Ocular Motility in Health and Disease

Contents:

- Extraocular Muscles
- Eye Movements
- Single Binocular Vision
- Strabismus
- Amblyopia

Objectives:

By the end of this course the undergraduate student should be able to:

- Detect strabismus by general inspection, the corneal light reflex test, and the cover test.
- Know the basic types of strabismus and be able to differentiate between them, and recognize which cases need prompt management.
- Understand the rationale of different lines of management, in order to point out their indications and limitations.
- Know when amblyopia should be suspected and when it must be prevented and treated.
The Ocular Motility and Squint (Strabismus)

THE OCULAR MOTILITY

EXTRAOCULAR MUSCLES

There are six extraocular muscles in each eye which control their movements.

I. The Recti Muscles.

Fig. (7.1) The extrinsic muscles of the eye

The four recti muscles originate from the common annular tendon of Zinn situated around the optic foramen at the apex of the orbit, and insert into the sclera by flat tendons at various distances from the limbus.

II. The Oblique Muscles

The superior oblique originates from the orbital apex above the annulus of Zinn and passes anteriorly and upwards along the superomedial wall of the orbit. It passes through the trochlea (a fibrous pulley), and inserts into the upper and outer part of the sclera behind the equator.

The inferior oblique originates from the floor of the orbit near the inferior orbital margin lateral to the orifice of the lacrimal fossa. It passes laterally, superiorly and posteriorly below the inferior rectus muscle, and inserts into the outer part of the sclera behind the equator.
The 3\textsuperscript{rd} cranial nerve (oculomotor), is the motor nerve supply of all the extraocular muscles except: 1) the superior oblique muscle, that is supplied by the 4\textsuperscript{th} cranial nerve (trochlear), and 2) the lateral rectus muscle supplied by the 6\textsuperscript{th} cranial nerve (abducent).

Muscular branches of the ophthalmic artery are considered as the main blood supply.

Action of the Extraocular Muscles

These muscles rotate the eye around the ‘centre of rotation’, situated 12-13 mm behind the cornea in a horizontal plane.

Three types of movements are possible around the centre of rotation.

1. Movements around the vertical axis whereby the eye is turned from side to side (abduction and adduction).
2. Movements around the horizontal axis whereby eye is turned up wards or downwards (elevation and depression).
3. Movements around the anteroposterior axis whereby the movement of ‘torsion’ occurs. Torsion includes: a) intorsion which means rotation of the upper pole of the cornea nasally and b) extorsion which means rotation of the upper pole of the cornea temporally.

The only action of the medial rectus is adduction. Similarly, the only action of the lateral rectus muscle is to abduction. For the rest of the muscles, more than one action is described with a main (primary) action and secondary (subsidiary) actions, depending on the position of the eye while initiating the
movement. This physiologic fact is based on the anatomical relationship of the muscle axis to the axis of the vision (visual axis). Figure(7.3)

![Figure (7.3): the relation between muscle axis and visual axis](image)

**Orbital axis, muscle axis and visual axis:**

The lateral and medial walls of the orbit make an angle of 45° with each other. The orbital axis therefore forms an angle of 22.5° (23 for simplicity) with either the lateral or medial walls. Because most extraocular muscles (5 out of 6) originate from the orbital apex, the orbital axis is considered to represent the muscle axis too. When the eye is looking straight ahead (primary position of gaze), its visual axis forms an angle of 23° with the orbital axis (muscle axis). The actions of the extraocular muscles depend on the position of the globe at the time of muscle contraction. The primary action of a muscle is its major effect when the eye is in the primary position and its subsidiary actions are the additional effects exerted on the eye in the same (primary) position.

Therefore, the **superior rectus elevates** the eye (primary action), as well as **adducts and intorts** it (subsidiary actions), and the **inferior rectus** acts as a **depressor** (primary action), together with **adduction** and **extorsion** (subsidiary actions).
The **superior oblique** is an eye intorter (**primary action**), as well as _depressor_ and _abductor_ (**subsidiary actions**), whereas, the **inferior oblique** extorts (**primary action**), as well as _elevates_ and _abducts_ (**subsidiary actions**) the eye.

