Case Report

Management of Aniridia with Ectopia Lentis

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We report a case of bilateral congenital aniridia with ectopia lentis and cataract in a female child of 14 years old in whom artificial iris and IOL complex was transplanted to attain cosmetic results and somehow visual outcomes. The surgery was successful in achieving desired outcomes.

Key words: Aniridia, artificial iris, artificial iris.

Aniridia is characterized by complete or partial iris hypoplasia, usually involving both eyes, with neonatal onset. May be with systemic association or isolated which is not limited to a defect in iris development, but is a panocular condition with macular and optic nerve hypoplasia, cataract, and corneal changes (often later onset).[1]

Aniridia may be familial or sporadic. Mutation on the PAX_6 gene on chromosome 11 is responsible for causing aniridia.[2]

A more recent study from Denmark reported the incidence as 1 per 96,000 live births with no gender and racial predilection.[2]

Vision may be severely compromised due to associated ocular complications such as: nystagmus, amblyopia, buphthalmos, and cataract.[1]

Very few lens opacities in infancy require lens extraction, but visually significant lens opacities eventually develop in 50 – 85% of affected individuals, often in the teens or early adulthood. Lens subluxation or dislocation occurs but is uncommon (in 18 – 35%), Nystagmus occurs in 85 – 92% of patients and glaucoma present in 70% cases.[2]

Symptoms of aniridia are treated accordingly and surgical intervention is carried out for cosmetic results which include transplantation of iris. It is also available in combination with intraocular lens (IOL) for the combine treatment of aphakia and aniridia.

CASE REPORT

A 14 year old female child resident of Quetta came to Hashmanis Hospital presented complaints of decreased vision and sensitivity to light (photophobia) with involuntary eye movements.

On examination unaided visual acuity was found to be counting finger at 1m in both eyes. There was no improvement with pinhole. Slit lamp examination revealed the absence of iris with opaque and subluxated lens in both eyes hence aniridia was diagnosed, associated with nystagmus, cataract and ectopia lentis. Relevant systemic examinations were carried out but no systemic association was found.

Surgical plan including artificial iris implant with IOL under general anesthesia was prepared with patient's consent to maximize cosmetic appearance and improve visual outcome. Prior to surgery IOL power was calculated and color of iris was chosen. Biometric calculations showed the resulting IOL powers of +31D and +32D for right and left eye respectively and green color of iris was chosen as per patient's choice. Both eyes were operated with a gap of three days.

Single piece artificial iris +IOL complex was used manufactured by Reper – NN Russia. The complex is made of the optic part with the diameter of 3.5 mm and painted peripheral part. The diameter of the block was 13 mm.

Post-operative cosmetically outcomes were remarkable along with improved visual status with decrease glare problem and photophobia. The best corrected visual acuity was improved to 6/60 in both eyes. The patient is happy and satisfies with the results.

The patient is kept on follow up to monitor intra ocular pressure and other associated conditions.

Fig. 1: Pre-Op. iris + complex implanted in both eyes.

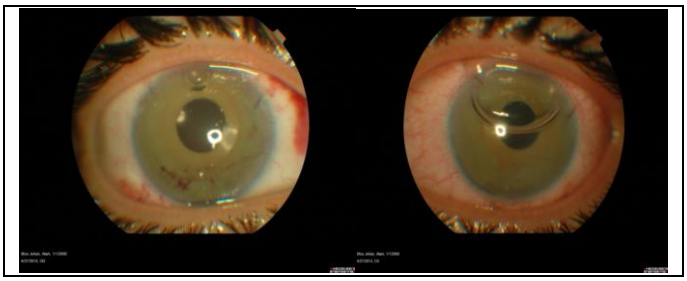

Fig. 2: Post-Op. iris + IOL complex implanted in both eyes

Surgical procedure:

- The eye was fixed by using a superior rectus suture. It was planned to secure the artificial iris + IOL complex with three sutures at 10, 2, and 6 O′ clock positions. The cornea was marked in clock wise fashion with a radial marker to ensure accurate suture placement for lens centeration. Conjunctiva and Tenon's were disected at 6 O′ clock position and sclera was exposed. A partial thickness, lamellar scleral flap was created at 6 O'clock position. Two incissions of 3.2 mm length were made at 2 O'clock and 10 O'clock positions. The Lensectomy and anterior viterectomy were done.
- Both the wounds were secured with sutures. The artificial iris + IOL complex was then prepared for implantation. Three sutures of, 10 - 0 polypropylene were tied in a fashion to be fixed in the eye at 10, 2 and 6 O'clock positions. A 5.2 mm limbal incission was placed superiorly and anterior chamber was reformed using a visco elastic. The long needle of suture was passed through superior incission and reterieved from 6 o′ clock position. The artificial iris + IOL complex was folded and implanted in ciliary sulcus. The inferior 6 O′ clock suture was pulled and the lens was fixed at 6 O′ clock position. For the superior fixation of the complex the needles were bent and passed from inside out at 10 and 2 O'clock positions and were fixed there. Superior incissions and scleral flap at 6 O'clock position were closed using 10-0 nylon sutures. The conjunctiva and Tenon were closed using 8 - 0 virgin silk. The residual viscoelastic material was removed by irrigation. Dexamethasone and tobramycin were injected subconjunctivaly and the eye was patched. Post operatively the patient was seen after 24 hours, 3 days, 1 week, 2 weeks and after 1 month. The patient was asked for a follow up check up at three monthly interval for a year and then to be seen every year. Post operatively she was given topical steroids and topical antibiotics 4 to 6 times a day , which gradualy tapered of over a period of 6 to 8 weeks.

