DOUBLE TROUBLE

• No financial disclosures
DOUBLE TROUBLE

Objectives:

- Discuss the most common causes of acquired diplopia in adults
- Review the management of these patients presenting with diplopia
- Review the indications for further systemic workup
CC: Double vision

1st Question: Does it go away with one eye covered?

- Yes: Binocular diplopia
- No: Monocular diplopia
DOUBLE TROUBLE
WHERE TO START

Monocular Diplopia
- Refractive
- Optical
- Retinal

Binocular Diplopia
- Optical
- CNS
- Orbital
- Systemic
- Binocular Vision Disorders
MONOCULAR DIPLOPIA
WHERE TO START

- Refractive
- Optical
- Retinal
MONOCULAR DIPLOPIA

CC: Double vision

1. Does it go away when you cover one eye?
   • No (monocular)

2. Right, Left, both eyes?

3. Constant or intermittent?
MONOCULAR DIPLOPIA

- Uncorrected refractive error
- Regular astigmatism
- Irregular astigmatism
- Anisometropia
- Diagnostic RGP
- Refraction
- Topography
- Exam

REFRACTIVE
MONOCULAR DIPLOPIA

- Optical Irregularities
  - Dry eye
    - Intermittent

- Corneal Opacities
  - Diagnostic RGP

- Iris/pupil
  - Cosmetic contact lens

- Cataract
MONOCULAR DIPLOPIA

- Retina
  - Macular Edema
  - Central Serous
  - Exudative ARMD
CC: Double vision

1st Question: Does it go away with one eye covered?

• Yes: Binocular diplopia
• No: Monocular diplopia
BINOCULAR DIPLOPIA

- Optical
- CNS
- Orbital
- Systemic
- Binocular Vision Disorders
BINOCULAR DIPLOPIA
WHERE TO START

• History
  • the double vision up-and-down or side-by-side?
  • Is the diplopia greater at distance or near?
  • Is the event variable?
  • Has this happened before?
  • Is there associated pain?

• Associated symptoms
  • Headache
  • GCA symptoms
  • Numbness/tingling
  • Motor deficits
BINOCULAR DIPLOPIA
WHERE TO START

• Systemic History
  • Vascular disease: HTN, HLD, DM
  • Graves Disease
  • Multiple sclerosis
  • Myasthenia gravis
BINOCULAR DIPLOPIA
WHERE TO START

- Acuity
- Motility
- Binocular Function
- Stereo
- Refraction
- Anterior and Posterior Segment
  - Signs of vascular risk factors
  - Optic disc edema or pallor
BINOCULAR DIPLOPIA
WHERE TO START

- Motility Grading
  -1 completes 75% of movement
  -2 completes 50% of movement
  -3 completes 25% of movement
  -4 does not move from primary

BINOCULAR DIPLOPIA
WHERE TO START

• Binocular Function
  • Cover Test
    • Cover-Uncover and Alternating
    • Distance and Near
    • Primary and cardinal positions of gaze
• Hirschberg
• Krimsky
• Stereo
BINOCULAR DIPLOPIA

• Optical
• CNS
• Orbital
• Systemic
• Binocular Vision Disorders
• Optical
  • Induced or unwanted prism
THIRD NERVE PALSY
SIGNS AND SYMPTOMS

- Binocular Diplopia
- Ptosis
- Exotropia and hypotropia
  - Down and out
  - Limitation of all fields of gaze except temporally
- Pupil sparing/involved
- With or without pain
- Complete and partial

https://eyerounds.org/cases/156/1-partial-eyelid-ptosis-adduction-supraduction-infraduction.jpg
THIRD NERVE PALSY
CN III PATHWAY

- Parasympathetic fibers to pupil on outside of nerve making them more susceptible to compression

https://myneurosurg.com/cranial-anatomy/cranial-nerve-3-oculomotor-nerve/
THIRD NERVE PALSY
CN III STRUCTURE

• Nuclei
  • Oculomotor – motor fibers
  • Edinger-Westphal – parasympathetic fibers

