

Psychogenic Nonepileptic Seizures (Pseudoseizures)

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Objectives After completing this article, readers should be able to:

1. Recognize the antecedent stressors associated with psychogenic nonepileptic seizures in children.
2. Identify pediatric nonepileptic seizures clinically.
3. Distinguish psychogenic nonepileptic seizures from epileptic seizures and other paroxysmal nonepileptic events.
4. Be aware of the comprehensive assessment needed to evaluate the child who has possible psychogenic nonepileptic seizures.
5. Become familiar with the management approaches used to treat psychogenic nonepileptic seizures in children.

Definition

Nonepileptic seizures are episodic behavioral events that mimic epileptic seizures but are not associated with abnormal cortical electrical discharges. Psychogenic nonepileptic seizures (PNES) are related to an underlying psychological stressor or conflict and differ from other paroxysmal nonepileptic events. A variety of terms have been used in the literature to describe these events, including hysterical epilepsy, hysteroepilepsy, psychogenic seizures, pseudoepileptic seizures, pseudoseizures, and nonphysiologic or functional seizures. The term psychogenic nonepileptic seizure is preferred because it is nonpejorative and neutral, although there is continuing discussion regarding the most appropriate terminology.

Demographics

PNES are common. Although population data are limited, one report suggested a prevalence of 2 to 33 per 100,000, basing the estimate on an assumption that 10% to 20% of patients seen in an epilepsy center would be found to have PNES. (1) Reviewing video-electroencephalography (EEG) monitoring records, Patel and associates (2) and Wyllie and colleagues (3) found that 3.5% and 7% of children, respectively, seen in clinic for assessment of persistent seizures had PNES. PNES occur in both elementary-age children and in adolescents as well as in all age groups of adults. (2)(3)(4)(5)(6) A female preponderance has been reported but is more pronounced in adolescents than children. (2)(5) Overall, this sex-related trend is more marked among adults, occurring in 75% to 80% of the patients. (6)

It can be difficult to distinguish PNES from epileptic seizures based solely on clinical features. In children, diagnosis often is delayed for 6 months or more. (2)(4) Because epileptic seizures are more common and practitioners are more familiar with them, a large percentage of patients who exhibit PNES are treated initially with antiepileptic drugs. Children seen in the emergency department may be started on antiepileptic drugs by the treating physicians before complete evaluation, and when the diagnosis is unclear, physicians may start antiepileptic drugs because they worry about not giving medication to a child who might have epilepsy.

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The cost of misdiagnosing PNES as epileptic seizures is high from both a financial and psychosocial standpoint. Misdiagnosis can result in medical costs of inappropriate and often costly treatment, unnecessary hospitalizations and emergency department visits, and loss of work as well as increased strain on interpersonal relationships within the family. In addition, patients may be exposed to a variety of iatrogenic complications, such as invasive procedures in prolonged PNES (eg, psychogenic nonepileptic status or pseudostatus epilepticus) and adverse effects from unneeded anti-epileptic drugs. Patients who have both PNES and epilepsy are at risk for invasive monitoring and evaluation for epilepsy surgery if they are misdiagnosed as having refractory epilepsy. Most importantly, misdiagnosis results in patients not receiving much-needed appropriate psychiatric treatment. From a psychiatric standpoint, achieving symptom reduction may be more difficult, treatment of underlying psychological concerns may be delayed, and possibly the dynamics that are responsible for the symptoms may be perpetuated.

Clinical Aspects

Obtaining a thorough history is the critical part of the evaluation. (7)(8) Whenever possible, the episode should be described by an eyewitness. Having the family bring in a video of the events may be very beneficial. Adolescents and children should be asked about possible stressors without their parents present. Key elements that must be explored in the history include:

1. Description of the episode. PNES often are associated with certain physical manifestations that may help differentiate them from epileptic seizures (Table). PNES has been described extensively in adults, (6) but there are fewer reports in children, (2)(3)(4)(5) with the largest study thus far involving 59 patients. (2) These investigators reported a statistically significant difference in the clinical description of spells in young children compared with adolescents. PNES were more commonly manifested as subtle motor activity in children younger than 13 years of age, such as prolonged staring with unresponsiveness, isolated head shaking, eye fluttering, generalized limpness, and behavioral changes or combativeness. On the other hand, in the group of children 13

Table. Differences in Physical Manifestations of Psychogenic Nonepileptic and Epileptic Seizures

