

UP YOUR KERATOCONUS GAME



Advice from an optometrist who sees patients with the disease regularly.

BY ZACHARY HOLLAND, OD, FSLS

y practice is unusual in that it is comprised primarily of specialty contact lens patients, many of whom are sent by referral. Roughly 50% of my patients have keratoconus or corneas that are in some way irregular. Most optometric practices see keratoconus far less often, which perhaps makes clinicians less alert or less comfortable identifying the early warning signs and taking the next steps to getting treatment for patients. As someone who has seen referrals from many primary care practices and cornea surgeons, I have some advice, which I'll share below.

KEEP KERATOCONUS ON YOUR RADAR

It can be hard to keep a running list of all potential ocular disorders top-of-mind during a busy day when you have 20 or 30 examinations to do. However, ruling out degenerative eye conditions is the primary purpose behind what we all do on a daily basis. That said, I still have patients referred to me with moderate to advanced keratoconus because it is so easily overlooked in the early stages. For those who are committed to proactively identifying this condition, it's important to know the early signs to look out for:

- · Increasing astigmatism
- · Presence of oblique astigmatism
- Maximum keratometry > 47
- · Patient who vigorously rubs their eves
- · Patient with a family member who has keratoconus

For clinicians who offer myopia management, it's important to keep keratoconus high on the differential diagnosis list, as patients can easily be misdiagnosed with progressive myopia.

BEWARE OF ATYPICAL PROBLEMS

By the time patients with keratoconus come to me, they typically have seen several other eye care providers

and feel discouraged. "No one has been able to correct my vision," is a complaint I commonly hear. In many cases, they have been saying for years that their vision isn't crisp. If an otherwise healthy and typical young patient doesn't correct to 20/20, you have trouble correcting their astigmatism with a toric contact lens, or they complain of odd visual symptoms, such as "shadowy vision" or monocular double vision, your suspicion of keratoconus should be raised. Even if your patient's refraction is 20/20, if the refraction doesn't make sense or is abnormally difficult, a topography or tomography analysis (if available) is pertinent. Remember, keratoconus is a condition of change. Every patient with keratoconus begins with a normal cornea that at some point becomes a pathological cornea. During this period of early change, the diagnosis isn't an easy one, but it often begins with a hunch that something just isn't "right."

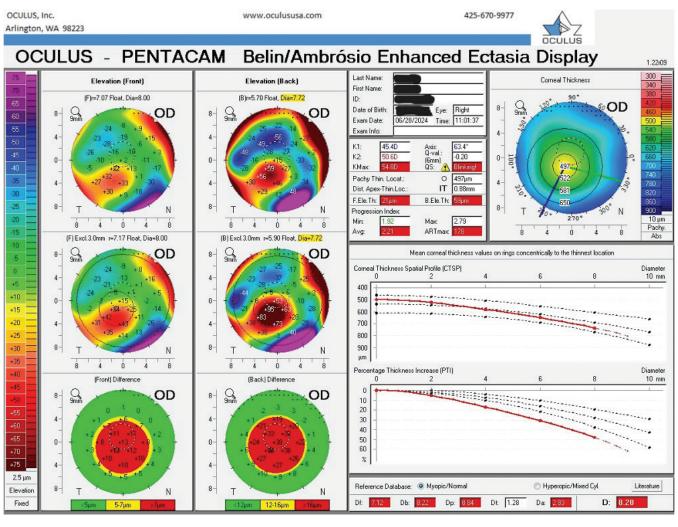


Figure 1. This patient was referred to me for a corneal consult after their primary optometrist suspected keratoconus. Their vision was 20/25 at the time of referral, and they have since had epithelium-off collagen CXL and are now a surprising 20/20 out of spectacles.

WHEN IN DOUBT, GET IMAGING

A lot of ODs may be hesitant to refer a patient to a cornea specialist based on mere mild suspicion. It can take a long time to get an appointment with a cornea specialist, the patient may have to travel, and you may not want to raise concern for a serious eye disease such as keratoconus when you aren't 100% sure. That said, I would still recommend not ignoring that seed of doubt. The threshold to order a topography and/or tomography exam, which can often be obtained from an optometrist in your community, can and should be lower than your threshold for a full cornea consult with a specialist.

Furthermore, imaging can help resolve doubts—or provide a much-needed baseline against which to measure future change. In fact, if your hunch turns out to be correct and the patient does have keratoconus (Figure 1), the sooner you obtain baseline imaging the better, as it may be needed to show disease progression. If the imaging comes back normal, you have done your job and fully evaluated the patient. I have yet to hear a patient complain about me being "too thorough."

