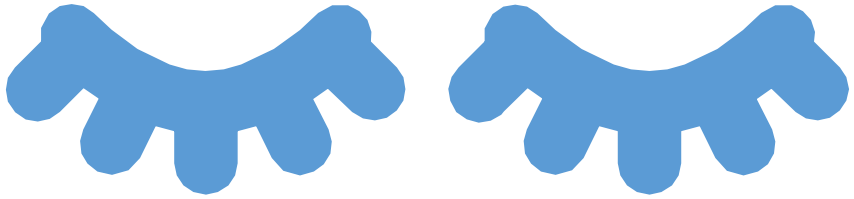


The outer layer of the eye
(cornea et sclera)

Sclera

- *It is a strong, opaque, white fibrous layer*

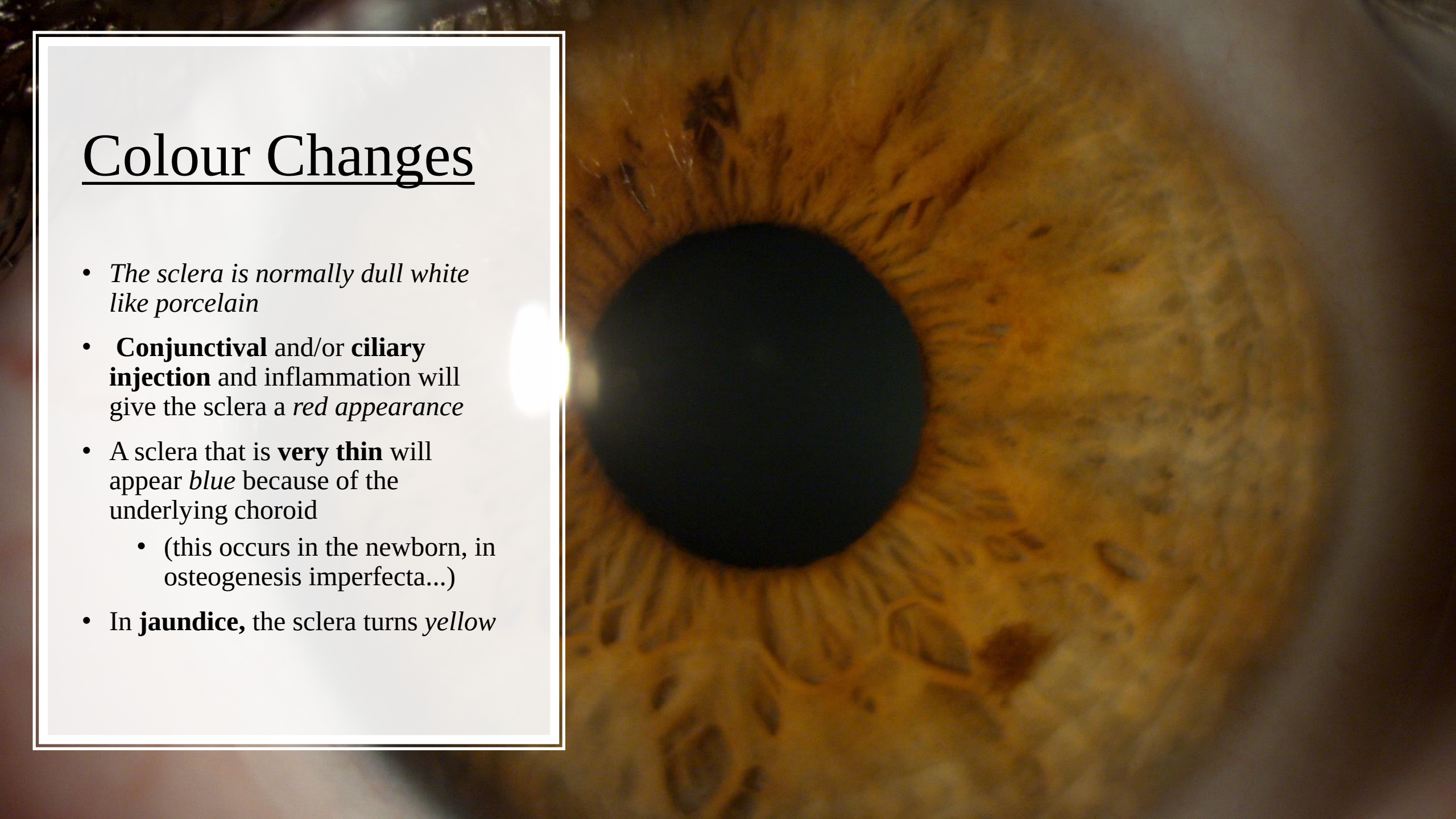
- **thickest (1mm)** anteriorly and at its posterior pole
- **thinnest (0.3mm)** at the equator and beneath the insertions of the rectus muscles
- The site where the fibers of the optic nerve enter the sclera is known as the *lamina cribrosa*
- In the angle of the anterior chamber, the sclera forms the *trabecular meshwork* and the *canal of Schlemm*
- *Layers:* episclera, stroma, lamina fusca

Examination Methods

- Inspection - slit lamp
- Ultrasound

Colour Changes

- *The sclera is normally dull white like porcelain*
- **Conjunctival** and/or **ciliary injection** and inflammation will give the sclera a *red appearance*
- A sclera that is **very thin** will appear *blue* because of the underlying choroid
    - (this occurs in the newborn, in osteogenesis imperfecta...)
- In **jaundice,** the sclera turns *yellow*



- ***Staphyloma***
- bulging of the sclera in which the underlying uveal tissue in the bulge is also thinned or degenerated

- ***Sclerectasia***
- thinning and bulging of the sclera without uveal involvement

Inflammation

Episcleritis

- *inflammation of the episclera*
- lymphocytic infiltration of subconjunctival and episcleral tissue
- common, usually idiopathic and benign, recurrent and frequently bilateral condition
- Females may be affected more commonly than males

- **Associated disease**
- ocular (e.g. dry eye, rosacea, contact lens wear)
- systemic (e.g. collagen vascular disorders such as rheumatoid arthritis, herpes zoster ophthalmicus)
- Infectious (very rare)

Episcleritis

- **Types**
  1. diffuse
  2. Sectoral
  3. nodular

- **Symptoms**
  - Discomfort
  - mild moderate pain
  - Epifora (watery eye)
  - Redness
  - Grittiness
  - photophobia

Episcleritis

- **Signs**
  - Localized / difuse redness
  - interpalpebral triangular configuration with the base at the limbus
  - nodule

- **Treatment :**
- usually resolves spontaneously within one to two weeks
- the nodular form can persist for extended periods of time
- cool compresses, artificial tears – mild cases
- A weak topical steroid - severe cases
- A topical / oral non-steroidal anti-inflammatory (NSAID) – severe cases

Scleritis

- *Deep, diffuse or localized inflammation of the sclera*
- anterior (95%), posterior (5%) scleritis

- **Etiology**
- In 50% cases is associated with connective tissue diseases such as :
  - *Rheumatoid arthritis*
  - *Polyarteritis nodosa*
  - *Systemic lupus erythematosus*
  - *Non-specific arteritis*
  - *Wagener's granulomatosis*
  - *Dermatomyositis*
  - *Polychondritis*
- It may be associated with prior episodes of herpes zoster

