Electroencephalograms in the diagnosis and management of the epilepsies

Introduction

The generic term 'epilepsy' embraces a particular group of disorders of the CNS but by itself is of limited clinical value. Intense clinical and genetic research over the last few decades has identified a large number of well-defined epilepsy syndromes with different clinical, electroencephalograms (EEG), neuropsychological and neuroimaging profiles, natural history and prognosis, conditions that ultimately require different management. There is evidence that early treatment can reduce the risk of seizure recurrence (First Seizure Trial Group, 1993), and its efficacy depends largely on the appropriate drug choice in relation to the particular clinical syndrome. Therefore, a diagnosis of epilepsy is no longer sufficient; identification of the particular form or syndrome is the cornerstone of meaningful, optimal management.

Epilepsy is principally diagnosed via clinical criteria based on patients' own accounts and descriptions by witnesses, but these are often insufficient for characterization of the epilepsy type. The EEG is the single most important test in the diagnosis and management of patients with epilepsies, first because it can support the clinical diagnosis by demonstrating epileptiform activity, and second because it can contribute to the diagnosis of its type or of a particular epilepsy syndrome. Not surprisingly, it is also an essential tool for nosological taxonomy. A study of 300 adult patients with a first unprovoked seizure showed that the EEG increased the accuracy of diagnosis (generalized *vs* focal epilepsies) from 47% (based solely on clinical grounds) to 77% (King et al, 1998).

Equally important is an understanding of what the interictal (in-between seizures) EEG cannot do. Notwithstanding the broad belief to the contrary, the interictal EEG on its own cannot diagnose or exclude epilepsy, and cannot indicate prognosis or the likelihood of seizure relapse after discontinuation of antiepileptic drugs (AEDs).

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International classification of epileptic seizures and epilepsy syndrome

The current classification of epileptic seizures (Commission on Classification and Terminology of the International League Against Epilepsy, 1981) and epilepsies (Commission on Classification and Terminology of the International League Against Epilepsy, 1989) of the International League Against Epilepsy (ILAE) is organized around two dichotomies: first between generalized and localized (partial or focal) disorders of brain function, and second between idiopathic epilepsies (genetically determined, unrelated to any structural brain pathology, and associated with normal neurological and neuropsychological status) and symptomatic epilepsies (those caused by cerebral pathology). Presumed symptomatic epilepsies, in which no certain aetiology can be demonstrated, are sometimes referred to as cryptogenic. The main syndromes within each type are listed (*Table 1*).

Generalized epilepsies are those that manifest with generalized seizures only, and localization-related (also called focal or partial) epilepsies are those that manifest with focal (or partial) seizures. Generalized seizures are those whose first ictal clinical changes reflect involvement of both hemi-

Table 1. Epilepsy types and main syndromes

Generalized	Idiopathic	Benign myoclonic epilepsy in infancy	
		Childhood absence epilepsy	
		Juvenile absence epilepsy	
		Juvenile myoclonic epilepsy	
		Epilepsy with generalized tonic–clonic syndrome only	
		Mixed phenotypes*	
	Symptomatic/cryptogenic	Myoclonic astatic epilepsy†	
		Epilepsy with myoclonic absences†	
		Infantile spasms (West syndrome)†	
		Lennox Gastaut syndrome†	
Focal/multifocal	Idiopathic	Benign rolandic epilepsy	
		Panayiotopoulos syndrome	
		Benign occipital epilepsy (Gastaut type)	
		Photosensitive occipital epilepsy	
	Symptomatic/cryptogenic	Lobar (frontal, temporal, parietal, occipital)	
		Specific syndromes	Rasmussen's encephalitis
			Mesial temporal epilepsy (atrophy)
		Multifocal	

* patients with phenotypes between juvenile absence epilepsy and juvenile myoclonic epilepsy (prominent myoclonic and absence components); † idiopathic forms of these syndromes also exist

spheres and in which initial EEG changes are bilateral (as for example the tonic-clonic seizures and the typical absences) (*Figure 1*).

