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Case Study

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# ACUTE CORNEAL HYDROPS SECONDARY TO UNDIAGNOSED KERATOCONUS

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## ABSTRACT

Acute corneal hydrops is a rare complication of keratoconus characterized by stromal edema due to leakage of aqueous through a tear in Descemet membrane. It can rarely be the first presentation of undiagnosed keratoconus. Here, we report a case of a 22-year-old man who presented with acute corneal hydrops and was diagnosed to have bilateral asymmetric keratoconus. He was successfully treated with Descemetopexy using intracameral perfluoropropane ($C_3F_8$) gas.

**KEYWORDS:** Hydrops, Descemetopexy, Pentacam.

## INTRODUCTION

Acute corneal hydrops is a rare complication with a reported incidence of 2.6 % to 2.8% in individuals with keratoconus.[1,2] It may also occur in other corneal ectasias, such as pellucid marginal degeneration, keratoglobus, and Terrien's marginal degeneration.[3] Corneal hydrops develops following acute rupture of Descemet membrane and overlying endothelium that allows aqueous humor to enter the corneal stroma and epithelium, resulting in severe corneal edema.[4] Unilateral involvement is common, and eye rubbing is the major risk factor.[5] Patients present with pain, photophobia, and decreased visual acuity, although clinical features can range from asymptomatic to marked symptoms.[5] Usually, the edema is self-limiting and will gradually resolve itself, often leaving residual scarring of the involved cornea.[6] However, in some cases, the presence of stromal clefts tends to delay resolution, and persistent edema can incite inflammation and vascularization.[7] Severe infrequent complications of corneal hydrops include extensive corneal scarring, severe neovascularisation, epithelial defects, microbial keratitis, corneal perforation, and glaucoma.[8,9] Acute corneal hydrops may be treated with conservative treatment or surgical intervention, which commonly includes intracameral injection of air/gas.[1,3] Rarely, acute hydrops can be the presenting feature of undiagnosed keratoconus.[10-14]

We report a case of bilateral asymmetric keratoconus with acute hydrops as the initial presentation that was managed with intracameral perfluoropropane ($C_3F_8$) gas injection.

## CASE REPORT

A 22-year-old male presented to our institute with complaints of sudden decrease of vision in the right eye (RE) of 2 days duration, associated with mild tearing and photophobia. He had a history of wearing corrective glasses for the past 12 years and had constant changes to his prescription. There was no history of contact lens wear, trauma to the eye, or excessive rubbing of the eyes. His medical history and family medical history were unremarkable, having no systemic or ocular illness. On examination, his best-corrected visual acuity (BCVA) in the RE was counting fingers close to face, and in the left eye (LE) was 6/6 (partial) with a manifest refraction of -7.00 -1.5 x 100 degrees. Slit-lamp biomicroscopic examination of the RE revealed a central cone with ectasia, a focal area of stromal and epithelial microcystic corneal edema, intrastromal clefts, and a Descemet membrane tear in the centre (Figure 1). LE showed the presence of prominent corneal nerves. Rest of the anterior segment was unremarkable. Intraocular pressure (IOP) and fundus examination were within normal limits.

Topographic and tomographic examination of LE was performed using the Pentacam rotating Scheimpflug camera (Oculus, Wetzlar, Germany). The refractive map showed an astigmatism of 1.5 dioptres (D), a maximum keratometry of 48.6 D, and borderline posterior elevation (Figure 2). The Belin/ Ambrosio Enhanced Ectasia Display (BAD) map revealed an average pachymetric progression index of 1.2, and final D value of 2.53 (Figure 3). Anterior segment- optical coherence tomography (AS-OCT; Optovue, Inc., Fremont, California) imaging of RE showed epithelial and stromal corneal edema with Descemet membrane tear and presence of intrastromal fluid clefts (Figure 4). Based on

clinical findings and investigations, a diagnosis of bilateral asymmetric keratoconus with RE acute hydrops was made. He underwent intracameral $C_3F_8$ injection in RE.

