The Royal College of Ophthalmologists

Guidelines for the Management of Strabismus in Childhood

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1. Overview

This guideline is designed for ophthalmologists managing children with strabismus (syn.squint), which is defined as a pathological misalignment of the visual axes. This is a broad subject and the reader is referred to comprehensive texts, for further information see bibliography (page 42).

The guidelines are intended to give general principles of management. It is assumed throughout this document that professionals dealing with common and uncommon cases of strabismus will have had adequate training and experience to manage children with these conditions. This document represents the current view of best practice endorsed by the College. Please also refer to the Royal College Quality standards document[1] and Ophthalmic services for children[2].

The management of strabismus in childhood is multidisciplinary matter and usually involves parents and children, ophthalmologists, orthoptists and optometrists. General practitioners, health visitors, paediatricians and paediatric neurologists may also become involved in the management of children with strabismus. It is desirable that these parties are committed to locally agreed care pathways, covering visual screening, referral, assessment, treatment and the monitoring of progress of children identified with strabismus. The latter is particularly important, as it is common that information will change with development, and multiple follow up visits will be required. Written information about strabismus should be available. Adequate time should be made available by the clinicians involved, in order to explain the terminology, the possible treatments, what they involve and for the consenting procedures that are required for surgical intervention.

2. Introduction

The classification of strabismus may be based on a number of features

Classification of Strabismus	
Intermittent	Constant
Infantile	Acquired
Accommodative	Non-accommodative
Comitant	Incomitant
Horizontal (eso/exo) – deviation	Cyclotorsional and vertical

Strabismus is a common condition in childhood affecting 2.1% of the population[3], with an increased prevalence associated with assisted delivery (forceps or caesarean section), low birth weight (including premature infants), neuro-developmental disorders. Neuro-developmental strabismus (associated with a neuro-developmental problem) is independently associated with maternal smoking later in pregnancy, maternal illnesses in pregnancy and low birth weight for gestational age. Approximately 60% have eso-deviations and 20% exo-deviations[4].

Strabismus may lead to a failure to develop binocular vision, and amblyopia, either of which may prevent an individual pursuing certain occupations. The appearance of ocular misalignment may interfere with social and psychological development with potentially serious effects for all patients with strabismus[5,6,7,8,9].

Timely diagnosis and appropriate treatment of children with strabismus can reduce the prevalence of amblyopia and ocular misalignment in later childhood and adult life. Correction of strabismus has been reported to improvement in motor coordination[10].

Strabismus may be the presenting symptom in children with a serious eye or brain condition (e.g. retinoblastoma, hydrocephalus or brain tumour). All professionals involved with the management of strabismus need to be able to recognise this, and either initiate onward referral or arrange for appropriate investigation and management.

3. Aims of management of Strabismus

The aim of strabismus management is to achieve good visual acuity in each eye, restore normal ocular alignment (as near as possible, which may be a small under or over correction) and maximise the potential for sensory cooperation between the two eyes (the development of binocular single vision, which includes 3D vision, or stereopsis). While normal binocular single vision is the goal, sub-normal levels may be useful and may prevent later recurrences.

Aims of management of Strabismus
To detect/exclude serious underlying ocular or neurological disease
To maintain or restore optimal visual acuity in each eye
To maintain or restore normal (or subnormal) binocular single vision
To restore appropriate ocular alignment
To eliminate double vision, or other induced symptoms (e.g. asthenopia)
To correct significant abnormal (compensatory) head posture
To improve binocular visual field (in the case of esotropia correction)

3.1 History

It is important to elicit a detailed medical history including details of the onset of symptoms and other visual problems or general health issues. Prematurity, birth history and developmental milestones should be noted. The onset of strabismus is generally before the age of 5 years[3,11,12]. Cases may be intermittent. A family history may be present.

Most cases of strabismus occur in children who are otherwise healthy and often appear asymptomatic. The presence of behaviour change or headache should be considered suspicious of serious neurological disease.

Parents and/or carers may report the presence of strabismus. However, small angle strabismus (as well as high refractive error and anisometropia causing amblyopia) may not be recognised unless the child is screened. Therefore screening of visual acuity at 4 years of age either by an orthoptist, or by an orthoptically managed service, with onward referral to an orthoptist as appropriate is recommended[1,2,13].

Strabismus is more common in children who have a positive family history and in those who have developmental ocular abnormalities or some specific systemic disorders. In such children a higher level of surveillance is recommended.

