The Necessity for Ocular Assessment in Atopic Children: Bilateral Corneal Hydrops in an 8 Year Old

abstract

Acute corneal hydrops is a rare complication of advanced corneal ectasia. This case report describes the clinical course of a child with severe atopy and no previous ocular examination who developed bilateral, acute corneal hydrops, secondary to keratoconus, by 8 years of age. The report demonstrates the rapidity of progression in the pediatric phenotype of the disease. This case also provides an important clinical lesson in highlighting the necessity for children with atopy to be referred for comprehensive ophthalmic examination, even in the apparent absence of visual symptoms, to ensure the timely diagnosis and management of any atopy-associated ocular disease. Prompt referral is particularly essential for pediatric corneal ectasia, in which the rapidity of progression may preclude stabilizing treatments, such as corneal collagen cross-linking, and result in significant childhood visual impairment, as was the case for this child. This case demonstrates the potential for significant ocular involvement in atopic children and identifies scope to enhance current international clinical guidelines relating to the management of childhood atopy through the inclusion of ocular screening for associated disease. Pediatrics 2014;134:e596–e601

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KEY WORDS
cornea, keratoconus, atopy, allergy, child, hydrops, corneal ectasia

ABBREVIATIONS
CXL—corneal collagen cross-linking
R—right eye
L—left eye
VA—visual acuity

Dr Downie undertook the clinical management of this patient, conceptualized the writing of this report, and undertook all aspects of the drafting of the manuscript.

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Atopic diseases, including eczema, asthma, and rhinoconjunctivitis, are the most common chronic conditions of childhood. Atopy is also an established risk factor for the bilateral, noninflammatory corneal ectasia keratoconus, which is characterized by progressive thinning of the corneal stroma. The prevalence of keratoconus is ~5.4 per 10,000, the condition affects all ethnicities, without gender predilection. Keratoconus classically manifests at puberty and is progressive until the third or fourth decade, when it usually arrests. Childhood diagnosis is relatively rare and considered a negative prognostic factor, with a heightened risk of more rapid ectasia and need for corneal transplantation, compared with late-teenage and adult-onset disease.

Furthermore, the presence of atopic disease is associated with more rapid corneal thinning, the need for earlier corneal grafting and a higher incidence of postgraft immunologic complications. Despite this known link between atopy and corneal degeneration, current international clinical guidelines relating to the management of childhood atopy do not include recommendations for ocular health evaluation.

This report describes the clinical course of a boy, who was 7 years old at initial presentation, with severe atopy and no previous ocular examination who rapidly developed bilateral, acute corneal hydrops. Acute corneal hydrops, resulting from spontaneous rupture of Descemet's membrane, is a rare complication of advanced ectasia, occurring in ~3% of keratoconic eyes. Recently, predictors for the development of acute corneal hydrops in keratoconus have been established to include Pacific ethnicity, history of eye rubbing, poor visual acuity at first presentation, and lack of family history. To the author’s knowledge, there are no previous published reports of corneal hydrops occurring in a child at such a young age. Furthermore, this case highlights the clinical necessity for children with atopy to be referred for ocular examination, even in the absence of significant visual concerns, to enable the timely diagnosis and management of any coexistent ocular conditions, in order to minimize the likelihood of incipient visual impairment.

**CASE REPORT**

In June 2009, a 7-year-old Polynesian boy accompanied his mother for her routine ocular examination in our clinic. During this consultation, the attending clinician (L.E.D.) observed the child frequently rubbing his eyes and eyelids. Upon directed questioning of the child's mother, it became evident that her child was severely atopic, with asthma, seasonal allergic rhinitis, and eczema affecting his eyelids and upper limbs. These conditions were being managed by his general medical practitioner; the child had previously attended a dermatologist for evaluation of his eczema, which was being treated with topical 1% hydrocortisone ointment, twice daily. A routine ocular examination had never been performed or recommended and so was suggested by the consulting optometrist. There was no known family history of ocular disease. Ocular history was negative for surgery; frequent, vigorous eye rubbing had been noted by the child's mother since an early age. The child was not aware of any vision difficulties; his only ocular symptom was that his eyes were often itchy.

Examination revealed a bilateral reduction in unaided vision (right eye [R] 20/300, left eye [L] 3/100). Visual acuities (VAs) remained abnormal (R 20/40, L 20/100) with spectacle correction. Slit-lamp biomicroscopy revealed bilateral, paracentral inferior stromal corneal thinning with positive signs of advanced ectasia (ie, Vogt striae and apical scarring) in the left eye (Fig 1A and B). Central corneal thicknesses were significantly reduced (R 482 μm, L 399 μm; compared with a mean normal value of ~536 μm). Computed videokeratography confirmed inferior-temporally positioned corneal apices, consistent with keratoconus (R < L; Fig 1C and D). The superior palpebral conjunctivae demonstrated mild generalized hyperaemia and a moderate papillary response, indicative of a nonspecific ocular immune reaction. Dilated ocular fundus examination confirmed normal posterior ocular health (R and L).

