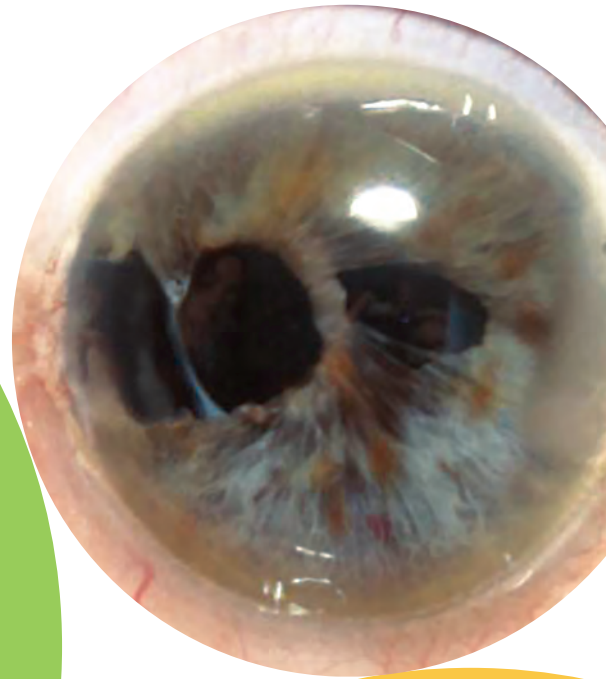


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# Focal Points®

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## Surgical Management of Congenital and Acquired Iris Abnormalities

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## Learning Objectives

Upon completion of this module, the reader should be able to

- 1 Summarize the common symptoms of patients with iris defects.
- 2 Discuss goals for treatment of an iris defect.
- 3 Gain proficiency in assessing iris defects and their impact on the patient.
- 4 Describe various techniques and devices to repair iris defects.
- 5 Describe complications that can arise from use of an iris prosthesis.

## Introduction

Iris abnormalities may either be pre-existing or occur iatrogenically during surgery. When iris damage occurs, careful assessment and treatment will directly impact the success of the procedure. Patients with iris defects present surgical challenges because the implantation of a traditional intraocular lens (IOL) may cause additional significant glare, photophobia, and polyopsia. Furthermore, in some iris defects, such as those associated with iridocorneal endothelial (ICE) syndrome, or in eyes with other comorbid conditions, such as retinal abnormalities or glaucoma, the overall success of the procedure may ultimately be compromised by disease progression.

Various techniques and devices can be used for iris repair. In general, repair is performed by cerclage, direct or indirect iris suture, or by implantation of a prosthetic iris. In addition, contact lenses and corneal tattooing, although infrequently performed, can be used to block unwanted excess light.

## Patient Assessment

In the patient with pre-existing iris defects, taking a careful history is important. One must understand how the defect occurred, what previous treatments have been tried, and what options exist for repairing the condition. It is most important to carefully ask patients about the extent of their visual disability. Consideration should be given to not repairing or replacing the iris tissue in patients who do not have significant symptoms. In general, larger iris defects cause more symptoms than smaller ones.

## Symptoms

Patients with iris defects can have symptoms that range from little or no visual compromise to severe

and debilitating. The most common symptoms are the following:

- *Photophobia.* Aversion to light can cause even normal indoor lighting to be troublesome.
- *Glare.* Starbursts, halos, or hazy vision occur particularly in bright light, but are possible even in poor lighting.
- *Decreased visual quality.* This is due to the transmission of higher order aberrations (particularly spherical aberration) from light travelling through the peripheral cornea, which is unguarded by the peripheral iris. This is particularly important in assessing patients with congenital aniridia. These patients also frequently have foveolar hypoplasia and have limited visual potential to less than 20/100. However, despite the fact that their Snellen acuity may not significantly improve after iris prosthetic implant surgery, these patients universally note a dramatic improvement in the quality of their vision.
- *Cosmetic dissatisfaction.* Patients with iris defects may experience poor self-esteem, self-consciousness, and depression due to their iris abnormalities. In addition, the presence of obvious iris abnormalities may affect how patients are perceived by others. Cosmetic improvement, particularly at the time of cataract surgery, can improve the emotional disposition of such patients.

## Iris Functions and Treatment Goals

In order to appropriately repair iris defects, a clear understanding of the iris/pupil function is crucial. Besides modulating light entrance through the pupil, the iris has several equally important functions, particularly in the context of cataract surgery. The goals of treatment of an iris defect are to

- Obliterate the pseudophakic-aphakic junction in patients with IOL implants.
- Reduce confusional focused/unfocused images.
- Reduce peripheral asphericity and optical aberrations.
- Decrease pupil size, thereby reducing photophobia.
- Improve cosmesis.

The repair should be designed to obliterate the pseudophakic-aphakic junction in patients with IOL implants;

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the absent or fully dilated pupil may be of a diameter in excess of 10 mm, while the largest IOL available in the United States is 6.5 mm in diameter (and most IOLs are 6 mm or less). Under these circumstances, light entering the eye will present both a focused (through the IOL) image and a superimposed defocused (around the edge of the IOL–aphakic) image. Similarly, patients with multiple iris defects may suffer from polyopsia, ghosting, poor contrast sensitivity, or reduction in visual quality. The repair of significant cosmetic defects as the sole complaint should be given due consideration, even in the absence of significant visual compromise.

## Causes of Iris Defects

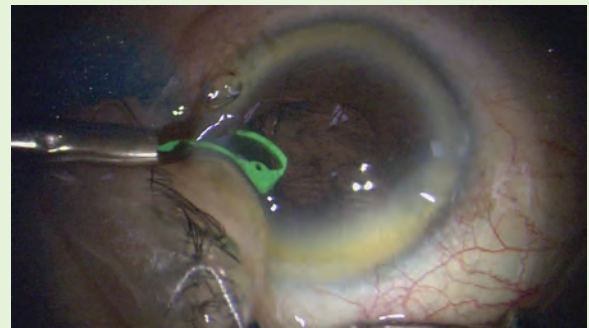
Iris defects may have traumatic or developmental etiologies (Table 1). In blunt and penetrating trauma, the surgeon should be careful of the likely concomitance of zonular dehiscence (particularly in blunt trauma), and of lens perforation (in penetrating trauma), as well as of corneal injury.

Iatrogenic conditions include surgical trauma, idiopathic postoperative mydriasis, or mydriasis associated with intraocular pressure (IOP)–related iris sphincter damage due to prolonged increased IOP, both in the postoperative period and as a sequela to untreated angle-closure glaucoma. Intraoperative snagging of the iris with instruments and devices may cause iridodialysis (Figures 1 and 2).

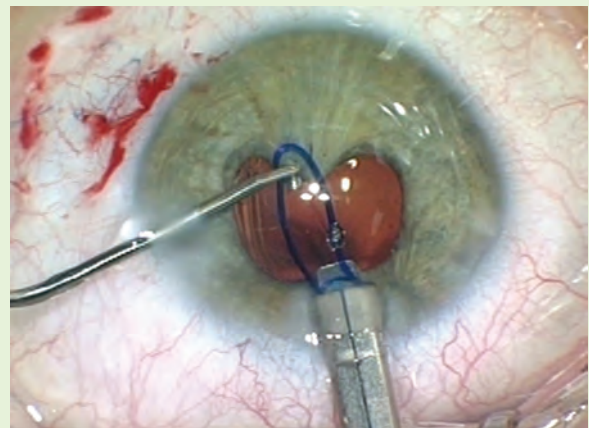
The preoperative use of  $\alpha_1$  adrenergic inhibitors such as tamsulosin (Flomax), doxazosin (Cardura), silodosin (Rapaflo), prazosin (Minipress), alfuzosin (Uroxatral), and terazosin (Hytrin)—drugs used in treatment of hypertension and urinary retention—may lead to an intraoperative floppy iris syndrome (IFIS).

Traumatic surgery can result in transillumination defects, iris prolapse, and chronic mydriasis (Figure 3). Iatrogenic traumatic aniridia has been reported in conjunction with endoscopic cyclophotocoagulation.

Congenital and developmental iris defects occur in a variety of conditions.



