Chair-side Reference: Corneal ectatic diseases and thinning disorders

Corneal ectatic and thinning disorders can be sight threatening. The addition of imaging technologies, such as corneal topography and/or optical coherence tomography (OCT), enable earlier diagnosis and an improved ability to monitor progression. Grading scales exist for more common ectatic diseases such as keratoconus; however there has not been a consensus on a grading scale.

<table>
<thead>
<tr>
<th>Keratoconus</th>
<th>Sagittal front corneal topography</th>
<th>Anterior OCT and/or colour photography</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age of onset: puberty. Progression typically occurs until 40 years of age. Bilateral, often asymmetric. Non-inflammatory, can be associated with systemic conditions including Down syndrome, Ehlers-Danlos syndrome, and osteogenesis imperfecta. There is also an association with Atopy and ‘knuckle-rubbing’</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Forme Früste**

- 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 term “keratoconus suspect” is also used
- In earlier research, this was also referred to as Forme Früste keratopathy

**Early**

- Slit lamp and OCT findings:
  - Subtle signs of corneal thinning
- Topography:
  - Abnormal elevation (steepening) of the anterior and posterior surfaces often resulting in an asymmetric bow-tie appearance
  - Posterior changes can occur prior to anterior
  - Typically results in an increase in astigmatism
- Pachymetry:
  - Abnormal corneal thickness distribution
  - Apical corneal thinning
  - Thinning typically presents inferior to the visual axis although can be present in other corneal locations

**Moderate to advanced keratoconus**

- Slit lamp findings:
  - Vogt’s striae (identified by arrow in corneal photo)
  - Fleischer’s ring
  - Corneal scarring at the conical apex (shown by arrow on OCT)
- OCT findings:
  - Atypical corneal thinning and forward bowing
  - Abnormal epithelial distribution
- Topography:
  - Marked corneal elevation and steepening
- Pachymetry
  - Prominent corneal thinning
  - Perforation or hydrops can occur
<table>
<thead>
<tr>
<th>Pellucid Marginal Degeneration</th>
<th>Age of onset: 20-50 years, Bilateral, non-inflammatory condition, Male preponderance, Presents with increasing against-the-rule astigmatism</th>
</tr>
</thead>
</table>
| **Early**                      | Spectacle visual acuity typically normal  
|                                |  • Difficult to distinguish from keratoconus  
|                                | 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” 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** | Usually bilateral and symmetric, but may be asymmetric. Males affected more than females. Can present in two forms:  
|                                | Quiescent type  
|                                |  • Affects older individuals  
|                                |  • Asymptomatic during early stages  
|                                | Inflammatory type  
|                                |  • Affects younger individuals between 20-30 years  
|                                |  • Can be associated with episcleritis or scleritis  
|                                | OCT findings:  
|                                |  • Thining of the superior peripheral cornea (peripheral gutter)  
|                                | Slit lamp findings:  
|                                |  • Yellow-white punctate opacifications in the stroma (earlier)  
|                                |  • Lipid deposition along the anterior edge of the limbus (later)  
|                                |  • Pseudo-pterigia (20% of cases)  
|                                | Topography:  
|                                |  • Against-the-rule astigmatism  
|                                |  • Steepening of the superior peripheral cornea  
| **Keratoglobus** (Imaging not available) |  • Age of onset: Usually at birth  
|                                |  • Rare bilateral condition  
|                                |  • Some association with connective tissue disorders such as Ehlers-Danlos syndrome, Marfan syndrome and Rubenstein-Taybi Syndrome  
|                                |  • Can occur in older patients following a hydrops in those with keratoconus or PMD  
| **Signs include:** |  • Generalised thinning of the cornea, especially in the periphery  
|                                |  • Pachymetry 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  |