Management of Keratoconus with Contact Lenses from Mild to Severe

Karen Lee, OD, FAAO, FSLRS
Rutvi Doshi, OD, FAAO
Susan Kovacich, OD, FAAO, Dipl
Options for Managing Keratoconus

- Spectacles/Soft spherical lenses/Soft toric lenses
- Conventional gas-permeables (GPs)
- Specialty GPs
- Intralimbal GPs
- Scleral GPs
- Hybrid lenses
- Specialty soft lenses
Case #1
Patient RR - 24yo Hispanic Female

- **CC:** distance blur OU
  - Onset: 4+ months ago
  - Gradually worsening
  - No improvement with soft contact lenses or spectacles
  - Denied eye rubbing

- **cc VA (specs)**
  - OD 20/40 PH 20/30
  - OS 20/70 PH 20/40

- **POHx:** keratoconus OU
- **FOHx:** unremarkable
Patient RR - Slit Lamp Findings

- (-) Vogt’s striae OU
- (-) Fleischer’s ring OU
- (-) Scarring OU
- (+) Mild stromal thinning OS

http://www.pacificu.edu/optometry/ce/courses/15167/images/Slide23.jpg
Patient RR - Corneal Topography

- OD sim K
  - 43.00@007 / 46.00@173

- OS sim K
  - 50.00@107 / 54.10@073
Patient RR - Corneal GP Lens Fitting

- Mild-moderate keratoconus
  - Reasonably centered cone on topography

- Choosing the first trial lens
  - Base curve (BC): between the steep K finding and the average K value, fudge towards the steep K

- Find the “steepest” trial lens BC that barely touches the apex of the cone
Patient RR - First Pair of Corneal GPs

OD CLEK 45.00 (7.50) / -1.87 / 8.6 OAD / 6.3 OZD  
OR: pl-0.50x105 20/20  
DVA: 20/25

OS CLEK 49.50 (6.82) / -5.00 /8.6 OAD / 6.3 OZD  
OR: -0.25-0.25x173 20/30  
DVA: 20/30
Patient RR - Second Pair of Corneal GPs

OD CLEK 44.62 (7.56) / -1.75 / 8.6 OAD / 6.3 OZD
- flattened by 0.37D, FAP, incorporated -0.25 DS SCOR, steepened PCS

DVA: 20/20-

OS CLEK 49.87 (6.77) / -5.37 / 8.6 OAD / 6.3 OZD
- steepened by 0.37, SAM, steepened PCs

DVA: 20/25+2
CLEK Study Group

Factors predictive of corneal scarring (Barr et al.)

1. Baseline corneal curvature

2. Contact lens wear

3. Corneal staining

4. Younger age
Corneal Cross-Linking?
Corneal Cross-Linking (CXL)

- Developed in Europe in late 1990’s
  - FDA approved April 18, 2016

- Halts progressive and irregular changes

- Improves corneal strength (300%) by strengthening and stabilizing bonds
Avedro Receives FDA Approval

Photrexa Viscous, Photrex and KXL system
Avedro CXL Candidates

Indications
- Keratoconus
- Pellucid marginal degeneration
- Terrien’s marginal degeneration
- Post-refractive ectasia

No information on patients:
- Younger than 14 years old, older than 65 years old
- Pregnant or breastfeeding

Recommended corneal thickness:
- No thinner than 400um

Maximum keratometric readings:
- None
CXL Contraindications

HIGHLIGHTS OF PRESCRIBING INFORMATION
These highlights do not include all the information needed to use PHOTREX A VISCOUS and PHOTREX A safely and effectively.
See full prescribing information for PHOTREX A VISCOUS and PHOTREX A.

PHOTREX A VISCOUS (riboflavin 5'-phosphate in 20% dextran ophthalmic solution) 0.146% for topical ophthalmic use
PHOTREX A (riboflavin 5'-phosphate ophthalmic solution) 0.146% for topical ophthalmic use

For use with the KXL System
Initial U.S. Approval: 2016

RECENT MAJOR CHANGES
Indications and Usage (1.2) 7/2016

INDICATIONS AND USAGE
PHOTREX A VISCOUS and PHOTREX A are photosensitizers indicated for use with the KXL System in corneal collagen cross-linking for the treatment of progressive keratoconus (1.1) and corneal ectasia following refractive surgery (1.2).

DOSAGE AND ADMINISTRATION
- Debride the epithelium using standard aseptic technique using topical anesthesia (3).
- Then instill 1 drop of PHOTREX A VISCOUS topically on the eye every 2 minutes for 30 minutes (3).

