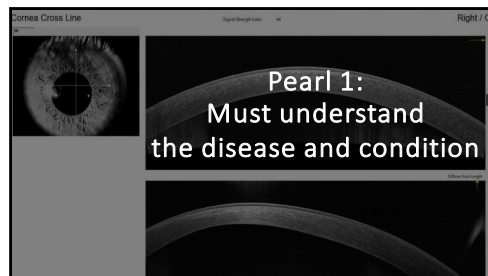


1

Disclosures

- Acculens, BostonSight, Aurion BioTech – Honoraria
- Glaukos – Consultant and speaker
- Oyster Point – Speakers Bureau
- Tarsus – Consultant
- All relevant financial relationships listed have been mitigated

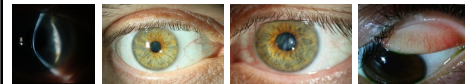
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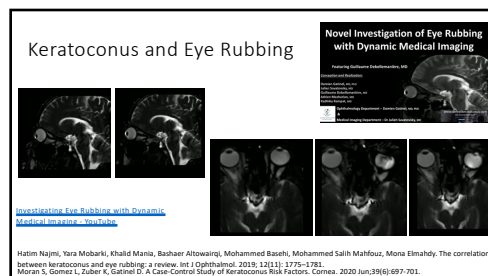
3

Keratoconus Overview

- Corneal Disorder
 - Central thinning & bulging
 - Bilateral
 - Asymmetric
 - Progressive
 - Non-infectious
 - Non-inflammatory
- Onset: Teens/puberty
- Prevalence and Incidence
 - Varies with geography, ethnicity, study
 - Prevalence 0.05 (US, 1986) to 4.79% (Saudi Arabia, 2018)
 - Better education and equipment, CK
- Risk Factors
 - Age
 - Family History
 - Eye rubbing



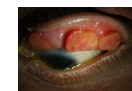
4



5

Keratoconus and Atopy

- Case-control study (1), (n=120) cases from 1985-1999
 - Atopy was defined based on the UK working group 1994 definition
 - History of **itchy skin** plus three or more of the following:
 - Involvement of skin creases
 - Personal asthma or hay fever (or Hx atopic disease in 1st degree relative)
 - General dry skin in the past year
 - Visible flexural eczema
 - Onset under the age of 2
 - Found an association between keratoconus and atopy
 - as well as eye rubbing and family history of keratoconus
 - Study supports hypothesis; most significant cause of keratoconus is eye rubbing
 - **Atopy may contribute to keratoconus but probably via eye rubbing associated atopic itching**

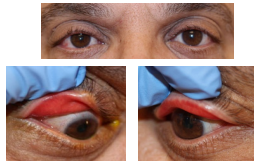


Rawazeer AM, Hodge WG, Lorimer B. Atopy and keratoconus: A multivariate analysis. Br J Ophthalmol. 2000;84:834-6.

6

Keratoconus and Floppy Eyelid Syndrome

- Loose and rubbery eyelids
- Easily evert
- Irritation upon awakening
- Associated with sleep apnea
 - Ask about snoring
 - Blocked airways, breathing stops
 - Refer for sleep studies
 - Treatment with CPAP

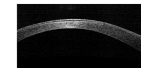


Razvan Cristescu Teodor, Florin Dumitru Mihaltan. Eyelid laxity and sleep apnea syndrome: a review. Rom J Ophthalmol. 2019 Jan-Mar; 63(1): 2-9.

7

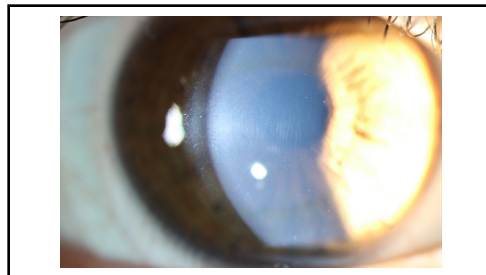
Corneal tissue is different in KC

- Cornea: connective tissue with cells and stromal extracellular matrix (ECM) that relies on the synergistic cooperation of many components
 - Needed to precisely transmit and refract visual information (1)
- ECM consists of organized lamellae composed of tightly distributed fibrils
- Collagen lamellae are altered in keratoconus →
 - Abnormalities in corneal shape
- KC is a progressive disease relative to defects in the corneal stroma
 - Development related to environment and genetics



(1) Zhou HY, Cao Y, Wu J, Zhang WS. [Role of corneal collagen fibrils in corneal disorders and related pathological conditions](#). Int J Ophthalmol. 2017 May 18;10(5):803-811. PMID: 28546981

