Scleral Lens Visual Rehabilitation After Selective Endothelial Removal in Peters Anomaly

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Introduction

- Peters Anomaly Type I is a rare congenital anterior segment dysgenesis. It is diagnosed on clinical findings of corneal opacity, corneal edema, and loss of posterior stroma, Descemet's membrane, and endothelium.
- The presentation is highly variable in severity and bilateral in 80% of cases.[1]
- Type I is classified with central corneal opacity and iridocorneal adhesions. Type II has central corneal opacity and cataracts or corneolenticular adhesions. Peters-plus syndrome is characterized with PA and development delay, short stature, dysmorphic facial features and cardiac and genital abnormalities.
- Children are at risk of amblyopia and sensory nystagmus. Nystagmus has been reported at 57.5%.[3,] was higher in bilateral disease and was found to be more common with history of PKP.[3]
- A new surgical technique, Selective Endothelial Removal (SER), has been shown to improve corneal clouding. Gas permeable corneal lenses (GPCL) or impression based scleral lenses (IBSL) can aid in visual maturation.

Patient presentation

History of Present Illness

- 0-day old infant male was seen for consultation in the hospital after uncomplicated spontaneous vaginal delivery for concern of corneal clouding
- He was born full term and otherwise healthy and doing well

Family history

- No family history of ocular disease and no maternal risk factors

Social history

- No prenatal drug exposure

Physical exam

- The right cornea is significant for central haze that extends to the inferior limbus (Figure A)
- The left cornea has inferocentral haze sparing superior, temporal and nasal cornea (Figure A)
- The anterior chamber of the right eye shows iridocorneal adhesions inferior, temporal and nasal regions
- The left eye has iridocorneal adhesions mostly inferonasal
- There is no sign of cataract in either eye
- Cycloplegic retinoscopy is +6.50 right eye and +7.50+1.00x180 left eye.

Lab results

- TORCH titers are negative

Imaging

- B-scan shows anterior synechiae from iris to cornea with flat and attached retina and normal posterior anatomy both eyes (Figure B)

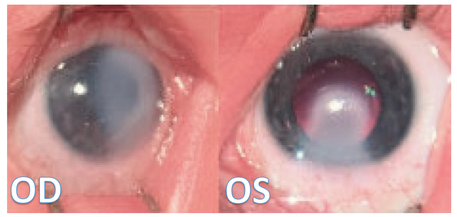

Figure A: External photograph taken at age 4 days of the initial presentation of corneal scarring and clouding

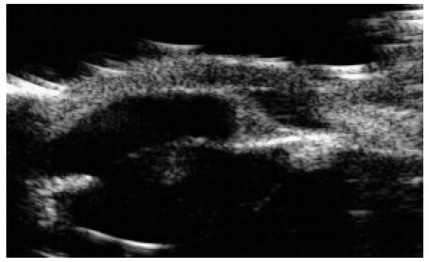

Figure B: Anterior segment B-Scan ultrasound OS at age 4 days with anterior synechiae from iris to cornea with anterior lens opacity underlying corneal opacity

