Neurology Clerkship “EMERGENCIES” Core Knowledge:

**Introduction:** Emergencies require quick and decisive action with very little time for introspection. Therefore, a physician must be able to recognize potential emergencies and act quickly in order to save a patient’s life. These actions must be reviewed continuously so that a physician acts virtually automatically in such a situation. Potentially lethal diagnoses must always be considered and excluded before any further action is contemplated. As noted in the clerkship handout, emergencies are “vascular”, “electrical” or “traumatic” in nature. This handout addresses the most common neurological emergencies regarding their recognition and the first steps to be taken in every instance.

**General:** The same course of action should be followed in every instance and is abbreviated A-B-C-D-E which signifies Airway, Breathing, Circulation, Drugs and Evaluation. For each and every neurological emergency discussed, it will be assumed that the A-B-C steps will have been taken. The patient’s orders should indicate NPO except medications until the emergency evaluation is complete. The remainder of this document will address drugs and evaluation. The first steps are as follows:

**Airway:** Does the patient have an open airway capable of gas exchange? If not, establish an airway by any means necessary including invasive procedures. Invasive procedures include intubation and tracheotomy. If you doubt your ability to perform these skills then STAT page a colleague who can.

**Breathing:** Is the patient effectively exchanging oxygen for carbon dioxide on their own? If not, then do it for them by whatever means are necessary. This will be confirmed in the “evaluation” stage.

**Circulation:** Does the patient have adequate blood pressure and pulse? If not, decide if the patient has hypovolemia or pump failure. Establish venous access with two large bore intravenous catheters as soon as possible.

**Drugs:** Yes, this includes oxygen! With few exceptions, such as carbon dioxide retention in COPD, most patients will either be helped or at least will not be harmed with supplemental oxygen. For patients who present with a change in mental status, these 3 drugs in this order should always be administered; THIAMINE 100 mg IV followed by Glucose (1 ampule of D50) IV followed by Naloxone (1 ampule) IV.

**Evaluation:** Ask yourself, “What potentially lethal diagnosis could my patient have?” If you cannot eliminate that emergency with 99.9% certainty, then perform the “gold standard” test to exclude that diagnosis.

**Neurological Emergencies:**

I. **Status Epilepticus:** While the standard textbook definition states that this occurs when a seizure has been ongoing for 30 minutes or that the patient does not fully regain consciousness between 2 seizures, this definition has been challenged recently. McDonald et al. suggest that an ongoing seizure lasting more than 5 minutes should be considered as incipient status epilepticus. The authors prefer this definition as irreversible brain damage will occur if a seizure continues for more than 60-80 minutes. Therefore, earlier intervention is preferred. The goal is to load one medication with rapid onset in order to stop the seizure followed by a second medication with a longer duration of action that will continue to prevent the seizure focus from propagating. BE PREPARED FOR EMERGENCY INTUBATION. All medications used will result in some degree of respiratory depression. STAT page anesthesia if you have any intubation concerns and explain why you are consulting this service.
The correct course of action is as follows:

**STOP THE SEIZURES ASAP BY BEGINNING:**
- LORAZEPAM 1-2 mg IV push, if ineffective
- LORAZEPAM 1-2 mg IV push, if ineffective; INTUBATE and administer
- LORAZEPAM 0.5-1 mg/kg IV FOLLOWED BY:
  - FOSPHENYTOIN 20 phenytoin equivalents/kg administered over 10 minutes

**CONSULT NEUROLOGY IF SEIZURES HAVE NOT STOPPED OTHERWISE:**

- STAT CT SCAN WITHOUT CONTRAST to evaluate for an intracranial anatomic abnormality
- CSF examination if head CT is NORMAL and there is ANY suspicion of meningitis

