Strabismus ,Amblyopia & Leukocoria

2017-2018

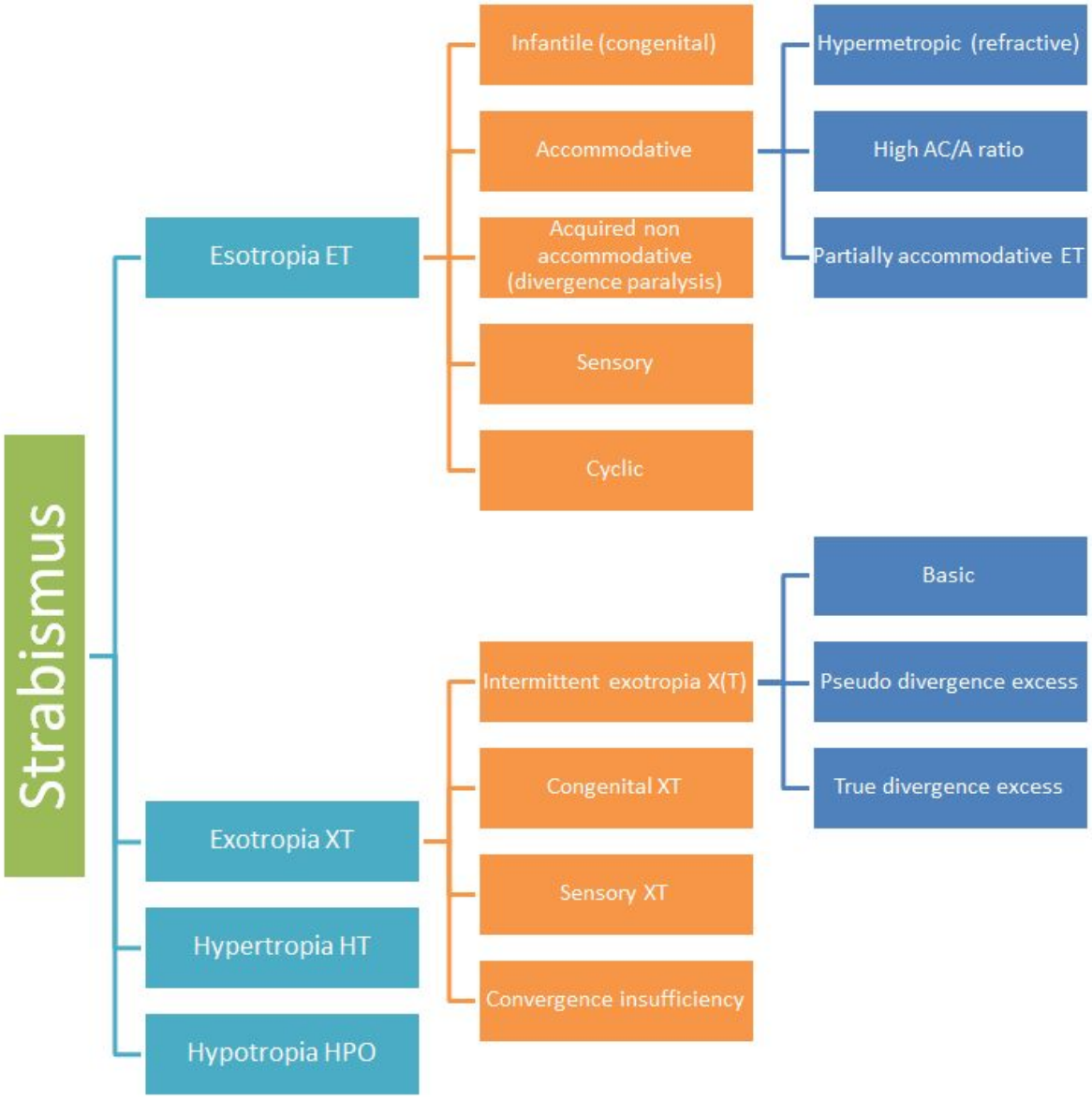
Done By: Lina Aljurf

Resources: Team 433, Doctors Notes, Ophthobook

slides
Important
book

Editing File

Objectives: Not provided



Abbreviations you need to know:

A or **Acc**: accommodation
AC/A: Accommodative convergence/accommodation ratio
N or **D**: Near and distant vision
ET: Esotropia
XT: Exotropia
X(T): Intermittent exotropia
HT: Hypertropia
HPO: Hypotropia
X: Exophoria
BLP: Bare light perception
Va: Visual acuity
PTO: Part-time occlusion
RD: Retinal detachment
EOM: Extraocular movements
PD: Prism diopter
BLR: Bilateral rectus muscle
R&R: Recess and resect

Strabismus

- Strabismus is ocular misalignment (Abnormal alignment of the eyes, the condition of having a squint)
- A strabismus can be tropia or phoria
- 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.
- 2%-3% of children and young adults.
- Incidence is equal in males and females.

Causes

1. Inherited pattern. Most patients fall under this category, so it is important to ask about family history.
2. Idiopathic.
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.

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.

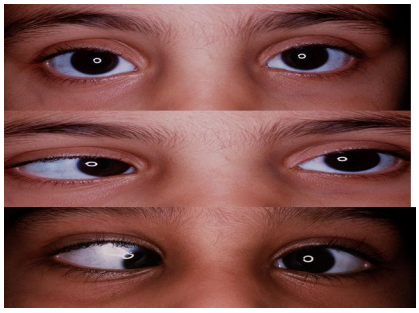
Every millimeter the corneal light reflex is off center, equals approximately 15 diopters of prism or 7 degrees. (1 mm=15PD or 7°)

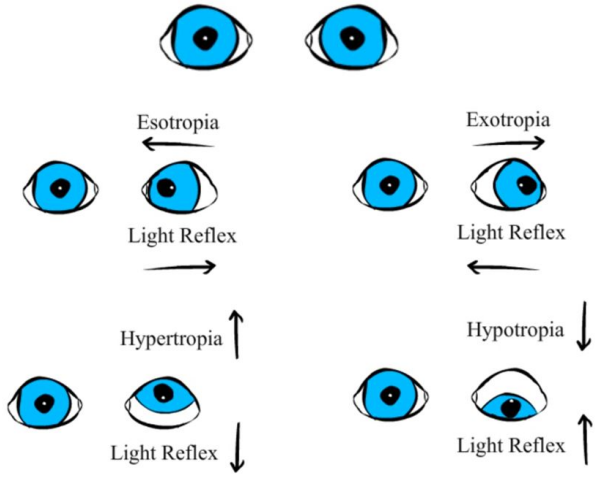
Note: roughly if corneal reflex is:

- **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)**





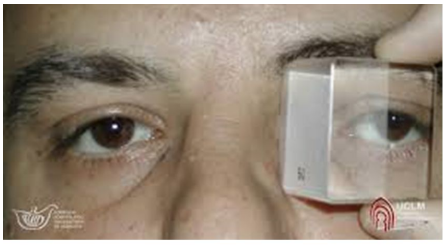
Krimsky test

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.

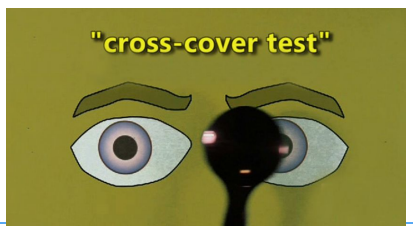
Cover test Watch this video and you'll understand everything!

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.**



Krimsky test



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).

Comitant strabismus

ET20

ET 25 ET30 ET25

ET30

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

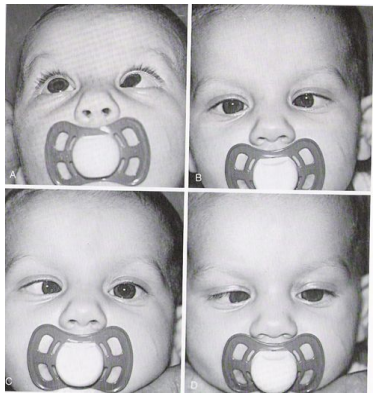
ET 20

ET 10 ET 12 ET40

ET18

Esotropia (ET) types:

1. Infantile (congenital) ET.
2. Accommodative ET: (most common)
   a. hypermetropic(refractive)
   b. high AC/A ratio.
   c. partially accommodative ET.
3. Acquired non accommodative ET (DIVERGENCE paralysis).
4. Sensory ET.
5. Cyclic ET.



