Strabismus (squint) classification and management

What is strabismus (squint)?

Strabismus, or squint, is any misalignment of the eyes. As a result the retinal image is not in corresponding areas of both eyes, which may result in diplopia in adult patients and can lead to amblyopia in childhood. Most strabismus results from abnormalities of the neuromuscular control of eye movement, although (less commonly) the cause may be disorder of the external ocular muscles. Strabismus is most commonly described by the direction of the eye misalignment:

- The prefixes '**eso**' and '**exo**' refer to an inward and outward ocular deviation respectively.
- The prefixes 'hypo' and 'hyper' refer to a downward or upward deviation respectively. This form of squint is less common.
- Some horizontal squints vary in severity with up/down gaze. If the deviation is greater in the upward gaze than in the downward gaze, it is said to follow a 'V' pattern. If it is greater in the downward gaze than the upward gaze, it is said to follow an 'A' pattern. These terms can be applied to both esotropias and exotropias.

A manifest quint is referred to as **heterotropia**. It is unusual for eyes to be perfectly aligned (orthophoria): most people have a very slight tendency to deviate their direction of gaze, particularly when gazing into the distance or daydreaming, so resulting in mild latent squint, or **heterophoria**. Heterophoria can develop into heterotropia if:

- Muscle strength is inadequate to maintain eye alignment.
- Stimulus to maintain eye alignment is weak (eg, blurred vision).
- There is a problem with the neurological pathway.

Classifying strabismus[1] [2]

There are a number of ways of classifying squints:

- **Congenital** (onset before six months of age) or **acquired**.
- **Right, left or alternating**: patients will squint with either the right or the left eye but never with both eyes simultaneously. In an alternating squint, the patient is able to alternate fixation between their right and left eye. This alteration between the left and right eye is mostly spontaneous but may be voluntary in some cases. Where a patient tends to consistently fixate with one eye and squint with the other, the eye that squints is likely to develop some amblyopia. Someone whose squint alternates is very unlikely to develop amblyopia because both eyes will receive equal visual stimulation.
- **Permanently or intermittent**: most intermittent esotropias are accommodative in origin.
- **Manifest** or **latent**: manifest squint is present when the eyes are open and being used; whereas in latent squint the eye turns only when it is covered or shut.
- **Concomitant (non-paralytic) or incomitant (paralytic)**: strabismus can be concomitant, where the size of the deviation does not vary with direction of gaze - or incomitant, where the direction of gaze does affect the size, or indeed presence, of the squint. Most esotropias are concomitant and begin early in childhood, typically between the ages of 2-4 years. Incomitant strabismus, or paralytic squint, occurs both in childhood and adulthood as a result of neurological, mechanical or myogenic problems affecting the muscles controlling eye movements.
- **Primary, secondary or consecutive**: most are primary. Secondary strabismus arises as a consequence of loss or impairment of vision. Consecutive strabismus arises following overcorrection - eg, exotropia resulting from overcorrection of esotropia.
- **Infantile (congenital or essential) strabismus**: is the common condition characterised by a squint (of various sorts as described above) in an otherwise normal infant with no refractive error.

- **Situational**: some squints are situational - eg, a patient may have a constant esotropia for reading but an intermittent esotropia for distance (but rarely vice versa).

There are other classifications which combine these elements with whether the problem is on near or distant gaze, whether there is accommodative ability or not and whether the problem changes over time (eg, exotropia becoming an esotropia).

Types of strabismus[1] [3]

Esotropia

This describes inward-turning squint. Esotropia is sometimes referred to - incorrectly - as lazy eye. (Lazy eye in fact refers to amblyopia, which can be a consequence of childhood esotropia.)

Accommodative esotropia is an inward turning of the eyes during the effort of accommodation. It is often seen in patients with significant hyperopic refractive error who over-converge in the effort to accommodate. This over-convergence associated with the extra accommodation required to overcome a hyperopic refractive error can precipitate a loss of binocular control and lead to the development of esotropia.

Exotropia

This describes an outward or divergent squint. It often begins intermittently with daydreaming or tiredness and it generally progresses in frequency and duration.

Convergence insufficiency is a common form of exotropia that responds well to orthoptic vision therapy including exercises. This disorder is characterised by an inability of the eyes to work together when used for near viewing, such as reading. Instead of the eyes focusing together on the near object, one deviates outwards.

Hypertropia/hypotropia

In hypertropia the visual axis of the affected eye is above that of the fixating eye (hypotropia is the converse). It is far less common than horizontal strabismus and usually develops after childhood. Squints are usually incomitant. Changes in head posture are common (to limit the resulting diplopia). It is most commonly due to superior oblique paresis.

