

Therapeutic Applications of Scleral Contact Lenses in Ocular Cicatricial Pemphigoid

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Background:

- Ocular cicatricial pemphigoid (OCP) is a rare autoimmune, sight-threatening disorder with an incidence anywhere between 1/8000 and 1/60,000
- Corneal and conjunctival cicatrization from ocular surface inflammation, compromised lid anatomy and function along with decreased tear production may lead to corneal opacification, ulceration, perforation, and devastating vision loss.

Symptom management in Ocular Cicatricial Pemphigoid (OCP) is challenging due to the progressive factors of dry eye, scarring, trichiasis and vision impairment. We set out to evaluate the benefits and therapeutic effect of scleral contact lenses in the management of OCP.

Methods:

A retrospective review was performed over a database of 20 patients (36 eyes) fitted with scleral lenses (SL) at the Department of Ophthalmology, Emory University from May 2018 to April 2021.

We evaluated, the time required for the ocular surface stabilization, vision rehabilitation success, and OCP-related ocular surface signs (including trichiasis and symblepharon **(Figures 3A & 3B)**).

Figure 3A (bottom left): Trichiasis and resultant corneal scarring from mechanical insult.
Figure 3B (bottom right): Symblepharon and fornix shortening resulting from scarring. Both complications of OCP may lead to reduced vision, poor comfort, and significant scarring if left untreated.

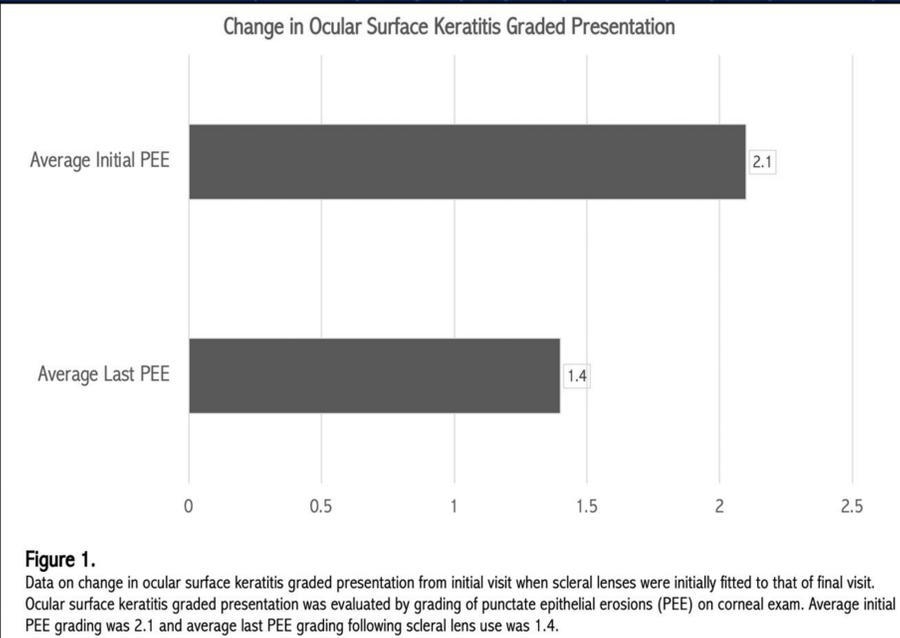
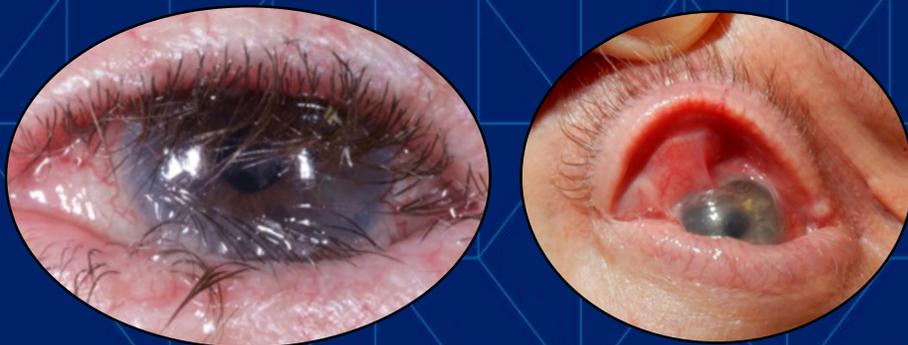


Figure 1.

