ONLINE FIRST

Descemet-Stripping Automated Endothelial Keratoplasty for Congenital Hereditary Endothelial Dystrophy

Massimo Busin, MD; Jacqueline Beltz, FRANZCO; Vincenzo Scorcia, MD

Objective: To describe the results of Descemet-stripping automated endothelial keratoplasty (DSAEK) for congenital hereditary endothelial dystrophy (CHED).

Methods: The medical records of all patients with CHED who underwent DSAEK at our institution were reviewed. A standard DSAEK was performed in all cases with the exception of the Descemet membrane not being removed in patients younger than 12 months. A thorough ophthalmic examination was performed preoperatively and at each postoperative visit in all patients.

Results: Fifteen eyes of 8 patients with phakic eyes (4 male and 4 female) were included. The mean age was 9 years (range, 6 months to 30 years). The average follow-up was 15.9 months (range, 3 to 48 months). There were 4 cases of graft detachment, all of which were managed by rebubbling. All corneas were clear within 1 week after surgery. Two of the three infants (6 eyes) could fix and follow preoperatively, while all 3 could do so as early as 1 week following surgery on the second eye. In older patients (9 eyes), preoperative best-corrected visual acuity was 20/200 or less in 6 eyes. Postoperatively, 8 eyes achieved a best-corrected visual acuity of 20/40 or better. Endothelial cell loss (=7 eyes) averaged 30.0% (range, 8.3% to 43.0%).

Conclusions: DSAEK performed in eyes with CHED allows rapid restoration of corneal clarity while minimizing intraoperative and postoperative complications. Our data suggest performing surgery at an earlier age, thus providing opportunity for improved visual development and potentially avoiding amblyopia.

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CONGENITAL HEREDITARY endothelial dystrophy (CHED) is characterized by bilateral corneal clouding of either congenital or infantile onset.[1] The course of the disease may be relatively stationary or may progress for a period of 1 to 10 years.[1] Symptoms include reduced vision, photophobia, and tearing, often resulting in amblyopia.

Although pathologic findings of CHED can be seen only in the Descemet membrane (DM) (diffuse thickening and lamination) and endothelium (sparse and atrophic cells),[1] for many years, penetrating keratoplasty (PK) has been the standard surgical treatment used in these patients.[2-8] However, to date, no definite evidence of the appropriate timing for PK has been found, as the risk of amblyopia must be weighed against the increased risk of complications described when this type of surgery is performed in children.

Today, Descemet-stripping automated endothelial keratoplasty (DSAEK) has become the treatment of choice in adults with corneal endothelial disease of various origin (Fuchs dystrophy, postsurgical endothelial decompensation, graft failure)[9] but there are few articles describing sporadic cases of DSAEK performed in the pediatric age group.[10-13] This may be because corneal diseases selectively affecting the endothelium in children, such as CHED, are rare in general and predominantly present in parts of the world with reduced access to tertiary care facilities or, most importantly, because the presence of a clear crystalline lens with normal accommodative function constitutes, for most surgeons, a contraindication to DSAEK surgery, particularly in children, as cataract formation has been reported to occur in up to 40% of phakic eyes after endothelial keratoplasty.[14]

Thanks to a simple modification of the surgical technique, during the last 3 years we have been able to routinely perform uneventful DSAEK in phakic patients, including children. Herein, we describe 15

