Results of Deep Anterior Lamellar Keratoplasty for Advanced Keratoconus in Children Less Than 18 Years

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• PURPOSE: To evaluate the outcomes of deep anterior lamellar keratoplasty (DALK) in children with advanced keratoconus.

• DESIGN: Retrospective noncomparative interventional case series.

• METHODS: A retrospective analysis was conducted of 20 eyes of 16 patients who underwent deep anterior lamellar keratoplasty at a tertiary eye care center for advanced keratoconus. The main preoperative parameters measured were decimal uncorrected visual acuity (UCVA), decimal best-corrected visual acuity (BCVA), and average keratometry. The average keratometry was taken as an average of the flat and steep keratometric readings. The follow-up ranged from 24 to 105 months. The main outcome measures were UCVA, BCVA, steep and flat simulated keratometry, refraction, graft clarity, and complications.

• RESULTS: At the final follow-up, 18 patients had clear grafts in the visual axis area. Mean decimal UCVA changed from 0.06 ± 0.08 to 0.24 ± 0.09 . Mean decimal BCVA changed from 0.12 ± 0.19 to 0.45 ± 0.24 . The average keratometry decreased from 64.15 diopter (D) to 45.7 D after surgery. The complications noted in the patients were graft rejection (1), shield ulcers (2), graft infection (2), and interface vascularization (4). They were managed accordingly and only 1 patient needed penetrating keratoplasty.

• CONCLUSIONS: DALK is an effective therapeutic modality for the management of advanced keratoconus in children. (Am J Ophthalmol 2016;162:191–198. © 2016 by Elsevier Inc. All rights reserved.)

ERATOCONUS IS A BILATERAL NONINFLAMMATORY progressive corneal ectasia with onset at puberty. It progresses until the third to the fourth decade and stabilizes thereafter. Diagnosis is commonly made at adolescence, despite the process of the disease beginning at a much younger age. The presentation of keratoconus in children is unique when compared to adults. The disease progresses at a rapid rate, very often presents in an advanced stage, and frequently has accompanying vernal keratoconjunctivitis (VKC).^{1,2} A higher rate of incidence and prevalence with an earlier onset and greater severity has been reported in certain Asian ethnicities as compared to the white population.³

The management of advanced keratoconus in children is mainly surgical and the visual outcomes after penetrating keratoplasty (PK) in these patients have been reported to be good.⁴ More than 75% of the penetrating grafts performed in adolescents aged between 13–19 years for keratoconus have been reported to achieve visual acuity of at least 20/40 and graft survival of more than 90% at 10 years postoperatively.⁵

The results of pediatric PK are poorer when compared to results in adults. This is mainly due to the reports of higher chances of graft rejection in pediatric corneal grafts.⁶

Deep anterior lamellar keratoplasty (DALK) is a newer alternative procedure for stromal corneal pathologies and has gained popularity over penetrating keratoplasty. The long-term visual outcomes, after penetrating keratoplasty and deep anterior lamellar keratoplasty for advanced keratoconus, in adults have been found to be comparable.^{7,8} The latter has obvious advantages in terms of retained host endothelium, lesser incidence of graft rejection, and the need for shorter duration of postoperative antiinflammatory medications.⁹ These advantages of DALK are likely to provide a definite edge over conventional PK in children, as the expected life of grafts is higher. There has been very scant literature on outcomes of DALK in children.^{10,11} Neither a large series nor any studies with a longer follow-up of DALK in keratoconus in children are available. We hereby report results of DALK in children with keratoconus from the Asian subcontinent with follow-up ranging from 2 to 9 years.

METHODS

AN INTERVENTIONAL CASE SERIES COMPRISING 20 EYES OF 16 patients (less than 18 years of age) with advanced keratoconus who underwent deep anterior lamellar keratoplasty between January 1, 2004 and March 1, 2014 was analyzed retrospectively. Approval to use the patients'

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data was taken from the Institutional Review Board/Ethics Committee of the Maulana Azad Medical College, affiliated with the University of Delhi. Prior informed consent for the surgical procedure regarding the details, benefits, and risks was obtained from the guardians of the children. In addition, assent was also taken from children in the age group of 13–18 years. The study was conducted in accordance with the tenets of the Helsinki Declaration.

The preoperative, intraoperative, and postoperative data of these patients were analyzed and the parameters that were recorded were age, unilateral/bilateral keratoconus, presence of VKC, uncorrected visual acuity (UCVA), best-corrected visual acuity (BCVA) with spectacles/contact lenses (pre- and postoperative), slit-lamp examination findings, automated keratometry (Shin Nippon Keratometer ACCUREF-K 9001; Vision Systems Inc, Tarpon Springs, Florida, USA), thinnest pachymetry on Orbscan IIz (Bausch and Lomb, Rochester, New York, USA), intraocular pressure (IOP) by Tono-Pen XL Applanation Tonometer (Reichert Technologies, Inc, Buffalo, New York, USA), intraoperative surgical technique, graft clarity, interface haze, postoperative complications and their management, duration of follow-up, and fellow eye management of the patients with unilateral operated DALK. The diagnosis of keratoconus was made based on conical protrusion, slit-lamp findings (stromal thinning, Vogt striae, Fleisher ring, and subepithelial scarring) and confirmed by elevation topography (Orbscan IIz; Bausch and Lomb) in all the cases.

