

# Iridocorneal Endothelial Syndrome in a Chinese Child

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## Abstract

**Purpose:** We reported a rare case of ICE syndrome in a Chinese child.

**Case report:** A 6-year-old child was admitted to the ophthalmology department of the Affiliated Wuxi Second Hospital of Nanjing Medical University, complaining of elevated intraocular pressure at his first diagnosis in a local hospital. The patient was initially treated with medication. During subsequent follow up, two iris holes were seen in the patient's right eye and he was then diagnosed with ICE syndrome. The child underwent surgery combined with mitomycin C intraoperatively. The intraocular pressure was stably controlled.

**Conclusion:** ICE syndrome has been rarely reported in children. Glaucoma filtering surgery combined with intraoperative administration of mitomycin is efficacious for intraocular pressure control. (*Eye Science* 2013; 28:153–156)

**Keywords:** children; ICE syndrome; glaucoma surgery

Iridocorneal endothelial syndrome (ICE) is a spectrum of disease characterized by primary corneal endothelial abnormality, progressive iris atrophy, synechia, and secondary glaucoma. ICE is rarely seen in clinical practice and mainly affects middle-aged and senior adults. Here, we reported a case of 6-year-old boy diagnosed with ICE syndrome.

## Patient data

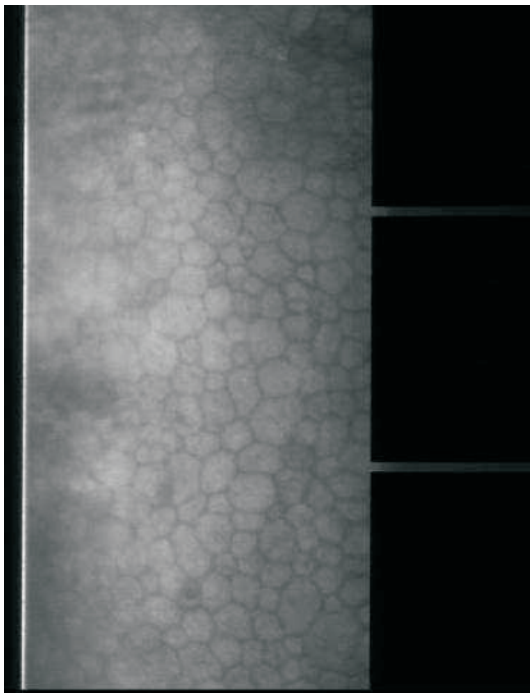
The boy was admitted to a local hospital at the age of 5, with a 5-year history of complaints of white lesions in his right eye, and was then diagnosed with “bilateral congenital glaucoma” (intraocular pressure: 29 mmHg OD, 23 mmHg OS). The intraocular pressure was still high after 1-month treatment using carteolol. He was switched to the

medication therapy of latanoprost but the high intraocular pressure was not effectively lowered after 6 months. He was admitted to our hospital on 10 January 2012. He had no family history of glaucoma. Ophthalmologic examinations revealed: visual acuity: 0.1 OD, intraocular pressure: 23.6 mmHg, conjunctival hyperemia (+) in the right eye, corneal diameter 14 mm in the right eye, strip-shaped opacity was observed in central corneal stroma, KP (+), Tyndall (–), peripheral corneal thickness > 1/3 central thickness, loose iris texture, round pupil with a diameter of 3 mm, sensitivity to light, and transparent lens. Fundus examinations revealed: in the right eye, optic disc with clear margins and light color, C/D=0.5, no patent abnormalities (obvious dellen and incisional mark, etc.) surrounding optic nerve, retinal nerve fiber layer thickness within normal range, macular foveal reflection (+); visual acuity: 0.4 OS, intraocular pressure: 18 mmHg, transparent cornea with a diameter of 13 mm, KP(–), Tyndall (–), peripheral corneal thickness > 1/2 central thickness, clear iris texture, round pupil 3 mm in diameter, sensitivity to light reflection, and transparent lens. Fundus examinations revealed: in the left eye, optic disc with clear margins and normal color, C/D=0.3-0.4, no patent abnormalities surrounding the optic nerve, normal retinal nerve fiber layer thickness, and macular foveal reflection (+). Auxiliary examinations revealed: corneal endothelial cell count: OD 1725.4/mm<sup>2</sup>, significantly enlarged size of the corneal endothelial cells, decreased cell density, a slight portion of endothelial cells had a regular hexagonal shape while most showed an irregular shape (Figure 1). OS 2314.5/mm<sup>2</sup>, endothelial cells displayed a regular hexagonal shape and no apparent abnormality was noted (Figure 2). OD axial length: 21.52 mm, OS axial length: 22.14 mm. Treatment

