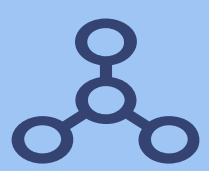


Strabismus, Amblyopia & Leukocoria

Objective

- not provided



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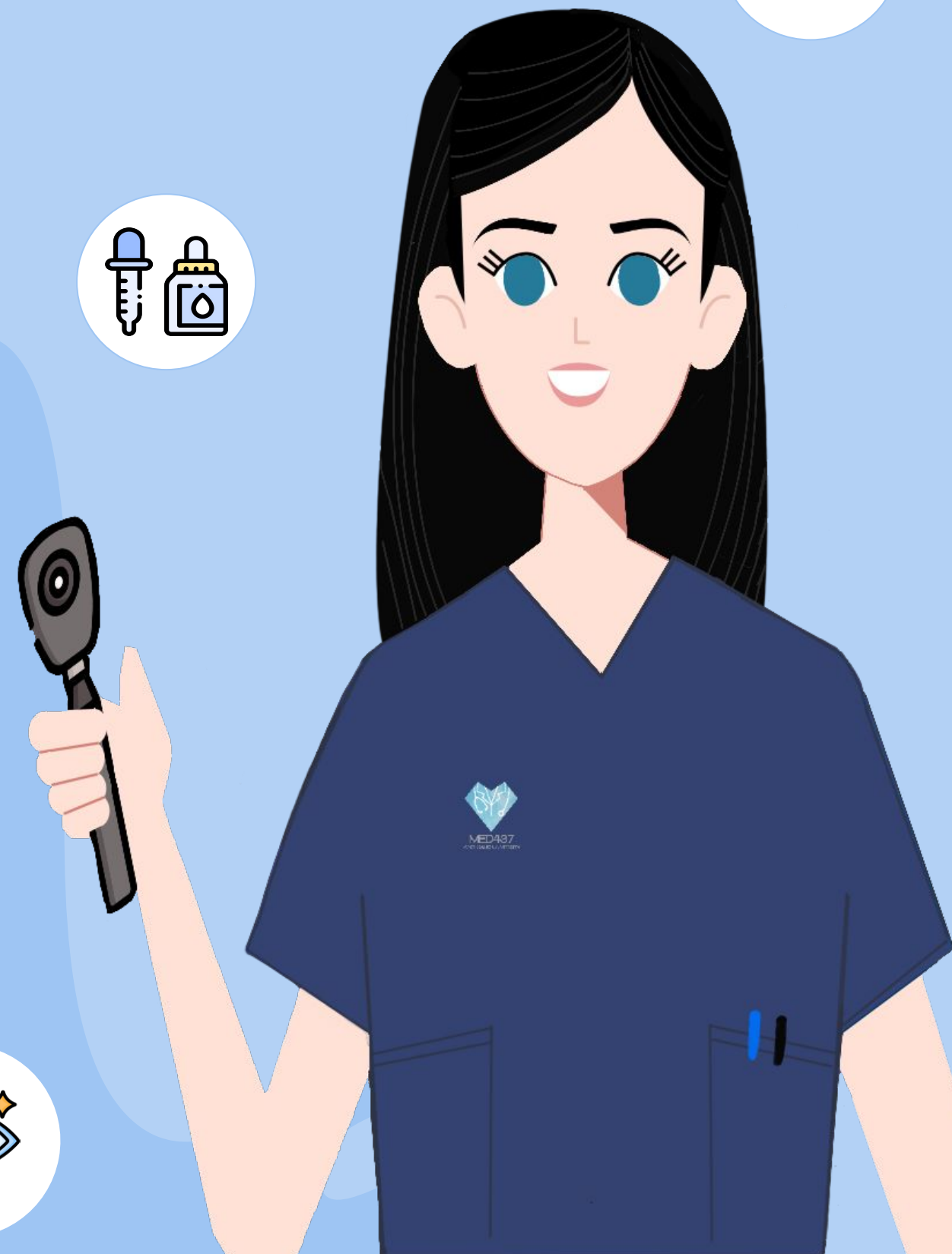
Resources :

Dr. Alotaibi slides and notes(F2), Dr. Adel
Alsuhaibani slides and notes(F1), 436 teamwork
Book (Lecture notes in Ophthalmology)

- Editing file

Color index

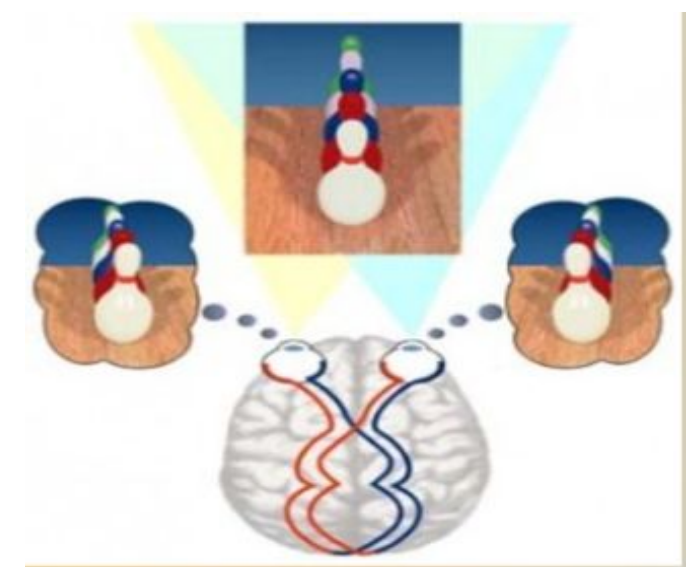
- **Important**
- **Notes**
- **Book**
- **Extra**
- **Golden note**



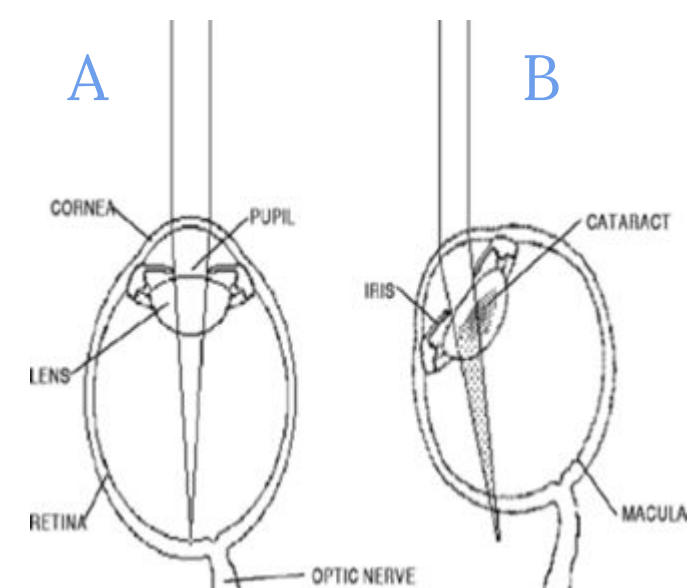
Strabismus

- ❖ Strabismus is a general term referring to ocular misalignment due to extraocular muscle imbalance (eyes are not aligned together).
- ❖ Strabismus occurs in approximately 3% of children and young adults. < 3 years
- ❖ 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.
- ❖ Why we are concerned about strabismus?
- ❖ It is important for functional and cosmetic reasons.
- ❖ Strabismus is associated with reduction of depth perception(binocular vision) and, if onset is in adulthood, double vision.
- ❖ Strabismus presents a cosmetic concern, especially for school-age children.
- ❖ When an individual's eyes are straight, they are said to have orthotropia. This indicates that both eyes are aimed at the same spot. The brain fuses the two separate images into one three-dimensional image.

- ❖ As we said before, when we use both eyes, we get 2 images; both images are shared by both eyes.
- ❖ Our retina is also divided into nasal and temporal parts; The temporal part will go to the same side of the brain, and the nasal will go to the opposite side. Then both images will go to the occipital cortex and get mixed there into a single image.



- ❖ 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".



Types of Strabismus:

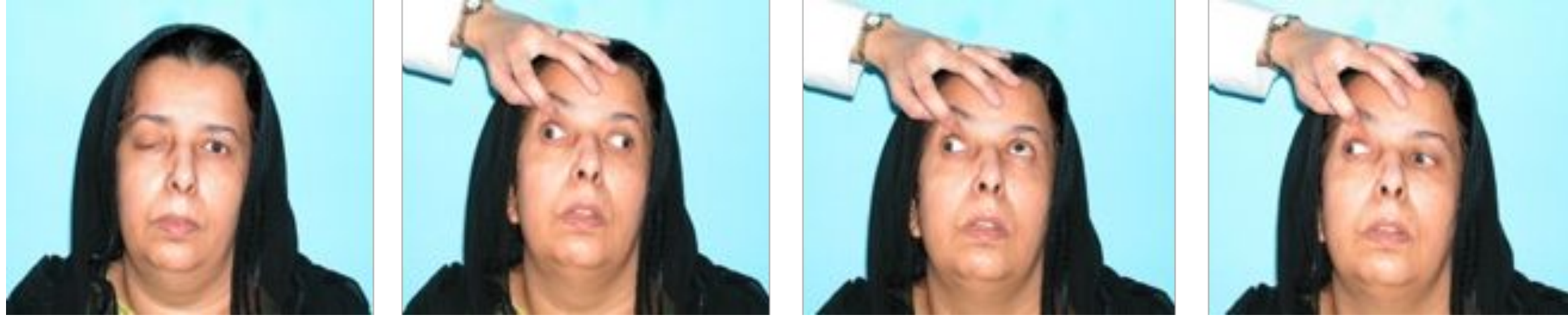
- ❖ **Comitant** → means the angle of deviation is the same wherever the patient look, and it's more common than non-comitant in children).
- ❖ **Non-comitant** → means there is deviation in certain gazes and is usually related to mechanical restriction (trauma) or neurological condition for example if the patient have right 6th nerve paralysis if he looks at the center he looks normal but if he looks at the right you will notice a deviation. Occurs in adults
- ❖ A.The eye is straight & the image is falling on the fovea.
- ❖ B.The eye is having inward deviation & the image is falling away from the fovea; therefore, the image will not be clear & the eye will be **amblyopic**



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.



- ❖ She has squint and ptosis in **the right eye**.
- ❖ In picture 2, she is looking to the left side, but medial rectus(3rd nerve palsy) of the right eye isn't working.
- ❖ In picture 3, she is looking up, but the right eye doesn't look up.
- ❖ But when she looks to the right in picture 3, she seems normal; thus, it is **non-comitant**.
- ❖ She has 3rd nerve palsy in the right eye (**the 3rd nerve supplies all muscles except lateral rectus (CN VI) and superior oblique (CN IV)).**
- ❖ This is neurological, she might've had stroke (this doesn't occur in children).



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



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



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

- ❖ Cause of strabismus? (in children)
 - Unknown. However, strabismus is certainly more common in families with a history of the disorder.
 - The **great majority of children** who present with strabismus have no other associated neurological abnormalities.
 - 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.
- ❖ Types of **comitant strabismus**:
 - Esotropia. More common, why? Because pediatric normally have hyperopia and with stress, fever, trauma and other risk factors the eye decompensated.

خلاص ما تتحمل اكثر من كذا فيصير لها انحراف، يقولون لك اهل الطفل: كان طبيعي الولد فجأة لفت عينه على جوا

- Exotropia.

Esotropia is divided into: (esotropia = means the eyes are deviated **toward** the nose)

- Infantile esotropia (develops during the first 6 months of life, not born with it. If born with it, we call it congenital).
- Acquired esotropia.

Infantile esotropia: The cause of this esotropia in children because medial rectus is pulling too much

- Infantile esotropia is the inward deviation of the eyes noted before the patient reaches **age 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.
- Infantile esotropia is not believed to be connatal but rather develops in the first few weeks or months after birth.
- Classic infantile esotropia is constant and involves a large angle of deviation exceeding 20 prism diopters (PD) on corneal light reflex measurement

Infantile esotropia (comitant)



white dot here means = corneal reflex by a torch light not ophthalmoscope and notice that the corneal reflex deviated temporally this mean the eye going inward nasally which is esotropia and we called this (corneal reflex or Hirschberg test).



Alternating esotropia
So this child will not have amblyopia

Left esotropia
Left eye will become amblyopic



Right esotropia

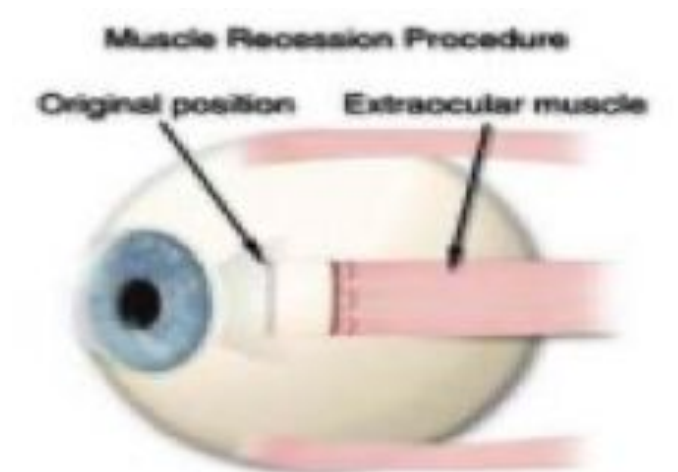
-Infantile esotropia (Right eye will become amblyopic).
- If left untreated → the brain will suppress the eye = amblyopia



-First day post-op - the surgery's purpose → 1) weakens both medial rectus 2) strengthen the right lateral rectus

Management of infantile esotropia:

- For infantile esotropia, 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.



Muscle recession procedure

-what will happen for this child if we don't treat him for long time? amblyopia
- Why the child will develop amblyopia? because the left (normal eye) will send the clear image to the fovea but the right eye with(esotropia) the image falls away from the fovea so will send blurred image to the brain, so these two images will go to the brain and the brain can't make them one as one image so here the brain get confused because physiologically each eye send an image to the brain and make them single image so in this child the brain will suppress the blurred image called suppression ,in response to that the eye become lazy and not working anymore. Also visual maturation will stop so we end up having amblyopia.
- if amblyopia isn't treated during the **first 5 years of age** it usually very hard to reverse it
-the fovea is the most sensitive area in the macula, and we see by our fovea very clear and sharp due to the high cones density in the fovea.

Prognosis after surgery for Inf ET?

- Better ocular alignment and visual prognosis can be achieved if surgical correction is performed before age 2 years.
- Long-term follow-up studies on esotropic infants by age 2 years have shown that close to 60% achieve a small angle (10PD) cosmetically acceptable strabismus.
- Although some binocular vision is achieved, it generally is subnormal, often involving peripheral fusion. **Brain will suppress the weak eye to obtain better vision**
- 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.

