

FIBROUS CONGENITAL IRIS MEMBRANES WITH PUPILLARY DISTORTION*

BY *Richard M. Robb, MD*

ABSTRACT

Background: In 1986 Cibis and associates¹ described 2 children with a new type of congenital pupillary-iris-lens membrane with goniodysgenesis that was unilateral, sporadic, and progressive. These membranes were different from the common congenital pupillary strands that extend from 1 portion of the iris collarette to another or from the iris collarette to a focal opacity on the anterior lens surface. They also differed from the stationary congenital hypertrophic pupillary membranes that partially occlude the pupil, originating from multiple sites on the iris collarette, but not attaching directly to the lens.

Case Material: The present report is an account of 7 additional infants with congenital iris membranes, similar to those reported by Cibis and associates, which caused pupillary distortion and were variably associated with adhesions to the lens, goniodysgenesis, and progressive occlusion or seclusion of the pupil. Six of the 7 patients required surgery to open their pupils for visual purposes or to abort angle closure glaucoma. A remarkable finding was that the lenses in the area of the newly created pupils were clear, allowing an unobstructed view of normal fundi.

Conclusion: This type of fibrous congenital iris membrane is important to recognize because of its impact on vision and its tendency to progress toward pupillary occlusion. Timely surgical intervention can abort this progressive course and allow vision to be preserved.

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INTRODUCTION

In 1986 Cibis and associates¹ described a new type of congenital pupillary-iris-lens membrane with goniodysgenesis that was unilateral, sporadic, and progressive. One of their 2 cases developed a secluded pupil with iris bombe, requiring peripheral iridectomy. In 1994 Cibis and coworkers² reported an additional case in which the pupillary-iris-lens membrane was dissected from the lens surface at 3 months of age without cataract formation. As noted by the investigators, these membranes appeared to be different from the congenital pupillary strands that are occasionally seen to cross the pupil from one portion of the iris collarette to another or from the iris collarette to a focal opacity on the anterior lens surface. They also differed from the congenital hypertrophic pupillary membranes that partially occlude the pupil, originating from multiple sites on the iris collarette but not attaching directly to the lens.³⁻⁵

The present report is an account of 7 additional patients with unilateral congenital iris membranes that caused pupillary distortion and were variably associated with adhesions to the lens, goniodysgenesis, and progressive occlusion or seclusion of the pupil. These iris membranes are important because of their impact on vision and because timely surgical intervention may alter their progressive course. The Committee on Clinical

Investigation of Children's Hospital, Boston, approved this medical record review.

CASE REPORTS

CASE 1

A 10-day-old full-term girl was noted by her pediatrician to have a white opacity in the left pupil. On ophthalmic examination at 11 days of age, she appeared to have a white plaque on the anterior surface of the left lens covering the nasal half of the pupil (Fig 1). The nasal portion of the pupillary margin was adherent to this membrane, but the temporal margin was free and there was a clear red fundus reflex in the temporal portion of the pupil. Both pupils constricted to light stimulus, and no afferent pupillary defect could be seen. There was a peripheral iris adhesion to clear cornea nasally at the 9-o'clock position. Retinoscopy revealed 2.50 diopters of hyperopia in each eye, and the fundi appeared normal by indirect ophthalmoscopy.

At 2 months of age, the infant had white plaque occupying three quarters of the pupillary area after dilation, and at 3½ months, less than 1 mm of clear pupil temporally. The child could now fix and follow well with the right eye but not with the left. The left fundus could no longer be seen. Examination of the infant under anesthesia revealed ocular pressures of 9 mm Hg in the right eye and 8 mm Hg in the left eye. The horizontal corneal diameters were 11.25 mm in each eye. On gonioscopy of the left eye,

