Cataract surgery and aniridia

Article *in* Current Opinion in Ophthalmology · November 2009

DOI: 10.1097/ICU.0b013e328333ea49 · Source: PubMed

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Current Opinion in Ophthalmology 2010, 21:60–64

Purpose of review

In the past years, several new options have been developed for the surgical management of aniridia in the course of cataract surgery.

Recent findings

The lens capsule may be altered – thinned – in aniridia, requiring particular attention in order to avoid complications with possible consequences for the surgical plan. Iris prosthetic devices for complete or partial restoration of an iris diaphragm have been developed and their use described. There are options for intracapsular placement as well as for fixation without using a capsular bag and options for improved cosmetic appearance have been created. Corneal tattooing is still an option for selected cases and has been refined technically. Complications attributable with the iris prostheses are relatively infrequent and manageable.

Summary

Cataract surgery offers an opportunity to manage associated partial or (sub)total aniridia of all origins with good to excellent functional and esthetic results with a relatively low and manageable complication potential.

Keywords

aniridia, artificial iris, iris diaphragm lenses, iris prosthetic devices

Curr Opin Ophthalmol 21:60–64

1040-8738

Introduction

Aniridia denominates the total missing or absence of the iris. In the strict sense of the term, that is, aniridia with no remaining remnants, aniridia is extremely rare. Clinically, aniridia is also used, when only minimal peripheral remnants of iris tissue can be identified, for example with gonioscopy.

The terms 'total', 'subtotal' and 'partial' aniridia are not sharply defined; they are used quite intuitively. Aniridia is usually called 'total', when no remnants of the iris can be seen with regular slit lamp examination, 'subtotal' when minimal iris remnants of less than 2–3 clock hours can be seen. With more iris remaining, aniridia is usually termed 'partial'.

Aniridia can have two main causes, both associated to a high degree with cataract.

Traumatic aniridia can be caused by avulsion of the iris in the course of a penetrating eye injury. The – fortunately – rare cases of iatrogenic aniridia by accidental removal of the iris as a complication of a surgical intervention are to be seen in this category.

Due to the invasive nature of trauma causing aniridia, associated (traumatic) cataract with the need for surgical intervention is frequent.

Congenital aniridia is a rare disease with an incidence of 1 : 64 000–100 000, caused by a variety of mutations in the paired box gene 6 (*PAX6* gene) on band p13 of chromosome 11. It is a 'panocular' disorder, affecting cornea, iris, lens, retina and the optic nerve. It may occur in isolation or be associated with other syndromes. Two-thirds of cases are familial, one-third is sporadic, but then passed on in an autosomal dominant way with variable expressivity. It is a profibrotic disease, which has implications for wound healing after surgical interventions. Lee *et al.* [1••] have recently published an updated overview.

Congenital aniridia is associated with cataract in 50–85% patients [2].

Other rare causes include irido-corneal-endothelial (ICE) syndrome, essential iris atrophy and secondary iris (sphincter) atrophy, such as after massive intraocular pressure increases.

For the present review, the literature from 2005 onward was reviewed and all articles found with contents relevant to the topic of this review were evaluated.

The consequences of aniridia for cataract surgery may be as follows:

(1) Reduced visibility of intraocular structures due to corneal opacities

(2) Fragile capsule
(3) Prosthetic replacement of an – artificial – iris
(4) Combined procedures for associated problems.

Corneal opacity

No report addresses this problem specifically in conjunction with aniridia. Oshima *et al.* [3] describe an elegant technique to facilitate cataract extraction in presence of severe corneal opacities in general. This technique can certainly be applied to corneal opacities as associated with aniridia of congenital, but also of traumatic or other origin.

It is obvious, that all visualization enhancing measures are to be considered, such as capsular staining, oblique illumination or endoillumination.

In congenital aniridia, it is always worth to try cataract extraction first, as long as visualization is considered sufficient, that it can be performed safely, because the options for restoration of corneal clarity are certainly considerably more demanding and carry a realistic risk for complications and failure.

If corneal clarity is not sufficient for safe performance of cataract surgery or if there is a doubt, the risk for complications outweighs the – limited – potential benefit. Then limbal stem cell transplantation is the best first approach; its result will guide the ensuing decisions.

Capsular fragility

In some eyes with congenital aniridia, the anterior capsule has clinically – intraoperatively – been noted to be particularly fragile [4]. The findings of the original report have been confirmed by Hou *et al.* [5]. The histological substrate is significant thinning of the capsule when compared with capsules from nonaniridic patients.

As some aniridic patients do not have such capsular fragility, their capsules being within the normal range for capsular thickness, it can at this time not be assessed, if this fragility is case or age dependent.

This fragility may cause tears in the anterior capsular opening either during capsulorhexis or by subsequent manipulations, even when intact and continuous at completion of capsulorhexis, with their consequences for capsular integrity [6]. As some iris prosthetic devices necessitate an intact capsular bag, such complication may require a significant alteration of the surgical plan [4].

Suggestions include careful performance of capsulorhexis with the mentioned fragility in mind, using supporting techniques, such as dispersive viscoelastic, capsular staining and keeping the diameter on the smaller side.

