

## Case report

# Misdiagnosis induced intraocular lens dislocation in anterior megalophthalmos

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Anterior megalophthalmos (AM) is an uncommon developmental anomaly of the anterior segment of the eye with a constellation of findings that includes enlarged cornea, deep anterior chamber, posterior positioning of the iris and lens, iris stroma atrophy, hypoplasia of iris dilator, pupil displacement, large capsular bag, lens subluxation, prematurely cataract and the tendency to retinal detachment. AM, especially when symptoms are mild, is not an easy disease to diagnose. We present 3 AM cases that were misdiagnosed as congenital cataract with weak zonule and megalocornea. Intraocular lenses (IOLs) dislocated after standard cataract surgeries and subsequent surgery (replacing the dislocated IOLs with iris-claw intraocular lenses) achieved satisfactory outcome. Although rare, AM should be included in the differential diagnosis of enlarged cornea and we recommend implanting Artisan lens in AM patients.

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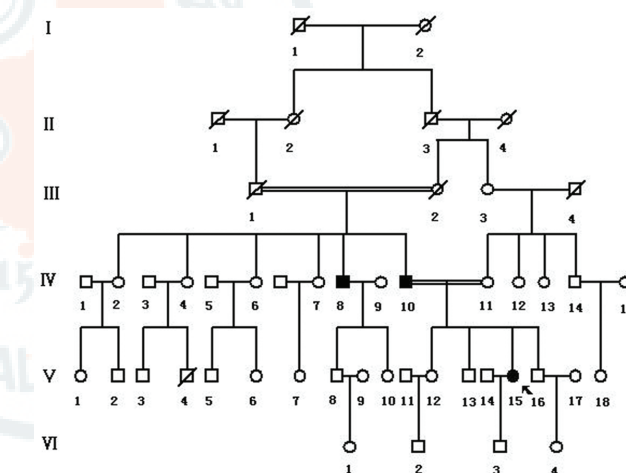
Anterior megalophthalmos (AM) is a rare, usually asymmetric inherited developmental anomaly of the anterior segment of the eye. Although X-linked recessive (possibly being located on Xq21.3-q22) inheritance is most common, all modes of inheritance (sporadic, autosomal recessive and autosomal dominant) have been reported. Abnormal findings include corneal diameter greater than 13.0 mm, corneal embryotoxon, mosaic corneal dystrophy, deep anterior chamber, posterior positioning of the iris and lens, pigment dispersion syndrome, iris hypoplasia, iris stroma atrophy, transillumination defects, hypoplasia of iris dilator, pupil displacement, large capsular bag, lens subluxation, premature cataract, and a predisposition for retinal detachment.<sup>1</sup> AM is easy to be clinically misdiagnosed as megalocornea. This report presents three cases of intraocular lens dislocation caused by misdiagnosis and inappropriately designed surgery.

### CASE REPORTS

#### Case 1

A 34-year-old woman with no history of systemic disease presented with gradually decreasing vision in the left eye. Ocular history included an uneventful extracapsular cataract extraction (ECCE) with posterior chamber intraocular lens implantation (Pharmacia 722c PC IOL, 13.5 mm in diameter) in the right eye, 3 years prior to presentation. She had a family history of premature cataract and of consanguineous marriage in two consecutive generations (Figure 1).

Best corrected visual acuity (BCVA) was 1.0 in the right eye and 0.08 in the left. Bilaterally, the corneas measured 13 mm horizontally. On slit-lamp examination, there were iris pigment deposits on the endothelial cells and the anterior chamber angle. It also showed a hyper-deep



**Figure 1.** Family pedigree. The family history revealed three affected members in two generations. The dark symbols represent the affected members of the family; the clear symbols indicate unaffected members. The circles and squares indicate female and male family members, respectively. The arrow points to the proband.

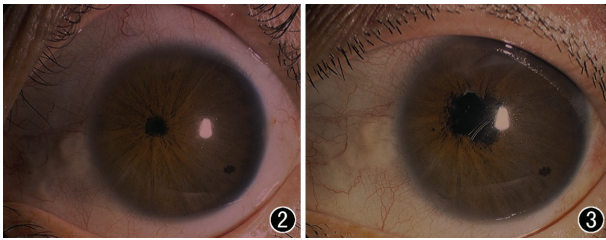
anterior chamber, pupil displacement (Figure 2), lens subluxation, and posterior subcapsular cataract. Fundoscopy could not be performed, as the pupil could only be dilated from 1.0 mm to 1.5 mm in diameter. Intraocular pressures (IOPs) in the two eyes were 15 mm Hg and 19 mmHg, respectively.

