Ictal Patterns in Generalized Epilepsy

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Summary: Ictal EEG may be of great benefit in facilitating accurate classification of the underlying seizure disorder in some patients and thus guiding further investigation and management. Ictal recordings in patients with generalized epilepsies are protean in their manifestations and yet may have considerable overlap. Classification is only possible through careful synthesis of all available clinical and electrophysiological data. Although the underlying pathophysiological mechanisms of the generalized epilepsies remain uncertain, evidence from EEG recordings tends to support Gloor's concept of corticoreticular epilepsy. Key Words: Ictal EEG—Corticoreticular epilepsy—Idiopathic generalized epilepsy—Symptomatic generalized epilepsy.

This article discusses ictal patterns recorded from scalp EEG in patients with various forms of generalized epilepsy. We emphasize the correlation of ictal semiology with electrographic changes, the value of ictal over interictal recordings in accurate classification of the epilepsies, some of the limitations of scalp EEG recording of ictal activities, and the major pitfalls in recording and interpreting ictal discharges.

CLASSIFICATION OF THE GENERALIZED EPILEPSIES

The Commission on Classification and Terminology of the International League Against Epilepsy has proposed revised classifications of both epileptic seizures (1981) and the epilepsies and epileptic syndromes (1989). It defines the generalized epilepsies and syndromes as epileptic disorders with generalized seizures, i.e., "seizures in which the first clinical changes indicate initial involvement of both hemispheres.... The ictal electroencephalographic patterns initially are bilateral." There are two major types of generalized epilepsies: idiopathic generalized epilepsy with an EEG expression of a generalized, bilateral, synchronous, symmetrical discharge with no known or suspected etiology other than a possible hereditary predisposition and those generalized epilepsies secondary to some underlying process either identified (symptomatic) or unidentified (cryptogenic). The classification utilizes interictal and in some cases ictal EEG data in describing these epileptic syndromes; yet, the vast majority of patients with seizures have their epilepsy classified and treatment instituted without resort to recorded ictal data. Despite some inherent limitations, e.g., the occurrence of complex partial seizures in some patients with symptomatic generalized epilepsy, or knowing whether patients with nocturnal generalized tonic-clonic seizures might have a focal onset or not, the classification represents the best available scheme for categorizing patients with seizures to guide investigation and management.
PATHOPHYSIOLOGY

An understanding of the mechanisms involved in the production of epileptiform discharges on scalp EEG would be helpful in interpreting interictal and ictal recordings, but, despite much clinical and experimental review, the responsible mechanisms remain uncertain. The bilaterally synchronous and symmetric spike-and-wave discharges on the scalp suggest a deep-seated generator. An early concept was that of centrencephalic epilepsy, arising from Morison and Dempsey’s (1942) work on the specific and nonspecific thalamic activating systems. Jasper and Droogleever-Fortuyn (1946) produced bisynchronous spike-and-wave discharges in the cat by electrical stimulation of the intralaminar nuclei of the thalamus at 3 Hz. Similarly, alumina implantaions into the intralaminar nuclei of the thalamus and reticular formation of the midbrain of the cat produced typical 3-Hz spike-wave activity with seizures (Guerrero-Gigueroa et al., 1963). In a study of depth recording in six children with absence seizures, Williams (1953) proposed that the 3-Hz spike-wave activity had its origin in the thalamus and then increased in voltage until it reached the cortex, which then set up a reverberating cortical-thalamic circuit. In some patients with symptomatic generalized epilepsy, Velasco et al. (1989) described spike-wave complexes in the centromedian nucleus of the thalamus preceding bilateral surface cortical discharges during symptoms of nonconvulsive generalized seizures.

Other clinical and experimental work, however, suggested that the cortex played a primary role. Tükel and Jasper (1952) described generalized spike-wave discharges in patients with lesions in anterior parasagittal areas and mesial lesions of the brain. Marcus and Watson (1966) using bilaterally symmetrical cobalt foci in the monkey cortex produced synchronous spike-wave discharges if the corpus callosum was intact. Niedermeyer et al. (1969), in depth recordings in patients with generalized spike-and-wave, found focal discharges in the frontal lobe in two patients. Bancaud et al. (1974) showed that electrical stimulation of human mesial frontal cortex produced clinical and EEG activity indistinguishable from spontaneous spike-wave bursts and absence. Working with the photosensitive West African baboon Papio papio, Naquet et al. (1972) showed that the discharges of the spontaneous and photically induced seizures have a frontorolandic origin.

