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Epileptic seizure VS Seizure mimickers

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Talk overview

- **Seizure mimics in children and adult**
- **Psychogenic non-epileptic seizures**
- **Pseudopseudoseizures**



Seizure VS Epilepsy

Seizure:

A transient occurrence of **signs** and/or **symptoms** due to abnormal excessive or synchronous neuronal activity in the brain

Epilepsy:

A disorder of the brain characterized by an enduring predisposition to generate epileptic seizures

ILAE OFFICIAL REPORT

A practical clinical definition of epilepsy

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SUMMARY

Epilepsy was defined conceptually in 2005 as a disorder of the brain characterized by an enduring predisposition to generate epileptic seizures. This definition is usually practically applied as having two unprovoked seizures >24 h apart. The International League Against Epilepsy (ILAE) accepted recommendations of a task force altering the practical definition for special circumstances that do not meet the two unprovoked seizures criteria. The task force proposed that epilepsy be considered to be a disease of the brain defined by any of the following conditions: (1) At least two unprovoked (or reflex) seizures occurring >24 h apart; (2) one unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years; (3) diagnosis of an epilepsy syndrome. Epilepsy is considered to be resolved for individuals who either had an age-dependent epilepsy syndrome but are now past the applicable age or who have remained seizure-free for the last 10 years and off antiseizure medicines for at least the last 5 years. "Resolved" is not necessarily identical to the conventional view of "remission or "cure." Different practical definitions may be formed and used for various specific purposes. This revised definition of epilepsy brings the term in concordance with common use.

KEY WORDS: Epilepsy, Seizure, Definition, Unprovoked, Recurrence.

Paroxysmal non-epileptic events

- Heterogenous group of time-limited events
- Changes in motor or behavioral activity beginning abruptly and ending in a short time
- Without abnormal **ictal EEG** changes
- Found in 5-10% of epilepsy patient



Seizure mimics in children

Seizure mimics in children

- Age-based approach
- The differentiation of seizure mimic from a seizure relies solely on a proper history and review of video recordings
- Investigations rarely help in the diagnosis
 - EEG for rule out epileptic phenomena

TABLE 1. Key features of seizures and seizure mimics.

Green Flags for Seizure Mimics	Red Flags for Seizures
Shaking is suppressible by tactile stimulation	Non-suppressible movements
Triggered by a specific event, location, or emotion	Occur when coming out of sleep
Staring or movements stop with distraction	Loss of consciousness during event
	Period of fatigue and confusion afterwards
	PMH of developmental delay, regression, or neurologic injury

