Strabismus for 5th yr medical students

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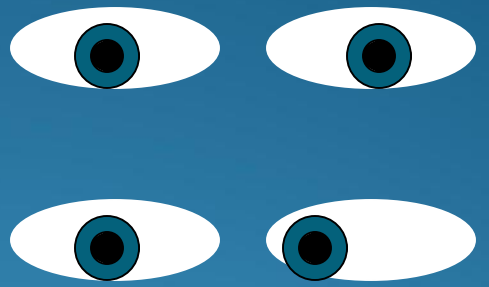
New words to Encounter

- Strabismus = heterotropias
- Esotropia = turn inward
- Exotropia= turn outward
- Hypertropia= turn upward
- Hypotropia = turn downward
- Amblyopia = Lazy eye (vision deficiency in an eye when the brain turns off the visual processing of one eye.
- Anisometropia= unequal refractive errors between the 2 eyes
- Diplopia = Double vision
- Monocular diplopia = diplopia persists when one eye is closed.
- Binocular diplopia= diplopia seen only when both eyes are open

Nomenclature

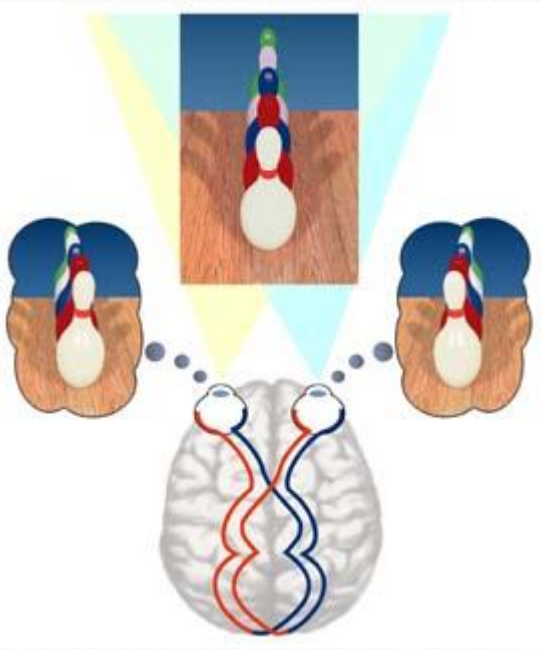
- Orthorphoria ϕ
- Esophoria E
- Esotropia ET
- Intermittent Esotropia E(T)
- At near X(T)'
- Exophoria X
- Exotropia XT
- Intermittent Exotropia X(T)
- Right Hypertropia RHT
- left Hypotropia LHoT



convergent

divergent

- Binocular single vision: slightly dissimilar images from both retinas are fused centrally to be interpreted by the brain as a single image.



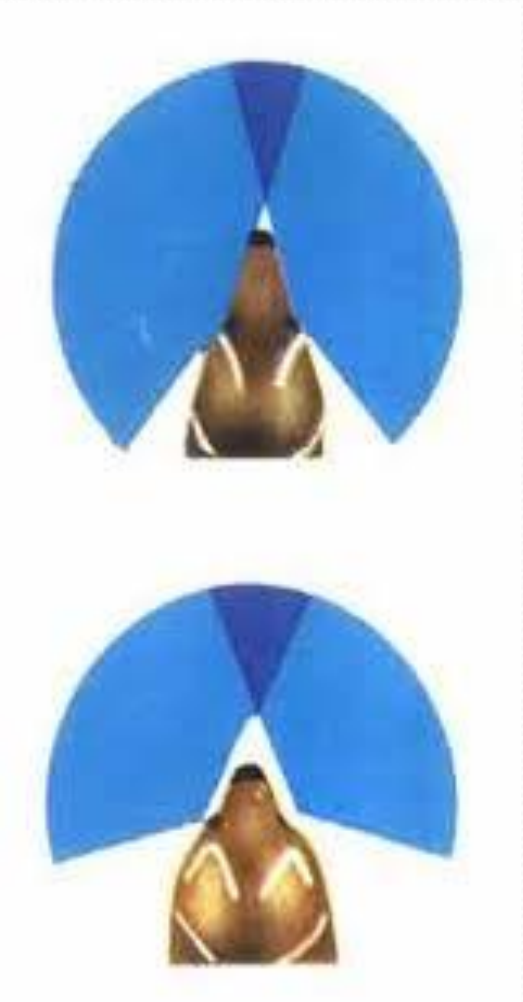
- Stereopsis: the construction of a 3D percept to the retinal images which have been taken from different angles.

- Who needs Stereopsis?

- PLEASE EXAMINE YOURSELF IN THE CLINIC

Importance of Stereopsis and Binocular single vision

- Increase field of vision
- Eliminate the blind spot since the blind spot of an eye fall on the opposite eye's visual field.
- Binocular acuity is greater than monocular
- Depth perception
- Estimation of Distance



- Normal movement of the eye (6 extraocular muscles)
- Binocular eye movements are called Versions
- Monocular eye movements with the other eye covered are called Ductions

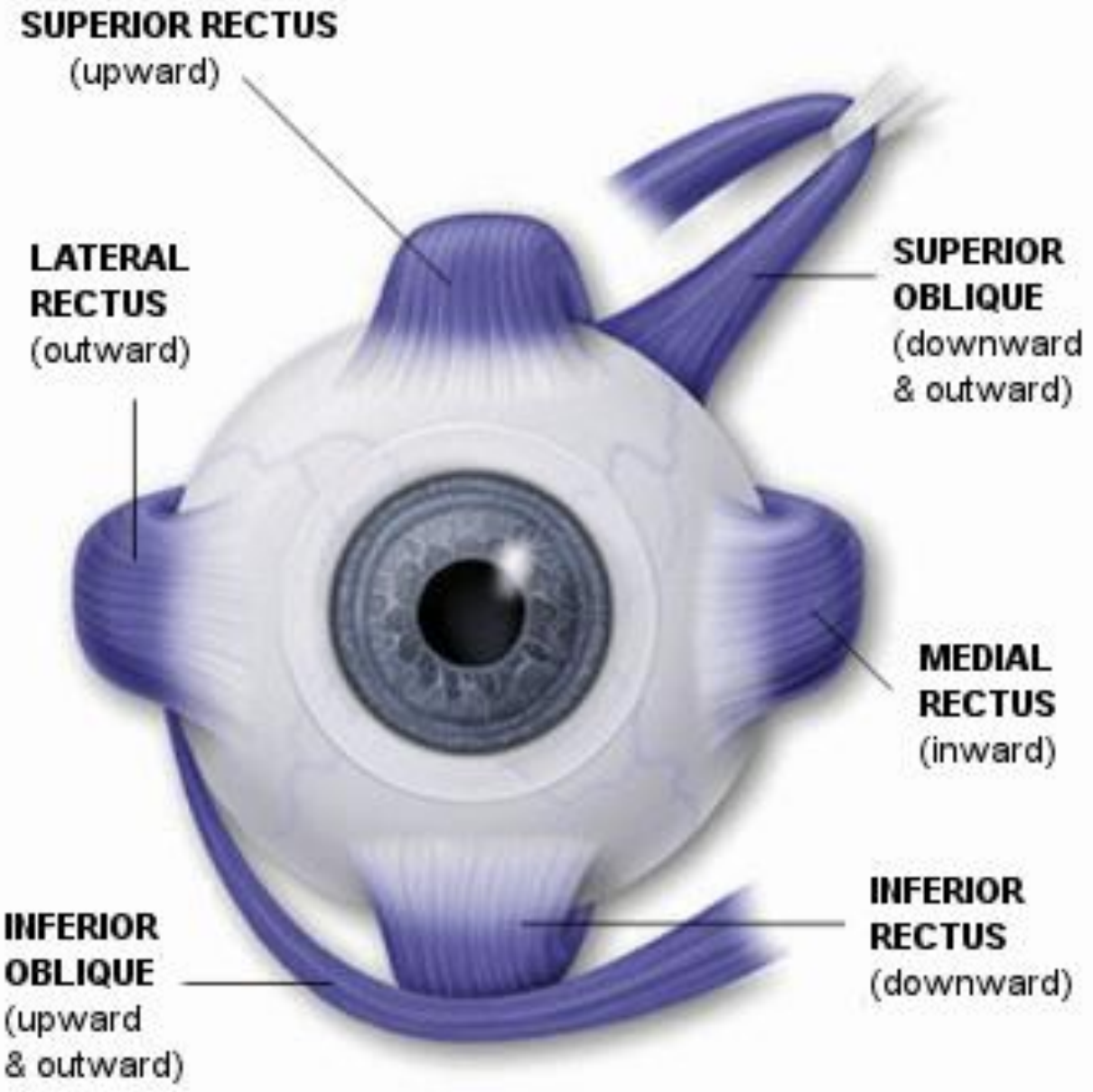
Nerve supply

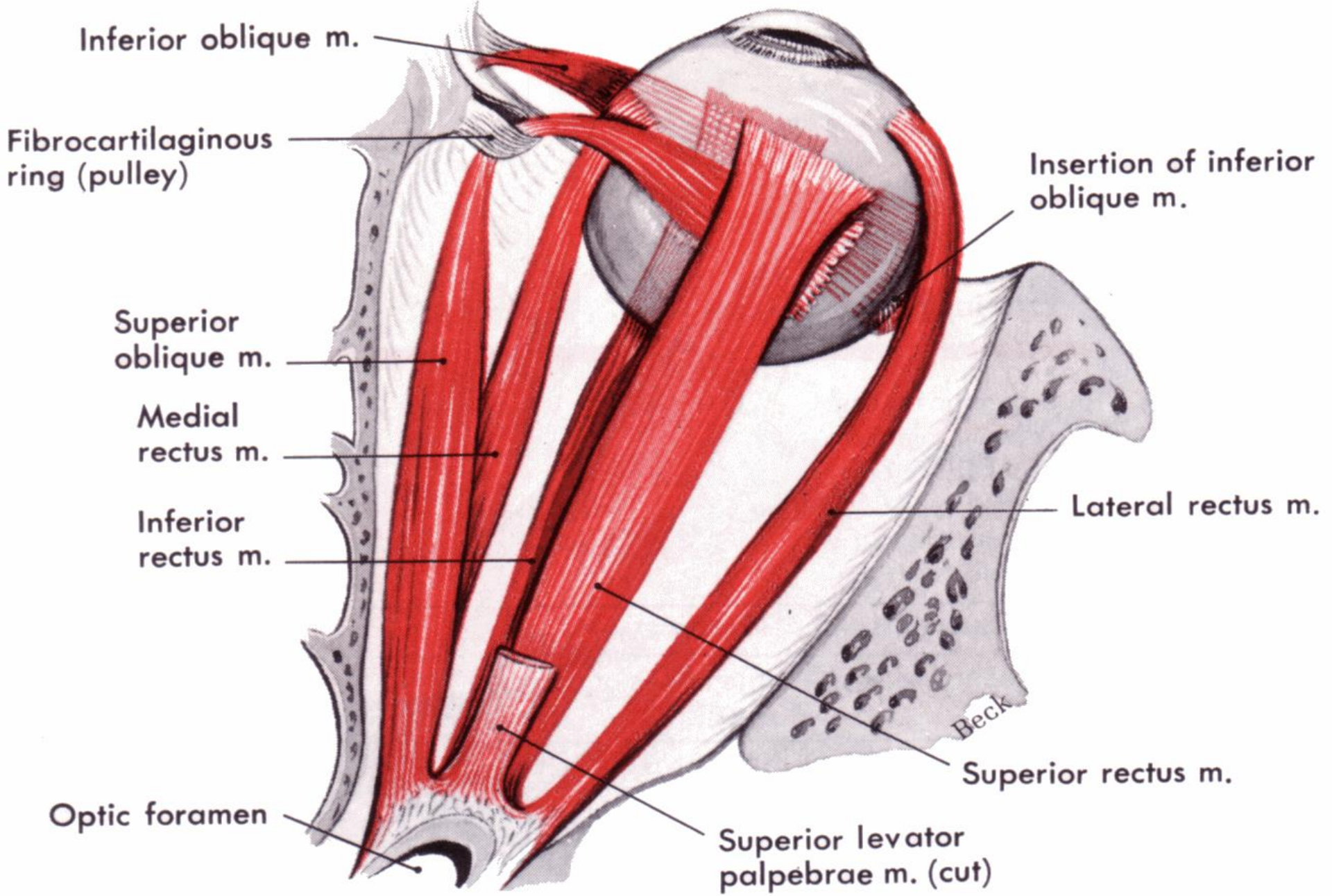
Third CN : MR, IR, SR, IO

Fourth CN : Superior Oblique

Sixth CN : Lateral Rectus

Eye movement





Muscles that move the right eye as viewed from above.