**Figure 1** The withdrawal of the BVI I-Ring pupil expander resulted in snagging of the temporal iris by the insertion and removal instrument, causing a subtotal iridodialysis. (Courtesy of Kenneth J. Rosenthal, MD, FACS.)

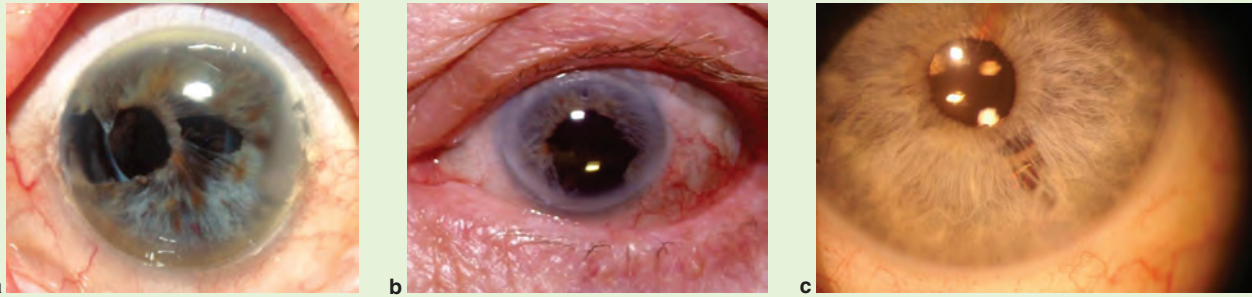


**Figure 2** Removal of the Malyugin pupil expansion ring caused a snagging of the distal iris by the loop. Slow and careful removal will avoid disinsertion or stretching of the iris. (Courtesy of Kenneth J. Rosenthal, MD, FACS.)

**Table 1. Causes of Iris Defects**

CATEGORY	TYPE OF DEFECT
Traumatic	Surgical: intraoperative floppy iris syndrome, iridodialysis, chronic elevation of intraocular pressure Nonsurgical: Blunt trauma Penetrating trauma
Developmental	Coloboma Iridocorneal endothelial syndrome Axenfeld-Rieger syndrome Congenital aniridia Ectropion uveae Albinism





**Figure 3** Examples of iris defects resulting from effects of intraoperative floppy iris syndrome (IFIS). **a.** Polycoria as a result of iris prolapse. **b.** Large defect involving iris sphincter and stroma. **c.** Transillumination defect. (Courtesy of Kenneth J. Rosenthal, MD, FACS.)

## Physical Presentation of Iris Defects

### Total or Subtotal Aniridia/Functional Aniridia

Complete functional absence of the iris occurs in patients with congenital aniridia and trauma, both surgical and penetrating. Complete iridodialysis rarely occurs as a surgical complication. Congenital and acquired ectropion uveae may also present as functional partial or full aniridia. Percussive injury (blunt trauma) may also cause partial or complete iridodialysis. Congenital aniridia should more accurately be named *hypoplastic iris syndrome* because almost all of these patients have an iris root present, although it serves no function. Most of these patients lack adequate iris tissue to permit optically useful surgical repair. Therefore, most patients with these abnormalities are likely to benefit from an iris prosthetic implant.

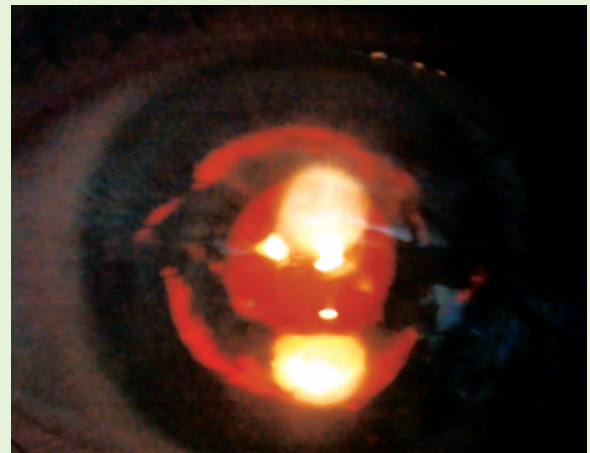
### Transillumination Defects

The examiner should carefully look for transillumination defects (TIDs). Patients with ocular albinism almost always have diffuse transillumination. Many other conditions may have focal or diffuse defects. Examples include Axenfeld-Rieger syndrome, iridocorneal endothelial syndrome, herpetic uveitis, Chandler syndrome, and Cogan-Reece syndrome. A unilateral presentation suggests surgical, blunt, or penetrating trauma, or a viral uveitic etiology. One must vigilantly check for ocular comorbidities such as glaucoma, corneal decompensation, cataract (particularly capsular and zonular abnormalities), and retinal issues. The placement of an iris implant to mask TIDs in these patients will markedly decrease photophobia and increase patients' visual potential.

Transillumination defects may also occur in patients who suffer from IFIS or intraoperative iris prolapse, due to stretching of the iris tissue. These defects have also been reported after blunt trauma, vitrectomy, and implantation

of IOLs (usually a single-piece IOL implanted in the sulcus or sutured to the posterior iris). We reported a case of TIDs in a patient with in-the-bag implantation of a single-piece acrylate IOL in conjunction with trabeculectomy using an EX-PRESS Glaucoma Filtration Device (Alcon Laboratories); this was due to iris trauma secondary to a shallow anterior chamber (Figure 4).

Many TIDs are asymptomatic, particularly those covered by the upper eyelid, and do not require treatment. Smaller, focal TIDs can be treated with direct surgical closure, whereas larger ones may require an iris prosthetic implant.



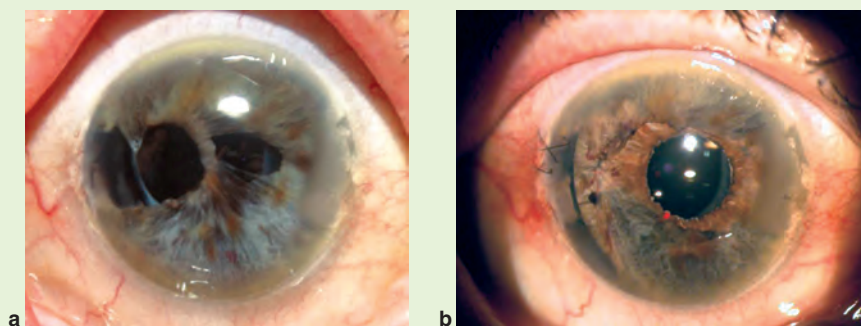
**Figure 4** A 1-piece intraocular lens (IOL) caused extensive transillumination defects that shadow the shape of the IOL, even though the lens was entirely within the capsular bag. Shallowing of the anterior chamber following this combined phaco-trabeculectomy may be implicated. The patient was completely asymptomatic and required no further intervention. (Courtesy of Kenneth J. Rosenthal, MD, FACS.)

## Polycoria

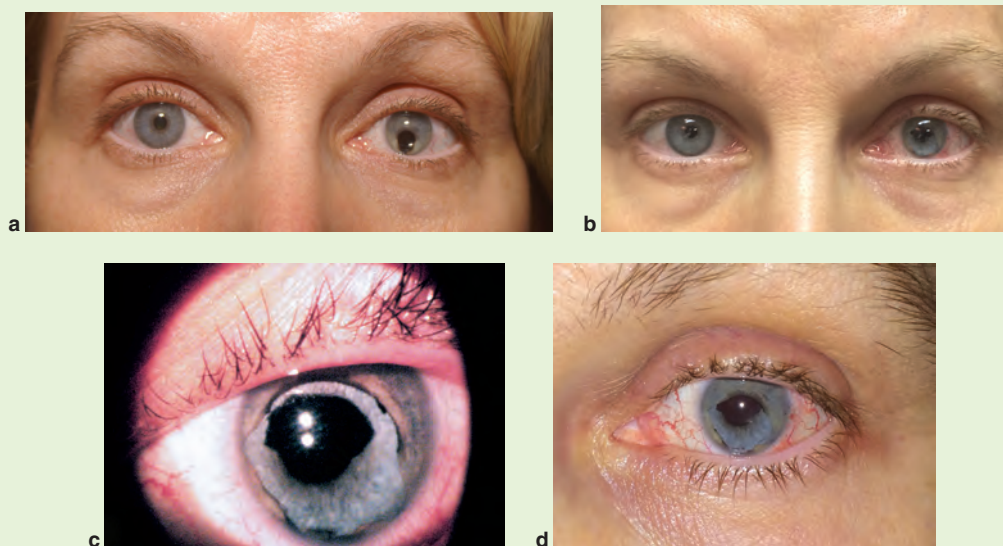
The presence of multiple pseudopupillary openings in the iris creates a “moth-eaten” appearance (polycoria), resulting in polyopia. This may occur in hereditary disorders such as Axenfeld-Rieger syndrome or posterior embryotoxon. The first case of modular/small-incision iris implantation, which I reported in 1996, was in a patient with Axenfeld-Rieger syndrome (Figure 5). The extent and location of the localized iris defects will determine their treatment. As with TIDs, smaller areas can be treated effectively with an imbrication suture, whereas the larger ones require prosthetic iris implantation.

