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OBJECTIVE

To present the diagnosis and management of unilateral posterior keratoconus with bilateral iris atrophy in a child.

INTRODUCTION

Posterior keratoconus (PKC) is a rare non-inflammatory ectatic corneal disorder. It is often present at birth and is unrelated to anterior keratoconus. PKC has been postulated as a mild form of Peter's anomaly. Corneal findings often include stromal thinning, steepening of the posterior corneal surface, and scarring. However, upon careful examination many other ocular features are present such as a shallow anterior chamber, iridocorneal adhesions, iris atrophy, anterior polar cataract, posterior embryotoxon etc. Currently, the etiology and pathogenesis of this disease is unknown. That is why case reports such as the one being presented is essential and can provide insight for future research.

CASE

A 4-year-old Hispanic female referred to the University Eye Center for CL fitting OS. Ocular history includes degenerative myopia OS, anisometropia, posterior keratoconus OS, and left esotropia. Patient had strabismus surgery at 3 years of age without complication.

Ocular medication includes atropine 1% QHS OU with poor compliance. Medical history includes heart murmur and iron deficiency anemia. Mother denies birth trauma or complications during pregnancy. No family history of keratoconus reported.

Visual acuity in spectacles:

OD: +2.00 -1.75 x 005 VA: 20/40
OS: -11.75 -4.00 x 011 VA: 20/800
ADD +3.00 (OS only)

CL Fitting:

Initially a GP lens was trialed (BC 9.0mm, DIA 10.50mm), which decentered superior with excessive edge lift. Overall, patient was unsuccessful with GP fit due to poor tolerance of lens comfort. As a result, a soft contact lens (-10.00sph) was utilized. BCVA OS was 20/200 in soft CL compared to 20/100 in GP. Polycarbonate spectacles to be worn over CL OS was prescribed: same Rx OD, plano with +3.00 ADD OS.

REFERENCES: 1. Gus, Patricia Ioschpe, et al. "Posterior Keratoconus and Iris Atrophy: a Fortuitous Association?" *Arquivos Brasileiros De Oftalmologia*, vol. 82, no. 1, 2019, doi:10.5935/0004-2749.20190014.

2. Olivo-Payne, Andrew, et al. "Optimal Management of Pediatric Keratoconus: Challenges and Solutions." *Clinical Ophthalmology*, Volume 13, 2019, pp. 1183–1191., doi:10.2147/oph.s183347.

3. Silas, Megan R, et al. "Posterior Keratoconus." *British Journal of Ophthalmology*, vol. 102, no. 7, Sept. 2017, pp. 863–867., doi:10.1136/bjophthalmol-2017-311097.

CLINICAL FINDINGS

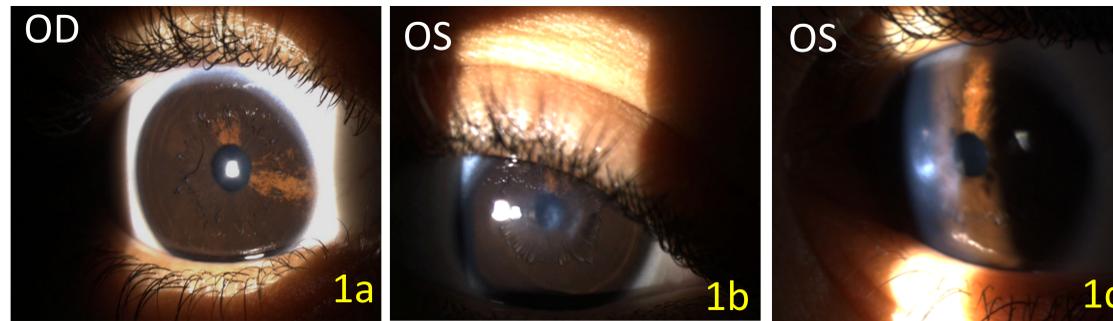


Figure 1: Slit lamp photos describing anterior segment findings: posterior embryotoxon located superior nasal OD (1a), bilateral iris atrophy OD>OS (1a, 1c) and central endothelial/stromal scar 3x3mm OS (1b, 1c)

