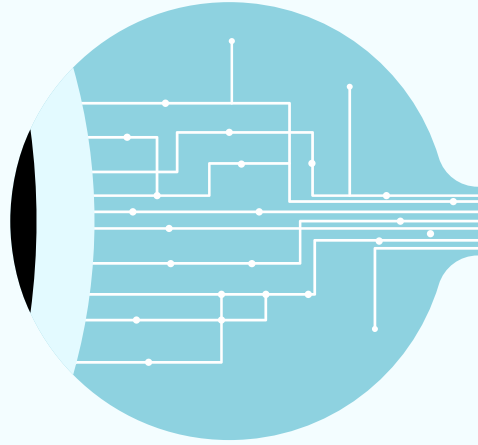


## Lecture 8



v1.00

Editing File



# Strabismus, Amblyopia & Leukocoria

Presented by: Dr. Abdullah Alotaibi

### Objectives of the course:

- no objectives were found

Doctor Abdullah Alotaibi didn't explain every thing in this file, any thing not mentioned by the doctor will be labelled in the slides as **extra text**, another doctor may come to A1 group with different slides in case of that we kept everything.

### Color index:

# Strabismus (حول)

## Definition

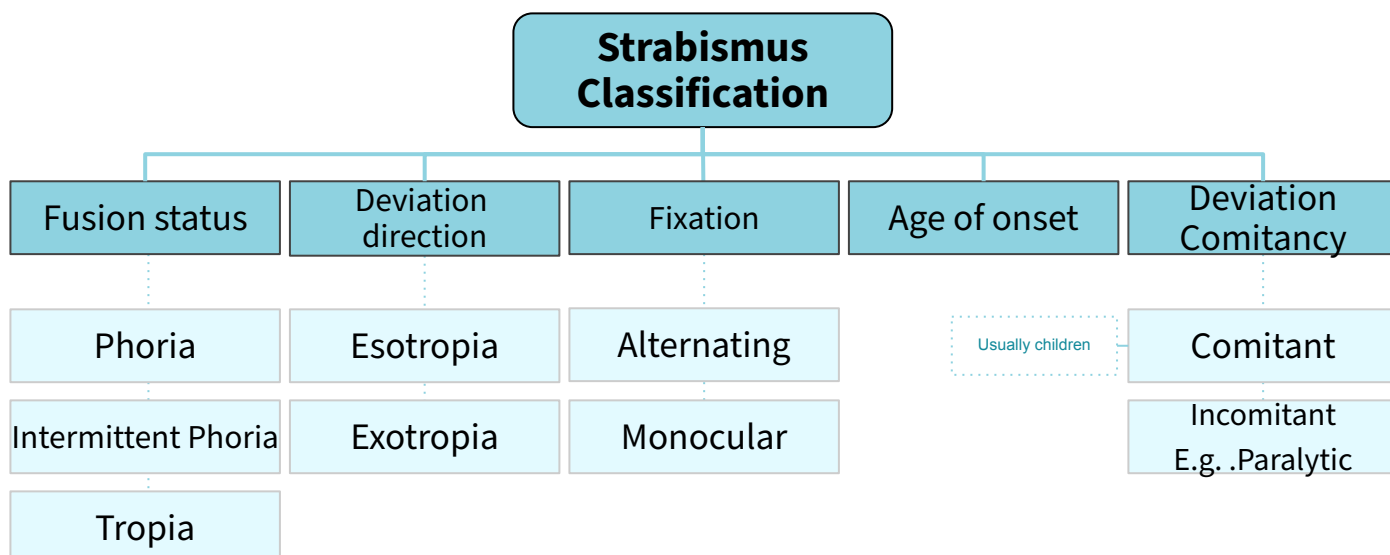
- Strabismus is a general term referring to ocular misalignment due to extraocular muscle imbalance.
- Strabismus occurs in approximately 3% of children under 3 years of age.
- Males and females equally affected.
- occurs when both eyes do not look at the same place at the same time; the eyes are unable to align properly under normal conditions. eye(s) may turn in, out, up, or down. often referred to as: cross-eyed, crossed eyes, cockeye, weak eye, wall-eyed, wandering eyes, and/or eye turn.
- Strabismus is associated with reduction of depth perception(binocular vision ) and, if onset is in adulthood, double vision occurs.
- 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 (binocular vision does not develop), and amblyopia (lazy eye) occurs .
- A cataract, eye tumor, or other eye disorder associated with reduced vision may also present with strabismus

## Causes of strabismus:

- Problems with the nerves that transmit information to the eye muscles
- Problems with the control center in the brain that directs eye movement
- In adults, can be caused by stroke, thyroid problems-tissue build up-, brain injury, or trauma

## Risk factors of strabismus:

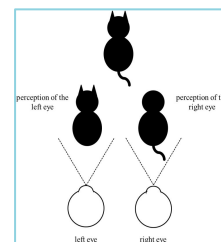
- Family history.
- Refractive errors: Hyperopia can cause strabismus.
- Medical conditions: People with down syndrome or cerebral palsy and people who have suffered a stroke or head injury



# Classification of Strabismus

## 1. Fusion status

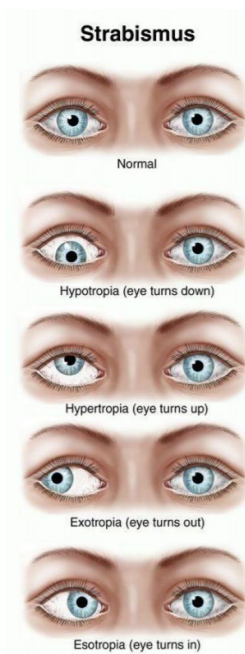
- **Orthophoria.**
- **Phoria:** Latent tendency of the eye to deviate and controlled by fusional mechanism. misalignment of the eyes that only appears when binocular viewing is broken by **Alternate cover test** which the examiner occludes one eye and then the other, switching the occluder back and forth to occlude the eyes without allowing the patient to fuse in between occlusion; to break fusion control **the squint will appear.** (438 note)
- **Intermittent Phoria:** Fusion control is present part of the time.
- **Tropia:** Manifest misalignment of the eye all the time.



