BrienHoldenVisionInstitute

# STRABISMUS ASSESSMENT

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## AIMS

Chapters 2, 3, 4 and 5 of this module aim to outline the diagnosis and management of strabismus via developing:

- A protocol for assessing the motor aspects of strabismus
- A protocol for assessing the sensory aspects of strabismus
- A framework for the differential diagnosis of strabismus
- Management guidelines for strabismus

## LEARNING OUTCOMES

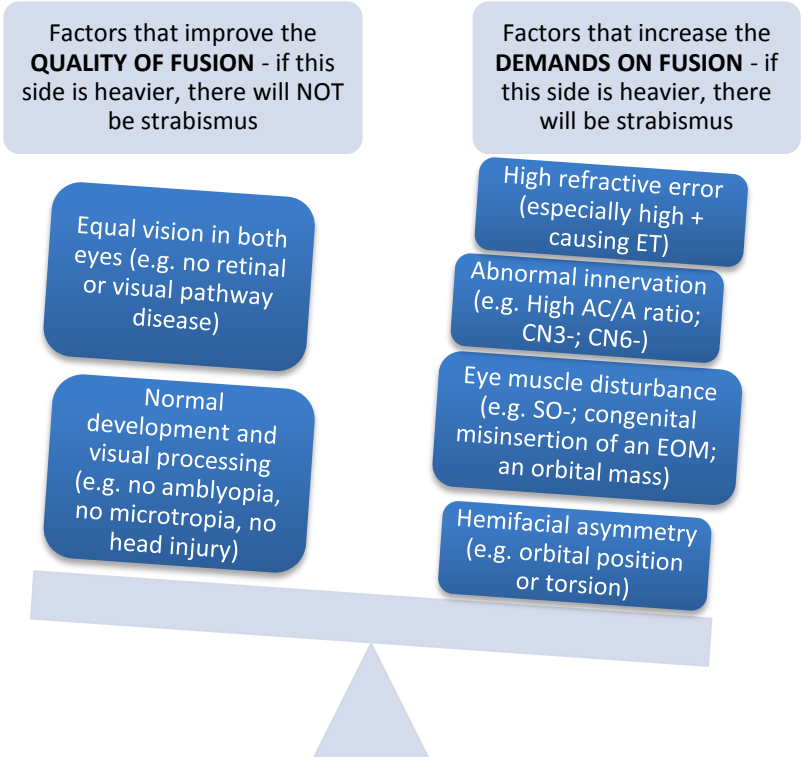
After completing chapters 2, 3, 4 and 5 of this module, students should be able to:

- Conduct a diagnostic assessment of the motor aspects of strabismus
- Conduct a diagnostic assessment of the sensory aspects of strabismus
- Differentially diagnose common strabismus
- Identify and appropriately refer potentially dangerous strabismus
- Manage common strabismus or aspects of strabismus that fit within their legal scope of practice.

## BACKGROUND

Strabismus (eye turn) and amblyopia (lazy eye) are common conditions, which often present to eye care practitioners for diagnosis, advice and management.

Normal single binocular vision occurs when the two monocular images are fused - motor vergence must be used to fuse images falling on non-corresponding points of the retina.Strabismus develops due to an imbalance of two factors; quality of fusion and demands on fusion.




The concept of supply and demand of fusional vergence supports this point. If the demand is more or the supply is less, both will lead to strabismus.

Strabismus can be described in terms of:

1) **Direction** – “exotropia ” means one of the eyes turns out relative to the point of fixation, “esotropia” means one eye turns in relative to the point of fixation, and “hypertropia” means one eye turns up relative to the other eye (can also be referred to as “hypotropia” in the other eye);

2) **Frequency** – “intermittent” means it is sometimes there and sometimes not, “constant” means it is always there;

3) **Magnitude** – the “size” of the strabismus (measured in prism dioptres or degrees);

4) **Laterality** – a “unilateral” strabismus will always be in either the right or left eye, whereas an “alternating” strabismus will change eyes;

5) **Comitancy (of direction)** – “comitant” means it is the same wherever a person looks, “incomitant” means it changes in size or direction depending on where a person looks;

6) **Comitancy (of distance)** – describes whether the strabismus changes with distance of observation.

