North Carolina Academy of Physician Assistants

Recertification Exam Review

Tuesday, February 21\textsuperscript{st}, 2017

10:15am-12:15pm

NEUROLOGY REVIEW

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Disclosures

• none
Learning Objectives

At the end of this session participants will be able to:

• Describe the epidemiology data, pathophysiology, and risk factors associated with select neurological disorders

• Discuss typical and atypical clinical features of select neurological disorders

• Explain an approach to assessing patients with neurological complaints, including pertinent history, physical exam findings, and diagnostic studies

• Describe principles of managing select neurological disorders, including both pharmacological and behavioral interventions
### The Neurologic System

**Diseases of Peripheral Nerves**
- Complex regional pain syndrome
- Peripheral neuropathies

**Headaches**
- Cluster headache
- Migraine
- Tension headache

**Infectious Disorders**
- Encephalitis
- Meningitis

**Movement Disorders**
- Essential tremor
- Huntington disease
- Parkinson disease
- **Vascular Disorders**
  - Cerebral aneurysm
  - Intracranial hemorrhage
  - Stroke
  - Transient ischemic attack

**Other Neurologic Disorders**
- Altered level of consciousness
- Cerebral palsy
- Concussion
- Dementias
- Delirium
- Guillain-Barré syndrome
- Multiple sclerosis
- Myasthenia gravis
- Post-concussion syndrome
- Seizure disorders
- Status epilepticus
- Syncope
- Tourette disorder
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Total: 100%
10:15am-11:00am

• Anatomy & Physiology of the Neurological System
• The Neurologic Evaluation
  – History
  – Physical Examination
  – Diagnostic Tests
• Vascular Disorders
  – Transient ischemic attack
  – Ischemic Stroke
  – Intracerebral Hemorrhage
  – Subarachnoid Hemorrhage
• Multiple Sclerosis
• Gullain-Barre Syndrome
• Myasthenia Gravis
• Cerebral Palsy
• Seizure Disorders
• Questions
• BREAK

11:15am-12:15pm

• Disorders of Peripheral Nerves
  – Peripheral Mono & Poly-Neuropathies
  – Complex regional pain syndrome
• Dementias & Delirium
• Syncope
• Concussion & Post-Concussion Syndrome
• Altered Level of Consciousness
• Movement Disorders
  – Essential tremor
  – Huntington disease
  – Parkinson disease
• Tourette Syndrome
• Infectious Disorders
  – Encephalitis
  – Meningitis
• Headaches
  – Cluster headache
  – Migraine
  – Tension headache
• Questions
Case Scenario

Visual Aids

Summary Slide

General Information – Pathophysiology – Clinical Features
Evaluation – Treatment – Prognosis

Updates from the Literature
Where is the dysfunction in the nervous system?
Brief Review of Neuro-Anatomy and Physiology
The Neurologic Evaluation:
History

• **Symptoms**
  - Acute, Subacute, and/or Chronic
  - Involvement of motor, including autonomic, and sensory systems

• **Clinical Course**

• **Aggravating & Alleviating** Factors

• **Medical History**
  - Past Illnesses and Surgeries; Family History; Social History

• **Medications & Allergies**

• Thorough **ROS**, including **psychiatric** symptoms

• Baseline Level of **Function** & Impact on Function
The Neurologic Evaluation:
Physical Examination

1. **Mental Status Examination**
   - Level of Consciousness, Memory, Orientation, Perceptions, Thought Processes, Thought Content, Insight, Judgment, Affect, Mood, Language, Higher Cognitive Functions

2. **Cranial Nerves**
   - I-XII (next slide)

3. **Motor**
   - Body Position, Involuntary Movements, Muscle Bulk, Tone, Strength

4. **Sensory**
   - Pain & Temperature, Position & Vibration, Light Touch, Discriminative

5. **Reflexes**
   - Deep Tendon Reflexes (DTRs), Cutaneous Stimulation [Abdominal, Plantar]

6. **Coordination**
   - Rapid Alternating Movements, Point-to-Point Movements, Stance, Gait
The Neurologic Evaluation: Cranial Nerves

I: **Olfactory**: Olfaction
II: **Optic**: Visual Acuity & Fields, Pupillary Light Reflex
III: **Oculomotor**: Pupillary Constriction, Extraocular Movements (EOMs)
IV: **Trochlear**: Infero-Lateral EOM
V: **Trigeminal**: Mastication, Facial Sensation, Corneal Reflex
VI: **Abducens**: Lateral EOM

- VII: **Facial**: Facial Movements, Corneal Reflex
- VIII: **Vestibulocochlear**: Hearing & Balance
- IX: **Glossopharyngeal**: Palate Movement & Speech
- X: **Vagus**: Parasympathetic
- XI: **Accessory**: Shoulder Movement
- XII: **Hypoglossal**: Tongue Movement
Neurologic Exam: Video Resources

• Screening Neurologic Exam
  https://www.youtube.com/watch?v=bMuOqlmf8bo
  https://www.youtube.com/watch?v=PpP16YS02p4
## The Neurologic Evaluation: Diagnostic Tests

### Central Nervous System
- Computed Tomography (CT)
- CT Angiogram (CTA)
- Magnetic Resonance Imaging (MRI)
- MR Angiogram (MRA)
- Cerebral Arteriogram
- Electroencephalogram (EEG)

### Peripheral Nervous System
- Electromyogram (EMG)
- Nerve Conduction Study (NCS)
- Ultrasound (U/S)

### PNS & CNS
- Cerebrospinal Fluid (CSF)
- Evoked Potential Studies (EPS)
- Laboratory Tests
- Biopsy
Approach to the Patient with Neurologic Complaints

Where is the dysfunction in the nervous system?
Case

- **Hx:** A 65yo male presents to the emergency department with complaints of slurred speech and right-sided weakness that have been improving since onset of symptoms approximately 30 minutes ago.
- **PE:** Mild dysarthria, slight right facial droop, strength right UE 5-/5 and LE 5/5.
- **Dx:** CT Head non-contrast: no acute process

Then...