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**Figure 2.** Macrocornea with pupil displacement. The pupil could be dilated from 1.0 mm to only 1.5 mm in diameter by pharmacological mydriasis.

**Figure 3.** Dislocation of the IOL in the left eye of case 1.

Congenital cataract with weak zonule and megalocornea in the left eye was diagnosed preoperatively. A capsular tension ring was inserted after phacoemulsification because of the weakness of the zonule, and a PC IOL (Bausch & Lomb ADAPT-AO, 10.7 mm in diameter) was implanted subsequently. Visual acuity improved to 0.8 immediately after surgery. However, the patient complained of vision fluctuation (VA 0.1–0.8) and monocular diplopia on the third day, and slit-lamp examination showed IOL dislocation (Figure 3). Ultrasound biomicroscopy revealed iris hypoplasia (Figure 4). Further examination showed that the endothelial cell density (ECD) was 3134 cells/mm<sup>2</sup> and the anterior chamber depth was 5.52 mm.

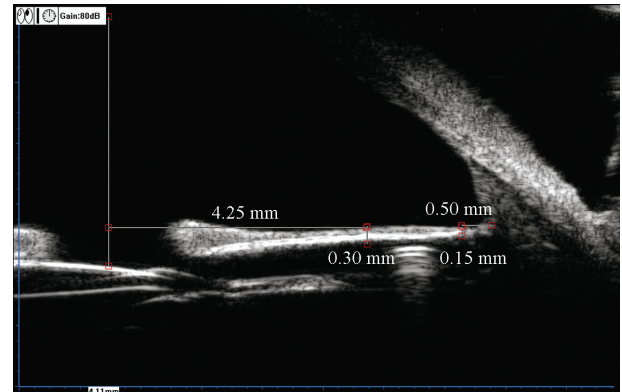
This further examination confirmed a diagnosis of AM. Therefore, the dislocated IOL was removed, and an iris-claw IOL (Artisan intraocular lenses, Ophtec BV) was implanted in the left eye, 1 month after the initial surgery. At last examination, 9 months after this surgery, BCVA was 0.9 and the IOL remained well centered (Figure 5). The IOP was 16 mmHg, and ECD was 3003 cells/mm<sup>2</sup>. After requesting additional medical history information for this patient, we identified cases 2 and 3.

**Case 2**

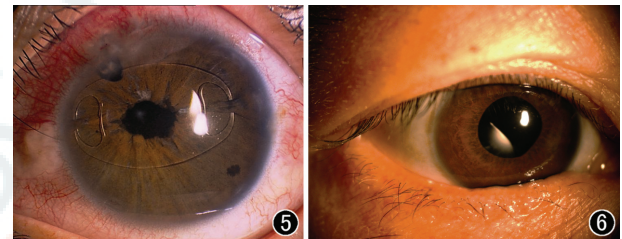
A 65-year-old man, father of case 1, had complained of blurred vision in both eyes. He had undergone cataract surgery on the left eye at his local hospital, but the clinical record was unavailable. The ocular manifestation of symptoms in his right eye was similar to that seen in his daughter. ECCE was performed, and a standard PC IOL (ORC Company, 13.5 mm in diameter) was implanted in his right eye. Upon last examination, 20 years after surgery, visual acuity in that eye was 0.8, and ultrasound biomicroscopy examination showed the IOL to be decentered slightly. He refused further surgery on the left eye, which had a dislocated IOL and poor VA (0.1).

**Case 3**

The uncle of case 1, a 49-year-old man, complained of blurred vision in both eyes and monocular diplopia in the right eye. He had had cataract extraction and PC IOL (PMMA, 13.5 mm in diameter) implantation of both eyes at the local hospital by an unspecified surgical technique. Visual acuity was 0.3 in the right eye and 0.25 in the left



**Figure 4.** Thickness at 4.25 mm from the central of the pupil was 0.3 mm; thickness at 0.5 mm from the periphery was 0.15 mm.



