

PEER REVIEWED

# Management of Acute Corneal Hydrops in a Patient with Keratoconus

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## Abstract

*This teaching case report is significant because there is no standard-of-care treatment for corneal hydrops. Both optometrists and ophthalmologists treat patients with this condition, either conservatively or surgically. With the advent of new technology, diagnosing keratoconus has increased, and procedures such as cross-linking are potentially lowering the incidence of corneal hydrops. However, some patients may not be able to receive such treatments, and the potential for hydrops remains an issue. Many providers of primary eye care do not feel comfortable diagnosing and/or managing corneal hydrops. This review outlines various management protocols, including conservative and surgical interventions, as well as protocol outcomes.*

**Key Words:** *keratoconus, hydrops, conservative treatment, surgical intervention*

## Background

Keratoconus is typically thought to be a bilateral disease that can present asymmetrically. It is associated with progressive corneal ectasia and scarring. With no definitive etiology, the corneal ectasia ultimately leads to irregular astigmatism, central anterior scarring and reduced vision.<sup>1</sup> Hydrops is a rare condition experienced by some keratoconic patients. Hydrops is characterized as a break in Descemet's membrane and underlying endothelium. This allows aqueous humor to leak into the stroma and cause stromal edema.<sup>2</sup>

## Student Discussion Guide

A 49-year-old Indian male presented for examination in a primary eyecare clinic with complaints of light sensitivity, mild ocular redness and a visible white spot in the left eye.

The History of Present Illness included a burning sensation OS with "foggy" vision, but the patient denied any discharge. The patient reported that the burning sensation started approximately one week prior to his visit. He had been self-medicating with Systane Lubricant Eye Drops (Alcon) three times a day in both eyes and Zaditor (ketotifen fumarate, Novartis) twice a day in both eyes, with mild relief. He denied any similar incidences in the past. He had no complaints at the time of his last vision and ocular health examination, which was nine days earlier with a different provider in the same clinic. At that visit, his best-corrected visual acuity was 20/20 OD and 20/60-2 OS (pinhole: no improvement).

The patient's Past Ocular History included keratoconus (oval cone in both eyes, OS larger than OD) with long-standing scarring of the left eye, dry eye syndrome, chronic allergic conjunctivitis and presbyopia. He was unsure when he was diagnosed with keratoconus but recalled his vision becoming poor in his early 20s. Piggyback contact lenses were self-discontinued approximately two years prior due to poor comfort. The patient denied any past ocular surgery or ocular trauma. His Past Medical History was

unremarkable, with no use of systemic medications. His Family History was significant only for keratoconus (brother). His Social History was negative for tobacco, alcohol or recreational drug use. He had no known allergies or drug allergies. He was oriented to time, place and person, and his mood was appropriate.

*Examination findings*

Entrance visual acuity (with spectacles):

OD: 20/20

OS: 20/400 (pinhole: no improvement)

Manifest subjective refraction:

OD: +1.75 -4.50 x 055 20/20

OS: +0.75 -4.50 x 120 20/400 ADD: +1.50 20/20 OU at 40 cm

Pupils were equal, round and reactive to light (with no afferent pupillary defect). Confrontation visual fields were full to static fingers in both eyes. Extraocular muscle motility was full in all gazes.

Slit lamp biomicroscopy:

Lids/lashes: few capped meibomian glands OU

Palpebral conjunctiva: trace papillae OU

Bulbar conjunctiva: trace injection OU

Tears: thin lacrimal lake OU

Cornea:

OD: moderate inferior thinning with no scarring; Fleischer's ring and striae present

OS: moderate inferior thinning; 6-mm area of dense and diffuse stromal edema with an underlying break in Descemet's membrane slightly below the visual axis

Iris: flat OU

Lens: clear OU

Anterior chamber: grade IV angle and free of visible aqueous cells or flare OU

Goldmann applanation tonometry:

OD: 11 mmHg

OS: 11 mmHg (with irregular mires)

@ 10:30 a.m.

The appearance of the cornea in the left eye and timing suggested a diagnosis of corneal hydrops. A 6-mm area of stromal edema with an underlying break in Descemet's membrane was present. There were

no obvious epithelial defects. Subjective complaints of blurry vision and a burning sensation were reported. No anterior chamber reaction or discharge were seen. The patient was started on Muro 128 ophthalmic solution (sodium chloride hypertonic solution 5%, Bausch + Lomb) three times a day along with Muro 128 ointment (sodium chloride hypertonic ointment 5%, Bausch + Lomb) at bedtime. He was also educated to continue using artificial tears three times a day in both eyes. Follow-up was scheduled for five days in an anterior segment specialty unit.

