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REVIEW OF CORNEA & CONTACT LENSES

SEPTEMBER/OCTOBER 2021

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REFERENCES: 1. Results of an online survey with Eye Care Professionals who completed an evaluation program for Bausch + Lomb ULTRA® Multifocal for Astigmatism contact lenses (n=219). Survey results include Eye Care Professionals who reported that successful fitting was extremely easy, easy, or somewhat easy (on a 6-point scale) during the first visit, with a margin of error ±4.1%. **2.** Results of an online survey with Eye Care Professionals who completed an evaluation program for Bausch + Lomb ULTRA® Multifocal for Astigmatism contact lenses (n=219). Survey results include Eye Care Professionals who strongly agreed, agreed, or slightly agreed (on a 6-point agreement scale) with the surveyed statement, with a margin of error ±2.8%.

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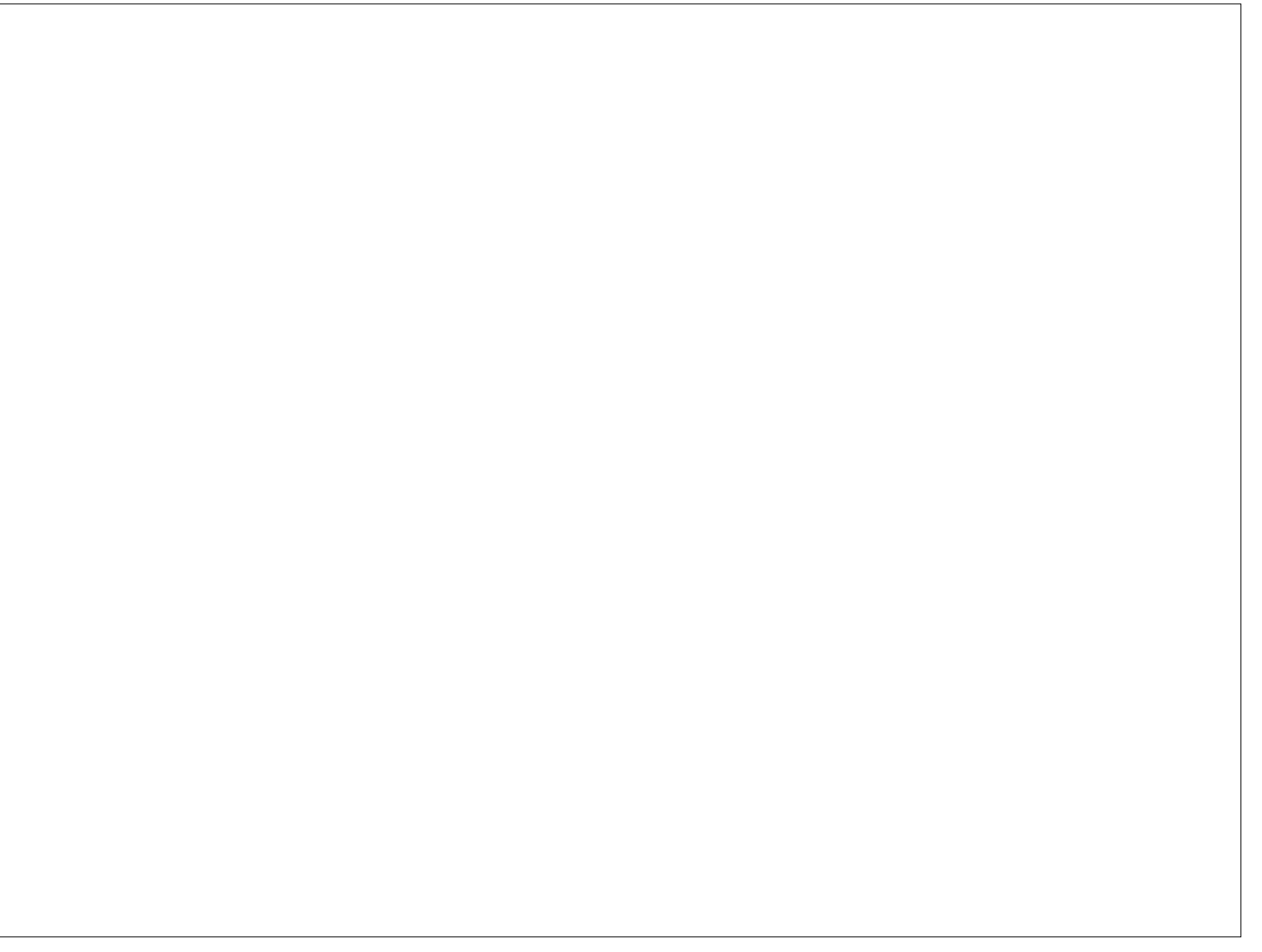

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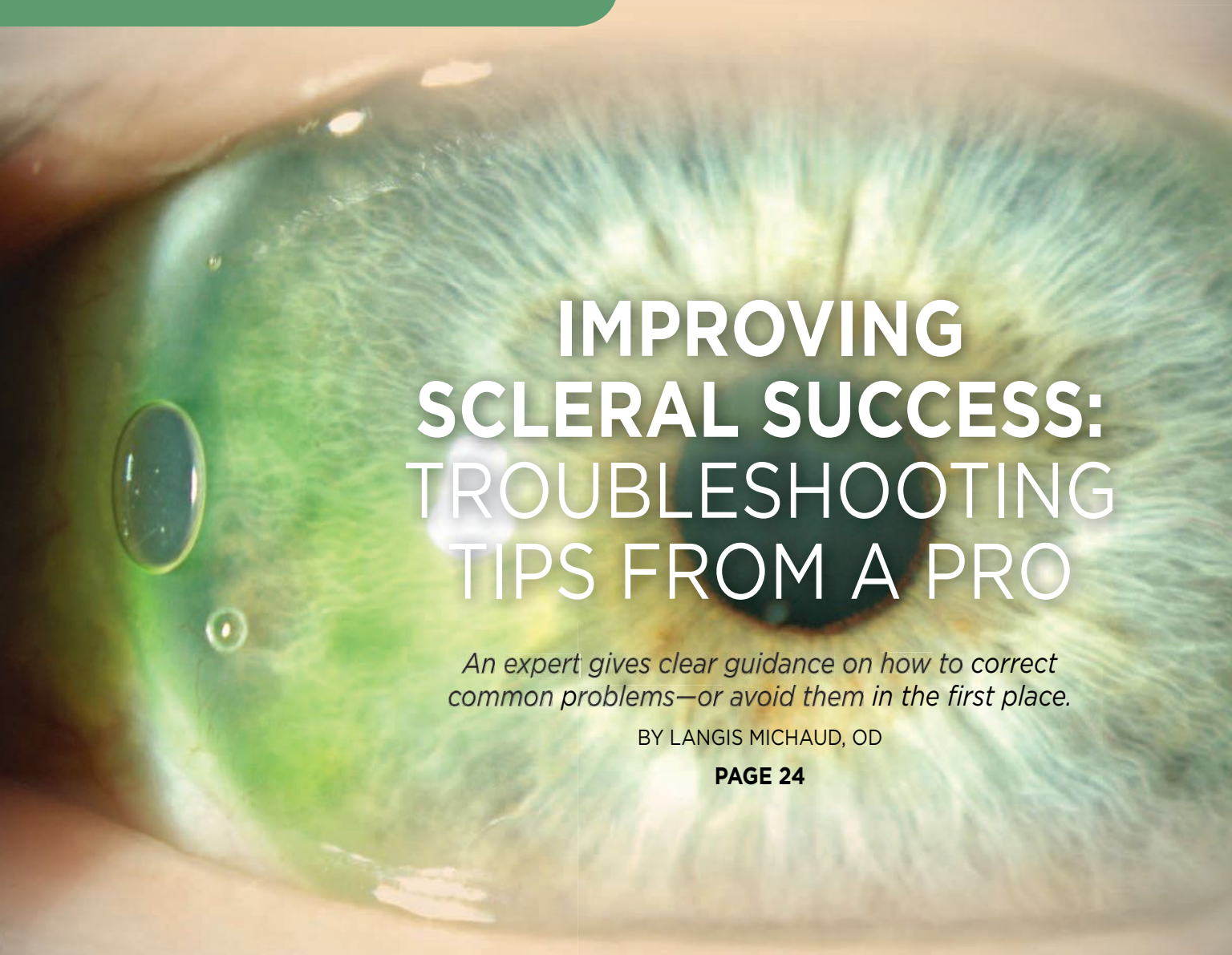

IMPROVING SCLERAL SUCCESS: TROUBLESHOOTING TIPS FROM A PRO

An expert gives clear guidance on how to correct common problems—or avoid them in the first place.

BY LANGIS MICHAUD, OD

PAGE 24

When it comes to myopia control in children who are 8-12 years of age at the initiation of treatment,

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***Indications for use:** MiSight® 1 day (omafilcon A) soft (hydrophilic) contact lenses for daily wear are indicated for the correction of myopic ametropia and for slowing the progression of myopia in children with non-diseased eyes, who at the initiation of treatment are 8-12 years of age and have a refraction of -0.75 to -4.00 diopters (spherical equivalent) with ≤ 0.75 diopters of astigmatism. The lens is to be discarded after each removal.

†Compared to a single vision 1 day lens over a 3 year period.

‡Fitted at 8-12 years of age at initiation of treatment.

References: 1. Chamberlain P, et al. A 3-year randomized clinical trial of MiSight® lenses for myopia control. *Optom Vis Sci.* 2019; 96(8): 556-67.
2. Chamberlain P, Arumugam B, Jones D, et al. Myopia Progression in Children wearing Dual-Focus Contact Lenses: 6-year findings. *Optom Vis Sci.* 2020; 97(E-abstract): 200038.

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IN BRIEF

■ Researchers found that **female sex and younger age may increase the risk of axial elongation in high myopia**. Eyes in females or eyes with prior choroidal neovascularization had a **greater annual rate of axial elongation**, especially when compared with men aged 40 to 70 years. **Women with high myopia were 1.51 times more likely to develop severe axial elongation** than men. Additionally, **patients aged 20 to 30 years were more likely to exhibit axial length growth than patients over 40**, possibly due to prolonged use of computers and mobile phones.

Du R, Xie S, Igarashi-Yookoi T, et al. Continued increase of axial length and its risk factors in adults with high myopia. JAMA Ophthalmol. August 26, 2021. [Epub ahead of print].

■ **Pregnancy may temporarily steepen crosslinked corneas** in some keratoconus patients (41.7% of pregnant women in this study). However, **by the end of the follow-up period (mean of 28 months post-delivery), corneas flattened by nearly the same amount.** Pregnancy did not alter any of the refractive or visual parameters in crosslinked corneas, suggesting that **the temporary change in corneal curvature could be attributed to a hormonal cause.**

Sarac O, Yesilirmak N, Caglayan M, et al. Dynamics of keratoconus progression after prior successful accelerated cross-linking treatment during and after pregnancy. J Cataract Refract Surg. August 24, 2021. [Epub ahead of print].

■ **Contrast sensitivity** (measured using the Pelli-Robson test) **can be useful in assessing visual dysfunction and severity of Fuchs' endothelial corneal dystrophy (FECD)**. This study found a moderate correlation between BCVA and contrast sensitivity, with some patients maintaining good BCVA but exhibiting reduced contrast sensitivity. This highly repeatable, accurate, inexpensive and rapid test could help **identify reduced vision in FECD patients that may be disguised by a good BCVA.**

Okumura N, Padmanaban V, Balaji J, et al. Clinical, morphological and optical correlates of visual function in patients with Fuchs' endothelial corneal dystrophy. Cornea. August 6, 2021. [Epub ahead of print].

Good DSEK Graft Survival Rates 10 Years Out

Several different options are available to treat corneal endothelial dysfunction, and techniques continue to evolve for these procedures. A recent study considered 10-year outcomes following Descemet stripping endothelial keratoplasty (DSEK) and found a high rate of graft survival, at 79%, including patients with complex grafts.

Additionally, the UK-based investigation reported an endothelial cell loss (ECL) of 73% at the 10-year mark, while low-risk Fuchs' endothelial dystrophy grafts showed a 92% survival rate over the same period.

"We propose that long-term prescribing of topical corticosteroids combined with early treatment of graft rejection contribute to reducing ECL and increased graft survival," the authors wrote in their paper.

The study analyzed the results of 356 consecutive DSEK grafts performed by 10 surgeons who used a standard protocol technique. The investigation's primary outcomes were cumulative graft survival and ECL from six months to 10 years, while secondary outcomes included risk factors for graft failure, postoperative complications, visual outcomes and central corneal thickness.

Indications included 209 Fuchs' endothelial dystrophy and 88 bullous keratopathy cases, 39 previous graft failures and 104 eyes with preoperative glaucoma. Cumulative graft survival rates in all eyes were 97% at one year, 90% at three years, 85% at five years and 79% at the 10-year study endpoint.

Considering topical steroid drops, the investigators suggested using long-term alternate-day dexamethasone 0.1% offers an optimum balance between reducing the graft rejection risk and minimizing steroid-associated complications.

The percentage of ECL loss of all grafts was 46.6±17.3 at year one, 54.9±18.7 at year three, 59.6±17.4 at year five and 73.1±9.7 at 10 years. Despite a 10-year endothelial cell count of only 692 cells/mm^2, graft survival remained high with good vision, the authors said.

Also of note: the researchers found that preoperative glaucoma, previous glaucoma surgery and regrafts were significant risk factors for graft failure.

DSEK remains a viable treatment option, especially in complex eyes with comorbidity, and outcomes can be maximized with a standardized protocol, the authors concluded.

Fu L, Hollick E. Long-term outcomes of Descemet stripping endothelial keratoplasty: ten-year graft survival and endothelial cell loss.

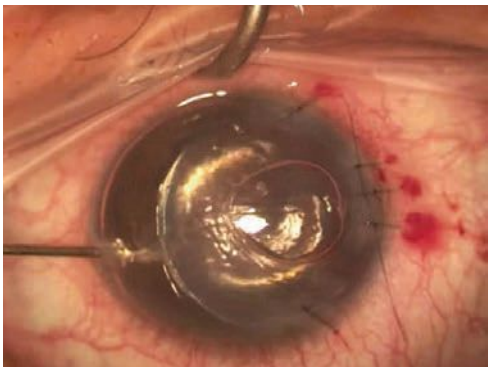

Photo: James Lewis, MD

While DSEK has proven itself to be a quantum leap up from penetrating keratoplasty, endothelial cell loss remains a concern.

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Topical Insulin Heals PEDs, Reduces Amniotic Graft Rates

Corneal defects that fail to improve in the two weeks following injury are known as persistent epithelial defects (PED). Causes of this rare instance of hindered epithelialization include altered epithelial adhesion, limbal stem cell deficiency, trauma, medications and infections. There are several noninvasive treatments for PED, such as autologous serum and topical insulin, the latter having less evidence of its efficacy, until recently. A new study demonstrates that topical insulin may actually be the more effective treatment option for this sight-threatening condition.

The retrospective data analysis included 61 patients treated with topical insulin (case group) and 23 treated with autologous serum (control group) for PED resistant to conventional treatment. The researchers sought to determine the percentage of patients who reached epithelization, as well as when and how quickly they did so.

Epithelization was achieved in 51 patients on insulin but only 11 on autologous serum. Re-epithelization also occurred much faster in patients in the insulin group with a mean time of 33 days (range four to 124), while the autologous serum group had a mean time of 83 days (range 13 to 231) until re-epithelization.

Amniotic membrane transplantation (AMT) is a surgical procedure often mandatory for patients whose eyes don't respond to treatment and face a risk of perforation. This study's results showed the need for AMT was significantly lower in the insulin group. In addition, PED recurrence was also much less common in patients treated with insulin compared with autologous serum (11% vs. 43%).

"A decrease in the need for AMT with topical insulin should be considered as an important step toward epithelization," the researchers wrote in their study. "For this reason, we have adopted insulin treatment as a first choice within second-line treatment options, that is, when epithelization is not achieved in two weeks with standard initial treatment. It would also seem to us an effective and valid first line of treatment option to avoid prolonging the re-epithelialization of PED and the appearance of possible associated complications."

In this dataset, topical insulin presented significantly better epithelization outcomes than autologous serum eye drops. Consider this treatment option for those rare cases of PED you may come across in your practice to help speed up the recovery process and prevent further damage while still achieving optimal visual outcomes.

Diaz-Valle D, Burgos-Blasco B, Rego-Lorca D, et al. Comparison of the efficacy of topical insulin with autologous serum eye drops in persistent epithelial defects of the cornea. Acta Ophthalmologica. August 18, 2021. [Epub ahead of print].

