Corneal Problems - Acute and Non-acute

The cornea is the avascular convex, anterior transparent front part of the globe which performs the dual functions of protection of the eye and refraction of light. It is a site of refraction of light entering the eye and provides a clear medium through which the light can travel. It is limited at its periphery by the corneal limbus, where the opaque sclera starts. It is intimately related to the conjunctiva via its epithelium which is continuous (although slightly different in nature) between the cornea and the conjunctiva. Infections, inflammatory conditions and trauma can all potentially extend from one to the other.

Other related separate articles include Recurrent Corneal Erosion Syndrome, Dry Eyes (keratoconjunctivitis sicca), Corneal Foreign Bodies, Injuries and Abrasions, Eye Injuries, Contact Lenses (Types and Care) and Contact Lens Problems.

Structure of the cornea

The cornea is a multilayered structure consisting of (from outside to inside):

- **Anterior corneal epithelium**: a thin, multicellular epithelial tissue layer composed of around six layers of cells (non-keratinised stratified squamous epithelium) of rapidly regenerating cells kept moist by tears. The air/tear film interface is the most significant component of the total refractive power of the eye, so disruption of this surface can reduce acuity. The corneal epithelium is continuous with the conjunctival epithelium. It regenerates constantly, from the bottom layer.
- **Bowman's layer** (also called the anterior limiting membrane): this is a protective acellular collagen layer.
- **Corneal stroma** (or substantia propria): a thicker, transparent layer of collagen and keratocytes which makes up around 90% of corneal thickness.
- **Descemet's membrane** (posterior limiting membrane): a thin acellular layer which acts as the basement membrane to the corneal endothelium, and consists mainly of collagen. There may be an even thinner protective membrane called **Dua's layer** on its surface. This membrane was described in 2013 but its existence is a matter of dispute.
- **Corneal endothelium**: a simple squamous monolayer of cells which regulate fluid and solute transport. These cells do not regenerate but instead stretch to compensate for dead cells.

An ABCDE mnemonic is:

**Anterior corneal epithelium, Bowman's membrane, Corneal stroma, Descemet's membrane, Endothelium.**

Symptoms in corneal problems

- **Pain**: occurs with most corneal problems unless there is gross neuropathy, in which case severe disease may cause minimal discomfort. Severe pain with only a very small apparent defect is a characteristic presentation in patients with the potentially devastating acanthamoebic keratitis.
- **Photophobia**: frequently accompanies pain.
- **Reduced visual acuity**: any lesion affecting the central visual axis or distorting the shape of the cornea will affect visual acuity. **Excess lacrimation** (epiphora) due to pain can also temporarily affect the vision.
- **Red eye**: this frequently accompanies the above symptoms.
- **Systemic symptoms**: these are not unusual in patients with acute corneal disease, particularly headaches, mild nausea and feeling generally run down.
Assessment of the cornea

The cornea in primary care

See separate Examination of the Eye article.

- Test visual acuity of both eyes.
- Observe the cornea in plain light. Note whether there are any areas of gross opacification.
- Check for sensation: twist a clean tissue or cotton ball to a tip and lightly touch the centre of the cornea. This should elicit a brisk and immediate response from the patient.
- Apply fluorescein to look for defects (if suspecting perforation, perform Seidel's test).
- Using a slit lamp, assess the cornea from the anterior (epithelial) surface, through the stroma and to the posterior (endothelial) surface. Look for defects (fluorescein uptake), oedema (area of haziness) and infiltrates (a well-demarcated white lesion within the stroma). Vascularisation may occur over the surface or through the stroma, indicating more long-standing disease.
- Examine the rest of the globe and its adnexae. If the symptoms warrant it, do a full systemic examination.

Further assessment of the cornea in a specialist unit

- Pachymetry - the measurement of corneal thickness, involving placing a measuring probe lightly on the surface of the anaesthetised cornea.
- Specular microscopy - a photographic investigation that enables the corneal endothelial cells to be accurately assessed.
- Corneal topography - maps the surface of the cornea, showing the gradient at each spot and therefore highlighting asymmetries, such as are found in the dystrophic conditions.
- Microbiological investigations - a corneal scrape (clinic) or biopsy (theatre) may be helpful.

Preservative-free ocular lubricants or artificial tears are a good first step in helping to relieve initial discomfort whilst awaiting specialist review. Pain can be addressed with oral analgesics; topical anaesthetics should be avoided as they are toxic to the corneal epithelium if used repeatedly.

