Introduction to Neurology & Neurologic Emergencies

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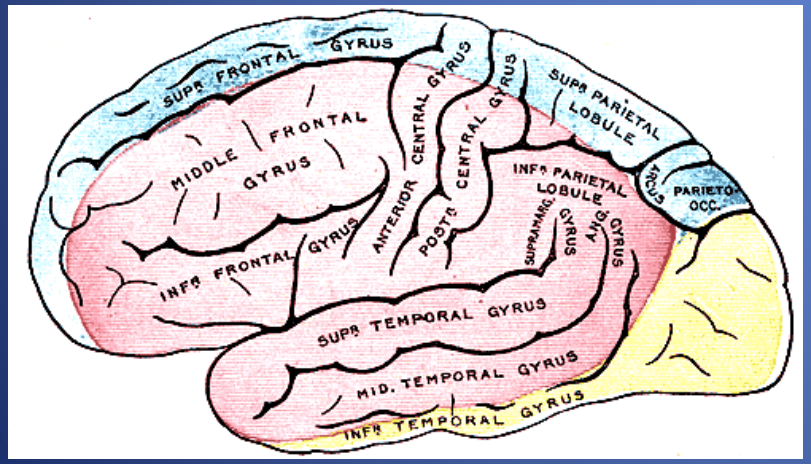
What is different about Neurology? *Localization*

Central nervous system

- Brain
- Spinal cord



Peripheral nervous system

- Nerve root
- Plexus
- Peripheral nerve
- Neuromuscular junction
- Muscle

The most important step in neurologic localization is differentiating a *central nervous system* lesion from a *peripheral nervous system* lesion

Localization
Aided by Neurologic Examination

Upper motor neuron

- Increased tone (spasticity)
- Hyperreflexia; pathologic reflexes (Babinski, Hoffman)

Lower motor neuron

- Reduced tone (flaccidity)
- Hyporeflexia
- Severely reduced bulk with fasciculations

Case Ching

- A 71 year old man presents for progressive difficulty walking
  - Developed back pain several weeks ago
  - Followed by numbness and weakness in the legs
- Physical Examination
  - Vitals stable
  - Cranial nerves normal
  - Hypertonia in LE with moderate weakness; UE normal
  - Hyperreflexia in LE; UE normal
  - Extensor plantar responses bilaterally
  - Sensation reduced to all modalities to mid-abdomen
  - Spastic gait

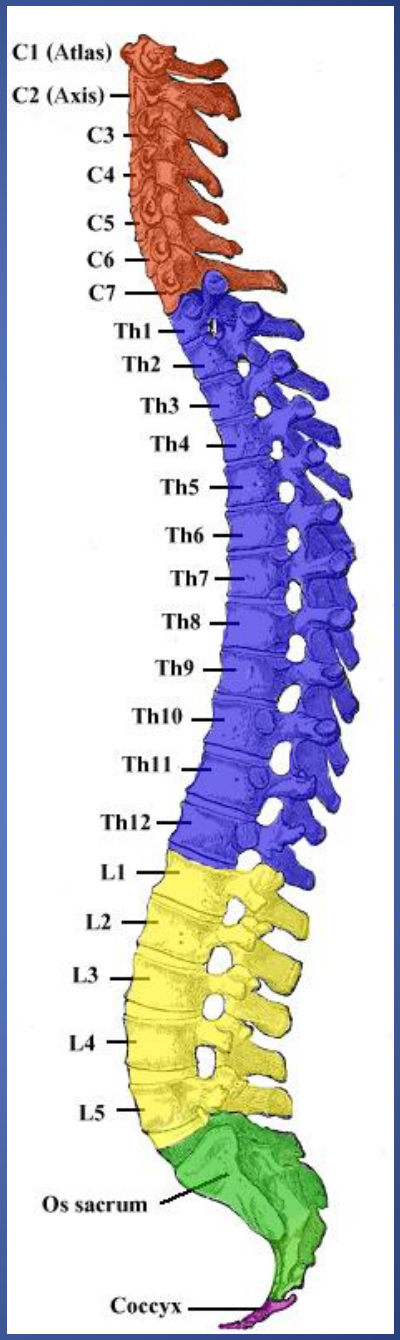
Case Ching

- Can you localize the lesion?

Case Ching

- What studies do you want to order and why?

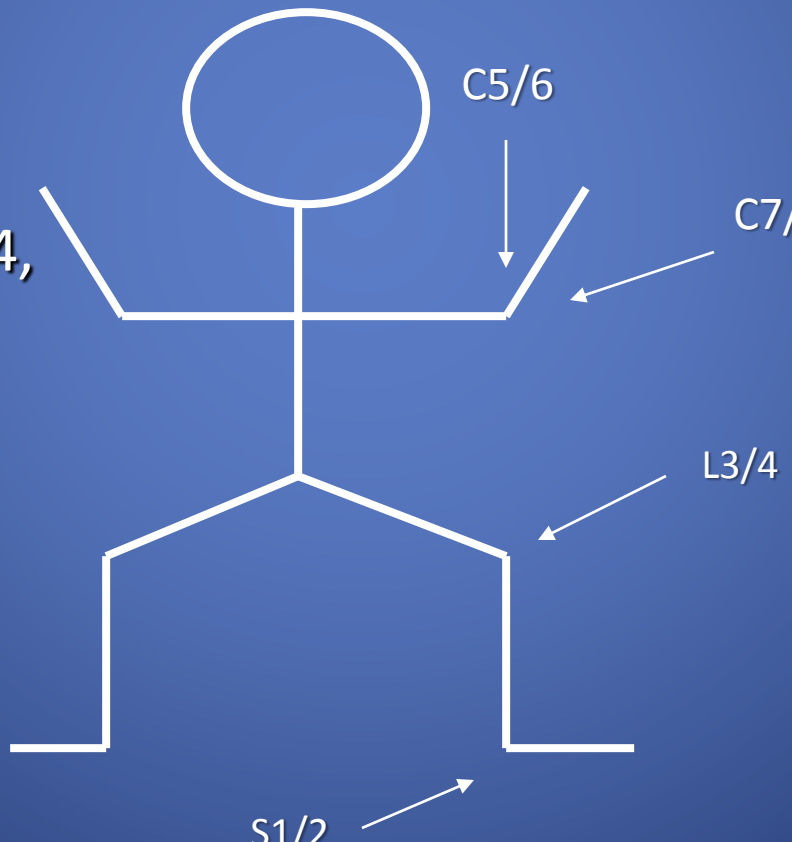
Where does the spinal cord end?



