

Deep anterior lamellar keratoplasty outcomes in macular and granular corneal dystrophy – A comparative cross-sectional study

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Purpose: To compare the functional and anatomical outcomes (in terms of graft uptake and rejection/failure) of deep anterior lamellar keratoplasty (DALK) in stromal corneal dystrophy (macular and granular). **Methods:** Sixteen eyes with macular corneal dystrophy (MCD; group A) and 10 eyes with granular corneal dystrophy (GCD; group B) underwent successful DALK by big bubble technique or layer-by-layer dissection. **Results:** Both groups showed significant improvement in their best-corrected visual acuity postoperatively (postoperative P value in MCD and GCD was 0.00001 and 0.0008, respectively) with no statistically significant differences between the two groups ($P = 0.77$) at 1 year. Postoperative endothelial count did not drop significantly in group A (MCD, $P = 0.1553$). Only in seven eyes preoperative endothelial count could be obtained (due to dense stromal corneal opacity), but there was a significant endothelial count difference between preoperative and postoperative count in group B (GCD, $P = 0.0405$) at the end of 1 year postoperatively which could be because of age and stage of disease (advanced granular dystrophy) and also because of small sample size of GCD compared with MCD. Intergroup comparison between the two groups showed no statistically significant difference ($P = 0.6353$) with good postoperative outcome in both groups. **Conclusion:** DALK can be successfully done in both groups and results are comparable. However, long-term outcomes on a large scale need to be further evaluated.

Key words: Deep anterior lamellar keratoplasty, granular corneal dystrophy, macular corneal dystrophy, stromal corneal dystrophy

Stromal corneal dystrophies cause significant visual morbidity in patients due to stromal opacity.^[1] Macular corneal dystrophy (MCD) is a progressive autosomal recessive disease which presents commonly in the second decade of life and the severity increases with age.^[2] Granular corneal dystrophy (GCD) also presents with stromal opacity and results in decrease in vision over time. It is inherited in an autosomal dominant fashion and can cause significant visual morbidity at third decade of life.^[3] Multiple surgical treatment modalities such as penetrating keratoplasty (PK) and deep anterior lamellar keratoplasty (DALK) have been advocated for treating corneal stromal dystrophies.^[4] Final visual rehabilitation is dependent on surgical procedure and the stage of disease.

Multiple studies have shown good outcomes in terms of visual acuity and graft clarity with both PK and DALK, and DALK can be attempted and successfully done if Descemet's membrane (DM) and endothelium are spared.^[5,6] However, risk of endothelial graft rejection in PK and recurrence of the disease, in addition to other complications, still remains one of the most feared problems while choosing the surgical procedure.^[1] Previous studies have individually shown good outcomes in patients undergoing DALK for both macular and GCD separately. But no previous studies have shown any comparative data on DALK in macular versus granular dystrophy. To the best of our knowledge, this

is the first study that has attempted comparing vision and endothelial count in a fairly large number of MCD and GCD.

Thus, we aim to show both functional and anatomical outcomes of DALK in macular and granular dystrophy independently and compare the outcome of DALK between the two conditions.

Methods

The study involved DALK in 16 eyes with MCD (group A) and 10 eyes with GCD (group B) and was conducted in a tertiary care eye hospital, by a single experienced senior surgeon, as a retrospective cross-sectional observational study from January 2009 to December 2016. Informed written consent was taken from all patients and the study protocol adhered to the tenets of the Declaration of Helsinki and was approved by the Institutional Review Board.

Visual acuity [uncorrected visual acuity and best-corrected visual acuity (BCVA)], slit-lamp examination, dilated fundus examination, refraction, and endothelial count (using Tomey EM-3000) were measured for all patients pre- and

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postoperatively. Inclusion criteria included all patients with clinically appearing stromal dystrophy (macular and granular only) and those having corneal involvement upto stromal involvement which was confirmed by anterior segment optical coherence tomography (AS-OCT) (Optovue RTVue XR100-2) and with progressive worsening of vision were included in the study. AS-OCT in all patients were obtained and demonstrated involvement only upto posterior stromal layer. It is an important deciding factor in choosing surgery (DALK vs PK) as very deep involvement and endothelial involvement can result in recurrence. Patients with no previous history of intraocular surgery were included. In few patients with macular dystrophy, preoperative endothelial count was not obtainable due to dense stromal opacity.

