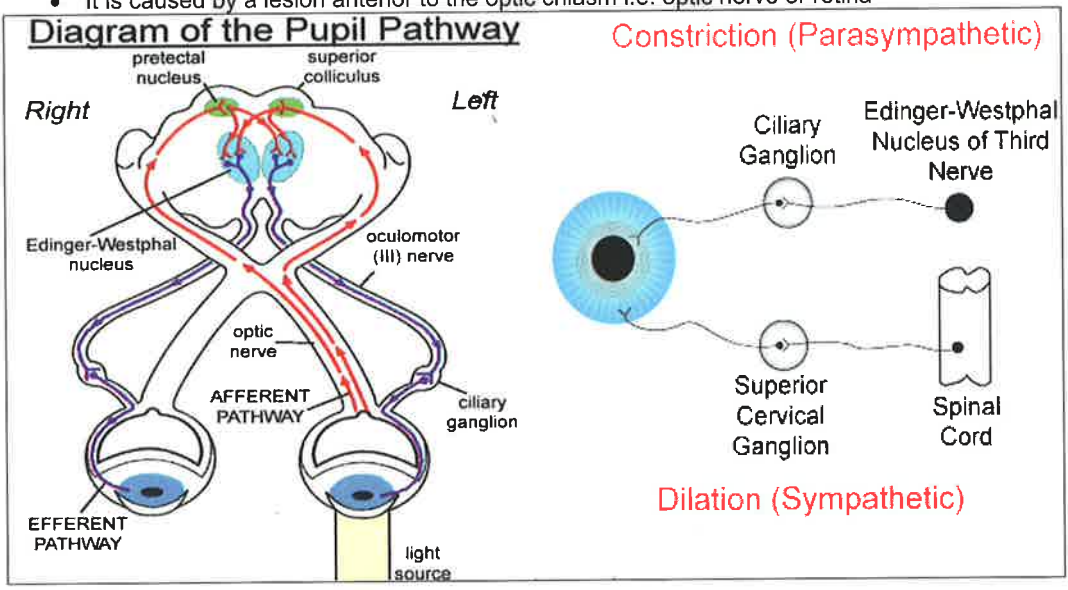
- high-dose steroids
  - ⇨ Methylprednisolone pulse therapy is the standard treatment
    - slightly shortens the time of recovery but does not prevent neurodegeneration and persistent visual impairment.
- recovery usually takes 4-6 weeks
- erythropoietin may have neuroprotective effects in autoimmune optic neuritis

Prognosis

- **MRI: if > 3 white-matter lesions, 5-year risk of developing multiple sclerosis is c. 50%**
- Retrobulbar neuritis has the same systemic implications as optic neuritis, in that an episode of optic or retrobulbar neuritis can contribute to a diagnosis of multiple sclerosis

Relative afferent pupillary defect

- Also known as the **Marcus-Gunn pupil**, a relative afferent pupillary defect is found by the 'swinging light test'.
- It is caused by a lesion anterior to the optic chiasm i.e. optic nerve or retina



Causes

- retina: detachment
- optic nerve: optic neuritis e.g. multiple sclerosis

Pathway of pupillary light reflex

- afferent: retina → optic nerve → lateral geniculate body → midbrain
- efferent: Edinger-Westphal nucleus (midbrain) → oculomotor nerve

Swinging flashlight test & Relative afferent pupillary defect RAPD (Marcus Gunn pupil)

- The Marcus Gunn pupil is a relative afferent pupillary defect indicating a decreased pupillary response to light in the affected eye
- In the swinging flashlight test, a light is alternately shone into the left and right eyes.
- A normal response would be equal constriction of both pupils, regardless of which eye the light is directed at. This indicates an intact direct and consensual pupillary light reflex.
- When the test is performed in an eye with an afferent pupillary defect, light directed in the affected eye will cause only mild constriction of both pupils (due to decreased response to light from the afferent defect), while light in the unaffected eye will cause a normal constriction of both pupils (due to an intact efferent path, and an intact consensual pupillary reflex). Thus, light shone in the affected eye will produce less pupillary constriction than light shone in the unaffected eye.
- A **positive** RAPD is due to retinal or optic nerve disease.

due to the consensual response of the pupillary light reflex, shining light in the unaffected eye will produce bilateral miosis.

