CME

Seizure Patterns in Childhood

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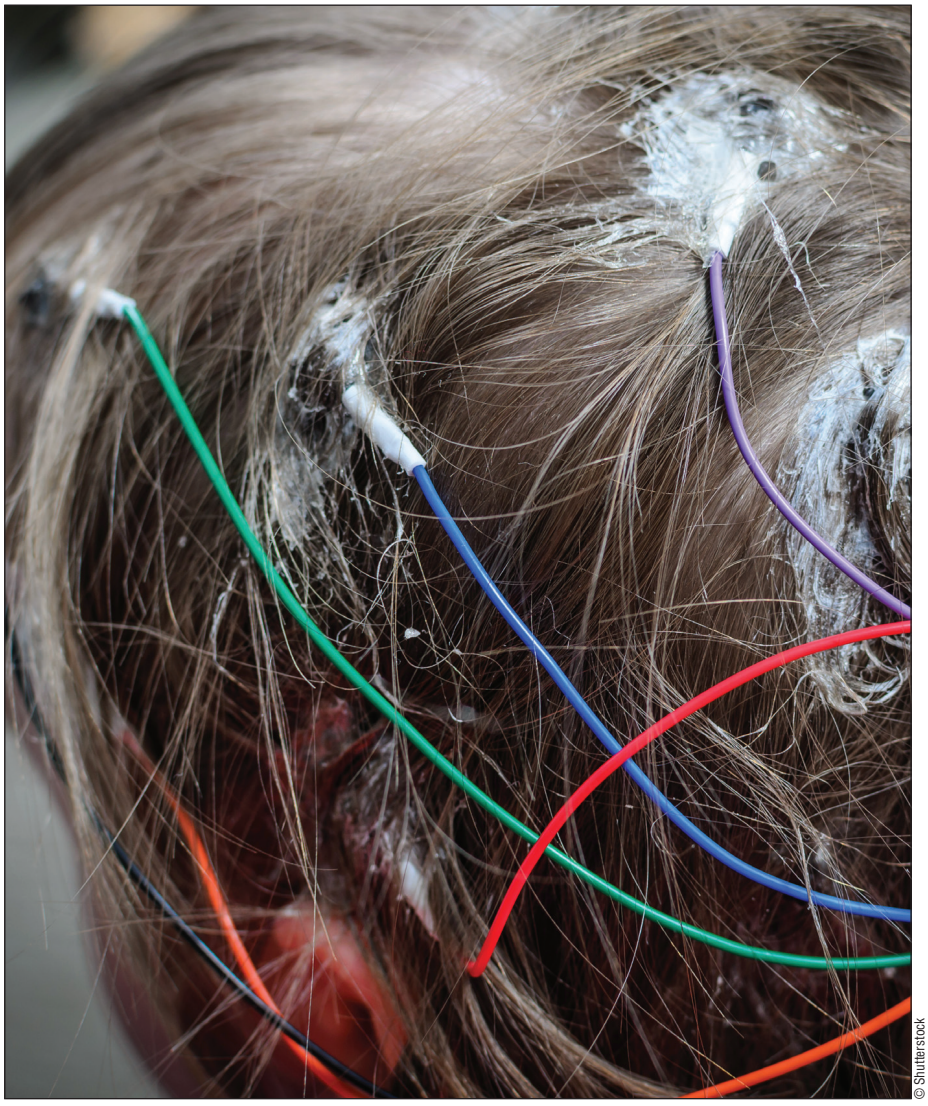
Abstract

Seizures are alarming and can be life-threatening. It is essential that pediatric providers be able to identify those paroxysmal events most likely to be seizures and to know which require immediate evaluation. Severity can range from childhood syndromes that are controlled relatively easily and usually outgrown to epileptic encephalopathies that are associated with severe developmental delay and sometimes death. Familiarity with seizure semiology can guide early diagnosis and treatment. **[*Pediatr Ann*. 2015;44(2):e24-e29.]**



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Disclosure: Steven M. Wolf and Patricia E. McGoldrick both report receiving consulting fees from INSYS Therapeutics and fees for non-continuing medical education services from Lundbeck A/S.

doi: 10.3928/00904481-20150203-08

Whether a child comes to your office with a clear story of a seizure or with odd complaints of strange sensations or unusual movements, such as head bobbing or episodes of "zoning out" and distractibility, it is important to obtain a full history and description of the events. The age of the child and the reliability of the observer will influence the description of the events; therefore, it is very important to gather as much information as possible regarding the episode(s) so that the seizure type can differentiated and managed properly. This article provides a simplified overview of the different ways in which seizures may present.

