Cornea

Anatomy and physiology

The cornea is a complex structure which, as well as having a protective role, is responsible for about three-quarters of the optical power of the eye. The normal cornea is free of blood vessels; nutrients are supplied and metabolic products removed mainly via the aqueous humour posteriorly and the tears anteriorly. The cornea is the most densely innervated tissue in the body, a subepithelial and a deeper stromal nerve plexus are both supplied by the first division of the trigeminal nerve.

Dimensions

The average corneal diameter is 11.5 mm vertically and 12 mm horizontally. It is 540 µm thick centrally on average, and thicker towards the periphery.

Structure

The cornea consists of the following five layers:

1- **The epithelium** is stratified squamous and non-keratinized, and is composed of:

1- A single layer of columnar basal cells attached by hemidesmosomes to an underlying basement membrane.

2- Two to three rows of 'wing' cells.

3- Two layers of squamous surface cells.

○ The surface area of the outermost cells is increased by microplicae and microvilli that facilitate the attachment of the tear film and mucin. After a lifespan of a few days superficial cells are shed into the tear film. Because of its ability to regenerate, the epithelium does no scar.

2- **The Bowman layer** is the acellular superficial layer of the stroma, and is formed from collagen fibers .It does not regenerate when damaged.

3- **The stroma** makes up 90% of corneal thickness. It is composed of collagen-producing modified fibroblasts (keratocytes), regularly orientated layers of collagen fibrils and ground substance. The stroma can scar, but cannot regenerate following damage.

4-Descemet membrane is a discrete sheet composed of a fine latticework of collagen fibrils. It has regenerative potential

5-The endothelium consists of a monolayer of hexaygonal cells. Endothelial cells maintain corneal deturgescence throughout life by pumping excess fluid out of the stroma. The young adult cell density is about 3000 cells/mm2. The number of cells decreases with age and neighbouring cells enlarge to fill the space; the cells cannot regenerate.

Signs of corneal disease

Superficial lesions

• **Punctate epithelial erosions (PEE)** tiny epithelial defects that stain with fluorescein stain, are generally an early sign of epithelial compromise. **Causes** include a variety of stimuli (Non specific sign); the location of the lesions may give an indication of etiology:

○ Superior – vernal disease, superior limbic keratoconjunctivitis.

○ Interpalpebral – dry eye (can also be inferior), reduced corneal sensation and ultraviolet keratopathy.

○ Inferior – chronic blepharitis, corneal exposure, eye drop toxicity, self-induced, aberrant eyelashes and entropion.

• **Punctate epithelial keratitis (PEK)** appears as granular, opalescent, swollen epithelial cells, with focal intraepithelial infiltrates. They are visible unstained but stain well with rose Bengal and variably with fluorescein. **Causes** include: Infections (mostly viral) other like eye drop toxicity.

• **Filaments** strands of mucus admixed with epithelium, attached at one end to the corneal surface, that stain well with rose Bengal). The unattached end moves with each blink. Grey subepithelial opacities may be seen at the site of attachment. **Dry eye is by far the most common cause**; others include superior limbic keratoconjunctivitis, neurotrophic keratopathy, long-term ocular patching and essential blepharospasm.

• **Epithelial oedema** manifest with loss of normal corneal lustre. The cause is endothelial decompensation, including that due to severe acute elevation of IOP. Epithelial vesicles and bullae are seen in severe cases.

• **Pannus** describes superficial neovascularization accompanied by degenerative subepithelial change.

Deep lesions

- **Stromal infiltrates** are yellow or grey–white opacities usually associated with limbal or conjunctival hyperaemia. They are stromal foci of acute inflammation composed of inflammatory cells, cellular and extracellular debris. This lesions may be infective or sterile lesions.
- **Stromal oedema** usually coexists with inflammatory Infiltration and associated with increased corneal thickness and decrease in transperanscy.Four important causes of central corneal oedema are disciform keratitis, keratoconus, fuchs' dystrophy and intraoperative damage to corneal endothelium**.**
- **Ulceration** refers to tissue excavation associated with an epithelial defect, usually with infiltration and necrosis.
- **Stromal vascularization** occurs in response to a wide variety of stimuli.
- **Folds in Descemet membrane** Causes include inflammation, trauma (including surgery) and ocular hypotony
- **Breaks in Descemet membrane** may be due to corneal enlargement, keratoconus and birth trauma.

