Diplopia
Common Causes and Management

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Overview
- Introduction
  - Anatomy/physiology review
- Conditions/Management
  - Common
    - Pediatric
    - Adult
    - Geriatric
  - Uncommon
- Case Review

EOM Anatomy
- 6 Extra ocular muscles
  - Controlled by 3 cranial nerves
    - CN III – SR, MR, IR, IO
    - CN IV – SO
    - CN VI – LR
- Other
  - 7th muscle controls eyelid
    - Levator palpebrae superioris
    - Innervation = Sup CN III

EOM Action Review

<table>
<thead>
<tr>
<th>Muscle</th>
<th>1st Action</th>
<th>2nd Action</th>
<th>3rd Action</th>
<th>Other Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>SR</td>
<td>Elevation</td>
<td>Intorsion</td>
<td>Adduction</td>
<td>Innervation = Sup CN III</td>
</tr>
<tr>
<td>MR</td>
<td>Adduction</td>
<td></td>
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</tr>
<tr>
<td>IR</td>
<td>Depression</td>
<td>Extension</td>
<td>Adduction</td>
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</tr>
<tr>
<td>LR</td>
<td>Adduction</td>
<td></td>
<td></td>
<td>Innervation = CN VI</td>
</tr>
<tr>
<td>SO</td>
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<td>Longed CN III</td>
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EOM Physiology
- EOM coordination
  - Herring’s Law:
    - Yoked muscles
  - Sherrington’s Law:
    - Antagonist muscles

EOM Testing
- Range of Motion
- Cover test
  - Unilateral
  - Alternating
- Other
  - Forced Duction
  - EMG: electromyography
Normal Binocular Vision
- Retinal correspondence
- Sensory fusion
- Motor fusion
- Stereopsis

Diplopia
- Due to absence of retinal correspondence
- Visual confusion
- Adaptations
  - Suppression
  - Monocular/alternating/intermittent
  - Abnormal retinal correspondence

Diplopia
- Monocular vs. Binocular
  - Monocular = Cataracts, CME, Bifocal Misalignment, uncorrected refractive error
  - Binocular = Needs further testing
    - Differentials:
      - Binocular vision dysfunction
      - Systemic etiology
      - Cranial nerve abnormalities
        - Palsy
        - Ischemic
        - Mass
        - Trauma

Initial Diplopia Case History
- Monocular/Binocular
- Horizontal/Vertical/Oblique
  - Systemic conditions

Initial Diplopia Work-up
- EOM’s
- Alignment evaluation
  - Cover test, Red lens, Maddox rod

Binocular Vision Testing
- Vergences
  - Von Graphe
  - Prism Bar
- NRA/PRA
- Fused cross-cylinder
- MEM
- Stereopsis
- Worth 4-dot
Testing for Misalignment

- Gross Evaluation
  - Corneal light reflex
  - Hirschberg/Kappa
    - 1 mm ~ 15-22 Δ
  - Krimsky
    - Place prism in front of fixating eye
    - Increase strength until reflex centers
  - Red reflex test/Bruckner
    - White reflex = strabismus/significant refractive error difference

- Cover test
  - UCT
  - ACT
- Parks 3 Step
  - Double maddox rod
  - Red lens test

Parks 3 Step aka Bielschowsky Test

- First, determine which muscles are under acting
  - I.E. - Right hyper ... either the R.E. inferior muscles are not pulling the eye down, or the L.E. superior muscles are not pulling the eye down.

  ![Diagram of Parks 3 Step](image)

- Next, determine if the hyper worsens in right or left gaze
  - I.E. - If the hyper worsens in left gaze (right head turn), we circle the muscles responsible for left gaze.

  ![Diagram of Parks 3 Step, Cont...](image)

- Finally, circle the head tilt that worsens the hyper
  - I.E. - If the head tilt worsens when tilted to the right shoulder, we make a circle in that direction.

  ![Diagram of Parks 3 Step, Cont...] (image)

***Which ever muscle has three circles touching it is the paretic/underacting muscle, therefore the above example would be a RSO Palsy.
- Don’t forget, this patient will most likely walk in with a left head tilt... “always trust the tilt”

Parks 3 Step Example

- 20 Δ L Hyper in primary gaze
- 10 Δ L Hyper in left gaze (right head turn), 30 Δ L Hyper in right gaze (left head turn)
- 15 Δ L Hyper with R head tilt, 40 Δ L Hyper with L head tilt

  ![Diagram of Parks 3 Step Example](image)

