Electroencephalography and video-electroencephalography in the classification of childhood epilepsy syndromes

C D Ferrie MRCP[1] A Agathonikou MD[2] C P Panayiotopoulos PhD FRCP[2]

J R Soc Med 1998;**91**:251–259 JOINT US–UK CONFERENCE ON PAEDIATRIC EPILEPSY, 26–27 JULY 1996

The electroencephalogram (EEG) is the single most useful investigation in children with seizure disorders but many clinicians use it only to help confirm their clinical suspicion of 'epilepsy'. This is misguided because epilepsy is a clinical diagnosis, because a normal EEG is frequently recorded in those with seizure disorders and because paroxysmal EEG changes occur in those who have never had a seizure. Other clinicians use it in monitoring response to therapy and as a guide to the discontinuation of antiepileptic drugs, although in this respect its role is limited. Its usefulness in the localization of epileptogenic zones in patients being considered for epilepsy surgery is unquestioned but this is a specialized area of little interest to most paediatricians. The EEG is probably at its most helpful in the categorization of epilepsies, including their syndromic classification. In this paper we illustrate the clinical utility of the syndromic approach and describe how the EEG can help the paediatrician classify a child's epilepsy.

EPILEPSY AND EPILEPSY SYNDROMES

The term epilepsy is becoming redundant: it is imprecise, it is often misleading, it has no prognostic value, it may lead to inappropriate treatment and it causes prejudice and discrimination. The epilepsies are probably hundreds of different conditions. The syndromic approach has allowed far greater accuracy in predicting prognosis than any previous classification and offers a rational guide to therapy.

Take, for example, a 9-year-old boy who is referred after a couple of nocturnal convulsions. He has also had attacks in the day during which he is unable to speak, 'twitches around the mouth' and then becomes vacant and unresponsive for 20–30 seconds. EEG reveals frequent high-amplitude sharp and slow wave abnormalities in the central and mid-temporal regions. Previous diagnoses might have been 'grand mal and petit mal' or 'psychomotor seizures'. A modern seizure classification would be 'simple partial seizures with motor phenomena evolving to complex partial seizures along with probable secondarily generalized seizures'. None of these allow accurate prognostication; at best the outlook would be guarded, with many doctors painting a gloomy picture based on their perception of the likelihood of a temporal lobe focus. Aggressive treatment with antiepileptic medication would then be appropriate, because of the notion that such children whose seizures are controlled quickly may be at less risk of developing 'chronic epilepsy'. In fact this child had benign childhood epilepsy with centro-temporal spikes (also known as rolandic epilepsy)[1]. The parents were told that his seizures would probably abate within a couple of years and would certainly stop before he left school. They decided against antiepileptic drug medication in the knowledge that this does not affect the ultimate outcome and that 'aggressive' treatment probably does more harm than good.

Another example is a girl age 14 years who has had a convulsion shortly after awakening on the day of an examination. The history reveals that for many years she has had brief 'blank spells', interpreted as daydreaming by her parents, and more recently has been 'clumsy' in the morning. Following an EEG which showed short, generalized, frontally predominant discharges of multiple spike and slow wave she was confirmed as having juvenile myoclonic epilepsy with typical absence seizures, myoclonic jerks and generalized tonic clonic seizures[2,3]. Treatment with sodium valproate is wholly successful in controlling her seizures. She is advised that, although she may never have another convulsive seizure in her life, her susceptibility is longlasting and medication should be continued well into adult life. She

Key to abbreviations
BECTS=Benign epilepsy with centro-temporal spikes
CAE=Childhood absence epilepsy
CEOP=Childhood epilepsy with occipital spasms
EBOSS=Early onset benign occipital seizure susceptibility
EMA=Eyelid myoclonia with absences
JAE=Juvenile absence epilepsy
JME=Juvenile myoclonic epilepsy
PMA=Perioral myoclonia with absences

[1]Department of Paediatric Neurology, Leeds General Infirmary, [2]Department of Clinical Neurophysiology and Epilepsies, St Thomas' Hospital, London, UK

Correspondence to: Dr C D Ferrie, Department of Paediatric Neurology, Clarendon Wing, Leeds General Infirmary, Leeds, LS2 9NS, UK

is warned not to switch medication during pregnancy but is advised on how the risk of teratogenicity may be reduced. Alternative diagnoses such as 'grand mal', 'grand mal and petit mal', 'centrencephalic' epilepsy or 'primary generalized' epilepsy could not have allowed such a clear prognosis or the syndrome-specific advice on avoidance of precipitating factors.

The current international classification of the epilepsies[4] starts with a division into whether seizures arise from a localized area of the cortex (*location-related or partial epilepsies*) or diffusely from the whole or the greater part of the cortex from the outset (*generalized epilepsies*). Further categories exist to accommodate cases in which partial and generalized seizures coexist or in which it is not clear whether the seizures are partial or generalized (*epilepsies undetermined whether focal or generalized*) and seizure disorders arising in special situations, such as febrile convulsions (*special syndromes*). The partial and generalized epilepsies are further divided according to whether a cause is known (*symptomatic*), unknown but suspected usually because of neurodevelopmental problems predating onset of seizure (*cryptogenic*), or unknown without suspicion of an underlying cause, and probably genetic in origin (*idiopathic*). Cryptogenic and symptomatic epilepsies will be considered together here. Although it may not be possible to offer a specific syndrome diagnosis one can categorize the patient broadly as to whether the epilepsy is idiopathic generalized, idiopathic partial, symptomatic/cryptogenic generalized or symptomatic/cryptogenic partial.

