

## Toric multifocal intraocular lens implantation in a case of bilateral anterior and posterior lenticonus in Alport syndrome

Jeevan S Ladi, Nitant A Shah

We report the first case of toric multifocal intraocular lens (IOL) implantation in both the eyes of a young patient of Alport syndrome with anterior and posterior lenticonus with a successful outcome. An 18-year-old female patient presented with progressively blurred vision in both eyes since 4–5 years not improving with glasses. Refraction showed high myopia with astigmatism; however, the vision did not improve beyond 6/60 with glasses correction. Clinical examination on slit lamp showed anterior and posterior lenticonus bilaterally with a classical oil droplet appearance. We performed clear lens extraction by phacoemulsification with toric multifocal IOL implantation in both eyes. Postoperatively, the patient achieved an excellent refractive outcome with the unaided vision of 6/9, N6 in both eyes.

**Key words:** Alport syndrome, lenticonus, toric multifocal intraocular lens

Lenticonus is an uncommon condition caused by bulging of lens capsule and underlying cortex. It can be anterior, posterior or both. Lenticonus anterior occurs bilaterally in Alport syndrome, a hereditary disease with a prevalence of 1 in 5000.<sup>[1]</sup> Alport syndrome is characterized by progressive renal failure, sensorineural deafness, and ocular manifestations (11–43% patients).<sup>[2]</sup> Ocular associations are polymorphous corneal dystrophy, lenticonus anterior, and flecked retina.<sup>[2,3]</sup> Rarely some cases have both anterior and posterior lenticonus.<sup>[4,5]</sup> Lenticonus is usually axial, 2–7 mm and causes myopia and astigmatism. Previously, it has been managed by clear lens extraction and intraocular lens (IOL) implantation.<sup>[3,4]</sup> We report the first case of bilateral anterior and posterior lenticonus in a young patient with Alport syndrome, in which clear lens extraction with toric multifocal IOL implantation was performed with a successful outcome.

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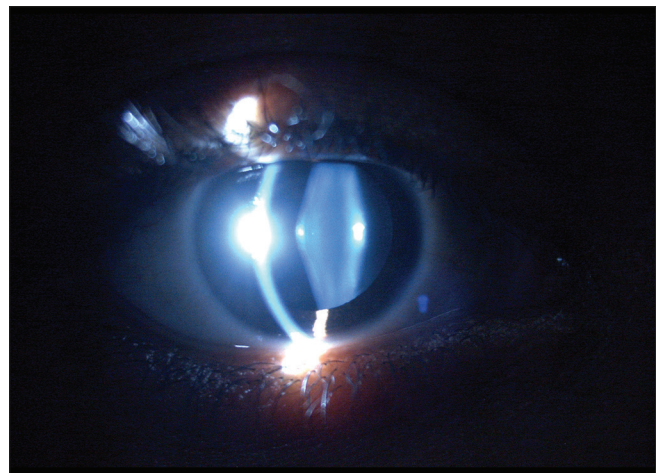
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## Case Report

An 18-year-old female patient presented with a progressive decrease in vision bilaterally since 4–5 years. Vision was 6/120 in the right eye with –19D sphere/–6.5D cyl at 10. The left eye improved to 6/60 with –19D sphere/–5D cyl at 160. In both eyes, Slit lamp showed anterior and posterior lenticonus [Fig. 1] with oil droplet reflex [Fig. 2]. Cornea was clear without dystrophy. Intraocular pressure was 14 right eye and 12 mm Hg in the left eye on Goldman applanation. Fundus was normal. The macula could not be focused by fundus camera (Zeiss Visucam Pro NM, Carl Zeiss Meditec AG, Jena, Germany). Biometry with IOL-Master (Carl Zeiss Meditec AG). Right eye axial length 22.78 mm, K1 43.38D at 17, K2 45.12D at 107. Left eye axial length 22.71 mm, K1 43.60D at 163, K2 45.98D at 73. Topography was done with the Galilei Dual Scheimpflug Analyzer (Ziemer, Switzerland). The patient had lenticular high astigmatism due to conus along with corneal astigmatism. It was decided to correct corneal astigmatism with toric multifocal IOL as it was more than 1.5D. Customized IOL (AT Lisa toric IOL, Carl Zeiss Meditec AG) was ordered.

