

Childhood Strabismus: Taking a Closer Look at Pediatric Squint

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S trabismus, colloquially known as squint, is a common pediatric problem with an incidence of three per cent to four per cent in the population. It is frequently associated with poor vision because of amblyopia and is occasionally a harbinger of underlying neurologic or even life-threatening disease. The family physician has a vital role in identifying strabismus patients and re-enforcing treatment, ensuring followup and compliance once treatment is started.

The different manifestations of strabismus derive their name from the direction of occular deviation, as well as whether it is latent or manifest (Table 1).

Congenital strabismus

Although babies will not outgrow strabismus, many infants have intermittent strabismus, which resolves by four months, due to their immature visual system. Therefore, it is best to delay referral for strabismus for the first four to six months of an infant's life. There are two common ages for the development of comitant strabismus: birth to six months and two to four years of age. Congenital or infantile esotropia is usually very obvious, with a large angle of deviation. A constellation of findings, such as latent vertical deviations, latent nystagmus, or a crossfixator with poor abduction are classic markers of the congenital esotrope. Since these children avoid lateral gaze, their condition resembles congenital VIth nerve palsy. This is ruled

Table 1 Strabismus manifestations		
	Latent (phoria)	Manifest (<i>tropia</i>)
Convergent	Esophoria	Esotropia
Divergent	Exophoria	Exotropia
Vertical (up)	Hyperphoria	Hypotropia
Vertical (down)	Hypophoria Comitant (the angle is the same in all directions of gaze) Congenital (very soon after birth)	Hypotropia Non-comitant (differs in all directions of gaze) Acquired

out by rotating the baby to elicit abduction nystagmus, or by "Doll's head" quick head turn, both of which will move the eyes into abduction. Congenital exotropia is seen infrequently, but is similar in features to congential esotropia.

Acquired strabismus

The second age group for the development of strabismus is between two to four years of age. Acquired esotropia is usually accommodative and responds to glasses. The onset can be intermittent, gradually becoming constant over months, weeks, or occasionally, days. In rare occasions, onset is sudden, which will often frighten parents, precipitating an ER visit.



VIth nerve palsy is ruled out by checking abductions, which are full. Amblyopia develops quickly and can be severe. Early recognition and treatment of accommodative esotropia can prevent loss of fusion and help maintain excellent vision. Exotropes whose condition was acquired, unlike esotropes, tend to remain intermittent and therefore, rarely develop amblyopia. They are more difficult to identify in the family physician's office, since they are often only manifest for distance. One clue is a history of the child shutting one eye when in sunlight. Among atypical acquired exotropes whose onset was sudden, large angle deviation is suspicious for underlying neurologic problems, like multiple sclerosis or leukodystrophy.

Non-comitant strabismus

Non-comitant strabismus can occur at any age: adult, childhood, or infancy. The deviation will be larger in the direction of action of the paretic muscle or in the direction away from the restricted muscle. The most common childhood cranial nerve palsy is VIth nerve palsy. This presents with torticollis. It is usually congenital, perhaps due to the trauma of birth. The patient has a tilt away from the paretic eye and upon straightening the head, the paretic eye is at a higher position (Figures 1a and b). Any abnormal head position torticollis, in particular, should have an ophthalmologic exam to rule out an ocular cause. When acquired VIth nerve palsy occurs in childhood, it is usually post-viral and clears in six weeks. The condition will need treatment with prisms and or patching to prevent loss of fusion and amblyopia. Congenital VIth nerve palsy is rare. Much more common is Duane's syndrome which is due to misdirection of the fibers of the VIth nerve.



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Figure 1a. A patient with a seemingly normal gaze.



Figure 1b. Examination demonstrates VIth nerve palsy.



Figure 2b. In pseudostrabismus, less sclera shows medially due to wide epicanthal folds



Figure 2b. In true esotropia, the light reflex is lateral in the left eye.

Examination

The examination for strabismus can be done fairly easily in your office with a few tools:

- a pen light,
- fixation toy,
- a hand or thumb as an occluder and
- an opthalmoscope.

The corneal light reflex test is done at arms' length, without disturbing the child. In pseudostrabismus, the light reflex is centered in each eye, but less sclera shows medially due to wide epicanthal folds (Figures 2a and b). In true esotropia, the light reflex is lateral in the left eye (Figure 3).

The cover test, while slightly more difficult to perform, gives more information and is more accurate. If the child or infant strongly objects to only one eye being covered, the uncovered eye may have poor vision. Your hand or thumb can be used instead of an occluder to break fixation. In the cover test, choose which eve looks abnormal, then cover the other eye, watching the uncovered eye for movement as it takes up fixation. False negatives occur if the child has very poor fixation or eccentric fixation due to poor vision. False positives occur if the child changes fixation at the wrong time (Figure 4). An important part of the examination is to look at the red reflex using the opthalmoscope set at 0, at a distance of 10 inches. A cataract or corneal opacity will show up as black against the red reflex from the opthalmoscope, while some large fundal lesions will appear white (leukocoria). Poor vision in one eye can be diagnosed as strabismus. When a child presents with strabismus and poor vision, it is important to differentiate whether there is an underlying cause, or if it is strabismus with amblyopia. A dilated fundus exam will answer this question.

Treatment

There is a window of opportunity for the treatment of amblyopia. The younger the age at diagnosis, the easier it is to treat, because the visual system has plasticity forming and reforming pathways until about age eight. Anisometropic amblyopia, due to unequal refractive error is often missed until later, when the amblyopia is much more refractory to treatment. The vision may be as poor as count fingers vision in an eye with a visual potential of 20/20. The best age for vision testing is three to four years of age. This is the most important screening eye test in this age group. It will pick up treatable vision loss that would otherwise be missed. Treatment of strabismus follows three steps:

- 1. Glasses to correct underlying refractive error
- 2. Glasses to correct occlusion, if there is amblyopia
- 3. Surgery



Figure 3. When the eyes are straight (top) the reflex is centered in both eyes; with esotropia (middle) the reflex is lateral, with exotropia (bottom) the reflex is medial.



It may be hard for parents to accept that surgery will not cure accommodative strabismus, which is correctable by glasses. Although many children will outgrow the need for glasses, some will require them into adulthood.

Final thoughts

Given current physician shortage and long waits for specialists, the family physician plays a vital role in recognizing and prioritizing referrals on the basis of urgency. If, in your assessment there is concern, do not hesitate to call an ophthalmologist directly.