Keratoconus: Clinical Decision Making and Patient Education

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Disclosures

• None
Objectives

• Review the clinical characteristics of Keratoconus
• Review current contact lens options
• Understand surgical options in Keratoconus
• Review Acute Corneal Hydrops
What is Keratoconus?

- Primary, progressive, clinically non-inflammatory* corneal thinning\(^1\)
  - *Recent studies show increase in inflammatory mediators in tear film and corneal cells\(^2\)
- Bilateral, Asymmetric
- Prevalence 1 in 2000\(^3\)
- Two-hit hypothesis\(^2\)
  - Genetic predisposition
  - Environmental influence
RefRACTive Signs

• Refractive:
  • Varying levels of myopia, astigmatism
  • Scissoring reflex on retinoscopy
  • Can present with multiple glasses remakes in early disease
Corneal Signs

- Apical thinning
- Fleischer ring: subepithelial iron deposition
- Vogt’s Striae: posterior, vertical; stress lines
- Apical Scarring
- Corneal Protrusion: Munson’s Sign
Imaging Signs

• Anterior Topography: Placedo Disc
  • Central/Paracentral steepening
Imaging Signs

- Tomography/Pentacam
  - Abnormal anterior/posterior elevation
Imaging Signs

- Tomography/Pentacam
  - Global pachymetry values
    - Rapid thickness change from center of ectasia to “normal” peripheral corneal thickness
    - Central thickness can be “normal”
Contact Lens Options in Keratoconus
Vision options: Early disease

• Spectacles
  • Minimal to no reduction in BCVA
  • Adverse to CLs
  • ~16% ⁴

• Commercial soft contact lenses
  • Early disease, adequate spectacle BCVA
Contact lens options: Specialty Soft

• Designs specific for Keratoconus
  • Increased center thickness to mask irregularity
  • Aggressive stabilization techniques

• Recent improvements in oxygen permeability may lead to increase in utilization of this modality

• Most successful in early/mild disease\textsuperscript{5}

• Poorer visual acuity and less control of higher order aberrations vs GP\textsuperscript{6}
Contact Lens Options: Hybrid

- Center: Gas permeable
- Periphery/Skirt: Soft
- New lens designs utilize silicone hydrogel skirt to improve oxygen permeability
- Moderately improved lens tolerance compared to corneal GP
Contact Lens Options: Gas Permeable

• Corneal Gas Permeable
  • Historical standard of care
  • Lens designs to match location/size of ectasia

• Scleral Lenses
  • Large diameter gas permeable lens
  • Avoids interaction with cornea
  • Bears weight on conjunctiva/sclera
  • Currently employed in moderate-advanced disease
  • Current area of extensive interest/research/publication
Surgical Options in the Management of Keratoconus
Surgical Options

- Collagen Cross-linking
- Intrastromal Corneal Ring Segments
- Combined PRK and Collagen Cross-linking
- Keratoplasty
Collagen Cross-linking (CXL)

• Ultraviolet activated riboflavin strengthens cornea

○ Primary goal:
  • Decrease/stop progression

○ Secondary goal:
  • Improve BCVA, uncorrected VA, and reduce spherical equivalent Rx
Collagen Cross-linking (CXL)

• Two treatment strategies
  • Epi-Off: original approach
  • Epi-On/ Transepithelial: Developed to address safety concerns
  • Community still undecided
    • Epi-Off: More effective?
      • Epi-Off: 93% stable
      • Epi-On: 40% stable
    • Epi-On: Safer?
      • Epi-Off: 0-3% microbial keratitis, ~10 % stromal haze, 2.5% sterile infiltrates
      • Epi-On: none of the above
Collagen Cross-linking (CXL)

- Supporting literature⁹:
  - 36 month randomized, controlled trial
  - Corneal Curvature: Kmax
    - Control: Steepened by 1.75 D
    - Study: Flattened by 1.03 D
  - BCVA and UCVA improved in study group
  - 2 non-vision affecting complications
Collagen Cross-linking (CXL)

- Patient Selection:
  - Progressive Disease
  - Likely risk of progression
    - Younger age
    - Steep K at diagnosis
    - Poor BCVA at diagnosis
Intrastromal Corneal Ring Segments (ICRS)

• INTACS®
  • Material: PMMA
  • Implanted in peripheral stroma

- Primary Goals:
  • Improve uncorrected and best-corrected VA
  • Improve CL tolerance
- Does not halt progression\textsuperscript{10}
Intrastromal Corneal Ring Segments (ICRS)

• Patient Selection\textsuperscript{11}:
  • Inadequate vision in spectacles and CLs
  • CL intolerance
  • Clear central cornea
  • Adequate mid-peripheral corneal thickness (~450 microns)

