2020

Clinical Optometry Update and Review

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DOUBLE TROUBLE

ACQUIRED DIPLOPIA IN ADULTS

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

DOUBLE TROUBLE
WHERE TO START

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
REFRACTIVE

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

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

DOUBLE TROUBLE

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

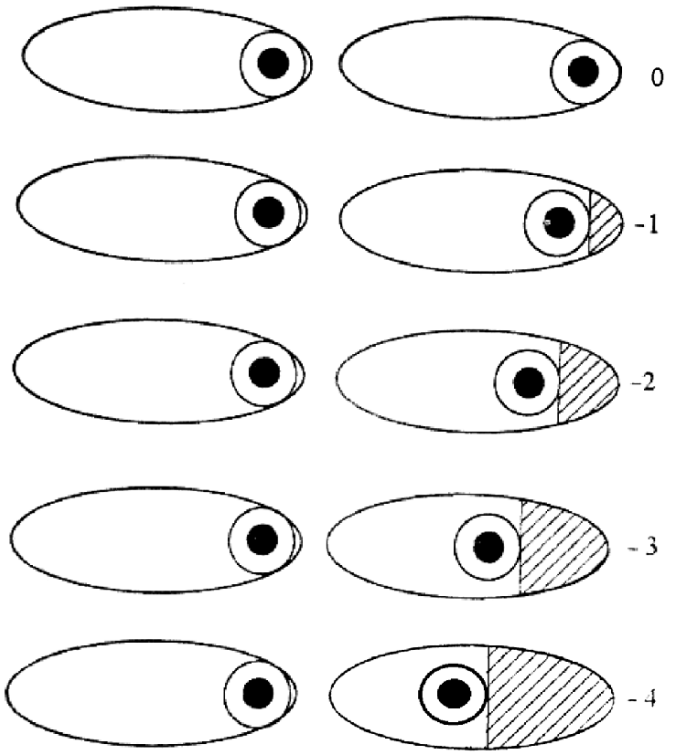
BINOCULAR DIPLOPIA
WHERE TO START

- 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

https://www.researchgate.net/figure/Drawing-and-designation-in-different-grades-of-abduction-deficit-No-abduction-deficit_fig1_323798591

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

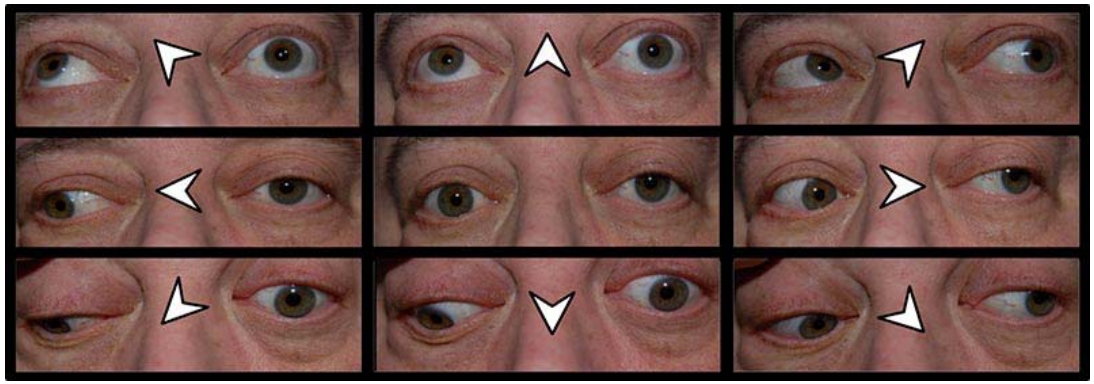
- Optical
- CNS
- Orbital
- Systemic
- Binocular Vision Disorders