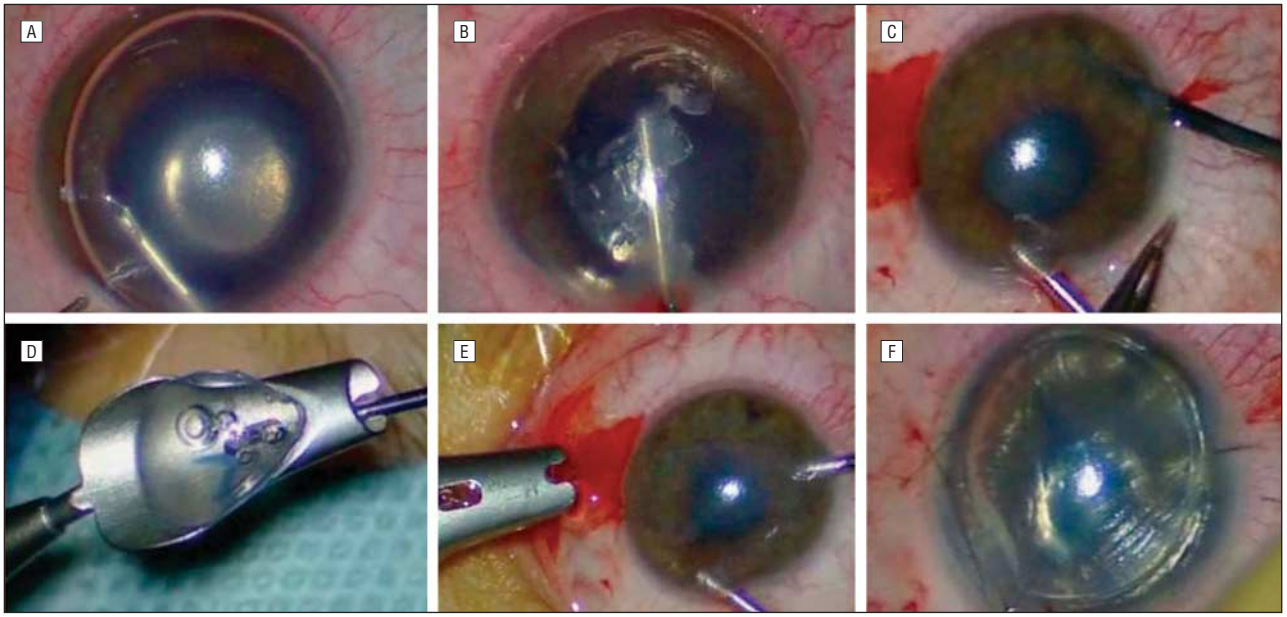

Figure 1. Descemet-stripping automated endothelial keratoplasty (DSAEK) standard technique. The procedure includes scoring of the Descemet membrane using a 25-gauge needle (A); stripping of the Descemet membrane with a blunt cannula (B); creation of an inferior peripheral iridotomy (C); loading of microkeratome-dissected donor tissue, 9.0 mm in diameter, on a Busin glide (D); bimanual DSAEK graft delivery with continuous irrigation (E); and after suturing all wounds, air injection with a 30-gauge needle inserted obliquely through the paralimbal cornea to achieve a complete fill of the anterior chamber (F).

cases of DSAEK in 8 patients with CHED. To the best of our knowledge, this is the first case series of DSAEK performed in patients, including children, who have CHED.

METHODS

We reviewed the medical records of all patients who underwent DSAEK for CHED by the same surgeon (M.B.) at our institution from January 2007 to July 2010.

All patients or legally responsible care takers provided informed consent for the procedures performed. Analysis of the data extracted from the medical records was performed using a standard spreadsheet program. A complete ophthalmological examination, including slitlamp examination, visual acuity and manifest refraction, applanation tonometry, ocular motility, and funduscopy, was performed preoperatively in all patients 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 from other, distant countries. 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 with additional peribulbar anesthesia (50% mixture of lidocaine, 2%, and bupivacaine, 0.5%) in 13 cases and under peribulbar anesthesia alone in the remaining 2 cases.

The surgeon sat at the 12-o'clock position in all cases. The main surgical steps are illustrated in **Figure 1**. We performed DSAEK using our standard, previously described technique.[15]

In 6 eyes of 3 patients, the DM could not be found; therefore, no tissue of any kind was removed from the corneas before attaching the donor graft. These 3 patients corresponded to the 3 patients younger than 1 year at the time of surgery.

In addition, owing to the phakic status of all of these patients, in the most recent cases (n=8), the incision sites were moved approximately 1 mm superiorly from our standard 9- and 3-o'clock positions. In doing so, the whole pull-through maneuver could be performed using the superior part of the iris to protect the underlying crystalline lens from accidental contact with the instruments (**Figure 2**).

Postoperatively, patients were instructed to lie supine for 2 hours, when possible for this young cohort of patients. All patients were examined 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.