Eye care practitioners need to use these descriptions to define the nature of a strabismus. Additionally, determining *cause* is critical to choosing an informed course of management for some types of strabismus.

An eye care practitioner may manage the strabismus themselves if they:

- can describe a strabismus
- can complete other necessary assessments
- can define a specific cause of strabismus by integrating assessments with history (e.g. when/how did the eye turn start) and associations (e.g. are there any central nervous system abnormalities)
- At least prove that the strabismus is free of sinister ocular, neurological or systemic causes,

When this is not the case, an eye care practitoner should refer for specific management or for aetiological diagnosis (i.e. to establish the cause of the strabismus, either to aid management or to prove the absence of sinister causes).

# CLINICAL EVALUATION OF STRABISMUS

Strabismus can be caused by an abnormality of sensory fusion, or motor control, or both. It is essential to be able to describe both sensory and motor aspects of the strabismus, and also be able to integrate these with case history and associations in order to establish the cause (aetiology) of strabismus whenever possible.

## DETAILED HISTORY OF STRABISMUS

### Symptoms

- Asthenopia - may imply difficulty controlling a large phoria (uncommon in children) or a conflict between accommodative and vergence demands upon prolonged near work (common with extensive computer usage)
- Diplopia - indicates recent AND sudden onset of a strabismus without development of suppression (uncommon in children)
- Cosmesis – patient's eyes "turn out, "turn in", "turn up", or "look funny" sometimes/ often/ always (this is the most common reason for parents to bring their children for an eye examination regarding strabismus)
- No symptoms – some people/families are unobservant; some accept strabismus as fate and/or otherwise unfixable so don't seek care until something else is wrong

### Onset and nature

- Is the strabismus constant or intermittent?
- How long has the strabismus been noticed?
- If intermittent, how often? when? while doing what?
- Type of deviation?

### Possible Aetiology (cause)

- Trauma - includes birth trauma
- Ischemia, stroke – includes birth hypoxia
- Neoplasms – includes orbital, and those affecting CN 3, 4 or 6 on either side
- Febrile illness
- Prematurity and low birth weight are predisposing factors to strabismus
- Heredity - history of strabismus or lazy eye among blood relatives
- Consecutive (due to previous treatment) - when, type, for how long, successful or not?

## MOTOR ASPECTS OF STRABISMUS

Clinical evaluation of the motor aspects of strabismus is generally performed in six parts. A manifest deviation of the visual axes of the two eyes needs to be **detected** (this can be harder than it sounds), then it needs to be analysed for **direction**, **magnitude**, **laterality**, **comitancy**, and **constancy**. The tests for each part are essentially simple, but constructing the aetiological diagnosis from the results can be difficult.

### A) Detection of the deviation

#### General inspection

- Take note of the apparent eye alignment/misalignment; head posture; lid fissures

#### Brückner's Method of inspection

- Shine a direct ophthalmoscope light on both eyes at once, look through the ophthalmoscope, focus on the patient's face, assess the relative brightness of the red reflex from each eye
- A strabismic eye tends to have a brighter reflex, while the fixating eye tends to have a darker reflex (because the macula is the darkest area of the ocular fundus)
- The difference is most marked on young children, and less pigmented people