As a primary care provider, you should have a relationship with one or more neurology teams in which you can you can pick up the phone and ask them about a case. Sometimes, having that conversation while the patient, family, or observer is sitting in the room with you will lead to a decision for immediate admission for a video electroencephalogram (EEG) or some other basic work-up to help exclude a confounding disorder.

The International League Against Epilepsy classifies epilepsies according to (1) whether or not consciousness is altered; (2) the location of the disease in the brain; (3) the cause of the epilepsy; and (4) the main symptoms (ie, what the seizures look like and how often they occur).

Seizures, however, are classified solely on the basis of their symptoms. **Table 1** lists the different types of seizures. The older seizure classification system was based on whether the seizures were partial (focal, denoting that they arose from one area of the brain) or generalized (arising from the whole brain). Under that classification system, focal seizures included simple (no change in consciousness) and complex partial (patient was not alert). Generalized seizures included myoclonic, absence, generalized tonic-clonic, and atonic, as well as spasms.

For the purposes of this article, we will discuss the seizure semiologies most frequently encountered in childhood, the syndromes in which they occur, and a brief note about treatment options. Syndromes and treatments are more thoroughly discussed in other articles in this issue.

SEIZURES WITH ALTERATION OF CONSCIOUSNESS

Dialeptic Seizures

Absence Seizures

Absence seizures are typically brief alterations in the level of consciousness. They last for 5-15 seconds and are often accompanied by an eye roll or an eye flicker or deviation. Typically, family members or teachers will describe them as "zone outs" or "staring spells." They are also described as "behavioral arrest." The diagnosis is confirmed by EEG findings. Routine EEGs are performed in the office and takes anywhere from 20 minutes to 1 hour, whereas a more thorough evaluation includes an overnight or 24-hour EEG that may be performed on an inpatient or outpatient basis and with or without accompanying video. The purpose of the longer recording is to quantify the frequency of the epileptiform discharges and the response to treatment, as well as to correctly diagnose either absence or atypical absence. This episode may be precipitated by hyperventilation. A prolonged test for 24 hours, with or without video, is useful in confirming a diagnosis when a routine 20-minute test does not.

Absence seizures are most commonly associated with a diagnosis of childhood absence epilepsy (CAE). Absence seizures may also occur in juvenile absence epilepsy, in which absence seizures present between ages 9 and 13 years. About 80% of these children and adolescents also have generalized seizures and may have myoclonic seizures.[1]

TABLE 1.

Classification of Seizures

Dialeptic seizures
- Staring spells
- "Zoning out"
- Confused

Motor seizures
- Myoclonic seizures (quick jerking)
- Tonic seizures (stiffening)
- Clonic seizures (repetitive rhythmic jerking involving both sides of the body)
- Tonic-clonic seizures (alternating jerking and stiffening)
- Versive seizures (sustained, forceful turning)
- Hypermotor seizures (large, violent movements)
- Automotor seizures (automatisms of the hands, mouth or tongue, consciousness is usually compromised)
- Spasms (sustained muscle contractions with flexion of the trunk and elevation of the arms)

Other seizure types
- Gelastic seizures
- Hypomotor seizures (inability to move)
- Astatic or atonic seizures (loss of tone, with falls or head drops)
- Aphasic seizures (inability to speak)
- Sensory seizures (unusual feelings or sensations)
- Autonomic seizures (piloerection, epigastric rising, bizarre heart racing)

Contrary to popular belief, absence seizures do not occur only in childhood absence epilepsy, but in several other less benign syndromes such as Doose syndrome and Dravet syndrome.

