Article Date: 2/1/2014

UNIQUE SCLERAL APPLICATIONS

Scleral Lens Applications in Unique Populations

In addition to correcting vision, scleral lenses can replace the function of a compromised ocular surface.

BY KAREN G. CARRASQUILLO, OD, PHD, FAAO, & MELISSA BARNETT, OD, FAAO

Dr. Carrasquillo is director of Clinical Care at the Boston Foundation for Sight, an adjunct clinical professor at the New England College of Optometry, and an advisory board member of the Gas Permeable Lens Institute. She has also authored various publications in her field.

Dr. Barnett is a principal optometrist at the UC Davis Medical Center, where she specializes in ocular surface disease and specialty contact lenses. She is on the Board of the Scleral Lens Educational Society and is an advisory board member of the Gas Permeable Lens Institute. She is an advisor to Alcon and Allergan, has worked as a consultant to Alden Optical, has received honoraria from Acculens and Essilor, and has performed research for Nidek.

Scleral contact lenses represent an area of major growth in the GP lens industry. More and more practitioners are realizing the benefits that these larger-diameter devices have to offer.

By definition, a scleral lens is one whose bearing is supported entirely by the sclera and does not contact the cornea. With a fluid-filled reservoir and choice of GP materials, these lenses allow the cornea to be submerged in an artificial pool of oxygenated tears. These scleral lens design features play an important role in managing refractive errors, including irregular astigmatism, and in managing diseases that affect the ocular surface ecosystem.

Protecting the Ocular Surface

The ocular surface ecosystem includes all of the anterior structures of the eye: the cornea, conjunctiva, and the lacrimal and accessory glands. A stable and healthy ocular surface ecosystem requires good eyelid function, a healthy tear film, and a balance of growth factors and nutrients to enable and promote healing and sustainability (Noble et al, 2004; Quinto et al, 2008). Ocular surface function is altered when this balancing act is compromised.

Severe ocular surface disease can lead to severe vascularization, conjunctivalization of the corneal epithelium, and/or opacification and scarring of the cornea, which can all impact

vision. The effects of ocular surface disease could also result in chronic dry eye—with resulting debilitating pain and photophobia—greatly affecting patients' quality of life.

Scleral lenses have demonstrated an ability to play a significant role in managing ocular surface diseases, primarily because they serve as prosthetic devices, replacing the function of a damaged ocular surface; a prosthetic device replaces function, whereas a prosthesis replaces a body part (Cressey et al, 2012; Rosenthal and Croteau, 2005; Schornack, 2011; Schornack and Baratz, 2009). Therefore, when using these devices in the management of complex corneal diseases, clinicians are really offering patients prosthetic replacement of the ocular surface ecosystem (PROSE) treatment.

Because of this prosthetic device function, scleral lenses have great applications in most realms of ocular surface disease. We will focus on the application of these lenses in cases involving pediatric patients, aphakia, glaucoma, and autoimmune conditions such as rheumatoid arthritis. We will also review options in handling these devices.

Scleral Lenses for Pediatric Patients

Damage to the ocular surface can be devastating in any age group, but one group that warrants special attention is the pediatric population. In recalcitrant cases, and especially when it has been reported that graft survival in infants (<5 years) is poor at best (Lowe at al, 2011), providing adequate support to the ocular surface in this population is essential not just to prevent the risk of amblyopia, but also to prevent visual loss in more aggressive cases.
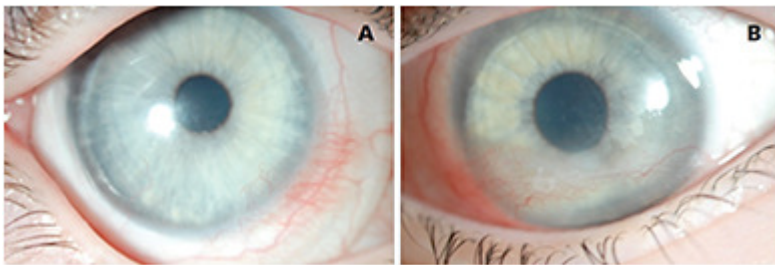

Figure 1. BostonSight devices OD (A) and OS (B) on the patient in Case 1 and the respective sequela of chronic exposure/neovascularization in each eye.