**II. Stroke** (Ischemic and hemorrhagic): A stroke is defined as an acute onset of neurological dysfunction due to an interruption in vascular perfusion. The vast majority of strokes are usually associated with NEGATIVE phenomena, (loss of strength, loss of sensation, loss of vision). POSITIVE phenomena (tingling, buzzing) may indicate loss of electrical transmission as experienced in migraine or demyelinating disease. The most important distinction to make is between ischemic and hemorrhagic stroke as ischemia stroke may respond to clot lysis whereas hemorrhagic strokes may worsen. The outcome from ischemic stroke is inversely related to intervention time. The faster one intervenes, the better the outcome. However, peripherally administered intravenous clot lysis agents are NOT effective after 3 hours of stroke onset and radiologically guided lysis is not effective after 6 hours. TIME IS BRAIN! The correct course of action within the first 3 hours of symptom onset is as follows:

- STAT HEAD CT WITHOUT CONTRAST, [contrast has the same appearance as acute blood]
- If bleeding is noted, STOP! Do not use clot lysis agents (e.g. TPA)
- If no bleeding is noted, determine if the patient is a candidate for TPA, (see stroke guidelines).
- If a TPA candidate, administer TPA per stroke protocol guidelines, (per hospital protocol)

**III. Worst Headache ever:** While it is true that one headache may be “the worst headache ever,” the physician must be concerned about any headache that is out of proportion to headaches experienced previously. The most important diagnosis not to be missed is that of meningeal irritation due to blood or infection. The penalty for missing such a disease process is so severe that any suspicion of a meningeal process should prompt the physician to perform appropriate testing. The classic meningeal signs described by Kernig and Brudzinski are very insensitive and should not be used for diagnosis. Any headache associated with a fever is meningitis until proven otherwise. In a patient who is immunocompromised, a fever may not be present. Any headache of acute onset, especially during vigorous activity is a subarachnoid hemorrhage from a ruptured aneurysm until proven otherwise. If there is ANY doubt, then these studies MUST BE performed:

- STAT HEAD CT WITHOUT CONTRAST
- ESR if there is any suspicion of temporal arteritis
- LUMBAR PUNCTURE if head CT is normal

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The lumbar puncture should include a cell count on tubes one and three. The normal range of WBCs in a non-traumatic lumbar puncture is 1-5. One WBC is expected for every 1,000 RBCs in cases of a traumatic lumbar puncture. The laboratory should spin down the fluid and decant the supernatant. This fluid is examined in a white light against a white background. The presence of a yellowish tinge indicated “xanthochromia” or yellowing of the CSF due to the presence of bilirubin from ruptured RBCs. This finding is present from 6 hours-3 weeks after an aneurysm has ruptured. The CSF protein should be 15-45 mg/dl but an increase of 1 mg/dl may be attributed to 1,000 RBCs. Elevated CSF protein values indicate a neuro-immunological reaction. It is important that the patient have a finger stick to check blood glucose at the time the LP is performed so that serum and CSF glucose can be compared to this value. The CSF glucose should be 2/3 or more of the CSF glucose or an infection should be suspected. Gram staining, CSF VDRL, bacterial antigen studies and a cryptococcal antigen should always be part of a routine CSF evaluation.

If “xanthochromia” is discovered then the following course of action is required:

- ELEVATE THE HEAD OF THE BED TO 30 DEGREES, (minimize intracranial pressure)
- ADMINISTER NIMODIPINE 60 mg p.o. q 4 hours, (reduce vasospasm)
- GIVE COLACE 100 mg p.o. b.i.d., (avoids straining while defecating)
- 4 VESSEL CEREBRAL ANGIOGRAM, (identify the aneurysm)
- NEUROSURGICAL CONSULTATION, (they will have to clip it)

If meningitis is discovered then the following course of action is required:

- CEFTRIAXONE 1 GRAM IV q 12 hours; FIRST DOSE STAT.
- VANCOMYCIN 1 GRAM IV q 12 Hours; FIRST DOSE STAT
- Consider the use of dexamethasone 0.4 mg/kg IV
- Consider an infectious disease consult

