

# ***Strabismus***

## Definitions

*Strabismus* (squint): is a misalignment of the eyes.

*Orthophoria*: is a perfect ocular alignment without effort.

*Heterophoria* (phoria): is a tendency of the eyes to deviate and the ocular alignment is maintained with effort.

*Heterotropia* (tropia): is a manifest misalignment of the eyes.

The prefixes *eso* and *exo* imply inward and outward deviations respectively e.g. an *esophoria* is a tendency of the eyes to deviate inward (converge). An *esotropia* is a manifest convergent squint. The prefixes *hyper* and *hypo* imply upward and downward deviation of the eye.

## Ocular movements

1-Ductions: are monocular movements around the vertical, horizontal, and anteroposterior axes. they consist of adduction, abduction, elevation, depression, intorsion and extorsion.

2-Versions: are binocular, simultaneous and conjugate eye movements (in the same direction) they include:

- dextroversion : binocular simultaneous movement of the eyes to the right.
- Laeversion : binocular simultaneous movement of the eyes to the left.
- Dextroelevation: gaze up & right.
- Dextrodepression: gaze down & right.
- Laelevation : gaze up & left.
- Laeodepression: gaze down & left.
- Dextrocycloversion: torsional movement of the superior limbi of both eyes to the right.
- Laeocycloversion: torsional movement of the superior limbi of both eyes to the left.

3- Vergences: are binocular, simultaneous movements of the eyes in the opposite directions. They include convergence and divergence.

## Positions of gaze

I- six cardinal positions of gaze : these are

- 1-dextroversion : right lat. Rectus + left med. Rectus.
- 2- laeversion : left lat. Rectus + right med. Rectus.
- 3- dextroelevation: right sup. Rectus + left inf. Oblique.
- 4- laelevation: left sup. Rectus + right inf. Oblique.
- 5- dextrodepression: right inf. Rectus + left sup. Oblique.
- 6- laeodepression: left inf. Rectus + right sup. Oblique.

II- nine diagnostic positions of gaze: they include the above six + the primary position, elevation and depression.

## ***Functional consequences of strabismus***

**Amblyopia:** is the unilateral or rarely bilateral decrease of best-corrected visual acuity caused by form vision deprivation and/or abnormal binocular interaction, for which there is no pathology of the eye.

Types of amblyopia:

- 1- strabismic: due to continued monocular suppression of the deviating eye.
- 2- Anisometropic: caused by a difference in refractive error of even as little as 1.0 dioptre sphere. It results from abnormal binocular interaction from the superimposition of focused and unfocused images or from superimposition of large and small images.
- 3- Stimulus deprivation; results from form vision deprivation due to opacities in the media e.g. cataract or severe ptosis.
- 4- Iso-ametropic: bilateral and caused by high symmetrical refractive errors, usually hypermetropia.
- 5- Meridional: caused by uncorrected astigmatism, could be unilateral or bilateral.

Diagnosis

- 1- visual acuity: a difference in best-corrected visual acuity of two lines or more is indicative of amblyopia. V.A. is worse when reading a row of letters than a single letter this is called crowding phenomenon.
- 2- Neutral density filter: it will decrease V.A. by two lines in a normal eye but not in an amblyopic eye.

Visual fields and color vision are normal in amblyopia.

Treatment

The sensitive period during which amblyopia could be reversed is up to 7-8 years in strabismic amblyopia and longer (up to 11-12 years) for anisometropic amblyopia.

- 1- occlusion of the normal eye to encourage use of the amblyopic eye.
- 2- Penalization: in which vision in the normal eye is blurred by atropine. used in mild amblyopia in association with hypermetropia.

## ***Confusion and diplopia***

Basic concepts

- 1- binocular single vision involves the simultaneous use of both eyes with bifoveal fixation. Conditions necessary for BSV are
  - i. overlap of visual fields
  - ii. accurate neuromuscular development and coordination, so that the visual axes are directed at the object.  
(visual axis is the line of vision which passes from the fovea to the point of fixation)
  - iii. normal visual pathway.
  - iv. approximately equal image clarity and size in both eyes.
  - v. corresponding retinal points.

*Confusion*: simultaneous appreciation of two dissimilar images caused by stimulation of corresponding points by images of different objects.

*Diplopia*: simultaneous appreciation of two images of the same object caused by stimulation of a non corresponding retinal points.

*Projection*: is the interpretation of the position of an object in space on the bases of stimulated retinal elements e.g. if an object stimulate the right fovea it will be interpreted by the brain as having originated from the straight ahead position and if another object lies in the nasal field it will stimulate the temporal retina and any object stimulate the temporal retina will be interpreted by the brain as having originated in the nasal field.

