CONTACT LENS MANAGEMENT OF KERATOCONUS

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Disclosures (TQ)

- Bausch & Lomb
- CooperVision
- GPLI

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- LENTECHS, LLC
- Menicon
- STAPLES Program

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Disclosures (EB)

- LENTECHS, LLC
- Consultant to the Contact Lens Manufacturers Association (i.e., Executive Director, GP Lens Institute)

"To the World You May be One Person, But to One Person You May be the World"

Anonymous

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DEFINITION

• Noninflammatory, self-limiting ectasia of the axial cornea. It is characterized by thinning, steepening and distortion of the apical corneal region

INCIDENCE

- What do we know about keratoconus???
- Most often begins in adolescents (10 20 y/o); 94% between 12 and 39; most severe cases are juvenile onset (i.e., 12 - 16y/o)
- Bilateral in 96 99.5%; one eye is more progressive
- 4 7 years progression
- 10 15% require a penetrating keratoplasty/some form of transplant; this % is decreasing due to scleral lenses and corneal cross-linking

Epidemiology

- Prevalence = 55/100,000 (about 1/2,000)
- Incidence = 2/100,000/year
- May be as high as 230/100,000 in 2016 (1/375)

IS IT ONLY BILATERAL???

- Recent studies have found little incidence of unilateral (0.5 - 4%) due to computerized videokeratography
- Li & Rabinowitz (2004, Am Acad Ophthalmol) initially found 14.9% unilateral in sample of 778 patients
- About 50 of "normal" eyes will progress to KCN within 16 years, most within 6 years
- If high inferior-superior dioptric asymmetry value = likelihood of KCN ______

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ETIOLOGY

- Corneal tissue change
- Atopic
- Systemic
- Contact lens wear
- Heredity & Genetic Predisposition

CORNEAL TISSUE CHANGE

- Epithelium or Stroma
- Although changes in basement membrane and Bowman's have been found, the earliest changes occur within the stromal lamellae
- Replacement of normal keratocytes by abnormal = abnormal collagen manufacture and ectasia

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APOPTOSIS

Kim, Rabinowitz, Meisler & Wilson (Exp Eye Res, 1999)

- Keratoconus accelerates keratocyte apoptosis
- Minor trauma (eye rubbing, poorly fitting CL, allergies) can release cytokines from the epithelium that stimulate apoptosis
- Begins in anterior stroma & is manifested by breaks in Bowman's layer & stromal thinning

Bergmanson (2018, GSLS)

Lamellar splitting = biomechanical weakening =
 ectasia

Subepithelial Fibrosis



#1 Disruptions in Bowmans



ETIOLOGY

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ATOPIC

- Primary factor associated with keratoconus
- Patients often have a history of dermatitis & hay fever
- Positive history of atopy in approximately 50% of keratoconics (CLEK Study); likewise raised serum levels of immunoglobulin E

ATOPIC

- Eye rubbing is very common; found in 70% of keratoconics in 600 patient sample approx.
 (50% rubbed eyes vigorously in CLEK Study)
- Trauma of eye rubbing may initiate response
- Eye rubbing with knuckles; may cause globe to give way at weakest point

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ETIOLOGY

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SYSTEMIC

- Connective tissue disorders such as Ehler's Syndrome, Rieger's Syndrome & Marfan's Syndrome have been associated with keratoconus as has Down's Syndrome
- CLEK Study does not support this relationship although a small but significant relationship is present with Down's Syndrome

ETIOLOGY

- Corneal tissue change
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CONTACT LENS WEAR??

- Possible association with long-term PMMA wear
- Possibly associated with low ocular rigidity combined with chronic hypoxia
- Some cases are actually corneal warpage syndrome (irregular astigmatism but no biomicroscopic signs or steepening present)

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ETIOLOGY

- Corneal tissue change
- Atopic
- Systemic
- Contact lens wear
- Heredity & Genetic Predisposition

IS IT GENETIC??