(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)

## 2. Type of deviation

- Horizontal**
  - Esodeviation
  - Exodeviation
- Vertical**
  - Hyperdeviation
  - Hypodeviation
- Torsional**
  - Incyclodeviation
  - Excyclodeviation
- Combined**

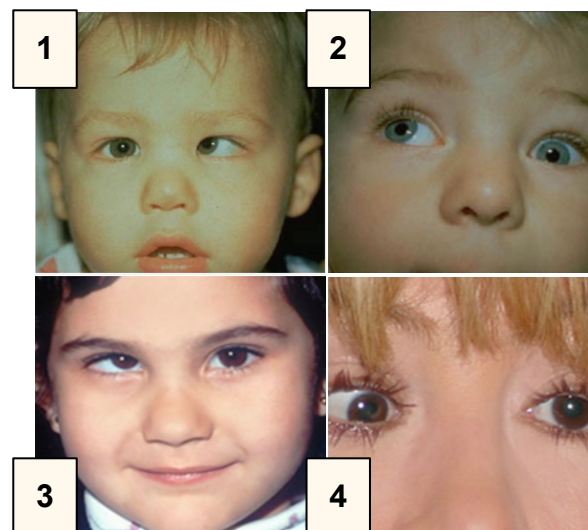


Vision at childhood differ from vision at adult; anatomy of eyes at birth is mature but physiological is not matured (visual acuity still grows (improves) until reaches 6/6 from birth to 6 or 7 age). Any abnormalities of eye in childhood period (like squint, cataract, ptosis and RE..) must treat them 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.(438 note)

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 ).(438 note)

## Type of deviation Examples :

1. **ESOTROPIA :** The left eye is turned inward note that the light reflection in the eyes is not symmetric
2. **EXOTROPIA:** The right eye is turned outward—again, not the light reflection in the eyes is not symmetrical
3. **HYPERTROPIA:** The right eye is turned upward—light reflection not symmetrical
4. **HYPOTROPIA:** The right eye is turned downward-light reflection in eyes is not symmetric
5. **Esophoria:** phoria nasally
6. **Exophoria:** phoria temporally



# Classification of Strabismus

## 3. Fixation

1. **Alternating:** Spontaneous alternation of fixation from one eye to the other.
2. **Monocular:** Preference of fixation with one eye, **Bad prognosis.** (fixed with patching the good eye, to stimulate the deviated eye)

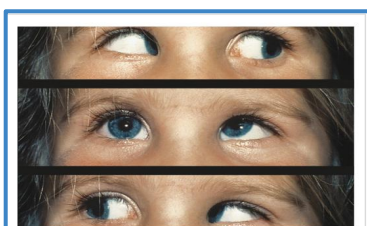
Why is it important to differentiate between alternating and monocular?

Child with Monocular may have amblyopia bc he just uses fixed eye and other eye will be neglected (not functioning); while Alternating switch between them so no amblyopia.

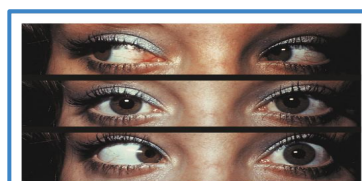
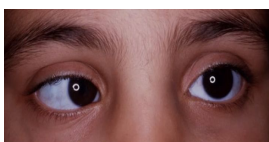
## 4. Variation of the deviation with gaze position or fixing eye

there is comitant type in kids, and the noncomitant type which is more in adult, the comitant is also 2 types: esotropia (common) and exotropia, eso is deviation of the eye towards the nose and there is risk of amblyopia and exo is lateral deviation & less risk.

1. **Comitant:** Angle of deviation that does not change with different gazes i.e. non paralytic or restrictive, most common in pediatrics, and deviation secondary to refractive error (most common), congenital muscle disease.
2. **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. If it was of neuronal origin, the squint is greatest in the field of the affected muscle. If it was, however, of mechanical origin, the squint is greatest in the direction away of the affected muscle (such as thyroid eye disease)



**FIGURE 6-3** Concomitant strabismus. In the views presented here, the misaligned eyes exhibit about the same degree of inward deviation (esotropia) in each position of gaze.



**FIGURE 6-4** Incomitant strabismus. The eyes appear straight in right gaze (top) and straight-ahead gaze (middle), but a misalignment is obvious in left gaze (bottom), indicating a paralysis of the left lateral rectus muscle or a restriction of the left medial rectus. These eye positions would be found in a left sixth cranial nerve palsy.

## 5. Age of onset

- **Congenital/infantile:** (early childhood-6 months)
- **Acquired:** (after 6 months, at 2 years): can also develop in adulthood as a result of lack of treatment during childhood, the affected individual usually experiences double vision. This occurs because the brain, which no longer has the "plasticity".
- Intermittent: expected until 4 months of age.

# Clinical evaluation

- **Patient history:** A comprehensive history is necessary to assess symptoms, health problems, and medications
- **Visual acuity:** Measurements are taken to assess extent to which vision is affected
- **Refraction:** Conducted to determine the lens power necessary to compensate for any refractive error
- **Alignment:** Assessing how eyes focus, move, and work together
- **Eye exam:** Assessing the structures of the eye (internal & external)

## Examination

1. **History** ask about age of onset, family history, history of trauma. symptoms, health problems, and medications..etc
2. **Inspection**
3. **Assessment of monocular eye function**
  - Visual acuity:(Measurements are taken to assess extent to which vision is affected)
    - Preverbal children within 1 -2 years can't take vision:
      - CSM (Central: no deviated, Steady: no nystagmus, Maintained: fixated).
      - OKN (Optokinetic nystagmus)
      - Preferential looking.
      - Visual evoked potential
    - Verbal children:
      - Symbol tests: single illiterate E.
      - Allen pictures.
      - HOTV letters.
4. **Assessment of binocular eye function:** (Hirschberg and krimski's tests are important according to first lecture doctor)

### Hirschberg test:

Simple test using light(torch) **Without Prism** 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

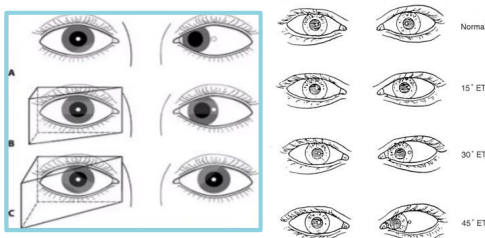
Cover- uncover test to **diagnose tropia**.  
(move from outside to center = exo, move from inside to center = eso)

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

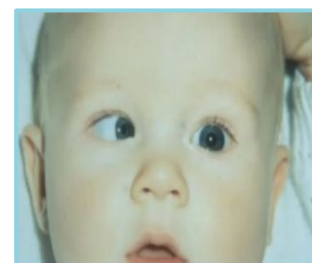
Breaking fusion control, for phoria.

### 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.



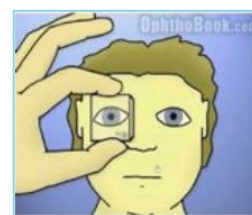
Hirschberg test

# Examination

## Cont.

### Hirschberg test:

- 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.**
- 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)



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

- **IMPORTANT (Always comes in exams)**
  - **Cover-uncover test → detect tropias.**
    - **Cover:** To see anything manifest (TROPIC) **لما تغطي العين الطبيعية العين الأخرى اللي مو مغطية اللي فيها حول بتصير في النص**
    - **Uncover:** uncover: To see anything hidden (phoria) **العين اللي تحت الغطاء بتبين اذا فيها حول خفي**
  - **Cross-cover test → detect phorias and tropias.**
- Ocular movement (motility) is different from eye alignment:(438 note)
  - **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.

### 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.(438 note)

### Cycloplegic refraction

To relax eyes and see **real refractive error (release accommodation)**(438 note)

- Tropicamide: Short half life (20min)
- Cyclopentolate: Intermediate two days to three
- Atropin: Two weeks

### General Management of squint:

- Rule out pathological causes like cataract by examining funds by fundoscope.
- Correct (refractive error,cycloplegic refraction), ( hyperopia may 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



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

- **Esotropia can be divided as :**
  - Infantile esotropia.
  - Acquired esotropia. (Accommodative, partially accommodative, viral illness, Diabetes)
- the esotropia divided Into 2 types infantile esotropia which is comitant type that means the nerve is normal and the abnormality is in the muscle which is the medial rectus muscle very strong and the eye is deviated to the nose and it comes to the kids who is below 6 month. Treated by surgery.
- acquired esotropia is the comitant type which is come to kids after 6 months specially on kids who have hyperopia and the hyperopia will cause accommodation and very long duration of accommodation will result in strabismus that we call it acquired esotropia treated by glasses.

