

## Strabismus

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Strabismus is a fascinating and complex topic. It fails to draw attention of the patient except for the cosmetic consideration. A small corneal scar will bring him running to the doctor but amblyopic visual loss does not bother him/her so much.

Treatment of strabismus is demanding for the patients as well as the doctor. That the apparently normal looking eye could be blind functionally is beyond comprehension of the patients.

Treatment is laborious time consuming and lengthy hence dropouts are frequent.

Treatment seems to be unrewarding and frustrating both for the patients and the doctors.

The nervous control of the ocular movements is complicated. The muscles are supplied by nerves arising from nuclei in the mid-brain.

Their action is coordinated by intermediate centers situated in this region by which reflex activities are governed.

Finally these intermediate centers are linked with the vestibular apparatus where by they became associated with the equilibration reflexes and with cerebral cortex.

These complexities run the ocular motor function. Any disruption will lead to strabismus of one of the many varieties that we are going to consider.

### **Prevalence**

Strabismus mostly develops within the first 6-7 years of life. Horizontal strabismus is far more common than vertical strabismus. Approximately 7% of children of 6 to 7 years age develop strabismus. Amongst infants about 1% may develop strabismus. And about 3% school children have amblyopia related to strabismus.

### **Risk factors for strabismus**

1. Cigarette smoking during pregnancy
2. Neurological disorders
3. Use of toxic drugs during pregnancy (opiates, marijuana, barbiturates)
4. Lead poisoning
5. Twins
6. Heredity
7. Ethnicity
8. Refractive errors
9. Low birth weight (prematurity)
10. Anatomic factors (cranio-facial anomalies)

### **Predisposition**

Under 5 yrs age, family history of strabismus, Hyperopia > 4.00 D. Delay in refractive error correction could precipitate strabismus.

### **Prevention**

Prompt, periodical and complete refractive correction has a strong preventive value. Refractive error and strabismus are close associates.

### **Average refractive error**

The average refractive error in children shows increasing amounts of hyperopia in the first 5—7 years of life. Average hyperopia in a 6-year child is approximately 2.00 D.

After 7 years of age, refractive errors move toward the permanent distribution. 33% of children and young adults are myopic worldwide

A relationship between esotropia and hyperopia was noted in 1864 by Donders , Lepard, and later Duke- Elder and many others. An exact threshold for esotropia to occur can not be predicted on the basis of refraction alone.

### **AC : A ratio**

High Convergence response to accommodation is more likely to produce esotropia. Excessive Accommodative Convergence, high AC:A ratio, and inadequate fusion amplitudes are risk factors for esotropia.

Hyperopia is more common in esotropic and even in most of the exotropic children. Only a few exotropic children are myopic

### **Age & Refractive status**

Rapid changes in refractive status of children under the age of 1 year occur. In addition, the globe size and muscle relationship changes. They stabilizes toward the end of the first year of life. Hence preference of delayed surgery for an year.

### **Risks of Hyperopia**

The risk of acquiring accommodative esotropia when hyperopia exceeds 4.00D is substantial. High hyperopia are best corrected by the earliest. Hyperopia in excess of 5.00 D is potentially amblyogenic and may lead to bilateral amblyopia .

## Amblyopia

It may be defined as vision after correction to be **less than 6/24**. The incidence of amblyopia is about 2.%, less than 1% amblyopes develop strabismus.

### **Strabismic amblyopia**

Decrease in vision in the non-fixing eye, as the result of deviation. Strabismus prior to 6 years of age is a risk for developing amblyopia. If free alternation of fixation continues, head tilt, face-turn, or chin position allows the patient to fuse in a non-concomitant strabismus, amblyopia may be avoided.

Every child with strabismus needs to be carefully, periodically and repeatedly evaluated for refractive error correction and visual acuity in the eyes. Equal vision in the two eyes is an insurance against amblyopia

Under the age of 6 yrs, once fixation preference has been established the non-fixing eye is at risk of amblyopia. In general, fixation preference is more likely to occur in the smaller-angle

esotropics than in the larger-angle.

There is no evidence that a retinal or optic nerve abnormality causes amblyopia. Instead, cortical defects of form and shape perception are at the root of the defect.