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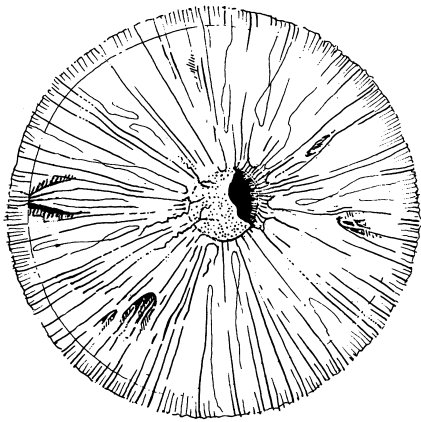


FIGURE 1

Case 1. Drawing of left iris showing white plaque covering nasal half of pupil and peripheral iris adhesion to clear cornea at 9-o'clock position.

the anterior chamber angle was open to the ciliary body band throughout, but there was a localized iris adhesion to Schwalbe's line at the 9-o'clock position. Radial iris vessels extended from the nasal iris onto the white pupillary plaque, which was in contact with the anterior lens surface.

With the infant under the same anesthesia, surgery was performed through a superior limbal incision. An attempt was made to cut the white plaque-like membrane with a Haab knife, but the membrane could not be cut or separated from the pupillary margin. An Ocutome tip was then inserted into the anterior chamber and passed through the small temporal pupillary opening. At this point, the membrane separated cleanly from the underlying anterior lens capsule, and the membrane and some of the adjacent iris were nibbled away to create a larger pupillary opening. A small crescent of iris pigment epithelium remained attached to the lens nasally, but elsewhere the lens was clear and has remained so postoperatively. Part-time patching of the right eye was undertaken between 4 and 9 months of age, and at the end of that time the preferential looking acuity was 20/130 in each eye. The eyes appeared straight and the ocular motility was full.

CASE 2

A 4-month-old boy was referred because of an abnormal pupil in the right eye. A small, eccentric right pupil had been noted at 2 weeks of age, but initial ophthalmologic examination had led to no recommendations for treatment. The child was generally well, and there was no family history of eye disease. At 4 months of age, he fixed and followed visual targets well with the left eye but only poorly with the right eye. A preferential looking test revealed visual acuity of 20/400 in the right eye and 20/180 in the left eye. The eyes appeared straight, and the corneas were similar in size. The left pupil was normal in size and configuration and reacted well to light. The right pupil was 1 mm in diameter. It was

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displaced nasally, and the nasal half of the pupil was occupied by a flat, white membrane that was adherent to the lens (Fig 2). A single iris strand extended from the white membrane above the surface of the iris to an attachment on Schwalbe's line, which could be seen through clear cornea, in the 2-o'clock meridian. A red fundus reflex could be seen through the temporal portion of the pupil, but this disap-

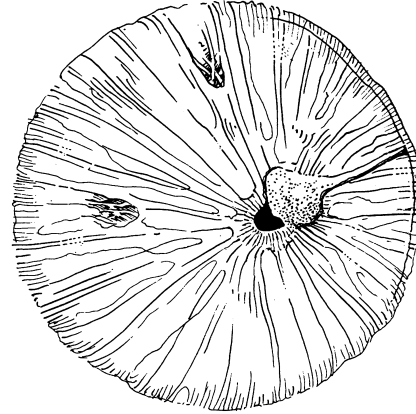


FIGURE 2

Case 2. Drawing of right iris showing nasal displacement of pupil by white membrane that was adherent to lens. A single iris strand extends from white membrane to an attachment to Schwalbe's line nasally.

peared when the pupil constricted to light.

When the infant was 4½ months of age, the pupil of the right eye was enlarged surgically with use of an Ocutome to nibble out the iris in an inferotemporal direction from the free pupillary margin. A pupil approximately 4 mm in diameter was created. The original iridolenticular adhesion remained intact, but the lens elsewhere was clear, and the fundus appeared to be normal. A peripheral iridectomy was performed at the 12-o'clock position.