From his experimental work, Gloor has provided a unitary hypothesis, termed the corticoreticular epilepsies. Large intramuscular injections of penicillin in the cat produced a transient epileptogenic state with generalized synchronous discharges during which the cat stares, blinks his eyes, and has myoclonic twitches of the face and brachial musculature. These findings could be reproduced by the application of a weak solution of penicillin to wide areas of the cortex bilaterally but not by its application to the thalamus or its intralaminar nuclei. Discharges and seizures can be triggered by low-frequency, thalamic stimulation, which, in the normal animal, produces spindles and recruiting responses. Gloor et al. (1977) proposed a state of mild generalized cortical hyperexcitability with epileptiform discharges and seizures triggered by incoming thalamocortical volleys. The report of Bickford et al. (1955) seems supportive of Gloor’s hypothesis. An 11-year-old girl with a story of absence seizures since age 2 developed other seizures more suggestive of a partial onset after head trauma. She showed typical 3-Hz spike-and-wave discharges and focal sharp waves in the right frontal region, some of which triggered generalized spike-and-wave activity. After depth electrode recordings from both frontal and subcortical regions and with one electrode implanted in the thalamus, it was concluded that there was no evidence to support the spike being initiated in the thalamic region. With brief electrical stimuli of the depth contacts, long episodes of spike-and-wave activity occurred accompanied by the typical symptoms of a petit mal seizure. There was no evidence to suggest that these discharges were being initiated exclusively from any single region, cortical, subcortical, or thalamic. These authors’ conclusions were that the findings in this patient supported an abnormally facilitated diffuse thalamocortical system as the basic “electro-pathology” in absence seizures.

Many spike-wave discharges seen during sleep occur in conjunction with K complexes, which are maximal over the Fz electrode rather than their more usual Cz maximum. It appears that arousal stimuli generate both the K complexes and the spike-wave discharges. The same mechanism is probably at play during wakefulness, but the K complexes are not available as visual guides. Gloor and Testa (1974) felt that this cortical hyperexcitability was particularly prominent at times when the ascending reticular activating system was inactive. Some clinical observations provide supportive data. Studying an 8-year-old girl with absence seizures, Cleeland and Booker (1967) found that the seizures were best developed during drowsiness and by partial sensory restraint.
In a study of 16 children with symptomatic generalized epilepsy, Papini et al. (1984) found that, of 406 recorded seizures, 53.9% occurred during inactive wakefulness, 31.5% during drowsiness, and only 8.1% during active wakefulness.

Thus, the corticoreticular epilepsies may depend on two anatomical areas—the modulating influence of the thalamus and mesencephalic reticular formation on a diffusely hyperexcitable cortex. This appears a likely mechanism, particularly in absence seizures. As Gloor emphasizes, it represents just one end of the spectrum with the diffuse hyperexcitability perhaps due to an inherited or biochemical trait. Diffuse cortical hyperexcitability could result from decreased inhibitory neurotransmission, as suggested by the observation in primary generalized epilepsy of a modest (approximately 15%) generalized decrease in cortical density of central benzodiazepine receptors, which are associated with the predominant cerebral mediator of inhibitory neurotransmission, the GABAergic chloride ionophore (Savic et al., 1990).

Other cases of corticoreticular epilepsies, arising from more localized disturbances such as a tumor, trauma, or following infection, may have a less important genetically determined influence. The underlying basis for the EEG findings in the symptomatic generalized epilepsies may be even more complex. In infantile spasms, a number of lines of evidence suggest that the primary abnormality may lie in the pons. Sleep studies have shown a marked decrease in rapid eye movement (REM) sleep in children with infantile spasms compared to normal infants and a decreased total sleep time, and reversal of the REM sleep abnormality only in infants whose EEG and clinical picture improved with adrenocorticotropic hormone or prednisone (Hrachovy et al., 1981). Neuropathological changes in the pons have been described in infants who had infantile spasms (Morimatsu et al., 1972), although this has not been substantiated by other workers (Jellinger, 1987).

