Project: Ghana Emergency Medicine Collaborative

Document Title: Pediatric Neurologic Emergencies

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Pediatric Neurologic Emergencies

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Washington University in St. Louis

Objectives

- General pediatric neurologic assessment
- Nonsurgical neurological emergencies
- Neurosurgical nontraumatic emerencies
- Neurosurgical traumatic injuries
- Initial stabilization and emergency management

General Approach

Symptoms

- Headache
- Changes in mental status
- Weakness (focal or generalized)
- Paresthesias
- Vomiting
- Visual changes
- Difficulty with speech
- Difficulty walking

History

- Trauma
- Ingestion
- Exposure history
- Fever/Infection
- Onset of symptoms acute or subacute?

Pediatric Neurologic Exam

- Depending on age, may not be able to go head-to-toe like adults
- Starts with initial observation, how they play and interact
  - Look at the face to see if there is any facial droop
  - Look for dysmorphic features or skin findings
- Use toys/shiny objects, test how they track for extraocular movements
  - Bring objects into their peripheral vision for visual fields

Pediatric Neurologic Exam

- Motor – watch for movement and use of all extremities
  - Any muscle atrophy
- Sensory – can test like adult after 5-6 years of age
- Tone
  - Abduction of hips at rest could suggest hypotonia (decreased muscle tone)
  - Persistent arching of back and neck could indicate hypertonia

Hypotonia (decreased muscle tone)

PD-INEL

Janelle Aby, Stanford University School of Medicine

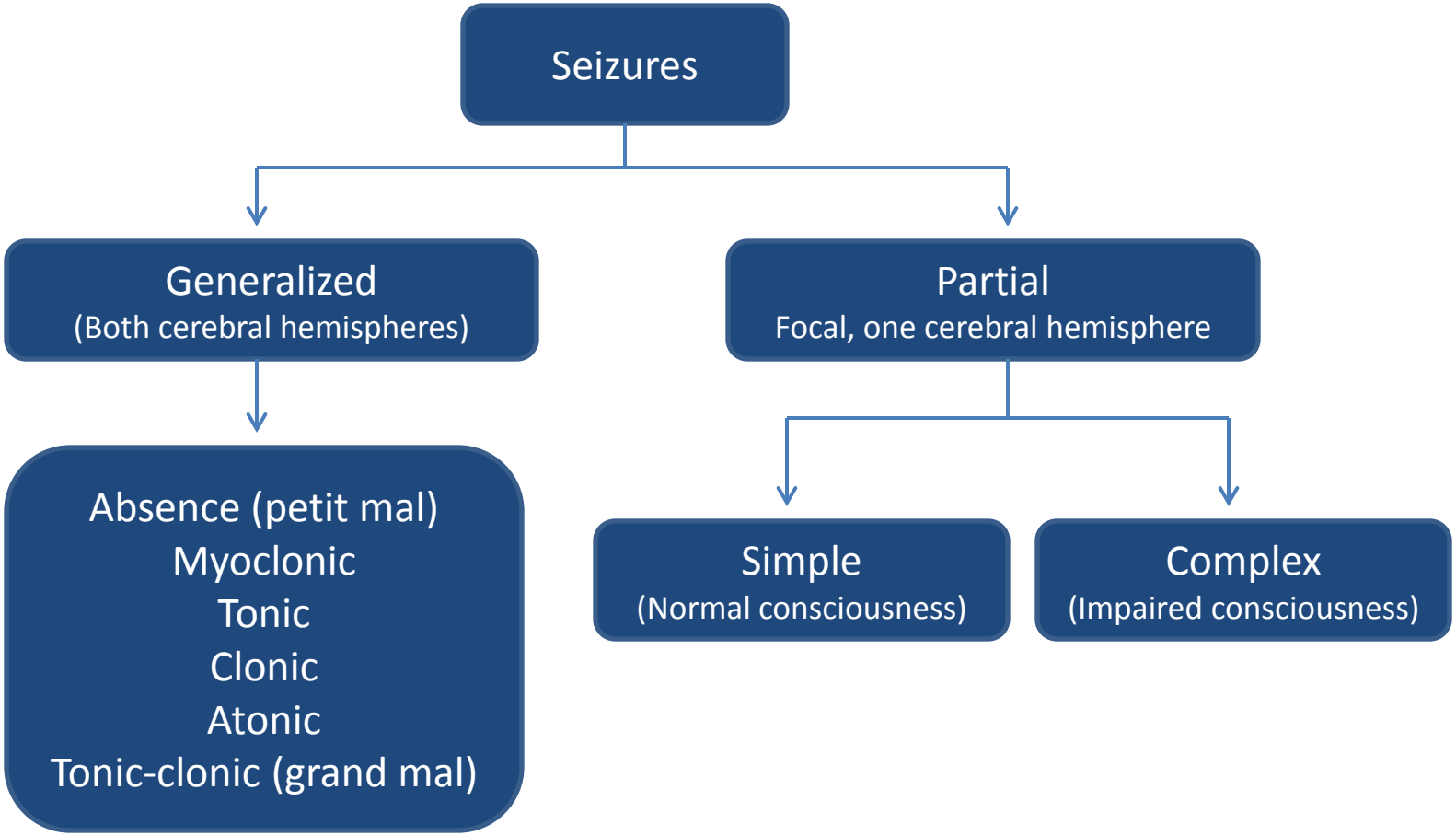
Glasgow Coma Scale

	Adult/Child	Infant	Score
Eye Opening	Same	Same	
Best Verbal	Oriented, appropriate	Coos and babbles	5
Response	Confused	Irritable cries	4
	Inappropriate words	Cries to pain	3
	Incomprehensible sounds	Moans to pain	2
	No response	No response	1
Best Motor	Obeys commands	Moves spontaneously	6
Response	Localizes to pain	Withdraws to touch	5
	Withdraws to pain	Withdraws to pain	4
	Flexion to pain	Flexion to pain	3
	Extension to pain	Extension to pain	2
	No response	No response	1

Seizures: The Terminology

- Cryptogenic (or idiopathic) = no known precipitating factor
- Provoked seizure = a precipitant can be identified
- Status epilepticus = prolonged seizure longer than 30 minutes (5 minutes) or recurrent seizures and patient does not regain consciousness in between

Categorizing a Seizure



Case 1

A mother runs into the emergency department holding her 3 year-old son in her arms. He has mostly jerking of his right side with his eyes also going to the left. He is foaming at the mouth. It has been going on for 15 minutes, and his lips are slightly blue. What do you do first?

A. Give diazepam 0.5 mg/kg rectally
B. STAT head CT
C. Suction his mouth and put him on oxygen
D. Start an IV

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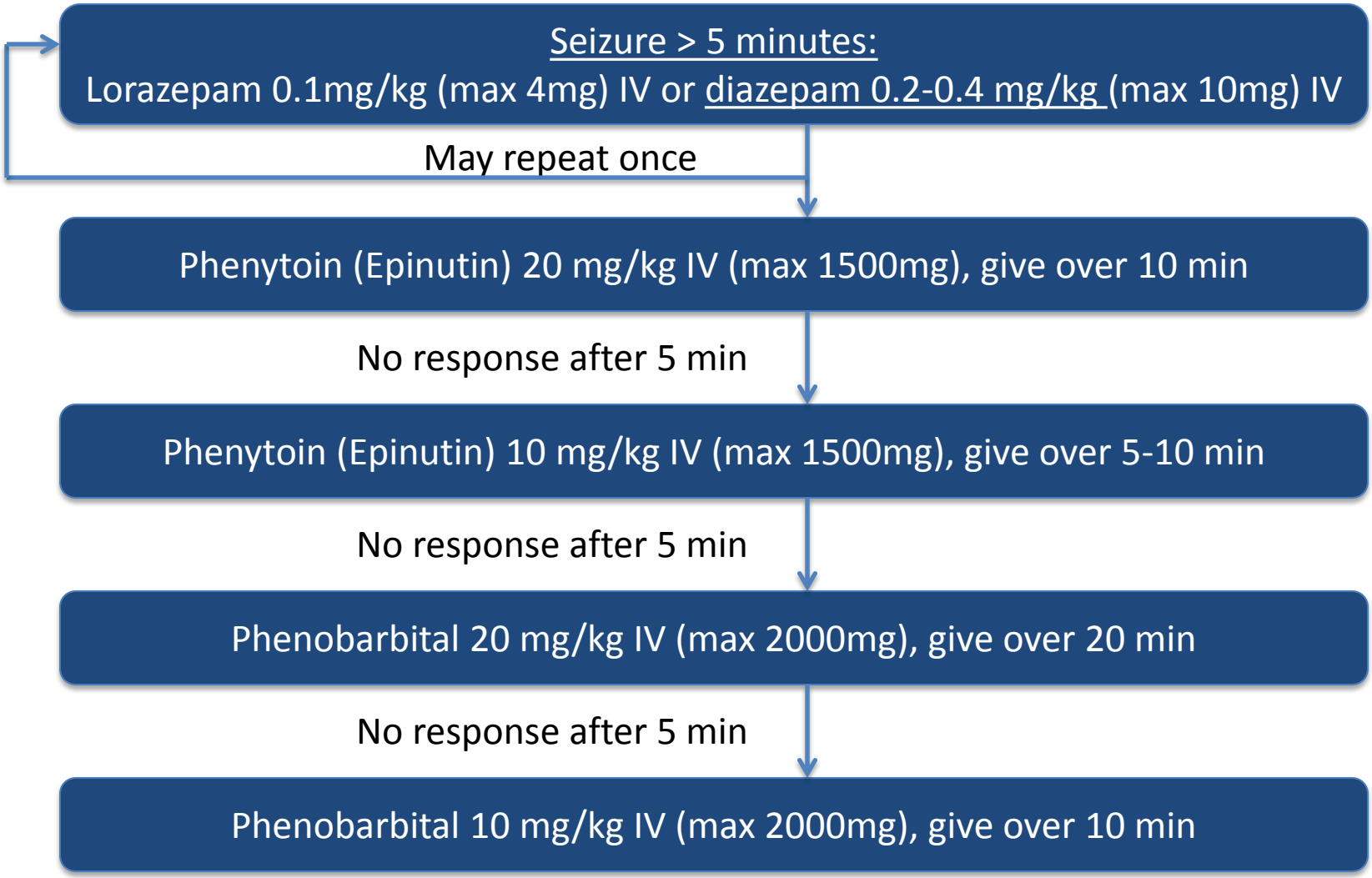
Seizure Management

- ABC!!!
  - Airway: clear secretions, place nasal or oropharyngeal airway if obstructing
  - Breathing: place patient on oxygen
  - Circulation: obtain IV or IO access
- Laboratory studies
  - Fingerstick blood sugar
  - Electrolytes (Na, K, Ca, Mg, P)
  - Full blood count (FBC)
  - Other tests: liver function panel, ammonia, toxicology screens, antiepileptic drug levels, lumbar puncture

Acute Treatment of Seizure



Acute Treatment of Seizure

- If refractory:
  - Continuous infusion of midazolam
- Evaluate the airway continuously
  - If need to intubate, prefer shorter-acting paralytic such as succinylcholine if no contraindications
  - Use end tidal carbon dioxide monitoring and pulse-ox if available

What are your options if you have no IV access?

