

PreDescemet's deep anterior lamellar  
keratoplasty as a treatment modality  
in severe granular corneal dystrophy  
type 2: a study with a long-term  
follow-up period

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Directed by Professor Eung Kweon Kim

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## ABSTRACT

### **Predescemetetic deep anterior lamellar keratoplasty as a treatment modality in severe granular corneal dystrophy type 2 : a study with a long-term follow-up period**

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This study was designed to assess the long term surgical outcomes of predescemetetic deep anterior lamellar keratoplasty (preDALK) in Korean patients with severe granular corneal dystrophy type 2 (GCD2).

Retrospective review of the medical charts of patients with GCD2 who had undergone preDALK was performed. DNA analysis was done to confirm the transforming growth factor- $\beta$  induced gene (*TGFBI*) R124H mutation. After preDALK was performed, visual acuity, refractive error, keratometry values, complications, and the presence of rejection and recurrence were evaluated.

Ten eyes of nine patients were followed up for more than 30 months, and the mean follow-up period was  $48.9 \pm 14.6$  months. Two eyes showed homozygous R124H mutations and the other eyes had heterozygous R124H mutation. The mean best corrected visual acuity after surgery was  $0.14 \pm 0.11$  (logMAR). The two eyes with homozygous mutations showed recurrence. However, all the eyes with heterozygous mutations showed no recurrence during the follow-up period ( $45.1 \pm 13.9$  months).

The outcome of preDALK which was conducted on the GCD2 patients seems to have at least an equivalent recurrence-free period in comparison to that of penetrating keratoplasty (PKP) and deep anterior lamellar keratoplasty (DALK). Furthermore, preDALK was predicted to provide a longer-term graft survival than that of PKP. Reoperation using the preDALK procedure may cause fewer complications than the DALK procedure. Therefore, preDALK should be considered as a primary surgical option for GCD2 patients with severe visual impairment who needs to undergo PKP and DALK.

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Key words: Predescemetic deep anterior lamellar keratoplasty, Granular corneal dystrophy type 2

**Predescemetometric deep anterior lamellar keratoplasty as a treatment modality in severe granular corneal dystrophy type 2 : a study with a long-term follow-up period**

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## **I. INTRODUCTION**

Granular corneal dystrophy type 2 (GCD2, Avellino corneal dystrophy) is a hereditary disorder that is aggravated with age and which sometimes threatens vision in heterozygous patients.<sup>1</sup> Hyaline granular deposits located in the anterior stroma are usually present at an early stage and are followed by lattice deposits located in the deep stroma. Diffuse stromal haze usually appears after the age of 50 years, is located at the sub-Bowman's layer, and often threatens central vision, requiring removal with phototherapeutic keratectomy (PTK).<sup>1-4</sup> In cases of patients with poor vision due to deeply located deposits such as lamellar deposits and interfacial deposits after LASIK with thick flap, superficial keratectomy is not sufficient to remove the opacities and keratoplasty may be required.

GDC2 homozygous patients have severe corneal deposits and decreased visual function from early childhood.<sup>5-8</sup> PTK can be attempted at this early age, but the recurrence appears more frequently and corneal transplantation is required much earlier than in heterozygous patients.

Deep anterior lamellar keratoplasty (DALK) is a surgical procedure that removes the corneal epithelium and stroma to the Descemet's membrane (DM), followed by transplantation of a donor graft while preserving the healthy DM and endothelium layers of the recipient. The procedure is widely used in patients with anterior corneal abnormalities nowadays because of longer graft survival, less endothelial rejection and lower postoperative complication rates than in patients with penetrating keratoplasty (PKP).<sup>9-13</sup> Predescemetic DALK (preDALK) is a similar surgical procedure to DALK, the difference between the two procedures is that preDALK leaves a very thin layer of the deepest stroma without complete exposure of the DM. Patients treated with DALK usually have better vision than patients treated with preDALK, due to less interfacial haziness and scarring. However, the DM baring technique is known to be more difficult and has more possibility of microperforation during the surgery compared to non-DM baring techniques.<sup>14-16</sup>

In this study, we report the surgical outcomes of 10 cases of patients with GCD2 who have undergone preDALK and whose follow-up period exceeded 30 months.

