Strabismus affecting children with multiple handicaps

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In recent years there has been an increasing appreciation of the ocular defects which affect handicapped children, and their management has become an important aspect of paediatric ophthalmic practice. With the advent of more sophisticated neurosurgical techniques during the last 15 years, particularly for the control of hydrocephalus, and with the general improvement in neonatal intensive care, an increasing number of children, especially those affected by serious developmental disorders of the central nervous system, who would previously have died in infancy, are now surviving into later childhood and into adult life. A considerable proportion of these patients suffer from ophthalmic disorders which in their most severe forms may render them blind or partially-sighted. There is evidence from recent surveys in Great Britain (Fine, 1968) that the proportion of visually handicapped children of school age who have other significant handicaps is very considerable and is continuing to increase, a fact which poses difficult long-term educational and social problems. A high proportion of such children have some form of squint as one aspect of their ophthalmic disorder, and this applies also to those multiply handicapped children who do not have any other serious ocular defect.

It is my intention to concentrate in particular on two common and serious disorders which occur at birth or in the neonatal period, and which are a major cause of severe handicap, often both physical and mental. These are hydrocephalus, especially when associated with spina bifida, and cerebral palsy. Squints of various types and other ocular disorders are very common indeed in children affected by these disorders. A discussion of their aetiology, diagnosis, and treatment amply illustrates many points which are equally applicable to children who exhibit strabismus associated with other general disorders.

Cerebral palsy

This very variable condition has been defined as “a persistent but not unchanging disorder of movement and posture appearing in the early years of life, and due to a non-progressive disorder of the brain” (MacKeith, Mackenzie, and Polani, 1959). Progressive disease of the brain (such as that which occurs, for instance, in Tay-Sachs disease) is specifically excluded from the diagnosis, as is disease of the spinal cord. Although the spastic diplegia of Little is the classical form of the disorder, other patterns of motor handicap can be discerned which have led to various methods of classification. That of Balf and Ingram (1955) is particularly helpful (Table I). The aetiology of the condition is also very varied (Table II), and these differing aetiological factors contribute to the variation in the type of squint which may affect any particular child. The incidence of squint in cerebral palsy has been variously estimated as 60 per cent (Guibor, 1953), 55 per cent. (Griffiths and Smith, 1963), 48 per cent. (Breakey, 1955), 43 per cent. (Schachat, Wallace, Palmer, and Slater,
Table I  Classification of cerebral palsy (after Ingram, 1964)

Hemiplegia
Bilateral hemiplegia (quadriplegia)
Diplegia (spastic, atonic, etc.)
Ataxia
Dyskinesia (choreoid, athetoid, etc.)

Table II  Aetiology of cerebral palsy

**GENETIC FACTORS**  Indirect evidence mainly from family studies

**ABNORMAL INTRAUTERINE DEVELOPMENT**  Arrest of development with vascular occlusion sometimes sufficiently severe to lead to the development of porencephalic cysts; especially affects “small-for-dates” babies.

**BIRTH INJURY AND NEONATAL ASPHYXIA**  Especially related to prematurity, first deliveries, twin births, and often associated with the formation of sub-dural haematoma.

**ACQUIRED INFANTILE DISORDERS**
- Encephalitis (specific or non-specific)
- Trauma
- Acquired disorders of unknown aetiology

1957), and 37 per cent. (Douglas, 1961). All these figures are extremely high when compared with reported incidences of between 2 and 6 per cent. in the normal child population (Lyle and Bridgeman, 1959). The ratio of incomitant to concomitant squint is also much higher than in normal children; in Griffiths and Smith’s series, 72 per cent. of the cerebral palsied children examined who had evidence of squint exhibited some significant degree of incomitance, whereas the relative incidence in the normal child population is approximately 10 per cent. of all squints. In addition to squint there is a considerable incidence of nystagmus, of gaze palsies, and of chaotic unco-ordinated eye movements in cerebral palsied children.

