

# Iridoschisis – A case report

Iridoschisis is a rare condition where a localised area of iris stroma is cleaved in two, with the anterior atrophic portion disintegrating into fibrils. The most common presentation of iridoschisis is an age-related iris atrophy in association with glaucoma. The association includes angle closure glaucoma, angle recession glaucoma and open angle glaucoma. A finding of iridoschisis is a warning to the clinician to assess the patient for pre-existing glaucoma and to monitor for glaucomatous changes, as it is unclear if iridoschisis is a cause or an effect of raised intraocular pressure (IOP).

## Definition

Iridoschisis is defined as a separation of the anterior iris stroma from the posterior stroma and muscle layers. The anterior layer, which contains blood vessels, splits into strands and the loose ends wave in the aqueous. Iridoschisis may appear unilateral in the first instance but is generally bilateral.

Schmitt first reported iris splitting into two layers in 1922<sup>1</sup> - but the term 'iridoschisis' was first proposed by Loewenstein and Foster in 1945<sup>2</sup>. Loewenstein described a 76-year-old woman with end-stage glaucoma who had an inferior localised cleavage of the iris into two layers, with the anterior fibrils floating freely in the anterior chamber.

Since then, there have been 72 reported cases of iridoschisis, of which 47 have been associated with different forms of glaucoma. Iridoschisis has been reported in association with angle closure glaucoma (28)<sup>3,4,5,6,7</sup>, angle recession glaucoma (2)<sup>8</sup>, open angle glaucoma (2)<sup>9,10</sup>, and in 15 cases the nature of the glaucoma was unclear<sup>4,5,11</sup>. Iridoschisis reported in patients with keratoconus (2)<sup>12,13</sup> and penetrating keratoplasty for keratoconus (1)<sup>14</sup> could be a coincidental finding given the high incidence of keratoconus.

However, there could possibly be a common connective tissue disorder. Iridoschisis has been reported in cases with microphthalmos (2)<sup>15,16</sup>, and syphilitic interstitial keratitis (1)<sup>17</sup>, but these are probably chance associations. Other cases of iridoschisis following ocular trauma (3)<sup>4,9,16</sup> have been reported. Iridoschisis may be an idiopathic iris atrophy in old age<sup>2,3,4,8,12</sup> as the majority of patients reported are in their 70s, but there have been three reports of juveniles with iridoschisis<sup>4,15,16</sup>.

Iridoschisis is not gender specific and there does not appear to be genetic involvement<sup>18</sup>. However, Danias et al<sup>18</sup> did report a mother with glaucoma and iridoschisis and a daughter without glaucoma but with iris stromal rarefaction, shown by high frequency ultrasound imaging (HFU).

## Case report

A 76-year-old male presented with a persistent swelling over the naso-lacrimal sac and epiphora in the right eye (OD) of several months duration. The left eye (OS) was normal. Visual acuity was count fingers (CF) OD and 6/6 OS. The history revealed that the patient had amblyopia (OD) and had contracted Tuberculosis (TB) at 26 years

of age. He was on medication for chronic obstructive airways disease (COAD), heart disease and fluid retention. His medications were Theodur, Digoxin, Dyazide and Solprin.

There was a firm non-tender mass over the right naso-lacrimal sac. There was no discharge and a probe could only travel 5mm down the lacrimal duct. A saline rinse of the duct did not produce saline to the pharynx. The Jones 1 test produced no dye. A diagnosis of a small right dacryocystocele was made.

