Caution Steep Hill Ahead: Diagnosing and Managing Keratoconus in Clinical Practice: Are you Ready to Conquer Keratoconus?

Melissa Barnett, OD, FAAO, FSLS, FBCLA

One Shields Avenue Davis, CA 95616 drbarnett@ucdavis.edu

Shalu Pal, HBSc, OD, FAAO, FSLA, FBCLA

5 Marine Parade Drive, Suite 913 Toronto Ontario M8V 4B4 shalupal@hotmail.com

Course description

Are you ready to conquer keratoconus? This lecture focuses on the modern management of keratoconus, early diagnosis, how to stop progression and rehabilitate vision. We will cover everything from methods to detect keratoconus earlier, an overview of corneal collagen cross-linking, and contact lens and surgical treatment options. Recognizing the methods to manage keratoconus sooner will shift the treatment paradigm for patients to improve quality of life, reduce the economic burden of the disease and avert vision loss.

Course Objectives

- 1. Be aware of different studies of keratoconus prevalence.
- 2. Learn how to identify key characteristics in patients with keratoconus.
- 3. Obtain knowledge of how to utilize common in-office diagnostics that are available in clinical practice.
- 4. Identify risk factors for keratoconus.
- 5. Identify goals with contact lens correction.
- 6. Learn when corneal collagen cross-linking is indicated for a patient with keratoconus.
- 7. Become knowledgeable of different surgical options for keratoconus.

Outline

- 1. Overview of keratoconus
 - a. Definition
 - b. Prevalence
 - c. Clinical signs
 - d. Hydrops and mitral valve prolapse
 - e. Pathogenesis of keratoconus
 - f. Risk factors that reduce the risk of keratoconus
- 2. Comorbidities in keratoconus
 - a. Vigorous eye rubbing
 - b. Atopy

- c. Ocular allergies
- d. Floppy eyelid syndrome
- e. Ethnicity
- 3. Risk factors for keratoconus
 - a. Age
 - b. Genetics
 - i. Family history
 - c. Comorbidities
 - i. Down syndrome
 - ii. Connective tissue disorders
 - 1. Marfan syndrome
 - 2. Ehlers-Danlos syndrome
 - iii. Pregnancy
 - 1. Change due to rise in pregnancy-related hormones (estrogen and relaxin)
 - 2. Alter corneal biomechanical properties and increase its refractive power
 - 3. No evidence of reversal after pregnancy
- 4. Importance of early diagnosis
 - a. Corneal collagen cross-linking
 - b. Strengthen the cornea to prevent progression
 - c. Avoid need for corneal transplantation
 - d. Economic burden of keratoconus
- 5. Diagnosing keratoconus in any practice without corneal imaging
 - a. Retinoscopy
 - i. Scissors reflex
 - ii. Al-Mahrouqi, et al. Retinoscopy as a Screening Tool for Keratoconus, Cornea: April 2019
 - 1. Retinoscopy is a sensitive and reliable test for detecting keratoconus including early disease
 - b. Refractive changes
 - i. Increase in astigmatism
 - ii. Increase in myopia
 - iii. Asymmetry in astigmatism between the eyes
 - iv. Frequent glasses prescription changes
 - c. Visual symptoms
 - i. Blurry vision
 - ii. Reduced best corrected vision
 - iii. Shadows / halos in vision
 - iv. Vision that is not crisp, even when 20/20
 - d. Keratometry
 - i. Steep corneal curvatures over 47.00D
 - ii. Irregular mires
 - e. Pachymetry
 - i. Thin corneal thickness less than $500\mu m$
 - f. Corneal findings with slit lamp examination
 - i. Vogt's striae
 - ii. Corneal stromal thinning
 - iii. Munson's sign

