Strabismus

Definition / Supporting Information

Strabismus (squint) is a misalignment of the eyes.

- In 'comitant strabismus', the angle of misalignment is the same in all positions of gaze.
  - This is not usually associated with neurological disease.
- In 'incomitant strabismus', the angle of misalignment changes in different positions of gaze; eye movement is limited due to:
  - Neuromuscular strabismus
    - Cranial nerve palsies
    - Myasthenia
    - Myopathic strabismus
  - Mechanical strabismus
    - Restriction syndromes such as Duane's syndrome, trauma, fibrosis, and thyroid eye disease

Phoria is a tendency for the eyes to drift apart, but alignment is maintained by fusion.

Tropia is a manifest deviation of the eyes, when fusion has failed or is absent.

- Esotropia
  - Eye turned in
- Exotropia
  - Eye turned out
- Hypertropia
  - Deviation upwards
- Hypotropia
  - Deviation downwards

Keywords / also known as: cross-eyed, crossed eyes, esotropia, misalignment, squint

Essential History

Ask about:

- Age of onset
  - Intermittent deviation of the eyes is a fairly common finding in healthy neonates.
  - Normal binocular co-ordination becomes evident at about 3 months, and strabismus (squint) after this age is significant.

- Known neurological disease
  - Strabismus is often found in association with cerebral palsy and in craniofacial developmental anomalies.
- Prematurity
  - Strabismus, amblyopia, and refractive error are much more common in children with treated or regressed retinopathy of prematurity (ROP).

'Red Flag' Symptoms and Signs

Ask about:

- Blurred / reduced vision
- Headaches
- Timing of onset
- Trauma eg, head injury
- Incomitant squint (angle of misalignment changing in different positions of gaze) with or without new onset nystagmus
  - Rule out neurological pathology (acute onset third nerve palsy can herald intracranial aneurysm and requires immediate inpatient care pending exclusion of aneurysm)
- Rapidly changing squint associated with variable ptosis
  - Possible myasthenia gravis
- Previously intermittent squint becoming constant
  - Risk of binocularity loss
- Loss of a previous abnormal head posture
  - Risk of binocularity loss
- New squint co-incident with other new neurological feature
- Absent or altered red reflex, or parent notices change in blackness of pupil or abnormal pupil reflex in photographs (sign of retinoblastoma)

Look for:

- Abnormalities on detailed neurological examination / cranial nerve assessment
  - Ptosis
  - Absent red reflex
  - Limited eye movements
  - Abnormal red reflex
  - Papilloedema
- Signs of systemic disease
  - Fever
  - Rash

Differential Diagnosis / Conditions

Comitant esotropias

- Pseudoestropia (common)
  - The infant has a wide nasal bridge and wide, prominent epicanthal folds, giving the appearance of esotropia.
  - Hirschberg's corneal light reflection test can document proper eye alignment (and a cover–uncover test will reveal a tropia if present).
- Infantile esotropia
  - Age of onset before 6 months
  - Usually large angle of deviation
  - Low risk of amblyopia due to cross fixation, but patients showing a strong fixation preference for one eye, are at risk
  - Requires intervention
    - Binocularity can be achieved if the eyes are aligned before 2 years of age.
- Accommodative esotropia
  - Age of onset between 1 and 5 years
  - Can be intermittent or constant
  - Amblyopia risk
  - Glasses for treatment of hypermetropia may fully correct or improve squint
    - Treat associated amblyopia
    - Consider intervention for residual angle in glasses
- Sensory esotropia
  - An esodeviation caused by unilateral reduced vision
  - Requires urgent referral and investigation
- Other esotropias
  - Divergence insufficiency
    - Angle of squint is greater at distance than near.
    - Important to rule out bilateral sixth cranial nerve palsies
  - Convergence spasm
    - Usually intermittent and associated with blurred vision and pupil constriction
  - Basic esotropia
    - Constant esotropia for near and distance

Comitant exotropia

- Intermittent distance exotropia
  - Age of onset 2–5 years
  - Worse for distance than for near, with variable control

