Differential diagnosis of Seizures in children

Sathida Poonmaksatit, M.D.
Division of neurology
Department of Pediatrics
King Chulalongkorn Memorial Hospital
Chulalongkorn University

Are these all epileptic seizures?

Outlines

- What are non-epileptic events
- How to approach
- Conditions that mimic sz and video examples

Epileptic seizures & Epilepsy

- Seizure: symptoms due to abnormally excessive or synchronous neuronal activity in the brain
- Epilepsy: the disease associated with spontaneously recurring sz by ILAE definition

1. A 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

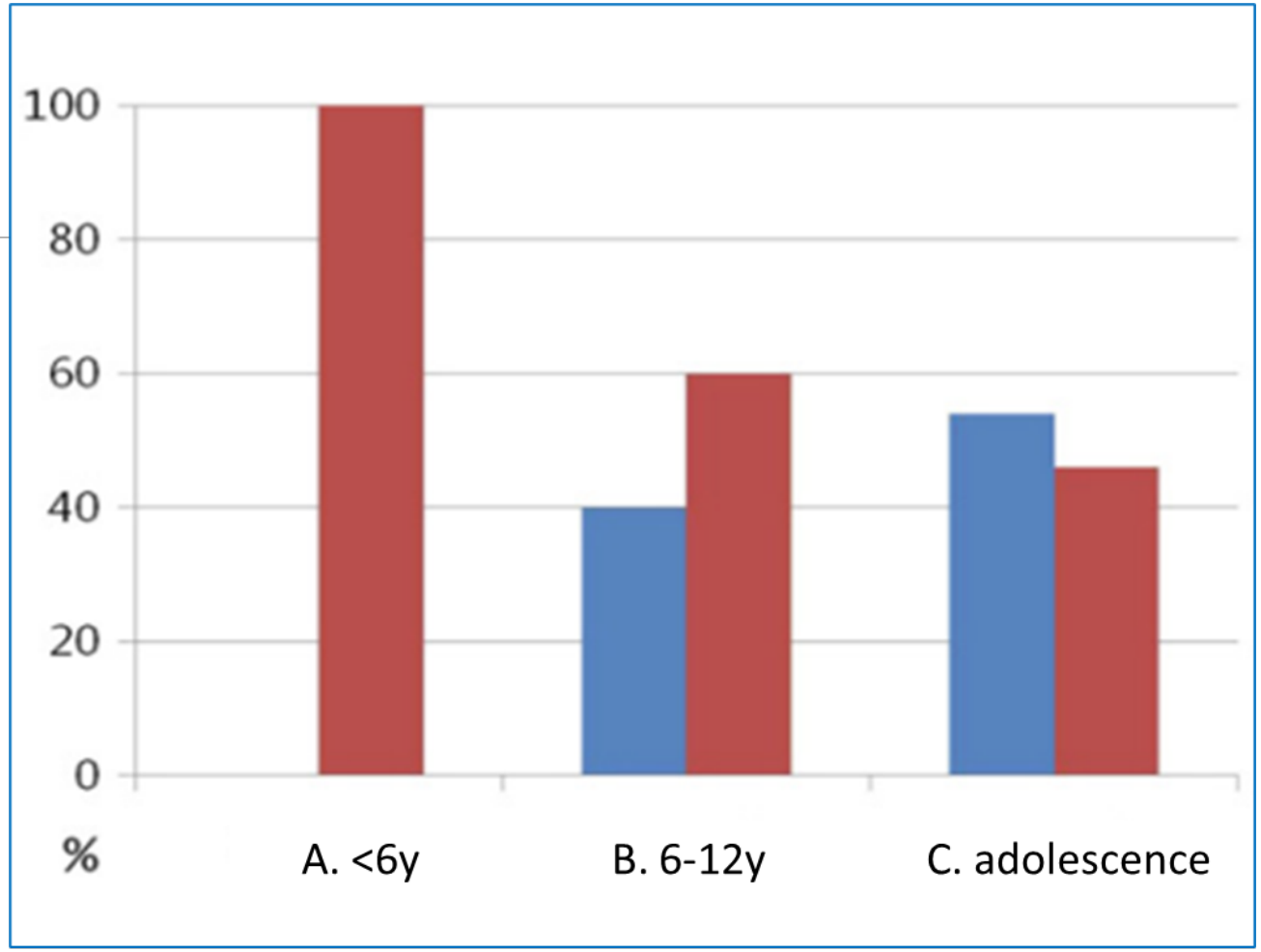
Epilepsia 2014

Paroxysmal nonepileptic events

- Paroxysmal events which resemble epileptic seizures
- Sudden, involuntary changes in behavior, sensation or consciousness
- Not accompanied by abnormal ictal EEG changes
- App. 20% of pts at epilepsy referral centers were PNEs
- 43% of children who underwent vEEG (Bye et al.)
- 5-10% of outpatient epilepsy have nonepileptic seizures compared with 20-40% of inpatient epilepsy

