Iris cysts in children: classification, incidence, and management

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Abstract

***Background*—Iris cysts in children are uncommon and there is relatively little information on their classification, incidence, and management.**

***Methods*—The records of all children under age 20 years who were diagnosed with iris cyst were reviewed and the types and incidence of iris cysts of childhood determined. Based on these observations recommendations were made regarding management of iris cysts in children.**

***Results*—Of 57 iris cysts in children, 53 were primary and four were secondary. There were 44 primary cysts of the iris pigment epithelium, 34 of which were of the peripheral or iridociliary type, accounting for 59% of all childhood iris cysts. It was most commonly diagnosed in the teenage years, more common in girls (68%), was not recognised in infancy, remained stationary or regressed, and required no treatment. The five mid-zonal pigment epithelial cysts were diagnosed at a mean age of 14 years, were more common in boys (83%), remained stationary, and required no treatment. The pupillary type of pigment epithelial cyst was generally recognised in infancy and, despite involvement of the pupillary aperture, also required no treatment. There were nine cases of primary iris stromal cysts, accounting for 16% of all childhood iris cysts. This cyst was usually diagnosed in infancy, was generally progressive, and required treatment in eight of the nine cases, usually by aspiration and cryotherapy or surgical resection. Among the secondary iris cysts, two were post-traumatic epithelial ingrowth cysts and two were tumour induced cysts, one arising from an intraocular lacrimal gland choristoma and one adjacent to a peripheral iris naevus.**

***Conclusions*—Most iris cysts of childhood are primary pigment epithelial cysts and require no treatment. However, the iris stromal cyst, usually recognised in infancy, is generally an aggressive lesion that requires treatment by aspiration or surgical excision.**

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The classification, incidence, and management of iris cysts has been covered in recent reports.[1–4] Iris cysts are classified as primary or secondary types, with the primary cysts being further divided into pigment epithelial or stromal types.[1] Most iris cysts arise from the iris pigment epithelium in adults but the iris stromal cyst characteristically appears in young children. Although there have been several reports on iris stromal cysts in children[5–17] little has been written about the incidence, natural course, and management of the various types of cysts that occur in childhood. This study was undertaken to determine the types of iris cysts that occur in the first two decades of life and to elucidate their natural course and management.

Patients and methods

A retrospective chart review was done on all patients coded in our computerised files with the diagnosis of primary iris cyst from 1 January 1974 to 31 December 1996. We extracted from that large series of primary iris cysts all cases of patients who were under 20 years of age at the time of diagnosis which we defined, for the purpose of this study, as childhood iris cysts. These childhood iris cysts were categorised according to a slight modification of a previously published classification of iris cysts (Table 1).[1] Follow up information was obtained on these children by chart review or by contacting the referring physicians. In addition to our comprehensive review of primary iris cysts, we identified in our files four secondary iris cysts that occurred in children under age 20 and they were included in this survey. We determined the incidence of the various iris cysts of childhood as well as the age and sex of the affected patient and the natural course and management of the lesions.

Results

Among 251 patients coded with the diagnosis of primary iris cyst, there were 53 in whom the lesion was diagnosed before age 20 years. In

Table 1 Classification of iris cysts

I Primary
A Primary cysts of iris pigment epithelium
1 Peripheral (iridociliary)
2 Mid-zonal
3 Central (pupillary)
4 Dislodged
a Fixed
b Free floating
(i) Aqueous
(ii) Vitreous
B Primary cysts of the iris stroma
1 Congenital
2 Acquired
II Secondary
A Traumatic
B Tumour induced
C Parasitic