Just as the state of cerebral palsy may be the end-result of many injurious influences acting during cerebral development, so any associated squint may also be attributed to a considerable number of possible factors (Table III, overleaf). If the squint is incomitant then this may be the result of damage to the oculomotor, trochlear, and abducent nerves or nuclei, either centrally or peripherally (trauma, for instance, may cause both nuclear palsy from cerebral damage, and peripheral cranial nerve injury). Duane’s syndrome has also been noted as occurring in some children affected by cerebral palsy (Douglas, 1961), and this is another possible cause of incomitant squint. Whether such a finding is coincidental, or whether cerebral damage is related to the anomalous neuronal firing patterns noted in this disorder, has not yet been determined. If the strabismus is concomitant, it may be only secondarily so, having originally been incomitant in very early life. Alternatively, it is primarily concomitant, in which case the main possible aetiological factors are poor vision affecting one or both eyes (sensory squint), poor fusion faculty, or significant refractive error.
Table III  Causes of squint associated with cerebral palsy

(A) INCOMITANT  
(1) Oculomotor, trochlear, and abducent nerve lesions  
   (a) Nuclear  
   (b) Intracerebral  
   (c) Extracerebral but intracranial  
   (d) Extracranial  
(2) Duane's syndrome

(B) CONCOMITANT  
(1) Primary concomitance  
   (a) Refractive error  
   (b) Absent or disturbed fusion faculty  
   (c) Visual sensory defect  
(2) Secondary concomitance  
   Affecting a squint of very early onset which was initially incomitant

Bearing all these factors in mind, it would seem likely that the incidence of squint would be greatest in those children most severely affected by cerebral palsy, and this is usually the case. Even if those children in which the squint might be the result of poor vision or of intellectual retardation are excluded, then the incidence of squint falls from 50 per cent. in cases of quadriplegia, to only 20 per cent. in cases of monoplegia (Douglas, 1961). The overall incidence of squint in cases of bilateral hemiplegia has been reported as 75 per cent., as against 43 per cent. in cases of diplegia, 27 per cent. in severe hemiplegia, and only 15 per cent. in mild hemiplegia (Ingram, 1964). Considering refractive error as a possible aetiological factor in the production of concomitant strabismus, it is interesting that Gardiner (1963) noted a very high incidence, at the age of 10 years, of hypermetropia in children with athetoid cerebral palsy (50 per cent.), but not in those with hemiplegia (15 per cent.), although this has not been statistically linked with any clear evidence of an increased incidence of accommodative convergent squint in the former group.

Hydrocephalus and spina bifida

In the British Isles spina bifida is the commonest major neurological congenital malformation compatible with life, and occurs in approximately 2.5/1,000 births (Nash, 1968). In a considerable proportion of cases an associated Arnold-Chiari malformation leads to the development of hydrocephalus, particularly after closure of a spinal meningomyelocele. Congenital hydrocephalus is also fairly common (1/1,000 live births) as an isolated defect (Butler and Alberman, 1969), and acquired hydrocephalus in infancy is a common consequence of intracranial trauma, infection, and neoplasia. Squint is a very frequent finding in these disorders, and decisions regarding its significance and management are therefore commonly encountered by the paediatric ophthalmic specialist. As in cases of cerebral palsy, the aetiology of squint in this present group of conditions is very varied, and is most easily considered under two headings:

(1) Cases in which the squint is the result of a primary developmental defect in the oculomotor nuclei associated with congenital hydrocephalus, either with or without spina bifida;
(2) Cases in which the squint is acquired as the result of post-natal factors which may or may not be related to the treatment of the basic neurological defect.

The congenital squints in such cases are at least initially incomitant, and usually alternating; convergent as the result of isolated abducent paresis, and less commonly mixed divergent and vertical from oculomotor or trochlear nerve palsies, the latter being much less common. After birth the progression of hydrocephalus may lead to traction and angulation of the oculomotor and optic nerves and to thinning of the visual cortex, as well as to pressure upon and distortion of the brainstem. As a result, acquired squints may arise, again most commonly from abducent palsies, but also from IIIrd nerve palsies, particularly from brainstem distortion; these may be accompanied by supranuclear vertical gaze palsy (the 'setting-sun' sign). Progression of the hydrocephalus, by leading to optic atrophy and to visual cortical thinning, may give rise to sensory incomitant squint from poor vision, or to squint from a lack of fusion faculty. When the hydrocephalus is the result of an acquired disorder, then any associated strabismus may be another direct effect of such a disorder, such as would be caused by meningitis, trauma, and intracranial tumour, on the oculomotor nuclei and nerves. Alternatively, it may be an indirect effect of damage to the retina, optic nerves, or visual cortex by the causative disorder (Table IV).

