

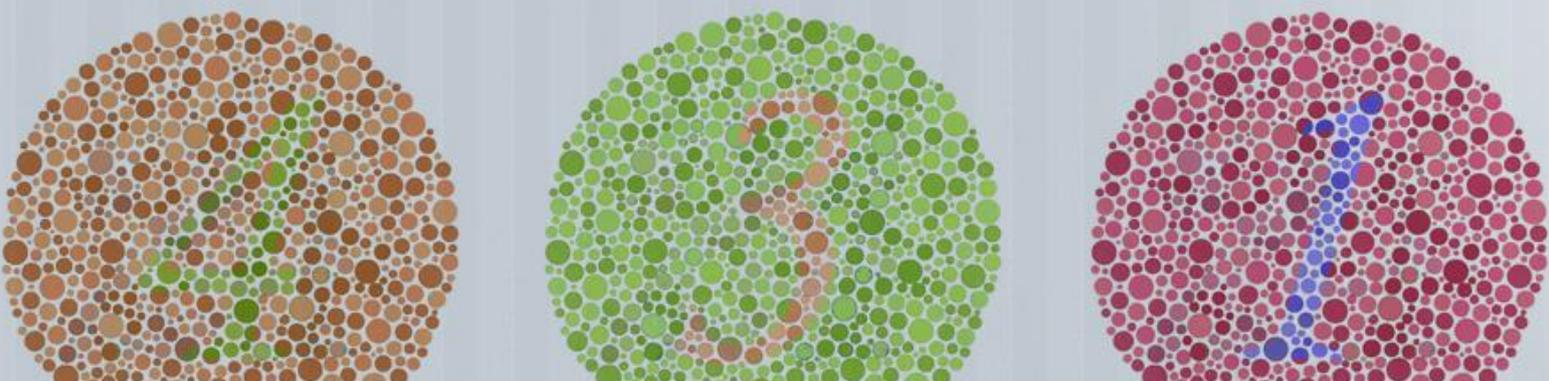
Strabismus, Amblyopia Management and Leukocoria



431
Teams
medical students

Ophthalmology Team

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From slides Doctor's Notes Team's Notes From the book Important

Lecture mostly contains pictures, but the doctor gave a lot of additional info which we added here.

Leukocoria

Leukocoria is **white opacity of the pupil**, and it is a **sign not a diagnosis**.



Causes will be presented going backwards through the eye structures:

1. Cataract

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



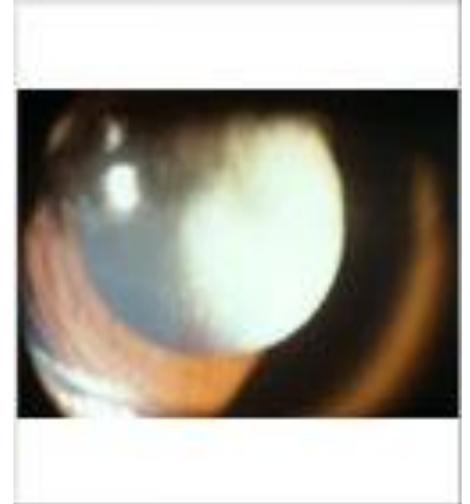
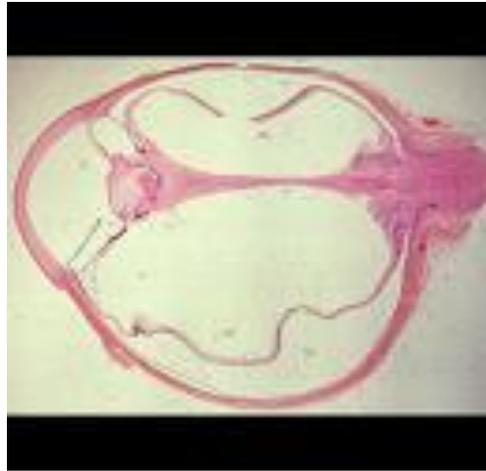
Using the ophthalmoscope if you see nice red reflex on both eyes (pic on right.) → unlikely to have any visual problems.

