

North American Neuro-Ophthalmology Society Neuro-Ophthalmology Curriculum

Proposed by the Curriculum Committee of the North American Neuro-Ophthalmology Society (NANOS)

Chair:	Valérie Biousse, MD	(N and O)*
Vice-chair:	Nancy J. Newman, MD	(N)
Members:	Kimberly Peele Cockerham, MD (O)	
	Bradley J. Katz, MD	(O)
	David Kaufman, DO	(N)
	Andrew Lee, MD	(O)
	Howard D. Pomeranz, MD, PhD (O)	
	Roger E. Turbin, MD	(O)
	Greg P. Van Stavern, MD	(N)
	Nicholas J. Volpe, MD	(O)
	Floyd A. Warren, MD	(O)

*(N: board certified in neurology; O: board certified in ophthalmology)

Neuro-ophthalmologists care for patients with visual problems related to the nervous system. Neuro-ophthalmology requires specialized training and expertise in diseases involving the eye and the brain. It is a subspecialty of both neurology and ophthalmology, and neuro-ophthalmologists are usually board certified in neurology, ophthalmology, or both.

The scope of practice of neuro-ophthalmologists may vary tremendously depending on their background (board certified in neurology or ophthalmology), and whether they perform surgery or not (ophthalmology is a surgical subspecialty, while neurology is not). In addition, a number of ophthalmology-trained neuro-ophthalmologists have additional training in oculoplastics or pediatric ophthalmology, allowing them to perform highly specialized surgical procedures.

Neuro-ophthalmology is still a “young subspecialty” and its definition varies from one place to another. Indeed, the subject material is extremely broad and the clinical experiences are fragmented and heterogeneous. Only a few fellows are trained in neuro-ophthalmology each year, and their knowledge and experiences vary depending on where and with whom they trained. Not all residency programs have a detailed neuro-ophthalmology curriculum and very few medical schools include our subspecialty in their curriculum. A plan of action is being developed by The North American Neuro-Ophthalmology Society to improve the quality of our training program, have the fellowships better recognized by both the American Academies of Neurology (AAN) and Ophthalmology (AAO), “approved” by the Association of University Professors of Ophthalmology (AUPO), and increase the number of trainees. The first step of such a plan of action is the development of a neuro-ophthalmology curriculum.

It is important to note that “curriculum” is defined as an educational plan rather than an all-inclusive list. The Curriculum Committee of the North American Neuro-Ophthalmology Society has developed this core curriculum in neuro-ophthalmology. This curriculum defines the minimum standards for the clinical neuro-ophthalmologist; its content should evolve as progress is made in our specialty. It should not be limiting, as the extent of this curriculum will vary depending on the neuro-ophthalmologist’s primary specialty (neurology or ophthalmology).

The goal of this curriculum is to outline “what the neuro-ophthalmologist should know”. It is the basis for the curriculum that should be taught in neuro-ophthalmology fellowship and should help define a more basic curriculum for ophthalmology and neurology residencies. Specific learning objectives, excerpting parts of this curriculum, need to be defined for fellows, residents, and medical students.

We also have attempted to weigh each part of this curriculum in order to emphasize what is really relevant to our specialty and to assist others in creating more focused curricula such as for fellows, residents or CME program planning. We have “weighted” each topic as “most relevant”, “more relevant”, and “relevant”. Our definitions of “most relevant”, “more relevant”, and “relevant” appear on the next page.

The first part of the curriculum includes a synopsis of anatomy and physiology and examination

techniques relevant to neuro-ophthalmology, as well as a number of “neuro-ophthalmic manifestations” of various ophthalmic, neurologic, and systemic disorders. Indeed, most diseases have neuro-ophthalmic manifestations. Some are common or particularly important as they may lead to early diagnosis and management of specific disorders. These disorders are emphasized in this outline.

The second part of the curriculum includes a list of disorders that are primarily diagnosed and treated by a neuro-ophthalmologist”, including disorders of the afferent visual pathways and efferent neuro-ophthalmic structures such as eye movement disorders and pupillary and eyelid abnormalities.

Weighing system proposal:

The curriculum includes a list of topics relevant to our specialty. Each topic is followed by the relevant score indicated in superscript.

We are proposing that topics be prioritized to indicate the depth and breadth of knowledge in the area that is required of a specialist in neuro-ophthalmology. We have used previous experience from the Curriculum in Emergency Medicine developed by the Society for Academic Emergency Medicine (SAEM) and the Council of Emergency Medicine Residency Directors (CORD), and from the Neuro-Ophthalmology/Orbit Knowledge Base Panel from the American Academy of Ophthalmology to develop the following weighting system:

1) **Most relevant (mastery of the topic):**

Knowledge or skills that are essential to the independent management of illness and injury in patients with neuro-ophthalmic disorders. This level of knowledge or skill is required to manage clinical problems that a) pose significant risks to patients’ health or visual function, b) require prompt diagnosis or management to insure optimal outcome, and c) are typically diagnosed and/or managed by neuro-ophthalmologists (i.e. common diseases seen on a daily basis by neuro-ophthalmologists). Because this knowledge/skill leads to important decisions and interventions, it must be comprehensive in breadth and depth, and accessible to the neuro-ophthalmologist without the benefit of consultation.

2) **More relevant (proficiency with the topic):**

Knowledge or skills that are used by the neuro-ophthalmologist, but are not essential for the independent and timely diagnosis and/or management of common neuro-ophthalmologic disorders. This level of knowledge or skills may be possessed by the highly trained neuro-ophthalmologist, but lacking in-depth knowledge/skill, a neuro-ophthalmologist may review reference texts, consult other specialists, or refer to other physicians without posing a risk to patient’s health or visual function. Typically, clinical problems requiring this level of knowledge/skill are managed in other settings or by other specialists. The neuro-ophthalmologist may commonly provide initial evaluation of these problems, but generally not definitive management. Rare disorders classically evaluated by neuro-ophthalmologists are also included in this category.