**IV. Acute Motor unit weakness to include botulism, myasthenia and Guillain-Barré syndrome:**

These disorders all manifest weakness with diminished reflexes and decreased tone unlike upper motor neuron lesions that will eventually manifest increased reflexes and increased tone. The goal is to localize the lesion and intervene appropriately. Step one, IS THERE ANY SENSORY DISTURBANCE? If the answer is “yes”, then the problem is in the peripheral nerves as NEITHER muscle diseases NOR neuromuscular junction disorders ever have any sensory symptoms. If there are sensory disturbances, then treat this patient as if they have GBS, (Guillain-Barré syndrome). The correct course of action is as follows:

- ADMIT TO THE PCU or the ICU
- Check forced vital capacity (FVC) q 6-8 hours
- INTUBATE when FVC < 15 ml/kg
- NEUROLOGY CONSULTATION
If there are no sensory signs or symptoms then consider neuromuscular junction [NMJ] and muscle diseases. NMJ disorders usually wax and wane in intensity and are rarely static and progressive. NMJ disorders commonly affect the cranial nerve muscles whereas muscle diseases hardly ever do. Muscle diseases may be associated with an increase in serum muscle enzymes, especially CPK (creatine phosphokinase) whereas this NEVER occurs with NMJ disorders. No patient should ever succumb to an acute NMJ disorder or to an acute muscle disease. The most important NMJ disorders can be divided into pre-synaptic and post-synaptic disorders. Pre-synaptic disorders IMPROVE with EXERCISE and WORSEN with rest, (e.g. botulism or LEMS). By contrast, post-synaptic disorders WORSEN with EXERCISE and IMPROVE with REST. The most commonly encountered post-synaptic disorder is myasthenia gravis, (MG). Unlike other post-synaptic disorders such as organophosphate poisoning, MG is NEVER associated with autonomic signs and symptoms whereas organophosphate poisoning would be associated with excess salivation, lacrimation, urination, perspiration and defecation. The correct course of action for NMJ disorders is:

ADMIT TO THE ICU

INTUBATE when FV < 15 mg/kg

NEUROLOGY CONSULTATION

Muscle diseases rarely occur acutely. Those that do are usually associated with acute muscle infarction from vascular compromise. The danger is that the myoglobin molecule which contains iron that may cause acute tubular necrosis and kidney failure. The diagnosis must be suspected and confirmed by evaluating the urine. The urine will test dipstick positive for “blood” as iron from myoglobin is present but the urinalysis will not reveal RBCs. The serum will also demonstrate elevated CPK values. The correct course of action is as follows:

ADMIT TO THE ICU

Infuse as much IV normal saline as can be tolerated

Alkalinize urine with one ampule of bicarbonate per liter of normal saline

NEUROLOGY AND NEPHROLOGY CONSULTATION

V. Neuroleptic malignant syndrome/Drug induced Parkinsonism: A simplistic, although clinically useful, view of Parkinson’s disease is that the signs and symptoms result from a relative deficiency of dopamine within the central nervous system. Dopamine’s effects are usually opposed by those of acetylcholine. Therefore, Parkinson’s disease could be viewed as being either a relative dopamine deficiency or a relative excess of acetylcholine activity. In fact, anticholinergic agents are more effective than dopamine agonists at reducing Parkinsonian tremor. By contrast, a simplistic model of psychosis is that schizophrenia and other delusional disorders represent the effects or excess dopamine or decreased effect of acetylcholine. This model is supported by the observation that anticholinergic agents, e.g. jimsonweed, present with acute psychosis. Conversely, dopamine blocking agents reduce the symptoms of psychosis but are prone to inducing Parkinsonism. The most extreme form of Parkinsonism is neuroleptic malignant syndrome or NMS. This is usually the result of exposure to dopamine blocking agents although paradoxical reactions to dopamine agonists such as metaclopramide (Reglan) have been reported. The patient presents with acute Parkinsonian features, (expressionless face, rigidity) but tremor is rare. The classic triad of elevated WBC count, elevated CPK and fever is not always present. Therefore, the diagnosis must be suspected in anyone with new onset Parkinsonism after having been administered a drug that affects dopamine. While this is usually a neuroleptic medication, be aware that this is not invariably the case.
The correct course of action is:

STOP the offending DRUG

Insert an NG tube as most patients cannot swallow

Bromocryptine (a dopamine agonist) 5 mg per NG tube q.i.d.