Strabismus: Associated Conditions
Family history of strabismus or amblyopia[14]
Prematurity[15,16,17,18]
Down's syndrome[3,19,20]
Developmental delay[3,20]
Craniofacial syndromes[21]
Fetal alcohol syndrome[22]
Unilateral ocular disease
Cerebral palsy[18,23]

3.2 General Examination

It is important to document any dysmorphic features and/or abnormal head posture before commencing examination of the eyes.

3.3 Visual function

The visual performance is measured in each eye (if possible). Many decisions depend on accurate and reproducible measurement of visual acuity (see table below). The standard measurement is visual acuity (or minimum resolvable), as opposed to minimum visible or minimum discriminable (also known as Vernier acuity or hyperacuity). Uniocular visual acuity should be assessed, using occlusion or occlusion glasses, where co-operation allows. It is important that the occlusion of each eye is complete to ensure that each eye is tested separately, therefore a patching each eye is preferable. It may be necessary to accept acuity with both eyes open if occlusion causes distress. Fixation pattern may be used as a gross comparison between the two eyes where formal testing is not possible. The two eyes can be separated using a vertically acting 10 or 12 dioptre prism (usually held base down). This is only necessary if there is no strabismus or the angle of strabismus is very small. A fogging lens (for example a plus lens) can be used instead of occlusion in the assessment of children with manifest or latent nystagmus.

In small children a variety of tests are available and the list (below) is a guide. Comparison to normal age adjusted examination findings is helpful for each test.

Vision Tests	**Age Guide**
Forced choice preferential looking grating	0-1 year
Cardiff cards	0-2 years
3 m logMAR (uncrowded) Kay pictures	18 months to 3 years
3 m logMAR crowded Kay pictures	2 – 4 years
3m logMAR crowded letters	Above 3 years

It is preferable to use a crowded test as early as possible for accurate detection of amblyopia.

Decisions based on visual acuity
Treatment with glasses
Amblyopia therapy (occlusion, penalisation)
Frequency of follow visits
Consideration for further investigations
Timing of strabismus surgery

It is also useful to document near visual acuity, particularly in school age children. Near chart letter matching may be a more appropriate measure of near acuity than reading text, if children are still learning to read. A comfortable size for reading is usually two sizes larger than threshold. In communications with parents and teachers, and documentation of the text size to be used is best given in terms of “font size” rather than either near Snellen or other near measurements that are commonly used in adults.

3.4 Binocular Vision

There are a number of age appropriate ways of assessing and measuring sensory binocular function in common practice.

Tests of Sensory Binocular Function		
	Near	Distance
Fusion	Worth 4 Dot	Worth 4 Dot
Fusion	Bagolini	Bagolini
Stereopsis	TNO	FD2 (Frisby Davis 2)[24]
Stereopsis	Frisby	
Stereopsis	Randot	
Stereopsis	Lang	
Fusion and stereopsis	Synoptophore	Synoptophore

The depth of suppression can be assessed using a Sbisa bar (or Bagolini Filter bar)[25]. Motor fusion can be assessed using the 20 base out test in preverbal children, and more formally in older children in a step wise manner using a prism bar, or smoothly using a Risley prism, at both near and distance fixation.

3.5 Ocular Alignment

Most childhood strabismus is concomitant. Incomitant deviations may occur with certain childhood conditions (e.g. Brown and Duane syndrome), but it is important to consider a cranial nerve palsy at all times, particularly if the eye movements are incomitant. Assessment of ocular alignment is done by carrying out a cover/uncover test and an alternate cover test for both distance and near targets, (for near this can be to a light initially, but an accommodative target must be used to increase accommodative stimulus). It is important to note if there is any fixation preference and the degree of control (if present). If appropriate, testing should be carried out with and without spectacles. Quantification of the deviation is done using a simultaneous prism cover test and an alternating prism cover test (APCT). These tests may be carried out in the nine positions of gaze where indicated. Alternatively, the deviation can be measured in the nine positions by using a synoptophore. The presence of “A” or “V” patterns and dissociated vertical deviation (DVD) should be documented (usually for distance fixation).

The accommodative convergence/accommodation ratio (AC/A) can be assessed where there is a large disparity between the distance and near angle. It is recommended that the gradient method is used at 6 meters.

The presence of any torsion can be assessed using a double Maddox rod, single Maddox rod, torsionometer, fundus examination or synoptophore.

3.6 Ocular Movements

The versions and ductions need to be assessed. In small children it may not be possible to bring the eyes into all areas adequately. Occlusion of an eye for a short period (up to an hour) or use of the vestibulo-ocular system may help in

demonstrating full abduction. Overacting and under acting muscles are documented. Convergence is tested using an accommodative target. In older children, the measurement can be facilitated by use of an RAF rule. If indicated jump and smooth convergence can be assessed individually. Saccadic and pursuit movements are occasionally of interest in neurological cases. The use of a diagram to document extra ocular movements is considered best practice.

