Strabismus, Amblyopia & Leukocoria

Saeed Alwadani, MD Assistant Professor Consultant Ophthalmologist Ophthalmology department King Saud University

What is Strabismus ?

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

According to fusion status

1. Phoria

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

2. Intermittent Phoria

Fusion control is present part of the time

3. Tropia

Manifest misalignment of the eye all the time

According to fixation

 Alternating Spontaneous alternation of fixation from one eye to the other

Monocular
 Preference of fixation with one eye

According to type of deviation

1. Horizontal Esodeviation Exodeviation

2. Vertical Hyperdeviation Hypodeviation

3. Torsional Incyclodeviation Excyclodeviation

4. Combined

According to age of onset

- 1. Congenital
- 2. Acquired

According to variation of the deviation with gaze position or fixing eye

1. Comitant

Same deviation in different direction of gaze

2. Incomitant

Variable deviation in different direction of gaze usually in paralytic or restrictive type of strabismus

- 1. History
- 2. Inspection
- 3. Assessment of monocular eye function

Visual acuity

Preverbal children

CSM OKN Preferential looking Visual evoked potential

Assessment of monocular eye function

70

60

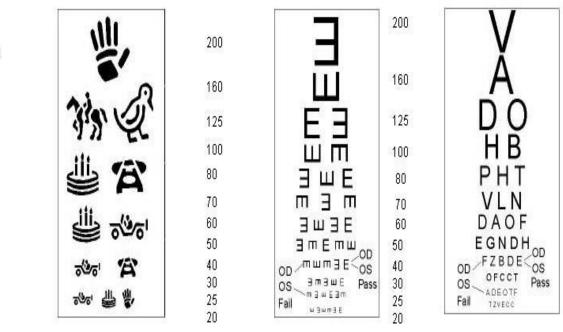
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Visual acuity

Verbal children

Symbol tests Single illiterate E Allen pictures H O T V letters



Assessment of binocular eye function

- 1. Hirschberg test
- 2. Krimski's test(measuring)
- 3. Cover test
- 4. Alternate cover test
- 5. Prism cover test



Fundoscopy

Cycloplegic refraction

- Tropicamide
- Cyclopentolate
- Atropin

Type of Strabismus

Esotropia

- Pseudoesotopia
- Infantile esotropia
- Accommodative esotropia
- Partially accommodative esotropia

Pseudoesotropia

- Occur in patients with flat broad nasal bridge and prominent epicanthal fold
- 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 and inferior oblique over action
- Treatment

Correction of refractive error Treat amblyopia Surgical correction of strabismus



Accommodative Esotropia

- Occur around 2 ¹/₂ years of age
- Start as intermittent then become constant
- High hypermetropia
- Treatment

Full cycloplegic correction Treat amblyopia



Partially Accommodative Esotropia

- Improve partially with glasses
- Treatment

Full cycloplegic correction Treat amblyopia Surgical correction of strabismus



Type of Strabismus

Exotropia

- Intermittent exotropia
- Constant exotropia
- Sensory exotropia

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

Correction of any refractive error Surgical correction of strabismus



Constant exotropia

- Maybe present at birth or maybe progress from intermittent exotropia
- Treatment

Correction of any refractive error Correction of amblyopia Surgical correction of strabismus



Sensory exotropia

- Constant exotropia that occur following loss of vision in one eye e.g trauma
- Treatment

Correction of any organic lesion of the eye Correction of amblyopia Surgical correction of strabismus