- ⇨ shining light in the affected eye will not produce miosis because the afferent limb of the pupillary light reflex pathway is damaged (eg: optic neuritis)
- ⇨ However, **due to the bilateral projections of nerves from the Edinger-Westphal nucleus**, light shined in the **unaffected eye** will produce **bilateral miosis**. This phenomenon is called a **consensual response**.

Herpes simplex keratitis

- Herpes simplex keratitis most commonly presents with a dendritic corneal ulcer

Features

- red, painful eye
- photophobia
- epiphora
- visual acuity may be decreased
- fluorescein staining may show an epithelial ulcer (dendritic corneal ulcer)

Management

- immediate referral to an ophthalmologist
- topical aciclovir

Herpes zoster ophthalmicus

- Herpes zoster ophthalmicus (HZO) describes the reactivation of the varicella zoster virus in the area supplied by the ophthalmic division of the trigeminal nerve.
- It accounts for around 10% of case of shingles.

Features

- vesicular rash around the eye, which may or may not involve the actual eye itself
- Hutchinson's sign: rash on the tip or side of the nose. Indicates nasociliary involvement and is a strong risk factor for ocular involvement

Management

- **Oral antiviral** treatment for 7-10 days, ideally started within 72 hours. **Topical antiviral treatment is not given in HZO**
- oral corticosteroids may reduce the duration of pain but do not reduce the incidence of post-herpetic neuralgia
- ocular involvement requires urgent ophthalmology review

Complications

- ocular: conjunctivitis, keratitis, episcleritis, anterior uveitis
- ptosis
- post-herpetic neuralgia

Blepharitis

- Blepharitis is inflammation of the eyelid margins.
- It may due to either meibomian gland dysfunction (common, posterior blepharitis) or seborrhoeic dermatitis/staphylococcal infection (less common, anterior blepharitis).
- Blepharitis is also more common in patients with rosacea
- The meibomian glands secrete oil on to the eye surface to prevent rapid evaporation of the tear film. Any problem affecting the meibomian glands (as in blepharitis) can hence cause drying of the eyes which in turns leads to irritation

Features

- symptoms are usually bilateral
- grittiness and discomfort, particularly around the eyelid margins
- eyes may be sticky in the morning
- eyelid margins may be red. Swollen eyelids may be seen in staphylococcal blepharitis
- styes and chalazions are more common in patients with blepharitis
- secondary conjunctivitis may occur

Management

- softening of the lid margin using hot compresses twice a day
- mechanical removal of the debris from lid margins - cotton wool buds dipped in a mixture of cooled boiled water and baby shampoo is often used*
  - ⇨ *an alternative is sodium bicarbonate, a teaspoonful in a cup of cooled water that has recently been boiled
- artificial tears may be given for symptom relief in people with dry eyes or an abnormal tear film

Keratitis

Definition

- Keratitis refers to inflammation of one or more of the three corneal layers, the most common of which is epithelial keratitis. This is **characterised by dendritic ulcers**. Rarer forms involve the stroma or endothelium.

Causes

- *Pseudomonas aeruginosa* is commonly associated with contact lens related infections.
- The management must also include advising the patient to discontinue wearing contact lenses and referral to a specialist ophthalmic unit.
- Recurrence is common.

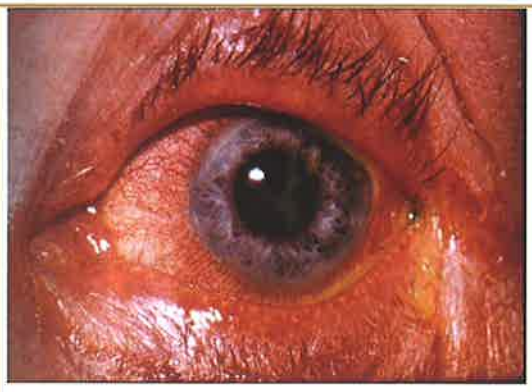
Keratitis overview		
	Characteristic features	**Therapy**
Bacterial keratitis (typically Staph. aureus Pseudomonas is seen in contact lens wearers.)	➲ **Most common form of keratitis** ➲ ↑ Risk with wearing contact lenses ➲ Purulent discharge and/or hypopyon ➲ Round corneal infiltrate or ulcer	Topical broad-spectrum antibiotics (e.g., ciprofloxacin)
Herpes zoster keratitis	➲ ↓ Corneal sensation ➲ **Punctate lesions** on the corneal surface (early disease) ➲ Vesicular eruption on forehead, bridge, and tip of the nose	➲ Oral acyclovir, valacyclovir, or famciclovir ➲ Topical steroids
Herpes simplex keratitis	Dendritic or geographic corneal ulcer	Topical trifluridine or ganciclovir
Acanthamoeba keratitis	➲ ↑ Risk with wearing **contact lenses** ➲ Corneal ring infiltrate	Topical antiseptic (e.g., chlorhexidine) with propamidine

