Involuntary Eye Movement Disorders

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Vestibular Nystagmus

Goal: To stabilize vision during brief head movements

How? The eyes makes a slow-phase counter-roll movement in the opposite direction of the head movement at the same speed followed by a quick fast-phase to resume straight-ahead position.

Example: If your head turns to the right = slow phase to left + fast phase to right

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Vestibular Nystagmus

Central Nervous System

- Purely vertical or purely torsional nystagmus
- R/L Posterior canal lesion → R/L anterior canal act unopposed → slow phase up + fast phase down = downbeat nystagmus
- Left anterior and right posterior canal lesion → Right anterior and right posterior canal act unopposed → slow phase counterclockwise + fast phase clockwise = torsional nystagmus beats clockwise

Peripheral Nervous System

- Mixed horizontal nystagmus
- Involves right horizontal canal, right anterior canal, and right posterior canal

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Testing for Vestibular Nystagmus

Method #1: Ask patient to fixate distant target with head held stationary in primary position:

- Purely vertical or purely torsional nystagmus → central vestibular disorder

Method #2: While fixating distant target, remove fixation by placing Frenzel goggles on patient:

- Mixed horizontal-torsional nystagmus → peripheral vestibular disorder
- Purely vertical or purely torsional nystagmus → central vestibular disorder

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Testing for Vestibular Nystagmus

Method #3a: Ask patient to fixate distant target.

To test the HORIZONTAL canal, the examiner briskly rotates the patient's head to the right (or left):

- In health: the patient will maintain fixation
- In disease: the patient cannot maintain fixation and will make 1-2 abnormal re-fixating saccades

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Testing for Vestibular Nystagmus

Method #3b: Ask patient to fixate distant target.

To test the VERTICAL canal, the examiner briskly rotates the patient's head to the right anterior canal-left posterior canal (or left anterior canal-right posterior canal) forward (or backward):

- In health: the patient will maintain fixation
- In disease: the patient cannot maintain fixation and will make 1-2 abnormal re-fixating saccades***

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Testing for Vestibular Nystagmus

In health, the eyes rotate at the same speed as the head movement:

Gain = eye speed/head speed = 1

To test VOR gain, ask the patient to read the Snellen chart while the examiner rotates the patient's head horizontally, then vertically:

- In health: visual acuity will decrease 1-2 lines
- In disease: visual acuity will dramatically decrease >> 2 lines

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Positioning Nystagmus versus Positional Nystagmus

Peripheral Vestibular Disease cause:

Positioning Nystagmus

- Caused by actual head movement
- Paroxysmal
- Subsides over time
- Examples:
  - Pathologic: Meniere's Disease, Benign Paroxysmal Positioning Vertigo (BPPV)
  - Physiologic: head extension vertigo

Central Vestibular Disease cause:

Positional Nystagmus

- Caused by a specific head position
- Does not subside and nystagmus remains until head changes to a different position
- Four types:
  - 1. Positional downbeat nystagmus
  - 2. Central positional nystagmus without vertigo
  - 3. Central positional nystagmus with vertigo
  - 4. Basilar insufficiency

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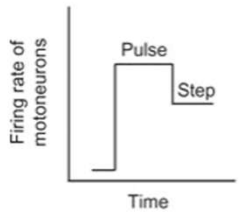
How do our eyes make a saccade?

There are two parts during an *innervation* for a saccade:

1. Pulse of Innervation
   - It is an eye velocity command
   - Burst of activity to propel eye toward target
   - High frequency burst in agonist muscle
   - The contraction of agonist muscles is responsible for rapid eye movements
2. Saccadic step of innervation
   - It is an eye position command
   - Once eye reaches new location after a saccade, new tonic innervation holds eye position
     - Motoneurons and agonist extraocular muscles assume higher level of tonicity
       - Elastic forces of orbit naturally wants to pull eye toward primary position



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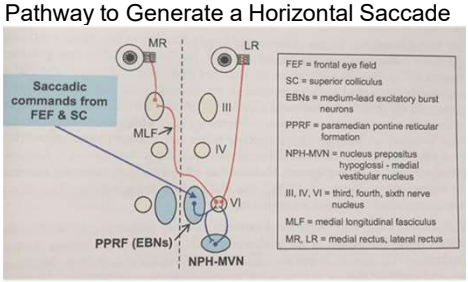
Neural Pathway to Generate a Horizontal Saccade



