NEUROLOGICAL DISEASE

SURGICAL CRITICAL CARE BOARD REVIEW

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Nothing To Disclose

Coma

- Coma is defined as “a state of extreme unresponsiveness, in which an individual exhibits no voluntary movement or behavior” (1).
  - Alternatively, coma is a state of *unarousable unresponsiveness* in which the patient lies with the eye closed and has no awareness of self and surroundings (2).
- Coma lies on a spectrum with other alterations in consciousness – from confusion to delirium to obtundation to stupor to coma and, ultimately, brain death (2).
- To be clearly distinguished from syncope, concussion, or other states of transient unconsciousness coma must persist for at least one hour (2).
- There are 2 important characteristics of the conscious state (3)
  - The level of consciousness – “arousal or wakefulness”
    - Regulated by physiological functioning and consists of more primitive responsiveness to the world such as predictable involuntary reflex responses to stimuli.
    - Arousal is maintained by the reticular activating system (RAS) - a network of structures (including the brainstem, the medulla, and the thalamus)
  - The content of consciousness – “awareness”
    - Regulated by cortical areas within the cerebral hemispheres,
- There are two main causes for coma:
  - Bihemispheric diffuse cortical or white matter damage or
  - Brainstem lesions bilaterally affecting the subcortical reticular activating systems.
- A huge number of conditions can result in coma. One way to categorize these conditions is to divide them into the anatomic and the metabolic causes of coma.
  - Anatomic causes of coma are those conditions that disrupt the normal physical architecture and anatomy, either at the level of the cerebral cortex or the brainstem
  - Metabolic causes of coma consist of those conditions that change the chemical environment of the brain.
  - The main causes are divided into:
    - Metabolic – Electrolyte abnormalities (hypo- or hyper-natremia, hypo- or hyper-calcemia), hypoglycemia, DKA, nonketotic hyperosmolar coma, hypothyroidism, uremia, hepatic encephalopathy, hypo- or hyper-thermia, hypercarbia, hypoxia/anoxia
    - Intoxications – Barbiturates, opiates, alcohol, benzodiazepines, other drugs of abuse, salicylates
    - Toxins – Carbon monoxide
    - Infections, both CNS and non-CNS – Meningitis, cerebritis, encephalitis, sepsis
    - Seizures – nonconvulsive status epilepticus, post-ictal states
    - Intracranial processes
      - Cortical - TBI, SAH, cerebral edema
      - Brain stem – high-grade SAH, severe TBI, posterior fossa hemorrhage
      - Herniation syndromes from any of the above or local effects from tumors and other mass lesions
- Diagnosis of unexplained coma is clinical, but the work up for etiology can be complex and should include the following as clinically indicated:
  - History (ie drug ingestion, trauma, anoxia)
  - Physical examination to evaluate brain stem involvement (pupillary response, etc.) and possible etiologies such as trauma
  - Standard laboratory testing to evaluate electrolyte abnormalities
  - Toxicology screen
  - ABG to evaluate for hypoxia or hypercarbia and for metabolic acidosis that may be associated with certain ingestions
  - Head CT to look for intracranial hemorrhage, cerebral edema, etc
  - EEG to R/O seizures
  - Lumbar puncture
  - MRI can be considered when other tests fail to demonstrate cause
- Treatment of coma is largely supportive.
  - Initial resuscitation will frequently include intubation either due to a failure to protect the airway or ventilator failure.
  - Administration of dextrose and naloxone (0.2-0.4 mg) for patients with unclear etiology and acute development of coma
  - Treatment of coma obviously is dictated by the underlying cause

