



Neuro-ophthalmology

Last Updated: July 6, 2021

Table 1: Diplopia
Differential list for diplopia

Neoplastic	<ul style="list-style-type: none">● Intraorbital meningioma● Pituitary adenoma (GH secreting, or apoplexy)● Metastasis● Carcinomatosis
Vascular	<ul style="list-style-type: none">● Giant cell arteritis● Giant intracranial aneurysm● Microvascular disease
Infectious	<ul style="list-style-type: none">● Meningitis● Sphenoid sinusitis (CN VI palsy)● Botulism
Metabolic	<ul style="list-style-type: none">● Wernicke's encephalopathy
Inflammatory	<ul style="list-style-type: none">● Multiple sclerosis● Myasthenia gravis● GBS● Miller-Fisher syndrome● Orbital pseudotumor● Orbital myositis
Miscellaneous	<ul style="list-style-type: none">● Traumatic neuropathy (orbital apex fracture, blowout fracture of orbital wall)● Increased intracranial pressure (CN III and VI palsy)● INO (lesion of MLF)● Graves' disease (thyroid orbitopathy)● Corneal disease

- Cataracts
- Idiopathic intracranial hypertension (CN VI palsy)

Table 2: Ptosis

Differential list for true ptosis

Neoplastic	<ul style="list-style-type: none"> • Infantile rhabdomyosarcoma • Hemangioma • Optic glioma • Dermoid cyst • Metastatic neuroblastoma
Neurologic	<ul style="list-style-type: none"> • Ophthalmoplegia • Unilateral ptosis • Mydriasis • Cranial nerve nuclear lesions
Endocrine	<ul style="list-style-type: none"> • Thyroid orbitopathy
Congenital	<ul style="list-style-type: none"> • Congenital Horner's syndrome • Marcus Gunn phenomenon • Jaw Winking syndrome • Blepharophemosis syndrome • Neonatal myasthenia
Inflammatory	<ul style="list-style-type: none"> • Myasthenia gravis • Orbital pseudotumor • Tolosa-Hunt syndrome • Polymyositis
Miscellaneous	<ul style="list-style-type: none"> • Trauma (post-surgical) • Third Cranial Nerve Palsy • Chronic topical steroid drops

Table 3: Monocular Blindness

Differential list for acute or chronic monocular blindness

Neoplastic	<ul style="list-style-type: none"> • Intraorbital neoplasm • Tuberculum sella meningioma
Vascular	<ul style="list-style-type: none"> • Amaurosis fugax (carotid bifurcation, great vessel, or cardiogenic thromboembolism) • Carotid cavernous aneurysm rupture • AION • Central or branch retinal artery occlusion • Central retinal vein occlusion • Takayasu's arteritis • Temporal arteritis • Hypoperfusion (hypovolemia, hypotension, hyperviscosity)
Inflammatory	<ul style="list-style-type: none"> • Optic neuritis • Retrobulbar neuritis • Multiple sclerosis
Miscellaneous	<ul style="list-style-type: none"> • Trauma • Ocular migraine • Retinal detachment • Papilledema • Vitreous hemorrhage

Table 4: Binocular Blindness

Differential list for binocular blindness. The distinction of cortical blindness from a pathway lesion is the preservation of the pupillary light reflex in the setting of cortical blindness.

Neoplastic	<ul style="list-style-type: none"> • Multifocal malignant glioma • Occipital lobe metastases
Vascular	<ul style="list-style-type: none"> • Anoxia (cortical infarction, hemorrhage) • AION
Infectious	<ul style="list-style-type: none"> • Multifocal abscess

Metabolic	<ul style="list-style-type: none"> • Hypovitaminosis (B1, folic acid, B12)
Toxic	<ul style="list-style-type: none"> • Methanol (acute loss) • Ethanol (progressive)
Inflammatory	<ul style="list-style-type: none"> • Optic neuritis
Miscellaneous	<ul style="list-style-type: none"> • Traumatic neuropathy

Table 5: Transient Blindness

Differential list for causes of transient blindness. Cerebrovascular occlusion often involves either a branch or the central retinal artery.