- **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 u remove the cover can he maintained his eye in the center)”.
- Check the extraocular movement: to rule out paralytic 6th nerve palsy, how to check? spinning راسه ندور 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 ≥ 1 yo).
- Check for the associations that we just mentioned above: cross fixation, DVD, IOOA, and latent nystagmus.
Fundus exam to check the optic nerve and retina.

- **Tests for deviation (how to test strabismus)**

❖ **Hirschberg test (Corneal light reflex)** 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 احفظوها بالنقطه مهمه.

❖ **Note: roughly if corneal reflex is:**

❖ At the pupil edge = 30 PD (15o)

❖ Midway between pupil and limbus = 60 PD (30o)

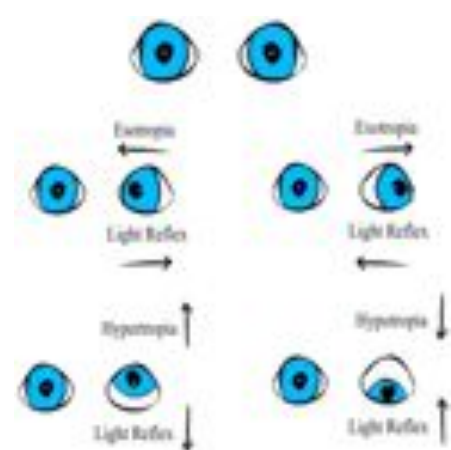
❖ At the limbus = 90 PD (45o)

❖ **When the corneal reflex is:**

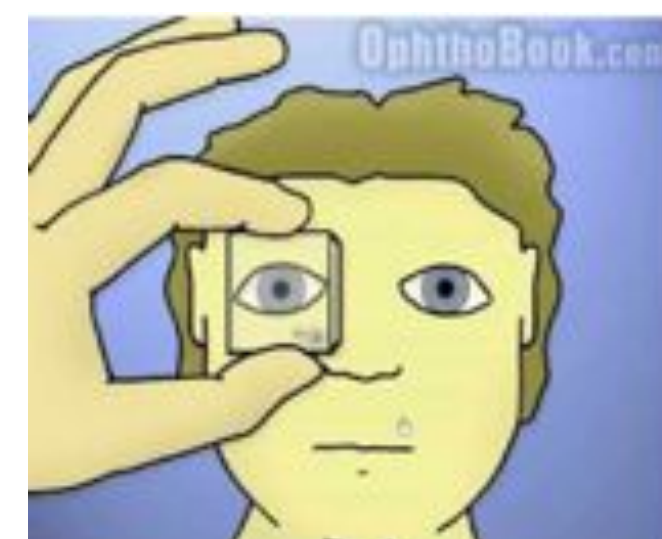
❖ In the center, we call it orthoptic and that's the normal case.

❖ Light is shifted temporally or laterally → esotropia (inward deviation).

❖ Light shifted nasally → exotropia (outward deviation).



Krimsky test



Name of the instrument: prism.
What is it used for: testing the deviation (strabismus).

❖ **Krimsky test**

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

■

❖ **Cover test** (most important test) Watch this video and you'll understand everything!

❖ There are 2 types of cover tests:

❖ **Unilateral cover test** (cover-uncover test): performed by having the patient focus on an object then covering the fixating eye and observing the movement of the other eye. If the eye was exotropic, covering the fixating eye will cause an inwards movement; and esotropic if covering the fixating eye will cause an outwards movement. It is used to **detect tropias**.

❖ **Alternating cover test** (cross-cover test): performed by moving the occluder from one eye to the other eye. Normally, the covered eye shouldn't move when the occluder is removed. It is the most accurate way to pick up subtle **phorias and tropias** since it breaks binocular vision.

❖ **IMPORTANT** (Always comes in exams).

➤ Cover-uncover test → detect tropias.

➤ Cross-cover test → detect phorias **and** tropias.

Pseudostrabismus

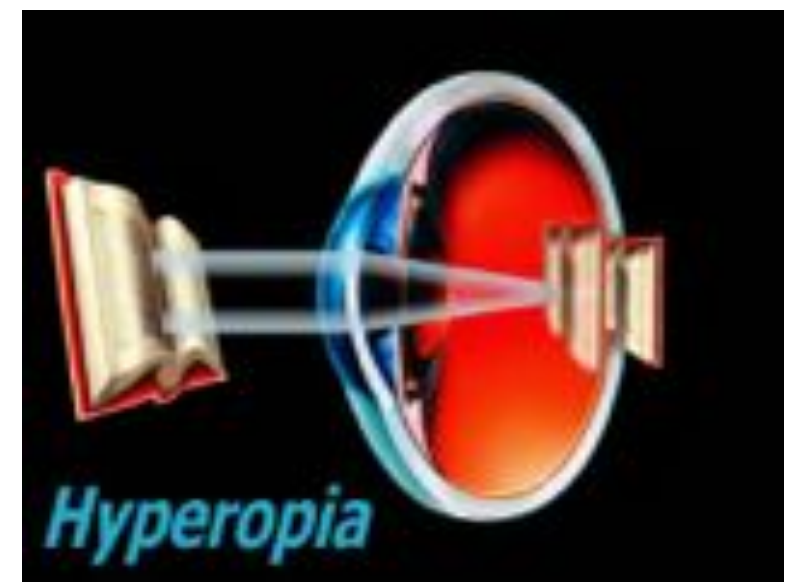
- ❖ Pseudoesotropia is a condition in which alignment of the eyes is straight (also known as orthotropic); however, they appear to be crossed.
 - ❖ This condition most commonly occurs in infants when a flat nasal bridge and prominent epicanthal folds tend to obscure the nasal portion of the sclera. **Skin covers the sclera**
 - ❖ A careful ocular examination (eg, pupillary light reflex) reveals that the eyes are straight.
 - ❖ Using the **cover-uncover test (only definitive test)**, the examiner finds that the patient manifests no deviation (**normal exam; because they are central**).
 - ❖ When to operate?
 - ❖ Hirschberg \ cover test differentiate it from true esotropia
 - Gradually disappear with age full examination & reassure
- Prognosis: It is accepted that better ocular alignment and visual prognosis can be achieved if surgical correction is performed before age 2 years.



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.

Accommodation

- ❖ Rays of light from the object diverge, these light rays are then converged by the crystalline lens in an attempt to focus them on the retina. For this to happen, the lens will accommodate, (become thicker centrally and optically more powerful) causing further bending of the rays of light until they focus on the retina. **We give convex glasses. We need more power.**
- ❖ Relationship of hyperopia to Acc Esotropia
- ❖ **Near reflex:** accommodation, convergence and miosis (involuntary).
- ❖ **How does accommodation occur at the level of ciliary body?** contraction of ciliary body, relaxation of the zonules and the lens becomes thicker providing more power (the main reason to accommodate is to get extra power of the lens).
- ❖ In children, if they have **hyperopia** (image behind the retina), they need to **accommodate excessively** to increase the lens power in order to make things clear, so the **convergence will be more** leading to esotropia due to muscle stretching (accommodative esotropia).
- ❖ Accommodation decreases with age that's why adults with hyperopia doesn't develop esotropia (newborns have the max accommodation).



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

Accommodative esotropia:

- ❖ This condition usually presents in patients aged 2-3 years (acquired after 6 months).
- ❖ Associated with hyperopia.
- ❖ Refractive error correction by glasses (convex) will treat the condition.
- ❖ Perform cycloplegic refraction on all children by using the retinoscope and loose lenses. **+ give glasses**
- ❖ Cycloplegia is the achieved with Mydriacyl 1% and cyclogel 1%. **No need to know.**

❖ **Clinical features of Acc ET:**

- o Refractive error usually +3_+4. **hyper**
- o May precipitated by acute illness or trauma.
- o Start intermittent and if not treated become constant.

❖ Do we do surgery for Acc ET?

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

❖ The angle of deviation is less the infantile esotropia.

❖ Treatment of high AC/A?

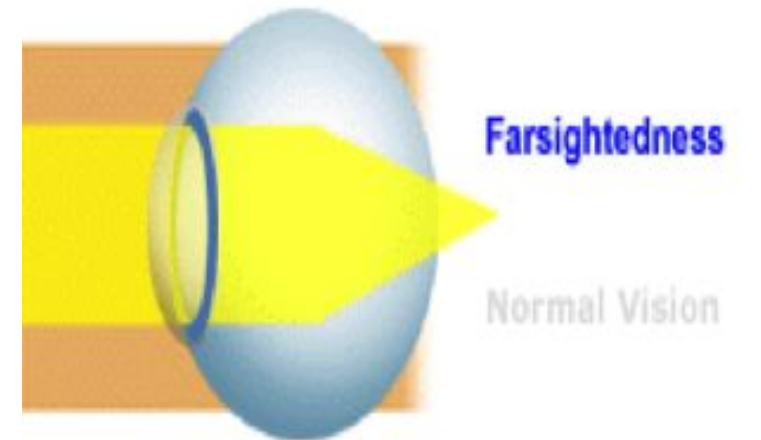
❖ With patients aged 4-5 years, one can attempt to reduce the strength of the hyperopic correction to enhance fusional divergence and to maximize visual acuity (**convex lens to relax the accommodation > back to normal**).

➤ When to stop glasses?

❖ Some children will no longer need their glasses at an earlier age while others will need the proper farsighted glasses or contact lenses to control the esotropia even as adults (**50% at 6-7 years of age they get rid of glasses**).

❖ Not every child with hyperopia will develop accommodative esotropia.

- ❖ أكثر من زيارة يعني في الزيارة الأولى نعطي المريض full cycloplegic بعد شهر - شهرين تكون الزيارة الثانية ونشوف تحسن ٨٠٪ للمريض، نكمل نعطيه cycloplegic.
- ❖ في الزيارة الثالثة بعد ٦ شهور بتكون عين المريض بإذن الله مستقيمة.



❖ When to stop glasses?

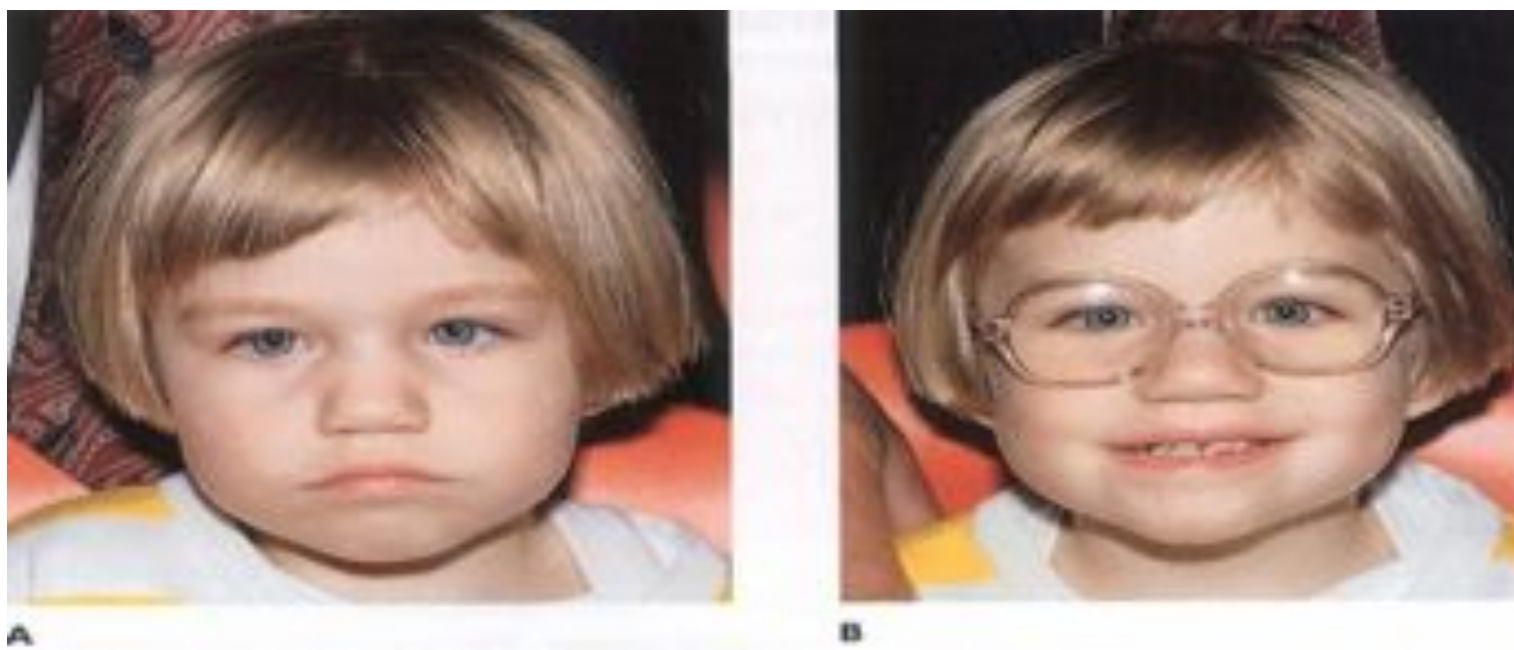
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* Wearing glasses for pediatric is MANDATORY, why? To prevent amblyopia * Adult = optional

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



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

Exodeviation

- ❖ Exodeviation is a horizontal form of strabismus characterized by visual axes that form a divergent angle. Deviated to the temporal part (outside). No need to know the other details about it,

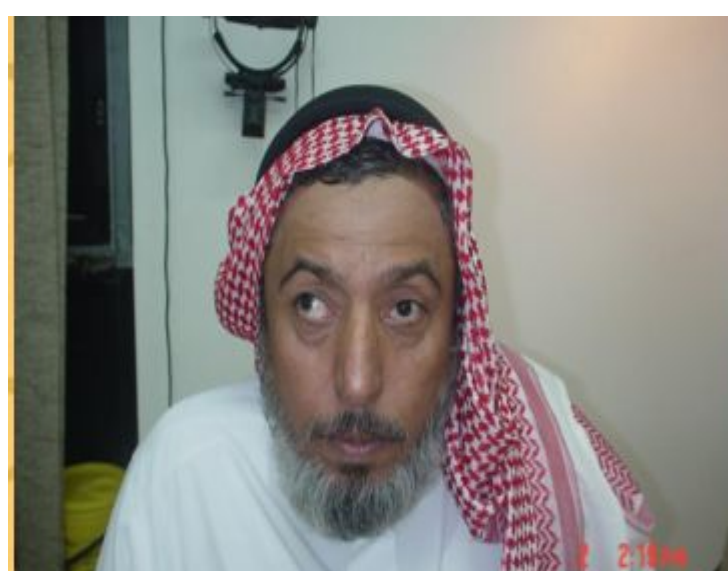
Exotropia (less risk of amblyopia)

- ❖ Esodeviations are more frequent than exodeviations, with a ratio of 3:1.
- ❖ Acquired exotropia is more common in the Middle East, Africa, and Asia and in those latitudes with higher levels of sunlight. It is less common in the United States and Europe.
- ❖ The deviation usually begins as an exophoria.
- ❖ This deviation may later progress to intermittent exotropia.
- ❖ When intermittent exotropia develops in a child whose visual system is still immature, bitemporal suppression develops, and the child does not perceive 2 separate images (diplopia).
- ❖ When intermittent exotropia develops in a child whose visual system is still immature, bitemporal suppression develops, and the child does not perceive 2 separate images (diplopia).
- ❖ As suppression increases, intermittent exotropia may finally progress to constant exotropia.
- ❖ Intermittent exotropia can have an early onset, with 25-40% of cases occurring before the second year of life.