Intraocular pressures (IOPs) in each eye were 15mmHg by applanation. Slit lamp biomicroscopic examination revealed bilateral iris atrophy in the inferio-nasal quadrant. A localised cleavage of the iris stroma into two layers was noted (**Figure 1**). The torn ends of the anterior stromal fibres were floating freely in the anterior chamber (**Figure 2**). Otherwise the iris appeared normal except for the area of atrophy. The patient was

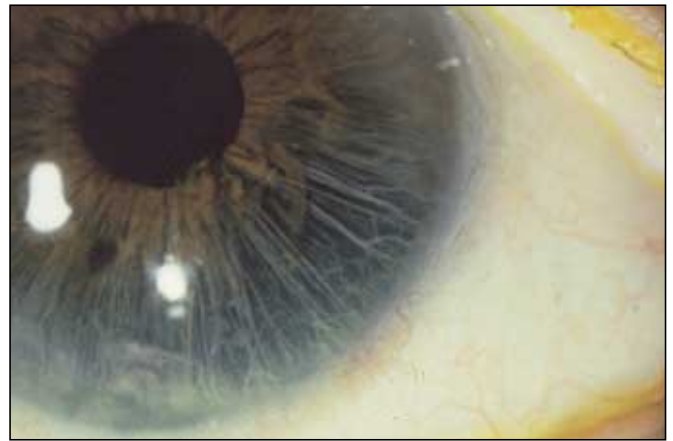
thoroughly assessed for glaucoma using visual field assessment, IOP, gonioscopy, optic nerve assessment and photo documentation. There was no evidence of glaucoma. Considering the above findings and the patient's age, a diagnosis of idiopathic iridoschisis was made.

As the patient could cope with his epiphora, no action was taken to remove the dacryocystocele. It was decided that the patient should be reviewed on a six-monthly basis for glaucomatous changes.

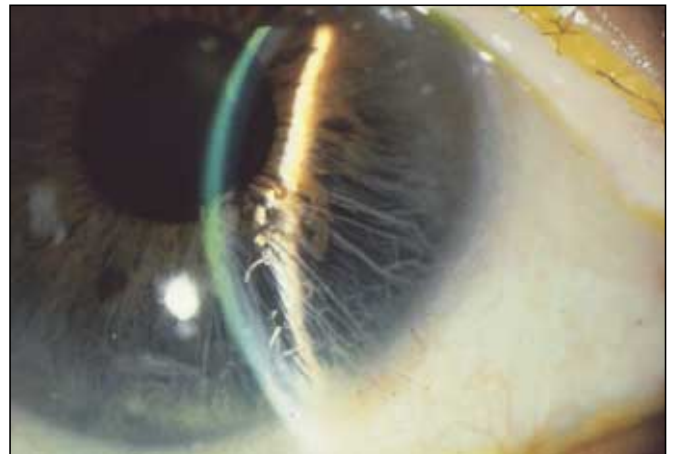
## Discussion

### Pathology of iridoschisis

Iridoschisis is characterised by separation of the anterior and posterior stromal sections of the iris. The anterior leaf usually splits into small strands, which wave freely in the anterior chamber. Larger free sections may contain blood vessels. Carnevalini et al



**Figure 1:** Low magnification view of the iris in iridoschisis showing anterior leaf degeneration, from 3 o'clock to 8 o'clock. The remainder of the iris is not involved



**Figure 2:** Slit view of iridoschisis showing loose ends of broken anterior stromal leaf fibrils floating freely in the anterior chamber

reported normal perfusion of blood vessels, from the inner pupillary margin to the outer iris of the affected sectors<sup>19</sup>. Occasionally, separation at the posterior stroma might occur with irregular pigmentation of the pigment layers<sup>5</sup>.

Electron transmission microscopy of the iris reported by Rodrigues et al<sup>5</sup> showed significant thinning of the stroma and reduced collagen fibrils in the affected area. The appearance of the stromal blood vessels and nerves in the affected area was normal. A change to the cornea was uncommon<sup>5</sup>. However, when present, degenerated corneal endothelial cells were localised over the area of iridoschisis, where the strands of degenerated iris might come into contact with the corneal endothelium<sup>5</sup>.

### Glaucoma and iridoschisis

Glaucoma is associated with iridoschisis in 65% of reported cases<sup>4,5,7,11</sup>. Angle closure glaucoma has been reported in approximately 40% of patients<sup>18,7</sup>. In a number of cases, iridoschisis preceded the angle closure episode<sup>7</sup>. Iridoschisis subsequent to angle closure glaucoma in patients has been reported by others<sup>2,3</sup>. Several reports did not make clear the order of events<sup>4,5,11</sup>.