Vascular	<ul style="list-style-type: none"> • Cerebrovascular embolism (carotid bifurcation, cardiogenic, calcific, atrial myxoma, air, adipose, amniotic fluid) • Hypoperfusion • Hemorrhage • Central retinal vein occlusion • AION • Takayasu's arteritis
Inflammatory	<ul style="list-style-type: none"> • Multiple sclerosis • Optic neuritis • Papilledema
Miscellaneous	<ul style="list-style-type: none"> • Typical migraine • Psychogenic

Table 6: Argyll Robertson Pupil

Differential list for the Argyll Robertson Pupil, which can be defined as bilateral miotic pupils that constrict to accommodation but are non-reactive to light.

Neoplastic	<ul style="list-style-type: none"> • Pinealoma

Infectious	<ul style="list-style-type: none"> • Meningovascular syphilis • Brainstem encephalitis
Endocrine	<ul style="list-style-type: none"> • Diabetes Mellitus
Toxic	<ul style="list-style-type: none"> • Alcoholism

Table 7: Marcus Gunn Pupil (Afferent defect)

Differential list for a Marcus Gunn pupil, which can be defined as a pupil which demonstrates greater pupillary sphincter contraction consensually than by direct exposure. The etiology of this finding is a prechiasmal disruption of the optic pathway.

Neoplastic	<ul style="list-style-type: none"> • Olfactory groove meningioma • Optic glioma
Vascular	<ul style="list-style-type: none"> • Hemorrhage (vitreous)
Inflammatory	<ul style="list-style-type: none"> • Multiple sclerosis • Optic neuritis
Miscellaneous	<ul style="list-style-type: none"> • Retinal detachment • Macular degeneration

Table 8: Adie Tonic Pupil

Differential list for Adie tonic pupil, which can be defined as a pupil that is tonically dilated and slowly reactive to light exposure. Tonic accommodation is commonly observed. Etiology of this condition relates to neurodegeneration within the ciliary ganglion.

Neoplastic	<ul style="list-style-type: none"> • Midbrain tegmental tumors
Vascular	<ul style="list-style-type: none"> • Ischemia • Amyloidosis

Infectious	<ul style="list-style-type: none"> • Meningovascular syphilis • Herpes zoster • Measles • Influenza • Cellulitis • Other viral agents
Toxic	<ul style="list-style-type: none"> • Alcohol
Congenital	<ul style="list-style-type: none"> • Familial dysautonomia • CMT disease
Inflammatory	<ul style="list-style-type: none"> • Sarcoidosis • Rheumatoid arthritis • GBS • Miller-Fisher syndrome • Sjogren syndrome • Shy-Drager syndrome • Orbital vasculitis
Miscellaneous	<ul style="list-style-type: none"> • Trauma • Ocular migraine • Post-orbital surgery • Paraneoplastic syndrome • Ross syndrome • Oculomotor nerve palsy

Table 9: Hippus

Differential list for Hippus, which can be defined as alternating miosis and mydriasis of the pupil under a consistent light source.

Vascular	<ul style="list-style-type: none"> • Infarction (contralateral)
Infectious	<ul style="list-style-type: none"> • Meningitis
Inflammatory	<ul style="list-style-type: none"> • Multiple sclerosis

Miscellaneous	<ul style="list-style-type: none"> • Idiopathic • Cataracts (during initial stages) • Oculomotor palsy recovery • Hysteria
---------------	--

Table 10: Miosis

Differential list for miosis. Those causes that induce a unilateral miosis are specifically noted.

Vascular	<ul style="list-style-type: none"> • Hemorrhage (pontine)
Infectious	<ul style="list-style-type: none"> • Neurosyphilis (rarely unilateral)
Toxic & Pharmacologic	<ul style="list-style-type: none"> • Cannabinoids • MAOI • Guanethidine • Opioids • Phentolamine • Cholinergics • Cholinesterase inhibitors • Barbiturates • Reserpine
Miscellaneous	<ul style="list-style-type: none"> • Horner's syndrome (unilateral) • Argyll Robertson pupil • Advanced age

Table 11: Mydriasis

Differential list for mydriasis. Those causes that induce a unilateral mydriasis are noted.