BINOCULAR DIPLOPIA

- Optical
  - Induced or unwanted prism

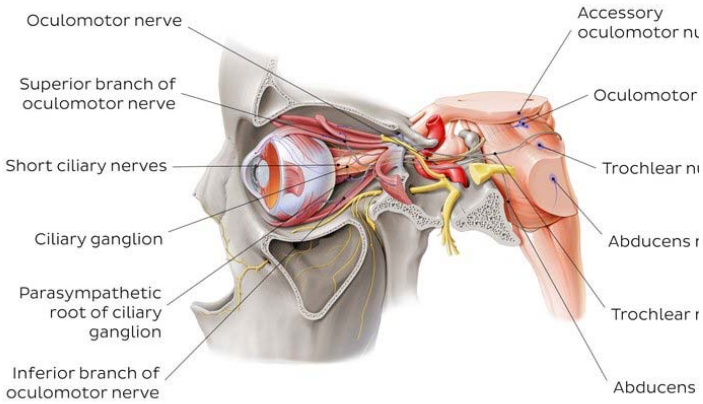
THIRD NERVE PALSY
SIGNS AND SYMPTOMS

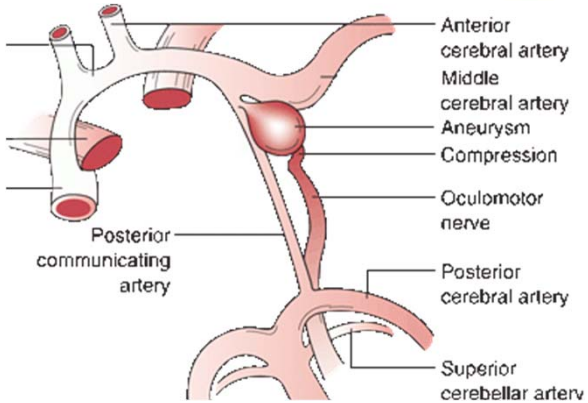
- Binocular Diplopia
- Ptosis
- Exotropia and hypotropia
  - Down and out
  - Limitation of all fields of gaze except temporally
- Pupil sparing/involved
- With or without pain
- Complete and partial



https://eyerounds.org/cases-i/case156/1-partial-eyelid-ptosis-adduction-supraduction-infraduction.jpg

THIRD NERVE PALSY
CN III PATHWAY



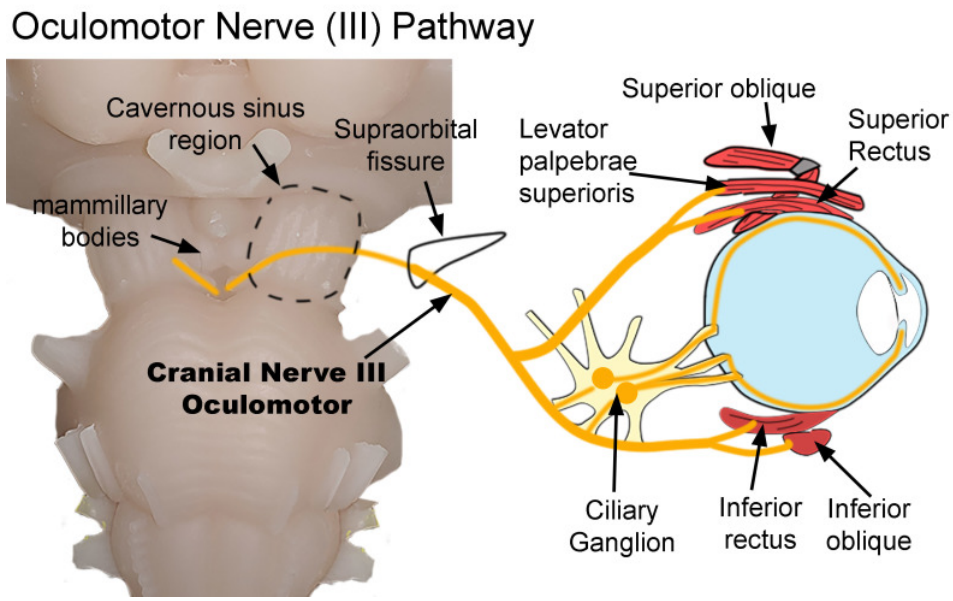


- Parasympathetic fibers to pupil on outside of nerve making them more susceptible to compression

https://www.kenhub.com/en/library/anatomy/the-oculomotor-nerve
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



https://www.ncbi.nlm.nih.gov/books/NBK549919/figure/article-43127.image.f3/?report=objectonly

