Centre for Eye Health

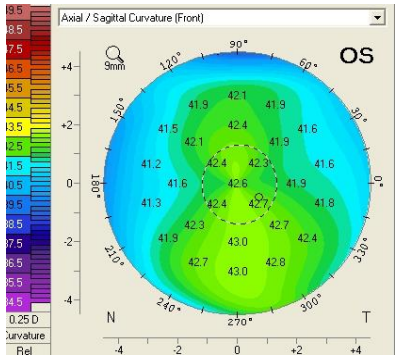
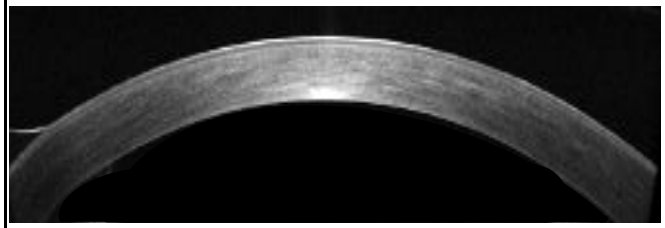
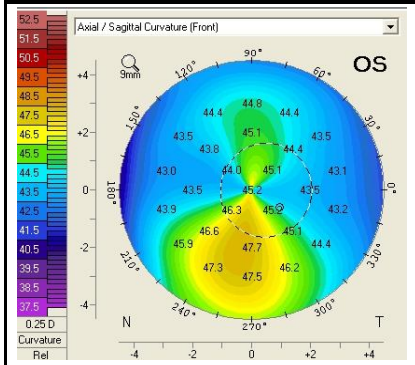
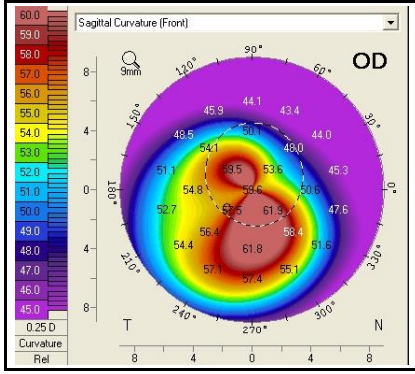
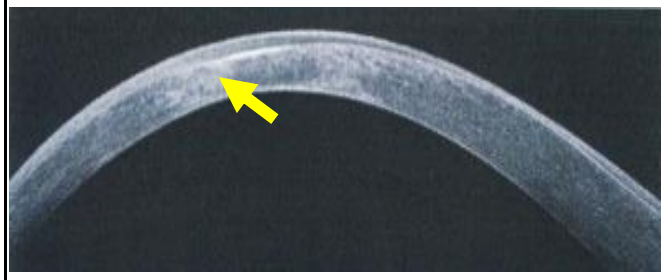
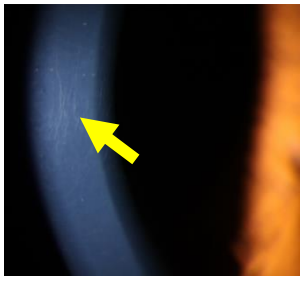
Chair-side Reference: Corneal ectatic diseases and thinning disorders

Optometry NEW SOUTH WALES AUSTRALIAN CAPITAL TERRITORY

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.

Keratoconus

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'

Sagittal front corneal topography	Anterior OCT and/or colour photography	Description
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 

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Optometry NEW SOUTH WALES AUSTRALIAN CAPITAL TERRITORY

Pellucid Marginal Degeneration

Age of onset: 20-50 years, Bilateral, non-inflammatory condition, Male preponderance, Presents with increasing against-the-rule astigmatism