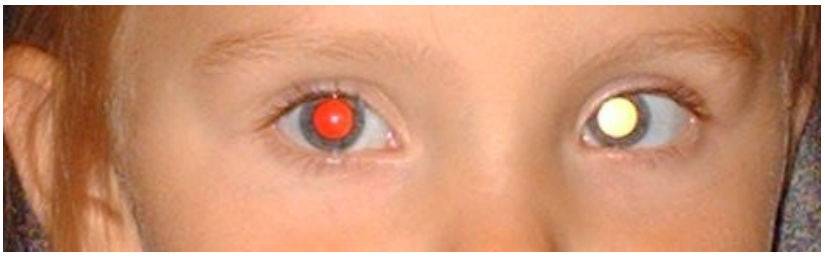


***Figure 12.1:*** *Brückner's Method of inspection*

#### Hirschberg test inspection

- The Hirschberg test is useful for detected lateral deviations of the eyes.
- Hold a penlight in front of your preferred eye, close your non-preferred eye, shine the light between your patient's eyes, assess the symmetry of position of the corneal reflexes in the right and left eyes
- The corneal reflex will be positioned slightly nasal to pupil centre in most fixating eyes, but shifted somewhere else (see sections on direction and magnitude to determine where) in a strabismic eye

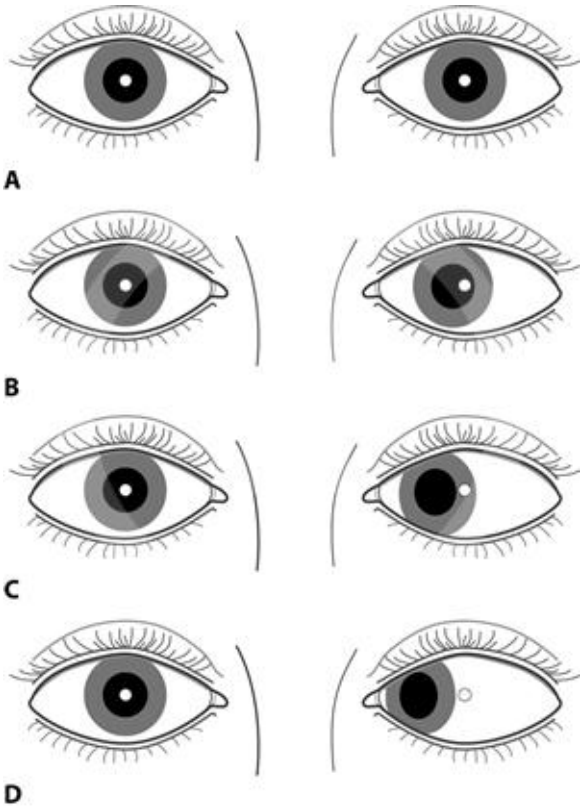



***Figure 12.2:*** *Hirschberg test: A: Straight eyes. B – D: Left esotropia of increasing magnitude (left corneal reflex is displaced laterally). Each 1mm deviation of the reflex indicates approximately 22$^{\Delta}$ of eye misalignment.*

Cover Test

- Perform at distance and near, as previously taught
- Note whether the deviation is always manifest or is latent – i.e. differentiate phoria from tropia – using the unilateral cover test
- Note the quality of fixation, and the speed/accuracy of refixation

Four Diopter (Base Out) Prism Test (used to detect monofixation syndrome ≈ small-angle strabismus ≈ micro-strabismus)

- Note that small-angle strabismus is a sensory adaptation
- Patient fixates a penlight at 40cm; examiner introduces a 4pd (Δ) BO loose prism from the temporal side into the spectacle plane of the eye suspected of having foveal suppression. If there is foveal suppression, the fellow eye will not move. If there is bifoveal fusion without suppression, the fellow eye will make a small corrective movement.
    - Note that the fusional vergence movement expected is quite subtle as the induced vergence demand is very small.