Doctor's notes:

- Congenital cataract is very important, because if you don't treat it in the first months of life → Irreversible amblyopia.
- For the brain to unify the 2 images → both should have the same shape, size and clarity. If one is clear and the other is not → brain gets confused → can't put them together → suppresses image from the cataract eye. If this continues for 2,3 or 4 months → amblyopia.
- For example: If the child presents with the problem at 1 year of age → already too late, you can't do anything. (Because amblyopia happens much earlier than 1yr)
- The eye is connected to the brain → Retina and optic nerve regarded as parts of the CNS → it's a neurological problem → difficult to reverse after 3 months of suppression.

2. Persistent hyperplastic primary vitreous

PHPV is a congenital condition caused by failure of the normal regression of the primary vitreous. It is usually associated with unilateral vision loss



Doctor's notes:

- During embryology, blood vessels come from the optic nerve to nourish the lens, they usually disappear → clear vitreous.
- If something happens, and they do not disappear → There will be a scar going from the optic nerve to the lens → lens will be opacified (cataract) → leukocoria

3. Organized vitreous hemorrhage

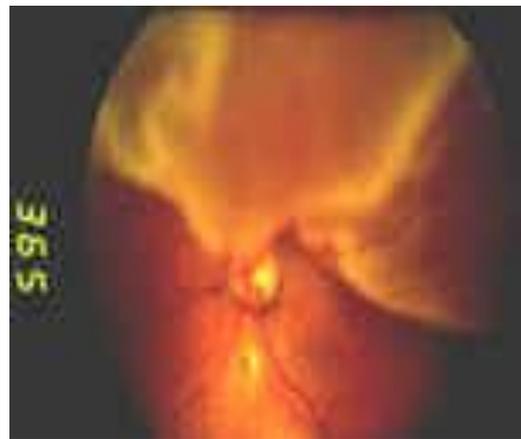
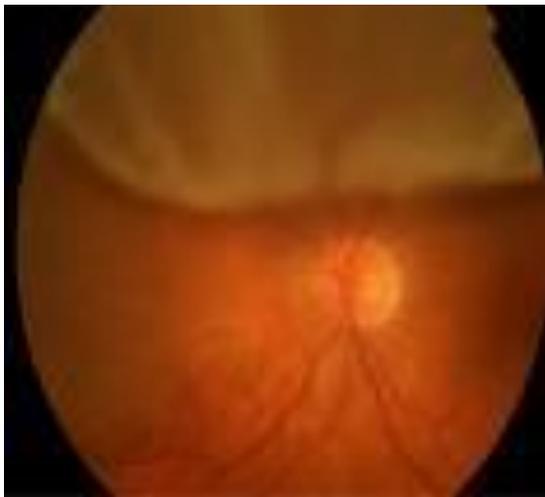
Vitreous hemorrhage is usually secondary to a neovascular membrane or to a retinal tear. Patients may complain of a red haze, blurred vision, or floaters. As it starts to resolve, color changes to yellow then white and some fibrous sheets may persist. A B-scan is usually diagnostic and vitrectomy is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



- You'll see the picture above when you use an ophthalmoscope on an eye with vitreous hemorrhage for 1-2 months.
- Blood will be removed, but iron stays for a long time → gives this golden color.
- If we don't know the reason behind this leukocoria → send the patient to ultrasound
- Sometimes it could be due to birth trauma (e.g. when the child is pulled using forceps).

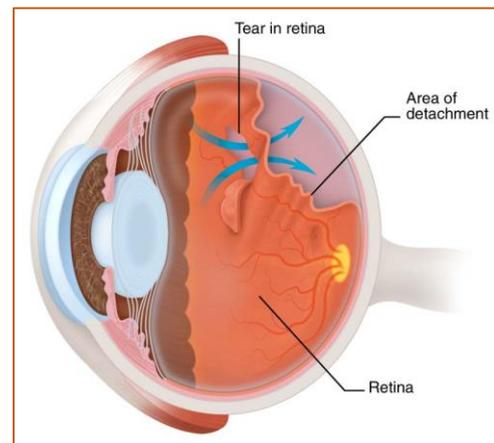
4. Retinal Detachment

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.