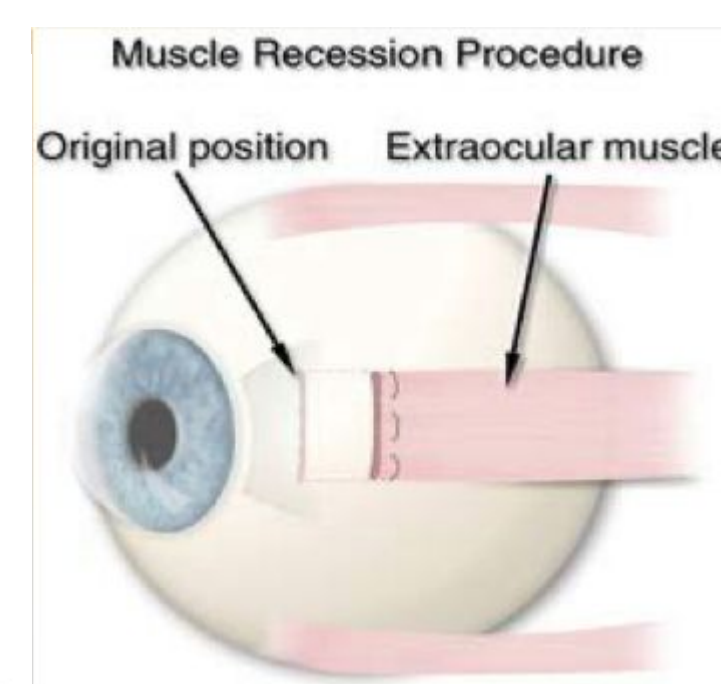
- Basic type of Exotropia

1- Dsc = 55 xt

2-Nsc = 55xt



This deviation may later progress to constant exotropia

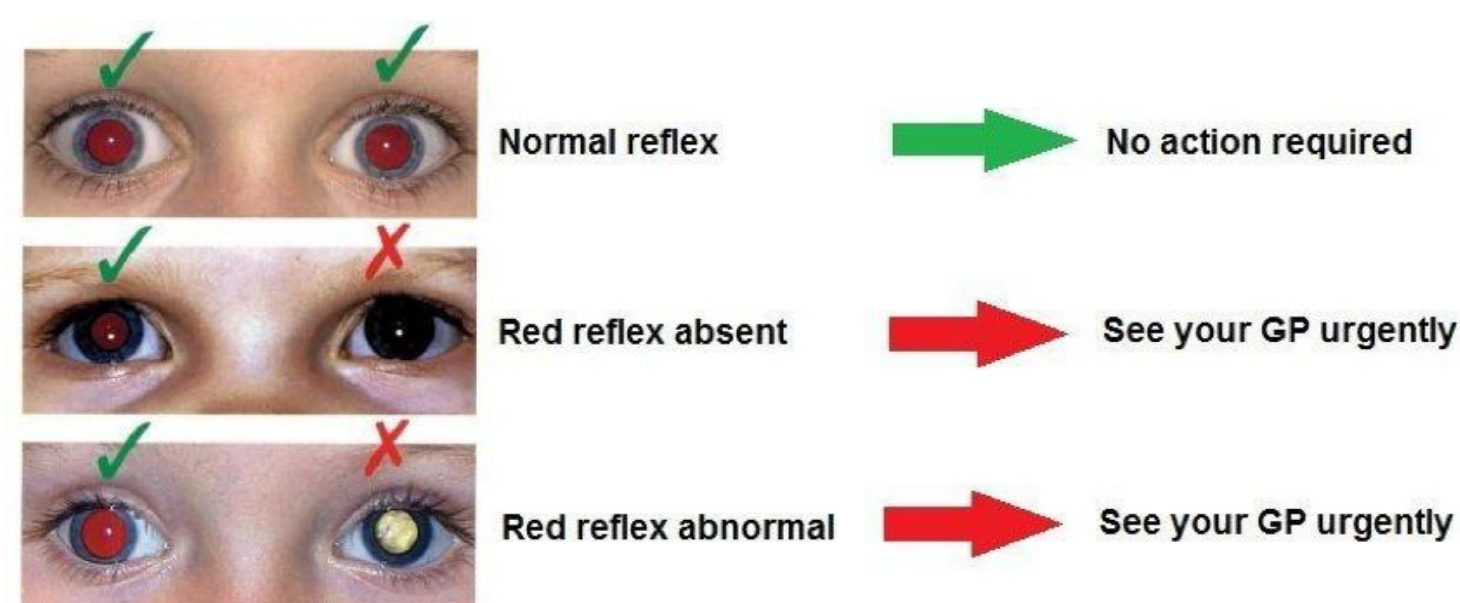
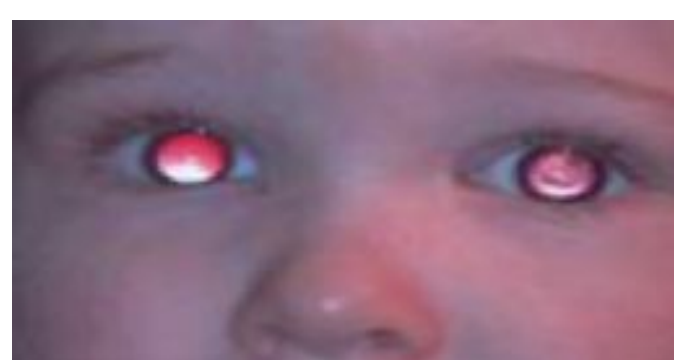


Leukocoria

- **Definition: white opacity of the pupil.**
- ❖ The color of the normal red reflex comes from the retinal pigment epithelium.
- ❖ How to diagnose the leukocoria? **direct** ophthalmoscope, shine light at pupils from 1 meter (you must get an equal red reflex)
- ❖ **Definitive diagnosis:** B scan ultrasound.



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



Normal red reflex

why we have red reflex? because of retinal pigmented epithelium it's immediately behind the retina as we shine the light the neurosensory retina usually is transparent also the vitreous and cornea are transparent. (from cornea until the retinal all transparent). So, when we shine the light, we get this reflex from pigmented epithelium

The pathology here can be anywhere: cornea, anterior chamber, lens, vitreous or retina.

Leukocoria

White opacity of the pupil. before elaborating in the causes remember that anything that oppose the transparency of (lens, vitreous and cornea until the retinal) could cause of leukocoria.

Causes:

Cataract (opacity of the lens).



It is **the most common cause of leukocoria** in children.

It can be congenital or acquired, usually causes blurred vision and glare.

- The most common causes of **acquired** cataract are TORCH & trauma.
- The diagnosis of **congenital** cataract is made by a direct **ophthalmoscope**.
- Treatment of congenital cataract must be performed within the first few weeks of life and be accompanied by a coordinated patching routine to the fellow eye to stimulate visual maturation in the amblyopic eye and minimize the risk of squint.

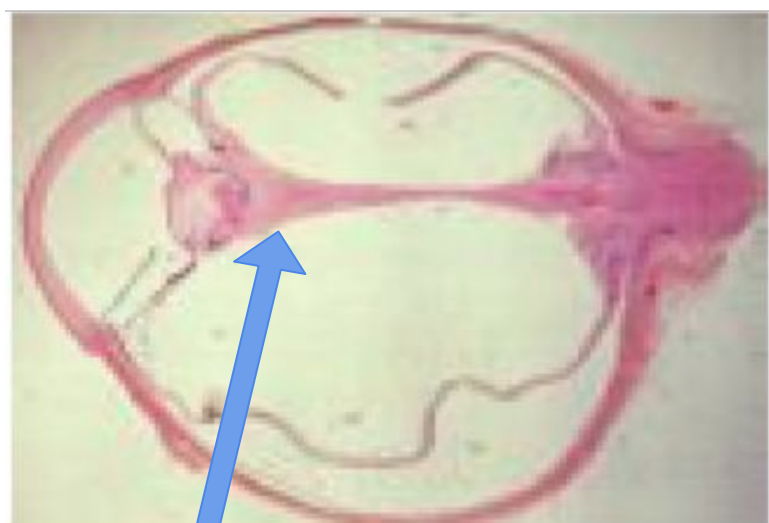
The cataract is worse in children and more dangerous than adults because the visual system mature during the first one year of life and the first 3 months of life are very sensitive if anything happen during this period the visual maturation will get affected and the patient will have amblyopia (lazy eye) and leads to blindness if not treated early. **That's why it's crucial if we treat the child in the 1st and 2nd month of life**, and if we leave this child to one year of age it's considered too late and the damage is irreversible. Very very imp to use the ophthalmoscope.

Persistent hyperplastic primary vitreous (PHPV) (also called persistent vascular fetus disorder)

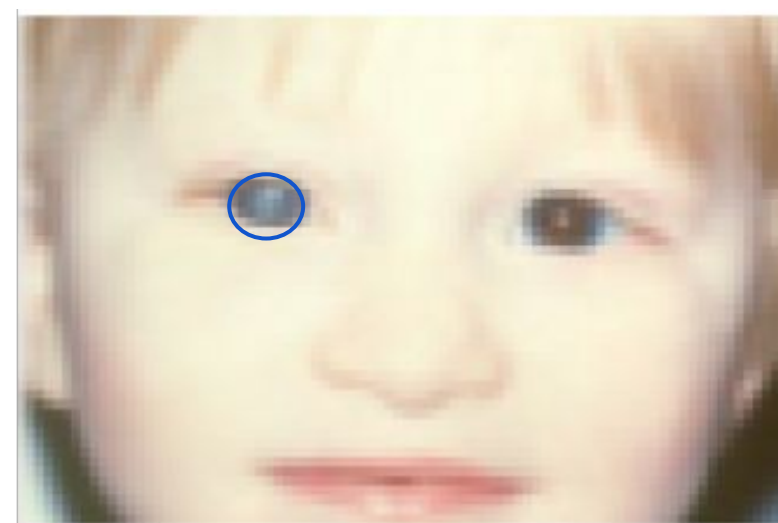
It is a congenital condition caused by a failure of the normal regression of the primary vitreous.

It is usually associated with **unilateral vision loss**.

- Caused by failure development of the eye during growth of embryo, the blood vessel will come out of the optic disc and nourish the development lens and this vessel **should be regress before birth**, if this physiological process fail and the blood vessel remain and attached to the lens the child will born with **persistent hyperplastic primary vitreous** leads to opacity of the lens and small eye (eye didn't devolve properly). **should treated in early months**. Otherwise he will develop **amblyopia**.



This should disappear before birth

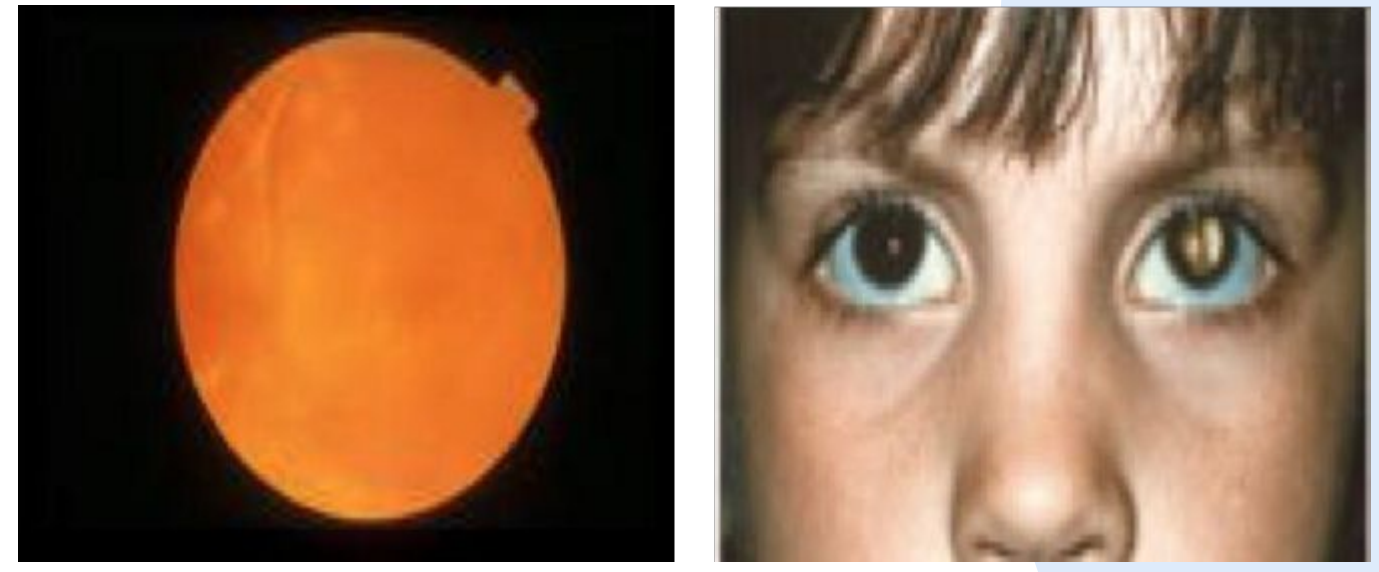


Opacity of the lens (circle). And smaller.