Features

- Red eye: pain and erythema (**sharp ocular pain**)
- photophobia
- blurred vision (in many cases).
- Microbial keratitis, **causing a white corneal infiltrate**
- foreign body, gritty sensation
- hypopyon may be seen

Dendritic ulcers

- **caused by** herpes simplex virus.
- **Presentation** is usually with pain, photophobia, blurred vision, conjunctivitis and chemosis.
- Steroid eye drops are contraindicated as they may induce massive amoeboid ulceration and blindness.
- **treated with aciclovir eye drops,** which should be continued for three days after the ulcer has healed.



Red eye

Red eye - glaucoma or uveitis?
- glaucoma: severe pain, haloes, 'semi-dilated' pupil
- uveitis: small, fixed oval pupil, ciliary flush

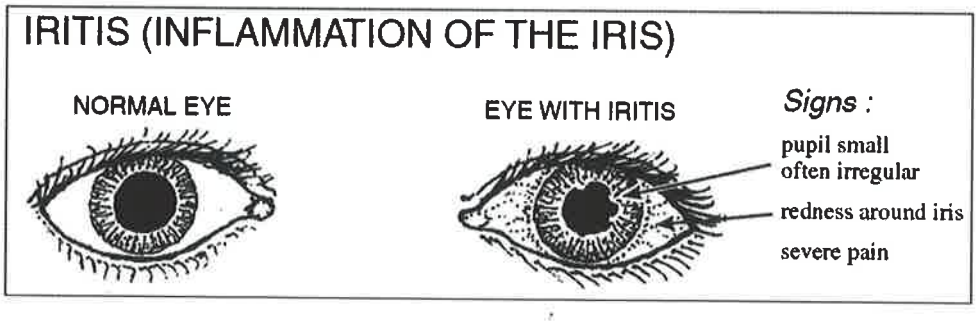
There are many possible causes of a red eye. It is important to be able to recognise the causes which require urgent referral to an ophthalmologist. Below is a brief summary of the key distinguishing **features**

Acute angle closure glaucoma

- severe pain (may be ocular or headache)
- decreased visual acuity, patient sees haloes
- semi-dilated pupil
- hazy cornea

Anterior uveitis

- **Features**
  - ⇨ acute onset
  - ⇨ pain
  - ⇨ blurred vision and photophobia
  - ⇨ small, fixed oval pupil, ciliary flush
  - ⇨ sign on ocular examination →Hypopyon
- **Iritis is associated with** conditions such as:
  - ⇨ Reiter's
  - ⇨ Behcet's
  - ⇨ **Psoriatic arthropathy (about 20%)**
  - ⇨ inflammatory bowel disease.
- **Signs of anterior uveitis**
  - ⇨ **Keratic precipitates**: (opaque aggregates of inflammatory cells deposited on the endothelium in anterior uveitis. They are typically located inferiorly.
  - ⇨ Cells +/- flare +/- fibrin in the anterior chamber
  - ⇨ Ciliary injection - localised conjunctival injection (redness) around the limbus
  - ⇨ Posterior synechiae - where part of the pupil margin becomes stuck to the lens
  - ⇨ Hypopyon (in severe anterior uveitis).