Localization

- Spinal levels

Its easy to remember…1/2, 3/4, 5/6, 7/8



ALWAYS ABNORMAL:

- Asymmetric reflexes
- Pathologic responses (Babinski, clonus = CNS)
- Absent reflexes (PNS)

Spinal Cord Compression

Management

- Dexamethasone (100 mg IV followed by 16 mg PO daily in divided doses)
- Surgical evaluation
- Radiotherapy

Differential

Metastatic disease

Epidural hematoma

Epidural abscess

Vertebral collapse (2' malignancy)

Aortic dissection

Case Gilmore

- A 28 year old man presents for progressive difficulty walking
  - “Stomach flu” 2 weeks ago
  - Recently developed low back pain and numbness in his feet
  - Difficulty standing and climbing stairs, tripping frequently
- Physical Examination
  - Vitals stable
  - Cranial nerves normal
  - Mild hypotonia, normal muscle bulk; mild distal LE weakness
  - Distal sensation reduced symmetrically to all modalities but normal in the trunk
  - Deep tendon reflexes absent
  - Slapping gait with bilateral foot drop

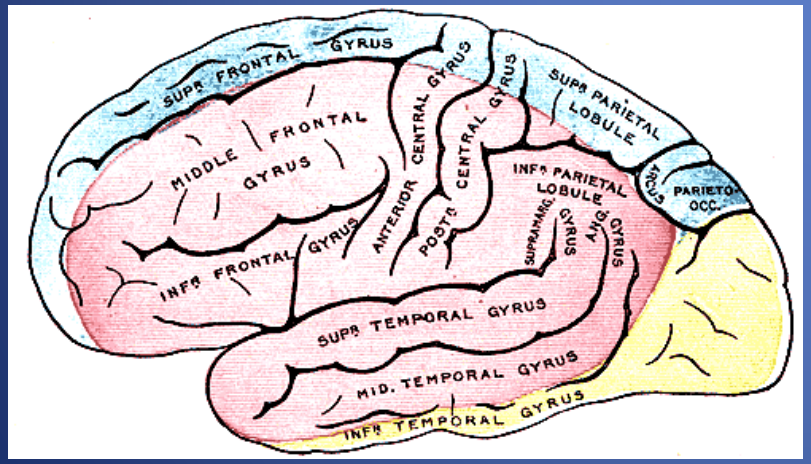
Case Gilmore

- Can you localize the lesion?

Localization

Central nervous system

- Brain
- Spinal cord



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The most important step in neurologic localization is differentiating a *central nervous system* lesion from a *peripheral nervous system* lesion

Case Gilmore

- How does the neurologic exam helps you in sorting out myelopathy, ,neuropathy, myopathy and NMJ?

Localization

	Neuropathy	Myopathy	Myelopathy	NMJ disorder
Weakness	Distal > proximal	Proximal > distal	Below level of lesion	Fluctuating/ muscle fatigue
Deep tendon reflexes	Severe reduction/ early loss	Mild reduction/ late loss	Increased	Normal or mildly reduced
Sensory	Distal/ ascending	Preserved	Sensory level	Preserved

Case Gilmore

- What studies do you want to order and why?

Guillain-Barre Syndrome

- CSF: cytoalbuminological dissocation (elevated protein with few or no mononuclear cells)
  - May be normal in the first week
  - If WBC count >10 consider Lyme, HIV, sarcoidosis
- Electromyography/nerve conduction study
  - Reduced nerve conduction velocities
  - Conduction block
  - Prolonged F-waves
- Antiganglioside antibodies
  - GM1 Abs (correlate with C. jejuni infection)
  - GQ1b associated with C. Miller Fisher variant (ataxia, areflexia & ophthalmoparesis)

Guillain-Barre Syndrome

- Typically follows an infectious process (2/3)
  - C. jejuni, CMV, EBV, M. pneumoniae
- Presents with ascending numbness/tingling, can be painful
- Weakness typically follows sensory disturbances
- ***Areflexia***
- Autonomic dysfunction
  - Labile BP, arrhythmia
  - Bowel and bladder function typically spared
- Symptoms should not proceed >8 weeks
  - 98% achieve “plateau phase” by 4 weeks
  - Duration of “plateau” 12 days

Treatment

- Telemetry, respiratory parameters, ICU monitoring for dysautonomia and respiratory compromise
- IVIG (0.4g/kg/day for 5 days) versus plasma exchange
  - Ease of administration, fewer complications, preferred in hemodynamically unstable patients
  - Should be started within 2 weeks
- Corticosteroids have not been shown to be beneficial
- Intubation criteria:
  - VC <15-20 mL/kg (<30% baseline)
  - PO_2<70 mmHg
  - Oropharyngeal weakness, weak cough, suspected aspiration

GBS vs ATM

TABLE 5. Distinguishing Features Between Guillain-Barré Syndrome and Transverse Myelitis