  - Amblyopia is uncommon
  - Treatment with glasses, orthoptic exercises, prisms, or surgery
- Sensory exotropia
  - Most common ocular misalignment caused by unilateral reduced vision
  - Requires urgent referral, investigation, and treatment
- Convergence insufficiency
  - An intermittent exotropia characterised by an exotropia at near fixation but straight eyes with distance fixation
  - Appropriate near convergence is insufficient.
  - Occurs in older children who may report blurred vision and headaches when reading

Incomitant strabismus

- Fourth nerve palsy
  - The tropia is worse when the patient's head tilts to the side of the weak superior oblique muscle and improves with head tilt to the side opposite the palsy.
    - Patients usually exhibit a compensatory head tilt to the opposite side of the paresis to help keep their eyes aligned.
  - Congenital superior oblique palsy (fourth nerve palsy) is the most common cause of a vertical deviation.
  - Acquired superior oblique paresis is seen in trauma (can be bilateral), vascular disease, multiple sclerosis, intracranial neoplasm, herpes zoster ophthalmicus, and diabetes with mononeuropathy.
  - Acquired or decompensated cases can experience torsional diplopia.
- Sixth nerve palsy
  - Limited abduction and an esotropia that is worse when looking to the side of the palsy
  - Neonates can have a transient sixth nerve palsy, often associated with a facial palsy which resolves spontaneously by 4–8 weeks.
  - Approximately one half of sixth nerve palsies resolve over a 6-month observational period.
  - Neurological causes of an acquired sixth nerve palsy include:
    - Intracranial tumours (see Increased Intracranial Pressure)
    - Meningitis
    - Mastoiditis (Gradenigo's syndrome)
    - Hydrocephalus
  - Bilateral sixth nerve palsy is a false localising sign in raised intracranial pressure.
- Third nerve palsy
  - The eye does not move up or down and is exotropic.

  - Ptosis is usually present.
  - The pupil is large and non-reactive in complete third nerve palsy.
  - Third nerve palsy may be congenital or acquired from trauma, intracranial tumour, viral illness, or posterior communicating aneurysm.
  - Acute, new cases are a medical emergency and managed as an evolving aneurysm unless / until alternative aetiology is discovered.
- Duane's syndrome
  - Co-contraction of the medial and lateral rectus muscles causes globe retraction and lid fissure narrowing on attempted adduction.
  - Some children with Duane's syndrome adopt a compensatory face turn to keep their eyes aligned with each other (ie, avoid a manifest squint).
  - Surgery is not usually indicated, but may be effective for correcting a significant compensatory face turn.
- Brown's syndrome
  - Clinical findings include:
    - Limited elevation in adduction
    - An exodeviation in attempted up-gaze
    - An ipsilateral hypotropia that increases in up-gaze
  - Most patients with Brown's syndrome have good binocular vision with a compensatory chin elevation and slight face turn away from the affected eye.
  - The management of true congenital Brown's syndrome is conservative unless the vertical deviation in primary position is significant.
- Möbius syndrome
  - A rare condition characterised by:
    - Facial (seventh) nerve palsy, sixth nerve palsy, often with a partial third nerve palsy
    - Distal limb abnormalities such as syndactyly (see Congenital Malformations) or even absent limbs
    - Craniofacial anomalies (see Congenital Malformations) can occur (eg, micrognathia, tongue abnormalities, facial or oral clefts)
- Myasthenia gravis
  - Limitation of eye movement and ptosis can vary throughout the day
  - Anti-acetylcholine receptor positive in only 50% of patients with ocular myasthenia
  - Has important systemic dangers (eg, weakness of muscles of respiration)
- Double elevator palsy
  - Congenital limitation of elevation of one eye
  - The term implies paresis of the superior rectus muscle and inferior oblique muscle.
    - However, in 70% of cases the deficient elevation results from a tight inferior rectus muscle.

  - May be mistaken for Brown's syndrome
    - The limited elevation in Brown's syndrome is worse in adduction than in abduction.
- Congenital fibrosis syndrome of extraocular muscles
  - Treatment is surgical recession of the fibrotic muscle and corrective eyelid surgery where needed.

Investigations

To be undertaken by specialist practitioners (eg, Paediatric / Paediatric Neurology / Paediatric Ophthalmology Team(s)):

- If no specific cause of an acquired superior oblique paresis can be found, then a neurological work-up, including neuroimaging, should be performed.