Table 2 Clinical data on 57 iris cysts in children

	Number	% Children with cysts	Age (years)		Sex	
			Mean	Range	Male	Female
I Primary iris cysts (53)						
A Primary cysts of iris pigment epithelium (44)						
1 Peripheral (iridociliary)	34	59	15	5–19	11	23
2 Mid-zonal	5	9	9	1–19	5	1
3 Central (pupillary)	3	5	13	1–19	3	0
4 Dislodged						
a Fixed	0	0	—	—	—	—
b Free floating	2	4	16	12–19	1	1
B Primary stromal iris cysts (9)						
1 Congenital	9	16	2	0–7	4	6
2 Acquired	0	0	—	—	—	—
II Secondary iris cysts (4)						
A Traumatic	2	4	8	2–13	0	4
B Tumour induced	2	4	1–	<1		
C Parasitic	0	0	—	—	—	—

addition, there were four secondary iris cysts, making a total of 57 iris cysts of childhood. The classification that we employed for childhood iris cysts is shown in Table 1.

The incidence and clinical data on these 57 patients is shown in Table 2. Of the 53 primary iris cysts, 44 were pigment epithelial and nine were stromal. The peripheral pigment epithelial cyst accounted for 34 cases or 59% of all childhood iris cysts. It was generally detected in the teenage years, was more common in girls (68%), and was usually found on routine slit lamp examination as an anterior bulging of the peripheral iris.[1] Since this cyst is difficult to visualise directly, we have recently used ultrasound biomicroscopy to confirm the cystic nature of the lesion (Fig 1). The mid-zonal cyst accounted for five cases (9%), was diagnosed in older children, was more common in boys (83%), remained stable and required no treatment. It appeared as a dark mass that became more fusiform in shape with dilatation of the pupil (Fig 2). The affected patient was usually referred because of concern about a ciliary body melanoma. The central (pupillary) cysts accounted for three cases (5%). All three were initially detected in infancy, remained stable, and the patient was referred to us during teenage years for a diagnostic opinion. Two were bilateral and multiple (Fig 3) and one was solitary. The multiple pupillary cysts often appeared to have ruptured or deflated (iris flocculi). There were two dislodged cysts, one of which floated freely in the aqueous (Fig 4) and one in the vitreous. All of these pigmented epithelial cysts either remained stable or regressed after they were diagnosed and none of them required treatment.

There were nine cases of primary iris stromal cysts, which accounted for 16% of all childhood iris cysts (Table 2). There was no history of amniocentesis or ocular trauma in these children. The iris stromal cyst was readily seen in the anterior chamber and contained clear fluid, allowing slit lamp visualisation of the iris pigment epithelium posterior to the lesion (Fig

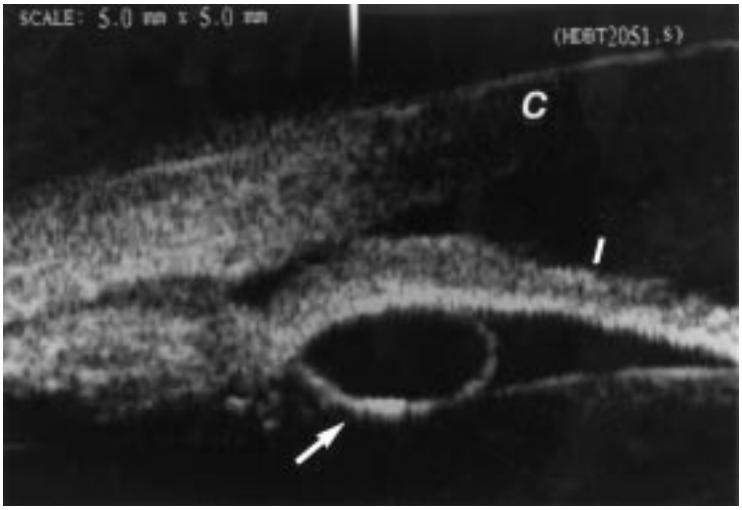

Figure 1 Peripheral iris pigment epithelial cyst. Ultrasound biomicroscopy, showing clear cyst (arrow) posterior to the iris (I). The cornea (C) is towards the top of the photograph.

