COMPREHENDING KERATOCONUS

The irregular cornea is seen more and more regularly than ever before. Our experts help to get you prepared.

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ON THE COVER
A 57-year-old white male with advanced keratoconus OU and central scarring as a result of the condition. He presented wearing corneal GPs (parameters unknown), but complained that the lenses dislodged very frequently. He was successfully fit with a scleral lens and can now see 20/25 with that correction.
Image and case courtesy of Stephanie Woo, OD, Havasu Eye Center, Arizona

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IN BRIEF

Epithelium-off corneal cross-linking (CXL) is effective in preventing worsening of keratoconus in pediatric patients; however, certain subtypes of the condition may be predisposed to continue progressing despite treatment, reports a study published online in the journal Cornea.1

Crosslinking—which increases the production of noncovalent bonds between collagen fibrils to improve the rigidity of corneal collagen—has previously been demonstrated to prevent the progression of keratoconus in adults.2-4 "To date, however, no controlled trials have been conducted on the procedure’s long-term safety and efficacy in children. Researchers in the Netherlands compiled data from patients under age 18 who underwent an epithelium-off CXL procedure for progressive keratoconus between January 2010 and December 2013 at the University Medical Center Utrecht. The procedure was performed in accordance with the Dresden protocol (30-minute isotonic riboflavin soaking time, 30-minute UVA irradiation, perpendicularly emission plane, 370nm at 3mW/cm²). Uncorrected distance visual acuity (UDVA), manifest refraction and Scheimpflug corneal tomography measurements were taken and a slit-lamp evaluation focusing on atopic/allergic eye disease and eyelid abnormalities was performed prior to the surgery and at one, three, six, 12, 24, 36, 48 and 60 months post-operation. Fifty-four eyes of 36 patients were involved. Results indicated that UDVA improved at all follow-up times with the difference reaching significance during the one-year, two-year and three-year visits. CDVA also improved similarly following the procedure at all time points except the five-year-postoperative visit. Patients also demonstrated improvement in both Kmax and Kavg values: the former improved significantly one year post-treatment with significant improvement throughout the follow-up period. Kavg similarly improved with levels reaching significance at the three- and four-year follow-up visits. “In our pediatric population, epithelium-off crosslinking can be considered as a late finding of VKC in our patients.”


Link Between Axial Myopia and Keratoconus Questioned

There may not be an association between keratoconus and higher levels of axial myopia, despite prior research suggesting otherwise, reports a study published online in the journal Cornea. The connection was first suggested in the 1970s as a way to explain the excessive degrees of myopia seen following penetrating keratoplasty; it was later confirmed by studies that identified significant axial elongation in patients with keratoconus. Of note, however, patients with emmetropic eyes were typically used as the control groups during these studies, which may have led to bias.

Researchers in Belgium set out to repeat prior statistical analyses of the corneal and noncorneal biometry in keratoconic eyes and compare these values to those of healthy eyes with a range of refractions and to a subgroup of emmetropes. Two hundred patients were divided into three groups: those with keratoconus, those with healthy corneas and a subset of the latter with emmetropic eyes characterized by a spherical equivalent refraction between ±0.75D. Uncylcleged refraction was determined using an autorefractometer, while keratometry values were measured using Scheimpflug tomography. The former was used to calculate the SE and Jackson cylinders J0 and J45, while the latter was used to calculate the anterior keratometry Kmax and anterior corneal Jackson cylinders J0,c and J45,c. Axial length, as well as central corneal thickness (CCT) and aqueous depth, were also measured.

“As one would expect, the differences seen for SE, J45, Kmax, J45,c, and CCT,” the researchers report. “Keratoconic eyes also had significantly deeper anterior chambers than the other two groups, but no significant differences were seen for either lens thickness or lens power.”

These results confirm that keratoconus has a minimal influence on axial myopia and that its pathology is purely corneal in nature, the researchers add. Additionally, the increase in anterior depth might be the result of combined thinning and outward expansion of the cornea. “Clinically, these findings may be important to better understand the refractive development in patients with keratoconus, as well as for lens power calculations in patients with keratoconus, where axial length is the most important factor for lens power,” they conclude.


CXL Approved in US
US-based corneal surgeons now have access to collagen crosslinking, following last month’s FDA clearance of the KXL System from Avedro for keratoconus. Three prospective, randomized, controlled, 12-month trials showed a decrease in Kmax among treated patients; untreated eyes continued to progress.

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Look Beyond the Slit Lamp

Are you doing everything you can to make life easier for your keratoconus patients?

This month’s issue of *Review of Cornea & Contact Lenses* highlights the trials and tribulations of managing patients who have keratoconus or another corneal thinning disorder. This group of patients needs our support not only from a clinical perspective, but also in the area of patient advocacy. Though a simple concept in theory, to properly advocate for this group of patients, we need to understand the factors that affect their lives beyond what we see at the slit lamp.

Several quality-of-life studies have demonstrated just how severely these patients’ lives are impacted. Lower quality-of-life scores on the NEI tool in keratoconic patients correlated with binocular visual acuity worse than 20/40. Additionally, steep keratometric readings (greater than 52D) were associated with lower scores concerning mental health, ocular pain tolerance and driving ability.

Unsurprisingly, patient vision in the best eye is the most significant predictor of quality-of-life scores. Additional findings demonstrate that patients in this group also have between category three and category four age-related macular degeneration (AMD), and that overall ocular pain was worse than AMD scores.

"Keratoconus is a disease of relatively low prevalence that rarely results in blindness," one group concluded, "but because it affects young adults, the magnitude of its public health impact is disproportionate to its prevalence and clinical severity." This decline in quality of life continues over time, with a plurality demonstrating a significant decline further on.

**MAKING A DIFFERENCE**

Where then, does advocacy fit in? Support groups for keratoconus patients include the National Keratoconus Foundation and Global Keratoconus Foundation. Both organizations provide resources such as written materials for eye care practitioners and information for patients.

Another resource, the International Keratoconus Academy—a recently-founded organization for eye care professionals—also offers an array of professional education and scientific development on this topic. According to Barry Eiden, OD, the organization’s co-founder and president: “Our mission is to promote and develop the knowledge of keratoconus and other forms of corneal ectasia, and further promote the awareness and understanding of the most appropriate and effective treatment strategies for the management of these diseases.” Those interested in further information can visit the organization’s website at www.keratoconusacademy.com.

**STRENGTH IN NUMBERS**

I’m constantly reminded that we play a personal and vital role in the management and support of every single patient we see daily. Having seen keratoconus and other ectasia patients in my practice for over 30 years, I can’t think of another group that would benefit more from our advocacy and support than this one.

For instance, patients with thinning disorders may need counseling from a mental health professional, as comorbidity beyond mental health is not uncommon. I recently sent a patient with keratoconus and a common case of eyelash ptosis/flabby eyelid syndrome for polysomnography; results from the pulmonologist demonstrated the presence of severe sleep apnea. For patients who suffer from severe ocular allergy—a source of misery unto itself and also a potential contributor to keratoconus progression if excessive eye rubbing occurs—we can help manage their experience of atopy with appropriate referrals.

Additionally, don’t forget the back of the eye—the retina must also receive special attention. A disproportionate number of our CLEK patients had one or more retinal breaks. Without a doubt, I can say I’ve seen more asymptomatic retinal breaks in this group of patients than any other.

Whether you see a large number of keratoconus patients or just a handful, be prepared to address the totality of this group’s needs. They rely on you not only for exceptional eye care, but also as a resource to help them cope with the many non-ocular aspects of their life that are compromised by the disease. Why not start by enrolling in the International Keratoconus Academy and attempting to become a more effective advocate? See where it gets you—and them.

Due to recent improvements in technology, scleral lenses are used more than ever to manage patients with irregular corneas, including presentations of keratoconus and other corneal ectasias. In moderate-to-advanced cases of keratoconus, a scleral lens may be required to help with lens centration.

Scleral lenses also accommodate corneas that are too steep to fit with corneal GP lenses or those that may have failed with other modalities. Vaulting the cornea minimizes risk of central scarring that can result from a poorly fitting corneal lens.

Although scleral lenses are a wonderful option, it is important to keep in mind the associated risks that may present with its use. There is still a large amount of evidence-based research necessary to determine the long-term side effects of scleral lens wear. The question is, however, should all patients be fit with scleral lenses? Is this arguably the new standard of care for this condition?

As part of the 2015 Gas Permeable (GP) lens annual report, 180 doctors were asked about their GP lens prescribing habits, reporting that though the majority of their fits were spherical lenses, one in five lens fits were achieved using scleral lenses (Table 1). Additional results reported scleral lenses were prescribed almost as often as corneal GP designs, with 50% of total fits being corneal GP designs and 44.7% of total fits being sclerals.

Interest in vaulting strategies also continues to represent an area of increasing current and future growth over traditional corneal GP lenses for patients with keratoconus. Trailing behind was the use of intra-limbal, hybrids, custom soft and piggyback lens designs.

Let’s now consider a few modalities to better understand their roles.

**SMALL DIAMETER GPs**

Gas permeable contact lenses continue to be the most commonly used contact lens type in keratoconus management. GP contact lenses mask irregular astigmatism to provide a more uniform anterior refractive surface. They also provide good tear exchange, but may be initially uncomfortable due to lid interaction with the edge of the lens. In mild cases, soft contact lenses or spectacle wear may suffice; however, as the condition progresses, GP lenses are generally required to correct the irregular astigmatism. Often, a corneal GP lens is used unless centration or comfort is compromised, or another lens modality proves more useful.

Three types of fitting relationship exist for corneal GP lenses: apical clearance, apical touch and three-point touch. Apical clearance provides central vault over the apex of the cornea with support of the lens bearing on the paracentral cornea. This condition results from bubbles entering underneath the lens that lead to excessive tear pooling that compresses against the cornea to create divots. These bubbles can result in glare or reduced vision.

Apical touch places all lens support on the apex of the cornea, creating a zone of bearing on fluorescein assessment. Patients report good visual acuity; however, there may be an increased risk of corneal scarring with flat fitting lenses. One study demonstrates similar wearing comfort with both apical touch and apical clearance lens fitting methods. The three-point touch fitting method allows the contact lens to bear lightly on the apex of the cornea, with heavier bearing on the paracentral cornea. This technique is associated with good visual acuity.