Focal (or partial) seizures are those in which the initial activation of a group of neurons (the so-called epileptic focus, usually a circumscribed structural change) is limited to a part of one hemisphere (Commission on Classification and Terminology of the International League Against Epilepsy, 1981) (*Table 2*); focal seizures are traditionally classified according to the lobe (frontal, temporal, occipital and parietal) or the anatomical area (e.g. mesial or lateral temporal) in which they reside and from where the seizures arise (also called area of ictal onset) (*Table 2*).

Figure 1. Typical absence seizure in childhood absence epilepsy. Note the abrupt bilateral and synchronous onset and the regularity of the discharge. Impaired consciousness is frequently associated with minor bilateral motor phenomena, such as eyelid movements and automatisms.

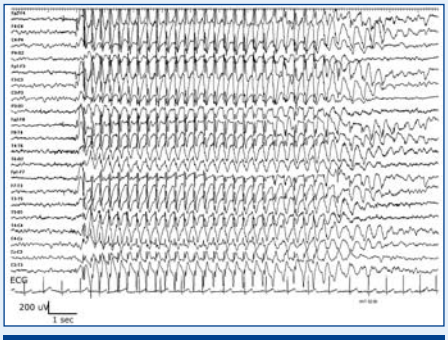


Table 2. Seizure types

Generalized	Tonic-clonic	
	Clonic – tonic-clonic	
	Clonic	
	Absences	Typical (>2.5 Hz)
		Atypical (<2 Hz)
		Myoclonic
	Myoclonic	
	Myoclonic – atonic	
	Negative myoclonic	
	Atonic	
	Spasms	
	Tonic	
Focal or partial	Frontal	Orbitofrontal
		Anterior frontopolar
		Supplementary motor
		Cingulate
		Dorsolateral
	Temporal	Mesial temporal
		Lateral temporal
	Occipital	
	Parietal	

The EEG in different types of epilepsy

Idiopathic generalized epilepsies

The EEG hallmark of the idiopathic generalized epilepsies (IGE) is a generalized spike-and-wave (GSW) discharge that shows an abrupt bilateral and synchronous onset, repeats itself regularly at >2.5 cycles per second, and is of maximal amplitude over the anterior areas. These are seen both interictally (also called subclinical discharges because they are not associated with any clinical accompaniments) and in association with the three main seizure types of IGE, namely typical absences, myoclonic seizures, and generalized tonic-clonic seizures (GTCS).

Typical absences

Typical absences are characterized clinically by impairment of consciousness (the actual absence) that occurs without warning and ceases suddenly without any ensuing (postictal) symptoms, such as confusion or weakness. Electrographically these attacks are accompanied by GSW discharges of ≥2.5 Hz, that terminate without subsequent electrical flattening (*Figure 1*). The term 'typical' distinguishes these seizures from the atypical absences characterized by slower (≤2.5 Hz) discharges seen in symptomatic or cryptogenic generalized epilepsies of children with learning difficulties, such as in Lennox-Gastaut syndrome (*Tables 1* and *2*).

Myoclonic seizures

Myoclonic seizures manifest as sudden, brief, clonic movements of distal, proximal or axial muscles, usually occurring in clear consciousness. The jerks may be symmetrical or asymmetrical, and may occur as single isolated jerks, or may be organized in rhythmic or arrhythmic clusters (sometimes called volleys). They may occur spontaneously, or in response to simple or complex stimuli, including movement, or intention of movement. Myoclonic seizures are the defining seizure type of juvenile myoclonic epilepsy (JME). On the EEG, they are characterized by brief and fast generalized spike wave discharges that may be symmetrical or show variable side emphasis (*Figure 2*).

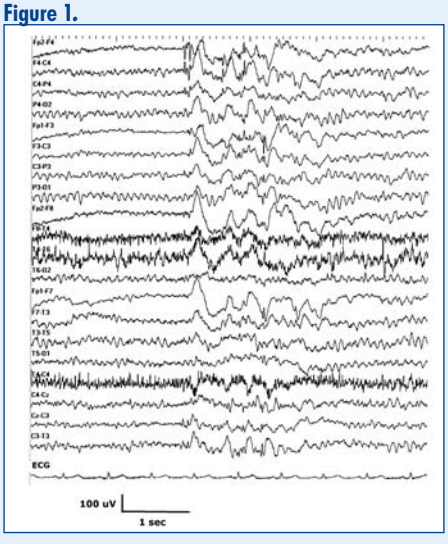
Generalized tonic-clonic seizures

GTCS are usually only recorded by chance during routine awake or sleep recordings. They may occur in isolation or, more frequently, may follow a volley of myoclonic seizures or a cluster of typical absences.

