

## Repeated phototherapeutic keratectomy (PTK) followed by PTK with photorefractive keratectomy for anterior granular corneal dystrophy

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Two preteen siblings with the anterior-stromal variant of granular corneal dystrophy type 1 underwent multiple phototherapeutic keratectomies (PTK) (due to recurrences of the dystrophy) with progressively increasing hyperopia after each procedure. The last procedure performed was an additional photorefractive keratectomy along with the PTK which led to a decrease in the hyperopia with better refractive outcomes. The addition of mitomycin C may have led to a delay in the recurrence of the dystrophy.

**Key words:** Granular corneal dystrophy, mitomycin C, photorefractive keratectomy, phototherapeutic keratectomy, stromal corneal dystrophy

Though symptoms of granular corneal dystrophy type 1 (GCD1) usually appear after the second decade, these may start as early as 18 months especially in homozygous child, with suboptimal vision at 4 years, which may potentially be amblyogenic.<sup>[1]</sup> We report two siblings with early-onset of clinically diagnosed anterior stromal GCD1, which was recurrent after repeated phototherapeutic keratectomies (PTKs) and ultimately needed PTK with photorefractive keratectomy (PRK) to treat both the corneal opacity and post-PTK hypermetropia. We believe that these cases are the youngest GCD1 patients to undergo PTK.

### Case Reports

#### Case 1

A 6-year-old girl had best-corrected visual acuity (BCVA) of 6/60 in both eyes (BE) due to GCD1 in May 2013 [Fig. 1a]. The

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opacities were noted at 105  $\mu$  (right eye/RE) and 110  $\mu$  (left eye/LE) from corneal surface [Fig. 1b].

Parents (nonconsanguineous marriage) had normal vision with minimal GCD1 changes [Fig. 2c] on 2020. Paternal grandfather had dense GCD1 changes (visual decline starting in the fifth decade) in the anterior and mid stroma.

PTK was done under total intravenous anesthesia (TIVA) – midazolam and propofol in BE (May 2013). BE regained BCVA of 6/18 (+2.0).

The dystrophy recurred (BCVA RE-3/60, LE-6/60) requiring repeat PTK in December 2014 (RE) and January 2015 (LE). BCVA improved to 6/18 BE (+4.0) after 1 month. In February 2016, central corneal thickness (CCT in ASOCT) was 545 (RE) and 538  $\mu$  (LE) respectively.

Recurrence necessitated third PTK (May 2016). BCVA improved to 6/18 (+6.0D BE) 3-weeks postoperatively.

When GCD1 recurred again [Figs. 1c, 3a, and 4a] (BCVA 6/60 BE), PTK + PRK with Mitomycin C (MMC) 0.02% for 30 s (to correct + 9D hyperopia, subjective refraction + induced hyperopia) was done (RE: November 2017, LE: January 2018), which improved corneal clarity and flattening [Figs. 1d, 3b, and 4b].<sup>[2]</sup> After initial PTKs, keratorefractive-surgical corrections were not done due to good BCVA with hyperopic glasses. However, with increase in hyperopic refraction in this possibly amblyogenic age, the case was discussed with peers, and PRK was performed. In September 2017, CCT (on OCT) was RE-478  $\mu$  and LE-534  $\mu$ .

BCVA was 6/9 (RE + 2.5/+1.50  $\times$  110) in December 2017 and 6/12 (LE + 2.75/+1.0  $\times$  90) in February-2018. In January 2018 (before LE PRK + PTK), CCT (on OCT) was RE-416  $\mu$  and LE-530  $\mu$ .

On the last visit (June 2020, 2 ½ year after the last surgery), BCVA was RE-6/18 (+2.50/+1.50  $\times$  10) and LE-6/12 (+2.75/+1.00  $\times$  90) with more severe recurrence [Fig. 2a and b].

#### Case 2

The elder brother of case 1 (8-year-old at presentation) had poor BCVA (RE-6/60, LE-6/18) due to similar opacities. After RE-PTK (August 2014), BCVA improved to 6/12 (+2.00). Recurrence necessitated repeat PTK (June 2016), which increased hyperopia (+6.0). The parents denied LE intervention.

In December 2018, RE PTK + PRK with MMC (to correct + 9.0 hyperopia, subjective-refraction and induced-hyperopia) was done under TIVA. BCVA improved to 6/12 (+1.25  $\times$  170) after 1 month, which was maintained till June 2020.

