

# Sccleral Lenses for the Treatment of Corneal Dystrophies

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## INTRODUCTION

Sccleral lenses have been increasing in popularity over the past several years for not only corneal ectatic disorders such as Keratoconus and post-refractive ectasia, but also for ocular surface disease. However, little has been documented on the use of scleral lenses for corneal dystrophies. Here we present three cases—one with lattice corneal dystrophy, one with reis-bucklers corneal dystrophy, and one with granular corneal dystrophy—to show that scleral lenses can be a practical therapeutic and visual rehabilitative option for success in patients with an anterior corneal dystrophy.

## Background

Many corneal dystrophies can be classified based on patient history, corneal location, and their unique appearance. However, a few of the dystrophies share similar characteristics and may be more difficult to differentiate between. In these cases, electron microscopy with staining or genetic testing can aid in giving a definitive answer.

DYSTROPHY	LOCATION	APPEARANCE
EBMD	Epithelial	Map-Dot-Fingerprint lesions in the epithelium
Meesman	Epithelial	Numerous intraepithelial cysts
Reis-Buckler	Bowman's	Curvilinear or reticular patterned grey opacities
Thiel-Behnke	Bowman's	Sub-epithelial honeycomb-like opacities
Lattice	Stromal	Fine branching and interlaced pattern
Avellino	Stromal	Combination of lattice and granular
Granular	Stromal	Discrete, white breadcrumb-like deposits
Macular	Stromal	Numerous small, circular indistinct grey-white spots
Schnyder	Stromal	Cholesterol and phospholipid crystal deposits in the central stroma

Table 1. Differentials of the more common anterior corneal dystrophies

## Case 1: Lattice Corneal Dystrophy

### Patient History.

A 46-year-old African American male presents with Lattice Corneal Dystrophy and severe dry eyes. He experiences recurrent corneal erosions and uses Celluvisc every hour.

### Anterior Segment Evaluation.

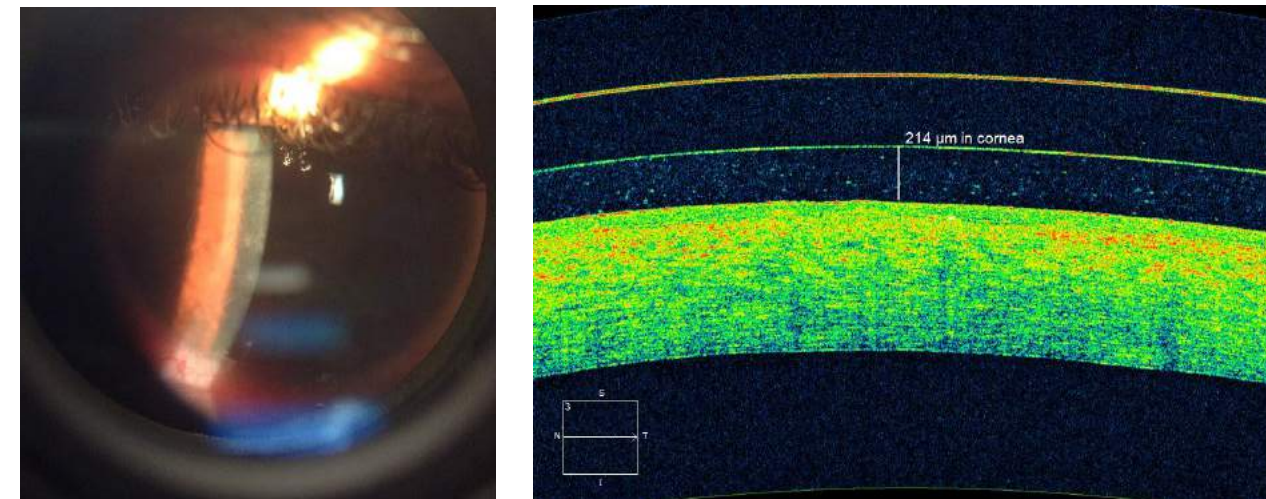


Fig 1 (a) anterior segment photograph OD (b) anterior segment OCT OD

Anterior segment evaluation showed branched opacities in the anterior stroma and 2-3+ of diffuse punctate epithelial keratitis that positively stained with sodium fluorescein.

