

Online brainmapping

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ROLE OF EEG IN EPILEPTIC SYNDROMES ASSOCIATED WITH MYOCLONUS

EEG is an essential component in the evaluation of epilepsy. The EEG provides important information about background EEG and epileptiform discharges and is required for the diagnosis of specific electroclinical syndromes. Such a diagnosis carries important prognostic information, guides selection of antiepileptic medication, and suggests when to discontinue medication.

EEG background frequencies - Following a seizure (ie, during the postictal period) the EEG background may be slow. However, interictal background EEG frequencies that are slower than normal for age usually suggest a symptomatic epilepsy (epilepsy secondary to brain insult). Normal background suggests primary epilepsy (idiopathic or possibly genetic epilepsy). Thus EEG background offers important prognostic and classification information.

Epileptiform discharges - They help separate generalized from focal (partial) seizures.

Epilepsy syndromes

1. Symptomatic epilepsy - Seizures resulting from an identifiable cerebral disorder
2. Cryptogenic epilepsy - Seizures occurring without identifiable cause in a patient with cognitive impairment or with neurological deficits. Examples of cryptogenic epilepsy include Lennox-Gastaut syndrome, infantile spasms, myoclonic astatic epilepsy of Doose
3. Idiopathic epilepsy - Seizures occurring in a patient without an identifiable cause and with an entirely normal neurological examination and of normal intelligence. Examples of idiopathic epilepsy include benign partial epilepsy of childhood with centrotemporal spikes; benign partial epilepsy of childhood with occipital paroxysms; juvenile myoclonic epilepsy

EEG characteristics of these specific electroclinical epilepsy syndromes will be discussed. Role of the EEG in temporal lobe epilepsy, frontal lobe epilepsy, etc will not be addressed here.

ROLE OF EEG IN NEONATAL SEIZURES

Generalized seizures are rare in neonates. Many of the so-called subtle, generalized tonic, and multifocal myoclonic seizures do not have an EEG correlate. These movements in the severely affected infant may represent brain stem release phenomena. Focal seizures, particularly clonic seizures, are highly associated with EEG changes. Thus EEG plays a crucial role in the evaluation of neonatal seizures. The EEG changes significantly with gestational age; therefore, calculation of gestational age and familiarity with age-specific norms is crucial in the interpretation of the EEG in infants.

There are 2 well-defined EEG seizure patterns seen in neonates. They include the following:

- Seizures with focal low frequency electrographic correlates - These may occur at 1-1.5 Hz frequency and are generally seen in severe cerebral insults, such as severe hypoxic-ischemic encephalopathy.
- Seizures with focal high frequency electrographic correlates - Typically evolve over 10-20 seconds and are usually seen with focal cerebral insults, such as strokes. Strokes in the neonate, unlike in the older individual, are typically associated with porencephalic cysts. Porencephalic cysts result from strokes that involve large portions of the cerebral parenchyma (i.e. loss of both gray and white matter leading to a communication between the subarachnoid space and the cerebral ventricles).

INFANTILE SPASMS & WEST SYNDROME

West syndrome is a triad of infantile spasms, developmental retardation or regression, and hypsarrhythmia on EEG. The syndrome presents between 6-18 months of age. Presence of a hypsarrhythmic EEG is diagnostic of infantile spasms. EEG patterns may evolve over a period; they initially appear in the sleep EEG record and subsequently present during the awake state. Hypsarrhythmia is seen in 75% of patients with West

syndrome. (click here to know more about west syndrome)

Hypsarrhythmia consists of diffuse giant waves (high voltage >400 microvolts) with a chaotic background of irregular, multifocal spikes and sharp waves. There is very little synchrony between the cerebral hemispheres. During sleep the EEG may display bursts of synchronous polyspikes and waves. There may be a pseudoperiodic pattern. Persistent slowing or epileptiform discharges in the hypsarrhythmic background may be present and may represent an area of focal dysfunction. There may be several variations to the hypsarrhythmic pattern, which are referred to as hypsarrhythmic variants. (click here to know more about hypsarrhythmia)

Clinical spasms are associated with a marked suppression of the background that lasts for the duration of the spasm. This characteristic response is called the "electrodecremental response."

