

Application of ultrasound biomicroscopy in the planning of cataract surgery in anterior megalophthalmos

Mohammad Ali Zare, Bahram Eshraghi,

Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences, Tehran, Iran

Correspondence to: Dr. Mohammad Yaser Kiarudi, Farabi Eye Hospital, Qquazvin Square, South Kargar Street, Tehran, Iran. E-mail: y_kia76@yahoo.com

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*Mohammad Yaser Kiarudi,
Ebrahim Azaripour Masoule*

Anterior megalophthalmos, a rare hereditary disorder, is macrocornea (horizontal corneal diameter more than 13 mm) in association with enlarged lens-iris diaphragm and ciliary ring. One of the major challenging issues in the cataract surgery of these patients is preventing intraocular lens (IOL) malposition, because of probable large capsular bag. Several approaches have been selected by previous surgeons, such as, custom-made anterior chamber IOLs. In this study, we show a normal capsular bag diameter despite ciliary ring enlargement, with application of ultrasound biomicroscopy (UMB). We suggest that in cases of anterior megalophthalmos without phacodonesis, UBM could measure the actual size of the capsular bag and obviate the need

for further procedures.

Key words: Anterior megalophthalmos, cataract, ultrasound biomicroscopy

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Anterior megalophthalmos, a rare hereditary disorder, is macrocornea (horizontal corneal diameter more than 13 mm) in association with enlarged lens-iris diaphragm and ciliary ring. Other features include anterior embryotoxon, iris hypoplasia, stromal atrophy, pigment dispersion, myopia, cataract, glaucoma, and lens subluxation or dislocation.^[1] It differs from megalocornea (isolated macrocornea), which is non-progressive and x-linked recessive.^[2] The patients with anterior megalophthalmos have a very deep anterior chamber and vitreous index (vitreous length / axial length \times 100) below 69%.^[3]

Cataract surgery in a megalophthalmic eye is challenging because of some technical aspects like limited view, cavernous chamber, thin peripheral cornea, and also the possibility of decentration of a standard intraocular lens (IOL) that is implanted in the large capsular bag.^[4] In previous studies different managements have been reported such as custom-made anterior chamber IOLs and suturing,^[5] a posterior chamber-placed iris-clip intraocular lens,^[6] and also a custom-made IOL.^[4]

In this case report we have introduced the advantages of preoperative ultrasound biomicroscopy (UBM) application, to determine the actual size of the capsular bag that may obviate

the need for further procedures.

Case Report

A 42-year-old man was referred to our clinic because of decreased visual acuity in both eyes. On examination, the best corrected visual acuities (BCVA) were 20/100 in the right eye and 20/60 in the left; intraocular pressures were within normal limits. The features of the anterior megalophthalmos were corneal diameter of 16.5 mm [Fig. 1], hyperdeep anterior chambers, and broad ciliary bands on gonioscopy. Funduscopy in both eyes showed no abnormality. Each lens showed moderate nuclear sclerosis cataract, and there was no sign of phacodonesis. The Keratometry measurements in the right and left eyes were 40.25@ 5°, 42.75@ 95°, and 40.25@ 175°, 42.5@ 85°, respectively. With the help of the IOLMaster (Carl Zeiss Meditec), the anterior chamber depth and axial length in the right and left eyes were determined to be 6.47 mm, 28.47 mm and 6.03 mm, 27.61 mm, respectively. The vitreous index was 64%. The diagnosis of the anterior megalophthalmos was confirmed with high anterior chamber depth and vitreous index less than 69%.^[3] The IOL power was calculated by the SRK / T formula. UBM (SONO MED, Vu Max II) was performed and the angle-angle distance was measured to be 16.25 mm in the right and 16.5 mm in the left eye [Fig. 2]. The capsular bag diameter was 10.21 mm and lens thickness was 3.59 mm. Although the angle-angle distance was increased, the capsular bag diameter was normal so there was no need for custom made or an iris-claw IOL. For cataract surgery, the surgeon (M.Z) followed a standardized procedure: topical anesthesia (lidocaine 2% in gel suspension) on the cornea, a 3.2 mm self-sealing temporal clear corneal incision, intracameral injection of 0.5 mL of preservative-free lidocaine 1%, followed by an injection of a dispersive ophthalmic viscosurgical device (OVD) (hyaluronate 3.0% [Vitrax]), capsulorrhexis, nuclear fracturing (during phacoemulsification there was backward shifting of the capsular bag, because of elongation of zonules, however, zonulolysis was not apparent and the capsular tension ring was not used), cortical cleanup, implantation of a foldable acrylic IOL (AcrySof MA60AT, Alcon Laboratories, Inc.), and

