Personal practice

The management of squint

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Squint may be the first sign of a serious ocular or systemic disorder, and yet it is also one of the most common ocular conditions of childhood. It follows that while most ocular deviations have no such sinister connotations, an understanding of the range of associations is essential to be prepared for this comparatively infrequent but important possibility.

Screening for squint has been covered recently¹ and in this article its presence will be assumed. The clinical evaluation of the child with a squint will be considered first, followed by a discussion of the aims and methods of treatment. The main thrust will be directed towards the squint itself, but as amblyopia is such a frequent and important association this topic will also be covered briefly. In order to avoid extensive and tedious repetition the associations of squint will be listed using a predominantly system based approach. Subdivision into paralytic and non-paralytic types is only done at a later stage as both may occur in certain categories. Inevitably this classification has limitations and inaccuracies, but hopefully it will help the paediatrician to consider the possible implications of a squint. For further reference the reader may wish to consult two excellent standard texts.^{2 3}

Squint and its associations

SQUINT AS AN ISOLATED ANOMALY

Recently emphasis has been placed on early referral of infants and children with squint, but this has not taken into account that during the neonatal period such a deviation may be a normal event. During

early neonatal life about 32% of infants are divergent and only 3% convergent.⁴ As the incidence of divergence is higher in preterm neonates it is likely that during development the eyes are initially divergent but that this resolves over the first few weeks of life.

Apart from those seen in the neonatal period, squints are comparatively uncommon in the first few months of life, and the term 'infantile' is preferred to 'congenital' because this type of esotropia is rarely if ever congenital but develops between 3 and 6 months of age. Characteristically the deviation is large, constant, and there is no appreciable refractive error.⁵

Apart from the infantile type, the onset of esotropia in childhood is most common between 1½ and 3 years of age. The deviation may range from the hardly detectable (microtropia) to the cosmetically obvious manifest squint, and may be intermittent or constant. In some the deviation increases—or is only present—on accommodation (partially and fully accommodative squints respectively). Many, but by no means all, are associated with refractive errors.

Exotropia is less common than esotropia at all ages and has less propensity to cause amblyopia. In infancy ocular divergence should alert the clinician to the possibility of a severe visual impairment.

Paralytic squints do occur as an isolated anomaly in an otherwise healthy child, but a careful search for an underlying cause must always be undertaken (see acute central nervous system disease). Most, but not all, paralytic squints in children are congenital rather than acquired, although the role of birth trauma as a causal factor has probably been greatly overemphasised. A sixth cranial nerve palsy may present at birth but characteristically this resolves over the ensuing few weeks. Apart from this clinically obvious and rare exception, the most common palsy, of the fourth nerve, is usually not diagnosed for a year or so as the ocular deviation is difficult to elicit in infancy and the presenting sign is usually a compensatory head posture. Third nerve

In this article the following terms will be used interchangeably: squint and strabismus, esotropia and convergent squint, exotropia and divergent squint, comitant and non-paralytic squint, and incomitant and paralytic squint. Comitance will be assumed unless the terms incomitant or paralytic are used.

palsy is less common but more obvious, and in children is more likely to be congenital than acquired. A form of the latter that usually commences in childhood is ophthalmoplegic migraine. Benign recurrent sixth nerve palsy, thought to be a post-infective mononeuropathy as it usually follows a febrile illness, is included here because at presentation there are no associated systemic or neurological signs.⁶ In the superior oblique tendon sheath syndrome (Brown's syndrome) elevation of the eye in adduction is limited. The aetiology of this condition may differ between cases from a congenital anomaly of the tendon sheath to an acquired swelling of the tendon.

SQUINT AND OCULAR DISEASE

Squint may be the presenting sign of serious ocular disease. The eyes of a blind infant are commonly, but not invariably, divergent. Most important, squint either convergent or divergent may be the first sign of a treatable and potentially life threatening condition such as retinoblastoma. Thus squint can be associated with unilateral or bilateral ocular disease, including cataract and retinal disorders such as regressed retinopathy of prematurity (see below). Esotropia also develops in some children with congenital nystagmus.

SQUINT AND SYSTEMIC CONDITIONS

Many systemic conditions can be associated with strabismus and it would not be sensible or indeed possible to list them all here. Instead only a few will be considered and these will be grouped into broad categories.

Prematurity

The incidence of comitant squint is increased in infants born prematurely with figures quoted from 11% to 19%.⁷⁻⁹ The pathogenesis of this association is not well understood but is almost certainly due in part to both the perinatal neurological insults and retinopathy of prematurity to which these infants are susceptible.⁸ A high incidence of squint has also been reported in term and preterm infants who have had phototherapy for the treatment of hyperbilirubinaemia,¹⁰ but a causal relationship between these two and squint has not been defined particularly as many of these babies will also have suffered the aforementioned complications.

