THE OHIO STATE UNIVERSITY
WEXNER MEDICAL CENTER

When Options Run Dry – Treatment of Ocular Surface Disease in a Patient with Stevens-Johnson Syndrome

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INTRODUCTION

Steven-Johnsons Syndrome (SJS) is a self-limiting inflammatory disorder affecting the skin and mucous membrane. It usually develops in response to an adverse drug reaction. The condition can affect the cornea, conjunctiva, and eyelids and can ultimately lead to severe dryness and visual impairment.

Background

A 49-year-old Caucasian female presents for a scleral lens follow-up. Her systemic history is significant for Stevens-Johnsons Syndrome caused by the drug, Lamitcal, resulting in severe ocular surface disease OD > OS. Prior failed treatments include artificial tears, ointment, Restatsis, autologous serum tears, amniotic membrane transplants, complete punctual cauterization, Elman radio-ablation/epilation of RUL and LLL, and partial tarsorrhaphy.

Case Presentation	
Chief Complaint	Eye dryness OD > OS, Scleral lens follow-up OU
Medical History	Stevens-Johnson Syndrome starting in 2015 (most recent flare-up 1/20/2017), Thrombocythemia, Depression
Systemic Medications	Alaprazolam 0.5mg, Lexapro 10mg, Lunesta 3mg, Melatonin 3mg, Trazodone 150mg
Ocular History	Ocular surface disease s/p SJS resulting in the following: puncta cauterization OU, Elman radio-ablation RUL, LLL, partial tarsorrhaphy OS, PEM IOL OU
Ocular Medications	Restasis BID, Lotemax ung TID, Genteal gel qhs, Pred Forte BID, Serum Tears q6h, preservative-free tears prn OU
Allergies	Lactimal, Neosporin, sulfa
Slit lamp Examination	OD: external- erythema; lids/lashes- cauterized puncta; conj/sclera- foreshortening of cul de sacs, 2+ injection; cornea- trace PEE, poor quality TF c irregular epi pooling OS: external erythema; lids/lashes- cauterized puncta, misdirected lashes; conj/sclera- cul de sac appears normal, 2+ injection; cornea- trace PEE, small sup scar
Fundus	OD: wnl OS: wnl
Acuity	OD: sc 20/200, PH 20/30-2 OS: sc 20/70, PH 20/30

Methods and Results

The patient is fit into a scleral lens in both eyes due to severe ocular surface disease following SJS. A large diameter lens is used to provide protection of the ocular surface and improvement in visual function. However, wear time is limited due to mechanical lid interaction following lash ablation OD. Quadrant-specific peripheral curve changes is used to improve comfort.

Office Visits for fitting OU	
Office Visit	**Chief complaint / Notes**
Initial Fitting	CC: Eye Dryness OD > OS Notes: Patient unable to tolerate 19.0mm scleral
Initial Scleral Dispense	Lenses not dispensed due to excess vault OD and edge lift OS; decrease sag and steepen quad 4 OD, steepen quad 1,2 OS (see figure 4)
Scleral dispense #2	BCVA OD 20/20-3, OS 20/20; Successful application and removal, Lenses dispensed
Follow-up #1	CC: OD lens getting caught during blink; Notes: Minimal vault OU; steepen quad 2 OD, increase SAG OU
ScCL disp #3- F/U #2	No changes, patient fully adapted to lenses but limited wear-time (4 hrs) due lid interactions

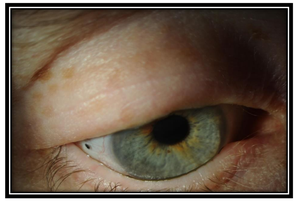

Figure 1. (Topcon, Nidek)

Shows right upper lid s/p Elman radio-ablation. This resulted in discomfort between upper lid and scleral lens.

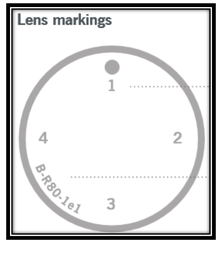

Figure 2.