Case 1: Chronic Exposure A 10-year-old Caucasian male had a history of congenital fibrosis of the extraocular muscles, multiple strabismic surgeries for a large angle exotropia, entropion status post-marginal lid rotation, and chronic corneal exposure in the left eye greater than the right. A corneal specialist performed the original examination in 2008; the patient was referred back to the corneal specialist in 2012 for progressive corneal neovascularization and scarring of the left eye. Medically, he had a history of epilepsy, for which he was taking 10mL levetiracetam b.i.d. Otherwise, past medical history was stable. Past ocular medications included artificial tears q.i.d. OD and OS and erythromycin ointment q.h.s. OD and OS.

Visual acuity without correction was 20/60⁻ OD and 20/50⁻ OS. Examination revealed meibomian gland dysfunction as well as lagophthalmos in the left eye, even with forced blink. There was no bells phenomenon noted. The right cornea exhibited mild neovascularization and punctate erosions (Oxford Grade I). The left eye had extensive neovascularization, with a very irregular corneal surface along the exposure zone and some trichiatic lashes on the left lower lid. Associated corneal punctate keratopathy (Oxford Grade III) was noted in the left eye. There was no frank epithelial defect on either eye. The anterior chambers were deep and quiet, with normal iris and lens structures.

The patient was referred for BostonSight PROSE treatment (Boston Foundation for Sight) to support the very fragile ocular surface and to reduce the impetus for further progressive neovascularization from chronic exposure. **Figure 1** shows BostonSight devices fitted over the right and left eyes and the respective sequela of chronic exposure/neovascularization in each eye. For this young patient, the diameter of the BostonSight devices was 17.5mm,

with a bitoric haptic alignment and a total sagittal depth of 460µm. After the fitting of these devices, best corrected visual acuity improved to 20/25^{-2} OD and 20/30^{-} OS. The goals for fitting were to support and protect the ocular surface from chronic exposure and from the effects of entropion, but this young patient also reported improved comfort.

Case 2: Familial Dysautonomia A 10-year-old Jewish male (back in 2009) had a history of neurotrophic keratopathy in both eyes and recalcitrant, persistent epithelial defects OD resulting from familial dysautonomia. Past ocular treatments included partial tarsorrhaphies and Refresh Lacri-Lube ointment (Allergan) q.h. OD and OS. Systemic medications included Zantac and Carafate.

The patient was referred for BostonSight PROSE treatment in 2009 to provide adequate ocular surface support and to prevent further epithelial surface breakdowns. Upon consultation, uncorrected visual acuities were 20/40^{+} OD and 20/40^{+2} OS. Anterior segment evaluation showed no geographical defect on the right eye, but superficial keratitis and punctate keratopathy in both eyes (Oxford Grade III OD and Oxford Grade II OS). There was corneal haze, opacity, and neovascularization just below the visual axis in the right eye. The left eye showed no evidence of haze, opacity, or neovascularization. Anterior chamber, iris, and lens structures were normal in both eyes.

The patient was fitted with 17.5mm-diameter devices with bitoric haptic alignment and a total sagittal depth of 470µm. He is now 13 years old, and three years after initiating PROSE treatment, his best corrected vision is 20/25 in both eyes, with an intact corneal epithelium ever since. Moreover, the prosthetic device function that these lenses have, and that practitioners can offer to patients who have ocular surface disease, is worth noting. **Figure 2** shows the patient's right cornea in 2009 immediately after sequela of persistent epithelial defects, showing opacity, and the same eye with a BostonSight device three years after initiating treatment in which the opacity has significantly faded.
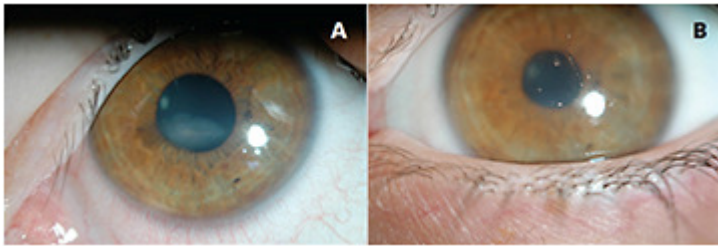

Figure 2. The Case 2 patient's right corneal opacity immediately after sequela of persistent epithelial defects (A) and with a BostonSight device three years after adequate support of the ocular surface in which the opacity has significantly faded (B).