- Early studies showed 3 4% had a family member with reports of 8 twins
- CLEK found 13.5% had at least one family member
- Rabinowitz via topography found 50% of family members showed topo abnormalities suspect of KCN

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IS IT GENETIC?

- Yes, very likely; work of Rabinowitz
- Nielsen, et al Cornea Aug., 2005
 - Isolated 56 genes and validated five which were altered in keratoconus
 - Bottom Line: There is a connection; ongoing research should tell us what genes are defective, how these defects cause KCN and the genetic therapy to retard progression.

PULLING IT ALL TOGETHER

 The "Cascade" Hypothesis by Kenney & Associates



The Cascade Hypothesis of KC

Cristina Kenney Ph.D.

- Young people tend to produce high levels of reactive oxygen species (ROS) or free radicals in the cornea
- ROS typically cleared by superoxide dismutase and other enzymes to prevent accumulation
- Some PTs unable to produce protective healthy enzymes; free radicals accumulate causing damage to structural integrity of cornea

The Cascade Hypothesis of KC

Cristina Kenney Ph.D.

- Free radicals increase due to exposure to Ultraviolet-B light, mechanical trauma (contact lenses, eye rubbing) and atopic disease.
- With these accumulative traumas, there is a *deposition of cytotoxic by-products* resulting in;
- Ruptures Bowmans
- Stromal thinning
- Corneal scarring



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Clinical Implications

- If our hypothesis is correct:
- Benefit patients to protect from sources of ROS/RNS
 - UV light
 - Atopy/allergies
 - Vigorous eye rubbing
 - Poorly fit contact lenses
 - Inflammation

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Recommendations for KC & Ectasia-Prone Patients

- UV Protection Glasses or Contact Lenses
- Contact Lenses Fit Properly
- Preservative Free Artificial Tears
- Allergy Medications



- Introduced in Feb. 2020
- First and only commercial genetic test to assess risk for keratoconus
- 4 quick non-invasive cheek (buccal) swab samples
- Results arrive in 2 4 weeks
- Not only tests for keratoconus but also corneal dystrophies



DIAGNOSIS

- History
 - Refractive shift/Subjective symptoms
- Ophthalmoscopy
- Biomicroscopic
- Topography

Detecting the Odd Shaped Cornea

- Subjective Symptoms
 - Regular astigmatism
 - Blur/headaches/"double vision"
 - Irregular astigmatism
 - Blur/headaches/"double vision"
 - Refractive changes with time
 - Refractive endpoint ill-defined
 - Visual acuity reduced

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OPHTHALMOSCOPY

- Shadow
- Photodiagnosis
 - Dilated Pupil

 Examine at 2 feet; cone viewed against red reflex



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BIOMICROSCOPE

- Best for detecting subtle changes
- Apical thinning via optic section or pachometry (i.e., CT = .38mm vs. .50mm)
- Increased nerve visibility
- Vogt's Striae: grouped thin vertical lines in posterior stroma due to corneal stretching



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INCREASED NERVE FIBER VISIBILITY





Keratoconus Signs • Vogt's Striae (easier to see with CL on)

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Keratoconus Signs • Corneal Scarring



- The Collaborative Longitudinal Evaluation of Keratoconus (CLEK) study is an 1209 patient study which evaluates the etiology, demographics and progression of this condition
- Primary investigators Karla Zadnik & Joe Barr
- 8 year study

CLEK STUDY: BIOMICROSCOPY RESULTS

- Vogt's Straie: 65% ≥ 1 eye; 30% both eyes
- Fleischer's Ring: $86\% \ge 1$ eye; 56% both eyes
- Scarring: 53% ≥ 1 eye; 22% both eyes

CORNEAL TOPOGRAPHY



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CORNEAL TOMOGRAPHY

- Thinnest pachymetry < 500 microns
- Anterior elevation > 10 to 15 microns
- Posterior elevation > 15 to 20 microns

Martinez-Abad A, Pinero DP. J Cataract Refract Surg 2017;43:1213-27 Ucakhan O et al. J Cataract Refract Surg. 2011;37:1116-24 Mihaltz K et al. Cornea. 2009;28:976-80

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- Where?
- How large?
- How steep?



