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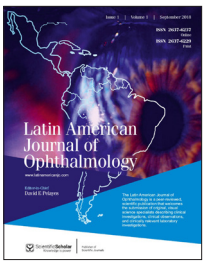

Case Series *Cornea*

Dual benefits of scleral lenses in collateral cases of Stevens-Johnson syndrome and Keratoconus-A case series

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ABSTRACT

Scleral lenses (SL) are larger diameter lenses that rest over the scleral, unlike the corneal lenses. These lenses are fitted to not touch the cornea by creating a space that will be filled with sterile isotonic fluid. Indications of SL include corneal ectatic conditions such as keratoconus (KC), pellucid marginal degeneration,and ocular surface disorders such as dry eye (DE), Stevens-Johnson syndrome (SJS), and graft-versus-host disease. Corneal ectasia in SJS has been rarely reported but still exists, which causes severe DE, photophobia, and blurred vision. In such a scenario, SL is an ideal treatment option as they provide constant lubrication to the ocular surface and better visual acuity. This report witnessed the effectiveness of SL in SJS associated with KC – a rare combination.

Keywords: Keratoconus, Stevens-Johnson syndrome, Scleral lenses, Prosthetic replacement of the ocular surface ecosystem

INTRODUCTION

Keratoconus (KC) is a bilateral, non-inflammatory, progressive thinning disorder of the cornea that results in high irregular astigmatism, distortion, and thereby poor vision. Spectacles, soft and rigid gas permeable (RGP) contact lenses (CL) are helpful in mild–moderate stages of KC. In advanced cases, these lenses may not be helpful in improving vision, and patients may have to use specialty lenses such as scleral lenses (SL). Prosthetic replacement of the ocular surface ecosystem (PROSE) is a food and drug administration (FDA) approved custom-designed SL used to improve vision and delay or obviate the need for keratoplasty in KC conditions. It has been shown to improve visual acuity, visual function, ocular surface comfort, healing, and quality of life in patients with ocular surface disorders.[1-3]

Stevens-Johnson syndrome (SJS) is an acute, systemic immune-mediated mucocutaneous disorder. Ocular involvement of SJS includes keratinized lid margins, symblepharon, severe dry eye (DE), and limbal stem cell deficiency. KC associated with SJS is uncommon but can occur due to multiple factors such as -blink-related micro trauma, persistent tear film disturbance, and ocular surface disease.[4-6]

This case series shows the usefulness of SL in KC associated with SJS for improving vision, comfort, and DE symptoms.

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CASE REPORT 1

A 32-year-old female reported with complaints of blurry vision and photophobia in her right eye. Consulted locally and diagnosed as SJS due to ingestion of penicillin seven years back. The patient underwent punctal cautery in both eyes. On examination, best corrected visual acuity (BCVA) in the right eye (OD) and left eye (OS) was 6/24, N12, and counting fingers (CF), <N36, respectively. Slit lamp shows OD symblepharon, distichiasis, superficial vascularization, scar, and inferior thinning of the cornea. OS was totally keratinized. KC was detected based on the Scheimpflug imaging system (Pentacam-HR; Oculus) in OD with the steepest keratometry readings of 67.8D. The minimum corneal thickness was 361 µm. Schirmer's was OD: 5 mm OS: 1 mm. Meibography shows OD: Grade 4 (>75% dropout) meibomian gland loss.[7] Tried prosthetic replacement of ocular surface ecosystem (PROSE) for vision and comfort. The final lens was dispensed with a vault of 220 microns and good haptics [Figure 1], which improved the vision to 6/9 and reduced DE symptoms.

CASE REPORT 2

A 16-year-old boy developed redness in both the eyes (OU), photophobia, and blurred vision after taking an injection for fever six years back. Diagnosed SJS associated with KC. BCVA was OD: 3/60, <N36, and OS: 6/60, N36. Slit lamp examination shows OU symblepharon, conjunctival inflammation, corneal scar, and vascularization. Schirmer's was OD: 25 mm OS: 30 mm. Meibography shows OU: Grade 3 (>67% dropout) Meibomian gland loss.[7] The patient underwent symblepharon release + Mucous Membrane Graft (MMG) and was referred for PROSE. Final PROSE with a vault of 237 and 249 microns in OD and OS, respectively, improves the vision to OD: 6/15, N6 OS: 6/12, N8. Symptoms of DE and photophobia are reduced significantly and the patient can able to open the eyes better.