Lower concentrations of the serotonin metabolite 5-hydroxyindoleacetic acid in the cerebrospinal fluid (CSF) of infants with infantile spasms compared to age-matched controls (Silverstein and Johnston, 1984) suggest a state of supersensitivity of serotonin receptors resulting in diminished presynaptic serotonin turnover and release from serotonin-containing neurons in the raphe region of the pons. However, the work of Chugani and colleagues (1992), using positron emission tomography in infants with infantile spasms, suggests that the primary lesion in infantile spasms is a focal or diffuse cortical abnormality, which at a critical stage of maturation causes abnormal functional interactions with brainstem raphe nuclei. Older infants and children with symptomatic generalized epilepsies, with or without preceding infantile spasms, are most likely to have conditions with diffuse or multifocal cortical insults. The presence of independent focal spikes in the frontal or temporal regions particularly and the manner in which some patients with symptomatic generalized epilepsy will display slow spike-and-wave activity on one EEG but multiple independent spike foci on another (Markand, 1977) supports the primary insult being cortical in these cases.

**SEIZURES OF THE IDIOPATHIC GENERALIZED EPILEPSIES**

The seizures that appear in these epileptic syndromes, which are also called primary generalized or intrinsic generalized epilepsies, typically present after the age of 4 years and consist of three major types: absence, convulsive seizures either tonic-clonic or clonic- tonic–clonic type, and myoclonic.

**Absence Seizures**

The classic interictal EEG of absence seizures is the presence of a normal background activity superimposed on which are bilaterally symmetric and synchronous 3-Hz spike-and-wave discharges usually with a superior frontal maximum. In some children with absence seizures, paroxysms of interictal occipital rhythmic delta activity provide an exception to the general rule that background activity is normal in the primary generalized epilepsies. These paroxysms should not be mistaken for partial seizures. Additionally, medication effects and other factors not inherent in the epilepsy itself may cause abnormalities of background activity. The generalized spike-and-wave discharges begin abruptly at a frequency of 3.5–4 Hz, gradually slowing to 2.5–3 Hz. As the burst progresses, the spike discharges may become lower in amplitude. The discharges are readily provoked by both hyperventilation and hypoglycemia. The distinction between interictal and ictal activities may be merely a measure of the duration of the discharge. Since the morphology and topography of the generalized spike-wave is identical in either case. Those discharges that last longer than 3 s have a readily recognized clinical accompaniment with an arrest of movement, a vacant appearance to the eyes, and then return to previous...
activities after the discharge has ceased (Fig. 1). On occasion, absence seizures may be associated with minor motor activity with either increases or decreases in postural tone, mild clonic activity, automatisms, autonomic phenomena, or a combination of some of these features. In testing auditory reaction times, Browne et al. (1974) found them to be normal in the 1 s before a paroxysm, but only 45% of them were normal at the start of a paroxysm and only 4% of reaction times were normal in the first second of the discharge. The stimulus mode may be a factor in some differences described in the literature in responsiveness to different sensory inputs. Orren quoted by Browne (1983) found several patients with greater responsiveness to auditory than visual clues during spike-wave paroxysms. Some authorities (Mirsky and VanBuren, 1965; Geller and Geller, 1970) have shown impaired visual attentiveness 0.5 s before the start of a discharge. Others (Orren, 1978) have shown decreased visual evoked potential amplitude before discharges. Absence seizures may also occur with other types of discharges, such as generalized irregular spike-and-wave bursts, with slow spike-and-wave activity (Gomez and Westmoreland, 1987), or with generalized paroxysmal fast activity (Lee, 1983).

**Generalized Tonic-Clonic Seizures**

In some forms of primary generalized epilepsy with generalized tonic-clonic seizures, the baseline EEG recording will be normal, whereas other patients will show bursts of generalized 3-Hz spike-and-wave, generalized irregular polyspike-wave or bursts of frontal intermittent rhythmic delta activity. Immediately preceding the tonic phase of the seizure, there may be preictal bursts of generalized polyspike-wave activity associated with bilateral massive myoclonus (Gastaut and Broughton, 1972). The seizure will commence with a brief period of extreme voltage attenuation with or without superimposed low-voltage, high-frequency generalized paroxysmal fast activity, which gradually becomes more discernable after being initially obscured by the prominent tonic phase of the seizure (Fig. 2, left). Often, the first detectable EEG change is the appearance of paroxysmal fast activity at about 10 Hz with rapidly increasing amplitude (Gastaut and Broughton, 1972). After approximately 10 s, the paroxysmal fast activity becomes intermixed with rhythmic slow activities, which gradually become more prominent as the paroxysmal fast activity wanes. This blend of rhythmic slowing and the fragments of generalized paroxysmal fast activity creates a polyspike-and-wave appearance. As the slower EEG rhythm reaches approximately 4 Hz, the tonic muscle contractions give way to interruptions in muscle tone manifesting the clonic phase of the seizure. The last clonic movement is followed by profound voltage suppression whose duration depends on the length of the

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**FIG. 1.** Recorded absence seizure in a 19-year-old woman with juvenile absence epilepsy. Spike-wave frequency slows from 3.5 Hz to 2.5 Hz during this simple staring spell.