## Crowding Phenomenon

Amblyopic eyes have reduced ability to see small objects, particularly with low contrast and more so if the objects are crowded as in letters in a line of Snellens Chart

### **Detection - Amblyopia**

In strabismus, measurement of visual acuity on all visits is the only way of early detection of amblyopia. Strabismic amblyopia is diagnosed **when the vision in one eye is two lines or more worse compared with the fellow eye.**

In amblyopia visual acuity may be reduced to 20/80(6/24) but is seldom worse. Visual acuity of 20/200(6/60) or less should raise the suspicion of anisometropic amblyopia or other ocular disease.

## Visual Acuity Measurement

Visual acuity measurements are still the gold standard for the evaluation of the patient with amblyopia. Of all the visual acuity measurements, Snellen measurements are preferred due to the "Crowding Phenomenon" value.

## Onset of amblyopia

From a clinical standpoint, strabismic amblyopia rarely develops after age 6 to 8 years

### **Management of Amblyopia**

#### Part-time Patching

Occlusion of the fixing eye is the standard therapy for equalizing visual acuity in amblyopic patients. In total occlusion the occluded eye may develop amblyopia if treated too long. Weekly follow up is advised.

#### Full-time, Alternate-Day Occlusion

This could keep the "binocular slate clean" and help to prevent amblyopia and anomalous retinal correspondence, both of which require some degree of binocular interaction.

Patching therapy needs to be continued in the management of strabismic amblyopia till visual acuity is equalized. Slippage of vision after discontinuance of patching is known. Re-patching is required for recovery.

#### Penalization

Penalization by cycloplegic drops in the fixing eye, may be enough in patients with mild to moderate amblyopia.

## Infantile Esotropia

Infantile Esotropia also called Congenital Esotropia and Essential Infantile Esotropia (EIE) was referred to in the past as congenital esotropia but is rarely, congenital and is more appropriately designated Essential Infantile Esotropia.

It is characterized by a large, stable deviation, a limited potential for single binocular vision, association with oblique muscle dysfunction and dissociated vertical deviation. In addition, may show latent or manifest nystagmus.

### **Presentation - I**

Affected children typically have an esodeviation of 20 to 35 degrees, with an average refractive status for their age. , rarely amblyopic. Good vision in either eye, as demonstrated by an ability to alternate fixation.

### **Presentation - II**

In addition, affected children show a habitual limitation of abduction and a "cross fixation" i.e. preferring to fix objects in their left visual field with their right eye, and vice versa, Features of "inferior oblique" over action.

### **Management of EIE**

These children are rarely amblyopic, have free alteration of fixation and they do not have any significant refractive error. If amblyopia or refractive error at all present, they be treated before surgery.

#### **EIE Surgical Management**

Is invariably surgical. Many authorities believe that unless, an anesthetic problem, early surgery by 2 years of age offers the best prospect for the development of single binocular vision and achievement of stable, long-term, ocular alignment

#### **Over-action inferior oblique**

Because EIE is so commonly associated with over-action of the inferior oblique muscles, it may be necessary to deal with this over-action either at the time of surgery of the horizontal muscle or as a separate surgical procedure

Simultaneous correction of oblique dysfunction will increase the frequency with which stable binocular single vision is achieved post operatively

#### **Divergent Vertical Deviation( DVD)**

It is worth bearing in mind that the child with EIE plus Divergent Vertical Deviation(DVD) is most unlikely to develop useful binocular vision, and that the surgeon is dealing with an essentially cosmetic defect.

## Esotropia Secondary to Ocular Disease

Strabismus in children with ocular disease is equally divided between esotropia and exotropia. So long as the ocular pathology is sufficient to degrade the vision in one eye below a level of 6/24 to 6/36 (20/80 to 20/120), strabismus is likely to result.

## Monocular deviation

With monocular pathology, the importance is on diagnosis and management of the disease rather than the strabismus. In children with retinoblastoma, 15% presented initially with strabismus (Parks, 1992).

In some older patients removing the underlying pathology will also eliminate the strabismus but in children it requires strabismus surgery if they are to achieve binocular vision. .

### **Cautionary note**

In children with deviation in one eye it is wise to operate only in the effected eye. Scleral perforation with its vision-threatening complications are reported in at least 3% of strabismus procedures (Morris et al., 1990),

## Accommodative Esotropia

Accommodative esotropias arise due to excessive accommodational demand, for a proportionate fixation distance (As in a hypermetrope)

### AC:A

An Emetrope, viewing an object at 1. Meter will converge one meter angle and accommodate 1 D. A hypermetrope of 4 D. will need to accommodate 4. D and yet need to converge 1 meter angle. This strains the synkinetic faculty of accommodation and convergence. binocularity will be broken and esotropia result.

