North American Neuro-Ophthalmology Society
Neuro-Ophthalmology Curriculum
Draft – September 2004

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. Afferent visual pathways[1]
1. Anatomy and physiology of the eye
2. Retina
3. Optic nerve
4. Optic chiasm
5. Optic tract
6. Lateral geniculate
7. Optic radiations
8. Calcarine cortex
9. Association areas

D. 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

E. Facial motor anatomy[2]

F. Sensory anatomy (trigeminal system) [2]

G. Autonomic anatomy[1]
1. Sympathetic
2. Parasympathetic
    a. Lacrimal
    b. Pupil

H. 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

II. OCULAR AND NEUROLOGIC EVALUATION

A. Ocular and neurologic examinations

1. Ocular evaluation
    a. Evaluation of visual function (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, exophthalmometry**(1)**
    c. Funduscopic examination**(1)**
2. Use of lensometer, phoropter, slit lamp, direct and indirect ophthalmoscopes 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)**
    b. Visual maturation of children**(1)**

B. Ancillary tests obtained in neuro-ophthalmology

1. Visual field testing
    a. Automated perimetry (familiarity with current perimeters and different testing strategies)**(1)**
    b. Goldmann perimetry (should be able to perform)**(1)**
    c. Tangent screen (should be able to perform)**(1)**
2. Electrophysiology
    a. Visual evoked responses**(2)**
    b. Electroretinogram**(3)**
    c. Multifocal electroretinogram**(3))**
    d. Dark adaptation**(3)**
    e. Eye movement recordings**(2)**
3. Ocular and orbital ultrasound(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[1]
2. Permanent[1]
3. Unexplained visual loss[1]
4. Non organic visual loss[1]

B. Positive Visual Phenomena[1]
C. Visual hallucinations[1]
D. Visual Field Loss[1]
E. Higher cortical dysfunction[1]
F. Normal and abnormal optic nerve[1]
1. Edema (disc swelling, papilledema) and pseudopapilledema/anomalous nerves
2. Optic atrophic
3. Optociliary shunt vessels
4. Cupped optic nerve
5. Optic disc anomalies/pseudopapilledema

G. Oscillopsia, nystagmus, ocular oscillations[1]
H. Double vision[1]
I. Abnormal extraocular movements[1]
J. Ptosis[1]
K. Lid retraction, lagophthalmos, lid lag[1]
L. Proptosis, enophthalmos[1]
M. Pupillary changes, anisocoria[1]
N. Abnormal facial movements[1]
O. Facial weakness[1]
P. Ocular pain, facial pain and headaches[1]

IV. DISORDERS OF THE AFFERENT AND EFFERENT VISUAL PATHWAYS

A. Diseases of the optic nerve

1. Ischemic optic neuropathy[1]
    a. Anterior
    b. Posterior
    c. Diabetic papillopathy
2. Inflammation[1]
    a. Non infectious
        1. Idiopathic optic neuritis
        2. Optic neuritis and multiple sclerosis
        3. Other inflammatory optic neuritides
            i. Sarcoidosis, orbital inflammation, lupus, etc...
    b. Infectious
        1. Optic perineuritis
        2. Neuroretinitis
3. Compression / Infiltration[1]
4. Paraneoplastic[1]
5. Traumatic[1]
6. Toxic[1]
7. Nutritional[1]

8. Metabolic[(1)]
9. Hereditary[(1)]
10. Congenital[(1)]
11. Glaucoma[(2)]
    a. Classification of glaucoma
    b. Evaluation and basic management of glaucoma
12. Raised intracranial pressure (papilledema)[(1)]
13. Decreased intraocular pressure (hypotony)[(2)]

B. Orbital pathology causing neuro-ophthalmic manifestations

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

C. Diseases of the chiasm

1. Chiasmal visual field defects[(1)]
2. Compression/infiltration[(1)]
3. Inflammation[(1)]
4. Trauma[(1)]
5. Ischemia/hemorrhage[(1)]

D. Diseases of the retrochiasmal visual pathways

1. Optic tract[(1)]
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"

E. 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
    b. Tadpole pupil[(2)]
    c. Argyll-Robertson pupil
    d. Correctopia[(2)]
    e. Physiologic anisocoria
    f. Horner's syndrome
    g. Third nerve palsy
    h. Afferent pupillary defect
    i. Light near dissociation

F. 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. Specific ocular motor syndromes[(1)]:
    a. Cranial nerve palsies
        1. Third, fourth and sixth
    b. 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
    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. Cyclic oculomotor paresis[(2)]
8. Classical brain stem syndromes[(2)]
    a. Foville
    b. Millard-Gubler
    c. Duane's syndrome
    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