EEG is useful in judging successful treatment of West syndrome. Typically, shortly after treatment with ACTH or vigabatrin is initiated, the spasms stop and hypsarrhythmia disappears.

Hypsarrhythmia rarely persists beyond the age of 24 months. It may evolve into the slow spike and wave discharges seen in Lennox-Gastaut syndrome.

Definition of Hypsarrhythmia

- The word Hypsarrhythmia is originally derived from the Greek word hypsolos which means high and it refers to high voltage arrhythmia with a disorganized EEG pattern that consists of chaotic admixture of continuous, multifocal, high amplitude spikes, polyspikes, sharp waves and arrhythmic slow waves. This EEG pattern is dynamic and highly variable from one patient to another and between one study and another study for a single patient. Background activity is often disorganized with frequent slow wave activity
- Marked change in the Hypsarrhythmia pattern also occurs during sleep. In REM sleep there is marked reduction to total disappearance of this EEG pattern. There is also normalization of this discharge pattern immediately following awakening from sleep.
- This discharge pattern is seen in children between the age of 4 months to 4 years and after the age of 4 years this pattern of discharge usually merges into the slow spike/slow wave complexes.
- Hypsarrhythmia pattern is frequently equated with infantile spasm (West syndrome), (characterized by massive flexion myoclonus of the head and neck called jack-knifing or Salaam attacks), however this pattern is not specific to any disease entity and is seen in response to any severe cerebral insult or severe multifocal disease process that occurs below the age of 1 year.
- Five different types of Hypsarrhythmia are present
 1. Hypsarrhythmia with increased interhemispheric synchronization.
 2. Asymmetrical Hypsarrhythmia.
 3. Hypsarrhythmia with a constant focus.
 4. Hypsarrhythmia with episodes of voltage attenuation.

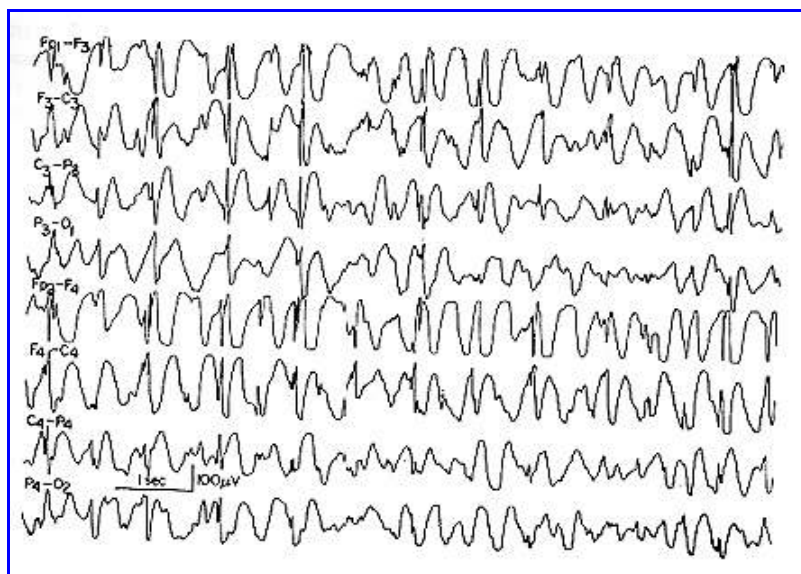


Figure 1. Hypsarrhythmia

LENNOX-GASTAUT SYNDROME (LGS)

Lennox-Gastaut syndrome is a childhood (onset 3-5 years) epileptic encephalopathy that manifests with atonic seizures, tonic seizures, and atypical absence seizures associated with mental retardation and a characteristic EEG pattern. Infantile spasms and West syndrome frequently transform into LGS. Unlike West syndrome, LGS tends to be a lifelong epileptic encephalopathy.