Doctor's Notes:

- Retina has 2 layers which are normally stuck together (neurosensory retina and retinal pigment epithelium). When separated → retinal detachment.
- High myopia → retina stretched → retinal break (hole) → vitreous gel (liquefied around age 40 or earlier in myopia) sweeps in → separates the 2 layers.
- Neurosensory retina not attached to the pigment epithelium → no red color seen, instead it'll be white.
- When you shine the ophthalmoscope → leukocoria.



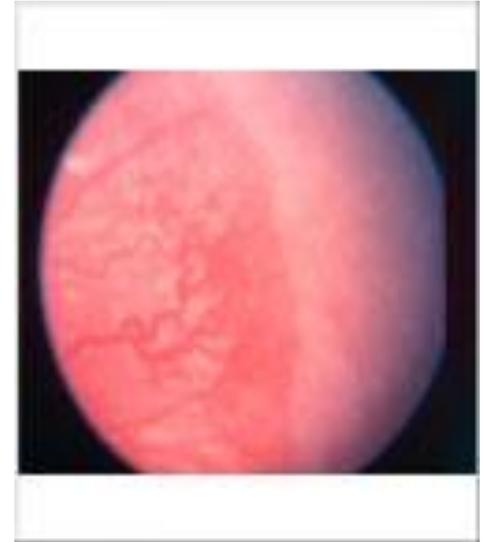
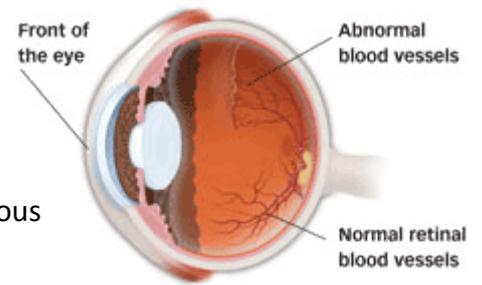
Retinal detachment, n.d. photograph, viewed 20 September 2014, <http://www.maireadoleary.ie/blog/wp-content/uploads/2014/04/retinal_detachment.jpg>.



Shadow, n.d. photograph, viewed 20 September 2014, <<http://www.maireadoleary.ie/blog/wp-content/uploads/2014/04/shadow.jpg>>.

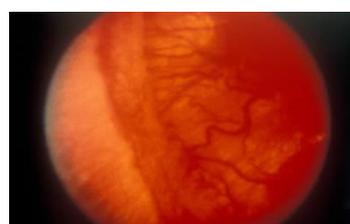
5. Retinopathy of prematurity

Occurs in **premature, low-birth-weight** (1500grams or less) infants maintained on oxygen therapy. Signs include neovascularization, fibrous bands, retinal detachments and vitreous hemorrhage. When advanced leukocoria can be present.



Doctor's notes:

- Usually in preterm (28 gestational weeks or less)
- So any child born preterm (≤ 28) or with low birth weight ($\leq 1500\text{gm}$) has to be screened before he leaves the NICU (Neonatal ICU).
- It can cause irreversible vision loss. If you go to a school of blind students, a good number will have a history of such case.
- During embryology, retinal blood vessels continue to grow till they reach the peripheries. This process continues till just immediately before birth.
- When the child is born preterm \rightarrow not enough time for vessels to reach all of the retina \rightarrow the remaining retina (with no blood vessels) becomes ischemic \rightarrow body will form new blood vessels (unfortunately fragile vessels) \rightarrow they will bleed and fibrosis will occur \rightarrow contraction of the retina and retinal detachment.
- This process can occur in the 4, 5 or 6 weeks following birth.
- If not managed early \rightarrow the child may go blind.
- Management:
 - We should kill this retina (the one with no blood vessels) \rightarrow \downarrow O_2 demand and \downarrow blood vessel proliferation.
 - This is done using laser, and we preserve the macula.
 - The patient wouldn't see the periphery but at least he's not blind and can manage his life.