Epilepsy&Behavior 2015, Seminars in neurology 2008

Distribution of PNEs by age



Epilepsy&Behavior 2015

Diagnosis of paroxysmal events

- History and careful observation
- Video recording
- Investigations
  - Video EEG monitoring → seizure
  - Polysomnography → sleep disorders
  - Brain imaging → structural brain abnormalities
  - EKG monitoring → syncope

Factor favoring epileptic vs nonepileptic sz

	Epileptic	Nonepileptic
Age of onset	Any age	Adolescent more common
Sex	Men and women about equal	Females > males
Psychiatric history	May be seen	Usually present (very important to screen for history of abuse)
Motor semiology	Rhythmic, nonsuppressible movements of extremities, usually has a crescendo–decrescendo phase if clonic phase is present	Nonrhythmic, asynchronous movements including side-to-side head movements or presence of pelvic thrusting
Duration	Usually less than 5 minutes	Can be more than 5 minutes
Injury	Can be present	Often not seen
Level of consciousness with bilateral involvement during event	Should have decreased level of consciousness, unable to follow commands	Often retains consciousness and able to follow commands
Triggers	Rare, except in certain type of epilepsies (such as startle or reading epilepsy)	Highly suggestible or provoked by nonepileptic triggers

Seminars in Neurology 2020

Further investigations

- Blood chemistry
- CSF analysis
- Toxicology screening
- EEG
- Neuroimaging
- Genetic testing

DDx of sz

BY CLINICAL SYMPTOMS

- Syncope and other generalized paroxysms
- Movement disorders
- Oculomotor and visual abnormalities
- Sleep related disorders

BY CAUSES

- Non-neurological conditions
  - Physiologic phenomenon
  - Behavioral phenomenon
  - Psychiatric disorders
  - Sleep disorders
- Neurological disorders
  - Migraine
  - TIA
  - Movement disorders

Nelson textbook of Pediatrics, 21ed. Aust Fam Phys 2005

Conditions that mimic seizures by age

Age	Syncope and other generalized paroxysm	Movement disorders	Oculomotor and visual abnormalities	Sleep related disorders
Neonate	Apnea	Jitteriness Tremor Increased startle reflex hyperekplexia	Paroxysmal tonic upward gaze	Benign neonatal sleep myoclonus Sleep transition disorders REM
Infant	Breath-holding spells Paroxysmal vertigo	Sandifer syndrome Benign myoclonus of infancy Shuddering attack Benign paroxysmal torticollis Jactatio capitis (head banging)	Paroxysmal tonic upward gaze Spasmus nutans Opsoclonus-myoclonus daydreaming	NREM partial arousal disorder REM sleep disorders Narcolepsy Sleep transition disorders
Children & adolescents	Benign paroxysmal vertigo Familial hemiplegic migraine Syncope Psychogenic seizure Hyperventilation spell Factitious disorder	Tics Tremor Paroxysmal dyskinesia Alternating hemiplegia Episodic ataxia Cataplexy Psychological disorder	Staring, daydreaming Drug reaction Hallucination Conversion reaction Factitious disorder	NREM partial arousal disorder REM sleep disorders Narcolepsy Sleep transition disorders Sleep myoclonus Restless leg syndrome

Nelson textbook of Pediatrics, 21ed.

Common paroxysmal nonepileptic events

NEWBORN	INFANT
• apnea • Jitteriness • benign neonatal sleep myoclonus	• breath holding spell • shuddering • spasmus nutans • Sandifer syndrome • hyperekplexia

Common paroxysmal nonepileptic events

CHILDHOOD	ADOLESCENT
• Tics	• Syncope
• self gratification	• paroxysmal dyskinesia
• sleep myoclonus	• migraine
• sleep disorders	• cataplexy-narcolepsy
	• pseudoseizure
	• hyperventilation syndrome

Jitteriness

- Most common in neonatal period
- Age at onset <1wk, Resolution before 6mo
- Generalized, symmetric, rhythmic oscillation
- Stimulus sensitive, but suppressible
- Cause
  - Idiopathic
  - Assoc with: HIE, ICH, hypoglycemia, hypo- Ca/Mg, drug withdrawal
- Outcome: depends on underlying cause
- No treatment needed

Benign neonatal sleep myoclonus

- Repetitive myoclonus only in sleep
  (most likely in quiet (non-REM) sleep)
- Onset during first week of life
- Prevalence 0.8-3 per 1000 births
- Diminish in the 2nd mo and usually gone by 6 mo
- Neuro exam and EEG: normal
- No treatment required and normal outcome

