It’s not rare…
if it’s sitting in your chair.
1 in 1,800
Kennedy et al. 1986

Keratoconus
- Noninflammatory?
  - Low grade inflammatory component
    - Lema et al. 2009
    - Tx with Pataday (Alcon) or Alrex (B+L)
- ectasia central/inferior cornea
- irregular astigmatism

Collaborative Longitudinal Evaluation of Keratoconus (CLEK) Study
- Observational (no intervention) study
- Funded by the National Eye Institute (NEI)
- 1,209 keratoconus patients were enrolled (1995 – 96) at 15 US sites
- 8 years of annual follow-up visits

Baseline Findings in the Collaborative Longitudinal Evaluation of Keratoconus (CLEK) Study
- Zadnik, Barr, Edrington et al.
  Investigative Ophthalmology & Visual Science
  December 1998

Age of Onset
- Puberty
- Earlier age of onset; more severe disease
- Progressive until the third to fourth decade
- CLEK Survey:
  - 90% diagnosed between ages of 10 and 39 years
  - Mean age of diagnosis = 27.3 ± 9.5 years
  - 98.7% of sample less than 50 years of age
- The Manchester KC Study: n = 29
  - Mean age of diagnosis = 21±8 years
Gender
- Literature reports relatively equal incidence between males and females
- CLEK Study sample: 56.4% male, 43.6% female
- Manchester KC Study 76% male

ASYMMETRIC CONDITION
<table>
<thead>
<tr>
<th>Flat Keratometry</th>
<th>Better Eye</th>
<th>Worse Eye</th>
<th>Difference</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>45.94 ± 4.10D</td>
<td>49.53 ± 5.83D</td>
<td>3.59 ± 4.46D</td>
</tr>
<tr>
<td>Steep Keratometry</td>
<td>48.51 ± 4.49D</td>
<td>52.86 ± 5.75D</td>
<td>4.35 ± 4.41D</td>
</tr>
<tr>
<td>High Contrast Best-Corrected Visual Acuity</td>
<td>7.30 ± 6.83 Letters</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Visual Acuity (CLEK Study)
- 88% were 20/40 or better through CLs
- 58% were 20/40 or better through manifest refraction
  - Is manifest refraction repeatable?
  - Is quality of vision adequate?

Vogt's Striae and Scarring

<table>
<thead>
<tr>
<th>Slit Lamp Biomicroscopic Sign</th>
<th>Number of Involved Eyes per Patient</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vogt's striae</td>
<td>427 (35%)</td>
</tr>
<tr>
<td>Fleischer's ring</td>
<td>169 (14%)</td>
</tr>
<tr>
<td>Corneal scarring</td>
<td>569 (47%)</td>
</tr>
</tbody>
</table>
Heredity

- Autosomal dominance with variable penetrance
- CLEK Study
  - 13.5% reported a family history of keratoconus at baseline
  - 18% reported a family history at Year 7
    - Parent, sibling, child, aunt or uncle

Common Associations

- Eye rubbing
- CLEK Study sample:
  - 49% rub both eyes vigorously
  - 1.8% rub only one eye vigorously
  - 3.2% were unsure whether they rubbed their eyes

Common Associations

- CLEK Study sample:
  - Hay fever or allergies – 53%
  - Asthma – 15%
  - Atopic dermatitis – 8%
  - 10 to 20% prevalence of atopy in the general population

Management of Keratoconus

Quality of Life in KC

Kymes, Walline, Zadnik et al.

American Journal of Ophthalmology

October 2004

Quality of Life (QOL) in KC

- Scores for CLEK subjects on all scales were consistent with patients with category 3 and 4 (advanced AMD) age-related macular degeneration patients, except for...
  - General health was better for CLEK subjects (they’re younger)
  - Ocular pain was worse for CLEK subjects (they are forced to wear GPs)
Keratoconus Patient Education

- You will not go blind from the condition
- Have a large and wonderful family
  - Average age of Dx 27.3 years in CLEK Survey
- LASIK is not a good idea
  - Both result in corneal thinning
- Back-off on the eye rubbing
  - However, do not induce paranoia

I fit CLs to enhance or improve vision, not to alter the progression of keratoconus.