This author instead favors larger openings, always within the limits of what is achievable in the individual situation. The profibrotic nature of congenital aniridia together with the consideration, that optic overlap is of reduced, if any, concern with the concomitant implantation of iris prostheses would seem to make larger openings preferable in avoiding excessive rhexis contraction and enabling less stress on the rhexis margin with manipulations associated with lens extraction and lens and prosthesis implantation.

In aniridia, most naturally, the main issue in conjunction with cataract surgery is reconstruction of an optic aperture.

Corneal tattooing

Corneal tattooing has been used for many decades, but never more than sporadically. New techniques have recently been described. Aliò *et al.* [7] describe the use of new mineral micronized pigments; Kim *et al.* [8•] have developed a technique using lamellar keratotomy with a femtosecond laser combined with extended manual dissection, where needed, to place the intracorneal pigments Two recent series – one with 14 cases [9], one with three cases [10] – have been published in France. All describe good tolerance, no serious complications (except one late perforation in an extremely pathological cornea) and good anatomical and cosmetic outcome, as well as functional improvements within the limits of the underlying condition.

Corneal tattooing is used in these, as in previously described, cases mostly in severely compromized eyes, in the great majority for posttraumatic aniridia of varying extent. The main purpose is attenuation of photophobia, cosmetic disfiguration or even occlusion for intractable diplopia in eyes with little residual function.

It has the main advantage of being noninvasive in the sense of not intraocular and being possible without lens removal or necessitating adequate intraocular visibility, as with intraocular surgical options. In the context of cataract surgery with aniridia it will therefore be of marginal importance only.

Prosthetic iris replacement

As mentioned above, the usage of the term 'partial aniridia' is not sharply defined. For the purpose of this overview, the authors define it as the missing of the iris to an extent, which does make reconstruction by other than prosthetic means impossible/insufficient.

In the past years, a number of prosthetic iris devices have been developed.

(1) Iris diaphragm lenses (Morcher, Stuttgart, Germany; Ophtec, Groningen, The Netherlands; proprietary designs)
(2) Segmental iris prosthetic devices [iris segment rings by Morcher; Hermeking Iris prosthetic system (IPS) by Ophtec]
(3) The artificial iris by Dr Schmidt Intraocularlinsen GmbH, St Augustin, Germany.

The iris diaphragm lens

First developed in 1991 and described in 1994 [11], it has gained increasing acceptance and international usage.

It consists of a plate of black polymethylmethacrylate (PMMA) with a plate diameter of 10 mm, with curved haptics, with or without fixation loops. It provides a central opening of variable diameter with or without the inclusion of an optically functional center.

Since its first description, different models have been developed for various different situations and needs.

The black diaphragm can be basically obtained for intracapsular and extracapsular implantation, with and without loops on the haptics for facilitation of suture fixation, different diameters of optic/'pupil' and haptic and different configuration to deal with partial and (sub)total aniridia.

Models with different diaphragm configuration and with different colors are offered by Morcher and Ophtec, both basically using comparable fixation haptics of a C/J-loop type.

Three recent case series with such diaphragm lenses have been published [12•,13•,14].

The vast majority of cases are of traumatic origin. They all show good success functionally, both with regard to photophobia and vision, taking the wide variety of preexisting associated traumatic alterations other than those of lens and iris into account. The alterations associated with congenital aniridia, such as glaucoma, corneal opacity and foveal hypoplasia limit the visual acuity outcomes in this group, so that the according result of Aslam *et al.* [2] is not surprising.

Complications reported are – not surprisingly, when considering the heterogenous composition of cases – of a wide variety, concerning almost all intraocular structures. Glaucoma is a frequent complication; whether it is caused or at least contributed to by the iris diaphragm portion of the implant itself is discussed by one author [2]. No complication with severe or complete permanent visual loss attributable to the implant has been reported.

An iris diaphragm lens with a different haptic design has been reported from Russia. It has a multipoint haptic, is foldable and comes in four different colors. Results of 20 eyes (19 patients) are described in detail and are functionally mainly dependent on preoperative abnormality. There were no complications leading to permanent visual loss [15].

The Artisan (Ophtec) iris reconstruction lens is a custom made PMMA-IOL with a PMMA-iris diaphragm, containing two or three 'lobster claw' fixation devices for fixation at residual iris. It is therefore only suitable for cases of partial aniridia with enough iris tissue left for enclavation of the haptic portions. Sminia *et al.* [16•] have published a series of five pediatric trauma cases. In all of them, implantation was secondary after previous trauma repair and extraction of traumatic cataract. Cosmetic and functional results were within the limits of the preexisting abnormality; complications could not be specifically attributed to the implant. The length of follow-up (5–12+ years) makes this small series of five cases noteworthy.

Segmental iris prosthetic devices

Although the iris diaphragm lenses require an incision size of a minimum of 10 mm for a full diaphragm, as needed in (sub)total aniridia, another option for prosthetic restoration of an 'iris' are devices, which allow the implantation through incision sizes of around 3–4 mm. They do, however, require an intact capsular bag for implantation.

