Neurology Review

Categories

- CNS Infections
- Auto-Immune Disorders
- Epilepsy and Sleep
- Vascular Diseases
- Headache and Pain Syndromes
- Trauma
- Degenerative Disorders/Dementia
- Altered mental Status
- Movement Disorders
- Structural Disorders
- Toxic/Metabolic Disorders
- Psychiatric
- Neuromuscular Disorders
- Localization/Anatomy
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Bacterial Meningitis

- Organisms: Streptococcus pneumonia, Neisseria meningitidis, Hemophilis influenzae
- Organisms in infants: Group B Streptococcus, E. coli, Listeria monocytogenes
- Symptoms: fever, nuchal rigidity, headache with photophobia, altered mental status, +/-focal neurological signs, seizures
- Lab: CSF shows high WBC with PMN’s predominating, Protein elevated, low Glucose, Gram Stain/Cultures positive
- Treatment: Ceftriaxone vs. Vancomycin
  - Chemoprophylaxis of contacts (N.m. and H.i.) with Rifampin
  - Do not delay antibiotics if LP cannot be immediately performed
- Complications: Sensorineural hearing loss, etc
Neurology Review

Aseptic Meningitis

- Viral meningitis is more common than bacterial meningitis
- Organisms: Enterovirus, Arbovirus, HSV
- Symptoms: Headache, Fever, Nuchal rigidity, no seizures, no altered mental status, no focal findings
- Diagnosis: CSF with a lymphocytic pleocytosis, Protein slightly elevated, normal Glucose, negative cx’s and gram stain
- Treatment: Supportive. Can start antibiotics until cx’s negative
Neurology Review

Chronic (fungal or mycobacterial) Meningitis

- Generally presents in a more sub-acute fashion
- Signs of raised ICP (papilledema, CN VI dysfunction)

CSF Profile:

- WBC: Very elevated, mostly lymphocytes
- Protein: Very elevated, especially with TB meningitis
- Glucose: Low
- Detection: India ink stain for fungi; Cryptococcal Antigen; fungal cultures; TB PCR and cultures

Organisms:

- TB: Basilar meningitis with CN deficits. Tuberculoma with focal CNS deficits.
- Cryptococcus, Coccidiodes, Histoplasma, Candida.
Encephalitis

- Inflammation of the brain parenchyma, usually viral
- Organisms:
  - Sporadic: HSV (anterior temporal lobe involvement), Arboviruses, West-Nile
  - Chronic: Measles, Rubella, HIV
- Symptoms: Altered MS, Seizures, focal deficits. Headache and fever less common
- Diagnosis: CSF with a lymphocytic pleocytosis, protein +/- elevated, normal Glucose, neg Cx’s G/S. PCR for enterovirus, HSV, viral titres, viral cultures (low yield)
- Treatment: Acyclovir (only for HSV), supportive care
Brain Abscess

- Organisms: Streptococci (anaerobic), Staphylococcus, opportunist organisms if immunocompromised
- Direct extension from sinusitis, mastoiditis, or hematogenous spread (Mycotic aneurysms may arise from septic emboli from the heart)
- Symptoms: Headache from raised ICP, focal deficits (including VI palsy), +/- fever
- Diagnosis/Treatment: MRI or CT with contrast. No LP! (risk of herniation) Broad spectrum antibiotics. Staph aureus coverage in Subacute Bacterial Endocarditis
- Neurocysticercosis (Taenia Solium)
  - Multiple cerebral calcifications, scolex formation
Neurology Review
AIDS and the Nervous System

- Primary HIV syndromes:
  - Subacute Encephalitis (AIDS Dementia Complex)
  - Vasculitis
  - Sensory Polyneuropathy
  - Meningitis
  - Immune Reconstitution Inflammatory Syndrome
    - Unmasking of an occult infection or symptomatic relapse of prior infection (especially TB or Cryptococcal meningitis)
Neurology Review
AIDS and the Nervous System

- Opportunistic Infections: (Can also be seen with T-Cell immunodeficiency)
  - Toxoplasmosis (vs CNS Lymphoma)
    - Rx Toxoplasmosis with Pyrimethamine and Sulfadiazole
  - Cryptococcal meningitis
  - Progressive Multifocal Leukoencephalopathy (JC virus)
  - Treponema Pallidum (Neurosyphilis)
    - Subacute Dementia (General Paresis), vasculitis, meningitis, gummas, Tabes Dorsalis (posterior column dysfunction, lancinating pains, dropped DTR’s, sensory ataxia, Argyll Robertson pupils (light-near dissociation)
  - CMV
    - Iritis, retinitis, optic neuritis, ventriculitis, meningitis, encephalitis, myelitis, radiculitis, polyneuritis
Neurology Review

Congenital “TORCH” Infections

- Toxoplasmosis, “Other” (Syphilis, VZV, HIV, etc), Rubella, CMV, HSV
- Typical presentation of IUGR, microcephaly, HSM, jaundice, seizures, retinitis, and sensorineural hearing loss
- Neonatal HSV can present with skin (rash), eye (retinitis, keratitis), or brain (encephalitis) symptoms. Risk of transmission is highest if primary maternal infection is acquired in 3rd trimester of pregnancy
- CMV associated with sensorineural hearing loss which can be progressive over the first year of life and may not be picked up at birth.
- Congenital syphilis is transmitted from maternal secondary syphilis during pregnancy
  - Early (<2 years): HSM, skeletal anomalies, bullous skin lesions, pneumonia and rhinorrhea
  - Late (>2 years): Blunted upper incisors (Hutchinson’s teeth), saddle nose, saber shins, interstitial keratitis, sensorineural deafness
Other Infections

- **Lyme Disease**
  - Meningitis, encephalitis, cranial neuritis (especially VII), radiculitis, mononeuritis multiplex
  - Diagnosis supported with CSF pleocytosis and ITAb’s to BB
  - Doxycycline or IV Ceftriaxone if severe disease
  - Treatment of Chronic Lyme disease is controversial and not supported

- **Rabies**
  - Early: Presents with fever, headache, weakness (non-specific)
  - Later: Delerium, dysphagia/drooling (hydrophobia) rapidly leading to death

- **VZV**
  - Shingles and post-herpetic neuralgia from reactivation of latent infection due to declining T-Cell immunity
  - Ear pain (external auditory canal rash) and Bell’s Palsy (Ramsay Hunt Syndrome)
  - Bell’s Palsy. Rx with oral steroids (oral acyclovir controversial)
Brain Abscess

HSV Encephalitis

Neurocysticercosis
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Multiple Sclerosis

