

Title: The Updated Prevalence of Pediatric Keratoconus; Much Greater Than You Thought!

Presenter:

Jennifer S. Harthan, OD, FAAO, FSLs, Dipl AAO

Description:

This 1-hour course will begin with a discussion of clinical cases of undiagnosed pediatric keratoconus. Further discussion will include a review of keratoconus and classic prevalence of the disease based on current literature. An in-depth review of the prevalence of keratoconus in a pediatric population, as found by a large-scale study that screened for the presence of keratoconus on objective metrics from Scheimpflug corneal tomography will be highlighted.

Learning Objectives:

At the conclusion of this course, the participant will be able to:

1. Understand the key history components and clinical features related to keratoconus.
2. Discuss the past and most updated prevalence data for keratoconus in the adult population and pediatric population.
3. Review objective metrics from Scheimpflug tomography to help identify keratoconus and keratoconus suspects in the pediatric population.
4. Understand the importance of identifying keratoconus early in the pediatric population.
5. Identify how to implement screening methods for keratoconus in the pediatric population, particularly those patients showing high risk factors.

Outline:

- I. **Case Examples**
 - a. 10-year-old presents for first eye exam presenting with blurry vision OD>OS
 - b. Has only had school screening
 - c. Entering VAs OD: HM, OS: 20/100
 - d. Diagnosis: KC
 - e. NOT a candidate for CXL
 - f. Why is this important?
 - i. The younger the onset of KC
 1. Poorer prognosis
 2. Faster progression
 3. Increased likelihood of PKP
 4. Higher risk of PKP rejection
- II. **KC Reviewed**
 - a. Progressive corneal ectasia
 - b. Bilateral, asymmetric, and progressive
 - c. Characterized by focal corneal thinning, steepening, and loss of biomechanical strength leading to irregular corneal astigmatism and decreased vision

- d. Genetic conditions, allergy, and environmental factors (e.g. eye rubbing, allergy, asthma, eczema) contribute

III. **Past**

- a. What is the difference between prevalence and incidence?
- a. Reports on the prevalence of KC vary based on the population studied and on how KC was defined.
 - i. Literature reporting on the prevalence of the disease in the adult population ranges between 1:50 to 1:750.
 - 1. Each of the previously published prevalence studies has used slightly different criteria to diagnose KC.
 - ii. Classic KC Prevalence Reference (Kennedy RH, Bourne WM, Dyer JA. A 48-year
 - 1. 1:2,000 – based on registration study in Olmsted County, MN
 - 2. Diagnosis based on detection of scissors reflex with retinoscopy and keratometry
 - iii. Comprehensive review of KC prevalence
 - 1. Netherlands study: Prevalence 1:375 (health insurance data base – topography utilized)
 - a. Godefrooij, D. A.; de Wit, G. A.; Uiterwaal, C. S.; Imhof, S. M.; Wisse, R. P., Age-specific Incidence and Prevalence of Keratoconus: A Nationwide Registration Study. *Am J Ophthalmol* **2017**, *175*, 169-172.
 - 2. Hashemi Meta-Analysis
 - 3. Raine Study
- b. Challenges with previous studies

II. **Present**

- a. Prior to the advancement of corneal diagnostic technology, the literature has reported KC to manifest clinical signs and symptoms during the second to third decade of life.
- b. Prevalence of keratoconus in the pediatric population is not well established
- c. IKA/ICO Pediatric Prevalence Study
 - i. Prospective, observational, single center study
 - ii. Subjects were between the ages of 3-18 years, predominantly low income and primarily two minority racial/ethnicity groups
 - iii. In addition to vision and ocular health assessment, image capture from the Pentacam® (OCULUS Optikgrate GmbH, Germany) tomographer was obtained on each eye
 - iv. Previous literature has reported the prevalence of KC in several populations using only the BAD3 Final D
 - 1. To improve specificity of KC detection using the BAD3 Final D, back elevation at the thinnest point (BETP) was added.
 - 2. Include Table of Criteria Utilized
 - v. Results
 - 1. Six of 2007 subjects were classified as KC, Prevalence = 1:334

2. Three of 2007 subjects were classified as KC suspects, Prevalence = 1:669
3. Nine of 2007 subjects classified as KC or KC Suspect, Prevalence = 1:223

vi. Significance

1. Prevalence found in this study is higher than previously reported highlighting the importance of earlier screening for KC.
2. Early diagnosis of KC with utilization of corneal tomography, recognition of progression, and timely intervention with corneal collagen crosslinking is imperative to prevent progression of the disease, avoid vision loss from KC and resulting impact on quality of life.
3. The frequency of abnormal corneal findings in this population warrants consideration to expand universal screening for KC with tomography, that can detect KC prior to vision loss as part of a pediatric comprehensive ocular examination.

III. Future – Next Steps

- a. Further large scale, multicenter studies are needed to understand prevalence in the general population.
- b. Screening for KC and KC suspects should be part of a routine pediatric eye examination, particularly for those patients showing risks factors such as high astigmatism, reduced best corrected visual acuity, family history of KC or aberrant corneal presentation.