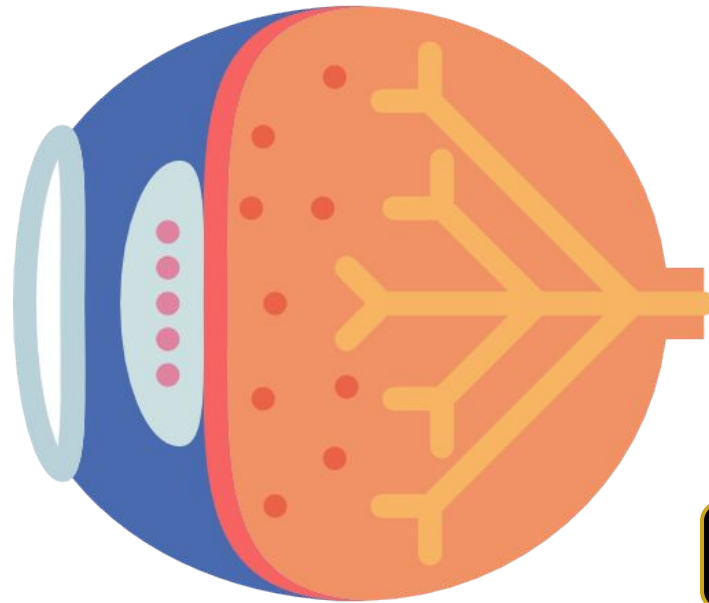


# Lecture: 3



[Editing file](#)

## Strabismus, Amblyopia & Leukocoria

- **Presented by Dr. Saeed Al Wadani**
- To understand the definition and types of strabismus and management options.
- Types of ocular deviation (concomitant and non-concomitant).
- Physiologic response to ocular misalignment:
  - Adaption: abnormal head position, suppression, amblyopia
  - Association of strabismus.
  - Therapy.
- To know extraocular muscles, innervations and function.
- To know the definition of amblyopia, different causes and management.
- The definition of leukocoria, causes and management.



Important



Doctor's notes



Golden notes



Extra



Book



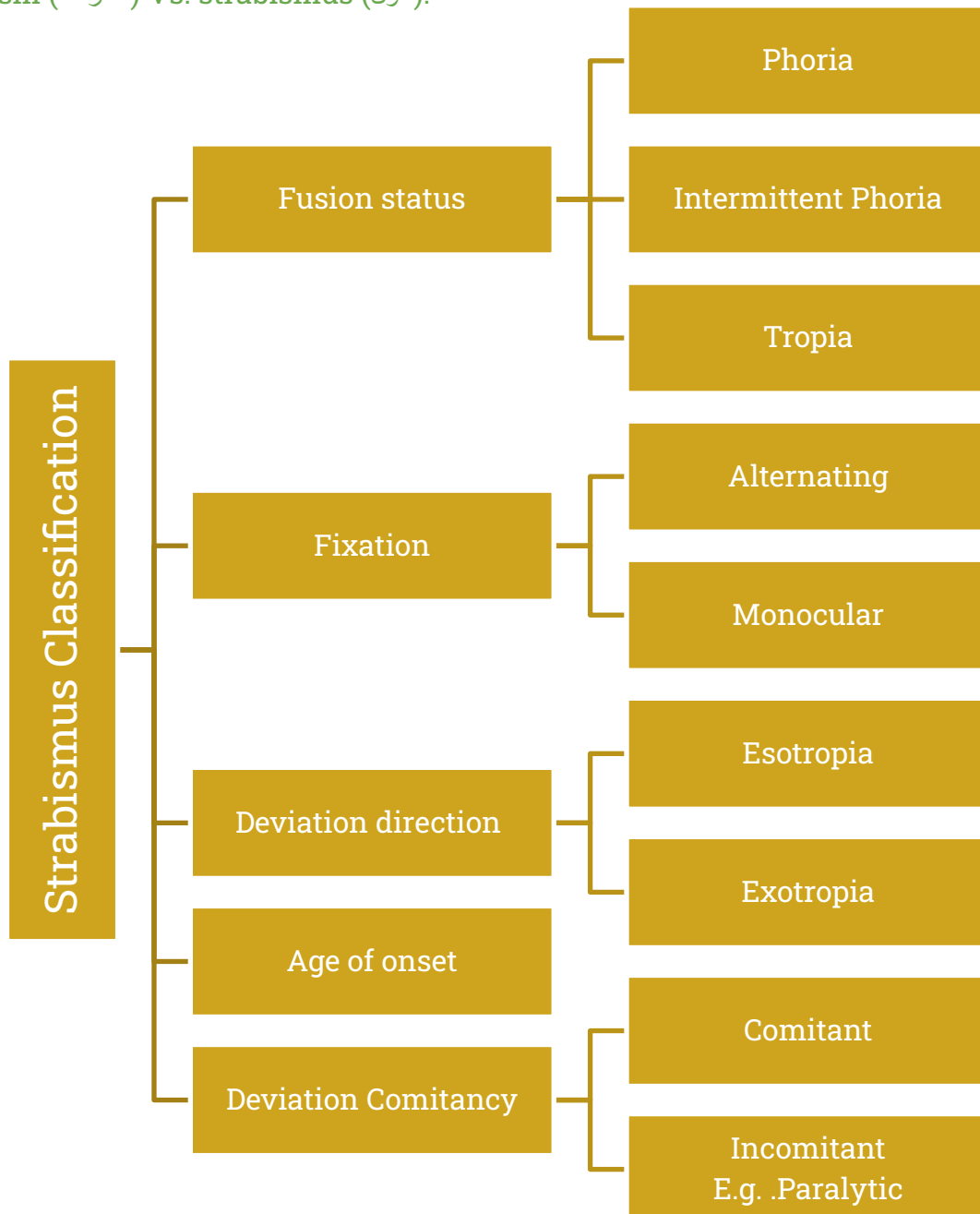
OPHTHALMOLOGY TEAM

# Strabismus (حول)

## Definition

Ocular misalignment due to abnormality in binocular vision or anomalies in neuromuscular control of ocular motility

