Use of Decellularized Descemet Membrane Anterior Keratoplasty to Facilitate Epithelialization of Pediatric Penetrating Keratoplasty for Total Sclerocornea

Radwa Elsharawi, MD, MSc, Daniel Lee, MD,† Abhiniti Mittal, MD,**
Winston D. Chamberlain, MD, PhD, Joseph Tauber, MD,‡ and Travis K. Redd, MD, MPH**

Purpose: To report the first known use of combined decellularized Descemet membrane anterior keratoplasty with penetrating keratoplasty (PKP) in a pediatric patient.

Methods: A 2-month-old boy with bilateral sclerocornea underwent bilateral sequential PKP. In the left eye, PKP performed at 2 months of age was complicated by a persistent epithelial defect postoperatively with a 1 month delay in epithelialization. As a result, the patient underwent PKP with a combined decellularized Descemet membrane corneal allograft implantation in the right eye at 3 months of age to enhance early postoperative healing. This was performed by creating a central 3-mm superficial keratectomy before placing Descemet membrane allograft onto the full-thickness graft.

Results: Compared to the 1-month delay in epithelialization after PKP in the left eye, the right eye, which underwent combined PKP and decellularized Descemet membrane corneal allograft, was fully epithelialized by the first postoperative week. The grafts remain clear and intact at 12 months of age. The patient exhibited significant improvement in visual behavior.

Conclusions: This case highlights the successful use of decellularized Descemet membrane anterior keratoplasty in preventing a persistent epithelial defect in the early postoperative period for a pediatric patient with sclerocornea. This may be a viable option for similar pediatric cases with delayed epithelial healing while promoting graft survival and minimizing the need for additional surgical interventions.

Key Words: sclerocornea, corneal transplant, corneal epithelium, Descemet membrane

(*Cornea* 2025;00:1–3)

Sclerocornea is a condition characterized by bilateral scleralization and opacification of the cornea. Vision can be preserved in cases with peripheral involvement sparing the central visual axis, but complete scleralization typically causes severe deprivation amblyopia and often warrants surgical intervention. However, penetrating keratoplasty (PKP) surgery is technically challenging in infants and carries a much higher rate of graft rejection and failure when compared with adults.[1–3] In addition to the risk of rejection, postoperative persistent epithelial defects are common. One study reviewing 165 cases of pediatric primary keratoplasties in children aged 12 and younger found that persistent epithelial defects occurred in 12.7% of cases and were associated with lower rates of graft survival.[4] A decellularized corneal Descemet membrane allograft has been shown to promote corneal reepithelialization.[5] To our knowledge, this is the first case report to describe the implantation of combined Descemet membrane anterior keratoplasty (DMAK) with PKP in a pediatric patient.

CASE PRESENTATION

A 2-month-old boy with bilateral total sclerocornea was born at 39 gestational weeks to a pregnancy complicated by fetal hydronephrosis. Wide genome testing revealed 7q3G.3 deletion and 5p15.32 duplication. Exam was notable for nystagmus and diffuse corneal opacification with fine vessels, an indistinct limbus and no discernible clear cornea in OU (Fig. 1). There was no view to the anterior chamber or any other intraocular structures. Ultrasound biomicroscopy and B-scan ultrasonography showed otherwise normal anterior and posterior segment anatomy. The patient underwent a bilateral sequential PKP in the left eye at 2 months of age, followed by the right eye at 3 months of age. In the left eye, a 6.5-mm donor punched PKP graft was placed in a 6.0-mm trephination site. Sixteen interrupted 10-0 nylon sutures were placed radially to secure the graft in place. A 9-mm epithelial defect was noted postoperatively at week 1. The patient was monitored with a steroid drop taper and antibiotic coverage, resulting in full epithelialization by postoperative month 1. At 3 months of age, patient was taken to the

Received for publication January 28, 2025; revision received March 30, 2025; accepted April 8, 2025.

