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n a recent series of observational studies called for by the FDA, researchers used a questionnaire to better understand patient-reported issues with laser in situ keratomileusis (LASIK) surgery, including the frequency of visual and dry eye symptoms, satisfaction with vision and overall satisfaction with the surgery.1

While the safety and efficacy of LASIK have been well documented, reports of dry eye symptoms, problems with vision postoperatively and generally dissatisfaction with results still exist, prompting the Patient-Reported Outcomes with LASIK (PROWL) questionnaire study. Researchers surveyed two groups of patients throughout their LASIK experiences: PROWL-1, which consisted of active-duty Navy personnel ages 21 to 52 from a single military center, and PROWL-2, which consisted of civilians ages 21 to 57 from five private practices and academic centers.

Although results were generally favorable, 28% of patients who had normal Ocular Surface Disease Index (OSDI) scores at baseline ended up with mild, moderate or severe dry eye symptoms at three months post-surgery. Although the total amount of visual symptoms and dry eye symptoms decreased from pre-surgery to post, 43% of PROWL-1 and 46% of PROWL-2 reported new visual symptoms three months after surgery. Additionally, 1% to 4% of patients were dissatisfied with their vision, and 1% to 2% were dissatisfied with the surgery overall.

“What’s surprising is the fact that preoperative OSDI results don’t correlate well with postoperative OSDI score,” says Clark Chang, OD, director of Clinical Services at TLC Vision and director of Cornea Specialty Lenses at Wills Eye Hospital Cornea Service in Philadelphia. According to Dr. Chang, this could stem from discord between clinical signs and patient symptoms in dry eye, the lack of a definitive clinical test to diagnose dry eye and postoperative dry eye being induced by something separate from the current clinical understandings.

Despite the number of patients who developed new visual symptoms postoperatively, only a few actually reported any kind of significant lifestyle impact. Still, conclusions from the study encourage doctors to counsel prospective LASIK patients about the possibility of postoperative visual symptoms.

The questionnaire is a step in the right direction towards a greater understanding of the risks associated with LASIK. “Patients and health care providers now have a well-defined tool to guide medical decisions and future research,” says Malvina Eydelman, MD, director of the Division of Ophthalmic and Ear, Nose and Throat Devices at the FDA’s Center for Devices and Radiological Health. “By continuing to listen to the patient’s perspective during the development, evaluation and use of medical devices, the FDA and manufacturers can work together to better assure that LASIK devices marketed in the United States address patients’ needs.”

Legal Changes in the Air

It’s been a combative time as various groups push back against challenges to contact lens distribution, citing potential safety risks. Here’s a review of the recent trends, news and nail-biters:

**UPP BATTLE**

In the wake of 1-800 Contacts’ victory in Utah, Johnson & Johnson Vision Care, Alcon and Bausch + Lomb have since reevaluated their policies. Johnson & Johnson discontinued the practice of UPP for its CL products in April 2016, and Alcon followed suit in December 2016 days after a federal appeals court allowed Utah’s anti-price fixing law to remain in effect.1 Bausch + Lomb was the last to fold, finally ending it’s UPP practice just last month.2

**FCLCA CHANGES**

The recent Contact Lens Consumer Health Protection Act of 2016 (S.2777) would amend the Fairness to Contact Lens Consumers Act (FCLCA) of 2004 by adding patient safety requirements and increased accountability for the online contact lens sales industry. The companion bill, H.R. 6157, would modernize prescription verification and clarify consumer protections regarding false advertising. It would disrupt the “robocalls” and other illegal sales tactics, says Joseph P. Shovlin, OD, of Scranton, Pa. “It requires that lenses be prescribed exactly as ordered from the prescriber and increases infraction fines to $40,000.”

**CONTACT LENS RULE OVERHAUL**

The comment period for the FTC’s proposed changes to the Contact Lens Rule—which would require eye care practitioners to obtain a signed acknowledgement from patients after receiving a prescription—ended January 30, with more than 4,000 commenters.1,4

While Johnson & Johnson Vision Care, the AOA and the Coalition for Patient Vision Care Safety drafted formal comments opposing the proposed changes, one comment signed by the attorneys general of 20 states suggests the proposed change is, in part, intended to “empower consumers to comparison shop for contact lenses, and to spur more competition.”4,5

**COMPETITION LAWSUITS**

In August 2016, the FTC sued 1-800 Contacts, alleging that it unlawfully entered into bidding agreements with at least 14 competing online contact lens retailers that eliminate competition in auctions to place advertisements on the search results page generated by online search engines.6 The agreements could restrict truthful and non-misleading internet advertising to consumers, resulting in some consumers paying higher retail prices for contact lenses.6

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4. 20 State AGs among 4,000 ‘commenters’ to FTC over proposed changes to contact lens rule. Vision Monday. February 14, 2017.
Amniotic Membrane: A Game-changer?

This treatment modality for recurrent corneal erosion might be the alternative you have been looking for.

Perhaps no corneal condition is more vexing than recurrent corneal erosion (RCE). While the causes vary, the treatment options have remained virtually unchanged for quite some time, at least until recently. Today, amniotic membranes have gained popularity as an alternate treatment for RCE.

THE PROBLEM

In RCE, disturbance to the underlying layers of the epithelium result in faulty hemidesmosome attachment of the epithelium to the basement membrane, and the loosely attached overlying epithelium easily erodes. Clinicians can gently pass a dry cellulose sponge over the suspected loose area of epithelium to look for epithelial movement.1

RCE can be the result of damage following trauma, known corneal dystrophy of the anterior cornea, corneal degeneration or surgical procedures such as refractive surgery, corneal transplants or even cataract surgery. Diabetes is also a well-known confounding factor.

THE TRIED-AND-TRUE

Traditional therapy for acute episodes includes antibiotic ointment to prevent infection, cycloplegia and proper lubrication. Once the epithelium closes, a hyperosmotic agent may aid in proper adhesion. For large abrasions, clinicians can use bandage lens therapy and pressure patching, but should avoid patching for contact lens wearers. Oral analgesics are often helpful as well.

When recurrent episodes are frequent, epithelial debridement, anterior stromal puncture, diamond burr polishing, excimer laser phototherapeutic keratectomy or extended bandage lens wear may be required. The key to minimizing recurrence is applying debridement and polishing beyond where the pathology appears to exist.

Due to its matrix metalloproteinase-9 inhibition, oral doxycycline 50mg BID with (and sometimes without) topical corticosteroid use for two to four weeks may be beneficial for some patients.

Often patients who have failed on other treatments do well with autologous serum tears as an adjunctive therapy. These affect the final phases of wound healing by supplying growth factors and extracellular matrix components that activate keratocytes.1 Always remember, medical treatment is aimed at minimizing inflammatory activity and managing meibomian gland disease.

THE NEW PLAYER

Amniotic membranes can create a so-called biological bandage and work as a temporary dressing, providing much-needed anti-inflammatory and anti-scarring effects. These membranes contain both antimicrobial properties and neurotrophic factors with low immunogenicity. They also inhibit angiogenesis.2,3

But new treatments always come with a host of unknowns: does the use of amniotic membrane after epithelial debridement and polishing significantly enhance healing to minimize recurrence? If so, how does this affect the overall healing time? Should we use this modality on most of our patients? At what point does it make the most sense? There’s not much information available to answer these questions, although a small series of retrospective reports show less recurrence with amniotic membrane use following diamond burr polishing.

Eye care providers still debate whether to use cryopreserved tissue or a dehydration preparation. While research shows fresh amniotic membrane is effective in clinical applications, it presents a significant risk of disease transmission, highlighting the importance of the processing methods that preserve biological effectiveness while ensuring safety.2

To complicate this new treatment further, some wonder if amniotic drops will eventually supplant the use of membrane tissue. So far, research shows morselized amniotic tissue is helpful in treating non-healing corneal defects.2

We all await a prospective, controlled study comparing bandage contact lenses with amniotic membrane application following debridement and polishing in patients who suffer from RCE. In the meantime, we must continue to use good judgment in caring for patients who are desperate for relief from the pain and inconvenience associated with erosive syndromes—which just might include applying an amniotic membrane to the cornea.

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April 20-23, 2017"
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Fitting Challenges
By Vivian P. Shibayama, OD

Foggy with a Chance of Clarity
Here are a few tricks you can use when battling scleral lens fogging.

For 20% to 30% of scleral contact lens wearers, lens fogging means removing their lenses and refreshing the fluid multiple times a day.1-4 Many scleral lens wearers suffer from ocular surface disease, and tear film debris is found to be more common in these patients.3,5

CASE
A 54-year-old female scleral lens wearer with a history of dry eye and s/p LASIK OU presented with complaints of constant lens fogging. Her medical history was positive for allergic rhinitis. She had no significant family history. Her medications included Flonase (GlaxoSmithKline) and Zyrtec (AstraZeneca), omega-3 fish oil supplements and Restasis (Allergan).

She reported that her scleral lenses fogged “constantly” and she had to remove and rinse them several times a day. For this reason, she limited her wear to three to four hours at night when her vision was at its worst. She would clean her lenses with Boston Advance cleaner (Bausch + Lomb) at night, store the lens in Boston conditioner and fill the lenses with Unisol 4 (Alcon) before insertion. Her presenting visual acuity in spectacles was OD -2.50 +0.50x067 VA 20/25- and OS -1.50 +1.00x118 VA 20/25- +2.00 add J1 OU.

Her pupils were round and reactive to light, with no relative afferent pupillary defect in either eye, and her extraocular movements were full OU. A slit lamp examination revealed clear lashes and 1+ inspissated meibomian glands bilaterally. Punctal plugs were in place in the inferior puncta OU. Examination of the corneal surface revealed 1+ inferior punctate staining, a tear break-up time of two seconds and a low tear lake OU.

The conjunctiva was white and quiet, irides and lenses were clear, and the anterior chamber was deep and quiet OU. Her intraocular pressure was 12mm Hg OU. Undilated posterior segment evaluation revealed a normal fundus OU with trace nuclear sclerosis OU.

Manifest refraction OD was -2.75 +0.50x060 VA 20/25 and OS -1.00 +0.50x118 VA 20/25 +2.25 add near acuity J1 OU. Topographical evaluation of the cornea revealed flat, plateau-shaped corneas OU and simulated keratometry read OD 36.76/34.23 at 175, OS 35.60/34.33 at 124 (Figure 1).

CONTACT LENS EVALUATION
The patient’s presenting contact lens parameters, measured in office, were OD 8.0BC/-11.00/15.7 CT 0.3mm diameter VA 20/20- and OS 8.0BC/-10.25/15.7 CT 0.3mm diameter VA: 20/25-.

Once she inserted her lenses, we instilled fluorescein to evaluate the fit. The fluorescein immediately entered the lens chamber at 10 o’clock in the right eye and two o’clock in the left eye, completely filling the lens within 10 seconds of instillation. Each time the patient blinked, fluorescein pumped into the lens chamber of both eyes along with mucous strands from the patient’s eye. The lenses were visibly flexing with each blink. This, combined with the fact that the patient had an apparent toric sclera, was causing tear film debris to be pumped into the lens.

DIAGNOSTIC CONTACT LENS FITTING
For the patient’s diagnostic contact lens fitting, we used a Europa (Visionary Optics) scleral fitting design. Since she was post refractive surgery, the first diagnostic lenses used were slightly flatter than average. A 42D/16.0 diameter lens was chosen for her right eye and a 43D/16.0 lens was chosen for her left eye, both of which were of standard spherical periphery.

Fluorescein was instilled and the lenses immediately filled with dye from the superior temporal edge of each lens (Figure 2). The right lens exhibited 250µm of central corneal clearance, while the left lens was roughly 350µm.
Both lenses had adequate limbal clearance, but excessive superior and inferior edge lift, causing the lens to move along the vertical meridian. The over-refraction of both lenses brought the vision to 20/20 OU.

To reduce lens flexure, we increased the center thickness of the lenses. A 2D toric periphery was added to both lenses to align them to the conjunctiva, create tighter seals and reduce lens movement. The initial lens measurements were OD 42.00/-11.25/16.0 2D toric periphery center thickness 0.35mm and OS 42.00/-9.25/16.0 2D toric periphery center thickness 0.35mm.

Finally, we addressed inferior band staining, which was likely due to nocturnal lagophthalmos. We advised the patient to add to her care regimen a gel tear at night and again in the morning, consider taping the lids to sleep and continue using Restasis OU. We also advised her to be diligent with daily lid hygiene and warm compresses.

