Differentiate between epileptic and non-epileptic events in children, but referring when the diagnosis is unclear or atypical
Recognize common patterns of non-epileptic events in children
Disclosures

- No financial disclosures
- Video credits:
  - Phillip Pearl, M.D.; Children's National Medical Center
  - www.medlink.com
  - www.youtube.com
  - Personal patients
    - Children's National Medical Center
    - San Antonio Military Medical Center

Definitions

- Paroxysmal = Sudden onset of a symptom
- Epileptic seizure = Abnormal excessive neuronal discharges with resultant symptoms (motor, sensory, alteration in mental status)
- If not epileptic:
  - Non-epileptic seizures
  - Spells
  - Movement disorders
Epileptic?

- Pattern / Exceptions
- Triggered/ Pre-event:
- Timing/rapidity of onset, length:
- Movements:
- Eyes:
- Mental status:
- Post-event:
- Other:
- EEG
  - Frequent → ictal
  - Non-frequent: Clinical

Epileptic?

- Triggered: usually not
- Pre-event: can have an aura
- Timing/rapidity of symptom onset: sudden, last seconds to minutes
- Movements:
  - Tonic
  - Clonic
  - T-C
  - Myoclonic
  - Atonic
- Eyes: usually open, deviated or nystagmus
- Mental status:
  - Partial vs. Complex partial
  - Generalized
- Post-event: Sleepy or confused
- Other: Incontinence, tongue biting

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Paroxysmal event

Non-epileptic

- Age
  - Neonates, 1-2 months
  - Infancy, <1-2 years
  - Childhood, 2 years to adolescence
  - Adolescence
- Sleep state
  - Awake
  - Asleep

Epileptic

Age?

Asleep or awake?

Good clinical history? (Pattern recognition)

Non-epileptic

Age: Neonates, <2 months

- Awake:
  - Jitteriness
  - Clonus
  - Startle/ Moro
  - GERD/Sandifer's
  - Hyperekplexia
  - Congenital nystagmus
  - Dyskinesias (from Bronchopulmonary dysplasia)
- Asleep:
  - Benign sleep myoclonus
### Age: Infancy, < 1-2 years

**Awake:**
- GERD related – Sandifer's
- Breath holding spells
- Benign paroxysmal vertigo
- Benign paroxysmal torticollis
- Shuddering spells
- Infantile gratification
- Spasmus nutans
- Paroxysmal dystonia

**Asleep:**
- Hyponic jerks

### Age: Childhood, 2 - 10 years

**Awake:**
- Breath holding spells
- Benign paroxysmal vertigo
- Paroxysmal dyskinesia
- Stereotypies
- Hyperventilation syndrome
- Narcolepsy (cataplexy)
- Staring – daydreaming
- Tic disorders
- Opsoclonus-myoclonus

**Asleep:**
- Head banging
- Night terrors
- Nightmares
- Sleep walking
- Confusional arousals
- Hypnic jerks

### Age: Adolescents

**Awake:**
- Confusional/basilar migraines
- Hyperventilation syndrome
- Narcolepsy
- Tic disorders
- Pseudoseizures
- Conversion Disorder

**Asleep:**
- Confusional arousals
- Sleep walking
- Hypnic jerks
- Restless legs syndrome
Awake
- Jitteriness
- Clonus
- Startle/Moro
- GERD/Sandifer’s
- Hyperekplexia
- Dyskinesias
- Younger
- Breath holding spells
- Benign paroxysmal vertigo
- Benign paroxysmal torticollis
- Shuddering spells
- Infantile gratification
- Spasmus nutans
- Startle disease
- Stereotypies
- Paroxysmal dyskinesias
- Opsoclonus/myoclonus
- Confusional/basilar migraines
- Hyperventilation syndrome
- Pseudoseizures
- Narcolepsy
- Tics
- Older