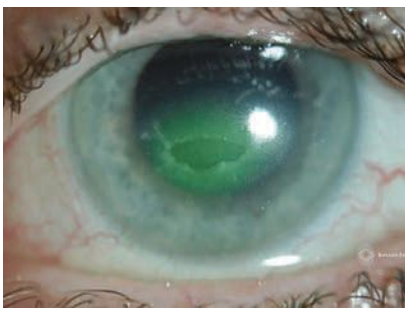

Photo: Alan Kwok, OD

Thanks to insulin-like growth factors, topical insulin can kick-start epithelial cell development from limbal stem cells.

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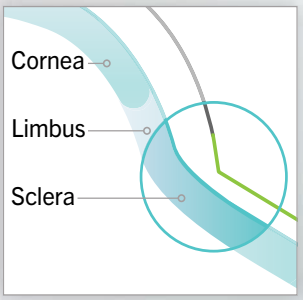
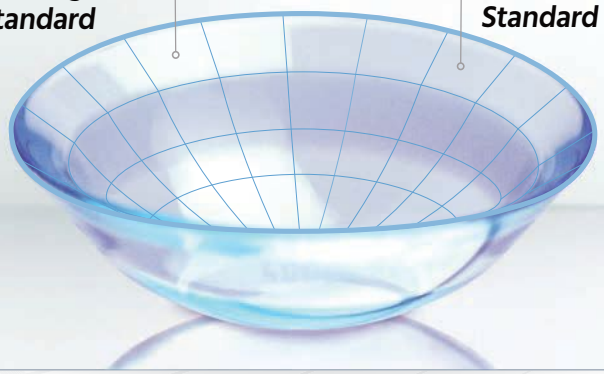


Linear landing zones follow the straight anatomy of the para-limbal sclera

Linear Scleral Landing Zone: ***Standard***

Toric Periphery: ***Standard***



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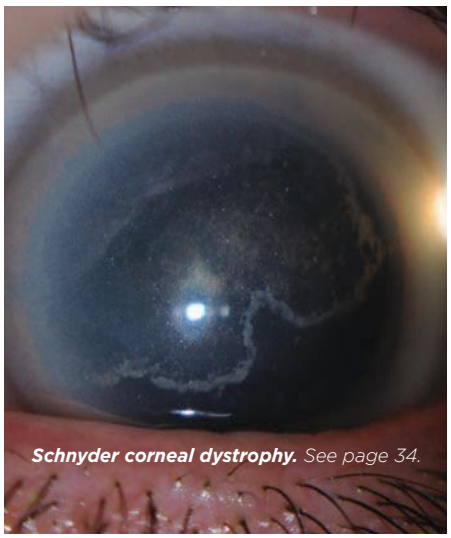

Schnyder corneal dystrophy. *See page 34.*

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By Joseph P. Shovlin, OD

Treating Myopia Today and Tomorrow

As the prevalence of this condition continues to increase, so does the effort to combat it.

The World Health Organization, in its Vision 2020 campaign for the prevention of blindness, declared myopia one of the top 10 eye diseases.[1] With the striking increases in myopia seen globally, modeling efforts with eventual targeted intervention appear to be a major thrust for future efforts.

The American Academy of Ophthalmology last year established a task force on myopia and recently published a summary of actions necessary to reduce the global burden of myopia.[2] They have focused on both delaying the onset of myopia and reducing progression.

These goals are laudable and should receive high priority for future public health initiatives and collaborative research for interventions for myopia control.[2] The task force has chosen to address the pillars of education, advocacy, research and public health.[2]

OPTIONS TODAY

Current treatment options show consistent evidence for being a benefit. They range from pharmaceutical use of a wide range of dosages of atropine to optical approaches (spectacles and contact lenses) that induce peripheral myopic defocus. Overall, practitioners must first weigh the risk/benefit before recommending each treatment option. Future studies should examine: (1) the optimum dosage of atropine, (2) frequency and best time of application, (3) duration of treatment and (4) any long-term consequences of treatment.[2]

There may be an additive effect using both pharmaceuticals and optical corrective devices, but unfortunately there is a lack of long-term follow-up, and how interventions (when used concurrently) interact is uncertain.[3,4] We welcome the addition of new devices such as Essilor's Stellest for myopia management, Cooper Vision's MiSight and Johnson & Johnson's Acuvue Abiliti.

Lifestyle changes may help reduce myopic progression and delay its onset. Interventions include diet, lighting conditions, posture, reading position and distance and time spent on near tasks, to name a few.[2,3]

A well-known and frequently cited modifiable risk factor is the time children spend outdoors. Several studies have shown a protective effect in essentially both delaying onset and reducing progression with increased outdoor exposure. More evidence is needed to quantify the ideal type of exposure outdoors, along with the amount and duration of exposure necessary to reduce progression.[2,3]

FIGHT FOR TOMORROW

Do we need more effective treatment options? Of course we do. However, in the meantime, don't hesitate to add these beneficial options now to your management plan when appropriate. I can appreciate and understand some reluctance, as I have often been slow to employ treatment options.

Atropine has greater efficacy at higher doses but also carries a higher rate of side effects. Reduced concentration atropine use requires formulations made by compounding pharmacies. Dilution results in a reduced concentration of preservative and buffers. But, there's not much tolerance for rare vision-threatening events in children, so careful screening and scrutiny is imperative.

By all measures, myopia is truly an epidemic, and there certainly are unmet needs in delaying and reducing progression. To move forward, we must first identify the major gaps that exist in knowledge. Surely, we'd like to know which kids are at significant risk and at what age, as well as what works best with currently available options and in what order (contacts then pharmaceuticals, or the converse). Also, is there an alternative pathway not yet known to slow progression and/or delay onset past the "magic age" of nine?

Even though myopia treatment presents challenges due to its multifactorial nature, I look forward to advances beyond what's available today. It would be interesting to be like the fictional character Rip Van Winkle, who comes back after sleeping for two decades to a changed world. Would it not be nice to come back to a changed world where myopia has been slowed in its progressive nature, or even cured? For now, take advantage of the viable options available to us today. You might just save someone from significant visual impairment years later. RCCL

1. Prousali E, Haidich AB, Fontalis A, et al. Efficacy and safety of interventions to control myopia progression in children: an overview of systematic reviews and meta-analyses. BMC Ophthalmol. 2019;19(1):106.

2. Modjtahedi BS, Abbott RL, Fong DS, et al. Reducing the global burden of myopia by delaying the onset of myopia and reducing the progression in children: the Academy's Task Force on Myopia. Ophthalmol 2021; 128(6):816-26.

3. Sánchez-González JM, DeHita-Cantalejo C, Baustita-Liamas MJ, et al. The combined effect of low-dose atropine with orthokeratology in pediatric myopia control: review of the current treatment status for myopia. J Clin Med. 2020; 9(8):2371.

4. Jonas JB, Ang M, Cho P, et al. IMI prevention of myopia and its progression. Invest Ophthalmol Vis Sci. 2021;62(5):6.

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1. CooperVision data on file 2021. Rx coverage database; 14 to 70 years. **2.** CVI data on file 2021. Based on number of prescription options available in the USA across all soft lenses reported by the 4 main manufacturers. **3.** Around the clock axis in 10° from Plano to -6.00DS in -0.75DC, 1.25DC and -1.75DC. **4.** CVI data on file 2021. Based on number of prescription options available in the USA across all soft 1-day toric lenses as reported by the 4 main manufacturers. **5.** CooperVision data on file 2020. Kubic masked online survey; n=404 US ODs who prescribe toric soft CLs.

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By Marcus R. Noyes, OD, and John D. Gelles, OD

Scleral Lenses: The Perfect Landing

Here's how to correct problems with conjunctival compression to achieve an ideal fit.

Scleral lenses offer significant visual and ocular health improvements to our patients, especially those with corneal or ocular surface disease. This article will focus on the scleral lens landing zone, specifically problems with tissue compression.

CAUSES AND EFFECTS

Tissue compression is the result of a misaligned scleral lens landing zone or a landing zone that is not wide enough to evenly spread the force the lens exerts on the conjunctiva. Both excessively steep and flat lenses are capable of creating these issues.

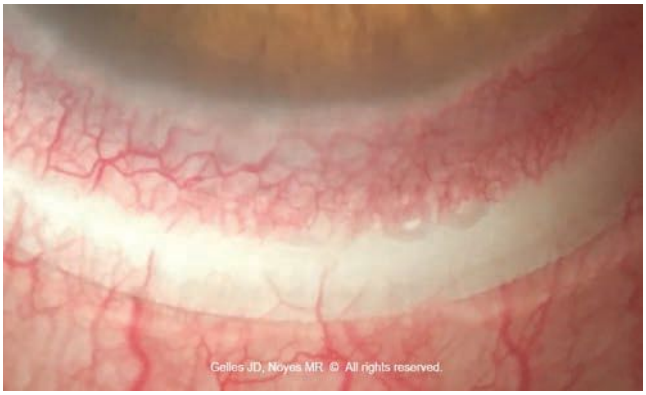
There are a variety of short-term effects of a misaligned scleral lens, including discomfort, arcuate staining, vascular compression (blanching/impingement) and rebound injection after lens removal. If scleral lens misalignment persists over the long term, the previously mentioned effects can be exacerbated and conjunctival hypertrophy development is possible.

IDENTIFICATION

Accurately identifying the region of vessel compression will aid in troubleshooting. Additionally, detailed documentation of this information will help in communicating with laboratory consultants to make lens modifications.

Techniques can be repeated for lens-on and lens-off evaluation, such as with diffuse white light, direct beam and cobalt blue (with and without vital dyes, with and without holding the lids) to observe findings such as pooling, injection, staining, compression and shadows.

First, note the position of the scleral lens markers, typically indicated by dots or dashes on the lens surface, by noting the axis they fall on (horizontal dashes are located at 0° and 180° and dots are located at 270°, for example).



This scleral lens landing zone displays severe (fine and large vessel impingement) conjunctival compression. The left edge of the lens is too flat (note the 0.5mm band of compression located 0.5mm from the lens edge), and the right edge is too steep (note the compression starts at the edge and extends 1mm inward). To improve this landing zone fitting relationship, steepen the left side and flatten the right side by using a toric periphery.

Document the vessel compression in relation to the following:

- lens edge (*e.g.*, "at the lens edge extending 1mm inward" or "2mm in from the lens edge and 1mm wide")
- severity of vessel compression (*e.g.*, "fine vessels are blanched" or "fine and large vessels are fully impinged")
- location of compression by axis (*e.g.*, "compression is located from 0° to 90°")
- periodicity of compression (*e.g.*, "even compression 360° around," "compressed in two areas 180° apart," "compressed in a single area" or "compressed focally over a conjunctival obstacle")
- conjunctival tissue movement (*e.g.*, "the conjunctiva is free moving under the landing zone edge" or "the conjunctiva does not move at the landing zone edge with digital movement through the eyelid").

Using direct white light illumination at a 45° angle to evaluate across the lens (beam on the nasal side to evaluate the temporal landing zone), note the appearance of the lens edge (*e.g.*, "a shadow is cast from the lens edge indicating the edge is lifted" or "no shadow is cast from the lens edge and conjunctival heaping is present indicating the edge is too steep").

Application of vital dye, either sodium fluorescein or lissamine green, will also aid in identification. With the lens on, note whether there are any locations of dye pooling vs. freely flowing under the landing zone edge and if there are adjacent signs of conjunctival folding/staining/heaping. After lens removal, note any rebound injection (both severity and pattern) and if a conjunctival impression ring is present (if so, indicate its severity, depth, width, location and periodicity). Repeat the use of vital


dye to assess conjunctival staining. All findings should corroborate the issues with the initial lens fit.

PROBLEM-SOLVING

If the scleral lens fitting relationship shows compressed vessels at the lens edge, the landing zone is too steep. This can be resolved by flattening the edge (*i.e.*, reducing the angle at which the landing zone contacts the globe). If the compressed vessels are not at the lens edge but rather located inward from the edge, the landing zone is too flat and can be fixed by steepening the edge (*i.e.*, increasing the angle of the landing zone).

If the area of compression is relatively even 360° around the landing zone, flattening or steepening the entire landing zone will help. If the compression has a pattern, such as equal compression at 0° and 180°, use a toric haptic. If the compression is unequal or located at a single quadrant, then a quadrant-specific design can be used.

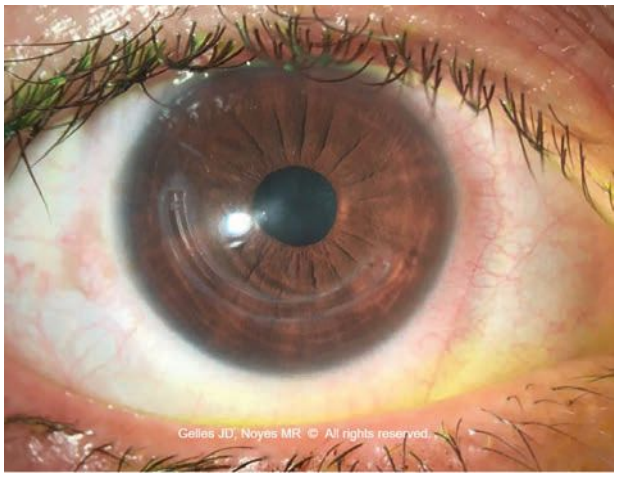
If there is focal compression due to pathology, such as pressing against a pinguecula, incorporating a peripheral edge elevation can help vault this pathology. In mild cases, a simple increase or decrease in lens diameter can overlay a very shallow pinguecula or avoid contact altogether. In some cases, notching a lens (focal removal of lens material) can be effective; however, this can be accompanied by adjacent tissue desiccation. Of course, the use of topography-guided or impression-based scleral lens designs may also aid in these situations. If there is deep circumferential conjunctival compression with rebound hyperemia on lens removal, consider increasing the landing zone width to distribute the lens force more evenly.

Additionally, note where the deepest point is located (inward from where the lens edge indicates an excessively flat landing zone and directly at the lens edge indicates a steep landing zone).

If the lens fitting relationship is left too steep for a longer period of time, noted by development of conjunctival hypertrophy, it is wise to prescribe a topical steroid and discontinue lens wear until improvement is observed. Once improved, refit the patient paying special attention to correcting lens alignment in the affected area. Keep the following in mind: a large-diameter lens to overlay the affected tissue or a small-diameter lens to avoid it.

Tips & Takeaways

- Compression at the lens edge = flatten
- Compression inside the lens edge = steepen
- Symmetric meridian of compression = add toric haptic
- Asymmetric meridian or quadrant of compression = use a quadrant-specific design
- Focal compression = elevate peripheral edge, decrease diameter or notch



Note the deep conjunctival impression on the nasal side of the scleral lens post-removal, with corresponding grade two nasal conjunctival rebound hyperemia. On the temporal side, the conjunctival impression and rebound injection signs are less impressive and the hyperemia is more focal. To improve the landing zone alignment, incorporate a quadrant-specific landing zone flattening the nasal quadrant more than the temporal, increase the landing zone width to spread the lens pressure more evenly and consider adding a focal peripheral elevation to the temporal side at the location of the hyperemia.

CONCLUSION

Keen observation, documentation and communication are key to successful scleral lens landing zone troubleshooting for conjunctival compression. Remember, laboratory consultants are not wizards; they can't magically make you a perfect lens based on poor descriptions. Use these tips for the best results all around. RCCL

Coming up at the CLMA

The Contact Lens Manufacturer's Association has a new president—one who aims to transform the custom lens industry.

This month I caught up with Josh Adams, vice president of Valley Contax, a custom specialty contact lens manufacturer in Springfield, OR. Adams currently serves as president of the Contact Lens Manufacturer's Association (CLMA), a trade organization for the gas permeable contact lens industry. Having just started a two-year term, Adams shares his experience in the industry and the lofty goals he holds for his presidential term and beyond.

Lindsay Sicks (LS): Valley Contax is a family-run company—congratulations on your 40th anniversary this year, by the way—but how did you personally get involved with the CLMA?

Josh Adams (JA): Well, thank you. We are very excited to celebrate our anniversary and reflect not only on how far the contact lens industry has come since 1981, but also look toward the future. I have been a part of this industry since I was 16 years old. My mother, Janice Adams, was president of the CLMA from 2006 to 2008. Following her tenure, I became the official CLMA representative for Valley Contax.

In 2011, I was inspired to join the CLMA board of directors—it was a time when the organization was in search of some fresh ideas. For two years, I served as the youngest board member and really tried to represent ideas for the next generation of contact lenses. I stepped away in 2013, but later realized that decision was probably a result of both naivete and impatience. A good friend helped me realize that it's much easier to effect change from within an organization than from the outside.

Josh Adams, president of the Contact Lens Manufacturer's Association (CLMA).

In 2015, the industry was widely affected by the Valeant Pharmaceuticals acquisition of Bausch + Lomb, Alden Optical and later Paragon Vision Sciences, which continued to motivate me and began to solidify my commitment to rejoin the board of directors. I have since moved up within the executive board over the years before becoming president. Back then, I knew the next decade in our industry was going to hold great promise in terms of innovation, but also include significant obstacles due to the increased interest in custom lenses shown by the big players in contact lenses.