- **Repeat PE:** normal exam
Cerebrovascular Disease

**Transient Ischemic Attack (TIA)**

**General Info**
- Definition: Transient episode of neurologic dysfunction caused by focal ischemia without acute infarction (AHA/ASA 2009)

**Pathophysiology**
- Large artery low-flow, Embolic, or Lacunar
- Neurologic Emergency

**Clinical Features**
- Focal neurological impairment with anterior or posterior circulation features; Temporary
- DDx: Ischemic Stroke, Seizure, Migraine Aura, Syncope, Hypoglycemia
- ABCD² may identify high risk patients, need for hospitalization

**Evaluation**
- Complete within 24 hours
- Labs: CBC, Lipid Profile, BMP with Glucose, PT/PTT, ESR
- CT; MRI w/ diffusion; Angiogram; Transcranial Doppler Ultrasound
- Carotid Duplex Ultrasound
- EKG +/- Telemetry or Holter
- Echocardiogram (TTE/TEE)

**Treatment**
- Antiplatelet; Anticoagulation if cardiac thrombus, Afib
- Carotid Endarterectomy (70-99% Stenosis)
- Risk Factor Reduction
- Stroke education

**Prognosis**
- 4-10% have ischemic stroke within 48 hours
• **Hx:** A 65yo male presents to the emergency department with complaints of slurred speech and right-sided weakness that started abruptly 30 minutes ago – these symptoms have been worsening since the onset.

• **PE:** Mild dysarthria, mild right facial droop, strength right UE 4/5 and LE 4/5, unsteady gait.
Cerebrovascular Disease

Ischemic Stroke

Pathophysiology
- Cerebral Infarction; 80% of all strokes
- Thrombotic (2/3), Embolic (1/3)
- Less commonly systemic hypoperfusion, venous occlusion

Clinical Features
- Focal neurological impairment with anterior or posterior circulation features; Persistent
- DDx: TIA, ICH, SAH, Seizure

Evaluation
- Hospitalization; ABCs; O2 Sat
- Labs: CBC, Lipid Profile, BMP with Glucose, PT/PTT, ESR
- CT; MRI w/ diffusion; Angiogram; Transcranial Doppler Ultrasound
- Carotid Duplex Ultrasound
- EKG +/- Telemetry or Holter
- Echocardiogram (TTE/TEE)

Treatment
- ABCs; Maintain BP, fluid volume, normoglycemia; Prevent Aspiration
- Thrombolysis with alteplase (tPA) within 3.0 or 4.5 hours
- Antiplatelet Therapy w/in 48h, unless Rt-PA then after 24h
- Early anticoagulation if cardiac thrombus, Afib; consider bleeding risk
- Carotid endarterectomy (70%-99%)
- DVT Prophylaxis
- Rehab (PT, OT, ST)

Prognosis
- Highly variable
- The prognosis for recovery is better in children than in older adults
Cerebrovascular Disease

Prevention

• Large Vessel Disease
  – **Internal Carotid Artery Stenosis**: Endarterectomy > Stenting; w/in 2 wks
  – **Extracranial Vertebral Artery Stenosis**: Rx, Risk Factor Reduction; Efficacy of surgical intervention unknown
  – **Vascular Dissection**: Anticoagulation vs. Antiplatelet

• Cardiogenic Source of Embolism
  – **Atrial Fibrillation**: anticoagulation (target INR 2.5); radio frequency ablation
  – **Thrombus**: anticoagulation (INR 2-3 for at least 3 months)

• Blood Disorders
  – **Antiphospholipid Antibody Syndrome**: Antiplatelet
  – **Thrombophilia**: Antiplatelet or Anticoagulation

• Risk Factor Reduction: Primary & Secondary Prevention
  – **BP Control; Tobacco Cessation; Weight Management; Normalize Cholesterol; Glycemic Control for DM and Metabolic Syndrome; Minimize ETOH Consumption**
  – **Female-Specific (AHA/ASA 2014)**: Pregnancy, Preeclampsia, Gestational DM, OCPs, Post-Menopausal Hormone Use, Changes in Hormone Status
Cerebrovascular Disease
Secondary Prevention

- Cardiogenic Source of Embolism
  - Anticoagulants more effective than Antiplatelets
    - Vitamin K Antagonist: warfarin PO Daily per INR
    - Direct Thrombin Inhibitor: dabigatran 110mg or 150mg PO Twice Daily
    - Factor Xa Inhibitor: rivaroxaban 20mg PO Daily
      apixaban 5mg PO Twice Daily

- Duration
  - Atrial Fibrillation: longstanding
  - Thrombus: at least 3 months
Case

• **Hx:** A 48yo female with history of AVM presents to the emergency department with complaints of severe bilateral headache and nausea that started 6 hours ago and has been getting progressively worse. She has vomited twice in the past hour. Patient notes new slurring of speech and poor coordination on right side of body. No history of migraine or head trauma.