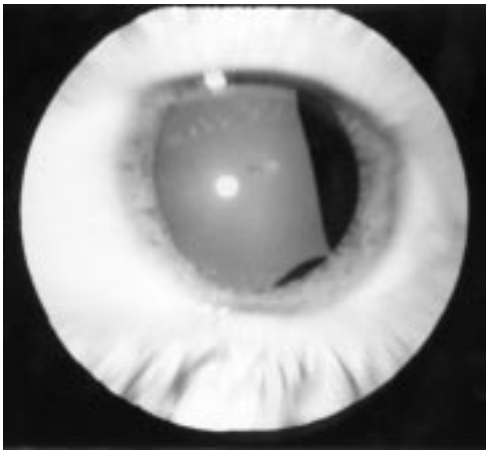

Figure 2 Mid-zonal iris pigment epithelial cyst. Note that there are two cysts, each of which has an elongated, fusiform shape.

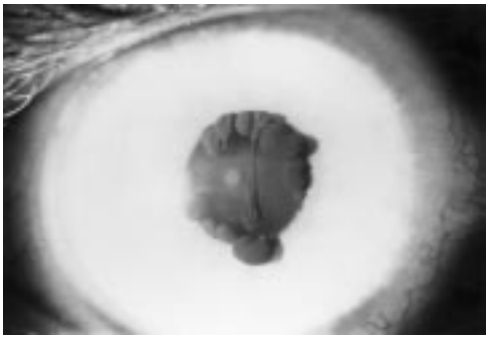

Figure 3 Pupillary iris pigment epithelial cyst. In this teenager, the lesions were bilateral and were noted shortly after birth. Note that some of the cysts are partly collapsed (iris flocculi).

Figure 4 Free floating iris pigment epithelial cyst in the anterior chamber in a 12 year old girl.

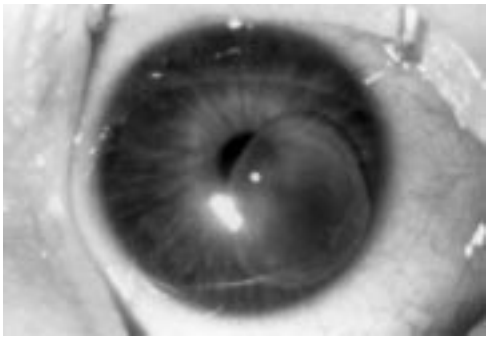

Figure 5 Iris stromal cyst located inferonasally in the right eye of an infant.

5). It was diagnosed in infancy and occurred in any quadrant. Eight of the nine cases showed slow progression after the initial diagnosis and covered a portion of the pupil, obstructing vision and requiring treatment. Two patients in the early part of the study were successfully managed by surgical excision alone by iridocyclectomy. Histopathology in each case demonstrated a cyst lined by non-keratinising epithelium similar to conjunctival epithelium (Fig 6). The remaining six children were managed by aspiration of the cyst and light cryotherapy at the limbal site of the collapsed cyst. There were no serious complications of this technique. This was done through the limbus with a 30 gauge needle with gentle suction of the cyst

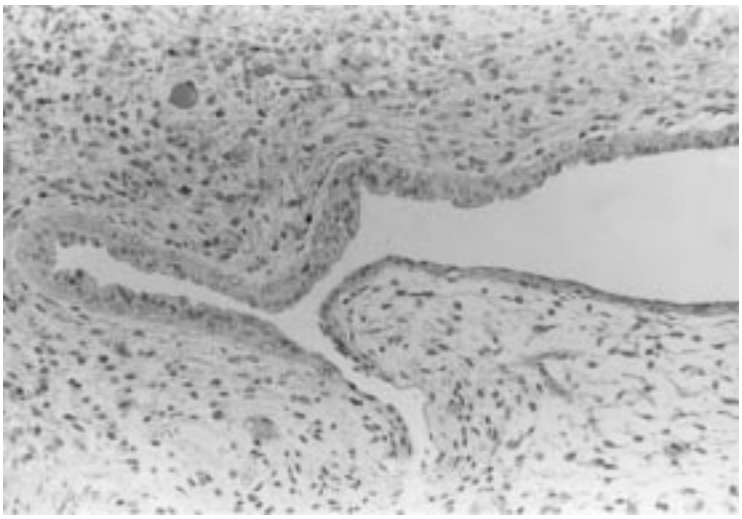

Figure 6 Histopathology of iris stromal cyst showing a thick wall and a lumen lined by non-keratinising epithelium similar to conjunctiva (haematoxylin and eosin, original magnification ×25).

