



Scleral lens for ocular surface disease

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ABSTRACT

Scleral rigid gas-permeable (RGP) lenses play an important and underappreciated role in the treatment of ocular surface disease. This review provides an update on scleral rigid gas-permeable lenses in the therapy of ocular surface disease (OSD). Several large cohort studies and case series have been published since 2008 that report on the use of scleral RGP lenses in the treatment of ocular surface disease as a sub-group of corneal disease, or in specific diseases such as Stevens-Johnson syndrome and ocular chronic graft-versus-host disease. In addition, there are case reports detailing the treatment with scleral lenses in OSD of specific etiology such as herpes zoster, limbal stem cell deficiency or ocular cicatricial pemphigoid. Two case series were published showing innovative utility of scleral RGP lenses in the management of OSD after glaucoma surgery and for drug delivery for corneal neovascularization. Reports continue to emerge on the impact of scleral RGP lenses in the treatment of patients with ocular surface disease. Advances in lens design and fitting techniques make scleral RGP lenses a practical option for a rising number and variety of patients with corneal disease. Clinicians who treat patients with ocular surface disease should be aware of scleral RGP lenses as a method to treat OSD.

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1. Introduction

Corneal disease, in particular ocular surface disease (OSD), is a well studied topic in scientific literature. One important and underappreciated modality to treat OSD is the use of large diameter, or scleral rigid gas-permeable (RGP) lenses. Recently, there have been a number of publications that pertain to the management of corneal and ocular surface disease with scleral RGP lenses.

History of scleral lenses and principles of design

The first description of scleral lenses in the medical literature was provided in a report on contact lenses by Adolf Eugen Fick in 1888 (1997). In a review of the history of contact lenses, Pearson (2007) reports Karl Otto Himmler as the first manufacturer of contact lenses. He also describes reports of contact lenses ranging in diameter from 15 to 22 mm in 1888, 1888, and 1889 by Fick, Kalt, and Muller, respectively, all made of glass. Pearson's report details the evolution of design and manufacture of current contact lenses, all of which are scleral or larger than 17 mm in size. Examining these early glass and then polymethyl methacrylate (PMMA) lens

designs retrospectively exposes reasons for failure as inadequate limbal clearance, problems with back surface junctions, and hypoxia related to the material.

In the 1980's, the advance of lenses manufactured with rigid gas permeable material (Ezekiel, 1983) provided a solution to the hypoxia created by earlier scleral lens designs. Throughout the decade, this design was improved by multiple centers around the world (Lyons et al., 1989; Schein et al., 1990; Visser, 1990). With material of sufficiently high Dk values serving to solve the difficulty of hypoxia, the issue of suction had to be resolved. Initially, suction was overcome by fenestration of the lens allowing for additional oxygen availability whether the device was made of PMMA or gas permeable plastic. Another method of reducing suction and increasing tear exchange can be achieved by creating grooves in the back surface of the lens. This technique was developed in the 1960's with PMMA and has been used in gas-permeable lenses (Romero-Rangel et al., 2000; Pullum and Buckley, 2007) (Fig. 1). Fenestration introduces air bubbles into the lens reservoir, which may improve oxygen availability for corneas with marginal endothelial function, but may

not be well tolerated by patients suffering from OSD (Jacobs, 2008).

Features of scleral rigid gas-permeable lenses

Current scleral RGP lenses are large diameter lenses (17.5-25mm) that completely vault the corneal surface. The bearing portion of the lens, called the haptic, is supported completely by the sclera and does not contact the cornea. The central, optic portion with its artificial tear filled reservoir, corrects front surface refractive error, including irregular astigmatism, and power can be added to correct axial refractive error (Fig. 2). Made from various polymers, all with high Dk values to promote oxygen permeability, scleral RGP lenses allow for the cornea to be submerged in an artificial pool of oxygenated tears. These unique design features provide an important role for these lenses in the management of ocular surface disease and refractive errors including irregular astigmatism.