Interictal EEG abnormalities in patients with IGE may include not only GSW discharges but also focal abnormalities that can involve multiple areas and may shift from side to side in different recordings (*Figure 3*) (Aliberti et al, 1994). These focal abnormalities are different to those that characterize focal epilepsies, and for further discussion on these differences readers are referred to the review by Koutroumanidis and Smith (2005).

In addition, the clinical ictal manifestations may include a range of symptoms and signs, such as lateralized or unilateral jerks (myoclonic seizures), rotatory GTCS (in which patients may turn their head or the whole body to one side before the generalized convulsion) and other features that ostensibly suggest focal rather than generalized epilepsies (Lancman et al, 1994). These are some of the arguments against the current simpli-

Figure 2. Generalized polyspike and wave discharge in juvenile myoclonic epilepsy. Note the polyspike component, the irregularity, brevity and the asymmetry of the discharge, and compare with Figure 1.



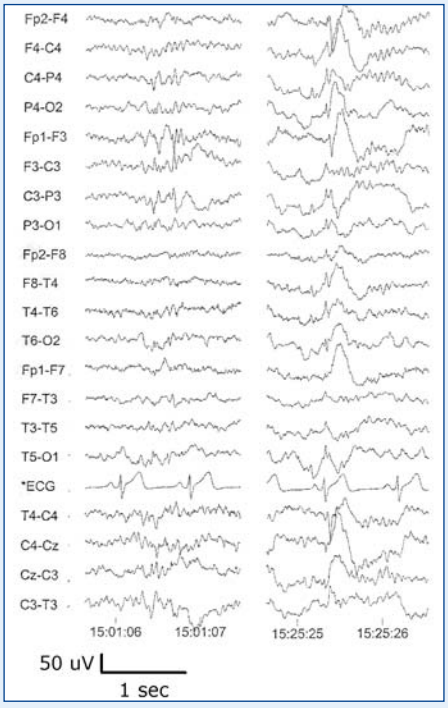
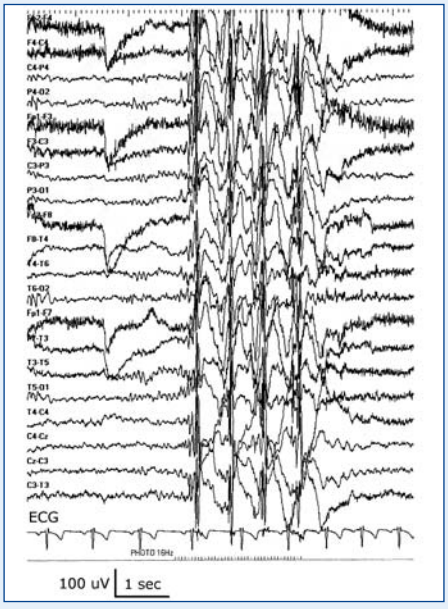


Figure 3. Focal (left frontal on the left trace) and bifrontal (abortive generalized on the right trace) spike-wave discharges during sleep in a 16-year-old girl with juvenile myoclonic epilepsy. Note the fast spike with the low aftercoming slow wave, and the normal regional background in the former (compare with focal discharges in symptomatic temporal and frontal lobe epilepsies in* Figures 6 and 7*).

Figure 4. Electroencephalogram on a 14-year-old girl with juvenile myoclonic epilepsy and photosensitivity. A generalized photoparoxysmal response was induced by intermittent photic stimulation at 16 Hz. It consists of rhythmic generalized polyspike and wave discharges, associated with myoclonic jerks of the body and arms.



fied dichotomy of the ILAE classification for seizures and syndromes (Commission on Classification and Terminology of the International League Against Epilepsy, 1981, 1989), but further discussion of these issues is beyond the scope of this article, and readers are referred to a more comprehensive review (Koutroumanidis and Smith, 2005).

