·Letter to the Editor·

Cataract surgery and intraocular lens implantation in anterior megalophthalmos

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Dear Sir,

I am Dr. Yun Li, from the Second Hospital Affiliated to Nanchang University, Jiangxi Province, China. I write to present a case report of IOL implantation in anterior megalophthalmos.

Anterior megalophthalmos (AM) is the term suggested by Vail [1] to define bilateral enlargement of the anterior segment of the eyeball in the absence of an increased intraocular pressure. It is a rare hereditary condition characterized by megalocornea (diameter greater than 12.0mm), enlarged ciliary ring, stromal atrophy, iris hypoplasia, lens subluxation or dislocation, and cataract formation at an early age.

The most common cause of visual impairment is the development of premature cataract, the extraction of which is prone to complications [2-6],especially when the crystalline lens is luxated or subluxated. The enlarged ciliary ring and capsular bag are deterrents to easy insertion of standard posterior chamber intraocular lenses (IOLs) which are likely to decenter.

Since its first description as a distinct entity by Vail in 1931, there have been a few reports about different techniques of cataract surgery and IOL implantation in Europe. However, to our knowledge, no reports of cataract extraction and IOL implantation in megalophthalmos have been reported in Asia.

Here we report a case of AM and describes how we managed the anterior megalophthalmos and present a new technique of standard posterior chamber IOL implantation which resulted in successful visual rehabilitation.

A 57-year-old man was referred to our department with bilateral progressive loss of vision and family history of ocular disease was negative. Visual acuity was light sensation without correction in the right eye and 20/40 with -6.50-2.00×85 in the left. Horizontal and vertical corneal diameters (measured with a pair of callipers under anesthesia) were 13.0mm and 12.0mm in both eyes, respectively. Axial lengths were 25.10mm in the right eye and 25.52mm in the left eye. The anterior chambers were 3.65mm deep in the right eye and 3.94mm deep in the left eye measured by UBM which showed that the anterior chamber angle was widely open. Bilaterally, localized corneal opacity was seen and irides showed marked moderate stromal atrophy and transillumination defects with iridodonesis. The pupil of either eye could not be fully dilated after pharmacologic mydriasis. The lens in the right eye was severely opaque and intumescent while the lens in the left eye showed moderate nuclear sclerosis (Figure 1). The fundus of the right eye could not be seen because of severe nuclear sclerotic cataract. The cup-to-disc (c/d) ratio in the left eye was 0.3 with healthy neuroretinal rim, and the retina was normal. Intraocular pressure was normal in both eyes.

An uneventful planned extracapsular cataract extraction was performed in the right eye.

A 4.0mm 2-plane limbus tunnel incision was constructed. DisCoVisc (Alcon) was used as the viscoelastic agent to maintain the anterior chamber, and can-opener anterior capsulotomy was performed. The nucleus was hydrodissected and delivered by manual expression, and the cortex was removed with a silicone irrigation/aspiration tip.

Because of the larger diameter of the ciliary ring of anterior megalophthalmos, it is suggested to implant a posterior chamber IOL with a lens diameter of 16.0 to 18.0mm [7]. However, such an IOL was not available. Under the circumstances, a posterior chamber IOL (Matrix Acrylic Aurium) with a 6.0mm optic and an overall length of 12.5mm was selected. The intraocular lens had a power of 19.50 diopters (D) determined by the SRK formula. After placing the IOL in the capsular bag, we let the patient sit up for 2 minutes, discovering that the IOL was dislocated temporally (Figure 2). As a result, we pulled the IOL from the capsular bag into the anterior chamber and made a haptic of the IOL outside the eye. From the limbus tunnel incision, one end of a double-armed 10-0 nylon suture was passed through the iris and the anterior capsule at 12 o'clock and passed from within the capsular bag, to emerge through

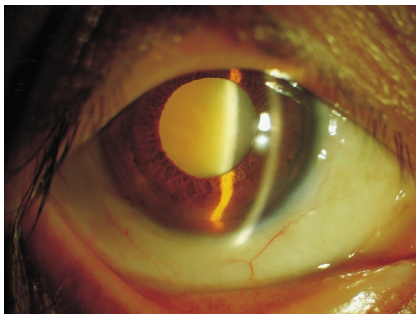

Figure 1 Localized corneal opacity can be seen and the anterior chamber is very deep in the right eye.Irides show marked moderate stromal atrophy.The lens is severely opaque and intumescent. The fundus could not be seen because of severe nuclear sclerotic cataract.

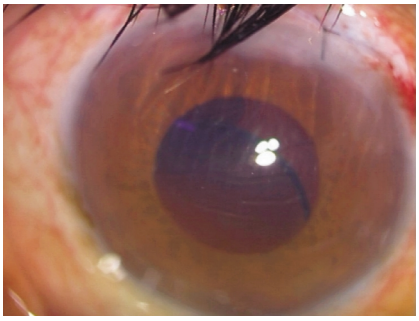

Figure 2 After placing the IOL in the capsular bag, we let the patient sit up for 2 minutes, discovering that the IOL was dislocated temporally.

the anterior capsule, the iris and limbus corneae at 6 o'clock. Then, the end of the suture was trimmed outside the eye and brought out of the capsular bag and the anterior chamber through the limbus tunnel incision. This suture was tied to the haptic of the IOL outside the eye. With gentle traction on the other end of a double-armed 10-0 nylon suture, the IOL was placed in the capsular bag. The suture was gradually pulled until there was no slack, tied to the sclera in limbus tunnel incision and trimmed. The limbus tunnel incision was closed with 10-0 nylon and the eye treated with topical steroid and antibiotic ointments. The postoperative course was uneventful, with a stable IOL.

One month after surgery, the visual acuity improved to 0.5 in the right. The IOL remained well centered without dislocation, and the retina was normal. The patient was very pleased with the visual result.

Anterior megalophthalmos is characterized by bilateral congenital enlargement of the cornea and anterior segment of the globe,and X-linked recessive is usually the most common mode of inheritance [8]. The condition is usually harmless, except for three complications that may appear late in life: dislocation of the lens, secondary glaucoma and cataracts [9-10]. The premature development of cataract and lens subluxation is the main cause of vision loss in megalophthalmos. These lenticular opacities usually develop by 30 to 50 years of age, and the cataract is of a complicated type.

Cataract surgery in megalophthalmos is challenging, especially given the option of IOLs. Implantation of a standard posterior chamber IOL is unsatisfactory because it is likely to decenter as a result of enlargement of the ciliary ring and capsular bag. Sergio Kwitko recommended a posterior chamber IOL with a lens diameter of 16.0mm to 18.0mm.

In this patient, we selected a standard posterior chamber IOL (Matrix Acrylic Aurium) with a 6.0mm optic and an overall length of 12.5mm, because the larger IOL was not available. The haptic of the IOL was sutured to posterior surface of the iris, anterior capsule and the sclera in limbus tunnel incision. Visual rehabilitation was successful. IOL dislocation did not appear. The way how we managed the anterior megalophthalmos demonstrated that extracapsular cataract extraction with a standard posterior chamber IOL sutured to the iris,anterior capsule and the sclera in limbus tunnel incision was a successful technique in megalophthalmos especially when the posterior chamber IOL with a lens diameter of 16.0mm to 18.0mm was not available.

Furthermore, we should pay more attention to retina examination in the postoperative period, because anterior megalophthalmic eyes are at greater risk of being affected by a type of vitreoretinopathy predisposing to retinal detachment[11].

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