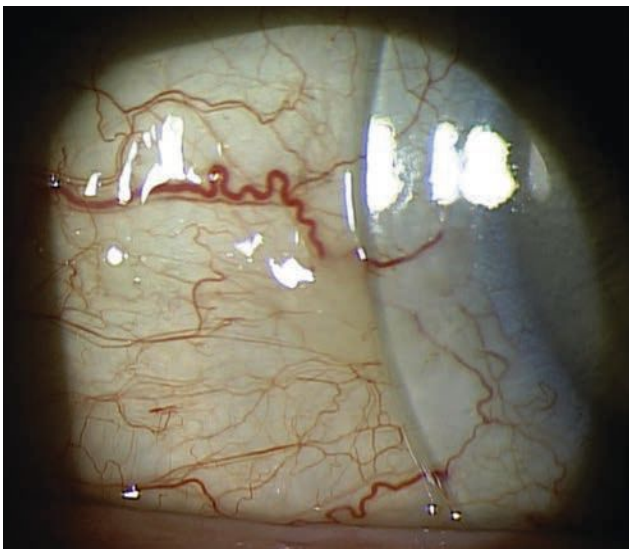
LS: What role do the individual manufacturers and laboratories play in the CLMA?

JA: The mission of the CLMA is to increase the awareness and uptake of custom-manufactured contact lenses. The individual manufacturers and laboratories are the lifeblood of the CLMA. We are all familiar with the nature of the custom lens industry—it's about quality, service and relationships. The CLMA is continuously looking to improve upon the success of our members, as that ultimately drives improved patient outcomes in the clinic. Our member laboratories play a key role in making all of that happen. We are a grassroots organization that still looks out for the independent laboratory. As long as we keep their best interests in mind, mutually rewarding relationships between individual practitioners and their manufacturing labs can thrive.

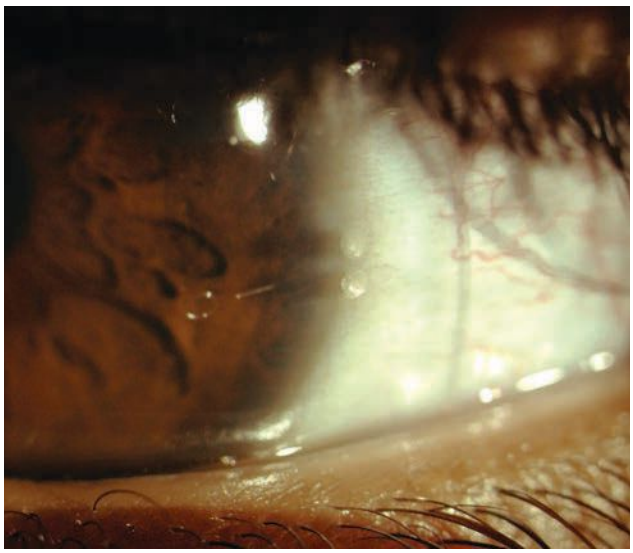
LS: Can you explain the role of the Gas Permeable Lens Institute (GPLI) in the CLMA?

JA: The GPLI has always been known as the educational arm of the CLMA. It was founded in 1985 by a small, independent group of active, contact lens–minded practitioners and forward-thinking industry leaders. The GPLI is so important to our members, as well as individual practitioners. It drives success at the clinical level due to the immense amount of instruction and education, which ultimately leads to positive outcomes for patients. The CLMA provides primary funding support for the GPLI and is very active in its strategic planning and future endeavors.

The GPLI is the primary budget line item for the CLMA, so we are very proud of its impact on the custom contact lens industry. At the end of the day, the GPLI's motto is "GP lenses change lives," and that's what we seek to help practitioners do.



Custom Stable Elite (Valley Contax) 16.8mm scleral lens fit on a patient at a GPLI Workshop, showing compression over a pinguecula.



Lens markings indicate the quadrant-specific nature of this Custom Stable Elite, fit as part of Valley Contax's Custom Stable Vision Project materials grant program.

LS: What are your priorities as CLMA president for the next two years?

JA: My primary goal is to unify the efforts of the custom contact lens industry by incorporating all US custom manufacturers into our organization. Over the past five years or so, the industry has seen many changes. The growth and excitement that exists in the custom contact lens space has also brought investment from more traditional players. While this is surely an exciting development, it also brings changes and challenges. My vision is to unite our industry and focus on technology and growth, as well as the clinical education that is key to practitioner and patient success.

LS: If you could do anything to transform the CLMA, what would it be?

JA: I will look to reinvent the CLMA to be prepared for future generations. We are a 60-year-old trade organization that has survived and thrived through so much change.

For the CLMA to continue to stay relevant in the coming decades, we must search for ways to support education and inspiration for all those involved in custom contact lenses. I am currently bringing ideas to the board of directors that can facilitate this transformation. We are prepared to look at all aspects of our operations to determine the best path forward.

Right now, we do a lot of good and impact many lives, but we have not yet reached our potential as an organization and have a lot more to provide for our laboratory members and the practitioners with whom they partner.

LS: What are the greatest lessons you have learned from your time with the CLMA?

JA: Patience and perseverance. With time and experience, I have learned that not everyone sees the same paths all the time—that can take some time to work through. Growing up in this industry and having progressed through the leadership ranks, I have learned that making a powerful impact through change takes time. The best way that I can support my fellow manufacturers is to listen, learn and figure out how we can shape this industry in the ways that benefit all of us.

Support your custom contact lens labs whenever possible! A list of all CLMA members can be found at www.clma.net/docs/2021-clma-member-directory.pdf. For more on the GPLI, visit www.gpli.info. RCCL

SCLERAL LENS FITTING ESSENTIALS

Keep these clinical pearls in mind when working through this process.

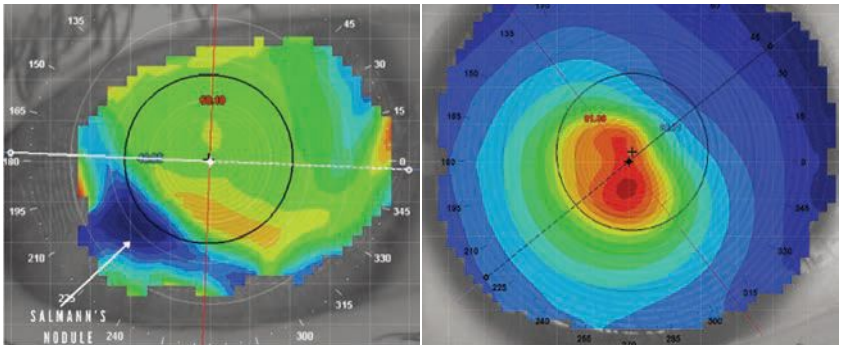
By Manveen Bedi, OD

Scleral lenses are large-diameter gas permeables that offer a wide range of parameters for patients with corneal irregularity from ectasia, post-surgical misadventure and trauma and ocular surface disease. The process of fitting scleral lenses may seem daunting at first; however, if a stepwise approach is followed, excellent success can be achieved with initial fits or refits. Here are some important considerations to keep in mind for achieving a successful fit and maximizing patient comfort and experience.

UNDERSTANDING CORNEAL TOPOGRAPHY

Scleral lenses are fit based on sagittal height, not base curve. First, locate the steepest point of the cornea, as the goal of the fitting is to vault over the cornea without causing corneal bearing. Data from topography can help determine if the corneal shape is prolate, oblate or asymmetric.

For patients with an ectatic corneal pathology, starting with a prolate lens can help achieve clearance over the central cornea. However, for patients with post-surgical eyes, pellucid marginal degeneration or peripheral elevations such as Salzmann's, using an oblate design from the fitting set will help achieve sufficient midperipheral clearance.



This patient has a midperipheral Salzmann's nodule, as indicated by the white arrow (left). Central corneal steepening is a classic sign of keratoconus (right).

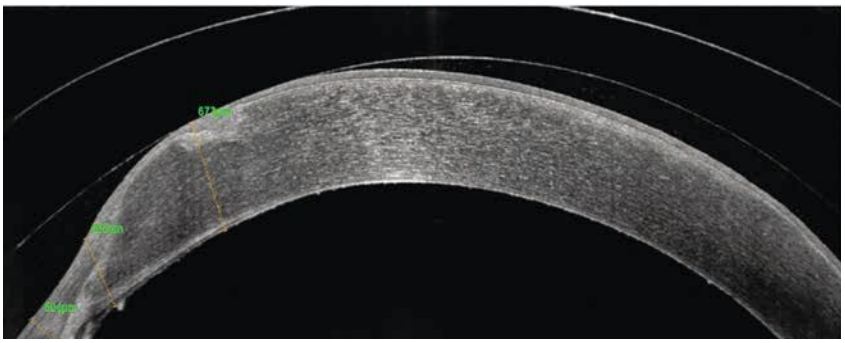
Once the appropriate trial lens design is selected and applied on the patient's eye, wait 20 to 30 minutes before assessing the lens fit. While the lens may look like a good fit upon initial application, the fit might change as the lens sinks into the conjunctival tissue during the settling process and reduces the clearance, the ideal amount of which is 150µm to 175µm after lens settling. Allowing for lens settling before evaluation provides useful information to guide further lens modification.

Also, after the first lens dispense visit, having patients return with at least four hours of wear on the day of the follow-up helps with lens evaluation, decreases chair time and reduces the number of modifications.[1]

It is important to closely monitor patients with significant ocular disease, especially endothelial disease. If the endothelial count goes below 800cells/mm^2, there is a greater risk of corneal edema secondary to endothelial dysfunction. Having a patient wear their lenses for four to five hours before evaluating for any sign of rainbow or glare during lens wear can provide a good indication of candidacy for scleral lenses. Choosing a hyper-Dk material, maintaining a reduced vault and

ABOUT THE AUTHOR

Dr. Bedi completed a residency in cornea and contact lenses at the Southern California College of Optometry at Marshall B. Ketchum University after graduating from the Illinois College of Optometry. She focused on specialty contact lens fitting for corneal pathologies, aphakia and prosthetics. She is a fellow of both the Scleral Lens Society and the American Academy of Optometry.



A highly prolate penetrating keratoplasty graft with nasal elevation in a small-diameter scleral lens.

reducing lens center thickness to maximize oxygen flux to the corneal tissue can aid in this process.[2]

SELECTING DIAMETER

Diameter is a key parameter in scleral lens selection. The majority of fitting sets come with standard diameters; however, making modifications to the diameter can only help maximize the range of parameters for your patient-specific scleral anatomy. Generally, starting with a lens diameter 5mm to 6mm larger than the horizontal visible iris diameter allows the lens to rest 2mm to 3mm past the limbus and offers smoother bearing.

The scleral lens landing area varies with the diameter and, with appropriate selection, it can help alleviate concerns of scleral lens compression, aid in achieving sufficient vault over asymmetric corneal surfaces, rehabilitate ocular surface disease and provide clearance from conjunctival lumps and bumps. For example, when working with patients with highly ectatic or asymmetric grafts in the midperiphery, it can be a challenge to clear the peripheral corneal elevations. Increasing vault in a small-diameter lens can help; however, this often adds more pressure on the haptic with issues of scleral compression. In such cases, a large-diameter lens can help achieve a sufficient vault to clear the peripheral cornea and allow for an even weight distribution at the landing zone.

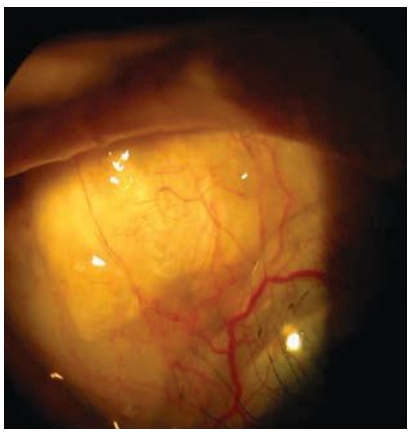
Conjunctival lumps and bumps, such as blebs, pingueculae and pterygia, can also pose a challenge to diameter selection. During the fitting process, if the lens edge abuts the pinguecula or bleb, reducing the diameter of the lens can help reduce irritation, hypertrophy and hyperemia. Notching and/or peripheral vaults can also help avoid constant friction over these areas of conjunctival elevation. However, if you are considering adding a notch or a microvault at the periphery, first ensure lens rotational stability and haptic alignment before ordering modifications. If the lens haptics are not aligned and the lens is unstable, then the notch or microvault may not land at the intended location.

MANAGING SCLERAL TORICITY

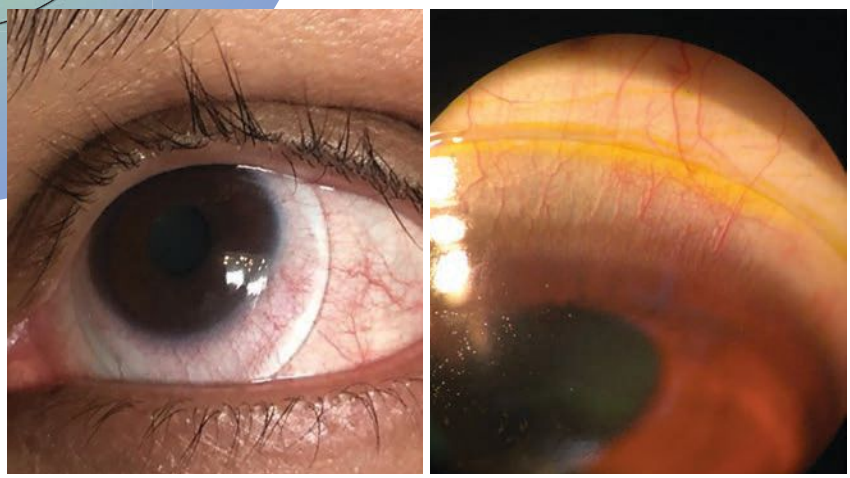
With advances in corneoscleral mapping, there is strong evidence that suggests the sclera is non-rotationally symmetrical, with increasing asymmetry past the corneal apex. The nasal area of the sclera is the flattest due to the proximal insertion of the medial rectus muscle in comparison with the lateral rectus muscle at the temporal portion. A toric or quadrant-specific haptic can help achieve a better scleral alignment, as it provides an even bearing across the scleral surface. Some of the common problems noted with a poor haptic alignment are blanching or fogging in the tear reservoir.[1,3,4]

Blanching occurs when there is localized pressure from the scleral landing zone that reduces conjunctival blood flow. Adding toricity to the lens haptic can help with even weight distribution at the landing zone and reduce compression of the conjunctival blood vessels. Blanching can occur at the lens edge and requires flattening the scleral landing zone. However, if compression takes place under the scleral lens landing zone and closer to the limbus, it indicates a heel effect in which the scleral lens lands too harshly on the conjunctiva, resulting in tissue irritation and hyperemia. In such cases, steepening the landing zone relieves compression and creates a smoother landing.

In addition, midday fogging or air bubbles trapped under the lens can arise from scleral edge lift off. Applying fluorescein after lens application can help assess lens edge lift off. If fluorescein seepage occurs in one of the meridians of the haptic after dye instillation, the lens is most likely flexing with each blink and pumping debris in the tear reservoir, resulting in midday fogging or bubble formation. Steepening the lens edge for better scleral alignment can help reduce patient symptoms



A notch in the scleral lens periphery helps avoid touch with the superior nasal bleb.



Blanching at the lens edge (left) and fluorescein seepage superiorly in a scleral lens due to edge lift off (right).

and prevent aggravated papillary reaction.

MEASURING OVER-REFRACTION

Once a trial lens with an adequate central clearance and haptic has been selected, the next step is to perform an over-refraction to improve the patient's visual outcome. Starting with an autorefraction over the lens can provide a good starting point for manifest over-refraction. A good spherocylindrical over-refraction is often useful to assess the best visual potential for the patient and rule out concerns with flexure and other causes of residual astigmatism.

There are two causes of residual astigmatism with rigid gas permeable lenses: lenticular cylinder and flexure. One way to identify the root cause is by performing topography over the scleral lens. If astigmatism is detected over the lens, flexure is the culprit. However, in a well-fitted lens, if there is no astigmatism noted on topography over the lens, then the cause of residual astigmatism is true lenticular cylinder. In this case, front surface toricity can help correct the refractive issue. In cases where the lens has no haptic toricity and cylindrical power is needed to improve visual outcomes, using a double slab-off ballasting design can help stabilize the front surface toric lens on the eye surface.

Routinely, we can correct for lower-order aberrations such as spherocylindrical refractive error, and defocus can be corrected with traditional glasses and contact lenses. However, patients with keratoconus have significant amounts of higher-order aberrations such as coma and trefoil. Scleral rigid gas permeable lenses provide a stable surface to correct for irregularities on the front surface of the cornea. Another recent technological advancement is wavefront-guided scleral lens correction that allows for superior control of aberrations.