Table IV Causes of squint associated with hydrocephalus (unrelated to shunting procedures)

<table>
<thead>
<tr>
<th>Category</th>
<th>Cause</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital incomitant</td>
<td>Primary developmental (III, IV and VI nuclear dysplasia)</td>
</tr>
<tr>
<td>Acquired incomitant</td>
<td>Meningitis</td>
</tr>
<tr>
<td></td>
<td>Tumours and trauma (Localizing)</td>
</tr>
<tr>
<td></td>
<td>Raised intracranial pressure (Non-localizing)</td>
</tr>
<tr>
<td></td>
<td>Stretching and angulation of brain-stem and cranial nerves.</td>
</tr>
<tr>
<td>Acquired concomitant</td>
<td>Secondary concomitance of a primarily incomitant squint</td>
</tr>
</tbody>
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Visual Defect

(a) From optic atrophy
   (1) Associated developmental defect
   (2) Following optic nerve or chiasmal traction or compression
   (3) Following papilloedema
   (4) Associated with chorioretinal damage (as in toxoplasmosis)
(b) From cerebral blindness

Fusion faculty defect

? from occipital cortical deficit

The management of hydrocephalus in early childhood has been revolutionized since the introduction of artificial valve-controlled mechanisms for the drainage of cerebrospinal fluid from the lateral ventricles of the brain either into the right atrium of the heart or into the peritoneal cavity. These procedures have increased very considerably the chances of affected children surviving beyond infancy, and this applies both to those cases in which hydrocephalus is an isolated defect and to those in which it is associated with spina bifida. Especially in the latter group, however, the quality of the survivors is very poor in a
proportion of cases (Table V), and in most recent years the previous policy of universal

**Table V**  Quality of meningomyelocele survivors
(after Lorber, 1971)

<table>
<thead>
<tr>
<th>Handicap</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>No handicap</td>
<td>2 per cent.</td>
</tr>
<tr>
<td>Moderate handicap, I.Q. 80+</td>
<td>17 per cent.</td>
</tr>
<tr>
<td>Severe handicap, I.Q. 61-79</td>
<td>45 per cent.</td>
</tr>
<tr>
<td>Extreme handicap, I.Q. 60-</td>
<td>20 per cent.</td>
</tr>
</tbody>
</table>

perinatal closure of meningomyeloceles without careful assessment of the child’s general state, and in particular of the likely permanent degree of paraplegia, has been placed in doubt. For severe lower limb deformities, recurrent urinary tract infections with pyelonephrosis, and mental retardation permanently affect a considerable proportion of the children who survive their life-saving operations in the first few weeks of life (Lorber, 1971). Interestingly, there is a considerable incidence of squint in cases of meningomyelocele without any evidence of secondary hydrocephalus, and this is presumably on account of associated defects in brainstem development; Clements and Kaushal (1970) reported the presence of squint in 53 per cent. of a series of 27 such cases. However, squint is an even more common finding in cases of meningomyelocele combined with hydrocephalus, and particularly when there have been complications affecting the artificial valve drainage mechanism; Stanworth (1970) reported an incidence of 74 per cent. in such complicated cases. There are a number of reasons for this (Table VI), an understanding of which makes

