



# Comprehensive Management of the Patient with Keratoconus

Jason Jedlicka, OD, FAAO  
Indiana University School of Optometry

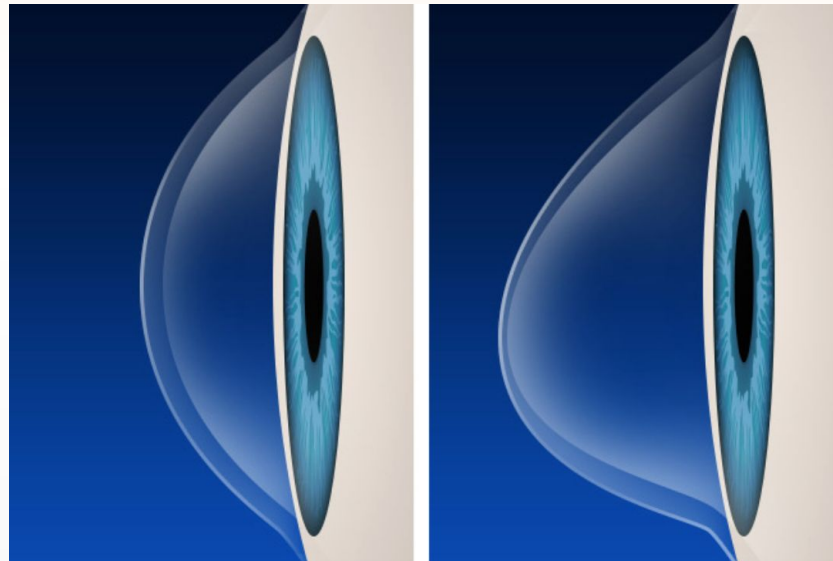


# Disclosures

- Consultant
  - Bausch and Lomb
  - Oculus
- Shareholder
  - Lens Design Associates

# What is Keratoconus?

- Definition: Keratoconus (KC) is a progressive, noninflammatory, bilateral (but usually asymmetric) **ectatic** corneal disease, characterized by **thinning** and weakening that leads to corneal distortion. Onset occurs between the second decade and sixth decade, with earlier onset correlated to more severe disease. Visual loss occurs primarily from irregular astigmatism and myopia, and secondarily from corneal scarring.



# How Does One Develop It?

- Genetic:
  - Monozygotic Twin Concordance
  - First Degree Relatives – increased risk (6-10%) vs. general population (.01%)
  - To date, 17 distinct genomic loci have been mapped for keratoconus indicating that there exists high degree of genetic heterogeneity
  - Keratoconus has been associated with numerous systemic and ocular disorders

# How Does One Develop It?

- Environmental:
  - Discordance for keratoconus in two pairs of monozygotic twins. McMahon T, Shin J, Newlin A, Edrington T, Sugar J, Zadnik K. *Cornea*. 1999 Jul;18(4):444-51.
  - Keratoconus, allergy, itch, eye-rubbing and hand-dominance. McMonnies CW1, Boneham GC. *Clin Exp Optom*. 2003 Nov;86(6):376-84.
  - *“The complexity of KTCN makes it difficult to identify factors influencing its development.”* Nowak D, Gajecka M. *The Genetics of Keratoconus*. *Middle East Afr J Ophthalmol*. 2011 Jan-Mar; 18(1):2-6.

# How Does One Develop It?

**Genetic**

(Inherited condition)

**Environmental**

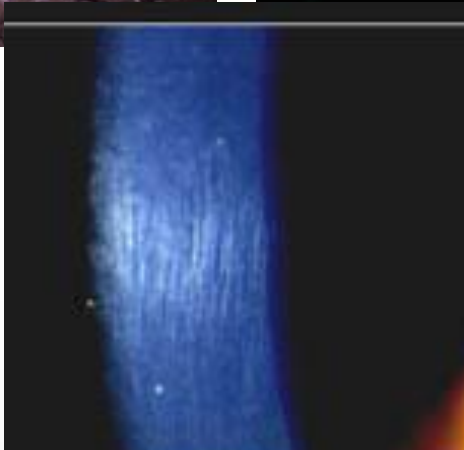
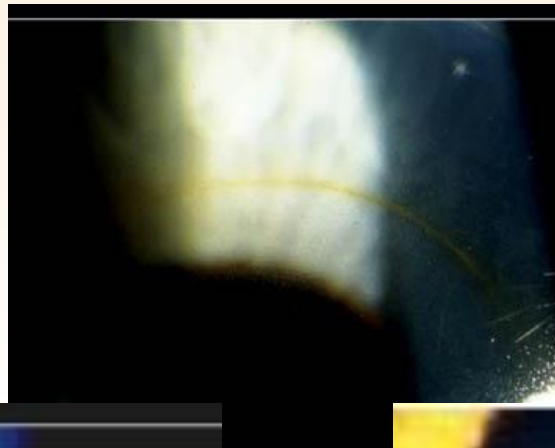
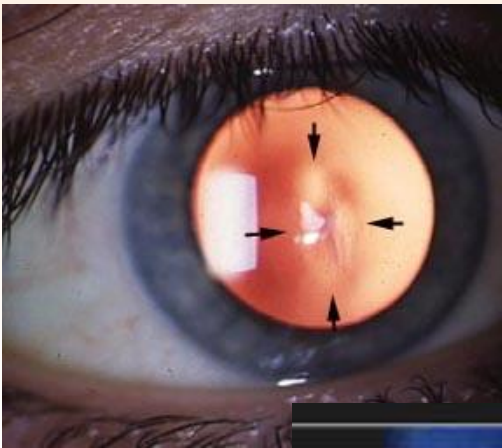
(Inherited risk / environmental stimulus)

# How is it Diagnosed?

- Initial suspicion of KC may arise from reduction in BCVA with an increase in astigmatism
- **Retinoscopic reflex should demonstrate scissoring**
- Slit lamp findings may not be present in earlier stages of disease
- **Corneal topography / tomography and pachymetry are the definitive diagnostic tests**

# Clinical Findings

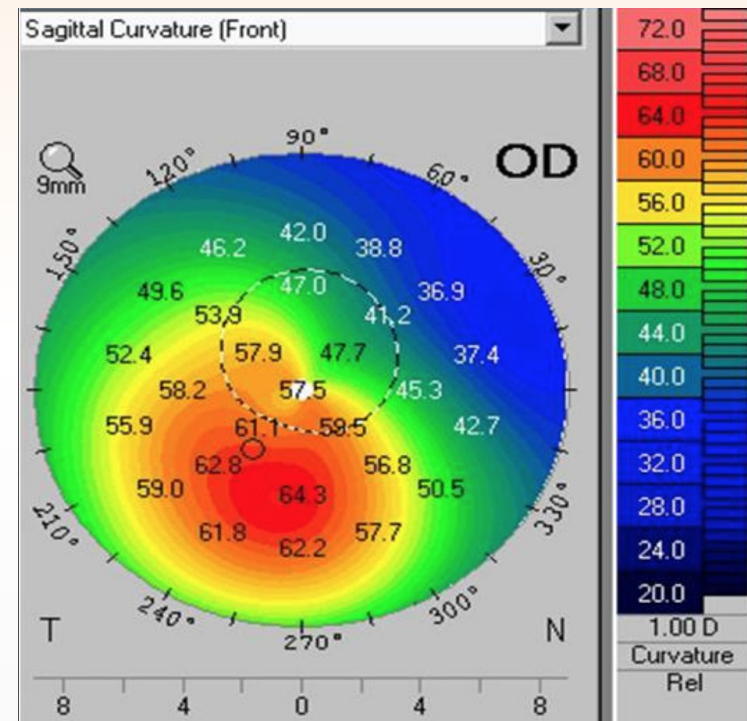
Often do not become apparent until later in the disease process





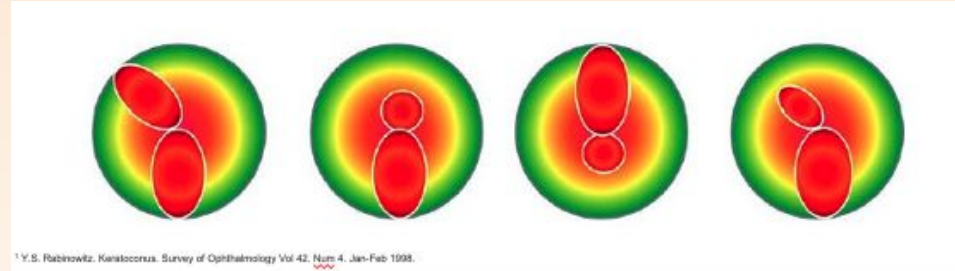
# Corneal Topography

- Classic keratoconic topography demonstrates an area of steepening, usually inferior

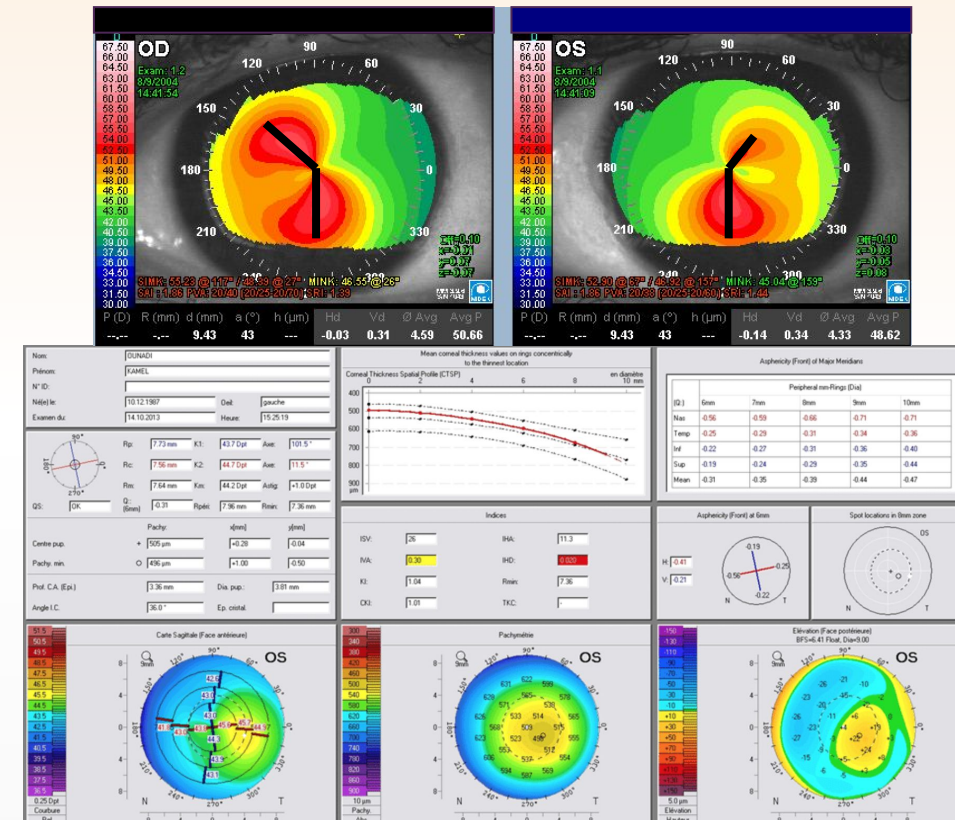


# Corneal Topography

- What are the important features of a topography to look at?
  - Steep Central K's
  - Asymmetric Bowtie**
  - Skewed Radial Axes > 21 degrees**

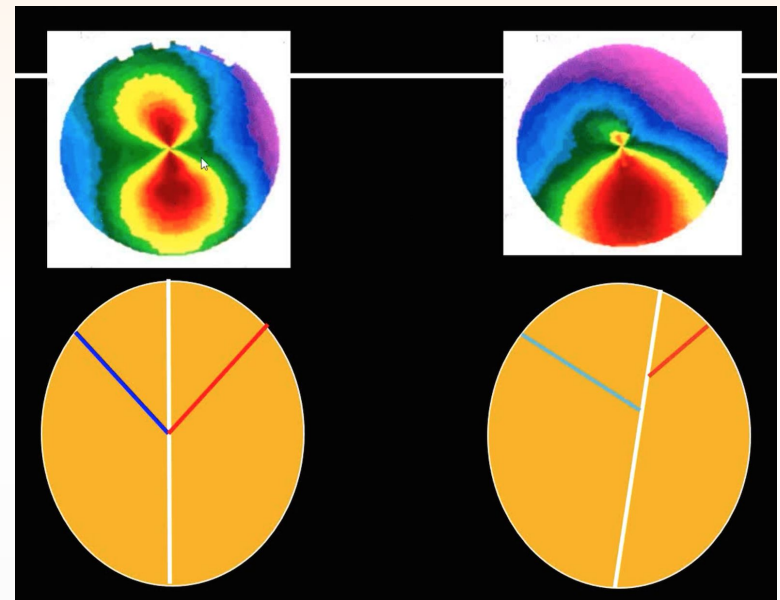


- Most topographers have Keratoconus screening Indices available



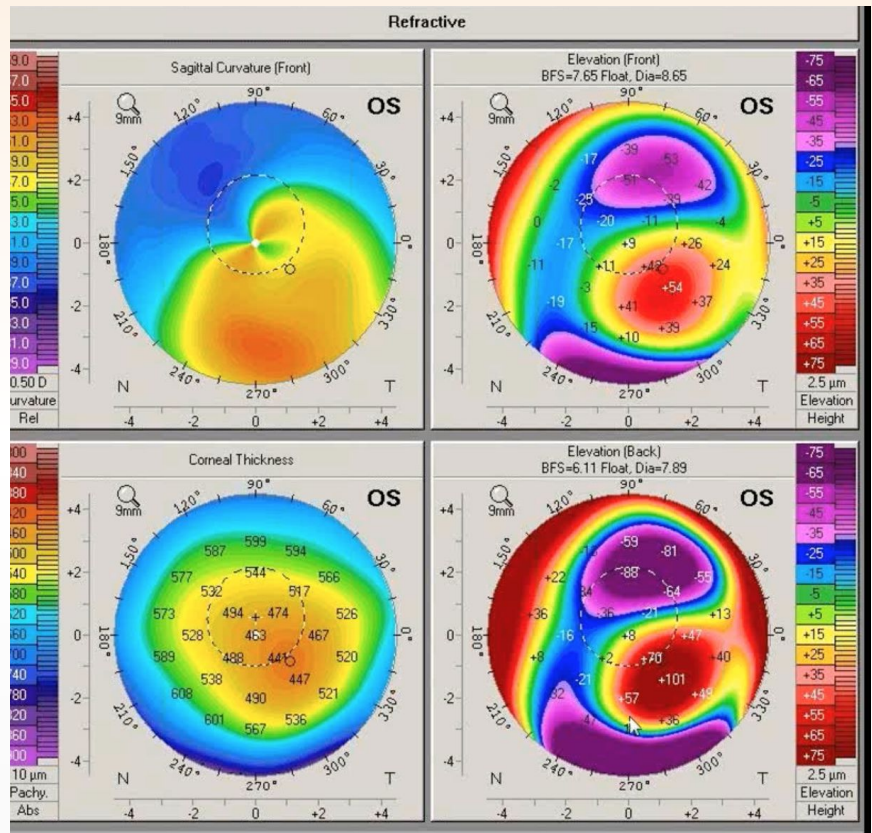
# Corneal Topography

- Problems with relying on corneal topography:
  - **Does not tell you anything about thinning / thickness profile**
    - **Keratoconus is a disease of thinning**
  - Does not tell you anything about the posterior corneal shape
    - **The posterior corneal changes occur earlier and more obviously than anterior corneal changes**
  - Can get false positives if visual axis and corneal apex are offset



