THEME

Epilepsy

The terms ‘convulsion’, ‘fit’ and ‘seizure’ are used interchangeably to describe episodes associated with involuntary motor activity. Not everything that twitches is a seizure and therefore it is important to consider other conditions before diagnosing epilepsy.

Nonepileptic events (NEE) can be due to physiological or exaggerated physiological responses, parasomnias, movement disorders, and behavioural or psychiatric disturbances (Table 1).

Epileptic seizures are ion channel disorders that share a common pathogenesis with other paroxysmal disorders including migraine, movement disorders and periodic paralyses. Seizures can result in positive neurological phenomena such as twitching and sensory symptoms or negative neurological phenomena such as collapse or unresponsiveness. The International League Against Epilepsy (ILAE) classification divides seizures into two broad categories: focal and generalised (Table 2). 1 Focal seizures are associated with activation of a limited number of neurones in one hemisphere and generalised seizures with synchronous activation of neurones in both hemispheres.

An eye witness account is essential to making a correct diagnosis in any child presenting with a possible seizure. Home video recordings are very useful in distinguishing between seizures and NEE. Overdiagnosis of epilepsy is more common than underdiagnosis. Incorrectly diagnosing epilepsy can have adverse effects on a child’s schooling and social life, and may result in iatrogenic side effects of anticonvulsant medications.

Important questions to ask

• Was the episode preceded by prodromal symptoms? Premonitory symptoms such as gradual visual loss, dizziness, and sweating are clues to a fainting episode
• Was the seizure provoked? Acute neurological insults such as fever, head trauma, intracranial infection, intoxication, hypoglycaemia and electrolyte disturbance can provoke seizures in the normal brain
• Were there precipitating factors? Sleep deprivation, stress, illness, flashing lights and fever can precipitate seizures in children with epilepsy
• What was the first thing that happened? An aura,
retained awareness at onset, and twitching beginning in one limb are clues to a focal seizure.

Modes of presentation of seizures include:
• collapse and loss of consciousness
• staring and altered behaviour
• limb movements and postures, and
• nocturnal events.

Episodic collapse and loss of consciousness

Tonic, tonic-clonic and atonic seizures
Sudden loss of consciousness and tonic posturing occur at the onset of generalised tonic-clonic seizures.

This is followed by rhythmic clonic jerking of the limbs, usually lasting less than 3 minutes. There is depression of consciousness in the postictal period.

In contrast, tonic seizures often occur from sleep and tend to be of brief duration; usually 10–15 seconds. They are associated with apnoea and stiffening of all muscle groups, especially the trunk and upper body. The postictal phase is much shorter than with tonic-clonic seizures. Atonic seizures or ‘drop attacks’ are associated with sudden loss of muscle tone and falling. More subtle atonic events can be associated with brief head nods.

Differential diagnoses of afebrile tonic-clonic seizures

Syncope is the most common condition confused with tonic-clonic seizures as it can be associated with anoxic convulsions (Table 3). Important clues to the diagnosis include prodromal symptoms such as light headedness, gradual loss of vision, pallor, nausea, and diaphoresis. Syncopal attacks are brief and are followed by rapid recovery of consciousness. Neurocardiogenic or vasovagal syncope is due to altered autonomic regulation of heart rate and blood pressure. Precipitants include standing for prolonged periods, pain and fear, however, they can also be caused by stretching, particularly in adolescent males.

Cardiac syncope due to structural heart defects such as aortic stenosis or arrhythmias is less common. Syncope during exercise, or while supine, is more suggestive of a cardiac aetiology; a 12 lead electrocardiogram (ECG) is indicated looking for a long QT interval or heart block. Referral to a cardiologist for echocardiogram, Holter monitoring and exercise testing may be warranted, particularly with recurrent attacks. Treatment of syncope includes recognition of presyncopal symptoms, avoidance of precipitants, increased intake of fluids and dietary salt. Beta blockers and fludrocortisone are rarely required.