FOLLOW-UP #1
The patient presented for a dispensing visit two weeks later. She reported that her eyes felt less dry in the morning as a result of the gel tears. When we inserted the lenses with fluorescein, they exhibited central 250µm of clearance OU on the initial insertion with good limbal clearance and centration. The right lens had slightly excessive superior edge lift and moved easily when the lid was pushed up. The left lens was aligned to the conjunctiva with minimal movement. The patient’s vision was measured at 20/20 OU with no over-refraction. We advised the patient to switch to a peroxide-based solution and use preservative-free sodium chloride to fill the lens.

FOLLOW-UP #2
At her next follow-up a week later, the patient had noticed a 50% improvement in fogging in the right eye and about 75% improvement in the left eye. She was able to wear the lenses for 10 to 12 hours a day while only rinsing them once or twice. Her right lens would become foggy faster than the left lens, however. A slit lamp examination revealed mild surface debris on the outside of both scleral lenses and some tear debris inside the chamber of the right lens. The central clearance was estimated at about 150µm to 200µm. After instilling, the right lens filled completely in 20 to 30 seconds, while the left lens did not fill within a one-minute period (Figure 3).

The left lens was finalized, but we recommended 3D of peripheral toricity in the right lens to reduce movement and improve alignment to the conjunctiva. We advised the patient to add Progent cleaner (Menicon) to her cleaning regimen every two weeks and remove surface debris with a moistened cotton swab or wipe the lens with a plunger edge midday instead of removing the lenses.

FOLLOW-UP #3
After another two weeks, the patient presented with no new complaints. She was removing her right lens two times a day to clear fog and cleared the surface debris...
on the left lens with a cotton swab. We placed the new right lens into the left eye to evaluate. A slit lamp examination revealed central 250µm clearance, and the lens did not move with blink. We then added fluorescein into the lens and there was no uptake within one minute of instillation (Figure 4). Her vision measured 20/20 OS. We sent her home with the new lens to try.

FOLLOW-UP #4
One week later, the patient was able to wear both lenses successfully with significantly less mid-day fogging. Occasionally she would have to remove the lenses to rinse the debris off, but most of the time she was able to wear the lenses all day without refreshing the fluid and she was happy with her vision and comfort. Her finalized contact lens prescription was OD 42.00/-11.25/16.0 3D toric periphery center thickness 0.35mm and OS 42.00/-9.25/16.0 2D toric periphery center thickness 0.35mm.

DISCUSSION
When troubleshooting midday fog, it is important to aggressively treat any lid disease or giant papillary conjunctivitis. Preservatives in solutions can also cause a buildup of matter in the lens, so preservative-free, peroxide-based solutions should be used. Introducing a regular deep cleaner like Progent can also be effective.

Central vaults should also not exceed 150 to 200 microns to minimize fogging. If a patient has a toric sclera, toric haptics can be used to create a greater seal.

Another consideration is the diameter of the lens. The farther sclera gets from the limbus, the more toric it becomes. Some believe the debris is related to the scleral lens placing pressure on the goblet cells of the conjunctiva, causing more mucin to be released into the eye. Smaller diameter lenses (16mm or less) will reduce the amount of misalignment of the conjunctiva, creating a better seal to prevent debris from entering the lens.

Lens flexure should be minimized to reduce movement of the lens against the conjunctiva by increasing the center thickness of the lens. In cases where fogging persists and the fit and care are optimized, patients can use a solution with higher viscosity to fill the lens and keep debris out of the lens chamber.

A NEW COATING ON THE HORIZON
Another option to consider is Hydra-PEG (Ocular Dynamics). The FDA recently approved this polyethylene glycol based polymer mix that bonds to the surface of contact lenses to improve their wettability and reduce the adherence of lipids and proteins. This coating would be ideal for patients with fogging issues and can improve discomfort related to ocular surface disease.

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pharmacological post-op care following cataract surgery has numerous objectives, including prophylaxis against infection (especially endophthalmitis, the most sight-threatening of possible complications), managing pain and inflammation and preventing cystoid macular edema (CME). Traditionally, patients are managed with three types of topical ophthalmic eye drops: an antibiotic, a steroid and a nonsteroidal anti-inflammatory (NSAID).

However, ophthalmic eye drops are not ideal for several reasons. First, they inherently have poor bioavailability, and drug level varies from one drop to the next. Second, eye drop schedule adherence can be problematic. In fact, most patients are also instructed to begin using eye drops a few days prior to cataract surgery, further extending the burden of drop instillation. Third, they have the potential for adverse responses, including toxicity or allergic reaction to either the active ingredient or preservative, as patients are often using the steroid and NSAID for several weeks. Finally, it can be expensive to use several medications at once, resulting in substitution for generic formulations.

THE COMPLIANCE ISSUE
Research suggests noncompliance with glaucoma medications can be as high as 40%. Adherence should be better for a shorter course of therapy with the objective of preventing complications after a surgery—but it’s not. In a recent study, eyedrop-naïve patients undergoing cataract surgery were recruited at the one-day postoperative visit. They completed a questionnaire about their postoperative medication use and were videotaped using their drops. The study found a major disconnect between subjective and objective reports. According to the subjects, 69% washed their hands before instilling drops, 42% were always accurate getting the drop in their eye and 58% never contaminating the dropper by touching their eye. But the videotape, reviewed by two independent researchers, showed that 93% demonstrated an improper instillation technique, 32% missed the eye, 57% contaminated the bottle and 78% did not wash their hands. Patients who had received instruction on drop usage did perform better.

With such poor compliance, it’s no wonder surgeons are looking for alternative methods of delivering postoperative medications including ophthalmic combination drops and injectable medications at the time of surgery.

COMBINATION MEDICATIONS
Using fewer drops means combining multiple therapeutic agents in one bottle. Pred-Gati (Imprimis Pharmaceuticals) consists of 1% prednisolone acetate and 0.5% gatifloxacin, Pred-Gati-Nepaf (Imprimis Pharmaceuticals) adds 0.1% nepafenac and Pred-Nepaf (Imprimis Pharmaceuticals) eliminates the antibiotic. All three are formulated as suspensions, available in 3mL bottles and cost $25.

These combination medications have several benefits: (1) the low cost makes it easier to prescribe different drops at different times in the postoperative regimen, (2) patients only have to use one bottle, (3) there is less exposure to preservatives, reducing the likelihood of toxicity, and (4) drug dilution is eliminated, as only one drop is instilled rather than several consecutive drops.

DROPLESS
Recent studies show moxifloxacin is as safe as a balanced salt solution when injected into the anterior chamber, with no toxicity. There was no statistically significant difference in acuity, intraocular pressure (IOP), corneal integrity (including endothelial cell counts) and anterior chamber reaction between either injection. Investigators also show intravitreally injected triamcinolone performs similarly to topical prednisolone in terms of reducing postoperative cells and flare, impact on IOP, patient symptoms and rate of complications. These findings set the stage for a new way of managing patients postoperatively—dropless.

Drugs injected transzonularly behave very differently, pharmacokinetically, than an intracameral injection, which is quickly washed out. A transzonular injection is retained in the vitreous matrix for an extended time, providing a sustained release of medications.

Some surgeons are compound-
ing their own injections. Imprimis has two injectable formulations available. Tri-Moxi contains triamcinolone acetonide and moxifloxacin. Tri-Moxi-Vanc adds vancomycin. They are injected through a cannula passed through the cataract incision site after the lens implant is in place but before the viscoelastic has been removed.6

In a chart review of 2,300 cataract-surgery eyes that received Tri-Moxi injections, there were no cases of endophthalmitis.6 A disadvantage of an intraoperative intravitreal injection is that the triamcinolone, which is formulated as a suspension, might cause initial visual blur and floaters.7 Despite this, several recent studies show that subjectively, patients are quite pleased with the dropless surgery.4,8 One recent study compared use of a Tri-Moxi-Vanc injection with a Pred-Moxi-Ketorolac (Imprimis Pharmaceuticals) single drop combination, and then Pred-Ketor (Imprimis Pharmaceuticals). Pred-Moxi-Ketorolac was used for one week and then discontinued, while Pred-Ketor was used for three more weeks. It was a small study of 25 patients with one eye receiving each treatment, so each patient’s contralateral eye served as the control. Outcomes were similar for the two groups in terms of IOP, central macular thickness and pain. Patients did prefer the injection for an improved overall cataract surgery experience, but there was similar visual acuity for the injected group vs. the topical group.4

A new study found that, of 1,541 eyes that received an intravitreal injection of Tri-Moxi-Vanc, none developed postoperative endophthalmitis, and 92% did not require supplemental medication following cataract surgery. Nine percent had breakthrough inflammation, requiring additional anti-inflammatory agents. The rate of CME was 2% and interestingly, there was no difference in the incidence in diabetic patients compared with non-diabetics. Less than 1% of patients manifested IOP greater than 10mm Hg at two-week or three-month follow up.8

While using fewer drops is a step in the right direction for improved postoperative care, dropless cataract surgery holds numerous advantages above and beyond this treatment advancement. Transzonular injections during surgery can provide predictable delivery of medication, excellent prophylaxis against infection and simplified postoperative care. Patients do not need to purchase drops or remember to use them, and physicians do not need to explain the medication schedule. Getting rid of drops could mean more satisfied patients.7

During transzonular drug delivery, the characteristic white pupil is evidence of quick diffusion throughout the eye and is the endpoint of uncomplicated cataract surgery.

The anatomical and optical irregularities caused by keratoconus present numerous challenges for clinicians looking to fit contact lenses. The increased higher-order aberrations (HOAs) in patients with keratoconus lead to impaired visual function, making the need for the best possible contact lens fit even more important. Although conventional ophthalmic lenses or standard soft contact lenses can’t effectively neutralize these induced aberrations, the same cannot be said for specialty lenses. Not only can specialty contact lenses correct for lower-order refractive error, they can also reduce HOAs by masking front corneal irregularities.

Eye care providers should have three goals in mind when fitting a patient with keratoconus. Each prescribed contact lens should provide: (1) adequate visual function for a patient’s daily activities, (2) acceptable comfort across the entire wearing period, and (3) proper fitting characteristics, such as sufficient dynamic tear exchange and oxygen transmission, to promote long-term ocular surface health. The patient doesn’t have to see 20/20 or have a lens that “fits perfectly” to have success, provided that these three goals are met.

Specialty lens fitters now have a range of lens designs to choose from, including specialty soft, corneal gas permeable (GP), piggyback, hybrid and scleral lenses. Although new options can increase a practitioner’s fitting success, it can be challenging to know when to use a particular lens design. Here, we discuss new specialty lens designs—and how clinicians can make the right choice for patients with keratoconus.

SMART STRATEGIES
One fitting strategy is to choose the initial lens type based upon the patient’s graded severity of keratoconus. Many clinicians start with specialty soft lenses if a patient’s presentation is considered mild based on clinical examination. If it’s more severe, fitting a scleral lens might prove more effective. Other variables to consider when choosing an initial design include topography findings, ocular comorbidities, previous lens experience, aperture size, manual dexterity and physical disabilities.

Adding to the fitting challenge is the presence of significant interocular asymmetry that often manifests with keratoconus. As a result, patients may ultimately have the best success with a different lens type in each eye, or may have to wear spectacles over their contact lenses to manage residual astigmatism, presbyopia or both.

SPECIALTY SOFT CONTACT LENSES
Patients who have mild keratoconus can sometimes successfully wear molded disposable soft contact lenses. Here, the primary benefit is patient convenience. These are prefabricated with multi-lens packaging to help reduce the anxiety of losing lenses and are available with various replacement schedules, including daily disposables. However, standard soft lenses are generally unable to provide the required visual improvements for patients with moderate keratoconus. In these cases, patients will need to be fit with a specialty soft keratoconus lens if they desire to stay with soft lenses.

Specialty soft contact lenses—custom designed and lathe cut for each individual eye—allow practitioners to adjust individual lens parameters to improve fit and vision. Soft keratoconus lens designs are manufactured by specialty lens laboratories with extended base curve and power ranges, which can accommodate many of the extreme fitting requirements of keratoconic eyes. This lens

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type’s center thickness is typically three to six times that of a traditional custom soft hydrogel lens (Figure 1). Enhanced center thickness helps to mask anterior corneal irregularities and decrease HOAs.

Pending disease severity, a soft lens may not mask all front surface irregularities, and many patients will continue to experience residual HOA. Future research into employing wavefront-guided correction technologies may offer a better clinical solution for managing residual HOAs when needed.1

Outside of the optical zone, a soft keratoconus lens is usually lenticularized to reduce lens profile thickness, improving the overall comfort of the lens and increasing oxygen transmissibility over the corneal limbus.