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Assessing the cone

• Where?

- Central: 28.7%
- Off-center:
 - Below horizontal: 87.8%
 - Average location: 262° (inf temp)
- How large?
 - Nipple (3 mm): 28.7%
 - Oval (3-5.5 mm): 44.3%
 - Globus (5.0 mm or above): 6.7%
- How steep?

HOW STEEP?*

- 95.4% steep K > 45D
- Form of Correction:
 - 73% corrected with GPs
 - 16% glasses

* CLEK Study

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Corneal Cross-linking (CXL) Aim of CXL is to halt or slow disease progression. Cross-linking is not a refractive Post-op evaluation for procedure. visual correction will be necessary.

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Corneal Cross-Linking

- FDA approved in April, 2016
- Has been found to result in 25% fewer corneal transplants over a 3 year period (Godefrooij, et al, 2016)



COLLEGEN CORNEAL CROSS LINKING (CXL)

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What is Corneal Cross-linking?

• Absorption of UVA by riboflavin • Creates new corneal collagen crosslinks

• UV light + riboflavin (vitamin B2) drops

- Results in a shortening and thickening of the collagen fibrils
- Leads to the stiffening of the cornea (2)

Beshtawi IM, O'Donnell C, Radhakrishnan H. Biomechanical properties of corneal tissue after ultraviolet-A-riboflavin crosslinking. J Cataract Refract Surg

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CXL Procedure: Epi-Off (FDA approved Dresden protocol) - Topical anesthetic instilled - Povidone-iodine wipe of outer orbit Lid speculum placed 9 mm debridement of central epithelium - Lid speculum removed Riboflavin instilled every 2 min for 30 min If not penetrated to 400 microns, hypotonic riboflavin instilled every 5-10 sec until achieved • Stromal cornea exposed to UV-A (365nm) for 30 min

- Post-Op:

• Prep:

- Irrigation and bandage lens topical antibiotics x 1wk
- Steroid x 2-3 wks

CXL Procedure: Epi-On (NOT FDA approved)

- May involve higher doses of UV light

 Shorter exposure time
- Effectiveness approx. 70% of epi-off ¹
- Faster recovery
- Less postoperative discomfort

1. Stulting RD et al. J Cataract Refract Surg 2018;44:1363-70

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OD Role is Critical!

- Early detection
- Prompt referral
- Proper post-Tx management
 - Haze is expected
 - Initial steepening, followed by flattening $(1.6^{1}-2D^{2})$
 - CL treatment?
 - Okay in 1-3 mo
 - Sclerals may be applied sooner than corneal lenses
 - Greenstein SA et al, J Cataract Refract Surg. 2011;37(7):1282-1290
 Wollensak G et al, Am J Ophthalmol.2003;135(5):620-627

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CXL Follow up Schedule

| VISIT | PLAN |
|-----------------|--|
| Day 1 to 1 Week | Topical antibiotic, steroid Frequent lubricants No eye rubbing Remove BCL once epithelium heals |
| Month 1 | OCT Imaging Tomography/Topography Vision assessment Contact lens refitting evaluation |
| Month 3, 6, 12 | Continued evaluation utilizing tomography/topography Vision assessment |

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Philosophy

- Either Epi-Off of Epi-On is better than no Cross-Linking at all
 - Today only Glaukos' (who purchased Avedro) Epi-off is FDA approved, and only CXL covered under some limited insurances
- Especially true in progressive young adults and pediatric patients
- Most important demographic to treat are young early progressive keratoconics
- You can save vision!

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Basic GP fitting concepts

- Small, central cone (eg.Nipple cone)
 → small diameter (<9.9 mm)
- Larger, more decentered cone (eg. Oval cone)
 → larger diameter (10.0-12.0 mm)
- REALLY BIG or extremely decentered cone (eg. Globus or PMD)
 - \rightarrow really big!
 - Scleral/ Mini-scleral design



Why Choose a Corneal GP?

