Rebounding From the Past: A Case Report
By Anthony Swanholm, OD

Case History

- Patient is a 28 year old male.
- Chief complaint was blurred vision with redness and discomfort while wearing hybrid lenses fit at a previous practice. Previous attempts of scleral lens wear were unsuccessful secondary to discomfort.
- The patient has a history of keratoconus OD>OS that was diagnosed at 21 years old. Vision demands include driving and work-related activities as an engineer that require correction with contact lenses versus spectacles. The patient has a history of seasonal allergies and sensitivity to penicillin and cephalosporin.

Pertinent Findings

- Entering VA with hybrid lenses: OD 20/50-1, OS 20/40-2 at distance.
- Slit lamp exam revealed 3+ injection temporally OU, Vogt’s striae OU, corneal thinning OU, central scarring OU, and temporal pinguecula OU. Goldmann applanation tonometry measured intraocular pressures of 10 mmHg OD and OS.
- The patient has substantial temporal pinguecula OU with significant hyperemia.
- Fundus exam showed no retinal pathology OU with a cup-to-disc ratio of 0.7/0.7 OD and 0.75/0.75 OS.
- Corneal topography revealed corneal irregularity beyond keratoconus OU, presumably due to corneal molding from hybrid lenses and corneal scarring. Centrally, the cornea is flattened due to corneal scarring and hybrid lens wear, as well as corneal steepening paracentrally. The horizontal visible iris diameter (HVID) is 11.4 mm.
- An optical coherence tomography (OCT) of the optic nerve head was performed due to large cup-to-disc ratio bilaterally. Multiple scans were attempted, however each scan was unreliable with limited useful information. Further glaucoma testing and evaluation is warranted at future follow-ups.

Differential Diagnosis

- Pellucid marginal degeneration
- Previous herpetic infection

Diagnosis and Discussion

This patient was diagnosed with keratoconus at a relatively young age. When keratoconus presents at an early age, the chance of progression to necessitate a corneal transplant is greater than those who develop keratoconus later in life.\(^1\) With the help of specialty contact lenses, corneal transplantation may be prevented or delayed. Scleral lenses may be selected as an initial lens or may be considered after the patient has become intolerant of other contact lens designs, prolonging corneal surgery.\(^2\) Some optometrists suggest keratoconus patients should initially be fit in scleral lenses due to better vision and comfort.\(^2\) In the 2012 Australian graft registry, they reported the average corneal transplant lasts 18 years.\(^3\) The rate of success for each subsequent full thickness replacement graft decreases with a second graft averaging 12 years,
while the third and fourth graft average 9 years. Graft survival rates also decrease each year with rates at 1, 10, and 20 years showing 97%, 89%, and 46%. By delaying a corneal transplant, the number of corneal transplants in a lifetime and the chance of failure and graft rejection are reduced. An alternative to a corneal transplant is deep anterior lamellar keratoplasty (DALK) which involves replacement of corneal stroma. DALK reduces graft rejection occurrence as it preserves the host’s natural endothelium. This procedure should be considered before a corneal transplant due to decreased rejection rates.

Keratoconus is associated with a variety of factors including genetic and environmental aspects. This patient has no family history of keratoconus but indicates seasonal allergies. Patients with allergies have a higher incidence of eye rubbing that is thought as a cause of keratoconus. Some research suggests that it may not be eye rubbing alone but it may initiate the process of keratoconus in those who are genetically predisposed. There is other speculation that keratoconus may not be allergy related. These sources describe distinct differences between allergic eye rubbing and keratoconic eye rubbing. Keratoconus patients often have a circular motion with their knuckle rubbing their eye, whereas in allergies, it is more eyelid and conjunctival rubbing with the back of the hand. What causes keratoconus is likely multifactorial and more research is needed to understand the true pathophysiology.

