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Case report

Endothelial keratoplasty for posterior polymorphous corneal dystrophy in a 4-month-old infant

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ABSTRACT

Purpose: To report a case of endothelial keratoplasties (EKs) performed in a 4 month old with a posterior polymorphous corneal dystrophy.
Observations: A 4 month old infant underwent Descemet membrane endothelial keratoplasty (DMEK) for posterior polymorphous corneal dystrophy. The graft was found to be dislocated on day 5 post-operatively and an attempt to unfold the DMEK scroll and re-bubble was not successful. The patient was then treated successfully with bilateral Descemet stripping automated endothelial keratoplasty (DSAEK). At 3 years of follow-up, her visual acuity was 20/70 in the right and 20/60 in the left eye with good endothelial cell counts.
Conclusions: and Importance: This study reports the youngest case of EKs performed at the age of 4 months in an infant. This is also the first reported case of attempted DMEK highlighting its challenges in infants. DSAEK remains a good treatment option for endothelial dysfunction secondary to posterior polymorphous corneal dystrophy in infants even after failed DMEK.

1. Introduction

Posterior polymorphous corneal dystrophy (PPCD) is one of the corneal endothelial dystrophies transmitted in an autosomal dominantly manner.[1] The clinical presentation of PPCD varies from asymptomatic corneal endothelial changes to congenital corneal edema, peripheral iridocorneal adhesions and glaucoma.[2] Corneal transplant is required in 20–25% of patients with PPCD who developed corneal edema.[1] Corneal edema caused by PPCD can have a huge impact on the visual development of children due to amblyopia. Therefore surgical interventions such as corneal transplant may be considered to restore corneal clarity and prevent amblyopia.

The classic surgical treatment of PPCD has been penetrating keratoplasty (PKP).[3] However, if the corneal clarity allows, full thickness transplantion has been superseded by partial thickness transplants. The procedure most widely applied now for endothelial dystrophies is Descemet stripping automated endothelial keratoplasty (DSAEK). DSAEK involves transplanting a thin layer of stroma, Descemet's membrane and endothelium and has been successfully performed in children.[4–8] In recent years, the technique of Descemet membrane endothelial keratoplasty (DMEK) has been developed, which involves transplanting only Descemet's membrane and endothelium. This technique has shown potential for faster and improved visual outcomes compared with DSAEK.[9]

We report the case of an infant with PPCD treated initially with DMEK which was unsuccessful followed by successful bilateral DSAEK. To the best of our knowledge this is the youngest case report of endothelial keratoplasty (EK) in the literature.

2. Case report

A 3 month old girl was referred for bilateral cloudy corneas since birth. The pregnancy was normal without any signs of infection and birth was spontaneous vaginal delivery at 41 weeks gestational age. Her family history was positive for PPCD in paternal grandfather necessitating a DSAEK procedure at age 62 years after persistent corneal edema post-cataract surgery. An examination of the

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patient's asymptomatic father revealed the characteristic endothelial changes of PPCD.

The patient's examination at presentation and under anesthesia revealed diffuse corneal epithelial edema, deep speckled opacities at the level of the endothelium and Descemet's membrane folds in both eyes (Fig. 1). Intraocular pressure (IOP), central cornea thickness (CCT) measured using hand-held spectral domain optical coherence tomography (Bioptigen Inc., Morrisville, NC, USA), the horizontal corneal diameters and axial length measured with immersion A-scan at presentation are shown in Table 1. The central cornea thickness was high at presentation, 958 μm in the right eye and 884 μm in the left eye. The dilated fundus examination permitted a hazy view of both fundi with no abnormality or disc cupping.