Acute corneal problems

Corneal injury

The cornea copes well with minor injuries or abrasions. If it is scratched, healthy cells slide over quickly and patch the injury before infection occurs and vision is affected. If the scratch penetrates the cornea more deeply, however, the healing process will take longer, at times resulting in greater pain, blurred vision, tearing, redness, and extreme sensitivity to light. Deeper scratches can also cause corneal scarring, resulting in a haze on the cornea that can greatly impair vision. See also separate Corneal Foreign Bodies, Injuries and Abrasions, Eye Injuries, Contact Lenses (Types and Care) and Contact Lens Problems articles.

Allergies

Allergies affecting the eye may affect the cornea, with pollen as the most common allergen. Other allergens include medications, animal hair and cosmetics, such as mascara and face creams. Touching or rubbing eyes after handling chemicals or soaps may also cause an allergic reaction. Treatment is to eliminate or minimise exposure to the allergen, where possible. See separate Allergic Conjunctivitis article.

Infections

Corneal Infection is referred to as keratitis. Trauma and foreign bodies may facilitate the entry of infectious material to the cornea. Contaminated contact lenses are another possible source and corneal infections are the most serious complication of contact lens wear. These infections can reduce visual clarity, produce corneal discharge and, in some cases, erode the cornea or lead to corneal scarring. As a general rule, the deeper the corneal infection, the more severe the symptoms and complications.
**Bacterial keratitis**

**Bacterial keratitis** is infection of one or more layers of the cornea. Most bacteria only produce keratitis once the integrity of epithelium is compromised, such as following corneal abrasion or prolonged contact lens wear. *Neisseria gonorrhoeae* and *Haemophilus influenzae* are the exception, being able to cross intact epithelium.

- **Risk factors** include:[2]
  - Extrinsic - eg, contact lens wear (especially prolonged or associated with poor hygiene), corneal trauma (accidental or surgical) and drug-related (eg, contaminated medication, prolonged steroid therapy and some glaucoma medication).
  - Ocular surface disease - eg, poor tear film.
  - Corneal epithelial abnormalities - eg, neurotrophic keratopathy, viral keratitis.
  - Systemic disease - eg, diabetes, debilitating disease, hypovitaminosis A.

- **Presentation** - redness, pain, photophobia, foreign body sensation and reduced visual acuity. There will usually be an epithelial defect ± the presence of white cell infiltrate ± oedema.

- **Management** - refer:
  - Patients will need intensive topical antibiotic treatment (often after microbiology cultures) ± cycloplegics.
  - Topical steroids may be added during the healing stage.[3] A balance has to be struck between reducing corneal scarring through inflammatory suppression and the possible adverse effects (including prolonging the infection, raising the intraocular pressure and inhibiting collagen synthesis).[2] For this reason, steroids should only be given under specialist supervision.
  - Patients with severe infection or in whom treatment compliance may be poor are admitted. A few others (eg, this is their only functioning eye) may also be admitted.
  - Patients should discontinue any contact lens wear and bring their contact lens equipment (lenses, storage box and cleaning solution) with them for microbiological assessment.

**Viral keratitis**[4, 5]

The most common pathogens are the herpes simplex virus (HSV), causing herpes simplex eye infections, and the varicella-zoster virus (VZV), causing herpes zoster ophthalmicus. These can cause damage at all layers of the cornea and the surrounding structures, either through direct viral invasion or as a result of secondary inflammation.

- **Presentation**:
  - HSV: primary infection is very mild and usually occurs in early childhood, usually characterised by an upper respiratory tract infection and slight rash. Secondary infection varies from superficial dendritic ulcers to deep stromal involvement. Patients present with typical features of corneal problems (see above). There is often decreased corneal sensation. Triggers for viral reactivation include ultraviolet (UV) light, trauma, cold, menstruation and psychological stress.
  - VZV: years to decades after the primary varicella infection there is an influenza-type illness, neuralgia, and macular-papular rash over the distribution of the ophthalmic branch of the trigeminal nerve. Keratitis develops in about 65% of these patients. Precipitating factors include physical trauma, surgery, immunosuppression and systemic illness.