Breath holding spells can be mistaken for seizures in preschoolers, toddlers and infants. The main clue to diagnosis is that they are provoked by trivial injury or emotional upset. More common cyanotic spells start with crying and progress to apnoea, cyanosis, loss of consciousness and tone. Rarely there is tonic stiffening or clonic jerking. Children rapidly recover consciousness following attacks. Breath holding may be genetically predetermined and can be associated with iron deficiency and therefore respond to iron supplementation. Resolution of attacks usually occurs

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Table 1. Differential diagnosis of epileptic seizures

<table>
<thead>
<tr>
<th>Non-neurological conditions</th>
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<tr>
<td>Normal physiological phenomenon</td>
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<tr>
<td>• Sleep myoclonus</td>
</tr>
<tr>
<td>Exaggerated physiological responses</td>
</tr>
<tr>
<td>• Startle</td>
</tr>
<tr>
<td>• Neurogenic syncope</td>
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<tr>
<td>Behavioural phenomenon</td>
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<tr>
<td>• Infantile shuddering attacks</td>
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<tr>
<td>• Breath holding spells</td>
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<tr>
<td>• Daydreaming</td>
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<tr>
<td>Psychiatric disorders</td>
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<td>• Pseudoseizures</td>
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<td>• Panic attacks</td>
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<tr>
<td>Sleep disorders</td>
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<tr>
<td>• Confusional arousals</td>
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<td>• Sleep talking</td>
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<tr>
<td>Neurological disorders</td>
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<tr>
<td>• Migraine</td>
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<td>• Transient ischaemic attacks</td>
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<td>Paediatric movement disorders</td>
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<td>• Tics</td>
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Table 2. Classification of seizures

<table>
<thead>
<tr>
<th>Generalised</th>
<th>Focal</th>
<th>Absence</th>
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<tbody>
<tr>
<td>Tonic-clonic</td>
<td>Focal sensory</td>
<td>Hemiclonic</td>
</tr>
<tr>
<td>Clonic</td>
<td>Experiential</td>
<td>Secondarily generalised</td>
</tr>
<tr>
<td>Tonic</td>
<td>Clonic</td>
<td></td>
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<tr>
<td>Myoclonic</td>
<td>Asymmetrical tonic</td>
<td></td>
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<tr>
<td>Spasms</td>
<td>With automatisms</td>
<td></td>
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<tr>
<td>Atonic</td>
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<td></td>
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<tr>
<td>Absence</td>
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by 3–4 years of age. Pallid breath holding, also known as reflex anoxic syncope (RAS), is associated with bradycardia and brief asystole with corresponding flattening on electroencephalogram (EEG). Reflex anoxic syncope is sometimes associated with anoxic seizures.

Psychogenic seizures mainly occur in adolescents. Clues to the diagnosis include gradual onset and build up of motor activity with quivering, flailing or rotatory limb movements rather than rhythmic clonic jerking. Other distinguishing features include prolonged attacks with variable features, intermittent staccato movements, lack of oral trauma and lack of postictal sedation. Video EEG monitoring may be required to confirm the diagnosis.

### Episodic staring or altered behaviour

**Absence and complex partial seizures**

Absence seizures are associated with brief episodes of unresponsiveness and behavioural arrest lasting less than 30 seconds. Subtle head nodding, eye flickering and facial twitching are often evident on close inspection. Absence epilepsy has a bimodal distribution with a peak onset at 7 years of age for the childhood form, and 13 years of age for the juvenile form. Absence seizures can be induced by asking the child to perform hyperventilation in the consulting room. An EEG showing bisynchronous 3 Hz spike wave confirms the diagnosis (Figure 1).

Symptomatology in complex partial seizures depends on the site of origin and spread patterns. Seizures arising from the temporal lobe are characterised by behavioural arrest, staring, unresponsiveness, head version, dystonic posturing and oromotor automatisms. A preceding aura is the main clue to diagnosis. Symptoms include epigastric rising sensations, unusual tastes, smells, auditory hallucinations, fear, autonomic, and psychic or experiential phenomenon. Seizures arising from the occipital lobe can be associated with positive visual phenomena such as flashing lights, rotating colours, complex formed hallucinations or negative phenomena such as scotomata.

### Differential diagnoses of episodic staring

Daydreaming is a common phenomenon, particularly in primary school aged children. Staring episodes noted in the classroom but not at home rarely turn out to be absence seizures. Psychogenic unresponsiveness can be seen in adolescents.

### Episodic limb movements and posturing

**Myoclonic and focal clonic seizures**

Myoclonic seizures are cortically generated shock-like involuntary muscle contractions. They can be generalised, focal, multifocal, symmetrical or asymmetrical. The diagnosis is confirmed by an EEG showing high amplitude polyspike slow wave discharges (Figure 2). Juvenile myoclonic epilepsy (JME) is the most
common epilepsy syndrome associated with myoclonic seizures, however they also occur in a wide range of idiopathic and symptomatic generalised epilepsies.