**B) Direction of the deviation**

Note whether the strabismus is an esotropia, an exotropia, a right hyper-/left hypo-tropia, or a right hypo-/left hyper-tropia

- Use general inspection, Hirschberg, and alternating cover test

- The Hirschberg reflex will be shifted nasally in an exotropic eye
- The Hirschberg reflex will be shifted temporally in an esotropic eye
- The Hirschberg reflex will be shifted downwards in a hypertropic eye

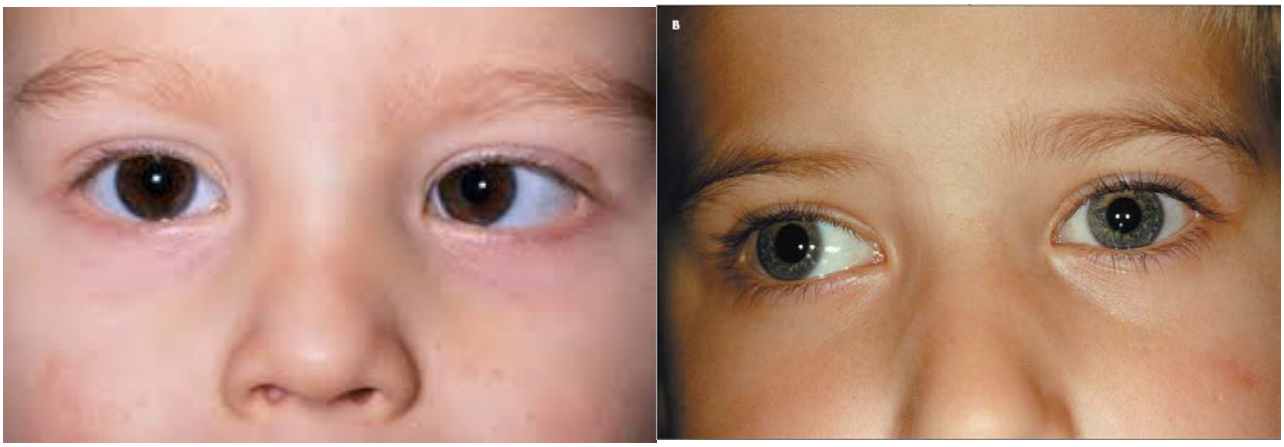


***Figure 12.3: (****Left): left esotropia (corneal reflex displaced laterally) (Right): The right eye of this child seems to have a combination of horizontal and vertical deviation as evidenced by the nasal and downward deviation of the Hirschberg reflex.*

**C) Magnitude of the deviation**

Can be defined as an angle (the degrees away from fixation), but more commonly defined as the amount of prism to realign a strabismic eye (in prism diopters = pd, $\Delta$)

Hirschberg test estimation (able to differentiate small, medium and large strabismus, i.e. $\pm$20pd). 1mm movement of the Purkinje image from the pupil center corresponds to ~12 degrees (or 22PD of eye deviation)

- In a small(ish) strabismus, the Hirschberg reflex will remain inside the pupil area
- In a medium-sized strabismus, the Hirschberg reflex will be around the pupil margin
- In a large strabismus, the Hirschberg reflex will be outside the pupil area

Krimsky test estimation (accurate to within $\pm$10pd)

- This variation of Hirschberg's Test increases the accuracy of magnitude measurements. The patient fixates a penlight held at 30cm. Prism is added in front of the strabismic eye until the corneal reflexes are symmetrically placed on the 2 eyes. Base direction is as per relieving prisms.
- In the **Modified Krimsky Test,** the patient fixates a pen light with the better eye and prisms are added into front of the better *eye* until the reflex is central in the deviated eye.

Neutralising cover test with prism bar (accurate to within $\pm$5pd)

- This is the most useful test for measuring the magnitude of strabismus – be good at it!
- Perform the alternating cover test, place prism in the spectacle plane of the strabismic eye (using a prism bar), change the power until there is no movement as you alternate your cover
- Base direction is as per relief prism (e.g. base in for exotropia, base out for esotropia)

- Sometimes neutralisation is marked by “equal and opposite movements” rather than “no movement”

Maddox Rod (accurate to within ±3pd for vertical deviations, but lack of accommodation control can make it variable for horizontal strabismus, particularly esotropia)

