

## **Pediatric Keratoconus Update**

Suzanne W. Sherman, OD, FAAO, Dip ABO  
Assistant Professor of Optometric Sciences (in Ophthalmology)  
Director of Optometric Service  
Columbia University Medical Center  
New York-Presbyterian Columbia  
635 W 165th St, New York, NY 10032  
Tel: (727) 698 1009  
[sw3178@columbia.edu](mailto:sw3178@columbia.edu)

Sharon P. Keh, OD, FAAO  
Assistant Clinical Professor  
SUNY College of Optometry  
33 West 42<sup>nd</sup> Street  
New York, NY 10037  
Tel: (917) 576-2315  
[skeh@sunyopt.edu](mailto:skeh@sunyopt.edu)

### Epidemiology, Prevalence, and Genetics

- Keratoconus - a progressive and asymmetric disorder associated with structural changes in corneal collagen organization
- Classically manifests in the second decade of life, the cornea assumes a conical shape, irregular astigmatism, progressive myopia, thinning, and poor visual acuity.
- 1 per 2,000, however recent epidemiologic findings show 1 in 375, a much higher prevalence
- Leoni-Mesplie et al found 27.8% of patients less than 15 years old had Amsler-Krumeich stage 4 disease compared to 7.8% of patients over 27 years old.
- The largest pediatric population study was done in Lebanon. 2,972 patients were younger than 14 years old.
  - The study reported an incidence of 0.53% (approx 1 in 200) over a 5 year period
  - El-Khoury S, Abdelmassih Y, Hamade A, et al. Pediatric Keratoconus in a Tertiary Referral Center: Incidence, Presentation, Risk Factors, and Treatment. J Refract Surg. 2016;32:534–541.
- Varies among populations
- Hereditary pattern not predictable, positive family histories reported 6%-8% of cases
- Genetic influences: Ethnic differences
  - Finding high volume KCN families and getting the complete family history
  - Involvement of family tree
  - Population trends (Hispanic and Ashkenazi Jewish families)
  - The current plan for genetic testing at Columbia University Medical Center
    - Search for the single strand variant
  - First-degree relatives of patients with keratoconus have a 15-67 times higher prevalence of keratoconus compared to the general population when subclinical forms are taken into account.

### Rates of Progression

- Case example
- Higher rate of corneal collagen remodeling compared to adults, possibly due to weak ectatic lamellae exceeding the capacity of the crosslinking process.
- Review of the pediatric cornea and how it differs from the adult cornea
- More debilitating progression with an increased likelihood of corneal opacities.
- Collaborative Longitudinal Evaluation of Keratoconus (CLEK) Study population - corneal curvature, 24% of cases, < 20-year-old was maximal.

- Known stiffening of the cornea with age (Kotecha et al).
- The rapidity of pediatric cornea evolving questions waiting or signs of progression commonly done for adults or warrants more often follow-up
- Frequent coexistence of ocular pathology (atopy, vernal keratoconjunctivitis (VKC), etc)
  - Fast progression and longer-term keratoconus
  - Discussion of Columbia University Medical Center pentacam screening study in the pediatric allergy population.

### Suspicion/Screening

- Case example
- Unable to achieve 20/20 vision, poor reflex, corneal warping (scissoring of red reflex), high myopia, astigmatism, anisometropia
- Family history
- Other known pathologies (VKC, atopy)
- Scheimpflug tomography (Pentacam, Oculus), Anterior segment optical coherence tomography
- Should we be screening all ophthalmic pediatric patients?

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### Therapy/Management

- Corneal Crosslinking (CXL)
  - Case example
  - FDA approved since 2016 patients older than 14
  - First publication of pediatric population in 2011
  - There are no randomized, contralateral eye studies comparing treatment at diagnosis to treatment upon detection of progression
  - The largest study to date Padmanabhan et al.
    - 194 eyes of 153 children ages 8-19 years underwent CXL for progressives KCN. 142 had standard Dresden protocol and 52 were treated using hypoosmolar riboflavin. CXL was associated with keratometric flattening and stabilization of pachymetry during 6.7 years follow-up. 59 eyes followed longer than 4 years.
  - What we have learned in 10 years of doing cross-linking at Columbia University Medical Center on patients 14+.
  - What we have learned in 5 years of doing cross-linking at Columbia University Medical Center on pediatric patients <14y/o, off label treatment studies.
- Contact lenses

- Soft toric lenses, RGP corneal lenses, hybrid lenses, scleral lenses
- Lens turnover and care

#### Clinical Takeaways

- Importance of screening for high-risk pediatric patients
- Role of Optometrist
- Encouragement/hope