

THE ROYAL COLLEGE OF
OPHTHALMOLOGISTS

GUIDELINES FOR THE MANAGEMENT OF
STRABISMNS AND AMBLYOPIA
IN CHILDHOOD

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Summary

Many professionals contribute to the management of children with squint and amblyopia including school nurses, health visitors, general practitioners, community physicians, orthoptists, optometrists and others in a variety of settings. It is therefore desirable to have locally agreed arrangements for the referral, assessment, treatment and monitoring of progress of children identified with relevant eye conditions, to which all parties contribute and which can be matched to national standards as these become established.

Guidelines are intended to be general principles, rather than specific protocols, regarding the best management of patients with a particular disorder. In drawing up these guidelines we wish to inform ophthalmologists and allied professionals as to the current view of best practice endorsed by the College. This is to enable the development of locally based protocols for the care of children with strabismus and amblyopia .

It is assumed throughout this document that those professionals dealing with common and uncommon cases of strabismus and amblyopia will have had adequate training and experience to manage children with these conditions.

1. Introduction

Strabismus (syn.squint) and amblyopia are common conditions in childhood, with strabismus affecting about 5% of five year olds of whom 60% have eso-deviations and 20% exo-deviations.⁽¹⁾

Amblyopia has an estimated prevalence in childhood of 1.2% to 4.4% depending on the defining criteria.^(2,3)

Strabismus or amblyopia may lead to failure to develop binocular vision which may prevent an individual pursuing certain occupations. The associated cosmetic disorder may interfere with social and psychological development with potentially serious effects for patients young and old.

Severe amblyopia persisting in adulthood is a significant risk factor for blindness in the case of an individual losing sight in the fellow eye.^(4,5)

Timely diagnosis and appropriate treatment of children with strabismus and/or amblyopia is likely to reduce the prevalence of persistent amblyopia and ocular misalignment in adults.

Rarely, strabismus and/or amblyopia may be the presenting symptom in children with a serious eye disease or systemic condition (e.g. retinoblastoma or hydrocephalus) when urgent referral to a specialist may be necessary.

2. Aims of management

- To maintain or restore optimal vision in both eyes
- To maintain or restore normal binocular vision
- To detect serious ocular pathology or neurological disease
- To achieve cosmetically satisfactory alignment of the eyes
- To correct significant abnormal (compensatory) head posture

The diagnostic aims may be achieved in a single consultation while some of the therapeutic aims may need to be pursued through childhood to 'visual maturity' e.g. by regular orthoptic assessment to the age of eight.

The use of the term 'cosmetic' in this context refers to the effect of improving the alignment of the eyes from an abnormal to a normal position. If achieved, this may provide both functional and psychological benefits.^(6,7)

3. Amblyopia

3.1 Definition

A deficit of vision, principally visual acuity, due to interruption of normal visual development during the sensitive period in childhood.

3.2 Types of amblyopia

3.2.1 Stimulus Deprivation Amblyopia

Stimulus deprivation amblyopia should be suspected in an infant or young child with eye disease which interferes with the eye's ability to form a focused retinal image. This may be a unilateral or a bilateral condition. Many eye conditions, such as corneal ulcer, glaucoma, ocular trauma or eye surgery in a young child may lead to stimulus deprivation and, if suspected, this should be specifically excluded on examination.⁽⁸⁾

In many cases of unilateral or asymmetrical 'organic' eye disease (e.g. retina or optic nerve lesion or partial cataract) there is an additional visual acuity deficit due to amblyopia which can only be ascertained retrospectively following successful amblyopia treatment.⁽⁹⁾

3.2.2 Strabismic Amblyopia

Strabismic amblyopia is suspected when a child shows either constant unilateral squint (without alternation of fixation) or a fixation defect with one eye. This may take the form of eccentric fixation, unsteady central fixation or fixation which on cover test is not maintained on removing the cover from the fellow eye.

3.2.3 Anisometropic amblyopia

Anisometropic amblyopia occurs when an interocular difference in spherical or cylindrical refractive error exceeds certain limits. In spherical anisometropia a minimum difference of 1.25 DS may be significant.^(10,11,12)

Unilateral high myopia may also cause amblyopia and bilateral amblyopia may result from high degrees of uncorrected hypermetropia such as occur in aphakia.