No IV Access

- Diazepam 0.5 mg/kg (max 20mg) per rectum
- Midazolam 0.2 mg/kg (max 7mg) intramuscularly
- Midazolam 0.2 mg/kg (max 10mg) intranasally

Unresponsive Seizures

- Hypoglycemia = 40 mg/dL or 2.2 mmol/L
  - 25% dextrose water (D25W) 2-4 ml/kg or for infants 10% dextrose water (D10W) 5-10ml/kg
  - Give dextrose until asymptomatic
- Hyponatremia = serum sodium (Na) < 135 mEq/L
  - Severe hyponatremia = Na < 120 mEq/L
  - 3% sodium chloride (NaCl) 2-5 ml/kg
  - Correct sodium acutely until seizures stop

Further Testing

- Emergent noncontrast head CT if:
  - Prolonged seizure
  - Focal neurological exam
  - Concern for trauma or hemorrhage
  - Prolonged decreased level of consciousness
  - Otherwise could wait for brain MRI if needed
- Lumbar puncture for concerns of meningitis or encephalitis
- Look for any signs of ingestion

Febrile Seizures

- 2-5% of children, most common convulsive disorder in young children
- Seizure occurring at 6 months to 5 years of age associated with fever
- Simple febrile seizure (85%) = generalized, once over 24 hours, less than 15 minutes
- Complex febrile seizure (15%) = focal, recur within 24 hours, greater than 15 minutes

Febrile Seizures

- 33% of children have one recurrence, 9% have ≥ 3 episodes
  - Increased risk of recurrence if younger and had lower fever with first seizure
- < 5% develop epilepsy
- No laboratory studies required except to find source of infection
  - Lumbar puncture if less than 6 months
  - Consider lumbar puncture if 6-12 months, pretreated with antibiotics, complex febrile seizure, altered mental status, signs of meningitis or encephalitis
- Generally do not need head CT

Stroke

Stroke: Pathophysiology

- Ischemic or embolic stroke
- Hemorrhagic stroke
  - 30-60% of strokes in children vs. 20% in adults are hemorrhagic

Ischemic Stroke

- 50% of children with ischemic stroke have underlying risk factor, most commonly congenital heart disease
- 94% present with hemiplegia
- Other clinical manifestations:
  - Hemianopsia
  - Dysphagia
  - Vertigo, ataxia
  - Headache in older children
  - Seizures as presenting symptom in younger children

Hemorrhagic Stroke

- Pathophysiology
  - Intraparenchymal hemorrhage
  - Nontraumatic subarachnoid hemorrhage, most often from intracranial aneurysm
- Most commonly presents with headache, altered level of consciousness, and vomiting

Stroke Management

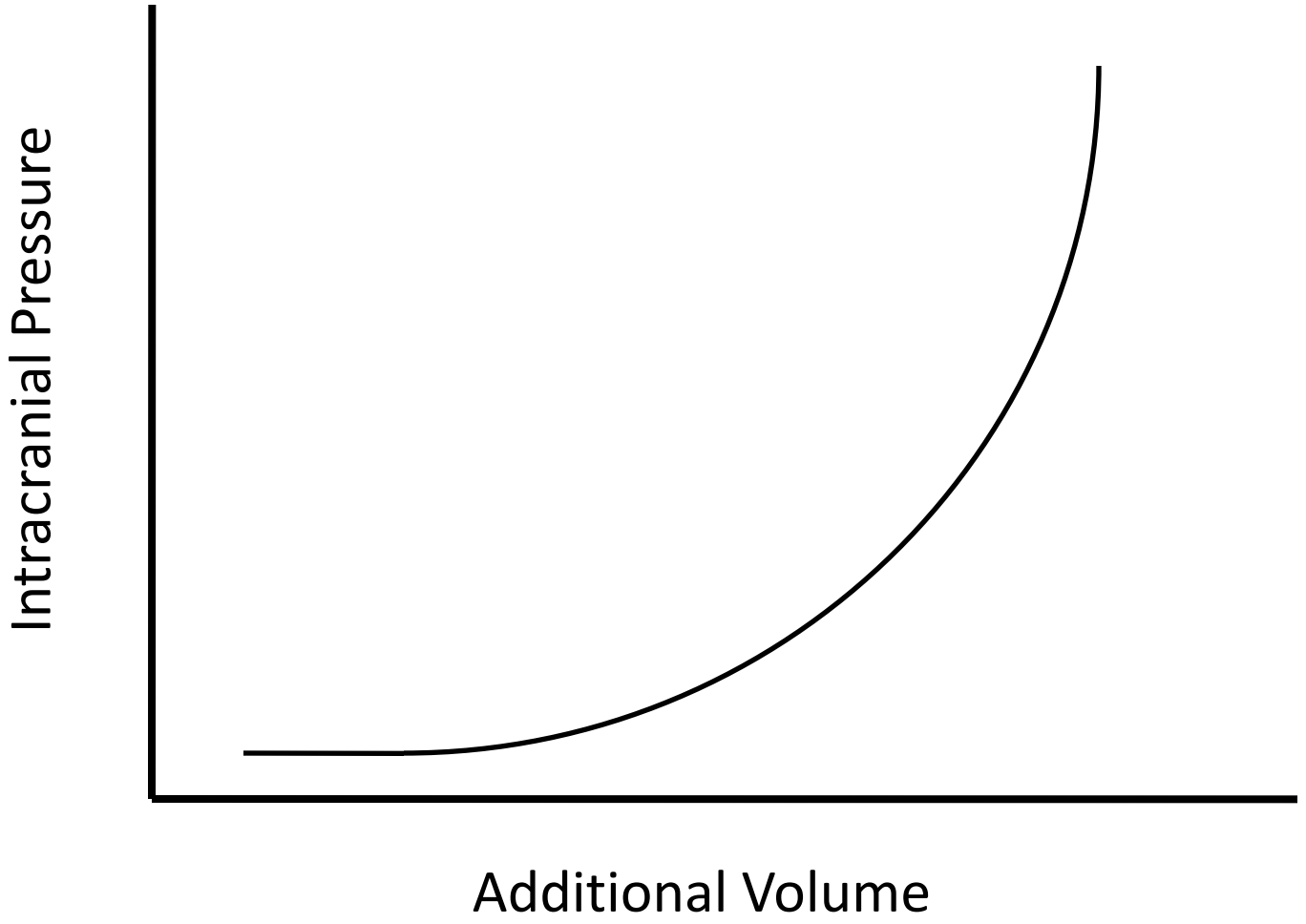
- Noncontrast head CT to rule out hemorrhage
- Manage blood pressure (beware of hypotension)
- Ischemic stroke
  - Anticoagulate with LMWH (enoxaparin) or heparin
  - Thrombolytics untested in children
  - Exchange transfusion if have sickle cell disease
  - Cardiac echo
- Hemorrhagic stroke
  - Correct any coagulation defects
  - Consult neurosurgery if rapidly expanding hematoma needs evacuation

Concerning Headaches

Increased Intracranial Pressure

- Cerebral perfusion pressure (CPP) = mean arterial pressure (MAP) – intracranial pressure (ICP)
  - Increase in ICP leads to decrease in CPP
- The noncompliant cranium can only accommodate a certain volume of intracranial contents and then have exponential increase in pressure
  - Can have delayed reaction in infants with open sutures and fontanelles

Effect of Increased Volume on Intracranial Pressure



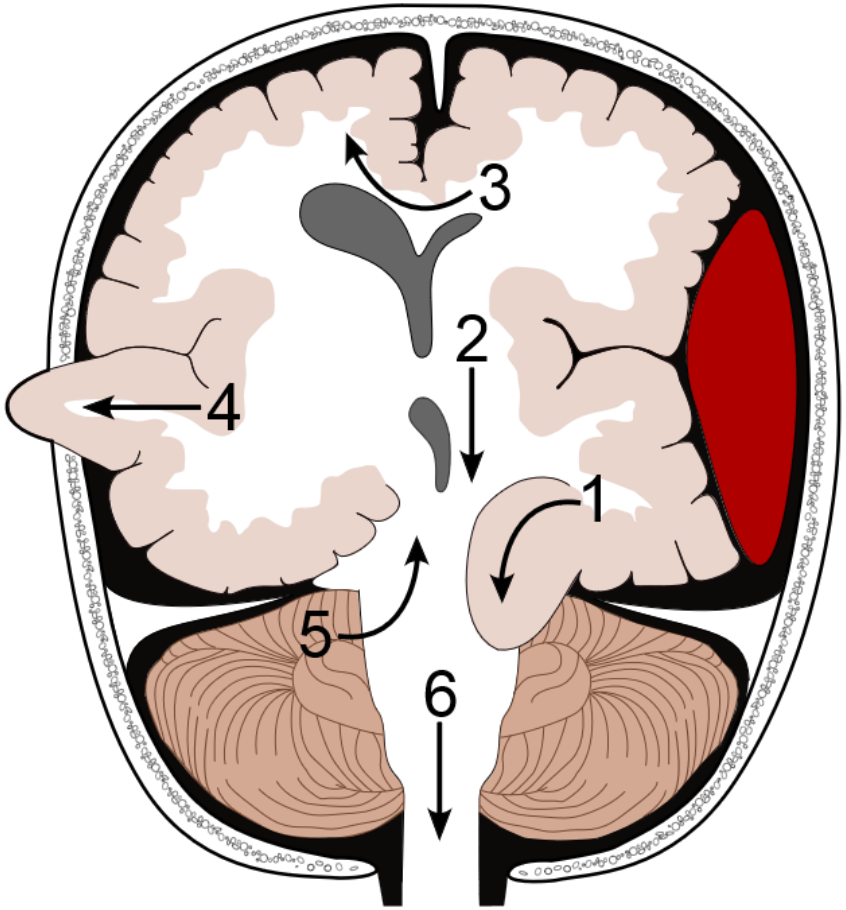
Causes of Increased Intracranial Pressure

- Tumors
- Increased cerebral spinal fluid production or decreased reabsorption
- Intracranial hemorrhage
- Cerebral edema
- Increased ICP will decrease blood flow to brain, leads to cerebral ischemia and secondary injury

The eventual result of increased intracranial pressure is herniation

Tentorial Herniation (1,2)

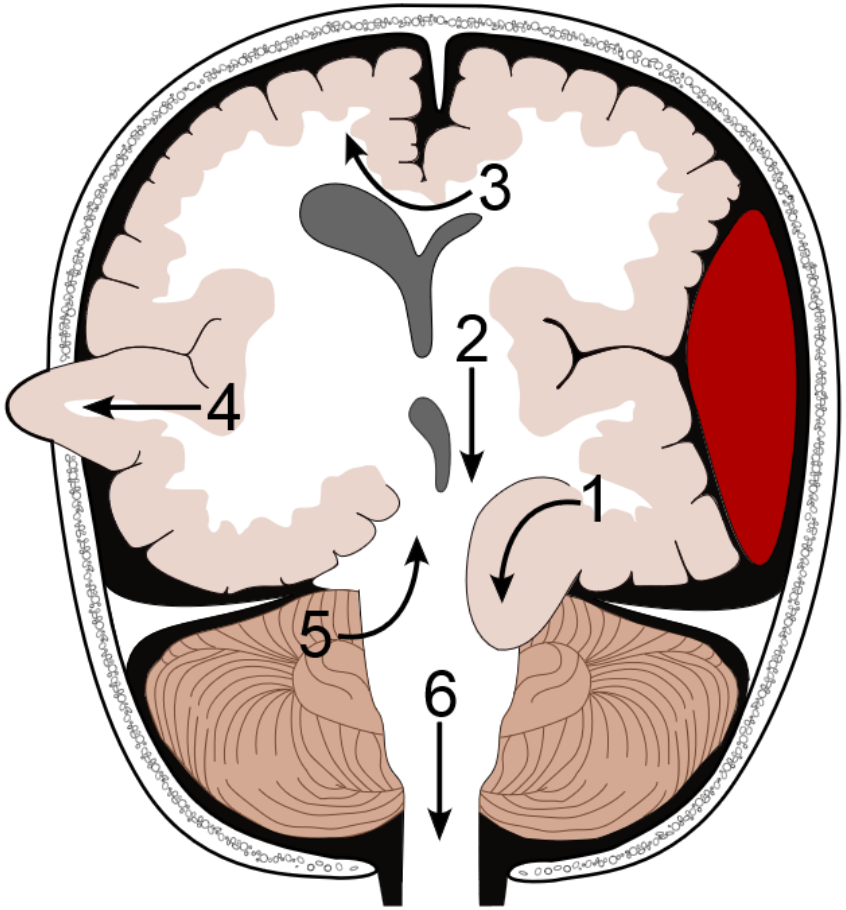
- Herniation of parahippocampal gyrus and uncus of temporal lobe through tentorial notch
- Temporal cortex presses against brainstem and cranial nerve III
- Headache, decreased mental status, blown pupil, ptosis, loss of medial gaze, decerebrate posturing
- Cushing's triad as brainstem compressed -> respiratory arrest



