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Original Article

Deep anterior lamellar keratoplasty for pellucid marginal degeneration

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Abstract

Purpose: To present the surgical outcomes of deep anterior lamellar keratoplasty (DALK) for pellucid marginal degeneration (PMD).
Methods: A retrospective review was performed in 16 eyes of 16 patients who underwent DALK at the King Khaled Eye Specialist Hospital, Riyadh, Saudi Arabia between January 1, 2006 and December 30, 2009. Baring of Descemet's membrane (DM) during DALK was achieved in 8 (50%) eyes; residual stroma was left intraoperatively in the remaining 8 (50%) eyes. The big bubble technique was performed in 10 (62.5%) eyes and manual dissection was performed in the remaining 6 (37.5%) eyes. Visual acuity (LogMAR notation), intraocular pressure, intraoperative complications and postoperative graft status were assessed.
Results: The mean follow up was 14.6 ± 8.2 months (range 6–35 months). The mean overall age was 31.4 ± 9.6 years (range, 19–50 years). Visual acuity increased statistically significantly from 0.9 ± 0.3 (range 0.5–1.6) preoperatively to 0.4 ± 0.2 (range 0.0–0.7) at last follow-up ($p < 0.0001$). There was a statistically significant improvement in postoperative sphere, cylinder, and spherical equivalent ($p < 0.035$, $p < 0.001$, and $p < 0.02$, respectively) compared to preoperative. Postoperative visual acuity was not statistically significantly related to gender, type of surgical technique, and baring or perforation of DM. The main graft-related complication was graft–host vascularization (2/16 eyes).
Conclusion: DALK reduces severe corneal astigmatism and results in good visual and refractive outcomes and is an effective alternative for patients with PMD.

Keywords: Lamellar keratoplasty, Pellucid marginal degeneration, Corneal ectasias

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Introduction

Pellucid marginal degeneration (PMD) is a progressive, noninflammatory peripheral corneal thinning disorder with onset between 20 and 40 years of age. PMD is characterized by a peripheral band of inferior corneal thinning with an adjacent 1–2 mm band of normal cornea to the limbus. The area of thinning typically is epithelialized, clear, avascular, and without lipid deposits.[1,2] Similar to keratoconus, PMD is a bilateral progressive disorder, although the disease can be asymmetric between eyes. Classic PMD occurs in the inferior cornea, however cases of superior PMD have been reported.[3] Clinically, PMD causes a flattening of the vertical meridian resulting in marked against-the-rule astigmatism. Typically, patients present with reduced visual acuity (VA) due to high irregular astigmatism. The etiology and prevalence of PMD remain unknown. Whether PMD, keratoconus, and keratoglobus are distinct diseases or phenotypic variations of the same disorder is unclear.[4]

Treatment of the early stage of PMD involves spectacles and contact lenses. As the disease progresses and patients

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cannot be adequately corrected with spectacles or become contact lens intolerant, surgical intervention is warranted.[5] Recently, deep anterior lamellar keratoplasty (DALK) has been reported as a viable alternative to penetrating keratoplasty for corneal ectasias.[6,7] In the current study, we present the surgical outcomes of DALK for PMD at a specialist center in Saudi Arabia.

Methods

Institutional Review Board approval was granted for this study and this study was conducted in accordance with the Declaration of Helsinki. A chart review was conducted for every patient who underwent DALK for PMD at the King Khaled Eye Specialist Hospital (KKESH) in Riyadh, Saudi Arabia between January 1, 2006 and December 30, 2009. Patients were included if the procedures were performed in eyes with a clinical diagnosis of PMD that were contact lens intolerant with no previous history of hydrops. Data were collected for age, sex, laterality, preoperative and postoperative visual acuity (logarithm of the minimum angle of resolution (LogMAR) notation) and refraction, preoperative and postoperative intraocular pressure (IOP), baring and perforation of Descemet's membrane during the procedure, the clinical course, including any episodes of rejection and/or complications.

DALK was performed by multiple surgeons according to surgeon's preference. Direct open dissection as described by Anwar[8] was performed in 6 eyes and the ''big bubble''[9] technique in 10 eyes.