**Muscle axis and the cardinal directions of gaze:**

Based on the aforementioned anatomical facts, the primary action of a specific EOM can only be tested when the eye is positioned so that the visual axis coincides with the axis of that particular muscle. Accordingly we have 6 directions of gaze where only one muscle is responsible for the specific eye movement. These are shown in (figure 7.4)

Moreover, the primary position, the straight upwards and straight downwards positions may be added i.e. 9 cardinal positions (Figure 7.5)

![Fig.7.4: The 6 cardinal gaze directions](image-url)
Monocular eye movements are called ductions and include, adduction, abduction, elevation, depression, intorsion and extorsion whereas binocular eye movements are either versions or vergences. Versions is the term describing situations when the two eyes move in the same direction with the visual axes parallel to each other. An example is right medial rectus and left lateral rectus, moving both eyes to the left gaze. Vergences describe situations when the two visual axes of the two eyes move opposite to each other as in convergence for near vision. The opposite of convergence is divergence.

**BINOCULAR SINGLE VISION**

The eye is a paired organ. Each eye forms a separate image. When the two eyes are aligned (have the same direction), both eyes will see the same object. The image received by the two eyes is similar but not exactly the same. This is due to the slight lateral displacement of the two eye globes. Each eye sees the object from a slightly different angle. The two images are fused together into one single image in order to avoid diplopia (double vision) which is extremely disturbing. Fusion occurs in the brain. The slight difference between the two images gives the fused image its 3 dimensional feature i.e. stereopsis which is the biggest contributor to depth perception. A large part of studying the diagnosis and the management of strabismus is directed towards preserving the important advantage of binocular single vision. It is noteworthy in this context to be aware
that, in the normal state, binocular single vision is acquired and reinforced during the first months of life.

Note:

Having the eyes set anatomically close together is a feature of humans and higher animals giving them the advantage of stereoscopic vision. Apparently, this is more important than having a wide field without stereopsis as observed in other members of the animal kingdom where the eyes are anatomically set to the sides of the head.
SQUINT (Strabismus)

Under normal conditions, both eyes move synchronously and meet at the object of regard (fixation point). Strabismus (squint) is the condition where one eye deviates away from the fixation point (ocular misalignment).

Examination of a case of squint:

Proper history taking is very important in every case of strabismus, that help in the diagnosis as well as for the prognosis.

<table>
<thead>
<tr>
<th>History taking in a case of strabismus</th>
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<tbody>
<tr>
<td>1. Age of onset</td>
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<td>• If convergent squint appears before 6 months of age it is termed congenital esotropia which is a special entity to be dealt with differently by the pediatric ophthalmologist. This particular situation requires surgery in the first year of life.</td>
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<tr>
<td>• Generally the later the onset the better the prognosis</td>
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<td>3. Is the squint intermittent or constant?</td>
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<td>4. Is there deviation of only one eye or alternate eye?</td>
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<td>5. Family history of squint or refractive error is useful.</td>
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<tr>
<td>6. History of glasses or previous strabismus surgery.</td>
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2) Testing the ocular motility in the 6 cardinal directions of gaze to exclude paralysis or paresis.

Assessment of ocular motility

- Monocular movements (Duction)
  Here ocular motility of each eye is assessed alone , by asking the patient to follow a moving target with one eye closed.
- **Binocular Conjugate movements (Versions):** Here ocular motility of both eyes are assessed together by asking the patient to look at a moving target (A pen or finger) in the 6 cardinal directions of gaze with both eyes moving towards the same target (figure 7.8)

- **Binocular disconjugate movements (Vergence):** Here ocular motility of both eyes are assessed together by asking the patient to look at a moving target (A pen or finger) as it comes closer and then moves farther from the patient's line of sight. Here both eyes will converge and then diverge, thus moving in opposite directions (figure 7.9)

![Fig 7.8: Assessing binocular ocular movements in the 6 cardinal directions of gaze](image)

![Fig 7.9: Testing for Convergence](image)

3) Observing the corneal light reflex in order to roughly measure the angle of deviation. This is more accurately measured by the use of prisms.(figure 7.11)
Assessment of angle of strabismus: (Hirschbirg test - Corneal light reflex test):

- The patient is asked to look at a distant target.
- Light is shone at both eyes.
- The position of the reflex of light on the surface of the cornea is observed in relation to the pupil.
- It should be symmetrical and centered in the pupils (Orthophoria)
- If the light is not centered, squint is present. (Heterotropia)
- According to the position of the corneal light reflex to the center of the pupil, the angle is roughly measured. (See figure 7.10)

Fig. 7.10: Corneal light reflex test
4) Performing the cover-uncover test in order to differentiate between unilateral and alternating squint (see cover test)
The monocular cover-uncover Test

Principle:
Fusion of the two eyes is broken by covering one eye.