### Topography.

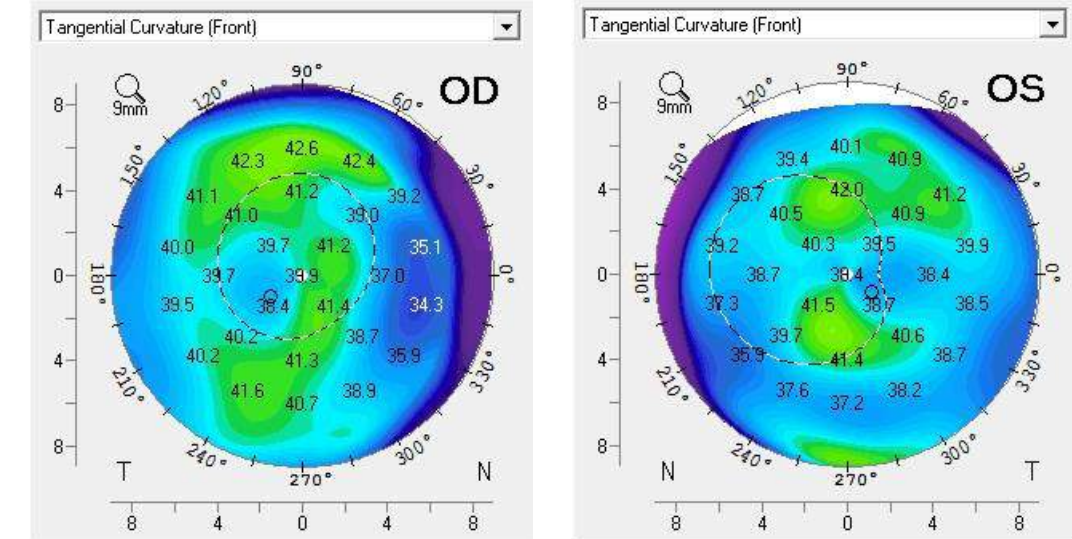


Fig 2 (a) (b) Irregular steepening corresponding with areas of amyloid deposits OD > OS

### Scleral Lens Fit.

A successful fit was achieved with Valley Contax Custom Stable Elite  
**OD:** +1.75 Sph/ 8.65 BC/ 15.80 Dia/ Std LCZ/ +2 L-Lite/ +4, -2 SLZ/ Clear  
**OS:** +0.25 Sph/ 8.65 BC/ 15.80 Dia/ Std LCZ/ +2 L-Lite/ +4, -2 SLZ/ Blue  
 Material: Optimum Extra with HydraPeg

### 1 Month Follow-Up.

	Initial	1 mo scleral lens wear
<b>Visual Acuity</b>	OD: 20/60- (BSCVA) OS: 20/50- (BSCVA)	OD: 20/20-3 OS: 20/20-1
<b>Drops used</b>	Celluvisc q1h OU Refresh PM QHS OU	Refresh PM QHS OU
<b>Corneal Staining</b>	2-3+ diffuse PEK OU	Tr diffuse PEK OU

## Case 2: Reis-Buckler's Corneal Dystrophy

### Patient History.

A 66-year-old Caucasian female is here for a scleral lens fitting for irregular astigmatism status-post LASIK. She had undergone uneventful LASIK surgery 5 years prior and noted gradually decreasing vision over the past few years. Additionally, the patient reported a constant burning in both eyes throughout the day that minimally improved with Systane artificial tears. Ocular history was positive for Reis-Bucklers.

### Anterior Segment Evaluation.

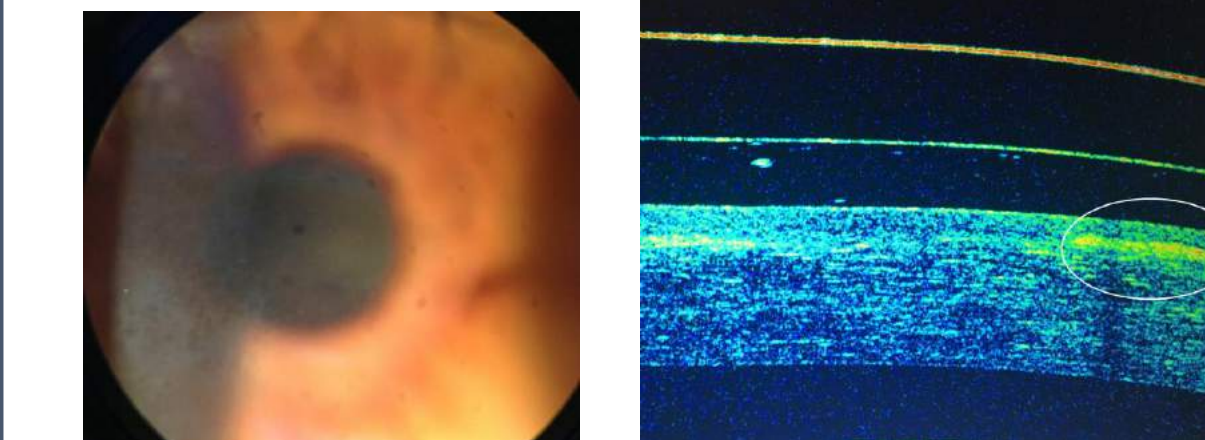


Fig 3 (a) anterior segment photograph OD (b) anterior segment OCT OD

Anterior segment evaluation revealed a diffuse central corneal haze in a scattered curvilinear or reticular pattern extending through the posterior epithelium and Bowman's layer. Anterior segment optical coherence tomography showed hyper-reflective deposits at the Bowman's layer.