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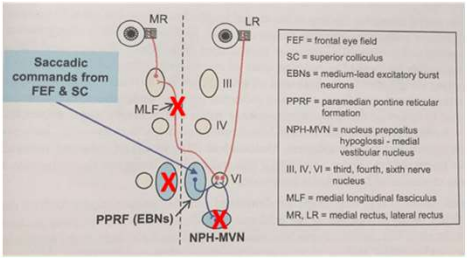
Lesions of Horizontal Saccades



Lesion in the:

- **MLF → ipsilesional adduction palsy (INO)**
- **PPRF → ispilateral conjugate horizontal palsy**
- **NPH-MVN → horizontal gaze-evoked nystagmus**

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Nystagmus occurring in Eccentric Gazes

Physiologic End-Point Nystagmus

Internuclear Ophthalmoplegia

One-and-a-half syndrome

Horizontal Gaze Evoked Nystagmus

Centripetal and Rebound Nystagmus

Bruns' Nystagmus

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Physiologic End Point Nystagmus

- Normal occurrence when looking in extreme gazes
- 3 types:
  - 1. Unsustained end-point nystagmus
  - 2. Sustained end-point nystagmus
  - 3. Fatigue-induced end-point nystagmus

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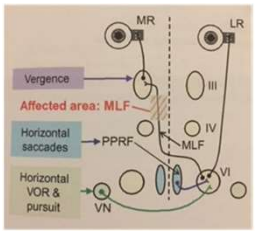
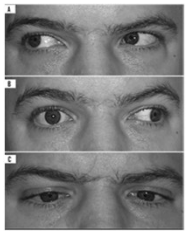
Internuclear Ophthalmoplegia





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Internuclear Ophthalmoplegia

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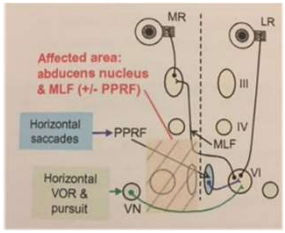
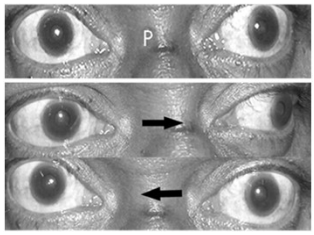
One-and-a-Half Syndrome





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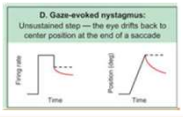
Horizontal Gaze Evoked Nystagmus

•After making a quick horizontal saccade contralesionally, the eyes slowly drift back to primary position

Thus, quick phases away from the side of the lesion, slow phase toward lesion

•Eyes slowly drift to primary position → gaze evoked nystagmus



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Centripetal and Rebound Nystagmus

Centripetal nystagmus - fast phase beating toward primary position

- Occur during sustained eccentric gaze > 30 seconds

Rebound nystagmus - fast phase beating in opposite direction to the gaze-evoked nystagmus when eyes are looking eccentrically

- only lasts for a few seconds

Seldom visually disabling unless it is severe.

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Gaze Evoked Nystagmus and Rebound Nystagmus

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Bruns' Nystagmus

- When looking ipsilesionally, nystagmus is low frequency with large amplitude
- When looking contralesionally, nystagmus is high frequency with small amplitude

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Acquired Pendular Nystagmus

- Combined horizontal, vertical, and torsional nystagmus
- Frequency is the same
- Can be binocular (conjugate), disconjugate (monocular), or disjunctive (convergent or divergent)
- One of the more common types of nystagmus
- Often associated with constant oscillopsia
- If the acquired pendular nystagmus appears:
  - Oblique nystagmus: horizontal and vertical are in phase
  - Elliptical nystagmus: horizontal and vertical are 180 out of phase
  - Circular nystagmus: horizontal and vertical are 090 out of phase

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Acquired Pendular Nystagmus

- Most likely caused by abnormal connections between brainstem and cerebellum
- Follows Alexander's Law
- Etiologies
  - Visual loss (unilateral optic nerve head disease)
  - Spasmus Nutans
  - Acute brainstem stroke
  - Spinocerebellar degeneration
  - Whipple's disease (oculomasticatory myorhythmia)
  - Central Myelin Disorder (most common: Multiple Sclerosis)

