



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:** 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.© 2017 The Authors. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Posterior polymorphous corneal dystrophy (PPCD) is one of the corneal endothelial dystrophies transmitted in an autosomal dominant 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been penetrating keratoplasty (PKP).³ However, if the corneal clarity allows, full thickness transplantation has been superseded by partial thickness transplants.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 (Biotigen 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 Sinsky 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 technique¹¹ 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

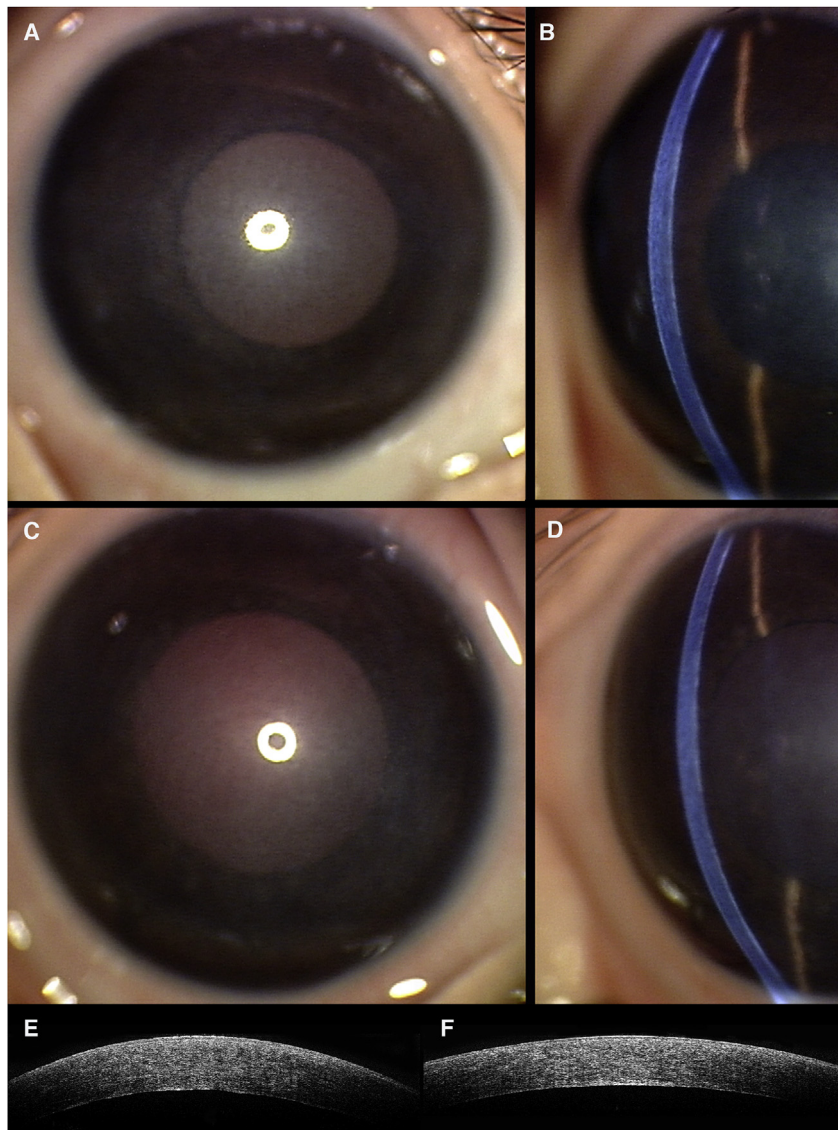


Fig. 1. A, B, C and D. Slit-lamp examination of the right eye (A) and left eye (B) at presentation showing diffuse corneal epithelial edema, and deep speckled opacity at the level of corneal endothelium. E and F. Spectral domain optical coherence tomography shows thick corneas and deep speckled opacity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

Table 1

Anterior segment characteristics at presentation and results at 3 years follow up after Descemet stripping automated endothelial keratoplasty.

	At presentation		after DSAEK	
	Right eye	Left eye	Right eye	Left eye
Cornea diameter (mm)	11.5	11.5	12.25	12.25
CCT (μm)	958	884	735 (donor tissue = 141)	706 (donor tissue = 129)
Anterior chamber depth (mm)	3.65	3.7	3.57	3.61
Axial length (mm)	18.7	18.85	20.40	20.60
IOP (mmHg)	19	15	15	15
Refraction	$-1.50/+1.50 \times 90$	$-0.75/+0.75 \times 90$	$-2.75/+2.25 \times 90$	-2.25

Abbreviations: CCT-central corneal thickness; DSAEK- Descemet stripping automated endothelial keratoplasty; IOP-intraocular pressure.

graft adhesion. At the end of the surgery only 60% of the air was left in the anterior chamber.