• **PE:** Patient is in distress. Mild dysarthria. Incoordination of right UE and LE. Unsteady gait.
Cerebrovascular Disease

Intracerebral Hemorrhage

Pathophysiology
• Bleeding directly into brain parenchyma; Increased ICP
• Common Causes: hypertensive vasculopathy, trauma, bleeding diathesis, amyloid angiopathy, illicit drugs, vascular malformation
• Risks: Hypertension, Adv. Age, Vascular Malformations, AntiThrombotic Rx, Tumor

Clinical Features
• Gradual onset over minutes to hours
• Focal neurologic impairment
• Eventual headache, nausea, vomiting
• Decreased LOC if elevated intracranial pressure
• Seizures in 5-30%

Evaluation
• Non-Contrast CT of Head to identify ICH
• In patients without HTN or known cause, consider MRI, CTA/MRA

Treatment
• Aggressive Full Care, ICU for 24hrs
• Goal: contain & limit bleeding; identify and treat cause
• Discontinue anti-platelets, Reverse Anticoagulation
• BP Monitoring & Control +/- ICP
  – HOB 30 degrees, sedation, analgesia
  – Inv: Osmotic diuresis, Vent. Cath, neuromuscular blockade, surgery
• Long-term rehab

Prognosis
• Mortality Rate: 35-52% at 30 days
Case

• **Hx:** A 72yo male with history of uncontrolled hypertension presents to the emergency department with complaints of “horrible”, severe headache without nausea and vomiting that started 20 minutes ago. No history of headaches or head trauma.

• **PE:** BP 164/100. Awake and fully oriented. No focal deficits.
Cerebrovascular Disease

Subarachnoid Hemorrhage

General Info
- Two most common causes: arterial aneurysms (80-85%), vascular malformations, dissection
- Aneurysmal type most common between 40-60yo

Pathophysiology
- Bleeding into CSF under arterial pressure leads to increased intracranial pressure
- Risk Factors: Tobacco, FamHx, HTN, Heavy ETOH Use

Clinical Features
- Headache: sudden, maximal at onset
  “Worst headache of my life”
- N/V, mental status change, elevated BP
- Meningismus, Vitreous Hemorrhage, LOC, seizure
- “Sentinel” Headache in 30% with minor bleed that precedes major bleed

Evaluation
- Emergent CT
- CSF Analysis if CT negative
- Cerebral Angiogram, CTA, MRI/A
- CBC, BMP, Coag, Toxicology

Treatment
- Aggressive Full Care, ICU
- Goals: Identify & treat cause to prevent re-bleeding; manage complications
- ICP & BP Control
  – Ventriculostomy for elevated ICP, acute hydrocephalus
- Surgical – clip or coil aneurysm
- Nimodipine improves outcomes

Prognosis
- Rebleeding common w/in 24h
- Complications: Rebleeding, Vasospasm, Hydrocephalus, Seizures
- Mortality Rate: 50% w/in 30d
Case

• **Hx:** A 22yo female presents to your outpatient clinic with complaints left vision loss associated with mild eye pain that start about 4 hours ago. Prior neurologic symptoms include 12 days of right vision impairment 2 years ago and 14 days of right arm incoordination 5 years ago.

• **PE:** Left visual acuity 20/70 with correction. Right visual acuity 20/25 with correction. No field cut. Fundoscopic exam reveals swelling of the right optic disk, blurring of disk margins. Otherwise non-focal.
Multiple Sclerosis

General Information
- Relapsing Remitting, Primary Progressive, or Secondary Progressive
- 18-45yo, F:M 2:1

Pathophysiology
- Etiology unknown
- T-cell mediated multifocal CNS inflammation and demyelination; then gliosis
- Possible viral precipitant
- Genetic factors

Clinical Features
- Episodic focal neurological impairment involving multiple CNS regions with or without complete recovery
- Separated by space & time
- Common presentations: optic neuritis, transverse myelitis

Evaluation
- MRI with Contrast
- CSF: protein, IgG, Oligoclonal Bands, Myelin Basic Protein
- Evoked Potential Studies

Treatment
- Acute Inflammation: high dose IV or Oral corticosteroids
- DMTs: interferons; glatiramer acetate; natalizumab (risk progressive multifocal leukoencephalopathy), fingolimod, teriflunomide, dimethyl fumarate, alemtuzumab, daclizumab
- Symptomatic

Prognosis
- Variable comorbid problems: fatigue, mood, cognition, pain, spasticity, gait impairment, bladder dysfunction
Case

• **Hx**: A 71yo male is admitted to your service through the ER with complaints of 3-day history of sensory loss in both lower extremities that is progressively ascending and now involves his hands. He feels strong, but says he is unsteady. No respiratory complaints.

• **PE**: Vital signs are stable. Reduced tactile, vibratory, and temperature discrimination throughout the entire lower extremities. Hands demonstrate reduced tactile sensation. Incoordination of bilateral LE and hands. DTRs absent in bilateral LE, trace at biceps.
Guillain-Barre Syndrome

General Information
- Acute Inflammatory Demyelinating Polyneuropathy (AIDP) Variant

Pathophysiology
- Autoimmune-mediated demyelination or axon injury → dysfunction of peripheral nerves
- Antecedents may include infection (2/3), immunization, surgery, BMT

Clinical Features
- Acute (hrs to days); Monophasic
- Progressive, Mostly Symmetric, Motor > Sensory Impairment
- Distal → Proximal
- Decreased or Absent DTRs
- Respiratory Impairment
- Miller-Fisher Variant: opthalmoplegia, ataxia, areflexia

Evaluation
- NCS: slow nerve conduction; denervation, axon loss
- CSF: elevated protein
- M-F Variant: Serum IgG to GQ1b

Treatment
- Inpatient Management
- Acute: Plasmapheresis, IV Ig
- Monitor Respiratory Status, BP
- Long-Term: PT, OT, ST

Prognosis
- Can be life-threatening if respiratory or swallowing muscles are affected
- Ventilatory Support (30%)
- Autonomic Dysfx (70%)
- Slow, often incomplete recovery; chronic type
Case

- **Hx:** A 40yo female presents for her annual physical examination. During ROS, the patient reports a 6 month history of muscle fatigue and double vision later in the day. Symptoms improve if she rests. No other deficits.

