Approach to neurology

• LOCALIZE the LESION!
  – “What means where”

• ELIMINATE EMERGENCIES FIRST!
  – VET those emergencies
    • Vascular
    • Electrical
    • Traumatic

• Short differential diagnosis

• Treat the underlying pathology
2 overriding concepts

- Ockham’s Razor for lesions
  - Put the lesion in *one site* if possible OR
  - Localize to a particular tissue such as:
    - Gray matter
    - White matter
    - Muscle

- “WIRE CROSSING IS BAD”
  - Tracts are PROPERLY laminated
  - KNOW where the tracts cross
4 Tracts for lesion localization

- MOTOR system [most important for localization]
  - Corticospinal
    - Cortical
    - Subcortical
  - Anterior horn cell starts the peripheral nervous system
- 2 sensory systems
  - DORSAL columns
  - Spinothalamic
- Cerebellar [least important for localization]
Motor system first!

[Strength, tone, atrophy, ±reflex]

• CNS lesion?
  – Hyper-reflexia (after initial shock)
  – Increased tone
  – NO ATROPHY (caveat: anterior horn cells)

• Peripheral lesion?
  – Decreased reflexes
  – Decreased tone
  – Atrophy (caveat: demyelinating neuropathies)
Reflex levels-count to 8

<table>
<thead>
<tr>
<th>REFLEX</th>
<th>ROOT LEVELS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ankle</td>
<td>S1 – S2</td>
</tr>
<tr>
<td>Knee</td>
<td>L3 – L4</td>
</tr>
<tr>
<td>Biceps brachii</td>
<td>C5 – C6</td>
</tr>
<tr>
<td>Brachioradialis</td>
<td>C5 – C6</td>
</tr>
<tr>
<td>Triceps brachii</td>
<td>C7 – C8</td>
</tr>
</tbody>
</table>
Central nervous system

• Cortical?
  – Aphasia?
  – Visual field cut?
  – Abulia? Apraxia?
  – Contralateral neglect?

• Subcortical?
  – Hemiparesis

• Brainstem
  – Cranial nerve lesion IPSILATERAL
  – Hemiparesis CONTRALATERAL

• Spinal cord?
  – Both sides of body usually affected
  – SENSORY LEVEL LOCALIZES TO SPINAL CORD
Peripheral nervous system

- Anterior horn cell/motor nerve roots
  - Invariant weakness with atrophy
  - NO SENSORY LOSS
- Peripheral nerve
  - Distal weakness WITH sensory loss
- NMJ
  - Waxing/Waning weakness
  - NO SENSORY LOSS
- Muscle
  - Invariant proximal>distal weakness
  - NO SENSORY LOSS
SENSORY SYSTEM (2 types)

- LARGE FIBER/DORSAL COLUMN
  - Vibration/Position sensation
  - Afferent portion of reflex arc
  - HIGH CROSS

- SMALL FIBER/SPINOThALAMIC
  - Light touch/Pin/Temperature sensation
  - LOW CROSS
Sensory tracts just below the cervicomedullary junction

- Dorsal columns: [CROSS at C1]
  - Sacral fibers are most MEDIAL
  - Cervical fibers are most LATERAL

- Spinothalamic: [IMMEDIATE CROSS]
  - Sacral fibers most LATERAL
  - Cervical fibers most MEDIAL
2nd Step: Sensory system

• If peripheral: NO DISSOCIATION
  – Sensory modalities lost together
  – No “splitting”
• If central: DISSOCIATION or LEVEL
  – Sensory level = SPINAL CORD
  – Dissociation
    • Pain and temperature affected on one side
    • Vibration and position affected on the other
Cerebellum

• ABOVE cervicomedullary junction
  – Usually contralateral to affected side

• BELOW cervicomedullary junction
  – Tracts cross twice
  – Lesion is ipsilateral to affected side
Major Neurology Topics

- Headaches/Brain Tumors
- Delirium/Dementia/Stupor and Coma
- Dizziness/Vertigo/Syncope/Hearing Loss
- Movement disorders
- MS/Spinal cord diseases
- Neuromuscular disease
- Epilepsy
- Stroke
Headaches

• RED FLAGS:
  – “Worst headache of life”
  – Headache with ANY neurological deficits
  – Headache with papilledema
  – Headache with fever
  – Headache with jaw claudication

• ALL other headaches are usually migraine
  – Cluster headaches are extremely rare
  – Tension headaches are rarer still
Headaches

• Any “RED FLAG” headache requires:
  – Head CT WITHOUT contrast (exclude bleed)
  – LP if CT is negative
  – ESR if temporal arteritis is suspected

• Non red-flag headaches
  – Migraine
  – Cluster
  – Chronic paroxysmal hemicrania
Migraine headaches

- Unilateral pulsatile HA with photophobophobia
- Pathophysiology—Low pontine serotonin
- Females > Males

Prophylaxis
- Tricyclic antidepressants
- Beta-blockers
- Anticonvulsants (Depakote, Topamax)