Postoperative treatment included dexamethasone/tobramycin ophthalmic (Tobradex, Alcon, USA) drops four times per day and atropine 1% (Bausch & Lomb, USA) at bedtime. Parents were encouraged to keep the baby in a supine position. The next day, the donor graft was attached, however at 5 days follow up, most of the graft was partially detached and scrolled in the inferior angle. Attempt was made to reattach the graft the next day without success. One week later, the patient underwent DSAEK in the left eye and 3 weeks later in the right eye. The DSAEK was performed in similar manner as described in the literature with an 8mm donor graft.¹² On follow-up examination under anesthesia 6 weeks after DSAEK, the corneas were clear and the grafts attached. At 12-month follow-up, specular microscopy during an exam under anesthesia showed the endothelial cell counts (ECC) were 1876 in the right eye and 1642 in the left eye and both corneas were clear (Fig. 2). This represents an ECC loss of 33% and 46% in the first year in the right and left eye respectively, compared with the initial ECC from surgical donor material. The remainder of the eye exam was normal.

At 3 year follow-up, her grafts remain clear and she has corrected visual acuity using the Lea chart of 20/70 and 20/60 (wearing refraction of $-2.75/+2.25 \times 90$ and -2.25) in the right and left eye respectively. The astigmatism in the right eye may be explained by a combination of a slight asymmetry in the thickness of the DSAEK and a longer corneal tunnel incision compared with the left eye. Usually one to two diopters of hyperopic shift is expected post DSAEK, whereas our patient had significant myopic shift. The myopic shift is caused by a combination of deprivational axial length growth from initial blur, and natural eye growth in the first two years of life. The increased of corneal diameter post-surgery (see Table 1) is also related to the normal growth seen in the first 2 years of life as well as the patient's mother also having corneas at 12.75mm. Her intraocular pressures were 17 in the right eye and 20 in the left eye with normal optic nerves. Since there was no unusual growth in the axial length, IOPs were never raised and discs were normal, we considered the increased of corneal diameter and myopic shift not being glaucoma related. She has been kept on prednisolone 1% drops to each eye on alternate days in the long term. The amblyopia was treated by glasses and patching up to five hours per day at six months of age when vision preference was detected.

3. Discussion

This case report describes DMEK in a 4-month-old patient with PPCD that was unsuccessful due to detachment of the graft, followed by uncomplicated DSAEK in both eyes with good outcome. To our knowledge, this is the only DMEK case reported in an infant, the youngest DSAEK treated patient with PPCD in the literature with long term follow up on visual acuity and ECC and only the second report of DSAEK in PPCD in an infant.⁴

DMEK is emerging as the keratoplasty of choice for the diseases which affect the endothelium. Maier et al. conducted a review looking at the advantages and disadvantages of DMEK versus DSAEK. DMEK had a better visual acuity, achieved more quickly and with a better patient satisfaction than DSAEK.¹³ Moreover, the rejection rate is lower in DMEK than DSAEK.¹³ However, DMEK is a more demanding surgical technique with higher rate of graft dislocation and re-bubbling as well as with more endothelial cell loss compared with DSAEK in early reports,¹³ though more recent papers are showing better endothelial cell counts with DMEK.¹⁴

Endothelial keratoplasty is more challenging in children than adults because of smaller cornea size, shallower anterior chamber, phakic status, soft sclera and difficulties maintaining post-operative supine positioning. Furthermore, any attempt at re-bubbling or even examination is difficult and may necessitate the use of general anesthesia. Gonnermann J et al.¹⁵ reported the first case of successful DMEK in a 12 year old with Kearns-Sayre syndrome and corneal edema. In this report the transplant remained clear 6 months after the surgery with a best-corrected vision of 20/100. Our case is the second report of DMEK in children and the first reported in an infant. Although our patient had a good cornea size, deep anterior chamber and a technically uncomplicated DMEK procedure, graft dislocation and unsuccessful re-bubbling resulted in surgical failure. Graft displacement in our patient is less likely to be due to inadequate wash of viscoelastic circulating in the anterior chamber because extensive wash of cohesive viscoelastic was performed with confidence that it was all removed. A clear cornea and good outcome with subsequent DSAEK was achieved in our patient even though the overall postoperative corneal thickness (see Table 1) was still relatively high. The use of ultrathin DSAEK could be potentially better and should be considered. There are only five reports^{4–7} including one series⁸ published in the literature of DSAEK in infants for CHED and PPCD with a follow-up from 3 to 24 months showing good graft survival without cataract formation. Lenhart et al.⁷ reported vision of 20/40 at two years follow-up after DSAEK performed in a 8 month old patient with CHED.

