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 ETIOLOGY

- Refractive
- Optical
- Retinal

CC: Double vision

- 1. Does it go away when you cover one eye?
 - No (monocular)
- 2. Right, Left, both eyes?
- 3. Constant or intermittent?



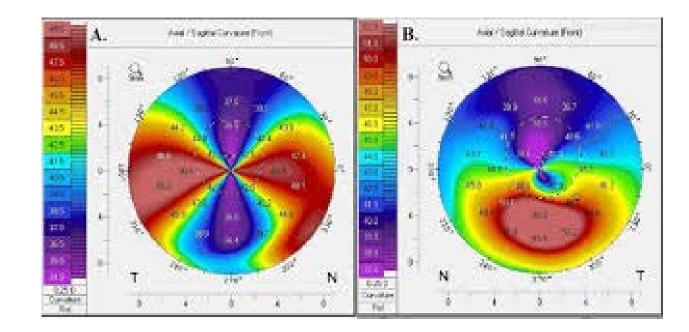
MONOCULAR DIPLOPIA ETIOLOGY

Refractive

- Optical
- Retinal

MONOCULAR DIPLOPIA REFRACTIVE

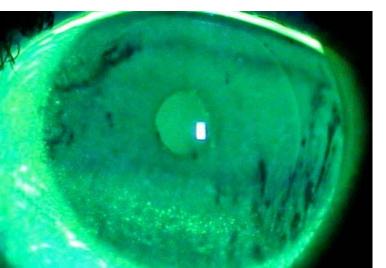
- Uncorrected refractive error
 - Regular astigmatism
- Anisometropia
- Irregular astigmatism
 - Diagnostic RGP
- Exam
 - Refraction
 - Topography



MONOCULAR DIPLOPIA ETIOLOGY

- Refractive
- Optical
- Retinal





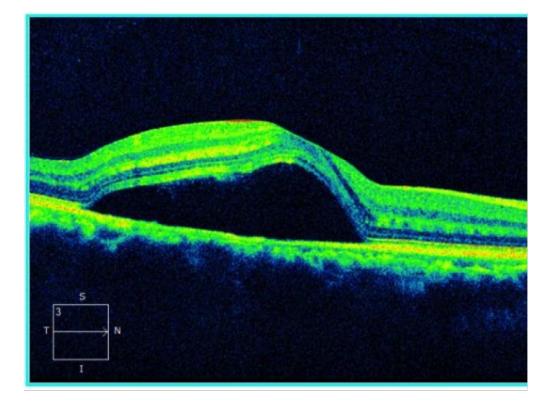
MONOCULAR DIPLOPIA

- Optical Irregularities
 - Dry eye
 - Intermittent
 - Corneal Opacities
 - Diagnostic RGP
 - Iris/pupil
 - Cosmetic contact lens
 - Cataract

MONOCULAR DIPLOPIA ETIOLOGY

- Refractive
- Optical
- Retinal

MONOCULAR DIPLOPIA



• Retina

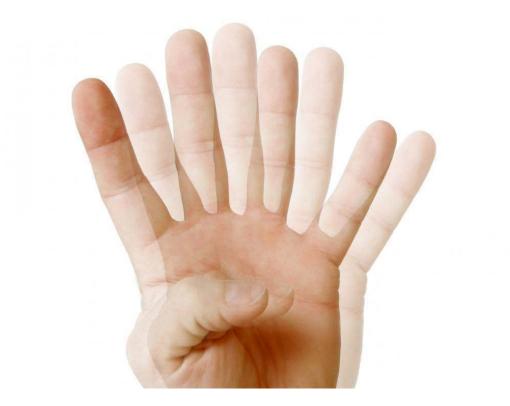
- Macular Edema
- Central Serous
- Exudative ARMD

DOUBLE TROUBLE

CC: Double vision

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

- Yes: Binocular diplopia
- No: Monocular diplopia



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

- 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

- Systemic History
 - Vascular disease: HTN, HLD, DM
 - Graves Disease
 - Multiple sclerosis
 - Myasthenia gravis

