Surgical Approach to a Case of Peters-plus Anomaly

Hana Mansour, MD; Alaa Bou Ghannam, MD; David Walton, MD; Helen Yeung, MD; Lulwa El Zein, MD; Julien Torbey, MD

Ophthalmology Department, American University of Beirut Medical Center, Beirut, Lebanon

Introduction

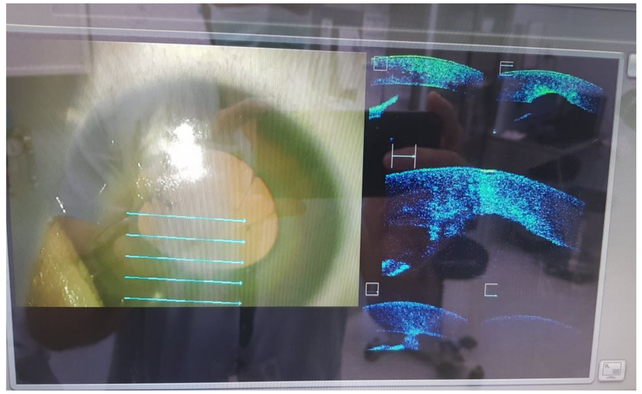
4-month-old girl had multiple congenital abnormalities including agenesis of the corpus callosum, cardiac defects, cherry red hemangiomas and gastrointestinal abnormalities presented for leukocoria.

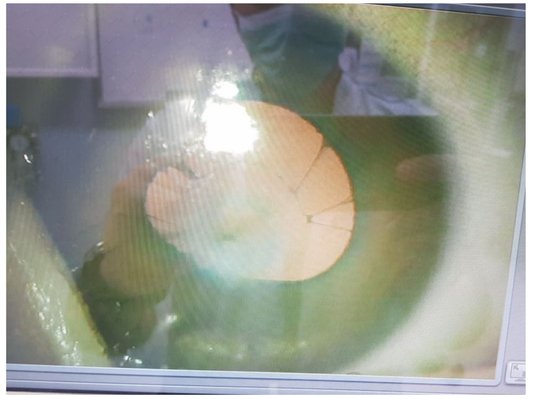
Eye Exam

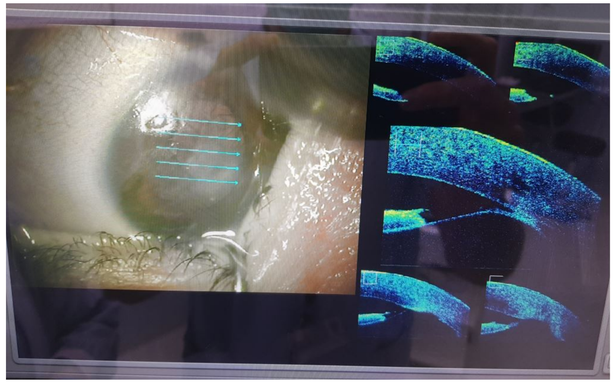
- Blinks to light but doesn't fix or follow.
- Bilateral corneal central opacities with scarring
- A/C: shallow with irido-corneal projections.
- Lenses are peripherally clear with central lenticulo-corneal touch.
- Pressures are in the upper twenties in clinic.
- OCT cornea: central loss of the endothelium + Descemet membrane with overlying stromal edema and scarring.
- AL: 17.7 mm in both eyes.
- Cup/Disc ratio: 0.2 OD and 0.15 OS, healthy.
- Genetic testing confirmed the diagnosis of Peters-plus syndrome: homozygous mutation in *B3GLCT* gene.

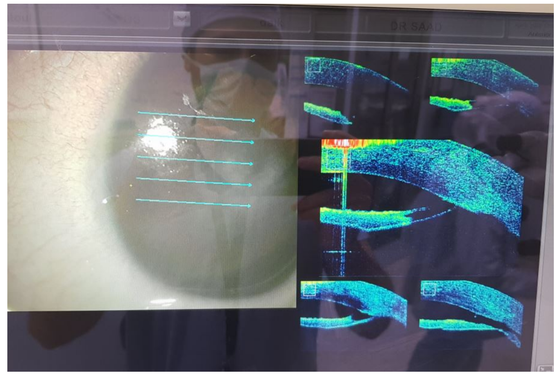
Analysis

Given the poor prognosis of corneal transplants at such a young age, it was decided to create optical iridectomies temporally in both eyes to allow some vision and avoid secondary glaucoma.









Surgical technique

Using a 23-gauge Microincision Vitrectomy (MVR) blade, the anterior chamber was entered temporally and viscoelastic was used to try to break the iridocorneal projections and to separate the lens from the cornea. Afterwards the 23-gauge ocutome was used to create a large temporal sector iridectomy of about 2.5 clock hours and to cut the thick iridocorneal projections.

Outcome

Postoperatively, the eyes had deep anterior chambers, large temporal sector iridectomy and intact anterior lens capsules. Pressures were normal. Patient was following objects in both eyes.

Conclusion

Sectoral iridectomies and severing iridocorneal adhesions are very useful and safe surgical alternatives to a low success penetrating keratoplasty to allow useful vision in eyes with Peter's anomaly.

References

Maillette de Buy Wenniger-Prick LJ, Hennekam RC. The Peters' plus syndrome: a review. Ann Genet. 2002;45(2):97-103.

Khatri D, Gosal JS, Das KK, Bhaisora KS. Peter Plus Syndrome: A Neurosurgeon's Perspective. J Pediatr Neurosci. 2019;14(3):148-153.