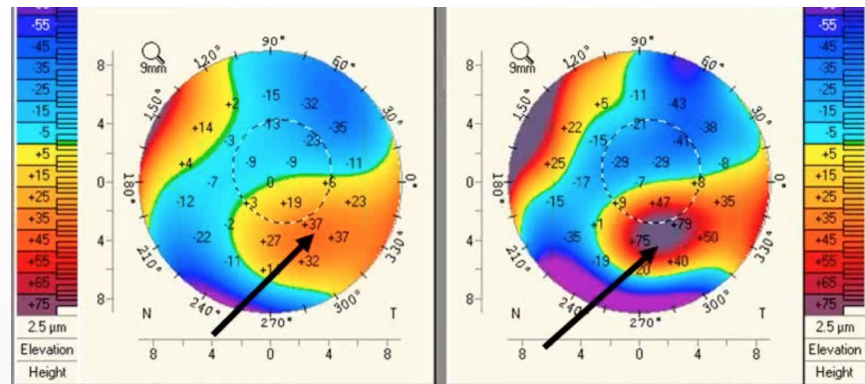
# Corneal Tomography

- Two dimensional “slice” through a three dimensional object
- Provides true elevation of the anterior and posterior cornea
- Corneal thickness
- Corneal curvature
- Best tool for detecting early KC as it can provide all data that is relevant for diagnosis



# Corneal Tomography

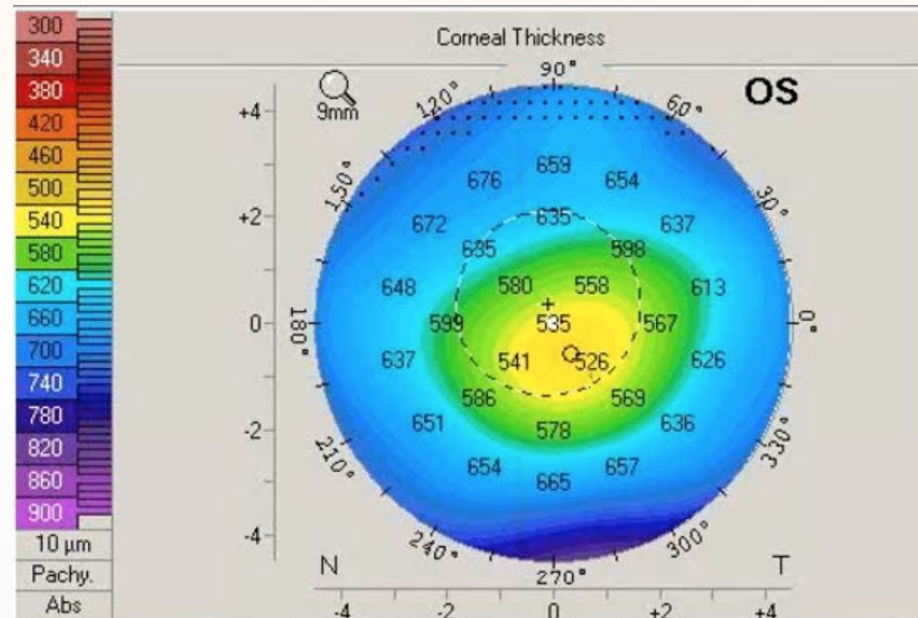
- Benefit of Corneal Tomography:
  - **DOES** tell you about thinning / thickness profile
    - Keratoconus is a disease of thinning
  - **DOES** tell you about the posterior corneal shape
    - The posterior corneal changes occur earlier and more obviously than anterior corneal changes
  - **DO NOT** get false positives if visual axis and corneal apex are offset





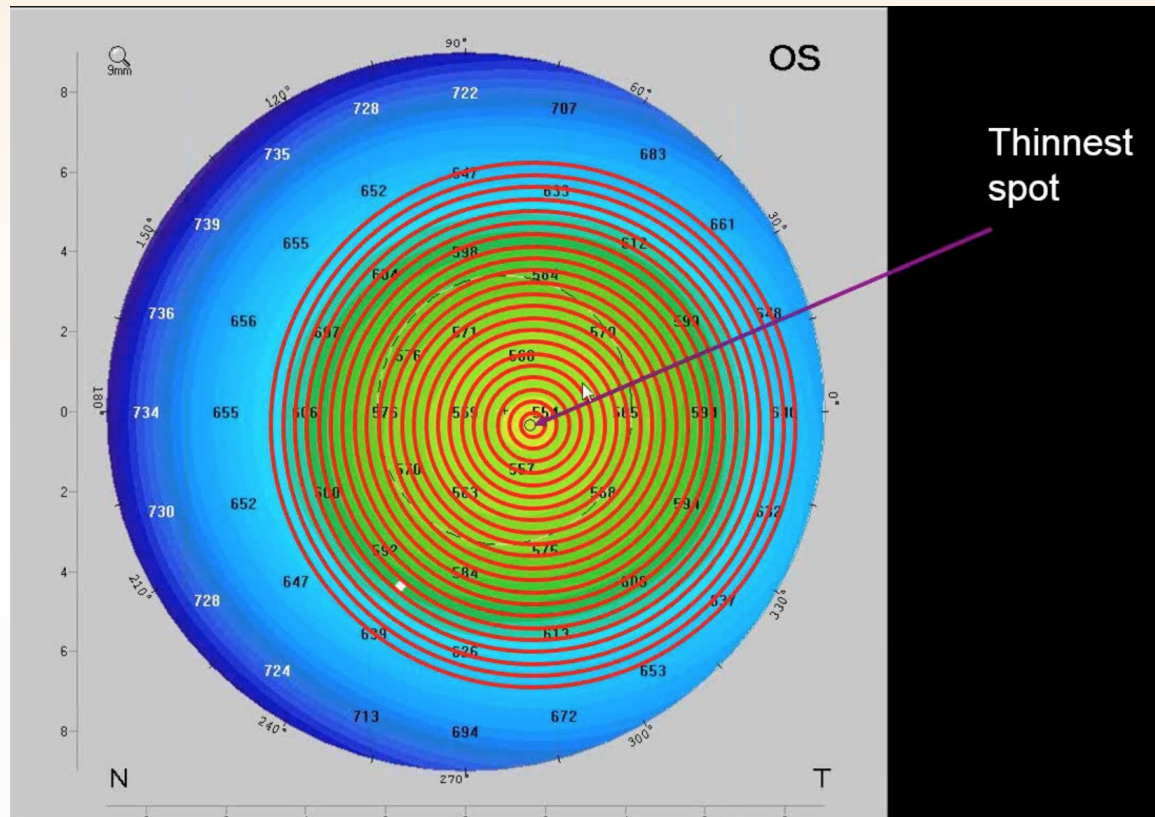
# Corneal Pachymetry

- Keratoconus is a disease of thinning
- How can we know if subtle thinning is early keratoconus or simply a thinner cornea?



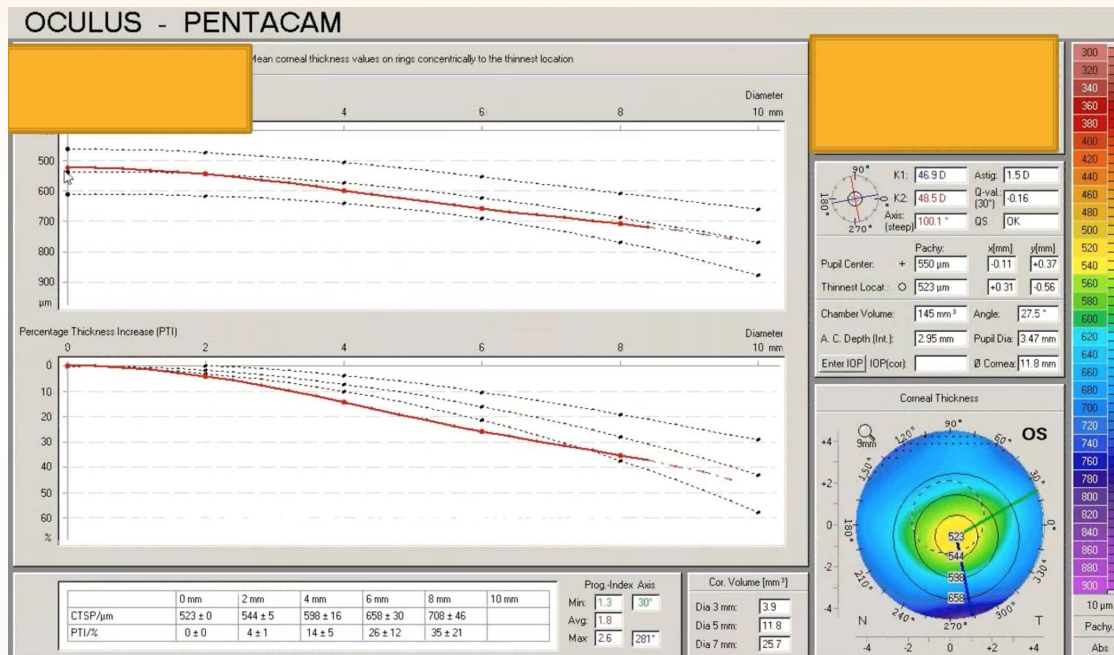
# Corneal Pachymetry

- Pachymetry Progression
  - Rate of increase in corneal thickness from thinnest point over a span of the cornea



# Corneal Pachymetry

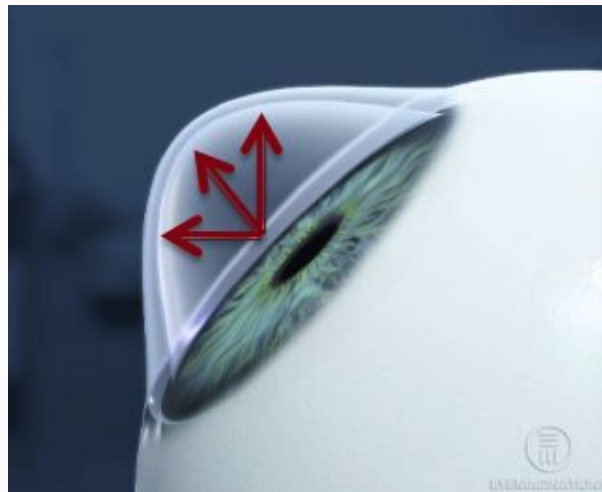
- Pachymetry progression plots out the rings compared to normative data
- If keratoconus exists, the thinning is typically a localized thinning, which will manifest in a greater rate of thickness CHANGE over the span of the cornea





# Diagnosing KC

- Abnormal POSTERIOR > Anterior Ectasia
- Clinical non-inflammatory corneal thinning leading to Abnormal corneal thickness DISTRIBUTION





# Management of KC in 2023

- Early detection if and when possible
- Slow / halt progression
  - Corneal collagen crosslinking
- Provide optimal visual correction while doing no harm
  - Spectacles
  - Contact lenses
  - Corneal rings
  - TG-PRK
- Monitor for further progression
- Corneal Transplant when other treatments are inadequate

# Why is Early Detection Crucial?

- With the ability to slow / or halt entirely the progression with Corneal Collagen Crosslinking, the earlier KC is detected, the better the outcome
  - Historically, it did not matter how early we diagnosed it since there was nothing to do about it other than correct the vision
- Keratoconus progresses much faster in younger individuals
  - Missing the opportunity to make a diagnosis at a yearly exam in a youth could be significant over the course of a year between exams

# How do we Detect KC Early?

- Screen those with a family history at a young age
  - Repeat screening as necessary to ensure early detection
- Test those with monocular vision issues (ghosting / diplopia / glare) as well as those with unusual changes to corneal astigmatism
- Screening / testing in these individuals should include corneal topography and pachymetry – corneal tomography if available

# OCULUS - PENTACAM

**Refractive**

Cornea Front		
Rf:	8.34 mm	K1: 40.5 D
Rr:	8.01 mm	K2: 42.1 D
Rm:	8.18 mm	Kav: 41.3 D
QS:	OK	Ax: 141.9°
		Astig: 1.7 D
Q-val:	0.37	Rper: 7.92 mm
		Rmin: 7.15 mm

Cornea Back		
Rf:	6.92 mm	K1: 5.8 D
Rr:	6.55 mm	K2: 6.1 D
Rm:	6.74 mm	Kav: 5.9 D
QS:	Model	Ax: 114.9°
		Astig: 0.3 D
Q-val:	0.28	Rper: 6.65 mm
		Rmin: 5.44 mm

Pachy		
	( $\mu\text{m}$ )	( $\mu\text{m}$ )
Pupil Center:	+ 504 $\mu\text{m}$	-0.22 +0.27
Pachy Apex:	497 $\mu\text{m}$	0.00 0.00
Thinnest Locat.:	475 $\mu\text{m}$	-0.68 -0.83
K Max. (Front):	47.2 D	0.75 -3.54

