



# Neuro- Ophthalmology





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## Basic Level Goals:

### A. Cognitive Skills

1. Describe the neuroanatomy of the visual pathways.\*\*
2. Describe the anatomy and functions of cranial nerves 2-8.\*\*
3. Describe the anatomy of the bony orbit.
4. Describe the pupillary and accommodative neuroanatomy.\*\*
5. Describe ocular motility and related neuronal pathways.\*\*
6. Describe the typical features, evaluation, and management of the most common optic neuropathies (eg, infectious, demyelinating, ischemic, inflammatory, hereditary, toxic, nutritional, compressive, infiltrative).\*\*
7. Describe the typical features, evaluation, and management of the most common ocular motor neuropathies (eg, third, fourth, sixth nerve palsy).\*\*
8. Describe the typical features of cavernous sinus syndrome and superior orbital fissure syndrome.
9. Describe and distinguish congenital nystagmus versus acquired nystagmus.
10. Describe the typical features, evaluation, and management of the most common efferent pupillary abnormalities (eg, Horner syndrome, third nerve palsy, tonic pupil, light-near dissociation).\*\*
11. Describe the typical features and evaluation of the most common visual field defects (eg, optic nerve, optic chiasm, optic radiation, occipital cortex).\*\*
12. Describe the clinical features and evaluation of ocular myasthenia gravis.
13. Describe the clinical features and evaluation of carotid-cavernous fistula.
14. Describe the differential diagnosis, evaluation, and management of congenital optic nerve abnormalities (eg, optic pit, disc coloboma, papillorenal syndrome, morning glory syndrome, tilted disc, optic nerve hypoplasia, myelinated nerve fiber layer, melanocytoma, disc drusen, Bergmeister papilla).
15. Describe the features of simple supranuclear and internuclear palsies (eg, internuclear ophthalmoplegia, vertical gaze palsy).
16. Describe the signs of nonorganic visual loss.
17. Describe the indications for obtaining neuroimaging studies, including computerized tomography (CT) scanning, magnetic resonance imaging (MRI), orbital

ultrasonography, and catheter angiography.

18. Describe the signs and symptoms of giant cell arteritis and the indications for performing a temporal artery biopsy.\*\*
19. Describe the clinical features, evaluation and neuro-ophthalmic aspects of thyroid ophthalmopathy.\*\*
20. Describe a systematic, sign-and-symptom-oriented neuro-ophthalmic patient interrogation (ie, history taking) and recording techniques.\*\*
21. Describe features of common headache and facial pain syndromes (eg, migraine, trigeminal neuralgia).

### B. Technical/Surgical Skills

1. Perform basic visual function tests (eg, color vision testing, Amsler grid, photostress test, contrast sensitivity testing).\*\*
2. Perform tests of binocularity and fusion (eg, polarized Titmus stereo test, Worth 4-dot test).\*\*
3. Perform a basic pupillary examination.\*\*
4. Describe indications for and perform basic pharmacologic pupillary testing for Horner syndrome, pharmacologic dilation, and tonic pupil.\*\*
5. Describe and detect a relative afferent pupillary defect.\*\*
6. Detect light-near dissociation
7. Perform a basic assessment of ocular alignment.\*\*
8. Use simple observational techniques (eg, Hirschberg test, Krimsky method).\*\*
9. Describe and perform basic cover/uncover testing for tropia.\*\*
10. Describe and perform alternate cover testing for phoria.\*\*
11. Perform simultaneous prism and cover testing.\*\*
12. Perform measurement of deviations with prisms.\*\*
13. Describe the indications for and apply Fresnel and grind-in prisms.
14. Describe the indications for and in a clinical setting perform forced duction and forced generation testing.
15. Perform a complete evaluation of the major ocular motor systems (eg, fixation, pursuit, saccades, convergence, vestibuloocular reflex).
16. Perform an evaluation of eyelids (eg, assess lid position, measure palpebral fissure, quantify levator function).\*\*