6. Coloboma

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).

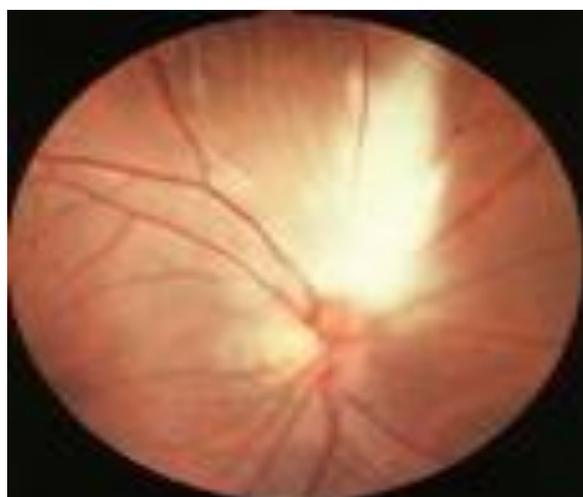


Doctor's notes:

- The iris will have a “cat eye” or “keyhole” appearance because part of the iris is not there → pupil (the opening) will not be rounded.
- Can extend backwards to affect the retina as well → there will be no retina, no retinal pigment epithelium → white sclera → leukocoria.
- The picture on the left shows choroidal coloboma (you see the bare white sclera under the choroid).

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.



Doctor's Notes:

- Nerve fibres here are normally unmyelinated → retina transparent.
- Congenital anomaly → myelinated → white color (like cat fur) appearance → white reflex (leukocoria).
- Fortunately not common.
- Only serious if it affects the macula.
- We can do nothing about it (no Tx).

8. Coat's Disease:

Typically a unilateral condition found in young boys. It is characterized by retinal telangiectasia (**dilated blood vessels**) and aneurysms (**and blood vessels**) leak that may cause exudative retinal detachments.



Doctor's note:

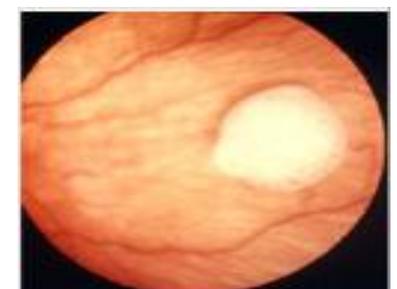
- Blood vessels leak lipids → yellow discoloration → leukocoria (if big enough)
- Also only serious if it affects the macula → affecting vision.
- Management: using laser to occlude the leakage in the blood vessels.

9. Retinoblastoma

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.

Doctor's notes:

- Tumor can invade the optic nerve and the brain → kills the child.
- When you find **leukocoria** → **you have to exclude Retinoblastoma.**
- Majority unilateral, but still happens bilateral sometimes.
- If you're not sure of the diagnosis → Ultrasound.
- Treat with chemotherapy, radiation, laser, enucleation (removal of the eye) ...



Amblyopia

Also called **Lazy eye**

Decrease in visual acuity of one eye without the presence of an organic cause that explains that decrease in visual acuity.

Usually results from: Cortical ignorance of one eye. (Cortical = at the level of the brain)

2-4% of the general population.

The most common cause of visual loss under 20 years of life.

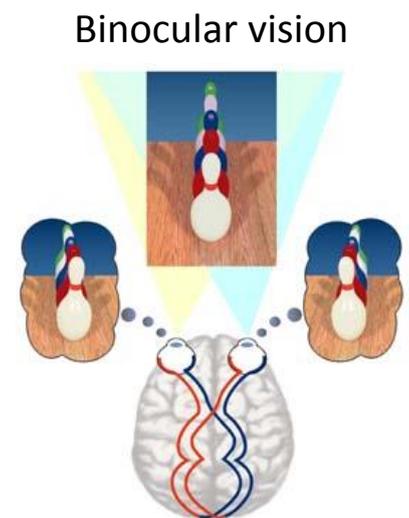
Amblyopia does not happen after 7 years of age.