Cornea Volume		
	$\text{mm}^3$	KPD: +1.3 D
Chamber Volume:	1.35 $\text{mm}^3$	Angle: 34.2°
A. C. Depth (Int.):	2.70 mm	Pupil Dia: 3.01 mm
Enter IOP	IOP (cor)	
		Lens TH:

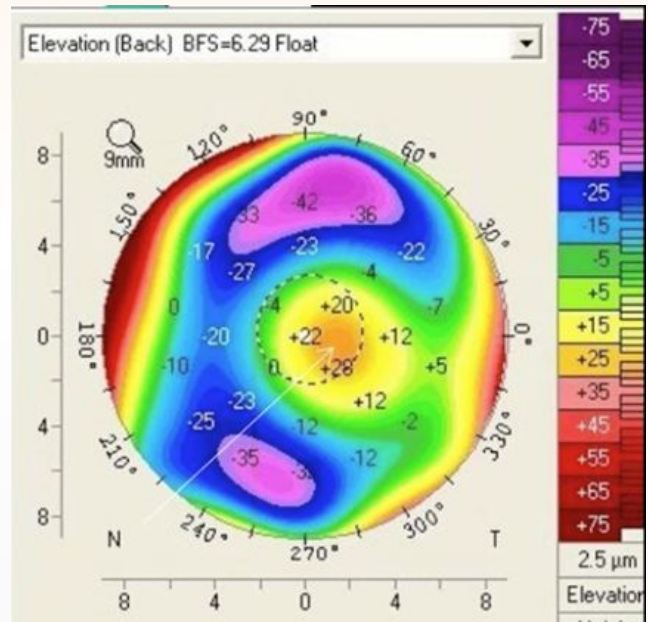
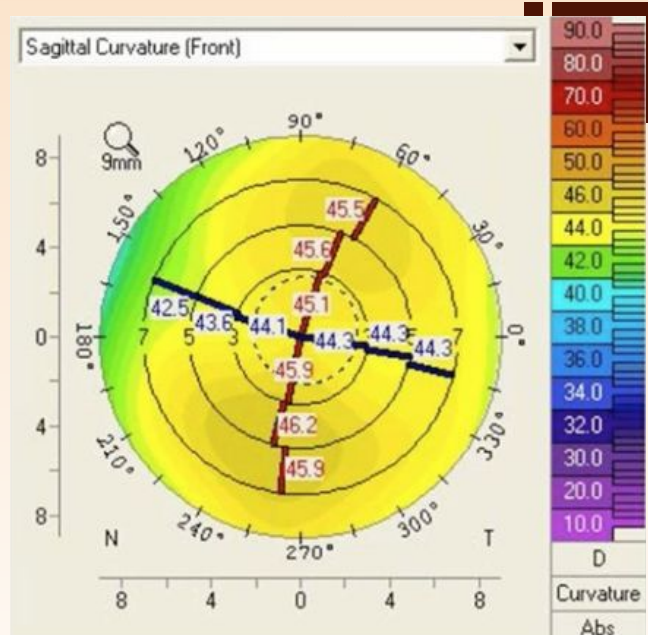
Sagittal Curvature (Front)  
OD

Elevation (Front)  
BFS=7.97 Float, Dia=8.00  
OD

Corneal Thickness  
OD

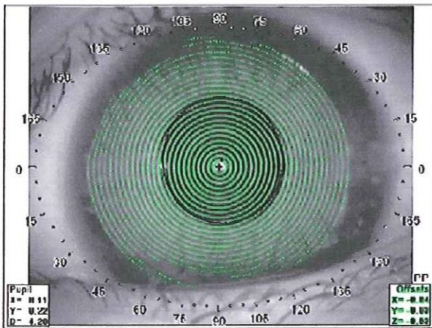
Elevation (Back)  
BFS=6.63 Float, Dia=8.00  
OD



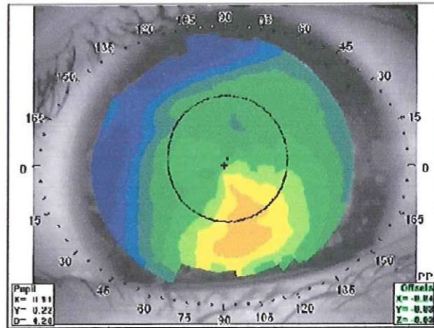


# Correctly Diagnosing KC – Is This KC?

Date: 3/3/2015 12:23:38 PM		Exam 4	
<b>Ks: 44.75 @ 131°</b>	<b>Kt: 44.20 @ 41°</b>	<b>Mink: 44.17 @ 48°</b>	<b>AveK: 44.48</b>
<b>SRI: 0.10</b>	<b>Es: 0.51 / Em: 0.30</b>	<b>PVA: 20/15-20/20</b>	<b>SAI: 0.50</b>
<b>Cyl: 0.55</b>			



Standard



Normalized

Diopters

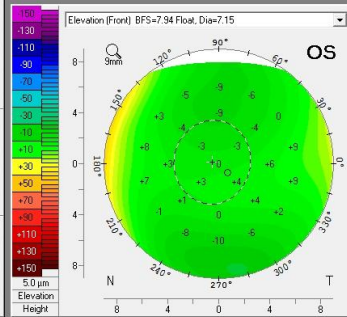
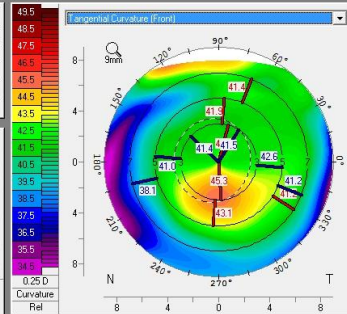
## OCULUS - PENTACAM 4 Maps Selectable

Last Name: Tseng	
First Name: Yung Fang	
ID: 184149	
Date of Birth: 06/26/1981	Eye: Left
Exam Date: 03/04/2019	Time: 16:50:22
Exam Info:	

Cornea Front	
R1:	8.07 mm K1: 41.8 D
R2:	7.80 mm K2: 43.3 D
Rm:	7.93 mm Km: 42.5 D
QS:	OK Axis: 175.7° Astig: 1.4 D
Q-val: -0.18	Rper: 8.08 mm Rmir: 7.54 mm

Cornea Back	
R1:	6.67 mm K1: -6.0 D
R2:	6.33 mm K2: -6.3 D
Rm:	6.50 mm Km: -6.2 D
QS:	OK Axis: 7.8° Astig: 0.3 D
Q-val: -0.14	Rper: 6.55 mm Rmir: 6.10 mm

Pupil Center:	+ 588 μm	x(mm): -0.26	y(mm): +0.06
Pachy Apex:	586 μm	0.00	0.00
Thinnest Local:	584 μm	+0.34	-0.34
K. Max. (Front):	44.7 D	0.00	-1.97
Cornea Volume:	64.3 mm <sup>3</sup>	∅ Cornea:	121 mm
Chamber Volume:	199 mm <sup>3</sup>	Angle:	42.7°
A. C. Depth (Int.):	3.31 mm	Pupil Dia:	3.14 mm
Enter IOP (IOP(Sum))	2.9 mmHg	Len Th:	



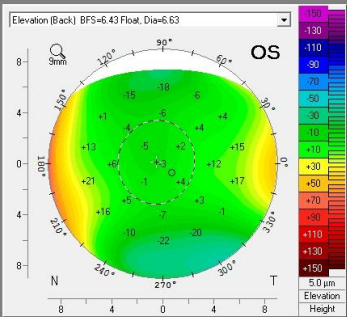
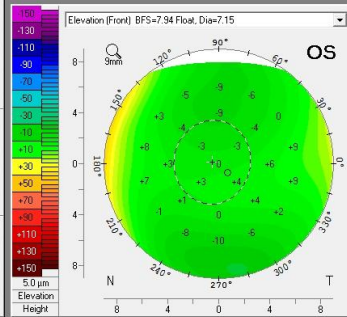
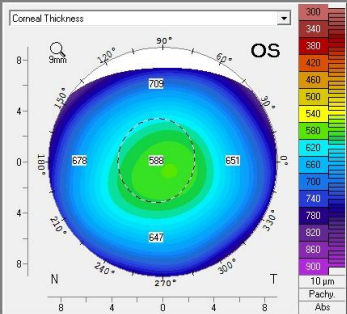
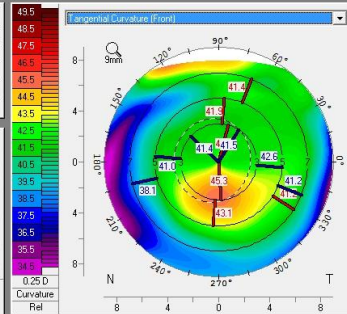
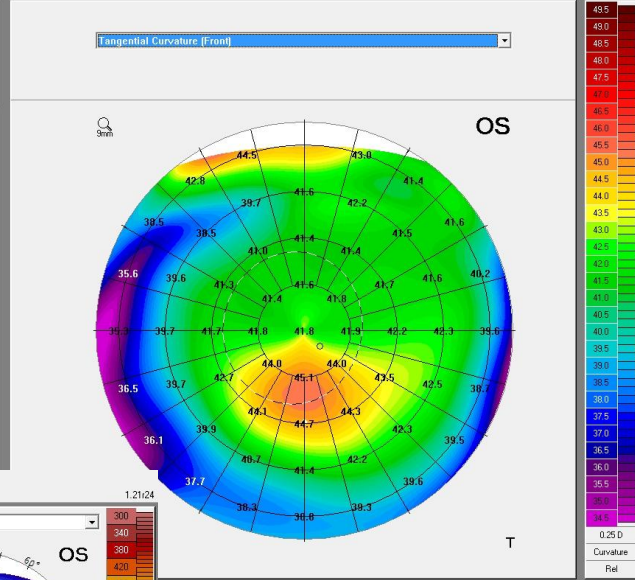
## OCULUS - PENTACAM 1 Large Color Map

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Date of Birth:	06/26/1981	Eye:	Left
Exam Date:	03/04/2019	Time:	16:50:22
Exam Info:			

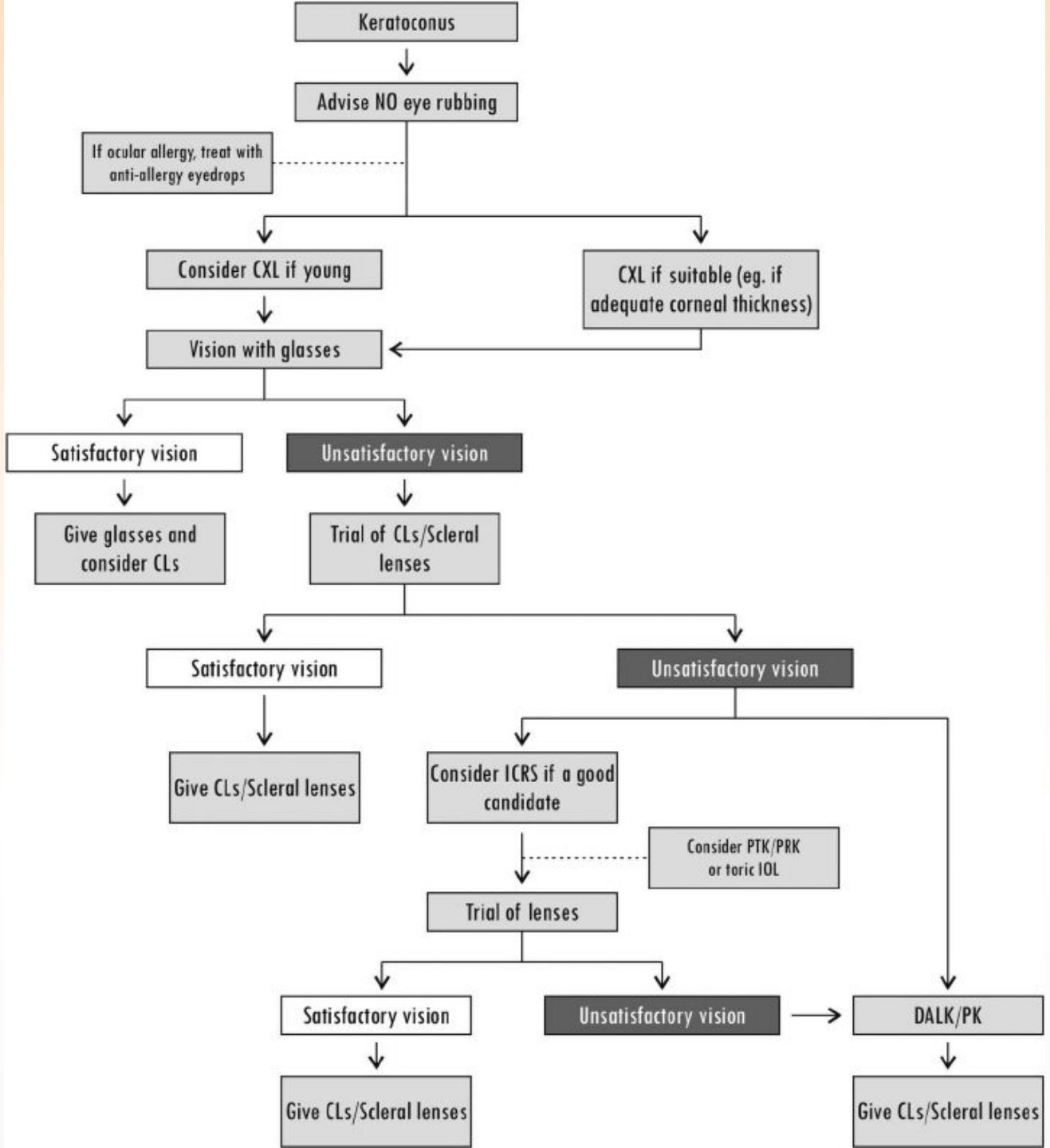
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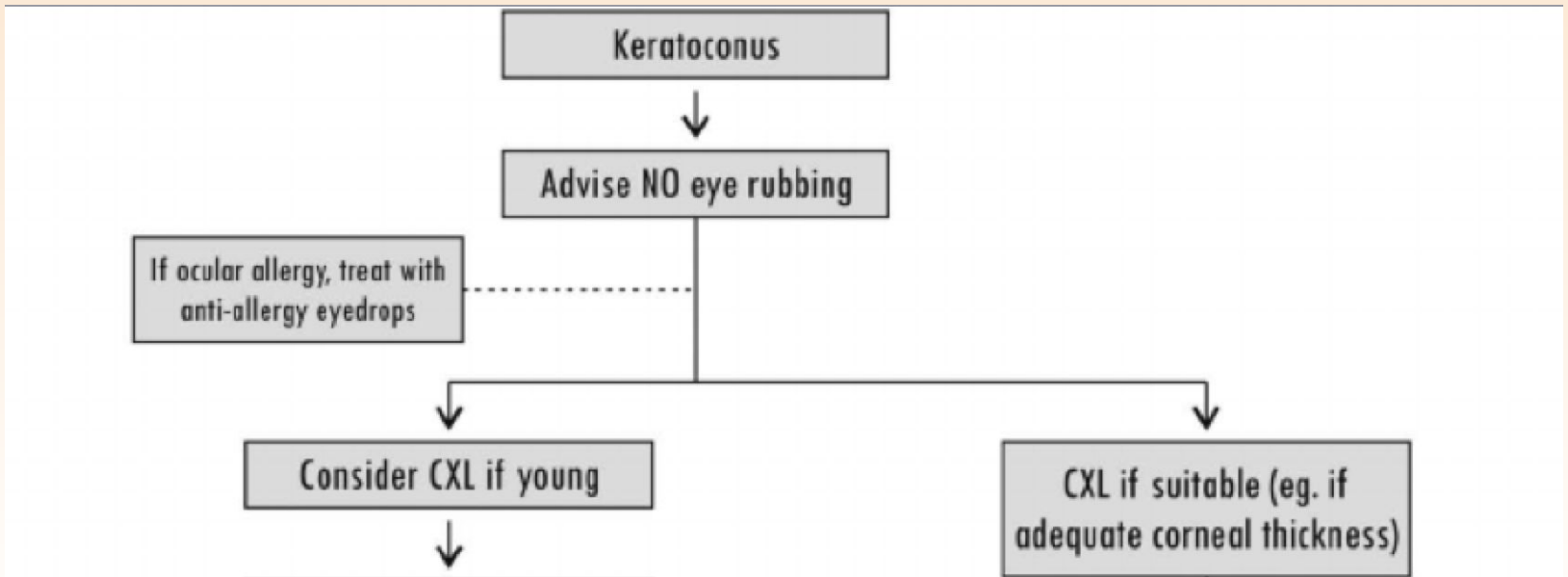
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Pachy:	x(mm)	y(mm)	
Pupil Center:	+ 588 μm	-0.26	+0.06
Pachy Apex:	586 μm	0.00	0.00
Thinnest Local:	584 μm	+0.34	-0.34



