Diagnosis and management of epilepsy

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Abstract

Epileptic seizures can be generalized or focal and take a variety of different forms: their recognition is important for both diagnosis and targeted treatment. A complete diagnosis of epilepsy involves the diagnosis of seizures (based on the clinical history), the epileptic syndrome and the underlying aetiology. The most common differential diagnoses are syncope and dissociative (psychogenic non-epileptic) seizures. All patients developing seizures should undergo an electrocardiogram, and neuroimaging should be undertaken in all patients in whom a focal cause is suspected. In most patients, seizures can be completely controlled, but antiepileptic medication should be tailored to the patient's specific needs, with particular reference to the seizure type(s) and epilepsy syndrome. Other needs, such as the likelihood of pregnancy and the presence of co-morbidities, including learning disability and psychiatric disorders, must also be considered. Patients not responding to antiepileptic drugs may be candidates for surgery. The need for continuing medication should be regularly reviewed in patients in long-term remission.

Keywords Antiepileptic drug; epilepsy; focal seizure; generalized seizure; seizure; status epilepticus

Key points

- Recognition of the different seizure types is crucial for diagnosis and treatment
- Complete diagnosis includes identifying the epileptic nature of the seizures, epilepsy syndrome and cause
- Investigation for seizures should include electrocardiography
- Neuroimaging should be undertaken for focal seizures of unknown cause
- Ambulatory electroencephalography (EEG) or EEG-videomonitoring can be helpful if the diagnosis of continuing seizures remains unclear
- Medication should be started in line with specialist advice
- Tailor the medication to the patient, seizure type and syndrome
- Consider the possibility of teratogenicity in girls and women of childbearing age
- Be aware of drug interactions, particularly with contraceptives and warfarin
- Consider the possibility of surgery in patients failing to respond to medication
- Consider the possibility of drug withdrawal in people who have been seizure-free for 2 years

Introduction

Epilepsy is defined as the occurrence of two unprovoked seizures more than 24 hours apart. A single seizure where the probability of a second seizure is greater than 60% has recently been added to the definition.[1] Provoked seizures (e.g. febrile seizures in infants) are excluded. The incidence in developed countries is approximately 50 per 100,000 per annum and is highest in infancy and old age. The prevalence is approximately 0.5–1%.

Epilepsy should be regarded as a symptom of an underlying condition, and a complete diagnosis includes confirmation of the epileptic nature of the seizures, the epilepsy syndrome and the aetiology. Epileptic seizures are diagnosed clinically from the descriptions of the patient and an eyewitness (Table 1): videos of seizures and electroencephalograph (EEG)-videotelemetry can also be informative. Familiarity with the various seizure types is important for both differential diagnosis and appropriate management.

Classification

In 2010, the Commission on Classification and Terminology of the International League Against Epilepsy (ILAE) proposed a new scheme for the organization of the epilepsies.[2] This conceptualizes generalized seizures as originating within, and rapidly engaging, *bilaterally distributed* networks. Focal seizures originate in networks *limited to one hemisphere*, although they can evolve to become bilaterally convulsive.

Generalized seizures

The three seizure types most commonly seen in genetic generalized epilepsies, which usually present in childhood or adolescence, are generalized tonic–clonic seizures, absence seizures and myoclonus. Tonic and atonic seizures are commonly seen in early-onset diffuse brain disease, usually associated with learning difficulties.

Generalized tonic–clonic seizures: usually occur without warning; an exception occurs when the seizure is heralded by repeated myoclonus. The patient becomes stiff (tonic phase) and, if standing, falls. Contraction of the jaw muscles can cause tongue-biting and as air is forced out of the lungs, the patient may let out a cry. Respiration ceases, and the patient becomes cyanosed. Shoulder dislocation and vertebral wedge fractures can be caused by the intense muscle spasm.