- Strabismus has an inherited pattern, i.e., it is much more likely to occur if one or both parents are affected. However, many cases occur without any family history of the disorder.
- Strabismus is associated with reduction of depth perception (binocular vision) and, if onset is in adulthood, double vision.
- If the eyes are misaligned, depth perception is substantially reduced. When one eye is deviated in early childhood, the brain may learn to ignore the image from that eye, and amblyopia (lazy eye).
- In adulthood, the affected individual usually experiences double vision. This occurs because the brain, which no longer has the "plasticity".
- Several neurological conditions are more commonly associated with strabismus, including Down's syndrome, cerebral palsy, hydrocephalus, and brain tumors.
- A cataract, eye tumor, or other eye disorder associated with reduced vision may also present with strabismus.
- **Astigmatism (انحراف) Vs. strabismus (حول).**



# Classification of Strabismus

**Fusion status** It describe how the strabismus appear or manifest to you.

## 1. Phoria

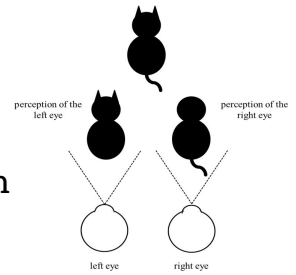
Latent tendency of the eye to deviate and controlled by fusional mechanism misalignment of the eyes that only appears when binocular viewing is broken by **Alternate cover test** which the examiner occludes one eye and then the other, switching the occluder back and forth to occlude the eyes without allowing the patient to fuse in between occlusion; to break fusion control, **the squint will appear**.

## 2. Intermittent Phoria

Fusion control is present part of the time.

## 3. Tropia

Manifest misalignment of the eye all the time. These patients show visible strabismus, no tests needed.



(Both eyes control together that called binocular fusion, or binocular single vision where the brain tries to produce one single image by fusing and controlling both eyes)

**Fixation** One eye is fixed

- Alternating:** Spontaneous alternation of fixation from one eye to the other, e.g pt has esotropia, sometimes left eye is straight and right is deviated inside and visa versa.
- Monocular:** Preference of fixation with one eye, e.g left eye is fixated and right eye is deviated all the time. **Bad prognosis.** (fixed with patching the good eye, to stimulate the deviated eye)  
Why is it important to differentiate between alternating and monocular?  
Child with Monocular may has amblyopia bc he just use fixed eye and other eye will be neglected (not functioning); while Alternating switch between them so no amblyopia.

## Type of deviation

### 1. Horizontal

- Esodeviation = eye is deviated inside
- Exodeviation = eye is deviated outside If eye deviates inside (to nose) we called it esophoria/esotropia, if it deviates outside we called it exophoria/exotropia.

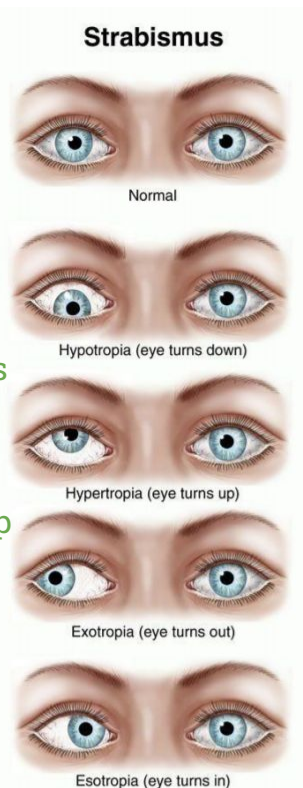
### 2. Vertical

- Hyperdeviation = eye deviated up
- Hypodeviation = eye deviated down

### 3. Torsional

- Incyclodeviation
- Excyclodeviation

### 4. Combined



Vision at childhood differ from vision at adult; anatomy of eyes at birth is mature but physiological is not matured (visual acuity still grows (improves) until reaches 6/6 from birth to 6 or 7 age). **Any abnormalities of eye in childhood period (like squint, cataract, ptosis and RE..) must treat them or will lead to amblyopia "lazy eye"; will never improve.** So decreasing vision if not treat will lead to amblyopia, thus, wearing eyeglasses is necessary and very important.

Usually non-acquired associates with hyperopia. In ophthalmology pediatrics clinic: 70 to 80% have esotropia squint and 90% of this case has hyperopia; that means the cause of squint is hyperopia, and 70% of this case only treat by eyeglasses; hyperopia they are accommodation so they suppose to be plus 1 at least plus 3 .in children are plus 6 or plus 7 (esotropia)

## Age of onset

- Congenital at birth
- Acquired (usually at 2 years)

## Classification of Strabismus:

### Variation of the deviation with gaze position or fixing eye

Angle of deviation of eye, how we measure it? By prism cover test.