Data on change in ocular surface keratitis graded presentation from initial visit when scleral lenses were initially fitted to that of final visit. Ocular surface keratitis graded presentation was evaluated by grading of punctate epithelial erosions (PEE) on corneal exam. Average initial PEE grading was 2.1 and average last PEE grading following scleral lens use was 1.4.

Results:

- The mean age was 67.4 (range, 43-81) years, with 4 to 1 female to male predominance
- Mean duration of follow-up was 17.5 months (range, 1.5-35)
- Mean wearing time was 10.9 hours a day, with no overnight wear permitted
- Mean keratopathy grading improved from 2.1 ± 0.8 to 1.4 ± 0.7 at the last documented visit **(Figure 1)**
- Mean visual acuity improved from 20/80 to 20/30 with SL (range, 20/400 to 20/20) **(Figure 2)**
- All fitted patients reported a subjective improvement in ocular comfort after initiations of SL wear **(Figure 4)**
- 96% of fitted eyes responded well to therapy and continued scleral lens wear
- Patients who initially presented with fornix shortening, symblepharon, and trichiasis remained stable

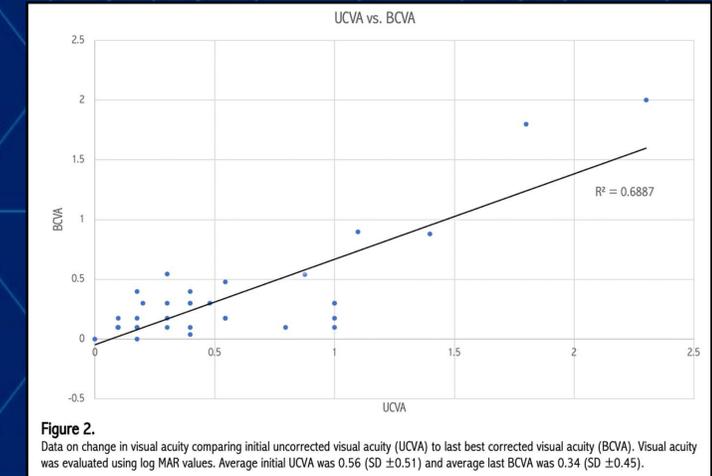


Figure 2.

Data on change in visual acuity comparing initial uncorrected visual acuity (UCVA) to last best corrected visual acuity (BCVA). Visual acuity was evaluated using log MAR values. Average initial UCVA was 0.56 (SD ± 0.51) and average last BCVA was 0.34 (SD ± 0.45).

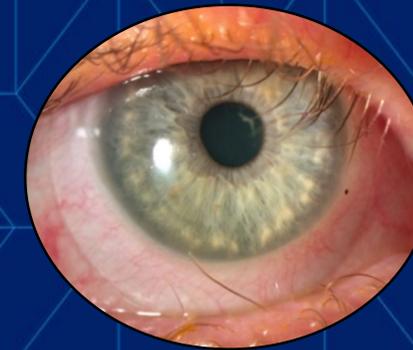


Figure 2 (above): Visual acuity in 42% of patient's eyes was improved by two or more lines between initial and final visits. The remainder of patients had stable visual acuity throughout the lens fitting process.

Figure 4 (left): A well-fitted scleral lens provided ocular surface protection, improvement in ocular surface disease, and increase in patient comfort.

Conclusions:

Patients presenting with fornix foreshortening, symblepharon, keratopathy, and trichiasis remained stable throughout the study period. Scleral lenses offer both ocular surface protection and improved visual acuity in patients with OCP. Scleral lenses should be given strong consideration as an ancillary therapy in patients with OCP.

References:

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- Zillikens D, Wever S, Roth A, et al. Incidence of autoimmune subepidermal blistering dermatoses in a region of central Germany. Arch Dermatol. 1995 Aug;131(8):957-8