The effect of corneal remodeling without the use of topical steroids or further surgical interventions has also been noted and reported as a result of using these devices in cases that had different disease etiology (Cressey et al, 2012).

It is worth noting that the effects/sequela of neurotrophic keratopathy in familial dysautonomia are similar to those noted in other congenital conditions associated with neurotrophic keratopathy, which we have also treated in our clinic (i.e., Hereditary Sensory Autonomic Neuropathy [HSAN] and Goldenhar Syndrome [Vaughn et al, 2004)], to name a few).

Scleral Lenses and Autoimmune Conditions

Autoimmune conditions can also have a devastating effect on the ocular surface. It has been well documented that dry eye is an ocular surface inflammatory disease, and a mechanism for inflammatory response has been proposed (Stern et al, 2013). Scleral lenses can serve as prosthetic devices, replacing the ocular system functions in these conditions, thereby providing dry eye symptom relief and stabilizing the ocular surface.

Case 3: Dry Eye Syndrome and Corneal Melt Secondary to Rheumatoid Arthritis (RA) A 70-year-old Caucasian male (back in August 2010), managed by a corneal specialist, had a history of an acute ulcer, hypopyon, and cultures positive for *Streptococcus pneumoniae* and coagulase-positive *Staphylococcus* in the right eye. He was developing corneal thinning along the corneal-scleral limbus, and a concern existed that the corneal thinning was associated with RA (RA melt). This patient had not been diagnosed with RA previously, but described joint pain and appeared to have significant ulnar deviation and significant enlargement at the proximal anterior phalangeal joints. A rheumatologist evaluated the patient to confirm or rule out RA as an underlying etiology, as that would determine the course of systemic medications. In the meantime, the corneal specialist started him on fortified Iquix (Santen) drops q.h., right eye only.

RA was confirmed. In September 2010, the thinning had progressed, and a corneal melt associated with RA was diagnosed. Following treatment with high-dose oral steroids and methotrexate, the corneal thinning stabilized. Ocular medications then included fluoromethalone ointment q.i.d. OD and q.d. OS.

Subsequent examination by the corneal specialist revealed uncorrected visual acuity of 20/200 OD and 20/30 OS, with stable corneal thinning and an intact epithelium. The plan was to discontinue fluoromethalone ointment and start Lotemax (Bausch + Lomb [B+L]) drops b.i.d. OD and q.d. OS. The patient complained of dry eye symptoms, in the right eye greater than the left, and poor vision due to an irregular cornea and opacity in the right eye. He was referred for corneal GP fitting OD to improve vision, but was intolerant due to dry eye symptoms.

The patient was then referred for BostonSight PROSE treatment in 2011. The goal for fitting was to improve vision, reduce dry eye symptoms, and support the ocular surface. Although both eyes showed sequela of dry eyes from RA, the patient was symptomatic only in the right eye. **Figure 3**A shows corneal opacity and neovascularization, **Figure 3**B shows pooling over the thinned area and punctate keratopathy, **Figure 3**C shows an area of ~70% stromal thinning associated with corneal melt, and **Figure 3**D shows ~40% stromal thinning inferior nasally OD.
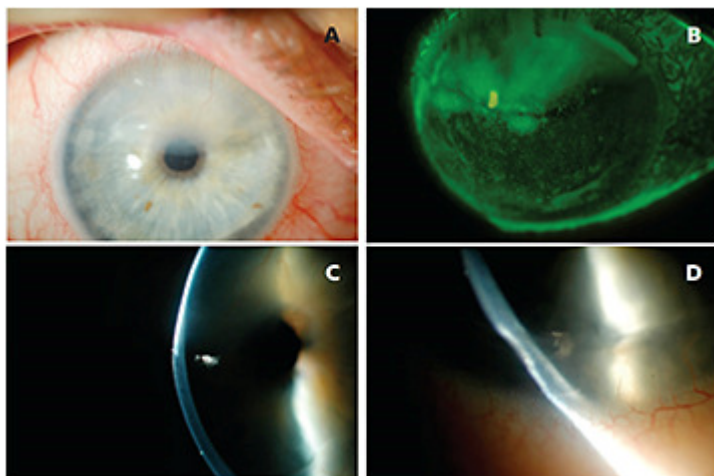

Figure 3. Corneal opacity and neovascularization in patient from Case 3 (A), pooling over the thinned area and punctate keratopathy (B), area of ~70% stromal thinning associated with corneal melt (C), and ~40% stromal thinning inferior nasally OD (D).

