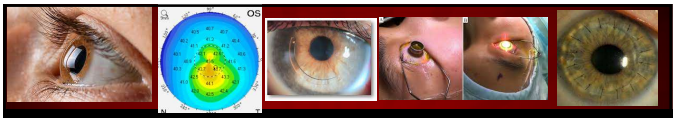


"WHAT'S NEW IN KERATOCONUS" 2022 SOUTHERN EYE CONGRESS TECHNICIAN LECTURE

Kamran M. Riaz, MD
Clinical Associate Professor
Director of Medical Student Research
Cataract, Cornea, External Disease, and Refractive Surgery
Dean McGee Eye Institute, University of Oklahoma
Kamran-Riaz@dmei.org

The Preferred Eyecare Provider for the OKC Thunder

FINANCIAL DISCLOSURES

- I have no financial interests in this lecture or any information discussed therein

- **Unrelated Disclosures:**
  Bausch & Lomb – Speaker
  CorneaGen – Speaker
  Imprimis Pharmaceuticals, Inc. – Research Support
  Ambrx – Consultant
  ImmunoGen - Consultant

FORMAT OF THIS LECTURE

- Discuss the classic/hallmark findings of keratoconus as 2nd last lecture of day… yeesh!
- 1st Emphasis: What's New?!
- 2nd Emphasis: What should an ophthalmic technician know?
- Items in yellow indicate one of these two emphasis points

INTRODUCTION

A Treatise of the Diseases of the Horny-coat of the eye, and the Various Kinds of Cataracts. Also, a Method, Entirely new, of Scarifying the Eyes for Several Disorders Hardcover – April 24, 2018

- From Greek:
  - Kerato: cornea
  - Conus: cone-shape
  - "Ochlodes" meaning "annoying" in Greek
- Duddell (1736): prominent corneas in a 14-yo boy
  - Various procedures: repositioning pupil away from cone, iris incarceration to make a slit pupil, cauterization of the cone, full thickness excision
- Nottingham (1854): KCN definition and associations

DEFINITION OF KERATOCONUS (KCN)

- Non-inflammatory, ectatic condition of the cornea in which there is progressive central thinning and steepening causing irregular astigmatism and decreased visual acuity

- Non-inflammatory: recent thoughts suggest otherwise
- Ectasia: next slide
- Cornea: you should know what this is –
  - most important part of the eye
- Thinning
- Steepening: dome-shape → cone-shape
- Irregular astigmatism
- Decreased visual acuity

KCN IS ONE MEMBER OF CORNEAL ECTASIA FAMILY

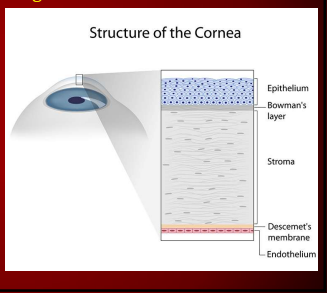
- Corneal ectasias: "dilation or expansion of a tubular organ".
- Clinically, thinning caused by corneal ectasias leads to protrusion or bulging of the affected region(s) of the cornea.
- Can be asymmetrical (one eye more affected) especially in early disease states
- In early disease, patients often 1) have minimal symptoms other than blurry vision 2) no symptoms
  - Diagnosis is increasingly dependent on 1) good history 2) diagnostic testing
  - Role of the ophthalmic technician

TYPES OF CORNEAL ECTASIAS

- Primary
  - KCN
  - Pellucid Marginal Degeneration
  - Keratoglobus
  - ?Terrien's Marginal Degeneration
- Secondary
  - Post-LASIK ectasia
    - Corneal strength decreases after both surface and lamellar ablation
  - Post-traumatic ectasia
  - Post-corneal scarring ectasia

KCN AFFECTS ALL 5 LAYERS OF THE CORNEA

- Epithelium: thinning early, thickening late
  - Role of ETM
- Bowman's: breaks
- Stroma: major area of thinning
  - Role of Tomography and AS-OCT
- DM/endo: break → hydrops



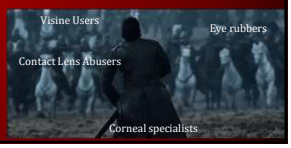
WHY SHOULD WE CARE?

- Most common corneal dystrophy/disease in the United States
- Approximately 1/1500-2000 in the USA
- Pts will come into comprehensive/optometry practice first
  - They will see YOU first

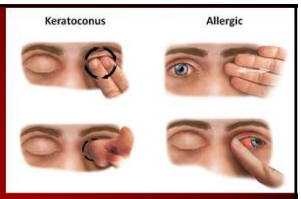
- Patients come in for:
  - "unhappy with glasses/contacts"
  - "I keep having to change my glasses"
  - "I want LASIK before my eyes get worse"

WHAT CAUSES KCN?