3) **Relevant (familiarity with the topic):**

Knowledge pertaining to clinical conditions that are either benign or not directly related to neuro-ophthalmology, and do not pose an imminent threat to patients’ health or visual function. Immediate diagnosis and management of these conditions by a neuro-ophthalmologist are beyond the scope of standard practice. This level of knowledge facilitates comprehensive and thorough diagnosis and/or management of complex clinical problems encountered in neuro-ophthalmology, but referral to other specialists is generally required for the diagnosis and/or management of these conditions.

(Note: This version is the result of the application of the curriculum outline to the organization of the NOVEL collections, as well as the contributions and suggestions of Howard Pomeranz, MD. 09-05)

**North American Neuro-Ophthalmology Society
Neuro-Ophthalmology Curriculum
Draft – September 2005**

I. ANATOMY AND PHYSIOLOGY FOR THE NEURO-OPHTHALMOLOGIST

- A. Bony anatomy**^(1 and 2)
 - 1. Orbit⁽²⁾
 - 2. Bony communications
 - a. Superior orbital fissure⁽¹⁾
 - b. Optic canal⁽¹⁾
 - c. Inferior orbital fissure⁽²⁾
 - d. Ethmoidal foramina⁽²⁾
 - 3. Skull⁽²⁾
 - a. Anterior cranial fossa
 - b. Middle cranial fossa
 - c. Posterior cranial fossa
- B. Anatomy of the orbit, the eyelids, and the lacrimal pathways**⁽²⁾
- C. Neuroanatomy**
- D. Afferent visual pathways**⁽¹⁾
 - 1. Anatomy and physiology of the eye
 - 2. Retina
 - a. Macula**
 - 3. Optic nerve
 - 4. Optic chiasm
 - 5. Optic tract
 - 6. Lateral geniculate
 - 7. Optic radiations (**Meyer's loop**)
 - 8. Calcarine cortex
 - 9. Association areas
- E. Efferent visual pathways**⁽¹⁾
 - 1. Supranuclear input
 - 2. Cerebellar connections
 - 3. Nuclear centers
 - 4. Ocular motor nerves
 - a. Abducens (VI)
 - b. Trochlear (IV)
 - c. Oculomotor (III)
 - 5. Extraocular muscles
 - 6. Vestibular pathways
- F. Facial motor anatomy**⁽²⁾
- G. Sensory anatomy (trigeminal system)**⁽²⁾
- H. Autonomic anatomy**⁽¹⁾
 - 1. Sympathetic
 - 2. Parasympathetic
 - a. Lacrimal
 - b. Pupil
- I. Vascular anatomy**⁽²⁾
 - 1. Arterial anatomy
 - a. Internal carotid arteries and their branches
 - b. Circle of Willis⁽¹⁾
 - c. External carotid arteries and their branches

- d. Vertebrobasilar system
- e. Aortic arch
- f. Blood supply of the orbit, eye and optic nerve⁽¹⁾
- 2. Venous anatomy
 - a. Cerebral venous sinuses and deep venous system
 - b. Cortical veins
 - c. Venous drainage in the neck
 - d. Venous drainage of the eyes and orbits

J. Fluids and secretions (Maybe better to move to II.B.7)

1. Cerebrospinal fluid

II. OCULAR AND NEUROLOGIC EVALUATION

A. Ocular and neurologic examinations

- 1. Ocular evaluation
 - a. Evaluation of visual function (Snellen and Rosenbaum card visual acuity, stereopsis, color vision, contrast sensitivity, basics of refraction, confrontation visual fields, Amsler grid, photostress testing)⁽¹⁾
 - b. Ocular examination, intraocular pressure, resistance to retropulsion, [Hertel] exophthalmometry⁽¹⁾
 - c. Pupillary examination**
 - i. Measuring the RAPD**
 - d. Funduscopic examination⁽¹⁾
 - i. Normal optic disc**
 - a) Disc anatomy**
 - b) Vascular features**
 - i) Venous pulsations**
- 2. Use of lensometer, phoropter, slit lamp, **transillumination**, direct and indirect ophthalmoscopy and slit lamp biomicroscopy of the fundus⁽¹⁾
- 3. Ocular motility, use of prisms, cover testing, red glass, maddox rod, forced duction test, sensory testing⁽¹⁾
- 4. Ocular examination of the young child⁽¹⁾
- 5. Neurologic evaluation
 - a. Neurologic examination (adult and child)⁽¹⁾
 - b. Basic cognitive evaluation (mini mental status)⁽¹⁾
- 6. Neuro-ophthalmic evaluation of the comatose patient⁽¹⁾
- 7. Examination of children:
 - a. Developmental milestones for children⁽²⁾
 - c. Visual maturation of children⁽¹⁾

Deleted: es

8. The Neuro-ophthalmologic history (maybe this should be moved to the top as number

one?)

B. Ancillary tests obtained in neuro-ophthalmology

- 1. Visual field testing
 - a. Automated static perimetry (familiarity with current perimeters and different testing strategies)⁽¹⁾
 - b. Kinetic perimetry
 - _____ Goldmann perimetry (should be able to perform)⁽¹⁾
 - _____ Tangent screen (should be able to perform)⁽¹⁾

Deleted: 1

Deleted: c.

c. Measure the Relative Afferent Pupillary Defect (RAPD)

Deleted: d

2. Electrophysiology
 - a. Visual evoked responses/potentials⁽²⁾
 - b. Electroretinogram⁽³⁾
 - i. focal
 - ii. full field
 - c. Multifocal electroretinogram⁽³⁾
 - d. Dark adaptation⁽³⁾
 - e. Eye movement recordings⁽²⁾
3. Ocular and orbital ultrasound [A scan, B scan]⁽²⁾
4. Retinal fluorescein angiography⁽²⁾
5. Nerve fiber layer analysis
 - a. Optical coherence tomography (OCT), HRT, GDx⁽³⁾
6. Imaging
 - a. Computed tomography⁽¹⁾
 - b. Magnetic resonance imaging⁽¹⁾
 - c. Vascular imaging (ultrasonography, CTA, MRA, CTV, MRV, conventional angiogram)⁽¹⁾
 - d. Functional neuro-imaging (MRI, SPECT, PET)⁽³⁾
7. Lumbar puncture with opening pressure⁽¹⁾