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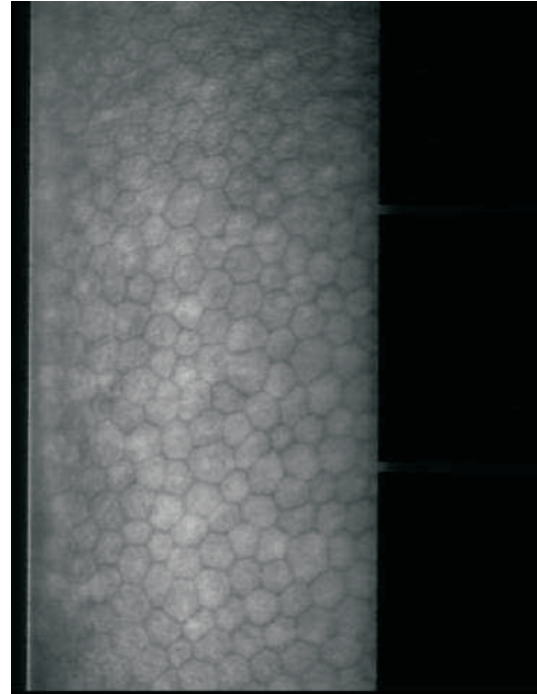
plan: during follow up, changes in the fundus optic nerve were observed and carteolol+brinzolamide was administered to reduce intraocular pressure. On 30 January 2012, a spindle iris hole, approximately 1 mm × 3 mm in size, was found at the 7 o'clock position in the right eye (Figure 3). Another spindle hole was seen at the 11 o'clock position in the right eye on a return visit on 5 March 2012, measuring approximately 1 mm × 2 mm in size (Figure 4). Diagnosis: 1. ICE syndrome in the right eye; 2. Secondary glaucoma in the right eye; 3. Bilateral macrocornea. Treatment plan: the patient underwent trabeculectomy combined with the administration of mitomycin C (0.2 mg/ml for 4 min). The patient had normal postoperative intraocular pressure and was advised to regularly undergo ocular examinations in terms of intraocular pressure and changes in the cornea, iris, and optic nerve.



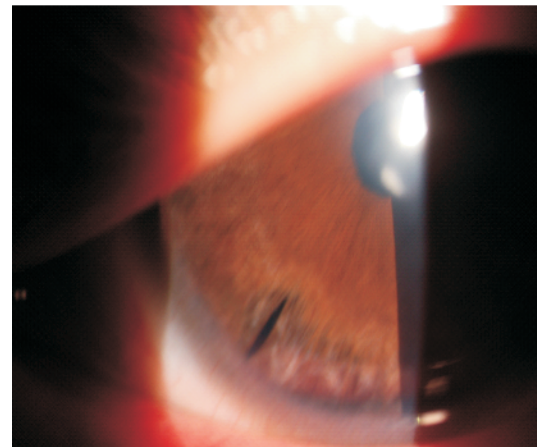
**Figure 1** Corneal endothelia in the right eye

## Discussion

ICE syndrome is a type of acquired disease with a 100% rate of blindness<sup>1</sup>. The underlying pathogenesis remains unexplained. Certain possible hypotheses have been proposed: the Campbell membrane theory, herpes simplex virus or Epstein-Barr (EB) virus

