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Case Report

Combined cataract phacoemulsification and aniridia endocapsular rings implantation in a patient with bilateral congenital aniridia and cataract: A case report

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Abstract

The objective of this article was to determine the long-term outcome of surgical treatment in a patient with bilateral congenital aniridia and congenital cataracts. The patient was treated by cataract removal and implantation of a single piece IOL in both eyes. Two aniridia rings were also implanted in the capsular bag. The best-corrected visual acuity (BCVA), intraocular pressure (IOP), stability of the intraocular lens (IOL) and subjective glare reduction were measured for two years after the surgery. After 10 months, the IOP in the left eye had increased to 26 mmHg despite the fact that anti-glaucoma medication was added. Therefore, the patient was scheduled for an Ahmed valve implantation. These results suggest that good visual outcomes can be achieved in patients with bilateral congenital aniridia and cataracts. Nevertheless, the IOPs must be continuously monitored, and glaucoma screening performed to prevent further complications.

Keywords: Congenital aniridia, Cataract, Aniridia ring, Corneal leucoma

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Introduction

Bilateral congenital aniridia is a rare disorder and is characterized by a partial or total absence of the iris associated with foveal hypoplasia which leads to a reduction of the visual acuity and nystagmus from an early age.[1] The prevalence of congenital aniridia is 1:64,000–1:96,000, and higher rates have been reported in patients who are younger than 20 years (1:47,000).[2] Congenital aniridia is associated with defects of the cornea, anterior chamber, iris, lens, retina, macula, and the optic nerve. Approximately, 85% of individuals with aniridia inherit it as an autosomal dominant trait and 13% as part of the autosomal-dominant WAGR syndrome characterized by Wilms' tumor, aniridia, genitourinary abnormalities, and mental retardation. The remaining patients are associated with other syndromes such as Gillespie syndrome and Peters anomaly.[3] Mutations or intragenic deletions of the *PAX6* gene represent the major causes of aniridia.[4]

Abnormalities frequently associated with aniridia include cataracts, glaucoma, and opacifications of the cornea.[5] Keratopathy develops secondary to limbal stem-cell deficiency and is believed to have an incidence of about 20%. Aniridia associated with glaucoma has a reported incidence of 6 to 75% with 91% of these primarily due to angle and trabecular developmental abnormalities. These patients respond poorly

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to treatment resulting in blindness due to failure of pharmacological therapy, and these patients require frequent glaucoma surgery to achieve an adequate control of the intraocular pressure (IOP).[5–7]

Congenital aniridia is associated with cataracts in 50–85% of patients.[5] Lens opacities combined with the lack of the irides result in glare, photophobia, and reduced visual acuity. However, cataract extraction presents significant technical challenges including reduced visibility of the intraocular structures due to corneal opacities and fragile anterior lens capsule due to a significant thinning of the capsule.[8] This fragility may cause tears in the anterior capsular either during capsulorhexis or by subsequent manipulations. Such complications may require significant alterations of the surgical plan because intraocular lenses and iris prosthetic devices require an intact capsular bag.[9]

Among the options for prosthetic restoration of the irides are devices which allow the implantation of Morcher aniridia rings through an incision of about 3 mm. The aniridia rings are capsular tension rings with multiple fins separated by the same size spaces. Two such rings are implanted into the capsular bag and placed in such a way that the fins of one ring overlap the spaces of the other.[10] Combined phacoemulsification cataract extraction with an implantation of a foldable intraocular lens and aniridia rings placement have been shown to be particularly effective in reducing glare and photophobia in patients with aniridia and albinsm.[10,11] We present our findings in a patient with bilateral congenital aniridia who was successfully treated with an implantation of an IOL and aniridia rings.

Case report

A 26-year-old man presented with complaints of a progressive decrease in his vision and glare in both eyes of 8 months duration. His medical history revealed that his father was diagnosed with open angle glaucoma. The patient's best-corrected visual acuity (BCVA) was 1.0 logMAR units in both eyes. Horizontal pendular nystagmus was present in both eyes. The IOP measured by Goldmann applanation tonometry was 12 mmHg OD and 14 mmHg OS.