- Comitant:**  
Same angle of deviation in different direction of gaze, **most common in pediatrics**, and deviation **secondary to refractive error (most common)**, congenital muscle disease.
- Incomitant:**  
Variable deviation in different direction of gaze usually in paralytic or restrictive type of strabismus. **e.g nerve palsy, thyroid eye disease causing restrictive myopathy and usually in adults.**

### Examination:

- History** ask about age of onset, family history, history of trauma...etc
- Inspection**
- Assessment of monocular eye function** check vision in every eye:
  - Visual acuity:
    - Preverbal children **within 1 -2 years can't take vision:**
      - CSM (**Central: no deviated, Steady: no nystagmus, Maintained: fixated**).
      - OKN.
      - Preferential looking.
      - Visual evoked potential
    - Verbal children:
      - Symbol tests: single illiterate E.
      - Allen pictures.
      - HOTV letters.
- Assessment of binocular eye function (very imp)**

#### Hirschberg test:

Simple test using light(torch) to see **corneal light reflex** in center (normal), if eye deviates inside the corneal reflex will be in temporal -opposite side-(abnormal), and it has angle.

#### Krimski's test (measuring):

It is Hirschberg test with Prism cover test, using for **measuring angle of deviation** and test fixated eye-bc deviated eye is amblyopia.

#### Cover test

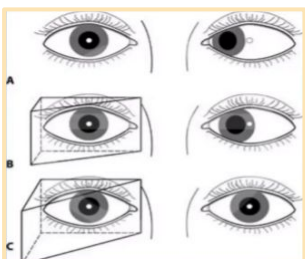
Cover- uncover test to **diagnose tropia**.  
(move from outside to center = exo, move from inside to center = eso)

#### Alternate cover test Prism cover test (measuring)

Breaking fusion control, for phoria.

#### Brückner Reflex

Trans illumination' test to see **red eye** reflex.



By Krimski's test: light reflex + prism, we put prism in front of seeing eye for example at 10 if modify then increase 20, 30... (A, B) to correct non-seeing eye (C) and see the angle.



Hirschberg test

# Examination

**Hirschberg test** please know how to do it very well because you will have it in the OSCE.

- A test used to assess alignment of the eyes by shining a light in the person's eyes "1 meter away" 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 مهمه هالنقطه احفظوها
- Note: roughly if corneal reflex is:
  - At the pupil edge = 30 PD (15o)
  - Midway between pupil and limbus = 60 PD (30o)
  - At the limbus = 90 PD (45o)
- 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)
- **IMPORTANT (Always comes in exams)**
  - Cover-uncover test → detect tropias.
  - Cross-cover test → detect phorias and tropias.
- Ocular movement (motility) is different from eye alignment:
  - **Eye movement ( 9 gaze):** ask the patient to look in all directions (up, down, left & right, oblique) without moving their head and ask them if they experience any double vision.
  - **Ocular alignment:** both eyes are straight, test by 2 tests: Corneal light reflex & cover-uncover test.



Name of the instrument?  
Prism.  
What is it used for?  
Testing the deviation (strabismus)

## Fundoscopy:

- It is important to examine the funds -by indirect and direct ophthalmoscopy- to rule out pathological causes like cataract, tumor, hemorrhage and **retinoblastoma**.
- Retinoscope or automated refractometry for measurement refractive error NOT for fundus.

**Cycloplegic refraction** To relax eyes and see **real refractive error (release accommodation)**

- Tropicamide: Short half life (20min)
- Cyclopentolate: Intermediate two days to three
- Atropin: Two weeks
- We put cycloplegic refraction in:
  - Pediatrics (infants come with small globe hyperopia (less axial length), then becomes myopia large globe)
  - Who has hyperopia because if you give them eyeglass with hyperopia will happen residual refractive error, what does this mean? lens by accommodation change the power of eye.
- More than 45 age they have presbyopia bc accommodation loss (stiffness) with aging, so need reading glasses.
- If a child comes to clinic with +3 and we put cycloplegic will change (increase) to +6, why? bc lens attached to ciliary muscle, and ciliary muscle attached to zonule, and contraction of ciliary muscles will increase the power, in another words, real refractive is +6 but bc lens by accommodation compensate but when kid feels fatigue (increase temperature, head trauma = feel weakness) so muscles are decompensated and eye will deviate inside (this is history)
- Near reflex is the action of the eye for adjustment to fixate on a near object, which consists of accommodation, convergence and miosis.

## Management of squint:

- Rule out pathological causes like cataract by examining funds by fundoscope.
- Correct refractive error, (cycloplegic refraction), (may hyperopia cause esotropia)
- Treat amblyopia: if present, cover the good eye to stimulate the poor eye (according to the age: 1 year old => cover it 1 hr\day, 2 years old => 2 hr\day ....etc) till 6-7 years old
- If not improved go to surgery



# Types of Strabismus

1 **Esotropia** Most cases (90%) in pediatrics are esotropia bc it is associated with Hyperopia.

## Pseudoesotropia حول كاذب

- Occur in patients with flat broad nasal bridge and prominent epicanthal fold like in children and Asian. corneal light reflex in center
- Gradually disappear with age. appears at birth
- **Hirschberg test** differentiate it from true esotropia.
- Using the cover-uncover test (only definitive test) the examiner finds that the patient manifests no deviation (normal exam; because they are central).