- Epidemiology: Prevalence: 100-150/100,000, Females>Males, North-South Gradient
- Diagnosis:
  - Typical demyelinating lesions/neurological deficits referable to the CNS separated in space and time
  - Clinically Isolated Syndromes
    - Optic Neuritis: central scotoma, pain with eye movement, papillitis or normal fundus if retrobulbar
    - Transverse Myelitis: flaccid paralysis, sensory level, B/B dysfunction
    - Other focal deficits: brainstem (INO) or cerebellar.
  - Single demyelinating “attack” with MRI features suggestive of multiple lesions of different age (MacDonald Criteria)
  - CSF profile showing intrathecal synthesis of IgG, Oligoclonal Bands +/- low grade lymphocytic pleocytosis
Multiple Sclerosis

Disease Progression
- Relapsing/Remitting
- Secondly Progressive
- Neuromyelitis Optica [multisegment transverse myelitis + optic neuritis + NMO (Aquaporin) Ab’s]

Treatment
- Glucocorticosteroids, plasmapharesis, or IVIg for acute attacks
- Disease Modifying Therapy...
**Neurology Review**

**Multiple Sclerosis**

**Disease Modifying Therapy**

**Immune Modulation Treatments**

- **Interferon β-1a (Avonex, Rebif)** and **Interferon β-1b (Betaseron)**
  - Inhibits T cell proliferation with antigen presentation by B cells
  - Reduces CXCR3 expression on CD4+ and CD8+ lymphocytes

- **Glutiramir Acetate (Copaxone)**
  - Binds MHC molecules on APC’s to compete with myelin antigens
  - Induces Suppressor T-Cells

- **Dimethyl Fumarate ("BG-12", oral)**
  - Glutathione depletion and induction of anti-inflammatory stress protein HO-1
  - May have anti-oxidative cytoprotective effects

**Multiple Sclerosis: Immune Depleting Treatments**

- **Natalizumab (Tysabri)**
  - Monoclonal antibody to the cell adhesion molecule α4 Integrin
  - Reduces the ability of lymphocytes to attach to and travel through endothelial cells lining the gut and the BBB
  - (Associated with PML)

- **Fingolimod (Gilenya, oral)**
  - Sphingosine 1-phosphate receptor modulator
  - Reduces lymphocyte egress from lymphoid tissues into the CNS

- **Teriflunomide (Aubagio, oral)**
  - Inhibits dihydroorotate dehydrogenase
  - Reduced pyrimidine synthesis important for rapidly dividing B and T-cells

- **Ocrelizumab**
  - Anti-CD-20 monoclonal antibody (similar to Rituximab)

- **Alemtuzumab (Campath)**
  - Anti CD52 monoclonal antibody
  - Depletes B and T-cell lymphocytes, macrophages, NK cells, monocytes and some granulocytes

- **Daclizumab (Zenapax)**
  - Monoclonal antibody blocking IL-2R receptors on lymphocytes
  - Reduces lymphocyte proliferation and differentiation
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Neurology Review

Definitions

- Seizure: Transient neurological dysfunction secondary to abnormal synchronous electrical discharges arising from the cortex.
- Epilepsy: A chronic condition characterized by recurrent, unprovoked seizures.
- Provoked Seizures: Due to acute irritation/disruption of the cortex. Provoked seizures do not necessarily lead to epilepsy (recurrent unprovoked seizures).
## Neurology Review

<table>
<thead>
<tr>
<th>Focal Seizures</th>
<th>Generalized Seizures</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Simple Partial Seizures</td>
<td>• Absence Seizures</td>
</tr>
<tr>
<td>• Complex Partial Seizures</td>
<td>• Atonic Seizures</td>
</tr>
<tr>
<td>• Secondarily Generalized Convulsive Seizures</td>
<td>• Tonic Seizures</td>
</tr>
<tr>
<td></td>
<td>• Clonic Seizures</td>
</tr>
<tr>
<td></td>
<td>• Myoclonic Seizures</td>
</tr>
<tr>
<td></td>
<td>• Primary Generalized Convulsive Seizures</td>
</tr>
</tbody>
</table>
Neurology Review

Clinical features of Focal Seizures

- Simple Partial Seizures
  - “Aura”

- Complex Partial Seizures
  - Any degree of impaired consciousness
  - Implies bilateral cortical hemisphere involvement

- Secondarily Generalized Convulsive Seizures
  - May begin with simple or complex partial seizure
  - May also rapidly secondarily generalize
<table>
<thead>
<tr>
<th>Clinical feature</th>
<th>Absence Sz</th>
<th>Complex Partial Sz</th>
</tr>
</thead>
<tbody>
<tr>
<td>Duration:</td>
<td>few to 15 sec’s</td>
<td>20 sec’s to minutes</td>
</tr>
<tr>
<td>Frequency:</td>
<td>Hundreds/Day</td>
<td>Intervals: days to wks</td>
</tr>
<tr>
<td>Aura:</td>
<td>Never</td>
<td>Possibly</td>
</tr>
<tr>
<td>Post-Ictal:</td>
<td>Never</td>
<td>Usually</td>
</tr>
<tr>
<td>Age of Onset:</td>
<td>Early school age</td>
<td>Any age</td>
</tr>
<tr>
<td>EEG:</td>
<td>3 Hz Generalized Spike/Slow Wave</td>
<td>Normal or focal</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Spikes or Background Changes</td>
</tr>
</tbody>
</table>
Neurology Review

Causes of Seizures

- Toxic/Metabolic: $\uparrow$ or $\downarrow$ Na$^+$, $\downarrow$ Ca$^{++}$, $\downarrow$ Glucose, uremia, liver failure, IEM’s, ETOh, drugs, medications, etc
- Neoplastic/Paraneoplastic: primary or metastatic brain tumors, limbic encephalitis
- Vascular: stroke, hemorrhage
- Structural: Developmental brain malformations
- Infection/Post-Infectious: Meningoencephalitis, abscess, ADEM
- Trauma: Early vs late Post-traumatic seizures
- Paroxysmal: Epilepsy
- Degenerative Disorders: NCL, lysosomal storage diseases, Neurodegenerative diseases (Alzheimers, Huntington’s, etc.)
- Psych: Non-epileptic seizures
Neurology Review

Febrile Seizures

- Seizures in setting of Fever, no evidence of CNS infection.
- Age 6 mo’s to 5 yrs.
- 2%-4% of Population
- Complex Febrile Seizures vs Simple Febrile Seizures
  - >15 minutes, Focal features, 2 or more within 24 hrs
- Risk of Recurrent Febrile Sz’s
  - Low temperature, young age (<12 months), Family Hx of Febrile Sz’s
- Risk of Epilepsy
  - Developmental delay, Complex Febrile Sz, Family Hx of Epilepsy
- Treatment
  - Risks outweigh benefits
Neurology Review

**Status Epilepticus**

- Unremitting or back-to-back Sz for >30 minutes
- Convulsive or Non-Convulsive Status
- Start Rx at 5 to 10 minutes
  - Benzodiazepine Therapy (Lorazepam or Diazepam)
  - AED Therapy (Phenytoin or Phenobarbital)
- Outcome depends on etiology
  - Remote symptomatic and neurodegenerative etiologies worse
  - Acute Symptomatic needs to treat the underlying cause and the seizure
  - Good prognosis for idiopathic etiology
Neurology Review

Sleep Disorders
Parasomnias

Nightmares vs Night Terrors
- Nightmares occur during REM sleep. Pt’s remember their dreams
- Night Terrors occur in younger children in stage III and IV sleep. Children have no recollection of the event.