3.3 Presentation and referral

Strabismic amblyopia is detected by the use of suitable tests performed on children who have presented with manifest squint (see below). Anisometropic amblyopia is usually discovered when a child presents having failed a screening test of visual acuity.

Amblyopia may be of mixed aetiology ; e.g. children with anisometropic amblyopia may present with strabismus and unilateral cataract may lead to secondary squint , in which case the amblyopia is likely to be severe.

The minimum criterion for diagnosing amblyopia on a test of visual acuity is accepted to be two lines difference between the eyes on the linear Snellen test. This is usually equivalent to a difference of one octave in spatial frequency resolution. On repeated testing it may be possible to detect amblyopia in an eye with visual acuity of 6/9 when the fellow eye sees 6/6 (i.e. one line difference).

3.4 Methods of examination

In preverbal children with squint, the observation of abnormal fixation behaviour (including an aversion to monocular occlusion) is the basis for diagnosing amblyopia.

This is often a difficult observation to confirm. The demonstration of a spontaneous shift of fixation from one eye to the other (alternation) under normal circumstances excludes amblyopia. In large angle esotropia with crossed fixation, alternation may be found when the fixation target passes the mid-line.

If on cover test the suspected amblyopic eye holds central fixation steadily through a blink after removing the cover from the fellow eye, significant amblyopia is thus excluded. Examination of fixation using vertical prisms is useful in diagnosing amblyopia in the absence of strabismus.^(13,14)

3.4.1 Visual Acuity Measurement

As children mature they are capable of more demanding tests of vision. This has the unfortunate effect of corrupting the analysis of serial measurements. As a result of the crowding phenomenon single optotype acuity tests underestimate the depth of amblyopia to an unpredictable extent.^(15,16) The standard test is a linear optotype test (e.g. Snellen) with a normal degree of crowding. Ideally, a log MAR chart provides a more uniform progression of difficulty which is most suitable for comparative studies of visual acuity.⁽¹⁷⁾

It is necessary to detect significant visual acuity deficits and, in the case of unilateral amblyopia, to measure interocular differences in acuity.

Visual acuity should always be measured with the appropriate optical correction in place. In general, one should use the most demanding test for the child's ability. It may be helpful to test the better eye first (if this can be ascertained) to encourage younger children to perform the test, while the worse eye should be tested first in older children since some may memorise the test letters.

A range of tests of visual acuity should be available and the most appropriate test employed for a particular child (see appendix).

3.5 Management of amblyopia

Amblyopia is a treatable condition in childhood, i.e. during a 'sensitive' period and the limits of this period are still being defined. Improvements in acuity in strabismic amblyopia have been reported in children up to nine years of age, but in most cases the age of eight is taken as the onset of visual maturity.⁽¹⁸⁾

Amblyopia is treated by preventing the use of the better eye and enforcing the use of the amblyopic eye for substantial periods of time. Occlusion of the normal eye with an adhesive patch is the mainstay of treatment in all forms of unilateral amblyopia. In anisometropic amblyopia it may be required if the vision fails to improve despite the provision of suitable optical correction.

The duration and intensity of occlusion therapy will depend upon factors such as:

- Age at onset of amblyopia
- Age at presentation
- Severity of the acuity deficit
- Initial response to treatment
- Compliance with prescribed treatment

In the case of stimulus deprivation in infancy appropriate treatment to prevent amblyopia may consist of surgical and optical intervention e.g. removal of cataract and fitting of a contact lens plus patching of the normal eye for extended periods.

The most important prognostic factor is compliance with treatment. In smaller prospective studies, when compliance is closely monitored, success rates are high and significantly better

with good compliance. In larger retrospective studies success rates are lower except when selection criteria tend to exclude poor compliance.^(19,20,21)

Compliance therefore needs to be accurately assessed and optimised.

A wide variety of protocols of occlusion has been described for use in various situations and none is universally applicable. If one should be found ineffective others may be tried. In the case of severe unilateral stimulus deprivation amblyopia, the presence of nystagmus and/or secondary squint may indicate a hopeless prognosis and intervention e.g. cataract may not be recommended, depending on the known duration of the disorder.