From the *Casey Institute, Cornea Division, Oregon Health & Science University, Portland, OR; †EyeHealth Northwest PC, Portland, OR; and ‡Tauber Eye Center, Kansas City, MO.

Supported by the National Eye Institute (P30 EY010572, K23 EY032639), Research to Prevent Blindness (Tom Wertheimer Career Development Award in Data Science and unrestricted departmental funding), and the Malcolm M. Marquis, MD, Endowed Fund for Innovation. The sponsor or funding organizations had no role in the design or conduct of this research.

The authors have no funding or conflicts of interest to disclose.

J. Tauber, MD, is the CEO of Brightstar Therapeutics. W. D. Chamberlain, MD, PhD, is a paid consultant of Brightstar Therapeutics.

Correspondence: Travis K. Redd, MD, MPH, Casey Eye Institute, Oregon Health & Science University, 515 SW Campus Drive, Portland, OR 97239-4197 (e-mail: redd@ohsu.edu).

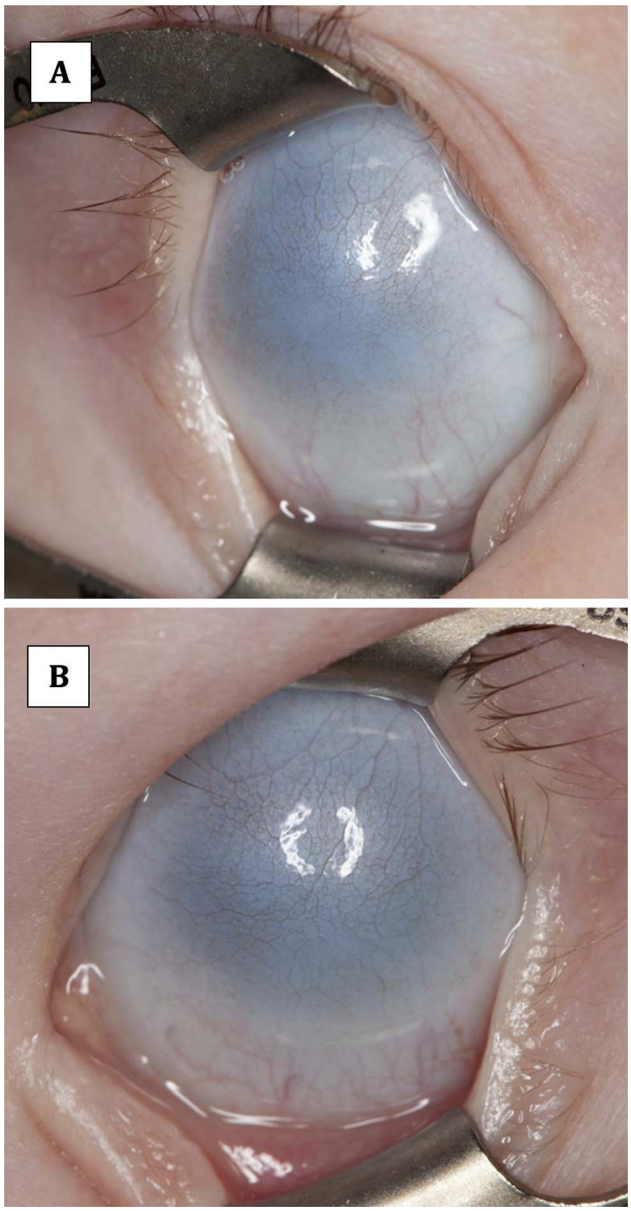

FIGURE 1. External photograph of the right (A) and left eye (B) preoperatively.