- **PE:** Normal cranial nerve examination, except of bilateral ptosis that is noted after prolonged eye closure. Strength initially 5/5, then 4/5 after demonstrating prolonged effort. Otherwise non-focal.
Myasthenia Gravis

General Information
• Generalized and Ocular Types
• More common in young women and older men

Pathophysiology
• Autoimmune destruction of acetylcholine receptors (AChR) on the surface of skeletal muscles
• Thymus involvement

Clinical Features
• Insidious onset; Fluctuating
• Muscle weakness, fatigability (CNs, Extremities, Respiration)
• Symptoms improve with rest
• Infection may exacerbate symptoms
• Normal Sensation, No DTR Change
• Myasthenic Crisis
• Ocular Type: ptosis, EOMs Weak

Evaluation
• Edrophonium (Tensilon) Test
• Chest CT: R/O Thymoma
• EMG/NCS with Rep Nr Stim: slow muscle response
• Serum AChR Ab; Serum MuSK Ab; TFTs (Ocular Type)

Treatment
• Cholinesterase inhibition: pyridostigmine
• Refractory Sx: Immunotherapy
• Rapid Tx: IVIG or Plasmapheresis
• Thymectomy if hyperplasia in AChR Ab positive MG

Prognosis
• Some Rx can exacerbate
• Maximal extent at 3yrs in majority of patient
• Course may be progressive or relapsing-remitting
Case

• **Hx:** A 2yo male is brought in for his routine well child check. The patient’s parent report that the child is still struggling with gait unsteadiness and incoordination. The other concern is that his vocabulary only consists of a couple words. Birth history is remarkable for prolonged labor and chorioamnionitis.

• **PE:** Remarkable findings include diffusely decreased muscle tone, incoordination of upper extremities, unsteady gait.
Cerebral Palsy

General Information
- 2-4/100,000 children

Pathophysiology
- Multifactorial
- Cerebral injury before, during, or after birth; Prenatal Risk Factors
- Types: Spastic, Dyskinetic, Ataxia

Clinical Features
- Chronic, non-progressive impairment of muscle tone, strength, coordination, or movements
- Clinical expression may change over time
- Spasticity (75%); Seizure Disorder (45%); Intellectual Disability; Specific Learning Disabilities

Evaluation
- Serial physical examinations
- Monitor if prenatal risk factors
- Classify type
- MRI Brain for etiology, prognosis
- Metabolic and Genetic testing
- EEG if seizure suspected

Treatment
- Supportive, Multidisciplinary Team
- Goal: Maximize Function
- Rx: Spasticity, Seizures
- Prev: MagSulf in PreTerm Labor

Prognosis
- Comorbidities: poor growth, orthopedic problems, osteopenia, urinary disorders, hearing, vision, cognition
Case

- **Hx:** A 21yo male with history of partial complex epilepsy presents to the emergency department after experiencing two seizures in the past 4 hours. Patient describes the seizures as similar to those in the past and verifies adherence with his prescribed anti-epileptic drug (AED). ROS is remarkable for sleep deprivation the past three nights due to studying.

- **PE:** Vital signs stable. Non-focal.

- **Dx:** Serum AED level is therapeutic.
Seizure Disorders

General Information
• Prevalence: 0.5-1%

Pathophysiology
• Not fully understood
• Epileptic Seizure: Inherent, recurrent abnormal paroxysmal neuronal discharges
• Secondary: Provoked
• Non-Epileptic: Psychogenic

Clinical Features
• Stereotypical, Random
• Generalized: convulsive or non-convulsive
• Focal: simple(partial) or complex
• Status epilepticus = medical emergency
  – Tx: benzodiazepam
• Post-Ictal State
• Secondary report from witness

Evaluation
• Determine if epileptic event
• EEG
• Neuroimaging (MRI)
• Labs for metabolic and toxic causes; Prolactin

Treatment
• Epilepsy: Antiepileptic Drugs (AEDs): Mono→Mono→Poly-therapy
• Surgery, Nerve Stimulation
• Women of Child-Bearing Age
  – AEDs may affect OCPs
  – Folate Supplementation
  – Caution with valproate, carbamazepine
• Manage secondary causes

Prognosis
• Drug Resistant Epilepsy: 20-40%
• Driving Restrictions
BREAK
Case

• Hx: A 42yo male with presents to an urgent care with complaints of drooling and “droopy face” on the right for the past 24 hours. He states this was preceded by a couple days of tingling on the right side of his face.
• PE: Moderate right facial droop and ptosis. Other CNs unremarkable. Remainder of exam is non-focal.
Disorders of the Peripheral Nerves

Mono-Neuropathies

Types
• Carpal Tunnel Syndrome
• Ulnar Neuropathy
• Radial Neuropathy
• Meralgia Paresthetica
• Femoral Neuropathy
• Sciatic Nerve Palsy
• Peroneal Nerve Palsy
• Tibial Neuropathy
• Cranial Nerve VII Palsy

Pathophysiology
• Most Common: Compression

Clinical Features
• Focal Sensory and/or Motor

Evaluation
• Occupational/Activity History
• Special Physical Exam Tests
• EMG/NCS

Treatment
• Conservative: modify activities, brace, PT/OT, Anti-Inflammmatories
• Invasive: Surgery, Steroid Injections
Case

• Hx: A 50 year-old female with history of HTN and DM presents to the office for routine follow-up. ROS is positive for numbness in the distal upper and lower extremities for the past 6-9 months that has been slowly progressive. No motor weakness or autonomic symptoms.