Nystagmus may be present and needs to be carefully assessed and documented in the primary position, secondary and tertiary positions viewing distance targets. Manifest/latent (fusional maldevelopment syndrome) nystagmus occurs in association with strabismus and often co-exists with dissociated vertical deviation (DVD). In children with nystagmus particular attention should be given to the visual acuity with both eyes open, near visual acuity (with both eyes open), fusion and any compensatory head posture to aid surgical planning.

3.7 Ocular Examination

The pupils must be examined. An afferent defect is particularly important in cases of constant unilateral squint with reduced vision. Following mydriasis it is necessary to examine the ocular fundus. This includes the optic nerve and quality of the nerve fibre layer, macula and retinal periphery. The use of the indirect ophthalmoscope is recommended as standard in small children (less than 5 years), however in older children, the direct or 90 dioptre/slit lamp examination can add information. If restraint in required, this is permitted to allow fundus examination to exclude life threatening examination findings such as papilloedema or optic atrophy. Parents should be requested to consent to have their child restrained. If the examination is inadequate for any reason, the difficulty in the examination should be documented and a date set for a repeat examination.

3.8 Other Tests

In those children in whom there is visual loss with or without strabismus, an electroretinogram may provide further information. A Lees Screen is rarely used in children, but may document limited movement in cooperative older children with acquired motility abnormalities.

Assessment of control using controlled binocular acuity (formally binocular visual acuity) can be helpful.

Use of the fixation circle in an ophthalmoscope can identify foveal fixation, or lack of it, which is particularly helpful in microtropia.

3.9 Abnormal Neurology

On rare occasions, a child with acquired strabismus or reduced vision may be found to have a primary neurological disorder such as optic nerve glioma, medulloblastoma, craniopharyngioma or hydrocephalus. This is more likely in the presence of features such as persistently reduced visual acuity, resistant to amblyopia therapy, deteriorating visual acuity or an ocular muscle under action. A careful examination should be performed to exclude an afferent pupil defect, papilloedema, optic atrophy or other cranial nerve abnormality. The finding of any abnormal neurological signs should prompt referral to a paediatrician and for cranial

imaging to be considered. This would normally be Magnetic Resonance Imaging (MRI) unless computerised tomography (CT) was specifically thought to be beneficial.

Possible Indications for Neuro-imaging
Headaches
Cranial nerve palsies
Afferent pupil defect
Optic nerve pathology
Neurological abnormality
Unexplained visual loss
Sudden onset strabismus

3.10 Refraction

About 6% of one year olds have a significant refractive error. [26] Hypermetropia and anisometropia greatly increase the risk of developing amblyopia and strabismus. [27, 28] Accurate refraction and appropriate prescription for ametropia are therefore essential in the management of strabismus. It is currently accepted practice that children with hypermetropic refractive errors (up to +4.00) do not need glasses as long as 1) there is no strabismus 2) visual acuity and binocular function are developing in an age appropriate manner, 3) there is no significant anisometropia or astigmatism.

Accurate refraction in children under 12 years old usually requires full cycloplegia. A subjective refraction may be possible in older children once they can read the chart letters (from age 8). However cycloplegia is indicated even in this group in the presence of an esotropia. A post cycloplegic subjective refraction is a useful exercise in selected cases.

Adequate cycloplegia for retinoscopy may be obtained 30 minutes following the instillation of cyclopentolate 1% eye drops. This is better tolerated if a topical anaesthetic such as proxymetacaine (0.5%) is instilled beforehand. Below the age of six months mydriatics are used in lower concentration to reduce the risk of toxicity (cycloplentolate 0.5%). The routine use of atropine for diagnostic cycloplegia or mydriasis is generally considered unnecessary. However, in patients with darkly pigmented irides, cyclopentolate may prove insufficient for full cycloplegia. In these situations, it may be necessary to use atropine.

Retinoscopy is carried out in a semi-darkened room using hand-held lenses, or trial frame, to neutralise the retinoscopic reflections along the visual axis. It is important to maintain the child's attention and fixation should be on the retinoscope light. It should not be necessary to use any restraint, and if it is, it is unlikely the refraction will be accurate.

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.

Accommodation can be assessed by using the near visual acuity, dynamic retinoscopy (suggest Nott method[29], for accommodative lag,) and pull away method (for amplitude). Accommodative facility can be assessed using the child's ability to

overcome flippers (+/-2.00 reading a near target). Children need to be able to cooperate with this latter test, and a lower age of 8 years old is suggested.