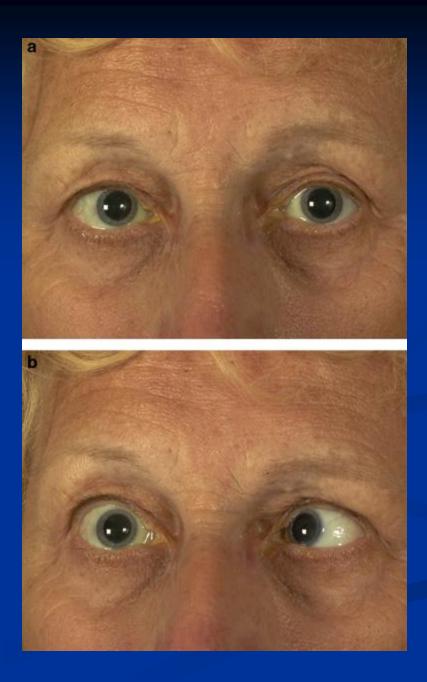
Types of Strabismus

Paralytic strabismus

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

6th Nerve Palsy

- Incomitant esotropia
- Limitation of abduction
- Abnormal head position

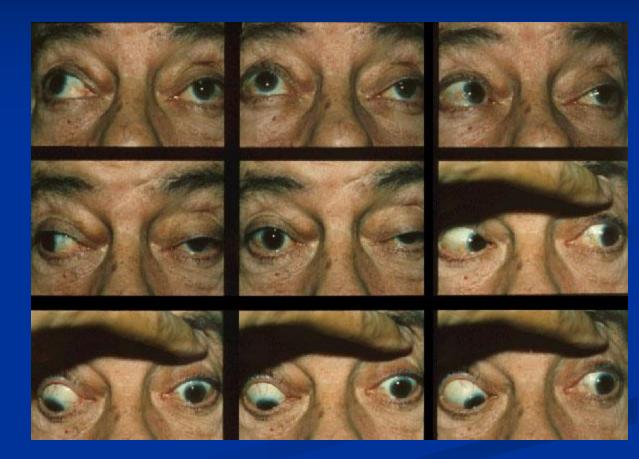


4th Nerve Palsy

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

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, diabetes, neuritis, trauma, infection and tumor



Special Types of Strabismus

- Duane strabismus
- Brown syndrome
- Thyroid opthalmopathy

Duane Syndrome

- Limitation of abduction
- Mild limitation of adduction
- Retraction of the globe and narrowing of the palpebral fissure on adduction
- Upshoot or downshoot on adduction
- Pathology faulty innervation of the lateral rectus muscle by fibers from medial rectus leading to cocontraction of the medial rectus and lateral rectus muscles



Brown Syndrome

- Limitation of elevation on adduction
- Restriction of the sheath of the superior oblique tendon
- Treatment needed in abnormal head position or vertical deviation in primary position

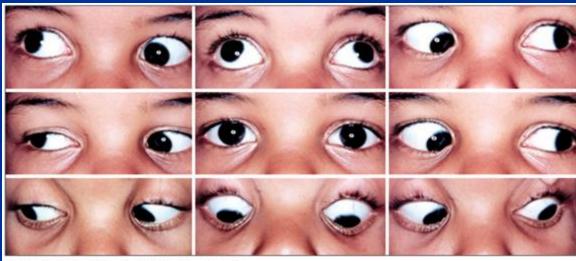


Figure 1 - The nine positions of gaze, boy

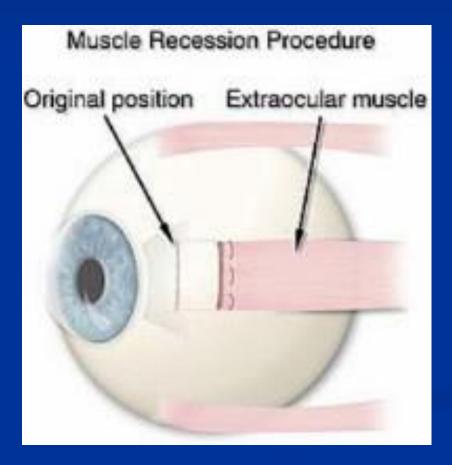
Thyroid Ophthalmopathy

- Restrictive myopathy commonly involving inferior rectus, medial rectus and superior rectus
- Patients presents with hypotropia, esotropia or both



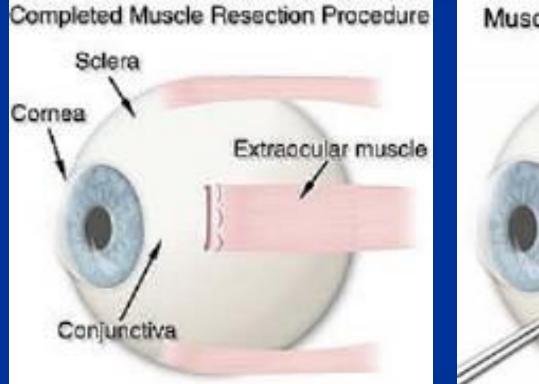
Surgery of Extraocular Muscle

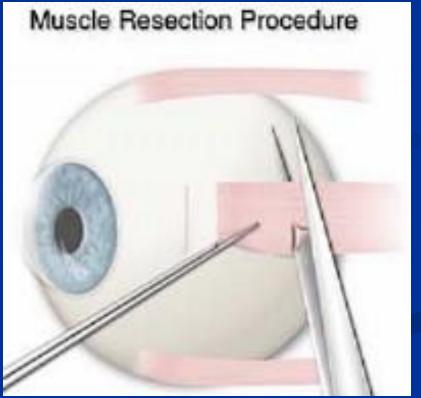
• Recession : weakening procedure where the muscle disinserted and sutured posterior to its normal insertion