Corneal special investigations

- ***Pachymetry*** ; measure corneal thickness.
- ***Specular microscopy***; It is mainly used to assess the endothelium, which can be analysed for cellular size, shape, density and distribution.
- ***Keratometry ;*** measure corneal curvature.
- ***Keratoscopy*** ;detects abnormalities of corneal shape. If corneal surface is regular, the reflected image will be consist of uniform concentric rings; if irregular, the reflection will be distorted. The following are main types of keratoscopy:

  ***1*-Plasido 's disc.**

  ***2*-Photokeratoscopes.**

  ***3*- computer-assisted Photokeratoscopes (Corneal topography).**

  - Anterior segment optical coherence tomography (OCT) and ultrasound biomicroscopy can also be used to image the cornea.

Microbial keratitis

- **BACTERIAL KERATITIS**

Predisposing factors

The pathogens able to produce corneal infection in presence of an **intact epithelium** are *Neisseria gonorrhoeae*, *Neisseria meningitidis*, *Corynebacterium diphtheriae* and *Haemophilus influenza and listeria sp* .Other bacteria are capable of producing keratitis only after loss of corneal epithelial integrity associated with the **following risk factors:**

- **Contact lens wear**, is the most important risk factor. *Pseudomonas aeruginosa* is responsible for over 60% of contact lens-related keratitis.
- **Trauma**, including surgery.
- **Ocular surface disease** such as herpetic keratitis, bullous keratopathy, dry eye, chronic blepharitis, trichiasis ,entropion, corneal exposure and severe allergic eye disease .
- **Other factors** include local or systemic immunosuppression, diabetes and vitamin A deficiency.

Common pathogens include: *Pseudomonas aeruginosa, Staphylococcus aureus* and *Streptococci.*

CLINICAL FEATURES

- **Presentation** is with pain, photophobia, blurred vision and mucopurulent or purulent discharge.
- **Signs**
  - An epithelial defect with infiltrate and significant circumcorneal injection .
  - Stromal oedema, folds in Descemet membrane and anterior uveitis, commonly with a hypopyon and posterior synechiae in moderate–severe keratitis., Chemosis and eyelid swelling in moderate–severe cases
  - Scleritis and increased IOP can develop,
  - Severe ulceration may lead to perforation and endophthalmitis.

MANAGEMENT

A bacterial corneal ulcer is a sight-threatening condition which demands urgent identification and eradication of the causative organism**.**

1- History for risk factors (e.g. contact lens wear, trauma. …)

2- **Hospital admission** should be considered for patients who are not likely to comply or are unable to self-administer treatment. It should also be considered for aggressive disease, particularly if involving an only eye.

3- **Corneal scraping for staining and Culture & sensitivity.**

4- **Antibiotics**

- **Initial (Empirical) topical Antibiotics treatment** ,while waiting the culture and sensitivity reports, either Fluoroquinolone or (cefuroxime + 'fortified' gentamicin), in order to cover common Gram-positive and Gram-negative pathogens, then choice of the antibiotics according to culture and sensitivity reports;
- ***Gram-positive organism*** Cefuroxime or vancomycin **.**
- ***Gram-negative organism*** 'Fortified' gentamicin , fluoroquinolone or ceftazidime.
- *There is no need to change the initial therapy if this has induced a favourable response, even if cultures show a resistant organism.*
- **Subconjunctival antibiotics** are usually only indicated if there is poor compliance with topical treatment.
- **Systemic antibiotics** are not usually given, but may be appropriate in the following circumstances: 1- Potential for systemic involvement(*N. meningitides*, *N. gonorrhoeae and H. influenza).* 2- Severe corneal thinning. 3- Scleral involvement

5- **Mydriatics** (cyclopentolate 1%, homatropine 2% or atropine 1%) are used to prevent the formation of posterior synechiae and to reduce pain.

