Epileptic vs. Non-epileptic Paroxysmal Events

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• No disclosures
Definitions:

• **Seizure**
  • abnormal excessive or synchronous neuronal activity in the brain

• **Epilepsy**
  • a condition characterized by the tendency for recurrent unprovoked seizure
Nonepileptic Paroxysmal Disorder

• 25% in monitoring units have no epilepsy

• Important for clinicians to determine if these events are truly epileptic
Non-epileptic paroxysmal events

- Common in the pediatric population
  - 20-25% of normal children presenting with ? seizure
  - Children with MR ? Epilepsy – 60% NEE

- Multiple etiologies:
  - other neurological d/o
  - other system d/o
  - sleep-related
  - behavioral / psychiatric

- Importance of diagnosis

- Age – specific differential
Is this an epileptic seizure?

• Strategic approach -
  • age based
  • key elements of event: “before, during, after”
    • abnormal movements, impairment of awareness,
    • autonomic or respiratory changes
    • precipitating & relieving factors
    • home video clip very useful

• Challenges in children with Intellectual disability, autism and severe cerebral palsy
Non-epileptic paroxysmal events

**Diagnosis:**
- Suspicion
- History
- Observation
  - direct
  - home-video
  - inpatient monitoring with EEG
- Induction
  - hyperventilation
  - photic stimulation
  - rotation
  - postural change
  - ? other
PNE in neonates

• Seizure semiology in neonates different –
  • no GTCS,
  • can be subtle (eye blinking, repetitive sucking/lip smacking, pedaling, clonic/tonic movements, eye deviation, autonomic spells, irritability, apneic spell) - Scher et al, 1989; Kramer, 1999

• Difficult to assess level of consciousness, aura or postictal manifestations - Hamer et al, 1999

• PNEs include jitteriness, benign neonatal sleep myoclonus, stiff baby/hyperekplexia

• Value of ictal EEGs –
  • majority (90%) PNE
  • focal clonic movements best correlation with seizures (44%)
  • jitteriness no correlation
  • apnea rarely sole manifestation, especially with bradycardia without motor component

Scher et al, 1989
PNE in infancy (2mths - 2 yrs)

Abnormal movements

- Myoclonus - benign myoclonus of infancy, benign sleep myoclonus, post CNS/spinal cord injury
- Tonic – hyperekplexia, withholding/constipation, shuddering attacks, spasmus mutans
- Dystonia – extrapyramidal drug reaction, Sandifer syndrome, paroxysmal torticollis
- Weakness – Alternating hemiplegia of childhood
- Stereotypies - head banging, rocking

Impaired awareness, autonomic or respiratory disturbances

- Masturbation
- Benign paroxysmal vertigo
- Breath holding attacks (cyanotic vs pallid)
Cyanotic
- 6 mths- 3yrs
- Precipitated by trauma, fear, anger
- Crying with prolonged breath holding in expiration
- Cyanosis, becomes limp ± stiffening or jerk (anoxic seizure)
- May sleep after attack
- Family history 30%
- Treatment is reassurance

Pallid
- Neonates & infants (infantile syncope)
- No precipitant ? Noxious stimuli
- Apnoea, pallor, bradycardia (vagal) ± seizure (anoxic-ischemic)
- EEG may show hypersynchronous slowing during cardiac asystole
- May need to investigate for underlying cause
**Non-epileptic event?**

- A 7-month-old boy became difficult to arouse, was limp and had blue extremities.
- He caught up on poliovirus, diphtheria-tetanus toxoids-acellular pertussis, Haemophilus influenzae type b-hepatitis B virus and pneumococcal vaccines.
- The hypotonic-hyloresponsive episode had resolved by the time the infant was seen in an emergency department 1 hour later.
Hypotonic-hyporesponsive episode

• The report describes hypotonic-hyporesponsive episode, encourages reporting of vaccine-associated adverse events and discusses prognosis and implications for subsequent immunization.

Abnormal movements

- Movement disorders
  - tics
  - paroxysmal dyskinesias (PKC/PNKC, PDC, PT)
  - drug induced reactions
  - other dystonias (DRD, ITD)
- Tonic - hyperekplexia
  - myotonia
- Atonia - periodic paralysis
  - AHC, cataplexy
- Sleep disorders/parasomnias
  - nightmares
  - sleep walking/”sit ups”
  - night terrors
- Others- Munchausen by proxy

Impaired awareness, autonomic or respiratory disturbances

- Syncope
- Migraine equivalents (BPV, cyclical vomiting, abdominal migraine)
- Staring spells

PNE in children (2-12 yrs)
Benign paroxysmal vertigo

- Attacks seen at 2-5 yrs of age
- Brief attacks (5-30 secs) – single episode or recurrent over days, well in between
- Appears frightened, pale, clutches parent
- + nystagmus, no LOC
- Quick immediate recovery
- Older children may be able to describe vertigo
- May develop migraine at later age
Staring spells

- Need to be differentiated from absence /complex partial seizures; difficult even when the child has neurologic abnormalities & epilepsy

  - noticed by teacher rather than parent
  - no family history of epilepsy
  - lack of motor movements (eye fluttering, lip smacking, hand movements)
  - responsive to touch, lack of interruption of play, can be “aborted” by a firm command
  - school difficulties

