Non-Epileptic Paroxysmal Events (NPE) in Children

Pediatric Board Review Course

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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/myoclonias
- Movement disorder
- Migraine equivalents
- Masturbation
- Drugs (Antiemetics)
- Withholding behavior
- Parasomnias (sleep)

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

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

Baby Catnap

2 month-old, male

- Normal term delivery
- Diagnosed with seizures on 1st day of life
  - Random arm & leg jerks, only during sleep, many times daily
- Was feeding normally at birth, normal awake exam, gaining weight
- Discharged on Phenobarb and Phenytoin
- Brain CT, CBC, CMP, Ca, P normal
- Now sleepy and more jerky
- Came back in the ER

The appropriate next step in the management of this child would be..

1. IV load of Phenobarb (levels likely low)
2. Ask ER to intubate and admit to intensive care unit
3. Stat EEG
4. Attempt to wake the child up
5. Ask nurse to start a “jerk” count
Benign Neonatal Sleep Myoclonus

- Myoclonus (vs. Seizures vs. Jitteriness)
  - Disappear when infant is woken up
  - Not seen during alert wakefulness
  - Does not stop on passive flexion (jitter stops)
- Neonatal 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)
  - EEG: Normal baseline and during events
  - Mostly disappear by late infancy

Breath E Wright
3 month-old, girl

- Daily episodes in sleep and wakefulness
  - Upper body/arms stiffens or arches
  - May stop breathing, and turn blue and sometimes pale
  - Duration: few seconds
- Slow weight gain, normal vital signs, alert infant with normal exam
- Parents ask for apnea monitor and oxygen tank at home

A reasonable first step in this child’s plan of care would be:

1. pH probe study/ medication for GERD
2. Polysomnogram and video EEG
3. Reassure parents for breath holding spells
4. Echocardiogram
5. Prescribe apnea monitor and oxygen tank
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

Ben D Bow
8 month-old, boy

- Episodes of
  - “forward head drops (bows)” since 3 months
  - Several times a day, during wakefulness
  - No change in behavior
  - Appropriate milestones for age
- Normal pregnancy and perinatal period
- Normal development, Normal exam
- Outside 30 minutes EEG normal

The events are likely to be..

1. Infantile spasms (West syndrome)
2. Tics
3. Opsoclonus-myoclonus syndrome
4. Benign infantile myoclonus
5. Lucky parents: child is learning to say “yes”
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

Tre Amble
4 year-old, boy

- Developmental delay
- Spells since last 3 months
  - Behavior arrest
  - Body tensing, trembling, hands close to chest
  - Duration: few seconds
  - Occur during day time, any time, every day
- Normal exam, no new findings

The most likely diagnosis is ..

1. Tics
2. Rett syndrome
3. Dysautonomia (temperature instability)
4. Tonic seizures (Lennox Gastaut syndrome)
5. Shuddering attacks
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

Will Power
14 month-old, boy

- Normal development
- Spells since the age of 7 months
  - Loud cry or screams when upset, stiffens, stops breathing, turns blue for few seconds
  - Last two episodes 1-2 minutes, followed by generalized clonic convulsions
- Normal in between spells
- Normal physical exam

The appropriate next step in the management of this child would be..

1. Start phenobarb due to motor convulsions in the last 2 episodes
2. Obtain video EEG to record spell
3. Reassure and educate parents that the spells will eventually disappear
4. Cardiac (Holter) monitoring
5. Remove all irritants from his daily life
Cyanotic Breath Holding Spell

- 6 months – 2 years (up to 5 years)
- Typically confused with epileptic seizure
- Always a trigger: fear, injury, frustration
- Cry → breath holding (expiration) → stiff, loss of awareness → clonic jerks
- Breath holding → Tachycardia and brain hypoxia → rare clonus or a grand mal seizure (pathophysiology not understood)
- Counseling, iron deficiency anemia
- Pallid spell (Reflex asystole)
  - parasympathetic dysregulation, pale and limp, with asystole

Chris N Cross
4 year-old, female

- Developmental delay
- New spells:
  - Mother finds her on floor laying on her belly, stiff or crossed legs, thrashing as if in pain
  - Few seconds to minutes duration, at times in clusters, almost daily
- Normal exam

The most likely possibility is ..

1. Complex partial seizures
2. Masturbation
3. Restless leg syndrome
4. Lactose intolerance
5. Renal colic
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

Stan D Still

13 months-old, boy

- Two weeks history of
  - Horizontal bobbing of eyes constantly
  - Body twitches, shakes and unable to stand or sit
  - Agitation, irritability, fretful behavior
- Previous normal history
- Up to date immunization
- Your exam reveal no other findings

The diagnostic test likely to confirm his diagnosis is..

1. Urine catecholamines
2. EEG and Observation in the hospital
3. Cerebrospinal fluid testing
4. Brain MRI with gadolinium
5. Dilated eye exam
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

Solo Tsunami

8 year-old, boy

- Spells began 2 years ago
  - Head, shoulder, arm, body and face jerks
  - Chronic dry cough
  - On bad days, touches and taps class mates around him, loud utterances in class room, show physical aggression
  - Stop for seconds when reprimanded
  - Worse when stressed, better in sleep
  - Waxing and waning course

- Academically struggling

Following statement is true about this child..

