

## Approach to Strabismus:

By John Hilhorst

### 1. Definitions

Strabismus is an anomaly of ocular alignment that can occur in any direction. It is characterized by a misalignment of one or both eyes that may turn inward/nasally (**eso-**), outward/temporally (**exo-**), upward (**hyper-**) or downward (**hypo-**). Further terms used to describe strabismus depend on when the condition is present and whether it changes with positions of gaze. A **latent** strabismus is present only when fixation is interrupted while a **manifest** strabismus is present without interruption of the visual axis. The terms "**phoria**" and "**tropia**" are used to describe latent and manifest strabismus respectively. Manifest strabismus is a visual defect that can be either constant or intermittent occurring for example only when the individual is tired. It can also involve only one eye or alternate eyes. **Comitant** is used to describe a deviation that is the same in all positions while **incomitant** strabismus is a deviation that changes with position.

### 2. General Presentation

In children, strabismus is a common condition but it can also occur in teenagers and adults. Strabismus is found to be equally established in males and females, and may be genetically influenced. Individuals who have strabismus experience a significant reduction in vision. Normally, children can focus on the same image allowing the brain to combine the two images from each eye to produce a three-dimensional image and thus the ability to perceive depth. By four months of age an infant's control of his or her eye movement should be completely aligned. Conversely, a child with strabismus will have an eye that is misaligned creating a discrepancy in the visual stimuli to the brain. The brain will ignore the image of the misaligned eye and focus on the eye that is straight resulting in a loss of depth perception.

### 2. Common etiologies

It is difficult to establish the exact cause of many cases of strabismus however many conditions that predispose children to developing strabismus have been identified. The development of strabismus is multifactorial with both genetic and environmental influences., Congenital and acquired forms of strabismus exist and it is very important to distinguish between the two as some of the acquired forms can be life or vision threatening. It has appeared to be most common in individuals with cerebral palsy, down syndrome, hydrocephalus, brain tumors and prematurity. Cataracts and eye injuries have also been correlated with individuals diagnosed with strabismus. The common risk factors associated with strabismus and children involve:

**Table 1: Risk factors for developing Strabismus:**

- Low birth weight (<1250 g), particularly premature infants who have developed retinopathy of prematurity
- Family history of strabismus
- Neuromuscular disorders (ie multiple sclerosis, myasthenia gravis, botulism),
- Congenital ocular abnormality
- Tumours of the brain or eye (ie retinoblastoma)
- Cataracts
- Head injury
- Infections (ie meningitis, encephalitis, measles)
- Systemic conditions with vision-threatening ocular manifestations (ie pauciarticular juvenile rheumatoid arthritis, which can predispose to iritis and cataracts)
- Drugs and toxins (ie lead and heavy metals)

Adapted from Weinstock et al. Screening for Childhood Strabismus by Primary Care Physicians (1998) and UpToDate 2008..

### **3. Questions to Ask**

All children should be screened for strabismus as it is essential to prevent visual and psychosocial dysfunction. High risk children should be referred to an ophthalmologist for a screening examination. It is very important to detect children with manifest strabismus as left untreated these children have a greater chance of

developing amblyopia. Amblyopia, also known as “lazy eye” is a functional reduction in visual acuity caused visual dysfunction during the critical period of eye development.

Questions to consider when screening a child for strabismus include:

- a. Onset of eye deviation? Acute vs. chronic? Has it been present since early infancy?
- b. When is the deviation present? Is it constant or intermittent? Does it occur with only certain positions of gaze?
- c. Is the deviation monocular, binocular or alternating?
- d. Associated symptoms? - (ex. headache for intracranial masses, any double vision?)
- e. Is there a family history of myopia, strabismus or amblyopia?
- f. Is there a history of toxin or medication exposure?
- g. Is there history of trauma?
- h. What is the age of the child? Refer to Table 1. for eye examinations.
- i. Birth and past medical history?

#### **4. Differential Diagnosis**

##### **- Primary Strabismus:**

##### **a) Esodeviations**

- Defined as convergent visual axes or “crossed” eyes
- Nasal deviation relative to the fixating eye
- Amblyopia and monocular blindness usually manifests as esodeviation in children 0–3 years of age
- Includes accommodative esotropia, idiopathic infantile esotropia, and abducens nerve palsies

##### **b) Exodeviations**

- Defined as divergent visual gaze
- Temporal deviation relative to the fixating eye

- Amblyopia and monocular blindness usually manifest as exodeviation in children older than 4 years
- Includes intermittent exotropia and oculomotor nerve palsies

**c) Ocular Instability of Infancy**

- Unsteady ocular alignment that may be present in normal newborns during the first few months of life

**d) Congenital esotropia**

- Pronounced medial deviation of one eye in the first year of life
- Occurs in otherwise healthy infants

**e) Accommodative strabismus**

- Result of visual discrepancy and favored use of the better eye
- Also occurs during accommodation when there is significant hyperopia
- The eye not in use is esotropic

