Neuro-Ophthalmology:

The evaluation and treatment of conditions affecting the brain, optic nerve, oculomotor system, and pupil.

Table of Contents

This handbook presents the discipline of neuro-ophthalmology in an organized and simplified format.

Mohawk Valley Retina has been lecturing on and caring for patients with neuro-ophthalmological disease for decades. We hope this monograph is useful in your management of these patients.

Please let us know if you need any assistance.

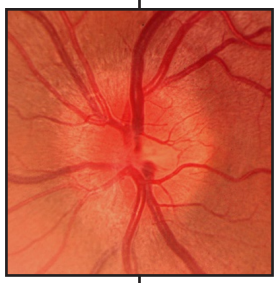
Team MVR

MVR

Mohawk Valley Retina

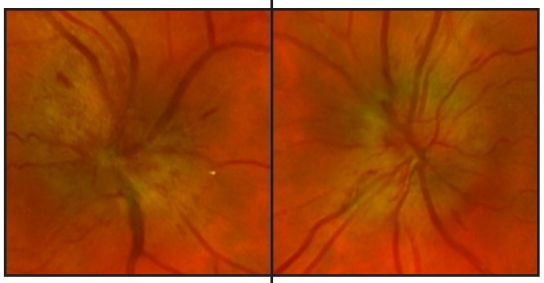
Optic Neuropathy

Typical Optic Neuritis:
MS



- Monocular
- Painful
- APD
- Obvious

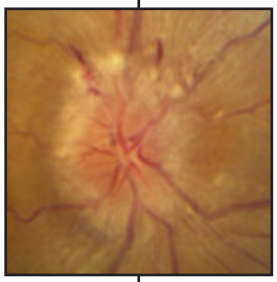
Vasculopathic:
HTN, DM, Lupus



- Blood, lipid, CWS
- Asymmetric
- Bilateral (usually)
- Uncontrolled systemic disease (always)

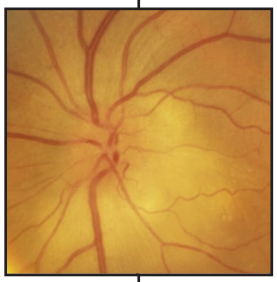
Atypical Optic Neuropathy

Infectious:
HSV, VZV, bartonella, syphilis, Lyme, tuberculosis, toxo, fungus



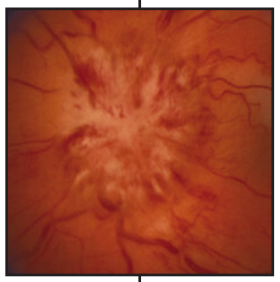
- Vitritis, vasculitis, retinitis commonly associated

Inflammatory:
Sarcoid, Lupus, HLA B27, Behçet's, Wegener's



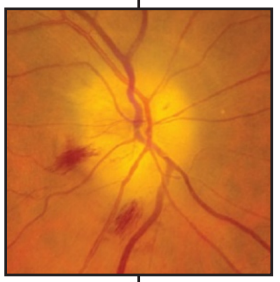
- Adjacent inflammation
- Vasculitis common
- Vitritis, retinitis less common

Infiltrative:
Leukemia, lymphoma, metastasis



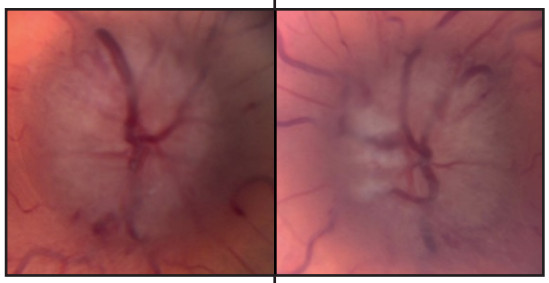
- Systemic disease
- Inflammation usually absent

Ischemic:
NAAION, AAION



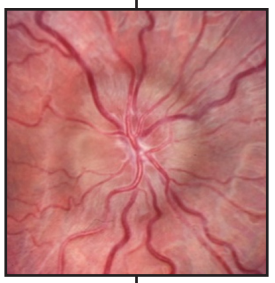
- Pallid edema
- CWS, lipid, heme
- Vasculopathic risk factors
- Inflammation absent

Papilledema:
Pseudotumor, tumor



- Bilateral
- Symmetric
- Swollen
- Increased ICP

Mechanical:
Compression, VP traction, ONH drusen



- Benign (unless compressive)

Optic Neuropathy: Acute disorders of the optic nerve.

Typical Optic Neuritis

Signs & Symptoms
- Monocular vision loss with spontaneous recovery, pain with movement, APD, VF deficit, swollen/inflamed nerve

Causes
- MS

Workup
- OCT, FA, HVF
- MRI brain (≈50% affected), MRI orbits (≈80% affected)
- LP: normal opening pressure, increased immunoglobulins, oligoclonal bands in 30-60%
- Labs: anti-AQP4, anti-MOG

Treatment
- Does not alter visual outcome, therefor use limited to decrease MS progression
- No CNS involvement on MRI: no benefit
- MRI ≥ 2 lesions: 1 gr IV methylprednisolone x 3 days followed by oral prednisone (1 mg/kg) for 14 days → Decreases clinical MS in 2 years from 36% to 16% → By 3 years benefit diminishes
- Avonex (beta interferon) injection may decrease or delay MS

Atypical Optic Neuropathy

Signs & Symptoms
- Monocular vision loss, non-resolving, painless, APD, VF deficit, swollen/inflamed nerve, adjacent inflammation

Causes
- Herpes, bartonella, syphilis, Lyme, tuberculosis, toxoplasmosis, fungus, sarcoidosis, Lupus, Crohn's, ulcerative colitis, lymphoma, Wegener's, Behçet's, AION, GCA

Workup
- OCT, FA, HVF ± VEP ± MRI
- Labs: CBC, ESR, ACE, ANA, RF, RPR, FTA-ABS, lysozyme, toxoplasma, Lyme, HSV 1 & 2, VZV, QuantiFERON, CMV, HLA A, HLA B, Toxocara, HIV, ionized calcium, SSA/SSB, bartonella, ANCA, Leptospira, HLTV
- LP in extreme cases

