Scleral Contact Lens Management of an Ectatic Microcornea

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Introduction

- Pellucid marginal corneal degeneration (PMD) is a bilateral, progressive, condition of inferior peripheral corneal thinning within 1 to 2 millimeters from the limbus.[1,2]
- Microcornea is a unilateral or bilateral condition when the adult horizontal corneal diameter is less than 11mm.[4,5]
- A monocular 66-year-old male with a diagnosis of PMD and microcornea of the right eye presented for a scleral gas permeable (GP) contact lens fitting.

Case History

- 66-year-old Caucasian male.
- CC: Glare and poor lens comfort with excessive lens movement and edge awareness with corneal GP contact lens.
- POH: OU: Microcornea, PMD.
- OD: Aphakia with cataract extraction, retinal detachment with scleral buckle, punctal cauterization, excisional biopsy with amniotic graft.
- OS: Correctopia, blind left eye.
- PMH: Hearing loss in the right ear, hypercholesterolemia, hypothyroidism, acid reflux, ear surgery, facial reconstruction surgery.
- Medications: 81mg Aspirin, Crestor, Lovazar, Synthroid, generic acid reducers.
- Allergies: None.
- Presenting Contact Lens: Corneal GP contact lens in the right eye only. No contact lens wear in the left eye.

Pertinent Clinical Findings

- Entering VA (through corneal GP contact lens right eye only): OD: 20/60+ OS: No light perception.
- EOMs: OD: Full range of motion. OS: Restriction noted in superior temporal gaze. OU: Small nystagmus noted in primary gaze.
- Pupils: OD: Irregular pupil opening surgical tear noted inferior with no response to light. OS: Inferior nasal coreotopia, no response to light. (see Figure 1)
- CVF: OD: Restriction noted in superior visual field. OS: No light perception.
- Corneal topography and Sim K readings OD only:
  OD: Irregular astigmatism with an atypical topographical pattern resembling PMD. (see Figure 2)
  OD: 35.9 / 40.4 @ 180
- Anterior segment: OD: 1mm corneal neovascularization 360°, 1-2+ diffuse corneal staining, small inferior paracentral scar. OS: 2+ endothelial pigment, 1+ diffuse corneal staining. OU: 0.5mm HVID, inferior corneal thinning consistent with PMD, 1+ papillae, mild hyperemia, and conjunctivochalasis.
- Posterior segment: OD: Scattered vitreal floaters, large CNVH with indistinct rim tissue, macular atrophy, diffuse inferior retinal whitening without signs of retinal detachment. OS: Unable to view.
- IOP: OD: 9 mmHg OS: 16 mmHg

Differential Diagnoses

Corneal Ectasia (see Chart 1)
- Primary: Pellucid Marginal Corneal Degeneration
- Others: Keratoconus, Keratoglobus

Chart 1: Explanation of Ectatic Cornea Differential Diagnoses

Pellucid Marginal Corneal Degeneration[1,2]	Keratoconus[1,2]	Keratoglobus[1,2]
• Rare bilateral, progressive, condition of inferior peripheral corneal thinning • Located 1-2mm from the limbus, extending from 4-8 o'clock • Onset early adulthood (2nd to 5th decade) • Signs include "sleeping dragon" against-the-rule pattern on corneal topography, hydrops can occur • Tx: Spectacle Rx, GP contact lenses, surgeries with reduced progress including: lamellar keratoplasty, penetrating keratoplasty, and inferior crescentic wedge resection	• Bilateral, asymmetric corneal thinning • Located in the central or paracentral cornea with protrusion at the apex of the thinning • Onset during adolescence and progressing until adulthood (4th decade) • Signs include Fleischer's ring, Vogt's striae, progressive myopia with irregular keratometry mires, and acute hydrops can occur • Tx: Spectacle Rx, GP contact lenses, surgeries including: lamellar keratoplasty, penetrating keratoplasty, and corneal cross-linking	• Extremely rare bilateral, severe thinning of the entire cornea • Onset shortly after birth • Signs include very thin global corneal thickness, very deep anterior chamber, hydrops can occur in severe cases • Tx: Spectacle Rx, GP contact lenses, surgery (can be difficult)

Small Horizontal Visible Iris Diameter (HVID) (see Chart 2)
- Primary: Microcornea
- Other: Microphthalmos, Sclerocornea

Chart 2: Explanation of Small HVID Differential Diagnoses

Microcornea[4,5]	Microphthalmos[4,5]	Sclerocornea[4,5]
• Uncommon autosomal dominant or recessive, bilateral or unilateral condition • Horizontal corneal diameter less than 10mm in infants and 11mm in adults • Signs include shallow anterior chamber, hyperopia, flat cornea • Tx: Manage refractive error, monitor for other ocular/systemic abnormalities	• Uncommon unilateral or bilateral condition of a malformed eye with a reduced axial length • Signs include small cornea, abnormal iris, and small overall ocular size • Tx: Manage refractive error, monitor for other ocular/systemic abnormalities	• Rare, congenital, asymmetric, bilateral, sporadic condition • Signs include opacification and vascularization of the cornea, can be partial or complete • Can appear to be microcornea if only peripheral cornea is involved • Tx: Manage refractive error, consider penetrating keratoplasty if bilateral/complete

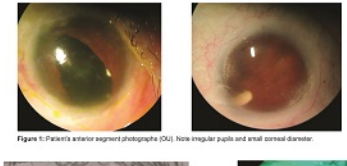

Figure 1: Patient's anterior segment photographs (OU). Note irregular pupils and small corneal diameter.