GP Fitting Philosophies

- Flat = touching the cone apex
  - Better vision?
  - Delay the need for surgery? NOT!
  - Increases risk of corneal scarring?
- Steep = vaulting the cone apex
  - Less disruption to cone apex

Rigid Contact Lens Fitting Relationships in Keratoconus

- Edrington, Szczotka, Barr et al. *Optometry and Vision Science* October 1999

How Flat They’re Fitted

- 88% fitted apical touch
- Mild kconus (<45 D) 1.18 D flat (SD±1.84)
- Moderate (45-52 D) 2.38 D flat (SD±2.56)
- Severe (>52 D) 4.01 D flat (SD±4.11)
- Overall 2.86 D flat (SD±3.31)

“Ideal” RGP Keratoconus Fit

- Sagittal height of base curve to equal or slightly exceed the sagittal height of the cornea
- No excessive areas of tear/debris pooling underneath optic zone
- Excellent exchange of tears
Keratometry
- Readings are challenging
- Extend range by using a +1.25 D spectacle trial lens and adding 8 to 9D to the drum reading
  (+2.25 D add ~16 D to the drum reading)
- Use steep K (or slightly flatter) to select initial diagnostic contact lens

Fluorescein Pattern Analysis
- Goal #1
  feather "three-point" touch or slightest amount of apical clearance

FDACL
first definite apical clearance lens

Fluorescein Pattern Analysis
- Goal #2
  minimize area of tear pooling around base of cone
Fluorescein Pattern Analysis

- Goal #3
  average (to maximum) peripheral clearance

  might not be uniform at all axis

Keratoconus GP Prescription

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>BC</td>
<td>6.50 mm (51.92 D)</td>
</tr>
<tr>
<td>CLP</td>
<td>-9.50 D</td>
</tr>
<tr>
<td>OAD</td>
<td>8.8 mm</td>
</tr>
<tr>
<td>OZD</td>
<td>6.5 mm</td>
</tr>
<tr>
<td>SCR</td>
<td>8.50 mm</td>
</tr>
<tr>
<td>TCr/TCw</td>
<td>11.00 mm/0.2 mm</td>
</tr>
<tr>
<td>ct</td>
<td>0.14 mm</td>
</tr>
<tr>
<td>blend</td>
<td>medium</td>
</tr>
<tr>
<td>material</td>
<td>DK ≥60</td>
</tr>
</tbody>
</table>

Other Options

- Aspherics / Biaspherics
  - Tip: start by using manufacturer’s fitting guide and personalize over time

- Toric GPs
  - Tempting, but remember KC results in irregular astigmatism

Large OADs for KC

- Option for decentered cone apex
- Option to enhance initial comfort

- Sclerals
  - Indicated for KC patients who are intolerant of GP lens wear
  - Indicated for KC patients whose GP lenses eject or decenter often
Soft Lens Options for Keratoconus

- Thick center (generally 0.30 to 0.60mm)
  - To "mask" irregular astigmatism
- Lenticular (thinner) mid-periphery and periphery
  - To enhance comfort and O2 transmission
- "Custom" base curve and peripheral curve systems
  - To enhance fitting

Now available in a silicone hydrogel material
- Kerasoft IC (Bausch + Lomb Boston Group)
  - 14.5mm OAD
  - 8.0mm OZD
- Made in Definitive silicone hydrogel material
  - Silicone hydrogel (Dk = 60)
  - Water content = 74%

Piggy Back

- When?
  - Comfort (initial); 3-9 staining; bandage
- How?
  - Minus, plus, how much?
  - Over-keratometry
- Optical considerations
  - ~23% effect
- Materials?
  - S-H? Which one?
- Care system?
  - Default (Optifree, H2O2)
  - Problem-solving

SynergEyes Hybrid CL

- FDA approved for astigmatism (A and Duette), post-surgical (PS), keratoconus (KC and ClearKone), and multifocal (M and Duette)
- 14.5mm OAD and 8.4mm GP center
- Paragon HDS 100 Dk
  (Duette 130 Dk)
- 27% water
  soft skirt
  (Duette S-H skirt)

ClearKone is available in 11 different Vaults

SynergEyes Fluorescein Patterns

The fit of the vault is independent of the fit of the skirt curve. Each should be fitted separately.
3-9 Staining

- Copious lubricants
  - Explain reason to patient
  - Explain how patient will benefit
- Prescribe soft KC lenses
- Piggyback with daily disposable or silicone-hydrogel SCL
- Scleral lenses

KConus Comfort

- Initial discomfort and continues throughout day
  - re-contour edge
  - Piggyback with a daily disposable or silicone hydrogel CL
  - SCLERAL
- Initially tolerable, but wearing time is decreasing
  - "open-up" peripheral curve system

Refractive Surgery for Keratoconus

- Keratoconus is a corneal thinning condition ... and so is LASIK
- Could perform PRK custom ablation?
- Could perform PTK to reduce central corneal scarring (raised or proud scar; nebula)
- IOLs
  - Correct manifest refraction OR or contact lens OR?