Treatment
- Systemic/intravitreal antiviral, antibiotic, antifungal, antiparasitic, anti-inflammatory, anti-VEGF

Ischemic Optic Neuropathy

Signs & Symptoms
- Monocular vision loss, painless, APD, altitudinal VF deficit, eventual pallor

Causes
- Arterial sclerosis (NAAION), temporal arteritis (AAION)

Workup
- OCT, FA, HVF
- BP, CBC, ESR, CRP, TABx, consider MRI, other labs if atypical optic neuritis considered

Treatment
- Non-Arteritic: control BP, aspirin, smoking cessation
- Arteritic: methylprednisolone 1-2 g/d for 3-5 days or/then prednisone 40-150 mg/day – taper prednisone 4-12+ months based on Sx, ESR, CRP

Vasculopathic Optic Neuropathy

Signs & Symptoms
- Vision loss variable (in part due to macular ischemia/edema), painless, no APD (usually), bilateral (usually), hyperemic, hemorrhagic with lipid, vascular findings predominate

Causes
- Hypertension, diabetes, Lupus

Workup
- OCT, FA, HVF
- BP, CBC, blood glucose, HgA1c, ANA, MRI/LP if uncertain

Treatment
- Control systemic disease, intravitreal anti-VEGF and/or Kenalog, laser

Papilledema

Signs & Symptoms
- Mild vision loss, headache, nausea, enlarged blind spot on HVF, bilateral optic nerve fullness, axoplasmic congestion, hemorrhage, exudates, eventual pallor

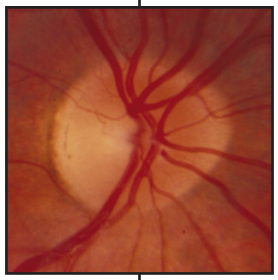
Causes
- Increased intracranial pressure: pseudotumor cerebri, tumor, venous sinus thrombosis, hydrocephalus, meningitis

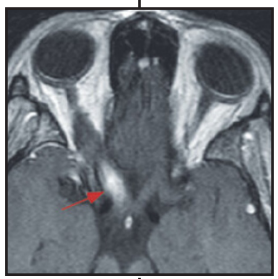
Workup
- OCT, FA, HVF
- MRI/MRV (flattening of posterior sclera 80%, empty sella 70%, perioptic subarachnoid space distension 50%, prelaminar disc protrusion 30%), lumbar puncture (increased ICP > 20 cm H_2O, normal CSF composition)

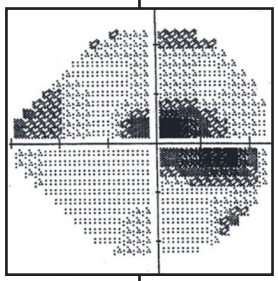
Treatment
- Observation with weight loss, CAI, repeat LPs
- Lumboperitoneal shunt, ON sheath fenestration in extreme cases

Chronic Optic Neuropathy

Compressive

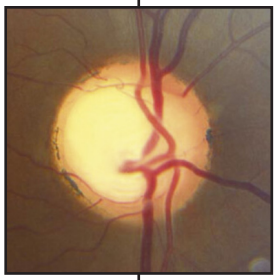


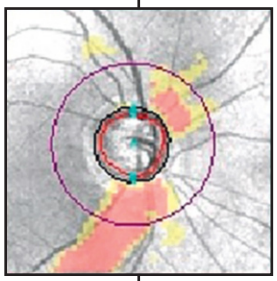


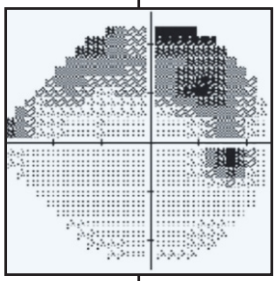


- Slow, progressive vision loss
- Unilateral/bilateral
- Any pattern of VF loss
- Color vision usually decreased

Glaucoma

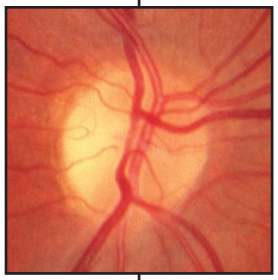


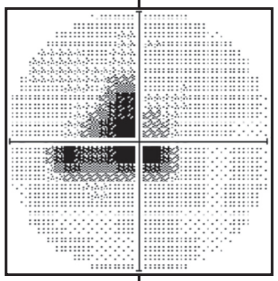


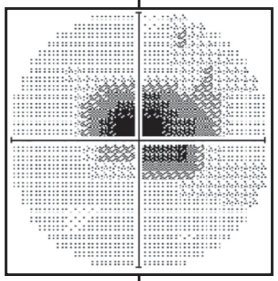


- VA and color vision normal (unless advanced disease)
- Disc cupping
- "Discs match the fields"

Nutritional/Toxic

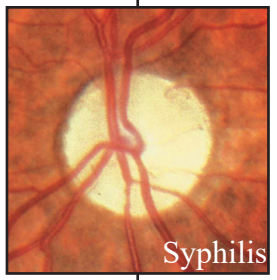


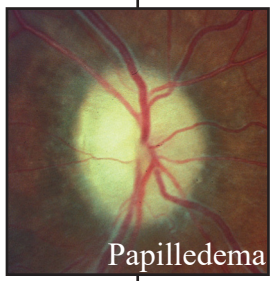


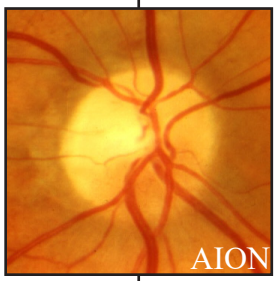


- Slow, progressive vision loss
- Bilateral
- Central or cecocentral scotomas
- Early color vision loss

Prior Acute Optic Neuropathy

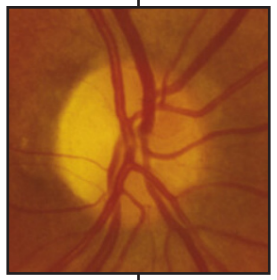


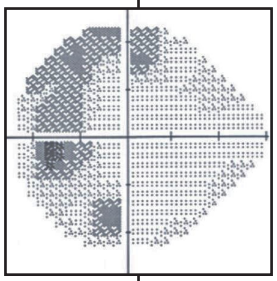


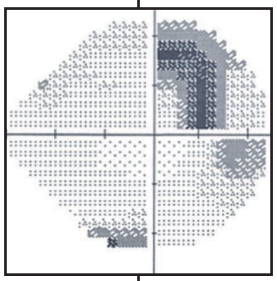


- Old (usually symptomatic) episode
- Unilateral/bilateral
- Pale disc

Hereditary







- Slow, progressive vision loss (except LHON)
- Bilateral
- Central, paracentral, cecocentral scotomas
- Color vision decreased

Chronic Optic Neuropathy: Peristant optic nerve disease resulting in atrophy.

