



Keratoconus: The good, bad, and the ugly

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Stats

- Prevalence:
 - 54.5 per 100,000
- Incidence:
 - 1 per 2,000 in the general population (some studies say 1 in 500)
 - Much higher among Asians (4-1) than the Caucasian population

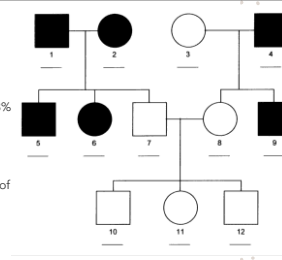
The severity of the disease also varies with race and was reported to be 4.4 and 7.5 times greater among Asians compared to Caucasians

Some studies have found it is more prevalent in men than women

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Heritage


- The hereditary pattern is not prominent or predictable
- Positive family histories have been reported in 6-8%
- Twin studies and pedigrees have demonstrated autosomal dominant transmission of the disease
- A majority of keratoconus cases occur as a result of the genetic predisposition triggered by environmental factors



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Genetic Testing



- AvaGen - The Genetic Eye Test
 - Gene sequencing for 75 keratoconus-related genes and over 2,000 variants
 - Utilizes next generation sequencing
 - Creates a polygenic risk score

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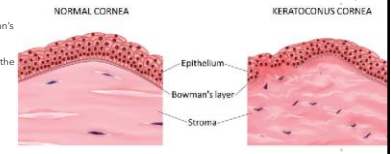
Associations

- Risk factors: Eye rubbing
- Leber's congenital amaurosis (oculodigital reflex)
- Vernal keratoconjunctivitis
- Atopic keratoconjunctivitis
- Floppy eyelid syndrome
- Contact lens wear
- Systemic associations:
 - Osteogenesis imperfecta
 - Down syndrome
 - Mitral valve prolapse
 - Marfan syndrome

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Pathophysiology

- Early biomicroscopic and histologic findings include fibrillations of Bowman's layer, leading to breaks followed by fibrous growth and dysplasia through the breaks.
- The cause of keratoconus is unknown however, it has been suggested that disturbances in protease regulation, wound healing, or IL-1-mediated apoptosis might be involved.



Semantic Scholar

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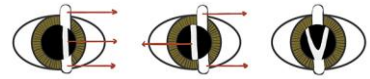
Clinical Signs

- Nearly all cases are bilateral but it is usually asymmetric
- Apical thinning of the central cornea progressively worsens and may cause extreme degrees of irregular astigmatism.

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Retinoscopy

Scissoring of the red reflex on ophthalmoscopy or retinoscopy is a very early sign of keratoconus

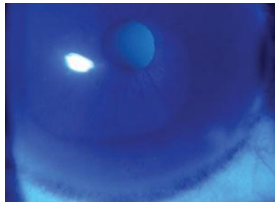
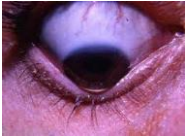


Mivision Education

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Slit Lamp Findings

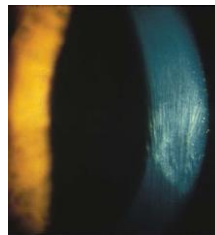
- Munson sign is a protrusion of the lower eyelid in downward gaze.



- Fleischer ring is an iron line present within the epithelium and located around the base of the cone.

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Slit Lamp Findings



- Vogt lines - Fine parallel rough striations of the posterior stroma, also called stress lines.

- Hydrops is a result of a tear in Descemet's membrane causing the sudden development of corneal edema.

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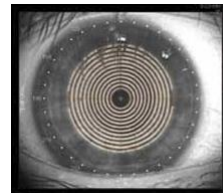
Hydrops Management

- Epithelial defect? - Moxifloxacin
- IOP elevation? - Timolol
- Corneal edema - Muro ointment, prednisolone acetate
- SF6 can be used in some instances to help reposition Descemet's membrane from the break

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Topography

- Placido disc based is the most commonly available form of topography
- Placido discs projected onto cornea, mires reflected from the tear film



Oculus Keratograp

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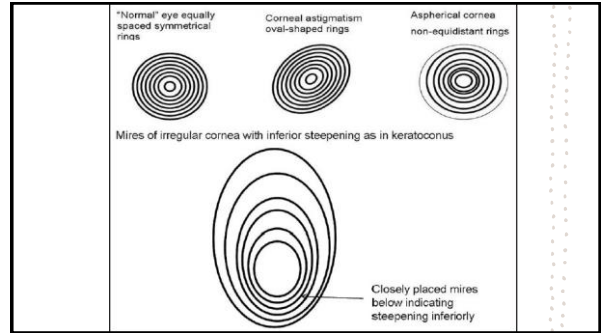
Images formed by Placido Disc

- Based on the overlay of concentric mires on the cornea.
 - The closer the mires, the steeper the axis.
 - The wider the rings, the flatter the axis.



The principles of Placido-disk technology. The larger the distances between the rings, the flatter the cornea, and the less dioptric power it has. The reverse is true with a steeper cornea, which shows smaller distances between the rings and a higher corneal power.

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Tomography

- Topography describes the shape
- Tomography ALSO describes thickness, in addition to anterior and posterior corneal curvature (scanning-slit and scheinplugg)
- This can reveal subtle changes in the posterior corneal curvature that can precede anterior changes
- Useful in distinguishing keratoconus vs pellucid

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Computerized videokeratography

- Computerized videokeratography algorithms are used to diagnose subclinical keratoconus
- Helps rule out refractive candidates who will be at high risk for post-refractive ectasia.

