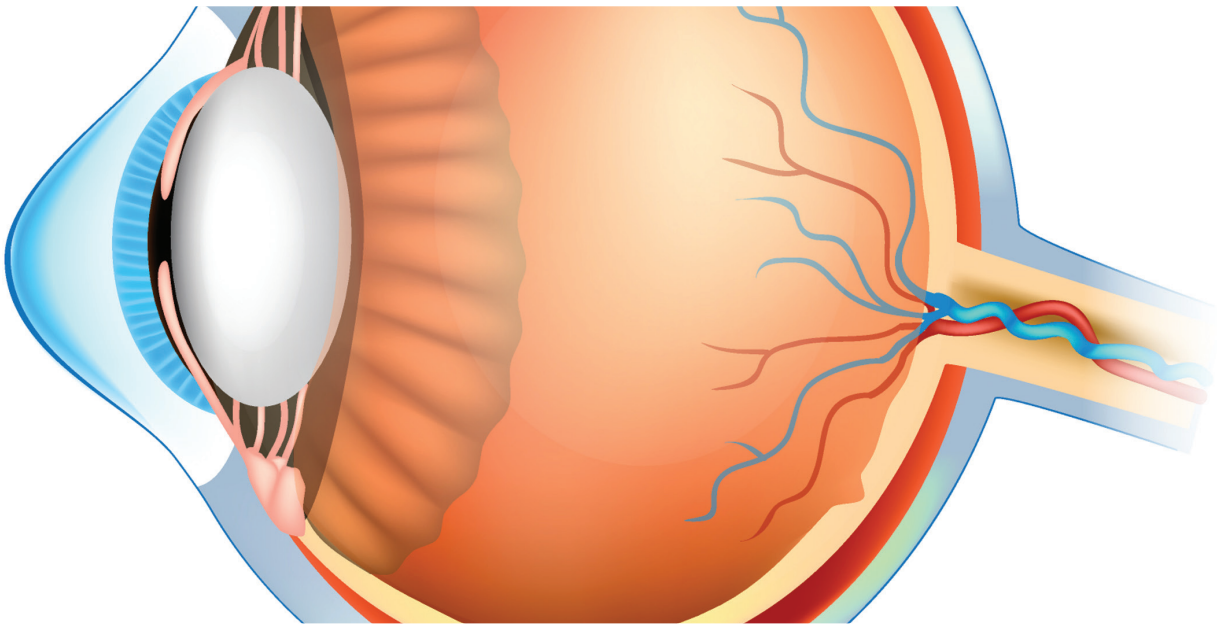

PRESERVING VISION IN PATIENTS WITH KERATOCONUS

As protocols for diagnosis and management evolve, early detection and treatment can help to protect your patients' vision.

BY TRACY L. SCHROEDER SWARTZ, OD, MS, FAAO, AND CLARK Y. CHANG, OD, MS, FAAO, FSLS

Keratoconus has been reported to affect about one in 2,000 people in the United States, but recent studies have shown that the prevalence may be higher in certain populations or age groups.[1-3] Data from a large cohort in Australia's longitudinal Raine Study, for example, found that when Pentacam (Oculus Optikgeräte) Scheimpflug tomography imaging was used to detect keratoconus, the prevalence in 20-year-olds was higher than expected.[1] The authors concluded, "This has important implications for screening individuals at a younger age, so treatment can be initiated before [further] disease progression."

We agree: The best way to preserve vision for patients with progressive keratoconus is to diagnose and treat early, with corneal collagen crosslinking (CXL), before they have lost vision that can't be recovered. Once a patient's vision drops below 20/40, he or she is significantly more likely to require penetrating keratoplasty (PKP).[4]

Unfortunately, it is a common misunderstanding that vision loss is necessary to document progression or to refer a patient for a CXL consultation. Members of a 2015 global consensus Delphi panel, later affirmed by the Cornea Society, noted that the preferred methods for documenting progression (corneal thinning and/or steepening of the anterior or posterior cornea) do not involve visual acuity.[5,6]

CATCH IT EARLY

Advanced tomography remains inaccessible for most primary eye care practices, but optometrists are increasingly investing in topography technologies.