The tonic phase usually lasts about 20 seconds, followed by clonic movement of the limbs. Salivation is common, and breathing is usually stertorous. The blood pressure and pulse increase, and the pupils dilate. Incontinence can occur. After about 2–3 minutes, the post-ictal phase ensues, lasting up to an hour or two; during this, the patient is confused and drowsy, and

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Questions for an eyewitness when diagnosing possible seizures

- What was the person doing just before the seizure?
- What alerted you to the seizure?
- What time of day did it occur?
- Describe what you saw during the seizure
  - Which parts of the body were affected?
  - Was one side affected more than the other?
  - Did the person go stiff?
  - What type of movements, if any, did the person make? (e.g. twitching, jerking, thrashing)
  - Were there any facial movements?
  - Was the person unconscious or was there a change in awareness?
  - Could they speak to you, and did their speech make sense?
  - Were their eyes open or shut?
  - Did their colour change? Were they flushed, clammy or pale?
  - Did their breathing change? If so, in what way?
  - How long did the seizure last?
  - Did the person injure themself?
  - Did you see anything else that you think might be important?
- How did the person behave after the seizure?
  - Were they sleepy, confused or alert?
  - How long did they take to recover completely?
  - Did they want to sleep afterwards?

Table 1

may complain of headache. Aching of the muscles can persist for days.

Absence seizures: cause the child to appear trance-like and unresponsive. Brief automatisms such as swallowing or fluttering of the eyelids can occur. True absences usually last only seconds, and longer episodes may represent focal dyscognitive seizures.

Myoclonic seizures: are brief muscle jerks that can affect one or several parts of the body. They may be repeated and are often subtle: 'shakiness' or clumsiness in the morning should be specifically sought as it is not always recognized as abnormal.

Tonic seizures: involve a sudden tonic contraction of the limb muscles and axial musculature, usually causing arm elevation and leg extension, which often causes the patient to fall backwards. The seizures typically last less than 30 seconds, with rapid recovery, but often result in injury because of the sudden fall.

Atonic seizures: involve sudden loss of postural tone, such that the patient falls limply to the ground. Although these seizures are typically very brief and recovery rapid, the sudden forward fall often results in severe injury to the face or teeth.

Clonic seizures: involve rhythmic generalized jerking and most commonly occur in neonates or infants.

Focal (formerly partial) seizures

These are defined according to their clinical features. Descriptive terms include 'without impairment of consciousness or awareness' (previously known as simple partial seizures), seizures involving subjective sensory or psychic phenomena, seizures involving loss of consciousness or awareness (focal dyscognitive seizures, previously complex partial seizures) and seizures evolving to bilateral convulsive seizures (formerly secondarily generalized seizures).

Focal seizures without loss of awareness: the symptoms typically reflect the function of the part of the brain in which the seizures arise. Thus, occipital seizures usually manifest as elementary visual symptoms such as blobs of coloured light lasting seconds, and must be distinguished from the much more common visual aura of migraine. Parietal lobe seizures can take the form of spreading paraesthesiae or pain; vertigo can also occur.

Frontal seizures take a variety of forms, including spreading focal motor activity (Jacksonian seizures), a 'fencing posture' or bizarre hyperkinetic motor activity such as cycling and kicking. Frontal seizures often occur as the patient is going off to sleep; they are typically brief, start and stop suddenly and may be repeated, sometimes leading to bilateral convulsive seizures.

Temporal lobe seizures can cause a rising epigastric sensation, *déjà vu* or *jamais vu*, gustatory or olfactory hallucinations, a feeling of fear or panic, and autonomic features such as pallor, tachycardia and a feeling of 'gooseflesh'. Auditory or visual hallucinations (often formed) can occur.

Focal dyscognitive seizures: these usually arise from the temporal lobe. The loss of awareness may be incomplete and may be preceded by the symptoms described above. There may be a motionless stare, followed by oro-facial automatisms such as lip-smacking or swallowing, or manual automatisms such as rubbing or picking. More complex automatic behaviour such as undressing can occur. The seizure typically lasts minutes, and the patient may be confused afterwards.

Bilateral convulsive seizures: it is sometimes impossible to distinguish whether or not there is a focal onset to convulsive seizures, particularly if they occur during sleep. Where the onset is gradual, symptoms attributable to the site of seizure onset can act as a warning ('aura'). Convulsive seizures first developing in adults more than 25 years old are likely to have a focal onset as genetic generalized epilepsies usually declare themselves in childhood or adolescence.

Epileptic syndromes

'Epilepsy' is the term given to a variety of different conditions characterized by the presence of seizures. The epilepsies can be organized into syndromes with particular characteristics, both clinical (e.g. age at onset, seizure type) and electrophysiological. Identifying the syndrome provides a guide to prognosis and appropriate therapy. The 2010 ILAE organization[2] is comprehensive and hence complex, including rare conditions as well as epilepsies occurring exclusively in infancy and childhood.

One important syndrome developing in adolescence is juvenile myoclonic epilepsy, which makes up 3–11% of epilepsy in