Non-Epileptic Paroxysmal Events (NPE) in Children

Pediatric Board Review Course

Ajay Gupta, MD
Pediatric Epilepsy/Neurology
Cleveland Clinic Foundation
Cleveland, OH

Age/State Based Approach

- Neonates (0-8 wks)
  - Jittery (Ca, Gl, HIE)
  - Startle reflex
  - Benign sleep myoclonus

- Infants (2mth-2yr)
  - Breath holding spells
  - Gastroesophageal reflux
  - Shuddering attacks
  - Stereotypy (Mannerisms)
  - Rhythmic movement disorder (sleep)
  - Factitious disorder by proxy

- Childhood (2-12yr)
  - Staring /day dreaming
  - Tics/Movement disorder
  - Migraine equivalents
  - Masturbation
  - Drugs (Antiemetics)
  - Withholding behavior
  - Parasomnias (sleep)

- Adolescents (>12 yr)
  - Syncope/Cardiac
  - Psychogenic seizures

Pathophysiological Approach

- Cardiovascular
  - Breath holding spell
  - Syncope
  - Arrhythmias

- Movement Disorder
  - Tics
  - Dystonias
  - Choreaathetosis
  - Shuddering attacks
  - Stereotypy
  - Nonepileptic myoclonus

- Neurobehavioral
  - Migraine variants
  - Sleep/arousal disorders
  - Substance use disorder
  - Masturbation
  - Panic attacks
  - Psychogenic seizures
  - Metabolic disorders

- Gastrointestinal
  - Reflux
  - Withholding/Constipation
Why Board Loves NPE?

- These are common in practice
- Diagnosis is possible by history and exam alone
- Most kids will do well with early recognition and management
- Costly investigations are avoidable

Clinical Approach

- Read the Vignette carefully, and note:
  - Age of onset and course of events
  - Description, frequency, awake/sleep/both, stereotypic nature, triggering or relieving factors
  - Consider pathophysiologic mechanisms based on event description
- Epileptic & non-epileptic events may coexist

Cyanotic Breath Holding Spell

- 6 months – 2 years (up to 5 years)
- Typically confused with tonic seizure
- Always a trigger: fear, injury, frustration
- Cry → breath holding (expiration) → stiff, loss of awareness → clonic jerks
- Patho-physiology not understood
- Correction of anemia, counseling
- Pallid type (Reflex asystole)
  - parasympathetic dysregulation, pale and limp, with asystole
Benign Neonatal Sleep Myoclonus

- Rapid, random, bilateral/asynchronous jerking, may be forceful and rhythmic
- Seconds-minutes or even hours in sleep
- All stages of sleep (Quiet sleep/NREM)
- Differential: Seizures and Jitteriness
  - Disappear when infant is woken up
  - Not seen during alert wakefulness
  - Does not stop on passive flexion (jitter stops)
- EEG: Normal baseline and during events
- Mostly disappear by late infancy

Head Drops
(Benign Infantile Myoclonus)

- Infants with sudden head nods/drops
  - No fall or interruption of activity
  - No change in facial expression/behavior
  - No extremity movements
  - Momentary, quick recovery
- Confused with infantile spasms
- History and Exam are benign
- Development, and EEG normal
- Management: Reassure

Gastro-esophageal Reflux

- Dystonic, abnormal movements of head, neck, upper trunk (*Sandifer’s syndrome*)
- Life-threatening events – apnea with cyanosis and/or pallor
- Vomiting, failure to thrive
- More common in delayed/hypotonic patients
- Management:
  - Confirm diagnosis, treatment of reflux
Shuddering Attacks (stereotypy)

- Onset 6mth – 10 yrs, gradually better
- Sudden tremulous contraction (shiver)
  - Flexion of head and trunk
  - Adduction and flexion of elbows
- Brief, up to 100/day, cluster
- Intervening several weeks of no spells
- Benign phenomenon
- No treatment, gradually disappears
- Specific stereotypy/mannerism