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I-S Ratio

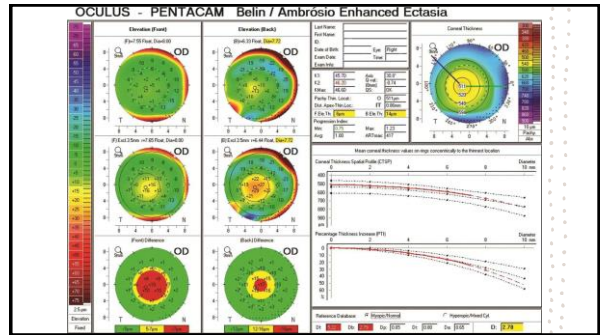
- Inferior - superior (I-S) values are useful for screening for KCN
- Derived by calculating the difference between the inferior and superior corneal curvature measurements at a defined set of 5 points above and below the horizontal meridian
- I-S values >1.4 are suggestive of corneal ectatic disorders

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Belin Ambrosio Enhanced Ectasia Display

- Combines elevation based and pachymetric corneal evaluation in an all inclusive display
- The difference between the standard and enhanced best-fit-spheres indicates a change in elevation, which is useful to identify ectatic protrusion on the corneal surface
- Analyzes deviation of normality for the front (df) and back (db), pachymetric distribution (dp) and also vertical displacement of pachymetry in relation to the apex (dy).
- A final "D" is calculated based on a regression analysis that weights differently each parameter
- Yellow = Suspicious = ≥ 1.6 SD from the mean
- Red = Abnormal = ≥ 2.6 SD from the mean

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Definition of Progression

Which of the following is **NOT** part of the criteria for progression?

- Increase in cyl by greater than or equal to 1D
- Increase in sphere greater than or equal to 0.25D
- Increase in spherical equivalent by greater than or equal to 0.5D
- Increase in steepest K by 1D



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- Increase in steep K by 1D



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Treatment

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Corneal Collagen Cross-Linking (CXL)

- A minimally invasive technique primarily used to halt further ectasia.
- The technique combines riboflavin and UV light to strengthen chemical bonds in the cornea, thereby stiffening a structurally weak and unstable cornea.



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Patient Selection

- Documented progression of keratoconus, iatrogenic post-laser surgery ectasia, and more recently pellucid marginal degeneration
- Thinnest Pachymetry >400um

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Contraindications




- Ocular herpes zoster or herpes simplex
- Autoimmune disorders
- History of poor epithelial healing
- Severe or central corneal scarring

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Surgical Techniques

Epi Off



Epi On

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Epi Off

- Conventional Procedure
- FDA Approved
- Benefits
 - Better riboflavin and UV absorption
 - Deeper and more effective therapy
 - More corneal stiffening and flattening
 - Lower risk of post-treatment ectasia progression

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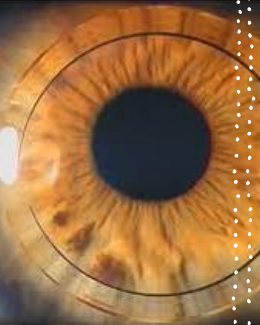
Epi On

- Leaves the corneal epithelium intact and achieves penetration of riboflavin in a variety of manners
- Followed by UVA irradiation with continued intermittent administration of the riboflavin solution during irradiation
- Benefits
 - Faster return to baseline vision and return into contact lenses
 - Lower risk for corneal infections and haze
 - Shorter duration of discomfort

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Intrastromal Corneal Ring Segments (ICRS)

- ICRS have also been approved for the treatment of mild to moderate keratoconus in patients who are contact lens intolerant



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Patient Selection

- A clear central cornea
- Corneal thickness of > 450 microns where the segments are inserted at 7 mm optical zone

Contraindications



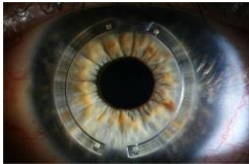
- Ocular herpes zoster or herpes simplex
- Severe or central corneal scarring
- Pachymetry <450um at the proposed incision site

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Intacs

- Hexagonal transverse shape with 8.0 mm external diameter and 6.8 mm internal diameter
- Predicted cylinder range of correction is +2.00 to +4.00 and higher



Ferrara Rings

- Available in European countries and South America
- The cross-section of a Ferrara ring is triangular in shape to reduce the photic phenomena by inducing a prismatic effect
- Corrects up to 7.0D of myopia

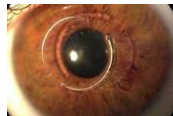


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Kera Rings

- Similar to Ferrara rings but have different options in arc length



Myo Rings

- A 360° continuous full-ring implant for myopia and keratoconus



Deep Anterior Lamellar Keratoplasty (DALK)

- Replacement of the central anterior cornea, leaving the patient's endothelium intact
- Advantages:
 - Risk of endothelial graft rejection is eliminated
 - Less risk of traumatic rupture of the globe in the incision
 - Faster visual rehabilitation
- Disadvantages:
 - Technically more difficult
 - Complications include interface haze, and double anterior chamber

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Penetrating Keratoplasty (PKP)

- Has a high success rate and is the standard surgical treatment with a long track record of safety and efficacy
- Risks of this procedure include infection, rejection, and risk of traumatic rupture at the wound margin
- Many patients after PK may still need hard or gas-permeable contact lenses due to residual irregular astigmatism

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Questions?



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thank you!

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