The Finseth Review
Brief Cases in Clinical Neurology

Sections (click on link to jump to section):

Note: Index includes rudimentary ratings of the relative importance of topics.

Statement of Purpose

This review is intended to supplement your medical education in neurology with the primary purpose of preparing you for the Clinical Neurology "Shelf" Exam and the Neurology portion of USMLE Step 2 Exam. It is also our hope that you come away with knowledge that will help you take care of patients with neurological problems in your future career. We have focused on the topics most likely to be tested on your "Shelf": 1)common problems, their pathophysiology, diagnosis and treatment 2)emergent problems and the next step and 3) clinical syndromes that have pathognomonic characteristics, making them easily testable. We hope that seeing these clinical syndromes in vignette format will help you recognize them on your shelf and accurately diagnose them in your future patients. While the diagnostic workup and treatment plans referred to in the review should suffice for your test and are up to date to the best of our knowledge (as of 1/2011), you should refer to medical references when diagnosing and treating patients. Best of luck to you in your clerkship and beyond.

Headache
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H1. A 23 yo M comes to the ER after he suddenly develops the worst headache of his life. He states he had a brief but terrible headache about a week ago while doing some yard work.

What is it? Subarachnoid Hemorrhage secondary to aneurysmal bleed. The prior self-limited headache was a sentinel bleed of the aneurysm. The hyperacute rapid onset

(thunderclap headache) is a clinical red flag for SAH rather than a migraine that evolves more slowly.
How is it diagnosed? CT scan without contrast. Contrast is held so that blood will show up bright white on the scan (be able to recognize blood in the subarachnoid space).
Treatment? Your next step would be angiography to locate the aneurysm and a neurosurgical consult. This is done because the aneurysm can continue to bleed leading to coma/death or if the bleeding has stopped, there is a significant chance of rebleeding for the next 2 weeks, and in particular in the next 24 hours. Nimodipine is given prophylactically to prevent the complication of cerebral vasospasm 4-14 days later (which can cause ischemic injury).

H2. A 23 yo M comes to the ER after he suddenly developed the worst headache of his life. He states he had a terrible headache about a week ago while doing some yard work. CT scan of the head is negative.

What is it? It's still a SAH due to aneurysmal bleed until proven otherwise. CT misses roughly 10% of SAHs.
Next step? Perform a lumbar puncture. A positive lumbar puncture would show xanthochromia (yellow color to fluid from RBC breakdown) or RBCs that do not clear from tube 1-4.
Treatment? As described above.

H3. A 75 yo female presents to the ER with pain in her right temple. She complains of some arthritis in her shoulders and hips as well for the past year. Her scalp is tender to the touch. Lab studies show a mild microcytic anemia and markedly elevated ESR.

What is it? Temporal arteritis in a patient with polymyalgia rheumatica.
Next step? High dose steroids. Steroids should be given immediately to prevent vascular occlusion of the temporal artery and permanent loss of vision to the eye. If vision loss has already occurred, steroids are still given to prevent vision loss on the contralateral side.
Diagnosis? Elevated ESR and temporal artery biopsy.

H4. A 27 yo M comes in with unilateral right neck and facial pain. The patient has trouble talking to you and has trouble finding words. His friend relates that they were playing tennis earlier in the day, and he was hit in the neck with a tennis ball. He had finished the game without much complaining but later was complaining of pain. On exam, you notice the patient has ptosis and a constricted pupil on the right.

What is it? Carotid Dissection resulting in Horner's syndrome and ischemic stroke. Horner's and stroke need not be present but can be sequelae.
Diagnosis? MR or CT Angiography or conventional angiogram.
Treatment? Probably best to just recognize this on your shelf. Anticoagulation vs antiplatelet therapy would be initiated to prevent formation of thrombus in the dissection.

H5. A 45 yo F migraine patient presents with a chronic daily headache for the past 3 months. The headache is diffuse and dull and occurs everyday. She states she was taking frequent analgesic pain meds for migraines and then slowly developed a daily, dull headache that won't go away. She is currently taking oxycodone every 4-6 hours, which only partially relieves her headache. Her pain has prevented her from achieving good sleep but does not wake her from sleep. Fundus exam shows no papilledema. No

nocturnal worsening or symptoms. No worsening of symptoms with straining. No focal neurological deficits or visual changes.

What is it? Analgesic rebound headache.
Diagnosis? Made clinically based on chronic, frequent use (3 days a week or more) of abortant or analgesic pain medications transforming intermittent headaches into a chronic daily headache. There also should not be any red flag symptoms for tumor/increased ICP or other secondary cause of headache.
Treatment? Cessation of frequent use of acute analgesic medications and transitioning to medications without analgesic rebound potential.

H6. A 45 yo M presents with a headache that has gradually worsened for the past two months. He describes a pressure like pain that is worse in the morning but persists throughout the day. Fundoscopic exam shows papilledema.

What is it? Intracranial mass leading to increased intracranial pressure. Any subacute progressive headache should raise suspicion for a brain tumor. Pain that is worse in the morning/middle of the night after being supine and papilledema are signs of increased ICP.
Diagnosis? MRI with contrast to look for tumor.
Treatment? If MRI +, therapy will potentially involve chemotherapy, radiation, surgery or hospice care. Dexamethasone would be indicated if symptomatic mass effect was present. If multifocal pattern on MRI suggesting metastasis, search for a primary cancer should follow with lung, melanoma, and breast as the most likely culprits.

H7. A 65 yo M presents to the hospital with headache for the past 12 hours and confusion. He has had a fever up to 103. On exam, patient complains of pain with neck flexion and flexes his hips when you perform this maneuver. No focal signs are present. Multiple ecchymotic/petechial patches are present over trunk and extremities.

What is it? Bacterial meningitis. Any patient with fever, headache, and neck stiffness should be suspected of having meningitis. His skin findings make N. Meningitidis as the likely culprit for his infection.
Diagnosis? Lumbar puncture with glucose, protein, WBC, RBC and gram stain, and cultures. Blood cultures would be drawn as well. If focal signs were present, an abscess might be present leading to increased ICP, and therefore a head CT should be performed prior to LP to avoid herniation, though empirically treating with antibiotics should not be delayed while waiting to be able to obtain LP with clinical suspicion of bacterial meningitis. Bacterial meningitis shows depressed glucose with elevated white count and a neutrophil predominance. Viral would have a near normal glucose with elevated white count and a lymphocytic predominance.
Treatment? Empiric IV antibiotics - specifically ceftriaxone for bacterial meningitis, with vancomycin initially to cover penicillin-resistant strains while csf and blood cultures are pending. Ampicillin added for certain populations, such as the elderly, immunocompromised, etc.

H8. A 33 year old male presents to the ED with a severe headache and confusion. The headache is severe and constant and has been progressing for the past 24 hours. It was submaximal at onset. He denies fever or neck stiffness. Vital signs reveal he is afebrile with pulse of 120, BP 210/130. Exam reveals papilledema. Lab studies are remarkable for creatinine of 2.3. CT and LP are negative.

What is the most likely diagnosis? Hypertensive encephalopathy. This is a diagnosis of exclusion but suspect in a patient with elevated blood pressure, papilledema and without signs of ICH, stroke or meningitis.
Treatment? Acute reduction in blood pressure by 25% and diastolic to 100-110. Labetolol and nicardipine are preferred agents.

Of note, acutely elevated BP/hypertensive encephalopathy may cause posterior signal on an MRI of the brain and is called PRES (posterior reversible encephalopathy syndrome). In addition to headache and confusion, cortical visual disturbance and seizures can be seen. The treatment is similar to above. This can be seen in eclampsia, as well, and even in normotensive patients on cytotoxic agents such as cyclosporine.

H9. A 25 yo obese F presents with a constant headache for the past few months. She reports that headaches are worst in the morning. She also reports sudden loss of vision for 1 to 2 seconds that occurs intermittently as well as ringing in her ears that coincides with her heartbeat. Fundus shows papilledema. An emergent head CT shows no abnormalities.

What is it? Pseudotumor cerebri (idiopathic intracranial hypertension). This condition tends to occur in young, obese females who will complain of chronic headaches and may have associated symptoms of transient visual obscurations (loss of vision lasting seconds) or pulsatile tinnitus or gradual visual field loss if severe. Look for Isotretinoin use in the vignette, as the drug can cause this condition as a side effect.
Diagnosis? Head CT is performed to rule out a mass lesion or hydrocephalus (dilated ventricles). LP with elevated opening pressure (greater than 20-25 for diagnosis and potentially much higher). Formal visual field evaluation could confirm visual field loss.
Treatment? LP can be temporarily therapeutic by withdrawing fluid to normalize pressure. Acetazolamide is a first line medical therapy that can decrease CSF production. Lasix may also be used. Weight loss may help. In cases with visual field loss, optic nerve sheath fenestrations should be considered to relieve pressure on the optic nerve and prevent further vision loss. Ventriculoperitoneal shunts may be considered in refractory cases.

H10. A 30 yo F patient is admitted for left sided paresthesias. Yesterday, while in the hospital, she had an MRI, which showed some demyelination and lumbar puncture which showed oligoclonal bands. While discussing with her the possibility of multiple sclerosis, she complains to you that she has been having a severe headache every time she gets up to use the restroom since yesterday. Once she returns to her bed and calls the nurse, her headache is always better when the nurse arrives.

What is it? Post-lumbar puncture headache (intracranial hypotension headache). The loss of fluid from the lumbar puncture and continued dural leak will cause a low pressure in the CSF that can lead to headache while sitting/standing that is relieved by lying flat.
Diagnosis? Usually clinical. An MRI could show diffuse meningeal enhancement +/- sagging of cerebellar tonsils (similar to Chiari malformation) on sagittal views.
Treatment? The process is typically self-limited and will generally resolve within 2-5 days. Fluids and bedrest will often be enough. Caffeine (cerebral vasoconstrictor) may provide some relief. A blood patch placed in the epidural space could be utilized for closing off refractory dural leaks.

H11. A 22 yo F complains of intermittent, severe throbbing headaches for the past year. She states the headaches are preceded by visual scintillations for 10 minutes, and then the pain occurs over her entire right skull. She states the headaches make her "incapacitated" and the only thing that helps is lying in a dark, quiet room for the next 5-10 hours.

What is it? Migraine
Diagnosis? Clinical. Criteria includes: 1) lasts 4-72 hours 2) has 2 of the following: unilateral, pulsating, moderate to severe, aggravated by physical activity 3) has 1 of the following: nausea, photophobia or phonophobia.
Treatment? As needed NSAIDs or acetaminophen/aspirin with caffeine (Excedrin) are often effective. Triptans (5HT1A agonists) can be an effective abortive medication in patients without contraindications (including CAD, stroke, uncontrolled hypertension). Preventive strategies to avoid migraine triggers such as stress, certain foods, sleep deprivation are important. If migraines are either severely disabling (missing work frequently) or occur greater than 4 times/month, daily prophylactic medication should be considered and can include propranolol, TCAs, depakote, or topamax.

H12. A 55 yo M with history of migraine headaches presents with a severe, sudden onset headache for the past 4 hours that is unrelieved by two doses of triptan medication. He states this headache is more diffuse than his usual migraines, was not preceded by his typical visual aura, and came on "all of a sudden" unlike his typical migraine pain that evolves to maximum intensity much more slowly.

What's the point? A secondary cause of headache should be suspected in a migraine patient when it is different in character from typical attacks (i.e. sudden, thunderclap onset, different location without typical aura, focal deficits that the patient has not had before from a diagnosis of complicated migraine etc for this patient) or has concerning features for secondary causes. A SAH is a potential for this patient.
Diagnosis? Emergent head CT should be performed to look for evidence of SAH or stroke.
Treatment? As discussed previously for SAH. (H1) DO NOT provide reassurance or administer sumatriptan.

H13. A 35 yo M complains of occasional headaches after work. He describes the headaches as if his head is in a "vice grip". He usually gets them towards the end of the work day, but is able to work through them as they are relatively mild and they last until after he has cooked dinner.

What is it? Tension-type headaches. While a very common type of headache, they are unlikely to show up on your exam.
Diagnosis? Clinical. These headaches are typically mild to moderate and described as a band of pressure around the head. Be sure to differentiate from analgesic rebound headache (H5).
Treatment? These medications typically respond well to prn OTC NSAIDS or a daily TCA. Avoidance of triggers and relaxation techniques can also be of help.

H14. A 33 yo M presents to your office in the midst of a severe headache. He describes his pain as a sharp, stabbing feeling behind his right eye. He states he has had these

headaches every day around the same time and they wake him up from his afternoon nap. On exam, you note tearing of his right eye and droopiness of his right eyelid.

What is it? Cluster headaches
Diagnosis? Clinical. The headaches will typically occur around the same time each day for a period of weeks or months and are severe. They last from around 15 minutes to 3 hours. They can be accompanied by autonomic symptoms on the ipsilateral face (tearing, runny nose, flushing, Horner's syndrome).
Treatment? 100% oxygen via a nonrebreather mask for 15 minutes will relieve this headache in ~70% of patients. It is unlikely to be asked but subcutaneous sumatriptan is also effective and there is evidence that certain medications may help in prophylaxis, such as a prednisone short term prophylaxis or verapamil for chronic prophylaxis among others.

H15. A 60 year old man complains of extremely severe, sharp, shooting pain on the right side of his face. The pain only lasts a few seconds but is so painful he has afraid to go outside because the wind across his face can set it off.
What is it? - Tic doloreaux (trigeminal neuralgia). It is a neuropathic pain that is commonly from an abnormal vessel loop contacting the trigeminal nerve at the base of the brain or rarely from other secondary causes like multiple sclerosis.
Diagnosis? Clinical.
Treatment? Treat with carbamazepine, which is successful for most patients. Refractory cases may require interventional block of the trigeminal ganglion (injections or gamma knife radiation). A craniotomy can be considered in refractory cases to separate the vessel loop from the trigeminal nerve.

H16. A 40 year old male auto mechanic presents for acute headache, dizziness and nausea. He states his symptoms gradually progressed over the past six hours as he worked in the auto shop. He states many of the other mechanics are feeling similarly. Exam shows tachycardia, tachypnea and pallor. Pulse oximetry reads 97%.

What is it? Suspect carbon monoxide poisoning.
Diagnosis? Measure percentage of carboxyhemoglobin in blood. Values vary among smoking status, but greater than 10% is typically more clearly abnormal (non smokers typically have levels less than 3% normally while smokers may have levels up to 10% or so). Note that pulse oximeters read falsely high in the presence of carboxyhemoglobinemia.
Treatment? 100% oxygen to displace CO from Hgb. Hyperbaric oxygen should be considered for significant elevations in carboxyhemoglobin, but no clear guidelines exist.

Traumatic Head Injury
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T1. A 30 yo M falls off his bike unhelmeted. He loses consciousness for approximately a minute but regains consciousness and finishes his ride home. While he is telling his wife about his accident, he gradually loses consciousness and is unarousable. In the Ed, his exam shows a fixed and dilated left pupil in the "down and out" position.

What is it? Acute epidural hematoma. The giveaway is the lucid period after initial loss of consciousness. Epidural bleeds are most commonly from trauma to the middle

meningeal artery. Ocular findings can be a late finding related to a compressive 3rd cranial nerve lesion from herniation from the pressure building from the expanding epidural hematoma.
Diagnosis? Head CT will show crescent biconcave (lens-shaped) bleed.
Treatment? Neurosurgical consultation with emergent craniotomy. High risk for death within hours if untreated in many cases. If craniotomy is done in time, patient may have full recovery.

T2. A 70 yo F is brought in by his out of town son for mental status changes. The patient has poor concentration and appears drowsy. The son notes that his mother had been in a car accident weeks before but seemed okay afterwards. He notes that she has seemed "out of it" on recent telephone calls.

What is it? Chronic subdural hematoma. This patient initially bled during her car accident. Subdural hematoma is due to bleeding from the bridging veins and is more common in the elderly from increased stress on the veins secondary to brain atrophy.
Diagnosis? CT scan shows a crescent-shaped bleed along the skull.
Treatment? Craniotomy

T3. A 50 year old male is involved in a high-speed motor vehicle accident. EMS finds him unconscious with bilateral orbital ecchymoses.
What is it? Basilar skull fracture. Any history of trauma with resultant "raccoon eyes", clear fluid dripping from the nose (rhinorrhea), clear fluid dripping from the ear (otorrhea) or ecchymosis behind the ear (Battle's sign) suggests basilar skull fracture.
Diagnosis? CT scan to view fracture and any intracranial bleed.
Treatment? Neurosurgical consult for evacuation of any bleed. Prophylactic IV antibiotics may be necessary given that there is a direct communication between the environment and the intracranial cavity.

T4. A 35 yo male is a restrained victim of a high-speed motor vehicle accident. He has a short loss of consciousness and short period of confusion but comes back to baseline quickly except he continues to have a severe headache with associated nausea and vomiting. A CT scan is performed, that is unremarkable. He is seen in clinic days later with continued headache, dizziness and photophobia.

What is it? Post-concussive syndrome induced by mild head injury. This syndrome is characterized by a spectrum of non-specific neurological findings, including headache, irritability, mild cognitive difficulties, dizziness, depression and/or anxiety.
Diagnosis? Clinical with exclusion of other more serious entities via exam or imaging.
Treatment? Brain rest until symptoms improve. This includes avoiding any activities requiring concentration including watching television, driving, or attending school. Patient will need to be cleared before resuming athletic events such as with a football injury.

T5. A 24 yo female complains of pain in her arm. She sustained a crush injury to her arm a month ago and had a throbbing pain related to that, but also has had a constant burning pain in her arm that won't go away that has not responded to NSAIDs or high-level opiates. On exam, she will not let you touch her arm because the slightest touch causes an agonizing pain. You note that the arm appears diaphoretic, erythematous, and swollen.

What is it? - Complex Regional Pain Syndrome (formerly known as Reflex Sympathetic Dystrophy)
Diagnosis? Clinical
Treatment? Complex without much evidence to support it. Sympathetic block may be considered or anticonvulsants/steroids for symptom control.

Vascular Neurology
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V1. – A 70 year old right handed man has had 2 transient episodes of weakness on his right side. The episodes completely resolve after about 5 minutes. Fundus shows cholesterol deposits in the left retinal artery.

What is it? – Transient ischemic attacks (TIAs) in the territory of the left carotid artery. It is most likely that the patient has stenosis/plaque of the carotid artery, most commonly found at the bifurcation of the common carotid artery.
How is the diagnosis confirmed? - CTA, MRA, carotid duplex, or angiogram.
Treatment: Carotid endarterectomy (CEA) should be considered and will depend on the grade of stenosis found on imaging. In symptomatic patients, CEA is clearly beneficial for patients with stenosis >70%. Antiplatelet agents and modification of vascular risk factors should also be undertaken.

V2. – A 70 yo M tells you about repeated episodes when he suddenly feels the room spinning, his vision becomes blurry, and his speech becomes slurred. He states that these episodes only last about 15 minutes and then completely resolve. He reports no associated headache with these episodes.

What is it? - TIAs again. This time his vertebral arteries are involved.
Diagnosis? CTA, MRA, or angiogram.
Treatment? Antiplatelet therapy, vascular risk factor modification, and consideration for stenting if refractory.

V3. A 30 year old right handed man has transient episodes of weakness in the right arm and leg. The episodes are quickly followed by a severe headache lasting several hours and associated with nausea and photophobia. When you see him in the office, his strength is normal.

What is it? Hemiplegic migraine.
Diagnosis? Clinical. MRI to rule out stroke.
Treatment? Treat as normal migraine but avoid triptan medications due to increased risk of stroke.

V4. – A 60 yo right handed F presents with abrupt onset of left-sided hemiparesis and difficulty speaking. There was no associated headache. She arrives at the ED 30 minutes after she first noticed symptoms.

What is it? – Ischemic stroke.
Diagnosis? - Head CT is done initially to rule out hemorrhage and may show signs of acute stroke. PT/PTT/INR, CBC, Chemistry are done for purposes of deciding on acute treatment. TPA contraindications are reviewed.