Preferential looking tests of visual acuity were equal in the 2 eyes postoperatively. Initially the infant's eyes appeared straight, but at 3 years of age a small-angle right esotropia became evident with near fixation. Part-time patching of the left eye was undertaken between the ages of 3½ and 5½ years. At age 6 years, glasses were prescribed as follows: right eye, +1.50 -0.50 x 180, and left eye, plano, with a bifocal add of +2.50 in each eye. The boy wore the glasses until age 9 years, at which time visual acuity was 20/25 in the right eye and 20/20 in the left eye with correction. A small-angle right esotropia with partial binocularity was still present. The right lens remained clear except at its original attachment to the nasal portion of the pupil.

CASE 3

A 3-week-old boy was noted to have a "speck" on his left iris. On examination the speck was found to be a small ectopic pupil pulled toward the upper nasal quadrant by a white iris membrane that attached to a prominent

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Schwalbe's line, visible through clear cornea between the 8- and 12-o'clock positions (Fig 3). A small red fundus reflex could be seen through the ectopic pupil, but the pupil dilated to only 1 mm with cycloplegic agents. The

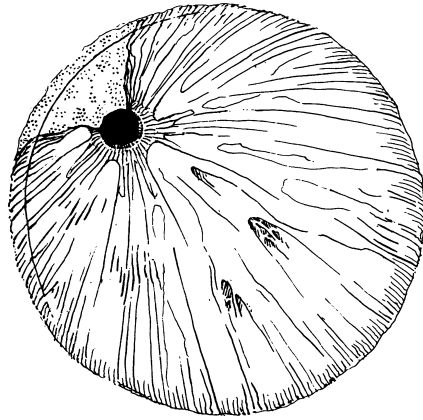


FIGURE 3

Case 3. Drawing of left iris showing small pupil pulled eccentrically by an iris membrane that attached to Schwalbe's line, visible through clear cornea between 8- and 12-o'clock positions.

right pupil was normal in size and configuration, and the lens was clear. Preferential looking test acuities were 20/700 in the right eye and 20/2000 in the left eye.

When the infant was 6 weeks of age, the left pupil was enlarged surgically. Gonioscopy revealed peripheral iris attachments to an anteriorly displaced Schwalbe's line between the 8- and 12-o'clock positions. The nasal pupillary margin was adherent to the lens. An Ocutome was used to extend the temporal margin of the pupil so that the horizontal pupillary diameter was approximately 6 mm. A peripheral iridectomy was performed at the 1-o'clock position. The lens was clear except at the point of the nasal iris adhesion, and the fundus was normal. At the time of surgery, the horizontal corneal diameters were 10.25 mm in the right eye and 9.50 mm in the left eye.

The left lens remained clear postoperatively. At 6 months of age, the preferential looking acuity was 20/88 in each eye. Mild myopia was noted when the infant was 2 years of age, but glasses were not prescribed until he was 7 years old, when refraction was -1.50 in the right eye and -0.50 -1.00 x 180 in the left eye. With this correction, the visual acuity with single letters was 16/16 in the right eye and 16/20 with single letters in the left eye. The eyes were straight, and the patient had binocular vision with stereopsis of 7 minutes of arc.

CASE 4

A 5-week-old girl was first seen because her left pupil was small and did not dilate. On examination she was able to fix and follow with each eye. The right iris and pupil were entirely normal. The left pupil was inferiorly displaced and

approximately 1 mm in diameter. A white membrane at the inferior margin of the pupil formed an attachment between the iris and the lens at that point (Fig 4). Under cycloplegia, the pupil dilated superonasally in an oblique, slit-like fash-

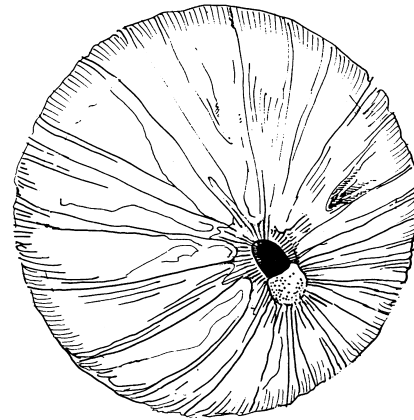


FIGURE 4

Case 4. Drawing of left iris showing inferior displacement of pupil by a white membrane that bound pupil to anterior lens surface.

ion. Retinoscopy through the slit opening revealed 1.25 diopters of hyperopia. The fundi appeared normal.