1-Infantile (congenital) ET

- Large Angle.
- Small hypermetropia.
- Before age of 6months.
- Cross fixation (turning the face to fixate the eye contralateral to the target).

Ocular association of infantile ET:

- 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 of infantile ET:

Surgically by weakening the medial rectus muscles at age of 10-11months to achieve monofixation syndrome.
(by disinserting the muscle from the original position and move it backward according to the degree of deviation i.e. if it was 30 you see in the table how many mm you have to move it back and place the insertion in the sclera)
Monofixation syndrome is a form of subnormal binocular vision without bifixation. It is often desired result of strabismus surgery
We have to make sure that there is no refractive error before doing the surgery.

Prognosis: gross stereopsis. (Stereopsis: the visual perception of depth, or the ability to see three dimensionally)

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: (The pics are not for this 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.**
- **Cyclоplegic retinoscopy is +1.25 D**
- **Fundus: normal**

Before surgery After surgery

2-Accommodative ET (most common)

A-Hypermetropic (refractive)

- **>+2.00 hypermetropia.**
- **age>6mo-7 years (4yrs).**
- **High risk of amblyopia.**
- **Intermittent at onset then constant.**
- **Corrected totally (<10PD residual N+D) with glasses. (N+D=Near + distant vision)**

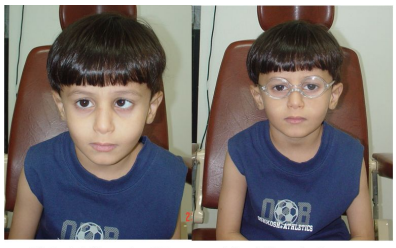
Acc ET due to hypermetropia corrected with glasses

Hypermetropic ET corrected with glasses

B-High AC/A ratio (Non-refractive)

The accommodative convergence/accommodation (AC/A) ratio gives the relationship between the amount of convergence (in-turning of the eyes) that is generated by a given amount of accommodation (focusing effort).
Esotropia with high AC/A ratio (also termed non-refractive accommodative esotropia).

Figure 7: Bifocals bisecting the pupil



High AC/A ratio ET treated by bifocal glasses

C- Partially accommodative ET

- >10 PD residual for D+N with full hypermetropic correction.
- Treatment: Surgery Sx for the residual deviation.

Scenario: a patient was given glasses. Two months later he comes with residual 10 PD or more for near+distant with full hypermetropic correction. You need to refract him again to see if there is any hidden hypermetropia. If esotropia >10 PD persists after full hypermetropic correction you go for surgery. Unlike fully accommodative type mentioned earlier which requires only glasses.

3- Divergence paralysis (Acquired non-accommodative ET) emergency

- ET at D>N (Distance > Near).
- MRI: arnold chiari, pontine tumor
- Adult
- First, treat the underlying cause then treat the esotropia with surgery

4- Sensory ET

- ET due to unilateral blindness. A patient with such a problem could have both ways: inward or outward deviation
- Treatment: Surgical usually for cosmetic purposes, does not treat the underlying problem

5- Cyclic ET

- Very rare.
- Acquired(2-6yrs).
- Cycle between straight and ET. (يجي ويروح)
- Treatment:
  - if hypermetropia treat with glasses
  - if NOT hypermetropia treat by surgery

Pseudostrabismus:

Pseudoesotropia is a condition in which alignment of the eyes is straight (also known as orthotropic); 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.

Exodeviation (Exotropia)

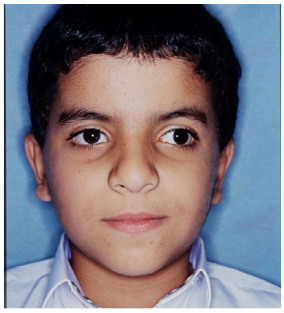
a horizontal form of strabismus characterized by visual axes that form a divergent angle (or in a simpler way: eyes turned outwards)

Exotropia types:

1. Intermittent exotropia X(T) .
2. Congenital XT.
3. Sensory XT.
4. Convergence insufficiency.

1-Intermittent exotropia X(T)

- 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.