operating room for exam under anesthesia, suture removal in the left eye, and PKP of the right eye. In the right eye, the decision was made to perform a combined decellularized DMAK (BrightMEM, Brightstar Therapeutics, Lexington, KY) given delayed epithelialization in the left eye. Similar to the PKP in the left eye, a 6.0-mm trephination and 6.5-mm donor punch were used in the right eye. After the full-thickness graft was secured with 16 interrupted 10-0 nylon sutures, a central 3-mm superficial keratectomy was performed and a 3-mm donor punch was selected for Descemet membrane allograft. The allograft was then placed over the epithelial defect with Descemet membrane facing posteriorly and pre-Descemet facing anteriorly. Thrombin/fibrinogen glue was then applied over the graft and left to dry. This was followed by the placement of a bandage contact lens. Pathology report of the corneal buttons from OU revealed an absent Bowman layer and the entire thickness of the stroma was replaced by irregularly organized fibers consistent with scleral tissue. Descemet membrane and the corneal endothelium were unremarkable. All sutures were removed by the end of the 2-month period after each surgery. The epithelial defect in the left eye resolved after 1 month while the right eye was fully epithelialized within 7 days. The grafts remained clear in OU at 10 months after PKP in the left eye and 9 months after PKP in the right eye (Fig. 2). The infant's visual behavior and nystagmus have improved, and he is tolerating rigid gas-permeable contact lenses.

DISCUSSION

PKP for infants with sclerocornea is necessary to prevent visual deprivation in early months of life. Although it remains challenging, PKP has yielded more successful outcomes in children with congenital corneal opacities in recent years.[3] In this case, bilateral sequential PKP was performed in an infant with sclerocornea where the postoperative course was complicated by a slow-healing epithelial defect in 1 eye, prompting combined decellularized DMAK with PKP in the fellow eye. To our knowledge, this is the first case of DMAK use in a pediatric patient. Delayed postoperative epithelialization is common in this population, which can increase susceptibility to infections and lead to unfavorable outcomes including keratolysis, neovascularization, graft thinning, rejection, and scarring.[6] This further increases the already substantial risk of graft failure in infants. A degree of limbal stem cell deficiency may be present in cases of sclerocornea,[7] although a study by Ma et al,[8] using transmission electron microscopy and immune-confocal microscopy found predominantly corneal epithelial cells in the corneal buttons of 4 cases with total sclerocornea. A study assessing PKP outcomes in 58 patients with sclerocornea,

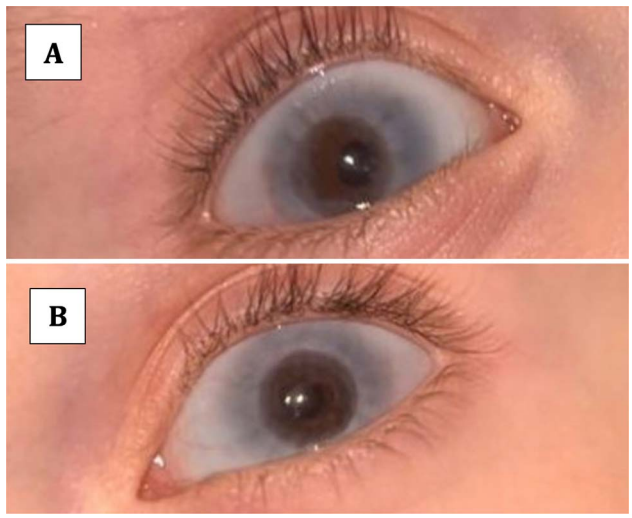

FIGURE 2. External photograph of the right (A) and left (B) eye taken at 9 and 10 months after PKP, respectively.

Peters anomaly and congenital glaucoma found 3 patients with delayed epithelial healing in the early postoperative period, all of whom were in the sclerocornea cohort.[2] Regrafting was performed in all 3 eyes because of high risk of vascularization and opacification.[2] We highlight a case where combined decellularized DMAK and PKP successfully avoided this complication.