#### *Follow-up visit #1*

The patient returned as scheduled for his follow up. He reported good compliance with the prescribed treatment regimen, but no change in his symptoms. The patient's spectacle-corrected distance visual acuity remained stable at 20/20 OD and 20/400 OS (pinhole: no improvement). Slit lamp biomicroscopy revealed no changes in the right eye. Evaluation of the left cornea showed a 5-mm area of dense and diffuse stromal edema with an underlying break in Descemet's membrane slightly below the visual axis. There were no evident epithelial defects. Intraocular pressure (IOP) measured with Goldmann applanation tonometry at 2:15 p.m. was 12 mmHg OD and OS (with irregular mires OS). The diagnosis of corneal hydrops was confirmed.

The patient was instructed to continue using Muro 128 ophthalmic solution 5% three times a day and Muro 128 ointment at bedtime in the left eye. The patient was extensively educated on his condition and possible long recovery time (average of three months). The option of a surgical consult vs. the topical medications was discussed. The patient elected to continue the topical/medical therapy, and a follow-up appointment was scheduled for one month.

#### *Follow-up visit #2*

The patient returned as scheduled for his follow-up appointment. He reported that the burning sensation had subsided, but that he occasionally felt his left eyelid "hitting something on the eye." He reported a new symptom of glare in bright sunlight. He had been using Muro 128 ophthalmic solution 5% three times a day and Muro 128 ointment at bedtime as instructed. The patient also reported that he felt a "gritty sensation" when waking up in the morning several days prior to this visit. His spectacle-corrected visual acuity was 20/20 OD and 20/400 OS (pinhole: 20/200 OS). Slit lamp biomicroscopy evaluation of the left cornea showed a 3-mm area of diffuse stromal edema with an underlying break in Descemet's membrane slightly below the visual axis. There were no evident epithelial defects. IOP measured at 1:30 p.m. with Goldmann applanation tonometry was 12 mmHg OD and OS (with irregular mires OS).

Because the patient reported having an ocular gritty sensation in the mornings, he was told to discontinue Muro 128 ointment and to use Soothe Lubricant Eye Ointment (Bausch + Lomb) instead. To help with the glare sensitivity he reported, he was started on Lotemax Gel 0.5% (loteprednol etabonate, Bausch + Lomb) four times a day in the left eye and advised to wear full-rimmed sunglasses when outdoors to block more sunlight. A follow-up visit was scheduled for one month.

#### *Follow-up visit #3*

The patient returned as scheduled for his follow-up visit. He reported having persistent glare but obtaining prescription sunglasses, which helped. He reported that he discontinued Lotemax because he experienced mild sensitivity after using it. He said he was using the Soothe ointment two to three times per day and at night. His spectacle-corrected distance visual acuity was 20/20 OD and 20/200 OS (pinhole: 20/100 OS). Slit lamp evaluation of the left eye revealed a 3-mm area of mild corneal stromal edema with early scarring below the visual axis. No break in Descemet's membrane was visible, and no sodium fluorescein staining was seen. IOP measured at 2:15 p.m. with Goldmann applanation tonometry was 13 mmHg OD and OS (with irregular mires OS).

The patient was instructed to discontinue Lotemax Gel 0.5%, and to use only Soothe Lubricant Eye Ointment in the left eye throughout the day and at night. A follow-up appointment was scheduled for one month. The patient was informed that at the next visit, scleral contact lenses would be used to determine his best-corrected visual acuity.

#### *Follow-up visit #4*

The patient returned as scheduled for his follow-up visit. He reported improvement in symptoms and that he had been using the Soothe ointment only as needed. His spectacle-correct distance visual acuity was 20/20 OD and 20/200 OS (pinhole: no improvement). Slit lamp biomicroscopy revealed no changes in the right eye. A 3-mm area of stromal scarring below the visual axis was present in the cornea of the left eye. There were no signs of sodium fluorescein staining. IOP measured with Goldmann applanation tonometry at 1:00 p.m. was 13 mmHg OD and OS (with irregular mires OS). An undilated 90D fundus examination revealed normal posterior segments (to the extent seen) that were noncontributory to this case. The cup-to-disc ratio was 0.45 vertically in each eye; the artery-to-vein ratio was 2:3 in each eye; and both maculas were clear with a positive foveal reflex in each eye.