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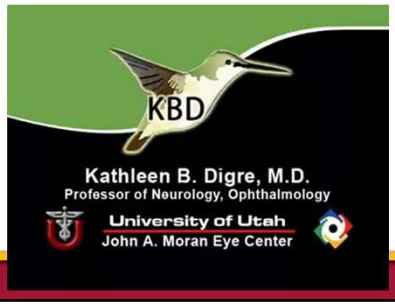
Acquired Pendular Nystagmus



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Infantile (Congenital) Nystagmus Syndrome

- Often horizontal (but can be vertical or elliptical)
- Commonly appears as jerk or pendular nystagmus
- Conjugate
- Nystagmus increases with fixation
- Nystagmus decreases with convergence or in the dark
- Can be seen with head oscillation or latent nystagmus
- Patient has no oscillopsia
- Null point: patient may assume a head turn to minimize nystagmus
- Reversal of OKN response (fast phase in same direction as drum rotation)

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Infantile (Congenital) Nystagmus Syndrome

How can we, optometrists, manage this condition?

- Cycloplegic refraction (astigmatism often found)
- Base-out prism to stimulate convergence
- Refer for EOM surgery (Kestenbaum procedure) to put eyes in null position

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Infantile (Congenital) Nystagmus Syndrome

Differential Diagnoses

- Spasmus Nutans
- Fusional Maldevelopment Nystagmus Syndrome (Latent Fixation Nystagmus)

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Infantile (Congenital) Nystagmus Syndrome

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Latent Nystagmus (Fusional Maldevelopment Nystagmus)

- Occurs or enhanced when monocular
- Appears as conjugate horizontal nystagmus beating toward fixating eye
  - Thus, direction of nystagmus switches direction when cover paddle moved to fellow eye
- Develops when binocular vision development is disrupted within the first 6 months of life:
  - Infantile esotropia (most common)
  - Monocular ophthalmic condition: cataract, glaucoma, significant anisometropia, hypertropia
- Associated with Down's syndrome

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Latent Nystagmus (Fusional Maldevelopment Nystagmus)

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Spasmus Nutans

- Triad of symptoms:
  - 1. Nystagmus
  - 2. Head bobbing
  - 3. Abnormal head position
- Usually appears within first 12 months of life
- No associated neurological abnormalities
- Good prognosis: can spontaneously resolve within 1-2 years after onset, but can persist for up to 8 years

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Spasmus Nutans

What to do?

- Order MRI of the head to rule out tumor along visual pathway
  - Emphasis on the optic chiasm for presence of glioma
- Differentiate spasmus nutans from infantile nystagmus
  - Spasmus Nutans: higher frequency, intermittent
  - Infantile Nystagmus: frequency increases with fixation, constant (jerk or pendular)

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Spasmus Nutans

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Saccadic Dyskinesia

Saccadic Intrusions

- Intermittent saccades disrupts vision/fixation

Saccadic Oscillations

- Continuous saccadic disruptions of fixation

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Three types of Saccadic Intrusions

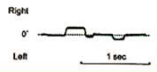
1. Square Wave Jerk

A small-amplitude saccade (0.5 to 3 degree) takes eye away from fixation (0˚)

~200 msec — *Intersaccadic Interval (visual feedback occurs)*

Corrective saccade returns eye back to fixation

- Normal SWJ ~4-6x's/min
- Pathologic SWJ > 15x's/min



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Three types of Saccadic Intrusions

2. Macro-Square Wave Jerk

A larger-amplitude saccade (4 to 50 degree) takes eye away from fixation (0˚)

Corrective saccade returns eye back to fixation

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Three types of Saccadic Intrusions

3. Saccadic Pulses

- Pulse bursts of saccades with defective steps of innervation
  - No saccadic step to maintain eye's fixation and position
- Can be conjugate or monocular

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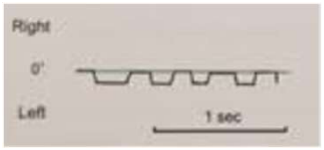
Three types of Saccadic Oscillations

1. Square Wave Oscillation
   - Similar to SWJ but occur continuously



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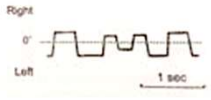
Three types of Saccadic Oscillations



