Debilitated to Functional: How Scleral Lenses Produced a Positive Outcome for a Patient with Stevens-Johnson Syndrome

Amanda Bancroft, B.S., Edward Boshnick, O.D., F.A.A.O

ABSTRACT

A Stevens-Johnson Syndrome patient was fit with a scleral lens for improved ocular surface integrity and visual function.

HISTORY

A 19-year-old male presented complaining of bilateral ocular discomfort and compromised vision for several years. Ocular history is remarkable for Stevens-Johnson Syndrome at age 5 secondary to an antimalarial treatment which led to a superficial keratectomy and buccal-mucosal graft at the limbus OS. Additionally, the patient suffers from long standing constant monocular and binocular diplopia secondary to a compromised corneal surface in both eyes.

VISUAL DEMAND

The patient is in the process of applying to law school, which has a high demand for near-work studying. Unfortunately, reading books and using digital devices are a challenge because of significant reduced visual acuity, diplopia, and constant ocular irritation. Currently, the patient attempts to use a digital magnifier with his one functional eye at an extremely close working distance to read on a phone or computer. In the past, the patient was told the only way to improve his vision is to have a corneal transplant, however, the patient traveled from Malawi Africa with hopes of finding an alternative solution. In order to ensure the patient is comfortable at achieving such intensive near-work in addition to improving ocular surface integrity, the proposed course of action is to utilize the superior optics and sustained wet environment provided by a highly oxygen permeable scleral lens.

EXAM FINDINGS

	OD	OS
BCVA (current refraction)	HM	CF at 3 feet
Pupils	Round, reactive, (-) APD	Round, reactive, (-) APD
Motility	Full	Full
CVF	Mild constriction 360	Mild constriction 360
SLE	Marked injection; corneal neovascularization with fibrosis; limbal stem cell deficiency	Marked injection; corneal neovascularization with fibrosis ; limbal stem cell deficiency

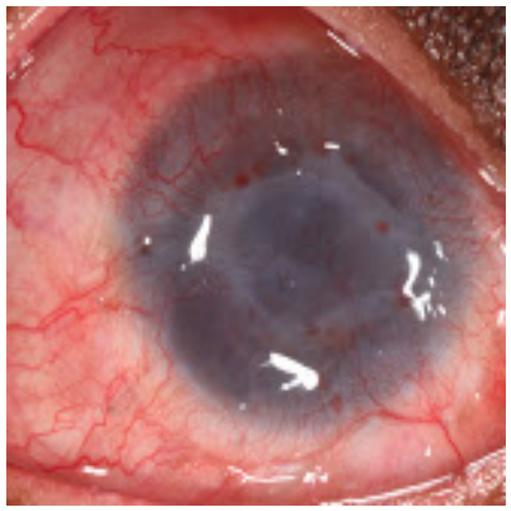

Figure 1: Left eye before application of a scleral lens or ocular surgery

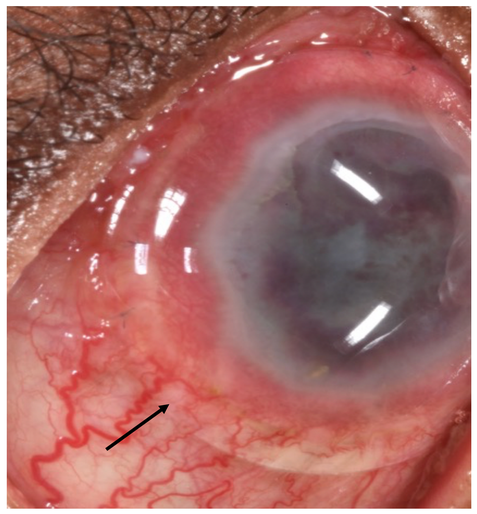

Figure 2: scleral lens properly vaulting the cornea two weeks post buccal-mucosal graft surgery.