The mechanism by which iridoschisis may cause angle closure is unclear. However, atrophy of the anterior stromal fibres of the iris may result in the free fibres bowing forward and coming into contact with the corneal endothelium. This could cause elevation of IOP with angle closure. Danias et al<sup>18</sup> have shown through HFU how the

intact posterior pigment epithelium may drape over the anterior capsule of the lens and cause a pupillary block and how the separated anterior stromal fibres may bow forward, obstructing the angle.

The causal relationship of raised IOP with iridoschisis is not known, but a few theories about the mechanisms have been proposed, which will be addressed in the following section.

### Aetiology of iridoschisis

The small number of cases of iridoschisis in the literature gives inconclusive evidence as to the aetiology of the disorder, but the following theories have been proposed. Loewenstein and Foster<sup>2</sup> suggest that the disorder is an idiopathic change of old age. They also propose that proteolytic enzymes resulting from the glaucomatous conditions may aggravate the disorder. Payne and Thomas<sup>10</sup> suggest that, as their patient was on prolonged miotic treatment for glaucoma, the mechanical shearing action caused tearing of the iris stroma. However, miotics have been the mainstay of glaucoma therapy for decades and few miotic users develop iridoschisis.

Iridoschisis resulting from trauma has been proposed by Loewenstein, Foster and Sledge<sup>20</sup>. They postulate that trauma may cause a spike of increased IOP which shears along the dilator fibres, splitting the anterior and posterior portions of the iris stroma. This is unlikely to be a common cause of iridoschisis given the bilateral nature of the condition. Bøjer<sup>6</sup> suggests that iridoschisis is a form of essential iris

atrophy in the elderly. As mentioned below, progressive iris atrophy begins in the third decade of life, whereas iridoschisis is seen mainly in the seventh decade. Albers and Klein<sup>21</sup> proposed that, as the sclerosis of blood vessels increases in the anterior iris stroma, a shearing action tears the tissue between the anterior and posterior sections during constriction and dilatation of the iris. Rodrigues<sup>5</sup> and Carnevalini<sup>19</sup> have shown that both the vasculature and its perfusion are normal in iridoschisis, in contrast to the vasculature of essential iris atrophy.

The authors conclude that the cause and the mechanism of iridoschisis remain unclear.

### Differential diagnosis of iridoschisis

Diagnosis of iridoschisis is based upon a slit lamp biomicroscopic examination and a history. Iridoschisis should not be confused with the two other main iris stromal anomalies, namely, the iridocorneal endothelial syndrome (ICE) and neurocristopathy of the iris and cornea (Axenfeld-Rieger)<sup>19</sup>. **Table 1** shows the main differential characteristics of iridoschisis.

### ICE syndromes

In the ICE syndromes, there are three rare, overlapping conditions - essential iris atrophy, iris naevus (Cogan-Reese syndrome) and peripheral anterior synechiae and corneal oedema (Chandler's syndrome)<sup>22</sup>. An abnormal corneal endothelium that migrates across the chamber angle on to the surface

**Table 1: Differential characteristics of iris stromal disorders**

ITEM	ICE <sup>1</sup>	IRIDOSCHISIS	AXENFELD-RIEGER
Iris	Stromal atrophy with or without hole, PAS <sup>2</sup> extending to Schwalbe's line, Naveus	Stromal splitting without hole, Sectorial & mainly inferior, Normal vasculature and perfusion, Anterior stromal atrophy and strands float in anterior chamber	Mild stromal thinning, Atrophy and hole formation, Hypoplasia, PAS <sup>2</sup> to prominent anteriorly displaced Schwalbe's line, Sphincter visible
Pupil	May be ectopic with PAS, <sup>2</sup> May be displaced towards PAS, <sup>2</sup> May be polycoria	Round to reactive to light and accommodation	Irregularity varies
Cornea	Abnormal endothelium Pleomorphism & cell loss	Changes uncommon, possible endothelial damage where anterior strands touch	Endothelial changes to PAS, <sup>2</sup> Oedema
Age of onset	3rd or 4th decade	Usually 6th or 7th decade	Birth
Laterality	Strictly monocular	Generally binocular, Symmetrical	Binocular and asymmetrical
Congenital	No	No	Yes
Progressive	Yes	Yes	No
Glaucoma	Occurs as disease progresses	Associated with angle closure ~40%, any type ~65%	~50% of cases
Other anomalies	None	None	Teeth, facial and musculoskeletal malformation of trabecular meshwork and Schlemm's canal

