

Strabismus

Nathalie Azar, MD

Pediatric Ophthalmology for the Non-Ophthalmologist

April 7, 2018

TERMINOLOGY:

Strabismus comes from the Greek word “Strabismos” which means to squint. For accuracy when describing the misalignment, we use the term –phoria or -tropia as a suffix and the prefixes Eso-, Exo-, Hyper-, Hypo, Incyclo- or Excyclo-.

A phoria is a tendency for the eyes to become misaligned when binocular viewing is disrupted but normal realignment is regained when the occluder is removed. A tropia on the other hand is a manifest misalignment. It can be intermittent or constant. When intermittent, there are times during the day when the eyes are straight and other times when the eyes are misaligned.

The prefixes used are the following:

Eso: inward deviation of the eye towards the nose.

Exo: outward deviation of the eyes.

Hyper: the deviated eye is higher than the fixing eye.

Hypo: the deviated eye is lower than the fixing eye.

Incyclo: the 12:00 meridian is turned towards the nose.

Excyclo: the 12:00 meridian is turned away from the nose.

In practice we use the following abbreviations:

E: Esophoria

X: Exophoria

ET: Esotropia

XT: Exotropia

E(T): Intermittent Esotropia

X(T): Intermittent Exotropia

X': Exophoria at near

ET': Esotropia at near

(Note: ' denotes near measurement)

EVALUATION OF STRABISMUS:

History:

- Date of onset of the deviation, constant or intermittent, deviation at distance or near or both, presence or absence of diplopia, inciting factors, factors relieving the symptoms,

family history of strabismus or amblyopia are some of the questions that need to be investigated among others.

Assessment of monocular Eye Function:

- Visual acuity distance and near, refraction, rule out of amblyopia

Assesment of Binocular sensory functions:

- Stereoacuity, fusion tests

Assessment of Binocular Motor Function Tests:

- Eye movements
 - Ductions: are the monocular rotations of the eye. For example, adduction is the movement or rotation of the eye towards the nose.
 - Versions: are the rotations of the fellow eye in conjugate eye movements.
 - _Vergences: are the movements of the eyes in opposite directions in disconjugate eye movements as in convergence and divergence.
- Ocular Alignment
 - Corneal Light reflex: used in patients who cannot cooperate with cover testing or in patients with extremely poor vision. Three tests of light reflexes are used:
 - 1) Hirschberg: 1mm of decentration of corneal light reflection corresponds to 7 degrees of 15 prism diopter of deviation
 - 2) The Krimsky method: a prism is placed over the fixing eye and the power is increased in order to achieve a centered light reflex or the non-fixing eye.
 - 3) The Brückner test: the direct ophthalmoscope is used with its largest aperture at a distance far enough to achieve a simultaneous red reflex in both eyes. In the presence of strabismus the deviated eye will have a brighter red reflex than the fixating eye. This is not a quantification test.
- Cover tests: requires patient cooperation, eye movement capability, image formation and perception. Four types of cover tests are used
 - 1) Cover-uncover test: detects the presence of a tropia (manifest strabismus) and differentiates a tropia from phoria. As one eye is covered the examiner observes for any movement in the uncovered eye; if a movement is present then a tropia is present.
 - 2) The alternate cover test: measures the total deviation both latent and manifest. The cover is placed alternately in front of each eye several times to dissociate the eyes and the eye movement in the uncovered eye is observed. If the eye is seen to move from out to center as it is being uncovered and the fellow is covered then an exo-deviation is present.
 - 3) The alternate cover prism test: a prism is placed over one eye and the alternate cover test is performed. The amount of prism is either increased or decreased in order to neutralize the movement of the eyes under the cover. The amount of prism required for neutralization corresponds to the

measurement of the deviation. This test does not differentiate between the measure of the amount of phoria and tropia.

- 4) Simultaneous prism test: this test is performed in the fixating eye at the same time a prism is placed in front of the deviating eye. The test is repeated using increasing prism power until the movement is neutralized. This test measures the tropia only.

Definitions:

- Comitant deviations: misalignment measures the same in all positions of gaze
- Incomitant deviations: misalignment measures differently in different positions of gaze, usually seen in paralytic and restrictive strabismus.
- A pattern strabismus: shows the eyes closer together in upgaze than downgaze with a 10 prism diopter difference between the two positions.
- V pattern strabismus: shows the eyes closer together in upgaze than downgaze with a 15 prism diopter difference between the two positions.