- Place a Maddox Rod in the spectacle plane of the strabismic eye, ask the patient to fixate a pen torch and tell you where the red line (seen by the eye looking through the Maddox Rod) is compared to the white light (seen by the other eye), then use prisms to place the red line and the white light together
    - Torsional deviations due to vertical recti or oblique muscle palsies could also be picked up from the orientation of the Maddox rod with respect to the vertical plane.
- Requires subjective responses ⇒ only useful in acquired, adult strabismus (children, or adults with longstanding strabismus are likely to suppress)

Hess-Lancaster Screen

- More cumbersome than Maddox Rod, without providing more information ⇒ historical use only

A note on magnitude of deviation pre- and post-cycloplegia

- In some esotropia cases, measurement of the magnitude of deviation pre- and post-cycloplegia can assist the clinician to distinguish between accommodative and non-accommodative esotropias. The idea is that as cycloplegia stops accommodation, it will decrease accommodative esotropia but have no effect on the angle of non-accommodative esotropia. The ambiguity in this is that cycloplegia stops accommodation at the end organ (the ciliary body); it does not necessarily change accommodative “drive” at the point of generation (brain). The accommodative drive is what feeds into the vergence system, not the action of the ciliary body.
- Hence, cycloplegia has an unpredictable effect on magnitude of deviation in accommodative esotropia
    - Sometimes it reduces because without any ciliary muscle outcome, the accommodative drive decreases, and so accommodative convergence decreases
    - Sometimes it increases because without any ciliary muscle outcome, the person tries harder to accommodate, i.e. the accommodative drive increases, and so accommodative convergence increases.

**D) Laterality of the deviation**

Use the unilateral cover test to determine which eye is deviated

- Is the strabismus right unilateral, left unilateral, R/alternating, L/alternating, or freely alternating?

## E) Comitancy of deviation

**Use ocular motilities for a *dynamic assessment of comitancy***

- Watch for incomitancies as you perform motilities– know how to use motilities to test all EOM pairs
- Incomitancies may be caused by muscle underactions, muscle overactions, muscle limitations, nerve palsies, or orbital factors (e.g. swellings in thyroid eye disease, neoplasms, shape factors such as orbital torsion)
- Use the UC Davis eye simulation web page until you can understand and recognize all common conditions
- Always note quality of pursuits when performing ocular motilities (particularly look for monocular naso-temporal smooth pursuit asymmetry (MNTSPA))
- Use near point of convergence to identify distance-near incomitancies

**Use objective cover testing (or subjective Maddox Rod) in 11 positions of gaze for a *static assessment of comitancy***

- Complete assessment requires measurement in nine cardinal directions of gaze, PLUS head tilt left AND head tilt right. However, the following abridged combinations are the most important for particular clinical entities:
    - Up/Primary/Down to describe alphabet patterns

      V-pattern describes an esotropia that is largest in down-gaze and smallest in up-gaze, OR an exotropia that is smallest in down-gaze and largest in up-gaze. The most commonly observed alphabet pattern is the V-pattern caused by oblique muscle dysfunction (inferior oblique-over action (IOOA) with superior oblique-under action(SOUA)) that often follows longstanding esotropia (described as V-pattern ET with IOOA/SOUA complex)

      A-pattern describes an exotropia that is largest in down-gaze and smallest in up-gaze, OR an esotropia that is smallest in down-gaze and largest in up-gaze. A-patterns can result from superior oblique-over action (SOOA) with inferior oblique-under action (IOUA) following longstanding esotropia (described as A-pattern ET with SOOA/IOUA complex)

      What would an X-pattern, a Y-pattern, and a $\lambda$-pattern look like?
    - Left/Primary/Right to describe CN6 palsies and Duane's syndromes
    - Park's Three-Step Technique to identify a paretic vertical muscle - this will most commonly be CN4 palsy – see Appendix 1 for description of Park's Three-Step test.

## F) Constancy of the deviation

Use the unilateral cover test to note whether the strabismus is constant or intermittent

- Can you encourage your patient to achieve bifoveal fixation under any circumstance?