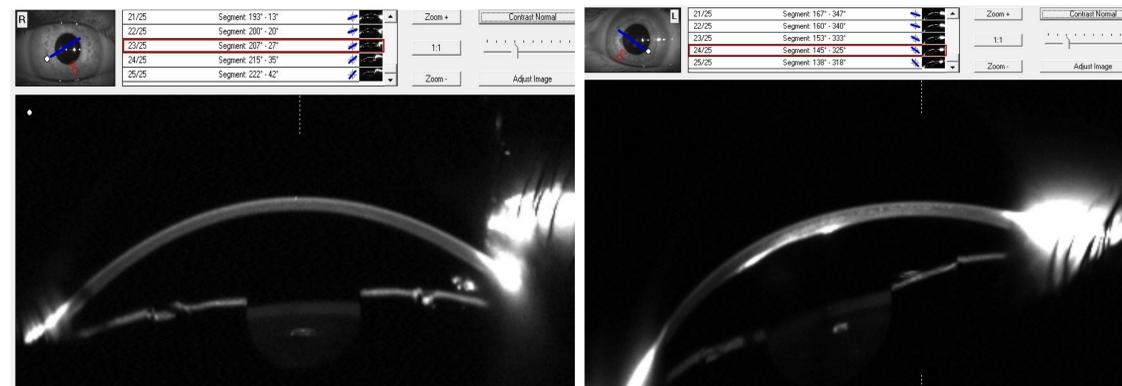


Figure 2: Scheimpflug image of the right eye show clear corneal layers. Image of the left eye reveal a hyperreflective thinner posterior corneal surface secondary to scarring. An anterior polar cataract can also be observed.

DISCUSSION

Due to patient's young age, adapting to a soft CL was less of a challenge compared to a GP lens and visual acuity was comparable between the two lens modality. The purpose of CL fitting OS was to reduce anisometropia with the current spectacle. Another priority was to initiate patching to reduce risk of refractive amblyopia. Reduction in vision can be attributed to central cornea scar and amblyopia. Due to the central location of the scar, surgical intervention such as penetrating keratoplasty is a plausible option. However, when considering the patient's age, the risks and benefits of surgery a conservative initial intervention is often favored.

CONCLUSION

PKC can occur early in childhood with a unilateral presentation. Typically it is a non-progressive corneal disorder; however, regular monitoring is warranted and anterior keratoconus should be a differential diagnosis. There have been reported cases of PKC in one eye and anterior keratoconus in the other eye. For patients with anisometropia and significant astigmatism, corneal topography is an important tool highlighting early corneal irregularities. Pediatric keratoconus tends to yield in poor visual outcomes, therefore early intervention such as corneal cross-linking (CXL) is warranted.

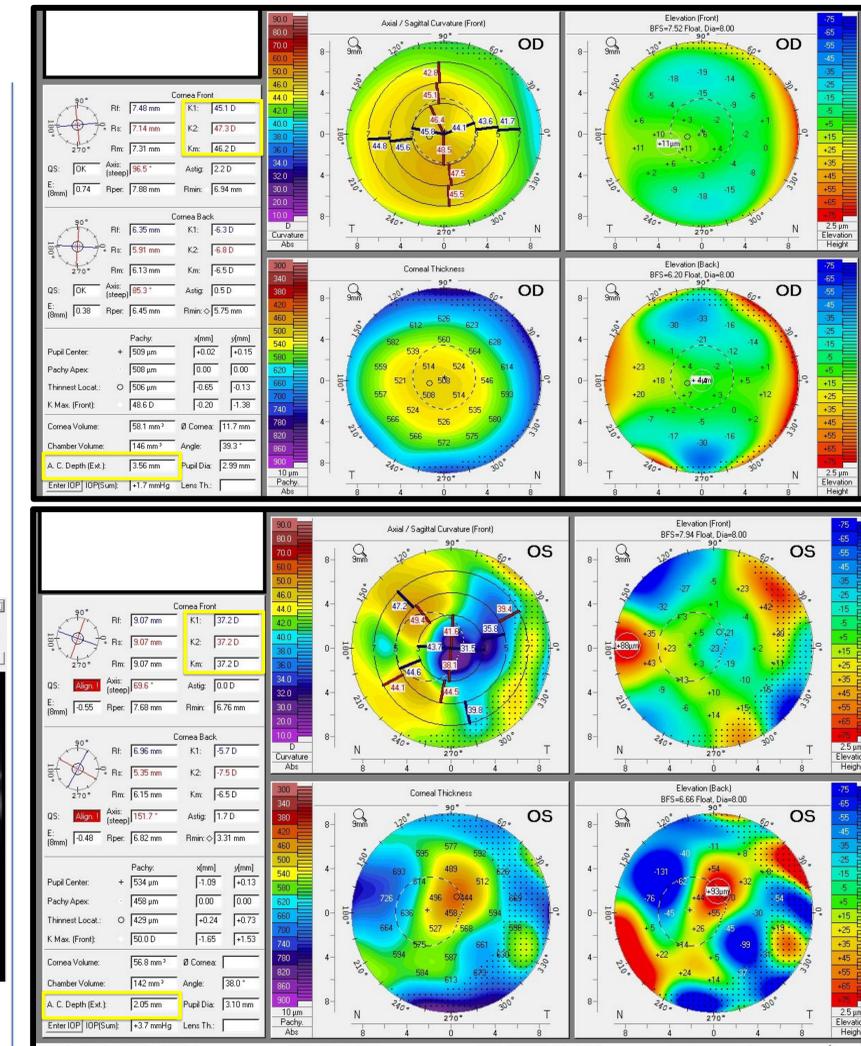


Figure 3: Baseline corneal topography reveals posterior steepening evident OS>>OD. A shallow anterior chamber depth is noted in the left eye.