## Sector Defects

**COLOBOMA.** Iris coloboma presents as a “keyhole-shaped” pupil and is a congenital disorder of the iris arising from failure of the embryonic optic plate to fully fuse in the fifth week of gestation. It may be associated with colobomas of the ciliary body, choroid, retina, or optic nerve. Iris colobomas are typically located in the inferonasal quadrant. Patients with this deformity are frequently very self-conscious about their appearance, and they often present for cosmetic improvement even in the absence of visual symptoms.



**Figure 5** Polycoria secondary to presumed intraoperative floppy iris syndrome (IFIS), with implantation of the HumanOptics CustomFlex iris prosthesis. **a.** Preoperative appearance. **b.** Postoperative appearance. The patient’s symptoms of polyopia and photophobia diminished markedly following this procedure. (Courtesy of Kenneth J. Rosenthal, MD, FACS.)



**Figure 6** Preoperative **(a)** and postoperative **(b)** photographs of a patient with coloboma of the iris and ciliary body. There was no posterior choroidal involvement. The patient had severe photophobia following cataract surgery, despite sequential opacification of the anterior capsule. Implantation of the CustomFlex artificial iris was performed, removing a portion of the superior iris prosthesis, to allow full pupillary aperture on dilation. **c, d.** Postoperative view of the coloboma: **(c)** close-up and **(d)** dilated. (Courtesy of Kenneth J. Rosenthal, MD, FACS.)

It should be kept in mind that patients undergoing simultaneous cataract surgery almost always have absent zonules in the area peripheral to the iris defect.

Closure of smaller colobomatous defects can be accomplished by the use of a Siepser knot or its variants. In iris colobomata that extend 2 or more clock hours, however, one should consider placement of an iris prosthetic implant—either a rigid “sector” iris implant or a flexible implant in which the superior, noncolobomatous area is cut out to allow access through the superior pupil when dilated (for dilated examination and low lighting conditions; Figure 6).

**OCULAR TUMORS.** Resections of iris tumors may result in variable degrees of residual iris defects. Primary repairs should be performed only if there are significant concerns about visual function or cosmesis. When necessary, repair should generally be done sequential to the tumor resection, in order to allow clinical healing as well as to ensure that the margins are tumor free, to assess the extent of the iris defect, and to prepare a surgical strategy.

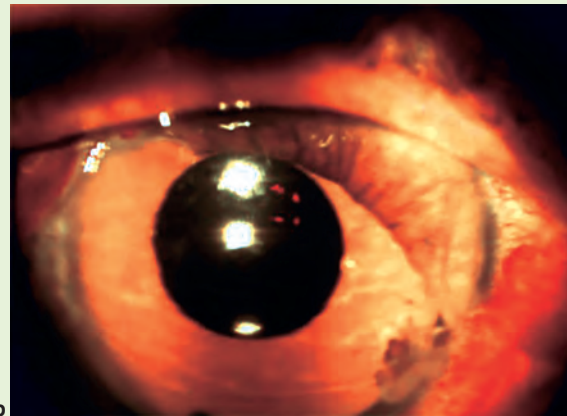
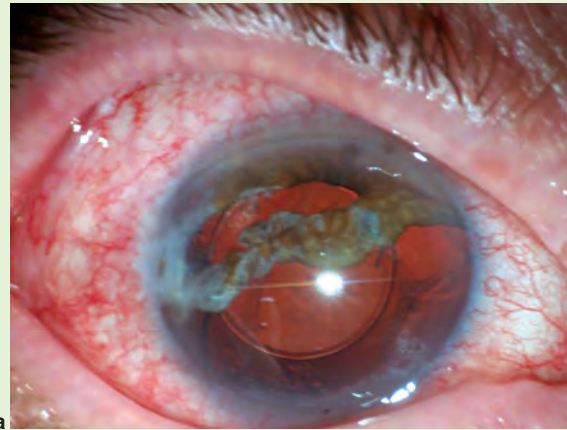
### Atonic Pupil

Chronic mydriasis is generally caused by failure of the iris sphincter due to mechanical damage to the myofibrils or by stretching of the iris stroma, or both. Common etiologies are blunt trauma (traumatic mydriasis), prolonged elevation of IOP (ie, acute angle-closure glaucoma or prolonged postoperative pressure spike), or inflammation.

Treatment of an atonic pupil overlaps with the strategy for congenital aniridia. However, it is much more likely that some degree of pupillary closure can be accomplished by cerclage or by quadrantic iris suture. For large or chronic mydriasis, particularly in patients with light-colored irises where the iris stromal pigment is scant to begin with, closure may result in inadequate pupillary closure and/or diffuse and visually significant TIDs. In these patients, as in all patients with inadequate iris stroma in general, an iris prosthetic device is the preferred strategy. Alternatively, in borderline cases, due consideration should be given to a stepwise approach: repair and then, if that fails, implantation of an iris prosthesis. In patients who may be at risk for poor healing after multiple surgeries, notably patients with poor corneal endothelial function, immune compromise, or diabetes, an iris implant is often the best choice.

### Iridodialysis

Iridodialysis (Figure 7) occurs most commonly due to trauma, although it may rarely occur secondary to degenerative disease. Surgically induced iridodialysis is commonly seen in conjunction with iris prolapse or with the inadvertent tethering of the iris with an instrument (see Figures 1 and 2). In such cases, prompt repair of these conditions may result in a good restoration of function. Delay may result in an ischemic and atrophic iris.



**Figure 7** Iridodialysis. **a.** This patient suffered a large iridodialysis with fixed, dilated, curled iris tissue throughout. **b.** After excision of the tissue, a partial iridodialysis repair was performed, and an Optec Model 311 inserted to reform the pupil. (Courtesy of Kenneth J. Rosenthal, MD, FACS.)

### Peripheral Iris Trauma

In cases of iris prolapse due to trauma or an iatrogenic cause (eg, during cataract surgery with shallow chambers, basal incisions, floppy iris, or increased IOP from choroidal hemorrhage or reverse pupillary block), repositioning the iris should generally be performed in preference to excision. However, in cases where there is prolonged external exposure, or strangulation or necrosis of the prolapsed iris, consideration should be given to excising the offending tissue prior to reconstructing the remaining iris using techniques described elsewhere in this paper.

Surgical iridectomy or iridotomy may cause visual distraction due to stray light entrance and visual symptoms. These problems may be alleviated by partial closure of the iridotomy or by corneal tattooing overlying the iris defect.



## Corectopia

Corectopia can be the source of visual challenges as well. The concentricity of the pupil with the central cornea and the macula (the “visual axis”) is important for optimal acuity. The ectopic pupil reduces vision because of spherical and other higher order aberrations as well as chromatic aberration, all of which increase toward the peripheral cornea, and poorer sensory resolution due to eccentric macular fixation. *Ectopia pupillae* is associated with myriad clinical syndromes, including penetrating trauma, iris coloboma, Axenfeld-Rieger syndrome, microphthalmia, irido-corneal-endothelial syndrome, and idiopathic congenital ectopia pupillae.

Medical treatment may be undertaken using mydriatics. However, these are often only temporary solutions due to suboptimal efficacy, ocular surface toxicity of the compounds, patient compliance, and tachyphylaxis.