Patients were given topical tobramycin, 0.3%, and dexamethasone, 0.1%, suspension (TobraDex; Alcon, Fort Worth, Texas) combination therapy every 2 hours after surgery; this was reduced as clinically indicated throughout the postoperative period. Patients were followed up at days 1 and 2 and week 1. Follow-up examinations at later times (month 1, and then at 3-month intervals) were possible only in 4 eyes (2 patients).

RESULTS

The results are summarized in **Table 1**.

Fifteen eyes of 8 patients with CHED (4 male and 4 female) who had DSAEK at our institution were identified. Preoperatively, all patients had diffuse, bilateral corneal edema rendering the details of the intraocular structures difficult to identify (**Figure 3**). Diagnosis of CHED was confirmed by exclusion of other pathologies such as congenital glaucoma and mucopolysaccharidosis. Also, histologic examination confirmed the clinical diagnosis in 3 eyes from which the removed DM and endothelium could be examined (**Figure 4**). The inheritance pattern of CHED was not determined; however, none of the patients had affected parents.

All patients had phakic eyes; at the time of presentation, the crystalline lens appeared to be clear in all cases on slitlamp examination. No patient had a history of ocular intervention. One infant exhibited strabismus, with alternating esotropia; nystagmus was not present in any patient. Fundus examinations and/or B-scan echogra-

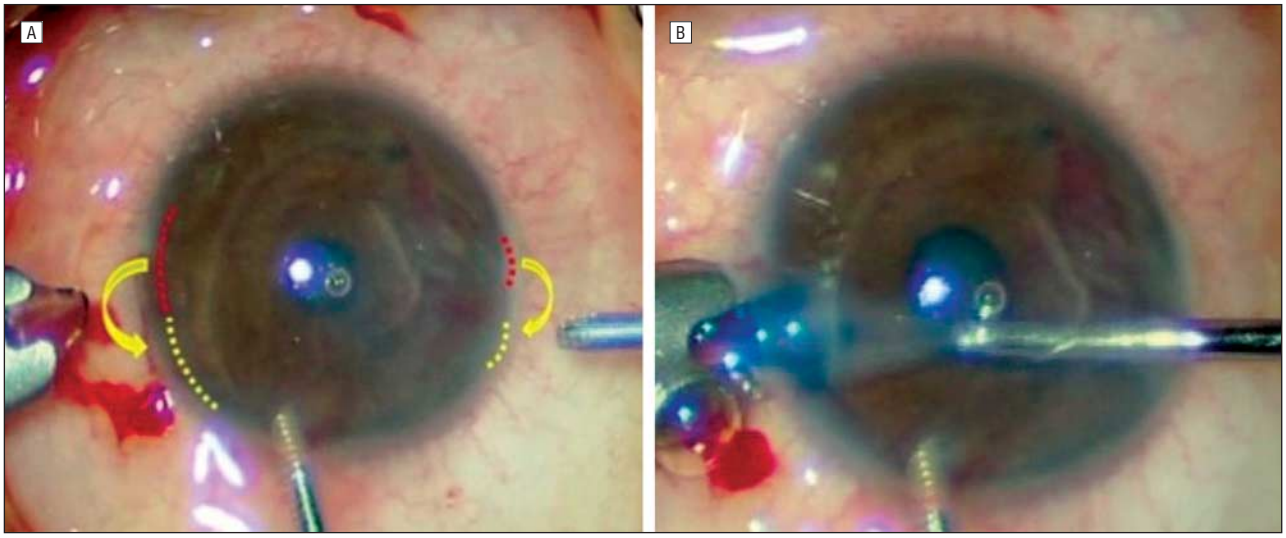

Figure 2. Modified graft delivery technique for Descemet-stripping automated endothelial keratoplasty in phakic eyes. A, Wounds for insertion of donor tissue are shifted superiorly (yellow dots) from their regular 9- and 3-o'clock positions (red dots). B, During graft delivery, forceps do not pass across the pupil; the iris protects the underlying crystalline lens from any possible contact with the instrument.