## Infantile Esotropia

- Common comitant congenital esotropia occur before 6 month of age. There is no accommodation.
- Deviation is often large more than 40 prism diopter.
- **Frequently associated with nystagmus (bad prognosis) and inferior oblique overaction.**
- Mostly these kids have low refractive error (not significant hyperopia +1,+2) bc have problem in muscles and eyeglasses is not useful and also within first two years of age not use accommodation.
- Classic infantile esotropia is constant and involves a large angle of deviation exceeding 20 prism diopters (PD) on corneal light reflex measurement.
- Treatment is usually surgical.
- **Treatment:**
  1. Correction of refractive error.
  2. Treat amblyopia.
  3. Surgical correction of strabismus.
  - **Infantile esotropia + large a angle of deviation + low refractive error (not significant)= surgery**
  - So, we do strabismus surgery (medial rectus recession) to stretch the muscle (medial rectus is pulling the eye toward the nose) > so we detach the medial rectus muscle from the anatomical insertion and we suture it back more posteriorly which will make it weaker and looser & patch the normal eye to treat amblyopia + Follow up the child until 8-9 years for the amblyopia not the squint.
  - ALWAYS Surgery for the extra-ocular muscles.
  - Treat the amblyopia by occluding the good eye.
  - Factors contributing to poor ocular alignment and visual prognosis include persistent preoperative amblyopia, latent manifest nystagmus, and myopia from -2.5 to 5.0 D.



The child looks like he has a squint, but he doesn't because he has flat nasal bridge and the epicanthus fold covers the sclera creating the illusion of eyes going in. we test it by cover-uncover test to see if the eye moves or not> in strabismus when you cover the good eye the deviated eye will look straight, but here it won't because it is pseudo.

# Types of Strabismus

## Accommodative Esotropia

- More common
  - Occur around 2 ½ years of age **when using accommodation.**
  - Start as intermittent then become constant.
  - High hypermetropia (+6, +7).
- 
- Type of squint: accommodative esotropia  
Type of refractive error: Hyperopia
- **Treatment:**
    1. Full cycloplegic correction.
    2. Treat amblyopia.
  - With patients aged 4-5 years, one can attempt to reduce the strength of the hyperopic correction to enhance fusional divergence and to maximize visual acuity (convex lens to relax the accommodation > back to normal).
  - Wearing glasses for pediatric is MANDATORY, why? To prevent amblyopia \* Adult = optional
  - **How to know if the person is hyperop or myop from their glasses? myopia glasses makes eyes seem smaller (-), while hyperopia makes eyes seem bigger (+)**



- Treatment = convex lenses
- Why? More power-> to reduce accommodation = treat esotropia
- Once treated eye > becomes straight

## Partially Accommodative Esotropia

- Improve partially with glasses.
- **Treatment: needs surgery**
  1. Full cycloplegic correction
  2. Treat amblyopia
  3. Surgical correction of strabismus



- If a child has squint gives eyeglasses with full correction and cycloplegic. If a child comes with +3 and after giving him cycloplegic refraction becomes +6, he is treated by giving +6. If it has monocular fixation, cover fixated eye to stimulate weak eye. If he returns after 3-6 months and the eye becomes straight this is Accommodative Esotropia bc squint modify by eyeglasses but if he returns after 3-6 months and eye is not straight but modify by 80% . This called Partially Accommodative Esotropia, treat of partial (20%) by surgery.

# Types of Strabismus

## 2 Exotropia

- Represent 5% to 10% cases of squint
- Intermittent exotropia the commonest type
- Constant exotropia
- Sensory exotropia
- Congenital
- Not related to hyperopia



Diagnosis: strabismus (exotropia).  
Examination: corneal light reflex shows outward deviation. Treatment: if indicated lateral rectus muscle recession.

### Intermittent exotropia

- Onset of deviation within the first year of age .
- Closing one eye in bright light.
- Usually not associated with any refractive error.
- Usually not associated with amblyopia.
- **Treatment:**
  1. Correction of any refractive error.
  2. Surgical correction of strabismus.

### Constant exotropia

- Maybe present at birth or maybe progress from intermittent exotropia.
- Treatment
  1. Correction of any refractive error.
  2. Correction of amblyopia.
  3. Surgical correction of strabismus.



### Sensory exotropia

- Not functioning eye for ex amblyopia (poor vision could be due to cataract, retinal detachment, optic atrophy...) => cause eye to deviate.
- Constant exotropia that occur following loss of vision in one eye e.g trauma, cataract.
- **Treatment:**
  1. Correction of any organic lesion of the eye.
  2. Correction of amblyopia.
  3. Surgical correction of strabismus.

## 3 Paralytic strabismus Incomitant (all above is comitant)/ usually in adults

### 6th Nerve Palsy in exam

- Incomitant esotropia
- Limitation of abduction
- Abnormal head position
- With ischemic or multiple sclerosis

Neuro ophthalmology lecture is very imp. esp 6th  
C- Paralytic strabismus nerve palsy "exam"





# Types of Strabismus

## 3rd Nerve Palsy

- Congenital or acquired.
  - Exotropia with Hypotropia of the affected eye.
  - In children caused by: trauma, inflammation, post viral and tumor.
  - In adult caused by: aneurysm (posterior communicating artery aneurysm), diabetes, neuritis, trauma, infection and tumor.
  - Pupil involved or intact.
- \*How can you differentiate between the cause of 3rd cranial nerve palsy is it surgical (related to nerve compression) or medical (related to blood supply)? by pupil examination\*

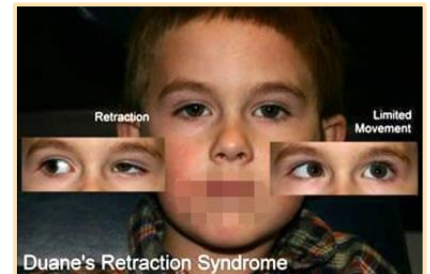
## 4th Nerve Palsy

- Congenital or acquired
- Hypertropia of the affected eye with excyclotropia
- Abnormal head position