Surgical intervention can be considered either in isolation or in conjunction with lens/cataract surgery. After synechiolysis, the pupillary ruff is incised up to a point slightly beyond the area of the desired reconstructed pupillary center. A vitrector or intraocular scissors can be used to shape a new “pseudopupil.” The pupillary sphincter should be preserved as much as possible to allow some pupillary movement postoperatively. The cut iris that lies between the original pupil and the new pseudopupil is then closed using 9-0 polypropylene sutures. In larger defects, it may be desirable to use an artificial iris implant behind the iatrogenic coloboma rather than suture it. This will largely depend on the width of the residual defect, the strength of the surrounding iris tissue, and the presence of concomitant transillumination defects.

In severe cases of ectopia pupillae, or in conditions such as progressive Adie pupil, or with total pupillary absence, one can consider making a de novo incision in the central iris and using a vitrector or curved vitreoretinal scissors to create a new pupil of about 4–5 mm in diameter. This allows for adequate light entrance in a variety of luminances, as well as adequate examination of the posterior segment.

## Treatment of Iris Defects

### Ocular Surface Strategies

Before considering surgical intervention, due consideration may be given to prescribing masking contact lenses. My experience with these contact lenses has been that they are generally uncomfortable and do not provide good quality optical correction. Nonetheless, in patients who wish to avoid surgery, or in whom surgery is otherwise not possible, this may represent an acceptable, if temporary, option.

Another method, though rarely employed, is to place permanent pigmentation in the corneal stroma, known as *corneal tattooing*. Some improvement in symptomatic glare and photophobia following peripheral iridectomy has been noted. Corneal tattooing has been practiced for almost 2000 years. With the increasing availability of implantable

iris devices, indications for the use of this procedure are narrow, except in instances of purely cosmetic rehabilitation in blind eyes. Some successes, particularly in improving cosmetic appearance, can be obtained. Alio and colleagues have had success using the femtosecond laser to place the tattoo. The procedure is not reversible and may cause difficulty in performing other ophthalmic examinations, particularly a fundus examination.

### Surgical Approaches

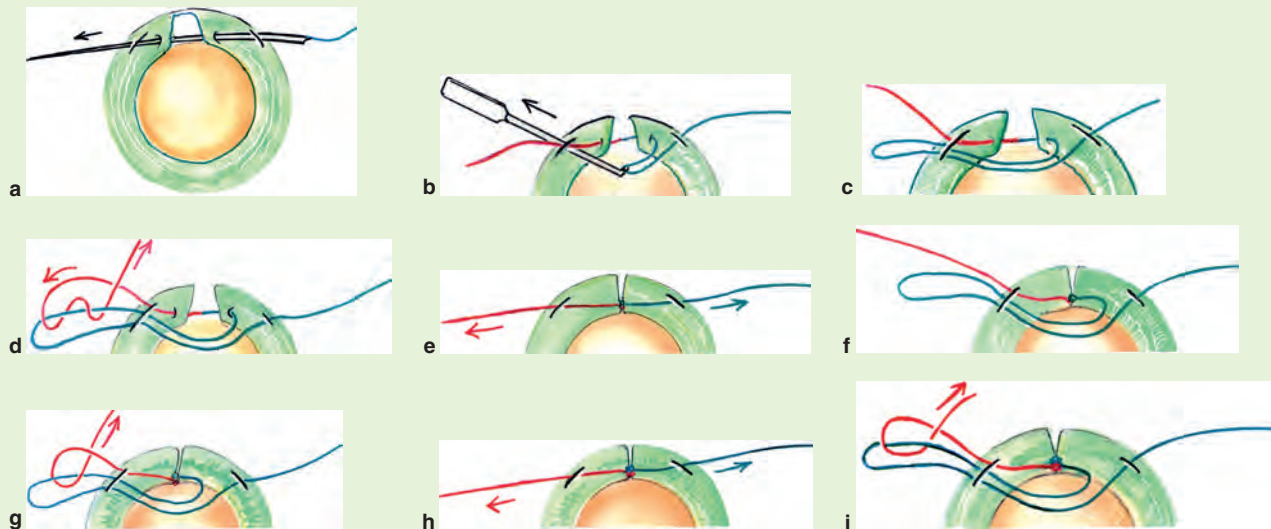
Because every iris defect is unique, the surgical approach must be tailored to the specific situation. Primary repair of an iris defect by means of techniques such as iris sphincterotomy, synechiolysis, iris suturing, and cerclage may be adequate in some patients. However, these techniques may be neither possible nor sufficient in eyes with more extensive iris defects. In these cases, overly aggressive iris suturing techniques should be avoided, because the iris tissue is almost universally abnormal, and there is a risk of iatrogenic iridodialysis and hemorrhage, as well as of sequential iris atrophy in areas under tension from suturing. Iris prosthetic implantation is preferable in the latter cases.

In evaluating the extent of the iris defect, one must consider not only the surface area of the iris that is deficient but also the elasticity of the remaining iris tissue, the eye color, and the presence of transillumination defects. Light-colored irises, especially light blue ones, are particularly susceptible to transillumination defects, either preoperatively or as a result of stretching of the iris tissue in the course of attempted repair. Similarly, albinotic irises almost always have total transillumination. Even when the iris tissue is intact, implantation of an iris prosthesis is likely to improve function by blocking light behind the pigment-free iris.

**IRIS DEFECT REPAIR OPTIONS.** Direct iris suture closure. The decision-making and application of iris repair are discussed in this module. Small areas of iris defects can be closed using either direct closure or a Siesper knot or its variants (Figure 8). My preference is to use direct closure for more peripheral defects, and the Siesper type of closure for more central closures. The latter will not necessitate undue pulling of the iris tissue to the periphery when tying the knot.

Direct closure employs the use of a McCannel-style suture, and of 9-0 or 10-0 polypropylene suture on a long curved or straight needle. From the area of iris defect, the suture is passed through the near clear cornea 2–4 mm adjacent to the defect, passed through the iris defect on each side, and then brought out through the near clear cornea on the opposite side. A straight through (ie, nonbeveled) paracentesis is placed overlying the defect. The suture ends are looped out through the paracentesis, using a Kuglen hook, and then tied, pulling the iris defect gently toward the cornea. The needle penetration can be accomplished with or without a paracentesis.





**Figure 8** The square-knot modification of the Siepser slipping suture knot technique with 3 throws (2-1-1) in a sectorial iris defect. **a.** The suture needle on 9-0 or 10-0 polypropylene is placed through the limbus. The needle is then placed through the adjacent areas of iris and out through a distal paracentesis. **b.** A Kuglen hook through the paracentesis retrieves a loop of suture from the side distal to the paracentesis. **c.** A loop of suture is created at the proximal paracentesis outside the eye. **d.** The iris side of the loop is then wrapped with a forceps (3 throws; forceps not shown) and the free end of the suture is grasped. **e.** The suture ends, outside of the eye, are each pulled such that the first throw of the knot slides inside the eye and is centered over the iris where the surgeon wants the knot to cinch. The iris will often twist temporarily to allow the throw to lie flat. **f.** The Kuglen hook (not shown) is used again to bring a loop of suture (blue) out in the same way as previously described, and the suture loop is oriented to commence the wrap of the second throw. **g, h, i.** Additional suture throws, creating a square knot, are performed. (Adapted from Osher RH, Snyder ME, Cionni RJ. Modification of the Siepser slip-knot technique. *J Cataract Refract Surg.* 2005;31:1099. Also Ogawa, GSH. Scleral and iris sutured posterior chamber intraocular lenses and intraocular knot-tying techniques. In: Fishkind WJ. *Complications in Phacoemulsification, Avoidance, Recognition, and Management.* New York: Thieme Publications; 2002.)

Ike Ahmed, MD, has developed a set of instruments that enable direct tying of the sutures within the eye without Siepser-type manipulation.

Direct, intraocular, or Siepser-style closure technique can also be employed in treatment of TIDs in which the defective iris is imbricated within adjacent areas of healthy iris tissue.