Brain Death

- In the setting of very severe neurological injury, brain death often occurs following coma.
  - The cycle of neuronal injury leading to neuronal swelling leading to increased intracranial pressure leading to decreased intracranial blood flow causing further neuronal injury leads to a situation where the intracranial pressure exceeds the systemic blood pressure resulting in brain death.
- The leading causes of brain death in adults are traumatic brain injury and subarachnoid hemorrhage (4). Hypoxic-ischemic insults and fulminant hepatic failure are also common causes.
- Brain death is defined legally by the Uniform Determination of Death Act (UDDA) from The President's Commission report on "guidelines for the determination of death" (5).
  - "An individual who has sustained either(5)
    - Irreversible cessation of circulatory and respiratory functions, or
    - Irreversible cessation of all functions of the entire brain, including the brain stem, is dead.
    - A determination of death must be made with accepted medical standards."
- The American Academy of Neurology (AAN) published a practice parameter in 1995 to delineate the medical standards for the determination of brain death (7).
  - There are several important principles
    - Brain death is a clinical diagnosis
    - To determine brain death, there must be absence of function of the entire brain, including the brain stem and irreversibility.
    - 3 clinical findings necessary to confirm irreversible cessation of all functions of the entire brain, including the brain stem: coma, absence of brainstem reflexes, and apnea.
  - The following is taken directly from these practice parameters (7) as well as a recent update to the 1995 practice parameters which clarified a few salient points about brain death determination (8):
    - Prerequisites. Brain death is the absence of clinical brain function when the proximate cause is known and demonstrably irreversible.
      - [illegible]
    - Pitfalls in the diagnosis of brain death
      - [illegible]
    - Clinical observations compatible with the diagnosis of brain death
      - [illegible]
    - Confirmatory laboratory tests
      - [illegible]
    - Documentation
      - [illegible]
- Policies surrounding the determination of brain death are typically State or Institution-specific.
  - Some policies require the clinical examination to be performed by neurologists, neurosurgeons, and/or intensivist.
  - Some require an apnea test with each clinical examination, while some only require one.
  - If a certain period of time has passed since the onset of the brain insult to exclude the possibility of recovery (usually several hours), 1 neurologic examination should be sufficient to pronounce brain death.
    - However, some US state statutes require 2 examinations, typically separated by 6 hours for adults.
    - Some require that 2 different physicians perform the examinations.
  - Most policies have different criteria for pediatric patients often requiring extended periods between clinical examinations or require confirmatory testing.
- Pathophysiological changes at the time of brain death
  - Cardiovascular effects
    - Progression of brain death causes rostral to caudal ischemia
    - Two phases in the process of brain death
      - [illegible]
    - Brain ischemia induces myocyte necrosis
    - Ultimately results in cardiac dysfunction and vasodilatation
      - [illegible]
    - Arrhythmias are also common
  - Pulmonary effects

CNS Infections - Bacterial meningitis

- Approximately 80% of bacterial meningitis cases are caused by the *Streptococcus pneumonia* and *Neisseria meningitides*
- Usually caused by encapsulated organisms in immonuocompetant hosts
  - Organisms typically enter the CNS via the bloodstream.
  - Direct inoculation of the CNS can occur in the setting of trauma, surgery, monitoring devices, or seeding through parameningeal structures
- The clinical presentation of patients with meningitis include rapid onset of fever, headache, photophobia, nuchal rigidity, lethargy, malaise, altered mentation, seizure, or vomiting
  - The ‘‘classic triad’’ of fever, neck stiffness, and altered mental status may be present in up to 2/3rds of patients, with fever being the most common
  - Immunocompromised may mount none of the classic symptoms and therefore meningitis should be in the differential diagnosis of any immunocompromised patient with altered mental status.
  - Physical examination findings classically include:
    - Focal deficits and increased intracranial pressure (ICP)
    - Nuchal rigidity
    - Meningeal irritation (Brudzinski’s and Kernig’s sign)
    - Purpura or petechia of the skin in meningococcemia
  - Diagnosis with lumbar puncture (LP) can make the diagnosis and speciate organisms to guide antibiotic choice.
    - LP can generally be safely performed without first obtaining a CT, although this is controversial. If concern about increased ICP or mass lesion exists, empiric antibiotics should be administered immediately so that head CT can be obtained prior to LP and not delay antibiotic administration.
    - CSF analysis in bacterial meningitis includes classically:
      - High opening pressure
      - High WBCs with a predominance of PMNs
      - Low glucose (<40)
      - High protein
- Treatment is rapid administration of a bactericidal antibiotic with good CNS penetration and the use of anti-inflammatory agents
  - Ceftriaxone or cefotaxime, and vancomycin is often recommended as first line therapy
  - In the very young, very old, and immunocompromised patients, empiric coverage for Listeria should be given with ampicillin
  - Current recommendations in adults include the use of dexamethasone to prevent the sequelae of bacterial lysis,
- Mortality rates range from 20-25% with higher rates in the elderly and those with major comorbidities.

