An approach to epilepsy in children and adolescents

The syndromal diagnosis of epilepsy, which is based on seizure type, clinical features and EEG data, directs the further investigation of the condition and determines the choice of medication.

ANNIE BYE
MB BS, MD, FRACP

Associate Professor Bye is a Senior Paediatric Neurologist, Sydney Children's Hospital, and Conjoint Associate Professor, School of Women's and Children's Health, University of New South Wales, Randwick, NSW.

The management of epilepsy begins with an analysis of the presenting seizure. The doctor must define the seizure type. Together with all details of the clinical history, including information such as whether the seizure occurred in wakefulness or in sleep, age of patient, previous neurological and developmental history and data from an interictal EEG, the specific epilepsy syndrome can be defined. It is the precise classification of the particular epilepsy syndrome that determines the patient management. Each syndrome has different consequences for the patient and influences choice of drug. This is why the correct syndromal diagnosis is critical. An excellent guide to the syndromal approach is given in Roger and colleagues' book *Epileptic syndromes in infancy, childhood and adolescence*.[1]

According to the International Epilepsy Classification, seizures are generalised or partial (Table 1).[2,3] Seizures and epileptic syndromes may be idiopathic (which have a probable genetic implication) or symptomatic. Below are descriptions of the different seizure semiologies with examples of epilepsy syndromes characterised by these particular seizure types. Recognising the seizure type is the first step to correctly diagnosing the syndrome (see Table 2).

Generalised seizures

Generalised seizures are classified as tonic–clonic, tonic, atonic, myoclonic or absence.

Tonic–clonic seizures

Generalised tonic–clonic seizures are characterised by the following phases:

- tonic phase – brief tonic flexion of axial muscles then longer tonic extension
- intermediatory phase of vibratory tremor
- clonic phase – alternating contraction and atonia (inhibition); expiratory grunts and tongue biting may occur
- brief second tonic phase (tongue biting)
- postictal phase – patient may be unconscious for a period; respiration may be compromised.

Primary generalised tonic–clonic seizures are often state related; for example, they may occur only when the patient is asleep. It is important to recognize early focal features as a partial seizure may secondarily generalise. A primary generalised

IN SUMMARY

- **The precise classification of the particular epilepsy syndrome determines the management of the patient. The seizure semiology, clinical features and EEG data define the syndrome.**
- **Seizures are classified as generalised or partial.**
- **Generalised seizures may be tonic–clonic, tonic, atonic, myoclonic or absence.**
- **Partial seizures are simple, complex or secondarily generalised.**
- **The syndromal diagnosis assists in determining the investigation plan, choice of drug management and prognosis.**

Table 1. Classification of seizure types[1,2]

Generalised seizures
Tonic–clonic seizures
Tonic seizures
Atonic seizures
Myoclonic seizures
Absence seizures
Partial seizures
Simple partial seizures
Complex partial seizures
Partial seizures evolving to generalised seizures

Table 2. Some epileptic syndromes in children and adolescents

Syndrome	Seizure type
Juvenile myoclonic epilepsy	Tonic–clonic, myoclonic, absence
Infantile spasms	Tonic
Lennox–Gastaut syndrome	Tonic, also others
Childhood absence epilepsy	Absence
Juvenile absence epilepsy	Absence, tonic–clonic
Benign rolandic epilepsy	Simple partial, may become secondarily generalised
Mesial temporal lobe	Complex partial

seizure never has a focal onset. The implications of a secondary generalised seizure are very different to a primary generalised event.

The neurophysiological hallmark of the patient with primary generalised epilepsy is a generalised spike and slow wave. Figure 1 shows the EEG of a boy of normal development and examination who has had three generalised tonic–clonic seizures. It is important to note that a normal interictal EEG does not exclude the diagnosis of epilepsy.

Pseudoseizures

An understanding of the sequence of phases in a tonic–clonic seizure helps to distinguish events that may be erroneously categorised as such. An example is a pseudoseizure, which may be characterised by apparent loss of awareness and shaking but on careful history it is clear the sequence of events is very different to that described above.

Tonic seizures

Tonic seizures generally have the following features:

- eyelid opening and upward eye deviation, in subtle forms
- rigidity of posterior neck, paraspinal and abdominal muscles
- limb contraction from tonic activity, which may cause posturing resulting in falls
- loss of consciousness during attacks
- brevity of events, lasting seconds to a minute (average, 10 seconds)
- activation by sleep.