## II. MATERIALS AND METHODS

### 1. Patient selection and analysis

This study was performed in accordance with the tenets of the Declaration of Helsinki and was approved by the Severance Hospital Institutional Review Board (No. 4-2012-0407). Medical records of patients who underwent preDALK for GCD2 from January 2006 to December 2011 at the Severance Hospital, Yonsei University College of Medicine were retrospectively reviewed. In each case, DNA analysis was performed on peripheral blood at the patient's first visits to confirm the Arg124His mutation in the transforming growth factor  $\beta$ -induced gene (*TGFBI*, formerly known as *BIGH3*) which is located on the long arm of chromosome 5.<sup>17-19</sup>

The clinical data of all enrolled patients were collected to include complete ophthalmic investigations, visual acuity, intraocular pressure, past history, previous ophthalmologic operation history, operation date, postoperative complications, presence of GCD2 recurrence, graft rejection, postoperative refractive error, best corrected visual acuity and keratometric values. The recurrence of GCD2 was defined when deposits were detected on the corneal central area which was approximately 6.0 mm in diameter and was not on the peripheral host-graft margin or at the suture related puncture site, as evaluated with slit lamp examination. Fourier domain optical coherence tomography (FD-OCT; RTVue-100: Optovue Inc., Fremont, CA) was used to evaluate preoperative and postoperative corneal morphology and depth of the deposits in some cases. The patients with ophthalmic diseases such as significant corneal thinning, edema, or uveitis that could affect the postoperative results, and whose follow-up period did not exceed 30 months, were excluded.

## **2. Predescemetic deep anterior lamellar keratoplasty**

All surgeries were performed by one surgeon (E. K. Kim) from January 2006 to December 2011 at the Severance Hospital (Seoul). Complete ophthalmic examinations were performed before and after the operations. General anesthesia was administered in all operations. Most surgeries were performed by using a layer-by-layer dissection technique with some modifications. In brief, after determining the size and measuring the depth of the recipient cornea with ultrasonic pachymetry and FD-OCT, the operator performed an approximately 70-80% partial thickness corneal trephination by using a Hanna trephine. Partial dissection of the anterior stroma was completed with a curved corneal scissors. The remaining stromal bed was carefully dissected, layer by layer with a crescent knife, a blunt spatula, and Vanna scissors until the residual stromal bed was not more than 5-10% of the preoperative full corneal thickness. The same sized trephine was used for donor preparations. The donor cornea was sutured with 10-0 nylon after stripping of the DM and the endothelium of the donor cornea.

Topical levofloxacin eye drops (Cravit; Santen Pharmaceuticals, Osaka, Japan) and topical prednisolone acetate 1% (Pred Forte; Allergan, Westport, Co. Mayo, Ireland) were administered to each patient, approximately six times per day. Corneal sutures were removed between 7 and 15 months after surgery.

### III. RESULTS

In all, 14 eyes of 12 patients received the preDALK procedure and 10 eyes of nine patients met the inclusion criteria. Two eyes were from two patients with homozygous mutations, and eight eyes came from the seven patients with heterozygous mutations. The characteristics of each patient and details of the surgeries are summarized in Table 1. The nine patients included five women and four men with a mean age of 40.9 years at the time of operation. The follow-up period was  $48.9 \pm 14.6$  (standard deviation) months and the mean graft size of donor and recipient corneas was  $8.35 \pm 0.16$  mm. The mean of the residual recipients' bed measured at the corneal center with FD-OCT, was  $36.8 \pm 9.7$   $\mu\text{m}$ . The mean of preoperative uncorrected visual acuity (UCVA) was  $0.43 \pm 0.24$  (logMAR) and the mean of postoperative best corrected visual acuity (BCVA) was  $0.14 \pm 0.11$  (logMAR). Six eyes had received LASIK (laser in situ keratomileusis) before undergoing preDALK and two eyes had no past ophthalmic operation history. The two eyes which were from the patients with homozygous mutations had previously received lamellar keratoplasty and deep lamellar keratoplasty before undergoing preDALK. Five eyes experienced one or multiple graft rejections however all of the rejections were well treated with proper management, and none of the cases had graft failure or another keratoplasty during the follow up period. The recurrence of corneal opacity related to GCD2 was only observed in homozygous GCD2 patients.

