

The Role of Corneal GPs in Managing Irregular Astigmatism Associated With Ehlers Danlos Syndrome

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Introduction

Ehlers Danlos Syndrome (EDS) is a genetic condition that affects connective tissue throughout the body, including the eyes. Common ocular findings associated with EDS include keratoconus and other corneal ectasias, blue sclera, high myopia, retinal detachment, and dry eye.¹ This report discusses how GP lenses can offer improved visual quality and ocular comfort without surgical intervention.

Case History

JK is a 28-year-old female referred for a scleral lens fit due to unstable vision with spectacle lens wear. JK was a former physical therapy student who relinquished her professional goals due to various systemic manifestations of EDS. JK has a complicated ocular history, symptomatic for blurry vision, migraines and history of idiopathic intracranial hypertension (managed with oral acetazolamide). JK has no history of ocular surgeries and she currently uses autologous serum drops to manage symptoms of dryness with little success. JK's best corrected VAs with spectacles were 20/100 OD and 20/80 OS. JK reported 20/15 OU VAs two years prior, but vision has progressively worsened and fluctuated since.

Clinical Findings

	OD	OS
Entering VAs sc	20/100	20/80
Conjunctiva/Sclera	White and quiet	White and quiet
Cornea	2+ inferior PEE, (-)Vogt's striae/scarring/Fleischer ring. Unstable tear film. Prominent vertical nerve fibers on central cornea	2+ inferior PEE, (-)Vogt's striae/scarring/Fleischer ring. Unstable tear film. Prominent vertical nerve fibers on central cornea
Pachymetry	489 microns	486 microns



Figure 1. Corneal topographies showed irregular superior steepening and associated thinning, indicating superior corneal ectasia on both eyes.

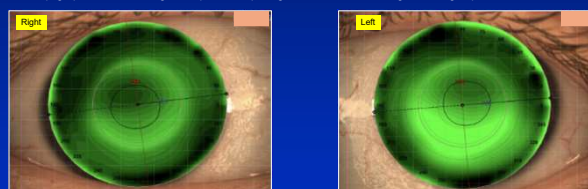


Figure 2. Custom designed toric corneal GP lens simulated NaFl patterns on JK's right and left eyes

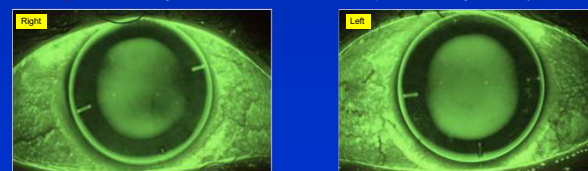


Figure 3. Quadrant specific, custom GP lenses on JK's right and left eyes

Management

JK's hand tremors and muscle weakness made scleral lenses difficult to handle. Larger diameter, quadrant specific corneal GPs were designed and dispensed at the 1 month follow up. Larger diameter corneal GPs were designed with an apical clearance of approximately 40 microns, with a peripheral alignment zone similar to that of an orthokeratology lens. The goal of this design was to improve GP lens comfort and create a tear reservoir that could minimize symptoms of ocular dryness. JK was elated to have corrected VAs with the lenses of 20/25-1 OD and 20/25-2 OS and reported a decrease in dry eye symptoms.

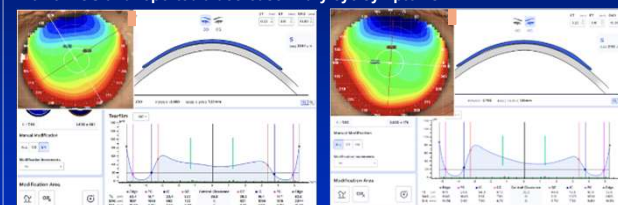


Figure 4. JK's elevation display maps and third party lens designing software used to design JK's custom quadrant specific GP lenses

Conclusion

Corneal GP lenses improve vision for individuals with irregular corneas or complicated refractive errors compared to spectacles. With advancements in design and lathing technology, today's corneal GPs can be highly customized to match a patient's corneal topography, providing the optimal health benefits and optics of a traditional GP while also maximizing comfort and fit.

References

- Villani E, Garoli E, Bassotti A, Magnani F, Tresoldi L, Nucci P, Ratiglia R. The cornea in classic type Ehlers-Danlos syndrome: macro- and microstructural changes. Invest Ophthalmol Vis Sci. 2013 Dec 11;54(13):8062-8. doi: 10.1167/iov.13-12837. PMID: 24168998.