Management of Strabismus

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Introduction

The term “strabismus” came from Greek, which means to squint or to look obliquely. Strabismus means ocular misalignment. One eye is misaligned in relation to the other when focusing on an object. It can be due to abnormalities in binocular vision or anomalies of neuromuscular control of ocular motility.

Classification of Strabismus

There is no single classification which is perfect or all-inclusive. The following are a few of the useful classifications:

According to age of onset:
- Congenital/infantile: a deviation documented prior to the age of 6 months;
- Acquired: an ocular deviation with onset documented after the age of 6 months.

According to the direction of deviation:
- Horizontal: esodeviation (convergent squint), or exodeviation (divergent squint);
- Vertical: hyperdeviation (upward) or hypodeviation (downward);
- Torsional: incyclotorsion or excyclotorsion;
- Combined

According to fusional status
- Phoria: a latent deviation that is controlled by the fusional mechanism so that under normal binocular vision the eyes remain aligned;
- Tropia: a manifest deviation in which fusional control is not present;
- Intermittent tropia: fusional control is present part of the time.

According to fixation/ laterality
- Monocular: definite preference for fixation with one eye;
- Alternating: spontaneous alternation of fixation from one eye to the other.

According to variation of the deviation with gaze position or fixation eye
- Comitant (concomitant): the deviation does not vary with the direction of gaze or fixating eye;
- Incomitant (non-comitant): the deviation varies with the direction of gaze or fixating eye. Most incomitant strabismus are paralytic or restrictive. If acquired, incomitant strabismus may indicate neurologic or orbital diseases.

Assessment of Strabismus

The assessment of strabismus is no different to other diseases. It starts with a good history taking, physical examination and then ordering appropriate investigations if needed.

History

Some special points to note in assessing strabismus include:
- The age of onset/ duration of the deviation or symptoms (e.g., diplopia). Old photographs are invaluable for this purpose;
- Is the deviation/ symptom associated with trauma or physical stress?
- Is the deviation/ symptom constant or intermittent?
- The past treatment should be reviewed.

Assessment of the visual acuity of each eye

Numerous tests are available to check for distance visual acuity, e.g., Snellen letters chart, illiterate E chart, Sheridan-Gardiner chart (for children). If the patient does not have corrective lenses, a pinhole may be used to try to ascertain the best visual acuity potential.

For pre-verbal children, preferential looking tests e.g., Cardiff picture cards can be used to estimate visual acuity. For uncooperative children/patients, each eye can be occluded with hand or occlude to test if any objection is demonstrated when one eye is occluded. The patient may attempt to manoeuvre around the occluder when the good eye is covered but not when the poorly seeing eye is covered.

If these tests fail, simple observation on fixation and following ability can give an idea on the visual function, e.g., “CSM method”. Corneal light reflex is “Central”; fixation on the examiner’s torch when held motionless and when slowly moved about should be “Steady”; the alignment of the two eyes is “Maintained”.

Assessment of stereo acuity

“Stereo acuity” is a sense of depth. Stereo acuity is appreciated when two simultaneous but slightly different images are fused and integrated by the brain.
Assessment of ocular alignment
Abnormal head posture: Before concentrating on examining the eyes, one should note if the patient has any “abnormal head posture”. Abnormal head postures may indicate restrictive or paralytic strabismus. Usually the patient places the head in a position that provides comfortable single binocular vision for the straightened view. Occasionally the head is placed to separate diplopic images maximally. Common “abnormal head postures” include face-turn, head tilt, chin-up or chin down, or any combination of the above.

Corneal light reflex tests: The patient fixates on a pen torch in front of him/her. In normal circumstances, the corneal light reflexes will be central and equal between two eyes. The corneal light reflex will be displaced nasally (inner-side) from an exotropic eye; displaced temporally (outer-side) from an esotropic eye; displaced upward from a hypertropic eye and vice-versa from a hypotropic eye.

The amount of deviation can be estimated by the “Hirschberg method”. Based on the assumption of a 4mm-pupil, the Hirschberg method assumes 1mm of displacement of light reflex across cornea corresponds to 7° of decentration or 15∆ (prism-dioptre), of ocular deviation of the visual axis. Therefore, a light reflex at the papillary margin is about 2mm from the papillary centre, which corresponds to 15°, or 30∆ of deviation. Similarly, a reflex in the midpoint of the iris is about 4mm from the papillary centre, which is roughly 30° or 60∆ of deviation; a reflex at the limbus is about 45° or 90∆ of deviation.

The “Krimsky method” or “Modified Krimsky method” quantifies the light reflex displacement using appropriately held prisms (Fig. 1). The original description suggested placing the prism before the fixating eye, but it can be modified to hold the prism before the deviating eye, which is easier. 3

Figure 1. Krimsky method to quantify the light reflex displacement with prism.