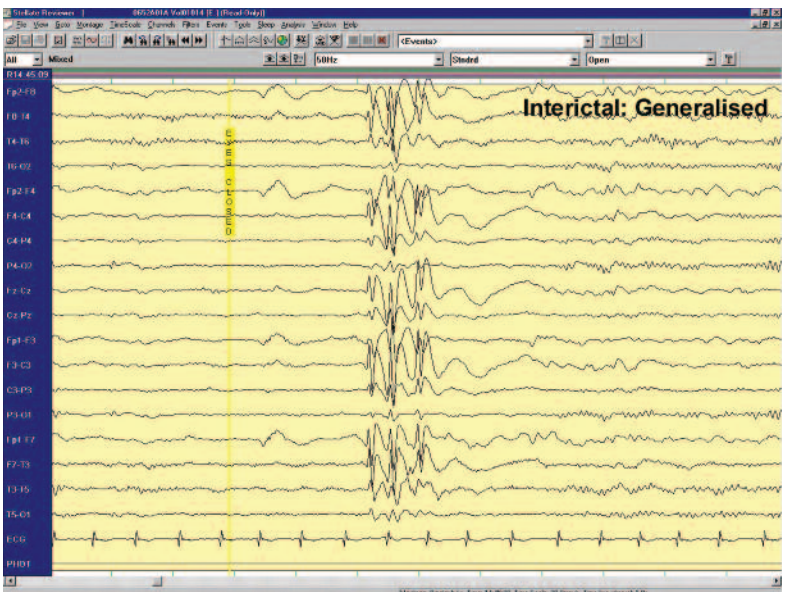

Figure 1. Interictal EEG in primary generalised epilepsy in a boy who has had three generalised tonic–clonic seizures. Note the generalised spike and slow wave.

Infantile spasms and Lennox–Gastaut syndrome

Examples of epilepsy syndromes in which tonic seizures may be a major seizure type are infantile spasms (also called epileptic spasms) and the Lennox–Gastaut syndrome. Both of these epilepsy syndromes are examples of catastrophic infantile and childhood epilepsies frequently associated with developmental regression. Again, the EEG in these syndromes may be extremely helpful. In both of these syndromes the background architecture is very abnormal, with spike and wave activity a major feature.

Atonic seizures

Atonic seizures involve a brief loss of muscle tone that may result in drop attacks. Consciousness is

not usually lost. These seizures can be seen in the symptomatic generalised epilepsies.

Myoclonic seizures

Myoclonic seizures are sudden, involuntary, shock-like muscle contractions, and can be focal or regional. They may be single, repetitive, rhythmic or arrhythmic.

Juvenile myoclonic epilepsy

Juvenile myoclonic epilepsy is a syndrome that presents in the early second decade of life and comprises a triad of seizure types: myoclonic, tonic–clonic and absence. The patient may present with a generalised tonic–clonic seizure in the early hours of the morning. In such a setting, it is important to ask whether the patient experiences myoclonic jerks soon after waking. These involuntary jerks may result in the patient spilling his or her breakfast, or lacerating his or her gum while cleaning teeth. Their presence is a big clinical clue to the diagnosis.

Absence seizures in this syndrome are usually brief. Sleep deprivation and alcohol may precipitate myoclonic seizures or generalised tonic–clonic seizures in juvenile myoclonic epilepsy.

Again in juvenile myoclonic epilepsy the EEG is helpful. The spike and slow wave is generalised, characteristically 5 to 6 Hz, with polyspike. Making a specific syndromal diagnosis is critical as it enables the doctor to give clear information about the particular epilepsy syndrome and to initiate appropriate drug management (for example, in this syndrome sodium valproate is efficacious, while carbamazepine may worsen the myoclonic seizures). Precise diagnosis enables the doctor to speak accurately about prognosis. The syndrome is regarded as lifelong although appropriate drug therapy usually maintains seizure freedom. The syndrome has particular implications for women of childbearing age concerned with pregnancy management and the care of young children.

