Successful Management of Corneal Hydrops and Intrastromal Corneal Ring Segment (ICRS) Migration Following ICRS Implantation for Keratoconus

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Patient: Male, 17-year-old
Final Diagnosis: Acute corneal hydrops • intrastromal corneal ring segment migration
Symptoms: Poor visual acuity • rigid contact lens intolerance
Medication: —
Clinical Procedure: Intrastromal corneal ring segment implantation • intrastromal corneal ring segment reimplantation
Specialty: Ophthalmology

Objective: Unusual or unexpected effect of treatment
Background: Acute corneal hydrops refers to a rare complication of keratoconus and other ectatic disorders with potentially sight-threatening sequelae. Intrastromal corneal ring segment (ICRS) implantation is a surgical procedure performed as therapy for keratoconus when there is contact lens intolerance. ICRS migration along the tunnel made for its insertion is one of its most frequent complications. We believe this is the first published case of acute corneal hydrops and ICRS migration unfolding shortly after an uneventful ICRS implantation, and successfully managed with medical treatment and ICRS reimplantation, respectively.

Case Report: A 17-year-old male, previously diagnosed with bilateral keratoconus 4 years earlier for which he underwent cross-linking surgery for both eyes 3 years prior, presented to our department for a first-time keratoconus assessment. His best-corrected visual acuity (BCVA) was 20/25 OD and 20/40 OS with rigid gas-permeable contact lenses. Due to contact lens intolerance in the OS and lack of custom-fit scleral lenses at the time, 2 ring segments (Ferrara, AJL Ophthalmic, Inc.) with an arc length of 160 degrees and 300 μm in size were implanted at a depth of 320 μm. Within 1 week, severe hydrops and ICRS migration emerged. Medical treatment provided resolution of hydrops and then ICRS reimplantation was performed without further complications, resulting in contact lens tolerance alongside a BCVA of 20/40 OS.

Conclusions: Corneal hydrops is a challenging complication in ICRS implantation due to its infrequent occurrence and the severity of its potential sequelae. Prompt medical treatment and close follow-up are essential to obtain the best outcome in these instances.

Keywords: Corneal Edema • Corneal Topography • Keratoconus • Prostheses and Implants • Refractive Errors • Refractive Surgical Procedures

Abbreviations: BCVA – best-corrected visual acuity; D – diopters; ICRS – intrastromal corneal ring segment; OD – right eye; OS – left eye; OU – both eyes; RGP – rigid gas-permeable; UCVA – uncorrected visual acuity

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Background

Acute corneal hydrops refers to an abrupt corneal edema, a rare complication of keratoconus and other ectatic disorders with potentially sight-threatening sequelae [1]. It can occur at any given time, but is mainly seen in advanced stages of keratoconus [2,3]. Corneal hydrops is thought to be elicited by Descemet membrane and endothelial rupture, causing infiltration of aqueous humor into the corneal stroma and for it to lose its clarity, thus decreasing visual acuity [1]. Topical hypertonic saline (5%) and steroids are the mainstays of medical treatment for hydrops, but with little evidence to support their actual effectiveness [1,4].

Intrastromal corneal ring segment (ICRS) implantation is a surgical procedure performed as a therapeutic for keratoconus when there is contact lens intolerance. This surgical technique consists of the placement of polymethyl methacrylate (acrylic) devices that act as a spacer for the corneal stroma, yielding a flattening of the conus [5]. It is a safe and well-tolerated procedure, but it is not exempt of complications [6]. ICRS migration along the tunnel made for its insertion constitutes one of its most frequent complications [5]. Despite this, it remains a safe procedure owing to its reversibility and good results [6].

To our knowledge, only 2 cases of corneal hydrops taking place during or briefly after ICRS implantation have been described in the literature: both failed to recover under medical therapy, ultimately requiring penetrating keratoplasty, and there was no concomitant ICRS migration present [7,8].

We report a first case of acute corneal hydrops and ICRS migration unfolding shortly after an uneventful ICRS implantation, and successfully managed with medical treatment and ICRS reimplantation, respectively.

Case Report

A 17-year-old male, previously diagnosed with bilateral keratoconus 4 years earlier for which he underwent cross-linking surgery for both eyes (OU) 3 years prior, presented with no particular concern to our department for a first-time keratoconus assessment. His last ophthalmic evaluation was after the cross-linking was performed. He had a history of atopy, but was otherwise healthy. His uncorrected visual acuity (UCVA) was counting fingers at 4 meters in the right eye (OD) and 2 meters in the left eye (OS). The best spectacle-corrected visual acuity was 20/50 OD and 20/200 OS with a manifest...
refraction of -10.00D sph -3.25D cyl ax 35º OD and -17.00D sph OS. The best-corrected visual acuity (BCVA) was 20/25 OD and 20/40 OS with rigid gas-permeable (RGP) contact lenses, but contact lens intolerance presented in the OS, and due to regional limitations, it was not possible to provide custom-made scleral contact lenses at the time.