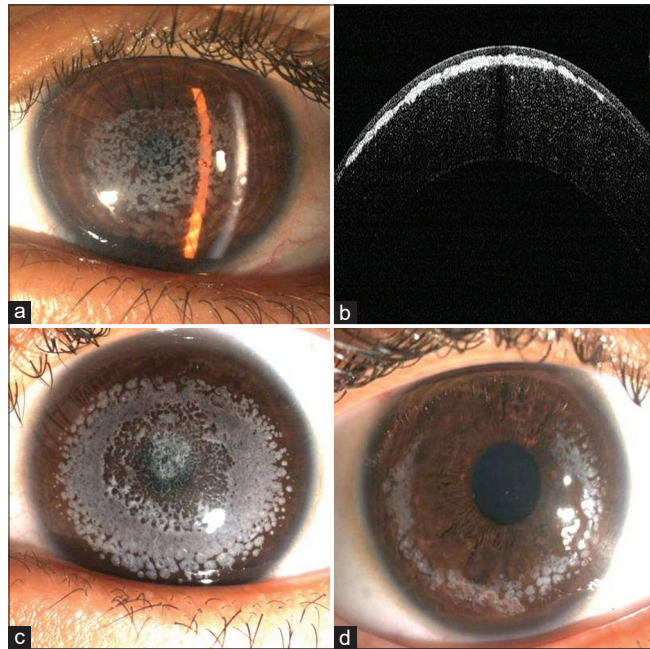
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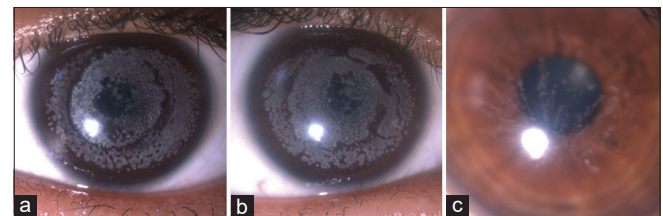
### Discussion

Definitive procedures like penetrating keratoplasty (PKP) in corneal dystrophies may have complications including recurrence, graft rejection, infection, and induced astigmatism.

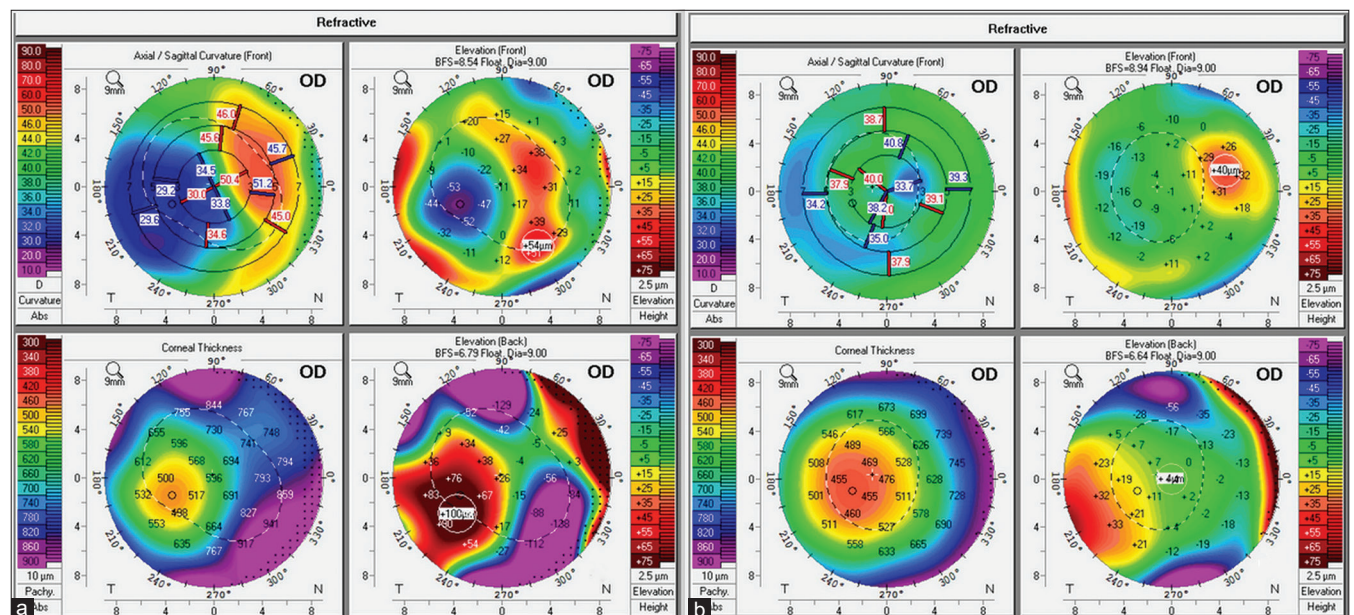


**Figure 1:** (a) Slit-lamp picture of the right cornea of the female sibling (case 1) showing typical features of GCD1. (b) Anterior segment-OCT image (case 1) showing granular opacities in the anterior corneal stroma at presentation. (c) Slit-lamp picture of the right eye (case 1) showing recurrence of dense bread crumb-like corneal opacities after PTK (just before PTK + PRK). There is superficial haze of intervening cornea, which may be related to recurrence or post-PTK haze. (d) Slit-lamp image of RE cornea of case 1 after PTK + PRK showing the clearing of the central cornea