Indications:
To detect manifest strabismus and for differentiating a heterophoria from a heterotropia.

Method:
The patient is asked to look at (fixate) a distant object. If the patient is a small child his attention may be drawn to a game or a TV screen. Ideally the cover-uncover test should be done again with the patient fixing a near (accommodative) object.

Steps:
1) After fixation, you start by covering the fixing eye or the eye which you think is fixing. You observe the eye movement of the uncovered eye if any.
2) Then you remove the cover and observe the eye under cover for the presence and the direction of eye movement
3) You then repeat the test again for the other eye.

Interpretation: Figure(7.7)
1. If there is no movement, both for the covered and the uncovered eyes, then the patient has orthophoria.
2. If the uncovered eye moves, while the other eye is covered, then the patient has manifest strabismus (Heterotropia)
3. In cases with heterotropias, if the patient started with one eye is deviated and after testing ends with the same eye, then it is a case of unilateral tropia, whereas in alternating tropia, the test will end with the opposite eye.
4. If the uncovered eye does not move, then observe the eye under cover, if it is deviated then the patient has latent strabismus (Heterophoria)
5. Observe the movement of the covered eye while removing the cover;
   • If there is outwards movement on removing the cover the patient has esophoria.
   • If there is inwards movement on removing the cover the patient has exotropia.
5) Testing for the presence of errors of refraction by retinoscopy or autorefractometry according to age. Cycloplegic refraction is a must.

The relationship between strabismus and errors of refractions:
Accommodation and convergence are closely related. They occur together for near work for every diopter of accommodation needed there is a corresponding amount of convergence exerted (accommodative convergence). It follows that when there is excessive accommodation as in hypermetropia there is also excessive convergence. The result is tendency of the eyes to deviate inwards (esophoria and/or esotropia). This is commonly seen in children because the child’s eye is still a small eye i.e. hypermetropic. The type of concomitant esotropia caused by this is termed **accommodative esotropia**. This type is often fully corrected by optical correction alone. Likewise, eyes with high myopia may present with exophoria or exotropia due to weak or absent accommodation with subsequent weak convergence and tendency for the eyes to deviate outwards.
6) Ocular examination especially fundus examination is an essential part of examination (see box).

- Fundus examination is an essential step in every strabismus case.
- Fundus lesions may be the primary cause of strabismus e.g. macular scars
- Strabismus may be the first presentation of other pathologies e.g. retinoblastoma, papilloedema.

**Classification of squint**

- Strabismus
  - False
  - True
    - Latent (heterophoria)
    - Manifest (heterotropia)
      - Concomitant
      - Non-concomitant
1. APPARENT STRABISMUS (Pseudo-strabismus)

It is a condition in which a false impression of ocular deviation is present but on examination the eyes are straight (orthophoric), with central corneal light reflex and no movement on doing cover test *(see later).* The situation is usually caused by some normal anatomical variations in early childhood such as the presence of epicanthus fold (figure 7.6) or the presence of a small or large interpupillary distance. The presence of high errors of refraction may give the false impression of squinting eyes. The condition requires no special treatment. The role of the physician is just to reassure the parents after making sure that the eyes are normal.

![Fig. 7.6; Epicanthus fold (Apparent convergent)](image)

2- LATENT STRABISMUS (Heterophoria)

Latent squint or phoria is the tendency of the eye to deviate. This tendency is, normally and unconsciously, prevented by the brain for the sake of maintaining binocular single vision. This condition is quite common in the general population and is usually asymptomatic (compensated). However, sometimes patients complain of eyestrain (asthenopia) especially after visual exertion or fatigue as in prolonged near work. Eyestrain symptoms include headache,
ocular or periorbital pain, chronic eye redness, feeling of tiredness or blurring of prints and overlapping of words or lines while reading (running of the letters). If these symptoms persist or when the latent squint turns into manifest we call it **decompensated**.

Still the role or errors of refraction to be considered in cases with decompansated phorias;

- **Hypermetropia** → Esophoria (Excess accommodation)
- **Myopia** → Exophoria (Weak accommodation)

There are many tests to diagnose and even measure phorias. The simplest and most commonly used test is the monocular cover/uncover test. This test is extremely important in diagnosing other types of squint as well. (see box for the cover-uncover test)

As a rule, asymptomatic cases of phoria require no treatment. Errors of refraction should be corrected. Exercises to increase the fusional ability (fusional reserve) may be advised. If exophoria deteriorates so that a manifest deviation occurs intermittently at frequent intervals (intermittent exotropia) surgical intervention is needed.