- "Two hit hypothesis"
- Genetic predisposition
  - Family members: first-degree relatives (7% with FH) – history important!
  - Genetic disease: Down's Syndrome, Ehlers-Danlos
- Inflammatory/Hormonal changes → increased tear film inflammation → eye rubbing
  - Eye rubbing also happens with asthma, hay fever, eczema, AKC/VKC
  - Sleep apnea/floppy eyelid syndrome



TANGENT ON EYE RUBBING



- Eye rubbing is a hallmark sign of both allergic eyes AND keratoconus
- All eye rubbing is not the same!
- Ask the patient how they rub their eyes
- Look for the patterns/complaints:
  - allergic eye patient: rubs eyes "**because it feels good**"
    - Rub eyes back and forth with PALM or BACK OF HAND
    - Rub the medial or lateral canthus
  - KCN patient: rubs eyes because "**it helps me see better**"
    - Rubs eyes with greater pressure on the center of the eyelid, using circular motions with a knuckle or two fingertips pads.

SYSTEMIC ASSOCIATIONS

- Atopic/Allergic Disease (30-40%)
- Downs Syndrome
- Ehlers-Danlos Syndrome
- Marfan Syndrome
- Cruzon's Syndrome
- Apert's Syndrome

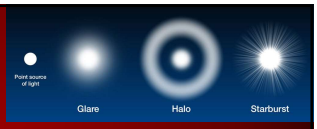
PREVENTION OF KCN

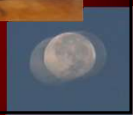
- No single strategy proven effective
- Avoiding eye rubbing
- Pressure on eye while sleeping (e.g., sleeping on belly)
- Treating allergies (to avoid eye rubbing)

- Theme: avoid mechanical pressure on the eye!

CLINICAL PRESENTATION

- "Disease of young people"
- Puberty onset/worsening most rapid (12-30 years of age)
- Decreasing visual acuity
- Frequent change of glasses
- Difficult refraction
- Glare and light sensitivity
- Complaints of "ghost images"





CLINICAL FEATURES OF TYPICAL KCN PATIENTS

- Heavy set/barrel chested
  - "breathe loudly"
  - Frequent straining/holding breath
  - "difficult to get into slit lamp"
- Frequent eye rubbing
- Atopic/allergic disease

MAKING THE DIAGNOSIS

- Clinical History
- Symptoms reported by the patient
- Clinical exam signs (often minimal)
- Corneal Imaging and Testing
  - Importance of high-quality imaging
  - Topography
  - Tomography
  - Pachymetry
  - Epithelial Thickness Mapping
  - Anterior Segment OCT
  - Corneal Biomechanics

MAKING THE DIAGNOSIS: ROLE OF THE TECHNICIAN

Important parts of the eye exam

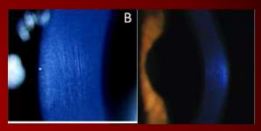
- Measurement of uncorrected visual acuity
- Measurement of corrected visual acuity (monocular/binocular)
- Pinhole visual acuity
- Manifest refraction, paying attention to
  - the amount of cylinder correction and axis location (WTR, ATR, oblique)
  - Asymmetric refractive error with high, progressive or mixed astigmatism
- Retinoscopy: look for scissoring reflex

SCISSORING REFLEX VIDEO

- https://www.aao.org/1-minute-video/retinoscopy-findings-in-keratoconus

CLASSICAL EXAM FINDINGS

- Vogt Striae
- Fleischer Ring
- Munson's Sign
- Rizzuti's Sign
- Oil Droplet Reflex

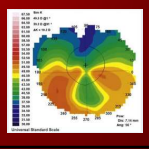


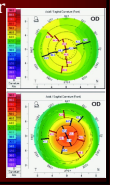
MAKING THE DIAGNOSIS

- Corneal topography: quantitative and qualitative evaluation of the **anterior corneal surface** and readily displays information using color-coded maps to aid clinical decision making.
- Corneal tomography: additionally measures **corneal thickness** and gives information about the **posterior cornea**.
- Information gained from both of these imaging techniques allows clinicians to diagnose ectatic disease, assess disease progression, and determine appropriate non-surgical/surgical interventions.
- Both are used to critically screen candidates who present for laser refractive surgery, especially to determine which candidates have an increased risk for post-refractive surgery corneal ectasia.
- Role of technician: alert doctor, especially in new patient with poor vision, about potentially getting imaging after the history/workup prior to being seen by clinician

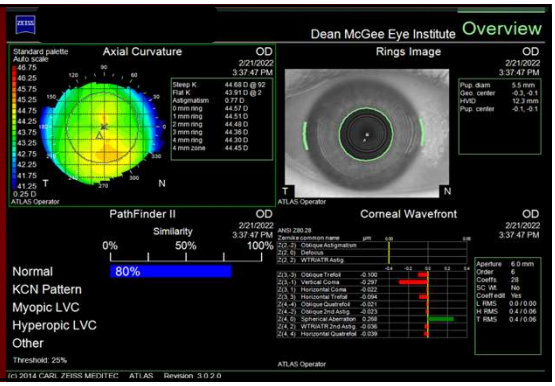
WHAT IS CORNEAL TOPOGRAPHY

- Characterizes the shape of the cornea, similar to describing the earth's surface using a topographic map ("mountains, plains, valleys and sea level")
- Historically, was used only to describe the anterior corneal surface; newer devices can give information about the posterior surface
- Three Main Methods
  - Placido-Disk Imaging
  - Scanning Slit Imaging
  - Scheimpflug Imaging