New programs, such as the iDetect KC partnership between Topcon and Glaukos, and devices such as the OPD-Scan III Wavefront Aberrometer (Marco Instruments) can also help to lower the financial entry point for optometric practices to incorporate or upgrade to a new corneal topographer. The same Topcon corneal imaging instruments enable users to voluntarily participate in a keratoconus data registry to help track the progress of patients. In reality, however, advanced

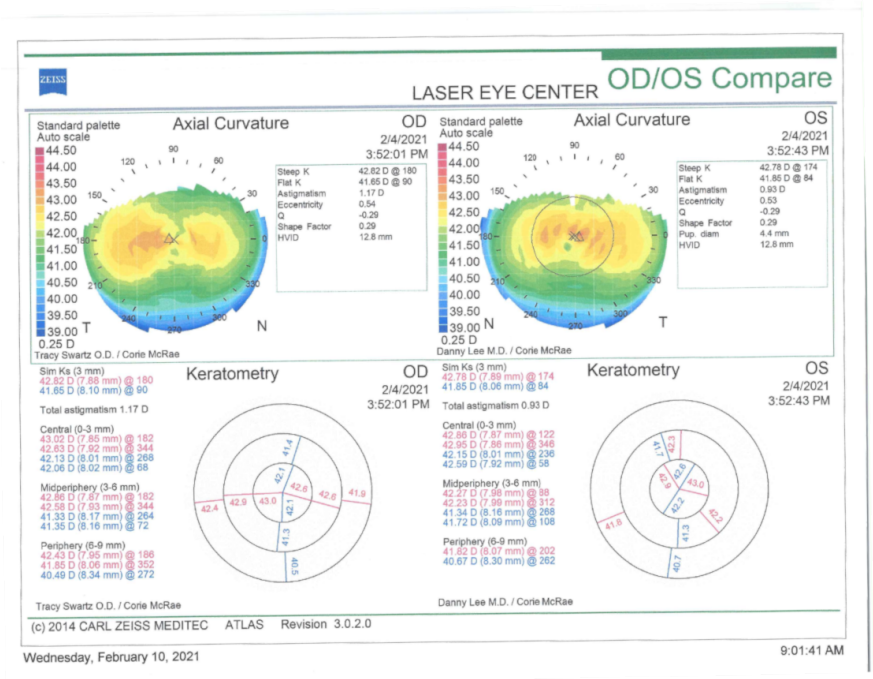

Figure 1. Topography shows an irregular horizontal steepening rather than the typical bowtie pattern (A). Epithelial mapping shows changes inferiorly that may be suspicious for keratoconus (B).

diagnostic imaging tools are only part of the answer. In a recent study in Belgium, where topography and tomography are widely available and patients have access to care within a national health care system, only 13% of 399 patients with keratoconus were detected by age 18.[7] By the time these patients were diagnosed, many had already reached a moderate or advanced stage of disease severity.

WHEN TO REFER

Similarly, in the United States, we see too many people diagnosed with keratoconus only after they have already lost vision. It is important that we increase the level of clinical suspicion of keratoconus to allow more patients with progressive disease to be treated early, preserving their best possible vision.

For primary care practices without access to topography or tomography, a constellation of factors can help to determine when a referral for advanced corneal imaging is needed; these include the patient's symptoms, refractive error, and corneal presentation. Early red flags for keratoconus suspects are listed in *Signs Your Patient May Have Keratoconus*.

Vigilance is particularly important with young patients, as their disease can progress quickly. A 13-year-old patient seen recently by one of us (T.L.S.S.) provides a great example.

The patient's manifest refraction was -3.00 -6.00 X 180° OU. That high astigmatism was cause for concern. Additionally, her topography showed mild irregularity in the astigmatism axes, with less corneal than refractive astigmatism, and mild but suspicious thinning inferiorly on epithelial mapping in each eye (Figure 1). For this patient, we will repeat axial length, tomography, and topography every 6 months to monitor for changes.