**FIG. 2.** Four discontinuous EEG samples recorded during a 65-s generalized tonic-clonic seizure in a 39-year-old man with post-traumatic symptomatic generalized epilepsy. The first sample shows frequent generalized spike-wave activity interictally, followed by generalized hypertonus. The second sample shows complete myogenic obscuration of EEG activity during generalized hypertonus, while myogenic and kinesogenic artifacts obscure the third EEG sample during generalized clonus. Lower-amplitude generalized jerks occur synchronously with spike-wave complexes during the fourth sample. The fourth sample also demonstrates severe postictal suppression of EEG activity (in artifact-free channels).
preceding seizure or seizures and any coexisting causes of encephalopathy (Fig. 2, right). The return of cerebral activity is initially in the delta frequency range and gradually increases in amplitude and frequency until restitution of the normal background activities of the patient. The duration of the postictal phase varies significantly and tends to be longer in young children or in patients who have had multiple seizures.

**Clonic-Tonic-Clonic Seizures**

In these seizures, the tonic and the second clonic phase are similar to the findings already described for the generalized tonic-clonic seizures. In eight patients with clonic-tonic-clonic seizures described by Delgado-Escueta and Enrile-Bascal (1984), the ictal EEG manifestations consisted of diffuse 10–16-Hz spikes during the myoclonic jerks, which evolved into a tonic extensor posture. During the later clonic phase, when extension periodically relaxed, diffuse high-voltage rapid spikes were interrupted by quiescence on the EEG.

**Myoclonic Seizures**

In the idiopathic generalized epilepsies, myoclonic jerks are typically associated with high-amplitude, very rapid 10–15-Hz spikes with or without accompanying slow waves. They may occur alone or precede generalized tonic-clonic seizures. In juvenile myoclonic epilepsy, the EEG during myoclonic jerks was reported by Delgado-Escueta and Enrile-Bascal (1984) as showing high-amplitude 10–16-Hz spikes, sometimes preceded by diffuse irregular 2–5-Hz spike-wave complexes and followed by irregular 1–3-Hz slow waves. Most children with absence seizures who have prominent myoclonus ultimately prove to have a condition other than an idiopathic generalized epilepsy. The two major syndromes that occur in this context, the myoclonic-astatic seizures of Doose (1970) and myoclonic absence (Tassinari and Bureau, 1985), will be discussed under the symptomatic generalized epilepsies.

**Syndromes of the Idiopathic Generalized Epilepsies**

The epileptic syndromes that constitute the idiopathic epilepsies are characterized by the occurrence of different types of generalized seizures, sometimes of more than one type, with an interictal EEG that typically shows normal background activities and the appearance of generalized spike-and-wave discharges of varying frequencies. The patients are usually neurologically normal, and in most instances the seizure disorder itself is a relatively benign one. The likelihood of an inherited basis for the generalized epilepsies is strongest in these groups of patients. The principal epilepsies that occur in this subset are childhood absence, juvenile absence, generalized tonic-clonic seizures, and juvenile myoclonic epilepsy.

The entity known as childhood absence or psychomotor epilepsy petit mal (Drury and Dreifuss, 1985) encompasses that subgroup of the primary generalized epilepsies with absence seizures as its fundamental and usually sole manifestation. It occurs in children aged between 4 and 8 years, who frequently have a family history of epilepsy. Intellectual and neurological examinations are normal, and there is usually a very favorable response to treatment.

Juvenile absence epilepsy has an onset around puberty with less frequent absence seizures than childhood absence (Wolf, 1985). Associated generalized tonic-clonic seizures are more common in this syndrome than in childhood absence. The EEG manifestations are similar to childhood absence (Fig. 1), although generalized fast spike-polyth waveform discharges may be more common.