# KC Management Decision Tree





Managing Keratoconus

# Stop the Progression



# Control Eye Rubbing and Allergies

- Eye rubbing has been shown to directly correlate to development of and progression of keratoconus
- Make KC patients aware of this, so they can avoid and tell their immediate family members as well
- If they have a desire to rub, offer alternatives
  - Cool compresses
  - Anti-itch drops for acute episodes (Opcon, Naphcon, etc...)
  - Allergy drops for daily management of allergens and itch

# Offer Strongly Recommend Corneal Collagen Crosslinking (CXL)

- Depending on the age and status of the individual, Corneal Collagen Cross-linking MUST be discussed as an option for management
  - For those under the age of 30, this should be almost mandatory
  - 30 – 45 requires evaluation of the state of disease and recent progression
    - Still a good idea but urgency may be less
  - Over age 45, offer procedure but ok to monitor if condition is mild or beyond severe
- In severe cases, it may be too late

# FDA-Approved Cross-Linking Procedure Summary



1. Remove epithelium.



2. Soak cornea with Photrex<sup>®</sup>  
Viscous (riboflavin 5'-phosphate in  
20% dextran ophthalmic solution).

✓ 30 minutes



3. Check for flare.



4. Once flare is observed, measure  
corneal thickness.

✓ If corneal thickness is less  
than 400  $\mu\text{m}$ , instill 2 drops of  
Photrex (riboflavin  
5'-phosphate in ophthalmic  
solution) until the corneal  
thickness increases to at  
least 400  $\mu\text{m}$ .

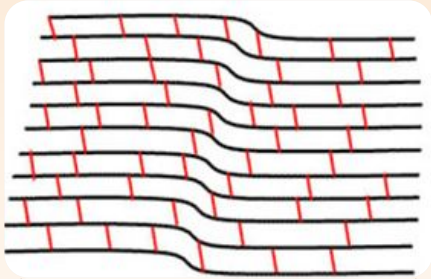


5. Irradiate for 30 minutes.

✓ Continue applying Photrex  
Viscous (riboflavin  
5'-phosphate in 20% dextran  
ophthalmic solution) during  
irradiation.

# Keys to Patient Counseling: Discuss Treatment Goals

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**Aim of cross-linking is to halt or slow disease progression**



**Cross-Linking is not a refractive procedure**



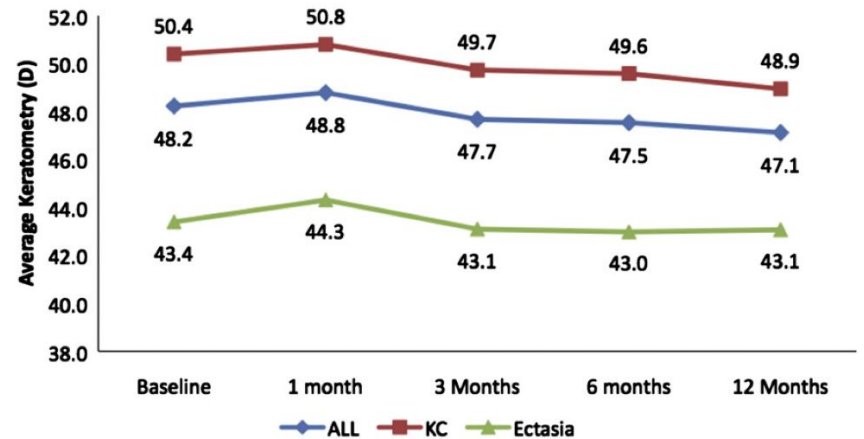
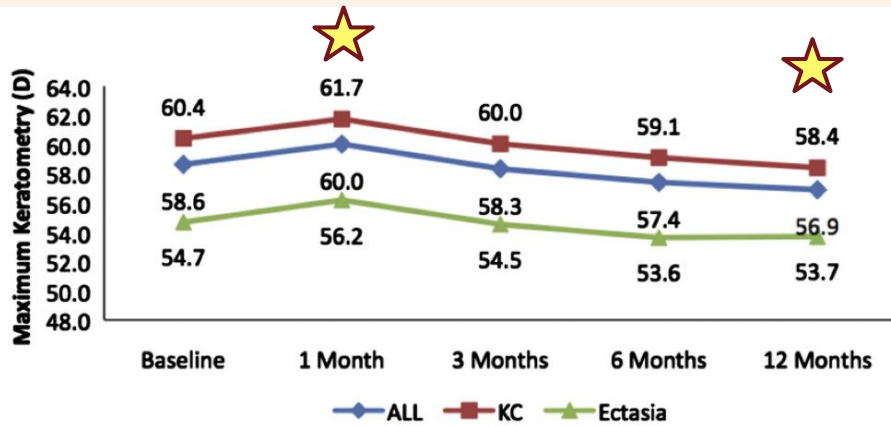
**Post-op evaluation for visual correction will be necessary**



# CXL: Best Candidates

- Evidence of progression of ectasia
- Absence of significant or full thickness scarring
- Minimum corneal thickness of 300 microns pre-tx
  - Preferably 400, but between 300 and 400 can swell the cornea
- Vision and function worth saving

# CXL – US Phase 3 FDA Studies



## Find an Ophthalmologist in Your Area Performing FDA-Approved Cross-Linking

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### Find a Cross-Linking Expert

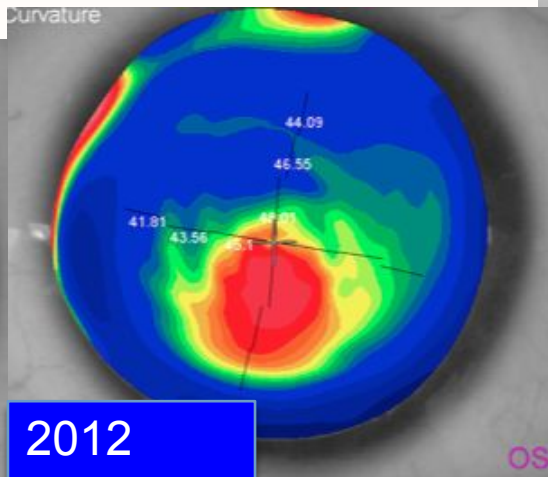
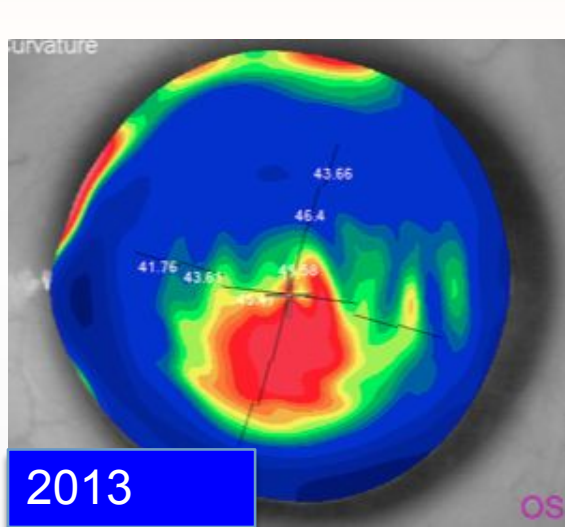
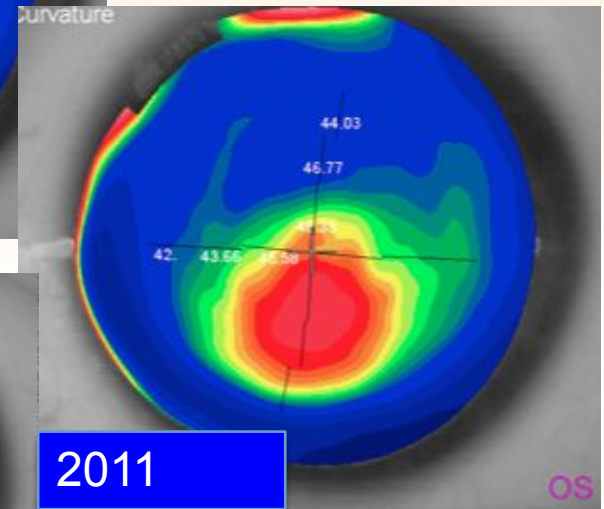
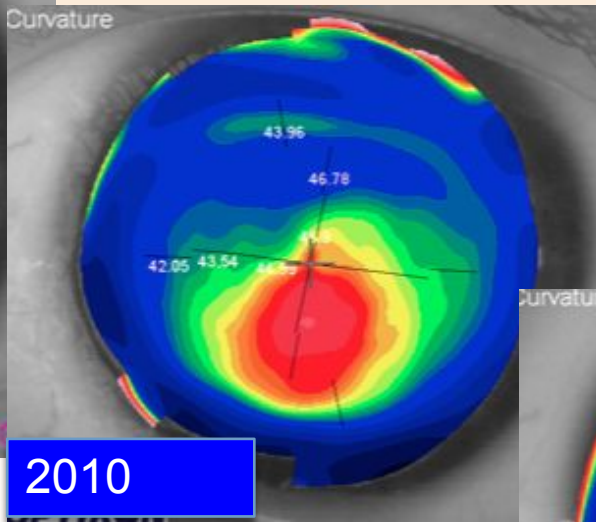
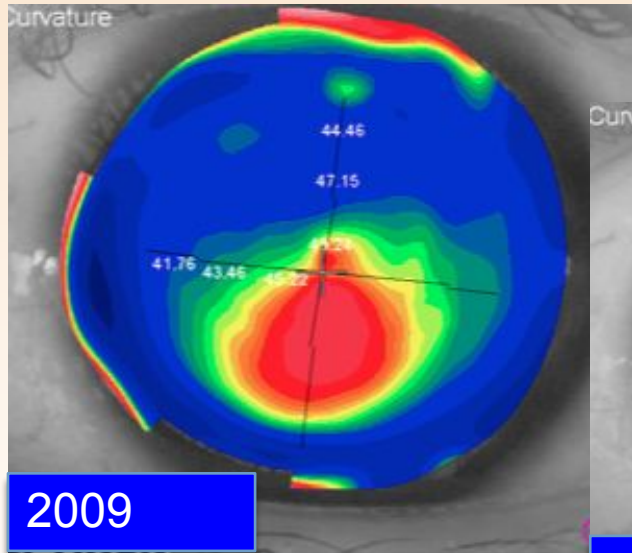
Search the directory below to find a cross-linking expert who is familiar with treating progressive keratoconus.