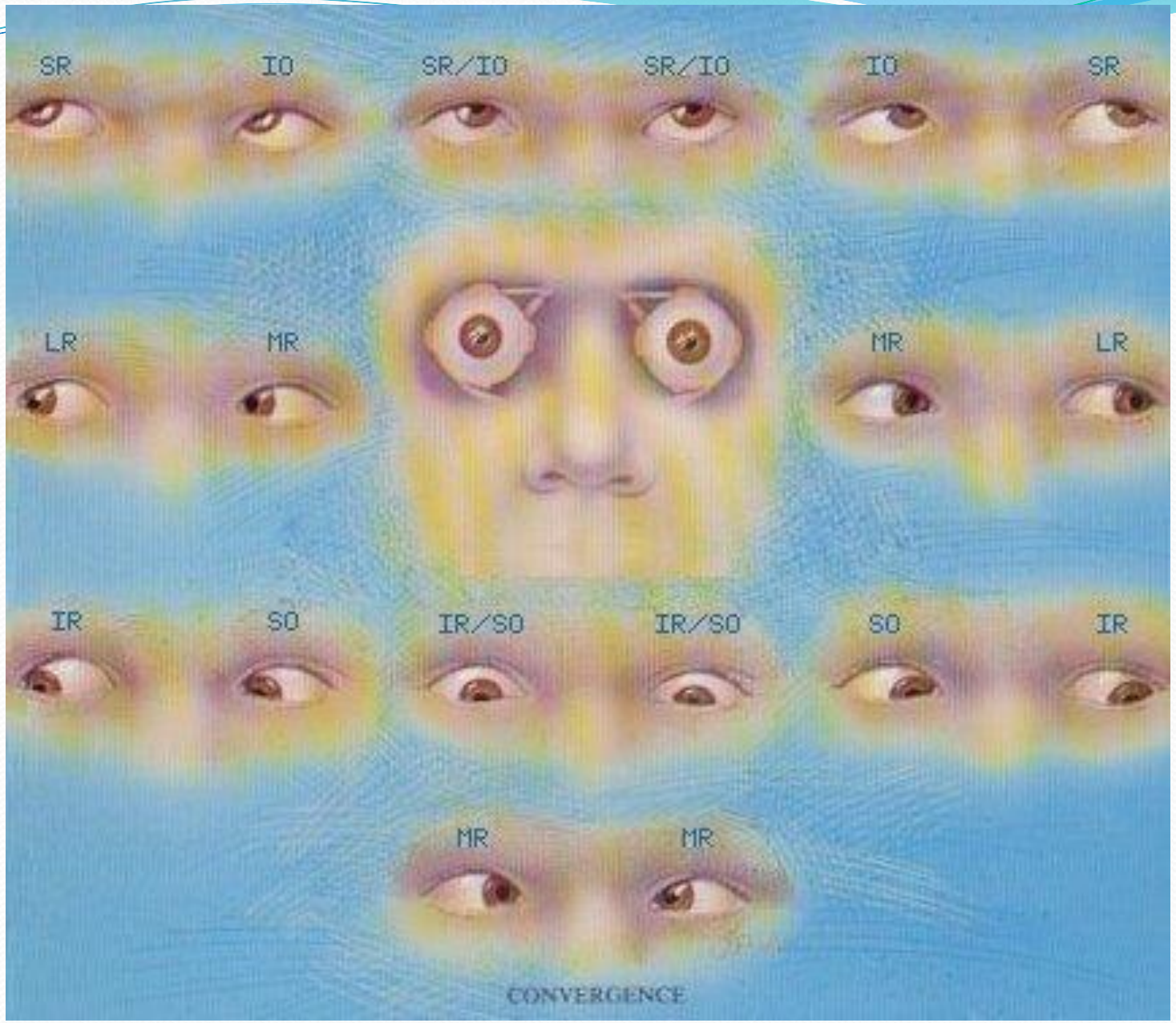
- These six positions of gaze are called the cardinal positions of gaze.
- In addition to these, there are another 3 position of gaze :

  the primary position – looking straight ahead

  Looking up

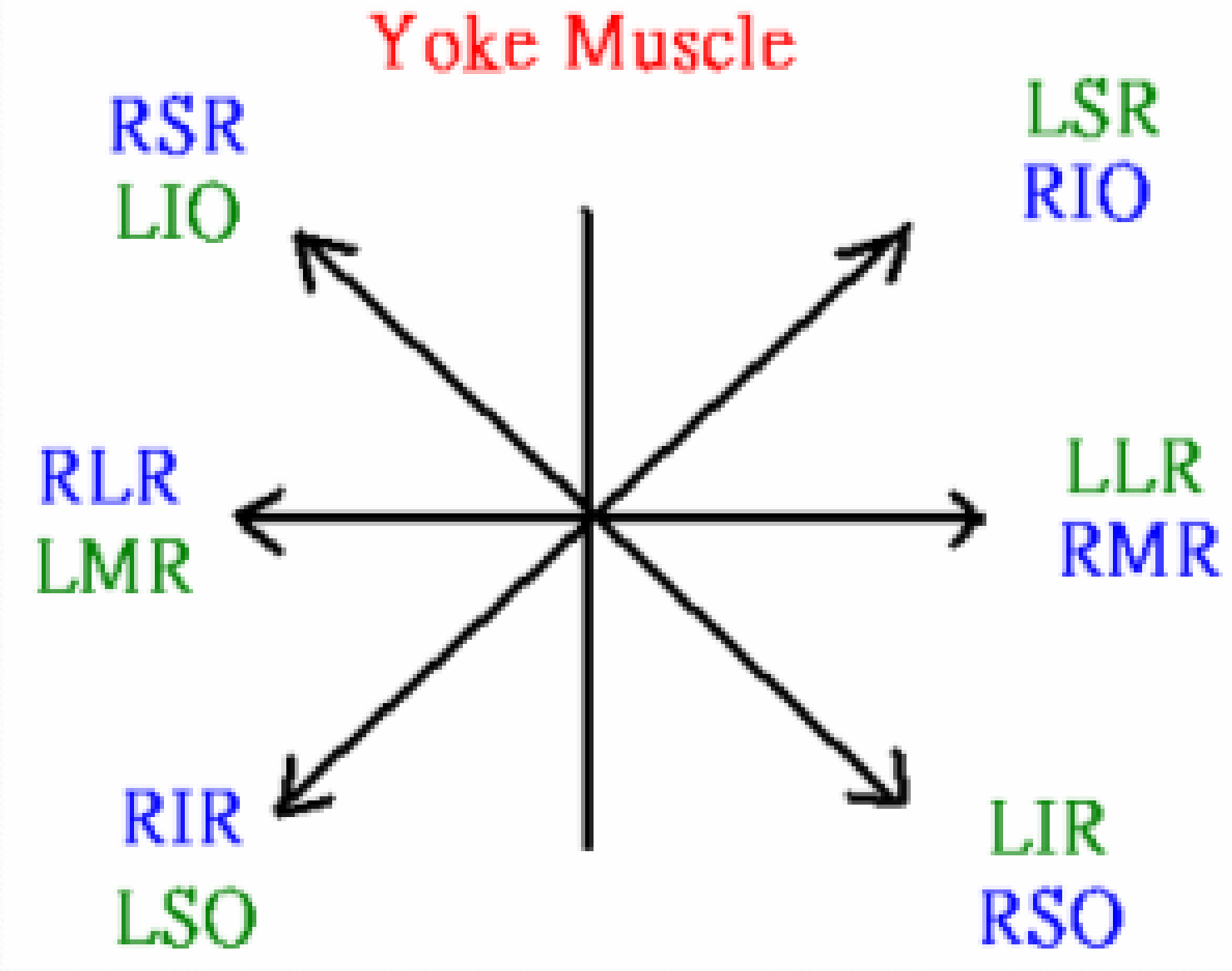
  Looking down

  Therefore the total number of the positions of gaze is 9


SR
IO
SR/IO
SR/IO
IO
SR
LR
MR
MR
LR
IR
SO
IR/SO
IR/SO
SO
IR
MR
MR
CONVERGENCE

- **Yoke muscles** are pair of muscles (one muscle in each eye) moving the eye into the same direction of gaze

- Rt lateral rectus & Lt medial rectus = to the right
- Lt lateral rectus & Rt medial rectus = to the left
- Rt superior rectus & Lt inferior oblique = to the right & up
- Rt inferior rectus & Lt superior oblique = to right & down
- Lt superior rectus & Rt inferior oblique = to left & up
- Lt inferior rectus & Rt superior oblique = to left & down


Yoke Muscle
RSR
LIO
LSR
RIO
RLR
LMR
LLR
RMR
RIR
LSO
LIR
RSO

- Evaluation of binocular eye movement
  ask the patient to follow your target in all positions of gaze

Under action of specific muscle could be :

- true paresis or paralysis
- restrictive myopathy
- underlying strabismus

What is squint (strabismus)?

- Squint is a misalignment of the two eyes so that both the eyes are not looking in the same direction.
- This misalignment may be constant, being present throughout the day, or it may appear sometimes and the rest of the time the eyes may be straight.(Intermittent)

  It is a common condition among children. It may also occur in adults.