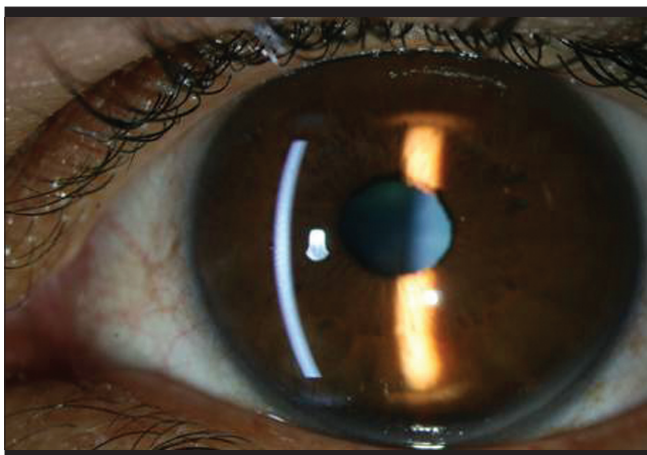


Figure 1: Slit-lamp appearance of macrocornea

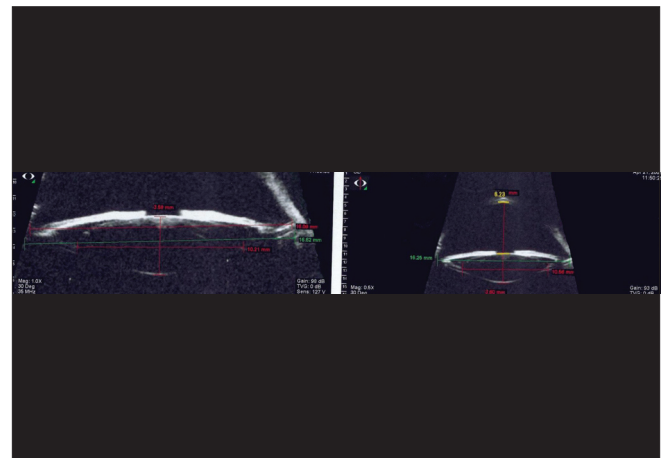


Figure 2: Ultrasound biomicroscopy in the radial section of the globe shows angle – angle distance, capsular bag diameter, lens thickness, and anterior chamber depth

aspiration of the OVD.

No sutures were applied. Postoperatively, the BCVA improved to 20/30 in the right eye, with a refractive error of + 2.00 diopter (D) sphere-2.50 D cylinder \times 5°. The IOL remained well-centered, without pseudophacodonesis, after five months follow-up.

Discussion

One of the major challenging issues in the cataract surgery of patients with anterior megalophthalmos is preventing IOL malposition, probably due to a large capsular bag. Kwitko *et al.*^[7] reported dislocation of a standard size posterior chamber IOL. They recommended using a larger IOL diameter in patients with megalocornea.

Marquez and Osher in their report, applied a custom IOL; an IOL with a 7.0 mm optic and an overall length of 16.0 mm.^[4] However, the process lasted for almost one year and the patient was unwilling to wait for the same period of time for the second eye.

To our knowledge, we have introduced the application of UBM the first time, to predict the risk of IOL decentration and for measurement of capsular bag. UBM has been used to examine the intraocular position of IOL haptics after phacoemulsification, extracapsular cataract extraction, and transscleral or iris fixation of the posterior chamber lenses.^[8] Kuchenbecker *et al.*^[3] described a case of anterior megalophthalmos with ciliary body dysplasia, diagnosed by using UBM.

Anterior megalophthalmos is a non-progressive, often symmetric inherited disorder (X-linked recessive, possibly located on Xq21.3-q22). On account of the enlargement of the ciliary ring, it is thought that the capsular bag is larger than normal and insertion of a standard posterior chamber IOL is unfavorable because of the likelihood of decentration.^[9]

In this study, we have shown a normal capsular bag diameter, despite ciliary ring enlargement, with application

of UBM. After surgery, the patient had a hyperopic shift. This phenomenon could be explained with a more posterior shifting of the IOL due to 'long' zonules. However, we do not want to generalize this case to all cases of anterior megalophthalmos. It is obvious that some of these patients have lens subluxation or dislocation that needs its own consideration, such as, insertion of capsular tension rings.^[10]

We suggest that in cases where there is no phacodonesis, UBM could measure the actual size of the capsular bag and obviate the need for a custom IOL to be designed.

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