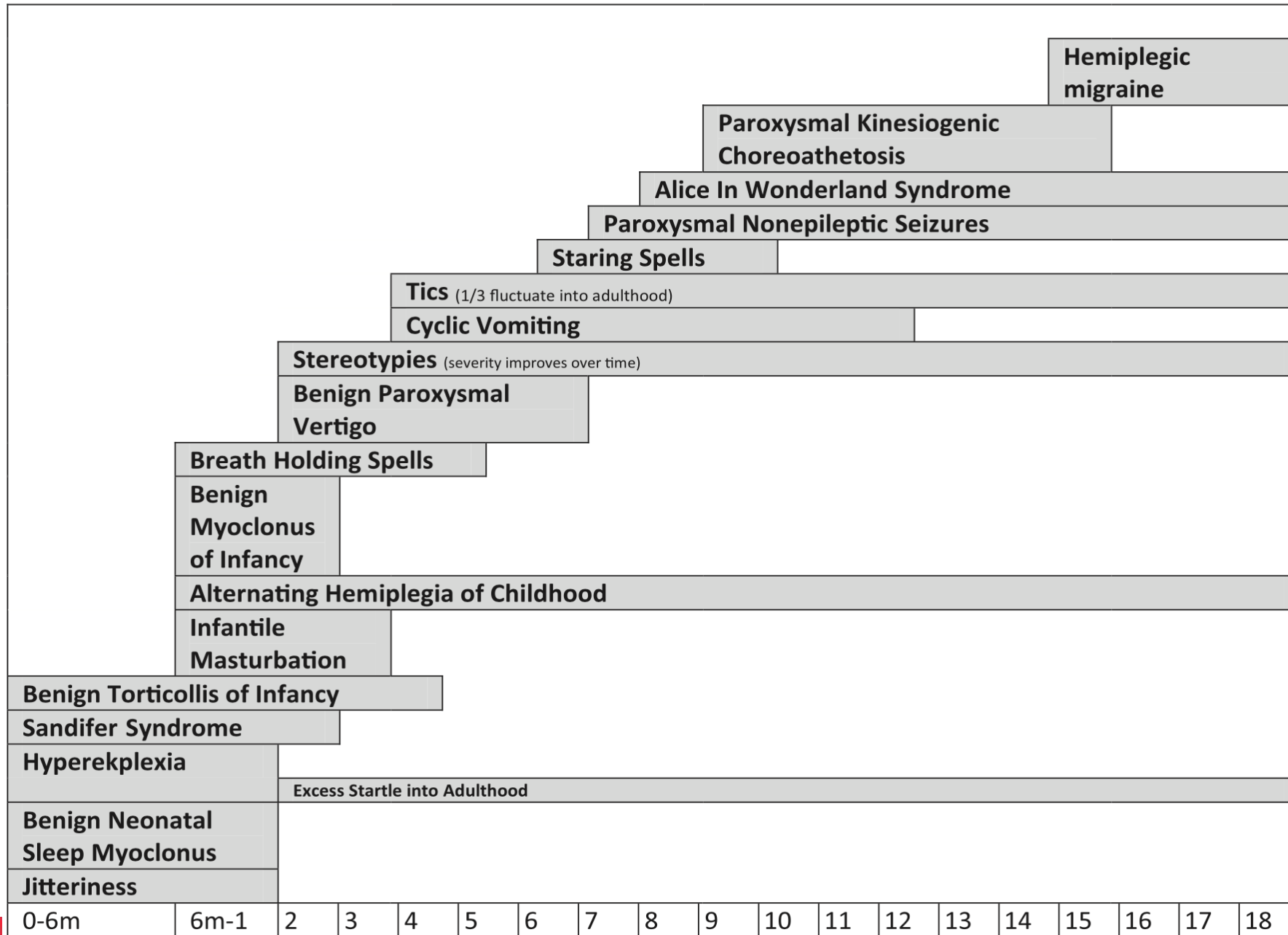


Figure 1. Age ranges (months and years) of seizure mimics, typical onset and resolution.

Seizure mimics in children

Neonates (Birth - 1 month)

Infants and young children (1 month – 2 years old)

School-aged children (2– 12 years old)




Neonates: birth - 1 month

- Jitteriness
- Benign neonatal sleep myoclonus
- Hyperekplexia (Excessive startle response)



Jitteriness

- Rhythmic, **oscillatory** movements
 - High frequency, low amplitude
- **Exacerbated by crying**, stress or sudden movements and resolve with change in position
- May be seen within the first days after birth and significantly improves by 2 weeks of life and resolves by 1 years
- Can be present as a sign of drug withdrawal, maternal CBD use, HIE, hypoglycemia, hypocalcemia or vitamin D deficiency



Benign Neonatal Sleep Myoclonus

- Bilateral or unilateral brief jerking movements
- Involve one, several or all limbs
- Movements can occur in clusters up to 30 minutes
- Only **occurs in sleep**
- Not always suppressible, do not stop with light touch
- May be seen as first few days of life
- Often resolve by 3-6 months



Hypererekplexia

- “Excess startle response”
- 3 components
 - Baseline generalized stiffness
 - Excess startle
 - Followed by transient stiffness
- Auditory, visual or tactile stimulation induces a startle episode
- Symptoms may appear at birth, should resolve by 1 year
- EEG helps to exclude seizures
- Imaging to rule out secondary conditions: brain stem compression, anoxic injury, tetanus
- Genetic defects: GLRA1, GLRB, SLC6A5
- Treatment: clonazepam


Infants and young children: 1 month – 2 years old

- Sandifer syndrome
- Benign paroxysmal torticollis of infancy
- Infantile masturbation
- Benign myoclonus of infancy



Sandifer Syndrome

- Back arching and stiffening of the neck or arms
- Resolve within three minutes
- Consistently occur after feedings, likely within 30 minutes
- Rarely occur in sleep
- Most cases occur in children under 2 years old
- Should evaluation for GERD



Benign Paroxysmal Torticollis of Infancy: BPT

- Painless torticollis
- Last minutes to weeks
- Side shifting
- The events occur on awakening or precipitated by changes in posture
- May be preceded by irritability, pallor, vomiting and ataxia
- Presents in first few months of life to 4.5 years old
- Strongly family history of migraine (55%)
- Self-limited by age 3-4 years
- Topiramate as an effective preventive agent
- Risk factors for developing BPPV, cyclic vomiting and migraine