ADVERSE REACTIONS
In progressive keratoconus patients, the most common ocular adverse reactions in any CXL-treated eye were corneal opacity (haze), punctate keratitis, corneal striae, corneal epithelial defect, eye pain, reduced visual acuity, and blurred vision (6.1). In corneal ectasia patients, the most common ocular adverse reactions were corneal opacity (haze), corneal epithelial defect, corneal striae, dry eye, eye pain, punctate keratitis, photophobia, reduced visual acuity, and blurred vision (6.1).

To report SUSPECTED ADVERSE REACTIONS, contact Allergan at 1-844-823-3575 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

See 17 for PATIENT COUNSELING INFORMATION

Revised: 7/2016
CXL Contraindications...

- Herpetic keratitis
- Corneal scarring or opacification
- Severe ocular surface disease
- Concurrent infection
- History of poor epithelial wound healing
- Autoimmune diseases
Avedro CXL Procedure

1. Remove corneal epithelium

2. Apply Photrex Visous drops

3. Once flare is observed perform pachymetry

4. Expose to KXL ultraviolet (UVA) light for 30 minutes

5. Apply bandage contact lens
CXL: What can we expect at 1 year?

United States Multicenter Clinical Trial of Corneal Collagen Crosslinking for Keratoconus Treatment (Hersh et al.)

- Kmax decreased by 1.6D
- CDVA improved by an average of 5.7 logMar
- UDVA improved 4.4 logMar
- No significant change in ECC 1 year after treatment
CXL: What can we expect at 1 year?

Corneal Tomographic Changes After UV Cross-Linking for Corneal Ectasia 1-Year Results (Baksoellah et al.)

- Reduced Kmax
  - Preoperative Kmax (59.3 ± 6.4D)
  - Kmax 1 month (59.7 ± 6.0D)
  - Kmax 3 months (58.3 ± 6.3D), stable thereafter
- Preop thinnest point thickness no different 12 months postoperatively (437 ± 29 µm)
- Stable CDVA
CXL: What can we expect at 7 years?

**Results at 7-years after cross-linking procedure in keratoconic patients**
(Nicula et al.)

- Decrease in Kmin (1.6D) and Kmax (2.0D)
- Decrease in cylinder -4.45 D to -3.50 D
- Increase UCVA, an average of 0.78 to 0.679 logMar
- Increase CDVA, a mean of 0.64 to 0.52 logMar
Most Common Adverse Reactions
(Koller et al.)

1. Temporary stromal edema (up to 70%)
2. Corneal haze
   • Temporary (100%)
   • Permanent (up to 10%)
3. Punctate keratitis
4. Corneal striae
5. Eye pain
6. Epi defect
7. Reduced VA or blurred vision
Common CXL Questions

“Natural” corneal cross-linking?
- **Aging**: oxidative deamination reaction
- **Diabetes**: glycation

Which eye to start with?

Epi on?
- Biomechanical effect <50% of the standard procedure
- 23% of cases continued KCN progression after 1 year
  (Soeters et al.)
Patient RR - Clinical Pearls

• Keratoconus does NOT lead to blindness!

• Flat-fitting lenses will lead to scarring; so will vigorous eye rubbing

• Contact lenses will NOT slow down or reverse KCN progression – goal is to provide best vision correction

• Support for patients: National Keratoconus Foundation
  ▫ http://www.nkcf.org/
Case #2

Patient LW - 35yo African-American Female

- CC: blurred vision, lost habitual corneal GP lenses, would like a new pair
- POHx: keratoconus OU (Dx at age 25)
- FOHx: unremarkable
Patient LW - Clinical Findings

- OU (+) KCN / (+) Vogt's striae / (+) central apical scarring

- sc VA
  - OD: 20/1000
    - PH: 20/150
  - OS: 20/1250
    - PH: 20/125
Patient LW

• Sim K’s
  ▫ OD: 67.5 @ 010 / 73.0 @ 100
  ▫ OS: 50.5 @ 126 / 59.9 @ 036

• Lenses ordered and dispensed
  ▫ OD 65.50 (5.15) / -25.25 / 8.8 OAD / 6.0 OZD / 20/25
  ▫ OS 58.00 (5.82) / -16.50 / 8.8 OAD / 6.2 OZD / 20/25+

Patient was pleased with comfort and vision.
Corneal Cross-Linking Referral?

• Probably not...