Fresh full-thickness donor corneas preserved in Optisol were used for all procedures. The corneal donor button was stripped of the Descemet's membrane and endothelium. Trephine sizes ranged from 8.25 to 9.5 mm in diameter, and the donor button was either the same diameter as the recipient or 0.25 mm larger. As inferior corneal thinning was present in all eyes, trephination was decentered inferiorly. The graft was sutured to the recipient with interrupted or combined interrupted-continuous 10–0 nylon sutures in 11 eyes and 5 eyes respectively. All eyes received subconjunctival injections of an antibiotic (cephazolin or gentamicin) and corticosteroid (methylprednisolone). Postoperative drops regimen included topical prednisolone acetate 1.0% used for 4 months followed by fluorometholone which was tapered over 4 months, antibiotics and artificial tears. The eyes were examined on 1st day, 1st week, 3–5 weeks, and 2–4 months postoperatively.

Selective suture removal for the reduction of astigmatism was performed as early as 9 weeks postoperatively. Otherwise, sutures were left in place up to 1 year as long as if they were not excessively tight, loose, exposed or attracting blood vessels.

Data were collected, reviewed and stored using Microsoft Excel 2007 (Microsoft Corp. Redmond, Wa., USA). Statistical analysis was performed using SPSS version 19 (IBM Inc., Armonk, NY, USA) and Stats Direct 7.2 (Stats Direct Ltd., Cheshire, UK). Descriptive and inferential analyses were performed to describe different indices and detect statistical differences between preoperative and postoperative data. The Wilcoxon signed rank test was used to compare preoperative and postoperative means. The Mann Whitney U test was used to compare means across different categories and groups, a p value less than 0.05 was considered statistically significant.

Results

The study cohort was comprised of 12 males and 4 females (16 eyes). The mean age of the cohort was 31.4 ± 9.6 years (range, 19 years to 50 years). The mean follow up was 14.6 ± 8.2 months (range, 6 months to 35 months). Baring of DM during DALK was achieved in 8 (50%) eyes; residual stroma that remained during surgery in the remaining 8 (50%) eyes with no interface opacity was noted. Perforation in DM occurred in 2 (12.5%) eyes, both of them had the big bubble technique. There was a statistically significant improvement in mean visual acuity from 0.9 ± 0.3 LogMAR (range, 0.5–1.6 LogMAR) preoperatively to 0.4 ± 0.2 (range, 0.0–0.7 LogMAR) at last visit postoperatively ($p < 0.0001$). The improvement in visual acuity was due to statistically significant improvements in sphere, cylinder, and spherical equivalent ($p < 0.035$, $p < 0.001$, and $p < 0.002$, respectively) (Table 1). The majority of astigmatism changes was following the DALK and prior suture removal ($p < 0.006$) compared to changes between prior suture removal and last visit ($p = 0.688$).

There was a statistically significant increase in IOP from 13.8 ± 1.9 mm Hg (range, 10–17 mm Hg) preoperatively to 15.8 ± 1.6 mm Hg (range, 13–19 mm Hg) postoperatively ($p < 0.0001$). Improvement in visual acuity was not statistically significantly related to gender, baring of DM, type of surgical technique, and DM perforation ($p > 0.05$, all cases) (Table 2).

The cohort was subdivided into 2 groups based on the surgical technique and the analysis was repeated. This analysis indicated that astigmatism correction was statistically significant with the big bubble technique ($p = 0.008$) compared to manual dissection ($p = 0.056$). However, crossing the two groups using the Mann Whitney test showed no statistical difference between groups ($p = 0.40$).

Intraoperative perforation of DM during DALK occurred in 2 (12%) eyes. However, this perforation had no statistical impact on final visual acuity ($p = 0.47$). These eyes were managed by injection of air or a mixture of perfluoropropane (C_3F_8) with air (14% C_3F_8, 86% air) into the anterior chamber to temporarily seal the microperforations. There was no formation of ''secondary anterior chamber'' postoperatively. Graft–host vascularization occurred in 2 (12%) eyes. In one eye, photodynamic therapy using verteporfin was used to treat the corneal neovascularization with complete regression. All (100%) eyes retained a clear graft at last follow-up.

Discussion

Several surgical procedures have been performed for visual rehabilitation of eyes with PMD. These include crescentic wedge resection,[10] crescentic lamellar keratoplasty,[11] large diameter penetrating keratoplasty,[12] and a modified procedure in which an inferior crescentic lamellar keratoplasty is combined with a central penetrating keratoplasty.[13] All these techniques have several disadvantages, including unpredictability, irreversibility, a long period of rehabilitation, and significant complication rates. Recently, one or two segments of intracorneal ring implants (ICR) have been inserted to correct

Table 1. Comparison of preoperative and postoperative visual acuity, intraocular pressure, sphere, cylinder, axis and spherical equivalent.