Needs to be dealt with or will develop lazy eye

Leukocoria Causes

Organized vitreous hemorrhage

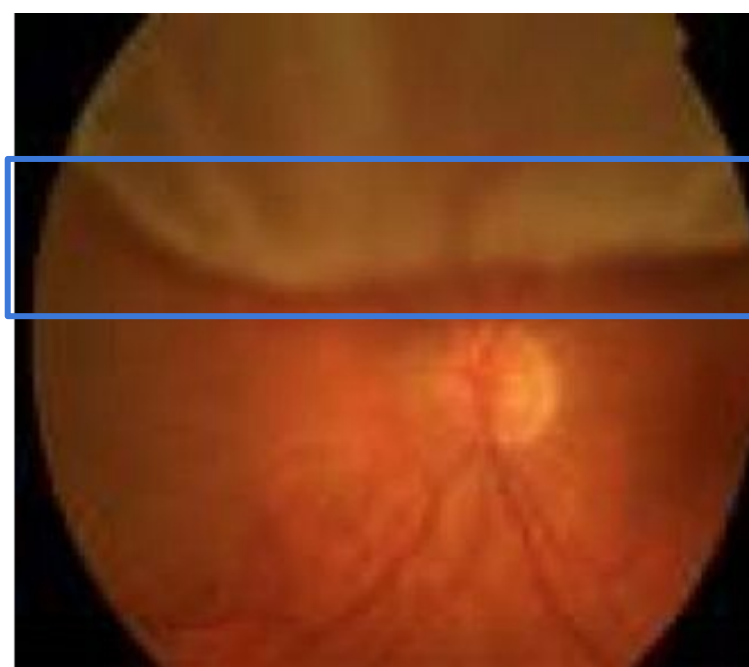
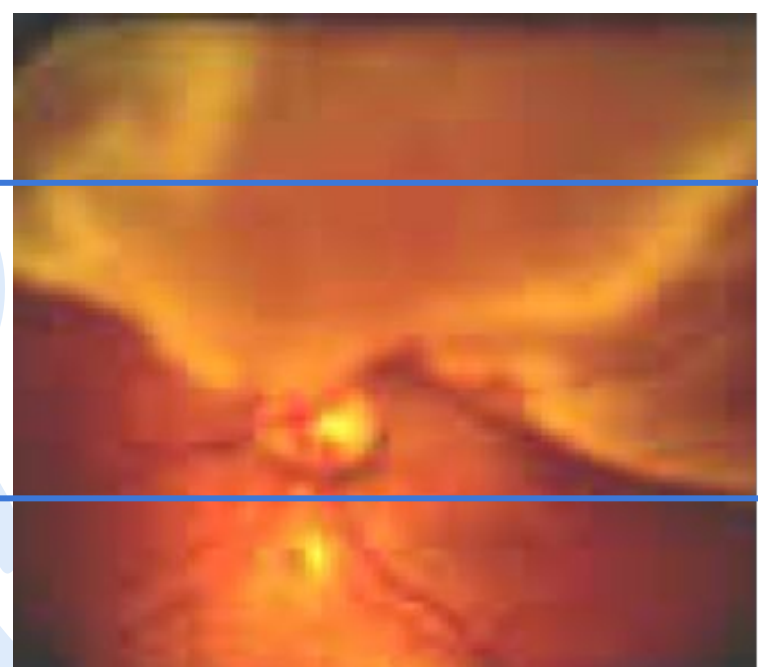


- A vitreous hemorrhage is usually secondary to a neovascular membrane or to a retinal tear.
If a child got trauma, there will be blood in the vitreous, and if the blood gets organized, this will prevent you from seeing transparent vitreous.
The most common cause of vitreous hemorrhage in babies is birth trauma(forceful birth(operative))
- Patients may complain of:
 - A red haze.
 - Blurred vision.
 - Floaters.
- As it starts to resolve, color changes to yellow then white and some fibrous sheets may persist.
- **A B-scan (ultrasound) is usually diagnostic**, and vitrectomy is usually required. [Send for US +make sure no retinal detachment](#)

Retinal detachment:

- Risk factors include:
 - Trauma ([mainly](#)) and surgery.
 - Vitreous detachment.
 - High myopia.
 - Retinal breaks or tears.
 - Retinal vascular disease.
 - History of detachment in the other eye.
- Symptoms include:
 - Flashes of light.
 - Floaters.
 - Curtain-like decrease in vision.

Retinal detachment is basically a separation of the neurosensory retina from the retinal pigment epithelium, so when this happens there will be no color behind the neurosensory retina resulting in a white appearance of the detached retina (curtain-like); But, you have to have a big retinal detachment to see leukocoria when you shine the light.

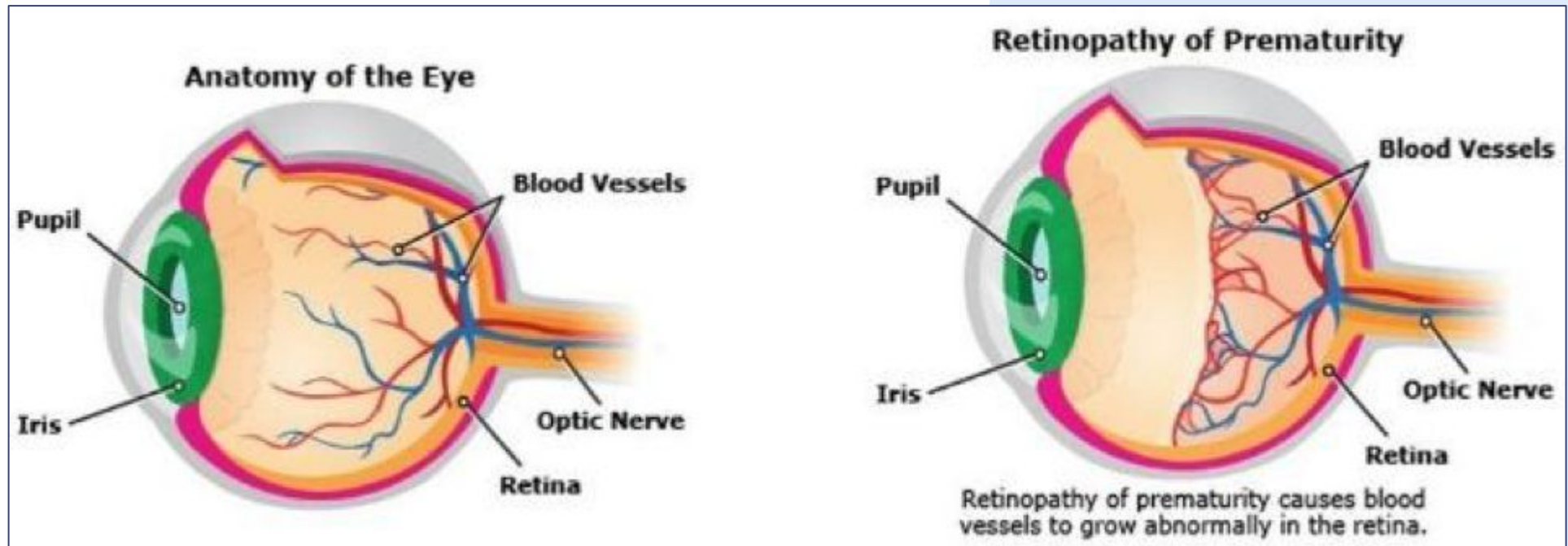


Abnormality:
leukocoria
2 causes: cataract & retinoblastoma

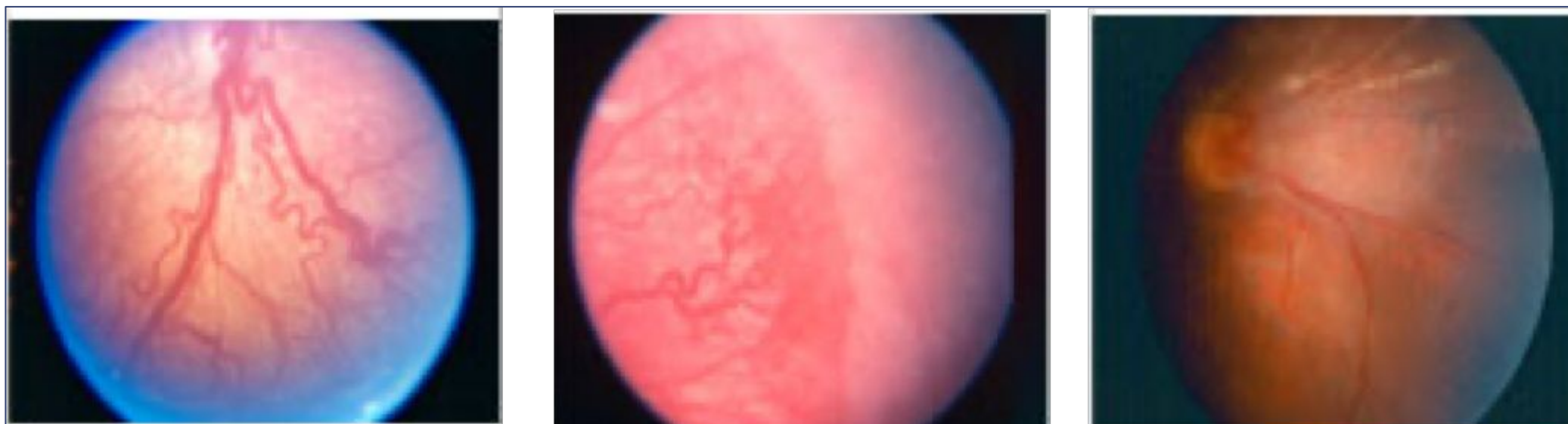


Leukocoria causes cont.

Retinopathy of prematurity (ROP):

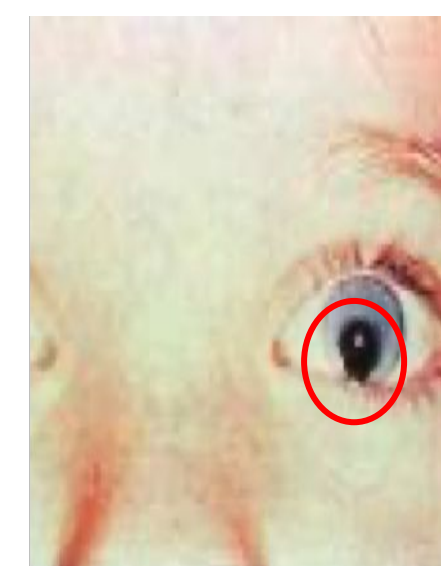


- It occurs in premature, low-birth-weight infants maintained on oxygen therapy.
 - Signs include:
 - **Neovascularization.**
 - Fibrous bands.
 - Retinal detachments.
 - Vitreous hemorrhage.
 - When advanced leukocoria can be present.
- During the embryology life, blood vessels will come from optic nerve and spread to the periphery of the retina to supply it.; This process doesn't stop before 37 weeks GA.
- 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**
- If you go to a blind school, you will find a good number of children with ROP.
- **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).
 - According to the book (any child born before 32 weeks of gestation should be screened for ROP).
- 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).**



Retinal fibrosis pulling to the side as a complication of ROP

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

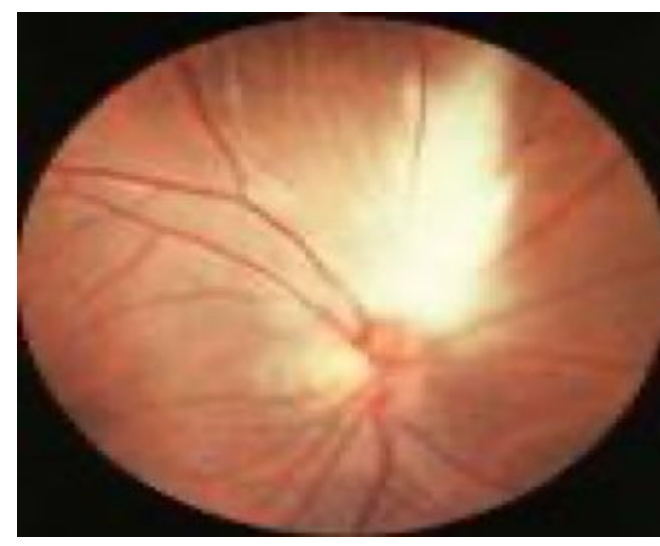


will result in "keyhole" or "cat eye" appearance. (Tissue is not formed)

Coloboma (occurs in uterus):

- A **congenital condition** caused by incomplete closure of the fetal fissure.
- Degree of visual loss to area affected (iris, retina, choroid, or optic nerve head).
Coloboma can affect the iris as well and will result in "keyhole" or "cat eye" appearance, notice that the iris isn't complete here there is still opening. It is an eye defect. Deficient structure of the eye Fetal fissure will give rise to the iris and retina.
Does coloboma causes problems? It depends if the macula and fovea are involved this will result in poor vision if it happens away from the macula and fovea the effect will be less.

Leukocoria causes cont.

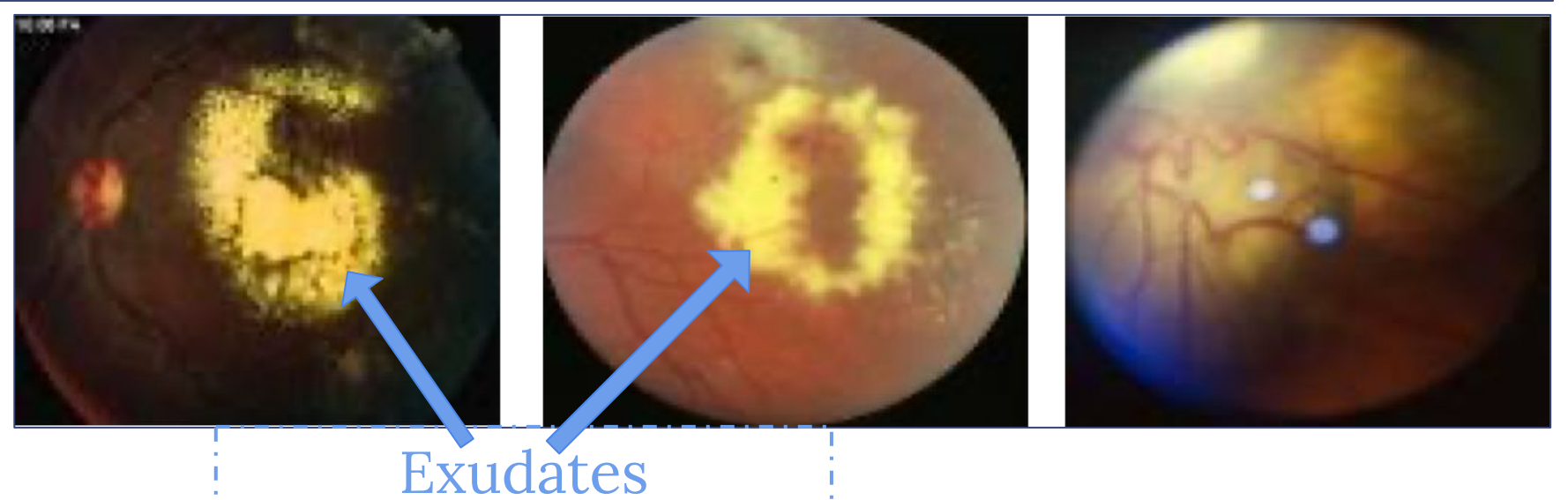


In the retina, the nerve fibers are unmyelinated to allow the passage of light “Cat fur appearance”

Medullated nerve fibers (occurs in uterus):

- **Congenital anomaly** caused by myelination of the retinal nerve fibers and usually asymptomatic **unless** the macula is involved.
- Involvement of the macula will cause decrease in vision because 95% of our visual acuity comes from the fovea.
- When large areas are involved, it can cause leukocoria.
- Normally, we have **non-myelinated axons** going to ON.
- We have myelination in our bodies for insulation, if there's myelin in the retina it will cover the macula and disrupt the vision.
- Myelin > Insulation > Fast conduction > white color

Coat's disease (congenital condition):



- Typically, a **unilateral** condition found in young boys.
- It is characterized by retinal telangiectasia and aneurysms that may cause exudative retinal detachment.
- Their blood vessels leak **exudates (lipids)** and will deposit.
- Quicker than BM and more profound
- It won't affect the vision unless involving the macula.
- The diagnosis is not by examining the retina only we inject the fluorescein stain.
- Rx: cauterization of leaking vessels by laser (like DM)

Retinoblastoma (RB):



- The most important cause of leukocoria because it doesn't affect the vision only, but it can be a life-threatening condition since it's malignant.
- Most common primary, **malignant, intraocular tumor** of childhood but still a rare tumor.
- Vast majority become apparent **before age of 3 yrs (2-4 y)**.
- It results from **malignant transformation of primitive retinal cells** before final differentiation.
- **Presentation is** most commonly (60%) with **leukocoria** and strabismus.
- **If you see leukocoria, always think of RB until proven otherwise.**
- **Retinoblastoma has to be ruled out because it is a life threatening disease**
- It is a lethal condition because it can **extend to optic nerve & reach the brain**.
- Rx: radiation, enucleation (removal of the eye as well as ON) or chemotherapy (the earlier the better).
- Think about the life of the patient, vision is not a priority in RB.
- We monitor the patient for 20 years, and we screen other children in the family.