  ▫ Patient is 35 yo
  ▫ Max K OD: 73.0 @ 100    Max K OS: 59.9 @ 036
  ▫ (+)Central apical scarring OU
  ▫ Financial burden
One Week Follow Up

“Not All Dimples Are Cute” - Dr. Brooke Messer

- CC: foggy vision after several hours of lens wear
- Modifying factors: blinking, artificial tears

OD

OS

DVA: 20/30-

DVA: 20/60-
Options?

• Adjust corneal GP parameters

• Intralimbal lenses?

• Scleral lens fitting?

• Piggyback with soft lenses
Goodbye, Dimples!

- Piggybacking OU
  - Air Optix Aqua Night & Day 8.4 / 13.8 /+0.50ds

OD

OS

DVA: 20/25

DVA: 20/25+
Piggybacking

- Lens selection
  - Dk
  - Replacement schedule
  - Modulus
  - Silicone hydrogel vs hydrogel

- Only 20.9% of lens power transmitted (Michaud et al.)

- What to look for:
  - Edge fluting
  - Independent movement of SCL from corneal GP
Soft Contact Lens Fluting

- **Modulus: Low vs High**
  - Acuvue Oasys: 0.72MPa
  - Air Optix Aqua N&D: 1.4MPa

- **Base curve**
  - Steeper can be better
Soft Contact Lens Fluting

Air Optix Aqua
N&D: 1.4MPa

Acuvue Oasys: 0.72MPa
8.4mm BCR

Acuvue Oasys: 0.72MPa
8.8mm BCR
Patient LW - Clinical Pearls

• Dimples are NOT cute!

• Consider piggybacking for:
  ▫ Poor lens centration / decentered cone apex
  ▫ Poor lens comfort; esp for first-time GP wearers
  ▫ Persistent corneal staining; esp 3-9 staining

• SCL selection for piggybacking:
  ▫ Low powers; do not have to refit or change power
  ▫ High Dk ie. silicone hydrogel
  ▫ Independent lens movement from corneal GP lens
## GP Classification

<table>
<thead>
<tr>
<th>Classification</th>
<th>Bearing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Corneal</td>
<td>Bears entirely on the cornea</td>
</tr>
<tr>
<td>Corneo-scleral</td>
<td>Lens bears on the cornea and sclera</td>
</tr>
<tr>
<td>Scleral</td>
<td>Lens bears entirely on the sclera</td>
</tr>
</tbody>
</table>

Table adapted from Scleral Lens Education Society
Benefits of intra-limbal GP designs?

- Improve comfort
- Enhance lens centration
- Stabilize visual acuity
- Modifiable peripheral curve system
- Reduce corneal desiccation and prevent 3 & 9 staining
Case #3
Patient PA - 67 yo African American Female

CC: Constant blurred distance vision OU and discomfort with current GPs x 1 year

PMHx
• Unremarkable

POHx
• PMD OU (15 yrs)
• Corneal ulcers OU (5 yrs ago)

VAs sc
OD: 20/200, ph 20/40+2
OS: 20/100, ph 20/30+1

Ocular Meds
• Refresh PFAT QID OU
Significant Slit Lamp Findings

• (+) Apical thinning
• (+) Inferior FR OS
• Subepithelial corneal scar OU
  ▫ OD: 2mm round w/marginal haze at 10:00 of pupillary margin
  ▫ OS: 1.5mm round superonasal to pupil
• Inferior SPK 1+ OS>OD
• Nuclear sclerosis cataract 1+ OU
Initial Visit

- Sim K’s from 1/19/2016
  - OD: 51.1@47/56.3@133
  - OS: 43.1@168/51.6@12
Irregular Cornea (IC)

- Post graft, keratoglobus, pellucid marginal degeneration and post-lasik ectasia fall into IC category
- Patient PA with (+)PMD OD>OS
  - Bilateral corneal disorder hallmarked by a thinning of the inferior peripheral cornea, 1-2mm above the inferior lumbus
  - Restricted to 4:00-8:00 region, exhibits a flat area inside this region and steepens rapidly as the cornea thins
  - The central flattening and peripheral steepening can make the GP fit difficult by creating poor lens stability and discomfort
Options Considered

- Adjust GP parameters
- Consider piggybacking
- Larger diameter – intralimbal lenses
  - Designed to sit inside the limbus to avoid limbal interference
  - Enhance lens centration on irregular corneas
  - Allows adequate tear exchange with adjustable peripheral curve system
Follow-up visit schedule and its importance
When should I re-fit
Case #4
Patient RL - 36 yo Hispanic Male