**Table 1.** Prescription for Custom Stable scleral contact lenses (Valley Contax) to determine the patient's best-corrected visual acuity. [Click to enlarge](#)

Custom Stable scleral contact lenses (Valley Contax) were fitted for both eyes to determine whether vision could be improved. Lens parameters and resulting vision are noted in **Table 1**. Despite the improved visual acuity provided by the scleral contact lenses in each eye, the patient declined to complete the fitting process due to the cost. In the meantime, he was educated about continuing to use artificial tears and ointment as needed. A follow-up appointment, to include a dilated fundus exam, was scheduled for four to six weeks.

The patient did not return for the scheduled follow-up visit but did return for a comprehensive examination a year later in the primary eyecare clinic. At that time he had no complaints and reported he was happy in his glasses and his vision in the left eye had improved over time.

#### *Examination findings*

Visual acuity (with spectacles):

OD: 20/20

OS: 20/50<sup>2</sup> (pinhole: no improvement)

Manifest subjective refraction:

OD: +2.00 -4.50 x 055 20/20

OS: +0.75 -4.50 x 120 20/50<sup>1</sup> ADD: +2.00 20/20 OU at 40 cm

Pupils were equal, round and reactive to light (with no afferent pupillary defect). Confrontation visual fields were full to static fingers in both eyes. Extraocular muscle motility was full in all gazes.

Slit lamp biomicroscopy:

Lids/lashes: few capped meibomian glands OU

Palpebral conjunctiva: trace papillae OU

Bulbar conjunctiva: trace injection OU

Tears: thin lacrimal lake OU

Cornea:

OD: moderate inferior thinning with no scarring; Fleischer's ring and striae present

OS: moderate inferior thinning; Fleischer's ring present; 4-mm area of stromal and endothelial scarring slightly below the visual axis

Iris: flat OU

Lens: grade 1 nuclear sclerosis OU

Anterior chamber: grade IV angle and free of visible aqueous cells or flare OU

Vitreous: syneresis OU

Goldmann applanation tonometry:

OD: 11 mmHg

OS: 11 mmHg (with irregular mires)

@10:30 a.m.

A dilated fundus examination revealed normal posterior segments (to the extent seen) that were noncontributory to this case. The cup-to-disc ratio was 0.45 vertically in each eye; the artery-to-vein ratio was 2:3 in each eye; the maculas were clear with a positive foveal reflex in each eye; and the retina was flat and intact with no breaks, tears or detachments in each eye.

Despite previous improvement in visual acuity with scleral contact lenses, the patient declined a new fitting due to having subjectively adequate vision with his current glasses. He was educated to continue utilizing artificial tears and ointment as needed, and a return visit was scheduled for one year.

### **Educational Guidelines**

This teaching case report can serve optometry students who have completed, or are completing, an anterior segment/ocular disease course and a specialty contact lens course, as well as optometrists in clinical practice.

#### *Key concepts*

1. The pathophysiology of corneal hydrops and its impact on the cornea and keratoconus
2. Critical thinking in the diagnosis, treatment and management of corneal hydrops
3. Ensuring that patients understand all treatment options and the anticipated time to resolution, and have realistic expectations after resolution
4. The use of specialty contact lenses following corneal hydrops

#### *Learning objectives*

1. At the conclusion of this case discussion, participants should be able to:
2. Understand corneal hydrops from an anatomical standpoint
3. Differentiate corneal hydrops from other corneal conditions
4. Understand the typical patient demographic for corneal hydrops
5. Understand the risk factors in association with the clinical presentation to best manage the patient, including if/when referral is appropriate
6. Provide patient education regarding all management options and expectations for those options

*Discussion questions*

1. Knowledge and concepts required for critical review of the case:
  - a. What are the typical clinical characteristics of corneal hydrops?
  - b. How can one distinguish corneal hydrops from other corneal conditions?
  - c. What would be the most appropriate management given the case provided?
2. Differential diagnosis:
  - a. What differential diagnoses make the most sense given the clinical characteristics?
  - b. What other factors need to be considered in this case?
  - c. Are there any ancillary tests that would have been helpful in this diagnosis?
3. Disease management:
  - a. How would you monitor this patient, if at all?
  - b. What topical therapy would you prescribe, if at all?
  - c. How would you determine whether you were going to manage this patient with conservative treatment only or refer for surgery?
  - d. What timeline is most appropriate for this patient?
4. Patient education:
  - a. How would you educate the patient regarding this diagnosis?
  - b. What is the long-term prognosis for this patient?
  - c. How would you discuss visual outcomes with conservative and surgical treatments?
5. Critical thinking:
  - a. How would you have managed this case?
  - b. Do you feel more prepared to manage or co-manage this condition?