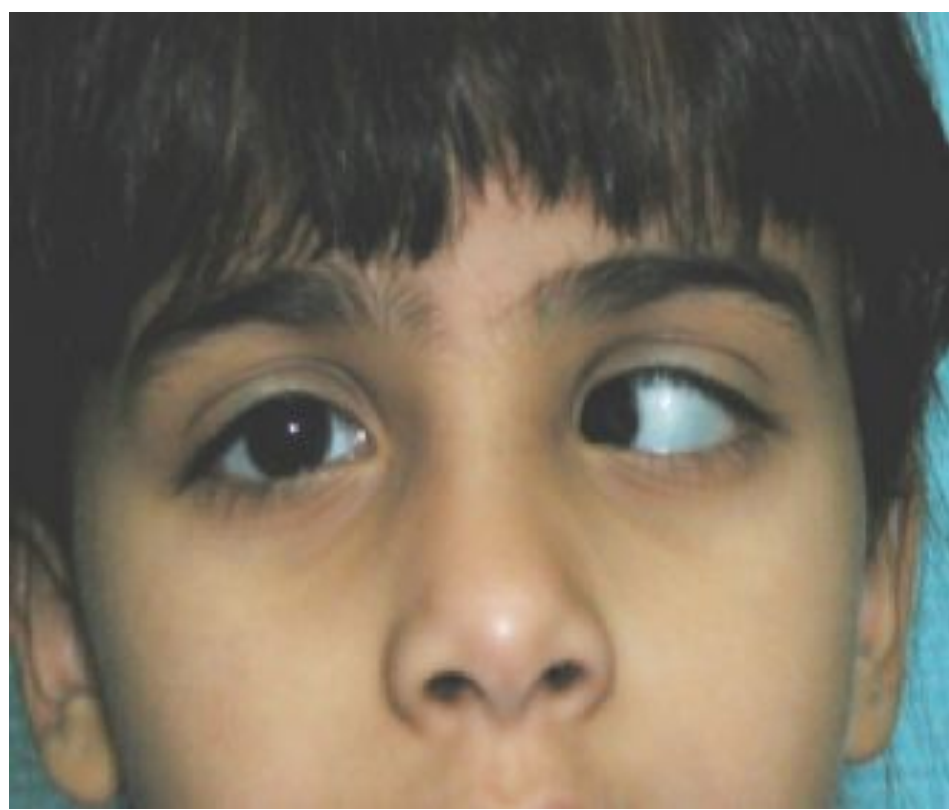
AMBLYOPIA (lazy eye)

- ❖ **Definition:** **reduction in visual acuity in one or both eyes**; without the presence of an organic cause that explains that decrease in visual acuity.
- ❖ This reduction cannot be completely accounted for by any clinically apparent organic abnormality.
- ❖ Cortical ignorance of one eye. .
- ❖ Always occurring before the age of 7 years.
- ❖ Prevalence of 2% to 4% in the general population.
- ❖ It is the single most common form of monocular vision impairment in the first 4 decades of life.

- Remember the eye is growing & will not get mature immediately after birth.
- The development of the visual acuity takes time (when a child is born the vision is around 20/200 and it improves over years).
- Amblyopia means that you have one functioning eye, you can't be a pilot, a surgeon or an ophthalmologist. The only way to prevent it is by early detection.

❖ Classification:

- According to the condition that induced it (**cause of amblyopia**).
- Strabismus is the most common amblyopia-inducing condition.
- 40% of children with a manifest strabismus develop amblyopia.
- Esodeviations are more common than exodeviations.
- **Strabismus will never resolve** by itself with growing, and it will develop to amblyopia.



The child is having strabismus (esotropia) & the left eye is the weak one; because the clarity of the image sent from the left eye is bad (the eye is already rotated & image isn't falling on fovea). While the image sent from the right eye is falling on the fovea & will be sharply clear. When the brain receives 2 images of different clarity, it will choose the clear one & neglect the other; resulting in amblyopia of the neglected eye.

❖ What if images were totally different or grossly different ?

1. **Diplopia.**
2. Then the brain will get **Confusion.**
3. The brain is smart, it will resolve the problem by **Suppression** (the end result of the received 2 images)> it will suppress the blurred image(the abnormal eye) > lead to amblyopia.

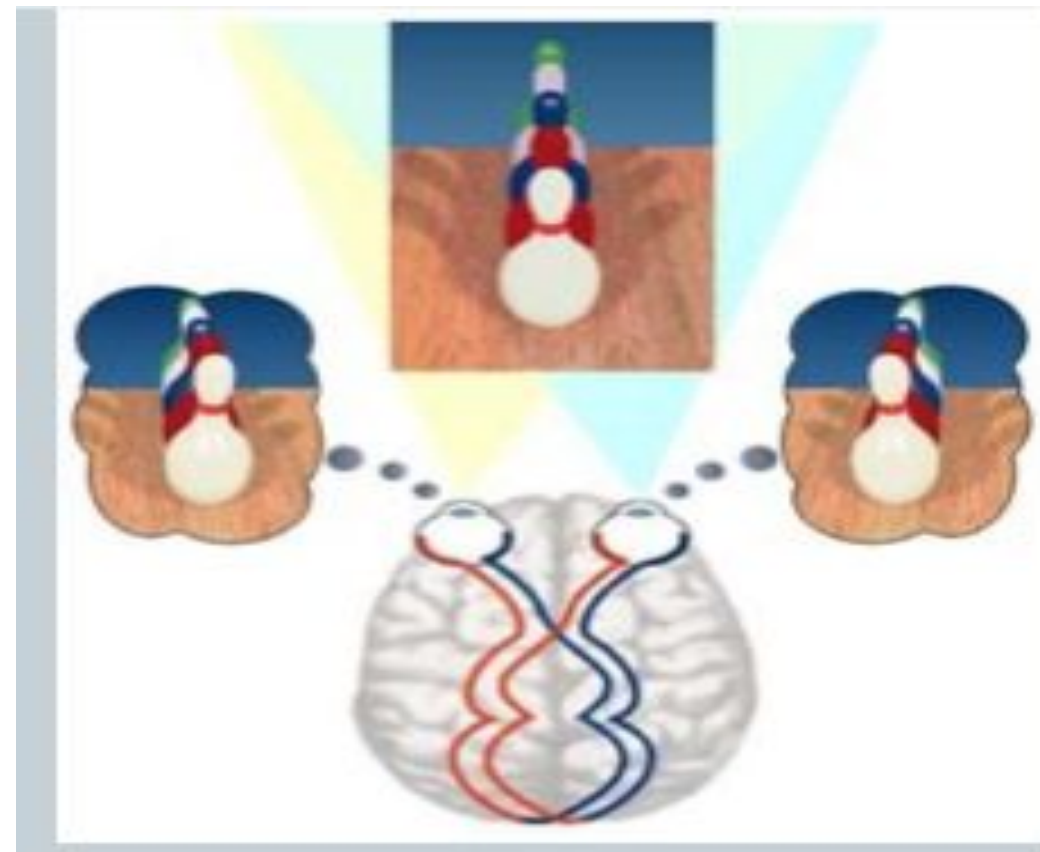
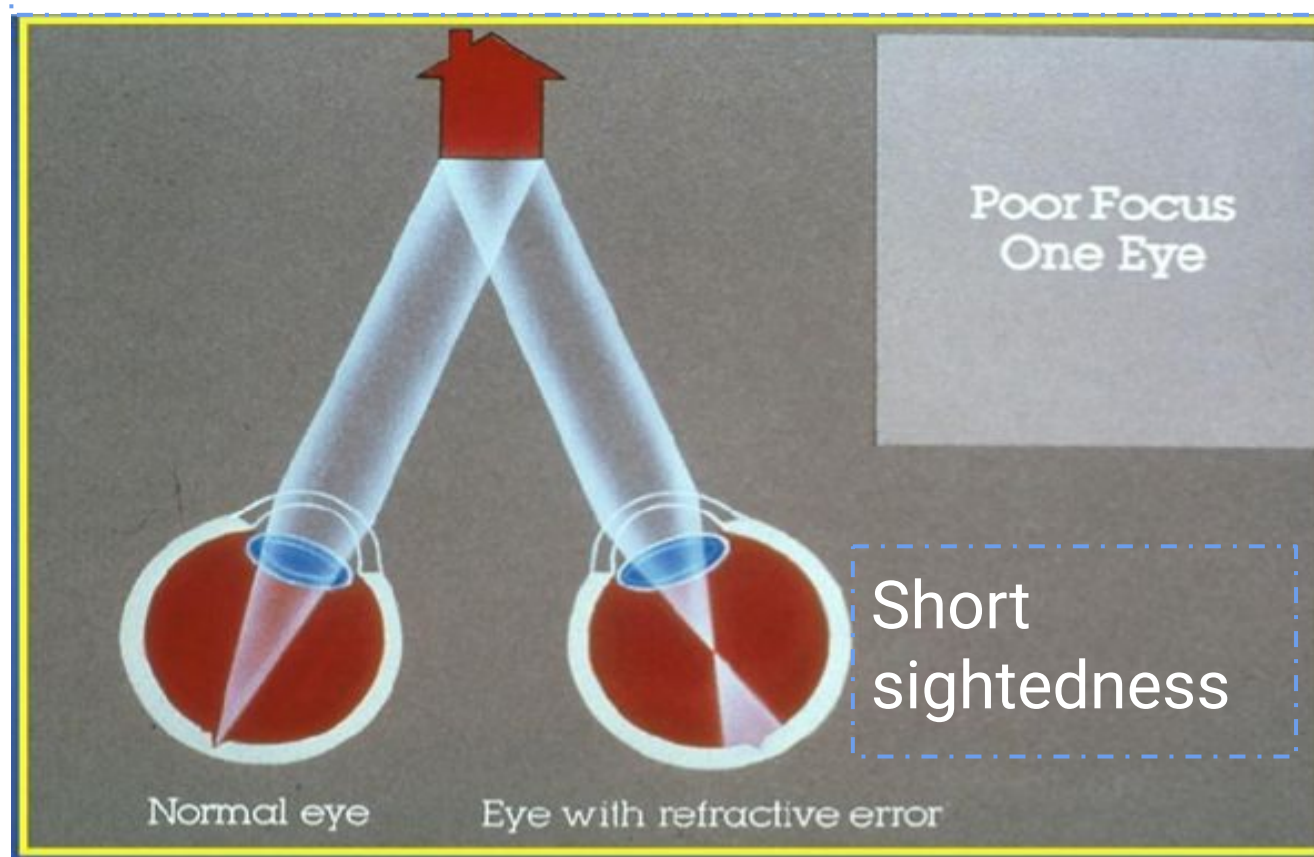
❖ Why fusion may fail ?

1. significant anisometropia The refractive error of both eyes are not the same.
2. significant aniseikonia (difference in image size)
3. Strabismus. Straight eye will send the good image, the eye with esotropia will send blurred image.
4. Higher cortical problems (e.g: head trauma, alcohol intake , drugs , ... etc).

No need to know this just know that not any difference we call it anisometropia and it is more common with hyperopia.

- ❖ Mild hyperopic or astigmatic anisometropia (1-2D) > mild amblyopia.
- ❖ Mild myopia anisometropia (less than -3D) > usually doesn't cause amblyopia
- ❖ unilateral high myopia (-6D) > severe amblyopia visual loss.
- ❖ The eyes of a child with anisometropic amblyopia look normally to the family and primary care physician.

This is anisometropia



Type of amblyopia

- 1- Strabismic
- 2- Deprivation amblyopia
- 3- Anisometropic amblyopia



This is alternating strabismus they alternating between two eyes and the won't have amblyopia.



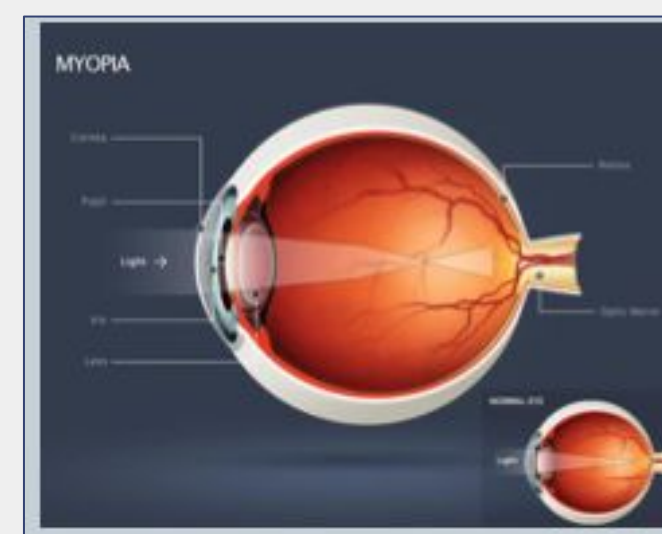
Anisometropic Amblyopia (Refractive Amblyopia):

- **Anisometropia**

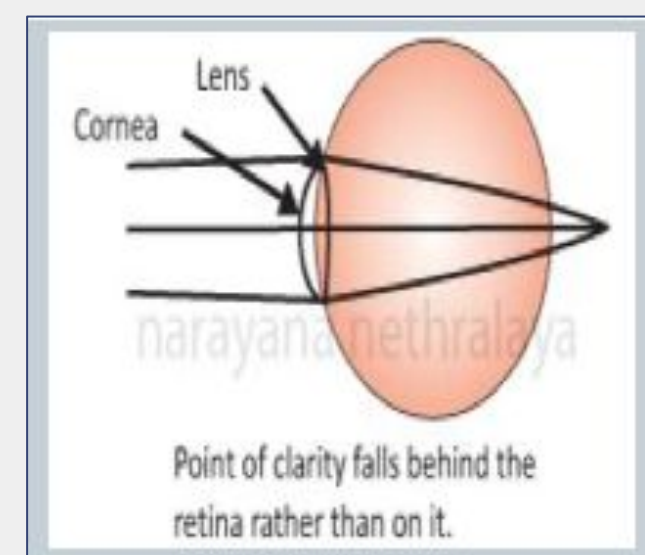
- 1.50 D of hypermetropia.
- 1.50 D of astigmatism.
- 3.00 D of myopia.

