What We Will Cover

- Basic definition
- Proposed causes
- Symptoms and signs
- Contact lens alternatives
- Fitting procedure
- Fluorescein pattern interpretation
- Problem solving
Definition

Non-inflammatory, self limiting, ectasia of the axial portion of the cornea
Appearance

- Occurs .15-.6% of population
- 5-7% of refractive surgery candidates had sub-clinical KC
- Usually manifested in adolescence approximately age 16
- Onset, found by Hall in 1967, ages 6-26
- Occurs slightly more in females, 50.6% vs. 49.4%
- Generally bilateral (96%), although asymmetrical
Etiology

- Corneal tissue changes
- Atopic conditions
- Heredity
- Systemic conditions
- Contact lens wear
Primary Site of Keratoconus

- Corneal stroma vs. epithelium and its basement membrane?
Stromal Defect

- Collagen studies
- Confocal corneal evaluations
- Enzymes studies
- Apoptosis
Collagen Studies

- Collagen in apex is altered
- Collagen in para-apical areas not
- Little interaction between 2 areas
- Result is slippage or thinning
Morphologic Changes in Keratoconus

- Confocal studies – normal epithelial cells in periphery
- Apical cells were elongated and arranged in whirl-like pattern
- Fold-like changes in basal cell layer indicating disruptions in layer
Enzymes

- Studies show enzyme collagenase present in both KC and normals
- Collagenase active in KC, latent in normals
- Active collagenase breaks down stoma resulting in thinning
Free Radical Theory And Apoptosis

- Cornea absorbs 80% of UVB entering eye
- UV generates free radicals
- Free radicals removed by antioxidants in cornea
- If not removed form aldehydes
- Decreased level of aldehyde dehydrogenase (ALDH) in cones
- Nitric oxide + superoxides = peroxynitrites
- Peroxynitrites cause tissue damage
- Causes apoptosis of anterior stroma and epithelium
Oxidative Damage/Mechanical Stress

FREE RADICALS

- Aldehydes
  - Aldehyde dehydrogenase (ALDH)
    - Reversible Damage
      - Wound Healing/Repair
  - if not enough ALDH
- Nitric Oxide
  - Peroxynitrites
  - Irreversible Damage
    - Apoptosis ← (LAR)

UVB
Atopy
Eye Rubbing
Atopic Etiology
Eye-Rubbing Theory
Heredity
Keratoconus Inheritance

- Hammersmith – 8% blood relatives
- Finnish studies – in 2 studies, 9% and 19% of blood relatives had KC
- CLEK Study – 13.5% family history
- Gonzales et al (1992) – 58% parents of KC had abnormal topography indices
- Rabinowitz (1998) – 50% abnormal topography
Systemic Conditions

• Down’s
• Reiger’s
• Ehlers-Danlos
• Marfan’s
• Cruzon’s
Contact Lens Wear
GASSET - 1978 study

- 162 keratoconics - 26.5% previous PMMA wearers.
- Control group - 1248 patients fit with soft lenses - only 1 developed keratoconus.
keratoconus

epithelium

heredity

atopic

connective tissue disorders (Ehlers-Danlos syn.)

ocular conditions (RP/vernal conjunctivitis)

contact lenses

stromal etiology

systemic conditions
Diagnosis
Case History

- Listen to patient’s symptoms; ask right questions
- Symptoms
  - Monocular diplopia or polyopia
  - Ghost images or halos
  - Things being distorted more than blurred
- Frequent eye exams and Rx changes
Vision

- Gradual decrease in VA may be first clinical sign
- Decrease in VA at both distance and close
- Initially only one eye is involved
- Increase in astigmatism at oblique axis
Clinical Pearl

Anytime you have a patient who has a reduction in myopia with an increase in astigmatism - think Keratoconus.
Retinoscopy

- Shows a “scissor-like” reflex
- Reflex secondary to conical distortion
Ophthalmoscopy