As we said before:

For the brain to unify the 2 images → both should have the same shape, size and clarity.

What if images were totally different or grossly different?

Diplopia → Confusion → Suppression



Why fusion may fail (causes of amblyopia):

1. Significant anisometropia

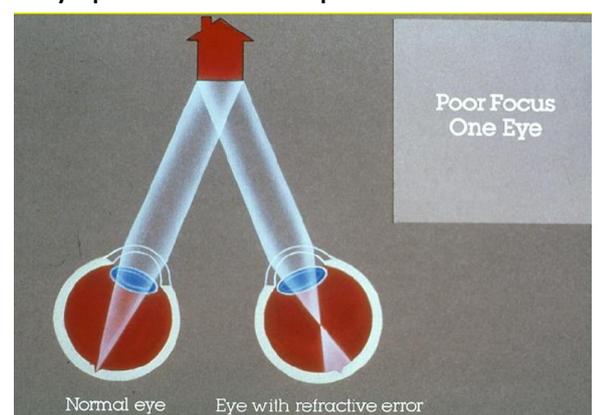
Mild hyperopic or astigmatic anisometropia (1-2D) --> mild amblyopia

Mild myopia anisometropia (less than -3D) usually doesn't cause amblyopia unilateral high myopia (-6D) --> severe amblyopia visual loss.

You don't have to remember the numbers, just know that anisometropia is more significant with hyperopia.

The eyes of a child with anisometric amblyopia look normally to the family and primary care physician.

Myopic anisometropia



Doctor's notes:

- Anisometropia: eyes have significantly different refractive errors. E.g. one eye has a refractive error of 1 and the other has 7 (difference = 6).
- Brain can't fuse the two images because of the difference in **clarity** → brain cancels the image from the unclear side. By doing this for a long time → suppression (amblyopia).
- We can only interfere and reverse this before 5 years of age. If child presents after that it's too late.
- Treatment:
 - We give the patient glasses for the refractive error.
 - Patch the good eye for a couple of hours every day to train the lazy eye.

2. Significant aniseikonia (difference in image size)

Team explanation: It's the same concept here, when you use glasses to correct eye refractive errors → the image size becomes smaller (myopia) or larger (hyperopia) → the brain receives images from the 2 eyes with different sizes → suppresses the abnormal one → amblyopia

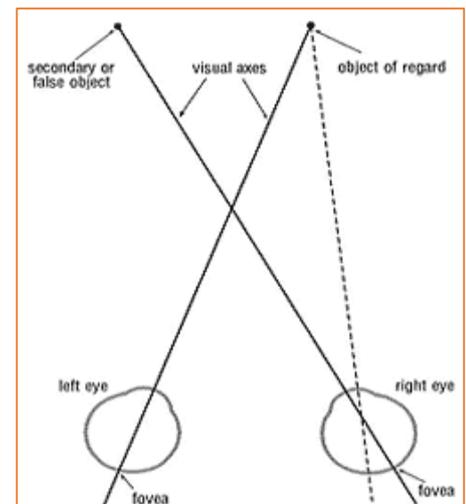
Doctor's notes:

- Imagine a situation in which we removed a child's lens due to cataract → we couldn't implant an intraocular lens because he was too young → we gave him glasses with +12D or +13D of power → image from the diseased eye will be clear now, but very big compared to the small image from the other eye → will be suppressed → That's why in this case we should use contact lenses instead (less difference in image size)

3. Strabismus



When the eye is squinted, the image will not fall on the fovea. The dashed line (on the image to the right) shows how the image of the object of regard is not falling on the fovea, while the image falling on the fovea is from another false point → blurred image in the squinting eye → squinted eye suppressed (amblyopia)



Diplopia charting, n.d. photograph, viewed 20 September 2014, <<http://www.answeringgenesis.org/assets/images/articles/tj/v16/i3/p111.gif>>.