STRABISMUS ENTITIES:

ESODEVIATIONS

Pseudoesotropia:

- Not a true esotropia
- There is an appearance of esotropia even though the eyes are well aligned. This is due to a flat and wide nasal bridge, prominent epicanthal folds, or a narrow inter pupillary distance.
- Observer sees less nasal sclera especially with gaze to either side, giving the appearance of esotropia
- The corneal light reflexes are well centered

Congenital Esotropia:

- Esotropia noted within the first 6 months of life. Esotropia is very rarely seen at birth, commonly presents at about 2 to 4 months of age. The term *infantile* esotropia more accurately describes this entity.
- Patients have a very large angle strabismus, usually greater than 30 prism diopters. The angle of misalignment is stable: when a fluctuation in the strabismus angle is seen, one should consider an accommodative component to the underlying deviation.
- Family history of esotropia is often present, but well defined genetic transmission is rarely seen.
- Normal hyperopic refractive error, usually less than 2 diopters

- Equal visual acuity is common. When present, the observer will find cross-fixation which is, in a large angle esotropia, the use of the adducted eye for fixation on objects in the contralateral temporal visual field. When the object of regard crosses midline the patient will pick up fixation with the fellow eye.
- Children with congenital esotropia are usually neurologically intact. Esotropia can be found in up to 30% of patients with neurological and developmental problems such as cerebral palsy and hydrocephalus.
- Etiology of congenital esotropia has not been elucidated.
- Need to differentiate congenital esotropia from bilateral 6th nerve palsy or Möbius syndrome. Abduction in each eye is tested by the doll's head maneuver, rotating the child and patching of the fellow eye.
- Management:
 - Cycloplegic refraction: rule out high hyperopic refractive error and underlying accommodative component.
 - Treat amblyopia if present
 - Surgery to achieve ocular alignment
- Surgery:
 - Goal of surgery is to achieve orthotropia and promote sensory fusion and hopefully stereopsis. Achieving a small angle esotropia of 8 prism diopters or less (Microtropia) is also desirable.
 - Timing is crucial: best within the first 24 months of age. Around age one is preferred.
 - Early surgery gives best chance for achieving binocular function after ocular realignment
 - Stereopsis and binocular function are often suboptimal even after the best ocular alignment is achieved.
 - Microtropia is a small angle esotropia of 8 prism diopters or less resulting in a stable ocular alignment with peripheral fusion, central suppression, and good appearance.
 - Surgery consists of weakening the medial rectus of each eye or weakening of the medial rectus and tightening the lateral rectus of the same eye.
 - It is not uncommon for children to require multiple surgeries to achieve the desired result.

Accommodative Esotropia

- Onset around age 2 and a half although it may range between 6 months to 7 years
- Often starts out as intermittent before becoming constant.
- Can be precipitated by an illness
- Family history of strabismus is common
- Amblyopia not uncommon
- Diplopia can be present initially and in younger children is easily suppressed

Refractive Accommodative Esotropia

- Moderate to high hyperopia with accommodative convergence and defective divergence. The patient needs to accommodate in order to correct the blur induced by the hyperopia. Since accommodation is coupled with convergence, the eyes would want to turn in. If the divergence mechanism is defective, the eyes would become esotropic.
- Esotropia is of a moderate amount ~ 20 prism diopter and measures essentially the same at distance and near.
- Hyperopia averages ~ 4 diopters
- Treatment:
 - Correct refractive error and give full hyperopic correction in glasses. Full time wear of glasses is essential. Over time, give the least amount of hyperopic power necessary to keep the eyes aligned. This will stimulate and strengthen fusional divergence mechanisms. Wean the patient off glasses over time with the aim of discontinuing spectacle correction and maintaining ocular alignment.
 - Correct the amblyopia by either patching or pharmacological penalization.

High Accommodative Convergence/Accommodation (AC/A) Esotropia:

- Patients have an abnormal relationship between accommodation and accommodative convergence, as they have excessive convergence per diopter of accommodation. This leads to an esodeviation that is greater at near than at distance.
- Patients with high AC/A ratio may have an underlying high hyperopia, moderate or low hyperopia, emmetropia, or even myopia.
- Treatment:
 - Correct the refractive error.
 - Correct the amblyopia.
 - For the near deviation bifocal add of +3.00 diopter is prescribed for each eye. This should be ground into the lenses in a flat top style with the bifocal line bisecting the pupil. The bifocal add eliminates the need for accommodation at near and keeps the eyes aligned. With time, the power of the bifocal add is reduced to simulate fusional divergence with the goal to eliminate the bifocal by age 10 to 12 years. If the bifocal power cannot be eliminated by that age, surgery for the near deviation is recommended.

Partially Accommodative Esotropia:

- Some patients may have a large angle esotropia and moderate hyperopia. With the full correction of hyperopia, the esotropia angle is reduced but not eliminated. These patients will require surgery aimed at correcting the residual angle while wearing their full hyperopic correction and glasses. The parents should be told in advance to expect the eyes to be straight with glasses after surgery, but to exhibit a smaller angle esotropia without their glasses. These patients will need to continue wearing glasses full time with the power of their correction decreasing as they get older. Some may ultimately outgrow their need for glasses and maintain good alignment without optical correction.