Table 1. Patient Demographic Data and Results

Case/ Sex/Age	Country of Origin	Eye	Preoperative BCVA	Postoperative BCVA and Refraction	Time of Follow-up, mo	ECL at Last F/U, %
1/M/9 y	Israel	Right	20/200	20/25 (+7/+1.5×80)	18	29.7
2/M/10 y	Israel	Left	CF	20/27.5 (+6.0/+0.5×90)	9	37.5
3/F/6 mo	Qatar	Right	No FF	FF	9	NA
4/F/7 mo	Qatar	Left	No FF	FF	9	NA
5/M/8 mo	Qatar	Right	FF	FF	4	NA
6/M/9 mo	Qatar	Left	FF	FF	3	NA
7/M/7 y	Egypt	Right	20/200	20/40 (+3.5/+1.0×40)	48	NA
8/F/16 y	Italy	Right	20/100	20/25 (+1.5/–2.0×60)	24	30.5
9/F/16 y	Italy	Left	20/70	20/22.5 (+0.5/–1.0×90)	30	34.8
10/M/6 mo	Iran	Right	FF	FF	3	NA
11/M/7 mo	Iran	Left	FF	FF	4	NA
12/F/29 y	Israel	Left	20/100	20/27.5 (+2.5/–2.5×150)	24	8.3
13/F/30 y	Israel	Right	20/200	20/25 (+1.5/–1.0×180)	12	NA
14/F/7 y	Macedonia	Right	20/200	20/40 (+2.0/–0.5×90)	18	43.0
15/F/7 y	Macedonia	Left	CF	20/70 (+1.0/–3.0×80)	24	25.9

Abbreviations: BCVA, best-corrected visual acuity; CF, count fingers; ECL, endothelial cell loss; FF, fix and follow; F/U, follow-up; NA, not available.

phy were unremarkable in all cases. Patient ages at the time of surgery ranged from 6 months to 30 years, with a mean age of 9 years. The average follow-up in this series was 15.9 months (range, 3 to 48 months).

The DSAEK was performed without complication in all cases. Four cases, all infant eyes, required reinjection of air under general anesthesia for graft dislocation that occurred within 2 days after surgery. In all cases, the donor tissue was successfully attached following this minor procedure.

There were no cases of pupillary block in this series. No late postoperative complications have occurred; in particular, there have been no cases of graft failure or rejection.

All corneas were clear by 1 week after surgery, and no cases have developed lenticular opacity during the follow-up time. Two of the 3 infants could fix and follow preoperatively, while all 3 could do so as early as 1 week following the second DSAEK surgery. In older cases (9 eyes), preoperative best-corrected visual acuity was better than 20/200 in 3 eyes and 20/200 or less in 6 eyes. At the last follow-up examination, 8 of these eyes had best-corrected visual acuity of 20/40 or better, and the remaining eye had best-corrected visual acuity of 20/70. The average postoperative spherical refractive error for these 9 cases was +2.44 diopters (range, –0.5 to +7.75 diopters). **Figure 5** illustrates an example of preoperative and postoperative slitlamp appearance.

Endothelial cell density could be determined in 7 eyes at different postoperative examination times (mean, 21 months; range, 9 to 30 months; Table 1); endothelial cell loss averaged 30.0% (range, 8.3% to 43.0%).

COMMENT

Penetrating keratoplasty in pediatric patients poses an additional challenge compared with the same procedure in adults.[16]

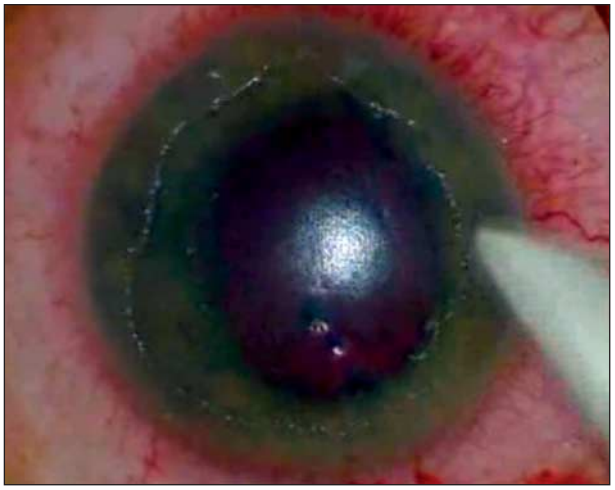

Figure 3. "Pitting edema" of the cornea of a patient with congenital hereditary endothelial dystrophy, following compression of the corneal tissue with a Weck-Cel spear.