Surgical technique

DALK was performed in all patients by Anwar's big bubble technique or layer-by-layer dissection.^[7] Host bed trephine size was decided based on the area of corneal involvement (7.75–8.25 mm; mean size of 8 mm) and donor graft was taken 0.25 mm more than the host size. All grafts were sutured using 12–16 interrupted nylon sutures depending on graft size (12 sutures for graft size <8 mm and 16 sutures for size >8 mm). One patient in granular dystrophy and two patients in the macular dystrophy group developed microperforation during separation of posterior stroma from the DM and were successfully managed by injecting air bubble in anterior chamber. Postoperatively, all patients were given topical antibiotics (moxifloxacin 0.5%) for 1 week and steroids (prednisolone acetate 1%) in tapered dose over 4–6 months. All patients were followed up for a minimum of 1 year postoperatively. The follow-up schedule is as follows:

(a) Day 1 postoperative, (b) first postoperative week, (c) at sixth week postoperative, (d) third month postoperative, (e) sixth month postoperative, and (f) first year postoperative and every fourth to sixth month postoperative thereafter. All sutures were removed beginning at sixth month and completely by the first year. Patients were on regular follow-up and no patients developed graft rejection or recurrence during the follow-up.

Results

The mean age (\pm standard deviation) of macular and granular dystrophy was 27.9 \pm 5.19 years (range 21–39 years) and 36.63 \pm 7.88 years (22–47 years), respectively. Both groups A and B (MCD and GCD) showed significant improvement in their BCVA at the end of 1 year (postoperative *P* value in MCD and GCD was 0.00001 and 0.0008, respectively) as shown in Tables 1 and 2. There was no statistically significant difference between postoperative mean BCVA when compared between the two groups [*P* = 0.77208, as shown in Table 3] at the end of 1 year.

There was a significant endothelial count difference between preoperative and postoperative count in group B (GCD, *P* = 0.0405) as shown in Tables 1 and 2. Preoperative endothelial count was not measurable in nine patients with macular dystrophy. When intergroup comparison of visual status was done between the two groups [group A vs B, as shown in Table 3], there was no statistically significant difference (*P* = 0.6353). Figs. 1-6 shows preoperative and post operative images of MCD and GCD patients. Fig. 7 shows AS-OCT image of a patient with advanced GCD.

Table 1: Pre- and postoperative BCVA and endothelial count of group A (macular corneal dystrophy)

	Preop	Postop	<i>P</i>
BCVA (LogMAR)	0.83 \pm 0.25	0.29 \pm 0.3	0.00001
EC (<i>n</i> =7) mm/cm ²	2330.71 \pm 255.33	2227.67 \pm 230.22	0.1553

BCVA=Best corrected visual acuity, EC=Endothelial count

Table 2: Pre- and postoperative BCVA and endothelial count of group B (granular corneal dystrophy)

	Preop	Postop	<i>P</i>
BCVA (LogMAR)	0.60 \pm 0.21	0.28 \pm 0.15	0.0008
EC (<i>n</i> =10) mm/cm ²	2476.80 \pm 203.72	2273.20 \pm 208.83	0.0405

BCVA=Best corrected visual acuity

Table 3: Postoperative BCVA and endothelial count intergroup comparison (group A vs group B, i.e., macular vs granular dystrophy)

	Group A	Group B	<i>P</i>
Postop BCVA (LogMAR)	0.29 \pm 0.3	0.28 \pm 0.15	0.7720
Postop EC mm/cm ²	2227.67 \pm 230.22	2273.20 \pm 208.83	0.6353

BCVA=Best corrected visual acuity

Discussion

DALK remains one of the preferred modality of treatment in cases where the corneal disease is limited to the anterior layers of cornea and has not involved the endothelial layer.^[8] Few studies have proven the superiority of DALK over PK even in stromal dystrophies due to lower risk of rejection, faster healing, and lesser complications like cataract and glaucoma,^[3,9-14] and hence it was preferred over PK in these cases. Macular corneal dystrophy, however, has slightly poorer prognosis as recurrence is possible even after successful surgery and has been described both in DALK and PK, and thus it makes more sense to prefer DALK in these patients.^[15-17] This study attempted to compare the outcomes of DALK in both the groups at 1-year follow-up which showed good visual outcomes in both groups, but recurrence rate has to be assessed in long-term follow-up.

Reddy *et al.* showed that there was no significant difference in outcome for patients with MCD between the DALK and PK groups in terms of postoperative visual and refractive outcomes, and in addition, DALK was superior to PK in its postoperative safety profile.^[18] The postoperative visual outcome at 1-year post DALK in MCD as shown by Cheng *et al.* is similar to our study.^[19] They also showed the advantage of DALK over outcomes of PK. Similar outcome was proven by Sogutlu Sari *et al.*^[2] Our study in addition included a large number of patients (16 eyes) with advanced macular dystrophy with good postoperative outcome.