Sleep Walking
- Stage III and IV sleep. Automatic motor activities. Risk of injury

REM Behavioral Disorder
- May be an early sign of Parkinson’s Disease, Lewy Body Dementia or Multiple Systems Atrophy
Dyssomnias

- **Restless Legs Syndrome**
  - Urge to move/stretch limbs. Impairs sleep onset. Excessive daytime somnolence (EDS). Responds to DA agonist medications (eg. Ropirinole)

- **Obstructive Sleep Apnea**
  - Common cause of EDS. Upper airway obstruction causes subclinical arousals. T and A or CPAP to Rx.

- **Narcolepsy**
  - Tetrad of EDS, Cataplexy, Hypnopompic hallucinations, Sleep paralysis
  - Multiple Sleep Latency Test (REM-onset sleep, short sleep latency)
  - HLA-DR2 and HLA-DQw1 association
  - Low CSF levels of Orexin (hypocretin)
  - Rx with Modafinil, Stimulants, Sodium Oxybate, TCA’s, scheduled naps

- **Idiopathic Hypersomnolence**
Neurology Review

Sleep Disorders

- Insomnia
  - Short-term/intermittent
    - Jet lag, shift work, acute illness, stimulant/caffeine, withdrawal of sedative/hypnotics, environment
  - Chronic
    - Dyssomnias, Medication/drug/EtOH, Mood/Anxiety disorders, Delayed sleep phase syndrome (teens and young adults)
    - Medical diseases: CHF, COPD, Asthma, GERD, arthritis/pain, fibromyalgia
    - Neurological diseases: Stroke, Neurodegenerative disorders, TBI, Neuromuscular disorders, Headache syndromes, ADHD
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Neurology Review

Stroke

• Thrombotic Stroke
  • Thrombosis of large vessels, often at points of bifurcation. Stuttering onset. Often occurs in sleep.

• Embolic Stroke
  • Occlusion of distal cortical vessels. Abrupt onset with maximal deficits at onset. Emboli are usually atherosclerotic plaques or come from cardiac sources.

• Hemorrhagic Stroke
  • Stroke due to cerebral hemorrhage of sudden onset. HTN infarction (Putamen, Thalamus, Pons, Cerebellum), AVM, Aneurysm, Amyloid Angiopathy

• Lacunar Infarction
  • Infarction of deep penetrating arteries (often secondary to lipohyalinosis due to hypertension and endothelial cell wall inflammation). Internal Capsule, Pons, Thalamus. Pure motor or pure sensory symptoms common
Neurology Review

Stroke Syndromes

- **ACA**
  - Leg > Arm weakness

- **MCA**
  - Arm = Leg weakness. Visual field cut. Higher cortical deficits (aphasia or hemi-neglect)

- **Ophthalmic Artery**
  - Amarosis fugax

- **PCA**
  - Visual field cut

- **Vertebro-Basilar**
  - Brain stem findings (vertigo, ataxia, dysphagia) with crossed long-tract signs (hemiparesis and/or hemisensory loss)

- **Lacunar**
  - Pure Motor (post limb internal capsule (IC), basis pontis or corona radiata).
  - Pure Sensory (thalamus).
  - Sensorymotor (post limb of IC and thalamus).
  - Clumsy Hand/Dysarthria (post limb of IC /corona radiata junction).
  - Leg Paresis/Ataxia (ant limb of IC or corona radiata).
Acute Anticoagulation (Heparin)
- Definite: Atrial Fibrillation and Arterial Dissection
- ? Progressive vertebrobasilar stroke, stroke-in-evolution, crescendo TIA’s
- Followed by Coumadin or LMW Heparin

rTPA
- 4 ½ window from onset. Contraindications...

Anti-Platelet
- Aspirin, Clopidogrel, Dipyridamol/ASA (Aggrenox)

Carotid Endarterectomy
- Mild stroke with ipsilateral severe carotid stenosis (70-99%)
- ? With moderate stenosis (50-69%)
- No benefit with mild stenosis (<50%)
Neurology Review
Subarachnoid Hemorrhage

- **Etiology:**
  - Ruptured congenital cerebral aneurysm (near circle of willis)
  - Other: AVM, mycotic aneurysm, trauma, intracerebral hemorrhage

- **Outcome:** Mortality 50% within 2 weeks. 30% survivors require lifelong care

- **Presenting Symptoms**
  - Thunderclap headache, nuchal rigidity, altered MS
  - III nerve palsy from p.comm aneurysm

- **Complications**
  - Cerebral vasoconstriction, SIADH, rebleeding, hydrocephalus, cardiac arrhythmias

- **Diagnosis**
  - CT scan. LP if nl CT (Tubes 1 and 4, xanthochromia)
  - Cerebral Angiography (MRA, CT angiogram, conventional angiogram)
Hypertensive Encephalopathy

- **Definition**
  - Diffuse cerebral dysfunction associated with sudden or severe elevations of systemic blood pressure

- **Signs/Symptoms**
  - Papilledema, Headache, Altered MS, Seizures, Focal neurological deficits

- **Treatment**
  - Avoid abrupt lowering of systemic blood pressure (use labetolol or nitroprusside drips)
  - Resolution of symptoms with Rx of blood pressure is diagnostic
Syncope

- Caused by reduced Cerebral Perfusion Pressure
- Vaso-Vagal syncope most common etiology
  - Brief LOC with rapid return to consciousness (unlike seizures)
  - Injuries are rare
  - Pre-syncopal symptoms common (light headedness, fading out of vision)
  - May be reflexive (site of blood or during micturation or defecation)
- Syncope with exertion or long-lasting syncope needs cardiac evaluation for structural or electrical conduction disorders
  - Prolonged QT syndrome
  - IHSS
  - Intermittent ventricular arrhythmias
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Secondary Headaches

- Intracranial Pain-Sensitive Structures
  - Dura, venous sinuses, proximal arteries, bones, sinuses, eyes, etc

- Pseudotumor Cerebri
  - Progressive postural HA, Diplopia (VI n. palsy), Papilledema
  - Idiopathic intracranial HTN, obesity, females>males, OP > 25 cm H2O
  - Rx with Acetazolamide
  - Need to exclude sagittal sinus thrombosis

- Temporal Arteritis
  - Inflammation of large intra and extracranial vessels in older adults
  - Headaches, jaw claudication, systemic sx’s, vision loss
  - May be part of Polymyalgia Rheumatica
  - Inflammation of Temporal Artery on biopsy
  - Rx with steroids
Neurology Review
Primary Headache Disorders

• Migraine Headaches
  • Common vs Classical vs Complicated Migraine
  • Often runs in families
  • Clinical features and triggers
  • Acute Symptomatic Rx: NSAIDs, ASA/Caffeine, Ergotamines, Triptans
  • Prophylactic Rx: TCA’s, AED’s, Ca-channel blockers, Beta-blockers, etc

• Cluster Headaches
  • Trigemino-vascular headache. Severe retro-orbital pain. Periodic attacks
  • Male predominance
  • Acute Rx with Oxygen, Indomethacin, Triptans, etc. Lithium as a preventative med.