*Assessment of learning objectives*

1. Case-based discussion of diagnosis and management of corneal hydrops
2. Clinical-thinking skills and knowledge of the clinical signs of corneal hydrops can be assessed in a

- small-group setting (e.g., seminar, Grand Rounds) and assessed as part of a written exam
3. Students should be evaluated on their knowledge of different corneal hydrops therapies and be able to create a treatment plan
  4. Practical assessment of clinical tests that are commonly used in the diagnosis of corneal hydrops, e.g., anterior segment optical coherence tomography (OCT) and corneal topography
  5. Written communications can be assessed by writing an information brochure for patients on corneal hydrops therapy
  6. Literature review on evidenced-based corneal hydrops management could be written up as a dissertation

## Discussion

Previously published data have shown that the estimated prevalence of keratoconus in the general population is 54 per 100,000.<sup>1</sup> A more recently published nationwide registration study in the Netherlands estimated the prevalence of keratoconus in the general population as 265 per 100,000.<sup>22</sup> Although the etiology of keratoconus is still not clear, it is believed that genetics, the environment (eye-rubbing, allergies) and the individual's endocrine system all play a role in the onset, progression and stabilization of the condition.<sup>6</sup>

The first case of corneal hydrops in the setting of keratoconus was reported by Plaut in 1900. It was described as a sudden opacity at the apex of the cornea due to a rupture of Descemet's membrane, which was later confirmed by Axenfeld in 1906.<sup>2</sup> Corneal hydrops occurs in 2.5-3.0% of the population with keratoconus. The majority of cases are unilateral, occur more frequently in males than in females, and typically present in the second or third decade of life.<sup>2</sup>

### *Differential diagnosis*

The differential diagnoses for this patient included:

- Corneal ulcer, depending on size and location, involves different modalities of treatment. The usual presentation includes ocular redness, pain and light sensitivity. A hypopyon and/or anterior chamber reaction may also be seen. An epithelial defect with positive sodium fluorescein staining is observed. Depending on location, vision may or may not be affected.<sup>3</sup>
- Corneal hydrops patients may present with acute light sensitivity, pain (depending on patient's pain threshold), and a well-demarcated area of edema associated with a break in Descemet's membrane. An epithelial defect is not usually seen. Vision is decreased, but location determines how variable the visual acuity will be.<sup>4</sup>
- Corneal degenerations are changes in various layers of the cornea that tend to be unilateral, asymmetric and peripherally located. They are frequently associated with systemic conditions, and the changes that tend to occur include deposition, thinning and/or vascularization of the corneal tissue.<sup>5</sup>
- Corneal dystrophies are changes that usually occur in a single layer of the cornea. The changes tend to be bilateral and centrally located. They are usually not associated with systemic conditions but are inherited. Onset tends to be at a younger age compared to degenerations.<sup>5</sup>
- Corneal scarring may occur secondary to corneal trauma or infection.

In this case report, the appearance of the cornea in the left eye and timing suggested a diagnosis of corneal hydrops. A 6-mm area of stromal edema and an underlying break in Descemet's membrane were present. There were no obvious epithelial defects. Subjective complaints of blurry vision and a burning sensation were reported. No anterior chamber reaction or discharge was seen.

Acute corneal hydrops is characterized as a break in Descemet's membrane (and underlying

endothelium) allowing aqueous humor to leak into the stroma and cause stromal edema. The breakage is spontaneous and most likely to occur in eyes with advanced thinning. Symptoms may include sudden onset decreased vision and irritation including pain and/or photophobia. Clinical signs include an observable break in Descemet's membrane, anterior and posterior stromal edema with possible epithelial involvement, and hyperemia of the conjunctiva.<sup>4</sup> The patient may also notice a visible "white spot" on their eye. Although usually associated with keratoconus, hydrops can also occur in other ectatic disorders such as pellucid marginal degeneration, keratoglobus and ectasia status post-LASIK, radial keratotomy or penetrating keratoplasty.<sup>2</sup> Complete resolution of signs and symptoms typically takes three to six months.