### **Types of Accommodative Esotropia**

**Fully accommodative esotropia** –A Full correction of Hypermetropia eliminates the deviation

**Partially accommodative esotropia** where full correction of hypermetropia does reduce the angle but does not eliminate it.

Fully accommodative esotropia can degrade to partially accommodative Esotropia if attention is not paid to refractive correction and to wards Amblyopia producing factors.

Not all children with hypermetropia will develop an accommodative esotropia. As the level of hypermetropia increases, so does the likelihood of strabismus and amblyopia. Why one child with a +3.00 D refractive error develops esotropia and not another remains obscure. Genetic influences may play a part, here.

### **Investigations**

A careful cycloplegic refraction is the critical step in diagnosis. Adequate cycloplegia is obtained with 2 drops of cyclopentolate 1%. But preferred regimen is atropine 1% ointment 3 times a day for 3 days prior to refraction.

Meticulous correction of both spherical and astigmatic errors in both the eyes is essential in children. An "off axis" refraction can be very misleading... All refractions should be accompanied by a careful fundus examination

### **Management Of Fully Accommodative Esotropia**

A. **Optical** By full refractive error correction of any error over 2D and in higher error near correction of 2to 4 D

B. **Pharmacological.** By use of drug like Pilocarpine (2%-4% ),2 to 4 Times to reduce accommodational demand. The drug should be given a trail of 2-4 months

C. **Surgery**

Discussion has centered on the role of surgery in fully accommodative esotropia. Even with surgery, many children still require spectacles for good vision and good alignment. As such in a fully accommodative strabismus preference is for a non-surgical method of treatment.

### Partially Accommodative Esotropia

When the angle of deviation is reduced (by at least 10 Prism D.) but not corrected with adequate hypermetropic correction, then these cases are considered as "Partial accommodative hypermetropia" This condition is usually seen at the age of 3 yrs or so They frequently need surgery.

### Surgery

The amount of surgery needed will depend on the size of the deviation and the surgical objective. Where amblyopia has been eliminated and the prospects for binocular single vision are good, the goal of surgery should be full correction of the deviation.

### Prognosis

Accommodative Esotropia if treated sufficiently early, carries a good prognosis. Care and management of Amblyopia and refractive error are the main stay for success in treatment of Accommodative Esotropia

## Exodeviation

1. Exotropia in first year of life
2. Exotropia later on in life

- Exophoria
- Intermittent exotropia
- Constant Exotropia

Exotropia may also be described as Excess divergence having greater divergence for distance than for near or insufficient convergence with greater divergence in near than at distance. Exotropia as group account for 25% of all cases of Strabismus in young children

### **Exotropia in the first year of Life**

Upon examination of ocular alignment in neonates, up to 60% have a transient constant exodeviation (for a short period), Up to 13 % have intermittent exotropia and 27% show orthophoria.

The majority of children with congenital exotropia show resolution of the strabismus by the age of 6 months spontaneously there after it may not recede spontaneously.

A link between exotropia, cranio-facial anomalies, homonymous hemianopia, cerebral palsy, congenital 3rd. nerve palsy has been well established

Therefore, the child with persistent exotropia should be evaluated carefully by the pediatrician and neurologist for the possibility of an orbital or skull defect or neurological deficiency.

### Management

Having excluded CNS, craniofacial and ocular pathology, Exotropia that is noted in the first several months of life should be watched without any particular treatment.

In the exotropia alternating fixation, which implies no amblyopia, is the rule. When exotropia persists beyond 5- 6 months of age, intervention may be required. Amblyopia is uncommon in congenital exotropia.

The exodeviations can be categorized as exophoria, intermittent exotropia, and constant exotropia. Exophoria denotes a tendency for the visual axes to diverge. This tendency is held in check by fusion, and binocularity.

Intermittent exotropia involves a condition in which the eyes may be diverged at times but aligned at other times. Constant exotropia describes a condition in which there is continuous divergence of the visual axes.

Exotropia can also be categorized as excess divergence (greater deviation in the distance than at near) or insufficient convergence (greater deviation at near than at distance). Excess divergence deviation is characteristic of intermittent exotropia.

