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EPILEPSY *in* CHILDREN

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Achieving MDG-4: Strategies & Action
PEDICON
PEDICON PUBLICATION 2014



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What is Not Seizure? How to Differentiate?

2

Renu Suthar • Naveen Sankhyan

INTRODUCTION

A large number of children who are diagnosed as epilepsy do not really have epilepsy. These children often have non-epileptic events (NEE) that are mistaken for epilepsy. Among children who are evaluated for refractory epilepsy prevalence of NEE is about 3.6–7%. With more and more reliance on technology rather than a thorough clinical evaluation the rate of misdiagnosis is likely to increase. Even when a clinical evaluation is done, it is important that the clinician is aware of the varied age appropriate causes of paroxysmal events in children. Paroxysmal events in childhood can be divided into three major categories: (1) Epileptic seizures, (2) Psychogenic non-epileptic seizures or (3) Physiologic non-epileptic events. Epileptic seizures are defined as transient occurrence of sign and symptoms due to abnormal, excessive, synchronous neuronal activity. Psychogenic non-epileptic seizures are paroxysmal time-limited, alterations in motor, sensory, autonomic, and/or cognitive signs and symptoms, but unlike epileptic seizures, psychogenic non-epileptic seizures are not caused by abnormal neuronal activity. Psychogenic non-epileptic seizures have psychologic underpinnings and causes. Physiologic non-epileptic events are events associated with systemic alterations that produce an event which can mimic a seizure (e.g. movement disorders, breath holding, shuddering) or cause a seizure through non-epileptic mechanisms (e.g. anoxic seizures associated with prolonged breath holding) (Table 2.1).

The best method to diagnose NEE is a thorough history from a reliable source, relevant examination, few investigations (occasionally), and if possible observation of the events or videos of the events. Few of the common paroxysmal non-epileptic events in children are discussed here.

BREATH HOLDING SPELLS (BHS)

These events are benign, but can be frightening to parents and other observing an episode. Breath holding spells are more common in boys. The typical age of onset is between 6 months to 18 months, and only 10% of children have onset after the age of 2 years. Two types of breath holding spells are described based on color change during the event.

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Table 2.1: Classification of non-epileptic events

Physiological	*Psychological*
Breath holding spells	Misinterpretation of physical symptoms
Reflex anoxic seizure	Anxiety disorder
Syncope	Conversion disorder
Paroxysmal dyskinesia	Dissociative disorder
Sleep disorders	Hypochondriasis
Childhood periodic syndromes	Psychosis
Stereotypies	Somatization disorder
Self-gratification disorder	Response to acute stress without evidence of psychopathology

Cyanotic Spells

These are most common of the two types of spells. They occur in neurological normal children, and are precipitated by anger, or frustration. When child is being denied of something or scolded, the child typically cries vigorously usually for less than 15 seconds and then becomes silent and holds the breath in expiration. Apnea is associated with rapid onset of cyanosis; there may be loss of consciousness and brief limpness followed occasionally by opisthotonus posturing. Rarely brief clonic movements may occur. Recovery usually occurs within 1 minute, with child having few gasping respiration and then a regular breathing and consciousness.

Pallid Spells

These are provoked by sudden fright or pain, by sudden striking of head or startle. The child may gasp and cry for a very brief period and then becomes quiet, lose consciousness and becomes pale. There may be associated bradycardia and transient cardiac asystole. In a severe episode, clonic movements of extremities and incontinence may occur. The diagnosis is established by a careful history elucidating the typical sequence of events. Questioning should focus on precipitating events, absence of breathing, facial color and family history. Laboratory test are not required. In cases where history is not clear, a video recording by parents may be helpful in confirming the diagnosis. Electroencephalogram (EEG) is generally not indicated, though electrocardiogram (ECG) should be considered to detect long QT syndromes.

The most important step in management of BHS is reassuring parents. Iron supplements are indicated if iron deficiency anemia is present and may be helpful in those without anemia as well. Though rarely indicated atropine (0.01 mg/kg two or three times daily) is effective for pallid BHS. Brief seizures associated with breath holding do not require antiepileptics.

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REFLEX ANOXIC SEIZURES

These are non-epileptic events resulting from cardiac asystole of vagal origin. Reflex anoxic seizures have been reported with minor blows to the occiput, expelling hard stools past an anal fissure, venipuncture, intramuscular (IM) injections, and seeing an intravenous (IV) scalp drip. Reflex anoxic seizures occur a few seconds after the provocation and are characterized by loss of muscle tone initially and later by tonic posturing. There may be opisthotonic posturing in some patients. A few jerks at the onset and end of an anoxic seizure may occur and probably represent myoclonic phenomena. A snoring type of inspiration or snort occurring close to the restoration of the cardiac rhythm is often noted. Urinary incontinence happens in approximately 10% of children with anoxic seizures, with bowel incontinence occurring less commonly. The color change seen with an anoxic seizure may be cyanosis or pallor, depending on the mechanism producing loss of consciousness. The duration of unconsciousness is almost always less than 1 minute. Most patients experience a rapid recovery of consciousness, but some will be dazed or disoriented for a short period. Some will be drowsy after an episode and may sleep. Diagnosis of reflex anoxic seizures is usually made by obtaining a history of the episode. Details about the precipitating circumstances and onset are essential. ECG is indicated to rule out cardiac arrhythmias, and management consists of reassuring the parents and explaining non-epileptic nature of events.