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**Table 1. 2015 GP Lens Rx Rates for Keratoconic Eyes**

<table>
<thead>
<tr>
<th>Lens Type</th>
<th>Rx Rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>CONVENTIONAL SPHERICAL GP</td>
<td>43%</td>
</tr>
<tr>
<td>TORIC</td>
<td>7%</td>
</tr>
<tr>
<td>HYBRID</td>
<td>9%</td>
</tr>
<tr>
<td>CORNEAL RESHAPING LENS</td>
<td>11%</td>
</tr>
<tr>
<td>MULTIFOCAL</td>
<td>11%</td>
</tr>
<tr>
<td>NON-SCLERAL GP FOR THE IRREGULAR CORNEA</td>
<td>14%</td>
</tr>
<tr>
<td>SCLERAL</td>
<td>20%</td>
</tr>
<tr>
<td>Source: GPLI</td>
<td></td>
</tr>
</tbody>
</table>
Case Report: How Allergy Can Influence the Course of Keratoconus

Though keratoconus has long been classified as a non-inflammatory disease with a variety of genetic and environmental factors, recent research has demonstrated increased inflammatory mediators in the tears of keratoconic patients, suggesting the pathogenesis of progression may involve chronic inflammatory events.\(^8\)\(^-\)\(^12\) As such, it may be appropriate to consider keratoconus as an inflammatory-related condition.\(^13\) Abnormal eye rubbing secondary to ocular allergic conditions is a commonly proposed pathogenetic factor in for keratoconus, as the action instigates the release of inflammatory mediators that may alter the corneal collagen and lead to corneal ectasia.\(^14\)

**Case in Point.** A 14-year-old Hispanic male presented to the clinic with reduced vision in his right eye that was uncorrectable to 20/20 with spectacle lenses. He reported wearing spectacle correction for myopia since age five, but had noticed the degradation in vision quality in the last two years. Patient history indicated he had been diagnosed with keratoconus at his previous comprehensive eye examination and noted a habit of eye rubbing due to ocular itching with no history of contact lens wear. The patient’s systemic history was unremarkable, as was his family’s systemic and ocular medical history. He denied taking any medications or having allergies to medications.

The patient presented with the following spectacle correction:

**OD:** -3.75 -7.75 x 013 with DVA 20/500, PH 20/100 and VVA RS200

**OS:** -400 DS with DVA 20/20-2 and NVA RS20-2

Corneal topography revealed an asymmetric bowtie with irregular astigmatism and inferior steepening present in both eyes. The overall presentation was worse in the right eye vs. the left (Figure 1). Mire distortion was evident during the patient’s corneal topography evaluation. His best-corrected spectacle distance visual acuity in the right eye was 20/150 and 20/20+2 in the left eye. Pinhole did not improve his vision in the right eye. Additionally, a biomicroscopy examination revealed the presence of Fleischer’s ring in his right cornea and Vogt’s striae in both. No other slit lamp findings commonly associated with keratoconus like stromal thinning, apical scarring or Munson’s sign were noted. No signs of ocular allergy were noted. An examination of the remainder of the anterior segment was unremarkable.

The patient was fit using a corneal GP lens from a fitting set that was designed based on the Collaborative Longitudinal Evaluation of Keratoconus study. The diagnostic lenses used were a tricurve design with an overall diameter of 8.8mm with base curve radii ranging from 7.18mm to 4.50mm and variable optic zone diameters that decrease in size as the base curve steepens. The contact lens powers are variable to provide low minus over-refractions for most of the mild-to-moderate keratoconic patients.\(^12\) The secondary curve is 8.50mm for each
lens, based on corneal curvatures beyond the cone being similar to the corneal curvatures of patients without keratoconus.\(^2\) The center thickness for all lenses is 0.14mm. One drop of proparacaine 0.5% ophthalmic solution was instilled in each eye to alleviate initial lens awareness during the contact lens fitting. Over-the-counter Alaway (ketotifen, Bausch + Lomb) was also prescribed for use twice a day in both eyes. No ocular allergy signs were noted at the time of the exam, though allergy drops were recommended for prophylactic use to minimize future rubbing. Preservative-free artificial tears were also prescribed to be taken as necessary.

**Results.** After a few follow-up visits and minor adjustments to the peripheral curves, the patient’s contact lenses were finalized. Fluorescein pattern assessment demonstrated fluffy central apical touch, with mid-peripheral pooling in the right eye (Figure 2). No bubbles were noted underneath the lens. The lens centered slightly inferiorly, but was picked up with a blink. There was 360 degrees of average peripheral edge clearance when the lens was centered on the eye. There was minimal lens movement on blink; however, movement was not a concern due to lack of corneal staining. The left lens demonstrated an alignment fit centrally with pooling noted superiority at the edge of the optic zone but centered slightly inferior temporally. The lens provided adequate movement on blink, and peripheral edge clearance was minimal at 9 o’clock and average at 3, 6 and 12 o’clock.

**HYBRIDS**

SynergEyes Ultrahealth is a hybrid lens specifically designed for keratoconic patients. When fit properly, these lenses vault the corneal apex to inhibit movement, minimizing mechanical interactions that could lead to scarring. These lenses also provide adequate oxygen supply to the cornea. With hybrid lenses, there is a tear pump under the lens as evidenced by the loss of sodium fluorescein underneath the lens over time.\(^4\) The beauty of hybrid lenses is that the GP center offers optimized vision, while the soft silicone hydrogel skirt centers the lens over the cornea to allow for longer-lasting comfort. Also, the GP center delivers a UVA and UVB blocker to protect eyes from the sun. Hybrids are suitable as a first-line therapy for mild-to-moderate keratoconic patients.

**PIGGYBACK LENSES**

Piggyback systems consist of a corneal GP lens fit over a highly oxygen permeable, low-powered soft contact lens. Low-powered lenses are typically used due to their marginal 20% contribution of marked lens power towards the optics of the lens system.\(^7\) However, now that several other options exist to manage corneal GP lens intolerance, piggyback lenses are not often used, though they can be a good option for centration and stabilization of GP lenses. They also provide added comfort for those suffering from corneal GP lens discomfort by minimizing mechanical eyelid interaction with the GP lens edge. For those who exhibit 3 and 9 o’clock corneal staining, a piggyback soft lens may be the best solution to provide protection to the cornea.

In summary, it is important to look at all the available options and decide when each is best for a given patient, taking into account both the severity of the condition and financial circumstances. In some cases, it may still be best to stick to traditional treatment methods.

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Bringing Clarity to

KERATOCONUS

The visual consequences of keratoconus—which has historically been defined as a noninflammatory condition with hallmark progressive corneal thinning and steepening—can be devastating for many patients.\(^1\) Corneal ectatic disorders often result in decreased acuity, increased ocular aberrations and, in some cases, the need for surgical intervention.\(^1\)

**TYPES OF ECTASIA**

Because *ectasia* describes a physiological finding rather than a particular disease process, it is used in the context of various clinical entities that can arise by mechanical, degenerative or genetic factors.

- **Post-Surgical Ectasia.** Iatrogenic ectasia can occur post-laser-assisted *in situ* keratomileusis (LASIK) or after a photorefractive keratectomy (PRK) procedure.\(^1,2\) Additionally, small-incision-lenticule extraction (SMILE) is a relatively novel refractive procedure that uses a femtosecond laser to remove a disc-shaped portion of stroma with no flap lift, achieving similarly successful results to those of LASIK. In two recent isolated case reports, ectasia following this procedure was documented.\(^11,12\)

  Common clinical findings in patients with post-refractive ectasia include a thicker resulting flap or thinner residual stromal bed than expected. However, post-refractive surgery ectasia can still occur in the absence of these findings.\(^1,6\) Overall, the prevalence of post-refractive surgery ectasia is estimated to be between 0.2% and 0.66%.\(^1,6\)

  For example, (i.e., subclinical) keratoconus is an often-undiagnosed latent form of biomechanical instability in the cornea, which can be induced by refractive surgery and result in post-refractive surgery ectasia.\(^6,8,9\) Other risk factors for iatrogenic corneal ectasia include thin baseline cornea, irregular corneal thickness, high myopia (due to the need for an increased amount of ablated tissue) and young age at the time of refractive surgery.\(^1,2\) In all cases, unilateral presentation of keratoconus contraindicates surgery for the other eye because keratoconus is currently understood to be a bilateral disease.\(^1,4\)

- **Keratoconus.** Considered to be the most common form of ectatic disorders, keratoconus affects at least one out of every 2,000 members of the general population.\(^1,2\) Usually, keratoconus presents in the second or third decade of life.\(^1\) Mandatory clinical findings to diagnose keratoconus include abnormal posterior ectasia, in addition to corneal thinning and abnormal corneal thickness distribution.\(^1,2\) According to the Global Consensus on Keratoconus and Ectatic Diseases, the pathophysiology of the condition likely includes genetic, biochemical, biomechanical and environmental components, but has no primary pathologic explanation.\(^2\) Induced ectasia may be unilateral or bilateral and secondary to mechanical processes in a predisposed cornea.\(^2\)

- **Pellucid Marginal Degeneration.** Considered the second most common form of ectatic disorders, pellucid marginal degeneration presents between the third and fifth decade of life.\(^1\) It is characterized by inferior peripheral corneal thinning and a 'bow-tie' topographical map appearance of the cornea.

**ABOUT THE AUTHOR**

Dr. Morrison is the cornea and contact lens resident at Pacific University College of Optometry. She specializes in medical contact lenses and has a current research interest in gas permeable contact lens design, scleral shape and contact lens solutions.

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By Sheila D. Morrison, OD
Keratoglobus. A rare corneal thinning disorder that is characterized by general thinning and protrusion of the cornea. Keratoglobus may be present early in life, or may be acquired. Like primary keratoconus, the pathophysiologic and genetic etiology of pellucid marginal degeneration and keratoglobus remains unclear.

Today, the majority of patients with keratoconus or other forms of corneal ectasia are managed with specialty contact lenses, such as corneal or scleral rigid gas permeable lenses, rather than surgery. As such, optometrists trained in medical contact lenses are often the first line of care for this affected population. Being knowledgeable about the causes of these conditions can help to enable better clinical decisions and increase the ease of delivering patient education.

BIOCHEMICAL INFLUENCES
Our knowledge about the causes of ectasia has come a long way since the condition’s discovery, largely because of advances in the way we measure the structure and composition of the cornea. Essentially, corneal thinning in keratoconus occurs when corneal collagen degrades. It has been suggested that this degradation could be related to alterations in enzyme activity from normal levels; for example, matrix metalloproteinase (MMP) levels have been shown to be elevated in keratoconic corneas. MMP levels have been shown to be elevated in keratoconic corneas, demonstrating a possible role for these enzymes in collagen degradation. Increased expression of MMPs is widely observed in human tissue in which inflammation is present. Elevated levels of other inflammatory mediators like interleukin-6 and tumor necrosis factor-a have also been found in the tears of keratoconus patients and an imbalance between pro-inflammatory and anti-inflammatory cytokines well. Aquaporins are cell membrane proteins that act as critical water channels for cells, and have been reported to be deficient in the epithelium of keratoconic corneas.