EEG shows an abnormally slow background and diffuse slow spike and wave (<2.5 Hz) activity. The slow spike and wave activity serves to differentiate (poor-prognostic) LGS from absence epilepsy, in which diffuse 3Hz spike and wave of benign absence is seen and the fast spike and wave (>2.5 Hz) activity often seen with some of the more benign myoclonic types of epilepsy. Prognosis of fast and slow spike and wave activity is dramatically different; it is poor for slow spike and wave activity seen in LGS. Many epilepsy syndromes overlap with LGS, including myoclonic astatic epilepsy of Doose and other severe myoclonic epilepsies.

EEG features of LGS may be divided into interictal and ictal.

- Interictal EEG features - Background slowing and diffuse slow spike and wave lasting from several minutes to a near continuous state are characteristic. Duration of epileptiform discharges tends to correlate with epilepsy control, with shorter durations occurring in patients with better control of seizures. Spikes, or more commonly sharp waves, are typically 200 milliseconds in duration and are followed by slow waves. Polyspike discharges are seen in those epilepsy variants with prominent myoclonic seizures or during non-REM sleep. (click here to know more about EEG in Lennox-Gastaut syndrome)
- Ictal EEG features - Electrographic accompaniment varies with the seizure type.

Electroclinical criteria of the slow 1-2.5c/s spike/wave discharge

- This EEG pattern is bilateral but asymmetrical and asynchronous with frequent lateralization and focalization.
- It has a frontal midline maximum.
- It is frequently continuous without any definite onset or offset and might extend through the whole record and is not associated with any clinical accompaniment.
- The discharge is not activated by hyperventilation
- The 1-2.5 c/s SWD is an age specific electrophysiological phenomenon. It usually start at the age of 6 months (earlier than the 3 c/s SWD) and disappear at the age of 16 years and is replaced by anterior temporal sharp activity and the clinical seizure manifestations merge into the main stream of temporal lobe epilepsies
- Background activity is often disorganized with frequent slow wave activity.
- The clinical correlate of this discharge is Lennox-Gastaut syndrome with multiseizure clinical presentation (grand mal fits, atonic fits, akinetic fits, atypical absence attacks, absence status). The occurrence of two or more than two types of seizures is almost the rule, mental retardation is very common.
- This discharge pattern could be idiopathic of genetic origin, cryptogenic with no overt cause , or symptomatic to a variety of brain diseases that include CNS infection, birth trauma, lipodosis, tuberous sclerosis, etc.

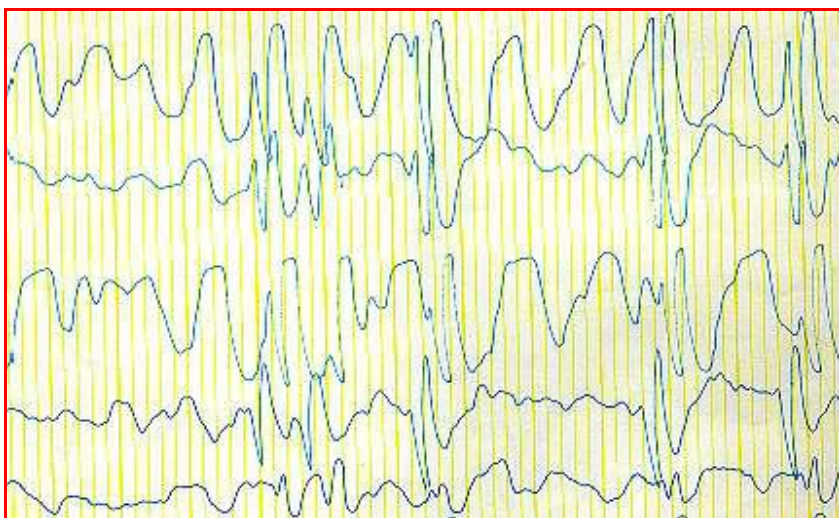


Figure 2. Slow 1-2.5c/s spike/wave discharge

JUVENILE MYOCLONIC EPILEPSY (JME)

JME is the most common epilepsy syndrome presenting with generalized tonic-clonic seizures between 12-30 years in a patient who is otherwise neurologically normal. It may account for up to 10% of all patients with epilepsy. Imaging studies are normal. In susceptible persons, sleep deprivation often precipitates seizures.