Morcher aniridia rings are capsular tension rings with multiple fins, separated by 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. A variant of this principle is an iris segment ring for the bridging of segmental iris loss ('partial aniridia').

The Hermeking prosthesis consists of elements of about 90°-segments, either double or single, for intracapsular or sulcus implantation; the double elements are 180° apart and connected with a spring-action, flexible PMMA rod; the single elements have haptic rod.

Two double elements are needed for a full iris prosthesis; they may be stabilized by a ring-clip element. For partial prosthetic replacement appropriate combinations of elements are used.

The recent literature contains a number of case series reports with these devices [14,17–20]. In these studies, to, the majority is in cases of traumatic aniridia, a few cases of congenital aniridia and ICE syndrome are also reported [17,18].

The results are very comparable with those of the diaphragm lenses, anatomic and functional results are within expectation in the vast majority of cases, as described above. There are no major complications described in those series; in some cases secondary repositioning may become advisable [17].

Artificial iris

A recent addition to the armamentarium is an iris prosthesis made from Gore-Tex material by Dr Schmidt Intraocularlinsen GmbH. It exhibits a surface structure resembling that of a natural iris and can be custom made to closely match the patient's iris (in which at least in part still existent). It is thin and rollable and can therefore be implanted through a small incision of 3–4 mm. It can be used as a complete iris prosthesis, implanted in the capsular bag or – usually suture fixated – in the ciliary sulcus.

It can also be used to close partial aniridic defects, by cutting out an appropriate segment from the artificial iris and suturing it to the edges of the defect in the patient's iris.

In the peer reviewed literature, the usage of this device is described and illustrated by authors in the 'Consultation section' of the *Journal of Cataract and Refractive Surgery* [21–24]. Otherwise, there have been anecdotal reports about the use of this system at different scientific meetings [25–27].

In existing anecdotal reports functional and anatomic, especially cosmetic, outcomes are reported as good, as the basic situation will permit. The surgical technique is under ongoing development. Among possible complications secondary glaucoma appears to occur with some frequency, especially with sulcus fixation of a full prosthetic diaphragm; techniques to reduce its incidence, such as circumferential notching of the outer margin of the implant, are under evaluation (H.R. Koch, personal communication).

In summary, the iris diaphragm lenses have their main indication in cases of total to subtotal aniridia, especially when sulcus fixation is mandated. The inclusion of the optical part in the implant is certainly an advantage over segmental capsular bag fixated prosthetic devices, which have to be placed under or over an additional intraocular lens. Their drawback is the requirement of a large incision (around 11 mm) and the practical impossibility to place them into a preserved capsular bag, due to their large diameter rigid plate (with exception of smaller diameter models for partial aniridia).

Segmental iris prosthetic devices have their best indication, when there is an intact capsular bag into which they can be implanted. They have the advantage of being implantable through small incisions, and can be custom selected in various ways in cases of partial aniridia. Their drawback is possible dislocation in the bag, resulting in only partial overlap of the elements with each other or with the iris defect, which can, however, in most cases be repositioned when diagnosed timely. They should be used when the capsular bag is perfectly intact and with segmental aniridia and when a small incision is of high priority; with friable capsules, such as in many cases of congenital aniridia, manipulations with the implantation may cause a capsular break, necessitating a change of the surgical plan.

Complications are common in cases with aniridia, but are mostly caused by the complicated nature of the cases, especially after trauma. A complication with a possible direct association with the implant may be glaucoma.

The artificial iris is a promising new device, the place of which will have to be established in further investigations.

Combined procedures

Associated problems with combined procedures are sporadically mentioned in the recent literature on the subject. In addition to the traumatic cases, in which combined measures with cataract extraction and iris replacement may be indicated by the underlying condition, there are two reports of such interventions combined with vitreoretinal surgery [28,29], and one with keratoplasty [30], all in trauma induced (partial) aniridia. Results and complications are within what is described for noncombined cases.

In this context, the statement of Lee *et al.* [1••], referring to congenital aniridia, may be repeated, 'Aniridia is a profibrotic disorder and as a result many interventions – including penetrating keratoplasty and filtration surgery – fail'.

In congenital aniridia, therefore, it appears advisable to err on the conservative side when considering combined procedures and to rather deal with one problem at a time.

Conclusion

Cataract surgery with aniridia is complicated by pathological alterations due to the underlying cause of aniridia. Challenges include corneal opacification, friable capsule and, above all, reconstruction of iris and pupil. In the past, a number of technical options have been developed for iris prosthetic replacement, each with specific advantages and disadvantages, which make differentiated indication mandatory. These options have greatly improved the possibilities for visual and anatomical rehabilitation, with

limited and controllable complication potential. Due to the great heterogeneity of the underlying conditions and associated abnormalities, a strongly individualized approach is needed and careful long-term follow-up is indispensable. Under such circumstances, however, good to excellent results can be obtained in these usually badly compromized eyes.

Acknowledgement

The authors have no financial interest with regard to the subject in this article.

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- • of special interest
- •• of outstanding interest

Additional references related to this topic can also be found in the Current World Literature section in this issue (p. 83).

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