### 3- MANIFEST STRABISMUS (Heterotropia)

In manifest strabismus the deviation of the eye is evident. It is of two main types namely **concomitant** and **incomitant (paralytic)** strabismus.

#### 1. Concomitant strabismus

In concomitant deviations, the deviation remains the same in all the directions of gaze. The condition may be **congenital** (esotropia is much more common than exotropia), or **acquired**. **Acquired deviation** may be attributed to **refractive** errors (*Hypermetropia is associated with esotropia & Myopia is*
associated with exotropia) or may be sensory (caused by impaired vision in one eye).

The hallmark of concomitant squint (as compared to paralytic squint), is the absence of limitation of ocular motility. Concomitant squint may be unilateral (when one eye is always deviating) or alternating if the 2 eyes alternately deviate. Unilateral deviations indicate presence of poor vision in the deviating eye (amblyopia).

**Treatment of concomitant strabismus:**

1) Correction of the error of refraction (reference to the box of relationship with errors). In many situations this is the only treatment needed e.g. accommodative squint.

2) Treatment of amblyopia if present by * occlusion of the sound eye (either full time or part time) or by * blurring its image by atropine (penalization). See later

3) Surgery

The principle is to weaken the strong muscle and/or to strengthen the weak muscle in order to achieve alignment of the 2 eyes. The most commonly used weakening procedure is recession and the most commonly used strengthening procedure is resection. As an example, in esotropia, the medial rectus is the muscle to be weakened and the lateral rectus is the muscle to be strengthened.
It must be noted that surgery and wearing eye glasses are not mere alternatives. Parents cannot decide to choose surgery because they find it difficult to make their child wear his correction. (Figure 7.12)

Fig. 7.12: Surgical technique for extraocular muscle surgery

2. PARALYTIC STRABISMUS

In paralytic strabismus, deviation of the eye is caused by paralysis or paresis of one or more of the extraocular muscles. The deviation of the eye varies in different directions of gaze. The patients commonly present with a manifest ocular deviation, binocular diplopia (seeing double objects), that is most marked in the direction of action of the paralyzed muscle, and limitation of ocular motility.
Causes of Paralytic Strabismus:

1. Lesions of the motor nerve nucleus.
   a. Congenital absence of the nucleus.
   b. Inflammations such as encephalitis, disseminated sclerosis.
   c. Degenerative and vascular lesions.

ii. Lesions of nerve trunk
   d. Trauma by direct injury or by pressure.
   e. Inflammations such as diabetes mellitus neuropathy.

iii. Lesions of the muscle.
   f. Congenital absence or mal-development of the muscle.
   g. Direct injury.
   h. Myopathy.

On examination, there is an ocular deviation (*The eye deviates to the opposite direction of action of paralysed muscle*). Limitations of movements, and false projection (*the patient is not able to grasp or point the object correctly*) are more evident in the direction of action of the paralyzed muscle.

Frequently, the patient may present with an abnormal head posture (the patient’s head and face is turned towards the direction of the action of the paralysed muscle) to avoid symptomatic diplopia. Diplopia often occurs in paralytic squint and not in concomitant squint.
Compensatory Mechanisms for Diplopia

1. Suppression- It is an adaptation which occurs mainly in children. The visual cortex in the brain neglects the blurred image it receives from the deviating eye. (the image is blurred because it does not fall on the fovea). Suppression occurs more easily in children because of plasticity of their brains. This is often the precursor of amblyopia.

2. Abnormal head posture

Identifying the affected muscle(s), is an essential step in examining a case with paralytic strabismus (examination of ocular duction & version) in order to identify the affected cranial nerve.

OCULAR CRANIAL NERVES PALSY

3rd Nerve Palsy

The 3rd cranial nerve (oculomotor) supplies the levator, superior rectus, inferior rectus, medial rectus, inferior oblique, sphincter pupillae and ciliary muscles. Complete paralysis presents with ptosis (prevents diplopia), with limitation of upwards, downwards and inwards movements. The eye adopts the abducted and slightly downward position, and the pupil is dilated and fixed, with accommodation completely lost. In incomplete paralysis, an individual muscle may be involved. (Figure 7.13)
Intracranial aneurysm (especially of the posterior communicating artery), has been considered as the most common cause of isolated third nerve palsy. Other causes include; trauma, cerebral herniation, and brain tumor. Moreover microvascular infarction within the nerve (commonly sparing the pupil), may be the cause in diabetic and hypertensive patients.