Brain_herniation_types.svg (Wikimedia Commons)

Tonsillar/Cerebellar Herniation (6)

- Cerebellar tonsils herniate through foramen magnum
- Usually from posterior fossa mass lesion
- Compresses aqueduct of Sylvius and causes hydrocephalus
- Neck pain, vomiting, decreased mental status, bradycardia, hypertension



General Management of ICP

- ABCs
  - May need to intubate if GCS ≤ 8, no gag, poor ventilation
  - Give supplemental oxygen
  - If normotensive, avoid excess fluid administration
- Once intubated, sedation if agitated to prevent spikes in ICP
  - Propofol would not be ideal due to risk of hypotension
- Head CT if signs of increased intracranial pressure

Management of ICP

- Elevate head of bed to 30 degrees to promote venous drainage of head
- Hyperventilation with PCO2 of 30-25
  - Hypocarbia leads to reflex vasoconstriction
  - Caution: if overhyperventilate, can cause excess vasoconstriction and decreased blood flow

Case 2

A 6 year-old boy is brought in after a car accident. He was unrestrained and flew into the front windshield. On arrival, vital signs are HR 68, BP 80/50. He has decorticate posturing and blown right pupil. You intubate him and elevate the head of the bed to 30 degrees. What is the best thing to do next?

A. Give mannitol 1gm/kg

B. Give 3% NS 3ml/kg

C. Hyperventilate

D. Take the child for emergent head CT

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Mannitol vs. Hypertonic Saline

IV Mannitol (0.5 to 1gm/kg)

- Increases serum osmolarity and draws free water into vasculature
- Blood less viscous so improves cerebral blood flow
- Onset in a few minutes
- Osmotic diuretic so will also lead to volume depletion

3% saline (2-5 ml/kg)

- Can also be given as a continuous drip
- Hyperosmolarity leads to decreased blood velocity and improved cerebral blood flow
- Does not have diuretic effect
- Preferred if not hemodynamically stable

Specific Causes of ICP

Brain Tumors

- Account for 20% of all cancers in children, second to leukemia
- Most commonly gliomas/astrocytomas in children
- Majority are located in the posterior fossa
- Ominous signs associated with headaches
  - Persistent vomiting
  - Vomiting on awakening

Brain Tumor Management

- Noncontrast CT if patient unstable
  - Can evaluate for hemorrhage, tumor-related obstructive hydrocephalus, mass effect
- If patient is stable, brain MRI is study of choice
  - More sensitive for identifying small brain tumors
  - Better visualization of the posterior fossa
- Neurosurgery consult

Hydrocephalus

- Due to either increased cerebral fluid production or decreased absorption
  - Noncommunicating hydrocephalus – CSF in the ventricular system is blocked by a congenital or an acquired defect
  - Communicating hydrocephalus – block in absorption at the meningeal surfaces
- Present with increasing head circumference, vomiting, sleepiness, irritability, downward deviation of the eyes ("sunsetting"), ataxia
- Noncontrast head CT -> dilated ventricles
  - Can do head ultrasound if fontanelle is open
- Neurosurgery consult for CSF shunt

Shunt Malfunction

- Many different types of shunts
  - All contain a ventricular catheter, one-way valve, and distal tubing into peritoneal cavity, pleural cavity, or right atrium
  - Some have reservoir available for percutaneous tapping
- Risk of shunt failure is highest first months after placement
  - 40% fail in first year, 80% by 10 years
- Evaluate for malfunction using head CT to evaluate ventricles and shunt series to evaluate shunt tubing
- If emergent, can try to access the shunt, but for definitive care, need neurosurgery consult for revision

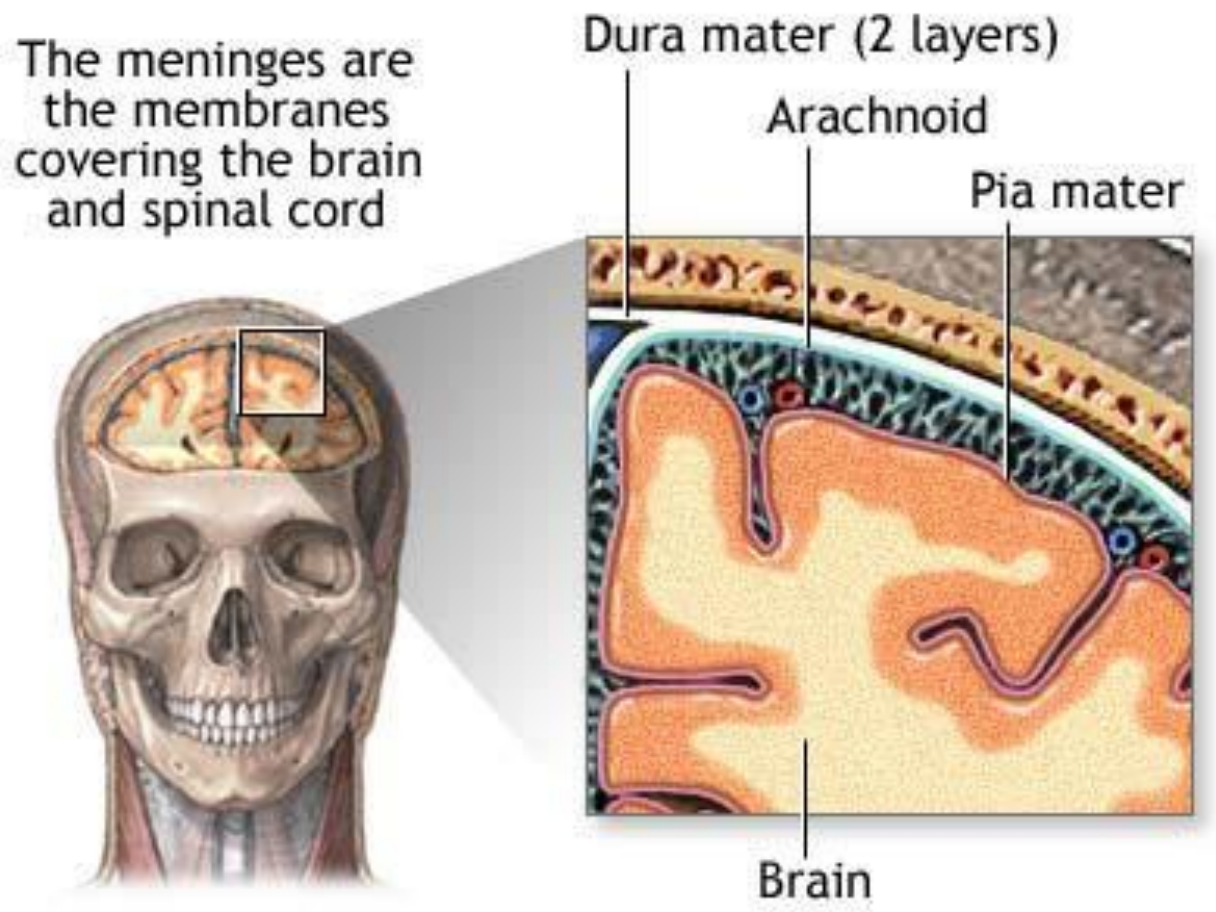
Traumatic Brain Injury

- In United States, 475000 children younger than 14 years old visit the ED annually for head injury
- Traumatic brain injury can be divided into 2 components:
  - Primary brain injury due to the traumatic event itself
  - Secondary brain injury from hypoxia, hypoperfusion, metabolic derangements during resuscitation

Parenchymal Injuries

- Cerebral contusion
  - Common in children due to the movement of a relatively mobile brain within a fixed skull
  - Most common in frontal and temporal lobes
  - Associated with risk of late intraparenchymal hematoma
  - Admit for observation
- Diffuse axonal injury
  - Diffuse primary injury to the white matter tracts of the brain
  - Due to severe acceleration and deceleration or angular forces to the brain
  - Usually from motor vehicle crashes or child abuse

Meninges of the Brain



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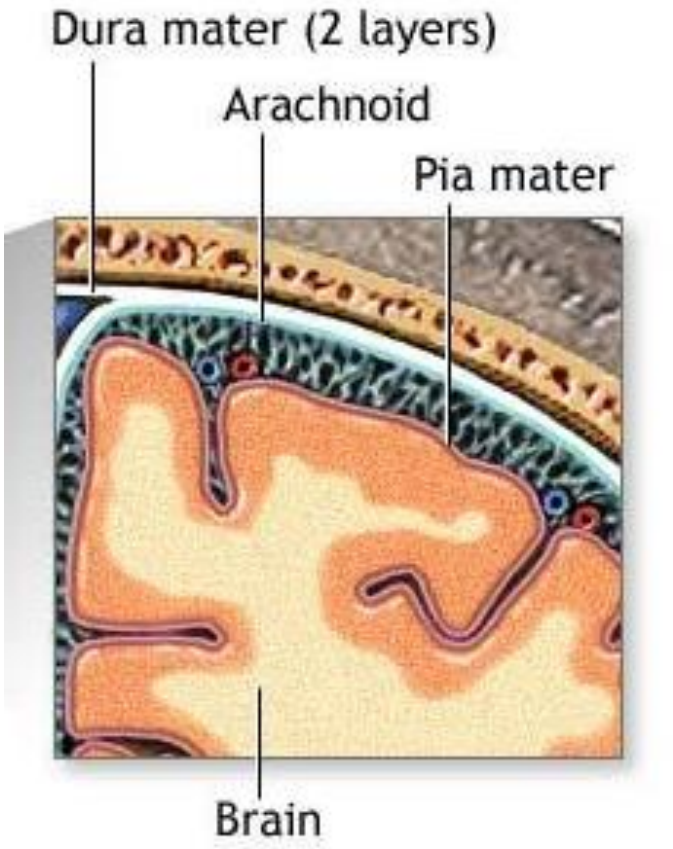
Epidural Hematoma



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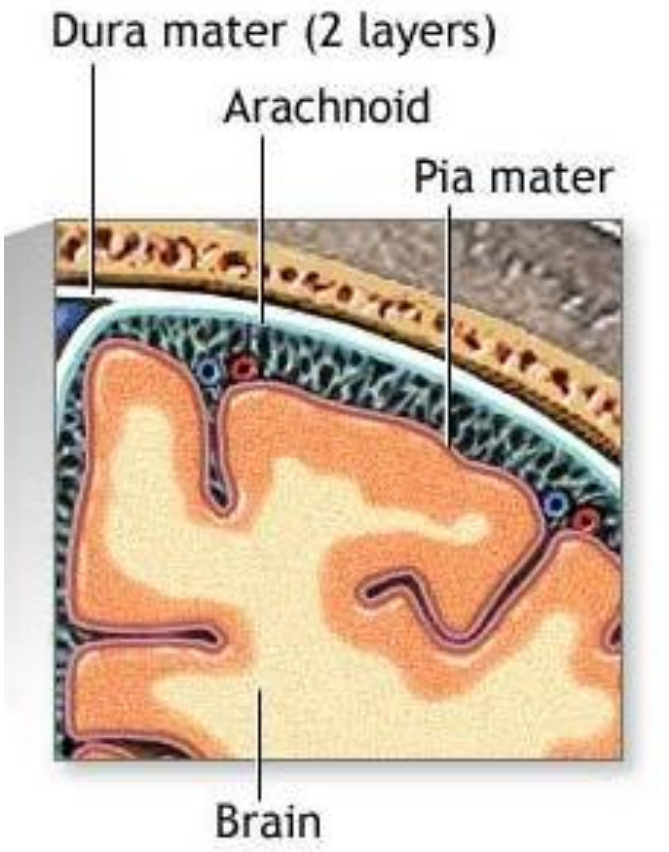
Epidural Hematoma