The patients with the VKC were classified into mild, moderate, and severe, according to the clinical findings. Severe VKC included cases with giant papillae and moderate cases included moderate-sized conjunctival papillae (Bonini grading).¹² The patients with severe VKC at presentation were managed medically with unpreserved topical 2% cyclosporine A solution for 2 weeks to control ocular surface inflammation, prior to the surgery.

• OPERATIVE TECHNIQUE: All the surgeries were performed by the same surgeon (R.A.). Of these, 12 surgeries were performed under general anesthesia and 8 surgeries under local peribulbar anesthesia. After host corneal partial-thickness trephination 8.5/9.0 mm, superficial keratectomy, and a circumferential peripheral 360 degree lamellar dissection, a big bubble was attempted in all the patients with a bent 27 gauge needle, bevel down. If this step was unsuccessful, manual layer-by-layer dissection and stromal delamination with air was performed. In cases with successful single big bubble, it was slowly decompressed with a pointed tip of a V-lance and the air was replaced with 1% sodium hyaluronate. Deep stromal layers forming the roof of the bubble were removed by divide-andconquer technique in 4 quadrants. A large lamellar corneal lenticule (8.5/9.0 mm), obtained after removing the Descemet membrane, was harvested from McCarey and Kaufman preserved cornea. The graft-host disparity of 0-0.5 mm was maintained. The graft was sutured to the host peripheral cornea with 16–20 interrupted 10-0 nylon sutures. Donor cornea was tucked into the peripheral host corneal pocket.

• POSTOPERATIVE MANAGEMENT AND FOLLOW-UP: All patients were examined every day for 4 days, then every week for the next 4 weeks, every 2 weeks for the next 12 weeks, every 4 weeks for 6 months, and then every 3 months. The topical antibiotic ofloxacin 0.3% (Ofax 0.3%; Optho Remedies Pvt Ltd, Allahabad, India) was stopped after 1 month and the predacetate 1% (PredForte 1%; Allergan, Irvine, California, USA) drops were tapered from 6 times a day to 2 times a day gradually over 3 months. Thereafter, they were continued on loteprednol etabonate 0.5% (Sun Pharmaceuticals Industries Ltd, Mumbai, India) twice- or thrice-daily regimen. In cases of suture-related complications (premature loosening, suture infiltration, exposed knot, and vascularization), early suture removal was done. Otherwise, suture removal was not done before 6–9 months. Graft clarity, UCVA, BCVA, IOP, and automated keratometry were noted at every visit. The UCVA, BCVA, IOP, keratometry readings, and endothelial count (Topcon SP 3000p; Topcon Corp, Tokyo, Japan) were recorded at the last follow-up after suture removal. A note was also made of associated VKC and treated, in case of flare-up.

RESULTS

DETAILS OF PATIENTS' DEMOGRAPHICS AND PREOPERATIVE data are presented in Table 1. The postoperative data are presented in Table 2.

• PATIENT CHARACTERISTICS: The mean age of our patients was 14.4 years (11–18 years). Of the 16 patients, 5 were female and 11 were male. Five patients had advanced bilateral keratoconus. Four patients underwent DALK in both eyes while 1 patient underwent DALK in 1 eye and PK in the fellow eye (Case 13). Cases 11 and 12 were twin brothers. Of the cases, 3 eyes had severe VKC, 8 eyes had moderate VKC, and 9 eyes had mild VKC. The mean follow-up was 44.5 months (24–105 months). All the patients were in Krumeich's stage 3 or more of keratoconus at the time of surgery.¹³ There was no hydrops in any eye with advanced keratoconus undergoing DALK. The mean thinnest pachymetry was 334.2 μ m (293–385 μ m).

• VISUAL OUTCOMES: The mean preoperative decimal UCVA was 0.06 \pm 0.08 (Snellen equivalent [SE] 20/ 320). The mean postoperative decimal UCVA was 0.24 \pm 0.09 (SE 20/80). At the final follow-up, 16 out of 20 eyes (80%) achieved UCVA of \geq 0.25 (SE 20/80). The mean preoperative decimal BCVA was 0.12 \pm 0.19 (SE 20/160). The mean postoperative decimal BCVA was 0.45 \pm 0.24 (SE 20/40). The preoperative BCVA in the

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