- Circular, oblong or dumbbell-shaped shadow
- “Oil or honey droplet”
- Looks like indistinct cataract
- Fundus blurred or indistinct
Keratometry

- Earliest sign is lack of parallelism
- Mires distorted
- Minification of mires
Minification of mires

normal

keratoconus
OPTIKON 2000

Point Values
- Power: 50.82 D
- Radius: 6.64 mm
- Distance: 0 mm
- Meridian: 270 deg
- Height: 0 μm

Simulated K
- 3 mm Zone:
  - 51.32D (6.58) @87°
  - 46.15D (7.31) @177°
  - diff. = 5.17D

5 mm Zone:

7 mm Zone:

Index
- BFS: 52.04 D
- BFC: 9.09 D
- TI: 5.26 D

Pupil
- Mean Ø: 3.395 mm
- Offset: 0.582 mm @ 101°
Axial vs. Tangential

- Tangential = true
- Smaller, more centrally located patterns
- More extreme curvature values
Biomicroscopy

- Can see signs of KC in cornea
- Observe corneal thinning - central vs. peripheral
- Normal - .5 mm
  Keratoconic - .25-.45 mm
Munson’s Sign

- Protrusion of cone
- Alters shape of lower lid down gaze
**Lines of Vogt**

- Series of sharp, single whitish vertical lines
- Brush-like
- Middle layer of the stroma
- Due to stretching of stromal lamellae
- Seen in apex before scarring
- Disappear if press on the globe
Fleischer’s Ring

- Pigmented line - yellow-brown to olive-green
- Partially or completely encircles base of cone
- Produced by depositing of hemosiderin in epithelium just above Bowman’s membrane
Apical Scarring

- Small, grayish opacities
- Occurs when ruptures in Bowman’s membrane are filled with connective tissues from stroma
Hydrops

- Occurs with break in Descemet’s and endothelium
- Aqueous invades the stroma
- Causes edema and opacification
- Self-limiting
- Incidence - 5-15%
Rigid Contact Lens Visual Acuity
(Log Minimum Angel of Resolution)

Carney L.G.: Contact lens correction of visual loss in keratoconus.

74% CLEK pts RGP wearers
Contact Lens Designs

- Three Point Touch
- Apical Clearance
- Aspheric
- Large Flat Lens
- Mini-Scleral
- Soper
- McGuire
- Rose K
- Soft Lens
- Piggyback
- SoftPerm
- SynergEyes
- Semi-Scleral
- Scleral lenses
Contact Lens Fitting Methods
(Prior to 1959)

- Apical clearance
- Extremely flat central touch
- Bearing apex and superior portion of cornea
Three Point Touch

- Most popular
- 2-3 mm. of bearing
- Bearing on apex and two additional areas at the cornea mid-periphery
Criteria for Success

- Good centration
- Good interchange of tears
Apical Clearance Design

- Original fitting philosophy (PMMA)
- Vaults central cornea
- Bears mid-periphery
- For central cones
- Less traumatic for cornea
- Possible reduced VA
Korb Study (1982)

- 7 patients fit
- 1 eye clearance fit (8.0), 1 eye fit flat (9.4)
- Flat lens – 4/7 pts scarred within 1 yr
- Steep lens – 0/7 pts scarred
Conclusion

- Larger bearing lenses accelerate apical changes with scarring in early keratoconus.
Large Flat Lens Design

- Useful on a displaced apex
- Must have a good peripheral system
- Tight peripheral systems may lead to increased corneal scarring
Rose K Lens

- Diagnostic fitting set of 26 lenses
- Standard diameter 8.7 mm,
  also 8.3 and 9.0
- Decreased OZD with increased B.C.
- Tighter P.C. system with increased B.C.
Disadvantage

- Unknown peripheral curves
- Costly diagnostic sets
- Possible flare from small OZDs
- Limited .1 mm steps in BC
Soft Lenses

