Refractive Outcome after Clear Lens Extraction with Posterior Chamber Intraocular Lens (IOL) Implantation for Anterior Lenticonus in A Patient with Alport Syndrome

Dr. Shifat Toufique[1], Dr. Mostafa Hossain[2], Dr. Sazzad Iftekhar[3], Dr. Mamunur Rashid Chowdhury[4], Dr. Halima Alimi[5], Bipul Kumer De Sarker[6]

Abstract

Aim : *To report the refractive and visual outcome after clear lens extraction with posterior chamber IOL implantation for anterior lenticonus in a patient with Alport syndrome.*

Method: *A 16year old male patient who was referred for amblyopia therapy on account of isoametropic amblyopia was found to have myopic astigmatism of -4.50 cyl and -3.50cyl, with BCVA of 6/60 and 6/36 respectively in the right and left. The triad of anterior lenticonus in both eyes, deafness and ultrasound and biochemical evidence of early kidney disease led to a diagnosis of Alport syndrome.*

Result: *Subsequent clear lens extraction with posterior chamber IOL implantation in both eyes restored vision to 6/6 with no significant refractive error.*

Conclusion: *Timely surgical intervention with clear lens extraction and IOL implantation is effective in restoring vision to normal in patients with poor spectacle-corrected vision in Alport Syndrome.*

Key Words: *Alport Syndrome, Anterior lenticonus, Clear lens extraction, deafness.*

Resume

Objetif: Pour signaler les résultats visuels et réfractifs après extraction d'une lentille claire avec l'implantation d'une IOL de chambre postérieure pour un lenticonus antérieur chez un patient atteint du syndrome de Alport.

Methode : Un astigmatisme myope de -4,50 cyl et de -3,50 cyl a été trouvé chez un patient âgé de 16 ans qui avait été dirigé pour amblyopie en raison d'une amblyopie isoamétropique, avec une BCVA de 6/60 et 6/36 respectivement à droite et à gauche. La triade du lenticonus antérieur dans les deux yeux, la surdité au cours des 12 années précédentes et des échographies et des preuves biochimiques de néphropathie précoce ont conduit au diagnostic de syndrome d'Alport.

Resultat: Une extraction de lentille claire ultérieure avec une implantation de LIO de chambre postérieure dans les deux yeux a rétabli la vision à 6/6 sans erreur de réfraction significative.

Conclusion : Une intervention chirurgicale opportune avec extraction de lentille claire et implantation de IOL est efficace pour rétablir une vision normale chez les patients présentant une mauvaise vision avec correction de la vision dans

1. Consultant and Assistant Professor, Ispahani Islamia Eye Institue and Hospital (IIEIH), Farmgate, Dhaka, Bangladesh.
2. Senior Consultant and Associate Professor, IIEIH, Dhaka, Bangladesh.
3. Consultant and Assistant Professor, Ispahani Islamia Eye Institue and Hospital (IIEIH), Farmgate, Dhaka, Bangladesh.
4. Consultant and Assistant Professor, Ispahani Islamia Eye Institue and Hospital (IIEIH), Farmgate, Dhaka, Bangladesh.
5. Consultant, Paediatric Ophthalmology, An-Nur Eye Hospital, Lagos, Nigeria.
6. Associate Professor and Consultant, Ispahani Islamia Eye Institute& Hospital

Address of Correspondence: Dr. Halima Alimi. halimaalimi@yahoo.co.uk. +2348185236748

le syndrome de Alport.

Mots-Clés: syndrome de Alport, lenticonus antérieur, extraction de cristallin claire, surdité.

Introduction

Alport syndrome is a rare, genetic disorder characterized by eye abnormalities in addition to other systemic affectations including renal failure and deafness.[1] It is inherited in various patterns, either X-linked (85%), Autosomal recessive (10%) or by Autosomal dominant pattern (5%). The characteristic ocular features include anterior lenticonus and cataract, corneal opacities, central perimacular fleck retinopathies, and temporal retinal thinning.[2] Whilst renal manifestation and hearing difficulties require management by the concerned specialists, poor vision can be treated by clear lens extraction and intraocular lens implantation to eliminate or reduce myopic astigmatism.

Case Report

A 16year old male with myopic astigmatism was referred with a diagnosis of amblyopia. He had experienced a gradual reduction in distance vision and in his hearing over 12 years before presentation.

Examination

Unaided Visual acuity (VA) was 2/60 and 6/60 in Right and Left respectively. Best corrected visual acuity (BCVA) was 6/36 and 6/24 with lens power of Plano-4.50cylinderx180 and Plano-3.50 cylinder x180 respectively in the right and left after objective retinoscopy. There was oil droplet retinoscopic reflex in both eyes. Slit lamp examination revealed an anterior lenticonus in both eyes. The cornea, retina, ocular motility and intra-ocular pressures were normal in both eyes.

Systemic examination revealed sensorineural deafness.

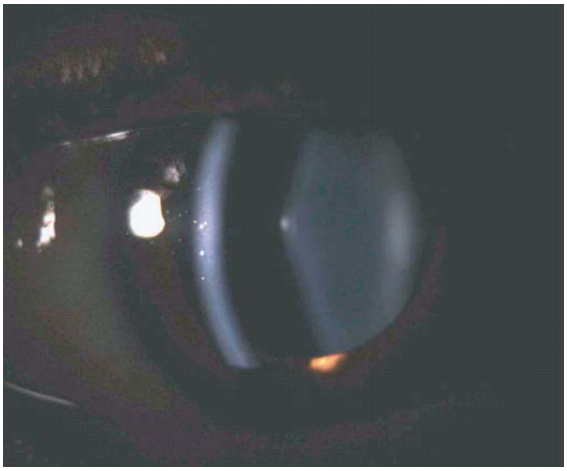
Investigations

An OCT of both anterior segments outlined outpouching of the anterior lens capsule centrally.