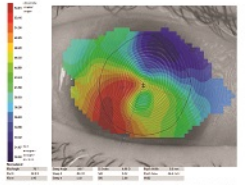

Figure 2: Patient's corneal topography: irregular astigmatism with pellucid-like pattern noted (OD).

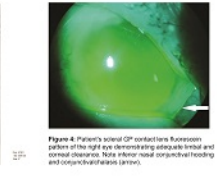

Figure 4: Patient's scleral GP contact lens fluorescein pattern of the right eye demonstrating adequate limbal and corneal clearance. Note inferior nasal conjunctival hooding and conjunctivochalasis (arrow).

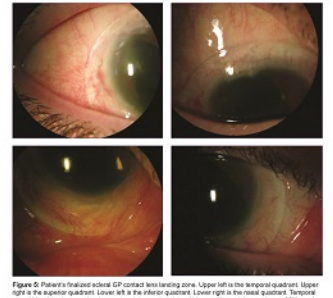

Figure 5: Patient's finalized scleral GP contact lens landing zone. Upper left is the temporal quadrant. Upper right is the superior quadrant. Lower left is the inferior quadrant. Lower right is the nasal quadrant. Temporal vessel blanching is secondary to scarring status post excisional biopsy with amniotic membrane. Mild nasal blanching is present in right gaze and disappears in primary gaze.

Treatment and Management

- Discontinued corneal GP contact lens wear in the right eye.
- Designed a small diameter scleral lens that would fit on the ocular surface similar to a full scleral lens on an eye with a normal corneal diameter using a corneoscleral lens fitting set. (see Chart 3)
- To achieve an acceptable fit from the initial corneoscleral lens design, the base curve was flattened to decrease the corneal clearance and the transition zone curves/landing curves were altered to allow the lens to align the ocular surface similar to a full scleral lens design. (see Figure 3)
- Final Scleral GP contact lens fitting relationship: Approximately 300µm of apical corneal vault, limbal clearance in all quadrants, variable scleral alignment with changes in fixation due to conjunctivochalasis. Trace to moderate blanching noted nasal, temporal, and inferiorly that changes with fixation. Nasal, inferior, and temporal conjunctival hooding. (see Figures 4,5)
- Best corrected visual acuity improved to 20/50+2 and the lens provided the patient good comfort with 8+ hours of uninterrupted wear time.
- Finalized scleral GP Lens and thoroughly educated the patient on proper lens care. Discussed lens overwear and importance to monitor closely due to monocular status.
- Prescribed Refresh preservative free artificial tears to be used as needed for ocular comfort in both eyes.
- Continued care with referring OD/OMD to monitor ocular health. Recommend low vision evaluation for near devices to help with activities of daily living.
- Return in three months to monitor the cornea and contact lens fit. Monitor for changes associated with conjunctival hooding and corneal edema.

Chart 3: Initial and Final Lens Parameters

Material	BC (Diam)	Dia (mm)	Power (D)	OZD (mm)	PC1 (mm)	PC2 (mm)	PC3 (mm)	PC4 (mm)	PC5 (mm)	CT (mm)
Boston XO2 dk:141	46.50/7.26	14.4	-1.00	7.4	7.56x1.8	7.95x1.0	9.60x0.5	13.50x0.5	14.70x0.2	0.21
Boston XO2 dk:141	44.00/7.67	13.8	+1.50	7.4	6.69x0.8	7.16x0.8	8.20x0.5	14.50x0.5	14.70x0.3	0.35

Figure 3: Lens profile illustrating how the peripheral curves of the lens differ from the initial contact lens design and the finalized contact lens. (Image Courtesy of Essilor Contact Lens)

Conclusions

- Scleral GP contact lens prescribing is becoming more prevalent in the optometric profession to improve the overall health, comfort, and vision for irregular corneas.
- Corneal ectasia in PMD is located inferiorly making contact lens fitting more challenging. The addition of a microcornea requires knowledge and experience with different specialty contact lenses.
- When fitting patients with specialty contact lenses, it is important to closely monitor corneal changes, as well as, changes in the fitting relationship. Patients with irregular corneas and wearing specialty contact lenses should be monitored every 4-6 months. In this case, the patient should be monitored every 3-4 months due to the multiple ocular conditions and his monocular status.
- It is important to find the appropriate balance that allows for a healthy fit providing good comfort and vision for the patient requiring specialty lenses.
- This challenging case demonstrates how specialty contact lenses can improve a patient's quality of life.

References

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