- Due to blunt impact to the cranium, often with skull fracture with laceration of epidural vessels
- In children, mostly from falls
- Classic presentation has initial loss of consciousness then “lucid interval” of several hours, but children often don’t have this presentation
- Management: craniotomy with drainage of hematoma and repair of lacerated epidural vessels
  - Sometimes observation if < 30 mL in volume or thickness < 2 cm with normal physical exam

Glitzy queen00 (Wikimedia Commons)

Subdural Hematoma



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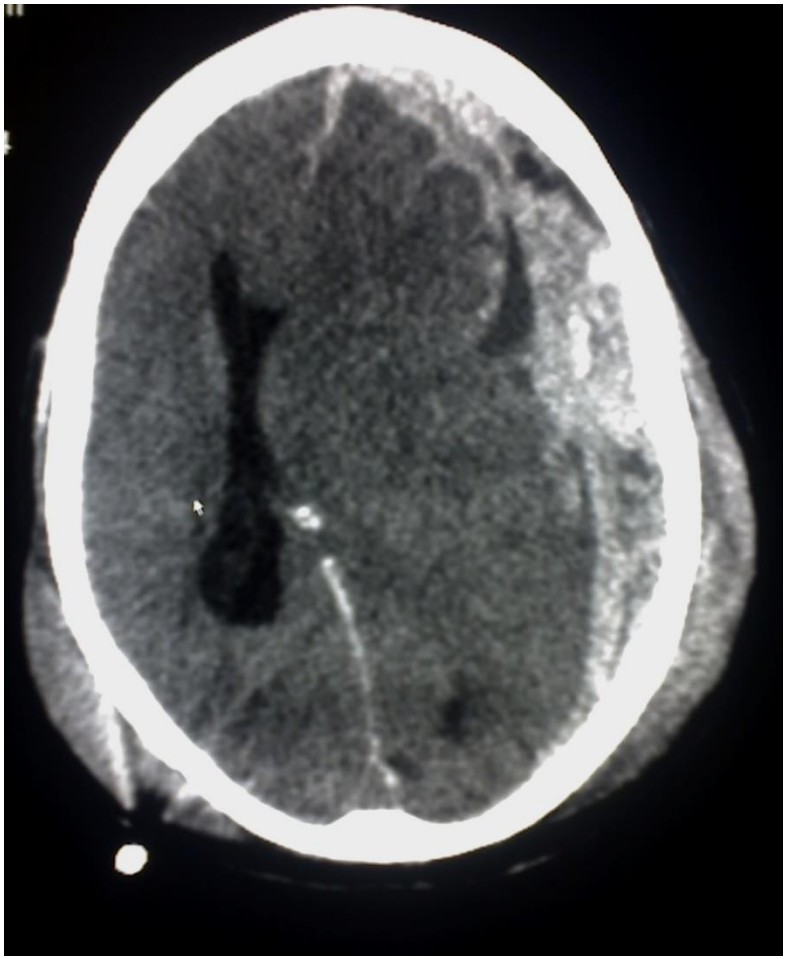

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Subdural Hematoma

- Usually from mechanisms that are associated with shear forces that tear bridging veins
  - In older children, likely MVC
  - Strong association with child abuse in younger children
- Have initial loss of consciousness and decreased mental status
  - Patients who present in coma or pupillary abnormalities have poorer prognosis
- Management:
  - Non-operative if not severely ill
  - Drainage if signs of increased ICP

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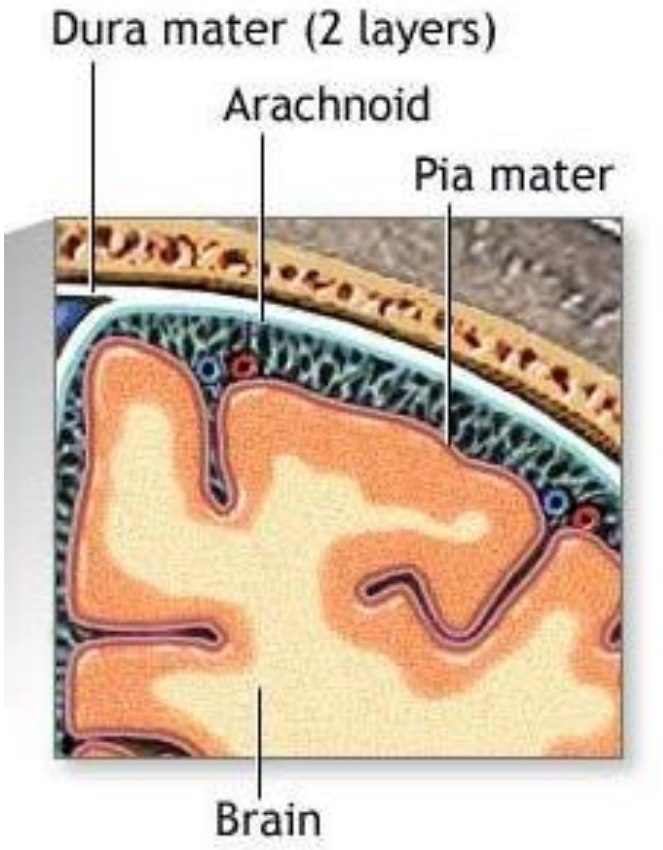
Subarachnoid Hematoma



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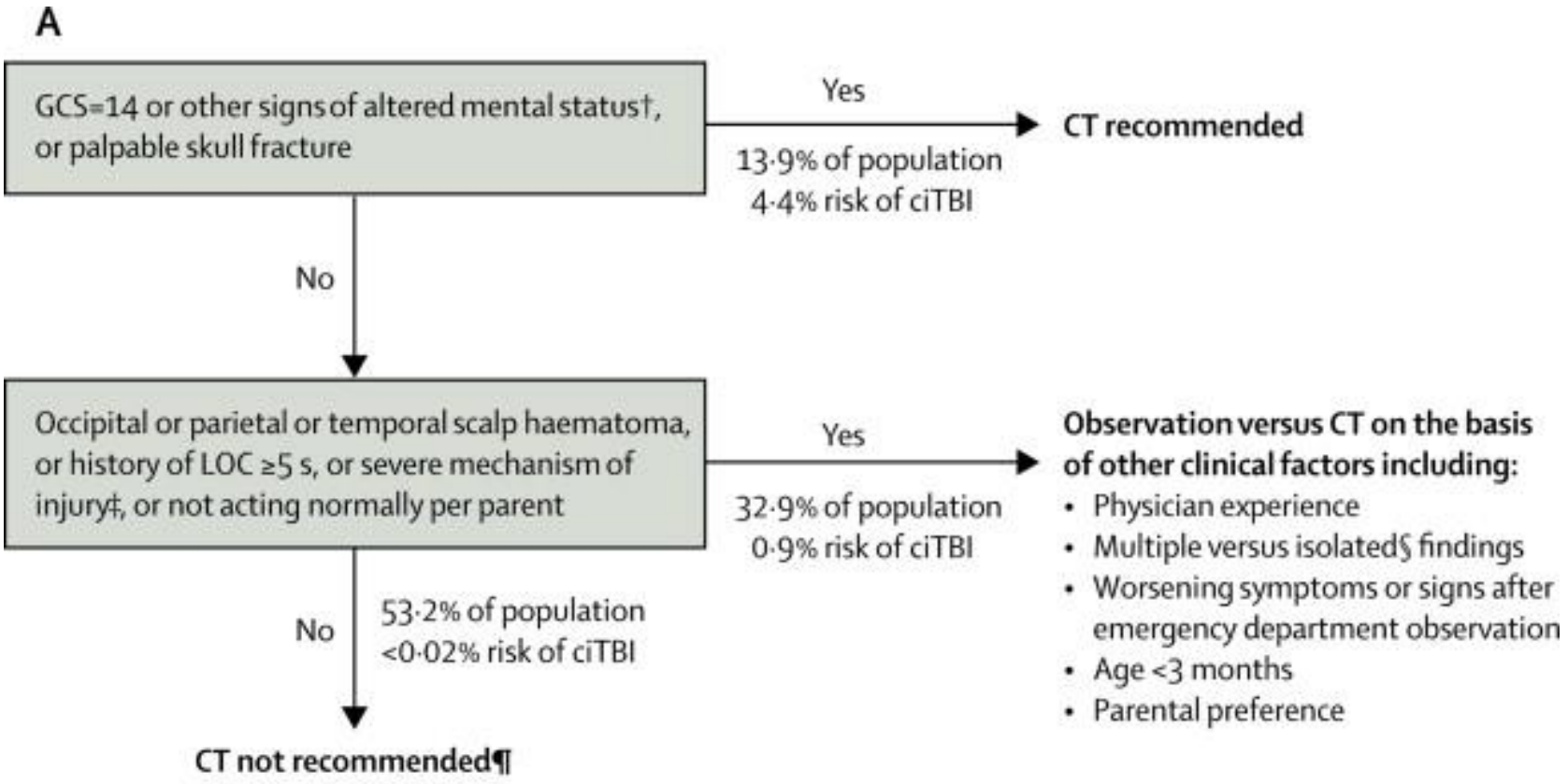
Subarachnoid Hematoma

- Tearing of small vessels in pia mater
- Subarachnoid space is large, so blood in SAH can distribute widely
- Presents with headache and signs of meningeal irritation (nausea, vomiting, neck stiffness)
- Can usually be detected on noncontrast head CT but sensitivity only 90%
- Management: admit for observation, generally non-surgical
  - May need prophylactic anticonvulsant (Epinutin, Levetiracetam)

Who gets a head CT?