17. List the indications for visual field testing and interpret standard clinical perimetry programs.\*\*
18. Perform confrontational field testing (eg, static and kinetic, central and peripheral, red and white targets).\*\*
19. Describe the indications for and perform basic kinetic perimetry and interpret results.\*\*
20. Describe the indications for and perform basic automated perimetry and interpret results.
21. Describe the format of standard clinical tests (eg, light stimulus, background illumination, test points).\*\*
22. Perform basic direct, indirect, and magnified ophthalmoscopy examination of the optic disc, macula, vessels, and periphery of the retina (eg, recognize optic disc swelling, optic atrophy, neuroretinitis, nerve head vascular abnormalities, and macular abnormalities, such as edema, pigmentary changes, subretinal fluid, vessel abnormalities, pigmentary changes) and use the findings to generate a differential diagnosis.\*\*
23. Describe the anatomy and indications for CT, MRI, and angiography.\*\*
24. Describe the indications for and interpret basic echography (ultrasound) of the orbits.
25. Perform exophthalmometry.
26. Check pulse, blood pressure in both arms, carotid bruit, and heart sounds.

### Standard Level Goals:

Describe the neuro-ophthalmic anatomy and physiology (ie, the orbit and adnexal structures, the afferent and efferent visual pathways with their intracranial projections, the sensory and motor anatomy of the face, and the autonomic nervous system, including their blood supplies) as it applies to the eye and visual system.

#### A. Cognitive Skills

1. Describe typical and atypical features, evaluation, and management of the most common optic neuropathies (eg, papilledema, optic neuritis, ischemic, inflammatory, infectious, infiltrative, compressive, hereditary optic neuropathies).\*\*
2. Describe features, evaluation, and management of the more complex supranuclear and internuclear palsies (eg, progressive supranuclear palsy and subtle internuclear ophthalmoplegia, one-and-half syndrome).
3. List the common causes of an acute versus chronic isolated ocular motor neuropathy and define general management of each.\*\*
4. List the common causes of cavernous sinus syndrome

and superior orbital fissure syndrome.\*\*

5. Describe and differentiate among different forms of acquired nystagmus (eg, downbeat, upbeat, pendular, gaze evoked, rebound, convergence, retraction).\*\*
6. List the different mechanism causing nonphysiologic anisocoria and describe characteristics features and evaluation of the less common disorders (eg, mixed sympathetic and parasympathetic denervation of iris, aberrant regeneration in third nerve palsy, pharmacologic miosis).
7. List mechanism and causes of central versus peripheral light near dissociation (eg, Argyll-Robertson pupil, diabetic neuropathy, tonic pupil, Parinaud syndrome).
8. Describe features and evaluation of the less commonly encountered visual field defects (eg, sectoranopia, checkerboard, monocular temporal crescent).
9. Describe more advanced aspects of visual field testing indications, selection, and interpretation (eg, artifacts of automated perimetry, testing, and thresholding strategies).
10. Describe neuro-ophthalmic aspects of common systemic diseases (eg, hypertension, diabetes, thyroid disease, myasthenia gravis, temporal arteritis, sarcoidosis, systemic infections, inflammation).\*\*
11. Describe neuro-ophthalmic findings that are common following head trauma (eg, traumatic optic neuropathy, bilateral fourth nerve palsy, traumatic brain injury).\*\*
12. Describe evaluation and management of inherited neuro-ophthalmic diseases (eg, Leber hereditary optic neuropathy, autosomal dominant optic atrophy, spinocerebellar degenerations).\*\*
13. Describe evaluation and management of ocular myasthenia gravis.\*\*
14. Recognize common pathologic findings of brain and orbits on CT and MRI related to neuro-ophthalmology.\*\*
15. Describe the typical features, evaluation, and management of urgent neuro-ophthalmic pathologies (eg, giant cell arteritis, cavernous sinus thrombosis, orbital apex syndrome, pituitary apoplexy).\*\*

#### B. Technical Skills

1. Describe the indications for intravenous edrophonium (ie, Tensilon) and prostigmin tests for myasthenia gravis.\*\*
2. Perform a detailed cranial nerve evaluation other than the oculomotor nerve evaluation (eg, trigeminal, and facial and acoustic nerve function).

