Diagnosis and management of childhood squints: investigation and examination with reference to red flags and referral letters

BACKGROUND
Strabismus (squint) is a condition in which the eyes are misaligned. While one eye is directed at an object, the other eye may turn in (esotropia), out (exotropia), up (hypertropia), or down (hypotropia). This misalignment may be persistent or intermittent. Newborn babies may have evidence of intermittent horizontal strabismus, which may be normal.

Any baby who continues to suffer from strabismus beyond the age of 3 months should be referred to secondary care for further investigations. The overwhelming majority of squints in children will be benign and can be referred for further investigation on a routine basis. However, if there are any red flags (Box 1) the child should be referred urgently (within a few days) to a hospital eye department (not an optician).

IMPORTANCE OF RAPID DIAGNOSIS
Timely referral and diagnosis is important because strabismus can be a sign of serious pathology, including:

- intracranial pathology, for example, brain tumours; and
- life- and sight-threatening ocular pathology, for example, retinoblastoma.

EFFECTS OF STRABISMUS
Squints in children <7 years old may impair visual development of the affected eye. Unless treated before the age of 7–8 years, this can become permanent. The fixating eye will see clearly, while the deviating eye will have reduced visual acuity. This is termed amblyopia (lazy eye). Vision loss occurs because nerve pathways between the eye and the brain are not properly stimulated. A further consequence of this may be a lack of stereopsis (depth perception). Children with strabismus may be stigmatised at school and suffer from a loss of self-esteem. Such psychological effects could continue into adulthood. They may also find that their career options are somewhat restricted because people suffering from squints are prohibited from certain professions.

HISTORY
The history tries to not only distinguish between causes and types of strabismus, but also to screen for red-flag features that could suggest serious pathology. The important aspects of strabismus history are:

- age of onset;
- which eye (left, right, or both);

Box 1. Red flags
Strabismus could be a sign of coexisting ocular pathology, for example, retinoblastoma or intracranial pathology, as where there is a tumour requiring urgent management. If this is suspected then an urgent specialist opinion should be obtained.

Other red flags are:

- abnormal red reflex;
- limited abduction;
- double vision;
- headaches;
- nystagmus (involuntary, repetitive, side-to-side oscillation of the eyes);
- face turned to the side; and
- other neurological features — strabismus can be associated with neurological disease such as cerebral palsy.

Finding abnormal neurological signs should prompt referral to a paediatrician.
Box 2. Clinical vignette

An otherwise healthy 3-year-old boy presents to the practice with a 4-month history of his left eye turning inward. His mother says it started on and off but is now present all the time. There is no history of double vision, headaches, or other neurological abnormalities. There is a strong family history of strabismus.

The examination shows:
- left eye turning in;
- normal red reflex;
- normal abduction bilaterally;
- no nystagmus;
- no head turn; and
- no other neurological signs.

Next step
Referral (routine) to local eye hospital for ophthalmic review.

Outcome
The child was given glasses for full-time wear, which helped his vision a bit and made the eyes straighter. He later needed 3 months of patching the right eye for 2 hours a day to treat left-eye amblyopia.

REFERENCES

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- frequency and direction of deviation;
- presence of diplopia;
- child or parental concerns about vision;
- birth history — including developmental history;
- glasses wear; and
- family history.

GENERAL ADVICE
Strabismus usually develops at some time in the first 3 years of life; however, it can also develop in older children. The duration and intensity of treatment will depend on factors such as age of presentation.

Although not always easy to decipher, the pattern of symptoms is important, such as whether the strabismus is constant or intermittent. If the pattern is reported as intermittent, the practitioner should note whether the frequency and symptoms are dependent on direction of gaze. Children may develop a very brief period where a strabismus develops when daydreaming or extremely tired.

In strabismus, the diplopia is binocular (present when both eyes are open). Horizontal diplopia would suggest eso- or exotropia. However, vertical diplopia is indicative of vertical strabismus.

Congenital esotropia tends to run in families, so it is important to check if anyone in the family has had eye surgery during childhood.

The list of red flags is shown in Box 1.

EXAMINATION
The patient should be examined for the following features:
- misalignment on inspection;
- red reflex;
- full range of eye movements (particularly abduction in cases of esotropia);
- dysmorphic features — especially craniofacial developmental abnormalities that can be linked to strabismus; and
- abnormal head posture — head posture develops when the child gains head control when upright. Hypertropia in the affected eye may be associated with a head tilt to the opposite side and depression of the chin. This may be due to weakness of the superior oblique muscle.

COVER TEST
The test is performed for near and distance vision with and without glasses while fixating on an accommodative target, for example, a letter of the alphabet.

The patient is asked to fix on the target. Cover one eye with an occluder for several seconds and observe the uncovered eye. If the uncovered eye moves, this is indicative of a manifest squint or tropia. For example, if the uncovered eye moves inwards then the patient has an exotropia, that is, the uncovered eye has moved inwards from its previously exotropic position to take up fixation. Conversely, if the uncovered eye moved outwards it would mean the patient had an esotropia. The test should be performed on each eye.

THE REFERRAL LETTER
The following information should be included in the referral letter:
- age of onset;
- direction of turn;
- concerns about vision and development; and
- presence of red flags — positive and negative findings [Box 1].

TREATMENT FOR BENIGN SQUINTS
Treatment for benign squints will arise from the hospital visit. This will be:
- glasses;
- patching of non-squinting eye [or blurring the vision with atropine drops]; and
- surgery.

Most children do not need surgery and there is a national trend of reducing the numbers of operations for childhood squint. When surgery is performed the eye will be red and sore which can take several weeks to settle. Children can normally go back to school after 2–3 days.

Box 2 represents a common clinical case.