Scleral rigid gas-permeable lenses for ocular surface disease

Since 2008, there have been twelve publications documenting utility of fluid-ventilated scleral RGP lenses for ocular surface disease. Three of these are large cohort studies that include analyses on OSD patient subgroups (Severinsky and Millodot, 2010; Stason et al., 2010; Gumus et al., 2011), while three more reports examined the effect of scleral RGP lenses in populations with one specific surface disease (Schornack et al., 2008; Tougeron-Brousseau et al., 2009; Rathi et al., 2011). Also included in the twelve publications are four case reports or series of single disease entities reporting outcome measures demonstrating the effectiveness of scleral RGP lenses (Schornack and Baratz, 2009; Grover et al., 2010; Gumus et al., 2010; Schornack, 2011), and two case series describing innovative uses of scleral lenses in patients with other ocular complications secondary to OSD (Lim et al., 2009; Tanhehco and Jacobs, 2010).

Scleral rigid gas-permeable lenses for general corneal and ocular surface disease

In a report from 2010 (Stason et al., 2010), 80 consecutive patients seen at the Boston Foundation for Sight (Needham, MA) from January to June 2006 are reviewed. The full spectrum of corneal disease, including OSD, is discussed, but nearly half, 38 of 80 patients, were treated for OSD. Those patients diagnosed with OSD are further classified by diagnosis into the categories dry eye syndrome (DES), chronic graft-versus-host disease (GvHD), and other. Mean visual acuity in the OSD cohort improved by -0.22 (logMAR). The authors report an increase in mean National Eye Institute (NEI) visual functioning (VFQ-25) scores for the entire cohort from 57.0 to 77.8. Findings were similar for surface disease group, with only two patients recording a decrease in visual function questionnaire (VFQ) score in this group. Greater benefits in the VFQ subcategories of eye pain, photophobia, and role difficulties were seen in the OSD group.

In another study published in 2010, Severinsky and Millodot (2010), from Spain, report on their experience with gas-permeable scleral lenses. All lenses had an overall diameter of 18.5 mm with an average optical zone of 12.5 mm. Over a six year period from January 2003 to December 2008, 97 consecutive patients were fitted. In contrast to the population reported in the United States report (Stason et al., 2010), near-

ly all patients fitted had either ectatic corneas or corneal irregularities following penetrating keratoplasty, only 6/97 patients had OSD, including Stevens-Johnson syndrome (SJS), GvHD, and exposure keratopathy. Criteria for fitting success was determined by the investigators to be daily wear time greater than 10 hours with no lens adhesion on removal, and partial success was considered achieved with daily wear time between 6 and 10 hours. The OSD group had a mean wear time of 9.3 hours with a range of 5-14 hours. Mean visual acuity for the OSD group was 20/50. Failure was reported in 21% of patients overall, with no breakdown by disease subgroup; it is unclear how many OSD subjects failed fitting. The authors in this report, as well as in the Stason study, conclude that those eyes with ectasia or astigmatism have increased visual acuity improvement over those eyes with a surface disease because improvement in visual acuity is not the main therapeutic indication for OSD, and because baseline visual acuity is generally higher in OSD patients.

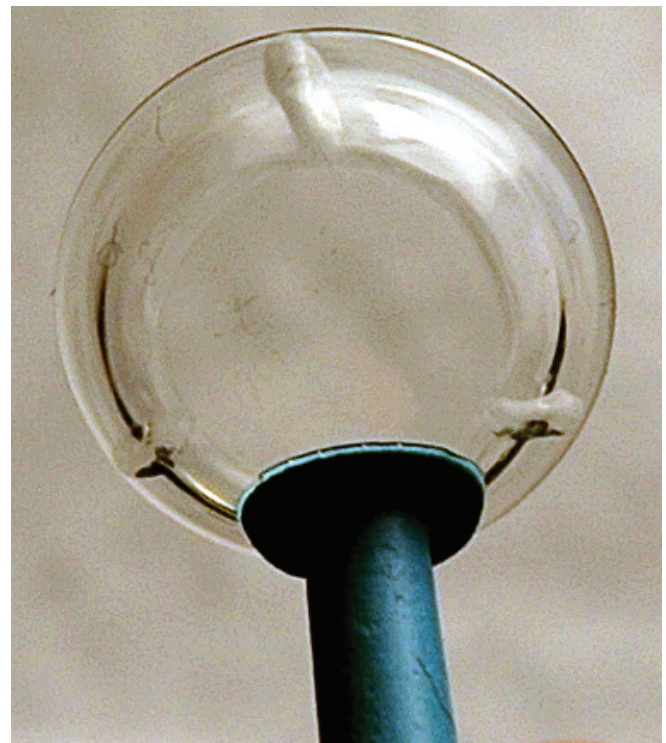


Fig. 1. Scleral rigid gas-permeable lens shown with grooves on the back surface to reduce suction.