3. Describe the interpretation of neuro-radiologic images (eg, indications and interpretation of orbital tumors, thyroid eye disease, pituitary adenoma, optic nerve glioma, optic nerve sheath meningioma).
4. Describe the evaluation, management, and specific testing (eg, stereopsis, mirror test, red-green testing, monocular prism test) of patients with “functional” (ie, nonorganic) visual loss (eg, recognize nonorganic spiral or tunnel visual fields).\*\*
5. Describe the indications for, perform, and list the complications of temporal artery biopsy.
6. Perform and interpret basic ocular coherence tomography (OCT) imaging of the eye (eg, optic disc, retinal nerve fiber layer, macula).\*\*
7. Describe the indications and interpret basic ocular electrophysiology (eg, visually-evoked potential [VEP], electroretinogram [ERG], electrooculogram [EOG]).
8. Perform basic neurologic screening examination (eg, tandem walk, sensory examination, cerebellar function testing, basic cognitive evaluation).
9. Identify patients with “functional” visual loss (ie, nonorganic visual loss) and provide appropriate approach and follow up.\*\*
10. Quantify relative afferent pupillary defect (RAPD) with neutral density filter and be able to detect RAPD in patients with only one working pupil.\*\*
11. Interpret fluorescein angiography images.

### Advanced Level Goals:

#### A. Cognitive Skills

1. Describe the typical and atypical features, evaluation, and management of papilledema and raised intracranial pressure due to a variety of causes (eg, sinus thrombosis, idiopathic, meningitis).\*\*
2. Describe the typical features, evaluation, and management of urgent neuro-ophthalmic pathologies (eg, giant cell arteritis, cavernous sinus thrombosis, orbital apex syndrome, pituitary apoplexy).\*\*
3. Describe typical features of the most advanced and least common optic neuropathies (eg, chronic recurrent inflammatory optic neuritis, posterior ischemic optic neuropathy, neuromyelitis optica, autoimmune optic neuropathy, toxic/nutritional).\*\*
4. Describe typical and atypical features, evaluation, and management of the most complex and least common ocular motor neuropathies and their mimics (eg, patterns of aberrant regeneration).
5. Describe typical and atypical features, evaluation, and management of the most complex and least common forms of nystagmus (eg, spasmus nutans,

see-saw nystagmus, periodic alternating nystagmus).

6. Describe typical and atypical features, evaluation, and management of the most advanced and least common pupillary abnormalities (eg, pupil findings in coma, transient pupillary phenomenon).
7. Describe features, evaluation, and management of the most complex and least common visual field defects and recognize pattern mimics (eg, combination of disc-related scotoma plus hemianopia, binasal hemianopia, sectoranopia, bilateral inferior altitudinal loss due to superior occipital lobe lesions and not bilateral anterior ischemic optic neuropathy).\*\*
8. Describe, evaluate, and treat the neuro-ophthalmic aspects of systemic diseases (eg, malignant hypertension, diabetic papillopathy, toxicity of systemic medications, paraneoplastic syndromes, HIV/AIDS).\*\*
9. Describe, evaluate, and treat the neuro-ophthalmic manifestations of trauma (eg, corticosteroid or surgical therapy in traumatic optic neuropathy).
10. Describe, evaluate, and provide appropriate genetic counseling for inherited neuro-ophthalmic diseases (eg, hereditary optic neuropathies, chronic progressive external ophthalmoplegia, neurofibromatosis, ataxia syndromes).
11. Recognize, evaluate, and treat transient monocular visual loss.\*\*
12. Describe indications and interpret blood test results for various systemic disorders with neuro-ophthalmic manifestations (eg, thyroid disorders, pituitary disorders, myasthenia gravis).
13. Describe syndromes of cortical visual dysfunction.
14. Detect early neuro-ophthalmic signs and symptoms of drug toxicity for commonly used medications.
15. Describe the neuro-ophthalmic complications related to pregnancy.

#### B. Technical/Surgical Skills

1. Perform and interpret the results of the intravenous edrophonium (ie, Tensilon) and prostigmin tests for myasthenia gravis; recognize and treat the complications of the procedures.\*\*
2. Perform and interpret the complete cranial nerve evaluation in the context of neuro-ophthalmic localization and diseases.\*\*
3. Interpret neuro-radiologic images in neuro-ophthalmology (eg, interpretation of orbital imaging for orbital pseudotumor and tumors, thyroid eye disease, intracranial imaging modalities and strategies for tumors, aneurysms, infection, inflammation, ischemia), and appropriately discuss, in advance of



testing, the localizing clinoradiological features with the neuroradiologist in order to obtain the best study and interpretation of the results.\*\*

