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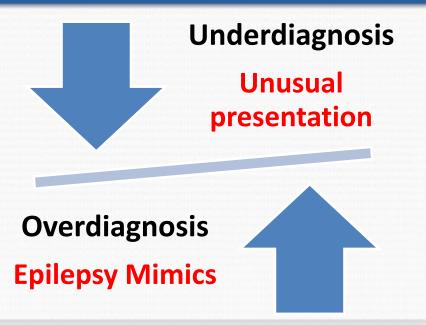




### **Diagnosis of Pediatric Epilepsy**

# AXIS | Is it epileptic seizure or not 1 → D.D. Of Pediatric epilepsy





#### **Jitteriness**

- The movement resembles a tremor
- Distinguished clinically from clonic seizures by:
  - No associated ocular movements
  - ✓ No Autonomic phenomena
  - ✓ Stimulus sensitivity ((eg, triggered by stimulation or easily stopped with passive movement of the limb).

### **Shuddering Attacks**

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

#### **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

### **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
- The most common stimulus is a painful event.
- The child turns pale (as opposed to blue) and loses consciousness with little if any crying.
- The EEG is also normal, and again there is no post ictal phase, nor incontinence.
- The child is usually alert within a minute or so.

# **D.D. Breath Holding Spells**

	Breath-holding spells	Epileptic seizures
Trigger	Crying, injury	Spontaneous, fever, sleep deprivation
Occurrence during sleep	No	May occur during sleep
Event	Sequence → provocation —apnea, cyanosis/ pallor, limpness.	Associated with stiffening and jerking of extremities
Postictal state	Usually brief	Maybe prolonged
Epileptiform abnormalities on EEG	Absent	Usually present
Treatment	Parental reassurance	Anticonvulsant therapy

### **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

### **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

### **Childhood Parasomnias**

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

### **Rhythmic Movement Disorder**

- Body rocking
- Head rolling
- Other less common muscle movements include:
- Body rolling
- Leg rolling
- Leg banging
- A combination of the aforementioned symptoms
- Head banging





### **Cataplexy**

- Cataplexy is a sudden and transient episode of muscle weakness accompanied by full conscious awareness
- Triggers: emotions (laughing, crying, or terror).
- Cardinal symptom of narcolepsy with cataplexy affecting roughly 70% of people who have narcolepsy

## **PseudoSeizures**

	Non-epileptic seizure	Epileptic seizure
Duration	Prolonged (several minutes)	Usually less than 2-3 minutes
Clinical features	<ul> <li>Fluctuating features</li> <li>Usually during wakefulness</li> <li>Preserved consciousness, avoidance behavior</li> <li>Side to side head movements</li> <li>Out of phase extremity movements</li> <li>Forward pelvic thrusting</li> <li>Emotional vocalization</li> </ul>	<ul> <li>Stereotypic features</li> <li>May occur in sleep</li> <li>Altered consciousness</li> <li>Head unilaterally turned</li> <li>In phase extremity movements</li> <li>Retropelvic thrusting</li> <li>Monotonous vocalization</li> <li>Pupillary reflex absent</li> </ul>
Incontinence	Rare	Present
Tongue bite	Occasional	Common
Postictal	None	Usually present
Affect	La Belle indifference	Concerned

# **Tics**

	Myoclonus	Tics
Onset	Any	School age
Pattern	Patterned, predictable, identical	Variable, wax and wane
Movements	Jerky, shock like, sudden	Blink, grimace, shrug
Rhythm	Maybe Rhythmic	Rapid, sudden, random
Duration	Brief, intermittent	Brief, intermittent
Premonitory	No	Yes
Trigger	No, action, stimulus sensitive	Excitement, stress
Suppression	No	Brief (causes inner tension).
Family History	May be positive	Frequently positive
Treatment	AED	Neuroleptics

### **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

### Syncope (Convulsive Syncope)

#### **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

### Paroxysmal Kinesigenic dyskinesia

- < 16 years, Duration: Secs. to few minutes</li>
- 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

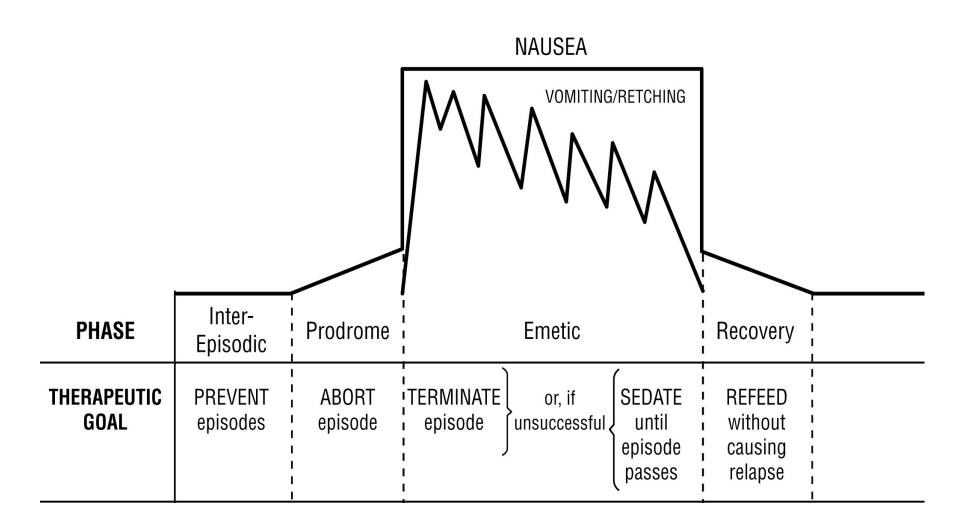
**Treatment:** If interferes with daily life