Anecdotally, many keratoconus patients tend to be a glaucoma suspect or have glaucoma. There are theories on why these two ocular conditions seem to be associated, but currently there is no definitive evidence. Keratoconus patients often have suspicious appearing optic nerves and low IOPs, making optic nerve appearance the rationale for further evaluation. However, optic nerve evaluation with biomicroscopy and OCT at initial and subsequent visits can be challenging due to irregular astigmatism and scarring causing limited views or unreliable OCT scans. Patients may also have reduced best corrected visual acuity, even with specialty contact lens wear, making traditional visual field testing challenging to interpret and reduces confidence in results.

In the literature, there are few formal studies about corneal molding due to improper contact lens fitting. One case study indicated corneal molding can occur with hybrid lenses even when lenses are properly fit at the initial visit. In these cases, ideally, the patient would discontinue contact lens wear, allowing the cornea to return to its natural shape. A newly designed contact lens would then be fit at a later date. It seems to happen too often in clinical practice that patients who require specialty contact lenses are so dependent on their contact lenses, they are unable to function without them. This forces the eye care practitioner to find a solution while continuing contact lens wear, often requiring multiple lens remakes and office visits.

**Treatment and Management**

The patient wore hybrid lenses for two and a half years that resulted in corneal molding. Scleral lenses were recommended to relieve any corneal bearing and allow his cornea to return to its natural state. The patient was fit in a Valley Contax Custom Elite 15.8 scleral lens designed to provide proper corneal vault, edge lift, comfort, and visual acuity. The scleral lenses were initially designed to have 300-350 microns of vault over the steepest part of the cornea, allowing additional clearance as the cornea returned to its natural state. Each lens had a sagittal depth of 4,870 microns with a standard toric peripheral curve. With a diagnostic lens over-refraction, the BCVA was 20/50 OD and 20/25 OS.
The patient returned for the one week contact lens follow-up with VA 20/50+2 OD and 20/60+2 OS in scleral lenses. Vision was acceptable for the first few hours but then became significantly reduced. The patient did not feel safe driving and returned to wearing hybrid lenses. The patient did not need to drive the following week for work, and was advised to wear his sclerals as often as he felt comfortable until next follow-up. In theory, as the cornea changes back to its natural state the tear lens underneath the scleral lens should shift accordingly as the cornea changes and the patient’s vision should not change throughout the day. This did not happen with our patient but there may be underlying factors contributing to his fluctuating vision such as incorrect lens power, clouding, or excessive vault. On follow-up exams, topography showed a gradual corneal change back to its natural keratoconic state with an unusual presentation due to central scarring.

The patient returned for his two week follow-up reporting blurred vision and discomfort with injection temporally at the pinguecula OU. To prevent pinguecula bearing, a quadrant specific peripheral curve was designed. Table 1.1 demonstrates the initial standard toric peripheral curve parameters. Table 1.2 demonstrates the quadrant specific peripheral curve lens that was designed. The lens edge lift was increased in the area of the temporal pinguecula and the lens power was refined with the next lens order. In this particular lens design each step beyond the standard peripheral curve is 30 microns with (+) as more flat and (-) as more steep. Lens quadrants (Q) are relative to the examiner and described here when there is no lens rotation. Q3 and Q1 are located on the flat meridian at 9 and 3 o’clock positions aligned with “O” lens etching. Q2 and Q4 are located at 12 and 6 o’clock positions. Again the quadrants are relative to the examiner; as an example, Q1 OD is located nasal while Q1 OS is located temporal.