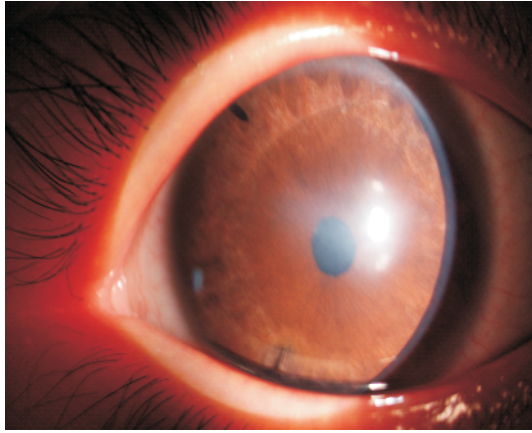


**Figure 2** Corneal endothelia in the left eye



**Figure 3** A subtemporal spindle-shaped iris hole in the right eye

theory, and inflammation theory, *etc.* At present, ICE syndrome can be clinically categorized into three types: progressive essential iris atrophy, Chandler's syndrome, and Cogan-Reese syndrome<sup>2</sup>. (1) Progressive essential iris atrophy is likely to affect young and middle-aged adults, mainly occurs as a unilateral episode characterized by widespread iris atrophy involving the full-thickness iris and accompanied by iris perforation, leading to pupil displacement, corneal endothelial abnormality and a mild



**Figure 4** A superior temporal iris hole in the right eye detected at the third return visit

degree of edema, and even secondary glaucoma in the advanced stages. (2) Chandler's syndrome frequently attacks middle-aged women, mainly as a unilateral episode, which is primarily characterized by abnormal corneal endothelium, a slight degree of iris atrophy and pupil involvement, without iris perforation, and constantly accompanied by intraocular pressure elevation. (3) Cogan-Reese syndrome is mainly seen in middle-aged female Caucasians, commonly has a unilateral involvement, and is clinically characterized by abnormal proliferation of corneal endothelium, iris atrophy, small nodules on the surface, and secondary glaucoma in the advanced stage.

No radical treatment targets ICE syndrome; the treatments mainly target secondary glaucoma and corneal edema. Denis<sup>3</sup> suggested that the success rate of filtering surgery was relatively high in patients with early ICE complicated with secondary glaucoma to lower intraocular pressure. The presence of corneal endothelial abnormality was likely to lower the success rate of filtering surgery. Lanzl et al<sup>4</sup>. found that the success rate of up to 80% for trabeculectomy combined with intraoperative use of mitomycin in 10 patients with ICE complicated with secondary glaucoma. Doe et al<sup>5</sup>. conducted a long-term follow-up of 26 patients with ICE complicated with secondary glaucoma who underwent trabeculectomy combined with intraoperative administration of antimetabolites or embedment of a decompression valve. Subsequent follow-up revealed that the efficacy of IOP control was significantly improved compared with tra-

beculectomy alone, hinting that the combination of intraoperative use of mitomycin and surgical methods may improve the success rate to certain extent. For patients with the symptoms of corneal edema, use of hypertonic agents or corneal contact lens should be considered as early treatments. For those presenting with corneal opacity and bullous formation, surgical treatment is the primary option. Price et al<sup>6</sup>. found that Descemet's membrane with selective replacement dysfunction can effectively alleviate corneal edema, delay visual loss and yields less injuries and recovers more rapidly compared with penetrating keratoplasty.

At present, cases of ICE syndrome in children have been rarely reported in China. In 2006, Salim reported 1 child with ICE syndrome. Initially, the patient underwent goniotomy and IOP elevated up to 30 mmHg 18 months later. The child subsequently underwent trabeculectomy combined with intraoperative use of mitomycin and IOP was effectively controlled<sup>7</sup>. The following ocular diseases should be identified from ICE syndrome: 1. Glaucoma complicated with congenital malformations, such as Axenfeld-Rieger syndrome, is a rare autosomal dominant disorder and commonly bilateral. Approximately 50% of patients had glaucoma with normal corneal endothelia and inactive iris malformations<sup>8</sup>. 2. Corneal endothelial diseases; Fuchs' endothelial corneal dystrophy is mainly characterized by multiple cornea guttata stretching from the posterior surface of central cornea to anterior chamber, seen with a slightly bronze color, Descemet's membrane thickened with focal posterior excrescences and irregular gray opacity. No signs of iridocorneal adhesion or iris atrophy were noted. In this case, the patient presented with significantly abnormal corneal endothelial count and morphology compared with normal subjects, and subsequently apparent iris atrophy and iris holes accompanied with IOP elevation. Therefore, the patient was diagnosed with progressive essential iris atrophy and treated with trabeculectomy combined with intraoperative use of mitomycin. The child's IOP is stable but remains to be further observed.

### Disclosure statement

There is no conflict of interest to declare.

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