The patient was fitted with a 19.0mm-diameter device with quad-toric haptic alignment and total sagittal depth of 480µm. **Figure 4** shows adequate vaulting over the thinned cornea. Best-corrected vision was 20/30[+2] OD. The patient also reported a significant reduction in dry eye symptoms and was able to wear the device for an average of 15 hours each day. After two years of adequate ocular surface support, together with adequate medical management for his RA, there has been no further epithelial breakdown, and the patient's quality of life has significantly improved.
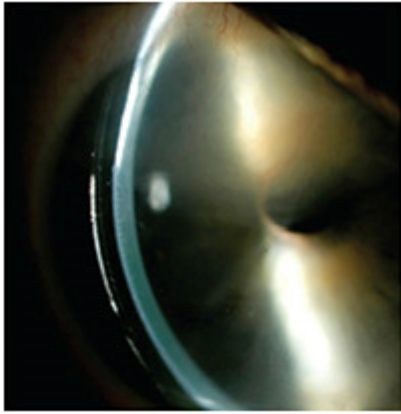
Figure 4. Adequate vaulting of a 19.0mm-diameter lens with a quad-toric haptic alignment over the thinned cornea of the patient in Case 3.

Scleral Lenses for Glaucoma

Scleral lenses are also a fantastic option for patients who have irregular corneas and glaucoma, and even more so if ocular surface disease compromises the ocular surface in such patients.

A history of glaucoma surgery, including trabeculectomy, shunt, stent, or glaucoma implant, may complicate the fitting of scleral lenses due to the resulting irregular conjunctival surface. The conjunctiva may be elevated or uneven in the area in which the glaucoma surgery was performed. Also, excessive pressure or rubbing over tube shunts or valves may compromise intraocular pressure and lead to conjunctival and/or tube erosion, which can increase the risk of further complications (i.e., endophthalmitis). A notch in the scleral lens can be created to avoid both pressure on the conjunctiva and contact with the surgical area.

Case 4: Dry Eye Syndrome and Primary Open Angle Glaucoma Status Post-Baerveldt Implant A 58-year-old Hispanic female was referred for a contact lens examination. She was experiencing irritated eyes with her current hybrid contact lens and had reverted back to a soft lens for the left eye. She complained of poor vision, especially at distance when driving at night. She also reported double vision when reclining, but not in straightahead gaze.

The patient's medical history was significant for diabetes, hypertension, hypothyroidism, and sleep apnea. Her ocular history was significant for dry eye in both eyes. The right eye had a stable intraocular lens, the left eye had a cataract. Primary open angle glaucoma was present in both eyes. Of note, she underwent a Baerveldt glaucoma implant six months prior to the examination.

Following the glaucoma implant, she developed a right hypertropia and alternating exotropia. This is not uncommon, as persistent restrictive strabismus may occur with glaucoma drainage implants due to scarring between the rectus and oblique muscles (Schwartz et al, 2006).

Systemic medications included insulin, metformin, glimepiride, lisinopril, hydrochlorothiazide, levothyroxine, and escitalopram. Ocular medications were Alphagan (Allergan) and Cosopt (Merck) b.i.d. OD. Entering vision was 20/50^{+2} OD without correction (sc) and 20/50^{-2} OS (with soft lens).

Anterior segment examination revealed a stable glaucoma drainage device implant located superior temporal in the right eye with a bleb over the plate. The tube was well covered and visible in the anterior chamber. Both eyes exhibited corneal staining (1+ inferior punctate epithelial keratopathy). The posterior chamber intraocular lens was stable in the right eye. Mild nuclear and cortical sclerosis was present in the left eye. Intraocular pressures were OD 25mmHg and OS 21mmHg at 1:57pm. Optic nerve examination revealed vertical elongation of the disc with peripapillary atrophy. Myopic degeneration was present in both eyes.