A detailed conversation with parents regarding the amblyogenic risk from her corneal edema was undertaken and management options discussed. Parents chose to proceed with DMEK in the left eye at 4 months of age. After instilling 4% Pilocarpine, the left eye was prepped and draped in a sterile manner. Four paracenteses were created at the 11, 1, 4 and 7 o'clock positions. Acetylcholine chloride (Miochol, Bausch and Lomb, USA) was injected into the anterior chamber and sodium hyaluronate (Healon OVD - Abbott Medical Optics Inc., USA) was used to fill the eye. An inferior periphery iridectomy was performed using Vannas scissors to avoid pupillary block from air bubble in the anterior chamber in the post operative care. The stripping of the endothelium was done with a reverse Sinskey hook in a circle of diameter 8 mm. The viscoelastic was thoroughly washed out of the eye. The donor cornea was placed on a punch block with endothelial side up and Descemet membrane was stripped using the Melles technique.[10] The membrane was then cut with 8mm trephine, the Descemet membrane stripped from the cornea and stained with trypan blue and injected into the anterior chamber as a double scroll. All wounds were sutured tightly and the scroll was unrolled in the anterior chamber, using the Yourek tapping technique[11] followed by air bubble. A full air fill was left in place for at least 30 minutes with the patient in the supine position whilst under general anesthesia to facilitate

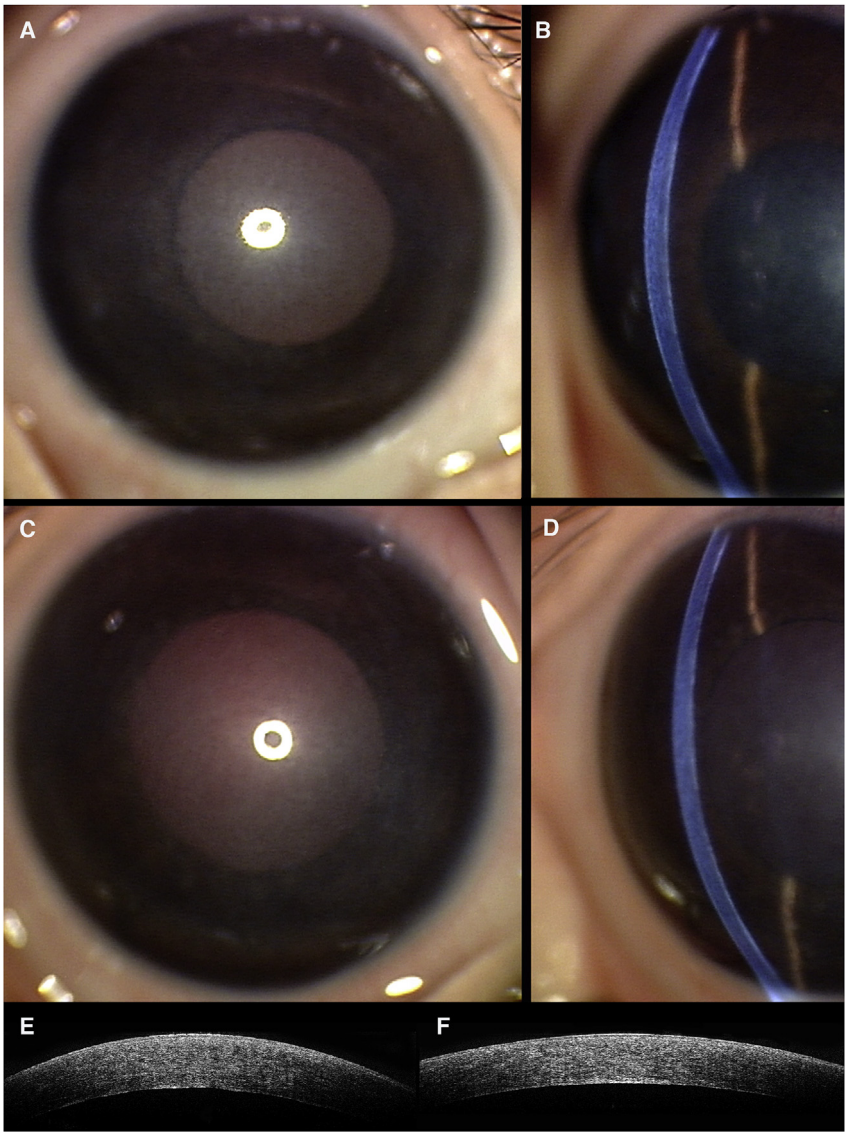

Fig. 1. A, B, C and D. Slit-lamp examination of the right eye (A) and left eye (B) at presentation showing diffuse corneal epithelial edema, and deep speckled opacity at the level of corneal endothelium. E and F. Spectral domain optical coherence tomography shows thick corneas and deep speckled opacity at the level of corneal endothelium in the right eye (E) and left eye (F).