Clinical & Refractive Optometry CRO
ONLINE CE A CE JOURNAL FOR OPTOMETRY

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Beneath the Shell: Managing Ocular Surface Health of a No Light Perception Eye with a Prosthetic Shell *CRO Journal*. Published online June 2025.
doi: 10.57204/001c.140574

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Beneath the Shell: Managing Ocular Surface Health of a No Light Perception Eye with a Prosthetic Shell

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CRO (Clinical & Refractive Optometry) Journal

ABSTRACT

Patients with a non-seeing eye and an intact globe may experience secondary psychological trauma due to cosmetic concerns as well as potential social ramifications. A prosthetic scleral shell fit over the intact, scarred globe is an important option to consider for improving cosmesis. Once a prosthetic shell has been successfully fit by an ocularist, the patient should still be monitored regularly as they are at risk for complications that could threaten the globe. Herein, we present a case of a patient with a scarred, non-seeing eye fit with a prosthetic shell. We will discuss the design and fabrication, proper fitting, and potential complications of prosthetic scleral shell wear.

CASE REPORT

A 65-year-old African American male presented with complaints of dull pain and light sensitivity in his right eye. The dull pain was present occasionally and he reported constant photophobia. His ocular history was remarkable for blunt force trauma to the right eye three finalyears prior resulting in no light perception (NLP) in this eye. He reported that he had been stabbed by a knife in the right eye and had undergone two reparative surgeries following the trauma. The exact procedures performed were unknown. His medical history was remarkable for hypertension, hyperlipidemia, cardiomyopathy, and seizure disorders.

Best-corrected visual acuities were NLP in the right eye and 20/20 in the left eye. Extraocular motilities revealed limited adduction of the right eye and no restrictions of the left eye. Cover test confirmed a large angle constant right exotropia. Confrontation visual fields were unable to be assessed in the right eye and were full to finger count in the left eye. The pupil reaction of the right eye was unable to be assessed due to scarring of the cornea and the left eye was noted to have normal pupil reaction with no reverse afferent pupillary defect (APD). Intraocular pressures were measured by Goldmann tonometry at 44 mmHg in the right eye and 8 mmHg in the left eye. Anterior segment evaluation revealed an intact right globe with diffuse corneal opacification and areas of corneal neovascularization while findings in the left eye were unremarkable. The posterior segment of the right eye was unable to be assessed due to the opacified cornea, while dilated fundus examination of the left eye revealed a healthy optic nerve, macula, and peripheral retina.

Findings of the left eye continued to be unremarkable at subsequent exams, and therefore the remainder of the discussion will concern the right eye only. Over the next four years, the patient presented with recurrent episodes of irritation in the right eye. It was noted at these visits that the eye was mildly proptotic with lower lid ectropion and an incomplete blink. The anterior chamber was shallow, and there was notable iris scar tissue as well as a mature cataract present. Diffuse superficial punctate keratitis and ruptured corneal bullae were noted at subsequent visits. This was addressed with lubricating ophthalmic drops and ointment. The patient was then lost to follow-up over the next two years. Upon his next presentation, the right cornea was noted to have diffuse opacification, neovascularization, stromal haze, and yellow keratinization. The patient had become very concerned about the poor cosmesis of his right eyeHe expressed interest in taking steps to improve the cosmesis of the eye, including enucleation followed by a prosthetic eye. The patient was then referred to an ocularist for a prosthetic evaluation.

The ocularist noted that the patient had a large globe and that she was concerned about retention of the shell as well as the possibility of corneal damage from the shell. She planned to design a scleral shell that would vault over the cornea. This shell would initially remain clear to determine the fit and allow time for adaptation with insertion and removal of the prosthetic. The ocularist reported that the patient demonstrated appropriate technique with insertion and removal of the scleral shell and that he was highly motivated for success in wearing the prosthetic.