III. NEURO-OPHTHALMIC SYMPTOMS AND SIGNS**A. Visual Loss**⁽¹⁾

1. Transient/Amaurosis⁽¹⁾
2. Permanent⁽¹⁾
3. Unexplained visual loss⁽¹⁾
4. Non organic visual loss⁽¹⁾

B. Positive Visual Phenomena [palinopsia, polyopia, macropsia, micropsia]⁽¹⁾**C. Visual hallucinations [formed, unformed]**⁽¹⁾**D. Visual Field Loss**⁽¹⁾**E. Higher cortical dysfunction**⁽¹⁾**F. Normal and abnormal optic nerve**⁽¹⁾

1. Edema (disc swelling, papilledema, pseudotumor cerebri) and pseudopapilledema/anomalous nerves
2. Optic atrophic
3. Optociliary shunt vessels
 - a. **Shunt Vessels (Glaucoma)**
 - b. **Shunt Vessels (CRVO)**
 - c. **Shunt Vessels (Congenital)**
 - d. **Shunt Vessels (Meningioma)**
4. Cupped optic nerve
5. Optic disc anomalies/pseudopapilledema

G. Oscillopsia, nystagmus, ocular oscillations⁽¹⁾**H. Double vision (diplopia)**⁽¹⁾**I. Abnormal extraocular movements**⁽¹⁾**J. Ptosis [reverse ptosis, brow ptosis]**⁽¹⁾**K. Lid findings**

1. Lid retraction
2. Lid lag
3. Lag ophthalmos⁽¹⁾
4. Lid twitch [fasciculations]

L. Orbital signs

- 1. **Proptosis**
- 2. **Enophthalmos⁽¹⁾**
- 3. **Pulsating exophthalmus**
- M. Pupillary changes**
 - 1. **Anisocoria⁽¹⁾**
 - 2. **Dilation Lag**
 - 3. **RAPD [Marcus Gunn pupil]**
- N. Facial Abnormalities**
 - 1. **Facial weakness⁽¹⁾**
 - 2. **Hemifacial spasm**
 - 3. **Titubation**
- P. Ocular pain, facial pain and headaches⁽¹⁾**
- Q. Speech disturbances [dysarthria]**

IV. DISORDERS OF THE AFFERENT AND EFFERENT VISUAL PATHWAYS

A. Diseases of the retina

- 1. **Cone**
- 2. **Rod**
- 3. **Ganglion cell**
- 4. **Macula**

B. Diseases of the optic nerve

- 1. Ischemic optic neuropathy [arteritic, non-arteritic]⁽¹⁾
 - a. Anterior
 - b. Posterior
 - c. Diabetic papillopathy
 - d. **Optic Neuropathy with Retinopathy [idiopathic enlarged blind spot syndrome]**

2. Inflammation⁽¹⁾ Formatted: French France

- a. Non infectious
 - 1. Idiopathic optic neuritis
 - 2. Optic neuritis and multiple sclerosis
 - 3. **Periphlebitis in Optic Neuritis**
 - 4. Other inflammatory optic neuritides
 - i. **Sarcoidosis, orbital inflammation, lupus, etc.**

Formatted: French France

- b. Infectious
 - 1. Optic perineuritis
 - 2. **Neuroretinitis [Bartonella, cat scratch disease]**
 - 3. **Optic Neuropathy with Sinus Disease**

Formatted: English U.S.

- 3. Compression / Infiltration⁽¹⁾
- 4. Paraneoplastic [CRMP5]⁽¹⁾
- 5. Traumatic⁽¹⁾

a. Traumatic Optic Neuropathy

- 6. Toxic⁽¹⁾
 - a. **BCNU Retinopathy [should this go under IV.A.4?]**

- 7. Nutritional⁽¹⁾
- 8. Metabolic⁽¹⁾
- 9. Hereditary⁽¹⁾

- a. **Leber's Optic Neuropathy**
- b. **Dominant/Kjer's optic atrophy**

Deleted: . . . a.

- 10. Congenital [anomalous]⁽¹⁾
- 11. Glaucoma⁽²⁾
 - a. Classification of glaucoma

Formatted: Bullets and Numbering

- b. Evaluation and basic management of glaucoma
- 12. Raised intracranial pressure
 - a. **Papilledema,**
 - b. **Pseudotumor cerebri/ Idiopathic intracranial hypertension**
- 13. Decreased intraocular pressure (hypotony)⁽²⁾

Deleted: Idiopathic intracranial hypertension

Formatted: Bullets and Numbering

C. Orbital pathology causing neuro-ophthalmic manifestations

Formatted: English U.S.

- 1. Trauma⁽²⁾
- 2. Mass lesions⁽²⁾
 - a. Neoplasms
- 3. Inflammation/Infection⁽²⁾
 - a. Orbital inflammation
 - i. Myositis**
 - b. Thyroid orbitopathy [Graves’]
 - c. Orbital cellulitis
 - d. Abscess
- 4. Orbital manifestations of dural fistulas⁽¹⁾

D. Diseases of the chiasm

- 1. Chiasmal visual field defects⁽¹⁾
- 2. Compression/infiltration⁽¹⁾
 - a. Chiasmal herniation**
 - b. Arachnoid cyst**
- 3. Inflammation⁽¹⁾
 - a. Chiasmal Optic Neuritis**
- 4. Trauma⁽¹⁾
- 5. Ischemia/hemorrhage⁽¹⁾

Formatted: English U.S.

Formatted: English U.S.