**Table VI**  Causes of squint associated with hydrocephalus (related to shunting procedures)

- **Post-operative infection** (meningitis)
- **Incomitant (various types)**
- **Secondary rise in intracranial pressure from blocked valve**
  - **(A) EARLY** Rapidly increasing incomitant convergent (abducent palsy)
  - **(B) LATE** Concomitant due to blindness (afferent visual pathway damage or post-papilloedema optic atrophy)

it clear that the acquired development of a squint may be of great importance (Harcourt, 1970). If the valve becomes infected, a resultant meningitis may produce various patterns of squint directly or indirectly. If the valve becomes blocked, then there may be a consequent dramatic rise in intracranial pressure, producing most frequently a rapidly progressive alternating incomitant convergent squint as the result of pressure upon the abducent nerves. Less commonly, the rise in intracranial pressure may be great enough to produce a severe and often permanent loss of vision either by pressure and traction effects upon the optic nerves or the optic chiasm or the visual cortex, or by producing post-papilloedema optic atrophy. These changes are likely to be most severe when there has been a premature fusion of skull sutures after the previous introduction of an artificial drainage system which has lowered the intracranial pressure to abnormally low levels. In children who are paraplegic as the result of their spinal meningomyelocele, a secondary blockage of the drainage system may be initially misdiagnosed, because the symptoms of malaise, fever, and vomiting due to
acutely rising intracranial pressure may mimic those which of severe recurrent urinary tract infection (Harcourt, 1968). In these puzzling cases, a rapidly increasing angle of convergent squint, possibly associated with papilloedema (to which attention may then be drawn), can be the first unequivocal sign of a blocked valve. The absence of papilloedema in such cases does not, of course, rule out such a possibility.

Assessment

The assessment of visual function and the orthoptic examination of multiply handicapped children poses difficult problems. Many such children are intellectually retarded or emotionally disturbed to a degree which makes co-operation even with simple subjective tests of vision impossible at an age when they would normally be applicable. Emotionally immature children usually strongly resent even temporary occlusion of an eye, so that the differentiation between true paralysis and pseudo-palsy of the lateral rectus muscles in cases of convergent cross-fixation is made particularly difficult, although vestibular stimulation of the horizontally-acting recti by rotation of the child, or the elicitation of horizontal "doll's eye" movements by sudden rotation of the child's head may offer alternative methods of stimulating abduction movements when the crossed eyes appear initially to have no ability to move laterally beyond the primary position.

Many of the handicapped children examined because of squint have defective vision, most commonly as a result of optic atrophy, and here again assessment has usually to be simple and objective. A difference in the degree of resentment of uniocular occlusion applied to one as compared with the other eye is a good indication of uniocular visual defect, and it should be remembered that strabismic amblyopia may be superimposed upon a unilateral organic afferent visual pathway defect in some such cases. In assessing the visual acuity of those older children who are able to co-operate with such testing, the Sheridan Gardiner Stycar testing system is the most appropriate. Problems may then be encountered with some cerebral palsied children, especially those with severe involuntary athetoid movements of the arms, who may not be able to point with their finger to the matching letter on the reference card, even if they are able to see the test letter perfectly clearly. If large plastic reproductions of the test letters are laid on a table in a semi-circle before the child, then he may be able to use his head or a steadier lower limb, or his gaze, to indicate which he believes to be the test letter that is being presented to him.

Assessment of visual function should include examination of the fields of vision in addition to measurement of the acuity of central vision, and this applies especially to cerebral palsied children, in whom homonymous hemianopia is a common feature. Such a defect in visual fields may cause additional visual problems to an affected child, particularly during any period of occlusion therapy for amblyopia, and it should always be sought for on initial examination and subsequently kept under consideration. Tizard, Paine, and Crothers (1954) estimated that homonymous hemianopia affected 25 per cent. of children with infantile hemiplegia, being more common in acquired than in congenital cases. The defect is most easily tested for by placing the child on the mother's knee facing the examiner. While he attracts the child's attention the mother introduces her hand from behind the child's head first from one side and then from the other into the periphery of the field of vision. A homonymous field defect may be suspected if there is a repeated and consistent difference between the child's refixation onto the hand appearing from one side as against the other. The two-target method described by Hoyt (1963) is very useful in eliciting a dense homonymous defect in a very young child. As the subject fixes the first
target, a second target is silently introduced from one side. The child does not attempt to look at the new object of interest until it crosses the midline into the seeing half of the field of vision.