Infantile Masturbation

- Self-stimulatory behaviors
- **Wide range of movements**
 - Torsional movement
 - Arched back
 - Body rocking
 - Vocalization such as grunting
 - Flushing or sweating
- No loss of consciousness
- Mean duration is 9 minutes (30s to 2h)
- Median age at onset is 12.5 months (3mo to 3y)
- Considered as **normal, healthy behavior**



Benign Myoclonus of Infancy

- Nearly identical to infantile spasms
- Brief jerking movements with flexion or extension of limbs and head
- Only occur during awake period
- Onset between 3 to 15 months
- EEG to rule out infantile spasm
- Benign condition, no long-term neurodevelopmental impairment

School-aged children: 2–12 years old

- Breath-holding spells
- Staring episodes





Breath-holding Spells

- Pallor or cyanosis from involuntary breath-holding episode which is normally preceded by a loud cry
- Can be **accompanying body stiffness or convulsions**, or transient loss of consciousness
- 0.1 – 4.6% of healthy children
- Onset between 6 to 18 months
- Spells gradually increase over time, reaching peak by 12 to 14 months
- Positive family history of breath-holding in 20 – 35%
- Association with **iron deficiency anemia** or iron deficiency
- Spells normally resolve spontaneously by around 5 years old

Staring Episodes

- Absence seizure like episode with **preservation of awareness**
 - Patient is easily distractable and responds to light touch or sound
- Typically occur in elementary school-aged children
- Occur in **specific situations** or settings: e.g. classroom
 - May resolve on weekends or vacations
- Self-limiting, no negative developmental outcome

Movement disorders

- Tics
- Motor stereotypies



Tics

- On-rhythmic brief rapid movements or vocalizations lasting several seconds
 - Eye blinking
 - Head jerking
 - Shoulder shrug
 - Vocal tics: grunting, throat clearing, vocalizing a syllable or word
- Often preceded by a **premonitory urge** that resolves after completing the tic
- Triggers: stress, excitement, fatigue, fever or infection

Tics

- Onset begin between 4 and 8 years old, more common in male (3-4:1)
- Significant comorbidities: ADHD, OCD, GAD, Sleep disorders, Mood disorders
- Treatment for patients with socially impacted: topiramate, clonazepam
- 1/3 will resolve, 1/3 will improve, 1/3 continue to fluctuate into adulthood



Motor stereotypies

- **Involuntary rhythmic purposeless** movements
- Lasting up to several minutes
- Present in a fixed pattern
 - Leg shaking
 - Thumb sucking
 - Nail biting
 - Teeth clenching
 - Head bobbing
 - Body rocking
- Children are typically unaware while doing the action



Motor stereotypies

- **Suppressible** by distraction or tactile stimulation
- **Specific emotional triggers:** excitement, boredom
- Onset earlier than tics, 24 to 36 months old
- High comorbidity rate for ADHD, learning disorders, tics, OCD
- Not harmful, reassurance is most important
 - Habit reversal training
 - Medications: fluoxetine, risperidone

Other seizure mimics

- Syncope
 - Cardiac
 - Reflex
 - Orthostatic
- Sleep disorders
 - Narcolepsy
 - Parasomnias
- Migraine
- Transient ischemic attack
- Movement disorders
- Hydrocephalus
- Psychogenic non-epileptic seizure



Non-REM parasomnias VS Nocturnal seizures

Table 2 Clinical Differences between Parasomnias and Frontal Lobe Seizures

	Non-REM Parasomnias (Somnambulism, Nocturnal Terrors)	Nocturnal Frontal Lobe Epilepsy
Age of onset	Usually < 10 years	Variable, generally childhood or adolescence
Positive family history	60–90%	< 40%
Mean number of attacks per day	1–2	> 3
Monthly frequency of episodes	Less than 1–4	20–40
Clinical course over time	Tendency to disappear in adolescence	Frequency of episodes generally stable over time
Mean duration of disorder	Around 7 years	Around 20 years
Duration of the episodes	Seconds to 30 minutes	Seconds to 3 minutes (usually less than 2 minutes)
Clinical features of the episodes	Variable complexity; not stereotyped in video recordings	Very stereotyped in video recordings, frequently vigorous or even violent movements
Triggering factors	Sleep deprivation, fever, alcohol, stress	Frequently no triggering factors are identified
Associated conditions	Obstructive apneas of sleep	Frequently none are identified
Ictal EEG	Slow waves, no epileptiform activity	Frequently normal or obscured by muscle artifact; clear EEG ictal pattern associated to episodes in < 10% of patients
Temporal occurrence during sleep	First half of the night, generally after 90 minutes of sleep	At any time during sleep, may happen in first 30–60 seconds
PSG stage	Stage III–IV	Generally stage II sleep, occasionally in stages III or IV