Pathophysiology of strabismus

Strabismus often occurs in children who are otherwise completely normal. However, children with disorders that affect the brain such as cerebral palsy, Down's syndrome, hydrocephalus and space-occupying lesions are more likely to develop strabismus.

Stroke is the leading cause of strabismus in adults. Neurological problems and Graves' disease (thyroid eye disorders) are common causes. Trauma may also cause strabismus through damage to the oculomotor cortex and nerves or through direct damage to the muscles of the orbit.

The pathophysiology is most easily considered by examining non-paralytic and paralytic squint separately.

Non-paralytic (concomitant) squint[3] [4]

Non-paralytic, concomitant squint is usually congenital. Affected children have full movement in both eyes if tested separately and extraocular muscles and nerves are grossly normal. There is no diplopia and the angle between the longitudinal axes of the eyes remains constant on testing eye movement.

Epidemiology

- Ocular misalignments are common in newborns: one study found the prevalence of these to be about 73% in 1-month-old babies, reducing to 50% in 2-month-old babies and virtually disappearing in normal 4-month-old babies[5] .
- Pathological misalignment affects about 5% of 5-year-olds (of these, 60% have eso-deviations and 20% have exo-deviations). This drops to around 3% of those aged 13-24.
- Esotropias appear to be more common than exotropias among Caucasians but, in the few studies looking at prevalence of this problem among non-white populations, the reverse appears to be true in West Indian children and patients of oriental descent.
- Intermittent exotropias are more common than constant exotropias, which tend to be associated with other abnormalities.

- Approximately 30% of patients with strabismus have a relative who is or was affected,and most families are concordant for the type of strabismus (eg, esotropia or exotropia).

Strabismus presentation in children [3] [4] [6]

- Parental concern due to the presence of a manifest squint.
- Some children (particularly those with intermittent exotropia) intermittently close one eye, especially when outside in the sunlight.
- Motor skills may be reduced in amblyopic children but particularly those with strabismus (fine motor tasks requiring speed and accuracy are most affected).
- At preschool screening.
- Compensatory head tilt or chin lift to minimise diplopia and enable binocular viewing.

- Most children have no associated disorders but there may be a history of risk factors raising the alert including:

    - Family history of strabismus or amblyopia.
    - Prematurity.
    - Neonatal jaundice.
    - Encephalitis.
    - Meningitis.
    - Cerebral palsy.
    - Craniofacial abnormalities.
    - Learning disabilities ± syndromes - eg, Down's syndrome and Turner syndrome.
    - Fetal alcohol syndrome.
    - A febrile illness may occasionally precede accommodative esotropia.

    There may also be associated ocular pathology including:

    - Refractive error, particularly anisometropia (the refractive error is different in both eyes) and high hypermetropia (very long sight).
    - Media opacities such as cataract.
    - Retinal abnormalities such as retinoblastoma.

Presenting features in adults [4]

- The patient usually notes the strabismus. It may also be obvious to others.
- Most often, adults complain of diplopia (double vision).

- Although not specific for strabismus, some patients complain of asthenopia (ocular discomfort) with 'eye strain' or headaches in certain situations. They may also note a 'pulling sensation' and be aware of when the eyes are misaligned.
- Some adults describe psychosocial implications, such as uncertainty in where to look, and that other people are confused as to whether the patient is looking at them or elsewhere.

Examination [2] [5]

- Determine on history and examination whether the symptoms are intermittent or constant and whether they are worsening. Early childhood eye deviation is common and settles by 4 months but this type is always intermittent.
- Parents commonly discover strabismus. In one study, strabismus or leukocoria as the presenting symptom of retinoblastoma was discovered by a family member in 75% of cases.
- The three screening methods needed to detect strabismus are:
    - Gross inspection.
    - Light reflex tests, including the Brückner test (inspection for a red reflex).
    - Cover tests.
- In clinic, ophthalmoscopy and measurement of visual acuity are also crucial.

