DJO Digital Journal of Ophthalmology
www.djo.harvard.edu

Digital Journal of Ophthalmology

Editorials

Boston Keratoprosthesis: expanding the boundaries

Mona Harissi-Dagher, MD, FRCSC, DABO

Author affiliations: Department of Ophthalmology, University of Montreal

Congenital corneal opacities (CCOs), occurring in approximately 3/100,000 newborns, result from many different disorders, including congenital hereditary endothelial dystrophy (CHED), Peters anomaly (PA), congenital hereditary stromal dystrophy (CHSD), and posterior polymorphous dystrophy (PPMD). CCOs cause visual deprivation during the early months of life that can result in long-term changes to the central nervous system.[1] This may result in profound and uncorrectable loss of vision that can negatively affect a child's development. Early detection is important to begin appropriate and prompt medical or surgical therapy and minimize amblyopia risk in these children.

In the past, penetrating keratoplasty (PK) has been performed to prevent these potentially devastating consequences of CCOs; however, traditional PK is associated with a high incidence of allograft rejection and complications.[2–5] Additionally, a poorer prognosis has been described in children with Peters anomaly and sclerocornea compared to those with acquired corneal opacities.[6] In fact, the concurrence of comorbid conditions such as of glaucoma, retinal disease, and anterior segment dysgenesis often requires additional intraocular surgeries, which are known to increase the risk of corneal decompensation.[7,8] The duration and severity of the initial CCO, the postoperative induced irregular astigmatism, the high risk of graft rejection and subsequent graft failure render children with CCOs at high risk for refractive and sensory deprivation amblyopia.

The Boston Keratoprosthesis (KPro) has enjoyed good results in the adult population, particularly through its rapid clearing of the visual axis, its excellent retention rate, and the paucity of postoperative complications in recent years.[9–12] Design modifications, enhanced surgical technique, and improved postoperative management have made keratoprosthesis surgery a reality in the twenty-first century and an excellent alternative for poor PK candidates.[13–18]

CCOs in children, although infrequent, continue to be challenging to manage.[19] The ability to achieve a quiet and comfortable eye with a clear visual axis and stable refraction within days following Boston KPro surgery is a significant advantage in pediatric corneal transplantation and plays an even more important role in children at high risk for amblyopia. The clear optical stem of the Boston KPro, with its spherical cut, eliminates regular and irregular astigmatism associated with PK and allows a best-corrected visual acuity soon after surgery. Conveniently, this refractive error can be corrected through the soft contact lens. The availability of aphakic powered KPros manufactured to conform to the axial length of the eye avoids the added complexity associated with intraocular lens (IOL) implantation in this age group. In addition, the Boston KPro is available in pseudophakic powers suitable for those children who already have intraocular lenses (IOLs). Furthermore, the Boston KPro is made out of polymethyl methacrylate (PMMA), an immunologically inert material, eliminating allograft rejection and its consequent inflammation, discomfort, and interference with amblyopia therapy. The Boston KPro may be a major step forward in corneal transplantation since children are known to mount an amplified inflammatory response and graft rejection may progress rapidly and be medically less responsive.

In their case report "Keratoprosthesis in congenital hereditary endothelial dystrophy after multiple failed grafts," Haddadin and Dohlman[20] discuss the outcome of KPro surgery for the management of CHED in a patient with multiple graft failures. The report demonstrates the favorable progress, over a 5-year span, of this 18-year-old patient with 20/30 vision and no glaucoma. CHED has historically been managed with penetrating keratoplasty, with moderate success, and, more recently,

Published November 03, 2011.

doi:10.5693/djo.04.2011.10.004

Correspondence: Mona Harissi-Dagher, MD, FRCSC, DABO, Department of Ophthalmology, CHUM, Notre Dame Hospital 1560 Sherbrooke East, Montreal H2L 4M1 (email: monadagher@hotmail.com)

Digital Journal of Ophthalmology

with Descemet's stripping endothelial keratoplasty (DSEK),[21] albeit a challenging surgical technique in this disease. As the authors note, the history of multiple failed grafts illustrates the lower success rate following PK for CCO. The likelihood of repeated graft failures with CHED, therefore, makes alternative surgical procedures a necessity. This case report represents successful management of CHED via KPro in an adult who had undergone a total of 13 PKs in hopes of visual rehabilitation. Certainly in CHED, KPro implantation deserves to be explored further, both in adult and pediatric patients and much earlier in time. As with congenital cataracts, clearing of the visual axis early on is crucial to avoid amblyopia. Theoretically, surgery at the youngest age possible would be best to avoid irreparable occlusion amblyopia and nystagmus. This is our impression as well with the Boston KPro. As in the case of Haddadin and Dohlman's patient, several PK surgeries can be avoided and visual rehabilitation can be accelerated if KPro surgery is considered early on.