Characteristics	Transverse Myelitis	Guillain-Barré Syndrome
Motor findings	Paraparesis or quadriparesis	Ascending weakness LE > UE in the early stages
Sensory findings	Usually can diagnose a spinal cord level	Ascending sensory loss LE > UE in the early stages
Autonomic findings	Early loss of bowel and bladder control	Autonomic dysfunction of the cardiovascular (CV) system
Cranial nerve findings	None	EOM palsies or facial weakness
Electrophysiologic findings	EMG/NCV findings may be normal or may implicate the spinal cord: prolonged central conduction on somatosensory evoked potential (SEP) latencies or missing SEP in conjunction with normal sensory nerve action potentials	EMG/NCV findings confined to the PNS: motor and/or sensory nerve conduction velocity reduced, distal latencies prolonged; conduction block; reduced H reflex usually present
MRI findings	Usually a focal area of increased T2 signal with or without gadolinium enhancement	Normal
CSF	Usually, CSF pleocytosis and/or increased IgG index	Usually, elevated protein in the absence of CSF pleocytosis

CSF Analysis

Table 1. Typical CSF Findings in Patients With and Without Meningitis

Parameter	Normal	Bacterial Meningitis	Viral Meningitis	Fungal Meningitis	Tuberculous Meningitis
Opening pressure (mm H_2O)					
WBC count (mm^3)					
WBC differential					
Protein (mg/dL)					
Glucose (mg/dL)					
Gram stain (% +)					

+: positive; –: negative; AFB: acid-fast bacilli; CSF: cerebrospinal fluid; L: lymphocytes; NA: not applicable; PMN: polymorphonuclear cells; WBC: white blood cells. Source: References 9, 10.

Case Montoya

- A 34 year old woman presents to the ER with complaint of eyelid drooping
  - A week earlier went to ER and given steroids for an asthma exacerbation and ciprofloxacin for an asymptomatic UTI
  - Feels “weak all over,” progressively short of breath
- Physical Examination
  - Speech is soft with a nasal quality
  - Fluctuating bilateral ptosis, pupils equal and reactive, “snarl” smile
  - Normal bulk/tone; strength can be overcome after <60s of effort
  - Normal reflexes, normal sensory examination
  - Gait normal, but cannot stand up 10 times consecutively

Case Montoya

- Can you localize the lesion?
- What studies do you want to order and why?

Acute treatment

- Respiratory parameters
  - 30% of pts develop respiratory muscle weakness and crisis occurs in 15-20%
- Telemetry: 14% of pts in myasthenic crisis have some degree of arrhythmias
- Intubation criteria
  - VC < 15 mL/kg
  - Stop anti-cholinesterase medication (causes excessive bronchial secretions and diarrhea)
- PE (improvement in 75%), IV Ig

Myasthenia gravis

- Ice pack test: improves ptosis in MG > 80%
- Tensilon test (edrophonium)
- Repetitive stimulation or single fiber EMG
- Autoimmune work up

- AchR Ab
  - 85% w/ gMG
  - 50-60% w/ oMG
- Chest CT
  - Thymectomy may increase chances of remission
  - May be less effective in ocular myasthenia and those >60

Case Cunningham

- A 32 year-old woman is found on the floor at work, unconscious, but spontaneously breathing.
- Exam:
  - BP 146/75, pulse 80, afebrile.
  - Non verbal, no grimace or movement to noxious stimuli.
  - L pupil 5 mm, sluggishly reactive to light; R pupil 2 mm , briskly reactive.
- Can you localize the lesion?

Coma

- Lowered level of arousal/consciousness
- Localization: reticular activating system, hypothalamus, bilateral or extensive hemispheric lesions (trauma, meningitis/encephalitis, bilateral infarction, metabolic)
- Etiology: structural or metabolic

Coma: respiratory pattern

- Cheyne-Stokes (crescendo-decresendo)
  - Periods of hyperpnea alternating with hypopnea
  - Bilateral hemisphere lesions, increased ICP
  - Also seen in sick elderly or CHF patients
- Central neurogenic hyperventilation
  - Rapid, regular hyperpnea
  - Paramedian reticular formation (midbrain/pons), other brainstem locations
- Respiratory ataxia
  - Irregular, variably shallow and deep respirations
  - Dysfunction of medullary respiratory centers
  - May be a preterminal pattern preceding respiratory arrest
- Abnormal respiratory patterns don't always localize "classically", may be seen in systemic disorders

Coma: level of responsiveness

- Coma: loss of consciousness, no awareness of self or environment, no voluntary movement or response
- Stupor: partial loss of responsiveness, variable impairment of consciousness, difficult to arouse but brief response to stimulation possible
- Lethargy: arousable and responsive, but unconscious/asleep without stimulation
- Examination – assess response to painful stimuli (suprorbital pressure, sternal rub, nasal irritation, nailbed pressure)

Coma: pupil size and reactivity

- Metabolic coma: small, reactive
- Structural lesion: asymmetric, unreactive
- Midbrain lesion: large, unreactive pupil(s)
  - parasympathetic defect
  - uncal herniation, p comm aneurysm, preterminal
- Pontine lesion: pinpoint, reactive pupil(s)
  - sympathetic defect
  - opiate toxicity, hypothermia
- Midposition unreactive: central transtentorial herniation

