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Visual Outcomes and Complications of Type I Boston Keratoprosthesis in Children

A Retrospective Multicenter Study and Literature Review

Simon S.M. Fung, MA, FRCOphth,[1] Samir Jabbour, MD,[2] Mona Harissi-Dagher, MD, FRCSC,[2] Reginald R.G. Tan, MD,[3] Patrick Hamel, MD, FRCSC,[4] Kashif Baig, MD, FRCSC,[3] Asim Ali, MD, FRCSC[1]

Purpose: To report outcomes and complications of Boston type 1 keratoprosthesis (KPro) implantation in children.

Design: Retrospective, multicenter case series.

Participants: All children 16 years of age or younger who underwent KPro surgery at 3 ophthalmology centers in Canada between January 2010 and November 2014.

Methods: Records of patients having undergone KPro implantation were reviewed. Data on preoperative characteristics, surgical procedure(s) performed, and postoperative outcomes were collected and analyzed.

Main Outcome Measures: Intraoperative and postoperative complications, device retention, and best-corrected visual acuity (BCVA).

Results: The KPro was implanted in 11 eyes of 11 patients 0.9 to 15.5 years of age, with 6 being primary corneal procedures. Best-corrected visual acuity recorded before surgery ranged from 20/125 to light perception (LP), and vision in 2 eyes was fix and follow. All patients had been diagnosed with glaucoma and 6 eyes had glaucoma drainage devices (GDDs) inserted before KPro implantation. At last follow-up (mean, 41.8 months; range, 6.5–85.0 months), 2 eyes retained BCVA of 20/400 or better, whereas 5 eyes lost LP. Postoperative complications included retroprosthetic membrane (9 eyes), corneal melt (5 eyes), infectious keratitis (3 eyes), endophthalmitis (3 eyes), GDD erosion (2 eyes), and retinal detachment (5 eyes). The initial KPro was retained in 4 eyes (36.4%).

Conclusions: Boston type 1 keratoprosthesis implantation in children is associated with a substantially higher rate of complications, higher chance of device failure, and worse visual outcomes than observed in adults. In view of these results, the authors do not recommend the use of the KPro in the pediatric population. *Ophthalmology 2017;■:1–8 © 2017 by the American Academy of Ophthalmology*

The Boston type 1 keratoprosthesis (KPro) has become a viable alternative to traditional penetrating keratoplasty (PKP) in the management of severe adult corneal pathologies.[1–4] Since its introduction, the KPro design and management of patients after implantation have improved,[5,6] which has translated into improvements in visual outcomes and reduction of complications.[2–4] In a recent review for the American Academy of Ophthalmology, Lee et al[7] summarized the results from 22 articles rated as level II (well-designed case-control and cohort studies and randomized clinical trials with substantial methodologic deficits) to level III (case series, case reports, and poor-quality cohort and case-control studies) clinical evidence. Among these reports, 69% to 81% of the study patients reached 20/100 or better best-corrected visual acuity (BCVA); remarkably, in a handful of studies, BCVA of 20/40 or better was achieved in 11% to 39% of patients. Furthermore, the KPro was retained in 65% to 100% of patients, with an average retention rate of 88%, although caution was advised by the authors in view of the relatively short follow-up in some of the reports with favorable outcomes.

Indications for the KPro have broadened beyond being a last resort procedure for eyes that have undergone multiple failed PKPs. Eyes with trauma, herpetic keratitis, limbal stem cell deficiency, aniridia, and cicatrizing diseases all have been reported to have successful visual rehabilitation after KPro implantation.[5,8] Consequently, clinicians have started using KPro as a primary corneal procedure in patients with a low probability of success with conventional PKP.[9–12]

The use of the KPro in the pediatric population is less well explored. Keratoprosthesis has the theoretical advantages of improving visual axis clarity rapidly while eliminating the risks and consequences of allograft rejection; thus, it could be a good alternative to an otherwise challenging PKP procedure in children.[13] However, currently there is a paucity of data regarding the outcomes and complications of pediatric KPro implantation. In the present study, we reviewed the

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visual outcomes, device retention rates, and complications after KPro implantation in a pediatric population at 3 tertiary university-based centers in Canada.

Methods

This study was approved by the institutional review boards at the 3 tertiary referral centers in Canada (Hospital for Sick Children, Toronto; University of Ottawa Eye Institute, Ottawa; and CHU Ste-Justine, Montréal) in accordance with the tenets of the Declaration of Helsinki. All pediatric patients (≤16 years of age at time of surgery) undergoing KPro implantation between January 2010 and November 2014 were included in this study. Before the procedure, a complete history was obtained and an ophthalmologic examination and A- and B-mode ultrasound scans were performed. In most patients, a preoperative examination under anesthesia was required.