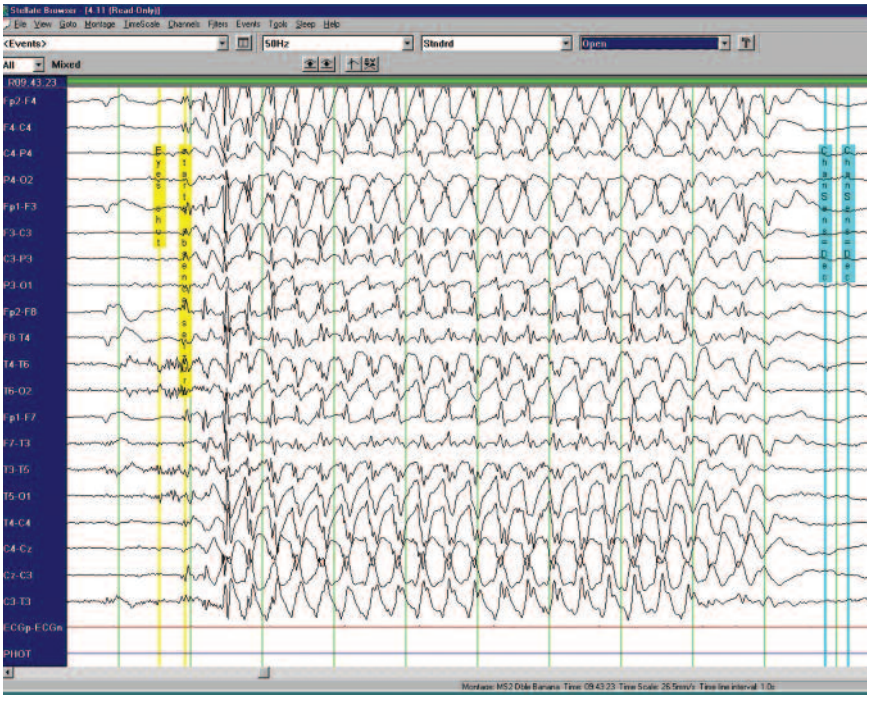

Figure 2. EEG during absence seizure in childhood absence epilepsy. Note the generalised spike and slow wave at 3 Hz.

Absence seizures

Absence seizures are characterised by a brief impairment of consciousness. There may be associated subtle tonic, clonic or atonic components. Automatisms may briefly be seen. Absence seizures are largely confined to the primary generalised epilepsies.

Childhood absence epilepsy

The most common recognised syndrome involving absence seizures is childhood absence epilepsy, which presents between ages 4 and 10 years with a peak incidence at 5 to 7 years. The seizures are brief (four to 20 seconds) and occur frequently throughout the day. They have a clear onset and offset and are associated with impairment of consciousness. Automatisms are frequent. The EEG at the time of the seizure is characteristic with a generalised spike and slow wave at 3 Hz (Figure 2).

Juvenile absence epilepsy and juvenile myoclonic epilepsy

Absence seizures are also seen in other primary generalised syndromes, such as juvenile absence epilepsy and the previously mentioned juvenile myoclonic epilepsy.

Partial seizures

Partial seizures represent the second major subdivision of seizure classification and are classified into three groups:

- simple partial seizures – consciousness preserved
- complex partial seizures – altered awareness and responsiveness
- partial seizures evolving to secondary generalised seizures.

Simple partial seizures

In a simple partial seizure the symptoms and signs arise from a limited area of the cortex. Features may be psychic, motor, autonomic or somatosensory.

Benign rolandic epilepsy

The seizures seen in benign rolandic

epilepsy, the most common school age epilepsy, are usually simple partial, and are predominantly nocturnal. The child or teenager, aged between 3 and 15 years, complains of an unusual sensation in the oral buccal cavity and may seek adult help. There is unilateral tonic or tonic–clonic activity of the face, a speech arrest and excessive pooling of saliva. Consciousness is usually preserved. The seizure may spread to become a hemiclonic seizure or a secondary generalised seizure. The EEG shows a classic spike morphology in the centrotemporal region activated by sleep.

This syndrome has an excellent prognosis for seizure outcome. It illustrates well the importance of seizure analysis and the need to make an accurate syndromal diagnosis for appropriate counselling and management. It is unfortunate if the family are not aware of the good seizure outcome and that the phenomenon is seen only for a few years, with often only one to five seizures occurring.

Complex partial seizures

Complex partial seizures may begin as a simple partial seizure or may have loss of awareness at onset. Characteristic features are an aura, automatisms, postictal confusion or deficit, and amnesia for the episode.

Mesial temporal lobe epilepsy

The seizures seen in the syndrome of mesial temporal lobe epilepsy are frequently complex partial seizures. They are characterised by an epigastric aura at onset. The patient may freeze, stare and demonstrate oral alimentary automatisms and dystonic posturing of an arm. In the postictal phase, the patient may be disorientated, appear confused and, if the dominant temporal lobe is involved, remain aphasic. An interictal EEG may provide supportive evidence of a partial seizure disorder, this time with a spike present in the anterior temporal leads (Figure 3).