Abortive
- Triptans
- Sedatives
- Ergotamines
Cluster headaches

• Similar pathophysiology
• Males > females
• Other differences
  – Wakes people from sleep in early AM
  – Relieved by 10-12 liters of oxygen by mask
  – Extreme agitation (migraineurs are reclusive)
  – Rhinorrhea and lacrimation
Brain Tumors

• Infiltrators
  – Primary CNS neoplasms
  – Neural crest derived tumors [melanoma]

• Displacers
  – Originate elsewhere
  – Supporting elements
    • Meningioma
    • Ependymoma
## Tumor Comparison

<table>
<thead>
<tr>
<th></th>
<th>Infiltrators</th>
<th>Displacers</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Size at detection</strong></td>
<td>Large</td>
<td>Small</td>
</tr>
<tr>
<td><strong>Usual presentation</strong></td>
<td>Seizures</td>
<td>Seizures/HA</td>
</tr>
<tr>
<td></td>
<td>Minimal deficits</td>
<td>Obvious deficit</td>
</tr>
<tr>
<td><strong>Location</strong></td>
<td>Diffuse</td>
<td>Gray-White junction</td>
</tr>
</tbody>
</table>
Delirium, Dementia, Stupor, Coma

• RED FLAGS:
  – Acute onset
  – Focal neurological deficits
  – Metabolic derangements

• DISEASES you should NEVER MISS:
  – Wernicke’s (always give Thiamine)
  – B12 or Folate deficiency
  – Thyroid disease induced encephalopathy
## Delirium, Dementia, Stupor, Coma

<table>
<thead>
<tr>
<th></th>
<th>Sensorium</th>
<th>Interaction with environment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Delirium</td>
<td>Agitated</td>
<td>Hostile but purposeful</td>
</tr>
<tr>
<td>Dementia</td>
<td>Clear</td>
<td>Normal on the surface</td>
</tr>
<tr>
<td>Stupor</td>
<td>Clouded</td>
<td>Minimal but purposeful</td>
</tr>
<tr>
<td>Coma</td>
<td>Obtunded</td>
<td>Reflex actions only</td>
</tr>
</tbody>
</table>
Approach-
Focal neurological deficit?

- Yes: The brain is likely at fault
  - Head CT WITHOUT CONTRAST-STAT
  - LP if head CT is negative
  - EEG if LP is negative
- No: The brain is secondarily affected
  - This is a TOXIC-METABOLIC process
  - Medication/drug abuse history
  - CBC, CMP, TSH, B12, Folate
  - RPR, HIV with high index of suspicion
Dementia

• Do not diagnose until you have excluded:
  – Pseudodementia (e.g. depression)
  – Treatable dementias (B12, Folate, TSH)
• Otherwise, it is probably Alzheimer’s
  – Procholinergics slow the rate of decline
  – They do NOT reverse the disease process
Delirium

- 99.9% is toxic-metabolic in origin
- Exception: Non-dominant hemisphere lesions
  - Acute stroke
  - Herpes simplex encephalitis

- Treatments
  - Find the underlying cause and address it
  - Tincture of time
Stupor and Coma

- **FOCAL DEFICIT** = Brain is at fault
- **LOCALIZE** the lesion
  - Examine ALL cranial nerve reflexes:
    - Pupillary
    - Blink
    - Oculocephalic [NOT Doll’s eyes]
    - Pharyngeal [NOT gag]
  - Posturing?
    - Decorticate or arm flexion = above red nucleus
    - Decerebrate or arm extension = below red nucleus
Dizziness, Vertigo, Hearing Loss

• Define vertigo
  – Illusion of motion
  – “Spinning” is not necessary

• Occurs from mismatch of sensory inputs
  – Vision
  – Posterior columns
  – Vestibular system

• MUST differentiate “central “ from “peripheral”
### Central versus peripheral

<table>
<thead>
<tr>
<th>Condition</th>
<th>Central</th>
<th>Peripheral</th>
</tr>
</thead>
<tbody>
<tr>
<td>Long tract signs</td>
<td>Presence confirms central</td>
<td>NEVER</td>
</tr>
<tr>
<td>Hearing loss</td>
<td>EXTREMELY RARE</td>
<td>Virtually diagnostic</td>
</tr>
<tr>
<td>Fatigues</td>
<td>No</td>
<td>Yes</td>
</tr>
</tbody>
</table>
Common causes of vertigo

• **Benign Positional**
  – Provoked in 1 position ONLY
  – Fatigues with each provoking maneuver

• **Meniere’s disease is TRIAD of**
  – Hearing loss [Localizes to periphery]
  – Tinnitus
  – Vertigo

• **Stroke**
  – Long tract signs
  – Cerebellar deficits
Movement disorders

- BALANCE of Ach and DA
  - Increased Ach = Decreased DA
  - Increased DA = Decreased Ach

- Excess DA leads to:
  - Psychosis
  - Chorea

- Reduced DA leads to
  - Parkinsonism
  - Dystonia/Rigidity
Movement disorders

• Parkinson’s Disease [Idiopathic]
  – Too little DA in Substantia Nigra
  – FOUR cardinal features
    • Tremor
    • Rigidity
    • Bradykinesia
    • Postural instability
  – Should respond to DA agonists

