Traumatic Aniridia

3

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Abstract

The aim of this chapter is to describe the nature of traumatic aniridia and the currently available therapeutic options.

Keywords

Eye injuries · Aniridia · Trauma

3.1 Introduction

The iris is a considerably vascularized smooth muscle sphincter covered with varying degrees of pigmentation. Its immediate and most evident function is to *regulate* the amount of light that enters the eyeball through the aperture of its central space, the pupil. Under high luminosity, the sphincter contracts and opens in dim conditions. Failure in this basic physiological effect results in glare and photophobia but also reduced depth of focus due to the loss diaphragm effect, much like the one used in pinhole visualization.

Alongside the main aid, the iris provides in the regulation of light entrance to the eye, and there are other more subtle but no less important aspects to iridian anatomy. Autonomic responses in the iris tissue cause it to dilate under stress or arousal situations alongside facial expressions and vascular dilatation (blushing); these involuntary responses carry much of the subtext in nonverbal communication, often being unnoticed under normal conditions but detected immediately under malfunction with an instinctive disgust response in the observer. The cosmetic importance of the iris is paramount to the patient and should thus be taken into consideration alongside functionality.

The Iris also has a particular property among the eye tissues in its ability to stick to damaged and inflamed structures generating synechiae which are undesirable to the ophthalmologist but act as a natural closing mechanism with tamponade and vascularization to the area essential if no medical treatment is available.

Partial or total traumatic aniridia constitutes an important challenge to the ophthalmologist, who must balance all of the aspects mentioned above with the different technical options for resolution on a case-by-case basis, as we will discuss in this chapter.

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H. Yan (ed.), *Management on Complicated Ocular Trauma*, Ocular Trauma,
https://doi.org/10.1007/978-981-16-5340-7_3

3.2 Definition of the Disease

Traumatic iris defects can be classified into four large groups:

Classification	Characteristics
Pupillary sphincter defects	Paralytic intermediate mydriasis.
Radial iridian defects	Wedge-shaped iridan tissue loss usually under 90°.
Iridodialisis	Iris separation at its base.
Aniridia	Large or total loss of iris tissue.

They can be produced by penetrating or blunt eye trauma. Iridian defects due to penetrating trauma can be repaired in the same surgical act or in a delayed manner. But as in all-penetrating ocular trauma, the primary objective is the hermetic closure of the eyeball; it might be advisable to defer the surgical repair of the iris to a second procedure when the best decision can be made with good visualization, the right tools, equipment, and properly trained personnel are present.

There is a wide range of possibilities in the resolution of iris defects. Some are easier and widespread, others more expensive or non-available due to government entities pending approval. These go from non-surgical to surgical approaches.

Non-surgical choices may offer the benefit of being reversible and economically accessible while usually providing only partial resolution to these cases. The use of spectacles might be an interesting recommendation, especially for patients until they receive their final surgical procedure since they can provide some alleviation to glare while masking the cosmetic issues. They may be simple sunshades, photochromatic glasses, and some patients might even prefer pinhole spectacles not only for their level of occlusion but also their added depth of field (Fig. 3.1). Cases with total aniridia and aphakia might choose to have a full occlusion with an opaque shade in the affected eye.

Another more invasive but reversible approach is the use of contact lenses. These have the bene-

Fig. 3.1 The use of slits and pinholes to reduce glare has been documented amongst the peoples of the north (left). Modern and commercially available examples exist available for the general public (right)

fit of tackling both optical solutions and cosmetic compensation while reducing the amount of light that enters the eye. They do, however, present the disadvantage of eye manipulation, tolerance requirement from the patient and a reasonably smooth cornea to be applicable.

Corneal tattooing (D Hirsbein [1]) provides an interesting alternative to intraocular surgery for localized iris defects and might even be a valid cosmetic approach, especially for patients without visual prognosis and severely damaged corneas since a trained physician might even imitate the characteristics of the fellow healthy eye with the use of various pigments. This is not, however, the best approach for full aniridia and also lacks the reversibility of the non-surgical methods.