Variable	Preoperative		Postoperative		Statistical significance (p value)
	Mean (± SD)	Range (min–max)	Mean (± SD)	Range (min–max)	
LogMAR visual acuity	0.9 (0.3)	(0.5–1.6)	0.4 (0.2)	(0–0.7)	<0.0001
Intraocular pressure (IOP)	13.8 (1.9)	(10–17)	15.8 (1.6)	(13–19)	<0.0001
Sphere	−3.2 (4.6)	(−15 to 3)	−0.3 (2.2)	(−5 to 3)	0.035
Cylinder	−8 (2.1)	(−11 to −2.5)	−4.3 (1.9)	(−8 to −1.5)	0.001
Axis	92.2 (8)	(80–105)	90 (41.9)	(40–180)	0.589
Spherical equivalent (SE)	−7.2 (4.1)	(−16.3 to −1)	−2.4 (2.2)	(−7 to 1.3)	0.002

$p < 0.05$ was considered statistically significant.

Table 2. Risk factors for visual acuity outcome after deep anterior lamellar keratoplasty.

Variable	p Value
Gender	0.583
Baring of Descemet's membrane	0.245
Type of surgical technique	0.703
Perforation of Descemet's membrane	0.472

$p < 0.05$ was considered statistically significant.

astigmatism associated with early PMD. Although, the result is promising, long term stability is questionable.[14,15]

To the best of our knowledge, a case of unilateral corneal perforation associated with PMD and the fellow eye with PMD were successfully managed with DALK.[16] Deep anterior lamellar keratoplasty (DALK) has a number of advantages over penetrating keratoplasty (PKP). For example, DALK is an extraocular procedure hence which diminishes or eliminates the chances of postoperative glaucoma, cataract formation, retinal detachment, cystoid macular edema, expulsive choroidal hemorrhage and epithelial ingrowth.[17–20] As a result of the preservation of host endothelial cells, endothelial graft rejection following DALK has not been reported in previous studies.[17–20] This is consistent with the current study which found no endothelial graft rejection. This observation is in contrast to PKP where endothelial graft rejection occurs in 20–30% of patients.[21,22]

A major advantage of DALK is the low rate of chronic endothelial cell loss compared to PKP. This endothelial cell loss is 11% within the first 6 postoperative months and then approaches a more physiologic rate of cell loss of 1–2%.[23] However, endothelial cell loss after PKP is 4.2% a year even 5–10 years after surgery.[21,22] A further advantage of DALK is the reduced need for topical steroids which may cause glaucoma, cataract and predispose to infection.

In the current study, we found a statistically significant reduction in severe corneal astigmatism after DALK ($p < 0.05$). The reduction in corneal astigmatism likely caused the statistically significant improvement in visual and refractive outcomes ($p < 0.05$, both cases). To date, there has been no recurrence of corneal thinning in the entire cohort. A drawback of this study is the small sample size. Despite the small sample size statistically significant differences were evident in this study. Furthermore, it seems that the ''big bubble'' technique enables better correction of astigmatism than manual dissection. Perhaps the retention of some portion of deep stromal corneal fibers can contribute to postoperative astigmatism.

There was a statistically significant increase in intraocular pressure postoperatively ($p < 0.000$). However this was clinically mild and within normal range. Most likely, this elevation was due to the change in the corneal thickness following DALK rather than a real elevation in the intraocular pressure.

DALK can be complicated by DM perforation which occurred in 2 (12%) eyes in this series. The rate of DM perforation in the current study compares well with previous reports of DALK for keratoconus, which range between 9%[9] and 15%.[18] However, we found that DM perforations did not have an impact on the final visual acuity. Injection of air, or a theoretically non-expandable mixture of air and C_3F_8, into the anterior chamber to allow completion of the dissection and to prevent a second chamber postoperatively, can be used to manage most small perforations. If these measures fail or if the perforation is large, conversion to PKP may be required.

In conclusion, DALK provides a safe and successful alternative to PKP for PMD but remains a challenging procedure. It reduces severe corneal astigmatism and results in good visual and refractive outcomes. Prospective studies with larger sample sizes are required to provide long-term analysis of DALK for PMD.

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