Types

Nodular

- The nodules consist of edematous swollen sclera and are *not mobile* (in contrast to episcleritis)

Diffuse

- The inflammation include the entire anterior sclera

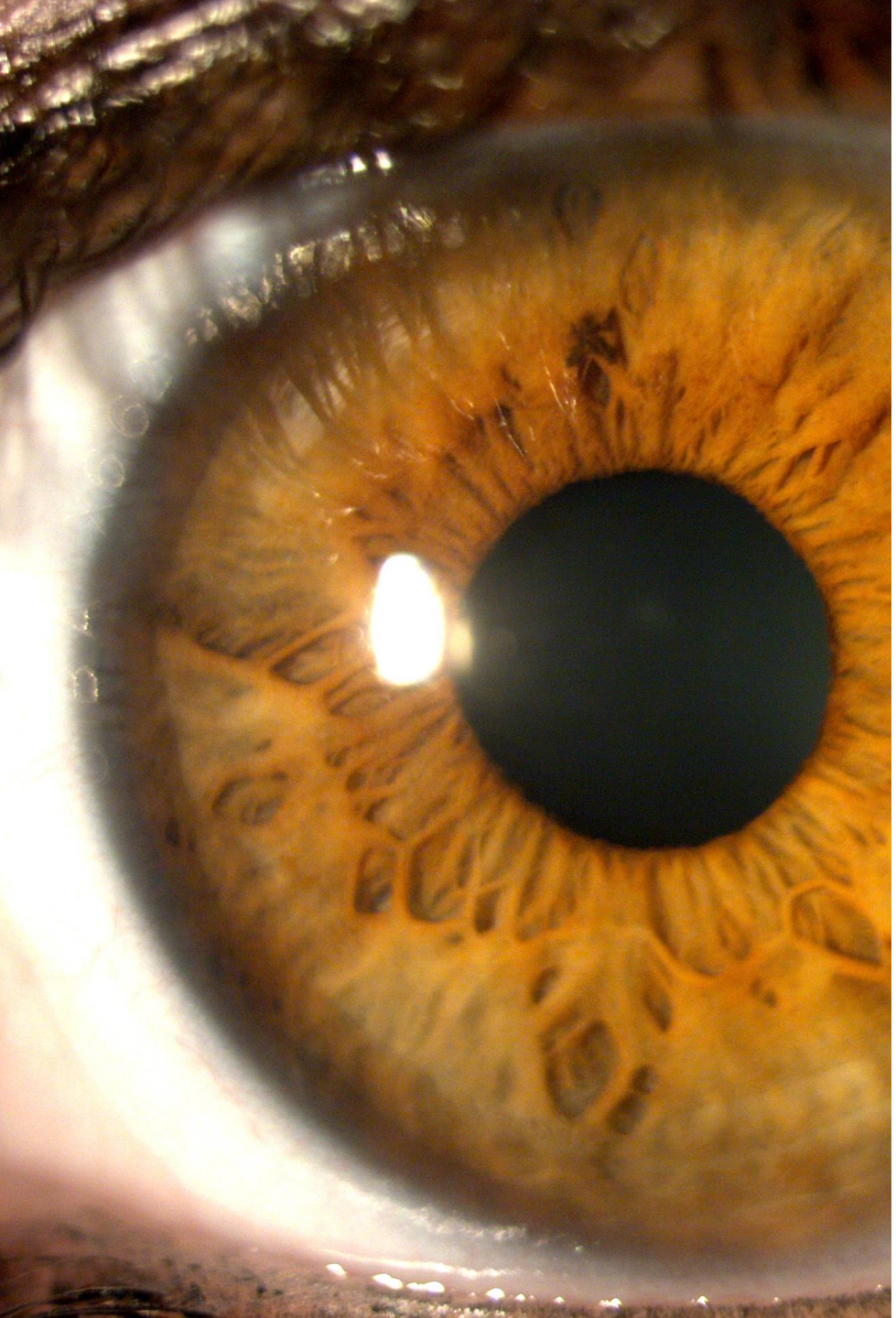
Necrotizing

- There are large areas of avascular sclera leading to necrosis
- There is exposure of the uveal pigment through a markedly thin sclera

Treatment

- **I. Medical therapy**
  - It is the first line of defence
  - *Local*: Corticosteroids
  - *General:* Systemic corticosteroids, Analgesics, anti-inflammatory drugs, Cytotoxic immunosuppressive drugs may be useful, e.g. cyclophosphamide

- **II. Surgical treatment**
- Surgical repair of scleral perforation (e.g. patch grafting)



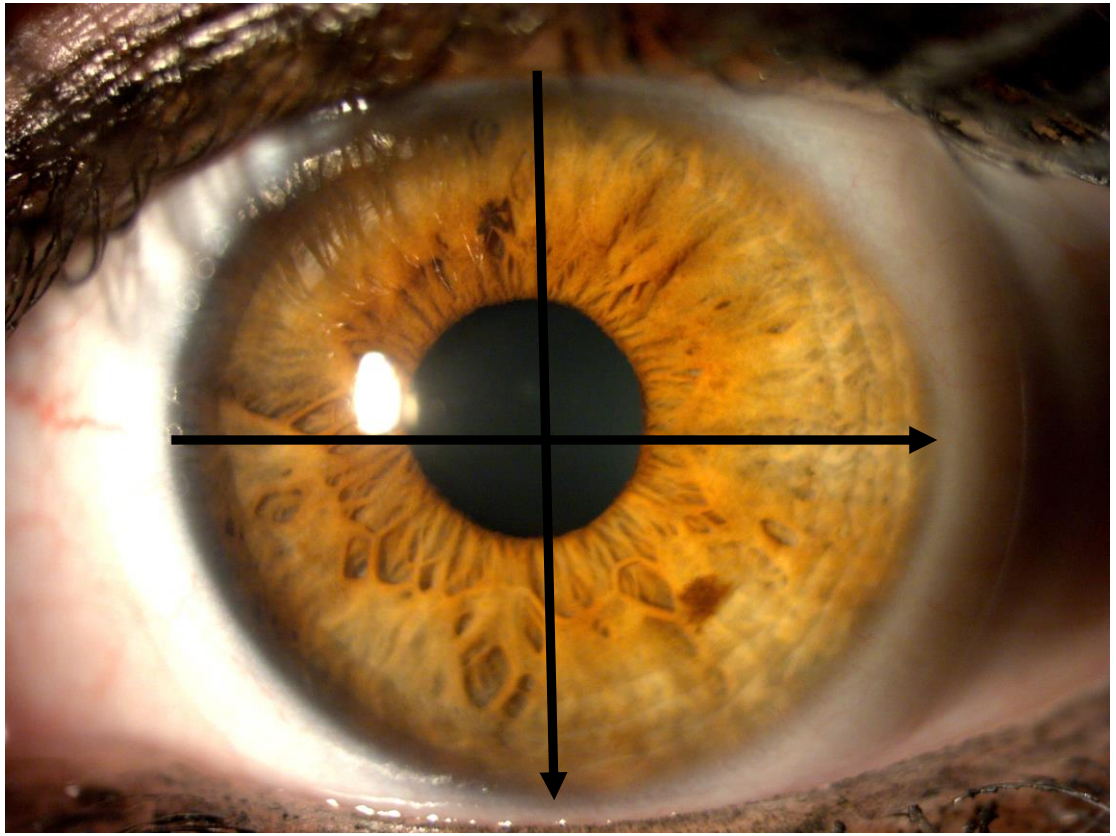
CORNEA

- *Avascular, transparent, smooth, shining structure*

- Responsible for +43D of the optical power of the eye
- Nutrients are supplied and metabolic products removed via *perilimbal blood vessels*, the *aqueous humour* posteriorly and the *tears* anteriorly
- The most densely innervated tissue in the body
- Innervation:
  - *1st division of the trigeminal nerve*