Asleep
- Head banging
- Night terrors
- Nightmares
- Sleepwalking
- Confusional arousals
- Hypnic jerks
- Younger
- Confusional arousals
- Sleepwalking
- Hypnic jerks
- Restless legs
- Older

History...
- Awake:
- Asleep:
  - Sleep transition? Within hours of sleep onset or later in night? Awaken from sleep? Can it be stopped? Videotape! Pattern!
Cases

- 13 Non-epileptic cases: Videos/History
  - Each age range
  - Some awake
  - Some asleep
  - Common, classic, important
- 2 Non-epileptic case "mimics"
  - i.e. epileptic

Age: Neonates, <2 months

- Awake:
  - Jitteriness
  - Clonus
  - Startle/Moro
  - GERD/Sandifer's
  - Hyperekplexia
  - Congenital nystagmus
  - Dyskinesias (from Bronchopulmonary dysplasia)
- Asleep:
  - Benign sleep myoclonus

Case

- Newborn with jerky arm movements while sleeping...
- Term
- No complications

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Case: Benign nocturnal myoclonus (sleep myoclonus)

- Seen especially in newborns, but seen some in all ages
- Term
- Early stages of sleep or sleep transition
- Especially: arms, fingers, wrists, elbows > legs
- Brief clusters, but can be repetitive and prolonged > 30 minutes
- Resolves by 3-4 months

Evaluation:
- Restraint: +/-
- Arousal – wake the child up!
- EEG: atypical cases

Treatment:
- Reassurance
- AED’s: NO - phenobarbital may decrease!

Prognosis:
- Tend to resolve over time
- Normal development

Case

- Newborn who seems stiff at baseline who is having paroxysmal episodes of tonic stiffening, especially when startled

*Courtesy of Phillip Pearl, M.D., Children’s National Medical Center*
Case: Hyperekplexia

- AKA “Stiff-baby syndrome”
- Abnormal response to auditory, sensory, visual stimuli:
  - Exaggerated startle
  - Generalized muscle rigidity
    - (*can be mistaken for tonic seizure*)
- At baseline neonates can still seem stiff and rigid
  - Clenched fists and anxious stare
  - Nocturnal myclonus and apnea

Testing:
- Bedside: Tapping of the nose or glabella
- Neurophys: EMG shows almost continuous activity
- Genetics: mutations in the glycine receptor

Treatment:
- Clonazepam/ GABA receptor agonists
- Forced flexion of the head and legs toward the trunk

Rare, but important:
- Apnea and sudden death from laryngospasm
- Confused as seizures
- Do not respond as well to phenobarbital

Age: Infancy, < 1-2 years

- Awake:
  - GERD related – Sandifer’s
  - Breath holding spells
  - Benign paroxysmal vertigo
  - Benign paroxysmal torticollis
  - Shuddering spells
  - Infantile gratification
  - Spasmsus nutans
  - Paroxysmal dystonia

- Asleep:
  - Hypnic jerks

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Case

- 16 month old who passes out when she gets upset...
- Usually awakens right away and cries

Case: Breath-holding spells

- Reflexive autonomic response: 5-27% infants
- 75% begin between 6–18 months, some as neonates
- Initial increase in frequency, peaks at 12-18 months
- Then declines, stops by 4-5 years
- FHx: 25% parents, 25% siblings (dominant)
- 2 types:
  - Cyanotic
  - Pallid

Cyanotic:

- 3x more common
- Crying... provoked by fear, frustration, etc.
- Breathing stops in expiration → Cyanosis →
  Limpness → Loss of consciousness →
  Awakens spontaneously after a few seconds
  → Awakens crying/ Seems normal
- May have some stiffening/ posturing/ twitches/ eye rolling with LOC

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Pallid:

- Provoked by sudden, unexpected stimuli, especially painful ones
- Provoking factor → Pale and limp → Loss of consciousness
- Lasts seconds, then patient awakens, may be normal or sleepy → normal after awakening
- May have some stiffening/ posturing/twitches/ eye rolling with LOC

Case: Breath-holding spells

Evaluation:
- CBC: association with iron-deficiency anemia
  - 80% vs 20% reduction at 3 months
- EKG: r/o long QT
- EEG/ Monitoring?: atypical cases only
- Ocular compression: not recommended

Treatment...


