

Lecture 23 – Developmental Strabismus

(Steinman Chapter 9; Adlers 9th Edition Chapter 22)

CONTEXT OF STRABISMUS

We have already mentioned three kinds of ocular misalignments that can affect binocular vision:

Fixation disparities

- very small—normally 5 arc minutes (0.15Δ) or less
- requires binocular fusion
- stereopsis expected
- some fixation disparity is normal and expected
- Large fixation disparities may indicate some binocular dysfunction.

Heterophoria

- normally less than 10Δ
- It is not present during binocular fusion, but
- is the rest position that the eye turn to *when binocular fusion is temporarily interrupted*.
- Small phorias are normal.
- Large phorias can lead to binocular eye strain.

Strabismus/Heterotropia

- Up to $30\Delta +$
- angle of deviation exceeds width of Panum's area
- binocular fusion impossible

Table 1. Three ocular misalignments.

Oculomotor deviation	Size	Binocular fusion?	Test condition
Fixation disparity	$< 5'$ ($\sim 0.15\Delta$)	yes	binocular fusion
Heterophoria	$< 10\Delta$	yes	interrupt fusion
Heterotropia	Up to $30\Delta +$	no	monocular fixation

We are concerned about strabismus for two reasons:

- If present during the critical period, it can prevent the development of normal binocular vision
- Cosmesis

EARLY ONSET STRABISMUS

The four major subcategories of developmental strabismus are summarized in **Adler's Fig. 24-49**. These differ in their time course, incidence and characteristics.

- **Infantile esotropia** is the dominant kind of strabismus from 6-12 months of age and makes up about 1% of the population.
- **Refractive esotropia** begins to increase in frequency from age 1 and peaks at about age 3.
- **Intermittent exotropia** has an onset at age 1 and gradually increasing, but relatively rare.
- **Constant infantile exotropia** is relatively rare, showing up at about age 6 months.

Quoting from Adler's Fig. 24-49,

Infantile esotropia is of major importance because it is associated with permanent deficits in stereopsis and binocular motion processing, and often requires multiple surgical procedures to

achieve stable eye alignment. Refractive esotropia (partially accommodative or accommodative) is often cured with spectacles and/or one surgery. Intermittent exotropia is often cured with one surgery. Constant infantile exotropia is distinctly rare.

As mentioned previously, during the first 6 months of life, it is not unusual for babies to have an intermittent strabismus, usually exotropia. Normal motor and sensory fusion should develop by 6 months of age, so by then, 98% of infants have aligned eyes, symmetric smooth pursuits and stereopsis. After about 6 months of age, the four main categories of strabismus begin to manifest themselves among the remaining 2% of the infants.

INFANTILE ESOTROPIA

The most common kind of developmental strabismus that is present in infants is infantile esotropia. This had been called, “congenital esotropia”, but this term is incorrect since it does not appear until after about 4-6 months of age. In addition to the large strabismus, usually about 15-30 prism diopters, the following anomalies are associated with infantile esotropia:

- loss of stereopsis
- asymmetric smooth pursuits
- incorrect motion perception
- latent nystagmus when fixating stationary targets
- face turn
- vertical deviation

Loss of stereopsis

Recall that stereopsis rapidly develops between about 3 and 5 months of age, so prior to the onset of infantile esotropia, these babies begin to develop normal stereopsis. After this, the infantile esotropia appears, but this is still within the critical period (infantile phase), so they rapidly lose whatever stereopsis they had developed prior to that time. (**Adler’s Fig. 24-51**)

Asymmetric smooth pursuits

Prior to about 3 months of age, infants prefer to follow targets moving nasally (from a temporal to nasal direction). With the development of normal stereopsis between 3 and 5 months, symmetric pursuits also develop. Children with infantile strabismus, however, continue to show a preference for temporal to nasal pursuits.

Motion perception

In addition to having better smooth pursuits in the nasalward direction, objects moving in this direction appear to be moving more quickly than if they are moving in the opposite direction. In these people, the pursuit asymmetry and incorrect motion perception persist into adulthood. This is illustrated by **Adler’s Fig. 24-52**.

Latent fixation nystagmus

These infants also show a nystagmus when attempting to fixate stationary objects. This is smaller or latent when both eyes are opened, but becomes most pronounced when the strabismic eye is covered. The covered eye will move with the fixating eye (**Adler’s Fig. 24-53**).

This is probably related to the maldevelopment of the smooth pursuit and motion perception mechanisms in the brain. This condition persists, but the infant soon adapts to the nystagmus and becomes unaware of any movement of the visual scene in spite of the nystagmus. The nystagmus, however, degrades visual acuity.