Anatomy

- The average corneal diameter is:
  - Vertically 11 mm
  - Horizontally 12 mm
- Average central thicknes: 0,55 mm
- Thicker towards the periphery



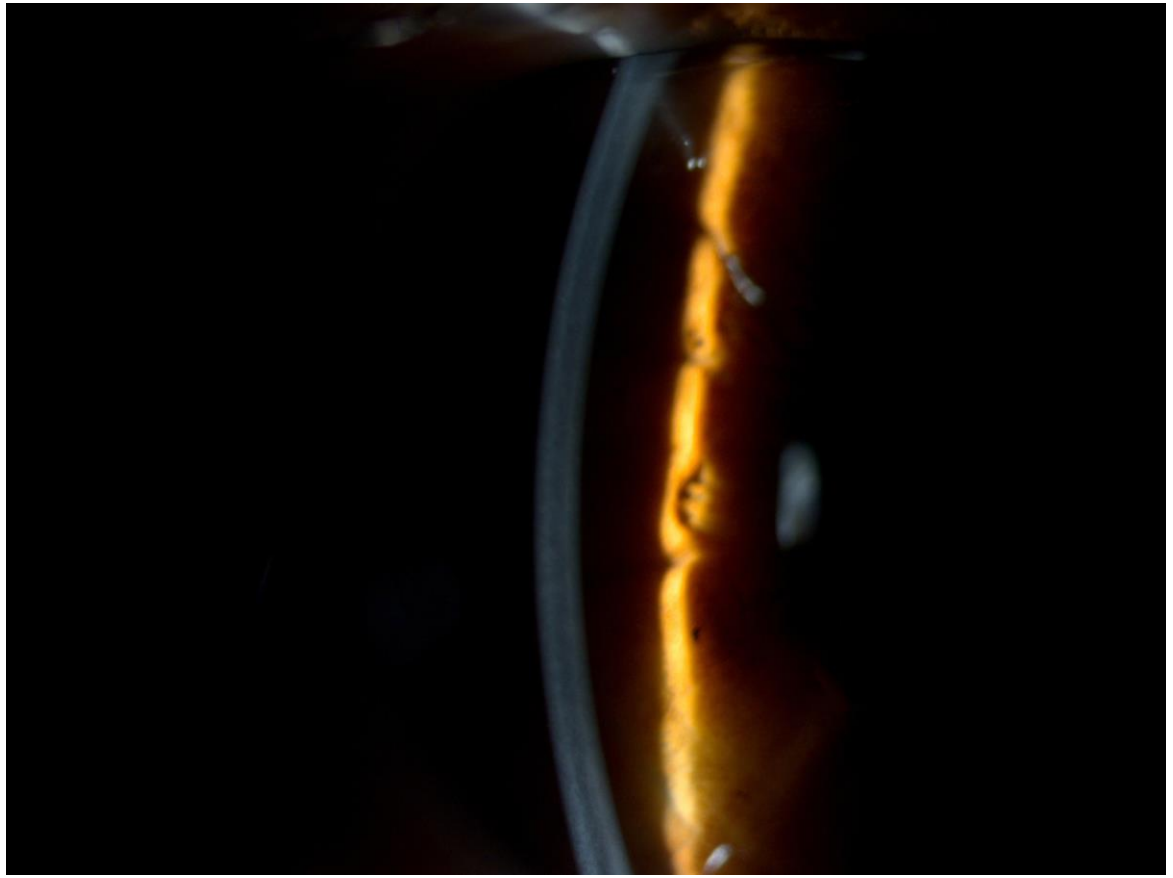
Histology

- **<u>Cornea consists of 6 layers:</u>**
  - Epithelium
  - Bowman layer
  - Stroma
  - Dua´s layer
  - Descement membrane
  - Endothelium