E. Diseases of the retrochiasmal visual pathways

- 1. Optic tract⁽¹⁾
 - a. Optic Tract Lesions**
- 2. Lateral geniculate⁽¹⁾
- 3. Radiations⁽¹⁾
- 4. Calcarine cortex⁽¹⁾
- 5. Association areas⁽²⁾
- 6. Specialized syndromes⁽²⁾
 - a. Anton's syndrome (cerebral blindness)
 - b. Riddoch's phenomena: Statico-kinetic dissociation
 - c. Balint syndrome
 - d. Gerstmann syndrome
 - e. Cerebral achromatopsia
 - f. Alexia without agraphia
 - g. Acalculia
 - h. Agraphia (with associated alexia)
 - i. Agnosias
 - j. Visual neglect
 - k. L-R confusion
 - l. Akinetopsia
 - m. Concept of "Blindsight"
 - n. cortical blindness**

Formatted: English U.S.

F. Pupillary pathology

- 1. Normal pupillary responses⁽¹⁾
- 2. Effects of drugs on the pupils⁽¹⁾
- 3. Congenital pupillary abnormalities⁽²⁾
- 4. Pupillary changes secondary to ocular diseases⁽¹⁾
 - a. Traumatic, foreign body
 - b. Inflammation
 - c. Neovascularisation
 - 1. Ocular surgery, laser
- 5. Traumatic pupillary changes⁽¹⁾
- 6. Evaluation and management of anisocoria⁽¹⁾
- 7. Evaluation and management of a large or a small pupil⁽¹⁾
- 8. Evaluation and management of specific pupillary disorders⁽¹⁾:
 - a. Adie's tonic pupil **Sector pupil palsy (LND & Sector palsies)**
 - b. Tadpole pupil⁽²⁾
 - c. Argyll-Robertson pupil
 - d. Correctopia⁽²⁾
 - e. Physiologic anisocoria (**Dilation lag**)
 - f. Horner's syndrome (**Dilation lag**)
 - g. Third nerve palsy

1. Cyclical [unless these conditions demonstrate pupil abnormalities it appears they belong only below in G.8.a.]

2. Subnuclear

- h. Afferent pupillary defect (**RAPD, RAPD not present, RAPD present**)
- i. Light near dissociation

j. Flynn Phenomenon, paradoxical pupillary light reflex (not in UMLS)

G. Eye movement systems pathology

- 1. Vestibular ocular system⁽¹⁾
- 2. Optokinetic nystagmus⁽¹⁾
- 3. Saccades⁽¹⁾
- 4. Pursuit⁽¹⁾
- 5. Convergence⁽¹⁾
- 6. Divergence⁽¹⁾
- 7. Titubation**
- 8. Specific ocular motor syndromes⁽¹⁾:
 - a. Cranial nerve palsies
 - 1. Third (Oculomotor)**
 - i. Cyclical**
 - ii. Subnuclear**
 - iii. Ophthalmoplegic Migraine**
 - 2. Fourth (Trochlear)**
 - 3. Sixth (Abducens)**
 - b. **Nuclear/Supranuclear** palsies
 - 1. Internuclear ophthalmoplegia
 - 2. One and half syndrome
 - 3. Horizontal gaze palsy
 - 4. Monocular elevation deficit
 - 5. Vertical gaze palsy
 - 6. Skew deviation
 - 7. Ocular tilt reaction

Formatted: English U.S.

Formatted: French France

Field Code Changed

Field Code Changed

Field Code Changed

Formatted: French France

- 8. Upgaze palsy
- 9. Downgaze palsy
- 8. Lateropulsion

- c. Ocular motor apraxia
- d. Spasm of the near triad
- e. Convergence insufficiency
- f. Divergence insufficiency
- g. Decompensation of phorias
- h. Restriction syndromes
- i. Ocular neuromyotonia

j. mitochondrial myopathy (chronic progressive external ophthalmoplegia/Kearns-Sayer)

k. Cyclic oculomotor paresis⁽²⁾

Deleted: i.

Formatted: Bullets and Numbering

Deleted: j

9. Classical brain stem syndromes⁽²⁾

- a. Foville
- b. Millard-Gubler
- d. Duane's syndrome

1. Synergistic Divergence

- d. Möbius syndrome
- e. Locked In syndrome
- f. Nothnagel
- g. Benedickt
- h. Weber
- i. Claude syndrome
- j. Wallenberg syndrome
- k. Syndrome of the anterior inferior cerebellar artery

10. Ocular motility disturbance by location⁽¹⁾

- a. Medulla
- b. Pons
- c. Mesencephalon
- d. Dorsal midbrain syndrome [Parinaud's]
- e. Cerebellar pathology

Formatted: French France

Formatted: English U.S.

H. Nystagmus and disorders of ocular stability

- 1. Jerk nystagmus⁽¹⁾
- 2. Pendular nystagmus⁽¹⁾ (**binocular & monocular**)
- 3. Congenital vs acquired nystagmus⁽¹⁾
- 4. Central vs peripheral nystagmus⁽¹⁾
 - a. **Vestibular nystagmus**
 - i. **Left beat nystagmus**⁽¹⁾
- 5. Specific types of nystagmus and their localizing value^(1 if common and 2 if rare)
 - a. Downbeat nystagmus⁽¹⁾
 - b. Upbeat nystagmus⁽¹⁾
 - c. **Gaze evoked nystagmus**
 - d. Rebound nystagmus⁽¹⁾
 - e. Brun's nystagmus⁽²⁾
 - f. Periodic alternating⁽¹⁾
 - g. Convergence retraction nystagmus⁽¹⁾
 - h. See saw nystagmus⁽¹⁾
 - i. Divergence nystagmus⁽²⁾
 - j. Sensory nystagmus⁽¹⁾
 - k. Congenital motor nystagmus⁽¹⁾
 - l. Spasmus nutans/dissociated nystagmus/monocular nystagmus⁽²⁾

- m. Rotary nystagmus
- n. Abducting nystagmus
- o. Latent nystagmus
- p. Vestibular nystagmus
- q. End-gaze nystagmus
- r. Voluntary nystagmus