Posterior polymorphous corneal dystrophy is one of the corneal endothelial dystrophies with wide range of clinical presentation. PPCD is usually an asymptomatic disease with normal vision, unless there is asymmetric disease, corneal edema, visual axis opacity or refractive error causing amblyopia at early age.^{16,17} Surgical management such as corneal transplant is required when the risk of amblyopia is high.

4. Conclusions

Our single case does not allow any conclusions to be drawn regarding the DMEK technique as a viable option for infants. The potential for better visual acuity and lower rejection rate in children would be a major advantage of DMEK over DSAEK. It may be that future refinements and increasing expertise in DMEK leads to better outcome for children. It would be advantageous to have

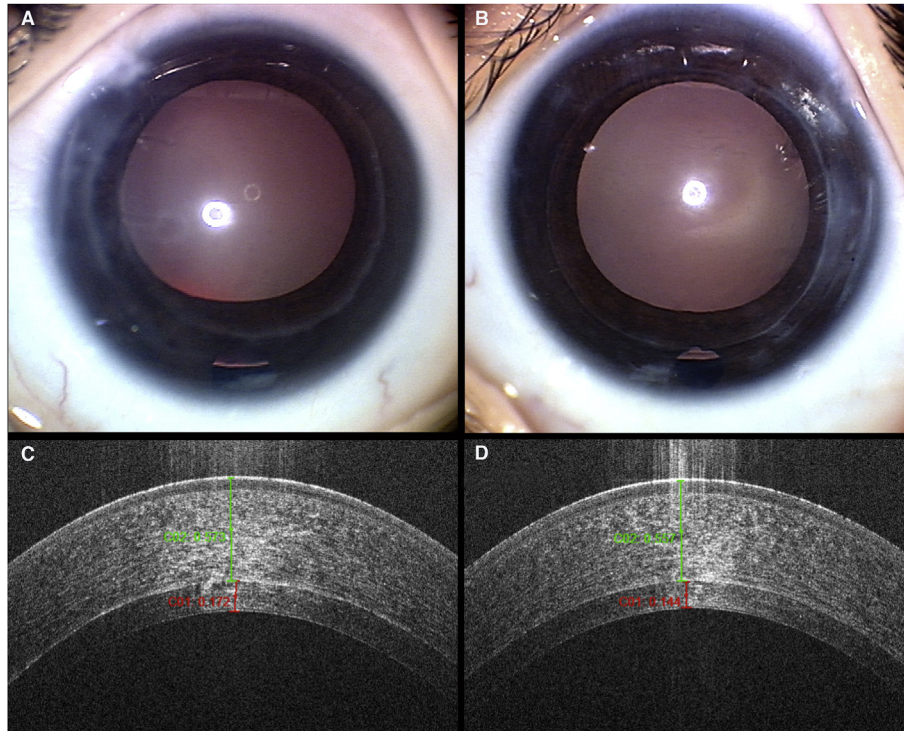


Fig. 2. A and B Slit-lamp examination of patient's right eye (A) and left eye (B) one year post Descemet's stripping automated endothelial keratoplasty (DSAEK) surgery showing clear cornea. C and D. Hand-held spectral domain optical coherence tomography of the right (C) and left eyes (D) shows attached grafts one year after DSAEK surgery.

ready access to hand held anterior SD-OCT and exam under anesthesia to better assess DMEK adhesion and perform re-bubbling procedure in infants. It is encouraging however, that good visual outcomes are achievable with subsequent DSAEK, even if DMEK is unsuccessful in infants.

Patient consent

Patient consent was taken in writing before preparation of this article.

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Conflict of interest

The following authors have no financial disclosures: MHS, DR, AA, KM.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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