6- **Topical Steroid is** controversial? Steroids reduce host inflammation, improve comfort, and minimize corneal scarring. However, they promote replication of some microorganisms, particularly fungi, herpes simplex and mycobacteria. Steroids also decrease fibroblast activity and inhibiting wound healing, thereby increasing the risk of perforation. For these reasons topical steroids may be initiated only when cultures becomes sterile and there is a clear evidence of improvement.

7- **Therapeutic l keratoplasty may** be considered in cases resistant to medical therapy, or for incipient or actual perforation.

- Improvement is usually heralded by a reduction in eyelid oedema and chemosis, shrinking of the epithelial defect, decreasing infiltrate density and a reduction in anterior chamber signs.
  - Subsequent scarring may be severe, including vascularization; in addition to opacification irregular astigmatism may limit vision.

FUNGAL KERATITIS

Two main types of fungi cause keratitis:

- **Yeasts** (e.g. *Candida*), ovoid unicellular organisms that reproduce by budding, are responsible for most cases of fungal keratitis in temperate climates.
- **Filamentous fungi** (e.g. *Fusarium* and *Aspergillus*), multicellular organisms that produce tubular projections known as hyphae. They are the most common pathogens in tropical climates, but are not uncommon in cooler regions; the keratitis frequently follows an aggressive course.

Predisposing factors

Common predisposing factors include chronic ocular surface disease, the long-term use of topical steroids (often in conjunction with prior corneal transplantation), contact lens wear, systemic immunosuppression and diabetes. Filamentary keratitis may be associated with trauma, often relatively minor, involving plant matter or gardening/agricultural tools.

Clinical features

The diagnosis is frequently delayed unless there is a high index of suspicion, and often bacterial infection will initially have been presumed.

- **Symptoms.** Gradual onset of pain, grittiness, photophobia, blurred vision and watery or mucopurulent discharge.
- **Candidal keratitis**
  - Yellow–white densely suppurative infiltrate is typical.
- **Filamentous keratitis**
  - Grey or yellow–white stromal infiltrate with indistinct fluffy margins.
  - Progressive infiltration, often with satellite lesions.
  - Feathery branch-like extensions or a ring-shaped infiltrate may develop.
  - Rapid progression with necrosis and thinning can occur.

- Penetration of an intact descemet membrane may occur and lead to endophthalmitis without evident perforation. **Other features** include anterior uveitis, hypopyon, endothelial plaque, raised IOP, scleritis.

Treatment

Improvement may be slow in comparison to bacterial infection.

- **General measures** are as for bacterial keratitis although hospital admission is usually required.

A culture result can be obtain within 48-72 hours but sensitivities take about a week.

1-Initial (empirical) treatment is with a topical broad –spectrum agent such as amphotericin B 0.15%, econazole 1%; and natamycin 5%, then according to culture and sensitivity report.

Because most antifungals are only fungistatic, treatment should be continued for at least 12 weeks.

2-A broad-spectrum antibiotic might also be considered to address or prevent bacterial co-infection.

3-Subconjunctival and Systemic antifungals may be given in severe cases.

4-Therapeutic keratoplasty is considered when medical therapy is ineffective or following perforation.

VIRAL KERATITIS

Herpes simplex keratitis

Herpetic eye disease is the most common infectious cause of corneal blindness in developed countries. As many as 60% of corneal ulcers in developing countries may be the result of herpes simplex virus.

Herpes simplex virus (HSV)

HSV is enveloped with a cuboidal capsule and has a linear double-stranded DNA genome. The two subtypes are *HSV-1* and *HSV-2*, and these reside in almost all neuronal ganglia.