Scleritis

Definition

- inflammation that occurs throughout the entire thickness of the sclera,

Aetiology

- may be underlying autoimmune disease e.g. rheumatoid arthritis
  - ⇨ Around 50% of patients with scleritis have an underlying disease, of which the majority are connective tissue disorders.
  - ⇨ **Rheumatoid arthritis is the most common.**

Features

- **severe pain** (may be worse on movement) and tenderness
  - ⇨ **pain** in scleritis is more evident and severe than episcleritis.
  - ⇨ **Tenderness** to palpation of the globe can differentiate it from episcleritis. After asking the patient to look down with eyelids closed, the physician gently presses the globe. Patients with scleritis have tenderness on palpation, while those with episcleritis do not.
  - ⇨ Unlike scleritis, patients with episcleritis do not complain of **blurred vision or photophobia.**
  - ⇨ Studies have shown that patients with RA-associated scleritis have **more widespread systemic disease** and a higher mortality rate than those episcleritis.
- 50% of cases are bilateral.
- Pain often radiates to the forehead, brow and jaw. This pain worsens with movement of the eye, and is classically worse at night.
- There is associated watering, photophobia and a gradual decrease in vision (sometimes with diplopia).
- Systemic symptoms such as fever, headache and vomiting can occur.
- On examination the globe is tender, and the sclera can have a bluish tinge.
- visual acuity is normal
- there is marked dilatation of the **deep and superficial** scleral vessels.
- Scleritis may cause thinning of the sclera (scleromalacia) and subsequent perforation.

Treatment

- Management ultimately depends on the underlying cause, but includes NSAIDs and prednisolone.
- **The patient should be referred urgently to the ophthalmology clinic**
- **Application of topical phenylephrine 2.5% leads to blanching of episcleral vessels in episcleritis but not in scleritis.**

Episcleritis

Scleritis is painful, episcleritis is not painful

- Results in ocular irritation with nodules.
- acute in onset, with **mild pain** or discomfort / grittiness.
- can be unilateral or bilateral, with localised or diffuse red eye.
- There may be **mild** photophobia and watering. **The lack of photophobia and discharge, and normal vision, makes episcleritis the most likely option**

Ocular manifestation of rheumatoid arthritis (see rheumatology)

Conjunctivitis

- Purulent discharge if bacterial, clear discharge if viral
- **Viral conjunctivitis**
  - ⇨ causes redness, soreness and watering.
  - ⇨ In severe cases it can cause a keratitis which may affect vision.
  - ⇨ It is highly contagious so patients should be advised to practise strict hand hygiene, to avoid sharing towels and to take time off work.
  - ⇨ It is a self-limiting disease which may take several weeks to resolve.
  - ⇨ Patients are treated with topical lubricants and some ophthalmologists give topical chloramphenicol to protect against secondary bacterial infections.

Subconjunctival haemorrhage

- history of trauma or coughing bouts
- **adverse effect of aspirin** therapy (and other antiplatelets).
- It usually resolves over 10-14 days.
- If the haematoma is large it may be worth considering prophylactic antibiotic eyedrops.

Posterior uveitis

- Posterior uveitis describes inflammation of the choroid, which can involve the retinal vessels.
- presents with gradual visual loss and floaters, which is often bilateral.
- Discomfort and erythema are rare.
- Slit light examination can demonstrate inflammatory lesions on the retina or choroid, with inflammation of the retinal vessels and oedema of the optic nerve.

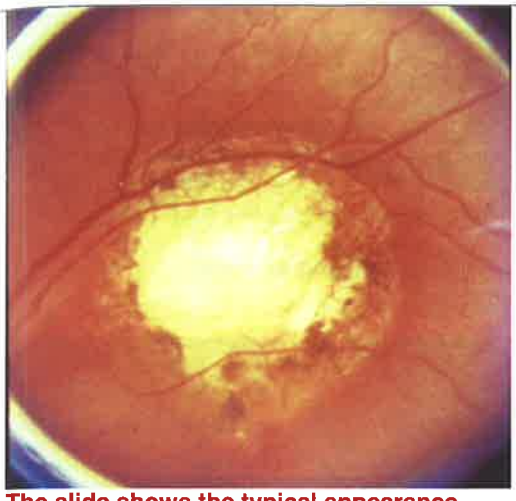
Retinitis

CMV Retinitis: causes hemorrhage at the edge of the area of retinal necrosis

- **Retinitis** is inflammation of the retina in the eye, which may lead to blindness.
- may be caused by several infectious agents, toxoplasmosis, cytomegalovirus and candida.
- Cytomegalovirus retinitis is the most common cause of vision loss in AIDS patients.