**Table 1. Clinical Characteristics and Summaries of Patients**

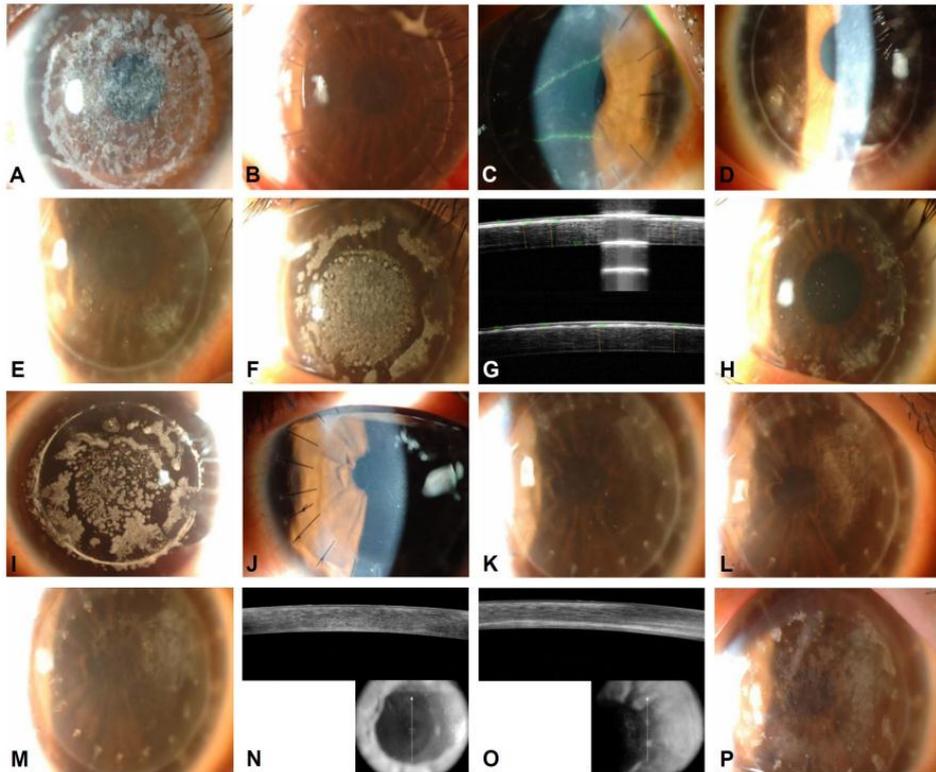
No.	Gene type	Gender /Age#	Ophthalmologic operation history	Operation date /Follow up period(month)	Preoperative UCVA / BCVA (logMAR)	Postoperative BCVA(logMAR) /POM	Postoperative SE / Cyl	Postoperative K - value mean(dioptor) /POM	Intraoperative complications	Recurrence /POM	Rejection /POM	Residual bed (µm)
1*	Hetero	Male / 50	LASIK 03'	2007.1.18 / 68	NA / 0.52	0.30 / 56	-12.125 / -4.25	43.995 / 30	None	None	None	27.0
2*	Hetero	Male / 50	LASIK 03'	2007.4.11 / 65	NA / 0.52	0.30 / 53	-13.25 / -3.00	45.5 / 27	None	None	None	36.7
3	Homo	Female / 24	LKP 90'	2006.9.20 / 65	0.70 / 0.70	0.22 / 16	-0.25 / -6.00	43.375 / 63	None	Yes / NA	Epi-Rej. / 3 Str-Rej. / 9	53.0
4	Homo	Female / 14	DLKP 01'	2007.3.9 / 63	0.70 / 0.22	0.15 / 48	+4.25 / +2.50	45.25 / 48	None	Yes / 24	Str-Rej. / 12, 13	43.0
5	Hetero	Male / 53	None	2008.11.17 / 34	NA / 0.22	0.10 / 34	+1.00 / -3.00	45.935 / 12	None	None	Str-Rej. / NA	44.0
6	Hetero	Female / 32	Blepharoplasty 02' LASIK 03' 2 times	2008.1.30 / 30	0.70 / 0.05	0.00 / 11	-9.50 / -1.00	49.00 / 18	None	None	Str-Rej. / 4, 8	NA
7	Hetero	Male / 59	None	2008.7.21 / 43	0.30 / NA	0.05 / 24	+1.00 / -3.00	43.81 / 24	None	None	None	24.0
8	Hetero	Male / 44	LASIK 99'	2008.10.27 / 42	0.70 / 0.52	0.00 / 36	-5.50 / -3.00	42.935 / 36	Micro perforation	None	None	30.3
9	Hetero	Female / 46	LASIK 02'	2008.8.11 / 40	1 / 0.70	0.10 / 34	+1.19 / -2.62	42.81 / 34	Micro perforation	None	None	36.0
10	Hetero	Female / 37	LASIK 00', 02', 04'	2009.3.6 / 39	1 / NA	0.15 / 30	-4.50 / -2.50	45.87 / 30	None	None	Str-Rej. / 35	NA