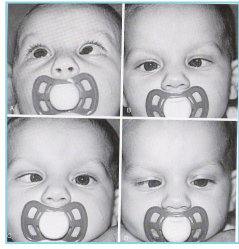
## A- Infantile Esotropia

- Inward deviation or the eyes noted before the patient reaches 6 months.
- When the eyes are misaligned in childhood, binocular vision, or the ability of the brain to use the two eyes together, does not develop.
- Classic infantile esotropia is constant and involves a large angle of deviation exceeding 20 prism diopters (PD) on corneal light reflex measurement.
- Infantile esotropia may be associated with a spectrum of clinical presentations, including amblyopia, impaired binocularity, central scotomas.
- **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.
- **Treatment:**
  - Treat amblyopia by **occluding the good eye.**
  - **Surgery** for the extraocular muscles **by weakening of medial recti**
  - Surgical correction of strabismus. **Infantile esotropia +a large angle of deviation + low refractive error (not significant)= surgery**



notice that she is looking to us from angle so without light reflex we think that she has Left eye Esotropia

But!! after doing light reflex it appears that the light reflex of left eye is in the middle and the real deviated eye is the right (The eye is inward and the light reflex outward so she has Esotropia ) and since she has it from 4 month old we call it **Infantile esotropia.**



Alternating esotropia refers to how a patient fixates. Patients with an alternating fixation pattern switch fixation between each eye. This will reduce the risk of amblyopia

Infantile Esotropia



### Depth perception:

The ability to judge how far away an object is as well as how far away objects are from each other.

# Types of Strabismus

## B- Acquired Esotropia.

Accommodation will cause convergence and miosis

### 1- Accommodative Esotropia

- Acquired after 6 months of age usually around 2 years of age.
- **Associated with hyperopia**
- Refractive error usually (+3,+4)
- May precipitated by acute illness or trauma
- Start intermittent and if not treated become constant

#### Treatment.

1. Perform cycloplegic refraction on all children by using the retinoscope and loose lenses. Cycloplegia is achieved with Mydracyl 1% and cyclogyl 1%. Example of cycloplegic agents to measure his refraction (drops):
  - a. Atropine
  - b. Cyclopentolate
  - c. Tropicamide
  - d. **phenylephrine** (will not cause cycloplegia it will cause dilation of pupil instead (**MCQ**) all can cause cycloplegia except phenylephrine)
2. Treat the cause: (mainly glasses for hyperopia).
  - Wearing glasses for pediatric is mandatory (convex lens to relax the accommodation > back to normal)
  - **Refractive error correction by glasses will treat the condition.**
  - **If the farsighted glasses control the crossing of the eyes, eye muscle surgery is never recommended!**



Type of squint:  
**accommodative esotropia**  
Type of refractive error: **Hyperopia**

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



**Accommodative Esotropia:** this kid has hyperopia of +5, when we look to him without glasses he has Esotropia and after wearing glasses it disappears (MCQ) Accommodative Esotropia happen in? **Hyperopia**

### 2- Partially Accommodative Esotropia

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





# Types of Strabismus

## B- Acquired Esotropia Cont.

### 3- Esotropia associated with Viral illness

- Acute, Often self limited, will spontaneously resolve in 3-6 months.
- Not improved with hyperopic glasses.
- Consider ruling out neoplastic causes.
- Treat/prevent amblyopia in the meantime



### 4- Esotropia associated with Diabetes

- CN 6 usually affected.
- Isolated unilateral ischemic palsy
- Usually resolves after 4-6 months.
- Consider Botox in the meantime, to The medial rectus

- If the case was **young** and diabetic think about imaging.
- What's the cause of Esotropia in viral illness and in diabetes?
  - nerve supplying LR (CN 6 ) abducens nerve palsy
  - sometimes due to increased intracranial pressure (hydrocephalus or tumor)
- Acute ET and you didn't find hyperopia and he has **6th** nerve palsy its an indication to do : Imaging **MRI**
- nerve that can easily be affected by head trauma in car accident and airbags = **4th nerve (Trochlear)**
- **So 4th and 6th are prone to be affected**
  - **4th = any trauma from outside**
  - **6th = any problems in the brain (inside)**

## Pseudo Strabismus حول كادب

- Occur commonly in patients with flat nasal bridge and prominent epicanthal fold which tends to obscure the nasal portion of sclera like in children and Asian.
- 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 with no deviation (normal exam; because they are central).**
- Pseudoesotropia is a condition in which alignment of the eyes is straight. (also known as orthotropic); however, they appear to be crossed.

### Hirschberg test = light reflex

You might think that his eyes are deviated inward but after light reflex it reveals that he has Pseudo-Strabismus (because of depressed nasal bridge and prominent epicanthal folds )

There is another test for detecting Pseudo-Strabismus other than light reflex :

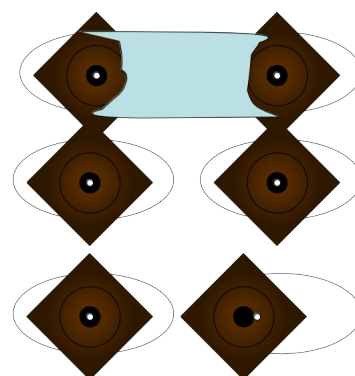
cover-uncover test (when you cover the normal eye, the deviated one will be corrected and it will move to the center ).



Pseudo ET

Orthophoria

Esotropia



# Types of Strabismus

## 2

### Exotropia

- horizontal form of strabismus characterized by visual axes that form a divergent angle
- not as common as esotropia
- Often familial
- Represent 5% to 10% cases of squint
- Make sure they have BCVA glasses
- Intermittent exotropia can breakdown over time, check serial stereo. If worsening think of surgery.
- Most common time of pediatric surgery is before 7 years old
- Congenital
- Not related to hyperopia



Exotropia



### Intermittent exotropia

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



Diagnosis: strabismus (**Intermittent** exotropia).

- The commonest exodeviation
- Large phoria, controlled by the strong fusional convergence.
- spontaneously breaks down into XT
- if child is tired, daydreaming or sick
- In adults, after taking alcohol or sedatives

Examination: corneal light reflex shows outward deviation.

Treatment: if indicated lateral rectus muscle recession.

### Sensory exotropia

- Not functioning eye for ex amblyopia (poor vision could be due to cataract, retinal detachment, optic atrophy...) => cause eye to deviate.(438 note)
- 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.



This patient had an injury to his right eye a long time ago and he lost his vision from it (at the beginning it was very minimal Exotropia and with time it became worse and worse )what's the cause?

Amblyopia or deprivation.

He has **Sensory Exotropia** because he doesn't use it.

# Types 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.

## Treatment of Exotropia :

### ◆ If amblyopia is present

1-Eye patching

2- treat other causes of amblyopia if present



The stronger eye is patched to force the brain to interpret images from the strabismic eye. Eye patches will not change the angle of the strabismus.