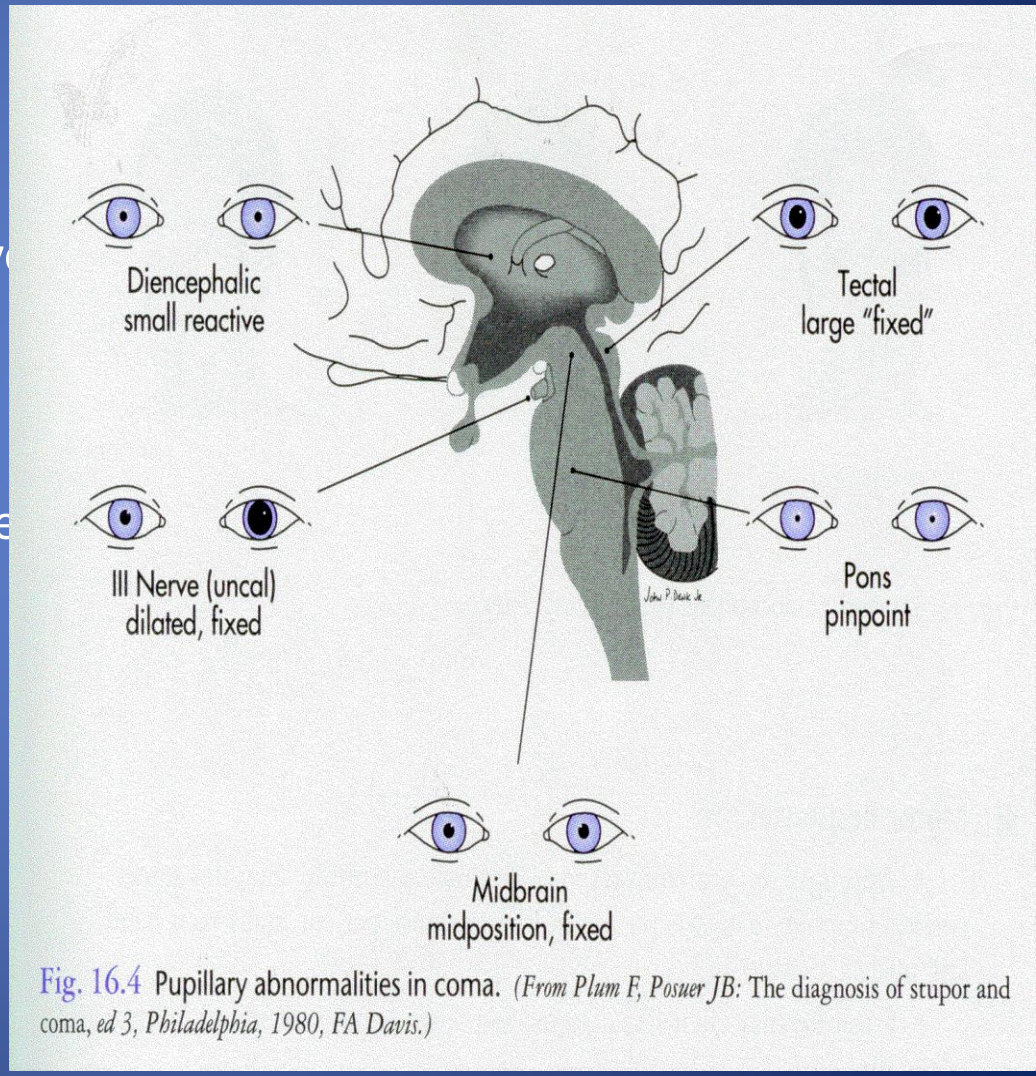


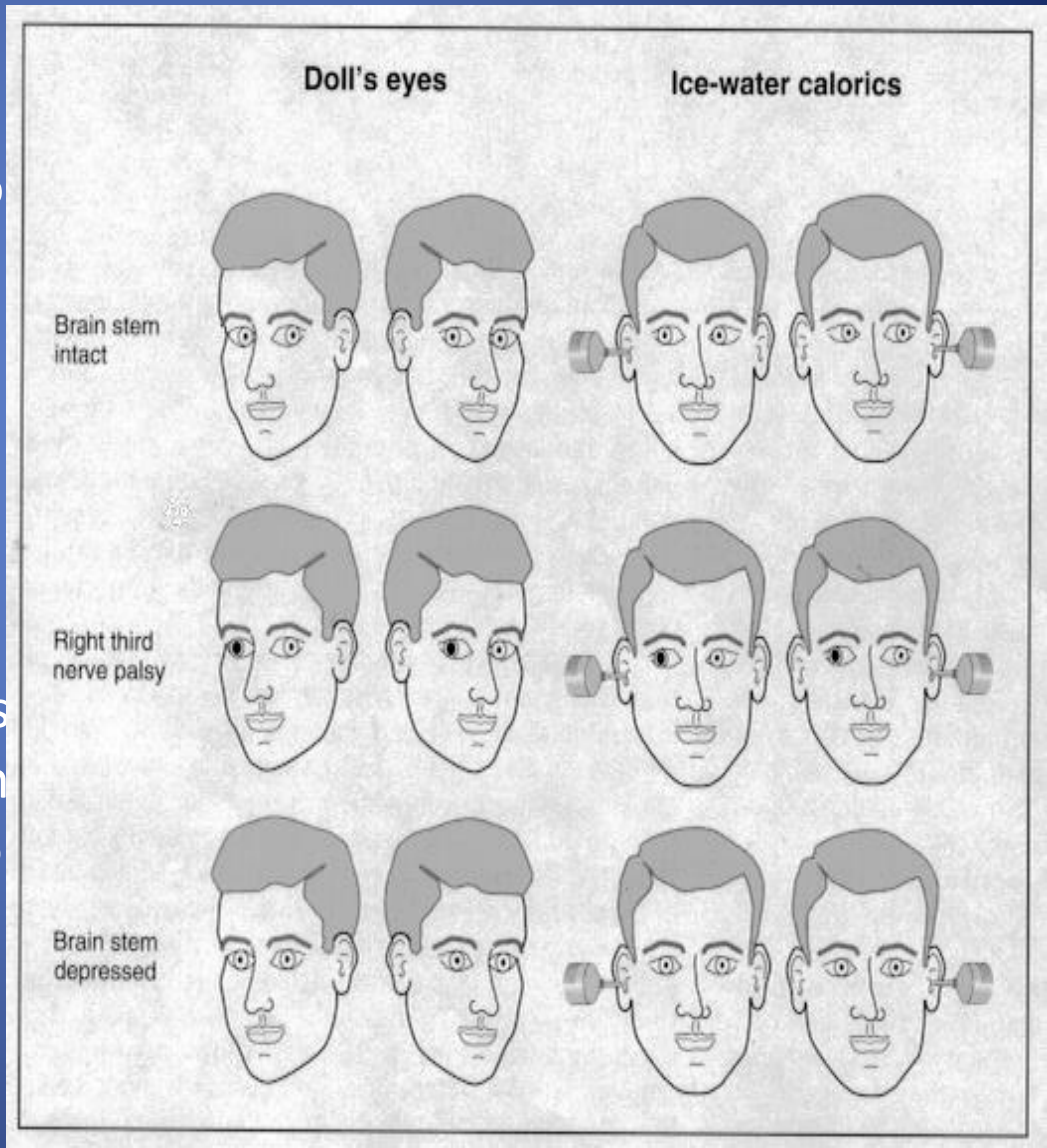
Fig. 16.4 Pupillary abnormalities in coma. *(From Plum F, Posner JB: The diagnosis of stupor and coma, ed 3, Philadelphia, 1980, FA Davis.)*