Given the high amount of astigmatic power often prescribed, most specialty soft keratoconus lenses will be ballasted with a double slab-off design to stabilize front surface toricity. They can now be manufactured in hydrogel or silicone hydrogel material. Silicone hydrogel, in particular, can increase oxygen permeability and avoid hypoxia associated with traditional hydrogel.2

The increased expense of lathe cut lenses largely determines lens replacement schedules as recommended by lens manufacturers, which are typically set at two- to three-month intervals.

Most designs are fit using diagnostic lenses by matching the sagittal height of the anterior eye with the sagittal depth of the soft keratoconus lens. High molecular weight sodium fluorescein can help clinicians assess flat or steep fits during the diagnostic fitting process. However, newer instruments capable of providing objective sagittal height data, such as a corneoscleral topographer, have dramatically improved fitting efficiency. Corneoscleral topography allows practitioners to measure mean sagittal height at a selected chord diameter, and a specific soft keratoconus design can then be matched to the sagittal height of the anterior ocular surface (Figure 2).

CORNEAL GP LENSES

The conventional standard for managing keratoconus has been corneal GP lenses. In fact, they are still the most commonly prescribed rehabilitation device for irregular cornea patients.3 In addition to significantly reducing HOAs, corneal GP lenses are usually least disruptive to corneal oxygenation due to their movement and smaller dimensions when compared with other lens types.1 However, the posterior GP surface bears weight on the cornea, causing longer adaptation time and greater lid-lens interaction.

Corneal GP lenses tend to center at or near the corneal apex and, therefore, can be an excellent choice for fitting keratoconus patients who have more centrally located apices such as central nipple cones and mild paracentral oval cones. Corneal GPs are often the only contact lens option for patients with deep sockets, small apertures or disabilities that prevent successful application of larger lens designs. Corneal GP lenses can be categorized into small (8mm to 9mm), medium (9mm to 10mm) and large (10mm to 11.5mm) diameters. Today, practitioners gravitate toward using corneal GP designs with large diameters, which tend to provide better centration, less unwanted lens movement and improved wearing comfort. Modern corneal GP designs can also offer benefits of aspheric optics, including back surface asphericity, which can improve the fitting relationship against an irregular corneal surface when compared with spherical posterior lens design fitting.

Alignment fitting is often only achieved in those with mild keratoconus, although some corneal touch may become unavoidable as the pathology progresses. Research shows flat-fitting lenses are associated with a higher rate of incidences of scarring.4 Although investigators have not shown a direct causal relationship between GP lens bearing and corneal scarring, it would be clinically prudent to minimize excessive apical bearing, since it can further lead to traumatic epithelial disruptions and associated lens discomfort (Figure 3).
PIGGYBACKS
To avoid apical bearing with corneal GP lenses in moderate to severe keratoconic cases, clinicians can piggyback a corneal GP lens on a soft contact lens. Due to the epithelial protection provided by the soft lens that is in immediate contact with the cornea, this modality also enables clinicians to troubleshoot short-term sequelae, such as contact lens discomfort from unnecessary mechanical trauma, thus ensuring long-term corneal health.

One of the most common patient objections to being fit for the piggyback lens system is the perceived inconvenience of maintaining two different lenses. Daily disposable lenses can frequently overcome this perception, provided the lens can center reasonably well. Daily replacement lenses can also provide better lens performance, as they are much less likely to develop chronic deposits or other issues with solution sensitivity and allergy-related wearing discomfort. Moreover, using silicone hydrogel materials will ensure enhanced oxygen transmissibility, which is an important consideration with tandem lens wear.

HYBRID LENSES
Many patients may not overcome their own perceived objections for piggyback lenses, and some may not adapt to piggyback lenses due to advanced staging of keratoconus. In these cases, newer hybrid lens designs may offer new hope. Hybrid lenses have a central GP disc surrounded by a soft material in the periphery. The GP center can offer good visual quality, while the soft skirt adds comfort by lifting the GP center from sensitive corneal tissue.

Many hybrid keratoconus lenses also have reverse geometry designs. This offers increased mid-peripheral sagittal height to better accommodate a decentered corneal apex and provide enhanced lens comfort. The added feature of reverse geometry also improves overall optics by approximating the optical zone closer to the corneal plane. In addition to reduced adaptation time from the incorporation of a soft lens periphery along with increased sagittal height in recent generations of hybrid lenses, this recombinant lens design reduces lens handling complexity and care compared with a piggyback lens system.

Not only does a built-in soft skirt assist with the centration of the hybrid lens on the eye, but it also helps to reduce complexity when fitting keratoconus patients with anatomical scleral obstacles (Figure 4). Research suggests the expanded range of fitting parameters in hybrid lenses can improve clinical outcomes in keratoconus patients, with a reported fitting success rate of 86.9%.6

Investigators reported hypoxia and difficulty with lens removal with early iterations of hybrid lenses; however, increased Dk value in GP centers and the incorporation of silicone hydrogel soft skirts (Dk of 84) in the newest hybrid lenses have largely alleviated clinical concerns of poor corneal oxygenation.2,4 Still, some patients have reported initial difficulty with proper lens removal techniques. Thus, it is wise to educate new patients regarding the early adaptation challenge prior to fitting and again at dispensing.

SCLERAL CONTACT LENSES
Keratoconus management is the most cited reason for using scleral contact lenses.5 Because the influence of corneal geometry is eliminated or significantly reduced, sclerals can achieve stable fitting patterns with improved comfort even in patients with advanced keratoconus. Most specialty lens manufacturers offer their own scleral lens designs that include fitting features such as bitoricity, notching, microvaults and multifocal optics. Bitoricity in posterior haptic surface can be used when scleral toricity is present, whereas notching or microvault can help to clear anatomical obstacles such as pinguecula or pterygium.

The majority of practitioners use diagnostic lenses for fitting with the clinical goal of providing a vault of approximately 200µm over the corneal apex.10 Although not an absolute fitting prerequisite, optical coherence tomography (OCT) can be invaluable in accurately measuring the corneal vault, especially when the cone apex is significantly decentered or when assessing a lens area with a shallow clearance. Patients often have better success with lens designs that incorporate back surface toricity when a front toric optic is desired, as it provides a stable lens platform to ballast the front toricity.11

Some scleral profiles are spherical, so not all patients require a back toric haptic surface. When a patient with a spherical sclera needs front surface toricity, a double slab-off prism can rotationally stabilize a
scleral lens. With that said, the ability to measure scleral topography to determine individualized back surface customizations in the haptic zone is an important step forward in improving the fitting efficiency and accuracy.

Commercially available instruments now map corneoscleral topography, and software will allow for simulated fitting of a scleral lens. Software algorithms can determine customized optic zone, limbal zone and haptic shapes to create a scleral lens that clears the cornea by a predetermined vault value set by the practitioner (Figure 5). Fitting software can also help dictate the geometry of a lens design. Although keratoconic corneas are prolate in geometry, some fits will result in more uniform vaulting with a reverse geometry scleral design. For example, keratoglobus, a special subcategory of keratocasisa, can challenge even the most experienced practitioner. Typically, these patients require large-diameter scleral lenses (between 20mm and 22mm) to create enough effective sagittal depth to vault over the extreme sagittal height manifested by this condition. Large optic zones and reverse geometry designs are needed to create extra mid-peripheral steepness to avoid peripheral corneal touch.

With recent manufacturing advancements in scleral lens technologies, multifocal optics have become reliable in correcting for presbyopia, even when fitting keratoconus patients. Most scleral multifocals are center-near designs that use simultaneous vision and are effective when two minimum conditions are met: (1) the central cornea is relatively clear of significant scar tissue, and (2) when the scleral lens is well centered.

Whether using a multifocal or single vision design, residual optical aberrations and lens decentration can be detrimental to the success of a scleral lens fitting. Clinicians can add wavefront correction to the front lens surface to correct for residual aberrations, including coma. To provide the best visual outcome, clinicians must ensure the anterior optical zone is centered so that the center of the front optical zone best matches the presumed location of a patient’s line of sight.

While scleral lenses can certainly be the first choice for patients who have severe keratoconus or keratoglobus, some experts debate whether they should be the first choice for patients presenting with mild to moderate irregularity. Since scleral lens fitting involves more ocular surface area than any other lens type and may lower oxygen bioavailability due to thicker tear layers, clinicians must objectively monitor for long-term corneal health, as a patient’s subjective comfort may not indicate a healthy ocular surface.

Expanded lens choices and improved designs have helped contact lens specialists better manage irregular cornea patients.

The clinical adoption of corneal collagen crosslinking as a first-line keratoconus treatment can serve to stabilize a majority of patients and reduce the need for constant lens refits. It’s essential to remember that even stable keratoconus patients can manifest different visual demands that may require clinical use of different contact lens technologies across various stages of life. Therefore, clinicians must keep an open mind about offering lens fitting options to achieve best possible outcomes.

It may be tempting to offer a universal solution to all problems by fitting everyone in a single lens modality. However, while scleral lenses may be successful for many patients, some eyes will do better with specialty soft lens designs, corneal GP lenses or hybrid lens designs. Clinicians must carefully assess the unique needs of each person and adhere to the three basic fitting goals. Furthermore, keeping up-to-date with advancements in lens design technologies and diagnostic instrumentation is also of paramount importance for the continual delivery of the best keratoconus care we can offer to our patients.
Newer, more sophisticated instruments are changing the way we view the cornea. A review of the latest diagnostic tools that assess anterior corneal shape and integrity will help clinicians better understand the healthy cornea and what can be considered an abnormal reading. These new instruments help practitioners define disease and the changes in corneal shape and integrity it causes—all of which can affect vision.

THE CORNEA UP CLOSE
Before proceeding, it’s helpful to refresh our memories on the basic corneal anatomy and physiology we all learned in school so that we can better recognize when problems occur. The corneal layers, totaling an average of 540µm in thickness, include the epithelium, Bowman’s layer, stroma, Descemet’s membrane and endothelium. The stroma and epithelium make up the majority of the cornea’s tissue thickness.

The tear film, while a distinct entity, interacts with the corneal epithelium; one cannot exist without the other, and their interactions are an important consideration when diagnosing corneal irregularities.

Due to eyelid friction, the superior epithelium is thinner compared with the inferior region. Greater frictional forces are applied to the superior cornea as the upper lid reaches its maximum speed as it crosses the visual axis. The lid then skims across the inferior part of the cornea. Due to the anatomical difference between the nasal and temporal aspects of the cornea, the epithelium is thinner on the temporal side as the outer canthus is higher than the inner canthus. In addition, the temporal portion of the lid is higher than the nasal side. The organization of collagen fibrils in the cornea is critical to corneal shape and clarity. The preferred orientation of collagen fibrils in the normal cornea is along the vertical and horizontal meridians. There is also a regular increase in collagen mass from the center of the cornea toward the periphery.

The corneal endothelium—a monolayer of hexagonal cells that cover the posterior surface of the cornea—maintains the transparency of the corneal stroma by controlling corneal hydration. These specialized cells do not regenerate, so when cellular abnormalities occur, the corneal stroma swells, changing the collagen arrangement and affecting transparency.

IDENTIFYING PATHOLOGY
The relationship between epithelial and stromal thickness during an episode of edema and corneal desiccation can be monitored with OCT global pachymetry imaging. The maps of the epithelium, stroma and total corneal thickness are useful during routine screening to identify corneal abnormalities such as dry eye disease (DED) and ectasia. OCT imaging is also helpful in observing corneal intrusion with foreign bodies, surgical sutures, corneal dystrophies, infiltrates and corneal ulcerations. Finally, revealing the interaction of the epithelium and stroma also highlights the process of healing and can improve surgical outcomes when the cornea is involved.

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Ectasia

The first stage of ectasia is the thinning of the stroma, although detection becomes difficult due to the corneal epithelium responding by becoming thicker, causing the total corneal thickness to remain unchanged. The only method of detecting the thickness of the epithelium and stroma using global pachymetry is with software that separates the layers—a feature that will become a modification on current anterior segment OCT devices as software improves. The epithelium becomes thicker over the depression and thinner over the elevations, smoothing over the corneal stromal irregularities and creating aberration-free visual acuity during the initial stages of the disease.12

As ectasia progresses, the epithelium begins to thin after the initial thickening stage, and the cornea distorts at the site of the stromal thinning. At the same time, the epithelium thins, forming a base encircling the area of stromal thinning giving support to the structure.13

In ectasia the collagen fibrils shift toward a preferred orientation of 20/160 degrees. There is also an increase in the irregularity of the scattering of the fibrils. The orientation of collagen fibrils inside the cone is altered considerably, while those in the periphery retain the preferred vertical/horizontal orientation.