Focal motor seizures cause rhythmic clonic jerking of one limb with preservation of consciousness. They arise from the primary motor strip, located in the precentral gyrus of the posterior frontal lobe. Electrical activity can spread to adjacent areas of motor cortex resulting in a ‘Jacksonian march’ of clonic jerking to other parts of the body.

Differential diagnoses of episodic limb movements and posturing

It is particularly difficult to clinically differentiate seizures from NEE in newborns and infants. Focal clonic jerking is highly likely to be a seizure, but other movements such as cycling, pedaling, eye blinking/deviation, extensor posturing and apnoea are not always epileptic in origin.10 Conditions that mimic seizures in newborns and infants are listed in Table 4.11

Motor tics are a common paediatric movement disorder, affecting approximately 3% of children. They are associated with eye blinking, facial grimacing, shoulder shrugging and neck movements that typically wax and wane in severity over time.12 They are exacerbated by anxiety, excitement and anger.

Consciousness is preserved and children can briefly suppress the movements. Rarer paediatric movement disorders are listed in Table 4.

Episodic nocturnal disturbance

Some forms of epilepsy are strongly associated with sleep (Table 5). Frontal lobe seizures are usually nocturnal. Seizures originating from the anterior or interhemispheric (mesial) frontal regions are associated with bizarre hyperkinetic motor behaviours including kicking and cycling movements of the legs, vocalisation, and complex upper limb automatisms. They often secondarily generalise.

Benign epilepsy with centro-temporal spikes (BECTS) is the most common type of childhood epilepsy. Seizures can be associated with facial twitching, tingling, drooling, inability to speak, and frequently, secondarily generalisation. The EEG shows characteristic unilateral or bilateral epileptic discharges over the low posterior frontal (‘central’) and temporal regions (Figure 3).

Differential diagnoses of nocturnal episodes

Parasomnias are defined as undesirable motor, autonomic or experiential phenomenon that occur exclusively during the sleeping state (Table 5).13 They
Fits, faints and funny turns in children

usually occur during the first half of the night when the child is in slow wave sleep. Confusional arousals are the most common parasomnia, affecting up to 20% of children. They are associated with confused behaviour and slow, nonsensical speech. Children have no memory of the attacks on waking the following morning. Other forms include night terrors and sleepwalking. Treatment strategies include avoidance of triggers such as sleep deprivation and teaching parents not to intervene.

Pharmacological treatment with benzodiazepines and imipramine are rarely required and parents should be reassured they are benign, self limited disorders. Features distinguishing seizures from parasomnias are outlined in Table 6.

Sleep myoclonus is a normal physiological phenomenon that occurs in the transitional state from wakefulness to sleep. It can be associated with prominent multifocal myoclonic jerking, particularly in infants.

Diagnostic investigations in suspected seizures

Biochemistry
Checking serum electrolytes, calcium, magnesium and glucose after a first seizure has a low diagnostic yield except in infants less than 6 months of age.

Electroencephalography
An EEG is usually indicated after a first febrile seizure. However an EEG should not be performed in febrile seizures or to ‘exclude epilepsy’ if the clinical diagnosis is in doubt as epileptiform abnormalities can be identified in a small percentage of normal children.

An EEG will help differentiate between focal and generalised seizures, guide the need for further investigation, and influence decisions to commence anticonvulsant treatment. An abnormal EEG is predictive of recurrence following a first afebrile seizure. For example, an evidence based review showed that 54% of patients with abnormal EEGs had further seizures compared to 25% of those with normal EEGs. A routine EEG has a sensitivity of around 60%, however the diagnostic yield can be increased by replicating the circumstances of the event (eg. sleep recording in suspected nocturnal seizures). Hyperventilation and photic stimulation also activate epileptiform abnormalities in some forms of generalised epilepsy.

Neuroimaging
Magnetic resonance imaging (MRI) is the investigation of choice because of its greater sensitivity in detecting epileptogenic lesions such as developmental brain malformations. It should be performed in children with focal seizures (other than BECTS), focal EEG abnormalities, or in children with associated neurological problems such as motor impairment or intellectual disability.

Conclusion
The circumstances of the attack, precipitants and prodromal symptoms will allow differentiation between seizures and nonepileptic events. Home video recordings of attacks may be useful in clarifying the diagnosis. Attacks provoked by pain or emotional stimuli are unlikely to be epileptic seizures. An EEG is usually indicated after a suspected seizure to confirm the diagnosis.

Conflict of interest: none declared.
References