CNS Infections - Viral encephalitis

- Causative agents include arboviruses, HSV, HZV, EBV, CMV, and rabies.
  - During epidemics, the arboviruses can account for 50%
  - Epidemics are often seasonal from June to September
  - In immunocompromised host, CMV and HZV are frequently seen
- Clinical manifestations include:
  - Rapid development of fever
  - Headache
  - Confusion
  - Localizing neurological signs
  - New-onset seizures
  - Meningeal irritation causing headache, photophobia, nuchal rigidity
- Diagnostic evaluation includes ruling out bacterial meningitis and subarachnoid hemorrhage
  - LP often appears “aseptic”
  - CT and MRI may demonstrated abnormalities
  - EEG is helpful for certain diseases such as HSV encephalitis
  - Open brain biopsies may be indicated for selected patients with atypical presentations, clinical deterioration despite treatment, or nondiagnostic PCR results
- Treatment is largely supportive
  - Empiric treatment with acyclovir while awaiting PCR results
  - HSV is treated with acyclovir (10 mg/kg q8h x 10-14 days). Must be renal-dose adjusted
  - Ganciclovir +/- foscarnet for CMV encephalitis
- Prognosis is dependent on the causative organism with excellent prognosis for EBV and Colorado tick fever, for example, and over 50-70% mortality for EEE and near 100% mortality for rabies.

CNS Infections - Brain abscess

- Traditional causes of brain abscess include otitis media and sinusitis.
  - With improvements in treatment for these common conditions, tooth abscesses, endocarditis, and pulmonary infections are becoming more common causes
  - Multiple abscesses usually suggest hematogenous spread
- Classic symptoms include headache, focal neurological deficit, and change in mental status
  - Also may have nonspecific indolent courses
  - Papilledema may occur which is rare with other CNS diseases
  - Focal symptoms depend on location of lesion
- Diagnosis is typically made with CT
  - CT with IV contrast demonstrates hypodense lesions with contrast-enhancing rings
  - MRI is highly sensitive
- Treatment
  - Antibiotics – empiric coverage with Nafcillin or a third-generation cephalosporin and Flagyl
    - In immunosuppressed patients, antifungals, anti-TB, antiparasitic, and atypical bacteria should also be instituted
  - Surgical drainage for large (>3 cm), superficial, or cerebellar abscesses.
  - Surgical drainage is also indicated for patients who are immunosuppressed, in coma, or have rapid clinical deterioration.
  - Medical management, at least initially, is advocated for patients with brainstem, multiple, or deep-seated lesions <3 cm

Cerebral edema

- Common causes include:
  - Traumatic brain injury (TBI)
  - Ischemic strokes
  - Non traumatic intracranial hemorrhage
  - Infections such as meningitis or encephalitis
  - Tumors
  - Fulminant hepatic failure
  - Hypoxia/anoxia

- Placement of intracranial monitoring devices to monitor ICP and trend therapy is indicated for the following conditions
  - TBI
    - “Salvageable patients” with a GCS 3-8 after resuscitation and an abnormal CT
    - Severe TBI with a normal CT scan with 2 or more of the following
      - Age >40
      - Unilateral or bilateral posturing
      - SBP <90
  - Non-traumatic intracranial hemorrhage
    - Lobar or gangionic hemorrhage with motor GCS ≤4 or midline shift on CT
  - Ischemic stroke
    - MCA infarct - Edema with midline shift on CT
    - Cerebellar infarct - Acute hydrocephalus
  - HSV encephalitis
    - Motor GCS ≤4
    - Necrotic mass
  - Aneurysmal subarachnoid hemorrhage (SAH) (21)
    - Acute hydrocephalus