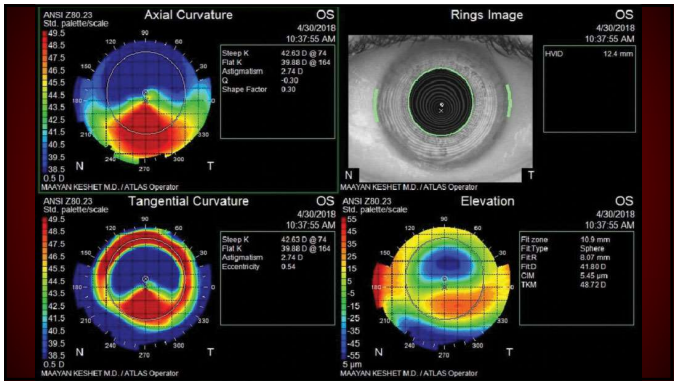




NORMAL

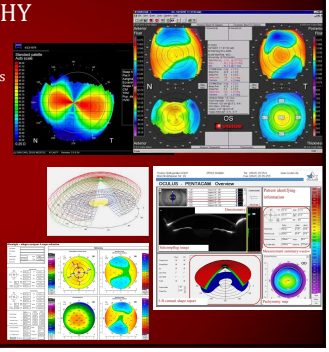


KCN



TOPOGRAPHY VS TOMOGRAPHY

- Topography
  - *Topo* ="to place", *graphein* ="to write"
  - Describes the shape of a surface (such as the earth's surface)
  - Can also evaluate tear film and ocular surface
- Tomography
  - *Tomos* means "to cut into sections"; CT scan: 3D image of organ
  - examination of the front and back surfaces of the cornea, along with pachymetric mapping → 3D image of cornea
  - Best device: high sensitivity and specificity for detecting ectasia



SCHEIMPFLUG IMAGING (OCULUS PENTACAM)

What All Does It Show?

- Topography of anterior and posterior surface
  - Back surface changes happen before front surface changes
- Pachymetry of anterior and posterior surface
- Calculation of AC angle, chamber volume, chamber height
- Images of the anterior segment (cornea, iris, lens)
- Densitometry of cornea and lens
- Images taken → data transferred to computer → 3d virtual model to derive additional information

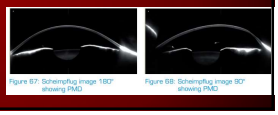


Figure 67: Scheimpflug image 180° showing PMD

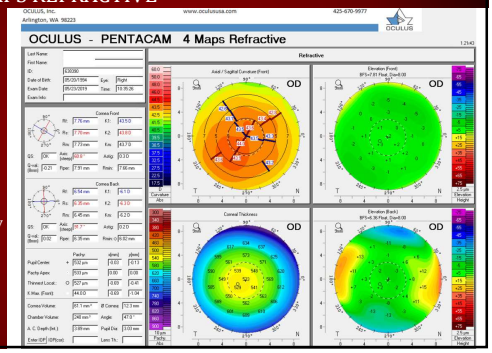
Figure 68: Scheimpflug image 90° showing PMD

PENTACAM: 4 MAPS REFRACTIVE

Normal Eye

Main Things to Look For

1. Equal colors in Axial Map
2. Amount of astigmatism
3. Kmax value
4. Elevation Maps ("sea of green")
5. Pachymetry values for local vs overall thinning (though ultrasound pachy is most important)



PENTACAM SETTINGS: DIFFERENT DOCTORS LIKE DIFFERENT SETTINGS

- K-value: Diopters; R-value: Base Curve
  - Blue is flat, Red is steep

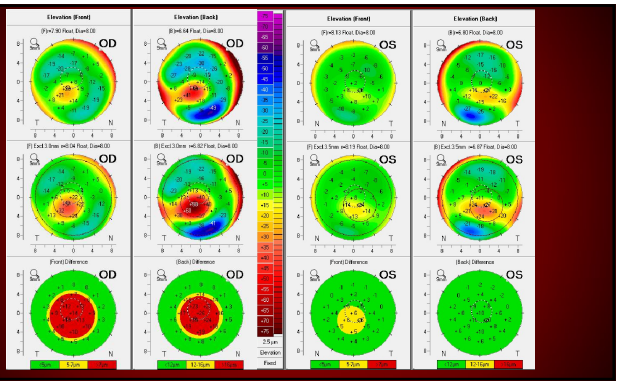
- Color Map Scale (can vary): look at numbers, not just colors!
  - Can adjust for **diameter** vs radius (default is diameter)
- Make sure it's a good scan:
  - No "blink" or "bad data"

BELIN-AMBROSIO DISPLAY (BAD)

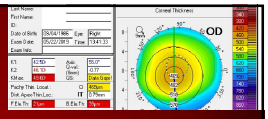
- Anterior curvature and pachymetry alone not enough to detect disease
- BAD: combining ant/posterior elevation and pachymetric data into one all-inclusive ("Megazord") display → quick/effective screening