• Branches
  • Superior – Superior Rectus and LPS
  • Inferior – Inferior Rectus, Medial Rectus, Inferior Oblique, Ciliary Ganglion

THIRD NERVE PALSY
ETIOLOGY

Pupil Sparing

• More common:
  • Ischemic/microvascular

• Less common
  • Cavernous sinus disease
  • GCA

Pupil Involving

• Aneurysm
• Tumor
• Trauma
• Cavernous sinus disease
• Ischemia
THIRD NERVE PALSY
MANAGEMENT

Pupil Sparing
• Management of vascular risk factors
• >50yo – w/ vascular risk factor
  • close observation
• <50yo
  • Neuroimaging
• Systemic Workup if GCA suspected
  • ESR/CRP

Pupil Involved
• Medical Emergency
  • CNS imaging (urgent)
  • Aneurysm must be ruled out
THIRD NERVE PALSY
MANAGEMENT

Pupil Sparing

- Occlusion patch
- Prism
- Pupil involvement can be delayed 5-7 days
- If ischemic...
  - Improvement in 4-8 weeks
  - Resolution in 3-6 months
- Strabismus surgery if deviation stable after 6 months

Pupil Involved

- Directed by neurology and/or neurosurgery
FOURTH NERVE PALSY
SIGNS AND SYMPTOMS

- Binocular, vertical diplopia
- Dizziness
- Difficulty Reading
- Hypertropia on affected side
  - Worse in downgaze
- Motility can look normal
  - Limited downgaze when adducted

https://webeye.ophth.uiowa.edu/eyeforum/atlas/pages/CN-IV-Palsy/index.htm
FOURTH NERVE PALSY
CN IV PATHWAY

- Longest intracranial pathway
- Exits from back of the brainstem
- Most vulnerable to trauma
### FOURTH NERVE PALSY

**ETIOLOGY**

<table>
<thead>
<tr>
<th>Common</th>
<th>Uncommon</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Trauma</td>
<td>• Tumor</td>
</tr>
<tr>
<td>• Microvascular/Ischemic</td>
<td>• Hydrocephalus</td>
</tr>
<tr>
<td></td>
<td>• Aneurysm</td>
</tr>
<tr>
<td></td>
<td>• GCA</td>
</tr>
</tbody>
</table>
FOURTH NERVE PALSY

WORKUP

• Three-Step Test:
  • Determine which eye is deviated upward in primary gaze
    • The higher eye comes down after being uncovered
  • Determine whether upward deviation is greater when looking left or right
    • Hyper deviation is worse on contralateral gaze
    • Unopposed action of inferior oblique
  • Determine whether upward deviation is greater when tilting head to right or left
    • Hyper deviation is worse when tilting head toward shoulder of affected side
    • IV is intorter

• Right IV palsy has a right hypertropia worse in left gaze and right head tilt
FOURTH NERVE PALSY
WORKUP

- Workup by PCP/internist
- MRI of the brain
  - Patients <45 years old with no history of head trauma
  - Patients 45-55yo with no vasculopathic risk factors or trauma
- ESR, CRP, platelets if GCA is expected
  - Top Normal ESR
    - Male: Age/2
    - Female: Age +10 / 2
  - CRP – does not rise with age
  - Platelets – may have thrombocytosis
FOURTH NERVE PALSY
MANAGEMENT

- Patching
- Prism
  - Fresnel
  - Wait 3-6 months for stabilization to prescribe
- Presumed vascular or idiopathic: 1-3 months
- Unresolved in 3 months – refer for imaging studies
- Additional neurological abnormalities – refer for imaging studies
SIXTH NERVE PALSY

SIGNS AND SYMPTOMS

- Binocular, horizontal diplopia
- Limited or complete loss of abduction on the affected side
- Esotropia distance > near
- Worse in direction of affected lateral rectus muscle
SIXTH NERVE PALSY
CN VI PATHWAY

