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**Case Report**

# Spontaneous Corneal Clearing after Descemet Membrane Rupture and Near-Total Detachment in Keratoglobus: A Case Report

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**Keywords**
Keratoglobus · Osteogenesis imperfecta · Hydrops · Descemet rupture · Corneal ectasia

**Abstract**
***Introduction:*** We present a case of a patient with osteogenesis imperfecta (OI) and keratoglobus (KG) who had a near-total rupture of Descemet's membrane followed by spontaneous corneal clearing. This case is unique in that it demonstrates the potentially excellent outcome of conservative treatment for Descemet's rupture in patients with KG and illustrates the impressive migratory potential of healthy endothelial cells. ***Case Presentation:*** An 11-year-old girl with OI and KG who had rupture and near-total detachment of Descemet's membrane presented for evaluation. This was managed conservatively and resulted in the eventual spontaneous clearing of the cornea. A similar process happened in the fellow eye some years later. Given the result of conservative management originally, the patient was once again treated conservatively, with significant improvement in corneal edema and visual acuity. ***Conclusion:*** Given the size of the ruptures, this case highlights the dynamic nature of the corneal endothelium and provides an extreme example of the migratory potential of corneal endothelial cells.



## Introduction

Keratoglobus (KG) is a rare corneal ectasia characterized by diffuse thinning of the cornea, disruption of Bowman's layer, and ruptures in Descemet's membrane [1]. These histopathological changes contribute to global protrusion and visual impairment secondary to myopia, irregular astigmatism, hydrops, and corneal scarring.

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Spontaneous rupture of Descemet's membrane, or hydrops, results in corneal edema secondary to the influx of aqueous humor from the anterior chamber into the corneal stroma. Occurrences have been reported in association with connective tissue disorders such as osteogenesis imperfecta (OI), Ehlers-Danlos type VI, and Leber's congenital amaurosis, and with corneal ectatic disorders including keratoconus, pellucid marginal degeneration, and KG [2]. The natural history of hydrops is well understood – a break, and partial detachment, of Descemet is followed by corneal edema, pain, photophobia, and decreased vision. Over many weeks, endothelial cells migrate over the previous area of rupture and re-establish a state of corneal deturgescence.

It is not unusual for a Descemet's rupture to be associated with partial membrane detachment [3]. In these instances, it is not entirely understood whether the migration of endothelial cells comes from those still attached to the cornea and in close proximity to the rupture or if cells on free-floating Descemet's have the migratory potential to travel around the edge of the rupture and back up onto the cornea. In other words, what are the migratory limits of the corneal endothelial cells?

We present a case of a patient with OI and KG who had a near-total rupture of Descemet's membrane followed by spontaneous corneal clearing, suggesting extensive migration or auto-transfer of endothelial cells and re-establishment of a new Descemet's membrane. This case is unique in that it demonstrates the potentially excellent outcome of conservative treatment for Descemet's rupture in patients with KG and illustrates the impressive migratory potential of healthy endothelial cells. Written informed consent was obtained from the parent of the patient for publication of this case report and any accompanying images. The CARE Checklist has been completed by the authors and is available as supplementary material.

## Case Report

An 11-year-old girl with a past medical history of OI and KG presented for evaluation. She reported an episode of acute corneal hydrops OS at age 7, treated at an outside facility. At the time, she was started on sodium chloride (Muro®) 128 ointment BID and prednisolone acetate 1% eye drops BID, both of which were slowly tapered over 3 months. An endothelial keratoplasty had been recommended when the cornea did not initially clear, but the patient's parents declined, and the patient was lost to follow-up until she presented to the outside facility again at age 10. At that time, her cornea had cleared OS. She was referred to our tertiary care facility for monitoring.