- Rigid optics → Good vision!
- Wide range of parameters
- Easily obtained



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Why Choose a Corneal GP?

• Corneal lenses are less costly and often less complex than other CL options



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Material considerations

- High oxygen, but stable, materials
 - Usually high minus powers
 - Movement may be compromised
 - Stability important for good vision
 - Flexure
 - Warpage

Diagnostic fitting

- Key performance characteristics
 - Centration
 - Pupil within back optic zone
 - Movement
 - "adequate" lens movement



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GP FITTING PHILOSOPHIES

- Apical Clearance
- Apical Bearing
- Three Point Touch

APICAL CLEARANCE

 Apical clearance centrally minimizes any further compromise to the fragile apex of the cone





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Apical Clearance

- Szczotka-Flynn (CLEK): fit 30 eyes of current GP wearers .2mm steeper than FDACL
- Used 8.6/6.5mm OAD/OZD
- Over 12 months, their wearing time increased from 10.5 - 13.7 hrs; no decrease in VA; only 1/22 who completed study developed scarring

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Corneal Scarring

(CLEK: Collaborative Longitudinal Evaluation of Keratoconus)

- Steeper corneal curvature increased risk of corneal scarring
- 26% per diopter of increased curvature
- 15% risk of developing a scar over 5 years
- Flat fitting lenses have a 3.5% increased risk of developing scar vs steep fitting lenses

 Each D fit flat increases risk of scarring by 19%

Comparison of flat and steep rigid contact lens fitting methods in keratoconus. CLEK Study Grp Optom Vis Sci. 2005 Dec;82(12):1014-21.

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Corneal Scarring

(CLEK: Collaborative Longitudinal Evaluation of Keratoconus)

Odds of scarring

- Contact lens wear: 3.51
- Corneal staining: 3.4
- Fleisher's ring: 1.63
- Age: 1.54
- Steeper cornea: 1.29

Factors associated with corneal Scarring in the (CLEK) Study Cornea. 2000 Jul;19(4):501-7.





Start with central (apical) fit Look for light (feather) touch

Diagnostic fitting



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Diagnostic fitting

- Start with central fit
- Second: peripheral fit

 Only after central fit achieved
 Flatten or steepen accordingly



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Corneal GP Comfort & Keratoconus

- No association with disease severity
- No association with apical (central) fitting relationship
- Minimal peripheral clearance may decrease comfort

Variables Affecting Rigid Contact Lens Comfort in the Collaborative Longitudinal Evaluation of Keratoconus (CLEK) Study Optom & Vis Sci, March 2004 81(3):182-188.



Diagnostic fitting

- Start with central fit
- Assess peripheral fit
 - Multiple curves that gradually flatten
 - Aspheric curve
 - Curves associated with standard design

Quadrant Specific Edge

- Flexible Edge Lift System
- Asymmetric Corneal Technology (ACT)



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Diagnostic fitting

- Start with central fit
- Assess peripheral fit
- Determine final lens diameter

 Nipple cones
 - Start small (8.5-9.0 mm)
 - Steeper cone, go smaller
 - Oval cones
 - Increase OAD to aid centration
 - Decrease OAD to aid removal

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FITTING GUIDELINES

 Generally the optical zone should be decreased in size as the BCR steepens; a rule of thumb is selecting OZD = BCR in mm; for example:

If BCR = 7.00mm; OZD = 7.00mm

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FITTING GUIDELINES (Bennett)

: Stage 1

Stage 1 Average K < 45.00D Conventional lens design Stage 2 Average K = 45 - 50D OAD = 9.0mm OZD = BCR Tetra or pentacurve PCR = 12.00mm PCW = .3 - .4mm

FITTING GUIDELINES

Stage 3 Average K = 50-55D OAD = 8.6-8.8mm OZD = BCR Tetra or pentacurve PCR = 12.00mm PCW = .3-.4mm Stage 4 Average K = 55+ OAD = 8.0mm OZD = BCR Pentacurve PCR/PCW = "3"

FITTING GUIDELINES

 Even though keratoconus patients often have larger amounts of corneal astigmatism as indicated by keratometry, bitoric lenses are rarely recommended.