In the repair of the ruptured sphincter of the iris, iridodialysis and in radial defects of the iris under 90°, the surgical technique of choice is suturing. Repair of the iris sphincter due to traumatic mydriasis using the loop technique with 10-0 polypropylene suture is the most used approach. The most popular surgical repair of radial defects of the iris and iridodialysis is the McCannel technique with 10-0 polypropylene double-stitched suture (Fig. 3.2).

In many cases of large defects of the iris, tissue retracts and becomes attached to the chamber angle, producing peripheral anterior synechiae, which can alter the flow of aqueous humor. Thus, through goniosynechialysis, in a careful way, the iris can be released for its subsequent iridoplasty with sutures. In those cases where repair of the iris is not possible due to great loss of iris tissue, there are different intraocular devices for its treatment.

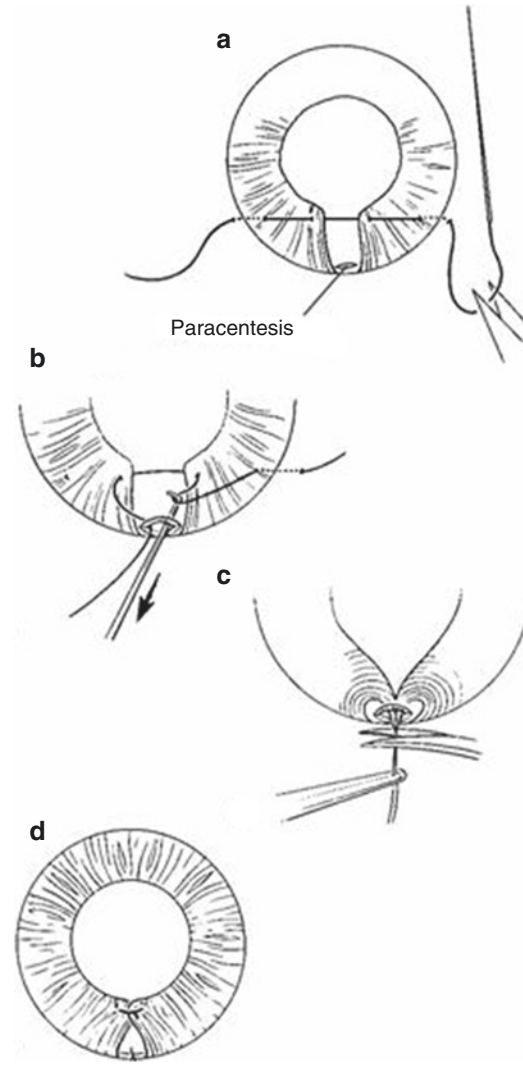

Fig. 3.2 The McCannel technique for repairing iris lacerations. With large lacerations, multiple sutures may be used. (a) A limbal paracentesis is made over the iris discontinuity. Then a long Drews needle with 10-0 polypropylene is passed through the peripheral cornea, the edges of the iris, and the peripheral cornea opposite, and the suture is cut. (**b**) A Sinskey hook, introduced through the paracentesis and around the suture peripherally, is drawn back out through the paracentesis. (**c**) The suture is securely tied. (**d**) After the suture is secure, it is cut, and the iris is allowed to retract. (Pending permission Reproduced with permission from Hamill MB. Repair of the traumatized anterior segment. Focal Points: Clinical Modules for Ophthalmologists. San Francisco: American Academy of Ophthalmology; 1992, module 1. Illustrations by Christine Gralapp)

3.3 Implants for Iris Defects

In 1964, Peter Choyce [2] reported the use of the first intraocular anterior chamber prosthetic iris device (PID). Clinically, PID can be categorized into three groups: (1) iris-lens diaphragm, (2) endocapsular capsular tension ring (CTR)-based PID, and (3) customized artificial iris [3].