MINIMIZING FLEXURE

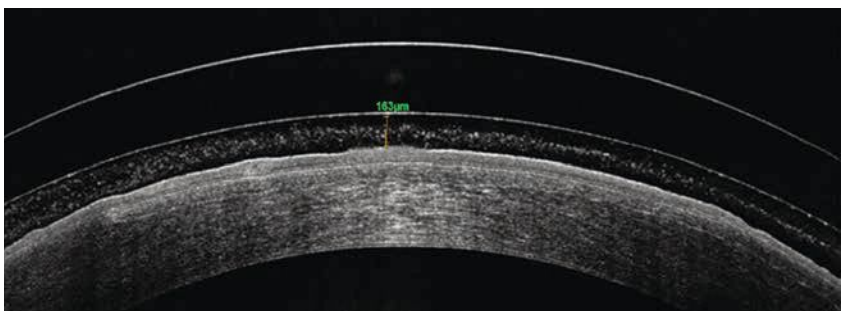
Flexure not only happens with corneal gas permeable lenses but also with sclerals. It can reduce visual acuity and result in ghosting of images. As discussed above, performing keratometry or topography over the scleral lens can help confirm flexure.

In situations of flexure, start by assessing the peripheral landing zone alignment. If the scleral lens landing zone does not correlate to the scleral toricity, the lens will bend, potentially resulting in residual astigmatism. However, if residual astigmatism is still present after a good scleral peripheral alignment and lens centration are achieved, the next step is to increase the center thickness of the lens with caution. Altering lens thickness helps; however, it also reduces oxygen transmissibility to the cornea, which must be taken into consideration.[5,6]

Lastly, front toricity is another option. However, it is important to ensure that the lens is rotationally stable before adding front surface toricity. If the lens is rotationally unstable, the additional refractive power may not land at the appropriate meridian and may exacerbate the ghosting. Once the haptic is finalized and the stability of the lens is established, front toricity can be added to the lens.[5,6]

CUSTOMIZING WETTING ANGLE AND LENS COATING

One of the issues that can be challenging during a new lens fitting or a refit is poor wettability of the front surface of the lens, especially in patients with ocular surface disease. It causes anterior lens surface fogging



Fogging in the tear reservoir due to improper scleral lens haptic alignment.

and deposits and can reduce the quality of vision and visual performance for patients.

Wetting angle is an important factor in the selection of scleral lens material. This refers to the ability of a fluid to spread across the lens surface. Normally, a low wetting angle helps with an even distribution of fluid over the lens surface. Managing anterior surface fogging requires a multistep approach of ocular surface treatment and appropriate lens material and lens coating selection. Material with sufficient Dk and a low wetting angle can help in improving anterior surface wetting concerns and lens optics. However, in cases where high Dk lenses, such as Boston XO and Boston XO2, are required for compromised corneas, adding hydrophilic coating such as Hydra-PEG (Tangible Science) can alleviate fogging concerns.[7,8]

TAKEAWAYS

As challenging of a task as scleral lens trial fitting (let alone successful fitting) seems, we must start somewhere. There are plenty of considerations to keep in mind as you work through the process; luckily, there are also many helpful resources at your disposal, including the suggestions provided in this article. RCCL

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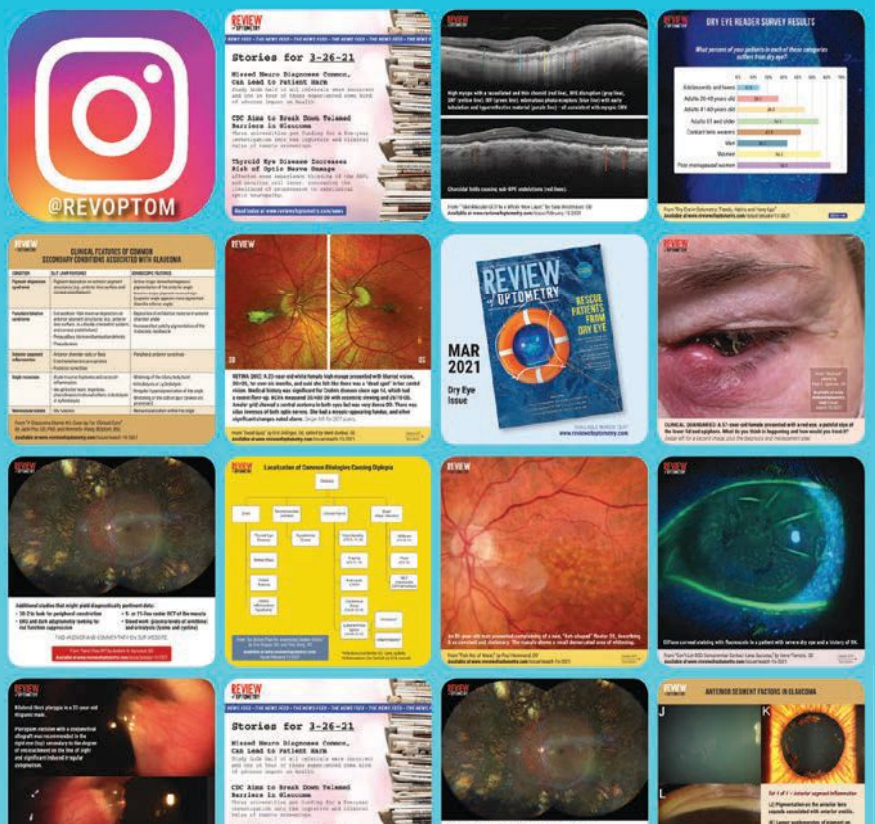
A NEW WAY TO EXPERIENCE *REVIEW OF OPTOMETRY*

Follow us on Instagram at @revoptom for striking clinical images, news headlines, issue previews and great content from the magazine—all formatted for mobile.



SCLERAL TOPOGRAPHY: MEASURING AND MATCHING ITS SHAPE

Taking advantage of this technology can improve your lens fits.

By Marcus Noyes, OD

While it's no secret that scleral contact lenses can provide massive visual benefits to patients, fitting these lenses can have a steep learning curve. With the emergence of new technologies, such as scleral topographers, the initial challenges are much easier to overcome, particularly for practitioners new to scleral lens fitting. Even for experienced fitters, the use of these topography systems can provide many benefits. This article aims to discuss the currently available scleral topographers and how to best use them to maximize fitting success by boosting clinical acumen, improving contact lens fits and generating revenue.

EMPIRICAL FITTING

Scleral lenses can vastly improve visual acuity in a variety of their indicated uses, particularly in conditions like corneal ectasia.[1] Before the fit, it can be easy to promise your patients a marked improvement in both vision and ocular comfort, only to have the patient discouraged by going through two, three, sometimes even six or more trial lenses while looking for the optimal fit and/or an improvement in vision. This issue is further compounded as recent studies have shown that a majority of scleras have asymmetric topography, completely independent of their corneal topographic counterparts.[2,3] That news can be discouraging to the practitioner as well, as dealing with this can take up valuable chair time. Scleral topographers can expedite the fitting process by providing the ability to fit empirically—similar to how many of us already fit corneal gas permeable (GP) lenses.

The first advantage to empirically guided scleral lens fits addresses the scenario previously outlined: you only need one diagnostic trial lens. Scleral topographers can map out the scleral surface and provide the patient with a custom-designed lens, unique to that particular eye. This effectively eliminates the need for the trial-and-error process of finding the trial lens that best fits the patient's eye. Once the scleral surface is mapped, a best-fit lens is generated and available to order. The only trial lens necessary is simply used to calculate over-refraction, trial lens fit be damned.

The second advantage to empirically guided scleral fits is chair time. Most practitioners have their ancillary testing instruments separate from the exam rooms, which means three things. First, the patient is no longer in the exam room as you test the many different trial lenses previously mentioned, meaning it frees up the time that would have been spent in the room with the patient doing trial-and-error. Next, the entire fitting process can be done before the patient enters the exam room. Both of these chair time–saving consequences not only save the practitioner's time but also the patient's.

Lastly, you no longer have to have the patient return to clinic for

ABOUT THE AUTHORS

Dr. Noyes is a clinical assistant professor of ophthalmology and visual sciences at the University of Iowa Carver College of Medicine. He is a Fellow of the American Academy of Optometry and of the Scleral Lens Society.

multiple repeat visits as you reorder and reorder lenses—the fit is nailed down within the first one or two. This not only saves chair time on the day of initial lens fitting but also lessens the demand for lens fitting follow-ups, which frees up future chair time.

PATHOLOGIC ELEVATION

Pingueculae, irregular corneal transplant graft tissues and other pathologic ocular surface elevations like Salzmann's nodular degeneration can all make scleral lens fits quite difficult. In some conditions, such as filtering blebs or tube shunts in glaucoma patients, traditional scleral lens fit can even be contraindicated. With the traditional best-fit trial lens approach, fitting over these ocular surface elevations often requires heavy lens modifications such as notching, peripheral lens elevations, quadrant-specific designs or truncations.

All these processes can require many trial lenses, frequent consultation with the manufacturing lab and sometimes even in-office lens modifications (*e.g.*, getting out a dremel and modifying the lenses yourself). Even if an adequate lens fit is achieved after all these steps, it can prove to be a frustrating and time-consuming process.

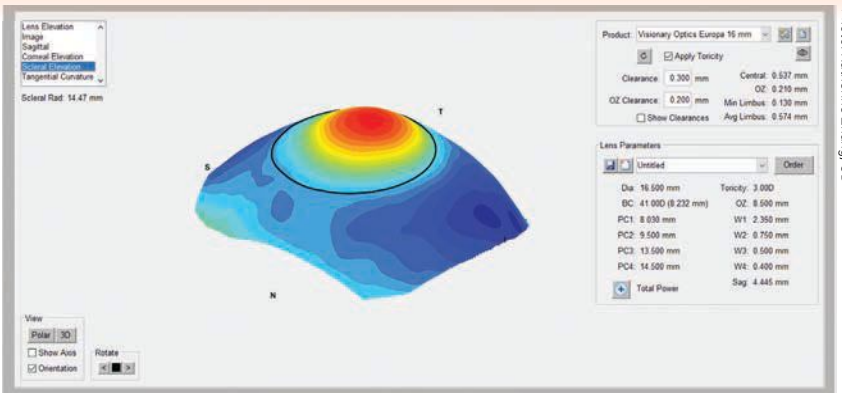


Photo: Katherine Zhang, OD

The sMap 3D scleral elevation map after image-stitching.

Perhaps one of the biggest selling points of scleral topographers is their ability to create freeform lenses. Such lenses are not spherical and are often asymmetric with multiple meridians of toricity or highly irregular surfaces. The first big advantage to freeform lenses is that they can vault these pathologic elevations, without sacrificing other lens parameters such as diameter or base curve. The second advantage: practitioners oftentimes no longer need to rely on complicated lens modifications such as notching or truncations because of freeform lenses' ability to vault these lesions.

CURRENT TOPOGRAPHERS

There are three currently available scleral topographers that can provide scleral profilometry data. Let's discuss how each device could benefit an optometrist and their practice.

sMap 3D (Visionary Optics). This device maps over one million data points on the ocular surface, covering an approximate 22mm range of the sclera, with a precision of 10µm.[4] It is integrated with Visionary Optics's Europa and Latitude lens designs. Europa is a standard scleral lens capable of many different modifications, such as toricity, quadrant-specific modifications, truncation and more. Latitude is the company's freeform lens design that allows for a more exact matching of the ocular surface.

This instrument makes use of sodium fluorescein (NaFl) for its image acquisition. So, from a protocol standpoint, it would go something like this: have the patient sit in front of the sMap 3D device, instill NaFl, perform image acquisition in three gazes (straight ahead, up-gaze and down-gaze). These images are then "stitched" together to one complete ocular surface profile. Lens parameters will be displayed, and you can adjust to your liking or continue with the recommended parameters. Whether that be a best-fit Europa lens or the free-form Latitude lens, the produce is available for order directly from the device.

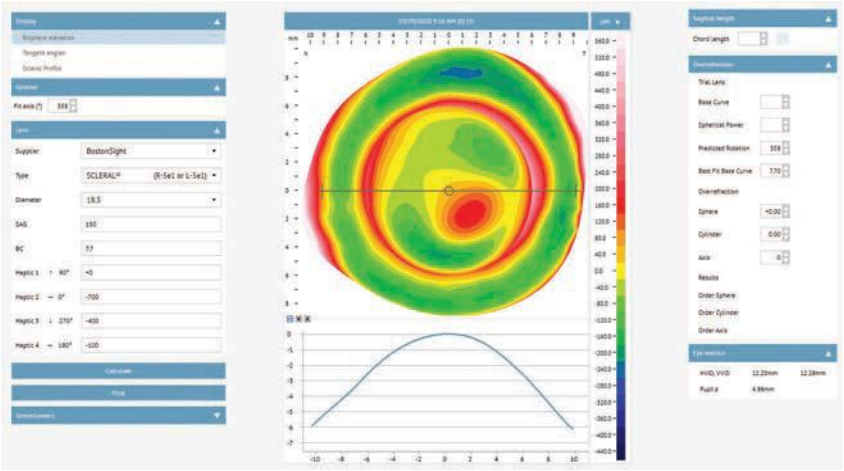
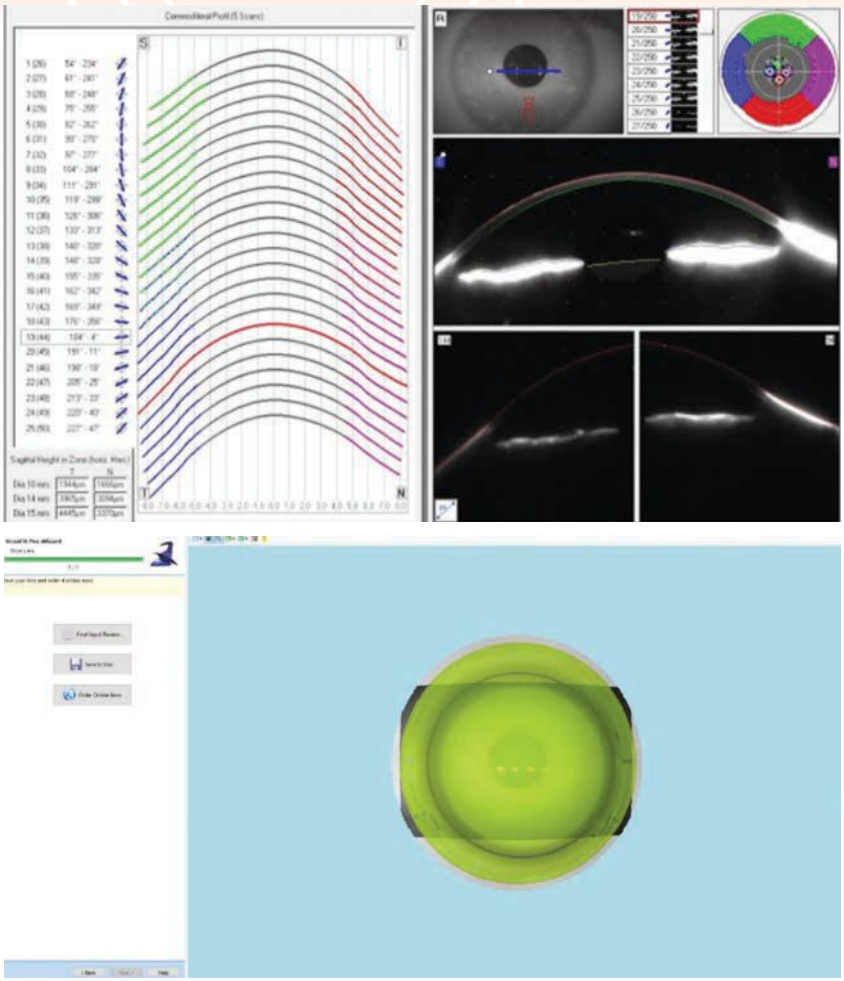


Photo: Michael Everson, OD

Eaglet-Eye ESP software image of a patient diagnosed with corneal ectasia.



Scheimpflug imaging of the corneoscleral surface (top). The same data translated through ScanFit Pro software (bottom).

Eye Surface Profiler (Eaglet-Eye). This instrument is a scleral topographer from Eaglet-Eye. It maps over 350,000 data points, covering a 20mm diameter of the sclera, with an accuracy of 10µm over the sclera (and 2µm to 3µm over the cornea).[5] Eaglet-Eye does not manufacture scleral lenses in-house, and instead has partnered with 19 scleral lens labs, some of which are listed here: ZenLens (all sclerals), BostonSight (BostonSight Scleral), Blanchard (all Onefit designs), EyePrint Prosthetics (ScanFit Pro), Acculens (Maxim), X-Cel (Atlantis), Advanced Vision Technologies (Naturalens Scleral), ValleyContax (Custom Stable) and SynergEyes (VS Scleral and Ultrahealth Hybrids). A complete list is available on their website.