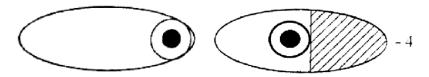
- Acuity
- Motility
- Binocular Function
- Stereo
- Refraction
- Anterior and Posterior Segment
 - Signs of vascular risk factors
 - Optic disc edema or pallor











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

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



Optical

- CNS
- Orbital
- Systemic
- Binocular Vision Disorders

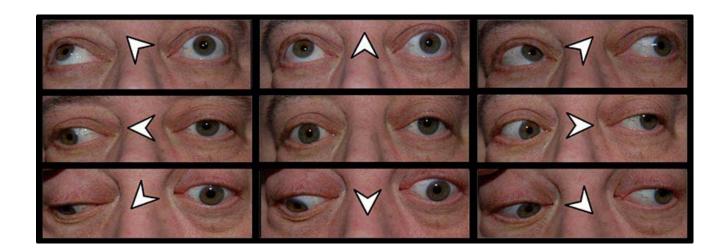


- Optical
 - Induced or unwanted prism

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

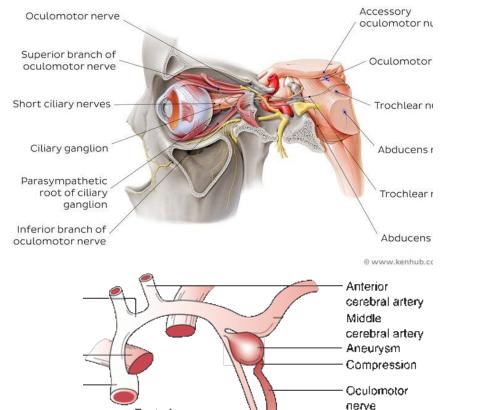
THIRD NERVE PALSY SIGNS AND SYMPTOMS

- 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



THIRD NERVE PALSY CN III PATHWAY

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



Posterior

Superior cerebellar artery

cerebral artery

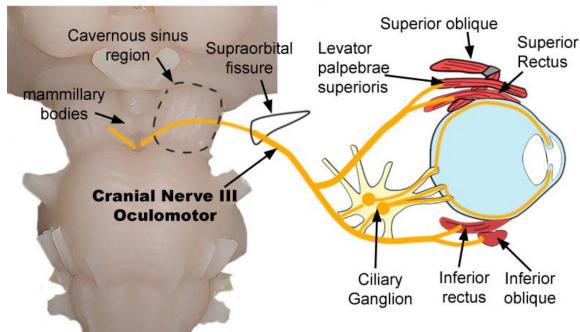
Posterior communicating

artery

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

Oculomotor Nerve (III) Pathway



THIRD NERVE PALSY ETIOLOGY

PUPIL SPARING

- More common:
 - Ischemic/microvascular
- Less common
 - Cavernous sinus disease
 - GCA

PUPIL INVOLVED

- Aneurysm
- Tumor
- Trauma
- Cavernous sinus disease
- Ischemia

THIRD NERVE PALSY MANAGEMENT

PUPIL SPARING

- >50yo w/ vascular risk factor
 - close observation
- <50yo
 - Neuroimaging
- Management of vascular risk factors
- 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

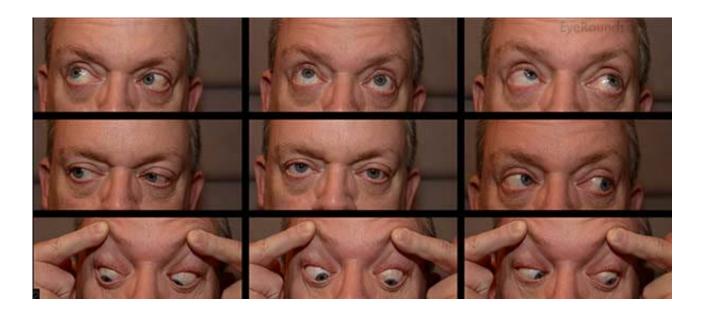
- Occlusion patch
- Prism
- Pupil involvement can be delayed 5-7 days
- If ischemic...
 - Improvement in 4-8 weeks
 - Resolution in 3-6 months
- Strabismus surgery if deviation stable after 6 months