Cover tests: Cover tests are objective tests that measure horizontal and vertical strabismus (Fig. 2). Cover tests require the patient to be attentive and cooperative. Each of the patient’s eye needs to be able to see the target and moves to take up fixation upon that target. If any of these requirements is lacking, the results of cover tests may not be valid. There are 3 types of cover tests: the “cover-uncover test”, the “alternate cover test” and the “simultaneous prism-cover test”.

The “cover-uncover test” detects tropia/manifest squint. The examiner observes the uncovered eye for movement to take up fixation as the fellow eye is covered with an occluder or the examiner’s hand. A movement towards the nose implies exotropia; a movement temporally, esotropia; an upward movement, hypotropia; a downward movement, hypertropia.

For the covered eye, the examiner should observe if it is deviated or moves when the cover is removed. The former implies that the tropia fixation preference shifts to the original deviated eye; for the latter, the eyes become straight again when not occluded and it implies that there is phoria (latent squint).

Phoria is better detected by the “alternate cover test”. Each eye is occluded alternately several times to dissociate the eyes. It is important to transfer the cover quickly from one eye to the other to prevent fusion. The direction of eye movement is noted when the occluder is swung between the two eyes. If no tropia was noted previously by the cover-uncover test, the eye movement elicited by the alternate cover test signifies phoria. If tropia was already present on the cover-uncover test, the alternate cover test measures the total deviation, both latent and manifest. When movement is detected, it can be quantified using prisms. Different powers of hand-held prisms are placed in front of an eye while doing the alternate-cover test until the eye movement is neutralised.

Tropia, when co-exists with phoria, can be measured using the “simultaneous prism-cover test”. This test is done by placing prisms in front of the deviated eye at the same time when the fixating eye is covered.

Assessment of ocular motility
After assessing ocular alignment, the eye movement needs to be thoroughly checked. Three cranial nerves innervate six extraocular muscles (some would consider the levator as the seventh extraocular muscle). The fourth cranial nerve (trochlear nerve) innervates the superior oblique muscle, the sixth cranial nerve (abducens nerve) innervates the lateral rectus muscle and the third cranial nerve (oculomotor nerve) innervates the rest. Each extraocular muscle has different actions in different gaze positions. In the primary position (looking straight ahead), each muscle has its primary action, secondary action and tertiary action. (Table 1).
There are six positions of gaze in which one muscle is the prime mover of the eye. These are called the six cardinal positions. The six cardinal gaze positions together with the primary gaze, straight up and straight down positions, a total of 9 gaze positions need to be examined.

### Investigations for Strabismus

If the visual acuity is subnormal, the reason must be sorted. 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.

If the eye movement is impaired, it can be caused by numerous reasons. The site of pathology can be at various levels including:
- Central nervous system, e.g. stroke
- Cranial nerves, e.g. diabetic mononeuritis
- Neuromuscular junction, e.g. Myasthenia gravis
- Extraocular muscles, e.g. thyroid eye disease

Appropriate blood tests or imaging are indicated to establish the cause for the impaired eye movement.

### Treatment of Strabismus

The treatment of strabismus must be tailored to the patient’s own functional and cosmetic needs. It can be non-surgical or surgical. Non-surgical treatments include simple observation, wearing appropriate corrective lenses or prismatic glasses, doing orthoptic exercise and fogging or partial occlusion of one eye. Each form of non-surgical treatment is discussed in more details below.

#### Non-surgical treatments

**Simple observation:** Some forms of strabismus will go away with time or when the root cause is treated. Examples are Myasthenia gravis, diabetic mononeuritis, temporary limited eye movement related to post-trauma periorbital soft tissue oedema. In cases of small angle strabismus, patients will try to avoid diplopia by a slight abnormal head posture. If this abnormal head posture is not bothering the patient, then conservative treatment can be adopted. Interestingly in cases of very large angle strabismus, patients may not experience any symptoms of diplopia. The second image from the deviated eye is so far away from the main image that the patient is able to ignore it in daily life.

**Correction of refractive error:** This is especially important for children with accommodative esotropia. In these patients, esotropia may be fully or partially corrected when hypermetropic (plus-lenses) glasses are worn (Fig. 3). Whereas for children with intermittent exotropia, control of exotropia is often improved when full myopic (minus-lenses) correction is prescribed. It has also been suggested that even over-minus lenses can be used to induce more convergence associated with accommodation. However the prescription of over-minus lenses is not widely practised locally.

**Orthoptic exercise:** A commonly prescribed orthoptic exercise is the “pencil push-ups” to train up the control of intermittent exotropia or exophoria. A fixation target is placed at an arm’s length and moved towards the nose. The patient needs to keep focusing on the target. Increasing accommodation is induced when focusing on progressively near target, thus convergence of the two eyes is also induced.

**Prismatic glasses:** Hand-held prisms are used to measure the angle of deviation clinically. Small angle prisms can be fitted into spectacle lenses for the patient to wear to neutralise the deviation (Fig. 4).

**Fogging / partial occlusion:** Especially in patients who are not willing to undergo surgery or cannot tolerate wearing prisms, fogging/ partial occlusion of one eye can be used to relieve diplopia. This method can also be used temporarily while waiting for surgery or to buy time for some forms of strabismus to resolve/ stabilise.