### **Epidemiology**

When the exotropias are grouped together they account for approximately 25% of cases of strabismus in young children Exotropia appears within the first few years of life. In most

cases, if left untreated, most of them are said to progress throughout the life-span.

About 15% are said to show improvement over time. Exotropia may begin with exophoria and, with the addition of binocular suppression, develop into intermittent and then constant exotropia.

Women develop exotropia slightly more commonly than men. Most children who have exotropia show refractive errors that are similar to those in age-matched children who do not have strabismus. Refractive error has little influence over exotropia..

### ***Management Of Intermittent Exotropia***

Amblyopia and Refractive error if present be treated though they do not constitute common causative elements. Amblyopia when it occurs it does so in the setting of anisometropia.

Part time Occlusion may help in constant uniocular exotropes. Deliberate over-misusing is said to be helpful in children and young adults who have intermittent exotropia or symptomatic exophoria by producing accommodative convergence.

### **Prism Therapy**

A role of a base-in prism to correct intermittent exotropia or symptomatic exophoria exists for deviations of approximately 20 prism diopters and less. Though difficult but if children are made to use prism glasses results are good.

### **Fusional Convergence Training**

There is a role for orthoptics training in the management of exophoria. The goal of orthoptics training is to increase fusional convergence with targets that do not allow the patient to use accommodative convergence.

Treatment should be directed toward symptoms, which usually are asthenopic in nature. The mere presence of exophoria (without symptoms) is not an indication for treatment.

### **Surgery**

Surgery for intermittent exotropia in children under 5 years of age is associated with the risk of an overcorrection and possible induction of microtropia and even loss of stereopsis.

## **Superior Oblique Palsy**

Most cases of superior oblique palsy (SOP) are traumatically induced. The trochlear nerves exit from the dorsal midbrain and can be damaged by contrecoup forces that are transmitted to the free tentorial edge

Another cause of SOP is ischemia, which can occur in patients with hypertension or diabetes mellitus. The palsy may be isolated or may occur in conjunction with other neurologic problems.

Occasionally, an acquired SOP will occur because of underlying and serious neurologic disease. Intrinsic midbrain disease, trochlear nerves damage, intracavernous sinus aneurysm can produce SOP, but other contiguous structures are also involved (e.g., oculomotor nerves, fifth cranial nerve).

### ***Diplopia***

Superior oblique palsy causes vertical, some degree of horizontal, and torsional diplopia. The last symptom is often most pronounced on down gaze, in the field of action of the superior oblique muscle.

Vertical diplopia is usually worse in gaze directed toward the uninvolved eye, due to inferior oblique overaction (IOOA), or on head tilt to the involved side.

## **Double Elevator Palsy**

Double elevator palsy refers to the inability to elevate one eye . The problem usually is monocular; seldom are both eyes involved except in cases of bilateral congenital inferior rectus fibrosis..

## **Divergent Vertical Deviation**

DVD almost never causes symptoms. ETIOLOGY the exact cause of DVD is unknown. White (1933) proposed that hyper-function of the superior rectus muscles.

Helveston concluded that "divergent vertical deviation results from a neurologic oculomotor imbalance mediated by a presumed vertical vergence center or centers." This remains the best explanation for DVD, but the exact mechanism whereby DVD develops remains elusive.

## **Strabismus management Inf. Oblique palsy**

Isolated inferior oblique palsy is a rare disorder. The innervation to the inferior oblique muscle travels with the inferior division of the oculomotor nerve, which also supplies the inferior rectus muscle and pupil.

Anything that damages the inferior division of the oculomotor nerve will therefore also affect these other muscles or functions.

Inferior oblique palsy usually occurs with damage to the peripheral portion of the nerve (Scott and Nanken, 1977). Inferior oblique palsy has been reported following viral illnesses and orbital trauma.

### ***Management***

As usual, management is indicated when the patient has symptoms that interfere with normal living activities. Transient double vision

## **Inferior Rectus Palsy**

Inferior rectus muscle dysfunction is seen most commonly as a component of a third cranial nerve palsy. Isolated inferior rectus palsy, is now a well-recognized condition.

The infrequent occurrence of isolated inferior rectus palsy reflects the complex neuroanatomy of the oculomotor nerve.

Mostly compressive, ischemic, or inflammatory third nerve lesions affect the nerve's fascicular portion, located between the oculomotor nucleus in the dorsal midbrain and the nerve's bifurcation into superior and inferior divisions in the anterior cavernous sinus.