Although the primary indication for scleral lenses in OSD is for comfort and support of the ocular surface, as opposed to improvement of visual acuity, investigators from Cullen Eye Institute, Baylor College of Medicine (Houston, TX) (Gumus et al., 2011) report on the impact of the Boston Ocular Surface Prosthesis (BOSP) (Boston Foundation for Sight, Needham, MA) on higher-order wavefront aberrations on their patients. Their series of 39 patients, 56 eyes, was broken down into 4 groups, with 18 eyes being categorized as having surface disease, including Sjögren's syndrome, neurotrophic corneal scarring, postherpetic corneal scarring, and dysfunctional tear syndrome. During the fitting process, optical eccentricity was added to the front surface of the lenses. If vision improved, then varying amounts of eccentricity were attempted (0.3, 0.6, 0.8) until vision no longer continued to improve. In the OSD subgroup, the investigators found a significant improve-

ment in best corrected visual acuity (BCVA) and a reduction in both cylindrical and higher-order error with this approach. The authors point out that scleral lenses, such as, the Boston Ocular Surface Prosthesis, can be effective for minimizing irregular astigmatism and higher-order aberrations and for significantly improving BCVA in eyes with corneal surface irregularity.

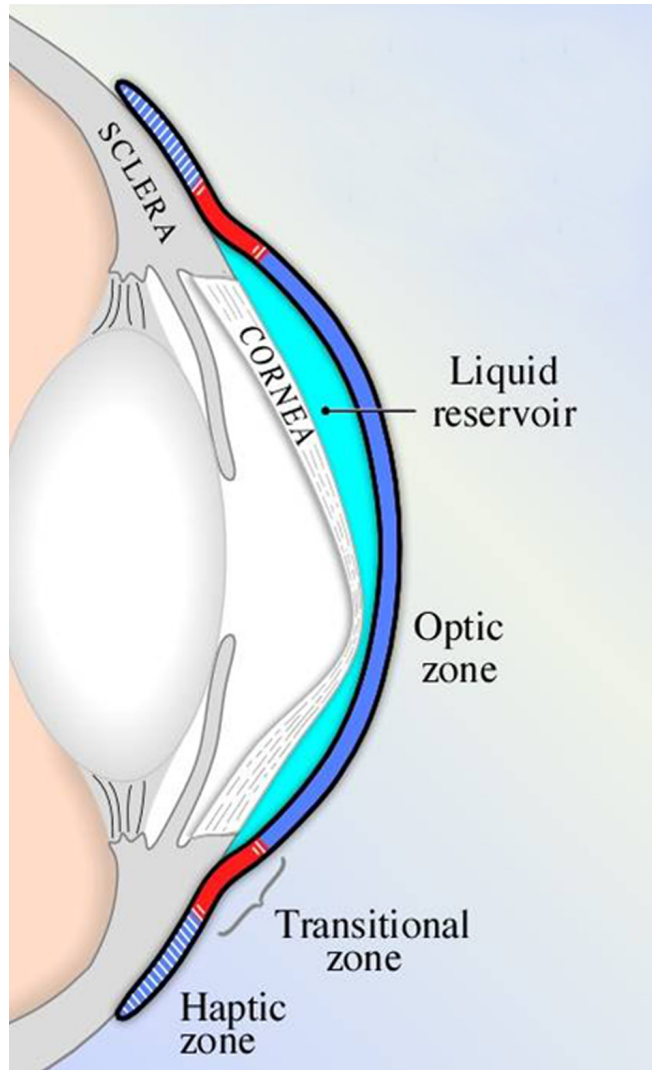


Fig. 2. Schematic diagram of modern scleral rigid-gas permeable lens.

Insight on specific diseases

Scleral rigid gas-permeable lenses for ocular chronic graft-versus-host disease

Schornack et al. (2008) report on 5 patients, 10 eyes, that present with keratoconjunctivitis sicca secondary to GvHD that was not responsive to standard ocular intervention. Their patients were fitted with Jupiter scleral lenses (Medlens Innovations, Front Royal, VA or Essilor Contact Lens, Inc., Dallas, TX). Outcome measures were ability to wear and tolerate lenses, patient reported ocular comfort following lens fitting, and visual acuity pre- and post-lens fitting. All five patients were successfully fit and wearing lenses without any complications, such as, infectious keratitis and corneal neovascularization at follow up (range 4-14 months). Subjective ocular symptoms were improved for all patients, however, no formal metric of ocular comfort was taken prior to or follow-