FIRST COMPONENT

- BFS (8.0mm zone): allows generation for standardized values but can minimize effects of elevation
- TL;DR: BAD will eliminate artifact and noise and better show an ectasia that may otherwise by hidden by standard reference maps using BFS 8.0mm

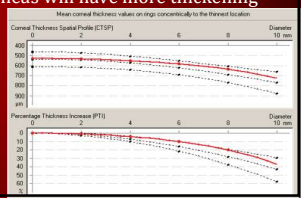


BELIN-AMBROSIO DISPLAY (BAD)



SECOND COMPONENT

- The displacement of the TP from the corneal apex, along with direction of displacement is given
- CTSP and PTI Maps included: ectatic corneas will have more thickening from the TP to the periphery



BELIN-AMBROSIO DISPLAY (BAD)

THIRD COMPONENT

- Progression Index

1. PI-min: minimum pachymetric progression index
2. PI-max: maximum pachymetric progression index
3. PI-avg: average pachymetric progression index
4. ARTmax: maximum Ambrosio relational thickness
   --importance of relative corneal thickness indices rather than point measurements
   --ARTmax passed on PI-max, PI-avg and thinnest point on the cornea
   --Lower numbers = no good → nada de bom
   --Cutoff: approximately <390 should alert you

BELIN-AMBROSIO DISPLAY (BAD)

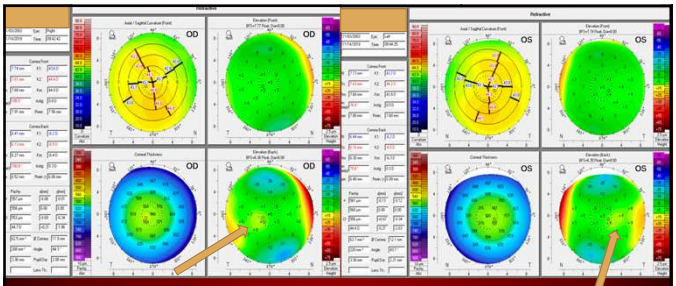
- 5 Additional Values to Review:

1. Df(front): change in anterior elevation from standard to enhanced refractive surface
2. Db(back): change in posterior elevation from standard to enhanced refractive surface
3. Dp: Pachymetric progression
4. Dt: thinnest value
5. Da: thinnest displacement (aka ArtMax)

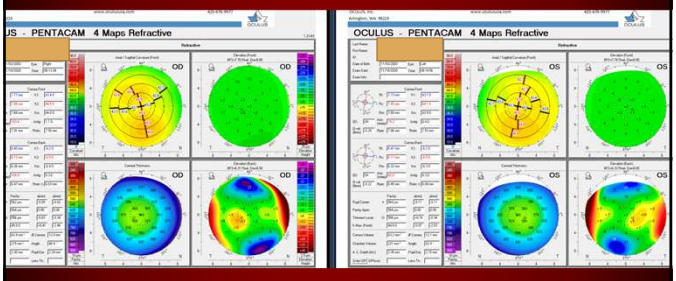
The final parameter "D" represents an overall reading of all five parameters compared to a database normal and keratoconic corneas. Alert the doctor if "D" is yellow or red.

One value may fall outside the norm, but "D" may still be normal

Conversely, multiple yellow parameters within the 5 may yield a red "D" value: HIGH RISK