If we start looking at kids as keratoconus suspects until proven otherwise, we are more likely to diagnose early and preserve their vision. Topography should be performed annually or more frequently for those at risk of progression (Figure 2).

DON'T DELAY CXL

When treatment choices for keratoconus were limited to specialty contact lenses or PKP, most optometrists managed patients using contact lenses for as long as possible to avoid the costly and invasive transplants. Even when it is successful, PKP requires a lifetime of complex follow-up corneal care, and there is the potential for rejection and repeat grafting.

Today, the treatment calculus is much different. By performing CXL early in patients with progressive keratoconus,

AT A GLANCE

- The best way to preserve vision for patients with progressive keratoconus is to diagnose and treat early before they have lost vision that can't be recovered.
- Topography should be performed annually or more frequently for those at risk of progression.
- Keratoconus can continue to progress in some patients into their 50s and beyond, but younger patients are at higher risk of progression.

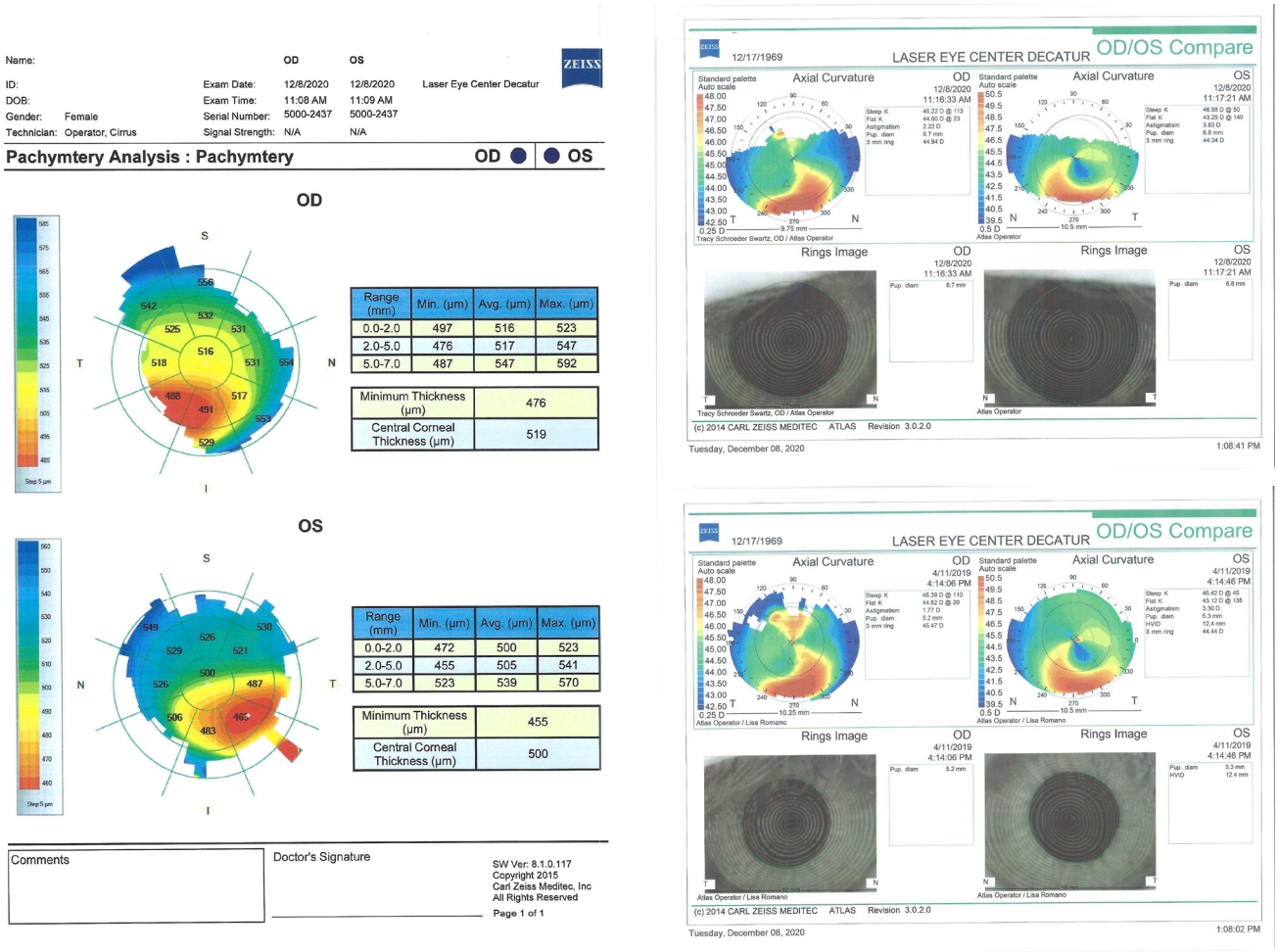

Figure 2. Pachymetry maps from anterior segment OCT can be used to monitor for keratoconus progression without tomography (A). Topography should be performed at least annually, with manual comparison of values such as steep/flat keratometry, corneal astigmatism, and central corneal power (B). This patient showed minimal progression (C).

we can slow or halt the progression of keratoconus, preferably in the mild stage, when patients can be still corrected well with glasses or soft contact lenses.