Compressive Optic Neuropathy

Signs & Symptoms
- Slowly progressive vision loss over months, visual field loss, monocular/binocular, acuity variably decreased, color vision usually decreased

Causes
- Tumor, aneurysm, thyroid eye disease

Workup
- OCT, FA, HVF
- MRI brain/orbits with contrast

Treatment
- Neurosurgical
- Orbital decompression

Glaucomatous Optic Neuropathy

Signs & Symptoms
- Asymptomatic until very advanced, normal color vision unless advanced, elevation of IOP, disc cupping, NFL loss, family hx
- “Discs match the fields” - visual field defect can be matched with focal thinning or notch in the neuroretinal rim

Workup
- OCT, HVF

Treatment
- Topical or surgical therapy

Prior Acute Optic Neuropathy

Signs & Symptoms
- Old episode, monocular/binocular, nerve(s) pale, APD possible

Causes
- MS, vasculopathy, infection, infiltration, ischemia, papilledema (see pages 1-2)

Nutritional/Toxic Optic Neuropathy

Signs & Symptoms
- Slowly progressive bilateral vision loss, color vision deficit, temporal or diffuse nerve pallor, central or cecocentral scotoma

Causes
- Nutritional: vitamin deficiency (B12, B1, folate, copper) due to poor diet, alcoholism, or bariatric surgery
- Toxic: tobacco/substance abuse, medications (ethambutol, isoniazid, linezolid, streptomycin, chloramphenicol, chlorpropamide, ethchlorvynol, disulfiram), heavy metals (lead, cobalt, chromium, thallium), ingestion or inhalation (methanol, ethylene glycol, carbon monoxide, toluene)

Workup
- Careful history
- Color plates, OCT, FA, HVF, ERG, MRI
- Labwork: CBC, serum B12, red blood cell folate, whole blood thiamine, heavy metal screening, ADOA/LHON mutation, urinalysis

Treatment
- Eliminate any causative agents
- Vitamin supplementation

Hereditary Optic Neuropathy

Signs & Symptoms
- Autosomal Dominant Optic Atrophy (ADOA): bilateral, symmetric, slowly progressive mild to moderate vision loss, color vision deficit, temporal nerve pallor, central, paracentral, or cecocentral field loss
- Leber Hereditary Optic Neuropathy (LHON): acute or subacute severe vision loss, bilateral sequential (within weeks or months) or simultaneous, color vision deficit, nerves initially appear normal or hyperemic, nerve pallor develops in several weeks, dense central or cecocentral field loss

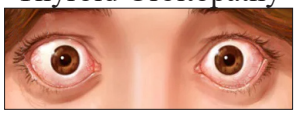
Causes
- ADOA: OPA1 gene mutation
- LHON: mutations in mitochondrial DNA, sites 11778, 14484, and 3460

Workup
- OCT, FA, HVF
- Genetic evaluation
- Consider other etiologies if no family history

Treatment
- None
- LHON: Cardiology consult due to increased incidence of cardiac conduction defects

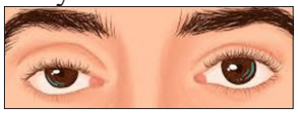
Binocular Diplopia

Muscle/Orbit Disease:
Restrictive Myopathy, Paretic Myopathy, Acute Orbitopathy

Thyroid Orbitopathy


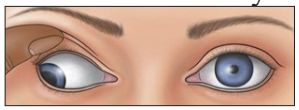
- Female > male
- Injection/exophthalmos
- Lid retraction
- IR > MR > SR
- Hypo/eso

Neuromuscular Junction Disease:
Myasthenia Gravis

Myasthenia Gravis


- Variable
- Fatiguable
- Ptosis

Nerve Disease:
Third Nerve Palsy, Fourth Nerve Palsy, Sixth Nerve Palsy, Unilateral Multiple Nerve Palsies, Bilateral Multiple Nerve Palsies

Third Nerve Palsy


- Any except esotropia
- Pupil enlarged (non-ischemic)

Causes: ischemia, compression, inflammation, infection

Fourth Nerve Palsy


- Vertical diplopia
- Hypertropia, increases with ipsilateral head tilt or nasal gaze

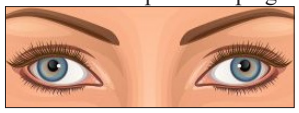
Causes: ischemia, congenital, trauma

Sixth Nerve Palsy


- Horizontal diplopia
- Reduced abduction

Causes: ischemia, compression, increased ICP, trauma, infiltrative, infectious

Brain Disease:
Internuclear Ophthalmoplegia, Skew Deviation, Supranuclear Ophthalmoplegias

Internuclear Ophthalmoplegia


- Horizontal diplopia
- Exotropia
- Weakness of adduction

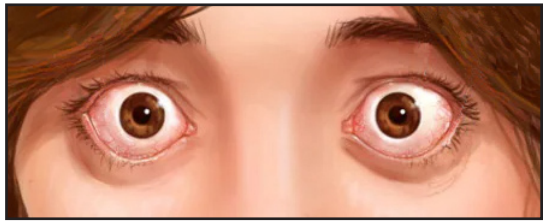
Other:
Decompensated Childhood Strabismus, Decompensated Phoria, Convergence Insufficiency, Convergence Spasm, Divergence Paralysis