THE EEG IN SYNDROMIC DIAGNOSIS

An epilepsy syndrome is defined by the non-fortuitous clustering of symptoms, signs and investigational results. The EEG is often central in the classification of a child's epilepsy and the electroencephalographer may well be the first to suggest a particular syndrome. However, whilst it is rarely possible to be certain of a syndromic diagnosis without EEG confirmation, no epilepsy syndrome can be diagnosed purely by electroencephalography. In this discussion we give the broad picture, emphasizing the idiopathic epilepsies since it is in these that the EEG is likely to be most useful.

Idiopathic generalized epilepsies

Beyond the neonatal period, the idiopathic generalized epilepsies are characterized by generalized tonic–clonic seizures, typical absences and myoclonic jerks alone or in combination. The latter two 'minor' seizure types are more important for classification. The international classification recognizes the following idiopathic generalized childhood epilepsies: benign myoclonic epilepsy; childhood absence epilepsy (CAE); juvenile absence epilepsy (JAE); and juvenile myoclonic epilepsy (JME)[4]. The first of these is rare and will not be considered further. In addition to these we shall discuss other proposed syndromes seen in childhood—eyelid myoclonia with absences (EMA)[5–7] and perioral myoclonia with absences (PMA)[8,9].

In the idiopathic generalized epilepsies the EEG background should be normal for the child's age. However, certain antiepileptic drugs, including carbamazepine and phenytoin, which like vigabatrin are inappropriate treatments in these epilepsies, can cause diffuse slowing of the EEG whilst others such as benzodiazepines and barbiturates can cause excess fast activity[10]. Furthermore, in CAE prolonged runs of rhythmic high-amplitude delta waves at around 3 Hz may occur posteriorly (Figure 1), and seem to be a good prognostic sign[11]. Another feature of these epilepsies is that paroxysmal abnormalities are generalized. Again, however, there are exceptions, most notably JME in which focal paroxysmal abnormalities occur in up to one-third of patients (Figure 1)[12]. Such focal abnormalities tend not to persist in one place and are not associated with focal EEG background abnormalities.

The major paroxysmal abnormality associated with the idiopathic generalized epilepsies is the 3 Hz spike and wave discharge[13]. This is seen in its most classical form in CAE and consists of repeating complexes of a spike (or spikes) and an aftercoming slow wave. Such discharges are generalized, bilaterally synchronous and usually have a frontal maximum. They are often associated with impairment of consciousness (i.e. a typical absence seizure) but may be subclinical. Therefore, such discharges can be considered as both ictal and interictal. Separation on the basis of the discharge duration (<3 seconds often being considered as interictal) has little rational basis; if one seeks subtle clinical events, including mild impairment of consciousness, during such discharges they will often be found[14]. The discharges are readily provoked by hyperventilation, and in children with idiopathic generalized epilepsies who are not on medication and who have adequately hyperventilated it is exceptional *not* to record paroxysmal EEG abnormalities. The occurrence of generalised discharges is also greatly influenced by the sleep–wake cycle. Recording during light sleep (we find partial sleep deprivation the most useful method of inducing this in children) and particularly on awakening will often precipitate them. In addition, intermittent photic stimulation is a powerful activator of EEG abnormalities in some idiopathic generalized epilepsies of childhood. EEG may prove particularly useful in demonstrating photosensitivity in children with TV or video-game induced seizures.

There is strong evidence that both the details of the EEG spike and wave discharge and the accompanying clinical events, particularly typical absence seizures, are syndrome