ing lens fitting. Visual acuity in this series also improved for all patients except one, who was 20/20 preceding lens fit. Two reports from 2007 (Jacobs and Rosenthal, 2007; Takahide et al., 2007) report on the role of scleral RGP lenses in chronic GvHD. The Jupiter lenses that are fit by these investigators (Schornack et al., 2008) are commercially available to eye care professionals, and “may therefore be more accessible to patients who could potentially benefit from this intervention.” The authors of this report conclude that contrary to the perception that the fitting process for scleral lenses is “...extraordinarily complex. We have found that the process is, in fact, relatively straight-forward for the majority of cases.” Silicone hydrogel soft-contact lenses (Focus NIGHT & DAY) (CIBA Vision, Duluth, GA) can also play a role in the management of (GvHD) (Russo et al., 2007). All four studies found improvement in symptoms with Russo et al. (2007) and Takahide et al. (2007) both noting significant changes in Ocular Surface Disease Index (OSDI) scores. (GvHD) is a disease for which scleral RGP lenses or soft lenses are an important option, since little else provides symptomatic relief for these patients.

Scleral rigid gas-permeable lenses for Stevens-Johnson syndrome and toxic epidermal necrolysis

A retrospective review published in 2009 by Tougeron-Brousseau et al. (2009) details the authors’ experience in France fitting scleral RGP lenses in a cohort of 39 consecutive patients, 67 eyes, diagnosed with SJS or toxic epidermal necrolysis (TEN) who were deemed candidates at consultation. OSDI, NEI-VFQ 25, and VA were all outcome measures with the investigators finding significant improvement in all three categories. Mean follow up was 33.3 months (range 16 to 54 months). Consistent with the Stason et al. (2010), a separation of the 32 available VFQ-25 scores into further subgroups revealed the highest improvements in ocular pain (97% of patients) and dependency (81% of patients). Of note in the study is the failure to fit only three patients, one of whom was 7 years old and had difficulty cooperating during the fitting process. However, one 9 year old patient was fit, confirming report from Gungor et al. (2008) that children are candidates for treatment with scleral RGP lenses. The main limitation of the French study developed from inclusion of only patients that were determined to be candidates, which the authors point out may have skewed the results of the study metrics and fitting success in a positive direction.

Rathi et al. from India (2011) report on a cohort of scleral RGP lenses for the management of surface disease resulting from SJS. Scleral lens prosthetic devices, Boston ocular-surface prosthesis (BOS-P), (Boston Foundation for Sight, Needham, MA) were used to fit 20 eyes primarily for relief of pain and photophobia, with improvement in vision a secondary benefit. A significant improvement in vision was noted, but nearly half of patients had debris build up under the lens, requiring them to remove, clean, and reinsert the lenses during wearing periods. Although no method was taken to quantify the symptomatic relief, all patients reported a reduction in symptoms, and the authors concluded that their findings of the BOS-P in SJS patients were encouraging.

Scleral rigid gas-permeable lenses for ocular limbal stem cell deficiency

Three case reports were published since 2008 that discuss the

management of ocular surface disease resulting from deficiency or dysfunction of limbal stem cells (LSCD) with scleral RGP lenses. Schornack (2011) presents a case in which a 36 year old male suffering from LSCD is fitted with Jupiter scleral lenses. Her findings include "...diffuse conjunctival injection in both eyes. Pronounced perilimbal injection was noted in the right eye. Whorl-like, layered corneal epithelial opacities that stained with fluorescein were noted in both eyes, with more in the right." along with a deterioration in vision.

Following nine months of scleral lens wear, the patient's vision had improved and slit-lamp examination showed no signs of conjunctival or corneal inflammation or epitheliopathy. He discontinued lens wear for 18 months with only a mild epitheliopathy noted at a routine follow up, which was resolved with one month of lens wear. Schornack (2011) concluded that the "intervention appears to have allowed for recovery of at least some of the regenerative potential of the patient's limbal stem cells."