When the infant was 4 months of age, a preferential looking test revealed an acuity of 20/270 in each eye, but by 4½ months the left pupil had become smaller, the iris was convex, and the anterior chamber was shallow. On examination under anesthesia, the ocular pressures were 12 mm Hg in the right eye and 32 mm Hg in the left eye. Gonioscopy revealed a closed anterior chamber angle in the left eye. Surgery was performed through a limbal incision at the 10-o'clock position. A peripheral iridectomy was made, and the anterior chamber was deepened with Healon. With an Ocutome, an enlarged pupillary opening was created superior to the iris-lens adhesion. The lens was found to be clear in the area of the newly created pupil. Unfortunately, after this surgery fixation with the left eye was unsteady and wandering.

Subsequent examination under anesthesia when the infant was 5½ months of age revealed ocular pressures of 11 mm Hg in the right eye and 10 mm Hg in the left eye. The optic disc could be seen clearly in each eye, and in both eyes the cup-to-disc ratio was 0.4. The refractive error by retinoscopy under cycloplegia was +3.50 -1.00 x 180 in each eye. Part-time patching of the right eye was begun.

At 1 year of age, the infant had a definite left esotropia. Patching of the right eye proved to be very difficult. At 3 years of age, the visual acuity with picture cards was 16/30 in the right eye and 1/30 in the left eye. The left esotropia persisted.

CASE 5

A 3-week-old boy was noted by his pediatrician and a

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local ophthalmologist to have a displaced pupil in his right eye. He was a full-term, otherwise healthy infant. On referral examination his corneas were found to be clear and symmetric. The left iris and pupil were normal in configuration and reaction to light. The right pupil was displaced superiorly. It was small and slit-like and appeared to be pulled toward the upper limbus by a white iris membrane that extended from the margin of the pupil to the anterior chamber angle (Fig 5). A red reflex could be seen

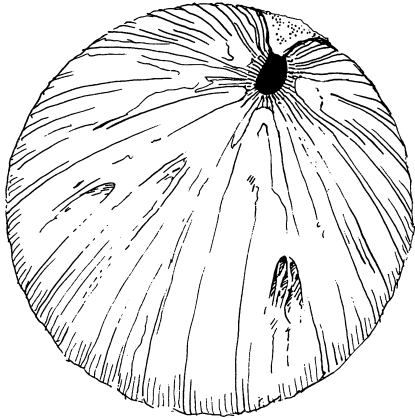


FIGURE 5

Case 5. Drawing of right iris showing displacement of pupil superiorly by a white iris membrane that extended from margin of pupil to anterior chamber angle.

through the slit pupil, and a small constriction to light was noted, but no view of the fundus could be obtained.

At 1 month of age, the child was examined while under anesthesia. The corneal diameters were 10 mm in the right eye and 10.5 mm in the left eye, and the ocular pressures were 10 mm Hg in both eyes. On gonioscopy, the right anterior chamber angle was normal except in the 1-o'clock meridian, where the iris was drawn up by a white membrane to the trabecular meshwork. A surgical incision was made at the 12-o'clock limbus, and with an Ocutome the pupil was enlarged in an inferior temporal direction. The underlying lens was clear, and through the enlarged pupil the fundus was seen to be normal.

