Stevens Johnson Syndrome, Keratoconus, and the Scleral Lens

Curtis Finch, OD; Brianna Ryff, OD, FAAO; Amy Schweikert

Introduction

In patients with concurrent presentations of Stevens Johnson Syndrome (SJS) and Keratoconus (KCN), a scleral lens can provide both preservation of the ocular surface integrity and the greatest benefit to vision.

SJS in the chronic phase is known to cause ocular complications including: progressive conjunctival shrinkage and scarring, corneal damage from trichiasis, cicatricial lid margins, severe ocular surface disease from exposure, and scarred lacrimal glands.

Due to the progressive nature of ocular changes in SJS and keratoconus, scleral lens patients with at-risk corneas should be monitored closely for changes that may alter the fit of the patient's lens. A poor fitting scleral lens may result in a progression of limbal stem cell deficiency (LCSD), keratitis, or neovascularization, and should be managed appropriately.

Case

This case involves a 45-year-old Caucasian female with a history of Stevens Johnson Syndrome (SJS), consequential ocular sequelae, and keratoconus.

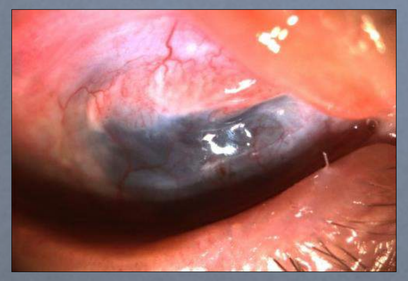
The patient presented with an unknown, 3-year old scleral lens OS, with an inadequate fit involving excessive blanching of conjunctival vessels.

BCVA of 20/40-1 OS through habitual scleral.

Upon initial presentation, the patient's left eye exhibited LSCD with pannus superiorly, inferior neovascularization, 3+ diffuse superficial punctate keratitis (SPK), and conjunctival thickening.

Figure 1. Patient's right eye with consequential ocular sequelae of SJS.

OD



Results

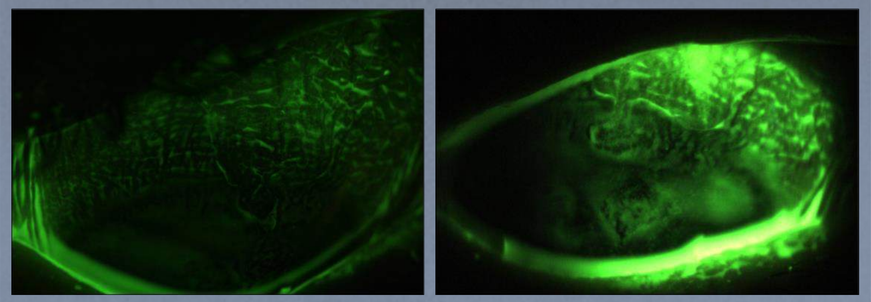


Figure 2. Limbal Stem Cell Deficiency (LSCD) observed via NaFl staining upon initial and final presentations.