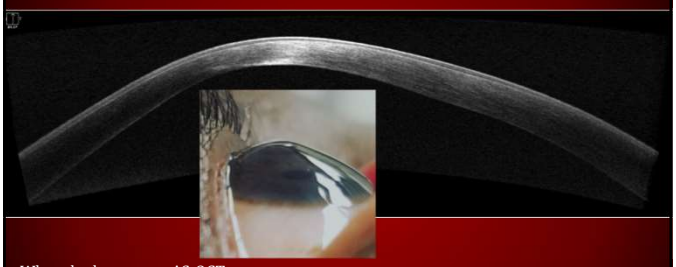


Patient interested in refractive surgery…

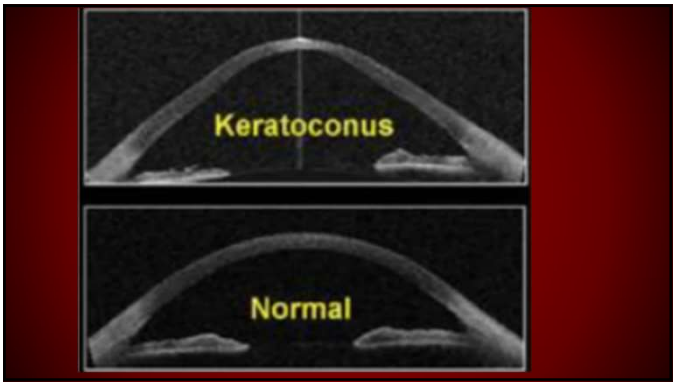


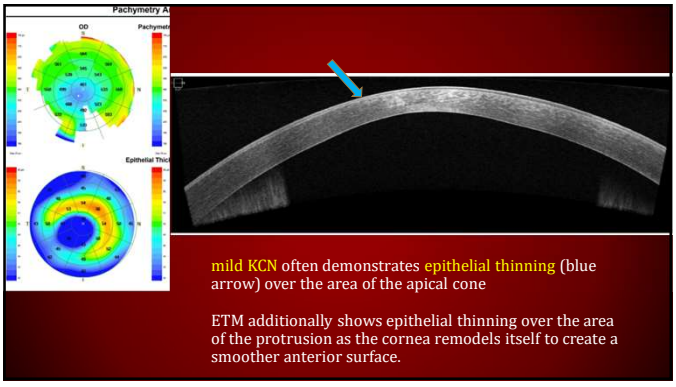
6 month f/u became 1 year (COVID) – Now 11/2020

Other Imaging: Anterior Segment OCT



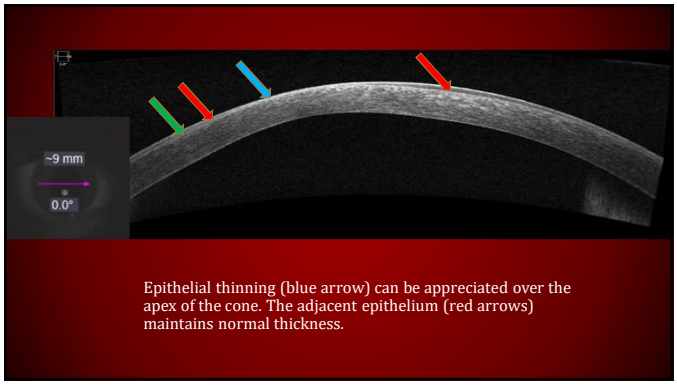
When do doctors get AS-OCT:
1. Baseline imaging
2. Assess presence of scarring
3. Effect of CXL





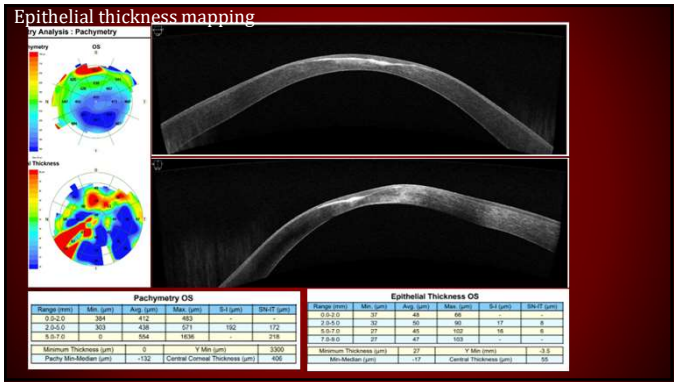
mild KCN often demonstrates epithelial thinning (blue arrow) over the area of the apical cone

ETM additionally shows epithelial thinning over the area of the protrusion as the cornea remodels itself to create a smoother anterior surface.



Epithelial thinning (blue arrow) can be appreciated over the apex of the cone. The adjacent epithelium (red arrows) maintains normal thickness.

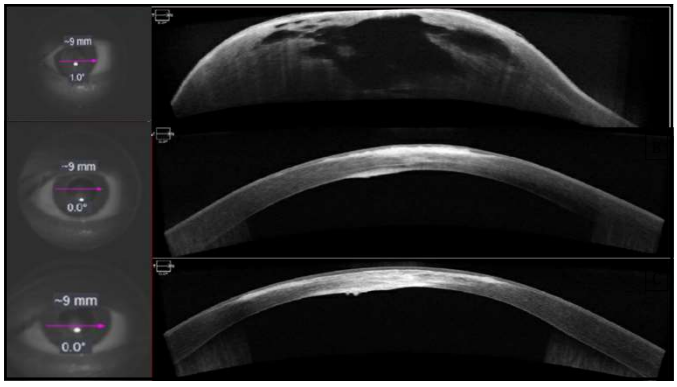
Epithelial thickness mapping

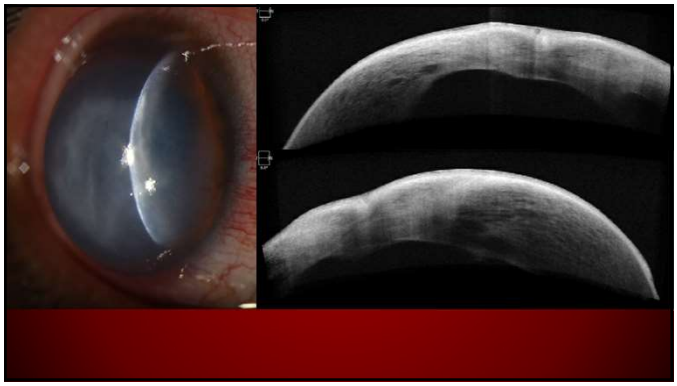


COMPLICATIONS OF KCN

- Hydrops: break in DM → fluid rushes into corneal stroma
- Patients present with PAINFUL, SUDDEN VISION LOSS with cloudy cornea, tearing, light sensitivity
  - Usually in a patient with known diagnosis of KCN
  - Can be some patients' first time seeing an eye doctor