GENETICS
Despite decades of research on the possible genetic causes of keratoconus, its pathogenesis and association with other diseases remains poorly understood. Support for the potential of genetics or the environment playing a role in the progression of keratoconus has been introduced by a series of twin studies describing cases in which both siblings are affected. However, not all twin studies demonstrate evidence of both individuals developing keratoconus. Additionally, some linkage studies report genetic susceptibility related to gene mutation, while others have found no link between keratoconus and mutated genes. It does appear, however, that a positive family history for keratoconus is a risk factor for development of the disease, indicating a possible familial inheritance. Studies have estimated relatives of those with keratoconus to have a risk of 15 to 67 times greater than those with no family history of keratoconus. A recent supported hypothesis also suggests that consanguinity is a significant risk factor for keratoconus. However, to the best of our current knowledge, there are no direct genetic causative factors identified yet.

Interestingly, keratoconus can develop in the absence of certain conditions or in the presence of others. The medical literature documents associations between the disorder and Down syndrome, Marfan syndrome, Ehlers-Danlos syndrome and Leber congenital amaurosis. These correlations could be postulated to relate to the eye rubbing behavior or connective tissue deficiencies that...
commonly accompany many of these conditions.\textsuperscript{17}

Thinning of other connective tissues of the body is not typically seen in keratoconus patients, however; the landmark Collaborative Longitudinal Evaluation of Keratoconus (CLEK) Study indicates no association with connective tissue disease.\textsuperscript{32}

**OXIDATION**

It is known that the cornea metabolizes oxygen to support its physiologic processes.\textsuperscript{17} Keratoconic corneas have been found to exhibit a decrease in levels of antioxidant enzymes such as glutathione, which are involved in the breakdown and elimination of reactive oxygen species.\textsuperscript{17,26-27} Relevant to keratoconus progression, several factors discussed in the literature that may lead to oxidative damage of corneal tissue—specifically, the stroma and Bowman’s membrane—are atopy, mechanical trauma and exposure to ultraviolet radiation.\textsuperscript{28}

- **Atopic Disease and Ocular Allergy.** Atopy is defined as the genetic predisposition to develop allergic diseases like allergic rhinitis, asthma and atopic dermatitis (i.e., eczema). Atopy is typically associated with heightened immune responses to common allergens, especially those that are inhaled or ingested. An allergy is defined as a chronic condition that involves an abnormal reaction to an ordinarily harmless substance that functions as an allergen for that individual. In an allergic reaction, the immune system views an allergen as an invader and directs white blood cells to produce IgE antibodies, which attach themselves to mast cells to trigger a release of potent chemicals like histamine.\textsuperscript{29}

Research suggests prevalence of keratoconus is associated with atopic disease and ocular allergy.\textsuperscript{1,3} Though there is evidence to suggest atopy may contribute to keratoconus, the association is most likely due to the tendency of patients with atopy and/or allergy to chronically rub their eyes to relieve irritation.\textsuperscript{2,30} As such, from a treatment standpoint, patients with coexistent keratoconus and atopic disease should be managed cooperatively by an eye care provider and immunologist.

- **Mechanical Insult.** The CLEK Study, which evaluated the connection between contact lens fitting techniques, apical changes and scarring in keratoconus, suggested that contact lens wear may be a risk factor for increased corneal scarring, implying that inadequate lens fit can negatively affect visual funcion.\textsuperscript{12} Equally important was the finding that because of the adverse effects poor vision can have on daily functionality, keratoconus patients who wear contact lenses (i.e., mostly rigid gas permeable lenses) generally have better scores on mental health and functionality measures than keratoconus patients who do not wear contact lenses.\textsuperscript{12}

As noted above, eye rubbing can trigger unilateral or bilateral induced keratoconus, a cause-and-effect relationship in which mechanical friction on the cornea causes oxidative stress and the production of damaging free radicals within the corneal tissue which may result in corneal thinning.\textsuperscript{10,36} This has been documented in repeated case reports in which patients who rubbed their eyes for reasons including ocular allergy, atopy, compulsive or nervous personality disorder, punctal agenesis (and subsequent chronic watery eye), stress or dry eye subsequently developed keratoconus.\textsuperscript{1-3,30,32-36} As such, it is the responsibility of eye care practitioners to keep modifiable risk factors in mind and educate all patients about the adverse effects of such behaviors. If possible it may also be necessary to educate family members or care providers about the importance of eliminating eye rubbing, as many patients will rub their eyes habitually and sometimes
without awareness that they are doing so.  

**Ultraviolet Light Exposure.**  
Free radicals, a byproduct of oxidative stress, are formed with exposure to ultraviolet (UV) light or when mechanical insult occurs in a tissue. As compared to unaffected corneas, keratoconic corneas have been found to have more damaging byproducts.17 Additionally, keratoconus prevalence and severity has been reported as highest in the Middle East, though no sun exposure study has been performed. It is notable to consider the role of UV in the prevention of keratoconus progression, as it is used to activate riboflavin for corneal collagen crosslinking, which has been shown to strengthen bonds in the cornea. Keratoconus is historically thought to be a noninflammatory disease process that involves thinning of the cornea. However, recent literature from a variety of sources suggests that due to increased MMP and other low-grade inflammatory markers, keratoconus may have an inflammatory component that results in damage to the structural integrity of the cornea. Because of its noninvasive nature, tear film analysis will remain a topic of great interest for the identification of biomarkers present to potentially aid in the etiology and early diagnosis of keratoconus.17

**Discussion**

Without a clear understanding of the reasons behind keratoconus and sleep apnea, diabetes or hormonal changes during pregnancy are also emerging in a number of studies. It remains highly likely that there is a genetic predisposition to keratoconus, which can be triggered by environmental factors related to oxidation. The progressive etiological elucidation of keratoconus and other corneal ectasias will require continued research efforts by vision scientists and clinicians.  

Most patients with keratoconus and other forms of corneal ectasia struggle to overcome visual aberrations induced by irregularity of the anterior corneal surface. Distortions induced by the irregular posterior corneal surface and visual limitations from corneal scar opacities within the visual axis in more advanced cases are other contributing factors. Contact lenses function predominantly to address these visual impairments in two ways: first, by correcting traditional refractive errors like myopia, hyperopia and astigmatism (also known as lower-order aberrations) and, second, by masking the irregularity of the anterior cornea to create a smoother and more regularly shaped anterior optical surface, which can significantly reduce the visual distortions known as higher-order aberrations.

For the most part, though, current contact lenses do not address the residual distortion induced by irregularity of the posterior corneal surface. This article will review current contact lens options with emphasis on how to select the best modality in certain cases. The advantages and limitations of each will also be covered.

**CORNEAL GAS PERMEABLE LENSES**

Rigid corneal contact lenses have been the gold standard of vision correction for keratoconus for over half a century. Though other options have been developed that address the vision needs of the keratoconic patient, these lenses still hold an important place in our contact lens armamentarium. Advantages include good visual rehabilitation via their ability to mask the anterior surface corneal irregularity, ease of handling, excellent tear exchange and oxygenation of the cornea, low rates of infectious keratitis and a decreased rate of inflammatory changes. Primary disadvantages include initial and ongoing discomfort and awareness by many patients, debris and foreign body accumulation under the lenses with associated discomfort and irritation, difficulty with centration of the optics in highly decentered cone apices and the inability of the lens to properly contour the corneal surface without corneal apical bearing in more advanced cases of the disease.

Considerations when evaluating corneal GPs for keratoconus include the location and size of cone, the shape and status of the peripheral cornea and the final visual needs of the patient. Due to highly suggestive evidence that lens bearing on the apex of the cone may increase the rate of scarring, apical bearing should be avoided as much as possible. We typically choose larger overall lens diameters for corneas with larger, decentered cones by first aligning the lens with the corneal periphery and then allowing for adequate tear exchange via appropriate peripheral edge clearance.

Additionally, we need to address the visual needs of our keratoconic patients by not only masking the anterior corneal surface irregularity as completely as possible but also addressing residual astigmatism. Often, we see patients who present in their corneal GP lenses with limited acuity, only to find that a cylindrical overrefraction can improve acuity by a number of lines. When prescribing corneal GPs for keratoconus, practitioners can either custom design a lens or use proprietary corneal GP designs specifically developed to address the topographic requirements of the keratoconic cornea. As a general guideline, we suggest the following for corneal GP design selection for keratoconus:

- When fitting small central cones (i.e., nipple types), use small-diameter multicurve designs with overall diameters of less than or equal to 9mm and optic zone diameters of less than or equal to 7mm.
- When fitting moderately sized and decentered oval cones, use

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moderate-diameter multicurve designs with overall diameters of 9mm to 11mm and optic zone diameters of 7.2mm to 9mm. Intermediate and peripheral curves can be spherical, aspheric or even toric if the peripheral cornea is regularly toric.

- When fitting large and highly decentered “globoid” and “pelucid-like” cones, use larger-diameter intralimbal designs with overall diameters of 11mm to 12.2mm (that typically measure 0.2mm less than the overall corneal diameter), along with larger optic zones (typically over 9mm). Patients wearing these designs can benefit most from asymmetric peripheral geometries, in which peripheral curvatures can be varied along different quadrants or meridians.

**SCLERAL GAS PERMEABLE CONTACT LENSES**

With the advent of highly oxygen permeable gas permeable lens materials, scleral lenses have gained significant popularity over the past decade. The general goal when fitting a scleral lens to virtually any eye—irregular cornea or not—is to completely vault the cornea and land on the bulbar conjunctival surface overlying the sclera. Avoidance of corneal bearing, especially in the}

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**Spectacles Can Still Play a Role**

Above all, the primary goal for correcting vision in patients with keratoconus is to provide clear, functional vision. There are many cases in which spectacles—the most classical form of vision correction—provide excellent outcomes. One of the key diagnostic methods to achieve acceptable acuity with spectacles is to perform corneal topography and observe the power distribution within the pupillary zone. If that pattern is unremarkable in terms of its astigmatic profile, one can expect the manifest refraction to result in good quality visual acuity, assuming that other elements of the ocular system are normal.

Patient success with spectacle wear in cases of keratoconus is limited by various factors of the disease, including adaptation to high astigmatism or significant anisometropia, and frequent changes in refractive results in cases of progressive keratoconus. Practitioners should keep in mind, however, that even in cases in which contact lenses provide better correction for patients with keratoconus, spectacles can still play a role, if only to allow for limited function when contact lenses are not actively being worn. All keratoconus patients should have a back-up pair of spectacles if possible.
limbal area, is critical for success. Additionally, peripheral landing should attempt to parallel the scleral surface without resulting in conjunctival impingement/compression or significant peripheral scleral lens lift or standoff.

When appropriately fit, these specialty lenses provide excellent visual performance and comfort independent of the degree of corneal astigmatism. Scleral lenses can be designed in a variety of sizes to accommodate a host of ocular shapes, and can be fabricated to include anterior surface optics that address residual astigmatism. Some are also available or are being developed in multifocal designs or to address residual higher-order aberrations.

