

leukocoria, amblyopia and strabismus

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material :

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Toronto notes

doctor record

Leukocoria (white pupil):

It is the absent of pigmentation in retinal pigmented epithelial (RPE) under the ophthalmoscope .

Causes of leukocoria:

CATARACT

1-Cataract :

The commonest cause of leukocoria in children : Cataract . can be congenital or acquired, usually causes blurred vision and glare .

Causes of cataract in the children:

30% unknown

30% inherited

30% Intrauterine infection, truma, radiation ,metabolic disorder like diabetes.

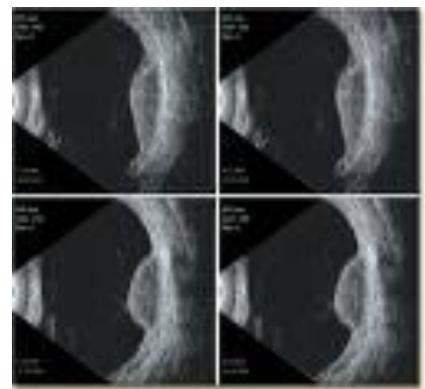
So it is important to perform red reflex for every neonate to eliminate cataract .

We use the ultra sound to make sure that the cause of the opacity of the eye related to the lens (in the figure the opacity was due to Retinoblastoma which is a life threatening).

Management :

Unilateral: immediate surgery before one month of age or he will lose his vision and develops amblyopia.

Bilateral :surgery in the first 2 or3 month of life>>less urgent because the brain will not prefer one image on another >>no amblyopia.



2- Persistent hyperplastic primary vitreous(PHPV) :

a congenital condition caused by failure of the normal regression of the primary vitreous. It is usually associated with unilateral vision loss. During embryology the blood vessels will come from the optic nerve to nourish the developing lens , this vessels should regress and disappear, if this process did not accomplish in a right way the blood vessels will persist and keep attached to the lens and the lens become pale, there will be an opacity in the lens and the eye become smaller **imp.**

(cataract ---< small eye ---< poor vision and leukocoria)

Treatment : remove the opacity in the lens and cut the blood vessels.



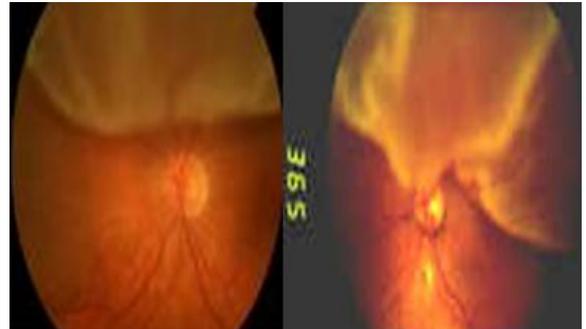
3- Organized vitreous hemorrhage:

vitreous hemorrhage is usually secondary to a neovascular membrane or to a retinal tear. Patients may complain of a red haze, blurred vision, or floaters. When the blood starts to resolve the hemoglobin will give different colors like whitish or yellowish color some fibrous sheets may persist. A B-scan (ultrasounds) is usually diagnostic and vitrectomy is usually required.



4- Retinal detachment :