- Interictal EEG helpful (hyperventilation and sleep activation of epileptiform discharges) but not confirmatory

- “Gold standard” is ictal video EEG; occasionally 24hr ambulatory EEG necessary
Syncope

- Occurs in specific environments (prolonged standing, sudden change in posture, heat exposure, hunger, dehydration, emotional or painful circumstances)
- Prodrome of lightheadedness, blurring of vision, nausea, pallor, sweating before “blackout”
- No oromotor manifestations (10% have convulsions - brief duration, myoclonic, tonic, tonic-clonic)
- “Warning bells” & recurrent unprovoked episodes warrant cardiac + autonomic evaluation (tilt test – drop in blood pressure & hear rate)
- Treatment is education & reassurance, sufficient fluid & salt intake, simple maneuvers to abort acute episode (supine, crossing legs/folding arms, tensing thigh & abdominal musculature), deconditioning, rarely need medication (fludrocortisone, alpha-agonists)
**PNE in adolescents (12 – 18 yrs)**

Other neurological:  
- Migraine  
- Movement disorders  
- Transient global amnesia

Other system:  
- Syncope, arrhythmia

Sleep-related:  
- Narcolepsy, sleep jerks,  
- Sleep walking, REM sleep

Behav/Psych:  
- Panic attacks, psychogenic seizures  
- Intoxications
**Pseudoseizures**

- Almost half of PNEs in later childhood & adolescence are pseudoseizures

- Female preponderance (70%) in those >12 yrs

- Underlying psychiatric diagnosis & psychosocial stress (“family in distress”) – *Wylie 1999; Palkanis 2000*

- Early diagnosis important (institute psychotherapy, withdraw AEDs, helps family comprehend problem)

- 9-26% abnormalities on interictal EEGs – *Holmes 1980*

- Early use of video EEG monitoring useful
Non-epileptic paroxysmal events

- Psychogenic seizures:
  Suspect when
  - ‘seizures’ refractory to medications
  - atypical clinical features
  - stress provokes/exacerbates
  - apparent ‘gain’

Early video-EEG evaluation
  - co-existent epilepsy
  - prevention of inappropriate medication,
    especially prevent treatment for status epilepticus
  - prevention of misdiagnosis as psychogenic
Non-epileptic event?

• 4 year old boy, presenting with increasing frequency of paroxysmal behaviors present for 2 years.

• Sudden hands-to-face movement, always same pattern

• Questionable interruption of attention with spell

• Frequency increased from occasional, to now sometimes multiple per hour

• Normal exam and neuroimaging

• EEG: bilateral occipital sharp waves
Non-epileptic event?

• 16 yo boy with new onset seizures

• Found unresponsive, or in generalized seizure on increasing number of occasions

• Onset never witnessed, so far had occurred only at home

• Normal exam, EEG, neuroimaging

• Failure to respond to Valproic acid at high doses
Non-epileptic event?

- 16 yo boy

- A single febrile seizure at 1 y

- 4 month history of paroxysmal nervousness, sweaty palms, chest sensation, L hand and then bilateral arm and ? Leg movements. Stop with reassurance from mother. Full recall. No post-ictal confusion.

- Neurology consultation – non-epileptic

- Psychiatry consultation – Anxiety disorder, treatment with hydroxyzine (paradoxical reaction to bzps in past)

- No change in events – represent to ED for 4th time
**Associated psychiatric disorders**

- No single entity

- Somatoform disorders
  - Conversion disorder
  - Somatization disorder

- Dissociative disorders?

- Factitious disorder - rare
- Malingering - rare
Psychiatric comorbidities

• Depression
  • not primarily a cause of PNES
  • May release psychopathologic behaviors including conversion

• Anxiety
  • Panic attacks / agoraphobia
  • PTSD

• Psychosis
Clinical approach

• **Pre-diagnosis:** Trust building, engage with family in diagnostic effort, discuss full range of differential diagnosis and approach to investigation, explore family theories both organic, non-organic. Ideally include psychiatric consultation as part of the investigation approach.

• **Post diagnosis:** Commit to ongoing support and follow up, discuss the subconscious process and lack of intent/malingering, relate to other somatic manifestations of emotional stress/nervousness etc., anticipate good outcome, refer for psychiatric consultation if not already in process.
Outcome

• Adults – disappointing.
  • Multiple studies ~ 1/3 of patients free of PNES 1-5 years after diagnosis.
  • Comorbid depression common

• Children – better outcome
  • 70-80% free of events at 1-3 years
  • Comorbid anxiety more common than depression

• Poor prognostic indicators:
  • Long duration PNES
  • Comorbid psychiatric disorder.
Many types - self stimulation, hyperventilation, staring spells, spasticity, clonus, dyskinesia, eye and head deviation, intermittent & inconsolable crying

– Desai 1992; Donat 1990; Holmes 1983

Very challenging – children unable to report symptoms, increased incidence of epilepsy & medical conditions that cause PNE

80-100% have abnormal interictal EEG

- Paolichi 2002; Donat 1990; Holmes 1983

Importance of early video EEG monitoring so that PNEs may be further evaluated and managed