Toxocara retinitis

- **In retinitis due to *Toxocara canis*, there is usually only a single, well demarcated lesion.**



The slide shows the typical appearance of *Toxocara* retinitis with a lesion at the macula.

Retinitis pigmentosa

Retinitis pigmentosa - night blindness + funnel vision

Definition

- Retinitis pigmentosa is a **degenerative** disease involving retinal receptors and pigment cells.

Pathophysiology

- **degeneration of rod photoreceptor cells** in the retina → night blindness and low peripheral vision
  - ⇨ There are two types of photoreceptors, called rods and cones.
    - **Rods** are in the outer regions of the retina, and allow us to see in dim and dark light.
      - ❖ Died early → night blindness
    - **Cones** reside mostly in the central portion of the retina, and allow us to perceive fine visual detail and color.
      - ❖ Died in the late stages

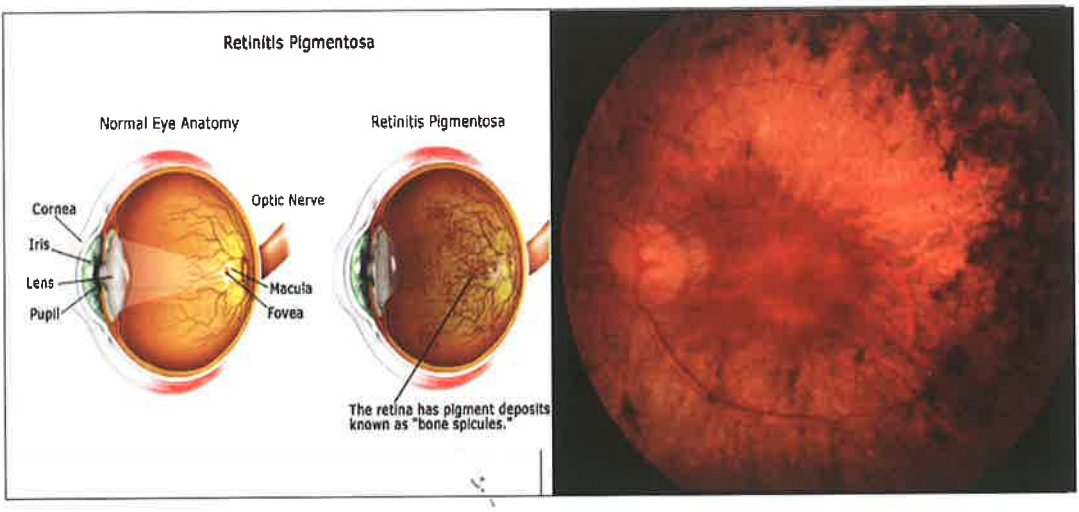
Features

- **night blindness is often the initial sign**
- funnel vision (the preferred term for tunnel vision)
- fundoscopy:
  - ⇨ black bone spicule-shaped pigmentation in the peripheral retina,
  - ⇨ mottling of the retinal pigment epithelium

Associated diseases

- Refsum disease:
  - ⇨ cerebellar ataxia, peripheral neuropathy, deafness, ichthyosis
- Usher syndrome
- abetalipoproteinemia
- Lawrence-Moon-Biedl syndrome
- Kearns-Sayre syndrome

- Alport's syndrome
- mitochondrial myopathy
- drug-induced
  - ⇨ Thioridazine
    - (typical antipsychotic drug belonging to the phenothiazine group and was previously widely used in the treatment of schizophrenia and psychosis; withdrawn worldwide in 2005 because it caused severe cardiac arrhythmias,)
    - It is important to differentiate this from corneal deposits that may develop with the use of chlorpromazine.
      - ❖ Thioridazine → **retinal** deposits (**retinitis pigmentosa**).
      - ❖ chlorpromazine → **corneal** deposits



Fundus showing changes secondary to retinitis pigmentosa

Sudden painless loss of vision

Causes	Notes
Central retinal vein occlusion	• Incidence increases with age • More common than arterial occlusion • Causes : glaucoma, polycythaemia, hypertension, DM • Features: ⇨ afferent pupillary defect ⇨ On fundoscopy: ▪ widespread dot-and-blot and/or flame-shaped hemorrhages in all four retinal quadrants ▪ Cotton wool spots characterized by yellow-white deposits on the retina caused by swelling of retinal nerve fibers due to ischemia ▪ Severe macular edema and papilledema • Fluorescein angiography: in order to differentiate ischemic from non-ischemic forms of retinal vein occlusion