Neoplastic	<ul style="list-style-type: none"> • Supratentorial mass induced rostrocaudal deterioration • Tectal mass inducing Parinaud's syndrome

Vascular	<ul style="list-style-type: none"> • Aneurysm (PCA, P-comm, SCA)
Infectious	<ul style="list-style-type: none"> • Acute ciliary ganglionitis
Endocrine	<ul style="list-style-type: none"> • Thyrotoxicosis
Toxic & Pharmacologic	<ul style="list-style-type: none"> • Epinephrine • Phenylephrine • Cocaine • Amphetamines • LSD • TCA • Anticholinergics • Cyclopentolate • Antihistamines • Thiopental
Miscellaneous	<ul style="list-style-type: none"> • Cluster headaches (unilateral) • Uncal herniation (unilateral) • Adie pupil syndrome (unilateral) • Pseudodilation (from a contralateral constriction syndrome) • Epilepsy (post-ictal) • Hyperemotional state

Table 12: Internuclear Ophthalmoplegia

Differential list for internuclear ophthalmoplegia, which is a syndrome resulting from a lesion of the medial longitudinal fasciculus between the 3rd and 6th cranial nerves nuclei. This disorder manifests as diplopia on lateral gaze and disconjugate eye movements.

Neoplastic	<ul style="list-style-type: none"> • Intra-axial brainstem tumors • Extra-axial brainstem tumors • 4th ventricular tumors • Metastasis
------------	--

Vascular	<ul style="list-style-type: none"> • Brainstem infarction
Infectious	<ul style="list-style-type: none"> • Brainstem encephalitis • Tertiary syphilis
Metabolic	<ul style="list-style-type: none"> • Hepatic encephalopathy • Maple syrup urine disease
Toxic	<ul style="list-style-type: none"> • TCA • Barbiturates • Phenothiazines • Phenytoin • Wernicke's encephalopathy (pINO)
Congenital	<ul style="list-style-type: none"> • Chiari II (syringobulbia associated) • Chiari III (syringobulbia associated)
Inflammatory	<ul style="list-style-type: none"> • Multiple sclerosis • SLE • Myasthenia gravis (pINO) • GBS (pINO) • Miller-Fisher syndrome (pINO)
Miscellaneous	<ul style="list-style-type: none"> • Trauma • Exotropia (pINO) • Progressive supranuclear palsy

Table 13: Vertical Gaze Palsy

Differential list for vertical gaze palsy

Neoplastic	Pineal tumors 3 rd ventricular tumors Intra-axial midbrain tumors Infarction (dorsal midbrain) Hemorrhage (dorsal midbrain)
Vascular	Infarction (dorsal midbrain)

	Hemorrhage (dorsal midbrain)
Endocrine	Thyroid-associated orbitopathy (mimic)
Metabolic	Hypovitaminosis B1 or B12 Niemann-Pick disease, type C Gaucher's disease
Congenital	Congenital oculomotor apraxia Congenital upward gaze limitation (mimic)
Inflammatory	Multiple sclerosis GBS (mimic) Miller-Fisher syndrome
Miscellaneous	Trauma Hydrocephalus

Table 14: Acute Ophthalmoplegia

Differential list for acute ophthalmoplegia. Oculomotor nerve palsy is most commonly observed with the presence of an aneurysm at the interface between the P-Comm and ICA. Abducens nerve palsy is more commonly observed with aneurysms of AICA or the basilar artery.

Neoplastic	<ul style="list-style-type: none"> ● Pituitary adenoma ● Brainstem glioma ● Craniopharyngioma ● Pineal tumors ● Lymphoma ● Nasopharyngeal carcinoma ● Optic glioma ● Orbital dermoid cyst ● Orbital rhabdomyosarcoma ● Cavernous hemangioma of the orbit ● Metastatic neuroblastoma
Vascular	<ul style="list-style-type: none"> ● Aneurysm (AICA, basilar, P-Comm, ICA terminus) ● Hemorrhage (brainstem) ● Inferior petrosal sinus thrombosis

	<ul style="list-style-type: none"> • Cavernous sinus thrombosis • Carotid-cavernous sinus fistula
Infectious	<ul style="list-style-type: none"> • Encephalitis • Botulism • Diphtheria
Endocrine	<ul style="list-style-type: none"> • Thyrotoxicosis
Toxic	<ul style="list-style-type: none"> • TCA • Antiepileptic drugs
Inflammatory	<ul style="list-style-type: none"> • Myasthenia gravis • Orbital pseudotumor • Pseudotumor cerebri • Tolosa-Hunt syndrome • Sarcoidosis
Miscellaneous	<ul style="list-style-type: none"> • Trauma • Idiopathic cranial nerve palsy • Ophthalmoplegic migraine