### ◆ Treat refractive errors with

1-Eyeglass or Contacts

Eyeglasses or Contacts are used to improved the positioning of the eye(s) by modifying the patient's reaction to focus. (accommodation)

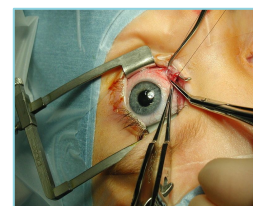


### ◆ Prisms

Prisms are used to modify the way light and images hit the eye, it can help in double vision

### ◆ Eye Surgery:

- Surgery may be necessary in an attempt to align the eyes
- During surgery, the muscle positions will be changed or the length of the muscles will be changed



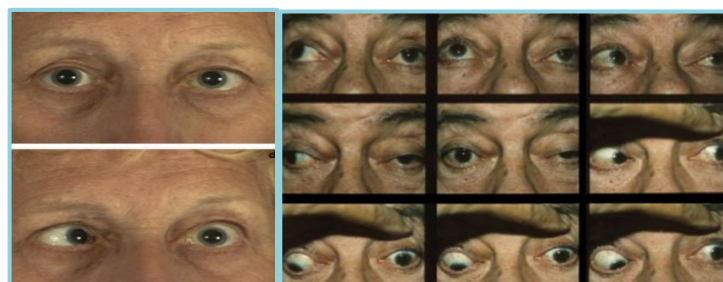
Surgery on medial rectus muscle of the left eye.

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

### 6th Nerve Palsy

- Incomitant esotropia
- Limitation of abduction
- Abnormal head position

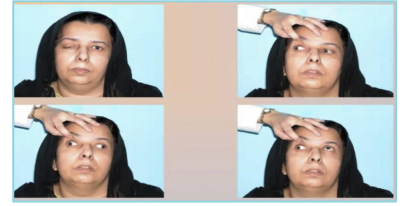
Neuro ophthalmology lecture is very imp. esp 6th  
C- Paralytic strabismus nerve palsy "exam"



# Types of Strabismus

## 3rd Nerve Palsy most common non-comitant strabismus is the third Nerve Palsy in adult

- 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.



## 4th Nerve Palsy

- Congenital or acquired
- **Hypertropia**(so Near reading ( downgaze)will cause double vision) of the affected eye with excyclotropia
- Abnormal head position

## 4 Special Types of Strabismus

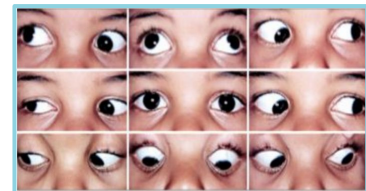
### Duane Syndrome (like 6th CN palsy)

- Limitation of abduction with mild limitation of adduction
- **Diagnostic test: On adduction, Retraction of the globe with narrowing of the palpebral fissure. This is how to differentiate between 6th CN palsy and Duane syndrome as in the pic duane syndrome in the left eye. Problem with horizontal movement**
- Upshoot or downshoot on adduction
- Pathology faulty innervation of the lateral rectus muscle by fibers from medial rectus leading to stimulus contraction 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.



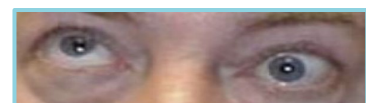
### Brown Syndrome

- Problem with vertical movement.
- **Limitation of elevation on adduction.**
- Restriction of the sheath of the superior oblique tendon.
- Treatment needed in abnormal head position or vertical deviation in primary position or has amblyopia.



### Thyroid Ophthalmopathy

- **Restrictive myopathy commonly involving inferior rectus, then medial rectus and superior rectus.**
- Patients presents with hypotropia, esotropia or both. **Orbital diseases are very important**



# Types of Strabismus

## Surgery of Extraocular Muscle

1. **Recession:** weakening procedure where the muscle disinserted and sutured posterior to its normal insertion.
2. **Resection:** strengthening procedure where part of the muscle is resected and sutured to its normal insertion.  
**(MCQ) -which procedure strengthens the muscle? / which procedure weakens the muscle?**
3. **Complications:**
  - Post operative diplopia.
  - Lost or slipped muscle.
  - Infection, Perforation of sclera
  - Anterior segment anesthesia
  - Conjunctival granuloma and cyst

### (MCQ)

- If someone has esotropia, what would you do as surgery?

we will do Medial Rectus Recession (weakening)

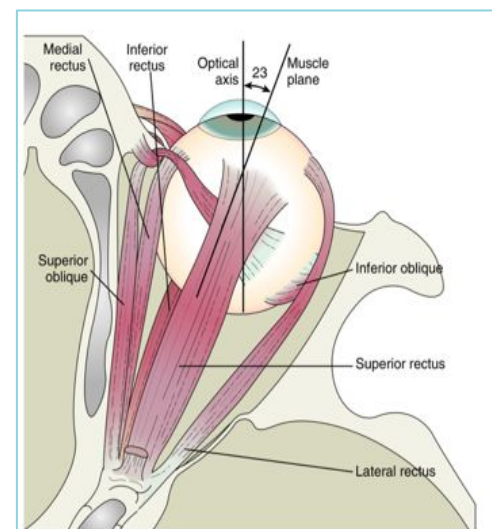
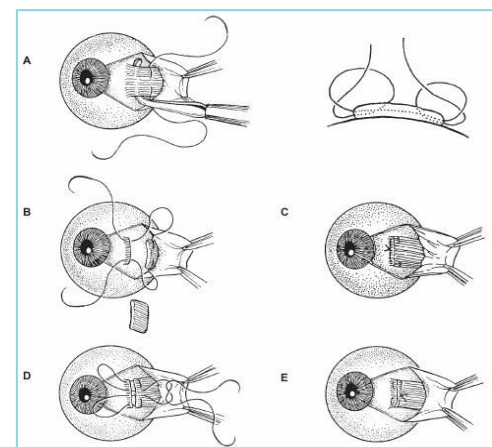
or

we can also do Lateral Rectus Resection (strengthening)

but we like to start with Recession (weakening) because it's reversible and we can put it back.

- If someone has exotropia, what would you do as surgery?

we will do Lateral Rectus Recession (weakening)

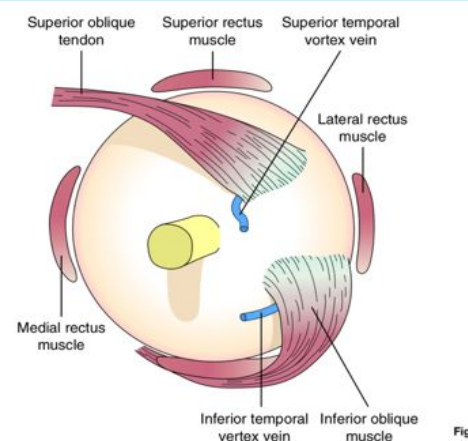
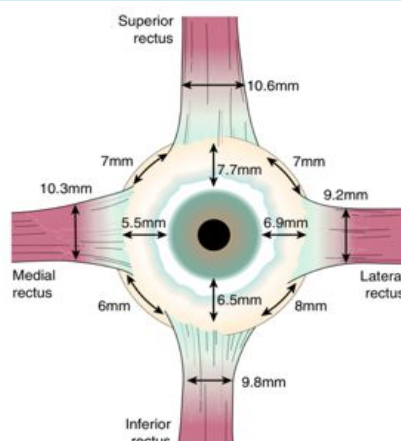


- **you should know the anatomy well**
- muscles / innervation/origin /insertion will help you to understand the move and how to treat
- If we took the eye as a ball and we divide it into two halves all extraocular muscles will be inserted in front of this equator except two muscles : obliques will be inserted posterior to the equator.