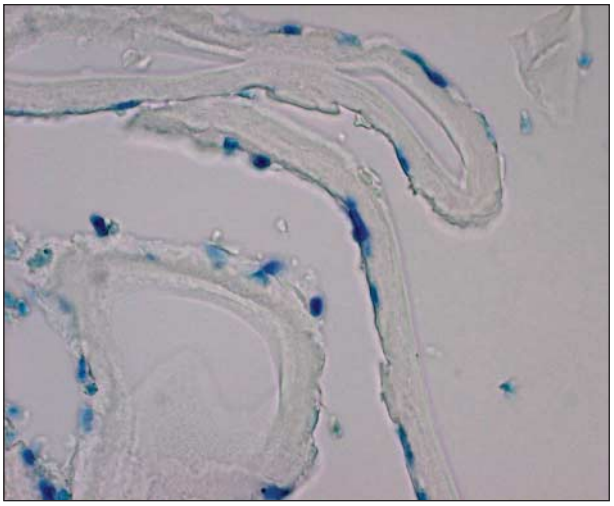

Figure 4. Light microscopic appearance of thickened Descemet membrane with sparse endothelial cells in a 29-year-old patient with congenital hereditary endothelial dystrophy (toluidine blue, original magnification ×40).

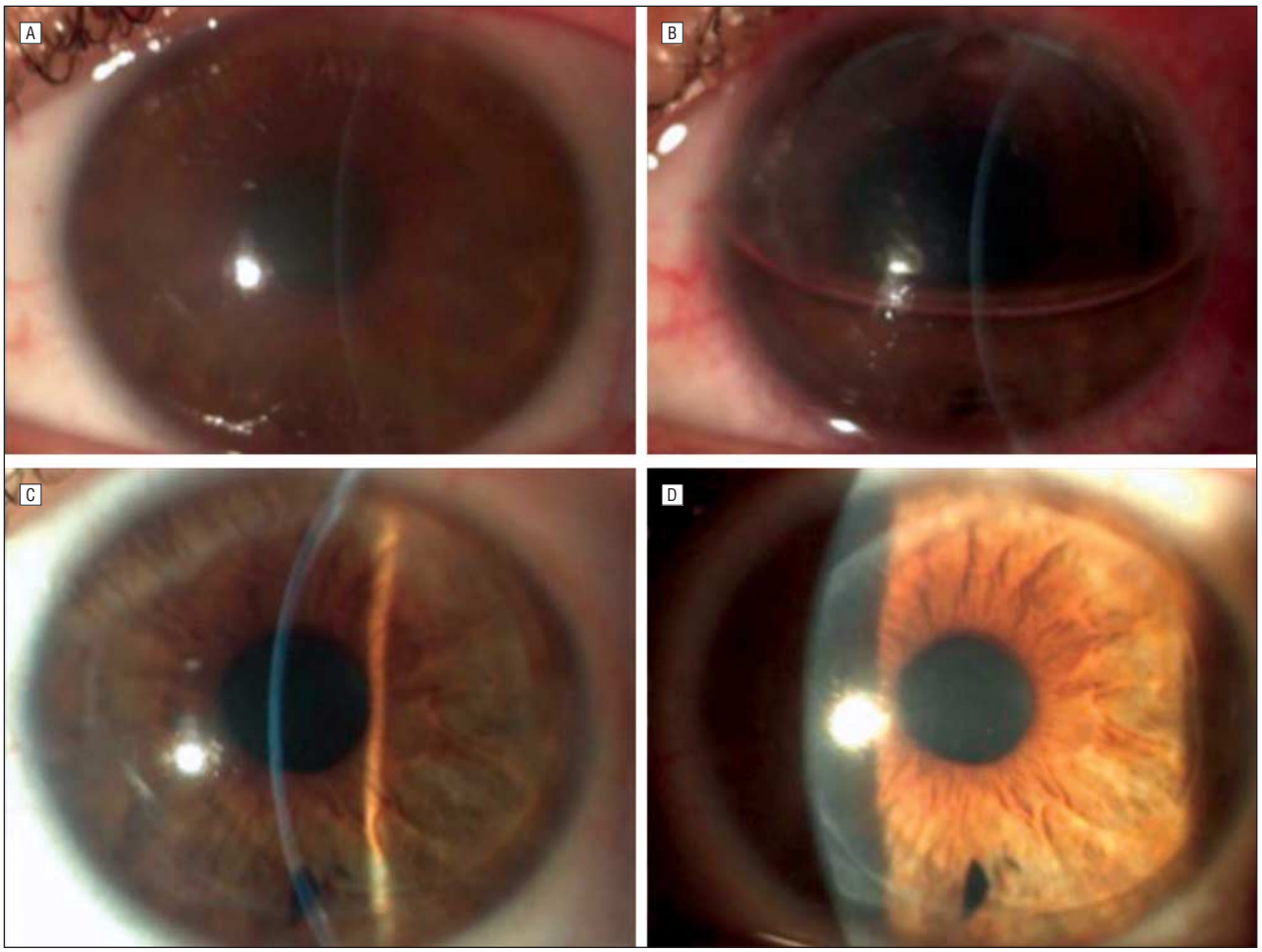

Figure 5. The cornea of a 29-year-old patient with congenital hereditary endothelial dystrophy before and after Descemet-stripping automated endothelial keratoplasty (DSAEK). A, Preoperative diffuse corneal edema is shown. B, Clear cornea is seen 1 day after DSAEK. C and D, Clear cornea with clear interface is seen 30 months after DSAEK.

In children, and even more so in infants, intraoperative vitreous back pressure is high, and open-sky intraocular surgery may result in possible damage to anterior segment structures (eg, iris or crystalline lens) or even devastating complications such as suprachoroidal hemorrhage. In addition, after surgery, a full-thickness corneal wound 7 to 8 mm in diameter is at higher risk of traumatic rupture in children than in adults, given the

Table 2. Summary of the Literature Describing Outcome of Penetrating Keratoplasty in Patients With Congenital Hereditary Endothelial Dystrophy

Source	Eyes/ Patients, No.	Mean Age at Surgery (Range)	Mean Follow-Up	Anatomical Success, %	Functional Success	Percentage			
						Graft Survival at 1 y	Graft Survival 2 y	Graft Survival 3 y	Graft Survival 5 y