CC: Blurred vision with soft CLs, D/C’d GP wear due to recurrent erosions

PMHx
- ?Herpes

POHx
- Keratoconus
- Pingueculae

VAs sc
OD: 20/100
OS: 20/100

Ocular Meds
- Valtrex 1g PO BID
Significant Slit Lamp Findings

- Pinguecula OS>OD
- (+)VS, FR and anterior stromal haze OU
- SPK OS, suspected early dendrites
  - Culture negative for herpes
Pingueculae

- Common, non-cancerous growth of the conjunctiva

- Exact etiology is unknown
  - Long-term sunlight exposure
  - Eye irritation
  - Welding

- No treatment is needed in most cases
  - Frequent lubrication may prevent irritation
  - Consider removal for comfort or cosmetic reasons

- Patient history is key!
# Scleral Lens Fitting

<table>
<thead>
<tr>
<th>OD</th>
<th>OS</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Trial lens:</td>
<td>• Trial lens parameters</td>
</tr>
<tr>
<td>▫ 16.0 / 48.00D / -3.00</td>
<td>▫ 16.0 / 51.00D / -4.50</td>
</tr>
<tr>
<td>▫ CT: 350um</td>
<td>▫ CT: 350um</td>
</tr>
<tr>
<td>• SCOR</td>
<td>• SCOR</td>
</tr>
<tr>
<td>▫ -3.50 20/30</td>
<td>▫ -7.25 DS 20/25-2</td>
</tr>
<tr>
<td>• Pt reported good initial comfort</td>
<td>• Pt reported good initial comfort with slight lens sensation nasally</td>
</tr>
</tbody>
</table>
Initial Scleral Lens Assessment OU

- Settling time 20 min
- 1.5 center thickness apical clearance
- Good temporal limbal clearance
- Minimal nasal limbal clearance
- Haptics were snug by the pingeuculae OS>>OD
First Pair of Lenses Ordered

<table>
<thead>
<tr>
<th>OD</th>
<th>OS</th>
</tr>
</thead>
<tbody>
<tr>
<td>OAD: 16.0 / BCR: 48.00D / -6.50 / increased limbal clearance and flattened outer haptics</td>
<td>OAD: 16.0 / BCR: 51.00D / -12.12 / increased limbal clearance and flattened outer haptics and 4x2 notch</td>
</tr>
</tbody>
</table>
Scleral Lens Dispensing Appointment

<table>
<thead>
<tr>
<th>OD</th>
<th>OS</th>
</tr>
</thead>
<tbody>
<tr>
<td>DVA: 20/20-2</td>
<td>DVA: 20/25-3</td>
</tr>
<tr>
<td>SCOR: pl DS</td>
<td>SCOR: +1.00 DS 20/20</td>
</tr>
<tr>
<td>Central fit is great</td>
<td>Central fit is great</td>
</tr>
<tr>
<td>Impingement over pinguecula</td>
<td>Notch is not large enough</td>
</tr>
</tbody>
</table>
Do we dispense?

- Warn the patient that things may get tight
- Slow buildup of wear time
- Do not exceed 6-8 hours a day
- Assure the patient that the next pair will be more comfortable and vision will be clearer
Second Pair of Lenses Ordered

**OD**

- OAD: 16.0 / BCR: 48.00D / -6.50 / 2x2 notch

**OS**

- OAD: 16.0 / BCR: 51.00D / -11.12 / truncated
Two Week Scleral Lens Follow Up

CC: Pt notes great improvement in vision
-eyes get red and irritated with lenses
-no problems with application and removal

cc DVA (sclerals):
-OD: 20/20-
-OS: 20/25-3

AWT: all waking hours
WTT: 4h

OS: Questionable corneal staining has resolved, severe impingement near pinguecula with staining
Second Pair of Scleral Lenses

- Added a 2x2 notch
- Full truncation of lens edge
Parting Words

Pt reports great improvement in vision
-no problems with comfort or injection

DVA cc (sclerals):
-OD: 20/20-
-OS: 20/20

No signs of corneal or conjunctival staining OU

RTC 2 weeks for cornea check
What if scleral lenses are not an option?
Prosthetic Devices

• BostonSight PROSE

• EyePrintPRO
PROSE?