**Figure 5.** Iris-claw IOL in place.

**Figure 6.** Decentration of the IOL in the right eye of case 3.

eye. Both of the IOLs had dislocated rightward (Figure 6), and his other ocular manifestations were similar to those of his niece, except that his pupils could be dilated to 5.5 mm in diameter. The dislocated IOL was removed, and an iris-claw IOL was implanted in his right eye. Upon last examination, 6 months after the surgery, BCVA was 0.9, and the symptoms of diplopia disappeared. IOP and ECD were normal. The patient is awaiting surgery on his left eye.

**DISCUSSION**

Enlarged corneas occur in one of three patterns: simple megalocornea unassociated with other ocular anomalies, anterior megalophthalmos, and buphthalmos due to congenital glaucoma.<sup>1</sup> Clinically, AM is likely to be misdiagnosed as megalocornea. The premature development of lens opacities is the most common cause of vision loss in AM patients. Enlargement of the ciliary ring and capsule, together with weakness of the zonule, increase the risks for cataract surgery. IOL dislocation caused by zonule weakness or a mismatch in diameter between the IOL and the capsule bag is the most frequent postoperative complication. Therefore, cataract complicated with AM should be properly differentiated from cataract with weak zonule and megalocornea, in order to avoid the implantation of a capsular tension ring<sup>2</sup> and conventional IOL, where mismatches can deteriorate and aggravate lens dislocation.

We include in this report relevant clinical features from the seven separate surgeries on the five eyes of the three cases, and suggest that implantation of an iris-claw IOL

would result in a better outcome than that with a routine PC IOL. The prevalent surgical approaches to AM treatment include implanting an angle-supported anterior chamber intraocular lens, anterior iris fixation of an iris-claw IOL,<sup>3</sup> posterior iris fixation of an iris-claw IOL,<sup>4</sup> a scleral-fixated IOL,<sup>5</sup> and a custom-made PC IOL.<sup>6</sup> Two eyes in the affected family (the right eyes of cases 1 and 2) achieved acceptable results with ECCE and 13.5-mm PC IOL implantation; the IOLs decentered slightly, without greatly affecting visual acuity. While it was reported that using a larger-diameter IOL in patients with AM could achieve perfect centration,<sup>6</sup> larger PC IOLs are no longer commercially available. There are certain advantages to a scleral-fixated IOL, including fewer disturbances to the corneal endothelial cells, but implantation is a complicated procedure. Improper surgical manipulations in the region of the ciliary body can be associated with lens tilt, suprachoroidal hemorrhage, retinal detachment, and even endophthalmitis.<sup>5</sup> Although the implantation of an iris-claw IOL has some disadvantages, complications can be avoided by meticulous planning before surgery. An iris-claw IOL rarely causes pigment dispersion as long as the lens is fixed appropriately, because the melanin granule density of the iris pigment epithelium is much greater than that of the anterior stroma of the iris.<sup>4</sup> Additionally, although the iris thickness 0.5 mm from its periphery is merely 0.15 mm and seems incapable of holding an IOL, the thickness of the iris at the enclavation site is much greater (0.30 mm), and the iris can stabilize the IOL with an adequate amount of enclavated tissue (Figure 4). Furthermore, a review of the literature found a negative correlation between anterior chamber depth and loss of endothelium in patients with an iris-claw IOL.<sup>7</sup> The corneal endothelial cells are relatively safe, because the anterior chamber is hyper-deep in these patients. Finally, the danger caused by pupil dilation is avoided, given the hypoplasia of the iris dilator seen in these patients.

Additionally, the surgical approach of cataract extraction is open to discussion. Phacoemulsification is an option for most cases. When the lens have dislocated dramatically, intracapsular cataract extraction may be a better choice in order to clean up the cataract safely and

thoroughly, decrease the incidence of severe complications such as damage to the endothelial cells and loss of the lens nucleus into vitreous cavity.

In summary, AM should be included in the differential diagnosis of an enlarged cornea, and its proper diagnosis is critical. Our study suggests that iris-claw IOL implantation is suitable for eyes with AM. In cases of significant IOL dislocation, secondary surgery, including dislocated IOL extraction and iris-claw IOL implantation, should be recommended. With the appropriate use of these procedures, visual outcomes for these patients could be improved and their suffering could be alleviated.

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