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Visual outcome after Descemet's stripping automated endothelial keratoplasty in an 8-month-old with congenital hereditary endothelial dystrophy

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Abstract

Descemet's stripping automated endothelial keratoplasty (DSAEK) has rapidly become the standard of care for endothelial dysfunction of the cornea in adults. There are few reports of DSAEK in children and infants, mainly because most pediatric corneal opacities are full-thickness and therefore not amenable to lamellar procedures but also because of the unique difficulties of performing this procedure in the youngest patients. We report the case of an 8-month-old girl who underwent DSAEK for congenital hereditary endothelial dystrophy. At 24 months' follow-up, her visual acuity was 20/40 in the operated eye. To our knowledge, this is the first report of an objective visual outcome in a child with DSAEK performed in infancy.

Case Report

A healthy 7-week-old girl presented to the Glaucoma Service at Emory Eye Center for a poor red reflex in both eyes. There was no epiphora or photophobia. Family history was noncontributory. Intraocular pressure (IOP) by applanation tonometry was 30–31 mm Hg in the right eye and 30–32 mm Hg in the left eye on 2% dorzolamide twice daily in both eyes, with pachymetry of 600–1100 μ m bilaterally. Corneal diameters were 11.0–11.5 mm in both eyes. Diffuse, bilateral corneal haze was noted, with no Haab's striae. The anterior segments and dilated fundus examination appeared otherwise normal. Because of dense corneal edema, high IOPs despite medical therapy, and concern for progressive ocular enlargement, the child underwent sequential, bilateral 360-degree trabeculotomies for presumed congenital glaucoma. Postoperatively, the IOP by applanation was 15–16 mm Hg in both eyes, with some subjective improvement noted in the corneal haze.

On reexamination at 3 months of age, her corneas were notable for limbus-to-limbus moderate diffuse haze (Figure 1). No inflammation or dystrophic deposits were noted. The child's parents elected to proceed with DSAEK in one eye when the girl was 9 months of age. Her vision before surgery was central, steady, and maintained in both eyes and her IOP

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was controlled on 0.25% timolol and latanoprost eyedrops. The corneal edema was still dense in both eyes.

Perioperatively, pachymetry in the right eye was 1115 μm and was too thick to measure in the left eye (Pachmate DGH 55 Exton, PA). Corneal dimensions were 10.5 mm vertically and 11.25 mm horizontally in the left eye. A paracentesis and a 3.5 mm superior limbal incision were made, and sodium hyaluronate was injected to form the anterior chamber. The posterior cornea was scored with a cystotome. Descemet's membrane was very adherent to the underlying corneal stroma, and forceps and the cystotome were used to peel the central tissue away in fragments. The anterior caplet of the pre-cut donor corneal tissue was removed. The posterior portion was placed stromal side down and a 7.75 mm trephination was performed. A Sheets glide was placed through the superior incision to prevent iris prolapse and the corneal tissue was placed on sodium hyaluronate over the Sheets glide and inserted through the incision using a 30-gauge needle. The tissue immediately unfolded. Interrupted 9.0 polyglactin 910 sutures were used to close both wounds. Air was injected into the eye behind the donor button, but keeping the air in the eye proved difficult. Ophthalmic viscoelastic plugs were placed at the paracentesis and the superior incision followed by multiple rebubbings. IOP by Tono-Pen XL (Reichert, Inc., Depew, NY) was 12 mm Hg at the end of the case.

On postoperative day 1, slit-lamp examination showed a round pupil, moderate corneal edema, and a hazy view of the anterior chamber and graft. There were superior iris transillumination defects. The child was placed on 1% prednisolone acetate eyedrops every 2 hours while awake, moxifloxacin 4 times a day, and neomycin/polymyxin B/dexamethasone ointment at bedtime. Two weeks after surgery, examination under general anesthesia revealed an IOP of 27 mm Hg on 0.25% timolol twice daily and pachymetry of 1069 μm in the left eye. The cornea was still edematous, but some iris details were visible. External cannula sweep across the corneal surface showed that the corneal button was adherent and slightly superiorly displaced. On examination under anesthesia at postoperative week 6, the left pupil was slightly eccentric and the donor corneal button was attached. There were 1-2 clock hours of iridocorneal touch to the donor-host junction and to a corneal vent incision created during surgery to promote graft adherence. A good red reflex was noted and cycloplegic refraction was plano +0.75 $\times$ 080 in the left eye. Prednisolone acetate was decreased. Three months later, the patient had mild diffuse haze in the operated eye, greater on the temporal side than on the nasal side, and a 360-degree ring of fibrosis at the graft-host junction. Ultrasound biomicroscopy confirmed graft adherence (Figure 2). Six months after surgery alternate patching of the eyes was recommended to prevent the child's (unoperated) right eye from becoming amblyopic, as a brighter red reflex was noted in the left eye than in the right eye (Figure 3). IOP was 14 mm Hg on 1% prednisolone acetate twice daily, 0.5% timolol twice daily, and 0.005% latanoprost every 4 hours. At 24 months' follow-up, she had an uncorrected visual acuity of 20/80 in the right eye and 20/40 in the left (operated) eye using isolated HOTV matching. IOP in the left eye was 13 mm Hg on the same dosage of timolol and latanoprost. The left corneal graft had residual mild, diffuse interface haze, and was significantly clearer than the unoperated right cornea. The left lens was clear.

Discussion

DSAEK has become a viable option for the treatment of pediatric corneal endothelial disease. It has been performed in children¹⁻⁵ but infrequently during infancy⁶⁻¹⁰ (Table 1). We report successful DSAEK in a 8-month-old child with CHED with uncorrected visual acuity of 20/40 in the left eye at 24 months' follow-up. The longest follow-up reported to date for a patient series including some infants is 15.9 months (range, 3-48 months).⁷

Whereas our patient's IOP is currently acceptable on two medications, her long-term risk for glaucoma in the operated eye is uncertain. Following lens clarity and obtaining an endothelial cell count will be important considering the multiple intraoperative rebubbings required.

The allure of DSAEK in children is clear in light of the many drawbacks of pediatric penetrating keratoplasty, including astigmatism, suture complications, prolonged steroid treatment, and the risk of traumatic graft dehiscence. Long-term outcomes of DSAEK in infants have not been established, but results in older children have been promising. There are also risks associated with DSAEK that may disproportionately affect younger children, who are generally phakic, with smaller anterior chambers and softer sclera. Technical challenges previously cited have included difficulties with Descemet's stripping² or inability to identify Descemet's membrane in infants < 12 months of age,¹⁰ inadequate visualization due to corneal opacity,² lenticular touch,⁸ graft dislocation,⁴ and the requirement for prolonged general anesthesia due to the difficulty of maintaining an air bubble tamponade.⁹ While early surgical intervention is advocated in children to avoid amblyopia, the optimal age for endothelial keratoplasty remains controversial. Our case demonstrates that good visual outcomes can be achieved in infants despite the difficulties in performing DSAEK in the first year of life. Further advances in surgical technique may improve the safety of DSAEK as a viable option for the treatment of pediatric corneal endothelial disease.

Literature Search

PubMed was searched last on July 15, 2013, using the search terms: *Descemet's stripping endothelial keratoplasty in children* and *Descemet's stripping endothelial keratoplasty AND pediatric*. All relevant articles published in the English language were reviewed for cases of Descemet's stripping endothelial keratoplasty performed in infancy.

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Fig 1.
Preoperative examination revealed bilateral, diffuse corneal haze in this 8-month-old girl with congenital hereditary endothelial dystrophy.

Fig 2.
Ultrasound biomicroscopy confirmed graft adherence 4.5 months after DSAEK in the left eye.

Fig 3.
Six months after DSAEK, a brighter red reflex was noted in the patient's left eye.

Reports of DSAEK in infants

Table 1

Author	No. eyes	No. patients ^b	Indication	Age, months	Postoperative VA	Follow-up, months
Jeng et al (2008) ⁶	1	1	Corneal edema	25	CSUM	6
Busin et al (2011) ^{7a}	6	6	CHED	6-9	FF	3-9
Belluci et al (2011) ^{8a}	2	1	CHED	3	Follows light OU	10, 9 (OD,OS)
Madi et al (2012) ^{10a}	6	6	CHED	6-9	FF	3-9
Sella et al (2013) ⁹	2	1	PPMD	8	Not recorded	6

CHED, congenital hereditary endothelial dystrophy; CSUM, central, steady, unmaintained; DSEK, Descemet's stripping endothelial keratoplasty; FF, fix and follow; OD, right eye; OS, left eye; OU, both eyes; PPMD, posterior polymorphous dystrophy; VA, visual acuity.

^aDescemet's membrane not identified/not removed.

^bInfants (< 25 months of age).