Treatment:
- Do not pick up child and hold upright
- Reassurance
- Consider iron supplementation
  - 91 children, 2/3 iron deficiency anemia
  - Iron supplementation 84% decrease vs 21%
- Cardiac pacemaker? Rare circumstances
- AED’s: no; Piracetam? Not available in U.S.

Prognosis:
- "Does anyone know an adult who has breath holding spells?" —Fenichel
8 month old with intermittent spells of “shivering”…
Normal otherwise

Case: Shuddering spells

- Spells of rapid shivering of the head, shoulder, and occasionally the trunk
- Usually start in infancy, around 4-6 months
- Can be provoked by fear, anger, frustration, with feeding
- Last few seconds, up to dozens/ day
- No change in consciousness

Evaluation:
- Consider VEEG - normal

Treatment:
- Reassurance
- Propanolol reportedly effective

Prognosis:
- Resolve spontaneously by a few years
- Some reports of subsequent essential tremor
Age: Childhood, 2 -10 years

- **Awake:**
  - Breath holding spells
  - Benign paroxysmal vertigo
  - Paroxysmal dyskinesia
  - Stereotypies
  - Hyperventilation syndrome
  - Narcolepsy (cataplexy)
  - Staring – daydreaming
  - Tic disorders
  - Opsoclonus-myoclonus

- **Asleep:**
  - Head banging
  - Night terrors
  - Nightmares
  - Sleep walking
  - Confusional arousals
  - Hypnic jerks

Case

- 4 y/o with inconsolable screaming within a couple hours of falling asleep, unconsolable but eventually falls asleep again but has no memory the next morning

Case: Night terror

- Onset usually between 3-6 years
- May have intercurrent illness in some
- Partial arousal from slow wave sleep: usually beginning of the night
- Child seems awake but is inconsolable, frightened, cries, thrashes around
- ½ sleepwalkers
- Autonomic sx: diaphoresis, tachycardia, tachypnea, flushing, increased muscle tone
- Eventually falls asleep again, (few minutes)
- No memory of event
Case: Night terror

- **Evaluation:**
  - Evaluate for illness or stressors
  - If stereotyped movements every time, consider EEG or PSG
  - Gentle restraint/ safety during episodes

- **Treatment:**
  - Reassurance
  - ½ will outgrow by 8 years
  - May consider benzo, ½ Diazepam mg or Clonazepam 0.5mg
  - Clonazepam, suppress slow wave sleep
  - Clonidine 0.05mg
  - Scheduled awakening

Case

- 3 year old with episodes of stiffening only during feeds

Case: GERD related movements

- Wide range of symptoms: Apnea, Irritability, Dystonic movements, Tonic-seizure like movements, Opisthotonic movements, Torticollis
- Pain, laryngospasm
- Sandifer: hiatal hernia + GER + dystonic neck posturing
- Common in patients with chronic encephalopathies with severe psychomotor retardation, up to 1/3
- Key to diagnosis: history
**Case: GERD related movements**

- **Evaluation**
  - pH probe monitoring
  - Barium swallow for anatomy
  - Milk scan
  - Video EEG non-epileptic
- **Treatment:**
  - Positioning
  - Thickening of feeds
  - Medications
  - Surgical

**Non-epileptic mimics (i.e. epileptic)**

- When cases are atypical...
  - Referral
  - Long term EEG monitoring

**Case**

- 6 month old with jerks mostly when falling asleep or when just waking up...
Case: Infantile spasms (*epileptic)

- Peak onset 4-7 months
- **Clusters**
- **Sleep transition**
- Flexor > extensor spasms, brief – few seconds
- EEG:
  - Hypsarrhythmia: interictal
  - Electrodecrement: ictal
- 2/3 Symptomatic: poor prognosis
- 1/3 Asymptomatic: 40% neurological normal outcome

Case: Infantile spasms

8 y/o boy is sent home from school because of strange episodes of swinging his arms around wildly...