Breath holding spells

- Involuntarily holding breath during expiration
- Incidence 4.6-4.7%

- Onset 6-18 mo
- 2 types: cyanotic vs pallid
- Resolution: 50% by 4 yr of age

  : almost all by age 7-8 yr
- 30% of these develop syncopal episodes in adolescence
- Treatment: reassure, iron therapy
- Reflex anoxic sz may follow spells

Shuddering attacks

- Shivering, rapid tremor of head, shoulders and arms (+face grimacing)
- Triggered by excitement, frustration or surprise
- Key: Preservation of consciousness, predictable triggers, ability to abort an episode when distracted and normal EEG
- Onset in infancy to early childhood
- Treatment is generally not needed

Sandifer syndrome

- Generalized stiffening and opisthotonic posturing, may be with apnea, staring, and minimal jerking
- Often occur after a feed, last 1-3 min
- Associated with reflux
- Incidence <1% of children with GERD
- Confirm diagnosis, treatment of reflux

Hyperekplexia

- 3 components: excess startle, transient stiffening, and baseline stiffness
- induced by auditory, visual or tactile stimuli
- appear at birth
- exclude secondary causes
  - acquired: brainstem compression, anoxic injury, tetanus
  - complex genetic neurodevelopmental: Tay Sachs, pontocerebellar hypoplasia type2, GLYT1 encephalopathy
- clonazepam for primary treatment

Spasmus nutans

- Triad: head tremor, nystagmus, and torticollis
- Onset 3-8 mo of age
- Unknown cause
- Visual abnormalities and posterior fossa lesion should be excluded
- Generally, resolves within several months

Self gratification

- Infantile masturbation
- Onset: 2months to 3 years
- Stereotyped posturing, pressure to pubic area
- Quiet, grunting, diaphoresis or facial flushing
- Lasting less than a minute (but can be hours)
- No alteration of consciousness
- Cessation with distraction
- Parent education and reassurance are important

Tics

- sudden, rapid, repetitive, and nonrhythmic movements or vocalizations
- Urge to do, briefly suppressible
- Exacerbated by stress, anxiety, excitement or fatigue
- Onset motor tics: 4-8 yr
- Waxing and waning course, remission can occur in later teen years
- Family Hx of tics or OCD

Tics

- Motor tics: simple
  (blinking, nose wrinkling, shoulder shrugging)
  : complex
  (facial and body contortion, hitting, jumping)
- Vocal tics : simple (grunt, bark, sniff, throat clearing)
  : complex (repetition of words, phrases)

- Treatment: behavioral therapy (CBIT), acupuncture, rTMS
  : pharmacotherapy (nonneuroleptic, typical and atypical neuroleptics, and others)

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

Syncope

- Transitory, brief loss of consciousness with loss of postural tone
- Caused by a decrease in global cerebral perfusion

Differences between sz and syncope

	Seizures	syncope
Precipitating factors	Sleep deprivation	Pain, fright, standing
Premonitory symptoms	No	Dizziness, vision loss, pallor
Duration of conscious loss	Minutes	Seconds
Clonic jerking	Yes	Sometimes
Recovery from attack	Delayed	Rapid
Postictal	Yes	No
EEG	Abnormal	Normal

Aust Fam Physician 2005

Pseudoseizure (PNEs, psychogenic nonepileptic sz)

	Psychogenic seizure	Epileptic seizure
Onset /offset	Slow onset and offset	Acute onset , abrupt offset
Duration	Usually > 3 minutes	Typically < 3 minutes
Movement	Flailing, thrashing, combativeness	Synchronous
Pelvic thrusting	More common	Never
Injury during event	uncommon	Frequent
Incontinence	Occasional	Frequent
Ictal Eye Closure	Frequent	Rare
Provocation	Common	Rare
Time of day	During wakefulness	During awake or asleep
Postictal behavior	None	Common(lethargy,confuse)
EEG	Normal	Abnormal

Cataplexy

- Sudden, transient muscle weakness in all skeletal muscles (exc. Extraocular and resp. muscles)
- Associated with emotional stimulus (e.g., laughter)
- Most specific symptom of narcolepsy (type1) which is childhood onset
- Dx: polysomnography and MSLT
- Treatment of cataplexy: antidepressants

Take home message

- **History and observation** is important
- **Video** recording is very helpful
- Epileptic & nonepileptic events may coexist!!

Thank you

ขอบคุณสำหรับวิดีโอ จาก
อ.นพ.วิโรจน์ พงษ์พันธุ์เลิศ
อ.พญ.ธนิตนันท์ ภาปราชญ์
อ.พญ.รับพร สุนทรโลหะนะกูล