• PE: Vital signs stable. Strength symmetric. 1+ DTRs in the UE and LE. Diminished vibratory, proprioception, and light touch in the distal extremities compared to proximally.

• Dx: HgA1C is 8.5.
Disorders of the Peripheral Nerves
Poly-Neuropathies

Etiologies
- Hereditary
  - Charcot Marie Tooth Disease
- Endocrine
  - Diabetes, Uremia, Alcohol, B12 Deficiency
- Infectious
  - HIV/AIDS, Leprosy, Lyme Disease
- Inflammatory
  - Sarcoidosis, Polyarteritis, Rheumatoid Arthritis
- Toxic
  - Industrial Agents, Pesticides, Heavy Metals, Medications
- Paraneoplastic Syndromes
- Neuropathy Associated with Critical Illness

Clinical Features
- Motor, Sensory, Autonomic
- Proximal or Distal
- Acute or Chronic

Evaluation
- Decreased DTRs
- Select Labs: CMP, HgA1C, B12, Toxicology, Paraneoplastic Panel
- EMG/NCS

Treatment
- Direct at cause
- Symptomatic to support motor activity, sensory discomfort, and autonomic function
- Refer when indicated

Prognosis
- Variable depending on etiology
Case

• Hx: A 45 year-old female with history of right metatarsal fractures 8 months ago presents for routine follow-up with complaints of persistent right foot pain, occasional swelling, and skin discoloration. She denies recurrent trauma and weakness.

• PE: Hyperesthesia of skin on the right foot with trace edema and discoloration. No orthopedic deformity. Gait guarded, steady.

• Dx: Plain radiograph of the right foot demonstrates healed fractures.
Disorders of the Peripheral Nerves

Complex Regional Pain Syndrome

General
• Type 1: un-definable nerve lesion, AKA reflex sympathetic dystrophy, 90% of cases
• Type 2: definable nerve lesion

Pathophysiology
• Unclear; may involve reflex arc
• Frequently begins following an injury, surgery, or vascular event such as a stroke

Clinical Features
• Involve specific body region, usually the extremities
• Characterized by pain, swelling, limited range of motion, vasomotor instability, skin changes, and patchy bone demineralization.
• 3 Stages: Early, Subacute, Chronic

Evaluation
• Clinical Diagnosis
• Autonomic Testing
  – Resting sweat output (RSO), resting skin temperature (RST), and quantitative sudomotor axon reflex test (QSART)
• Consider bone scintigraphy, plain radiographs, or MRI
• Response to nerve block

Treatment
• Prevention: early mobilization, rehab
• Prevention: Vitamin C supplementation for fractures
• Pain management
• Pharmacologic and invasive procedures

Prognosis
• Variable
Case

• Hx: A 68 year-old male with history hypertension presents to the clinic with his spouse for his annual physical. His spouse reports that the patients memory is “less sharp than ever.” The patient says he often forgets where things are and has gotten lost while driving a few times recently.

• PE: Disorientation, poor short-term recall, poor attention span, delay in naming objects. No other focal deficits.
Dementias

General Information
- Alzheimer (AD): 60-80%
- Lewy Body; Vascular Type; Mixed Type
- Diagnostic Criteria: DSM IV TR, DSM V

Pathophysiology
- Unknown; Genetic & Environmental

Clinical Features
- AD: Intellectual impairment with compromise in at least two of the following: language, memory, visuospatial skills, emotional behavior, personality, and cognition
- Lewy Body Dementia: impaired cognition with Parkinsonian features
- Vascular: forgetfulness, attention maintained
- DDx: Parkinson Dementia, Alcohol-Related Dementia, Frontotemporal Dementia, Cerebrovascular Disease, Delirium, Depression

Evaluation
- History, Physical Exam, Secondary Reports, Screen with MMSE (<24)
- Neuropsychological evaluation
- TSH, B12 deficiency, CBC, CMP, heavy metals, drug/alcohol screen; consider imaging

Treatment
- All Types: acetylcholinesterase inhibitors - donepezil, rivastigmine, and galantamine; Vitamin E
- AD: N-methyl-D-aspartic acid (NMDA) receptor antagonist; Vitamin E
- Lewy Body: dopamine agonists
- Vascular: control BP, metabolic abnormalities
- Supervision & Safety
- Behavioral Management
- Caregiver support

Prognosis
- Variable progression

http://www.alzinfo.org/top-alzheimers-symptoms.asp 07/26/10
Case

• Hx: A 77 year-old female patient has been admitted through the emergency department with a diagnosis of urosepsis. Throughout the 2-day hospitalization, the patient has had a fluctuating mental status and is agitated at times. Nursing staff reports that the patient has been hallucinating at night. There is no history of cognitive disorder.

• PE: Oriented to self, mild distress, poor attention span, distractible, without hallucinations. Otherwise non-focal.

• CT Scan Head, CBC, CMP: Unremarkable
Delirium

General Information
- AKA: acute confusional state, encephalopathy

Pathophysiology
- Poorly understood
- Multiple risks & precipitants
- May be caused by medical condition, medicine, or intoxication; if not, NOS

Clinical Features
- Disturbance of consciousness with reduce ability to focus, sustain, or shift attention
- A change in cognition or new perceptual disturbance
- Sx develop over hours to days, tend to fluctuate
- Atypical Symptoms: focal or lateralizing deficits

Evaluation
- History & Physical Exam
- Confusion Assessment Method (CAM)
- Medication review, electrolytes, infection, intoxication, withdrawal, metabolic, low perfusion, postoperative
- Select tests based on DDx

Treatment
- Direct at the cause; Safety measures; Close follow-up
- Consider Thiamine; Caution with Psychotropic Rx
- Prevention: minimize risk factors, environmental modification, orientation protocols, sensory aids, no restraints

Prognosis
- May persist for days to months
- Increased mortality

Case

• Hx: A 25 year old female presents to an urgent care after an episode of passing out while at a crowded conference. The patient reports she felt lightheaded before passing out. The patient denies incontinence and post-event confusion. Witness report is that there were no convulsions. The patient says this has happened before once while in church.