Decompensated Phoria

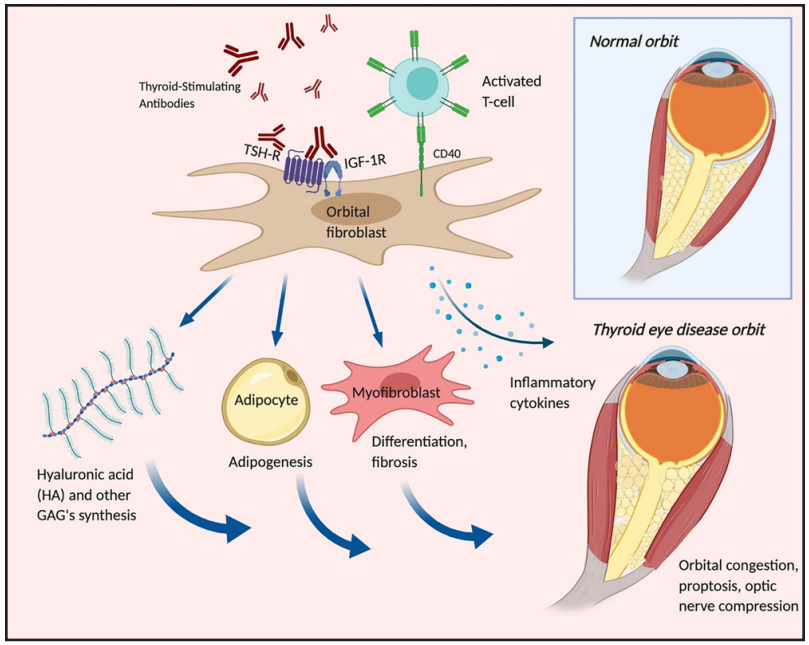

- Horizontal diplopia, initially intermittent

Thyroid Orbitopathy:

An autoimmune disorder characterized by enlargement of the extraocular muscles and an increase in orbital fat volume.



Signs & Symptoms
- Lid retraction with lid lag in downgaze
- Proptosis - unilateral or bilateral
- Orbital congestion with periorbital edema, puffiness of the eyelids, chemosis (all worse in the morning)
- Diplopia with restricted eye movements (restricted elevation and esotropia most common)
- Injection of blood vessels over insertion sites of horizontal rectus muscles
- FB sensation, redness, tearing, photophobia
- Elevated IOP (especially in upgaze)
- Decreased vision
- APD, reduced color vision, decreased visual field can be seen with compressive optic neuropathy



Causes
- Stimulatory auto antibodies leading to fibroblast activation, infiltration of T helper cells, B lymphocytes, macrophages and mast cells
- Inflammation/infiltration of extraocular muscles with extracellular matrix components and inflammogenic molecules

Differential Diagnosis
- Third nerve palsy with aberrant regeneration, myasthenia gravis, orbital pseudotumor, Parinaud's syndrome

Workup
- Forced ductions
- Hertel ophthalmometry
- Visual field if optic nerve involvement suspected
- MRI/CT of orbit
- Blood tests: thyroid-stimulating immunoglobulin, thyroid autoantibody tests (thyrotropin-receptor antibody, thyroid peroxidase antibody, thyroglobulin antibody), thyroid function tests (TSH, free T4, free T3)

Treatment
- Anti-inflammatory therapy with corticosteroids, methotrexate, or biological agents such as infliximab (Remicade), rituximab (Rituxan) or teprotumumab (Tepezza)
- Endocrinologic management of systemic thyroid disease
- Lubrication and nighttime eye lid taping
- Orbital decompression → strabismus surgery → eyelid surgery
- Orbital irradiation
- Smoking cessation

Myasthenia Gravis:

Acquired autoimmune disease in which antibodies bind to acetylcholine receptors at the postsynaptic neuromuscular junctions, resulting in weakness and rapid fatigue of skeletal muscle.

Signs & Symptoms

- Diplopia: variable and fatigable
- Ptosis: unilateral, bilateral, or alternating, fatigable
- Strabismus with any pattern of motility limitation
- Altered facial expression
- Difficulty speaking, swallowing, or breathing
- Weakness and fatigability in arms, legs, neck, and hands

Causes

- Autoimmune antibodies against: acetylcholine receptors, muscle-specific receptor tyrosine kinase (MuSK), and lipoprotein receptor related peptide 4 (LPR4)
  - Thymus gland is often implicated as the source
  - Idiopathic or drug-induced (penicillamine, aminoglycosides, beta blockers, chlorpromazine)

Differential Diagnosis

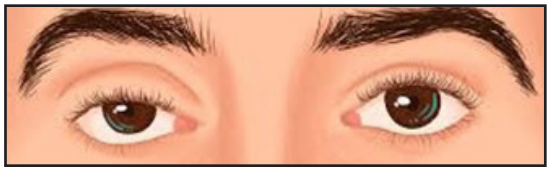
- Eaton-Lambert syndrome, chronic progressive external ophthalmoplegia, Kearns-Sayre syndrome, third nerve palsy

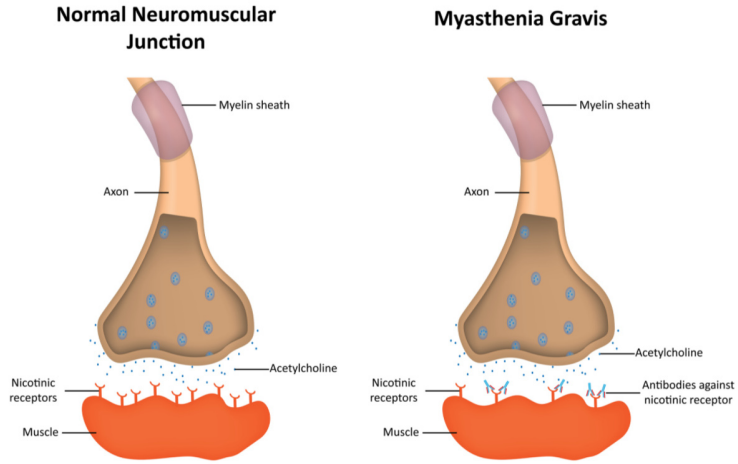
Workup

- Sleep/rest test: myasthenic ptosis and diplopia lessen with 30 minutes of rest
- Ice test: Ptosis improves following 2 minutes of ice application
- Tensilon (edrophonium) test: short-acting acetylcholinesterase inhibitor given intravenously; useful in patients with diplopia but no significant ptosis
- Prostigmin (neostigmine bromide) test: long-acting acetylcholinesterase inhibitor given intramuscularly; useful in patients with diplopia but no significant ptosis
- Blood tests: Acetylcholine receptor blocking and binding antibodies and MuSK antibodies
  - Detectable in 90% of generalized myasthenia cases, but only 50% in those with only ophthalmic symptoms
- CT or MRI of chest searching for thymus enlargement or thymoma