Examination of the anterior segment in the OD revealed an incomplete Fleischer ring, and Vogt striae were observed next to a Fleischer ring in the OS. Both corneas were transparent. Fundus examination was unremarkable in OU and intraocular pressure was normal in OU.

Preoperative corneal topography (Orbscan, Orbtek Bausch & Lomb, Inc.) of OD (Figure 1) and OS (Figure 2) revealed points of maximum elevation in line with keratoconus at the posterior float (OD 144 μm, OS 153 μm) and anterior float (OD 65 μm, OS 59 μm), an irregular keratometric map, and a central corneal thinning (OD 440 μm, OS 395 μm).

On account of RGP contact lens intolerance in the OS and lack of custom-fit scleral lenses, ICRS implantation was agreed upon to provide better contact lens tolerance. The implantation of 2 ring segments (Ferrara, AJL Ophthalmic, Inc) with an arc length of 160 degrees and 300 μm in size employing manual dissection technique was decided on.

An initial procedure using topical anesthesia was performed in the first day of month 1. It began as the corresponding corneal markings were placed, followed by an incision with a diamond knife on the 95-degree meridian, the most curved meridian of the cornea, and at a depth of 320 μm (80% of the corneal thickness) in the thinnest zone at the implantation site (400 μm). The implantation site, meridian, and depth were based on a single, preoperative corneal topography obtained with the Orbscan. Tunnel creation within the corneal stroma using semicircular dissectors proceeded, the ICRS were implanted, and the incision was sutured with 10-0 nylon. Once completed, an attempt to gently express aqueous humor from the cornea to check for inadvertent perforation into the anterior chamber was made, but was unsuccessful and highlighted an unscathed microscopic corneal morphology. Lastly, a drop of gatifloxacin and prednisolone were administered, and thus concluded the procedure without any setback.

The day after surgery, our patient presented to follow-up with mild corneal hydrops and unaltered ICRS. Topical gatifloxacin every 6 h for 10 days and topical prednisolone in a tapering schedule (12, 8, 6, and 4 drops, each for 5 days) for 20 days.
were continued. Within 1 week, corneal hydrops was significantly exacerbated, and ICRS migration had emerged: both segments shifted inferiorly and the nasal segment sat above the temporal segment (Figure 3). Intraocular pressure was normal and remained normal throughout the case, with no further findings on examination. Here, topical sodium chloride (5%) was added every 6 h for 8 weeks. A follow-up 15 days later revealed no change in the condition.

By month 2, 7 weeks having elapsed since the onset of severe hydrops, the corneal hydrops resolved, as a transparent and otherwise unaffected cornea was now present during inspection. After careful consideration, ICRS reimplantation was decided on in month 3. Preexisting tunnels were accessed to reimplant the ICRS with the hook; during ICRS manipulation, the nasal segment was easily movable, whereas the temporal segment exhibited some adhesion. Once in position, 2 additional 10-0 nylon sutures were placed inferiorly to keep both ICRS in place (Figure 4).

The following day, UCVA was 20/200 in OU, and the ICRS were securely fastened. Our patient returned to us in month 4 with a concern of foreign body sensation coming from the upper incision, so the upper suture was removed. This case concluded in month 6, as the 2 lower sutures were also removed. A final follow-up was performed nearing the end of the month with the ICRS anchored and our patient achieving tolerance to RGP contact lenses in the OS, alongside a BCVA of 20/50 OD and 20/40 OS.

We offer the following supplement to orient the reader to the events that took place in this case (Figure 5).

**Discussion**

Keratoconus is a non-inflammatory ectatic disorder that consists of a progressive steepening and thinning of the cornea, producing its characteristically conical shape. These changes
in corneal morphology eventually lead to visual impairment: mainly myopia and astigmatism [2]. Although uncommon, this disorder can even lead to irreversible blindness [9]. The onset of keratoconus is usually during puberty or early adulthood, with a higher rate of progression the earlier the onset. Its etiology is complex but it has strong associations with atopy, eczema, eye rubbing, and, especially, genetics [10]. Hydrops in keratoconus, even if rare, can happen at any time, particularly in advanced stages of the disease [3]. Spontaneous hydrops has a reported incidence of 2.8-3.15% in patients with keratoconus [11].