NA, not available; SE, spherical equivalent; UCVA, uncorrected visual acuity; BCVA best corrected visual acuity; POM, postoperative month; K-value, keratometric value; Epi-Rej., epithelial rejection; Str-Rej., stromal rejection; LASIK, laser in situ keratomileusis; LKP, lamellar keratoplasty; DLKP, deep lamellar keratoplasty

\*Case 1 and 2 is each eye of one patient. # Age at which preDALK was performed.

## **1. Homozygous GCD2 patients (Cases 3 and 4)**

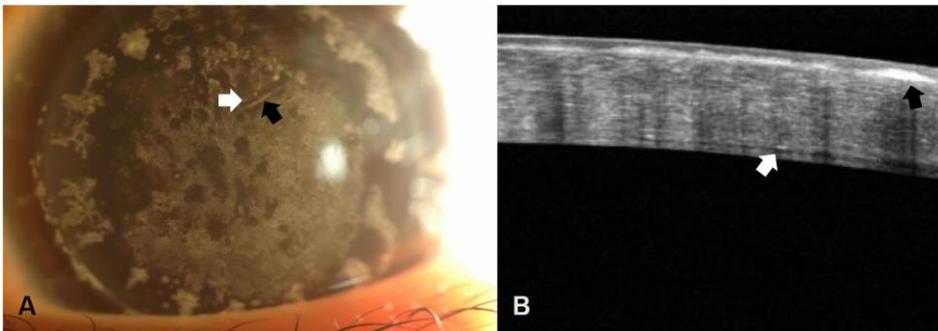
Two eyes of two patients with homozygous mutations were enrolled in this study. The case 3 patient was followed up regularly until postoperative month (POM) 16, and showed no recurrence. However, the patient did not visit the clinic after POM 16 and returned with severe recurrence at POM 62, as shown in Figure 1E and 1F. Gray-white, poorly demarcated granular deposits were observed in the sub-Bowman layer, as examined with FD-OCT (Figure 1G). The patient received PTK on POM 63 (Figure 1H). The case 4 patient visited our clinic regularly and the early signs of GCD2 recurrence were noted at POM 24 (Figure 1M). Both patients experienced two episodes of graft rejection which were well controlled with proper management, though some stromal opacity remained in the deep stroma (Figure 1C, D, E, K, and L).



**Figure 1.** Slit-lamp photographs of GCD2 homozygous eyes. A-H are from case 3, and I-P are from case 4. A, Preoperative photograph of case 3 with thin diffuse opacities at the central cornea. B, One day after preDALK. C, Epithelial rejection was noted approximately 3 months after preDALK. D-E, Stromal rejection appeared and regressed with some stromal opacity. F, The gray-white, poorly demarcated granular deposits were observed at 62 months after preDALK. G, FD-OCT showed that corneal deposits were located in the sub-Bowman layer. H, Photographs after receiving PTK showed almost no corneal deposits at the central area. I, Well-demarcated, discrete granular pattern of corneal opacities was observed in a preoperative photograph of case 4. J, One day after preDALK. K-L, Stromal opacities with ghost vessels caused by

stromal rejection. M, Recurrence of GCD2 was detected as thin, diffuse deposits at the corneal center at 24 months after preDALK. N, FD-OCT examination on the corneal center at 24 months after preDALK showed subtle whitish deposits at the sub-Bowman layer. O, FD-OCT examination of the stromal rejection site showed opacities which were located in the deep stroma. P, The progression of recurrence at 40 months after preDALK.