Treatment

- Pyridostigmine: inhibits acetylcholinesterase, stopping the breakdown of acetylcholine in the neuromuscular junction
- Immunosuppression: prednisone, azathioprine, mycophenolate mofetil, cyclosporine, or in select cases, monoclonal antibodies
- Thymectomy: indicated for those with thymoma, but may also improve symptoms in those without





In myasthenia gravis, AChR antibodies attack the acetylcholine receptors and prevent the acetylcholine neurotransmitter from binding to the muscle

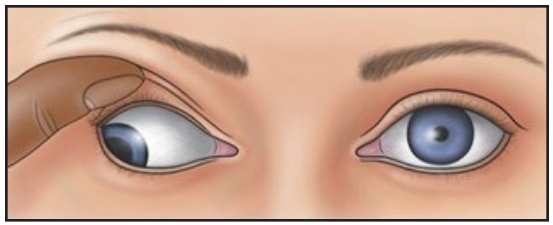
Myasthenia Gravis Palsy Criteria

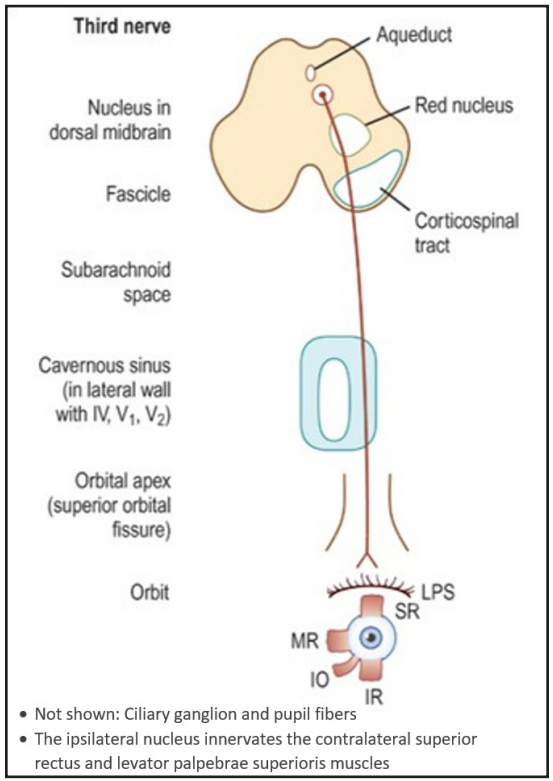
- No severe headache, tinnitus, or neurologic symptoms
- No pain
- Normal corneal and facial sensation
- Normal pupils and CVF

One or more of the following:

- Diplopia: variable in direction or severity from day to day, significantly worse at night or when tired
- Hoarse voice, problems swallowing, or problems breathing
- Ptosis appears or worsens on upgaze for 2 minutes
- Weakness of eyelid closure or facial muscles on testing
- Cogan lid twitch
- Improvement in ptosis and/or strabismus with ice test

Third Nerve Palsy: The third nerve innervates MR, SR, IR, eye lid, and pupil.





Signs & Symptoms
- Diplopia
- Palsy (any except esotropia)
- Pupil enlarged (non-ischemic)

Causes
- Ischemia (most common): HTN, DM, GCA
- Compression: aneurysm (1/3 of all third nerve palsies, can rupture within hours), tumor
- Inflammation: MS
- Infection: viral or post viral

Differential Diagnosis
- Partial third nerve palsy with pupil spared: myasthenia gravis, skew deviation, INO
- Complete third nerve palsy with pupil spared: myasthenia gravis

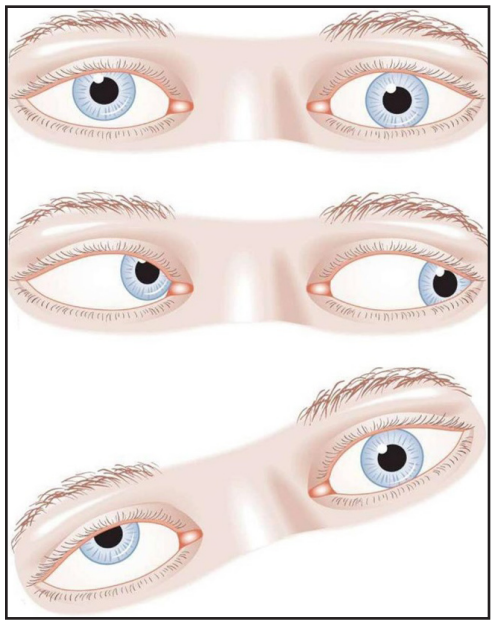
Workup
- Ischemic → observe → MRI/MRA if no improvement in 3 months or new neurologic signs/symptoms develop
- GCA suspected → ESR/CRP/CBC
- Ischemic criteria not met → urgent MRI/MRA