## 4 Special Types of Strabismus

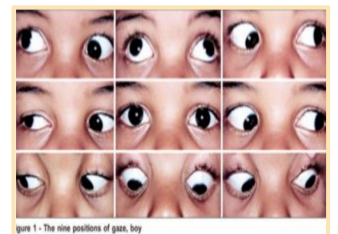
### Duane Syndrome (like 6th CN palsy)

- Limitation of abduction مايقدر يشوف براء، ماتطلع العين براء
- Mild limitation of adduction
- **Diagnostic test: when adduct the retraction of the globe** تخيل في لميتيشن في الابدكشن والادكشن فالعضلتين يشدون فتدخل العين الجوا **And narrowing of the palpebral fissure on adduction** العينة فتدخل الجوا
- this how to differentiate between 6th CN palsy and Duane (diagnostic test) as in the pic duane syndrome in the left eye.
- Upshoot or downshoot on adduction العضلة مشدودة وعند الادكشن مشدودة فالعين تروح فوق أو تحت
- **Type1: one eye, type 2: both eyes.**
- Pathology faulty innervation of the lateral rectus muscle by fibers from medial rectus leading to co-contraction (miscommunication/miscontraction) of the medial rectus and lateral rectus muscles
- Vision is 20/20, Duane Syndrome don't have amblyopia. Problem when eye move horizontally.
- Treatment: no need bc does not cause amblyopia and cosmetic in childhood is not important the important is visual acuity to avoid amblyopia.



### Brown Syndrome Common to be asked in the exam

- Problem when eye move vertically.
- **Limitation of elevation on adduction** ماتطلع العين
- Restriction of the sheath of the superior oblique tendon.
- Could be unilateral or bilateral but usually unilateral.
- Treatment needed in abnormal head position or vertical deviation in primary position or has amblyopia.



### Thyroid Ophthalmopathy

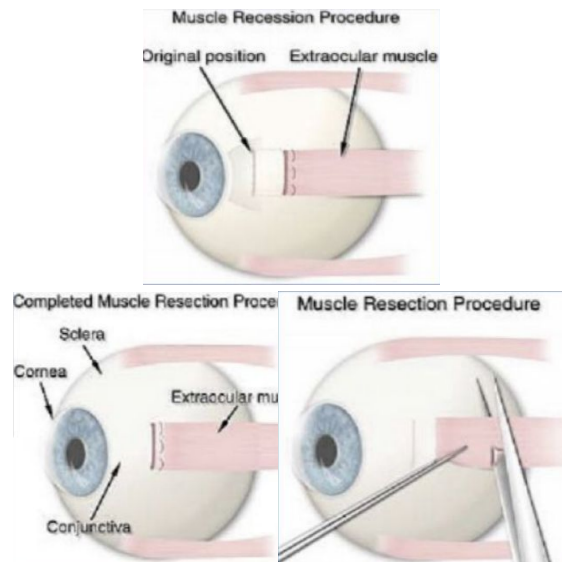
- **Restrictive myopathy commonly involving inferior rectus** commonest to be affected, then **medial rectus** 2nd most common and superior rectus.
- Patients presents with hypotropia, esotropia or both. **Orbital diseases are very important**



# Types of Strabismus

## Surgery of Extraocular Muscle

1. Recession ارخي عضلة : weakening procedure where the muscle disinserted and Sutured posterior to its normal insertion.
  2. Resection اشد عضلة : strengthening procedure where part of the muscle resected And sutured to its normal insertion
- In General, if we have deviated eyes inside, tightened lateral rectus muscles and relaxed medial rectus.
- **Complication of Extraocular Muscle Surgery**
    - Perforation of sclera
    - Lost or slipped muscle insertion of muscle comes from optic nerves so dis-insertion will happen
    - Infection
    - Anterior segment anesthesia
    - Post operative diplopia common
    - Conjunctival granuloma and cyst



Normal vision at birth (1-2 hrs old)

- Blurry vision (barely counting fingers).
- Anatomically (build is complete).
- Physiologically (incomplete) upto to 3-5 years.

# Amblyopia

## Definition

Amblyopia or lazy eye refers to decrease of vision, either unilaterally or bilaterally, for which no cause can be found by physical examination of the eye.

- 2%–4% of U.S. population affected
- **Three critical periods** of human visual acuity development have been determined, during these time periods, vision can be affected by the various mechanisms to cause or reverse amblyopia.
  - These periods are as follows:
    - Development of visual acuity from the 20/200 range to 20/20, which occurs from birth to age 3-5 years.
    - The period of the highest risk of deprivation amblyopia, from a few months to 7 or 8 years.
    - The period during which recovery from amblyopia can be obtained, from the time of deprivation up to the teenage years or even sometimes the adult years.
- **Any abnormalities ( cause abnormal development of vision) during this period will lead to amblyopia if not treat after age 8.**
- **Vision develop normally: 1- both eyes are straight 2- No refractive error 3- Clear media means light inside clear cornea, clear anterior chamber, clear lens, clear vitreous. So any problem in these 3 and not detected during this period will lead to amblyopia.**
- **Diagnosis of amblyopia:** usually requires a 2-line difference of visual acuity between the eyes, for example eye is 6/6 the other eye is 6/8