- A young baby should be examined for the presence of epicanthic folds (crescenteric folds of skin on each side of the nose) which could give rise to pseudoesotropia (the impression that the eyes are turned inwards when in fact they are not). The corneal reflection test (Hirschberg's test) can help to rule this out:
    - **Hirschberg's test**: this gives a rough estimate of the degree of strabismus. Hold a pen torch about an arm's length (c. 33 cm) away from the patient and shine it in front of their eyes. If the patient is able to understand instructions, ask them to look at the light (babies will tend to look towards it anyway, even if briefly). Observe where the reflection of the pen torch lies with respect to the cornea. It should be central bilaterally. If it lies at the inner margin of the pupil, there is an outward deviation (exotropia) of the eye. If it lies at the outer margin, an esotropia is present.
- Look for facial asymmetry (either craniofacial abnormalities or head tilt) and obvious eye abnormalities - eg, ptosis or proptosis.
- Perform the cover/uncover test. If this appears to be normal, try the alternate cover test:
    - **Cover/uncover test**: an object to focus on is held in front of the patient who is instructed to focus on it. One eye is completely occluded for several seconds and the uncovered eye is observed for movement as it focuses on the object. This eye is then covered and the other eye is observed for movement. Movement of the eye outwards confirms that there is an esotropia (ie the eye was turned inwards initially) and vice versa for exotropia. The test is repeated for objects at six metres and far distance, which may also reveal a vertical squint.
    - **Alternate cover test**: this is done in a similar fashion to the previous test but the occluder is rapidly switched from one eye to the other. There is now no longer bifoveal stimulation (so each eye is seeing a separate image). Observing the eye movement as the occluder is removed, note whether it moves inwards (ie there is a latent exophoria and the eye has to move in to see again) or outwards (revealing a latent esophoria).
- Assess the patient for evidence of any other ocular abnormality or systemic abnormality (see risk factors and associations, above).

- If the visual acuity is subnormal, investigation is needed. It may be as simple as a refractive error but it could be due to more sinister causes, such as retinoblastoma, congenital malformations, cataract, optic neuropathy or cortical blindness.

Referral

- A neonate with a constant squint or with a squint that is worsening from 2 months of age should be referred to an ophthalmologist [7] .
- Any older child with a suspected squint should be seen in the Eye Clinic. The earlier the referral, the better chance the child has of avoiding the possibility of amblyopia.

Investigation

There will be an orthoptic assessment (to assess the visual acuity and ascertain the presence and nature of the squint) as well as a medical review to ensure that the eye is otherwise healthy. Tests may include assessment of motility, accommodation, fixation, binocularity, stereopsis and refraction. If there is suspicion of associated diseases, the relevant investigations will be carried out according to clinical findings.

Strabismus management

- Treatment is guided by the exact nature of the squint and by the patient's age.
- Correction of refractive errors is the first step in the management.
- If the patient is less than 8 years old, any concurrent amblyopia will need treating (eg, eye patching ± cycloplegic drops).
- Practical considerations include using plastic instead of glass lenses for spectacles made for children and ensuring that the lenses are large enough to prevent the child from looking over them.
- Some patients are treated with prisms (placed on spectacle lenses).
- Adaptation to refraction may take up to 18 weeks. Follow-up by an orthoptist usually occurs at six weeks and between four and ten times a year following this.

- If treatment attempts have failed or if the squint is large enough, patients may go on to have surgical alignment (particularly for esotropias). A combination of muscle recession (it is moved backwards on the globe and so its action is weakened) and antagonistic muscle resection (a segment of muscle is removed, so strengthening its action) is used with the aim to restore binocular function. Sometimes, adjustable sutures are used to enable minor corrections to be made without having to go through a further full surgical procedure. It may take more than one procedure to achieve the satisfactory result but few surgeons would operate more than two or three times.

Esotropia: specific treatments [8]

- Infantile esotropia (defined as esotropia constant by 6 months of age, although some authorities use a cut-off point of 12 months) is best managed with early surgical intervention to optimise outcome. There is evidence that early surgery is associated with better binocular outcome.
- New developments include chemodenervation by injection of botulinum toxin in one or more extraocular muscles. A Cochrane review produced mixed results and suggested that further trials were needed to compare the cost-effectiveness of this treatment compared to surgery.
- Another approach has been to use miotic agents (eg, cholinesterase inhibitors) which reduce accommodative effort and convergence by stimulating ciliary muscle convergence. However, side-effects limit this use.

Exotropia: specific treatments [9]

- Where this is intermittent, treatment is more commonly sought by the patient or parents for cosmetic reasons than due to a visual need to realign the eyes. One study found that if visual acuity is of concern, surgery is best carried out between the ages of 5-8 years [10] .
- If the problem is mild, eye exercises may suffice. One UK study found that the majority of patients with intermittent exotropia did not need surgery [11] .