HSV-1 causes infection above the waist (principally the face, lips and eyes),usually acquired by kissing or close contact with a person who either has a cold sore (herpes labialis) or is shedding the virus asymptomatically

HSV-2 causes venereally acquired infection (genital herpes). Rarely *HSV-2* may be transmitted to the eye through infected secretions, either venereally or at birth (neonatal conjunctivitis). HSV transmission is facilitated in conditions of crowding and poor hygiene.

Primary infection, without previous viral exposure, usually occurs in childhood and is spread by droplet transmission, or less frequently by direct inoculation. Due to protection by maternal antibodies, it is uncommon during the first 6 months of life, most primary infections with HSV are subclinical or cause only mild fever, malaise and upper respiratory tract symptoms. Rarely immunodeficient patients, the infection becomes generalized and even life threatening. **After primary infection** the virus is carried to the sensory ganglion for that dermatome (e.g. trigeminal ganglion for HSV-1, spinal ganglion for HSV-2) where latent infection is established. In some patients clinical reactivation may occur when the virus replicates and is transported in the sensory axons to its target tissue, causing recurrent infection (herpes labialis, herpetic keratitis, genital herpes).

Primary ocular infection

Typically occurs in children between the ages of 6 months and 5 years, and may be associated with generalized symptoms of a viral illness.

Clinical features

1-Blepharoconjunctivitis is usually benign and self-limited and, in children, it may be the only manifestation of primary herpetic infection. The skin lesions typically involve the lids and periorbital area.Initially, they consist of vesicles which rapidly form superficial crusts and then heal without scarring. The conjunctivitis is unilateral, acute, follicular, and associated with a watery discharge and preauricular adenopathy.

2- keratitis develops within a few days in about 50% of patients with Blepharoconjunctivitis.

Treatment

In patients with Blepharoconjunctivitis, topical antiviral ointment should be applied prophylactically to the eye five times /day for about 21 days to prevent keratitis.

Epithelial keratitis

Clinical features

- Epithelial (**dendritic or geographic**) keratitis is associated with active virus replication.
- Early cases shows opaque epithelial cells arranged in a coarse punctate or stellate pattern.
- Central desquamation results in a linear-branching (**dendritic**) ulcer, most frequent located centrally; the branches of the ulcer have characteristic **terminal buds** and its bed stains well with **fluorescein**.
- **Corneal sensation is reduced**.
- Inadvertent topical steroid treatment may promote progressive enlargement of the ulcer to a **geographical or 'amoeboid' configuration.**

Differential diagnosis of dendritic ulceration includes;

1- Herpes zoster keratitis,

2-healing corneal abrasion
3-soft contact lenses.
4-toxic keratopathy secondary to topical medication.

Treatment

The majority of dendritic ulcers will eventually heal spontaneously without treatment. Treatment of HSV disease is predominantly with nucleoside (purine or pyrimidine) analogues that disrupt viral DNA.

1. **Topical antiviral.** The most frequently used drugs are **aciclovir 3% ointment** and **ganciclovir 0.15% gel**, each administered five times daily. The drugs are relatively non-toxic, they have approximately equivalent effect, acting preferentially on virus-laden epithelial cells, and penetrating effectively into the stroma; 99% of ulcers heal within two weeks.
2. **Debridemen**t may be used for resistant cases. A topical antiviral agent should be used in conjunction.

HERPES ZOSTER OPHTHALMICUS

Herpes zoster is a common infection caused by varicella-zoster virus. Herpes zoster ophthalmicus (HZO) is the term used for shingles involving the dermatome supplied by the ophthalmic division of the fifth cranial (trigeminal) nerve. Varicella-zoster virus (VZV) causes both chickenpox (varicella) and shingles (herpes zoster); VZV belongs to the same subfamily of the herpes virus group as HSV – the viruses are morphologically identical but antigenically distinct. After an episode of chickenpox the virus travels in a retrograde manner to the dorsal root and cranial nerve sensory ganglia, where it may remain dormant for decades, with reactivation thought to occur after VZV-specific cell-mediated immunity has faded, virus migrates back down the sensory nerves to the skin and eye where it cause the characteristic lesions.