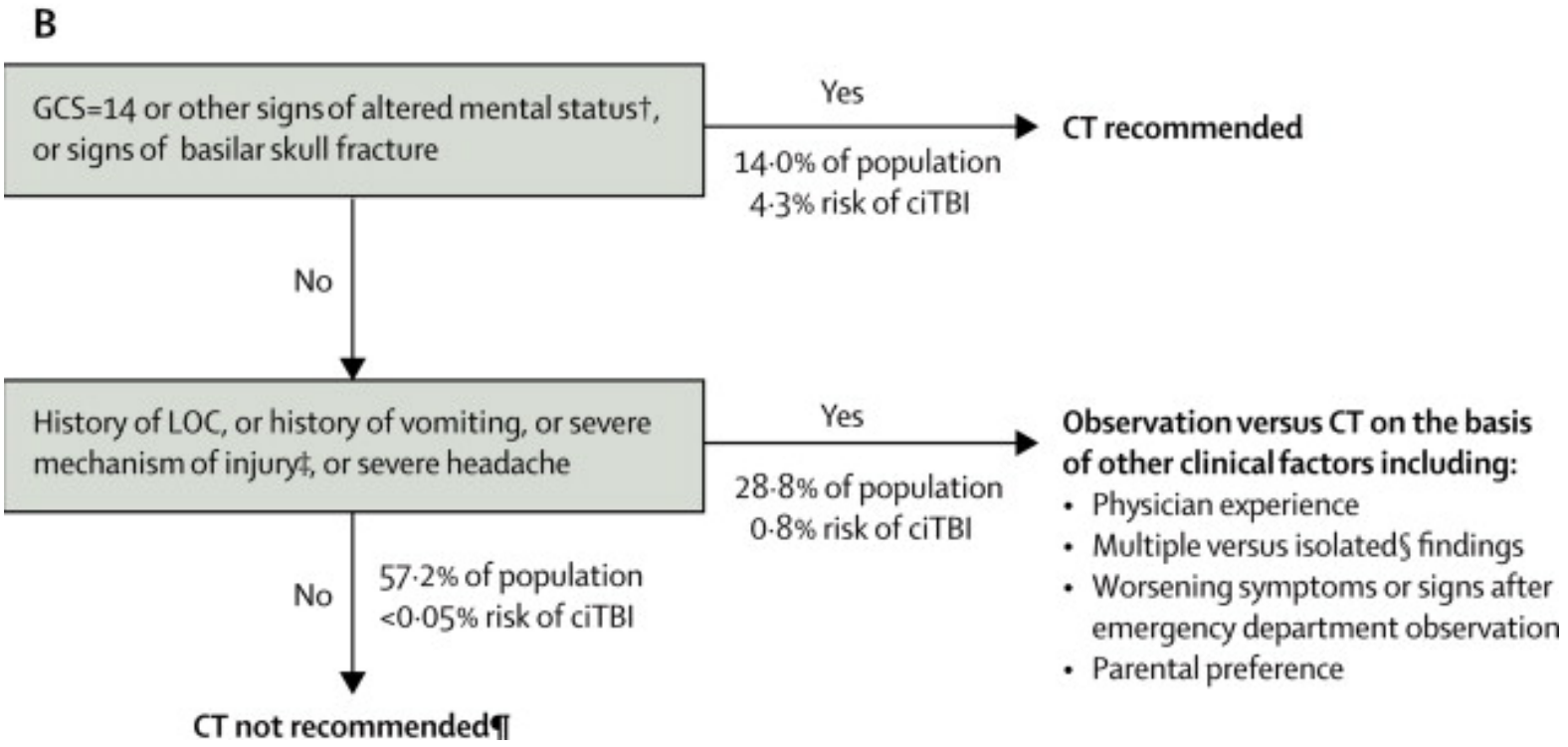
- Can be difficult to decide in children with minor head trauma (GCS of 14-15)
- Lethal malignancy from pediatric head CT is between 1 in 1000 and 1 in 5000, with higher risk at a younger age
- Prospective multi-center study involving 42412 patients used to develop a prediction tool (Kuppermann *et al*, 2009)

Head CT algorithm for children < 2yo and GCS 14-15



 Kupperman N, et al. *Lancet* 2009.

Head CT algorithm for children ≥ 2yo and GCS 14-15



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Disorders of motor function or weakness

Guillain-Barré

- Acute inflammatory demyelinating polyradiculoneuropathy
- The most common cause of motor paralysis in children
  - Uncommon prior to 3 year old
- Antecedant viral infection triggers inflammation and demyelination from autoimmune process
  - Adenovirus, Ebstein-Barr virus, cytomegalovirus, human immunodeficiency virus, varicella-zoster virus, vaccines, *Mycoplasma pneumoniae* and *Campylobacter j*ejuni

Guillain-Barré Signs and Symptoms

- Weakness
- Leg and back pain
- Abnormal gait in younger children
- Sensory loss
- Decreased deep tendon reflexes
- Bowel or urinary incontinence
- Autonomic dysfunction (hypotension)
- Cranial nerve involvement in 30-40% of cases
- Respiratory paralysis occurs in 20-30%
- Can progress for days to weeks

Guillain-Barré

- Diagnosis:
  - Cerebrospinal fluid has elevated protein without elevated white count (pleocytosis)
- Management:
  - Hospitalized and observed due to potential respiratory compromise
  - Generally self-limited, 90% have complete or near-complete recovery
  - Consider plasmapheresis and IVIG in the more severely effected children

Myasthenia Gravis

- Due to autoantibody against acetylcholine receptor proteins
- 3 types:
  - Transient neonatal
  - Infantile
  - Juvenile (most common)
- Mean age of onset 8 years old
- 4:1 females

Myasthenia Gravis

- Clinical manifestations:
  - Mostly affects cranial nerves, ptosis
  - Generalized truncal weakness in 50%, worsens by end of day
- Diagnosis: Tensilon test (edrophonium)
- Management:
  - Check for respiratory compromise, may need ventilatory support
  - Admit if severe respiratory compromise or weakness
  - Treat with Mestinon (cholinesterase inhibitor)

Acute Cerebellar Ataxia

- Most common cause of ataxia in children
- Typically presents 1-4 years of age
- Parainfection or postinfection demyelinization:
  - Most commonly after varicella
  - EBV, enterovirus, HSV, influenza, Mycoplasma, Q fever

Acute Cerebellar Ataxia

- Symptoms 5-10 days after illness onset
  - Acute truncal unsteadiness, possibly some tremors and dysmetria
  - Dysarthria
  - Nystagmus
- Management:
  - Head CT or brain MRI to rule out cerebellar mass
  - Supportive treatment, resolves within 2 weeks

Infantile Botulism

- Due to intestinal colonization of *Clostridium botulinum*
- Toxin impedes acetylcholine release from nerve terminals
- If ingest contaminated honey or poorly canned foods
  - Honey given in United States as dietary supplement, for cough
  - Rare in Africa

Botulism: Clinical Manifestations

- Acute weakness in well infants < 12 mo old
- Constipation, then lethargy and poor feeding
- Poor suck and gag
- Poorly reactive pupils
- Ptosis, facial weakness, oculomotor palsies
- Decreased deep tendon reflexes

Botulism: Management

- Diagnose by identifying *C. botulinum* spores in the stool
  - Have serum assay for toxin but often negative in infants
- Manage airway, may not be ventilating appropriately
  - 60-80% require intubation and mechanical ventilation
- Human-derived botulinum immune globulin (need Center for Disease Control approval)

Spinal Cord Trauma

- Spinal cord injury is rare in pediatrics, approximately 2 of 100,000 children per year
- In children with cervical spine injury, less than 5% are in children less than 2 years old
- Most common cause is motor vehicle crash or being hit by car

The Immature Spine

- Pediatric spine typically matures around 8 years
- Ligaments of the spine are more lax and facet joints are more horizontal, allowing for subluxation of vertebrae
- Paraspinous musculature less developed
- Fulcrum of C-spine is C2-C3 in young children rather than C5-C7
  - Fulcrum migrates to C5-6 by 14 years of age
  - Upper cervical spine more prone to injury

Spinal Cord Injury

- Associated with significant mechanisms of injury, often have evidence of other injuries
- If high cervical cord injury, often have spinal shock (bradycardia and hypotension)
- Suspect whenever have complaints or exam findings of decreased motor strength, decreased or absent reflexes, rectal tone

Risk Factors Associated with C-spine Injury in Children

- Case-control study of children < 16 years old after blunt trauma, included 540 children
- 8 factors associated with C-spine injury (having 1 or more 98% sensitive, 26% specific)
  - Altered mental status
  - Focal neurologic findings
  - Neck pain
  - Torticollis
  - Substantial torso injury
  - Conditions predisposing to cervical spine injury (trisomy 21)
  - Diving
  - High risk MVC

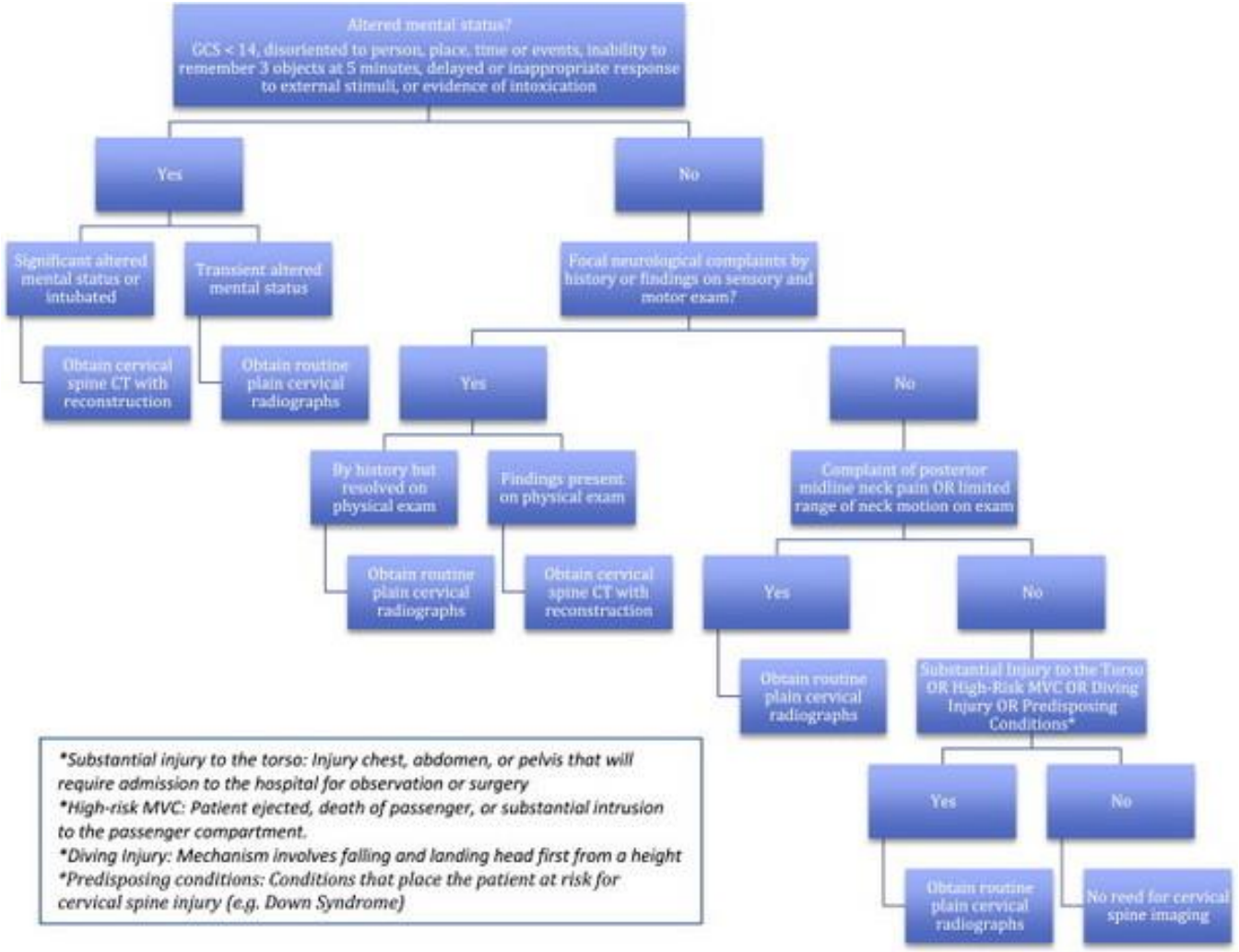
Spinal Injury Management

- ABCs
- Immobilization is mainstay of therapy
  - Use rigid cervical collar and also rigid long board for transport, but take off board as soon as possible
- Consult neurosurgery
  - May need emergent laminectomy if have compressive lesion
- Use of steroids in pediatric spinal cord injury is controversial