Treatment? Intravenous TPA can be given within 4.5 hours (6 hours for Intraarterial TPA) given that there are no contraindications to its administration. If contraindications exist, or patient presents between 6 and 8 hours, clot retrieval could be considered at advanced centers. Of note, reperfusion therapy like TPA or clot retrieval is aimed at restoring flow to save the area surrounding an infarct called the "ischemic penumbra" and prevent it from becoming infarct itself.

V5. A 60 yo right handed F awakens yesterday with mild left-sided hemiparesis and mild neglect on the left and some slurring of speech but no aphasia. Her symptoms improved and she is left with minor residual deficits. There was no associated headache. A brain MRI shows a small ischemic stroke in the right MCA territory. Carotid duplex shows 80% stenosis on in the right carotid artery.

What is it? Large vessel ischemic stroke, likely secondary to carotid artery disease.
Treatment? CEA should be performed. Again, CEA should be considered in any patient with symptomatic stenosis (>50%) but is more clearly beneficial with stenosis >70%. Patient should be started on antiplatelet therapy – aspirin, clopidogrel (plavix) or aspirin + dipyridamole (aggrenox) are all options. Typically aspirin is given as it is cheapest and the alternatives only show marginal decreases in stroke risk compared to aspirin alone. Plavix or Aggrenox would be considered if a patient was at especially high risk of stroke or if the patient was on aspirin when they had a stroke.
Of note, in asymptomatic patients with carotid stenosis, indications for CEA will depend on degree of stenosis (>60% generally used) along with surgical risk (<3%) and life expectancy (>5 years). See AHA guidelines for a full discussion.

V6. A 60 yo right handed F awakens with left-sided hemiparesis and difficulty speaking. There was no associated headache. A brain MRI shows a fully evolved ischemic stroke in the right MCA territory. Investigations show atrial fibrillation.

What is it? Large vessel ischemic stroke, likely secondary to cardioembolic disease.
Treatment? Anticoagulation (Coumadin or dabigatran) rather than antiplatelet therapy for secondary stroke prevention. IV heparin should be initiated in these special cases even in setting of acute stroke.

Other indications for Coumadin for secondary prevention of stroke due to a cardioembolic source would include low ejection fraction (less than or equal to 25%), DVT with PFO, hypercoagulable disorder, mobile severe aortic arch atherosclerosis. Other neurologic indications for Coumadin could include sinus venous thrombosis or potentially carotid dissection. Anticoagulation would never be used for embolic endocarditis due the high CNS bleed risk from possible mycotic aneurysm.

V7. – A 70 yo right handed M suddenly develops a severe headache on the right side of his head with weakness of the left extremities. Over the next 30 minutes he vomits twice and becomes more somnolent. His wife volunteers his past medical history of untreated hypertension and diabetes.

What is it? - Suspect an intraparenchymal hemorrhage originating from the region of the right basal ganglia affecting the right side of the brain. Hemorrhage vs. ischemic stroke is difficult to distinguish on clinical exam alone. Both will have localizing signs depending on their location. Clues to hemorrhage would include a progressive deficit after initial onset, decreasing consciousness and signs of increased ICP.

Diagnosis? Head CT. The "clues" above are unreliable. CT is necessary.
Treatment? Initial management involves blood pressure control to keep it in the 140-160 range (contrary to ischemic stroke which is left untreated up to ~220 systolic). Measure INR and correct if >1.5. Manage increased ICP with head elevation and hyperventilation or mannitol if acutely needed. Neurosurgery on board in case surgical intervention necessitated (best indication for craniotomy would be a cerebellar hemorrhage larger than 3 cm that would cause pressure on the brainstem but large cortical hemorrhage especially in a younger patient causing herniation concern could be another candidate. Typical cause of hemorrhage especially in older patients with posterior fossa, thalamic, or basal ganglia hemorrhages would be hypertension. In cases of younger patients, patients lacking hypertension, or cortical strokes, evaluation for a vascular malformation or other secondary cause would be warranted. In old patients with a history of multiple cortical hemorrhages, consider cerebral amyloid angiopathy.

V8. A 28 yo pregnant F at 20 weeks presents to the antepartum unit following a seizure. She states she has had a severe headache for the past day. MRI shows bilateral thalamic infarction on DWI.

What is it? Venous Sinus Thrombosis. Clot forming in the dural sinuses causes venous congestion and can cause ischemia in odd places not following a typical vascular territory (bilateral thalamus, corpus callosum, etc.). Tends to occur in hypercoagulable patients, such as this pregnant female.
Diagnosis? MR Venogram would show clot in sinuses.
Treatment? IV heparin would be started initially followed by coumadin. If clot does not resolve, thrombolytic injection into the sinuses may rarely be necessary.

Seizure/Syncope
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S1. A 7 year old boy is brought in by his mother for staring spells. She reports that the boy's teacher has complained the child has trouble paying attention in school and is noted to stare off into space frequently. The mother noted an episode yesterday when the child would not respond to her for a few seconds while staring off with eye fluttering. He was not confused afterwards.

What is it? Childhood Absence Epilepsy. Suspect this in a child of age 5-10 with brief "staring spells" without postictal confusion.
Diagnosis? EEG will show the classic 3 Hz spike-and-wave pattern.
Treatment? Ethosuximide is first line. Lamictal or depakote are used for more resistant cases.

S2. A 30 year old female is brought to the hospital by her husband following an episode of loss of consciousness. She was found in the bathroom on the floor with rhythmic shaking. On exam, she is confused and smells of urine. She gradually becomes less confused over 30 minutes. At that time, she states she doesn't remember the episode of shaking but recalls an unusual smell of "burned candy" immediately prior to it.

What is it? Partial seizure with secondary generalization. The patient presents with an aura of a burnt candy smell localizing to the temporal lobe that secondarily generalized to involve her entire cerebral cortex resulting in loss of consciousness and generalized

tonic-clonic seizures. Another classic presentation of partial seizures with secondary generalization is the “Jacksonian March” where shaking starts in the hand and gradually expands to involve the arm and then whole body.
Diagnosis? EEG and MRI would be performed as above to look for a precipitating cause, most likely in the temporal lobe in this case.
Treatment? If EEG/MRI were normal, AEDs would not be started given she would only have a 33% chance of developing epilepsy after one isolated seizure. Driving cessation until seizure free would be recommended.

S3. A 30 year old female presents to you complaining of an episode of right arm rhythmic shaking yesterday. She notes she was totally aware during the entire episode, which lasted two minutes.

What is it? This history is suggestive of a partial seizure localized to the motor strip in the left frontal lobe. The key to partial seizures is that the patient has no alteration in consciousness and the symptoms can often be localized. The shelf may ask you to localize and common questions would be (smell - temporal lobe, visual symptom - occipital lobe, motor symptom - frontal lobe). While this presentation is similar to a TIA, ischemia tends to present as “negative” symptoms like weakness or vision “going black”, whereas seizures tend to present as “positive” symptoms like shaking or seeing flashes of light.
Diagnosis? EEG and MRI would be performed as a standard part of the initial seizure evaluation to evaluate for a precipitating cause (tumor, vascular lesion, etc.), which are common in partial seizures.

S4. A 33 year old is being evaluated for spells. She has had unusual spells where she will suddenly become unresponsive that lasts for a minute or two, stare off, and pick at things in the air (per witnesses but the patient does not remember the time during the events). She states she is confused and sleepy for up to an hour afterwards. She noticed some lip smacking at the beginning of his last spell.

What is it? Complex partial seizure. The patient has an alteration in consciousness, automatisms (finger rubbing, lip smacking, chewing or swallowing) and postictal confusion is present. It also must be differentiated from absence seizures in children - the key is the presence of postictal confusion in complex partial seizures and longer duration of the seizure activity.
Diagnosis? EEG and MRI.
Treatment? This patient would be started on an AED given that he has had multiple, seemingly unprovoked seizures qualifying for the diagnosis of epilepsy. Generally speaking, all the AEDs are effective in treating focal-onset seizures (not the case with primary generalized seizures) and so therapy would involve choosing by cost and side effect profiles.

S5. A 70 year old male with history of hypertension treated with HCTZ is brought in by his family. He had been at his grandson’s graduation. It was a hot day outside. At the end of the ceremony, the patient stood up to leave but suddenly became lightheaded and lost consciousness and fell to the ground. The patient’s son noted a few brief jerks after he fell to the ground. There was no tongue-biting or loss of urine. The patient quickly regained consciousness afterwards without any confusion.

What is it? Syncope from orthostatic hypotension. The patient has risk factors for dehydration (hot day, diuretic), is elderly and presents with sudden dizziness (lightheadedness) upon standing. Do not let the "few brief jerks" fool you! "Convulsive syncope" is common and is differentiated from true tonic-clonic movements by the short duration of convulsions and other historical clues suggesting syncope. Note there is no post-ictal confusion, urinary incontinence, or tongue-biting to suggest seizure.
Diagnosis? Perform orthostatic blood pressure measurement to confirm. A positive test is a drop of >20 systolic/10 diastolic when measured supine vs. erect. A tilt table test is a more sensitive test for orthostatic hypotension if needed. A screening EKG would also likely be performed in this elderly man with HTN to rule out a cardiac cause.
Treatment? Rehydration for this case.

If a patient has refractory orthostatic hypotension from autonomic dysfunction, then compression hose, leg strengthening exercises or pharmacological agents (examples being fludrocortisone or midodrine) can be used. Elevating the head of the bed 20 degrees can help by keeping the renin-angiotensin system active during sleep.

S6. An 18 year old female is brought to the ER by her mother immediately after an episode of loss of consciousness. The girl was at a football game when her friend tripped and cut her lip open and was bleeding significantly. The girl states she felt lightheaded and then lost consciousness and fell to the ground. She regained consciousness after a few seconds and reports no confusion after the event. The patient reports she had a similar episode last year while at a haunted house.

What is it? Vasovagal (vasodepressor) syncope. This is caused by a vagal (parasympathetic) reaction resulting in either cardiac inhibition and/or vasodilation and subsequent CNS hypoperfusion leading to loss of consciousness. Common precipitants include fear, emotional response, sight of blood, post-tussive, or straining going to the bathroom. Patients tend to have recurrent episodes starting in their teens and a past episode may be given as a clue.
Diagnosis? History as above.
Treatment? Trigger avoidance.

If no trigger is associated with these, the pharmacological options would include SSRIs for a trial as an example.

S7. A 55 year old male reports to the ED with nausea, vomiting and severe epigastric pain radiating to his back. An NG tube is placed and the patient is made NPO with IV fluids and meperidine. When you see the patient the next day, he appears improved but looks mildly tremulous. An hour later, the nurse calls you when she finds the patient unconscious and making tonic-clonic movements.

What is it? Alcohol withdrawal seizures. These can occur 12-48 hours after alcohol cessation. Sometimes the vignette will state the patient is an alcoholic; other times it will give clues like a diagnosis of acute pancreatitis or tremulousness (sign of withdrawal) and sometime it will only give risk factors - college student, homeless. This should be high on the differential for post-operative seizures (because patient has no access to alcohol).
Diagnosis? Clinical suspicion.
Treatment? Benzodiazepines both acutely for seizure and give scheduled benzodiazepine with gradual taper for withdrawal. Patients are often placed on CIWA

protocol (a scoring system to measure signs of withdrawal and used to determine dose of prn ativan). Note: CIWA does not determine if a patient is in withdrawal. You must decide that. It only measures the severity of symptoms, not their cause. No AEDs are indicated in provoked seizures.

S8. A 55 year old male reports to the ED with nausea, vomiting and severe epigastric pain radiating to his back. An NG tube is placed and the patient is made NPO with IV fluids and meperidine. When you see the patient the next day, he appears improved but looks mildly tremulous. The following day the patient looks increasingly tremulous with a mild tachycardia. He has pulled out his NG tube, and was seeing things that weren't there in the room. The next night, the patient has develops a fever of 104, BP 210/120 and HR 120. When you enter the room, the patient is obtunded and diaphoretic.

What is it? Delirium Tremens. This is the most feared stage of the alcohol withdrawal syndrome, occurring about 3-7 days after the patient's last drink. It is marked by autonomic instability, impaired mental function, as well as agitation and sympathetic symptoms. Note, this patient also went through the stage of alcoholic hallucinosis first (visual hallucinations most common- start 12-24 hours out and tend to resolve 24-48 hours out).
Diagnosis? Clinical.
Treatment? Benzodiazepines as above for withdrawal.

S9. A 25 year old male with type I DM is found on the living room floor unconscious and rhythmically jerking. The patient had been feeling sweaty, tremulous, anxious, a little confused with some palpitations the hour prior to this.

What is it? Hypoglycemia-induced generalized seizures. This patient is an insulin-dependent diabetic with concurrent illness and likely decreased oral intake.
Diagnosis? Finger-stick blood glucose will be low - There is no strict cut off value that will induce seizures but it will typically be below 60 mg/dL.
Treatment? IV dextrose. No AEDs are indicated in provoked seizures.

S10. A 35 year old female with history of seizures presents to your office with complaints of increasing frequency of seizures. She states she has had seizures occasionally for the past five years but she is now having seizures "all the time". Her alcoholic husband is present with her and he states they can last up to an hour. He videotaped her last seizure, which shows her on the ground with eyes closed, with her arms flailing and her head shaking side to side. She states she is usually "out of it" for a couple minutes afterwards.

What is it? This history is suspicious for psychogenic nonepileptic spells (formerly known as pseudoseizures). This is a form of conversion disorder thought to be an unconscious reaction to psychological stress. Hints that the diagnosis is PNES include stress at home (alcoholic husband), long seizures (>30 minutes) without significant sequelae, little post-ictal confusion, alternating movements, shaking head side-to-side, closed eyes, intact consciousness while shaking bilaterally, or pelvic thrusting. However, long-term EEG monitoring is required to confirm the diagnosis.

Diagnosis? If PNES is suspected, patients are placed on long-term EEG monitoring in an Epilepsy Monitoring Unit. If a typical spell is witnessed without corresponding epileptiform discharges, the diagnosis is confirmed.

Treatment? No AEDs are necessary (unless patient has concurrent epilepsy which is not that uncommon). The patient should be told that she does not have epilepsy and that her spells are secondary to a processing error of the mind from stressors affecting the subconscious mind (it is stressed these spells are not voluntarily produced) but that the prognosis can be good with suggestive positive reinforcement and an attempt to deal with any underlying stressors. How the diagnosis is delivered has been shown to be very important in effecting patient outcomes. Cognitive behavioral therapy may be effective for PNES.

S11. A 25 year old female with history of refractory epilepsy starts to have a seizure while in the hospital. When you arrive, she is 5 minutes into a generalized tonic-clonic seizure.

What is it? While not technically status epilepticus (>30 minutes), it is treated like status epilepticus certainly by 5 minutes of continuous seizing.
Next step? 2 mg IV ativan. If that does not break the seizure, try another dose of IV ativan and load with 20mg/kg fosphenytoin. If unsuccessful, consider another dose of IV ativan and then consider intubation to protect airway with using more definitive treatments to break the seizure if still ongoing at that point such as propofol or Phenobarbital/Pentobarbital.

S12. A 25 year old female with history of refractory epilepsy starts to have a seizure while in the hospital. When you arrive, she is 5 minutes into a generalized tonic-clonic seizure. She is given 2 mg IV ativan but continues to seize for the next 5 minutes. The patient is loaded with fosphenytoin and given another dose of IV ativan. 15 minutes in, respiratory rate is 4/min.

What is it? Status epilepticus (again not technically) with respiratory depression from benzodiazepines.
Next step? Endotracheal Intubation. Also be aware that a seizing patient cannot protect their airway and endotracheal intubation may be indicated even before high doses of benzodiazepines are given.

S13. A 4 year old female is brought in by EMS after having generalized convulsions. Her parents state the child has recently had a sore throat and suddenly became unconscious with bilateral rhythmic jerking last 10 minutes. Her temperature in the ED is 103.4 F. Neurological exam is normal.

What is it? Febrile seizures. These typically occur in patients 6 months to 6 years old during an acute febrile illness. There is no specific threshold temperature but it is thought that patients are more susceptible if they have a rapid rise in temperature.
Next step? Treat with Tylenol. Reassure parents. If seizure had lasted >15 minutes, was a partial seizure, there were focal deficits, or the child experienced more than one seizure in a 24 hour period, then a more thorough neurological workup would be necessary and the patient would be at a higher risk for developing epilepsy.

S14. A 25 year old female is started on Depakote for recurrent seizures. She asks you the side effects.

What's the point? Depakote is a common AED and it has three black box warnings - and they are all testable. 1) Hepatotoxicity 2) Teratogenicity - Neural Tube Defects (Depakote is the worst AED in pregnancy) 3) Life-threatening pancreatitis.

Common seizure side effects to consider for your shelf:

Lamotrigine (lamictal)- Stevens-Johnson rash
Carbamazepine (tegretol)/Oxcarbazepine (trileptal)- Hyponatremia
Phenytoin (dilantin)- gingival hypertrophy, hypertrichosis, ataxia
Levitiracetam (keppra)- agitation/neuropsychiatric disturbance
Topiramate (topamax)- kidney stone, mild cognitive difficulties/word finding difficulties
Valproic acid (depakote)- weight gain, hair thinning, tremor, hepatotoxicity, pancreatitis, teratogenicity.

All seizure medications, if toxic/overdosed on them, could cause nystagmus, n/v, ataxia.

Cognitive Disorders
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C1. A 75 year old male is brought in by his wife for "memory problems," worsening over the past year. She states that she has taken over bill paying after her husband kept paying their utility bills too many times because he had forgotten he had already paid it. When asked, the patient denies a problem and jokes "just trying to stay ahead of the bills". She also notes that he recently got lost while driving in the neighborhood where they've lived for 40 years. She notes that she has felt his memory has declined for the past few years but is increasingly severe for the past year.

What is it? Early Alzheimer's Disease. In early stage Alzheimer's, patients have difficulty with anterograde memory formation and visuospatial skills resulting in functional impairment. Classically they will forget recent events and get lost. The patients often deny there is a problem and use humor to minimize their problems. They are typically brought in by family members.
Diagnosis? The approach would be to rule out reversible causes of dementia as Alzheimer's is a clinical diagnosis and would want to ensure not missing treatable conditions. This workup standardly consists of depression screen, TSH, B12, RPR, and MRI (to rule multiinfarct, hydrocephalus, or inflammatory lesions). Neurocognitive testing would also be helpful.
Treatment? Acetylcholinesterase inhibitors are first line therapy to improve cognitive function and slow cognitive decline, such as donepezil (Aricept). An NMDA antagonist namenda (memantine) can be added on for moderate to severe cases. .

C2. A 75 year old male comes to your office complaining of "memory problems". He states that he doesn't have the sharpness he used to and cites examples like not being able to answer Jeopardy questions before the contestants as easily anymore. He also states that he has trouble remembering as many details about his clients at work. He continues to work as a banker without problem. His MMSE is 30/30.

What is it? With no real significant impairments, these mild memory/processing issues that are not quite as sharp without any real functional impairment could represent normal

aging. Reassurance could be given. This needs to be differentiated from mild cognitive impairment and dementia.

C3. A 75 year old reports progressive decline in memory for the past 12 months. He states he is more easily disoriented, loses things and forgets things. He states he often repeats questions and e-mails because he forgets that he already asked or wrote them, respectively. He also states he has had trouble recognizing his own car sometimes in the parking lot. He is still able to perform well at work and screening of ADLs shows that he is completely independent. MMSE is 27/30, losing two points in delayed recall and one point in stating today's date. Lab workup is unremarkable, and MRI shows cortical atrophy consistent with age.

What is it? Mild cognitive Impairment. MCI consists of a more significant impairment in a cognitive domain than normal aging, but without significantly affecting activities of daily living. For example, a patient may have noticeable memory difficulties but still be able to maintain typical activities of their daily living. On the other hand, dementia represents significant impairment in at least 2 cognitive domains- memory and learning new information plus another domain such as visuospatial function, language function, executive function, or personality.
Diagnosis? Clinical, often with cognitive testing. MCI patients often miss a couple points on recall on MMSE testing, while scores in the below 27 or certainly below 24 would be more suggestive of an early dementia picture.
Treatment? Observation with the understanding that elderly patients with MCI have a 5-16% annual risk of progressing to dementia versus 1-3% annually in the general elderly population.