The patient presented for a follow-up exam in our clinic having worn the clear scleral shell for eight hours per day on average. He reported no pain but did state he had minor irritation and dryness. He also reported no complications with insertion and removal of the shell. Anterior segment evaluation revealed 2+ diffuse conjunctival injection along with stable corneal findings and no signs of corneal or conjunctival abrasions. The scleral shell showed good central clearance with some edge lift and bubbles at the nasal and temporal edges (Figure 1). Anterior segment OCT measured central corneal thickness to be 600 microns with approximately 180 microns of clearance between the corneal surface and the prosthetic (Figure 2).

Figure 1. Anterior segment photography shows corneal scarring, opacification, keratinization, and neovascularization of the right eye underneath the clear scleral shell

Preservative-free artificial tears were prescribed three times a day before, during, and after use of the prosthetic device. After a few weeks of successful wear with the clear prosthetic shell, the ocularist finalized and painted the shell (Figures 3 & 4). The patient was very pleased with the cosmesis of his new prosthetic and reported comfortable wear for roughly 8-10 hours a day. We plan to have the patient return to our clinic regularly to monitor the ocular surface health underneath the final prosthetic scleral shell.

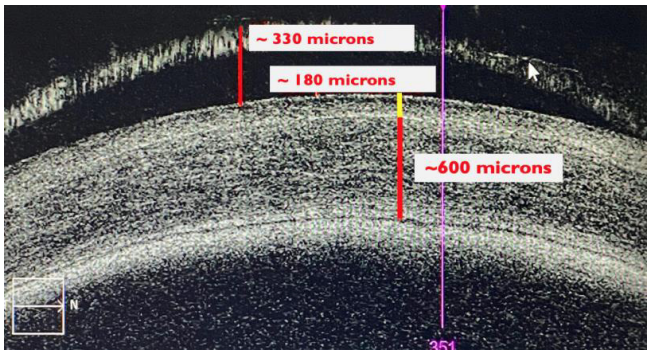
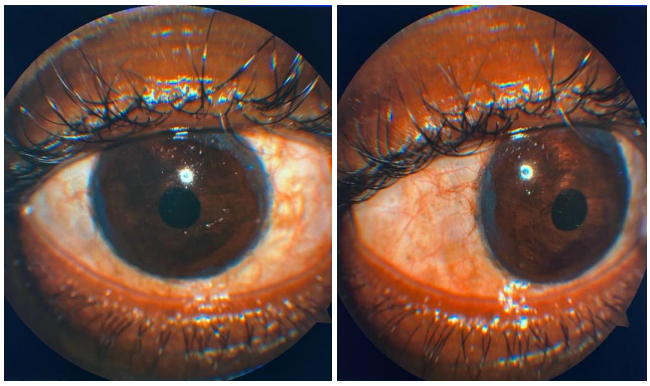


Figure 2. Anterior segment OCT of the right eye shows central corneal thickness as well as clearance between the corneal surface and the prosthetic shell



Figures 3 & 4. Anterior segment photography shows the final, painted prosthetic scleral shell

DISCUSSION

For individuals with blind and disfigured eyes secondary to trauma, concern for ocular surface health as well as cosmesis is important in providing the best quality care for these patients. Education on the importance of regular eye exams along with the various options for improved cosmesis should always be addressed.[5] Once a patient has made the decision to be fit with a prosthetic device, proper education regarding care of the device and maintaining follow-up visits should be discussed as well. The different types of prosthetics, the fitting process, assessment of ocular surface health, and proper care and maintenance of prosthetic devices will be reviewed in the following discussion:

Types of Ocular Prosthetic Devices

For eyes loss of functional vision, there are many options for improved cosmesis including artificial eyes and ocular implants. This section names a few commonly encountered artificial eyes including custom ocular prosthesis, conformers, stock eyes, glass eyes, custom painted contact lenses and scleral shells.