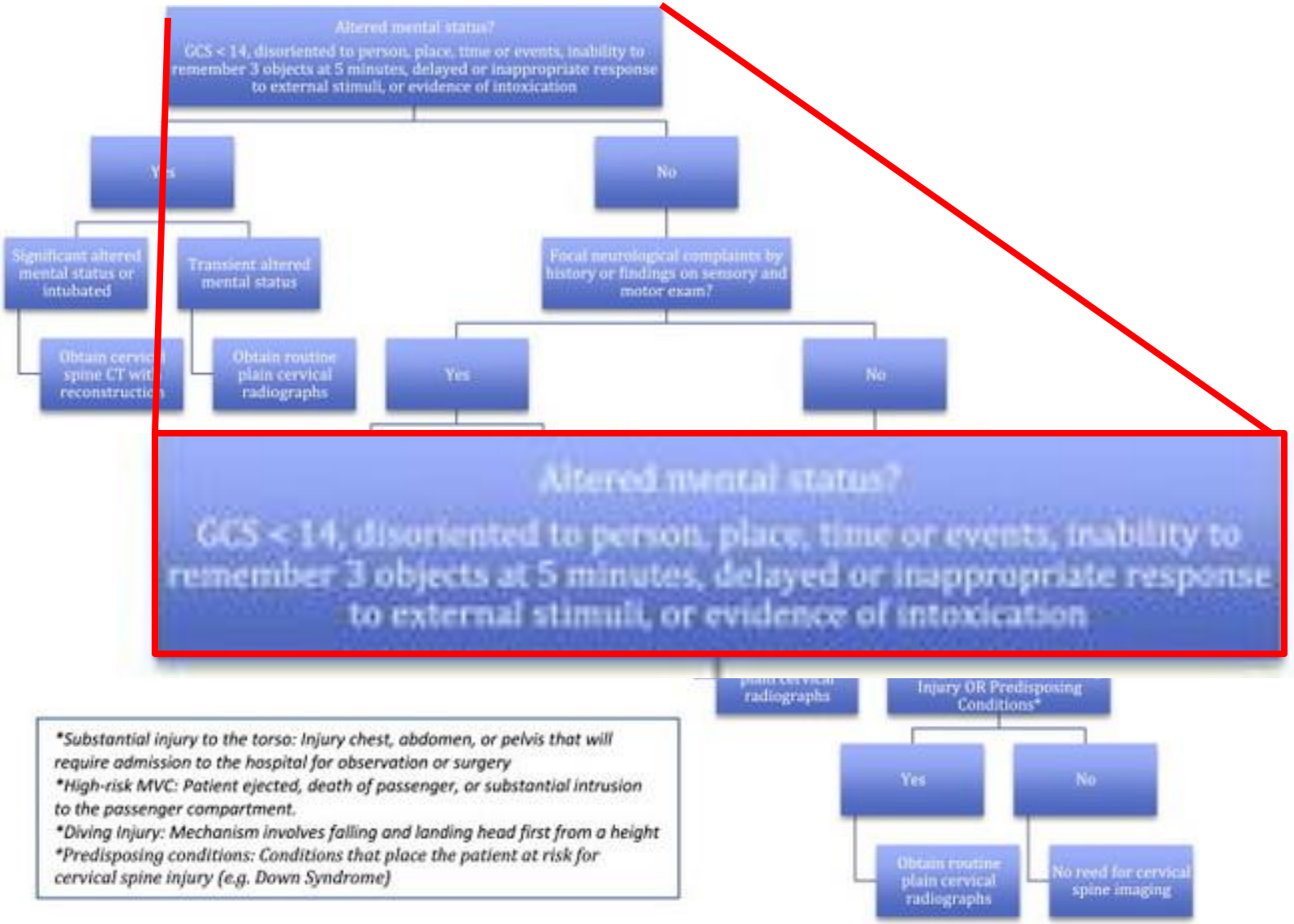
Spinal Imaging

- Obtain plain radiographs to evaluate for fractures or subluxations
  - 90% sensitive for bony cervical spine injury
  - Good option for screening in alert patient
  - Flexion-extension plain radiography helps evaluate for ligamentous stability but not useful acutely
- CT 100% sensitive for bony cervical spine injury
  - First choice for critically injured children
  - To avoid unnecessary radiation, algorithms available for C-spine injury
- MRI 100% sensitive for bony, ligamentous, and cord injuries but takes more time

Algorithm for C-spine Imaging



 Leonard JC. *Pediatr Clin N Am*. 2013; 60:1123-1137.



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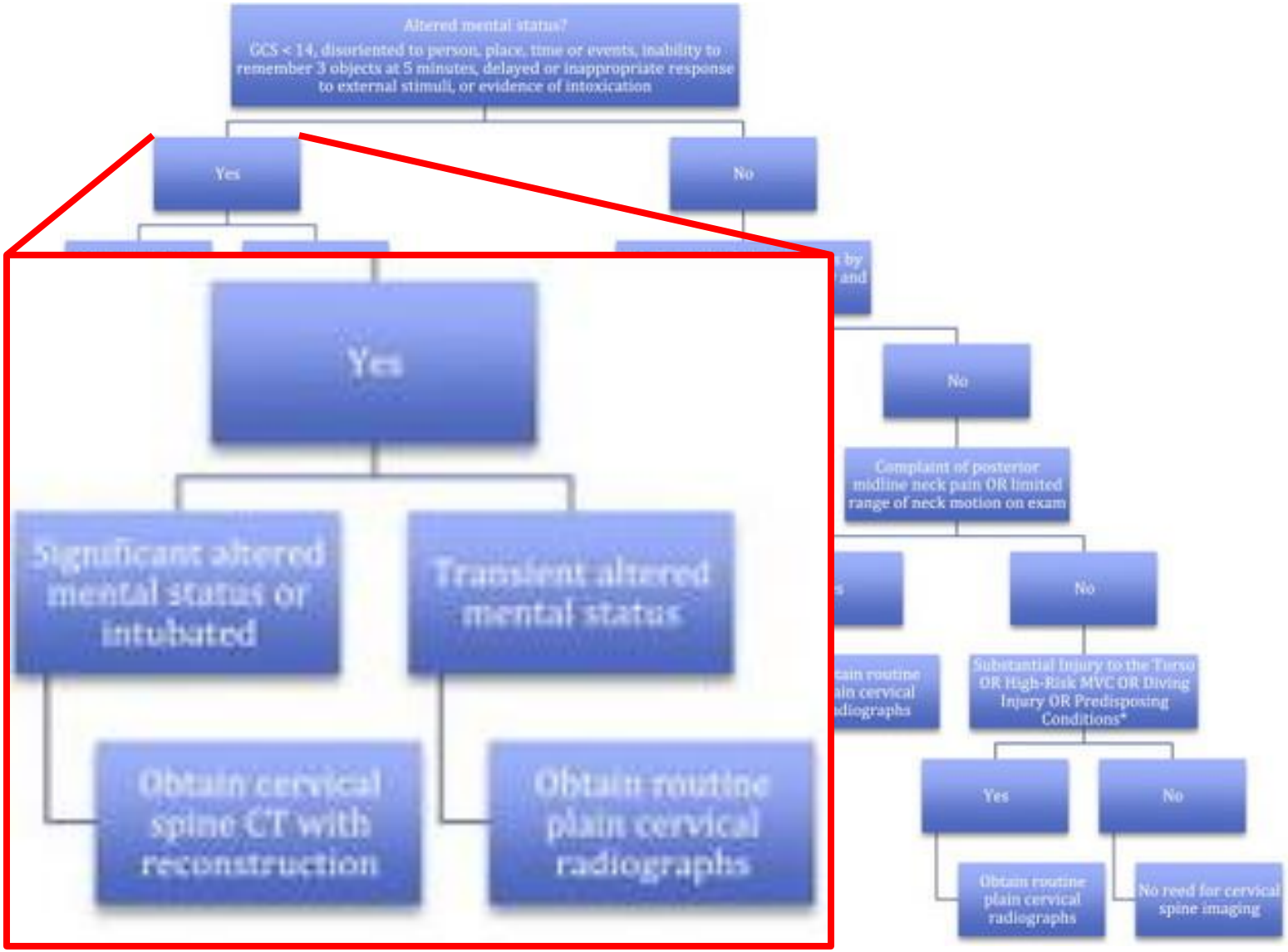

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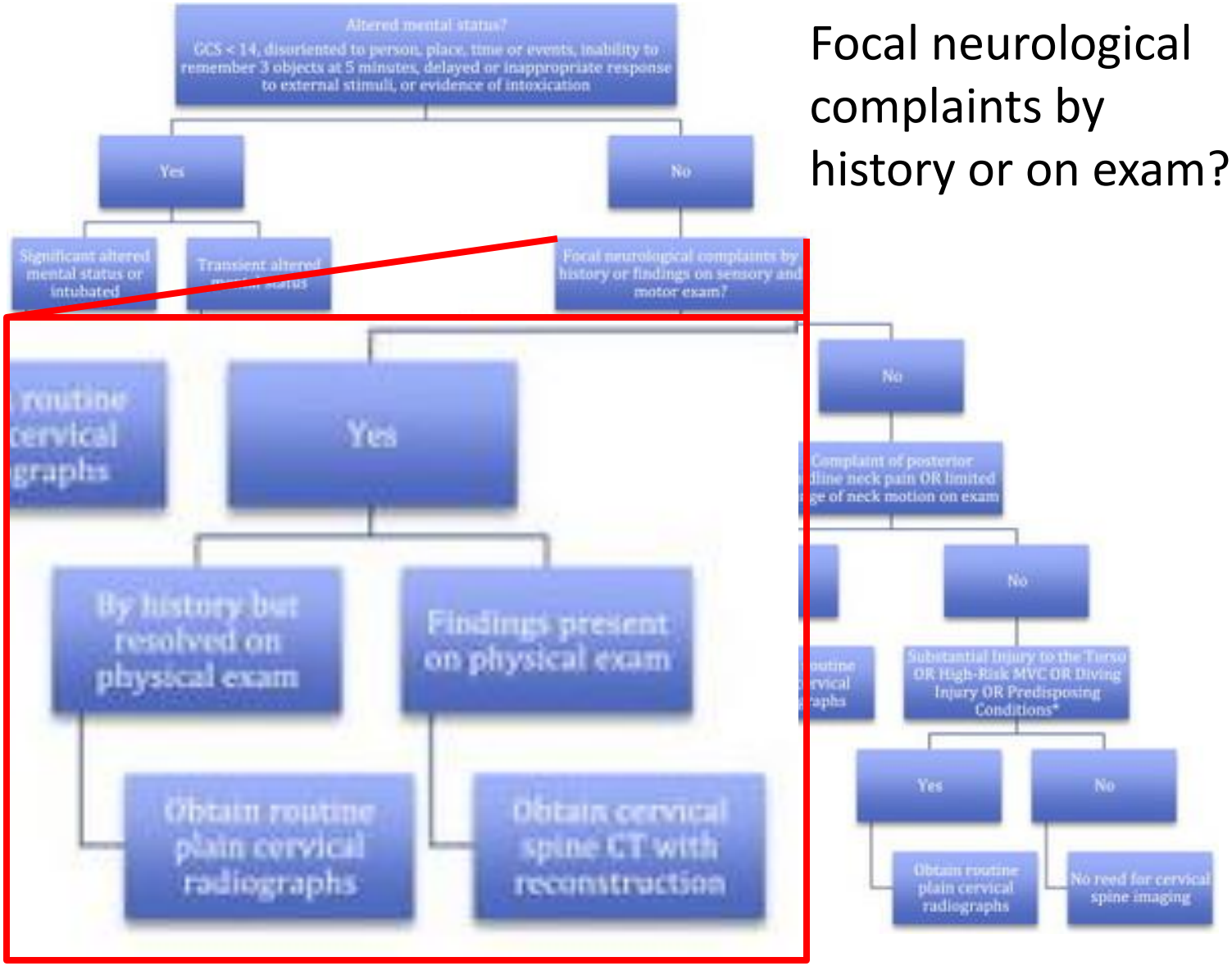
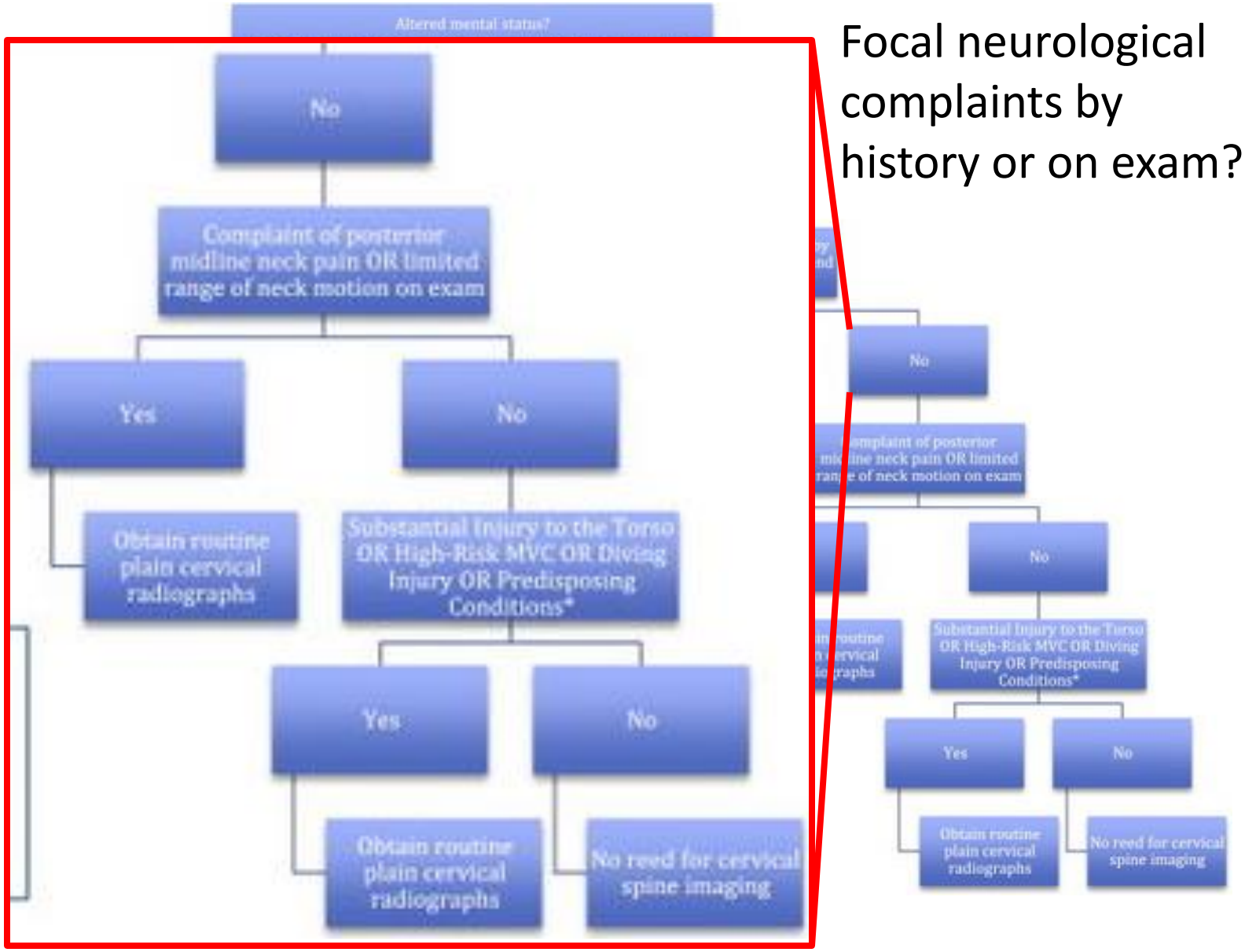
Focal neurological complaints by history or on exam?