In addition to scleral lenses, this device can also be used for GP, hybrid, orthokeratology and even custom soft lens designs (with a compatible lens manufacturing lab).

The Eye Surface Profiler also uses NaFl for image acquisition, as it is usually applied to both superior and inferior sclera. From there, the eyelids are opened and a single image is acquired while the patient is in primary gaze. Once the corneoscleral topography plot is displayed, the practitioner then has the option of adjusting parameters and choosing which manufacturing lab and lens design to proceed with. The device will automatically generate a best-fit lens based on the design of your choosing. Many of the manufacturing labs allow for direct data transmission from the device to the lab through the Eaglet-Eye ESP "DirectConnect" software module.

Pentacam (Oculus). The Pentacam is familiar to most eye care practitioners as a Scheimpflug corneal topographer. However, Oculus now offers a cornea/scleral profile (CSP) module that can detect scleral topographic properties in addition to corneal ones. There are currently three models of the device: Pentacam, Pentacam HR and Pentacam AXL Wave. At the time of publication, the HR and AXL Wave models do not support the CSP module.

The CSP module acquires 250 images for over 100,000 data points, covering an 18mm diameter of the cornea and sclera.[6] Pentacam also supports a variety of external manufacturing labs such as EyePrint Prosthetics, Valley Contax, Bausch + Lomb and others. This technology also supports RGP and other specialty contact lens designs on a per-lab basis.

As mentioned above, the Pentacam relies on Scheimpflug imaging for image acquisition, so it does not require the use of fluorescein. This may be advantageous for fitting patients with dry eyes or with low tear production. To begin, the patient puts their chin in the rest, and the Pentacam takes 250 images throughout a series of five gazes (up, down, left, right and primary gaze). These images are then stitched together to create an ocular surface profile.

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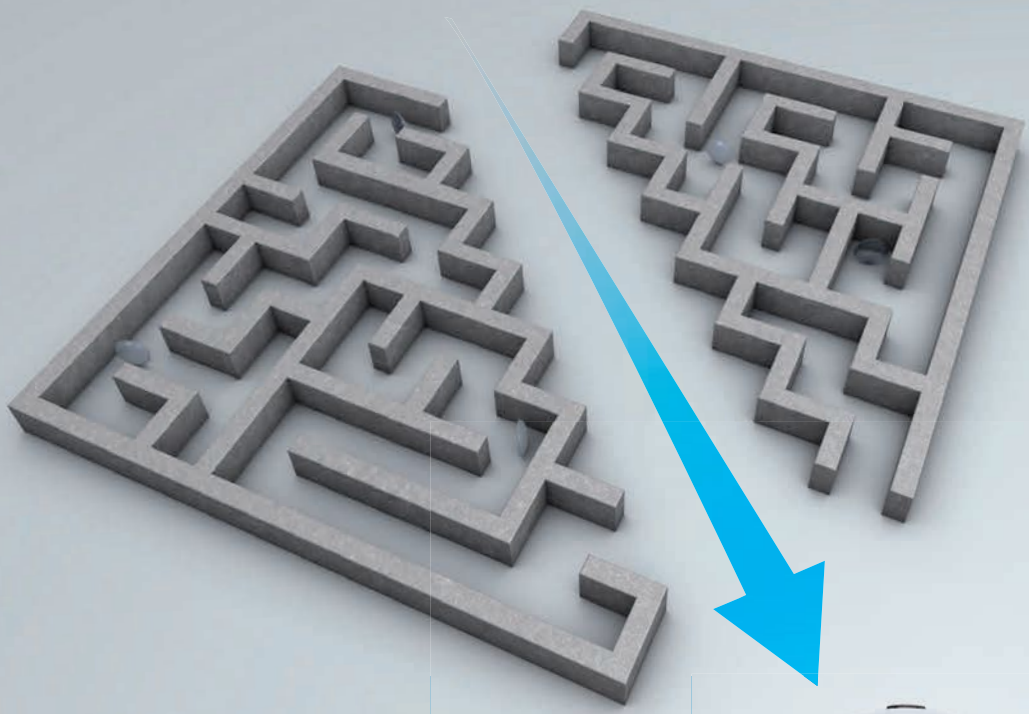

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via compatibility with external software such as BostonSight's BostonSight Scleral, EyePrint Prosthetics's ScanFit Pro or Valley Contax's Custom Stable, for example.

An EyeFit Pro impression of the ocular surface.

IMPRESSION-BASED LENSES

While not technically scleral topography, impression-based technology can offer similar benefits. The most common impression-based technology is from EyePrint Prosthetics (EyePrint Pro). EyePrint Prosthetics offers three lens designs. The first is the EyePrint Pro lens, capable of constructing an exact replica of the ocular surface, regardless of pathology like filtering blebs, tube shunts and even symblepharon.

The company's middle tier offering is the EyeFit Pro—also an impression-based design. This lens also creates an exact replica of the ocular surface, but is for slightly less irregular surfaces, such as corneal ectasia, post-traumatic ocular surfaces, irregular corneal transplants and Salzmann's nodules. Finally, its newest offering is the ScanFit Pro. This one is the most affordable lens design and is used in conjunction with the aforementioned scleral topographers, foregoing the impression process completely. It is typically used for corneal ectasias, pinguecululae and other small surface nodules. All lens offerings from EyePrint Prosthetics are freeform technology.

COMBINING TECHNOLOGIES

Scleral topographers can also be used in tandem with other instruments to create some truly unique designs, which further widens the breadth of what can be corrected. One example is decentered optics. This concept is useful in patients with irregular pupils (*e.g.*, post-trauma patients) or abnormal visual axes (*e.g.*, scarring, corneal ectasia, corneal transplants).

Another example of combining technologies is higher-order aberration (HOA) correction. We know that many ocular surface conditions, most notably corneal ectasia, cause an increase in HOAs. Because freeform scleral lenses typically do not show marked rotation on the ocular surface, this makes them an ideal solution for HOA wavefront correction, which in turn can be quite significant to improving visual acuity in many patients.[7]

TAKEAWAYS

Scleral topography and profilometry can be excellent tools both for those practitioners new to scleral lens fitting and those who are expert fitters already. These devices allow for the elimination of the traditional "trial-and-error" process of scleral lens fitting—empirically providing either the best-fit lens or freeform lenses that match the ocular surface on a micron scale. This can help you save time and money while ensuring optimum comfort and vision for your patients. They can also be combined with existing technologies such as HOA correction and decentered optics to create truly personalized options per patient, per eye.

Traditionally, fitting scleral contact lenses can be complicated. While the best way to learn more about scleral lens fitting and the treatment of ocular surface conditions is to get involved by going to conferences, talking with colleagues and seeking out mentors and using and understanding emergent technology like scleral topographers can greatly reduce the steep learning curve, making life easier for both the patient and the practitioner alike. RCCL

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IMPROVING SCLERAL SUCCESS:

TROUBLESHOOTING TIPS FROM A PRO

An expert gives clear guidance on how to correct common problems—or avoid them in the first place.

By Langis Michaud, OD

Scleral lenses have always been part of the scene, especially when treating diseases of the ocular surface or restoring vision in patients with irregular corneas or ectasia. While they were once reserved for an elite of prescribers, technological development has made it possible to democratize their use over the past 10 years. Now, there are thousands of practitioners around the world who offer this modality, which is slowly but surely becoming a standard offering in a contact lens practice.

While it is true that, not long ago, fitting scleral lenses was more an art than science, multiple research and clinical studies made a difference. We have now a better understanding of the anatomy of the ocular surface and its relationship with scleral lenses. Using the devices at our disposal, it has never been easier to fit scleral lenses. Today, it's even common to proceed with a totally empirical approach by deriving lens parameters from the profiles obtained via mapping devices, which are increasingly reliable. The lens design itself has been refined, now offering adjustment possibilities by quadrant and zones, all in order to minimize physiological impacts on the eye and optimize the vision provided.

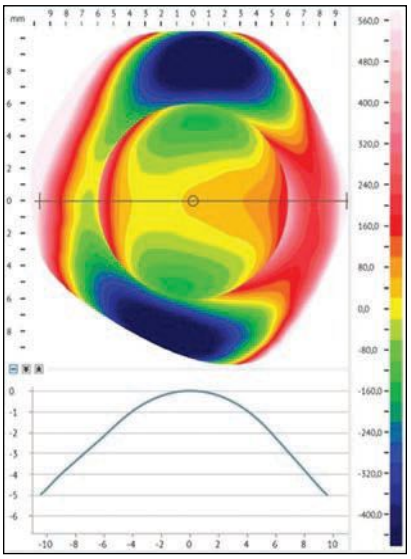

Fig 1. Profilometry of the conjunctiva shows irregular toricity of more than 450 um.

All of these changes resolved a number of issues that existed in the past. They also generated other effects with which we have to learn to fix. Here, we discuss the most common problems and attempts to provide practical solutions to solve them.

FITTING ISSUES

It is now known that the sclera and the conjunctiva are asymmetric, toric, non-rotational surfaces.[1] Almost three out of four eyes present a conjunctiva with irregular toricity (unrelated to the corneal form), which increases further away from the limbus.[2] It's not uncommon to see differences of 300µm to 500µm between ocular surface peaks and valleys. There is no such thing as two perpendicular principal meridians to describe conjunctival profile. Very rarely, the surface varies according to a predictable sinusoidal pattern. As a result, it is difficult to perfectly align the lens with the ocular surface unless a landing zone is derived from an optical or physical molding.[3]

ABOUT THE AUTHOR

Dr. Michaud graduated from L'École d'optométrie de Université de Montréal and is currently the dean of the School of Optometry. He is a Diplomate of the American Academy of Optometry and a Fellow of the British Contact Lens Association, the Scleral Lens Education Society and the European Academy of Optometry. He has received honoraria from Bausch + Lomb, CooperVision and Acculens.

The issues depend on the misalignment that occurs in a given quadrant. When the lens is flatter than the surface, it lifts off, causing discomfort (lens-to-lid interaction). This open door also allows entry of debris in the reservoir, attracted by the sub-atmospheric pressure acting as a suction force. When substantial, debris contributes to mid-day fogging.[4] One way to limit its presence is to better draw the peripheries of the lenses in order to obtain an impeccable alignment in all the meridians.

On the other hand, when the lens is too steep relative to the ocular surface, it generates compression of the tissue and blood vessels.[5] Adjacent tissues can also become more swollen, increasing the compressive effect as hours of wear accumulate. The lens becomes difficult to remove at night, and rebound redness occurs within seconds of removal. Besides the aesthetic concern, this type of compression is associated with marked discomfort—a feeling of dry eyes and constant pressure felt in the eye.[6] Usually, the patient cannot tolerate this type of long-term effect.

At the slit lamp, it is extremely difficult to assess the adjustments, in microns, that must be made when the lens-to-conjunctiva alignment is deficient. Another issue arises when we qualify these defects in terms of steps. A "step 2" curvature for one lab will imply a difference of 60µm from standard, while for another manufacturer it will mean 100µm. Therefore, it is strongly recommended to qualify in numbers (microns) and not in steps (step 1-2-3, flat 1-2-3) the changes required, so that all speak the same language.[7]

For reference, conjunctival toricity becomes significant when it reaches 300µm or more. To match this, scleral lens toricity must be designed with equivalent toricity. Change in peripheral curve toricity must reach at least 100µm to become clinically

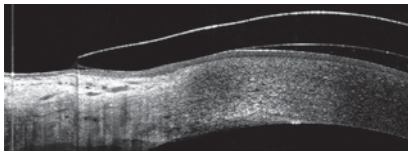

Fig 2. Edge of the lens lifting off the conjunctival surface.

significant. With less than that, the lens will behave like one designed with spherical peripheral curves.

The best approach is to perform a conjunctival scan—before wearing scleral lenses—and to establish a reliable profile. Three devices exist for this purpose.[8] They not only provide a topography of the conjunctival surface, but their software can also suggest the parameters of the first lens to be fitted. Some also offer the option of forwarding the scans to the lens manufacturer where consultants can help interpret them and design an optimal lens for the patient.

Another advantage: by analyzing these scans, it is possible to predict lens rotation, and therefore its stabilization axis (usually the steeper). This makes it possible, in advance, to compensate for the orientation of the frontal toric power when the latter is required. It is therefore becoming increasingly easy and obvious that scleral lens fitting can and should be done empirically.[9] As a result, practitioners gain efficiency and precision, while minimizing the chair time required for the multiple trials that the use of diagnostic lenses entails. This saved chair time amply justifies the investment in reliable instruments that allow the ocular surface to be measured with precision.

The use of conjunctival topographers also makes it possible to better define the limbus. The most recent research data indicates that the cornea is oval and that the elevation of the limbus is not symmetrical, like a potato chip.[10] These differences generate variations in the thickness of the reservoir above the limbus. The vault over the limbus is a critical element in the presence of mid-day fogging and also of conjunctival prolapse.[11] Therefore, it's necessary to minimize the vault to limit the entry of debris and prevent the conjunctival folds from forming. Scans can indicate the need for a quadrant-specific limbal vault, which improves the overall fit of the lens.

Some of the manufacturers now offer the option of designing a toric, oval-shape optic zone, accommodating variations of the limbal elevation and diameter, as well as quadrant-specific peripheral curves that are independently designed.[12] A better designed, better aligned lens will tend to be less off-center. However, the centration of the lens is a fundamental element in the visual correction of patients.

VISUAL ACUITY ISSUES

One of the biggest frustrations I have when fitting scleral lenses is not being able to correct the patient's visual acuity optimally. This is often the case with patients showing nipple cones or low/emerging oval cones, whose visual acuity in glasses is correctable to 20/30 or better. Fitted with sclerals, these patients complain of shadowing, ghosting of images and stretching of letters. Most of the time, they prefer their glasses to these new devices supposedly prescribed to improve their visual outcome.

Intuitively, these symptoms relate to the presence of residual astigmatism, coming either from the posterior cornea, crystalline lens or generated by a presumed flexure of the scleral lens.

Clinical studies have shown that it is quite different. Although a common recommendation from consultants, using a front toric lens will not help to improve visual acuity of these patients. Some practitioners may find signs of astigmatism when performing retinoscopy, but subjectively, if vision is slightly improved with

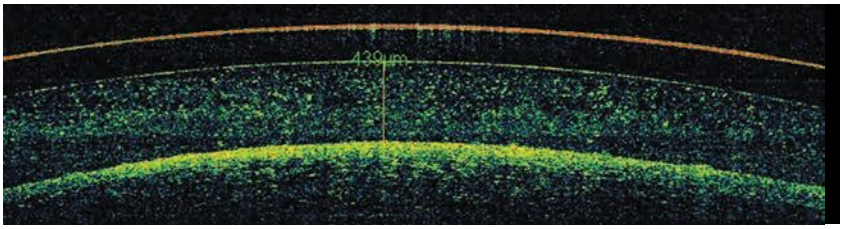

Fig 3. Debris is accumulated in the reservoir (mid-day fogging).

toric over-refraction, there is never a "wow" moment associated with it. Increasing lens thickness to fix flexure does not provide any good outcome as it was also proven. Thin lenses fitted with low vault have the same behavior as thick lenses fitted with greater fluid reservoir, and the same symptoms are found.[13]

Why is this? Simply because the true nature of the visual symptoms is neither physiological nor related to the lens parameters, but indeed manifestations of higher-order aberrations (HOAs), in particular vertical coma.[14] They are highly disturbing because the patient never experienced them before, or most often HOAs naturally present in their eyes are of the opposite direction and partly suppressed, with the patient being used to it. The new ones, generated by lens wear, are consequently more disturbing and obvious.

There are several reasons for the presence of these aberrations. First, the multiple indices change at each interface of the media (air, tears, anterior lens surface, posterior lens surface, fluid reservoir, front cornea, back cornea and to a lesser extent aqueous humor), influencing the light passing through the eye.

The second reason is linked with the lens centration. Most scleral lenses—in particular the larger ones—will tend to decenter down and temporal, following gravity and the natural slope/shape of the conjunctival surface. Consequently, the optical axis of the lens is no longer positioned in front of the visual axis, generating HOAs. The fluid reservoir is also modified, now shaped as a prism, and contributing to the degradation of the image perceived.

Finally, in keratoconus, posterior and anterior cornea tend to compensate for each other in aberrations generated. When a scleral lens is worn, most of the anterior corneal surface is compensated and HOAs from the back surface are now fully expressed.[15]

There are several ways to fix these issues. We identified adding a front toric power or increasing lens thickness as not effective. Another, more promising avenue is to generate an optical surface that will compensate HOAs in the system. Topo-guided wavefront optics can be (not so easily) generated; a few manufacturers are beginning to offer this option.