## MONOCULAR SENSORY ASPECTS OF STRABISMUS

Clinical evaluation of the monocular sensory aspects of strabismus generally considers three areas: vision, fixation, and accommodation.

### A) Vision

Assess monocular vision and visual acuity using a developmentally appropriate test

- Contour interaction is increased in amblyopia – this causes a crowding effect, which often results in “single symbol acuity” being different than “line acuity” or “logMAR acuity”. These differences in acuity are strongly suggestive that any vision reduction is due to amblyopia rather than eye disease
- Record single letter AND logMAR acuities – note the significance of any difference
- Note mis-counting and mis-ordering of letters/symbols on a line
- Note the pattern of letter misrecognition on a logMAR chart

### B) Fixation

Monocular fixation in amblyopia can be either central or eccentric, and steady or unsteady. These qualities can be measured using any of the following four tests. Clinically, the visuoscope is by far the most valuable of these tests.

i. **Visuoscope**: this is an ophthalmoscope with a target inserted in the illumination system. This target is imaged at infinity by the condensing system of the ophthalmoscope.
   - Start by asking the patient look at a point in the room, find a known spot somewhere in the fundus (e.g. the optic nerve head), focus your view, then ask the patient to “look at the target NOW”
   - The test is most accurately performed through a dilated pupil with the other eye occluded, but can be done is less ideal conditions. This will ensure that the view of the fundus is not precluded by pupil miosis when the subjects fixate on the ophthalmoscope’s light.
   - The target (depending on the manufacturer of the ophthalmoscope) will either be a star with two concentric rings, or a circle with a graticule
   - The examiner notes the position of the foveal reflex in relation to the target that is projected on the fundus
     - Fixation is central if the foveal reflex is in the centre of the target, or eccentric if the foveal reflex is somewhere else
     - Fixation is steady (i.e. one retinal point is used for fixation) if the foveal reflex stays on the same point of the target, or unsteady (i.e. an area of the retina is used for fixation) if it moves about
   - Note that illumination of the visuoscope should be kept low (or green) so the patient can see the target
   - Visuoscopy takes practice: it is worth using your visuoscope on several normal and as many abnormal subjects as possible, and drawing or making brief notes on your observations.

ii. **Haidinger's Brushes**: these are of little more than historical significance. Visuoscopy is a simpler, faster and more direct measure of fixation.

iii. **Macula Integrity Tester**: this utilizes the phenomenon of Haidinger's Brushes. When a rotating polaroid filter is viewed through a deep blue filter this gives rise to the sensation of a rotating propeller. The perception of this propeller is the result of polarization of light by Henle's fibre layer at the fovea. The position of the propeller must coincide with the position of the fovea because of its anatomical structure. If the propeller position coincides with fixation, fixation is central. If not, fixation is eccentric. Failure to appreciate the brushes shows an inhibition of macular function.

iv. **After Image Transfer (AIT)**: This test assumes normal correspondence (i.e. that both foveae give rise to the same visual direction). The fovea of the non-amblyopic eye is tagged monocularly with an afterimage (AI). The non-amblyopic eye is then occluded and the patient is directed to look at a fixation point with the amblyopic eye. Central fixation is represented by the AI being superimposed on the fixation point. Eccentric fixation is represented by displacement between the AI and the fixation point.

### C) Accommodation Response

The functional deficit of amblyopia includes decreased amplitude of accommodation and increased lag of accommodation. This is best quantified using MEM retinoscopy (refer to Accommodation-Vergence notes for details). In the case of strabismus and amblyopia, it is most important to note the differences between the right and left eyes.

The causative role of accommodation in accommodative esotropia, either in achieving clear vision for a hyperope, or when there is a high AC/A ratio, or both, is also worth consideration.