One of the proposed etiologies of keratoconus is eye-rubbing. There are several theories as to how rubbing causes keratoconus including increased concentrations of inflammatory mediators in the pre-corneal tears, large intraocular pressure spikes, a reduction in shear strength, and cone-forming deformation.<sup>23</sup> The corneal thinning along with some sort of corneal trauma (e.g., rubbing of the eyes) may be considered as an underlying cause of hydrops.<sup>2,4</sup> A 2013 study conducted in New Zealand showed that hydrops typically developed approximately four years after the diagnosis of keratoconus, and the subjects with hydrops were more likely to have a history of eye-rubbing (but less likely to have a family history of keratoconus). There was also no statistically significant differences in the prevalence of atopic disease or contact lens wear between the keratoconus with hydrops and keratoconus without hydrops groups.<sup>7</sup> It is thought that race does not play a role, but a population-based study done in the United Kingdom showed that the proportion of South Asian and black patients with acute corneal hydrops was significantly higher than in the general population.<sup>8</sup> Although theirs was a small study, Grewal and Laibson reported 21 of 22 (95%) eyes with hydrops seen by a referral cornea service during a 2.5-year period had seasonal allergies, 20 of 22 (91%) eyes had allergy-associated eye-rubbing behavior, and six patients were able to identify a traumatic inciting event (vigorous eye-rubbing in four and traumatic contact lens insertion in two).<sup>9</sup>

Muro 128 was a topical therapy used for the patient in this case report. Muro 128 is designed to draw out excess fluid from the cornea and reduce swelling. It is available as an eye drop (2% or 5%) or an ointment (5%) that is traditionally used at bedtime. The 2% ophthalmic solution is conventionally used for less severe cases. The 5% solution was chosen by the previous provider given the degree of edema. There are mixed opinions about the use and efficacy of Muro 128. A common misconception is that if a keratoconic patient presents with corneal hydrops, topical therapy with Muro 128 ophthalmic solution is required to resolve the edema. If epithelial edema is present, there would be some validity in its use. The hypertonicity allows for an osmotic gradient to occur, allowing fluid to be drawn out of the corneal layers above Bowman's membrane. If the edema is localized in the stroma, Muro 128 drops will have less of an effect. For stromal edema, the endothelial pumps are the main mechanism for drawing out fluid.<sup>10</sup> Nonetheless, some clinicians use a hypertonic solution to resolve any corneal edema, and reports of efficacy are mixed. In retrospect, I would have not prescribed Muro 128 because the patient did not have any frank epithelial edema.

Patients commonly complain of irritation with Muro 128, as did this patient ("gritty sensation"). Therefore, it is important to make an informed decision about prescribing it based on its tolerability and efficacy. A sensation of burning and stinging (or in this case a "gritty sensation") can be attributed to the hypertonic solution creating an unstable tear film. Hyperosmolar levels in the tear film can result in corneal inflammation and trigger sensory neurons.<sup>11</sup> Given the patient's complaints while using the drop, it was determined best to discontinue.

For patients who present without evidence of epithelial compromise, prescribing a topical nonsteroidal anti-inflammatory or steroid drop three-to-four times per day may provide relief from pain and inflammation. A topical cycloplegic could be prescribed in lieu of, or together with, a steroid to reduce pain and reduce the possibility of a secondary anterior chamber reaction. One may opt not to prescribe a

steroid as it may slow corneal healing and lead to a rare corneal perforation. However, in the later stages of healing, when edema has been reduced, a steroid may aid in decreasing scarring.<sup>12</sup> After a trial of Muro 128, a steroid was prescribed for this patient due to his complaints of glare and sensitivity, which were attributed to the dense stromal edema. A less potent steroid, Lotemax Gel, was used instead of the traditional prednisolone acetate ophthalmic suspension 1%, as the patient was told to use it four times a day over the next month. Lotemax was developed to rapidly metabolize to inactive metabolites with the goal of minimizing side effects, so its versatility and safety allows its use for chronic therapy.<sup>13</sup>

Concerns to consider regarding longer-term use of topical steroids include increased IOP, cataract formation and overall suppression of the immune response. A review by Jones and Rhee showed that when treated with topical steroids for four to six weeks, 5% of the population demonstrated a rise in IOP greater than 16 mmHg and 30% had a rise of 6-15 mmHg. A study included in the review showed that IOP returned to baseline or normal approximately one week after discontinuation of steroid treatment.<sup>14</sup> As the patient in this case report did not have any predisposing conditions (a personal or family history of glaucoma, younger child or older adult, type-1 diabetic, history of connective-tissue disease, or high myopia), there was less concern about use of a steroid eye drop for this period of time.