Juvenile myoclonic epilepsy has its onset in early teenage years and is characterized by absence, myoclonic, tonic-clonic, and clonic-tonic-clonic seizures. The interictal EEG pattern displays a normal background frequency and frequent bursts of generalized, bisynchronous polyspike-wave activity with a frequency greater than 3 Hz (Delgado-Escueta and Enrile-Bascal, 1984; Janz, 1985). A photoparoxysmal response is seen in about one-third of patients (Asconape and Penry, 1984; Janz, 1985). The absence seizures in this syndrome (Fig. 3) are associated with either 3-Hz spike-wave complexes similar to those
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seen in pyknolepsy or rapid polyspike-wave complexes slowing to 3 Hz during unconsciousness (Delgado-Escueta and Enril-Bascal, 1984; Janz, 1985). In a clinical and video/EEG study of these absences (Panayiotopoulos et al., 1989), the clinical manifestations were subtle even during prolonged discharges and often apparent only if the patient was involved in some type of activity during the ictus. The EEG manifestations were variable but usually consisted of polyspike-wave complexes of rather irregular frequency rather than the smooth 3-Hz spike-wave complexes of pure absence. Myoclonic jerks in this syndrome are most likely to occur in the transition from sleep to wakefulness and during photic stimulation.

Other types of idiopathic generalized epilepsies occur, and many patients who appear to have an idiopathic generalized epilepsy may not fit neatly into any specific category. For instance, the Commission on Classification and Terminology of the International League Against Epilepsy (1989), which proposed the revised classification of epilepsies and epileptic syndromes, recognizes the entity of epilepsies with grand mal seizures on awakening. These patients have generalized 3-Hz or more rapid spike-wave discharges on EEG but no evidence of absence seizures or myoclonic jerks.

SEIZURES OF THE SYMPTOMATIC GENERALIZED EPILEPSIES

These epileptic syndromes may present at any age but commonly occur in infancy or early childhood. Their clinical and EEG features may vary significantly, to a much greater extent than the idiopathic generalized epilepsies and are chiefly dependent on the age of the child. Seizures that occur in these syndromes may be considered under the following categories: convulsive, nonconvulsive, and infantile spasms and myoclonic seizures.

Convulsive Seizures

The EEG appearance of tonic-clonic seizures has already been described.

The classic EEG accompaniment of tonic seizures is generalized paroxysmal fast activity (GPFA) (Blume et al., 1973; Markand, 1977; Beaumanoir, 1985). This pattern typically occurs during sleep when many bursts are not associated with any clinical change, whereas in wakefulness most bursts are ictal. The discharges consist of bursts of 10-25-Hz spikes averaging 3-5 s, preceded or followed by generalized sharp-and-slow-wave complexes (Fig. 4) (Brenner and Atkinson, 1982). There is little change in the frequency of the spikes during the bursts, but their amplitude may decrease as the discharge progresses (Fig. 5). The discharges typically have an amplitude maximum at the superior frontal electrodes, are bilaterally synchronous but may have a shifting voltage emphasis between the two hemispheres, and rarely may occur unilaterally. Activation procedures such as hyperventilation and photic stimulation do not provoke GPFA. Blume et al. (1973) also described tonic seizures occurring with electrodecremental responses.

Nonconvulsive Seizures

Atypical absence seizures are characterized by a more gradual onset and cessation than pure absence seizures and are often accompanied by a slight decrease in body tone (Beaumanoir, 1985). Their EEG accompaniment consists of slow spike-and-wave activity unchanged from the baseline tracing, or more hypersynchronous slow spike-and-wave activity (Fig. 6) (Markand, 1977).

Atonic and myoclonic seizures that may be clinically indistinguishable may be associated with a more rapid high-voltage spike-and-wave activity sometimes accompanied by brief electrodecremental responses (Fig. 7) (Gastaut and Broughton, 1972, Blume, 1987) or without any identified change on scalp EEG (Gastaut and Broughton, 1972; Markand, 1977; Blume, 1987). In an analysis of 239 drop attacks in 45 patients with symptomatic generalized epilepsy recorded with CCTV-EEG techniques, Egli et al. (1985) observed that the majority of these drop attacks were not in fact tonic seizures but pure tonic seizures, which they termed axial spasms. A sudden
flexion of the hips, upper trunk, and head led to a fall produced by the rapidity and severity of the hip flexion. These episodes could occur alone or in connection with other seizure manifestations, such as absences. In the pure axial spasms, there were no disturbances of consciousness and no significant EEG changes. When axial spasms were combined with other seizure manifestations, they were preceded by an absence seizure and generalized spike-wave activity with the onset of the spasm coinciding with the last generalized spike. Egli et al. (1985) believe that axial spasms represent a more mature form of infantile spasms. Akinetic seizures characterized by a complete lack of mobility despite preservation of muscle tone and associated with an impairment of consciousness have similar or identical EEG discharges to those seen in atonic seizures (Gastaut and Broughton, 1972).

