Assessing babies and pre-school children for strabismus

Dr Simon Barnard, **Ellis Johnson** and **Alex Levit** present an overview of strabismus or heterotropia, also known in the UK as a squint. It is the condition in which only one of the visual axes is directed towards the fixation object

This article will discuss some of the methods that optometrists use to detect the presence of strabismus during case finding examinations or screening. Management protocols for strabismus will not be discussed but, needless to say, within the profession there is a range of skill sets. Some optometrists might refer all strabismus patients for ophthalmological management. Others, who have the clinical knowledge and experience to treat those types of strabismus that are likely to respond to the therapeutic interventions available to optometrists, will manage the patients themselves by prescribing for refractive errors, and carrying out occlusion therapy and orthoptic exercises on suitable patients.

Why is strabismus important? Strabismus is a common condition with a prevalence of 3.9 per cent in pre-school children and is a risk factor for amblyopia.[1]

Amblyopia is defined as a visual loss resulting from an impediment or disturbance to the normal development of vision.[2] Amblyopia is generally known by lay persons as 'lazy eye', a term which can belie the depth of visual loss. The visual loss is not associated with any recognisable pathological cause but can be attributed to an amblyogenic (amblyopia-causing) factor, most commonly strabismus and/or anisometropia.[3]

The prevalence of amblyopia is 3-4 per cent.[4,5,6] In the population under the age of 20 years, amblyopia is 10 times more common a cause of visual loss than all others taken together, whether caused by trauma or disease.[7] It is difficult to define the respective prevalence of amblyopia secondary to strabismus and to anisometropia as these two conditions often occur together.

Unless a large angle squint is present, it is unlikely to be noticed either by the parent or a non-eye care professional. Optometrists have an important role in detecting strabismus. Conventional wisdom is that early detection of strabismus is important as early treatment and management is more likely to lead to a more successful outcome.

TABLE 1

Vision assessment methods recommended for use based on age

Age	Vision test
6-12 months	Forced choice preferential looking (Keeler gratings)
12-18 months	Cardiff acuity cards (preferential looking)
18-24 months	Kay pictures
2-3 years	Keeler crowded cards
	Crowded Kay pictures
3+ years	LogMAR tests

Figure 1 **Infant with no manifest strabismus, symmetrical corneal reflexes and positive angle lambda in each eye**

Investigation of babies and pre-school children

As with any thorough eye examination, it is essential that a detailed history is taken and documented. Important components include the age of onset of any signs or symptoms, details regarding the strabismus characteristics (which eye, deviation direction, constant or intermittent). Crucially here, parents may fail to offer up information regarding their child, as they may not feel they are relevant, so detailed and exact questioning is vital from the practitioner. For example, any birth problems, forceps or prematurity. Equally important is knowledge of family history of strabismus or amblyopia. Throughout the history, the clinician must be attentive to patient information and be aware that presenting signs may not always be manifest to the parent. It is not uncommon for children to present with the sensory adaptation of suppression, which is not apparent to the parent and only revealed through optometric examination.

Vision assessment

Ideally, both vision and visual acuity should be assessed monocularly. This is far from always being possible. For those children who are very uncooperative, an attempt to obtain binocular vision and visual acuity should be made. For babies and infants, a meaningful assessment of vision may sometimes be achieved, but the clinician will invariably rely on results of tests of eye position and refraction to determine risk factors.

This article is not intended to review methods of assessing vision but some examples of appropriate methods of visual assessment should be chosen according to patients' age and reading ability (Table 1). The importance of the use of crowded test types is stressed here, as these are more sensitive to amblyopia detection due to the crowding phenomenon.

Ocular examination

The assessment of pupillary reflexes ocular adnexa (including eyelid position) and examination of the ocular media and fundi is vital and is especially important if strabismus is present. All patients diagnosed with strabismus should undergo an examination of the ocular media and fundi under the mydriasis induced by the cycloplegic drug used for refraction. Indirect ophthalmoscopy will provide the adequate field of view required. The author has found the Optomap laser scanning ophthalmoscope to be invaluable for capturing a wide field image. Cycloplegic drugs should not be instilled until other tests including that of eye position have been first carried out.

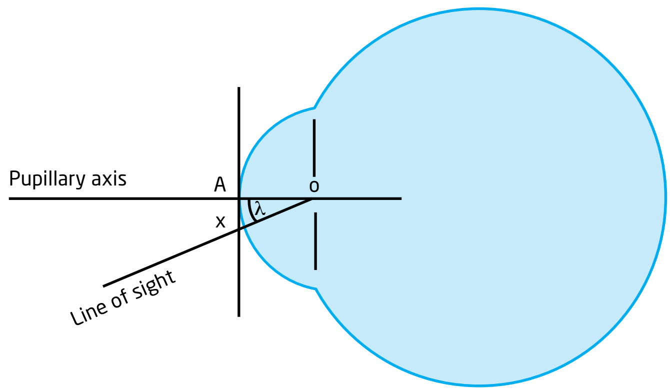

Figure 2 Schematic representation of the components of angle lambda.[10] AO is the distance from the front surface of the cornea to the entrance pupil, measured along the pupillary axis; the pupillary axis is the line perpendicular to the cornea which passes through the centre of the entrance pupil; the distance along the line of sight from the cornea to the entrance pupil is XO; angle AOX is angle lambda

Refraction

It is not the remit of this article to discuss refraction methods in young children. However, the importance of retinoscopy under cycloplegia should not be underestimated.