Disorders of the central nervous system

Acute diseases of the central nervous system—The association between squint and acute serious disease of the central nervous system is well known, and it must always be remembered that it may be the first sign of serious neurological disease. The deviation

may be unilateral, bilateral, and concomitant or paralytic, or both. Causes include raised intracranial pressure, intracranial tumours, trauma, degenerative and inflammatory diseases.^{6 12}

Neurodevelopmental disorders—In general, the incidence of ocular abnormalities in children with neurodevelopmental disorders including Down's syndrome and cerebral palsy ranges from about 40% to 90%.¹² Of these squint is the most common with an average incidence of about 40%,¹²⁻¹⁵ depending both on patient selection and the nature of the neurodevelopmental disorder. Although both comitant and incomitant deviations occur the former are far more common.

Brain stem dysfunction—Two conditions need to be considered in this category: Duane's and Möbius' syndromes. Duane's syndrome is relatively common and superficially resembles a sixth nerve palsy and its identification can obviate the need for extensive investigation. Often an isolated anomaly, it can be associated with other congenital abnormalities such as Goldenhar's syndrome, spina bifida, and deafness. Möbius' syndrome is either a developmental defect or an acquired hypoxic insult to the cranial nerve nuclei and is characterised by bilateral facial weakness, horizontal gaze palsies, and esotropia in some cases. Affected children are usually mentally retarded, deaf, and exhibit limb malformations.

Muscle disease

Uncommon in routine clinical practice and can easily therefore be misdiagnosed. Extraocular muscle involvement in myasthenia usually involves the levator muscle of the eyelid resulting in ptosis and may also affect any other extraocular muscle leading to diplopia. Characteristically myasthenic symptoms increase with fatigue and the signs can be unilateral or bilateral. Ocular motility may be affected in other muscle disorders and esotropia may occur in myotonia congenita. Ophthalmoplegia is seen in a number of muscle disorders including congenital and other myopathies, dystrophia myotonia and mitochondrial cytopathy. As in these last mentioned conditions the limitation of ocular movements is usually symmetrical, diplopia is not a common symptom. Rarely only the extraocular muscles are involved as in the congenital fibrosis syndrome.

ORBITAL CONDITIONS

In this group of conditions certain ocular movements are limited, due not to a neurogenic cause, but to mechanical restriction of the muscle movement. Examples include injury such as orbital blow

out fractures and infections. Squint may be the first sign of an orbital tumour such as rhabdomyosarcoma.

Investigation

Even now the evaluation of a squint, and its effect on the visual system, remains essentially clinical. The obvious priority is to identify first those conditions that require urgent treatment, and investigation should consequently be directed initially towards three aspects. First the type of squint, whether it is paralytic or non-paralytic, and either congenital or acquired; second, the identification of amblyopia; and third, the presence of associated pathology in the eye, orbit, or elsewhere. The extent of this spectrum has already been indicated and clearly the prevailing clinical conditions in an individual patient will dictate the relevance of each of these topics.

DETERMINING THE TYPE OF SQUINT

Differentiating the paralytic and non-paralytic squint is important and sometimes difficult. Exemplified by sudden onset squint a relatively common clinical dilemma in which determining the type (or even types) of squint is often difficult and can be at times impossible, particularly in the young fractious child.¹⁶ A sudden onset is suggestive, but not pathognomonic, of paralysis and can also occur with non-paralytic deviations, and on occasion both types may coexist.

It is pertinent to review the natural history of paralytic strabismus, which at its onset is characterised by a deviation greatest in the direction of action of the paretic muscle. Over the ensuing weeks and months there is tendency for 'spread of comitance' to occur, with the deviation spreading to all positions of gaze, so that eventually it becomes indistinguishable from a comitant squint and the paretic muscle cannot be identified. The following points may help in distinguishing a recent from an old paresis, or comitant squint. Diplopia is a feature of a recent paresis but not of other types. Amblyopia is present in longstanding but not recent deviations and past pointing is a feature only of the latter. A compensatory head posture is often present in both new and old pareses, but rarely in a primary comitant squint, and the former two can readily be differentiated by scrutinising old photographs.

HISTORY

Ocular history

Diplopia indicates a recent onset deviation, usually paralytic. If spontaneously volunteered by the patient it can usually be relied upon, but this symptom extracted by direct questioning must be treated with

caution as children can confuse physiological and pathological diplopia. The term diplopia is also sometimes used to describe blurred vision, as in the uncorrected myope. As children relatively rarely complain of diplopia, indirect evidence such as closing one eye when looking in a particular direction can be helpful. Often there is a disparity between history and findings concerning the following signs: squint, ptosis, compensatory head posture, proptosis, and pupil inequality. In this situation a selection of photographs can be invaluable in distinguishing old from fresh pathology and obviate the need for extensive investigations.

