Type I Lattice Corneal Dystrophy s/p Penetrating Keratoplasty: A Scleral Lens Success Story

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Background

- **Type 1 lattice corneal dystrophy (LCD1):** a rare hereditary bilateral disorder of the cornea
  - An autosomal dominant mutation of the TGFBI gene leads to accumulation of amyloid deposits in stroma[1]
- **Variable clinical appearance** depending on the stage of the disease[2]
  - At the onset (usually by age 10): small filamentous anterior stromal lesions, mostly central
  - With progression: lesions affect posterior stroma and opacify, eventually assuming a ground glass appearance throughout the cornea
  - By the third decade, most patient will experience recurrent corneal erosions (RCEs) and reduced VA
- **Diagnosis**:
  - Biomicroscopy examination alone is often sufficient to diagnose LCD1
  - Corneal biopsy: can be used for definitive diagnosis[3]
- **Treatment**: RCE management, phototherapeutic keratectomy and penetrating keratoplasty (PKP) in later stages[3]

Case Description – Patient MF

- 64-year-old Caucasian female, diagnosis of LCD1 OU confirmed by corneal biopsy with corneal specialist
- **Past surgical history**: penetrating keratoplasty (PKP) OD (2016), cataract extraction (CE) OU (OD 2015, OS 2022)
- **Chief complaint**: blurred vision OU with PALs despite PKP OD, difficulty adjusting to new glasses since OS CE a few months prior
- **Habitual glasses Rx with VA**:
  - OD -4.75-6.25x075 20/30- (PH 20/20-)
  - OS plano 20/50 (PH NI)
  - Add +2.50
- **Topography**
  - OD tilted corneal graft with inferior elevation and steepening (Figure 1)
  - OS irregular astigmatism (Figure 2)
- **Slit lamp examination of cornea**
  - OD: tilted corneal graft with inferior elevation, clear centrally (Figure 3)
  - OS: hazy ground glass-like lesions involving anterior and posterior stroma from limbus to limbus (Figure 4)

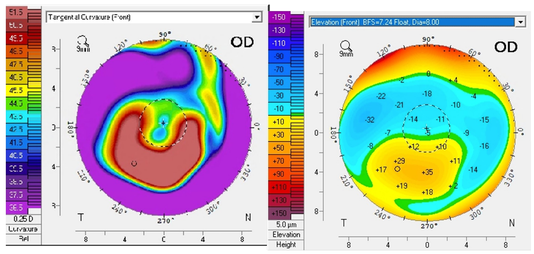

Figure 1: Topography of right eye at initial consultation visit

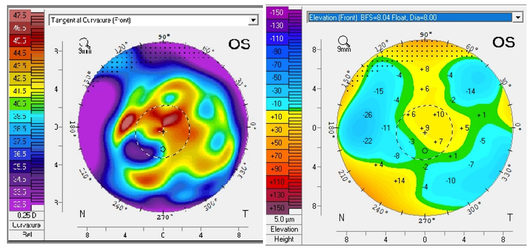

Figure 2: Topography of left eye at initial consultation visit

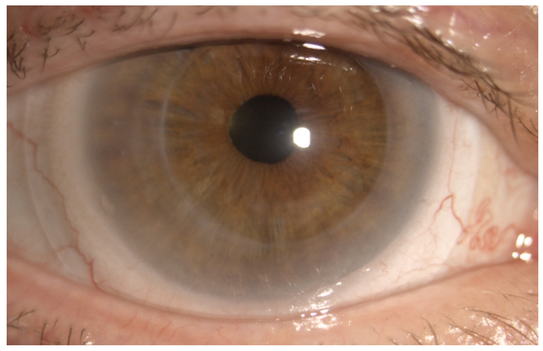

Figure 3: anterior segment photo of right eye wearing a scleral lens

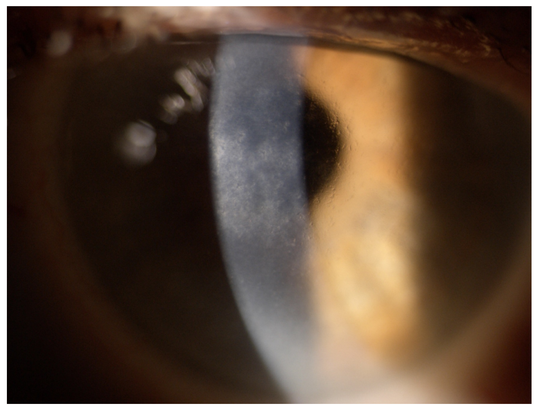

Figure 2 anterior segment photo of left eye showing lattice dystrophy

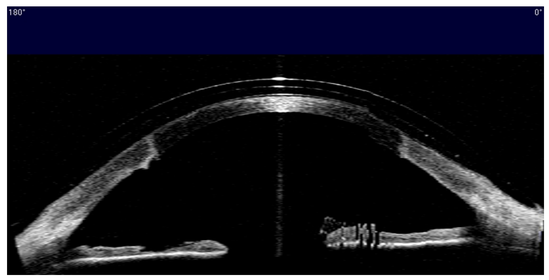

Figure 5: AS-OCT image of right eye wearing a scleral lens at two-week follow-up: excellent fitting relationship from limbus to limbus

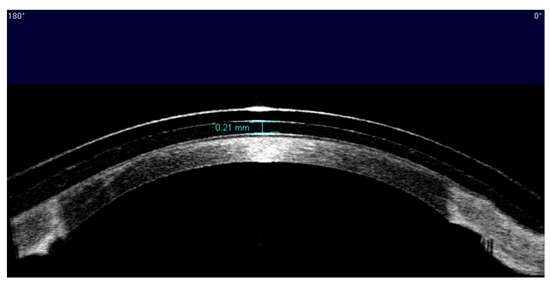

Figure 6: AS-OCT of right eye wearing a scleral lens at two-week follow-up: central clearance of 210 µm after 4 hours of wear

Scleral Lens Management OD

- A Onefit MED scleral lens (Blanchard Contact Lenses) was ordered for OD only
- **Parameters**: 4300 / 7.78 / -5.37 / 15.6 / M-100 / L +25 / TPC +25/-125 / Optimum Infinite / 0.20
- **Two-week follow-up**:
  - BCVA OD: 20/20-
  - Excellent fitting relationship (Figure 5 and 6): central clearance of 210 µm after 4 hours of wear, good mid-peripheral and limbal clearance 360, edges aligned 360
  - Patient reported clear and comfortable vision all day OD

Discussions

- For patients with advanced LCD1, scleral lenses can offer superior vision and comfort after PKP
- Scleral lenses may improve vision in an eye with corneal dystrophy: significant improvement in BCVA (0.492 logMAR) in a study with 17 eyes with corneal dystrophy[4]
  - However, patient MF was already considering having PKP done on OS before scleral lens fitting, due to having worse BCVA OS post-CE compared to pre-CE
- When only one eye has undergone PKP, scleral lens fitting success in the post-operative eye can help accelerate the decision to operate on the other eye.

References

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