Hirschberg Test

Eye movement disorders:

1- Concomitant (non-paralytic)

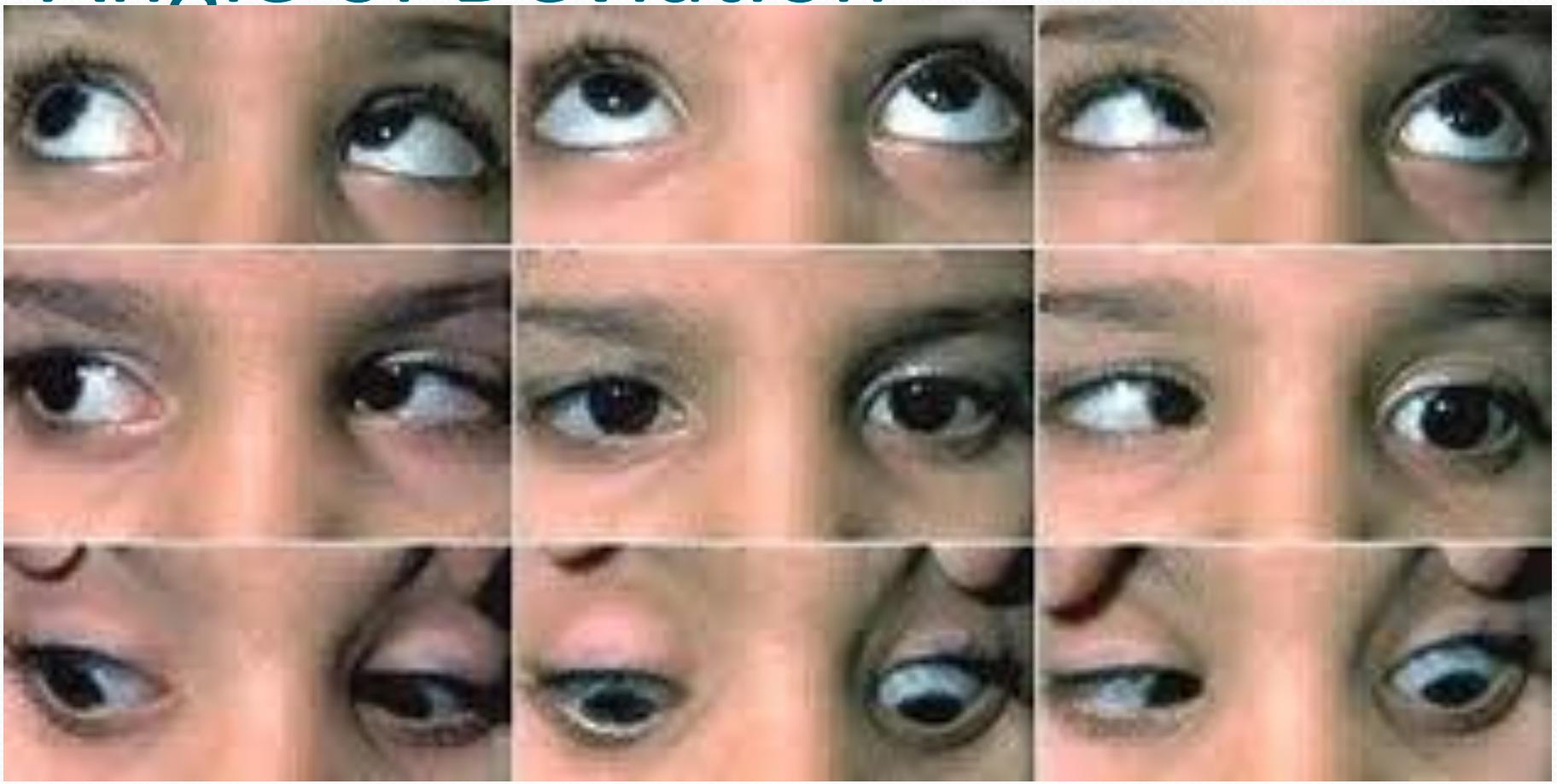
2- Incomitant (paralytic)

3- Gaze palsies

4- Disorders of the brainstem nuclei or vestibular input

5-…

Angle of Deviation



Concomitant (non-paralytic)

- Usually congenital
- both eyes have full movement if tested separately
- Diplopia is absent
- Only one eye is directed towards the fixated target .
- The angle of deviation is constant and unrelated to the direction of gaze .
- Extraocular muscles and nerves are grossly normal
- Most has its onset in childhood .

Aetiology of Concomitant squint

- .....
- Refractive error which prevents the formation of a clear image on the retina .
- Opacities in the media of eye blurring or preventing the formation of the retinal image .(i.e. : amblyopia)
- Abnormalities of the retina that prevent the translation of a correctly formed image into neural impulses .
- ...

Incomitant squint (paralytic)

- usually acquired .
- Diplopia is present (if occurs after the first 10 years of life). Diplopia is maximal when attempting to look in the direction requiring the action of the weak muscle.
- The degree of misalignment varies with direction of the gaze.
- One or more of the extra-ocular muscles or nerves may not be functioning properly , or normal movement may be restricted mechanically by tethering of the globe.
- This type of strabismus may indicate either a nerve palsy or an extra-ocular muscle disease .

Palsies

- 6th nerve: Failure of Abduction.
- 4th nerve: defective depression of the eye when in adduction.
- 3rd nerve: failure of adduction, elevation and depression of the eye, ptosis and in some cases dilated pupil.

Causes of isolated nerve palsies

- Vascular disease (DM, HTN, Aneurysm, CST)
- Orbital disease
- Trauma
- Neoplasia
- Raised intracranial pressure (3^{rd} or 6^{th} , False localizing)
- Inflammation (Sarcoidosis, Vasculitis, Infections, GBS)
- …

CST: Cavernous Sinus Thrombosis
GBS: Guillain-Barre Syndrome

Extraocular muscles disease

- Dysthyroid eye disease
- Myasthenia gravis
- Ocular myositis
- Ocular myopathy
- Browns Syndrome
- Duane's Syndrome
- …

Dysthyroid eye disease

- Due to infiltration of the extraocular muscles with lymphocytes and the depositions of glycosaminoglycans.
- Both Hyper and Hypo-Thyroidism.

Thyroid eye disease:Mechanical vertical and divergent squint

Dysthyroid eye disease

Symptoms & signs:

1. A red painful eye.
2. Diplopia.
3. ⇩ Visual acuity.
4. Exophthalmous.
5. Chemosis.
6. Lid retraction.
7. Lid lag.
8. Restricted eye movement/ squint.

- The inferior rectus is the most commonly affected.
- Mechanical limitation of the eye in up gaze.
- Involvement of the medial rectus→limitation of abduction. (DDx→6th nerve palsy)

- Complications:

1. Chemosis & corneal ulcers→corneal perforations.
2. Compressive optic neuropathy→blindness.

- Treatment:

1. Systemic steroids.
2. Radiotherapy.
3. Surgical orbital decompression.
4. Prisms.

Myasthenia Gravis

- Acetylcholine receptor targeted antibodies
- Females > males, 15-50 years of age
- 40% show involvement of Extraocular muscles only.
- Variable diplopia and ptosis due to fatigue.
- Diagnosis: Edrophonium test
- Treatment: neostigmine (acetylcholine esterase inhibitor), thymectomy.

Ocular myositis

- Inflammation of the extraocular muscles
- Pain, diplopia and restriction of movement.
- Systemic Disease, R/O thyroid disease.

Ocular Myopathy

- (Chronic) Progressive External Ophthalmoplegia(COPE)
- rare condition
- Mitochondrial DNA mutation
- Associated ptosis
- Movement of the eyes is slowly and symmetrically reduced
- Worst case, eye movement can be lost completely
- Pathology : ‘ragged red fibers’

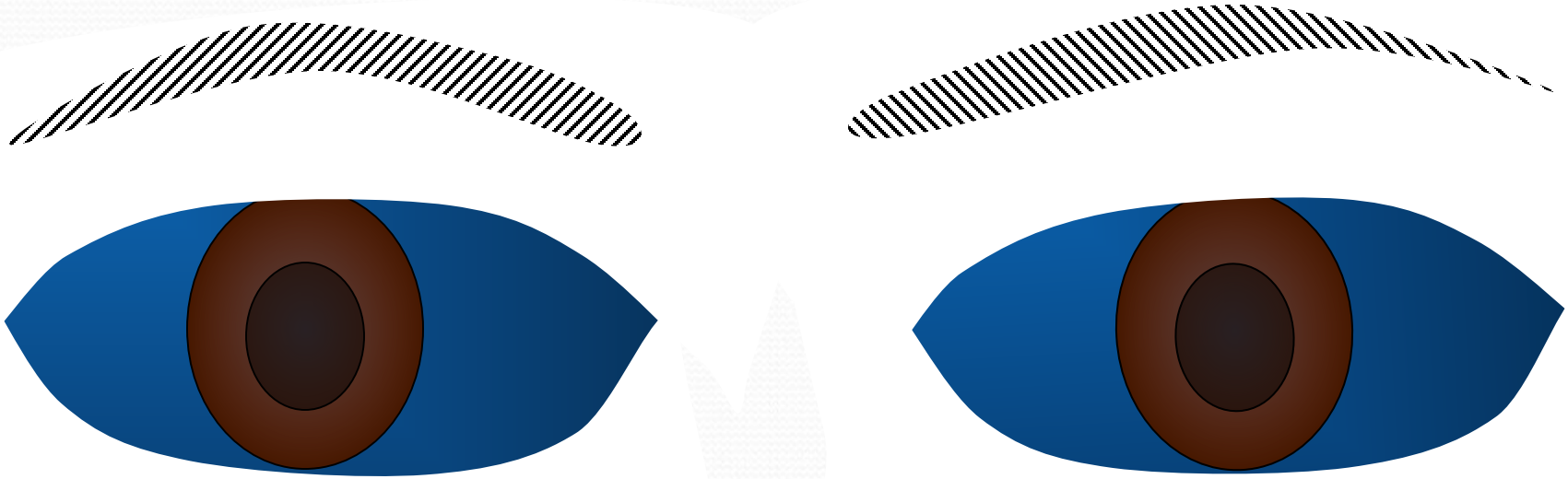
Brown's Syndrome

- 'superior oblique tendon sheath syndrome'
- Movement of IO muscle is restricted by the SO muscle tendon failing to pass smoothly through its trochlear pulley or a stiff inelastic tendon.
- Restriction of elevation in adduction
- Cause is unknown, maybe congenital or due to orbital trauma.

Duane's Syndrome

- Faulty innervation of the MR and LR muscles.
- 'Congenital Miswiring'
- LR works for ADDuction, MR works for ABDuction
- Children do not usually develop amblyopia because binocular alignment is normal is some gaze positions.
- Surgery is not often required.

Duane's Syndrome Type I



Gaze Palsies

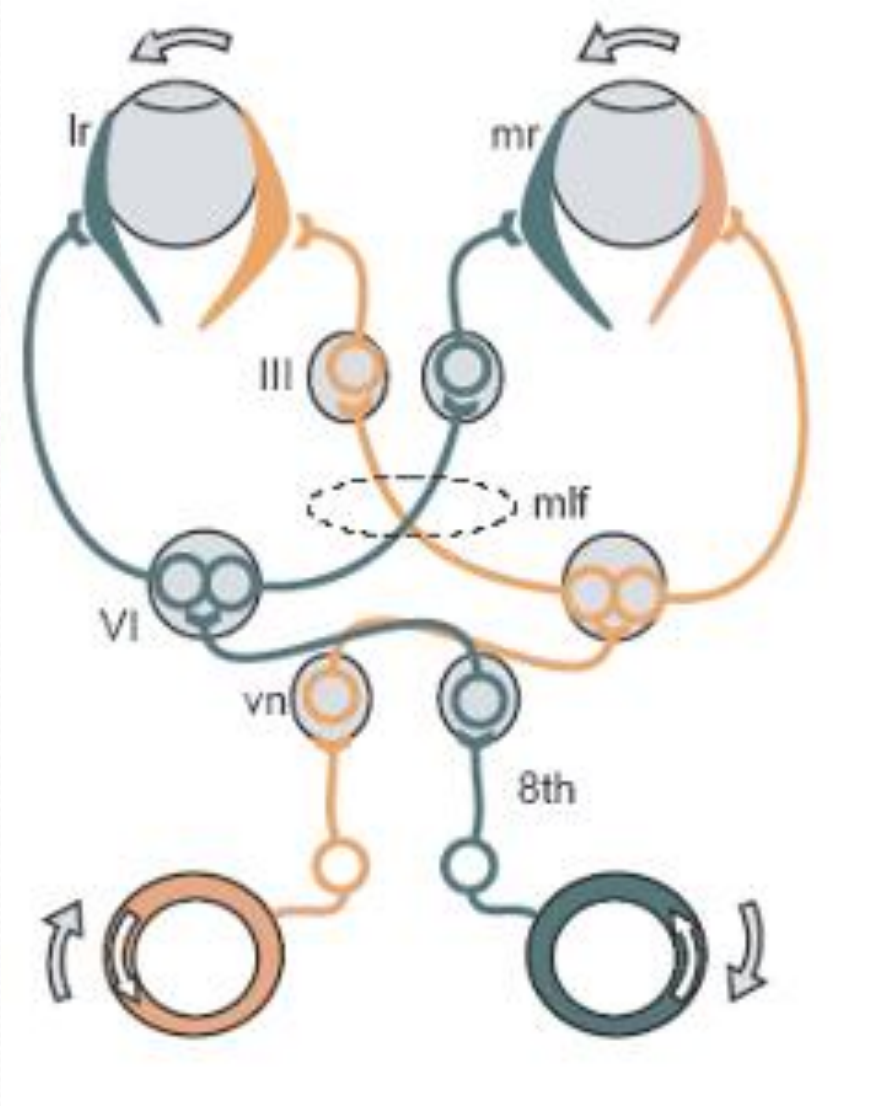
- 2 eyes acting in concert
- Connections between nuclei