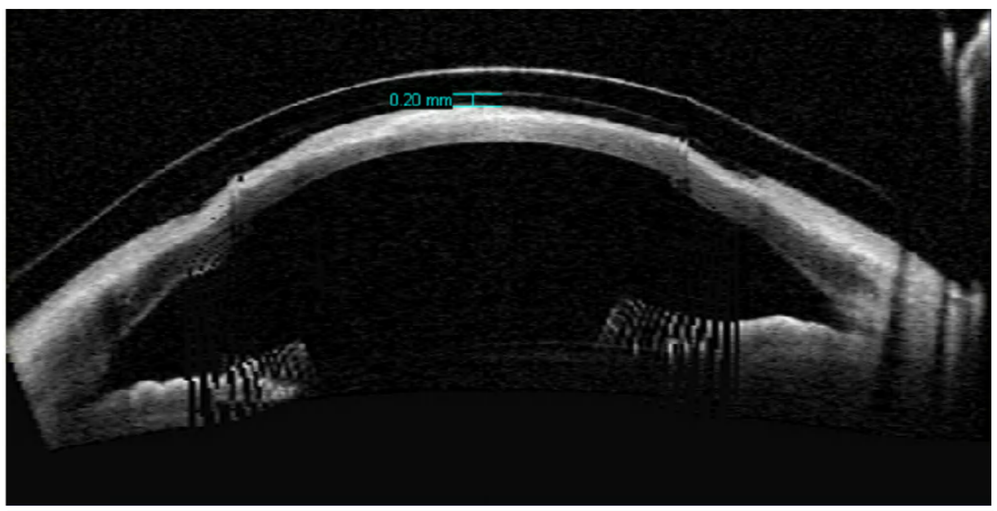

Figure 3: Anterior Segment OCT view of custom EyePrintPRO™ vaulting over the entire cornea. The irregular cornea can be visualized as well as the buccal-mucosal graft at the limbus

Figure 4: EyePrintPRO™ impression of the left eye 2 weeks post buccal-mucosal graft surgery

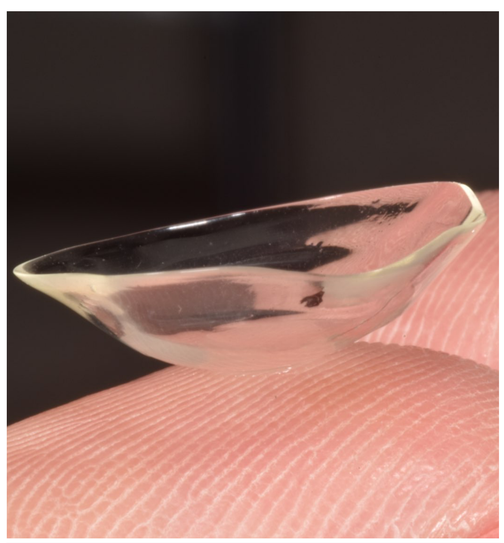

Figure 5: Profile view of EyePrintPRO™ scleral lens for the irregular ocular surface.

DIAGNOSIS AND DISCUSSION

Stevens-Johnson Syndrome is a Type IV hypersensitivity reaction primarily resulting from medication use. Other less common causes include viral/bacterial infections or collagen vascular diseases. SJS can potentially lead to vision-threatening ocular manifestations. Patients experience extreme ocular discomfort and reduced vision due to immune-mediated insult to ocular mucosal tissue. Conventional visual corrections fail to address the compromised ocular surface and traditional wetting agents alone are ineffective due to the constant demand for lubricating the dry, inflamed ocular surface. Corneal procedures such as a penetrating keratoplasty are not ideal because of the high risk for graft rejection secondary to inherent inflammation present on the ocular surface. These procedures also fail to address the damaged conjunctiva. Thus, a lens that vaults over the cornea is indicated. To achieve optimal time in a wet environment, a scleral lens modality could be utilized.

TREATMENT AND MANAGEMENT

Considering this patient's need for ocular surface rehabilitation and the presence of an irregular sclera, an EyePrintPRO™ scleral lens modality was chosen. Rather than standardized curves, a polymer impression was taken of the patient's cornea and sclera. Using EyePrint Designer Software, a scleral lens was successfully created and fit to the patient's irregular ocular surface. The lens provided adequate central and limbal clearance with no signs of blanching or edge lift. Visual acuity of 20/80 was achieved OS. After the initial lens dispense, the patient returned for multiple follow-up visits to evaluate symptomatology. At each visit, the patient reported significant improvement in ocular comfort and visual function. The patient is able to achieve all-day wear. A scleral lens will be made in the near future for the right eye to improve acuity and achieve binocularity.

CONCLUSION

Scleral lenses are a novel solution to rehabilitate and maintain the ocular surface in patients with Stevens-Johnson Syndrome.

REFERENCES

Available upon request