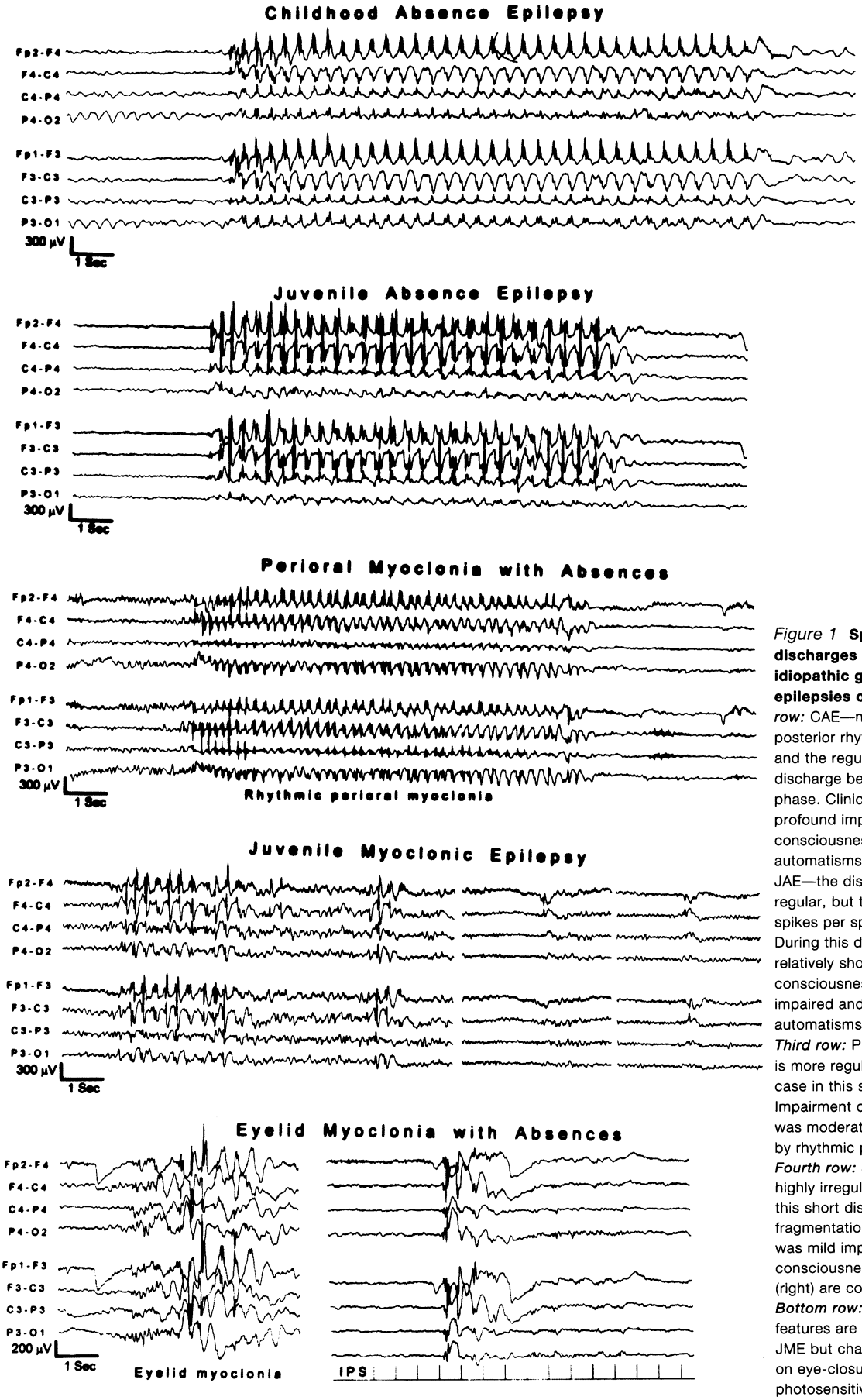

Figure 1 **Spike wave discharges in various idiopathic generalized epilepsies of childhood.** ***Top row:*** CAE—note the preceding posterior rhythmic delta activity and the regulatory of the discharge beyond the opening phase. Clinically there was profound impairment of consciousness and simple automatisms. ***Second row:*** JAE—the discharge is still fairly regular, but there are multiple spikes per spike-wave complex. During this discharge, which is relatively short for JAE, consciousness was severely impaired and simple automatisms occurred. ***Third row:*** PMA—this discharge is more regular than is often the case in this syndrome. Impairment of consciousness was moderate and accompanied by rhythmic perioral myoclonia. ***Fourth row:*** JME—note the highly irregular morphology of this short discharge and the fragmentation. Clinically there was mild impairment of consciousness. Focal features (right) are common in JME. ***Bottom row:*** EMA—the EEG features are similar to those of JME but characteristically occur on eye-closure. Patient is highly photosensitive (right)

related[15–17]. Video-EEG recordings and detailed analysis of spike wave discharges are therefore prerequisites for full evaluation. Different phases of the spike and wave discharge have been defined—opening (1st second), initial (the following 3 seconds) and terminal (the last 3 seconds)[15–17]. The opening phase is often highly irregular and its diagnostic value is limited. The duration, frequency and morphology of the remainder of the discharge should be assessed. In general, the frequency is fastest in the initial phase and slowest during the terminal phase; slowing may be gradual and regular, or there may be abrupt changes in frequency. Spike wave discharges differ in the number of spikes per spike wave complex (single, double or multiple), their position in relation to the aftercoming slow wave (constant or varying) and the amplitude of the spikes (constant or varying). Multiple spikes may cause the 'w' appearance seen particularly in JME. Finally, discharges may be continuous or contain fragmentations (Figure 1).

Standard EEG performed by a competent technician allows only a basic evaluation of consciousness during discharges, and the association of clinical events with EEG changes may be doubted. Video-EEG, with its facility for replay and precise timing of clinical and EEG events, allows far superior analysis. Amongst the most useful events detected are the sudden jolts of limbs constituting myoclonic jerks (it is surprising how often a history of these is not obtained clinically despite detailed questioning), rhythmic myoclonic movements of the face, particularly of the eyes/eyelids and around the mouth, the timing of events such as eye opening and the occurrence of automatisms. Impairment of consciousness is manifested by interruption of on going activities and by reduced responsiveness to external stimuli. Breath counting during at least two sessions of hyperventilation is useful for evaluating the former; it simultaneously tests attention, concentration, memory, and numerical and language function. Subtle impairment of consciousness is manifested by interruption, hesitation, repetition or stuttering[18,19]. Response to external stimuli is evaluated by manoeuvres such as giving names/numbers to patients during discharges and asking them to recall them after the discharge is over, and also by calling to or touching the patient during the discharge and monitoring response. Impairment of consciousness can then be classed as profound (awareness, perception, responsiveness, memory and recollection deeply disturbed), or moderate or mild (detected only by minor errors such as hesitations during breath counting, for example).