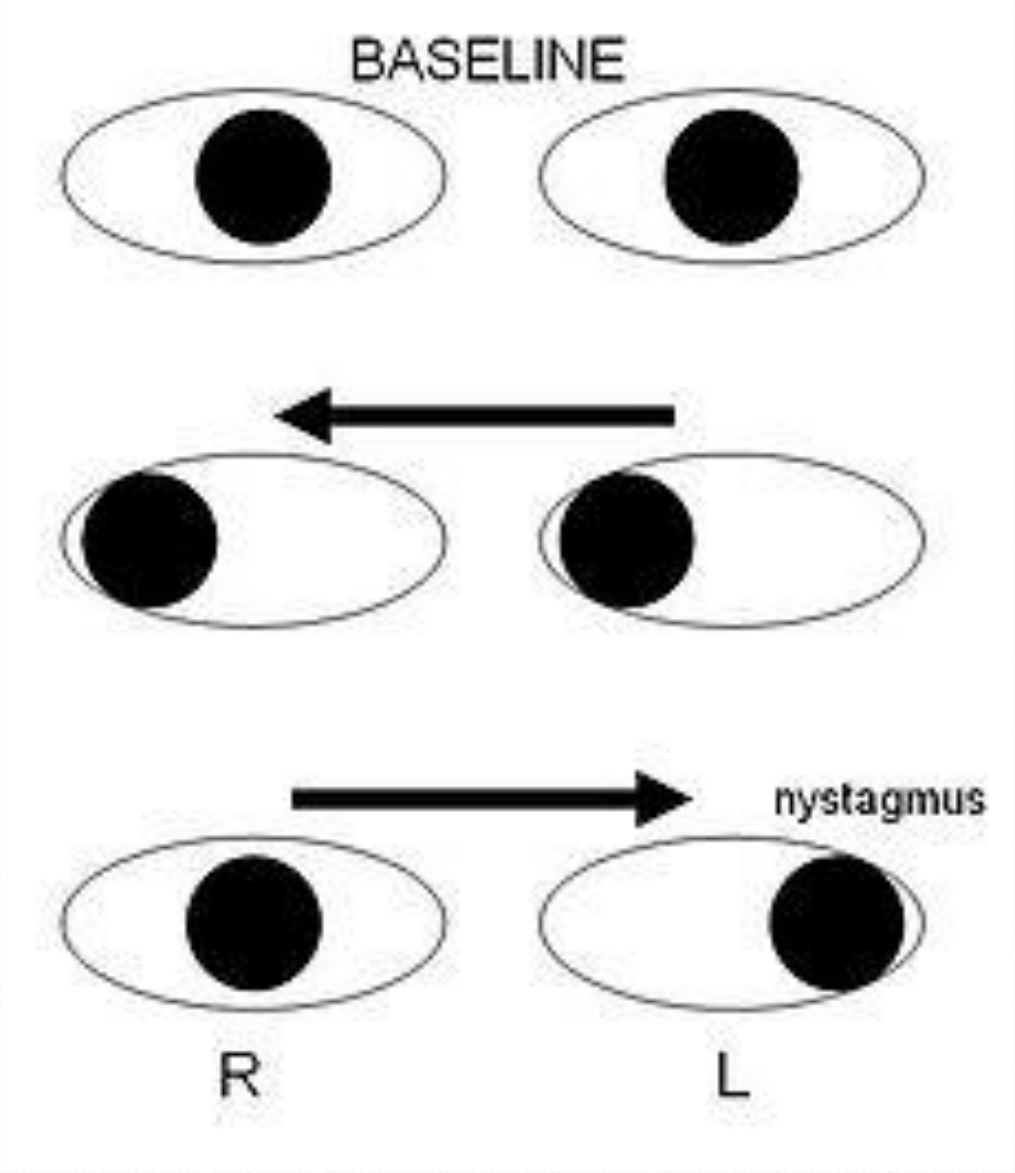
Parapontine Reticular Formation (PPRF)

- Controls the horizontal movements of the eyes.
- Occurs with other brainstem disease, vascular and tumours.
- Horizontal gaze palsy to the side of the lesion.

Internuclear ophthalmoplegia

- conjugate lateral gaze in which the affected eye shows impairment of adduction.
- if the right eye is affected the patient will "see double" when looking to the left
- divergence of the eyes leads to horizontal diplopia.
- Convergence is generally preserved.
- Injury to MLF (medial longitudinal fasciculus)


lr
mr
III
mlf
VI
vn
8th


BASELINE
nystagmus
R
L

Cover test

- A test to detect strabismus; the patient's attention is directed to a small fixation object, one eye is covered and after a few seconds, uncovered; if the uncovered eye moves to see the picture, strabismus is present

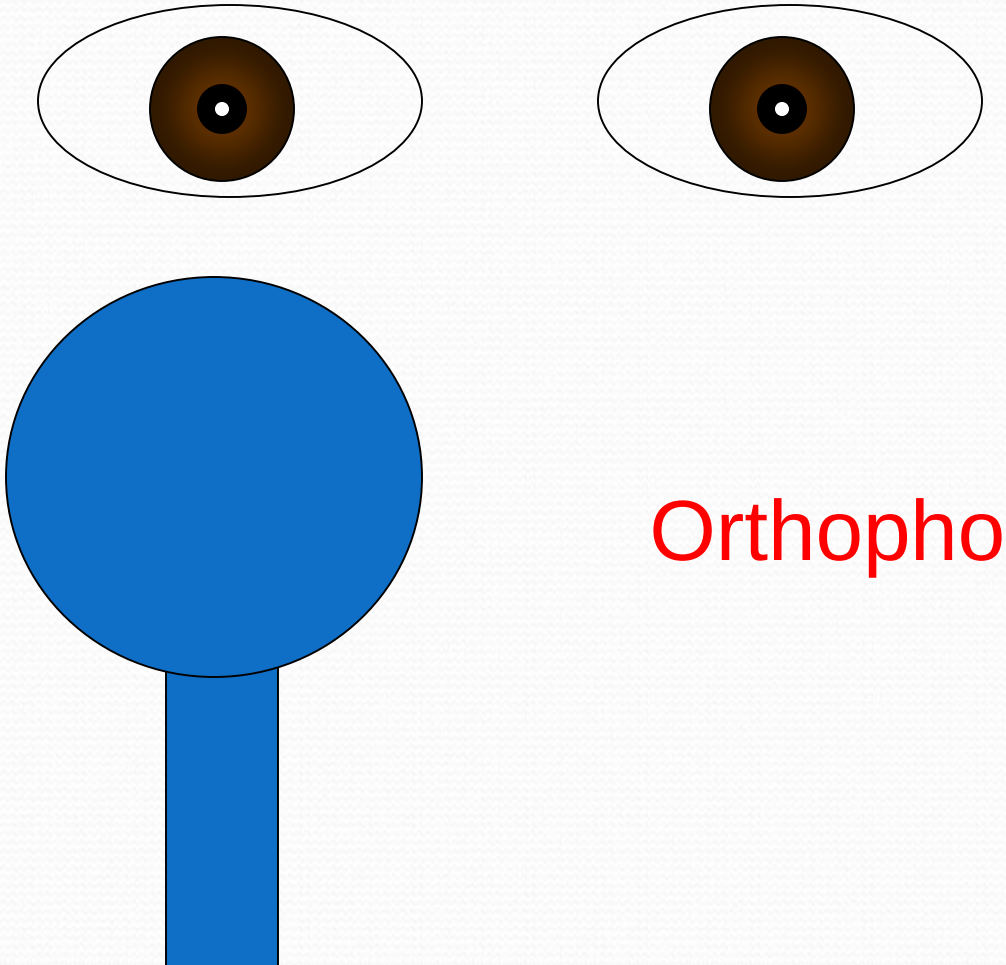
What if after you uncover?

- If it moves inward => exotropic
- If it moves outward =>esotropic
- If it moves up => hypotropic
- If it moves down => hypertropic

- Each eye should be examined separately because there is no way of knowing which eye may be expressing the deviation

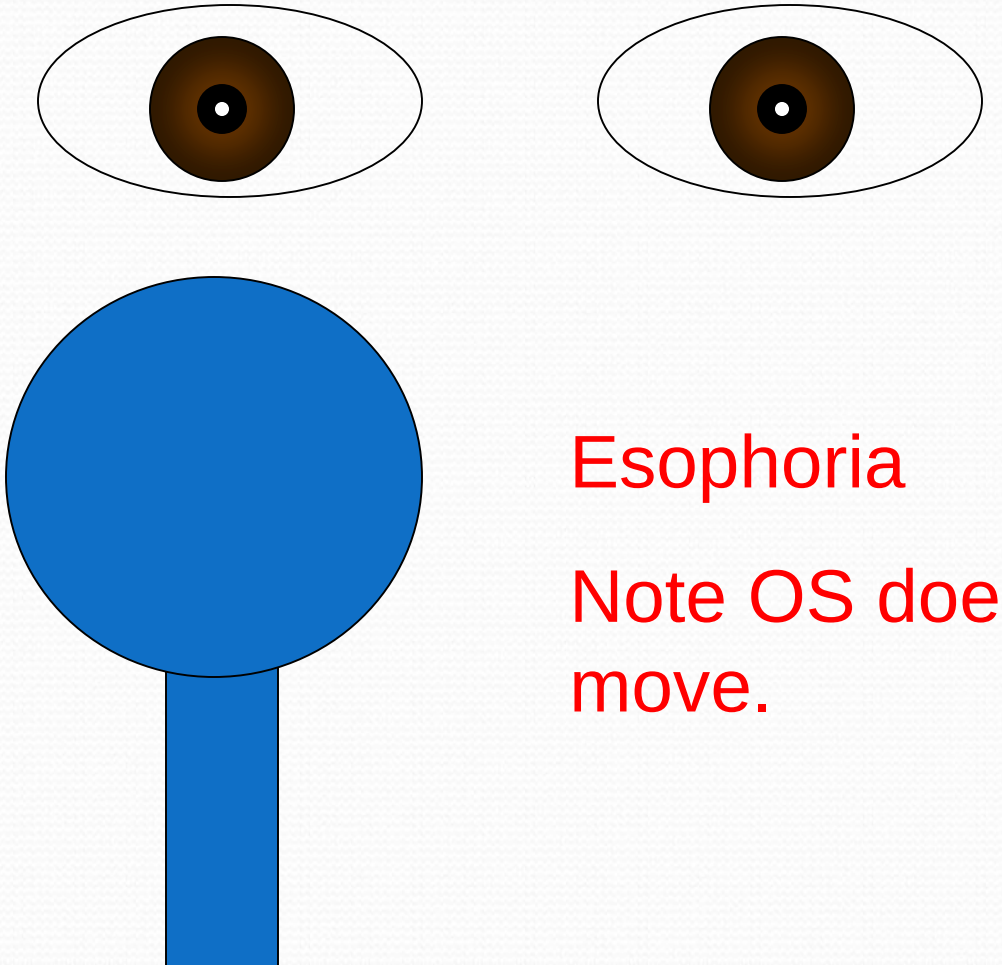
- No shift on cover testing means there is NO tropia
- Very small angle deviation may be difficult to detect so visual acuity testing is important in all cases of suspected strabismus for detecting amblyopia