• Chronic Tension-Type Headache
  • Chronic daily headache. Mild to moderate. Band-like non-throbbing pain
  • May have mood or anxiety disorder
  • Needs Preventative medication. Address Medication Overuse headache
• Trigeminal Neuralgia
  • Stabbing Facial Pain
  • Rx with AED’s (carbamazepine 1st-line), TCA’s, Duloxetine
  • Surgical decompression vs ablation

• Complex regional Pain Syndrome
  • Type I (Reflex Sympathetic Dystrophy):
    • Severe Pain. Vasomotor changes, sudomotor changes, bone demineralization
    • Late atrophy, dystrophic skin and nail changes
  • Type II (Causalgia)

• Neuropathic Pain
  • Caused by spontaneous firing of small fibre sensory nerves
  • Treat with TCA’s or AED’s, Duloxetine
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Head Trauma

Epidural Hematoma
- Associated with fx of temporal bone and tearing of middle meningeal artery
- Convex appearance on CT limited by cranial sutures
- LOC (initial head injury), followed by “Lucid Interval”, followed by LOC with uncal herniation

Subdural Hematoma
- Tears in subdural bridging veins. Affects older people (brain atrophy)
- May have delayed symptomatic presentation
- Crescent shape on CT not limited by cranial sutures
- Can evolve into a subdural hygroma (CSF density) over time
Neurology Review

Head Trauma

- **Subarachnoid Hemorrhage**
  - May be seen with other types of hemorrhage with head trauma
  - Complications include: Hydrocephalus, SIADH, cerebral vasospasm

- **Intraparenchymal Hemorrhage**
  - Due to damage to deep penetrating cerebral vessels
  - Cerebral contusions arise from translational forces and are commonly seen at the frontal, temporal or occipital poles of the cortex (coup countercoup injury)

- **Basilar Skull Fracture**
  - CSF otorrhea/rhinorrhea (glucose will be high on sample)
  - Hemotympanum
  - Racoon eyes
  - Battle sign
Neurology Review
Head Trauma

• Concussion
  A concussion (or mild traumatic brain injury) can be defined as a complex pathophysiologic process affecting the brain and induced by either direct or indirect traumatic biomechanical forces applied to the head in the setting of typically normal neuroimaging studies. Symptoms are a constellation of physical, cognitive, emotional, and/or sleep-related disturbances and may or may not include an initial loss of consciousness. Duration of symptoms is highly variable generally lasting from minutes to days or weeks and occasionally even longer in some cases.

• Post-Concussion Syndrome
  Lasts weeks to months
  Symptoms may include: headaches, poor attention and concentration, fatigability, memory problems, anxiety/mood changes, sleep disorders

• Second Impact Syndrome
  Impact before concussion resolved.
  Malignant High ICP due to loss of autoregulation leads to herniation
## Graded Return to Play Protocol

<table>
<thead>
<tr>
<th>Rehabilitation stage</th>
<th>Functional exercise at each rehabilitation stage</th>
<th>Objective</th>
</tr>
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<tbody>
<tr>
<td>1. No activity</td>
<td>Complete physical and cognitive rest</td>
<td>Recovery</td>
</tr>
<tr>
<td>2. Light aerobic exercise</td>
<td>Walking, swimming, stationary bike with heart rate less than 70% maximum</td>
<td>Increased heart rate</td>
</tr>
<tr>
<td>3. Sport-specific exercise</td>
<td>Running drills without contact</td>
<td>Add movement</td>
</tr>
<tr>
<td>4. Noncontact training</td>
<td>More complex drills without contact</td>
<td>Exercise, coordination, and cognitive load</td>
</tr>
<tr>
<td>5. Full contact</td>
<td>Normal training</td>
<td>Restore confidence</td>
</tr>
<tr>
<td>6. Return to play</td>
<td>Game play</td>
<td></td>
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*Source: Dr. Jordan*
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Neuropathology (mix and match)

A. Lewy Bodies
B. Pick Bodies
C. Cowdry Type A bodies
D. Microglial Nodules
E. Negri bodies
F. Spongiform changes
G. Senile Plaques
H. Alzheimers Type II Astrocytes
I. Balloon Cells
J. Neurofibrillary Tangles
Neuropathology (mix and match)

A. Lewy Bodies 1) Herpes Virus
B. Pick Bodies 2) Parkinson’s Disease
C. Cowdry Type A bodies 3) Rabies
D. Microglial Nodules 4) Prion Disease
E. Negri bodies 5) Alzheimer's Disease
F. Spongiform changes 6) HIV
G. Senile Plaques 7) Hepatic Encephalopathy
H. Alzheimers Type II Astrocytes 8) Cortical Dysplasia
I. Balloon Cells 9) Pick’s Disease
J. Neurofibrillary Tangles

Neurology Review

Dementia

Definition:
- A global impairment of cognitive function without impaired alertness
- Impairs normal social and occupational functioning
- Subacute to chronic onset and often irreversible

Mild Cognitive Impairment
- Deficits in memory beyond those expected for age that do not significantly impact daily functioning (remembering names of people or misplacing items)
- These deficits tend to remain stable over time (unlike AD) and are apparent to the individual
- Reported memory problems by a knowledgeable informant, poor performance on standardized cognitive testing, inability to perform some activities of daily living (such as correct hygiene/grooming) may suggest progression to AD
Dementia

Differential Diagnosis:

- **Metabolic:**
  - Thiamine deficiency (Wernicke’s Encephalopathy: Korsakoff’s psychosis, ophthalmoplegia, ataxia)
  - B12 deficiency: Incr Homocysteine and MMA, megaloblastic anemia, subacute combined degeneration of the cord (dorsal columns and descending corticospinal tract), delerium/dementia
  - Chronic EtOH abuse
  - Hepatic or renal failure
  - Hypothyroidism or Cushing’s syndrome

- **Vascular:** Multi-infarct dementia

- **Infection:** Syphilis, AIDS, Creutzfeld-Jakob ds (triphasic waves on EEG)

- **Structural:** Normal Pressure Hydrocephalus

- **Degenerative Disorders:** Alzheimer’s ds, Parkinson’s ds, Dementia with Lewy Bodies, Picks ds, Huntington’s Chorea

- **Pseudodementia:** Depression
Neurology Review

Cortical Dementias

- Alzheimer’s Disease
- Picks Disease: Fronto-temporal dementia with Pick Bodies
- NPH: HCP without increased ICP, Dementia, Gait Apraxia, Urinary Incontinence (Wet, Wacky, Wobbly). Partially reversible with VP shunt
Neurology Review