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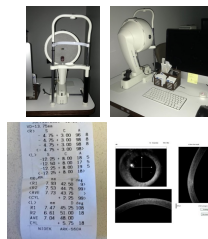
Pearl 2: Must be adequately equipped
Must "Go All In"



10

Equipment

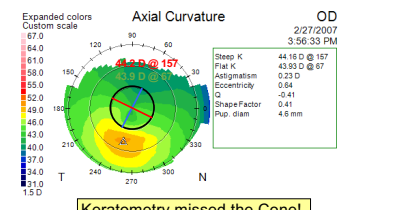
- **Scheimpflug Tomographer**
 - 2-D imaging
 - Maps front and back corneal surface, thickness
- **Autorefractor (great screener)**
 - Look for asymmetry in Ks between eyes
 - Unusual RE
- **Corneal Topographer (not detailed enough)**
 - 2-D imaging
 - Placido disc based
 - Maps surface curvature
- **Need Basics: Slit Lamp and Phoropter**
- **Optional:**
 - Keratometer
 - Anterior Segment Optical Coherence Tomography (AS-OCT)
 - Described in 1994 (Izatt JA), initially for retina
 - High resolution cross-sectional imaging



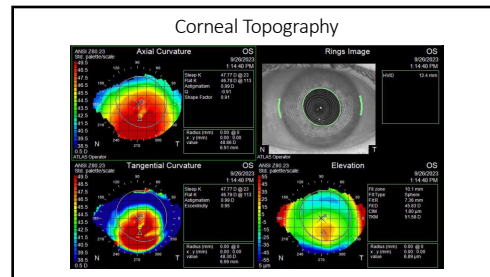
Izatt JA, Hee MR, Swanson EA, et al. Micrometer-scale resolution imaging of the anterior eye in vivo with optical coherence tomography. Arch Ophthalmol. 1994;112:1584-1589.

11

Clinical Advantages: Topography vs. Keratometry



12



13

Corneal Tomographer

- Best for **early** detection of keratoconus
- Comparison, normative data features, other:
 - Corneal optical densitometry
 - Zernike Analysis
 - Show 2/4 exams, comparison maps
 - Belin/Umbrasio Enhanced Ectasia Display
 - Belin ABCD Progression Display
- Can be upgraded to offer scleral profilometry to aid in scleral lens fitting
- Comparison Maps
 - Belin ABCD Progression Display
 - A (anterior radius of curvature in 3 mm Zone)
 - B (back/posterior radius of curvature in 3 mm Zone)
 - Posterior elevation often changes in keratoconus before anterior changes
 - C (corneal thickness at thinnest point)
 - D (distance corrected visual acuity - DCVA)
 - Reveal trends of progression or stability

14

Keratoconus Classification: Belin ABCD staging and classification system

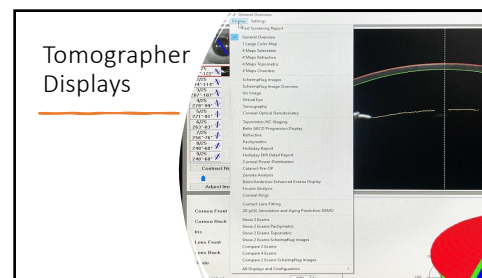
TABLE 1. Belin ABCD Staging/Classification Parameters

ABCD Criteria	A ABC (3-mm Zone) Anterior Radius of Curvature (mm)	B PBC (3-mm Zone) Posterior Radius of Curvature (mm)	C Thinnest Point (µm)	D DCVA
Stage I	<+25 (-4.0 to 0.0)	<+50	>550	>20/20 (1/10)
Stage II	<+25 (-4.0 to 0.0)	<+50	>470	<+20/20 (1/10)
Stage III	<+15 (-0.5 to 0.0)	<+35	>425	<+20/40 (1/20)
Stage IV	<+15 (-0.5 to 0.0)	<+35	>400	<+20/60 (1/30)
Stage V	<+15 (-0.5 to 0.0)	<+35	>400	<+20/100 (1/50)

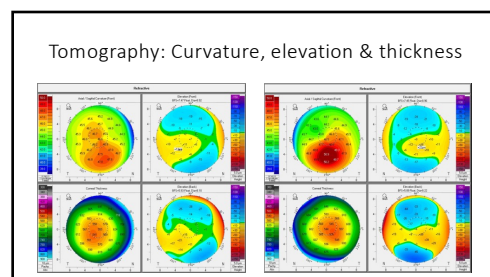
The ABCD system uses 4 parameters: "A" anterior radius of curvature from a 3.0-mm optical zone centered on the thinnest point, "B" posterior radius of curvature from a 3.0-mm optical zone centered on the thinnest point, "C" minimal corneal thickness, and "D" unaided best spectacle-corrected vision and 5 stages (I-V). The first 3 parameters (ABC) are machine-generated (Contact Lens).