General history

Information on pregnancy, perinatal history, and general development are obviously important, but for the readers of this article need not be stressed. Nevertheless as squint may be the first sign of a serious neurological or ocular abnormality, the history must take these possibilities into account.

EXAMINATION

Ophthalmic examination

Measurement of visual acuity is the first step, but this remains the Achilles heel of paediatric ophthalmology. Even at the age of 3½ years accurate measurement is often not possible using standard tests.¹⁷ Preferential looking based techniques are opening important new horizons in clinical vision assessment, but as yet caution must be expressed regarding their accuracy in amblyopia measurement in the very young.¹⁸ Therefore in this population, evaluation remains qualitative, relying on such observations as objection to the occlusion of one eye.

Look for any facial, orbital, eyelid, or ocular asymmetry. Surprisingly when this is present determining which side is abnormal may not be simple. Thus a mother's comment that the right eye is smaller could denote just that. It could also indicate the presence of a right ptosis, right enophthalmos, or left proptosis, all of which could, by different mechanisms, be associated with strabismus.

Assessment of the squint and ocular motility, including the cover test, is mandatory. Ocular movements must be tested in all positions of gaze, and while many paralytic squints are obvious, during the recovery phase limitation of movement may be almost undetectable. In this instance further tests (for example, Hess chart) of overaction of the synergistic muscle of the other eye can be invaluable.

No ophthalmic examination is complete without assessment of the pupillary reactions, visual fields (when indicated) and fundoscopy. As many squints are associated with an abnormal refractive status,

except where urgent clinical problems preclude, mydriatic refraction is an essential part of the assessment.

Systemic examination

Most squints occur as an isolated anomaly; however, the possibility of coexistent or causal systemic pathology must not be forgotten, and a detailed paediatric assessment undertaken if indicated.

ADDITIONAL INVESTIGATIONS

Having already indicated the range of conditions that can be associated with strabismus, the need for additional tests such as a Hess chart, tensilon test, muscle biopsy, or computed tomogram is dictated by the prevailing clinical conditions and will not be considered here.

Treatment

The aim of treatment is to restore binocular single vision. This single aim encompasses: rectifying and refractive error, the treatment of amblyopia, and finally correction of the ocular deviation. As in almost all young children with strabismus there is a risk of amblyopia, treatment is directed initially towards prevention or correction of this aspect, and then to the management of the deviation. Although complete restoration of normal visual functions is often not possible, significant and worthwhile improvement can usually be achieved.

TREATMENT OF AMBLYOPIA

Not all types of amblyopia behave similarly and here only general guidelines are included. The principle of all modalities of amblyopia treatment is to promote the use of the amblyopic eye by compromising the vision of the better eye. This is usually achieved by occlusion, but other commonly used methods include penalisation and the CAM stimulator. Treatment should commence at as early an age as possible, but a prerequisite to all amblyopia treatment is a clear retinal image, and consequently accurate optical correction is essential using either spectacles or contact lenses.

Occlusion of the better eye, by an adhesive occluder, is the most commonly used method of amblyopia treatment and this can be worn full time or part time. Full time occlusion requires careful monitoring as there is a danger of transient amblyopia developing in the initially non-amblyopic eye. Part time occlusion is generally preferred for durations of 30 minutes to three hours or more. One study that compared conventional (more than three hours a day) and minimal (30 minutes) occlusion, and the CAM stimulator (see below) found all three

methods equally effective, although there was no improvement in 23%.¹⁹ If no improvement was achieved with one method it was worth trying one of the alternatives.¹⁹ If the amblyopia is refractory to treatment, as a last resort a short period of intensive occlusion on an inpatient basis may be undertaken. Occlusion is continued until either the visual acuity is restored to normal, or failing this, remains stable for at least three months. After cessation of treatment vision may deteriorate again in about 50% of cases,²⁰ and further periods of occlusion may be required. If amblyopia is to recur this usually occurs soon after the completion of treatment, but can be much later at about 12 to 13 years of age. Compliance is a common problem not surprising as occlusion is unpleasant particularly when the amblyopia is dense. Reported success rates vary: 77%,¹⁹ 92%,²¹ or 100%.²⁰ Flynn and Cassady reported success in only 56%, but their series included patients with form deprivation amblyopia, a particularly difficult group to treat.²² Despite these varying success rates occlusion remains the mainstay and most effective form of treatment.