On exam, her best corrected visual acuity (BCVA) was 20/100 OD and 20/60 OS, with a manifest refraction of −7.75 + 0.75 × 175 OD and −9.50 + 2.00 × 110 OS. There was a blue/gray discoloration to the sclera OU. Peripheral scarring was noted in the right eye. In the left eye, Descemet's membrane was noted to be mostly free floating in the anterior chamber; it was adherent only in the far periphery. A large "cat-eye"-shaped Descemet rupture was noted centrally within the flap (Fig. 1). Anterior segment OCT of the left eye confirmed an old, large Descemet detachment, with Descemet seemingly free-floating within the anterior chamber (Fig. 2). The overlying cornea was clear and compact at the time of examination.

The patient presented acutely at age 11 with pain, photophobia, and decreased vision in the right eye. BCVA was "count fingers" in the right eye and 20/60 in the left eye. An anterior segment OCT was obtained (Fig. 3), but further photographs at this timepoint were not possible due to the patient's extreme light sensitivity and discomfort. Slit lamp exam was notable for limbus-to-limbus corneal edema suggestive of corneal hydrops. Given the history of an excellent response to conservative management in the left eye, treatment with Muro 128 drops TID and Muro 128 ointment QHS was elected.

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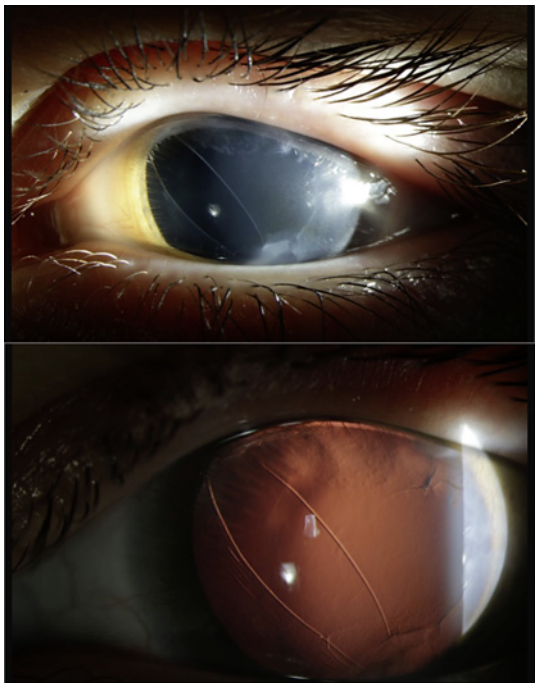

**Fig. 1.** Slit lamp photos of the patient's left eye in the slit beam and retro-illumination. A large Descemet's rupture is seen with the membrane hanging in the anterior chamber. The overlying cornea is clear and compact.

At the patient's most recent visit, 5 months after acute hydrops OD, BCVA was 20/125 OD and 20/50 OS. There was central clearing of the cornea OD but with persistent microcystic edema peripherally. Now, with the cornea clearer, it was immediately evident that the patient had experienced a similar event to that of the left eye, inclusive of a large rupture and near total Descemet detachment (Fig. 1, 2). Descemet's membrane was attached only at the peripheral limbus; it had not re-attached, yet the cornea had cleared. Her pain had resolved, and she had tapered off all topical treatment. Continued observation was elected given the slow clearing of the cornea and improved VA.

## Discussion

Acute corneal hydrops is an uncommon complication of keratoconus and is estimated to occur in approximately 3% of patients with keratoconus and 11% of patients with KG [4, 5]. The severity of corneal hydrops is divided into 3 grades according to the extent of corneal edema: grade 1 describes edema within a diameter of 3 mm, grade 2 describes edema within a diameter of 3–5 mm, and grade 3 is edema within a diameter greater than 5 mm [6]. Several studies have attempted to determine the clinical risk factors associated with corneal hydrops in keratoconus, which include [4] earlier age of onset, male gender, advanced corneal ectasia [5], poor visual acuity at time of diagnosis, atopy and severe allergic eye disease, and a history of eye rubbing [4–6]. Several conditions have been found to be associated with KG, including atopic conditions, chronic blepharitis, idiopathic orbital inflammation, and trauma [7].