Cover – Uncover test



Orthophoria

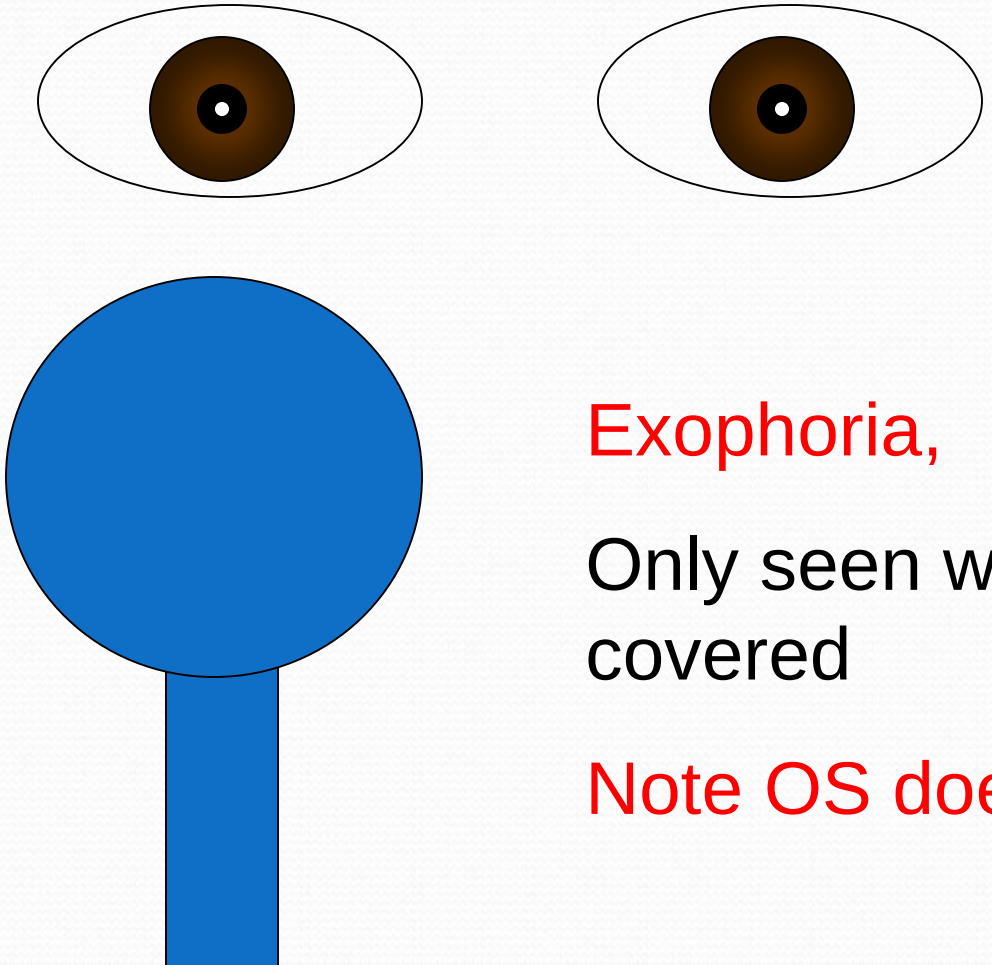
Cover – Uncover test



Esophoria

Note OS does not move.

Cover – Uncover test

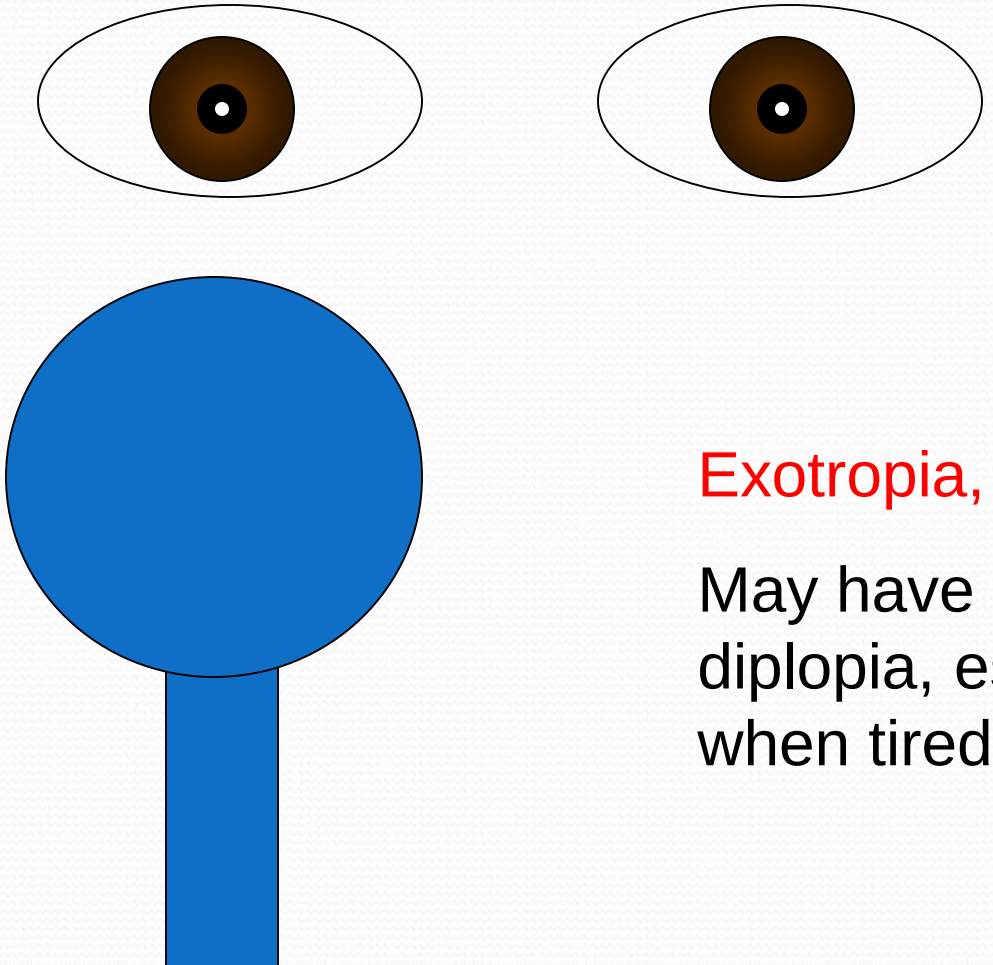


Exophoria,

Only seen when eye is covered

Note OS does not move

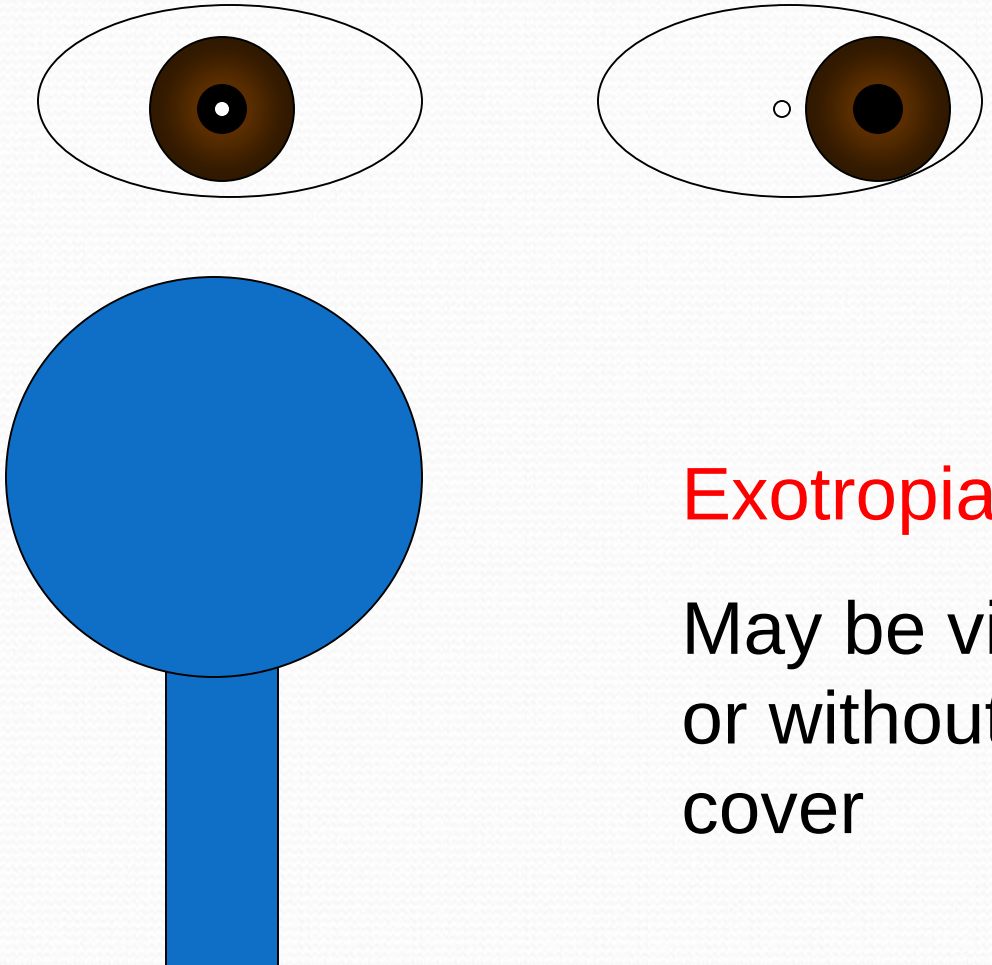
Alternate Cover test



Exotropia, intermittent

May have intermittent diplopia, especially when tired or sick

Alternate Cover test



Exotropia, Constant

May be visible with or without alternate cover

Hirschberg corneal light reflex

- Objective assessment of ocular alignment
- In newborn and often in young children , it may be the only feasible method
- Normally the light is reflected on each cornea symmetrically and in the same position relative to the pupil (i.e. centrally) and visual axis on each eye.