• PE: Normal mental status examination. Non-focal. No evidence of trauma.

• Dx: Serum Glucose 95, Pulse Oximetry 92%
Syncope

Pathophysiology
• DDx: arrhythmia, aortic stenosis, carotid sinus hypersensitivity, MI, hypoglycemia, orthostatic hypotension, postprandial hypotension, psychogenic, PE, vagal faint, TIA, seizure

Clinical Features
• Sudden, transient loss of consciousness (LOC) with loss of postural tone; not due to trauma; rapid spontaneous recovery
• Differentiate from vertigo and pre-syncope
• Exertion
• Witness report helpful

Evaluation
• History & Physical Examination; PMHx CAD, CVD
• Stratify risk for sudden cardiac death
• EKG
• Consider: Ambulatory Cardiac Monitoring, Echocardiogram, Tilt Table Test, Electrophysiologic Studies, CT or MRI Brain

Treatment
• Direct at the cause
• Consider driving restrictions

Prognosis
• Recurrence is variable based on cause and treatment success
Case

- **Hx:** An 18 year old high school senior presents to your office one day after “taking a hard hit to the head” while playing ice hockey. He denies remembering what happened in the couple minutes preceding the head injury. He denies passing out, though describes himself as “dazed” and not quite himself today. **ROS** is positive for mild nausea and frontal headache.

- **PE:** Normal mental status examination, except for reduced attention span and difficulty naming 2 of 5 objects. Otherwise non-focal
Concussion (Traumatic Brain Injury)

General Information
• Annual Incidence Sports-Related Concussion (US): 1.6-3.8 million

Pathophysiology
• Due to contact and/or acceleration/deceleration force
• Coup/Contracoup contusions; axonal rupture

Clinical Features
• Concussion: trauma-induced change in mental status +/- loss of consciousness
• Early: Amnesia, HA, awareness, dizziness, N/V
• Late: mood, cognition, sleep, sensitivity to light and noise
• Seizures (5%)

Evaluation
• Medical or Trained Professional
• Standardized Assessment of Concussion Monitor for Complicated Concussion

Treatment
• Sports-related: remove from play that day
• Identify immediate neurologic emergencies and high-risk individuals for second impact syndrome
• Full physical Rest
• Manage neurologic sequelae
• Prevention of cumulative and chronic brain injury (i.e. limit repeated injury)
• Monitor for 24 hours; Hospitalization for GCS <15, abnormal CT, Seizures, or predisposition for bleeding
• Neurobehavioral Evaluation

Prognosis
• Second Impact Syndrome: diffuse cerebral edema that occurs after 2nd concussion; rare and controversial
• Return to Play Protocol: graduated approach, when full recovery evident
Post-Concussion Syndrome

General Information
• May occur following any type of TBI; 30-80% of cases
• More common in females and elderly

Pathophysiology
• Theories: structural, biochemical, psychogenic

Clinical Features
• Headache, dizziness, fatigue, neuropsychiatric Sx, cognitive impairment, insomnia, anxiety, irritability, noise sensitivity

Evaluation
• Clinical Diagnosis
• Judicious testing, neuropsych
• MRI to exclude other causes
• Referral for Prominent Symptoms, Psychiatric D/O

Treatment
• Individualized & Symptomatic
• Education & Reassurance

Prognosis
• Symptoms most severe: 7-10 d
• Majority subclinical: 30 d
• Vast majority recover: 3 mos
• 10-15% have persistent PCS
Case

• **Hx:** A 21 year male presents to the emergency department with decreased level of consciousness. The patient is accompanied by a friend who states the patient was hit in the head by “some idiot at the bar.” The patient is mumbling to himself and denies complaints.

• **PE:** Lethargic, oriented to self, inconsistently follows simple commands, poor attention span, slurred speech, names 2 of 3 objects. Contusion along left temple. Otherwise non-focal.

• **Dx:** CT Head – no acute process, left scalp swelling

  UDS – Positive for ethanol
Altered Level of Consciousness

General Information
• Alert ↔ Coma Spectrum
• Coma: “unarousable unresponsivness”

Pathophysiology
• Impairment of the Reticular Activating System (RAS)
• Common Etiologies: trauma, CVA, intoxication, metabolic, infection

Clinical Features
• Abnormal vital signs, ventilatory pattern, trauma
• Decreased LOC, Motor Responses, Cranial Nerve Reflexes

Evaluation
• Glasgow Coma Scale (GCS)
• Detailed Neuro Exam
• Labs: CBC, CMP, ABG, aPTT, PT, Toxin Screen
• Neuroimaging, CSF, EEG, EKG

Treatment
• Airway, Breathing, Circulation
• Manage cause
• Close Monitoring

Prognosis
• Dependent upon cause, severity of brain injury
• GCS
• FOUR Score
Case

- **Hx**: A 48 year old male presents for his annual physical. He has no new complaints and is not on any medications.

- **PE**: Bilateral action tremor in the upper extremities, “to and fro” in quality, that is absent at rest. Normal speech. No head titubation. No rigidity or increased tone. Otherwise non-focal.