Pearce et al,[7] 1969	21/10	42.7 y (22-71 y)	...	19	...	...	15	...	...
Kirkness et al,[3] 1987	31/20	13.5 y	39 mo	90	66% ≥ 20/60	...	...	...	...
Sajjadi et al,[4] 1995	37/21	9.5 y (2-30 y)	3 y	92	57% ≥ 20/80	...	...	...	...
al-Rajhi and Wagoner,[5] 1997	56/40	11.8 y (2 mo-35 y)	37 mo	62.5	20.8% ≥ 20/80	92	72	...	56.5
Schaumberg et al,[2] 1999	16/9 (21 grafts)	Median, 40 mo (3 mo-10 y)	70 mo	69	27% ≥ 20/80	...	71	...	...
Javadi et al,[8] 2003	24/15	8.1 y (SD, 2.5 y)	35.5 mo (SD, 36.2 mo)	79.1	52% ≥ 20/80	...	...	88	74
Al-Ghamdi et al,[6] 2007	35/22	...	Median, 51 mo (range, 12-120)	85.7	57.2% ≥ 20/160	97.1	90.9	83.6	83.6

Abbreviation: ellipses, data not available.

unpredictable nature of their daily activities. For these reasons, surgery has often been avoided or delayed for as long as possible, not only in eyes with severe anterior segment abnormalities (eg, Peter anomaly), but also in eyes with relatively normal anatomy such as CHED, often compromising the visual development of these patients.[16]

In DSAEK, the surgeon uses a "closed-system" technique throughout the procedure and performs all maneuvers through a short clear cornea tunnel. Vision-threatening intraoperative complications, in particular suprachoroidal hemorrhage, are therefore extremely rare and more easily managable.[9] Also, structural integrity of the globe is maintained closer to the natural state, and postoperative wound dehiscence is less frequent[9] and, in any case, unlikely to compromise visual outcome.