Psychogenic non-epileptic seizure

PNES

- **Definition**

- Paroxysmal changes in behavior, consciousness and autonomic function that resemble epileptic seizures but lack the electroencephalographic (EEG) signature of epileptic seizures

- **Challenging in diagnosis**

- Average of 7 years between the manifestation of spells and definite diagnosis

- **Often misdiagnosed and consequently are exposed to unnecessary antiepileptic drugs**

PNES

- Estimated incidence around 1.4-3 per 100,000
- Commonly in young adulthood (20-40 years)
- 75% of patients are females
- Increase the risk in patients with epilepsy

Psychogenic non-epileptic seizure

Table 1 Behaviors to distinguish psychogenic nonepileptic and epileptic seizures

Behavior	Psychogenic nonepileptic seizures	Epileptic seizures
Eye movement	Eyes closed during seizure onset for approximately 20 s; geotropic eye movements may be observed	Eyes open during seizure onset; may close for approximately 2 s
Weeping	May be present	Not present
Postictal nose rubbing and cough	Not present	May be present
Stertorous breathing postictally	Not present	May be present
Ictal stuttering and postictal whispering	May be present	Not present
Body movements	Pelvic thrusting; out-of-phase or side-to-side oscillatory movements; chaotic and disorganized thrashing	Pelvic thrusting; quick, tonic posturing; vocalization
Self-injury	May be present	May be present
Tongue laceration	May be present	May be present
Incontinence	May be present	May be present



Clinical clues

TABLE 2. Six-item tool for differentiating PNES from ES²³

Item	Epileptic seizures	PNES
Eyes	Opened	Closed
Head	Fixed/unilateral	Side-to-side movements
Limbs	In phase/same direction	Out of phase
Body (axis)	Straight	Opisthotonus
Body (movement)	No rotation	Intense rotation in bed
Evolution of seizure	Continuous	Fluctuating

PNES, psychogenic non-epileptic seizures; ES, epileptic seizures.

Psychiatric comorbidities in PNES

TABLE 1. Psychiatric comorbidities in PNES compared with ES²²

Comorbidity	Rates of comorbidity with PNES (%)	Relative risk of comorbidity when compared with ES
Total	53 - 100	1.3, statistically significant
Depression	8.9 - 85	1.6, statistically insignificant
Anxiety	4.5 - 70 (higher number from panic disorder)	1.82, statistically significant
PTSD	7 - 100	3.21, statistically significant
Personality disorders	5.4 - 74.3	1.73, statistically significant
Psychosis	0 - 15	
Substance abuse	9.8 - 29.5	

PNES, psychogenic non-epileptic seizures; ES, epileptic seizures.

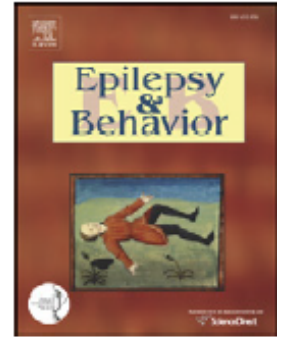
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The yield and clinical utility of outpatient short-term video-electroencephalographic monitoring: A five-year retrospective study

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Yield of vEEG and PNES

- Highest yield in psychogenic nonepileptic seizures (37.1%), interictal epileptiform discharges (17.2%) and epileptic seizures (6.9%)
- The provisional diagnosis was epilepsy 77.7% and PNES 22.3% before vEEG
- Changed the pre-test diagnosis in 30.9% of patients



Pseudopseudoseizures



Take home messages

- History taking and physical examination are still the most important
- Smartphone always helpful
- Both epileptic and non-epileptic phenomena may coexist

THANK YOU

GRACIAS
ARIGATO
SHUKURIA

GRAZIE
MEHRBANI
PALDIES

BOLZIN
MERCI

TASHAKKUR ATU
SUKSAMA
EKHMET

YOUSPAGARATAM
TINGKI
BIYAN
SHUKRIA

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