Face turn

Infants with infantile esotropia tend to rotate their head so that the fixating eye is positioned in the nasal canthus. For example, in a left esotropia (OD fixating), the child will tend to turn his face to the right (**Adler's Fig. 24-54**). This helps to reduce the velocity of the nystagmus and improves acuity. They also have difficulty moving their eyes temporally (abduction).

Vertical deviation

Fifty to 90% of these infants also show a vertical deviation that manifests itself when one eye is covered. When either eye is covered it will begin to drift upward; when the cover is removed, it will drift downward. When the other eye is covered, it will also drift upward, then return downward when uncovered. This condition is known as a **dissociated vertical deviation (DVD)**.

Q. How does this differ from a normal hypertropia or hyperphoria?

A.

In addition many patients who have had infantile esotropia show an over-action of one or both inferior obliques. The eye with the overacting inferior oblique will over-elevate during nasal versions.

Neural mechanism

Over 90% of all infantile strabismus and 75% of later onset strabismus are esotropia. The etiology appears to be abnormal development in the visual areas of the brain rather than a problem with the extraocular muscles.

We know that normal visual input is required for the development of stereopsis during the infantile phase of the critical period (to 8 months), and it appears that the development of motion perception and eye alignment are closely related to stereopsis during this early developmental period. The failure of one of these functions to develop interferes with the development of the others.

Studies with monkeys suggest that abnormal neuronal development begins with an imbalance in the ocular dominance columns at layer IVC (recall **Adler's Fig. 24-47**). The second order neurons, which contain the first disparity selective neurons, receive anomalous input. This can affect neurons in magnocellular tract as well, and that will retard development of motion perception in area V5 (MT) (**Adler's Fig. 24-56**).

Tychsen proposes that, since most newborns have a tendency to drift toward intermittent exotropia, the oculomotor and motion systems develops a strong compensating nasally bias. However, in order to develop well-balanced ocular alignment, the visual system requires feedback, some of which is provided by stereopsis. Disparity detectors signal to the brain that the eyes have either over-converged, or are still underconvergent. If the stereoscopic feedback system fails to develop, the infant may be left with an overconvergence (esotropia), and the unbalance preference for nasal motion persists. This is illustrated in **Adler's Fig. 24-58**. The parallel development of stereopsis, symmetric pursuits and motion perception all support each other. This is just one of several theories about how infantile esotropia develops.

Management options

Early intervention, before age 2, is critical in cases of infantile esotropia. If treatment is delayed beyond age 2, it is unlikely that the person will ever develop completely normal binocular function.

Refractive correction. The first step in treatment is to correct any refractive error that may be present. If hyperopia is present, it may contribute to the tendency for an eso deviation. Prism, up to about 23 prism diopters, may also be prescribed to allow binocular fusion.

Vision therapy. This should be supported by a program of vision therapy. Vision therapy may include occlusion combined with training techniques to stimulate monocular fixation and abduction.

Surgery. If the child still cannot develop binocular fusion, surgery may be necessary to assist with the ocular alignment and motor fusion. If performed before the age of 2, the probability of developing stereopsis is better, but patients who have had surgery often have a small residual strabismus after surgery.

ACCOMMODATIVE (REFRACTIVE) ESOTROPIA

Beginning from about age 1, some infants begin to show accommodative esotropia (**Adler's Fig. 24-49**). Most have an uncorrected hyperopia of at least 3 diopters and strong accommodative convergence. In order to see clearly at all distances they must accommodate, and the excessive accommodative convergence leads to esotropia.

The time of onset and basic etiology of this condition is different from those of infantile esotropia. Often the strabismus can be successfully treated by simply correcting the refractive error. When strabismus surgery is required, the success rate (defined as long term alignment and stereopsis) is relatively high (80-90% according to Tychsen).

There are fundamental differences between children who develop infantile esotropia and those who develop esotropia after age 1. Many of the infantile esotropes are babies who were born prematurely. They usually have very little refractive error and are difficult to treat. Following surgical correction of infantile esotropia, only about 50% are able to develop stereopsis.

Tychsen (Adler's 9th ed. p. 827) suggests the reason for the basic differences between children with infantile and refractive esotropia.

The difference is more likely related to the degree to which connections for normal binocular motion processing and stereopsis were established before the onset of strabismus. Infants who develop strabismus [infantile esotropia] have arrested development of stereopsis during the critical period for development of both of these binocular functions, whereas children who develop [accommodative] esotropia after infancy have enjoyed a period of normal binocular development.