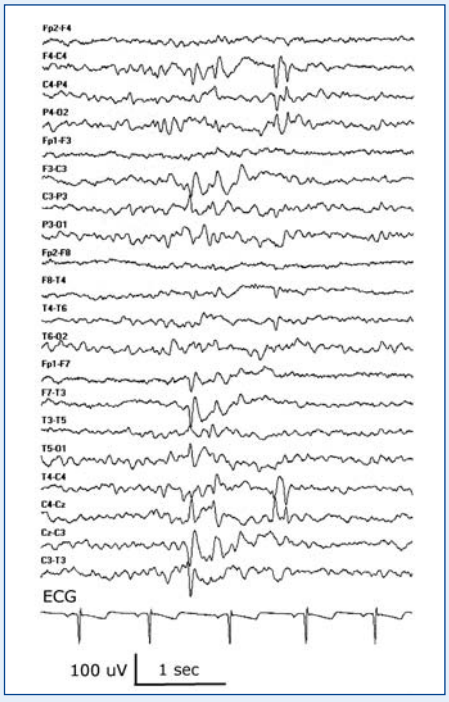
The IGEs are also sleep-sensitive epilepsies: GSW discharges are activated during drowsiness and light sleep and practically disappear during rapid eye movement (active) sleep. Sleep deprivation seems to activate GSW discharges independently (Koutroumanidis and Smith, 2005).

Epileptiform activity and seizures may also be provoked by hyperventilation (especially typical absences) and specific triggers (Koutroumanidis and Panayiotopoulos, 2004) (i.e. photic or pattern stimulation (*Figure 4*), video games, thinking, reading and language-induced activities).

Idiopathic focal epilepsies

These epilepsies are represented mainly by rolandic, autonomic and occipital seizures and, as a rule, the prognosis is excellent.

Figure 5. Bilateral independent functional spikes in benign rolandic epilepsy. Regional background rhythms remain normal, and the aftercoming slow wave is small (compare with focal discharges in symptomatic temporal and frontal lobe epilepsies in* Figures 6 and 7*).



The EEG is characterized by focal spikes or spike-wave complexes in the central or centro-temporal region of one or both hemispheres, or by multifocal spikes of similar morphology in occipital and other areas. There is no disturbance of the regional background activity (*Figure 5*).

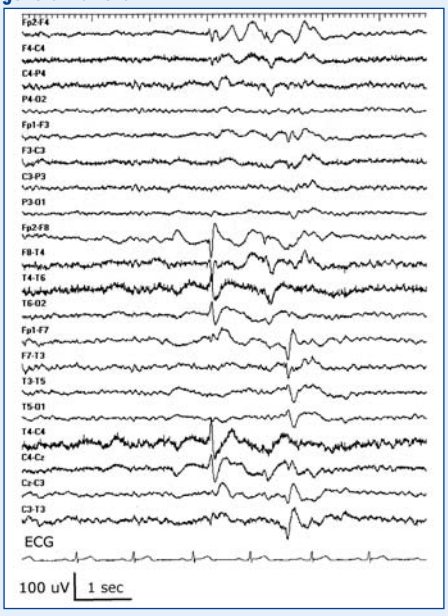
The EEG findings are fundamental to the diagnosis of these benign conditions. Often the spikes occur many times per minute but their frequency typically increases during sleep, and thus a sleep recording may be advisable if the diagnosis is suspected on clinical grounds. Another reason for undertaking a sleep recording is that some children may display almost continuous GSW activity during slow wave sleep.

Symptomatic and cryptogenic focal epilepsies

These are caused by a localized structural abnormality of the brain and ictal (seizure) clinical manifestations depend on the topography of the focus and the propagation of the ictal discharge.