Recommended treatment included nonpreserved artificial tears, frequent breaks when reading and using a computer, good water intake, and daily omega-3 fatty acid intake. Due to elevated intraocular pressure in the right eye, an appointment was scheduled with the glaucoma surgeon.

Medical management and options were discussed, and the patient was fitted with Maxim Scleral (Acculens) lenses in Boston XO_2 material (B+L). Lens parameters were OD 46.00D base curve (BC), 15.0mm overall diameter (OAD), 8.00mm optical zone diameter (OZD), +0.50D power, sag 4.35, 4mm notch (to insert superior temporal) and OS 46.00D BC, 15.4mm OAD, 8.0mm OZD, −13.00D power, sag 4.46 (intermediate/near). The right lens was targeted for distance, the left lens for intermediate/near to eliminate diplopia. A superior temporal notch was made in the right scleral lens (**Figure 5**).

Visual acuity at distance was 20/30 OD, 20/30^{+2} OS, 20/25^{-2} OU, and the patient had good computer and near vision, with J1+ OS and J1+ OD at near. The patient reported incredible comfort with the scleral lenses. Both lenses fit well with good central apical clearance and good peripheral alignment. No blanching was present in either eye. The scleral lens notch was correctly positioned superior temporal in the right eye and did not touch the glaucoma implant. The patient was able to wear the devices for 15 hours each day. Intraocular pressures checked multiple times during a three-month period ranged from 16mmHg to 18mmHg in each eye.
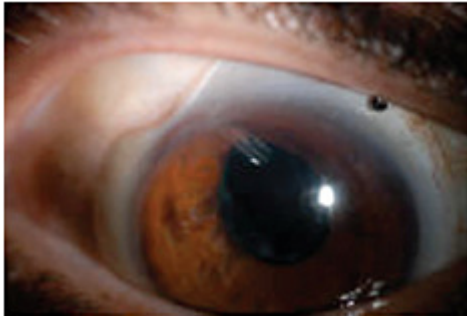

Figure 5. A superior temporal notch in the right scleral lens of the glaucoma patient in Case 4.

With scleral lenses, it is possible to fit inside of conjunctival abnormalities by decreasing the lens diameter or to fit over abnormalities by increasing the lens diameter. Alternatively, as we did in this case, it is possible to go around the abnormality by putting a notch in the lens. In cases of glaucoma surgeries or implants, it may be beneficial to avoid the abnormality altogether and to create a notch in the scleral lens. Scleral lens notches are also advantageous when there are other types of conjunctival abnormality such as an elevated pinguecula or conjunctival cyst.

Putting a notch in a scleral lens may sound complicated, but it is not. The first step is to measure the size (both height and width) of the conjunctival abnormality using a slit beam. Next, measure the height and width of the conjunctival abnormality while the scleral lens is on the eye. Then, mark the scleral lens with a permanent (e.g., Sharpie) or dry erase marker while the lens is on the eye. Next, measure the tracing on the lens after removing it from the eye. Finally, call the laboratory consultant to discuss the plan and send the lens to the laboratory.

When applying the lens, it is important to place it on the eye with the correct orientation. Make sure to inform the staff person who is training the patient on scleral lens application and removal, as well as the patient, about the need for proper lens orientation.

Scleral Lenses for Aphakia

Scleral lenses are an excellent option for patients who have aphakia, especially in the case of a concomitant irregular cornea or intolerance to corneal GP lenses.