Slit-lamp examination of the right eye showed a 360° peripheral corneal vascular proliferation, inferior corneal leucoma, superficial punctate keratitis demonstrated by diffuse epithelial staining of the central cornea with fluorescein, total aniridia, and a NO2NC2 cataract according to the Lens Opacities Classification System II (LOCS II).[12] Slit-lamp examinations of his left eye showed a 300° peripheral corneal vascular proliferation, mild superficial punctate keratitis, total aniridia, and a NO2NC2 cataract (Fig. 1). A fundus examination revealed absence of macular reflex and a normal optic disk in both eyes. Specular microscopy showed an endothelial cell density of 3289 for the right eye and 2906 cells/mm^2 for the left eye.

Small incision (2.2 mm) phacoemulsification was performed with an implantation of an acrylic foldable intraocular lens (Alcon Laboratories, Inc., Fort Worth, TX, USA). Additionally, a Morcher type 50C endocapsular aniridia ring was placed (Morcher® Type, Morcher Company, Germany), and both the IOL and the aniridia rings were inserted in the capsular bag in both eyes by the same surgeon with an interval of two weeks.

After two months, the BCVA improved to 0.89 logMAR units in both eyes with no signs of inflammation, and the BCVA in the right eye improved to 0.69 logMAR units at the 6 months visit; additionally, a subjective mild improvement in glare perception was referred (Fig. 2).

At the 10th month visit, the patient complained of decreased vision in the left eye (1.3 logMAR units) and was found to have an elevation of the IOP to 26 mmHg. There was also a displacement of one of the rings due to capsular fibrosis and contraction without IOL displacement (Fig. 3). Combined topical dorzolamide and timolol treatment was initiated in the left eye, and at the one year follow-up, the IOP was not significantly decreased despite the fact that a third anti-glaucoma medication was added. Therefore, the patient was treated with an implantation of an Ahmed glaucoma valve in the superotemporal quadrant and the removal of the displaced ring.

Six months later, the BCVA was 0.92 logMAR in the right eye, and the IOP in the left eye was maintained at 14 mmHg with topical timolol.

Discussion

Cataract is found in 50–85% of the patients with congenital aniridia. Phacoemulsification cataract extraction in patients with aniridia will improve in the BCVA[8–10]; nonetheless, the surgical procedure is often challenging due to abnormalities associated with congenital aniridia, including shallow chamber, corneal leucoma and limited visualization. In addition, the outcome can be limited by a worsening of the keratopathy or the development of glaucoma in more than 50% of the cases.[13] Recent studies have demonstrated a significant association of intraocular surgery and progression of corneal opacities. Moreover, Reinhard et al.,[14]

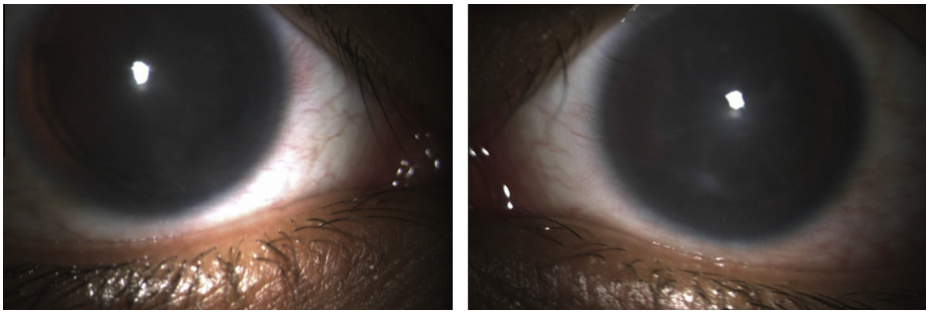

Fig. 1. Slit-lamp photographs of the right and left eyes showing 300° peripheral corneal vascular proliferation, mild superficial punctate keratitis, total aniridia, and cataracts.