عبر كذا ان الابسرسى حوها مو حاي بحت واحد حاي بلوه ولدلاله الاكسى حوها 3 رى ما موجود بالحدول فى الصفحة النباليه.

- so all muscles have actions similar to its name except obliques different from its name (because of its insertion)

- recti have a space between it and the limbus.
- The muscle that have the shortest distance between it and the limbus: Medial rectus=5.5
- The muscle that have the longest distance between it and the limbus: Superior rectus=10.6



# Types of Strabismus

Table 5-9. Extraocular muscles

Muscle	Muscle length (mm)	Tendon length (mm)	Arc of contact (mm)	Anatomic insertion from limbus	Action from primary position	Origin	Innervation
Medial rectus (MR)	40	4.5	7	5.5 mm	Adduction	Annulus of Zinn	CN 3, (inferior division)
Lateral rectus (LR)	40	7	12	6.9 mm	Abduction	Annulus of Zinn	CN 6
Superior rectus (SR)	40	6	6.5	7.7 mm	1. Elevation 2. Intorsion 3. Adduction	Annulus of Zinn	CN 3, (superior division)
Inferior rectus (IR)	40	7	6.5	6.5 mm	1. Depression 2. Extorsion 3. Adduction	Annulus of Zinn	CN 3, (inferior division)
Superior oblique (SO)	32	26	7-8	Posterior to equator in superotemporal quadrant	1. Intorsion 2. Depression 3. Abduction	Orbital apex above annulus of Zinn	CN 4
Inferior oblique (IO)	37	1	15	Posterior to equator in inferotemporal quadrant	1. Extorsion 2. Elevation 3. Abduction	Behind lacrimal fossa	CN 3, (inferior division)
Levator palpebrae	40	14-20	—	Septa of pretarsal orbicularis and anterior surface of tarsus	Lid elevation	Orbital apex above annulus of Zinn	CN 3, (superior division)

- usually we ask about the primary movement
- MCQ: what's the movement of obliques??
- primary action of it: Intorsion and Extorsion

Superior oblique Superior rectus Superior temporal

Superior



- He has Right eye esotropia (big angle 60)
- So what kind of surgery did we do ? we worked on 3 muscles
- **Medial Rectus Recession (weakening) For right and left**
- **Lateral Rectus Resection (strengthening) For right.**
- **If not solved right away -> amblyopia will develop.**

## Other Treatment Modality:

- **Botox Therapy**
  - Used as an alternative to eye muscle surgery
  - The drug will temporarily relax the eye muscle ( paralyze), which will allow the opposite eye to tighten and straighten.
  - The effects are short term about 3 to 8 weeks.

(MCQ)

**If someone has esotropia, where would you inject it?** we will do it on the Medial Rectus (The botox weakening the muscle)

# Amblyopia

## Definition

Ambly=lazy / opia=eye

- Reduction in visual acuity in one or both eyes that cannot be completely accounted for by any clinically apparent organic abnormality.
  - Amblyopia is a form of reversible cerebral visual impairment (reversible if catch in early age)
- (MCQ): It's a unilateral condition **ONLY** answer: **NO! It can be Bilateral**)

- Always occurring before the age of 7
- Prevalence of 2%-4% in the general population
- It's the single most common form of monocular vision impairment in the first 4 decades of life.
- Always associated with disease of the visual system
- For Vision to develop normally: 1- both eyes are straight 2- No refractive error 3- Clear media( So any problem in these 3 and not detected during the developmental period it will lead to amblyopia)

## Diagnosis

- ↓ BCVA (Best Corrected Visual Acuity)  $\leq 20/40$ .
  - **OR**; A difference of 2 lines of Snellen acuity between the amblyopic eye and the normal eye
- E.g. Right eye 20/20, Left Eye 20/60 (more than two lines). We consider the left eye amblyopic.
- Pre-verbal kids: Use preferential looking test, fixation preference if associated with strabismus
  - Older kids (3 and older): Allen pictures or Tumbling E.

Clinical characteristics of amblyopia include : (اسماء اصاوبه عسان يحقو عبر اللي اندكر بالعريف) (MCQ!!!)

-No change or even improvement in BCVA through a 3.0 neutral density filter( normally changes in non-amblyopic eye)

-Improvement in vision when tested with **single** optotypes  
(single optotype :when we use **single** optotype without anything around it)  
(the amblyopic patient if you test him with **single** optotype he will score more-vision will improve)  
(-Crowding phenomenon: if we use multiple optotypes he will score less-vision will be less-)

## Classification of Amblyopia:

### 1. Abnormal binocular interaction:

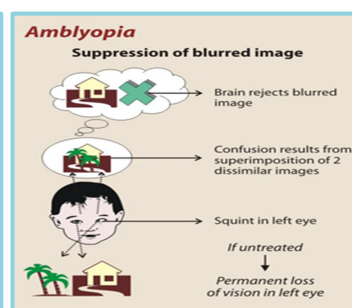
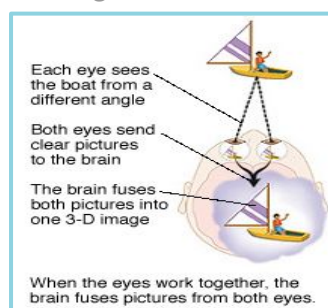
- In strabismus amblyopia occur in at least 40%

### 2. Blur/distortion of the visual image due to :

- Uncorrected refractive errors / Media opacities ( as lens opacity or corneal scarring)  
cataract/vitreous hemorrhage/edema in cornea

### 3. Multiple mechanisms:

Normal



what happen if patient have deviated eye (strabismus): brain won't be able to fuse the 2 images so the brain will neglect one image (that's why amblyopia occurs in strabismus)

# Amblyopia

## Classification of Amblyopia:

### 1. Strabismus.

In this pt, image from the left eye will fall away from the fovea -> distorted image -> brain will suppress left eye -> eye will not be used -> lazy eye (amblyopia)



- Strabismus is the most common amblyopia-inducing condition, 40% of children with a manifest strabismus will develop amblyopia. (because we have one eye which is straight and normal so light will come to it and go to the centre of retina (macula and fovea) which has big amount of Neuro sensitive receptor and then will be carried by the optic nerve and will be very clear image that goes to the brain, but the other eye which is deviated the light will be in the peripheral of the retina and that will cause Blurred image to go to the brain, and the brain as we know will take these two images and convert them into one but in this case one is clear and the other is a blurry so the brain will cancel the blurry vision, and also will cancel any image that comes from the deviated eye, with time the eye will be called lazy eyes (amblyopia) because the brain cancel any image that comes from it, so we need to treat them earlier before 7 years or their will be a permanent damage and the brain will not depend on.
- Esodeviation is more common than exodeviation, so Incidence of amblyopia is greater in esotropic patients than in exotropic patient

### 2. 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
- type of amblyopia **is more common** in patients with anisohypermetropia than anisomyopia. Small amounts of hyperopic/astigmatic anisometropia, such as 1.5 diopters, can induce amblyopia. In myopia, mild myopic anisometropia up to - 3.00 diopters usually does not cause amblyopia.
- Inhibition of the fovea occurs to eliminate the abnormal binocular interaction caused by one defocused image and one focused image.
- Treatment is by prescribing glasses to correct refractive errors
  - The only way to find out anisometropia is to screen the child, at 6m, 2 y, 4 y, 5 years (4 times).