Atypical Absence Seizures

Atypical absence seizures last longer (up to 20 seconds) than absence seizures and are not as easily controlled. The beginning and end of the episodes are not as clearly defined, and the spike and wave abnormalities on the EEG are of lower voltage (<2.5 Hz).

Atypical absence seizures occur in individuals who are diagnosed with Lennox-Gastaut syndrome. This syndrome represents about 1%-10% of childhood epilepsies and is diagnosed when there is a triad of multiple seizure types, mental retardation with behavioral disturbances, and an EEG characterized by generalized slow spike and wave complexes with a slow background. There is no single etiology but it is often diagnosed after an individual has suffered from infantile spasms or a birth injury such as hypoxic-ischemic injuries. The other seizure types noticed in Lennox-Gastaut syndrome include tonic, atonic, and myoclonic. Seizures do not remit over time and the cognitive impairments can worsen. These seizures are most often treated with rufinamide, lamotrigine, valproic acid, felbamate, and clobazam.

Atypical absence seizures also occur in Doose syndrome (myoclonic astatic epilepsy), a rare but often intractable epilepsy that is characterized by the presence of several different seizure types, including atonic or drop attacks, absence seizures, and myoclonic jerks. It typically presents from age 7 months to 8 years, and child development is normal until the seizures begin. Seizures occur many times each day and are associated with EEG findings of bursts of spike and wave complexes or polyspike and wave complexes. Doose syndrome has an unpredictable course and may become intractable. The seizures respond most favorably to treatment with steroids (including high doses of prednisone or adrenocorticotropic hormone) or the ketogenic diet. These children can have a fairly positive outcome (elimination of seizures and improvement in development) if treated properly and promptly.

Another syndrome (this one genetically mediated) in which absence seizures can occur is Dravet syndrome (severe myoclonic epilepsy of infancy), which occurs when there is a mutation on the sodium channel gene *SCN1A* (the same gene defect that occurs in generalized epilepsy with febrile seizures). Seizures begin between ages 2 and 12 months and initially are generalized tonic-clonic or one-sided tonic or tonic-clonic, are prolonged, and can be provoked by fever and illness. Later in life, myoclonic seizures are the most common type, but other types, most commonly atypical absence, also occur. The child, who may have developed normally, begins to exhibit slowed development and the gait is awkward and often unsteady. The EEG may show fast spike and wave or polyspike and wave or can be multifocal. The seizures occurring in Dravet syndrome are treated with a variety of antiepileptic medications such as valproate. Avoidance of lamotrigine, phenytoin, and carbamazepine is key.

> Myoclonic seizures are quick jerks that may be mistaken for a shiver or chill.

Motor Seizures

Myoclonic Seizures

Myoclonic seizures are quick jerks that may be mistaken for a shiver or chill. They last for less than 1 second and may occur repetitively but not rhythmically. They can easily be mistaken for the brief muscle jerking that occurs in children with cerebral palsy and spasticity. They are also found in Doose syndrome, Lennox-Gastaut syndrome, and juvenile absence epilepsy (all of which were described previously).

In addition, myoclonic seizures figure prominently in several epileptic syndromes whose names include the word "myoclonic."

Benign myoclonic epilepsy is rare and more common in boys, and about 40% of the time it is associated with a family history of febrile seizures or epilepsy.[2] Seizures begin between ages 5 months and 5 years, with head drops, eye deviation, and arm jerks. However, the outcome is usually favorable.

Otohara syndrome (early infantile epileptic encephalopathy) is a severe epileptic encephalopathy occurring in early infancy. The EEG shows a distinct "burst-suppression pattern." The term "burst-suppression pattern" is used to describe high-voltage spiking activity on EEG (looking like a mountain) then sudden flattening (looking like a valley) (**Figure 1**). The infant suffers frequent seizures of a variety of types, including tonic, myoclonic, and partial seizures. The syndrome has not been found to respond to any treatment, although some treatment options can slow certain types of seizures. It is associated with severe developmental disability, and these patients require prolonged and involved care and their families need extensive support.

