

Congenital glaucoma—a preventable cause of blindness

A few years ago two leading articles were written which encouraged paediatricians to detect and refer children with congenital cataract within the first days or weeks of life.^{1 2} These and other articles, together with an increasing awareness of the problem by paediatricians, has led to a significant decrease in the age at which treatment can be completed, so that it is within a critical but finite period of development. Congenital glaucoma is also important because it can be treated and because it is one of the major causes of preventable blindness in children. It too requires early treatment for a successful visual outcome but referral to appropriate centres is often late.

Congenital glaucoma is responsible for 4% of childhood blindness and in western Europe the incidence has been estimated at 1:12 500 live births.^{3 4} The risk is higher if the parents are related, suggesting an autosomal recessive inheritance. If there is no consanguinity, however, the risk to future siblings of a single affected child is only 1:20 and it seems more likely that inheritance is multifactorial.⁵

Congenital glaucoma is due to a developmental abnormality of the trabecular meshwork so that aqueous humour fails to drain from the eye. In young children this causes enlargement of the eye (buphthalmos), raised intraocular pressure, corneal oedema, and optic nerve damage. More than half the cases are due to an isolated defect of the trabecular meshwork whereas others are the result of various forms of maldevelopment of the anterior chamber of the eye. Seventy per cent of cases are bilateral. A wide variety of systemic diseases are associated with an increased incidence of congenital glaucoma including neurofibromatosis, homocystinuria, Marfan's syndrome, Lowe's syndrome, Reiger's syndrome, and congenital rubella infection. A number of children with chromosomal disorders have been described with congenital glaucoma but none of the associations are strong enough to give a clue to the genetic basis of the disease.

The most severe cases present with a dramatic picture at birth or within the first few hours or days of life with opaque corneas, photophobia, and blepharospasm. When less severe, the signs are more subtle, and the condition may not become apparent for weeks or months. During this time

raised intraocular pressure causes enlargement of the eye, corneal oedema, cupping of the optic disc, and the gradual development of visual field defects. When detected early, and when successful surgery is performed, optic disc cupping is reversible and the visual prognosis is good.^{6 7} The outlook is worse the larger and more opaque the cornea.

Most children with congenital glaucoma present within the first six months of life and corneal clouding is the usual presenting symptom.⁸ Buphthalmos, the 'ox eye' appearance so characteristic of congenital glaucoma, seldom itself worries parents as large eyes may be considered attractive. In severe or neglected cases the cornea becomes white but in the early stages all that may be noticed are hazy streaks which may have a very sudden onset. As the eye enlarges the cornea is stretched and Descemet's membrane splits and aqueous seeps into the cornea showing as linear streaks of corneal oedema that are visible to the naked eye. With oedema comes photophobia, blepharospasm, and watering.

In mild cases surgery by goniotomy is curative in 90% of cases with one or more operation. This procedure takes only a few minutes to perform and involves passing a blade through a small corneal incision and sweeping it around 180° of the anterior chamber angle. If the cornea is opaque, more complex surgery is required. Children with associated ocular developmental disorders and those requiring repeated surgery have a much poorer prognosis.

All children should ideally be screened for eye disease, not only in the nursery by a paediatrician, but again at 6 weeks, 8 months, 18 months, and 3 years.⁹ At these ages screening may be by the health visitor, general practitioner, or paediatrician. Examination should include holding an ophthalmoscope 18" from the cornea to look for opacities in the media. The red reflex should be seen and if it cannot, there is some opacity: corneal, lenticular, or vitreoretinal. If the reflex is hazy or absent, a check should be made to see if the iris details are clear. If not, the problem resides in the cornea and could represent corneal oedema from congenital glaucoma.

Any asymmetry of the size of the eyes should be considered suspicious of congenital glaucoma, although a difference of 1 mm is within normal

limits. Deciding whether the larger or smaller eye is abnormal can be difficult. Microphthalmos is commoner than buphthalmos and clues from the history such as photophobia and watering should be asked for. If in doubt, examination under anaesthetic with measurement of corneal diameters and intraocular pressure and fundoscopy may be necessary to make the diagnosis.

Photophobia sufficient to cause discomfort on a dull day or in artificial light is always a sinister symptom in childhood. If acute illnesses such as meningitis are excluded, ocular conditions such as injuries, albinism, retinoblastoma, corneal crystals, oedema, or dystrophies as well as congenital glaucoma should be considered as possible causes. Examination with a hand torch provokes a typical response with the child screwing up the eye and turning away as a tear falls onto the cheek.

Watering of the eyes without stickiness in infancy is unusual and merits careful consideration. Blockage of the nasolacrimal duct leads to a complaint of watering with stickiness. If the eye is not sticky a careful examination for other causes of tearing should be made so that the benign condition can be distinguished from the potentially blinding one.

The suspicion of glaucoma in a child should be

treated with urgency if serious visual handicap is to be avoided.

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