### 3. Visual deprivation

- The least common type of amblyopia, <3% of cases BUT it has the most potential to cause severe amblyopia. more than strabismic or refractive amblyopia.
- Amblyopia results from disuse or under-stimulation of the retina.
- This condition may be unilateral or bilateral. Examples include cataract, corneal opacities, eyelid ptosis, eyelid hemangioma, and surgical lid closure.
- A newborn with **cong. Cataract** only needs 1-2 weeks to develop amblyopia

below are examples for causes of deprivation :

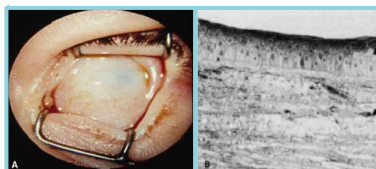
What do you feel about the size of the eye ?

Large and also its opaque -and it is secondary to congenital glaucoma. They call it bull's eye (عين البقر)



Where is the cornea ?

The cornea replaced by adjacent tissue (sclera) so we call it sclerocornea



ptosis affect visual pathway



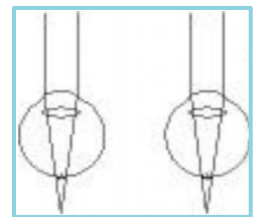


# Amblyopia

## 4. Ametropic Amblyopia (Refractive AMBLYOPIA):

- **Hyperopia:** will cause **Bilateral** Amblyopia if the patient have:
  - 1- Greater than +4.50 diopters
  - 2- Highly hyperopia may causes strabismic amblyopia
- **Myopia:** Rare why? because he sees near things and childrens world is near to them so he wont have it unless it very high number -8 or -7
  - 1- Amblyopia is rare if symmetric myopia, because these patients simply decrease the working distance to focus the image.
- **Astigmatism (Meridional).**

mostly with one of above either Astigmatismal Hyperopia or Astigmatismal Myopia  
it can cause it if more than 3 or 3.5
- Uncorrected high hyperopia is an example of this bilateral amblyopia
- If both eyes were +7 we call it Ametropic Amblyopia.(438 note)
- Every refractive error can cause different Amblyopia; for example:(438 note)
  - 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.(438 note)



## 5. Organic:

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

## SCREENING: IMPORTANCE

- How to treat them? How to know if they even have it or not? By Screening.
- Amblyopia is usually preventable or treatable, half of the cases undetected until age 5 years.
- All newborn infants should be screened, Early detection is key to effective treatment:
  - Assess red reflex to check for media opacities
  - Determine visual acuity
  - Evaluate fixation preference, ocular alignment, and eye diseases by 6 months of age.
- Life-threatening disorders may present as amblyopia e.g. **Retinoblastoma**

### The various causes of Amblyopia among schoolchildren in Qassim province, Kingdom of Saudi Arabia:

Causes	Percentage
Anisometropia	77.72
High bilateral refractive error	16.84
Esotropia	3.46
Exotropia	1.98

- **Anisometropia** : بقاوب المجاس بين عيني وعيني
- it's the most common cause of Amblyopia around 80%
- Here in ksa we are opposite to western
- In our region Anisometropia is the most common cause whereas in western countries is strabismus
- why? because in western countries they have good health care system and they can screen kids and catch kids with refractive error early and treat it.
- For us we don't have screening .

# Amblyopia

## SCREENING:



Direct ophthalmoscope: assessing red reflex



Direct ophthalmoscope: examining retina



Asymmetric red reflex



Normal red reflex



Direct ophthalmoscope

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

- 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

## Treatment

- The first thing to do is to treat the underlying cause correct the refractive error to provide a clear retinal image for the amblyopic eye, 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)
  - (occlude the good eye to force the brain to give attention to weak eye)
    - 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.
  - The first 5,7 years of child age is the sensitive period because of brain plasticity in children, where amblyopia can be reversed, after that it becomes more difficult.  
After 7yrs of age it's very difficult to correct, you can't force the brain to use the amblyopic eye.



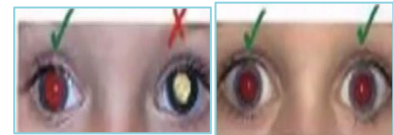
# Leukocoria

## Definition

Leuko : White / Coria :Pupil

white opacity of the pupil.

- ◆ 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.
  - White pupils are Abnormal
  - Normal pupils are Red
  - Why is it important to have red reflex? It indicates clear cornea, clear vitreous, normal retina, Anterior chamber,lens
  - Red reflex comes from pigmented epithelium in the retina



Rt: normal  
Lt: Congenital cataract

## Leukocoria work-up

1. Family history -because cataract can be congenital -
2. Prenatal and postnatal history
3. Trauma history -unilateral cataract-
4. Parents examination when indicated
5. **Outpatient** Department examination vs. examination under sedation or examination under anesthesia
6. Vision assessment
7. IOP
8. Alignment and nystagmus
9. SLE-slit lamp examination-
10. Fundus examination

## Causes of Leukocoria

### A-External and Corneal causes STUMPED

★ **STUMPED: -IMP They will ask about it- (SAQ: What are the causes of corneal opacity)**

causes of congenital clouding of the cornea:

Sclerocornea

Tears in the Descemet membrane secondary to birth trauma or congenital glaucoma

Ulcers-Keratitis

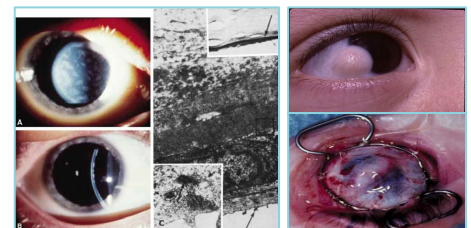
Metabolic

Peters anomaly

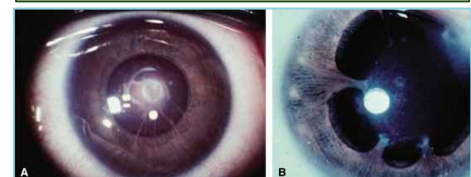
Edema (CHED)

Dermoid cyst

**shown below:**



Dermoid cyst

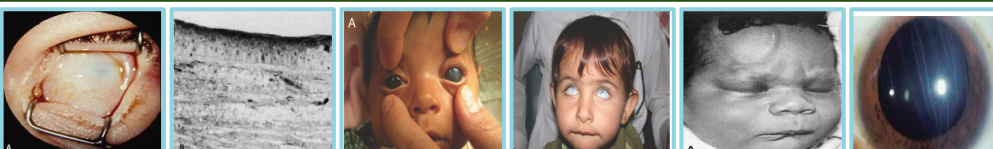


Peters anomaly

Tears in the Descemet membrane secondary to birth trauma (Trauma during delivery) usually in past when they have obstructed delivery they use forceps and it might press on corena and there will be tear in the descemet membrane (Vertical)