A. Esotropia

B. Exotropia

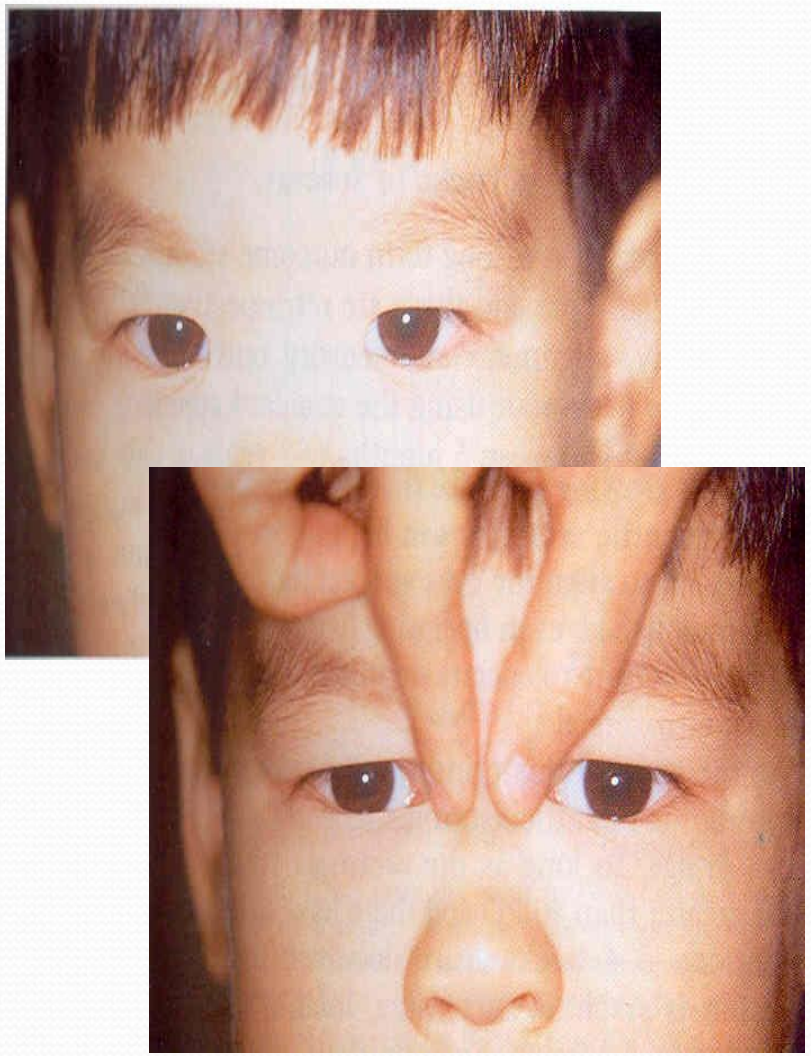
C. Hypertropia

D. Hypotropia

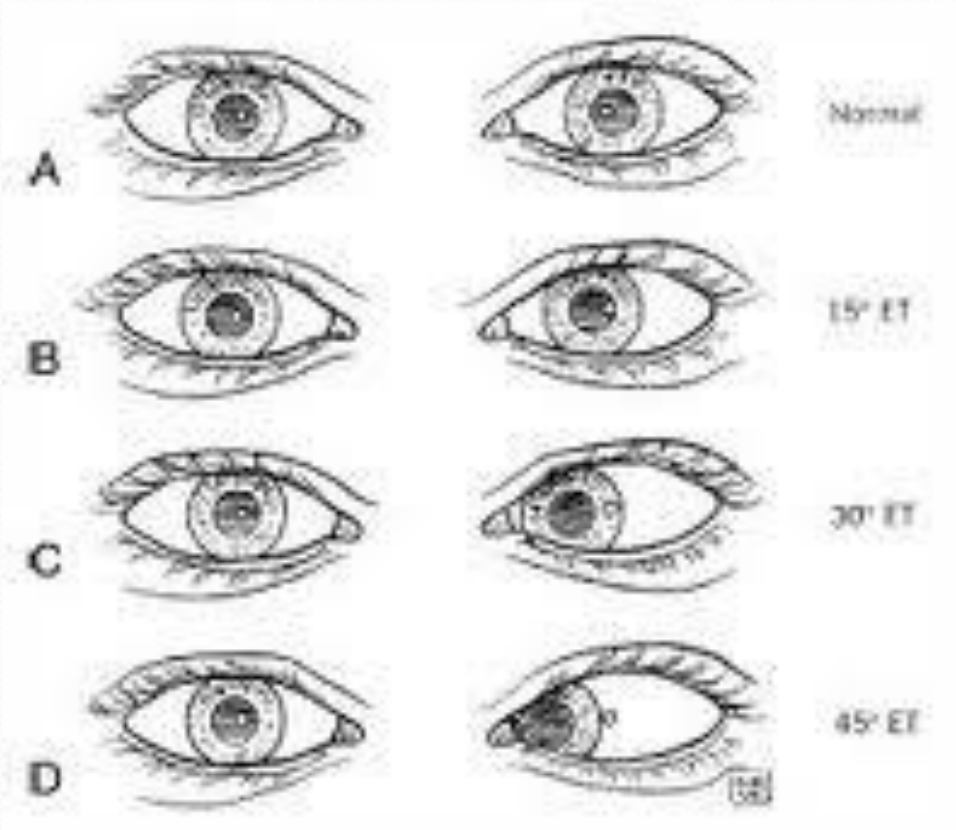
pseudoesotropia

- Small IPD
- Epicanthal folds
- Flat nasal bridge

- Be aware that this diagnosis is a DIAGNOSIS OF EXCLUSION.



- In deviating eye the light reflection will be eccentrically positioned and in the direction opposite to that of the deviation
- the size of deviation can be estimated by the amount of displacement of the light reflex


A
Normal
B
15° ET
C
30° ET
D
45° ET

Work up

- History:
  - Frequency
  - Onset
  - Family history
  - Past medical/surgical history

- Examination:
  - Visual acuity
  - Epicanthus (Be very cautious as its presence doesn't exclude strabismus)
  - Facial asymmetry
  - Cover/uncover test
  - Alternate cover test(latent squint→phoria)
  - Refractive error (topical atropine/cyclopentolate)

Classification of Esotropia

- Right, left or alternating(variable fixation)
- Concomitant or Incomitant
- 1ry, 2ry or Consecutive(overcorrection)

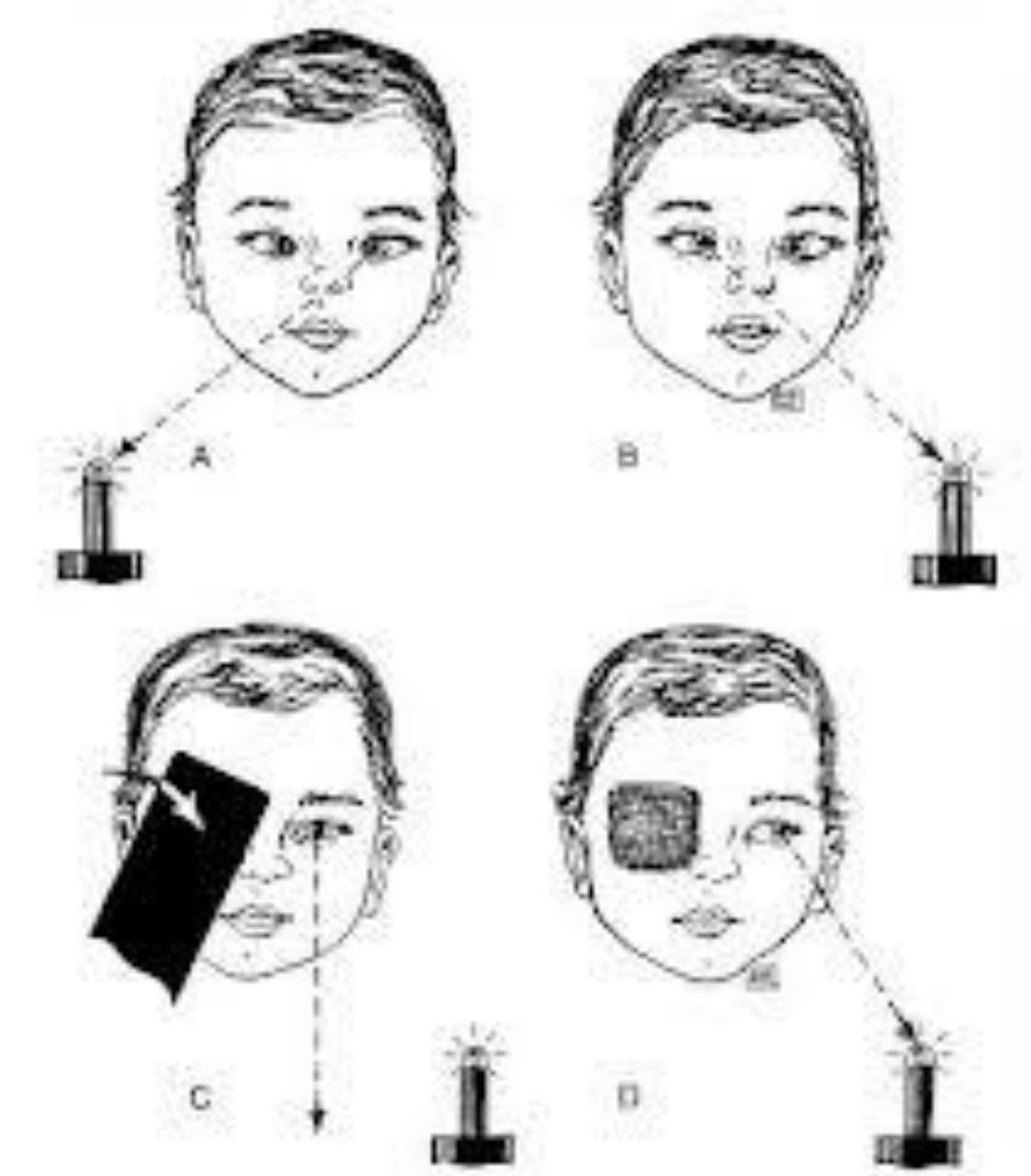
Concomitant Esotropia

- 1)Congenital (Infantile) esotropia
- 2)Accommodative
- 3)Non-Accommodative

- Constant esotropia: present all the time, with or without glasses, may have an accommodative effect
- Intermittent esotropia: not always present, Near esotropia, Distance esotropia and Cyclic esotropia (one day on, one day off).