Alzheimer’s Disease

- **Prevalence**
  - Causes 50% of Dementia in older pts. 20% of 80 year-olds have AD
  - Seen in 100% of Down syndrome patients over 40 years of age

- **Clinical Stage**
  - Early: Mild forgetfulness, misplace items, personality changes
  - Later: Disorientation, unable to work, worsening language and memory, severe personality changes with anger/agitation, delusions
  - End-stage: Severe cog impairment, incontinence, risk for aspiration, extrapyamidal signs, vegetative

- **Pathology**
  - NF Tangles, Senile Plaques, Brain Atrophy

- **Treatment**
  - Cholinesterase inhibitors (Aricept, Cognex, Exelon)
  - Glutamate Antagonists (Namenda)
Neurology Review

Subcortical Dementias

- Parkinson’s Disease
- Dementia with Lewy Bodies
  - Fluctuating memory/cognitive problems with extra-pyramidal symptoms. Diffuse Lewy Bodies seen throughout cortex and brainstem
- Shy-Drager Syndrome (Multiple system atrophy)
  - Bradykinesia and rigidity without tremor
  - Orthostatic hypotension and/or cerebellar ataxia may be present
  - Poor response to levodopa/carbidopa
- Progressive Supranuclear Palsy
  - Falls and postural instability
  - Impaired vertical gaze
  - Poor response to levodopa/carbidopa
- Huntington’s Disease
  - AD triplet repeat (CAG) on chromosome 4
  - Disinhibition followed by dementia. Choreoathetoid movements
  - Caudate heads atrophic on imaging
Parkinson’s Disease

Clinical Symptoms
- Tetrad of Rigidity, Bradykinesia, Resting Tremor, Postural Instability
- Sub-Cortical Dementia

Pathology
- Diffuse Gliosis with Lewy Bodies
- Degeneration of DA-containing Neurons within the Substantia Nigra leads to depletion of DA within the Striatum

Treatment
- Sinemet (Levodopa + Carbidopa (inhibits DOPA decarboxylase)
  - Dopamine itself cannot cross the BBB. Carbidopa is a dopamine decarboxylase inhibitor
  - Carbidopa allows for more absorption of levodopa from the gut
  - Carbidopa does not cross BBB so Levodopa can be decarboxylated within the CNS to allow increased concentrations of Dopamine to reach the CNS
- Dopamine Agonists (Pramipexole, Ropinirole, Bromocriptine, Pergolide) with or without Catechol-O-methyltransferase inhibitors (entacapone)
  - May cause impulse control disorder in some patients with PD
- Anticholinergics (Benztropine, Trihexiphenidyl)
- Antiviral (Amantidine)
- Deep Brain Stimulation/Pallidotomy
Neurology Review

Categories

- CNS Infections
- Auto-Immune Disorders
- Epilepsy and Sleep
- Vascular Diseases
- Headache and Pain Syndromes
- Trauma
- Degenerative Disorders/Dementia
- Altered Mental Status
- Movement Disorders
- Structural Disorders
- Toxic/Metabolic Disorders
- Psychiatric
- Neuromuscular Disorders
- Localization/Anatomy
Neurology Review

Altered Mental Status

- **Localization**
  - Brainstem (ascending RAS) or Bilateral Cortical Hemispheres

- **Depressed LOC vs Delerium**
  - Delerium has normal level of alertness but altered content of consciousness. Includes agitation, disorientation, poor concentration, hallucinations, etc. Same as acute psychosis. Consider drug screen

- **Coma**
  - Unarousable Unresponsiveness. GCS scale 3-15 (D.Dx. next slide)

- **Persistent Vegetative State**
  - EEG with wake and sleep states. Spont eye opening. Grunts/groans
  - Minimally Conscious State: some awareness of self or environment

- **Locked in Syndrome**
  - Normal consciousness. Corticospinal and corticobulbar tracts affected
  - Some vertical eye movements and blinking preserved
Neurology Review

Causes of Coma

- **Toxic/Metabolic**
  - Carbon Monoxide, chemotherapy, radiation, EtOH, sedative/hypnotic medications and drugs heavy metals, hyper/hypoglycemia, DKA, hyponatremia, IEM’s, renal failure, liver failure, hypercapnea, hypoxia, porphyria, hypothyroidism

- **Structural**
  - Herniation syndromes, hydrocephalus, cerebral edema

- **Infectious/Post-Infectious /Autoimmune**
  - Meningoencephalitis, brain abscess, sepsis, ADEM, CNS vasculitis, SLE

- **Neoplastic/Paraneoplastic**
  - Primary or metastatic brain tumors, paraneoplastic limbic encephalitis

- **Paroxysmal**
  - Seizures, non-convulsive status epilepticus, post-ictal state

- **Trauma**
  - Concussion, intracranial hemorrhage (epidural, subdural, subarachnoid, intraparenchymal)

- **Vascular**
  - Ischemic or hemorrhagic stroke, SAH, venous thrombosis, hypoxic-ischemia, hypertensive encephalopathy, cerebral hypoperfusion

- **Degenerative/Genetic**
  - Neurodegenerative disorders

- **Psych**
  - Conversion, catatonic schizophrenia
Brain Death

- **Establish irreversible and proximate cause of brain death**
  - Exclude effects of CNS depressant drugs, paralysis of neuromuscular blocking agents, severe electrolyte or glucose disturbances, hypothermia

- **Achieve normal systolic blood pressure**

- **Demonstrate lack of cortical function**
  - No evidence of responsiveness. No eye opening or movement in response to noxious stimulus.

- **Demonstrate lack of brainstem reflexes**
  - Absence of pupillary light responses
  - Absence of ocular
  - Absence of corneal reflex.
  - Absence of facial muscle movement to noxious stimulus.
  - Absence of the pharyngeal and tracheal reflexes

- **Apnea Test**
  - Pre-oxygenate
  - No spontaneous respiration with $\text{PCO}_2 > 60$ mm Hg (or $>20$ mm Hg above baseline)

- **Ancillary Testing**
  - Only if cannot complete physical exam
  - Cerebral angiography or Cerebral scintigraphy (technetium Tc 99m hexametazime)-No Intracranial perfusion
  - EEG-Isoelectric

- **Repeat Examination**
  - 2 exams 24 hours apart with 2 apnea tests for term to 30 days old
  - 12 hours apart for older children up to 18 yrs old
Neurology Review

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Neurology Review

Movement Disorders

- Involuntary Movements Described by their Features
  - There may be overlapping clinical features

- Hyperkinetic MD’s
  - Tremor, Chorea, Athetosis, Tics, Myoclonus, HemiBallismus

- Hypokinetic MD’s
  - Rigidity, Dystonia, Parkinsonism

- Tremor
  - Postural Tremor (Physiological, Essential, Hyperthyroid)
  - Intention Tremor (Cerebellar)
  - Resting (Parkinson’s)