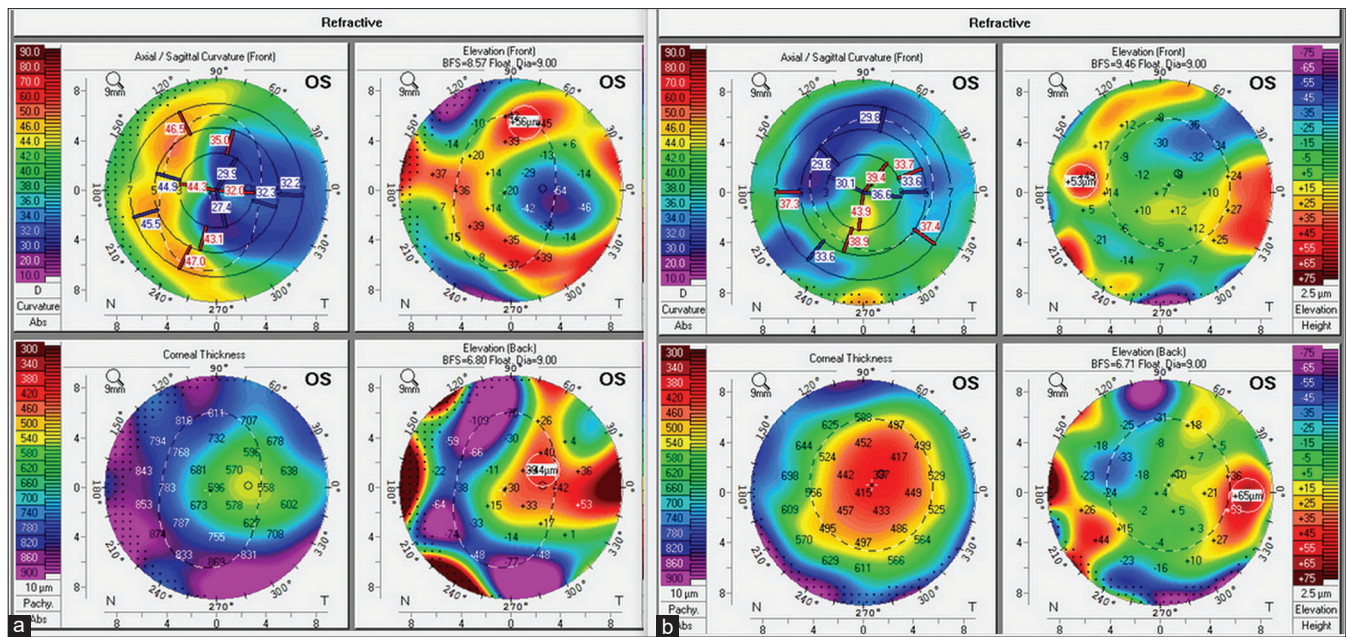
A large series on GCD recommended use of lamellar keratoplasty (LK) as primary procedure especially in young patients who may need multiple procedures due to the lower morbidity rate.<sup>[3]</sup> LK includes anterior LK (ALK) (femtolasers or microkeratome-assisted) or deep ALK. Excimer laser procedures are easier to perform, less aggressive, repeatable, relatively safer, have faster recovery, and do not have interface problems like ALK.<sup>[4,5]</sup> Also, the long wait for suitable cornea because of scarcity of cornea may be avoided. Though PTK has been traditionally used in epithelial and subepithelial dystrophies, it has been used with success in GCD before or after keratoplasty for recurrences.<sup>[6-8]</sup> Repeated deep PTK can cause postoperative haze/scarring, corneal flattening, and hyperopia.<sup>[2]</sup> PTK and PRK with MMC has been used successfully in Thiel-Benke corneal dystrophy, GCD and even in macular corneal dystrophy.<sup>[5]</sup> Recurrence after PTK is known.<sup>[4,6,8]</sup> In a study of 50 eyes which underwent PTK, significant recurrence was noted in 47% eyes with Reis-Buckler corneal dystrophy (RBCD) after an average of 21.6 months, 23% in GCD at postoperative mean 40.3 months, and 14% in lattice corneal dystrophy at 6 months postoperatively.<sup>[8]</sup> GCD was the slowest to recur of all the corneal dystrophies in this study.<sup>[8]</sup> Recurrence after a second PTK was noted in RBCD and GCD.<sup>[8]</sup> Another study on PTK for recurrent GCD after PKP noted that mean duration between PKP and first PTK, first PTK and second PTK, second PTK, and third PTK was 85.1, 62.12, and 42.8 months, respectively.<sup>[7]</sup>



