Strabismus ,Amblyopia & Leukocoria

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slides
Important
Book
extra

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Resources:
Team 433,
Doctors Notes,
Vaughans and Asbury's General Opthalmology

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Contents

Definitions:

● **Primary gaze**: the position of the eye when the body in normal position and looking straight forward.

● **Binocular vision**: ability of the eyes to focus on one object and then to fuse two images into one.

● **Exo-**: deviation outwards

● **Eso-**: deviation inwards

● **Hyper-**: deviation upward

● **Hypo-**: deviation downward

● **Tropia**: misalignment that is **always there**, even when both eyes are open and attempting to work together. Large angle deviations are obvious. If small angle, you can detect it with the cover-uncover test.

● **Phoria**: misalignment that only occurs **some of the time**, usually under conditions of stress, illness, fatigue or when binocular vision is interrupted i.e. When the two eyes are no longer looking at the same object such as when the synchronization between the eyes is broken by covering one eye. You can "break fusion" using the cross-cover test.

● **Cover test**: determination of the presence and degree of heterotropia by covering one eye with an opaque object and examining for any movement of the uncovered eye to fixate a target.

● **Diopter**: unit of measurement of refractive power of lenses.

Strabismus (squint)

Under normal binocular viewing conditions, the image of the object falls simultaneously on the fovea of each eye (bifoveal fixation) and the vertical retinal meridians are both upright. Any ocular misalignment, such that only one eye views the object of regard with the correct vertical orientation, is called “strabismus.

Strabismus is subdivided into tropia or phoria.
Gold standard procedure for detection: Cover testing (subdivided into two tests)

1- Unilateral cover test (cover-uncover test): detect phoria
2- Alternating cover test (cross-cover test): detect both phoria and tropia

More explanation is found in the examination section.

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Causes

1. Idiopathic
2. Inherited pattern. Most patients fall under this category, so it is important to ask about family history.
3. Neurological conditions (cerebral palsy, hydrocephalus & brain tumors).
4. Down syndrome.
5. A congenital cataract, eye tumor.

Why are we concerned about strabismus?

- Binocular single vision*
- Double vision. Mainly in adults because children and infants have a suppression feature which is not found in adults.
- Cosmetic

Strabismus consequences

- Lazy eye (amblyopia) in children
- Double vision usually in adults but you may see it in children. E.g. if they have a tumor and present with sudden esotropia and diplopia

*Binocular single vision: the state of simultaneous vision, which is achieved by the coordinated use of both eyes, so that separate and slightly dissimilar images arising in each eye are appreciated as a single image by the process of fusion. Thus binocular vision implies fusion, the blending of sight from the two eyes to form a single percept.
It can be:

1. Normal – Binocular Single vision can be classified as normal when it is *bifoveal* and there is no manifest deviation.
2. Anomalous - Binocular Single vision is anomalous when the images of the fixated object are projected from the fovea of one eye and an extrafoveal area of the other eye i.e. when the visual direction of the retinal elements has changed. A small manifest strabismus is therefore always present in anomalous Binocular Single vision.

Esotropia (convergent strabismus):

Before surgery After surgery

Its divided into two types:

1- **Comitant** (paralysis of lateral rectus muscle)
2- **Non-comitant**. Most common in children
    a. Partially accommodative
    b. Accommodative
    c. Nonaccommodative esotropia
        i. Infantile esotropia: Onset at 6 months
        ii. Acquired esotropia: After the age of 6 months

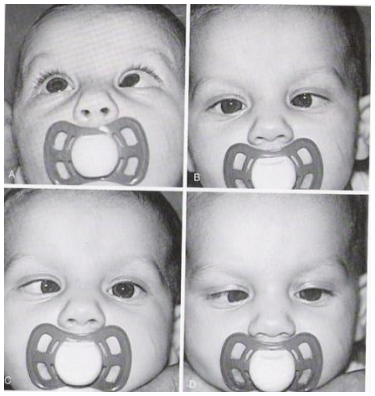
❖ **Comitant esotropia**: occurs at any age due to paralysis of both or one lateral rectus muscle. Paralysis may be due to:

1- Muscle fibrosis
2- tumors
3- neurological. E.g. abducens nerve palsy

This type requires more attention when presented at the clinic.

❖ **Infantile esotropia (congenital):**

Onset by the age of 6 months old. The angle of deviation is unchanged in all direction of gaze. The cause is not related to refractive error or paresis of an extraocular muscle. Most likely cause is due to faulty innervational control, involving the supranuclear pathways for convergence and divergence and their neural connections to the medial longitudinal fasciculus. A smaller number of cases are due to anatomic variations such as anomalous insertions of horizontally acting muscles, abnormal check ligaments, or various other fascial abnormalities.



