



Management Options For Pediatric Advanced Keratoconus

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PURPOSE

Keratoconus is a chronic, progressive, non-inflammatory, bilateral but often asymmetric corneal ectasia. Although the exact cause of Keratoconus is debatable, genetics and environment play major roles. Symptomology and treatment vary depending on severity of the disease and the level of visual acuity lost. Options for visual improvement include: spectacles in the early stages of the disease and a growing array of specialty contact lenses. Medical and surgical interventions include: corneal collagen crosslinking to slow disease progression, INTACS to alter corneal shape and, in most severe cases, lamellar and full-thickness corneal transplants. This case will discuss management of advanced Keratoconus in pediatric patient suffering from Vernal Keratoconjunctivitis

PERTINENT HISTORY & EXAM FINDINGS

11-year-old middle-eastern male with history of advanced Keratoconus and unsuccessful corneal gas-permeable lens wear presents for consideration of various surgical and contact lens treatment options. Ocular history is also significant for chronic allergic conjunctivitis that has been treated in the past with unknown topical drops. Patient denies any systemic history. Family history is positive for older sister with mild Keratoconus in both eyes and history of allergic conjunctivitis..

Best corrected spectacle acuity is 20/400 PH 20/100 in both eyes. Patient presents with 4x4mm central apical corneal scarring in the right eye and dense Vogt's striae and severe stromal thinning centrally in both eyes. Slit lamp examination also reveals peripheral arcuate opacification in superficial stroma (Figure 3). This peripheral corneal finding known as pseudogerontoxon confirmed that this patient has been suffering from Vernal Keratoconjunctivitis.¹ Dilated fundus exam was unremarkable.

Treatment options such as corneal cross-linking, scleral contact lens fitting, and corneal transplant were discussed with patient and parents. This case was co-managed with corneal specialist who, despite advanced stage of the disease and corneal scarring in the right eye, elected to perform collagen-crosslinking on both eyes.

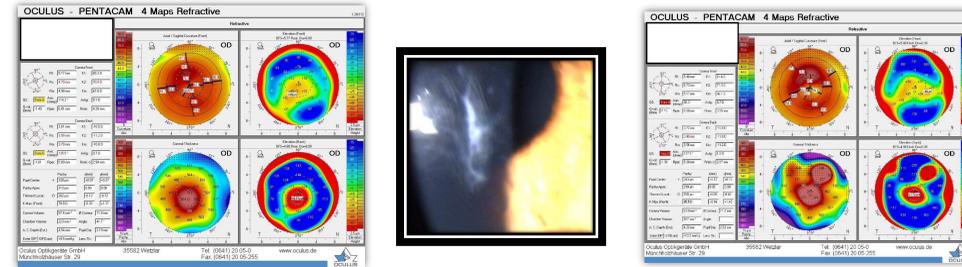


Figure 1. OD: corneal topography pre-cross linking(left) and post- cross linking (right). Center images shows apical scarring and Vogt's striae

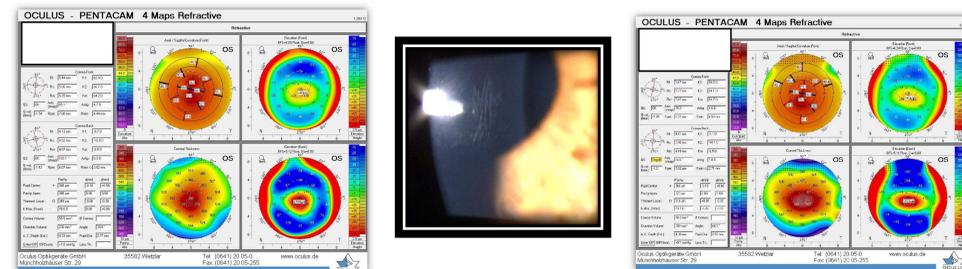


Figure 2. OS: corneal topography pre-cross linking(left) and post- cross linking (right). Center images shows Vogt's striae and absence of scarring

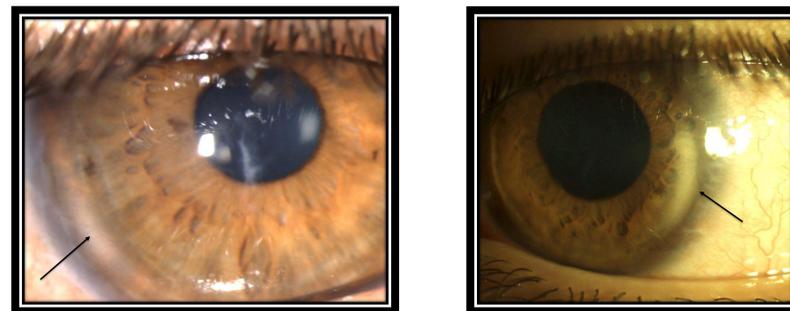


Figure 3. Inferotemporal pseudogerontoxon OD and OS consistent with Vernal Keratoconjunctivitis

To improve visual acuity, the patient was fit with small diameter scleral lens in both eyes achieving best corrected vision of 20/70 in the right eye and 20/50 in the left eye. A regular cornea scleral lens design was used as opposed to irregular cornea design due to patient's small palpebral aperture size and difficulty with insertion of lenses.

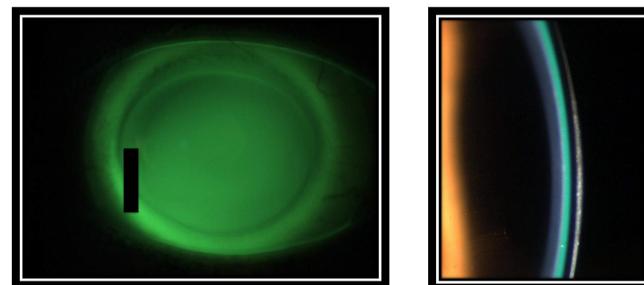


Figure 4. Scleral lens vaulting the cornea

Due to visual improvement with scleral lens and patient's young age, corneal transplant surgery was not advised at this time.

DISCUSSION

Keratoconus is multifactorial in nature. Although pathogenesis of Keratoconus is still unknown, both environmental and genetics play a role in this primary corneal ectasia. In the case of our patient, both him and his older sister have been diagnosed with Keratoconus. Both siblings also suffered from Vernal Keratoconjunctivitis as confirmed by ocular history and presence of pseudogerontoxon on slit lamp examination in both patients. As the disease waxes and wanes, Vernal Keratoconjunctivitis can pose a contact lens fitting challenge and active episodes lead to temporary discontinuation of lens wear.

One of the main symptoms of Vernal Keratoconjunctivitis is ocular pruritus which often leads to eye rubbing. There is evidence that shows eye-rubbing associated with allergen exposure and high levels of immunoglobulin E contributes to the progression of Keratoconus.² This puts our patient at increased risk for progression.

The patient underwent corneal collagen-crosslinking in both eyes to slow down the rapid progression of his Keratoconus. Standard epi-off corneal collagen-crosslinking has been shown to be safe and effective to halt the progression of Keratoconus in a pediatric population at 3-year follow-up.³

CONCLUSION

Despite the advanced nature of Keratoconus and history of Vernal Keratoconjunctivitis, the patient successfully underwent collagen-crosslinking in both eyes and was fit with scleral lens. Due to advancements in surgical and contact lens options, this patient did not have to undergo a corneal transplant at such a young age. However, close monitoring of lens fit is warranted due to progressing Keratoconus and presence of Vernal Keratoconjunctivitis.

REFERENCES

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