Early myoclonic epilepsy is characterized by myoclonus, usually in combination with other seizure types. The EEG shows a burst-suppression pattern. Infants who have this EEG pattern have a dire outcome. These seizures can be associated with an inborn error of metabolism, cerebral dysgenesis, and hypoxic-ischemic insults.

Epilepsy with myoclonic absences occurs from age 5 months to 12 years, with a peak incidence at age 7 years. Development is normal until seizures begin, and then the child has myoclonic absence seizures lasting from 10-60 seconds, occurring multiple times each day, with myoclonus of the shoulders and limbs. The EEG is typically abnormal during the event; thus, it is important to capture the episode while recording the EEG to confirm the diagnosis.

Juvenile myoclonic epilepsy usually presents in adolescence or young adulthood, with peak onset between ages 14 and 16 years. It is described in detail in

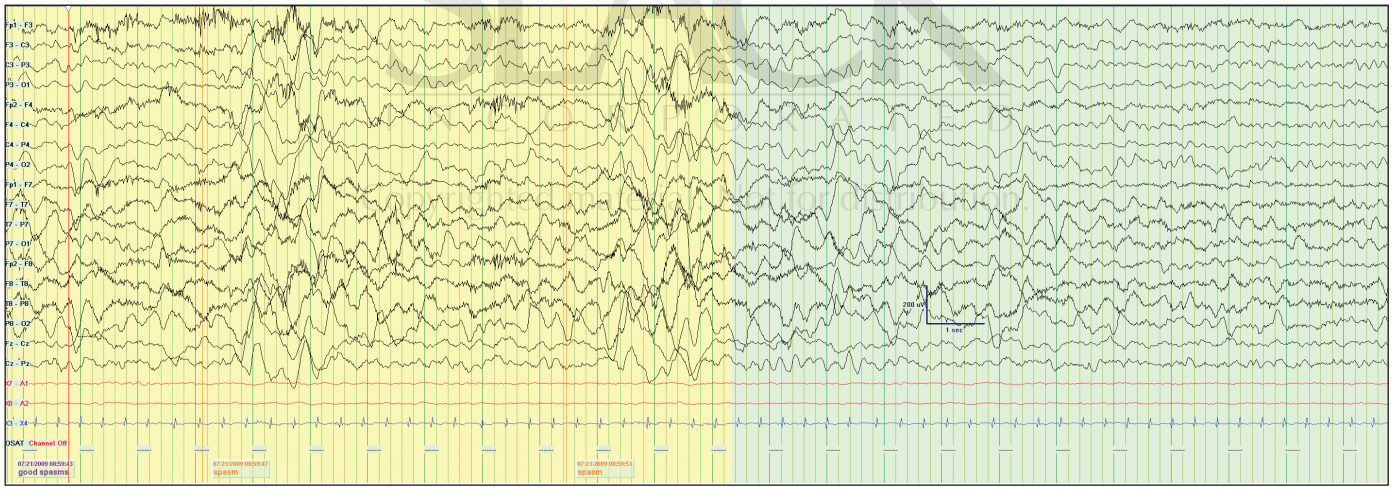

Figure 1. Electroencephalogram (EEG) of a baby with Otohara syndrome, showing the typical burst-suppression pattern. Paroxysms of EEG activity are interspersed with periods of quiescence.

this issue in the article titled "Common Pediatric Epilepsy Syndromes." Myoclonic seizures are also a prominent feature in Dravet syndrome or severe myoclonic epilepsy.

Tonic Seizures

Tonic seizures are stiffening of one or more body parts, occur often at night, and occur frequently in Lennox-Gastaut syndrome. Tonic seizures by themselves are typically a very difficult seizure type to treat. The most commonly used antiepileptic medications for this type of seizure include valproic acid, benzodiazepines, felbamate, levetiracetam, and vigabatrin.

Clonic Seizures

Clonic seizures consist of repetitive jerking involving both sides of the body and occur at regular intervals of less than 1-2 seconds. They occur frequently in Dravet syndrome, juvenile myoclonic epilepsy, and Otohara syndrome.