• Most commonly affected ocular motor nerve in adults
SIXTH NERVE PALSY
ETIOLOGY

More Common
- Microvascular/ischemic
- Trauma
- Idiopathic

Less Common
- Increased ICP
- Cavernous sinus mass
- Multiple sclerosis
- Stroke
- GCA
SIXTH NERVE PALSY
WORKUP

• MRI of the brain
  • <45 yo
  • 45-55 with no vascular risk factors
  • History of cancer?
  • Does not resolve after 3-6 months
• ESR, CRP, platelets for any suspicion of GCA
SIXTH NERVE PALSY
MANAGEMENT

• Patching
• Prism
  • Fresnel
  • Wait 3-6 months for stabilization
• Re-examine every 4-6 weeks
INTERNUCLEAR OPHTHALMOPLEGIA (INO)

SIGNS AND SYMPTOMS

• Diplopia
  • Intermittent or constant

• Adduction deficit in affected eye
  • Partial to complete

• Left INO = cannot left eye cannot adduct

https://www.slideserve.com/polly/neuro-ophthalmology
Damage to the interneuron between VI (LR) and III (MR)

This interneuron is called the medial longitudinal fasciculus (MLF).

The MLF can be damaged by any lesion (e.g., demyelinating, ischemic, neoplastic, inflammatory)

The MLF is supplied blood by branches of the basilar artery and ischemia in the vertebrobasilar system can produce an ischemic INO
INO
ETIOLOGY

• Elderly
  • Stroke

• Young
  • Multiple Sclerosis
INO WORKUP

- Head CT or MRI
INO MANAGEMENT

• Ischemic and demyelinating typically recover
• If XT, patching or surgery for unresolved deviations
ASSOCIATED SYNDROMES

• WEBINO
  • Wall Eyed Bilateral INO
  • (Bilateral INO with bilateral XT)

• WEMINO
  • Wall eyed monocular INO
  • Unilateral INO with XT

• One and a half
  • INO with damage to PPRF
  • Ipsilateral conjugate gaze palsy
  • Ipsilateral INO
  • Right 1 1/2 = Cannot look to the right; cannot adduct in the right

• Eight and a half
  • 1 ½ plus a VII palsy
SKEW DEVIATIONS

• Vertical deviation not due to any single muscle or nerve
• Damage to brainstem or cerebellum
  • Damage to vestibular nerve inputs to brain stem
• Usually caused by stroke
  • multiple sclerosis
  • Inflammation
  • Trauma
  • Tumor
• Usually accompanied by other neurological symptoms
• Need to differentiate from IV palsy
  • Upright – Supine Test
  • Double Maddox Rod
BINOCULAR DIPLOPIA

- Optical
- CNS
- Orbital
- Systemic
- Binocular Vision Disorders
BINOCULAR DIPLOPIA

ORBITAL ETIOLOGIES

- Orbital Tumors
- Orbital Pseudotumor
- EOM Restriction
BINOCULAR DIPLOPIA
ORBITAL ETIOLOGIES

• Tumor
  • Proptosis
  • Numbness or tingling around eye
  • Vision loss
  • Pain
Orbital Pseudotumor
- Non-infectious, inflammatory process of the orbit without a known local or systemic cause
- Associated with a variety of rheumatologic conditions
- Painful

Management
- Rheumatology labs
- CT
- Steroids (mild)
- Surgical resection

DDx:
- Orbital cellulitis
- Thyroid eye disease
BINOCULAR DIPLOPIA
ORBITAL ETIOLOGIES

- EOM Restriction
  - Trauma
    - Entrapment
  - Age
    - Functional
    - Structural
BINOCULAR DIPLOPIA

- Optical
- CNS
- Orbital
- Systemic
- Binocular Vision Disorders
SYSTEMIC ETIOLOGIES

• Thyroid Eye Disease
• Myasthenia Gravis
• Multiple Sclerosis
THYROID EYE DISEASE

• What is it – Autoimmune disease caused by antibodies against receptors present in thyroid cells and extraocular muscles and soft tissues of the orbit