Non-Accommodative Acquired Esotropia

Basic Esotropia

- Esotropia that develops after 6 months of age and is not associated with an accommodative component.
- Deviation is same at distance and near
- CNS lesions may need to be ruled out although those patients are often neurologically intact.
- Management entails treating the amblyopia and surgical correction

Sensory Esotropia:

- In the presence of monocular vision loss such as optic atrophy or cataract, there is absence of fusion and therefore the stimulus to keep the eye aligned is no longer present. A deviation ensues with younger children having a tendency of developing an esotropia and older children and adults developing exotropia.
- The treatment is aimed improving the visual acuity if possible (i.e. cataract surgery), then eye muscle surgery to realign the eyes.

Consecutive Esotropia:

- Results following surgery for exodeviation. Small angles often resolve spontaneously, larger angles require surgical correction.
- If fusion was present prior to the exotropia surgery, then prompt surgical correction of the large consecutive esotropia is required.
- Consecutive esotropia may develop many years after exotropia surgery if there was no development of fusion.

Sixth Nerve Palsy:

- Paralysis of the sixth nerve which elevates the lateral rectus will result in an incomitant esodeviation with a greater esotropia angle on the side of the paralysis.
- Diplopia is common in older patients.
- Face turn towards the side of the paretic muscle is adopted in order to avoid diplopia.
- Majority due to infectious or immunologic processes
- Approximately one third of cases associated with intracranial lesions
- Amblyopia can develop
- Management:
 - Careful history
 - Imaging if intracranial lesion suspected
 - Patching to maintain vision and avoid amblyopia
 - Botulinum Toxin to the antagonist medial rectus has been used in older patients.
 - Spontaneous resolution occurs often
 - Surgery indicated if no significant resolution occurs after at least 6 months

○

EXODEVIATIONS

Pseudoexotropia:

- Appearance of an outward deviation of the eyes when the eyes are properly aligned.
- This may be seen with wide interpupillary distance, positive angle kappa, ocular abnormalities such as temporal dragging of the macula seen in retinopathy of prematurity.

Exophoria:

- A tendency for an outward drift of the eyes that is controlled by fusion under binocular viewing conditions.
- This condition can be detected by the alternate cover test.
- It is fairly common.
- It is often asymptomatic, unless the angle of the latent deviation is large requiring large convergence amplitudes.
- A large exophoria becomes symptomatic when near requirement increases such as increased reading assignments in high school and college.

Convergence Insufficiency:

- Presents as asthenopia (pain with reading), blurred near vision, and or double vision at near.
- Patients have a large exophoria at near only
- Treatment consists of orthoptic exercises
- Base-in prisms in reading glasses when exercises fail, eye muscle surgery on the medial rectus when nonsurgical treatments are unsatisfactory

Intermittent Exotropia:

- The most common type of exodeviations.
- It is latent at times and manifest at others.
- Usually occurs early on, before age 5
- Diplopia is not a common feature, unless onset is after age 10.
- Parents usually report that the outward deviation occurs late in the day, during illness, day dreaming, or drowsiness.
- More frequently seen at distance than near.
- Can be progressive to a constant exotropia
- Treatment: glasses or prisms are temporizing measures. Ultimately surgery is the treatment of choice for a progressing problem.
- Surgery consists of symmetric recession of the lateral rectus in each eye or a recession of the lateral rectus and a recession of the medial rectus in the same eye.

Constant exotropia:

- Often the result of decompensated intermittent exotropia or manifest sensory exotropia.
- Treatment is surgical

Congenital exotropia:

- Much less common than congenital esotropia
- Onset is within the first 6 months of age
- Large angle constant deviation
- Many children will have associated neurologic or craniofacial disorders
- Treatment is correction of refractive error, correction of amblyopia and early surgery. As with congenital esotropia, the potential for binocular vision is not great.

Sensory exotropia:

- Results from loss of vision in one eye which causes loss of fusion. This can be seen in high and anisometropia, corneal lens opacities, optic atrophy, and macular lesions among others.
- More common in older children and adults.
- Treatment: try to improve vision and then surgery for ocular realignment

Consecutive exotropia:

- Exotropia resulting from surgery for esotropia
- Treatment is surgical

VERTICAL DEVIATIONS:**Superior oblique palsy or paresis:**

- Most common vertical misalignment
- Findings:
 - Head tilt to one side with eyes aligned,
 - Vertical misalignment seen when the head tilts to the contralateral side.
 - Vertical diplopia can be noted by some patients.
 - Inferior oblique overaction in the field of action of the paretic superior oblique muscle
- Congenital
 - Congenitally lax, attenuated or even absent superior oblique tendon
- Acquired
 - most likely due to closed hit trauma, less commonly due to CNS vascular problems, diabetes, or brain tumors
- Can be unilateral or bilateral
- Underaction of the superior oblique muscle and overaction of the ipsilateral inferior oblique.