Coma: eye/eyelid movements

- Check C-spine if any history of trauma
- Rule out vestibulotoxic drugs
- Conjugate eye deviation
  - frontal lobe/ brainstem lesions
  - seizure
- Oculocephalic response
  - Hold eyelids open , briskly rotate head side to side
  - Positive response: contraversive conjugate eye deviation
- Corneal reflex

Coma: oculovestibular reflex

- Oculovestibular reflex
  - Caloric test: inject 20 cc o more ice water into external auditory canal, wait up to 60 seconds for response
  - Performed if oculocephalics absent
  - Absent response suggests sedative drug intoxication brainstem structural lesio or brain death

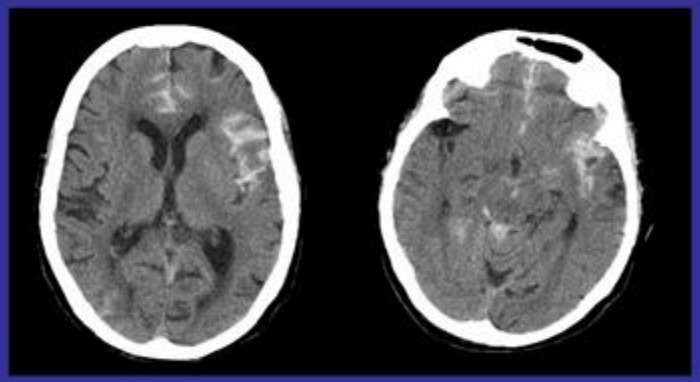


Coma: motor exam

- Tone: flaccidity versus spasticity, asymmetry
- Reflexes: asymmetry, pathologic reflexes
- Response to noxious stimuli
  - Localization and withdrawal
  - Flexion withdrawal
  - Decorticate rigidity
  - Decerebrate rigidity
  - No response
- Decorticate (flexor) posturing
  - flexion of UE
  - extension of LE
  - cortical level
- Decerebrate (extensor) posturing:
  - extension of UE and LE
  - midbrain level
- Myoclonus: anoxia, metabolic coma

Coma: management

- Protect airway, ensure oxygenation, maintain blood pressure
- Neck immobilization, if indicated
- Correct deficiencies in glucose (50% glucose w/ IV thiamine)
- Consider nalaxone/ flumazenil
- History/examination
- Urgent, noncontrast brain CT
- Check metabolic panel, drug screen



Subarachnoid hemorrhage

- Berry aneurysm most common cause if no history of trauma
- Verify blood by CT, or LP if CT normal
- Emergent angio and surgical or interventional management

Intubate if GCS <8 or hypoxemia

IV Fluids (2L 0.9% NS)

MAP ≤120 mm Hg and systolic <180 mg Hg

Nimodipine 60 mg 6xs/daily

Phenytoin (if seizures occur)

EVD for acute hydrocephalus

Case Sanchez

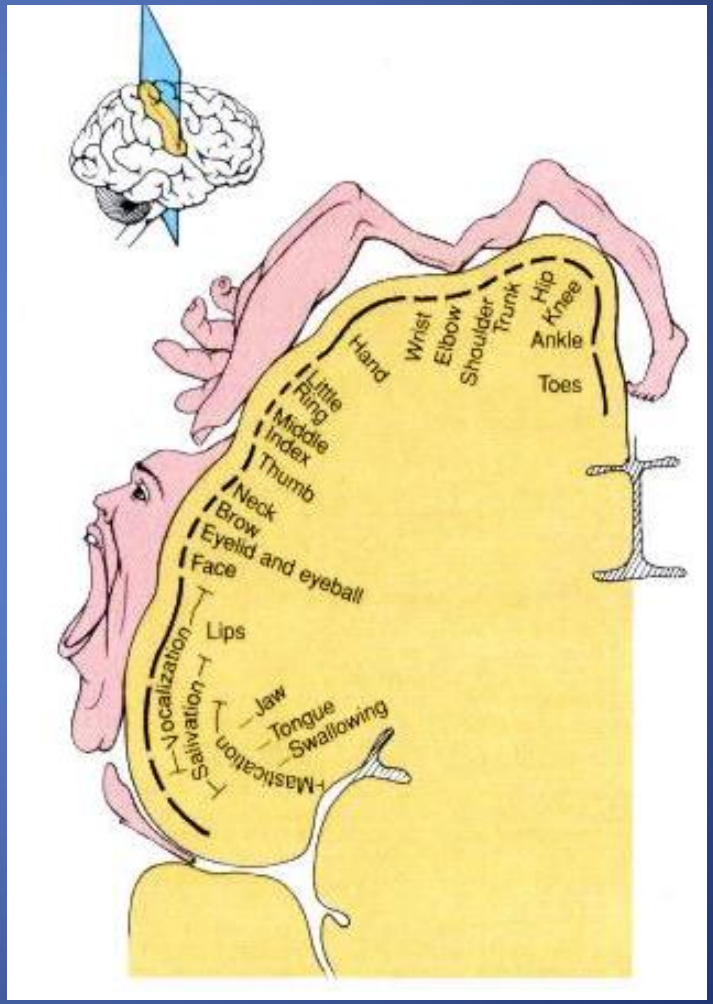
- A 27 year old woman is brought to the ER for “sudden onset of confusion”
- Physical Examination
  - Global aphasia: unable to produce spontaneous language, repeat phrases or follow commands
  - Right lower facial weakness
  - Right arm weakness

Case Sanchez

- Can you localize the lesion?