In Table 1 we detail the EEG and video-EEG features that help in syndromic classification of CAE, JAE, JME, EMA and PMA[4–7]. The definitions used for CAE, JAE and JME are ones proposed by Panayiotopoulos and others, which make use of clinical and EEG exclusion as well as inclusion criteria[20–24]; they are stricter than the current definitions proposed by the International League against Epilepsy[4]. Representative EEGs are shown in Figure 1. The reader is referred elsewhere for the clinical details which are essential for diagnosis of these conditions[2,3,6–9,20–24].

The other paroxysmal abnormality frequently encountered in childhood idiopathic generalized epilepsies is polyspike or polyspike and wave discharges[13]. These are generalized, symmetrical, of high amplitude and often accompanied by myoclonic jerks. Such discharges do not occur in CAE, are rare in JAE, but are characteristic of JME. In EMA similar discharges occur after eye closure and are associated with myoclonia of the eyelids[7]. Polyspike and polyspike and wave discharges are also encountered in symptomatic/cryptogenic myoclonic epilepsies.

Symptomatic/cryptogenic generalized epilepsies

These epilepsies are characterized by mixed generalized seizures, especially tonic, atonic, myoclonic and atypical absence seizures[4]. The exception is West syndrome, characterized by infantile spasms. The EEG findings in West syndrome will not be discussed further. The nosology of the symptomatic/cryptogenic generalized epilepsies of childhood is controversial, but useful division can be made between the Lennox–Gastaut syndrome and the symptomatic/cryptogenic myoclonic epilepsies[25]. The former clinically is characterized by tonic, atonic and atypical absence seizures, but not by the myoclonic seizures which occur in the latter conditions. The EEG is useful in classification of these epilepsies. Since seizures are frequent and seizure types are often difficult to identify from the history alone, video-EEG recordings are particularly helpful. The background EEG is usually diffusely slow[11]; however, it can be normal, especially early in the evolution of the epilepsy. Asymmetries of the background are common and may shift both within a recording and between recordings. One must not put much weight on such asymmetries.

The Lennox–Gastaut syndrome is characterized by the slow spike and wave discharge[11,13,26,27]. This superficially resembles the 3 Hz spike and wave discharge discussed previously but the frequency is <2.5 Hz, the discharge tends to be less well formed, and asymmetries, frequently shifting, are common. Both atypical absences and atonic seizures may accompany such discharges. Slow spike and wave discharge also occurs in other symptomatic/cryptogenic generalized epilepsies which lack the clinical features required for diagnosis of Lennox–Gastaut syndrome. Runs of generalized rhythmical 10 Hz spikes (sometimes called fast spike discharges) seen during sleep and sometimes accompanied by tonic seizures are more specific for the Lennox–Gastaut syndrome and some

Table 1 **Syndrome-related electroclinical features in idiopathic generalized epilepsies**

	EEG features of spike-wave discharge				***Clinical events during spike-wave discharges***	
	Duration of discharge	***Frequency***	***Morphology***	***Associated EEG features***	***Improvement of consciousness***	***Other features associated with absences***
CAE	Moderate – mean 12+/– 2 seconds	3–4 Hz; regular slowing from start to finish	Single, double or occasionally up to 3 spikes per discharge. Spikes are well formed and maintain a constant relation with the slow wave. No abrupt changes in amplitude. No fragmentations	Normal background apart from runs of posterior slow at around 3 Hz. Photosensitivity probably never seen.	Severe	Automatisms common—associated principally with the severity of impairment of consciousness. Eyes characteristically open and overbreathing stops within 3 seconds of discharge commencing. Other ictal features are confined to mild, unsustained eyelid fluttering or perioral movements
JAE	Long – mean 16+/– 7 seconds	3–4 Hz; regular slowing from start to finish	Spike and/or multiple spike and wave. Spikes are well formed and maintain a constant relation with the slow wave. Minimal changes in amplitude seen but no fragmentations	Normal background. Focal features rare. Photosensitivity rare	Severe, but less than in CAE	Automatisms common—associated principally with the severity of impairment of consciousness. If patient is counting breaths during hyperventilation this usually stops, but it, and other activities, may restart before cessation of the discharge.

Continued

continued

JME	Short – mean 6.8+/– 4.8 seconds	3–5 Hz (sometimes even faster). No smooth decrease in frequency	Multiple spike and wave discharges more commonly seen than spike and wave. Number and amplitude of spikes show considerable inter and intra discharge variation. Abrupt changes in amplitude and fragmentations common	Focal abnormalities in around 1/3 of patients. These consist of focal single spikes and spike-slow wave complexes or focal slow waves. They occur independently right or left but often show a unilateral preponderance Moderate excess of background theta often seen. Photosensitivity in around 27% of patients	Mild	Absences are simple, i.e. generally unaccompanied by other phenomena besides impairment of consciousness
EMA	Brief (generally 3–6 seconds)	3–6 Hz. No smooth decrease in frequency	Multiple spike and slow wave. More like those of JME than CAE or JAE	Absences are more likely to occur in a well lit room on eye closure. Total darkness eliminates eye-closure related abnormalities. Photosensitivity universal	Mild	Absences are accompanied by marked rhythmic eyelid myoclonia and often retropulsion of the eyeballs/ head and with a tonic component of the involved muscles
PMA	Brief (2–9 seconds)	3–4 Hz. No smooth decrease in frequency	Multiple spike and wave discharges. Number and amplitude of spikes show inter and intra discharge variation. Often no regular relationship between spikes and waves.	Focal abnormalities common. Photosensitivity not seen	Variable	Absences accompanied by rhythmic perioral myoclonia

authorities consider these a prerequisite for its diagnosis[11,13,28]. However, sleep recordings may be difficult to obtain and fast spike discharges may not be present throughout the evolution of the condition.