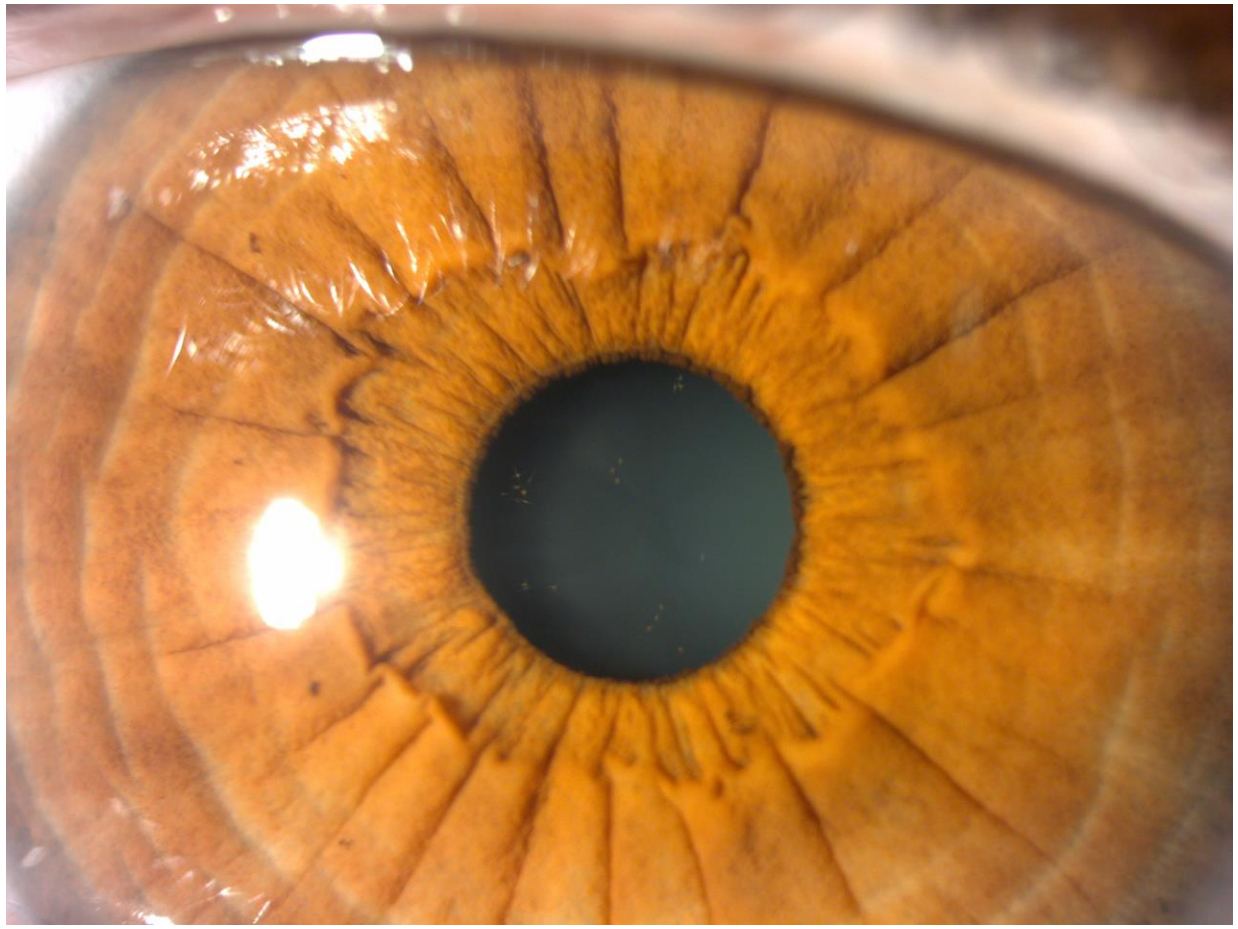
Examination methods

1. Slit lamp

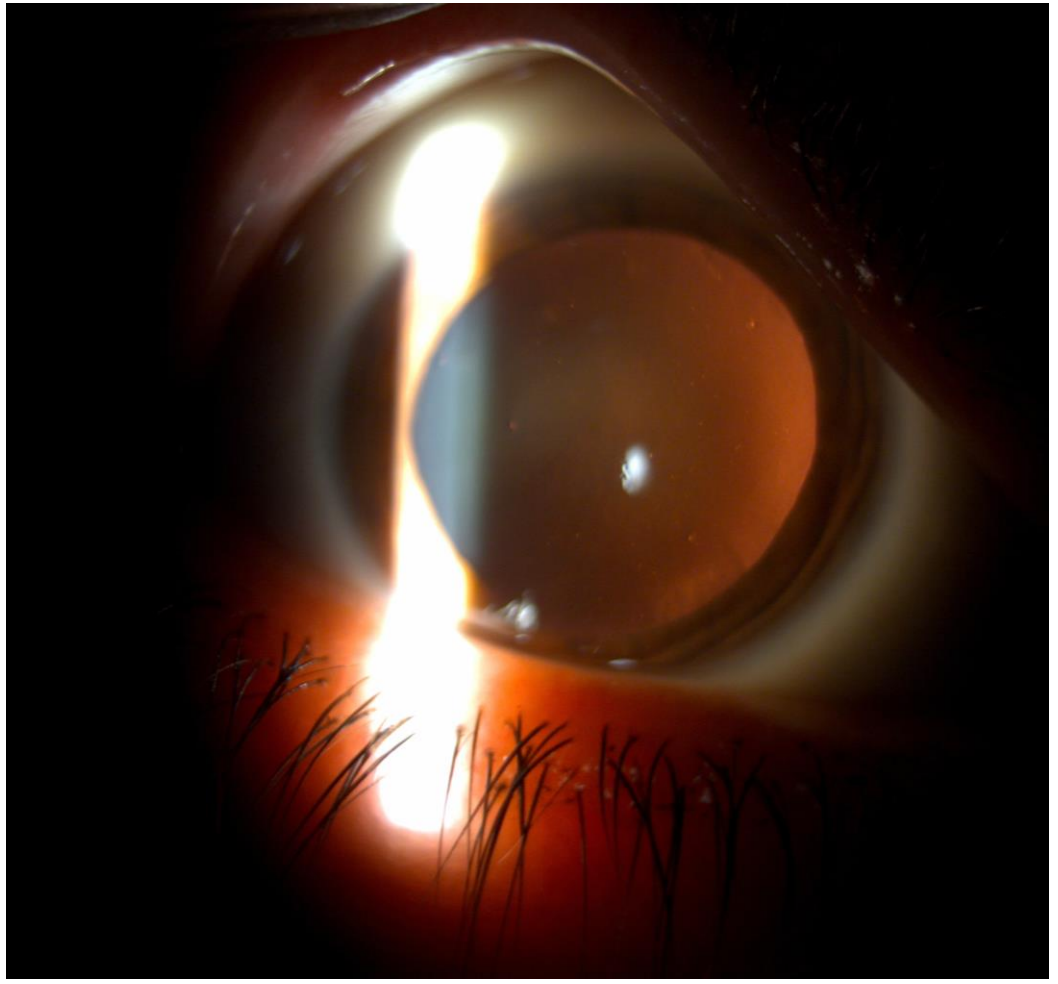
- position, depth and size of the abnormalities
- ***Direct illumination***
  - Diffuse or narrow slit-beam
  - Cobalt blue filter- conjunctiva or cornea stained with fluorescein, bengal solution
- ***Retroiluumination***
  - Uses reflected light from iris or fundus after pupil dillatation to illuminate the cornea



narrow slit beam



diffuse slit-beam



retroillumination

Symptoms of corneal disease

- The 4 main symptoms of corneal disease are:

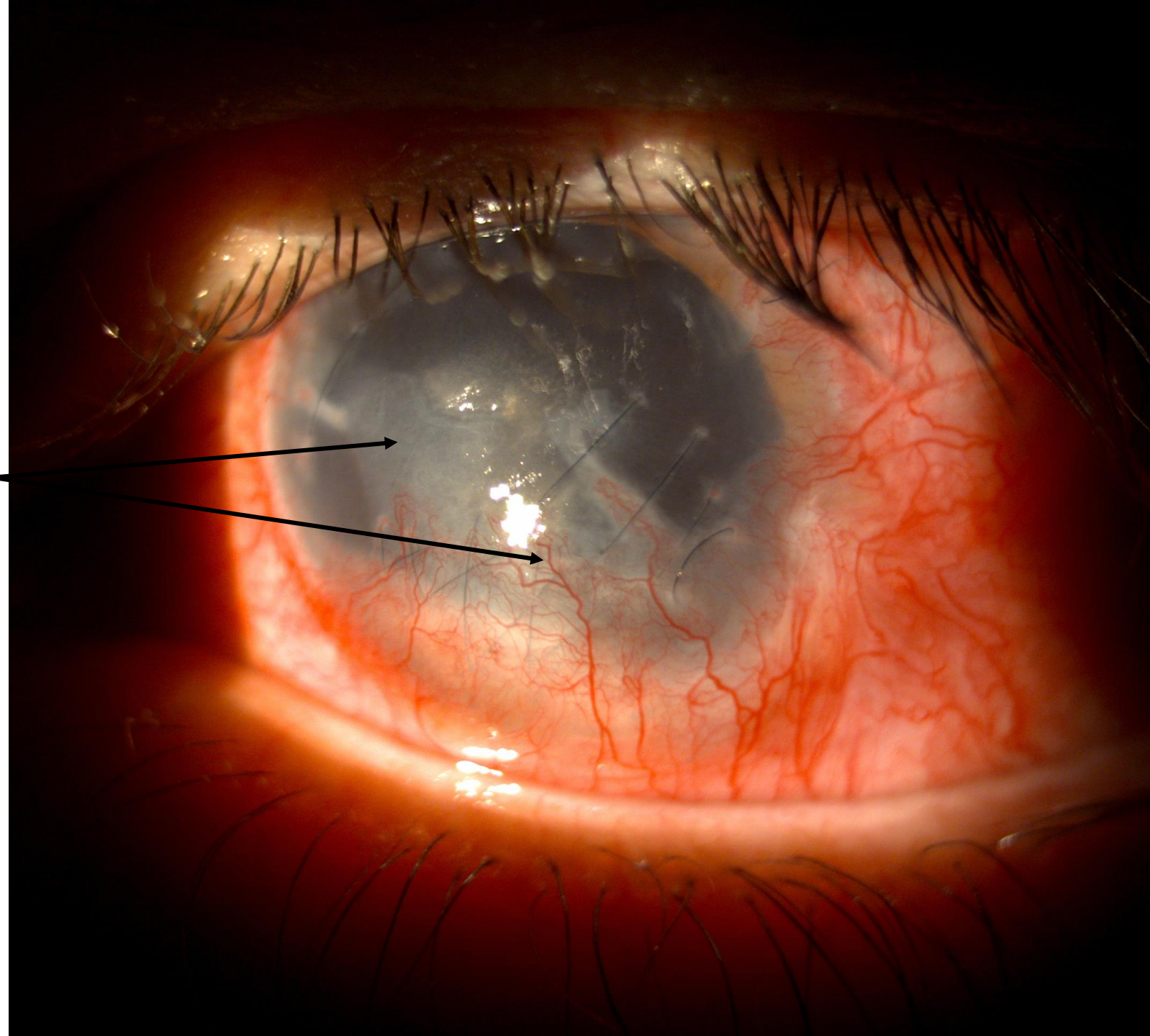
1. Reduced vision
2. Pain
3. Iridescent vision
4. Lacrimation / tearing