Corneal topography assumes this change in collagen structure that affects the shape of the cornea.14 In Scheimpflug topography, a spinning mirror provides an enhanced depth of focus without distorting the image. Clinicians can image not only the curvature of the anterior cornea (as with placido-based topography), but also elevation of the anterior cornea, pachymetry and curvature of the posterior cornea. Indices such as center surround index, surface regularity index and standard deviation of the power quantify the change in corneal shape.

The most common index used with Scheimpflug topography is the Belin-Ambrosio Enhanced Ectatic Display, which uses a modified best-fit sphere to classify the shape of the cornea. In a central 8mm zone, the thinnest part of the cornea is excluded (so that it does not skew the best-fit sphere). This enhanced best-fit sphere is compared to the standard best-fit sphere.15 If there is no pathology, those spheres will agree, but the difference between those spheres will be significant when ectasia is present. Studies have found that the Belin-Ambrosio index can predict risk of ectasia prior to refractive surgery.16,17 But, perhaps most impactful for optometrists, research suggests it can be extremely predictive for distinguishing a normal cornea from keratoconus or other non-inflammatory thinning disorders, such as pellucid marginal degeneration.18

Dry Eye

Corneas with DED have exhibited corneal epithelium thickness variation due to inflammation and desquamation of the cellular layer. During the initial stages of DED, the inflammatory processes and epithelium proliferation causes epithelium thickening. As the disease progresses, limbal stem cell atrophy results in epithelial thinning. Because superior and inferior stem cell population in the normal eye is greater in the superior and inferior cornea than the nasal and temporal regions, epithelial thickness change is more dramatic in the peripheral cornea than the central.19

As the tear film becomes more deficient and lubrication is reduced, greater frictional force upon the superior cornea results in an increase of cell loss. Research also shows tear deficient patients increase their blinking rate, causing even greater frictional force on the epithelium and additional cell loss and thinning.20 Investigators suggest long-term soft contact lens wearers also experience a thinning epithelium.21
Corneal Endotheliopathies
Since the endothelium is a 5µm monolayer, it is difficult to observe pathology without the use of a specular microscope. A specular microscope provides useful information when observing dystrophies and effects of contact lens wear. It is also helpful for ensuring the endothelium is functioning properly before presurgical intervention.

Normal cell loss occurs every year but can be exaggerated by uveitis, glaucoma, Fuchs’ dystrophy, contact lens wear and intraocular surgery. When cells die, they slough off the posterior surface of the cornea—compromising the endothelium mosaic and affecting function.

During contact lens wear and presurgical evaluations, visualizing the condition of the endothelial cellular arrangement is crucial to avoid potential damaging effects. Endothelial cell loss can render the cornea ineffective in controlling corneal hydration, resulting in pathological edema.

Contact lenses can induce endothelium change with high pleomorphism and polymegethism but also a decrease in endothelial cell density. Loss of endothelial cell density is correlated with length of time wearing contact lenses. The only method to observe changes in the corneal endothelium while wear contact lenses is with a specular microscope. This becomes important for future positive outcomes from ocular surgery such as cataract extraction and refractive surgery.

Patients might experience clinical signs of contact lens intolerance associates with hypoxia, especially while wearing large diameter scleral lenses or hybrid lenses. In addition to photophobia, contact lens intolerance, foggy, hazy or fluctuating vision, indicate the endothelium might be compromised. Any patient who exhibits signs of reduced vision after a few hours of wear without any other explanation should be examined with a specular microscope. Many of these patients are experiencing endotheliopathies.

CORNEAL INTEGRITY
Corneal hysteresis is a measure of elasticity of the tissue and the ability of the cornea to dissipate energy when the shape is disrupted. The one commercially available device for measuring corneal hysteresis, the Ocular Response Analyzer (ORA, Reichert), records IOP as a puff of air indents the cornea and then as the cornea rebounds to its original shape. The difference in these two IOPs is recorded as corneal hysteresis (CH), which is actually a measure of corneal viscosity (how the cornea resists applied force). Clinical trials have used these two measurements to develop a constant for corneal thickness known as corneal resistance factor (CRF).

Studies show CH and CRF are somewhat variable in healthy subjects, although there is a general trend toward greater CH as healthy patients age. When the cornea is thinned in either a pathological process such as keratoconus, or in a surgical process such as LASIK or LASEK, CH decreases. When the cornea is strengthened in corneal crosslinking, CH increases.

Research shows eyes with pseudophakic bullous keratopathy have significantly reduced CH and CRF, which increase after DSAEK. Low CH is also associated with different forms of glaucoma, including primary open-angle, primary angle-closure, pseudoexfoliative and congenital. Because IOP measurements with Goldmann applanation tonometry may be different in patients with low CH and CRF, some researchers recommend glaucoma patients with low CH and CRF be treated more aggressively.

THE INS AND OUTS OF REFRACTION
Patients come in every day with 20/20 vision, yet still discuss their inability to function at night or see clearly during the day—and the shape of the cornea is often the key.

The normal corneal curvature range is no more than 10D; anything greater than that suggests for corneal thickness known as corneal resistance factor (CRF).

Although ultrasound pachymetry is still the standard for measuring corneal thickness, OCT pachymetry is rapidly taking its place. The inherent error in ultrasound’s hand-driven measurement is hard to overlook compared with a computer-driven tool. Research shows a statistically significant variance between ultrasound and OCT, with ultrasound measuring greater thickness compared with other techniques.

Corneal shape abnormalities that can affect vision. If the power change created by the curvature across the cornea cannot be adequately corrected with spectacles due to the difference between the flattest and steepest curve, contact lenses (often rigid lenses) are indicated.

A combination autorefractor/keratometer can help clinicians uncover corneal shape and surface abnormalities useful for refractive and surgical outcomes.

Depending on the level of distortion seen on an autorefractor’s point spread function, two prescriptions may be necessary, and the power map is key to understanding why. As the power map changes from the apex of the cornea during dim illumination, the prescription will change during dilation.

The axial map measures the range of curvature distributed across the corneal surface. They are spherical biased and calculated from light rays projecting through the optical axis as a reference point. This smooths out the data, which can hide corneal abnormalities.

The instantaneous display or the tangential curvature map, best for understanding the true corneal shape and defining an ectatic cornea, does not use a reference axis and illustrates transitions in curvature with greater sensitivity.

Diagnostic instruments help us identify corneal abnormalities within the ocular system by first defining the initial changes from the normal structure—far sooner than can be seen through a slit lamp. Armed with more knowledge of both normal and abnormal corneas, clinicians can readily identify the initial stages of disease and promptly institute treatment strategies.

Specular microscopy, as seen here, can help clinicians observe changes in the corneal endothelium caused by dystrophies and contact lens wear.

STUMPED: Demystifying Congenital Corneal Anomalies

Understanding the history of corneal anomalies and where they occur are important steps in preventing long-term complications.

By Stephanie Fromstein, OD, and Luis Trujillo, OD

Any can attest to the anxiety that comes with nearly any corneal abnormality, let alone an anomalous finding in a pediatric patient. The sympathetic nervous system causes the influx of a dizzying cocktail of chemicals that evokes a “fight or flight” response. As responsible practitioners tasked with the care of our patients, we have to resist flight, choose to fight and hone in on two critical questions: (1) How can I get this child to sit still long enough for me to examine them, and (2) Why is the cornea not clear?

While the answer to the first question is beyond the scope of this article, several methods exist for organizing congenital corneal anomalies. The acronym STUMPED can be a successful tool to help clinicians recall the various diagnoses:

- S — Sclerocornea and other anomalies of Size
- T — Tears and Trauma
- U — Ulcers
- M — Metabolic
- P — Peters’ anomaly and other mesodermal dysgenesis syndromes
- E — Edema
- D — Dermoids and Dystrophies

These corneal anomalies are almost always diagnosed on clinical findings. Additionally, obtaining adjunct testing on pediatric patients can be quite challenging. A firm grasp on the clinical presentations of many of these corneal irregularities is key to a proper diagnosis.

SCLEROCORNEA AND ANOMALIES OF SIZE

Clinically, sclerocornea is a rare, bilateral clouding of the cornea. The white appearance is consistent with that of the sclera and may be limited to the periphery or spread centrally toward the visual axis. The central cornea tends to be clearer than the limbus, which is nearly indistinguishable from sclera.1,2

Megalocornea and microcornea also represent rare, often bilateral corneal abnormalities. Megalocornea is diagnosed by a corneal diameter greater than 13mm. It is most often seen in males, as it is linked to the X-chromosome. While the cornea is histologically normal in these cases, clinicians should take care to distinguish the condition from buphthalmos, in which the cornea bulges forward secondary to high intraocular pressure (IOP).4 Microcornea, conversely, presents with a corneal diameter of less than 10mm. While the cornea is also normal in this case, the condition is often accompanied by cornea plana and a shallow anterior chamber. Both anomalies require long-term monitoring for glaucoma due to concurrent angle anomalies.4,5

UCELS

Corneal ulcers should be ruled out in any patient presenting with a corneal opacification.1 Pediatric microbial keratitis is rare but potentially devastating, as pediatric eyes mount a severe inflammatory response. These ulcers are most often caused by herpetic or bacterial infections, and they warrant immediate referral for treatment. While pediatric treatment is generally the same as adult treatment, it requires extra care to ensure nontoxicity due to blood absorption.8

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METABOLIC CONDITIONS

Children with corneal deposits may present with several congenital metabolic disorders:

**Cystinosis.** This rare autosomal recessive disease results in widespread deposition of cysteine crystals in the cornea, conjunctiva, iris, lens and retina. Aside from potentially fatal renal failure, corneal crystals are often the first extra-renal findings.4,5,9

**Mucopolysaccharidosis.** Caused by a missing enzyme, this lysosomal disorder breaks down glycosaminoglycan.1-3 Patients present with bilateral corneal opacities, which can be diffuse. Additional findings of glaucoma, retinal abnormalities and optic nerve swelling, all of which can be confirmed with laboratory studies or conjunctival biopsies, may aid in diagnosis.1

**Fabry’s disease.** This lysosomal storage disorder results in pathognomonic vortex keratopathy and wedge-shaped cataracts. Corneal findings can be differentiated from medication-induced deposits of similar appearance based on pharmaceutical history. Conjunctival and retinal vascular changes, third nerve palsy and nystagmus may support the diagnosis, and classic skin findings such as angiokeratoma corporis diffusum can confirm diagnosis.4,6

**Tyrosinemia type 2.** High plasma tyrosine levels that cause a recalcitrant pseudodendritic keratitis characterize this condition. Palmar and plantar hyperketatotic lesions are common and can aid in diagnosis.4,6

**Wilson’s disease.** This is a ceruloplasmin deficiency that leads to widespread copper deposition throughout the body. Clinical signs include Kayser-Fleischer rings, which are darker versions of arcus or sunflower cataracts or both. These patients also suffer from liver disease, basal ganglia dysfunction and psychiatric disturbance.4,6

**PETERS’ ANOMALY AND OTHER MESODERMAL DYSGENESIS SYNDROMES**

Peters’ anomaly is the final stage in a group of bilateral hereditary disorders known as the mesodermal dysgenesis syndromes, which also includes posterior embryotoxon and Axenfeld-Rieger syndrome. All three entities arise from faulty cleavage of angle structures and, as such, predispose patients to glaucoma.5,6

In isolation, posterior embryotoxon, a thickened and anteriorly displaced Schwalbe’s line visible at the corneal periphery, common and mostly benign. It can, however, be seen in conjunction with Axenfeld-Reiger syndrome in a spectrum of disorders including peripheral anterior synechiae and iris defects. Posterior embryotoxon can have associated dental, facial and skeletal abnormalities. Patients with Peter’s anomaly, a rare and serious bilateral condition, can present with features of both posterior embryotoxon and Axenfeld-Reiger, as well as central corneal opacification and iridocorneal and iridolenticular adhesions. Patients with any of the aforementioned conditions should be monitored closely for glaucoma.5,7

**EDEMA**

See discussion of congenital hereditary endothelial dystrophy (CHED) below.