- Advantages of soft lenses
  - Better comfort
  - Protects cornea from abrasions
  - Reduces irregular astigmatism
  - Allows for spectacle over-correction
  - Can put minus power in contact lens
Specialized Soft Cone Lenses

- Flexlens
- Freeflex
- Hydrokone
• **Advantages**
  
  Protect cornea
  
  For RGP intolerance

• **Disadvantages**
  
  Two lens system
  
  Expense
SynergEyes High Dk Hybrid

- Paragon HDS 100
- 27% H₂O skirt
- 14.5 mm diameter
- 8.4 mm rigid center (9.0 mm in KC)
- 7.8 mm optic zone
- Four skirt radii choices for each base curve radius (3 for KC)
Fitting Procedure
All Patients Should Be Refracted
Keratoconic Fitting
Philosophy

Flat vs. Clearance

Flat = comfort, vision
Steep = less scarring
Use Diagnostic Lenses

- No formula approach
- Each patient different
- Consider using anesthetic
## Keratoconus Trial Lens Set

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<th>OZR</th>
<th>Power</th>
<th>Dia.</th>
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Choose a B.C. Midway
Between K Readings
Clinical Pearl

If patient wears a contact lens, use this as your first diagnostic lens.
Analyze Central and Peripheral Areas Separately
Check Consistency of Your Findings
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Blend: Heavy

Light blend = 0  
Medium blend = .1  
Heavy blend = .2
Problem Solving
Reduced Vision Acuity

- Incorrect power
- Residual astigmatism
- Lens flexure
- Corneal scarring
- Lens deposits
- Poor centration
Lack of Contact Lens Movement

- Peripheral curve “seal-off”
- Secondary curve “seal-off”
- Sharp junction OZ border
Lens Discomfort

- Flat lens
- Steep lens
- Excessive edge lift
- Lens intolerance
Corneal Staining

- Central
- Peripheral
  - Tight P.C.
  - Poor blend
- Dryness
- Partial blink
Fitting Technique

- Take k readings
- Initial lens - split the k’s
- Second lens - equal to the steep k
- Compare the consistency of the lenses
- Order the lens with flat enough PC’s
Fitting Tips

- Look for adequate movement and centration
- Distribute lens mass so no one corneal point is providing total lens support
- Use small optical zones and flat peripheries
- Allow for good tear venting and exchange
Indications for Surgery

- Decreased vision
- Dense corneal scarring
- Contact lens intolerance
- Progressive thinning
- Perforation
Surgical Alternatives

- Penetrating keratoplasty
- Lamellar keratoplasty
- Intacs
Penetrating Keratoplasty

- Incidence 10-20%
- 95% successful
- 50% of time contact lens required for best VA
- Large amount of astigmatism present after surgery
Lamellar Keratoplasty (DLK)

- Incision to 3/4th corneal depth
- Cornea removed to Descemets
- Donor cornea sutured in place
Advantages

- Not an intraocular technique
- Avoids complications of PKP
- Criteria for acceptable donor tissue not as stringent
- Post-op astigmatism is less
Disadvantages

- Technically difficult with large amount of thinning
- Post-op VA generally worse than with PKP
Intacs Corneal Ring

- Differential thickness
- Placed vertically
- VA improves 3 line sc, 2 lines cc
- Best for mild KC without scarring
Conclusions

- No specific etiology
- Bilateral, asymmetrical
- Classical signs aren’t always present
- Use rigid lenses for definitive diagnosis
- No “cookbook” fitting method
- Allow adequate setting time
- Check consistency when doing fittings
- Whenever possible, use apical clearance philosophy
- More advanced, use larger diameters
- Use referral criteria, not just VA
- PKPs have high success rates
Websites For Keratoconus

- National Keratoconus Foundation
  http://www.nkcf.org/
- KC information in Spanish
  http://www.nkcf.org/indexSpanish.html
- KC link discussion board
  keratoconus-link@cshs.org
Thank You For Your
Attention