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

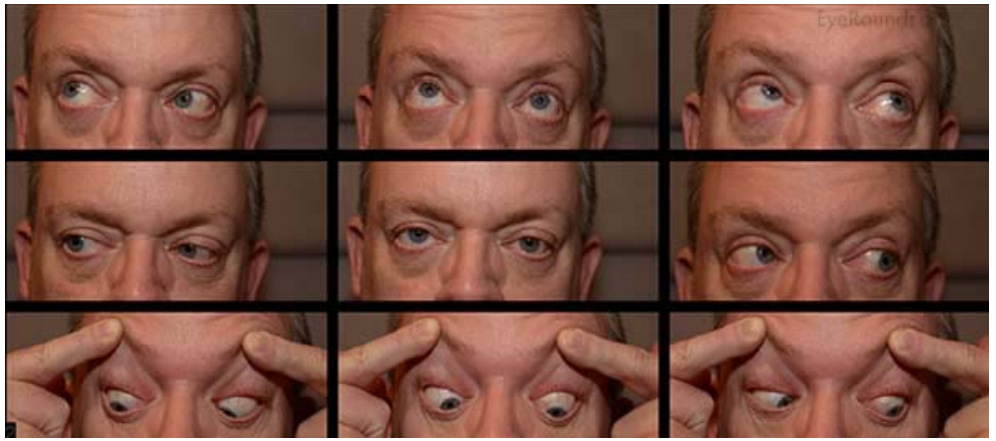
- Directed by neurology and/or neurosurgery

FOURTH NERVE PALSY
SIGNS AND SYMPTOMS

- Binocular, vertical diplopia
- Dizziness
- Difficulty Reading

- Hypertropia on affected side
  - Worse in downgaze

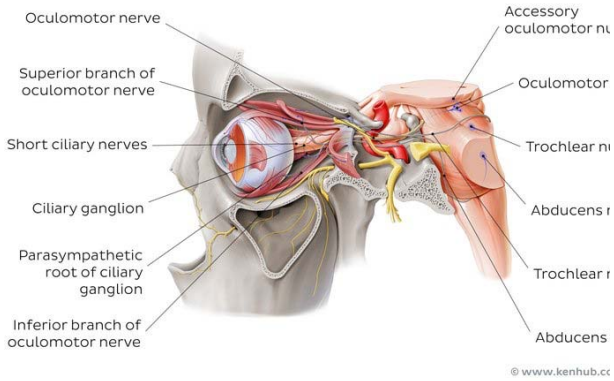
- Motility can look normal
  - Limited downgaze when adducted

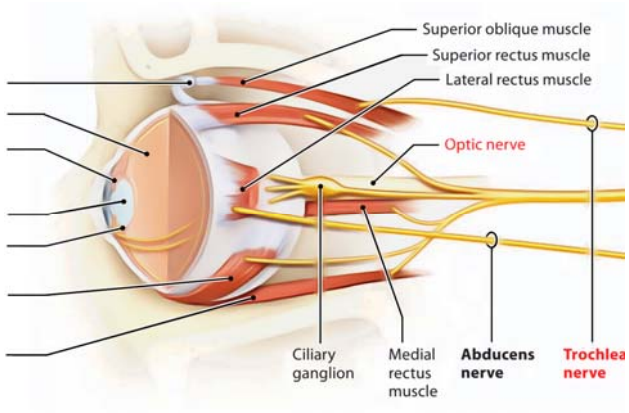


https://webeye.ophth.uiowa.edu/eyeforum/atlas/pages/CN-IV-Palsy/index.htm

FOURTH NERVE PALSY

CN IV PATHWAY





- Longest intracranial pathway
- Exits from back of the brainstem
- Most vulnerable to trauma