**DERMOIDS AND DYSTROPHIES**

Limbal dermoids, one of three types of conjunctival choristomas, are mostly comprised of collagen and are typically located inferior-temporally at the limbus. Under slit lamp examination, a dermoid is well circumscribed, elevated and can appear fleshy to pale yellow in color. Treatment calls for excision, as the growth is superficial.3 Although limbal dermoids are present unilaterally, a bilateral presentation is typical of children with Goldenhar’s syndrome, who will also present with colobomas of the lid and iris or total aniridia.3

Corneal dystrophies are bilateral, symmetric and slowly progressive. Some present in a neonatal population, while others classically manifest after the first decade of life. They show a hereditary pattern—mostly autosomal dominant—and often can be identified based on a strong family history. This familial preponderance represents the easiest method of diagnosis in the pediatric population, as well as some insight into the probable natural course of the condition.

Another strategy for diagnosis is location: dystrophies tend to target the central portion of a single corneal layer, dramatically narrowing the list of differentials from innumerable to several. Within a given layer,
STUMPED: DEMYSTIFYING CONGENITAL CORNEAL ANOMALIES

dystrophies can often be differentiated based on clinical presentation along with any accompanying signs or symptoms; ability to describe these will, of course, be dependent on patient age. In addition, optical coherence tomography can be a useful tool in localizing corneal changes and deposits and further aiding in diagnosis.

Generally, there are few other associated ocular or systemic findings to complicate the picture with corneal dystrophies. Here is a summary of the most common dystrophies affecting the pediatric population:4-7

**Epithelial.** Meesmann and Lisch epithelial dystrophies are similar entities presenting with diffuse (Meesmann) or localized (Lisch) epithelial microcysts and vesicles. As these rupture onto the epithelial surface, minor pain and opacification may be noted. The conditions are relatively benign and management is often palliative. In contrast, gelatinous drop-like dystrophy presents with pronounced central raised mounds of amyloid in the epithelium, resembling myriad fruit from mulberry (early childhood) to a late-stage yellow kumquat appearance. This condition is more likely to warrant keratectomy referral.4-7

**Bowman’s layer.** The three conditions classified under Bowman’s layer dystrophy were once thought to be the same etiology and only recently have been distinguished, based on microscopic appearance and clinical presentation.

**Stroma.** Congenital stromal dystrophy—a cousin of CHED—is a rare condition causing congenital opaque, flaky clouding of the corneal stroma. The condition often results in marked corneal edema.4-7 Granular dystrophy and lattice dystrophy are common autosomal dominant conditions leading to stromal deposition of hyaline and amyloid deposits in a diffuse and branching pattern, respectively. A third condition, Avellino dystrophy, combines features of both and is named after the original group of affected individuals from Avellino, Italy.4-7 Central cloudy dystrophy of Francois and Schnyder’s dystrophy both involve diffuse central deposition of cholesterol in the corneal stroma early in life, and the latter should be worked up for elevated serum cholesterol levels.4-7

**Endothelium.** Similar to congenital stromal dystrophy, CHED is also characterized by congenital bilateral corneal clouding—from mild haze to a milky appearance—with an accompanying thickened Descemet’s membrane and generalized corneal edema. This is another condition that should be differentiated from congenital glaucoma, as these patients often have high IOP readings secondary to endothelial dysfunction and thickened corneas.4-7 Posterior polymorphous dystrophy is also relatively common in the pediatric population. In this condition, endothelial cells act like epithelial cells, propagating in linear, band-like or grouped configurations. These cells invade the angle and the anterior iris, causing elevated pressure, iris distortion and abnormalities.4-7

While corneal anomalies in a pediatric population may seem daunting, an understanding of the history of the findings, as well as a clinical examination to determine the layer in which the condition is found, can significantly narrow the list of differentials. While many conditions are slowly progressive and largely benign, keeping a close eye on sight-threatening conditions, both direct (elevations in IOP) and indirect (form-deprivation amblyopia, especially in unilateral conditions), is critical in preventing long-term complications.

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Corneal dystrophies are typically defined as hereditary, bilateral, progressive alterations to the cornea not associated with systemic disease or prior inflammation. IC3D classification of corneal dystrophies is anatomically based and divided into four categories: epithelial and subepithelial dystrophies, epithelial-stromal transforming growth factor beta-induced (TGFBI) dystrophies, stromal dystrophies, and endothelial dystrophies. This article provides an overview of signs and symptoms associated with various corneal dystrophies and explores options to manage patient symptomology.

EPITHELIAL AND SUBEPITHELIAL DYSTROPHIES
Epithelial basement membrane dystrophy (EBMD), also known as map-dot-fingerprint or Cogan’s microcystic dystrophy, is common and is caused by abnormal epithelial basement membrane adhesions usually occurring as a consequence of degenerative changes or trauma. Rarely, it may be due to an inherited mutation. Clinical signs include irregular, hazy areas of epithelium in map, dot or linear patterns that negatively stain with fluorescein. Patients may be asymptomatic or complain of photophobia, pain secondary to erosions or decreased visual acuity (VA) due to irregular astigmatism or corneal opacity.

Meesmann corneal dystrophy (MECD) is a less common epithelial layer dystrophy inherited autosomal dominantly that presents clinically as hazy epithelium with microcysts. Of those affected, 85% will show microcysts covering the entire cornea. Although MECD patients may be asymptomatic, some may complain of symptoms similar to those found in EBMD such as decreased VA, light sensitivity or pain.

EPITHELIAL-STROMAL TGFBI DYSTROPHIES
Reis-Bucklers dystrophy is inherited in an autosomal dominant manner and leads to damage and focal opacities in Bowman’s membrane and the anterior stroma. Clinically, these opacities will be ring-shaped and most dense centrally, although they can involve the entire cornea. VA is usually reduced from a young age and will slowly deteriorate over time. Patients may also complain of pain secondary to corneal erosions.

Lattice dystrophy is also an autosomal dominant inherited dystrophy. This gene mutation leads to amyloid protein deposits in the anterior stroma that present clinically as linear opacities. Patients may complain of pain secondary to erosions or decreased VA.

Granular dystrophy is caused by an autosomal dominant gene mutation that leads to hyaline protein deposits in the anterior stroma. These deposits are bread crumb-like and increase in size and number with age (Figure 1). Patients may complain of glare, photophobia, painful erosions or decreased VA.

Differentiating the various etiologies can be difficult, but with the right training and diagnostic tools, you can find the right diagnosis and manage the patient accordingly.

ABOUT THE AUTHORS

Dr. Frantzis is a Cornea and Contact Lens resident at SUNY College of Optometry. She specializes in complicated contact lens fittings and diagnosing and managing patients with corneal anomalies.

Dr. Duchnowski is the section chief of Contact Lens Service at the University Eye Care Center and the director of Cornea and Contact Lens externship at SUNY College of Optometry.
Avellino dystrophy, also known as granular type 2 or combined granular-lattice dystrophy, is inherited in an autosomal dominant fashion and leads to both hyaline and amyloid deposits in the stroma.1 Bread crumb-like opacities are seen similar to those in granular dystrophy, but in combination with deeper stromal refractile lines such as those found in lattice dystrophy (Figure 2). Patients may complain of glare, photophobia, painful erosions or severely decreased VA.

STROMAL DYSTROPHIES

Macular dystrophy is inherited in an autosomal recessive manner.1 This gene mutation leads to a defect in corneal glycosaminoglycans, which presents clinically as anterior stromal opacities similar in shape to those found in granular dystrophy, but with greater severity.1 Patients may complain of glare, photophobia, painful erosions or severely decreased VA.

Schnyder crystalline corneal dystrophy, autosomal dominantly inherited, presents before the third decade of life.1 Corneal findings vary with age: patients 23 and younger present with a circular central stromal haze, those between 23 and 38 present with arc-shaped bands of haze in the midperipheral cornea and patients older than 38 often have dense stromal haze in the limbal region.1 VA decreases over time and glare symptoms increase.

ENDOTHELIAL DYSTROPHIES

Fuchs’ endothelial dystrophy may be inherited or sporadic.1 Endothelial cells slowly die, disrupting normal fluid gradients in the cornea. Clinical signs include endothelial guttata, stromal or epithelial edema, bullae, low endothelial cell counts or even corneal scarring with chronicity (Figure 3). Patients may complain of painful erosions or decreased VA worse in the mornings.

Posterior polymorphous dystrophy is caused by a gene mutation that affects Descemet’s membrane and the endothelium.1 Clinical findings may be asymmetric and manifest as linear or vesicular changes with irregular, scalloped edges at the level of Descemet’s membrane. Of these patients, 25% will also have iridocorneal adhesions, while 15% will have elevated intraocular pressure (IOP).1 Patients may be asymptomatic or present with decreased VA due to corneal edema and pain if corneal bullae develop.

Congenital hereditary endothelial dystrophy, inherited in an autosomal recessive manner in the majority of cases, presents at birth with bilateral cloudy corneas.2 Additional findings include thicker than average corneas, highly reduced endothelial cell counts and nystagmus. Less frequently, patients present with band keratopathy and elevated IOP.1

MANAGEMENT

As there is no cure for these dystrophies, treatment should focus on managing patient symptoms.

Many superficial corneal dystrophies present with recurrent corneal erosions (RCE), the management of which may be frustrating for doctors and patients. Initial topical treatments include lubricants, antibiotics and hyperosmotics. Doxycycline and corticosteroids, which inhibit the action of metalloproteinases that break down the epithelial basement membrane, may be useful.2

Fig. 2. This 38-year-old female with Avellino corneal dystrophy was symptomatic for decreased vision, with BCVA of 20/30.

Fig. 3. At left, corneal guttata of a 43-year-old female as seen on slit lamp. At right, corneal guttata as seen with a specular microscope.
Autologous or umbilical cord serum are pharmacologic options for patients who exhibit minimal RCE improvement with topical treatments. Umbilical cord serum has a high concentration of growth factors, and research shows it decreases the frequency of erosions compared with lubricants alone. Contact lens options are also available for patients who fail with other topical treatments. Bandage lenses can help protect the epithelium from mechanical forces with efficacy similar to topical lubricants, and patients may experience earlier pain relief. Scleral lenses can potentially resolve epithelial defects by providing mechanical protection and continuous hydration to the epithelium (Figure 4). Research also suggests amniotic membranes can promote ocular surface healing with minimal adverse effects. For recalcitrant cases, surgical options include micropuncture, diamond burr debridement, phototherapeutic keratectomy and alcohol delamination.

When patients present with decreased VA, clinicians should take corneal topographies to evaluate for the presence of irregular astigmatism. In these cases, gas permeable, scleral and hybrid lenses are viable options for visual improvement. For patients with more severely decreasing VA, surgical options include phototherapeutic keratectomy, a safe and effective technique, especially for the anterior corneal dystrophies. It removes superficial corneal tissue, and research shows it reduces corneal erosions and improves VA. Traditional penetrating keratoplasty (PK) or deep lamellar keratoplasty (DLKP) are other surgical options. Investigators showed DLKP had favorable outcomes in lattice dystrophy, while it progressively decreased endothelial cell density in patients with macular dystrophy. It is possible some dystrophies will recur after any of these surgical procedures, and if indicated, the procedure may be repeated. Research suggests Reis-Bucklers, granular and lattice dystrophies have high rates of recurrence.

Many of the corneal dystrophies are capable of resulting in corneal edema. Traditional treatment of corneal edema includes the use of topical lubricants and hypotonic drops or ointments. The surgical options available for these patients aim to replace the dysfunctional endothelium. Descemet’s stripping automated endothelial keratoplasty or Descemet’s membrane endothelial keratoplasty is preferred over traditional PK for patients with only endothelial dysfunction because of decreased likelihood of serious adverse effects such as graft rejection.

Rho-associated kinase (ROCK) inhibitors may play a role in the future management of corneal edema, as these drugs have been shown to promote endothelial cell proliferation. Case reports found ROCK inhibitors to be successful in improving corneal clarity and vision, while also significantly decreasing corneal edema.

Corneal collagen crosslinking has been proposed as yet another way of managing corneal edema. Crosslinking is currently used for preventing the progression of corneal ectasia, but also has the ability to reduce stromal swelling. Case studies show a modified corneal crosslinking procedure was successful in decreasing corneal thickness and improving visual fluctuation in association with corneal edema.

Corneal dystrophies can be challenging to diagnose and manage. It’s imperative we handle these cases in a timely fashion, as they can profoundly impact our patients’ lives. Fortunately, our knowledge and treatment options continue to expand, and we look forward to further developments to better our patients’ quality of life.