Symptomatic generalized/cryptogenic myoclonic epilepsies (as well as severe myoclonic epilepsy in infancy, which is undetermined whether partial or generalized) can usually be suspected on EEG by the occurrence of generalized spike (more usually polyspike) and wave abnormalities at frequencies above 3.5 Hz (fast spike and wave)[13,25,29]. Such discharges may coexist with slower spike and wave discharges. Photosensitivity is common in the symptomatic/cryptogenic generalized myoclonic epilepsies but is not seen in the Lennox–Gastaut syndrome.

Many children with symptomatic/cryptogenic generalized epilepsies defy precise syndromic diagnosis. Their EEG may show combinations of the abnormalities discussed previously. In addition, multifocal spike and sharp waves are common. These should not be taken as indicating a partial seizure disorder unless a consistent focal paroxysmal abnormality is accompanied by a consistent focal abnormality of the EEG background.

Finally, one cryptogenic generalized epilepsy illustrates the importance of using combined clinical and EEG criteria in classification. In myoclonic absence epilepsy the EEG is identical to that of CAE, with 3 Hz spike and wave discharges. Clinically, absence seizures are accompanied by severe rhythmic myoclonia, predominantly of the upper limbs. Unlike in CAE, response to medication is poor and mental decline is the rule[30].

Idiopathic partial epilepsies

The idiopathic partial epilepsies of childhood are benign epilepsies easily confused with more sinister ones, and the EEG is central to their diagnosis. Ictal recordings are only exceptionally obtained and the EEG diagnosis rests on interictal features. Therefore, video-EEG is not essential for full evaluation. EEGs, especially if recorded whilst the patient is awake, may be normal. Paroxysmal abnormalities are greatly enhanced by light sleep, but recording on awakening does not usually offer additional information. Hyperventilation and intermittent photic stimulation are not generally useful. Currently the international classification recognizes two idiopathic partial epilepsies of childhood (excluding