Belin MW, Jang HS, Borgstrom M. [Keratoconus: Diagnosis and Staging](#). *J Ophthalmol*. 2022 Jun 1;41(1):1-11. PMID: 34116536

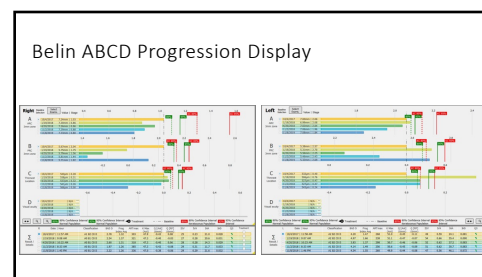
15



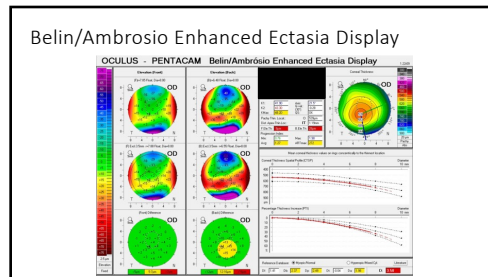
16



17



18



19

Pearl 3:
Strive to be a Detective

Many clues that point to early KC

Follow the clues!

20

Autorefractor

- Error messages
- Erratic results
- Asymmetry in RE or KS
- Ks greater than 47D
- High cylinder
- Oblique axis
- Mismatch in AR and MR

The screenshots show data for Right Eye (RE) and Left Eye (LE) with various parameters like S, C, A, and K. Some values are highlighted in red, indicating potential errors or abnormalities.

21

Refraction and Retinoscopy

- Refraction
 - BCVA is not crisp 20/20
 - Inconsistent responses, weak endpoints
 - Frequent changes from past
 - Over months
 - Age appropriate norms
 - High or asymmetric astigmatism
- Retinoscopy
 - Scissoring reflex (1)

The photographs show a patient being examined by a healthcare professional using a retinoscope. The patient is wearing a fixation target.

(1) Al-Mahrouti H, Orzba SB, Al-Habib S, Mundemkattil N, Babu J, Panchacharam SM, Al-Saidi R, Al-Ratni A. [Retinoscopy as a Screening Tool for Keratoconus](#). *Cornea*. 2019 Apr;38(4):443-445.

22

Patient Complaints

- Glare and starbursts
 - Worse at night
- Monocular shadows & diplopia
- Eye Strain
- Headaches
- Itchy Eyes

Rule out other pathology

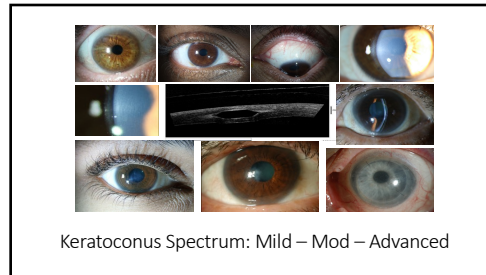
23

Slit Lamp

- Early findings:
 - NOTHING, then subtle changes**
 - Corneal thinning
 - Fleischer's ring
 - Vogt's striae
- Later findings:
 - Advancing ectasia and thinning
 - Munson's sign
 - Central cornea scars
- Don't forget Eyelids:
 - Papillae, lid laxity, floppy eyelids

The photographs show slit lamp views of the cornea and eyelids. One image shows a bright reflection on the cornea, and the other shows a patient's eyelids being examined.