Features:

- Large Angle. Deviation is often large 40 diopter
- Small hypermetropia.
- Before age of 6months.
- Cross fixation (turning the face to fixate the eye contralateral to the target).

Ocular association of infantile ET: -team 433-

- DVD (dissociate vertical deviation)
- IOOA (inferior oblique oval overaction)
- Latent nystagmus
- Smooth pursuit asymmetry (slow lagging temporally directed on OKN (Optokinetic nystagmus))
- DHD (dissociate horizontal deviation)

Treatment:

Surgery **after** correction of retractive errors. Remember this type cannot be corrected by wearing glasses.

Extra notes:

- Surgically by weakening the **medial rectus** muscles at age of 10-11months to achieve monofixation syndrome.
- surgery must be done as early as possible.
- Surgery should be preformed before the age of 2.
- Monofixation syndrome is a form of subnormal binocular vision without bifixation. It is often desired result of strabismus surgery.
- We must make sure that there is **no refractive error** before doing the surgery.

❖ Accommodative esotropia:

Onset at age 2-3 years or at any age.

Occurs when there is overreaction to the normal mechanism of accommodation.

two mechanism contributes to this type:

1- **Due to hyperopia (refractive).** Refraction errors plays a role in this type. In a child with high hyperopia, the eye requires large accommodation power to bring the object at the fovea to Clarify the image. As a result, they eye deviates inward.

2- **Due to High AC/A Ratio (non-refractive).** The eye deviates the more accommodation power it requires. The eye deviation is prominent at near distance; focusing at a near object required more accommodation. (bring a pencil and put it near you nose. You eye will cross)

Treatment

wearing full cycloplegic refraction glasses.

Aim: relax the ciliary muscle

Note: If the farsighted glasses control the crossing of the eyes, eye muscle surgery is never recommended!

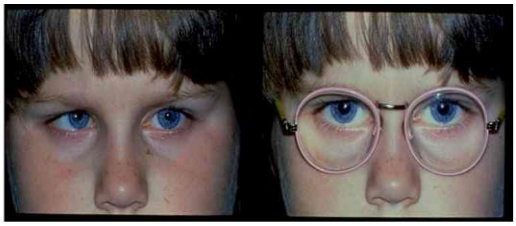
❖ Partially accommodative esotropia:

A mixed mechanism—part muscular imbalance and part accommodative/convergence imbalance—may exist.

To make it simple - A patient was given cycloplegic glasses for his esotropia. In the next few visits he comes back and you find that the angle of deviation is decreased but persist.

Treatment:

Surgery is the choice to **eliminate** the deviation completely.



Hypermetropic ET corrected with glasses

High AC/A ratio ET treated by bifocal glasses

Ophtobook
Treatment of strabismus: Before taking anyone to surgery, correct all the non-surgical causes of strabismus: check for refractive error and treat any amblyopia - many cases of strabismus will improve or resolve by just doing these things. Eye surgery consists of shortening or relaxing the extraocular muscles that attach to the globe to straighten the eye.

Strabismus surgery: To correct simple esotropias (cross-eyed) or exotropias (wall-eyed) we can weaken or strengthen the horizontal rectus muscles. A recession-procedure involves disinserting the rectus muscle and reattaching the muscle to the globe in a more posterior position. This effectively weakens the action of this muscle and turns the eye accordingly.

Infantile ET clinical example:

A 4-month-old healthy child presents with a history of his eyes turning in most of the time, since about 8 weeks of age.
Examination:

- ET for both distance and near 60 PD.
- Extraocular movement (EOM) is Full.
- Cycloplegic retinoscopy is +1.25 D
- Fundus: normal

Pseudostrabismus:

is a condition in which alignment of the eyes is straight; however, they appear to be crossed due to:

- a flat nasal bridge
- prominent epicanthal folds

This illusion of crossed eyes is found in many Asians and young children.
A careful ocular examination (eg, pupillary light reflex, cover test) reveals that the eyes are straight, and the person will have normal corneal light reflexes and no refixation on cover testing.
This appearance gradually disappears as the child ages.

Exotropia (divergent strabismus)

A horizontal form of strabismus characterized by visual axes that form a divergent angle (or in a simpler way: eyes turned outwards)

Exotropia types:

1. Intermittent exotropia
2. Constant exotropia
3. Convergence insufficiency.

1- Intermittent Exotropia

Most **common** type of extropian. Onset maybe at the first year.

Characteristic sign: closing one eye in the bright light.

