The Application of Scleral Contact Lenses in the Management of Coexisting Keratoconus and Stevens-Johnson Syndrome

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INTRODUCTION

Scleral contact lenses are large-diameter lenses that vault over the cornea and rest on the sclera. Indications for these lenses include preventing desiccation due to ocular surface disease or improvement of vision due to irregular astigmatism; there is a greater indication for the use of scleral lenses when treating these conditions concurrently. Stevens-Johnson syndrome (SJS) is a complex immune-mediated mucocutaneous disease. It is characterized by the acute blistering and keratinization of various mucous membranes leading to sloughing of the mucosal surfaces, including the ocular surface. Keratoconus (KCN) is a noninflammatory corneal disorder where the central or paracentral corneal tissue progressively thins and steepens, ultimately causing decreased vision. This case highlights the therapeutic use of scleral lenses in KCN coexisting with SJS.

CASE REPORT

A 25-year-old African American male presented for blurry vision at distance and near without correction OU, along with ocular irritation OU. He previously wore glasses, but they offered little improvement of vision. His medical history was positive for SJS secondary to phenytoin use. He also reported burns secondary to SJS that gradually healed. Treatment included Restasis BID OU, Prednisolone Acetate QID OU, and artificial tears.

Entering uncorrected distance acuities were 20/80 OD and 20/200 OS. Trichiasis and thickening of the upper and lower eyelid margins in addition to diffuse conjunctival injection and scarring of the palpebral conjunctiva of both eyes was noted. The corneas of both eyes had peripheral neovascularization 360 OD>OS and inferior corneal steepening OS>OD as highlighted in Figure 2. Corneal tomography confirmed KCN OS>OD (Figure 1). The patient was diagnosed with SJS with concurrent KCN. Dilated fundus exam was deferred.

DIAGNOSIS AND DISCUSSION

There are only a few case reports on patients with SJS and concurrent KCN with no known associations between the two conditions at this time. However, it is postulated that the corneas of SJS patients may be susceptible to corneal ectasia due to eye rubbing, blink related micro-trauma, and increased inflammatory mediators. Regardless of the pathophysiology both conditions can benefit from treatment with scleral lenses.

FIGURE 1
Tomography showing anterior and posterior corneal steepening OU.

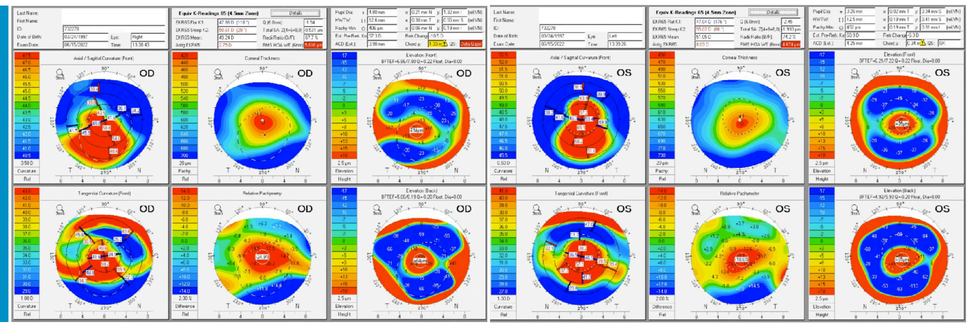

FIGURE 2
OU on the day of presentation showing trichiasis and thickening of the upper and lower eyelid margins OU, 1+ conjunctival injection OU, and peripheral corneal neovascularization 360 OD>OS.

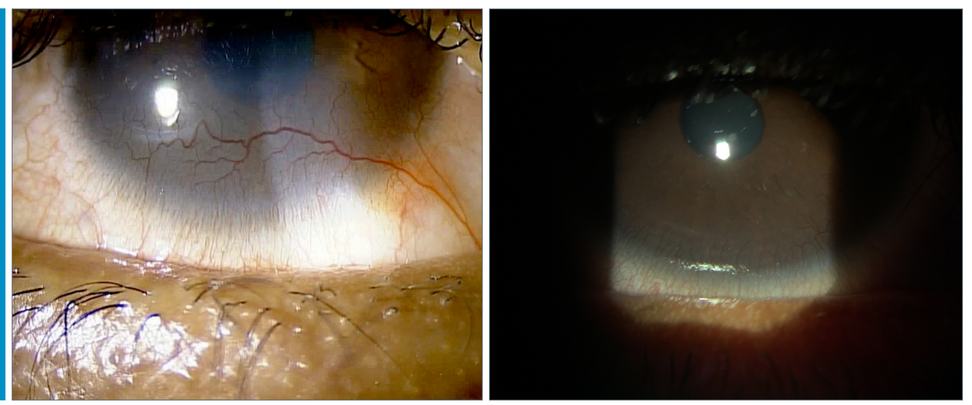

FIGURE 3
OU after 1.5 months of full-time scleral lens wear showing improved peripheral corneal neovascularization 360.

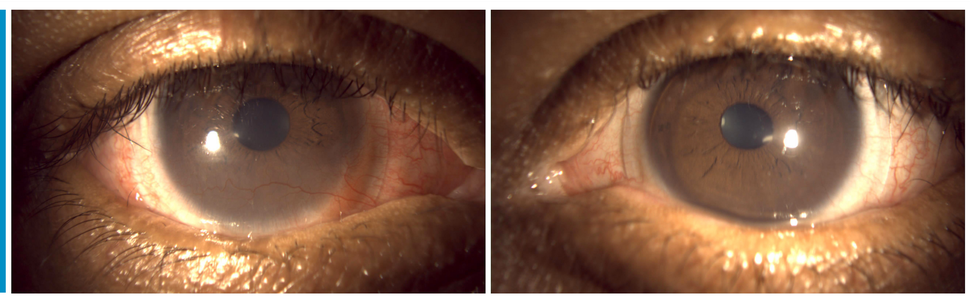

FIGURE 4
Onefit MED scleral lens parameters

	Power	Sag	OAD	M	L	Edge	Material
OD	+0.50 DS	4150	16.0	STD	STD	+100/-50	Optimum Infinite
OS	+2.00 DS	4350	16.0	STD	STD	+125/-25	Optimum Infinite

TREATMENT/ MANAGEMENT

The patient was fit into Onefit MED (CooperVision Specialty EyeCare) scleral contact lenses of 16.0mm diameter (Figure 4) that were selected based off corneal irregularity and horizontal visible iris diameter. The scleral contact lenses vaulted the cornea allowing the tissue to be continuously bathed in sterile saline. This allowed for healing of the ocular surface and protection of the cornea from aberrant eyelashes/keratinized eyelid margin trauma as highlighted in Figure 3. The scleral lenses also allowed for a smooth refractive surface which improved the acuity from 20/80 to 20/40 OD and from 20/200 to 20/40 OS. The patient also reported symptomatic relief of the ocular irritation OU.

CONCLUSION

Although rare, clinicians should be aware of the co-existence of SJS and KCN. Patients with concurrent KCN and SJS can be fitted with scleral lenses to improve quality of vision and prevent corneal desiccation. Management with scleral lenses can prevent further scarring and keratinization which reduces the risk of vision loss while improving comfort.

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