- Most Movement Disorders Localize to The Basal Ganglia
  - Extrapyramidal System
Neurology Review

Tourette Syndrome

- **Tics**
  - Rapid, stereotyped motor movements or vocalizations
  - Usually Begin in Childhood
  - Corprolalia is rare

- **Chronic Tic Disorders**
  - Chronic Motor Tic Disorder of Childhood
  - Chronic Vocal Tic Disorder of Childhood
  - Tourette Syndrome

- **Frequent Co-Morbid Disorders**
  - ADHD
  - Anxiety Disorders
  - OCD
Neurology Review

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Neurology Review
Structural Disorders

- Herniation Syndromes
- Hydrocephalus
- Spinal Cord Disease
- Neurocutaneous Disorders
Regions of Brain Herniation
Neurology Review

Hydrocephalus

- Communicating HCP
  - Impairment of reabsorption at the arachnoid granulations
  - May be a late finding in bacterial meningitis or subarachnoid hemorrhage

- Non-Communicating HCP
  - Obstruction most commonly at The Aqueduct of Sylvius (pineal gland tumors) or IVth Ventrical (foramen of Luschka and Magendie)

- Signs/Symptoms of Hydrocephalus
  - Progressive postural headache, VI nerve palsy (diplopia), Papilledema

- Treatment
  - CSF diversion through a ventricular shunt (VP most common)
Neurology Review

Spinal Cord Disease

- **Myelomeningocele**
  - Neural Tube Defect (normal closes at day 24)
  - Folic Acid Deficiency, Genetic etiologies
  - Open defect at birth. Closure to prevent meningitis
  - Weak legs, neurogenic bladder, constipation/incontinence
  - Latex Allergy
  - Chiari II Malformation

- **Occult Spinal Dysraphism**
  - Overlying skin abnormality (tuft of hair, dimple, hemangioma, lipoma)
  - May be associated with a tethered spinal cord

- **Spina Bifida Occulta**
  - Midline defect of the posterior vertebral bodies (incidental, 10% of population)

- **Syringomyelia**
  - Dilation of central canal of cord
  - Loss of pain/temperature sensation (anterior commissure)
  - Chiari I, Trauma, Tumors are etiologies
Neurology Review

Spinal Cord Disease

**Acute Myelopathy**
- Combination of flaccid paralysis, dermatomal sensory level (pain/temp and/or posterior column) and autonomic dysfunction (Horner’s, bowel/bladder incontinence)
- Needs emergent neuroimaging with MRI (mass lesion until proven otherwise)

**Cauda Equina Syndrome**
- Compression of the lumbar/sacral nerve roots below level of conus medullaris
- May be caused by tumor, spinal stenosis, degenerative disc disease
- Sx: LBP, sciatica, urine retention/incontinence, saddle anesthesia, sexual dysfn.

**Spinal Stenosis**
- Narrowing of the spinal canal caused by protruding discs, bone spurs, osteoarthritis, or thickening of the ligamentum flavum
- Sx: Back pain when standing, neurogenic claudication, radicular pain, weakness, incontinence, or cauda equina syndrome

**Spondylosis**
- Osteoarthritis of the vertebral body joints or degenerative changes of the vertebral discs resulting in nerve root compression
- Spurling’s test (pain in ipsilateral shoulder when pressing down on rotated head)
Neurology Review

Neurocutaneous Disorders

???
Neurocutaneous Disorders

???????
Neurology Review

Neurocutaneous Disorders
Neurology Review

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Neurology Review

Electrolyte Abnormalities

- Sodium
  - ↓Na⁺ may be caused by SAH, meningitis, head trauma, brain tumors.
  - ↓Na⁺ may cause seizures or encephalopathy
    - **SIADH** (↓serum Na⁺, ↓serum Osm, ↑urine Osm, ↑ urine Na⁺ excretion, ↓UOP, normovolemia) Rx with fluid restriction or 3% saline if symptomatic.
    - **Cerebral Salt Wasting** (↓serum Na⁺, ↓serum Osm, ↑urine excretion of Na⁺, ↑UOP, hypovolemia). Rx with fluid and NaCl replacement
  - Rapid correction of hyponatremia may lead to **Central Pontine Myelinolysis**

- Calcium
  - ↓Ca⁺⁺ can lead to delerium, seizures, and neuronal hyperexcitability (carpopedal spasm, Chvostek’s sign)
Neurology Review

Glucose Abnormalities

- Hyperglycemia
  - DKA
    - Polyuria, polydipsia, dehydration, and metabolic acidosis lead to AMS, focal deficits, or coma
    - Cerebral edema
  - Hyperosmolar Nonketotic Hyperglycemia
    - Dehydration, significantly ↑ serum glucose and Osm, Sz’s, and coma

- Hypoglycemia
  - Endogenous (infants)
  - Secondary to medications (insulin), alcoholism, etc
  - Initial agitation, tachycardia, sweating leading to coma, seizures, posturing etc
  - Rx with D25W 2-3 cc/kg
Neurology Review

Ethanol

- Acute Intoxication
  - Pancerebellar symptoms and encephalopathy
  - ↑ serum Osmolality

- Seizures
  - Due to EtOH withdrawal
  - Prophylaxis with Benzo’s may be helpful

- Thiamine Deficiency
  - Wernicke’s Encephalopathy (Ophthalmoplegia, confusion and ataxia)
  - Rx with 100 mg Thiamine before or concurrent with Dextrose

- Delerium Tremens
  - Delerium, tremor, sweating, tachycardia
  - Rx with Benzodiazepines, manage hypoglycemia, give Thiamine
Neurology Review
Medication/Drugs

• **Sedative/Hypnotics**
  - Includes Benzodiazepines, Opiates, Barbiturates and others
  - Intoxication: Depressed MS to coma, Respiratory Depression, Small but Reactive pupils. Rx: Nalaxone (opiates), Flumazenil (Benzo’s)
  - Withdrawal: Delerium, Agitation, Insomnia, tachycardia, HTN, Dilated but reactive Pupils, Seizures. Rx: Benzodiazepines

• **Sympathomimetics**
  - Includes Cocaine, Amphetamines, PCP, Stimulants, etc
  - Intoxication: Delerium, Agitation, Insomnia, tachycardia, HTN, Dilated but reactive Pupils, Seizures. Rx: Haloperidol, Benzodiazepines

• **Anticholinergics**
  - Includes: Anticholinergics, TCA’s, Antipsychotics, Antihistamines
  - Delerium, Dry skin, Urine Retention, Tachycardia, Fever, Flushing. Large, Dilated pupils. Rx: physostigmine

• **Organophosphate Poisoning**
  - Diaphoresis, Salivation, Lacrimation, Bradycardia, Small Reactive Pupils
Neurology Review

**Thyroid**

- **Hypothyroid (myxedema)**
  - Confusion, Dementia, delayed relaxation of DTR’s
  - Can progress to Seizures and Coma
  - Cretinism in Congenital Hypothyroidism