- **Management** - referral in both cases, as the degree of corneal involvement needs to be assessed to determine the need for (usually) topical antivirals ± cycloplegia (HSV), systemic antivirals (VZV) and topical steroids, and to monitor for complications (such as necrosis, ulceration/perforation, scarring). Immunosuppressive drugs such as ciclosporin A are new alternatives to corticosteroid use in select HSV patients.[6] Most patients also benefit from lubricants. Whilst it is best practice to refer the patient with suspected HSV, in a well-established case of recurrent infections, treatment may exceptionally be started in the community, where there is an agreed written plan with the local specialist.
Fungal keratitis (keratomycosis)[7, 8]
A rare but potentially devastating infection most commonly caused by Aspergillus and Fusarium species and typically seen in agricultural settings or where an injury has occurred involving organic matter such as wood or plants. Candidal keratitis is sometimes seen in patients with AIDS. Immunosuppressed patients and those with pre-existing corneal disease are also at risk.

- **Presentation** - similar to bacterial keratitis but onset is gradual and less severe. It should be suspected following non-response to treatment of 'bacterial' keratitis.
- **Management** - refer. The cornea is scraped and topical antymycotic therapy initiated (although doubt has been cast as to the effectiveness of current available therapies). Treatment may last many weeks: unresponsive cases may require systemic treatment or therapeutic penetrating keratoplasty (corneal transplant).

Protozoal keratitis[9]
Acanthamoeba species can cause a devastating, sight-threatening infection. It is a ubiquitous free-living protozoan found in air, water (fresh, salty and tap and in swimming pools and hot tubs) as well as dust, soil or sewage. It survives freezing to boiling temperatures and the chlorination of swimming pools. Contact lens wearers are at greater risk.

- **Presentation** - ranges from asymptomatic to a foreign body sensation, reduced visual acuity and extreme pain (disproportionate to mild clinical findings). Punctate or dendritiform defects may be present with small, white satellite lesions.
- **Management** - refer. Topical amoebicides are used in association with topical steroids. In severe cases, a therapeutic penetrating keratoplasty may be needed to preserve the globe.

Other infectious keratitis

- **Luetic interstitial keratitis** - stromal inflammation associated with syphilis infection.
- **Microsporidial keratitis** - bilateral diffuse keratitis or unilateral deep keratitis seen in the immunocompromised.
- **Infectious crystalline keratopathy** - a rare indolent infection associated with HSV, acanthamoebic keratitis, Streptococcus viridans and long-term topical steroid therapy.

Acute problems following keratoplasty[3, 10]
Corneal grafting (keratoplasty) is a common and successful procedure. It can be performed as an elective procedure to improve vision, or as an emergency in the case of corneal perforation.

- Early postoperative complications include:
  - Wound leak.
  - Raised intraocular pressure.
  - Persistent epithelial defect (>2 weeks).
  - Endophthalmitis.
  - Graft failure.
  - Graft rejection.
  - Urrets-Zavalia syndrome (iris ischaemia).

- **Presentation** - patients who have undergone a keratoplasty and present with the symptoms outlined above should be assumed to have one of the above complications until assumed otherwise. Corneal graft rejection most often occurs within the first two years following the procedure.
- **Management** - refer, ideally to the operating surgeon.

Non-acute corneal problems

Dry eye
Dry eye results when the eye produces fewer or poorer-quality tears and is unable to keep its surface lubricated and comfortable.
The tear film consists of three layers:

- An outer lipid layer that slows evaporation.
- A middle (aqueous) layer that nourishes the cornea and conjunctiva.
- A bottom (mucin) layer that helps to spread the film across the eye.

Tear production and quality reduce with age. Dry eye is more common in postmenopausal women. It is associated with some connective tissue diseases. Other risk factors include dry climates and some medications. See separate Dry Eyes, Rheumatoid Arthritis and Sjögren’s Syndrome articles. Artificial tears, which lubricate the eye, are the principal treatment and are available over the counter. Lubricating ointments are sometimes used at night to help prevent the eye from drying. Humidifiers, protective glasses when outside and avoiding windy and dry conditions may be helpful.

Congenital problems\textsuperscript{[11, 12]}
Babies with suspected corneal abnormalities should be referred urgently, even if they appear otherwise well.