CXL has been shown to effectively stabilize keratoconus in long-term studies[10-13] and to substantially reduce lifetime economic burden and increase quality-of-life years for patients with keratoconus in the United States.[14] Most studies to date in the United States have been conducted with the standard epithelium-off protocol that is used with the iLink system (Glaukos) for CXL.

Change may be on the horizon, however. In a phase 3 study of an investigational transepithelial, epithelium-on CXL procedure, a 1.00-D difference was reported between treatment and placebo arms in Kmax treatment effect from baseline to 6 months.[15] An epithelium-on CXL procedure, if eventually approved, would be a welcome addition to our treatment armory for keratoconus.

Optometrists should keep in mind that there is no global period for CXL. The OD can expect to get the patient back after CXL and can bill postoperative follow-up visits as office visits, along with serial topography and prescribing post-treatment spectacles or contact lenses when appropriate.

MAKE THE MOST OF WHAT IS LEFT

What about those patients who have already lost vision? For patients who continue to progress, CXL is designed to slow or stop further corneal steepening and the associated visual deterioration. After stabilizing the cornea with the CXL, we have good nonsurgical options to improve visual function with an increasingly wide variety of specialty contact lenses.

In our experience, most patients with keratoconus can achieve better corrected vision with contact lenses, provided they haven't developed corneal scarring or significant warping of the posterior corneal surface. Once patients' keratoconus is stable and the need for frequent contact lens refitting is reduced, not only do they require less chair time to manage, but they are also more likely to report