SYNCOPE

Syncope is defined as a temporary loss of consciousness and postural tone resulting from transient and diffuse cerebral hypoperfusion, followed by spontaneous recovery with no neurological sequelae. Syncope is a common clinical problem affecting 15–25% of children and adolescents prior to adulthood. The event is typically preceded by a prodrome lasting several seconds to 1–2 minutes, which has distinctive premonitory features such as nausea, epigastric discomfort, blurred or tunnel vision, muffled hearing, tinnitus, dizziness, light-headedness, diaphoresis, hyperventilation, palpitations, pallor, cold and clammy skin, or weakness. Although syncope is commonly a benign self-limiting event, rarely it may be the first warning sign of a serious underlying cardiac or noncardiac disease. Syncope may result from cardiovascular or neurological causes (Table 2.2).

Table 2.2: Non-epileptic events as seen at different ages

Neonates and infants	*Toddlers*	*Children*
Jitteriness	Breath holding spells	Syncope
Benign sleep myoclonus	Benign paroxysmal vertigo	Migraine
Self-gratification behavior	Self-gratification behavior	Day dreaming
Shuddering attacks	Sleep disorders	Tics
Gastroesophageal reflex	Gastroesophageal reflex	Pseudoseizures
Breath holding spells	Stereotypies	Sleep disorder

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Cardiovascular-mediated Syncope

Among cardiovascular causes aortic stenosis, hypertrophic obstructive cardiomyopathy, sick sinus syndrome, long QT syndrome, Wolff-Parkinson-White (WPW) syndrome, tachyarrhythmia and heart block are important ones. Risk factor associated with cardiovascular syncope are presence of heart murmur and congenital heart disease. Acute attacks associated with hyperpnea or cyanosis, abrupt loss of consciousness during exercise and absence of usual premonitory symptoms or precipitating factors associated with neurally mediated syncope. Prolonged ECG and Holter monitoring are needed for confirmation.

Neurocardiogenic Syncope

Neurocardiogenic syncope can occur at any age. It is precipitated by upright posture, prolonged standing, sudden change in posture, heat, fatigue, hunger and a concurrent illness or stress. A detailed history usually reveals contributory environmental factors before the loss of consciousness and postural tone. The loss of consciousness is usually brief, lasting from a few seconds to 1–2 minutes, followed by rapid spontaneous recovery without neurological deficits. During the ictus the patient may have tonic posturing or a brief clonic seizure, rarely associated with incontinence. Distinguishing between neurocardiogenic syncope and seizure is a common clinical dilemma and a frequent source of diagnostic error. Typical features like precipitated by prolonged standing, noxious stimuli, gradual onset with premonitory symptoms, associated with facial pallor, diaphoresis and immediate recovery suggest neurocardiogenic syncope. An ECG should be obtained on all patients who present with syncope, especially if it is recurrent or occurs with exercise.

SLEEP DISORDERS

Sleep disorders and other sleep events can be confused with seizures. The common sleep disorders confused with seizures are divided into those occurring in non-rapid eye movement (NREM) and REM sleep.

Non-REM Sleep Disorder

NREM parasomnias include confusional arousals, sleepwalking, and sleep terrors. These are most commonly seen during N3 sleep and are thus prevalent in the first third of night, when this type of sleep is most abundant. NREM parasomnias occur during attempted transition from N3 into N2 or N1 sleep, with consequent partial arousal and non-epileptic bizarre behaviors. Confusional arousals are seen most often in toddlers, whereas sleepwalking and sleep terrors may occur throughout the first decade. Children with confusional arousals may sit up in bed, moan or whimper inconsolably, and utter words like “no” or “go away.” Autonomic activation,

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characterized by flushing, sweating, and piloerection, is minimal to absent. The event generally last 5–30 minutes, after which the patient gradually goes back to sleep and has no recollection of the events the subsequent morning.

Night Terror

These events occur during partial arousal from NREM sleep, typically occurs once in a night, within 1–2 hours of falling asleep. They are characterized by marked autonomic features including tachycardia, tachypnea, tremulousness, mydriasis, and sweating. The typical episode lasts for a few minutes, and stops abruptly and child rapidly returns to deep sleep. Typically child does not remember the event next morning. Night terrors can be confused with nocturnal frontal lobe epilepsy, however, the seizure events are shorter in duration (often 20–30 seconds), and are accompanied by more prominent head segment automatisms, such as lip smacking, eye deviation or chewing. Further, the patient may have partial recall of the event and headache upon awakening in the morning.