As an example of a treatment regimen, Scott advocates full-time occlusion in the first instance for a period of one week per year of life (up to a maximum of four weeks) before re-assessment. If no improvement has occurred following three consecutive age-related periods of treatment, then 50% occlusion of the preferred eye is prescribed. If improvement occurs to parity on fixation assessment or visual acuity test then maintenance occlusion of 50% daily wearing is instituted, with gradual reduction in this percentage.⁽²⁰⁾

An alternative approach, advocated by Watson et al is to use minimal occlusion therapy of as little as 20 minutes per day combined with active use of the amblyopic eye in a visually demanding game. We feel this method is better suited to maintenance of acuity gained by more energetic treatment and in the older cooperative child.⁽²²⁾

Another successful method of treating amblyopia is penalisation of the non-amblyopic eye using optical defocus. This may be achieved by the use of topical atropine with or without defocusing lenses. Penalisation has advantages, which include the relatively easy application and prolonged effect of atropine as compared to occlusion and the possible benefit of maintaining low spatial frequency stimulation of the penalised eye. On the other hand, the impact of cycloplegia alone may be insufficient to cause a switch of fixation to the amblyopic eye and atropine may cause toxic and allergic reactions.⁽²³⁻²⁶⁾

Occasionally, amblyopia treatment causes a reversal of amblyopia to affect the occluded eye while the amblyopic becomes the fixing eye with a good visual prognosis. This is more likely to occur in younger children (< 2 years of age).

The response to treatment of amblyopia is quicker in younger children.⁽²⁷⁻²⁹⁾

Although treatment may safely be discontinued when serial assessments exclude the persistence of amblyopia, it is necessary to monitor children in this situation up to the age of visual maturity and many will require 'maintenance' treatment at times.⁽³⁰⁾

Success in the treatment of amblyopia is highly dependent upon compliance. As the parent or guardian is usually the principal therapist, they will want to know the proposed treatment, its duration and likely outcome. It is therefore essential that the nature of amblyopia and its impact on the child's present and future vision is fully explained. It is then necessary to outline and agree a treatment plan with the parent(s). This is a joint responsibility of the ophthalmologist and orthoptist who should collaborate closely in advising and supervising treatment.

If attempts at treatment of amblyopia achieve no improvement after substantial efforts by all parties, it is then wise to agree to discontinue treatment after full discussion with the parents. The potential benefits of treatment must be considered in the context of the particular child and the family.

4. Refraction and spectacle prescription

About 6% of one year olds have a significant refractive error.⁽³¹⁾ Hypermetropia and anisometropia greatly increase the risk of developing amblyopia and strabismus.^(32,33) Accurate refraction and appropriate prescription for ametropia are therefore essential in the management of strabismus.

4.1 Cycloplegia and retinoscopy

Accurate refraction in children usually requires full cycloplegia. Adequate cycloplegia for retinoscopy may be obtained 20 to 30 minutes following the instillation of cyclopentolate 1% eye drops. This is better tolerated if a topical anaesthetic such as proxymetacaine (0.5%) is also used. Below the age of three months mydriatics are used in lower concentration to reduce the risk of toxicity

The routine use of atropine for diagnostic cycloplegia or mydriasis is unnecessary and may cause harmful side-effects. However, in patients with darkly pigmented irides cyclopentolate may prove insufficient for full cycloplegia and it may be necessary to use atropine eye drops or ointment. This will achieve cycloplegia after 90 minutes, and so may be suitable for use prior to an appointment.

Retinoscopy is carried out in a semi-darkened room using hand-held lenses to neutralise fundus reflections along the visual axis. It is important to maintain the child's attention for fixation and it should not be necessary to use any restraint.

Prior to cycloplegia it is useful to examine the pupils in cases of constant unilateral squint and following mydriasis it is necessary to examine the fundi with direct and indirect ophthalmoscopy in all cases to exclude pathology (e.g. optic disc hypoplasia).

This exercise should be repeated in the case of failed amblyopia therapy.

It is rarely necessary to perform an examination under anaesthesia in order to carry out refraction and fundus examination and its routine use should be discouraged. If general anaesthesia is to be employed for another purpose, then this may offer an opportunity to examine the eyes more fully.

Regular refraction is the rule in children with amblyopia, squint or high refractive errors especially when amblyopia persists despite apparently adequate treatment.

4.2 Correction of Refractive Errors

The management of refractive errors in children with squint and/or amblyopia requires a team approach and is best carried out under the supervision of a consultant ophthalmologist. The prescription of spectacles for children with uncomplicated ametropia is the responsibility of ophthalmologists or optometrists in practice, subject to local agreement.