Abnormalities of size

- **Megalocornea** - the cornea is too large - this is an uncommon, bilateral and non-progressive condition which is usually X-linked recessive. It is associated with myopia, astigmatism, cataracts and, later on in life, lens dislocation and glaucoma. It may be associated with Marfan’s syndrome, Apert’s syndrome, Ehlers-Danlos syndrome, Down’s syndrome and osteogenesis imperfecta.
- **Microcornea** - may be unilateral or bilateral and the rest of the eye may be normal (although there are reports of associations with optic nerve hypoplasia, scleroderma, cataract formation, iris abnormalities and secondary angle-closure glaucoma). It may be associated with fetal alcohol syndrome, Turner syndrome, Ehlers-Danlos syndrome, Weill-Marchesani syndrome, Waardenburg’s syndrome, Nance-Horan syndrome and Cornelia de Lange’s syndrome.

Abnormalities of shape

- **Cornea plana** - is a flat cornea: this rare bilateral condition shows autosomal dominant and recessive patterns of inheritance and is associated with peripheral sclerocornea, severe refractive errors, cataracts and colobomata.
- **Keratoglobus** - is an abnormally thin, round cornea. It is one of the corneal ectasias (see ‘Corneal ectasias’, below) and may be associated with Ehlers-Danlos syndrome type IV.\textsuperscript{[10]}

Corneal opacities
The cornea may be cloudy at birth for a number of reasons: these babies should be referred for urgent ophthalmological opinion. The opacity may be:

- **Diffuse** - caused by congenital glaucoma, birth trauma, fetal alcohol syndrome and, rarely, other causes (small print).
- **Focal and central** - caused by birth trauma, Peter’s anomaly (corneal dysgenesis) or neonatal keratitis.
- **Focal and peripheral** - caused by scleroderma (opacification and vascularisation of the cornea), presence of a limbal dermoid or neonatal keratitis.

Disorders of the periphery of the cornea

Marginal keratitis\textsuperscript{[13]}
This is caused by hypersensitivity to staphylococcal toxins, more common in patients with chronic staphylococcal keratitis or blepharitis. It is characterised by peripheral infiltrates and multiple epithelial defects which eventually coalesce. It is a recurring condition.

- **Presentation** - typically, the patient is familiar with their symptoms of mild irritation and discomfort associated with a red, watery eye. Occasionally, discomfort is severe.
- **Management** - refer for confirmation of diagnosis and a short course of topical steroids.
Rosacea keratitis
This occurs in patients with acne rosacea. The severity of the ocular condition does not correlate with that of the skin condition. Like acne rosacea it is more common in middle-aged, fair-skinned females.

- **Presentation** - nonspecific irritation, burning and redness associated with inferior punctate epithelial defects and peripheral neovascularisation. There may be lid and conjunctival involvement.
- **Management** - refer for topical steroids and a course of systemic antibiotics (eg, doxycycline 100 mg once daily for twelve weeks). Concurrent blepharitis also needs addressing. In very severe cases where there is the threat of corneal perforation, systemic immunosuppression is used.

Ulcerative keratitis in systemic disease
This is particularly associated with rheumatoid arthritis where there is severe, progressive corneal thinning (perforation may occur). It also occurs in other conditions such as granulomatosis with polyangiitis (Wegener's granulomatosis), systemic lupus erythematosus, relapsing polychondritis and polyarteritis nodosa. It may be referred to as 'peripheral ulcerative keratitis' (PUK).

- **Presentation** - acute unilateral/bilateral exacerbations characterised by decreased visual acuity, with variable pain and redness (there may be none).
- **Management** - treatment involves systemic immunosuppression, topical immunosuppression, ocular lubricants and globe protection (such as an eye shield). Both ophthalmologists and rheumatologists are involved.

Pterygium
A pterygium is a pinkish, triangular-shaped growth of tissue which encroaches on to the cornea. They rarely grow over the pupil. Pterygia are more common in sunny climates and in the 20-40 age group; they are thought to relate to environmental 'wear' to the eye surface, including from dry air, dust and solar radiation. If surgically removed they may grow back, particularly if the patient is less than 40 years of age. Lubricants reduce the redness and provide relief from the chronic irritation.

Other disorders
- **Mooren's ulcer** - this is an ulcerative condition (usually unilateral) which arises as a result of an autoimmune response to corneal antigens. It is rare but serious, particularly in young Africans in whom an aggressive form is seen. Treatment depends on the subtype (ranges from topical steroids/antibiotics to aggressive systemic steroid treatment).
- **Dellen** - localised saucer-shaped thinning of the cornea caused by localised tear film instability. Managed with lubricants; it is a transient condition.
- **Phlyctenulosis** - small pinkish-white nodule with an associated red eye. It occurs as a result of a nonspecific delayed hypersensitivity reaction to bacterial and viral antigens. It may resolve spontaneously or be treated with a short course of antibiotics or topical steroids.
- **Terrien's marginal degeneration** - uncommon, idiopathic bilateral thinning of the cornea, usually occurring in the third to fourth decade of life, more often in males. There may be pain; there is decrease in visual acuity. Surgery may be needed to excise the diseased tissue.