Two other types of amblyopia treatment in current use are penalisation and the CAM visual stimulator. In penalisation the vision of the better eye is blurred with either atropine or glasses, or a combination of these two. It can be used in amblyopia in the following situations: as the first line of treatment (not common), as an alternative when occlusion has failed, for maintenance of vision after successful occlusion, for recurrent amblyopia, and for children who refuse to wear an occlusive patch for cosmetic reasons.^{2 3} The CAM visual stimulator consists of a rotating high contrast grating with a transparent cover over which the child draws. Each treatment session lasts only a few minutes. Although improvement does occur this cannot be attributed to the gratings per se, but to other factors, such as minimal occlusion of the better eye during treatment.²³

TREATMENT OF SQUINT

As already mentioned the aim of squint treatment is the restoration of binocular function. Unfortunately, particularly in comitant deviations, this is often not possible, and the cynic could easily regard treatment as entirely cosmetic. Of course cosmesis is an important goal in itself but in some paralytic deviations normal binocular function can be restored, while in other types of squint useful but subnormal binocular function can be achieved. In this section the deviation will be assumed to be comitant unless otherwise specified, and because of the large range of conditions within this range treatment will only be discussed in broad terms.

Despite the knowledge that most squints are without any serious systemic or ocular overtones, this possibility must not be forgotten, and before correction of the ocular deviation is considered, the child should be adequately investigated.

Nonsurgical treatment

Optical—Refractive errors and squint are commonly associated, and for this reason every strabismic child should have a meticulous mydriatic refraction. Spectacles are prescribed for two distinct reasons: first to correct a visually important refractive error and to provide a clear retinal image—essential before embarking on amblyopia treatment. Second, to ensure reasonable balance between accommodation and convergence. In this latter situation spectacles are prescribed to rectify the ocular misalignment and not to correct a visual deficit. Certain deviations, such as fully accommodative esotropia, can be adequately controlled by spectacles, while for others the result is less satisfactory and surgery may then be indicated. The effect of refractive correction depends on the age of the patient and squint type, but even so is often difficult to predict and it is common practice to correct any 'significant' refractive error as an initial step before embarking on surgery. Prisms are rarely used in paediatric practice except perhaps temporarily to control diplopia in a paralytic squint.

Surgical treatment

Surgery to align the eyes is considered for the following reasons: to cure diplopia, correct a compensatory head posture, or most commonly to correct a squint either not associated with a refractive error, or when spectacles have not had the desired effect. General comments only will be made.

Paralytic squint—Whether the palsy is congenital or acquired, treatment is essentially surgical (having first corrected any amblyopia) aiming to correct either a compensatory head posture or diplopia. For an acquired palsy at least six months of recorded stability is mandatory before surgery is contemplated.

Comitant squint—Any significant refractive error is corrected first, even in the presence of normal visual acuities. The adage, amblyopia correction before surgical intervention, generally holds true and every parent must understand that surgical alignment will not improve visual acuity. Surgery for infantile esotropia is generally undertaken between 6 months and 2 years of age as correction after this time achieves less in terms of binocular function. Surgery

for cosmetic reasons can be performed at any age although the needs of an individual child will dictate the timing of this.

A group which requires particular attention is that with neurodevelopmental problems. As with all cases of strabismus the appropriate refractive correction should be considered first otherwise the result after surgical intervention can be disappointing.²⁴ If surgery is undertaken the amount of extraocular muscle surgery necessary to achieve the same result needs to be less in the retarded compared with the normal child. In the past there has been a general reluctance to consider cosmetic surgery for these children, but the results can be rewarding for the child, parent, and others in daily contact.¹² Under these circumstances it is not the role of the surgeon to advise surgery, but simply ensure that parents understand the various therapeutic options, their benefits and limitations.

Treatment of special types of squint—The management of other types such as Duane's syndrome, superior oblique tendon sheath syndrome, and those due to muscle or orbital disease, cannot be considered in detail here. Some of these conditions produce no symptoms or little cosmetic defect (for example, most cases of Duane's syndrome) and clearly do not require treatment, while for others surgery may be considered to reduce or correct the strabismus or head posture, or increase the field of binocular single vision.

Botulinum toxin

This neurotoxin produces a temporary paralysis when injected into muscle and has been used as an alternative to extraocular muscle surgery over the past eight years. It is particularly effective in palsies, but has also been used in comitant deviations. To date its use in children is limited,^{25 26} but in the future it may well find an important niche in the management of paediatric strabismus.

Conclusion

While in most children squint is an isolated abnormality, it can also be the consequence of, or associated with, a large variety of other conditions. Clinical evaluation is not always straightforward and it is imperative that when there is the possibility of additional pathology the appropriate multidisciplinary assessment is adopted. In general, treatment is directed first to the correction of amblyopia and secondarily to the deviation itself.

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