PUPIL INVOLVED

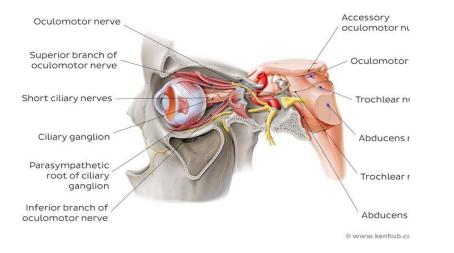
 Directed by neurology and/or neurosurgery

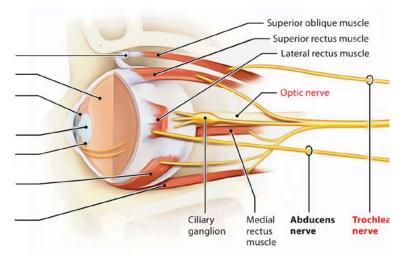
FOURTH NERVE PALSY SIGNS AND SYMPTOMS

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



FOURTH NERVE PALSY CN IV PATHWAY





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

FOURTH NERVE PALSY ETIOLOGY

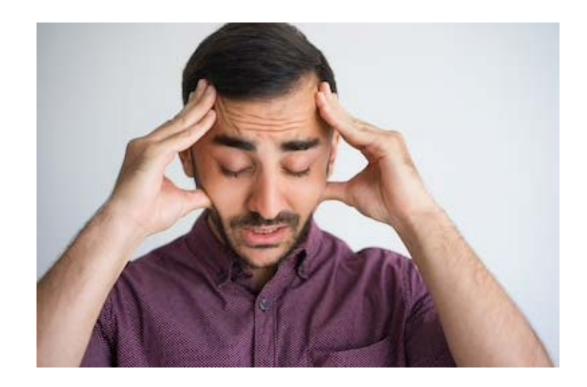
Uncommon

Common

- Trauma
- Microvascular/Ischemic

- Tumor
- Hydrocephalus
- Aneurysm
- GCA

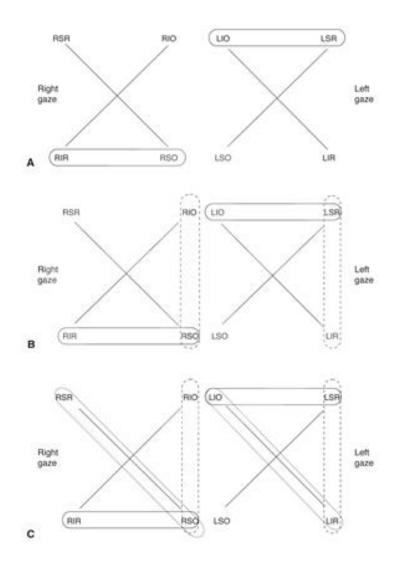
- Three Step Test
 - Process to determine affected EOM in cases of vertical tropias
 - 8 possible muscles responsible for vertical tropia
 - Superior and inferior recti
 - Superior and inferior obliques



- Three-Step Test:
 - 1. Determine which eye has hypertropia in primary gaze
 - 2. Determine whether hypertropia is greater when looking left or right
 - 3. Determine whether hypertropia is greater when tilting head to right or left

• Right IV palsy has a right hypertropia worse in left gaze and right head tilt

- Right Hypertropia
 - Weakness in right depressors (RIR, RSO)
 - Weakness in left elevators (LIO, LSR)
- ...Worse in left gaze
 - Weakness in RSO or LSR
- ... Worse with right head tilt
 - Weakness of RSO