A repeat refraction every 12 months is advised. If visual acuity fails to improve or deteriorates, it may be necessary to repeat the refraction, (along with a repeat fundus examination) or consider alternative diagnoses.

3.11 Management

A team approach is recommended, with the majority of follow up visits occurring with the orthoptist.

Intermittent deviation of the eyes is a quite common finding in healthy neonates and should not cause undue concern.[30] Normal binocular coordination becomes evident at about three months and any persistent strabismus, after this age, is significant.

In many cases, the management of strabismus in children commences with glasses. Children's spectacles should always be provided with plastic lenses to reduce the risk of injury. Advise careful fitting, using adequate support for the nasal bridge and sufficient size of lens to avoid children looking over the top of the glasses.

In all forms of esotropia, full correction of hypermetropia is the treatment of choice (having corrected the retinoscopy for working distance only). This is often known as the 'full correction'. There is no requirement to subtract any lens power for cycloplegia. A reasonable lower limit for glasses prescription is +1.50 diptre sphere. In exotropia, uncorrected hypermetropia might be preferable to aid exotropia control, assuming visual acuity is good.

The main aims of further review are to further diagnose and classify the strabismus, monitor visual development (visual acuity and binocular function), treat amblyopia and manage the strabismus to obtain, maintain or restore binocular single vision where potential is present (e.g. prisms, alteration of / or addition of lenses, exercises). A period of refractive adaption is recommended after glasses have been prescribed, until the vision is stable, as the visual acuity can improve with glasses alone, even in strabismic amblyopia. This may take up to 18 weeks. [31-36] It is necessary to monitor the visual acuity during this time, to know when to introduce further strategies (such as occlusion) to improve the visual acuity. The frequency of follow up visits will depend on many factors, such as age and change in treatment and treatment effect. Follow up visits are usually scheduled with the orthoptists, and usually occur initially 6 weeks after glasses have been prescribed, and can by expected to occur approximately between 4 and 10 times a year following this.

An annual refraction is required to monitor changes in the glasses prescription. An earlier repeat is indicated if the vision fails to improve in what would be an expected way (for example with compliant occlusion), the visual acuity is worse with the spectacles, or there is a small residual esotroipa. Accurate vision assessment dictates many of the management decisions in strabismus and, it is the view of this College, that childhood strabismus is best managed by the orthoptist, in conjunction with an ophthalmologist in all children less than 5 years of age. Annual refractions are carried out either by an optometrist working in partnership with the orthoptist, or an ophthalmologist.

The location of follow up visits does not have to be in a hospital eye department, as long as the location is set up in accordance with the standards and guidance as outlined in references 1, 2.

See appendix 2 (Tiffin personal communication) for one possible care pathway. The different professional groups carry out the following

Parent Carer	Case finding
Community optometrist	Case finding (less than 5 years of age) Spectacle dispensing (less than 5 years of age) Refractions
School Nurse	Case finding
Orthoptist school screening	Case finding
GP	Case finding Authorisation of referral
Health visitor	Case finding
Paediatricians	Case finding Management of associated paediatric conditions Overseeing investigations such as cranial imaging
Orthoptist	History Measurement/assessment binocular function Measurement of visual acuity Measurement of strabismus Assessment of eye movements Related tests (synoptophore, Lees screen) Diagnosis (in conjunction with refraction information) Management plan Amblyopia therapy Orthoptic exercises Continued follow up, monitoring change Consideration of surgical intervention
Hospital Optometrist	Cycloplegic refractions (yearly) Non cycloplegic refraction Contact lens management Low vision aids Fundus examination
Ophthalmologist	History Measurement/assessment binocular function Measurement of visual acuity Measurement of strabismus Assessment of eye movements Anterior segment examination Cycloplegic refraction Non cycloplegic refraction Fundus examination Co-morbidity Diagnosis Management plan Consideration of surgical intervention Perform surgical intervention

Further treatment of any residual strabismus that persists (despite the correct glasses and following amblyopia treatment) may be indicated to improve appearance

and increase the potential for binocular development. Treatment is usually surgical although there may be reasons to consider prism wear (e.g. acquired sixth nerve palsy), botulinum toxin (e. g. VI nerve palsy[37] infantile esotropia[38,39,40]) and exercises (e. g. convergence insufficiency, distance esotropia and symptomatic phorias). In general there is a more profound degradation on visual acuity and binocular vision of strabismus with younger children, and so increased potential gains and risks with their management.

3.12 Facilities

The appropriate facilities for children in hospital are defined in 'Ophthalmic Services for Children'.[1,2]

Whether the consultation takes place in the community or in hospital 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 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/or pictures or TV screens to attract visual attention.