Childhood Parasomnias

<table>
<thead>
<tr>
<th>Features</th>
<th>Nightmares</th>
<th>Night terrors</th>
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</thead>
<tbody>
<tr>
<td>Age of onset</td>
<td>2-5 years</td>
<td>4-8 years</td>
</tr>
<tr>
<td>Duration</td>
<td>&lt;1-2 minute</td>
<td>&gt;5 minutes</td>
</tr>
<tr>
<td>Semiology</td>
<td>Cling, verbalize</td>
<td>vary/autonomic</td>
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<tr>
<td>Stage sleep</td>
<td>REM</td>
<td>NREM III &amp; IV</td>
</tr>
<tr>
<td>Time</td>
<td>early am</td>
<td>first third of night</td>
</tr>
<tr>
<td>Recall</td>
<td>usually able</td>
<td>not able</td>
</tr>
</tbody>
</table>

Tics

- 1st decade, confused with myoclonic jerks
- Repetitive, Purposeless movements
  - Sudden, brief, stereotypic, wax/wane
  - Brief voluntary inhibition possible
  - Worsen with anxiety, Disappear in sleep
- Motor Tics
  - Simple: Face twitch, head shake, eye blink, sniff
  - Complex: Facial distortion, jump, fiddle, sway
- Vocal: cough, clear throat, words (profane)
- Common: Tourette syndrome
Treatment of Tourette

- Tics, hyperactive, aggressive behavior
  - Clonidine, Pimozide
  - Other dopamine antagonists
- Inattention
  - Methylphenidate, D-amphetamine, Atomoxetine
- Anxiety, depression, obsessive compulsive
  - SSRI, Bupropion
- Behavior counseling, child guidance

Childhood Masturbation

- Infants/young children, min to hours
- Episodes of genital and self stimulation
- Paroxysmal, Stereotypic, rhythmic
  - Tightening of thighs and rocking
  - Pressure to pubic/supra-pubic areas
  - Irregular breathing, flushing, diaphoresis
- Mimic complex partial seizures or pain
- Reassure and inform parents

Opsoclonus Myoclonus Syndrome

- Syndrome of myoclonic encephalopathy
  - Age: Infants and young children
  - Eye: Constant eye bobbing
  - Polymyoclonia of body and extremities
- Etiology:
  - Neuroblastoma (Para-neoplastic syndrome)
  - Post infectious (acute cerebellar ataxia)
- Management:
  - Investigate for neuroblastoma
  - Steroids for acute Rx, self-limiting, long term outcome not always favorable
Benign Paroxysmal Vertigo

- Benign condition, healthy toddlers
- Spells
  - Sudden, few minutes (uncommonly ~hour)
  - “As if disequilibrium”
  - Key: An alert child who is unable (refuses or frightened) to walk
  - 1-2/ week to 1 every 1-2 month
  - Stable course, improve by 6 years age
- Family history of migraines
- No Rx., reassurance
- Rare: Anti vertiginous agents

Oculogyric Crisis

- Acute dystonic reaction: Eyes, upper body or whole body
- Could be distressing
- Mechanism: Dopamine blockade
  - Metoclopramide, antipsychotic drugs, overdose of some antiepileptic drugs
- Rx: IV Diphenhydramine, IV Diazepam

Syncope (Convulsive Syncope)

- Transient global cerebral hypoperfusion
- Pre-syncopal symptoms (aura)
  - light head, nausea, visual blurring, distant hearing, vertiginous
- Syncope:
  - Fall/slump, pale, loss of awareness, stiff, clonic jerking (confused with GTCS)
- Three most common in children
  - Vasovagal (Neuro-cardiogenic)
  - Orthostatic (Drugs, autonomic dysfunction)
  - Cardiogenic (Arrhythmia)

Benke et al., Eur Neurol 1997;37:28
Treatment - Syncope

- Treat cause if found – cardiac, reduce drugs or stop (listed under dizziness)
- Conservative – neurocardiogenic syncope
  - Avoid precipitating factors, hydration, adequate salt intake, compression stockings
- Meds: Weak evidence in trials:
  - Midodrine – alpha-1 agonist, 10mg tid
  - Fludrocortisone (0.1mg/day to up to 1mg/d)
  - Pseudoephedrine, Paroxetine, DDAVP

Paroxysmal Kinesigenic Choreoathetosis

- 4-16 years, Duration: Secs. to few minutes
- Dystonic or Choreo-athetotic movements
  - Induced by activity or emotion
  - Involve extremities, face, head/neck, inability to speak or fall, Intact sensorium
- Idiopathic sporadic, symptomatic, familial
- Stable or improving course
- Treatment: If interferes with daily life
  - Low doses of Carbamazepine or phenytoin

Thanks