Prosthetic Replacement of the Ocular Surface Ecosystem
PROSE Treatment

- FDA approved in 1994
- Custom designed and fabricated prosthetic devices
- UCSF is one of thirteen PROSE centers around the country and one of two west of Texas
Profile of Scleral Lens: Spherical
Profile of Scleral Lens: Toric
PROSE Profile Possibilities
PROSE Profile Possibilities
Creation of Scleral Lens from Virtual Eye Model

Christine Sindt OD1,2  Bruno Lay 3

1. University Of Iowa Ophthalmology 2. EyePrint Prosthetics 3.ADCIS

christine-sindt@uiowa.edu
When to Refer for a Corneal Transplant

- Recurrent corneal hydrops
- Dense central corneal scarring
- Unsuccessful with all contact lens modalities
- Pt education is key!
Corneal Graft Indications

Current indications and surgical approaches to corneal transplants at the University of Toronto: A clinical-pathological study
Australian Graft Registry (2012)

**Penetrating and Endothelial Corneal Graft Survival**

**Fuchs’ Dystrophy and Bullous Keratopathy (Post 2003)**

**Penetrating:**

- **Mean Survival 63.51 months** (SE=1.48; 95% CI: 60.65, 66.37)
- **94% at 1 year**
- **88% at 2 years**
- **74% at 4 years**
- **51% at 6 years**
- **Median Survival 84 months (7 years)**

**Endothelial:**

- **Mean Survival 27.60 months** (SE=0.90; 95% CI: 25.04, 29.36)
- **77% at 1 year**
- **70% at 2 years**
- **Median Survival 38 months (3.2 years)**

**Penetrating Corneal Graft Survival**

**1st Graft for Keratoconus**

**1st Graft for Keratoconus:**

- **Mean Survival 18.22 years** (SE=0.29; 95% CI: 17.66, 18.78)
- **97% at 1 year**
- **95% at 5 years**
- **89% at 10 years**
- **77% at 15 years**
- **45% at 20 years**
- **Median Survival 19 years**
Graft Details

- Graft status
  - Endothelial cell count
    - No scleral lenses <800 cells/mm²
  - Age of graft

- Corneal profile
  - Prolate vs oblate
  - Irregularity indices
  - Protrusion areas

- Presence of sutures

- Contact lens history
### Post-Graft Fitting Principles

1. High oxygen permeability (highest Dk material)
2. Minimize lens center thickness and apical clearance
3. Reverse geometry design
4. Vault over graft-host junction and sutures
Case #5
DW 41- yo Caucasian Male

CC: Ocular discomfort OS
- OD: would like a new GP, no problems
- OS: GP ejection and discomfort especially when eye is dry

PMHx
• None

Ocular Meds
• None

VA cc (GPs)
OD: 20/20-2
OS: 20/20-2

POHx
• Keratoconus
• PKP OS
PK: Sweat the Details!

Age of graft: 1998 \textit{18 years old!}

Current drops: None

Slit lamp
- OS: Clear graft, frond of neovascularization at 12

Baseline pachymetry
- OS: 658um, 667um

Spectral microscopy
- OS: 761 cells/mm²

IOP
- OS: 12 mmHg GAT
Spectral Microscopy OS
Habitual GP OS

- Parameters: 10.8 / 8.6 / 7.26 / -5.50 / reverse curve
- Fit: Apical touch with midperipheral pooling and excessive edge lift inferiorly

Trial Scleral Lens Parameters OS

- Parameters: 16.0 / 7.18 (47.00) / -2.50 / rev geo / sph hapt
- SCOR: -7.25 DS 20/20
- Fit: 1 ct AC, nasal MP touch due to lens drop / LT 360 / EL at 6 and 12
Trial Scleral Lens Fit OS

- Lens drop
  - Commonly down and out causing superior nasal MP

- Haptics were not aligned
  - Incorporated a toric periphery
Two Week Follow Up

- VA CC: 20/20  AWT: 16h  WTT: 9h
- Fit: 1 ct AC, thins nasally MP / 360 LC / haptic alignment
- Pachy OS: 678um, 687um
- IOP: 14mmHg
PK After 9 Hours of Wear Time
Monitor CLOSELY

• Corneal edema
  ▫ 50um of swelling is considered significant
  ▫ Determine adequate wearing schedule
Graft Edema with Scleral Lens Wear
Fenestrating Against Corneal Edema
Monitor CLOSELY

- Neovascularization
  - Especially near sutures
  - Baseline photos help!
12 out of 31 (39%) experienced “serious graft complications”
  • Microbial keratitis (2)
  • Graft rejection (10)
    • 8 controlled with steroid, 2 re-grafted
  • Total failure rate: 19.4%
References


• Hersh PS, Stulting RD, Muller D, Durrie DS, Rajpal RK. United States Multicenter Clinical Trial of Corneal Collagen Crosslinking for Keratoconus Treatment. Ophthalmology. 2017;


• Nicula C, Nicula D, Pop RN. Results at 7 years after cross-linking procedure in keratoconic patients. J Fr Ophtalmol. 2017;


