

## Congenital cataracts presenting as a childhood squint

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### Abstract

**Aim:** A timely reminder that a small posterior subcapsular cataract could present with a squint.

**Methods:** A case series is reported of 4 patients who were referred to the paediatric ophthalmology service for management of a squint.

**Results:** In all 4 cases the strabismus was secondary to the undiagnosed posterior subcapsular cataract.

**Conclusion:** These cases emphasise the importance of carefully examining the red reflex in any child presenting with a squint. If the squint does not fit into the typical pattern of presentation, or a smudge appears to be present when performing retinoscopy or indirect ophthalmoscopy, a further assessment of the media using a direct ophthalmoscope or a slit lamp should be undertaken to check for a congenital cataract. A portable slit lamp can be very helpful in examining small children.

**Key words:** Cataract, Retinoscopy, Squint

We report on 4 children recently referred to our service who had presented with a squint, and on further assessment were found to have a unilateral posterior central cataract. All 4 children were male, 3 had the cataract in the left eye and 1 in the right, and their ages ranged from 6 months to 3 years at the time of diagnosis. Two of the children had an intermittent exotropia (XT), one an esotropia (ET) and the other a dissociated vertical deviation (DVD) of the eye. These cases emphasise that there must be a high index of suspicion when assessing children presenting with strabismus to exclude congenital cataract as an underlying cause for the onset of squint.

Congenital cataract presenting with a squint is not uncommon. In a study looking at how congenital cataracts were diagnosed it was found that 63% of the children presented with a squint.<sup>1</sup>

Although newborn screening is intended to detect congenital cataract, these discrete, posteriorly situated cataracts may be missed because of their small size.

These lens opacities are not identifiable to the naked eye and are usually detected as an abnormality in the red reflex, particularly when undertaking retinoscopy. Because of their position, posterior cataracts are more likely to have a negative impact on focusing and hence vision.<sup>2</sup>

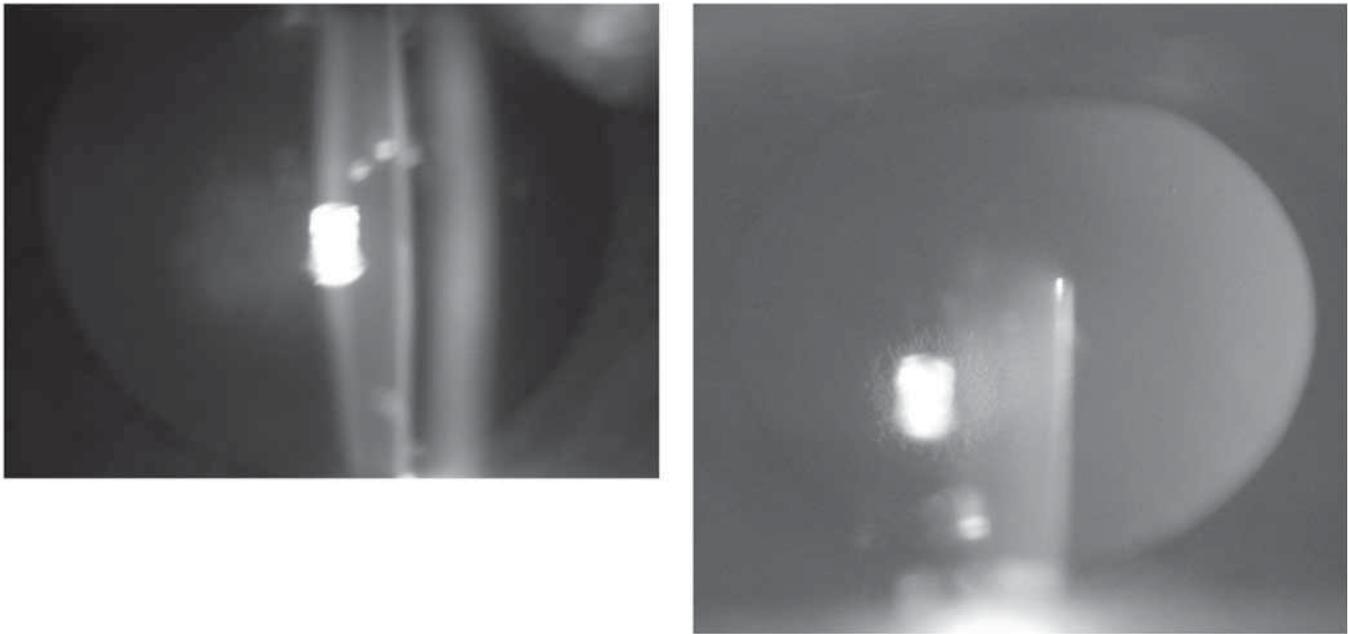
The diagnosis was made at our institution on the initial ophthalmic visit for 2 of the children; 1 of these children had been examined previously elsewhere and had come to our clinic for a second opinion. The other 2 children were referred for an opinion from colleagues once cataract had been identified. One child had their diagnosis made on the fifth attendance at a paediatric ophthalmology clinic and in the other child the lens opacity was identified on the fourth visit after an examination under anaesthesia (EUA) was performed.

Congenital cataracts occur about the rate of 248 cataracts per 734 000 deliveries (0.0003%).<sup>3</sup> They can be unilateral or bilateral, congenital or acquired, isolated or associated with a systemic condition.<sup>4</sup> Furthermore congenital cataracts can be small and discrete, yet visually significant because of their location. A central posteriorly located cataract tends to be visually significant as it is located at the nodal point of the eye and all images pass through this area.

Strabismus can present at any age; however, the common types of childhood squints usually follow a pattern. In retrospect, each of these cases had some atypical features which should alert the ophthalmologist to the possibility of an underlying pathology.

In the 2 cases of an intermittent XT, the squint had been present from birth, rather than the typical age of presentation of between the second and third year of life.<sup>5</sup> Isolated intermittent hypertropia is not a typical presentation of a childhood squint, although this can occur as a DVD or inferior oblique over-action in association with infantile ET; however, this child did not have any horizontal deviation or prior history of squint surgery.

The fourth case was a 3-year-old child with a sudden-onset esotropia. He was systemically well and fundoscopy was normal. The initial diagnosis was of VI nerve palsy, although at subsequent examinations he had a full range of eye movements. The child was sent for urgent investigation to a paediatrician, underwent blood testing and MRI under general anaesthetic. Perhaps if the



**Fig. 1.** The posterior cortical cataract that was detected in the child with an acute-onset esotropia.

child's congenital cataract had been detected at this initial assessment, the general anaesthetic could have been avoided.

None of the 4 children has required cataract surgery to date because of the small size of their lens opacity. They have been managed with glasses where required and patching to maximise their visual potential. Although no child has required squint surgery, the cataracts can progress, so these children need close monitoring. Each child was assessed by a paediatrician and no underlying systemic illness was identified.

These cases emphasise the importance of carefully examining the red reflex in any child presenting with a squint. All 4 children in this series were reported to have had a normal retinal examination and to have undergone retinoscopy at their initial consultation. Two of the children required a general anaesthetic (1 for the EUA, the other for the MRI), which although relatively safe does carry a small risk and is a significant cost to the health system. It is easy to overlook a small opacity when performing retinoscopy, which may be assumed to be a speck of dirt in the graticule. If the squint does not fit into the typical pattern of presentation a further

assessment of the media using a direct ophthalmoscope or a slit lamp should be undertaken to check for a congenital cataract. A portable slit lamp can be very helpful in examining small children.

The orthoptist and ophthalmologist examining the child presenting with squint should always consider the possibility of underlying pathology.

The authors declare they have no competing interests.

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