Six weeks postoperatively, the child's preferential looking acuity was 20/1,000 in the right eye and 20/270 in the left eye. Part-time patching of the left eye was begun. By 4 months of age, the preferential looking acuity had improved to 20/180 in each eye, and at 7 months it was 20/130 in each eye. Patching was discontinued. The eyes appeared straight. A refraction at 14 months of age revealed mild hyperopic astigmatism, +2.50 -1.00 x 80 in the right eye and +2.00 -1.00 x 90 in the left eye. No glasses were prescribed. When the patient was last seen at age 7 years, the visual acuity was 20/25 in each eye. The eyes were straight, and there was binocular vision with stereopsis of 7 minutes of arc on the random dot E. The elliptical

pupil in the right eye showed only a slight constriction to light, but there was no relative afferent pupillary defect.

CASE 6

A 3-month-old boy was referred for ophthalmologic evaluation because of an irregular left pupil. He had been delivered with the aid of forceps, but at birth there were no forceps marks around the eyes. The left pupil was displaced temporally, and a strand of tissue extended from the pupil to the anterior chamber angle at the 3-o'clock position. Between the 2- and 4-o'clock positions, an anteriorly displaced Schwalbe's line could be seen through clear cornea. A small red fundus reflex could be seen through the oval eccentric pupil, but there was no view of the fundus and retinoscopy could not be performed. The anterior chamber was of average depth. The anterior segment and fundus of the right eye were normal. Ocular pressures were 15 mm Hg in each eye.

Because the left pupil appeared to be getting smaller with time, an examination of the infant under anesthesia was done at 6 months of age. The horizontal corneal diameters were found to be 11 mm in the right eye and 10.5 mm in the left eye. Gonioscopy revealed peripheral iris adherent to the anterior chamber angle from the 2- to 4-o'clock positions, a fibrous pupillary membrane, and a band extending from the membrane to Schwalbe's line temporally. Through a limbal incision, a full iridectomy was created with scissors from the 9- to 12-o'clock positions, extending centrally to the displaced pupil. In the area of the iridectomy, the lens was clear. The fundus was normal.

By 7 months of age the left eye was noted to be exotropic. Part-time patching of the right eye was begun. Initially the left eye was mildly hyperopic, but by age 1½ years, the refraction was plano in the right eye and -1.50 in the left eye. Glasses were prescribed, and efforts to patch the right eye were continued. By age 6 years, the left exotropia had increased to 40 prism diopters. The Snellen acuity with correction was 20/25 in the right eye and 20/50 in the left eye. Recession of the left lateral rectus muscle and resection of the left medial rectus were performed. Patching was not resumed postoperatively, and a small residual exotropia remained.

By age 15 years, myopic astigmatism had developed in both eyes: -3.25 -2.00 x 180 in the right eye and -3.25 -2.00 x 180 in the left eye. The corrected visual acuity was 20/20 in the right eye and 20/50 in the left eye. Ocular pressures were 17 mm Hg in both eyes. The patient chose to wear contact lenses, and when he was last seen at age 20, visual acuity was unchanged.

CASE 7

A 2-month-old girl was referred by her pediatrician and a

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local ophthalmologist because of anterior segment abnormalities in the right eye. The child was a full-term healthy infant, and there was no family history of eye disease. On examination, the right cornea appeared slightly smaller than the left. A slightly elevated dermoid, 3 mm in diameter, at the inferior limbus extended only 1 mm onto clear cornea. The pupil was displaced toward the upper temporal quadrant, and from the 9- to 11-o'clock positions, the margin of the pupil was devoid of its pigmented layer and was attached to the lens by a white membrane (Fig 6). From this membrane a thin band of fibrous tissue extend-