• How does it affect the eyes
  • NOSPECS
  • VISA
    • Vision (Optic Nerve)
    • Inflammation (Congestion)
    • Strabismus (EOM)
    • Appearance (Exposure)
THYROID EYE DISEASE

• How does it cause diplopia
  • Thickening and enlargement of EOM causes motility restrictions
  • Elevation and abduction are most commonly affected

• What do you do for treatment of diplopia
  • Prism or strabismus surgery
  • Normalize thyroid levels
  • teprotumumab (Tepezza)
    • Reduce proptosis and improve diplopia
    • FDA approved JAN 2020
MYASTHENIA GRAVIS

• What is it
  • Antibodies against acetylcholine receptors
    • ACh release is normal, but fewer receptors may not be able to trigger an action potential
MYASTHENIA GRAVIS

• How does it affect the eyes/cause diplopia
  • Most common presenting symptoms are ocular muscle weakness
    • Ptosis
    • Strabismus
    • EOM deficits
      • MR is most common
  • Affects small muscle groups first
  • Ocular symptoms before systemic

• How do we treat it
  • Prism or strabismus surgery
  • Acetylcholinesterase inhibitors
  • Pyridostigmine - long acting cholinesterase inhibitor
INTRACRANIAL HYPERTENSION

• What is it
  • Elevated pressure in CSF surrounding brain and spinal cord
    • Pseudotumor cerebri
    • Tumor
    • Chiari Malformation

• How does it affect the eyes
  • Headache
  • Episodes of vision loss
  • VF loss
  • Horizontal diplopia
  • Optic disc edema
How does it cause diplopia
- Most commonly VI palsy

How do we treat it
- Acetazolamide – CAI to decrease ICP
- Diplopia resolves with ICP
MULTIPLE SCLEROSIS

• What is it?
  • Autoimmune disease against CNS myelin
  • Females > Males (2:1)
  • 15-45 yo
  • Sensory – pain, numbness, tingling, pins and needles sensation
  • Motor – muscle weakness, muscle spasms, impaired coordination and balance, difficulty with speech and swallowing
  • Autonomic – bladder and bowel dysfunction
MULTIPLE SCLEROSIS

• How does it affect the eyes
  • Optic Neuritis
    • 20% present with optic neuritis
    • 75% will have episode during lifetime
    • Decreased VA
    • Orbital Pain (92%) worse with eye movement
    • Desaturated color vision
    • VF loss
    • APD
  • Internuclear Ophthalmoplegia (30%)
  • Rarely VI
MULTIPLE SCLEROSIS

• How does it cause diplopia
  • INO
  • Rarely CN palsies

• How is it treated
  • Diplopia resolves on its own
  • Systemic therapy focuses on recovering from attacks, slowing progression and managing symptoms
BINOCULAR DIPLOPIA

- Optical
- CNS
- Orbital
- Systemic
- Binocular Vision Disorders
ACQUIRED EXOTROPIA

- Convergence Insufficiency
  - Exo deviation Near > Distance
  - Head Injury
  - CNS degenerative disorders
- Age
  - Changes in fusional response
ACQUIRED ESO TROPIA

- Divergence Insufficiency
  - Eso deviation distance > near
    - Age
      - Degeneration of pulley system
    - Stroke
    - Demyelinating disease
    - High ICP
ACQUIRED DIPLOPIA

- Decompensating phoria
  - Can be horizontal or vertical
  - Usually intermittent

- Break down in fusion vergence response
  - Large phoria

- Full EOM’s
WHEN WORKUP IS NEEDED...

- Cranial Nerve Palsies
  - Young patient (<45)
  - Patient with no vascular risk factors
  - Unresolving episode (3-6 months)
  - Presence of other neurological symptoms
  - Multiple Cranial Nerve Palsies

- Concern for Systemic Etiology
  - Giant Cell Arteritis
  - Myasthenia Gravis
  - Multiple Sclerosis
  - Intracranial Hypertension