Cerebral edema – Key Concepts

- Monroe-Kellie Doctrine
  - The intracranial contents are brain tissue, blood, and CSF
  - The total volume of the contents of the intracranial vault must remain constant because the skull is rigid and incompressible
  - If there is an increase in volume in any of the 3 components, it must occur at the expense of the other 2 or herniation will occur
- Cerebral perfusion pressure
  - Management of cerebral edema is not just about management of ICP, but also cerebral perfusion pressure (CPP = MAP – ICP)
  - CPP should be maintained to maximize perfusion but minimize the risks of systemic hypertension
  - The optimal CPP is a matter of some debate, but clearly CPP <50 should be avoided
  - Aggressive attempts to raise CPP >70 with fluids and vasoactives have been associated with an increased risk for ARDS
  - The optimal CPP for patients with TBI is thought to be between 50 – 70 mm Hg
- Autoregulation
  - The intact cerebrovascular system normally maintains cerebral blood flow (CBF) at CPP from 50 to 150 mm Hg or MAP of 60 to 160 mm Hg via autoregulation via reflexive vasodilatation and vasoconstriction in response to alterations in cerebral perfusion.
  - Similarly, in the normal brain, there is pCO2 responsiveness (and to a much lesser degree pO2 reactivity) whereby elevated pCO2 causes vasodilatation and lower pCO2 induces vasoconstriction.
  - In the setting of intracranial insults, autoregulation may be lost and responses to alterations in systemic pressure (or alterations in PCO2) may be markedly abnormal in the injured brain
    - Loss of autoregulation causes a linear increase between CBF and CPP

Cerebral edema - Management

- General measures for all patients at risk of cerebral edema
  - Maintain adequate oxygenation and ventilation
  - Mechanical ventilation for all patients who are at risk of loss of ability to protect the airway
  - Prevent hypotension. Keep SBP >90 mm Hg
  - Maintain normothermia as fever causes an increase in CBF via an increase cerebral metabolic rate
  - Semi-upright head position at 30◦
  - Adequate analgesia and sedation to minimize agitation and pain
  - Maintain euvolemia
  - Provide seizure prophylaxis when appropriate (essentially all CT verified TBI except isolated SAH or IVH)
  - Avoid noxious stimuli

- Management of elevated ICP targets the “3 compartments”
  - CSF compartment
    - Drainage of CSF via intraventricular catheters (IVC) or external ventricular drains (EVD)
    - Lumbar drains should be avoided except in rare cases due to the risk of induced herniation syndromes
  - Blood compartment
    - Sedation
      - Adequate sedation should be provided to not only prevent systemic and intracranial responses to stimuli, but also to lower CBF via decreasing cerebral metabolic rate
      - Sedatives should preferably be short acting
      - Propofol is recommending for acute ICP management, but has not been shown to improve outcome (22)
      - Benzodiazepines are frequently used in many institutions
      - Barbiturates
    - Hyperventilation
  - Brain tissue
    - Surgical evacuation
    - Osmolar therapies
      - Both mannitol and hypertonic saline are effective at lowering intracranial pressure by decreasing brain tissue water content
      - Mannitol
      - Hypertonic saline
  - Decompressive craniectomy
    - Commonly used in the management of TBI and increasing in the management of severe cerebral edema in patients with territorial infarction, decompressive craniectomy allows for the intracranial contents to herniate out of a surgical defect and relieve intracranial pressure
    - Indications are typically institution-specific, but generally this procedure is reserved for patients who have intracranial hypertension refractory to conventional medical therapy
    - The recently published DECRA trial demonstrated more unfavorable neurological outcomes in patients with severe TBI treated with early bifrontotemporoparietal decompressive craniectomy
  - Therapeutic hypothermia