Surgery of Extraocular Muscle

• Resection : strengthening procedure where part of the muscle resected and sutured to its normal insertion





Complication of Extraocular Muscle Surgery

- Perforation of sclera
- Lost or slipped muscle
- Infection
- Anterior segment anesthesia
- Post operative diplopia
- Congectival granuloma and cyst

Amblyopia

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

Amblyopia

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

Amblyopia

Diagnosis of amblyopia usually requires a 2-line difference of visual acuity between the eyes

Causes of Amblyopia

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

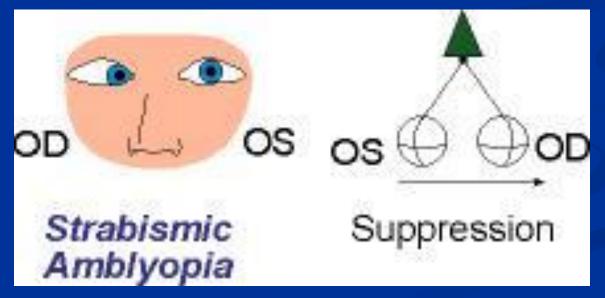
Anisometropia

- 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.

Strabismus

The patient favors fixation strongly with one eye and does not alternate fixation. This leads to inhibition of visual input to the retinocortical pathways.

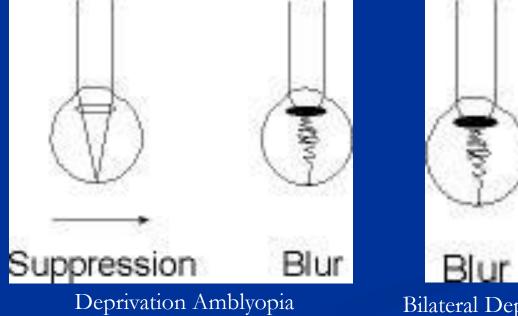
Incidence of amblyopia is greater in esotropic patients than in exotropic patients

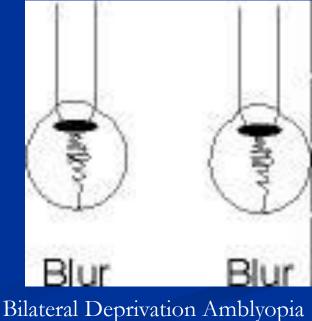


Alternation with alternate suppression avoids amblyopia

Visual deprivation

Amblyopia results from disuse or understimulation of the retina. This condition may be unilateral or bilateral. Examples include <u>cataract</u>, corneal opacities, <u>ptosis</u>, and surgical lid closure



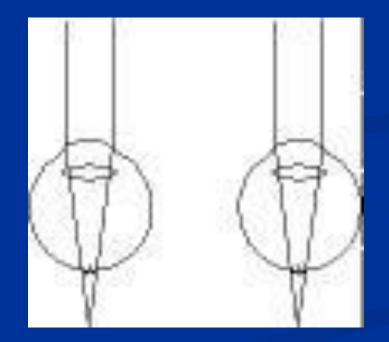


Organic

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

Ametropic Amblyopia

Uncorrected high hyperopia is an example of this bilateral amblyopia.



SCREENING: IMPORTANCE

- Amblyopia is usually preventable or treatable
- Early detection is key to effective treatment
- Life-threatening disorders may present as amblyopia
- Screening responsibility rests with primary care physician

AMBLYOPIA: EARLY DETECTION

Assess red reflex
Determine visual acuity
Evaluate ocular alignment



Direct ophthalmoscope



Direct ophthalmoscope: assessing red reflex



Direct ophthalmoscope: examining retina



Normal red reflex



Asymetric red reflex

Treatment

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



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.