Scleral lenses, however, are not a panacea. There are concerns regarding the oxygen transmission properties of these typically thick lenses, as they are fit with significant corneal vault and minimal tear exchange. We are still not certain what long-term physiological implications could be identified after years of wear on diseased eyes, especially on those patients with compromised corneal endothelial function, such as those who have undergone penetrating keratoplasty.

The scleral lens fitting process has advanced tremendously over the last few years, with the introduction of technologies like anterior segment OCT and various forms of corneo-scleral profile measurement instruments. The ability to “virtually” design scleral lenses without a required diagnostic lens evaluation and the capacity to measure their physical fitting characteristics immediately following dispensing has reached high levels of precision. When the degree of corneal and scleral surface irregularity is substantial enough to resist correction by even a vaulting scleral lens, an ocular prosthetic scleral device may be necessary. These are approximately the same size and fabricated from similar materials as scleral lenses, but are highly customized to address the unique and individual characteristics of more complex corneal cases.

The Prosthetic Replacement of the Ocular Surface Ecosystem (PROSE) device by the Boston Foundation for Sight is one example; fitting this device involves the use of proprietary diagnostic lenses and a software program that measures the shape of the individual patient’s ocular surface.

Another example of a custom-izable ocular prosthetic device is the EyePrintPro by EyePrint Prosthetics, an optically clear prosthetic scleral cover shell that matches the contours of the eye to provide good vision and comfort. When creating this prosthetic, an impression is taken of the patient’s eye at the practitioner’s clinic, then shipped to EyePrint Prosthetics to be digitized by computerized topographical scanners. Using three-dimensional scanning technology and computer-controlled machining systems, an exact match is created to each individual cornea and sclera. In effect, instead of using a series of standardized curvatures to create the lens as is done with traditional scleral lenses, the EyePrintPro is generated to exactly match the unique irregularities of the individual eye. Because of the precise nature of the back surface fit, high quality and individualized
optics such as toric corrections, multifocals, high-order aberration correction and prismatic powers can be placed on the front surface of the device.

**COMBINING SOFT AND RIGID MATERIALS**

Patients and practitioners alike frequently prefer to combine the crisp optics of rigid lenses with the comfort of a hydrogel material. A number of interesting approaches allow us to do just that.

- **Piggyback or Tandem Contact Lens Systems.** This approach traditionally incorporates a corneal gas permeable lens over a soft lens. Advantages of this setup include improved initial and ongoing patient comfort as compared with direct corneal rigid lens wear, potential reduction of mechanical physical compromise to the ocular surface induced by the corneal rigid lens and possible improvement of GP lens centration for improved optics. The soft “carrier” portion of the system...
provides assistance with GP lens centration (especially if higher plus power soft lenses are used, which create a relatively central steeper element to the anterior ocular surface) and a barrier against direct contact between the rigid lens and the ocular surface.

The GP lens portion of the system, meanwhile, provides the necessary optical correction. Any design and optical configuration of GP lens can be used in piggyback/tandem lens systems; however, disadvantages include potential limitation of oxygen transmission through the system (depending upon lens material DK/t and thickness profiles) and the inherent difficulty or inconvenience of using such a system.

Today, the issue of corneal hypoxia has been largely addressed by the availability of multiple contact lens materials with high oxygen transmission properties. Additionally, use of frequent replacement soft lenses (especially daily disposable modalities) as the base carrier in these systems means practitioners can provide a more convenient and healthy option for piggyback/tandem lens wear as compared with options in the past (Table 1). In cases of corneal irregularity like keratoconus, the fitting characteristics of the soft lens may be influenced by the modulus of the material used. Higher modulus lenses may at times result in edge fluting with associated lens awareness or discomfort. As such, high Dk/low modulus lens designs are often preferred.

The fitting methods of these systems are typically quite simple and result in use of GP lenses with the same fitting and vision parameters as would be used when fitting the GP lens directly on the cornea. The exceptions to this would be cases in which high-power soft lenses are used to influence lens centration. It has been estimated that only 20% of the soft lens power manifests in a piggyback/tandem lens system; as such, since low power soft lenses are typically employed most, their influence on the total power of the system is quite insignificant.

In addition to the use of conventional disposable soft lenses as a carrier for the GP lens in piggyback/tandem lens systems, it is occasionally advantageous to consider custom soft lenses that have a recess or cut-out in the anterior surface in which the GP lens can be placed. Recessed soft lenses help better hold the GP lens in place for optimal centration; as such, in this case, the soft lens provides the system's centration and movement, while the GP provides the optics. Examples

### Table 1. Soft Lenses Typically Used in Piggyback/Tandem Contact Lens Systems

<table>
<thead>
<tr>
<th>Design</th>
<th>Dk/t</th>
<th>Modulus (MPa)</th>
<th>Replacement Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Night &amp; Day</td>
<td>175</td>
<td>1.50</td>
<td>Monthly</td>
</tr>
<tr>
<td>Acuvue Oasys</td>
<td>147</td>
<td>0.2</td>
<td>Two-Week</td>
</tr>
<tr>
<td>Biofinity</td>
<td>160</td>
<td>0.75</td>
<td>Monthly</td>
</tr>
<tr>
<td>Ultra</td>
<td>163</td>
<td>0.70</td>
<td>Monthly</td>
</tr>
<tr>
<td>Dailies Total 1</td>
<td>151</td>
<td>0.70</td>
<td>Daily</td>
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<tr>
<td>MyDay</td>
<td>100</td>
<td>0.40</td>
<td>Daily</td>
</tr>
<tr>
<td>AcuVue Oasys One-Day</td>
<td>121</td>
<td>0.96</td>
<td>Daily</td>
</tr>
</tbody>
</table>

**Fig. 4.** An anterior segment optical coherence tomography scan of a scleral contact lens on the eye.
of recessed or cut-out soft lenses include the Flexlens Piggyback (X-Cel) and the Recessed Pillow Lens System (Fusion Technologies and EyeVis Vision Research).

- **Hybrid Contact Lenses.** Based on the initial concept introduced by piggyback/tandem lens systems, hybrids are single lenses that combine the attributes and advantages of both soft lenses and rigid gas permeable lenses. The Saturn lens, introduced in 1984, was the first hybrid design; since then, various developments in hybrid lens technology have led to the contemporary version now manufactured and distributed by SynergEyes. Today’s hybrid lenses are fabricated from high oxygen transmission materials and constructed to avoid the fragility of the junction between the soft skirt and rigid center of former hybrid designs.

  Along with advances in materials and manufacturing improvements, hybrid lens designs themselves have also moved forward, with more indications than ever. This includes their use on irregular corneas, with the most current lens being the UltraHealth lens (Synergeyes). The UltraHealth lens is a reverse geometry “vault” design configured to leap the apex of the keratoconic cornea. When appropriately fitted, the lens centers fairly well and maintains adequate movement during wearing time. It also provides acceptable comfort and good visual performance. An oblate design version of the lens, known as the UltraHealth FC, was also recently introduced. This reverse geometry design is most appropriate for corneas characterized by oblate topography, which is most commonly found in patients who have undergone refractive surgery or penetrating keratoplasty. However, some practitioners have also used this design to successfully fit certain cases of keratoconus via incorporation of the more pronounced reverse curve as a way to “lift” the lens in order to achieve adequate apical vault.

  Contraindications for hybrid lens wear for keratoconic patients include scenarios in which appropriate physical fitting characteristics cannot be achieved and thus results in one or more of the following: apical bearing, lens landing areas with excessive bearing/compression or lack of movement either initially or after a period of wear. These issues are more commonly present when the cone is either too advanced in elevation or curvature and/or when the cone is significantly decentered. Thus a rule of thumb is that those cones that are mild to moderate and more centralized tend to respond much better to wear of vaulting hybrid contact lenses. Optical limitations can also present with hybrids when significant residual astigmatism is found due to the lack of availability of anterior toric options at this time, so this must also be kept in mind when evaluating a patient.

  **All in all, keratoconus is a challenging disease that presents in a wide range of severity and visual disturbances. A detailed understanding of the phenomenon and extensive clinical experience in its management allows the practitioner to tailor the most appropriate vision correction solution for each patient. Contact lenses have been and continue to be the mainstay for this patient population; fortunately, both old and new lens options abound. Integrating the most appropriate contact lens treatment with a comprehensive medical and surgical approach will allow the keratoconic patient to receive the highest quality of care, which should result in optimal vision along with appropriate control of the disease.**
Keratoconus patients typically experience wide-ranging visual fluctuations and disease progression from onset, typically at puberty, until the fourth decade of life, during which the condition becomes relatively stabilized. Though symptoms of visual aberrations are unwelcome at any point during one’s lifetime, the manifestations during these formative years are especially inopportune.

While refractive correction with gas permeable contact lenses is often the standard of care in keratoconus, management during the condition’s progressive years necessitates frequent contact lens adjustments, resulting in a significant reduction in quality of life and a substantial long-term economic burden for affected patients.^1^ According to conventional evidence, up to 20% of cases may go on to require penetrating keratoplasty.2^ To date, despite steady advancements in contact lens technologies, keratoconus still remains the most common cause for such surgical intervention.

What if these patients could have their vision stabilized years or even decades earlier? Such is the game-changing potential that corneal crosslinking (CXL) represents. Despite having only recently obtained FDA clearance in April 2016, CXL with ultraviolet-A (UVA) and riboflavin photosensitizer has already been implemented around the world as a first-line treatment for keratoconus since the late 1990s (Figure 1).^3^-^6^ Its ability to stiffen corneal tissue makes CXL the first treatment to stabilize underlying stromal weakness and halt or slow corresponding progression of ectasia. Additionally, a global Delphi panel comprised of representatives from each of the four supranational corneal societies—the Asia Cornea Society, the Cornea Society, EuCornea and PanCornea—recently published a consensus report that recognized the importance of incorporating CXL as part of the new standard of care in managing keratoconus and ectatic diseases.7

CORNEAL BIOMECHANICS INFLUENCE SHAPE

The ideal outcome for a patient with keratoconus is to arrest the condition’s continual progression before visual function is compromised. Hence, the potential benefits of early intervention with corneal crosslinking have sparked resurgent clinical interest in exploring diagnostic instrumentation that could more easily allow for early disease detection.