The eye converges when looking at near fixation (looking at near objects). But the squint becomes more prominent with distance fixation (looking at far objects)

Features:

- Acquired.
- Early childhood.
- Intermittently controlled by fusional convergence.
- Close one eye in the bright light parents notice that. This happens due to the outward drifting the eye which created a double vision.

Treatment:

- **Medical**
  - Alternate patching,
  - Over minus lenses (e.g. if the patient has myopia -1, we give him -2 lenses to accommodate more)
- **Surgical**. Indicated when the eyes is deviated 50% of the time. Involves lateral rectus muscle

This deviation may later progress to constant exotropia

Surgery indications:

- poor control.
- The deviation occurs more than 50% of time.
- Lost distance stereopsis.

2- Constant Exotropia

Constant exotropia is less common than intermittent exotropia. It may be present at birth or may occur when intermittent exotropia progresses to constant exotropia.

Treatment:

- Correction of any refractive error
- Correction of amblyopia
- Surgical correction of strabismus

3- Sensory exotropia

Constant exotropia that occur following loss of vision in one eye e.g trauma

Treatment

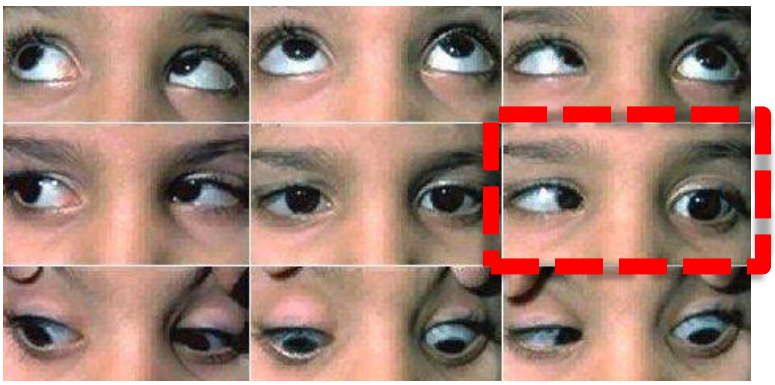
Correction of any organic lesion of the eye
Correction of amblyopia
Surgical correction of strabismus

Basic	Pseudo divergence excess	True divergence excess
• XT D=N • Tx: BLR weakening	• Xt D>N • Patch 30-60min • XT D=N • Tx: BLR weakening	• XT D>N • Patch 30-60min • XT D>N • Tx: R&R

Special Types of Strabismus

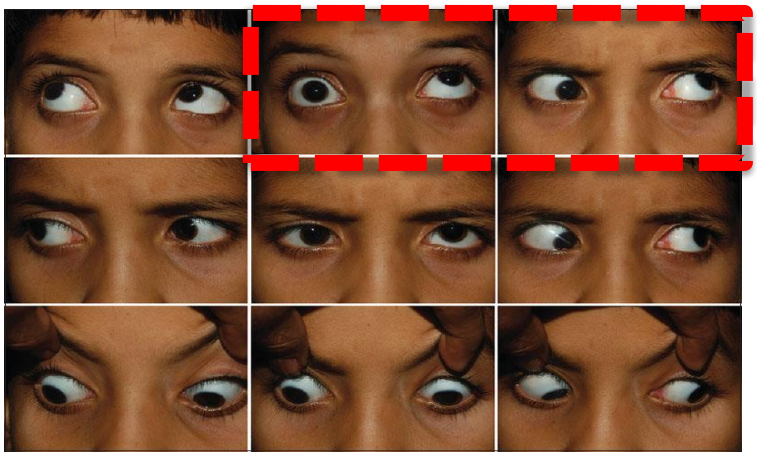
❖ Duane's strabismus

- **Limitation of abduction** (diagnostic test)
- Mild limitation of adduction
- Retraction of the globe and narrowing of the palpebral fissure on adduction
- Upshoot or downshoot on adduction
- Pathology **faulty innervation of the lateral rectus muscle by fibers from medial rectus leading** to co-contraction of the medial rectus and lateral rectus muscles



❖ Brown's syndrome

- Limitation of **elevation on adduction** (diagnostic)
- Restriction of the sheath of the superior oblique tendon
- Treatment needed in abnormal head position or vertical deviation in primary position
- The main differential diagnosis is paresis of the **inferior oblique muscle**.
- The condition is usually unilateral and idiopathic, though rarely it may be due to trauma, inflammation, or tumor.



❖ Thyroid opthalmopathy

- Restrictive myopathy commonly involving inferior rectus, medial rectus and superior rectus
- Patients presents with hypotropia, esotropia or both

Amblyopia (lazy eye)

Decreased visual acuity that is uncorrectable with glasses or contact lenses, as a result of prolonged abnormal visual experience in a child under the age of 7 years and without organic causes.