Occurring during day and during sleep

Case: Frontal lobe seizure (*epileptic)

- Exception: bilateral motor involvement with preservation of consciousness: unilateral supplementary motor area, usually on the right
- Some are unusual nocturnal seizures: posturing, vocalizations
- Pseudoseizures/Conversion or Factitious: diagnosis of exclusion
  - Rare under 5 years
  - 12-18 years, females > males

Cases: Migraine variants

- 15 month old who on occasion will suddenly look frightened and grab onto her parent’s legs....
- Healthy otherwise
- Mother has migraine headaches

Case: Benign paroxysmal vertigo

- Early childhood, 1-7 years
- Sudden onset of vertigo with sudden stereotyped appearance of fright with maybe pallor and nystagmus without alteration in consciousness
- Child may either hold onto parent’s leg, or lie on the floor
- Rare vomiting
- Last minutes, rapid and complete recovery
- Irregular intervals, daily to every couple months

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Case: Benign paroxysmal vertigo

- Migraine?:
  - 70% FHX migraine
  - 10% coexisting typical migraine
  - 21% eventually develop migraines

- Evaluation:
  - Consider...

- Treatment:
  - Reassurance
  - Decrease and eventually stop in older childhood
  - Meclizine? Periactin?

Case

- 6 month old with episodes of head turning, lasting minutes to hours, before resolving spontaneously

Case: Benign paroxysmal torticollis

- Onset 2-8 months
- Intermittent neck twisting, abnormal head positioning; occasional trunk posturing
- Side can vary
- May have pre-event irritability, distress, or vomiting
- Last 10 minutes to 14 days
- Recur 2-3 times a month to once every 2-3 months
- Resolves by 2-3 years

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Case: Benign paroxysmal torticollis

- ? Labyrinthine involvement (eye rolling)
- ? Migraine variant in children
  - FHx migraine 73%
  - Coexisting typical migraine hx 18%

Evaluation
- Consider MR imaging of brain → c-spine
- Consider GERD

Treatment
- Reassurance
- Screen for delays → improves over time

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Case

- 6 month old with episodic spells of leg extension and fast breathing...
- Normal infant otherwise
- Distractable...

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Case: Infantile gratification

- Seen in all age groups, but manifestations difficult to recognize in infants/toddlers
- Suspected as either seizures or dystonia
- Rubbing thighs together, quiet grunting, irregular breathing, facial flushing, diaphoresis
- Context: boredom, excitement, anxiety
- Consciousness preserved... Distraction
  - Videotaping

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Case: Infantile gratification

- Evaluation:
  - Abuse history
  - Perineal exam (local irritation: cause or result?)

- Treatment:
  - Reassurance and education
    - Normalcy of behavior
    - Normalcy of development
    - "Gratification" vs. "Masturbation"
    - No scolding … use redirection

Case

- 8 y/o autistic boy who has strange sudden flapping movements of his hands when excited...

Case: Stereotypies

- Repeated, purposeless movements
- Can be hand flapping, clapping, running aimlessly, rocking, spinning, bruxism, rubbing the hands, etc ….
- Tend to occur with excitement or fear
- No alteration in consciousness
- Seen in normal children but usually diminishes by age 2 years
- Persistence seen in patients with ASD’s or chronic encephalopathies

www.youtube.com
Case: Stereotypies

- **Evaluation:**
  - Consider EEG if atypical or alteration in consciousness, urination, fall, etc.
  - Screening for ASD's and cognitive dysfunction
  - Also consider: tics?