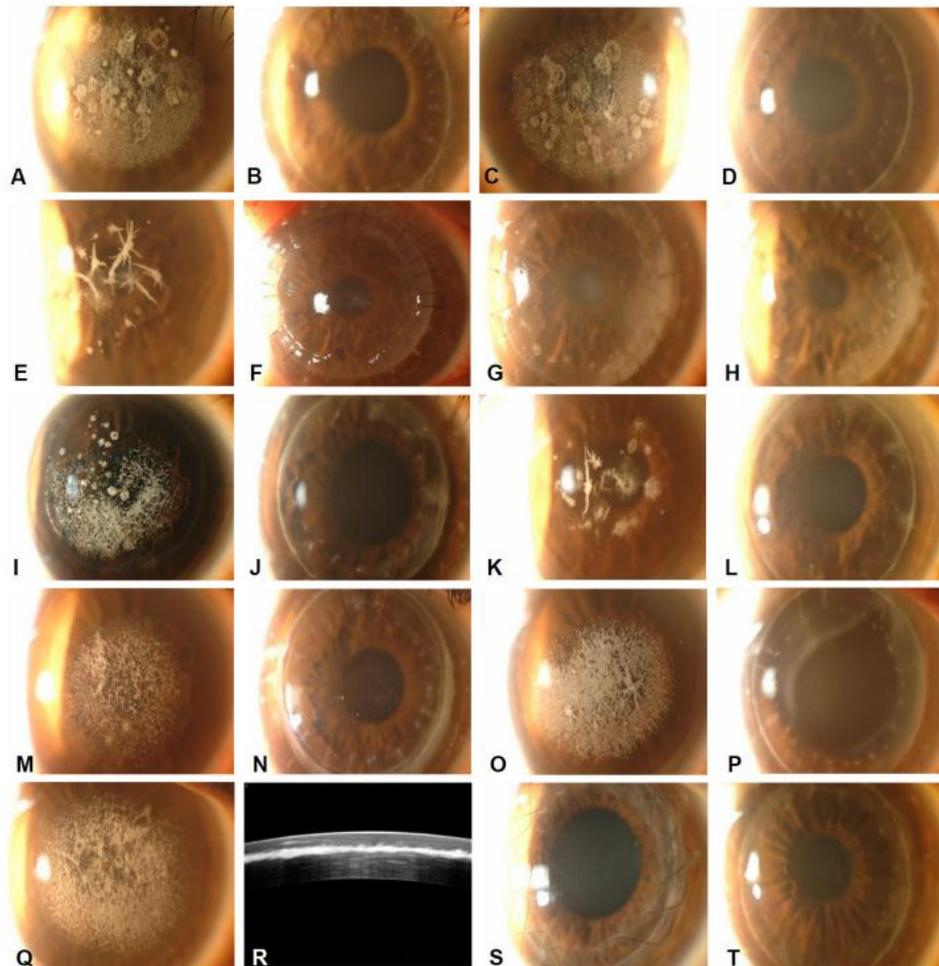
The case 4 patient had tiny dot-like opacities within the interfacial space on POM 63, and the opacities were also observed on FD-OCT examination (Figure 2 A, B).



**Figure 2.** Slit-lamp photograph and FD-OCT of case 4 at POM 63. A, Small numbers of tiny dot-like lesions were observed in the stromal interface. B, FD-OCT examination confirmed the location of tiny dot-like lesions which were located in the stromal interface. White arrow, tiny dot-like lesion; Black arrow, linear deposits in the sub-Bowman layer.

## **2. Heterozygous GCD2 patients**

Eight eyes of seven patients were recruited to this study. Six eyes had received one or more LASIK surgeries before undergoing preDALK. Two eyes of patients who did not receive LASIK surgery had lattice deposits at the deep stromal layer of the corneal center. Three eyes experienced episodes of one or more stromal graft rejections and stromal opacities similar to those observed in the homozygous patients described above (Figure 3 G and H). The mean and longest follow-up periods were  $45.1 \pm 13.9$  months and 68 months, respectively, and slit lamp examination revealed no granular shaped deposits or diffuse subepithelial deposits in all eight eyes.