**f) Idiopathic childhood exotropia**

- May be alternating, or may be secondary to visual discrepancy and favored use of the better eye
- The eye that is not fixed on an object is exotropic

**g) Congenital Cranial Nerve III palsy**

- Familial, usually unilateral, ptosis

**h) Strabismus Associated with Syndromes**

- Includes Angelman's, Down's, Prader-Willi, Cri-du-chat, Noonan's, Marfans, and others

**Secondary Strabismus:**

**a) Retinoblastoma**

- Frequent early finding in retinoblastoma
- May be due to visual impairment or space- occupying lesion

**b) CNS etiologies**

- Associated with hydrocephalus and periventricular leukomalacia, especially in premature infants
- Associated with increased intracranial pressure and intracranial masses due to cranial nerve VI compression
- Cranial nerve palsies and variable limb anomalies
- Head trauma with nerve palsies or orbital fractures
- CNS infections (meningitis, encephalitis, orbital cellulitis, measles)

**c) Neuromuscular disorders**

- Includes Guillan Barre Syndrome, Myasthenia Gravis, Multiple Sclerosis, Botulinism

**d) Graves disease (thyrotoxicosis)**

**e) Drugs/Toxins**

- Lead and Heavy metals

**Pseudostrabismus:**

- Pseudoesotropia is most common
- It is an apparent esotropia that may occur in the first year of life in children with wide-spaced eyes, flat nasal bridge, or prominent epicanthal folds; it is not a true strabismus, but rather an optical illusion
- Pseudoexotropia may also occur; this may be caused by retinopathy of prematurity and other vascular disorders

**5. Physical Examination**

The goal for examination of strabismus is to develop as much knowledge about the diagnosis without physically touching the child. Toys or detailed flashy objects can be used to entertain the child while observing eye alignment and movement. Recall high risk children should be referred to an ophthalmologist. Always remember to perform an assessment of general health and neurologic status in addition to the ophthalmologic exam. Examination of the eyes should assess visual function, pupillary reactivity, eyelid position, and extraocular movements in addition to special tests. Adhere to the following table, adapted from Wright et al. (2006), to administer the proper order of examination:

Table 2:

Order of Examination
1. Inspection: Evaluation and measurement of face turns
2. Amblyopia assessment / visual acuity
3. Sensory tests
4. Ductions and versions
5. Measurement of deviation

Adapted from Wright et al. Handbook of Pediatric Strabismus and Amblyopia, 2nd edition. (2006)

### **1. Inspection:**

- Do the eyes appear straight?
- Is their face turn or head posturing?
- The presence of straight eyes with a face turn in a patient who has strabismus can indicate the presence of binocular fusion.

### **2. Amblyopia assessment / Visual acuity:**

- Amblyopia can be evaluated by using linear acuity. In younger children single optotype testing is quicker, easier and more accurate than linear. There are many tests that can be administered in regards to visual acuity. Some of the best examples for young children include: wright figures, Allen picture cards, HOTV and the illiterate E game.

### **3. Sensory tests:**

- Sensory tests should be a part of every strabismus examination. It should incorporate a haploscopic fusion/suppression test (e.g. Worth 4-dot test) and a test for stereo-acuity (e.g. Titmus).

### **4. Ductions and Versions:**

- Ductions test monocular movement and involve occluding one eye and forcing fixation to the eye being examined. They evaluate the ability of the eye to move into extreme fields of gaze.
- Versions test binocular movement and involve observing how the eyes move together. The key is to look for imbalances of eye movements and oblique muscle dysfunction missed on ductions. Version tests should include eye movements through nine cardinal positions of gaze: from primary position to straight right, straight left, straight up, straight down, up to the right, up to the left, down to the right, and down to the left. Unordinary versions should be described with a scale of +4 to -4, with 0 being normal and +4 indicating the maximum over-action and -4 indicating severe under-action.

### **5. Measurement of Deviation:**

- You can measure deviation using the following methods:
  - Light reflex tests

- Cover tests

- **Light reflex tests:**

- **Hirschberg Test** or corneal light reflex test assesses eye alignment by observing the location of the corneal light reflex within the pupil. It should be administered by directing a light into the patient's eyes and having the patient look directly into the light to assess the location of the light reflex in each eye. Individuals who are diagnosed with strabismus will have an eccentric light reflex in the deviated eye. Temporal displacement of the light reflex indicates esotropia, nasal displacement indicates exotropia, and inferior displacement indicates hypertropia.