In a patient with 3rd nerve palsy, unless the clinical evidence overwhelmingly supports microvascular disease, emergent cerebral imaging with angiography must be obtained,

1) The presence or absence of pupillary dilatation is very important to differentiate between surgical causes of 3rd nerve palsy where the pupil is involved (e.g. aneurysms) and medical causes (e.g. diabetic neuropathy) where the pupil is spared.

4th Nerve palsy

The 4th nerve (the trochlear nerve), supplies the superior oblique muscle. Forth nerve paralysis causes limitation of downwards and inwards movement as well as extortion of the eye. It may be congenital (more common) or acquired
Congenital cases may present in adulthood with difficulty in reading binocularly. For acquired cases, microvascular disease (*in diabetic and hypertensive patients*) is considered the most frequent cause in unilateral fourth nerve palsy, whereas, closed head trauma, is the most frequent cause of bilateral cases.

Examination will reveal vertical diplopia (and hypertropia) more in the downward position. The patient may tilt his or her head towards the opposite shoulder to minimize the diplopia. (Figure 7.14 A&B)

![Fig.7.14 : (A)Rt hypertropia (B) Left head tilt](image)

### 6th Nerve Palsy

The 6th nerve (abducent nerve) supplies the lateral rectus muscle, that is considered as the most common type of isolated ocular muscle paralysis. Paralysis of the lateral rectus muscle causes limitation of abduction, with esotropia in the affected eye in the primary position. The patient commonly presents with horizontal diplopia more on attempting abduction. (Figure 7.15)
Fig 7.15: A case of left 6th nerve palsy

In adults older than 55 years of age, especially those who give history of DM and hypertension, microvascular causes should be considered. Intracranial space occupying lesion is responsible for 20% of cases presenting with 6th nerve palsy, especially in adults younger than 45 years. Increased intracranial pressure due to pseudotumor cerebri or hydrocephalus can present infrequently with bilateral sixth nerve palsies. Trauma has been considered as the most common etiology of a sixth nerve palsy in children (42%). Post-viral/para-infectious disease, or inflammation of the petrous ridge from severe otitis media are other causes in children. Because 6th nerve palsy can accompany any increase of the intracranial pressure, it is does not, by itself point to the presence or the site of the space occupying lesion, It is therefore described as a false localizing sign.

- Patients at risk for microvascular disease and those over 55 should initially be managed by observation, as 98% of microvascular palsies spontaneously recover within 3 or 4 months. If recovery does not ensue or if other neurologic abnormalities are present, neuro-imaging is required.
- In adults younger than 45, MRI is indicated initially, as the incidence of a space-occupying lesion increases with decreasing age

Treatment of the basic underlying cause and occlusion of the affected eye (If the diplopia is troublesome) are carried out while waiting for spontaneous recovery.

If no recovery appears after 6 months, surgical intervention is needed.
In patients with incomitant strabismus due to restrictive causes; the condition is mainly due to muscle affection rather than nerve palsy e.g restrictive myopathy in thyroid eye disease, OR due to mechanical entrapment of the muscle, e.g. orbital floor fracture with inferior rectus muscle entrapment.

**Amblyopia**

Amblyopia is defined as a condition of impaired vision in the absence of organic disease. It is usually unilateral.

This may be due to constant unilateral strabismus occurring early in life (*Strabismus amblyopia*), that may lead to suppression which in turn causes amblyopia. Uncorrected anisometropia is another cause (*Anisometropic amblyopia*). Amblyopia may also occur as a result of media opacities e.g. congenital cataract or fundus lesions present at an early age (*Sensory deprivation amblyopia*)

Amblyopia should be considered only when the difference in the best corrected visual acuity between the two eyes exceeds 2 chart lines.

In cases where amblyopia is suspected, patients should be referred for proper evaluation and management, with the main principles of management are 1) *Treatment of the cause*, 2) *Occlusion of the normal eye to force the patient to use the amblyopic eye* and/or 3) *Penalization* (blurring the normal eye by instilling cycloplegic drops thus giving advantage for the weaker eye in near vision). The protocol for occlusion and / or penalization to be set by a specialist according to the age of the patient and the extent of the amblyopia.