24



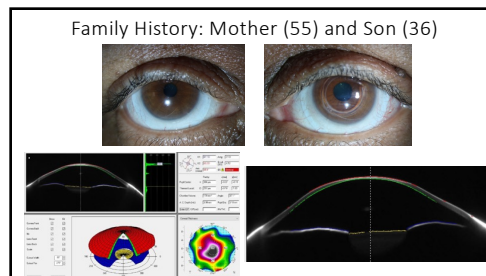
25

Family History

- Make sure to ask about it
- Older generations
 - KC likely not diagnosed
- Study evaluating parents of KC patients (1)
 - 44 KC patients and biological parents (88) recruited (Shandong, China)
 - Parents of KC pts exhibited early signs of KC in corneal tomographic & biomechanical parameters
 - Although findings did not manifest clinically, parents' corneas had reduced stiffness & thickness compared to healthy individuals
- Genetic testing
 - Family
 - Children

Li J, Zhang BN, Jhanji V, Wang X, Li D, Du X. *Parental Corneal Tomographic and Biomechanical Characteristics of Patients With Keratoconus*. *Am J Ophthalmol*. 2023 Aug 9 PMID: 37567431

26

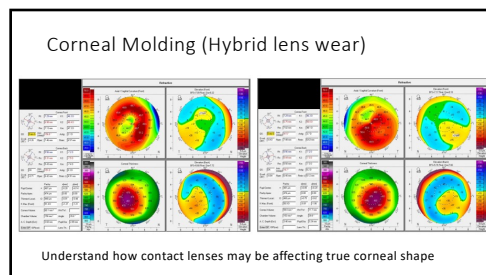


27

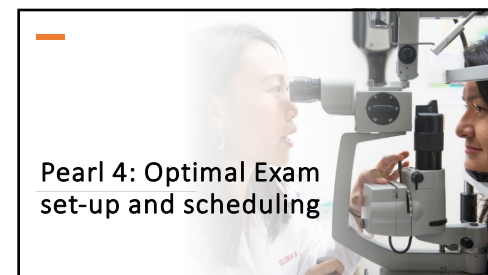
Contact Lens Effects on Cornea

- Corneal Molding with RGPs, hybrid lenses, Ortho-K
 - Need lens holiday for most accurate K measurements
- Flattening even seen with scleral lenses
 - Scleral lens influence on corneal curvature and pachymetry in keratoconus patients. Soeters N, Visser ES, Imhof SM, Tahzib NG. *Cont Lens Anterior Eye*. 2015 Aug;38(4):234-7.
 - curvature parameters significantly flatter compared to ≥1 week after lens removal
 - Avg K(max) 1.1D lower (P<0.001)
 - Effect of Scleral Lenses on Corneal Topography in Keratoconus: A Case Series of Cross-Linked Versus Non-Cross-Linked Eyes. Severinsky B, Fadel O, Davelman J, Moulton S. *Cornea*. 2019 Aug;38(8):986-991.
 - Short-term scleral lens wear in KC patients may cause flattening of anterior cornea.
 - CKL treatment does not guarantee corneal shape stability after scleral lens wear
 - K max dec from 56.7 to 55.8 D

28



29



30

Exam Components

- Need more time for initial consultation
- Need to (re-)educate patient and family on condition
 - Must LISTEN to patient
 - Offer supportive feedback
- Educate on community resources and support groups
 - National Keratoconus Foundation (NKF)
 - Online: Keratoconus Group, Living with KC
 - Provide brochures, handouts
- Discuss:
 - Avoidance of bad habits: Eye rubbing and implications
 - Treatment for ocular allergies, FES, Dry eyes
- Ask about family history



31

Ultimate Consult Goals

- Need to gain patient's trust
- Offer realistic expectations and timeline
- Assurance of partnership and support
- Patient needs to understand treatment of disease vs vision correction



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Pearl 5: Know what to do when keratoconus is detected

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What To Do

- Best to detect early, **before** corneal damage done
- If progression noted, offer corneal cross-linking (CXL)
 - Closer follow-up & lower threshold for CXL should be adopted in pts < 17 years & steeper than 55 D Kmax
- Insurance coverage has improved



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Corneal Cross-linking (CXL)

- FDA approved in April 2016
 - IOL UV System, with Photrexa and Photrexa Viscous (riboflavin)
- Indications:
 - Progressive Keratoconus & Post-Refractive Surgery (LASIK) Ectasia
- Procedure involves:
 - Epithelium removal (Epi-off)
 - 30 min riboflavin application
 - Corneal thickness minimum: **400 microns** (Photrexa if needed)
 - 30 min exposure 365 nm UV-A light, 3.0 mW/cm²
- No specific age range limitations
 - Patients ages 14-65, included in FDA studies
- Activated riboflavin and reactive oxygen species interact in cornea to form crosslinks: stiffens cornea¹
 - 328.9% increase in biomechanical rigidity²



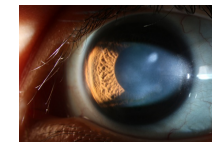
1. Besharwan IM, O'Donnell C, Radhakrishnan H. Biomechanical properties of corneal tissue after ultraviolet A-riboflavin crosslinking. J Cataract Refract Surg. 2013;39(3):403-42.