3.13 Communication

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

Groups involved:
Patient and family
Hospital administration
Medical staff, orthoptists and optometrists in hospitals
Community paediatricians
General practitioners, health visitors
Allied Services (e.g. teachers, school nurses, community optometrists)

Good communication between staff is essential in order to provide coherent advice to parents. A clear and detailed medical, orthoptic and optometric record should be kept and be mutually available when patients attend clinics and on admission for surgery. Letters should be sent to general practitioners and parents. It is good practice to copy correspondence to the community paediatrician and other allied services with the parents/patients permission.

We recommend the local provision of information sheets for parents and children explaining the nature of the conditions concerned and their treatment and expected

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

List of Appendices:

- **Appendix 1 - Clinical Examples**
- **Appendix 2 - Care Pathway**

Appendix 1: Clinical Examples

Infantile Esotropia

Definition: An esotropia that is constant by 6 months of age.

Alternative Terminology: Congenital Esotropia; Essential Infantile Esotropia

Incidence: Estimates vary from 8% of childhood esotropia[11] and 1 in 400 live births[41]

Age: Onset before 6 months of age

Underlying Cause: Idiopathic (Unknown)

Presenting Scenario: Parents/carers see inwardly turned eye from an early age.

Classic Findings:

Good visual acuity each eye (in the majority of cases)
Amblyopia 13-33%[42,43]
Binocular vision absent
Refractive error uncommon[44]
Large angle of esotropia
Cross fixation
Asymmetrical optokinetic response[45]
Latent nystagmus
Over elevation in adduction (develops later)
Dissociated vertical deviation (develops later)

Differential Diagnosis:

Early onset Accommodative Esotropia
VI nerve palsy
Duane Syndrome
Nystagmus block esotropia
Sensory Esotropia

Treatment Aims:
Correction of amblyopia[42,43]
Surgically align eyes
Development of binocularity (normal or subnormal)[46-51]
Correction of persistent over elevation in adduction and “V” pattern
Treatment of DVD[52-56]

Controversies: Definition: some use an esotropia by 12 months of age[57]

Effect of surgery on prevalence of amblyopia[42, 43, 44]
Age at Surgery[46, 47, 48,51,58,59,60,61,62, 63]
Indications for Toxin[38, 39, 40, 64]
Surgical Procedure: Bilateral medial rectus recession vv unilateral medial rectus recession and lateral rectus resection
Two or Three muscle surgery[65, 66, 67]

Outcome: In eyes aligned within 10 prism dioptres of orthophoria up to one third of patients develop subnormal binocular vision. Some evidence suggests early surgery is associated with a better binocular outcome.[62]

Fully Accommodative Esotropia

Definition: An esotropia that is acquired, is either constant or intermittent (before treatment), which is straightened by correcting the associated hypermetropia.

Alternative Terminology: Refractive accommodative esotropia

Incidence: 36.4% of Childhood Esotropia[11]

Age: Onset usually between ages 2-5 years old

Underlying Cause: Uncorrected hypermetropia[11]

Presenting Scenario: Parents see inwardly moving eye when tired, or concentrating on objects close by. Children occasionally exhibit signs of distress when the eye is squinting.

Classic Findings:

Good Vision in each eye (at time of onset of strabismus)
Development of amblyopia over time (if uncorrected)
Hypermetropia (usually more than +2.0 D)
Esophoria or esotropia
Normal AC/A ratio
Family history often present
Restoration of binocular single vision with spectacle correction
Or may control to a microtropia

Differential Diagnosis:

Non accommodative esotropia
Constant esotropia (binocularity not re-established with spectacle correction)
Also known as partially accommodative esotropia or constant esotropia with an accommodative element
VI nerve palsy
Congenital esotropia
Cyclical Esotropia
Convergence Excess Esotropia
Near Esotropia

Treatment:
Full Cycloplegic hypermetropic correction
Orthoptic treatment to expand base in fusion range

Controversies: Role of surgery
Indication for miotic drops

Outcome: Restoration of binocular function (by definition).
Continued Glasses (or contact lens) wear required, possibly life long
Minority loose binocularity despite apparent good glasses compliance and develop constant esotropia
Rarely patients progress to myopia in adolescence, with breakdown to esotropia with myopic correction.

Constant Accommodative Esotropia

Definition: A group of esotropias that are helped, but not cured, with glasses for hypermetropia

Alternative Terminology: Partial Accommodative esotropia
Constant Esotropia with an accommodative element

Incidence: 10% of Childhood Esotropia[11]

Age: 2-5 years old

Underlying Cause: Hypermetropia. Poor Fusional Reserves. Esophoria. Not fully understood

Presenting Scenario: Esotropia seen when tired or concentrating on close objects.