Degenerative conditions

Age-related degenerations
- **Arcus senilis** (sometimes referred to as corneal annulus or anterior embryotoxon) - this is the most common peripheral corneal opacity, which may occur alone or in association with hyperlipidaemia (especially if present in younger individuals). It is caused by lipid droplets in the corneal stroma. Rarely, it is unilateral, in which case it is associated with carotid disease or ocular hypotony.
- **Vogt's limbal girdle** - a common, innocuous age-related finding characterised by peripheral chalky-white crescents at the 3 o'clock and 9 o'clock positions.
- **Cornea guttata** - an innocuous change in endothelial cells which can occasionally be a precursor of early Fuchs' endothelial dystrophy (see 'Dystrophic conditions', below).
Lipid keratopathy
This involves deposits of lipid within the cornea, which may be idiopathic or associated with previous keratitis or disordered lipid metabolism. There are two types, one of which is associated with corneal vascularisation if left untreated. It appears as a bright white, well-defined patch on the cornea, sometimes associated with a ‘feeding’ vessel. It requires laser or surgical removal.

Band keratopathy
This is the deposition of calcium salts within the cornea, which looks like a grey band of opacitiy crossing the cornea horizontally. It is most commonly found in chronic uveitis but also with a number of other causes - eg, prolonged glaucoma, long-standing corneal oedema and corneal dystrophies. It can also arise in the context of systemic disease such as hypercalcaemia, hyperuricaemia and in chronic kidney disease. Chelation is the treatment of choice: sodium edetate is applied until all the calcium is removed. Ultimately, the underlying condition needs to be addressed.

Other degenerative conditions

- **Spheroidal degeneration** - a bilateral condition of unknown cause, mostly occurring in men working outdoors. Small golden-brown lesions accumulate in the cornea, associated with generalised haziness. UV protection helps but patients may need surgical removal of the lesions.
- **Salzmann’s nodular degeneration** - discrete grey opacities arise in the cornea, secondary to chronic keratitis (especially trachoma). They may be associated with a red eye, irritation and blurred vision. The treatment is as for spheroidal degeneration.
- **Crocodile shagreen** - this describes a faint network of stromal opacities resembling crocodile skin. It is innocuous.

Dystrophic conditions

These are a group of progressive, usually bilateral, conditions which affect one of the various layers of the cornea, leading to a loss of normal clarity due to a build-up of cloudy material. There are over 20 corneal dystrophies that affect all parts of the cornea. They are usually inherited, bilateral and gradual in progression. They generally begin in one layer and then spread to affect others. Some cause severe visual impairment, while a few cause no vision problems. Some cause repeated episodes of pain without permanent loss of vision. Examples include:

- **Fuchs’ endothelial dystrophy** - an autosomal dominant inherited condition which usually appears in the sixth decade. Fuchs’ dystrophy begins with deterioration in the endothelial layer. Eventually the endothelial cell loss affects the layer’s function, leading to swelling of the cornea and impairment of vision. Swelling also causes painful blisters on the epithelial layer. Corneal transplant may be needed to restore vision.
- **Lattice dystrophy** - involves an accumulation of amyloid deposits in the stroma. These gradually cloud the cornea and may accumulate under the epithelium, causing recurrent painful epithelial erosion. The condition usually arises in children between the ages of two and seven.
- **Map-dot-fingerprint dystrophy** - an abnormality of the basement membrane of the epithelium, so that epithelial cells cannot properly adhere to it. This causes recurrent epithelial erosions, periodic blurred vision and severe pain. It tends to occur in both eyes and usually affects adults between the ages of 40 and 70. Typically, map-dot-fingerprint dystrophy will flare up occasionally for a few years and then go away on its own.

Corneal ectasias

**Keratoconus**
This is the most common primary corneal ectasia, a bilateral, progressive conical distortion of the cornea, starting in puberty. The aetiology is not clear but repeated trauma (eg, eye rubbing) and connective tissue disorders may contribute.