**Iris cerclage.** This technique is used when reduction in pupil size is desired, and when there is good iris stromal tissue with good pigmentation. The presence of TIDs or other iris defects, especially in lightly colored eyes, will result in a good anatomic closure and an unhappy patient, due to persistent translucence of the remaining repaired iris. It should therefore be avoided in favor of iris prosthetic implant surgery, when possible.

Cerclage is performed by passing sutures through multiple paracenteses, using a 9-0 polypropylene suture on a long, curved needle (or a comparable material), weaving the pupillary edge either through multiple individual bites (about 2 or 3 per quadrant), or with a “baseball stitch” technique, in which the suture weaves around the pupil

with each stitch. Once passed 360°, the suture ends are tied, either by direct closure through a straight through paracentesis (ie, not beveled as with traditional paracenteses) within true clear cornea, and as close to the proposed pupil size as possible, or with a Siepser or modified Siepser knot.

**Quadrantic imbrication.** Quadrantic imbrication can be accomplished by placing a McCannel/Siepser suture at each quadrant, thus producing a smaller pupil in a square configuration. Four paracenteses are performed. Through each, a 9-0 or 10-0 polypropylene suture on a long needle is passed, grasping the para-adjacent pupillary edge, about 1.5 mm peripheral to the pupil, then passed across several millimeters, and the iris is regrasped. The needle is passed through the next paracentesis, and tied using a Siepser, or Siepser-variant technique, thus imbricating the iris. This is repeated for each quadrant.

**IRIDODIALYSIS REPAIR.** Small iris root dehiscence, typically less than 2 clock hours, is often asymptomatic and can be left untreated, particularly when in areas normally covered by the upper eyelid. Repair can be

accomplished by a variety of means. My favored repair technique is as follows:

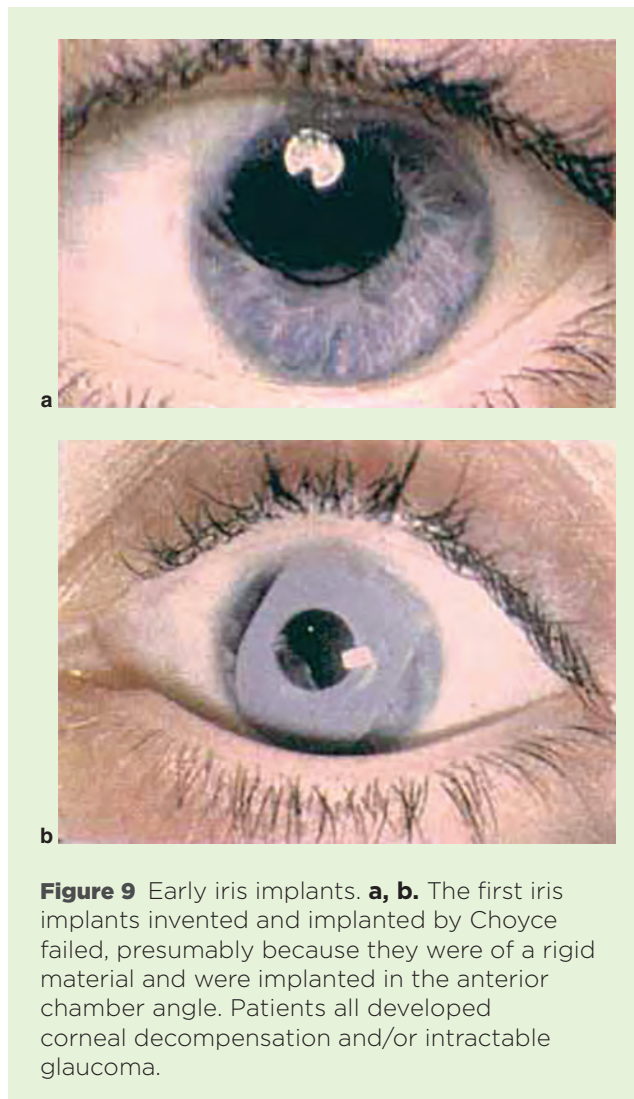
1. Make a 2–3 mm scleral groove beginning 2.5 mm posterior to the sclera overlying the area of dialysis.
2. Through a paracentesis, grasp the iris with a double armed 9-0 or 10-0 polypropylene suture near its peripheral, dehiscent root (some authors advocate using 8-0 polypropylene).
3. Pass a 25-gauge bent hypodermic needle on a 3 cc or smaller syringe through the scleral groove so that it pierces the sclera at 1.5–2 mm posterior to the limbus and passes parallel to the iris plane into the anterior chamber.
4. Engage the suture needle within the 25-gauge needle and pass it out through the sclera; pass the second arm of the suture 1–2 mm adjacent to this and tie the sutures loosely, leaving a small area of gap between the iris root and the internal scleral wall to ensure that the trabecular meshwork is not occluded.
5. When possible, rotate the suture knot into the eye, but if there is an adequate depth groove and the knot is covered, and when rotation is not mechanically possible, this may not be necessary.
6. If the suture is exposed, cover it with an adjacent scleral graft or with eye-banked sclera or cornea. Confirmation using a gonioprism or an intraocular endoscope is helpful in cases where the placement of the suture cannot be clearly determined. For larger areas of iridodialysis this can be repeated as many times as required, leaving some space in between the suture passes, to minimize the number of sutures required and to allow some space peripheral to the dialysis. When adequate Tenon and conjunctiva can be mobilized, the suture site should be covered and secured with either tissue glue or absorbable sutures such as 7-0 or 8-0 polyglactin.

In moderate-sized dialyses, imbrication of the adjacent iris tissue may create an adequate closure, although a peripheral—and generally optically innocuous—space may persist.

An alternative technique purposefully creates a visible space, a so-called “hang back” technique to minimize the chances of trabecular occlusion. A sutureless technique involves the entrapment of the iris within a sclerotomy.

### Iris Prosthetic Implants

The implantation of a masking artificial iris implant can potentially reduce glare and improve patients’ visual function. Such devices may also mask the optical aberrations prevalent in the peripheral cornea and eliminate visual competition between focused and unfocused images. Newer developments have allowed a near perfect cosmetic match between eyes as well.

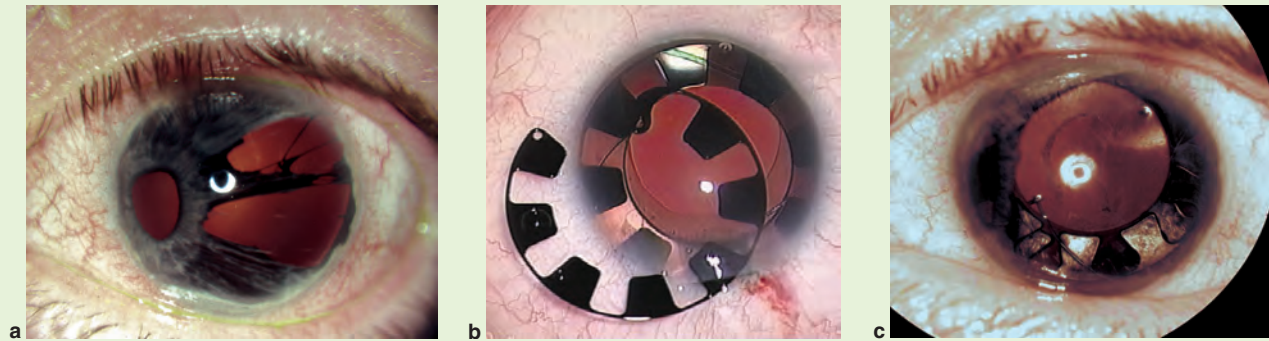


**Figure 9** Early iris implants. **a, b.** The first iris implants invented and implanted by Choyce failed, presumably because they were of a rigid material and were implanted in the anterior chamber angle. Patients all developed corneal decompensation and/or intractable glaucoma.