• Parkinson’s Plus syndromes do NOT respond to DA therapy
Tremors

• Essential
  – Action tremor
  – Disappears at rest
• Parkinson’s tremor
  – Tremor at REST
  – Disappears wit movement
• Cerebellar Tremor [RARE]
  – Worsens as the target is approached
  – Clinically obvious
Movement disorder therapy

• “Move too much…give them sedatives”
  – Beta-blockers
  – Barbiturates
  – Benzodiazepines

• “Move too little…try some Sinemet”
  – Use DA agonists below age 60
  – Start Sinemet at age 60 or greater
Some exceptions

- Acute dystonic reaction from DA blocker
  - Block Ach to restore balance
  - Use diphenhydramine IV

- Parkinson’s tremor does NOT respond to DA therapy
  - Non-anticholinergic sedatives required

- Focal dystonia responds best to Botulinum toxin injectins
Multiple Sclerosis

- A WHITE MATTER CNS disease
- Two distinct clinical episodes separated in SPACE and TIME
  - Space: Must affect DIFFERENT white matter locations
  - Attacks must be more than 30 days apart
- MRI criteria can be used instead of an actual SECOND clinical attack
1 lesion ≠ Multiple Sclerosis

• Diagnosis based on location
  – Optic tract = Optic Neuritis
  – Spinal cord = Transverse myelitis
  – Cerebellum = Acute cerebellitis

• Risk of MS increased after first attack

• IV steroids hasten recovery but have NO EFFECT on probability of second attack

• IV steroids are superior to oral steroids which are = placebo
MS therapy

• Acute attack- IV methylprednisolone
  – Given as 500 mg IV bid for 3-5 days
• Reduce relapse frequency/progression
  – Interferons
  – Copaxone
• When above agents fail:
  – Cyclophosphamide
  – Mitoxantrone
Spinal cord disorders

- ALWAYS eliminate cord compression:
  - Plain X-rays to exclude bony pathology
  - MRI
- CSF exam if previous studies are normal:
  - Transverse myelitis
  - Meningomyelitides
- Other odd causes
  - Spinal cord strokes
  - B12 deficiency
Neuromuscular

• LOCALIZE the lesion
  – Motor, Sensory or both?
  – Trunk, shoulder or hip girdle involved?
  – Face involved?

• If motor affected:
  – Constant or variable weakness?
  – Atrophy?
  – Tone increased or decreased?
Nerve structure

• Myelin sheath
  – For rapid conduction of impulses
  – Vibration and position sense
  – Motor function

• Vasa nervorem
  – Blood vessels end on nerve surface
  – Loss of perfusion affects small fibers in nerve center

• Axons
  – Most metabolically active
  – Atrophy when the motor axons are injured
Neuropathy vs. myopathy vs. NMJ

• Neuropathy vs. myopathies
  – Most neuropathies cause distal before proximal dysfunction
  – Muscle diseases affect proximal before distal

• NMJ disorders usually involve the CNs
  – NO sensory loss
  – NO atrophy
  – Extraocular muscles affected early
Myelinopathy versus axonopathy

- Reflexes lost early
- Large fiber sensory loss
- Weakness with minimal atrophy
- CSF protein may be increased
- Patterns other than distal to proximal

- Reflexes retained until late
- Small > large fiber sensory loss
- CSF protein normal
- Distal to proximal dysfunction
Myopathies

• Inflammatory usually have increased CPK
  – Polymyositis: Cellular infiltrate
  – Dermatomyositis: A vasculitis
• Non-inflammatory have normal to minimally elevated CPK
• Diagnosis can only be established by biopsy
Epilepsy

- A disorder of the cortical gray matter
- Definition:
  - 2 UNPROVOKED seizures
  - A CLINICAL diagnosis
- Provoked seizure examples:
  - Syncope
  - Sleep deprivation
  - Alcohol withdrawal
Epilepsy diagnosis

• Clinical to classify the epilepsy
  – Reliable witness
  – Tongue laceration
  – Prolonged LOC

• EEGs do NOT diagnose epilepsy
  – Many patients have normal EEGs
  – Spikes on an EEG do NOT prove epilepsy
Evaluation

• Elicit complete history to exclude a provoked seizure
• CBC, CMP, UDS at a minimum
• Imaging
  – Generalized only requires head CT
  – Focal onset REQUIRES MRI
• EEG
  – Highest positives within 24 hrs
  – Anticonvulsants do NOT suppress abnormalities
Treatment

- Treatment of a first unprovoked seizure is controversial but most will do so if any of the following are noted:
  - Focal onset
  - EEG abnormalities noted
  - MRI abnormality in cortical gray noted
- All anticonvulsants have the same efficacy for most seizure types—they differ in side effects
Treatment exceptions

- Juvenile myoclonic epilepsy
  - Valproic acid
  - Lamotrigine
- Absence seizures
  - Valproic acid
  - Ethosuximide