Recent investigative work in the field of corneal biomechanics may hold the potential for earlier identification of patients who could benefit from CXL, and could also aid in the analysis of CXL outcomes to further improve treatment parameters. These research findings have also provided insights into the implications of redistributing biomechanical stress in the cornea, with the intention of not only stabilizing the disease but also normalizing corneal shape.10^-^13^ The stiffness of the human cornea relies upon the lamellar organization of the stromal collagen fibers, which are regulated by an interconnecting network of proteoglycans.8 While there are still unanswered questions about the precise combination of molecular, genetic and environmental factors that contribute to the pathogenesis of keratoconus, it is believed that the interaction of these factors leads to the loss or slippage of collagen fibrils and changes to the extracellular matrix in the corneal stroma.9

Studies using x-ray scattering techniques reveal a disorganization of the collagen lamellae in the region of the cone, with a more normal organization of collagen in the surrounding regions.10^ While studies from the 1970s and 1980s revealed bulk abnormalities in mechanical strength

SEARCH FINDINGS HAVE ALSO PROVIDED INSIGHTS INTO THE IMPLICATIONS OF DISTRIBUTING BIOMECHANICAL STRESS IN THE CORNEA, WITH THE INTENTION OF NOT ONLY STABILIZING THE DISEASE BUT ALSO NORMALIZING CORNEAL SHAPE.10^-^13^ THE STIFFNESS OF THE HUMAN CORNEA RELIES UPON THE LAMELLAR ORGANIZATION OF THE STROMAL COLLAGEN FIBERS, WHICH ARE REGULATED BY AN INTERCONNECTING NETWORK OF PROTEOGLYCANs.8 WHILE THERE ARE STILL UNANSWERED QUESTIONS ABOUT THE PRECISE COMBINATION OF MOLECULAR, GENETIC AND ENVIRONMENTAL FACTORS THAT CONTRIBUTE TO THE PATHOGENESIS OF KERATOCONUS, IT IS BELIEVED THAT THE INTERACTION OF THESE FACTORS LEADS TO THE LOSS OR SLIPPAGE OF COLLAGEN FIBRILS AND CHANGES TO THE EXTRACELLULAR MATRIX IN THE CORNEAL STROMA.9

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of keratoconic corneas relative to normal eyes, more recent work based on biomechanical modeling and Brillouin optical microscopy further demonstrates that these abnormalities may be attributed to focal weakening over the region of the cone, rather than across the entire cornea.\textsuperscript{11-13}

The focalized reduction in elastic modulus within the affected corneal region deforms to a greater extent than the surrounding tissue when subjected to the strain of the normal intraocular pressure, manifesting in the conical protrusion. A proposed biomechanical cycle of decompensation suggests that the initial pathological changes triggered by genetic predisposition and environmental factors result in this focal weakening of the cornea.\textsuperscript{11} The initial asymmetry in elastic modulus is thought to initiate a cycle of stress redistribution in which the focal thinning results in increased biomechanical stress, leading to deformation and a further increase in stress, driving disease progression.\textsuperscript{11} If such theories are borne out by future research, a comprehensive assessment of corneal stress distribution would be essential to understanding the biomechanical factors at play in the progression of the ectasia and the efficacy of treatment options.

Currently, two commercially available devices measure corneal biomechanical properties by analyzing corneal behavior in response to a pulse of air. The Ocular Response Analyzer (Reichert) measures the difference in tissue response between in- and outgoing applanation pressures to provide a measurement of energy loss due to viscous damping. This clinical parameter is represented as corneal hysteresis (CH). Another device, the CorvisST (Oculus), uses a high-speed Scheimpflug camera to evaluate the deformation of the cornea in response to these pressures.\textsuperscript{14}

Though both of these systems provide information about the overall biomechanical properties of the cornea, neither is able to map the regional differences in these properties. As such, the sensitivity of the devices may be insufficient to diagnose early or forme fruste keratoconus, due to the overlap in the range of values for normal and weakened corneas. Additionally, the absence of spatial information limits the applications of these systems in determining focal weakening of the cornea or measuring regional effects of corneal crosslinking within a treated cornea.\textsuperscript{14}

Several techniques are in development to address the potential benefit of spatial measurement of corneal tissue properties, including supersonic shear imaging, Brillouin optical microscopy and optical coherence elastography. All hold promise to aid in the development of patient-specific corneal crosslinking procedures through improved understanding of regional differences in corneal stiffness.

**CROSSLINKING PARAMETERS IMPACT CLINICAL OUTCOME**

When crosslinking was first introduced, a single treatment approach was applied in all cases. Termed “the Dresden protocol,” this conventional technique is performed following removal of the central 7mm to 9mm of epithelium. The stroma is saturated with riboflavin for 30 minutes, then irradiated with 365µm UVA at 3mW/cm\textsuperscript{2} for another 30 minutes for a total UVA dose of 5.4J/cm\textsuperscript{2} (Figure 2). Additional riboflavin drops are instilled at five-minute intervals during the irradiation phase.\textsuperscript{15}

Several randomized, controlled trials have demonstrated statistically significant improvement in maximum keratometry (K\textsubscript{max}) or cone apex power in CXL-treated eyes compared with the untreated controls.\textsuperscript{16,17} Progression of K\textsubscript{max} of 2D or more has been observed in 0% to 4.3% of CXL-treated eyes.\textsuperscript{16-18} A long-term study of patients treated with the conventional protocol reveals persistence of the treatment effect through a 10-year follow-up period.\textsuperscript{19}

Fig. 1. Peter Hersh, MD, of the Cornea and Laser Eye Institute performing the conventional (i.e., Dresden) corneal crosslinking procedure.

Fig. 2. Slit lamp photograph illustrating riboflavin in the corneal stroma following the loading phase.
Though further investigation is still necessary, significant advancements have been made in the last decade in regards to understanding the photochemical mechanisms that result in the formation of new crosslinks in the cornea. Under the right conditions, the interaction of UVA and riboflavin sets off a complex chain of photochemical reactions, resulting in the formation of covalent bonds within the intracellular matrix of the collagen lamellae, which effectively stiffens the cornea in the treated zone.\(^5\) This increase in stiffness may break the cycle of biomechanical weakening that results in ectasia, further limiting progression of the disease.\(^11\)

With an improved understanding of corneal biomechanical behavior and the mechanisms of CXL comes a wave of new clinical efforts aimed at optimizing the procedure’s treatment parameters. Namely, this ongoing research will focus on deriving new CXL delivery protocols to decrease treatment time, increase both intra- and postoperative comfort, and improve efficacy, which could result in more meaningful corneal reshaping in addition to disease stabilization.

One such modification is the introduction of transepithelial crosslinking, which is intended to improve patient comfort and minimize infection risk.\(^20\) While standard formulations containing 0.1% riboflavin and 20% dextran show minimal penetration through an intact epithelium, new riboflavin formulations with added corneal-enhancing compounds like BAC and/or EDTA, can improve riboflavin diffusion despite epithelial presence.\(^21-23\) Attempts to disrupt the epithelial tight junctions without debriding the epithelium have also been carried out using disruptive devices, surgical sponges or iontophoretic delivery through the use of a mild electrical current.\(^24-26\)

Another procedural modification is the use of accelerated CXL, which uses higher irradiance UVA (7mW/cm\(^2\) to 43mW/cm\(^2\)) to shorten the amount of time necessary to deliver the equivalent total energy dose (Figure 3). Clinical studies evaluating the efficacy of accelerated CXL demonstrate stability or flattening of \(K_{\max}\) comparable to results achieved with the Dresden protocol.\(^27,28\) However, some investigators have examined the stromal demarcation lines following accelerated CXL—interpreted as an indirect indicator of the relative CXL treatment depth, commonly observed in the first one to three months post-op—and reported a trend towards shallower depth of the line following accelerated vs. conventional techniques.\(^27,29-32\)

This difference in the demarcation line depth may be modulated by oxygen bioavailability within the stroma. Oxygen levels in the cornea are depleted by the photochemical reactions of CXL, with rapid oxygen replenishment once the UVA source is removed.\(^33\)

One method to increase oxygen concentration in the cornea during accelerated CXL is to program the UVA emission to turn on and off at repeated time intervals to allow for diffusion of oxygen into the stroma during pauses in UV exposure.\(^34\) Hence, pulsed irradiation may have the potential to increase the corneal stiffening effect obtained with the same UVA dose, and may potentially lead to a reduction in procedure time by increasing the treatment efficiency of high irradiance CXL. Preliminary results with pulsed irradiation are promising, indicating safety equivalent to the continuous UV exposure protocols, albeit with greater demarcation line depth.\(^35,36\) However, longer follow-up is necessary to determine whether demarcation line depth significantly impacts clinical outcomes.

Interestingly, this correlation between the depth of the de-
marcation line and the different CXL protocols applied suggests a possible opportunity to customize CXL treatment parameters to target a specific depth—for example, to accomplish a shallower CXL effect in cases of thinner corneas by varying irradiance, total dose or pulse interval.37

CUSTOMIZED CROSSLINKING TO RESHAPE THE CORNEA

Though corneal flattening and visual improvement have been reported, the primary goal of conventional CXL protocols is in fact to stabilize the cornea against progression and prevent further visual loss (Figure 4). Conventional CXL achieves this effect by uniformly stiffening the central 9mm of the anterior stroma. With the new understanding of the distribution of biomechanical forces within the keratoconic cornea comes the potential to customize crosslinking treatment plans based on an individual patient’s characteristics and corneal topography to achieve not just stabilization but also improvement in corneal shape.

In addition to varying the depth of crosslinking by modifying parameters like UVA irradiance, total energy dose and/or pulse interval, the lateral distribution of crosslink formation in the cornea may be better controlled through the use of customized UVA illumination patterns that would allow surgeons to focally stiffen the weakest region of the cornea rather than the conventional approach of uniformly stiffening the entire central cornea.12 Three-dimensional modeling of this concept suggests a potential for greater normalization of the cornea (inferior flattening and superior steepening), presenting the opportunity to combine visual rehabilitation and biomechanical stabilization into a single procedure. Photorefractive intrastromal corneal crosslinking (PiXL) is the clinical product of this theoretical approach.

PiXL uses a topographically customized UVA pattern to induce a variable distribution of crosslinking in the stroma (Figure 5). The CXL treatment depth (Z-axis) is controlled by the UVA energy parameters, while the lateral aspect (X-axis and Y-axis) is controlled by the specific UVA pattern applied using a crosslinking system containing a digital micromirror device (a semiconductor comprised of adjustable microscopic mirrors that can create a desired beam profile).13 The first clinical case report of PiXL application in a keratoconus patient used the KXL II System (Avedro) and demonstrated improvement in uncorrected visual acuity from 20/40 to 20/25 and reduction in corneal astigmatism of 0.8D at six months post-op.39

Though PiXL is not yet available in the United States, a number

Fig. 4. A case example of topographic flattening after corneal crosslinking. Preoperative axial topography is shown at left, three-month postoperative topography at center, and a difference map revealing the change between the two time points is shown at right.