Children's spectacles should always be provided with plastic lenses to reduce the risk of injury.

4.2.1 Hypermetropia

In all forms of esotropia, full correction of hypermetropia is the treatment of choice. In practice, a reasonable lower limit for spectacle correction is + 1.50 dioptres (+ 3.00 ret. @ 2/3 metre). When prescribing, 'full correction' means that only the working distance is allowed for with no subtraction for cycloplegia.

In children without strabismus the precise indication for treatment of spherical errors is ill defined and will depend on the age of the child and the magnitude of the error.

For instance, in infants with hypermetropia, emmetropisation may occur naturally and this should be monitored.

In bilateral balanced hypermetropia, without strabismus, some refractive correction is advisable for errors greater than about + 4.00D even in the presence of normal uncorrected visual acuity, since this may prevent asthenopia when the demands of school increase.⁽³⁴⁾

In convergence excess esotropia bifocals may be considered. High-top executive bifocals are prescribed aiming to fully correct the near deviation and allow fusion with the least addition needed up to + 3.00 DS add. Bifocals are not suitable if only partial reduction in the squint angle is obtained. Once binocular fusion has become established on this treatment, gradual weaning is carried out to avoid long-term dependence.

4.2.2 Anisometropia and astigmatism

Anisometropia and astigmatism are potent causes of amblyopia in childhood which may be missed in younger children in the absence of squint. Hypermetropic anisometropia appears more likely to cause amblyopia than anisomyopia.

The need to correct the refractive error will depend on its magnitude and the age of the child. For instance, anisometropia of greater than 4.00 D is likely to need correction at any age, whereas correction of 1.50 to 2.00 D may only be desirable in children of school age. When amblyopia is found, prescribe for spherical or cylindrical anisometropia of more than 1.00 D. If spectacles are prescribed for hypermetropia and/or correction of squint, one may prescribe to correct any amount of anisometropia.

In balanced (symmetrical) astigmatism without squint in children less than four years old, serial refraction may reveal 'normal' emmetropisation with time.⁽³⁵⁾ Spectacles are not usually required but the child should be reviewed to confirm that the astigmatism is no longer significant.

4.2.3 Myopia

High myopia (-6.00 D or more) may require correction in infancy and moderate myopia (-4.00 D or more) in two year olds and older children. Lesser degrees of myopia do not usually cause problems in small children and prescription can be based on subjective refraction over the age of six years.

5. Strabismus (Syn: squint)

5.1 Definition

Strabismus is a misalignment of the eyes in which the visual axes deviate from bifoveal fixation.

5.2 Classification

The classification of strabismus may be based on a number of features including the relative position of the eyes, whether the deviation is latent or manifest, intermittent or constant, concomitant or otherwise and according to the age of onset and the relevance of any associated refractive error. The type of strabismus is established by a detailed history and orthoptic examination.

- Infantile esotropia (syn: congenital or essential esotropia) is an idiopathic syndrome in which an esodeviation is present before the age of six months. It is variably associated with other clinical features including dissociated vertical deviation, inferior oblique overaction, latent nystagmus, crossed fixation, asymmetrical monocular optokinetic responses (OKN) and, usually, no refractive error.⁽³⁶⁾
- Acquired strabismus includes fully and partially accommodative refractive esotropia, convergence excess esotropia, cyclic esotropia, occlusion esotropia and various forms of parietic squint.⁽³⁷⁾
- Exotropia may also occur in congenital and acquired forms, both concomitant and incomitant.
- Vertical strabismus includes dissociated deviations, cyclovertical muscle anomalies and restrictive conditions (e.g. Brown's syndrome) as well as rarities such as double elevator palsy.

These broad categories of strabismus are distinguished by having various aetiologies and usually differ in prognosis with and without treatment.

5.3 Presentation and referral

Intermittent deviation of the eyes is a quite common finding in healthy neonates and should not cause undue concern. Normal binocular coordination becomes evident at about three months and strabismus after this age is significant.

Constant squint is generally recognised early by the family, health visitor or general practitioner. A positive family history of squint or amblyopia should alert those in primary care when carrying out routine checks or immunisations.⁽³⁹⁾

Strabismus is often found in association with neurological disease such as in cerebral palsy and in craniofacial developmental anomalies.