The three main causes of amblyopia are

1- **Strabismus**,
2- **Unequal refractive error (anisometropia)**; one eye is not equal to the other.
3- **Ametropic amblyopia**; high refractive error in both eye. Prescribe glasses if:
   hyperopia: +5 and more
   myopia: -10 and above
   Antistigamitsm: 1.5
4- **Visual deprivation** (eg, congenital cataract, ptosis or optic nerve hypoplasia).

- Almost during visual immaturity till the 9th BD. The visual pathway is a plastic system that continues to develop during childhood until around 6-9 years of age. During this time, the wiring between the retina and visual cortex is still developing. Any visual problem during this critical period, such as a refractive error or strabismus (misaligned eyes) can mess up this developmental wiring, resulting in permanent visual loss that can't be fixed by any corrective means when they are older.

Major risk factors: prematurity, developmental delay, and family history

Avoidance: screening children during vaccine visits.

Diagnosis criteria: -team 433-

- Visual acuity <20/40 both eyes (oculus uterque) or in one eye.
- Family history
- History of visual deprivation during infancy like cataract

Amblyopia is diagnosed when the following criteria are met and a cause is identified: (433 Team)	
Unilateral	Bilateral
• Asymmetric objection to occlusion of 1 eye • Unequal fixation preference behaviour • Best corrected visual acuity difference between the 2 eyes of 2 more lines	• Best corrected visual acuity less than 20/40 in either eye in a child age 4 or older • Best corrected visual acuity less than 20/50 in either eye in a child age 3 and younger

Classification

● Strabismus amblyopia ● Refractive amblyopia ● Occlusive: organic, cataract ● Idiopathic, 2ry to nystagmus

Treatment


Patching

- Correct refractive error.
- Part-time occlusion / patching, as a treatment option, you patch the healthy eye and let the patient only use the diseased one.
    - It is not used during sleep
    - Patch duration depends on the patient's age
    - do NOT stop putting patch suddenly, it will get worse
    - If the vision became equal in both eyes, we give the patient maintenance patches, then reduce them one hour till he reaches the maturity of his vision at 9 or 10 years old
- Defocusing (penalization). We can use it if the patient refused patching

  Penalization is a therapeutic technique performed by optically defocusing the eye having better vision by using cycloplegia (pharmacological penalization) or by altering the eyeglass lens (optical penalization) to cause decreased vision in the non-amblyopic eye. If pharmacologic penalization is used it should be used for a short period as atropine has side effects.

  Or we use atropine to dilate pupil and cause blurry vision in the good eye that's why the patient will be forced to use the lazy eye.

Leukocoria **(important topic for OSCE)**

Definition: white pupil and media opacity under opthalmascope.
Media opacity: fancy name for absent red reflex.

It's a sign not a diagnosis.

Differential diagnosis:

1- **Retinoblastoma**.
   Common primary malignant ocular tumor in children. Ultrasound is used to rule it out.
   Treatment:
   If small tumor: laser removal
   If big tumor: enucleation (eye removal)

2- **Kocuria**
3- Persistent hyperplastic primary vitreous (**phpv**): persistent hyaloid vessels
4- Coloboma
5- **Congenital cataract**. unilateral, associated with TORCH infections
6- **Inherited cataract**. bilateral
7- **Retinopathy of prematurity**. Common in premature babies whom received extensive oxygen therapy.
8- Others: Astrocytoma Coat's disease, uveitis

Dark is important

Former teams

Tests for deviation (How to detect strabismus?)

Hirschberg test (Corneal light reflex)

A test used to assess alignment of the eyes by shining a light in the person's eyes and observing where the light reflects off the corneas.

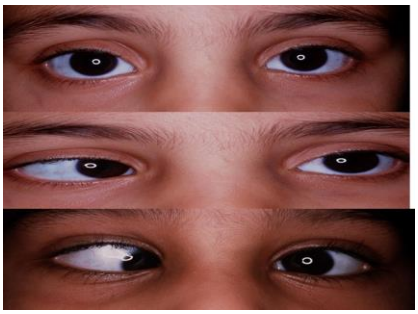
Every millimeter the corneal light reflex is off center, equals approximately 15 diopters of prism or 7 degrees. (1 mm=15PD or 7°)