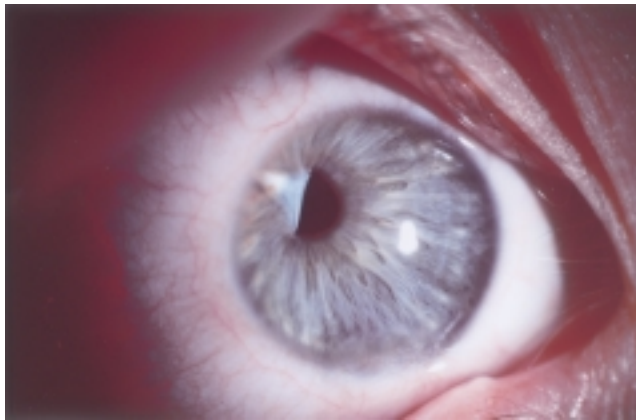


FIGURE 6

Case 7. Photograph of right eye showing displacement of pupil toward upper temporal quadrant and a white membrane attaching iris to lens and extending peripherally to anterior chamber angle. Photograph was taken after a small inferior limbal dermoid had been removed. No iris surgery was performed.

ed peripherally above the surface of the iris to the anterior chamber angle. The portion of the pupil that was not bound to the lens could be dilated, and through the dilated pupil the lens appeared to be clear and the fundus normal. The anterior segment of the left eye was normal. The eyes were straight, and the child fixed and followed visual targets well.

At age 1 year, fixation with the right eye was noted to be reluctant, and a myopic retinoscopic reflex was seen in the right eye. Examination with the infant under anesthesia was therefore carried out. The corneal diameters were 11 mm in the right eye and 11.25 in the left eye. Ocular pressures were 9 mm Hg in the right eye and 11 mm Hg in the left eye. Cycloplegic retinoscopy revealed refractive errors of -4.00 in the right eye and +1.00 in the left eye. Gonioscopy confirmed that the anterior chamber angle of the right eye was open throughout its circumference except where the band extended from the iris surface to attach to the angle structures at the 10-o'clock position. On either side of the band the peripheral iris stroma was thinned, and several knuckles of the underlying pigmented iris could be seen. Both fundi were normal

except for a slightly less prominent foveal reflex in the myopic right eye.

After this examination the right pupil was kept dilated with atropine. Glasses were prescribed, and the left eye was patched part-time. This treatment was carried out with varying success until age 4 years. A right exotropia developed, and the myopia in the right eye increased to -6.00 diopters. Visual acuity at age 4 years was 3/30 in the right eye and 20/30 in the left eye with single picture cards. At this point, the limbal dermoid was excised from the inferior cornea. Despite efforts to continue with glasses and occlusion of the left eye, the visual acuity of the right eye at age 8 was only 20/300 with correction. Glasses were discontinued, and no further attempts at patching were made.

DISCUSSION

The iris membranes described in these 7 cases are congenital, unilateral, and sporadic. They arise in the iris but not specifically from the iris collarette. They involve only a sector of the iris and create an attachment of the pupil to the anterior surface of the lens in that sector, distorting the pupil and pulling it into a variably eccentric position. The pupil may be small and slit-like, dilating only in that portion of its circumference not occupied by the membrane. A strand of the membrane often extends from the iris surface peripherally toward the anterior chamber angle, attaching to an anteriorly displaced segment of Schwalbe's line. In some cases, the membrane appears to be progressive. It may extend over the pupil (case 1) or may constrict the pupil (case 6) to the point that no useful optical path remains. In other cases, progression of the membrane leads to blockage of aqueous flow from the posterior chamber to the anterior chamber, and iris bombe results (case 4).

Pupillary dilatation may be useful in some patients to provide a better optical opening (case 7),⁶ but more often surgical enlargement of the pupil has been necessary to restore vision or to treat angle closure glaucoma. Forceps and scissors may be used to perform the surgery (case 6),⁷ but a suction-cutting instrument that can be inserted into the anterior chamber allows a more controlled iridectomy and protection of the underlying lens (cases 1 through 5).⁸ Except for the point at which the membrane attaches to the lens, the lens has been clear in the area of the newly created pupil, and it has remained so. The fundi have been normal when viewed postoperatively. A small limbal dermoid in 1 patient (case 7) was the only other ocular abnormality found in our cases, except for a slightly smaller corneal diameter compared with the fellow eye (cases 3, 5, 6, and 7). No systemic disorders have been associated with the condition, nor have any abnormalities of pregnancy been identified.