***Surgical technique:*** Preoperatively, the pupil was constricted by topical application of 2% Pilocarpine nitrate eye drops, one drop every 15 min, 1 hour before surgery to avoid intraoperative injury to the lens, and also to prevent the gas from escaping behind the iris. The procedure was performed under topical anesthesia and all aseptic conditions. Anterior chamber (AC) paracentesis was done with a 26-gauge needle. 0.1ml of aqueous humor was aspirated, and 14% non-expansile concentration of $C_3F_8$ was injected enough to fill two-thirds of AC. Postoperatively supine position was advised along with topical antibiotics (Moxifloxacin 0.5%), corticosteroids (Prednisolone acetate 1%), hyperosmotics (Sodium chloride 5%), and preservative-free lubricants (Sodium hyaluronate 1.5%)

On postoperative day 1, $C_3F_8$ gas was found to fill the upper third of the anterior chamber, and IOP was normal. One week later, the BCVA in RE improved to 6/60 with a reduction in epithelial and stromal edema, IOP was normal, and the AS-OCT showed a reattached Descemet membrane (Figure 5). At a follow-up of 3 weeks, the RE BCVA was 6/18 with complete resolution of corneal edema, normal IOP, and a nebular grade corneal scar (Figure 6). The patient was also advised to undergo corneal collagen cross-linking in LE.

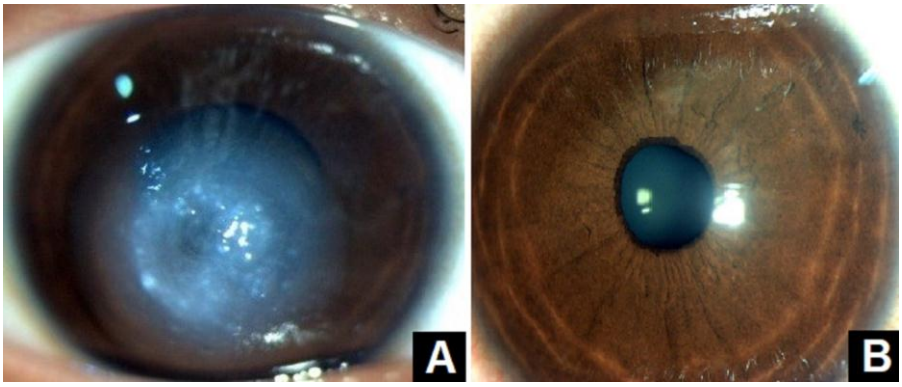


**Figure 1: Slit-lamp photograph of the anterior segment of the (A) right eye showing a focal area of macrocystic and microcystic corneal edema with a central Descemet membrane tear (B) normal left eye.**