### Topography.

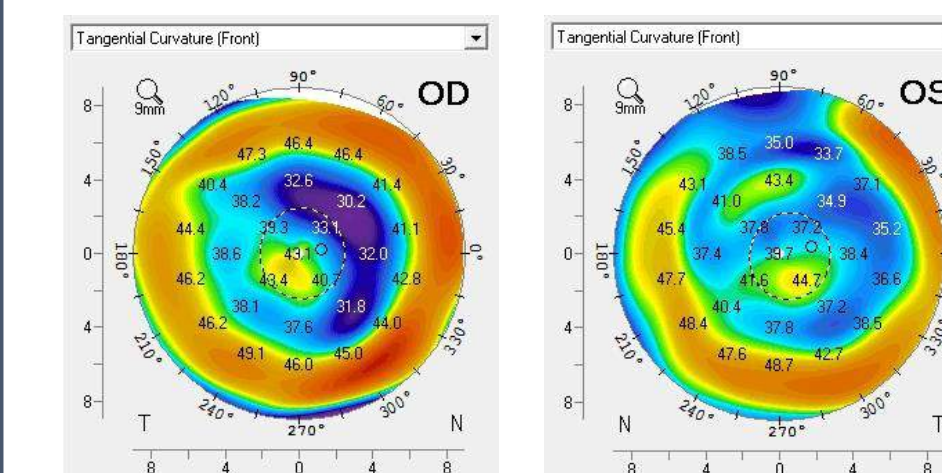


Fig 4 (a) (b) infero-central island of steepening OD > OS

### Scleral Lens Fit.

A successful fit was achieved with Blanchard OneFit 2.0  
**OD:** +2.75 Sph/ 8.10 BC/ 14.90 Dia/ XLC/ 1 Flat Edge Lift/ Clear  
**OS:** +4.50 Sph/ 8.10 BC/ 14.90 Dia/ XLC/ 1 Flat Edge Life/ Blue  
 Material: Optimum Extra

### 1 Month Follow-Up.

	Initial	1 mo scleral lens wear
<b>Visual Acuity</b>	OD: 20/60- (BSCVA) OS: 20/50- (BSCVA)	OD: 20/20-3 OS: 20/20-1
<b>Corneal Staining</b>	2+ central and infero-central PEK OU	Tr inf PEK OU

## Case 3: Granular Corneal Dystrophy

### Patient History.

A 62-year-old Hispanic male is referred for a scleral fitting for irregular astigmatism. Ocular history was positive for Granular Corneal Dystrophy Type I. He has undergone Phototherapeutic Keratectomy in both eyes in 2004 and in the right eye again in 2012. He presented complaining of photophobia and a gradual decrease in vision over the past few years. He was interested in vision improvement to allow him to drive again.

