Scleral Lens Fitting and Management in an Endothelial Corneal Dystrophy Patient

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Background

- Posterior polymorphous (PPD) and Fuch's (FD) are endothelial corneal dystrophies (ECD)
- Both have autosomal dominant inheritance
- Signs: guttata, edema, reduced best corrected visual acuity
- Scleral lenses (SL) enhance vision
- SL research on ECD patients is limited
- Thinnest, smallest SL with highest Dk is warranted for enhanced oxygen permeability & maintaining corneal integrity
- Baseline testing & close monitoring with specular microscopy & tomography ensure quality care

Clinical Findings

Case Description:

- 33-year old Caucasian female
- Presented for SL fit
- Current SL: 2 years old, discomfort & fogging reported

Ocular History:

- ECD (Characteristics of PPD & FD) OU
- Keratoconjunctivitis Sicca (KCS) OU

Topical Medications:

- Xiidra bid, Systane Ultra & Refresh Mega-3 prn OU

Medical History & Medications:

- Non-contributory

Family ocular History:

- ECD – mother, aunt & grandmother (maternal side)

Family Medical History:

- Non-contributory

Presenting SL distance visual acuity:

- Right Eye (OD): $20/60^{-1}$ & Left Eye (OS): $20/40^{-2}$

Biomicroscopy:

- Mild mucous debris OU
- Trace diffuse punctate epithelial erosions OU
- Significant diffuse deep stromal speckled opacities OD > OS (Figure 1)
- Grade 4 guttata OD > OS (Figures 1 & 3)

Pentacam Tomography:

- Central Corneal Thickness – OD: 707µm & OS: 670µm (Figure 2)

Specular Microscopy:

- Endothelial Cell Count (ECC) – OD: 1915 cell/mm^2
  OS: 2065 cell/mm^2
- Coefficient of Variation (CV) – OD: 73% & OS: 45%
- Hexagonal Cells (Hex) – OD: 58% & OS: 76%

Finalized Scleral Lens Parameters:

- OD: Zenlens RC/-8.75DS/7.11mm/14.8mm/Steep 2 PC/4400µm Sag/-150um LC/250µm CT/BXO2/HydraPEG
- OS: Zenlens RC/-6.50DS/7.22mm/14.8mm/Flat 3H & Flat 1V PC/4400µm Sag/-125um LC/250µm CT/BXO2/HydraPEG
- Adequate fit OU (Figures 3 & 4)
- Patient to return to clinic in 3-4 months for follow-up

Clinical Imaging

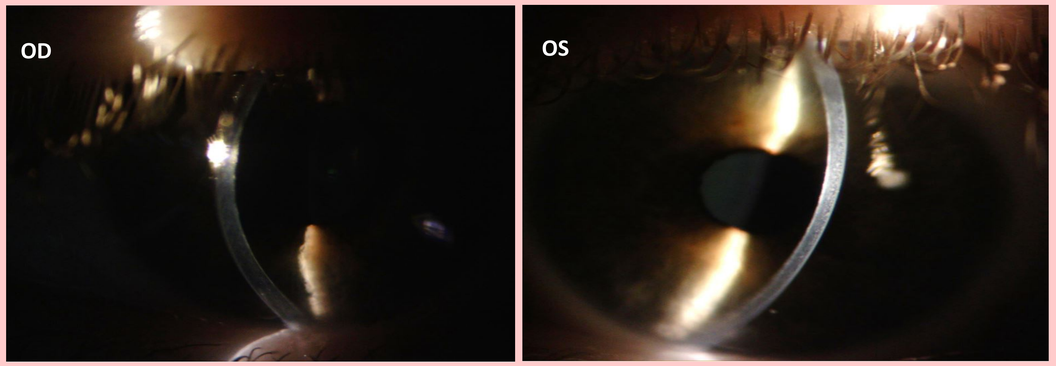

Figure 1- Corneal Images: Deep stromal speckled opacities & grade 4 corneal guttata OU