 Leonard JC. *Pediatr Clin N Am*. 2013; 60:1123-1137



Focal neurological complaints by history or on exam?

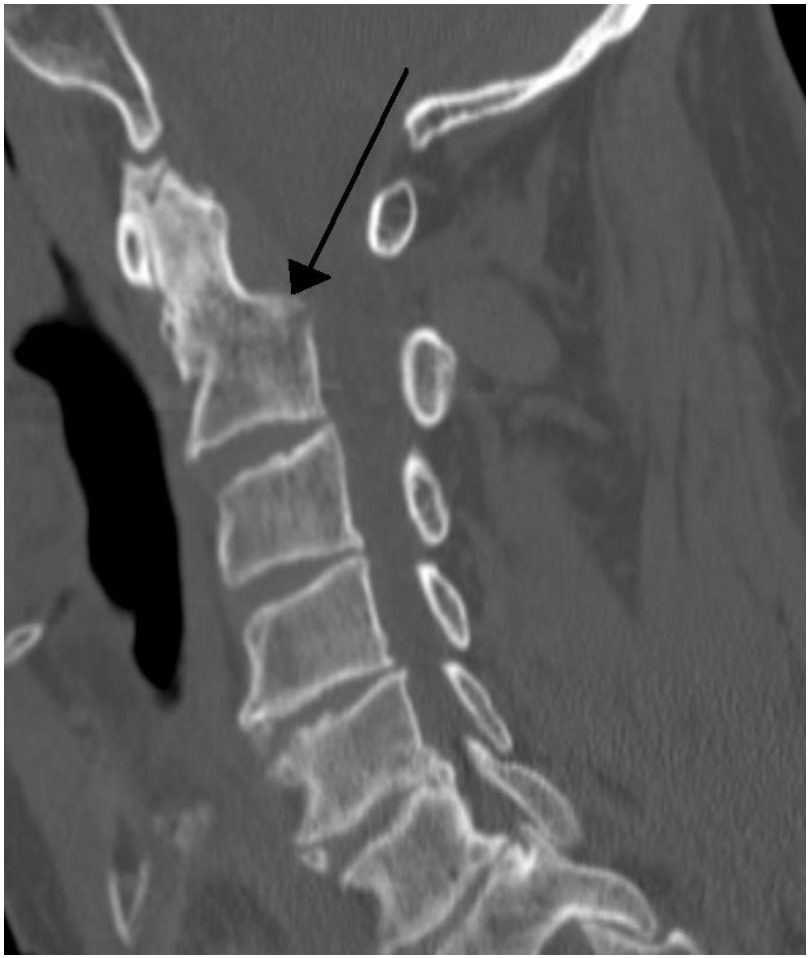
 Leonard JC. *Pediatr Clin N Am*. 2013; 60:1123-1137

Specific Spine Injuries

Atlantoaxial Dislocation

- “Cock robin” torticollis – chin rotation to the contralateral side and flexion of neck
- Children predisposed due to ligamentous laxity and robust synovium
- C-spine x-rays show only asymmetric positioning of dens relative to lateral masses and have a normal neurologic exam
- Management: rigid collar, pain medication and muscle relaxant
  - See spine specialist in 2 weeks

Dens Fractures



PD-INEL James Heilman, MD (Wikimedia Commons)

- In children, weakest point in the axis (C2) is cartilaginous subdental epiphysis
  - Present until 7 years old
  - Can be hard to tell if there is fracture at epiphysis
- From forceful neck flexion
- If significantly displaced, can cause neurologic deficit

Chance Fractures

- Hyperflexion injury over a seat belt during sudden deceleration in a motor vehicle accident
  - Children at risk because the lap belt rides higher
- Leads to anterior vertebral compression with rupture of ligaments
- Associated with intraabdominal injuries
  - Tears and transections of duodenum, jejunum, and mesentary

Spinal Shock

- Flaccid below level of lesion
- Absent reflexes
- Autonomic dysfunction leading to hypotension, bradycardia, and hypothermia
  - Hypotension characterized by low diastolic blood pressure and wide pulse pressure from loss of vascular tone
  - Refractory to fluids
- Treat with primary alpha-agonists such as norepinephrine and phenylephrine

Summary

- Neurologic emergencies in pediatrics can be caused by many reasons
- Ensure ABCs are the first thing to be managed
- Let the history and exam help you determine lab testing and imaging

Resources

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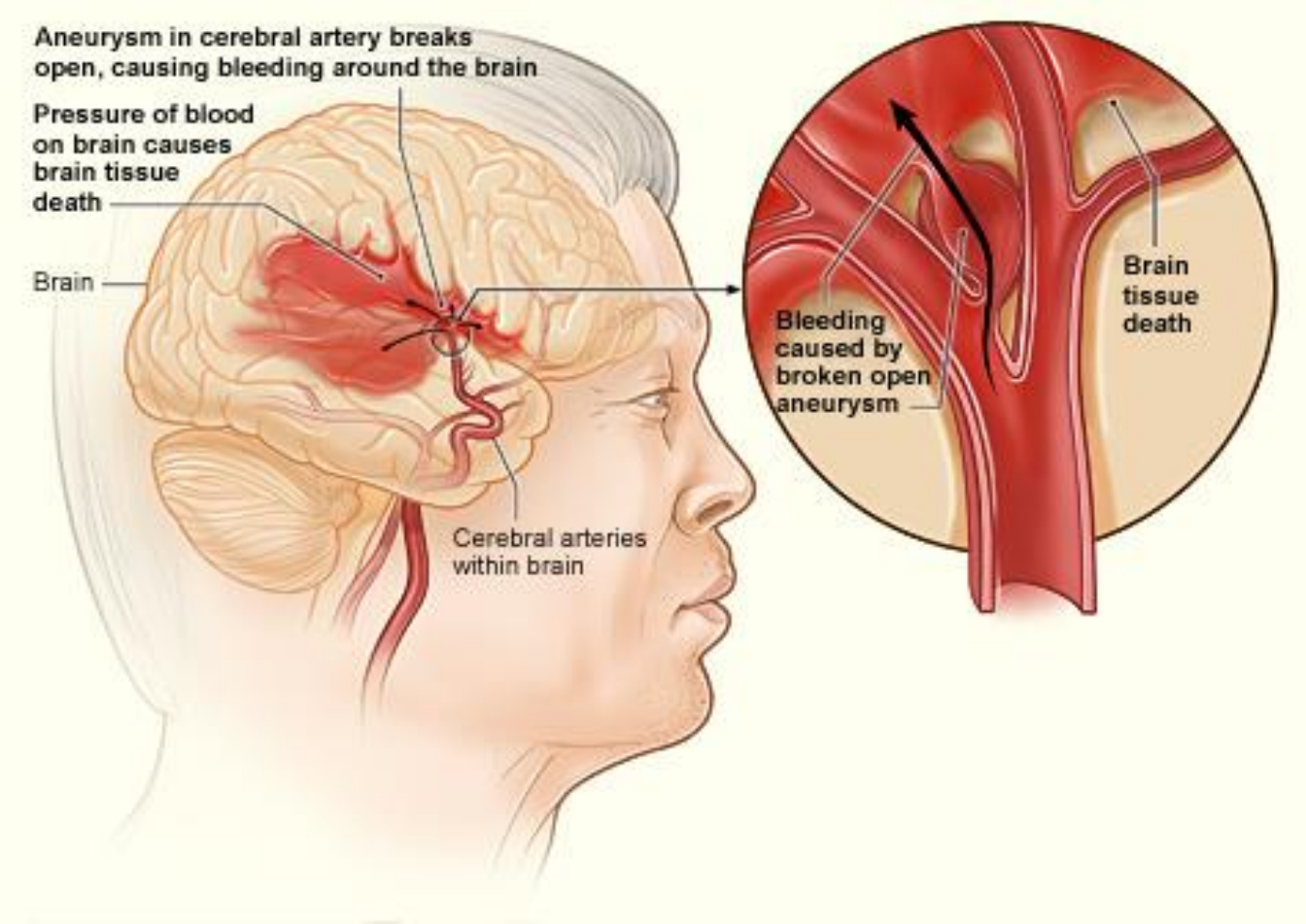
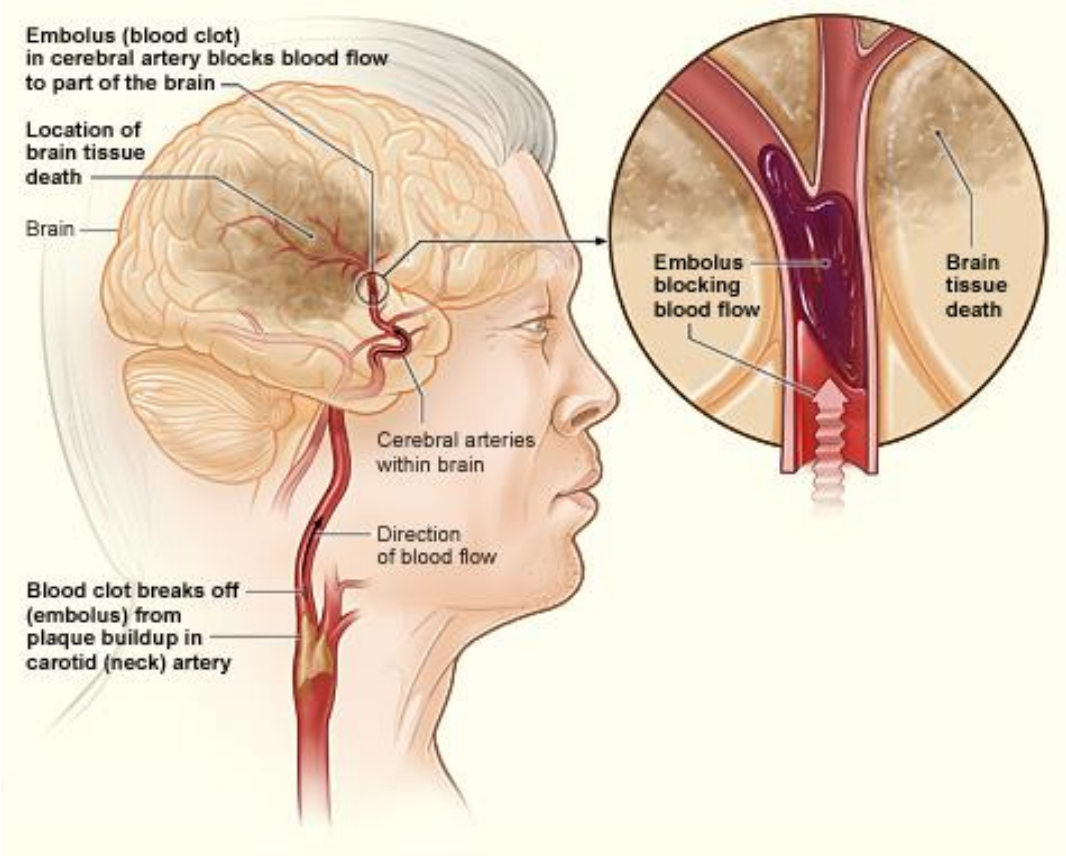
Subcommittee on febrile seizures. Clinical Practice Guideline – Febrile seizures: Guideline for the neurodiagnostic evaluation of a child with simple febrile seizure. *Pediatrics.* 2011; 127:389-394.