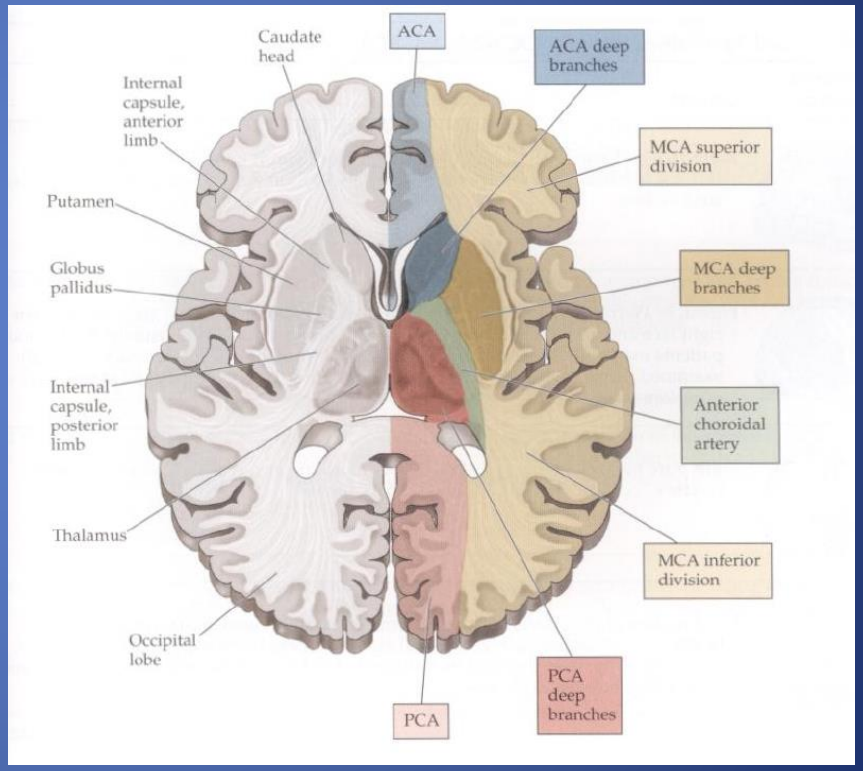
Localization

- Central nervous system
- Left hemisphere
  - Dominant in 99% of R handers, 60-70% L handers
- Cortex: perisylvian language area
  - Expressive, nonfluent aphasia (frontal lobe)
  - Receptive aphasia (temporal lobe)



Acute stroke symptoms

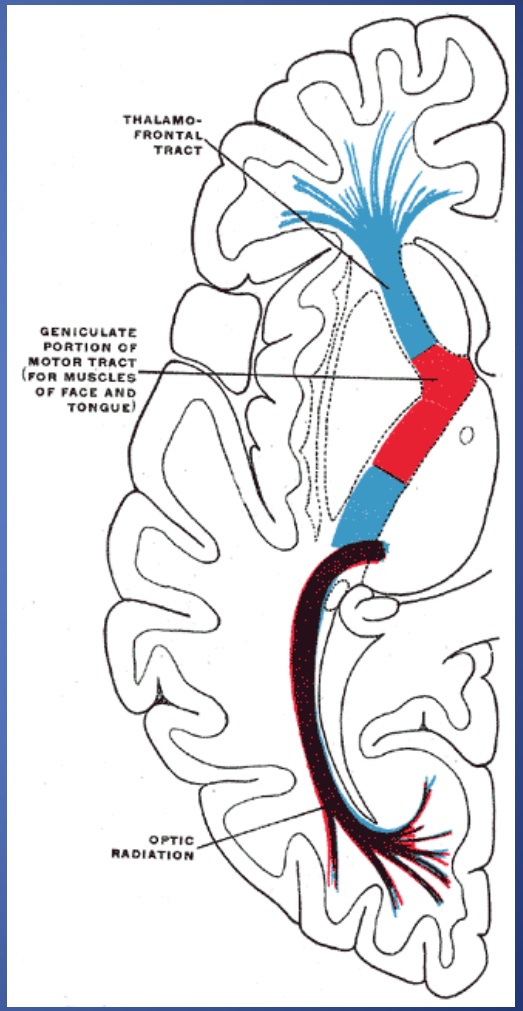
- Hemiparesis or isolated limb weakness
- Hemisensory deficit
- Monocular or binocular (typically homonymous) vision loss
- Brainstem deficits (diplopia, dysphagia, deafness, crossed sensory or motor signs)
- Ataxia of limbs or gait
- Cortical signs (aphasia, neglect, apraxia…)



Subcortical strokes

- Caused by occlusion of small penetrating branches of cerebral arteries
- Chronic HTN>DM>emboli

Syndrome	Presentation	Localization
Pure Motor	Face, arm, leg	Internal capsule
Pure Sensory	Face, arm, leg	Thalamus
Sensorimotor	Face, arm, leg	Thalamocapsular
Ataxic-hemiparesis	Hemi-ataxia & hemiparesis	Basis pontis
Clumsy hand dysarthria	Dysathria, incoordination	Genu of the IC



Acute stroke syndrome

- A-B-C's
  - NPO, intubate for inadequate airway, ventilate if needed
  - Correct hypotension, rule out acute MI or arrhythmia (a-fib)
- Rule out hypoglycemia
  - Blood glucose is between 50 and 400 mg/dl
  - Minimize hyperglycemia by running an IV of 0.9% normal saline initially at a TKO rate
- Use parenteral antihypertensive Tx only for sustained, very high BP (>220/120; or >185/110 for IV tPA)
  - BP maintained under 185/110