Nonetheless, other methods for measuring IOP, such as the Icare tonometer (Icare USA), should have been utilized. This tonometer would have allowed the student intern or provider to precisely and repeatably measure an area of the cornea that may correlate closely with topography (e.g., the thinnest location). A study from Northern Ireland explored the relationship between IOP measurements and topographical variations in corneal curvature and corneal thickness. For the 49 keratoconic eyes studied, the median central pachymetry and IOP values were 495  $\mu\text{m}$  and 10 mmHg, respectively. The median temporal and nasal pachymetry and IOP values were 621  $\mu\text{m}$  and 641  $\mu\text{m}$ , and 14 mmHg and 13 mmHg, respectively.<sup>24</sup> This study suggests that the Icare tonometer could perhaps offer more precise measurements of IOP because it can be utilized over the thinnest area of the cornea.

Other topical drops that could be used in this scenario are artificial tears and antibiotics (if the epithelium is compromised). With epithelial compromise, a topical, broad-spectrum antibiotic (such as ciprofloxacin hydrochloride ophthalmic solution 0.3% up to four times-a-day) for prophylactic coverage could be prescribed. A broad-spectrum antibiotic ointment, such as erythromycin, also could be recommended for overnight use. Research indicates that use of topical medications only is considered a conservative approach. Corneal perforation is a rare occurrence, and various case reports have shown that patients can be managed conservatively over several days with aqueous suppressants, pressure patching and bandage soft contact lenses until resolution.<sup>15</sup>

During the resolution phase of corneal hydrops, corneal neovascularization is a potential complicating factor. Neovascularization of the cornea is a serious concern when the site of hydrops is near the limbal vasculature. Topical steroid drops should be prescribed to inhibit the reaction, and, should worsening occur, systemic steroids may be indicated. In some of the cases reported, the neovascular response began two to four weeks after the onset of hydrops, an important reason for perhaps monitoring these patients monthly.<sup>16</sup>

In cases of acute hydrops, a relatively new management approach — intracameral injection of gas/air to reduce the duration of corneal edema — has been used. The purpose of injecting gas/air is creating a barrier to prevent the aqueous humor from passing through the ruptured Descemet's membrane into the stroma. Blocking the intrusion of aqueous humor into the stroma would allow for faster healing of the corneal endothelial cells over the exposed stroma, and deposition of the new Descemet's membrane.<sup>17</sup> Miyata and Tsuji evaluated air injection as a barrier between the endothelial cell layer and aqueous humor. They monitored measures such as how long the corneal edema lasted, the length of time between the onset of hydrops and when the eye could wear a gas permeable (GP) contact lens again, and the best-corrected visual acuity with a GP lens after the edema had subsided. A control group was

utilized for comparison. The results showed that the average duration of corneal edema was 20.1 days in the intracameral air injection group and 64.7 days in the control group. The average length of time between the onset of hydrops and when the eye could wear a GP contact lens again was 33.4 days in the intracameral air injection group and 128.9 in the control group. Best-corrected visual acuity with a GP lens after the edema had subsided ranged from 20/50 to 20/25 in the air injection group and from 20/100 to 20/25 in the control group. The researchers noted the intracameral air injection induced no complications.<sup>17</sup>

Gas, either perfluoropropane ( $C_3F_8$ ) or hexafluoride ( $SF_6$ ), is another potential barrier injection. Basu and Vaddavalli studied utilizing  $C_3F_8$  gas injected between the endothelial cell layer and anterior aqueous face. Their primary outcome measure was average time to resolution of corneal edema, calculated both from the date of onset of hydrops and the date of initiation of therapy. Results showed the average time to resolution, both from the date of onset of symptoms and from the date of initiation of therapy was significantly lower in the study group than in the control group, which received no surgical intervention (90.5 days vs. 125 days and 78.7 days vs. 117.9 days, respectively).<sup>18</sup>

Panda and Aggarwal studied utilization of  $SF_6$  gas as a barrier. Although they studied a smaller patient population, corneal edema resolved at four weeks in the treatment group and not until 12 weeks in the control group.<sup>19</sup> Despite the development of complications such as pupillary block and increase in IOP, the  $SF_6$  gas injection treatment was deemed effective. It has been found that surgical intervention is a relatively safe and successful therapy for the early reduction of corneal edema, whether with air or gas.