Hemorrhagic Stroke



Ischemic Stroke

Clot leads to reduction in cerebral blood flow and hypoxic damage

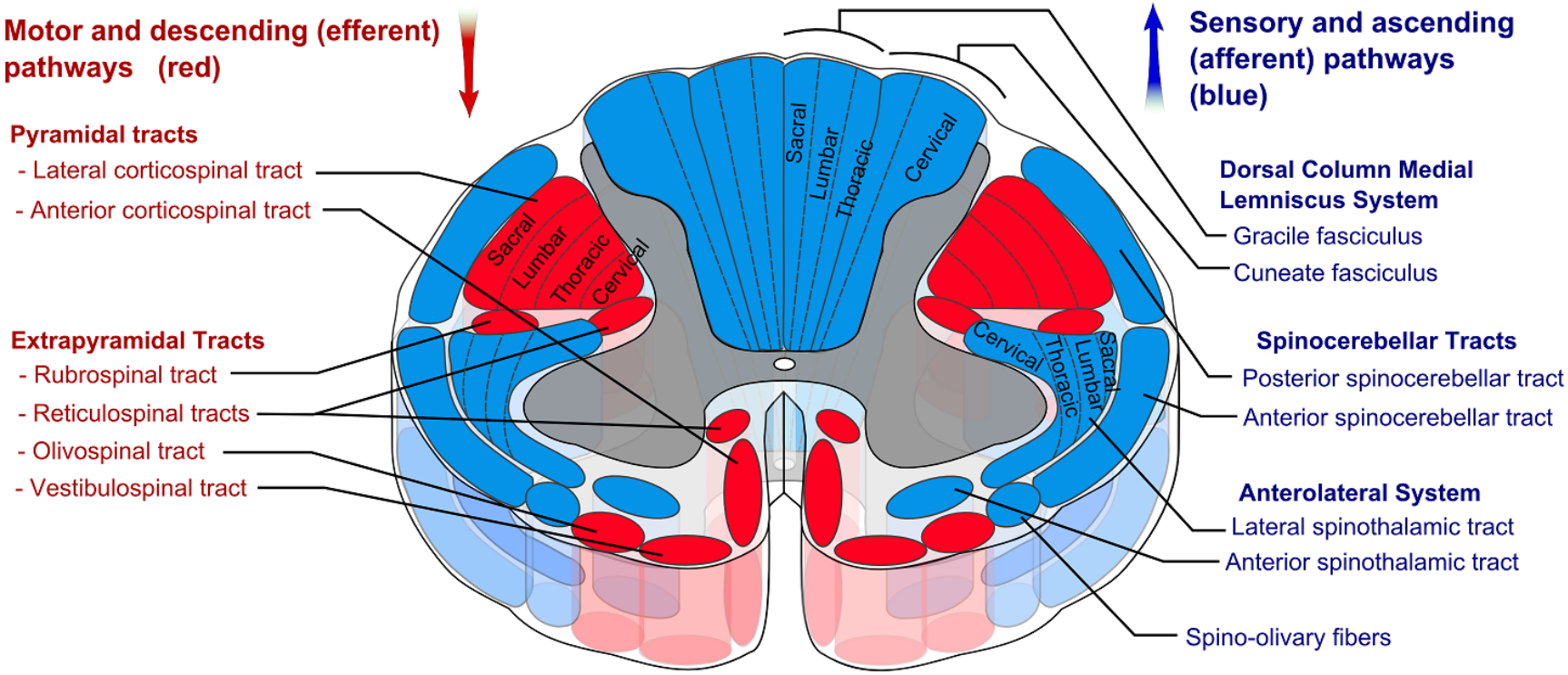


 National Heart Lung and Blood Institute (Wikimedia Commons)

Spinal Cord Injury: Anatomy Review

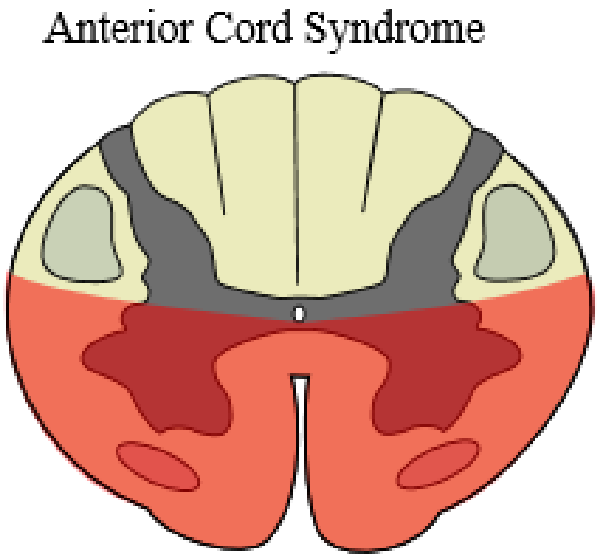
- Cord has ventral (motor) compartment and dorsal (sensory) compartment
- Upper motor neurons originate on one side of cerebral cortex, then cross at level of medulla before entering spinal cord
- Sensory neurons originate at one side and immediately cross other side before being entering cord

Spinal Cord Anatomy



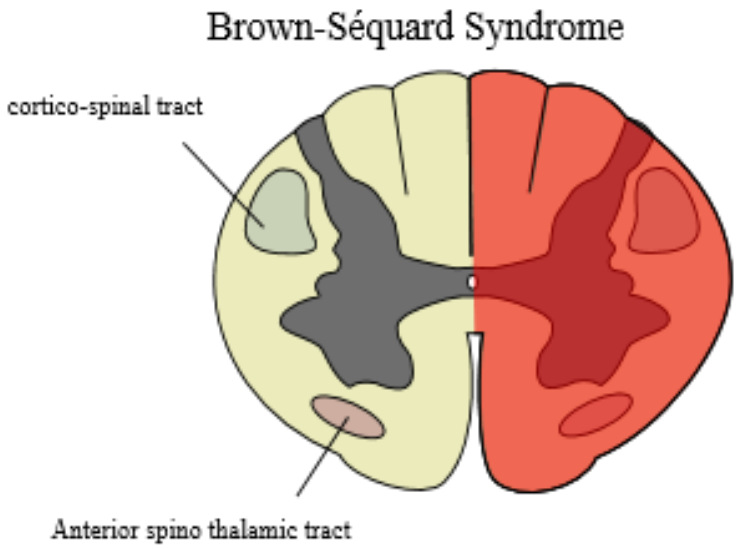
Polarlys and Mikael Häggström (Wikimedia Commons)

Anterior Cord Syndrome



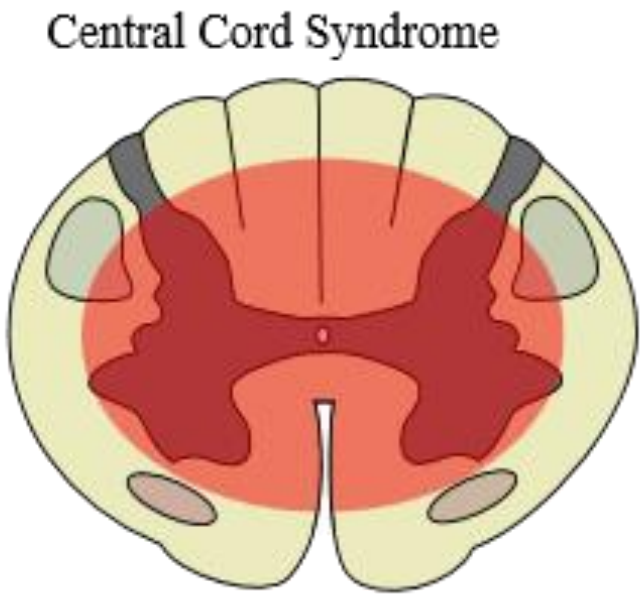
- Complete motor paralysis
- Loss of pain and temperature sensation
- Position and vibration sense preserved
- Associated with severe flexion injury

Brown-Sequard Syndrome



- Injury to one of the two lateral sides of the cord
- Ipsilateral (same side of injury) loss of motor function and position and vibratory sensation
- Contralateral (opposite side of injury) loss of pain and temperature sensation

Central Cord Syndrome



- Hyperextension injury may cause more severe injury to central regions of cord
- Diminished or absent upper extremity function
- Preserved lower extremity function

SCIWORA

- Spinal cord injury without radiographic abnormalities
- Children at high risk because of the flexibility of their spinal column
  - Can sublux transiently and cause compression, then return to normal position prior to x-ray
- Some have positive MRI,

Subfalcine Herniation (3)

- One cerebral hemisphere herniates beneath the falx cerebri to the opposite side
- Usually from unilateral supratentorial mass lesion
- Bilateral leg weakness, disturbances of bladder control