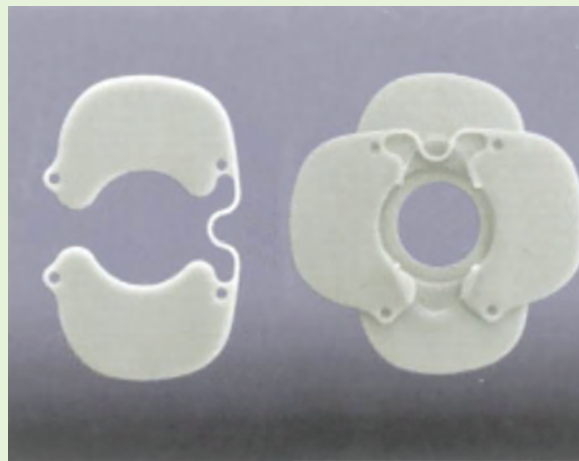
For eyes that have iris defects too extensive for primary surgical repair, implantable iris prosthetic devices have been developed that will help the surgeon compensate for the deformed or imperfectly functioning iris. Several companies produce these iris prosthetic devices. The HumanOptics CustomFlex artificial iris, discussed later in this module, was recently approved by the United States Food and Drug Administration (US FDA). Other devices are available outside the US.

**EARLY IMPLANTS.** The first iris implants were developed and implanted by Choyce as iris/lens combinations, fixated in the anterior chamber angle, as early as the 1960s (Figure 9). However, they universally failed due to corneal compensation and/or intractable glaucoma.

In 1991, Sundmacher developed a polymethyl methacrylate IOL surrounded by an opaque black segment that serves as an artificial iris diaphragm for implantation in patients with iris defects. In patients with absent or inadequate capsular support, this lens was effective, though the black optic carrier measures up to 10 mm in diameter



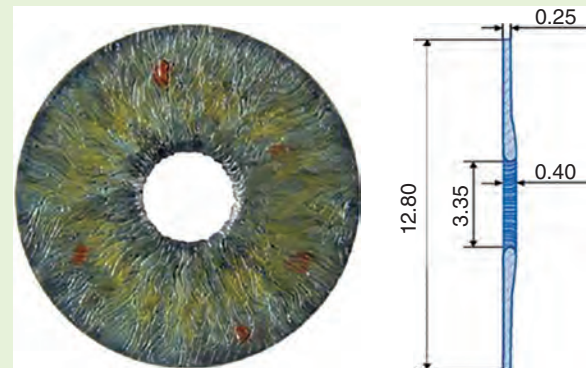
**Figure 10** The Rasch-Rosenthal iris diaphragm ring, the first small-incision iris device, is implanted within the capsular bag in pairs and dialed so that the fins slightly overlap. **a.** Preoperative photograph: Axenfeld-Rieger syndrome with polycoria. **b.** Second Rasch-Rosenthal implant being inserted. One modular implant is already in the capsular bag. **c.** Postoperative appearance showing continuous iris prosthesis, obliterating the pseudophakic-aphakic junction. (Courtesy of Kenneth J. Rosenthal, MD, FACS.)



**Figure 11** The Ophtec Iris Prosthetic System designed for in-the-bag implantation through a small incision. First, 2 elements are implanted 90° apart. A clip is then placed (shown behind the 2 primary elements) to create a round pupil and bind the elements in a stable fashion.

(and encompasses varying optic sizes), thus requiring a large incision. It is sutured to the scleral wall, through eyelets on the terminus of each haptic.

**OTHER SYSTEMS.** In response to the incision size limitation, Volker Rasch, MD, of Potsdam, Germany, and I invented a series of aniridia and coloboma aperture rings (Morcher GmbH). The Rasch-Rosenthal iris diaphragm rings are designed to be implanted in an intact capsular bag, with or without zonular instability and with a separately implanted IOL (Figure 10). These devices have a 6-mm pseudopupil and were designed to interface with the most popular IOL designs of 6 mm, thus obliterating



**Figure 12** The HumanOptics CustomFlex iris prosthesis is a handcrafted device on a silicone base. It uses silicone paint to copy a photograph matching the fellow eye, or eye subject of the patient's choice. The artisan takes into account the effect of the corneal magnification and replicates the unique coloration and contour of the photograph supplied.

the pseudophakic-aphakic junction. I implanted the first of these devices (the first iris implant of any kind in the US) in July 1996 in a patient with Axenfeld-Rieger syndrome. Masker has developed a similar modular iris prosthesis designed with a 4-mm pupil that is thought to be more physiological in average lighting conditions.

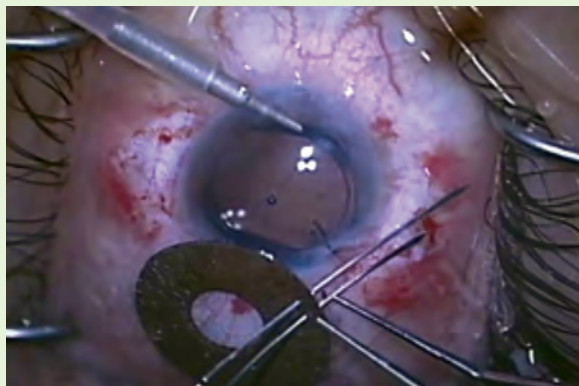
Ophtec manufactures a similar device, the IPS (Iris Prosthetic System), but it is not currently available in the US (Figure 11). Ophtec also offers aniridic IOLs that are available in 3 homogeneous standard colors.

The HumanOptics CustomFlex artificial iris was invented by Prof. Dr. med. Hans-Reinhard Koch. As noted earlier, the device was recently FDA approved. It is composed of a



flexible silicone disk with a 3.5-mm central “pseudopupil” aperture (Figure 12). The devices can be implanted within the capsular bag or placed in the sulcus, with or without suture fixation. An optional fiber mesh backing is available

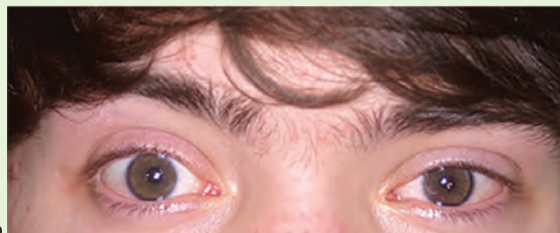
to provide further structure for suture fixation (Figure 13). The device can be trephined to the correct size or trimmed to be used as a partial device. It is flexible and can be inserted through an incision as small as 2.4 mm.



**Figure 13** This patient had sequelae of congenital rubella with microphthalmia, glaucoma, and hypoplastic iris. An intraocular lens and iris prosthesis were sutured in place. This photograph shows the needles of a double-armed polypropylene suture placed through the substance of the device, which will then be fixated to the sclera. This device was produced with a fiber backing, to add support to the suture fixation. (Courtesy of Kenneth J. Rosenthal, MD, FACS.)

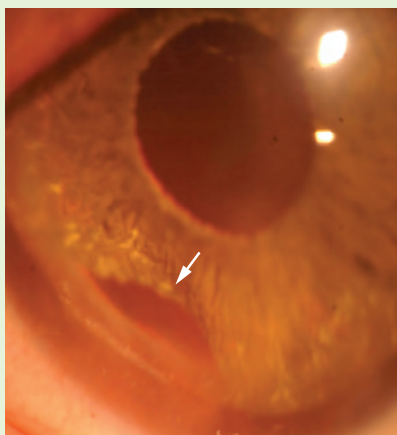


a

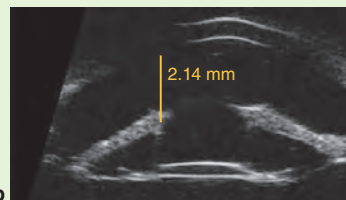


b

**Figure 14** A 17-year old patient with congenital aniridia—with foveolar hypoplasia—after implantation of the first (a) and then fellow eye (b) with the HumanOptics CustomFlex iris prostheses. Preoperative best-corrected visual acuity (BCVA) was 20/100 in both eyes, with nystagmus. Postoperatively nystagmus is virtually absent, and the visual acuity is BCVA 20/30. (Courtesy of Kenneth J. Rosenthal, MD, FACS.)



a



b



c

**Figure 15** Artificial iris implantation. **a.** Slit-lamp photograph taken 1 month postoperatively after implantation of the HumanOptics CustomFlex artificial iris, showing the inferonasal region of the prosthesis bowing forward (arrow). **b.** Ultrasound biomicroscopy (UBM) scan taken of the initial eye approximately 2 months postoperatively. Measurement of the anterior chamber depth from the periphery of the pupil at the apex of the vaulted prosthesis to the corneal endothelium was 2.14 mm. **c.** UBM scan taken of the patient's left eye 3 months postoperatively. Measurement of the anterior chamber depth remained at 2.14 mm and had not progressed by clinical evaluation. Some investigators think that implantation of a capsular tension ring at the time of the primary surgery may deter this problem. (Courtesy of Kenneth J. Rosenthal, MD, FACS. From Rosenthal KJ, Venkateswaran N. Clinical and ultrasound biomicroscopic findings in a patient with anterior vaulting of a customized, flexible artificial iris. *J Refract Surg.* 2013;29(1):663–664.)