There are several case series in the literature with follow-up for the treatment of large iris defects of different types, and their choice will depend on each case [4–6]. In the majority of penetrating or blunt ocular trauma in which there has been great damage to the iris, other intraocular structures will surely have been affected, such as the lens, chamber angle and retinal injuries. That said, most ocular trauma patients have or will develop a lens opacification and therefore, when deciding to implant an intraocular device for the treatment of large iris defects, the extraction of the lens is often essential. When deciding to treat an iris defect, we must evaluate the size of the defect to be corrected. As previously mentioned, radial defects smaller than 90° should, as far as possible, be sutured. For those cases where the iris cannot be sutured and for defects greater than 90° there are different devices for aniridia (partial or total) on the market, and the choice will depend on whether the patient is phakic, pseudophakic, or aphakic. Currently, there are several implants available to treat aniridia, as well as aphakia or cataracts, marketed by Morcher GMBH (Germany), HumanOptics (Germany) [7] and Ophtec Inc. (USA) [8] (see Fig. 3.3 below). All companies present different models of implants to treat iris defects.

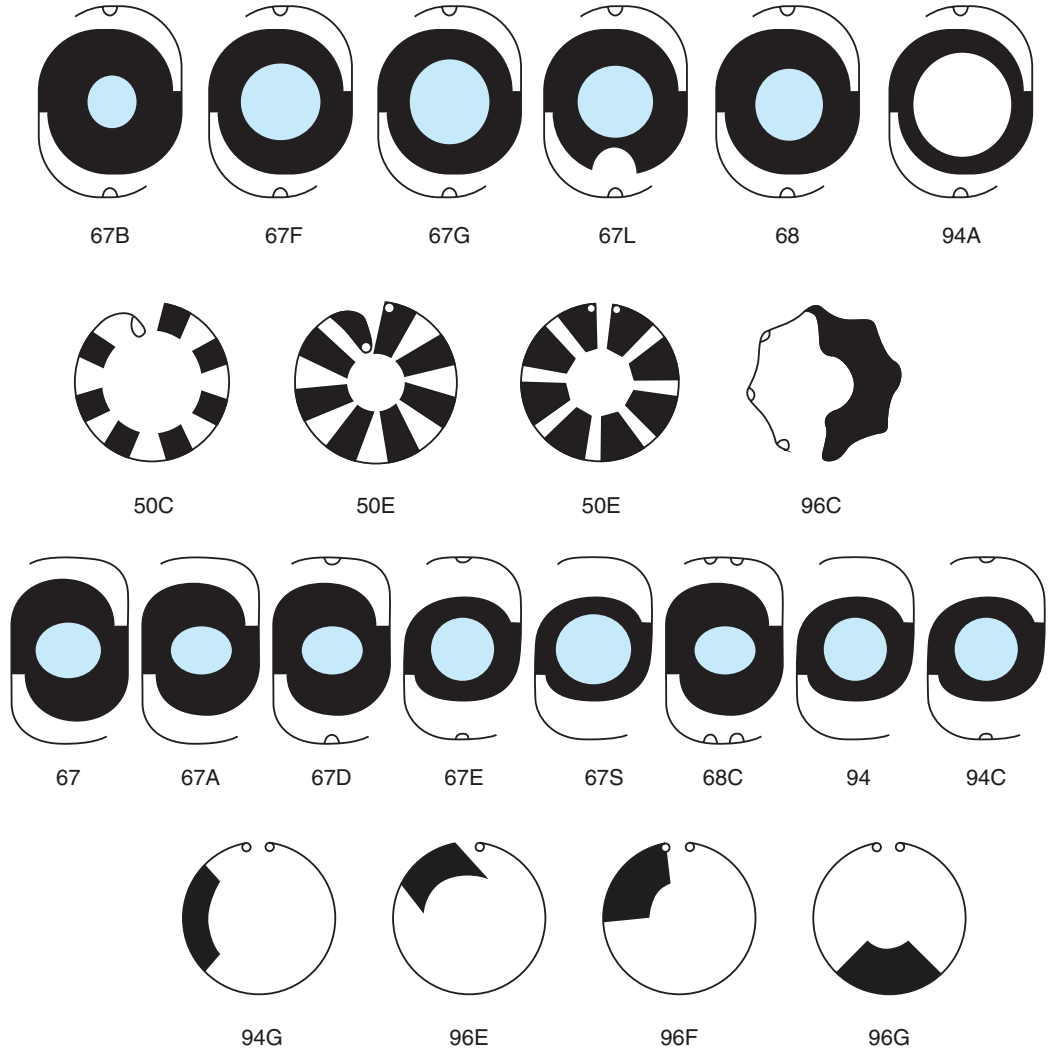

Fig. 3.3 Currently available models of aniridia implants from Morcher GMBH (Stuttgart, Germany) [9]

