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Corneal Lenses in Recurrent Granular Corneal Dystrophy

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BACKGROUND

Granular corneal dystrophy (GCD) is a rare inherited corneal condition that manifests as bilateral stromal opacities in the first decade of life.[1] Penetrating keratoplasty (PKP) and deep anterior lamellar keratoplasty (DALK) are considered when the stromal opacities become visually significant but can result in irregular astigmatism. Rigid lenses provide the benefit of correcting irregular astigmatism.[1]

CASE HISTORY

Chief Complaint:

- A 33-year-old Hispanic male presented for a contact lens fitting due to blur in both eyes worsening four months ago.

Ocular History:

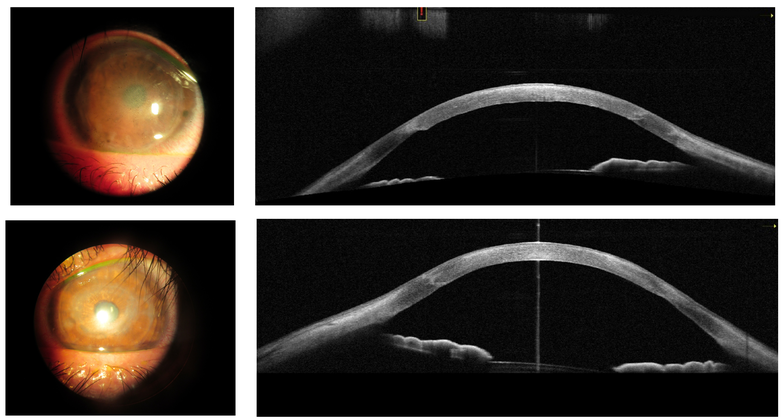
Ocular history
- Diagnosed with granular corneal dystrophy (type unknown) at age 15
- PKP OD and DALK OS at age 25 due to reduced vision from significant central stromal opacities

Family History
- Granular corneal dystrophy (type unknown): mother, brother, maternal grandmother

EXAMINATION FINDINGS

	OD	OS
DVA sc	20/150	20/150
Manifest refraction	-1.75 -2.50 x155 (DVA 20/80)	-4.75 -2.00 x105 (DVA 20/40)
Pentacam tomography	Irregular astigmatism: 48.2D/44.4D @130	Irregular astigmatism: 51.4D/49.7D @090
Anterior Segment	PKP graft with scattered neovascularization fronds encroaching 3-4mm 360 2+ central stromal granular opacities (-) corneal erosions	DALK graft with scattered neovascularization fronds encroaching 3-4mm 360 Paracentral stromal scarring in ring-like pattern (-) corneal erosions
Posterior Segment	Unremarkable	Unremarkable

OD



OS

Figures 1 and 2. Anterior segment photo of OD (L) and anterior segment OCT of OD with central stromal opacities (R)

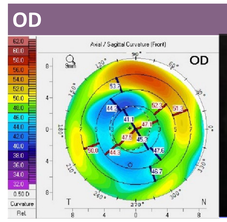
Figures 3 and 4. Anterior segment photo of OS (L) and anterior segment OCT of OS (R)

MANAGEMENT

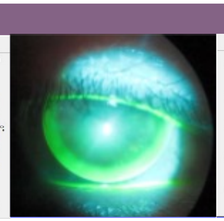
- The patient was fit in large diameter corneal lenses in both eyes.
- Scleral lenses were not pursued due to risk of exacerbation of corneal neovascularization and good potential for successful corneal lens wear.
- Best corrected DVA through the right corneal lens was limited by the recurrence of central granular stromal hyaline deposits in the graft tissue.
- Repeat penetrating keratoplasty was not pursued due to good vision achieved with corneal lenses OU.

Final Corneal RGP lens parameters					
	Base Curve	Diameter (mm)	Optic Zone (mm)	Peripheral Curves (mm)	DVA
OD	47.75D (7.07mm)	10.4	7.8	9.00 x 1.1 12.00 x 0.2	20/40 (PHNI)
OS	49.00D (6.88mm)	10.4	8.6	9.00 x 0.7 12.00 x 0.2	20/20

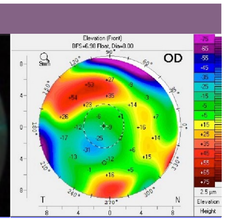
OD



OD Axial Map

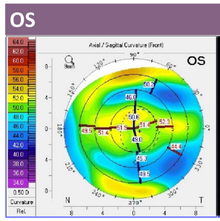


OD Final Corneal Lens

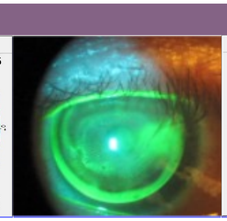


OD Elevation Map

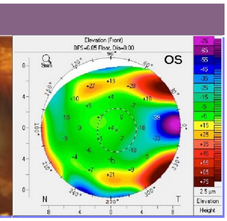
OS



OS Axial Map



OS Final Corneal Lens



OS Elevation Map

CONCLUSION

- Granular corneal dystrophy (GCD) results from a mutation in the TGFBI gene and is inherited in an autosomal dominant pattern.[1,2]
- Two subtypes of GCD exist:
  - Type 1 (GCD1): granular deposits (composed of hyaline) with ”breadcrumb” appearance[2]
  - Type 2 (GCD2): granular deposits (composed of hyaline) with later onset of lattice deposits (composed of amyloid)[2]
- In GCD1, stromal opacities begin to form usually in the first decade of life and enlarge over time, taking on a granular appearance and causing a reduction in vision.[2]
- Surgical management of GCD involves penetrating keratoplasty (PKP) or deep anterior lamellar keratoplasty (DALK) to remove the stromal opacities and improve the patient’s vision.[3]
- Phototherapeutic keratectomy (PTK) has been utilized in recent years to improve vision in patients with GCD by removing superficial stromal opacities.[2]
- Rigid contact lenses can improve vision in patients with PKP or DALK due to their ability to mask irregular astigmatism.
- GCD recurrence rate has been found to be 22-90% in patients with post-PKP corneas, with a median time of 2.6 years to recurrence.[3,4]
- Limitations in visual correction with corneal lenses may exist depending on the presence and severity of recurrent GCD in the graft tissue.
- Repeat PKP or DALK may be considered when the recurrent GCD leads to visually significant stromal opacities.

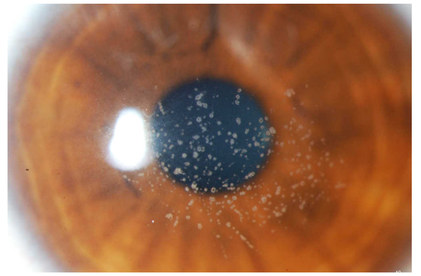


Figure 5. Granular Corneal Dystrophy Type 1[5]

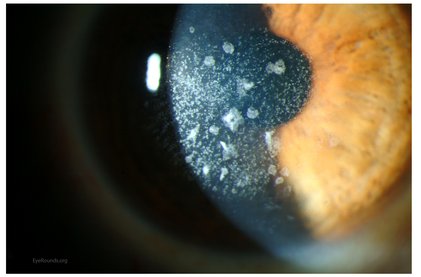


Figure 6. Granular Corneal Dystrophy Type 2[6]

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