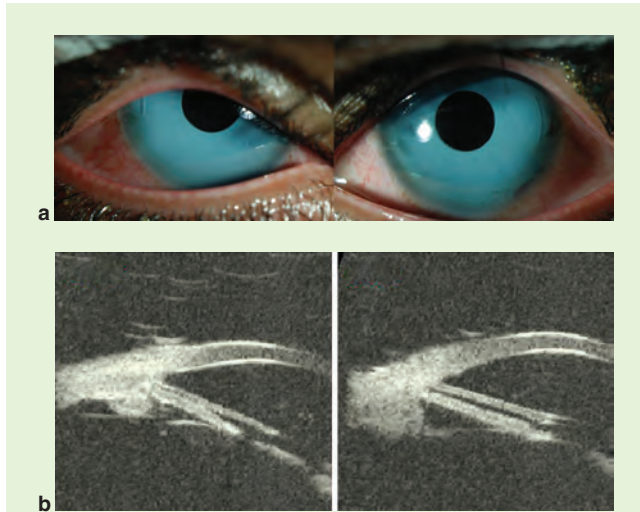
## Complications

Many patients, if not most, who suffer from iris abnormalities have significant comorbidities. The congenital aniridic patient frequently has concomitant limbal stem cell deficiency with its attendant corneal vascularization and pannus, as well as foveolar hypoplasia and nystagmus. Nonetheless, implantation of the iris prosthesis in such patients frequently will produce better-than-anticipated

results, presumably because presence of the new iris causes improvement in visual quality and allows the patient's neuroadaptive processes to progress. The device can be inserted through a sub 2.5-mm incision so there is minimal impact on the already deficient stem cell population in these patients (Figure 14).

When the HumanOptics device is placed in the capsular bag, it may eventually "pea-pod" out of the bag due to capsular phimosis. We reported such a case, which stabilized and did not affect the outcome (Figure 15). Shifting or dislocation of the device may occur but can almost always be repaired.

The BrightOcular Cosmetic Iris implant is a device made of silicone and fixated in the anterior chamber angle. It has been used primarily to change eye color, although it has been used for functional iris repair as well. Along with other surgeons, we have reported devastating ocular complications—in particular, iris depigmentation, pigmentary glaucoma, end-stage and advanced glaucomatous cupping, and corneal decompensation in individuals who received this implant. In some cases, explantation of the device was helpful in preventing further advancement of the disease (Figure 16). These devices, although not available in the US, have been implanted elsewhere, including a concentration of cases in the Middle East, India, and Mexico, and there are patients in the US who have travelled abroad for this surgery.



**Figure 16** Complication of a cosmetic iris implant. **a.** A patient developed corneal decompensation, pigmentary dispersion, and ocular pain after insertion of the Nulris/BrightOcular cosmetic iris. **b.** Ultrasound biomicroscopy demonstrates contact with the iris as well as the corneal endothelium. The devices were explanted. Despite this, corneal decompensation and glaucoma persisted. (Courtesy of Kenneth J. Rosenthal, MD, FACS.)

## Conclusion

Iris abnormalities, both congenital and acquired, may cause significant visual challenges. Knowledge and proficiency in assessment of iris defects, their impact on the patient, and various techniques and devices to repair these problems represent a significant step toward improving visual function and comfort.

# Clinicians' Corner

**Samuel Masket, MD**, and **Francis W. Price Jr, MD**, weigh in with their clinical perspectives on “Surgical Management of Congenital and Acquired Iris Abnormalities.” Clinicians' Corner consultants respond to questions without reading the module or each other's responses.

## 1. What surgical tips do you have for a surgeon learning to implant the CustomFlex artificial iris (Human Optics), approved by the United States Food and Drug Administration (US FDA) in 2018?

**Dr. Masket:** Surgeons wishing to add this device to their treatment for cases of partial or total aniridia must learn several steps of the procedure in order to achieve good results. Properly used, this prosthetic iris can be a life-changing experience for the patient and most rewarding for the physician. Those steps include taking appropriate photos of the fellow eye in order to obtain an aesthetically valid color match. Additionally, presurgical evaluation should determine the best means for “harnessing” the artificial iris to the eye, recognizing that there are potentially 5 specific methods:

- within the capsule bag in combination with cataract surgery or with an existing “in the bag” intraocular lens (IOL)
- unsutured in the ciliary sulcus
- sewn to the sclera
- fixated (sewn or otherwise) to an IOL and implanted as a unit with either the device or IOL sutured to sclera
- sewn to a remnant of the native iris

Understanding these options and when to best apply them is essential, given that eyes requiring an artificial iris tend to have multiple preexisting abnormalities. At present, a teaching curriculum is under development (VEO Ophthalmics) and will be comprehensive and vital for surgeon education. However, until that time, ophthalmologists are urged to take courses that are offered at the annual meetings of the American Academy of Ophthalmology, the American Society of Cataract and Refractive Surgery, and the European Society of Cataract & Refractive Surgeons. In addition, visiting a colleague who is familiar with the device will be of immense benefit.

**Dr. Price:** The most important tip is to take an introductory course to gain a basic knowledge about the handling and use of the implant. In-the-bag placement at the time of cataract surgery is by far the safest, least complicated way to use the implant. All other methods require some sort of secondary fixation because the implant has no haptics. It is important to prevent the edge of the implant from contacting any intraocular surface (other than the capsular bag) because that could cause inflammation and other problems.

We learned with an earlier implant, the Ophtec model 311, that a 9-mm diameter artificial iris provided substantial improvement in glare and light sensitivity for most patients. As a result, I seldom use an implant diameter larger than 9 mm unless the corneal diameter exceeds 12.5 mm. It is important to keep in mind that if an endothelial keratoplasty is required, these implants are not in contact with the wall of the eye, and the surgeon should not let go of the graft until it is either firmly anchored to the cornea or is firmly pressed against the cornea by an air bubble; otherwise, the graft will sink back to the retina.

## 2. For small peripheral iris defects, are there any additional tests (ie, colored contact lens trial) to assess whether surgical repair will more than likely cure the patient's complaints of glare and photophobia?

**Dr. Masket:** In my experience, peripheral iris defects are most commonly iatrogenic and associated with intraoperative floppy iris syndrome in cataract surgery; as

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**Francis W. Price Jr, MD**, is a practicing ophthalmologist at Price Vision Group in Indianapolis, Indiana.

such, they occur temporally, beneath the main incision. These defects can have functional (glare) and cosmetic issues, particularly as the eyelids do not cover them. Prior to a consideration of surgery, I refer the patient to an optometrist familiar with specialized contact lens fitting. In cases of this type, a stock peripherally opaque contact lens (OCL) can be tried in the office, if only to determine whether the OCL alleviates the glare issue. If the trial is successful, a custom OCL can be designed to provide aesthetic as well as functional “cures.” That said, OCLs of this nature are often difficult to wear over long periods, as they are thick and can be uncomfortable. Moreover, having the OCL artificial pupil projected onto the anterior cornea can induce vision problems if the lens slides off center. My success with long-term use of OCLs for managing iris defects is at best 10%; however, they are an excellent option for determining whether iris surgery of some form has a likelihood for success.

**Dr. Price:** Any treatment of an iris defect is a mechanical repair of the normal light-blocking properties of the iris—and in some cases an improvement in cosmetics as well. The artificial irises are quite expensive and, as with any implant, they pose some risks. Therefore, I think it is important to determine the least invasive and least risky treatment. Small peripheral defects can often be treated with suture repairs, either pulling the edges of the defect together or repairing a dialysis. A careful slit-lamp examination is often the best way to determine the best option. The most difficult defects to fix are those where a substantial portion of iris tissue has been lost or the iris is scarred into the cornea so that closing the defect will either place too much tension on the iris attachment to the wall of the eye or cause the sutures to “cheese wire” through the iris. Once inside the eye, intraocular forceps are a good way to judge the ability to pull the edges of the iris into position.