In our case, a patient with OI presented with corneal hydrops secondary to a near-total detachment of Descemet's membrane. While corneal hydrops itself is a known occurrence in this population, the severity observed here is unusual. We observed resolution of corneal edema, presumably achieved through migration of endothelial cells over a great distance, and re-establishment of the endothelial pump function. Note that in our photos, Descemet membrane never re-attached, and so the most plausible explanation is that of endothelial migration.

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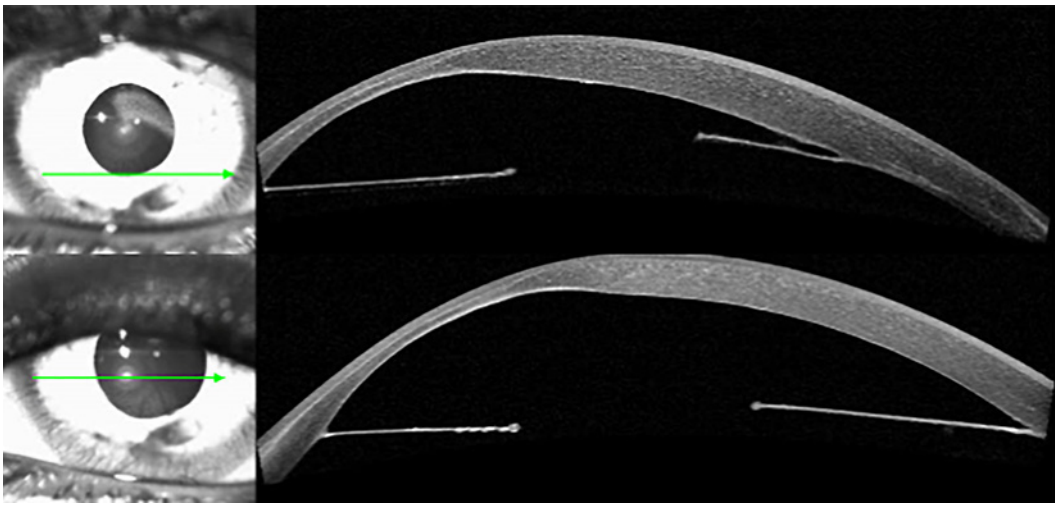

**Fig. 2.** Anterior segment OCT of the left eye demonstrating the large Descemet's rupture with membrane flap in the anterior chamber. The compactness of the overlying cornea and the hyper-reflectivity of the posterior surface demonstrate endothelial cell migration with the re-establishment of Descemet's membrane on the posterior cornea.

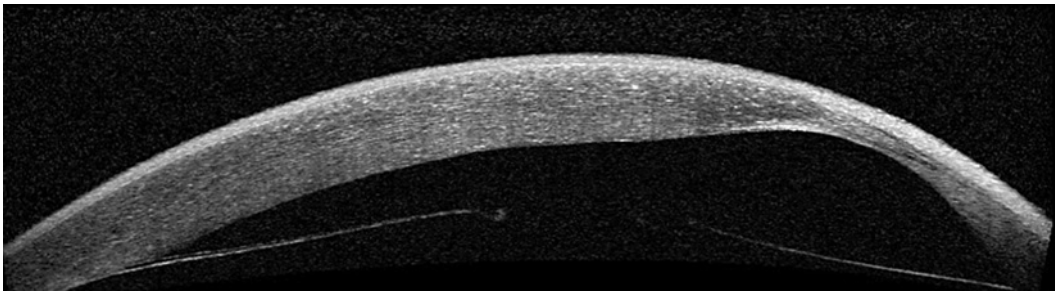

**Fig. 3.** Anterior segment OCT of the right eye shortly after an episode of acute hydrops. A large Descemet's rupture and detachment is noted in the anterior chamber. Lack of a hyper-reflective band at the level of the posterior stroma indicates that endothelial cell coverage has not yet occurred.