Complications [2] [12]

- An uncorrected squint can lead to amblyopia (lazy eye).
- Surgical under- or over-correction may happen during the initial procedure, necessitating further surgery.
- Inferior oblique overactivity may sometimes occur (usually at about 2 years of age) so patients may need further surgery despite an apparently good initial result.
- Dissociated vertical vision (the eye drifts up and out during periods of inattention) can occur years after initial strabismus surgery and may warrant surgical intervention if it becomes cosmetically unacceptable.
- One study found that strabismus was associated with significantly worse general health-related quality of life in preschool children [13] . Another study found a significantly increased risk of strabismic children going on to develop adult mental health problems [14] . There are social ramifications too, such as poorer chances of success in job interviews (see 'Psychosocial aspects of squints', below) [15] .

Prognosis

This depends on the nature and degree of the squint and whether there are any associated underlying problems. Generally, early intervention should produce good alignment and limit any amblyopia but perfect stereopsis (3-D vision) is rarely achieved.

Paralytic squint [1]

Paralytic squint is usually acquired through damage to the extraocular muscles or their nerves. Diplopia is usual and maximal in the direction of gaze produced by the weak muscle. The angle between the longitudinal axes of the eyes varies through the range of eye movements.

The III, IV and VI cranial nerves are involved. A nerve palsy may be isolated or there may be multiple nerves involved. Each nerve may be affected at any point along its course from the brainstem to the orbit. Myopathies may give rise to diplopia and restriction of eye movement; in severe cases there may be a degree of paralytic squint. Myopathies, unlike neuropathies, tend to be bilateral. Below is an overview of diplopia and cranial nerve palsies; more detail about these conditions is given in the separate Diplopia and III, IV and VI Cranial Nerve Lesions article.

Diplopia

This is the term used when a patient sees an image in two different places. They are most commonly side by side (horizontal diplopia) but may be one on top of the other (vertical diplopia) or, unusually, oblique to each other. The image from the paralysed eye is always peripheral to the image from the normal eye.

A patient presenting with diplopia should be advised to stop driving and inform the Driver and Vehicle Licensing Agency (DVLA) [16] .

Monocular diplopia

This term is used when the double vision remains on occlusion of the uninvolved eye; it is not caused by strabismus. Common causes include the presence of a refractive error, incorrect spectacle alignment and some media opacities (eg, cataract). Less commonly, it can arise as a result of a dislocated lens, retinal detachment and central nervous system (CNS) disease.

Binocular diplopia

This term is used when the double vision is corrected when either eye is occluded. It may be intermittent, such as in myasthenia gravis and when there is intermittent decompensation of an existing phoria. Constant binocular diplopia is more typical of an isolated cranial nerve palsy (III, IV or VI cranial nerves), orbital disease (eg, thyroid eye disease), post-surgery or post-trauma and with various CNS problems.

Specific causes of paralytic squint

The direct causes are divided into nerve disorders, muscle disorders and systemic disorders. Each of these may have one of several possible underlying causes and thorough investigation with appropriate imaging is essential.

If strabismus occurs acutely, the patient must be urgently referred to an ophthalmologist, as it may represent an intracranial process such as a mass lesion, aneurysm, raised intracranial pressure, CNS infarction, or inflammation in the CNS. Children with craniosynostosis and craniofacial syndromes also require evaluation by an ophthalmologist for the detection of possible strabismus [4] .

Third cranial nerve palsy [17]

- **Presentation**: there may be external ophthalmoplegia with partial or complete motility problems, resulting in varying degrees of squint, or internal ophthalmoplegia (partial or complete impairment of pupillary reactions). There may also be a ptosis.
- **Aetiology**: pupil-sparing causes tend to relate to ischaemic microvascular disease (and rarely, cavernous sinus syndrome). Pupil-involving disease usually arises as a result of an aneurysm but can also occur as a result of a tumour, trauma, pituitary apoplexy, herpes zoster and leukaemia. Children may exhibit third nerve palsy as part of an ophthalmoplegic migraine.

Fourth cranial nerve palsy

- **Presentation**: binocular vertical diplopia, difficulty in reading and the sense that things appear to be tilted.
- **Aetiology**: trauma, vasculopathy (often related to diabetes and hypertension) and demyelinating disease. This may also be congenital or idiopathic.

Sixth cranial nerve palsy

- **Presentation**: horizontal diplopia which is worse for distance than near vision and most pronounced on lateral gaze on the affected side.
- **Aetiology**: vasculopathy (usually diabetic, hypertensive or atherosclerotic) and trauma are the most common causes but it is also often idiopathic. Less common causes include an increase in intracranial pressure, cavernous sinus mass, multiple sclerosis, giant cell arthritis, inflammation and infection. Children may also get this as a benign, post-viral (or post-vaccination) condition as well as due to increased intracranial pressure and Gradenigo's syndrome (multiple cranial nerve palsies associated with complicated otitis media).