Despite these advantages, until recently, only 2 case reports dealing with DSAEK in patients with CHED have been reported worldwide.[17,18] However, the authors of the first of these case reports[17] converted to PK owing to poor visualization through the edematous cornea and the impossibility of stripping the abnormal DM, thus not answering the question of whether DSAEK is a treatment suitable for CHED. The second case report described a successful DSAEK in 1 eye of an adult patient with CHED but visual outcome was poor in the presence of amblyopia.[18]

Our series represents the first report of DSAEK performed in a relatively large number of patients with CHED. The population of this study can be divided into 2 groups with distinct features, namely infants younger than 12 months and older children together with young adults.

In all of the infants, we could not identify any DM and therefore attached the donor graft onto the posterior surface of the recipient cornea, which was left intact. Other authors have reported that normal DM does not hinder attachment of donor tissue to the recipient cornea,[19-21] nor does it seem to influence visual outcome.[19-21] In all our cases, although DM of these young children is thought to be abormal,[1] donor tissue attached to it and the cornea cleared completely within 1 week from surgery. Although in 4 eyes rebubbling was necessary, this may have been owing to many factors including poor or no compliance with postoperative supine posturing. To date, visual outcome in these 6 infants could not be properly evaluated because of the young age, as would also be the case following PK,[16] but fixation was also present early after surgery in the patient who failed to fix and follow preoperatively. However, the rapid achievement of corneal clarity in these patients will hopefully allow for good visual development, and the long-term results and survival of the donor tissue will be closely monitored.

In the other group, including older patients, both surgery and postoperative course did not differ substantially from those of patients with Fuchs dystrophy.[22] Not only could the DM be identified and easily removed, but also restoration of cornea clarity was rapid and paralleled by early visual rehabilitation leading to best-corrected visual acuity of at least 20/40 in 8 of 9 cases, and 20/70 in the remaining case. Our study shows, therefore, that excellent best-corrected visual acuity is possible following DSAEK for older patients with CHED, possibly supporting the theory that CHED does not have such a strong amblyogenic drive as other corneal pathologies do.[3] It is also possible, however, that by selecting older patients with CHED, we have inadvertently chosen patients with milder disease, as patients who are more severely affected may have already presented and undergone surgery at a younger age.

All 15 eyes in this series were phakic, requiring modification of our standard technique for DSAEK. For phakic eyes, many corneal surgeons elect to perform cataract surgery prior to or concurrently with DSAEK. There are, however, many patients with phakic eyes who may benefit from DSAEK without cataract surgery. This particularly applies to children and younger adults with normal accommodative function that can be performed for many years after surgery, such as those in this study. The outcome of DSAEK in adult phakic eyes with endothelial failure described in the literature does not differ substantially from that of DSAEK performed in pseudopha-

kic eyes with postoperative bullous keratopathy.[18,22,23] We have had similar experiences with phakic eyes requiring DSAEK surgery because of primary endothelial failure (Fuchs dystrophy).

In addition to having phakic eyes, children with CHED share with other pediatric patients undergoing intraocular surgery the intraoperative problem of a high vitreous pressure. To help deal with the former of the 2 issues, in most cases in our series, we chose to place the wounds for graft delivery 1 mm above the horizontal meridian, thus avoiding the pass of intraocular instruments across the pupil and using the iris as a barrier to prevent lenticular contact with the instruments should the anterior chamber accidentally shallow or collapse. Furthermore, in contrast to PK, even high levels of posterior pressure were neutralized by the combination of closed-system conditions and continuous irrigation through the anterior chamber maintainer used in DSAEK, creating enough space to perform the surgery safely. To date, no patient included in our series has developed cataract.

Suturing is minimized in DSAEK, providing a further advantage of this procedure compared with PK in the pediatric population. The rapid healing response in children results in frequent loosening of or neovascularization around sutures and may result in irreversible graft failure owing to infection and/or immunologic rejection.[16] Following DSAEK in this population, corneal suture removal can be performed as early as 1 week after surgery and is frequently combined with surgery of the other eye and/or an examination under anesthesia of the eye that already received surgery, thus reducing the total number of general anesthesias.

