Chief Complaint: Weakness	
Guillain Barre[1]	
PowerPlans	N/A
Pathophysiology	Monophasic demyelinating neuropathy. Immune system attacks peripheral nerves. At least half of cases are preceded by viral infection (respiratory > GI illnesses). C jejuni enteritis is an infamous example
Presentation	Progressive motor weakness (ascending) & areflexia +/- autonomic dysfunction
Differential	Spinal cord lesion (transverse myelitis), acute flaccid myelitis, tick paralysis, toxic neuropathy
Red Flags	Weakness of muscles of respiration can indicate need for intubation.
Workup	CSF profile classically w/ albuminocytologic dissociation (elevated protein w/o leukocytosis). EMG is not helpful early in the disease course.
Treatment	IVIG or plasmapheresis; consult PT
Miller-Fisher variant of Guillain Barre[2]	
PowerPlans	N/A
Pathophysiology	Antibody-mediated (anti-Gq1b) demyelination of the cranial nerves w/ or w/o peripheral nerve involvement.
Presentation	Defined by the presence of areflexia, ophthalmoplegia and ataxia; viral illness usually precedes symptoms. Sensorium remains intact.
Differential	Guillain-Barre Syndrome, myasthenia gravis, spinal cord lesion, MS
Red Flags	Weakness of muscles of respiration can indicate need for intubation
Workup	MRI of the brain and spine; LP if no space-occupying lesion. CSF profile similar to that of GBS w/ albuminocytologic dissociation (elevated protein w/o leukocytosis)
Treatment	IVIG 2g/kg over 2-5 days
Multiple Sclerosis[3]	
PowerPlans	N/A
Pathophysiology	T lymphocytes attack oligodendrocytes à damaged axons (autoimmune-mediated demyelination); known genetic (HLA subtypes) and environmental (smoking, latitude, vit D) risk factors
Presentation	• Repeated episodes focal deficits (optic neuritis, weakness, numbness) separated in time. • Imaging often shows lesions separated by space w/i the CNS
Differential	ADEM (often a first presentation of MS- multiple lesions causing altered sensorium), NMO spectrum disorder (neuromyelitis optica), MOG-antibody associated demyelinating disease, malignancy, nutritional deficiency, leukodystrophy, mitochondrial disorder, CNS vasculitis
Red Flags	• Presentation is broad and variable • Seizure (indicating gray matter involvement), fever should lead you to rethink the diagnosis • Weakness of muscles of respiration and/or mental status changes can indicate need for intubation

Chief Complaint: Weakness	
Multiple Sclerosis[3]	
Workup	• Definitive diagnosis requires repeated episodes over time. • LP reveals CSF w/ elevated protein count +/- presence of oligoclonal bands (must be compared w/ serum); MRI is imaging modality of choice. • The presence of 3 or more white matter lesions on T2 imaging especially if perpendicular to the ventricles sensitive for diagnosis (Dawson's fingers)
Treatment	Acute exacerbations require short-course of steroids. Load w/ methylprednisolone (30 mg/kg; maximum 1 g) treat for 3-5 days. Neuroimmunology consultation for disease-modifying drugs.
Infantile Botulism[4]	
PowerPlans	N/A
Pathophysiology	• C. botulinum produces toxin that interferes w/ release of acetylcholine at NMJ (disrupts vesicle binding to the pre-synaptic membrane). • In infancy, C. botulinum colonizes intestinal tract in situ. • Contamination of honey or corn syrup, dusty environments near construction/agricultural soil disruption are culprits. • In adults, paralysis results from ingestion of the toxin.
Presentation	Descending paralysis: often starting w/ ophthalmoplegia (may involve pupillary response), followed by weak cry, dysphagia and progresses to weakness of respiratory muscles
Differential	GBS Miller Fisher variant, hypermagnesemia, SMA, Myasthenia Gravis
Red Flags	Weakness of muscles of respiration can indicate need for intubation
Workup	Isolation of organism in stool; EMG: short-duration, low-amplitude motor unit potentials
Management	• ICU care for severe presentation, may require ventilator support • Immune globulin • Avoid aminoglycosides (produce pre-synaptic neuromuscular blockage) • Treat w/ BIG prior to confirmation of stool/EMG if clinical suspicion is high
Complications	Apnea, respiratory failure, sudden infant death
Myasthenia Gravis[5]	
PowerPlans	None
Pathophysiology	Antibody blockade of the post-synaptic ACh receptor at the neuromuscular junction
Presentation	• Fatigable weakness (symptoms worse at the end of the day) • Diplopia and ptosis can be provoked by sustained upgaze, arm weakness can be provoked w/ repetitive arm pumps. • Weakness tends to present in the muscles of the face, causing dysphagia, dysphonia, drooling, dysarthria (bulbar symptoms) • **Myasthenic Crisis**: Presents w/ inability to clear secretions or maintain oxygenation (precipitated by infection, surgery, stress, meds, etc)
Differential	Botulism, Miller Fisher variant of GBS, brainstem lesion, thyroid ophthalmopathy
Red Flags	Check how high the patient can count in a single breath, NIFs, check sustained up-gaze; evaluate neck flexion/extension (sensitive test for diaphragmatic strength) to assess need for intubation

Weakness continued on next page →

Chief Complaint: Weakness

Myasthenia Gravis[5]

Workup	Ice pack for eval of ptosis (should improve as cold slows acetylcholinesterase activity; check for antibodies (anti-AChR, anti-MuSK), EMG: decrement in muscle potentials on repetitive nerve stim
Management	Avoidance of drugs which may exacerbate MG (see uptodate table). Monitor FVC/NIF and intubate for FVC < 15 mL/kg and NIF < -20. Suctioning, NG tube.
Treatment	See below: IVIG (0.4 g/kg/d x 5d), plasmapheresis if severe
Complications	Respiratory failure, death