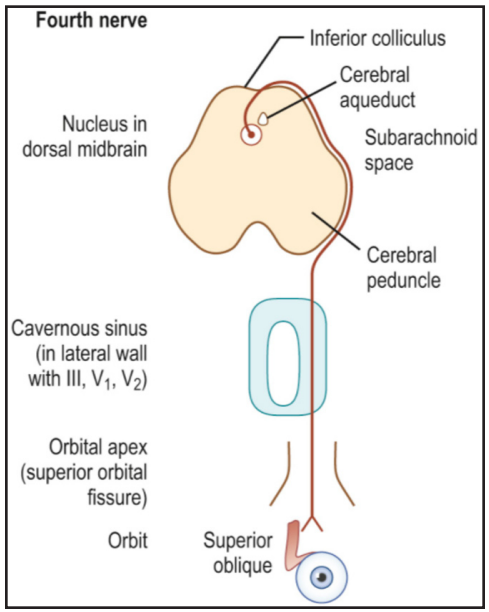
Treatment
- Occlusion, prism, strabismus surgery

Ischemic Third Nerve Palsy Criteria

- Over age 40
- One or more vasculopathic risk factors
- No h/o cancer, vasculitis, or autoimmune disease
- Sudden onset of diplopia and unilateral ptosis, with ptosis becoming complete in 24 hours
- No pain
- No other systemic neurologic symptoms
- Complete palsy: complete ptosis, no movement on attempted elevation, depression, or adduction
- Normal pupil: pupils are equal in size, no APD
- Fourth nerve function intact (intorsion of eye is seen on attempted depression in abduction)
- Sixth nerve function intact (full abduction)
- No signs of abberant regeneration (no changes in pupil size and no lifting of ptosis on attempted elevation, depression, or adduction)
- Other eye: normal motility and no ptosis
- Both eyes: normal VA, full CVF, no redness, no proptosis, no chemosis, normal intraocular exam (no iritis, vitritis, or optic nerve abnormalities)

Fourth Nerve Palsy: The fourth nerve innervates the superior oblique.





Signs & Symptoms
- Vertical diplopia: Effected eye hypertropic
  - Increases with nasal gaze
  - Increases with ipsilateral head tilt

Causes
- Ischemia: HTN, DM, GCA
- Congenital
- Trauma
- Infiltrative (rare): tumor, inflammatory, infectious

Differential Diagnosis
- Skew deviation, partial third nerve palsy, myasthenia gravis

Workup
- Congenital → observe
- Ischemic → observe → MRI if no improvement in 3 months
- GCA suspected → ESR/CRP/CBC
- Ischemic/congenital criteria not met (traumatic, bilateral) → MRI

Treatment
- Observe to resolution (ischemic: 3 months; traumatic: 6 months), strabismus surgery

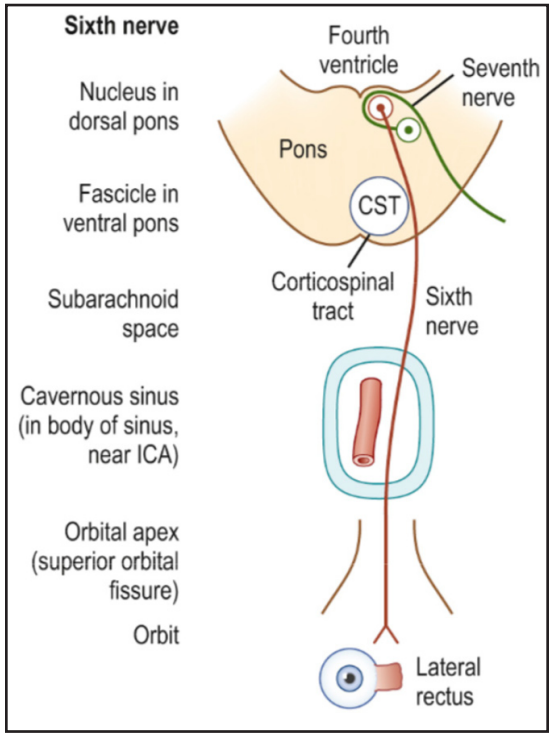
Ischemic Fourth Nerve Palsy Criteria

- Over age 40
- One or more vasculopathic risk factors
- No h/o cancer, vasculitis, or autoimmune disease
- Sudden onset of diplopia that does not vary
- No pain
- No other systemic neurologic symptoms
- Vertical or oblique deviation in primary position on cover test
- Hypertropia in primary position that increases with nasal gaze and on ipsilateral head tilt
- No change in deviation when assessed in sitting vs supine position (r/o skew deviation)
- Other eye: normal motility
- Both eyes: normal VA, full CVF, normal pupils, no ptosis, no redness, no proptosis, no chemosis, normal intraocular exam (no iritis, vitritis, or optic nerve abnormalities)
- Vertical prism fusional amplitude testing: less than 5 prism diopters
- Torsion testing: fundoscopy shows extorsion, double Maddox rod shows subjective extorsion of less than 10 degrees

Congenital Fourth Nerve Palsy Criteria

- Any age
- No h/o cancer, vasculitis, or autoimmune disease
- Evidence of head tilt from early childhood
- Diplopia with any timecourse (sudden/gradual, intermittent/persistent), no subjective torsion
- No pain
- No other systemic neurologic symptoms
- Vertical or oblique deviation in primary position on cover test
- Hypertropia in primary position that increases with nasal gaze and on ipsilateral head tilt
- Other eye: normal motility
- Both eyes: normal VA, full CVF, normal pupils, no ptosis, no redness, no proptosis, no chemosis, normal intraocular exam (no iritis, vitritis, or optic nerve abnormalities)
- Vertical prism fusional amplitude testing: greater than 5 prism diopters
- Torsion testing: fundoscopy shows extorsion, double Maddox rod shows no torsion or less than 10 degrees of extorsion

Sixth Nerve Palsy: The sixth nerve innervates the lateral rectus.