This is perhaps the first study which compared the MCD and GCD and evaluated their outcome after DALK. Salouti *et al.* showed that granular dystrophy can recur post DALK, but complications associated with PK can be avoided if DALK compared with PK is chosen for visual rehabilitation.^[3] Postoperative visual outcomes for stromal dystrophy as shown by Unal *et al.* were very similar to our study.^[1] MCD progresses with age and requires surgical intervention at advanced stage. Our study did not have patients above sixth decade, and thus

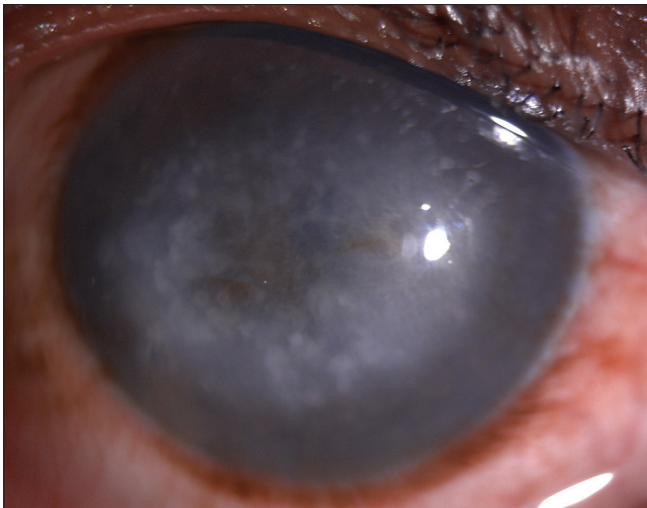


Figure 1: Preoperative picture of a patient with macular corneal dystrophy (MCD)

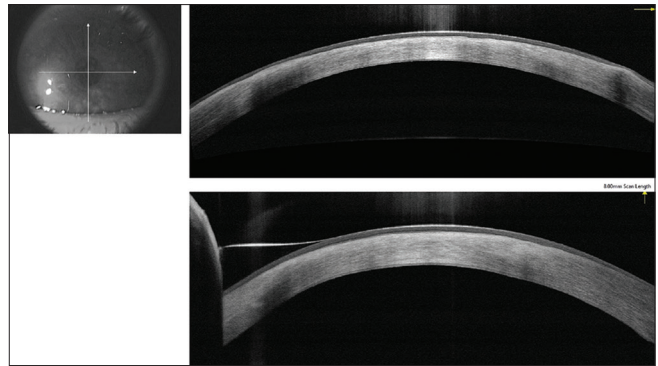


Figure 2: Anterior segment optical coherence tomography (OCT) of the same patient with MCD

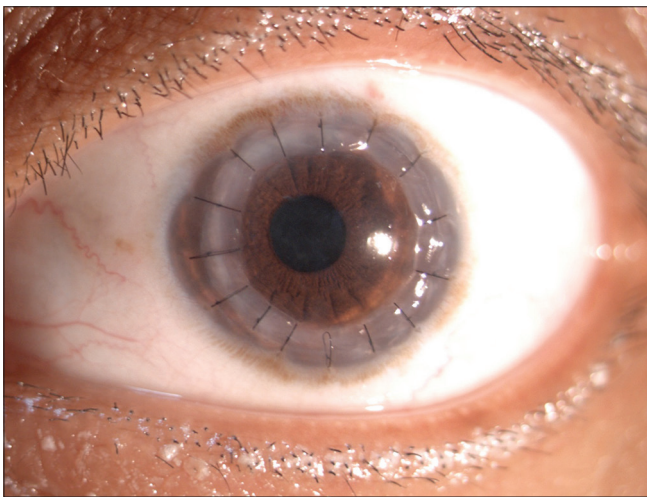


Figure 3: Postoperative picture of the same patient with MCD after DALK



Figure 4: Picture of a patient with granular corneal dystrophy (GCD)