Causes	Notes
Branch retinal vein occlusion	• Features: ⇨ Usually asymptomatic ⇨ No afferent pupillary defect ⇨ the hemorrhages are found in a single zone.
Central Retinal artery occlusion	• Causes: thromboembolism (from atherosclerosis) or arteritis (e.g. temporal arteritis) • Features: ⇨ Sudden, painless loss of vision in one eye (often described as a "descending curtain") ⇨ afferent pupillary defect, ⇨ history of amaurosis fugax, often describes as a 'black curtain' descending over the vision. ⇨ On fundoscopy: 'cherry red' spot on a pale retina, ▪ Grayish-white (cloudy) discoloration of the entire retina ▪ Cherry-red spot at the fovea centralis
Branch retinal artery occlusion	• Features: ⇨ Sudden onset of visual field defects (scotomas) in the affected eye ⇨ No afferent pupillary defect, ⇨ On fundoscopy: Grayish-white discoloration of the retinal quadrant supplied by the affected vessel
Retinal detachment	• Risk factors: Previous intraocular surgery (e.g., cataract surgery), posterior vitreous detachment • Most commonly due to retinal tears → retinal fluid, which is formed by vitreous degeneration, seeps into the subretinal space → retinal detachment • Features: ⇨ Prodromal symptoms: result from posterior vitreous detachment (floaters, flashes of light (photopsia) ⇨ Localized retinal detachment: scotoma (visual field defect): Dense shadow that starts peripherally progresses towards the central vision ⇨ Straight lines appear curved ⇨ Extensive retinal detachment and/or macular involvement: Central visual loss (often described by patients as a curtain descending over their field of vision) ⇨ Fundoscopy: A freshly detached retina has a grey color instead of the normal pink color and may appear crinkled. A retinal tear may be visible
Vitreous haemorrhage	• Causes : bleeding disorders, DM → Proliferative retinopathy → rupture fragile neovascular vessels (most common cause) • Features: ⇨ Large bleeds cause sudden visual loss ⇨ Moderate bleeds may be described as numerous dark spots ⇨ Small bleeds may cause floaters ⇨ Fundoscopy: inability to visualise the retina

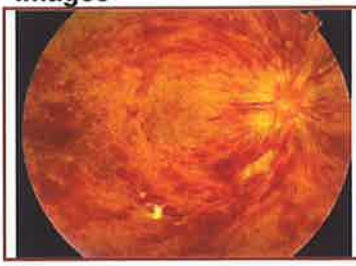
Amaurosis fugax

- Definition: sudden, painless loss of vision that lasts for seconds to minutes and is followed by spontaneous recovery (mostly unilateral)
- Cause: retinal ischemia following transient occlusion of the central retinal artery by microemboli
- Complications: Transient ischemic attacks (TIA)

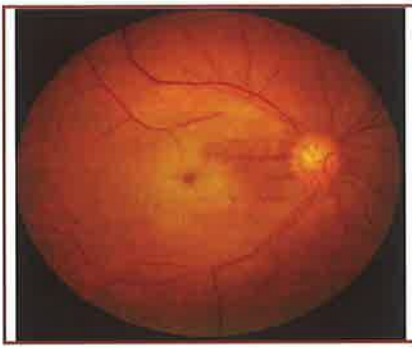
Posterior vitreous detachment

- Occur in up to 50-75% of the population over 65 years
- Features:
  - ⇨ Flashes of light (photopsia) - in the peripheral field of vision
  - ⇨ Floaters, often on the temporal side of the central vision
- Complications: Retinal tears/holes, retinal detachment, Vitreous hemorrhage

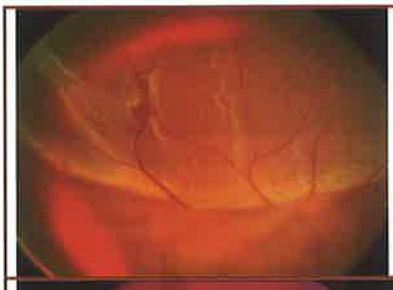
Images



Central vein occlusion:
Flame-shaped hemorrhage is visible in all four retinal quadrants.



