

# PEARLS FOR KERATOCONUS SCREENING AND TREATMENT



Greater awareness of the early signs of keratoconus and how best to treat the disease are still needed.

BY KATHRYN M. HATCH, MD

**O**f the many patients with keratoconus seen in my practice, a substantial percentage are teenagers, young adults, and adolescents as young as 8 or 9 years old. In most cases, these patients are referred to me by other eye care professionals for advanced keratoconus treatment, and their diagnosis and referral might have been delayed. If their primary eye care professionals put protocols in place to screen for the signs of keratoconus routinely, it could increase the rate at which keratoconus is identified and treated before the disease reaches an advanced stage.

## AWARENESS

**Screening.** Primary eye care professionals should identify the signs of keratoconus, especially in younger patients, and understand the impact that early intervention has on visual outcomes. Mass screening programs for keratoconus should ideally be performed for every middle school student or by their pediatrician. Until this becomes an attainable goal, every eye care professional should screen patients for keratoconus starting at age 12, especially children and adolescents with changing vision or increasing astigmatism. The earlier keratoconus is diagnosed, the better the chance of optimizing these patients' visual outcomes.

**Eye rubbing.** Increasing public awareness of the impact of eye rubbing and its association with keratoconus, including the weakening of corneal tissue, as well as awareness of the disease should be a priority. Patients with keratoconus who rub their eyes vigorously are likely to cause their condition to progress. Flagging the issue may deter them from continuing the unhealthy habit.

**CXL.** If patients are aware of their progressing keratoconus symptoms, the disease is typically in the advanced stages when vision is altered. CXL is a safe and effective procedure that can stabilize the cornea in this situation. CXL can improve the corneal shape, but it cannot reverse the effects of the disease.<sup>1,2</sup> Providers should therefore either perform CXL on patients with mild keratoconus or refer them for treatment. For some practitioners, this may require a paradigm shift away from waiting until the disease progresses to an advanced stage before initiating CXL treatment. This is in part because most health insurance policies require proof of disease progression before CXL is covered. A better approach to patients with keratoconus is to treat them with CXL as soon as the disease is diagnosed to prevent it from progressing and vision from worsening.

## SIGNS OF KERATOCONUS AND STAGES OF DISEASE PROGRESSION

Patients with keratoconus may not be identified if eye care professionals don't perform routine screenings. In other words, a keratoconus diagnosis can easily be missed if no one is looking for signs during routine examinations.

**Early disease.** Keratoconus may not have affected the vision of young patients yet, but they might have experienced a mild change in their prescription or an increase in astigmatism. It also may not be possible to correct their VA to 20/20.<sup>4</sup> Patients with early stages of keratoconus often describe symptoms such as glare, halos, and double or triple vision that cannot be measured with a vision exam. Further, some patients may squint routinely to see clearly without realizing it or complaining. When patients present with these symptoms and there is no discernible explanation for the changes in their vision—especially if they are young and with no signs of cataract—topography or tomography must be performed to assess them for keratoconus.

**Advanced disease.** The clinical signs of advanced keratoconus are generally more noticeable at the slit lamp. These signs include Vogt striae, Fleischer rings, and visible thinning and bulging in the characteristic cone shape. Distant direct ophthalmoscopy

can reveal an oil-droplet reflex, and retinoscopy can show a scissoring reflex; both are indicative of the disease. In advanced keratoconus, apical scarring and the Munson sign can be seen. These patients may require keratoplasty or the use of scleral lenses or BostonSight PROSE lenses (BostonSight) for optimal vision.<sup>3</sup>

**Disease progression.** The goal is to diagnose and treat keratoconus with CXL before advanced signs of the disease develop. It is hard, however, to predict which patients will experience disease progression. Adolescents and young adults are known to be at increased risk of progression because the body's tissue is more elastic in youth and a level of crosslinking occurs naturally with aging.<sup>4</sup> It is

important to recognize that patients of any age can experience disease progression.

#### THE BOTTOM LINE

Eye care professionals must be aware of the early signs of keratoconus and screen all patients, especially younger ones, for the disease. When keratoconus is identified, patients should be treated with CXL in a timely manner. CXL was approved by the US FDA in 2016. Because it is the only treatment known to prevent keratoconus progression, patients should be informed of CXL and offered a consultation with a specialist who performs it. Eye care professionals who do not perform CXL should refer patients with signs of early

keratoconus to another provider for possible treatment. ■

1. Raiskup F, Theuring A, Pillunat LE, Spoerl E. Corneal collagen crosslinking with riboflavin and ultraviolet-A light in progressive keratoconus: ten-year results. *J Cataract Refract Surg.* 2015;41:41-46.
2. Wittig-Silva C, Chan E, Islam FMA, et al. A randomized, controlled trial of corneal collagen cross-linking in progressive keratoconus: three-year results. *Ophthalmology.* 2014;121:812-821.
3. Davidson AE, Hayes S, Hardcastle AJ, Tuft SJ. The pathogenesis of keratoconus. *Eye.* 28:189-195.
4. Mukhtar S, Ambati BK. Pediatric keratoconus: a review of the literature. *Int Ophthalmol.* 2018;38(5):2257-2266.

---

#### KATHRYN M. HATCH, MD

- Director, Refractive Surgery Service, and Site Director, Massachusetts Eye and Ear, Waltham, Massachusetts
- Assistant Professor of Ophthalmology, Harvard Medical School, Boston
- Member, *CRST* Editorial Advisory Board
- kathryn\_hatch@meei.harvard.edu
- Financial disclosure: Consultant (Glaukos, CXL Ophthalmics)