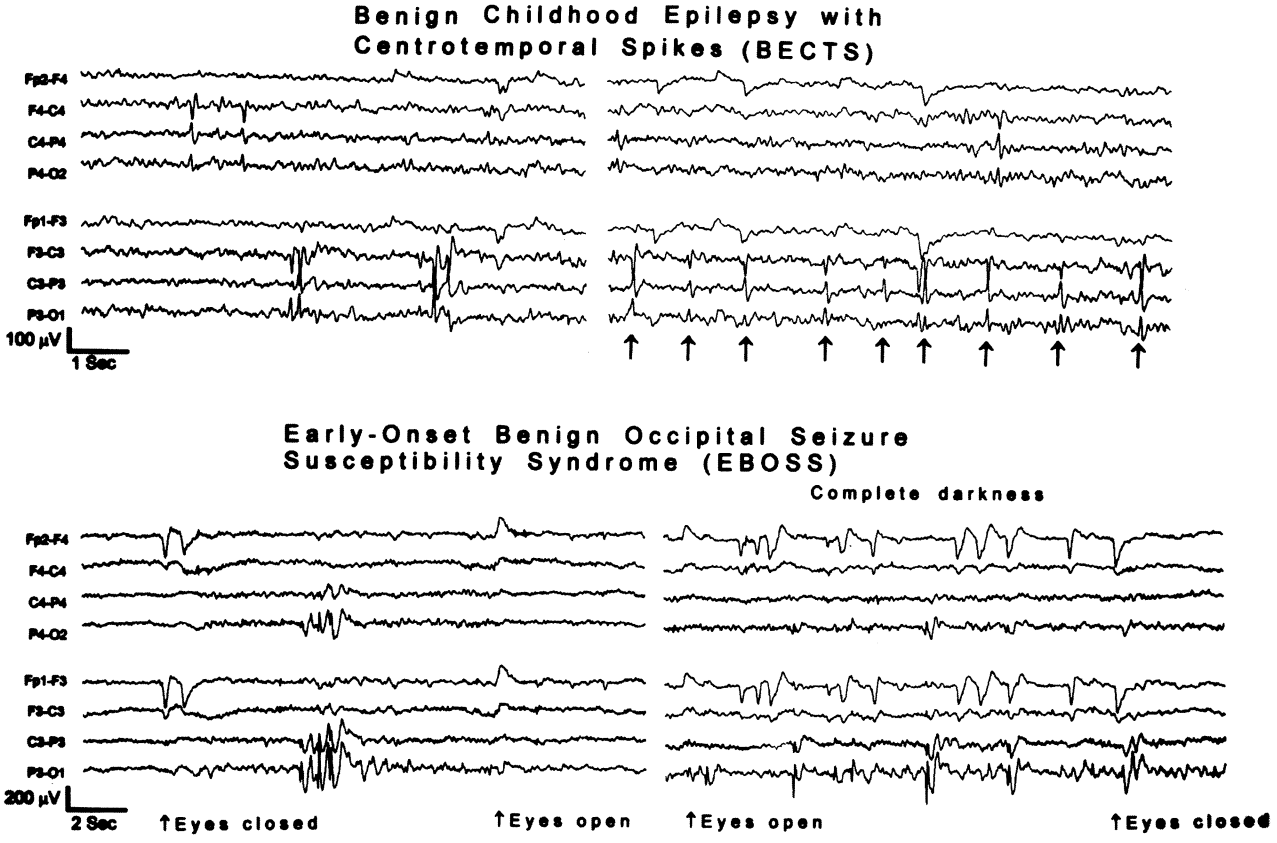

Figure 2 **Paroxysmal discharges in idiopathic partial epilepsies of childhood.** *Top:* BECTS—The trace on the right shows giant somatosensory evoked spikes induced by tapping the contralateral fingers (arrows). ***Bottom:*** GBOSS. Occipital paroxysms are present when the eyes are closed but attenuate when open to the light (left). In complete darkness, they are present with the eyes open and closed (right). They were also seen with the eyes open and closed when spherical lenses had eliminated central fixation (fixation-off sensitivity)

primary reading epilepsy)—benign epilepsy with centro-temporal spikes (BECTS) and childhood epilepsy with occipital paroxysms (CEOP)[4].

BECTS is the commonest idiopathic partial epilepsy, constituting up to 20% of all new-onset epilepsies in school-age children. It is characterized by distinctive, mainly nocturnal, simple hemifacial seizures with and without secondary generalization. The characteristic interictal EEG abnormalities consist of frequent, high-amplitude, sharp and slow wave complexes in the central and mid-temporal areas which may be unilateral or independently bilateral[31] (Figure 2). These have an unusual dipole with focal negativity in the mid-temporal regions and simultaneous positivity in the frontal regions. Demonstration of these in a child with a history compatible with BECTS and normal findings on examination is virtually diagnostic of the syndrome and further investigations are not required.

CEOP is probably two separate but related conditions—the classical late-onset variety characterized by simple diurnal seizures with visual symptoms, and the more common early-onset variety (also known as early onset benign occipital seizure susceptibility syndrome [EBOSS])[32], characterized by mainly nocturnal complex partial seizures with ictal vomiting and tonic eye and head deviation[33–35]. The characteristic interictal EEG abnormalities in both variants of CEOP are occipital paroxysms. These consist of long runs of high-amplitude sharp and slow wave complexes which are often bilateral (Figure 2). In shape and amplitude they resemble centro-temporal spikes[31]. Occipital paroxysms characteristically attenuate when the eyes are open and when the patient is visually fixating[36,37]. This can be tested for by recording in a completely darkened room: occipital paroxysms will occur both with the eyes open and the eyes closed. Introducing a tiny source of light onto which the patient can fixate will attenuate the occipital paroxysms. An alternative method of eliminating central fixation when the eyes are open is to place +10 spherical lenses in front of the patient's eyes. This phenomenon, known as fixation-off sensitivity, is characteristic but not universal in CEOP. There has been controversy concerning the specificity of occipital paroxysms for CEOP. This arose from the description of patients with occipital abnormalities only superficially resembling those seen in CEOP, in whom there was some attenuation with the eyes open[38,39]. The occurrence of occipital spikes in children with visual impairment and in those who do not have seizures has been known for over 40 years. These facts emphasize that the syndromic approach requires integration of many electro-clinical features; when undue emphasis is given to one feature, mistakes can arise.

Other idiopathic partial syndromes have been proposed but are less common and less well defined[40–42]. Clinically there may be overlap between the benign partial childhood epilepsies (e.g. a child with occipital seizures and other features of CEOP or EBOSS may later have a rolandic seizure), and there is also considerable overlap of EEG features, with many patients having additional sharp and slow wave complexes in cortical locations other than those characteristic of their syndrome[32]. In recognition of shared clinical and electrical features the term benign childhood seizure susceptibility syndromes has been proposed as an all embracing term[31]. Even if a patient cannot be classified into either BECTS or one of the variants of CEOP, an idiopathic partial epilepsy should be strongly suspected in a child with partial seizures who is neurodevelopmentally normal and whose EEG has a normal background but contains frequent paroxysmal abnormalities, similar to those described for BECTS, in any brain region.

Cryptogenic and symptomatic partial epilepsies

This group of epilepsies includes seizures arising from various brain regions including the medial temporal lobes, temporal neocortex, and the frontal, parietal and occipital lobes. Seizure semiology depends crucially on the site of origin and subsequent propagation. The interictal EEG is generally less useful in the classification of these epilepsies than for the other conditions. This is because the EEG findings compatible with them are very wide, normal EEGs are common and, whilst the location of unifocal abnormalities may reveal the origin of the seizures, accurate topographical localization may be difficult[43]. Sleep often activates abnormalities in the cryptogenic and symptomatic partial epilepsies and a sleep recording should always be attempted when one of these epilepsies is suspected and the awake EEG is normal.

Abnormalities of the background EEG are as important as paroxysmal epileptiform abnormalities in diagnosing these epilepsies. Focal or hemispheric slowing or amplitude asymmetries may point to the origin of the seizures (and to an underlying structural abnormality responsible for the epilepsy). Paroxysmal abnormalities which may be seen include unifocal spike or sharp waves or spike and wave complexes, multiple spike and waves and particularly in children, bilateral spike and wave abnormalities, synchronous or asynchronous.