2. Macrosaccadic Oscillations
   - Series of large saccades that straddle fixation
     - Fovea overshoots target each time and never fixates it
   - Intersaccadic intervals when eye is stationary in eccentric fixation
   - Amplitude of saccades increases and decreases within a pulse burst
     - A large amplitude saccade takes eye away from fixation (0˚)
     - ~200 msec
     - *Intersaccadic Interval (visual feedback occurs)*
     - Corrective saccade overshoots and passes to other side of target
   - Disappear in the dark and replaced by irregular saccadic eye movements

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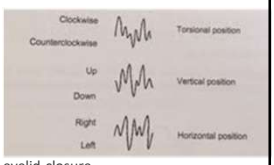
Three types of Saccadic Oscillations

3. Opsoclonus and Ocular Flutter
   - Saccadic oscillations with no intersaccadic intervals
   - High-frequency oscillations (10-15 Hz) with large and variable amplitude



Ocular flutter: one plane (usually horizontal)

Opsoclonus: all three planes (horiz, vert, torsional)

   - Present during fixation, pursuit, convergence, sleep, and eyelid closure

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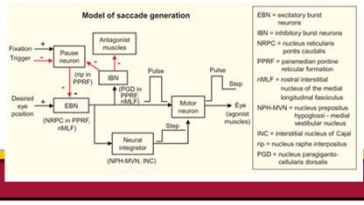
Pathophysiology of Opsoclonus/Ocular Flutter

In health, omnipause neurons inhibit excitatory burst neurons.

In opsoclonus/ocular flutter, the omnipause neurons are no longer inhibited and thus excitatory burst neurons can act uninhibited.



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Etiologies for Opsoclonus/Ocular Flutter

Most common:

- Paraneoplastic syndrome***
  - Neuroblastoma in kids
- Parinfectious brainstem encephalitis
- Metabolic-toxic conditions
- Idiopathic

Other etiologies:

- Meningitis
- Intracranial tumors
- Thalamic hemorrhage
- Multiple sclerosis
- Hydrocephalus
- Hyperosmolar Coma
- Lyme Disease
- AIDS

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Types of Sudden Onset Strabismus

Third Cranial Nerve Palsy

Fourth Cranial Nerve Palsy

Myasthenia Gravis

Multiple Sclerosis

Brain tumor

Aneurysm

Stroke

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Third Cranial Nerve Palsy

Intact left gaze only
(abduction OS only)

Ptosis, mydriasis, and eye position is "down and out",

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Fourth Cranial Nerve Palsy

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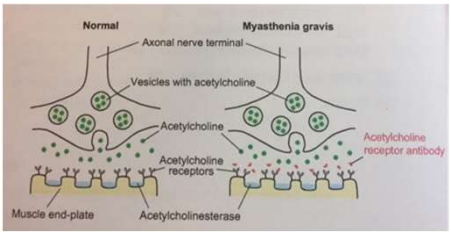
Myasthenia Gravis



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Neuroanatomy of Extraocular Muscles

Cranial nerve III (two divisions)

- Inferior division - inferior rectus, inferior oblique, medial rectus sphincter
- Superior division - superior rectus and levator palpebrae superioris
- Extraocular muscle fibers internal, pupillary fibers external

Cranial nerve IV

- Superior oblique
- Very long pathway, susceptible to trauma

Cranial nerve VI

- Lateral rectus
- Susceptible to increased intracranial pressure

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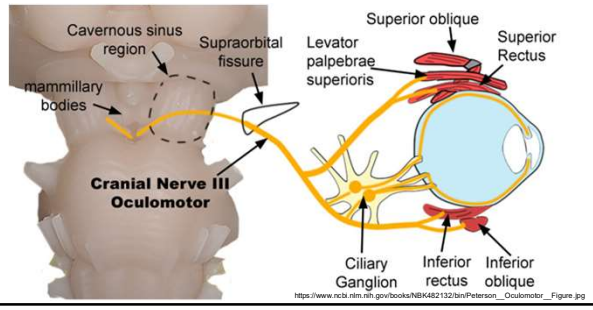
Cranial Nerve III Anatomy



https://www.ncbi.nlm.nih.gov/books/NBK482132/bin/Peterson__Oculomotor__Figure.jpg

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Cranial Nerve III - Compressive vs Ischemic Lesion