Fig. 4. At left, a 50-year-old female with EBMD. Her BCVA was 20/60. At right, this patient was successfully fit in scleral lenses and achieved 20/20. The optic section of the fit shows, from left to right, the front surface of the lens (white hyper-reflective beam), contact lens thickness (black band) and corneal vault (green band)—the space between the contact lens back surface and the cornea.

CE TEST - MARCH 2017

1. Which of the following dystrophies is most likely to present with reduced vision in childhood?
   a. Epithelial basement membrane dystrophy.
   b. Fuchs' endothelial dystrophy.
   c. Reis-Bucklers dystrophy.
   d. None of the above.

2. Amyloid protein deposits are the cause of opacities in which corneal dystrophy?
   a. Lattice dystrophy.
   b. Epithelial basement membrane dystrophy.
   c. Granular dystrophy.
   d. Posterior polymorphous dystrophy.

3. What is the most common cause of epithelial basement membrane dystrophy?
   a. Degenerative changes or trauma.
   b. Inheritance.
   c. Systemic disease.
   d. Systemic medication.

4. Which corneal dystrophy is best described as a combination of bread crumb-like and linear opacities in the corneal stroma?
   a. Granular dystrophy.
   b. Lattice dystrophy.
   c. Posterior polymorphous dystrophy.
   d. Avellino dystrophy.

5. Which corneal dystrophy is most likely to present with iridocorneal adhesions?
   a. Epithelial basement membrane dystrophy.
   b. Posterior polymorphous dystrophy.
   c. Granular dystrophy.
   d. Fuchs' endothelial dystrophy.

6. Which of the following is an option in the management of RCE?
   a. Scleral contact lenses.
   b. Soft contact lenses.
   c. Umbilical cord serum.
   d. All of the above.

7. Why may a modified collagen crosslinking play a potential role in the management of Fuchs' endothelial dystrophy?
   a. It reduces swelling capacity.
   b. It prevents progression of corneal ectasia.
   c. It restores endothelial cell density.
   d. It thickens the damaged endothelium.

8. Scleral contact lenses may be indicated in the management of corneal dystrophies for:
   a. Improving visual acuity.
   b. Decreasing frequency of erosions.
   c. Promoting endothelial cell function.
   d. Both a and b.

9. Which surgical procedure would most likely be performed on a patient with Fuchs' endothelial dystrophy?
   b. Phototherapeutic keratectomy.
   c. Diamond burr debridement.
   d. Descemet stripping automated endothelial keratoplasty.

10. Phototherapeutic keratectomy is a procedure that:
    a. Removes superficial corneal layers with a laser.
    b. Transplants a donor epithelium and stroma.
    c. Transplants a donor Descemet's membrane and endothelium.
    d. Strengthens corneal bonds.

EXAMINATION ANSWER SHEET

Get to Know Your Dystrophies
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3. __ __ __ 8. __ __ __
4. __ __ __ 9. __ __ __
5. __ __ __ 10. __ __ __

Evaluation questions (1 = Excellent, 2 = Very Good, 3 = Good, 4 = Fair, 5 = Poor)
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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: Complex __ __ __ __ __
18. How long did it take to complete this course? _________________________
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   __________________________________________________________________
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Take a **New Look** at

Colored Lenses

New colored contact lens modalities and expanded technologies offer opportunities for both patient care and practice growth.

By Jane Cole, Contributing Editor

Today's colored contact lenses have evolved beyond the days of special occasion or weekend-only novelty for many patients. Thanks to technological advances, these lenses now have healthier, more breathable options with silicone hydrogel material. They are also now available in monthly and daily options, distance vision correction and plano. Still, some optometrists don’t actively market colored contact lenses, creating a void of untapped potential. Practice management consultant Gary Gerber, OD, says the historical baggage that comes with this line of products creates an impediment for doctors to prescribe these lenses as often as they could.

But doctors should make a point to stay up-to-date on the new advances in this category. “Those docs willing to break old habits of ‘no one wants colored lenses’ will do great once they actually start using them,” says Dr. Gerber.

In recent years, manufacturers have rolled out new designs that have redefined the way many think about colored contact lenses. Some of the newer generations of lenses even have technology that enhances the limbal ring. “I have some people in these lenses who love them, and they think the colored contact lenses make them look younger,” says Mile Brujic, OD, of Premier Vision Group.

Here, your colleagues offer practice management tips on how to best market colored contact lenses and maximize their potential in your practice.

**IT’S AS SIMPLE AS ASKING**

Dr. Brujic and his staff take a proactive approach to colored contact lenses and mention the option at the beginning of the eye exam instead of waiting until the end of the visit, which could result in increased chair time. “If you go through the whole exam and then ask the patient, ‘Do you have any other questions?’ and the patient says, ‘Yes, I’m interested in color contact lenses,’ it’s like you are ready to walk out of the room, and now you’re in a reset mode. So being proactive on the front end is important. Have a discussion with patients at the beginning of the exam and let them know whether or not they’re candidates,” says Dr. Brujic.

His staff plays a critical role in marketing contact lenses. During the pretest, they are encouraged to ask patients if they are interested in colored contact lenses. “Ideally, practitioners should never have to ask. They should be able to walk into the exam room and know already if the patient is interested in colored lenses.”

Kiranjit Bedi, OD, of America’s Best Contacts & Eyeglasses, part of National Vision, says one of the first questions her staff asks during contact lens visits is whether they are interested in clear or colored lenses. “This plants the seed at the front desk for patients to start thinking about questions they may have, or for them to consider something new,” says Dr. Bedi.

Sometimes, patients simply have never considered the possibility of wearing colored lenses, don’t know that a separate exam isn’t necessary or don’t realize colored lenses are an option for them.

**HEALTHIER MODALITIES**

In terms of colored contact lens technology, there hadn’t been a great deal of change in the industry until a recent silicone hydrogel lens with a Dk value of 110 came on the market, Dr. Bedi says. This has provided a healthier alternative for her patients, especially ones tempted to sleep or nap in their lenses, despite discussions about associated risks, she adds.

“I used to shudder when patients would ask to be fit in
colors, only because the technology was not keeping up with the advancements clear contact lenses were making,” says Dr. Bedi. “For some time, I noticed patients were asking for colors less frequently or were purchasing a clear counterpart to their colored lenses. When I probed this pattern, my patients would simply explain they wanted more comfortable contacts for the majority of the week, and ‘save’ the discomfort and color change option for the weekends or special events.”

**PATIENT SELECTION**

Sometimes patients like colored contact lenses for cosmetic reasons. “Women especially appreciate the opportunity to try tint if the options are available in their current lenses,” says Glenda Secor, OD.

However, it’s not the only reason to fit colored lenses. “What they make their eyes look like is just a fringe benefit. I have patients who wear colored contacts because they are easier to see in the case,” says Dr. Brujic.

While women make up the majority of patients who want colored contacts, some men are interested in them as well. For example, Dr. Brujic says he recently fit a male patient into a plano-power colored contact lens post-cataract surgery because the patient liked the way the lenses made him look.

Sometimes patients like colored lenses because they make them look younger, he adds. However, Dr. Brujic also had a patient with mild peripheral iris atrophy after cataract surgery who is now wearing a color contact lens to stop peripheral stray light entering the eye through the area of atrophy.

So with colored contacts, “you will never know unless you ask,” Dr. Brujic says.

“Men also like the option, but are more hesitant sometimes to ask,” echoes Dr. Secor. “Just suggesting an option is always helpful.”

Some doctors see every patient as a candidate for colored lenses. “Our patients wearing colors range in age from 16 to 75,” says Dr. Bedi. “Repeat patients you have established a relationship with are easier to recommend to. After seeing patients on a consistent basis for a few years, you become aware of how adventurous they may be. We, as providers, have always thought this way when we recommend glasses, but we often don’t consider patients may want different options with their contacts.”

Perhaps we know a patient likes to spend time at the beach, and therefore we offer a sunglass prescription,” Dr. Bedi explains. “We should also have this mindset when prescribing contacts as well. Changing your look is easy with color contacts, and it’s something patients often don’t think of unless you point it out.”

**MAKE YOUR MARKETING COLORFUL**

With colored contact lenses, seeing, often, really is believing. Your staff can promote colored contact lenses simply by wearing them. “Having your staff wear the contacts helps patients visualize themselves wearing them,” says Dr. Bedi. “We forget how commonly we compliment each other on our shoes, clothes, hair and other things, and this often inspires our own choices. We forget how we make our decisions of what to buy based on others’ experiences. We read reviews, we ask questions, and what better than to have a member of your staff able to express exactly what they feel while wearing the lenses.”

Another marketing plus is that the manufacturers already provide material to practices at no cost to the doctor. Contact lens representatives can help with trials, pamphlets and rebates, in addition to providing a wealth of knowledge as to what new advances are on the horizon and, conversely, what may be less popular. Dr. Bedi says one particular representative was instrumental in her spreading the news about a new silicone hydrogel colored lens before they were available on the market. “I started planting the seed with information, encouraging my patients to come back when the lenses were available.”
TAKE A NEW LOOK AT COLORED LENSES

available.”

Many manufacturers also offer the opportunity for patients to see themselves in the lenses through their online color studios, allowing patients to upload a photo of themselves and virtually “try on” different colored contact lenses and then share via e-mail or through their social media sites. “Our office encourages patients to try the lenses and take pictures to post or share with their friends on social media,” says Dr. Bedi. “This not only helps them make a decision on which color to pick, but is a great way to spread the word about colored contacts.”

When it comes to pricing, doctors shouldn’t be concerned about setting a premium on colored contact lenses, says Dr. Brujic. “It’s remarkable what people will pay for ‘want’ vs. what they need. And contacts in general are a want for most individuals, and cosmetic lenses are just a high level of want.”

PART-TIME WEARERS

If a patient isn’t interested in cosmetic or colored contact lenses for full-time wear, the opportunity is still there for part-time wear, including daily disposable options. This can also be an option for serial contact lens wearers who occasionally may want to wear colored contact lenses for special occasions. “Just keeping those options in mind is important, so have them in the bag just in case anyone wants colored lenses on a part-time basis, or you think it would fit a patient’s lifestyle,” says Dr. Brujic.

NO ADDITIONAL CHAIR TIME

Colored contact lenses shouldn’t necessarily take any extra time, Dr. Gerber says, as the process is often delegated to staff, and the doctor just confirms the final selection. “All that’s necessary is to put a colored lens on (one different color on each eye) and ask the patient which one they like better,” says Dr. Gerber.

Dr. Bedi agrees. “Additional chair time is not an issue when you know your patient and if this is a suitable modality for them.” A quick way to minimize chair time is to have the majority of information available through the staff and pamphlets in the waiting area. “By the time the patient gets to your chair, it should be a simple discussion of expectations and answering any leftover questions.”

Another smart strategy to ensure colored contact lens fittings are patient-friendly is to keep trial colored lenses in your office. That way, when patients settle on a color they prefer, you can have them try on the trial lens and check to make sure their vision isn’t impaired before they leave the office. “Although we talk about opportunity, there are still individuals for whom the lens won’t center properly, which interferes with the color and the pupil and results in poor visual outcomes,” says Dr. Brujic. “Night vision is sometimes not as good for the same reason. All other issues are similar to those with wearing other types of contact lenses.”

Colorful
Fitting Pearls

When fitting colored contact lenses, Dr. Secor offers these tips:

Eye color matters: Certain tints work better on certain eye colors. Brown eyes need an opaque design, while translucent lenses, or lighter tints, work well on lighter eyes.

Ask around: Dr. Secor suggests asking all patients if they are interested in enhancing their eye color. “This can be on the intake form or staff pretesting to indicate to the doctor any patient interest,” she says.

Dig a little deeper: If the patient answers “yes,” the doctor can ask this follow-up question, “Do you want a subtle or dramatic change?” Dr. Secor says. This narrows options and saves chair time, she adds.

Women vs. men: Women are usually easy because they understand fashion, she says. Men can be more elusive, but they may be interested in the lens if you suggest it.

Choices are good: Prescribing contact lens designs that also have color choices and visibility tints can make fitting colored lenses even easier because you can fit the lens and then simply change to the tinted version, she adds.

Colored contact lenses offer practice-building opportunities, and you might not know a patient is interested in colored contact lenses unless you ask. “It always shocks me when patients surprisingly ask, ‘I can wear colors?’ In this day and age, we want what’s cool, the newest and latest, and what is on trend,” says Dr. Bedi.
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Cosmetic Contact Lenses for the Deformed Iris

This simple alternative provides added benefits.