Formatted: German Germany

- 6. Induced nystagmus⁽³⁾
 - a. Valsalva
 - b. Sounds (Tullio's phenomena)
 - c. Calorics: hot or cold water in ear⁽¹⁾
- 7. Ocular oscillations⁽¹⁾
 - a. Superior oblique myokymia
 - b. Square wave jerks
 - c. Opsoclonus
 - d. Flutter
 - e. Ocular bobbing
 - f. Oculopalatal myoclonus
 - g. Oculomasticatory myorhythmia⁽²⁾

I. Eyelid position abnormalities

- 1. Eyelid retraction.⁽¹⁾
- 2. Ptosis.⁽¹⁾
 - a. Pseudoptosis⁽³⁾
 - b. Congenital⁽²⁾
 - c. With elevator palsy⁽²⁾
 - d. Marcus Gunn Jaw Wink⁽²⁾
 - e. Blepharophimosis⁽³⁾
 - f. Levator dehiscence⁽²⁾
 - g. Myopathic⁽¹⁾
 - h. Neuro-muscular transmission⁽¹⁾
 - 1. Ocular myasthenia**
 - i. Neuropathic⁽¹⁾
 - 1. Apraxia of eyelid opening
 - 2. Third nerve dysfunction
 - 3. Aberrant regeneration/**reinnervation** of third nerve
 - 4. Horner's syndrome
 - j. Blepharospasm⁽¹⁾
- 3. Eyelid nystagmus⁽³⁾
- 4. Lid twitch**
- 5. Lid bobbing**

Formatted: English U.S.

Formatted: English U.S.

J. Facial nerve dysfunction

- 1. Central and peripheral facial palsy⁽¹⁾
- 2. Blepharospasm⁽¹⁾
- 3. Hemifacial spasm⁽¹⁾
- 4. Facial myokymia⁽²⁾
- 5. Oculomasticatory myorhythmia (Whipple's)⁽²⁾
- 6. Facial tics⁽³⁾
- 7. Facial myotonia⁽³⁾
- 8. Facial dystonia (Meige's syndrome)**

V. SYSTEMIC, NEUROLOGIC, AND OPHTHALMIC DISORDERS COMMONLY ASSOCIATED WITH NEURO-OPHTHALMIC MANIFESTATIONS

A. Developmental and congenital anomalies with neuro-ophthalmologic consequences

- 1. Visual maturation⁽¹⁾
- 2. Complications of prematurity⁽²⁾
- 3. Cerebral palsy⁽³⁾
- 4. Complications of birth injuries⁽³⁾
- 5. Congenital hydrocephalus⁽¹⁾
- 6. Cranial dysostoses (craniosynostosis)⁽³⁾
- 7. Amblyopia⁽²⁾
- 8. Other disc anomalies
 - a. Congenital optic nerve anomalies⁽²⁾
 - 1. Bergmeister papilla
 - 2. Optic nerve dysplasia and aplasia/hypoplasia
 - 3. Septo-optic dysplasia
 - 4. Optic nerve coloboma
 - 5. Optic nerve pit
 - 6. Morning glory syndrome
 - 7. Optic nerve drusen
 - 8. Tilted disc
 - 9. Myelinated nerve fibers
 - 10. Staphyloma
 - 11. Congenital Optic Atrophy**
 - 12. Laurence-Moon-Biedl Syndrome**
 - b. **Optic disc anomalies/pseudopapilledema**
- 9. Common malformations of the eye and orbit⁽³⁾
- 10. Skull base malformations.
 - a. Chiari malformation⁽¹⁾
 - b. **Basal encephaloceles**
- 11. Fibrous dysplasia⁽³⁾
- 12. Ocular manifestations of child abuse⁽²⁾

Formatted: English U.S.

Formatted: English U.S.

Formatted: English U.S.

B. Systemic disorders commonly associated with neuro-ophthalmologic manifestations.

- 1. Specific hereditary ocular and neurologic diseases with neuro-ophthalmic presentation⁽²⁾
 - a. **Pseudoxanthoma Elasticum**
 - b. **Bassen-Kornzweig Syndrome**
 - c. **Ehlers-Danlos Syndrome**
 - d. **Olivopontocerebellar Degeneration (Spinal cerebellar ataxias)**
- 2. Neurocutaneous syndromes⁽²⁾
 - a. Neurofibromatosis (**Transillumination – ciliary vessels and lisch nodules**)
 - 1. NF-1 [von Recklinghausen]
 - 2. NF-2
 - b. Tuberous sclerosis [Bourneville's disease]
 - c. von Hippel-Lindau [angiomatosis of the retina and cerebellum]
 - d. Sturge-Weber [encephalotrigeminal angiomatosis]
 - e. Ataxia telangiectasia [Louis-Barr syndrome]
 - f. Wyburn-Mason [racemose angioma of the midbrain and retina]
- 3. Vascular disease

Formatted: English U.S.

Formatted: English U.S.

Field Code Changed

Formatted: English U.S.

Field Code Changed

Field Code Changed

Formatted: English U.S.

Field Code Changed

Formatted: English U.S.

Field Code Changed

Formatted: English U.S.