Routine blood investigations were normal. Urine examination showed 3 + protein. The patient had partial hearing loss. Anterior and posterior lenticonus with urine protein and partial hearing loss indicated Alport syndrome.

Temporal clear corneal phacoemulsification with foldable toric multifocal IOL implantation was done in both eyes with 5 days interval. Preoperatively, reference marks were placed on the cornea with the patient sitting upright to correct for



**Figure 1:** Slit lamp photograph of the right eye showing anterior and posterior lenticonus

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cyclotorsion [Fig. 3]. Intraoperatively, the implantation axis was decided using reference marks and the alignment axis obtained from IOL calculation. Mendez degree gauge and Nuijts Toric axis marker (American Surgical Instruments Corp) were used.

The capsule was noted to be thin, and capsulorrhexis was done using DisCoVisc (Alcon-Couvreur, Belgium). The anterior capsule in area of the conus was fragile, hence, a two stage capsulorrhexis was performed, 3 mm was done first and was enlarged to 5.5 mm with forceps. Hydrodissection was not performed to avoid posterior capsular dehiscence. Hydrodelineation was done. The central lens matter was removed by phaco aspiration followed by cortical cleanup. Capsule polishing was done cautiously using flow rate 6 and vacuum 15. Foldable toric multifocal IOL (AT LISA Toric 909M, Carl Zeiss Meditec AG) was implanted. Right eye + 21 spherical power/+2.5D AT 104 and left eye + 20 spherical power/+3.5D at 75. After IOL implantation, the posterior capsular bulge disappeared hence posterior capsulorrhexis was not performed.

Postoperatively, the patient was given eye drops prednisolone (Alcon) 4 times a day for 3 weeks, moxifloxacin (Vigamox,

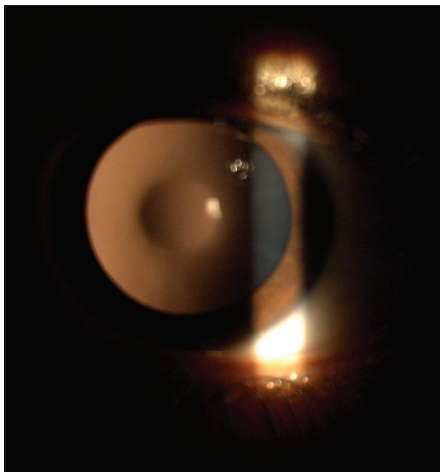
Alcon) 4 times a day for 1 week, and Nevanac (Alcon) 3 times a day for 6 weeks.

At 4 weeks uncorrected vision was 6/9, N6 in both eyes. Right eye 6/6p with +0.25D/-0.75D at 25 and left eye 6/6 with -0.25D. Both eyes showed a well-centered IOL [Fig. 4]. Retina could be focused well with fundus camera. At 1 year follow-up, uncorrected vision was 6/9 N6 in both eyes with no posterior capsular opacification [Fig. 5]. The vision was right eye 6/6p with +0.25D/-1D at 40 and left eye 6/6 with +0.25 D/-1 at 80.

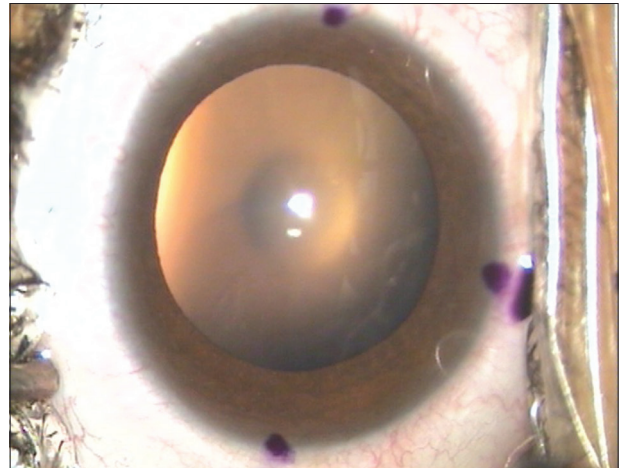
## Discussion

Alport syndrome is a rare disorder characterized by ocular abnormalities, progressive nephritis, and sensorineural hearing loss. Progressive deterioration of vision is caused by myopia induced by lenticonus. Our patient showed high myopia (-19D) due to the presence of both anterior and posterior conus which is rare. Lenticular astigmatism was significant (5D in right and 2.6D in left eye).