Factor	Psychogenic Nonepileptic Seizures	Epileptic Seizures
Duration	Prolonged	Briefer (usually <5 min)
Clinical features during episode	Fluctuating	Stereotypic
Time of day	Usually during wakefulness in the presence of an audience	May occur in sleep whether or not anyone is present
Consciousness	Preserved even with generalized motor activity	Usually altered (exception is supplementary motor area seizures)
Onset	Gradual, with slow escalation in intensity	Abrupt
Head movements	More frequently side-to-side	Usually unilaterally turned, with staring expression
Extremity	Out-of-phase movements, unusual posturing	In-phase movements, rhythmic muscle contractions
Vocalizations	Emotional (crying) in the middle or end of episode	Cry at the onset of episode
Eyes	Closed during the episode	May be open during the episode
Pelvic thrusting	Forward direction	Retrograde direction
Incontinence	Rare	May be present
Related injury	Inconsistent with fall	Consistent with fall
Tongue bite	Occasional (usually at the tip)	Common (at the side)
Postictal change	None or brief, even after prolonged generalized convulsive event	Prolonged, with confusion and exhaustion (although maybe absent after frontal lobe seizures)

years and older, the clinical features were similar to those seen in adults, characterized more commonly by prominent motor activity with generalized arrhythmic jerking or flailing of extremities, side-to-side head movements, and forward pelvic thrusting.

2. Presence of precipitating factors. Do the episodes occur in a specific setting, such as at school or home or in the presence of a specific person? Particular attention should be placed on the timing and setting of the first episode.

3. History of traumatic events or stressors. Several risk factors have been found to be associated with an increased tendency for PNES. (2)(3)(7)(9) In children, the most commonly identified stressors include:

- School difficulties, such as difficulty with learning, (2)(10) poor school performance, stress with school work, change in school environment, and behavioral problems at school with detention or suspensions.
- Family discord, including parental or sibling hostility, parental divorce or separation, domestic physical abuse, and financial stress.
- Interpersonal conflicts with teachers and social difficulties with peers or friends.
- Sexual abuse, often difficult to discover in the initial acute setting, has been reported more frequently in adults who have PNES. (6) A history of sexual abuse is less frequent in children who have PNES, with ranges from 5% to 32%, (2)(3)(9) and further study is necessary to elucidate the true contribution.
- Other stressors include bereavement, prior or concurrent somatic illness, and illness in the family.

4. History of coexistent psychiatric disorders. Many adults who have PNES have depression, anxiety disorders, or borderline personality disorders. (6) Children who have PNES also have more behavioral and emotional problems than children in the general population. Wyllie and associates (3) reported mood disorder in 32% and anxiety disorder in 24% of children exhibiting PNES, and Vincentiis and colleagues (9) reported mood/anxiety problems in 62%. Studies of the presence of severe psychopathology in children who have PNES present conflicting results. The prevalence is higher in adolescents than children. We found comorbid psychopathology in 16% of children younger than 13 years of age and 48.6% of adolescents who had PNES. (2)

5. History of epilepsy and other neurologic illness. A positive history of epilepsy or other neurologic illness should not exclude consideration of a comorbid diagnosis of PNES. Coexisting neurologic illness, includ-

ing cognitive dysfunction such as learning disabilities, (10) headaches, and attention-deficit/hyperactivity disorder (ADHD), (2) have been reported in children who have PNES. Concurrent epilepsy was found in 12% of the affected children seen by Wyllie and colleagues, (3) 14% of patients studied by Kramer and coworkers, (5) and 44% of the children reported by Patel and associates. (2) Nonepileptic status may occur and is mistaken easily for epilepsy in children.

6. A family history of epilepsy (2)(4)(9) and somatization. These findings may serve as a behavioral model for children to shape the expression of their own symptoms.

Differential Diagnosis

Other paroxysmal nonepileptic events are included in the differential diagnosis and can be mistaken for epileptic seizures or PNES. Particularly in younger children, many paroxysmal disorders can mimic seizures. Salient clinical features of some of the more common paroxysmal events can help differentiate them from PNES.

Breath-holding spells occur at a peak age of 6 to 18 months, decreasing in frequency in the second postnatal year. Cyanotic episodes are more common than pallid episodes. These spells always are preceded by brief, vigorous crying for usually less than 15 seconds due to some emotional stimulus such as anger or frustration. The child then becomes silent, holds the breath in expiration, and develops cyanosis. There may be loss of consciousness with a brief period of limpness, followed by opisthotonic posturing, stiffening, or brief clonic movements. Recovery usually is within 1 minute, with a few gasps, followed by return to baseline. Parents should be reassured that these episodes are self-limited.