### Anterior Segment Evaluation.

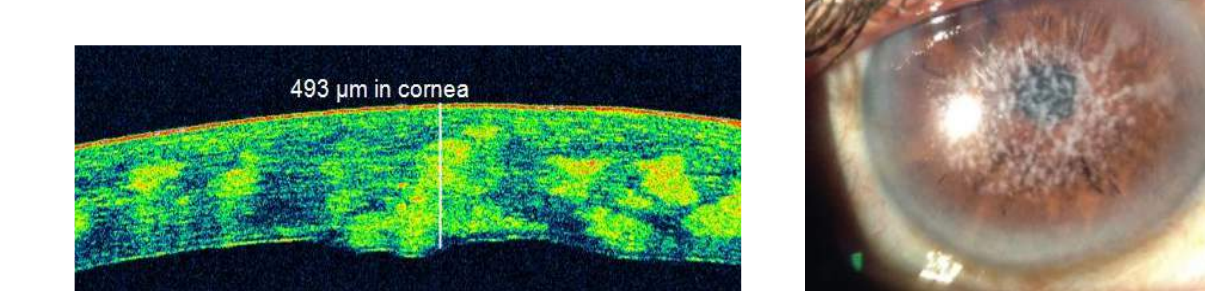


Fig 5 (a) anterior segment OCT OD (b) anterior segment photograph OD

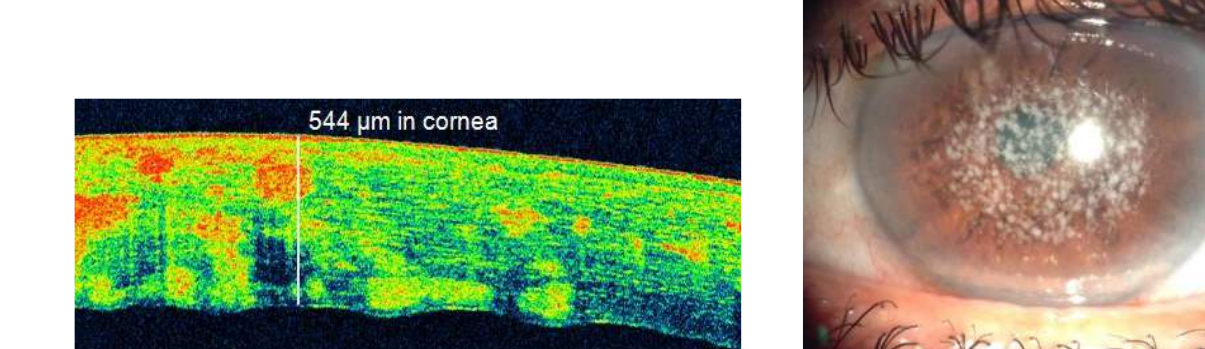


Fig 6 (a) anterior segment OCT OS (b) anterior segment photograph OS

Anterior segment evaluation revealed discrete white deposits in the central cornea. Anterior segment optical coherence tomography showed scattered hyper-reflective deposits within the stroma.

### Topography.

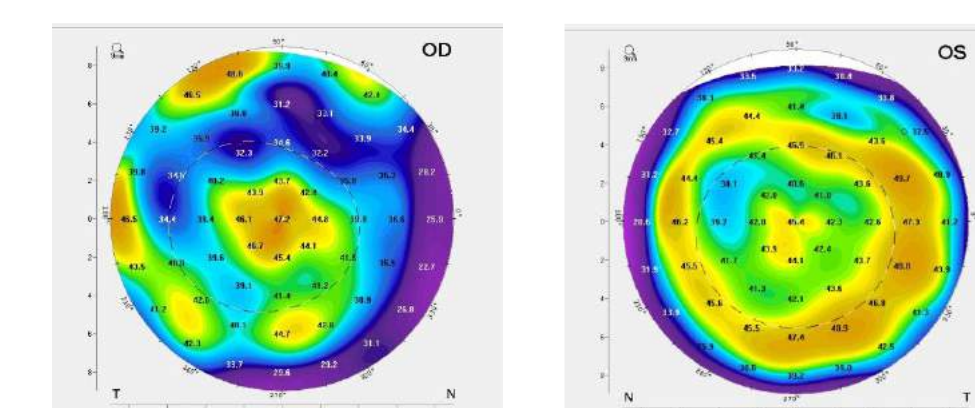


Fig 7 (a) Irregular surface OD (b) Central oblate area with irregularities OS

### Scleral Lens Fit.

A successful fit was achieved with Xcel Atlantis  
**OD:** +1.50 -2.25 x040/ 7.67 BC/ 15.0 Dia/ 2 Steep LZ/ Std SZ/ Black Dot  
**OS:** +0.50 -1.00 x095/ 7.67 BC/ 15.0 Dia/ 1 Steep LZ/ Std SZ/ Red Dot  
 Material: Optimum Extreme

### 1 Month Follow-Up.

	Initial	1 mo scleral lens wear
<b>Visual Acuity</b>	OD: 20/100 (BSCVA) OS: 20/100 (BSCVA)	OD: 20/60-2 OS: 20/60+2

## Conclusion

Treatment for anterior corneal dystrophies is mainly aimed at preventing recurrent corneal erosions and dryness. The irregular corneal surface due to the deposits can cause uneven tear film distribution and pockets of desiccation. Initial therapy may include artificial tears and nighttime ointments. A bandage contact lens and antibiotics may be added depending on the severity of the corneal erosions. In certain cases, surface debridement may help clear the corneal epithelium of irregular tissue, paving the way for improved healing. In severe cases, surgical intervention such as superficial keratectomy or phototherapeutic keratectomy may be necessary to remove the deposits. However, there can be recurrence of the deposits.

Scleral lenses can help relieve the dryness by providing a fluid reservoir of continuous lubrication for the cornea. Additionally, the lens can act as a barrier to protect against mechanical shearing forces. Lastly, the scleral lens can correct any resulting irregular astigmatism by providing a new, smooth refracting surface.

## Take-Home Points

- ❖ Corneal dystrophies can be classified based on their location and appearance
- ❖ A definitive diagnosis can be made with staining on electron microscopy or genetic testing
- ❖ Corneal dystrophies can contribute to ocular surface disease and dry eyes due to uneven tear film distribution
- ❖ Corneal dystrophies can lead to corneal irregularities from deposits and recurrent corneal erosions
- ❖ Superficial keratectomy or phototherapeutic keratectomy can temporarily alleviate the symptoms by removing the deposits, but in many cases they will recur over time
- ❖ Scleral lenses can be an effective treatment for both dryness and irregular corneal astigmatism

## REFERENCES

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