• Moderate-Severe most likely to benefit
Combined ICRS and CXL

• Combines refractive management and corneal stabilization\textsuperscript{12}
• Comparable refractive outcome to ICRS alone
• Ideal sequence of treatment undecided
Combined Photorefractive Keratectomy and CXL

- Combines subtractive technique (PRK) for vision correction with stabilizing technique (CXL)
- Same-day PRK followed by CXL has less complications and is more effective than sequential procedure\textsuperscript{13}
- Primary Goals:
  - Stop progression
  - Reduce spherical equivalent to improve BCVA and UCVA
Combined PRK and CXL

- Treatment strategies
  - Topography guided PRK: better regularization of topography\(^\text{14}\)
  - Non-topography guided: less tissue removed\(^\text{15}\)

- Both improve UCVA, decrease SE and decrease progression
Combined PRK and CXL

• Patient Selection
  • Progressive disease
  • Inadequate vision in spectacles and contact lenses
  • Contact lens intolerance
  • Clear cornea
  • Mild-Moderate disease
Keratoplasty$^{16}$

- Penetrating Keratoplasty
  - Historical standard of care
  - Full-thickness corneal transplant
  - Indicated when endothelium is compromised (Hx of Hydrops) or scarring is posterior

- Deep Anterior Lamellar Keratoplasty
  - Anterior, partial thickness corneal transplant
  - Indicated with anterior scarring and in presence of uncompromised Descemet’s membrane/endothelium
PKP vs. DALK\textsuperscript{17}

**TABLE 1. Incidence of Postoperative Complications in Keratoconus Patients Who Had Undergone DALK or PKP**

<table>
<thead>
<tr>
<th></th>
<th>DALK (n = 294)</th>
<th>PKP (n = 79)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Double chamber</td>
<td>22.0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Rejection</td>
<td>1.3</td>
<td>11.8</td>
<td>0.0002</td>
</tr>
<tr>
<td>Infection</td>
<td>2.5</td>
<td>4.1</td>
<td>0.45</td>
</tr>
<tr>
<td>Secondary glaucoma</td>
<td>5.1</td>
<td>18.7</td>
<td>0.0002</td>
</tr>
<tr>
<td>Wound dehiscence</td>
<td>1.3</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Endothelial decompensation</td>
<td>0</td>
<td>1.4</td>
<td>0.21</td>
</tr>
</tbody>
</table>
Keratoplasty

- **Patient Selection**
  - Central corneal opacification
  - Contact lens intolerance
  - Inadequate BCVA
  - Advanced disease with high risk of perforation/hydrops (~200 central thickness)
Acute Corneal Hydrops
Clinical Presentation

- Sudden onset\textsuperscript{18}
  - Corneal edema
  - Vision loss
  - Epiphora
  - Pain
  - Photophobia
Epidemiology

• Affects 3% of keratoconus patients\textsuperscript{19}

• Risk Factors\textsuperscript{20,21}
  • Early age at Dx
  • Poor BCVA at Dx
  • Hx of eye rubbing
  • Males>Females
  • Family Hx protective

• Typically unilateral at presentation\textsuperscript{22}
  • 40% chance of fellow eye within 10 years
Pathophysiology

• Localized break in Descemet’s Membrane\textsuperscript{22}

• Inciting incident\textsuperscript{23}
  • Eye Rubbing
  • Valsalva
  • Idiopathic
Treatment Approaches

• Observation
  • Self-limiting with resolution in 5-36 weeks\textsuperscript{24}

• Topical\textsuperscript{25}
  • Hypertonic solution
  • Antibiotic
  • Corticosteroid
  • Cycloplegic
Treatment Approaches

• Surgical
  • Intracameral gas injection\textsuperscript{26,27,28}
    • Air, Sulfur Hexafluoride, Perfluoropropane
    • Pre-Descemet’s Sutures\textsuperscript{29}
• Improved recovery time: as little as 7 days
• Final visual recovery: equivalent
Outcome\textsuperscript{25}

- Varying corneal opacification
- Flattening of involved area
  - CL re-fitting
- 20% elect PKP within 6 months
- BCVA trends downs

![Box plot showing visual acuity comparison before and after hydrops. Median: 20/80 before hydrops, Median: 20/400 after hydrops.](image)
Treatment Paradigm

- Every Keratoconus Patient
  - Discourage Eye-Rubbing, prescribe topical MCS/AH PRN
  - Identify risk for progression
    - If progression documented or likely: Recommend CXL
    - If stable: monitor with serial topography
- Attempt non-invasive visual improvement
  - Spectacles → CLs
- Discuss surgical options
Citations