Eye alignment

An early test for checking eye position and alignment was initially described to by Hirschberg in 1881 with further case studies published in 1885.[8,9]

Hirschberg test

The corneal reflexes (first Purkinje image) are observed from a 50cm working distance using a centrally located pen torch. For a subject whose visual axes are aligned, no asymmetry will be apparent.

It should be understood that the corneal reflex of most fixating eyes is not normally positioned in the centre of the pupil. Generally, the fovea is temporal to the posterior pole, so that the reflex is nasal to the corneal apex, producing a positive angle lambda. Angle lambda is the angle subtended at the centre of the entrance pupil of the eye by the intersection of the pupillary axis and the line of sight. Figure 1 shows an infant with no manifest strabismus and a positive angle lambda in each eye.

Although routinely measured clinically, the angle is commonly wrongly designated as angle kappa. Angle kappa is actually the angle between the visual and pupillary axes measured at the nodal point, a point not clinically accessible.[10] It is angle lambda (Figure 2) that is measured clinically.[11]

If a deviation is present, corneal reflexes may no longer be viewed as being symmetrically placed with regard to the pupil centre. Hirschberg found

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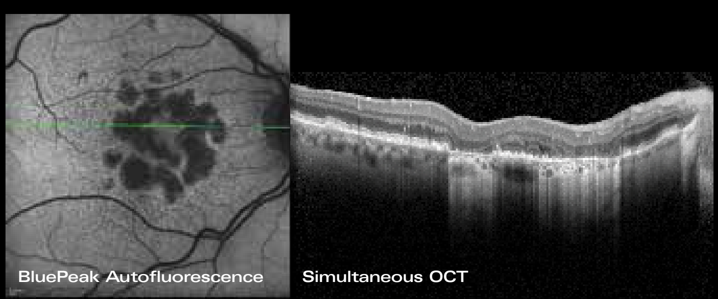

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that each 1mm of decentration of the corneal reflection corresponded to 7° of deviation of the visual axis. Thus if the corneal reflection in the deviated eye is found to be at the limbus (equal to a displacement of approximately 6mm), the deviation is estimated at about 45°.[12] Various subsequent studies re-evaluated the conversion factor and a 1mm displacement of corneal reflex is generally accepted as translating to about 22Δ deviation.[13,14] Figure 3 shows a right esotropia of an estimated 20Δ amplitude. Note that the left corneal reflex is slightly nasally displaced compared to that of the right eye.

It should be noted that angle lambda is usually large in infants, giving the appearance of exotropia but is often counteracted by epicanthal folds seen at this age.[15]

In very young children, uncooperative patients or for those with a markedly reduced visual acuity, this is often the only method of assessing the angle of deviation. Due to the gross nature of this test, it is only suitable for moderate to large manifest deviations. Refinement using the Krimsky technique,[16] requires a prism to be place over the fixing eye of increasing strength until the corneal reflexes are symmetrical. Base out prism should be used for an eso deviation whereas base in prism should be used for an exo deviation. It has been argued that these methods of deviation angle evaluation are much less accurate than a prism cover test, even when conducted by an experienced clinician.[17]

It should be noted that a disadvantage of using a pen torch to produce the corneal reflex is that such a gross target is unlikely to induce accommodation effectively.

Cover test

The cover test is the most commonly performed and arguably the most important test relating to binocular vision and enables the clinician to objectively detect the presence of heterophoria or strabismus and to measure the size of deviation.[18,19] This is a vital component of any optometric examination. The patient should be directed towards a suitable fixation target at 6m for distance and similarly for near. The target chosen should be selected to stimulate fixation and accommodation.

The cover test on infants is not easy. Keeping the infant's attention on a distance target is rarely successful and the introduction of the cover causes further distraction. Covering the eye with the thumb while resting the fingers on the head is sometimes more successful. However, when a baby is uncooperative the clinician may be forced to rely upon the Hirschberg test despite its crudeness.

In the presence of strabismus, on covering the fixing eye, the deviated eye should, if it has some degree of vision, move to uptake fixation. When the cover is removed, the behaviour of the eye underneath the cover can be examined. If the strabismus is alternating, the patient will be able to sustain fixation through the previously deviated eye through to the next blink. When fixation cannot be maintained and fixation reverts to original position, this highlights a fixation preference for the undeviated eye. The clinician will then record which eye manifests the deviation or whether the strabismus alternates, the direction of deviation, and an estimation of the amplitude (in prism dioptres or degrees).