2. Wolfersnk G. Crosslinking treatment of progressive keratoconus: New hope. Curr Opin Ophthalmol. 2006;17:356-60.

35

CXL Potential Complications

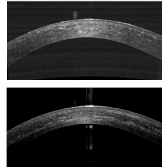
- Infections
- Infiltrates (sterile or infectious)
- Non-healing epithelium
- Corneal Haze
 - Usually resolves over months
- Corneal scarring
- Endothelial cell damage
- Continued progression
- Epi-on vs Epi-off?
 - Current studies evaluating new eye drops, supplemental oxygen, higher energy, pulsed UV



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CXL Post-operative Considerations

- **After procedure**
 - Topical Antibiotic, Steroid (sometimes NSAID)
 - Lubrication
 - Placement of bandage SCL – No eye rubbing!
- **Week 1:**
 - Topical meds, lubrication
 - Remove bandage SCL once epithelium healed
- **Month 1:**
 - Assess vision
 - Corneal Imaging – stromal remodeling
 - Consider CL fitting, getting back into CLs
- **Months 3, 6, 12:**
 - Assess vision – MR and BCVA often change
 - Corneal Imaging



• **Zero Global Period: Visits can be billed to insurance**

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Pregnancy and Keratoconus: Recommendations

- Regular eye exams before pregnancy
- Monitor VA, refraction and topographies closely
- May consider CXL in high risk patients before pregnancy
 - Family Planning
- **Do not perform CXL during pregnancy**
 - No large scale studies, unknown effects on fetus
- Use available correction during pregnancy, VA may flux
- Consider CXL or surgical options after delivery



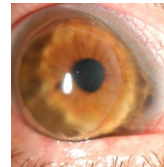
Keratoconus & Pregnancy: a case for pro-active CXL

<https://www.nkef.org/heratoconus-pregnancy-a-case-for-pro-active-cxl/>

38

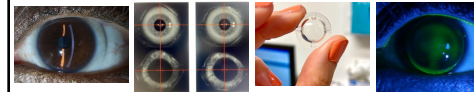
Importance of Early KC Intervention

- **Preserve vision**
 - Maintain ability to wear contact lenses
 - Maintain quality of life
- **Prevent progression**
 - Corneal striae and scars reduce BCVA
- **Protect against need for surgical intervention**
 - Potential complications
 - Graft rejection
 - Cost



39

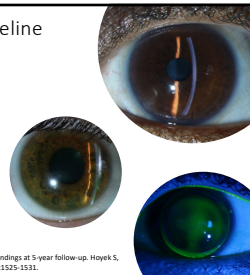
Pearl 6: Understand concurrent management of disease and vision correction



40

Vision Correction Timeline after CXL

- **SCL (toric)**
 - Months (3-6-12), follows changes in Sbx
 - Can wear after few weeks: More flux
- **Hybrid**
 - 1-2 months
 - Minimal changes after CXL
- **RGP**
 - 1-2 months
 - Minimal changes after CXL
- **Scleral**
 - 2-4 weeks in established wearer
 - 1-2 months in new wearer
- Less flux with specialty contact lenses



Corneal flattening following collagen crosslinking for keratoconus: Findings at 5-year follow-up. Hoyer K, Arri N, El Rami H, Saba P, Antoun J. Eur J Ophthalmol. 2021 Jul;31(6):1525-1531.

41

BLOG: How to prepare patients for contact lens wear after cross-linking

Timeline to initiate contact lens wear after cross-linking

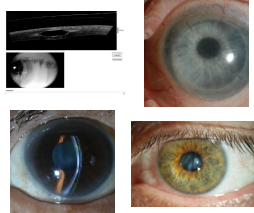
	Soft contact lenses	Hybrid contact lenses	Scleral contact lenses
Start wear	1 month	2 months	Establish at 3 months and begin at 6-12 months
Gas permeable	1 month	2 months	6-12 months
Hybrids	1 month	2 months	6-12 months
Sclerals	2-3 months	2 months	6-12 months

Healio

Recommendations for wearing contact lens wear after corneal crosslinking. Healy, David, MD, PhD, FRCO, FRCO

42

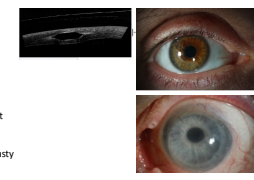
Pearl 7:
Know how to correct vision depending on Keratoconus Stage