- **Treatment:**
  - Reassurance
  - Ignore

Case

- 7 y/o irritable girl with funny eye movements
- Eventually has myoclonic jerks also

Case: Opsoclonus-myoconlus

- 6 months - 3 years
- Opsoclonus: rapid, conjugate, involuntary, chaotic eye movements
- Myoclonic jerks
  - Non-epileptic
- Ataxia
  - Truncal and gait > extremity
- Behavioral changes
  - Irritability
- "Dancing Eyes, Dancing Feet"
### Case: Opsoclonus-Myoclonus

- **Autoimmune**
- Associated with neuroblastoma
  - 50% of children with OMS
  - 1-2% of children with neuroblastoma present with OMS
- Usually low grade with good prognosis
- OMS also seen with infections
- Urine for VMA, HMA; MRI or CT, MIBG looking for neuroblastoma
  → Surgical resection
- ACTH, IVIG; others: cyclophosphamide, azathoprine
  - More recently Rituxumab (CSF B-Cells)
- Opsoclonus usually resolves, but behavioral, learning difficulties persist

### Case

- 6 y/o who slumps to the floor when she is startled or gets excited...

### Case: Narcolepsy

- **Cataplexy**: sudden loss of muscle tone from strong emotion or stimulus... laugh, excitement, startle
  - Partial paralysis of face or hands
  - Sudden head drop/knee bends
  - Fall to floor
  - No alteration of consciousness

- **Narcolepsy**
  - Daytime sleep attacks
  - Cataplexy
  - Sleep paralysis
  - Hypnagogic hallucinations

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**Case: Narcolepsy**

- Usually 2nd - 3rd decade, may occur after age 5 years → **adolescents**
- Sleep disorder of REM sleep... short latency from onset of sleep
- HLA DR2, etc.

**Evaluation:**
- PSG + MSLT (5 nap opportunities)

**Treatment:**
- Methylphenidate
- Modafinil
- TCA’s: suppress REM sleep, reduce cataplexy
- Naps
- ?Driving

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**Age: Adolescents**

**Awake:**
- Confusional/basilar migraines
- Hyperventilation syndrome
- Narcolepsy
- Tic disorders
- Pseudoseizures
- Conversion Disorder

**Asleep:**
- Confusional arousals
- Sleep walking
- Hypnic jerks
- Restless legs syndrome

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**Case**

- Adolescent with quick involuntary movements that are temporarily suppressible...
### Case: Tics/Tourette’s

- Stereotyped movement: sudden and quick, recurrent but non-rhythmic, purposeless but can be made to appear purposeful
  - Motor
  - Vocal
- Urge or sensation is suppressible temporarily
- Exacerbated by stress
- Should disappear during sleep

### Case: Tics/Tourette’s

- Reports up to 20% of children will have motor tics, at some point, usually mild and transient
- Onset 2-15 years, most severe 8-12 years, ½ resolve by adulthood
- M>F
- Wax and wane
- Type may shift over time
- **Transient tic disorder:** >4 weeks
- **Chronic tic disorder:** > 1 year
- **Tourette’s syndrome:** motor + verbal tics > 1 year

### Case: Tics/Tourette’s

- **Evaluation:**
  - Good history and exam
  - If explosive onset may consider PANDAS
  - Screening for comorbidities
    - ½ ADHD: Tourette’s Study Group, 2002: worsening of tics in Ritalin 20%, Placebo 22%
    - OCD
- **Treatment:**
  1. Reassurance and education
  2. Reassurance and education
  3. Alpha-2 adrenergic agonists: Clonidine (start 0.05mg)
  4. Neuroleptics: Risperdal (start 0.25mg)
  5. TCAs

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Conclusions

- We have reviewed a system of differentiating epileptic from non-epileptic paroxysmal events in children - realizing exceptions to “rules” exist
- We have reviewed classical clinical presentations and used visual examples in a case-based format to enhance pattern recognition of non-epileptic childhood spells
- We have reviewed when further evaluation is needed, when treatment might be indicated, and for atypical cases, when referral is appropriate

Questions?

- Finally, we have established that...
  - Kids do the funniest things!