Management of cataract in a patient with anterior megalophthalmos

Syed Bilal Hassan Zaidi,1 Kirk A J Stephenson,1 Hafiz Muhammad Ejaz-Ul-Haq,2 Hassan Massanna3

DESCRIPTION
A healthy 40-year-old man presented with reduced (counting fingers) right eye visual acuity (VA) due to a dense unilateral cataract (posterior subcapsular and nuclear sclerosis) (figure 1A). Examination revealed bilateral enlarged but thin corneas and deep anterior chambers (ACs) (table 1). Bilateral iridotomies and phacodonesis were noted with no significant subluxation of the right crystalline lens. Iridocorneal angles were open bilaterally with normal intraocular pressures (12 and 14 mm Hg, right and left, respectively) (figure 1C). Corneal diameters were 14.5 mm/14.5 mm and 15.0 mm/15.0 mm, right and left, respectively, in horizontal and vertical meridians (figure 1D). There was no view of the fundus in the right eye, but the left eye posterior segment was normal without stigmata of vireorectal degeneration. B-scan ultrasonography excluded major right eye posterior segment pathology. Megalocornea, extremely deep AC and zonular laxity, without sequelae of congenital glaucoma (ie, raised intraocular pressure and optic neuropathy), suggested a diagnosis of anterior megalophthalmos (AM). Cataract surgery for visual improvement was discussed and consented for by the patient.

Cataract extraction in AM presents a number of surgical challenges. Extreme AC depth makes access to the cataract difficult, but this was bypassed by supracapsular prolapse of the lens nucleus out of the capsular bag also minimising shear stress on the weakened zonules of Zinn. Intraocular lens (IOL) choice in such a large eyes and deep ACs with weak, stretched zonules. The main lens issues in AM are instability (crystalline lens and IOLs).2 IOL choice is critical in such a large eye with deep ACs with weak, stretched zonules. The main lens issues in AM are instability of standard diameter IOLs (ie, 13 mm) within the abnormally large capsular bag, lack of optic centration and postoperative posterior dislocation of IOLs.3–5 Cases of IOL dislocation with endocapsular fixation have been described.4 Multiple IOL strategies have been employed, including iris-claw

Table 1 Biometric parameters of the eyes in this anterior megalophthalmos case

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Right eye</th>
<th>Left eye</th>
<th>Normal range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Axial length</td>
<td>24.16 mm</td>
<td>24.19 mm</td>
<td>22–26 mm</td>
</tr>
<tr>
<td>AC depth</td>
<td>6.39 mm</td>
<td>5.81 mm</td>
<td>1.5–4 mm</td>
</tr>
<tr>
<td>Lens thickness</td>
<td>4.00 mm</td>
<td>4.04 mm</td>
<td>4 mm</td>
</tr>
<tr>
<td>White to white distance</td>
<td>15.0 mm</td>
<td>14.5 mm</td>
<td>11–12 mm</td>
</tr>
<tr>
<td>Central corneal thickness</td>
<td>475 µm</td>
<td>493 µm</td>
<td>555 µm</td>
</tr>
<tr>
<td>K1</td>
<td>45.2 D</td>
<td>44.3 D</td>
<td>42.00–44.00 D</td>
</tr>
<tr>
<td>K2</td>
<td>47.2 D</td>
<td>47.9 D</td>
<td>42.00–44.00 D</td>
</tr>
<tr>
<td>Preoperative refraction</td>
<td>Precluded by cataract</td>
<td>–2.50 D to –0.50 D x 120</td>
<td>–6.00 D to +3.00 D</td>
</tr>
</tbody>
</table>

The axial lengths (ultrasound for the right eye), anterior chamber depths, lens thickness, white to white distance, central corneal thickness and keratometry (K1 and K2), as measured by IOLMaster 700 V1.1.14 (Carl Zeiss Meditec AG, Jena, Germany) and refraction (dioptre sphere, DS, and dioptre cylindrical, DC) are shown relative to normal reference ranges. AC, anterior chamber.
### Learning points

- Consider a diagnosis of anterior megalophthalmos in the presence of megalocornea, ultradepth anterior chamber and zonular laxity (ie, iridophacodonesis, ectopia lentis) without signs of congenital glaucoma or increased axial length.
- Preoperative diagnosis allows appropriate surgical planning to ensure appropriate intraocular lens (IOL) choices and promote optimal refractive and visual outcomes while avoiding IOL dislocation into the vitreous cavity.
- The surgeon must consider intraoperative challenges in anterior megalophthalmos, including accessing the cataract in an ultradepth anterior chamber, baseline zonular instability with difficult capsulorhexis and IOL considerations, in addition to standard cataract risks.

IOLs, iris-sutured IOLs and custom-made larger-diameter IOLs. AM is associated with vitreous abnormalities (eg, syneresis, synchysis and optically empty vitreous with strands) and vitreoretinal pathology (eg, lattice degeneration, associated with retinal detachment); thus, annual dilated fundal examination is recommended. Other notable ocular associations include anterior embryotoxon, mosaic corneal dystrophy, glaucoma due to goniodysgenesis, irregular astigmatism, and amblyopia due to strabismus and myopia. Mutations in the Xq12–q26 region have been implicated previously. Systemic associations include Marfan syndrome, trisomy 21, Apert syndrome, mucolipidosis type 2 and Walker-Warburg syndrome.

Recognition and diagnosis of AM allows planning for the monitoring of the posterior segment and preparation for the unique challenges of cataract surgery in the complex anterior segment.

### REFERENCES