Because squint is particularly likely to be found in association with other visual system defects in multiply handicapped children, a very thorough ophthalmic examination is required. Particular attention should be paid to the pupillary light reflexes, the optic disc appearances, and other fundus features. Optic atrophy is difficult to diagnose with certainty in young children on account of the considerable physiological variation in the colour of normal optic discs which may occur in this age group. Any significant degree of pallor affecting one optic disc compared with the other, in the absence of a marked anisometropia, should strongly suggest that the nerve is atrophic, and such a unilateral optic atrophy may be the cause of a concomitant non-alternating “sensory” squint. Where the diagnosis is in doubt, the “pupillary-escape” phenomenon of Marcus Gunn, in which the pupil of the affected eye appears to dilate under the effect of direct light when the light is transferred rapidly from before the opposite eye, is often a helpful clinical test. These investigations are particularly relevant when occlusion therapy is planned in an attempt to overcome suspected strabismic amblyopia.

Aims of management

The basic aims of management of any squint, whether affecting a child or an adult, are always the same; that is, an appraisal of the possible cause of the squint, energetic treatment of any strabismic amblyopia which has developed, and attempted eradication of the ocular deviation in order to allow the development or restoration of bifoveal fixation where possible. In dealing with handicapped children there is, as already indicated, a particular importance in discerning the cause of any squint, especially the possibility of raised intracranial pressure. As such children’s squints commonly develop as the result of congenital ocular palsy, or of defects in the afferent visual pathways, or of defects in the potential fusion faculty, it is to be expected that the proportion in which treatment leads to the development of normal binocular function is very small. Nevertheless, this should remain the ideal against which all lesser aims are measured. First priorities are refraction under cycloplegia, which also allows a thorough examination of the media and fundi, a correction of any significant refractive error as soon as glasses can be tolerated, followed by treatment of any amblyopia. When young patients cannot be persuaded to wear an eye patch, then the long-term instillation of atropine drops to the good eye, possibly accompanied by an optical over-correction of 2 or 3 dioptres to the amblyopic eye to stimulate its use for near fixation (penalization for near), or associated with an under-correction of 5 dioptres to the normally fixing eye (total penalization), has been suggested as a useful alternative method of treatment (Catros and Garrec, 1972; Pouliquen, 1972). The use of prisms to produce an over-correction of the original deviation and a change to fixation of the previously non-fixing eye, possibly to be followed by binocular stimulation with after-images, has also recently been advocated for use with cerebral palsied children at special centres in conjunction with other forms of motor co-ordination readaptation (Pigassou, Le Moigne, and Cahuzac, 1972). On account of the associated problems of unco-ordinated eye movements, of nystagmus, and of organic ocular defects, in addition to those of general motor and intellectual and emotional dysfunction such as have already been stressed, little progress has been reported with these techniques, but they are clearly worth considering if they hold out any hope at all of allowing the development of normal binocular vision without
interfering with a child’s general developmental progress by the laying of too much stress on one particular aspect of his general readaptation.

In practice, and particularly when older handicapped children are being considered, the principal concern is whether or not cosmetic squint surgery should be advocated. There is now a widespread willingness to carry out such surgery, and I agree wholeheartedly with this provided that the adverse factors are also fully considered. The first of these is that the long-term results of such surgery tend to be unpredictable, particularly in the very young, and especially in cases of cerebral palsy. In some such cases severely unco-ordinated eye movements are associated with a very variable angle of squint, so that the exact amount of surgery which requires to be undertaken is difficult to assess. This is true also of the children who exhibit cross-fixation, owing to the difficulty, already mentioned, of distinguishing in the handicapped between true and apparent lateral rectus palsy in such cases. As the associated physical condition may not be a static one, there is also the danger that after surgery some further acquired ocular palsy may occur, or conversely that the original weakness may show further spontaneous improvement.