Severity of visual acuity reduction depends on the size and location of the break in Descemet's membrane and subsequent edema. Most patients, before the development of hydrops, wear some form of GP contact lens correction. When hydrops develops, contact lens wear must be discontinued due to the irregular corneal surface and edema. If epithelial defects are present, any abrasiveness from a flat fitting or low clearance contact lens can exacerbate discomfort. As the area of edema heals and scar tissue forms, the previous area of steepening (usually somewhere within in the cone) will flatten. This flattening effect can sometimes aid in vision if the scarring does not occur centrally, and it can make contact lens fitting easier.

The Collaborative Longitudinal Evaluation of Keratoconus study showed that rigid contact lens wear at baseline, regardless of how the lenses were fitted (steep, aligned or flat), was associated with incident corneal scarring. Although a greater proportion of the corneas wearing flat-fitting contact lenses were scarred, after controlling for disease severity, the risk of corneal scarring did not increase with flat vs. steep rigid contact lens fit.<sup>25</sup> Due to fit and comfort issues, clinicians may elect to proceed with scleral lenses, which allow for complete vaulting of the cornea. This fit can protect the cornea from irritation secondary to a contact lens. If hydrops caused a central scar, penetrating keratoplasty (PK) may be a suitable option to maximize visual potential.

Tuft and Gregory showed that the development of hydrops in eyes with keratoconus was a significant risk factor for subsequently receiving a PK, and at the end of the study period 87 of the 147 eyes studied (59%) had surgery for visual rehabilitation.<sup>20</sup> Grewal and Laibson showed that various medical therapies did not differ significantly with regard to outcome, and ultimately 4 of 22 patients (18%) underwent a PK.<sup>9</sup> Tuft and Gregory also showed that the same studied eyes that underwent a PK had a greater rate of graft rejection than eyes grafted without hydrops.<sup>20</sup> But Fan Gaskin and Good with the Auckland keratoconus study found no statistically significant differences in the overall corneal transplantation rate between the two studied groups (subjects with keratoconus and corneal hydrops over a 17-year period compared with an age- and gender-matched control group of subjects with keratoconus but no prior history of corneal hydrops).<sup>7</sup> Basu and Reddy showed that the risk of endothelial rejection episodes was greater in eyes with longer duration of corneal hydrops and co-existent ocular allergy. Also, although endothelial rejection episodes are more common in eyes with resolved corneal hydrops, long-term

allograft survival and visual results after PK are similar in eyes with keratoconus with and without prior corneal hydrops.<sup>21</sup>

## Conclusion

A standard of care for the management of corneal hydrops, employing either topical ophthalmic medications or surgical intervention in the form of an intraocular injection, has yet to be established. A logical approach to treatment is to reduce the patient's symptoms and expedite corneal healing.

Conservative management of corneal hydrops with topical treatment may take two to four months for complete resolution, while a surgical approach may shorten that time. Although surgical intervention leads to a faster resolution, topical management is less invasive and presents minimal complications. With either modality, the final vision result will be similar. Perhaps size, location and amount of edema should dictate which method of treatment would be most effective. If the hydrops is off-center and patient complaints are minimal, a conservative approach of topical therapy may suffice. Conversely, if the edema is large in diameter and central, and central scarring affecting visual acuity status post healing is a concern, referral to a cornea specialist for surgical intervention may be warranted.

This condition can initially be quite uncomfortable and visually disabling to the patient. However, once the area of hydrops has resolved and the previous ectatic area is flatter, vision may actually be better than prior to onset (as it was for the patient in this case report, who had post-hydrops spectacle-corrected visual acuity of 20/50<sup>+1</sup> vs. 20/60<sup>-2</sup> pre-hydrops). Post-hydrops fitting with scleral contact lenses — bridging the patient's unique corneal curvature — may be the best opportunity to increase vision potential, although corneal GP lenses may be easier to fit with a new, flatter curvature. If scarring is severe centrally once the edema has resolved, vision may not improve regardless of the contact lens type selected. A referral to a cornea specialist for a cornea transplant may be warranted.

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