- **Hyperthyroid**
  - Agitation to acute confusional state
  - Seizures
  - Heat intolerance, hair loss, dry skin, weight loss, tachycardia
  - Brisk DTR’s, Postural Tremor
Neurology Review

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Neurology Review

Psychiatry

- **Serotonin Syndrome**
  - AMS, ↑BP, ↑HR, sweating, flushing, fever, n/v, ↑DTR’s, myoclonus

- **Tardive Dyskinesia**
  - Seen in elderly female schizophrenics with long-term neuroleptic use
  - Oral-buccal dyskinesias persist after offending medication withdrawn
  - Difficult to treat. ? Benefit of prophylactic anticholinergics with neuroleptics

- **Drug-Induced Parkinsonism**
  - Caused by too much DA blockade. Responds to lowering/removing drug

- **Drug-Induced Dystonias**
  - Oculogyric Crisis, Torticollis, etc. Responds to IV Diphenhydramine

- **Neuroleptic Malignant Syndrome**
  - Rare life-threatening idiosyncratic side effect of DA-blocking drugs
  - High Fever, Muscle Breakdown, Myoglobinmuria
  - Generous Hydration, Alkalinize Urine, Dantrolene
Neurology Review

Psychiatry

- Malingering vs Conversion Disorders
  - Non-Epileptic Seizures
  - Unable to Walk
  - Psychogenic Blindness

- Munchausen Syndrome
  - A form of Malingering. Intentional production of symptoms to meet some psychological need
  - Examples include injection one’s self with feces to cause fevers, surreptitiously taking insulin, applying mydriatic eye drops into one eye
  - May result in unnecessary surgeries and medical interventions
Neurology Review

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- Localization/Anatomy
Neurology Review

Anterior Horn Cell Disorders

- Clinical features
  - Weakness, atrophy, and fasiculations

- Polio
  - Initial Encephalitis, followed by asymmetric limb weakness/atrophy
  - Post-Polio Syndrome

- Spinal Muscular Atrophy
  - Type I, II, III
  - AR, SMN-1 gene exon 7 and 8 deletions

- Amyotrophic Lateral Sclerosis
  - Lou Gherig’s Disease
  - Anterior Horn Cell along with Corticospinal tract degeneration
  - Supportive Rx only
Neurology Review

Neuropathies

- General Features
  - Often length-dependent weakness, sensory loss (polyneuropathies)
  - Early loss of DTR’s
  - Large fibres (vibration and position sense), Small fibres (pain/temperature)

- Guillain Barre Syndrome
  - Albuminocytological disociation
  - AIDP conduction block, demyelination on NCV’s
  - Rx with Plasmapharesis or IVIg
  - CIDP: Rx with steroids

- Charcot-Marie-Tooth Disease
  - Hereditary Motor and Sensory Neuropathy
  - Symmetric distal weakness and large fibre sensory loss
  - CMT1A caused by duplications PMP-22 gene (autosomal dominant)
Neurology Review

Neuropathies

- **Diabetic Peripheral Neuropathy**
  - Small fibre painful polyneuropathy or focal neuropathy
  - Rx with AED’s, TCA’s, SNRI’s

- **Critical Illness Polyneuropathy**
  - Seen with sepsis, multi-organ failure, respiratory failure
  - Difficult to wean patient off ventilator
  - NCV’s show sensory and motor axonal neuropathy
  - Recovery may take weeks to months

- **Toxic Neuropathies**
  - **Lead**
    - Microcytic anemia with basophilic stippling. Distal (m/s) axonal
  - **B-6 Deficiency**
    - Paresthesias, decreased pain/temp. (s>m) axonal
  - **Acute Intermittent Porphyria**
    - Altered MS, GI pain, diminished DTR’s. (m>s) axonal
Neurology Review
Focal Traumatic Neuropathies

• Median Neuropathy at the Carpal Tunnel
  • Weakness of thumb abduction Abductor Pollicis Brevis (median, C8, T1) and thumb opposition to palm Opponens Pollicis (median, C8, T1)
  • Thenar atrophy
  • Pain and loss of sensation to palmar surface including the thumb, thenar eminence, index finger, middle finger and medial aspect of ring finger
  • Splinting of wrists and neuropathic pain meds is first line of RX if weakness absent
  • Carpal tunnel release procedure if weakness present
Focal Traumatic Neuropathies

Ulnar Neuropathy at the Elbow

- **Weak** pinky abduction *Abductor Digiti Minimi* (ulnar C8, T1) and flexion of DIP joints *Flexor Digitorum Profundus III and IV* (ulnar C7, C8)
- Hypothenar atrophy
- Sensory loss on palmar side of pinky and lateral half of ring finger

Ulnar Neuropathy at Guyon’s Canal

- **Weak** pinky abduction *Abductor Digiti Minimi* (ulnar C8, T1), Hypothenar atrophy and sensory loss in ulnar hand distribution
- **Spared** flexion of DIP joints *Flexor Digitorum Profundus III and IV* (ulnar C7, C8)
Neurology Review
Focal Traumatic Neuropathies

Radial Neuropathy at the Upper Arm
- **Weak** wrist extension Extensor Carpi Radialis (C5,C6), elbow flexion Brachioradialis (C5,C6), wrist extension and finger extensors Extensor Digitorum (C7,C8)
- **Weak** elbow extension if lesion is in the axilla Triceps (C6,C7,C8)
Upper Brachial Plexus Stretch Injury (Erb’s Palsy)

- Weak C5, C6, >C7 nerve root innervated muscles
  - Deltoid (Axillary n. C5>C6)
  - Supraspinatus and Infraspinatus (Suprascapular n. C5>C6)
  - Brachioradialis (Radial n. C5<C6)
  - Biceps (Musculocutaneous n. C5, C6)
  - Wrist Extension (Radial n. C6)
- Absent Biceps DTR (C5, C6)
Neurology Review

Foot Drop

**Sciatic neuropathy**
- **Weak** Sciatic Nerve: Hamstrings (sciatic L5,S1,S2)
- **Weak** Peroneal Nerve: Weak Dorsiflexion: Tibialis Anterior (L4,L5) and Foot Eversion: Peroneus Longus and Brevus (L5,S1)
- **Weak** Tibial Nerve: Weak Foot Inversion: Tibialis Posterior (L5)

**Peroneal Neuropathy**
- **Weak** Peroneal Nerve: Weak Dorsiflexion: Tibialis Anterior (L4,L5) and Foot Eversion: Peroneus Longus and Brevus (L5,S1)
- **Spared** Foot Inversion: Tibialis Posterior (tibial L5)

**L5 Radiculopathy**
- **Weak** Dorsiflexion: Tibialis Ant. (L4,L5, peroneal nerve)
- **Weak** Foot Eversion: Peroneus Longus and Brevus (L5,S1, peroneal nerve)
- **Weak** Foot Inversion: Tibialis Posterior (L5, tibial nerve)
- **Spared** Hamstrings (L5,S1,S2, sciatic nerve)
Neurology Review