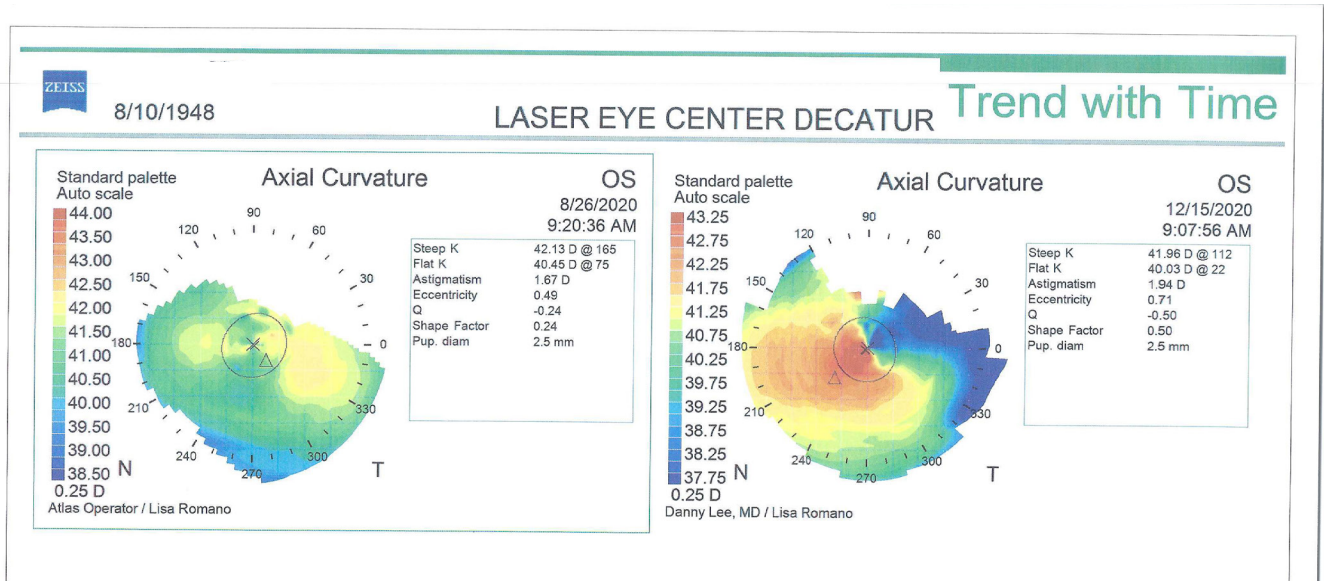

Figure 3. This patient with keratoconus underwent laser cataract surgery. Irregular astigmatism with vision loss (20/30 BCVA) resulted from a limbal incision created to address the corneal astigmatism.

SIGNS YOUR PATIENT MAY HAVE KERATOCONUS

The following factors should be considered early red flags for keratoconus suspects.[8,9]

- Myopic shift or frequent refractive changes beyond what is expected with age;
- High astigmatism or frequent changes in the amount or axis of astigmatism;
- Unexplained quality-of-vision complaints;
- Scissor reflex or an irregular retinal reflex on retinoscopy;
- Placido ring warpage on manual keratometry or topography;
- Family history of keratoconus or corneal transplant;
- Atopic or allergic eye disease or chronic eye rubbing;
- Conditions that have been associated with keratoconus, such as connective tissue disease and Down syndrome;
- Higher-order aberrations (especially vertical coma) and related visual symptoms.

clinical satisfaction and less anxiety about their disease.

It is important to aggressively control ocular surface problems in patients with keratoconus because a healthy tear film is necessary for successful contact lens wear. Lid disease, dry eye, ocular allergies, and other ocular surface disease must be controlled to prevent inflammation and itching. Eye rubbing is a risk factor for keratoconus progression and must be avoided before and after CXL. The use of daily topical antihistamine use may be required to reduce ocular symptoms.

KERATOCONUS THROUGHOUT THE DECADES

Although keratoconus slows or stops progressing in most patients by the fourth decade of life, that is a norm, not a guarantee. We have plenty of patients who have continued to experience progression into their 50s and beyond, so ongoing monitoring is necessary.

As cataracts develop, our expertise in addressing corneal astigmatism associated with phacoemulsification surgery will also benefit our patients with keratoconus. In irregular and keratoconic corneas, limbal relaxing incisions may not lead to predictable astigmatic correction; stable corneal astigmatism may be better addressed using toric IOLs (Figure 3).[16,17]

When you refer patients for cataract surgery, be sure to include their refractive and corneal histories. Patients wearing contact lenses of any kind must discontinue wearing their lenses before evaluation. Because this can be a difficult conversation it

is best to start the discussion early, when the cataract is still developing, so that the patient won't be shocked when discontinuation is requested. In severe cases, this can be done monocularly to allow the patient to function with the one corrected eye.

As our diagnostic and treatment options for keratoconus continue to expand, we can enjoy the opportunity that CXL provides to better preserve the vision of our patients with this condition. ■

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