- Workup by PCP/internist
 - DM, HTN, HLD
- 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

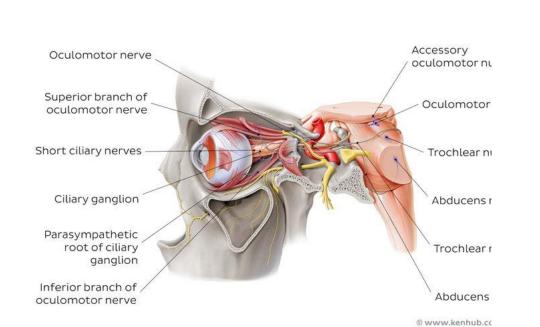
SIXTH NERVE PALSY SIGNS AND SYMPTOMS



- 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



Trochlear

nerve

Oculo

nerve

Superior oblique muscle - Superior rectus muscle — Lateral rectus muscle

Midbrain

Pons

edulla

Optic nerve

Abducens

nerve

Ciliary

ganglion

Medial

rectus

muscle

Trochlea Eyeball

Sphincter

Ciliary

muscle

Inferior oblique

muscle Inferior

rectus

muscle

pupillae muscle Lens

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 Prism
 - Wait 3-6 months for stabilization
- Re-examine every 4-6 weeks
- Strabismus surgery if large angle remains

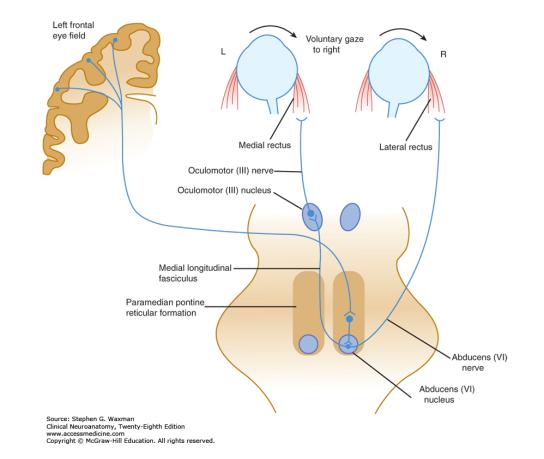
INTERNUCLEAR OPHTHALMOPLEGIA (INO) SIGNS AND SYMPTOMS

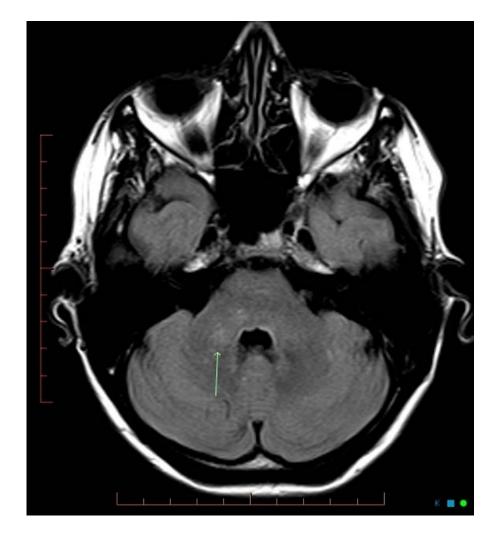


- Diplopia
 - Intermittent or constant
- Adduction deficit in affected eye
 - Partial to complete
 - Left INO = cannot left eye cannot adduct

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





INO etiology

- ElderlyStroke
- Young
 - Multiple Sclerosis



• 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 Syndrome
 - 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 Syndrome
 - 1 ½ plus a VII palsy