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The kind of iris membrane described in the cases presented here is different from what has been called a hyperplastic pupillary membrane.^{3,5} The latter arises from the collarette of the iris and usually does not attach to the lens but covers the central pupil to a varying degree. Attachments to the anterior chamber angle are not found. These hyperplastic membranes are occasionally familial in nature.³ Arguments for and against surgery for them have been advanced, but improvement of vision with surgery has been modest when preoperative and postoperative visual acuities have been available for comparison.⁴ Another congenital pupillary abnormality, called congenital idiopathic microcoria by Lambert and associates,⁸ is similar to but not identical with the abnormality in our patients. Their 5 patients had a unilateral white fibrous membrane present at the pupillary margin, and 4 of the 5 had eccentric pupils. The lenses, however, were reported to be uninvolved in the 4 patients who had pupil surgery, and no anterior chamber abnormalities were described. The investigators recommended early surgical treatment and vigorous occlusion therapy. Reynolds and colleagues⁷ reported 2 patients with congenital pupillary membranes, the second of which had a white pupillary membrane, a tiny vertically eccentric pupil, and a filamentous adhesion to Schwalbe's line inferiorly. Surgery was performed in this case at 6 months of age, and the lens was described as clear postoperatively. Finally, the 3 cases described by Cibis and associates^{1,2} with a congenital pupillary-iris-lens membrane with goniodysgenesis, and 1 additional case in the German literature⁹ referred to by Cibis, all seem to have had the condition described in this paper. One of Cibis's patients had surgical removal of the membrane from the lens without cataract formation; another required a peripheral iridectomy for impending angle closure glaucoma.

The etiology and pathogenesis of the membranes described are uncertain. They are present at birth, but are unilateral and nonfamilial. Fluorescein angiography has shown vessels in the membrane and abnormal leakage over the membrane and at the pupillary margin.¹ The histology of 1 pupillary membrane revealed stromal fibroblasts, blood-bearing vessels, and perhaps aberrant smooth muscle, indicating that the tissue was derived from iris and not lens.² Cibis postulated disruption of the normal evolution of the embryonic pupillary membrane, possibly due to a vascular occlusion, as the cause of the abnormal membrane.² Mann¹⁰ suggests that fetal iritis may lead to pupillary membranes that cause an attachment of the margin of the pupil (rather than the collarette) to the lens, but her illustrative cases do not resemble the cases described here. It does seem likely that some kind of aberrant persistence of the embryonic anterior tunica vasculosa lentis, other forms of which have been described by Goldberg in an account of persistent fetal vasculature in the eye,¹¹ is involved in the formation of this distinctive iris membrane.

CONCLUSION

The condition I have termed fibrous congenital iris membrane with pupillary distortion is an entity that has now been recognized by a number of investigators. It frequently interferes with the development of normal vision. Occasionally, it leads to seclusion of the pupil and angle closure glaucoma. In either circumstance, early surgery to create an enlarged pupil should be performed. Occlusion or seclusion of the pupil may be progressive in the early months, and cases in which the pupil can initially be dilated successfully should be observed closely for progression. An important feature of the condition is that the lens is clear except in the small area of iris-lens adhesion, and good vision can be obtained through a surgically enlarged pupil if amblyopia is not allowed to progress to an irretrievable level.

ACKNOWLEDGEMENTS

The author thanks Anne B. Fulton, MD, for contributing case 6 for this study, and Robert D. Gross, MD, for providing the photograph of case 7. Harriet R. Greenfield drew the illustrations for cases 1 through 5 using sketches and verbal descriptions from their medical records.

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DISCUSSION

DR MORTON F. GOLDBERG. I, too, have seen patients with fibrous congenital iris membranes with pupillary distortion.