Risk of ocular involvement

1- The Hutchinson sign. describes involvement of the skin supplied by the external nasal nerve, a branch of the nasociliary nerve supplying the tip, side and root of the nose.The sign correlates strongly with ocular involvement
2- Age. HZO occurs most frequently in the sixth and seventh decades. In the elderly, signs and symptoms tend to be more severe and of longer duration.
3- AIDS patients. tend to have more severe disease .The development of shingles in children or young adults classically has prompted a search for immunodeficiency or malignancy.

Acute shingles

A prodromal phase precedes the appearance of the rash. It lasts 3–5 days and is characterized by tiredness, fever, malaise and headache. Symptoms involving the affected dermatome vary from a superficial itching, tingling or burning sensation to a severe boring or lancing pain that is either constant or intermittent. Older patients with early severe pain and a larger area of involvement are at particular risk of post-herpetic neuralgia**.**

Skin lesions

Painful erythematous areas with **a maculopapular rash** develop. Within 24 hours, groups of **vesicles** appear and these become confluent over 2–4 days. The vesicles often pass through a **pustular phase** before they **crus**t and dry after 2–3 weeks. The lesions heal to leave residual skin destruction and **depigmented scars.**

Treatment of skin lesions

1-Oral antiviral treatment

Aciclovir (800 mg five times daily for 7–10 days) has been the mainstay of treatment. Optimally given within 72 hours of rash onset, reduces the severity and duration of the acute episode and the risk of post-herpetic neuralgia. The incidence of late ophthalmic complications is also reduced by about 50%.

2-Topical aciclovir cream, and a steroid-antibiotic combination can be used three times daily. until the crusts have separated.

Clinical features of Eye disease

1-Acute eye disease 2-Chronic eye disease 3- Relapsing eye disease

<u>Acute eye disease</u>

1-Acute epithelial keratitis develops in over 50% of patients within 2 days of the onset of the rash and usually resolves spontaneously within a few days. It is characterized by dendritic lesions that are smaller and finer than herpes simplex dendrites, and have tapered ends without terminal bulbs .The lesions stain better with rose Bengal than with fluorescein. Treatment, if required, is with a topical antiviral.

2-Conjunctivitis (follicular and/or papillary) is common; it often occurs in conjunction with lid margin vesicles. Treatment is not required in the absence of corneal disease, though some practitioners give topical antibiotic and/or antiviral prophylaxis.

3-Episcleritis occurs at the onset of the rash and usually resolves spontaneously.

4- Scleritis uncommon but may develop at the end of the first week. Treatment is with oral flurbiprofen 100 mg three times daily. Oral steroids with antiviral cover may be required for severe involvement.

5-*Nummular keratitis*. It is characterized by fine granular subepithelial deposits surrounded by a halo of stromal haze). The lesions fade in response to topical steroids.

6-*Stromal (interstitial) keratitis* *develops* in about 5% 3 weeks after the onset of the rash. It usually responds to topical steroids.

7-**Disciform keratitis** (immune-mediated endotheliitis), Treatment is with topical steroids.

8-Anterior uveitis.

9-Posterior uveitis.

Chronic eye disease

1-Neurotrophic keratopathy.

2- Mucous plaque keratitis.

3- Lipid degeneration

4- Scleritis.

5-Subconjunctival scarring.

6-Eyelid scarring.

Relapsing eye disease

In the relapsing phase lesions may reappear years after an acute episode; eyelid scarring may be the only diagnostic clue. Reactivation of keratitis, episcleritis, scleritis or iritis can occur.

Neurological complications may require intravenous antivirals and systemic steroids.

○ Cranial nerve palsies affecting the third (most common), fourth and sixth nerves usually recover within 6 months.

○ Optic neuritis is rare.

○ CNS manifestations are rare but include encephalitis, cranial arteritis, and Guillain–Barré syndrome.