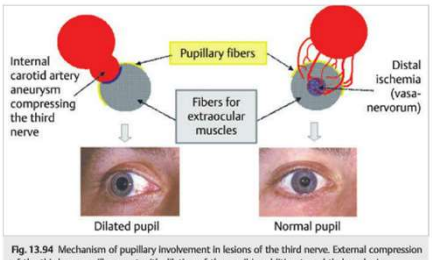

Fig. 13.94 Mechanism of pupillary involvement in lesions of the third nerve. External compression of the third nerve will present with dilation of the pupil in addition to ophthalmoplegia.

https://neuro-ophthalmology.stanford.edu/wp-content/uploads/2018/05/556.png

Ischemic lesion:
-Compromises EOM fibers FIRST
-Pupil is spared
-Typically vascular disease (DM/HTN)
-Imaging still recommended

Compressive lesion
-compromises pupil fibers first
-Expect dilated/blown pupil
-Typically an aneurysm
-Requires prompt referral

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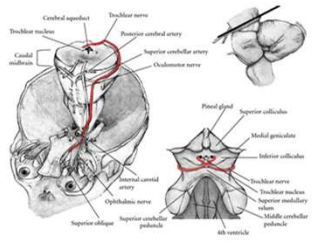
Cranial Nerve IV Anatomy



https://www.researchgate.net/publication/225302226/figure/download/fig5/Pathway-of-the-Trochlear-Nerve-Elevation-and-enlargement-of-the-brainstem-exposes-fibers.png
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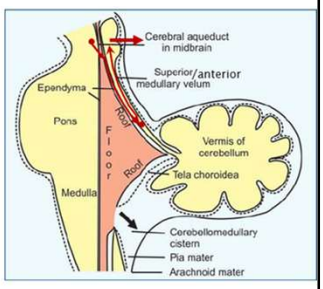

The trochlear nerve travels down to the roof of the cerebellum, crosses, then travels back up to the midbrain before exiting at the level of the midbrain

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Cranial Nerve VI Anatomy

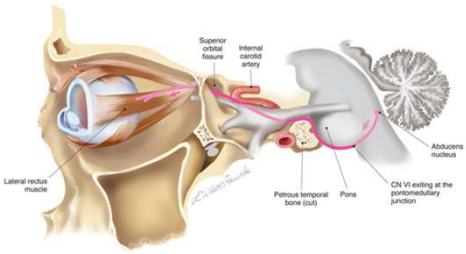

Figure VI–1 Overview of the abducens nerve.

From Cranial Nerves 3rd Ed. ©2010 Wilson-Pauwels, Stewart, Akesson, Spacey. PMPH-USA

The abducens nerve travels over a sharp ridge in the temporal bone (petrous portion of the temporal bone). This makes the abducens nerve susceptible to high intracranial pressure which presses the nerve into the ridge

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Mechanical Restriction versus Neurological Palsy

Forced Duction Test

(+) Forced Duction Test - mechanical restriction

(-) Forced Duction Test - neurological palsy

https://media.springernature.com/lw685/springer-static/image/chp%3A10.1007%2F978-1-4939-2745-6_57/MediaObjects/273339_1_En_57_Fig3_HTML.gif

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Etiologies of Nerve Palsies

Third Cranial Nerve Palsy - Diabetes, compressive lesions

Fourth Cranial Nerve Palsy - Trauma

Sixth Cranial Nerve Palsy - Increased intracranial pressure

Internuclear Ophthalmoplegia - Multiple Sclerosis

Sinus Infections

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Parks' 3 Step

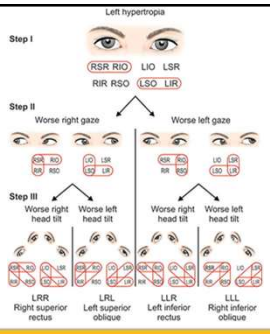
Used for evaluating vertical muscle weakness

First step based on hyper eye

Second step based on worsened horizontal gaze position

Third step based on which head tilt worsens the vertical diplopia

Cannot be used for muscle restriction.