C4. An 85 year old female from a nursing home with advanced Alzheimer's is in your office. What would be clinical features seen in late stage Alzheimer's?

Early Alzheimer's presents with memory complaints, some word finding difficulties, and visuospatial difficulties (i.e. might get lost while driving). Alzheimer's steadily progresses with the loss of abilities to maintain typical activities of daily living due to cognitive decline. Late stage Alzheimer's expands its clinical sequelae. It can involve hallucinations, delusions, sleep disorders, aggression, or paranoia. Atypical antipsychotics can be used for agitation, though limited use as needed as they may increase mortality overall. Incontinence develops. Patients develop apraxia and lose the ability to use utensils for eating and gait difficulties occur and eventually the patient ends up bed bound with poor motor control, poor swallowing, and minimal speech production. Secondary infections- i.e. from pneumonia are a common source of mortality. Comfort and palliative care are the mainstay of therapy in very advanced Alzheimer's.

C5. An 80 year old female with Alzheimer's Disease is sent to the ED from her nursing home for decreasing mental status. The patient has moderate Alzheimer's disease that is treated on stable doses of donepezil and memantine. A nurse had noticed the patient appeared more drowsy and was mumbling to herself 2 days ago, but when she checked back later that afternoon, the patient appeared to be better. However, today she once again appears drowsy and confused. A head CT in the ED shows diffuse cortical atrophy.

What is it? Delirium. The waxing and waning acute change in mental status is suggestive of delirium. Altered sensorium is a clue to this. Delirium is more common in patients with underlying neurologic disease like strokes and dementia and is commonly caused by infection, electrolyte imbalance, and medications (especially anticholinergics).
Next step? Urinalysis & electrolytes to search for common causes of delirium.

C6. A 65 yo male with history of CAD, hyperlipidemia and HTN is brought to your office by his wife for "memory problems". The wife states his husband all of a sudden started having trouble with his memory 3 years ago and she has taken care of most their finances since. His memory just recently got worse on Tuesday of this past week and he now has some slurring of his words. On exam, the patient has right-sided weakness, uneven palatal elevation and an upgoing babinski on the right.

What is it? Multi-infarct Dementia. Note that this is a patient with vascular risk factors who presents in a step-wise cognitive decline with focal signs on neurological exam.
Diagnosis? MRI will show multiple infarcts.
Treatment? Cholinesterase inhibitors may improve cognitive function. Controlling vascular risk factors will help to prevent further strokes.

C7. A 59 year old male is brought in by his wife for "strange behavior". She states the patient has had a gradual change in his personality and is far more likely to yell and become upset. She also notes that he often places objects in his mouth, as if he is examining them with his mouth. On exam his memory and visuospatial skills are intact. A brain MRI shows atrophy in the frontal and temporal lobes bilaterally.

What is it? Frontotemporal dementia. This common dementia presents with changes in personality and sometimes includes hyperorality or language deficits. Memory and visuospatial skills are typically retained early though a global dementia is part of the late stage of this disorder.
Diagnosis? MRI would show frontotemporal atrophy.
Treatment? Supportive psychiatric medications as needed.

C8. A 78 year old male has been having visual hallucinations for the past two years. His wife states that he has good days and bad days with significant confusion on certain days. She also notes that he has fallen several times lately. She has also woke up to find her husband out of bed walking around the house but appears to still be asleep. Exam shows mild bradykinesia and a resting tremor of his hands bilaterally.

What is it? Dementia with Lewy Bodies (DLB). This dementia is characterized pathologically by the presence of Lewy bodies in the brain. A patient with DLB will present initially with a dementia characterized by fluctuating levels of alertness, visual hallucinations, falls and often REM sleep behavior disorders.
Diagnosis? Clinical. Another clue to diagnosis would be the onset of a delirium-like state with anticholinergics or visual hallucinations after administering L-dopa for a patient's parkinsonism. Typical antipsychotics should be avoided as DLB patients have neuroleptic sensitivity with common reactions of sedation, Parkinsonism, or neuroleptic malignant syndrome. Atypical antipsychotics should only be used at a low dose with caution.

Treatment? Anticholinesterase inhibitors such as donepezil (Aricept) are first line medications for this disorder. Careful titration of L-dopa (to avoid mental status effects/visual hallucinations or orthostasis in these sensitive patients).

C9. A 78 year old male with a 10 year history of Parkinson's Disease presents with visual hallucinations. His wife states that he has good days and bad days with significant confusion on certain days. She also notes that he has fallen several times recently, and his levodopa therapy has gradually decreased in efficacy over the past few years. Exam shows masked facies, bradykinesia and a resting tremor in his right hand.

What is it? Parkinson's Disease Dementia (PDD). This is a closely related dementia to DLB, but is differentiated by the development of cognitive impairment greater than one year after the onset of motor symptoms (average length is 10 years). The dementia that develops in PDD is characterized by the same cognitive symptoms and neuropathologic features (lewy bodies) as in DLB (see above DLB question).

C10. A 38 year old female is referred to you for dementia. Over the past four months she has had increasing confusion, difficulty with memory, gait incoordination, and she has frequent quick jerks of her limbs especially when startled.

What is it? Creutzfeldt-Jakob Disease. This presents with a subacute dementia presentation with startle myoclonus and potentially gait incoordination. The boards will likely make the patient in the vignette young to help you realize this is not a more typical neurodegenerative process driving the dementia.
Diagnosis? EEG will show periodic generalized 1Hz discharges that are characteristic. CSF may show 14-3-3 protein.
Prognosis? The disease is fatal, typically within one year of onset.

C11. A 73 year old female presents due to recent falls. She has had a slow gait for the past several months, which has worsened recently resulting in falls. Upon further questioning, her husband reports that she has had difficulty performing routine tasks around the house like paying bills or remembering to turn off the stove and accidental loss of urine.

What is it? Suspect Normal Pressure Hydrocephalus. While relatively uncommon, NPH is a favorite on the boards due to its classic triad and because it can be a reversible cause of dementia. The triad consists of a 1) apraxic gait 2) dementia and 3) urinary incontinence.
Diagnosis? CT or MRI of the brain should reveal enlarged ventricles that are not caused by cortical atrophy. High volume LP with a pre and post gait assessment. If gait significantly improves with LP, considered diagnostic.
Treatment? Approximately half of patients will improve after a ventricular shunting procedure. Good prognostic indicators for a shunt response include gait disorder appearing first and being more prominent than cognitive dysfunction and short duration of symptoms- i.e. less than 6 months.

C12. A 55 year old male is brought to the ED after being found on the street confused. Police report the patient has difficulty walking a straight line. The patient reports he's fine but "needs a drink" because he's "seeing double" when he looks to the right or left. On exam, the patient has bilateral lateral rectus palsy.

What is it? Suspect wernicke's encephalopathy in this alcoholic with the classic triad of confusion, ataxia, and oculomotor involvement (nystagmus, gaze palsy).
Next step? Give IV thiamine. It is especially important (and often tested) to give thiamine prior to glucose administration in an alcoholic as glucose can displace thiamine and induce wernicke's encephalopathy. (Note: It's been suggested that the old "thiamine before glucose rule" is just mythology passed down from generation to generation of physicians without much evidence to back it up. Regardless, give thiamine before glucose on your board exams).

C13. A 55 year old female reports her "memory stinks" for the past month. She states she is having trouble at work because she cannot remember what tasks she has to perform and has difficulty concentrating while there. She is constantly forgetting where she placed items, like her keys, etc. She feels it may be due to her recent difficulty sleeping and she states she's so stressed about all this that she "can't eat a thing" and has no energy. On MMSE, she scored a 29/30, missing just one point on one of the three recall items. Her affect is flat.

What is it? "Pseudodementia" from depression. Depression screens are routinely performed to rule out depression as a cause of cognitive impairment. The depression in an elderly patient can affect concentration and cause the patient to have memory issues because they are not attending to processing items for memory well. Patients' cognitive testing generally is better objectively than their subjective complaints, and they are oftentimes the one bringing up their issues with this rather than family members, etc.
Next step? Treat the depression with combination of SSRI and psychotherapy.

C14. A 55 year old female reports her "memory stinks" for the past two months. She states she is having trouble at work because she cannot remember what tasks she has to perform and has difficulty concentrating while there. She also reports feeling sleepy and easily fatigued. She has gained 15 lbs over the past two months and says she's constipated.

What is it? Hypothyroidism. Hypothyroid patients can have difficulty with cognition including memory and concentration.
Next step? TSH is best test. Free T4 would likely accompany TSH. Treat hypothyroidism if confirmed.

C15. A 68 year old male has been in the hospital for the past day after a pulseless arrest lasting 30 minutes. You are asked to evaluate the patient neurologically. On exam the patient is on the ventilator and does not breath over the vent. He does not respond to voice or sternal rub, pupil, oculocephalic with cold caloric testing, corneal and gag responses are absent, and patient has absent reflexes bilaterally in his upper and lower extremities.

What is it? Brain death secondary to hypoxia-ischemia. Brain death is defined by 1) unresponsiveness (coma) 2) brainstem death as evidenced by absent BST reflexes and absent spontaneous breathing (including failed apnea test). These observations must be made after other factors (temperature, electrolytes, oxygenation) are corrected. Note: the presence of spinal cord reflexes does not have any significance on determining brain death. Ancillary tests can be performed to look for a flatline EEG or lack or perfusion of a nuclear medicine scan but these are not required routinely in typical cases meeting the above criteria.

C16. A 68 year old male has been in the hospital for the past 3 days after a pulseless arrest lasting 10 minutes. You are asked to evaluate the patient neurologically. On exam the patient has been weaned off the vent and is breathing spontaneously. He does not respond to voice or sternal rub. Pupils are sluggish but reactive. Oculocephalics and corneal are minimally reactive and gag responses is present, and patient has 2+ reflexes bilaterally in his upper and lower extremities.

What is it? Coma. Differentiated from brain death by presence of respiratory drive and presence of some cranial nerve reflexes.

C17. A 68 year old male has been in an extended care facility for the past 5 weeks after a pulseless arrest lasting 10 minutes. You are asked to evaluate the patient neurologically. On exam the patient has been weaned off the vent and is breathing spontaneously. He does not respond to voice or tactile stimuli. He opens his eyes spontaneously in the morning, appears awake during the day, and closes his eyes at nighttime. He cannot follow any commands or interact with his environment meaningfully. Pupil, oculocephalic, corneal and gag responses are present, and patient has 2+ reflexes bilaterally in his upper and lower extremities.

What is it? Persistent Vegetative State. Differentiated from coma in that patients have sleep-wake cycles. Patients have no observable, repeatable, meaningful responses to external stimuli. Must be no improvement for 3 months to be considered "persistent" in non-traumatic cases and 1 year in traumatic cases. Coma will typically transition to a recovering state vs a vegetative state within about 2 weeks.

Infectious Disease
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I1. An 18 year old male reports to his college health center with complaints of 8 hours of fever, headache and neck stiffness. He jerks in pain when his hip is flexed and knee extended. Exam shows raised red/purplish spots on his trunk.

What is it? Bacterial meningitis. Any patient with fever, headache, and neck stiffness should be suspected of having meningitis. His age, living conditions (college), and ecchymotic rash point to N. meningitidis as the likely culprit for his infection.
Diagnosis? Lumbar puncture with glucose, protein, WBC, RBC and gram stain, and cultures. Blood cultures would be drawn as well. If focal signs were present, an abscess might be present leading to increased ICP, and therefore a head CT should be performed prior to LP to avoid herniation. Do not delay empiric treatment waiting for study results. Bacterial meningitis shows depressed glucose with elevated white count and a neutrophil predominance. Viral would have a near normal glucose with elevated white count and a lymphocytic predominance.
What looks like lancet-shaped diplococci on gram stain? S. pneumoniae
What looks like gram-negative diplococci on gram stain? N. meningitidis
Treatment? Empiric IV antibiotics - specifically ceftriaxone for bacterial meningitis, with vancomycin initially to cover penicillin-resistant strains while csf and blood cultures are pending. Ampicillin added for certain populations, such as the elderly, immunocompromised, etc.

I2. A 30 year old male reports to the ER with complaints of fever, headache and neck stiffness. He jerks in pain when his hip is flexed and knee extended. No focal deficits are presents. LP shows normal glucose, lymphocytic pleocytosis, and negative gram stain.

What is it? Viral (Aseptic) meningitis. Any patient with fever, headache, and neck stiffness should be suspected of having meningitis. His LP is characteristic of viral meningitis.
Diagnosis? PCR for HSV and enterovirus are commonly done.
Treatment? HSV in the CSF would be treated with acyclovir. Other etiologies would be treated supportively.

I3. A 30 year old male reports to the ER with complaints of fever, headache and neck stiffness. He was confused and lethargic when the ambulance arrived and had a seizure while in riding in the ambulance. Exam is negative for meningeal signs. MRI shows abnormalities in the bilateral temporal lobes. LP shows normal glucose, lymphocytic pleocytosis, and negative gram stain.

What is it? HSV encephalitis. Any patient with fever, headache, and neck stiffness should be suspected of having meningitis, but this patient additionally has signs of encephalitis - seizures, severe mental status changes, and temporal lobe abnormality on MRI. His LP is characteristic of a viral infection. Encephalitis involving the medial temporal lobe is characteristic of HSV. Pure encephalitis can lack any meningeal signs of irritation, though a meningoencephalitis can have signs of both encephalitis and meningeal signs.

I4. A 70 year old female with history of Aortic Valve replacement and recent dental procedure reports with complaints of fever, headache and difficulty expressing her thoughts. Exam reveals papilledema. CT shows a ring-enhancing lesion in the left frontal lobe.

What is it? Brain Abscess. The classic triad of brain abscess is subacute fever, headache, and focal neurologic signs. There may be increased ICP. CT/MRI will show a ring-enhancing lesion. Risk factors include any causes of bacteremia, preceding infections adjacent to the brain (sinusitis, otitis) or an immunocompromised state.
Diagnosis? You would likely start treatment based on this history. Aspiration or blood cultures could be done to target therapy.
Treatment? Treatment would depend on the size and location of the lesion but would certainly involve IV antibiotics with possible surgical intervention or CT-guided aspiration.

I5. A 45 year old with long-standing HIV reports to your clinic for forgetfulness. He states he has had a recent pneumonia treated with TMP-SMX but is not on antiretrovirals due to cost and compliance difficulty. He states he has gradually become more forgetful and has had difficulty concentrating over the past year. On exam, the patient is slow to process your questions and respond. An MRI shows mild diffuse atrophy with prominent sulci and ventricles.

What is it? HIV-associated Dementia (HAD). This dementia typically presents in patients with AIDS secondary to the toxic immune response from the HIV virus in the CNS. It is typically insidious and later in the course of HIV illness.

Diagnosis? MRI to rule out focal pathology and opportunistic infection. In addition, imaging may show the characteristic diffuse atrophy as above.
Treatment? HAART

I6. A 35 year old with HIV reports right-sided progressive hemiparesis for the past week. He reports one seizure, which caused him to come to the ED. He appears confused and lethargic. CD4 is 50. An MRI shows multiple ring-enhancing lesions.

What is it? Cerebral toxoplasmosis. This is the most common intracranial pathology in AIDS patients and usually occurs only when CD4<100. It can present with a variety of focal signs/symptoms as well as symptoms of increased ICP. Ring-enhancing lesions on MRI/CT with contrast are characteristic.
Diagnosis? A presumptive diagnosis is made by a combination of clinical presentation, history of exposure (positive serology (IgG)) and imaging. CSF PCR for toxoplasma can be helpful as well. A therapeutic trial of pyrimethamine-sulfadiazine is often used even if still considering other causes (like CNS Lymphoma).
Treatment? Pyrimethamine-sulfadiazine for 6 weeks will usually result in a response, especially if treated early. Clinical response is expected within the first week.

I7. A 35 year old with HIV reports right-sided progressive hemiparesis and slurring of his speech for the past few months. CD4 is 50. An MRI shows left confluent subcortical white matter disease without mass effect.
What is it? Progressive Multifocal Leukoencephalopathy. Suspect this condition in an HIV patient with insidious onset of focal signs/symptoms and multiple areas of demyelination on MRI that do not enhance or produce mass effect.
Diagnosis? CSF JC virus PCR is 80% sensitive.
Treatment? HAART therapy.

I8. A 35 year old with HIV reports right-sided progressive hemiparesis for the past few months and complaints of morning headaches. An MRI shows a single enhancing periventricular lesion. CSF is positive for EBV.

What is it? CNS Lymphoma related to AIDS. This can present as either a single or multiple lesions that may or may not enhance and so can present like toxoplasmosis.
Diagnosis? EBV in the CSF is relatively specific for this condition.
Treatment? Poor prognosis but HAART therapy, steroids, and radiation are utilized, as well as chemo in selected patients may be considered.

I9.A 35 year old with HIV reports diffuse headaches for the past few months. He also reports numbness over the right side of his face. CD4 is 50. An MRI shows enhancement of the base of the brain. CSF reveals decreased glucose, increased protein and mononuclear pleocytosis.

What is it? Either TB or fungal meningitis. These will present subacutely in immunocompromised individuals and are characterized by headaches or cranial neuropathies, basal MRI enhancement and mononuclear pleocytosis in the CSF.
Look for clues in the history to suggest different causes: TB (homeless, immigrant, history of TB), cryptococcus (soil/pigeon droppings), histoplasma (Ohio/Mississippi river valleys – bird/bat droppings), coccidiomycosis (southwest US), blastomycosis (also associated with vertebral osteolytic lesions), candida, or aspergillosis.

Diagnosis? Depends on etiology – crypto has india ink stain or crypto antigen. TB may have positive PPD or +CXR findings. Candida may show budding yeast and pseudohyphae.
Treatment? Liposomal amphotericin for fungus. Systemic Antituberculosis therapy for TB.

I10. A 35 year old male from Connecticut presents with headache, bilateral facial palsy, and he also reports some numbness in his feet. He notes that developed a circular, enlarging rash that had since gone away a month ago.

What is it? Lyme disease. Lyme disease on the boards will classically present in patients from the Northeast with some mentioned outdoor/tick exposure followed initially by erythema migrans rash, headaches and arthralgias/myalgias. Neurological manifestations may include meningitis, radiculopathy, neuropathy, encephalopathy, or Bell's palsy. Bilateral Bell's palsy could be clues for Lyme disease or Sarcoidosis.
Diagnosis? Test for serum Lyme antibodies and CSF studies if concern for central nervous system involvement or meningitis.
Treatment? IV ceftriaxone. Oral doxycycline can be used in cases with only isolated Bell's palsy.

I11. A 40 year old "chimney sweep" presents with acute confusion and hallucinations. He is drooling. You offer him water and when he drinks it, he coughs it up and groans in pain. His wife states he has complained of pain and tingling for the past week at a site on his leg where he was bitten by a bat 1 month earlier.

What is it? Rabies encephalitis. Note that he has "hydrophobia" - painful contraction of laryngeal, pharyngeal, and diaphragmatic muscles in response to swallowing liquids - as well as hypersalivation. Patients may have paresthesias, pain or pruritis at the inoculation site that develop prior to the encephalitis and are thought to manifest from viral replication at the dorsal root.
Prognosis? Almost universally fatal.

I12. A 12 year old girl is bitten by a wild dog. Her parents bring her to the ER. She has no symptoms except pain at the wound site. She has never been vaccinated against rabies.

What's the point? Be concerned that she could develop Rabies. Concern for rabies is warranted when bitten by a bat, fox, raccoon, skunk or wild dog.
Next step? If the dog was captured, monitor it for signs of Rabies. If dog got away or appears rabid, give post-exposure prophylaxis as soon as possible with Rabies Immunoglobulin. If a patient had previously been vaccinated (due to high risk of exposure), provide two booster doses of vaccine on days 0 and 3.

I13. A 55 year old female with poorly controlled type II DM presents with facial swelling. She has had sinusitis for the past week. Hours ago, she acutely developed the onset of painful swelling of her right eye, diplopia and numbness over the upper part of her right face. Serum glucose is 400.

