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Corneal Hydrops Secondary to Advanced Keratoconus

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A 12-year-old boy with Down syndrome presented to the emergency department (ED) with a 2-month history of redness, swelling, irritation, and decreased vision of his right eye. Review of systems was negative for headaches, fevers, recent trauma to the eye, or other illnesses. According to his mother, the patient had been treated in Mexico with prednisolone eye drops 1 week prior to presentation to the ED, but after his condition worsened, his family had brought him to the United States for further workup and care.

Eye examination findings were significant for briskly and equally reactive pupils bilaterally, and bilateral conjunctival injection that was worse in the right eye compared with the left eye (**Figure 1**). Examination findings were also remarkable for a translucent area of swelling covering the iris of the right eye (**Figure 2**). Visual acuity was 20/50 for the left eye, and the patient counted fingers at 5 feet for the right eye.

The ophthalmology service was consulted on arrival to the ED, and a protective eye shield was placed over his right eye. The ophthalmologist diagnosed corneal hydrops secondary to advanced keratoconus.

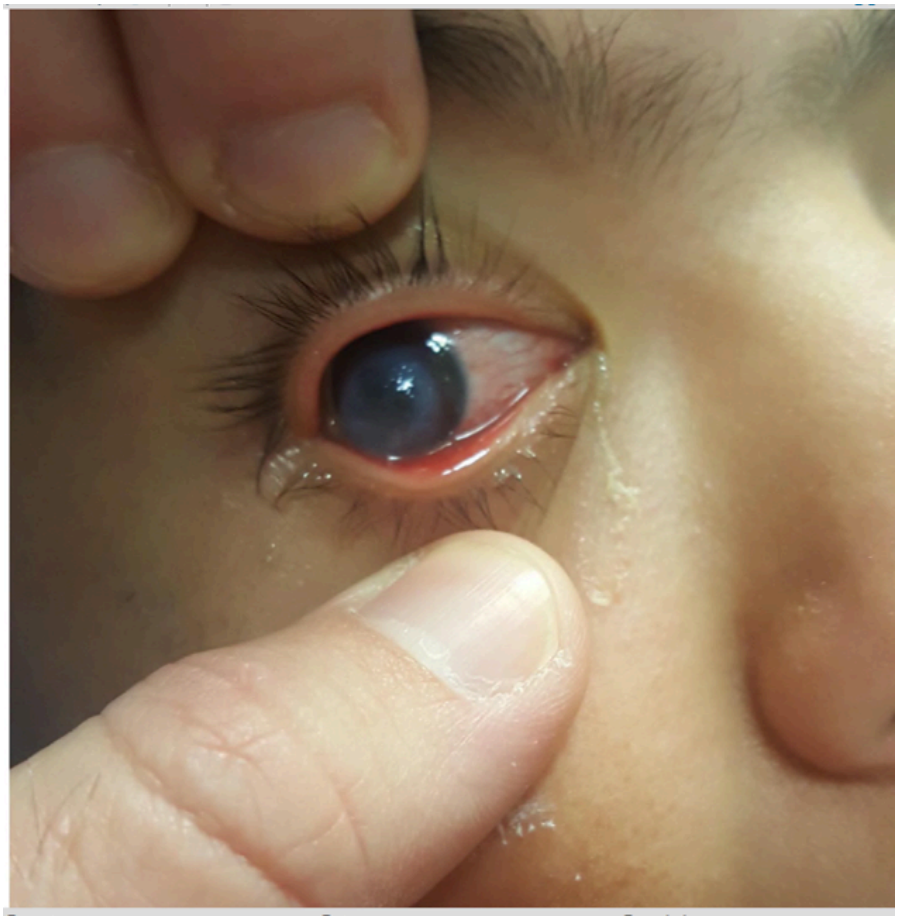

Figure 1.