Foot Drop Sensory Deficits

Sciatic Nerve

Common Peroneal Nerve

Leg Dermatomes
Neurology Review

Neuromuscular Junction

• Clinical Features
  • Ophthalmoparesis, respiratory and bulbar weakness, fatigable weakness, preserved DTR’s

• Myasthenia Gravis
  • Auto-immune humoral attack of muscarinic Acetylcholine receptors
  • Fatiguable weakness
  • Dx confirmed with Tensilon Test (Edrophonium), Electrodecrement of > 10% CMAP amplitude with repetitive nerve stimulation, or the presence of antibodies to the ACHR at the NMJ
  • Co-Morbid autoimmune thyroiditis, malignant thymoma
  • Symptomatic treatment with Mestinon with (acetylcholinesterase inhibitor)
  • Immune supression with Steroids, Immuran, mycophenylate mofetil, cyclosporin, cellcept.
  • Acute attacks treated with IVIg, IV steroids, or Plasmapharesis
Neurology Review

Neuromuscular Junction

- **Botulism**
  - Pre-synaptic release of Acetylcholine at NMJ and parasympathetic NS
  - Light chain of toxin proteolytically cleaves SNAP-25 and synaptic vesicle docking protein leading to reduced release of Ach
  - Clinical scenarios may include:
    - Ingestion of pre-formed toxin (rapid respiratory and bulbar weakness and death)
    - Wound botulism from IVDA (slower-onset limb weakness followed by bulbar weakness)
    - Infantile Botulism (subacute onset of weakness, dysphagia, and constipation)
  - Cosmetic and medical indications for use
    - Dystonia
    - Hyperhidrosis
    - Wrinkles

- **Lambert Eaton Myasthenic Syndrome**
  - Presynaptic release of Acetylcholine impaired. Paraneoplastic syndrome
  - Antibodies to voltage-gated Ca channel
Neurology Review

Myopathies

- Clinical features
  - Proximal weakness. +/- ↑CPK. May need muscle biopsy to make diagnosis

- Congenital Myopathies
  - Structural Myopathies: nemaline rod, central core, etc
  - Metabolic Myopathies: Pompe’s, McArdles, mitochondrial, etc
  - Muscular Dystrophies: Duchene’s, Limb Girdle, Emory Dreyfuss MD’s

- Acquired Toxic Myopathies
  - Statin myopathy

- Acquired Inflammatory Myopathies
  - Dermatomyositis
  - Polymyositis
  - Inclusion Body Myositis

- Rhabdomyolysis
  - ↑CPK, ↑K+, Myoglobinuria, Renal failure
  - EtOH, heat stroke, sympathomimetics, malignant hyperthermia, trauma, etc
Neurology Review

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Neurology Review

Vertigo

- **Peripheral Vertigo:** Due to damage or malfunction of the peripheral vestibular apparatus
  - Vertigo is often severe, positional, fatigable, of short duration with a lag time of a few seconds following movement of the head
  - **Acute Vestibulitis:** Viral etiology, lasts weeks and resolves spontaneously. Dysfunction of the labyrinth causes imbalance of firing with more output from unaffected labyrinth. Nystagmus fast beat away from affected ear. No hearing loss. Rx with meclizine.
  - **Benign Positional Vertigo:** Older pt’s. Severe vertigo lasting a few seconds brought on by head turning. Ca++ otoliths in utricle and saccule that migrate into the ampule of one semicircular canal. Treatment with the modified Epley Liberation maneuver.
  - **Meniere’s Syndrome:** Recurrent vertigo, tinnitus, and hearing loss. Lasts hours. May be caused by endolymphatic hydrops. May result in permanent hearing loss. Treatment with hydrochlorothiazide.
  - **Perilymphatic Fistula:** Due to trauma. Intermittent or positional vertigo with conductive hearing loss. Usually heals on own.
Neurology Review

Vertigo

- **Central Vertigo:** Due to dysfunction of the VIII nerve or central brainstem connections
  - Less severe than peripheral vertigo. Less related to changes in head position. Non-fatigable. Longer lasting than peripheral vertigo.

- **Acoustic Neuroma**
  - Hearing loss, tinnitus and vertigo. May also involve cn’s V and VII resulting in facial numbness and weakness. Ataxia from CPA involvement
  - Bilateral Acoustic neuromas seen in NF-2

- **Vertebro-Basilar Insufficiency**
  - Episodic brainstem dysfunction due to vascular insufficiency
  - Spells of diplopia, vertigo, dysarthria, ataxia, facial and limb weakness and numbness lasting minutes. Exam between attacks may be normal.
  - Usually seen in older pt’s with atherosclerotic disease elsewhere
II (Optic Nerve)

- Homonymous Hemianopsia: Anything behind optic chiasm
- Bitemporal Hemianopsia: Lesion of the optic chiasm
- Superior Quadrantanopsia: Meyers Loop of optic radiations anterior Temporal lobe
Neurology Review

Cranial Nerves

- III (Oculomotor Nerve) palsy
  - Posterior Communicating Artery Aneurysm
  - Uncal Herniation

- VI (Abducent Nerve) palsy
  - Non-localizing VIth nerve palsy with any cause of raised ICP

- VII (Facial nerve)
  - LMN = whole face. UMN = spares upper face
Neurology Review

Brainstem Reflexes

- Pupillary Light Reflex:
  - II is Afferent. III is Efferent
  - Horner’s Sign (meiosis, ptosis, anhydrosis) due to SNS dysfunction to head/face. Long Pathway.
  - Afferent Pupillary Defect: CN II dysfunction (ie optic neuritis)

- Oculocephalic Reflex
  - Oculocephalic Reflex: Slow drift component of nystagmus ipsilateral to ear with cold H2O. Fast-Beat saccadic component to contralateral side
  - VIII is Afferent. III, IV, VI are Efferent

- Corneal Blink Reflex
  - Ophthalmic Division of the Trigeminal Nerve (V-1) is Afferent. VII is Eff.

- Gag Reflex
  - IX is Afferent. X is Efferent
# The Neurological Motor Examination

<table>
<thead>
<tr>
<th></th>
<th>Upper Motor Neuron</th>
<th>Lower Motor Neuron</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Strength</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Tone</strong></td>
<td>Spasticity</td>
<td>Hypotonia</td>
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<tr>
<td><strong>DTR’s</strong></td>
<td>Brisk DTR’s</td>
<td>Diminished or Absent DTR’s</td>
</tr>
<tr>
<td><strong>Plantar Responses</strong></td>
<td>Upgoing Toes</td>
<td>Downgoing Toes</td>
</tr>
<tr>
<td><strong>Atrophy/Fasciculations</strong></td>
<td>None</td>
<td>+/−</td>
</tr>
</tbody>
</table>