43

Procedures that affect shape and vision

- Corneal Intacs
 - FDA approved for KC in 2004
 - "Flattens" cornea
- Topography Guided PRK (TG-PRK)
 - "Smooths" cornea, touch-up
- Cataract surgery and IOL selection
 - Avoid toric IOL in highly irregular K
 - Might consider in CL intolerant patient
- Corneal Transplantation
 - Penetrating (PK) vs Lamellar Keratoplasty
 - 12-20% may require ¹
 - Rate of PK for KC decreasing in US ²
 - * Potential complications

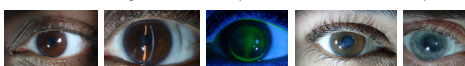


1. Jhanji V, Sharma N, Vajpayee RB. Management of keratoconus: current scenario. Br J Ophthalmol. 2011;95(8):1044-50.
2. Suresky D, Orkin SE, Pan W, VanderBeek BL. Trends in Corneal Transplantation in Keratoconus. Cornea. 2017 Feb;36(2):131-137.

44

Treatment depends on Keratoconus Stage

- Early/mild KC → CXL to prevent progression; SRx, SCL, RGP
 - **Must manage corneal disease and vision, separately**
- Mild → Intacs or combo with TG-PRK to minimize need for GP lenses
- Mod with Scars: Poor Intacs candidate
- Mod-advance → Specialty/Scleral lenses
- Advanced → Specialty/Scleral lenses
- Advanced with significant scars and poor CL corrected VA → K Transplant



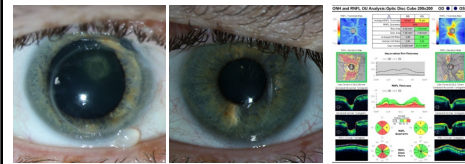
45

Corneal Transplants for KC can lead to Complications

Study: Physicians should maximize use of scleral or RGP CL because patients who successfully use CL have almost one-fifth the risk of undergoing keratoplasty.

Ling JJ, Mar SJ, Stein JD, Rahman M, Polaksky J, Woodward MA. Impact of Scleral Contact Lens Use on the Rate of Corneal Transplantation for Keratoconus. Cornea. 2021 Jan;40(1):39-42.

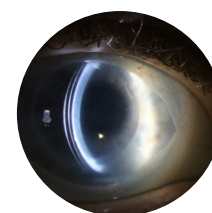
Blindness unlikely with KC itself, but possible if glaucoma/retinal disease develops as complication of PK



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Challenges in fitting post-graft corneas

- Corneal edema/ Hypoxia
- Graft failure
- Graft rejection
- Irregular shape and ectasia
- Chronic topical steroids
- Dry eyes



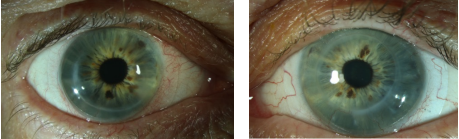
47

Keratoconus and Contact Lenses: Life Changing!



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Scleral lenses s/p PK OU for Keratoconus



- Patient doing great! Daily Scleral lens wear OU in 2023
- K grafts (30 yrs old) cl OU, no rejection, IOP/retina stable
- 20/25+ VA OD and OS

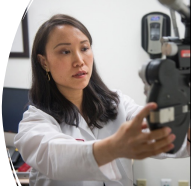
Evolution of Contact Lens Design Over a 30-Year Period and Comparison of Scleral Techniques: A Cohort Study
I. Pringle A, Montard R, Gauthier AS, Desmarests M, Mariet AS, Ratajszak C, Binda D, Saleh M, Delbosq B. Am J Ophthalmol. 2016 Mar;163:59-69. PMID: 26706619

49



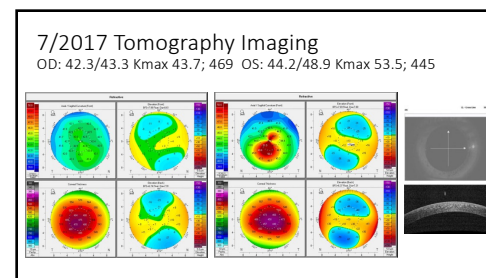
50

Case 1: KC Detected!