In cases where the defects are necessary peripheral iridotomies, asking the patient to ignore the symptoms is best.

### 3. Please compare the advantages and disadvantages of the Morcher iris devices, Ophtec iris implants, and CustomFlex artificial iris.

**Dr. Masket:** At present, neither Morcher nor Ophtec iris implant devices are available in the US. Furthermore, the CustomFlex artificial iris has only recently received US FDA approval and has not been “rolled out,” making it only available to surgeons who participated in the FDA trial. In the near term, additional surgeons will be trained and approved to use the device.

Nevertheless, at present, Morcher aniridic devices are made solely of black polymethyl methacrylate (PMMA), limiting their cosmetic benefit only to patients with dark irises, although they may be very helpful functionally.

However, a number of these are designed to go into the capsular bag and may fit through a 3.5 mm clear corneal incision. Morcher also manufactures a series of combination IOL/aniridia devices in black PMMA with a clear optical center; these require large incisions. These devices are generally sutured to the sclera and perform well. They cost significantly less than the CustomFlex device.

Ophtec also offers PMMA aniridic IOLs that are available in 3 distinct colors, blue, green, and brown. Nonetheless, the colors are rather vivid and have no iris markings, limiting their cosmetic benefit. Similar to the Morcher single-piece aniridic IOL combinations, the devices are intended to be suture fixated to the sclera. Another shortcoming of the Ophtec device is that the opaque colored area is somewhat small and allows significant light to come around the periphery. Similar to the Morcher aniridic/IOL combination devices, they require large incisions, as they are made of rigid PMMA. Ophtec also manufactures multipiece colored PMMA “buildable” devices that are intended to fit in the capsular bag and fasten together to make a single unit.

The HumanOptics CustomFlex has the advantage of being aesthetically barely distinguishable from the normal human iris, as it is matched to the color and markings of the fellow eye by skilled artisans who “paint” the device by hand. It also is manufactured from medical grade silicone, allowing it to be folded or rolled and inserted via a 3.0-mm corneal incision. The device has an optional fiber meshwork embedded to facilitate suturing to IOLs or the eye wall. As it is only an artificial iris, one must combine it with an existing IOL or mate it with one at the time of surgery. The chief advantages of this product are near perfect aesthetics and the ability to be passed through small incisions. It is significantly more expensive than the Morcher or Ophtec models.

**Dr. Price:** The biggest advantage of the CustomFlex artificial iris for those of us in the US is that it has been approved by the FDA. Secondary advantages are the stunningly normal cosmetic appearance, foldability allowing injection through a 2.8-mm incision, ease of punching to the appropriate diameter, and ability to be used with many different IOL implants. Disadvantages are that it has no haptics and must be sutured to the eye or fixated to a lens implant when not placed in the capsular bag. It is not part of an IOL, so it must be combined with one in aphakic patients unless the patient is to be left aphakic.

The Ophtec lenses only come in 3 solid colors, none of which look natural in darkly pigmented eyes. These implants are made of PMMA and therefore require larger incisions if the model 311 is used. Advantages are that the implant can come with or without an optic; it has haptics that are 13.75 mm in diameter with eyelets that can be fixated to the wall of the eye. The Morcher lenses are solid black and made of rigid plastic. To the best of my knowledge, the company has never sponsored a US clinical trial.

### 4. Cosmetic iris implants used to change eye color have been reported to cause devastating ocular complications. These implants are only available outside the US. How do we educate all patients (in the US and internationally) to avoid these implants?

**Dr. Masket:** Artificial iris implants that are designed to change eye color appearance (BrightOcular) are placed in the anterior chamber angle in otherwise normal eyes. They are used outside the US; they are distinctly not FDA approved. These devices are very commonly associated with corneal decompensation, iris atrophy, cataract, uveitis, and potentially blinding glaucoma. As such, they do not belong in the marketplace and can, unfortunately, taint public opinion against appropriate use of other artificial iris devices that are approved for functional and aesthetic purposes. It should be noted that the latter are only implanted in combination with lens-based surgery and never in the anterior chamber angle. I hope that national and supranational organizations will bring pressure to manufacturers and surgeons who are inducing major eye injury for a profit motive. “Buyer beware” is apparently ineffective, as the as websites continue to solicit unknowing patients.

**Dr. Price:** Artificial iris implants, as discussed earlier, are used to fix mechanical problems with the functional ability of the iris to block excess light coming into the eye. Secondary advantages of these lenses are to provide a normal appearance to an eye that has been essentially disfigured either by trauma or a congenital problem, like aniridia. These implants are intended to be used only at the time of cataract surgery or after the eye has had the natural lens removed. They basically are placed where the natural crystalline lens is.

Cosmetic iris implants typically can be compared to an anterior chamber lens sitting on the iris and in the anterior chamber angle. They are associated with chronic inflammation—as most anterior chamber lenses have been—as well as disfigurement of the normal iris, decompensation of the cornea, and glaucoma. Physicians who place these implants prey upon people seeking to change their appearance and obviously downplay the potential consequences. Education is the best prevention for the public.

### 5. What are the options for treating a patient with aniridia?

**Dr. Masket:** Aniridia can range from small peripheral defects and minor sphincter irregularities to total aniridia and can vary from acquired defects to congenital disorders. There is no single remedy for all conditions. Prior to a consideration for iris surgery, should symptoms warrant, nonsurgical approaches (eg, custom [iris-imprinted] contact lenses or miotic therapy, as appropriate), should be attempted. Surgeons may choose to acquire skills for all types

of iris defects. These include a variety of suture methods (most typically the Siepser sliding knot technique) and use of artificial iris devices, as may be available. Moreover, congenital aniridia has the potential to involve virtually all aspects of the globe (as well as renal tumors) and may require corneal and glaucoma surgery, in addition to management of corneal stem cell failure. Surgeons who encounter aniridic patients infrequently and do not possess the experience or tools for management should consider referral to qualified colleagues. Finally, corneal tattooing may be considered for small peripheral iris defects or for cosmetic management of scarred corneas in eyes with no vision potential.

**Dr. Price:** The options for treating a patient with aniridia are constantly changing and improving. Gene therapy studies have begun for at least the subset of individuals with a nonsense genetic mutation. Further study is needed to determine what that therapy has to offer and when it needs to be initiated. Congenital aniridia entails a number of complex developmental problems, including macular hypoplasia, limbal stem cell dysfunction, loss or disfigurement of the iris, glaucoma, and nystagmus. Some individuals have some but not all of these problems, so the treatment depends on what individual issues each patient or eye has. I have used artificial iris implants in many patients with congenital aniridia with excellent results. Interestingly, even the nystagmus may sometimes improve.

### 6. Please describe the advantages and disadvantages to corneal tattooing as well as the technique.

**Dr. Masket:** I have no personal experience in performing corneal tattooing. It is an old art that is practiced by few colleagues. Over a long career, I have seen less than a handful of patients with corneal tattoos. The technique involves injecting carbon-containing dyes (for black pigment) into the corneal stroma via fine needles or by instillation within a lamellar dissection. It can be used for aesthetic benefit for leucocoria in eyes unsuitable for surgery. I have seen 1 case of peripheral corneal tattoo to remedy dysphopia secondary to laser peripheral iridotomy. Downsides include permanence and negative impact on vision should the treatment extend too close to the entrance pupil. My sense is that it is an underutilized art form.

**Dr. Price:** Corneal tattooing has been used for decades, if not centuries. It essentially has the same limitations of all tattoos: over time (decades). The pigments can migrate and become more diffuse. Duke-Elder had very nice descriptions of these issues. Therefore, patients should be advised that the pigment can change over time, just like a faded tattoo on an elderly person that looked good 50 years earlier. The pigments can be applied in the traditional ways with needles, or now in circumscribed locations created by a femtosecond laser or manual lamellar dissection.



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