NEUROLOGY CONSULTATION

This should not be confused with an acute dystonic reaction in which the patient suddenly becomes contorted into a rigid posture although the underlying cause is similar. The correct course of action is to administer DIPHENHYDRAMINE 50 mg IV once.

VI. Hypertensive Encephalopathy/Eclampsia: Hypertensive encephalopathy is defined as an elevated mean arterial blood pressure associated with cognitive changes and/or neurological deficits. The exact pathophysiology is incompletely understood but the course of action requires gradual blood pressure reduction. Most patients with this disorder have been chronically hypertensive. Thus, the brain has compensated for increased intra-arterial pressure such that aggressive reduction of blood pressure may precipitate a stroke. The goal is to reduce the blood pressure slowly over 24 hours to achieve a mean arterial pressure of 110 mm Hg. Since treatment of hypertensive encephalopathy varies by institution, consultation with a critical care specialist is essential.

Eclampsia is usually preceded by pre-eclampsia. Pre-eclampsia is defined as the triad of weight gain, edema and hypertension in a pregnant woman. Eclampsia occurs when pre-eclampsia leads to tonic-clonic seizures. Magnesium has proven superior to anticonvulsants in the situation so anticonvulsants should not be administered as a first line agent. The definitive treatment for eclampsia is to deliver the baby.

VII. Stupor and Coma/Change in Mental Status: Stupor defines a state of decreased consciousness resulting in a decreased ability to interact with the environment. Coma is defined as a state wherein the patient is so obtunded that there is no purposeful interaction with the environment and that any movements observed are essentially disinhibited reflexes. While the boundary between stupor and coma is indistinct, the clinical approach is essentially the same. A patient with decreased consciousness should be evaluated in a similar manner. The first course of action is to determine if the patient’s examination is symmetric or asymmetric. Asymmetric or focal deficits are usually the result of central nervous system dysfunction whereas symmetric deficits usually indicate that the brain is not functioning due to some metabolic derangement. Restated, “If there is a focal deficit then blame the brain. If there is no focal deficit then the brain is an innocent bystander to some metabolic derangement.” You will observe that “change in mental status” accounts for numerous unnecessary neurological consultations. If there is a focal deficit then the following course of action should be pursued so that the cause of primary brain dysfunction is identified:

HEAD CT WITHOUT CONTRAST

LUMBAR PUNCTURE if there is any suspicion of CNS infection and head CT is normal

EEG (provides an electrical map of brain function)
If there is NO FOCAL ABNORMALITY, then the brain is likely an innocent bystander to a metabolic derangement.  The following course of action is recommended:

- HEAD CT WITHOUT CONTRAST (exclude occult frontal lobe pathology)
- LUMBAR PUNCTURE if there is any suspicion of CNS infection and head CT is normal
- EEG if there is ANY suspicion of non-convulsive status epilepticus
- CBC to evaluate for infection
- TOXIC SCREEN to evaluate for licit and illicit drugs
- LIVER PANEL and METABOLIC PANEL
- ABG
- Serum TSH, B12 and Folate levels

Dementia should NEVER be diagnosed in anyone who has a change in mental status.  Dementia assumes that the person has impaired cognitive function when maximally awake and alert.