What is it? Cavernous sinus syndrome likely from mucormycosis. Preceding sinusitis and poorly controlled DM are clues. Mucormycosis is usually due to infection with either

Mucor or Rhizopus and is feared for its aggressiveness and ability to infect arteries and cause infarction.
Next step? Urgent action is required. Multiple actions must be taken but the best bet is to choose surgical debridement. Surgery would also involve biopsy to confirm mucormycosis. A CT would likely be done to assess soft tissue involvement. Liposomal amphotericin would be given as well.

I14. A 65 year old former commercial sex worker presents due to difficulty walking. She states she had a progressive difficulty in walking for several years. On exam, she has a wide-based ataxic gait with prominent footslap. Proprioception at the big toe is absent bilaterally. Her knee and ankle appear hypertrophied with significant crepitus but the patient has no complaints of pain. Her pupils are small and irregular and react to accommodation but not to light.

What is it? Tabes dorsalis from late stage neurosyphilis.
Diagnosis? CSF VDRL reactivity would confirm.
Treatment? IV aqueous crystalline Penicillin G. If penicillin-allergic, desensitize and treat with penicillin anyway. (Note: desensitization is only used in allergic patients with NEUROsyphilis. For any other form of syphilis, use doxycycline or tetracycline)

I15. A 21 year old male presents to the ED following a seizure. The seizure reportedly started with arm twitching followed by convulsions and loss of consciousness. There is also a few month history of headaches. The man reports he had been working in Latin America last year working on an organic farm. MRI shows multiple enhancing cystic lesions with scolex visible within the cysts.

What is it? Neurocysticercosis. Clues include travel to an endemic area (Latin America), partial seizures with secondary generalization (suggesting focal pathology) and headaches, and the enhancing cystic lesions are due to the cysticerci lodging in the brain parenchyma. The scolex (head) of the cysticerci can be visible on MRI.
Diagnosis? Criteria for diagnosis can be complicated but presence of scolex or biopsy confirmation secure the diagnosis. Otherwise clinical manifestations, neuroimaging (either cysts or calcification from resolved lesions) and serum antibody tests are used to make a diagnosis.
Treatment? Albendazole and praziquantel are used for living cysticerci (they are assumed to be living if cysts are present on neuroimaging). Antiepileptics for seizure prevention.

Neuroophthalmology
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NO1. A 55 year old African American female presents with sudden onset of intense pain in her right eye. She reports seeing halos around lights. The eye appears red. It is mid-dilated and does not react to light.

What is it? Acute angle closure glaucoma. This typically presents as acute, intense unilateral pain with redness. The pupil is classically mid-dilated and fixed. Anticholinergics or sympathetics can precipitate an attack,
Diagnosis? Ocular tonometry is best tool and will show elevated pressures.

Treatment? Stat acetazolamide and beta-blocker to rapidly reduce IOP. About an hour later (once IOP reduced), give pilocarpine to constrict pupil. Consult ophthalmology urgently if not already done.

NO2. A 60 year old diabetic female presents with diplopia. On exam, there is right-sided ptosis and her right eye is deviated inferolaterally. Both pupils are reactive to light.

What is it? Ischemic 3rd nerve palsy likely secondary to diabetes. Key point: benign causes spare the pupil.
Treatment? Palsy will likely resolve over time. Medical management of diabetes and other vascular risk factors.

NO3. A 60 year old diabetic female presents with diplopia. On exam, there is right-sided ptosis and her right eye is deviated inferolaterally. Her right pupil is dilated and non-reactive.

What is it? Compressive 3rd nerve palsy. An emergency! It is most likely secondary to an aneurysm in the posterior communicating artery.
Next step? Angiography followed by neurosurgical clipping or coiling of aneurysm

NO4. A 60 year old diabetic female presented with a large right ischemic MCA stroke causing left sided weakness and neglect. On day 3 of hospitalization you examine her and she has become somnolent with altered mental status. On exam, the patient is lethargic and GCS is 6. There is right-sided ptosis and her right eye is deviated inferolaterally. Her right pupil is dilated and non-reactive.

What is it? Uncal herniation with compressive 3rd nerve palsy.
Next step? Edema from a large CVA maximizes 3-5 days after insult. Significant mass effect with concern for herniation would be treated with mannitol, intubation and hyperventilation, as well as stat neurosurgical consult for possible craniotomy to relieve pressure. Steroids are not effective for cytotoxic edema as from ischemic CVA; they are effective for vasogenic edema such as related to a tumor.

NO5. A 60 year old female with diabetes complains of double vision, most prominent when looking down. On exam, the diplopia is relieved when covering one eye and you notice she preferentially tilts her head to the side.

What is it? Trochlear nerve palsy. Trochlear nerve palsy has a characteristic feature that the patient will tilt their head to the contralateral side to try to compensate for the abnormal torsion of the eye from the trochlear palsy.

In approaching diplopia, note whether covering one eye relieves the double vision. If it does not, this is monocular diplopia, which is not neurological, and is due to inherent ocular disease or conversion disorder. If it does, the double vision is due to lack of coordination between the eyes. The commonly testable causes of diplopia are myasthenia gravis, oculomotor palsy (discussed above), and trochlear or abducens nerve palsies
Next step? Reassurance and await recovery. It is likely a “microvascular” cause to the palsy given her history of diabetes and no head trauma. Trauma is another common cause of trochlear nerve palsy.

NO6. A 60 year old female with diabetes complains of double vision, most prominent when looking to her right. On exam, the diplopia is relieved when covering one eye and you notice she cannot fully abduct her right eye.

What is it? Abducens nerve palsy.

NO7. A 65 year old female with a 40 pack-year smoking history presents with pain in her right shoulder. You note anisocoria. The pupil is smaller on the right and the difference in size is greater when the patient is in darkness. There is ptosis on the right. Eye movements are full. Chest x-ray reveals a well-defined opacity in her right lung apex.

What is it? Horner's syndrome secondary to pancoast tumor. While anhydrosis was not mentioned, the patient has the other two components of ptosis and miosis as well as a lung mass. Note that the other major neurogenic cause of ptosis (oculomotor palsy) will have either a normal (microvascular cause) or dilated pupil (compressive cause) and eye movements will be affected. Also note the anisocoria is greater in darkness, which suggests the normal dilating sympathetic response is not intact in the diseased eye. The other testable cause of ptosis is myasthenia gravis, which does not involve the pupil, is fatigability and the vignette will likely mention diplopia.
Next step? Chest CT and oncologic consultation.

NO8. A 30 year old woman presents to your office. On exam, eye movements are full and no ptosis or diplopia are noted. Her pupils are slightly asymmetrical. The asymmetry increases in bright light. Her right pupil constricts minimally to bright light. When presented with a near stimulus, the right pupil constricts fully, but is slower than the left to redilate when the near stimulus is removed. Lower extremity reflexes are absent.

What is it? Adie's syndrome. This is a benign dysautonomia present in some young, healthy women which is defined by the presence of 1) tonic pupils and 2) weak/absent lower extremity reflexes.
Need some help with eye exam terminology? The patient has anisocoria (pupillary asymmetry) that increases in bright light, suggesting a parasympathetic palsy. Her pupil is slow to redilate after accommodating, termed a tonic pupil. She could also be said to have light-near dissociation in her right eye because her eye does not react (constrict in response to light) but accommodates (constrict in response to near stimuli).
What's the point? No treatment necessary. Don't mistake this for other entities with tonic pupils: Argyll Robertson Pupils (light-near dissociation bilaterally secondary to syphilis) or oculomotor palsy (will also have eye movements disturbed or diplopia complaints).

NO9. A 35 year old female with a past history of transient right sided weakness presents for blurry vision. On exam, the patient when looking laterally to either side and you notice the patient cannot adduct either eye past the midline.

What is it? Bilateral internuclear ophthalmoplegia in a patient with likely multiple sclerosis. INO is secondary to a lesion in the medial longitudinal fasciculus, which is responsible for carrying a signal from the abducens nucleus in the abducting eye to the contralateral oculomotor nucleus in the adducting eye.
Next step? MRI for suspected multiple sclerosis.

NO10. A 7 year old male is brought in by his mother for strange behavior. On exam, his eyes are deviated downward, he cannot look upward and his eyes accommodate but do not react. CT reveals a pineal tumor.

What is it? Parinaud's syndrome. This syndrome is characterized by a paralysis of upward gaze with possible tonic deviation of the eyes downward ("setting sun" sign). Light-near dissociation can also be present. Several entities can cause this syndrome, most commonly hydrocephalus and pineal tumors. Note: be able to recognize a picture of this (young child with bulging fontanelles and "setting sun" sign).
Next step? MRI and Surgical evaluation

NO11. A 70 year old right handed man has a transient episode of "blurry vision". He reports his vision went black in his right eye for 10 minutes. The episode completely resolved. Fundus shows cholesterol deposits in the right retinal artery.

What is it? – Amaurosis Fugax. This term is used to describe transient monocular blindness secondary to emboli to the central retinal artery of one eye (a specific type of TIA). It is most likely that the patient has a cholesterol plaque at the bifurcation of the right common carotid artery and a small piece broke off and temporarily occluded the vascular supply to his right eye.
How is the diagnosis confirmed? - Imaging of the carotid artery.
Treatment: Carotid endarterectomy is indicated given that the patient is symptomatic. Antiplatelet agents will follow.

NO12. A 35 year old female with a remote history of transient right sided weakness presents for pain in her right eye, worse with eye movement. On exam, the patient has full vision in her left eye but reports blurriness in all visual fields of her right eye. When rapidly alternating your penlight from eye to eye, you note the right eye will dilate slightly when you switch from the left to the right eye. Patient has difficulty discriminating red colors.

What is it? Optic neuritis of the right eye in a patient with likely multiple sclerosis. The exam reveals a relative afferent pupillary defect in the right eye.
Next step? MRI for suspected MS.
Treatment? High dose IV steroids for symptom relief and to shorten the duration of the acute attack and reduce the short term risk of MS (though does not affect long term risk). *Oral* steroids should be avoided as they negatively affect long term vision.

NO13. A 35 year old male presents to the ER after a motor vehicle collision. The patient reports the car "came out of nowhere" and hit him on his passenger side as he was pulling out of his driveway. On exam you note the patient has poor peripheral vision bilaterally, although he claims "I can see fine". Review of systems reveals galactorrhea and hypogonadism.

What is it? Prolactinoma. The patient's bitemporal hemianopsia is characteristic of a chiasmal pattern of blindness, most commonly caused by pituitary adenomas. Galactorrhea and hypogonadism are results of hypersecretion of prolactin.
Next Step? MRI to confirm a sellar mass and likely transsphenoidal removal.

NO14. A 70 year old with a history of MI and TIAs has an abnormal visual field test. You view the report, which shows deficits in the superior temporal quadrant of his left eye and superior nasal quadrant of his right eye.

What is it? Left superior quadrantanopsia likely from a stroke affecting Meyer's loop (optic radiations in temporal lobe carrying information from the inferior retina). You should be familiar with the various patterns of postchiasmal blindness and be able to localize the deficit. A review of the visual field defect diagram should be sufficient. Deficits to be familiar with are inferior quadrantanopsia (optic radiations in parietal lobe called Baum's loop) and homonymous hemianopsia from either optic tract lesion or from occipital stroke (will have macular sparing with PCA territory infarct of occipital lobe).

NO15. A 65 year old man presents with a facial rash. The rash was preceded by pain over the right forehead and nose by one day. There are now multiple crops of vesicles over his right forehead and on the tip of his nose.

What is it? Herpes Zoster Ophthalmicus.
Diagnosis? Usually clinical diagnosis only. For atypical cases could look at a Tzanck smear or viral culture/PCR of vesicular fluid.
Next step? Slit Lamp exam! If HZO is suspected, consult ophthalmology and make sure the patient gets a corneal exam with fluroscein staining to look for herpetic lesions on the cornea. Remember that if herpes zoster involves the nose, it may involve the eye as well.
Treatment? Oral antivirals. Other topical eye treatments may be necessary depending on extent of involvement.

NO 16. A 55 year old female presents for a routine eye examination. She has no complaints. Her tonometry shows intraocular pressure is 26 OD, 29 OS. Fundoscopic examination shows a cup-to-disc ratio of 0.4 OD, 0.8 OS with inferior notching. Visual field testing is performed showing peripheral vision loss in both eyes. The left eye shows most severe loss in the superior and nasal fields.

What is it? Primary Open Angle Glaucoma.
Treatment? Pressure-lowering medications include topical beta blockers, carbonic anhydrase inhibitors, alpha-2-agonists, and prostaglandin analogs among others.

Movement Disorders
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M1. A 30 year old male presents with complaints of tremor. He states he has been having increasing difficulty drinking from a glass and notices the shaking especially while holding a newspaper in the morning. His father has had the same problem and his father additionally has a head nodding tremor. The patient has started using his "dad's trick" of drinking alcohol, which eliminates his tremor temporarily.

What is it? Benign Essential Tremor. Diagnosis is clinical based on typical progressive action tremor and head nodding, that temporarily responds to alcohol. 50% have a positive family history.
Treatment? Beta-blockers (propranolol) or primidone. Deep Brain Stimulation can be effective in medication-refractory cases.

M2. A 65 year old female presents with a tremor. It began gradually over years in her left hand and eventually began in her right hand as well. It occurs at rest. Exam shows limited facial expression, decreased eye blinking, cogwheel rigidity and short stride with decreased arm swing while walking. She has difficulty maintaining balance when pulled backwards. MRI is unremarkable.

What is it? Parkinson's Disease. This vignette shows all the hallmark signs of idiopathic Parkinson's disease. A mnemonic TRAP is useful - Tremor (resting), Rigidity, Akinesia (bradykinesia), and Postural Instability.
Diagnosis? Clinical. Can be made with some confidence in presence of 2 of 3 characteristic features (TRA of TRAP).
Associated pathology? (This is testable!) Loss of dopaminergic neurons in the substantia nigra and deposition of Lewy bodies in the brain in this neurodegenerative disorder. Lewy bodies are comprised of the alpha-synuclein protein.
Next step? Therapy usually begins with Levadopa/carbidopa or a dopamine agonist. Several other drug classes can be added as well as adjuncts. Deep Brain Stimulation is considered if failing maximal medical therapy.

M3. A 65 year old female presents with a tremor. She was recently hospitalized for recurrent vomiting and diagnosed with diabetic gastroparesis. She has been on medication since discharge. Soon after discharge, she developed a resting tremor in both hands. She is also is slow in her movements and shows decreased facial expression and eye blink.

What is it? Drug-induced parkinsonism. Common offending drugs include dopamine-blocking agents like metoclopramide (as in this case) or neuroleptics (haldol is classic). Historical clues to suspect secondary causes are abrupt onset, bilateral onset (PD usually starts on one side), and recently starting a new offending medication.
Next step? Stop the metoclopramide and symptoms will typically resolve within days to weeks. An anticholinergic medication can be considered (such as cogentin), especially if one cannot be taken off of the offending medication completely, symptoms persist or symptoms are severe.

M4. A 50 yo male presents with a resting tremor for the past year. His wife notes he has decreased facial expression and slowed walking for this time period as well. Recently, the patient started feeling dizziness upon standing and has even "fainted" a few times. The patient also complains of impotence for past year and paroxysms of intense sweating and flushing of his skin. He also has incoordination walking. On exam, the patient exhibits masked facies, cogwheel rigidity, and also an ataxic gait/dysmetria of limbs. His blood pressure is 130/85 while supine and 90/60 with standing.

What is it? Multiple System Atrophy (MSA). This one of the Parkinson's plus syndromes that can be difficult to learn. Think of MSA as typical parkinsonism with the addition of autonomic failure or cerebellar dysfunction. Signs of autonomic failure present in this vignette include orthostatic hypotension, impotence and autonomic storms resulting in diaphoresis and flushing. Signs of cerebellar dysfunction include gait ataxia and limb dysmetria.

M5. A 65 yo female presents with unsteadiness and falls of recent onset. Her husband also states she seems more disinhibited. On exam, she has a prominent stare and

furrowed brow. She has slowed movements and cogwheel rigidity. She is unable to look down. MRI shows midbrain atrophy.

What is it? Progressive supranuclear palsy (PSP). This is one of the Parkinson's plus syndromes that can be difficult to learn. Think of PSP as parkinsonism (rest tremor very infrequent though) and with addition of downward gaze palsy (vertical gaze palsy). The disease is also characterized by early falls, dysarthria, frontal-lobe cognitive problems and rapid progression. The true hallmark is paralysis of downward gaze, and it'd be unfair to ask you to recognize it without mentioning this in the vignette.

M6. A 65 yo female presents for loss of control of her right hand. She states the hand has a "mind of its own" and will often perform purposeful movements without her initiating the movement. On exam, the patient is noted to have significant difficulty performing organized motor movements and cannot demonstrate blowing out a match or brushing her teeth. She has masked facies and significant rigidity.

What is it? Corticobasal Degeneration (CBD). This is one of the Parkinson's plus syndromes that can be difficult to learn. Think of CBD as parkinsonism (rest tremor very infrequent though) plus "alien limb". The very characteristic "Alien Limb" phenomenon, where a limb performs involuntary purposeful movements is your key to diagnosis.

M7. A 40 year old female comes to your office. Her head is turned to the right with a hypertrophied left sternocleidomastoid muscle. She states that her neck is getting more and more "stuck" in that position and she has neck pain. If she performs a "sensory trick" of lightly touching her head, it relieves the symptoms some, but this has become very bothersome for her.

What is it? Spasmodic torticollis (cervical dystonia).
Next step? Treat with botox injection of tonically active SCM.

M8. A 20 year old male is brought in by his roommate for strange behavior. The patient is stating he is being chased by several demons that are lurking around the corner. The patient is refusing to stay in his hospital bed because he is not safe. He is given an IM dose of haloperidol. He calms down. When you return to his bed he is no longer hallucinating but his head is hyperextended in a spasm position and his arms are stuck in an extended position with his eyes rolled back, and mouth open with tongue protruded. He states in a quiet voice that his arms are "stuck" there. His neck paraspinal muscles are hard to the touch.

What is it? Acute dystonia secondary to neuroleptic use.
Next step? Treat with anti-cholinergic medications like benztropine or trihexyphenidyl or diphenhydramine (and antihistamine which also has anticholinergic properties).

M9. A 20 year old male is diagnosed with schizophrenia and started on typical antipsychotics. When you see him in the office three weeks later the patient's hallucinations have diminished but he has had trouble sitting still since being on the medication. You ask him to stay in his seat. He attempts to remain still but looks very uncomfortable and eventually states he has to get up and move around.

What is it? Akathisia secondary to adverse reaction to haloperidol or other typical antipsychotic. This develops subacutely after drug exposure.

Next step? Stop offending drug. Consider an atypical antipsychotic.

M10. You see a 28 yo male in clinic for schizophrenia follow up. He has been well-controlled on haldol since his diagnosis at age 24. On exam, you note he is making strange movements with his mouth including lip smacking and waving his tongue inside his mouth as well as facial jerks.

What is it? Tardive dyskinesia secondary to chronic neuroleptic use. It typically develops after months to years of drug exposure and is more common in the elderly. It is characterized by choreiform movements of the mouth/face and less commonly the trunk/limbs.
Next step? Wean the haldol and consider an atypical antipsychotic. Tardive dyskinesia usually takes months to years to remit but can be permanent.

M11. A 43 year old man presents to you for change in mood and involuntary movements. He states his father died when he was 55 after 10 years of similar movements and a progressive dementia. He is afraid he may be suffering of the same condition. The patient notes he feels more depressed lately and he is no longer able to perform his job as a banker because he was making mistakes and acting impulsively. On exam, you note involuntary writhing movements in his hands.

What is it? Huntington's Disease.
Next step? MRI would reveal caudate atrophy. Genetic counseling prior to considering testing for a CAG expansion on chromosome four.

M12. A 10 year old girl is brought in by her mother for abnormal movements and painful joints. They immigrated from rural China last year. The girl has been experiencing joint pain for the past week, starting in her elbow but now in her wrist and hands. Yesterday, she started making rapid jerking movements with her hands and facial grimacing. On exam, you note a 2/6 systolic murmur and a pink ringed rash on her trunk, which she states start as a small bump.

What is it? The girl is suffering from acute rheumatic fever and exhibits the motor abnormality of Sydenham's chorea. She also is exhibiting a migratory polyarthritis, erythema marginatum, and a murmur suggestive of carditis. The fact that she is an immigrant from a rural area would make her more likely not to have received antibiotics for a past Group A strep infection.