SKEW DEVIATIONS



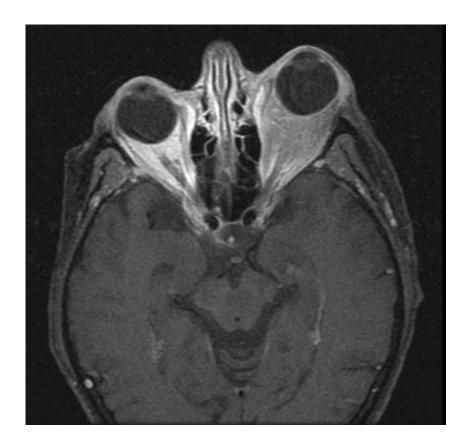
- Vertical deviation not due to any single muscle or nerve
- Damage to brainstem or cerebellum
 - Damage to vestibular nerve inputs to brain stem
- Usually caused by stroke
 - multiple sclerosis
 - Inflammation
 - Trauma
 - Tumor
- Usually accompanied by other neurological symptoms
- Need to differentiate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

BINOCULAR DIPLOPIA

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

- Orbital Tumors
- Orbital Pseudotumor
- EOM Restriction

- 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



• EOM Restriction

- Trauma
 - Entrapment
- Age
 - Functional
 - Decrease in accommodative response
 - Structural
 - Degeneration of pully system



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
 - Decompensated congenital IV palsy
 - Normally have full EOM's

WHEN WORKUP IS NEEDED...

- Cranial Nerve Palsies
 - CN III Palsy with pupil involvement
 URGENT!!
 - 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

RESOURCES

Bagheri, N & Wajda, B (Eds.). (2017) The Wills Eye Manual (7th ed.). Baltimore, MD: Wolters Kluwer. Boisvert, C.J. (2016, March 4). Multiple Sclerosis. Retrieved from https://eyewiki.org/Multiple_sclerosis Gupta, A. et al. (2019). Cranial Nerve 4 Palsy. Retrived from https://eyewiki.org/Cranial_Nerve_4_Palsy. Colon-Acevedo, Betsy. (2020). Aquired Ocuomotor Nerve Palsy. Retrieved from https://eyewiki.org/Acquired_Oculomotor_Nerve_Palsy. DelGiodice, M. and Trottini, M. (2017). Every Picture Tells a Story. Review of Optometry. July 2017. Triantafilou, D. et al. (2020). Abducens Nerve Palsy. Retrieved from https://eyewiki.org/Abducens_nerve_palsy. Freedman, Kenn. DDx. May 2013. PowerPoint Presentation. https://webeye.ophth.uiowa.edu/eyeforum/tutorials/Corneal-Imaging/Fig6-LRG.jpg https://www.eyecentre.com.au/wp-content/uploads/2017/04/OCT-of-central-serous-chorioretinopathy.png https://everounds.org/cases-i/case156/1-partial-evelid-ptosis-adduction-supraduction-infraduction.jpg https://www.kenhub.com/en/library/anatomy/the-oculomotor-nerve https://myneurosurg.com/cranial-anatomy/cranial-nerve-3-oculomotor-nerve/ https://webeye.ophth.uiowa.edu/eyeforum/atlas/pages/CN-IV-Palsy/index.htm Karpecki, Paul. (2016). Practice Pearl of the Week. Review of Optometry. April 2016 Koetting, C. and Mangan, R. (2017) Find the Nerve to Fight Diplopia. Review of Optometry. April 2017 Nitescu, Raluca. (2016). Age Related Strabismus. Romanian Journal of Ophthalmology. Vol 60, Issue 2, Apr-Jun 2016 54-58. Noel, Marie-Eve C. (2019). Myasthenia Gravis. Retrieved from https://eyewiki.org/Myasthenia_Gravis. Rodriguez, S. et al. (2020). Skew Deviation. Retrieved from https://eyewiki.org/Skew_deviation. Saenz, B. et al (2019). Thyroid Eye Disease In Your Exam Lane. Review of Optometry. October 2019. Swisher, J. et al. (2018). Internuclear Ophthalmoplegia. Retrieved from https://eyewiki.org/Internuclear_Ophthalmoplegia.