CASE REPORT 3

A 31-year-old homemaker was diagnosed with OU SJS in 2017 due to intake of chloroquine and complaints of severe DE, blurred vision, and underwent MMG and punctal cautery in 2018. Developed KC in OU since 2020. BCVA was OU: 6/36, N36. Schirmer's was OD: 4 mm OS: 2 mm. Meibography shows OU: Grade 4 (>75% dropout) Meibomian gland loss.[7] Dispensed PROSE with a vault of 224 and 215 microns in OD and OS improved the vision to OD: 6/12, N10 OS: 6/15, N12.

Table 1 shows the details of all three patients fitted with PROSE.

DISCUSSION

KC is a non-inflammatory corneal thinning disorder that results in irregular astigmatism and poor vision. Spectacles and soft toric lenses can be useful in the early stage of KC. Other options include corneal lenses, piggyback, hybrid lenses, and SL. SL are larger diameter lenses that rest on the sclera and do not touch the cornea and limbus by creating a space between the lens and cornea, which will be filled with normal saline (0.9% sodium chloride). (PROSE; Boston Foundation for Sight) is a FDA-approved custom-designed RGP scleral lens used in the management of irregular corneal conditions and ocular surface disorders.[1]

SJS is a rare, severe immune-mediated mucocutaneous disorder that results in lid margin keratinization,

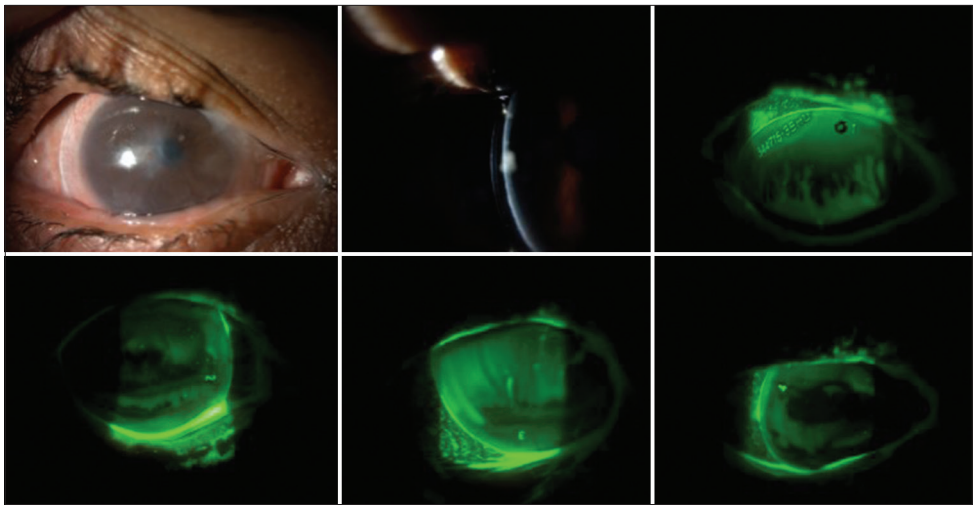

Figure 1: Prosthetic replacement of ocular surface ecosystem fit in Stevens-Johnson syndrome with Keratoconus.

Table 1: Details of patients fitted with PROSE.