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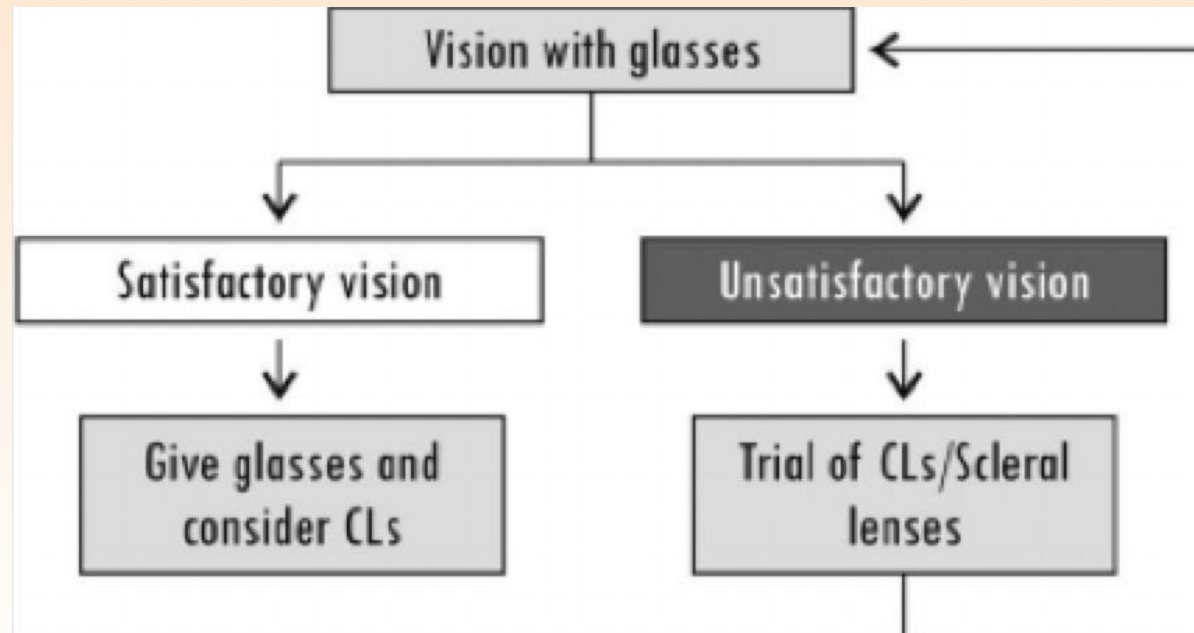
Visit the **Find a Physician Locator** on the Living with Keratoconus website

**[LivingwithKC.com](http://LivingwithKC.com)**









Managing Keratoconus

# Correct the Vision: Corrective Lenses

# Spectacles

- Should not be overlooked in mild cases
- Patient should not be told that they **MUST** use contact lenses just because they have KC
- When vision is functional with spectacles and patients are satisfied with their BCSVA, glasses may offer advantages over contact lenses
  - Less “trauma” to the cornea
  - Less apt to lead to eye rubbing
  - More protection from harmful wavelengths of UV



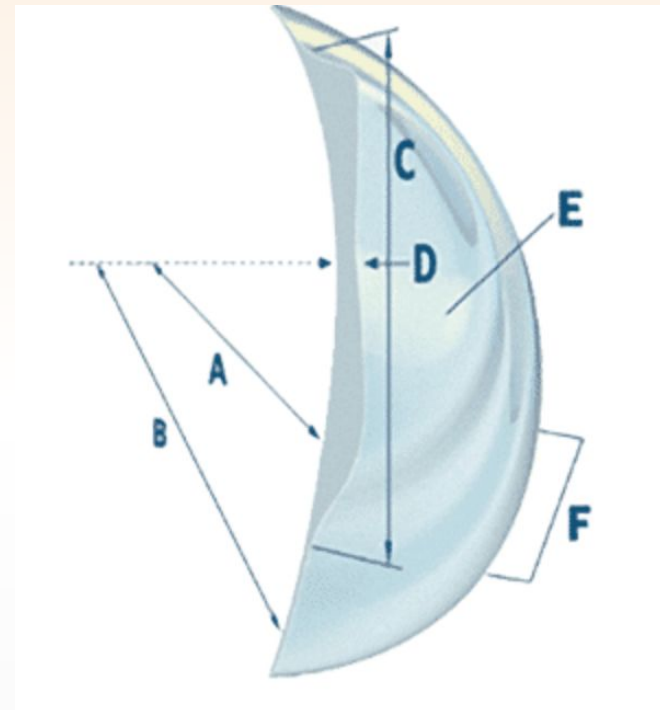
# Soft Lenses

- Milder forms of KC can sometimes be managed with regular soft toric disposable lenses
- Biofinity torics are a nice option – a little firmer lens masks some cylinder



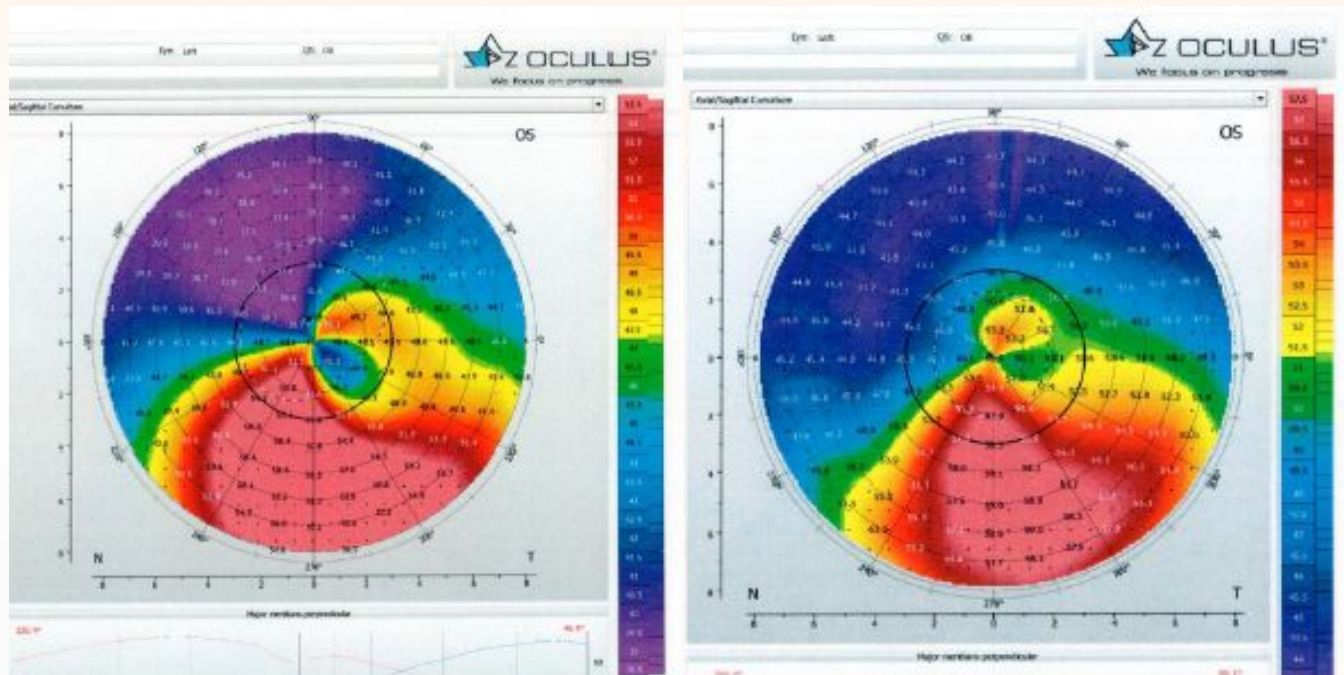
# Soft Lenses for KC

- Designed with a thicker optic zone to mask irregularity
- Multiple BC and Diameter options to optimize fit
- Can add toric optics
- Can be more comfortable than GPs and easier on the epithelium
- Can cause edema due to thickness if poorly fit and visual acuity is typically not as good as GPs



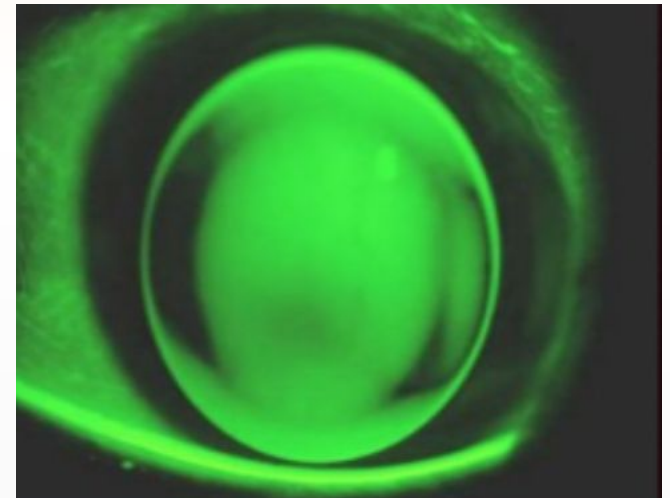
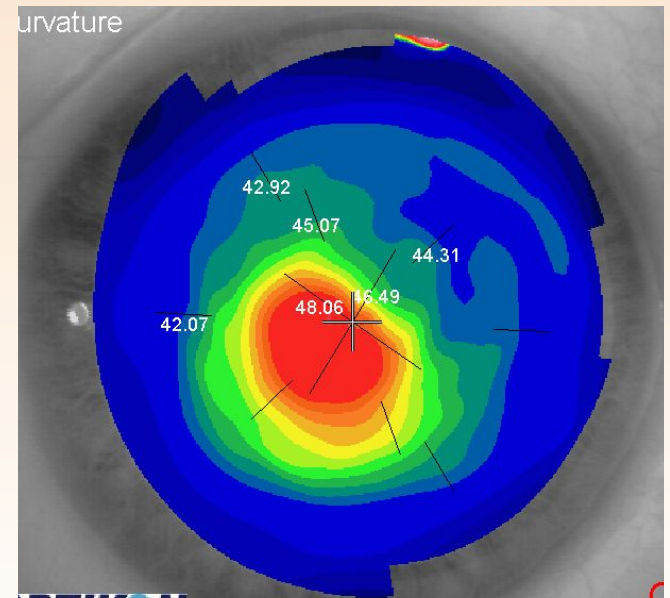
# Soft Lenses for KC

- Work best when refraction yields a “Decent Outcome” or central 3 mm is not overly irregular

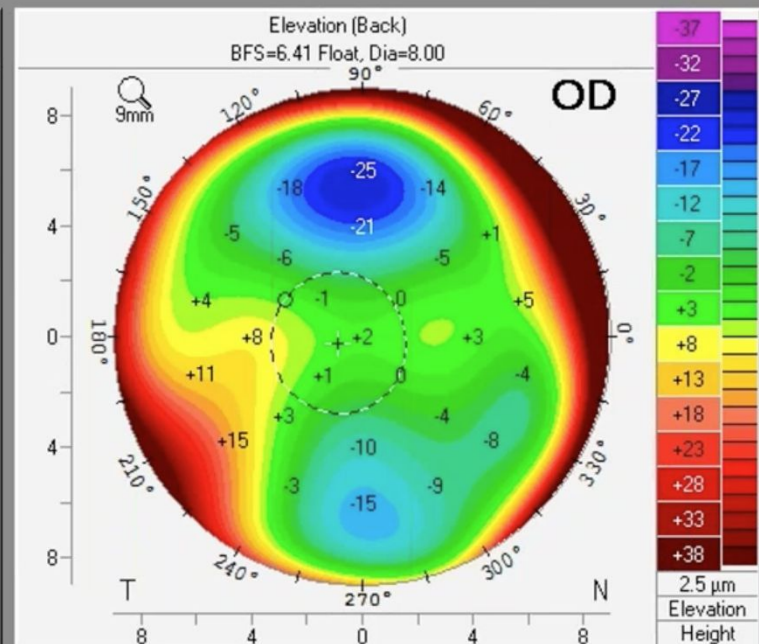
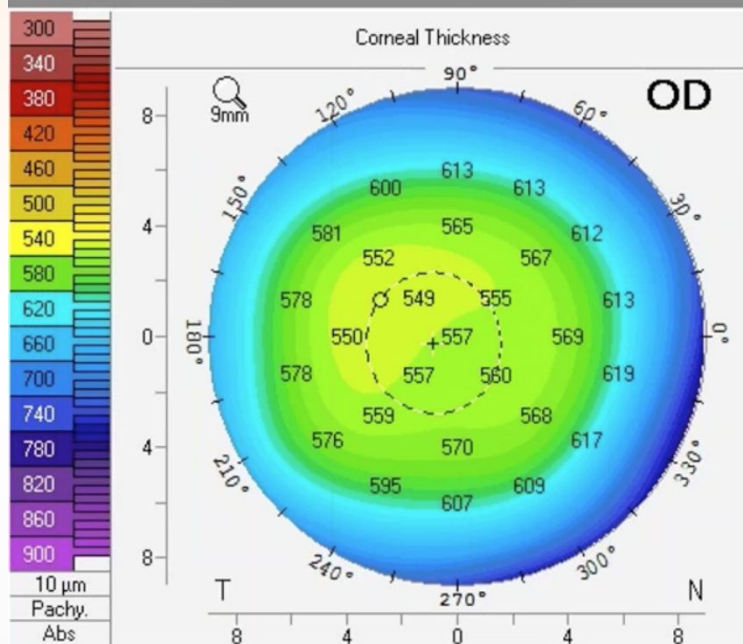
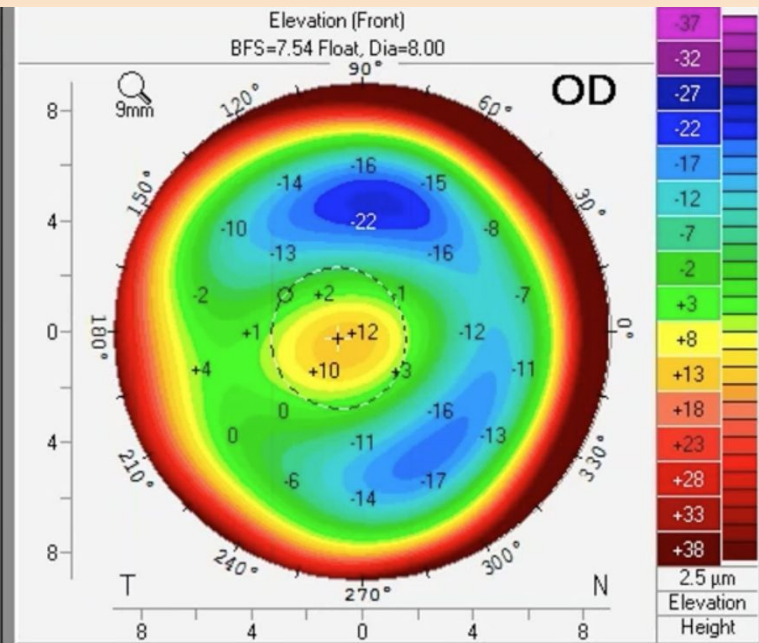
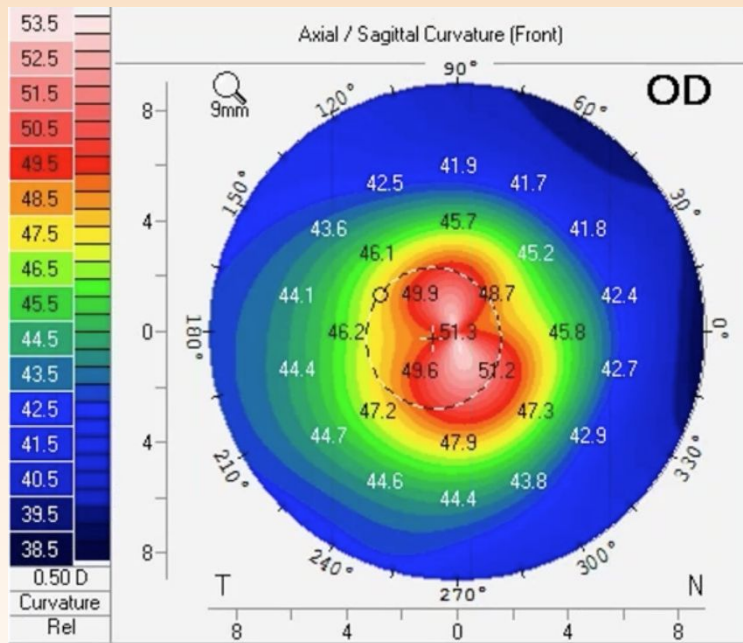