A custom ocular prosthetic eye is a custom painted and impression fit device made with polymethyl methacrylate (PMMA) material. These prostheses are typically made to fit over an implant and can be worn for weeks or months without removal. Conformers are also impression fit and made with PMMA material. These devices are used to expand the socket in the presence of anopthalmos or microopthalmia prior to fitting a prosthetic eye. Stock eyes are also made with PMMA but are not impression fit. These devices can have very poor motility and are to be removed nightly. Glass eyes are no longer commonly used in the United States due to the significant disadvantage of breakability and increased risk of injury to the eye or orbit. Custom painted contact lenses are silicone hydrogel or hydrogel lenses used to mask disfigured eyes and can be made to occlude the iris and/or pupil. A scleral shell is a painted PMMA device made for blind disfigured eyes. These shells fit directly over the patient's own eye and provide excellent cosmesis as well as motility. Scleral shells can also correct volume loss and are beneficial for photophobic patients.[1,5]

Design/Fabrication Process

The process of obtaining a custom ocular prosthetic for a patient begins with the initial consultation with an ocularist. The ocularist will review the overall process, make assessments about different options for prostheses, and will come to a decision with the patient about which device will best meet his or her needs. Next is the impression phase in which the patient's tissue contour is accurately captured to properly fabricate the posterior side of the prosthetic. Once the impression is captured, the modeling/sculpting phase begins. This is followed by the painting phase where the ocularist works on matching the fine details on the prosthetic device to the patient's companion eye before the final fitting.[1,6] Common complications an ocularist may encounter during the fitting and fabrication process include exposed implants, ptosis of the upper lid, laxity of the lower lid, socket contraction, and superior sulcus deformity.[8] Most of these complications require a specialized adaptation of the prosthetic device or surgical intervention.[1]

Proper Prosthetic Shell Fit & Potential Ocular Surface Health Complications

Evaluation of the Scleral Shell Fit

The key to a healthy ocular surface while using a prosthetic shell begins with a properly fitted device. Proper fit of a prosthetic scleral shell should be similar to that of a scleral contact lens. One major difference in assessing the fit for a shell versus a lens is that the shell is not filled with a saline reservoir. The shell is vaulted only using the natural tear film and possibly a few drops of lubricating artificial tears. There is similarity in that both a scleral shell and scleral contact lens should entirely vault the cornea with adequate central clearance. However, in regards to the amount of desired central clearance, the goal slightly differs. With a typical scleral contact lens fit on a relatively normal or healthy eye, the initial goal of central clearance is around 200 to 300 microns. This amount of clearance is expected to settle throughout the day ranging from 50 to 200 microns. If there is minimal clearance or corneal bearing present, there is an issue with the sagittal height of the lens or shell. Corneal bearing in both normal and diseased or NLP eyes is a complication that can cause corneal microcysts, staining, abrasions and epithelial bullae. Excessive clearance in relatively normal eyes can cause reduction of clear vision from formation of air bubbles beneath the lens.[11] It is important to keep in mind that with NLP eyes, scleral prosthetic shells are fit for comfort and cosmesis and not for clear vision. For this reason, excessive clearance resulting in air bubbles in scleral shell fits are not of concern.

Another important factor to consider when assessing overall scleral shell fit is the amount of edge lift. Adequate edge lift will avoid conjunctival blood vessel impingement and allow for proper tear exchange for better comfort and less lens awareness. Other indications that there is not enough edge lift include the presence of conjunctival drag, conjunctival redness, and conjunctival staining. Other factors to assess include rotation and overall retention of the shell on the eye. Both rotation and lack of retention indicate the need for an adjustment to the device or possible lid surgery. Significant problems in the fit of the scleral shell should prompt a referral back to the ocularist for modifications to the prosthetic device.[4,7]

Case History and Evaluation of the Eye

After assessing the scleral shell on the eye it should be removed and evaluated along with the underlying ocular surface as part of routine eye examinations and follow-up visits. In addition to the case history questions asked of all patients, scleral shell wearers should be asked about pain or irritation they may be experiencing, the number of hours the shell is worn daily, and how the patient is caring for the scleral shell. Symptoms such as discharge, excessive mucous, or pain could indicate a serious underlying complication of scleral shell wear. Extreme socket sensitivity may occur with these patients and could indicate an issue with the fit of the prosthetic device or possible infection of the eye. Itching is one of the most frequent but least serious symptoms that typically occurs with scleral shell wear. It is commonly caused by dryness and/or allergies. Itching may also result from giant papillary conjunctivitis (GPC) caused by friction and mechanical irritation of the conjunctiva or socket, especially in the presence of a shell with heavy protein deposits.