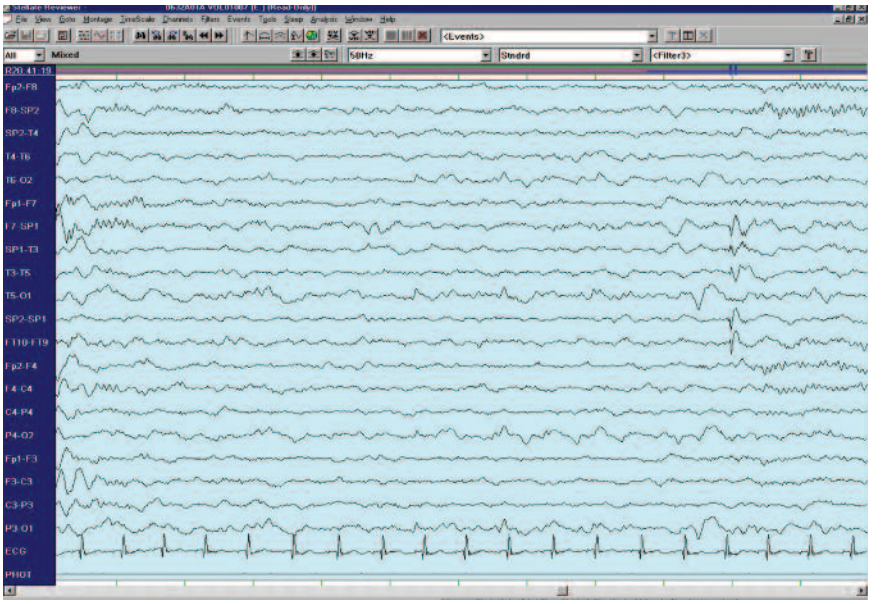

Figure 3. Left anterior temporal spike in mesial temporal lobe epilepsy. Note the left anterior temporal spike.

In clinical practice

The initial step in managing a patient presenting with a fit is to identify the seizure type. As seen above, not all seizures are the same, and old jargon such as 'grand mal' and 'petit mal' is no longer used. Patient demography is then gathered, and the interictal EEG provides further data. All this information is used in syndromal classification. Management is based on this.

Two examples of different presentations of epilepsy and the subsequent syndromal classification are given in the box on page 34.

Syndromal diagnosis assists in investigation and determines what should be done. In primary generalised epilepsies, the yield of magnetic resonance imaging (MRI) is low. In the partial symptomatic epilepsies, it is critical to determine whether a lesion is present. It is important to determine the cause of a new onset partial or symptomatic seizure disorder. For example, in mesial temporal lobe epilepsy the most common MRI finding is hippocampal atrophy and signal change. The MRI is a sophisticated test and it is important that physicians who interpret it are experienced in doing so. Depending on the syndromal diagnosis, metabolic and other tests may be indicated.

The aim of treatment is seizure freedom without unacceptable toxicity. Failure to achieve this is termed medical intractability. The principle of anticonvulsant medication use is the appropriate choice of drug, based on the syndromal diagnosis. In nonemergency situations, it is often beneficial to give an initial low dose of an anticonvulsant and to titrate upwards slowly. It is important to give a well-conducted trial of each anticonvulsant. A drug fails if at appropriate dosage or level, seizures are not controlled. A drug may also fail if unacceptable side effects occur.

Drugs often used in the primary generalised epilepsies are:

- sodium valproate (Epilim, Valpro)
- ethosuximide (Zarontin)
- lamotrigine (Lamictal).

In the partial epilepsies, drugs commonly used are:

- carbamazepine (Tegretol, Teril)
- sodium valproate

continued

Syndromal classification: two cases

There is a variety of specific epilepsy syndromes, all with differing consequences for the patient. Two examples are given here.

Juvenile myoclonic epilepsy

A child of 12 years presents with a tonic–clonic seizure in the early hours of the morning. History reveals myoclonic jerks in the early morning, which are aggravated by sleep deprivation. An interictal EEG shows fast spike and slow wave with polyspike. The doctor appropriately reaches a diagnosis of juvenile myoclonic epilepsy.

Lennox–Gastaut syndrome

A 3-year-old boy presents with frequent falls and blank episodes. The child has bruised his face. His mother comments that his behaviour has deteriorated and he has less mature play. The doctor recognises that the falls are tonic seizures. Regression of development is occurring. The EEG shows a very abnormal record with spike and slow wave. A diagnosis of a symptomatic epilepsy, such as Lennox–Gastaut syndrome, is made.

- lamotrigine
- topiramate (Topamax).

This list is not exhaustive but represents the commonly used drugs. Levetiracetam (Keppra) has recently become available. Its major indication is intractable partial seizure disorders.

Combinations of drugs are used in patients whose epilepsy is difficult to control. A particularly helpful combination is that of sodium valproate and lamotrigine, which have a synergistic effect.

The endpoint of treatment is seizure freedom. It is, however, important to remember that comorbid features are often seen in children and teenagers with epilepsy. These include learning disorders, behavioural issues and a suboptimal quality of life. For adolescents, there are major issues such as drug compliance, driving, career choice, and some restriction of life choices. Pregnancy and contraception require special consideration. Epilepsy associations in each State and Territory and at a national level can offer major support. Accurate information applicable to the particular syndrome places the patient and family in the best position to manage the epilepsy.

Conclusion

The particular epilepsy syndrome the patient has determines the management of the epilepsy. The syndromal diagnosis is made from the seizure semiology, clinical features and EEG data. MT

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