- a. Vascular risk factors⁽²⁾
 - 1. Factor V Deficiency**
- b. Prevention of vascular disease⁽³⁾
- c. Neuro-ophthalmic manifestations of vascular diseases⁽¹⁾
 - 1. Temporal /giant cell arteritis**
 - 2. Carotid Occlusive Disease**
 - 3. Vasospastic Amaurosis Fugax**
 - 4. Central/Branch Retinal Artery Occlusion**
 - 5. Saturday Night Retinopathy**
 - 6. Carotid Dissection**
 - 7. Moyamoya Disease**
 - 8. Dolichoectasia**
- d. Hypercoagulable states⁽²⁾
 - 1. Factor V Deficiency**
- 4. Systemic hypertension⁽²⁾
- 5. Metabolic diseases including diabetes mellitus⁽²⁾
 - 1. Diabetic Retinopathy**
- 6. Autoimmune diseases, classification of vasculitides⁽³⁾
 - a. Specific vasculitides with ocular and neurologic manifestations
 - 1. Giant cell arteritis⁽¹⁾ (**Temporal arteritis**)
 - 2. Sarcoidosis⁽¹⁾
 - 3. Lupus⁽²⁾
 - 4. Antiphospholipid Antibody Syndrome**
 - 5. Wegener granulomatosis⁽²⁾
- 7. Complications of cancers, paraneoplastic syndromes; principles of treatment⁽²⁾
 - a. Leukemia**
 - b. Lymphoma**
 - c. Histiocytosis**
 - d. Metastatic Carcinoma**
 - e. Multiple Myeloma**
 - f. Paraneoplastic Disease**
 - g. Lymphomatoid Granulomatosis**
- 8. Neuro-ophthalmic complications of chemotherapy, and radiation therapies⁽¹⁾
 - a. BCNU Retinopathy**
 - b. Radiation retinopathy**
- 9. Complications of infections⁽²⁾
 - a. Specific infections with common neuro-ophthalmic complications
 - a. AIDS
 - b. Syphilis
 - c. Presumed Ocular Histoplasmosis**
 - d. Toxoplasmosis**
- 10. Inflammatory**
 - a. Keratitis**
 - 1. Exposure Keratitis**

Deleted: arthritis

C. Neurologic disorders commonly associated with neuro-ophthalmologic manifestations.

- 1. Head and ocular injury
 - a. Recognize and evaluate neurological complications of head injury⁽³⁾
 - b. Neuro-ophthalmic complications of brain injury (acute and late)⁽¹⁾
 - c. Traumatic optic neuropathies (direct and indirect)⁽¹⁾
 - d. Traumatic cranial nerve palsies (III, IV, and VIth)⁽¹⁾
 - e. Diagnose and evaluate orbital and facial fractures⁽³⁾

- f. Recognized the complications of ocular trauma⁽³⁾
- g. Evaluate post-traumatic visual loss⁽¹⁾
- h. Diagnosis of post-concussion syndrome⁽³⁾
- 2. Increased intracranial pressure
 - a. Differential diagnosis and management of intracranial hypertension⁽¹⁾
 - 1. Intracranial mass, infection or bleed
 - 2. Meningitis
 - 3. Subarachnoid hemorrhage
 - 4. Hydrocephalus
 - 5. Cerebral venous thrombosis
 - 6. Idiopathic intracranial hypertension.
 - b. Neuro-ophthalmic manifestations and complications⁽¹⁾
- 3. Vascular disease of the brain and the eye
 - a. Classification of stroke⁽²⁾
 - 1. Thalamic infarct**
 - 2. Pontine infarct**
 - 3. Midbrain infarct**
 - b. Mechanisms⁽²⁾
 - 1. Vein vs artery
 - 2. Hemorrhage vs ischemia
 - 3. Embolism vs thrombosis vs hemodynamic
 - 4. Large artery vs small artery
 - c. Diagnosis and evaluation of stroke⁽²⁾
 - 1. Vascular evaluation of stroke**
 - 2. Cardiac evaluation of stroke
 - 3. Hypercoagulable states
 - d. Basics of acute treatment and secondary prevention of stroke⁽²⁾
 - e. Neuro-ophthalmologic manifestations of stroke⁽¹⁾
 - 1. Pituitary apoplexy**
 - f. Ocular ischemia⁽¹⁾
 - 1. Transient visual loss
 - 2. Central and branch retinal artery occlusions
 - 3. Ocular ischemic syndrome
 - 4. Central and branch vein occlusions
 - g. Venous sinus thrombosis⁽¹⁾
 - 1. Superior ophthalmic vein thrombosis**
 - 2. Cerebellar venous thrombosis**
 - h. Hemorrhage⁽¹⁾**
 - 1. Subarachnoid**
 - 2. Thalamic hemorrhage**
 - 3. Pontine hemorrhage**
 - 4. Midbrain hemorrhage**
 - i. Intracranial vascular malformations, diagnosis and neuro-ophthalmologic manifestations⁽¹⁾
 - 1. Aneurysms
 - i. Ophthalmic Artery Aneurysm**
 - ii. posterior communicating artery aneurysm**
 - 2. Arteriovenous malformations
 - 3. Dural fistulas
 - 4. Carotid cavernous fistulas (direct, indirect)
 - 5. Cavernous hemangiomas
- 4. Seizures with neuro-ophthalmologic manifestations
 - a. Occipital seizures⁽¹⁾

Deleted: . i.

Formatted: Bullets and Numbering

- b. Pupillary changes, ocular movement changes during seizures⁽²⁾
- 5. Neuro-ophthalmic manifestations of neoplasms:
 - a. Intracranial tumors⁽²⁾
 - b. Skull base tumors⁽²⁾
 - 1. Chordoma**
 - c. Orbital tumors⁽²⁾
 - 1. Choroidal Folds From Orbital Mass**
 - d. Optic nerve tumors⁽¹⁾
 - e. Benign vs malignant neoplasm⁽²⁾
 - f. Primary vs secondary⁽²⁾
 - g. Pediatric vs adult tumor^{(2)s}
- 6. Demyelinating disease
 - a. Multiple sclerosis⁽¹⁾
 - 1. Relationships between optic neuritis and multiple sclerosis⁽¹⁾
 - 2. Treatment strategies⁽²⁾
 - 3. Vision assessment in multiple sclerosis⁽¹⁾
 - b. Devic's disease⁽¹⁾
 - c. Acute disseminated encephalomyelitis⁽¹⁾
- 7. Infections (neuro-ophthalmic manifestations of localized and systemic infections)
 - a. Intracranial infections⁽²⁾
 - 1. Abscess
 - 2. Cerebritis
 - 3. Empyema
 - 4. Meningitis
 - 5. Meningo-encephalitis
 - b. Whipple's disease⁽²⁾
 - c. Orbital infections (cellulitis)⁽²⁾
 - d. Optic nerve infection (infectious optic neuritis and **neuroretinitis**)⁽¹⁾
 - e. Creutzfeld-Jacob disease⁽²⁾
 - f. Toxoplasmosis**
 - g. Lyme disease**
- 8. Metabolic diseases⁽³⁾
 - a. Wilson's disease
 - b. Vitamin deficiencies (vitamin A, B1, B12)
 - c. Metabolic storage diseases
 - d. Amyloidosis
- 9. Neuro-degenerative diseases⁽²⁾
 - a. Parkinson syndromes
 - 1. Parkinson's disease
 - 2. Progressive supranuclear palsy
 - b. Alzheimer, Frontotemporal dementia
 - c. Vascular dementias⁽³⁾
 - d. Amyotrophic lateral sclerosis⁽³⁾
 - e. Ataxia**
 - 1. Hereditary ataxias⁽³⁾ (**Spinal cerebellar ataxias**)
 - f. Tauopathy**
- 10. Polyradiculopathies
 - a. Guillain-Barré⁽²⁾
 - b. Miller Fisher variant⁽¹⁾
- 11. Neuro-muscular transmission deficits⁽¹⁾
 - a. Physiology of the neuromuscular transmission⁽¹⁾
 - b. Myasthenia gravis⁽¹⁾
 - c. Lambert-Eaton myasthenic syndrome⁽³⁾