4. Identify patients with “functional” visual loss (ie, nonorganic visual loss) and provide appropriate counseling and follow-up.\*\*
5. Quantify RAPD with neutral density filter and detect small RAPD in patients with only one working pupil.\*\*
6. Perform optic nerve sheath decompression, if trained, for papilledema.\*\*
7. Perform neuro-ophthalmic evaluations for people with special needs (eg, comatose patients, children, children with developmental and visual maturation evaluations).
8. Describe indications, dose, and administration of Botox for neuro-ophthalmic disorders (eg, hemifacial spasm, blepharospasm, paralytic strabismus).

### Very Advanced Level Goals: Subspecialist

#### A. Cognitive Skills

1. Describe the arterial circulation in detail and know the general venous drainage along the entire anterior visual pathway (eg, optic disc, retrobulbar optic nerve, intracranial segment of optic nerve, chiasm, lateral geniculate body).
2. Describe evaluation, give differential diagnosis, and outline a management plan of the most advanced and least common optic neuropathies (eg, chronic recurrent inflammatory optic neuritis, posterior ischemic optic neuropathy, neuromyelitis optica, autoimmune optic neuropathy, rare toxic optic neuropathies).\*\*
3. Describe the cortical visual syndromes and know the localization of the causative lesion (eg, akinetopsia, prosopagnosia, simultagnosia).
4. Be able to discuss strengths and weaknesses of current treatment options (eg, steroids for acute nonarteritic anterior ischemic optic neuropathy, hyperbaric oxygen treatment, neuromyelitis optica antibodies in optic neuritis).\*\*
5. Describe typical and atypical features, evaluation, and management of rare eye movement disorders (eg, differential diagnosis of monocular oscillations, localization of lesion and purported mechanism of oculopalatal myoclonus).
6. Describe typical features, pathophysiology, evaluation, and management of rare pupillary syndromes (eg, tadpole pupil, paradoxical pupillary constriction).
7. Describe the advantages, disadvantages, indications, and pitfalls in special perimetric methods (eg, blue-yellow perimetry, automated kinetic perimetry, motion perimetry, microperimetry).

8. Describe and differentiate among various kinds of unusual positive visual phenomena and know their possible causes (eg, palinopsia, persistent photopsia).\*\*
9. Know the differential diagnosis and evaluation for acute or progressive homonymous hemianopsia in a patient with a normal MRI.\*\*
10. Describe the various prion diseases and their management.
11. Describe the various mitochondrial syndromes that have neuro-ophthalmic manifestations, and provide appropriate genetic counseling for inherited neuro-ophthalmic diseases (eg, Kearns-Sayre and related syndromes, mitochondrial encephalomyopathy, lactic acidosis, stroke-like episodes [MELAS], neuropathy, ataxia, and retinitis pigmentosa [NARP]).\*\*
12. Describe evaluation, give differential diagnosis, and outline a management plan for patients with headache and facial pain presenting as neuro-ophthalmic manifestations.\*\*
13. Describe the features, evaluation, and differential diagnosis of dizziness and vertigo from neuro-ophthalmic problems.\*\*

#### B. Technical/Surgical Skills

1. Recognize pitfalls in interpretations of unusual results of pharmacologic tests used for diagnosis of pupillary disorders.\*\*
2. Know techniques that reveal the most subtle manifestations of eye movement disorder (eg, slow medial rectus saccade as the only sign of internuclear ophthalmoplegia, fundus photos for excyclotorsion, head shaking test).\*\*
3. Perform and interpret the complete neurologic examination.
4. Be able to detect symptomatic lesions overlooked by the neuroradiologist (eg, small lesion in optic canal, carotid dissection).\*\*
5. Be able to perform specific maneuvers that definitively reveal nonorganic visual loss or overlay (eg, 4-diopter prism test, rocking mirror).\*\*
6. Perform and interpret spectral-domain OCT (eg, outer retinal disorders, detection of drusen).
7. Describe the indications and interpret laboratory results for seromarkers, antibodies, and antigen levels for various systemic diseases with neuro-ophthalmic manifestations (eg, paraneoplastic syndromes, autoimmune disease, inflammatory disorders).\*\*
8. Interpret indocyanine green angiography and autofluorescence imaging.