M13. A 6 year old boy with ADHD presents for follow up. While interviewing the boy, you notice he frequently sniffs. You ask him about it. He says he does it all the time and can't help himself. You ask him to stop. He is able to stop sniffing, but tells you that it makes him very uncomfortable, and he has difficulty controlling his urge to sniff.

What is it? Complex motor tic. A tic is a rapid, recurrent stereotyped movement. These can be classified as simple (one muscle group) or complex (coordinated involvement of multiple muscle groups).
Next step? This tic is likely not affecting the patient's life too much and thus only reassurance should be offered. Most people "outgrow" their tics by adulthood.

M14. A 9 year old boy with ADHD presents for follow up. While interviewing the boy, you notice he frequently sniffs and grunts. You ask him about it. He says he does it all

the time and can't help himself. You ask him to stop. He is able to stop sniffing and grunting, but tells you that it makes him very uncomfortable, and he has difficulty controlling his urge to sniff. He states this has been going on for the past two years and he has had other repetitive behaviors in the past including blinking and repeating words and grunting.

What is it? Tourette's syndrome is characterized by the presence of multiple motor tics including one that is a vocal tic (grunting, echolalia (repeating words), or coprolalia-obscene language) that are present for at least one year. Common comorbidities include anxiety, depression, ADHD, and OCD.
Next step? If the tics are interfering with the patient's life, treatment should be started with an alpha agonist (clonidine or guanfacine) or even an atypical antipsychotic or tetrabenazine.

M15. A 65 year old male comes to clinic for 1 month of falls to his right. You observe his gait to be broad-based with an irregular, lurching stride. He has a slow, coarse nystagmus worse when looking to his right. He is slightly unsteady when standing with his feet together that is not affected with closing his eyes. Finger-to-nose testing reveals past-pointing and intention tremor on the right. He also displays an erratic pattern with alternating hand taps on the right.

What is it? Suspect cerebellar pathology given the patients nystagmus, intention tremor, dysdiadochokinesis and cerebellar gait. Given its subacute presentation and lateralizing findings, suspect a right cerebellar tumor/mass. Gait problems could imply cerebellar vermis involvement but the limb dysmetria would relate to the ipsilateral cerebellar hemisphere.
Next step? MRI with contrast to look for cerebellar pathology.

M16. A 65 year old alcoholic male comes to clinic for chronic unsteadiness. You observe his gait to be broad-based with an irregular, lurching stride. He has a slow, coarse nystagmus. He is slightly unsteady when standing with his feet together that is not affected with closing his eyes. Finger-to-nose testing reveals past-pointing and intention tremor. He also displays an erratic pattern with alternating hand taps.

What is it? Alcoholic cerebellar degeneration. Chronic alcoholic cerebellar degeneration can have a preference for the cerebellar vermis and chronic gait ataxia as the cardinal feature.
Next step? MRI should reveal cerebellar atrophy.

M17. A 60 year old former prostitute presents complaining of falls. She has a narrow-based gait and frequently looks down at her feet while walking. She stands steadily with her feet together but quickly loses her balance when asked to close her eyes. Exam is negative for dysmetria or dysdiadochokinesis. There is bilateral loss of vibration sense in the lower extremities.

What is it? Tabes dorsalis secondary to tertiary syphilis. The positive Romberg is a sign of sensory ataxia.
Next step? Lumbar puncture with testing of CSF for the usual things plus VDRL.
Treatment? Penicillin G IV q4h or IM q6h for two weeks.

M18. A 20 year old female presents with yellow skin and changes in motor behavior. The patient had a proximal arm resting tremor. She is also having difficulty walking, speaking and swallowing. Her movements are slow, tremulous, and incoordinated. Labs reveal elevated liver transaminases. Exam reveals greenish brown pigment in her iris.

What is it? Wilson's Disease. Copper deposits in the basal ganglia and causes the neurologic signs of Wilson's disease, which may include "Parkinsonism," slowed movements, tremor, ataxia, dysarthria, dysphagia, and dystonia.
Next step? Confirm diagnosis by liver biopsy with quantitative copper assay. Elevated free copper, low ceruloplasmin or elevated urine copper are suggestive (and clues to diagnosis if in vignette), but are not definitive. MRI can show changes of the basal ganglia, as well as a couple other sites.
Treatment? Copper chelation is indicated and is classically done with penicillamine (but has been replaced by the chelator trientine). Note: If not so severe (hepatic compensation and no neurologic symptoms), zinc is the therapy of choice (blocks intestinal copper absorption).

M19. A 65 year old male with history of hypertension, CAD and recent ischemic stroke presents with involuntary violent flailing of his left arm and leg.

What is it? Hemiballismus.
Next step? MRI will show a lesion in the contralateral (right) subthalamic nucleus. (They may give you the MRI report in the vignette).

Neuromuscular Disorders
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NM1. A 26 yo female comes to your office for complaints of double vision, worse at night. She also noticed her eye lids will droop some nights but are always better in the morning. When you have her smile, she looks like she is snarling. Her shoulder shrug weakens with repeated attempts against resistance. Reflexes and sensation are normal.

What is it? Myasthenia Gravis. Know this disease! Its hallmark clinical signs are fatiguable weakness that improves with rest, with common initial manifestations of ptosis and diplopia.
Pathophysiology? Autoantibodies to acetylcholine receptors (AChRs) on the postsynaptic nerve terminal.
Diagnosis? Acetylcholine receptor antibodies- 85% sensitive (If negative MUSK antibodies picking up an additional 40% of cases beyond that). EMG with repetitive nerve stimulation 70% sensitive. Tensilon (edrophonium) test could be used to see if an objective marker like ptosis transiently improves with infusion- though could be tested about, less commonly used in clinical practice at this point.
Next step after diagnosis? Screening CT of chest for thymoma. 10% have a thymoma, while 75% have an abnormality of their thymus (usually hyperplasia). Remove thymoma if suspected. Could still consider thymectomy as a treatment option with a young, generalized myasthenia patient even without a thymoma.
Treatment? Acetylcholinesterase inhibitors (pyridostigmine). Immunosuppressants are often used as well.

NM2. A 60 yo male with myasthenia gravis on pyridostigmine and prednisone presents for fever. He has recently felt feverish up to 102 and is coughing up green sputum. On exam, he has ptosis, difficulty speaking and decreased facial movement, and is much weaker than baseline, unable to lift his arms or legs off of the bed. His lung sounds are significantly diminished bilaterally. 02 sat has only dropped some to the low 90's but clinically is having significantly labored breathing.

What is it? Myasthenic Crisis. Could be secondary to a trigger like an infection in this case.
Next step? Many things would happen at once. First, evaluate respiratory parameters. Functions of respiratory strength like vital capacity (VC) and negative inspiratory flow (NIF) will demonstrate significant neuromuscular respiratory weakness prior to significant drop in oxygenation and intubation to be considered in this patient if having severe respiratory weakness. Second (and next most likely to be tested) is to start either plasmapheresis (most often) or IVIG to control the disease itself. In addition (but unlikely to be tested), antibiotics would be started for presumed infection (caution with certain antibiotics such as aminoglycosides which could exacerbate the weakness). Very high doses of pyridostigmine could also potentially cause cholinergic crisis with excessive secretions, diarrhea, fasciculations, and paradoxical weakness. This is very rare with doses typically used, though.

NM3. A 60 yo M smoker presents with hemoptysis and weakness. He complains of weakness in his legs especially while climbing stairs. In addition, he states he has noticed dry mouth and impotence. Exam reveals weakness of hip flexion and knee extension. His reflexes are absent. Sensation is intact. CXR reveals a mass near his left mainstem bronchus. Repetitive nerve stimulation shows an incremental muscle response.

What is it? Suspect Lambert-Eaton Myasthenic Syndrome (LEMS). It is characterized by weakness classically of the proximal lower limbs that improves with repetitive movement. Reflexes are often hyporeflexic. Patients may have ptosis or diplopia like MG (less common and pronounced than typical MG though), and Lambert Eaton also may have autonomic symptoms (impotence, dry mouth) unlike MG. It is typically a paraneoplastic condition and the most common associated malignancy is small cell carcinoma of the lung.
Pathophysiology? Autoantibodies against calcium channels at presynaptic motor nerve terminal.
Diagnosis? Autoantibodies to P/Q type calcium channels can be detected in 85% of patients. Incremental response to repetitive nerve stimulation is given in the vignette.
Treatment? Treatment of the underlying malignancy if present and symptomatic therapy with 3,4 Diaminopyridine (3,4 DAP), as well as immunosuppressive treatment are the top choices.

NM4. A 35 yo M presents with weakness. His wife speaks for him as he is slurring his speech. She states her husband had been previously healthy until he had acute onset of double vision, drooping of his eyelids and slurring of his speech. Soon afterward, he started to develop weakness of his whole body. On exam, the patient appears to be mentating well. He is taking shallow breaths but pulse oximetry is 95%. His pupils are dilated and poorly reactive. His mouth is dry. The patient has diffuse symmetric

weakness. Reflexes are absent. Sensation is intact. The patient is a farmer and cans his own vegetables.

What is it? Botulism. Botulism classically presents with bulbar symptoms followed by generalized weakness with diminished reflexes. There also may be autonomic symptoms like dry mouth, pupillary dilation/fixed.
Pathophysiology? A preformed toxin made by C. botulinum is ingested and inhibits presynpatic ACh release. Look for historical clues like home canning where C. botulinum can grow.
Diagnosis? Clinical diagnosis is preferable given need for quick treatment. Nerve conduction would reveal presynaptic neuromuscular blockade and reduced compound muscle action potentials (CMAPs)- similar to Lambert Eaton. Serum assays may identify the toxin.
Treatment? Equine antitoxin is given immediately! Respiratory monitoring with early intubation.

NM5. A 20 yo new mother comes in hysterical over her "floppy baby". She states the baby has been less active and had a weaker cry than usual. The baby is no longer tracking her with his eyes. She admits to giving the baby honey sometimes when the baby won't stop crying. On exam, the baby has ptosis, dilated poorly reactive pupils and no eye movement. The baby has blue lips. Respiratory effort is weak. There is trace movement of the lower extremities.

What is it? Infantile botulism. Botulism presents similarly in infants with bulbar symptoms and descending paralysis. Autonomic symptoms are helpful as well. Look for a history of feeding the infant honey, which can contain C. botulinum spores that can germinate in the GI tract and produce botulinum toxin. (Note that adults must take in preformed toxin as they can fend off the spores from germinating).
Diagnosis? Same as above, though stool is higher yield for detection of the toxin.
Treatment? Immediate intubation, ventilatory support and botulism immune globulin.

NM6. A 23 yo M presents with weakness. He states he recently went on a camping trip in the Northern Appalachian Mountains. After returning from the trip, he noticed progressive weakness that started in his feet, spread proximally and now involves his entire lower extremities and hands. On exam, the patient is wheel chair bound. You note pupils are equal and reactive. Reflexes are absent. Sensory exam is WNL. LP is performed and CSF is normal.

What is it? Tick paralysis. This is characterized by tick attachment followed by ascending paralysis within a week that will typically take about 5-7 days to potentially involve all muscles including bulbar/facial/respiratory.
Diagnosis? Finding the tick will make your diagnosis. Check hair-laden areas.
Treatment? Removal of tick will resolve symptoms. Supportive care as needed.

NM7. A 35 yo M presents with acute onset of weakness. He first noticed his weakness this morning when his legs felt "rubbery". He has noticed the weakness spreading to involve his hands then upper arms. He also notes some painful tingling in his legs. He denies fever, neck pain or back pain. He was previously healthy except for a bout of diarrhea 2 weeks prior that resolved spontaneously. On exam, his cranial nerves are intact. He has flaccid paralysis of his upper and lower extremities. Reflexes are absent. Sensation is decreased in all modalities, distal greater than proximal.

What is it? Guillian-Barre Syndrome (AIDP). This syndrome is characterized by areflexic "ascending" weakness/paralysis involving both distal and proximal muscles over the course of a few hours to days with an antecedent respiratory or GI illness weeks earlier. Cranial nerves are often involved as can sensory and autonomic nerves. Please note that while "ascending" is a term often used to describe the weakness observed in GBS, it is not actually what you would classically think of as ascending. The immune system will "hit" nerves randomly and block or slow conduction distal to that point. Therefore, distal sites are more likely to be affected, but the typical pattern of weakness is concurrent distal AND proximal weakness.
Pathophysiology? GBS is thought to be an autoimmune acute inherited demyelinating polyneuropathy (AIDP). It is thought that autoimmunity occurs after exposure to infectious/vaccine non-self antigens that mimic antigens present in myelin. C. Jejuni can be an associated pathogen.
Diagnosis? Often made clinically given characteristic pattern and due to fact that treatment is most effective when given as soon as possible. LP will characteristically show albuminocytologic dissociation (high protein, NL WBC) but protein may not rise for a week after weakness starts. Nerve conduction studies would show acute demyelination with or without axonal damage.
Treatment? Early IVIG or plasmapharesis. Supportive care with monitoring of vital capacity, blood pressure and heart rhythm. Ventilation is required in about 30% of patients and may be needed for a few weeks. Do NOT pick steroids - they are ineffective.

NM8. A 60 yo M presents with subacute onset of weakness. He first noticed his weakness eight weeks ago when he started stumbling and found his feet to feel weak. He has noticed over the past 2 months that the weakness is gradually spreading up his legs, and he is having more trouble walking. He also is getting some weakness in his arms- he now has both proximal and distal weakness. He also notes some painful tingling in his legs. He denies fever, neck pain or back pain. He was previously healthy. On exam, his cranial nerves are intact. He has 3+ strength in his lower extremities. Reflexes are absent at the patella and ankle. Toes are downgoing. Sensation is decreased to vibration in the feet. C-spine imaging is normal. LP is performed. CSF is acellular and shows protein is 5x the normal value.

What is it? Chronic Inflammatory Demyelinating Polyneuropathy (CIDP). This disease entity is very similar to GBS but can be different in a couple ways: 1) It usually presents subacutely but may be indistinguishable from GBS initially 2) It may take either a chronic progressive course or take a relapsing-remitting course. Progressive symptoms greater than 8 weeks would be consistent with CIDP rather than GBS. GBS is usually not progressive more than 4 weeks. 4-8 weeks is a gray zone.
Diagnosis? LP should show albuminocytologic dissociation as in vignette. NCS/EMG will show demyelination.
Treatment? IVIG or plasmapheresis.

NM9. A 60 yo F presents with numbness and tingling in her hands and feet for the past several years. She states the tingling is gradually progressing up her hands and legs. She states she has burning pain associated with the numb areas. Her past medical history includes hypertension, CAD and DMII. Neurological exam shows full strength in all limbs. Reflexes are absent at the ankle. Toes are downgoing. Sensory exam

reveals distal loss of pin, temperature, touch and vibration sense in hands and legs up to the knee. Serum B12, TSH, and Serum protein electrophoresis are normal.

What is it? Diabetic Sensorimotor Polyneuropathy (DSPN). This is characterized by a gradually progressive sensory loss in a stocking glove distribution. Motor symptoms may occur later and autonomic symptoms can occur.
Diagnosis? This can be diagnosed in a patient with chronic progressive stocking glove sensory loss with impaired glucose metabolism (either DM or abnormal Glucose Tolerance Test) and no other identifiable cause (nutritional (B1, B6, B12), immune/hematological disorder (paraprotein such as monoclonal gammopathy of undetermined significance, multiple myeloma, waldenstrom's, amyloid, etc), toxic (EtOH, chemo), inherited, or other metabolic (thyroid).
Treatment? Tight glucose control can help halt progression. No treatments are currently effective in reversing nerve damage. Painful dysesthesias can be controlled with antiepileptics or TCAs or other SNRIs utilized for neuropathic pain.

NM10. A 60 yo M presents with increasing dizziness upon standing. The patient reports he has DMII for the past 30 years. He had had numbness and tingling in his hands and feet for the past 20 years He also has gastroparesis. In recent years, he has impotence, urinary incontinence, and has been seen in the ED for unconsciousness secondary to hypoglycemia several times. Sensory exam reveals distal loss of pin, temperature, touch and vibration sense in hands and legs up to the thigh. Serum B12, TSH, and serum protein electrophoresis are normal.

What is it? Autonomic neuropathy secondary to diabetes. You should know the various signs of autonomic dysfunction: orthostasis, impotence, urinary incontinence, abnormal sweating, resting tachycardia, gastroparesis, constipation, dry mouth and loss of sympathetic symptoms with hypoglycemia predisposing to hypoglycemia.

NM11. A 28 year old pregnant female with hyperemesis gravidarum is experiencing paresthesias in her feet for the past week. She has had little oral intake for the past 6 weeks. On exam, she has decreased sensation past mid-calf, decreased ankle reflexes, and strength on dorsiflexion and plantar flexion of the ankle. Oral glucose tolerance test is normal. Nerve conductions show axonal damage in the distal LEs.

What's the point? Suspect a nutritional neuropathy in this patient with decreased oral intake. B1 deficiency (dry beriberi) or B6 deficiency are most likely. B1 (thiamine) is most often depleted in alcoholics but can occur in any patient with protracted vomiting. B6 (pyridoxine) is hypermetabolized in pregnancy but is classically associated with isoniazid use.
What other nutritional neuropathy should I know about? B12 deficiency. This is commonly tested, especially its association with an elevated MCV. It may present as just a distal sensory neuropathy, or more classically as Subacute Combined Degeneration of the spinal cord. (see Spinal Cord section for a vignette of this condition).

NM12. A 60 year old female was previously healthy until being diagnosed with Breast Cancer. She is undergoing treatment with paclitaxel. She has noticed progressive numbness in her distal legs after her last two treatments. On exam, no weakness is noted. Ankle reflexes are absent. Distal sensory loss to pin, temperature, light touch and proprioception are noted up to mid calf.

What is it? Drug-induced neuropathy. Many pharmaceuticals can cause nerve damage, most notably chemotherapeutics (taxol, vincristine, thalidomide, cisplatin) and antiretrovirals. They may cause damage in a variety of patterns but length-dependent neuropathy is the most common.
Diagnosis? Clinical. Key historical clues to attribute the cause of neuropathy to a toxin is by their temporal relationship and dose-response relationship.
Treatment? Cessation of the drug.

NM13. A 15 year old female presents for evaluation with her Mother. She has always been a clumsy walker and trips frequently since early in life. Her mother is concerned because she had similar symptoms early in life and her disease has progressed so that she has weakness in both her legs and hands. On exam, she has significant ankle dorsiflexor weakness bilaterally. Ankle reflexes are diminished. All sensory modalities are diminished in her feet. On inspection, she has hammer toes and high arches. She walks with a steppage gait.

What is it? Charcot Marie Tooth (type I). You only need to recognize this one. It is the most common heritable neuropathy and, depending on type, will present in early adulthood or childhood. It is characterized by distal sensorimotor loss. Wasting occurs as weakness progresses and hammer toes/high arches will be present if symptoms occur at an early enough age. Genetic testing is available. CMT type 1 is a demyelinating neuropathy and type 2 is an axonal neuropathy.

NM14. A 40 yo M with DMII presents for numbness and tingling in his right hand. His symptoms are worse first thing in the morning and after using his hand repetitive movements like chopping vegetables or working in his woodshop. On exam, he has no neck pain, full strength except for some weakness in thumb abduction. Some atrophy of his thenar eminence is noted. There is sensory loss in his thumb, 2nd and 3rd digits. Flexing his wrists and placing the dorsums of his hands together reproduces his symptoms after 20 seconds.

What is it? Carpal Tunnel Syndrome. Know this! This patient has all the classic signs of carpal tunnel syndrome. Also note that symptoms may be bilateral. The main differential for his sensory disturbance is radiculopathy so keep an eye out for neck pain, weakness or myotomal/dermatomal findings that conform to a nerve root rather than the individual peripheral median nerve.
Diagnosis? Nerve Conduction Studies to establish diagnosis and grade severity.
Treatment? Wrist splints to be worn at night (this is very high yield for the exam). Surgery is reserved for significant disease. This case with weakness and atrophy of the thenar eminence would be a severe case.