Post-herpetic neuralgia

Post-herpetic neuralgia is defined as pain that persists for more than one month after the rash has healed. It develops in up to 75% of patients over 70 years of age. Pain may be constant or intermittent, worse at night and aggravated by touch and heat. It generally improves slowly over time. Neuralgia can impair the quality of life, and may lead to depression of sufficient severity to present a danger of suicide. Patients severely affected should be referred to a specialist pain clinic.

CORNEAL DEGENERATIONS

Arcus senilis

Arcus senilis is the most common peripheral corneal opacity; it frequently occurs without any predisposing systemic condition in elderly individuals, but may be associated with dyslipidaemia in younger patients (arcus juvenilis).

Signs

○ Bilateral stromal lipid deposition, initially in the superior and inferior perilimbal cornea, progressing circumferentially to form a band about 1 mm wide.

Lipid keratopathy

1-**Primary** lipid keratopathy is rare and occurs spontaneously in avascular cornea. It is characterized by white or yellowish stromal deposits consisting of cholesterol, fats and phospholipids.

2-**Secondary** lipid keratopathy is much more common and is associated with previous ocular injury or disease that has resulted in corneal vascularization. The most common causes are herpes simplex and herpes zoster keratitis

Treatment

1- control of the underlying inflammatory disease. then

2- Photocoagulation or needle cautery of feeder vessels.

3- Penetrating keratoplasty may be required in advanced but quiescent disease.

Band keratopathy

Band keratopathy is a relatively common disorder characterized by deposition of calcium salts in the Bowman layer, epithelial basement membrane and anterior stroma.

Causes

1- Ocular; Chronic anterior uveitis (particularly in children), phthisis bulbi,
2- Age-related; affects otherwise healthy individuals.
3- Metabolic; this is rare and includes increased serum calcium and phosphorus.
4-Hereditary; include familial cases and ichthyosis.

Signs

Interpalpebral distribution of calcification (a band-like chalky plaque) containing transparent small holes and occasionally clefts.

Treatment is indicated if vision is threatened or if the eye is uncomfortable. It is important to recognize and treat any underlying condition.

1- Chelation is simple and effective for relatively mild cases and is performed using a microscope. The corneal epithelium overlying the opacity and a solid layer of calcification are first scraped off with forceps. Followed by application a solution of ethylenediaminetetraacetic acid (EDTA).

2- Other modalities: diamond burr, excimer laser keratectomy and lamellar keratoplasty for more extensive and deeper lesion .

CORNEAL DYSTROPHIES

The corneal dystrophies are a group of bilateral, symmetrical, inherited corneal opacifying disorders. Dystrophies begin early in life but may not become clinically apparent until later. They tend to be slowly progressive. Many of which are associated with decreased vision and discomfort.

Classification

Anterior dystrophies

Epithelial basement membrane (map-dot-fingerprint) is the most common corneal dystrophy
Reis–Bücklers dystrophy
Meesmann dystrophy

Stromal dystrophies

Lattice dystrophy
Macular dystrophy
Granular dystrophy

Posterior dystrophies

Fuchs endothelial corneal dystrophy
Posterior polymorphous corneal dystrophy
Congenital hereditary endothelial dystrophy

Cogan (epithelial basement membrane) dystrophy

Epithelial basement membrane (map-dot-fingerprint) is the most common corneal dystrophy.
Signs Bilateral dot-like , microcystic , map-like, fingerprint-like lines epithelial lesions.
Complications in the form of recurrent corneal erosions develop in about 10% of patients, usually after the age of 30 years. The remainders are asymptomatic throughout life.

Granular corneal dystrophy

Inheritance is autosomal dominant.
Onset is in the 1st decade but vision is usually not affected in the early stage of the disease. Recurrent erosions are uncommon**.**

Signs

○ Discrete white central anterior stromal deposits resembling sugar granules, breadcrumbs ***separated by clear stroma.***

○ Gradual increase in number and size of the deposits with deeper and outward spread, ***sparing the limbus***

Treatment by penetrating or deep lamellar keratoplasty is usually required by the fifth decade.