Risk Factors and Prognosis for Corneal Ectasia after LASIK – Randleman et al. (Ophthalmology 2003)

- Preoperative risk factors
  - forme fruste keratoconus (88%)
  - Residual stromal bed thickness of <250 microns (70%) – mean 223 microns
    - Flap thickness ~ 160 microns + 12 microns per diopter of correction
- Average time to development of ectasia = 16.3 months

INTACS (rebar for the cornea)

- For mild / moderate KC patients (K < 57 D)
- If patient can’t tolerate GP lens wear
- Goal is to reduce K distortion and flatten K and to center apex
  - (Makes it easier to fit GP? I don’t think so! Generally Rx sclerals.)

Intracorneal ring segments for KC correction:
Long-term follow-up
Alio et al., J Cataract Refract Surg, June ’06

<table>
<thead>
<tr>
<th></th>
<th>Best spectacle Corrected VA</th>
<th>Steep K</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-Op</td>
<td>0.46 = 20/50</td>
<td>51.07 D</td>
</tr>
<tr>
<td>Mean 6 month</td>
<td>0.66 = 20/30</td>
<td>47.15 D</td>
</tr>
<tr>
<td>Post-Op</td>
<td>0.62 = 20/32</td>
<td>48.92 D</td>
</tr>
</tbody>
</table>
Corneal Collagen Cross-linking (C3-R)

- **Remove corneal epithelium** (or not)
  - Corneal thickness should be 455 microns (including epithelium)
- **Apply riboflavin 0.1%** (Vitamin B2) drops
  - Every 3min for 30min pre-op; then every 5min for 30min tx
- **Expose to ultraviolet (UVA) light** (370nm)

Coskunseven et al.  
*Journal of Refractive Surgery*, April 09

<table>
<thead>
<tr>
<th></th>
<th>Best spectacle Corrected VA</th>
<th>Max K</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-Op</td>
<td>0.29 = 20/70</td>
<td>54.02 D</td>
</tr>
<tr>
<td>Mean 9 month Post-Op</td>
<td>0.40 = 20/50</td>
<td>52.45 D</td>
</tr>
</tbody>
</table>

Complication and failure rates after corneal crosslinking  
Koller et al., *J Cataract Refract Surg*, August 09

- 117 eyes of 99 patients
- Baseline, 6 mos, and 12 mos follow-up
- Percentage of eyes losing ≥2 lines of VA
  - 2.9% (95% CI, 0.6-8.5%)
  - **Risk factors included >35 years of age**
- Percentage of eyes exhibiting progression
  - 7.6%
  - **Risk factor of steep K >58 D**

Post-Penetrating Keratoplasty

- Up to 50% of post-PK pts benefit from CL wear
- Over 50% of post-PK pts have 4 or more diopters of astigmatism
- Residual astigmatism is common
- Irregular astigmatism is most common
Corneal Buttons
- IntraLase femtosecond laser creates smoother graft edges
  - New cut patterns improve button fit
  - Reduces surface irregularities
  - Better outcomes

IntraLase Advanced Keratoplasty
- Increased surface area of contact.
- Early fibrosis leading to early healing.
- Slide courtesy of Dr. Dan Tran, TLC

Indications for Scleral GP CLs
- Pellucid Marginal Degeneration
- Keratoconus
  - If lens ejection cannot be resolved
  - If patient is intolerant to GP contact lenses
- Intolerance to GP lenses, but GP necessary for acceptable vision
- Severe Dry Eyes
  - Sjogren's syndrome
  - Graft-host disease

Scleral GP Contact Lenses
- >18mm in diameter?
- Fitting relationship
  - Vault cornea
  - Clear limbus
  - "Align" sclera
    - May Rx toric peripheral curves to assist in aligning
- Use high Dk (>100) GP material

"The vascular compression pattern is to scleral lenses what fluorescein patterns are to corneal GP lenses." - Perry Rosenthal, MD

When in doubt...
- Call the pros
  - Most GP labs will have a "guru" to help you trouble-shoot