CONCLUSIONS

The syndromic method takes more time and requires more detailed knowledge than the traditional approach. However, it offers considerable advantages in terms of targeted therapy and prognostic accuracy. Detailed EEG and video-EEG studies of the kind outlined here are a central component of the syndromic method and the role of the electroencephalographer is vital. However, this expertise is not universally available. All children with seizures should have at least one EEG examination by an electroencephalographer with expertise in the syndromic approach.

REFERENCES

1 Lerman P. Benign partial epilepsy with centro-temporal spikes. In: Roger J, Bureau M, Dravet C, Dreifuss FE, Perret A, Wolf P, eds. *Epileptic Syndromes in Infancy, Childhood and Adolescence*, 2nd edn. London: John Libbey, 1992:189–200

2 Grunewald RA, Panayiotopoulos CP. Juvenile myoclonic epilepsy: a review. *Arch Neurol* 1993;**50**:594–8

3 Panayiotopoulos CP. Juvenile myoclonic epilepsy: an underdiagnosed syndrome In: Wolf P, ed. *Epileptic Seizures and Syndromes*. London: John Libbey, 1994:221–30

4 Commission on Classification and Terminology of the International League Against Epilepsy. Proposal for revised classification of epilepsies and epileptic syndromes. *Epilepsia* 1989;**30**:389–99

5 Jeavons PM. Nosological problems of myoclonic epilepsies in childhood and adolescence. *Dev Med Child Neurol* 1977;**19**:3–8

6 Appleton RE, Panayiotopoulos CP, Acomb BA, Beirne M. Eyelid myoclonia with typical absences: an epilepsy syndrome. *J Neurol Neurosurg Psychiatry* 1993;**56**:1312–16

7 Giannakodimos S, Panayiotopoulos CP. Eyelid myoclonia with absences: a clinical and video-EEG study in adults. *Epilepsia* 1996;**37**:36–44

8 Panayiotopoulos CP, Ferrie CD, Giannakodimos S, Robinson RO. Perioral myoclonia with absences: a new syndrome. In: Wolf P, ed. *Epileptic Seizures and Syndromes*. London: John Libbey 1994:143–53

9 Panayiotopoulos CP, Ferrie CD, Giannakodimos S, Robinson RO. Perioral myoclonia with absences. In: Duncan J, Panayiotopoulos CP, eds. *Typical Absences and Related Epileptic Syndromes*. London: Churchill Communications, 1995:221–30

10 Bauer G. EEG, drug effects and central nervous system poisoning. In: Niedermeyer E, Lopes da Silva F, eds. *Electroencephalography: Basic Principles, Clinical Applications, and Related Fields*, 3rd edn. Baltimore: Williams & Wilkins, 1993:631–42

11 Niedermeyer E. Epileptic seizure disorders. In: Niedermeyer E, Lopes da Silva F, eds. *Electroencephalography: Basic Principles, Clinical Applications and Related Fields*, 3rd edn. Baltimore: Williams & Wilkins, 1993:461–564

12 Aliberti V, Gruenwald RA, Panayiotopoulos CP, Chroni E. Focal electroencephalographic abnormalities in juvenile myoclonic epilepsy. *Epilepsia* 1994;**35**:297–301

13 Niedermeyer E. Abnormal EEG patterns: epileptic and paroxysmal. In: Niedermeyer E, Lopes da Silva F, eds. *Electroencephalography: Basic Principles, Clinical Applications, and Related Fields*, 3rd edn. Baltimore: Williams & Wilkins, 1993:217–40

14 Panayiotopoulos CP, Giannakodimos S, Chroni E. Typical absences in adults. In: Duncan J, Panayiotopoulos CP, eds. *Typical Absences and Related Epileptic Syndromes*. London: Churchill Communications, 1995:289–99

15 Panayiotopoulos CP, Obeid T, Waheed G. Absences in juvenile myoclonic epilepsy: a clinical and video-electroencephalographic study. *Ann Neurol* 1989;**25**:391–7

16 Panayiotopoulos CP, Obeid T, Waheed G. Differentiation of typical absence seizures in epileptic syndromes. *Brain* 1989;**112**:1039–56

17 Panayiotopoulos CP, Chroni E, Daskalopoulos C, Baker A, Rowlinson S, Walsh P. Typical absence seizures in adults: clinical, EEG, video-EEG findings and diagnostic/syndromic considerations. *J Neurol Neurosurg Psychiatry* 1992;**55**:1002–8

18 Panayiotopoulos CP, Baker A, Grunewald R, Rowlinson S, Walsh P. Breath Counting during 3 Hz spike and wave discharges. *J Electrophysiol Technol* 1993;**19**:15–23

19 Giannakodimos S, Ferrie CD, Panayiotopoulos CP. Qualitative and quantitative abnormalities of breath counting during brief generalised 3 Hz spike and slow wave 'subclinical' discharges. *Electroencephalog Clin Neurophysiol* 1994

20 Panayiotopoulos CP. The clinical spectrum, of typical absence seizures and absence epilepsies. In Malafosse A, *et al.*, eds. *Idiopathic Generalized Epilepsies*. London: John Libbey, 1994:75–85