# Corneal GP Lenses

- Mainstay of KC fitting for decades
- Steep base curves with peripheral curves that flatten more rapidly than lenses for normal corneas
- Usually smaller diameters but increasingly getting larger
- Useful for CORRECTING ACUITY, NOT TO IMPACT PROGRESSION OF THE DISEASE
  - You cannot think this way any longer, the science has disproven it
- **Work best on more central cones**

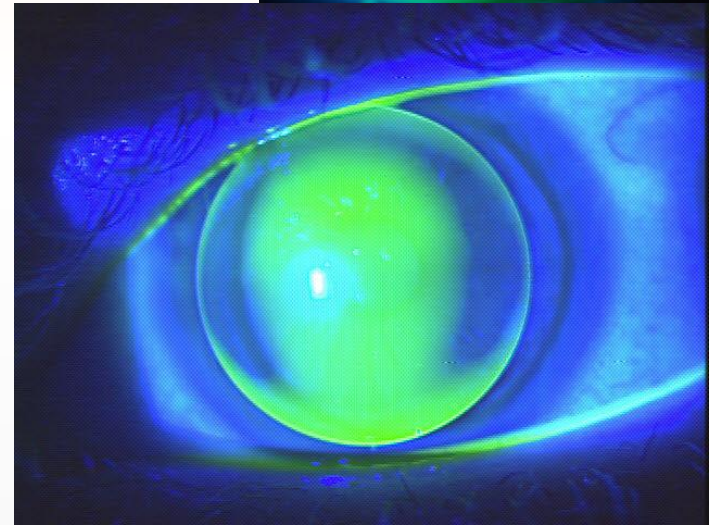
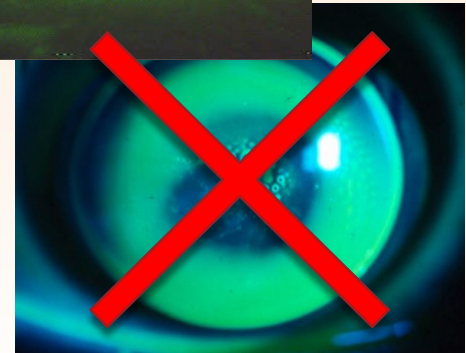
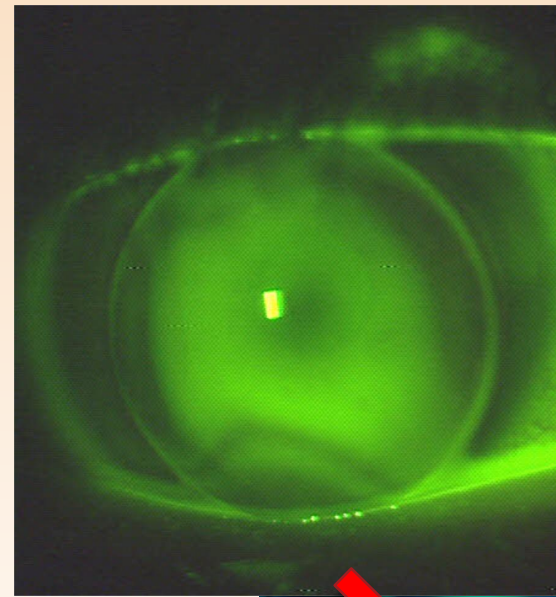






# Corneal GP Lenses

- Apical Bearing versus Apical Touch versus Apical Clearance
  - CLEK study demonstrated that corneal bearing causes scarring – not recommended
  - Apical clearance is safest to prevent scarring, but some feel VA is reduced with this strategy
  - Where the apex is located can dictate the type of lens to fit – smaller lenses are better on central steep areas, for decentered apex, a larger lens usually is better



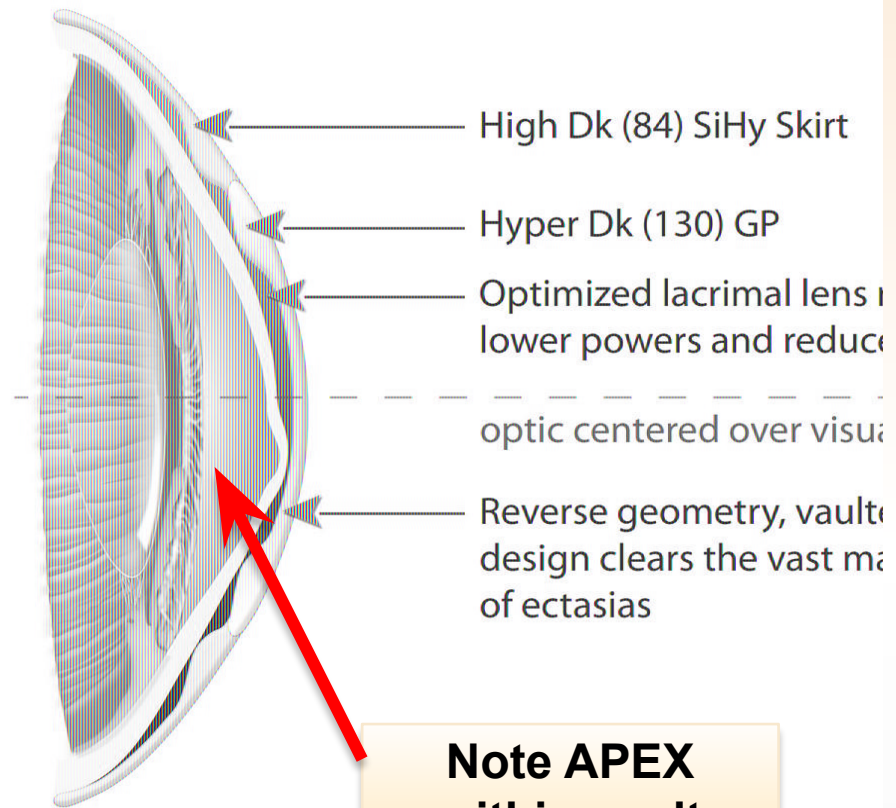


# Hybrid Contact Lenses

- GP lens with bonded soft “skirt”
- Provides GP optics with edge awareness more like a soft



## UltraHealth™ Vaulted Lens



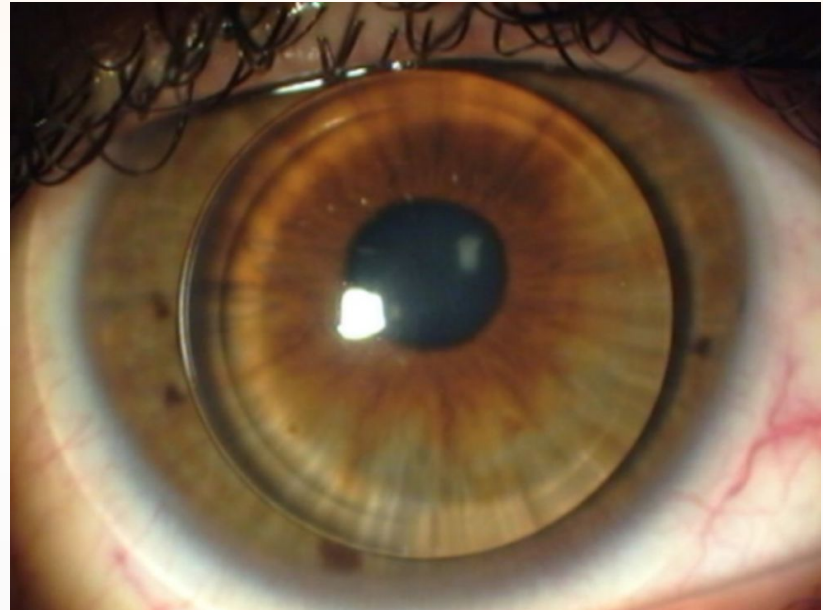
**Note APEX  
within vault  
zone**

# Hybrid Contact Lenses

- Can be more comfortable initially than a GP while providing GP optic
- Soft skirt provides good centration and stability
- Can tighten with wear, important to get fit correct!
- If the soft / GP junction crosses the apex, hybrids can cause epithelial breakdown and scarring / discomfort
- **IDEAL FOR CENTRAL CONES WHEN STABILITY AND COMFORT ARE A PRIORITY**

# Piggyback Contact Lenses

- Can be used to aid in adaptation for a period of time
- Can be used to protect the epithelium from GP related erosions

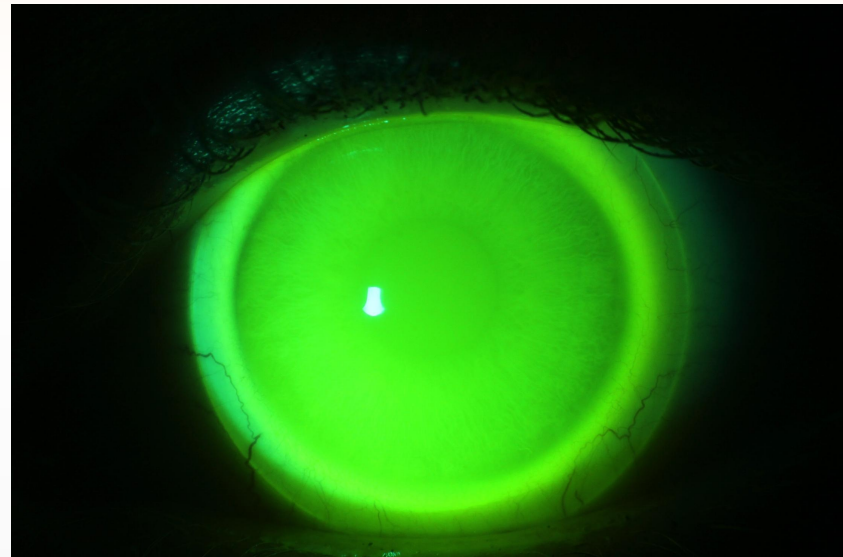


# Piggybacking Contact Lenses

- Soft lens will age more rapidly due to GP lens use over it
- Solutions need to be GP and soft lens compatible
- More expensive, more hassle, and taxing to the tear layer
- Typically best to fit the GP to the cornea then let the SCL be neutral from a fit and VA standpoint
  
- Power effect of soft lens = 20%
  - $-.50\text{ D lens} = -.10\text{ D power effect}$
  - Really do not need to factor in unless using higher power lens for some reason

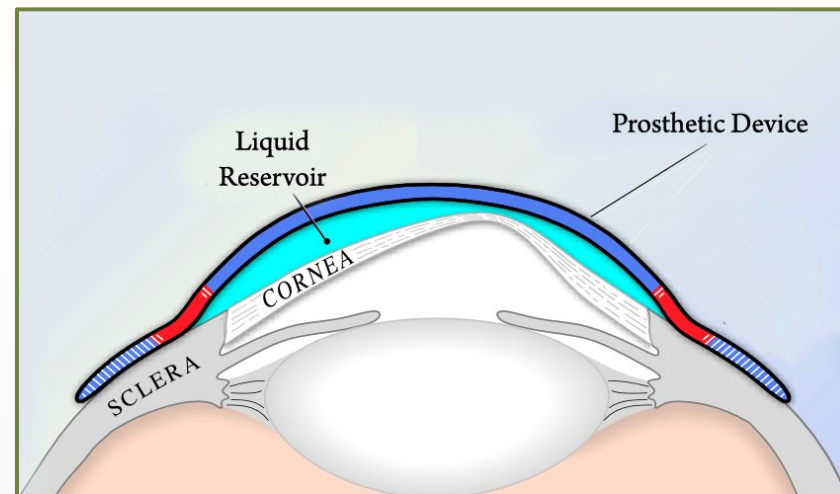
# Scleral Contact Lenses

- Provide GP vision correction
- Provide stable, comfortable lens wear
- Do not bear on the epithelium so no CL related scarring