*Corresponding points*: are areas in each retina stimulation of them gives the same projection of the object in the space i.e. no diplopia will occur. The right nasal retina has corresponding points in the left temporal retina. Both foveae are corresponding pointsw.

*Streopsis*: is the perception of depth (3<sup>rd</sup> dimension)

### ***Sensory adaptation to strabismus***

This occurs in children under 6-8 years but rarely in adult in order to get rid of diplopia and confusion. It involves:

- 1- suppression: active inhibition, by the brain of an image from one eye when both eyes are open.
- 2-Abnormal retinal correspondance: a condition in which the fovea of the normal eye will be the corresponding point of extrafoveal area in the deviated eye.so that no diplopia will occur.

### ***Motor adaptation to strabismus***

It involves the adoption of an abnormal head posture and occurs in adults who cannot suppress or in children with good binocular vision.

## **Clinical evaluation**

history:

- 1-age of onset may indicate aetiology.
- 2-variability: intermittent strabismus indicates some degree of binocularity, an alternating deviation idicates symmetrical visual acuity in both eyes.
- 3-general health e.g. cerebral palsy is associated with increased incidence of strabismus.
- 4-birth history.
- 5-family history: strabismus is frequently familial although there is no definitive inheritance pattern.

*visual acuity*: is the ability to distinguish separate element of a target.

### **Testing of VA in preverbal children:**

- 1-occlusion of one eye, if strongly objected by the child indicates poorer acuity in the other eye.
- 2-fixation test by using a 16 prism diopter base-down prism.
- 3-hundreds & thousands: if the child is able to see and pick up small sweets at 33 cm, visual acuity is at least  $6/24$ .
- 4-rotation test by rotating the child briskly through 360 degree this will induce nystagmus. If nystagmus stops when rotation is stopped then VA is good.
- 5-preferential looking test infants prefer to look at a pattern rather than a homogenous stimulus.
- 6-visual evoked potential.

### **Testing VA in verbal children**

- 1-at age 2 years: VA is tested by picture naming tests.
- 2-at age 3 years: matching of single letter charts.
- 3- at age 4 years: Snellen acuity test.

*Tests for stereopsis:* e.g. Titmus test by using two plates in the form of a booklet which are viewed through special spectacles on the right there is a large fly which should appear solid and the child is encouraged to pick up one of its wings. On the left are a series of circles and animals one of the circles and one of the animals appears forward in the presence of normal stereopsis.

*Tests for sensory anomalies e.g.:*

- 1-Worth four-dot test: the patient wears a red lens in front of the right eye, a green lens in front of the left eye & views a box with one red, two green and one white light. A normal person should see the 4 lights.
- 2-synoptophore: an instrument used for assessing strabismus, testing binocular vision and detection of suppression & abnormal retinal correspondence.

*Grades of binocular vision (by using the synoptophore)*

- 1-first grade (simultaneous perception).
- 2-second grade (fusion) is the ability of the two eyes to produce a composite picture from two similar pictures.
- 3- third grade (stereopsis)

*Measurement of deviation*

- 1- Hirschberg test: a pen torch is shone to the eyes from arm's length and the patient is asked to fixate the light. The corneal reflection of the light will be centered in the pupil of the fixating eye but will be decentered in a squinting eye every millimeter deviation is equal to 7 degrees e.g. if the reflex is situated at the temporal border of the pupil the angle is about 15 degrees if it is at the limbus, the angle is about 45 degrees.

2- cover tests: the most accurate they include:

a- cover-uncover test; the cover test is to detect heterotropia. If right deviation is suspected the examiner covers the left eye and notes any movement of the right eye no movement indicates orthophoria, adduction of the right eye to take up fixation indicates exotropia & abduction indicates esotropia. The uncover test is to detect heterophoria in which the patient fixates a distant target and the examiner covers the right eye and after few seconds removes the cover. No movement indicates orthophoria. Adduction indicates exophoria, and abduction indicates esophoria. the test is repeated for the opposite eye

b- alternate cover test: interrupts binocular fusion mechanisms and reveals the total deviation (tropia + phoria). The right eye is covered for about two seconds then the occluder is quickly shifted to the opposite eye for two seconds, then back and forth several times.

c- prism cover test: precisely measure the angle of deviation.

3-dissimilar image tests:

a- Maddox wing to measure heterophoria for near.

b- Maddox rod it measures the total deviation (tropia+phoria).

#### *Refraction and fundoscopy*

Dilated fundoscopy is mandatory in strabismus to exclude macular scar, optic disk hypoplasia, and retinoblastoma. Strabismus is often secondary to refractive errors like hypermetropia, astigmatism and myopia.