Clinical signs of corneal disease

- **Corneal opacities**
  - ***Nebula*** is a mild loss of corneal transparency
  - ***Macula*** is an intermediate opacity
  - ***Leucoma*** is a totally opaque portion of the cornea

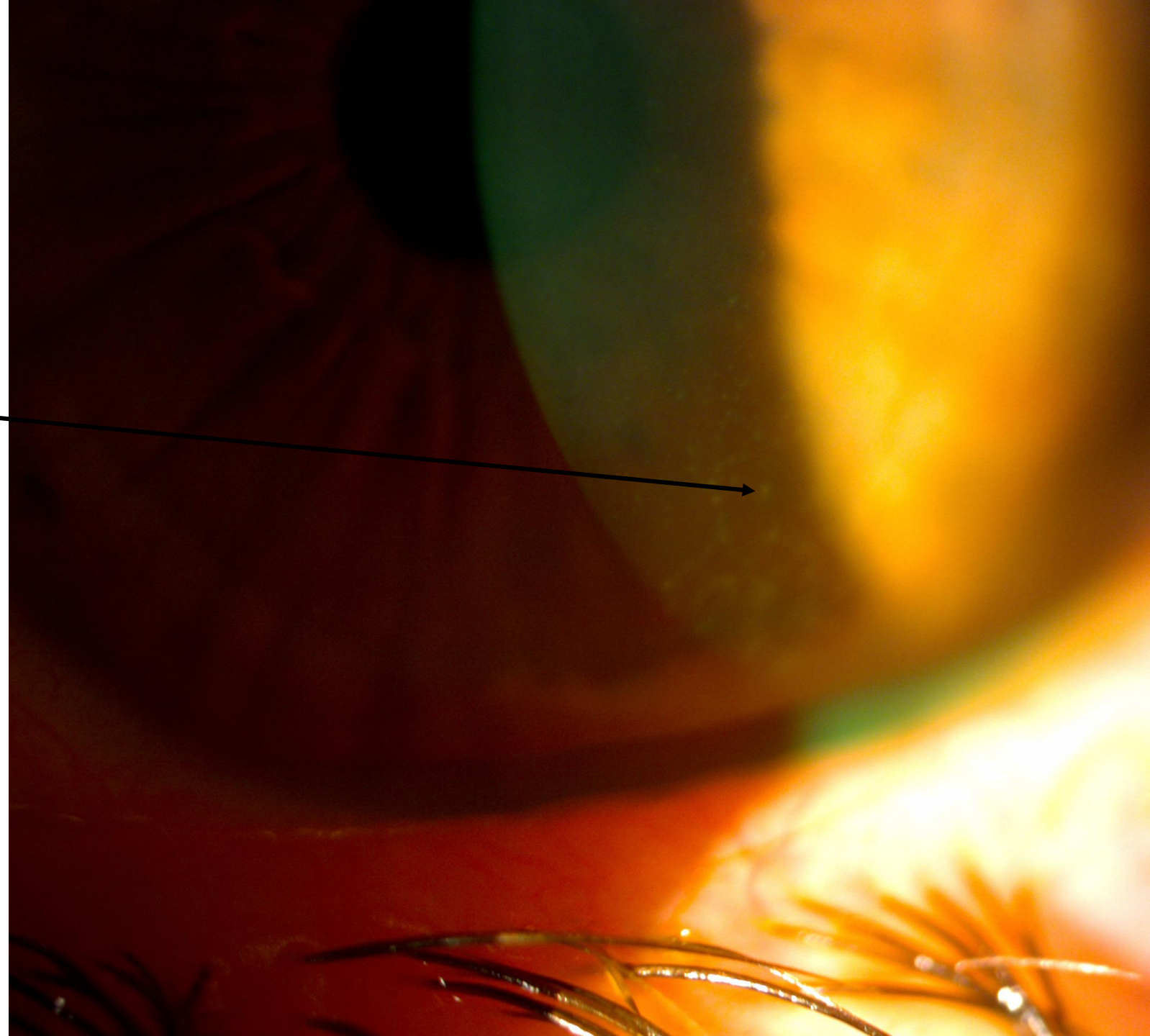
Clinical signs of corneal disease

- Corneal vascularisation: Superficial / Deep
- Corneal edema
- Striae
- Infiltration: superficial / deep
- Pigmentation