Stabilization of the lens is crucial here. Any rotation or movement will modify the level and pattern of HOAs, and their compensation on the lens will then be useless. Another possibility is to increase anterior surface asphericity without generating convex power. It works well, but the difficulty is to determine the right level of asphericity for a given eye. Again, many trial-and-error sessions may occur and can generate frustration on both patient and practitioner sides. The easiest method to limit HOAs' influence on acuity is to make sure the lens is not decentered. This means to align the lens with the conjunctiva after proper mapping of its surface, as discussed previously.

It may be intuitive as well to rely on a smaller diameter lens, which would be fit with a lower vault. For instance, a 15mm lens would—in theory—deal with less conjunctival toricity than a 17.5mm one; this is not true. The important factor to consider here is the primary functional diameter (PFD) of the lens rather than its overall diameter. PFD is determined by the first contact of the lens with the ocular surface. Manufacturers can determine this value and must disclose it.

It is not rare to see a 16mm lens landing at 14mm and a 15.5mm OAD landing at 13.5mm. Consequently, there is not much difference between these two lenses and both will behave about the same on the same ocular surface. The rest of the lens, over PFD, can be considered just plastic with very limited impact on the lens behavior and its support from the ocular surface, except if the lens is not designed with curves but with tangents (like PROSE or Visser's design). For the latter, peripheral curves are more closely aligned and match the conjunctival profile whilst with curves, the lens is generating a series of touch point on the landing area, the first one supporting most of the lens mass. In the case of tangent designs, diameter matters, which is not the case for 90% of the other lenses in the market.

PHYSIOLOGICAL ISSUES

An article published in 2012 suggested a theoretical model to evaluate hypoxic stress related to scleral lens wear.[16] Based on this, the cornea remains free of hypoxic stress if the lens was made thin (250µm or less) and fitted with lower vault (200µm or less), assuming that it was made with the highest Dk. This was a simple model looking at direct diffusion of the oxygen through the lens and the fluid reservoir, assuming that there was no tear exchange, no tear mixing and no lateral diffusion of oxygen in the cornea.

The article generated an ongoing debate and has since been cited more than 100 times, which says a lot about the importance of this topic and the extensive research that was carried out in the last 10 years.

We now know that the model is more complicated that was suggested. Oxygen diffusion is more complex and we have to take in account the carbon dioxide exchange as well, and the impact of both on the corneal metabolism.[17] Limbal vessels may play a role that was not considered in the previous model. Consequently, the model must be revisited.

What became obvious is that corneal edema occurs with scleral lens wear very rapidly after lens insertion and stabilizes to reach a plateau after 90 minutes post insertion. Edema is stromal in nature, not epithelial, and is better evaluated through OCT, while the lens is worn compared to pachymetry once the lens is removed. The level of corneal edema never exceeds 4%, which is considered physiological, except if the lens is worn under closed-eye conditions.

On average, edema reaches 1% to 2%, which is considered safe for a normal cornea by most authors. This is probably where the debate begins. The first mistake is that we compare apples and oranges. Physiological edema reaches 4% after night eye closure but fades away over one hour upon awakening. The cornea is then restored to its normal metabolism. When scleral lenses are worn, they are applied after a few minutes of eye opening in the morning. Cornea is still edematous and will remain in this status during all wearing hours. Chronic hypoxic stress occurs with no known long-term impact.

Though a normal, young, healthy cornea can probably sustain such stress—even in the long term—this is a different story for fragile corneas, penalized by a weakened endothelial cell function. The perfect example of

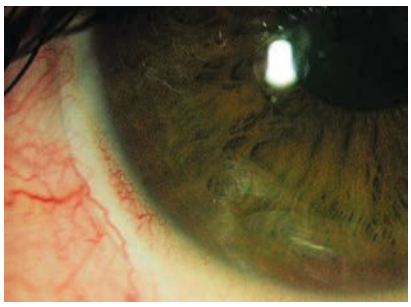

Fig 4. Conjunctival compression with blanching.

such a case is a post-graft patient.[18] It is now the standard of care to consider every other option than sclerals, if possible, on eyes in which the cornea displays less than 800 cells/mm^2.[6] If there is no other option to restore vision or to treat the ocular surface, a close follow-up of the cornea is mandatory. Fenestration and channels must be considered to improve tear exchange. Lens fitting and design must be optimized.

Oxygen permeability of a lens is evaluated by dividing the material permeability by its thickness. It is now possible to manufacture scleral lenses with very high Dk (up to 200), which must be favored. The lens thickness varies according to its design and its power. Minimal thickness is mandatory to alleviate lens break and to keep its geometrical stability. Optimal lens thickness varies from 200µm to 300µm.[19] Another factor in play is the fluid reservoir. The same logic applies here: oxygen permeability of the reservoir is dictated by its thickness and the fluid permeability, which is very low (around 80).[20]

Fig 5. Corneal edema is visible under the scleral lens.

Studies showed that, between the two factors and up to 4% edema, lens material permeability is the most important one.[17] Under chronic, severe hypoxia or when the fluid thickness is highly excessive (600µm to 800µm), the reservoir increases its impact on the corneal metabolism.[20] However, at a very high vault (>800µm), there is a gradient between the temperature of the fluid at the corneal surface and the one lying just under the lens surface. This difference generates a tear mixing that does not exist otherwise, which can contribute a marginal increase in the level of oxygen delivered to the cornea.

Consequently, best practices these days strongly suggest prescribing scleral lenses with the highest DK/t possible, at least on fragile corneas. I am not comfortable leaving any other eyes under hypoxic stress, as minimal as it is, especially when you can fit better with zero hypoxic stress and still keep all the benefits of the scleral lenses.[22]

The last element to discuss showed up on our radar a few years ago. Charles McMonnies suggested that, in theory, scleral lenses can induce a rise in intraocular pressure (IOP) by compressing episcleral veins and the subatmospheric pressure can cause lens tightness over time. Several clinical studies were carried out to validate this statement.[23]

Some of these studies tend to evaluate IOP during lens wear, either before complete lens settling (<4hrs), which may underestimate the real impact of the lens on the pressure, or with equipment that is highly variable (transpalpebral) or not practical (pneumotonometry).[24,25] Others evaluated IOP just after lens removal, which led to normal findings.[26] As soon as the pressure on the episcleral veins is removed, aqueous outflow is restored rapidly and IOP goes back to normal level in a few seconds.

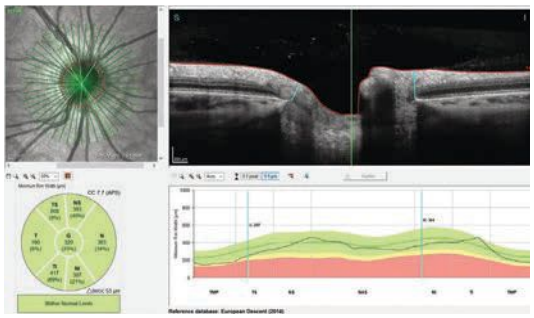

Fig 6. BMO-MRW assessed by OCT (Spectralis).

Other studies looked at the influence of the lens diameter, assuming that a larger lens would compress the ocular structures.[27] This is not true, and again, PFD of the lens is the key element to consider. A 16.5mm lens landing at 13.8mm will generate the same pressure on the episcleral veins as a 15.0 lens landing around the same area (13.5mm). Both lenses will contribute the same to vein compression. The suction effect will be the same if the reservoir vault is kept constant.

Finally, two manuscripts were published about the optic nerve reaction during lens wear, especially the Bruch's membrane opening minimal rim width (BMO-MRW), which was found—at least in animal models—to be a valid marker of any IOP variation. One of these studies did find a slight variation of IOP (increase) but without reaching statistical significance.[28] Just one eye was fit, the fellow one acting as a control, which may be not the state-of-the-art (synergistic effect). The other study took into account diurnal IOP variation and found a significant increase of IOP over six hours of lens wear (+5mm Hg).[29] The same eye was compared to itself.

These conflicting results show the high variability among lens wearers. Nobody can predict who will spike in IOP and who will not be affected, but the reality is that some patients will indeed present a significant increase during scleral lens wear.

To fix this issue is quite simple: relieve some pressure by alleviating lens tightening over time. One thing is certain—scleral lenses would not generate glaucoma, at least on healthy optic nerve patients. For those already affected by glaucoma or at risk, caution is in order. In such cases, other options must be explored; if sclerals are finally selected, a close follow-up is mandatory, including optic nerve photo/scan analysis and visual field.

TAKEAWAYS

The last 10 years have given us incredible technological advances in terms of clinical data acquisition as well as the possibilities of manufacturing lenses. These innovations have multiplied the possibilities of adaptation to sclerals, which is beneficial for patients but can prove problematic for the practitioner, who must therefore juggle numerous options. It may be hard to master, especially for those fitting a limited number of cases per year. Some issues of the past persist, some have been solved and new ones are emerging. On each occasion, the practitioner must find ways to fix them. Hopefully, technology is there to help us. RCCL

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DETAILING THE DYSTROPHIES

Sort out the subtle differences among these corneal anomalies with this overview of presentation and management.

By Christina Cherny, OD, and Suzanne Sherman, OD

Corneal dystrophies are rare ocular disorders that often present in early childhood and arise independent of systemic and environmental factors; they are characterized by a hereditary, bilateral and symmetric nature.[1,2] Dystrophies are typically slowly progressive and non-inflammatory, with some cases resulting in corneal opacification.[1-3] Clinicians need to be able to distinguish these disorders from degenerations—changes from natural aging or disease that result in loss of normal corneal properties—although the distinction between the two is not always clear.[2] Careful examination of every corneal layer in both eyes is required in order to confirm bilateral disease.[2,4]

Dystrophies are often confined within a specific layer and have traditionally been classified in this manner. More recently, it has been discovered that certain dystrophies may affect multiple layers simultaneously, resulting in clinicians using unique, characteristic landmarks to identify the individual dystrophies once they are fully developed.[1,4]

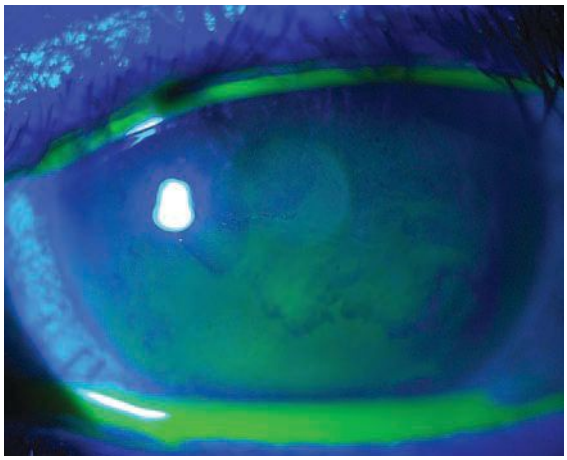

Fig. 1. A resolving erosion due to epithelial basement membrane dystrophy.

Here we dissect each corneal dystrophy, layer by layer, to aid clinicians in conducting a workup and arriving at a correct diagnosis.

EPITHELIAL AND SUBEPITHELIAL DYSTROPHIES

This category includes some of the more common forms. While troubling to the patient, they typically cause minimal vision loss.

Epithelial basement membrane dystrophy (EBMD). Also known as map-dot-fingerprint dystrophy, this condition is sometimes classified as a corneal degeneration due to minimal documented cases of autosomal dominant familial inheritance; many cases appear isolated or secondary to corneal trauma (*Figure 1*).[1,2,4] EBMD occurs due to poor basal epithelial cell adhesion to the basal lamina, which predisposes the eye to recurrent corneal erosions (RCE).[1]

Corneal landmarks include grayish dots and lines in a characteristic pattern as well as cysts and blebs that may appear as thickened, hazy and gray epithelium with scalloped borders.[1,2,4] While some patients may be asymptomatic, others can present with decreased

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visual acuity and/or monocular diplopia, as well as painful, recurring episodes of corneal erosion that may wake the patient.[1,2]

Initial RCE treatment is typically conservative, consisting of copious lubrication particularly during episodes of acute RCE.[2] If corneal erosions persist or the dystrophy becomes visually significant, consider surgical RCE treatment options; these include phototherapeutic keratectomy (PTK) and anterior stromal puncture (ASP).[2]

Epithelial recurrent erosion dystrophies (EREDS). These entail episodes of painful epithelial erosions lasting under one week, with erosive episodes often commence at night.[1,2,4] Although the frequency of erosive episodes slowly diminishes with advancing age, it's possible for diffuse, central subepithelial opacities to develop and impair visual acuity—with keloid formations developing in certain cases.[1,2] Subepithelial pannus of the inferior cornea, intraepithelial bullae, amyloid deposition and microscopic gelatinous buds within the basal layers may be observed in EREDS variants.[1,2,4]

Conservative treatment of RCE may be sufficient during recurrent episodes; surgical RCE treatment, such as debridement or stromal puncture, is often necessary to address the fibrinous pannus and remove corneal opacities.[2,4]

Meesmann corneal dystrophy (MECD). This condition may follow a static or slowly progressive course resulting in moderately decreased in visual acuity.[1,2,4] Signs of MECD include tiny, translucent intraepithelial vesicles extending from limbus to limbus that are more densely concentrated within the interpalpebral area, as well as diffuse gray opacities.[1,2] Patients may be asymptomatic but can be symptomatic for decreased vision, glare, light sensitivity and foreign body sensation, with superficial punctate keratopathies or RCEs contributing to surface pain.[1,2,4]

Treatment consists of ocular lubrication, bandage contact lens wear in cases of epithelial defects or surgical RCE treatment in severe instances.[2,4]

Lisch epithelial corneal dystrophy (LECD). Characterized by clear microcysts as well as gray-white opacities in whorl, radial, band, feather and club-shaped patterns, LECD progresses slowly from periphery to the center, with an appearance similar to that of band keratopathy.[1,2,4] Patients may be asymptomatic or present with decreased vision or photophobia.[1,2,4] Treatment options include bandage contact lens wear, PTK and limbal stem cell transplantation.[4]

Gelatinous drop-like corneal dystrophy (GDLD). This progressive epithelial dystrophy displays a higher prevalence in the Japanese population.[1,2,4] GDLD may present similarly to band keratopathy with multiple mulberry-like opacities that stain with fluorescein.[1,2,4] Other signs include superficial vascularization, stromal opacification and large kumquat nodules.[1,4] Patients may be symptomatic for decreased vision, photophobia, tearing, redness and foreign body sensation.[1,2,4] Although surgical options are available, including superficial keratectomy, lamellar keratoplasty and penetrating keratoplasty, recurrence is common within a few years and may result in visual disability.[1,2]

EPITHELIAL-STROMAL TGFBI DYSTROPHIES

Conditions in this category all arise from an autosomal-dominant mutation in the gene responsible for production of transforming growth factor β-induced (TGFBI), an extracellular matrix protein involved in cellular adhesion. Some of these disorders can be difficult to distinguish from one another, given their similar symptoms.

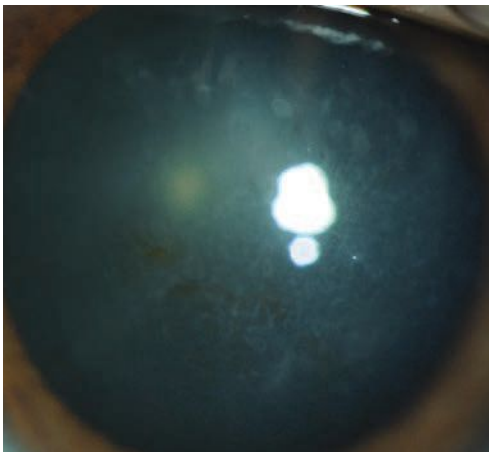

Fig. 2. Signs of Reis-Bucklers corneal dystrophy include confluent geographic-like opacities at Bowman's layer that become less discrete.