There are also the potential dangers of general anaesthesia in such children; the cerebral cortex may be very thin with a poor blood supply in some cases of hydrocephalus, and any prolonged hypoxia may lead to further permanent damage to the cerebral function. Secondary rises in intracranial pressure have also been reported after anaesthesia in hydrocephalic children whose condition had previously been well controlled by ventriculocisternal shunts, and it may be that this is related to some displacement or derangement of the valve itself, or to a fracture of the ventricular catheter due to a lack of gentleness and care in positioning the child’s head during and immediately after operation. Spina bifida children with severe kyphosis are difficult to intubate and they are also very liable to develop postoperative chest infections.

Finally, there is the problem of the recurrent hospitalization of multiply handicapped children to be considered. Lorber (1971) has given a distressing account of the total numbers of operations carried out on a series of children suffering from spina bifida during the first 10 years of their lives, and this amounts to an average of ten operations per child during the period. If to this is added the possibility of numerous additional admissions to hospital on account of urinary tract or chest infections, then any further admission for a cosmetic squint operation has to be weighed up very seriously. Nevertheless, there are many handicapped children in whom squint surgery can safely be undertaken without risk of producing physical or psychological ill-effects, and such surgery should never be denied solely on the grounds that the child is intellectually or severely physically retarded, bearing in mind the beneficial effect upon the anxious and unhappy parents of the cosmetic amelioration of one obvious aspect of their child’s deformity.

Discussion

FELLS What is the usual cause of the squint in toxoplasmosis?

HARCOURT All the cases I have seen had concomitant squints, which I think were most probably due to the defects in their afferent visual pathways from macular scarring and optic atrophy. It is possible, however, that some squints originate from extrinsic ocular muscle palsies and become secondarily concomitant.

FELLS What is the best time to operate for squint in hydrocephalic and other seriously handicapped children?
This is a matter for individual clinical judgement. It depends particularly on the individual child's general health, and on the standard of general anaesthesia which is available. For instance, children with spina bifida and hydrocephalus tend to be recurrently ill with urinary tract infections, and those with severe hydrocephalus and thinned cerebral cortex may have their residual cerebral cortical function at risk unless they are particularly well oxygenated under general anaesthesia. Although results can be unpredictable, I have not produced over-correction of convergent squint with true cross-fixation and limitation of abduction in more than a very few young hydrocephalic children, so that I am not dissuaded from operation by the risks of such a result, particularly as the postoperative cosmetic improvement is usually very striking and has a beneficial effect on the whole family. I am guided very much by the parents' wishes regarding the timing of such surgery, provided that I think that there is no particular medical contraindication.

I feel that the results are unpredictable and I do not operate unless I am pushed into it.

What are the functional results obtained in strabismic multiply-handicapped children?

This depends very much upon the severity of the handicap and the time of onset of the squint. Even minimally handicapped "clumsy" children have an increased incidence of squint and in such mild cases of handicap it is certainly possible to obtain a reasonable functional result. Severe multiple handicap, usually involving the central nervous system, tends to produce squint of very early onset from oculomotor nerve palsies, because of afferent visual pathway disorder and cerebral dysfunction affecting the binocular reflexes; the last factor is particularly important in cerebral palsy. Because of these adverse influences, a functional cure is very unlikely to occur, and this is particularly so in cases of cerebral palsy where there is often a widespread and bizarre derangement of the supranuclear control of ocular movements in addition to a paralytic strabismus of very early onset. More important than the cosmetic surgery is the detection and treatment of amblyopia which can certainly be successfully treated in early childhood provided the defective vision is indeed due to strabismic amblyopia and not to organic afferent visual pathway difficulties, which is unfortunately often the case.

What is the incidence of defects in vertical ocular movements in children with hydrocephalus?

I limited myself to the discussion of squint and not of gaze palsies, and I cannot give specific figures, particularly as there is more often a mixed vertical/horizontal deviation in residual strabismus which is difficult to interpret in terms of specific ocular muscle pareses. I certainly believe that modern very early surgical treatment of hydrocephalus by ventricular drainage has much reduced the incidence of the "setting-sun" sign, with loss of conjugate elevation and possible associated IIIrd and IVth nerve palsies, which was previously commonly seen in such severely affected children.