FOURTH NERVE PALSY
ETIOLOGY

Common

- Trauma
- Microvascular/Ischemic

Uncommon

- Tumor
- Hydrocephalus
- Aneurysm
- GCA

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

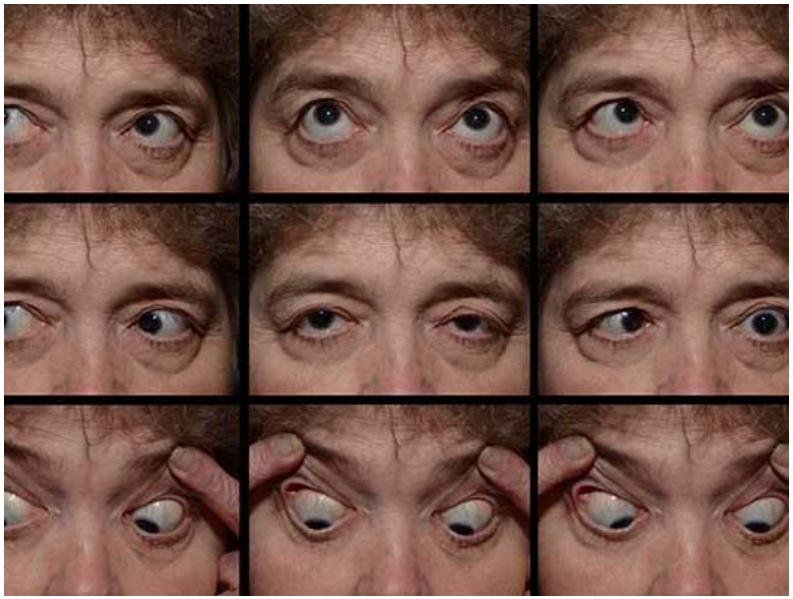
FOURTH NERVE PALSY
WORKUP

- 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

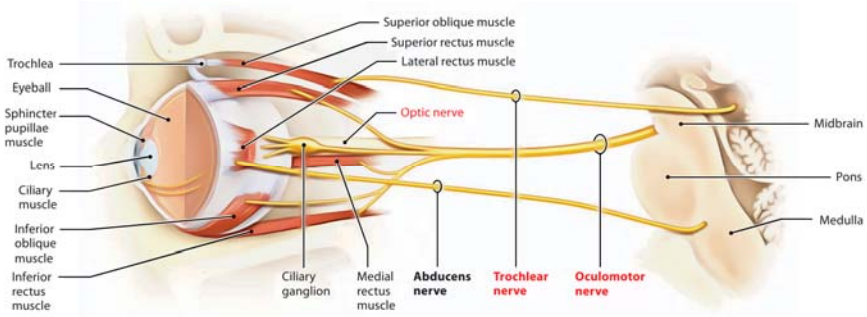
- Patching
- Prism
  - Fresnel
  - Wait 3-6 months for stabilization to prescribe
- Presumed vascular or idiopathic: 1-3 months
- Unresolved in 3 months – refer for imaging studies
- Additional neurological abnormalities – refer for imaging studies