Shows quadrant marked lens with a dot at quadrant 1. "Courtesy of BostonSight"

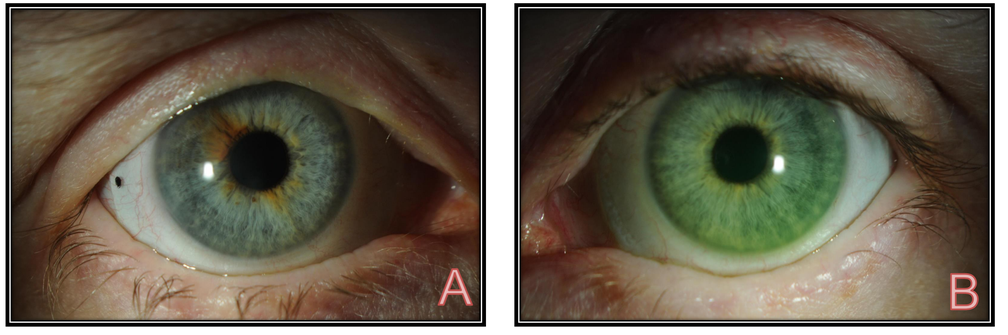

Figure 3. (Topcon, Nidek)

Figure 3A and 3B shows 18.0 mm BostonSight lenses providing adequate protection of the ocular surface. Quadrant specific changes were made to improve comfort. However, the patient is only able to wear the lenses for a limited time due to mechanical lid interactions following lash ablation.

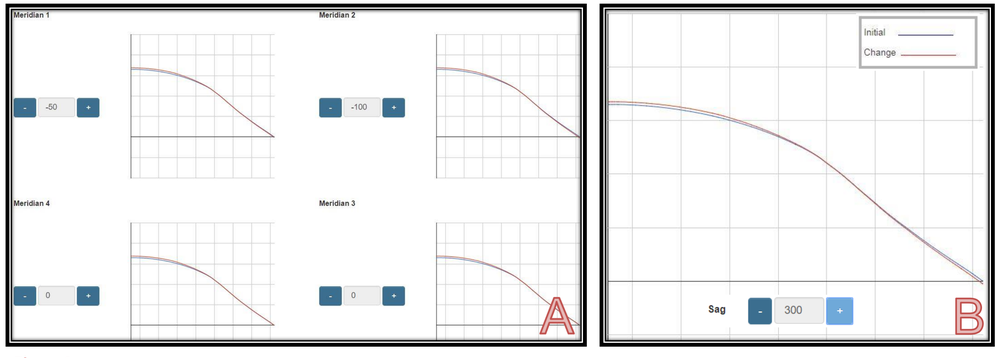

Figure 4.

Figure 4A shows the BostonSight Fitconnect feature allowing visualization of quadrant specific changes of the patient's left eye. It demonstrates increases in sag by 300um and steepen edges 1 by 50um and 2 by 100um. Figure 4B shows a zoomed-in view of quadrant 2 from figure 4A. "Courtesy of BostonSight"

Final Scleral Lens Prescription OU							
Brand	**Overall Diameter (mm)**	**Base Curve (mm)**	**Power (D)**	**CT (mm)**	**Dot placement (Deg)**	**Material**	**BCVA**
BostonSight Scleral OD	18.0	8.0	+6.25	0.35	180	CXtra100	20/20-3
BostonSight Scleral OS	18.0	8.0	+0.25	0.43	060	CXtra100	20/20

Discussion

Stevens-Johnson Syndrome is a rare acute inflammatory condition usually in response to adverse drug reactions. It is an acute epithelial blistering and membrane erosion of the skin that can be fatal. Ocular involvement in the acute phase has been reported in 50-88% of patients. This includes acute conjunctivitis with corneal involvement. Signs following the acute stage can result in persistent epithelial erosions, ulceration, and perforation. This can ultimately lead to corneal, conjunctival, and eyelid cicatricial changes such as neovascularization, keratinization, opacification and symblepharon resulting in severe dryness and visual impairment.

Studies show that treating ocular symptoms aggressively during the acute phase reduces the risk for chronic ocular symptoms. Various treatments used in the management include non-surgical intervention such as lubrication, topical steroid/cyclosporine, autologous serum tears, and punctal occlusion. Surgical intervention include amniotic membrane transplantation, fornix reconstruction, etc.

A study has shown PROSE treatment has durable effectiveness in the treatment of patients with ocular surface disease related to SJS. Scleral lenses are often used as early intervention. Advancements in lens design and surface coatings have improved outcomes.

CONCLUSIONS

Scleral lenses provide successful management of ocular surface disease in patients with Stevens-Johnsons Syndrome. Advanced fitting techniques such as quadrant specific designs and lens coatings can help fit these complex ocular surfaces. Tangible Hydra-PEG will be considered in the future for this patient to increase lubricity and help alleviate the mechanical lid interactions.

ACKNOWLEDGEMENTS

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