Solution = Left Superior Oblique Palsy

Double Maddox Rod Test

Evaluates patient for excyclotorsion

Possible Patient Responses

- If the patient reports the lines are parallel, there is no excyclotorsion
- If the patient reports the lines are not parallel, rotate the trial frame axis until the lines are parallel. Greater than 10° of rotation is a positive test.
Strabismus

- Ocular misalignment
  - Non-corresponding retinal points
  - Disrupts binocularity

- Comitancy
  - Comitant
    - Magnitude consistent in all gazes
  - Non-comitant
    - Magnitude varies in different gazes

- Hyper deviations
  - Phoria (HP or HyperP)
  - Tropia
    - Intermittent (IET)
    - Constant (CHP)

- Torsion deviations

- Eso deviations
  - Phoria (EP)
  - Tropia (ET)
    - Intermittent (IET)
    - Constant (CET)

- Exo deviations
  - Phoria (XP)
  - Tropia (XT)
    - Intermittent (IET)
    - Constant (CET)

Strabismus Nomenclature

- Hyper deviations
  - Phoria (HP or HyperP)
  - Tropia
    - Intermittent (IET)
    - Constant (CHP)

Pattern Strabismus

- Non-comitant deviations

- Exotropia
  - 'A' pattern
  - 'V' pattern
    - Less symptomatic

- Esotropia
  - 'A' pattern
  - 'V' pattern

Unique Forms of Strabismus

- Duane's retraction syndrome
  - Rarely diplopic (suppression)

- Type I
  - Exotropia
    - Adduction deficit
    - Enophthalmos with Adduction
    - Esotropic

- Type II
  - Adduction deficit
    - Enophthalmos with Adduction
    - Exo T/P

- Type III
  - Ab and Adduction deficit
    - Enophthalmos with Adduction

- Treatment
  - Surgical if large angle in 1° gaze
  - Asymptomatic = monitor

Comitant Deviations

<table>
<thead>
<tr>
<th>Esotropia</th>
<th>Exotropia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Basic ET</td>
<td>Basic XT</td>
</tr>
<tr>
<td>Acute ET</td>
<td>Divergence excess (DE)</td>
</tr>
<tr>
<td>Sensory ET</td>
<td>Convergence insufficiency (CI)</td>
</tr>
<tr>
<td>Divergence insufficiency (ET)</td>
<td></td>
</tr>
<tr>
<td>Near reflex spasm</td>
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Pattern Strabismus

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- Esotropia
  - 'A' pattern
  - Less symptomatic
  - 'V' pattern

Type I

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<thead>
<tr>
<th>Exotropia</th>
<th>Enophthalmos with Adduction</th>
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<td>Esotropic</td>
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Type II

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- Duane’s retraction syndrome
- Rarely diplopic (suppression)

- Treatment
  - Surgical if large angle in 1° gaze
  - Asymptomatic = monitor
Unique Forms of Strabismus

- Moebius Syndrome
  - Congenital CN VI and VII palsies
  - Esotropia and corneal exposure
  - 10% have developmental delay

Vertical Deviations

- Two common etiologies
  - Neurologic
    - Congenital
      - CN IV palsy – weakened sup. Oblique
      - + Head tilt to opposite shoulder
    - Dissociated Vertical Deviation
      - (+) Hypodeviation
      - Associated with infantile ET
  - Mechanical
    - Mass (orbital)

Unique Forms of Strabismus

- Brown's Syndrome (can be bilateral)
  - Inability to elevate while in Adduction
  - Sup oblique tendon obstruction

- Treatment
  - Symptomatic
    - Prism
    - Monitor
  - Surgical – if torticollis/improved binocularity

1° vs. 2° Deviations - Paresis

- Primary
  - Deviation angle with functioning eye fixating
- Secondary
  - Deviation angle with paretic eye fixating
- Hering's Law
  - Secondary angle > primary angle

Symptomatic Strabismus

- Intermittent
  - Diplopia when deviation present

- Acquired
  - Decompensated phoria
  - Cranial nerve palsy
  - Other systemic etiology

Acquired Vertical Strabismus

- CN III
- CN IV
  - Decompensated congenital
  - Post-trauma
  - Ischemic
  - Acute acquired (CVA, mass)
- Other
  - Skew, Myasthenia, Graves
Management

- Best Correction
- Patching/Medical therapy
  - PEDIG Review
- Orthoptics
- Surgical evaluation

Management, cont...