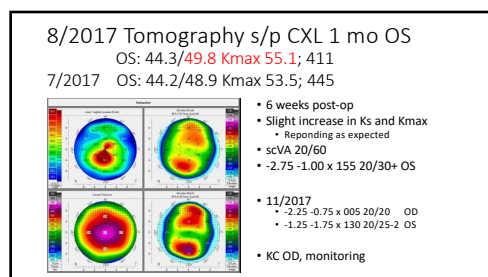


- Presented in 2017, age 17
 - Had "drastic change" in VA + SRx OS over last 2 yrs
- 2015
 - -1.50 -1.75 x 155 OS
- 2017
 - -2.25 -0.75 x 033 20/20 OD
 - -1.75 -6.50 x 160 20/40 OS
 - OD reported to be stable
- Outside optometrist referred for CXL
 - Performed within 2 weeks of referral

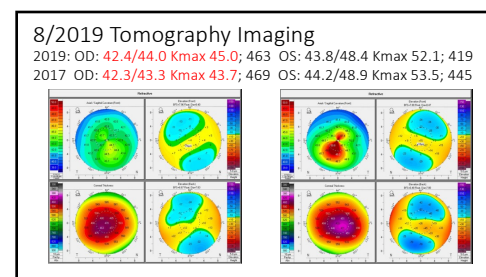
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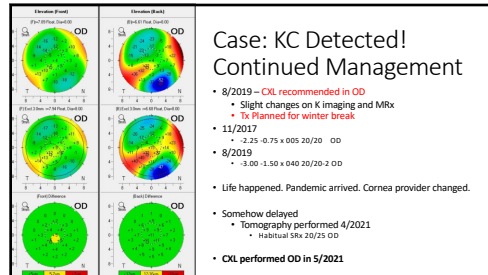
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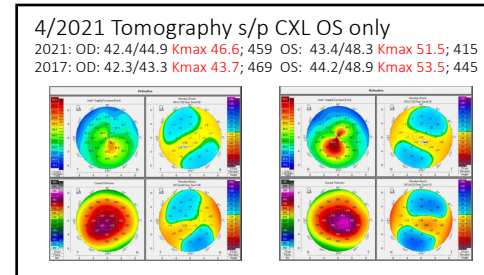
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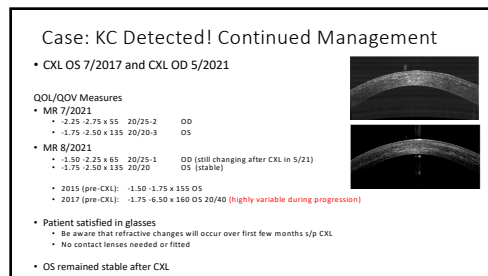
54



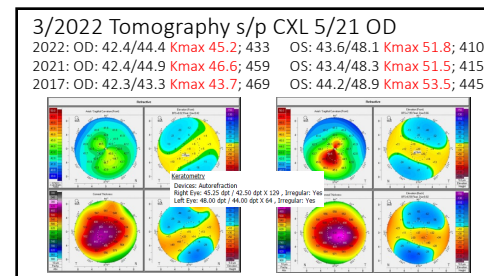
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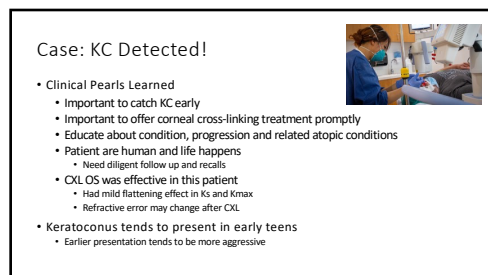
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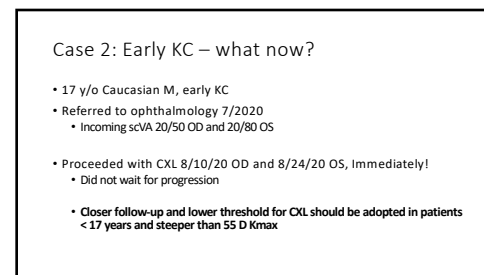
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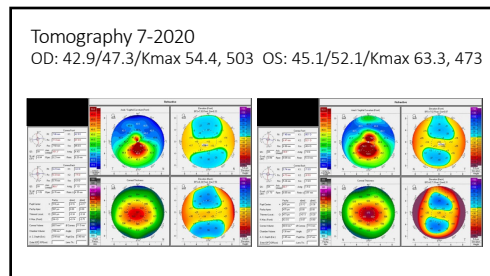
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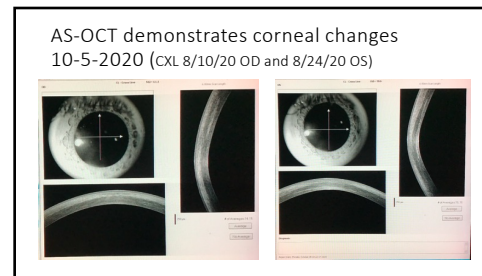
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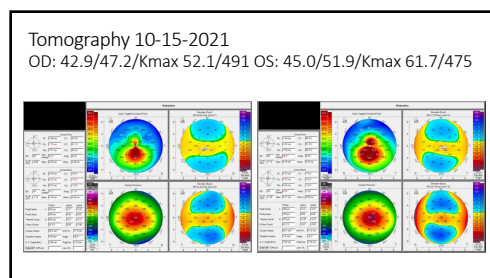
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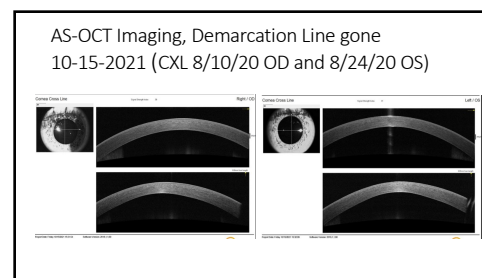
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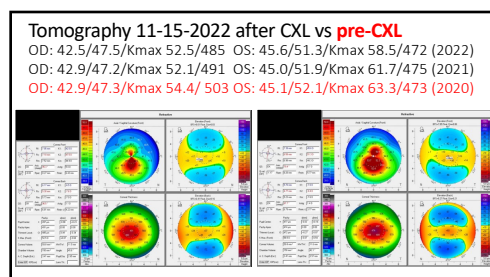
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Case 2: Early KC – what now?