## Causes of Amblyopia

### 1. Anisometropia

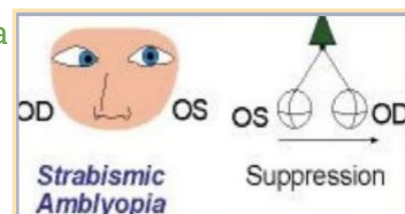
- Is varying of refractive power in both eyes, plain eye has focused image and ametropia has anti focused, and amblyopia happens in defocused images.
- Inhibition of the fovea occurs to eliminate the abnormal binocular interaction caused by one defocused image and one focused image.
- This type of amblyopia **is more common** in patients with anisohypermetropia than anisomyopia. Small amounts of hyperopic anisometropia, such as 1-2 diopters, can induce amblyopia. In myopia, mild myopic anisometropia up to - 3.00 diopters usually does not cause amblyopia.
- **Anisohypermetropia:** الطفل مايشوف القريب وهذا يسبب له امبلوبيا
- **Anisomyopia:** الطفل إللي عنده مشكلة فقط بالبعيد فما يسبب امبلوبيا  
 → The only way to find out anisometropia is to screen the child, at 6m, 2 y, 4 y, 5 years (4 times).  
 → child will never complain of anything , the family would not notice anything because both eyes look normal.

### 2. Strabismus

- The patient favors fixation strongly with one eye and does not alternate fixation (monocular fixation). This leads to inhibition of visual input to the retinocortical pathways.
- Incidence of amblyopia is greater in esotropic patients than in exotropic patient, because esotropia more common to happen than exotropia.

### 3. Visual deprivation

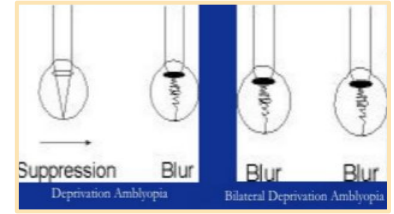
- Amblyopia results from disuse or under-stimulation of the retina.
- This condition may be unilateral or bilateral. Examples include cataract, corneal opacities, ptosis, and surgical lid closure.
- **Must be corrected before 5 years of age or will develop amblyopia**



# Amblyopia

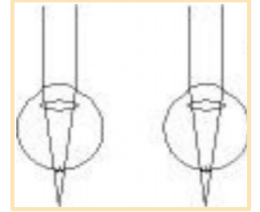
## 4. Organic

- Structural abnormalities of the retina or the optic nerve may be present.
- Functional amblyopia may be superimposed on the organic visual loss.



## 5. Ametropic Amblyopia

- If both eyes were +7 we call it Ametropic Amblyopia.
- Uncorrected high hyperopia is an example of this bilateral amblyopia
- Every refractive error can cause different Amblyopia; for example:
  - Myopic -10 or more
  - Hyperopic was +5 or more
  - Astigmatism 1.5 or more
  - Or if it's less than these numbers but the child has squint
- All will lead to Amblyopia, so the child MUST wear eyeglasses to prevent it.



## SCREENING: IMPORTANCE

- How to treat them? How to know if they even have it or not? By Screening.
- Amblyopia is usually preventable or treatable
- Early detection is key to effective treatment:
  - Assess red reflex
  - Determine visual acuity
  - Evaluate ocular alignment
- Life-threatening disorders may present as amblyopia e.g. Retinoblastoma
- Screening responsibility rests with primary care physician.
- Direct ophthalmoscope : 15x magnification, picture is erect
- Indirect ophthalmoscope: 3.5x magnification, picture is inverted



Direct ophthalmoscope: assessing red reflex



Normal red reflex



Direct ophthalmoscope

Pupil examination.  
It magnifies the optic disc 15 times. The image is erect and real.



Direct ophthalmoscope: examining retina



Asymmetric red reflex

- Left eye: normal red reflex and clear media.
- Right eye: absent red reflex & media opacity.
- Mention 2 causes of absent red reflex
  1. Congenital cataract
  2. Retinoblastoma
  3. Significant refractive error

# Amblyopia

## Treatment

- The first thing to do is to treat the underlying cause correct the refractive error, remove the media opacity surgically (is it ,for example, anisometropia, cataract or strabismus).
- The younger the child, the better outcome of amblyopia therapy.
- Occlusion Therapy (2nd thing)
  - The “gold standard” treatment for unilateral amblyopia is occlusion of the dominant eye to force fixation to the amblyopic eye.
  - Opaque adhesive patch applied to the skin.
  - Opaque contact lenses, cloth occluders applied over the glasses, and graded transparent filters.
- Amblyopia can be treated only during the first 5 years of life because of brain plasticity in children.
- The first 5 years of child age is the sensitive period where amblyopia can be reversed, after that it becomes more difficult. That's why we don't say it will go away
- It is too late to treat amblyopia at the school age.
- Strabismus and squint need to be treated.





# Leukocoria

## Definition

A white pupillary reflex. It differs from absent red reflex.

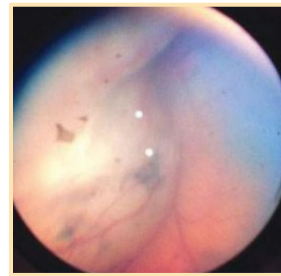
- ◆ Leukocoria in a child requires **urgent** attention , primarily because in most patients with retinoblastoma it is the first sign noticed .
- ◆ Secondly , a white pupil indicates a severely amblyopiogenic condition , which may be treatable. This is bad prognosis.
- ◆ Anatomic location is important in the differential diagnosis of Leukocoria, **parents notice it.**