Self-stimulation is masturbatory behavior that occurs in infants and young children. The episodes are characterized by stereotypic movements, usually involving the lower trunk, such as tightening of the thighs and gluteal muscles and rhythmic pelvic thrusting. These movements may continue for minutes to hours and may be associated with irregular breathing, flushing, and grunting. The episodes may be mistaken for bouts of abdominal pain, partial seizures, or movement disorders such as dystonia, resulting in unnecessary evaluation. The condition is benign and self-limited. Reassurance and counseling should be provided. The term “gratification behavior” may be more helpful because parents are occasionally taken aback by the term “masturbation.”

Gastroesophageal reflux in babies and in children who have disabilities (eg, cerebral palsy) may be associated with back arching that occurs after feedings. Dyspeptic

dystonia, or Sandifer syndrome, is a rare condition that is associated with gastroesophageal reflux or hiatal hernia. The condition occurs commonly in children 4 to 14 years of age, although such children often had feeding problems in infancy. The disorder is characterized by spastic torticollis and dystonic body movements, such as neck extension, head nodding, gurgling sounds, and writhing movements of the limbs. The child may appear quiet during the posturing. These behaviors can be differentiated from seizures by the signs generally appearing after feeding.

Neurocardiogenic or vasovagal syncope commonly is confused with epilepsy. The syncope is precipitated by prolonged standing, change in posture, heat, fatigue, or hunger. The syncopal episode is characterized by a classic prodrome of lightheadedness, blurred vision, pallor, or sweating, followed by loss of consciousness from a few seconds up to 1 or 2 minutes. Convulsive syncope can result from cerebral ischemia and is not indicative of predisposition to epilepsy. Tonic posturing or brief jerking of the extremities and, rarely, even incontinence results from cerebral hypoxia. Following the episode, the patient appears pale and diaphoretic without clearly obvious disorientation. Other causes of syncope include cardiovascular-mediated syncope due to structural or conduction heart defects. If there is any question of an arrhythmia, it is prudent to obtain EEG. Some episodes of cardiovascular-mediated syncope are associated with a Valsalva-like maneuver, such as micturition, trumpet playing, and weight-lifting. Differentiation from epilepsy is based primarily on the clinical features.

Tics are sudden, brief, repetitive, rapid involuntary movements of the face, neck, and shoulders. Motor tics can be simple or complex. Simple motor tics consist of isolated movements, such as head jerking, darting of the eyes, or twitching of the nose. Complex tics are characterized by more coordinated, sequential movements, such as head shaking associated with shoulder shrugging. Tics occur in clusters, are exacerbated by emotional stress and excitement, and are absent during sleep. Efforts to suppress the tics volitionally result in an increasing urge to perform them, with relief after doing so. Most tic behavior terminates spontaneously, although in some cases, the patient may progress into Tourette syndrome. Tics occasionally may be mistaken for myoclonic seizures, which also are characterized by brief isolated jerks of one or more extremities without associated change in responsiveness.

Migraine has several manifestations. Migraine without aura is characterized by severe throbbing headache that is most frequently frontal or temporal; more fre-

quently bilateral; commonly associated with nausea, vomiting, photophobia, and phonophobia; and often relieved by sleep in a dark, quiet place. Migraine headache with aura is preceded by an aura such as blurred vision, brightly colored lights, scotomata, fortification figures, or distortion of body image (Alice in Wonderland syndrome). Other migraine syndromes, such as basilar, confusional, ophthalmoplegic, and hemiplegic migraine, which are associated with neurologic dysfunction and occasionally altered sensorium, now are included in the category of migraine with aura. Childhood "periodic syndromes," such as cyclic vomiting syndrome, abdominal migraine, benign paroxysmal vertigo of childhood, and benign paroxysmal torticollis, are considered precursors of migraine. Further testing, including EEG and neuroimaging studies, may be indicated if structural abnormalities or epilepsy are in the differential diagnosis.

Parasomnias are sleep-related phenomena that may occur during all the stages of sleep, including sleep onset (a rhythmic movement disorder characterized by gratification phenomena such as head banging or body rocking), nonrapid eye movement (REM) sleep (confusional arousals, night terrors, sleepwalking), and REM sleep (nightmares, REM sleep behavior disorder). A good clinical description by the parents and, if possible, a home video of the event help differentiate these episodes from seizures.