- 1. Anxiety and obsessive compulsive disorder is likely
- 2. EEG is likely to be abnormal
- 3. One parent has similar illness
- 4. Brain MRI will likely show basal ganglia hyperintensities
- 5. This is ‘Saint Vitus Dance’ disease
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

The treatment of tics in Tourette syndrome is indicated..

- 1. At the time of diagnosis to prevent progression
- 2. If tics occur > 6 hours/day
- 3. If tics persist for > 2 years
- 4. If tics affect child’s social, occupational, and academic life
- 5. All of the above

Tourette syndrome is commonly associated with all of the following except...

1. Attention deficit hyperactivity disorder
2. Anxiety disorder
3. Obsessive compulsive disorder
4. Oppositional defiant disorder
5. Seizures
First Line Treatment of Tourette

- Tics, hyperactive, aggressive behavior
  - Clonidine, Pimozide, Guanfacine, other dopamine antagonists
- Inattention
  - Methylphenidate, D-amphetamine
  - Attention to tics for worsening
- Anxiety, depression, obsessive compulsive
  - SSRI, Bupropion
- Behavior counseling, child guidance

Dugin Knight, 6 year-old, male

- Spells since 5 years of age
  - Out of sleep, usually an hour after going to bed, occur 1-2 nights per month
  - Screams, restless, sweats profusely, flushed, heart racing, pupils big (per mom), inconsolable, 5-8 minutes, last one mom timed at 11 minutes
  - Goes to bed, not recall the event next day
- Healthy child, no other concerns
- Normal exam

The most likely diagnosis is ..

1. Frontal lobe seizures
2. Nightmares
3. Panic attacks
4. Acute confusional migraines
5. Night terrors (Pavor nocturnus)
### Childhood Parasomnias

<table>
<thead>
<tr>
<th>Features</th>
<th>Nightmares</th>
<th>Night terrors</th>
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<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>
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<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>

### Bob N Reel

3 year-old, female

- Spells since last 3 months
  - During wakefulness, 1-3 per week,
  - Frightened look, guards her head/neck motion, holds parents or objects, eyes appear to bob around, freezes without motion for ~ 5 minutes
  - May fall or lay down if standing, remains alert
  - Recovers quickly to play again
- Healthy child, no other concerns
- Normal exam

### The most likely diagnosis is ..

1. Basilar migraines
2. Benign paroxysmal vertigo
3. Transient ischemic attacks (Vertebro-basilar)
4. Vertiginous seizures
5. Spasmus nutans
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

Rip U Torn
10 year-old, boy

- Has autism and intellectual disability. Takes Ritalin, Clonidine, and Risperidone
- Taken to ER for violent aggression in school. Given I/M Haloperidol with good response. Home medications were increased at discharge on same day.
- Later at home, developed episodes of upper body stiffness, neck extension with up-rolling of eyes.
- Exam shows alert, but restless and distressed, unable to control episodes. Afebrile, normal vitals, and no other acute systemic findings

The most appropriate next step in the management of this child is ..

1. Intravenous 20ml/kg saline bolus
2. Lumbar Spinal fluid examination
3. Stat EEG
4. Intravenous Diphenhydramine
5. Intravenous Phenytoin
Oculogyric Crisis
Acute Dystonic Reaction

- 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

Kneil Down
16 year-old, male

- Spells since 11 years of age
  - Nausea, dizzy, “sounds” appear distant, collapses to ground, stiffens, unconscious, few body jerks. Duration 30-60 secs
  - Common after pain, shots, head hang when playing soccer
  - Takes Carbamazepine for seizures but not improved
  - No other medical or health issue
  - Normal exam, no exercise intolerance

The most likely diagnosis is ..

1. Convulsive neuro-cardiogenic syncope
2. Cardiac arrhythmia
3. Grand mal seizures
4. Complex partial seizures with auditory auras
5. Pseudo–seizures (non-epileptic seizures)
Neurocardiogenic 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)
- Other causes of syncope in children
  - Orthostatic (Drugs, autonomic dysfunction)
  - Cardiogenic (Arrhythmia)
- May be confused with seizures

Treatment

- Avoid triggers when possible
- Hydration, adequate salt intake, compression stockings
- Physical counterpressure (isometric activity)
  - Leg-crossing with simultaneous tensing of leg, abdominal, and buttock muscles
  - Handgrip on a rubber ball or similar object
  - Arm tensing gripping one hand with the other while simultaneously abducting both arms
  - May not avoid but delay syncope enough to lay supine
- Medications are rarely necessary
  - Midodrine – alpha-1 agonist
  - Fludrocortisone (0.1mg/day to up to 1mg/d)
  - Pseudoephedrine, Paroxetine, Beta blockers, DDAVP

Thank You