- **FHx**: No family history of tremor.
Movement Disorders

Essential Tremor

General Information
• Most common cause of tremor: 5% worldwide
• 50% of Cases are Familial, Autosomal Dominant

Pathophysiology
• Unclear; Genetic Factors
• Possible involvement of brainstem, cerebellum

Clinical Features
• Longstanding, Progressive
• Chronic Bilateral Action Tremor; Absent at Rest
• To-and-Fro Tremor
• UE > LE; Head Titubation; Speech Impairment
• No other deficits; no dystonia

Evaluation
• Clinical Diagnosis
• Assess for Family History of Tremor
• Evaluate for neurologic deficits, Parkisonian Sx
• Beneficial response to alcohol

Treatment
• AAN Guidelines [2011]
• 1\textsuperscript{st} Line: propanolol or primidone
• 2\textsuperscript{nd} Line: Beta Blockers, gabapentin, topiramate
• Refractory: Botox, DBS, Thalamotomy
• Alcohol: discouraged

Prognosis
• Variable
Case

• Hx: A 40 year old male with a family history of Huntington’s Disease presents for follow-up with complaints of an increase in involuntary movements. The patient’s partner reports new behavioral changes, including irritability and poor memory.

• PE: Impaired short-term recall, otherwise mental status normal. Frequent choreiform movements of the upper extremities and neck. Slight dystonia in upper extremities. Otherwise non-focal.
Huntington Disease

General Information
• Rare: 5-8/100k (U.S., Europe)

Pathophysiology
• Progressive Neurodegeneration
• Trinucleotide (CAG) expansion in the Huntington gene on chromosome 4p
• Autosomal-Dominant

Clinical Features
• Choreiform Movements & Dystonia
• Psychiatric & Behavior Changes
• Dementia
• Slowly Progressive

Evaluation
• Clinical Diagnosis
• Family History
• Genetic Testing
• Neuroimaging: late stage caudate atrophy

Treatment
• Supportive, Interdisciplinary Care; No Cure
• Movement: tetrabenazine; otherwise atypical then typical neuroleptics
• Psychosis: neuroleptics
• Depression

Prognosis
• Usually fatal within 15-20 years
• Family Genetic Testing
Case

• Hx: A 70 year old male reports to the urgent care after falling at home. He states “I lost my footing.” His wife says that he propelled forward and was unable to stop. He has no other complaints.

Movement Disorders

Parkinson’s Disease

General Information
- Mean Onset: age 70
- Prevalence: 0.3% (general pop.); 3% population over > 60 yo

Pathophysiology
- Degeneration substantia nigra
- Decrease dopamine, imbalance of acetylcholine

Clinical Features
- Progressive Symptoms
- Cardinal Features: Resting Tremor, Bradykinesia, Rigidity
- Also Postural Instability, Shuffling Gait, Masked Facies, Stooped Posture, Micrographia
- Non-Motor: Depression, Fatigue, Cognitive Impairment, Psychosis

Evaluation
- Clinical Diagnosis
- Sx respond to dopamine
- Rule out secondary causes (drug induced)

Treatment
- Dopaminergic: levodopa/carbidopa, bromocriptine
- Anticholinergic: amantadine, benztropine
- Adjuncts: COMT, MAO B Inhibitor; Deep Brain Stimulation
- PT; Psych; Family Support

Prognosis
- Variable Progression
Case

• Hx: A 5 year old male presents to the office with his mother for a “problem visit.” The mother reports that her son “has been blinking too much.” There is no associated neurologic deficits, mental status change, or developmental issues.

• PE: Non-focal examination, except for 3 episodes of frequent blinking with associated facial grimacing.
Tourette Syndrome

General Information
• Disorder manifested by motor and phonic tics
• 2-8/1,000 (worldwide)
• Onset: Childhood (2-15yo)

Pathophysiology
• Genetic predisposition
• CNS dis-inhibition of the motor and limbic system

Clinical Features
• Tics: sudden, brief, intermittent movements (motor) or utterances (vocal or phonic)
• Simple or Complex
• Comorbidities: ADHD, OCD

Evaluation
• Clinical Diagnosis
• Other behavioral disorders
• Family History

Treatment
• Education for patient, family, teachers, peers, etc.
• Rx when Function Impaired
  – Dopamine Agonists (fluphenazine, pimozide), Antagonists (tetrabenazine)
  – Botulinum toxin injection
• Habit Reversal Training
• Treat Comorbidities

Prognosis
• 50% of cases resolve by age 18
• Severity diminishes during adulthood
Case

• Hx: A 25 year old female is admitted through the emergency department with complaints of headache, fever, and lethargy, that started in the last 12 hours.


• Dx: CT Head non-contrast – Normal Study CSF – elevated protein, WBC; decreased glucose; Cx pending
CNS Infection
Bacterial Meningitis

General Information
• Strep pneumoniae, Neisseria meningitidis, group B streptococci (infants), ↓Hflu since ↑HIB Immunization
• Medical Emergency
• Prevention: Immunization against N. meningitidis, S. penumoniae

Pathophysiology
• Inflammation, Cytokine mediated
• ↑ Intracranial Pressure (ICP)
• Tissue Necrosis

Clinical Features
• Triad: Fever, Nuchal Rigidity, Mental Status Change
• Headache – generalized, severe

Evaluation
• CT: R/O Space-Occupying Lesion if risk factors present
• CSF: ↑ICP, ↑WBC (1-10k), ↑Prot, ↓Glucose, turbid-purulent, Gram Stain & Culture
• Blood Cx: Positive in 50-90%

Treatment
• Empiric Antibiotics after LP
• Dexamethasone to reduce neurologic complications
• Pathogen-Directed Antibiotics

Prognosis
• Mortality Rate 100% w/o Tx
• Monitor for Response and Complications
Case

• Hx: A 32 year old male is admitted through the emergency department with complaints of headache, nausea, and malaise, that started the yesterday. No focal complaints. Last week he had gastroenteritis that has resolved.