This deviation may later progress to constant exotropia

Treatment:

- surgical involves lateral rectus muscle
- Non-surgical: alternate patching, over minus lenses (e.g. if the patient has myopia -1, we give him -2 lenses to accommodate more)

Surgery indications:

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

Types of X(T):

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

Type	Definition
Basic	Distance and near measurements are equal.
Pseudodivergence Excess	Distance measurement initially exceeds near, but the near measurement approaches distance after 30–60 minutes of monocular occlusion.
True Divergence Excess	Distance measurement exceeds near measurement by ≥ 10 prism diopters even after monocular occlusion for 30–60 minutes.

2- Congenital XT

Very rare.
Constant large angle between the two eyes which is assessed by the prism
Poor fusion prognosis
High risk of amblyopia
Associated with craniofacial abnormalities,albinism,CP.
Treatment: Bilateral rectus muscle (BLR) Weakening.

3- Sensory XT

Blind eye drifts outward
Treatment: Surgery (cosmetic purpose)

4- Convergence insufficiency

- Inability to maintain the convergence on objects approaching from distance to near (moving pencil from distance to near your eyes will cross normally. In such patient their eyes will drift out)
- Complain of C/O: (symptoms): asthenopia (frontal headache), diplopia
- Exophoria X or exotropia XT at near N ,Straight at distance D.
- Remote near point of convergence (normal 5-6cm).
- Tx: orthoptic exercise.

Amblyopia (lazy eye)

Definition: Amblyopia refers to reduced vision, uncorrectable with glasses or contact lenses, due to failure or incomplete development of the visual cortex of the brain.

- VA is <20/40 or 2 lines below the good eye.
- 2 ~4%.
- 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.
- Unilateral or bilateral.

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

Diagnosis criteria:

- VA <20/40 OU or in one eye.
- Family history (FHx)
- Hx 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

- Optical correction.
- PTO (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.

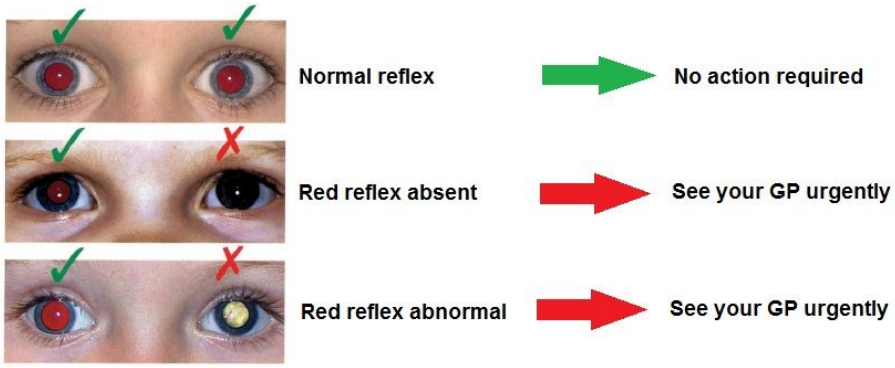

Patching

Leukocoria (important topic for OSCE)

Definition: white pupil and no red reflex

Potential causes:

- Cataract (most common) A cataract in a newborn can occur by several mechanisms. They can be idiopathic, genetic, from metabolic disorders, child abuse trauma, or caused by one of the maternal TORCH infections during fetal development. Whatever the cause, it is important to remove these cataracts as soon as possible as they are amblyogenic and will lead to permanent vision loss from visual neglect.
- Retinoblastoma
  - Eye tumor, highly aggressive and needs immediate intervention
  - Children less than 4 years, average 18 months
  - It's the most common primary malignant ocular tumor in children.
  - Due to random somatic mutation or several germline inheritance pattern
  - spreads by extension to the optic nerve toward the brain
  - Ultrasound is used to rule it out
  - Treatment: often involves enucleation (removal) of the entire eye to avoid seeding tumor cells into the orbit
- Persistent hyperplastic primary vitreous (PHPV)
- COLOBOMA a developmental defect of the eye occurring at embryonic stage
- Retinal detachment (RD)
- Astrocytoma
- Coat's disease, uveitis
- Retinopathy of prematurity (ROP



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