- **Krimsky Test** - Is an adaptation of the Hirschberg test but involves the addition of a prism to measure strabismus in patients who are uncooperative or whom have worse than 20/400 vision. Place the prism in front of one eye, with the base oriented appropriately (esotropia, base out; exotropia, base in; hypertropia, base down) to standardize the deviation. Shine a penlight into both eyes, as done so in the Hirschberg test, and direct the patient to fixate on a target (toy, picture etc.). Placing a prism over the fixing eye in a patient with tropia will cause a version movement in which both eyes move in the direction of the apex of the prism, which moves the light reflex in the deviated eye. Placing the prism over the non-fixing eye directly moves the light reflex to the center of the pupil without a version shift.

- **Bruckner Reflex Test** - Is a test performed by using the direct ophthalmoscope to obtain a red reflex from both eyes simultaneously. Patients who are diagnosed with strabismus will show asymmetrical reflexes with a brighter reflex coming from the deviated eye, reflecting more light in the deviated eye.

- **Cover and Cover/Uncover Tests**

- The cover test is one of the most important tests in detecting strabismus

- The Cover and Cover/Uncover tests assist in determining the binocular relationship of the eyes. When performing the cover test, ask the child to fixate on a distinct object (detailed picture, toy etc.) while covering one eye and observing the movement of the uncovered eye.. In a normal child, no movement in the uncovered eye will be observed. Manifest strabismus is present if the uncovered eye moves to refixate on the target.
- The Cover/Uncover test is performed to detect latent strabismus and does not need to be done if the cover test has already revealed manifest strabismus. Again the child is asked to visually fixate on an object, near or far. One eye is covered for a few seconds and then rapidly removed. This time, the movement of the eye that was under the cover is observed. If the previously covered eye shifts back into position to refixate as it deviated while covered, the patient is said to have latent strabismus. By observing the direction of refixation of each eye, you will be able to determine the problem and measure the deviation using prisms
- Be aware that most prism and cover measurements cannot be used on infants or very young children as it requires them to be able to fixate accurately on a object, and remain still long enough to get a proper evaluation.

The following table, adapted from Weinstock et al. (1998), displays the general exams that should be assessed to determine eye disorders at various ages:

**Table 3:**

Age	Eye Screening by Primary Care Physician
Neonate	<ul style="list-style-type: none"> <li>- External examination</li> <li>- Ocular alignment</li> <li>- Corneal light reflex test</li> <li>- Ophthalmoscopy</li> <li>- Clear media</li> <li>- Normal red reflex</li> </ul>



Age	Eye Screening by Primary Care Physician
6 months	<ul style="list-style-type: none"> <li>- Above tests, plus:</li> <li>- Ocular motility</li> <li>- Fix upon and follow a small toy</li> <li>- Pupillary reactions</li> </ul>
3 Years	<ul style="list-style-type: none"> <li>- Above tests, plus:</li> <li>- Visual acuity</li> <li>- Sheridan Gardner (single letter matching game) or picture chart</li> <li>- Cover test</li> </ul>
5-6 years	<ul style="list-style-type: none"> <li>- Above tests, plus:</li> <li>- Visual acuity.</li> </ul>

Adapted from Weinstock et al. Screening for Childhood Strabismus by Primary Care Physicians (1998).

## 6. Prognosis

With early detection, the prognosis of strabismus is excellent. Thus, it is extremely important to know when to refer a child to an ophthalmologist. (See table 4) Under conditions of accurate diagnosis and administering proper treatment before the ages of 6 years the outlook for children diagnosed with strabismus has been positive. Unfortunately, however, once a child reaches the age of 8 to 10 years, treatment of strabismus has been found to be unsuccessful and can result in permanent decrease in vision.

**Table 4:**

<b>Indications to Refer to an Ophthalmologist</b>
<ol style="list-style-type: none"> <li>1. Constant esotropia at any age</li> <li>2. Persistent esodeviations at 4 months of age</li> <li>3. Positive corneal light reflex or cover test that reveals deviation</li> <li>4. Asymmetry of appearance on Bruckner test</li> <li>5. Incomitant deviations</li> <li>6. Parental concern about ocular alignment</li> </ol>

Adapted from Coats and Paysse, Evaluation and management of strabismus in children. Up To Date, 2008.

## **7. Note on examinations:**

Many infants and young children are very difficult to assess and it is therefore difficult to obtain a valid diagnosis. Further, older children who have developmental delays or are non-verbal can be challenging to examine. Currently, ocular photo-screening has been used to screen patients for factors such as strabismus because it requires little cooperation from the patient; the need to draw attention for fixation on an object is not required as used in standard evaluation techniques. With the current technology showing promising results, the use of photo-screening could potentially improve the ability to diagnose individuals who are at the highest risk of amblyopia conditions. Current use of ocular photo-screening involves a unique camera and video system that acquires images of pupillary reflexes and red reflexes. Upon examination, children that display abnormal findings are further evaluated with techniques used today (see Table 3). Two photo-screener systems are available which include the MTI Photo-screener and the Visiscreen 100. Although current research reports have shown the ability for photo-screening to be a valid assessment for amblyopia, further research and evaluations need to be administered to ensure proficient results.

## References

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