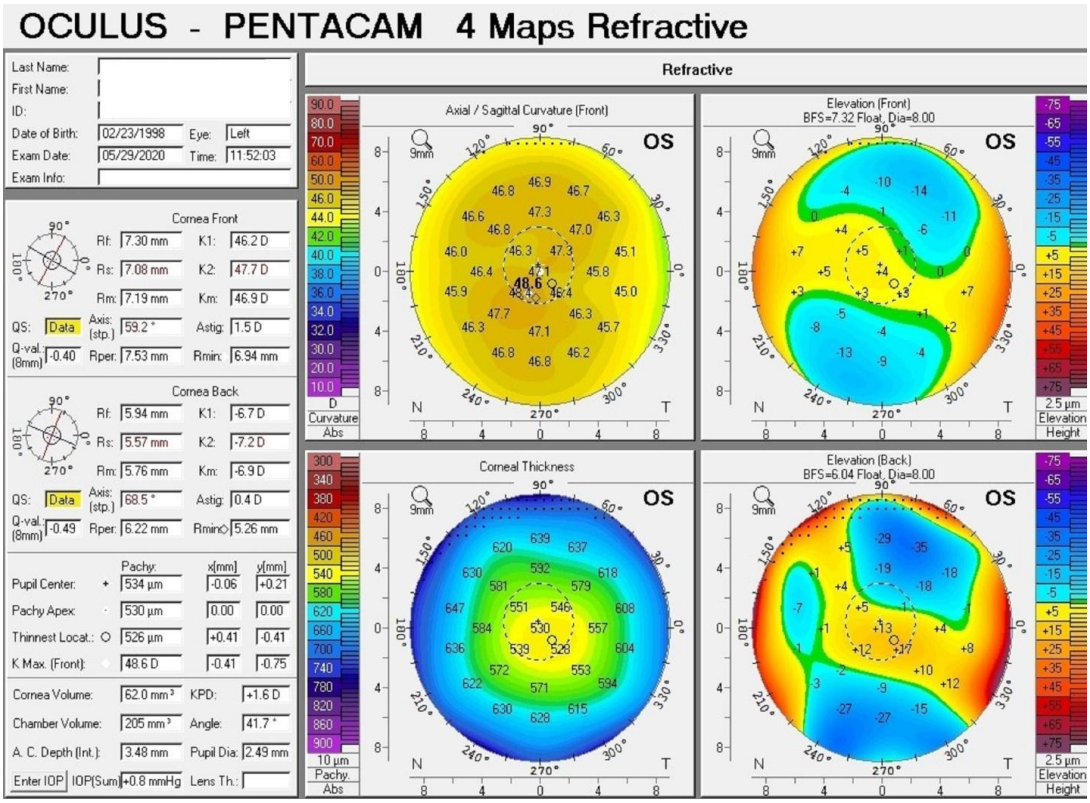


**Figure 2: Left eye Pentacam corneal refractive map showing a maximum keratometry of 48.6 D and borderline posterior elevation.**

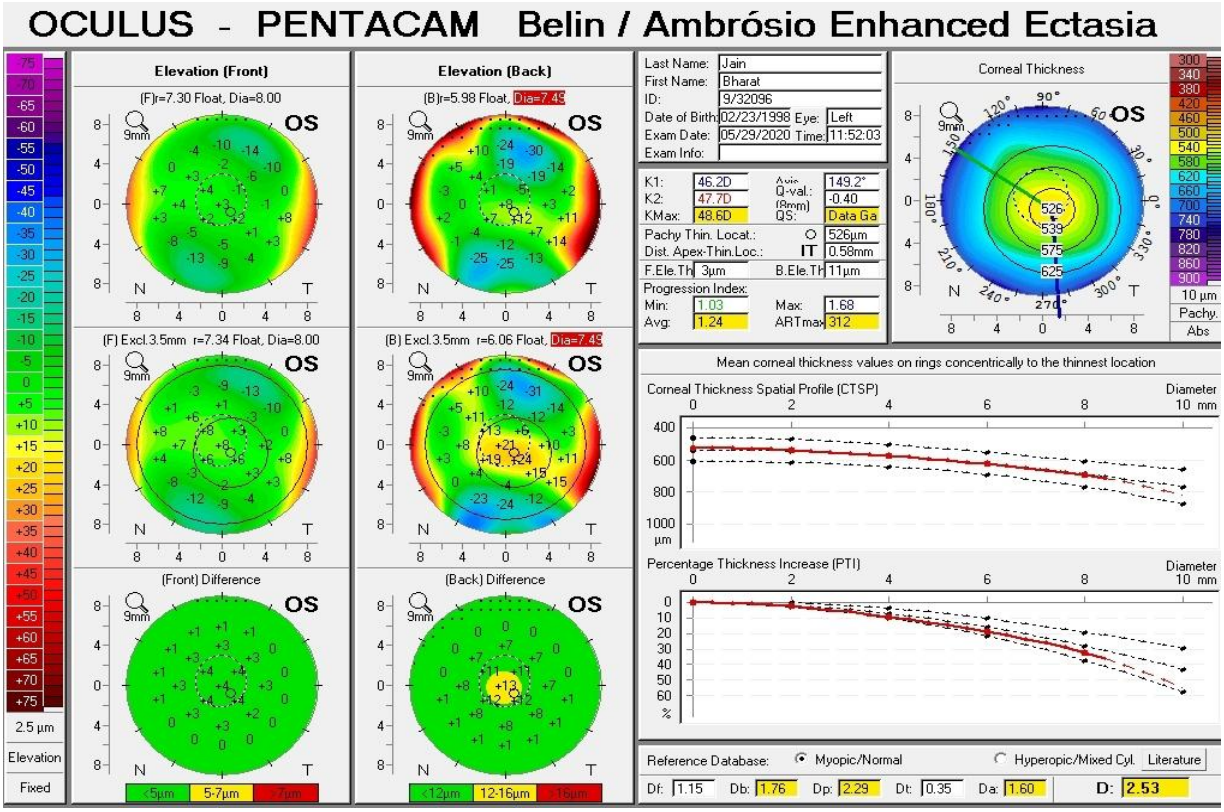


**Figure 3: Left eye Pentacam Belin/ Ambrosio Enhanced Ectasia Display map showing an average pachymetric progression index of 1.2, and final dioptre value of 2.53.**