Fig. 5. Customized, dual-arc UVA treatment pattern applied to the central cornea of a patient with intracorneal ring segments, using the KXL II system.
Corneal cross-linking (CXL) has revolutionized keratoconus management by targeting underlying corneal instability and successfully stopping or slowing down disease progression. This innovation has energized research efforts to better understand the mechanisms driving keratoconus progression and to develop new diagnostic instrumentation to allow for earlier diagnosis. In addition, the next generation of CXL protocols is expected to attempt to further normalize the irregular contour of treated corneas and potentially improve patients’ visual function.

The author thanks Grace Lytle, OD, for her contributions to this article. Dr. Lytle is employed by Avedro, the manufacturer of the KXL crosslinking system.

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The structural instability brought on by corneal ectasia and thinning makes keratoconus notoriously difficult to manage long-term, as the cornea becomes—literally—a moving target. Visual fluctuations and Rx changes in response to corneal shape change are common.

Treatments for keratoconus have significantly advanced and diversified since the disease was first called to attention by Burchard Mauchart in 1748. First-line solutions include spectacles and contact lenses, with one caveat: soft lenses may be adequate to correct both myopic and astigmatic errors early in the disease process, but rigid gas permeable lenses will often become necessary as it progresses and corneal irregularity increases. Newer customizable lenses—such as sclerals, Rose K or Boston lenses—offer higher oxygen permeability and better comfort for these patients. However, in some cases, simple vision correction methods may not always be adequate. What other choices do patients have then?

Curtailing Progression

Ultraviolet A riboflavin-mediated corneal collagen crosslinking (CXL) may help prevent keratoconic disease progression in patients who exhibit a clear central cornea and minimum corneal thickness of 400µm. This surgical technique involves the biomechanical strengthening of the collagen fibers of the cornea by increasing the collagen crosslinks. Evidence for its efficacy and safety continues to be reviewed. Available globally since the late 1990s, the procedure has just received FDA clearance for use in the US.

Other surgical interventions should be considered when corneal irregularity is so great that spectacle or contact lens wear is no longer tolerated, when central scarring is limiting vision, or when significant corneal thinning may preface a possibility of traumatic perforation (a rare but devastating potential consequence of steroid use). Other FDA-approved surgical options include intrastromal corneal ring segments (ICRS) and corneal transplantation.

ICRS are minimally invasive plastic polymer arc-shaped segments inserted into the cornea of keratoconus patients with mild cases of irregular astigmatism and preserved corneal thickness. Successful implantation requires a 6mm diameter zone of central cornea that measures more than 450µm in thickness. The ring segments are inserted into a pocket that can either be made manually or with a femtosecond laser. The incision is made based on the steep axis, as determined by corneal topography.

The goal of ICRS is to reduce corneal steepening as well as irregular astigmatism, so that patients can use contact lenses or spectacles with improved tolerability. Long-term research has demonstrated improvements in corrected distance visual acuity (CDVA) and spherical equivalent and a reduction of topometric astigmatism over the first few years in 60% of patients. Note, there does exist a possibility for clinical regression over the long term, as this treatment modality does nothing to blunt the progression of the disease, based on patients followed over five years. Currently, ongoing studies are evaluating the potential benefit of combining ICRS with corneal crosslinking as ultimately—even after ICRS implantation—the patient’s disease may continue to progress and eventually require corneal transplantation.

About the Authors

Dr. Behshad is a clinical instructor at the Gavin Herbert Eye Institute at UC Irvine California. His research interests include the use of the femtosecond laser for corneal surgery as a method to improve endothelial transplantation techniques.

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Dr. Farid is director of the cornea, cataract and refractive surgery department and vice-chair of ophthalmic faculty at the Gavin Herbert Eye Institute at UC Irvine, where she also serves on the residency education committee and is director of the cornea fellowship program. She also serves as an associate medical director for Sight Life Eye Bank.
ADVANCED STAGE OPTIONS

Patients with more pronounced presentations of keratoconus who are unable to tolerate contact lenses, who have significant scarring on the central visual axis or who fail to improve in visual acuity with corneal crosslinking or ICRS may require keratoplasty.

The choice of employing lamellar vs. full-thickness keratoplasty will depend on the extent of corneal scarring. Types of corneal transplantation techniques include conventional full-thickness penetrating keratoplasty (PKP), deep anterior lamellar keratoplasty (DALK) and femtosecond-enabled keratoplasty (FLEK), which can be performed as either a full-thickness graft procedure or in combination with the DALK procedure (known as fs-DALK).

In conventional full-thickness PKP, manual trephination can lead to misalignment of host and donor tissue, which may ultimately heal in such a way as to limit visual outcome.\(^9\) Patients most likely to benefit from a conventional PKP rather than a FLEK operation include those who possess a mechanical issue like a history of glaucoma implant surgery with bleb, or anterior staphyloma that prevents the laser interface from having direct contact with the corneal limbus due to the irregular ocular surface. As such, contraindications to FLEK include a history of glaucoma filtering surgery, the presence of an aqueous drainage device or narrow palpebral fissures.

The advent of the femtosecond laser has allowed for greater precision of wound architecture in both the host and donor tissue, allowing for quicker healing of the incision, improved approximation of tissue and quicker visual recovery with FLEK.\(^10\) During the procedure, the femtosecond laser pushes microcavitation bubbles to a precise corneal depth through photodisruption. These bubbles combine in a manner that results in planar cuts of tissue, a technique that was originally pioneered for use in laser in-situ keratomileusis (LASIK) flap creation.\(^5\) Various configurations for keratoplasty incisions have been designed with the femtosecond laser, including but not limited to “top hat,” “mushroom” and “zig-zag” (Figure 1).

Evidence has demonstrated these wound configurations allow more surface area for tissue healing—thus decreasing rates of wound leakage and enabling faster visual recovery—lower levels of postoperative astigmatism and earlier removal of sutures (Figure 2).\(^11\-17\)

Although no head-to-head comparisons have been conducted between the various configurations of wound architecture, the zig-zag-shaped incision appears to have some advantage over the others, as it leaves room for a smoother graft/host interface transition as well as decreased vertical and torsional misalignments due to its ability to create a hermetic wound seal (Figure 3).\(^10\)

Corrective lenses can serve patients well for years, but a lasting solution may require a trip to the OR.

By Soroosh Behshad, MD, Priscilla Q. Vu, and Marjan Farid, MD

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**Fig. 1. From top to bottom:** schematic of “top-hat” incision; schematic of “mushroom” incision; and schematic of “zig-zag” incision.

**Fig. 2.** Left: A one-month postoperative slit lamp photo demonstrating a 24-bite running 10-0 nylon suture technique with zig-zag incision. Right: High magnification of graft/host interface with running suture demonstrating smooth surface transition at one month following zig-zag incision.
Regarding FLEK—an alternative to conventional PKP—some studies have demonstrated an improvement in CDVA, spherical equivalent and topographic astigmatism. Additionally, the advanced precision provided by the femtosecond laser allows for adequate scarring at four months post-op, creating the opportunity for possible early suture removal and topical steroid tapering.8

The deep anterior lamellar keratoplasty surgical approach to anterior corneal disease allows for selective replacement of diseased corneal epithelium and stroma after separation from healthy host Descemet’s membrane and endothelium. DALK is beneficial for keratoconus in that it provides the safety of extraocular surgery, the elimination of endothelial rejection, the potential for shorter post-op steroid regimens and increased graft longevity.18 Of note, stromal rejection is still a risk and should be routinely monitored for.

The DALK “big bubble” technique, combined with the femtosecond laser, was first described in 2009.19,20 Femtosecond DALK offers the advantages of FLEK, including better donor/host fit, increased surface area apposition and faster wound healing, as well as the inherent benefits of DALK in treating stromal and anterior corneal disease (Figure 4).21–23

Additionally, fs-DALK is easier to perform than manual DALK because surgeons have the ability to customize the posterior ablation depth, allowing for more accurate dissection just anterior to Descemet’s membrane and reducing risk for perforation. Since a presentation of corneal hydrops can involve the Descemet’s membrane, keratoconus patients with a history of hydrops are not good candidates for DALK; instead, full-thickness grafts are recommended in these cases. Additionally, extreme corneal thinning increases the risk for Descemet’s membrane perforation, requiring intraoperative conversion of fs-DALK to a full-thickness keratoplasty. Fortunately, doing so maintains the advantages of femtosecond laser incision.19

There is a lack of reported data on the best surgical option at any given stage of keratoconus; as such, ultimately, the clinician must decide based on exam findings, patient symptoms and disease state (Figure 5). As always, initial management should first involve spectacles and/or contact lens wear. Corneal crosslinking may play a role in stabilizing disease progression, but further research is needed to determine efficacy and safety. ICRS can decrease refractive error and irregular astigmatism. Corneal transplantation is reserved for more advanced disease; however, patient outcomes have been vastly improved by recent improvements in technology and technique. The advent of the femtosecond laser has given surgeons a new tool for achieving better and faster visual recovery. Future options may also include novel artificial or engineered corneas for keratoplasty.


Fig. 3. OCT scan of a zig-zag incision taken one month post-op shows good alignment of donor and host tissue at both the anterior and posterior surfaces.

Fig. 4. The fs-DALK graft technique.
(a) Baring of Descemet’s (highlighted in red) using “big bubble” technique after standard non-penetrating femtosecond laser zig-zag incision.
(b) “Big bubble” technique with bubble dissecting tissue anterior to Descemet’s membrane.
(c) Removal of entire stroma, preserving host DM and endothelium.
(d) Suturing of donor tissue after donor endothelium has been removed.

Fig. 5. Surgical management of keratoconus decision tree.
One of the most sensitive yet underused instruments in practice today for identifying and characterizing corneal disorders is the corneal topographer. This device provides us with an array of analysis options that can accurately reveal conditions that warrant our attention or treatment. However, as with most advanced instrumentation available to practitioners today, understanding and keeping track of the wide range of options for interpreting results can be overwhelming. For example, which analysis display is best at uncovering the earliest signs of a given disease? How should we differentiate one condition from another? Which functions of the technology help to monitor patients if long-term changes are occurring?

The corneal topographer can be a powerful tool in the hands of both the experienced and neophyte user when properly armed with an understanding of the fundamentals.

This article will discuss the various capabilities and functions of the technology in disease screening, particularly as it relates to keratoconus and corneal thinning disorders.

**TYPES OF UNITS**

Though dozens of corneal mapping systems exist, all can be categorized as either reflection or projection units.\(^1\) Reflection systems map the anterior surface of the cornea and provide noninvasive tear film breakup and stability testing as added features.\(^1,2\) Newer reflective-based topographers are capable of measuring beyond the cornea to provide information on scleral shape and sagittal depth, which can be beneficial when fitting large diameter contact lenses.\(^3,4\) However, regardless of additional features, all reflective-based systems are only capable of measuring the anterior surface and therefore cannot describe the posterior cornea or its thickness.

