Management of Keratoconus
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Outline:

- Keratoconus
  - Epidemiology
  - Genetics
- Diagnosis
- Medical Management
- Surgical Management
  - Cross-linking
  - Intacs/CK
  - PK
  - IEK
  - DALK

Keratoconus Background

- Noninflammatory ectatic degeneration
  - Corneal findings include:
    - Paracentral cone
    - Progressive thinning
    - Irregular astigmatism
    - Scarring
  - Onset at puberty with progression through 30's
  - Asymmetrically bilateral
- Prevalence 54.5/100,000 people
  - True rates hard to determine due to varying definitions of disease along a spectrum from forme fruste keratoconus, pellucid marginal, and true keratoconus
  - No gender/race predilection
  - More common in youth
- Etiology
  - Multiple theories, likely some combination of all
    - Connective tissue disorder
      - Is known to be associated with Ehlers-Danlos, Marfans, and others
    - Enzymatic defect
      - Especially protease, tear lysozyme, lipid peroxidase, or nitric oxide
      - Possibly cause increased oxidative stress leading to broken collagen crosslinks and weakened corneas
    - Environmental/mechanical
      - Eye rubbing known to increase risk
      - Possible link between rubbing leading to trauma and inflammation
  - Genetics
    - Complex and poorly defined genetics
    - 6-20% of cases with known family history, but some studies suggest up to 60% of cases have family members with some topographical abnormality
Still most cases considered isolated

**Diagnosis**

- Symptoms:
  - Early cases easily missed as often asymptomatic at first
  - Later cases have decreased vision
    - progressive refractive change (increasing myopia and cylinder)
    - Inability to correct to 20/20
- Signs:
  - Scissoring of reflex on retinoscopy
  - Charleux oil drop sign
  - Fleisher ring
  - Munson’s sign
  - Rizzuti phenomenon
  - Vogt’s Striae
  - Stromal thinning, typically at apex
  - Scarring (later finding)
    - Subepithelial (breaks in Bowman’s)
    - Deep stromal (breaks in Descemet’s membrane, AKA hydrops)
- Testing
  - Topography
    - Highly sensitive, useful for monitoring progression
    - Topographic findings:
      - Focal area of increased corneal power
      - Inferior/superior power asymmetry
      - Skewed steep axis
  - Tomography
    - More sensitive than topography, gives information on posterior cornea and pachymetry that topography alone misses
    - Tomographic findings:
      - Posterior float (very early sign, not seen on topography)
      - Anterior float
  - Wavefront
    - Measures higher order aberrations
    - Keratoconus often has increased coma and spherical aberration
Treatment - Medical:

- Early/mild cases can be corrected with glasses
- Mild-to-moderate cases with increased irregular astigmatism need rigid gas permeable lenses
  - RGPs can increase scarring (if flat fit) or increase rate of progression (if tight fit). Fit is important!

Treatment - Surgical:

- Corneal cross-linking newest treatment option for all except the most end-stage cases
  - Does not reverse changes to a significant degree, but does prevent progression
  - Early treatment leads to better outcomes
  - Goal of treatment to increase strength of cross-links between collagen in cornea to prevent future ectasia
  - Concept has been around since 1990’s, treatment under investing since later 90’s early 2000’s, FDA approval in 2016!
  - Uses a photosensitizing agent (riboflavin) soaked onto cornea. Then expose cornea to UVA light (365nm wavelength) and riboflavin will increase cross-links
  - Essentially 100% effective at halting progression
  - Two procedure options: Epithelium-off (FDA approved) vs epithelium-on (still investigational)
- Implantable intraconal ring segments (aka ICRS or INTACS)
  - PMMA rings that are inserted into laser cut channel in the stroma
    - Approved for treatment of low myopia (flatten the cornea and thus reduce myopia)
    - Have been approved for use in KCN since 2004 under a humanitarian device exemption from FDA
    - Do not change disease progression, may flatten cornea, reduce I/A, reduce myopia, or improve contact lens fit
  - Conductive keratoplasty
    - A radiofrequency probe that causes focal collagen shrinkage
• Actually, increases corneal steepening, FDA approved for low hyperopia
• In KCN used to steepen flat axis to improve I/A and move apex of cone centrally
• Does not alter disease progression
• Often combined with INTACS
  o Combo INTACS/CK for mild to moderate cases with borderline contact lens tolerance

○ Penetrating keratoplasty
  ▪ Goal standard surgical treatment
  ▪ ~20% life-time risk in KCN population of needing a PK
  ▪ Indications
    • RGP intolerance
    • Scar
    • Thinning/perforation
  ▪ 80% 5-yr survival of graft (KCN patients do better than most with PKs)
  ▪ Younger patients and those with more ectasia are more likely to progress to needing PK
  ▪ PK enhancements
    • Deep Anterior Lamellar Keratoplasty (DALK):
      o Maintains original host cornea’s Descemet’s and endothelial layers
      o Less rejection and endothelial cell loss
      o Technically more challenging, may need to convert to standard PK intraoperatively (15-40% conversion rate)
    • Intralase Enabled Keratoplasty (IEK)
      o Utilizes femtosecond laser to cut host and donor corneas
      o Allows for more precise match between tissue
        ▪ Less astigmatism, quicker visual recovery