Tonic-Clonic Seizures

Tonic-clonic seizures alternate between stiffness and shaking, occur exclusively in generalized seizures, and are commonly referred to as grand mal seizures. They occur in Dravet syndrome, juvenile myoclonic epilepsy, and Otohara syndrome, as well as other generalized epilepsies. They may also present with secondary generalization, as the continuation of a seizure that begins focally or with one part of the body.[3]

Versive Seizures

Versive seizures are seizures that begin with sudden, sustained, forceful turning of the eyes and head to one side.[4] These are focal or localization-related seizures. They are usually treated with oxcarbazepine or carbamazepine but sometimes with newer medications such as lacosamide.

Hypermotor Seizures

Hypermotor seizures are large movements that are executed rapidly and appear violent. They are usually short, occur during sleep, and have a brief post-ictal period. Often, they arise from one or both frontal lobes of the brain and are notoriously difficult to control.[5]

Automotor Seizures

Automotor seizures are a form of dialeptic seizures (with impairment of consciousness) with prominent motor movements. The person performs movements that imitate natural movements and are relatively complex, involving different body parts in different planes. These occur particularly in the fingers, hands, tongue, and lips. Again, these are focal seizures and typically treated with oxcarbazepine or carbamazepine but sometimes with newer medications such as lacosamide. Medications for generalized seizures also may be used, and epilepsy surgery can be considered.

Spasms

Spasms are sustained muscle contractions with flexion of the trunk and elevation of the arms, sometimes accompanied by a cry or a head bob, often at sleep transitions and often in clusters. When they occur in infancy they are part of the triad that defines West syndrome, which is also referred to as infantile spasms. This is one of the epileptic encephalopathies of childhood. Onset most frequently occurs between ages 4 and 6 months, and the diagnosis is made by the presence of Salaam seizures (spasms) coupled with behavioral changes (regression in eye contact, use of hands, and loss of tone) and a hypsarrythmic pattern on the EEG (**Figure 2**). About 30% of the children who develop infantile spasms have previously had normal development.[6] These spasms are frequently mistaken for colic and can be missed on a routine EEG. Time is of the essence for diagnosis and treat-

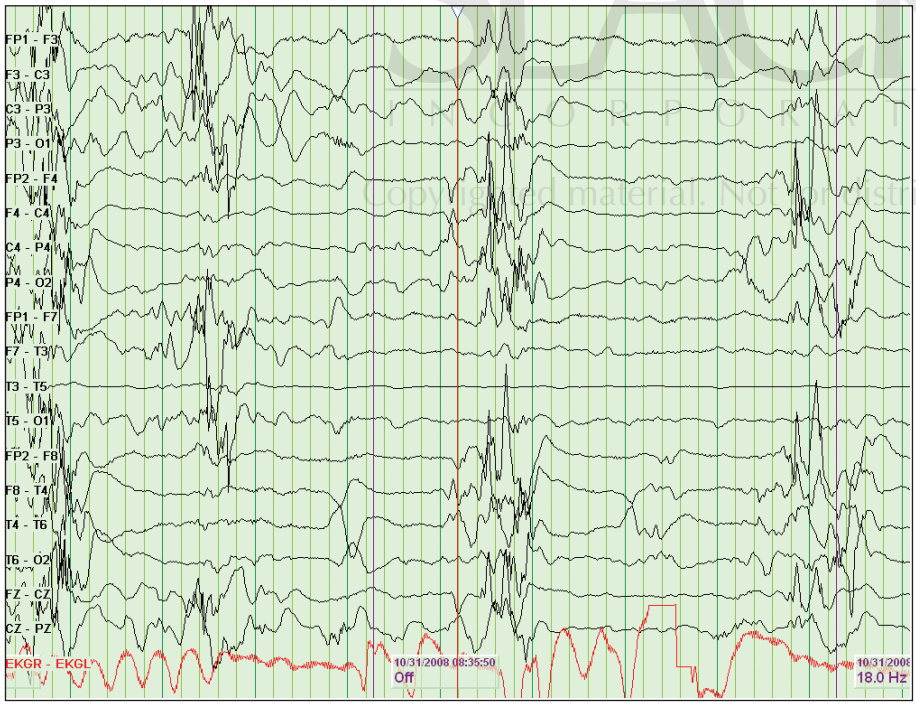

Figure 2. Electroencephalogram depicting hypsarrhythmia in a child with infantile spasms. The high-amplitude chaotic pattern is highly specific to this type of seizure.