Typically, the patient may experience myoclonic jerks in the morning, although many patients do not mention that they are having myoclonic

seizures until asked specifically about body jerks.

Approximately 15% of patients have associated juvenile absence epilepsy or generalized tonic-clonic seizures upon awakening. Often the diagnosis is not made in a definitive fashion, which is unfortunate, since a correct diagnosis helps guide management, which, in turn, affects prognosis as the drugs used in this entity differ from those used in most other seizure types.

- EEG

Interictal EEG shows a normal background with frequent generalized polyspike and wave discharges that may be anteriorly dominant or diffuse. Polyspike and wave discharges by definition have at least 3 spike-like components in them.

Photosensitivity is present in at least 30% of patients. Photic stimulation, commonly at a frequency of 10-20 Hz, will elicit a photoparoxysmal

Electroclinical criteria of the fast 4-6 c/s spike/wave discharge

- This discharge occurs in patients older than 16 years.
- It is bilateral but less symmetrical and synchronous compared with the 3 c/s SWD and usually takes the morphological feature of polyspike wave discharge.
- It has a frontal midline maximum
- It has a sudden onset and sudden offset and lasts for a very short periods (usually less than 3 seconds)
- This discharge pattern is not activated hyperventilation, however phobic stimulation is a potent activator of this discharge pattern.
- The clinical correlate of this discharge pattern is myoclonus and grand mal fits (juvenile myoclonic epilepsy).
- Studies using video monitoring combined with EEG recording revealed that the spike components of this discharge coincide with the myoclonic jerks and the slow waves coincide with periods of relaxation between the myoclonic fits, accordingly the number of spikes in this polyspike/wave complexes were found to be proportional to the severity of the myoclonic fits.
- The fast spike/wave complexes of juvenile myoclonic epilepsy has a strong genetic background. The gene locus was mapped on the short arm of chromosome 6.

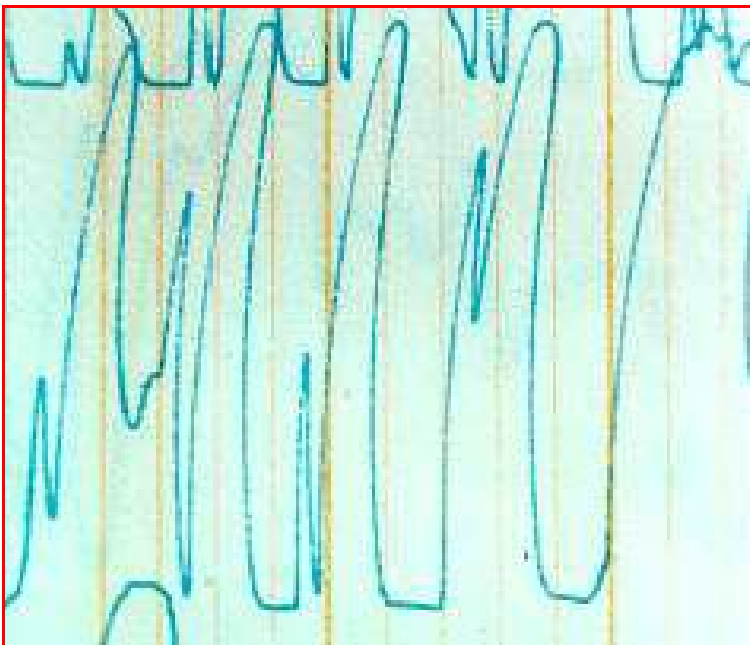


Figure 3. The fast polyspike/wave complexes

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