Classic Findings:

Amblyopia
Absent binocular function
Suppression of squinting eye
If small angle, development of abnormal retinal correspondence (ARC)
Hypermetropia
Constant esotropia even with “full correction”
Family history

Differential Diagnosis

Fully accommodative esotropia
VI nerve palsy
Congenital esotropia
Cyclical Esotropia
Convergence Excess Esotropia

Treatment
Full Cycloplegic hypermetropic correction
Amblyopia treatment (occlusion, atropine, other penalisation techniques) [1,34,68-81]
Prism adaption prior to surgery[82]
Surgery to restore ocular alignment
Restoration of binocular function rare

Complications: Treatment of children in whom amblyopia treatment may reduce their suppression leading to intractable double vision. The density of suppression can be monitored with a Sbisa Bar to reduce risk of diplopia[25].

Outcome: Amblyopia can be refractory despite apparent good compliance.

Good alignment is usually possible. Restoration of binocular function is rare, with continued suppression a more usual (and favourable) outcome.

Non Accommodative Esotropia

Definition: An esodeviation occurs after 6 months of age that is not improved with hypermetropia correction.

Alternative Terminology: Acquired non-accommodative esotropia

Incidence: 16% of Childhood Esotropia[11], 17.7/100,000 live births[83]

Age: 2-5 years old

Underlying Cause: Not fully understood

Presenting Scenario: Esotropia seen when tired or concentrating on close objects.

Classic Findings:

Onset maybe acute
Family history of strabismus 34%[83]
Reduced binocular function
Suppression of squinting eye
Amblyopia 41%[83]
Low hypermetropia or emmetropia (Mean of +1.42[83])
Esophoria breaking down to esotropia
Family history

Differential Diagnosis

Accommodative esotropia
VI nerve palsy
Cyclical Esotropia
Other neurological disease[84]

Treatment
Trial of full hypermetropic correction
Amblyopia treatment (occlusion, atropine, other penalisation techniques) [1,34,68-81]
Prism adaption prior to surgery[82]
Surgery to restore ocular alignment (73%)[83]

Outcome: Good outcome is usually possible. Restoration of binocular function is unusual, but good alignment with continued suppression is a favourable outcome.

Convergence Excess Esotropia

Definition: An intermittent esotropia with binocular single vision present at distance fixation but esotropia on accommodation for near fixation.

Terminology: The term convergence excess is sometimes used to include patients with a constant esotropia and no binocular vision. In the UK this would be called a **constant esotropia with an accommodative element**.

A **near esotropia** is a condition with an increased angle for near viewing but not associated with a high AC/A ratio. The term **non-accommodative convergence excess** is sometimes used for this condition.

Near distance disparity is considered relevant if over 8-10 prism dioptres.

Incidence: 27% of esotropia have near distance disparity[73], but the prevalence of convergence excess is less.

Age: Onset is usually between 1-4 years old, but can be up to aged 10 years.

Underlying Cause: High AC/A ratio: not fully understood what underlies this.

Presenting Scenario: Parents see inwardly moving eye when tired, or concentrating on near objects. Children occasionally exhibit signs of distress when the eye is squinting. Older children may report double vision at near.

Classic Findings:

Good vision each eye
Amblyopia rare
Binocular in the distance, reduced at near unless corrected with near add
May control to a fully accommodative esotropia at distance (with hypermetropic glasses)
Variable control at near
Esotropia at near, which may be phoric to a non accommodative target
High AC/A – leading to deterioration of control[85]
Poor near controlled binocular vision (CBA)

Differential Diagnosis:

Non accommodative esotropia
Constant esotropia
Near Esotropia
Hypo accommodative Esotropia[86]
Congenital esotropia

Treatment:
Full cycloplegic hypermetropic correction where present
Bifocal glasses – split pupil[87,88]

Orthoptic exercises[89]
Miotics[90]
Surgery[91,92]

Indications for Surgery:

Reducing binocularity at near
Reducing control with other forms of treatment (e.g. bifocals)
Parent/Doctor preference over other treatments (e.g. bifocals).
Orthoptic exercises not progressing.
Consider prism adaptation to the near angle[93,94, 95]

Type of surgery: large bilateral medial rectus muscle recession[96], pulley surgery[97], slanted recession[98], posterior fixation[99, 100, 101], medial rectus recession with resection.[102]

Controversies: The terminology
Use of bifocal glasses.
Indications for surgery.
Surgical Procedure

Outcome: Is generally good. Most data suggest consecutive exotropia at distance occurs in approximately 10% of cases[92, 95]

Intermittent Distance Exotropia

Definition: An intermittent exotropia, with a larger angle at distance.

