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Background:

Congenital aniridia is often accompanied with cataract formation and corneal scarring with age. In this case a simultaneous reconstructive procedure in the anterior segment would be desirable in order to improve visual function and, consequently, the quality of life of a suffering patient.

Methods:

In a male 61 years old patient (visual acuity: HM/LP, IOP = 12/14 mmHg (OD/OS)) congenital aniridia complicated with photophobia, amblyopia, partial ptosis, corneal scarring with neovascularisation, Brunescens cataract, slight limbal deficiency and glaucoma was diagnosed in both eyes. The left eye of the patient was undergone the simultaneous quadruple surgery, consisting of perforated keratoplasty (diameter of 9 mm), extracapsular cataract extraction, intracapsular implantation of 2 aniridia rings, forming an artificial iris diaphragm (50F, Morcher) and intracapsular IOL implantation (AR40E, AMO).

Results:

Early and late postoperative periods run on without any complications. 5 months postoperatively the visual acuity far and near amounted to 0,05/0.1 (OD/OS) without correctors, which was the best visual acuity that ever was measured in this patient. Photophobia was decreased. Ptosis was reduced in comparison to the non operated eye. A similar procedure in the conjugate eye is planned 12 months after the first surgery. The reported surgery allows to avoid additional sulcus- or scleral IOL and iris diaphragm fixation. The possibility of secondary intraocular hypertension might be reduced thereby.

Conclusions:

The reported case confirms the possibility of successful rehabilitation of patients with congenital aniridia with cataract and corneal scarring by means of the quadruple procedure. The procedure can be recommended in complicated aniridia in case of sufficient capsular support and mild limbal deficiency. It might be performed even in case of partially damaged or slightly weak zonula since aniridia rings act also as a capsular ring providing additional stabilization of the capsular bag.

BACKGROUND:



congenital aniridia
+
age



cataract formation
corneal scarring
glaucoma



Methods of surgical treatment proposed earlier elsewhere:

corneal scarring
+
cataract
+
aniridia



keratoplasty
+
cataract extraction
+
implantation of an IOL
with iris diaphragma
and sulcus fixation (e.g.
OPHTEC, Morcher)

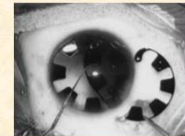


Miller AR et al., J Cataract Refract Surg. 2007
Phillips PM et al., J Cataract Refract Surg. 2008

cataract
+
aniridia



cataract extraction
+
implantation of two
aniridia rings and an
IOL in the capsular bag



Osher RH & Burk SE,
J Cataract Refract Surg. 1999

Our idea of surgical treatment: combination of both methods

corneal scarring
+
cataract
+
aniridia
+
glaucoma

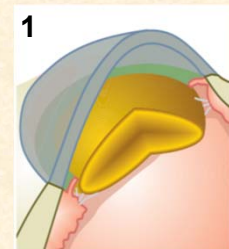


keratoplasty
+
extracapsular cataract extraction
+
implantation of two aniridia rings
and an IOL in the capsular bag

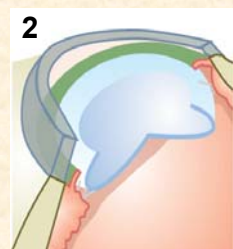


METHODS:

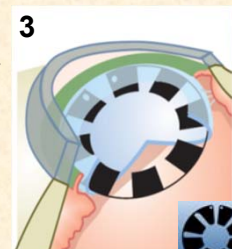
Surgical technique: simultaneous quadruple surgery



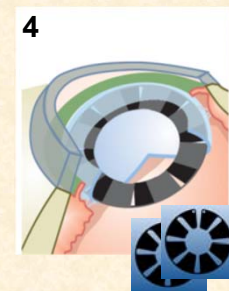
1 patient's eye before the surgery



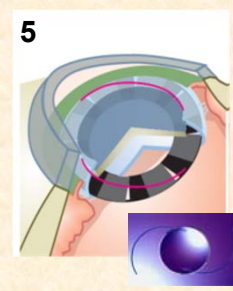
2 cornea is trepanated, extracapsular cataract extraction is performed



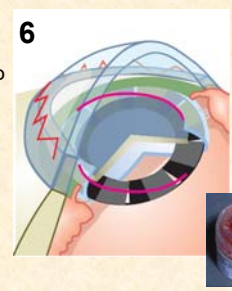
3 1st iris diaphragm (50E, Morcher) is implanted into capsular bag



4 2nd iris diaphragm is implanted into capsular bag and rotated towards the 1st one to create complete diaphragm

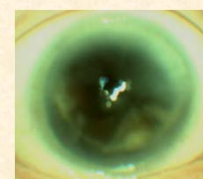


5 IOL (AR40E, AMO) is implanted into capsular bag

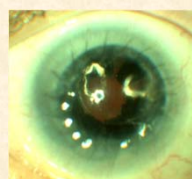


6 corneal transplantation is performed

RESULTS (one-year follow-up):



pre-operatively



post-operatively



preoperatively the palpebral fissure in the left eye was similar to those in the right eye

- early and late postoperative periods run on without any complications
- 12 months postoperatively UCVA far and near amounted to 0.05 (20/400) and 0.1 (20/200) respectively
- IOP: 16 mmHg (OS) under medication
- photophobia was decreased
- palpebral fissure becomes larger in comparison to the non-operated eye, which might be due to decreased photophobia