Signs & Symptoms
- Horizontal diplopia
- Reduced abduction

Causes
- Ischemia: HTN, DM, GCA
- Compressive: CCF, meningioma
- Infiltrative: Sarcoid, cancer
- Infectious (rare)
- Elevated ICP
- Trauma

Differential Diagnosis
- Myasthenia gravis, restrictive esotropia from "tight" medial rectus (thyroid eye disease), congenital Duane syndrome (absence of sixth nerve)

Workup
- Ischemic → observe → MRI if no improvement in 3 months
- GCA suspected → ESR/CRP/CBC
- Ischemic criteria not met/traumatic → MRI

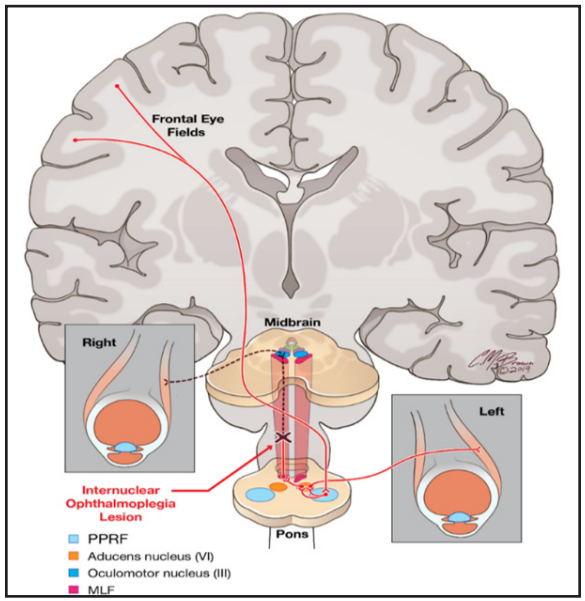
Treatment
- Observe, prism, strabismus surgery if >6 months duration and stable, botulinum toxin

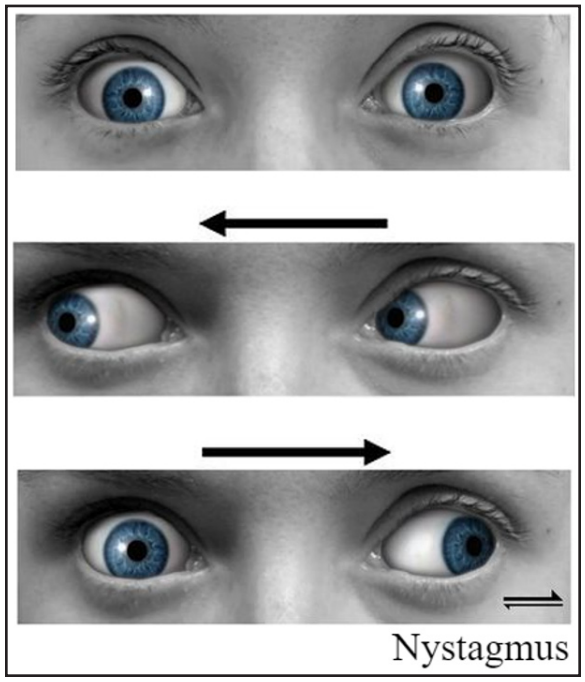
Ischemic Sixth Nerve Palsy Criteria

- Over age 40
- One or more vasculopathic risk factors
- No h/o cancer, vasculitis, or autoimmune disease
- Sudden onset of diplopia that does not vary
- No pain
- No other systemic neurologic symptoms
- Esotropia in primary position
- Unilateral restriction of abduction
- No other motility limitations on the affected side
- Other eye: normal motility
- Both eyes: normal VA, full CVF, normal pupils, no ptosis, no redness, no proptosis, normal intraocular exam (no iritis, vitritis, or optic nerve abnormalities)

Internuclear Ophthalmoplegia:

Secondary to lesion in the MLF. Damage to the MLF disrupts its ability to conduct signals sent from the PPRF.





Paramedian Pontine Reticular Formation (PPRF): Involved in the coordination of eye movements

- Horizontal gaze
- Saccades

Medial Longitudinal Fasciculus (MLF): A set of myelinated composite fiber tracts found in the brainstem

- Connects CN VI nucleus on one side of pons to the medial rectus subnucleus of CN III in contralateral midbrain

Signs & Symptoms

- Horizontal diplopia
- Exotropia
- Weakness or paralysis of adduction
- Horizontal jerk nystagmus of the abducting eye

Causes

- Brainstem (midbrain or pons) stroke, tumor, infection, inflammation, MS
- Wernicke encephalopathy (thiamine deficiency)
- Pernicious anemia causing B12 deficiency

Differential Diagnosis

- Myasthenia gravis, partial third nerve palsy

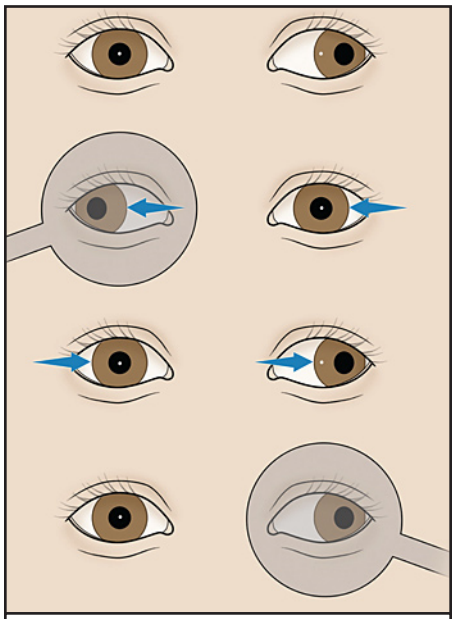
Workup

- Urgent MRI of brainstem and midbrain

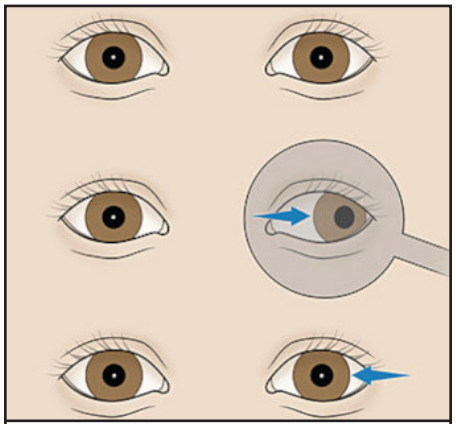
Treatment

- Treat underlying condition

Decompensated Phoria: A latent binocular misalignment that becomes symptomatic.