**Figure 3.** Slit-lamp photographs of GCD2 heterozygous cases. A-B, case 1; C-D, case 2; E-H, case 5; I-J, case 6; K-L, case 7; M-N, case 8; O-P, case 9; Q-T, case 10. A-B, Preoperative and postoperative slit-lamp photographs at POM 44. C-D, Preoperative and postoperative slit-lamp photographs at POM 41. E, Multiple lattice deposits with diffuse stromal haze which covered the visual axis were observed in a preoperative slit lamp photograph. F, One day after preDALK. G-H, Stromal rejection appeared and regressed with some stromal opacity. I-J, Preoperative and postoperative slit-lamp photographs at POM 30. K-L, Preoperative and

postoperative slit-lamp photographs at POM 30. M-N, Preoperative and postoperative slit-lamp photographs at POM 24. Postoperative photographs were taken wearing rigid gas permeable lens. O-P, Preoperative and postoperative slit-lamp photographs at POM 34. Q-R, The preoperative slit-lamp photograph after three LASIK surgeries showed diffuse, dense, ill-defined corneal deposits. FD-OCT revealed dense corneal deposits located in the stromal interface. S, One day after preDALK. T, Twenty-five months after preDALK.

## IV. DISCUSSION

The optimal treatment of GCD2 has not been established because GCD2 is a genetic disorder and has no valid animal model. Moreover, GCD2 is a very slow progressing disease, so it is difficult to assess the therapeutic efficacy and safety of treatments. Confirmation of the disease progression is time consuming, and multiple factors can affect the progression of GCD2.

In this study, we have defined the recurrence of GCD2 as deposits on the central area of the cornea, detected with slit lamp examination. This was because the clarity of the central area of the cornea mainly affects visual acuity. The deposits usually accumulated much faster in the margins of the donor cornea, and at suture lines which are far from the visual axis, because the abnormal deposits adhered to the corneal wound easily where the collagen fibers of the corneal stroma were cut.<sup>20-22</sup> Therefore, the peripheral corneal recurrence was not thought to be suitable for evaluation of the surgical outcome.

As mentioned above, abnormal deposits of GCD2 accumulated faster in the lesion where the collagen fibers of corneal stroma were cut. Therefore, the preDALK procedure, which leaves a thin layer of the deepest stroma, has the possibility of severe exacerbation of GCD2. This was observed in the eyes of GCD2 patients who formerly received LASIK. Thus, leaving behind the stroma of the recipient can hinder the preDALK procedure for patients with stromal dystrophy. However, Moon *et al.*, found that the abnormal deposits did not accumulate as quickly in the deep stromal interface of the cornea as in the superficial stromal interface.<sup>23</sup> Consequently, our group performed preDALK on GCD2 patients to reduce intraoperative complications which would necessitate conversion to a PKP, and to leave place for another DALK when unexpected severe exacerbation of GCD2 occurred. As was expected, no such interstromal exacerbation occurred during the follow-up periods.

The reason why the abnormal deposits are not observed in the deep host-graft stromal interface in GCD2 is not well understood. One possible theory is that abnormal deposits are produced only in the epithelium so that the abnormal deposits could not migrate a great distance in a short time.<sup>24-26</sup> An alternate possible explanation is that abnormal deposits are produced in the keratocytes of the stroma<sup>27-29</sup> and the keratocytes in the deep stroma are at a low density. Therefore, opacities would not occur in the deep stroma. Another hypothesis is that some unknown substances which are present in the aqueous humor block the accumulation of abnormal deposits located in the deep stroma near the aqueous humor. These unknown substances might have antioxidant effects because the oxidative damage induced by decreased catalase is known to be related to pathogenesis of GCD2<sup>30</sup>. Melatonin, an antioxidant, was reported to have some beneficial effect on GCD2 corneal fibroblasts, *in vitro*.<sup>31</sup> This hypothesis should be further verified.

GCD2 homozygous case 4 showed a recurrence at POM 24, and case 3 showed no recurrence until POM 16. Case 4 also showed small dots of abnormal deposits in the stromal interface on POM 63. However, the opacities at the stromal interface were thought to have little effect on the visual quality because the opacities were smaller in number and size than that of the opacities in the sub-Bowman layer. Therefore, the presence of opacities at the stromal interface would not provide reason to not perform preDALK on homozygous GCD2 patients.