SHORTEN THE EXAM INTERVAL

Instead of—or in addition to—getting imaging, doctors can also choose to see

the patient back in 3 to 6 months rather than waiting for their next annual exam. This allows a closer follow-up to detect a changing refraction if there is uncertainty at the previous visit and can truly be sight-saving. One group of researchers found that COVID-related delays of approximately 6 months resulted in continued progression and a 1-line loss of visual acuity, on average.1 Waiting a whole year to confirm your suspicion of a corneal problem could allow unnecessary progression and permanent vision loss for the patient.

DON'T MAKE ASSUMPTIONS

The hurdle to establishing keratoconus disease progression and

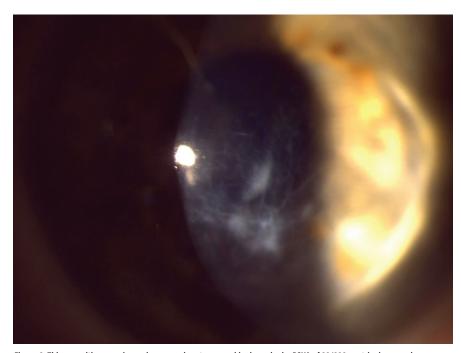


Figure 2. This eye with corneal scarring secondary to corneal hydrops had a BCVA of 20/200 post-hydrops and was recommended for a corneal transplant. The patient sought a second opinion with me, and their VA improved to 20/25 with a scleral lens. Their transplant surgery has been cancelled for now.

qualifying for coverage of corneal crosslinking (CXL) isn't as high as many practitioners think, but documentation of progression is important for insurance to cover CXL. Insurance requirements vary by geographic location and carrier, however, in most cases, a myopic shift of as little as -0.50 D in the manifest refraction spherical equivalent, a -1.00 D change in astigmatism, or an increase in maximum keratometry of 1.00 D within 24 months is often sufficient when it occurs in conjunction with corneal irregularities on topography or keratometry. Referring ODs can document progression on their charts to establish medical necessity.

I find that, too often, patients aren't referred for care until their vision is 20/40 or worse. The fact of the matter is that patients do not have to lose vision to qualify for CXL. Although we can certainly fit them in scleral lenses to improve some of their lost vision, we may have already lost ground in terms of visual potential at that point.

CONSIDER SCLERAL LENSES

Practitioners who don't fit scleral lenses may not be aware of the tremendous advances that have been made in this category, as well as with other specialty lenses in the past 10 years. They are no longer a "long shot" or a "modality of last resort." Although scleral lenses don't treat keratoconus per se (ie, they don't halt the disease process), we can almost always help a patient see better with a specialty lens fit no matter what stage keratoconus they have. In fact, there are many cases where even a patient with late-stage keratoconus who might be considered a candidate for corneal transplantation may be able to postpone or prevent the need for a transplant with just a new scleral lens fit (Figure 2).

Also, don't give up on those who say, "I tried those in the past; it didn't work." There have been many changes to the lens designs and optics that show significant improvements in vision and comfort, even over the past 5 years.

DON'T SWEAT ABOUT WHICH TREATMENT TO PRESCRIBE FIRST

CXL and specialty contact lenses aren't an either/or proposition. For a patient who can't see well with glasses or traditional contact lenses and whose disease is still progressing, both management options are appropriate. And, in many cases, both avenues can be pursued simultaneously.

While we are waiting for documentation of progression, insurance authorization, and an opening in the surgeon's schedule for CXL to stabilize the cornea, we can fit the patient in a scleral lens or another specialty lens to correct their vision. In fact, because a scleral lens rests on the conjunctiva, outside of the central cornea, any minor corneal shape changes that are induced by CXL are unlikely to affect the scleral lens power or fit. There is nothing lost by starting the lens fitting process while the patient is being evaluated for CXL.

BECOME A KERATOCONUS SLEUTH

CXL and specialty lenses are amazing advances that can protect against keratoconus progression and maximize your patients' vision. However, cornea surgeons and specialty lens providers are dependent on the vigilance of primary care optometrists to identify patients with the disease in the course of routine care. Be your patients' hero and find pathology that others might overlook.

1. Shah H, Pagano L, Vakharia A, et al. Impact of COVID-19 on keratoconus patients waiting for corneal cross linking. Eur J Ophthalmol. 2021;31(6):3490-3493.

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