EEG findings are focal and usually correspond well with the localization of the epileptic focus (*Figures 6–8*). Interictal spikes are usually followed by a large aftercoming slow wave, and the background activity

Figure 6. Bilateral independent spike wave foci in symptomatic temporal lobe epilepsy. This patient has magnetic resonance imaging evidence of bilateral mesial temporal atrophy, and presents with complex partial seizures with secondary generalization.



around the focus is usually disturbed, as opposed to idiopathic focal epilepsies in which background rhythms around focal spikes are normal. They are frequently associated with rhythmic or arrhythmic delta activity, which is regarded as a marker of focal epilepsy (Koutroumanidis et al, 2004) (*Figure 9*). These interictal abnormalities are activated by sleep.

GSW discharges are sometimes seen in focal epilepsies. They may either follow focal discharges (a phenomenon called secondary bilateral synchrony) (*Figure 9*) or, infrequently, may occur on their own, thus mimicking IGE (see below). Such findings are rare (Koutroumanidis and Smith, 2005).

Seizures arising in the temporal lobe can originate in mesial regions or more lateral neocortical areas of the temporal lobe and the EEG findings often localize appropriately.

Owing to the large volume of the frontal lobe and the fast propagation of the epileptic discharges, seizures arising from here are more difficult to localize by means of scalp-EEG. Indeed, ictal EEG may even be 'normal', and because of their frequently bizarre, hypermotor (excessive non-rhythmic motor activity, such as arm or leg flailing movements) manifestations, frontal lobe seizures may be misdiagnosed as non-epileptic phenomena.

When limited to brain areas far from the midline structures that are involved in consciousness, focal seizures may not be associated with impaired awareness (and are called simple partial seizures); frontal and occipital seizures that do not propagate outside the lobe of origin are good examples.

When the seizure discharge invades both hemispheres, seizures are accompanied by clouding of consciousness (and are called for this reason complex partial seizures), or evolve into secondary GTCS. It is important to note that very circumscribed interictal or ictal focal epileptic activities may remain undetected by scalp EEG recording, and a typical example of this type is a focal motor seizure that only involves a small group of muscles such as in Rasmussen's syndrome. As with some frontal lobe seizures, it is worth noting that the lack of associated ictal EEG change does not necessarily rule out the diagnosis of focal epilepsy.

Figure 7. Post-traumatic (symptomatic) right frontal epilepsy.

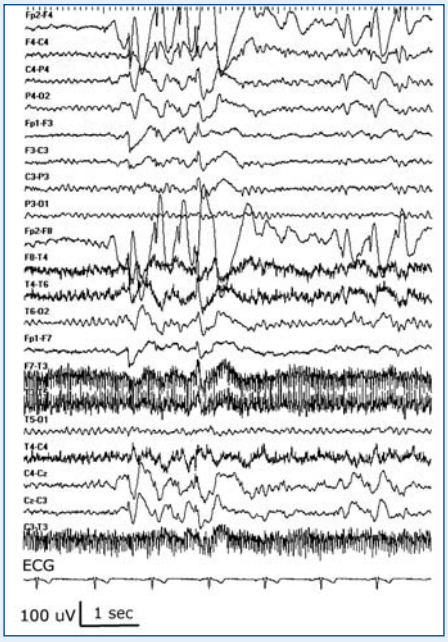


Figure 8. Right occipital spike discharges in a young woman with right occipital cortical malformation.

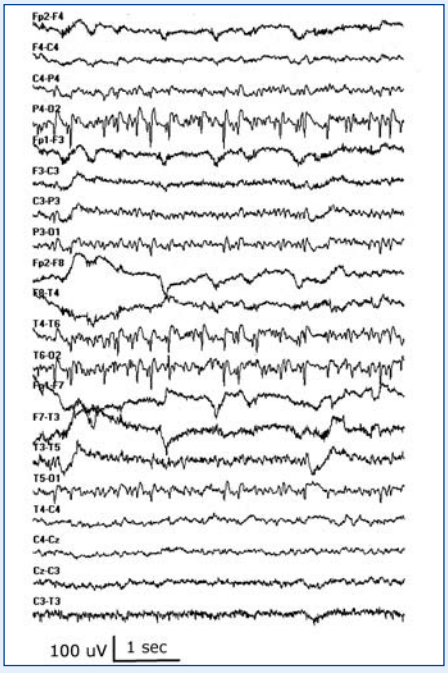
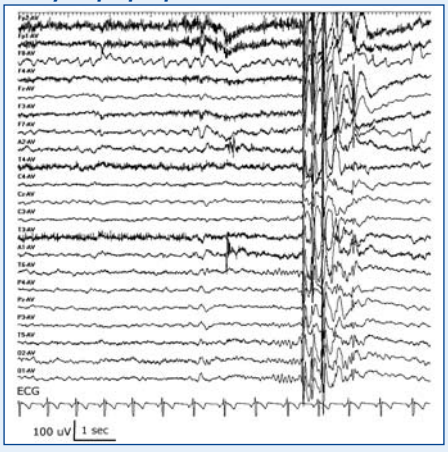