- Refraction changes
 - CXL 8/10/20 OD & 8/24/20 OS
 - AR 10-19-2020
 - -5.75 +4.25 x 99
 - -12.25 +9.00 x 77
 - MR 10-19-2020
 - -3.25 +1.50 x 150 20/25-2 OD
 - -5.00 +2.00 x 100 20/40-2
 - AR and MR do not match
 - MR 11-12-2020
 - -1.50 +1.50 x 150
 - -5.25 +2.50 x 080
 - MR 12-3-2020
 - -1.00 +1.75 x 160 20/25-2
 - -4.50 +2.75 x 080 20/40 (w/ effort)
- 2020: Fit in toric SCL OD and RGP OS
 - 20/20 -2 OD and 20/20-2 OS
- 6-22-2021
 - SCVA 20/20- OD
 - Toric SCL 20/30+ OS
 - -2.50 -2.25 x 010 SCL
- MR 6-20-22
 - -0.50 +0.75 x 120 20/25+2
 - -0.50 +3.25 x 015 20/40
- Eventually wanted scleral lenses (20 yrs old)
 - Blur in Sfx, glare at night
 - Dispensed lenses, needing front toric Rx
 - 20/20 OD and OS, 2-2023

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AR Clues
2-9-2023

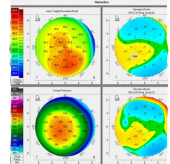
AR still erratic
AR does not
match MR



	S	C	A	
<R>	-5.75	+5.00	93	5
	-5.00	+4.25	95	5
	-5.50	+4.75	94	5
	-6.00	+4.75	95	5
<L>	-5.50	+4.75	95	
	-8.75	+6.25	76	5
	-7.25	+5.25	78	5
	-11.75	+9.50	77	5
Err	-8.50	+6.25	74	E
	-8.25	+5.75	74	E
	-8.75	+6.25	77	
PD	63			
<R>	mm	D	deg	
<R1>	7.83	43.00	5	
<R2>	7.16	47.25	95	
<AVE>	7.50	45.00		
<CYL>		+4.25	95	

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Review of Practical Pearls to Optimize
Keratoconus Care for Patients and Practitioners



- Pearl 1: Must understand KC disease
- Pearl 2: Must be adequately equipped
- Pearl 3: Strive to be a Detective
- Pearl 4: Optimal exam components & goals
- Pearl 5: Know what to do when KC detected
- Pearl 6: Understand concurrent management of disease & vision correction
- Pearl 7: Know how to correct vision depending on Keratoconus Stage

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Thank You!



Gloria B Chiu, OD, FRAO, FSLs
Associate Professor of Clinical Ophthalmology
USC Roski Eye Institute
Adjunct Faculty at Southern California College
of Optometry/MBKU

Department of Ophthalmology
Keck Medicine of USC
T: (323) 442-6335
uscyeve.org
gloria.chiu@med.usc.edu
Instagram: @gloriachiuod

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