Ideally a cover test with and without refractive correction is advised to assess the effect of refractive correction on angle of deviation.

The inter-observer repeatability of the cover test in childhood esotropia has been investigated and caution should be advised when monitoring change with this method. In children with esotropia with angles >20Δ, differences of 12Δ or more and for smaller strabismus angles between 10-20Δ, 6Δ changes are likely to be genuine. Anything less than these differences, even for experienced clinicians, are likely due to measurement error although smaller change in angles can be indicative of real change.[20]

Figure 3 Child with an estimated 20Δ right esotropia

Prism cover test

The prism cover is an objective way to measure the angle of the deviation, either in the horizontal or vertical plane. The patient should be directed to the appropriate distance target at 6m or 33cm for near. For very young children it is likely that only a near cover test will be possible. An initial alternating cover test is performed to estimate angle and direction of deviation. The direction of the prism base employed is dependent upon the direction of deviation with base out interposed for eso deviations, base in for exo deviations, base down right for right hyper and base down left for left hyper. Using a prism bar, the strength of the prism is gradually increased during alternating cover test until all eye movements are neutralised. To confirm that this is indeed the end point, the power of the prism is increased to check the eye movement is reversed. When there is both a horizontal and vertical component to the deviation, the largest deviation should be measured first and a second prism bar used over the same eye to measure the remaining deviation. Needless to say, when a patient is not cooperative for the cover-uncover or the alternate cover test, it is unlikely that the prism cover test can be meaningfully employed.

Another difficulty with this test occurs with infants because the practitioner will have to invite assistance from the parent or assistant to hold a fixation target while the clinician holds the cover with one hand and the prism bar with the other.

Motor fusion

Prism reflex test

For infants, motor fusion can be demonstrated by interposing a large aperture prism in front of one eye and observing if a fusional movement occurs to restore binocular vision. A 5Δ or 10Δ base out is normally overcome by the cooperative infant of six months.[15]

In young children this test is a convenient objective method for use to assess if there is a presence of binocular single vision and if motor fusion is present. An accommodative near target is chosen and a 20Δ base out prism is placed before each eye separately. The prism shifts the image of the object on the fovea so the eye must move to maintain fixation. By definition of Hering's law, the other eye will move, abducting then adducting to maintain the image of the fovea of this eye. The converse of this sequence should be seen when the prism is removed. If these movements are not observed, this may indicate a lack of binocular single vision and/or abnormal motor fusion.

As with the cover test, the prism reflex test can be very difficult to carry out meaningfully in anything but a cooperative baby or young child. In these circumstances the practitioner may have to rely on the Hirschberg test.

Motility

To determine whether a strabismus is concomitant or incomitant an assessment of ocular motility should be carried out to test the functions of the extraocular muscles. The amplitude of a concomitant strabismus will be unchanged in different directions of gaze.

Stereopsis

The presence and quality of binocular single vision can be established by use of stereotests. In the consulting room the Lang stereo test can be used with occasional success to show the presence of stereopsis in infants from the age of about six months.[21]

Global stereotests use random dots whereas local stereotests make use of contours. An example of a global stereotest for use on slightly older children is the TNO random dot test which depends on complex patterns of displaced random dots to create disparity and therefore depth. The Titmus fly is a local stereotest which uses displaced contoured targets to create disparity and an impression of depth. As local stereotests are prone to monocular clues, global stereotest are considered more accurate as these are truly binocular and therefore can identify strabismus and amblyopia.

Conclusions

Strabismus is a common condition and is a major risk factor for amblyopia. Currently there are three commonly employed tests used to assess eye alignment and diagnose the presence of strabismus. All of these tests require considerable clinical skills.

- The cover test is the most important as it enables diagnosis of the type of strabismus and its amplitude. However, even in skilled hands inter-practitioner reliability with regards to accuracy of strabismus amplitude has been shown to be poor
- Prism fusion tests can be useful but co-operation from the infant or young child is required and a meaningful result may not always be attainable
- The Hirschberg test is probably the easiest test to employ but it is very crude and even if the practitioner can determine accurately a 1mm asymmetry this would detect a strabismus of over about 20Δ. Smaller amplitudes of strabismus will not be detected. It is likely that the conventional Hirschberg test is not very specific or sensitive for strabismus.

Recent developments

A recent innovation is the IRISS Eye Check, a hand held device developed by IRISS Medical Technologies which enables a fully automated Hirschberg test to be carried out and the results displayed in real time within a few seconds. Initial results show a sensitivity of 95 per cent and specificity of 92 per cent for strabismus.[22] Early indications suggest that it is able to detect differences as small as 0.04mm both in terms of corneal reflex asymmetry and pupil size. A further article discussing IRISS Medical Technologies Eye Check will be published in *Optician* next month

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