In phakic patients with iris defects under 180°, a surgical option is cataract surgery with routine intraocular lens implantation and placement in the capsular bag of one or two partial rings for aniridia with a 90° diaphragm (Morcher GMBH), positioning them in such a way as to cover the iris defect (see Case 2). Another option is to place a 180° diaphragm ring in the sulcus [10]. The size of the incision for the insertion of these devices will depend on the chosen model, ranging from 3 mm to 4.5 mm, approximately. In pseudophakic patients with iris defects, the same options can be chosen with the placement of one or two implants at 90° or 180°, depending on the iris defect to be covered.

In cases where aphakia occurs, there are several paths to choose from. It is possible to opt for the placement of an implant for aniridia with optics with a 360° diaphram of black PEMA sutured to the sclera (see Case 1). Ophtec USA Inc. has iris prostheses (with 360° diaphragms) of different colors with the possibility of incorporating a lens for optical correction through a 3.5 mm incision assisted by an injector. The HumanOptics CUSTOM FLEX artificial iris is a custom foldable iris prosthesis, without optics, for insertion in the posterior chamber sutured to the intraocular lens or sclera (see Case 3).

In all cases of partial or total iris defects, the choice of the type of implant and the surgical technique will depend on the ophthalmologist, as well as their experience in these cases.

3.4 Brief Case Reports

3.4.1 Case 1

Fifty-four-year-old male with work-related blunt ocular trauma to his OS 2 years prior with sphincter iris rupture, generalized iris atrophy, low inferior lens subluxation and mild cataract.

Preoperative uncorrected visual acuity OD 10/10 OS 2/10.

Best-corrected visual acuity improving to 8/10 in OS. (sph. +1.25 cil. −2.00 at 70°).

Normal IOP and fundus examination in both eyes.

The patient's main source of discomfort was a glare in his left eye.

Our first approach was the placement of a corrected iris print contact lens (CL) for refractive, cosmetic, and functional purposes, reaching a 8/10 VA with reduced glare. Unfortunately, the patient reported intolerance to the CL, choosing not to use it.

Six months later, the patient's cataract worsened into a mature white cataract, reducing the glare but increasing the esthetic concerns. With a normal ultrasound evaluation, the surgical option was presented to remove the cataract and place a lens with aniridia implant 67F from Morcher GMBH (Stuttgart, Germany) (Fig. 3.4).

A superior scleral approach was used to perform the phacoemulsification of the lens with the posterior widening of the incision for placement within the lens bag. The implant's wide nature allowed for proper tension without the need for fixation despite some superior zonular rupture.

Final visual acuity in the left eye was 10/10 with sph +1.00 cyl. −2.50 at 10° ADD +3.00.

Best tolerance and glare reduction were obtained with multifocal progressive aerial spectacles with photochromatic transition treatment.

3.4.2 Case 2 (Courtesy of Dr. Ariel Blanco)

A 22-year-old male suffering a penetrating corneal wound to his right eye with iris tissue loss. First surgical intervention achieved the correct closure of the eye. A secondary procedure was indicated when a white cataract developed.

Phacoemulsification of the lens was performed with the assistance of ocular tension rings placement. Two 50C aniridia implants from Morcher GMBH (Stuttgart, Germany) were placed in the bag to achieve occlusion alongside an aspheric foldable IOL (Fig. 3.5).