The main location of corneal deposits observed in the recurrent homozygous GCD2 patient was in the sub-Bowman layer, which was the same location as in the non-treated GCD2 homozygous patients (Figure 1G). This pattern of recurrence could be helpful in patient treatment because the deposits were located in the anterior of the cornea, and could be removed with PTK more easily than the deposits located in the posterior of the cornea. In addition multiple PTK would delay another keratoplasty. In the same context, preDALK

procedure could allow the GCD2 patients to receive multiple PTK by providing available corneal stroma, which would be inevitably lost during a PTK procedure. We could not assess the recurrence patterns of heterozygous GCD2 patients because there were no recurrent cases in our study. Additional long-term follow-up data is required for clarification.

The recurrence of GCD2 after therapeutic corneal procedures has been a major concern. The pattern and the time of recurrence have been studied after PKP and DALK. Lyons *et al.*<sup>32</sup> reported the results of PKP and lamellar keratoplasty for granular corneal dystrophy, and both procedures resulted in favorable postoperative vision and similar recurrence rate. The recurrence of granular corneal dystrophy within the graft material was almost universal within 4 years, independent of the performed procedure. Marcon *et al.*<sup>33</sup> reported the recurrence of various corneal stromal dystrophies after PKP in seven eyes with granular dystrophy. No clinically significant recurrence during the 5-year follow-up period was noted with in the patients with granular corneal dystrophy.

Vajpayee *et al.*<sup>34</sup> reported the results of DALK in 10 eyes with corneal stromal dystrophies, including one case of granular corneal dystrophy. Park *et al.*<sup>20</sup> also reported on DALK in four eyes of GCD2 patients. The two reports revealed good short-term results with no recurrence or with clinically insignificant recurrence. In the study of Salouti *et al.*<sup>22</sup>, five of seven eyes had simple recurrence and three of seven eyes had clinical recurrence, which is a similar definition of 'recurrence' used in our study, after DALK with the Melles technique at,  $34 \pm 2$  months. However, previous reports on DALK in eyes of granular corneal dystrophy have limitations, such as an unclear distinction of the type of granular corneal dystrophy, no description of genetic mutation type, and short follow-up periods.

In the present study, there was no recurrence during the follow-up periods in heterozygous cases ( $45.1 \pm 13.9$  months). On the other hand, the recurrence

time of homozygous patients was at least 16 months and 24 months. It is difficult to compare the recurrence rates of GCD2 patients because previous studies have used different definitions for recurrence and because there is a lack of genetic information. The recurrence times and patterns, however, were quite similar between the PKP group in the study of Marcon *et al.*<sup>33</sup> and the preDALK group with heterozygous mutations in the present study. Furthermore, the recurrence pattern was also similar with the DALK group reported by Park *et al.*<sup>20</sup>, Salouti *et al.*,<sup>22</sup> and the preDALK group in the present study. Therefore, we recommend that the recurrence of GCD2 should not be the reason for conducting PKP or DALK rather than preDALK.

In two cases of GCD2 patients, microperforation and rupture of DM occurred during surgery. This complication, however, was well resolved with air injection. Other possible postoperative complications such as ocular hypertension, double anterior chamber and infectious keratitis were not observed in any of the cases.

Half of the enrolled patients had epithelial or stromal rejections. All rejections were well treated with topical steroid eye drops, despite some remaining corneal opacity in the stroma (cases 3, 4, 5, 6, and 10). As known in the previous literature,<sup>35-37</sup> none of the cases suffered from endothelial rejection, and any rejections were well controlled without severe damage to the graft. It is known that keratoplasty procedures which reserve the endothelium of the recipient provide longer graft survival than PKP procedures.<sup>38</sup> In cases of DALK and preDALK, the preDALK procedure provides decreased risk of intraoperative complications such as DM perforation. In addition, reoperation with preDALK could be conducted more easily in the technical aspect than DALK due to the thicker corneal residual bed. PreDALK, therefore, is thought to be more suitable than DALK for GCD2, a lifelong, inextirpable disease. In summary, the preDALK procedure should be considered as a first choice of surgery in patients who need keratoplasty due to the progression of GCD2.