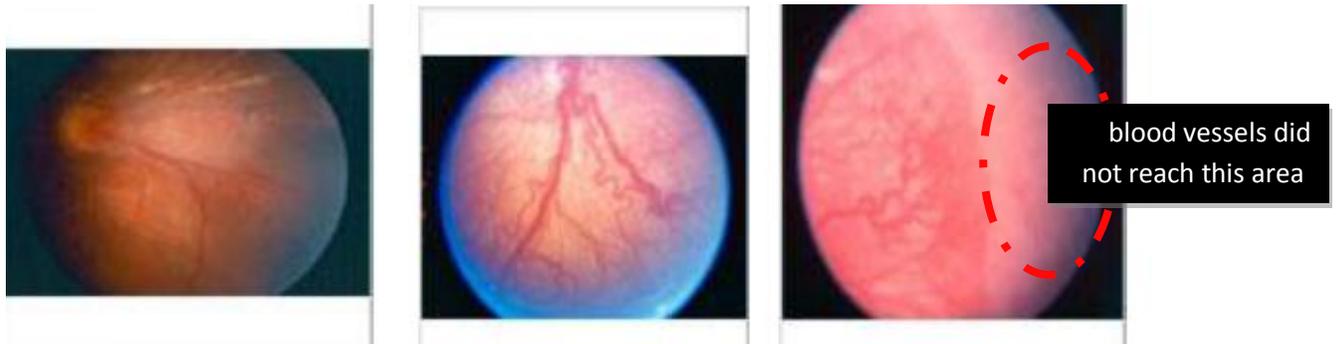
Why retinal detachment cause leukocoria?
Neurosensory retina are transparent behind it is the retinal pigmented epithelium which give the color of the retina, if those two layers got separated then we will have white color (leukocoria). If it is big you will see leukocoria and this a bad sing . Causes of RD: high myopia and trauma.



5- Retinopathy of prematurity:

During development, the blood vessels grow from the optic nerve and go continuously to the peripheral of the retina and keep going until the 37th week of gestation .
When the baby born prematurely before term the blood vessels will not have the time to complete the growth to the peripheral area>> no blood supply >> ischemic>>> formation of new blood vessels but these blood vessels are fragile so they will bleed. The blood contains WBCs, macrophages and fibroblast all this will cause fibrosis and contraction of the retina so the retina will detached.
If we did not detect it and treat it, it will cause blindness.
Treatment: by laser to kill the area and reduce the demand of blood.
Detection by screening .

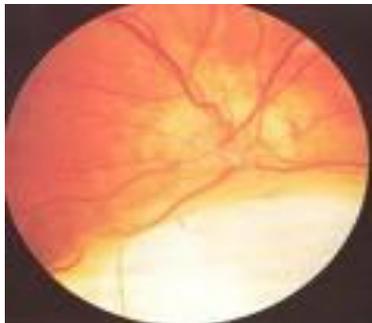
who to screen? Gestational age of 28 weeks or less and Birth weight of 1500 g or less. **imp**



6- Coloboma (absent of tissues) :

Congenital condition caused by incomplete closure of the fetal fissure . Degree of visual loss related to area affected (iris, retina, choroid, or optic nerve head)

- When it is anterior (iris coloboma)it will cause absence of the iris tissue (the iris become incomplete) It will give : key hole OR cat eye appearance **but it will not give leukocoria .**
- When the coloboma extend backward to the retina and retinal pigmented epithelium (posterior or choroidal coloboma) so there will be no retina no retinal pigmented epithelium, there is only sclera (whit area) , if it is big it will give leukocoria.



leukocoria in coloboma



the most common presentation is inferior ,nasal

- If the it is involve the macula it will cause poor vision.

7- Medullated nerve fibers :

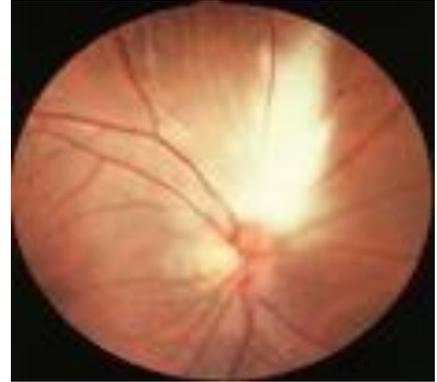
congenital anomaly caused by myelination of the retinal nerve fibers and usually **asymptomatic**. When large areas are involved it can cause leukocoria.

Cats Fur appearance in Medullated nerve fibers

There are 1.2 million nerve fibers and they should not be myelinated.

If the myelin involve the nerve fibers in the macula it cause poor vision.

Treatment : no treatment.