- d. Toxic neuromuscular transmission defect (Botulism, medications)⁽²⁾
- 12. Myopathies (involving the extraocular muscles)
 - a. Congenital myopathies/ oculopharyngeal muscular dystrophy
 - 1. dystrophies/ Ion Channel Disorders (Myotonia)⁽³⁾
 - b. Mitochondrial diseases⁽²⁾
 - 1. Chronic progressive external ophthalmoplegia (CPEO)
 - c. Ischemic⁽¹⁾
 - 1. Giant cell arteritis (**Temporal arteritis**)
 - 2. Orbital ischemic syndrome
 - d. Metabolic - Toxic⁽³⁾
 - 1. Drug induced, toxic
 - e. Inflammatory⁽¹⁾
 - 1. Thyroid orbitopathy
 - 2. Orbital inflammatory disease
 - f. Neoplasm/infiltration⁽²⁾
 - g. Congenital syndromes involving the extraocular muscles
 - 1. Anomalous muscle insertions⁽³⁾
 - 2. Brown's syndrome⁽¹⁾
 - 3. Congenital fibrosis of the extraocular muscles (CFEOM) ⁽²⁾
 - 4. Duane's syndrome⁽¹⁾
 - i. Synergistic Divergence**
 - 5. High myopia⁽²⁾
- 13. Headache and facial pain
 - a. Classification of headaches and facial pain proposed by the International Headache Society (IHS)⁽²⁾
 - b. Migraine⁽¹⁾
 - 1. Migraine without aura⁽²⁾
 - 2. Migraine with visual aura⁽¹⁾
 - 3. Hemianopia after migraine**
 - c. Tension headaches⁽³⁾
 - 1. Episodic
 - 2. Chronic
 - d. Cluster headache⁽¹⁾
 - e. Headache associated with increased intracranial pressure⁽¹⁾
 - f. Headache and facial pain of vascular origin⁽²⁾
 - g. Ocular pain related to ocular or optic nerve disease⁽¹⁾
 - h. Trigeminal neuralgia (Fifth nerve)⁽¹⁾
 - i. Herpes zoster (zoster ophthalmicus)⁽¹⁾
 - j. Referred ocular pain⁽¹⁾
 - 1. Vascular (i.e. dissection)
 - 2. Ocular ischemic syndrome
 - 3. Cavernous sinus syndrome

Formatted: German Germany

14. Psychiatric illness

D. Ocular diseases commonly associated with- or mimicking neuro-ophthalmologic disorders.

- 1. Ocular neoplasms⁽³⁾
 - a. Benign vs malignant (**Transillumination – ocular melanoma**)
 - b. Primary vs secondary
 - c. Pediatric vs adult neoplasms
- 2. Ocular infections⁽³⁾
 - a. External
 - b. Endophthalmitis

c. Neuroretinitis⁽¹⁾

3. Ocular inflammation (uveitis)⁽³⁾
 - a. Classification of uveitis
 - b. Neuro-ophthalmic disorders associated with uveitis⁽¹⁾
 - c. Optic neuritis and uveitis⁽¹⁾
 - d. Meningo-uveitis⁽²⁾
4. Retinal disorders⁽³⁾
 - a. Vascular⁽¹⁾
 - b. Degenerative/hereditary⁽³⁾
 - 1. Acquired Hyperopia**
 - c. Inflammatory/infectious⁽³⁾
 - d. Outer retinopathies/White dot syndromes⁽³⁾
5. Glaucoma⁽³⁾
6. Ocular causes of acute and chronic visual loss⁽²⁾

Formatted: French France

Formatted: French France

E. Neuro-ophthalmic manifestations of iatrogenic diseases

1. Radiation⁽²⁾
2. Chemotherapy⁽³⁾
3. Various drugs with specific neuro-ophthalmologic complications, including
 - a. Cyclosporine⁽¹⁾
 - b. FK-506 (tacrolimus)⁽¹⁾
 - c. Amiodarone⁽¹⁾
 - d. Hydroxychloroquine⁽²⁾ (**Plaquenil**)
 - e. Ethambutol⁽¹⁾
 - f. Vasoconstrictors⁽¹⁾
 - g. Steroids⁽¹⁾
 - h. Facial and orbital injections⁽²⁾
4. Alcohol
 - a. Thiamine deficiency⁽¹⁾
 - b. Wernicke encephalopathy⁽²⁾
5. Neuro-ophthalmic complications of surgical procedures
 - a. Post operative visual loss⁽¹⁾
 - b. Epidural anesthesia⁽²⁾
 - c. Ocular and orbital surgery⁽¹⁾
 - d. Neurosurgery⁽¹⁾
 - e. Endovascular procedures⁽¹⁾