21 Panayiotopoulos CP. Juvenile absence epilepsy. In: Wallace S, ed. *Epilepsy in Children*. London: Chapman & Hall, 1996:325–32

22 Panayiotopoulos CP. Juvenile myoclonic epilepsy. In: Wallace S, ed. *Epilepsy in Children*. London: Chapman & Hall, 1996:333–48

23 Loiseau P, Duche B. Childhood absence epilepsy. In: Duncan J, Panayiotopoulos CP, eds. *Typical Absences and Related Epileptic Syndromes*. London: Churchill Communications, 1995:152–60

24 Duncan J, Panayiotopoulos CP. Juvenile absence epilepsy: an alternative view. In: Duncan J, Panayiotopoulos CP, eds. *Typical Absences and Related Epileptic Syndromes*. London: Churchill Communications, 1995:167–73

25 Aicardi J. *Epilepsy in Children*. New York: Raven Press, 1994

26 Aicardi J, Levy Gomes A. The Lennox–Gastaut syndrome: clinical and electroencephalographic features. In: Niedermeyer E, Degan R, eds. *The Lennox–Gastaut Syndrome*. New York: Alan R Liss, 1988:25–46

27 Blume WT. The EEG features of the Lennox–Gastaut syndrome. In: Niedermeyer E, Degan R, eds. *The Lennox–Gastaut Syndrome*. New York: Alan R Liss, 1988:159–76

28 Beaumanoir A, Dravet C. The Lennox–Gastaut syndrome. In: Roger J, Bureau M, Dravet C, Dreifuss FE, Perret A, Wolf P, eds. *Epileptic Syndromes in Infancy, Childhood and Adolescence*, 2nd edn. London: John Libbey, 1992:115–32

29 Dravet C, Bureau M, Guerrini R, Giraud N, Roger J. Severe myoclonic epilepsy in infants. In: Roger J, Bureau M, Dravet C, Dreifuss FE, Perret A, Wolf P, eds. *Epileptic Syndromes in Infancy, Childhood and Adolescence*, 2nd edn. London; John Libbey, 1992:75–88

30 Tassinari CA, Bureau M, Thomas P. Epilepsy with myoclonic absences. In: Roger J, Bureau M, Dravet C, Dreifuss FE, Perret A, Wolf P, eds. *Epileptic Syndromes in Infancy, Childhood and Adolescence*, 2nd edn. London: John Libbey, 1992:115–32

31 Panayiotopoulos CP. Benign childhood partial epilepsies: benign childhood seizure susceptibility syndromes. *J Neurol Neurosurg Psychiatry* 1993;**56**:2–5

32 Ferrie CD, Beaumanoir A, Guerrini R, *et al.* Early-onset benign occipital seizure susceptibility syndrome. *Epilepsia* 1997;**38**:285–93

33 Panayiotopoulos CP. Benign childhood epilepsy with occipital paroxysms: a 15-year prospective study. *Ann Neurol* 1989; **26**:51–6

34 Gastaut H. Benign epilepsy of childhood with occipital paroxysms. In: Roger J, Bureau M, Dravet C, Dreifuss FE, Perret A, Wolf P, eds. *Epileptic Syndromes in Infancy, Childhood and Adolescence*, 2nd edn. London: John Libbey, 1992:201–18

35 Panayiotopoulos CP. Benign nocturnal childhood occipital epilepsy: a new syndrome with nocturnal seizures, tonic deviation of the eyes, and vomiting. *J Child Neurol* 1989;**4**:43–8

36 Panayiotopoulos CP. Inhibitory effect of central vision on occipital lobe seizures. *Neurology* 1981;**31**:1331–3

37 Panayiotopoulos CP. Fixation-off-sensitive epilepsy in eyelid myoclonia with absence seizures. *Ann Neurol* 1987;**22**:87–9

38 Aicardi J, Newton K. Clinical findings in children with occipital spike-waves suppressed by eye opening. In: Andermann F, Lugaresi E, eds. *Migraine and Epilepsy*. London: Butterworth, 1987:111–24

39 Cooper GW, Lee SI. Reactive occipital activity: is it benign? *Epilepsia* 1991;**32**:663–8

40 Panayiotopoulos CP. Epilepsies characterized by seizures with specific modes of precipitation (reflex epilepsies). In: Wallace S, ed. *Epilepsy in Children*. London: Chapman & Hall, 1996:356–75

41 Tassinari CA, De Marco P. Benign partial epilepsy with extreme somato-sensory evoked potentials. In: Roger J, Bureau M, Dravet C, Dreifuss FE, Perret A, Wolf P, eds. *Epileptic Syndromes in Infancy, Childhood and Adolescence*, 2nd edn. London: John Libbey, 1992:225–30

42 Dalla Bernardina B, Sgro V, Fontana E, Colamaria V, La Selva L. Idiopathic partial epilepsies in children. In: Roger J, Bureau M, Dravet C, Dreifuss FE, Perret A, Wolf P, eds. *Epileptic Syndromes in Infancy, Childhood and Adolescence*, 2nd edn. London: John Libbey, 1992:173–88

43 Curatolo P. Epilepsies symptomatic of structural lesions. In: Wallace S, ed. *Epilepsy in Children*. London: Chapman & Hall, 1996:399–415