Stereotypies are complex, repetitive, rhythmic, seemingly purposeful movements that are suppressible by distraction and do not affect daily activities. They occur in brief clusters, especially when the child is excited or stressed. Stereotypies most frequently involve the arms and head and present as arm flapping, hand rotation, or head nodding. They are seen in children who have normal development and also in children who have autistic spectrum disorder, learning disabilities, and ADHD. Such children usually have a chronic course, with complete resolution of stereotypies occurring in only about 5% by 11 to 12 years of age. Reviewing a home video of the event often helps differentiate these episodes from seizures.

Paroxysmal movement disorders such as paroxysmal dystonic (nonkinesogenic) choreoathetosis and paroxysmal kinesogenic choreoathetosis constitute a group of sudden disturbances of neurologic function associated with a variety of unusual movements such as dystonia, chorea, or athetosis, many of which are related to channelopathies that can be mistaken for seizures. These conditions frequently are inherited. Paroxysmal dystonic choreoathetosis begins in early childhood and is characterized by episodes of dystonia and choreoathetosis in-

volving the face, trunk, and extremities, often associated with dysarthria and dysphagia, that last from minutes to several hours and occur several times a week. The attacks start spontaneously at rest; after alcohol intake; or with stress, hunger, or excitement. This condition responds to clonazepam. The attacks of paroxysmal kinesogenic choreoathetosis are brief (usually <1 or 2 minutes) and consist of dystonic or choreoathetotic movements affecting one or more extremities, occurring several times a day, and usually precipitated by sudden movement after rest. The movements respond to low doses of carbamazepine or phenytoin. Consciousness is retained in both of these disorders.

PNES also should be differentiated from other behavioral nonepileptic disorders. Somatoform disorders, a group of conditions characterized by physical symptoms suggesting a medical disorder but representing a psychiatric condition, also may be considered. Malingering, the willful production of symptoms for gain, and factitious disorder, a need to assume the sick role, are seen predominantly in adults and rarely in children. Seizures are a frequent presenting complaint in factitious disorder by proxy. In this disorder, the caregiver falsely presents symptoms or signs of seizures in a child and assumes the sick role by proxy.

Evaluation

Distinguishing between PNES and epileptic seizures can be one of the more challenging tasks facing the clinician. (6) Accurate diagnosis of PNES requires a high degree of suspicion. PNES should be suspected whenever:

- The events have atypical clinical features, such as occurring only in the presence of an audience or at unusual times. The PNES may be associated with certain stressors, although often the stressors are not readily apparent.
- The events occur frequently despite adequate concentrations of appropriate antiepileptic medication.
- There is a history of repeated hospitalizations or emergency department visits.
- There appears to be a lack of concern about psychosocial stresses in the child's life and an excessive emotional response to the PNES episodes. (8)
- Several routine EEG tracings are normal.

Routine interictal EEG has some limitations. A random normal interictal EEG result neither rule out epilepsy nor confirms the diagnosis of PNES. Similarly, an abnormal interictal EEG tracing with epileptic activity does not by itself confirm epilepsy. Interictal epileptiform activity, such as generalized spike-slow wave discharges,

may be seen in 2% to 3% of asymptomatic individuals. An abnormal EEG result may be seen in patients who have epilepsy, and if these patients also exhibit PNES, difficulty may arise in ascertaining the true nature of any given episode.

Prolonged video-EEG monitoring remains the gold standard for definitively diagnosing PNES. Although this study is performed in an inpatient setting, thus separating the patient from routine environmental and social triggers, it has the benefit of simultaneous video recording and less artifact. Therefore, it is preferable to a 24-hour ambulatory EEG. By extending the recording time, prolonged video-EEG increases the likelihood of capturing the events of concern. A family member or friend who has witnessed the episode should be in the room at all times. If spontaneous spells do not occur, provocative procedures such as suggestion, hyperventilation, and photic stimulation may be used to elicit the event.

Confirmation by the family that the recorded event is the "habitual event of clinical concern" is essential. If multiple types of episodes are reported, all the different episodes should be recorded. A definitive diagnosis of PNES is made only if the recorded event is confirmed to be the habitual event of clinical concern and is not associated with epileptiform activity on EEG. If typical episodes are not recorded, a conclusive diagnosis of PNES becomes more difficult.

Lack of epileptiform changes on EEG during the episode does not indicate conclusively that the episode definitively is PNES. Clinically, frontal lobe seizures often are misdiagnosed as PNES. These spells commonly manifest with bizarre bilateral motor activity that may be clinically mistaken for PNES. In addition, the muscle and movement artifact associated with these seizures may obscure EEG activity, making it difficult to discern any ongoing ictal pattern. Also, some partial seizures associated with deep-seated foci may not produce an ictal pattern on routine surface EEG. For this reason, recording several of the events of clinical concern can be helpful in distinguishing between PNES and difficult-to-diagnose epileptic seizures.