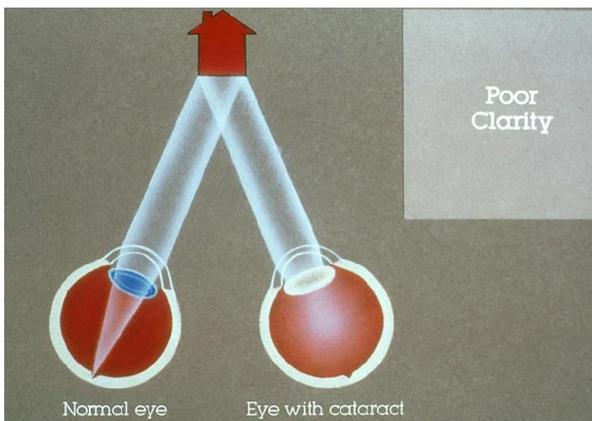
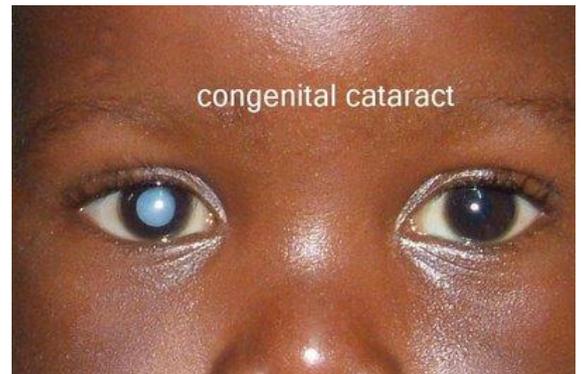
Doctor's notes:

- Eye is not straight.
- In the abnormal eye the image of interest will not fall on the fovea → blurred image to the brain → brain suppresses the blurred image → amblyopia.
- **Don't tell the parents that when the child grows up it'll go because it won't.**
- If left untreated till 5 years of age → irreversible amblyopia.
- Sometimes hyperopia may cause a squint (strabismus), we call this "accommodative esotropia" → treat using glasses → eye becomes straight and we should patch the good eye for certain hours to reverse the maluse of the amblyopic eye.
- If refractive error is not the cause → we should go for surgery → to make it straight again → image will fall on the fovea.

4. Deprivation amblyopia



- Cataracts (**congenital cataract**) and corneal opacities.
- Eyelid ptosis, eyelid hemangioma.
- Results in more severe visual impairment than strabismic or refractive amblyopia.
- Anything which prevents the light from going inside the eye is known as deprivation amblyopia or (occlusive amblyopia).
- If you leave any of the cases in the images untreated → they will develop deprivation amblyopia.



Types of amblyopia

In the lecture the doctor talked about amblyopia by cause, but in the slides it's also listed by type (it's almost the same)

1. Strabismic
2. Deprivation amblyopia
3. Anisometropic amblyopia (refractive amblyopia)
 - More common with hypermetropic anisometropia
 - Astigmatic = Meridional amblyopia

Treatment:

- Treat the cause of amblyopia: correct the refractive error, remove the media opacity surgically.
- The **younger the child, the better outcome** of amblyopia therapy.
- The first five years of child age is the sensitive period (we call it the golden period) where amblyopia can be reversed, after that it become more difficult.
- **Again don't tell the parents that when the child grows up it'll go because it won't.**
- For operating on congenital cataract:
 - Unilateral → more urgent → operate in the first month of life
 - Bilateral → OK to make it 2 months



Occlusion therapy

- It's the mainstay of treatment.
- Patch the good eye after removing the cause.

Strabismus

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 are we concerned about strabismus?

Of course we care about cosmetic issues, but the **most important thing is that it may lead to amblyopia.**



In this image we see the corneal reflex. On the left eye it's straight on the pupil, while on the right eye the light is deviated temporally → pupil deviated nasally. We call this type of squint → "esotropia".