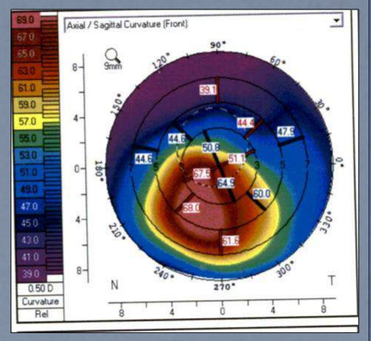


Figure 3. Oculus Pentacam Axial/Sagittal Topography

K1: 57.4D Pachy Apex: 315um
K2: 58.5D Thinnest Loc: 279um
Kmax: 73.2D

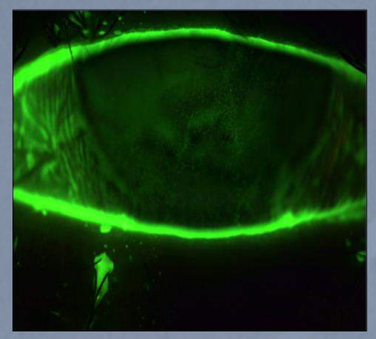
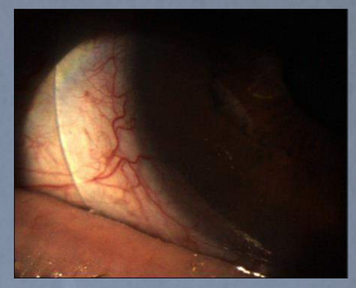
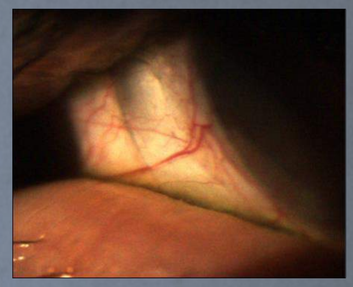


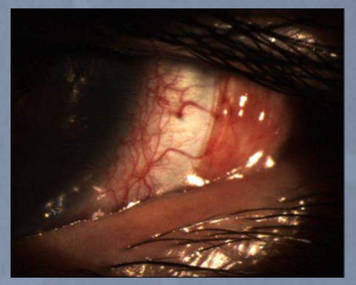
Figure 4. Moderate punctate keratitis staining with NaFl s/p habitual scleral lens removal.



Habitual



Final Scleral



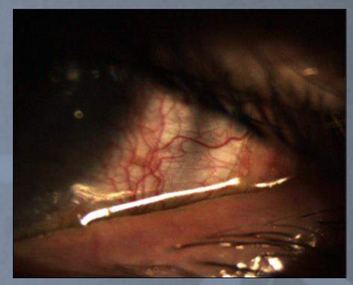


Figure 5. Conjunctival blanching, impingement, and vessel impingement observed with the patient's habitual lens is minimized via a proper scleral lens fit.

Final Scleral Lens Parameters

Type	Power	BC	DIA	SAG	LCC	APS
Z-11 Prolate 17	-21.25 – 2.00 x 142	6.90 mm	17.0 mm	5425 um	H +100 V-150	H + 5 V + 4

The patient was successfully re-fit with a ZenLens to maintain ocular surface integrity.

Conjunctival blanching of the lens secondary to conjunctival thickening associated with SJS was minimized through several adjustments of Limbal Clearance Curve (LCC) and Advanced Peripheral System (APS) parameters.

Proper limbal and central clearance was achieved with adequate vault over the cone (375um clearance), and the signs of SPK mildly improved.

A proper fit increased comfort for the patient, allowing her to wear the lens for an extended period of time (~6-8 hours). The patient was optically corrected to 20/30+1 OS.

Conclusion

Scleral lenses have become a staple tool in managing keratoconus and ocular surface disease, but an improper fit may lead to the cornea becoming compromised. Due to progressive conjunctival shrinkage and scarring associated with SJS, monitoring the peripheral lens alignment more frequently is recommended.

Corneal changes such as LSCD and neovascularization, should be closely monitored with anterior segment photography, epithelial mapping, slit lamp exam, and the use of vital dyes.

Early detection of potential complications from SJS in conjunction with scleral lenses can prevent the need for future corneal surgeries (transplants) and provide superior comfort and visual correction for these complex patients.

MIDWESTERN UNIVERSITY

Educating Tomorrow's Healthcare Team

References:
HN, S., S, K., HG, L., J, C., & DS, J. (2016). Stevens-Johnson Syndrome and Corneal Ectasia: Management and a Case for Association. *American Journal of Ophthalmology*, 276-281.
Wang, J. W., Rixen, J. J., Goins, K. M., & Kitzmann, A. S. (2014, August 18). *Ocular Manifestations of Stevens-Johnson Syndrome*. Retrieved from Ophthalmology and Visual Sciences - University of Iowa Healthcare: https://webeye.ophth.uiowa.edu/eyeforum/cases/192-Stevens-Johnson.htm