Bell's Palsy

PowerPlans	Facial Palsy EBG
Pathophysiology	Acute paralysis of the peripheral facial nerve. Pathogenesis viral (most commonly HSV) but also may be post-viral or immune-mediated (VZV, Hepatitis, HIV, Lyme, EBV)
Presentation	Weakness in the upper and lower face**,** pain, tingling in ipsilateral ear canal, taste changes, impaired lacrimation and hypersensitivity to sound
Differential	Otitis media**,** trauma, tumor, TB, Ramsay Hunt Syndrome, Malignant Hypertension, Mastoiditis
Red Flags	HTN, other cranial neuropathies
Workup	Exclude other cause (i.e. HTN, trauma, active herpetic lesions c/w RHS), Lyme serologies
Management	• **Watchful waiting**: eye ointments/artificial tears to maintain hydration, eye patch or taping eyelid closed while sleeping, use of corticosteroids controversial (most kids have complete spontaneous recovery); valacyclovir/acyclovir if HSV suspected, doxycycline if Lyme is suspected May-November; consider MRI if other symptoms present . • **Empiric corticosteroids**: = Prednisone 2 mg/kg once daily x 5 days w/ 5-day taper (max 60 mg/ dose). Start w/i three days of symptom onset.
Complications	Corneal ulcers if absent blink reflex/incomplete closure of palpebral fissure

CNS Manifestations of Lyme Disease

PowerPlans	N/A
Pathophysiology	B. burgdorferi from animals via tick vector
Presentation	fatigue, malaise, headache, facial palsy, peripheral neuritis, meningitis
Differential	Aseptic meningitis
Workup	Clinical diagnosis; lumbar puncture (elevated opening pressure, lymphocytic pleocytosis), screening test serum antibodies; confirmatory testing w/ western blot

Stage	Treatment
Early localized	**Ages 8 and older**: Doxycycline 4mg/kg/day divided BID x14 d **All ages**: Amoxicillin 50 mg/kg/d divided TID x14 d
Early disseminated and late disease	Same as early but for 21-28 d Ceftriaxone 75-100mg/kg IV or IM daily for 14-28d OR Penicillin 300K units/kg IV given in divided doses q4hr 14-28d

Chief Complaint: Weakness	
CNS Manifestations of Lyme Disease	
Management	See previous
Complications	Complications of meningitis, facial palsy, peripheral neuritis
Stroke[6]	
PowerPlans	Please call a code stroke if symptom onset < 5 hours prior (x52170); Neuroscience ICP admit plan or Neuro stroke plan, See Neurology Card
Pathophysiology	Acute onset neurologic dysfunction due to impaired blood supply to the brain; ischemic or hemorrhagic
Presentation	Acute onset unilateral weakness or numbness, acute onset altered mental status, new-onset focal seizures
Differential	Todd's paralysis following focal seizure, hemiplegic migraine, venous sinus thrombosis
Red Flags	Risk factors include infection, pro-thrombotic state, leukocytosis and anemia Risk factors for arterial ischemic stroke include Sickle Cell Disease and Cardiac Disease Risk factors for venous stroke are IBD, auto-immune disorders, infections and dehydration
Workup	Brain MRI/MRA w/ stroke protocol (includes DWI/ADC, FLAIR, T2, T1, susceptibility sequences) +/- MRV. TTE look for cardiac causes, serum labs to look for coagulopathy, if newborn add metabolic studies
Management	ABC's! Head of bed flat; IVF at maintenance, target SBP 50-90th percentile for age. Maintain euglycemia and normothermia, treat seizures, consider PICU admission and neurosurgical consult
Complications	Malignant edema which may lead to herniation, hemorrhagic conversion **(consider STAT CT for change in exam)**

1. Jones, H. Guillain-Barre Syndrome: Perspectives w/ Infants and Children. Seminars in Pediatric Neurology June 2000.
2. Shahrizaila, N, and Yuki, N. Bickerstaff brainstem encephalitis and Fisher Syndrome: anti-GQ1B antibody syndrome. Journal of Neurology, Neurosurgery and Psychiatry 84(5). 2013.
3. Krupp et al. International Pediatric Multiple Sclerosis Study Group criteria for pediatric multiple sclerosis and immune-mediated central nervous system demyelinating disorders: revisions to the 2007 definitions. Multiple Sclerosis Journal. April 2013.
4. Thompson et al., Infant Botulism in the age of botulism immune globulin. Neurology. June 2005.
5. Peragallo, J. Pediatric Myasthenia Gravis. Seminars in Pediatric Neurology. May 2017.
6. Lehman, et al., Transient focal neurologic symptoms correspond to regional cerebral hypoperfusion by MRI: A stroke mimic in children. American Journal of Neuroradiology. July 2017.

Chief Complaint: Altered Mental Status	
Meningitis: Inflammation of the leptomeninges secondary to infection **Encephalitis**: Infection of brain parenchyma secondary to infection (altered mental status, focal neurologic deficits)	
Bacterial Meningitis	
PowerPlans	**Fever in infant < 30 days**
Pathophysiology	Bacterial infection of the meninges. Caused by hematogenous spread or direct spread from sinuses or mastoids
Presentation	• Fever, headache, vomiting, meningismus, seizures • **Kernig Sign**: Stretching of hamstring w/ knee extension + back pain • **Brudzinski Sign**: passive neck flexion, involuntary hip/knee flexion
Differential	Viral meningitis/encephalitis, brain abscess, increased ICP, neoplasm, ADEM

Altered Mental Status continued on next page →