KERATOCONUS

Condition is bilateral, often asymmetric. Progression typically occurs until 40 years of age. Non-inflammatory, although inflammatory mediators can hasten progression.

**Age of onset:** Puberty.

**Typical clinical findings:** Myopic astigmatism, scissor reflex on retinoscopy and/or distorted mires on keratometry.

**Established associations:** Knuckle rubbing, eczema, asthma, allergy and positive family history.

**Associated systemic conditions:** Down syndrome, Ehlers-Danlos syndrome, Marfan’s syndrome, osteogenesis imperfecta, Leber’s congenital amaurosis and floppy eyelid syndrome. There is also a possible association with mitral valve prolapse.

### Sagittal front corneal topography

#### Subclinical Keratoconus

- Typically refers to an “incomplete form” of keratoconus
- The term can describe either of the following:
  1. Where the contralateral eye has keratoconus and the eye in question has:
     - Normal corneal topography
     - OCT findings are indistinguishable from a “normal” cornea
  2. Some suspicious corneal topographical changes with no other clinical findings conclusive for a diagnosis of keratoconus
- The terms “keratoconus suspect” and “Forme Fruste keratopathy” have also been used to describe this in the literature

#### Early Keratoconus

- Subtle signs of corneal thinning (yellow arrow).
- Subtle signs of epithelial thinning overlying the cone.

**Topography:**
- Abnormal elevation (steepening) of the anterior and posterior surfaces often resulting in an asymmetric bow-tie appearance and increase in astigmatism.
- Posterior changes can occur prior to anterior changes

**Pachymetry:**
- Abnormal corneal thickness distribution
- Apical corneal thinning - typically presents inferior to the visual axis but can be present in other corneal locations.

#### Moderate to advanced keratoconus

- Vogt’s striae
- Fleischer’s ring
- Perforation or hydrops can occur
- Corneal scarring at the conical apex

**OCT findings:**
- Atypical corneal thinning and forward bowing
- Abnormal epithelial distribution (Yellow arrow)

**Topography:**
- Marked corneal elevation and steepening

**Pachymetry:**
- Prominent corneal thinning

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CHAIR-SIDE REFERENCE: CORNEAL ECTASIA

This reference is based on the current literature and evidence at the time of writing. This reference is designed a guide to aid diagnosis and management decisions however individual cases must be assessed in the context of all available clinical data.
**PELLUCID MARGINAL DEGENERATION**

Condition is bilateral and non-inflammatory. Male predominance. Presents with increasing against-the-rule astigmatism. Age of onset: 20-50 years. There is currently debate as to whether pellucid marginal degeneration, keratoconus and keratoglobus are separate entities or different phenotypes of the same condition.

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<th>Sagittal front corneal topography</th>
<th>Anterior OCT and/or colour photography</th>
<th>Description</th>
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| Early                             |                                      | Spectacle visual acuity typically normal. May be difficult to distinguish from keratoconus without multimodal imaging. Slit lamp and OCT findings:  
  - A 1-2mm thick band of peripheral corneal thinning extending from 4 o’clock to 8 o’clock  
  - Cornea is clear at the area of thinning (yellow arrow)  
  Topography:  
  - Inferior corneal thinning and steepening  
  - Superior corneal flattening along the vertical meridian  
  - “Crab claw” or “kissing dove” pattern  
  Pachymetry:  
  - Possible inferior thinning however typically minimal change in central corneal thickness |
| Advanced                          |                                      | Spectacle visual acuity is typically reduced but can remain normal with large cylindrical corrections. Slit lamp findings:  
  - Protrusion of the inferior cornea above the area of thinning  
  - Inferior peri-limbal striae  
OCT findings:  
  - Peripheral stromal thinning (yellow arrow)  
  - Epithelial changes overlying the area of thinning  
Topography:  
  - Marked “Crab claw” pattern, Sometimes better appreciated on a Tangential map  
  Pachymetry:  
  - Perforation or hydrops can occur |
| TERRIEN’S MARGINAL DEGENERATION  |                                      | Males are affected more than females. May present in 2 forms:  
  1. Quiescent type (affects older individuals, asymptomatic in early stages)  
  2. Inflammatory type (affects younger individuals 20-30 years of age, associated with episcleritis or scleritis). |
| KERATOGLOBUS                      |                                      | Rare bilateral condition usually present at birth. Some associations with connective tissue disorders such as Ehlers-Danlos syndrome, Marfan syndrome and Rubenstein-Taybi Syndrome  
Clinical signs:  
  - Generalised thinning of the cornea, especially in the periphery  
  - Pachymetry is reduced to up to one fifth of normal corneal thickness  
  - Globular protrusion of the cornea  
  - High myopia with irregular astigmatism  
  - Perforation or rupture of cornea  
Keratoglobus can occur in older patients following a hydrops in those with keratoconus or pellucid marginal degeneration. |