DISCUSSION

Iris implant surgery is a new modality treatment for people in whom iris could not develop normally or who lack natural iris (for e.g: in conditions of aniridia). Though these patients who undergone such procedure are also at the risk of complications but the benefits of gaining an iris may outweigh their risks.[3]

The artificial iris is a thin prosthesis made of the silicone which is also used for intraocular lenses that is very flexible and can be folded and inserted into the eye by making a peripheral corneal surgical incision, about 2.8mm in length.[4]

The first artificial iris implant was introduced in the 1980s for patients with a cataract and an intact capsule while the first implantation under FDA clinical trial was done in US in 2002.[5] Since then it has been used in many patients in whom there was cosmetic disfigurement due to absence of iris.

A study in march 2000 evaluated the long term results of implantation of a black diaphragm intra ocular lens (IOL) in eyes with congenital aniridia which carried out a 46 months follow up and reported

improvement of visual acuity in 73.68% (14 of 19 eyes) However, post-operative problems were glaucoma (2.73%), cystoid macular edema (18.18%) and chronic endothelial cell loss (27.27%).[6] A study from Pozedyeva NA et al, also reported improvement in corrected visual acuities for 75% eyes. Post-operative complications include 2 cases of hyphema, 1 case of ciliochoroidal detachment, 4 eyes with exaggerated immediate post-operative reaction and 1 eye with persistent low grade cyclitis out of 20 eyes under study.[7]

A study by I Mavrikakis et al reported the efficacy of prosthetic iris devices along with IOL in patients with iris deficiencies. The best corrected visual acuity found to be improved in 90% of patients while glare reduced in 80% of patients.[8]

A study in European journal of ophthalmology, 2011 describes the efficacy of elastic artificial ILD (iris-lens diaphragm) from Reper – NN for combination of aphakia and iris defects of various extent.[9]

Iris implantation is not much common in Pakistan because of very low incidence of aniridia and low socio economic conditions.

CONCLUSION

Artificial Iris + IOL transplantation provides good management option for aniridia patients either congenital or traumatic as it restores cosmetic appearance and resolve visual associated symptoms so it should be promoted by surgeons and made available to the patients.

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REFERENCES

1. **Hingorani M, Moore A.** Aniridia. 2003 May 20 [Updated 2013 Nov 14]. In: Pagon RA, Adam MP, Ardinger HH, et al., editors. Gene Reviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2014. Available from: http://www.ncbi.nlm.nih.gov/books/NBK1360/.
2. **Bakri S, Simon JW, Cibis GW.** Aniridia in the Newborn Clinical Presentation, Medscape[internet] Dec 3, 2013 [cited on 2nd July 2014] available from: http://emedicine. medscape.com/article/1200592-clinical#a0217.
3. Cosmetic Iris Implants: Risks Outweigh Benefits, eye smart [internet], updated on Nov-01-2013 [cited on 05-july-14]
4. Artificial iris implant surgery, Vistar eye center [home page on the internet], Virginia [cited on 05-july-2014]
5. Jules Stein Eye Institute, Clinical Update JANUARY 2011 Vol. 20|NO. 1 [cited on 06-July-2014].
6. **Reinhard T, Engelhardt S, Sundmacher R.** Black diaphragm aniridia intraocular lens for congenital aniridia: long-term follow-up. J Cataract Refract Surg. 2000; 26 (3): 375-81.
7. **Pozdeyeva NA, Pashtayev NP, Lukin VP, Batkov YN.** Artificial iris – lens diaphragm in reconstructive surgery for aniridia and aphakia. J Cataract Refract Surg. 2005; 31: 1750–9.
8. Mavrikakis I, Mavrikakis E, Syam PP, Bell J, Casey JH, Casswell AG, Brittain GP, Liu C. Surgical management of iris defects with prosthetic iris devices. Eye (Lond.) 2005; 19: 205-9.
9. **Pozdeyeva NA**, **Pashtayev NP**, **Viktorova EA, Treushnikov VM**, **Starostina** OV. Artificial iris – lens diaphragm design modifications for surgical correction of iris defects combined with aphakia, Eur J Ophthalmol. 2011; 00(00).