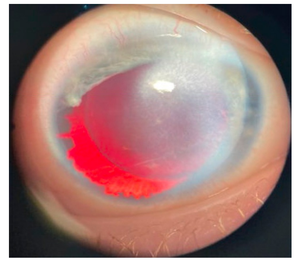

Figure C: Image illustrating patient's optical iridectomy right eye

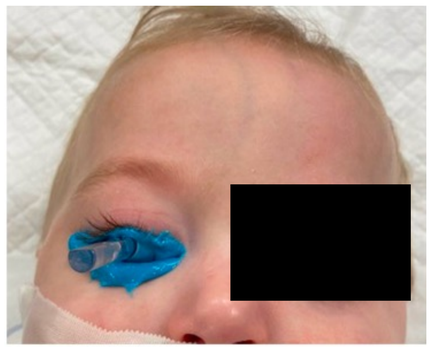

Figure D: Imprinting for scleral lenses at age 10 months

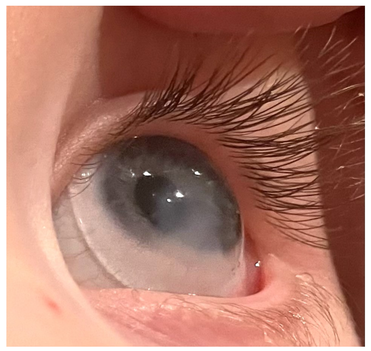

Figure E: Left eye with scleral lens

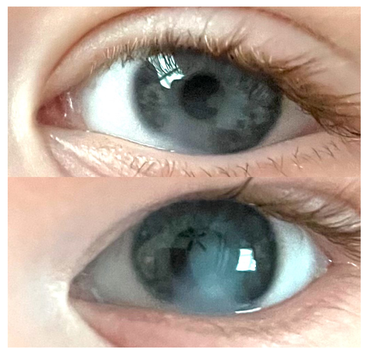

Figure F: Left eye top image 14 months. Left eye bottom image 6 months

Clinical Course

- Started on Cyclopentolate/phenylephrine 2 times per day both eyes in the hospital and continued when discharged
- Developed sensory nystagmus by 3 months of age
- Fit with gas permeable contact lenses at 5 months of age and wore glasses when not wearing contacts, patch therapy started based on asymmetric presentation – patch OS 4 hours per day, later increased to 2 days/week
- At 6 months of age, there was no improvement of R corneal opacity, parents elected to have an optical iridectomy from 6:00 to 8:30 to create an alternative visual axis (Figure C). Underwent selective endothelial removal (SERM) of the right eye to aid in clearing scarring.
- At 9 months of age (3 months post-SERM and iridectomy), the right cornea showed significant improvement of the corneal opacity and improved red reflex
- At 10 months old he was fit with impression based scleral lenses (IBSL) to optimize the optical zone given the optical iridectomy of the right and pharmacological dilation of the left. (Figure D)
- The right lens had a base curve of 7.979, power +9.75, and diameter 15.0 mm. The left lens had a base curve of 7.911, power +11.25, and diameter 15.0 mm
- 2 weeks post distribution of IBSL (Figure E), patient's parents reported more independence in the patient
- At 12 months old his corneal clarity continued to improve in both eyes, his nystagmus improved, and he became more visually independent
- At 13 months reverse amblyopia and esotropia developed in the OS (patched eye) and OD started to be patched 1 full day per week
- At 14 months, corneal clouding continued to improve with use of IBSL (Figure E)
- At 19 months old he is tolerating his ISBL very well and has continued visual rehabilitation with increased independence

Discussion

- Selective endothelial removal (SERM) is a newer procedure that has shown to reduce the stromal opacity.[4] Unhealthy endothelium is debrided while preserving intact Descemet's membrane. Healthy endothelial cells enlarge and migrate allowing normal functionality of the endothelium.[5]
- Penetrating Keratoplasty (PKP) is the main treatment for PA. Graft survival and visual acuity are related to the indication with corneal neovascularization and lens abnormality posing the greatest risk[6]
- Gas permeable contact lens or scleral lenses can correct high ametropias and irregular astigmatism. Impression-based scleral lenses are an alternative to smaller contact lenses that may better suit those with high refractive error and nystagmus in order to achieve better visual performance
- An essential part of visual rehabilitation is monitoring of visual development, which includes altering contact lens parameters and patching regimen to control amblyopia, nystagmus, and strabismus.

Conclusion

- Selective endothelial removal shows to be a safe alternative to PKP for PA
- Gas permeable or scleral lenses have shown in this case to improve vision and greatly increase independence and function
- Treatment options need to be tailored to the severity of the disease. Gas permeable or scleral lenses provide superior optical correction, and should be considered in management of PA.

References

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