(refractive amblyopia)

- More common with **hypermetropic anisometropia**
- (Astigmatic (Meridional) amblyopia=



Hypometria



Hypermetropia. More Amblyopagenic

- It means both eyes are not sending clear images because of different refractive errors in each eye.
- One eye will send clear image and the other is blurry, the brain will receive 2 different images. The brain will prefer the clear image and tell the other eye not to send images > amblyopia.
- **The only way to find out anisometropia is to screen the child**, at 6 months, 2 years, 4 years, 5 years (4 times).
- **The child will never complain of anything** and the family would not notice anything because both eyes look normal.

Deprivational Amblyopia



Diagnosis: unilateral congenital ptosis
Complication: amblyopia
Treatment should be started ASAP

- **Cataracts, corneal opacities, eyelid ptosis, eyelid hemangioma:**
- These conditions will develop amblyopia because it prevent the light to go inside the eye > amblyopia.
- Result in more severe visual impairment than strabismic or refractive amblyopia.
- Why is the cornea a good medium for bacteria? Because it is an avascular structure, so no WBCs coming, and the infection will become severe.
- Amblyopia can't be prevented in trauma patients, but it can be reversed when it occurs.
- If a child is born today with cataract and you leave him for one month he will develop amblyopia..
- Eye with cataract will prevent access to light. No light entrance



Cataract secondary to trauma



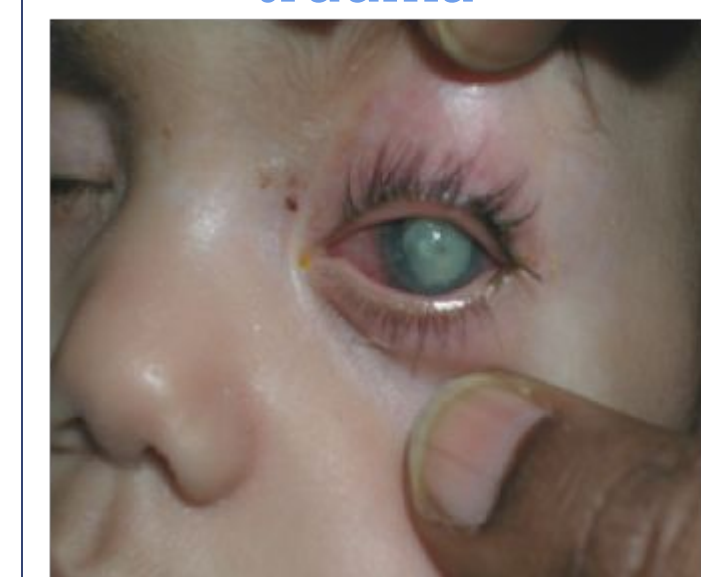
Hemangioma causing drop of the eyelid



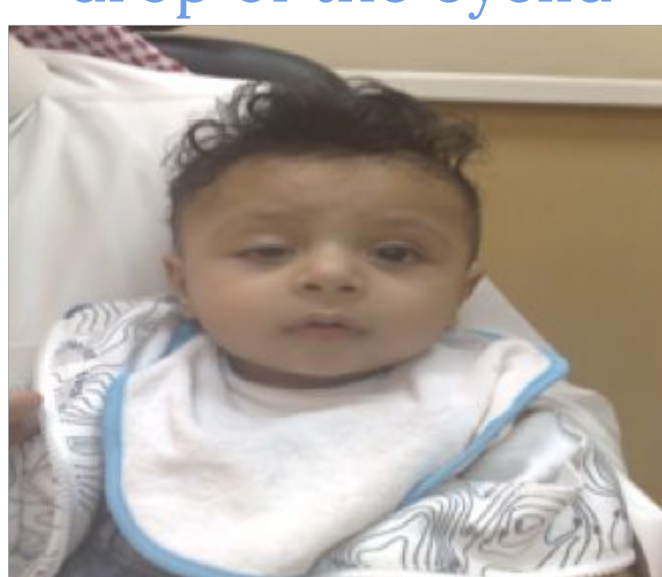
Diagnosis: capillary hemangioma of right eye
Complication: amblyopia



Bilateral congenital cataract



Infection (corneal ulcer) secondary to trauma > cornea become opaque



Congenital ptosis; prevent air and light



Occlusion amblyopia from excessive patching

❖ Screening for Amblyopia

- Half of all amblyopia cases undetected until age 5 years.
- All newborn infants should be screened in the nursery with the use of a red reflex test to check for media opacities.

Management of Amblyopia:

- ❖ **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.

These points are very important for the exam!!!

- Causes of amblyopia are : strabismus, anisometropia, deprivation amblyopia(cataract,hemangioma,ptosis ...)
- 2 main types is strabismus: comitant (more in children) and non-comitant.
- Types of comitant :(esotropia, exotropia).
- We covered the esotropia more because it is related to amblyopia.
- Types of esotropia:
 1. Infantile: happens before 6 month and caused by contracted middle rectus > treated by surgery (treat the strabismus) + occlusion (treat the amblyopia).
 2. Accommodative : happens after 6 months , caused by hyperopia> convergence > amblyopia , so we treat them by convex glasses (NO surgery) and if there is amblyopia treat by occlusion. (less accommodation > less convergence)
- how do know the child is having amblyopia ? visual acuity.
- Pseudostrabismus : caused by flat nasal bridge > diagnose first with corneal reflex then confirm (most confirmation) by cover test.



For your level you must know these things:

- Amblyopia is very important to detect and treat.
- We have two types of squints:
 - A. Comitant (children).
 - B. Non-comitant (adults due to neurological causes).
- Also, know that in children we **care more about esodeviation** more than exodeviation because it is more related to amblyopia (lazy eye). **Don't say it will go away!**
- Esodeviation types:
 - A. Infantile → first 6 months of life, treated by surgery.
 - B. Accommodative: treat by convex glasses.
- If you see a child with squint know if it's real or pseudo.
- Can we know the anisometropia in children? Yes, by screening.

Strabismus حول

- 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.

Classification of Strabismus:

- According to many things:

1- Fusion status: Both eyes control together that called binocular fusion.

- Phoria: حول مخفي

Latent tendency of the eye to deviate and controlled by fusional mechanism.

Both eyes control together and they are straight, when you break binocular fusion by test called **Alternatives 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 appears.**

- Intermittent Phoria:

Fusion control is present **part** of the time.

- Tropia:

Manifest misalignment of the eye **all** the time.

2- Fixation:

- 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.

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.

3- 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.

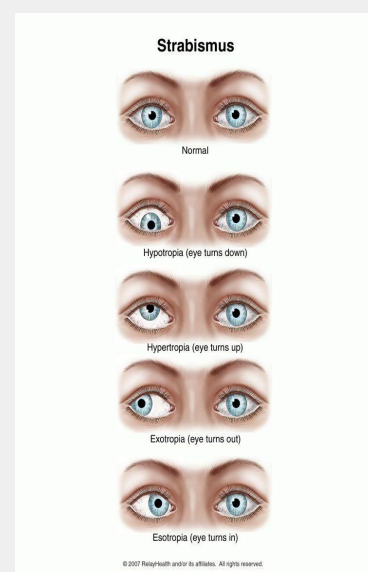
2. Vertical

- 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 acommedation so they suppose to be plus 1 at least plus 3 .in children are plus 6 or plus 7 (esotropia)

4- Age of onset

- Congenital at birth
- Acquired

Examination cont...

- ❖ **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 funds.
 - ❖ **Cycloplegic refraction** = to relax eyes and see real refractive error
 - Tropicamide 'short half life 20min'
 - Cyclopentolate 'intermediate tow days to three'
 - Atropin 'two weeks'
- "Dr said: side effects in pharmacology and toxicology lecture are very important".

We put cycloplegic refraction in

- 1- Pediatrics
 - 2- 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 (تشدد فتحاول تعوض +3 فتصير +6) but when kid feels fatigue (increase temperature, head trauma = feel weakness) so muscles are decompensated and eye will deviate inside (this is history)

- كان نظره ممتاز وطاح على رأسه وتعب وعينه دخلت على طول ليه صار كذا؟ زي ما شرحنا فوق العضلة ما عااد تتحمل تعوض

In summary: Pathophysiology: kid has +7 and tries to compensate with time when there is fatigue can't compensate and inside deviated happens and by adding cycloplegic refraction doctor can see real refractive not pseudo refractive.

Types of Strabismus

- In general, management of squint
- 1- rule out pathological causes like cataract by examining funds by fundoscope.
 - 2- correct refractive error حتى لو بسيط (cycloplegic refraction), (may hyperopia cause esotropia)
 - 3- 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\dayetc) till 6-7 years old التغطية ماتنفع خالص بعدها
 - 4- if not improved go to surgery.

A- Esotropia

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

1. Pseudoesotropia حول كاذب
2. Infantile esotropia
3. Accommodative esotropia
4. Partially accommodative esotropia

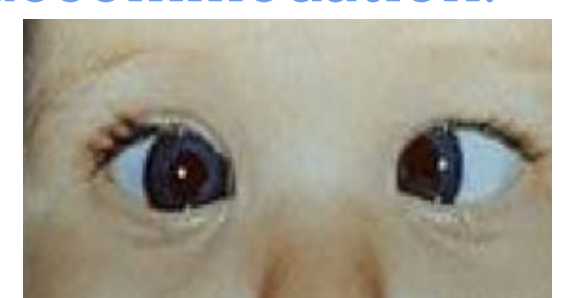
❖ Pseudoesotropia

- Occur in patients with flat broad nasal bridge and prominent epicanthal fold like in children and Asian.
- Gradually disappear with age.
- **Hirschberg test** differentiate it from true esotropia.



❖ Infantile Esotropia

- Common comitant esotropia occur before six month of age.
 - 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.
 - **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



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).

➤ Treatment:

1. Full cycloplegic correction.
2. Treat amblyopia.



❖ Partially Accommodative Esotropia

- Improve partially with glasses.

➤ Treatment:

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 gives 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.

B- Exotropia represent 5% to 10% cases of squint

- Intermittent exotropia the commonest type
- Constant exotropia
- Sensory exotropia
- congenital

❖ 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 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.

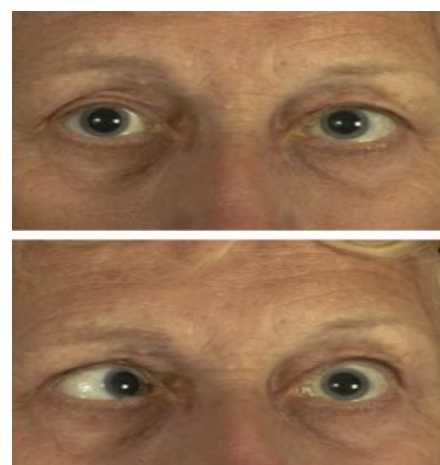
C- Paralytic strabismus "Incomitant"

Neuro ophthalmology lecture is very important esp 6th nerve palsy "exam"

- 6th nerve palsy
- 4th nerve palsy
- 3rd nerve palsy

❖ 6th Nerve Palsy in exam

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



❖ 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 or medical? by pupil examination*



❖ 4th Nerve Palsy

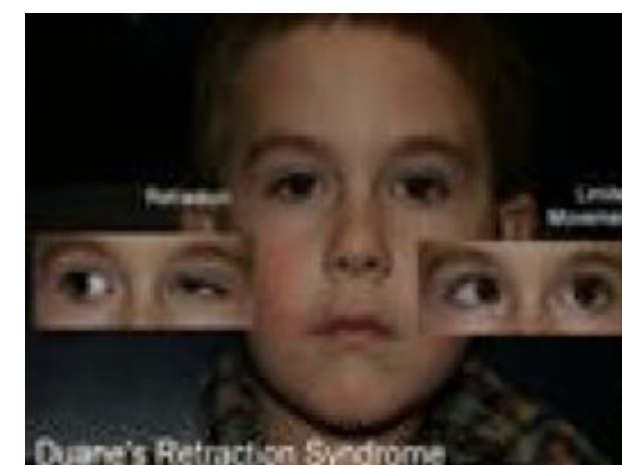
- Congenital or acquired
- Hypertropia of the affected eye with excyclotropia
- Abnormal head position

D- Special Types of Strabismus

1. Duane strabismus
2. Brown syndrome
3. Thyroid ophthalmopathy

❖ 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 (العين تدخل جوا) and narrowing of the palpebral fissure on adduction.** (تخيل في لمتيشن في الابدكشن والادكشن فالعضلتين يشدون فتدخل العين)
- 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 الابدكشن 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** العين ماتطلع .
- 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.