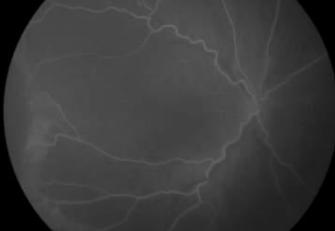
Anatomic location is important in the differential diagnosis of Leukocoria

Causes of Leukocoria

- Cataract
- Retinoblastoma
- Toxocariasis
- Coat´s disease
- **ROP**
- PHPV
- Retinal detachment
- Coloboma
- Retinal dysplasia
- Norrie´s disease







1- History

- 2- Complete ocular examination
- 3- B Scan ultrasonoghraphy
- 4- Intravenous fluorescein angiogram (coats disease, ROP, retinoblastoma)
- 5- CT or MRI
- **6- Serum ELISA**

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.

•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



RETINOBLASTOMA

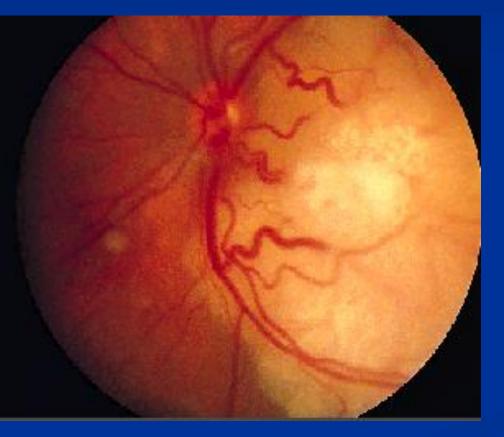
 Retinoblastoma is the most common intraocular tumor of childhood.



CLINICAL
MANIFESTATIONS
Leukocoria (60%)
Strabismus (20%)

 OTHER- Uveitis, Orbital cellulitis, Hyphaema, Heterochromia, Glaucoma, Bupthalmos

RETINOBLASTOMA





Retinoblastoma



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.

MANAGEMENT

EMPIRICAL GENETIC COUNSELLING

- ENUCLEATION
- 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</p>
- 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.

RETINOPATHY OF PREMATURITY (ROP)

Vasoproliferative retinopathy affecting premature infants

exposed to high oxygen

INCIDENCE

Prematurity (<32/40)</p>

Birth weight (30% < 1000gm affected)

Oxygen duration

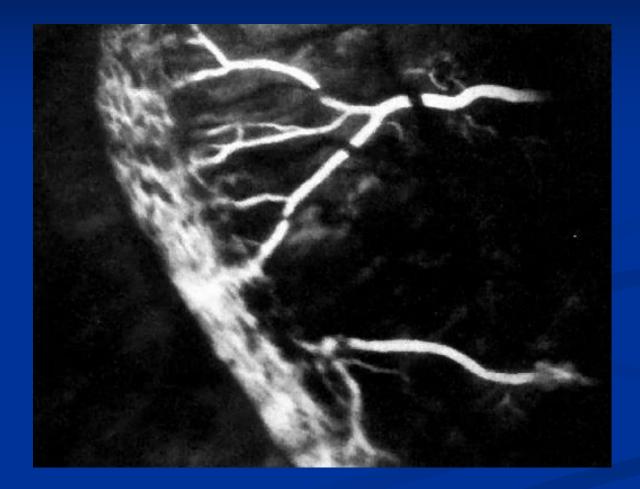
90% ROP regresses spontaneously, 5% blindness

RETINOPATHY OF PREMATURITY (ROP)



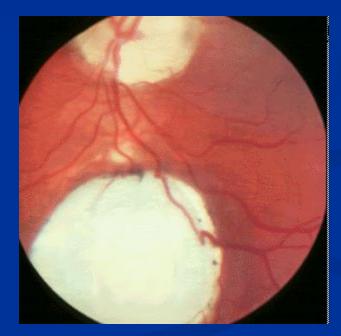
Signs include:

- neovascularization,
- fibrous bands
- retinal detachments
- vitreous hemorrhage
- leukocoria



COLOBOMA

- Optic Disc Coloboma
- Due to failure of closure of foetal fissure inferiorly
- May be isolated disc or associated chorioretinal coloboma
- Usually sporadic, some AD
- Can be bilateral
- Visual acuity varies from normal to NPL.



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