Terminology: Intermittent exotropia, distance exotropia, exotropia of divergence excess type.

Incidence: Up to 1% of all children[4], or 32 per 100,000 of children under 19[103].

Age: Onset usually between 2-4 years old

Underlying Cause: unknown

Presenting Scenario:

Parents see outwardly moving (or turned) eyes when tired, daydreaming or in bright sunlight.
Uniocular eye closing in bright sunlight.
Double vision rare. Usually few if any child reported symptoms.

Classic Findings: (104)

Good visual acuity
Refractive error – same as population
Amblyopia in 16% (VA = 6/9 or worse)[12]
Control variable – worse for distance fixation
Good sensory single vision for near
Hemi-field suppression (not universal – some have double vision)
Poor motor fusional reserves
Exophoria/exotropia on cover testing
Convergence normal
AC/A normal, true high or false high (tenacious proximal fusion)[105]

Differential Diagnosis

Infantile Exotropia (usually constant, onset before 6 months of age)
Sensory Exotropia (poor unilateral vision)
Convergence weakness (bigger angle for near)
Convergence Insufficiency (poor convergence)

Treatment [12,106,107,108]
Correction of refractive error[104]
Maintain/treat visual acuity[104]
Monitor control[107,109,110,111,112]
Unilateral or alternating occlusion[113,114]
Minus lens therapy[115,116]
Exercises[108]
Surgery[104]

Indications for Surgery: [107,108,117,118]

Reducing control
Surgeon/parent/patient preference
To improve or maintain Binocular single vision[119]

Type of Surgery: unilateral recess resect, unilateral or bilateral lateral rectus recession[120,121,122]

Controversies: The value (if any) of orthoptic exercises/vision therapy
Indications for surgery.
Ideal age for surgery[123]
Natural History
Long term prognosis.

Complications: Post operative over corrections.[124]
Managed by alternate patching, temporary prisms, toxin or re-operation

Outcomes: Most studies suggest a favourable outcome for surgery can be achieved in around 80-90% of cases[121, 125-129]

Ongoing Studies: Pilot RCT comparing Surgery to Observation for intermittent exotropia. ISRTN:44114892; Decision making in intermittent distance exotropia (X(T)).

Convergence Insufficiency

Definition: Convergence insufficiency (CI) is diagnosed where convergence is less than 10cms from the eyes, and where convergence is absent or cannot be maintained without symptoms. Some authors refer to more than 6 cm from the nose.

Terminology: The term has a different definition according to the Convergence Insufficiency Treatment trials (CITT), which seems to represent how the term is used in the USA. In the CITT, the term also includes exophoria at near of at least 4 PD greater than at far, and insufficient positive fusional convergence at near, or minimum positive fusional vergence of 15 PD base-out break.[130, 131]

Prevalence: The prevalence of this condition is quoted as being between 2 and 33% in a recent systematic review[132]. The wide variation is partly based on different definitions, but also reflects that this is often treated in different settings (for example, hospital based orthoptic practice and community optometrists). Based on the UK definition it is likely that the true prevalence is nearer the lower value.

Age: Onset (in children) usually between age 8 and 16 years old.

Underlying Cause: primary idiopathic, or secondary to exophoria, some drugs, small vertical deviation, accommodative or refractive problems, head injury or uncorrected myopia.

Presenting Scenarios: These can be varied. Symptoms can vary widely. These may trigger a visit to an optometrist. Many optometrists treat effectively with simple pen convergence, and only refer more complex or unresponsive cases to hospital services. This would be the commonest presentation in the UK.

Classic Findings:

Symptoms – “eye strain”, asthenopic symptoms associated with close work
Good visual acuity
No specific refractive error, although can occur secondary to uncorrected myopia
Reduced convergence (less than 10 cm from the eyes)

Differential Diagnosis

Near exophoria (convergence weakness type)
Reduced base out fusion range (which can be associated with CI)

Treatment :
Convergence exercises (at home)[130, 131, 133, 134, 135]

Indications for Surgery:

None

Controversies: Terminology

Convergence weakness exophoria: In the UK, exophoria with a discrepancy between near and distance prism cover test measurement would not be considered significant unless it was 10 prism dioptres or more. A 4PD difference would be classed as a non-specific exophoria, and would not be considered large enough to make a diagnosis of convergence weakness exophoria. In the case of a convergence weakness exophoria (where there is an increase of 10PD or more at near), where this is considered to be causing symptoms, treatment would be offered. Orthoptic exercises would include improving positive fusional vergence (base out/convergence range) and positive relative vergence in addition to convergence exercises of push-up and jump vergence. The order of the different exercises would depend on the patient and individually adjusted to their results. These would be conducted as an outpatient, with the expectation of exercises being carried out at home.