<sup>1</sup>ICE = Iridocorneal endothelial syndrome

<sup>2</sup>PAS = Peripheral anterior synechiae

of the iris is the common feature of the ICE syndromes<sup>23</sup>. Specular microscopy has shown the corneal endothelium to have variable cell numbers, to be pleomorphic and to resemble corneal guttata<sup>24</sup>.

In essential iris atrophy, there is progressive angle closure as peripheral anterior synechiae (PAS) form. As the iris stroma atrophies, a full thickness iris hole forms opposite the PAS. This causes the pupil to be displaced towards the area of PAS<sup>22</sup>. Normal stroma may be seen between the atrophied stroma.

Iris naevus shows a diffuse solid mass covering the anterior iris. It replaces the stroma locally, but does not show prominent vessel structure, ectropian iridis or progression<sup>25</sup> (i.e. it is not malignant). When the naevus is close to the anterior angle, it may close the angle with PAS, in a similar manner to that of essential iris atrophy<sup>26</sup>. Iris hole is not normally a feature in this presentation.

Chandler's syndrome is characterised by PAS, corneal oedema at the point of attachment of the PAS to the corneal endothelium and monocular glaucoma. Iris hole formation is not a feature in this syndrome<sup>26</sup>. The iris may appear normal or it may display mild stromal atrophy and corectopia<sup>27</sup>. This condition can be part of the above syndromes in complicated cases and may be difficult to differentiate.

### Neurocristopathy of the iris and cornea

In neurocristopathy of the iris and cornea, there is hypoplasia of the iris stroma with filaments connected to abnormal peripheral cornea<sup>28</sup>. Variants of this condition include Axenfeld's, Rieger's and Peter's anomalies. The Axenfeld's anomaly has a prominent Schwalbe's ring (posterior embryotoxon) at the peripheral cornea, iris strands extending to the ring, normal anterior iris stroma and pupil. Rieger's anomaly displays an indistinct corneoscleral limbus, an irregular, centrally displaced, prominent Schwalbe's ring visible nasally and temporally, gross iris strands connecting to Schwalbe's ring with a normal appearance to the angle behind the iris strands, an oval pupil, thin iris stroma with loss of normal crypts and folds, and prominently visible iris sphincter<sup>27, 28</sup>. Peter's anomaly has a central corneal defect, with stromal thinning and absence of Descemet's membrane and endothelium. There are usually iris strands adherent to the edge of the posterior corneal defect<sup>28</sup>. The neurocristopathies<sup>27</sup> are typically bilateral and the above anomalies have associated defects of teeth, facial bones, and/or musculoskeletal system.

### Management of iridoschisis

The detection of iridoschisis does not indicate that any intervention is necessary. However, as iridoschisis is associated with

the family of glaucomas, it is essential that the clinician check thoroughly for glaucoma. The standard glaucoma tests should include visual acuity, visual field assessment, gonioscopy, biomicroscopy, and ophthalmoscopy with a careful assessment of the optic nerve head and tonometry. Iridoschisis is progressive and glaucoma may develop subsequently, so that six monthly or annual follow-up for glaucomatous change is required. Photo-documentation will assist the longitudinal assessment of iridoschisis.

### Conclusion

An elderly patient presented with a small right dacryocystocele and iridoschisis. The finding of iridoschisis is a warning for the practitioners to be vigilant at the regular review and to do standard glaucoma tests. Glaucoma was not a feature in this patient, but as 65% of patients with iridoschisis have glaucoma, it is possible that this patient could develop glaucoma in later years.

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