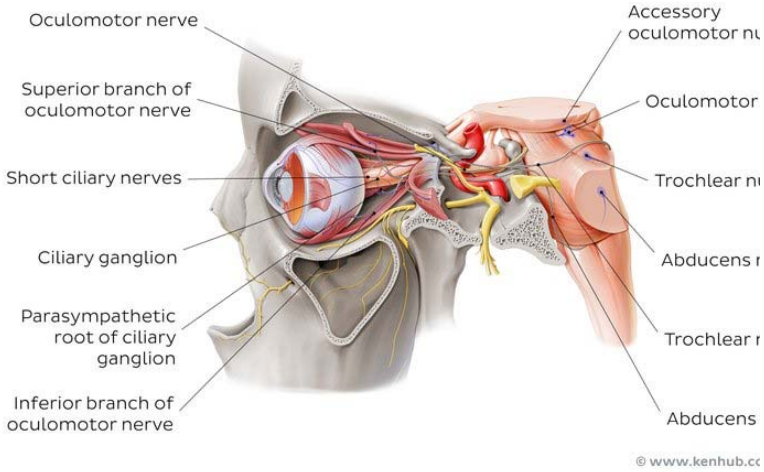
SIXTH NERVE PALSY
SIGNS AND SYMPTOMS



- 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

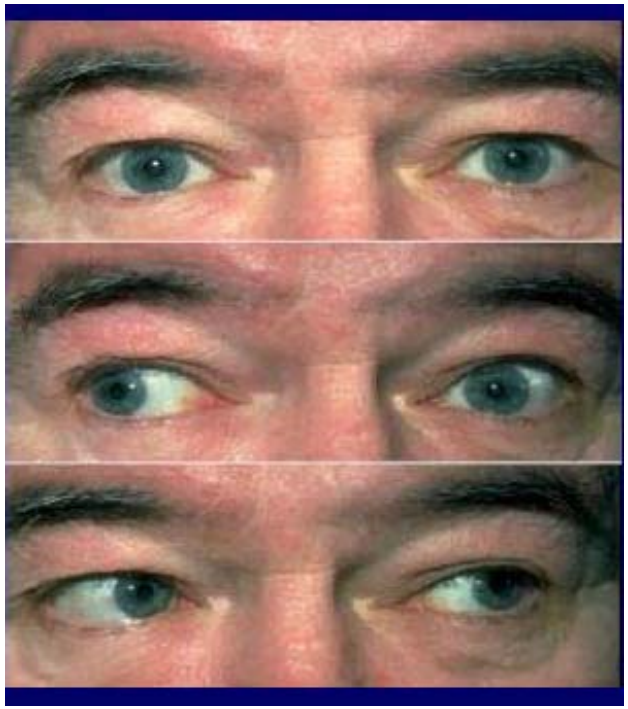
- MRI of the brain
  - <45 yo
  - 45-55 with no vascular risk factors
  - History of cancer?
  - Does not resolve after 3-6 months
- ESR, CRP, platelets for any suspicion of GCA

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

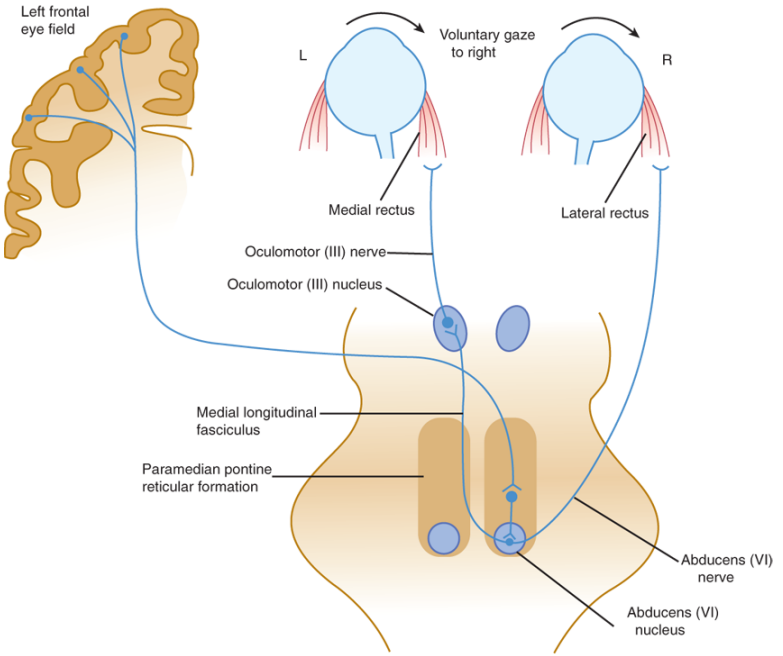


- Diplopia
  - Intermittent or constant
- Adduction deficit in affected eye
  - Partial to complete
  - Left INO = cannot left eye cannot adduct