This pic is without ophthalmoscope

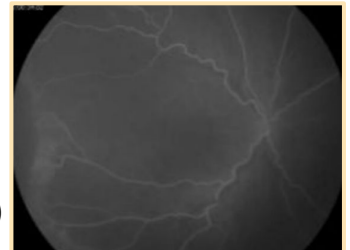
## Causes of Leukocoria

- **Retinoblastoma MOST IMPORTANT**
- Cataract
- Toxocariasis
- Coat's disease
- ROP
- PHPV
- Retinal detachment
- Coloboma
- Retinal dysplasia
- Norrie disease
- **Vitreous Hemorrhage doesn't cause white pupil; it happens behind the retina.**



## Work-up

1. History
2. Complete ocular examination
3. B Scan ultrasonography
4. Intravenous fluorescein angiogram (coats disease , ROP, retinoblastoma )
5. CT or MRI
6. Serum ELISA
7. Anterior chamber paracentesis



## 1. CATARACT

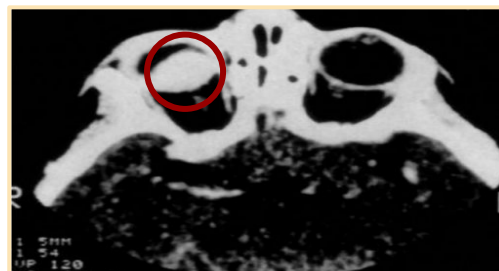
- Opacification of the lens.
- Congenital cataracts usually are diagnosed at birth.
- Unilateral cataracts are usually isolated sporadic incidents
- Bilateral cataracts are often inherited and associated with other diseases.
- **Cataract happens behind the pupil don't mix it with corneal ulcer which is anterior to it.**
- They require a full metabolic, infectious, systemic, and genetic workup.
- The common causes are hypoglycemia, trisomy (eg, Down, Edward, and Patau syndromes), myotonic dystrophy, infectious diseases (eg, toxoplasmosis, rubella, cytomegalovirus, and herpes simplex [TORCH]), and prematurity
- **Rubella triad: SNHL, cataract "hyperopia" and pepper salt retinopathy. Hearing loss, visual loss.**



# Leukocoria

## 2. RETINOBLASTOMA

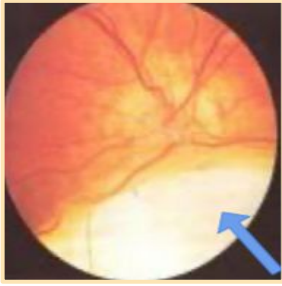
- Retinoblastoma is the most common intraocular tumor of childhood.
- **CLINICAL MANIFESTATIONS:**
  - Leukocoria (60%) is the 1st most common (MCQs)
  - Strabismus (20%)
  - If we see a child with squint we have to examine the fundus to rule out RB.
  - Vast majority become apparent before age of 3 yrs (2-4 y).
  - OTHER: Uveitis, Orbital cellulitis, Hyphema, Heterochromia, Glaucoma, Buphthalmos.
  - **Calcification** is another feature of retinoblastomas, usually occurring in necrotic areas. Calcium stains with H&E. It is worth identifying calcium in suspect eyes by ultrasound, or CT scan to differentiate retinoblastomas from other tumours.



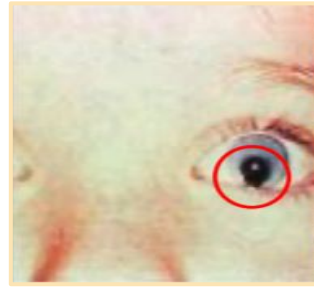
Why the pupil is white?  
b.c. of the retinal tumor.

- **MANAGEMENT:**
  - If diagnosed early laser is enough, late end stage diagnosis requires **enucleation**.
  - EMPIRICAL GENETIC COUNSELLING
  - ENUCLEATION removal of all the globe
  - Unilateral, poor visual prognosis
  - PLAQUE
  - 4-12mm +/- vitreous seeding
  - EXTERNAL BEAM
  - >12mm, multiple foci, only eye
  - LASER
  - Consider- indirect, xenon arc
  - Cryotherapy if <2dd in size
  - CHEMOTHERAPY, if intracranial extension
- If a child decides to come early (premature), the blood vessels have no time to reach the periphery, so he will be born with part of his retina not supplied by blood vessels > ischemia > neovascularization (to supply the ischemic parts, but they are fragile) > bleed > fibrosis > fibrosis shrinks > pull the retina causing RD.
- If a child is not caught at an early stage, he will end up with a blind state because there will be total retinal detachment!! Child will become blind.
- Any child born less than 1500g or 28 weeks must be screened in the NICU before going home to provide early intervention if needed (the screening is usually 4-6 weeks after birth).
- The less the weight, the earlier the birth, the sicker the baby > the more likely to get ROP.
- Rx: laser to kill (burn) the ischemic retina (to stop the demand), but it will destroy the peripheral vision (tunnel vision); however, central vision is preserved (20/20).

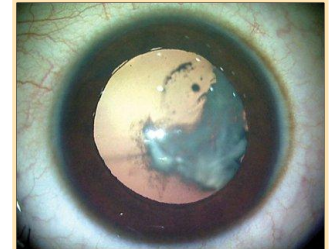
# Leukocoria



Leukocoria from scleral shining (Arrow) because of absence of retina.



will result in "keyhole" or "cat eye" appearance. (Tissue is not formed)



## Persistent hyperplastic primary vitreous (PHPV)

- A gray-yellow retrolental membrane may produce leukocoria, with the subsequent suspicion of retinoblastoma.
- In PHPV, the globe is white and slightly microphthalmic (small globe). Patients have no history of prematurity or oxygen administration.

b.c. If the baby premature it's possible to have retinopathy of prematurity. As in embryology primary vitreous usually disappears but if its still there it becomes hyperplastic.