NM15. A 40 yo M with DMII presents for numbness and tingling in his right hand. His symptoms are worse first thing in the morning and after leaning on his elbows. On exam, he has no neck pain. He also has some very mild weakness in spreading his fingers and flexing his 5th digit. There is sensory loss in his fifth digit and medial aspect of 4th digit.

What is it? Ulnar nerve entrapment at the elbow. This nerve is commonly entrapped as it passes through the cubital tunnel at the elbow and patients get a feeling similar to “hitting their funny bone”.

Diagnosis? Nerve Conduction Studies.
Treatment? Conservative treatment with elbow pads or behavior modification. Surgery if conservative treatment fails or in significant cases.

NM16. A 50 year old male presents with severe neck pain radiating to his left shoulder and extending to the arm, forearm, and dorsum of the hand. On exam, you note very mild weakness of his left triceps, finger extensors, and wrist flexors, as well as decreased sensation of the third digit and a diminished triceps reflex.

What is it? Cervical radiculopathy (C7).
Diagnosis? MRI/EMG.
Treatment? Conservative treatment involves analgesics with rest and possible cervical collar or cervical traction. Surgery may be considered with significant motor deficit. Also, intractable pain despite conservative management for at least 6 weeks could be another indication.

NM17. A 20 yo M dislocates his shoulder playing basketball. He was reaching up for a rebound when his arm was hit backward and the shoulder popped out. On exam, you note a sulcus sign.

What nerve is susceptible to injury? Axillary nerve.
How can you test function of the axillary nerve? Test shoulder abduction and sensation of lateral shoulder.

NM18. A 20 yo M is mugged and beat with a baseball bat. He was hit in the right arm and it appears his humerus has been fractured along the middle of its course.

What nerve is susceptible to injury? Radial nerve as it runs in the spiral groove.
What is the best way to test function of the radial nerve? Test wrist/finger extension and sensation of the dorsolateral hand.

NM19. A 28 yo pregnant female complains of pain and numbness over her right anterolateral thigh. No motor symptoms are present. The exam is normal except for numbness in the area described.

What is it? Meralgia paresthetica. This is secondary to entrapment of the lateral cutaneous nerve as it crosses the inguinal ligament and is frequently seen in patients with rapid weight gain or obese patients or tight clothes.
Next step? Reassurance. Avoid tight clothing and standing for long periods. Treatment options in the non-pregnant include NSAIDs, drugs for neuropathic pain, or local corticosteroid injections.

NM20. A 50 yo F with long-standing rheumatoid arthritis presents with fever, a flare in her hand joints and new pain in her right foot. She had seen you last week for weakness of her left wrist and finger extensors and neuropathic pain consistent with a left radial neuropathy. She states that severe burning/electrical pain in her foot started two days ago. She complains of weakness at her ankle joint and some difficulty walking. On exam, she is weak in dorsiflexion and eversion of the right foot. She also has some tingling in both feet at this point with reduced sensation distally.

What is it? Mononeuritis Multiplex. This is a disease entity characterized by multifocal mononeuropathies due to an inflammatory cause such as vasculitis. The patient in this vignette has Rheumatoid Arthritis, but several other diseases are associated including SLE, Churg-Strauss, PAN, Wegener's, etc. If anyone has symptoms explainable by multiple mononeuropathies with an inflammatory disorder and significant pain, suspect this disease.
Localize the patients' current mononeuropathy:
Treatment? Aggressive immunosuppression with high dose steroids and potentially cyclophosphamide.

NM21. A 30 year old male presents with right sided facial weakness. He states he felt retroauricular pain two days ago followed by slowly developing weakness of his right face. He notes sensitivity to sound in his right ear and some altered taste. He is now unable to close his right eye. On exam, pupils are equal and reactive, EOMI, and facial sensation is normal. There is right facial droop and absence of a forehead furrow on the right. The ear canal is clear of any lesions. Hearing is intact. Palate elevates symmetrically. No parotid enlargement. No focal weakness is noted. Gait, cerebellar testing are normal.

What is it? Idiopathic Bell's Palsy. The patient has signs of peripheral seventh nerve palsy (forehead affected) without a clear cause. Hyperacusis and altered sensation may also be present secondary to the seventh nerve's innervation of stapedius muscle and sensory role for taste of anterior two thirds of tongue.
Diagnosis? Clinical. Forehead sparing of weakness might indicate imaging (see below), as well as atypical progressive lesions overtime to rule out local infiltrative processes.
Treatment? High dose prednisone with taper +/- antiviral (acyclovir or valtrex). Symptomatic treatment for eye protection may include lubricating drops, an eye patch, night time eye ointment or instructions to tape eyelid closed while sleeping depending on severity. Prognosis is generally very good.

NM22. A 60 year old male presents with right sided facial weakness. He states he abruptly developed weakness of his right face this morning. On exam, pupils are equal and reactive, EOMI, no nystagmus and facial sensation is normal. There is right facial droop and presence of a forehead furrow bilaterally when asked to raise his eyebrows. The ear canal is clear of any lesions. Hearing is intact. Palate elevates symmetrically. No parotid enlargement. No focal weakness is noted. Gait, cerebellar testing are normal.

What is it? Suspect a L cerebral lesion as the cause of R supranuclear facial weakness. Note that there is sparing of the forehead muscles on the affected side. This suggests a "supranuclear" cause of facial weakness. To understand why, you must understand the anatomy (best to find a picture - see Figure 2 in the following AAFP article http://www.aafp.org/afp/2007/1001/p997.html).
Diagnosis? Would recommend brain imaging with forehead sparing facial weakness.

NM23. A 60 year old man complains of extremely severe, sharp, shooting pain on the right side of his face. The pain only lasts a few seconds but is so painful he has afraid to go outside because the wind across his face can set it off.

What is it? - Tic doloreaux (trigeminal neuralgia). It is a neuropathic pain that is commonly from an abnormal vessel loop contacting the trigeminal nerve at the base of the brain or rarely from other secondary causes like multiple sclerosis.
Diagnosis? Clinical.
Treatment? Treat with carbamazepine, which is successful for most patients. Refractory cases may require interventional block of the trigeminal ganglion (injections or gamma knife radiation). A craniotomy can be considered in refractory cases to separate the vessel loop from the trigeminal nerve.

NM24. A 45 year old male presents with gradual symmetric proximal muscle weakness for the past three months. He has noticed difficulty climbing steps. He has no rash, no facial or extraocular weakness, no family history of a neuromuscular disease. He is otherwise healthy and on no medications. On exam, there is four out of five strength in the proximal muscles of the arms and legs. Repeated testing does not result in increased weakness. Involved muscles are nontender. Reflexes are intact. Serum CK is greater than 20X the normal limit. Needle EMG of the deltoid shows increased spontaneous activity with positive sharp waves and short duration, low-amplitude polyphasic units on voluntary activity (irritable myopathy findings).

What is it? Polymyositis. This is an inflammatory myopathy characterized by subacute progressive symmetric proximal muscle weakness.
Diagnosis? Muscle biopsy after EMG. This is likely beyond what you need to know, but biopsy would show CD8 + T-cell infiltrates within muscle fascicles/fibers bound to MHC-I molecules on the sarcolemma. Consider screen for underlying malignancy especially in older patients.
Treatment? Glucocorticoids and other immunosuppressants like azathioprine or methotrexate.

NM25. A 35 year old female presents with gradual symmetric proximal muscle weakness for the past month. She has noticed difficulty combing her hair and getting up from a chair. On exam, the patient has some purple discoloration of her upper eyelid, patches of red over her upper trunk, and raised purple scaly lesions over her MCPS and interphalangeal joints. She has 4 out of 5 strength in her proximal muscles with normal reflexes.

What is it? Dermatomyositis. This is also an inflammatory myopathy with similar weakness to polymyositis but occurs more often in females and has the pathognomonic heliotrope rash and gottron's papules.
Diagnosis? CK would be elevated and EMG will show findings consistent with myopathy. Muscle biopsy shows inflammation (MAC positive) and perifascicular atrophy. Consider screen for underlying malignancy especially in older patients.
Treatment? Same as for polymyositis.

NM26. A 65 yo male presents with gradually worsening weakness for the past few years. He states he first noticed some weakness in his grip in the past year and that has gradually worsened. He has also had some recent falls and states his knees will give out on him, and he has difficulty climbing stairs. Reflexes and sensation are normal. Babinski is downgoing. EMG shows an irritable myopathy.

What is it? Inclusion Body Myositis. IBM is an inflammatory myopathy that occurs in older persons (most common myopathy in patients > 50 yo) and is typically results in

gradually progressive weakness. In addition to proximal weakness, prominent muscles involved would be the finger flexors and the quadriceps. The weakness can be somewhat asymmetrical.
Diagnosis? Muscle biopsy shows inflammation with "rimmed vacuoles" and amyloid staining.
Treatment? Unfortunately, IBM does not respond well to immunosuppressive medication treatment (unlike polymyositis and dermatomyositis).

NM27. A 50 yo female with HTN and SLE presents with subacute proximal muscle weakness. She has had trouble combing her hair and climbing stairs. She denies muscle aches. She takes HCTZ 25 mg and prednisone 50 mg each day. On exam, she has severe acne. She has 4 out of 5 weakness in her proximal muscles. Reflexes and sensation are normal. Babinski is downgoing. CK is normal. EMG shows a myopathy without irritability.

What is it? Steroid-induced myopathy. They may induce a proximal muscle weakness that is similar to inflammatory myopathy but without irritability on EMG (no spontaneous discharges – which are more consistent with an inflammatory process) and usually a normal or just mildly elevated CK.
Treatment? Adjustment of steroid medication.
What other major drug-induced myopathy should I know? Lipid-lowering agents (fibrates, statins, niacin, and ezetimibe) can all cause myalgia and proximal muscle weakness with a rare occurrence of rhabdomyolysis (check CK and urine).

NM28. A 6 yo boy is brought by his mother for weakness. The mom states her son has always had trouble keeping with the other kids while they were playing. She has noticed an even bigger difference in the past year. On exam, the patient has proximal weakness. You have the boy lay supine on the floor and watch him get up. You note that he tents himself up with his arms to get to his feet and once on his feet, has to use his arms to push off his knees to bring his torso upright. You also note hypertrophied calf muscles. Serum CK is 50X normal.

What is it? Duchenne muscular dystrophy. This is the most common inherited muscular dystrophy, affects males with typical onset about age 3-5. It has a predictable course with difficulty walking around age 12 and difficulty breathing in the 2nd or 3rd decade. Cardiomyopathy also is a complication that develops.
Pathogenesis? Mutation in dystrophin gene on X chromosome.
Diagnosis? Dystrophin deficiency on muscle biopsy or mutation analysis of peripheral leukocytes.
Treatment? Glucocorticoids may slow progression up to 3 years. Treatment is supportive otherwise.

NM29. A 15 yo boy is brought by his mother for weakness. The mom states her son has always had trouble keeping with the other kids while they were playing. The boy notes that he has had more and more trouble climbing stairs. On exam, the patient has 4 out of 5 strength in his proximal extremities and 4+ in distal extremities. No facial weakness is present. You note hypertrophied calf muscles. Serum CK is 50X normal.

What is it? Becker Muscular Dystrophy. This is a related but less severe disorder compared to Duchenne and is characterized by reduced quantity of or reduced size of

the dystrophin gene. By definition, patients can walk past age 15 and most survive into the fourth of fifth decade.
Diagnosis? Muscle biopsy and DNA analysis.
Treatment? Supportive. Similar to Duchenne MD, cardiomyopathy is a complication that requires monitoring for and treatment when it develops.

NM30. A 35 year old male presents for weakness. He states he has weakness mainly in his neck, hands and feet. He also complains of muscle stiffness. On exam, you note frontal balding, temporal wasting and cataracts in his eyes. After you have him close his eyes tight, there is a considerable lag time to opening them. He has noticeable ankle dorsiflexor weakness on exam with a foot drop appearance. Reflexes are normal. When you hit his thenar eminence with your reflex hammer, his thumb involuntarily abducts and then slowly relaxes. Serum CK is normal. EKG shows first-degree AV block.

What is it? Myotonic Dystrophy.
Diagnosis? EMG usually shows myotonia. Muscle biopsy may show atrophy of type I fibers.
Treatment? (Low yield for shelf) Few patients require treatment for myotonia but phenytoin and mexiletine are preferred if treatment is desired. Treatment for cardiac conduction abnormalities depends on severity.

NM31. A 35 yo female complains of muscle weakness and aches for the past month. She states she has had difficulty combing her hair and climbing stairs. She also endorses a 10 lb weight gain, feeling cold and constipation over the past month. On exam, the patient has weakness of her proximal muscles. In reflex testing, you notice the relaxation phase is prolonged. Sensation is normal. Serum CK is 5X normal.

What is it? Hypothyroid myopathy.
Diagnosis? TSH and Free T4. Treatment? Levothyroxine.

NM32. A 68 yo M presents for follow up of muscle weakness. He had initially presented with weakness and muscle twitching of his right arm that was gradually progressing over the past 3 months. He had also noticed a change in the timbre of his voice. Since that time, the patient had a normal LP and serum protein electrophoresis. His swallowing has worsened and he now has stiffness and weakness in his right more than left leg. He denies any changes in cognition, vision or bowel/bladder function. On exam today, he has atrophy of his right upper extremity. Fasciculations are noted in the extremities and tongue. Reflexes are 3+ in the right arm and bilateral lower extremities. Sensation is intact. Plantars are upgoing.

What is it? Amyotrophic Lateral Sclerosis.
Diagnosis? Clinical based on the presence of UMN and LMN signs and progressive weakness with EMG confirmation of at least 3 of 4 levels of the neuraxis with motor neuron denervation: 3 of 4 of the following: bulbar, cervical, thoracic and lumbosacral. CNS imaging and lab testing to rule out other possibilities.
Treatment? Riluzole (rilutek) can produce a modest lengthening of survival. Care is supportive otherwise.

Neuroimmunology

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NI1. A 30 yo female presents with sudden onset of pain and blurry vision in her left eye. The vision is reported as “dim”. The pain is worse with eye movement. The patient denies ever having similar symptoms or any weakness, numbness, tingling in the past. On exam, pupillary constriction is decreased when light is presented to the left pupil compared to the right. Visual acuity is 20/100 in the left and 20/20 in the right. EOMI are intact but elicit pain. No periorbital swelling.

What is it? Optic neuritis. The hallmark of optic neuritis is blurry vision, pain with eye movement and relative afferent pupillary defect. An MRI can look for optic nerve enhancement or MS lesions. Other infectious/inflammatory syndromes can cause this and testing for those are based on other clinical clues.
Treatment? IV steroids or supportive care. Do NOT just give oral steroids. If there are MS-like lesions on the MRI (even without other history than the optic neuritis alone), you would treat with an MS disease modifying drug (such as interferons or glatiramer acetate). FYI, the rationale for treating a patient with “clinically isolated syndrome” (one clinical event due to demyelination) but at high risk for developing definite MS (lesions on MRI) is supported by the CHAMPS trial.

NI2. A 30 yo female presents with sudden onset of pain and blurry vision in her left eye. The vision is reported as “dim”. The pain is worse with eye movement. The patient denies ever having similar symptoms in the past. She does report some transient numbness/tingling and weakness in the her left leg two years ago that resolved after a couple weeks. On exam, pupillary constriction is decreased when light is presented to the left pupil compared to the right. Visual acuity is 20/100 in the left and 20/20 in the right. EOMI are intact but elicit pain. Motor strength is 5/5 diffusely. Reflexes are 3+ in the left leg. Babinski is positive on the left.

What is it? Multiple Sclerosis, relapsing remitting type. The patient fits the classic picture of dissemination in time (>1 month apart) and space (two different CNS sites) with UMN signs on exam. She would currently fit the pattern of relapsing remitting since she had a discrete episode of left leg symptoms that fully recovered.
Diagnosis? MRI to evaluate for MS lesions, classically appearing as periventricular ovoid white matter lesions perpendicular to the ventricle or white matter lesions affecting the corpus callosum or infratentorial region. LP with CSF analysis for oligoclonal bands.
Treatment? IV steroids for acute flare. Patient would be placed on maintenance therapy with interferon-beta or glatiramer acetate.

NI3. A 50 year old male presents with increased difficulty walking. He states he has had slowly worsening weakness in his bilateral lower extremities, R>L over the past 5 years. He also reports gradually worsening numbness and tingling for the past 3 years. He states his symptoms are especially bad when taking a hot shower or after physical exertion. He denies visual changes or back pain. On exam, there is a mild relative afferent pupillary defect. There is decreased strength in the lower extremities. Reflexes are 3+ in the upper extremities and patella and clonus is elicited at the ankles. Plantar response is upgoing. There is moderate deficit on finger to nose pointing and heel to shin testing.

What is it? Suspect primary progressive multiple sclerosis. This subset of MS occurs in older patients (mean age 40) and the male: female ratio is 1:1. It is characterized by

gradually progressive MS symptoms as compared to the typical relapsing-remitting type with discrete attacks.
Diagnosis? This patient would certainly get an MRI of the brain and spine to confirm the diagnosis and rule out other causes. LP with oligoclonal bands would be helpful.
Treatment? Immunosuppressant medications could be considered, although PPMS is often poorly responsive to treatment.

NI4. A 30 yo female presents with sudden onset of pain and blurry vision in her left eye. The vision is reported as "dim". The pain is worse with eye movement. Her right eye already has had some decreased vision as well. She has also had an episode of acute paralysis "from her belly button down" this past year that has recovered completely. On exam, pupillary constriction is decreased when light is presented to the left pupil compared to the right. Visual acuity is 20/200 in the left and 20/80 in the right. EOMI are intact but elicit pain. Strength and sensation are fully intact. 3+ reflexes are present in the lower extremities and upgoing babinskis. MRI of the brain is normal. MRI of the spine reveals a focal nonenhancing lesion from T9-L2. CSF is negative for oligoclonal bands. Serum is positive for antibodies against aquaporin-4.

What is it? Neuromyelitis Optica. This is an MS variant classically characterized by bilateral optic neuritis and myelitis without brain involvement.
Diagnosis? Aquaporin-4 antibodies (NMO antibodies) are present in many patients.
Treatment? IV steroids for the acute flare. Immunosuppressants or rituximab may be used.

NI5. A 30 year old female presents with fever, headache and paralysis. She had been recovering from a viral illness when her fevers started to recur, she developed headache, neck stiffness and became progressively more sleepy and confused over the period of six hours. She was noted to have a seizure on transport to the hospital. On exam, the patient is unable to move her upper or lower extremities. Reflexes are 3+ with clonus at the ankles. Babinski +. LP is performed and shows lymphocytic pleocytosis. MRI shows extensive white matter disease with large "fluffy" lesions of the brain and also lesions of the spinal cord, which enhance with contrast.

What is it? Acute Disseminated Encephalomyelitis (ADEM). This disease typically presents after vaccination or viral illness with diffuse demyelination of the brain and spinal cord leading to very severe symptoms, including headache and encephalopathy, which help differentiate it from a MS picture. Note that the entire area of white matter disease was enhancing on MRI. This indicates that the disease was active in all areas, an attribute not typical of MS.
Diagnosis? MRI and LP.
Treatment? IV steroids for 5-7 days or longer with an oral steroid taper for 2 weeks after that. If patient not showing response to the IV steroids then plasma exchange/IVIG should be considered.

NI6. A 20 year old male presents with fever, headache and neck stiffness. Labs show an elevated WBC and sodium of 115. Hypertonic saline is started. LP is performed and patient has a neutrophilic pleocytosis with gram positive lancet shaped diplococci. He is started on ceftriaxone. The next day, the patient wakes up paralyzed, except for his eyes and eyelids. Stat labs are drawn and show an mildly improved WBC and sodium of 135.

What is it? Central pontine myelinolysis leading to “Locked-In” Syndrome. It is characterized by acute quadriplegia in the setting of brisk sodium correction or hyperosmolar states.
Diagnosis? Brain MRI will show symmetric high signal in the basis pontis.
Treatment? Stop sodium correction immediately! The best is to prevent it by slowly correcting sodium deficits (no more than 10 meq/L in 24 hours).