Infantile Spasms and Myoclonic Seizures

Gibbs and Gibbs (1952) coined the term hypsarhythmia to describe the high-amplitude irregular asynchronous delta activity with multiple independent and shifting spike foci, which is the classic EEG finding in this condition. Gibbs and Gibbs (1952) described in their atlas diffuse spike discharges associated with the jerking and quivering movements of the infantile spasms. It was quickly appreciated that the most common EEG accompaniment to the spasms was an abrupt generalized reduction in voltage, termed electrodecremental by Bickford and Klass (1960). The complex motor phenomena that may occur during infantile spasms were first documented by Pampiglione (1964). He demonstrated that these spasms were associated with activity in both flexor and extensor muscles, and that, even when the muscle activity appeared to be extremely brief, it was usually accompanied by a group of muscle action potentials, thus differing from the activities seen in myoclonic seizures. Kellaway and colleagues (1979) reported an analysis of over 5,000 spasms in 24 infants with a synchronized video and polygraphic recording system. In their study, mixed spasms with both flexor and extensor activity were the most common type, accounting for 42% of the total, followed by spasms in flexion (34%) and extension (22%). These authors were the first to describe the akinia and attenuated responsiveness that could either follow a spasm or occur independently as a seizure without a preceding spasm. These events they termed an "arrest."

Spasms in West's syndrome tend to cluster and are especially likely to occur soon after arousal from sleep. The spasms seem least likely to occur when the child has been stimulated through measurement associated with the jerking and quivering movements of the infantile spasms. It was quickly appreciated that the most common EEG accompaniment to the spasms was an abrupt generalized reduction in voltage.
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and application of EEG electrodes. For these reasons and because seizure activity may be very subtle in these children, it is the policy in our laboratory to record EEGs in children with known or suspected infantile spasms only as 2-h-long video/EEG recordings. This allows both for an adequate transition through states of wakefulness, drowsiness, sleep, and arousal and also facilitates the close correlation of behavioral activities with EEG changes.

Kellaway et al. (1979) identified 11 different ictal EEG patterns accompanying infantile spasms. About 80% of these events, however, consisted of a generalized, frontally dominant slow-wave or sharp-and-slow-wave complex with or without an aftergoing period of abrupt voltage attenuation (Fig. 8). Almost 38% of the over 5,000 events were accompanied by a generalized slow-wave transient followed by voltage attenuation. There was no correlation between the type of spasm and the ictal pattern seen.

However, “arrest” attacks and asymmetrical seizures were most likely to be accompanied by a diffuse voltage attenuation with superimposed fast activity, although the number of these events seen was very small. Electrodecremental episodes may occur in the awake or sleeping state without evidence of a clinical seizure or any recorded change in the electromyographic or other polygraphic channels (Kellaway et al., 1979), and, conversely, in the presence of marked spasms, no obvious EEG changes may be seen on occasion (Pampiglione, 1964). In our experience and that of others (Westmoreland and Gomez, 1987), it is relatively common to see an alternate pattern during sleep in children with hypsarrhythmia that approximates a burst-suppression pattern (Fig. 9). Bursts of high-amplitude, rather asynchronous slowing are followed by 1–2 s of abrupt voltage attenuation with this pattern repeating itself regularly throughout the sleeping tracing. Such activity should not be viewed as indicative of underlying seizure activity. Transient normalization of the EEG activity for age may occur after a flurry of spasms (Kellaway et al., 1979).

Patients with infantile spasms may also have myoclonic jerks. As illustrated in Fig. 10, the myoclonic jerks are associated with a different EEG pattern than infantile spasms, with high-amplitude rapid spike or polyspike discharges without an ensuing electrodecremental pattern. Partial seizures may
FIG. 11. Recorded partial seizure in the same child as in Fig. 10. This partial seizure was associated with the child rolling both her eyes back and appearing unresponsive. Its EEG accompaniment consisted of high-amplitude spike-and-wave activity widely distributed over the right hemisphere.

also occur in children with infantile spasms (Fig. 11).