**VIII. Increased intracranial pressure:**  The Monroe-Kellie hypothesis states that the sum of brain tissue pressure, CSF pressure and intravascular pressure is a constant.  Signs and symptoms of increased intracranial pressure (IIP) will occur when the brain can no longer compensate for an increase in pressure in any of these factors.  Since venous and CSF drainage is improved when upright, many patients with IIP will have their most severe symptoms while lying down.  These symptoms can include headache, nausea and vomiting.  Earlier stages of IIP are hardly ever associated with visual loss.  The most common sign of IIP is that of papilledema wherein the optic disk appears swollen.  The treating physician must determine the cause of IIP and decide if it is intravascular, CSF related or brain substance related.  The most common brain substance related cause of IIP is a neoplasm.  CSF overproduction rarely occurs whereas CSF obstruction is more common.  Causes include ventricular obstruction due to tumors, intracranial hemorrhage and meningitis.  Vascular causes can include malignant arterial hypertension or a reduction in venous drainage due to an intracerebral venous thrombosis.  Malignant hypertension has been previously discussed.  The causes of venous thrombosis include adjacent infection and hypercoaguable states.  The immediate course of action is as follows:

- HEAD CT WITHOUT CONTRAST
- LUMBAR PUNCTURE if there is any suspicion of CNS infection and head CT is normal
- MRI of the brain with MR angiography and MR venography
- If a venous thrombosis is identified, then investigate for hypercoaguable states
- If no cause is identified, consider the diagnosis of idiopathic intracranial hypertension

**IX. Acute MONOCULAR Visual Loss:**  Patients with acute visual loss usually consult an ophthalmologist but all physicians should know how to evaluate these patients.  The physician should determine if one eye or both eyes are affected.  In a patient who had normal vision in both eyes before the event, monocular disorders indicate a lesion ANTERIOR to the optic chiasm.  Loss of vision in both eyes, even if incomplete, indicates a lesion behind the optic chiasm and in the brain.  Most instances of acute visual loss will affect only one eye.  At the very least, the physician should examine the red...
reflex, the pupillary reflex, extraocular movements and measure intraocular pressure. If the physician
is uncomfortable with this, then an ophthalmologist should be consulted STAT. The physician must
identify the site of visual dysfunction by dividing the pathway into the orbital structures (eye and
surrounding tissues), the retina and the optic nerve. Intra-ocular causes include corneal opacities,
acute glaucoma, and eye trauma (e.g. hyphema). Acute monocular visual loss WITH extraocular eye
muscle dysfunction identifies that the lesion is within the orbit. Isolated monocular visual loss
WITHOUT extraocular muscle abnormalities may be located in any site anterior to the optic chiasm.
Do not dilate the pupil unless advised to do so by the ophthalmologist. If the ophthalmologic exam is
unremarkable, then the lesion is behind the retina and falls within the neurologist’s purview. The
correct course of action for MONOCULAR VISUAL LOSS and a normal examination performed by an
ophthalmologist:

PALPATE THE TEMPORAL: ARTERIES AND OBTAIN AN ESR (temporal arteritis?)

MRI OF THE AFFECTED ORBIT AND NERVE

If all the above are normal, consider a diagnosis of optic neuritis

X. Acute BINOCULAR Visual Loss: The lesion is usually posterior to the chiasm and should be
evaluated as if it were a new onset stroke. There are exceptions to this rule such as anterior ischemic
optic neuropathy but this can be evaluated after a stroke has been excluded.