- iv. Conical protrusion
- v. Fleischer ring
- vi. Hydrops
- vii. Subepithelial or anterior stromal scars
- g. Genetic testing
 - i. How to incorporate genetic testing
 - ii. Testing in children
- 6. Collaborative care
 - a. Refer a keratoconus suspect for corneal topography / tomography
 - i. Family history
 - ii. Any risk factors, especially vigorous eye rubbing
 - b. Practice management benefits to keep patients in your practice
 - c. Patient evaluation after corneal stabilization
- 7. Benefits of referring patients for corneal imaging
 - a. Prevent progression
 - b. Early treatment
 - c. Easier to fit glasses and contact lenses
 - d. Decrease economic burden of the condition
- 8. Correction and treatment of unstable/ectatic conditions:
 - a. Keratoconus/Corneal Ectasia old mantra vs. new mantra.
 - b. Early diagnosis is critical
 - c. Treatment
 - i. Corneal collagen cross-linking
 - ii. Corneal surgery
 - iii. Contact lenses
- 9. Halt progression with corneal cross-linking (CXL)
 - a. Review CXL procedure
 - b. Epi off/Dresden protocol
 - c. Three ingredients for CXL; Riboflavin, UV (A) light and Oxygen
 - a. FDA approval trial data published for progressive KC
 - b. FDA approval trial data published for progressive post-refractive surgery ectasia.
 - c. Seven-year follow-up of CXL. Review Kmax line-graph
 - i. Corneal Cross-linking to Halt the Progression of Keratoconus and Corneal Ectasia: Seven-Year Follow-up
 - ii. Age considerations with cross-linking. When evidence of progression is needed.
 - d. Post-operative care and important factors for co-management
 - e. Epi-on vs. epi-off CXL
- 10. Further KC worsening may occur while progressive KC eyes wait to receive CXL
 - a. Review of studies
- 11. Risk factors associated with further KC progression while waiting for CXL treatment
 - a. Higher Kmax at baseline
 - b. Worse tomography/topography KC indices at baseline
 - c. Age < 18 Yrs & Romano et al proposed an age-dependent maximum wait time
 - d. Pregnancy
- 12. Intacs (Intrastromal ring segments)
 - a. What they are
 - b. How they work

- 13. Corneal transplantation
 - a. Incisional
 - b. Laser
 - c. Complications
 - d. Collaborative care
 - e. When to fit contact lenses
- 14. Goals with contact lens correction
 - a. Provide good vision
 - i. By masking the irregular corneal surface
 - 1. Reduces aberrations
 - a. Improves acuity
 - b. Provide good corneal health
 - c. Provide adequate comfort for all hours of wear
- 15. Diagnosis
 - a. Subjective symptoms
 - b. Refractive shift
 - c. Keratometry mires
 - d. Corneal topography
- 16. Treatment
 - a. Corneal collagen cross-linking
 - b. Corneal surgery
 - c. Contact lenses
- 17. Corneal GP Contact Lenses
 - a. Benefits
 - i. Provides rigid optical surface
 - 1. Good optics
 - ii. Widely available
 - iii. Lower cost than many other options
 - iv. Wide range of lens design parameters available
 - b. Challenges
 - i. Poor success if corneal elevation difference is greater than 350 microns (scleral lens indicated)
 - ii. Areas of touch may lead to:
 - 1. Staining
 - 2. Scarring
 - 3. Corneal warpage
 - c. Fitting tips
- 18. Piggyback (PB) Lens System
 - a. Definition: Corneal GP over a soft contact lens
 - i. GP provides good vision
 - ii. Soft protects cornea
 - b. Indications
 - i. Poor comfort with GP
 - ii. Minor corneal abrasion with GP wear (temporary)
 - c. Challenges
 - i. Extra cost
 - ii. Extra hassle
 - 1. Daily disposable soft minimizes added care

- d. Fitting tips
 - i. Fit corneal GP vs soft contact lens first
 - ii. Soft lens with central excavation
 - 1. GP sits in excavation
 - 2. May improve GP lens centration
 - iii. Oxygen considerations
 - 1. Due to double barrier (GP + soft), use high O2 materials
- 19. Custom Soft Lenses
 - a. Indications
 - i. Those who can't tolerate a corneal GP
 - ii. Those with mild to moderate corneal irregularities
 - b. Challenges
 - i. Vision may not be as good as that provided by rigid optics
 - ii. Added thickness may lead to corneal hypoxia
- 20. Hybrid Lenses
 - a. Definition
 - i. Rigid center
 - ii. Soft skirt
 - b. Indications
 - i. Those who can't tolerate a corneal GP
 - c. Challenges
 - i. Proper lens movement
 - d. Empirical fitting
- 21. Scleral Lenses
 - a. Indications
 - i. Often the preferred option if irregularity is advanced
 - b. Benefits
 - i. Better comfort than many corneal GPs
 - ii. Better vision than many soft lens options
 - iii. Very customizable
 - c. Challenges
 - i. Some patients have difficulty with lens handling
 - ii. Can be more costly than some other options
 - iii. May induce hypoxia in those with poor endothelial function
 - d. Fitting tips
 - i. Central clearance
 - 1. More if suspect progression
 - 2. Impact of settling
 - ii. Limbal clearance
 - iii. Edge clearance
 - iv. Solutions
 - e. Care and handling