Seizures

- Status epilepticus (SE) is defined as
  - “5 minutes or more of continuous clinical and/or electrographic seizure activity OR recurrent seizure activity without recovery (returning to baseline) between seizures”
  - The more traditional definition is “more than 30 minutes of continuous seizure activity or two or more sequential seizures without full recovery of consciousness between seizures”
  - SE is classified as either
  - Generalized - Convulsive (GCSE) or Nonconvulsive (NCSE).
  - Partial - Simple or Complex
  - Refractory SE (RSE) is SE that does not respond to standard treatment such as an initial benzodiazepine and an anti-epileptic medication
  - Convulsive seizures can lead to significant metabolic effects including rhabdomyolysis, acute respiratory failure, lactic acidosis, hyperglycemia, renal failure, etc.
  - NCSE can cause significant neuronal injury if left untreated
- Diagnosis
  - The diagnosis of seizure is made with EEG
    - Ideally the EEG should be performed without benzodiazepines or antiepileptics to better identify a seizure focus
    - Can diagnose both partial seizure via identification of an abnormal seizure focus in the cortex and generalized seizures by identification of diffuse cortical activity or postictal slowing and/or depressed amplitude
    - Mandatory in any patient with an abnormal mental status after a generalized convulsive seizure to R/O partial treatment and NCSE
  - Workup for cause of seizure should include CT and MRI, evaluation for metabolic disorders, CSF analysis
- Treatment
  - As a general rule, a single or few self-limited seizures in an ICU patient do not warrant specific treatment.
  - The treatment of SE
    - Airway protection
    - IV hydration if fever or concern for rhabdomyolysis from extended convulsions
    - Benzodiazepines – typically lorazepam 0.1 mg/kg IV
    - Antiepileptic (AED) administration - no specifically preferred agent (phenytoin, fosphenytoin, levetiracetam)
    - If SE persists, subspecialty consultation should usually be requested
    - RSE is typically treated with the following via high-dose infusion. End-points of treatment are cessation of seizures on EEG or burst suppression.
      - High-dose benzodiazepines (midazolam or lorazepam)
      - Propofol
      - Phenobarbital
      - Ketamine

Ischemic Stroke - Evaluation

- Initial Management
  - Airway support and ventilatory assistance are recommended for patients who have decreased consciousness or who have bulbar dysfunction causing compromise of the airway
  - Supplemental O2 should be administered to all patients with any degree of hypoxia to maintain an O2 saturation >94%.
  - Fever should be aggressively treated
  - Both hyperglycemia and hypoglycemia should be aggressively treated
  - Hypertension
    - There is conflicting data on management of hypertension for ischemia stroke. A ”cautious approach” to the treatment of arterial hypertension should be recommended
    - If the patient is a candidate for rTPA, they should have their blood pressure lowered to systolic blood pressure is ≤185 mm Hg and diastolic blood pressure is ≤110 mm Hg. The blood pressure should be stabilized and maintained below 180/105 mm Hg for at least the first 24 hours after intravenous rtPA treatment.
    - Patients with markedly elevated blood pressure may have their blood pressure lowered.
      - Suggested algorithm is to lower blood pressure by ~15% during the first 24 hours after onset of stroke.
    - If the patient is not a candidate for thrombolytics, antihypertensive medications should be withheld unless the systolic blood pressure is >220 mm Hg or the diastolic blood pressure is >120 mm
    - Labetalol and Nicardipine are recommended agents based on expert consensus
- Imaging
  - CT scanning of the brain is recommended before initiating any specific therapy to treat acute ischemic stroke
  - Other than hemorrhage and evidence of large volume of infarction, there are no specific CT findings that should preclude treatment with rtPA within 3 hours of onset of stroke
  - Multimodal CT and MRI may provide additional information that will improve diagnosis of ischemic stroke
  - Emergency treatment of stroke should not be delayed in order to obtain multimodal imaging studies however