Figure 9. Runs of irregular slow activity over the right anterior to mid temporal area (F8 and T4 electrodes) triggering a high voltage bilateral and synchronous polyspike and wave discharge (secondary bilateral synchrony). This focal slow activity is epileptic phenomenon.



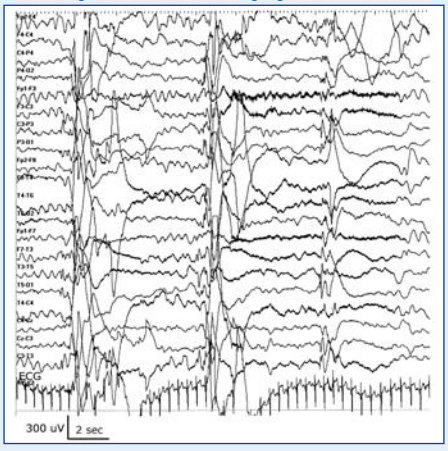
Symptomatic or cryptogenic generalized epilepsies

This is a group of epilepsies and syndromes (*Table 1*) that most often start during infancy and childhood, and manifest with generalized seizures whose clinical and EEG features are different from those in IGE. These seizures include atypical absences, spasms (*Figure 10*), myoclonic, tonic and atonic seizures, and frequently focal seizures. The background EEG activity and brain imaging are typically diffusely or multifocally abnormal.

Potential diagnostic uncertainties

Misinterpretation of IGE for focal epilepsies and vice versa is possible and may seriously affect patient treatment and management, clinical and genetic research, and AED. From the clinical viewpoint, absences with automatisms may mimic complex partial seizures, asymmetric myo-

Figure 10. Epileptic spasms: high voltage diffuse slow waves are followed by brief periods of diffuse low voltage fast activities. Epileptic spasms usually occur in children with severe symptomatic or cryptogenic generalized epilepsies, but may also be idiopathic. This example is from a 16-month-old boy with idiopathic flexor spasms that responded to treatment. Brain magnetic resonance imaging is normal.



clonic seizures may resemble focal motor seizures, and absence status epilepticus (a very prolonged absence seizure that may last for hours, days and even weeks) may sound (and behave) like complex partial status epilepticus.

Misdiagnosis may be further compounded by the presence either of asymmetric GSW discharges and focal spikes in patients with IGE (*Figure 3*), or by the apparently symmetric and regular GSW that may occasionally occur in symptomatic focal epilepsies. For further discussion see review by Koutroumanidis and Smith (2005).

Conclusions

The EEG must always be interpreted in the context of the patient's history and clinical characteristics of the patient's seizures. In isolation, the EEG is of limited help and sometimes it may even be misleading. There are no golden rules for EEG diagnosis; the diagnosis of the epilepsy type should always be based on the whole electroclinical picture of the individual patient. **BJHM**

Conflict of interest: none.

KEY POINTS

- The electroencephalogram (EEG) can support the clinical diagnosis of epilepsy and identify possible triggers, refine the diagnosis by suggesting a specific epilepsy type or syndrome, help with the selection of the optimal antiepileptic treatment and assist with the definition of prognosis.
- The EEG on its own cannot diagnose or exclude epilepsy. Full clinical information is mandatory for optimal interpretation of the EEG.

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