Serum prolactin concentrations rise severalfold 5 to 20 minutes following a generalized tonic-clonic seizure of more than 30 seconds' duration and decline to normal within 1 hour. This peak value can be compared with a sample obtained 90 to 120 minutes later. Patients who experience PNES do not demonstrate the expected rise in serum prolactin values. However, this technique has several limitations. There is a high false-negative rate in patients who have complex partial seizures, in which prolactin concentrations are raised in only 60% of cases,

and the elevation is not as high as is seen following a generalized tonic-clonic seizure. Also, because of the rapidity with which the values peak and decline following a seizure, the timing for blood collection is critical. The data have been studied predominantly in adults, and the value of this testing in children and adolescents is uncertain. Therefore, this method is not used routinely.

Management

It is important to convey the diagnosis of PNES to the patient and family in an appropriate, understanding, and nonjudgmental manner without making them feel that the child is “faking it,” “is crazy,” or that “it is all in his or her mind.” (7)(8) Clinicians should remind families that stress can precipitate a variety of symptoms, such as headaches, increased heart rate, and fatigue, and similarly can cause symptoms and signs of PNES. The video of the episodes should be reviewed with the family to help confirm that the episodes recorded are similar to those witnessed by the family and to educate the family about the episodes that clearly are PNES. Clarity in diagnosis is essential.

If the episodes are referred to as seizures, the parents should be told that the events are not epileptic, ie, “there is no abnormal brain cell firing associated with the events.” Parents should understand that because EEG results are normal during the episode, these events are not epilepsy or caused by brain damage. Although video-EEG does not determine the cause of the episode, it is important that epilepsy be ruled out because antiepileptic drugs are ineffective for PNES.

The clinician should explain that the spells may have emotional causes, of which the patient may not be aware, and that the patient may benefit from assessment and treatment by trained mental health personnel, such as a psychiatrist or psychologist. It is important to maintain physician-patient contact even after the referral to a mental health professional because of the need for a multidisciplinary management approach. (7)

The objective of management is early diagnosis to prevent the episodes from becoming the patient’s primary coping mechanism for dealing with stress. (7)(8) In the outpatient setting, antiepileptic drugs are weaned very slowly, only after psychiatric care has been initiated and the patient and parents have accepted and understand the PNES diagnosis and treatment approaches. The child or adolescent is referred to a psychiatrist or psychologist who is knowledgeable about PNES for evaluation and treatment of the underlying psychopathology.

In a small number of cases that have uncomplicated histories, the PNES stops after explanation and educa-

tion about PNES. Treatment should center on teaching the patient new coping skills with stress management techniques. Good evidence indicates that cognitive behavioral therapy is beneficial for adults who experience PNES. (11) Family involvement is important to manage continued nonepileptic events using a behavioral approach and to ensure the success of the patient’s individual therapy. (7)(8)

Psychopharmacologic agents may be indicated to treat associated psychiatric disorders such as anxiety, depression, or ADHD. Given the important role of learning and social difficulties in triggering PNES related to emotional causes, patients often need psychoeducational testing and intervention. School personnel also need to be guided on how to reintegrate the child into school, address possible learning or social difficulties, and respond to PNES that might occur at school. Children who have social problems also can benefit from social skills training.

Outcome

Overall, children and adolescents who have PNES have a better prognosis than adults. (12) This difference may be related to earlier diagnosis and treatment of the previously described stressors or coping disorders in children. In adults, PNES occurs in the context of more chronic psychological maladjustment, such as chronic depression and personality disorders.

Summary

- Based primarily on consensus due to lack of relevant clinical studies, it is difficult to distinguish PNES from epileptic seizures. Failure to make this distinction may result in lack of appropriate management and a decline in the quality of life for the child and family.
- Based on some research evidence as well as consensus, common associated stressors in children include school difficulties, family discord, and interpersonal conflicts with peers and friends, with sexual abuse being less common than in adults. Anxiety and depression commonly are associated with PNES. (2)(3)(4)
- PNES should be differentiated from other paroxysmal nonepileptic events. Based on strong research evidence, prolonged video-EEG monitoring is the gold standard in making a definitive diagnosis. (5)(6)
- Based on some research evidence as well as consensus, early diagnosis and referral to a psychiatrist or psychologist for treatment of the underlying psychopathology with individual and family therapy are the mainstay of successful management of pediatric NES due to psychological causes. Children who have PNES have a better prognosis than adults. (3)(8)(11)(12)

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