Infantile Esotropia

- First 6 months of life
- Not associated with hypermetropia
- Large angle of deviation
- Both eyes are convergent (crossed fixation)
- Left fovea fixes right field & vice versa


A
B
C
D

Infantile Esotropia

- Assessment:
  1- Fixation reflex
  2- Cover uncover test
  3- Refraction by cycloplegic drugs
  4- Fundoscopy to evaluate any organic disease (retinoblastoma)
- Rx.: Surgery (recession of both medial recti)

Pre-surgery

Post-surgery

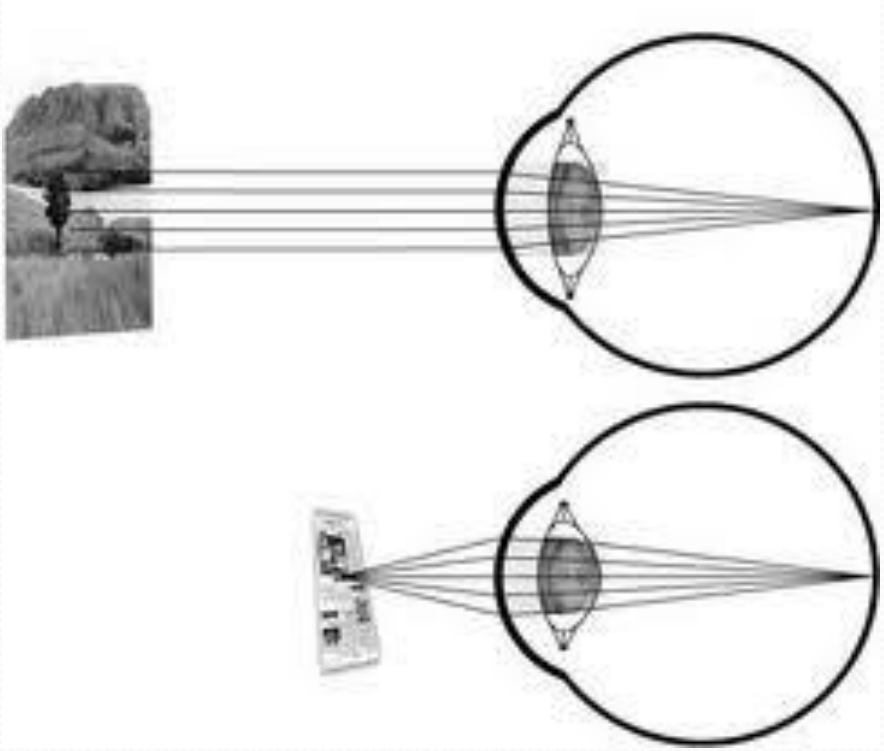
Mobius Syndrome

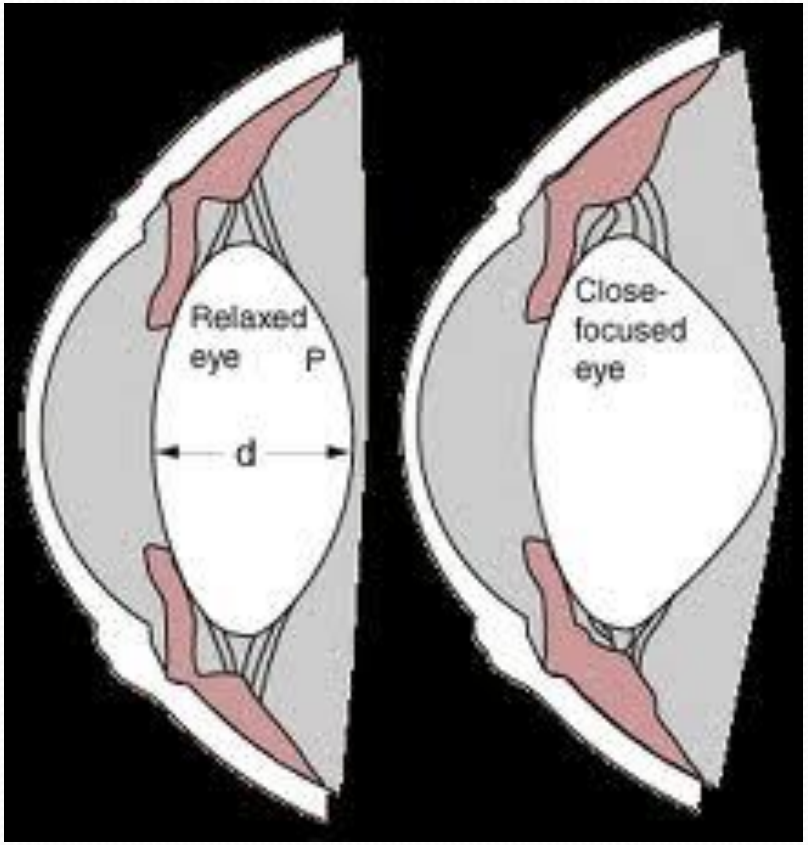
- 6^{th} and 7^{th} nerve underdevelopment
- Crossed eyes (bilateral 6^{th})
- Lack of facial expression (facial palsy)
- Clubbed feet
- Missing fingers or toes
- Chest wall anomalies

Accommodative esotropia

- **Accommodation** is the process by which the human eye changes optical power to maintain a clear image (focus) on an object as its distance changes
- Accommodation acts like a reflex, but can also be consciously controlled.
- The combination of these three movements (accommodation, convergence and miosis) is under the control of the Edinger-Westphal nucleus and is referred to as the *near triad*.

- occurs as a consequence of a reduction in zonular tension induced by ciliary muscle contraction.
- It is normally accompanied by a convergence of the eyes to keep them directed at the same point, sometimes termed *accommodation convergence reflex*




Relaxed
eye
P
d
Close-
focused
eye

Accommodative esotropia

-*Accommodative esotropia* is often seen in patients with a moderate amount of hypermetropia.

-The hypermetrope, in an attempt to "accommodate" or focus the eyes, converges the eyes as well, as convergence is associated with activation of the accommodative reflex..

Types of Accommodative Esotropia

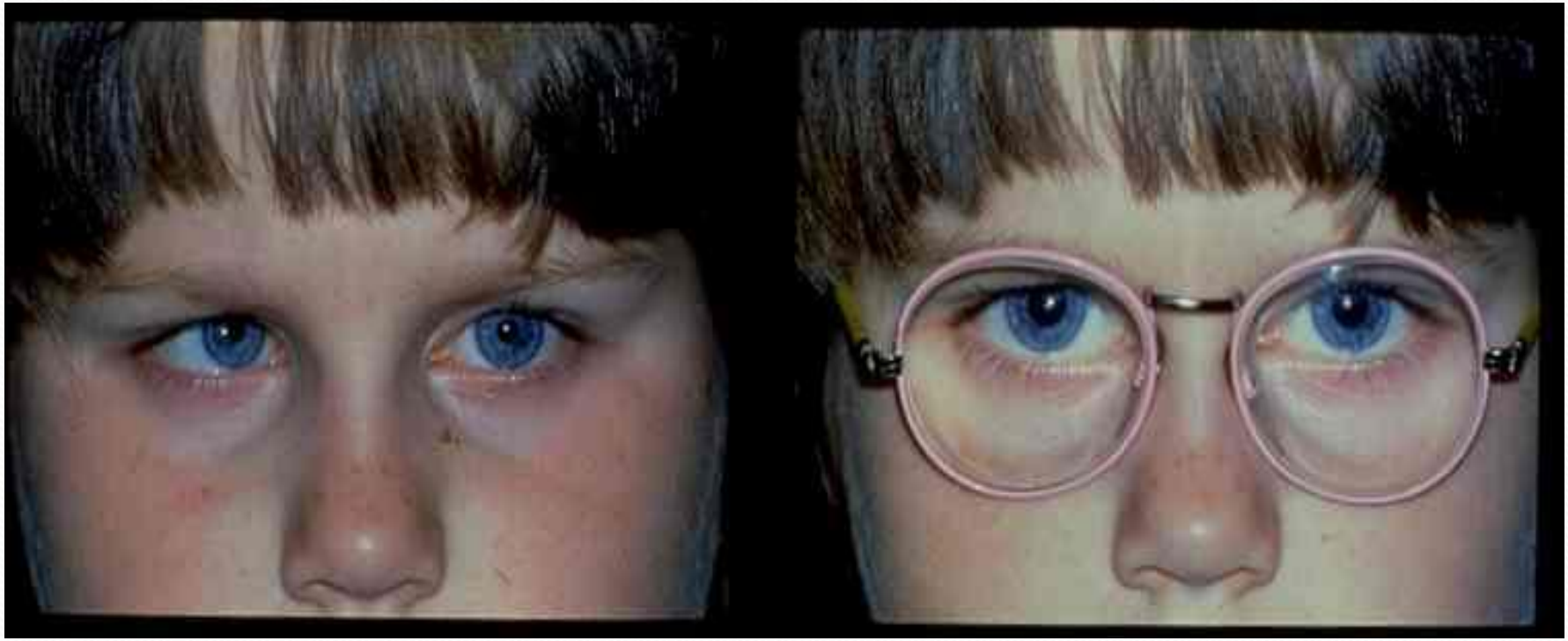
- **1)fully accommodative esotropia:**

  correct glasses is enough to control deviation

- **2)convergence excess esotropia.**

  In this condition the child exerts excessive accommodative convergence relative to their accommodation.

  -In such cases an additional hyperopic correction is often prescribed in the form of bifocal lenses, to reduce the degree of accommodation, and hence convergence.



Glasses are not an alternative to surgery or visa versa

Non- accommodative esotropia

- Induced by :

1- Emotional or physical stress (illness)

2- Sensory deprivation (untreated congenital cataract, optic atrophy)

3- Retinoblastoma

4-…

Exodeviations

1. Intermittent

. divergence excess

. convergence weakness

. Basic

2. Constant

. Congenital

. Sensory

. Consecutive

Intermittent Exotropia

- Onset before 5 years.
- Manifests during times of :
- visual inattention.
- Fatigue
- Stress
- During illness
- If exposed to bright light causes reflex closure of one eye

Concomitant exotropia

- Usually adults or > 5 years
- Types:

  1- Accommodative exotropia

  2- Non-accommodative exotropia

  3- Consecutive exotropia

Accommodative exotropia

- Rare
- Associated with uncorrected myopia
- Can be seen when the child look to a far distance
- Intermittent & later becomes constant
- Rx.: Correct myopia

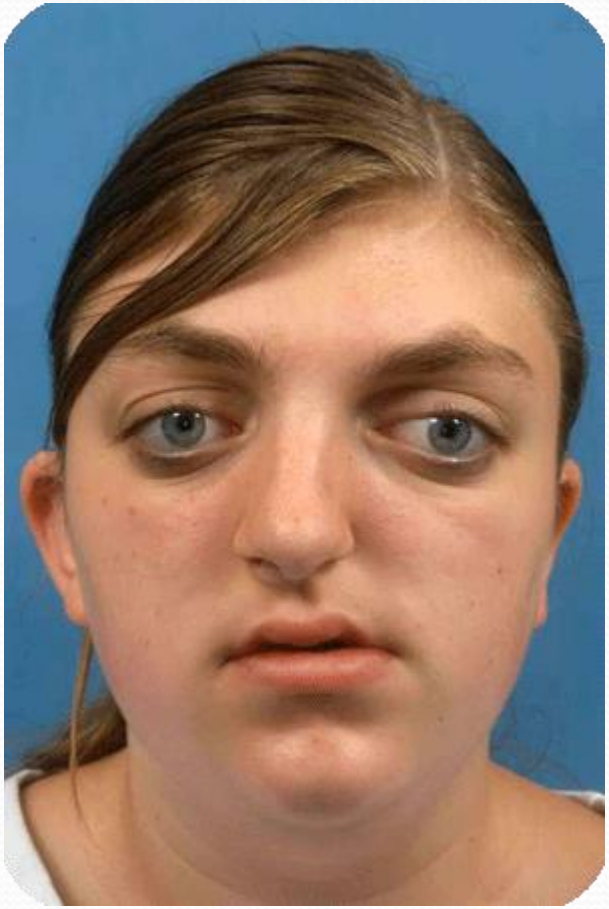
Non-accommodative exotropia

- More common

Crouzon's syndrome(branchial arch syndrome)

Defect in Fibroblast growth factor receptor 2
Autosomal dominant, chromosome 10
shallow eye sockets after early fusion of surrounding bones
Cranial synostosis
Hypertelorism (greater than normal distance between the eyes)
PDA and aortic coarctation