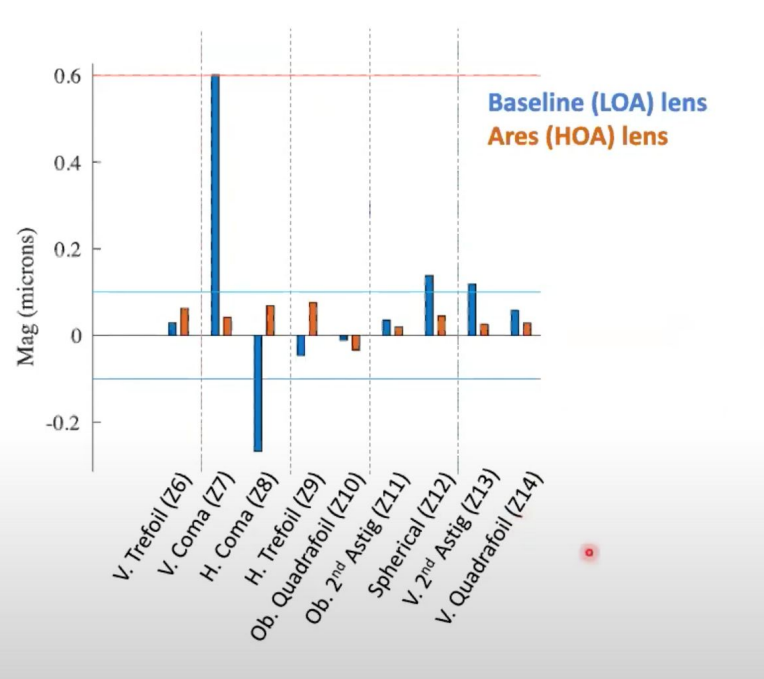
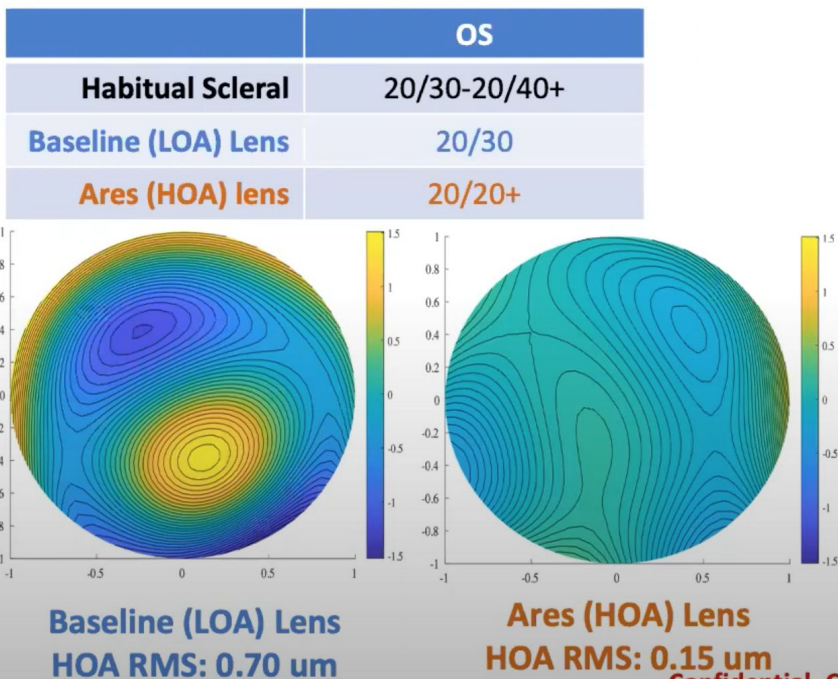
# Scleral Contact Lenses

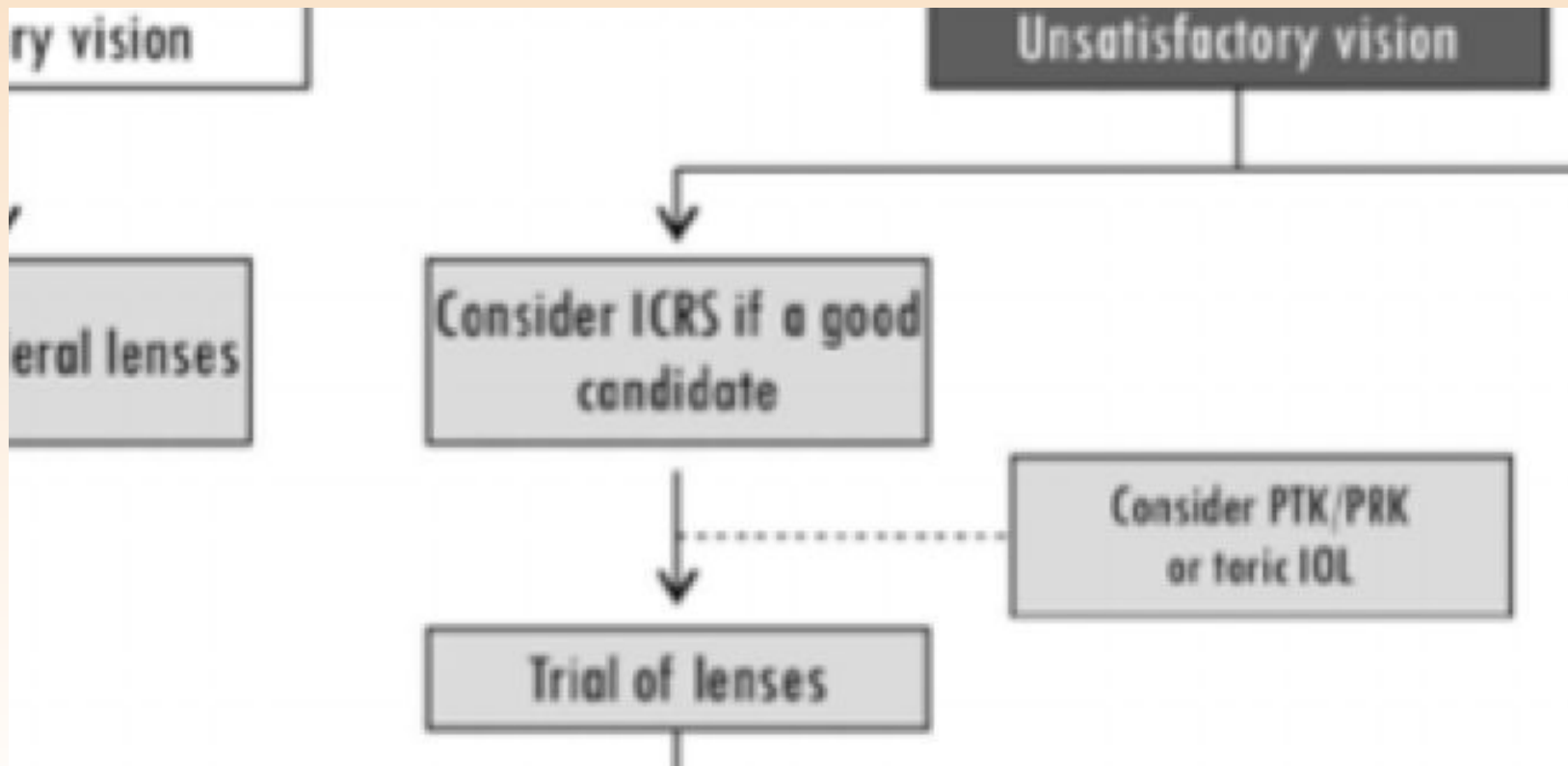
- Fit with full corneal clearance as well as limbal clearance
- Align lens to the scleral curve to achieve a lens with little or no movement
- **Best suited to those with large or highly off center cones, or those that have failed other options due to discomfort, or those that absolutely require a stable fit due to occupation or avocation**



# Scleral Contact Lenses

- Improving vision through correction of Higher order aberrations with scleral lenses
  - Most KCN patients have significant HOA even with their GP lenses
  - Leads to reduced BCVA
  - Correcting HOA now is possible with scleral lenses





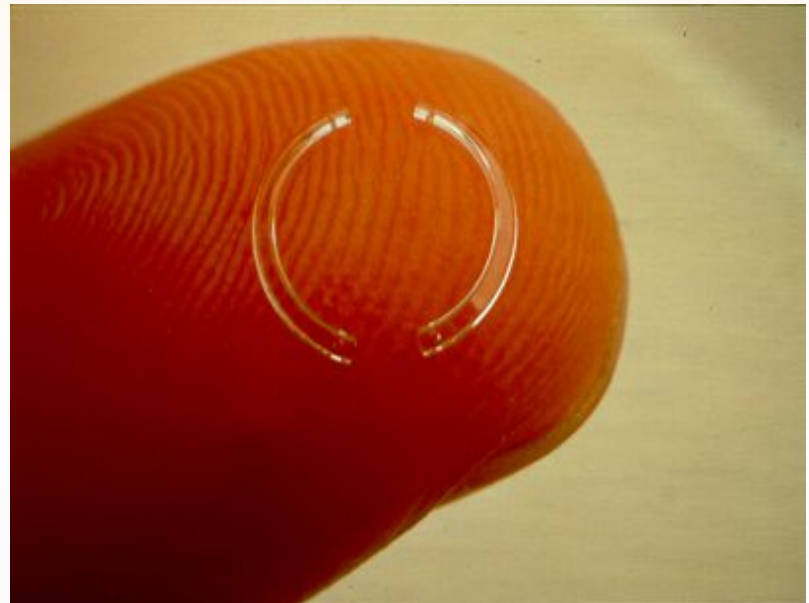
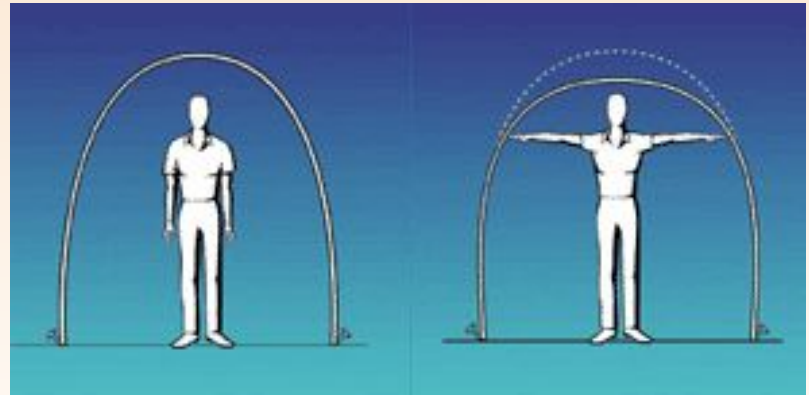
Corneal Rings

**When Lenses Don't Correct the Vision Adequately**



# Corneal Rings

- **Concept:**
  - Add support to thinned area of cornea to prevent forward bulging
- INTACS provide a ***structural supporting element*** to the thinned and abnormal keratoconic tissue



# Corneal Rings

- *J Cataract Refract Surg. 2008, Ertan, et al.*
- Retrospective study of Intacs implanted in 306 keratoconic eyes of 255 patients
- 10 month mean results:

UCVA	BSCVA	Spherical RE	Cylindrical RE	Keratometry
75.7% of eyes improved	71.6% of eyes improved	-6.04 D to -3.09 D <b>(2.94 D)</b>	-4.11 D to -3.82 D <b>(0.29 D)</b>	50.07 D to 47.9 D <b>(2.17 D)</b>

- Intacs treatment with a femtosecond laser was effective for management of keratoconus of all stages
- Improvement in UCVA may be less in severe keratoconus than in moderate keratoconus

# Corneal Rings

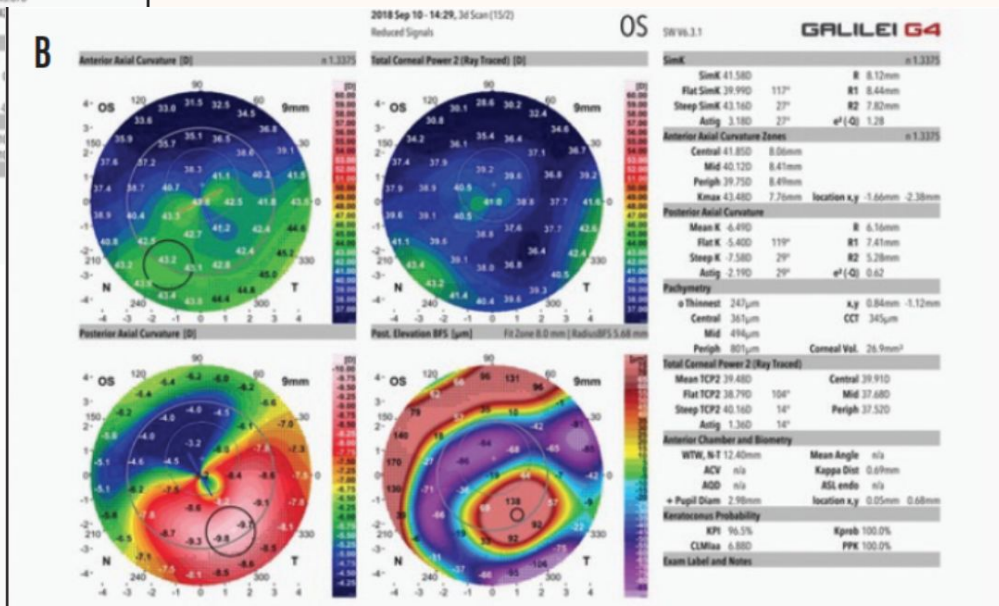
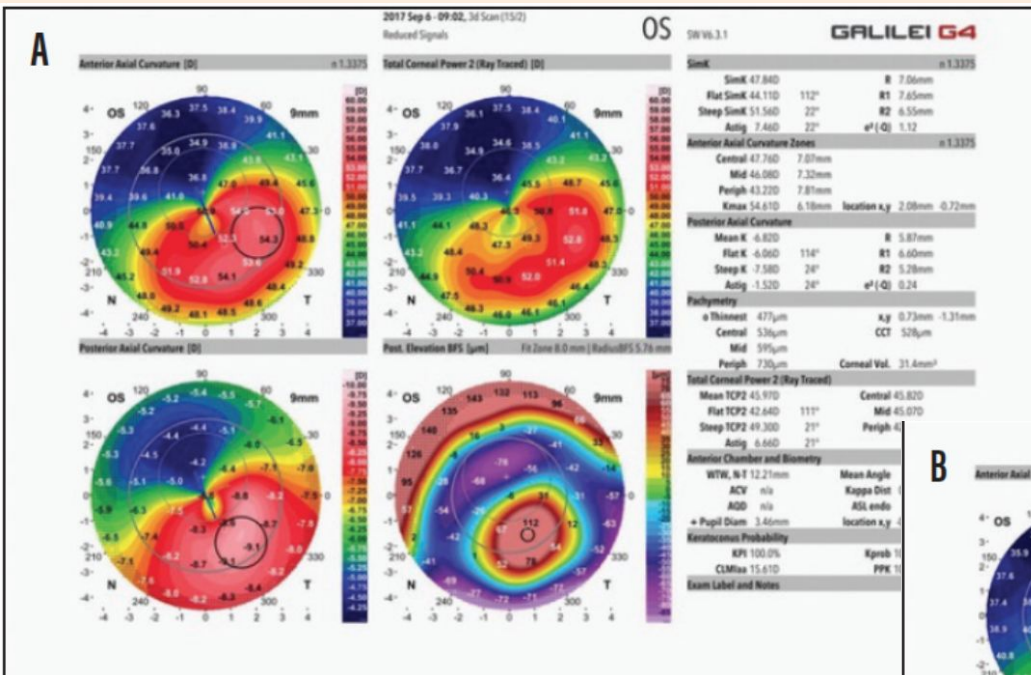
- When is the best time to use them?
  - When best corrected acuity with GP's is worse than expected by looking at the contact lens fit and corneal clarity
  - When manifest refraction reveals low to moderate myopia
- **No reason to suggest that intacs SLOW or STOP progression**
  - Only turn back the clock on the condition
  - Corneal Rings can improve the corneal regularity, cross linking can lock it in – this is why they are often done in conjunction