Table 15: Papilledema

Differential for papilledema

Neoplastic	<ul style="list-style-type: none"> • Melanocytoma • Hemangioma • Hemangioblastoma • Metastatic (lung, breast, leukemia) • Meningeal carcinomatosis • Tumors that compress the intraorbital optic nerve
Vascular	<ul style="list-style-type: none"> • Hematoma (epidural, subdural, parenchymal) • AION
Infectious	<ul style="list-style-type: none"> • Cerebral abscess • Meningoencephalitis

	<ul style="list-style-type: none"> • Lyme disease • Neurosyphilis
Congenital	<ul style="list-style-type: none"> • Congenital disk elevation (pseudopapilledema)
Endocrine	<ul style="list-style-type: none"> • Juvenile diabetes (unilateral)
Inflammatory	<ul style="list-style-type: none"> • Sarcoidosis • Papillitis (unilateral) • Arteritis (SLE)
Miscellaneous	<ul style="list-style-type: none"> • Post-traumatic cerebral edema • Foster-Kennedy syndrome (unilateral) • Acute obstructive hydrocephalus

Table 16: Optic Nerve Enlargement
Differential for enlargement of the optic nerve

Neoplastic	<ul style="list-style-type: none"> • Optic nerve sheath meningioma • Intracranial meningioma (extension to optic nerve) • Optic nerve glioma • Plexiform neuroma • Metastasis (breast, prostate, lung, Ewing's sarcoma, leukemia, neuroblastoma)
Infectious	<ul style="list-style-type: none"> • Lyme disease • Syphilis • Tuberculosis • Whipple's disease • Toxoplasmosis • Histoplasmosis • Cryptococcus • Viral encephalitis • Poliomyelitis • Herpes zoster • Measles • Mumps

	<ul style="list-style-type: none"> • Rubella • Mononucleosis
Metabolic	<ul style="list-style-type: none"> • Adrenoleukodystrophy
Inflammatory	<ul style="list-style-type: none"> • Multiple sclerosis • Optic neuritis • Neuromyelitis optica • Sarcoidosis • Idiopathic inflammatory pseudotumor • Inflammatory bowel disease (CD, UC) • Reiter's syndrome

Table 17: Horner's Syndrome

Differential list for Horner's syndrome. The specific neuron involved in the pathway (first, second, or third order) is listed along with each etiology as 1, 2, or 3, respectively. The first-order neuron connects the hypothalamus with the intermediolateral cell column in the upper thoracic cord. The second-order neuron connects the intermediolateral cell column to the superior cervical ganglion. The third-order neuron completes the pathway from the superior cervical ganglion to the orbit.

Neoplastic	<ul style="list-style-type: none"> • Pontine glioma (1) • Upper cord glioma (1) • Upper cord ependymoma (1) • Upper cord neurofibroma (2) • Upper cord schwannoma (2) • Pancoast tumors of the lung (2) • Cervical paravertebral mass (2) • Axillary metastasis (2)
Vascular	<ul style="list-style-type: none"> • Infarction (lateral medullary, dorsolateral pontine, hemisphere) (1) • Cavernous sinus lesion (3) • Carotid artery dissection (3) • Carotid artery trauma (3)

Infectious	<ul style="list-style-type: none"> • Encephalitis • Bulbar poliomyelitis (1)
Congenital	<ul style="list-style-type: none"> • Cervical rib syndrome
Inflammatory	<ul style="list-style-type: none"> • Multiple sclerosis (1) • ALS (1)
Miscellaneous	<ul style="list-style-type: none"> • Cervical cord trauma (1) • Lower brachial plexus trauma (2) • Cervical disk herniation (2) • Cluster headache (3) • Syringobulbia (1) • Superior orbital fissure lesions (3) • Iatrogenic surgical injury during ACDF, pharyngeal, laryngeal, or thyroid surgery (2)

DOI: <https://doi.org/10.18791/nsatlas.v2.02.1.2>