#### Surgical treatments

Surgical treatments of strabismus include operating on the extraocular muscles or paralysing the extraocular muscles with Botulinum toxin injection.

**Muscle surgery:** Extraocular muscles are inserted into the sclera. The action of extraocular muscles can be manipulated by: 1. recession (weakening), 2. resection/ plication (strengthening), or 3. transposition (changing the vector of force). The extraocular muscles are accessed via the conjunctiva (i.e. no skin wound on the lids) (Fig. 5A). International tables are used as references to guide the amount of recession/ resection/ transposition to correct the measured deviation. Absorbable sutures are used in most cases...
of strabismus surgery for both muscles (6/O Vicryl) and conjunctival wound closure (8/O Vicryl) (Fig. 5B). The risks of strabismus surgery include residual/recurrent/consecutive strabismus. Rare but important complications include globe penetration (through-and-through sutures), ocular ischaemia (especially after multiple muscles surgeries).

Botulinum injection: Botulinum toxin can be injected into the extraocular muscle(s) under electromyography guidance to temporarily paralyze the antagonist of a weak muscle, such as in the case of a weak lateral rectus in sixth cranial nerve palsy, Botulinum toxin can be injected into the medial rectus as a form of treatment.

Some Examples of Different Types of Strabismus

Congenital/infantile esotropia

“Congenital” esotropia is rarely truly congenital. In fact, many babies have a moderate exodeviation at birth and this would go away by 6 months. Exodeviation can be occasionally seen and these babies may become orthophoric (straight eyes), usually by about 2 months. One cannot predict if a baby will have a deviation at 2 to 4 months. So the documented presence of esotropia by 6 months of age has been arbitrarily defined as “congenital” or “infantile” esotropia. This type of esotropia is typically of a large angle, >30D. Cross-fixation is frequent. Usually there is no significant refractive error. Associated vertical deviations e.g. inferior oblique overaction and dissociated vertical deviation are common. Nystagmus may be present. Monocular smooth pursuit is often asymmetrical. Treatment of congenital esotropia is essentially by surgery. Early surgery before 2 years of age is advocated. 5,6

Acquired accommodative esotropia

Accommodative esotropia is always acquired. The onset of accommodative esotropia is generally between 6 months and 7 years. It usually begins as intermittent esotropia and then becomes constant. Most of these children have high hyperopia (long-sighted) of at least +3.0 dioptres. Atropine drops or ointment is often needed to relieve all accommodation before the true refractive status can be fully revealed. One other feature is that the esodeviation is more at near than at distance fixation. The treatment for accommodative esotropia is to prescribe maximal plus-lenses for constant wear as fixation. The treatment for accommodative esotropia is that the esodeviation is more at near than at distance fixation. Atropine drops or ointment is often injected into the medial rectus as a form of treatment.

Intermittent exotropia

Exotropia is more common than esotropia in Hong Kong. Intermittent exotropia is the commonest among all types. Intermittent exotropia typically presents between the ages of 18 months and 4 years. These children’s eyes are mostly straight during the day. But when they get tired or are looking at distant objects, the exophoria breaks down to exotropia and one eye will deviate out. Often the deviation corrects itself after one or two blinks. Some would progress and the frequency of exotropia increases with time. The initial treatment of intermittent exotropia is to correct all myopia with glasses. Often the frequency of exotropia would reduce as the visual acuity at distance after wearing glasses improves. At near fixation, accommodative convergence is also induced. Accommodative convergence can also be enhanced by orthoptic exercises, such as “pencil push-ups”. To ensure this test is done correctly, an observer should watch for the convergence movements of both eyes. Apart from “pencil push-ups”, sometimes monocular occlusion is used to prevent suppression. If non-surgical treatments have failed and the angle of exotropia is significant (>20D), muscle surgery may be indicated.

Graves’ ophthalmopathy

Graves’ ophthalmopathy is an immunological disorder that affects the orbital muscles and fat. Patients with Graves’ ophthalmopathy are not necessarily hyperthyroid. A minority of patients (<10%) are euthyroid or even hypothyroid. Smoking is a known risk factor that increases the severity of Graves’ ophthalmopathy. 7 Lid retraction and proptosis cause corneal exposure symptoms of grittiness, tearing and photophobia; enlarged or fibrotic extraocular muscles can cause diplopia. The inferior rectus and the medial rectus muscles are most commonly affected. Graves’ ophthalmopathy can be a blinding condition when there is severe corneal complication or compressive optic neuropathy. Overall, there is also a cosmetic issue apart from functional eye problems. When troublesome diplopia due to restrictive strabismus is present, non-surgical treatments can be used first. The thyroid status needs to be controlled and smokers are strongly advised to quit smoking. Strabismus surgeries should be delayed until the overall ophthalmopathy is stable for at least 6 months and after orbital decompression surgery if both procedures are required.

References