Ischemic Stroke - Treatment

- Thrombolytics
  - Intravenous rtPA (0.9 mg/kg, maximum dose 90 mg) is recommended for selected patients present within 3 hours of onset of ischemic stroke. Blood pressure must be controlled prior to administration. A subset of patients may benefit if administered up to 4.5 hours after onset of symptoms
    - exclusions are age >80, oral anticoagulation, NIHSS score >25, evidence of ischemia on imaging of >1/3 of the MCA territory, history of previous stroke and diabetes.
  - Intra-arterial thrombolysis is an option for treatment of selected patients who have major stroke of <6 hours' duration due to occlusions of the MCA and who are not otherwise candidates for intravenous rtPA.
    - Intra-arterial thrombolysis or mechanical thrombectomy is reasonable in patients who have contraindications to use of intravenous thrombolysis, such as recent surgery
- Anticoagulation - Urgent anticoagulation is not recommended for patients with moderate to severe strokes because of an increased risk of serious intracranial hemorrhagic complications
- Antiplatelet agents - Aspirin (initial dose is 325 mg) within 24 to 48 hours after stroke onset is recommended
- Reperfusion devices/mechanical thrombectomy devices (such as MERCI) can be used in selected patients although improvements in patient outcomes have not been established.

Ischemic Stroke - Management of complications

- Aggressive treatment of edema and close monitoring of the patient for signs of neurological worsening during the first days after stroke are recommended
- Patients with acute hydrocephalus secondary to an ischemic stroke should be treated with placement of a ventricular drain
- Decompressive surgical evacuation of a space-occupying cerebellar infarction is a potentially life-saving measure and clinical recovery may be very good
- Recurrent seizures after stroke should be treated
- Osmotherapy, has been recommended for treatment of deteriorating patients with malignant brain edema after large cerebral infarction, with the recognition that these measures are unproven
- Hyperventilation should be used as a short-term intervention.
- Decompressive surgery for malignant edema of the cerebral hemisphere is effective and potentially life-saving.
  - Both the age of the patient and the side of the infarction (dominant versus nondominant hemisphere) may affect decisions about surgery.
  - Although the surgery may be recommended for treatment of seriously affected patients, the physician should advise the patient's family about the potential outcomes, including survival with severe disability
- Corticosteroids are not recommended
- Prophylactic administration of anticonvulsants is not indicated

Intracranial hemorrhage (ICH) - Evaluation

- Initial Management
  - Initial management of airway, oxygenation, etc. is the same as for any patient with a neurological emergency
  - Corticosteroids are not recommended
  - Normoglycemia should be maintained
  - Blood pressure control is first line therapy for these patients and based on Class C recommendations:
    - If SBP is >200 mm Hg or MAP is >150 mm Hg, then consider aggressive reduction of BP with continuous intravenous infusion, with frequent BP monitoring every 5 min.
    - If SBP is >180 mm Hg or MAP is >130 mm Hg and there is the possibility of elevated ICP, then consider monitoring ICP and reducing BP using intermittent or continuous intravenous medications while maintaining a cerebral perfusion pressure ≥60 mm Hg.
    - If SBP is >180 mm Hg or MAP is >130 mm Hg and there is no evidence of elevated ICP, then consider a modest reduction of BP (eg, MAP of 110 mm Hg or target BP of 160/90 mm Hg) using intermittent or continuous intravenous medications to control BP and clinically reexamine the patient every 15 min
  - Rapid correction of coagulaopathy is essential
    - Patients with a severe coagulation factor deficiency or severe thrombocytopenia should receive appropriate factor replacement therapy or platelets
    - Patients with ICH whose INR is elevated due to warfarin should have their warfarin withheld, receive therapy to replace vitamin K–dependent factors and correct the INR, and receive intravenous vitamin K
    - PCCs have not shown improved outcome compared with FFP but may have fewer complications. They are reasonable alternatives to FFP
    - rFVIIa is not recommended as a sole agent for oral anticoagulant reversal in ICH

- Imaging
  - CT scan or MRI is essential in the management of these patients to differentiate ischemic from hemorrhagic stroke
  - CT angiography and contrast-enhanced CT may help to identify patients at risk for hematoma expansion
  - CT angiography, CT venography, contrast-enhanced CT, contrast-enhanced MRI, magnetic resonance angiography, and magnetic resonance venography can be useful to evaluate for underlying structural lesions, including vascular malformations and tumors