Treatment Approach

To be undertaken by Paediatric Ophthalmology Team:

- Infantile esotropia
  - Best treated with early surgery
    - Surgery is usually performed when the patient is between 6 months and 2 years of age.
    - Peripheral fusion can be achieved if the eyes are aligned before 2 years of age.
- Accommodative esotropia
  - Accommodative estropia is usually associated with farsightedness.
  - The first line of treatment is to prescribe spectacles.
  - If spectacles do not correct the esotropia, then surgery will be needed.
  - Early treatment is critical to achieving the best results.
- Sensory esotropia
  - An esodeviation caused by unilateral blindness
  - Loss of vision may cause an eye to drift
    - If the vision loss occurs before 2 years of age, patients develop esotropia.
    - If the vision loss occurs after 2 years of age, patients develop exotropia.
  - Requires urgent intervention
- Intermittent exotropia
  - Treatment is elective
    - Eye muscle surgery is the treatment of choice for most forms of intermittent exotropia.
    - Usually requires strabismus surgery if the deviation is poorly controlled

  - Indications for surgery include:
    - Increasing exotropia
    - Exotropia is present > 50% of the time
    - Poor fusion control of the exotropia
- Duane's syndrome
  - Strabismus surgery
    - Effective for correcting the compensatory face turn
    - Improves abduction slightly, but does not result in full abduction capabilities
- Brown's syndrome
  - The management of true congenital Brown's syndrome is conservative unless the vertical deviation in primary position is significant.
  - In most cases, waiting until the child's vision is mature before performing surgery is advised because an induced strabismus after surgery is not uncommon and can lead to the loss of binocular vision.
  - Acquired Brown's syndrome
    - Usual treatment is treating sinusitis, if present, and providing oral non-steroidal anti-inflammatory drugs
    - Surgery is not usually indicated for acquired inflammatory Brown's syndrome.
- Congenital fibrosis syndrome
  - Treatment is surgical recession of the fibrotic muscle

When to Refer

Refer patients with new onset comitant strabismus to local optician for:

- Assessment of vision
- Formal measurement of strabismus
- Refraction

Refer urgently to specialist practitioners (eg, Paediatric / Paediatric Neurology / Paediatric Ophthalmology Team(s)) if:

- New onset squint and systemically unwell (look for papilloedema)

Refer urgently to Paediatric Ophthalmology Team if:

- Acute third nerve palsy (arrange emergency transport)
- Diplopia
- Limited eye movements (incomitant strabismus)
- Poor vision and abnormal red reflex
  - Suspect retinoblastoma (NB. the child will likely be systemically well)

  - Reports by parents of leukocoria should be checked with a red reflex test, and if an abnormal red reflex is detected, an urgent referral should be made
- New squint and ptosis
- Sensory esotropia / sensory exotropia

'Safety Netting' Advice

- Patients with acute onset strabismus and associated neurological signs including nystagmus require an urgent systemic evaluation to exclude intracranial pathology.
- New onset strabismus in an infant may also be an important sign of vision loss.
  - Disorders such as congenital cataracts and retinoblastoma can present with strabismus and absent or altered red reflex.

Patient / Carer Information

Please note: whilst these resources have been developed to a high standard they may not be specific to children.

- Information about retinoblastoma (Web page), Childhood Eye Cancer Trust
- Squint (Web page), the NHS website
- Childhood squint video (Web page), the NHS website
- Squint (strabismus) in children (Web page), Patient

Resources

National Clinical Guidance

Suspected cancer: recognition and referral (Web page), NICE guideline NG12, National Institute for Health and Care Excellence

Suggested Resources

Please note: these resources include links to external websites. These resources may not have national accreditation and therefore PCO UK cannot guarantee the accuracy of the content.

Taylor K, Elliott S. Interventions for strabismic amblyopia. Cochrane Database of Systematic Reviews. 2014,7

Royal College of Ophthalmologists. Guidelines for management of strabismus in childhood. London: RCOphth; 2012

Archer SM, Sondhi N, Helveston EM. Strabismus in infancy. *Ophthalmology*. 1989;96(1):133-137 [PubMed]

Childhood squint (strabismus) (Web page), Royal National Institute of Blind People (RNIB)

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