**ABSTRACT**

This case report illustrates how silicone hydrogel cosmetic lenses can improve the appearance of disfigured eyes while also providing the benefits of disposability, low cost and alleviation of any ocular hypoxia from low Dk hydrogel lens wear. Following the report is a discussion of the treatment’s success and the additional advantages it provides.

**Keywords:** Disfigured eyes, prosthetic lenses, silicone hydrogel, cosmetic contact lenses, ocular hypoxia

**INTRODUCTION**

Prosthetic contact lenses can improve the appearance of a disfigured eye, whether due to disease, trauma, or congenital disfigurement or defect. However, they also come with some disadvantages, including non-disposability, relatively high cost and the lack of silicone hydrogel material. Clinicians can often overcome these disadvantages by prescribing silicone hydrogel color contact lenses. These contact lenses, normally used to change eye color, can aid disfigured eye appearance with the convenience of disposability without inducing ocular hypoxia.

**CASE REPORT**

**History**

A 54-year-old female presented without complaints with a history of a perforating corneal infection 18 years earlier in the left eye, which resulted in a cataract extraction with intraocular lens (IOL) implantation, iris reconstruction and penetrating keratoplasty (PK). She habitually wore Freshlook Colorblends contact lenses (Alcon) and used OTC readers for deskwork, but did not own any prescription glasses. Systemic health history was noncontributory, and she denied taking any regular medication. She reported an allergy to Methiolate (thimerosal, Eli Lilly).

**Diagnostic Data**

The patient’s extraocular motilities and automated screening visual fields were full. No afferent pupil-lary defect was noted and color vision was normal. Intraocular pressures with noncontact tonometry were 16mm Hg OD and 18mm Hg OS. Automated keratometry was 44.00 at 156/45.00 at 066 OD and 44.00 at 085/47.25 at 175 OS. Manifest refraction was -9.75 -0.50x150 OD yielding 20/20 and -1.25 -3.25x099 OS yielding 20/25+. Accommodative testing yielded a +2.25 near add.

Biomicroscopy showed normal lids and lashes, with moderate conjunctival hyperemia in each eye. The right cornea was unremarkable, while her left cornea showed a clear penetrating corneal graft. Anterior chambers were free of inflammation. Her right iris was normal, but her left iris showed previous traumatic damage with suture loop (Figure 1). The right crystalline lens was clear. A posterior chamber IOL implant in the left eye was centered and free of capsular opacification. Funduscopy showed healthy optic nerves with cup-to-disc ratios of 0.3 and 0.4 with normal maculae, vasculature and retinal peripheries.

**Diagnosis and Treatment**

My clinical impression was status-post PK OS, pseudophakia OS and anisometropia. Due to generalized conjunctival hyperemia, I re-prescribed the patient silicone hydrogel colored contact lenses (Air Optix Colors, Alcon), moving her from a relatively low-Dk hydrogel material, phemfilcon A (Dk/t: 20 at -3.00D) to a higher one in lotrafilcon B (Dk/t: 138 @ -3.00D).

The lenses had 8.6 base curve and 14.2mm diameter OU with -8.00 DS OD yielding 20/25 and -2.00 DS OS yielding 20/80. Spherical over-refraction was -0.75 DS OD yielding 20/20- and spherical-cylindrical over-refraction was PL-2.25x090 OS yielding 20/25. The patient understood the right lens power was at the parameter limit of -8.00 and that there was residual astigmatism in the left eye.

**ABOUT THE AUTHOR**

Dr. Chou is a partner at EyeLux Optometry in San Diego, CA, where he directs a referral-based keratoconus clinic.
**Progress Visit**
At a subsequent progress visit, the patient reported a significant reduction in the conjunctival hyperemia and improved comfort. She was also pleased with the cosmetic concealment of the iris disfigurement (Figure 2). The contact lens prescription was finalized with no further changes and near-variable focus spectacle lenses were prescribed for use over the contacts.

**Discussion**
Prosthetic contact lenses can improve cosmetic appearance of disfigured eyes, social relationships and quality of life. Hand-painted lenses are a common treatment option for these patients. My patient, who had previously received a hand-painted prosthetic lens for the left eye, said she didn’t wear it because she felt it did not sufficiently match the appearance of her right eye.

In addition to the potential for mismatching, their lack of reproducibility, disposability and availability in silicone hydrogel materials are all drawbacks, further compounded by relatively high cost.

Commercially available silicone hydrogel colored contact lenses can overcome most, if not all, of these drawbacks. The higher Dk silicone hydrogel material can minimize complications from ocular hypoxia. Research shows a reduction in conjunctival hyperemia when patients move from hydrogel to silicone hydrogel lenses—which was certainly the case for this patient. Although she did not demonstrate corneal neovascularization from hypoxia, silicone hydrogel lens wear is preferable to hydrogel lens wear to reduce the risk of neovascularization, which is associated with an increased risk of corneal allograft rejection.7

The patient was pleased with her visual outcome, in part because I set realistic expectations prior to re-prescribing, including awareness that the goal was not a perfect cosmetic match but a general improvement. In the same manner that oculoplastic surgeons typically perform blepharoplasty bilaterally even when the patient complains of unilateral blepharochalasis, a better cosmetic outcome likely results from prescribing the same colored contact lenses bilaterally. Of course, if the patient is functionally monocular, it is appropriate to counsel the patient about the risk of microbial keratitis in the good eye. If the patient consents to bilateral contact lens wear, he or she should be reminded of the necessity of minimizing the risk of infection, including hand washing, proper disinfection and daily lens removal.6

In addition to the cosmetic benefit and reduction of ametropia, the patient felt there was reduction of her light sensitivity, likely attributable to the color print of her new contact lenses blocking light that would otherwise enter the enlarged and irregular pupil—and she wasn’t the first patient of mine with this benefit. Two other patients complaining of glare following laser peripheral iridotomies found some relief when wearing silicone hydrogel colored contact lenses. Research shows exposed and partially exposed iridotomies are more frequently associated with glare-type symptoms than iridotomies covered by the superior lid. These experiences suggest that colored contact lenses may bring functional benefit by blocking out light for patients complaining of glare after iridotomy.

To be sure, colored contact lenses are not suitable for all patients with disfigured eyes, and prosthetic contact lenses will continue to play an important role. But some patients are happy with the cosmetic improvement provided by silicone hydrogel colored contact lenses—not to mention their disposability, silicone hydrogel materials and relatively low cost.

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Dendrites: Misdiagnosis Not Permitted

Their presence could play an important role in diagnosing and treating herpetic disease.

Imagine a non-contact lens wearer presents at your clinic with unilateral keratitis. If I were to ask you for the likely diagnosis, most of you would (appropriately) ask for more information. But you actually don’t need more information to create your primary differential because for any unilateral keratitis without a compelling history, herpes simplex virus (HSV) keratitis will always be near the top of your differential until examination and patient history prove otherwise.

If, after examining the patient, you can’t think of a reason it’s not HSV—if there isn’t another, more likely culprit—then call it herpes simplex and treat it as indicated and observe for a response.

A VIRUS WORTH KNOWING

This is an unspoken rule in cornea clinics because of how widespread the virus is and its wide range of clinical manifestations. First, in the United States alone, there are 500,000 cases of ocular HSV per year.1 Just about every adult carries HSV serotype 1, and a smaller percentage carries serotype 2, so most adults have some risk. Next, the virus has significant predilection for the eye. HSV keratitis is the most common cause of infectious blindness in industrialized countries.1 Finally, HSV keratitis can present as essentially any form of keratitis, including vesicular pre-dendrites, dendritic epithelial disease, sub-dendritic stromal keratitis, neurotrophic epithelial disease, marginal keratitis, nummular stromal keratitis, diffuse stromal keratitis, necrotizing stromal keratitis, interstitial keratitis (with this, instead of thinking syphilis, think HSV), corneal rings, disciform endothelitis and linear endothelitis.

All corneal tissue is at risk for developing HSV involvement, and it can look like just about anything. Unless the exam and history are suggestive of an alternate pathology, you’ll rarely be wrong in treating a unilateral keratitis as HSV and assessing for a response.

Because it is such an important diagnosis, my next few columns will discuss some of the finer elements of identifying and working with patients you suspect might have HSV. As dendritic keratitis theoretically sets the stage for all other forms of HSV keratitis, let’s discuss the dendrite—its clinical behavior, role in the natural history of other HSV keratitides and treatment, which is unique compared with all other forms of HSV keratitis.

DENDRITE UP CLOSE: DIFFERENTIAL

Obviously, having successfully completed optometry school (sorry, student readers), you are aware of dendrites and would never, ever misdiagnose them (Figure 1). Despite the consistency with which infectious epithelial HSV forms dendriform lesions, other conditions may occasionally produce branching epithelial lesions as well, which can lead to some clinical head scratching. Thygeson’s superficial punctate keratitis, for example, generally produces small, round lesions, but may occasionally take on a dendritic appearance. In these cases, the greater number of lesions the smaller size and the (typical) involvement of both eyes suggests Thygeson’s. Zoster-related pseudodendrites or zoster mucous plaque keratitis can each mimic HSV dendrites on gross examination, but on closer evaluation neither of these lesions is ulcerated. HSV keratitis is the most frequent initial misdiagnosis of Acanthameoba keratitis (AK), which in its early stages takes on a pattern epitheliopathy that may have a dendritic distribution; once again, however, these lesions will be non-ulcerated and may have perineuritis. Other rarer causes of dendriform lesions also exist, but given the unique elements of each, the diagnosis of HSV dendritic keratitis should generally be straightforward.
forms are inflammatory sequela or a mix of inflammatory and infectious mechanisms. This is important for two reasons:

1. Because dendrites are true infectious ulcerations, we treat them with a topical or oral antiviral and leave out the steroid. Perhaps less known, however, is how unique this treatment is in the spectrum of HSV keratitis. For nearly all other forms of HSV keratitis, the appropriate treatment is pairing a steroid and antiviral, even in cases when a preceding dendrite has not fully resolved (Figure 2). If it’s HSV and not ulcerated, you add a steroid.

2. HSV is an epithelial-based pathogen, meaning initial attacks have to occur within epithelial cell populations, which open the door for alternate, deeper pathologies. In theory, this means every patient with a deeper form of corneal HSV must have experienced an epithelial episode at one point. In practice, however, this rule is not preserved. I would estimate that around 50% of the stromal and endothelial keratitides we treat as HSV at our clinic have no compelling historic or objective clinical evidence of previous dendritic keratitis. To reconcile the science with the clinical manifestation, you may look for any telltale irregular anterior scar, which could be an indication of a past episode of dendritic keratitis. When scarring is also absent, however, it does not rule out the possibility of stromal HSV as the underlying etiology. It’s widely speculated that a high percentage of HSV patients will have bouts of subclinical infectious keratitis as their initial corneal exposure rather than a dendritic keratitis.

**ANCILLARY TESTING**

Without the need for a preceding dendrite, you are free to keep HSV on the differential list for just about any type of keratitis. However, this does require supporting the diagnosis in other ways. In most cases, the clinical picture itself will tell you what you need to know. When further evidence is warranted, clinicians can use ancillary testing.

As HSV tracks along sensory nerves, patients will frequently develop varying degrees of corneal neuropathy and the involved eye will become relatively less sensitive than the non-involved eye. With no esthesiometer at our clinic, we use dental floss to touch the cornea across quadrants to assess sensitivity and compare the involved eye to the non-involved eye. Although an indirect way to assess HSV risk, in a patient with unusual unilateral keratitis and ipsilateral reduced or absent sensitivity, it’s a strong indicator of a viral source or history.

Another ancillary test is a viral swab and culture. This also is only an indicator of active virus, so it will only be positive with a dendrite (which you probably shouldn’t need a culture to identify) or necrotizing stromal keratitis, which diagnostically could benefit from a culture.

When the diagnosis of HSV is more difficult to confirm, a blood test for IgG or IgM viral antigens, while not diagnostic of corneal disease, can support exposure to the underlying etiology. If you are treating for viral keratitis and the antibodies are both negative, you likely need to reconsider your diagnosis. Again, most of our diagnoses of HSV keratitis are based on clinical features, and it is relatively uncommon that we send out for labs.

I infectious epithelial HSV keratitis, which we see clinically as a dendrite, is a fairly simple clinical diagnosis, and its treatment is the most straightforward of any of the manifestations of HSV keratitis. More interesting is the science behind how these episodes set the stage for myriad other, deeper forms of HSV even though, in many cases, the initial infectious bouts are asymptomatic. This, when paired with the extremely varied manifestations of HSV keratitis, frees us to keep it on the differential for virtually any form of corneal inflammation, which is why for any unilateral keratitis without a compelling history, HSV keratitis will always be near the top of your differential until examination and patient history prove otherwise.