XI. Intracranial aneurysm: Intracranial aneurysms that have ruptured usually present as acute onset
of the worst headache that the patient has ever experienced. However, the physician must diagnose
unruptured aneurysms as well. Aneurysms result from a defect within the tunica media of the arterial
wall. The vast majority of intracranial aneurysms are located within the circle of Willis. Two common
locations are in the anterior communicating artery [ACA] joining the two anterior cerebral arteries and in
the posterior communicating artery [PCA] that joins the middle and posterior cerebral arteries. ACA
aneurysms are usually discovered accidentally. However, large ACA aneurysms may present with
symptoms of optic chiasm or pituitary dysfunction. By contrast, PCA aneurysms are adjacent to the
oculomotor nerve and may present with signs or symptoms of III nerve dysfunction. The
parasympathetic fibers are located on the outside of the III nerve. Thus compressive lesions produce
pupillary abnormalities. By contrast, ischemic lesions usually spare the parasympathetic fibers and
thus pupillary function is normal. A complete pupil-sparing III nerve palsy is not likely to be due to a
PCA aneurysm. However, any pupillary abnormality with oculomotor dysfunction is a PCA aneurysm
until proven otherwise. The correct course of action for any suspected aneurysm is as follows:

ELEVATE THE HEAD OF THE BED 30 DEGREES

COLACE 100 mg p.o. b.i.d.

NIMODIPINE 60 mg p.o. q 4 hours

4 vessel cerebral angiogram (aneurysms may be multiple)

NEUROSURGICAL CONSULTATION

XII. Acute Myelopathy: Spinal cord compression should always be suspected in any acute
myelopathy. While many patients have a known history of a primary carcinoma, this may be the first
indication of a cancer. Spinal cord compression can also occur from trauma or centrally herniated disks
to name causes other than cancer. Most patients present with acute neck or back pain followed by
symptoms of acute spinal shock. Acute spinal shock presents with some combination of bilateral limb
paralysis, urinary retention and often a sensory level. Unilateral limb weakness is NOT a common
manifestation of spinal cord compression. The immediate course of action is as follows:
PLAIN X-RAYS OF THE SUSPECTED LEVEL, (bony lesions/fractures can be noted)

MRI WITHOUT THEN WITH GADOLINIUM AT THE SUSPECTED LEVEL

NEUROSURGICAL CONSULTATION IF ANY ABNORMALITY IS NOTED

DISCUSS THE USE OF IV METHYLprednisolone WITH THE NEUROSURGEON

IF NO LESION IS NOTED THEN CONSULT NEUROLOGY

XIII. Acute Wernicke’s encephalopathy: While most commonly seen in “skid row alcoholics”, Wernicke’s encephalopathy actually results from a thiamine deficiency. Therefore, alcoholism is not a prerequisite for this disorder. Patients who are thiamine deficient for any reason may be prone to this condition. The classic triad of inability to form new memories, ataxia and extraocular muscle palsies is rarely seen in its entirety. Since the outcome is so devastating, any patient with an acute change in mental status ought to be administered the relatively benign medications of thiamine 100 g IV followed by glucose. Thiamine is always administered first because it is a cofactor for glucose metabolism. It is thus theoretically possible to precipitate Wernicke’s encephalopathy with initial glucose administration. Since these agents are inexpensive, it is a good practice to administer thiamine followed by glucose to anyone with an acute change in mental status.

XIV. Sedative withdrawal and delirium tremens: Sedative withdrawal is the preferred term as abrupt discontinuation of barbiturates, benzodiazepines and alcohol manifest in a similar manner. When the patient is no longer receiving large doses of these medications, a withdrawal state occurs. The patient becomes tremulous, agitated and irritable. In many instances, reinstitution of the agent at a slightly reduced dose followed by a gradual taper is all that is required. This is NOT the same as delirium tremens in which a patient becomes extremely agitated and experiences hallucinations. These hallucinations vary from insects crawling on the skin (formication) to more complicated delusions similar to schizophrenia. The correct course of action is to keep the patient comfortably sedated so that he or she cannot act on these delusions. There are no evidence based medicine guidelines but most physicians tend to use lorazepam as it can be administered IV, IM or p.o. The dose of lorazepam should be titrated such that the patient is somnolent but arousable. The patient should be placed in a PCU or other such environment where close observation is possible. Delirium tremens is fatal in approximately 10-15% of patients.

Daniel L. Menkes, MD, FAAN
University of Connecticut