The management options for corneal hydrops include conservative management with symptomatic relief, intracameral air or gas injection, compressive sutures alongside gas injection, amniotic membrane transplantation with cautery, cyanoacrylate tissue adhesive with bandage contact lens, and penetrating keratoplasty [8]. Most cases of corneal hydrops resolve over 2–4 months following the migration of adjacent endothelial cells [9]. Milder cases of hydrops tend to be treated more conservatively, with invasive procedures reserved for cases where the affected corneal area is larger and the expected visual outcomes are thought to be more guarded without a higher degree of intervention.

Intracameral gas injection, or pneumodescemetopexy, has established itself as a mainstay of treatment for Descemet's membrane detachment (DMD) and the treatment of corneal hydrops [8]. The injected gas or air acts as a barrier against aqueous humor entering the stroma and aids in the re-approximation of the torn edges of Descemet's membrane. This technique has been applied to DMD in KG with or without compression sutures; reports show resolution of edema within 3 months [10]. However, gas tamponade may not be suitable for all

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cases of KG. There is some evidence that the difference in resolution time between patients treated with intracameral gas injection was not significant for patients with KG [11]. In some cases, attempted pneumodescemetopexy was not successful due to a larger tear with thickened and fibrosed edges [12]. Keratoplasty has also been implicated in the treatment of DMD secondary to KG. DSAEK has been used in cases of KG complicated by acute hydrops in patients not deemed good candidates for pneumodescemetopexy [12].

Another technique used to treat DMD involves stripping the patient's own Descemet's membrane and allowing healthy endothelial cells to migrate and fill the defect. This technique is usually referred to as Descemet's stripping only (DSO) and is often used for the treatment of Fuch's endothelial dystrophy (FED), where there is a relatively small area of dysfunctional endothelium [13]. Although the mechanism of endothelial cell repair is unknown, successful DSO has been shown to have good visual outcomes [13]. Some reports show comparable results between DSO and DMEK for the treatment of selected patients with FED, although the time to corneal clearing is significantly longer with DSO [14].

One of the first clinicians to report using DSO [13] noted the self-healing capacity of endothelial cells in FED. Although much is still unknown about the minimum cell density needed for success, it is largely accepted that there must be an adequate cell count and preserved endothelium over most of the periphery for DSO to be successful. As such, there is a maximum amount of central endothelium that can be removed during DSO with a reasonable expectation of corneal clearing.

There is some literature that suggests there may be other mechanisms at work other than the migration of peripheral cells. There are reports of failed endothelial keratoplasty with free floating grafts in the early postoperative period in which no effort was made to re-attach the graft; the area of stripped native endothelium was large, yet the cornea spontaneously cleared [15]. The theory is that healthy endothelial cells from the failed graft may have been populated into the anterior chamber; these healthy cells were then spontaneously transferred to the bare posterior stroma at the site of the intended graft. This has been termed Descemet membrane endothelial transfer, but the results have been quite mixed and inconsistent, and this is not considered to be an acceptable treatment for endothelial disease [15].

This case outlines an instance where a large area of corneal endothelium detached, yet the cornea spontaneously cleared. Because of the large nature of the rupture, the patient experienced an "auto-DSO" with a large volume of corneal endothelium involved. Over several weeks, this spontaneously improved, and the cornea once again became clear and compact. The affected area was larger than the typically accepted 4 mm central zone, where peripheral endothelial cell migration is thought to account for central corneal clearing in DSO. Our patient demonstrated re-establishment of Descemet's membrane on the posterior cornea with a large flap of original Descemet's membrane in the anterior chamber (Fig. 2); if the re-establishment of healthy endothelial cells occurred purely from migration of healthy peripheral cells, this suggests a remarkable migratory capability of native endothelial cells. However, there may also have been a component of auto-transfer of healthy endothelial cells that were liberated into the anterior chamber from the native flap after the rupture occurred.