8- Coat's disease:

Typically a unilateral malformation of the blood vessels condition found in young boys. It is characterized by retinal telangiectasia and aneurysms that may cause exudative retinal detachments.

Telangiectasia of the blood vessels, the blood vessels will leak out lipid and cause exudates (exudates are lipid) that give leukocoria if it is big enough .

Treatment : by laser the leaking point.



After laser therapy

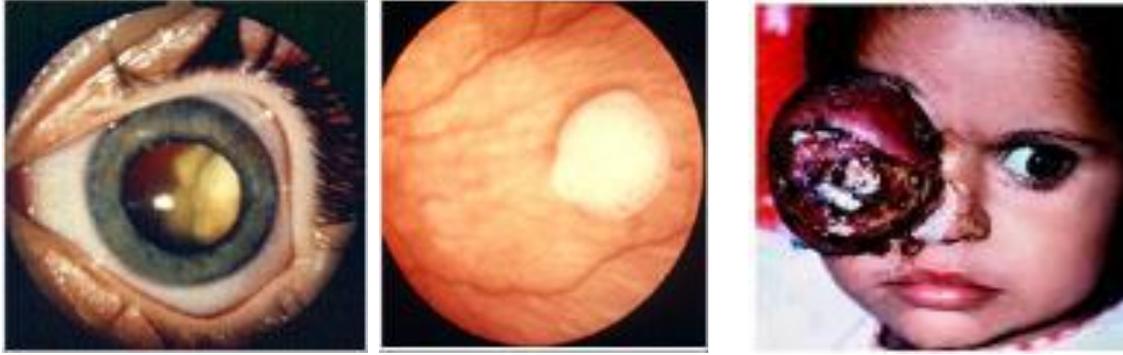
9- Retinoblastoma:

Life threatening , most common primary, malignant, intraocular tumor of childhood but still a rare tumor. Vast majority become apparent before age of 3yrs. It results from malignant transformation of primitive retinal cells before final differentiation.

Presentation is most commonly(60%) with leukocoria and strabismus because it will block the fovea the eye will deviate.**imp**

Treatment: by Chemotherapy, radiation or Enucleation of the eye (removing of the eye) depend on: the size of the tumor and the distance from the optic nerve.

If the tumor involves the optic nerve , it will go back to the brain and this what kill the patient.



Late stage of Retinoblastoma

AMBLYOPIA

We need two straight eyes to have Binocular vision to appreciate the depth of the image.

In order to have binocular vision we have to have :

- Two eyes.
- The image should fall on the fovea in both eyes.

Then both eyes will send both images (from the right and the left) to the cortex.

In order for the brain to unify the two images, the two images from both eyes should be similar in : size, shape and clarity.

if the brain comes to unify the two images and find them different from each other so the brain will take the good one and cancel the other image, with time the brain will continue to cancel the image from the abnormal eye and this will lead to lazy eye or Amblyopia.

DEFINITION of AMBLYOPIA:

Reduction in visual acuity in one or both eyes. This reduction cannot be completely accounted for by any clinically apparent organic abnormality.

Always occurring before the age of 7 years but it is reversible if we catch the child in 5 or 6 year of age .

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.

Classification :

According to the condition that induced it:

Strabismus : Any ocular misalignment or tendency toward misalignment. is the most common amblyopia-inducing condition . 40% of children with a manifest strabismus develop amblyopia . Esodeviations are more common than exodeviations.

ANISOMETROPIA :

different in refractive error between the two eyes.

The amount of different is:

1.5 D in hypermetropia

1.50 D in astigmatism

3.00 D in myopia

the brain cannot unify them so the brain will cancel one eye and become amblyopic.

It is difficult to detect because the child will not complain because he use his normal eye without noticing the problem in the other one >>>so we need a good screen program .



Deprivational Amblyopia (the visual axis is not clear) :

Results in more severe visual impairment than strabismus or refractive amblyopia .