In this image we can see "exotropia". In adult cases it's usually due to neurological causes, in this case **3rd nerve palsy.**

We have 2 types of strabismus:

1. Esotropia (towards the nose) (the commonest type)
2. Exotropia (temporally)

1. Esotropia:

- a. Infantile esotropia
- b. Acquired esotropia

Slides also say

Eye deviations known as strabismus are divided into two main types:

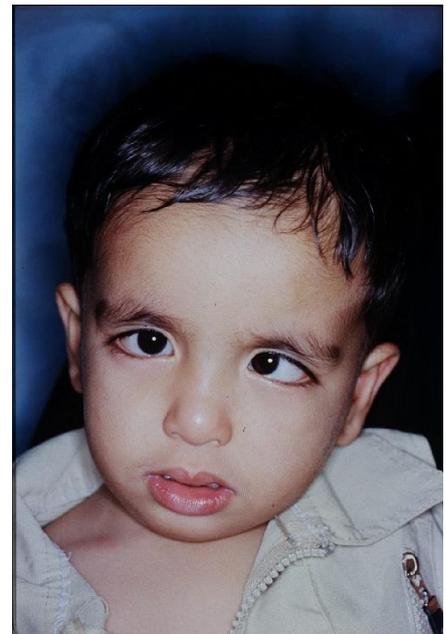
1. Comitant.
2. Noncomitant.

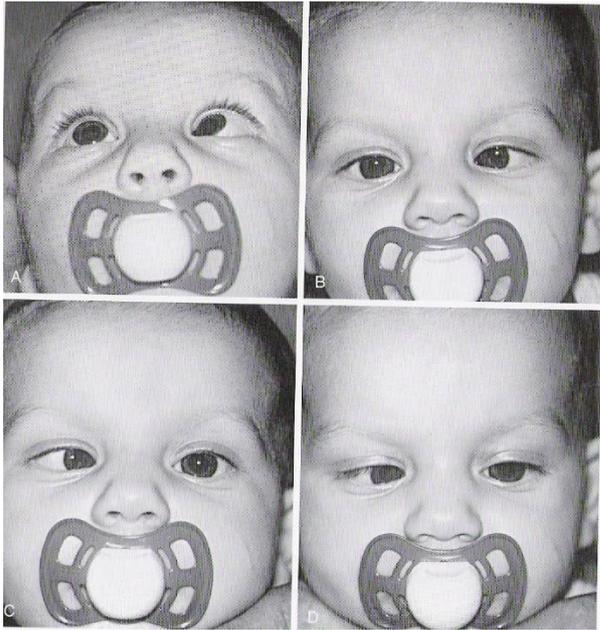
But doctor didn't mention them.

Infantile esotropia

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. In infantile esotropia we don't know the cause.

- 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, and central scotomas.





In the image on the left the child has "alternating esotropia" → no amblyopia because the suppression is alternating.

A and B → esotropia in the left eye.

C and D → esotropia in the right eye.

Always squinting in the right eye → will become amblyopic.

Management:

- Treat the amblyopia by occluding the good eye.
- Surgery for the extra ocular muscles.

We go to the medial rectus → make it weaker

We go to the lateral rectus → make it stronger

Muscle recession procedure: we move the insertion of the muscle → muscle longer + weaker.

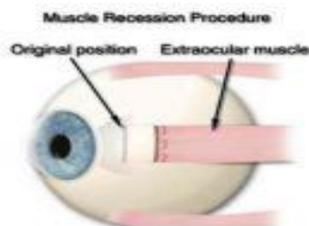
We should follow up with this patient to treat her amblyopia.



Before surgery



After surgery



Mom was told to place a patch 3hrs/day, but she misunderstood and placed a new patch every 3 hrs → too much occlusion → occlusive amblyopia → patch the other eye.

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.