Formatted: French France

F. Functional disorders

1. Terminology⁽³⁾
 - a. Factitious (Münchhausen's)
 - b. Malingering
 - c. Conversion reaction
 - d. Exaggeration
 1. Hypochondriasis
 2. Somatization disorder
2. Clinical presentations⁽¹⁾
 - a. Visual loss⁽¹⁾
 - b. Visual field defects⁽¹⁾
 - c. Spasm of near triad⁽¹⁾

- d. **Voluntary** Nystagmus⁽¹⁾
- 3. Specific techniques of evaluation⁽¹⁾

VI. PROCEDURES COMMONLY PERFORMED/OBTAINED IN NEURO-OPHTHALMOLOGY

A. Surgical and endovascular procedures and their complications

1. Temporal artery biopsy⁽¹⁾
2. Principles and complications of strabismus surgery⁽²⁾
3. Canthotomy, cantholysis⁽²⁾
4. Approaches for orbital biopsies and orbital tumors⁽²⁾
5. Optic nerve sheath fenestration⁽¹⁾
6. Orbital decompression⁽¹⁾
7. CSF shunting procedures⁽¹⁾
8. Monitoring of intracranial pressure⁽¹⁾
9. Pituitary surgery (transphenoidal adenomectomy)⁽²⁾
10. Interventional neuroradiology techniques⁽²⁾
 - a. Cerebral angiography and venography⁽²⁾
 - b. Embolization⁽²⁾
 - c. Angioplasty⁽²⁾
 - d. Stenting⁽²⁾
 - e. Intravenous and intra-arterial thrombolysis⁽²⁾

11. Tensilon test

12. Cataract surgery

a. Intraocular lenses

B. Botulism toxin therapy

1. Treatment of blepharospasm, hemifacial spasm, Meige syndrome⁽²⁾
2. Treatment of strabismus⁽³⁾

VII. PATIENTS' COUNSELING

A. Definition of legal blindness⁽¹⁾

B. Legal requirements for driving (visual) in the state in which you practice neuro-ophthalmology⁽¹⁾

C. Counseling of the visually impaired patient⁽¹⁾

1. Liaison with rehabilitation services (neurologic and for the visually impaired)
2. Liaison with a low vision service

D. Genetic counseling⁽²⁾

1. Principles of genetics, genetic testing, and genetic counseling

VIII. RESEARCH, ADMINISTRATION, EDUCATION RESOURCES

A. Administration

1. Credentialing, career development, recruitment, budgeting, health care financing, managed care, public relations, personnel management, marketing, hospital administration, practice management, contracts, work schedule.⁽³⁾
2. JCAHO requirements relating to neuro-ophthalmology (staffing, equipment and supplies, facilities, quality insurance).⁽²⁾

B. Research

1. Read and analyze scientific articles⁽¹⁾
2. Research funding.⁽²⁾
3. Development of a research project.⁽²⁾
4. Ethical issues in research, including consent and researchers' interactions with corporate funding sources (conflict of interest)⁽²⁾
5. Write and publish a manuscript⁽³⁾

C. Large previous and ongoing studies addressing specific neuro-ophthalmic issues

1. Optic Neuritis Treatment Trial (ONTT)⁽¹⁾
2. Longitudinal Optic Neuritis Study (LONS)⁽¹⁾
3. CHAMPS study⁽¹⁾
 3. Ischemic Optic Neuropathy Decompression Trial (IONDT)⁽¹⁾

D. Ethics and professionalism

- A. Proper documentation in medico-legal proceedings⁽²⁾
- B. Knowledge of cost, resource allocation, quality of care and access to care issues.⁽²⁾
- C. Basic familiarity with medical malpractice.⁽²⁾
- D. Physician-physician relationships.⁽²⁾
- E. Laws relating to drug dispensing, regulation, and abuse.⁽¹⁾
- F. Role of expert witness in medico-legal procedures.⁽³⁾

E. Neuro-ophthalmologic resources

Topics:

A. Organizations⁽²⁾

- NANOS⁽¹⁾
- INOS
- EUNOS
- AAN
- ANA
- AAO
- ARVO
- Regional organizations
- NEI
- NINDS

B. Journals⁽²⁾

C. NOVEL database⁽³⁾

http://medstat.med.utah.edu/neuroophth.NOVEL_updates/

Formatted: English U.S.

Formatted: French France

IX. REFERENCES

References:

- American Academy of Neurology. Neuro-Ophthalmology/Neuro-otology Fellowship core curriculum. <http://www.aan.com/about/sections/fellowship/nono.pdf>
- American Academy of Ophthalmology. Basic and Clinical Science Course. Neuro-Ophthalmology. Section 5. 2003.
- American Academy of Ophthalmology. Basic and Clinical Science Course. Orbit, Eyelids, and Lacrimal

System. Section 7. 2003.

-Continuum. American Academy of Neurology. Newman NJ, Galetta SL, eds. Neuro-Ophthalmology. 2003.

-Glaser JS. Neuro-Ophthalmology. Lippincott Williams & Wilkins, 3rd edition. 1999.

-Headache Classification Subcommittee of the International Headache Society. The International Classification of Headache Disorders: 2nd edition. Cephalalgia. 2004; 24 Suppl 1: 9-160.

-Liesegang TJ, Hoskins HD Jr, Albert DM, O'Day DM, Spivey BE, Sadun AA, Parke DW, Modino BJ. Am J Ophthalmol 2003; 136: 114-121.

-Liu GT, Volpe NJ, Galetta SL. Neuro-Ophthalmology. Diagnosis and Management. WB Saunders, 2001.

-Miller NR, Newman NJ, Bioussé V, Kerrison JB. Walsh & Hoyt's Clinical Neuro-Ophthalmology. Lippincott Williams & Wilkins, 6th edition. 2005.

-Spivey BE. A technique to determine curriculum content. J Med Educ 1971; 46:269-274.

-Society for Academic Emergency Medicine (SAEM) webpage. How to design a curriculum ?

<http://www.saem.org/model/intro.htm>

X. HISTORY OF NEURO-OPHTHALMOLOGY