The Boston KPro is appropriate for implantation in pediatric cases and may sometimes be the procedure of choice to quickly establish a clear optical pathway, reduce the potential for reoperation and complications, and assist in the process of amblyopia prevention and therapy. The increased ocular morbidity associated with concurrent glaucoma and vitreoretinal diseases continue to put children with CCOs at high risk for failure of visual restoration. Furthermore, strict control of ocular inflammation is essential. These abnormally developed eyes limit visual potential. Since the corneal leukoma precludes an accurate evaluation of the rest of the eye, lifting the CCO is the crucial step in visual rehabilitation.

While adult KPro surgery has been performed for decades, dealing with the multiple associated pathologies common in children with CCOs often require additional expertise and resources. Thus a team approach is needed, with close coordination among corneal, vitreoretinal, glaucoma, and pediatric specialists for preoperative evaluation, surgery, and postoperative care. In addition, the commitment of the parents to their child's long-term care after surgery is crucial to a successful outcome.

References

1. Rezende RA, Uchoa UC, Uchoa R, et al. Congenital corneal opacities in a cornea referral practice. Cornea 2004;23:565-70.
2. Aasuri MK, Garg P, Gokhle N, Gupta S. Penetrating keratoplasty in children. Cornea 2000;19:140-4.
3. Comer RM, Daya SM, O'Keefe M. Penetrating keratoplasty in infants. J AAPOS 2001;5:285-90.
4. Michaeli A, Markovich A, Rootman DS. Corneal transplants for the treatment of congenital corneal opacities. J Pediatr Ophthalmol Strabismus 2005;42:34-44.
5. McClellan K, Lai T, Grigg J, Billson F. Penetrating keratoplasty in children: visual and graft outcome. Br J Ophthalmol 2003;87:1212-4.
6. Yang LL, Lambert SR, Lynn MJ, et al. Long-term results of corneal graft survival in infants and children with Peters' anomaly. Ophthalmology 1999;106:833-48.
7. Dana MR, Moyes AL, Gomes JAP, et al. The indications for and outcome in pediatric keratoplasty. Ophthalmology 1995;102:1129-38.
8. Dana MR, Schaumberg DA, Moyes AL, et al. Corneal transplantation in children with Peters anomaly. Ophthalmology 1987;104:545-6.
9. Ma JJ, Graney JM, Dohlman CH. Repeat penetrating keratoplasty versus the Boston keratoprosthesis in graft failure. Int Ophthalmol Clin 2005;45:49-59.
10. Aquavella JV, Qian Y, McCormick GJ, Palakuru JR. Dohlman Keratoprosthesis: current techniques. Cornea 2006;25:656-62.
11. Dohlman CH, Harissi-Dagher M, Khan BF, Sippel KC, Aquavella J, Graney JM. Introduction to the use of the Boston Keratoprosthesis. Expert Review of Ophthalmology 2006;1:41-8.
12. Robert MC, Harissi-Dagher M. Boston type 1 keratoprosthesis: the CHUM experience. Can J Ophthalmol 2011;46:164-8.
13. Bothelo PJ, Congon WG, Handa JT, Akpek EK. Keratoprosthesis in high-risk pediatric corneal transplantation: first 2 cases. Arch Ophthalmol 2006;124:1356-7.
14. Aquavella JV, Gearinger MD, Akpek EK, McCormick GJ. Pediatric keratoprosthesis. Ophthalmology 2007;114:989-94.
15. Khan, B.; Harissi-Dagher, M.; Dohlman, CH. Chapter 67, Keratoprosthesis. In: Albert, DM.; Miller, JW.; Azar, DT.; Blodi, BA., editors. *Albert and Jakobiec's Principles and Practice in Ophthalmology*. 3rd ed ed. Philadelphia, PA; Edinburgh: Saunders/Elsevier; 2008.
16. Harissi-Dagher M, Khan BF, Schaumberg DA, Dohlman CH. The importance of nutrition to corneal grafts when used as a carrier of the Boston. Keratoprosthesis 2007;26:564-8.
17. Khan BF, Harissi-Dagher M, Dohlman CH. Advances in Boston Keratoprosthesis: enhancing retention and prevention of infection and inflammation. Int Ophthalmol Clin 2007;47(2):61-71.
18. Harissi-Dagher M, Dohlman CH. The Boston Keratoprosthesis. Contemp Ophthalmol 2006;5:1-8.
19. Harissi-Dagher M, Colby K. Anterior segment dysgenesis: Peters anomaly and sclerocornea. Int Ophthalmol Clin 2008;48(2):35-42.
20. Haddadin R, Dohlman CH. Keratoprosthesis in congenital hereditary endothelial dystrophy after multiple failed grafts. Epub September 23, 2011. Digital J Ophthalmol 2011;17(3)
21. Fernandez MM, Buckley EG, Afshari NA. Descemet stripping automated endothelial keratoplasty in a child. J AAPOS 2008;12:314-6.