Cycloplegia: is the paralysis of the ciliary muscle. Hypermetropia is the commonest refractive error causing strabismus and accurate measurement of hypermetropia necessitates cycloplegia to eliminate accommodation which masks the true degree of this refractive error. Drugs used to achieve cycloplegia are:

1-cyclopentolate drops 0.5% under age 6 months and 1% thereafter.

2-atropine drops 0.5% under age of 1 year and 1% thereafter. The drops are given three times daily 3 days before refraction.

It is important to repeat the refraction every 6 months.

#### *Investigations of diplopia*

The Hess test and Lees screen plot ocular position as a function of the extraocular muscles and enable differentiation of paretic strabismus (caused by neurological pathology) from restrictive myopathy (as in thyroid eye disease or blow-out fracture of the orbit).

### ***Esotropia (manifest convergent squint)***

It may be comitant or incomitant. In a comitant esotropia the angle of deviation is the same in different horizontal gaze positions. In an incomitant deviation the angle differs in various positions of gaze as a result of abnormal innervation or restriction.

**Accommodative esotropia.** Near vision involves both accommodation and convergence. Accommodation is the process by which the eye focuses on a near target, by altering the curvature of the crystalline lens. Simultaneously, the eyes converge, in order to fixate bifoveally on the target. Both accommodation and convergence are quantitatively related, and have a fairly constant relationship to each other (AC/A ratio) abnormalities of this ratio are an important cause of certain types of esotropia.

Types of accommodative esotropia:

1. refractive type here the AC/A ratio is normal and esotropia is due to excessive hypermetropia usually between +4.00 and +7.00 diopters. The considerable degree of accommodation required to focus clearly on even a distant target is accompanied by a proportionate amount of convergence and a manifest convergence squint results. The deviation presents at about the age of 2.5 years.
2. non-refractive type: this is associated with a high AC/A ratio in which a unit increase of accommodation is accompanied by a disproportionately large increase of convergence in the absence of significant hypermetropia.

#### **Management**

- 1- Refractive error should be corrected.
- 2- bifocals may be prescribed if there is high AC/A ratio.
- 3- miotic therapy can be used short term in children with accommodative esotropia due to high AC/A ratio, who will not wear spectacles. miotic therapy works by inducing peripheral accommodation (direct stimulation of the ciliary muscle). Less accommodative effort is therefore required by the patient for near vision and thereby less accommodative convergence is induced.
- 4- treatment of amblyopia.
- 5- surgery if spectacles do not fully correct the deviation. the principle of surgery involves weakening of medial recti.

#### **Essential infantile esotropia**

An idiopathic esotropia developing in the first 6 months of life in an otherwise normal infant with no significant refractive error and no limitation of ocular movement.

#### **Signs**

- 1- the angle is usually large >15 degrees.
- 2- Cross fixation in which the child uses the right eye in left gaze and the left eye in right gaze. Such cross-fixation may give a false impression of bilateral abduction deficits, such as with bilateral 6<sup>th</sup> nerve palsy which can be excluded by rotating the child or patching of one eye for a few hours.
- 3- Nystagmus.
- 4- Inferior oblique overaction.

5- Poor potential for binocular single vision.

#### Management

Ideally the eyes should be surgically aligned by the age of 12 months, but only after amblyopia have been corrected. the initial procedure is recession of both medial recti. Under correction may require further recession of the medial recti or resection of one or both lateral recti. Surgery for inferior oblique overaction may be required.

### ***Exotropia***

Is a manifest divergent squint which may be constant or intermittent.

#### *Constant exotropia:*

##### Types

- 1-congenital exotropia is present at birth and characterized by normal refraction, large and constant angle of deviation. It is treated surgically by bilateral lateral rectus recession, usually combined with resection of one or both medial recti.
- 2-sensory exotropia is the result of monocular or binocular visual impairment by acquired lesion, such as cataract or opacity in the media, in children over the age of 5 years or in adults. Treatment by correction of vision if possible then surgery.
- 3-consecutive exotropia follows surgical correction of an esotropia.

#### *Intermittent exotropia*

It presents most frequently at around 2 years with exorhoria, which change to exotropia under conditions of visual inattention, fatigue or ill health. with time the deviation will be more evident.

##### Types

- 1-basic exotropia where the angle of deviation is the same for near and for far vision.
- 2-convergence weakness: the angle is greater for near.
- 3-divergence excess: the angle is greater for distance.

#### Management

- 1-spectacle correction in myopic patients may control the deviation by stimulating accommodation and, with it, convergence.
- 2-orthoptic treatment e.g. occlusion.
- 3-surgery is necessary in most patients by about the age of 5 years in the form of recession of both lateral recti.