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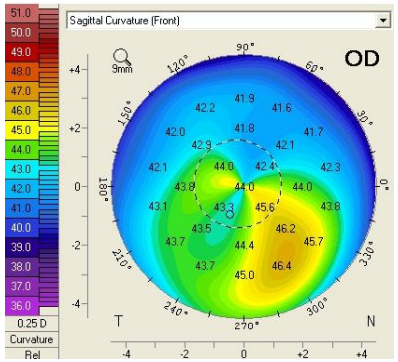
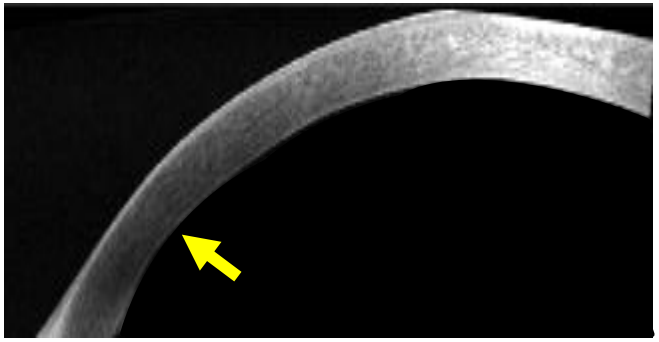
Early





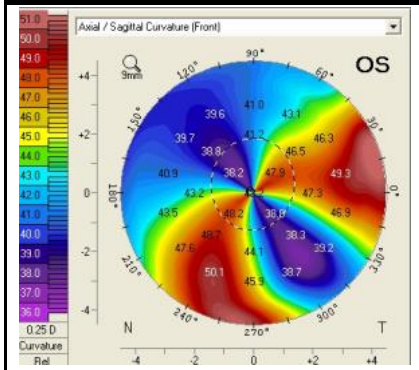
Spectacle visual acuity typically normal
- Difficult to distinguish from keratoconus

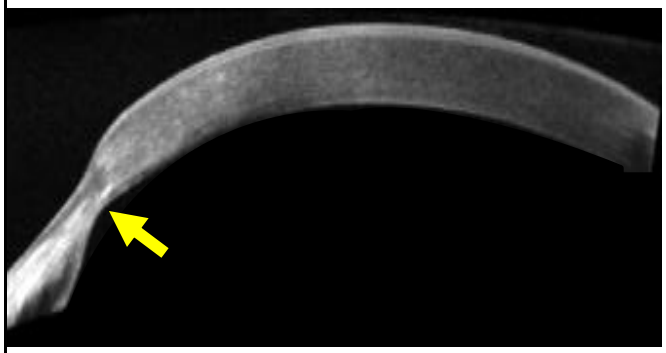
Slit lamp and OCT findings:
- A 1-2mm thick band of peripheral corneal thinning extending from 4 o'clock to 8 o'clock
- Cornea is clear at the area of thinning (yellow arrow)

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

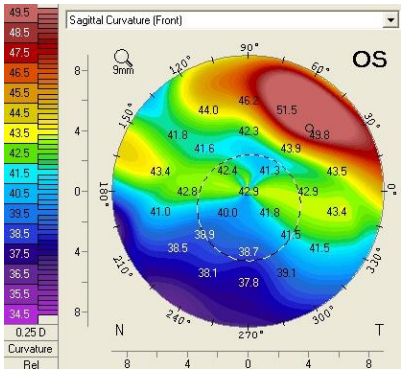
Slit lamp findings:
- Protrusion of the inferior cornea above the area of thinning
- Inferior peri-limbal striae

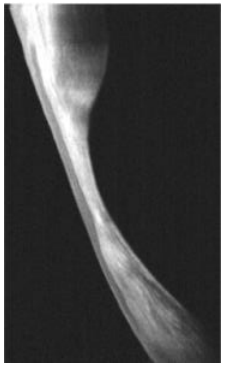
OCT findings:
- Peripheral stromal thinning (yellow arrow)
- Epithelial changes overlying the area of thinning

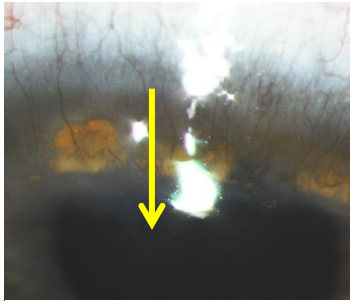
Topography:
- Marked "Crab claw" pattern
  Sometimes better appreicated on a Tangential map

Pachymetry
- Perforation or hydrops can occur

Terrien's marginal degeneration







OCT corresponds to the line shown on anterior eye image.

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:
- Thinning 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-pterygia (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