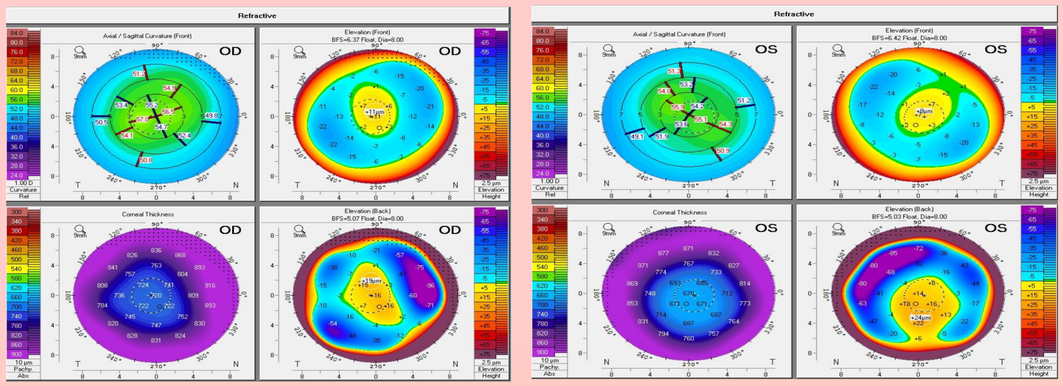

Figure 2- Pentacam Tomography: Central Corneal Thickness – OD: 707µm & OS: 670µm

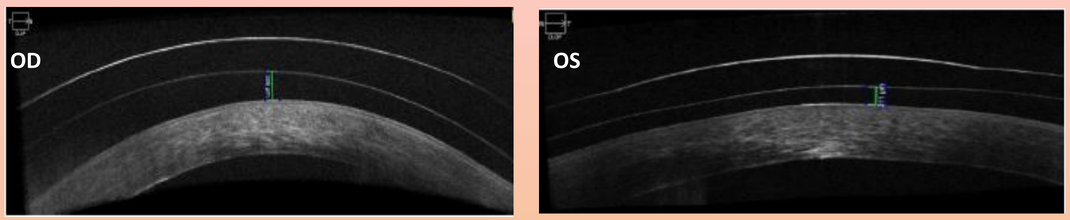

Figure 3- Final Scleral Lens OCT: Adequate central clearance OU

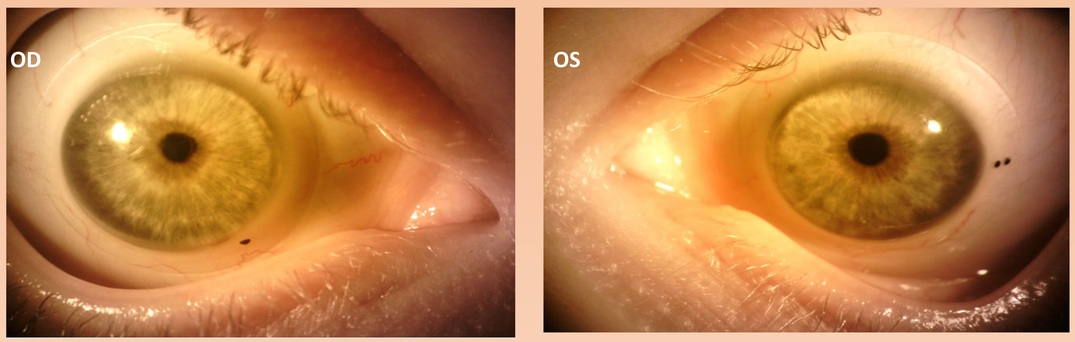

Figure 4- Final Scleral Lens Images: Adequate fit with no impingement or blanching 360 OU

Discussion

- Fitting ECD patients with SL improves vision & quality of life.
- Minimum ECC of 700 cells/mm2 (Normative ECC shown in table 1) and 50% Hex along with low CV are needed for adequate SL fitting.
- Careful SL fitting and management is warranted to maintain corneal integrity and delay keratoplasty.
- This case demonstrated the importance of fitting ECD patients with the smallest diameter, thinnest, and highest Dk SL along with ensuring adequate clearance (100-300µm) to maintain corneal health.
- This case also demonstrated the use of HydraPEG coating to manage fogging.
- Tomography and specular microscopy for baseline testing and SL management are warranted for adequate monitoring.
- Close monitoring may be necessary to avoid negative sequalae such as: corneal edema and increased intraocular pressure.

Relation between endothelial cell density and age (source: Edelhauser, 2006; Niederer, 2007).

AGE	CELL DENSITY (cell/mm^2)
10-19	2900–3500
20-29	2600–3400
30-39	2400–3200
40-49	2300–3100
50-59	2100–2900
60-69	2000–2800
70-79	1800–2600
80-89	1500–2300

Table 1: Normative Age-related Endothelial Cell Density: Decreases with age increase
Reference: Fadel D, Kramer E. Potential contraindications to scleral lens wear. Contact Lens Anterior Eye. 2019;42(1):92-103.

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