In patients with progressive myoclonus epilepsy, the myoclonus may be very prominent during the later stages of the illness. Although associated with very frequent generalized spike-wave activity, there need not be a precise temporal correlation between the myoclonus and epileptiform activity, at least in Lafora disease (Yen et al., 1991), and probably in other forms of progressive myoclonic epilepsy also.

Myoclonus may also be a prominent manifestation in two other epileptic syndromes, the myoclonic-astatic seizures of Doose (1970) and myoclonic absence (Tassinari and Bureau, 1985). In the Doose syndrome, the interictal EEG is dominated by theta activity with a parietal maximum and bisynchronous frontally dominant fast polyspike-and-wave activity. Myoclonic jerks may be prominent and are associated with short bursts of high-frequency polyspike-and-wave activity. The astatic or myoclonic-astatic seizures are associated with 2-3-Hz spike-and-wave activity. GPFA occurs in sleep. Myoclonic absence is characterized by absence seizures with prominent myoclonus and sometimes with tonic activity but with the typical 3-Hz spike-wave activity that is seen in pyknolepsy. The attacks are precipitated by hyperventilation. No other seizure types or EEG discharges such as GPFA occur initially. Ultimately, myoclonic absence seizures disappear and other seizure types typical of the symptomatic generalized epilepsies emerge.

 Syndromes of Symptomatic Generalized Epilepsy

The clinical manifestations in patients with symptomatic generalized epilepsies are commonly age-dependent. In West's syndrome, first described by Dr. W. J. West, a general practitioner in England, in his own son in 1841, there is a strong correlation between the occurrence of infantile spasms and the appearance of hypsarrhythmia on EEG, but infantile spasms may appear without a hypsarrhythmic EEG. Nevertheless, the concordance of infantile spasms and hypsarrhythmia together is higher than any other epileptic electroclinical syndrome irrespective of age or etiology.

Symptomatic generalized epilepsies commonly manifest themselves in older children and adults as the Lennox-Gastaut syndrome characterized by mental retardation, multiple generalized seizure types often refractory to anticonvulsant therapy, and slow spike-and-wave discharges on EEG. The interictal EEG manifestations of this syndrome, however, are imprecise, vary from patient to patient, and may even vary within the same patient depending on the state of arousal or from one study to another. The major seizure types seen in this syndrome are atypical absence (Fig. 6), tonic (Fig. 5), atonic (Fig. 7), and myoclonic (Fig. 12), but generalized tonic-clonic, akinetic, clonic (Fig. 13), and complex partial seizures (Fig. 14) also occur. Many patients show frequent prolonged trains of slow spike-and-wave activity. In most instances, these are not accompanied by any recognizable clinical change, although response testing may be limited due to mental dullness. There may even be significant variation of the latency of responses during periods with no slow spike-and-wave activity (Erba and Cavazzuti, 1977).

The epileptic syndromes of Doose (1970) and myo-
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clonic absence (Tassinari and Bureau, 1985) that are considered by some authorities to be distinct entities, represent in our opinion forms of symptomatic generalized epilepsy that begin in the latter half of the first decade of life in children who were previously neurologically normal but then evolve to look similar to those children with Lennox-Gastaut syndrome.

Children with multiple independent spike foci on their interictal EEG usually have the same clinical features and many of the same ictal events as children with slow spike-and-wave discharges. In some instances, as reported by Markand (1977), children with symptomatic generalized epilepsy may show slow spike-and-wave discharges on one EEG recording or in a particular state and multiple independent spike foci on another. Burnstine et al. (1991) reported scalp ictal EEG recordings in four children with multiple independent spike foci. Almost all seizures were manifested by a tonic fencing posture. The ictal onset consisted of a generalized electrodecremental pattern in 48% followed by rhythmic activity such as repetitive sharp waves or spikes or repetitive theta activity either focally or over both hemispheres, or in 50% began with rhythmic activity either focal, hemispheric, or bihemispheric and in 2% of cases with a focal electrodecremental pattern. The head and arm movements associated with the seizures were stereotyped for each patient and did not vary with the site of ictal onset or the site of subsequent rhythmic activity. Three of these four patients subsequently underwent epilepsy surgery with improved seizure control in all three.