In conclusion, KG with endothelial rupture has no consensus on best practices for management. Current practice ranges from intracameral gas injection to endothelial transplant. This case highlights the potential for excellent visual outcomes with observation alone in a patient with a large endothelial rupture. Further evaluation with specular microscopy could aid in elucidating the health and function of endothelial cells after such an event.

## Statement of Ethics

This study was granted an exemption from requiring ethics approval by the Penn State Institutional Review Board. Written informed consent was obtained from the parent of the patient for publication of their medical case and any accompanying images.

## Conflict of Interest Statement

S.P. is a consultant for Bausch & Lomb, Carl Zeiss Meditec, and Hoya Surgical Optics and has received research support from Alcon, Bausch & Lomb, and Carl Zeiss Meditec, all unrelated to the present work. None of the other authors have made any disclosures.

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## Author Contributions

H.E.: writing – original draft, review, and editing and conceptualization. D.R.: writing – review and editing and conceptualization. S.P.: conceptualization, supervision, and writing – original draft, review, and editing.

## Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

## References

1 Meghpara B, Nakamura H, Vemuganti GK, Murthy SI, Sugar J, Yue BYJT, et al. Histopathologic and immunohistochemical studies of keratoglobus. Arch Ophthalmol. 2009;127(8):1029–35.
2 Biglan AW, Brown SI, Johnson BL. Keratoglobus and blue sclera. Am J Ophthalmol. 1977;83(2):225–33.
3 Mulhern M, Barry P, Condon P. A case of Descemet's membrane detachment during phacoemulsification surgery. Br J Ophthalmol. 1996;80(2):185–6.
4 Fan Gaskin JC, Good WR, Jordan CA, Patel DV, McGhee CN. The Auckland keratoconus study: identifying predictors of acute corneal hydrops in keratoconus. Clin Exp Optom. 2013;96(2):208–13.
5 Fan Gaskin JC, Patel DV, McGhee CNJ. Acute corneal hydrops in keratoconus—new perspectives. Am J Ophthalmol. 2014;157(5):921–8.
6 Tuft SJ, Gregory WM, Buckley RJ. Acute corneal hydrops in keratoconus. Ophthalmology. 1994;101(10):1738–44.
7 Wallang BS, Das S Keratoglobus Eye. 2013;27(9):1004–12.
8 Chow VWS, Agarwal T, Vajpayee RB, Jhanji V. Update on diagnosis and management of Descemet's membrane detachment. Curr Opin Ophthalmol. 2013;24(4):356–61.
9 Watson SL, Abiad G, Coroneo MT. Spontaneous resolution of corneal oedema following Descemet's detachment. Clin Exp Ophthalmol. 2006;34(8):797–9.
10 Özcan G, Uçakhan ÖÖ. Surgical management of corneal hydrops: case series. Turk J Ophthalmol. 2022;52(1):64–8.
11 Basu S, Vaddavalli PK, Ramappa M, Shah S, Murthy SI, Sangwan VS. Intracameral perfluoropropane gas in the treatment of acute corneal hydrops. Ophthalmology. 2011;118(5):934–9.

12 Sharma S, Fernandes M. Descemet stripping automated endothelial keratoplasty: an alternate surgical modality for Descemet's membrane detachment following hydrops in keratoglobus. Indian J Ophthalmol. 2020;68(3):513–4.
13 Colby K. Descemet stripping only for fuchs endothelial corneal dystrophy: will it become the gold standard? Cornea. 2022;41(3):269–71.
14 Huang MJ, Kane S, Dhaliwal DK. Descemetorhexis without endothelial keratoplasty versus DMEK for treatment of fuchs endothelial corneal dystrophy. Cornea. 2018;37(12):1479–83.
15 Dirisamer M, Ham L, Dapena I, van Dijk K, Melles GRJ. Descemet membrane endothelial transfer: "free-floating" donor Descemet implantation as a potential alternative to "keratoplasty". Cornea. 2012;31(2):194–7.