9.Ocular motility disturbance by location[(1)]
a. Medulla
b. Pons
c. Mesencephalon
d. Dorsal midbrain syndrome
e. Cerebellar pathology

G. Nystagmus and disorders of ocular stability

1. Jerk nystagmus[(1)]
2. Pendular nystagmus[(1)]
3. Congenital vs acquired nystagmus[(1)]
4. Central vs peripheral nystagmus[(1)]
5. Specific types of nystagmus and their localizing value[(1 if common and 2 if rare)]
   a. Down beat nystagmus[(1)]
   b. Upbeat nystagmus[(1)]
   c. Rebound nystagmus[(1)]
   d. Brun's nystagmus[(2)]
   e. Periodic alternating[(1)]
   f. Convergence retraction nystagmus[(1)]
   g. See saw nystagmus[(1)]
   h. Divergence nystagmus[(2)]
   i. Sensory nystagmus[(1)]
   j. Congenital motor nystagmus[(1)]
   h. Spasmus nutans/dissociated nystagmus/monocular nystagmus[(2)]
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)]

H. 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]
i. Neuropathic[1]
1. Apraxia of eyelid opening
2. Third nerve dysfunction
3. Aberrant regeneration of third nerve
4. Horner's syndrome
j. Blepharospasm[1]

3. Eyelid nystagmus[3]

I. 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]

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. Congenital optic nerve anomalies[2]
a. Bergmeister papilla
b. Optic nerve dysplasia and aplasia/hypoplasia
1. Septo-optic dysplasia
c. Optic nerve coloboma
d. Optic nerve pit
c. Morning glory syndrome
d. Optic nerve drusen
e. Tilted disc
f. Myelinated nerve fibers
g. Staphyloma
9. Common malformations of the eye and orbit[3]
10. Skull base malformations. Chiari malformation[1]
11. Fibrous dysplasia[3]
12. Ocular manifestations of child abuse[2]

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

1. Specific hereditary ocular and neurologic diseases with neuro-ophthalmic presentation[2]
2. Neurocutaneous syndromes[2]
    a. Neurofibromatosis
        1. NF-1
        2. NF-2
    b. Tuberous sclerosis
    c. von Hippel-Lindau
    d. Sturge-Weber
    e. Ataxia telangiectasia
    f. Wyburn-Mason
3. Vascular disease
    a. Vascular risk factors[2]
    b. Prevention of vascular disease[3]
    c. Neuro-ophthalmic manifestations of vascular diseases[1]
    d. Hypercoagulable states[2]
4. Systemic hypertension[2]
5. Metabolic diseases including diabetes mellitus[2]
6. Autoimmune diseases, classification of vasculitides[3]
    a. Specific vasculitides with ocular and neurologic manifestations
        1. Giant cell arteritis[1]
        2. Sarcoidosis[1]
        3. Lupus[2]
        4. Wegener granulomatosis[2]
7. Complications of cancers, paraneoplastic syndromes; principles of treatment[2]
    a. Neuro-ophthalmic complications of chemotherapy, and radiation therapies[1]
8. Complications of infections[2]
    a. Specific infections with common neuro-ophthalmic complications (AIDS or syphilis for example)

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]
    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
2. Cardiac evaluation
3. Hypercoagulable states

d. Basics of acute treatment and secondary prevention of stroke[2]
e. Neuro-ophthalmologic manifestations of stroke[1]
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]
h. Subarachnoid hemorrhage, neuro-ophthalmic manifestations[1]
i. Intracranial vascular malformations, diagnosis and neuro-ophthalmologic manifestations[1]
1. Aneurysms
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]
b. Pupillary changes, ocular movement changes during seizures[2]

5. Neuro-ophthalmic manifestations of neoplasms:
a. Intracranial tumors[2]
b. Skull base tumors[2]
c. Orbital tumors[2]
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]

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. Hereditary ataxias[3]
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
        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]
        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]
    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 of optic nerve disease[(1)]
h. Trigeminal neuralgia[(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

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

1. Ocular neoplasms[(3)]
a. Benign vs malignant
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)]
c. Inflammatory/infectious[(3)]
d. Outer retinopathies/White dot syndromes[(3)]
5. Glaucoma[(3)]
6. Ocular causes of acute and chronic visual loss[(2)]

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 (tacrolismus) [(1)]
c. Amiodarone[(1)]
d. Hydroxychloroquine[(2)]
e. Ethambutol[(1)]
f. Vasoconstritors[(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]

F. Functional disorders

1. Terminology[3]
    a. Factitious (Münchausen'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. 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]

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)]
4. Ischemic Optic Neuropathy Decompression Trial (IONDT)[(1)]