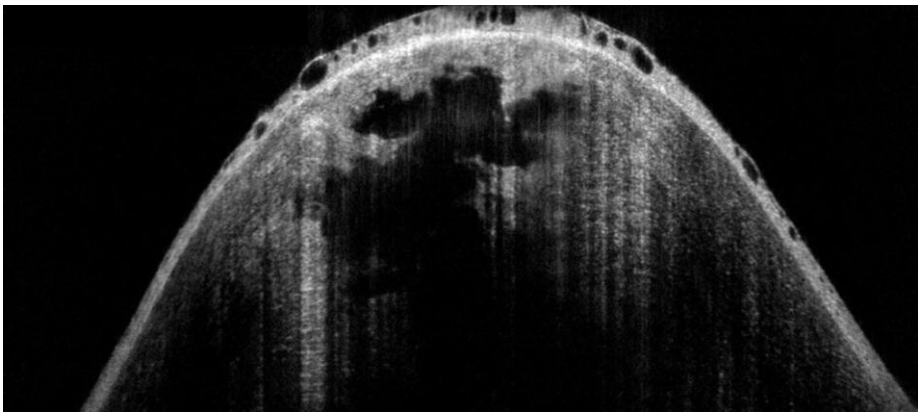

**Figure 4: Anterior segment- optical coherence tomography imaging of right eye showing epithelial and stromal corneal edema with Descemet membrane tear and presence of intrastromal fluid clefts.**

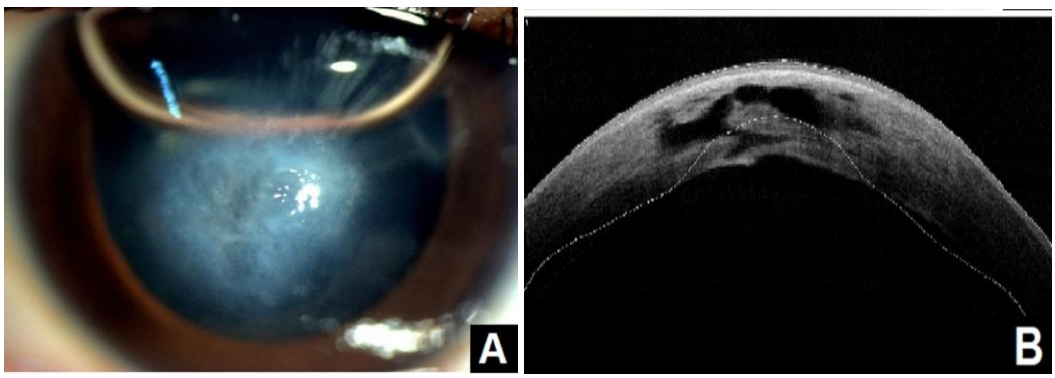


**Figure 5: (A) Slit-lamp photograph of the anterior segment of the right eye showing a reduction in corneal edema with gas bubble occupying less than one-third of the anterior chamber (B) Anterior segment- optical coherence tomography imaging of right eye showing reattached Descemet membrane at 1 week follow-up.**

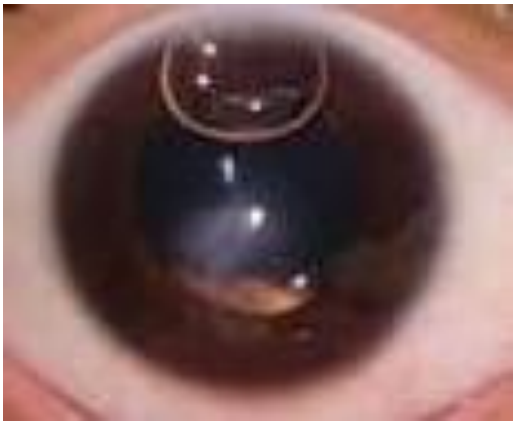

**Figure 6: Slit-lamp photograph of the anterior segment of the right eye showing corneal nebular grade scar at the centre with rest of the cornea and otherwise normal anterior segment at 3 weeks follow-up.**

## DISCUSSION

Keratoconus is generally considered a bilateral condition, though the presentation may be markedly asymmetric. It may take years after the initial diagnosis of keratoconus in one eye for the condition to become apparent in the fellow eye.[15] In our case, one eye had advanced keratoconus with acute hydrops, and the contralateral eye had borderline parameters on Pentacam.