https://www.slideserve.com/polly/neuro-ophthalmology

INTERNUCLEAR OPHTHALMOPLEGIA
MEDIAL LONGITUDINAL FASCICULUS

- 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



https://eyewiki.aao.org/Internuclear_Ophthalmoplegia

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

INO
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

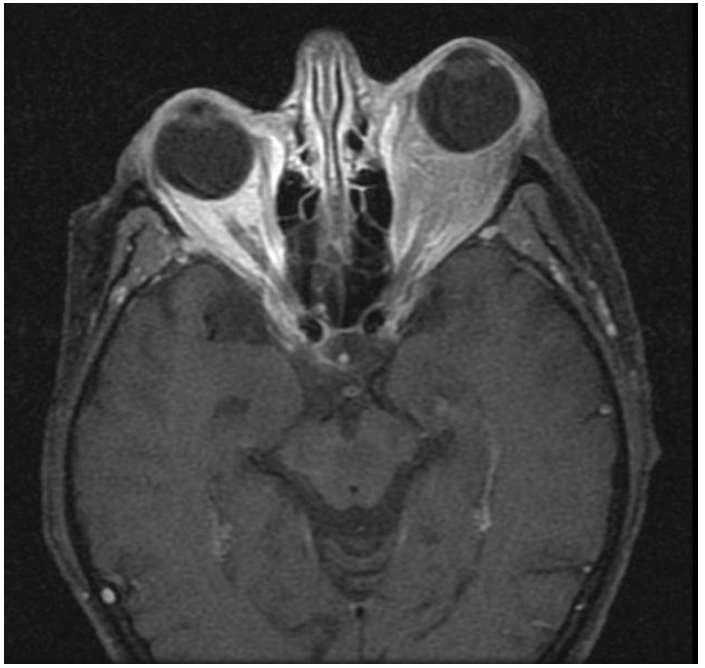
- Optical
- CNS
- Orbital
- Systemic
- Binocular Vision Disorders

BINOCULAR DIPLOPIA
ORBITAL ETIOLOGIES

- Orbital Tumors
- Orbital Pseudotumor
- EOM Restriction

BINOCULAR DIPLOPIA
ORBITAL ETIOLOGIES

- Tumor
  - Proptosis
  - Numbness or tingling around eye
  - Vision loss
  - Pain

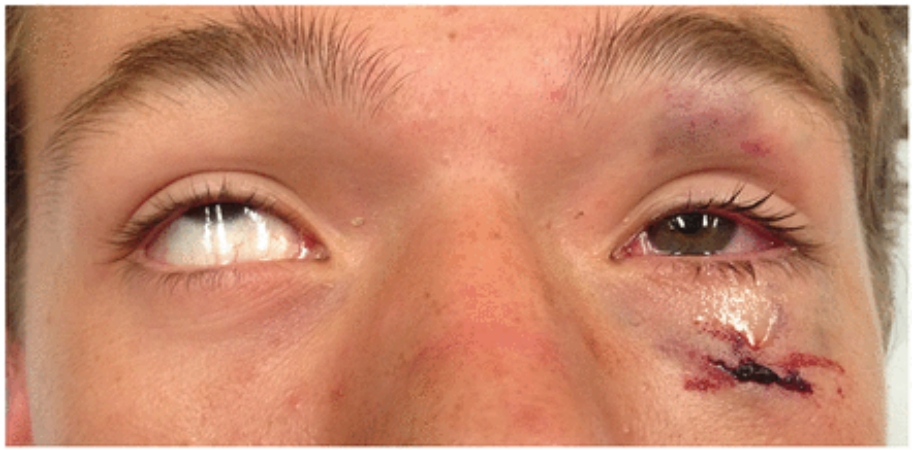


BINOCULAR DIPLOPIA
ORBITAL ETIOLOGIES

- 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 if 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 if affect the eyes
  - Headache
  - Episodes of vision loss
  - VF loss
  - Horizontal diplopia
  - Optic disc edema

INTRACRANIAL HYPERTENSION

- 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 ESOTROPIA

- 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