Low doses of Carbamazepine or phenytoin

# **Paroxysmal Kinesigenic Choreoathetosis**

	Paroxysmal Kinesigenic Dyskinesia	Paroxysmal Nonkinesigenic Dyskinesia	Paroxysmal Exertion-Induced Dyskinesia
Age of onset	Childhood/adolescence (majority of cases)	Childhood/adolescence (majority of cases)	Variable (depending on cause)
Episodes			
Triggers	Sudden movements or intention to move	Coffee, tea, alcohol (more consistent), anxiety, excitement, fatigue	Prolonged exertion (more frequently), fasting, stress, anxiety
Duration	Brief, majority of episodes <1 minute	Typically more prolonged, 10 minutes to 4 hours	Episode ends with rest
Frequency	Up to hundreds of episodes a day <sup>b</sup>	Weekly episodes more common <sup>b</sup>	Dependent on triggers
Treatment	Good response to antiepileptics; carbamazepine is drug of choice	Avoid triggering factors; poor response to benzodiazepines, phenytoin, acetazolamide, levodopa	Avoid triggering factors treat underlying cause when applicable
Etiology			
Gene associated with primary form <sup>c</sup>	Proline-rich transmembrane protein 2 (PRRT2)	Myofibrillogenesis regulator 1 ( <i>MR-1</i> ) <sup>d</sup>	Glucose transporter 1 (SLC2A1) <sup>e</sup> (in 20-25% of cases)
Secondary causes	Secondary to brain injury (vascular, trauma, multiple sclerosis)	Secondary to brain injury, symptomatic cases are rare	Parkinson disease, dopa-responsive dystonia

# **Cyclic Vomiting Syndrome**



### **Cyclic Vomiting Syndrome**

### New Diagnostic Criteria for Children

All of the criteria must be met to meet this consensus definition of cyclic vomiting syndrome.

- 1. At least 5 attacks in any interval, or a minimum of 3 attacks during a 6 mo period
- Episodic attacks of intense nausea and vomiting lasting 1 hr-10 days and occurring at least 1 wk apart
- 3. Stereotypical pattern and symptoms in the individual patient
- 4. Vomiting during attacks occurs at least 4 times/hr for at least 1 hr
- 5. Return to baseline health between episodes
- 6. Not attributed to another disorder

### To sum up

#### Neonates (0-8 wks)

- Jittery
- Startle reflex
- Benign sleep myoclonus

#### Infants (2mth-2yr)

- Breath holding spells Attacks
- Stereotypy (Mannerisms)
- Rhythmic movement disorder (sleep) -

Age/State Based Approach

#### Childhood (2-12yr)

- Staring /day dreaming
- Tics/Movement disorder
- Migraine equivalents
- Masturbation
- Drugs (Antiemetics)
- Stereotypy (Mannerisms)
- Parasomnias (sleep)

#### Adolescents (>12 yr)

- Syncope/Cardiac
- Psychogenic seizures

# **Epilepsy mimics during sleep/ wake**

	Wake	Sleep
Neonates	Jitteriness	Benign neonatal sleep myoclonus
<8wks	Stiff baby/hyperexplexia	
Infants	Breath holding spells	Rhythmic movement disorder
2 mo -2 ys	Shuddering attacks Spasmus Nutans Stereotypies Benign myoclonus of early infancy Hyperexplexia	

# **Epilepsy mimics during sleep/ wake**

	Wake	Sleep
Children 2-12 years	<ul> <li>Stereotypies</li> <li>Syncope</li> <li>Migraine/variants (cyclic vomiting, benign paroxysmal vertigo)</li> <li>Tics</li> <li>Paroxysmal choreoathetosis (kinesiogenic, dystonic)</li> <li>Gastroesophageal reflux</li> <li>Pseudoseizures</li> </ul>	<ul> <li>Head banging</li> <li>Parasomnias- Sleep terrors, sleep walking</li> <li>Hypnic jerks</li> </ul>

# **Epilepsy mimics during sleep/ wake**

	Wake	Sleep
Adolescents >12 years	<ul> <li>Syncope</li> <li>Migraine and variants</li> <li>Paroxysmal choreoathetosis (kinesiogenic, dystonic)</li> <li>Pseudoseizures</li> <li>Tremor</li> <li>Tics</li> <li>Transient global amnesia</li> </ul>	<ul> <li>Narcolepsy—steep paralysis, hypnagogic hallucinations</li> <li>Parasomnias</li> </ul>

# To sum up

Epilepsy mimic	Imitating epileptic condition/s
Syncopes	Generalised Tonic Clonic Seizures, Focal seizures, Absences, Drop attacks, myoclonic Epilepsies
Breath holding attacks	Tonic Spasms
Cataplexy	Atonic fits
Day dreams/Childhood Preoccupation	Absence epilepsy
Tics	Myoclonus, stereotypy
Sleep disorders	Frontal lobe seizures, Benign rolandic epilepsy
Benign Sleep myoclonus	Myoclonic Epilepsies, Focal Epilepsies
Migraines	Occipital lobe seizures, temporal lobe seizures
Pseudo seizures	Status epilepticus, tonic, tonic clonic seizures, absences, parietal lobe sensory seizures, Status non-convulsicus