Case	Steep K-OD (Diopter)	Steep K-OS (Diopter)	Pachymetry-OD (microns)	Pachymetry-OS (microns)	UCVA OD	UCVA OS	BCVA with PROSE (OD)	BCVA with PROSE (OS)	Fluid Reservoir Thickness-OD (microns)	Fluid Reservoir Thickness-OS (microns)	OSDI Score-Before PROSE	OSDI Score-After PROSE
1	67.80	-	361		6/24	CFCF	6/9	-	220	-	65.9	15.9
2	64.50	58.75	314	328	3/60	6/60	6/15	6/12	237	249	68.7	18.3
3	57.75	60.12	357	312	6/36	6/36	6/12	6/18	224	215	75.6	20.7

PROSE: Prosthetic replacement of ocular surface ecosystem, K-OD: Keratometry of right eye, K-OS: Keratometry of left eye, OD: Right eye, OS: Left eye, UCVA: Uncorrected visual acuity, BCVA: Best corrected visual acuity, OSDI: Ocular surface disease index

symblepharon, DE, and limbal stem cell deficiency. Corneal ectasia in SJS has been reported in a few studies suggesting that it may occur due to multiple factors, such as blink-related microtrauma, which leads to persistent inflammation of the ocular surface. Other factors include persistent tear film disturbance and ocular surface diseases that may contribute to corneal ectasia. In such cases, PROSE is an ideal treatment to provide better vision by reducing corneal aberration and constant lubrication to the ocular surface.[2-4]

PROSE (PROSE; Boston Foundation for Sight) is a FDA-approved custom-designed RGP scleral lens used in the management of irregular corneal conditions and ocular surface disorders.[3] Many studies showed that PROSE devices having a high success rate in providing vision and it also delays the need for corneal transplant in advance KC.[5,6]

Pushker *et al*. showed that MMG for SJS helps to reduce the friction on the ocular surface, thus reducing the ocular surface inflammation and improving the eyelid closure, thereby improving the ocular surface.[8] All three patients in this report also had difficulty in eyelid closure, especially during the initial SL trial, and were advised for MMG, which improves eyelid closure and comfort before dispensing PROSE.

SJS associated with KC can be successfully fitted with SL to improve vision.[2-4] This case series also showed a significant improvement in vision and comfort with PROSE in all the patients. It also compares the ocular surface changes and quantifies the dryness symptoms using ocular surface disease index (OSDI) questionnaire before and after PROSE wear. OSDI survey has 12 validated questions based on ocular symptoms, vision-related function, and environmental triggers. The total score of the OSDI questionnaire ranges from 0 to 100 (0–12 representing normal; 13–22 representing mild DE; 23–22 representing moderate DE; and >33 representing severe DE).[9,10] OSDI survey was given to this patient before and after PROSE wear. OSDI score representing severe DE was significantly improved to mild DE after PROSE use. Considering the steep cornea, fluid reservoir thickness was maintained between 200 and 250 microns. The combination of SJS and KC takes more chair time, fitting challenges result in more than one modification with the PROSE device. The final device was dispensed with optimal fit and produced a remarkable vision improvement, reduced DE symptoms, thus improving the quality of life.

Other treatment options of SJS

Management options for ocular sequelae in SJS patients include topical lubricants, topical steroids, punctal occlusion, tectonic procedures, fornix reconstruction, and limbal allografts. Surface stabilization and fornix reconstruction

include punctal occlusion (collagen or silicon plugs), adnexal corrective procedures, and symblepharon release with amniotic membrane grafting.

MMG for lid margin keratinization has an important role in improving comfort and vision but also in preventing a deterioration of the ocular surface. Visual rehabilitation procedures include limbal allografts and optical iridectomy beneath the area of clear cornea. Various keratoprosthesis procedures include modified osteo odonto kerato prosthesis (MOOKP), Boston type 1 Kpro, and Boston type 2 Kpro.[11]

CONCLUSION

Corneal ectasia in SJS has been rarely reported but still exists. The etiology of corneal ectasia in SJS is unknown but responsible for unexplained low visual acuity. All cases of SJS must be evaluated for corneal ectasia, and PROSE can be a safe and effective treatment option for such patients. The PROSE fitting process involves the use of a proprietary software program that allows for a highly customizable device that precisely fits the curvature of the patient's eye to improve vision and comfort. Improved accessibility to these devices would be necessary to offer this therapy to more patients in need.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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