By comparison, projection-based instruments measure both the anterior and posterior corneal surface.\(^1\) This also allows for pachymetry measurement of the cornea’s thickness. Projection topography provides a more comprehensive picture of the cornea as a whole and is generally favored by surgical sites when considering LASIK candidacy.

When attempting to determine if a particular cornea is normal or abnormal, one of the simplest analyses possible is to assess the range of power distributed across the corneal surface.\(^1\) Using an “axial” interpretation, determine the flattest dioptric power that the topographer can read on the selected cornea. Then, find the steepest reading on the surface. The dioptric difference between the two is the scale range of power (Figure 1). Normal corneas distribute less than 10D of power from the flattest to steepest readings on the axial display. This analysis option may be described by some units as the power or sagittal display.

Another analysis option of diagnostic value is the tangential display, which is known in some instruments as the instantaneous or true curvature map.\(^1\) This interpretation is more sensitive to finite

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**Fig. 1.** Analyze the scale range of power distributed across the corneal surface on the axial map. Distributions of less than 10D of power are typical of normal eyes.
Mapping the irregular cornea reveals much about disease status and its amenability to treatment. Here’s a review.

surface changes in the eye, which the axial display can miss due to the smoothing effects inherent in its formula. A tangential map clearly defines the size, shape and position of the anomaly or diseased tissue. This may be helpful when characterizing the presentation of one condition over another.

To help us simply and efficiently classify the various types, one study identified the following criteria: nipple cones measure less than 3mm in diameter and generally are located more central to the pupil, while oval keratoconic presentations had diameters of 3.0mm to 5.5mm and are usually present inferior to the visual axis (Figure 2a and 2b).6 Cones greater than 5.5mm are considered globic and involve much more of the corneal surface, but present in a smaller percentage of cases (Figure 2c).6 Lastly, pellucid and Terrien’s marginal degenerations are other forms of corneal thinning disorders that are rare and involve tissue closer to the limbus than keratoconus typically does (Figure 2d).6

**DISEASE INDEXING**

Though corneal topography axial and tangential maps can be used to identify when the condition is moderate or severe, neither interpretation can provide a definitive diagnosis when the disease is mild or in the early-onset stage. For this reason, a series of disease detection indices have been developed; these eliminate the need for operator experience in comprehending the subjective appearance of topography contours and instead provide results using a mathematical analysis of the corneal shape.

The first and possibly most relied-upon technique for assessing possible signs of disease is the inferior-superior (I-S) value (Figure 3).7 This is a comparison of the average power of the inferior cornea against the average of the superior cornea: if one hemisphere is significantly different from the other, this is a strong indicator of an abnormal and possibly diseased eye.
WHAT CORNEAL SHAPE REVEALS ABOUT CORNEAL HEALTH

Considering approximately 90% of the corneal thinning disorders present in the inferior hemisphere, it’s logical for the inferior cornea to be significantly steeper than the superior. As such, I-S values greater than 1.5D are indicative of an abnormal eye.

Another commonly employed disease detection index is the surface asymmetry index (SAI), which compares the principal oblique meridians for symmetry (Figure 4). For example, the curvature from the apex along axis 45° is compared to the opposing curvature from the center along axis 225°. Then, the same hemi-chord comparison is completed between axis 135° and 315°. A completely symmetric eye would have an SAI value of zero, but if the oblique meridians are significantly different from each other, this indicates an asymmetric surface instead. SAI values greater than 1.0D are indicative of a diseased or irregular eye.

The last disease detection index typically employed is the surface regularity index (SRI), a center-weighted analysis that determines the relative smoothness of the cornea. High power distributions within the pupil may be indicative of an irregular cornea: though a normal, healthy eye would have a smooth central cornea to allow for high quality visual acuity, a diseased eye with radical surface power changes within the pupil is more likely to degrade vision. SRI values greater than 0.80 are considered abnormal; suspect corneal disease in such eyes. Simply put, the higher the value, the more abnormal the surface. The scale range, I-S, SAI and SRI would indicate a symmetrical or normal eye is closer to zero values while a more irregular or diseased eye would be a higher value.

The position and size of the steepest curvature, or apex, can also provide insight when diagnosing disease. For example, a normal, healthy eye typically exhibits an axial or tangential topography, with the steepest curvature near the center and a gradual rate of flattening towards the periphery (Figure 5a). By comparison, a keratoconic patient typically exhibits an apex inferior of center, with a higher rate of curvature change within a smaller surface area of the topography (Figure 5b). In central or nipple keratoconus, the apex might present central to the pupil, but regardless of the position of the apex, it’s common in diseased and irregular eyes to see radical power changes within tighter distances as compared with normal, healthy eyes.

Another interpretation that topography software allows is the elevation display. This map overlays a best-fit spherical surface on the anterior surface and defines where the tissue is above the surface (red) and below (in blue) in microns. This can be helpful in determining how high the elevation changes in the eye surface are. Projection topographers have the added benefit of also being able to image both the posterior and anterior surfaces (Figure 6).
Research has suggested that the posterior elevation analysis may be more accurate in identifying the earliest signs of thinning disorders, where at times the anterior surface may not reveal irregularity.9,10

Often, characterizing the difference between keratoconus, pellucid (PMD) and Terrien’s marginal degeneration (TMD) can be difficult. Corneal topography can help to make distinguishing between the conditions relatively easier. For example, keratoconus appears on the axial or tangential map (in red) as a round or oval acute area of steep curvature, generally within the central or paracentral cornea.11 By comparison, PMD or TMD present with red curvature more peripheral and closer to the limbus.12 Additionally, PMD takes on a “kissing doves” or “butterfly wing” appearance on the axial map, while TMD presents in various other patterns. Both PMD and TMD, however, are more typically associated with lower flat K (Kf) readings than keratoconus is. Using PMD as an example, the Kf value is usually less than or equal to 40D, whereas in keratoconus the Kf value is usually greater than or equal to 45D.

When monitoring corneal disease cases, the subtractive or difference maps can be helpful in defining the smallest changes over time. Typically, this analysis function is used mostly in orthokeratology cases to assess the corneal changes produced following overnight wear. However, the subtractive map—when employed in a diseased eye—can determine if the condition is stable or advancing. Comparing two scans, a subtractive map demonstrating all green indicates no changes over time, but red and blue areas on the same map indicate steepening and flattening, respectively (Figures 7a and 7b). If there is steepening over the cone, this likely indicates disease progression, though it can also be the result of contact lens molding. Conversely, we may often see more blue over the cone on the subtractive map following corneal crosslinking.

CONTACT LENS FITTING

Ultimately, if we are diagnosing corneal thinning disorders in our patients, there is a high likelihood we will be fitting specialty contact lenses to improve the patient’s visual quality of life. Though many time-tested custom soft lens options exist for keratoconus today, the axial map can help us better understand the power of the anterior surface and whether a soft lens is the right choice for a particular patient. For example, if the distributed power within the pupil shows a range greater than 10D, this could result in too much aberration for a custom soft lens to mask. Additionally, the presence or absence of symmetry within the pupil can be telling: the more extreme the power distribution, the less likely a conventional or custom soft lens will be able to mask the asymmetry.

Another question we face is whether to fit a particular eye with a corneal GP or scleral lens. One study attempted to answer this question by identifying the highest point of elevation in microns (red area) and the lowest point of depression (blue area) on each eye in a topographical elevation map. The difference in height between the two was considered to be the elevation differential (Figure 8). The study determined that patients with less than or equal to 350µm of elevation change could achieve a successful corneal GP lens fit 88% of the time, while those with greater than 350µm of elevation change required a scleral lens to fit over the high asymmetry of the eye.13 In addition to steering us toward the right modality, corneal topographers also come with contact lens fitting modules. These programs allow for simulated fluorescein modeling of specific lens designs to accurately determine the initial trial or custom parameters that are best suited to the eye (Figure 9). Research demonstrates that corneal topography fitting software can predict the fluorescein pattern 74% of the time, regardless of the map.
WHAT CORNEAL SHAPE REVEALS ABOUT CORNEAL HEALTH

quality. However, with good quality images—which can be dependent on the eye’s surface and fissure size—this fitting accuracy can be improved to 95%. These modules can help reduce chair time and lab costs by improving the efficiency of the trial process and accuracy of the initial custom parameters.

TOMOGRAPHY VS. TOPOGRAPHY

The term *topography* refers to a detailed representation or description of the surface characteristics of a structure. Corneal topography is typically performed with Placido ring technology that provides curvature data of the anterior corneal surface. In contrast, *tomography* is the process of generating a two-dimensional cross-sectional image of a slice through a three-dimensional object. One method to achieve this is through the use of Scheimpflug imaging (Figure 10). This technology is used by instruments like Pentacam (Oculus) and Galilei (Zeimer) to measure the anterior and posterior corneal surface, as well as other anterior segment structures. A rotating Scheimpflug camera (single in Pentacam; dual cameras in Galilei) creates a 360° representation of the anterior segment.

Scheimpflug tomography of the anterior segment provides true elevation-based data that allows for measurements of anterior corneal elevation, posterior corneal elevation, full corneal thickness (global pachymetry) and high quality imaging and structural measurements of the anterior segment. Elevation data can also be used to derive corneal curvature measurements analogous to the curvature data obtained by Placido-based corneal topography. True elevation corneal measurements and data of both the anterior and posterior cornea, along with global pachymetry data, allows for highly accurate detection of corneal ectatic diseases like keratoconus. Specifically, posterior corneal elevation anomalies will precede and be more advanced than anterior corneal elevation and curvature anomalies in keratoconus.

There are many cases in which the anterior corneal surface is found...
to be normal—both in terms of curvature and elevation—but a posterior corneal elevation anomaly still exists. This can result in what would be a false negative for keratoconus detection, if only the anterior corneal surface were analyzed (such as with Placido-based corneal topography). Conversely, some conditions or situations can result in what would appear to suggest keratoconus, when, in fact, the cornea is quite normal. Displaced corneal apices and large deviations of the visual axis (large angle Kappa) are two instances in which Placido topography, or any representations of anterior corneal surface curvature, would suggest keratoconus while elevation-based Scheimpflug would suggest an otherwise normal cornea.\(^{16}\)

Additionally, anomalies of global pachymetry are also highly diagnostic for keratoconus. Though many cases of keratoconus are characterized by the thin point of the cornea being below that normal range of minimum corneal thickness, it is the rate of change of corneal thickness from the thin point out peripherally that is the most sensitively diagnostic, even in cases where the thickness of all points in the cornea are statistically normal.\(^ {17}\)

Scheimpflug tomography can also aid in the differential diagnosis of true pellucid marginal corneal degeneration. Traditionally, the classic “kissing dove” or “crab claw” pattern seen on curvature topography maps is considered characteristic of PMD; however, because true PMD exhibits localized peripheral corneal thinning inferiorly 1 mm from the limbus, in the majority of cases the corneal thin point on global pachymetry and corneal elevation maps is actually significantly more central in regular PMD than in true PMD. In fact, the global pachymetry maps and both the anterior and posterior corneal elevation maps are quite typical of true keratoconus rather than pellucid.\(^ {18}\)

One of the newest developments in Scheimpflug tomography is the technology’s ability to image and measure corneo-scleral profiles. These measurements can be used to detect and quantify asymmetries of scleral shape when performed in 360°. Software is under development to enable this data to be used in the virtual design of both corneal and scleral contact lenses (Figure 11).