Multiple nerve palsies [18] [19]

- **Presentation**: there may be a combination of unilateral III, IV and VI cranial nerves resulting in limitation of eye movement (and therefore diplopia), facial pain corresponding to one or more branches of the fifth cranial nerve, a ptosis and small pupil (Horner's syndrome) or a dilated pupil if the third cranial nerve is affected.
- **Aetiology**: there are a number of conditions and syndromes which can give rise to this clinical picture:

    - Arteriovenous fistula (carotid-cavernous or dural-cavernous).
    - Tumours within the cavernous sinus (primary or metastatic).
    - Intracavernous aneurysm.
    - Mucormycosis (particularly in those patients with uncontrolled diabetes and in immunocompromised patients).
    - Pituitary apoplexy.
    - Herpes zoster.
    - Cavernous sinus thrombosis.
    - Tolosa-Hunt syndrome.
    - Rare causes: sarcoidosis, granulomatosis with polyangiitis, tuberculosis.

Ocular myopathies

Ocular myositis [20]

This is an idiopathic, nonspecific inflammation of one or more of the extraocular muscles, usually presenting early in adult life and associated with acute pain on moving the eye. It is treated with non-steroidal anti-inflammatory drugs (NSAIDs) or steroids. It either spontaneously resolves after six weeks or follows a protracted course of recurring episodes. Immunosuppressants and radiotherapy are sometimes effective.

Ocular myopathy (progressive external ophthalmoplegia)

This rare condition is characterised by progressive, bilateral reduction in eye movement, associated with a ptosis, usually before the age of 20. It occurs as a result of mutations in the mitochondrial DNA. Although it may occur in the absence of any other clinical sign, it is usually associated with skeletal muscle weakness. Ultimately, ocular movement may be lost altogether. Treatment options are limited.

Brown's syndrome [21]

This condition may be congenital or acquired (iatrogenic or inflammatory: rheumatoid arthritis, pansinusitis or scleritis) and is characterised by malfunction of the trochlear nerve or the superior oblique. Congenital cases are occasionally treated with surgery and acquired cases may respond to a course of steroids along with treatment of the underlying cause.

Duane's syndrome [22]

In this condition, there is unilateral or bilateral lateral rectus activity during adduction and reduced activity in abduction. This results in a limited ability to abduct the eye and a narrowing of the palpebral aperture on adduction. Most cases are managed conservatively, as there is no amblyopia due to the eyes being straight in the primary position.

Myopathies due to systemic disease [23]

Dysthyroid eye disease

See the separate Thyroid Eye Disease article. Hallmarks of advanced disease are a painful red eye with diplopia, a reduced visual acuity, proptosis, lid retraction and lid lag. Patients may also have restricted eye movements (particularly elevation and abduction) giving rise to a squint - this is known as restrictive thyroid myopathy, exophthalmic ophthalmoplegia, dysthyroid eye disease or Graves' disease.

Myasthenia gravis

See the separate Myasthenia Gravis article. About 40% of patients show involvement of the extraocular muscles with muscle fatigue resulting in intermittent diplopia ± squint.

Management and prognosis of paralytic squint

Patients should be referred to the local ophthalmology team (occasionally, clinical indicators suggest a neurology referral would be more appropriate) for further investigation and treatment. Depending on clinical suspicion, this is likely to involve orthoptic confirmation of the paralytic squint, blood tests and imaging with subsequent management of the underlying cause. If no cause is found or whilst the underlying problem is being corrected, patients may be fitted with prisms (fixed on to glasses) to alleviate the diplopia. Surgery is an alternative if this is unsuccessful. Adjustable sutures are often used to ensure great precision. Prognosis depends on the primary problem.

Psychosocial aspects of squints

Strabismus is usually managed with the aim of correcting double vision in the adult or preventing amblyopia in the child. Whilst these functional outcomes are obviously important, the psychosocial consequences of strabismus are arguably as important. A study has found that both adults and children find strabismus disturbing to look at (regardless of gender, age or whether the respondents had strabismus in their own family) [24] . Curiously, adults found a squinting right eye more disturbing than a squinting left eye. In the same study, children perceived esotropia as more disturbing than exotropia.

The psychosocial aspects go beyond a subjective appreciation of an individual's appearance: individuals may be adversely affected in many aspects of their lives, including finding a partner, job prospects and interaction with peers [15] [25] . As such, a strabismus-specific quality of life questionnaire has been devised but is not widely used.

Dr Mary Lowth is an author or the original author of this leaflet.

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