- IV tPA must be/may be given within 4.5 hrs of stroke onset
- Neuro deficit (NIHSS score 5 to 22) must not be rapidly improving (TIA) or post-ictal
- Normal PTT, PT≤15 sec, platelets ≥100,000
- No bleeding, recent surgery, MI, arterial puncture or LP
- No blood, or edema/infarct > 1/3 of MCA territory on CT

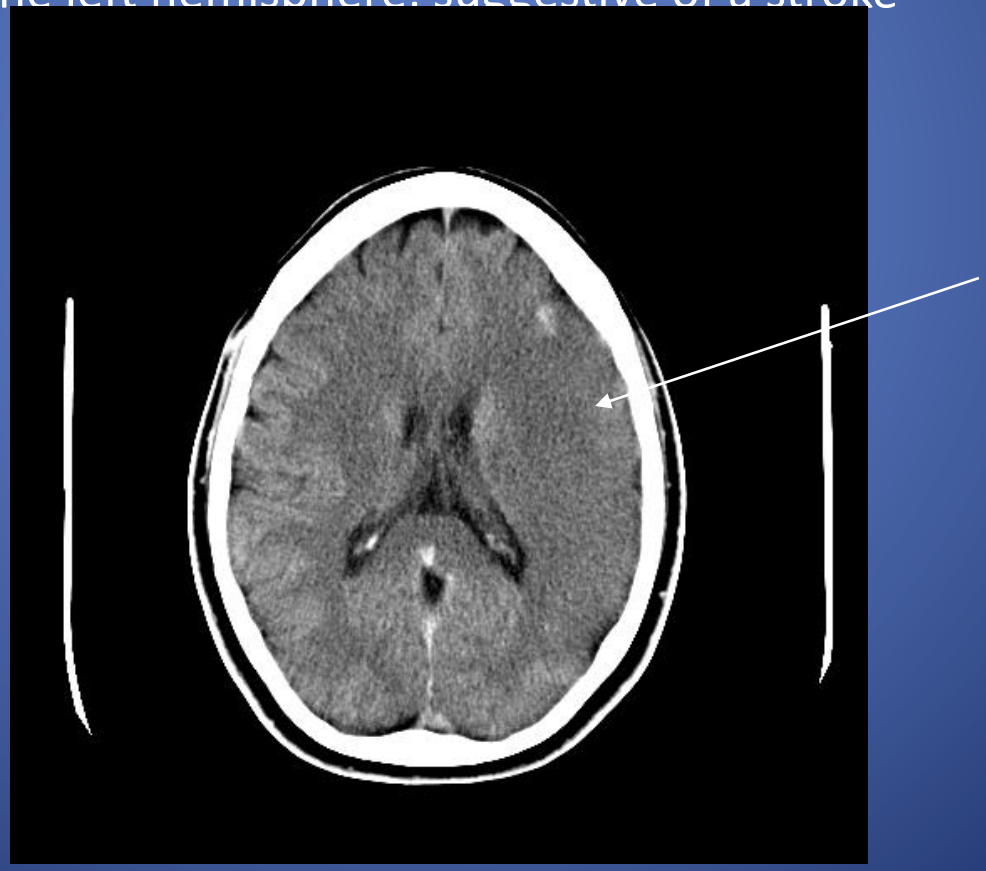
TIME IS BRAIN!!!

Diagnostic tests

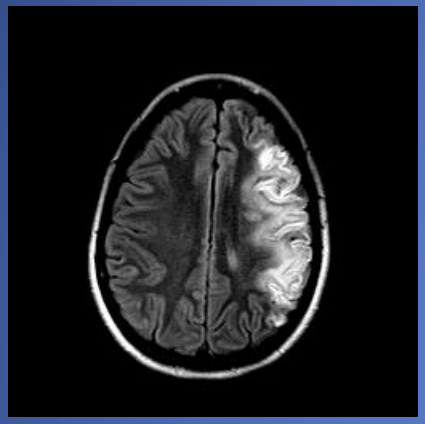
- What imaging test do you want to order?
- CT scan
  - in deteriorating patient, quickly rules out hemorrhage, mass (tumor, abscess) or early infarct edema
  - shows cortical infarcts by 1-2 days, may miss lacunar infarcts
- MRI scan
  - highest resolution scan, but longer scanning time
  - DWI (diffusion weighted imaging) detects impaired movement of water in infarct immediately
  - non-invasively view arterial supply (MRA)
  - contraindications: pacemaker

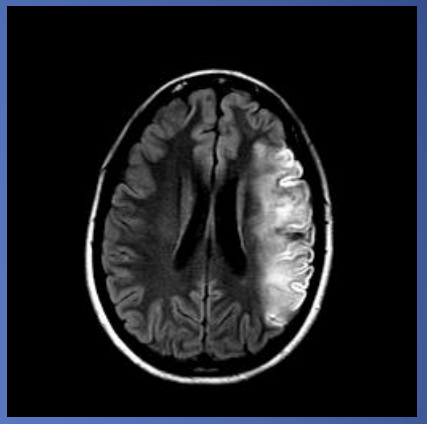
Non Infused Head CT

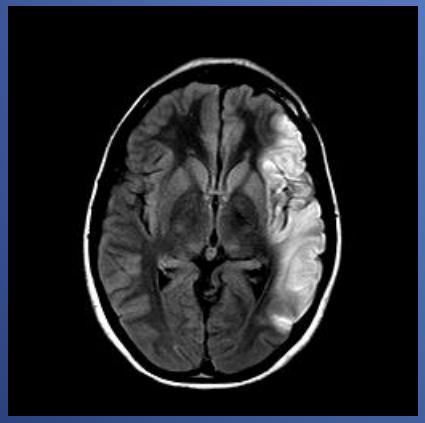
- What is your impression of the head CT?
  - The CT shows loss of gray-white junction and effacement of the sulci throughout the left hemisphere, suggestive of a stroke