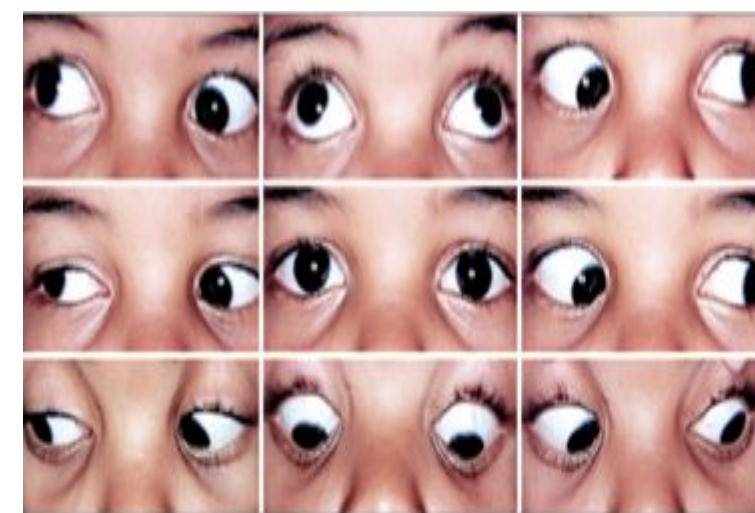


Figure 1 - The nine positions of gaze, boy

❖ Thyroid Ophthalmopathy

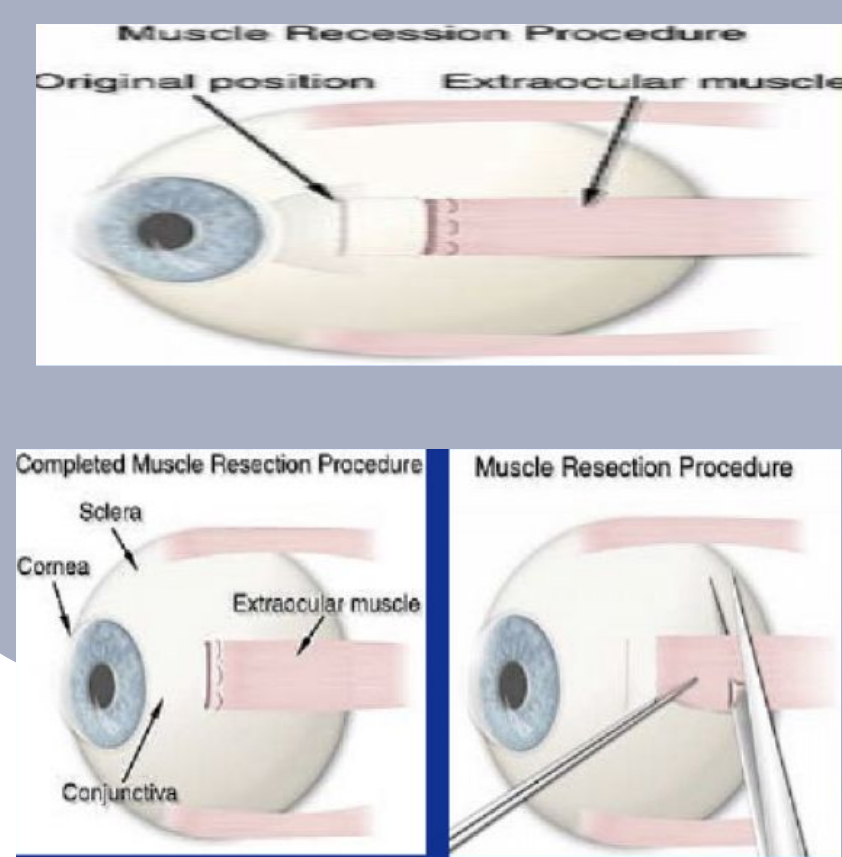
- **Restrictive myopathy commonly involving inferior rectus** commonest to be affected, then medial rectus 2nd most common and superior rectus.
- Patients presents with hypotropia, esotropia or both. **Orbital diseases are very important**



Surgery of Extraocular Muscle

1. Recession **عضلة ارخي**: weakening procedure where the muscle disinserted and Sutured posterior to its normal insertion
2. Resection **عضلة اشد**: 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

What is Amblyopia ?(Lazy Eye)

- Amblyopia refers to a 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:

- The 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 detect 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

Many causes of amblyopia exist; the most important causes are as follows:

Anisometropia

- Anisometropia is different 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 اللي عنده هذه المشكلة مايشوف القريب وهذا يسبب له امبلوبيا**.
- anisomyopia: **الطفل اللي ماعنده مشكلة بالقريب المشكلة عنده فقط بالبعيد فما يسبب امبلوبيا**

Visual acuity of normal person is 6/6, abnormal visual acuity: 6/+6 (7,8...) = he sees from 6m but normal person sees from 7,8...m causes of abnormal visual acuity may cataract, refractive error, glaucoma, amblyopia...

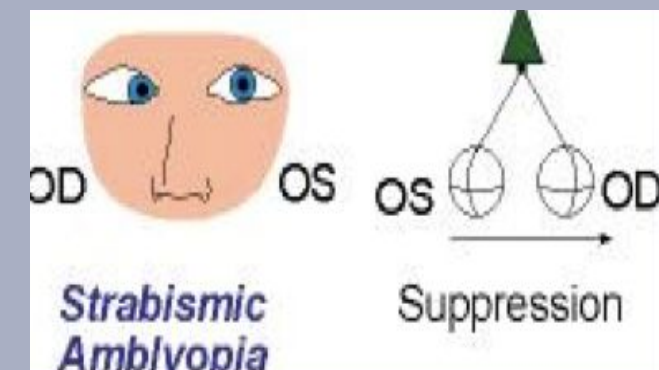
Normal refractive error in pediatrics are hyperopia
hyperopia = small globe and myopia = large globe

So kids have small eyes (e.g. length of children's are 16 with growing will be 24) so hyperopia with growing transfer from hyperopia to plain and some of them transfer to myopia. Physiological -refractive error- hyperopia will be plus 1,2 to 3,

❖ Causes of Amblyopia Cont

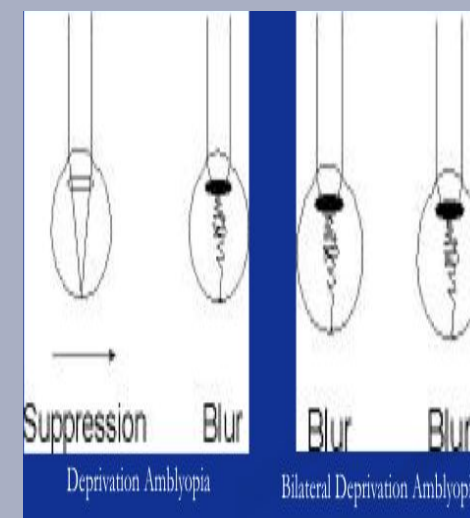
Strabismus

- 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.**



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.



Organic

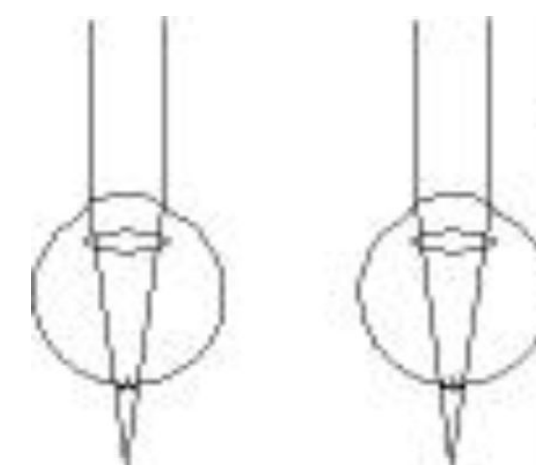
Structural abnormalities of the retina or the optic nerve may be present. Functional amblyopia may be superimposed on the organic visual loss

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

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



Direct ophthalmoscope: assessing red reflex



Normal red reflex



Direct ophthalmoscope: examining retina

& fundus



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

❖ Treatment

- You have to treat the cause and patch the good eye to stimulate the weak one.
- The clinician must first rule out an organic cause and treat any obstacle to vision (eg, cataract, occlusion of the eye from other etiologies).
- Remove cataracts in the first 2 months of life, and aphakic correction must occur quickly
- Treatment of anisometropia and refractive errors must occur next
- The next step is forcing the use of the amblyopic eye by occlusion therapy



Leukocoria

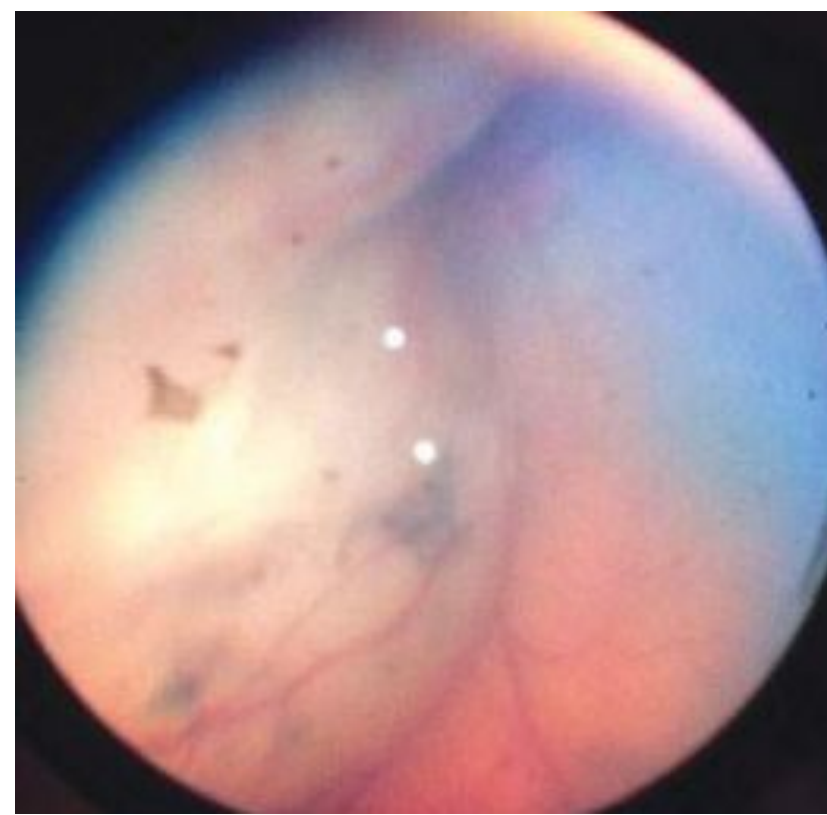
- A white pupillary reflex. It differs from absent red reflex.
- Leukocoria in a child requires **urgent** attention, primarily because in most patients with retinoblastoma it is the first sign noticed.
- Secondarily, a white pupil indicates a severely amblyopiogenic condition, which may be treatable. This is bad prognosis.
- Anatomic location is important in the differential diagnosis of Leukocoria



Abnormal left eye. This pic is without ophthalmoscope, the child is walking like this.

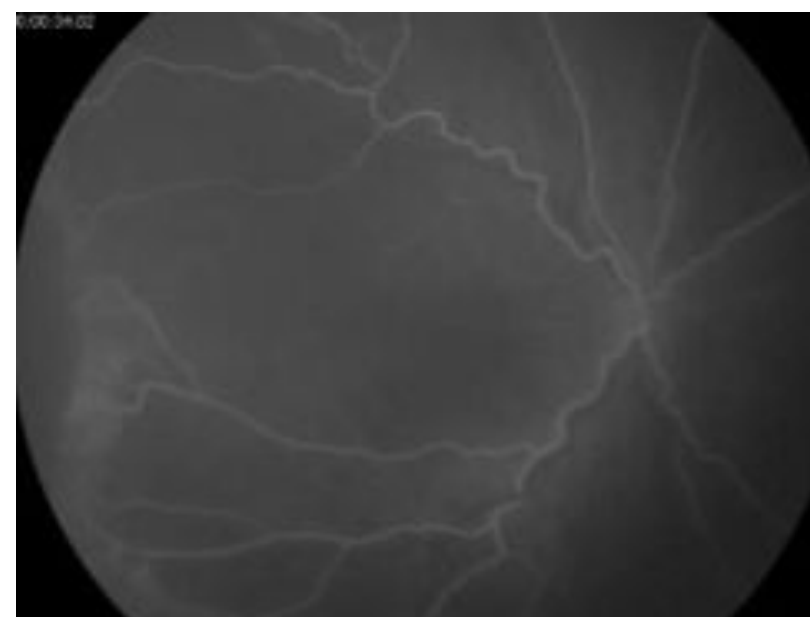
❖ Causes of Leukocoria

- Cataract
- Retinoblastoma
- Toxocariasis
- Coats' 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



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.

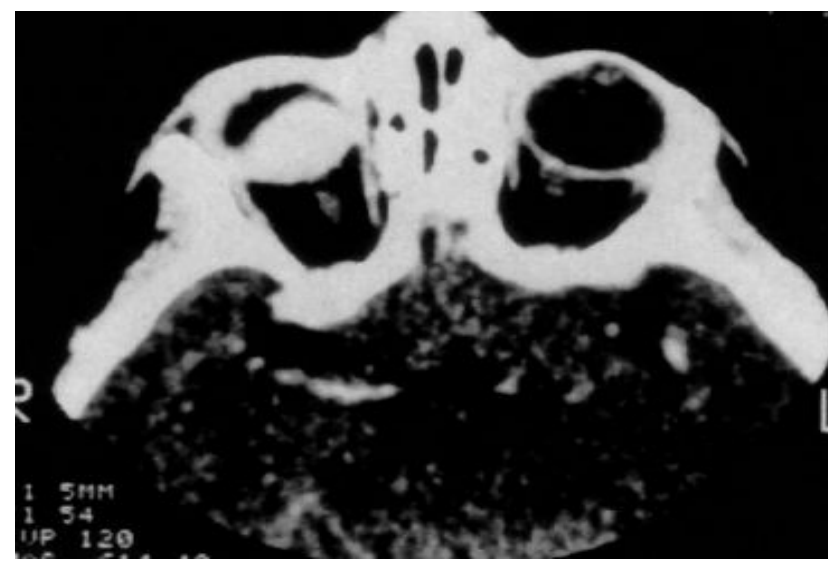
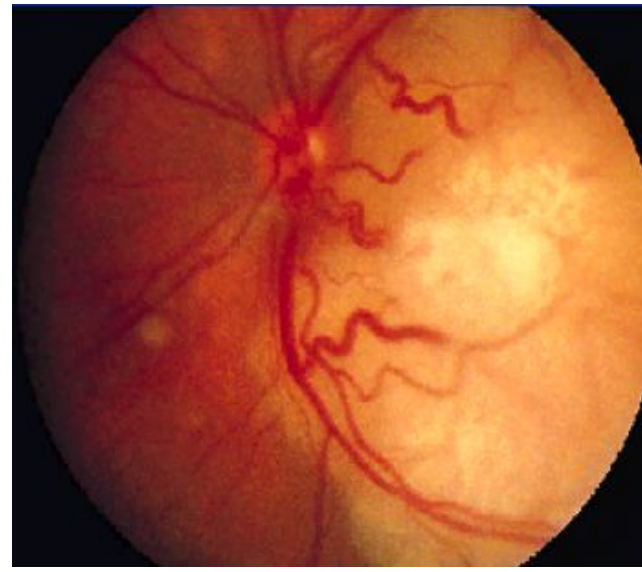
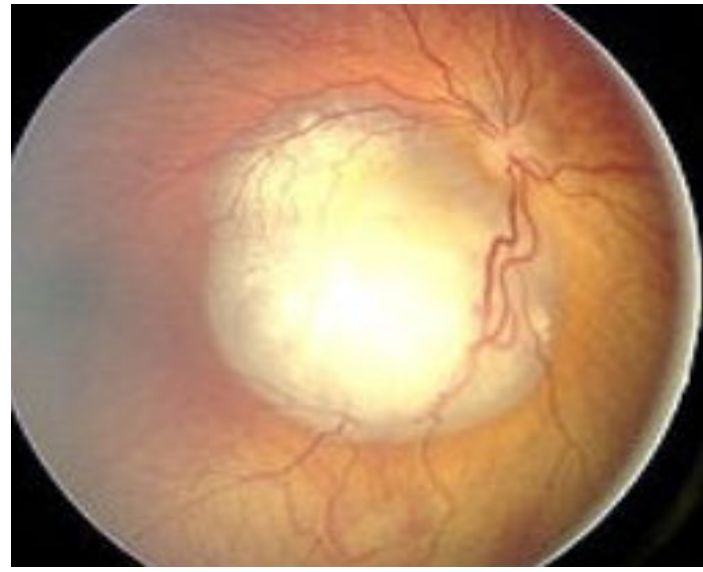


RETINOBLASTOMA

- 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.
- 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

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. Patients have no history of prematurity or oxygen administration. b.c. If the baby premature it's possible to have retinopathy of prematurity.