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INTRALIMBAL DESIGNS



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LARGE OAD DESIGNS

- 10 11.5mm designs used in cases where small designs are unsuccessful (i.e., inferior cones, globus cones)
- Examples: Dyna Intra-Limbal (Lens Dynamics) & Rose K-IC (Blanchard); the latter has an aspheric periphery and aberration control optics

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Dyna Intra-limbal Lens

- Lens Dynamics
- 11.2 mm OAD
- 9.4 mm OZ
- Steep, median, or flat periphery
- Menicon Z = primary material of choice

DIL Fitting

- With topographies Start 4-5mm temporal reading
- Manual K's Start .2mm flatter than mean K
- Alignment fit with light feather touch.
- Movement is only 1/2 1mm



ROSEK2

Fitting Guidelines The use of the Rose K2 IC diagnostic fitting set is the only way to properly assess the correct Rose K2 IC lens fit.

Topography: Define Steepest Corneal Meridian Initial Lens = 3 Diopters Flatter.

Trial Lens = Standard Edge Lift Geometry

Slight Peripheral Seal = Standard Flat Significant Peripheral Seal = Double Flat Excessive Lift = Standard Steep Over Refract for Best achieved V/A

Initial Base Curve Selection:

Evaluate Edge Lift:

Options:

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- Indications
 - Poor comfort with a corneal GP
 - Minor corneal abrasion with GP wear
 - Temporary use of soft lens



Piggyback (PB) Lens System

- Challenges
 - Extra cost
 - Extra hassle
 - Daily disposable soft minimizes added care



Minus Rx

Plus Rx

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Piggyback (PB) Lens System

- Fitting Tips- Approach #2
 - Fit soft lens first
 - If steep cornea, fit minus lens — Provides flatter fitting surface
 - If flat cornea, fit plus lens
 Provides steeper fitting surface
 - Then fit GP lens to front surface of SCL

Piggyback (PB) Lens System

- Fitting Tips- Approach #1
 - Fit corneal GP first
 - Then fit "near plano" soft lens underneath
 - Good for temporary use (ie. abrasion)



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Piggyback (PB) Lens System

- Power Determination
 - Approximately 21% of labeled soft lens power translates*
 - Perform O.R. over PB System to arrive at final GP power

* Michaud L, Brazeau D, Corbeil ME, et al. Cont Lens Anterior Eye. 2013;36(6):318-23.

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Piggyback (PB) Lens System

- Soft Lens Fitting Tips
 - Try DD SiHy first
 - If loose or excessive edge lift:
 - Steepen base curve
 - Increase diameter
 - Decrease center thickness (improve drape)
 - May need custom soft lens

Piggyback (PB) Lens System

- GP Fitting Tips
 - Central excavation design
 - GP sits in excavation
 - To improve GP centration



Piggyback (PB) Lens System

- Oxygen considerations
 - Double barrier system
 - Use high O2 materials



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FITTING SCLERAL LENSES

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Taking it to the Bank

- 46 yo w, m, banker
- Keratoconus OU
- Wearing piggyback lenses

 Instilling wetting drops approximately 16x/day
- Whatcha gonna do?

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Basics of Scleral Lens Fitting

Corneal clearance

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Bubble Trouble

- Lens application error!
 - Fill bowl with preservative-free saline
 - Patient face parallel with floor
 - "Elevator Technique"



















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DescriptionDescriptionOptimizing the provided of the p

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Basics of Scleral Lens Fitting

- Corneal clearance
- Limbal clearance
- No edge lift or scleral blanching



If complaints of fogging...

- Bubble?
- Surface wetting?
- Chamber debris?
- Corneal edema?