Case 5: Aphakia with Corneal GP Intolerance A 70-year-old Caucasian female presented for a contact lens examination complaining that her contact lens tended to slide off of her right eye.
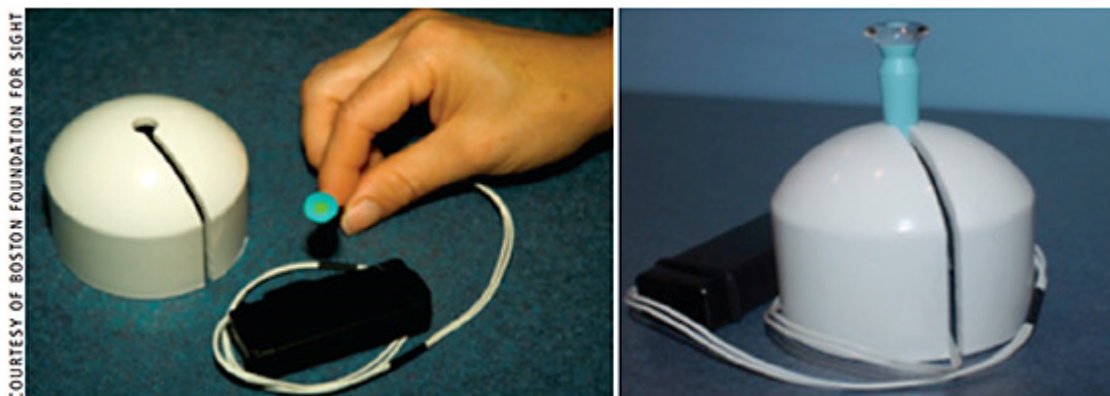

Figure 6. See Green Lens Inserter.

Her medical history was significant for hypothyroidism, osteopenia, and vitamin D deficiency. Her ocular history was significant for aphakia in the right eye. The left eye ocular history included status post-penetrating keratoplasty and glaucoma status post-shunt, cystoid macular edema, anterior chamber intraocular lens, and an epiretinal membrane. Systemic medications included lisinopril and levothyroxine, and ocular medications included prednisolone acetate 1% q.d. OS.

Entering vision was 20/25 OD (GP) and count fingers at four feet OS (sc). Both eyes had irregular astigmatism. Anterior segment examination revealed meibomian gland dysfunction in both eyes as well as a stable glaucoma drainage device implant located superior temporal in the left eye. The cornea of the right eye was clear. The penetrating keratoplasty of the left eye was stable. A peripheral iridotomy was stable in both eyes. The right eye was aphakic. There was an anterior chamber intraocular lens in the left eye.

Figure 7. EZi Scleral Lens Applicator.

Recommended treatment included good eyelid hygiene, including lid scrubs and warm compresses with diluted baby shampoo or commercially available eyelid scrubs. We discussed medical management options and fit the patient with a Jupiter scleral lens (Essilor, Visionary Optics) manufactured in Optimum Extra (Contamac) material. Lens parameters were 42.50D BC, 16.6mm OAD, 8.14mm OZD, +15.50D power, and center thickness of 0.49mm for the right eye. She deferred lens fit for the left eye.

The patient reported good distance vision (20/20) and amazing comfort with the scleral lens. The lens fit well with good central apical clearance and a good peripheral alignment. No blanching was present. Wearing time was 14 hours per day. At the follow-up appointment, the right eye exhibited temporal microcystic edema. The chamber of the scleral lens was increased to 14.4mm, and the microcystic edema resolved.

Scleral Lens Handling Options

Sometimes it is difficult for a monocular patient who has aphakia to see the scleral device for application, or for a patient who has dexterity issues to successfully apply the devices.

There are several tools that are beneficial for such patients. One is the See Green Lens Inserter by Dalsey Adaptives (**http://dalseyadaptives.net/store/**), which is available with and without a stand (**Figure 6**). The lighted plunger helps center the device for application. The stand holds the plunger and lens in place prior to application. This is helpful for patients who have unsteady hands or for those who need both hands to hold their eyelids open.

Another tool is the EZi Scleral Lens Applicator by Q-Case Inc. (**http://ezibyqcase.com/**) (**Figure 7**). This device is placed on the finger like a ring and has a base for scleral lens application. This design provides stability and allows patients to apply scleral devices with one finger.

A third option is a #8 O ring that is available at any hardware store. The scleral lens device rests on the O ring on a patient's finger, which can allow for stable application.

Conclusions

More and more clinicians are appreciating the benefits of scleral lenses and their unique applications in the management of refractive errors associated with irregular corneas, but also in the management of ocular surface disease. It has been shown that these lenses serve a prosthetic device function, replacing the function of compromised ocular surfaces. PROSE treatment can be achieved with any large-diameter lens that bears its weight entirely on the sclera and does not contact the cornea. **CLS**

To obtain references for this article, please visit
http://www.clspectrum.com/references.asp *and click on document #219.*

Contact Lens Spectrum, Volume: 29 , Issue: February 2014