NONCONVULSIVE GENERALIZED STATUS EPILEPTICUS

Nonconvulsive generalized status epilepticus (NCGSE) is a state of altered mental status characterized chiefly by a slowing of behavior sometimes progressing to stupor, often with prominent psychiatric manifestations and with an EEG accompaniment of nearly continuous generalized epileptiform activity. The occurrence of status epilepticus in children with pyknoleptic petit mal is exceptionally rare. When it occurs in young persons with idiopathic generalized epilepsy, most will have generalized tonic-clonic seizures in addition to absence seizures (Niedermeyer and Khalifeh, 1965). It may also present without a prior history of seizures, particularly in middle to later life and in women. It is in this age group that psychiatric manifestations tend to be most common, sometimes resulting in admission to psychiatric services before the organic basis of the condition is recognized (Lee, 1985).

The ictal EEG manifestations are remarkably consistent across a number of different case series (Lee, 1985; Guberman et al., 1986; Dunne et al., 1987). Discharges consist of continuous, or else intermittent but frequent, generalized spike-wave or polyspike-wave discharges varying in frequency from 1 to 4 Hz. The discharges usually abate promptly with the administration of intravenous benzodiazepines along with a rapid return of the patient's mental status to baseline (Fig. 15).

FIG. 13. Recorded generalized clonic seizures in a 23-year-old man with generalized epilepsy secondary to early head trauma. Atypical absence seizures predominated in childhood, but exclusively generalized clonic seizures have occurred since adolescence. During briefer seizures, there is low-amplitude faciocervical clonus synchronously with higher-amplitude, symmetric somatic jerks, so that this unobscured EEG recording demonstrates generalized spike-wave complexes that are synchronous with the jerks.

FIG. 14. Recorded partial seizure in a patient with symptomatic generalized epilepsy, a 14-year-old boy who had herpes simplex encephalitis at age 9. Interictal EEG showed generalized spike-and-wave and multiple independent spike discharges. This seizure commenced with a generalized myoclonic jerk followed by a rhythmic interictal discharge in both temporal regions without any clinical accompaniment.
tems are often adequate to demonstrate generalized ictal patterns. Widespread myogenic ("muscle"), kinesogenic ("movement"), and other artifacts are a significant problem in recording generalized seizures (Fig. 2). Recording with greater numbers of electrodes can provide more sites that are not obscured by artifacts during some portion of a generalized seizure.

Difficulties in distinguishing interictal from ictal generalized EEG discharges have been noted in the preceding sections. Behavioral testing performed during and after discharges can often permit this distinction. Simultaneous audio and video monitoring during scalp EEG recording is required for adequate documentation of unresponsiveness and other aspects of ictal and postictal behavior. Behavioral testing during monitored events is usually most competently performed by EEG technologists or other medical professionals (Riley et al., 1980). Parents, spouses, and other companions often can be trained rapidly to perform adequate ictal and postictal behavioral testing. Behavioral testing during seizures should at a minimum include assessment of responsiveness to voice and touch, presentation of a memory item, and subsequent evaluation of memory for the item after any postictal behavioral alteration has resolved. It is also useful for the observer to record observations of ictal behaviors, including unelicited as well as tested behaviors.

Adequate documentation of behavior is not provided by ACR, because notes kept by patients and their companions may not be consistently reliable in documenting the quality of behavioral testing and often are not sufficiently objective in reported observations. ACR may be particularly useful for patients whose spells do not readily occur in the hospital environment and who have attentive parents or others to follow them about with a notebook in which to record behavioral observations. Even in the best of circumstances, uncertainty as to the precise temporal relationship of behavioral and ACR-recorded EEG changes is likely. Outpatient short-term and inpatient long-term EEG audio and video monitoring remain the techniques of choice for determining ictal behavioral-electrographic correlations.

Induction of seizures during monitoring is usually indicated in order to obtain a favorable number of seizures per day. Tapering of antiepileptic drugs (AEDs) is widely employed in order to induce epileptic seizures and perhaps also psychogenic pseudo-epileptic seizures in some individuals. Considerable knowledge of AED pharmacokinetics and pharmacodynamics is desirable in order to obtain diagnostically useful recordings safely. In particular, overly rapid withdrawal of barbiturates and benzodiazepines is capable of provoking epileptic seizures in persons who have never previously experienced epileptic seizures. Such withdrawal seizures are generalized from onset, so could lead to mistaken diagnosis of primary generalized epilepsies. Other means of physiological induction of epileptic seizures include sleep deprivation, hyperventilation, photic stimulation, and sleep induction. Despite all such measures, some patients will not have seizures during any reasonable period of monitoring.

REFERENCES
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