If the near angle was 20 PD or larger, exercises may not be successful in isolation. It is suggested that intervention such as prisms, botulinum toxin or surgery may be required.

Insufficient positive fusional vergence: If a reduced base out fusion range was part of the examination findings, exercises would be aimed at improving the fusion range, in addition to exercises as mentioned above.

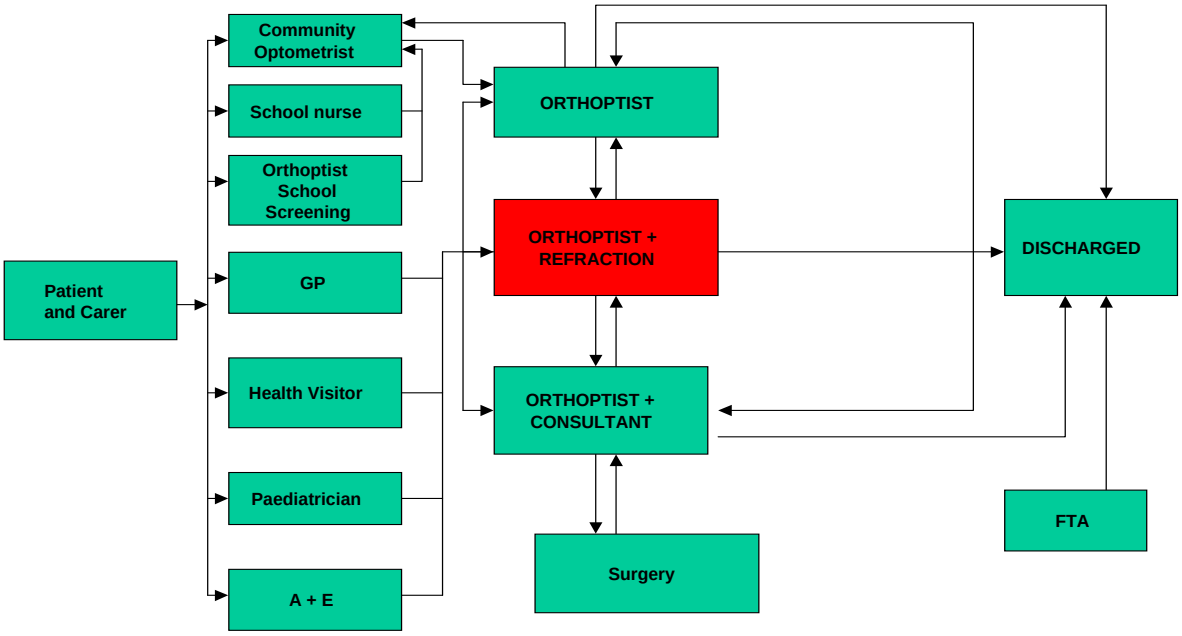
The main conclusion of the CITT was that office based intensive orthoptic exercises achieved a significantly greater improvement in symptoms and clinical measures of near point of convergence than home based pencil push ups. The paper has been criticised on many fronts. Kushner[136] carried out a survey that concluded that most paediatric ophthalmologists and orthoptists do not use unmonitored home treatment or pencil push ups only. He documented that in addition to pencil push ups to one target, additional exercises are used to a variety of targets, additional prisms, jump convergence exercises and stereogram convergence exercises can be introduced. Handler commented that the control group did not represent the standard of care. [137] The exercises are performed at home, but follow up is meticulous and not as outlined in the home arm of the CITT. Kushner quoted a small audit that showed that such methods carried out at home are successful without the need for office based exercises. Wicks published an audit that showed that home based convergence exercises (including stereograms), were successful in 88%. [138] Wallace noted in the editorial that accompanied the 2008 CITT study that the total time was less in the home treatment group, [139] a point echoed by Granet. [140] Lastly Handler has criticised the symptom score used in the CITT, as being too vague and in some instances repetitive.[137]

Complications: No complications of treatment are commonly reported.

Outcomes: There is a paucity of outcome data published, but anecdotal data is that treatment is successful in the majority of patients. Exercises can be reduced or abandoned usually before 6 months.

Ongoing Studies: None known.

Appendix 2: Care pathway



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No relevant declarations of interest

8. Review Date of these Guidelines

These guidelines will require revision in the light of new information and the proposed review date is December 2013.