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Evaluating Sudden Onset Strabismus - Hess Lancaster Screen

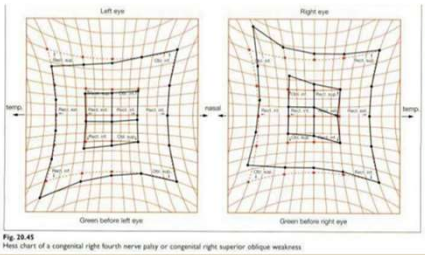
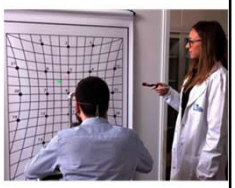


Fig. 20.45
Hess chart of a congenital right fourth nerve palsy or congenital right superior oblique weakness



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Maddox Rod

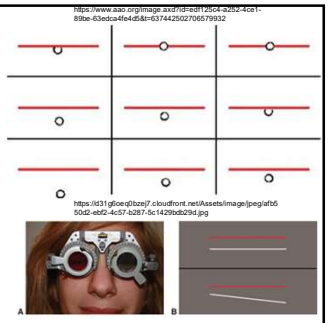
Maddox rod can easily be used to find which gaze position makes diplopia worse

- Hyper eye sees image below
- Eso sees crossed diplopia

Can be used to measure deviation

Double maddox rod can be used to identify a torsional diplopia

- B: left hyper with extorsion



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Evaluating Sudden Onset Strabismus - 9 Gaze Cover Test

Two methods

- Dolls head (VOR)
- Move target

Most important for measuring non-comitant strabismus (>5 difference in some gaze positions compared to others)

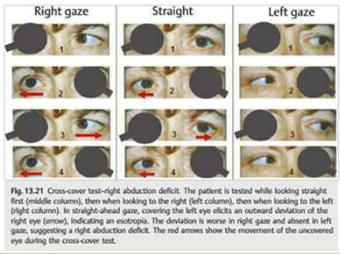

Fig. 13.21 Cross-cover test–right abduction deficit. The patient is tested while looking straight first (middle column), then when looking to the right (left column), then when looking to the left (right column). In straight-ahead gaze, covering the left eye elicits an outward deviation of the right eye (*arrow*), indicating an esotropia. The deviation is worse in right gaze and absent in left gaze, suggesting a right abduction deficit. The red arrows show the movement of the uncovered eye during the cross-cover test.

https://neuro-ophthalmology.stanford.edu/wp-content/uploads/2018/03/620.png

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Differentiating Benign and Malignant Causes of Strabismus

Benign?

- Consider previous cover test findings (high phoria?) and compensating vergence ranges
- Onset of presbyopia

Malignant?

- Acute onset
- Underlying systemic history
- Non comitant

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When to refer?

- Acute onset
- Pupillary involvement
- Pain on eye movements

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Case #1

17 year old male complains of new onset (9 days) binocular vertical diplopia

- No history of trauma
- No recent illness

VAs: 20/20 OU

Stereo testing: Stereo E disappears in superior right gaze

Cover test: Right hypertropia

Park's Three Step Testing: Right hyper, worse on left gaze & left tilt

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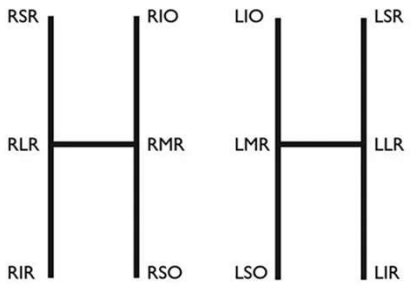
Case #1

Suspect - LSR or RIO

Neither muscles makes sense in isolation.

No ptosis OS to support damage to superior division of oculomotor nerve



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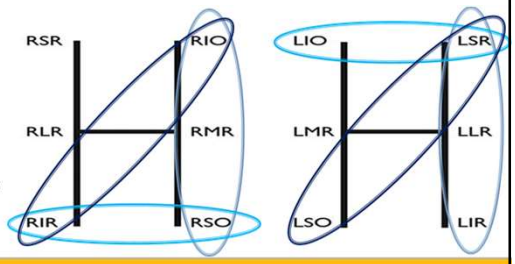
Case #1 - Park's Three Step

LSR damaged in isolation (no ptosis) is suspicious

Consistent with loss of stereo in superior left gaze



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Case #1

Diagnosis: Pineal Gland Germinoma

Initial doctor wanted to refer for vision therapy

Park's three step repeated with consistent results led to referral to pediatrician who sent patient for immediate imaging

Inoperable tumor but good prognosis due to small size

Sudden onset vertical diplopia is always concerning especially when the involved muscle is NOT the superior oblique

Congenital SO palsies are common

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Case #2

28 year old male with complaint of sudden primarily vertical diplopia OU for 4 days