• Dx: CT Head non-contrast – Normal Study
  CSF – normal protein, WBC, glucose; Cx pending.
CNS Infection
Viral Meningitis/Encephalitis

General Information
• AKA “Aseptic”, Cx Negative
• ↑ Incidence Summer, Fall
• Coxsackievirus A/B, echoviruses, mumps

Pathophysiology
• Inflammation from virus or from perimeningeal area (ears, sinuses)
• Risk Factors: travel, ticks, STIs

Clinical Features
• Meningitis: headache, fever, nausea or vomiting, malaise, photophobia, and meningismus
• Encephalitis: mental status change, motor/sensorhy deficits, behavior change
• Seizures can occur with either

Evaluation
• History to ID possible exposures
• CT: R/O Space-Occupying Lesion
• CSF: ↑WBC (1k); Prot, Glucose, & ICP WNL; clear; Gram Stain & Culture Neg
• HSV PCR; CSF and serum IgM Ab for West Nile Virus
• Rule bacterial cause

Treatment
• Acyclovir if + HSV, encephalitis
• Symptomatic
• Consider Empiric ABX in elderly, immunocompromised, or if on ABX

Prognosis
• Benign and self-limited
Case

• Hx: A 48 year old female presents for a problem visit with complaints of frequent headaches, which she describes as a “band around” her head that lasts for 2-3 hours approximately twice per week. No associated features or triggers. OTC NSAIDs help “some.” These headaches have been present for years, though have been more bothersome lately due to work-related stress.

• PE: Non-focal examination except for mild scalp tenderness.
Headache Disorders

Tension Type Headache

General Information
• Most common type of headache
• Females > Males

Pathophysiology
• Unknown, Multi-factorial; Genetic
• Abnormal neuronal sensitivity in CNS misinterprets normal stimuli as noxious

Clinical Features
• Mild to moderate intensity, bilateral, non-throbbing headache without other associated features (phonophobia, nausea, vomiting) or aggravation with physical activity
• Pericranial muscle tenderness
• Triggers: psychosocial stress, migraine

Evaluation
• International Headache Society (IHS) Classifications [2004]
  – Infrequent Episodic TTH: HA 1d/mo
  – Frequent Episodic TTH: HA 1-14 d/mo
  – Chronic TTH: HA 15 or more d/mo
• Neuroimaging if indicated

Treatment
• Abortive: ibuprofen, naproxen, acetaminophen; ketorolac IM; caffeine as adjunct
• Preventive: amitriptyline, SSRIs, anticonvulsants; behavioral including stress management, EMG biofeedback, PT, acupuncture

Prognosis
• Variable
• Avoid medication overuse headache: limit acute Treatment to 9 days/month, 2 doses/day
Case

• Hx: An 18 year old female with a history of migraine headache presents for her college physical. She continues to experience 2-3 headaches per month, which she characterizes as “throbbing on the left side” with associated nausea, photophobia, and malaise. The headaches typically last 3-4 hours and are effectively aborted with prescribed “triptan” and rest. She inquires if there is a medication she can take to prevent the migraines. No current headache

• PE: Non-focal examination.
Headache Disorders

Migraine Headache

General Information
• Prevalence: 12%; F>M
• Migraine without Aura (75%)

Pathophysiology
• Multi-factorial; Genetics
• Common Triggers: Stress, Hormones, Not Eating, Weather, Sleep Disturbance, & Others

Clinical Features
• Prodrome: affective or vegetative symptoms
• Aura: focal neurologic symptoms
• Headache: unilateral, “throbbing”, +/- nausea, vomiting, phonophobia, photophobia
• Postdrome: exhaustion

Evaluation
• IHS Diagnostic Criteria for w/ Aura and w/o Aura
• Imaging if atypical symptoms, focal deficits, immunocompromised

Treatment
• Abortive: NSAIDs, acetaminophen, triptans, DHE, anti-emetics; prompt Treatment
• Prophylaxis if >4 HA/Month, HA > 12 hrs, HA \( \rightarrow \) disability
• Prophylactic: BBs, CCBs, ACE/ARBs, TCAs, Anticonvulsants; Behavioral Therapy

Prognosis
• Variable
• Education; Avoid Triggers

Case

• Hx: A 38 year old male presents to the emergency department with complaints of recurrent episodes of severe, “overwhelming” and “shock-like” pain behind his right eye in the past 48 hours. There is associated tearing of the right eye during the pain, but no other associated symptoms. He has never had these headaches before. He denies having a headache currently.

• PE: Non-focal examination.
Cluster Headache

General Information
• Most common type of trigeminal autonomic cephalalgias
• Prevalence: <1%;
• M:F Ratio - 4.3 :1

Evaluation
• Clinical Diagnosis
• IHS Diagnostic Criteria for episodic & chronic types
• Imaging if mass lesion suspected

Pathophysiology
• Unclear
• Genetic factors

Clinical Features
• Bouts & Remissions
• Short-lasting (15-180 minutes)
• Unilateral: Periorbital, Supraorbital, Temporal
• Severe Pain
• Autonomic Symptoms

Treatment
• Abortive: oxygen , if no effect then triptans
• Prophylactic: begin at diagnosis; verapamil, low dose prednisone
• Surgical options under investigation

Prognosis
• Recurrence 80% at 15 years after Diagnosis
• Variable
References


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