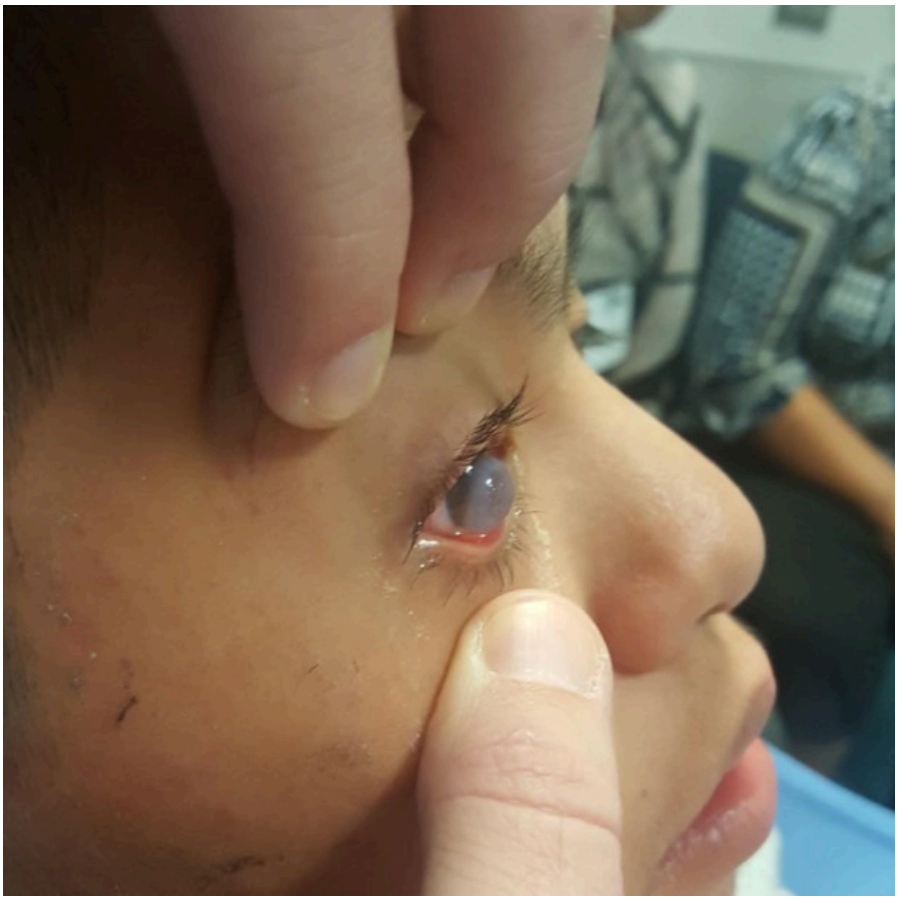

Figure 2.

Discussion. Keratoconus is a progressive condition that is associated with structural corneal collagen changes.[1] The onset is usually around puberty, and the condition worsens during early adulthood. The prevalence of keratoconus is widely variable, with an estimate of 1 in 2000 individuals.[2] Patients with various conditions, including Marfan syndrome, Ehlers-Danlos syndrome, Down syndrome, and retinitis pigmentosa, have a higher risk of developing keratoconus.

Pediatric keratoconus tends to be more aggressive than adult keratoconus, and although it usually starts as a unilateral condition, it has a 50% chance of becoming bilateral.[3,4] Management in adults typically includes visual rehabilitation with glasses, contact lenses, and corneal cross-linking. However, with pediatric keratoconus, the diagnosis often is not made until the disease has progressed beyond mild disease. Rapid deterioration of this condition is concerning for an unfavorable prognosis. Corneal cross-linking has shown great promise in the management of pediatric keratoconus.[5,6]

Keratoconus is usually a clinical diagnosis, and the differential diagnosis includes corneal abrasion, corneal edema, and, in the earlier stages, sometimes even conjunctivitis. If keratoconus is suspected, an ophthalmologist should be included as soon as possible in the care of the patient.

Outcome of the case. The patient was discharged home on atropine, prednisolone, and dorzolamide hydrochloride-timolol maleate eye drops, with a follow-up visit scheduled with the ophthalmology clinic for the following day. The patient was advised to wear a Fox shield for eye protection in the interim. Due to the very advanced nature of the disease, the cornea was found to be almost completely opacified with extreme thinning. With use of the prescribed medication and frequent visits to the ophthalmology clinic, the patient's condition improved. However, the prognosis for complete recovery of his vision remains grim, and the boy is undergoing evaluation for a possible corneal transplant.

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