Some doctors subdivide refractive esotropia into two subcategories:

Accommodative esotropia with a normal AC/A

- Etiology: uncorrected refractive error.
- Refractive error of +2-6 diopters
- Similar angle of deviation at far and near
- Usually intermittent
- Good prognosis with spectacle or contact lens correction of hyperopia

Accommodative esotropia with a High AC/A

- Etiology: high AC/A
- Refractive error of < 2 diopters
- Larger eso deviation at near than far
- Usually constant at near, may be intermittent at far
- Correct any hyperopia and Rx bifocals (seg height at mid-pupil), plus VT. In some cases surgery may be needed, but it can be deferred until age 5 or 6 if the deviation is intermittent.

INTERMITTENT EXOTROPIA

This is a relatively small subgroup compared to the esotropes. Onset begins at about age 1 and increases over the succeeding years (**see Adler's Fig. 24-49**).

The disruption to binocular vision seen in these exotropes is usually less severe than that seen in esotropes. Since the onset is after 1 year of age, many of the important binocular visual functions have had time to develop (stereopsis, motion perception, smooth pursuits, etc.). The deviation is usually intermittent, so there are periods of time when the eyes are correctly aligned, giving the visual system regular exposure to normal binocular input. This helps to preserve stereopsis and prevent amblyopia.

In exotropia, the deviation is seen more often at far than at near. Accommodative convergence, which comes into play during near fixation, helps to align the eyes, and since near objects are larger, they are easier to fixate. Near objects also provide better disparity cues.

Vision training for exo deviations is more successful since the eyes can more easily converge than diverge. For deviations greater than about 25 prism diopters, surgery may be required in addition to vision training to allow the patient to achieve ocular alignment all day. Success rates for surgery are also relatively high--85% achieve long term ocular alignment and stereopsis.

CONSTANT INFANTILE EXOTROPIA

The fourth major category of strabismus is constant infantile exotropia. It has an onset at about 6 months of age and is extremely rare (< 0.1%). Constant infantile exotropia is usually associated with retinal or optic nerve disease, or other anomalies at higher levels of the visual system. This patient population may include a small number normal infants with a slowly developing oculomotor system in which intermittent exotropia is still present. These may be holdovers or "late bloomers" from the intermittent exotropia group.

A, V and X patterns

In addition to the basic horizontal deviations discussed above, those patients may also have a differing angle of deviation depending on their vertical gaze.

- A larger eso (or smaller exo) deviation with superior gaze and larger exo (smaller eso) inferiorly is known as an A pattern.
- A larger exo (smaller eso) superiorly with a greater eso (smaller ex) inferiorly is known as a V pattern.
- Some patients will show greater exo (less eso) deviations for both superior and inferior gaze. This is known as an X pattern.

These appear to be caused by mechanical rather than paretic factors such as under or over actions of the oblique or vertical recti muscles. They are most commonly seen in esotropia with the V pattern being more common than the A pattern.

MICROTROPIA

Following surgery, some patients will have a cosmetic cure, yet they will be left with a small constant strabismus of 1-9 prism diopters. The residual strabismus is big enough that it interferes with normal bifoveal fixation, and they will probably have central suppression, while they are able to fuse peripherally. This is because Panum's area is larger in the periphery. These patient may develop some stereopsis, but usually not the high quality of stereopsis that would come with normal bifoveal fixation.

This is also sometimes called **microstrabismus** or **monofixation syndrome**. These patients are usually asymptomatic, and are usually not treatment.

SURGERY AND/OR VISION THERAPY?

Opinions differ as to when a strabismic child should be referred for surgery. This will depend on the type and characteristics of the strabismus. Surgeons are generally biased toward a surgical solution and downplay the benefits of vision therapy. Some may view vision therapy as “unscientific”, and this may be partially due to the fact that much of vision therapy rests on the foundation of psychophysics, which is less predictable than biomechanics. In addition, vision therapy uses tools that may appear unsophisticated compared to surgery.

Among optometrists there is also a range of opinions, but ODs generally recognize that some patients can benefit from vision therapy. Since vision therapy is non-invasive, and surgery is irreversible, it is prudent to take maximum advantage of vision therapy and turn to surgery only when absolutely necessary.

Many vision therapy procedures emphasize cortical stimulation and cortical development, while surgery is purely mechanical, fixing the oculomotor end of the system. Keep in mind, however, that cortical and oculomotor aspects of vision are closely related. In the cases in which surgery is required, it helps align the eyes, and this gives sensory fusion a chance to develop. It will then be very important to support sensory fusion, which also helps the visual system sustain motor fusion. Vision therapy therefore can play an important role in helping patients after strabismus surgery.

You should learn as much about VT as you can while you're here, because there are a wealth of educational resources at the NSU College of Optometry. Your future patients will be depending on you for your expertise and clinical judgment in designing the treatment plan which is best for them.