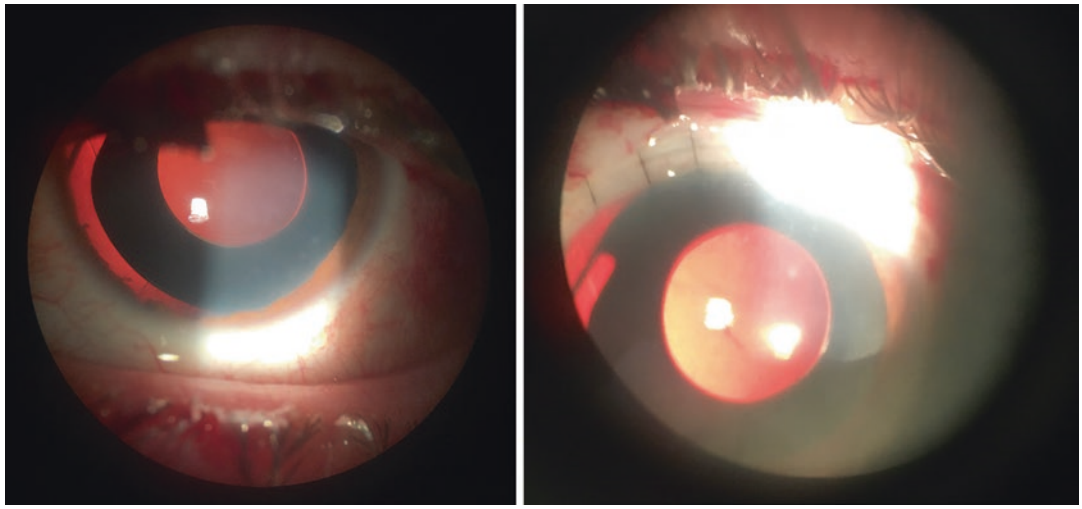

Fig. 3.4 Postop 67F lens from Morcher GMBH (Stuttgart, Germany)

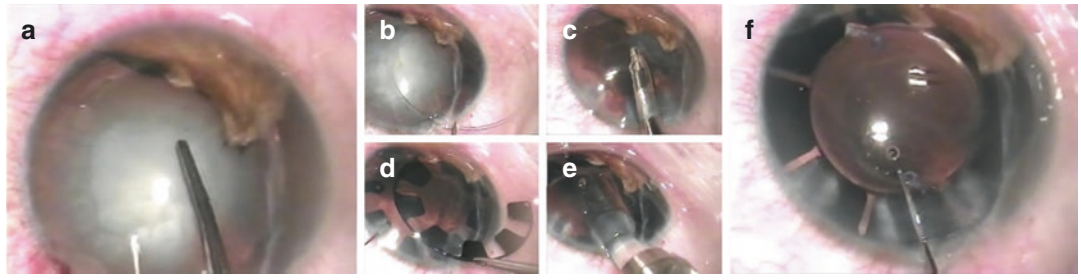

Fig. 3.5 (**a**) Initial conditions, (**b**) Capsular tension ring placement, (**c**) phacoemulsification, (**d**) Morcher implants, (**e**) IOL placement, (**f**) final results. Images Courtesy of Dr. Ariel Blanco

3.4.3 Case 3 (Courtesy of Dr. Ariel Blanco)

A 50-year-old male suffering post-traumatic aniridia with iris atrophy in his left eye.

After lens removal and IOL placement in the bag the Artificial Iris is cropped to size, and a peripheral "artificial iridectomy" is performed to guarantee aqueous flow (see Fig. 3.6a–d). Two points of fixation from prostheses to sclera are taken with polypropylene suturue before the implantation through the main corneal incision (see Fig. 3.6e, f). After implantation, the sutures are tightened and fixed, correcting for centration of the prostheses (see Fig. 3.6g, h).

3.5 Important Signs, Examinations, Diagnosis, Surgical Procedures and Skills, or Postoperative Treatment for Complications

Appearances might be deceiving, and a trauma case severe enough to cause some degree of aniridia is not to be underestimated. Whether the iris is lost due to extraction in prolapsed tissue, atrophied, desinserted, or ruptured, the energy involved in any of these traumatic mechanisms rarely limits its damage solely to the sphincter.

A thorough fundus examination is mandatory in any, and all trauma cases, second only to ultra-