Strabismus, amblyopia and refractive error are much more common in children with treated or regressed retinopathy of prematurity (ROP).⁽⁴⁰⁾ Premature infants with a history of stage III ROP or worse should be followed up after the neonatal period to screen for these complications.^(41,42)

If squint or amblyopia is suspected in the primary care setting, it is appropriate for local protocols to provide for direct referral to an optometrist or an orthoptist to exclude refractive error and strabismus. If no abnormality is detected, such patients may be discharged. Cases with intermittent or constant manifest squint should be referred to an ophthalmologist without delay. In all children referred with strabismus or amblyopia the possibility must be considered that this is the presenting feature of a serious ophthalmic or systemic disease requiring urgent management.

5.4 Strabismus management

5.4.1 Infantile esotropia

There are five broad considerations in planning management.

5.4.1.1 Development of binocular vision

Children with untreated infantile esotropia, when assessed at school age, will commonly show equal visual acuity and a dense alternating suppression with no form of demonstrable binocular cooperation. Early surgery is advocated on the basis that the primary defect is a motor one and alignment of the eyes before some critical age might permit the development of binocular function.⁽⁴³⁾ This is the most controversial issue in the management of the condition and has the greatest influence on the timing of any surgery. As yet, there is not enough evidence to decide the matter and contrasting policies are followed by different surgeons. To illustrate the variety of results that have been obtained, three studies have been selected:

- (i) In a widely cited study that stimulated further work on early surgery, 93% of 106 children operated upon and successfully aligned before the age of two years, had Worth 4-dot fusion or gross stereopsis. Of the children aligned after this age, 31% demonstrated a similar outcome.⁽⁴⁴⁾
- (ii) In a series of 358 patients who had undergone surgery for infantile esotropia, 20% were orthotropic with fusional amplitudes and normal retinal correspondence. None had stereopsis with TNO test and only three had low-grade stereopsis on the Titmus test. The probability of achieving subnormal binocular vision appeared to decrease with increasing age at surgery and was consistent with the view that surgery before the age of two produces better results but these may still be obtained in patients at a later age.⁽⁴⁵⁾
- (iii) In a recent prospective study of 98 out of 118 patients who underwent surgery for infantile esotropia, and who had remained aligned to within 8 PD of straight five years later, one third had stereopsis on the Titmus test (range 200-3000 seconds of arc). Thus 22% of the original group had obtained some form of binocular function and 68% had remained well aligned. No patient with a divergent squint of any degree demonstrated stereopsis.⁽⁴⁶⁾

In attempts to improve results, other studies have aimed for alignment by twelve or even six months of age, but no better stereopsis has been achieved.⁽⁴⁷⁾

The most common outcome of successful surgery is the monofixation syndrome with subnormal binocular vision. The surgical target in infantile esotropia is, therefore, usually within 10 PD of straight, this being the maximum angle at which monofixation is possible.

The advantages cited for various forms of subnormal binocular vision over complete suppression are: simultaneous binocular perception, fusional vergence, intact binocular field, normal distance judgement and, sometimes, gross stereopsis.^(48,49) An advantage to later surgery is a lower risk of subsequent amblyopia.

As an alternative to surgery, botulinum toxin injection into the medial recti has been reported but is not at present in general use.⁽⁵⁰⁾

5.4.1.2 Correction of amblyopia

While many children with infantile esotropia demonstrate balanced alternating fixation, amblyopia may occur. In untreated squint this is reported as between 13% and 33%, rising to 20-80% after surgery.^(51,52) It is therefore important to monitor infants following squint surgery and to treat any amblyopia detected.

Provided that suitable orthoptic supervision is carried out, it is not necessary to delay surgery until completion of amblyopia therapy.

5.4.1.3 General health issues

Systemic disorders which increase the risks of anaesthesia should be regarded as a relative contraindication to early surgery. The angle and direction of squint in infants with cerebral palsy and other neurological disorders is often unstable. In such patients surgery for presumed infantile esotropia may be better deferred at least until two years of age.⁽⁴⁹⁾ However, in a prospective study of surgery for essential esotropia, the outcome was no worse in the neurologically impaired or premature infants.⁽⁵³⁾

5.4.1.4 Surgical treatment

The definition of satisfactory cosmesis and the optimum age for surgery in a given case are a matter for discussion between the parents, orthoptist and surgeon. As regards the type of surgery, published evidence suggests that bi-medial rectus recession is the most effective procedure, perhaps combined with simultaneous resection of one lateral rectus for large angle squint.⁽⁵⁴⁻⁵⁶⁾

The type and amount of surgery to perform for a particular squint is a decision for the experienced surgeon. Parents need to be advised that, whilst accuracy in measuring and operating upon strabismus is essential, the response to surgery is variable and cannot be guaranteed. It is good practice to agree the objectives and discuss the actions necessary if the desired surgical outcome is not achieved.