Patients frequently proclaim refractive surgery was “the best decision they ever made.” Modern technologies such as the femtosecond laser and wavefront analyzers have made bladeless procedures and custom ablations possible, typically resulting in better outcomes.

Despite these advances, patient satisfaction may wane over time. Accommodative and lenticular changes may cause refractive error shifts, requiring vision correction. Worse complications—including severe dry eye, corneal scarring and other corneal irregularities—can dramatically impact vision quality. Although patients may hesitate to pursue contact lenses, gas permeable (GP) lenses are often the best choice to meet their visual needs.

RK: ONGOING ISSUES
The National Eye Institute’s 1980 Prospective Evaluation of Radial Keratotomy (PERK) study found that, after one year, 60% of eyes that underwent radial keratotomy (RK) were within +/- 1.00D of emmetropia, with 78% having uncorrected visual acuities (VA) of 20/40 or better.¹ In 1994, the 10-year study results found that, of the 88% of patients who returned, 60% were still within 1.00D of emmetropia, but 43% of eyes had a hyperopic shift of 1.00D or more.²

Patients who underwent RK in the study are currently 40 or older. In addition to significant hyperopic shifts, they are now presbyopic and require full-time vision correction for both distance and near. Some are now ready for cataract surgery.

Radial incisions weaken the overall stability of the cornea, creating diurnal variations in intraocular pressure, which contribute to corneal curvature and VA changes.³

The further incisions extend into the clear cornea, the greater the risk of instability.² In addition, research shows irregular astigmatism is a likely complication around the incisions.⁴ These patients not only are more likely to experience a hyperopic shift, but will also complain of distortion, glare and halos at night and overall poor quality of vision.²,³ Other adverse events such as neovascularization, split incisions, scarring and inclusion cysts may further complicate the situation.

POST-LASIK CHALLENGES
Although laser in situ keratomileusis (LASIK) is the most common elective surgical procedure in the world today, it has complications.⁶ Patients may struggle with severe dry eye, decreased quality of vision and glare or starbursts at night.⁷ One visually devastating complication is post-LASIK ectasia, even though research reports the incidence ranging from only 0.2 to 0.6%.⁸,⁹ GP lenses are commonly used to correct irregular astigmatism induced by ectasia.

CORNEAL TOPOGRAPHY
GP lenses are a good option for post-surgical eyes in need of further vision correction. When fitting such patients with lenses, clinicians can use a corneal topographer to assess corneal shape and help determine appropriate lens design. RK patients have flatter central corneas due to the radial cuts extending toward the central cornea, in addition to a varying degree of mid-peripheral steepening (knee effect) and irregularity. Meanwhile, the non-ectatic myopic LASIK patient will have a defined ablation zone within the central cornea.

Both types of post-surgical eyes can be described as oblate in shape, with a flatter central curvature and steeper mid-periphery. If a flat base curve (BC) is chosen to match the central cornea, it may cause excessive edge lift or corneal molding. A steeper BC may fit the mid-periphery better, but could result in central bubbles or peripheral seal-off.

To properly fit the entire cornea, reverse geometry GP lenses are often the best choice for alignment. For GP intolerant patients or difficult fits, hybrid or scleral lenses can also be used to vault the cornea entirely. Post-hyperopic LASIK patients,
however, have steeper central corneas more characteristic of a prolate shape. While aspheric GPs often provide acceptable fits, keratoconic lens designs can also be used. If ectasia is suspected but not clearly evident in the slit lamp, corneal topography will reveal it. As with keratoconus, corneal steepening typically occurs inferiorly. Centration can be difficult to achieve, so larger diameter GPs, hybrids or scleral lenses that vault the entire cornea are ideal.

**GP FITTING OPTIONS**

GP lenses for the oblate cornea can be fit both diagnostically and empirically. As with any GP lens, the main goal is good centration and sodium fluorescein alignment. In addition to diameter, which typically ranges from 10mm to 11mm, reverse geometry lenses have two curves that control the lens fit. First, the BC aligns the lens with the central cornea. The second curve moving outside of the optic zone is the reverse curve, commonly referred to as the fitting curve. Steepening or flattening this curve can manipulate the sagittal depth to align with the steeper mid-peripheral cornea.

When choosing the initial lens, practitioners should follow the lens’s fitting guide. If the curves must be adjusted, avoid changing both the BC and the reverse curve simultaneously. Once the central and mid-peripheral cornea is aligned, the peripheral edge can be refined.

In the event that a diagnostic set is not available, reverse geometry lenses can be designed empirically using corneal topography and refractive data. Several lens designs have front-surface multifocal capability that can be ordered with add power, pupil size and eye dominancy.

**BEYOND GPs**

Hybrid and scleral lenses are becoming the first-line choice because of their wide variety of indications. Their ability to vault the cornea can mask larger amounts of irregularity for oblate and ectatic corneas alike. Both now have expanded prolate and oblate shape designs. Their larger diameter and on-eye stability can improve comfort for many patients. Research proves scleral lenses to be therapeutic for ocular surface disease, which is concomitant in many post-refractive surgery patients.

**PATIENT EXPECTATIONS**

One of the most important aspects of fitting post-refractive surgery patients is evaluating and managing expectations. Some successful patients have regressed over time, leading to decreased visual acuity and quality. They often desire better vision without spectacles. While their goals may be achieved, especially with newer multifocal options, the fitting process can be challenging. Proper patient education about fitting process and cost is essential.

**Fig. 3. Post-RK axial map.**

**Fig. 4. Post-LASIK ectasia axial map.**

Patients who have struggled with less-than-ideal outcomes from RK or post-LASIK may present frustrated and angry at their current situation; but the significant vision improvements GP lenses can provide often leave them ecstatic. No matter which patient is in your chair, you have an arsenal of lenses to meet their needs.

Seeing the Cornea with OCT Eyes

This diagnostic tool can provide advanced analysis of the anterior segment, corneal pachymetry and more.

The cornea—a structure that is both incredibly simple and fascinatingly complex—is the centerpiece of our slit lamp examinations. Yet, we seem to forget its intricacies because we see it every day. Like the complexity of a computer or car, so long as it is working well, all is well. But sometimes it is important for us to sit back and re-examine the cornerstone of sight, especially as vision scientists. One of our newer technologies, optical coherence tomography (OCT), is the perfect tool to take a closer, and more careful, look at the cornea.

Seen largely as instruments for the posterior segment, most clinicians and industry partners have overlooked its utility in visualizing the anterior segment. We, however, have been using OCT to view and analyze the anterior segment for years.

In this column, we review some technological advancements that are currently available or are in development, as well as postulate where the technology may go in the future. In our minds, OCT can be as much a part of our anterior segment assessment as it is for the posterior segment, and it may possibly become a daily practice mainstay similar to the tonometer or autorefractor.

**EARLY OCT USE**

We first began using OCT instruments to measure sagittal depth of our custom scleral contact lens fits. Early to the scleral lens world in 2004, we traditionally used the estimation method to view how much clearance the lens had from the cornea. Soon, we realized we could use the anterior chamber feature of our OCT to image the lens, tear film and cornea. Although the estimation method was suitable in the past, its variability begged for a better way—leaving room for OCT, which has now become the mainstay when fitting scleral lenses.

A study comparing the estimation method with OCT use found a margin of error ranging from 53µm to 207µm. For the right eyes in particular, the difference between the observer and OCT averaged 128µm (Figure 1).

Initially, the anterior OCT was designed to view the anterior chamber angle in patients with glaucoma. Today, the value of an anterior OCT can range far beyond glaucoma alone.

An early OCT model that gained attention in cataract and glaucoma circles was the Zeiss Visante, which produced widefield cross-section tomography of the anterior chamber. Its major benefit was the capability to produce images of the anterior chamber beyond limbus to limbus. Unfortunately, the system’s limited software capabilities and advancements stifled its adoption by the eye care market (Figure 2).

**EXPANDING VIEWS**

Following the glimpse into the anterior chamber the Zeiss Visante provided, many OCT companies began looking for ways to expand their own anterior views. Current OCT hardware does not allow for wider-angle views of the anterior chamber. As such, all current OCT systems require the use of ancillary lenses to capture these additional views.

While newer hardware has expanded OCT imaging with much wider views, they still do not provide the full limbus-to-limbus or conjunctiva-to-conjunctiva views provided by the original Zeiss Visante. Clinicians can steer the instrument to spread the view beyond this area of the cornea, but the technology isn’t ideal. We predict this will be one of the biggest opportunities for differentiation in future OCT instruments (Figure 3).
Improvement is progression software. Although today’s OCT instruments have reached beyond what our expectations were several years ago, we know they can do more.

MEASURING CORNEAL THICKNESS

Originally, we used specialized calipers to measure corneal thickness on our small-field OCT scans. Now, advanced OCT technologies allow for pachymetry maps of the cornea. The newer Zeiss instruments, for instance, can reach 9mm in diameter, and the maps can show various areas of corneal thickness at one time. As with topography, we can visualize and measure changes in patients with corneal anomalies such as keratoconus.

Corneal topography remains the mainstay for detecting and measuring keratoconus in many practices, largely because it has a greater adoption rate than anterior segment OCT or specialized instruments that measure posterior and anterior curvatures such as Orbscan (Bausch + Lomb) or Pentacam (Oculus). However, topography has its limitations, including variability when imaging the tear film and capturing highly irregular surfaces. As such, we have found topography is limited in its ability to show corneal change for advancing keratoconus patients. OCT, on the other hand, allows us to measure thickness across the central cornea. Future software developments and advanced hardware systems could help OCT become the hallmark for keratoconus and other corneal diagnoses (Figure 4).

MEASURE, CALCULATE AND TREAT

Other exciting advancements in OCT imaging relate to power and curvature calculations and the measurement of epithelial thickness mapping, which can help with the detection and management of corneal anomalies and dry eye (Figure 5).

Some advancements are limited to research settings or are only available outside the United States, but we hope to see them progress into clinical practice soon. We predict OCT advancements with nerve assessment will one day make it a useful tool for other corneal diseases and dry eye issues. For example, software that can map cataract density and location may help determine cataract severity and type, while length measurements will help with intraocular lens calculations and future refractive error calculations from all areas of the cornea, possibly providing a better understanding of refractive error and myopia control.

Lastly, partnering point-of-care diagnostics with dynamic OCT imaging may improve our ability to determine where a patient is in their acute disease state. With the most accurate diagnosis in hand, we can better tailor treatment options and improve patient care.

It is time to bring OCT to the forefront of eye care. It has applications for both posterior and anterior segment assessment—far more than what we might imagine possible. It is important for us to drive our industry partners to push the limits and expand the parameters of our current technological capabilities.

Double Trouble?
Suspected amoebic infection plus fungal superinfection—in a diabetes patient, no less.

This 59-year-old female with a history of diabetes was referred for a non-healing corneal ulcer. At symptom onset five weeks prior, she was seen in the emergency department for a small (1mm) central corneal ulcer and given tetracaine and bacitracin ointment. Two days later, she saw her local optometrist and was switched to Vigamox (Alcon) QID and Nevanac (Alcon) BID; also, a bandage contact lens (BCL) was applied. Two days later, the central ulcer was resolving. The Nevanac was discontinued but Vigamox was continued.

A week later, vision had improved to 20/60 and the epithelium was nearly resolved. The BCL was removed and Pred Forte (Allergan) added to control scarring. After another week, the nasal central corneal ulcer remained. Current meds were discontinued and she was started on fortified vancomycin, tobramycin and ofloxacin and referred for tertiary care.

The clinical image below shows a small, ~1.5mm x 0.5mm paracentral area of subepithelial haze with feathery edges and surrounding corneal haze without stromal thinning. Confocal microscopy demonstrated multiple areas suspicious for trophozoites near Bowman's membrane and several areas of branching elements in the mid to deep stroma.

Concern is for an amoebic keratitis as well as possible branching fungal elements. Recommend treatment for Acanthamoeba keratitis is PHMB and oral ketoconazole. The decision was made to hold off on treating for a superimposed fungal keratitis. A corneal biopsy was performed for further pathogenic characterization that will further guide treatment. Topical antimicrobials will continue until there is clinical improvement on anti-amoebic therapy.

**CALL FOR ENTRIES:** If you have great clinical images of fascinating cases to share, send large, high resolution photos (corneal disease or contact lens wear only) and a brief case description to: photos@jobson.com.
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