Spinal Cord
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SC1. A 67 year old male with heavy smoking history reports the development of mid-back pain for the past two weeks that is worsening in severity. The pain is relatively constant but worsens with movement and often awakens him at night. Neurological exam shows some mild weakness in his right lower extremity, 3+ reflexes in the right lower extremity and upgoing babinski on the right.

What is it? Myelopathy in thoracic region, likely from neoplastic spinal cord compression given time course.
Next step? MRI with contrast to evaluate for compressive etiology.
Treatment? Decompressive surgery is undergone when neurological deficits are present. Radiation is used as an adjunct. IV steroids are also given acutely.

SC2. A 33 year old male IV drug abuser reports the development of mid- back pain for the past two weeks that is worsening in severity. The pain is relatively constant but worsens with movement. He reports a low-grade fever for the past few weeks as well. The patient just noticed weakness in his lower extremities this morning. Neurological exam shows some mild weakness in his bilateral lower extremities, 3+ reflexes at the knee and ankle and upgoing babinskis bilaterally.

What is it? Myelopathy in thoracic region, likely from spinal epidural abscess.
Next step? MRI with contrast to evaluate for compressive etiology.
Treatment? Urgent decompressive laminectomy with debridement. IV antibiotics right away and for 6-8 weeks to complete therapy.

SC3. A 75 year old male reports the development of neck and shoulder pain for the past few years that is worsening in severity. He also reports pain over his upper outer arms bilaterally. He has stiffness in his legs for over a year but more recently, the man has had trouble walking and some imbalance. On exam, the man has point tenderness at the neck, shooting pains down his arm with neck rotation, and some atrophy of his hand muscles. Vibratory sensation is diminished in his feet. His legs are mildly weak, right greater than left. His legs have increased tone and 3+ reflexes. Toes are upgoing bilaterally.

What is it? Suspect spondylitic myelopathy. This is essentially osteoarthritis of the cervical spine leading to radicular arm symptoms and compressing the cervical cord (corticospinal tract) leading to his lower extremity symptoms.
Next step? MRI of C-spine.
Treatment? Neurosurgical evaluation for decompression.

SC4. A 33 year old female reports the development of bilateral lower extremity weakness and numbness over the past six hours. She also reports a painful band of skin involving the T7-T9 dermatomes. She has had flu-like symptoms for the past couple days. She is incontinent with absent rectal tone. An MRI shows focal signal and enhancement at T7-T9.

What is it? Transverse myelitis. Her symptoms describe a myelopathy and the MRI confirms an inflammatory cause - therefore a myelitis. Transverse myelitis can be postinfectious, acute infectious or related to systemic disorders or demyelinating disorders.
Next step? MRI was your first step to characterize and rule out a compressive cause. A workup to search for the cause would be appropriate including LP, serum studies, immune studies and brain MRI.
Treatment? IV methylprednisolone. Supportive care should be provided such as a urinary catheter.

SC5. A 70 year old patient undergoes repair of an abdominal aortic aneurysm. 6 hours after the surgery, the patient complains of weakness and numbness in his legs. On exam, the patient cannot move his legs and has diminished light touch, pain, and temperature sensation up to T8. Vibration and proprioception are intact throughout.

What is it? Anterior spinal artery syndrome. Diminished flow in the anterior spinal artery due to surgery/embolism/stenosis leads to extensive infarct of the spinal cord but spares the dorsal columns.

SC6. A 25 year old female presents to the ED after burning her hands in the kitchen at work. She notes she didn't know the pot was hot and didn't feel much pain. On exam, her hands reveal second degree burns. She has diminished pain and temperature sensation over her shoulders and upper extremities. Her grip strength and intrinsic hand muscles are weak. Lower extremity sensation and strength are intact.

What is it? Central cord syndrome due to syringomyelia.
Next step? Do MRI of cervical spine if not already done in vignette; it will show cavitation/csf signal of the central cord with.

SC7. A 16 year old male loses control of his car and slams into a tree. When the ambulance arrives, he reports his "legs don't work". The ED physician performs an exam and notes the patient has decreased vibration, proprioception, and 2-point discrimination on the right side below the nipples. He has decreased pin prick and temperature sensation on the left side two levels below the nipples. He has trace movement of his right lower extremity. Lower extremity reflexes are absent.

What is it? Right Brown-Sequard Syndrome (hemisection of spinal cord) at level T4.
Worth noting: Note pain and temperature are lost contralateral to the lesion and are spared for approximately two levels below due to the tract of Lissauer that runs rostral prior to crossing over. Also be aware that the nipples are at T4 and umbilicus at T10. Also, reflexes can absent despite an UMN lesion in the acute phase due to spinal shock.

SC8. An 80 year old female complains of chronic, progressive lower back pain that radiates to her legs. She also complains of leg weakness and numbness in a saddle-like distribution between her thighs. She is incontinent of urine. Exam reveals 1+ reflexes at the knee and absent at the ankles.

What is it? Cauda Equina Syndrome likely from lumbar spinal stenosis.
Diagnosis? MRI of lumbar spine to evaluate for stenosis or other compressive etiology.
Treatment? Neurosurgical intervention. Give IV steroids if oncologic compression.

SC9. A 65 year old male presents 10 months after gastric bypass with complaints of weakness, confusion and numbness in his feet. On exam, the patient has distally decreased proprioception and vibration sense. He has a broad-based gait and appears unsteady. He is able to stand on his own, but starts to fall when he closes his eyes. Reflexes were 3+ in the arms/patellars and trace at the ankles. CBC reveals an MCV of 110 and hypersegmented neutrophils.

What is it? Subacute Combined Degeneration secondary to Vitamin B12 deficiency. This syndrome is characterized by dorsal column and corticospinal/spinocerebellar tract dysfunction as well as a stocking-glove neuropathy.
Next step? Serum B12 (and methylmalonic acid and homocysteine levels if equivocal). MRI may show hyperintense lesions in the dorsal columns. Evaluate for cause of B12 deficiency once confirmed (gastric bypass in this patient).
Treatment? Intramuscular Vitamin B12 - first qday, then qweek to build up stores. Patient will then be transitioned to a qmonth schedule for chronic maintenance.

SC10. a 75 yo male complains of leg pain and paresthesias radiating down from his back bilaterally with walking. It is often relieved with rest but more reliably relieved by flexing at the waist. Ankle-Brachial Index is 0.9.

What is it? Neurogenic claudication.
Diagnosis? MRI of lumbar spine.
Treatment? Neurosurgical consultation for decompression.

SC11. An 18 year old suffered a C5 complete traumatic spinal cord injury one year ago. His caretaker notes the patient has an unusually high blood pressure of 150/100, up from the typical 85/55. In addition, the patient is sweating profusely and complains of an intense headache.

What's the point? This patient may be suffering from autonomic dysreflexia. Patients with SCI may have baseline low blood pressures and even modest elevations should cause you to think of autonomic dysreflexia. Autonomic dysreflexia can occur in SCI patients with lesions at T10 or above and is a response to a noxious stimulus below the lesion level that will activate sympathetic tone and increase blood pressure. Your body's blood pressure "censors" are above the lesion and cannot communicate back to halt the sympathetic reflex. This can lead to life-threatening hypertension.
Next step? Search for an inciting noxious stimulus. Common stimuli that can cause autonomic dysreflexia include bladder distention, hangnail, tight fitting clothing or stool impaction. You can also sit the patient up and give clonidine (alpha2 agonist) to help lower the blood pressure.

Sleep Disorders
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SL1. A 35 year old female presents with complaints of difficulty sleeping. She states that when she gets in bed, she has a "creepy crawly" sensation in her legs that is relieved when she moves her legs.

What is it? Restless Legs Syndrome (RLS).
Diagnosis? Clinical based on four criteria: 1) Urge to move limbs 2) Uncomfortable feeling relieved with movement 3) Symptoms worse at night 4) Symptoms worse with rest
Next step? Check for secondary causes such as iron deficiency anemia. Treat with dopamine agonists like ropinirole, pramipexole or levodopa.

SL2. A 50 year old obese male presents with excessive daytime sleepiness. He states he feels tired all the time and reports feeling in a "fog". He will often doze off on the bus or while sitting at his desk at work. He reports 8-10 hours of sleep each night but wakes up twice to use the restroom. His wife complains that he has always been a snorer. He denies nighttime caffeine, trouble falling asleep, cataplexy, or hallucinations. On exam, he has hypertrophied tonsils. CBC reveals a hemoglobin of 18.

What is it? Suspect Obstructive Sleep Apnea. This is the most common medical cause of excessive daytime sleepiness. The airway closes when upper airway muscles relax during sleep. These dilating muscles no longer oppose the negative pressure produced during inspiration, and the airway closes, producing apnea. The body's normal response is to arouse, which causes the muscles to fire and open the airway but leads to sleep impairment. Suspect it in middle aged obese males who snore loudly with polycythemia, a "crowded" airway (large tonsils or mallampati class IV), and/or nocturia (due to release of ANP from R atrial enlargement from pumping against higher lung pressures at night). Patients may have medical complications of hypertension and diabetes mellitus.
Next step? Polysomnography (Sleep Study). It is difficult to predict clinically who will have OSA.
Treatment? CPAP at night. The positive pressure will help keep the airway open.

SL3. A 17 year old female presents with excessive daytime sleepiness with frequent brief naps. She states she gets approximately 6-8 hours of sleep with occasional awakenings. She decided to see a doctor after awakening yesterday morning and not being able to move any of her limbs. Upon questioning, she has noticed a similar phenomenon of paralysis when she has been tickled in the past. She has also noticed "seeing things" when falling asleep.

What is it? Narcolepsy.
Diagnosis? Mean sleep latency < 8 minutes with demonstration of early-onset REM + symptoms of cataplexy (paralysis with emotion), hypnogogic/hypnopompic hallucinations, and sleep paralysis (paralysis upon awakening). Associated with a reduction of hypocretin/orexin in the csf though testing for that is primarily in the research setting rather than clinic setting.
Treatment? Stimulants like modafinil for daytime sleepiness. Antidepressants may be used for REM symptoms like cataplexy/hallucinations/sleep paralysis. Gamma hydroxybutyrate can be used in refractory cases.

SL4. A 23 year old college student complains of difficulty sleeping. He just recently returned from a study abroad trip to Siberia. He states he has difficulty falling asleep until 3 or 4 am and then has difficulty arising in the morning, leading him to sleep until around 11 am. He denies naps, snoring, episodic paralysis or abnormal leg movements.

What is it? Circadian Rhythm Sleep Disorder. This disorder results from changing time zones or performing shift work.
Treatment? Light therapy or sleep aids may be beneficial.

Neurootology
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NT1. A 35 year old female presents with vertigo. She has had a "cold" for the last couple days and has been suffering severe constant vertigo for the past six hours. She also complains of difficulty hearing out of her left ear. She is nauseated and has vomited twice. The vertigo is worse with head movement. On exam, her tympanic membrane is slightly erythematous on the left, she has nasal erythema, no meningismus. All other cranial nerves are intact. There is some horizontal nystagmus to the right.

What is it? Acute viral labyrinthitis. Suspect this condition in acute onset, severe vertigo with hearing loss. The vertigo will typically be constant for the first few days and then become periodic, exacerbated by sudden movements.
Diagnosis? Clinical generally without need for testing. Audiogram could show sensorineural hearing loss or electronystagmogram would show caloric hypofunctioning. MRI would rule out central cause.
Treatment? Supportive with antiemetics and hydration. Benzodiazepines or meclizine may reduce vertiginous symptoms.

NT2. A 35 year old female presents with vertigo. She has been suffering severe constant vertigo for the past six hours. She denies hearing loss. She is nauseated and has vomited twice. Vertigo is worse with head movement. On exam, her tympanic membrane is normal on the left, no meningismus. All other cranial nerves are intact. There is some horizontal nystagmus to the right.

What is it? Vestibular neuronitis. This condition is an idiopathic neuropathy of the vestibular nerve (CN VIII). It presents similarly to labyrinthitis in that it is sudden, constant and severe but is differentiated in that there is no associated hearing loss (because cochlea not inflamed).
Diagnosis? Clinical generally without need for further testing. Electronystagmogram shows caloric hypofunctioning. MRI may be performed to rule out central cause.
Treatment? Benzodiazepines, anticholinergics, and antihistamines for vertigo, and antiemetics for symptom control of vertigo/nausea. A short course of steroids could be given.

NT3. A 65 year old female presents with vertigo. She has had retroauricular pain for the last couple days and has been suffering severe constant vertigo for the past six hours. She also complains of difficulty hearing out of her left ear. She is nauseated and has vomited twice. On exam, you note vesicular lesions in the left ear canal and around the

pinna. She has no meningismus. All other cranial nerves are intact. There is some horizontal nystagmus to the right.

What is it? Herpes Zoster Oticus. The syndrome is similar to labyrinthitis but differentiated by the presence of vesicular lesions.
Treatment? Use antivirals early with supportive measures to prevent pain.

NT4. A 35 year old female presents with recurrent vertigo. She has been suffering severe constant vertigo for the past six hours. She also complains of difficulty hearing out of her left ear with a sense or aural fullness and tinnitus. She is nauseated and has vomited twice. She states she has had two similar episodes in the past. On exam, her tympanic membranes are clear. All other cranial nerves are intact. There is some horizontal nystagmus to the right.

What is it? Meniere's Disease. This is characterized by recurrent unilateral vestibular dysfunction with associated hearing loss, tinnitus, and aural fulness.
Diagnosis? Same as above. Audiogram also can have a distinctive feature of low tone hearing loss.
Treatment? Symptomatic treatment of acute vertigo as above. Low salt diet to prevent recurrent attacks. Also, low nicotine, caffeine, and alcohol intake recommended. Diuretics if low salt diet not sufficient. Hearing loss can become a chronic issue. For refractory vertigo spells, ablative procedures or endolymphatic sac surgery could be considered.

NT5. A 35 year old female with history of migraine headaches presents with recurrent vertigo. She has been suffering severe constant vertigo for the past six hours. She denies hearing loss. She is nauseated and has vomited twice. She states she has had two similar episodes in the past. On exam, her tympanic membranes are clear. All other cranial nerves are intact.

What is it? Vestibular migraine. This is characterized by recurrent unilateral vestibular dysfunction WITHOUT associated hearing loss. In other words, suspect this if a patient would qualify for meniere's disease but has normal hearing.
Diagnosis? Excluding other pathology.
Treatment? Migraine prevention.

NT6. A 55 yo male is being treated in the hospital for Klebsiella pneumoniae with IV gentamicin. Four days into his hospital stay he complains of decreased hearing. He also states his vision is blurry and 'bouncy'. He denies nausea or vomiting. On exam, visual acuity is 20/20 bilaterally, tympanic membranes are clear, hearing is decreased bilaterally, otherwise cranial nerves are intact. No nystagmus. Gait exam reveals a wide-based gait.

What is it? Acute bilateral vestibular toxicity secondary to gentamicin (aminoglycoside). Bilateral vestibular toxicity typically does not present with severe vertigo and nausea/vomiting. Instead it presents with hearing loss, oscillopsia (bouncy and blurry vision) and difficulty walking, especially in the dark.
Treatment? Stop the offending drug. Unfortunately, hearing loss is typically permanent but the patient may see some improvement in oscillopsia and gait.

NT7. A 48 year old female presents with vertigo. She states she is having frequent attacks of vertigo that last approximately 20 seconds and tend to occur at night when she rolls over in bed. She denies hearing loss, nausea or vomiting. On exam, you have her sit with her head rotated 45 degrees and then lay her down with her neck slightly extended. This maneuver brings on the vertigo and you note rotatory nystagmus.

What is it? Benign Paroxysmal Positional Vertigo (BPPV).
Diagnosis? The Dix-Hallpike maneuver may bring on the symptoms.
Treatment? The Epley maneuver for canalith repositioning.

NT8. A 65 yo male presents with gradually progressive unilateral hearing loss on the right. He also states he has a mild chronic headache and some ringing in his right ear. When prompted, he reveals a very mild disturbance in his balance. The Rinne test shows depreciation of air more than bone conduction on the right, and the Weber test is heard best in the left ear. Tympanic membranes appear normal.

What is it? Suspect an acoustic neuroma (vestibular schwannoma) until proven otherwise in a patient with progressive unilateral sensorineural hearing loss. This is a tumor on the CN VIII at the cerebellopontine angle, explaining the hearing issues. While the classic presentation is gradual hearing loss due to nerve compression, sudden changes may occur due interruption of the nerve's blood supply. Tinnitus and headache are common accompanying symptoms. Although patients may have subtle balance issues, it is typically not an issue due to central compensation. More advanced disease may involve CN VII, causing facial weakness.
Diagnosis? MRI with gadolinium shows an enhancing lesion at the cerebellopontine angle.
Treatment? Surgical removal vs. Stereotactic radiotherapy vs. observation are the options depending on the clinical scenario.

NT9. A 22 yo male presents with gradually progressive unilateral hearing loss on the right. He also states he has a mild chronic headache and some ringing in his right ear. When prompted, he reveals a very mild disturbance in his balance. The Rinne test shows depreciation of air more than bone conduction on the right, and the Weber test is heard best in the left ear. Tympanic membranes appear normal. Audiometry is performed and shows diminished hearing in both ears, right greater than left. An MRI with gadolinium enhancement shows bilateral enhancing lesions at the cerebellopontine angles.

What is it? Bilateral acoustic neuromas in a young patient are the hallmark presenting sign of neurofibromatosis type II. See ONC section for diagnosis and treatment.

NT10. A 69 year old female with history of hypertension and atrial fibrillation reports sudden onset diplopia and vertigo since yesterday. She states the vertigo is mild and is not worse with head movement. She reports no hearing loss. She is not nauseated or vomiting. On exam, she is mildly dysarthric. She is unable to deviate her right eye laterally and reports diplopia on rightward gaze. Both vertical and horizontal nystagmus are noted. Palatal elevation is asymmetric. Her tympanic membranes are clear.

What is it? Central vertigo likely secondary to embolic stroke in the posterior circulation. Note that central vertigo tends to be a constant mild sensation generally without hearing

loss. The presence of other cranial nerve deficits is highly suggestive of a central process and requires imaging investigation. Vertical nystagmus is specific for central vertigo but not always present. Nystagmus in all gaze directions or nystagmus that does not extinguish are other red flags for central vertigo.
Diagnosis? Stat CT if presents within acute stroke window to screen for tpa treatment. MRI brain otherwise.
Treatment? Acute stroke treatment. H1 receptor antagonists and benzodiazepines may improve vertigo as they act centrally. Vestibular rehabilitation therapy may be indicated if persistent.

NT11. A 65 yo male presents with difficulty hearing. His wife states he has trouble hearing her, especially in public places like a shopping mall. She states the problem has progressively gotten worse over the years. He reports no imbalance, headache or tinnitus. Weber test shows no laterality. Rinne test shows air over bone conduction bilaterally. Audiometry shows bilateral sensorineural hearing loss in the high frequency range.

What is it? Presbycusis. This is age-related hearing loss.
Diagnosis? Audiometry as above with clinical history of gradually progressive bilateral hearing loss.
Treatment? Hearing aid.

Neurooncology & Neurocutaneous Disorders
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A few words on brain tumors. They can present in three main ways: 1) progressive focal deficit 2) seizure 3) symptoms of ICP or a combination of any three. Primary brain tumors will tend not to have systemic symptoms (weight loss, fevers, fatigue), while malignant brain tumors will. Primary brain tumors will arise from the "support cells" of the CNS: either astrocytes, oligodendrocytes, Schwann cells, ependymal cells, lymphocytes, meninges or primitive neuroectodermal cells. The key to differentiating them is to know the characteristic biopsy appearances.

ONC1. An 11 year old boy presents with progressive imbalance, headache and vomiting. The boy has had more difficulty walking for the past few weeks. His headaches are worse in the morning and have woken him from sleep. On exam, he has unilateral ataxia and a positive Romberg sign. MRI shows an enhancing mass in the cerebellum with little surrounding edema.

What is it? Pilocytic astrocytoma. This is the most common primary brain tumor in children. It is a low-grade astrocytoma that typically occurs in the cerebellum.
Diagnosis? MRI as above. Biopsy will show "spindle-shaped" cells and Rosenthal fibers.
Treatment? Complete surgical resection is typically curative for this low grade malignancy.