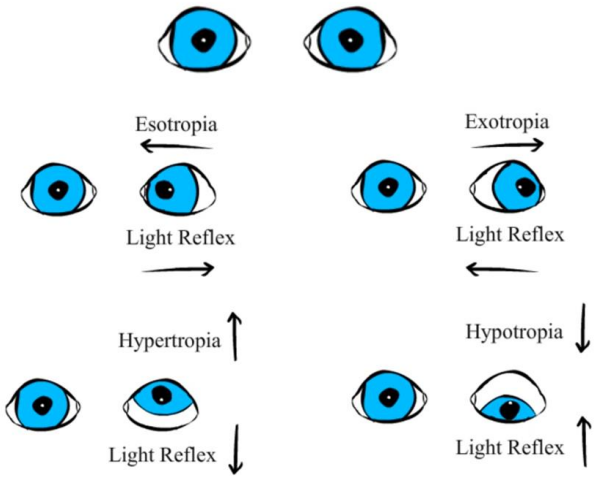
Note: roughly if corneal reflex is:

- **at the pupil edge = 30 PD (15°)**
- **midway between pupil and limbus = 60 PD (30°)**
- **at the limbus = 90 PD (45°)**

When the corneal reflex is:

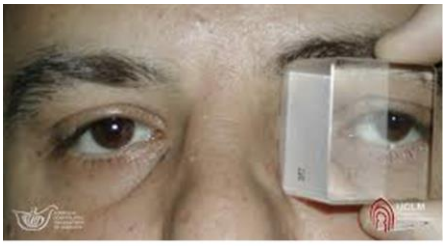
- **in the center we call it orthoptic and that's the normal case.**
- **Light is shifted temporally or laterally → esotropia (inward deviation)**
- **Light shifted nasally → exotropia (outward deviation)**





Krimsky test

The Krimsky test is essentially the same as Hirschberg test, except that we quantitate it better by using a prism. The prism is placed in front of the deviating eye and is used to move the light reflex to the center of the pupil. The apex of the prism is directed towards the direction of deviation i.e. laterally if exotropic and medially if esotropic.

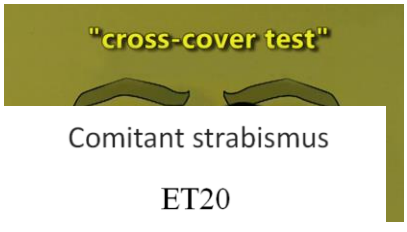


Krimsky test

Cover test **Watch this video and you'll understand everything!**

There are 2 types of cover tests:

- **Unilateral cover test (cover-uncover test): performed by having the patient focus on an object then covering the fixating eye and observing the movement of the other eye. If the eye was exotropic, covering the fixating eye will cause an inwards movement; and esotropic if covering the fixating eye will cause an outwards movement. It is used to detect tropias.**
- **Alternating cover test (cross-cover test): performed by moving the occluder from one eye to the other eye. Normally, the covered eye shouldn't move when the occluder is removed. It is the most accurate way to pick up subtle phorias and tropias since it breaks binocular vision.**



Comitant strabismus

ET20

ET 25 ET30 ET25

ET30

Prism and cover test

Types of strabismus (Two classifications)

First:

- **Comitant: can be XT or ET**
  Almost same angle in any direction of gaze (within 10 PD range).
- **Non-comitant: can be XT or ET**
  angle change with direction of gaze (Paretic as in 6th and 3rd nerve palsy, restrictive as in thyroid disease).

Second:

- **Esotropia (ET): The eyes are turned inwards (crossed-eyed) all the time**
- **Exotropia (XT): The eyes are turned outwards (wall-eyed) all the time**
- **Hypertropia (HT): The eyes are turned upwards**
- **Hypotropia (HPO): The eyes are turned downwards**

Non-comitant strabismus

ET 20

ET 10 ET 12 ET40

ET18

Summary

Esotropia				
	Age	**Hypermetropia**	**Treatment**	**Special characters**
Infantile	**<6 months**	**Small**	**weakening the medial rectus muscles at age of 10-11 months**	**- Large angle - Cross fixation**
Hypermetropic	**>6 months - 7 years (average 4 years)**	**>+2.00**	**Glasses**	**- Risk of amblyopia - Intermittent at onset then constant - <10 PD residual with glasses**
High AC/A ratio	**_____**	**_____**	**Bifocal glasses**	**crossing of eyes with focusing efforts on near objects**
Partially acc	**_____**	**full hypermetropic correction**	**Surgery for residual deviation**	**>10 PD residual**
Divergence paralysis	**Adult**	**_____**	**Treat underlying cause then surgery**	**- Emergency - D>N**
Sensory	**_____**	**_____**	**Surgery**	**- blindness - can cause ET or XT**
Cyclic	**2-6 years**	**might be present or absent**	**Hypermetropia → glasses NO hypermetropia → surgery**	**- acquired - cycle between straight and ET**