Exam Date: 6/21/2017
Exam Time: 1:34 PM
Gender: Male
Technician: Operator, Cirrus
Serial Number: 5000-2853
Signal Strength: N/A
HD Cornea Analysis : HD Cornea
OD
OS
~9 mm
205.0°


~9 mm
1.0°
~9 mm
0.0°
~9 mm
0.0°



GOAL 1: HALTING THE DISEASE PROCESS

- How can we do this?
- Educating the patient re: eye rubbing
- Treating any allergic disease, ocular surface disease, etc.
- Oral riboflavin with UV light exposure
- Corneal Cross Linking

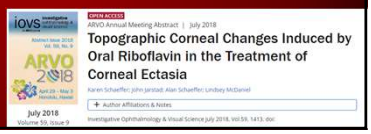
ORAL RIBOFLAVIN

- 400 mg twice daily – used by neurologists for treatment of migraine headaches
- 10-15 minutes UV light exposure during peak UV time (~10 am to 2 pm)
  - DON'T LOOK AT THE SUN DIRECTLY!

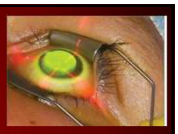
Supplement Facts:
Serving Size: 1 Softgel

	Amount Per Serving	% Daily Value
Riboflavin (Vitamin B2)	133 mg	7824%
Magnesium (from Magnesium Oxide and Magnesium Citrate)	57 mg	14%
Proprietary Blend	101 mg	*
PA Free Butterbur Extract (Petasites hybridus)		*
Coenzyme Q10		*
Bioperine™		*

*Daily Value not established.



CORNEAL CROSSLINKING (CXL)

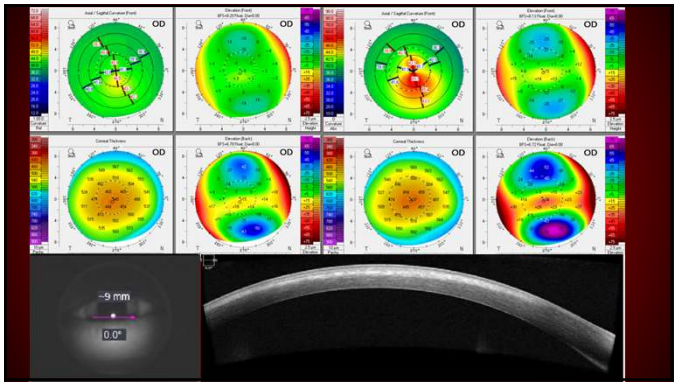


- Since mid2000's; USA approval 2016
- procedure used to strengthen the cornea
- Liquid riboflavin is used to "marinate" the cornea
- UV light is used to active the riboflavin
- causes new corneal collagen cross-links to develop.
- Those cross links cause the collagen fibrils to shorten and thicken, leading to a stiffer, stronger cornea.
- "Epithelium-off" (Dresden protocol): only FDA-approved method
- Other protocols: epithelium on, accelerated CXL, etc.

ROLE OF OPHTHALMIC TECHNICIANS IN CXL

- Invaluable!
- Application of riboflavin
- Checking pachymetry
- Performing UV light treatment
- Making sure I sign the forms ☺
- Making sure I send in the meds ☺
- Patient Education re: drops

CROSSLINKING VIDEO



line of demarcation appears as a prominently hyper-reflective line within the stroma. In this patient, the treatment has affected approximately one-half of the stromal thickness.

OTHER FUTURE THERAPIES

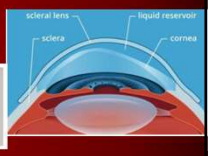
- KCN corneas have reduced lysyl oxidase (LOX) → opical medications (IVMED-80) to increase LOX activity in the cornea
- CXL at the slit-lamp
- On-Eye Cross-Linking: scleral lens-based device filled with fiboflavin is connected via thin fiber optic cable to a small, portable UV delivery device
  - The riboflavin doesn't have to be readministered during the treatment, nor does the treatment require additional oxygen

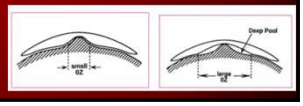
GOAL 2: HELPING PATIENTS SEE BETTER (NONSURGICAL)

- Glasses:
  - Importance of excellent, meticulous, patient refraction
  - May need to utilize streak refraction, Jackson cross cylinder, etc.
  - Patience with patients!