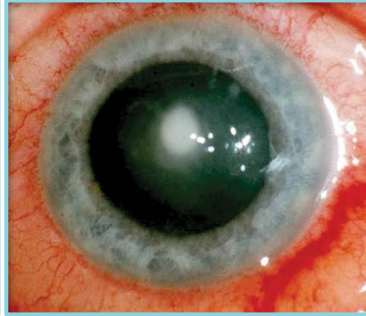
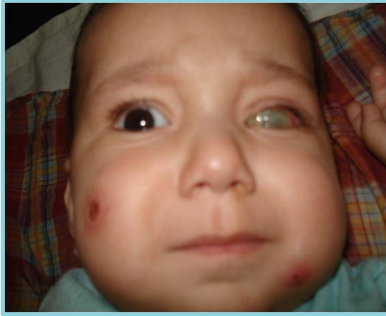
**(MCQ) What's the difference between birth trauma and congenital glaucoma in the orientation of the scar**

birth trauma: it will be Vertical  
congenital glaucoma: Horizontal



# Leukocoria

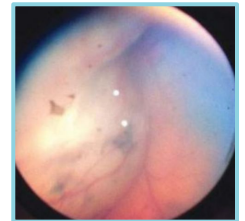
## A-External and Corneal causes STUMPED



(signs of infection)  
red eye ,edematous,patient in pain,  
swelling ,loss of clarity  
This is ulcer

## B-Anterior segment and Lens causes & Posterior chamber and Retinal causes :

- Cataract
- Retrolental mass (PHPV, ROP, Retinal detachment),
- Tumor **Retinoblastoma MOST IMPORTANT**
- Exudates (FEVR, Coats' disease)
- Change in retinal pigment (high myopia, myelinated nerve fiber, retinal dysplasia)
- Infections (toxoplasmosis,endophthalmitis)
- Coloboma
- Retinal dysplasia
- Norrie disease
- **Vitreous Hemorrhage doesn't cause white pupil; it happens behind the retina.**



### 1. CATARACT

- Opacification of the lens.
- can be congenital or acquired, usually causes blurred vision and glare.
- Congenital cataracts usually are diagnosed at birth.
- Unilateral cataracts are usually isolated sporadic incidents
- Bilateral cataracts are often inherited and associated with other diseases.
- 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.**

### Congenital cataract :



Bilateral cataract

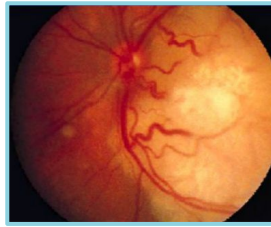
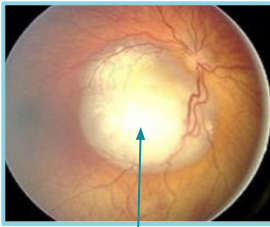


Unilateral cataract  
cornea is intact  
the problem is  
retrocorneal  
at the level of pupil

# Leukocoria

## 2. RETINOBLASTOMA

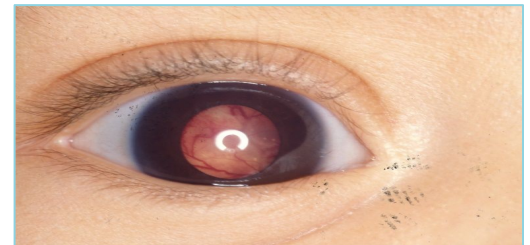
- Retinoblastoma is the most common primary, malignant intraocular tumor of childhood, but still rare tumour
- **CLINICAL MANIFESTATIONS:**
  - Life threatening cause.
  - Leukocoria (60%) assume that every patient with leukocoria is having retinoblastoma until proven otherwise
  - Strabismus (20%)
  - 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
  - It results from malignant transformation of primitive retinal cells before final differentiation.
  - Uveitis, Orbital cellulitis, Hyphema, Heterochromia, Glaucoma, Buphthalmos.
  - **Calcification**, 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



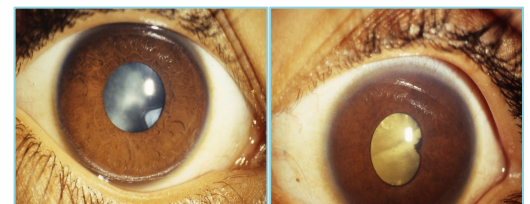
Why the pupil is white?  
b.c. of the retinal tumor.

- **MANAGEMENT:**
  - EMPIRICAL GENETIC COUNSELLING
  - **ENUCLEATION** removal of all the globe (resection)
  - **PLAQUE** radiotherapy, 4-12mm +/- vitreous seeding
  - **EXTERNAL BEAM** >12mm, multiple foci, only eye
  - **LASER**: indirect, xenon arc
  - Cryotherapy if <2dd in size
  - **CHEMOTHERAPY**, if intracranial extension

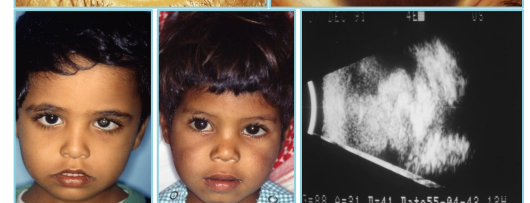
**Cornea is intact**  
**Nothing in the level of pupil**  
**lens is clear**  
**But in the level of retina there is blood vessels which is so clear**  
**Most likely :Retinal detachment or Retinoblastoma**



**Cornea is intact**  
**What's happening is the level of lens**  
**this is cataract**  
**But!! When we did a B-scan for him we found that vitrus is full of**  
**retinoblastoma and its stuck to the lens from behind and the lens lost**  
**its clarity**



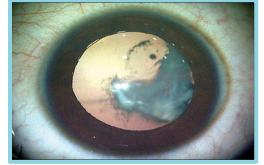
**So when a child comes to you with cataract You should examine his**  
**eyes with ultrasound to know what's the cause of cataract ?is it**  
**congenital or secondary to vitreous problem (retinal detachment or**  
**retinoblastoma )**



# Leukocoria

## 3. Persistent hyperplastic primary vitreous (PHPV) (old name) new: persistent fetal vasculature.

- congenital condition caused by failure of the normal regression of the primary vitreous. usually associated with unilateral vision loss. (When it attached to lens it will cause opacity)
- Patients have no history of prematurity or oxygen administration.