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I believe that they are similar, and probably identical in pathogenesis, to the cases reported by Cibis and colleagues in 1986,¹ and to those reported by Dr. Robb. I do not believe, however, that this entity represents a new or unique disease. Rather, I believe it to be a manifestation of persistent fetal vasculature (PFV) in the anterior segment.

A review of ocular embryology shows that the key vascular structures in the anterior segment include the anterior tunica vasculosa lentis, the posterior tunica vasculosa lentis, and the intervening radial anastomoses between these two structures, known as iridohyaloid arteries.² Abnormal persistence of any of these vessels, causes well-known congenital anomalies such as persistent pupillary membrane, which may infrequently bleed and become scarred, or which may persist unchanged, or (usually) regress. Indeed, 30-95% of normal individuals have some persistence of the so-called pupillary membrane, a vascular remnant of fetal life, although visually disabling hyperplasia and scarring of this tissue are far less common. If these tissues do become scarred, a variety of physical signs may occur, including white membranes, posterior synechiae, and seclusion or occlusion of the pupil, leading, in some cases, to pupillary distortion or to secondary glaucoma of various types.

Clues to the underlying fetal and vascular origin include the radial configuration of the iris bands and associated posterior synechiae, as seen in several cases presented by Robb and Cibis et al., as well as the hairpin, recurving configuration of scar or vascular tissue around the sphincter of the pupil in any meridian. The presence of such congenital radial bands or scars implicates persistence of fetal vasculature, even if these structures are not confined or attached to the collarette of the iris.

Occasionally, additional clues include shorter axial lengths, smaller corneas, and asymmetries of the discs, maculas, or posterior poles. In fact, in Dr Robb's series, 4 of 7 cases showed such asymmetry in corneal diameters, a frequent finding in persistent fetal vasculature.

In summary:

1. I believe Dr Robb's observations are indeed similar to those of Dr Cibis and his colleagues.
2. Clues, such as congenital micro-cornea and vascular remnants of fetal life (which are often radially oriented), are helpful in making the correct diagnosis of persistent fetal vasculature that affects primarily, or only, the anterior segment.
3. Postnatal changes of a tertiary nature, such as scar tissue formation or remodeling, may occur, necessitating surgical intervention.
4. The presence of a clear lens is encouraging, but does not guarantee good visual results following anatomically successful anterior segment surgery, due to

associated PFV or other malformations in the posterior segment of the eye.

5. Finally, and perhaps most importantly, the simple presence of a defect in the eye at the time of birth does not necessarily relegate such an eye to permanently reduced vision. If surgery is done in a timely and uncomplicated fashion, and is followed by successful therapy of organic, anisometric, or strabismic amblyopia, good visual results may occur. Thus, Dr Robb has provided a useful service in stressing that these uncommon, late, tertiary events occasionally require surgery and that the surgery can be both anatomically and visually successful.

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[Editor's Note] DR GERHARD W. CIBIS feels that these cases are much more than persistence of primary fetal vasculature and in some cases may involve abnormalities of migration, proliferation, and regression of iris tissue; he showed cases to support this hypothesis.

DR RICHARD M. ROBB. Thank you, Dr Goldberg, for your careful reading of the manuscript and interpretation of how persistent fetal vasculature might be involved in the formation of these membranes. With regard to visual potential I would add that, so far, in the cases I have seen there have been no fundus abnormalities that I have recognized, except in 1 case where there was marked anisometropia and a flattened fovea in the more myopic eye. So I am still encouraged by the potential for good vision in these cases, and I agree with Dr Cibis that it is important to recognize the distinctive iris abnormality and to follow the patients carefully. Most congenital pupillary membranes that we see as pediatric ophthalmologists are looked at once and discounted. But these fibrous membranes are really different, and the fact that some of them are progressive means that you have to look repeatedly in the first weeks and months to be sure that you don't lose the opportunity to obtain a good visual result. These cases are uncommon, but the fact that several of us have seen them in some number, and that there are others reported in the literature, means that they are not rare. I believe many of you will encounter them. Thank you.