The use of decellularized DMAK is an innovative approach that may facilitate corneal epithelialization. Decellularized Descemet membrane provides a substrate for corneal epithelial cell migration and prevention of stromal degradation. By expressing limbal basement membrane–specific proteins such as vitronectin and SPARC/BM-40, it may create a niche-like environment that enhances stem cell longevity and migration to the central cornea, maintaining a reservoir of epithelial stem cells on the ocular surface.[9] BrightMEM is a decellularized Descemet membrane tissue product that is classified by the US Food and Drug Administration as an Human Cells, Tissues, and Cellular and Tissue–Based Product (HCT/P). As an HCT/P, BrightMEM is processed by an US Food and Drug Administration–registered facility that is accredited by the Eye Bank Association of America. The tissue is processed using a validated and proprietary procedure to ensure a consistent HCT/P with a specific purpose of facilitating reepithelialization. Intermediate-term storage solutions are used in conditions similar to whole corneas or tissue processed for DMEK or Descemet stripping automated endothelial keratoplasty. The allograft is processed within a few days of surgery and shipped in validated coolers. After removal of superficial scarring, pannus, and/or neovascularization, the allograft is placed on the corneal surface with anterior side up, smoothed and allowed to adhere. A small amount of tissue glue is applied to seal edges before a bandage contact lens is applied.

Although there are other conventional treatments for delayed epithelial healing including lubrication with preservative-free tears and autologous serum tears,[10] increasing the drop burden in pediatric patients may limit adherence, which can subsequently threaten graft survival. Amniotic membrane transplantation has also been shown to be beneficial in treating persistent epithelial defects after PKP,[6] although with a high risk of recurrence, likely secondary to its rapid breakdown.[5] When compared with amniotic membrane, Descemet membrane submerged in high-dose collagenase was significantly more resistant to degradation.[5] This makes it a more durable substrate for epithelial healing.[5] Descemet membrane is also more optically transparent and provides additional structural integrity to the cornea.[5] One study showed successful use of DMAK with allogenic simple limbal epithelial transplantation for limbal stem cell deficiency. The membrane was present 1.5 years after transplantation.[5]

In conclusion, this report demonstrates the potential benefit of adjunctive decellularized DMAK in pediatric keratoplasty patients to prevent delayed epithelial healing. Future prospective studies are warranted to further evaluate this approach.

REFERENCES

1. Stulting RD, Sumers KD, Cavanagh HD, et al. Penetrating keratoplasty in children. *Ophthalmology*. 1984;91:1222–1230.
2. Frueh BE, Brown SI. Transplantation of congenitally opaque corneas. *Br J Ophthalmol*. 1997;81:1064–1069.
3. Comer RM, Daya SM, O'Keefe M. Penetrating keratoplasty in infants. *J AAPOS*. 2001;5:285–290.
4. Al-Ghamdi A, Al-Rajhi A, Wagoner MD. Primary pediatric keratoplasty: indications, graft survival, and visual outcome. *J AAPOS*. 2007;11:41–47.
5. Cheung AY, Reinisch CB, Hou JH. Decellularized Descemet membrane anterior keratoplasty with allogeneic simple limbal epithelial transplantation for partial limbal stem cell deficiency following partial keratolimbal allograft failure. *Cornea*. 2025;44:108–112.
6. Seitz B, Das S, Sauer R, et al. Amniotic membrane transplantation for persistent corneal epithelial defects in eyes after penetrating keratoplasty. *Eye (Lond)*. 2009;23:840–848.
7. Skeens HM. Congenital stem cell deficiency. *Ocular Surface Disease: Cornea, Conjunctiva, and Tear Film*. Elsevier Health Sciences; 2013: 251–259. E-Book: Expert Consult-Online and Print.
8. Ma DH, Yeh LK, Chen HC, et al. Epithelial phenotype in total sclerocornea. *Mol Vis*. 2014;20:468–479.
9. Shukla R, Tauber J. *Booth talk and surgical demonstration. BrightMEM anterior keratoplasty*. Presented at: ASCRS; 2024; Boston, MA. https://www.youtube.com/watch?v=96_FVSX-36s&t=95s
10. Chen YM, Hu FR, Huang JY, et al. The effect of topical autologous serum on graft re-epithelialization after penetrating keratoplasty. *Am J Ophthalmol*. 2010;150:352–359.e2.