- Some patients with accommodative esotropia can choose to either relax their accommodation to straighten their esotropia but blur their vision, or accommodate to clear their vision but manifest their esotropia – you can relate changes in accommodation (monitored with retinoscopy) with changes in their angle of esotropia (monitored with cover test or any of the other methods discussed in the section on magnitude of deviation)

- Some patients with accommodative esotropia lack this kind of conscious control of their accommodation – they might present with habitual accommodative lead or lag.

## BINOCULAR SENSORY ASPECTS OF STRABISMUS

Clinical evaluation of the binocular sensory aspects of strabismus generally considers three areas: sensory fusion, stereopsis (technically high level use of fusion), and binocular correspondence.

### A) Status of Sensory Fusion

Having diagnosed a strabismus, the next step is to determine whether fusion is possible. There are 4 basic levels of fusion - superimposition, flat fusion, contour stereopsis and random-dot stereopsis.

Central suppression indicates lack of central fusion (although peripheral fusion may cause superimposition). Diplopia indicates lack of fusion and no suppression.

There are many possible tests for suppression. Each of these tests uses a different level of dissociation to enable the level of suppression can be assessed. The more the testing method mimics natural seeing conditions, the more likely suppression will be found. Tests with minimal dissociation are more likely to detect suppression.

i. **Worth Four Dot Test**: consists of four lights in a diamond pattern, two green, one red, and one white. The patient wears red/green glasses so that the white light is visible to both eyes, the green light is visible through the green filter and the red lights are visible through the red filter. For a strabismic patient, four responses are possible. If five lights are seen, diplopia is present. If four lights are seen, but there is a manifest strabismus to cover test, anomalous correspondence is present. If three green lights are seen, the eye behind the red filter is being suppressed. If two red lights are seen, the eye behind the green filter is being suppressed.

   Remember that the perception of coloured lights through the filters differs from the perception of coloured targets on a white background.A red light is transmitted through a red filter, not through a green filter. A red target on a white page seen through a green filter appears black, because the red is not transmitted through the green filter. A green target seen through a green filter ‘disappears’ because the white background also appears green.

ii. **Polaroid suppression check**: Some distance visual acuity charts (e.g. AO projector charts) have a suppression check. When the AO Projector chart is viewed through polaroid filters, one letter from a line is seen by both eyes, two letters are seen only by the left eye and two letters are seen only by the right eye.

   All the commonly used polaroid stereotests have suppression checks (usually the “R” and the “L”).

iii. **Red/green bar readers**: Checks for near suppression, but beware of alternating suppression.

iv. **Brock string**: this can be performed in free-space or with red/green filters.

v. **Mirror polar test**: when a patient wears polarizing glasses and looks in a mirror, only the right eye can see the right eye, and only the left eye can see the left eye: i.e. the right eye is not visible through the left lens and vice versa.

   Put on a pair of polaroid lenses and look at yourself in a mirror. Close one eye and see what happens.

   A strabismic patient, when suppressing, will see only one eye. The strabismic eye will appear blacked out.

vi. **Coloured Filters**: when a patient with strabismus views a penlight through red and green glasses in a darkened room, she or he should see two lights, one red and the other green. When suppression is present, only a red OR a green light is seen.

### B) Presence and Level of Stereopsis

Stereopsis is the perception of 3-dimensional space that arises from binocular disparity cues only. Disparity can be presented via contour stereograms (where two dimensional shapes can be seen with each eye individually, but a three-dimensional perception is possible when the disparity information between right and left eyes is available), or via random-dot stereograms (where no regular shapes can be seen with the right or left eye individually, but a three-dimensional perception is possible when the disparity information between right and left eyes is available). Random-dot stereopsis places a higher requirement on fusion than contour stereopsis does. Random-dot stereopsis is therefore better

at detecting strabismus; however, contour stereopsis may be useful in monitoring treatment of strabismus.