RETINOPATHY OF PREMATURE (ROP)

Vasoproliferative retinopathy affecting premature infants exposed to high oxygen

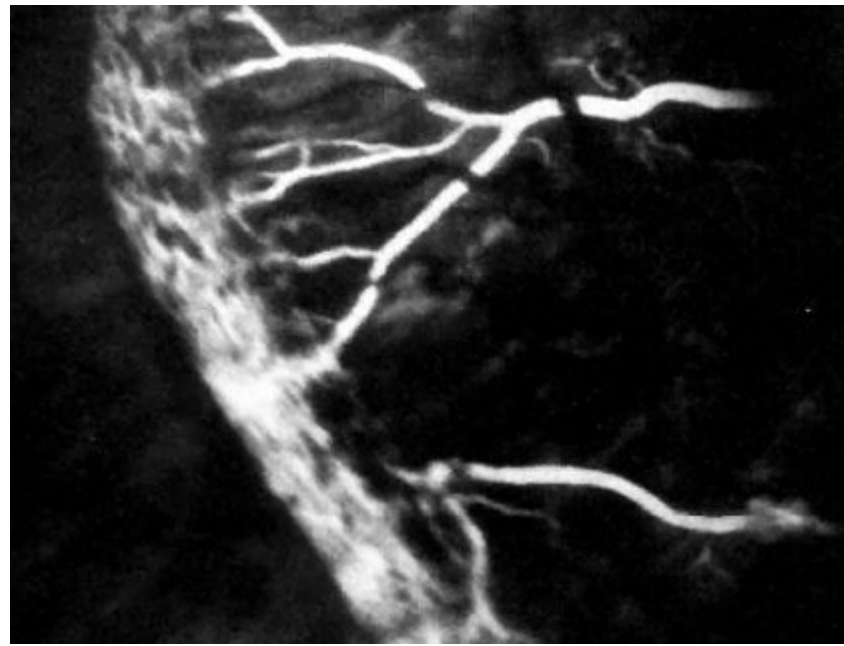
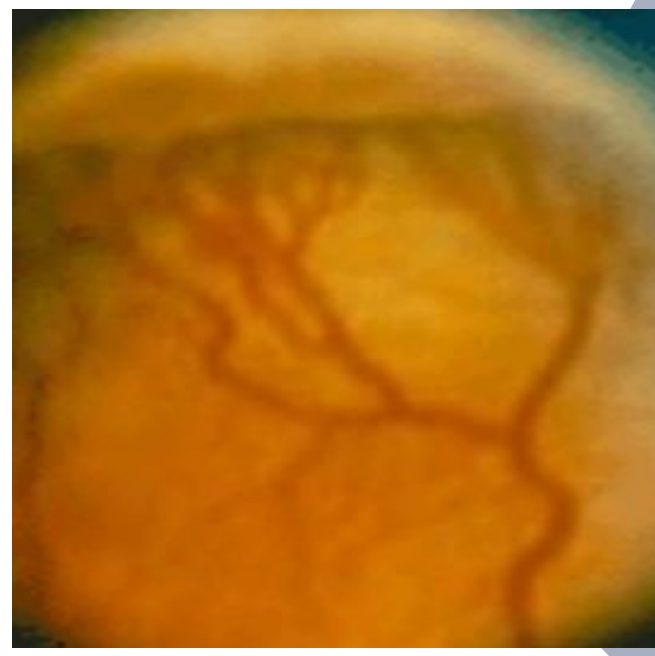
- Normally: in the 8th month half of the retina have blood vessels & the other half doesn't, it continues growing till delivery where it becomes fully mature, at that time when the baby exposed to oxygen it stops growing. Premature baby <32 weeks: blood vessels of the retina is on the middle didn't reach 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

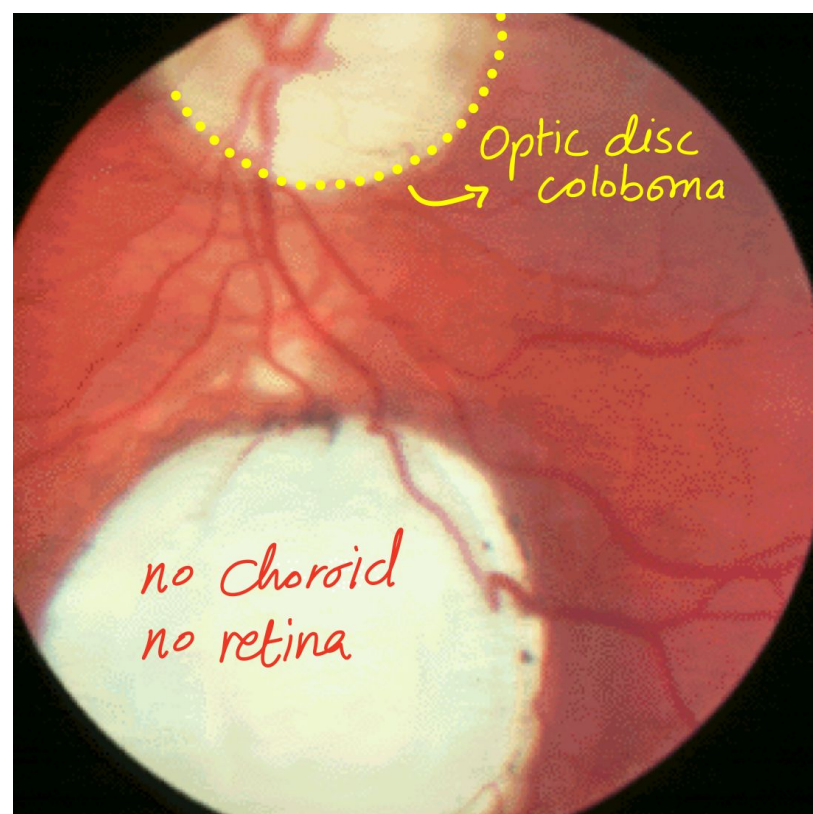
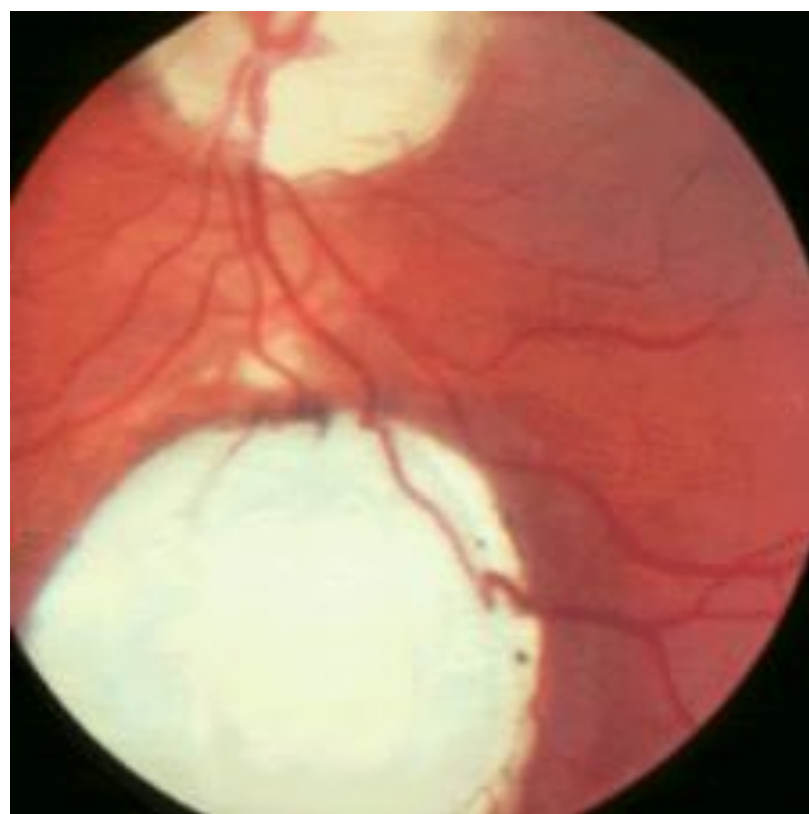
- Retinal Ischemia > ↑no. of VEGF > neovascularization
- fibrous bands
- retinal detachments
- vitreous hemorrhage
- leukocoria
- no hard exudates (no proteins) it founds in diabetic retinopathy or vein occlusion.



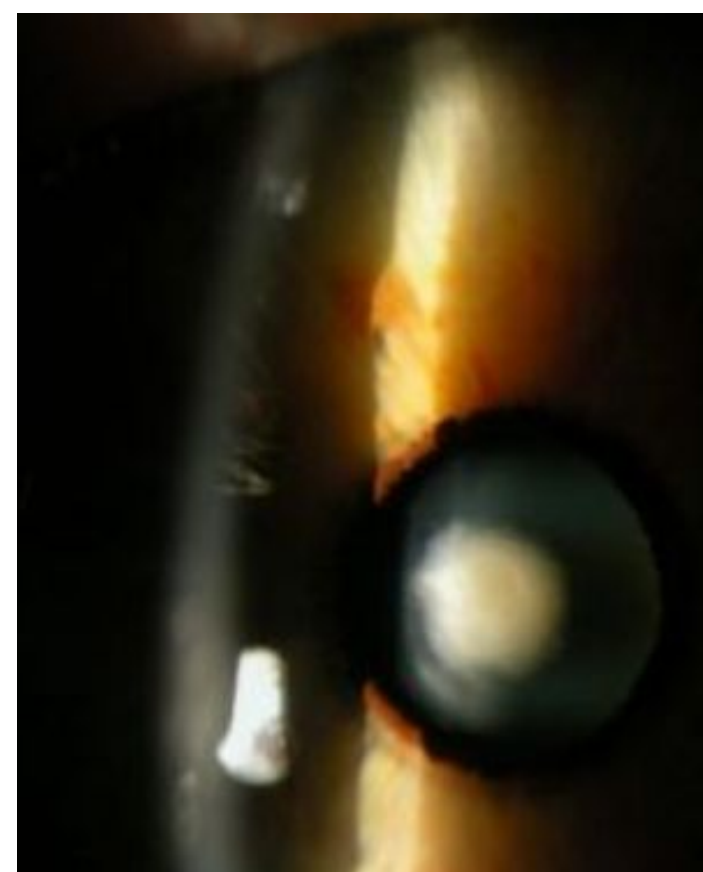
Fundus with fluorescein angiography, we do laser in the area with no blood vessels.

COLOBOMA

- Coloboma means no tissue development or agenesis.
- It can also be in the iris > iris coloboma > no iris tissue.
- Optic Disc Coloboma
- During 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 NPL if the coloboma was away from optic disc & macula.
- If it was in the optic disc or the macula there might be no light perception.



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



Practice Questions

- Q1. The mother of one- -and- -a- -half- -year- -old child gives history of a white reflex from one eye for the past 1 month. On CT scan in the orbit there is calcification seen within the globe. What is the most likely diagnosis?**
- A. Congenital cataract.
 - B. Retinoblastoma.
 - C. Persistent hyperplastic primary vitreous.
 - D. Coat's disease.
- Q2. A child with accommodative esotropia, after complete correction of the the esotropia with glasses. The left eye had decreased visual acuity compared to the right. What is your next management?**
- A. Patch the eye.
 - B. Increase the lenses.
 - C. Correct the amblyopia.
 - D. Refractive surgery
- Q3. You were asked to examine a preterm newborn in the NICU. Fundus examination revealed engorged blood vessels at the retina. Which of the following should you rule out?**
- A. Hyperplastic primary vitreous.
 - B. Congenital cataract.
 - C. Retinoblastoma of prematurity.
 - D. Retinoblastoma.
- Q4. A premature infant with a gestational age of 20 weeks and birth weight of 1200 gram. The mother noticed white pupil what is most likely diagnosis?**
- A. Retinoblastoma.
 - B. Coats disease.
 - C. ROP.
 - D. Coloboma.
- Q5. A 65 years-old patient w/ uncontrolled DM, recently diagnosed w/ HTN came to ER complaining of double vision. On examination, the doctor noted esotropia in her right eye. What is the name of affected nerve?**
- A. Trigeminal.
 - B. Oculomotor.
 - C. Abducent.
 - D. Optic.

Practice Questions

- Q6. A 9-year-old girl presented with outward deviation of the eye, with normal fundusoscopic examination. Which of the following tests is the most appropriate to confirm the diagnosis?**
- A. Corneal light reflection test.
 - B. Cover uncover test.
 - C. Visual acuity.
 - D. Extraocular muscle motility.
- Q7. A 12-year-old child came for pre-school routine examination. His best corrected visual acuity in the right eye is 20/20, and in the left eye 20/100. Ocular examination showed normal anterior and posterior segments. Which of the following best describes the patient's situation?**
- A. Astigmatism.
 - B. Myopia.
 - C. Hyperopia.
 - D. Amblyopia.
- Q8. A premature infant with a gestational age of 20 weeks and a birth weight of 1200 gram. On retina screening examination. What eye problem is he more likely to develop?**
- A. Coat's disease.
 - B. Neovascularization of optic nerve head.
 - C. Macular ischemia.
 - D. Retinal detachment
- Q9. A 2 months old child came with his parents to the clinic complaining of Congenital cataract, what is the proper time to do cataract surgery?**
- A. At age of 2 years to insert the IOL.
 - B. At age of 1 year to avoid the General anesthesia complication.
 - C. As soon as possible.
 - D. At any age until 6 years.
- Q10. A child came to the clinic upon light reflex the light was shifted temporally in the right eye and central in the left eye. What is the diagnosis?**
- A. Esotropia.
 - B. Exotropia.
 - C. Ephotropia.

Answers:

1: B 2: C 3: C 4: C 5: C 6: B 7: D 8: D 9: C 10: A