Therapeutic management of the allergic conjunctivitis was initiated with a topical anti-histamine/mast-cell stabilizer (ketotifen fumarate 0.025%, twice daily). The importance of ceasing eye rubbing was emphasized. The child underwent fitting for custom, rigid gas permeable contact lenses to mask the underlying corneal irregularity, enabling a significant improvement in VA (R 20/25, L 20/40). Ophthalmologic referral confirmed the child’s suitability for right corneal collagen cross-linking (CXL); the advanced nature of the ectasia precluded treatment in the left eye.

The child shortly relocated with his family to their Polynesian homeland. In December 2009, e-mail correspondence was received advising of a sudden “large, white spot” on the child’s left eye. The family returned to Australia for emergency eye care. Five days later, examination confirmed a left acute corneal hydrops; severe stromal edema and epithelial bullae involved two-thirds of the cornea (Fig 2). VA in the affected eye was count-fingers at 50 centimeters. Supportive treatment was initiated with nonpreserved lubricants (hourly, q1h), fluorometholone acetate (0.1% q2h) and topical prophylactic antibiotic therapy (chloramphenicol 0.5%, 4 times daily). After 1 month, topical therapy was ceased; the child returned overseas.
In March 2010, the family returned to Australia. A full-time spectacle correction enabled satisfactory vision for school-related tasks. Central corneal thicknesses were R 392 µm, L 422 µm, contraindicating right CXL. Despite concerted effort to control the child's eye rubbing through the use of topical antiallergy therapeutics, cold compresses (as required) and parental reminders at home, further rapid, progressive corneal ectasia was documented in the right eye over the following months. In October 2010, the child presented with a right acute corneal hydrops (Fig 3A, day 1). The

FIGURE 1
Baseline anterior eye photographs and corneal topography. Slit-lamp photographs of the left eye at baseline, showing characteristic signs of keratoconus: A, Vogt striae; B, apical stromal scarring. Corneal axial power maps with normalized scale (in diopters, D) for the right (C) and left (D) eyes at baseline showing inferiorly decentered corneal apices with more advanced steepening in the left eye.
clinical course and treatment was prolonged compared with the fellow eye. Ocular surface rehabilitation required intensive topical antiinflammatory treatment with prednisolone acetate 1.0%/phenylephrine acetate 0.12% q2h and nonpreserved ocular lubricants, cyclopia to improve comfort, and prophylactic topical antibiotics for several weeks. Over the first 21 days, the area of stromal edema expanded to involve the entire cornea (Figs 3 B, C, and D). Stromal neovascularization and opacification were present by day 48 (Fig 3E). Topical corticosteroids were tapered over 7 months; intraocular pressures remained within normal limits throughout treatment. By day 112, the peripheral neovascularization had reduced, with formation of a central stromal scar and relative stabilization of the anterior ocular surface (Figs 3 F, G, H, and I). Over this period, the left eye had stabilized with significant regression of the central opacity. Corneal transplantation was discussed with the child's mother but rejected because conservative management

FIGURE 2
Left eye: 5 days postacute corneal hydrops. Slit-lamp photographs of the left acute corneal hydrops with white diffuse light (A), sodium-fluorescein (B), and in optic section (C) showing severe corneal stromal edema and epithelial bullae over most of the cornea.

FIGURE 3
Right eye: the clinical course of acute corneal hydrops from day (D) 1 to D150. A series of slit-lamp photographs showing the biomicroscopic appearance of the right acute corneal hydrops from initial presentation (D1) until stabilization (D150). An expansion in the area of involved cornea was evident from D1 (A) to D7 (B), through D21 (C and D). Early peripheral corneal pannus (black arrow) at D48 (E) progressed to significant stromal neovascularization (white arrow) by D55 (F). By D82 (G), the central edema had resolved and the pannus had regressed, with progressive stabilization of the cornea (D112) (H) until the formation of a residual central stromal scar (D150) (I).
was preferred. By May 2011, VA had improved to L 20/30°. A left hybrid contact lens (consisting of a rigid gas permeable center bonded to a soft hydrogel skirt) fitting was undertaken to facilitate participation in sporting activities; this modality was preferred for ease of adaptation and its excellent on-eye stability. At the most recent attendance (March 2012), further bilateral resolution to the corneal opacification and recovery of VA were documented (Table 1).