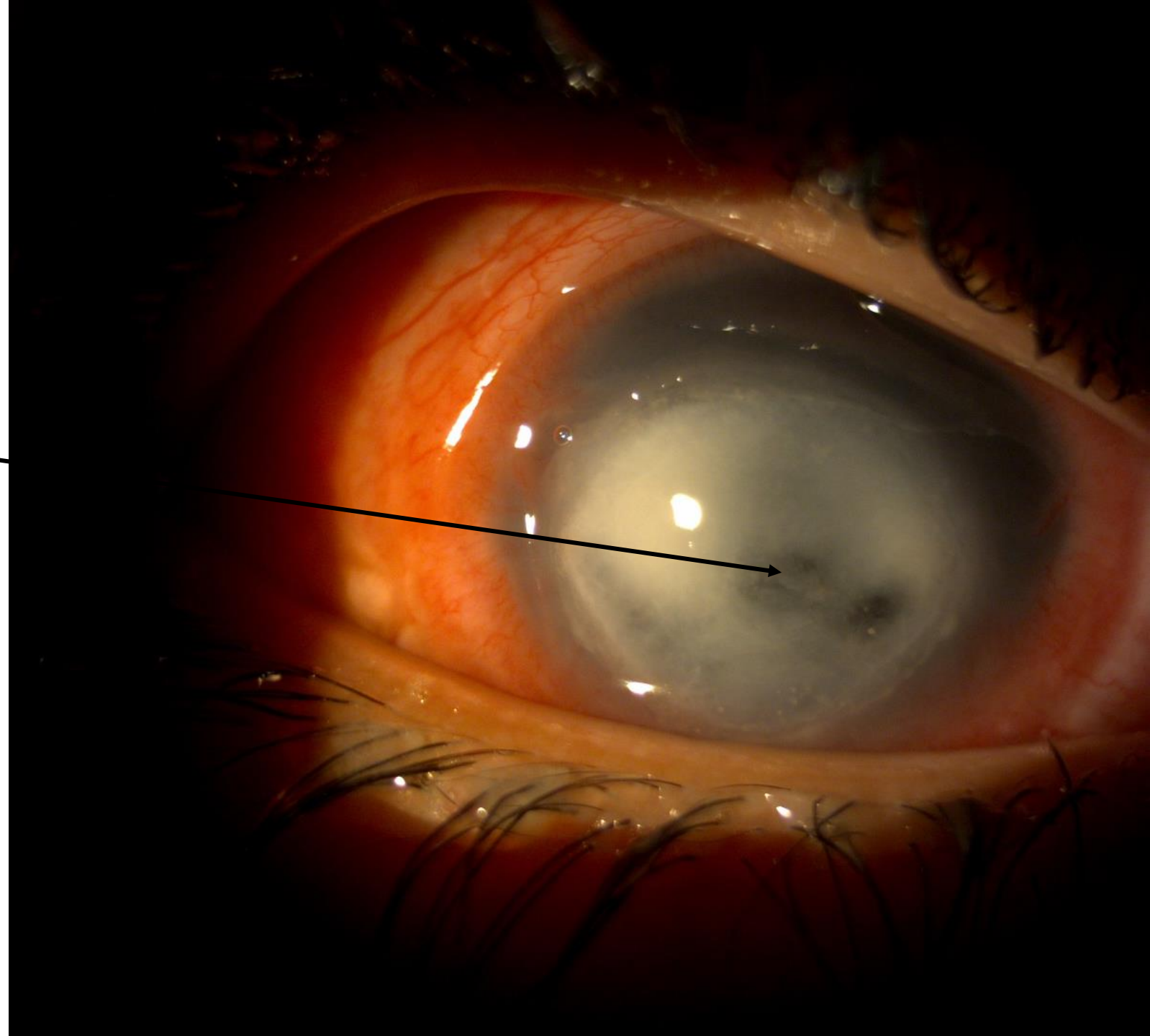
Clinical signs of corneal disease

- Keratitis superficialis punctata



Clinical signs of corneal disease

- Ulcer



2.Corneal topography (Keratometer)

- Provides a colour-coded map of corneal surface
- The power in dioptries of the steepest and flattest meridians and their axes are calculated and displayed
- Steep curvatures (high diopries) are coloured red and orange
- Flat curvatures (low dioptries) are coloured viollet and blue
- Normal cornea - yellow-green spectrum

Indications:

- To quantify irregular astigmatism
- to diagnose early keratokonus
- to evaluate changes in corneal shape after refractive surgery, corneal grafting or cataract extractions

3. Pachymetry

- display the **thickness** of the cornea, usually in micrometres / 550 mikrom./

4. Specular microscopy

- *Studies the changes in different layers of the cornea under magnification which is 100 times greater than the slit-lamp biomicroscopy (corneal endothelium)*
- Celular shape, size, density and distribution
- Normal endothelial shape: *hexagonal*
- **Indications**: prior to intraocular surgery evaluation of donor corneas before penetrating keratoplasty demonstrate corneal diseases and dystrophies (corneal oedema, cornea guttatta, etc.)

5. Corneal Sensitivity

- simple preliminary examination of corneal sensitivity with a distended cotton swab

- This examination confirm the diagnosis in the presence of a suspected *viral infection* of the cornea or *trigeminal or facial neuropathy* (these disorders are associated with reduced corneal sensitivity)

Corneal degenerations and dystrophies

Arcus senilis (*GERONTOXON)*

- *Extracellular lipid infiltration at the corneal periphery*
- is the most common peripheral corneal opacity
- it frequently occurs without any predisposing systemic condition in elderly individuals
- may be associated with dyslipidaemia in younger patients (arcus juvenilis)

- **It first appears** inferiorly, then superiorly, and eventually encircles the cornea
- It appears as a ***grayish-white infiltrate*** separated from sclera by a clear interval of Vogt (1 mm)
- It does not require any treatment as it does not affect the vision or vitality of the cornea

Band keratopathy

- *deposition of calcium salts in the subepitelial space*
- **Causes**
  - association with hyperthyroidism, vitamin D poisoning or sarcoidosis
  - hypercalcaemia, chronic uveitis, chronic glaucoma, interstitial keratitis etc.

- A continuous band lies in the interpalpebral area starting in the inner and outer side

- **Treatment:**

- repeated application of calcium binding agent EDTA (ethylenediaminetetraacetic acid)

- Excimer laser phototherapeutic keratectomy has been effectively performed to treat more extensive cases

Fuchs endothelial dystrophy

- *Bilateral accelerated endothelial cell loss*
- More common in women
- Onset: old age
- **The clinical features are divided into four stages**
    - **a)** **Stage of cornea guttata:** excrescences“ of Descemet membrane secreted by abnormal endothelial cells
    - **b)** **Oedematous stage :** endothelial decompensations leads to central *stromal oedema* and blurred vision
    - **c)** **Stage of bullous keratopathy:** epithelial oedema, microcysts, bullae
    - **d)** **Stage of scarring**

Fuchs endothelial dystrophy

- **Treatment**
- topical sodium chloride 5% drops or ointment
- Ruptured bullae can be made more comfortable by the use of bandage contact lenses, cycloplegia, antibiotic ointment and lubricants
- Posterior lamellar (e.g. Descemet membrane-stripping endothelial keratoplasty – DSAEK – or Descemet membrane endothelial keratoplasty – DMEK)
- Penetrating keratoplasty (PKP)

Ectatic conditions

Keratoconus

- *Ectatic, progressive disorder in which the cornea assumes a conical shape secondary to stromal thinning and protrusion*

- Most patients do not have a positive family history
- **Onset:** in puberty with unilateral impairment of vision due to progressive myopia and astigmatism
- *50% of fellow eyes will progress to keratokonus within 16 years*

Associatons:

- Down, Turner, Ehlers Danlos, Marfan syndrome
- Osteogenesis imperfecta, mental retardation

Clinical signs:

- ***Oil droplet reflex***
- ***Vogt striae-*** deep stromal stress lines
- ***Fleisher ring-*** epithelial iron deposit (hemosiderin)
- **„ scissoring“ reflex**
- ***Rizutti’s sign*** or a conical reflecton on nasal cornea when a penlight is shone from the temporal side
- ***Axenfeld's sign* -** hyposensitive cornea
- ***Munson sign*** – bulging of the lower lid in downgaze

ACUTE HYDROPS

- *Rupture in Descement membrane – influx of aqueous into the cornea - corneal oedema*

- **<u>Acute episodes are treated with:</u>**
  - hypertonic saline
  - soft contact lenses
  - KP should be deferred till the oedema has resolved
- Heals within 6 -10 weeks
- *Healing may result in improved visual activity as result of scarring and flattering of the cornea*

Keratoconus

- **Therapy**
- Soft contact lenses - early cases
- Rigid contact lenses - for higher degree of astigmatism
- Keratoplasty- corneal transplantation
- Intracorneal ring segment implantation
- Corneal collagen cross-linking- riboflavin + ultraviolet-A light

CORNEAL INFLAMMATION

(keratitis)

Morphological and etiological classification

- Superficial
  - Infectious
    - Bacterial
    - Fungal
    - Viral
    - Acanthamoeba keratitis
  - Non-infectious
    - Allergic k.
    - Exposure k.
    - Traumatic
- Deep
  - Ulcer (bacterial, fungal, viral, ...)
  - Corneal abscess
  - Stromal k.