- **Presentation** - it can occur in the second to third decade of life, with progressive visual impairment and occasional sudden transient corneal oedema as the weakened Descemet's membrane of the cornea cracks. The changes are nearly always bilateral.
- **Management** - it is treated with spectacles initially, then contact lenses. Ultimately, patients benefit from keratoplasty. Acute oedema is managed with topical hypertonic sodium chloride and topical atropine; it may take weeks to months to resolve.
Pellucid marginal degeneration
This condition is similar to keratoconus but occurs later in life (second to fourth decade). Treatment is the same.

Keratoglobus\textsuperscript{[17]}
See description under ‘Abnormalities of shape’, above; it may also be acquired (probably as an end-stage keratoconus). It arises as a result of thinning of the stromal layer of the cornea. Treatment may include protection from trauma, specialised contact lens wear or surgery.

Iridocorneal endothelial (ICE) syndrome\textsuperscript{[18]}
This unilateral syndrome typically affects women aged 30-50. Endothelial cells migrate from the cornea to the iris and, in doing so, obstruct the angle and cause both glaucoma and corneal swelling and cloudiness which may eventually require transplant. The cause is unknown. The three main features are corneal oedema, iris atrophy and secondary angle-closure glaucoma. ICE syndrome can result in substantial visual impairment both from corneal oedema and from neuropathy secondary to glaucoma.

Neurokeratopathies

Exposure keratopathy\textsuperscript{[19]}
This is caused by damage to the cornea as a result of improper tear cover/wetting of its surface. The tear film may be reduced or normal but the blink rate is reduced (facial nerve palsy, severe proptosis, eyelid scarring).

- **Presentation** - a progressively painful red eye. Look for loss of shiny reflection from the corneal surface.
- **Management** - if recovery of the underlying problem is anticipated, aggressive lubrication is sufficient. Otherwise, tarsorrhaphy (partial suturing together of the lids) may be effective.

Neurotrophic keratopathy
This occurs when there is loss of sensation in the cornea (eg, acoustic neuroma, diabetes, HSV affecting the fifth cranial nerve) followed by secondary intracellular oedema (the pathogenesis is unknown).

- **Presentation** - variable: a painless red eye with mild visual impairment secondary to corneal oedema through to epithelial defects leading to corneal ulceration. A decrease in corneal sensation is the key factor.
- **Management** - this depends on the severity but lubrication ± protection overnight will usually be enough.

Miscellaneous conditions

Astigmatism
In this condition, the corneal shape is slightly rugby ball-shaped rather than truly spherical, causing a refractive error. It is most commonly treated with corrective spectacles and contact lenses (the latter may be the best way of improving sight). Surgery is also an option.

Drug-induced keratopathies
The cornea can be affected by a number of systemically administered drugs, including gold (causing chrysis - deposition of gold deposits), antimalarials and amiodarone - both of which give rise to vortex keratopathy characterised by whorl-like corneal deposits. Vortex keratopathy has also been associated with indometacin, tamoxifen and clofazimine. Drug-induced keratopathy has been described in the use of some oriental herbal medicines.\textsuperscript{[20]}

Thygeson's superficial punctate keratopathy\textsuperscript{[3]}
This rare, idiopathic condition usually arises in the younger population (under 24 years). It is characterised by recurrent episodes of pain and foreign body sensation (± blurred vision, red eye, photophobia and tearing) and crumb-like, non-staining white opacities scattered over the corneal surface. It is usually bilateral but asymmetrical. It is managed with topical steroids. Although visual prognosis is good, patients have to be monitored for complications of treatment.
**Metabolic keratopathies**

- **Cystinosis** - ocular features include progressive deposition of cystine crystals causing photophobia, blepharospasm, epithelial erosions and reduced visual acuity. Later the iris, lens and retina are also involved.
- **Immunoprotein deposits** - eg, multiple myeloma, Waldenström's macroglobulinaemia, monoclonal gammopathy. Uncommonly, these cause bilateral corneal deposits which, when severe, may require penetrating keratoplasty.
- **Mucopolysaccharidoses** - corneal deposits typically (except Hunter’s syndrome and Sanfilippo syndrome) and tend to be most severe in Hurler’s syndrome. The retina and optic nerve may also be affected.
- **Wilson’s disease** - the Kayser-Fleischer ring occurs when copper is deposited around the periphery of the cornea. It may only be visible by gonioscopy, where it may be found to be variable in colour (eg, brown, red, green or yellow).

**Further help & information**

**Keratoconus Group**

**Further reading & references**

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