Urinalysis revealed 3+ of proteinuria and 20-25 red blood cells per high power field. Kidney ultrasound revealed features suggestive of early renal parenchymal disease. Serum Urea, creatinine and electrolyte levels were however normal.

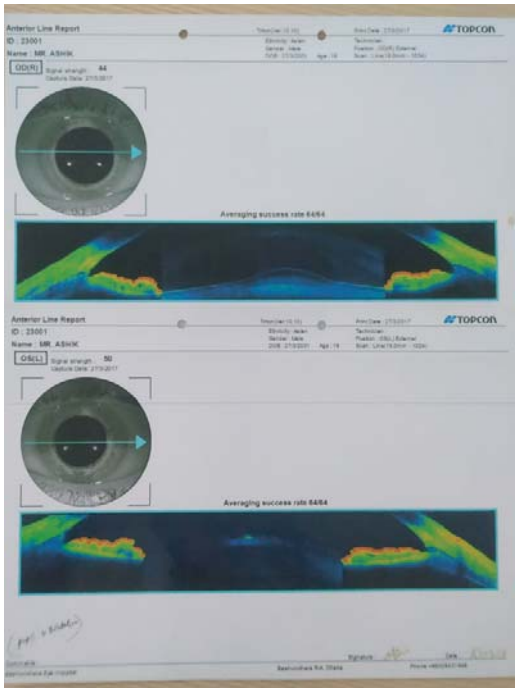
Cornea topography was within normal limits in both eyes. His Keratometry readings were 42.9D and 45.1D in the right and 44D and 45.2D in the left. Axial lengths were normal at 22.03mm and 22.04mm in the right and left respectively.

Anterior Segment Photograph (Right Eye)



Anterior Segment Photograph (Left Eye)

Anterior Segment Ocular Coherence Tomography (Right And Left Eye)



Surgical procedure

He had bilateral clear lens extraction with PC IOLs inserted. A superior scleral tunnel was made and AC entry made. Anterior capsulorrhexis was done using the capsulorrhexis forceps. Hydrodissection was done and the nucleus and cortical matter removed by irrigation and aspiration using a simcoe cannula. A Polymethyl methacrylate lens was inserted into the bag. Subconjunctival dexamethasone and gentamicin were given post operatively. He was placed on Topical Moxifloxacin and Dexamethasone eyedrops and ointment, tailed off over a 6week period.

Results

Unaided VA was 6/12 in either eye on the first post-operative day with quiet anterior segments and stable IOL position in the bag. Final BCVA was 6/6 in either eye with refraction of Plano–0.75 cylinderx180 and Plano-0.50cylinderx180 in the right and left respectively by the fourth postoperative week. Visual acuity has been maintained for 2 years of follow up so far, and no post-operative complications have occurred.

Discussion

Alport syndrome is a rare, genetic inherited disorder characterized by deficiency in mature collagen 4∝3∝4∝5 subunits in basement membrane structure of the eye, ear and kidneys.[2] This occurs due to mutations affecting the COL4A5 gene, which codes for Collagen 4. There is a persistence of immature (embryonic) Collagen 4∝1∝1∝2 in kidneys (Glomerular membrane), Cornea (Bowman and Descemet membrane), Retina (Internal limiting membrane and nerve fibre layer), Lens (anterior lens capsule) and inner ear (stria vascularis). This results in weakness which manifests as deafness, early-onset renal failure, corneal erosions and posterior polymorphous corneal dystrophies, central retinal flecks and peripheral thinning, and anterior lenticonus, a forward bowing of the anterior lens capsule centrally with resultant induced index myopic astigmatism,. This severely reduces visual acuity, even with spectacle wear. Less commonly, posterior lenticonus may occur.[3-5]

Affected males usually by X-linked inheritance from their mothers, usually have severe manifestations with early onset renal failure whereas females exhibit relatively mild disease with only microscopic heamaturia and normal renal function.[2]

Progressive myopia in Alports syndrome is amenable to treatment by clear lens extraction with intra ocular lens implantation, documented to be undertaken successfully mainly by phacoemulsification with foldable intraocular lens implantation.[6-10] In this case study, extraction was safely done by irrigation and aspiration through a scleral tunnel. This was enabled by the softness of the nucleus, being of a peadiatric age patient. However nucleus removal through a scleral tunnel in an adult would be a good option when phacoemulsification is either unavailable or unaffordable, as is the case in many developing nations. Implantation of a PMMA lens rather than a foldable one significantly cut dowm on cost of

surgery. In addition, the process of capsulorrhexis on a fragile anterior capsule may be challenging. The inherently fragile capsule develops partial splits that may rupture. Mohan has described a flower-petal rupture pattern with extension of the anterior capsulorrhexis and subsequent posterior capsular rupture.[7] Ecsedy et al successfully performed cataract surgery in two patients with anterior lenticonus, Femto-laser technology for anterior capsulorrhexis.[8] However anterior capsulorrhexis was successfully done in this patient utilizing the conventional utrata forcep cautiously. This is important, as the cost of Femto laser procedures are likely to be unaffordable for most people in developing nations. His corrected visual acuity improved significantly with a reduction in his degree of myopic astigmatism after lens removal. Hence, a carefully performed lens aspiration with implantation of hard lens through a sclera tunnel is a viable option in developing nations and low resource settings.

Conclusion

Clear lens extraction with PMMA IOL implantation can produce excellent refractive outcome and visual results in Alport syndrome.

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