Crouzon’s syndrome



secondary exotropia

seen in cases of unilateral loss of vision

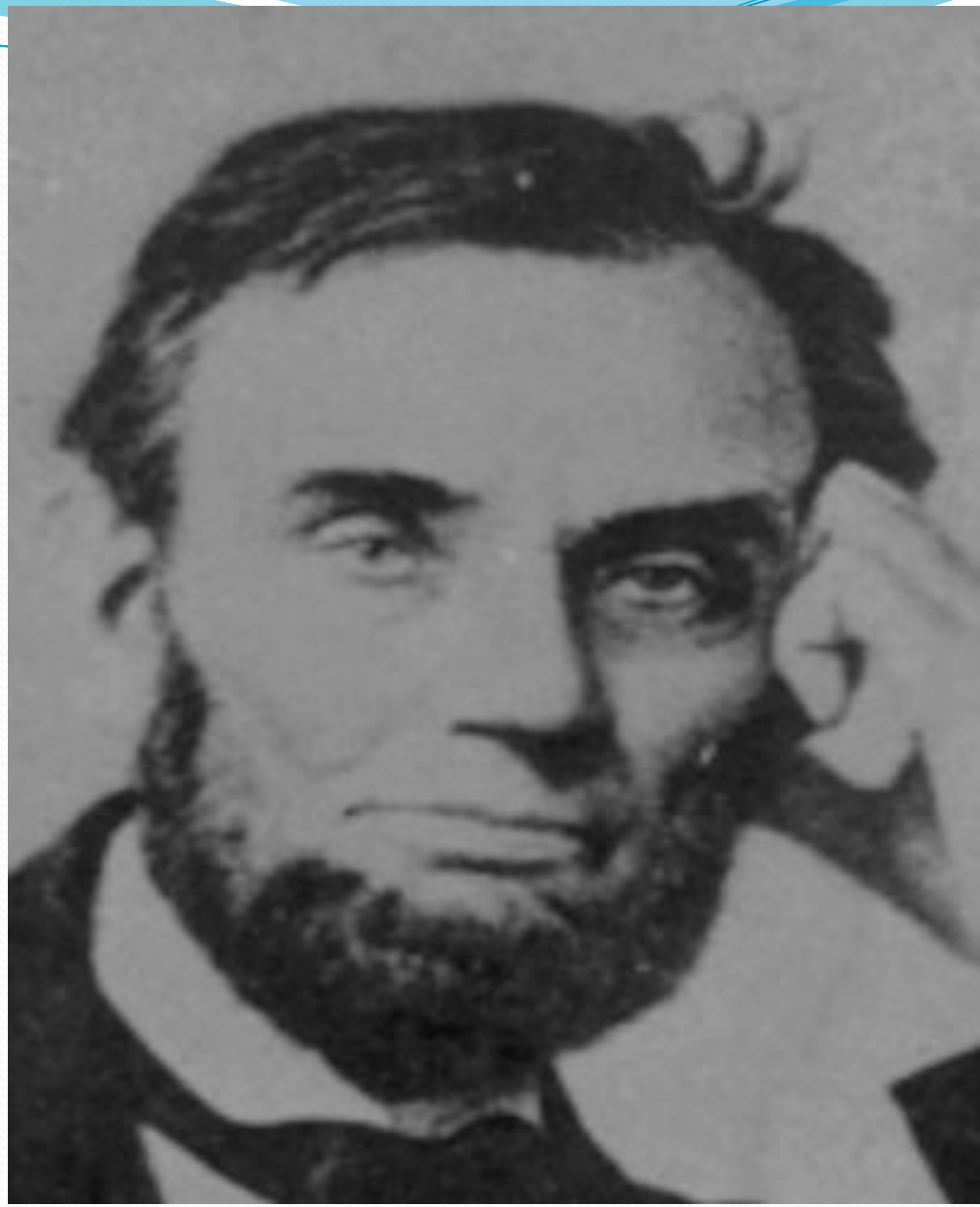
Consecutive exotropia:

Consecutive exotropia:

due to surgical overcorrection of an esodeviation.

Hypertropia,hypotropia

- **Hypertropia** is a condition of misalignment of the eyes (strabismus), whereby the visual axis of one eye is higher than the fellow fixating eye.
- **Hypotropia** is the similar condition, focus being on the eye with the visual axis lower than the fellow fixating eye



Right pseudo-ptosis secondary to right hypertropia

Management(overview)

- Early detection

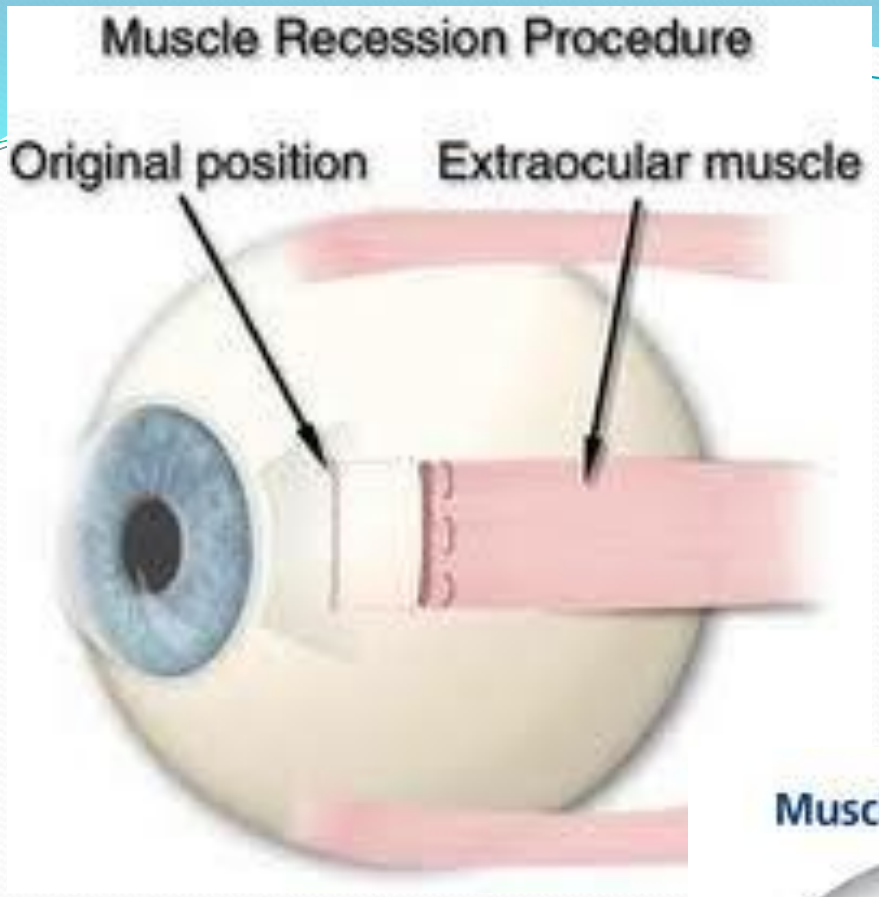
- Glasses can treat some or all of the esotropia in farsighted (hyperopic) and may decrease deviation in a myopic individual with exotropia

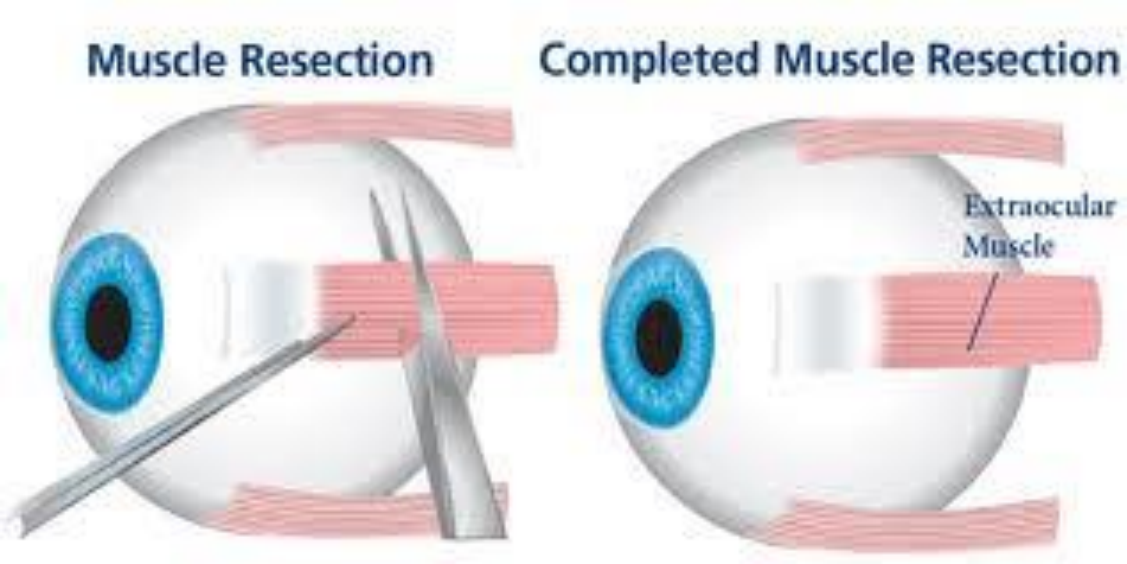
- Tell parents that eyes will continue to cross every time glasses are off.
- Glasses are not an alternative to surgery or visa versa.

- Surgical correction of misalignment may still be necessary for functional or cosmetic reasons .
- It must be stressed that surgery is not an alternative to glasses and patching when amblyopia is present .
- In paralytic strabismus treatment is directed to the underlying pathology

- Surgical Intervention:

1) Recession: incision in the conjunctiva to expose the muscle, muscle is then disinserted on the globe.
2) Resection: cutting and shortening of the muscle and attaching it to its original position


Muscle Recession Procedure
Original position
Extraocular muscle


Muscle Resection
Completed Muscle Resection
Extraocular
Muscle

Amblyopia

- Amblyopia: a unilateral reduction of best corrected central visual acuity in absence of visible organic lesion corresponding to the degree of visual loss.

- Etiology: Suppression (monocular or cortical process producing absolute scotoma) or non use of retino-cortical pathway

Types of Amblyopia

- Strabismic
- Anisometropic
- Form Deprivation

Strabismic amblyopia

- **Adult-onset strabismus** usually causes double vision rather than amblyopia, since the two eyes are not fixated on the same object.

- **Children's** brains, however, are more neuroplastic, and therefore can more easily adapt by suppressing images from one of the eyes, eliminating the double vision.

- This plastic response of the brain, however, interrupts the brain's normal development, resulting in the amblyopia.

- Strabismic amblyopia is treated by clarifying the visual image with glasses, and/or encouraging use of the amblyopic eye with an eye-patch to cover the dominant eye.

As a general practitioner , you are NOT allowed to cover an eye of a child under the age of 10 years, whatever the cause is .

. The ocular alignment itself may be treated with surgical or non-surgical methods, depending on the type and severity of the strabismus.

- The younger the age at which amblyopia is treated; the better is the chance of recovery of vision

Refractive amblyopia

- Refractive amblyopia may result from anisometropia (unequal refractive error between the two eyes).
- The eye which provides the brain with a clearer image (closer to 20/20) typically becomes the dominant eye.
- The image in the other eye is blurred, which results in abnormal development of one half of the visual system

- Refractive amblyopia is usually less severe than strabismic amblyopia and hence commonly missed by General practitioners.

- Frequently, amblyopia is associated with a combination of anisometropia and strabismus

Strabismus and amblyopia

Form-deprivation amblyopia

- results when the ocular media become opaque such as is the case with cataracts or corneal scarring from forceps injuries during birth.

Form-Deprivation Amblyopia

- These opacities prevent adequate visual input from reaching the eye, and therefore disrupt development.
- If not treated in a timely fashion, amblyopia may persist even after the cause of the opacity is removed.

Take home messages

- Strabismus is a symptom/sign (similar to fever) which might be the presenting sign of life threatening conditions.
- Parents are always true about their complaint of presence of squint.
- There is nothing called Pseudo strabismus.
- Never patch the eye of a child.

Thank You