# Management of Ocular Surface Disease in a Patient with Ehlers-Danlos Syndrome

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## BACKGROUND

Ehlers-Danlos Syndrome (EDS) is a connective tissue disorder that affects several organs and tissues throughout the body. Although the prevalence varies based on the form of Ehlers-Danlos, the most common forms, the hypermobile type and the classic type, show a prevalence of 1 in 5000 to 20,000 and 1 in 20,000 to 40,000 respectively.[1]

Several ocular structures are affected by this condition including the lids, cornea, lens, vitreous and retina.[2-3] Although the structural effect of this condition on the eyes is often the topic of interest, it is important to recognize the effect it can have in causing ocular surface disease.

In two separate studies, Magda Gharbiya et al[2] and Edoardo Villani et al[3] (Table 1), compared the eyes of hypermobile type EDS and classic type EDS against a control of normal eyes respectively. Each study found that patients with their respective type of EDS had quicker tear break up time, decreased Schirmer I test, and higher scores on the Ocular Surface Disease Index (OSDI) as compared to their control subjects. Magda Gharbiya et al theorized that there may be an autonomic component in the hypermobile type EDS in causing a decrease in tear production. There also may be a developmentally impaired mechanism of regulating lacrimal gland secretion. Edoardo Villani et al[3] theorized that these differences in classic EDS are brought on by "altered fibroblasts and collagen matrix on the proliferation and differentiation of ocular surface epithelial cells."

Several studies, including one by Alipour et al[4], have shown that scleral lenses are effective in treating moderate to severe dry eye. This treatment therapy could also provide needed relief for patients dealing with dry eye secondary to EDS.

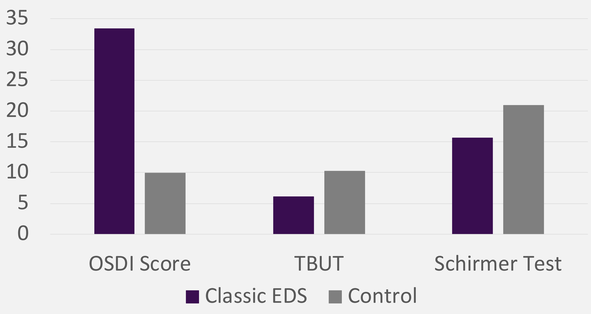
## CASE HISTORY

- A 55-year-old white female presented to our clinic with a chief complaint of dry eye
  - Location: Both eyes
  - Duration: Longstanding
  - Symptoms reported as severe and getting worse
  - Modifying factors: see Table 2
  - Associated symptoms: migraines
- Pertinent patient medical history:
  - (-) for Sjogren's Syndrome
  - Recently diagnosed with Ehlers-Danlos Syndrome

## EXAM FINDINGS

- Entering VAcc:
  - OD: 20/20
  - OS: 20/25
- Anterior Segment Findings:
  - Occluded puncta OU
  - 2+ punctate staining OU
  - Decreased TBUT OU
  - Schirmer's Test w/o anesthesia:
    - OD: 5mm after 5 minutes
    - OS: 5mm after 5 minutes

**Table 1* - Dry Eye in Classic EDS vs Control**




*Data taken from the study performed by Edoardo Villani et al[3]

**Table 2 - Pre-Scleral Lens Therapy**

| Treatment Therapies: | Reported Dry Eye Symptoms: |
|---|---|
| • Nighttime ointment QHS<br>• Artificial tears 4-6x/day<br>• Restasis BID<br>• Hot compresses x 10 min BID<br>• Silicone punctal plugs* | • Severe:<br>-Gritty<br>-Burning<br>-Pain<br>-Migraines from dryness |

*Patient requested the punctal plugs be removed due to irritating her eyes

## TREATMENT

- Patient was successfully fit in 15.6mm Onefit MED scleral lens with toric peripheral curves
- The final lens demonstrated good clearance and landing

## FOLLOW UP

At follow up our patient reported that her eyes were feeling much better after wearing her scleral lenses.

## CONCLUSION

Our patient found significant relief from her dry eye symptoms through scleral lens therapy. Use of scleral lenses is an effective treatment option for patients with EDS and should be considered if other treatment options are failing.

## REFERENCES

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2. Magda Gharbiya, Antonietta Moramarco, et al.; Ocular Features in Joint Hypermobility Syndrome/Ehlers-Danlos Syndrome Hypermobility Type: A Clinical and In Vivo Confocal Microscopy Study, American Journal of Ophthalmology, Volume 154, Issue 3, 2012, Pages 593-600.e1, SSN 0002-9394, https://doi.org/10.1016/j.ajo.2012.03.023.
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