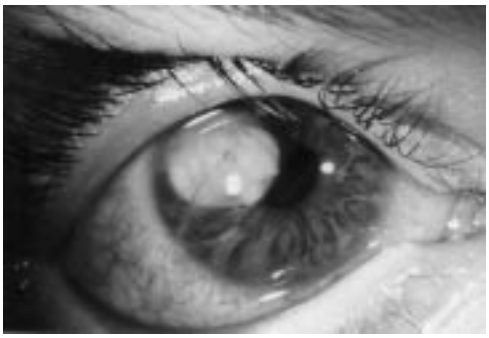

Figure 7 Epithelial downgrowth cyst secondary to perforating corneal trauma in an 11 year old girl. The progressively enlarging lesion was removed by a sector iridectomy.

fluid until the cyst was collapsed in the anterior chamber angle. The cryotherapy was then applied lightly to the limbus near the base of the collapsed cyst.

There were four secondary iris cysts in children. Two were of the epithelial downgrowth type and were managed by local excision (Fig 7). Two were tumour induced cysts, one secondary to a lacrimal gland choristoma of the iris[18] and the other secondary to a peripheral iris naevus.

Discussion

Although there have been a number of publications on iris cysts, to our knowledge there are no previously reported large series of iris cysts of childhood. It was somewhat surprising to find that of the 231 cases of primary iris cyst in our files, 53 (23%) were diagnosed in patients under age 20 years. We had previously believed that the iris stromal cyst, to be described shortly, represented the most common iris cyst of childhood. We generally consider the peripheral iris pigment epithelial cyst to be a disorder of adulthood, occurring most often in women between 20 and 40 years of age. In this study of childhood iris cysts, however, pigment epithelial cyst proved to be more common than iris stromal cyst. This may reflect referral bias, since iris pigment epithelial cysts are more likely to simulate melanoma and thus are more likely to be referred to our service.[19]

In this series, the 34 peripheral iris cysts accounted for 59% of all childhood iris cysts. However, most of these were diagnosed in teenagers with a mean age at diagnosis of 15 years. Like the adulthood lesion, they were more common in females, with 23 of the 34 cases (68%) occurring in girls. Although we have previously classified this cyst as an acquired lesion of adulthood, it is possible that some of them are congenital but are not detected in young children, who are less likely than adults to undergo detailed slit lamp biomicroscopy. It is almost always detected on routine slit lamp examination as an anterior bulging of the iris stroma nasally or temporally near the horizontal meridians. It usually prompts a referral to rule out iris or ciliary body melanoma. The office manoeuvres that help differentiate it from melanoma have been previously reported and include careful slit

lamp biomicroscopy with proper tilting of the light source, gonioscopy, ocular transillumination, and indirect ophthalmoscopy with scleral depression.[1 20] In cases where the clinical diagnosis remains uncertain, ultrasound biomicroscopy can be helpful confirming the cystic nature of the lesion (Fig 1).