GOAL 2: HELPING PATIENTS SEE BETTER (NON-SURGICAL)

- Contact Lenses
  - Soft toric
  - Rigid gas permeable contact lens
  - Piggyback CL options – pt first wears soft contact lens, then RGPCL
  - Rose K lens: multicurve lens to vault over apex
  - Hybrid CL – lens with a soft "skirt" and hard center
  - Scleral contact lenses
  - Custom geometry lenses
  - Bitoric contact lenses



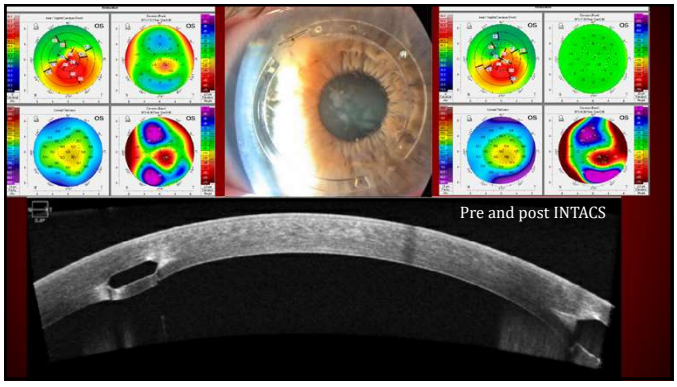


GOAL 2: HELPING PATIENTS SEE BETTER (SURGICAL)

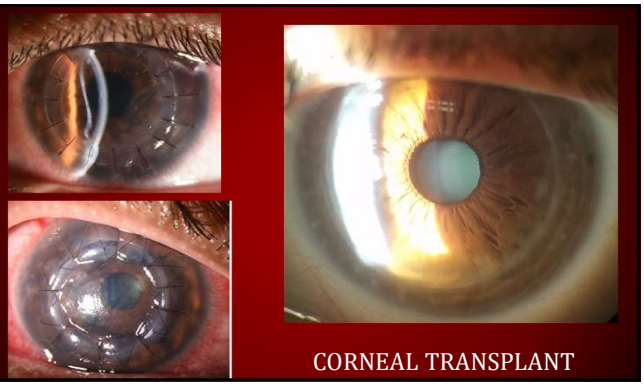
- Corneal ring segments: INTACS
- PRK combined with or after CXL: controversial!
- Conductive keratoplasty (CK) after CXL: controversial
- Phakic IOL
- Bowman's membrane transplantation
- Corneal transplantation: DALK or PKP
  - Both can be used to achieve good VA
  - DALK lower rejection rate
  - Good prognosis for KCN patients

INTACS

- Semi-circle PMMA rings placed in the midperipheral corneal stroma
- Channel made mechanically or with a laser
- Segment placement → corneal flattening → correction of astigmatism and myopia
- INTACS can be used in mild-moderate keratoconus with CXL to "flatten and freeze" an ectatic cornea. First, INTACS can be placed to reverse corneal steepening and then CXL can be used to halt disease progression.
  - Several strategies have been proposed, including combining INTACS with CXL as a single procedure, or to stage the two procedures by several months.
  - When staging, some surgeons prefer CXL followed by INTACS, while others prefer the opposite.



Pre and post INTACS



CORNEAL TRANSPLANT

KERATOCONUS AND CATARACT SURGERY

- Patients with stable KCN may come in for cataract surgery later in life
- What do you need to know as an ophthalmic technician?

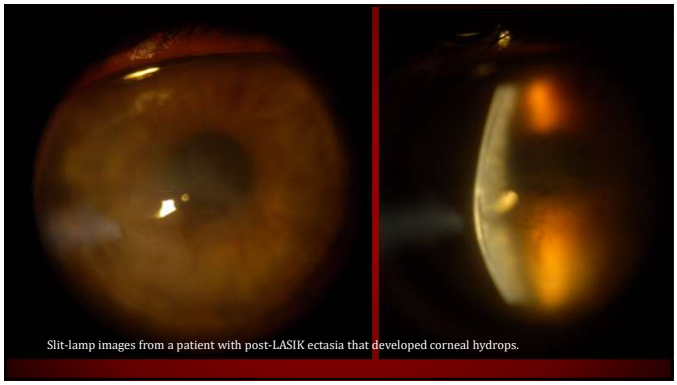
KERATOCONUS AND CATARACT SURGERY

- Recall that KCN progression tends to plateau in middle-age years, and these patients may have good vision with glasses or contact lenses until they develop visually-significant cataracts.
- Two groups of KCN patients with cataracts:
  - KCN patients who are accustomed to wearing **glasses only**
  - KCN patients who are accustomed to primarily wearing **contact lenses** (e.g., soft toric contact lenses (STCLs), rigid gas permeable (hard) contact lenses (RGPCLs), and scleral contact lenses (ScCLs))
- crux of the problem in KCN corneas: *the steeper the cornea, the greater the risk for postoperative hyperopia.*