**Though we cannot rely on just one instrument, corneal topography itself can be sensitive to the earliest presentation of corneal thinning disorders, and can provide us with the ability to categorize the type of condition its severity. Understanding its functions can greatly aid practitioners both in the characterization of the condition and the selection of a contact lens modality to best suit the patient.**

4. Precision Ocular Metrolgy, USA. sMap 3D.

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**Fig. 10.** Scheimpflug image of a case of advanced keratoconus along an oblique meridian.

**Fig. 11.** Pentacam corneo-scleral profile software.
## EXAMINATION ANSWER SHEET

### What Corneal Shape Reveals About Corneal Health

Valid for credit through May 1, 2019

**Online:** This exam can also be taken online at [www.reviewofcontactlenses.com](http://www.reviewofcontactlenses.com). Upon passing the exam, you can view your results immediately. You can also view your test history at any time from the website.

**Directions:** Select one answer for each question in the exam and completely darken the appropriate circle. A minimum score of 70% is required to earn credit.

**Mail to:** Jobson Optometric CE, Canal Street Station, PO Box 488 New York, NY 10013

**Payment:** Remit $20 with this exam. Make check payable to Jobson Medical Information LLC.

**Credit:** COPE approval for 1 hour of CE credit is pending for this course.

**Sponsorship:** Joint-sponsored by the Pennsylvania College of Optometry

**Processing:** There is an eight-to-10 week processing time for this exam.

### Answers to CE exam:

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**Evaluation questions (1 = Excellent, 2 = Very Good, 3 = Good, 4 = Fair, 5 = Poor)**

- Rate the effectiveness of how well the activity:
  - 11. Met the goal statement:  
  - 12. Related to your practice needs:  
  - 13. Will help improve patient care:  
  - 14. Avoided commercial bias/influence:  
  - 15. How do you rate the overall quality of the material?  
  - 16. Your knowledge of the subject increased:  
  - 17. The difficulty of the course was:  
  - 18. How long did it take to complete this course?  
  - 19. Comments on this course:  
  - 20. Suggested topics for future CE articles:  

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By submitting this answer sheet, I certify that I have read the lesson in its entirety and completed the self-assessment exam personally based on the material presented. I have not obtained the answers to this exam by fraudulent or improper means.

**Signature:**

**Date:**

**Please retain a copy for your records.** LESSON 12914, RO-RCL-0516

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### CE TEST - MAY 2016

**WHAT CORNEAL SHAPE REVEALS ABOUT CORNEAL HEALTH**

1. **Reflection-based topography systems:**
   - a. Map the anterior surface of the cornea and, in some cases, provide non-invasive tear film break-up and stability testing.
   - b. Measure both the anterior and posterior corneal surface to allow for pachymetry measurement of the cornea’s thickness.
   - c. Can definitively determine whether the disease is mild or in the early-onset stage.
   - d. Are favored by surgical sites when considering patients for LASIK.

2. **The inferior/superior value:**
   - a. Compares the principal oblique meridians for symmetry.
   - b. Determines the relative smoothness of the cornea.
   - c. Is a comparison of the average power of the inferior cornea against the average of the superior cornea.
   - d. Denotes a more abnormal corneal surface if higher in value.

3. **Keratoconus patients typically exhibit which of the following?**
   - a. Peripapillary curvature closer to the limbus and a kissing ditches appearance on the axial map.
   - b. Flattening over the peripheral cornea area of thinning and steepening opposite to the mid-point of thinning, resulting in against-the-rule or oblique astigmatism.
   - c. Axial or tangential topography with the steepest curvature near the center and a gradual rate of flattening toward the periphery.
   - d. An area inferior of the center with a higher rate of curvature change within a smaller surface area of the topography.

4. **When monitoring corneal disease, which topography map demonstrates if the condition is stable or advancing?**
   - a. Difference map.
   - b. SAD/ADD map.
   - c. Axial map.
   - d. Tangential map.

5. **The elevation display defines where the tissue is above and below the surface in what colors, respectively?**
   - a. Red and green.
   - b. Red and blue.
   - c. Red and purple.
   - d. Green and red.

6. **Which of the following conditions is Scheimpflug tomography best suited to identify?**
   - b. Terrien’s marginal degeneration.
   - c. Pellucid marginal degeneration.
   - d. Keratoebulbus.

7. **What is the measurement criteria for distinguishing between the cones of nipple, oval and globic keratoconus?**
   - a. Less than 4mm/4mm to 5mm/greater than 5mm.
   - b. Less than 2mm/2mm to 6mm/greater than 6mm.
   - c. Less than 2mm/2mm to 5mm/greater than 5mm.
   - d. Less than 4mm/4mm to 5mm/greater than 5mm.

8. **SRI values greater than what are considered abnormal?**
   - a. 0.60.
   - b. 0.70.
   - c. 0.80.
   - d. 0.90.

9. **One of the newest developments in Scheimpflug tomography is its ability to:**
   - a. Create simulated fluorescent patterns.
   - c. Use displaced corneal apices and large deviations of the visual axis to identify keratoconus.
   - d. Track corneal disease progression.

10. **Approximately what percentage of corneal thinning disorders present in the inferior hemisphere?**
    - a. 10%.
    - b. 50%.
    - c. 70%.
    - d. 90%.
Boost Your Contact Lens Fits With Help from Your Staff

A group effort can mean a stronger, more informed choice.

The typical contact lens business may be missing a substantial opportunity to grow if its staff members are not adequately informed of the value that contact lens wearers bring to an eye care practice. Totaled up over their lifetime, this population represents a considerable source of potential revenue. As such, it is important for practitioners to consider how they currently incorporate contact lenses into their repertoire and what steps should be taken to continue moving forward—whether it's securing new wearers or upgrading existing ones. This strategic plan can't be implemented by the doctor alone, however. Instead, it all starts and ends with the practice’s staff.

DELEGATION
Ensuring each patient receives enough attention from the doctor during the overall encounter is key to a successful visit. As in any business, adequate preparation and reliance on support from your staff can help make this happen. Though some practitioners may find it challenging to allow others to help with the contact lens exam, it can free the doctor up to investigate another concern, interpret data from scans or even move on to the next patient. Encouraging your contact lens technicians to gain further education with contact lenses and achieve certification allows them to record patient history, assess lens fits and perform overrefractions, potentially even smoothing the patient exam process enough to allow for an extra exam or two.

THE HISTORY
Start by making an effort to understand how the patient uses their eyes throughout the day. Proper questioning should include asking how many hours per day they spend on a digital device, as technology is now an integral part of our daily life. Next determine the number of hours each patient wears their contact lenses per day, including how many of those are “comfortable” hours. Inquire specifically about their “end of day” comfort.

Other small steps that may improve efficiency with contact lens wearers include:
1. Institute in-office messaging. Whether to help with diagnostic lens fitting or first-time contact lens training courses, the ability to communicate with staff members makes the adoption of any changes smoother and more effective. For example, the use of signaling systems, texting or computer programs can help improve the flow of information. Additionally, having a point person outside the exam room ready to relay any necessary directions can ensure clarity.

2. Keep diagnostic lens banks up to date. Not having the necessary diagnostic lens in your stock means that certain patients cannot be fit—and if they’re already waiting in the exam chair, this constitutes a serious problem. Thus, when training staff members on contact lens exams, also check which diagnostic lenses you have on hand and order the missing ones.

3. Spread the knowledge. Make sure that multiple staff members have the capacity to help with contact lens insertion and removal. This ensures someone is always there to jump in and help with this sometimes time-consuming process.

TIPS FOR TRAINING
First and foremost, all staff members should know how to talk to patients about their contact lenses. This starts in the front office with the answering of a patient’s phone call. Have them mention contact lenses at some point during this call: whether it’s to remind the patient to bring in their old prescription or to consider the possibility of trying lens wear, introduce them to the concept of exploring contact lenses during their next visit.

This conversation then continues with the technician, so get them on board to discuss new opportunities specific to the patient’s prescription or introduce other lens possibilities to them. For example, if the patient is a current wearer who uses monthly disposables and complains of discomfort towards the end of their wear cycle, daily disposables or lenses made from more oxygen permeable materials are alternatives the technician could suggest.

On the other hand, if the patient has never worn contact lenses before, the technician can ask them the during the pre-test if they’ve ever considered trying them, and to talk to the practitioner if they’re
Boost Your Contact Lens Fits with Help from Your Staff

(Continued from page 37.)

interested. Ask the technician to make a note in the patient’s chart if this is the case, so that the desire can be followed up on. The technician may also be the ideal person to provide the patient with contact lens fitting fees and annual wear cost estimations.

For the practitioner, the key to a successful visit is for them to listen to the patient’s concerns, their needs and desires for a contact lens. Always offer them the best lens choice to meet those requirements, but also consider individualizing prescribing habits to meet a patient’s lifestyle, even if they don’t explicitly ask for it. Let them know you wish to ensure their lenses are the clearest and most comfortable option for their circumstances and, above all, state the expectations for success up front. They may thank you.

CONSIDERING THE ENVIRONMENT

In addition to strengthening your staff’s education, offer them the chance to work with the most innovative products. Though some diagnostic equipment may be reserved for those patients who are complaining of symptoms, many who present simply looking to refill their old prescription can also benefit. Patients are often busy in their own world and are not up to date on events or developments in contact lens technology. This allows for an opportunity to not only demonstrate the newest advancements to the patient, but also for staff members to use these pieces of equipment.

If the patient does not opt to move forward with contact lens wear, have your opticians make one final offer. Consider quickly re-evaluating your protocol and make that offer while they are choosing frames. A study analyzing the fitting process with non-contact lens wearers found that fitting all suitable patients with contact lenses prior to spectacle dispensing not only makes the frame selection process easier, but also is a good way to get patients interested in contact lenses.1

In conclusion, communication is vital. Constant communication between all members of the office staff—from the receptionist to the practitioner—will enable better cooperation towards the end goal of growing the contact lens business. Each staff member should extend the office philosophy in each patient encounter, delivering a consistent message. Take note: this does not mean that everyone in the office needs to be an expert in contact lenses; rather, they should simply be kept aware of the fitting process and the newest lens technologies available. This approach may help you own the contact lens market in your area.

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