The mid-zonal pigment epithelial cyst accounted for 9% of all childhood iris cysts. It is occasionally detected without pupillary dilatation but is best seen with the pupil dilated when it appears as a round or oval lesion that becomes increasingly more fusiform in shape upon pupillary dilatation. It is the cyst that is most likely to be confused clinically with a ciliary body melanoma.[1 21] The mid-zonal cyst is more likely to be multiple or bilateral. The presence of mid-zonal cyst in childhood suggests that this cyst may be congenital. The central or pupillary margin cyst is congenital since it is generally recognised in infancy. It is sometimes inherited in an autosomal dominant pattern.[1 22 23] Although there are generally no systemic manifestations, a recent association with familial aortic dissection has been reported.[23] It can be solitary but it is more characteristically bilateral and multiple, with lesions extending for 360 degrees around the pupillary margin. These lesions can be large enough to completely cover the pupil leaving only a small central pinhole that usually permits excellent central vision. Some cysts are collapsed and others are large and round (iris flocculi).[1] The two patients in this series were both referred during the teenage years for another opinion even though the parents had noted the lesions shortly after birth. In general, no treatment is indicated.

In some instances, an iris pigment epithelial cyst can become dislodged from its primary site and float freely in the vitreous or aqueous (Fig 4).[24] These lesions move freely with changes in position of the head or eyes and generally sink to a dependent position with gravity. Although these free floating cysts can occasionally become fixed, usually in the angle inferiorly, neither of the two in this series had become fixed.

The primary iris stromal cyst accounted for nine cases, or 16% of all childhood iris cysts in this series. It is commonly seen in infancy or early childhood as a solitary clear lesion of the iris stroma through which the iris pigment epithelium can be visualised at the posterior margin of the lesion.[1 4–17] Although it has been classified as a stromal cyst because of its anatomical location, it is actually lined by surface epithelium with goblet cells like conjunctival epithelium. Its pathogenesis is usually uncertain, although displacement of the surface epithelium into the eye at the time of amniocentesis has been incriminated in some cases.[16]

The stromal cyst is usually progressive and enlarges slowly to fill much of the anterior chamber and cover the pupil. Although the term "stromal cyst" has been traditionally used to describe this lesion because of its anatomical location, it is lined by surface epithelium compatible with conjunctiva, and the term "surface epithelium cyst" is possibly preferable. The best treatment is not yet determined and it is unclear whether aspiration, surgical removal, or laser treatment offer the best chance of control. We have generally employed the least invasive measure by performing an aspiration of the cyst allowing it to collapse into the angle and then to perform light cryotherapy to the overlying limbus in order to create an adhesion between the collapsed cyst and the overlying limbal tissue. This procedure can be repeated if the cyst recurs. The aspiration technique leaves the child with a round pupil and avoids a large corneoscleral incision and possible astigmatic amblyopia. If there are two or more recurrences using the aspiration technique, removal of the tumour by an iridocyclogoniectomy approach can be performed. Some authors have reported success with block excision techniques.[25–28] As an alternative to surgical removal, laser treatment to rupture the cyst, or piecemeal removal of the cysts by a vitrectomy technique can be attempted. We have not usually recommended these methods because they can cause a severe inflammatory reaction and can theoretically lead to a sheet of epithelial downgrowth in the anterior chamber.

A secondary iris cyst appears to be less common in children, accounting for only four cases in this series. Two were epithelial ingrowth cysts secondary to penetrating ocular trauma, both of which required surgical excision. The other two were tumour induced cysts, one of which arose from an ectopic lacrimal gland choristoma in the iris and was similar to the lacrimal gland cyst (dacryops) that occurs in the orbit.[18] Because of progressive enlargement and secondary glaucoma, this cyst may require surgical excision by iridectomy or iridocyclectomy. The second tumour induced cyst was a pigment epithelial cyst that arose immediately posterior to a peripheral iris naevus.[26] Another tumour that can produce an anterior chamber cysts is the medulloepithelioma of the ciliary body. Although we have evaluated and managed a number of cystic medulloepitheliomas,[29] none of them has presented primarily as a cystic lesion in the anterior chamber or iris and therefore medulloepithelioma was not included in this study. Parasitic cysts secondary to echinococcosis or cysticercosis are also known to occur in the anterior chamber, but they are extremely rare and were not encountered in this series.

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