Central retinal artery occlusion:
Narrow retinal arteries and a pale retina with early signs of nerve fiber layer edema are visible.
The fovea centralis appears red (cherry-red spot; due to the transparency of the well-vascularized choroid, as no nerve fibers are present in the fovea avascular zone. Therefore, there is no edema.



Retinal detachment:
The retina is visible as a yellow-grey, bullous elevation in the upper part of the image.
- Green overlay: detached retina
- Red overlay: tear

Fundus Photograph of Vitreous Haemorrhage

Central retinal vein occlusion - sudden painless loss of vision, severe retinal haemorrhages on fundoscopy

Flashes and floaters - vitreous/retinal detachment

An elderly patient with acute visual loss has giant cell arteritis until proved otherwise

The history of diabetes, complete loss of vision in the affected eye and inability to visualise the retina point towards a diagnosis of vitreous haemorrhage.

Nasal branch retinal vein occlusion → sudden blurring (not total visual loss) of the temporal field in the affected eye.

Of all types of retinal vessel occlusion, ischemic Central Retinal Vein Occlusion is most commonly associated with neovascularization.

Tunnel vision

Tunnel vision (also known as **Kalnienk vision**) is the loss of peripheral vision with retention of central vision, resulting in a constricted circular tunnel-like field of vision.

Causes

- papilloedema
- glaucoma
- retinitis pigmentosa
- choroidoretinitis
- optic atrophy secondary to tabes dorsalis
- hysteria

Ectopia lentis

Ectopia lentis/subluxation of the lens is associated with:

- Ehlers-Danlos syndrome
- Marfan's syndrome
- Weill-Marchesani syndrome (short stature, skeletal abnormalities and ectopia lentis), and
- Refsum's disease.

Fundoscopic features in eye infections

- ***Cytomegalovirus* (CMV) retinitis**
  - ⇨ secondary to human immunodeficiency virus (HIV)
  - ⇨ Fundoscopy of the left eye revealed an extensive **'brushfire-like' lesion** in the major superior temporal arcade with a large patch of white fluffy lesion mixed with extensive **retinal haemorrhages.**
- **Ocular histoplasmosis and syphilitic choroiditis** would give a fundus picture of **multiple whitish lesions**.
- **Syphilitic neuroretinitis** would normally give a picture of a **macular star exudation**.
- **Tuberculous periphlebitis** gives a picture of **perivenous sheathing and minimal retinal haemorrhages**.

Eye signs in Systemic diseases

- **Lisch nodules of the iris are golden nodules occurring bilaterally in the teenage years onwards in Neurofibromatosis type 1 (NF-1). Axillary freckles** appear at 10 years of age, while cafe au lait spots increase in size and number throughout childhood.
- **Brushfield spots** of the iris are found in people with Down syndrome.
- **Kayser-Fleischer rings** are due to copper deposition in Descemet's membrane of the cornea.
- **Band keratopathy is caused by calcium deposition in Bowman's layer of the cornea. Patients who present with band keratopathy should have a serum calcium and phosphate level**
- **Ectopia lentis with aortic regurgitation → Marfan syndrome (Lens dislocation (classically upwards)).**
  Inferior dislocated lens → consistent with a diagnosis of homocytinstinuria.
- **Roth's spots haemorrhages in the retina → associated with subacute bacterial endocarditis.** also, seen in leukaemia.
- **'black sunburst' - a chorioretinal scar, which is one of the commoner retinal manifestations of Sickle cell disease (SCD) and pathognomonic.**

Hyphaema

Overview

- Occurs when bleeding from iris vessels fills the anterior chamber with blood and if there is enough blood
- the main risk in the acute stage is of raised intraocular pressure (IOP).
- It is usually caused by trauma - often small objects (champagne corks, squash balls) hitting the eye.

Treatment

- Strict rest is vital if a hyphaema is present, as there is an increased risk of a second bleed in the initial period.
- **Intravenous carbonic anhydrase inhibitors is the most appropriate treatment**
- Aspiration may be required to prevent loss of vision.
- avoid drops that dilate the pupil (such as anticholinergics) the iris remains stable and a second bleed is therefore less likely.

The slide shows hyphaema: blood in the anterior chamber.