Upon removal of the prosthetic, it is important to thoroughly examine the shell for surface deposits, chips, scratches, or cracks.[4] If the device is found to have any irregularities or damage the patient will need to be referred back to the ocularist for possible cleaning, repairs or replacement.

The physical assessment of the eye without the scleral shell begins with evaluating the lids for swelling, abrasions, and keratinization of the lower palpebral conjunctiva from ectropion. Careful assessment for lid infections, such as preseptal cellulitis, should be performed and managed with care to prevent orbital cellulitis. There will be no symptoms of decreased vision or diplopia in these eyes as compared to a seeing eye. One should also evaluate the lashes for mucous or discharge, and the palpebral conjunctiva for papillae or abrasions.[11]

Staining the corneal surface with sodium fluorescein is an important tool to aid in detecting abrasions and keratitis that may correspond to areas of damage or deposits on the prosthetic device. Abrasions may result from improper insertion or removal techniques or from an improperly fitting device. It is also important to look for signs of conjunctivitis which may result from infection, inflammation, or mechanical irritation by the prosthetic device.[4] Other possible complications of scleral shell wear include microbial keratitis, corneal edema, corneal neovascularization, corneal bullae, and corneal ulcers. Corneal ulcers are of particular concern because of the risk for corneal perforation and eventual loss of the globe.

The follow-up frequency for scleral shell wearers depends on the patient's symptoms, the fit and condition of the scleral shell, and the health of the eye and orbit. Evidence of a poor fit and damage to the prosthetic shell require a referral back to the ocularist. Depending on the ocular surface appearance, associated symptoms, care of the prosthetic, and clinical findings, more frequent follow-up examinations may be warranted for patients wearing prosthetic devices. If all ocular health findings are determined to be normal and the patient is compliant with appropriate insertion, removal, and cleaning, and there are no patient complaints, annual examination is the standard.

Care and Maintenance for Prosthetic Scleral Shells

A clean and well maintained ocular prosthetic device is vital to help ensure the best prognosis for a healthy ocular surface long term. Scleral shells should be removed and cleaned every night. A simple, yet effective option, for cleaning the shell is warm water and a gentle soap or baby shampoo. Another option for cleaning the shell is a non-abrasive gas permeable contact lens cleaner followed by a saline rinse. This cleaning regimen should be implemented each night following removal of the prosthetic scleral shell to prevent infection and other complications.[1]

Aside from regular cleaning, scleral shells need to be periodically polished. The frequency depends on how often the patient removes their prosthetic device. If the patient removes and cleans the scleral shell daily as advised, the recommended frequency for polishing the shell is six to twelve months. If the patient does not remove and clean the shell nightly, the recommended frequency is every three months.[1,10]

The ocular surface, lid structure, and surrounding tissues may change over the years leading to an ill fit of the prosthetic. The replacement for scleral prosthetic shells is typically three to five years. In contrast, most forms of artificial eyes require replacements every five to seven years.[1,4] Appropriate patient education on the importance of removing and cleaning their device each night along with good ocular hygiene is vital to the maintenance and longevity of the device.

CONCLUSION

Scleral shells are an excellent option to consider in those patients with non-seeing eyes and intact globes. However, a scarred, non-seeing eye is still at risk for major complications and sequelae, especially when fit with a prosthetic shell. A host of ocular disease conditions and complications of prosthetic shell wear can develop that may lead to discomfort and damage to the ocular surface. Monitoring an NLP eye regularly for development of ocular complications that could result in pain, infection, or even loss of the eye is imperative. Ocular disease and complications should be managed appropriately, and the patient's symptoms should be addressed at each visit. The cleanliness, intactness, and fit of the shell must also be assessed periodically to prolong the life of the device as well as the health of the eye. Loss of an eye is both a physical and psychological traumatic experience. It is important to remember that as eyecare providers we play a crucial role in being able to offer options for improved cosmesis to individuals with non-seeing and disfigured eyes.

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