5.4.1.5 Correction of associated features

Correction of overacting inferior oblique muscles found in association with 'V' pattern strabismus may be required on cosmetic grounds, including a compensatory abnormal head posture and, if marked, is usually carried out at the same time as the esotropia surgery. If binocular function is present after surgery, persistent inferior oblique overaction may disrupt it.

In summary, there is no series of cases reported in which successful alignment in infantile esotropia has allowed the development of high grade stereopsis associated with bi-foveal fixation (40 seconds of arc or better). If the eyes are aligned to within 10 PD of orthotropia, up to one third of patients develop subnormal binocular vision. There is evidence to suggest that this binocular vision provides functional advantages. However, there are no accurate means of predicting pre-operatively which patients will enjoy this outcome.

5.4.2 Acquired strabismus in early childhood

5.4.2.1 General principles

Most of the preceding recommendations in infantile strabismus management also apply in acquired strabismus. The important differences are:

- An assumed history of possibly normal binocular vision prior to the onset of squint.
- A greater likelihood that optical treatment alone will be required.
- The related risk of loss of binocular vision if treatment for the squint is delayed.

In view of these factors, treatment should aim to restore ocular alignment and binocular vision as soon as possible. It is therefore necessary to consider whether a given case of childhood strabismus has a chance of a good functional result following therapy on the basis of the history, with particular regard, e.g. to age, and findings such as the presence and severity of any associated amblyopia and / or suppression. It may also be necessary to inform general practitioners that delay in the referral of young children with strabismus serves no useful purpose. Information regarding the mode (i.e. constant or intermittent) and time of onset of a squint is helpful in assigning appropriate urgency to appointments.

5.4.2.2 Management sequence

It is important to measure and fully correct significant refractive error before planning any surgical correction of strabismus. It is also desirable to have corrected any amblyopia present.

5.4.2.3 Pre-operative prism adaptation in acquired strabismus

There is good evidence to show an improved predictability and outcome of surgery in acquired esotropia following adaptation using Fresnel prisms on spectacles. Briefly, the method is used to discover patients with fusion potential and may disclose a larger angle of squint than that first measured. Surgery carried out on this larger angle has a greater chance of success without an increased risk of over-correction and prism-responders so treated are less likely to require re-operation. This technique is recommended where practicable, particularly if measurements of squint angle are variable.⁽⁵⁷⁾

5.4.3 Exotropia

Exotropia may be constant or intermittent and may present as a primary condition or be consecutive (following esotropia) or secondary to unilateral visual loss.

Constant primary exotropia is much less common in this country than esotropia. It is thought to be more commonly associated with other developmental abnormalities. The deviation is usually large with alternating fixation and a low risk of amblyopia and the squint is present on near and distance fixation even when accommodation is stimulated.

In contrast, intermittent exotropia, which may begin in infancy, is noted when one eye drifts outward at times, particularly in bright conditions, on distance fixation and when the patient is tired or unwell. When the deviation is manifest there may be suppression or diplopia, typically overcome by closing one eye. Intermittent exotropia may be found to measure the same angle at near and distance. More commonly, the eyes are straight at near and divergent in distant and far distant fixation. In convergence weakness, the angle is larger on near fixation. Children who have straight eyes on near testing demonstrate good stereopsis (60 seconds of arc or better) when old enough to perform detailed tests.

Treatment aims are generally the same as for esotropia, namely eradication of amblyopia, restoration of fusion where possible and re-alignment where necessary to achieve satisfactory function and appearance.

Orthoptic treatment is useful in improving control of residual intermittent exotropia in children with good fusion who are old enough to learn how to be aware of the deviation of one eye. Training is then aimed at improving fusional amplitudes.