ment. The most efficacious treatments are adrenocorticotropic hormone or vigabatrin.[7]

Misleading Symptoms that Are Clues to Seizures

There are many childhood behaviors and events that can mimic seizures. In such cases, a video of the events supplied by the parents is often very useful. Noted in the following text are various seizure types that may be easily missed. They are usually stereotyped and repetitive, and are an odd collection of symptoms that should make you think twice.

Gelastic Seizures

Gelastic seizures are "laughing seizures." This laughter is unusual in its presentation, inappropriate for the situation, and stereotyped. They occur as the result of hypothalamic hamartomas but can also be neocortical in origin. These are often missed. Treatment is resection of the hypothalamic hamartoma, which can be accomplished by a variety of different surgical techniques.[8]

Behavioral Arrests

Inhibitory (negative motor seizures) seizures are also known as hypomotor seizures, in which there is immobility or a marked decrease in movements. These may go unnoticed, particularly in infants. They can be seen in seizures of either generalized or focal onset. If the onset is focal, they most likely arise from the supplementary motor area, a consideration that must kept in mind when performing epilepsy surgery.[9]

Reflex Seizures

Seizures precipitated by certain stimuli include reading epilepsy and praxis-induced seizures. They used to be lumped together under the term "reflex seizures," which is now thought to be misleading. Reading seizures are benign but difficult to control. The clinical seizure is an abnormal sensation or tonic (stiffening) or myoclonic movements of the muscles involved in reading and talking. There is no change in the level of consciousness. Seizures occur after a certain amount of reading and may secondarily become generalized tonic-clonic seizures. There are other rarer, event-induced or praxis seizures that have been triggered by doing math, drawing, playing cards or chess, or even thinking. These usually begin during adolescence and consist of absence seizures, bilateral myoclonus, or generalized tonic-clonic seizures.[10]

Gastrointestinal Symptoms

Sometimes seizures are associated with vomiting and can be mistaken for upset stomach and the flu. Onset is in early childhood and vomiting is followed by head and eye deviation, sometimes progressing to a generalized tonic-clonic seizure. They are common in early-onset benign childhood occipital epilepsy (also known as Panayiotopoulos syndrome), and usually remit after 1-2 years.

Sudden Falls and Syncope

Astatic or atonic seizures occur when there is a sudden loss of tone. If the person is standing, they fall. If the person is seated, there is a (sometimes subtle) head drop or nod. These often result in injury. It is important to differentiate these from motor seizures that can begin with a focal presentation and then proceed to a fall. These are most frequently associated with Lennox-Gastaut syndrome.

Change of Speech

Aphasic seizures are characterized by an interruption in speech or the inability to talk. These seizures arise from Broca's area (anterior language area in the brain) and present as an interference with language output, including speech arrest or slowing of speech. Seizures that interfere with speech present a little differently, often manifested by the inability to name objects or follow two-step commands, or trouble with spontaneous speech. These localize to Wernicke's area (posterior language.)

Unusual Sensations

Sensory seizures are described by patients as tingling, numbness pain, a feeling of heat or cold, or more complex sensory hallucinations. These usually localize to the primary somatosensory area of the brain.

Autonomic Findings

Seizures are classified as autonomic when autonomic signs are the main clinical feature. These autonomic signs include tachycardia, sweating, epigastric sensations, and piloerection.[10]

CONCLUSIONS

Seizures are among the most alarming things a parent will ever observe. Basic knowledge of the semiology and the syndromes will help the clinician decide when to initiate further evaluation and when to be more reassuring. This will also enable one to provide some guidance regarding treatment options.

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