Pediatric Neurology: What the heck do we do?

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Disclosures

Relevant Financial Relationship(s)

None

Off Label Usage

None

Learning Objectives

- Develop an understanding of what a Pediatric Neurologist does on a typical day
- Review the key findings of febrile seizures and benign seizures with gastroenteritis
- Appreciate the distinguishing features of seizures and nonepileptic spells
- Differentiate the difference between postconcussive syndrome and migraine exacerbated by head injury

A Little About Me

- Grew up in Petersburg, ND
- Undergraduate degree at UND
- Medical school at UND
- Pediatric Neurology residency at Mayo Clinic
- Here at Altru for a little over 1 year
  - Clinic and hospital coverage
  - Outreach
- Enjoy spending time with my family, music, and almost anything outdoors

Overview

- Perinatal acquired/congenital neurologic disorders
- Genetic, Metabolic, Neurocutaneous disorders
- Neurodevelopmental disorders
- Epilepsy
- Nonepileptic paroxysmal disorders
- Headaches
- Movement and balance disorders
- Brain injury and disorders of consciousness
- Hydrocephalus
- Infections of the nervous system
- Tumors of the brain and spine
- Cerebrovascular disease
- Neuromuscular disorders

Perinatal Acquired/Congenital Neurologic Disorders

- Neonatal seizures
- Hypoxic ischemic encephalopathy
- Neonatal brain injury
  - Stroke, CVST, birth trauma
- Intraventricular hemorrhage and other hemorrhages
- Perinatal metabolic encephalopathies
- Congenital structural defects
  - Forebrain
  - Spinal cord

Genetic, Metabolic, Neurocutaneous Disorders

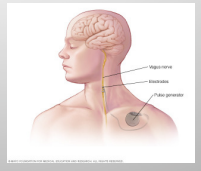
- Genetic syndromes
  - Down syndrome 1/1000
  - Fragile X syndrome 1/4000-7000
  - Prader-Willi and Angelman syndromes
- Inborn errors of metabolism 1/800-2500
- Neurocutaneous disorders
  - Neurofibromatosis 1/2600-3000
  - Tuberous sclerosis 1/5000-10000

Neurodevelopmental Disorders

- Developmental delay and Intellectual disability
  - Plateau and regression
  - Global and more specific
  - Etiology if possible
  - Multidisciplinary team
- ADHD
  - Common comorbidity
  - Tx: Tourette syndrome
- Autism spectrum disorders
  - No formal diagnostics
  - Autism diagnostic clinic

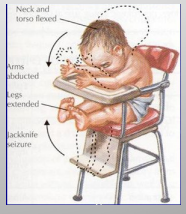
Epilepsy

- Diagnosis
  - Clinical exam (ex. hyperventilation)
  - EEG with video monitoring
- Treatment
  - Medication
  - Vagal nerve stimulator
  - Surgery (postop management)
- Other epilepsy evaluations and management
  - cEEG monitoring
  - PICU
  - Ketogenic diet



Infantile Spasms

- Onset 3-7 months, after 18 months rare
- Developmental plateau and regression
- EEG: Hypsarrhythmia
- Tx: ACTH and Vigabatrin



18 mo F with febrile seizures

- Hx global developmental delay since birth
- 3 seizures with fever requiring multiple seizure rescue medications, 2 of them lasting hours
- Often GTCS, may have focal jerking of the RUE at the start
- Family history of epilepsy

Simple vs. Complex Febrile Seizures

Simple	Complex
• 6 months to 6 years	• 6 months to 6 years
• GTCS, atonic, tonic	• Focal seizures
• < 15 minutes, self-resolves	• > 15 minutes, requires Tx
• Don't recur w/in 24 hrs	• Recurrence w/in 24 hrs
• Return to baseline quicker	• Longer postictal phase
• Normal development	• Developmental delay
• Family Hx febrile seizures	• Family Hx epilepsy

Febrile status epilepticus: > 30 minutes

Benign seizures with gastroenteritis?

Nonepileptic Paroxysmal Disorders

- Psychogenic nonepileptic spells
- Sleep-wake disorders
- Syncope/POTS
- Breath-holding spells
- Headaches and variants
- Benign shuddering attacks
- Inattention/staring spells
- GERD/Sandifer syndrome
- Benign myoclonus
- Tics
- Stereotypies
- Self-stimulation/soothing

Psychogenic Nonepileptic Spells

- Stress response
- No EEG correlate
- Are NOT under the control of the child
  - Not malingering, attention seeking, "faking it"
- Do not respond to anti-seizure medications

Epileptic events

- Stereotyped, rhythmic, synchronous
- Not distractible or suppressible
- Eyes open
- More often associated with injury
- More often associated with incontinence
- More likely to have post-ictal confusion
- Can occur during wakefulness or sleep

Non-epileptic behavioral event

- Arrhythmic, asynchronous shaking of limbs
- No-no head shaking
- Pelvic thrusting ("hips-arrhythmia")
- Intermittent responsiveness
- Forced eye closure
- Prolonged without post-ictal confusion
- Do not arise out of sleep

Sleep Disorders

- Parasomnias
  - Confusional arousals
  - Somnambulism
  - Sleep terrors
- Insomnia
  - Sleep hygiene
  - Medication
- Restless legs syndrome
  - Iron supplementation