5.4.3.1 Surgery in childhood exotropia

The question of optimal timing of surgery in intermittent exotropia is not settled. It is appropriate to consider a variable plan according to the age of the child. Indications for surgical intervention include increasing frequency of manifest deviation with symptoms and deteriorating binocular function as demonstrated by serial orthoptic assessment.⁽⁵⁸⁻⁶⁰⁾

Various types of horizontal muscle surgery are effective in treating childhood exotropia. These include unilateral and bilateral lateral rectus (LR) recessions and LR recess/ MR resect procedures.⁽⁶¹⁾ In basic exotropia, recess/resect surgery appears more effective, whereas in simulated distance exotropia, bilateral lateral rectus recession has a higher success rate.⁽⁶²⁾

In exotropia with convergence insufficiency, a lateral rectus recession and relatively large ipsilateral medial rectus strengthening procedure may also reduce the difference between distant and near angle.⁽⁶³⁾

In young children, Pratt-Johnson recommends an amount of surgery to fully correct the exotropic angle measured in distant fixation. A small esophoria is the ideal immediate post-operative state. In children over the age of three, the aim of surgery is to produce a small initial over-correction of the deviation (up to 10 PD) measured in far distant fixation, this being likely to lessen with time but to reduce the frequency of recurrence of exotropia.⁽⁶⁴⁾

Significant esotropia persisting after one week requires attention to prevent amblyopia and suppression. Alternate occlusion may be used or prisms fitted to spectacles to establish and maintain binocular fusion. Once this is achieved, further surgery may be necessary to improve the ocular alignment and remove the need for prisms.

5.4.4 'A' and 'V' Patterns in Horizontal Deviations

If an exotropia increases on upward gaze, or an esotropia on downward gaze, a 'V' pattern is said to exist. Similarly, an 'A' or 'X' pattern may be found. 'V' pattern horizontal deviations are usually found in association with overaction of the inferior oblique muscles. Assessment of these patterns on prism/cover testing should be made with the child wearing full refractive correction and on distance fixation with either elevation or depression of the chin.

Surgery to weaken overacting inferior oblique muscles in significant 'V' pattern deviations may improve abnormal head posture and expand the field of binocular fusion. In patients with 'V' exotropia without fusion the same procedure may improve alignment on downward gaze.

'A' pattern deviations may be due to overacting superior oblique muscles and may be corrected by superior oblique weakening procedures. 'A' and 'V' patterns may also be treated by vertical displacement of the insertions of the horizontal recti if there is no evidence of oblique muscle dysfunction.

5.4.5 Vertical strabismus

The two most common causes of vertical strabismus in childhood are superior oblique underactions and dissociated vertical deviations.

Superior oblique weakness may be due to paresis or to maldevelopment of the muscle tendon.⁽⁶⁵⁾ Typically, a hypertropia in the affected eye will be associated with a compensatory head tilt to the opposite side with chin depression and overaction of the ipsilateral inferior oblique. The head posture develops as soon as the infant gains head control when upright. It may not be noticed by the family and photographs are useful evidence. There is usually evidence of fusion in the presence of the head posture in primary gaze. Fusion is prevented if the hyperdeviation or cyclodeviation is large.

The aim of surgery is to allow normal binocular fusion in primary gaze and on looking down without abnormal head posture.

Surgery to weaken an overacting inferior oblique muscle may be achieved by myectomy, recession or disinsertion.

Surgery to 'strengthen' the superior muscle by tucking the tendon on the temporal side of the superior rectus is appropriate if it is found to be lax and if it is normally inserted in the globe.

Dissociated vertical deviation (DVD) typically develops later on in children who have previously undergone treatment for large angle infantile esotropia. Correction of DVD may be required if a marked cosmetic defect is present. Suitable surgical procedures include posterior fixation sutures or large recessions of the superior rectus or in the case of co-existing inferior oblique overaction, disinsertion and anterior transposition of this muscle.^(66,67)

These forms of surgery are usually the province of the strabismus specialist. As with more common operations for squint, it is necessary to discuss the rationale for a particular operation and the possible untoward effects which may occur such as post-operative Brown's syndrome after shortening of the superior oblique tendon or masked bilateral superior oblique weakness following unilateral surgery.⁽⁶⁸⁾

5.4.6 Neurological disease

On rare occasions, a child with acquired strabismus or amblyopia may be found to have a primary neurological disorder such as optic glioma or medulloblastoma. This is more likely in the presence of features such as nystagmus, persisting amblyopia or deteriorating visual acuity. A careful examination should be performed to exclude an afferent pupil defect, papilloedema, optic atrophy or other cranial nerve disorder. The finding of any abnormal neurological signs should prompt referral to a paediatrician and consideration of the need for cranial imaging and electrophysiology.