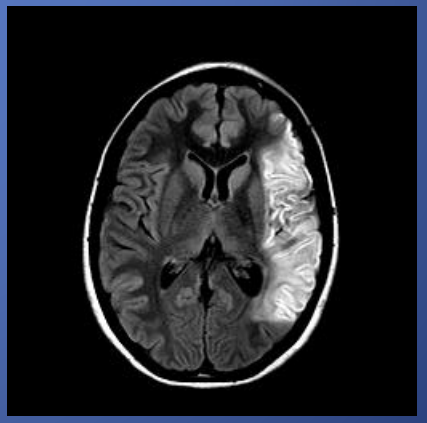


MRI (Diffusion Weighted Image)







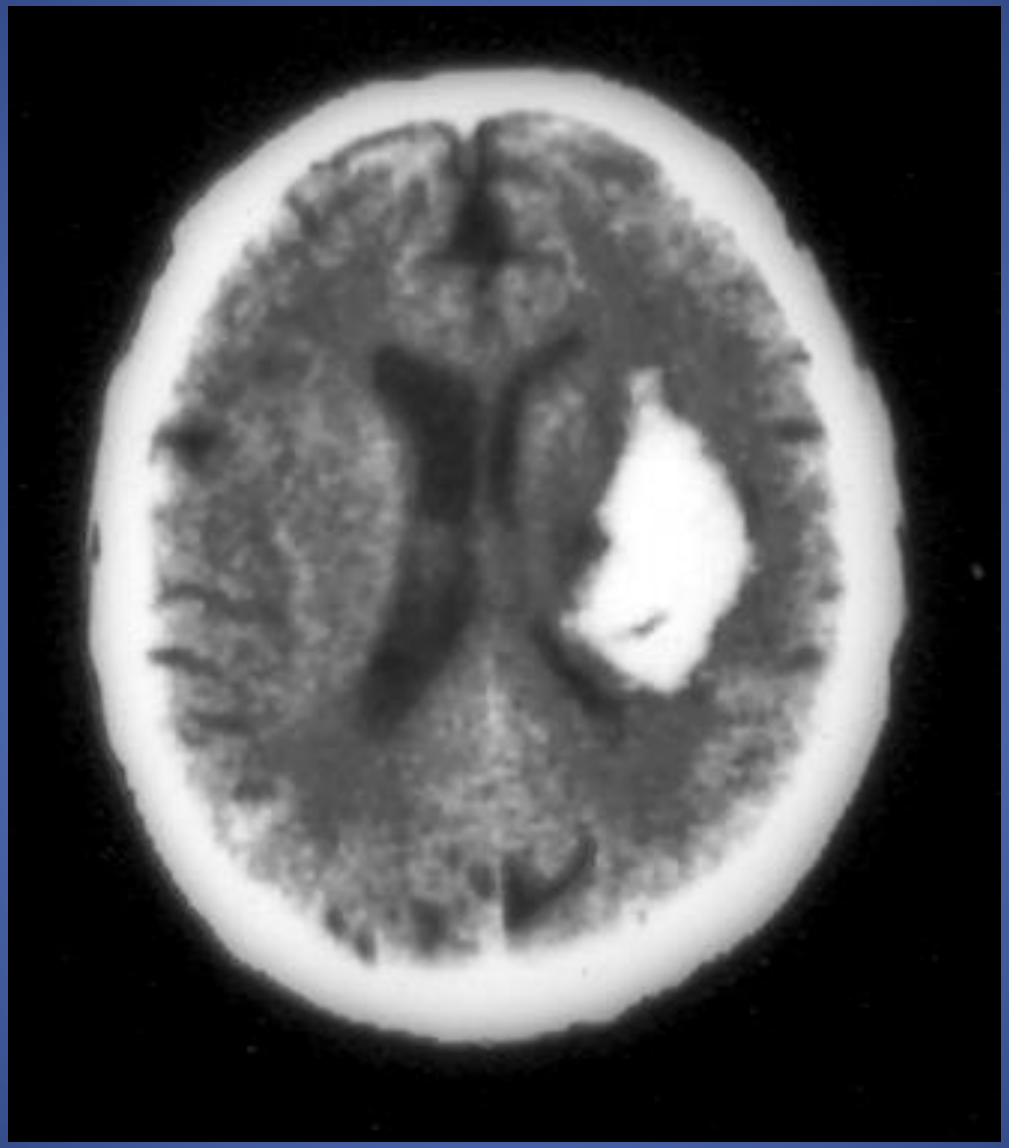


Acute stroke in younger patients

- Arterial
  - Dissection (spontaneous, traumatic)
  - Fibromuscular dysplasia, Marfan's, vasculitis
  - Vasoactive drugs: cocaine, amphetamines
  - Migraine
- Paradoxical cardiac embolus (PFO)
- Hypercoagulable states

Case Hong

- A 75 year-old man is verbally unresponsive in the ER, moving only his left limbs to painful stimuli. BP is 210/106, pulse 85/min and afebrile.
- He gradually becomes less responsive.
- What do you do?



Intracranial hemorrhage

- Approximately 10% of all strokes
- Most common cause hypertension
- Lobar hemorrhage: consider anticoagulation, amyloid angiopathy, AVMs, cavernous malformations, metastatic or primary tumors
- Neurologic deterioration primarily due to hematoma expansion and worsening cerebral edema
- Treatment: maintain airway/treat hypoxia, cardiac monitoring, avoid hypotonic solutions, treat glucose disorders, treat seizure, generally treat MAP > 150 mmHg (gradual reduction)

Increased intracranial pressure

- General medical treatment of increased ICP:
  - ICP monitoring
  - Hyperventilation (pCO_2 < 33 mm) – vasoconstriction with reduction of blood volume; aggressive hyperventilation may cause worse outcome
  - Hyperosmolar therapy: mannitol 20% (0.25 gm/kg q6 hrs if S_{osm} <310)
- Specific treatment of increased ICP:
  - CSF drainage
  - Surgical evacuation of hematoma
  - Tumor, encephalitis, abscess (vasogenic edema): dexamethasone 4 mg IV q6 hrs