**DISCUSSION**

This unique case describes the clinical course of a young atopic child with previously undiagnosed, progressive corneal ectasia, who developed bilateral corneal hydrops within months of diagnosis. This report is significant for the following:

1. Highlighting the necessity for atopic children to undergo routine ophthalmic examination to ensure a timely diagnosis and management of any associated ocular disease;
2. Demonstrating the rapidity of progression in pediatric keratoconus and bilateral acute corneal hydrops, with significant visual impairment;
3. Detailing the natural history of keratoconus, including a high-resolution pictorial sequence from pre– to post–acute corneal hydrops, both in relation to the anterior ocular signs and corneal topographic changes; similar documentation of the disease in this manner apparently does not exist in the literature.

Although the etiology of keratoconus is unknown, it can be strongly associated with atopy. Nevertheless, routine ocular examination is not currently included in clinical guidelines for childhood atopy. In the present case, the child had severe atopy but few visual symptoms other than ocular itchiness. The importance of avoiding eye rubbing, a risk factor for the development and progression of keratoconus, had not been discussed. Such factors led to a delayed diagnosis of keratoconus and potential disease progression due to unidentified environmental risk factors. Prompt diagnosis of pediatric patients with keratoconus is imperative to ensure that there is an opportunity for treatments, such as corneal CXL, to be administered within the necessary therapeutic window. CXL involves the photosensitizing agent riboflavin and ultraviolet-A irradiation to induce photooxidative cross-linking of the stromal collagen to increase corneal biomechanical stability. For safety, a minimum corneal thickness of 400 μm is required after epithelial debridement; apical corneal scarring must also be absent. As such, the severity of the keratoconus in this 7-year-old child’s left eye was beyond suitability for CXL at diagnosis; the fellow cornea developed ectasia that contraindicated treatment over the next few months. Medium- to long-term clinical data suggest the capacity for CXL to stabilize the disease. Furthermore, significant and rapid functional improvement has been demonstrated in pediatric keratoconic patients.

A future consideration for this child relates to the implications of both the hydrops and the presence of corneal neovascularization on corneal graft viability. The association between hydrops and corneal allograft survival per se is controversial; conflicting data exist regarding whether eyes with resolved hydrops have a relatively higher or lower incidence of endothelial rejection. Most recently, it was found that keratoconic eyes with prolonged hydrops (ie, >3 months) and/or those with coexistent ocular allergy were more prone to postgraft endothelial rejection. Neovascularization also compromises the physiologic corneal immune privilege, potentially doubling the risk of allograft rejection. The presence of seasonal allergic conjunctivitis, protracted clinical course of the hydrops, and development of corneal neovascularization are therefore significant negative prognostic factors for the longevity of any transplant for this child.

This case highlights the potential to provide improved care for atopic children in relation to appropriate screening for comorbid ocular disease. Ideally, corneal stabilization with treatments such as CXL, rather than the late-stage management of hydrops documented in this case, will be the mainstay of treatment of pediatric patients with atopic-related keratoconus in the future.

### TABLE 1 Summary of Best-Corrected VA in Each Eye, Pre– and Post–Acute Corneal Hydrops

<table>
<thead>
<tr>
<th>Date</th>
<th>Age</th>
<th>Refractive Correction</th>
<th>VA</th>
</tr>
</thead>
<tbody>
<tr>
<td>June 2009</td>
<td>7 y 8 mo</td>
<td>Spectacles</td>
<td>20/40</td>
</tr>
<tr>
<td>December 2009</td>
<td>8 y 1 mo</td>
<td>Spectacles</td>
<td>20/25</td>
</tr>
<tr>
<td>March 2010</td>
<td>8 y 4 mo</td>
<td>Spectacles</td>
<td>20/50</td>
</tr>
<tr>
<td>August 2010</td>
<td>8 y 9 mo</td>
<td>Spectacles</td>
<td>20/100</td>
</tr>
<tr>
<td>October 2010</td>
<td>9 y 11 mo</td>
<td>Spectacles</td>
<td>CF</td>
</tr>
<tr>
<td>November 2010</td>
<td>9 y</td>
<td>Spectacles</td>
<td>20/200</td>
</tr>
<tr>
<td>January 2011</td>
<td>9 y 2 mo</td>
<td>Hybrid contact lens</td>
<td>n/a</td>
</tr>
<tr>
<td>May 2011</td>
<td>9 y 6 mo</td>
<td>Spectacles</td>
<td>20/200</td>
</tr>
<tr>
<td>August 2011</td>
<td>9 y 9 mo</td>
<td>Spectacles</td>
<td>20/160</td>
</tr>
<tr>
<td>March 2012</td>
<td>10 y 4 mo</td>
<td>Spectacles</td>
<td>20/80</td>
</tr>
</tbody>
</table>

**CF**, counting fingers; **GP**, rigid gas permeable; **n/a**, not applicable.

*Acute corneal hydrops.
REFERENCES

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