Owing to the short follow-up available in our preliminary study, we are not able to comment on the survival of DSAEK grafts performed for CHED. In the adult population, Price et al[24] recently reported 5-year survival rates for DSAEK of 95% vs 93% for PK in Fuchs dystrophy and 76% vs 73% for PK in pseudophakic or aphakic bullous keratopathy. No similar data exist for the pediatric group. Although the survival rate for PK in infants is relatively low,[16] PK in CHED is generally accepted to have a much better prognosis,[2-8,16] with a survival rate varying from a low 19% (in 1969[7] when the surgical procedure was much less sophisticated) to 90%[3] (**Table 2**). It is, therefore, conceivable that DSAEK grafts may have the same survival rates as PK for this specific indication.

Despite these high PK survival rates, visual acuity still remains a problem for patients with CHED, and some degree of amblyopia is generally accepted as inevitable.[2-8] Our visual results compare favorably with those previously published for PK performed in patients with CHED.[2-8] However, it is difficult to compare trials directly, given the differences in patient ages and disease severity and/or amblyopia as well as the differing proportions of patients being of verbal age and therefore able to comply with visual acuity testing. The published visual results of PK for CHED are included in Table 2.[2-8]

Randomized controlled trials comparing the visual outcomes of DSAEK and PK for children with CHED, while ideal, are unlikely to be performed given the rarity of this condition and the improved safety profile of DSAEK compared with PK. Likewise, the best timing for surgery is unfortunately not likely to be statistically determined. It is therefore necessary to apply best clinical judgment when considering surgical plans for these patients.

In our experience, DSAEK performed in eyes with CHED reduces intraoperative as well as postoperative risks and consequently allows the surgeon to comfortably face the surgical challenge for younger patients. Rapid achievement of corneal clarity, with earlier initiation of amblyopia treatment, may contribute to improving the visual outcomes of these patients, while early suture removal and retained structural integrity of the eye eliminate most late postoperative complications. These advantages are numerous and, while further studies are required, particularly to determine the long-term survival rates for DSAEK in this population group, we believe that the decision regarding type and timing of surgery for patients with CHED has become easier.

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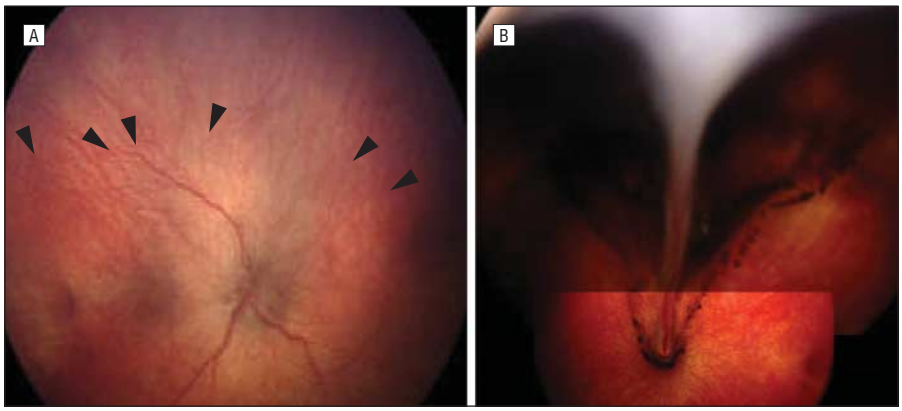
Archives *Web Quiz Winner*

Congratulations to the winner of our April quiz, George Magrath, MD, Storm Eye Institute, Medical University of South Carolina, Charleston. The correct answer to our April challenge was septo-optic dysplasia (de Morsier syndrome). For a complete discussion of this case, see the Research Letters section in the May *Archives* (Kiernan DF, Al-Heeti O, Blair MP, et al. Peripheral retinal nonperfusion in septo-optic dysplasia [de Morsier syndrome]. *Arch Ophthalmol*. 2011;129[5]:671-673).



Be sure to visit the *Archives of Ophthalmology* Web site (http://www.archophthalmol.com) and try your hand at our Clinical Challenge Interactive Quiz. We invite visitors to make a diagnosis based on selected information from a case report or other feature scheduled to be published in the following month's print edition of the *Archives*. The first visitor to e-mail our Web editors with the correct answer will be recognized in the print journal and on our Web site and will also be able to choose one of the following books published by AMA Press: *Clinical Eye Atlas*, *Clinical Retina*, or *Users' Guides to the Medical Literature*.