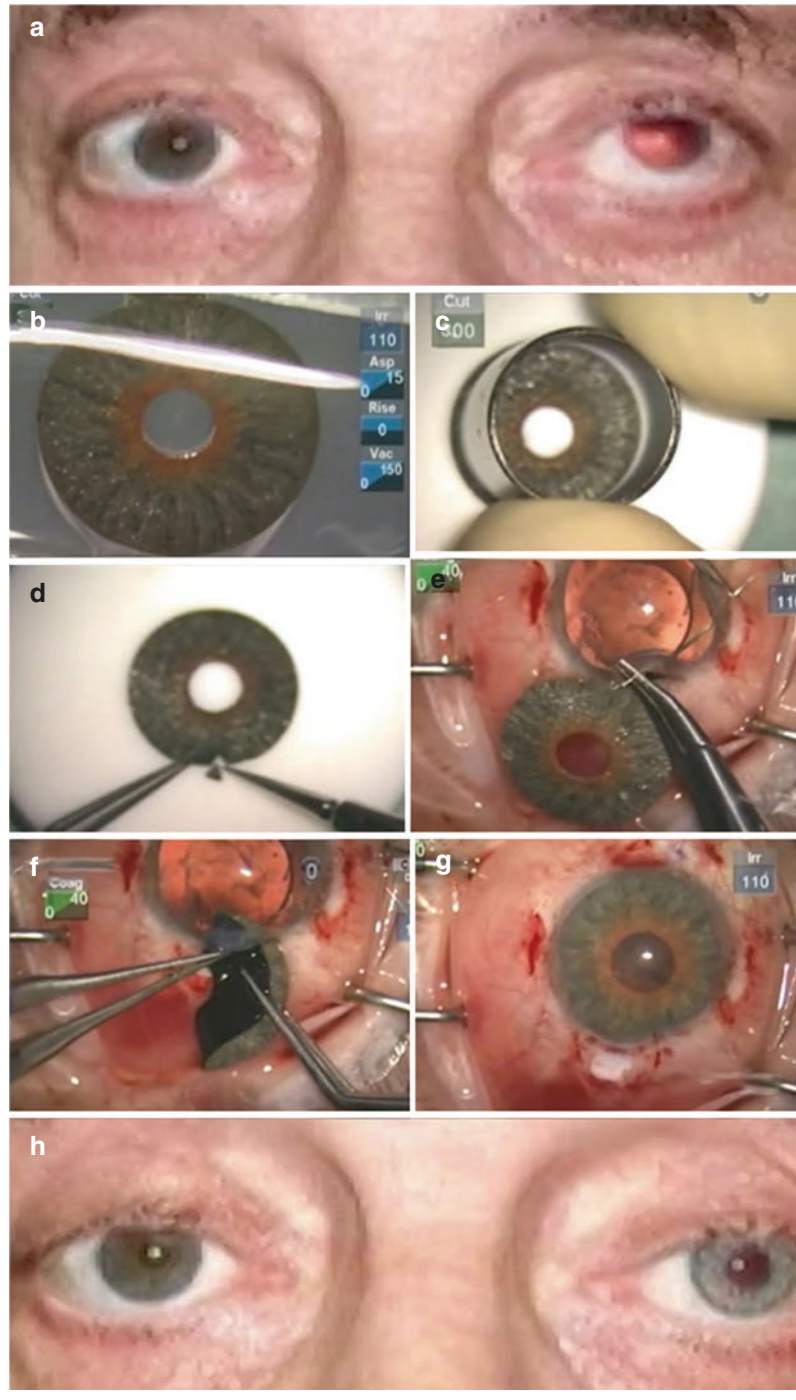

Fig. 3.6 Implantation procedure for CUSTOMFLEX® ARTIFICIALIRIS by HumanOptics. (**a**) Pre-operatory, (**b**) iris implant, (**c**) implant size adjustment, (**d**) suture fixation, (**e**) first intraocular suture placement, (**f**) folded implant insertion, (**g**) fully placed implant, (**h**) post-operatory. Images Courtesy of Dr. Ariel Blanco

sound evaluation (in closed globe scenarios). If no retinal detachment (RD) is present special attention must be taken to examine both the posterior pole for macular holes and the periphery for any retinal tears. Retinal damage takes priority after eye closure is obtained when needed, and all anterior segment interventions are secondary to the retina expert. Mixed procedures such as phacovitrectomy with anterior segment repair can be attempted under the proper conditions of visual-

ization, resources, and personnel available. Posterior segment evaluation does not stop if the initial evaluation is normal since some patients might experience RD in mid to late terms after ocular trauma. Patients should be observed in all follow-ups as well as educated in alarm signs such as novel floaters, metamorphopsia, and photopsia.