Reis–Bucklers corneal dystrophy (RBCD). This condition results in early visual impairment.[1,2,4] Signs include confluent geographic-like opacities at Bowman's layer that become less discrete and later extend to the limbus and deeper into the stroma[1,2] (*Figure 2*). Vision slowly deteriorates and painful RCEs occur, although they diminish with time.[1,2] RCEs are treated conservatively, while PTK and automated lamellar keratoplasty (ALK) are used to treat visual impairment. Recurrences are often seen after one year post-op.[4]

Thiel–Behnke corneal dystrophy (TBCD). Often difficult to distinguish from RBCD, this dystrophy's signs include flecks or diffusely scattered irregular opacities at Bowman's layer, with later symmetrical subepithelial honeycomb opacities.[1,4] Although the opacities are often central, they can progress to the corneal periphery and deeper

into the stroma with age.[1] Patients may be symptomatic for RCEs during earlier decades with slowly progressive visual impairment due to scarring.[1,2] Treatment includes lubrication, PTK or ALK, although there is a risk for recurrence.[4]

Lattice corneal dystrophy, type 1 (LCD1). Amyloid deposition within basement membranes in LCD1 patients gives rise, early on, to superficial central fleck opacities and deeper fine, translucent lattice lines within the superficial stroma. These opacities later spread throughout peripheral cornea and within deeper layers, avoiding far peripheral stroma.[1,2] The deposits contribute to neuropathy and lead to corneal hypoesthesia, while RCEs also occur and contribute to pain and visual impairment.[1,2]

By the fourth decade, patients are often symptomatic with significant visual impairment.[1,2] Deposition within the trabecular meshwork may contribute to ocular hypertension and glaucoma, while deposition within other organs can lead to numerous systemic complications.[2] Treatment involves PTK, DALK or PKP, although deposits may recur post-op; intraocular pressure (IOP) must continue to be monitored.[1,2,4]

Granular corneal dystrophy, type 1 (GCD1). Hyaline deposition produces a radiating vortex pattern superficial to Bowman's layer in these patients.[1,4] Over time, dense crumb-like and snowflake-like granules arise but do not extend to the limbus, although they may extend to the deeper stroma.[1,4] Symptoms include glare, photophobia and severely decreased vision; RCEs may transpire.[1,4] Treatment includes conservative and/or surgical RCE treatment, although the deposits may recur.[4]

Granular corneal dystrophy, type 2 (GCD2). Also known as Avellino dystrophy, this condition presents

Table 1. Corneal Dystrophies: Inheritance and Onset

Dystrophy Name	Inheritance	Onset
Epithelial basement membrane dystrophy (EMBD)	Sporadic	Adulthood
Epithelial recurrent erosion dystrophies (EREDs)	Autosomal dominant	First decades
Meesmann corneal dystrophy (MECD)	Autosomal dominant	Early childhood
Lisch epithelial corneal dystrophy (LECD)	X-linked dominant	Childhood
Gelatinous drop-like corneal dystrophy (GDLD)	Autosomal recessive	First or second decade
Reis–Bucklers corneal dystrophy (RBCD)	Autosomal dominant	Childhood
Thiel–Behnke corneal dystrophy (TBCD)	Autosomal dominant	Early childhood
Lattice corneal dystrophy, type 1 (LCD1)	Autosomal dominant	First two decades of life
Granular corneal dystrophy, type 1 (GCD1)	Autosomal dominant	Early childhood
Granular corneal dystrophy, type 2 (GCD2)	Autosomal dominant	Early childhood
Macular corneal dystrophy (MCD)	Autosomal recessive	Childhood
Schnyder corneal dystrophy (SCD)	Autosomal dominant	Childhood
Congenital stromal corneal dystrophy (CSCD)	Autosomal dominant	Birth
Fleck corneal dystrophy (FCD)	Autosomal dominant	Congenitally or in the first years of life
Posterior amorphous corneal dystrophy (PACD)	Autosomal dominant	Congenitally or in the first decade
Central cloudy dystrophy of François (CCDF)	Autosomal dominant	First or second decade
Pre-Descemet corneal dystrophy (PDCD)	Isolated	After 30 years of age
Fuchs' endothelial corneal dystrophy (FECD)	Isolated or autosomal dominant	In or after the fourth decade; early variant presents in the first decade
Posterior polymorphous corneal dystrophy (PPCD)	Autosomal dominant	Early childhood
Congenital hereditary endothelial dystrophy (CHED)	Autosomal recessive	Birth
X-linked endothelial corneal dystrophy (XECD)	X-chromosomal dominant	Birth

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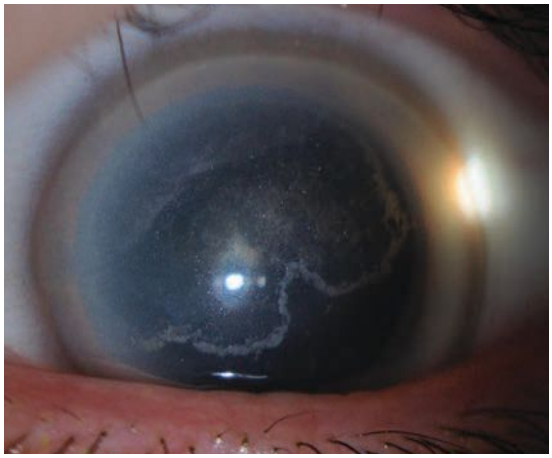

Fig. 3. Signs of Schnyder corneal dystrophy include decreased photopic visual acuity, glare and decreased corneal sensitivity.

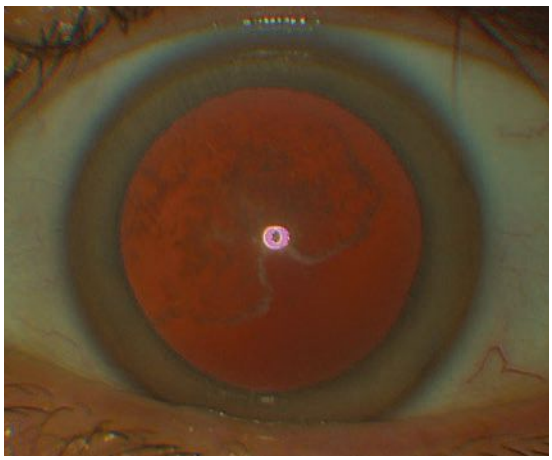

Fig. 4. Schnyder corneal dystrophy as seen on retroillumination.

with subtle, superficial stromal white dots due to hyaline and amyloid deposition; these progress to dense superficial patches with moth-eaten centers that may appear star- or ring-shaped and accompany branching stromal opacities.[1,4] Patients experience RCEs and progressively decreased vision with age.[1,4] Therapy includes conservative RCE treatment or PTK, although recurrences may take place.[4]

STROMAL DYSTROPHIES

Given that the stroma comprises the lion's share of the cornea, there is a wide array of dystrophies affecting this layer. Systemic associations are possible—be sure to investigate any risk factors noted in the medical history.

Macular corneal dystrophy (MCD). Keratin sulfate deposition leads to the development of superficial, central, white snowflake opacities and fog involving the limbus and deep stroma in these patients.[1,2,4] With time, corneal sensitivity diminishes, diffuse stromal haze evolves and Descemet's membrane grays and develops guttae.[1,2] Patients may develop RCEs and photophobia, with vision diminishing severely by the second decade.[1,2,4] Lamellar keratoplasty is often required to treat deep stromal deposition, although PKP is indicated if there is endothelial involvement; recurrences may be observed.[2,4]

Schnyder corneal dystrophy (SCD). Although this condition often begins in childhood, diagnosis is usually made by the third decade, with identification further delayed in crystalline phenotypes, which comprise 50% of cases *(Figures 3 and 4)*.[1] Cholesterol is deposited within the corneal layers and leads to ring-like central opacities, comma-shaped subepithelial crystals, arcus lipoides and, with age, midperipheral stromal haze.[1] Patients are symptomatic for decreased photopic visual acuity, glare and decreased corneal sensitivity, and may have associated dyslipidemia and xanthelasma.[1,2]

Ocular treatment includes PTK, deep ALK (DALK) or penetrating keratoplasty (PKP), and systemic cholesterol-lowering agents are often indicated.[1,2]

Congenital stromal corneal dystrophy (CSCD). Over a slowly progressive course, these patients will demonstrate bilateral diffuse corneal clouding with densely scattered white stromal opacities throughout the entire cornea.[1,2,4] Patients may be symptomatic for photophobia and severe vision loss, which may be accompanied by strabismus.[1,2]

Ocular treatment options for this dystrophy include DALK and PKP, as well as treatment for amblyopia and strabismus. Systemic associations include type 1 diabetes or slowly progressive chronic renal involvement, which necessitates evaluation of blood glucose and renal function.[2]

Fleck corneal dystrophy (FCD). This non-progressive disorder consists of tiny translucent discoid opacities or white stromal flecks, which may extend to the limbus.[1,2,4] Patients may be asymptomatic, although mild photophobia and diminished corneal sensitivity may occur.[1] Due to stable visual acuity, treatment is seldom indicated.[2]

Posterior amorphous corneal dystrophy (PACD). This condition presents as diffuse white, sheet-like opacities of the posterior stroma that may extend peripherally and to the limbus.[1,4] Decreased corneal thickness, corneal flattening and hyperopia are often associated, as well as cornea plana.[1,2] Additional reported associations include scleralization of the peripheral cornea, iris malformations, prominent Schwalbe's line, iris processes, pupillary remnants, irido-corneal adhesions, corectopia and pseudopolycoria, although there is no association with glaucoma.[1,2]

Patients may be asymptomatic

Table 2. Corneal Dystrophies: Characteristic Findings on OCT

Dystrophy Name	OCT Findings[7-16]
Epithelial basement membrane dystrophy (EMBD)	• Increased reflectivity of epithelial basement membranes • Basement membrane duplication • Intraepithelial hyporeflective cysts
Epithelial recurrent erosion dystrophies (EREDs)	• NA
Meesmann corneal dystrophy (MECD)	• Diffuse hyporeflective microcysts throughout epithelium
Lisch epithelial corneal dystrophy (LECD)	• Epithelial and subepithelial hyper-reflectivity
Gelatinous drop-like corneal dystrophy (GDLD)	• Hyperreflective nodular formation in anterior stroma
Reis–Bucklers corneal dystrophy (RBCD)	• Hyperreflectivity within Bowman's
Thiel–Behnke corneal dystrophy (TBCD)	• Hyperreflective saw tooth material on Bowman's, extending into epithelium • Ridges and furrows within stroma • Irregularly thickened or thinned epithelium
Lattice corneal dystrophy, type 1 (LCD1)	• Hyperreflective material in mid stroma
Granular corneal dystrophy, type 1 (GCD1)	• Merging hyperreflective deposits in epithelium and anterior stroma • Individual hyperreflectivities in middle stroma • Shadows posterior to deposits
Granular corneal dystrophy, type 2 (GCD2)	• Hyperreflective material in anterior stroma with clear intervening spaces
Macular corneal dystrophy (MCD)	• Elongated hyperreflectivity in epithelium, Bowman's and stroma • Hyperreflectivity of entire stroma
Schnyder corneal dystrophy (SCD)	• Epithelial hyperreflectivity. • Diffuse hyperreflectivity of anterior, mid and posterior stroma • Maximum reflectivity of anterior stroma • Hyporeflective stromal striae
Congenital stromal corneal dystrophy (CSCD)	• NA
Fleck corneal dystrophy (FCD)	• NA
Posterior amorphous corneal dystrophy (PACD)	• Diffuse stromal thinning • Posterior stromal hyperreflectivity
Central cloudy dystrophy of François (CCDF)	• NA
Pre-Descemet corneal dystrophy (PDCD)	• NA
Fuchs' endothelial corneal dystrophy (FECD)	• Thickening of cornea and Descemet's membrane • Epithelial microcysts and bullae in moderate disease • Corneal folds
Posterior polymorphous corneal dystrophy (PPCD)	• Irregular endothelium bridging anatomic angle structure
Congenital hereditary endothelial dystrophy (CHED)	• Progressive Descemet's membrane thickening, particularly of the non-banded portion
X-linked endothelial corneal dystrophy (XECD)	• NA

or experience mild photophobia and impairment of visual acuity, and the disorder is stable or slowly progressive.[1,2] Treatments include PKP and DALK.[2]

Central cloudy dystrophy of François (CCDF). Presentation here resembles crocodile shagreen (a degeneration) within the anterior central cornea (*Figure 5*).[1,2,4] Signs include translucent, scaly polygonal opacities within the deep stroma surrounded by clear intervening tissue.[1,2] Treatment is seldom indicated for CCDF, as patients are asymptomatic and the course is nonprogressive.[1,2]

Pre-Descemet corneal dystrophy

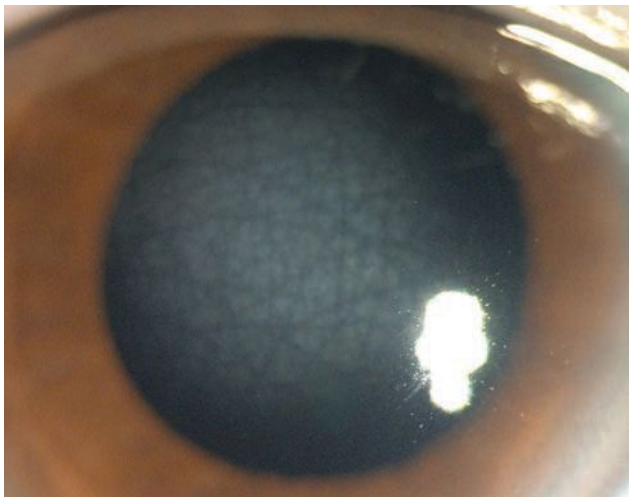

Fig 5. The appearance of central cloudy dystrophy resembles that of crocodile shagreen within the anterior central cornea.

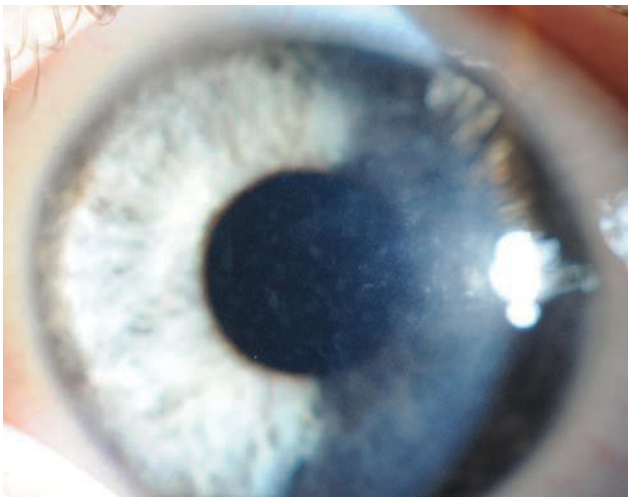

Fig. 6. Signs of posterior amorphous corneal dystrophy start out with asymmetric gray opacities of Descemet's membrane and endothelium.

(PDCD). This generally presents with punctiform, focal gray opacities in the deep stroma anterior to Descemet's membrane, which may resemble corneal farinata.[1,2,4] Many forms of PDCD are nonprogressive and patients remain asymptomatic, with no indicated treatment.[1] Due to common systemic association with ichthyosis, dermatologic work-up may be warranted.[2]

ENDOTHELIAL DYSTROPHIES

These disorders, characterized by opacities or lesions, can threaten corneal function as well as vision, given their location.

Fuchs' endothelial corneal dystrophy (FECD). The most prevalent of this category of dystrophies, this condition displays a female predominance and presents with characteristic corneal guttae—wart-like deposits on the inner aspect of Descemet's membrane resembling beaten metal that start centrally and disseminate peripherally.[1,2] In later stages, endothelial decompensation occurs with subsequent stromal edema, epithelial bullae, subepithelial fibrosis and superficial vascularization.[1,2,4]

Patients are intermittently symptomatic for decreased vision during episodes of epithelial and stromal edema, which is often worse upon waking, and vision may progressively deteriorate with time.[1,2,4] Bullous keratopathy may cause pain, photophobia and epiphora, with ruptured bullae resulting in epithelial erosions.[2,4]

Treatment involves hyperosmolar agents in the edematous stage to promote stromal deturgescence, bandage contact lens wear to control pain in the bullous keratopathy stage and surgical replacement of the endothelium in severe disease with Descemet's membrane endothelial keratoplasty (DMEK), posterior lamellar keratoplasty (typically Descemet's stripping automated endothelial keratoplasty) or PKP.[2,4]

Posterior polymorphous corneal dystrophy (PPCD). This condition begins with asymmetric, geographic gray opacities of Descemet's membrane and endothelium, as well as vesicular zones surrounded by gray circular opacities and/or white bands of flaky material resembling railroad tracks (*Figure 6*).[1,2,4] It's common for patients to develop corneal edema with subsequent visual impairment, which can necessitate keratoplasty.[1] Peripheral iridocorneal adhesions exist in 25% of cases and may lead to elevated IOP.[1,4] Patients may be symptomatic for foreign body sensation, decreased vision or photophobia.[2] Systemic associations include herniation and warrant a gastrointestinal work-up.[2]

Congenital hereditary endothelial dystrophy (CHED). Developing in the fifth month of gestation, with a generally nonprogressive course thereafter, CHED leads to a decrease in endothelial cell count with dysfunction.[2] The cornea appears opacified with a diffused, bluish edema and warrants differentiation from congenital glaucoma.[2,4] Additional clinical signs include a thickened Descemet's membrane, pseudo-bullous keratopathy, corneal thickening and nystagmus.[2]

CHED patients are typically symptomatic for severe visual loss and may be treated with hyperosmolar drops for edema or surgical endothelium replacement in severe disease.[2,4] Systemic association includes neurosensory hearing loss, and audiology/ENT work-up is warranted.[2]

X-linked endothelial corneal

dystrophy (XECD). This condition presents in males with congenital corneal clouding wherein the cornea may be diffusely hazy or milky-white in appearance.[1] Additional clinical signs include moon crater–like endothelial changes with possible secondary subepithelial band keratopathy and nystagmus.[2,4] Patients are symptomatic for decreased vision, although the course is minimally progressive.[1]

PTK can be performed in certain cases of subepithelial band keratopathy, while surgical endothelial replacement is performed in visually significant congenital haze.[4]

MANAGEMENT

Many corneal dystrophies produce pain and vision loss by means of RCE, and preventative measures such as topical lubrication and hypertonic saline are often necessary to protect the epithelium from erosion and bacterial infection.[3] During acute RCE episodes, bandage contact lenses may be used along with cycloplegics and antibiotic ointment.[3] Autologous serum drops, oral doxycycline, topical steroids, superficial keratectomy, PTK and ASP may be considered for recalcitrant RCEs.[3,5] Keratoplasty may be used in extreme cases, although deposits may recur post-graft, particularly in stromal dystrophies.[3,6] In certain cases, visual improvement may be achieved with the use of rigid gas permeable contact lenses.