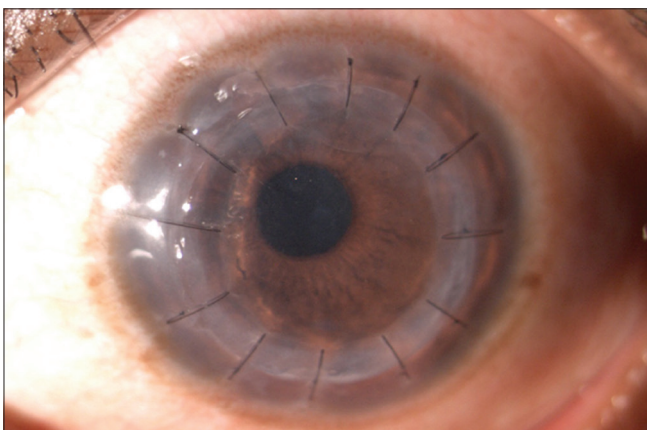


Figure 5: Postoperative picture of the same patient with granular corneal dystrophy (GCD)

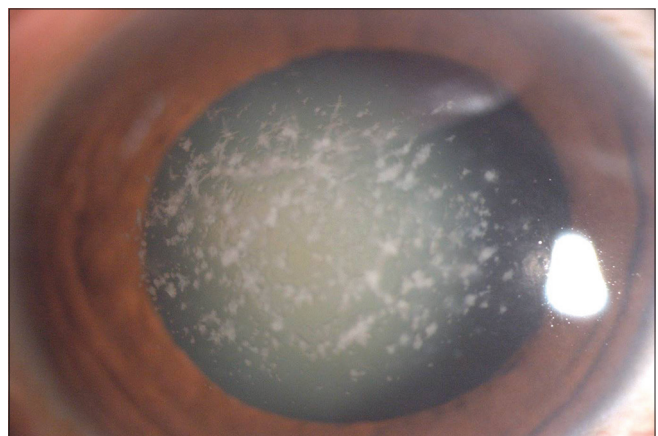


Figure 6: A patient with advanced granular dystrophy

results could be more favorable. Hence, a larger sample size and more variable age group analysis could be done in future.

Big bubble technique is useful for DALK in stromal dystrophies and is one of the good predictive factors in terms of outcome as shown by Vajpayee *et al.*^[12] Patel *et al.*^[20] showed that big bubble technique of DALK can be successfully carried

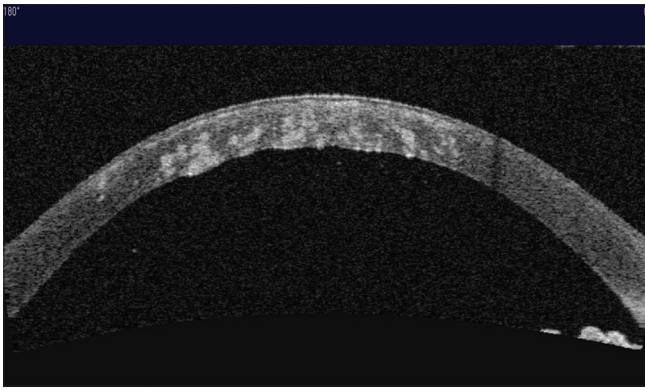


Figure 7: As-oct image of a patient with advanced granular dystrophy

out for stromal dystrophy. Careful and complete removal of the posterior stromal layer is important for successful visual outcome in these cases. Some believe that DALK is not a treatment of choice for MCD as it can involve the deeper layers of the cornea and possibly the DM^[2] and can lead to DM thinning and increased chances of intraoperative DM perforation and postoperative endothelial decompensation.^[14,21] This result could be due to certain factors and limitations such as (a) advanced age in GCD compared with MCD group, (b) small sample size of GCD compared with MCD group, (c) advanced stage of dystrophy and dense stromal opacity where preoperative accurate endothelial count could not be determined, and (d) retrospective study design. Cheng *et al.* showed that though PK may result in immediate improvement, it has associated complications, whereas DALK provides a better stability and safety profile and there was no significant difference in graft survival between the eyes treated with PK and DALK for MCD.^[19] We found that posterior scarring was more in MCD group in few cases (two eyes) compared with GCD which could be due to the advanced and deeper involvement seen in MCD. However, there was no statistically significant difference in outcomes between the two after DALK.

Conclusion

DALK is a successful surgical preference for both MCD and GCD as shown in the study; however, long-term outcomes in terms of recurrence are yet to be assessed. Though this study has shown successful outcomes even in patients with advanced MCD and GCD, future studies aiming to compare outcomes in different stages of disease can help in better decision-making with regard to the choice of surgery. This study also shows that outcome of DALK for both MCD and GCD is good and comparative.

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Conflicts of interest

There are no conflicts of interest.

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