Arousal parasomnias

Clinical Feature	Confusional arousal	Sleep terror	Somnambulism
Age of onset	2-10 years	2-10 years	5-10 years
Frequeny	1-2/mo to 3-4/wk	1-2/mo to 3-4/wk	1-2/mo to 3-4/wk
Peak onset	First 3rd of night (N3)	First 3rd of night (N3)	First 3rd of night (N3)
Behavior	Whimper, some articulation, sitting up in bed, inconsolable	Screaming, agitation, flushed face, sweating, inconsolable	Walking around, quiet or agitated, unresponsive to verbal commands
EEG	Rhythmic theta/delta activity	Rhythmic theta/delta activity	Rhythmic theta/delta activity
Duration	10-30 min	10-20 min	10-20 min

Syncope

- Neurocardiogenic (aka vasovagal)
  - Positional
  - Often triggered by stimulus (heat, blood, etc)
  - Lightheaded, visual blurriness, "blacks out" or "curtain falls", tunnel vision
  - Fall limply
  - Regain consciousness shortly after falling
  - May be disoriented after, but not confused
  - Can have convulsions

Orthostatic Hypotension and POTS

- Orthostatic hypotension (OH): drop in BP upon standing
- Postural (orthostatic) tachycardia syndrome (POTS): increase in HR upon standing
- Clinical features: lightheadedness, "weakness", blurred vision, and fatigue upon standing (POTS), syncope (OH)
- Dx: Orthostatic vitals, tilt-table test
- Treatment
  - Water, salt, positional tactics, exercise
  - Fludrocortisone

Breath-Holding Spells

- 6 months to 6 years
- Pathogenesis unclear
  - Autonomic nervous system dysfunction?
  - Iron deficiency anemia more prevalent
- Fam Hx in 20-35% patients
- Cyanotic and Pallid
- Dx: Clinical +/- EKG
- Tx: Education, iron supp

Headaches

- MIGRAINES > post-traumatic headaches > medication overuse headaches > tension-type headaches > TACs > others
- Diagnosis: clinical +/- MRI brain
- Treatment
  - Headache hygiene
  - Medication
  - Cefaly
  - Botox
  - Nerve blocks

Headaches: Red Flags

- Occipital location
- Nocturnal onset
- "Worst headache of my life", new HA, different HA, persistent/progressively worsening
- HA triggered by change in position, coughing/sneezing, bowel movement
- Systemic s/s (fever or other infectious s/s)
- Focal neurologic deficits, papilledema, ataxia, seizures

Postconcussion Syndrome vs. Migraine Exacerbated by Head Injury

- Postconcussion syndrome can present w/ headache, dizziness, cognitive impairment, and psychological symptoms
- Differentiating factors
  - Previous migraine headaches
  - Family history migraine
  - Headache is the predominant symptom
  - Improvement or resolution of other symptoms
- Return to play?

Movement and Balance Disorders

- Movement disorders
  - Tics and Tourette syndrome
  - Stereotypies
- Balance disorders
  - Vertigo
  - Ataxia

Tics and Tourette Syndrome

- Involuntary, but can be voluntarily suppressed
- Sudden, brief, repetitive movements or utterances
  - May need to exclude myoclonic seizures
- Stereotyped, but not rhythmic
  - Helps differentiate from focal seizures
- Build up sensation before tic, relieved once tic is performed
- Typically nearly daily, fluctuate, new tics
- Tx: Education/reassurance → CBT/HRT → medication

PANDAS: Fact or Fiction?

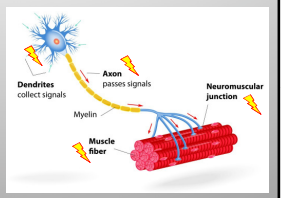
- Pediatric autoimmune neuropsychiatric disorder associated with group A streptococci (PANDAS): symptoms of obsessive-compulsive disorder (OCD) or tic disorders exacerbated by GAS infection
- High prevalence of GAS infections/carriers, tic disorders
- My treatment plan
  - Treat acute GAS infection
  - Treat OCD and tics
  - No chronic abx

Medical Management of:

- Hydrocephalus
- Brain injury and disorders of consciousness
  - Concussion, postconcussive syndrome, posttraumatic headaches
  - Determination of brain death
- Infections of the nervous system
  - Meningitis
  - Encephalitis
  - Abscesses
- Tumors of the brain and spine
- Cerebrovascular disease

Neuromuscular Disorders

- Motor neuron unit
  - Anterior horn cell
  - Peripheral nerve
  - Neuromuscular jxn
  - Muscle
- Diagnostics
  - EMG



Hypotonia

Central

- 70-80%
- Dysmorphic features
- Abnormalities of other brain function
- Other organ malformations
- Spasticity (fisting, scissoring)
- Normal to brisk DTRs

Peripheral

- 20-30%
- Facial muscle weakness/jaw underdeveloped
- Bone/joint deformities
- Muscle atrophy
- Fasciculations
- DTRs absent or depressed
  - DTRs > weakness → neuropathy
  - DTRs = weakness → myopathy

Infantile Botulism

Summary

- Actively seeing patients in the clinic and hospital
- Diagnosis through clinical exam, laboratory evaluation, imaging, EEG
- Treatment through healthy lifestyle/behavioral, medications, Botox injections, nerve blocks, VNS
- Part of a multidisciplinary team

Resources

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