## RETINOPATHY OF PREMATURITY (ROP)

- Vasoproliferative retinopathy affecting premature infants exposed to high oxygen
- Normally: in the 8th month half of the retina have blood vessels & the other half doesn't, it continues growing till delivery where it becomes fully mature, at that time when the baby exposed to oxygen it stops growing. Premature baby <32 weeks: blood vessels of the retina is on the middle didn't reach the periphery or even the macula so when the baby exposed to oxygen it will stop growing > ROP.

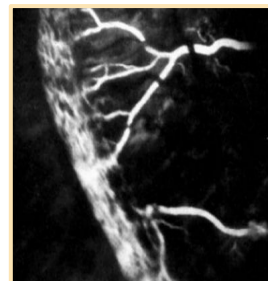
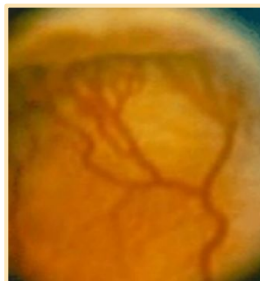
## INCIDENCE:

- Prematurity (<32/40)
- Birth weight (30% < 1000gm affected)
- Oxygen duration
- 90% ROP regresses spontaneously, 5% blindness



## Signs:

- Retinal Ischemia > no. of VEGF\* > neovascularization. ←IMP!! Pathophysiology
- Fibrous bands
- Retinal detachments
- Vitreous hemorrhage
- Leukocoria
- No hard exudates (no proteins) it founds in diabetic retinopathy or vein occlusion

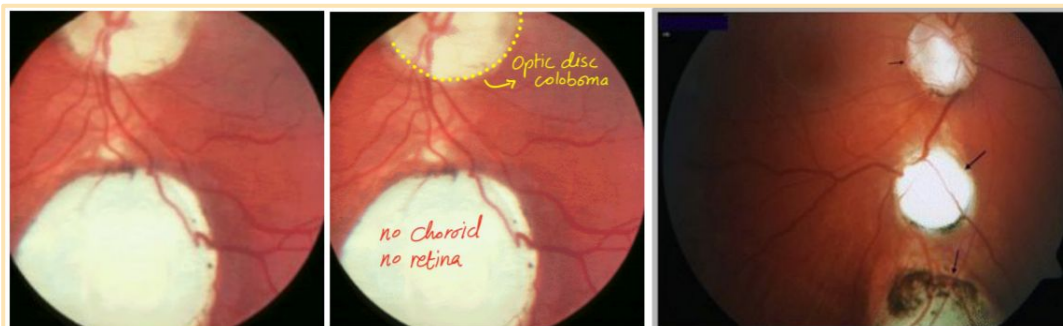


Fundus with fluorescein angiography, we do laser in the area with no blood vessels.

# Leukocoria

## 3. COLOBOMA

- Coloboma means no tissue development or agenesis.
- It can also be in the iris > iris coloboma > no iris tissue.
- Optic Disc Coloboma
- In embryogenesis the optic cup continues until it fuses inferiorly and the fissure closes.
- Due to failure of closure of foetal fissure inferiorly
- It always **inferior nasal** never superior nor temporal
- May be isolated disc or associated chorioretinal coloboma
- Usually sporadic, some AD
- Can be bilateral
- Visual acuity varies from normal to NLP if coloboma was away from optic disc & macula.
- If it was in the optic disc or the macula there might be no light perception.
- Degree of visual loss to area affected (iris, retina, choroid, or optic nerve head).



Extra:  
Right eye illustrating optic disc coloboma (small arrow) and two retinochoroidal colobomas (large arrows).



# Lecture Quiz



**CASE:** could come in the exam (MCQ)

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. How to approach this child:

## ❖ History taking:

- Family history, born at which week of gestation, mode of deliver

## ❖ Examination:

- Check the visual acuity, how to check the visual acuity in infant? fix and follow(following an object)
- CSM test "Central (no deviation) Steady (no nystagmus) Maintained (if you remove the cover can he maintain his eye in the center)".
- Check the extraocular movement: to rule out paralytic 6th nerve palsy, how to check? spinning *دور راسه*  
*اذا طبيعي عينه بتناظر على برى باتجاه دوران الراس .*
- Check the amount of deviation: Hirschberg test, Krimsky test.
- Check the refraction: cycloplegia is achieved by dilated drop called cyclopentolate (0.5% if child age less than 1, 1% if  $\geq 1$ yo).
- Check for the associations that we just mentioned above: cross fixation, DVD, IOOA, and latent nystagmus.

## ❖ Fundus exam:

- Check the optic nerve and retina.



# Lecture Quiz

1



Diagnosis: esotropia of the right eye. What happened after wearing the glasses? The esotropic eye is corrected & back to normal. What to do if the left eye becomes amblyopic? Cover the right eye to force the left & improve its vision.

2



Diagnosis: capillary hemangioma of right eye. Complication: amblyopia.

3



Lesion: right 6th nerve palsy. Cause: intra-cranial tumor.

4



Nerve injured: left oculomotor nerve. 2 most important systemic causes: HTN & DM.



The child looks normal in this picture.

5

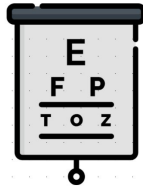


The faulty eye is the left eye because the lateral rectus is not working (no abduction) causing non-comitant strabismus.

6



This is a case of Brown syndrome (mechanical problem in the superior oblique).



OPHTHALMOLOGY TEAM

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