Bacterial keratitis

- Over 90% of all corneal inflammations are caused by bacteria.
- Is uncommon in a normal eye, usually develops when the ocular defences have been compromised

- **Protective mechanisms of the cornea are:**
  - Reflexive eye closing
  - Flushing effect of tear fluid (lysozyme)
  - hydrophobic epithelium which forms a diffusion barrier
  - Epithelium that can regenerate quickly and completely

Etiology

The most common pathoges are:

- *(Pseudomonas aeruginosa, SPA, Str. Pyogenes, pneumoniae,..)*

Bacteria that can penetrate an apparently normal corneal epithelium are:

- *(N.meningitis, gonorrhoeae, Cl diphtheriae, H.influenzae)*

Corneal defect :

- contact lens wearer, trauma, foreign body

ocular surface disease

- (dry eye, chronic blepharitis, etc.)

Systemic disease

- immunosupression, DM, etc.,

Clinical manifestation

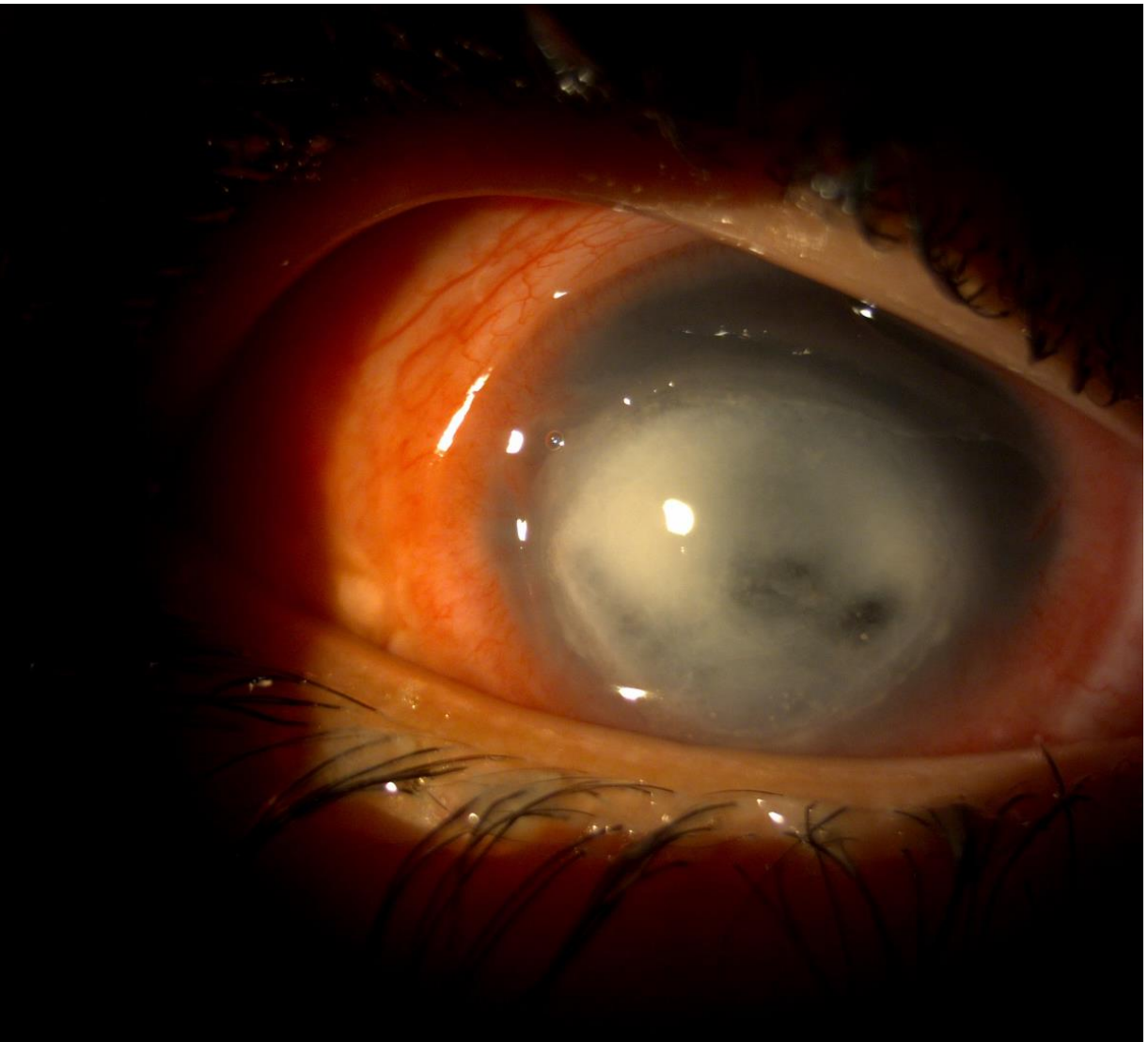
Symptoms:

- Pain
- photophobia
- blurred vision
- lacrimation

Signs:

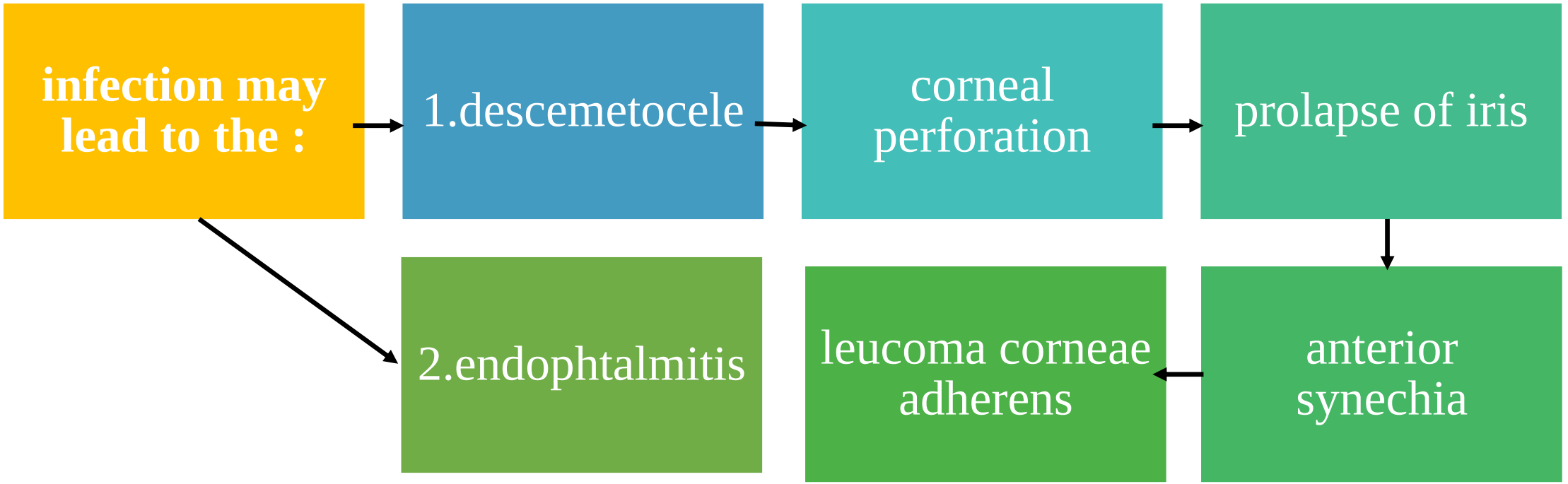
- The lids are red and swollen
- blepharospasm
- epithelial defect
  - circumcorneal injection
  - stromal oedema
  - Precipitates, hypopyon
  - posterior synechiae

Corneal ulcer



- *Deep corneal defect*
- **Etiology:**
  - Bacterial, Viral, fungal,...
- ***Signs***
  1. Cornea is dull and hazy
  2. A greyish white or yellow disc is in the centre
  3. Hypopyon pus in anterior chamber (reaction of the uvea)
  4. conjunctival and ciliary congestion is usually present.
  5. The lids are red and swollen, blepharospasm
- ***Ulcer + hypopyon = ulcus corneae serpens (bacterial)***

Corneal ulcer : Progression



Corneal ulcer

Diagnosis: history, symptoms, signs, cultivation, Gram staining

Healing process:

- Superficial /deep vascularisation
- Leucoma
- Anterior synechiae
- ***leucoma corneae adherens***

leucoma + ant. synechiae

Corneal ulcer

Treatment:

- Topical antibiotics - hourly for 24-48 hours
- Oral antibiotics (threat of perforation)
- Mydriatics (are used to prevent the formation of posterior synechiae)
- Topical steroids ? (fungal infection excluded)
- Contact lens

- Surgical:
  - tarsoraphy
  - Conjunctival flap
  - amniotic membrane transplantat

Fungal keratitis

- *Major cause of visual loss in tropical and developing countries*

Main types are:

- Filamentous fungi: *Aspergillus,Fusarium,..(produce hyphae)*
- Yeasts- *Candida (unicellular)*

Risc factors:

- trauma
- chronic ocular surface disease and epithelial defects
- DM
- Imunosuppression
- hydrofilic contact lenses
- diagnosis often delayed

Fungal keratitis

Symptoms:

- foreign body sensation, blurred vision, photophobia, discharge

Signs:

- grey-yellow stromal infitrate with indistinct margins, progressive, satellite lesions, hypopyon
- yellow-white infiltrate, dense suppuration

Investigations: (before antifungal therapy) Gram and Giems staining, cultures, histology

Treatment: removal of epithelium

- Topical antifungals agents : natamycin, econazol, amphotericin B, miconazol
- systemic antifungals: severe keratitis or endophtalmitis

Viral keratitis (herpes simplex keratitis)

- Major cause of unilateral corneal scarring worldwide
- Most common infectious cause of corneal blindness in developed countries
- HSV-1 – waist, lips, eye / HSV-2- genital herpes- occasionally transmitted to the eye through infected secretions or at birth
- **Risk of reccurence:** 10% at 1 year, 50% at 10 years
- **Clinical signs:** - **epithelial keratitis**
  - **disciform keratitis**
  - **stromal necrotic keratitis**
  - **metaherpetic ulceration**

Epithelial keratitis *(Dendritic, geographic)*

- the most common presentation
- Result of virus replication

- **<u>Symptoms :</u>**
  - discomfort
  - blurred vision
  - watery eyes

Epithelial keratitis *(Dendritic, geographic)*

Signs:

- begins as an superficial punctate keratitis
- central desquamation results in linear branching ulcer (located mostly centrally)
- bed of the ulcer stains well with fluorescein
- *reduced corneal sensation !!!*
- topical steroids may allow progression

- **Diagnosis:** history, symptoms and signs, debridement, viral culture
- **Treatment:** topical antiviral agents (ointment, drops)

Disciform keratitis *(endothelitis)*

Signs:

- stromal oedema with overlying epithelial oedema
- immune ring of stromal haze
    - (viral antigen plus host antibody complexes)

Treatment:

- topical steroids with antiviral cover

Stromal necrotic keratitis

Signs

- stromal necrosis
- melting
- anterior uveitis
- scarring
- vascularization

- NB
- *Acute deterioration and melting might indicate secondary microbial infection*

Metaherpetic ulceration

- *result of a non-healing epithelial defect after prolonged topical treatment*

Varicella- zoster virus

- **Varicella- zoster virus(VZV) causes:**
    1. chickenpox (varicella)
    2. shingles (herpes zoster)
- After an episode of chickenpox the VZV travels in a retrogarde manner to the dorsal root and cranial nerve sensory ganglia, where it may remain dormant for decades
- **Reactivation**
    - when the VZV specific cell-mediated immunity has faded
- **HZO**
    - describes shingles involving the dermatome supplied by the ophtalmic division of the 5th tranial(trigeminal) nerve

Varicella- zoster virus

- **Signs**
- **Hutchinson's rule**
  - Ocular involvement is usually associated with eruption of vesicles on the skin of tip of the nose (nasociliary branch) during the acute stage.
- **Acute epithelial keratitis**
  - Micro dendritic epithelial ulcers, Unlike herpes simplex, these ulcers are small, peripheral, stellate and with tapered ends, i.e. without rounded bulbs

Th: oral acyclovir, systemic steroids, symptomatic treatment

- *Acute epithelial keratitis develops in 50% patients within 2 days of the onset maculopapular rush*

Interstitial keratitis

- *Interstitial keratitis (IK) is an inflammation of the corneal stroma without primary involvement of the epithelium or endothelium*
- *is a non-ulcerating inflammation of the corneal stroma, the host's allergic / immune reaction to foreign antigens*

- **Bacterial etiologies:**
  - Syphilis, Lyme Disease, Tuberculosis
- **Viral etiologies:**
  - Herpesviridae - Herpes simplex, Herpes zoster, Epstein- Barr

Interstitial keratitis

- **Signs**
  - Conjunctival injection
  - Corneal haze - either diffuse, sectoral, central or circumferential.
  - White cell infiltration without significant necrosis or suppuration
  - Stromal neovascularization
  - Ghost vessels - when the disease is quiescent

- **Symptoms**
  - Decreased vision highly dependent on the extent an location of involvement
  - Significant photophobia and pain are highly characteristic

Interstitial keratitis

- **Medical therapy:**
  - Topical /Systemic corticosteroids
  - Topical /Systemic ATB
  - Topical /Systemic antivirals
  - Cycloplegics