Intracranial hemorrhage (ICH) - Treatment

- Treatment
  - Intraventricular administration of recombinant tissue-type plasminogen activator is considered investigational, but appears to have a good safety profile
  - Surgical intervention
    - Patients with cerebellar hemorrhage who are deteriorating neurologically or who have brainstem compression and/or hydrocephalus from ventricular obstruction should undergo surgical removal of the hemorrhage as soon as possible
    - For patients with lobar clots >30 mL and within 1 cm of the surface, evacuation of supratentorial ICH by standard craniotomy should be considered

- Management of complications
  - Seizures
    - Clinical seizures should be treated
    - Continuous EEG monitoring is indicated in ICH patients with depressed mental status out of proportion to the degree of brain injury
    - Patients with a change in mental status who are found to have seizures on EEG should be treated with antiepileptic drugs
    - Prophylactic anticonvulsant medication should not be used
  - Cerebral edema
    - Patients with a GCS score of ≤8, those with evidence of transtentorial herniation, or those with significant IVH or hydrocephalus might be considered for ICP monitoring and treatment.
    - A CPP of 50 to 70 mm Hg may be reasonable depending on the status of cerebral autoregulation
  - Ventricular drainage as treatment for hydrocephalus is reasonable in patients with decreased level of consciousness

Aneurysmal Subarachnoid Hemorrhage (aSAH) - Evaluation

- A high level of suspicion for aSAH should exist in patients with acute onset of severe headache, the so-called “worst headache of my life.”
  - Other symptoms may include localizing neurological findings which may indicate the location of aneurysm , neck stiffness, 3rd nerve palsy (PCOM aneurysm)and generalized tonic-clonic seizures
- Common grading systems include:
  - World Federation of Neurological Surgeons (WFNS) which is based on GCS and motor deficit on admission
  - Hunt and Hess which is based on degree of neurological symptomatology (from asymptomatic or minimal headache to deep coma)
  - Fisher Grade based on CT findings

- Initial Management
  - Initial management of airway, oxygenation, etc. is the same as for any patient with a neurological emergency
  - Fever should be aggressively managed
  - Hyponatremia should be avoided and treated
  - In patients with unsecured aneurysms, BP control is one of the mainstays of therapy
    - Blood pressure should be controlled with a titratable agent to balance the risk of stroke, hypertension-related rebleeding, and maintenance of cerebral perfusion pressure
    - Current recommendations are to maintain a SBP < 160mm Hg to reduce the risk of rebleeding
- Imaging
  - Diagnostic workup should include noncontrast head CT which, if nondiagnostic, should be followed by lumbar puncture
  - CTA may be helpful, but if inconclusive, angiography is still recommended
  - DSA with 3-dimensional rotational angiography is indicated for detection of aneurysm in patients with aSAH and for planning treatment to determine whether an aneurysm is amenable to coiling

Aneurysmal Subarachnoid Hemorrhage (aSAH) - Treatment

- Treatment
  - Definitive treatment of aSAH is surgical clipping or endovascular coiling. Which procedure is used is largely dependent on the anatomy of the aneurysm
  - Surgical clipping or endovascular coiling of the ruptured aneurysm should be performed as early as feasible in the majority of patients to reduce the rate of rebleeding after aSAH with complete obliteration of the aneurysm when possible
  - For patients with ruptured aneurysms judged to be technically amenable to both endovascular coiling and neurosurgical clipping, endovascular coiling should be considered
  - Repeat imaging following aneurysm clipping or coiling should be done to evaluate for completeness of obliteration