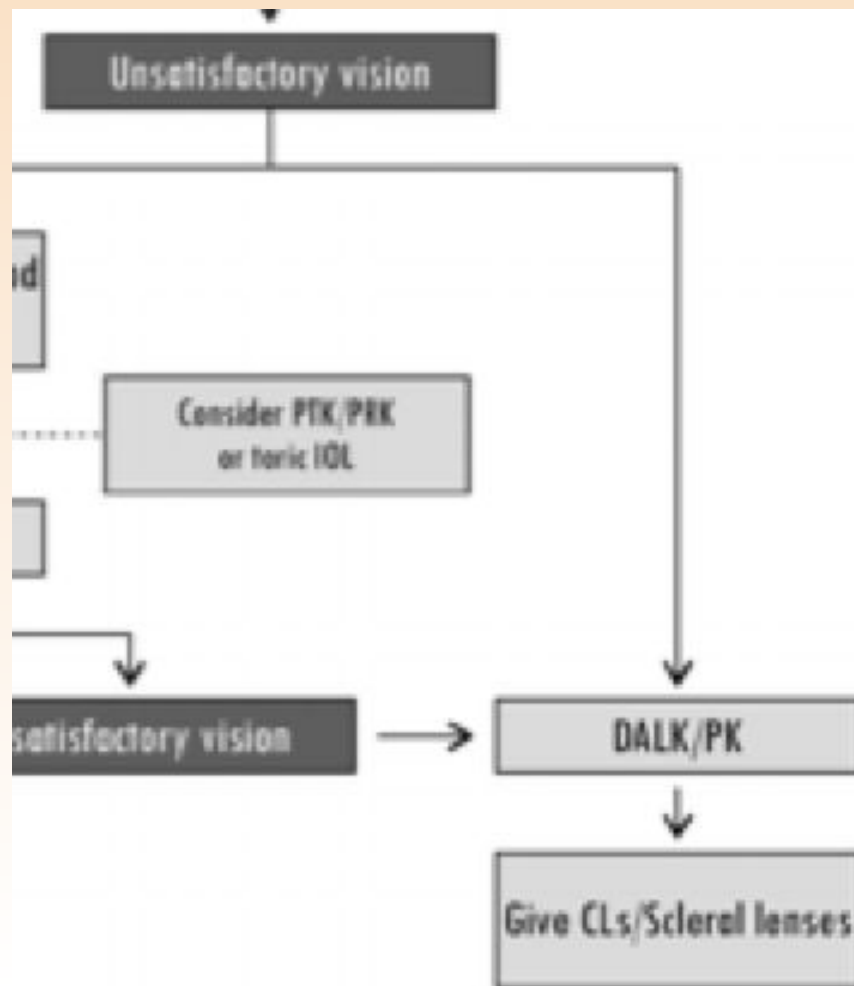
# CXL with Topo Guided PRK (TG-PRK)

- Use of custom PRK to smooth corneal shape irregularities after CXL (not to treat refractive error - not to remove more than 50 microns of tissue)
- Sequential
  - TG PRK is done 1-2 yrs post CXL
- Simultaneous
  - TG PRK is done first and immediately followed by CXL
- Kanellouplous found better outcomes with Simultaneous Treatment
- Nattis in 2019 published data on Sequential TG-PRK at 30 months to smooth AND correct refractive error and found good outcomes (removing MORE than 50 microns)

# CXL with Topo Guided PRK (TG-PRK)

- Is it as safe in the long run?





Corneal Transplants

## When Lenses Don't Correct the Vision Adequately

# Penetrating Keratoplasty

- Transplant of the Entire Corneal Thickness
  - Usually 6-9 mm area depending on corneal pathology
- Provides a CLEAR cornea layer
- Often the new cornea is IRREGULAR
- Risk of glaucoma, cataracts, rejection, etc...

# Penetrating Keratoplasty

- Traditionally done with Trefphine
- Good: Creates smooth cut
- Bad: Minimal surface area between graft and host means less stability and strength and more unevenness to the tissue
  - Sutures may need to be left in a LONG time



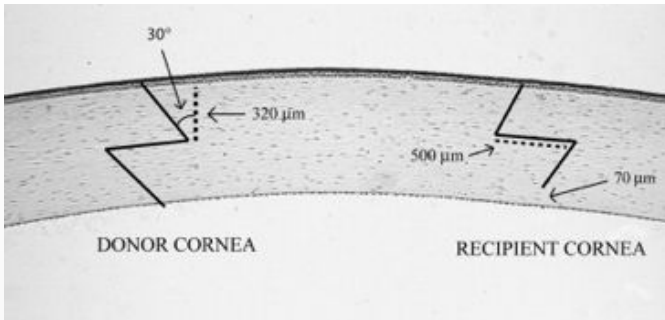
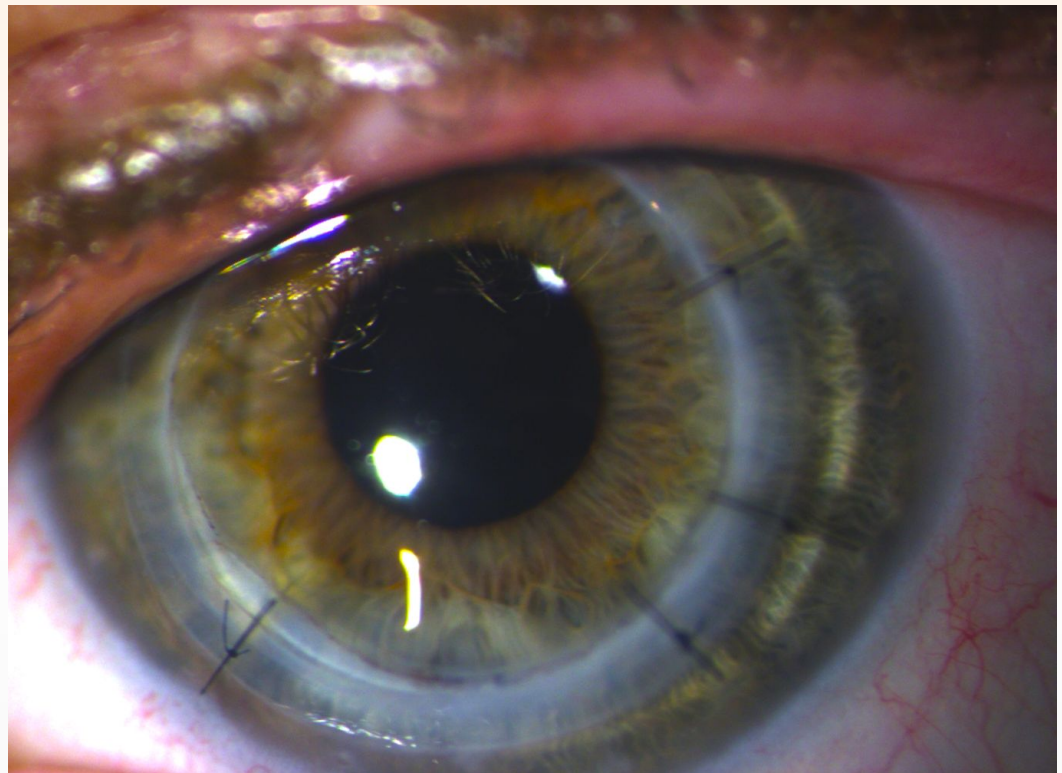
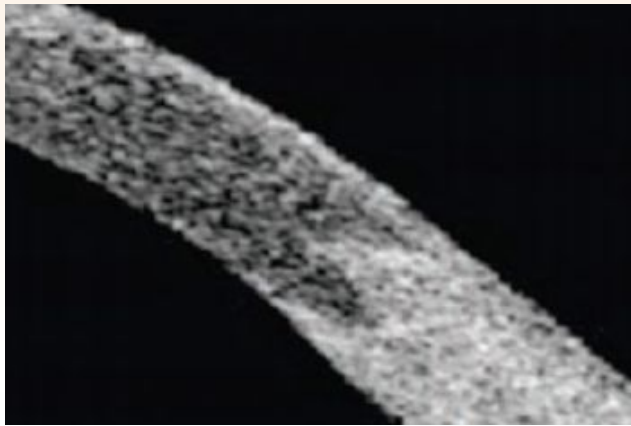


# IEK (Intralase Enabled Keratoplasty)

- Good
  - Suturing may not need to be as precise
  - Sutures do not need to be as tight (less distortion to the cornea)
  - Better front-to-back alignment means more consistent surface
    - Fewer issues with sunken or bulging grafts
  - Broader surface area between graft and host means sutures can come out sooner and contact lenses can be fit earlier
- Bad
  - Cost and availability of lasers

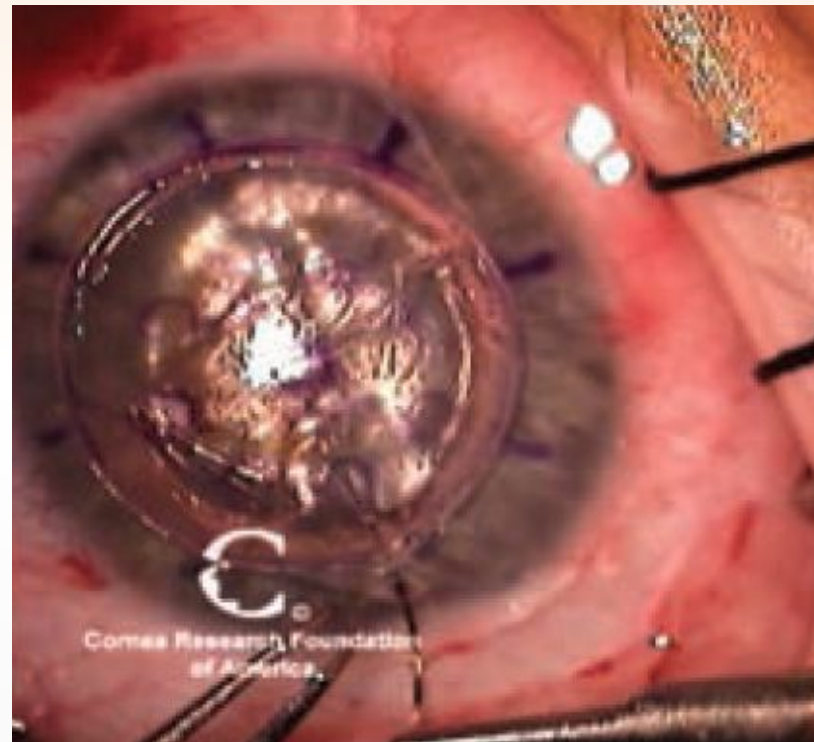


# IEK (Intralase Enabled Keratoplasty)



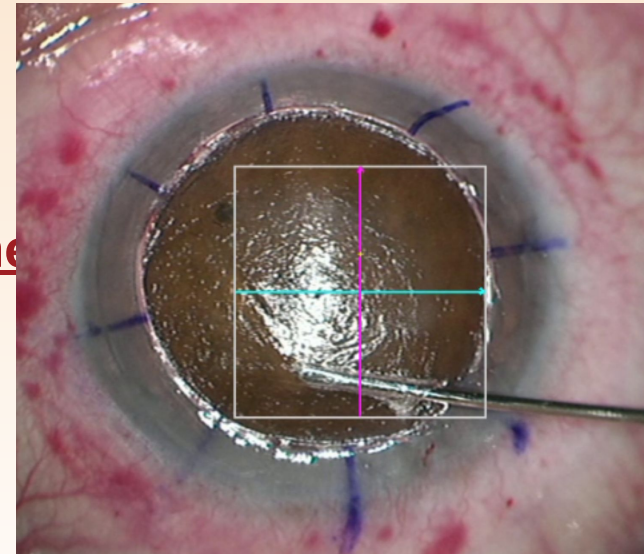
# Deep Anterior Lamellar Keratoplasty (DALK)

- Transplants the anterior 95% of the cornea, leaving Endothelium, Descemet's Membrane and sometimes a sliver of stroma behind
- Historically advantageous when quality corneal tissue is not as available



# DALK vs PK

- DALK has been shown to have similar best-corrected visual acuity outcomes compared with PKP when baring of Descemet's membrane occurs surgically
  - When graft is too shallow, VA can be reduced
- DALK spares transplantation of the endothelium, which eliminates the possibility of endothelial rejection
  - Epithelial, stromal, and mixed epithelial and stromal graft rejection, however, can occur at a rate of approximately 8 to 10%
  - Less than 5% of grafts fail secondary to rejection
  - The risk of graft loss following blunt trauma is also markedly reduced as compared to PK



# Other Surgical Interventions

- ICL
  - Can be used to reduce myopia and reduce dependence on corrective lenses
- IOL
  - Can be used to improve VA when cataracts develop or to reduce refractive error
- Toric IOL
  - Use with caution and clear understanding of how this makes contact lens fitting later more difficult

# Managing Keratoconus in 2023

- Detect early!
- Manage eye rubbing tendencies
- Refer for cross linking when appropriate
- Manage vision with proper corrective lenses depending on the stage of the condition
- Do no harm to the cornea with contact lenses
- Consider Corneal Rings when BCVA is reduced
- When all other options are exhausted and cornea is scarred, consider corneal transplant

# Resources

- IKA: International Keratoconus Academy
- [LivingwithKC.com](http://LivingwithKC.com)

jajedlic@indiana.edu