6. Facilities

The appropriate facilities for children in hospital are defined in 'Ophthalmic Services for Children'.⁽⁶⁹⁾

Whether in a community clinic or in a hospital eye department there should be adequate provision of space, time and equipment to allow the clinician to properly examine the patient and provide any necessary treatment. Many factors influence the ease with which assessment in hospitals and clinics is achieved. These include comfortable surroundings in waiting and play areas for children and their attendants, minimal delay in seeing the clinician and a friendly, professional approach by staff to the parents and child.

The optometrist and orthoptist should have easy access to the ophthalmologist, ideally in adjacent accommodation or with the opportunity to jointly examine the child (i.e. concurrent clinics where possible).

It is important to be able to maintain the child's attention for examination, especially if accurate retinoscopy is to be achieved. It is helpful to have easy control of the lighting in the examination room to prevent distraction and to have access to a variety of toys and pictures to attract visual attention.

7. Communication

The treatment of children with strabismus involves a number of disciplines and may take place in a variety of locations and adequate communication between staff and patients and parents may be difficult to achieve.

Groups involved:

- Hospital and patient (+family)
- Medical staff and orthoptists and optometrists in hospitals
- Community-based orthoptists and medical officers
- Community paediatricians
- General practitioners and health visitors
- Allied services (e.g. teachers, school nurses, non-hospital optometrists)

Good communication between staff is essential in order to provide coherent advice to parents. Clear and detailed medical and orthoptic records should be kept and be mutually available when patients attend clinics and on admission for surgery. Letters should generally be sent to general practitioners on all new cases and whenever there is a change in the clinical condition or treatment of a patient. It is good practice to copy correspondence to the community paediatrician concerning children undergoing treatment for squint or amblyopia when other conditions such as developmental delay coexist.

We recommend the local provision of information sheets for parents explaining the nature of the conditions concerned and their treatment and expected outcomes in simple clear language. These should be available in out-patient departments and wards.

To aid communication in the treatment of amblyopia we recommend the development of record books held by parents, in which the serial prescriptions of e.g. occlusion therapy and a diary of treatment carried out and the visual acuity achieved are recorded .

Regular case discussions should be encouraged. Staff should be supported to attend relevant academic meetings and maintain appraisal of the literature.

8. Audit

There are many aspects of strabismus and amblyopia management which require audit from time to time in order to be sure of the quality and efficacy of care provided. Audit requires clear objectives and adequate resources in order to be carried out successfully.

In order to audit the results of treatment in strabismus, reference should be made to the aims of treatment which are:

- i) Optimum visual acuity in each eye
- ii) Optimum binocular function
- iii) Good cosmetic appearance

The first two aims may be quantified on examination, while the third requires questions to be asked of the patient and parents, although alignment of the eyes to within 10 prism dioptres of straight is usually compatible with an acceptable appearance and with peripheral fusion.

Suggested data to be assessed in audit are the following:

- i) Status**
 - a) at presentation
 - b) during and after amblyopia treatment
 - c) pre-operative
 - d) post-operative
 - e) on discharge
- ii) Patient variable**
 - a) age
 - b) visual acuity
 - c) refraction/prescription
 - d) diagnostic category
 - e) associated diagnoses
 - f) strabismus angle
 - g) surgery
 - i) target angle
 - ii) surgical dose
 - iii) technique
 - iv) complications
- iii) Event**
 - a) out-patient appointment
 - b) out-patient attendance
 - c) under-correction
 - d) over-correction
 - e) re-operation

Results of audit carried out locally should be compared to those published in relevant literature. National audit initiatives, when deemed appropriate, might be carried out under the auspices of the College.

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Over sixty fellow consultants and orthoptist colleagues contributed helpful information prior to final drafting.

12. Appendix

Tests of visual acuity

Age most suitable

Preferential-looking (PL) Gratings-based (Teller, Keeler)

3-12 months

Vanishing Optotypes (Cardiff)

12-30 months

Picture Matching (Kay, Elliott)

2-4 years

Single letter (Sheridan-Gardiner, Sonksen-Silver)

3-5 years

Linear Snellen and log MAR (Bailey-Lovie, Glasgow)

4 years and on

13. Expiry date

These guidelines will require revision in the light of new information.

Proposed expiry date: December 2004.