- Deviation is greatest with a head tilt to the ipsilateral side, which explains why patients tilted their head to the contralateral side to maintain ocular alignment and binocularity.
- Treatment is surgical
 - Surgery on one vertical muscle if the deviation is less than 15 prism diopters
 - Surgery on two vertical muscles if the deviation is more than 15 prism diopters

Dissociated vertical deviation (DVD):

- Common innervational disorder
- Seen in patients with congenital esotropia and other forms of strabismus
- Associated with early disruption of binocular development
- When an eye is occluded or during periods of inattention, the fellow eye drifts up and outward with extorsion. As the vertically deviated eye moves down to fixate, the fellow eye will not make a downward movement. Therefore, this is not a true hypertropia as the fellow eye will not make a downward movement into a hypotropic position of equal magnitude.
- The condition is bilateral and often asymmetric
- Often seen after correction of the horizontal deviation in congenital esotropia and not before.
- Indication for treatment:
 - If the deviation happens spontaneously, frequently, or is of a large magnitude so that it is cosmetically significant
- Treatment :
 - Changing the fixation preference by optically or pharmacologically blurring the eye with the lesser vertical deviation in asymmetric DVD.
 - Surgery on both eyes with asymmetric displacement of the muscles in asymmetric DVD.

Monocular Elevation Deficiency (Double Elevator Palsy):

- Limitation of elevation in adduction and abduction and straight upgaze.
- Hypotropia of the involved eye
- Chin up position to maintain fusion in downgaze.
- Ptosis or pseudoptosis may be present
- Treatment is surgical
 - Indications: large vertical deviation in primary gaze, abnormal head position
 - Surgery: transposition of the medial and lateral rectus to the superior rectus or recession of the tight inferior rectus

Orbital floor fractures:

- Most commonly from motor vehicle accidents
- Direct impact on the orbit, closing the orbital entrance causing acute increase in orbital pressure and leading to fracture of the orbital floor, hence the term blowout fracture.

- Clinical presentation: ecchymosis of the involved eye, diplopia in some or all positions of gaze, paresthesia or hypoesthesia, enophthalmos, hypotropia may or may not be present in primary gaze increasing in upgaze.
- Diagnosis confirmed by orbital imaging, CT, and MRI and forced duction testing
- Surgical management of the fracture: controversial, immediate exploration versus waiting a few days to two weeks.
- Management of the strabismus: wait 6 months for any signs of recovery, eye muscle surgery for the residual deviation.

SPECIAL FORMS OF STRABISMUS:

Duane syndrome:

- Spectrum of motility disturbances: all have retraction of the globe on attempted adduction, horizontal eye movements are limited.
- Limitation of abduction (Type I): more commonly seen than limitation of adduction (Type II). Limitation of both adduction and abduction to a certain degree is called Type III.
- Syndrome is sporadic, high prevalence in females, more common in the left eye
- Sixth cranial nerve nucleus is absent, aberrant branch of third nerve innervating the lateral rectus which leads to co-contraction of the medial and lateral recti muscles and retraction of the globe on adduction
- Can be associated with Goldenhar syndrome (hemifacial microsomia, ocular dermoids, ear anomalies, preauricular skin tags, upper eyelid colobomas), and Wildervanck syndrome (sensorineural hearing loss and Klippel-Feil anomaly)
- Management:
 - Indications: abnormal head position/exaggerated head turn, marked globe retraction, upshoots and downshoots of the involved eye
 - Treatment is surgical with recession of the medial rectus on the involved side coupled with surgery on other muscles if indicated.

Brown Syndrome:

- Deficient elevation of the eye in adduction
- Forced duction testing to differentiate it from inferior oblique palsy
- Patients may present with a head tilt or chin up position
- Management depends on whether the etiology is inflammatory or not. Local injection of corticosteroids near the trochlea or bringing the underlying systemic disease into remission may correct the deviation in inflammatory cases.
- Surgery is indicated in non-inflammatory cases, when there is a hypotropia in primary gaze, significant abnormal head position, or both.

Möbius Syndrome:

- Rare condition, both 6th and 7th nerve palsy, mask like faces, esotropia
- Gaze palsies attributed to abnormalities in the pontine reticular formation, indicating that the lesion is not only nuclear
- Limb, chest, and tongue defects in many patients
- Poland syndrome exhibits absent pectoralis muscle

SUMMARY

- Recognize ocular misalignment
- Treatment consists of:
 - Correcting refractive errors
 - Patching, pharmacologic or optical penalization for amblyopia
 - Bifocals for high AC/A ratio
 - Manipulating power in the glasses to control the alignment
 - Surgery when optical correction does not correct the misalignment
 - Botulinum Toxin for specific indications