Acute corneal hydrops is the sudden onset of gross corneal edema in an eye with keratoconus, mostly seen in the second or third decade.[1,16] Several factors have been reported to be associated with a higher predisposition for corneal hydrops, including younger age, male gender, vernal keratoconjunctivitis, and eccentric (rather than central) cone location.[1,17] In the present case, the patient was a 22-year-old male without any history of repeated episodes of itching or rubbing of eyes. Also, the location of the cone was central. Fan Gaskin et al., in their study, found that 41% and 25% of patients with acute corneal hydrops did not have a history of atopy and eye rubbing, respectively.[10]

In our case, acute corneal hydrops was the initial presentation of keratoconus. Multiple case studies have reported that keratoconus may be first diagnosed at the presentation of an acute corneal hydrops.[10-14] Cheung et al. reported a case of keratoconus in which the diagnosis was made only when the patient developed corneal hydrops at the age of 21 years.[11] Corneal hydrops may be misdiagnosed as infectious keratitis or other corneal pathology, and keratoconus may be overlooked, especially when there is no history of itching or rubbing of eyes. A late diagnosis may predispose to severe complications, including corneal perforation, microbial keratitis, and glaucoma.[8,9] Hence, it is worth pointing out that a detailed examination of both eyes, including refraction and evaluation of contralateral eye using investigative modalities like Pentacam, is crucial in such unusual presentations, especially in young adults and children, to avoid misdiagnosis. It is of note that other environmental or genetic factors may have played a role in the development and progression of keratoconus in otherwise healthy individuals.[18]

A wide range of medical and surgical options are available to treat acute corneal hydrops. Medical therapy includes topical hyperosmotics (help draw fluid), antibiotics (prevent secondary infection), cycloplegics (reduce pain and photophobia), steroids (reduce inflammation and subsequent neovascularization) , preservative-free lubricants, and anti‑glaucoma medications (lessen the hydrodynamic force on the posterior cornea).[1,3] Other conservative options include a bandage contact lens (relieve pain) and oral non-steroidal anti-inflammatory drugs.[1,3] Surgical interventions include injection of air/gas, sulfur hexafluoride ($SF_6$) or $C_3F_8$, into the anterior chamber, which shorten the recovery period and prevent aqueous penetration into the stroma by tamponade effect and also by unrolling the torn ends of ruptured Descemet's membrane.[19-21] In the present case, the choice of $C_3F_8$ for Descemetopexy was made as it is the longest acting (6 weeks) among all, and usually repeat injections are not needed. Air remains for a short time, and $SF_6$ though, is long-acting (2 weeks) compared to air, repeat injections may still be required. Possible intraoperative complications are elevation of IOP due to pupillary block, inadvertent intrastromal gas injection, infection, and endothelial damage.[5] Other surgical options that are useful in special situations include compressive sutures along with gas injection, cyanoacrylate tissue adhesive with bandage contact lens, AS-OCT guided intrastromal fluid drainage with air tamponade, penetrating keratoplasty, and amniotic membrane transplantation with cauterization.[1,5,22-24] The hydrops may leave a residual scar once it heals. The patient may require a contact lens or keratoplasty for visual rehabilitation, subsequently.

In conclusion, unilateral acute corneal hydrops may be the first presenting feature of undiagnosed keratoconus and should always be considered in the differential diagnosis of corneal leucoma, especially in young adults and children. The contralateral eye in such a case, needs close monitoring and management at the appropriate time. Every practitioner should strive to detect keratoconus early with the latest diagnostic imaging devices and manage subsequently to avoid devastating complications like hydrops. Treatment of acute corneal hydrops by descemetopexy using $C_3F_8$ is an effective and safe procedure for rapid recovery.

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