All surgeries were performed by experienced corneal surgeons at 3 tertiary centers (A.A., K.B., M.H.-D.). The KPro devices were obtained from Massachusetts Eye and Ear Infirmary (Boston, MA) and were implanted using the standard technique.[14] Lensectomy and anterior vitrectomy were performed concurrently in phakic patients. In aphakic patients, anterior vitrectomy was performed as part of the procedure. When required, subtotal iridectomy also was performed to allow KPro insertion. In addition, pars plana vitrectomy and repeat silicone oil tamponade was performed in patient 10, who had undergone vitrectomy for retinal detachment repair previously, and redetachment of the retina was noted after silicone oil removal during KPro insertion. After surgery, all patients received a bandage contact lens for comfort (Kontur [Kontur Kontact Lens Co., Hercules, CA] or Acuvue Oasys [Vistakon, Jacksonville, FL]).

After surgery, all patients were maintained indefinitely on topical prednisolone acetate 1% (Sandoz Canada, Inc., Boucherville, Canada), ranging from 1 to 4 times daily, and moxifloxacin 0.5% (Vigamox; Alcon Canada, Inc., Mississauga, Canada). In 5 patients (patients 2−6), additional topical vancomycin (14 mg/ml from a compounding pharmacy) or polymyxin B sulfate 10 000 units and trimethoprim 1 mg combination (Polytrim; Allergan Canada, Markham, Canada) also were part of the postoperative regimen. Preoperative glaucoma medication was continued, and the regimen was modified as needed according to glaucoma progression. Follow-up visits were scheduled at minimum every 3 months. The contact lens was changed at each follow-up visit.

The main outcome measures in this study were visual outcome, device retention, and complications. Information from each patient, including demographics, clinical course, and visual acuity (measured as BCVA in logarithm of the minimum angle of resolution units [logMAR]) with age-appropriate testing methods, was collected retrospectively and entered in a uniform Microsoft Excel spreadsheet (Microsoft Corp., Redmond, WA) at each site. Visual acuity testing methods included Teller acuity cards for preverbal children, Lea symbols for preschool children, the HOTV vision test for 3- to 5-year-olds, and Snellen or logMAR visual acuity testing chart in children 5 years of age or older. De-identified data were reviewed for completeness and consistency by the first author (S.S.M.F.). The visual acuity data collected included last preoperative, best postoperative, and final BCVA at last follow-up. Statistical analyses were performed using Microsoft Excel software and SPSS software version 22 (IBM SPSS Statistics, Chicago, IL). Statistical significance was determined at $P < 0.05$ with the Fisher exact test.

Results

Preoperative Characteristics

Eleven eyes of 11 patients were included in this study. Median follow-up was 26.7 months (range, 6.5−85.0 months). The baseline characteristics are summarized in Table 1. The median age of the patients at the time of surgery was 4.7 years (range, 0.8−15.6 years). Seven of the patients (64%) were male. All patients underwent unilateral surgery, with 6 eyes being the right eye.

The most frequent indication for KPro surgery was prior donor graft failure in 5 eyes (45%), 4 of which had undergone prior PKP and 1 of which had undergone prior anterior lamellar keratoplasty. The remainder of the 6 eyes (55%) underwent KPro implantation as a primary corneal procedure in view of high risk of donor graft failure with conventional PKP. The primary diagnoses were Peters anomaly type II, in which lenticular abnormalities were present in conjunction with the congenital corneal opacity (5/11), aniridia (5/11), and sclerocornea (1/11). All eyes had a history of glaucoma: 5 eyes had advanced glaucomatous optic neuropathy, whereas 6 eyes had prior glaucoma drainage device (GDD) insertion. Five eyes were aphakic after prior combined lensectomy and anterior vitrectomy procedures. Visual acuity before KPro surgery included the following: BCVA of 20/600 (n = 1), counting fingers (n = 1), hand movements (n = 2), and light perception (LP; n = 7).

Intraoperative Variables

All patients received the aphakic KPro model with the same 3-piece threadless snap-on design along with C-shaped locking ring. Patient 2 received a 16-fenestration titanium back plate, whereas all others received the 16-fenestration polymethyl methacrylate back plate model. In 2 eyes (patients 4 and 5), adult backplates (8.5 mm) were used, whereas the others received 7-mm pediatric backplates. The size of the carrier corneal donor graft ranged from 7.25 to 8.5 mm. Concomitant procedures at the time of KPro implantation included lensectomy, anterior vitrectomy, GDD insertion, goniosynechialysis, and repair of retinal redetachment with pars plana vitrectomy and silicone oil tamponade.

Visual Outcomes

Best postoperative BCVA was achieved at a mean of 4.0 months (range, 0.0−24.7 months), with 6 eyes (55%) achieving better vision than the preoperative BCVA. However, at the last follow-up, vision in 6 eyes (55%) was worse than the preoperative level (Fig 1). Moreover, 5 eyes (45%) lost LP (no light perception visual acuity) and 2 eyes (18%) were phthisical (patients 1 and 4). Between patients who underwent KPro implantation after failed keratoplasty and those who had undergone KPro as a primary corneal procedure, no statistical differences could be found in terms of visual acuity loss ($P = 0.57$) or loss of LP ($P = 0.24$).

Postoperative Complications and Management

Table 1 summarizes the postoperative complications and secondary procedures. Nine eyes (82%) had formation of retroprosthetic membrane (RPM), 5 of which were treated with neodymium:yttrium−aluminum−garnet laser. In the other 4 patients treated with surgical membranectomy, 3 procedures were combined with vitreoretinal surgery. The second most common