Couses :

1-Cataracts and corneal opacities



Corneal Abrasions

corneal ulser

2-Eyelid ptosis, eyelid hemangioma



Screening for Amblyopia:

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

Infants should be evaluated for fixation preference, ocular alignment, and eye diseases by 6 months of age.

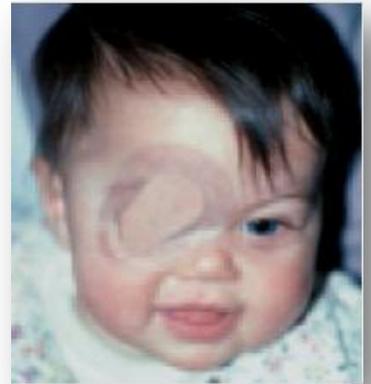
Management:

Occlusion

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.



Strabismus

- is a general term referring to ocular misalignment due to extraocular muscle imbalance. In short, the eyes are "crooked".
- Strabismus occurs in approximately 2% of children under 3 years of age and about 3% of children and young adults.
- The condition affects males and females equally.

Why we are concerned about Strabismus?

1- is associated with reduction of [depth perception](#) and, if onset is in adulthood, double vision.

2-a cosmetic concern, especially for school-age children.

Squint in children will not cause diplopia because the brain can suppress the eye but in adult the brain cannot suppress the eye and the visual system are mature so they do not have diplopia.

Orthotropia: When an individual's eyes are straight, This indicates that both eyes are aimed at the same spot. The brain fuses the two separate images into one three-dimensional image.

strabismus are divided into two main types :

Noncomitant or paralytic squint : there is underaction of one or more of the eye muscles due to a nerve palsy, extraocular muscle disease or tethering of the globe.

Comitant or non-paralytic squint : the movements of both eyes are full (there is no paresis) but only one eye is directed towards the fixated target .

Types of comitant strabismus:

Esotropia

Exotropia

Esotropia: inward deviation of the eye and divided into :-

Accommodative or Infantile esotropia: the inward deviation of the eyes noted before the patient reaches age 6 months.

Age: This condition usually presents in patients aged 2-3 years.
the commonest type of esotropia.



- Near reflex 3 things:
- 1- Accommodation.
 - 2- Meiosis.
 - 3- Convergence.

Hyperopic patients accommodate in order to increase the refraction power to make image fall on the retina. In 2 years old child who have hyperopia he need accommodation and convergence if not treated it will lead to accommodative esotropia. But not all hyperopic patient will have esotropia



Perform cycloplegic refraction on all children by using the retinoscope and loose lenses. Cycloplegia is achieved with Mydracyl 1% and cyclogel 1%.

Clinical features of Acc ET:

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

Do we do surgery for Acc ET ?

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

Acquired non-accommodative:

Not common in children, the patient has esotropia but it is not congenital esotropia and does not have hyperopia. Causes : Tumor in the brain or neurological cause.

Alternating esotropia: esotropia moving from eye to the other , not fixed in one eye . It is good and it will not cause amblyopia.



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 .



A careful ocular examination (eg, pupillary light reflex) reveals that the eyes are straight. Using the cover-uncover test, the examiner finds that the patient manifests no deviation.

Exodeviation or Exotropia:

- Exodeviation is a horizontal form of strabismus characterized by visual axes that form a divergent angle.

-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 (the presence of a latent squint). This deviation may later progress to intermittent exotropia . intermittent exotropia can have an early onset, with 25-40% of cases occurring before the second year of life.



Intermittent. So Exotropia is not associated with amblyopia like esotropia.

Types:

Congenital : very very rare.

Intermittent : hirshberg test normal but cover uncover test shows mild exotropia.

This is the commonest type of exotropia.



All the previous types of strabismus are horizontal strabismus.

We have vertical strabismus:

□ acquired : in case of trauma there will be fracture in the floor of the orbit so the eye will not move , when the patient try to look u one eye will move but the other will not so he will have diplopia.



Other example of vertical strabismus : myasthenia graves, multiple sclerosis and thyroid eye disease.