Two previous reports detail the occurrence of hydrops during ICRS implantation using the manual dissection technique for keratoconus [7] and subsequent to femtosecond laser-assisted implantation for pellucid marginal degeneration [8], but recovery was not achieved with conservative or medical management. Thereafter, both cases required penetrating keratoplasty as definitive management for corneal hydrops [7,8], whereas our patient experienced recovery nearly within 8 weeks of hydrops first appearing, resorting to topical gatifloxacin every 6 h for 10 days and topical prednisolone in a tapering schedule (12, 8, 6, and 4 drops, each for 5 days) for 20 days at the beginning of the hydrops. Treatment was continued by adding topical sodium chloride (5%) every 6 h for the next 8 weeks when hydrops aggravated 1 week later after its onset to mitigate it.

There is a vast array of treatment options for corneal hydrops, ranging from conservative to surgical procedures aimed to alleviate this ectatic complication. Medical treatment often comprises topical hypertonic saline to reduce edema and topical corticosteroids to prevent neovascularization and reduce inflammation, but, despite widespread use for acute hydrops, evidence of its benefit is limited [1,4]. The efficacy of topical hypertonic saline in hydrops seems to depend on the underlying condition causing it and its severity, being provably useful in cases of mild hydrops [12]. Further studies regarding the effectiveness of topical hypertonic saline and steroids as therapy for hydrops are required to determine whether these treatments are actually useful or not.

In this case, neither ICRS explantation nor any other surgical intervention for hydrops was necessary. Furthermore, ICRS migration presented along with worsening of corneal hydrops. We believe that the intrastromal structure swelling up with aqueous humor after the corneal hydrops had exacerbated may have precipitated the ICRS migration. Once the former completely subsided, and the corneal stroma returned to its normal configuration, ICRS reimplantation was secured by positioning 2 sutures inferiorly besides the superior main incision suture without any additional migration. This approach resulted in an outstanding outcome for our patient, in terms of tolerance to the RGP contact lenses and as such, a substantial improvement in BCVA.

Our main limitations were the reliance on a single preoperative Orbscan pachymetry during this case, and the lack of accurate readings with ultrasound pachymetry, intraoperative measurements, and of optical coherence tomography after hydrops established. During the procedure, there was no noticeable corneal perforation or change in its microscopic morphology, and we were unable to express aqueous humor out of the cornea using gentle pressure.

It is presumed that a rupture in the corneal endothelium and Descemet membrane allows for an inflow of aqueous humor into the corneal stroma, begetting corneal hydrops [13]. Current theory suggests that during the course of keratoconus, the cornea is subject to stretching, and in hand with Descemet membrane disruption, develops intrastromal fluid-filled clefts [14]. Nevertheless, as Parker et al reported, a Descemet membrane disruption, even a total descemetorhexis in keratoconic eyes, is not enough to induce an acute hydrops, but it requires a serious defect in the Descemet membrane in addition to the posterior stroma to give rise to corneal hydrops [14].

Due to the restrictions this case was subjected to, it is impossible to determine the exact cause, but we believe that a non-optimal ICRS implantation site or stress due to manual tunnel dissection technique or during suturing may have led to an inadvertent perforation of the endothelium, Descemet membrane and Dua layer with the anterior corneal stroma remaining intact, as no deflation of the anterior chamber was present and aqueous humor was unable to be expressed.

Despite the limitations involved in this case, these do not change the outcome: penetrating keratoplasty was avoided and corneal hydrops was managed exclusively with medical treatment. Regarding the concomitant ICRS migration, an ICRS reimplantation without further complications was performed, which resulted in the initially desired improvements in contact lens tolerance and visual acuity. This case is of great relevance as it describes a previously unseen scenario of concurrent corneal hydrops and ICRS migration, along with its successful management, providing a valuable experience in the treatment of these complications, especially in this unique scenario.

**Conclusions**

Corneal hydrops is a challenging complication in ICRS implantation due to its infrequent occurrence and the severity of its potential sequelae. Being aware of this possible complication during or after surgery may provide greater foresight, and knowing the clinical course and successful management fortunately reached here should give the surgeon a sense of preparedness and security dealing with these unusual yet serious complications.
To our knowledge, this is the first case report of the resolution of corneal hydrops in the setting of ICRS implantation without the need for surgical intervention, alongside ICRS migration and successful reimplantation, and with the expected contact lens tolerance and an excellent refractive outcome.

We emphasize that prompt medical treatment and close follow-up are essential to obtain the best outcome in these instances. It is challenging to identify the precise cause of this event: whether it was a spontaneous rupture of the Descemet membrane and other posterior corneal layers or a surgically-induced disruption of these posterior corneal structures is unknown, but its scarcity in the literature, with only a few reports, still attests to the safety offered by ICRS implantation.

Declaration of Figures’ Authenticity

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