**Figure 2:** (a and b) The slit lamp image of the anterior segment (case 1) of the right eye (a) and the left eye at final follow-up (June 2020). (c) The magnified slit lamp picture of the anterior segment of the father at final follow-up (June 2020)



**Figure 3:** (a) Schiempflug image (Pentacam) refractive 4 map (after the third PTK) and before PTK + PRK of the right cornea of case 1 showing flat cornea with keratometry values of 33.1 D and 37.5 D and an irregular surface. (b) Schiempflug image (refractive 4 map) after PTK + PRK of RE of case 1 showing a more regular surface and improved D keratometry values (35.9/38.1 D)



**Figure 4:** (a) Schiempflug image (refractive 4 map) of the left cornea of case 1 after the third PTK and before PTK + PRK showing flat keratometry (30.3/37.5 D) with an irregular surface. (b) Schiempflug image (refractive 4 map) after PTK + PRK of LE of case 1 showing a more regular surface and reduced flattening. The keratometry was 33.9/39.8D

The rapid recurrence in our cases may be due to homozygous status. The recurrence pattern after PTK may be different in homozygous (early, more severe, diffuse and peripheral between epithelium, and ablated area) and heterozygous (late, milder, granular opacities at central cornea) mutations of R124H (BIG-H3).<sup>[9]</sup> Our limitation includes avoidance of masking agent during PTK, absence of histopathology/genetic analysis/aberrometry, and limited follow-up period.

PTK was done each time using 6 mm ablation zone and 0.5 mm transition zone and depth was 60  $\mu$  after the mechanical scraping of the epithelium. PRK was done to correct both preoperative and expected postoperative hyperopia after simultaneous PTK. MMC was used only in the last procedure in both patients, which may reduce the postoperative haze and possibly reduce or delay the recurrence, though this effect is debatable.<sup>[10]</sup> In the Excimer laser suite, general anesthetic gases may impede working of the laser. Hence, TIVA was used.

## Conclusion

In conclusion, we report two children with anterior stromal GCD, who were first treated with PTK but required repeated procedures due to the recurrent disease. Also, repeated PTK was noted to cause hyperopia, which needed an additional PRK in both the cases leading to good visual outcome.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

## References

- Møller HU, Ridgway AE. Granular corneal dystrophy Groenouw type I. A report of a probable homozygous patient. *Acta Ophthalmol (Copenh)* 1990;68:97-101.
- Rapuano CJ. Excimer laser phototherapeutic keratectomy for corneal dystrophies. *US Ophthalmic Rev* 2006;30-2.
- Lyons CJ, McCartney AC, Kirkness CM, Ficker LA, Steele AD, Rice NS. Granular corneal dystrophy. Visual results and pattern of recurrence after lamellar or penetrating keratoplasty. *Ophthalmology* 1994;101:1812-7.
- Stewart OG, Pararajasegaram P, Cazabon J, Morrell AJ. Visual and symptomatic outcome of excimer phototherapeutic keratectomy (PTK) for corneal dystrophies. *Eye Lond Engl* 2002;16:126-31.
- Hsiao C-C, Hou Y-C. Combination of phototherapeutic keratectomy and wavefront-guided photorefractive keratectomy for the treatment of Thiel-Behnke corneal dystrophy. *Indian J Ophthalmol* 2017;65:318-20.
- Hahn TW, Sah WJ, Kim JH. Phototherapeutic keratectomy in nine eyes with superficial corneal diseases. *Refract Corneal Surg* 1993;9:S115-118.
- Rathi VM, Taneja M, Murthy SI, Bagga B, Vaddavalli PK, Sangwan VS. Phototherapeutic keratectomy for recurrent granular dystrophy in postpenetrating keratoplasty eyes. *Indian J Ophthalmol* 2016;64:140-4.
- Dinh R, Rapuano CJ, Cohen EJ, Laibson PR. Recurrence of corneal dystrophy after excimer laser phototherapeutic keratectomy. *Ophthalmology* 1999;106:1490-7.
- Inoue T, Watanabe H, Yamamoto S, Inoue Y, Okada M, Hori Y, Maeda N, Inoue Y, Hayashi K, Shimomura Y, Tano Y. Different recurrence patterns after phototherapeutic keratectomy in the corneal dystrophy resulting from homozygous and heterozygous R124H BIG-H3 mutation. *Am J Ophthalmol* 2001;132:255-7.
- Yuksel E, Cubuk MO, Eroglu HY, Bilgihan K, Yuksel E, Cubuk MO, Eroglu HY, Bilgihan K. Excimer laser phototherapeutic keratectomy in conjunction with mitomycin C in corneal macular and granular dystrophies. *Arq Bras Oftalmol* 2016;79:69-72.