Table 1.1 Standard toric peripheral curve

<table>
<thead>
<tr>
<th>Eye</th>
<th>Q1</th>
<th>Q2</th>
<th>Q3</th>
<th>Q4</th>
</tr>
</thead>
<tbody>
<tr>
<td>OD</td>
<td>+5</td>
<td>-2</td>
<td>+5</td>
<td>-2</td>
</tr>
<tr>
<td>OS</td>
<td>+5</td>
<td>-2</td>
<td>+5</td>
<td>-2</td>
</tr>
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</table>

Table 1.2 Patient’s Quadrant Specific Peripheral Curve

<table>
<thead>
<tr>
<th>Eye</th>
<th>Q1</th>
<th>Q2</th>
<th>Q3</th>
<th>Q4</th>
</tr>
</thead>
<tbody>
<tr>
<td>OD</td>
<td>+5</td>
<td>-2</td>
<td>+8</td>
<td>+1</td>
</tr>
<tr>
<td>OS</td>
<td>+8</td>
<td>-2</td>
<td>+5</td>
<td>+1</td>
</tr>
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</table>

The patient returned for his 4 week follow-up and reported very comfortable lenses but still has blurry vision with VA of 20/100 OD and 20/80 OS. Upon examination, scleral lens fit after all day wear was 150 microns OD and 200 microns OS vault over the cone with no peripheral impingement or lift-off. The patient had significant deposits on his lenses after 1.5 weeks of wear. After cleaning lenses in office, VA improved to 20/80 OD, 20/50 OS with no significant over-refraction. The patient was already using a hydrogen peroxide cleaning regimen but due to lens deposits we added Boston Advanced cleaner for use prior to hydrogen peroxide.
cleaning. We discussed with the patient we are currently at or close to reaching the best visual acuity with specialty contact lenses. Surgical options of DALK and corneal transplant were discussed as options if patient is unhappy with vision. Patient will return for follow-up appointment in 2 weeks to determine if he is happy with current level of vision in sclerals or if he wants surgery.

Corneal topography measurements were taken at initial appointment after hybrid lens wear for 2.5 years, as well as, after 1.5 months of scleral lens wear. In table 1.3, the keratometry values demonstrate significant corneal steepening as the cornea rebounded to its natural state. There still remains central corneal flattening due to central corneal scars OD and OS. Similarly, paracentral steepening remains but now presents to a greater degree.

Table 1.3

<table>
<thead>
<tr>
<th></th>
<th>OD</th>
<th>OS</th>
</tr>
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<tbody>
<tr>
<td>Initial</td>
<td>58.30 @ 166</td>
<td>58.91 @ 050</td>
</tr>
<tr>
<td></td>
<td>62.51 @ 076</td>
<td>67.85 @ 140</td>
</tr>
<tr>
<td>1.5 months Scleral Wear</td>
<td>OD 67.40 @ 020</td>
<td>OS 69.68 @ 043</td>
</tr>
<tr>
<td></td>
<td>74.59 @ 110</td>
<td>76.91 @ 133</td>
</tr>
<tr>
<td>Keratometric Change</td>
<td>OD +4.21</td>
<td>OS +10.77</td>
</tr>
<tr>
<td></td>
<td>+7.19</td>
<td>+9.06</td>
</tr>
</tbody>
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This case is currently ongoing and results will be updated as patient returns for follow-up appointments. New case information will presented at the Academy meeting.

Conclusion

Scleral lenses are a viable option for keratoconus patients who have not had success with other lenses but it is essential correct fitting techniques are appropriately applied. With keratoconus and other corneal ectasias, some argue scleral lenses should be the initial lens modality selected. Quadrant specific peripheral curves require a more advanced fit but may be necessary for those patients who have abnormally shaped scleras or pathologies.

Glaucoma and glaucoma suspects with keratoconus are difficult to manage due poor views, unreliable OCT scans, and unreliable visual fields, specifically those with advanced keratoconus and scarring. Extra care should be taken in their glaucoma management and a glaucoma referral may be indicated early in the disease process in these patients.

Specialty contact lenses are able to provide better vision in those with corneal pathologies but specialty lenses have their limitations. We are often able to achieve drastically improved visual acuity with specialty lenses compared to spectacles or soft contact lenses thus delaying other surgical options. As optometrists, it is difficult to assess at what point surgery is the best option. Eventually, the vision to perform daily activities with specialty lenses becomes unsatisfactory. Surgical options need to be explored.
Citations:


