Endothelial Keratoplasty for Congenital Corneal Edema

Massimo Busin MD, Silvana Madi MD, Paolo Santorum MD

Congenital corneal edema is due to either primary (ie, endothelial dystrophies[1,2]) or secondary (ie, congenital glaucoma[3]) endothelial failure. Loss of corneal transparency due to any of these etiologies causes visual deprivation and consequent amblyopia of various degrees.

Penetrating keratoplasty (PK) has been the gold standard for treatment of congenital corneal edema in the pediatric age group. However, PK is very challenging in children because of both low scleral rigidity and high vitreous pressure, which increase the incidence of vision-threatening intraoperative complications.

Descemet-stripping automated endothelial keratoplasty (DSAEK) offers several advantages over PK.[4] It is performed under "closed system" conditions, thus minimizing the incidence of intraoperative complications. DSAEK's small corneal incision requires fewer sutures and is less likely to dehisce than PK. In addition, the sutures of DSAEK are completely removed in children as early as 2 weeks postoperatively, thus allowing prompt treatment of amblyopia and more rapid visual recovery.

To date, few reports have been published about DSAEK in congenital corneal edema. Two reports concerned DSAEK in congenital hereditary endothelial dystrophy (CHED).[5,6] More recently, several children, together with adults, were included in a series of DSAEK procedures performed for CHED.[7] We report herein the outcomes of DSAEK performed at our institution in 17 eyes with congenital corneal edema.

Subjects and Methods

We reviewed the medical records of all pediatric patients who underwent DSAEK at our institution from January 2007 to March 2012.

All patients or legally responsible caretakers provided informed consent for the procedures performed. Analysis of the data extracted from the medical records was performed using a standard spreadsheet program. Preoperatively, all patients underwent a complete ophthalmologic examination, including slitlamp examination, visual acuity and manifest refraction, applanation tonometry, and ocular motility, as well as funduscopy, when possible and appropriate. Visual acuity was measured by Snellen chart or assessment of fixation patterns in infants. Follow-up examinations were not possible at regular intervals at our institution, as most patients were referred. However, each patient was seen at our facility at least once after suture removal, and additional information was retrieved from the referring ophthalmologists.

Surgery was performed using general anesthesia according to the standard technique described in a previous report[7] and shown in Figure 1.

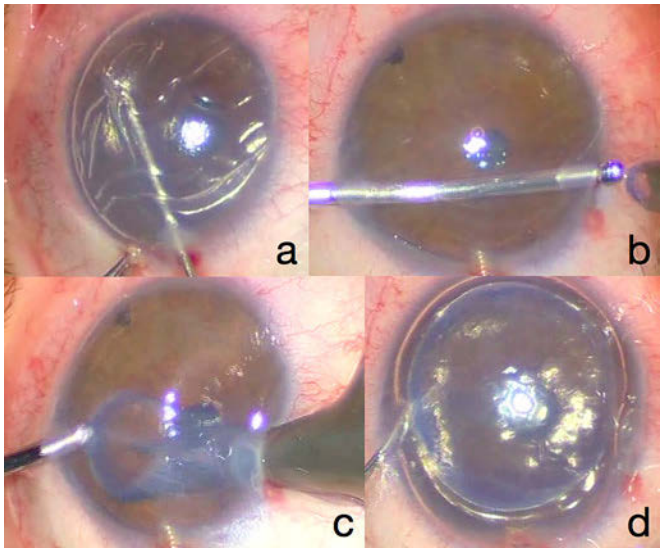

Figure 1. DSAEK standard technique. The procedure includes scoring and stripping of the Descemet membrane using a 25-gauge bent needle (a), bimanual DSAEK graft delivery under continuous irrigation through incisions shifted superiorly by 1 mm to avoid contact with the crystalline lens (b and c), complete air fill at the end of the procedure to tamponade the graft and secure attachment to the posterior corneal surface after airtight suturing of all incisions, including the side entries (d).

The Descemet membrane could not be identified in infants (age < 12 months) and therefore was not stripped in these eyes. In all phakic eyes (n = 15) the incisions sites were shifted 1 mm superiorly from the standard 3 and 9 o'clock position. This was done to protect the crystalline lens from accidental trauma with the instrument, while performing the pull-through maneuver for the insertion of the graft.[7] In the 2 aphakic eyes venting incisions were used to drain fluid from the interface while the air tamponade was taking place.

Postoperatively, patients were instructed to lie supine for 2 hours, when possible. All patients were examined not later than 2 hours after surgery at the slitlamp or again using the operating microscope, and some air was removed when the air level failed to lie above the inferior peripheral iridotomy by this time.

All patients were seen at Days 1 and 2, as well as Week 1 after surgery. Later follow-up examinations were scheduled at Months 1, 3, 6, and 12 and were performed elsewhere for all patients referred elsewhere.

Results

Seventeen eyes of 9 patients 16 years old or younger (6 male, 3 female) who underwent DSAEK at our institution were identified. Patients' age ranged from 6 months to 16 years. The average follow-up in this series was 14.5 months (range: 3 to 48 months). Causes of congenital corneal edema included CHED (n = 15) and congenital glaucoma (n = 2). Fifteen eyes had clear crystalline lens at the time of presentation, 2 eyes were aphakic. Table 1 summarizes the demographics of population.

Table 1. Demographic Data

Patient	Eye	Age/Sex	Diagnosis	Lens Status
1	O.D.	6 m/F	CHED	Phakic
1	O.S.	7 m/F	CHED	Phakic
2	O.D.	6 m/M	CHED	Phakic
2	O.S.	7 m/ M	CHED	Phakic
3	O.D.	8 m/M	CHED	Phakic
3	O.S.	9 m/M	CHED	Phakic
4	O.D.	10 m/M	CHED	Phakic
4	O.S.	10 m/M	CHED	Phakic
5	O.D.	7 y/M	CHED	Phakic
6	O.D.	7 y/F	CHED	Phakic
6	O.S.	7 y/F	CHED	Phakic
7	O.D.	9 y/M	CHED	Phakic
7	O.S.	10 y/M	CHED	Phakic
8	O.D.	16 y/F	CHED	Phakic
8	O.S.	16 y/F	CHED	Phakic
9	O.D.	15 y/M	Buphthalmus	Aphakic
9	O.S.	15 y/M	Buphthalmus	Aphakic

Abbreviations: m = months; y = years; M = male; F = female; CHED = congenital hereditary endothelial dystrophy.

All surgeries were uneventful. Graft dislocation occurred in 4 eyes (all infantile) within the first 2 postoperative days and was managed successfully in all eyes by rebubbling under general anesthesia.

All corneas cleared by 1 week postoperatively and remained so for the whole period of follow-up.

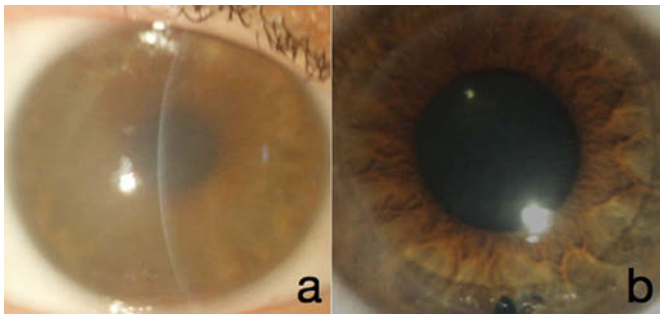

Figure 2. Slitlamp appearance of an eye with CHED before (a) and few weeks after (b) DSAEK.

The only late complication observed was an immunologic rejection episode, easily reverted with topical and systemic steroids. No lenticular opacities were seen postoperatively in any eye.

The outcomes of DSAEK in our pediatric population are summarized in table 2.

All 8 eyes of the 4 infants included in this series could fix and follow as early as 1 week after surgery, whereas 2 of the 6 eyes

Table 2. DSAEK Outcomes in Pediatric Population

Patient	Eye	Preop. BCVA	Graft size (mm)	Postop. BCVA	Refraction	Follow-up (months)	ECL at Last Follow-up
1	O.D.	No FF	8.5	FF	FF	9	NA
1	O.D.	No FF	8.5	FF	FF	9	NA
2	O.D.	FF	8.5	FF	FF	3	NA
2	O.S.	FF	8.5	FF	FF	4	NA
3	O.D.	FF	8.5	FF	FF	4	NA
3	O.S.	FF	8.5	FF	FF	3	NA
4	O.D.	FF	9.0	FF	FF	5	NA
4	O.S.	FF	9.0	FF	FF	3	NA
5	O.D.	20/200	9.0	20/25	+7.0/+1.5×80	48	19%
6	O.D.	20/200	9.0	20/40	+2.0/-0.5×90	18	43.0%
6	O.S.	CF	9.0	20/70	+2.0/-3.0×80	24	25.9%
7	O.D.	20/200	9.0	20/25	+7.0/+1.5×80	18	29.7%
7	O.S.	CF	9.0	20/27.5	+6.0/+0.5×90	9	37.5%
8	O.D.	20/100	9.0	20/25	+1.5/-2.0×60	24	30.5%
8	O.S.	20/70	9.0	20/22.5	+0.5/-1.0×90	30	34.8%
9	O.D.	<20/200	9.5	20/200	+10 sphere	18	53%
9	O.S.	<20/200	9.5	20/200	+10 sphere	12	49.7%

Abbreviations: BCVA = best-corrected visual acuity; CF = count fingers; HM = hand motion; ECL = endothelial cell loss; FF = fix and follow; NA = not available.

could not preoperatively. In older children, whose visual acuity could be assessed by means of Snellen charts, best-corrected visual acuity (BCVA) improved to 20/40 or better in 6 of 9 eyes (66.6%). Reasons for vision worse than 20/40 were glaucomatous damage ($n = 2$) and amblyopia ($n = 1$).

Mean postoperative refractive astigmatism was 1.1 ± 0.99 D, ranging from 0.5 to 3 D.

Endothelial cell density could be evaluated in 9 eyes. At the time of this review, the average endothelial cell loss from the cell density measured at the eye bank was 36% (range: 19%-53%).

References

1. Waring GO III, Rodrigues MM, Laibson PR. Corneal dystrophies: II. Endothelial dystrophies. *Surv Ophthalmol*. 1978; 23(3):147-168.
2. Pearce WG, Tripathi RC, Morgan G. Congenital endothelial corneal dystrophy: clinical, pathological, and genetic study. *Br J Ophthalmol*. 1969; 53:577-591.
3. Vanathi M, Panda A, Vengayil S, Chaudhuri Z, Dada T. Pediatric keratoplasty. *Surv Ophthalmol*. 2009; 54(2):245-271.
4. Lee WB, Jacobs DS, Musch DC, et al. Descemet's stripping endothelial keratoplasty: safety and outcomes—a report by the American Academy of Ophthalmology. *Ophthalmology* 2009; 118(10):1944-1949.
5. Pineda R II, Jain V, Shome D, Hunter DC, Natarajan S. Descemet's stripping endothelial keratoplasty: is it an option for congenital hereditary endothelial dystrophy? *Int Ophthalmol*. 2010; 30(3):307-310.
6. Anwar HM, Eldanasoury AM, Hashem AN. Descemet's stripping automated endothelial keratoplasty for congenital hereditary endothelial dystrophy. *Clin Ophthalmol*. 2012; 6:159-163.
7. Busin M, Beltz J, Scorcia V. Descemet-stripping automated endothelial keratoplasty for congenital hereditary endothelial dystrophy. *Arch Ophthalmol*. 2011; 129(9):1140-1146.
8. Busin M, Bhatt PR, Scorcia V. A modified technique for Descemet membrane stripping automated endothelial keratoplasty to minimize endothelial cell loss. *Arch Ophthalmol*. 2008; 126(8):1133-1137.