REM Sleep Disorder

The REM sleep disorders include:

- Nightmares, wherein the child gets up frightened after a bad dream, and then becomes fully awake.
- Sleep paralysis, the child is unable to move for a brief period and feels very frightened.

Shuddering Attacks

These events generally starts in infancy and can begin as early as 4–6 months of age. These episodes are characterized by brief shivering like movements, stiffening, adduction of arms and thighs, flexion of head and flexion or extension of neck. There is no change in level of consciousness during these movements. There may be several attacks per day which can occur spontaneously or after certain precipitating factors such as fear, anger, frustration or embarrassment. Most children improve spontaneously before the age of 10 years. The management of these events requires reassurance of parents and avoiding any antiepileptic drugs.

PAROXYSMAL NON-EPILEPTIC SEIZURES (PNES)

These events are known by various terms like pseudoseizures, non-epileptic seizures, hysterical seizures, and psychogenic seizures. PNES is preferred terminology as it is non-judgmental and more acceptable to parents. They are commonly seen in adolescents and are commoner in girls. They can occur in various forms and combinations such as convulsive movements, sudden slow limpness, tonic posturing, unresponsiveness and bizarre clonic jerks. It may be difficult at times to differentiate

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true epileptic seizures from psychogenic non-epileptic seizures. Important feature of PNES that help differentiate them from epileptic seizures are: bizarre, rhythmic coarsely clonic movements, pelvic thrusting, tendency to occur in presence of others, no history of incontinence, self-injury or typical ictal tongue bite (sides). Children with PNES may have the ability to enact the event and at times stop the event on command. The duration of events is prolonged as compared to true seizures and frequency is generally high. Psychosocial stress is more common in children with PNES. Diagnosis can be confirmed with a video EEG monitoring, where the clinical event can be captured. Normal ictal EEG in a typical clinical event confirms the diagnosis. Efforts must be made to find out the precipitating stress factor. Appropriate psychology and psychiatry consultation should be sought.

STEREOTYPIES AND SELF-STIMULATION

Repetitive stereotyped movements are very common among children with autism, mental retardation, and sensory deprivation; however, they are also observed in non-autistic children with normal cognition. These movements involve rhythmic motor behaviors, self-stimulating movements, and gratification phenomena, an extreme example of which is masturbatory behavior. Examples of self-stimulating movements or gratification phenomena include body rocking, head banging (jactatio capitis), and head rolling. Rocking motions of the trunk observed among mentally and visually impaired children may represent a form of vestibular stimulation akin to the maternal rocking that effectively comforts a crying, tired, or sleepy infant. More difficult to recognize is masturbatory behavior, particularly when this occurs in young children and infants. The repetitive movements usually involve the lower trunk and may be accompanied by pelvic thrusting or contractions of the gluteal muscles. Thighs may adduct and rub against each other. The event if uninterrupted may be prolonged. The primary differentiating feature is that the child can be distracted out of the event. Management again involves reassurance and explanation to the parents and avoiding unnecessary investigations and drugs.

POINTS TO REMEMBER

- The best method to diagnose non-epileptic events is a thorough history from a reliable source, relevant examination, few investigations (occasionally), and if possible observation of the events or videos of the events
- Physiologic non-epileptic events and psychogenic non-epileptic seizures must always be considered and excluded before making a diagnosis of epilepsy
- Breath holding spells and reflex anoxic seizures are often confused with epilepsy in young children, particularly when associated with brief clonic movements
- Syncopal events are commonly confused with seizure in older children. Precipitating events, warning symptoms, pallor and sweating favors diagnosis of syncope
- Pseudoseizures are not uncommon in older children, very frequent events, preserved consciousness, bizarre movements and ability of children to enact the event and at times stop the event on commands are some of the features helps in differentiation from true seizures.

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EPILEPSY *in* CHILDREN

Epilepsy in Children is a common yet incompletely understood problem by general pediatricians. An exhaustive list of causes and multiple classifications add to the complexities of the issues. A plethora of new antiepileptic drugs has come in market and often we face difficulty in choosing appropriate molecule. Silver lining is that most of pediatric epilepsies are amenable to treatment with an excellent long-term outcome, provided the workup and management plan is executed methodically. Hence, this book, where Pediatric Neurologists from Indian Academy of Pediatrics (IAP) have given a knowhow of Pediatric Epilepsy in a simplified way. So, this book becomes an essential companion of pediatricians and residents.



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Shelving Recommendation
PEDIATRICS

ISBN 978-93-5152-134-1

Available at all medical book stores
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