Doctors notes:

- Asians might look like having strabismus but they don't.
- Pseudostrabismus can be proven by:
 - Light falling on the pupil (just like in the image, if there was strabismus it wouldn't fall on the center)
 - **Cover test** (the most reliable test to prove strabismus): e.g. somebody has a squint in the left eye → cover the right eye:
 - If the left eye moves to the middle → strabismus
 - If the left eye doesn't move → no strabismus (pseudostrabismus in this case)

Accommodative esotropia:

- **Acquired** (acquired esotropia) after 6 months of age usually around 2 years.
- Associated with **hyperopia**.
- Refractive error correction by **glasses** will treat the condition.
- Perform cycloplegic refraction on all children by using the retinoscope and loose lenses. Cycloplegia is achieved with Mydracyl 1% and cyclogel 1%. (doctor never mentioned anything in this point).
- Refractive error usually +3_+4
- May be precipitated by acute illness or trauma
- Start intermittent and if not treated become constant
- This child has hyperopia → image behind the retina → child accommodates (to correct the image) → as a consequence he'll also converge → squint.



- Commonest type of esotropia.

Do we do surgery for Acc ET ?

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



Exodeviation:

- Exodeviation is a horizontal form of strabismus characterized by visual axes that form a divergent angle.
- Not as common as esotropia.
- Starts gradually (intermittent) → doesn't cause amblyopia.
- Has to be treated with surgery.



Summary

- Leukocoria: white opacity of the pupil.
- If you don't treat congenital cataract early → irreversible amblyopia.
- Persistent hyperplastic primary vitreous (PHPV): failure of the normal regression of the primary vitreous, mostly unilateral, scar → leukocoria.
- Organized vitreous hemorrhage: Vitreous hemorrhage (may be due to birth trauma), yellowish appearance, vitrectomy is usually required.
- Retinal detachment: Due to HIGH MYOPIA → retinal tear → fluid leaks → detachment.
- Retinopathy of prematurity: in premature, low-birth-weight. Retina not fully vascularized at birth → new fragile vessels → fibrosis → detachment.
 - Kill this retina (laser) → save what's left (preserve macula)
- Coloboma: incomplete closure of the fetal fissure, "cat eye" or "keyhole" appearance.
- Medullated nerve fibers: myelination of the retinal nerve fibers. Looks "like cat fur"
- Coat's Disease: retinal telangiectasia and aneurysms --> exudative retinal detachments.
- Retinoblastoma is a life-threatening disease → has to be ruled out. Mostly unilateral.
- Anisometropia: significantly different refractive errors → amblyopia. Give glasses + patch.
- Don't reassure the parents in case of strabismus.
- Strabismus is important because it may lead to amblyopia.
- The younger the child, the better outcome of amblyopia therapy.
- Esotropia (medial deviation) is the most common type of strabismus.
- Accommodative esotropia is the most common type of esotropia. Usually acquired after 6 months of age. No surgery.
- Alternating esotropia causes no amblyopia because the suppression is alternating.
- Treat infantile esotropia by occluding the good eye + surgery for the extra ocular muscles.
- Exodeviation starts gradually --> doesn't cause amblyopia. Needs surgery.

One last point regarding why the doctor said we have to fix congenital problems obstructing the view as early as possible, while in anisometropia he said before 5 years. I found these two points which might explain:

- From the American Academy of Ophthalmology: “When the visual axis is blocked by a lens opacity during the sensitive period of visual development, irreversible amblyopia and permanent nystagmus may result. **The first two months of life** are the most critical for visual development; amblyopia resulting from visual deprivation **after the age of 2 to 3 months can often be reversible to some degree**. Visual development continues until at least 7 years of age.”
- “For example, anisometropia does not have much affect on acuity **unless it is persistent for 3 years**; ie, a short period of anisometropia after birth does not lead to acuity deficits.”¹

¹ Daw NW. Critical Periods and Amblyopia. Arch Ophthalmol. 1998;116(4):502-505. doi:10.1001/archophth.116.4.502.