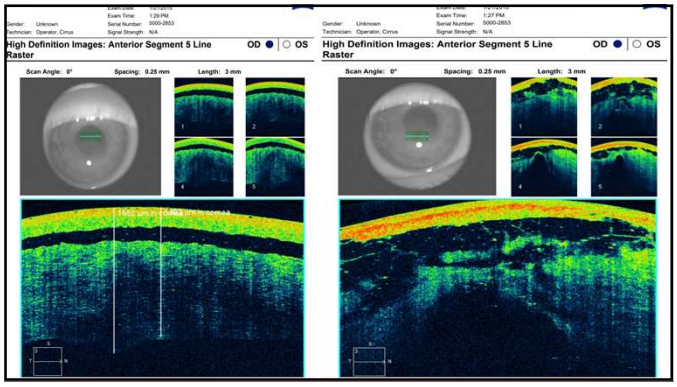
KERATOCONUS AND CATARACT SURGERY

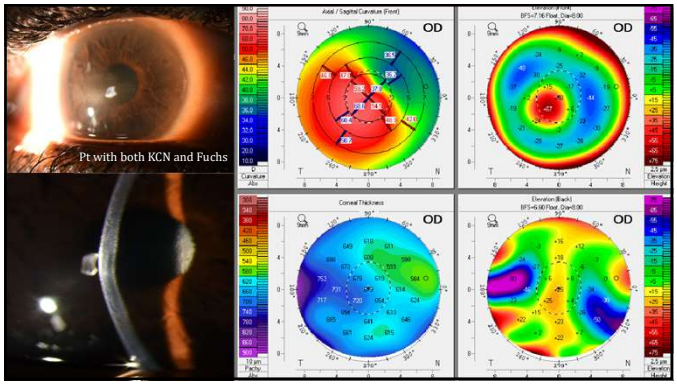
- Surgeons may want to get multiple device measurements
  - Manual K, Auto K, Biometry Ks, Topography, Tomography, Ray-Tracing Devices
- IOL options:
  - Monofocal or toric – if they are happy in contacts, monofocal
  - Tell your surgeon not to use a premium IOL! ☺
- Surgery expectations
  - They can't compare their results to "normal" corneas
  - Extra time to heal

POTPOURRI

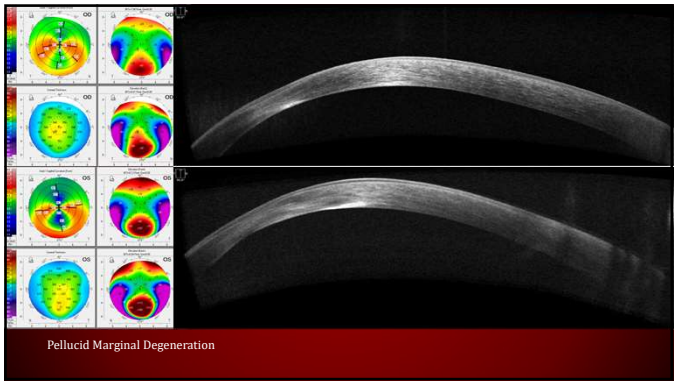


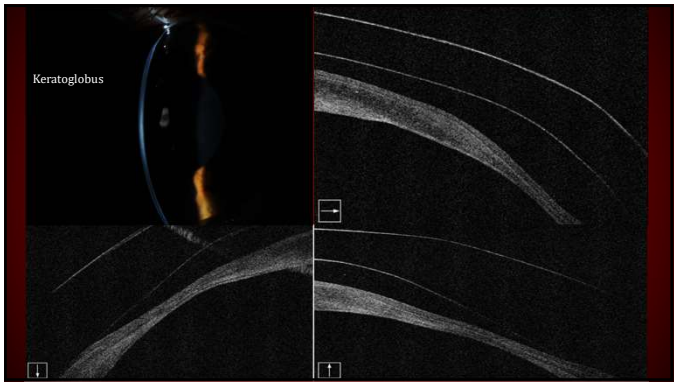
Slit-lamp images from a patient with post-LASIK ectasia that developed corneal hydrops.

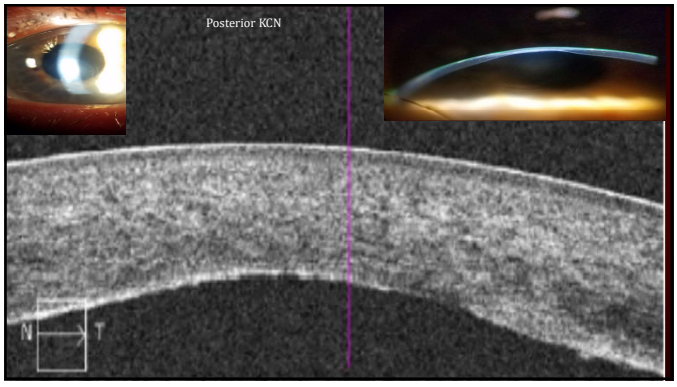




Pt with both KCN and Fuchs


Pellucid Marginal Degeneration


Keratoglobus


Posterior KCN
N
T

FINAL THOUGHTS

- KCN is commonly seen in eye care practices
  - Ophthalmic technician may be the first clinical care person a KCN pt has ever seen
- Role of technicians is vital
  - 1) history taking
  - 2) refraction
  - 3) obtaining good imaging
  - 4) assisting with treatments
- Knowledge of treatment options can help you
  - Answer some patient questions
  - Understand what the clinician/surgeon does for these patients
- If nothing else, please tell EVERY PATIENT: NO EYE RUBBING!!!!!!

THANK YOU

- Questions/Comments?