Given the spectrum of visual disability that corneal dystrophies may cause and the possible need for surgical intervention, interdisciplinary care with cornea specialists is imperative, and systemic associations need to be evaluated by the respective specialties. Furthermore, examination of family members may be recommended to establish inheritance pattern and solidify diagnoses.

And now you're up to speed with every corneal dystrophy! Although dystrophies are rare and sometimes challenging to diagnose, doing so can make a big difference with your patients. RCCL

Is There a Role for Genetic Testing?

With the advent of genotyping, the molecular genetic basis of numerous corneal dystrophies has been described and we are now able to genetically screen for certain suspected corneal dystrophies and confirm diagnosis.[17] Online-based genetic testing services allow the clinician to order gene panels specifically for corneal dystrophies, although limited information exists regarding the quality or utility of the sequencing information. The long-term goal of genetic testing is to employ this knowledge for the development of targeted therapeutic approaches by means of gene therapy.[17,19]

The cornea may be an appealing target for gene therapy due to easy access, corneal immune privilege and often monogenic or Mendelian inheritance of corneal dystrophies.[18] However, to this day, most corneal gene therapy studies have been conducted only in animal models or *in vitro*.[18] Additionally, the mutational heterogeneity inherent to corneal dystrophies presents an obstacle to gene therapy development, which generally targets a single, primary gene defect.[19] This compels further research into mutation-independent approaches, such as by means of RNA interference followed by gene replacement.[19] Therefore, although extensive research is being performed to develop a targeted treatment for corneal dystrophies and circumvent surgical therapy, this is not the standard of care at this time.

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By Aaron Bronner, OD, and Alison Bozung, OD

Don't Be "Dup"d

Be on the front lines of managing ocular complications of medications such as Dupixent.

A 42-year-old woman requested a virtual eyecare visit for symptoms of chronic redness, dryness, tearing and itching of both eyes (*Figure 1A*). She had been treated in the past with OTC topical antihistamine and mast cell stabilizer eye drops without resolution of symptoms. She was using topical triamcinolone 0.1% cream nightly for periocular erythema and topical neomycin-polymixin B-dexamethasone eye drops once per day, both of which improved but did not resolve symptoms with the recommended dosage. She was also using artificial tears daily and Lumify (brimonidine tartrate 0.025%, Bausch + Lomb) as needed. Her medical history was significant for atopic dermatitis, for which she was prescribed Dupixent (dupilumab, Sanofi and Regeneron Pharmaceuticals) two years prior to presentation.

A tentative diagnosis of dupilumab-associated ocular surface disease was made, and the patient was asked to visit the clinic for a thorough eye exam. Use of topical steroids and Lumify was stopped until evaluation. On examination, her best-corrected visual acuity was 20/20 in each eye with no afferent pupillary defect. Her extraocular motilities and confrontation fields were full, and intraocular pressures (IOP) were 17mm Hg OD and 18mm Hg OS. Her slit lamp exam revealed bilateral periocular erythema and meibomian gland dysfunction with thickened and mildly keratinized lid margins. There was 2-3+ diffuse bulbar hyperemia with 3+ papillae in both eyes. There were multiple white focal eosinophilic aggregates at the limbus and a slightly reduced tear breakup time of seven to eight seconds bilaterally. Her dilated fundus exam was unremarkable.

The patient was started on prednisolone acetate 1% eye drops four times daily, Restasis (cyclosporine 0.05%, Allergan) twice daily, hypochlorous acid eyelid cleanser twice daily and warm compresses once per day. Upon return, her clinical findings and symptoms had improved significantly, but IOPs measured 26mm Hg OD and 29mm Hg OS. Due to steroid-induced ocular hypertension, prednisolone 1% was discontinued in favor of fluorometholone 0.1%. Dorzolamide 2% twice daily was added temporarily.

At her next follow-up, symptomatic improvement had been maintained and her IOP had improved to the mid-teens. The fluorometholone eye drops were tapered to once daily and the patient continues to use cyclosporine twice daily, eyelid scrubs and warm compresses once daily, and artificial tears as needed. With this dedicated ophthalmic management, she has been able to continue Dupixent (*Figure 1B*).

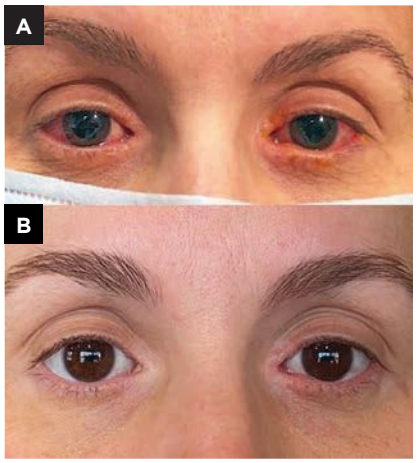

Fig. 1. (A) Initial clinic visit reveals bilateral conjunctival injection, epiphora and blepharitis. (B) Patient-provided photograph with significant improvement three months after initiation of dedicated ophthalmic therapy.

WHAT IS DUPIXENT?

A first-in-class medication for difficult-to-manage allergic or inflammatory conditions, Dupixent was FDA-approved for moderate-to-severe atopic dermatitis in 2017. Since then, the drug's indications have been extended to include moderate-to-severe asthma and inadequately controlled chronic rhinosinusitis with nasal polyposis.[1] It is currently being evaluated for use in eosinophilic esophagitis. Dupilumab is a monoclonal antibody that serves as a dual inhibitor of interleukin-4 (IL-4) and interleukin-13 (IL-13) signaling pathways. This mediation is administered by subcutaneous injection and is typically dosed every two weeks. It has been approved for use in individuals as young as six years of age.

SIDE EFFECTS

So why, as optometrists, do we need to know about this drug? Potential adverse effects include ocular side effects such as conjunctivitis and keratitis. According to a literature review, conjunctivitis was seen in anywhere from 9% to 38% of patients.[2-4] The higher incidence in more recent studies may be due to a selection bias, smaller study sizes, increased recognition of disease or exacerbations of previously undiagnosed ocular surface disease.

Don't Be "Dup"d (Continued from p. 39)

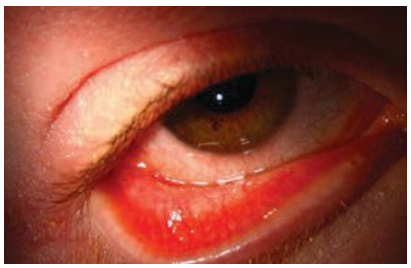

Fig. 2. This 17-year-old male with severe atopic dermatitis was started on dupilumab. There was a marked skin improvement which he called "life-changing," but shortly thereafter he developed itching, redness, epiphora, photophobia and ocular discomfort. The clinical exam revealed bulbar and palpebral conjunctival injection with a mixed papillary and follicular response, punctate erosions and filamentary keratitis.

The pathophysiology behind the development of ocular surface side effects related to dupilumab is still a point of ongoing research. Multiple studies have shown a link between the IL-13 signaling pathway and the proper maintenance of goblet cells. With IL-13 blockage, conjunctival goblet cell density is drastically decreased, which can lead to increased ocular surface inflammation, decreased tear film stability and epithelial barrier dysfunction.[5,6] Literature has also demonstrated an increase in OX40 ligand activity, eosinophilia and *Demodex* infestations, which can all negatively impact the ocular surface.[2]

A history of ocular surface disease—such as atopic keratoconjunctivitis (AKC)—and more severe atopic dermatitis prior to treatment with dupilumab are considered to be the most common risk factors in the development of dupilumab-associated ocular surface disease.[2] Though there is often phenotypic overlap in the clinical diagnoses of this disease and AKC, the histopathologic findings are quite different. AKC and allergic conjunctivitis typically manifest increased density of goblet cells and mucus production (*i.e.*, ropy discharge), whereas dupilumab reveals the opposite.

It's worth mentioning that dupilumab therapy was not associated with an increased incidence of conjunctivitis compared with the placebo when used for other conditions, such as asthma, rhinosinusitis and eosinophilic esophagitis.[7] Because these last three conditions are not directly associated with ocular surface disorders themselves, there may be an avenue for further research here.

TREATMENT

Since its debut, use of this drug has continued to climb due to its clinical success and increasing list of indications. Many severe cases of atopic dermatitis otherwise recalcitrant to treatment have shown significant skin improvement with dupilumab. Therefore, these patients are often reluctant to discontinue this life-changing therapy. In most cases, management of the ocular surface complications can allow a patient to continue their systemic treatment.

After diagnosing dupilumab-associated ocular surface disease, initiate treatment. One study suggested a treatment algorithm in which mild cases may respond well to topical ocular lubricants and antihistamine-mast cell stabilizers alone.[8] Moderate-to-severe cases will likely require topical steroid drops as a first-line treatment option. The researchers also recommended starting a steroid four times daily until symptoms resolve, then gradually tapering by one drop every two weeks to the lowest dose tolerated. Patients need to be advised of the ocular risks of steroids, which include steroid-induced ocular hypertension or glaucoma and cataract formation. Therefore, long-term steroids should be avoided if possible, especially for children or those who have these adverse effects. As seen in our patient, other steriod-sparing agents may also be used successfully in this population of patients. Studies have shown improvement in with topical cyclosporine, lifitegrast and tacrolimus.[10]

WHAT TO LOOK FOR

The presentation of ocular surface and periocular effects varies drastically between patients, so it is important to be aware of all possible findings. Clinical ocular findings include conjunctival hyperemia, follicular conjunctivitis, limbal nodules, filamentary keratitis, dry eye, blepharitis and *Demodex* infestation. More severe complications include conjunctival cicatricial changes, madarosis, punctal stenosis, cicatricial ectropion and limbal stem cell deficiency.[8,9]

MOVING FORWARD

It's important to recognize that this class of medications is not going away. In fact, ongoing research has delivered promising results so far for at least two other monoclonal antibodies that also block IL-13 signaling pathways. Both lebrikizumab and tralokinumab are currently in clinical trials, indicated for atopic dermatitis.[11,12]

As primary eyecare providers, we should be on the front lines of

managing the ocular complications of these medications. Optometrists should consider reaching out to their local dermatology practices and offering their clinical expertise to help manage these patients. This new drug class has been life-altering for so many individuals—we can't lose sight of the fact that our profession can help them in more ways than one. RCCL

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By Christine W. Sindt, OD

Open for a Surprise

Dilation reveals a rare, ring-shaped sequela of cataract surgery.

A 24-year-old male presented with bilateral aphakia, having undergone extracapsular lensectomies and anterior vitrectomies at six months of age. Acuities were 20/40 OD and OS while wearing soft contact lenses and over-refraction glasses to correct astigmatism and near vision. Dilation did not improve retinal viewing due to the presence of a dense Soemmering's ring, an opaque white layer that forms when the anterior capsule incision adheres to the posterior capsule after cataract surgery.

The phenomenon was first described in 1828 by D.W. Soemmering, who wrote of a ring-like substance behind the iris of cadaver eyes that had undergone cataract removal. It is generally only seen when the pupil is displaced or dilated.

Posterior capsular opacification (PCO) occurs about 20% to 50% of the time after cataract surgery. During the process, a wound healing response transforms residual lens epithelial cells to myofibroblasts and is associated with collagen deposition. While we normally associate the term PCO with posterior fibrous opacities, it also encompasses opacities such as Elschnig's pearl and Soemmering's ring.[1]

PCO formation is influenced by type of surgery performed and IOL implanted, history of trauma or aphakia, age of the patient (more aggressive in younger patients) and any comorbidities.[1] This patient's history of lens extraction during infancy surely contributed to the finding.

While PCO that occurs in the line of sight may be treated with Nd:YAG capsulotomy, Soemmering's rings are generally left untouched and are of little consequence. However, there are reports in the literature of pupillary block, corneal decompensation and uveitis-glaucoma-hyphema (UGH) syndrome secondary to dislocated rings. In these cases, iridotomy or surgical removal of the ring is necessary.

Our patient is followed every six months and remains stable, with average intraocular pressures and no evidence of glaucoma, inflammation or corneal disease. RCCL

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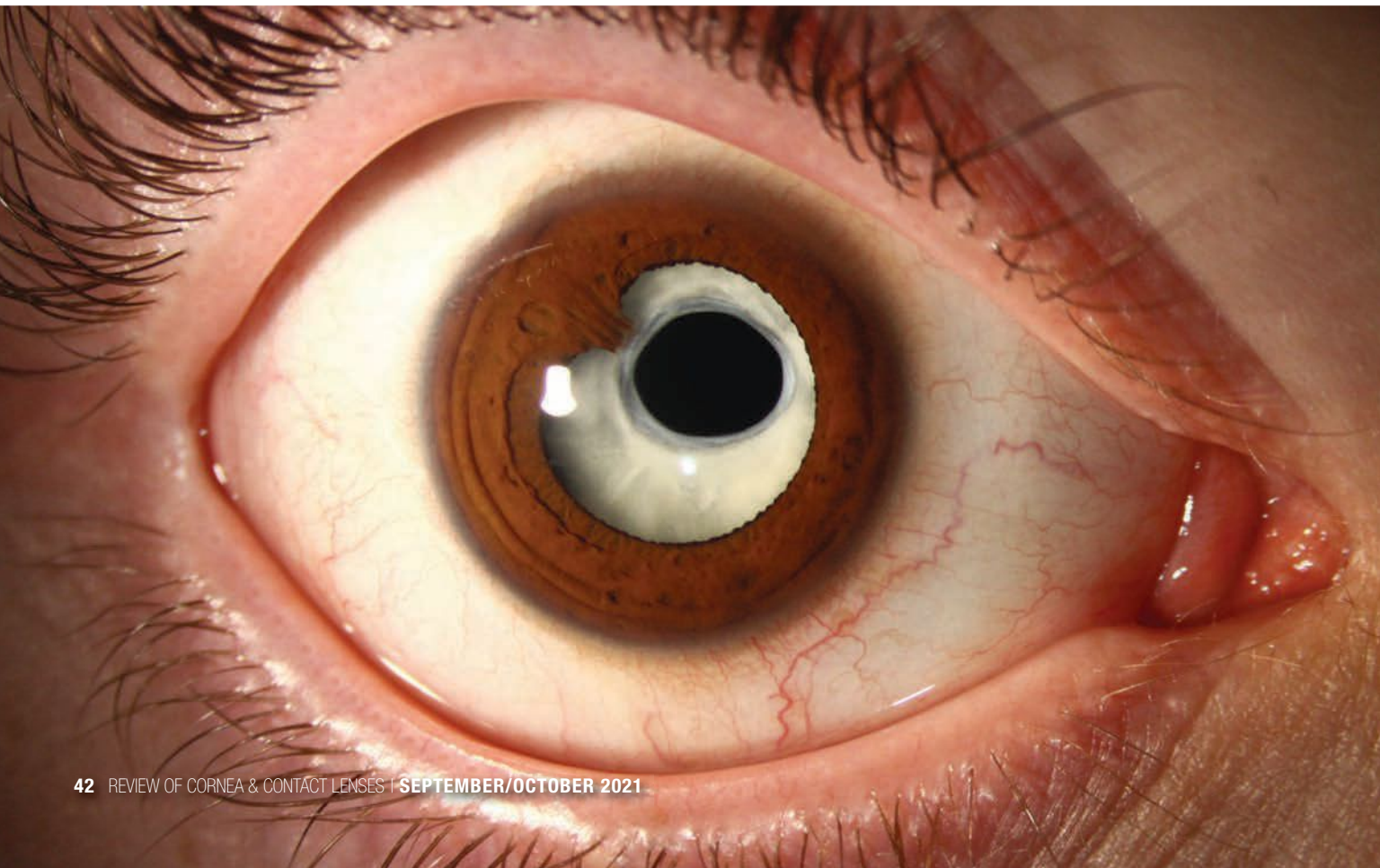

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