Intraocular pressure (IOP) increases, and glaucoma secondary to anterior segment trauma is frequent. Pigment dispersion, synechiae, angular scarring, and hyphema are possible causal mechanisms and need to be addressed since the initial evaluation all the way to postop follow-up. Same as with retinal damage, not all ocular trauma patients who develop glaucoma experience it since their initial exam and most are only detected if examined.

When examining the anterior segment, phacodonesis is one of the most informative signs to be looked for. Even if no evident subluxation is observed, knowing the zonular structure might be damaged can make the difference in proper planning for intervention with alternative IOLs and capsular support such as tension rings. A simple way to examine the inevident cases is, under slit lamp evaluation with the patient's head weight fully rested on the supports, to knock on top of the examination table upon which the slit lamp is placed. The vibrations will travel through the lamp's structure into the eye and cause a subtle but observable movement of the lens. Whenever in doubt, repeat the process observing the healthy eye and confirm no such changes occur.

As in all intraocular procedures, follow-up consultations need to evaluate for endophthalmitis and RD with prompt referral to retina experts if any of these are detected. A common complication, especially with in bag or sulcus placement of prostheses, is the luxation to the posterior segment. In such cases, a full vitrectomy is indicated with removal and/or scleral fixations of the devices.

3.6 Personal Experiences

As suggested at the beginning of this chapter, there are multiple sources of discomfort for the patient suffering aniridia. Especially in post-trauma patients who experienced a totally normal life until the event and now have to cope with the consequences. A sincere and thorough interrogation will go a long way in identifying the main concern with each patient for whom functionality might not be the top priority.

Some patients pose glare as their main concern without much care for esthetics and even stating that the functionality of their fellow healthy eye is compromised since they can no longer work comfortably outdoors. It is often useful to consider the need for fundus examination while tackling glare with occlusive means since the "new pupil" will be static; therefore some compromise with minor glare reduced by opaque or changing spectacles might be advisable.

Other patients want to regain visual acuity at all costs and as soon as possible, especially if the fellow eye is not at 20/20 for some other reason. Patients with a relatively healthy posterior segment and corneal transparency might achieve good visual outcomes provided the correct IOL measurements are obtained, and the prosthesis is correctly placed. But, for most, there needs to be serious talk before surgery to manage expectations. A post-traumatic cataract with aniridia is not a run of the mill procedure, lens measurements can be troublesome and sometimes can only be estimated via ultrasonography, the anatomy of the anterior segment is drastically changed, leading to corrective mistakes, and the zonular diaphragm might not resist the in bag option leading to sulcus placement or scleral fixation (all less precise). An honest talk with the patient and their family needs to address these issues and explain the case at hand with the myriad of alternatives both in treatment and possible complications, always keeping the door open for ulterior interventions or the assistance of contact lenses and/or glasses (this is particularly true for younger patients who are not used to reading glasses).

Finally but not least important is the esthetic aspect concerning the patient. This often disregarded issue by the physician might be the center of the patient's psychological discomfort and is a subject that needs to be handled with every bit of honesty as the ones mentioned above with an

extra layer of tact not only with young and impressionable patients but also with the occasional heavy-hearted parent. When available, offer the cosmetic contact lens option as soon as possible, even when surgical treatment is under consideration or scheduled for the intermediate future. This will provide not only the alleviation of the patient's concern; but also a valid alternative in case surgical outcomes do not meet the subject's expectations.

As far as surgical options, the CUSTOMFLEX® ARTIFICIALIRIS by HumanOptics provides one of the best alternatives in handling these two aspects (no commercial interest from any of the authors). We do find, however, two limitations to be considered with this alternative.

1. *Pupil Size*: ArtificialIris is 360°, 12.8-mm diameter disks with fixed pupils of 3.35 mm, which present a limitation in the fundus examination. However, modern Non-Mydriatic Fundus Cameras can help overcome this issue, and an ultrasound evaluation can provide a comprehensive assessment of the posterior segment.
2. *Price and Availability:* the implant was granted approval by the FDA on May 30, 2018, extending its legality to other, but not all, regulatory agencies around the world that follow the American standards. Yet, price can be a significant limiting factor.

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