Schornack and Baratz published a case report (2009) in detailing their experience managing ocular cicatricial pemphigoid (OCP) with scleral rigid gas permeable (RGP) lenses. A 74 year old woman presented with OCP, and, on slit lamp evaluation, showed severe keratoconjunctivitis sicca, entropion with trichiasis, symblepharon, and central corneal epithelial defects. She was fit with Jupiter scleral lenses and instructed to wear them day and night. After six weeks of lens wear the epithelial defects in both eyes had resolved and vision in the right eye improved from 20/150 to 20/50 while acuity in the left remained stable (hand motion). There is likely increased risk of infection due to overnight wear in this case, which the authors acknowledge. The authors advised the patient to continue overnight wear, even though there was a potential risk of microbial keratitis, to "reduce the risk of additional permanent surface damage." In conclusion, the investigators state "scleral contact lenses may play a role in visual rehabilitation and ocular surface protection in patients whose OCP cannot be managed with traditional systemic and topical therapy."

Grover et al. (2010) reported two cases of persistent epitheliopathy following treatment of conjunctival melanoma. Both patients underwent surgery to excise a conjunctival melanoma and later developed corneal epitheliopathies due to presumed LSCD that resulted from "a combination of damage from the underlying neoplasia, surgical excision or the limbal stem cells, exposure to 18% alcohol, cryotherapy, and topical chemotherapy." Neither patients' persistent epitheliopathy responded to conventional therapeutic measures, and each was referred to be fit with the BOSP.

The first patient was fit with a BOSP and at 11 months had improvement in vision and in comfort with the lenses, and improvement in the appearance of the ocular surface on slit lamp exam. Case 2 was fit with a BOSP and had immediate improvement in vision and symptoms, but daily wear was delayed due to coexisting medical issues. The authors concluded that "the BOSP can be a useful nonsurgical option for these patients"

All three reports involving epitheliopathies resulting from LSCD utilized the unique features of scleral RGP lenses in the management of their respective diseases. Schornack (2011) suggests that not only do scleral RGP lenses protect

the cornea from shear forces from blinking and bath the cornea in fluid to aid in the epithelial healing process, but protective support of the limbus by scleral lenses may also be an important factor in the resolution of these conditions.

Scleral rigid gas-permeable lenses for neurotrophic keratopathy

Gumus et al. (2010) published a case report in 2010 on the use of the BOSP in the management of a persistent corneal epithelial defect resulting from herpes zoster ophthalmicus. At the time of presentation, the patient had developed a 5.6mm x 3.4mm epithelial defect OD and had absent corneal sensitivity. Following extensive therapeutic options, including oral doxycycline, autologous plasma, punctal plugs OD, multiple therapeutic contact lenses, and double layer amniotic membrane transplantation and a temporary lateral tarsoraphy; the authors fit the patient with a BOSP.

The patient was instructed to wear the BOSP daily and a soft contact lens overnight after removal of the BOSP. The epithelial defect healed within 3 weeks of this regimen and vision improved from 20/400 to 20/50. Many cases of epithelial defects resulting from a neurotrophic cornea can be challenging to manage as they may not respond well to therapeutic measures. Although the BOSP has, until recently, had limited availability, the authors propose it as an alternative therapy in the management of persistent epithelial defects resulting from corneal hypoesthesia that do not respond well to conventional measures.

Frontiers and innovations in scleral rigid gas-permeable lenses

Two recently published reports by Lim et al. (2009) and Tanhehco and Jacobs (2010) highlight the novel use of scleral RGP lenses in the management of ocular surface diseases complicated by secondary ocular issues. In the Lim et al. report (2009), the authors describe five cases of ocular surface disease accompanied by corneal neovascularization. All patients were fitted with BOSP lenses for therapeutic indications and, upon establishment of epithelial integrity, were prescribed topical bevacizumab. The BOSP acted as a drug delivery system by utilizing the reservoir as a depot for a dose of 1 drop of 1% bevacizumab. All patients had improved vision and reduction in neovascularization and /or corneal haze. No serious ocular or systemic effects were found in any of the five cases. Tanhehco and Jacobs (2010) detail their experience fitting the BOSP in 5 patients with glaucoma tubes and trabeculectomies. Lens modification after manufacture was the primary approach to dealing with post-operative limbal variants that would affect fit.

2. Conclusion

Herein, we have presented how modern designs, materials, and approaches to fitting scleral lenses have impact in the treatment of patients with OSD. These lenses create an environment at the ocular surface that improves visual function for these patients in terms of both improved comfort and improved visual acuity. Through the mounting body of evidence reviewed here, and continued innovations, it is clear that scleral RGP lenses are an important adjunct in the treatment of ocular surface disease.

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