ONC2. A 5 year old boy presents with progressive headache and vomiting for the past 3 months of unclear etiology and recent onset of imbalance. Exam shows papilledema, ataxia and a positive Romberg. MRI shows a large enhancing midline cerebellar mass

with compression of the fourth ventricle. Biopsy reveals small cells with hyperchromatic nuclei in a “pseudorosette pattern”.

What is it? Medulloblastoma. Medulloblastoma accounts for 30% of pediatric intracranial tumors. It most commonly presents with hydrocephalus and symptoms of increased ICP and cerebellar findings.
Diagnosis? Most often made by pathology after surgery.
Treatment? Combination of surgery, radiation and chemotherapy based on staging and pathology. Ventricular shunt may be considered as well.

Note: An ependymoma will present very similarly in young children with symptoms of hydrocephalus. This makes sense since ependymal cells line the ventricles and so overgrowth could lead to obstruction of flow. Clues to suggest an ependymoma over a medulloblastoma would include 1)mass confined to ventricle on imaging 2) calcifications on CT or 3) blepharoplasts on biopsy/pathology.

ONC3. A 60 year old male presents for seizures. He has had two secondarily generalized seizures in the past week. He also reports a three month history of diffuse, progressive headache. Exam shows papilledema and mild LE weakness with increased reflexes and upgoing toes on the right. MRI shows a mass with heterogenous contrast enhancement and surrounding vasogenic edema of the left side of the brain extending across the corpus callosum. Biopsy shows focal necrosis with pseudopalisading of malignant nuclei and endothelial proliferation, resembling a glomerulus in structure.

What is it? Glioblastoma Multiforme. This is an all-too-common Grade IV astrocytoma. It typically presents in middle-to-late adulthood.
Diagnosis? MRI and biopsy. Key clues to suggest GBM are a large tumor that crosses the corpus callosum, heterogenous contrast enhancement, “pseudopalisading” cells and a “glomeruloid” appearance.
Treatment? Dexamethasone to decrease edema and improve symptoms. Surgery, radiation, and chemotherapy (such as temodar, etc) but poor prognosis of around a year or so on average.

ONC4. A 40 year old female presents with new onset seizures. Exam shows papilledema but no focal neurologic deficits. MRI shows a well-defined calcified mass near in the right temporal lobe. Biopsy reveals sheets of regular cells with spherical nuclei surrounded by a halo of clear cytoplasm, resembling “fried eggs”.

What is it? Oligodendroglioma.
Diagnosis? Imaging may show calcifications and biopsy shows “fried egg cells”. 1p19q status portends better response to treatment.
Treatment? Surgery with systemic chemotherapy with a median survival around 7 years.

ONC5. A 35 year old with HIV reports right-sided progressive hemiparesis for the past few months and complaints of progressive headache worse with lying down. An MRI shows a single enhancing periventricular lesion. CSF is positive for EBV.
What is it? Primary CNS Lymphoma. This can present as either a single or multiple lesions that may or may not enhance and so can present like toxoplasmosis.
What is Secondary CNS Lymphoma? This occurs in a patient with progressive B-cell lymphoma with involvement elsewhere that has metastasized to the CNS (leptomeninges is the most common site).

Diagnosis? EBV in the CSF is relatively specific for this condition.
Treatment? See infectious disease section.

ONC6. An 8 year old boy presents with growth delay. The boy had been growing normally but has sharply fallen off the growth curve over the past year. The parents state the boy has been eating normally. ROS reveals a chronic headache for the past six months. On exam, you note decreased peripheral vision. Lab investigation shows low IGF-1 and Growth Hormone levels show negligible rise after insulin administration.

What is it? Craniopharyngioma. Craniopharyngiomas are benign suprasellar masses that can present with visual field defects, hypopituitarism and/or headaches. There is a bi-modal age of presentation, most frequently between 5-10 yo or 60-70 yo.
Diagnosis? MRI will show a cystic suprasellar mass.
Treatment? Transcranial or transphenoidal surgical resection followed by postoperative radiation if incomplete resection.

ONC7. A 65 year old female smoker presents with progressive R sided weakness, headache and seizure. On exam, she has papilledema and weakness of her R UE and LE with increased reflexes. She also complains of fatigue, weight loss and fevers for the past 2 months. An MRI shows multiple enhancing spherical lesions at the gray-white junction, one in the left frontal lobe, one in the left temporal lobe and one periventricular.

What is it? Brain metastases in a patient with no known primary cancer. Up to one third of patients with brain metastases do not have a known underlying malignancy at the time of discovery.
What is the mechanism of brain metastasis? Hematogenous
What is the next step? Search for the primary cancer with exam/history clues and further studies- i.e. CT (chest/abdomen/pelvis). Most likely primaries to metastasize to the brain are lung, breast, melanoma, and renal cell carcinoma.

ONC8. An 8 year old female presents with painless slowly progressive vision loss and proptosis of the right eye. She has optic atrophy on fundoscopic examination. She has inguinal and axillary freckling with multiple rubbery subcutaneous nodules presents on her back and extremities and also hyperpigmented macules on her trunk. Her mother has similar nodules.

What is it? Optic nerve glioma in a patient with Neurofibromatosis, type I (NFI). The syndrome is characterized by cafe-au-lait patches, neurofibromas, axillary/inguinal freckling, iris hamartomas, sphenoid dysplasia (long-bone abnormalities), and high risk of optic nerve glioma (~12%). Optic nerve gliomas present with gradually progressive vision loss, proptosis and possibly strabismus.
Diagnosis? MRI will reveal a fusiform enlargement of the optic nerve.

ONC9. A 65 year old male with Neurofibromatosis type II presents with progressive weakness and numbness in her right leg over the past couple months. She also complains of a headache with nausea and vomiting. Exam shows hyperreflexia and spasticity on the right with an upgoing Babinski. MRI shows a large uniformly enhancing dural-based mass along the sagittal sinus compressing the temporal lobe.

What is it? Meningioma in a patient with Neurofibromatosis type II (NFII). Meningiomas are almost always benign and only cause symptoms by compression - either focal deficits, seizures, or nonspecific neurologic signs. Patient with NFII are at increased risk of vestibular schwannomas (otherwise known as acoustic neuromas - see Neurootology section for vignette), meningiomas, spinal tumors or children with the disease may get posterior subcapsular cataracts. Cutaneous manifestations may be seen.
Diagnosis? MRI showing a "dural-based" lesion is characteristic. Biopsy may be necessary to rule out dural metastasis.
Treatment? Surgical resection is curative. Observation if asymptomatic.

ONC10. A 2 year old male with mild mental retardation and epilepsy presents for continued seizures despite treatment with levitiracetam. On exam, you note several hypopigmented lesions on the child's neck and trunk. You perform an MRI, which shows subependymal nodules. At this point, you question the father, who reveals he also has seizures. You note he has numerous papules on his face concentrated around the bridge of his nose and medial cheeks.

What is it? Tuberous sclerosis. Tuberous sclerosis is characterized by cutaneous lesions, seizures, and mental retardation. Cutaneous manifestations include ash-leaf shaped hypopigmented macules, adenoma sebaceum (facial angiofibromas), and shagreen patches (lumbosacral skin thickening and yellowing) in that order of frequency.

Diagnosis? Clinical based on a combination of findings. MRI will show cortical tubers and/or subependymal nodules, which may be calcified.
Next step? Echocardiogram and renal ultrasound to look for cardiac rhabdomyoma and renal angiomyolipoma, respectively.
Treatment? Infantile spasms are a common seizure manifestation and treatment may include vigabatrin (concern for retinal toxicity) or ACTH. Periodic MRI is typically done to look for subependymal giant cell astrocytomas, which are benign but may cause hydrocephalus.

ONC11. A 25 year old female presents with progressive imbalance. She states she difficulty walking and tends to fall to the left. She has no other complaints. On fundoscopic exam, you note a dilated artery leading from the optic disc to a peripheral tumor with an engorged vein. You note difficulty with finger-to-nose and heel-to-shin testing on the left. Her gait is ataxic. MRI reveals a well-defined hypervascular enhancing mass in the left cerebellum.

What is it? Cerebellar hemangioblastoma in a patient with Von Hippel-Lindau Disease (VHL). VHL is characterized by CNS hemangioblastomas (retinal, cerebellar, spinal) and renal/liver cysts. Patients are at increased risk for developing pheochromocytoma and renal cell carcinoma.
Next step? Renal imaging.
Treatment? Surgical excision.

ONC12. A 1 yo male presents for management of seizures. The child has had seizures since shortly after birth that are poorly controlled on carbamazepine. On exam, the child has a maroon patch of skin in the V1 and V2 distribution on the right side of his face. CT

scan shows calcifications in the right temporoparietal lobe.

What is it? Sturge Weber Syndrome (encephalotrigeminal angiomatosis). This syndrome is characterized by the classic Port-wine stain in V1 and V2 with or without leptomeningeal angiomas. Be able to recognize in a picture.
Diagnosis? CT may show calcifications or MRI with contrast may show pial angiomas.
What to watch for? Most patients have seizures, which may be intractable to medications. Patients may also have developmental delay, have "stroke-like" episodes or develop glaucoma. This disorder is not inherited unlike the autosomal dominant inheritance of Tuberous Sclerosis and Neurofibromatosis.
Treatment? Seizure control +- glaucoma therapy.

ONC13. A 5 year old female presents for difficulty walking. The parents state she has had an abnormal gait since starting to walk and the diagnosis of cerebral palsy had been considered. On exam, you note dilated conjunctival vessels against a white conjunctival background. The patient has dysarthric speech. She walks with a broad-based unsteady gait. She is equally off balance with her eyes open or closed. Lab results show a low lymphocyte count and low IgA levels. An MRI shows cerebellar atrophy and a dilated fourth ventricle.

What is it? Ataxia-telangiectasia. This autosomal recessive disorder is characterized by cerebellar findings (ataxia, dysarthria), ocular telangiectasias, and eventually early death from recurrent pulmonary infections or cancer (Hodgkin's lymphoma most common).
What is the pathophysiology? Defect in DNA damage repair.

ONC14. A 14 month old male presents for "crooked eyes". The mom says his right eye has been "lazy" for the past two weeks. On exam, you note leukocoria in the right eye. Fundoscopic exam reveals a white retinal mass. CT scan reveals a retinal-based intraocular mass with calcifications.

What is it? Retinoblastoma.
Diagnosis? Ultrasound or CT.
Treatment? Radiation or chemotherapy. Enucleation (removal of eye) is reserved for cases where vision cannot be salvaged.

ONC15. A 24 year old female with presents headache, amenorrhea and milky breast discharge for the past few months Exam reveals decreased peripheral vision. Lab testing reveals elevated prolactin and calcium levels. PTH is high.

What is it? Suspect Multiple Endocrine Neoplasia type I. The syndrome is characterized by the 3 P's: Pituitary adenomas, Parathyroid hyperplasia, and Pancreatic (and duodenal) neuroendocrine tumors. Suspect this condition in anyone with two of the three P's or one of the P's with a positive family history.

Pediatric Neurology
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Pediatric neurology is a difficult field for shelf preparation. It's low yield; you're likely to see only a few questions on it. Yet it is filled with many clinical syndromes that students

have little clinical exposure to and have trouble keeping straight. Luckily for you, mere recognition (diagnosis) of the syndrome is all that would typically be expected, unless otherwise indicated.

P1. A 12 year old female presents with chronic headache and neck pain. The headache is worse with coughing. On exam you note papilledema and diminished pain and temperature sensation in the bilateral upper extremities. Back exam is normal. An MRI reveals no mass lesions, no meningeal enhancement, but the inferior portion of the cerebellar tonsils lie below the level of the foramen magnum.

What is it? Arnold-Chiari Malformation type I. This can be present in older children or in adults. Low lying cerebellar tonsils may obstruct CSF flow causing hydrocephalus, headache and possibly syringomyelia of the spinal cord.
Diagnosis? MRI.
Treatment? Decompressive surgery for symptomatic patients.

P2. A one month old male presents with progressive head enlargement. The head circumference is greater than the 99th percentile. He has had poor feeding and vomiting for the past few days. On exam, you noted dysjunction of the sutures, a tense fontanelle, and dilated scalp veins.

What is it? Hydrocephalus.
Next step? CT or MRI.

P3. A 15 year old male presents after having a generalized seizure with LOC. This is his first seizure. Further history reveals he has had quick occasional bilateral arm jerks when he wakes up in the morning for the past two years. He retains consciousness during those jerking episodes.

What is it? Juvenile Myoclonic Epilepsy (JME). Recognize this syndrome by onset in early adolescence and the presence of myoclonic jerks preceding development of generalized seizures a couple years later.
Treatment? Depakote has traditionally been considered first line.

Other newer, “cleaner” AEDs that cover for generalized epilepsy would include levetiracetam (keppra), topiramate (topamax), and lamotrigine (lamictal).

P4. A 7 year old male presents for management of seizures. The boy has had two seizures, one was noted while he was sleeping at a sleep over. The other seizure occurred during wakefulness. The boy states he felt “funny sensations” along the left side of his tongue and inner cheek and was suddenly unable to talk, followed by rhythmic jerking of his left face. He retained consciousness during the episode.
Interictal EEG reveals high voltage centrotemporal spikes.

What is it? Benign rolandic epilepsy. Recognize this syndrome by hemifacial or secondarily generalized seizures which are preceded by a somatosensory aura in that area and occur more frequently during sleep. EEG shows centrotemporal spikes.

P5. A 4 year old female with developmental delay presents for management of seizures. She has multiple types of seizures: generalized, atonic, and atypical absence seizures. EEG is notable for 2 Hz spike-and-wave discharges.

What is it? Lennox-Gastaut Syndrome. Recognize this syndrome by a triad of 1) multiple types of seizures 2) cognitive dysfunction and 3) <3 Hz spike-and-wave discharges.

P6. A 6 month old male with developmental delay presents for “spasms”. The parents note that the child will suddenly flex his trunk, bringing his head toward his knees, and then slowly relax back over a few seconds. This is then repeated 10-20 times during his typical attacks. He typically has 2-3 attacks per day. Interictal EEG shows chaotic disorganized rhythms with superimposed multifocal spikes, termed hypsarrhythmia.

What is it? West Syndrome (Infantile spasms). Recognize this syndrome by the triad of 1)spasms 2)hypsarrhythmia and 3)mental retardation.
Treatment? ACTH or prednisolone are first-line. They are thought to work by suppressing CRH.

P7. A 20 month old male presents for developmental delay. The boy was born prematurely at 30 weeks. The boy began to sit at 10 months and is unable to walk. On exam, his lower extremities exhibit increased tone with 3+ patellar reflexes and clonus at the ankles bilaterally. MRI shows periventricular leukomalacia.

What is it? Cerebral palsy. This is characterized by a nonhereditary, nonprogressive (fixed) movement and postural disorder. Look for clues of perinatal neurologic insult paired with abnormal movements (spastic paresis most common).

P8. A 30 year old female with history of bipolar disorder gives birth to a healthy male at 40 weeks gestation. The child is asymptomatic but a small tuft of hair is noted in the midline lumbosacral area. An x-ray reveals bony deficits at L5 and S1.

What is it? Spina Bifida Occulta.

P9. A 10 year old child presents for learning difficulty. He has never done well in school and deficits are noted across all subject areas. On exam, you note he has a long narrow face with large ears and a prominent jaw. Testicular exam reveals macroorchidism.

What is it? Fragile X Syndrome.
What is the genetic abnormality? Increased CGG repeats on the X chromosome.

P10. A 6 month old male of Ashkenazi Jewish ancestry presents for developmental delay. On exam you note a cherry red macula. No hepatosplenomegaly is present.

What is it? Tay-Sachs disease.

P11. A 6 month old male of Ashkenazi Jewish ancestry presents for developmental delay. On exam you note a cherry red macula. Hepatosplenomegaly is present.

What is it? Niemann-Pick disease.

P12. A 5 year old female presents for developmental delay. The parents report a prominent speech delay with minimal use of words. The child had motor delays growing

up and now has an ataxic gait. The child's demeanor appears is characterized by happiness and easy excitability, and she makes frequent hand-flapping movements.

What is it? Angelman syndrome.
Pathophysiology? Loss of maternal contribution to Chromosome 15.

P13. A 5 year old male presents for developmental delay. The parents report a prominent speech delay with minimal use of words. The child had hypotonia at birth and now is generally uncoordinated. He also had poor feeding in infancy but now is "always eating" and has gained weight excessively.

What is it? Prader-Willi Syndrome.
Pathophysiology? Loss of paternal contribution to Chromosome 15.

P14. A 1 yo female presents with regression of developmental milestones. She had been developing normally until approximately 10 months of age. She no longer will crawl, use her hands purposefully, or say "mama" or "dada". You note hand-wringing and teeth grinding on exam.

What is it? Rett Syndrome.

P15. A 40 year old female gives birth to a male at 38 weeks gestation. The child is noted to have brachycephaly, bilateral epicanthal folds, small ears, and a heart murmur.

What is it? Trisomy 21 (Down's Syndrome).
What are the potential neurologic complications? 1)Mental retardation 2)Atlantoaxial instability may lead to odontoid compression of spinal cord 3) Early onset Alzheimer's Disease in 5th decade.

P16. A 24 year old female alcoholic gives birth to a female at 35 weeks gestation. The child is noted to have microcephaly, a smooth philthrum, a thin upper lip, and microophthalmia.

What is it? Fetal Alcohol Syndrome

P17. A 2 year old male presents for evaluation of language delay. The child has minimal use of words. On exam, the child avoids eye contact and does not exhibit a social smile. You note hand flapping, and he appears preoccupied with the clasp on his mom's purse.

What is it? Autism.

P18. A 7 year old male presents for abnormal behavior. His mother states that he has not made many friends, in part due to his "obsession with trains". She states he rarely engages conversation unless it is about trains. She has attempted to sign him up for sports or take him to group activities, but he will throw tantrums until she allows him to go back to his regular routine. He has performed well in school and showed normal language development.

What is it? Asperger's syndrome. It is an autism spectrum disorder with impaired social development but normal language and cognitive abilities, otherwise.

Neuropharmacology
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PH1. A 25 year old male undergoes an appendectomy at 6am. Approximately one hour into the surgery, the surgeons complain to the anesthesiologist that the patient is "tensing up" and is tachycardic. The patient's temperature spikes to 106F in the PACU.

What is it? Malignant hyperthermia.
Next step? Administer dantrolene.
What lab abnormalities can be present? Combined respiratory and metabolic acidosis, increased serum CPK and potassium, and urine myoglobin.

PH2. A 22 year old male is started on haloperidol for treatment of schizophrenia. One week later he presents to the ED with a "fever" and confusion. On exam, you note a temperature of 41C (106F), BP 200/110, HR 120. He is diaphoretic with diffuse muscle rigidity and tremor. Lab results reveal increased CPK and urine myoglobin.

What is it? Neuroleptic Malignant Syndrome.
Next step? Cooling measures and IV hydration with alkalinization to prevent renal failure.
Treatment? Cessation of haloperidol. Bromocriptine or dantrolene are classic medication treatment answers. ECT for refractory cases could be considered.

PH3. A 16-year-old female with depression treated with citalopram presents to the ER nonresponsive after taking an overdose of her mother's phenelzine tablets. On exam, her temperature is 103F, pulse 150, BP 180/110. She has rigid, tremulous extremities with clonus at the ankles.

What is it? Serotonin Syndrome.
Next step? Cyproheptadine is a good answer (serotonin antagonist). Supportive care measures like cooling or benzodiazepine for muscle relaxation would be appropriate as well.

PH4. A 35 year old female with bipolar disorder type I presents for memory problems, tremor, and imbalance for the past 10 days. She has taken lithium for the past 10 years and hydrochlorothiazide, started two weeks ago for hypertension. On exam, she has a bilateral coarse tremor and ataxic gait.

What is it? Lithium toxicity.
Treatment? Temporarily cease taking lithium. Change to nondiuretic antihypertensive.

You're done. Give your brain a rest.

While I hope this was helpful, I realize it could be better.
You may send any feedback to taylorfinseth@gmail.com.
Thanks.

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** = very high yield, know all you can about it /// * = important /// No * = good to know

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