Limitations of this study include its retrospective design, small case number, and the absence of controls. However, as the GCD2 patients who require keratoplasty are uncommon, and as the most common type of GCD2, heterozygous GCD2 is a slow progressing disease, it is difficult to design a prospective type of study.

In conclusion, preDALK conducted on GCD2 patients seems to give an equivalent recurrence-free period in comparison to that of PKP and DALK. Furthermore, preDALK is predicted to provide a longer-term graft survival than PKP and reoperation after the preDALK procedure is thought to be less complicated when compared to the reoperation after the DALK. Therefore, preDALK should be considered as a primary surgical option for GCD2 patients with severe visual impairment who need to undergo PKP and DALK.

## V. CONCLUSIONS

In this study, we reviewed 10 eyes of nine GCD2 patients who underwent preDALK and were followed for more than 30 months. The mean preoperative UCVA was  $0.43 \pm 0.24$  (logMAR) and the mean postoperative BCVA was  $0.14 \pm 0.11$  (logMAR). Five eyes experienced one or multiple graft rejections but all of the rejections were well treated with proper management, and none of the cases had graft failure or required another keratoplasty during the follow-up period. The recurrence of GCD2 was only observed in homozygous type GCD2 patients.

The preDALK of GCD2 patients seems to have at least an equivalent recurrence-free period in comparison to that of PKP and DALK as reported in previous studies. Furthermore, preDALK is predicted to provide a longer-term graft survival than that of PKP, and reoperation after the preDALK procedure can be conducted with fewer complications than after the DALK procedure. Therefore, preDALK should be considered as a primary surgical option for GCD2 patients with severe visual impairment who need to undergo PKP and DALK.

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## ABSTRACT (IN KOREAN)

중증의 제2형 과립형 각막이상증환자들의 치료방법으로써  
데스메막 직전까지 시행한 심층층판각막이식술의 장기임상결과

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최 영 준

본 연구는 한구의 중증 제2형 과립형 각막이영양증환자를 대상으로 데스메막 직전까지 시행한 심층층판각막이식술의 장기임상결과를 보고하고자 계획되었다.

본 연구는 데스메막 직전까지 시행한 심층층판각막이식술을 시행 받은 환자들의 의무기록을 바탕으로 후향적으로 시행되었다. 환자들은 모두 수술 전 유전자 검사를 통하여 transforming growth factor- $\beta$  induced gene (TGHBI)의 R124H 변이를 확인하였다. 심층층판각막이식술을 시행한 후 시력, 굴절력, 각막굴절률, 합병증, 거부반응 및 재발을 평가하였다.

총 9명의 환자의 10안이 수술 후 30개월 이상 경과관찰되어 본 연구에 포함되었으며 평균 경과관찰기간은  $48.9 \pm 14.6$  개월이었다. 2안은 동형접합성 변이를 보였으며 나머지 8안은 이형접합성 변이를 보였다. 수술 후 최대교정시력은 평균  $0.14 \pm$

0.11 (logMAR)였으며 동형접합성 변이를 지닌 2안은 경과관찰기간 중 재발하였으나 이형접합성 변이를 지닌 8안은 경과관찰기간 ( $45.1 \pm 13.9$ 개월) 중 모두 재발하지 않았다.

제2형 과립형 각막이영양증환자를 대상으로 데스메막 직전까지 시행하는 심층층판각막이식술은 재발하지 않는 기간은 최소한 전층각막이식술과 데스메막까지 시행하는 심층층판각막이식술과 동등하다고 생각된다. 더욱이, 데스메막 직전까지 시행하는 심층층판각막이식술은 전층각막이식술에 비하여 이식편의 장기생존을 기대할 수 있으며 데스메막까지 시행하는 심층층판각막이식술에 비하여 재수술시 부작용이 적다. 그렇기 때문에 데스메막 직전까지 시행하는 심층층판각막이식술은 심한 시력저하로 각막이식술이 필요한 제2형 과립형 각막이영양증환자들에게 우선적으로 고려되어야 한다.

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핵심되는 말 : 심층층판각막이식술, 제2형 과립형 각막이영양증