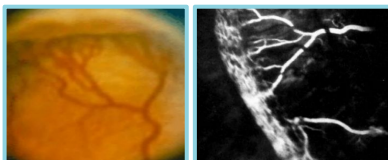
## 4. RETINOPATHY OF PREMATURITY (ROP)

- Blinding disease if you don't catch early, screening is important
- Vasoproliferative retinopathy affecting premature, low birth weight infants exposed to high oxygen (usually before 38w)
- Normally in fetal life the vessels developed from central to peripheral part of the eye if the fetus delivered early at 24 gestational age the development of these vessels is incomplete and the peripheral part of the eye will be empty with no vessels so we treat it by killing the peripheral retina !! yes we apply laser to the peripheral and by that the oxygen demand will decrease. The patient may develop VF defects but we prevent the ischemia and neovascularization and blindness from happening. neovessels (unhealthy) they bleed > fibrosis > blindness

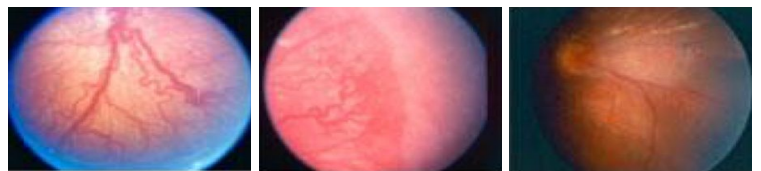
### Signs:

- Retinal Ischemia > no. of VEGF\* > neovascularization. ←IMP!! Pathophysiology
- Fibrous bands
- Retinal detachments
- Vitreous hemorrhage
- Leukocoria

When you catch it early-> laser cauterization(peripheral)-> stop formation of neovessels. (Central is preserved)



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



No developed blood vessels in this area -> ultimately leads to retinal detachment. Prophylactic is Laser

## 5. COLOBOMA

- Congenital condition caused by incomplete closure of the fetal fissure inferiorly. (no iris)
- Degree of visual loss related to area affected (iris, retina, choroid, optic nerve head)
- If it was in the optic disc or the macula there might be no light perception and the vision will be poor
- In embryogenesis the optic cup continues until it fuses inferiorly and the fissure closes.
- May be isolated disc or associated chorioretinal coloboma
- Usually sporadic, some Autosomal dominant
- Can be bilateral

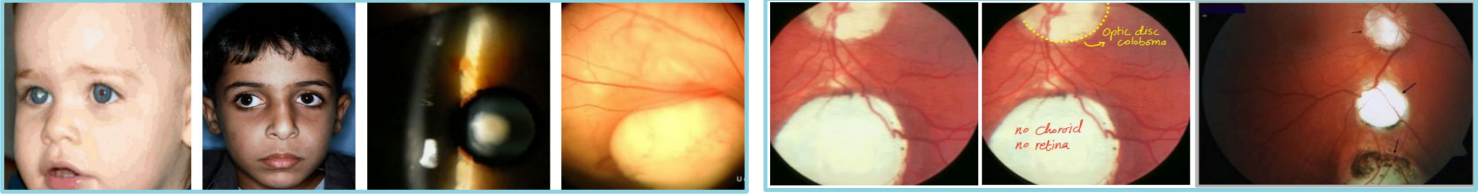


Leukocoria from scleral shining (Arrow) because of absence of retina.

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

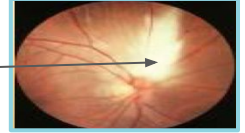


# Leukocoria



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

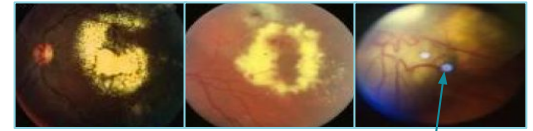
Myelinated (normally unmyelinated)



## 6. Medullated nerve fibers:

congenital anomaly caused by myelination of the retinal nerve fibers and usually asymptomatic. When large areas are involved it can cause leukocoria.

## 7. Coat's disease: (blood vessels disease)



typically a unilateral condition found in young boys. It is characterized by retinal telangiectasia and aneurysms that may cause exudative retinal detachments.

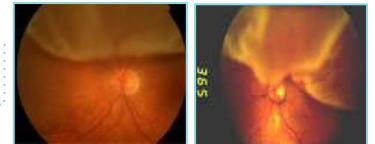
Tx: Laser therapy

Leaks = lipids

Go for leaky blood vessels and cauterize

## 8. Retinal detachment:

2 Layers separated



risk factors include trauma and surgery, vitreous detachment, high myopia, retinal breaks or tears, retinal vascular disease, and history of detachment in the other eye. symptoms include flashes of light, floaters, **curtain-like decrease in vision**

## 9. Organized vitreous hemorrhage

Happens when suction is used during giving birth



vitreous hemorrhage is usually secondary to a neovascular membrane or to a retinal tear. (trauma to the eye) Patients may complain of a red haze, blurred vision, or floaters. As it starts to resolve, color changes to yellow then white and some fibrous sheets may persist. A B-scan (ultrasonography) is usually diagnostic and vitrectomy is usually required.

### Summary of Leukocoria

- Leukocoria is white opacity of pupil.
- Can be caused by: Cataract, PHPV, ROP, Coat disease, Vitreous hemorrhage.
- Retinoblastoma has to be ruled out because it is life threatening disease.

# 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 imp systemic causes: HTN & DM

5



The child looks normal in this picture



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)



# Lecture Quiz

**Q1-6 years old patient presented to you in the clinic. His mother complaining of her child eye that is deviating inward. After completing history and physical examination, he was diagnosed with accommodative esotropia .What is your initial and best treatment plan?**

- A. Surgically correct the eye by medial rectus recession
- B. provide the child a convex lens to overcome hyperopia
- C. provide the child a concave lens to overcome myopia
- D. correct astigmatism with cylindrical lens

**Q2- which of the following causes of leukocoria should be ruled out first?**

- A. congenital cataract
- B. persistent hyperplastic primary vitreous
- C. prematurity of retinopathy
- D. Retinoblastoma

**Q3-After performing krimisky test, the following patient has 2 mm of eye deviation. How many diopter should be used in prism to overcome the misalignment?**

- A. 15d
- B. 30d
- C. 45d
- D. 60d



**Q4-which of the following is false regarding comitant strabismus?**

- A. common among pediatric patients
- B. the angle of deviation is not changing with different gazes
- C. nerve palsy is common to cause it
- D. congenital muscle diseases may cause it

**Q5-3 years old presented to you with his mother in the clinic. Complaining Of abnormality in her child eyes. After completing physical examination, the pediatrician noted anisometropia causing amblyopia “lazy eye”. What is your first treatment plan?**

- A. occlude the dominant eye
- B. treat the refractive error in the patient
- C. reassure and observation for 4 years then do the surgery
- D. Apply opaque adhesive patch in the skin

## Short Answer Questions

### Case 1



A: Name the condition shown in pictures above?

B: What is the cause for this condition?

C: What diagnostic test should be done?

### Case 2

**A 33 year old male complaining of gradual visual loss in peripheral field, difficulty in night vision, eye examination shown below:**



A: what is the abnormal finding in this patient?

B: Mention 2 common causes?

Answers:

Case 1

A: pseudostrabismus

B: flat nasal bridge and prominent epicanthal fold which tends to obscure the nasal portion of sclera

C: 1.cover-uncover test (only definitive test) the examiner finds that the patient manifests with no deviation 2.Hirschberg test (these tests used to differentiate pseudo from tru strabismus)

Case 2

A: White pupils (leukocoria)

B: 1.Retinoblastoma (most important deferential to rule out), 2. Congenital cataract

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