There are a number of stereotests designed to measure random-dot and contour stereopsis. The most clinically useful are hand-held tests such as the Randot. However, synoptophores are historically interesting as they allow disparity information to be presented at the angle of a strabismus. This provides information on the sensory changes in a strabismus with an uncorrectable motor anomaly. This information has little clinical relevance, and so synoptophores are seldom used.

Examine the available tests of contour stereopsis (e.g. the Titmus stereo fly, animals and Wirt circles; the animals and modified Wirt circles of the Randot test), and random-dot stereopsis (e.g. the Lang stereotest; the Randot shapes). Note that random-dot tests usually present a pass-fail criterion, rather than a facility for thresholding to a stereoacuity like the contour tests. This is consistent with the respective function of each type of stereotest – random-dot stereotests detect strabismus, contour stereotests allow monitoring of treatment progress. Note that routinely available clinical stereotests are calibrated only for one viewing distance and one inter-pupillary distance.

***Figure 4**: Randot and contour stereotests, Worth 4-dot test (below to the left)*

**C) Status of Binocular Correspondence**

Strabismus can cause a restructuring of the way information from one eye relates to information from the other eye. The restructuring is called anomalous retinal correspondence (ARC). ARC may be reversible if the condition that caused it is treated.

The level of dissociation produced by tests of binocular correspondence varies. The more the test mimics the real world, the more likely that ARC will be evident. The depth of ARC is not the same in all patients. The longer the ARC has been present, the better established it becomes.

i. **Worth Four-Dot**: Fusion on the W4D (perception of 4 dots under binocular conditions with the red/green filters in place) in the presence of a manifest strabismus (diagnosed on unilateral cover test) implies ARC.

ii. **Bagolini Lenses**: these lenses have fine striations on their surface that diffract light, meaning that when a point source of light is viewed through one lens, a streak of light is seen passing through the light source.

To perform the test, the patient wears Bagolini lenses in the spectacle plane of both eyes, one with striations at 45 degrees and one with striations at 135 degrees (Bagolini frames can be bought in this arrangement). The patient then fixates a penlight held at 33 cm in a normally lit room. When a manifest strabismus is present, three responses are possible:

- *Suppression* – only one streak is seen going through the light
- *Normal correspondence* (without suppression) – two streaks are seen but they are separated
- *Anomalous Correspondence* – two streaks are seen crossing through the light source

# REFERENCES

## GENERAL REFERENCES

von Noorden GK, Campos EC. Binocular Vision and Ocular Motility (6th ed). Mosby: St Louis 2001. This is available for free download through ORBIS (http://telemedicine.orbis.org/bins/content_page.asp?cid=1-2193) - it is long and complicated (don't expect to understand all of it on the first reading) but it is a useful reference book to have as problems present.

Pratt-Johnson JA, Tillson G. Management of Strabismus and Amblyopia. Thieme, 2001

# APPENDIX 1 - PARKS THREE-STEP METHOD

The Parks three-step method is used for differentiating between palsies of the four vertically-acting EOMs, (when a vertical deviation is present).

For simplicity the technique only considers OD so treat L hyper as R hypo or L hypo as R hyper.

Use cover test or Maddox rod to measure the deviation.

- A muscle crossed out 3 times => paretic muscle in OD
- A muscle not crossed out at all => paretic muscle in OS

## STEP 1

If hypo cross out elevators (SR and IO)

If hyper cross out depressors (IR and SO)

## STEP 2

If deviation increases on R gaze cross out SR and IR

If deviation increases on L gaze cross out IO and SO

## STEP 3

If deviation increases on tilting head to right hand side cross out SR and SO

If deviation increases on tilting head to left hand side cross out IO and IR

## EXAMPLE:

1. **In the primary position is the deviation R/L or L/R?** ANS: R/L => cross out IR, SO
2. **Does the vertical deviation increase on left or right gaze?** ANS: left gaze=> cross out IO, SO
3. **Does the vertical deviation increase on head tilt to left or right shoulder?** ANS: to Right=> cross out SR, SO
4. As the SO has been crossed out three times: the defective muscle = RSO