- Temporary Support
  - Occlusion
  - Fresnel Prism
  - Injections
    - Botox®

Systemic Causes for Binocular Diplopia

- Thyroid: The “can cause everything” diagnosis
  - Anytime you suspect thyroid disorder TSH/T3/Free T4
  - Forcedduction test will be (+) in most cases (due to ECM infiltration, most often IR)

  - Autoimmune: Variable and transient symptoms
    - Ocular myasthenia gravis: order Anti-AChR, anti-striated muscle test, single fiber EMG
    - Dyspnea/Dysphagia/SOB = ER immediately

- Ischemia: Must r/o GCA in older patients
  - Immediate ESR and CRP

Cranial Nerve III Palsy

- Patients will present with a ptosis, dilated pupil, and an eye that appears down and out (only the SO and LR are acting)

- Patients may not complain of diplopia until the upper lid is elevated if a complete ptosis is present

Cranial Nerve III

- Cranial Nerve 3 innervates;
  - Contralateral SR
  - Ipsilateral MR, IR, and IO
  - Bilateral innervation to the Levator muscles

- CN 3 splits into two divisions just before entering the superior orbital fissure
  - Superior: SR and Levator
  - Inferior: MR, IR, IO; Iris sphincter (miosis), and the ciliary body (accommodation)
Cranial Nerve III Palsy, cont…

- Must differentiate Ischemic vs. Aneurysm/Neoplasm
  - Ischemic- typically pupil sparing and resolve over 3-4 months without treatment
    - Associated with DM/HTN
    - Work-up: GGT (glucose tolerance test), FTA-Abs, ANA
  - Aneurysm/Neoplasm - pupils typically affected and will worsen with time.
    - EMERGENCY: Pupil affected CN III palsy along with the worst HA of their life *** Impending Aneurysm***
    - This needs an immediate referral to the ER
    - Work-up: MRI/MRA

Cranial Nerve IV

- CN 4 is responsible for the SO muscle
- Patients CC: **Oblique Diplopia**
- Isolated CN 4 palsy most often congenital or traumatic etiology.
  - Typically have a head tilt to OPPOSITE shoulder
  - Many congenital cases will decompensate in 5th-6th decade of life
    - Consider Vertical Vergence testing or double Maddox rod
    - Acquired cases; evaluate patients with a parks-3 step test.

Cranial Nerve VI Palsy

- Cranial nerve 6 is responsible for the LR muscle
- Nuclear palsy causes an ipsilateral horizontal gaze palsy.
- Most often due to ischemic events in elderly patients (only 1 LR affected) – think diabetes and hypertension.
  - Work-up includes: b.p., GGT, Sed rate, FTA-Abs, ANA
- In kids; most often seen post-viral infection... no recent sickness must r/o neoplasm and increased ICP.

Cranial Nerve VI Palsy

- Patients typical chief complaint: **Horizontal diplopia**
- Presentation:
  - Esotropia in primary gaze
  - Limited/absent Abduction

Multiple CN’s Affected

- Cavernous sinus syndrome- lesion in either the Cav sinus OR the SOF (superior orbital fissure)
  - Patient presents with: periorbital pain, ipsilateral EOM paresis, sensory loss along V1 and V2
  - **EMERGENCY** - must r/o ICA aneurysm, Cavernous Carotid Fistula, Tolosa-Hunt (Granulomatous inflammation) and a nasopharyngeal carcinoma**
- Orbital Apex Syndrome – Looks like a Cav sinus syndrome, but CN II also involved | VF changes/swollen ONH

Observe vs. Image

- Isolated?
  - Traumatic?
    - Congenital?
      - Neuroimage & further evaluate
  - Non-Isolated?
    - Neuroimage & further evaluate

- Traumatic?
  - Congenital?
    - Vasculo-pathic?
      - Observe
      - Progressive or not improved

- Non-vasculo-pathic?
  - Neuroimage & further evaluate
Cranial Nerve Example
- This is a complex cranial nerve 6 acute palsy, the patient was sent for a CT STAT
  - CT was WNL
- Over the next 4 months the traumatic palsy completely resolved
  - 4mo f/u – CT in 1° gaze = 4△ EP(‘)

Abnormal EOM patterns
- Supra/Intra Nuclear Pathways
  - VI = CN6 Nucleus aka – the lateral gaze center.
  - VN = Vestibular Nucleus – information from the inner ear – semicircular canals and ampulla.
  - NPH = Nucleus prepositus hypoglossi - tells the eyes where they are in space.
  - PPRF = paramedian pontine reticular formation – coordinates horizontal saccades.
  - MLF = Medial longitudinal fasciculus – tells eyes which direction they

Internuclear Ophthalmoplegia
- A lesion of the IPSILATERAL MLF
  - Named for the side of the lesion/aka the eye with the paretic EOM
  - Convergence will be intact because CNIII nucleus is not affected