Taking it to the Bank

- Good vision and comfort with scleral lenses
- Using rewetting drops <u>1-2 x per week</u>



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Who Will Benefit From UltraHealth?

- Select patients with the following conditions:
 - Keratocon
 - Ectasias
 Intacs®
 - Post-Surgical
 - Other Irregular Corneas
- Corneal GP patients wanting better comfort and/or acuity
- Piggyback patients wanting improvements
- Scleral patients where eye health is especially important
- Where does lens not work as well?
 Too much lenticular culie does
 - Too much lenticular cylinderGlobus and tilted grafts

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If pooling (clearance) is seen

Once First Bearing is Observed

If feather touch or light bearing, add an additional 100µ to lens.
 If heavy bearing, add an additional 150µ to the lens.
 Goal: Vault 100-150µ above apex of the comea

• Decrease the vault in 100µ increments until **the first bearing is observed**

Inserting the Diagnostic Lens

 Fill the bowl of the lens completely to the top with 1 drop of fluorescein (or use a fluorescein strip) and nonpreserved saline.

- High molecular fluorescein not required

Have the patient lean forward and tuck their chin to chest

Nose should be perpendicular to the floor

Retract the upper and lower lids and gently place the lens on the cornea

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WHAT ABOUT SOFT LENSES?

- Increasing in popularity, notably for early/mild keratoconus
- Several designs available including Hydrokone (Visionary Optics), KeraSoft (B + L), and NovaKone (Alden)
- They also have a toric lens available with any cylinder or axis. Steep base curve radii
- KeraSoft has the benefit of the Definitive Si-Hy material

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Bottom Line on Contact Lens Selection: Type of Cone

- Nipple: Small OAD/OZD
- Oval: Small design or IL (if large oval)
- Globus: IL or Mini-Scleral(possibly PB or hybrid)
- Marginal/Inferior: Same as globus



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Bottom Line on Contact Lens Section: Progression

- Mild: Cross-linking, Conventional GP, small Kcone design or soft Kcone lens
- Moderate: Small Kcone (nipple), IL (Scleral if IL decenters)
- Severe: Scleral

WHAT PERCENTAGE OF YOUR IRREGULAR CORNEA PATIENTS DO YOU FIT INTO EACH OF THE FOLLOWING MODALITIES?

| LEN | S TYPE | ≥ 20% OF PATIENTS | ≥ 50% OF PATIENTS | | | |
|-----------|--|-------------------|-------------------|--|--|--|
| T A Scler | ral Lenses | 73% | 52% | | | |
| E Sma | ll-Diameter GPs | 48% | 18% | | | |
| Intro | limbal | 26% | 10% | | | |
| Hyb | rid | 22% | 5% | | | |
| Cust | om Soft | 28% | 4% | | | |
| Pigg | yback | 9% | 1% | | | |
| | | | | | | |
| | | | | | | |
| CONTACT L | CONTACT LENS SPECTRUM OCTOBER 2020 cls | | | | | |

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Useful Websites

- GP Lens Institute: (www.gpli.info)
- National Keratoconus Foundation: (www.nkcf.org)
- International Keratoconus Academy of Eye Care Professionals: www.keratoconusacademy.com)
- Scleral Lens Education Society (www.sclerallens.org)

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MODULE COMPONENTS

- Coding and Billing Webinar: Dr. Clarke Newman
 (new updated now being posted)
- FAQs: 34 common questions and the answers
- Ten Most Common Errors
- 3 sample letters of medical necessity
- Medically Necessary Contact Lens brochure
- Codes in common use
- Quarterly student webinar GPLI in April: coding and billing for specialty CLs: Dr. Stephanie Woo

N K C F National Keratoconus Foundation

- National Keratoconus Foundation: An organization that supports research and education about keratoconus:
- Newsletter for patients
- Educational Booklet for patients
- Referrals

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SUMMARY

- Advancements in locating the "Keratoconic Genes" as well as effective treatments are occurring rapidly
- Specialty GPs and hybrid lenses are the primary treatment options
- Topography is important