Macular corneal dystrophy

Is the least common, but the most serious of the three classic stromal dystrophies.

Inheritance. Autosomal recessive

Onset. Early (end of first decade) visual deterioration; recurrent erosions are very common.

Signs

○ Dense poorly delineated greyish-white spots centrally and peripherally. **There is no clear zone between opacities.**

○ There is eventual involvement of full-thickness stroma, **extending to the limbus.**

Treatment. Penetrating keratoplasty. Recurrence is common.

CORNEAL ECTASIAS

1- Keratoconus

2- Pellucid marginal degeneration

3- Keratoglobus

Keratoconus

Keratoconus is a progressive disorder in which the cornea assumes a conical shape secondary to stromal thinning and protrusion. **The hallmark of keratoconus** is central or paracentral stromal thinning, accompanied by apical protrusion and irregular astigmatism. The condition starts at around puberty and progresses slowly thereafter, although it may become stationary at any time. Nearly all cases are bilateral, but the severity is asymmetrical. The role of heredity has not been clearly defined and most patients do not have a positive family history. Offspring appear to be affected in only about 10% of cases and autosomal dominant transmission with incomplete penetrance has been proposed.

Associations

1- ***Systemic disorders*** include: Down, Turner, Ehlers–Danlos and Marfan syndromes, atopy, osteogenesis imperfecta, mitral valve prolapse.

2- ***Ocular associations*** include: vernal keratoconjunctivitis, aniridia, ectopia lentis, retinitis pigmentosa. persistent eye rubbing.

Histopathologically, keratoconus shows the following:

1. **Fragmentation of Bowman layer**
2. **Thinning of the stroma and overlying epithelium**
3. **Folds or breaks in Descemet's membrane**
4. **Variable amounts of diffuse scarring**

Presentation

Presentation is **typically between the ages of 10 and 20 years**, with unilateral impairment of vision due to progressive myopia and astigmatism. The patient may report the need for frequent changes in spectacle correction or a decreased tolerance to contact lens wear.

Early signs of keratoconus.

1. ***Ophthalmoscopy***: Oil droplet reflex
2. ***Retinoscopy*** shows an irregular Scissoring of the red reflex.
3. ***keratometry*** : It can be graded as mild (<48 D), moderate (48–54 D) or severe (>54 D).
4. ***Photokeratoscopy or plasido's disk*** shows irregularity of reflected ring counours.
5. ***Slit lamp biomicroscopy*** shows very fine, vertical, deep stromal stress lines (Vogt striae), which disappear with pressure on the globe.
6. ***Corneal topography*** shows irregular astigmatism and is the most sensitive method of detecting early keratoconus and for monitoring progression

Late signs of keratoconus.

1. Progressive central or paracentral corneal thinning. This associated with poor visual acuity resulting from marked irregular myopic astigmatism with steep keratometry (K) readings.
2. Bulging of the lower lid in down gaze when the patient looks down (Munson sign).

3. Epithelial iron deposits may surround the base of the cone (Fleischer ring).
4. Central and paracentral corneal scarring in severe cases.
5. Acute hydrops is caused by a rupture in Descemet membrane that allows an influx of aqueous into the cornea .Although the break usually heals within 6–10 weeks and the corneal oedema clears, a variable amount of stromal scarring may develop. Acute episodes are initially treated with cycloplegia, hypertonic (5%) saline ointment and patching or a soft bandage contact lens.

Treatment

1. **Spectacles** or **soft contact lenses in** early cases.
2. **Rigid contact lenses** are required for higher degrees of astigmatism.
3. **Corneal collagen cross-linking (CXL)** using riboflavin drops and ultraviolet-A light to stabilize or even reverse ectasia. CXL is commonly used only after progression has been documented.
4. **Intracorneal ring segment implantation.**
5. **Keratoplasty (corneal transplant) is** indicated in patients with advanced progressive disease, especially with significant corneal scarring.