Patient reports diplopia improves with left head tilt, worsens with right tilt

Patient reports having cold/sinus infection for a week

BCVA: 20/20 OD, OS, OU

Maddox rod & double Maddox rod: see images

Park's Three Step: Left hyper, worse w/right tilt & gaze

Hess Lancaster Screen: See related slide

CC Dist		
2 EP 2 LHT	2 EP Ortho	2 EP 2 RHT
2 EP 2 LHT	2 EP Ortho	2 EP Ortho
2 EP 4 LHT	2 EP Ortho	2 EP Ortho

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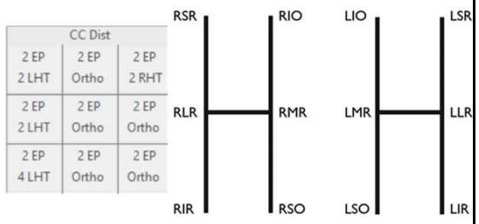
Case #2 - Diplopia worse down and right - Maddox Rod

Suspect - RIR or LSO

LSO makes sense due to long trochlear pathway, but is NOT consistent with improvement on left head tilt. SO palsy tilt away.

CC Dist		
2 EP 2 LHT	2 EP Ortho	2 EP 2 RHT
2 EP 2 LHT	2 EP Ortho	2 EP Ortho
2 EP 4 LHT	2 EP Ortho	2 EP Ortho



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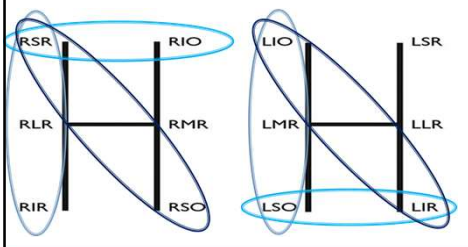
Case #2 - Left hyper, worse on right gaze & right tilt



RSR is not consistent with our findings on 9 gaze Maddox rod.

Likely a restriction not muscle weakness
Positive forced duction

Unlikely for RSR to be damaged in isolation (with no ptosis)
Superior division of CN III

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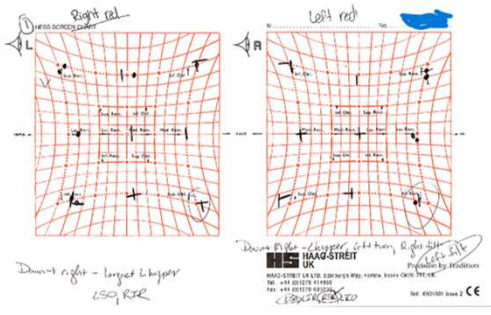
Case #2

Note the eso shift in both eyes.

LIO is slightly over acting

RSR is under-acting

Pretend you are the patient



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Case #2 - Bonus Testing - Double Maddox Rod

Red over right eye

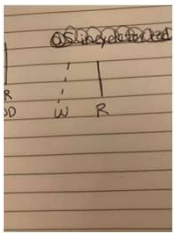
Left eye sees image tilted inward which indicates the left eye was actually extorted

Left inferior muscle is overacting

Inferior muscles extort

Left superior muscle is underacting

Superior muscles intort



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Case #2

Diagnosis: Over acting LIO due to sinus infection/congestion

The inferior oblique is most inferior muscle in orbit

Patient noted pain when doctor tapped left maxillary bone

Parks three step results not consistent with Maddox rod results - restriction

Slight overacting of LIO on Hess Lancaster Screen

Slight underacting of RSR on Hess Lancaster Screen

Overacting of LIO explains left hyper that worsens with right gaze

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Conclusions & Clinical Pearls

If patients complain of oscillopsia, you can determine if the vestibular system is involved if visual acuity is decreased in addition to observing if the nystagmus is unidirectional or mixed.

Acquired pendular nystagmus is one of the more common types of nystagmus and patients have constant oscillopsia.

Patients with infantile nystagmus do not have oscillopsia.

Paraneoplastic syndrome is the common etiology for opsoclonus, specifically neuroblastoma for kids.

Ischemia is the most common cause of third cranial palsy, specifically diabetes.

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Conclusions & Clinical Pearls

Most likely muscle palsy in isolation is superior oblique muscle

Make sure patient has head straight during muscle testing

If Park's 3 step doesn't match suspected muscle from EOMs, CT, Maddox rod, consider muscle restriction instead of palsy

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Thank you!!!

Questions?

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