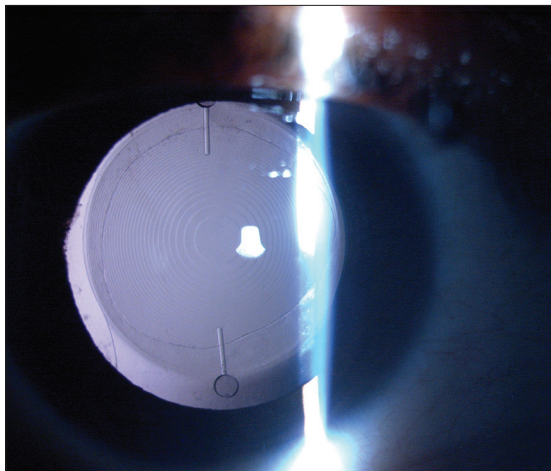
Clear lens extraction by phacoemulsification with foldable IOL implantation has been reported in lenticonus



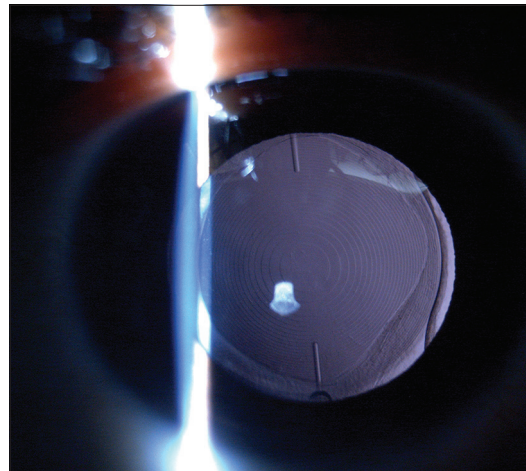
**Figure 2:** Slit lamp photograph showing oil droplet reflex in lenticonus



**Figure 3:** Reference marks placed on the cornea with a marker pen preoperatively



**Figure 4:** Postoperative slit lamp photo of the right eye showing well centered multifocal toric intraocular lens with good axis alignment



**Figure 5:** Slit lamp photo of 1 year follow-up of the right eye showing well centered intraocular lens with no change in axis alignment

with success.<sup>[4-6]</sup> Fragile lens capsule makes capsulorrhexis difficult, and it tends to go to periphery.<sup>[3,4,6]</sup> Hence, we performed it under DisCoVisc in two stages, initially, a small capsulorrhexis of about 3 mm and then enlargement to 5.5 mm.

Hydrodissection was not performed due to risk of posterior capsule dehiscence. Cortical cleanup and capsule polishing were done carefully. We chose to implant toric multifocal IOL as the young patient wanted spectacle independence and had significant corneal astigmatism (>1.5D). A recent study shows that postoperative astigmatism of >1.5D compromised distance and intermediate vision in eyes with multifocal IOL.<sup>[7]</sup> This decrease was significantly more prominent in eyes with a multifocal IOL than in eyes with a monofocal IOL, indicating that astigmatism management is crucial when implanting multifocal IOLs. Although there are other options to manage coexisting corneal astigmatism such as limbal relaxing incisions, clear corneal incisions, and postoperative laser refractive correction;<sup>[8]</sup> these have disadvantages such as limited predictability, dry eye, problems in wound healing, and need for additional surgery (in case of laser refractive correction).<sup>[9]</sup> Toric multifocal IOL offers the opportunity to correct astigmatism and achieve spectacle independence as shown by high satisfaction in our patient. High patient satisfaction with toric multifocal IOL's is also highlighted in a previously published study.<sup>[10]</sup>

In our patient, the vision did not improve preoperatively beyond 6/60 even with the correction of myopia and astigmatism, raising doubts of amblyopia. However, lenticonus is known to develop and progress in the second decade, and hence amblyopia is usually not seen; as has also been reported in previous studies.<sup>[5]</sup> The cause of nonimprovement even with glasses correction is due to the inability of the eye to focus rays at the fovea due to irregular refraction by the conus. This was illustrated by inability of the fundus camera to focus macula preoperatively.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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