Perspectives on Keratoconus and Pediatric Eye Care Populations

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Optometrists should be at the forefront of the early diagnosis of keratoconus in pediatric populations, but currently the standard of care does not encourage specific diagnostic testing for keratoconus. The American Optometric Association publishes clinical practice guidelines (CPG) as a formal process of patient care recommendations based on current scientific evidence and expert clinical opinion. According to the CPG titled “Pediatric Eye and Vision Examination” the measurement of intraocular pressure (IOP) should begin in the school age child which starts at 6 years old. However, in an otherwise “healthy” pediatric patient, when was the last time that you uncovered or made the primary diagnosis of glaucoma from elevated IOP’s? The prevalence and incidence of primary glaucoma in the pediatric patient population is amazingly small. Actually, the incidence of childhood glaucoma is estimated to be 2.29 per 100,000 patients younger than 20 years old based on a defined U.S. Population study in Olmstead County. Clearly, keratoconus has both a higher incidence and prevalence in the same population. In 1998, Rabinowitz estimated the incidence of keratoconus to be 1 in 2,000. So if we are encouraged to screen IOP on every member of this population, shouldn’t we be at least as aggressive, if not more so, in screening for keratoconus in the pediatric population?

Keratoconus is a bilateral, asymmetric and progressive disease of the cornea that is characterized by localized corneal thinning (anterior and posterior surface elevation changes) with resulting irregularity of the anterior corneal surface that can ultimately induce significant visual distortion. Clinical evidence of keratoconus takes place in the first or second decade of life and commonly progress until about the fourth or fifth decade and it may progress longer. As the disease advances, the ability to see clearly with the use of traditional glasses and contact lenses becomes more and more difficult. At some point along the continuum of the disease, specialty contact lenses can possibly be required in order to “mask” the keratoconic irregularity of the cornea and provide adequate visual function. Many cases, estimated to be over 10% of keratoconus progress to where corneal transplantation, either partial of full thickness, is required due to visually significant scar formation along the visual axis or contact lens intolerance. Although corneal transplantation methods and technologies have greatly improved over the years, it is still a complex surgical procedure which requires many months of recovery and has tremendous impact on activities of daily living. A main goal of management of the individual with keratoconus is outright avoidance of the corneal transplant if possible. Early detection, management and treatment via the newest technology is the least invasive and most effective method to avoid a corneal transplant. It is important to note that a corneal transplant has significant risks such as graft failure, graft rejection, glaucoma, infection, and the likely need for repeat grafts over the course of a patient’s lifetime. All of these risks have the potential to cause blindness.

Over the past decade there has been a significant paradigm shift in the management of keratoconus. With the advent of corneal
Cross linking we can now control and halt the progression of the disease with high degrees of success. As such, early detection of keratoconus has become critically important in order for us to be able to preserve vision and avoid a lifetime of visual compromise and functional challenges. With improvements in cross linking technologies we have treatment methods that have very low risk while providing tremendous benefit for our patients over the course of their lifetime.

Newer and more sensitive diagnostic technologies such as corneal topography, corneal tomography, anterior segment Optical Coherence Tomography (OCT) and corneal biomechanical evaluation among others, has led us to understand that keratoconus is more prevalent than we initially thought. Current estimates, based on expert consensus are as common as 1 in 700-800. With this new technology, we can likely diagnose at pre-clinical stages much earlier in life. Figure 1 shows the Pentacam tomography of a young individual we screened with a parental history of keratoconus at a stage when the child had 20/15 uncorrected visual acuity. Although he had no other clinical signs of the disease with traditional examination, tomography was positive and the child was cross-linked with no progression of keratoconus over the past few years.

Should we consider screening for keratoconus in our pediatric populations? The answer is Yes! This disease and its potential impact on vision and quality of life presents a challenge to eye care practitioners, patients and those who care for our patents. Since we...
can halt the progression of the disease with a safe and effective treatment (Cross-linking was FDA approved in April of 2016), shouldn’t we recommend methods that will allow us to detect keratoconus at the earliest point possible? Again, the answer is, Yes!

If we know all of this information and there is an FDA approved procedure and device, what are the obstacles that are holding us back? With new technology comes new equipment, and that equipment comes with a cost. As with any new technology, there will be acquisition costs for the doctor and procedural fees to the patient. Patients should be made aware that early keratoconus detection is now possible, and doctors should give serious consideration to obtaining the technology now available to detect keratoconus at its earliest stage.

Currently small minorities of eye care providers (ECP’s) have access to the most sensitive diagnostic technologies and, at this point in time, corneal cross linking is typically not covered by medical health insurance. However, Aetna insurance recently announced that they will cover epithelium off cross linking, so the landscape is rapidly changing. Out of pocket expense for the cross linking procedure can currently be in the range of thousands of dollars per eye.

In our practices, we are fortunate to have access to many of the most advanced keratoconus diagnostic technologies and we are involved in corneal cross linking studies that allow us to provide safe and effective methods to halt keratoconus progression. Recently we have instituted a screening protocol for keratoconus detection in our pediatric population beginning at age 8. For children with no other risk factors or development of symptoms we will repeat keratoconus screening every 2 to 3 years until age 30. For individuals with family history of keratoconus we will perform screening annually during that time period.

Our practices are in the small minority, and we acknowledge that challenges are significant in bringing these technologies to the majority of ECPs. We feel that it is time we begin to give thought to this challenge and explore ways to address it in the best interests of pediatric patient care.

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Dr. Press has no disclosures. Drs. Eiden and Morgenstern have done consulting and lecturing for Oculus, manufacturer of Pentacam tomography, and Corvis that measures biomechanical properties of the cornea. Dr. Eiden also consults and lectures for OptoVue, manufacturer of the OCT that incorporates software for measuring corneal properties useful in the early diagnosis of keratoconus.

References


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