Internuclear Ophthalmoplegia, cont...
- Unique EOM pattern-
  - (+) Beat Nystagmus OS
  - No ADduction OD secondary to lack of innervation to RMR… therefore we name the INO for the side with the palsy, convergence is spared, so we know the lesion is behind the CNIII nucleus… we have a Right INO

Internuclear Ophthalmoplegia, cont...
- Differentials-
  - Lesion aka multiple sclerosis – Order MRI with FLAIR to r/o paraventricular white matter lesions
  - Highly likely etiology because this would be a small lesion in a small space.
  - Usually younger patients (under 40)
  - Mass- typically a mass will cause more signs, but early on it is possible to have only an INO – order MRI
  - Cardiovascular accident (CVA)- think stroke. If acute symptoms immediate ER referral and order a CT scan. If long standing, MRI.
  - Usually older patients, also typically have a Hx of HTN

Binocular Internuclear Ophthalmoplegia
- A lesion of BOTH MLF pathways
  - Convergence may/may not be spared.
  - Do not need to specify a side, BOTH sides are affected.
Binocular Internuclear Ophthalmoplegia, cont...

- **EOM Pattern**:
  - Intact convergence suggests a lesion is lower in the brainstem (below the midbrain, usually in the pons).
  - Lack of convergence suggests a lesion is higher in the brainstem within the midbrain.
  - AKA a WEBINO (Wall-Eyed BINO)

- **Differentials**:
  - Lesion - aka multiple sclerosis – less likely because there would have to be similar lesions bilaterally in the MLF which is possible, but less likely.
  - Usually younger patients (under 40)

  - **Mass**: Because there’s little respect to the vertical midline with a BINO, a space occupying lesion must be ruled out – order MRI with and without contrast.

  - Cardiovascular accident (CVA) - less likely with a BINO because the defect from a stroke will typically respect the vertical midline, but a hemorrhagic leak will spill over to bilateral – order CT scan STAT.

  - Usually older patients, also typically have a Hx of HTN

One and One-Half Syndrome

- Lesion of either the CN6 nucleus AND the ipsilateral MLF

- OR

- A lesion of the PPRF AND the ipsilateral MLF

- **EOM pattern**:

- (+) complete gaze paralysis when looking ipsilateral to the lesion, and an INO in the other direction.

- Again, Name the INO... looking at the pattern in left gaze, the right eye fails to ADOuct, therefore we have a lesion of the Right MLF – in this example we have a RIGHT 1 and 1/2 syndrome.

One and One-Half Syndrome, cont...

- **Differentials**:

  - Lesion - aka multiple sclerosis – Order MRI with FLAIR to r/o paraventricular white matter lesions – less likely etiology because typically these lesions are not that large, but it is possible.

  - Usually younger patients (under 40)

  - **Mass**: May also have other complaints consistent with a space occupying lesion, i.e. Headache, Nausea, Vomiting, Visual loss, or Visual field cuts – order MRI with and without contrast.

  - **Cardiovascular accident (CVA)** - think stroke. If acute symptoms immediate ER referral and order a CT scan. If long standing, MRI.

  - Usually older patients, also typically have a Hx of HTN

Variable Diplopia

- **Myasthenia Gravis** – usually worst in the evening

  - Intermittent symptoms

  - Age of onset

    - Women – 2nd to 3rd decade of life

    - Men – 6th to 7th decade of life

- **Decompensated Phoria**

  - Typically purely horizontal, without associated lateral gaze restrictions
Myasthenia Gravis

- Autoimmune attack of acetylcholine receptors
  - Associated with thymoma (thymus gland tumor)

- Ocular and Systemic Components
  - 90% will have ocular findings at some point
  - Many begin as OMG (ocular)
    - Some convert to OMG (generalized) within 2 years
    - Goal = to prevent conversion

- (+) Systemic involvement, must R/O: SOB, trouble talking/swallowing

Clinical Findings

- Cogan’s lid twitch
- Improvement with ice pack, rest
- Worsening by (AKA enhancement)
  - Elevating the contralateral eyelid
  - Prolonged up look

Blood work

- Anti-AchR
- MuSK
- Single fiber electromyography (EMG)
- Chest X-ray/Chest CT

Oral Prednisone

- Oral acetylcholinesterase inhibitors
- Lid crutches/Sx
  - For persistent ptosis

Diagnosis Treatment

Diplopia Summary

- Most common causes of diplopia
- First line treatment
- Conditions requiring emergent/urgent referral
- When to consider surgical evaluation

Clinical Case Review