If "tropic" (the latent strabismus has become manifest): comitant esotropia or exotropia on cover test



If "phoric" (latent strabismus): no movement on cover test, but a latent esophoria or exophoria is demonstrated on alternate cover test

Signs & Symptoms
- Horizontal diplopia initially intermittent, may become persistent
  - Often worse with fatigue or alcohol
  - Onset with advancing age, illness, or trauma
- History of covering one eye

Causes
- Previous strabismus, now with loss of motor control and subsequent diplopia

Differential Diagnosis
- Any other cause of diplopia: diagnosis of exclusion
- Must be certain other neurologic disease absent

Workup
- Orthoptic measurements
- Rule out nerve palsy

Treatment
- Observe if occasional
- Prism, strabismus surgery, botulinum toxin

Decompensated Phoria Criteria

- h/o intermittent horizontal diplopia
- No severe headache, pulsatile tinnitus, or neurologic symptoms
- No arm or leg weakness, hoarse voice, problems swallowing or breathing, or history of intermittent ptosis
- No eye or orbital pain
- No symptoms of GCA
- No limitation of eye movement in any direction
- Normal velocity horizontal and vertical saccades
- Deviation measures the same on prism cover test in all directions of gaze
- Decreased horizontal prism fusional amplitude
- No increase in diplopia on sustained side or upgaze
- Normal visual fields to confrontation

Anisocoria: Unequal pupils.

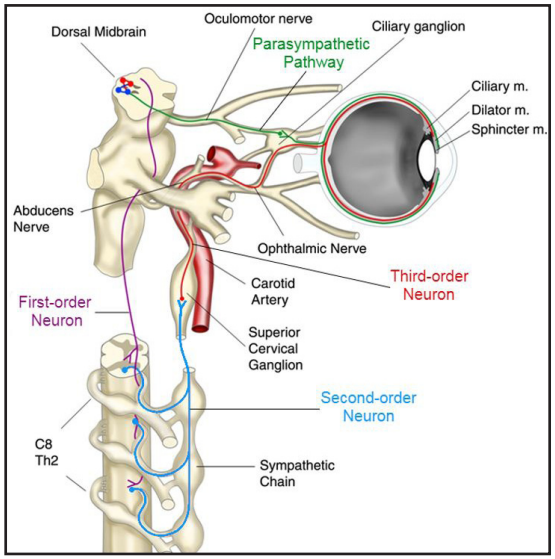
Adie (Tonic) Pupil: Results from damage of the post-ganglionic fibers of the parasympathetic pathway. Involved pupil is larger.

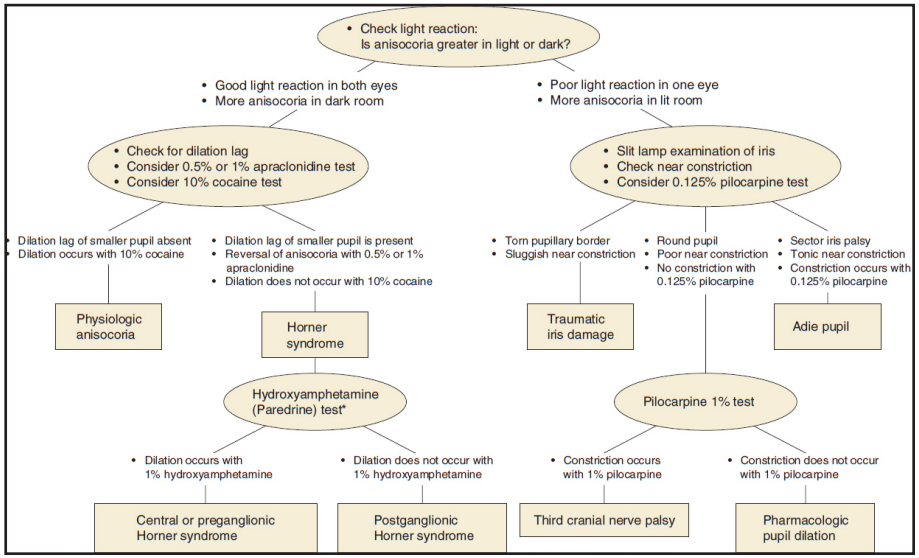
Signs & Symptoms
- Anisocoria greater in bright illumination
- Dilated pupil has minimal or no reaction to light, and slow, tonic constriction with convergence
- Spiraling of iris on slit lamp exam
- Absence of other neurologic symptoms/signs

Causes
- Idiopathic, orbital trauma, surgery, VZV infection

Differential Diagnosis
- Third nerve palsy, pharmacologic, traumatic iris damage

Workup
- Observe





Horner Syndrome: Results from damage to sympathetic pathway. Involved pupil is smaller.

Signs & Symptoms
- Anisocoria greater in dim illumination
- Mild upper lid ptosis and lower lid elevation on the affected side
- May have anhidrosis

Causes
- Central (first-order neuron): brainstem stroke, brainstem tumor, spinal cord tumor/compression
- Preganglionic (second-order neuron): apical lung tumor, internal carotid artery dissection, iatrogenic (neck/chest surgery)
- Postganglionic (third-order neuron): internal carotid artery dissection, cavernous sinus tumor or inflammation/infection, autonomic trigeminal cephalalgias, iatrogenic (neck surgery)

Differential Diagnosis
- Physiologic anisocoria

Workup
- Central or preganglionic → x-ray cervical spine, CT chest, MRI brain/brainstem, and MRI/MRA or CT/CTA neck
- Postganglionic → MRI brain/brainstem, and MRI/MRA or CT/CTA neck

Visual Pathway Disorder:

Damage to the visual pathway somewhere between the optic nerve and visual cortex, including optic chiasm, optic tract, and optic radiations.

Optic Chiasmal Disease

Signs & Symptoms

- Partial or complete bitemporal field loss

Causes

- Pituitary tumor
- Aneurysm
- Rare: infiltration, infection, ischemia, MS

Workup

- Urgent MRI of brain with contrast

Treatment

- Neurosurgery and endocrinology

Retrochiasmal Disease

Signs & Symptoms

- Partial or complete homonymous hemianopia

Causes

- Stroke
- Tumor
- Rare: inflammation, infection

Workup

- MRI of brain

Treatment

- Stroke evaluation
- Neurosurgery for tumors