- Management of complications
  - Rebleeding
    - Strict BP management and repeat intervention for aneurysm obliteration are indicated for patients with rebleed
  - Vasospasm/Delayed cerebral ischemia (DCI)
    - Vasospasm and subsequent cerebral ischemia are one of the leading complications following aSAH
    - Oral nimodipine should be administered to all patients with aSAH
    - Maintenance of euvolemia and normal circulating blood volume is recommended to prevent vasospasm
    - Daily monitoring for arterial vasospasm with transcranial doppler is “reasonable”
    - Perfusion imaging with CT or MRI can be useful to identify regions of potential ischemia
    - When vasospasm occurs:
      - Induction of hypertension is recommended for patients with DCI unless blood pressure is elevated at baseline or cardiac status precludes it
      - Cerebral angioplasty and/or selective intra-arterial vasodilator therapy is reasonable in patients with symptomatic cerebral vasospasm, particularly those who are not rapidly responding to hypertensive therapy
  - Hydrocephalus
    - Generally, aSAH-associated acute symptomatic hydrocephalus should be managed by cerebrospinal fluid diversion with IVC/EVD or lumbar drain
    - Seizures
      - The use of prophylactic anticonvulsants should be used in the immediate posthemorrhagic period, but routine long-term use of anticonvulsants is not recommended except for patients with known risk factors, such as prior seizure, intracerebral hematoma, intractable hypertension, infarction, or aneurysm at the middle cerebral
  - Medical complications are known to occur frequently in patients with aSAH

Neuromuscular disorders - Myasthenia Gravis (MG)

- Myasthenia gravis is an autoimmune disease whereby antibodies destroy the neuromuscular function
- Defective neurotransmission leads to fatigable muscle weakness
- Most patients have thymic hyperplasia or thymomas
- Diagnosis
  - Anti-AchR antibodies
  - Edrophonium test
    - Distinguishes myasthenic crisis from cholinergic crisis (too much anticholinesterase)
    - Dangerous in cholinergic crisis though
  - EMG with repetitive stimulation
  - CT to R/O thymoma
- Treatment
  - Respiratory failure
    - Follow VC, NIF, PImax
    - Hypercapnea is late sign
    - Intubation is indicated if VC <12 or significant bulbar dysfunction
  - Anticholinesterases (pyridostigmine)
  - Immunosupressives (e.g. steroids)
  - Thymectomy
  - Plasma exchange
  - IV Ig
  - Avoid drugs that exacerbate condition such as aminoglycosides, macrolides, lidocaine, oral contraceptives, phenytoin, and propranolol among others

Neuromuscular disorders - Guillain-Barre syndrome (GBS)

- An acute self-limited inflammatory, demyelinating polyneuropathy
- Commonly precipitated by an infection (CMV, EBV, or *Campylobacter jejuni)*
- Limb paresthesias are typically the presenting signs with flaccid paralysis that can remain localized or spread to entire body
- Ptosis and bulbar dysfunction are ominous signs for severity of the disease and need for intubation
- Diagnosis
  - Must R/O other causes of flaccid paralysis such as spinal cord injury
  - MRI is typically nonspecific
  - EMG is the most sensitive test with motor neuron conduction block being most typical
  - CSF analysis is nonspecific
  - Ganglioside antibodies are frequently detected
- Treatment
  - Initial steps depend on clinical presentation including, severity of weakness, rapidity of progression, respiratory muscle weakness, dysautonomia, systemic complications, and comorbid disease (23).
  - Intubation is indicated if VC ≤20 mL/kg and PImax ≤-30 mm Hg (23)
  - IV Ig 0.4 g/kg for 5 days
  - Plasma exchange for patients who cannot tolerate IV Ig

Neuromuscular disorders - Critical Illness Polyneuropathy

- Relatively recently describe phenomena that is classically described as
  - Presence of sepsis, multi-organ failure, respiratory failure, or SIRS.
  - Difficulty weaning from ventilator
  - Limb weakness
- Part of a spectrum of disease that has been found in 25-85% of critically ill patients
- Other types include acute myopathy of intensive care and combinations of myopathy and neuropathy
  - 46% of patients admitted with sepsis, multi-organ failure or on prolonged mechanical ventilation go on to develop acquired neuromuscular disorders
- Diagnosis
  - Causes a flaccid paralysis, often predominantly distal
  - Failure to wean from mechanical ventilation is a hallmark
  - Cranial nerves are typically spared
  - EMG studies are diagnostic and demonstrate axonal neuropathy
- Treatment is supportive
  - No specific therapy
  - Aggressive rehabilitation
  - Prevention of SIRS
- Prognosis
  - Recovery is weeks to months
  - No medication therapy, only conservative
  - 50% have complete recovery