EEG in Childhood Epileptic Syndromes

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Awareness of Revision of Terminology & Classification

Communication
Article reviews
Further studies

EPILEPSY SYNDROMES AND RELATED CONDITIONS

- Benign familial neonatal seizures
- Early myoclonic encephalopathy
- Ohtahara syndrome
- * Migrating partial seizures of infancy
- West syndrome
- Benign myoclonic epilepsy in infancy
- Benign familial and non-familial infantile seizures
- Dravet's syndrome
- HH syndrome
- * Myoclonic status in nonprogressive encephalopathies
- Benign childhood epilepsy with centrotemporal spikes
- Early onset benign childhood occipital epilepsy (Panayiotopoulos type)
- Late onset childhood occipital epilepsy (Gastaut type)
- Epilepsy with myoclonic absences
- Epilepsy with myoclonic-astatic seizures
- Lennox-Gastaut syndrome
- Landau-Kleffner syndrome
- Epilepsy with continuous spike-and-waves during slow-wave sleep (other than LKS)
- Childhood absence epilepsy
- Progressive myoclonus epilepsies
- Idiopathic generalized epilepsies with variable phenotypes
  - Juvenile absence epilepsy
  - Juvenile myoclonic epilepsy
  - Epilepsy with generalized tonic-clonic seizures only
- Reflex epilepsies
  - Idiopathic photosensitive occipital lobe epilepsy
  - Visual sensitive epilepsies
  - Primary reading epilepsy
  - Startle epilepsy
- Autosomal dominant nocturnal frontal lobe epilepsy
- Familial temporal lobe epilepsies
- * Generalized epilepsies with febrile seizures plus
- * Familial focal epilepsy with variable foci
- Symptomatic (or probably symptomatic) focal epilepsies
  - Limbic epilepsies
    - Mesial temporal lobe epilepsy with hippocampal sclerosis
    - Mesial temporal lobe epilepsy defined by specific etiologies
    - Other types defined by location and etiology

Interim Organization ("Classification) of Epilepsies

Epilepsia, 51(4):676–685, 2010
doi: 10.1111/j.1528-1167.2010.02522.x

SPECIAL REPORT

Revised terminology and concepts for organization of seizures and epilepsies: Report of the ILAE Commission on Classification and Terminology, 2005–2009

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Interictal EEG & Clinical seizures

Interictal epileptiform pattern	Clinical seizure type
3 Hz spike-and-waves or polyspike-and-waves	Absences
Polyspike-and-waves, spike-and-waves, mono-and polyphasic sharp waves	Myoclonic seizures
Hypsarrhythmia & variants	Infantile spasms
Spike-and-waves or polyspike-and- waves	Clonic seizures
Slow spike-and-waves and other patterns	Tonic seizures
Spike-and-waves or polyspike-and- waves	Tonic-clonic seizures
Polyspike-and-waves or spike-and-waves	Atonic seizures
Polyspike-and-waves, spike-and-waves	Long atonic seizures
Polyspike-and-waves	Akinetic seizures

Primary Epilepsy Syndrome
"Primarily generalized seizure"

Absence epilepsy

Juvenile myoclonic epilepsy

Absence Epilepsy

- Absence seizure: a generalized, non-convulsive epileptic seizure predominantly disturbance of consciousness with relatively little or no motor activity with 3-Hz spike-wave bursts

EEG Findings in Absence Epilepsy

- Normal background
- Abrupt onset of synchronous spike-wave complex
- Frequency of complex: 3 Hz
- Induced by hyperventilation
- Associated clinical manifestation vary with duration of complex

Duration of Ictal Spike-wave

- CAE:
  - duration range 4 – 20 seconds

    < 4 or > 30: less likely to be CAE
  - Mean duration
    - 8 +/- 0.2 s (Hirsch et al)
    - 12 +/- 2.1 s (Panayiotopoulos et al 1989)
- JAE :
  - duration 16.3 +/- 7.1 s)

Interictal EEG

- Normal background, some may be slightly slow
- Paroxysmal of rhythmic slow wave activity 2.5 – 3.5 Hz in background or occipital region
- Synchronous burst of spike-and-wave complexes varies between beginning & later

Observation in Absence Epilepsy

1. Asymmetry of paroxysm or lateralization misleading to misinterpretation
2. Continuation or overlapping with juvenile myoclonic epilepsy results into inappropriate couselling

Asymmetry or Lateralization

- Misinterpretation of typical absence seizures as focal seizures, especially as temporal lobe seizures, is a relatively common error
- 3-Hz spike-waves may be founded in other epilepsy syndromes
- Variant absence epilepsy with focal discharge

Ferrie CD. Epilepsia 2005
Grosso S. Epilepsia 2005
Yoshinaga H. Seizure 2004

Ictal EEG

- Similar to interictal
  - Duration of burst of generalized 3 Hz spike-and-wave activity usually < 30 second
  - Clinical manifestation might not detectable, if duration less than 2.5 second

EEG in Absence Epilepsy

- Focal with synchronous 3-Hz spike-wave
  - 56% of 23 pts with absence epilepsy
    Yoshinaga H, Seizure 2004
  - 27% of 124 pts with typical absence epilepsy
    Covanis A, Seizure 1992
- A variant of absence epilepsy and is associated with refractory to initial treatment
  Ferrie CD. Epilepsia 2005
  Grosso S. Epilepsia 2005
  Yoshinaga H, Seizure 2004

EEG in childhood absence epilepsy

- Clinico-encephalographic study of 23 patients
- Classification according to response to treatment into 3 group
  - Group A: 8 patients who responded well to the therapy
  - Group B: 13 patients who suffered from relapse of epileptic discharges on EEG despite clinical seizure cessation
  - Group C: 2 patients who continue to suffered from seizure

Yoshinaga H, Seizure 2004

EEG in Childhood Absence Epilepsy

- 56% had focal epileptic discharges, including a surprising 63% of patients in group A
- "Lead-in" in the ictal EEGs and automatism were most common observed in the patients group B (not different in 3 group)
- 1 patient in group C evolved into complex partial seizure or absence status but the initial EEG did not show any abnormal focalities

Yoshinaga H, Seizure 2004

Absence Epilepsy: Early prognostic signs

- 27% of124 children with absence seizure had lateralized (frontal) spikes
- They could show that this represented a poor prognostic sign for relapse after withdrawal of therapy

Holmes D, Epilepsia 2004

Primary Epilepsy Syndrome "Primarily generalized seizure"

Absence epilepsy

Juvenile myoclonic epilepsy

Juvenile Myoclonic Epilepsy

- Sudden, mild to moderate myoclonic jerks(shoulder & arm) during awake, with secondary GTC
- Sleep deprivation, alcohol intake, fatigue
- Chromosome 6 , AD
- Onset 12 - 18 years (mean 14.6 years)
- Normal examination

Juvenile Myoclonic Epilepsy Interictal EEG Findings

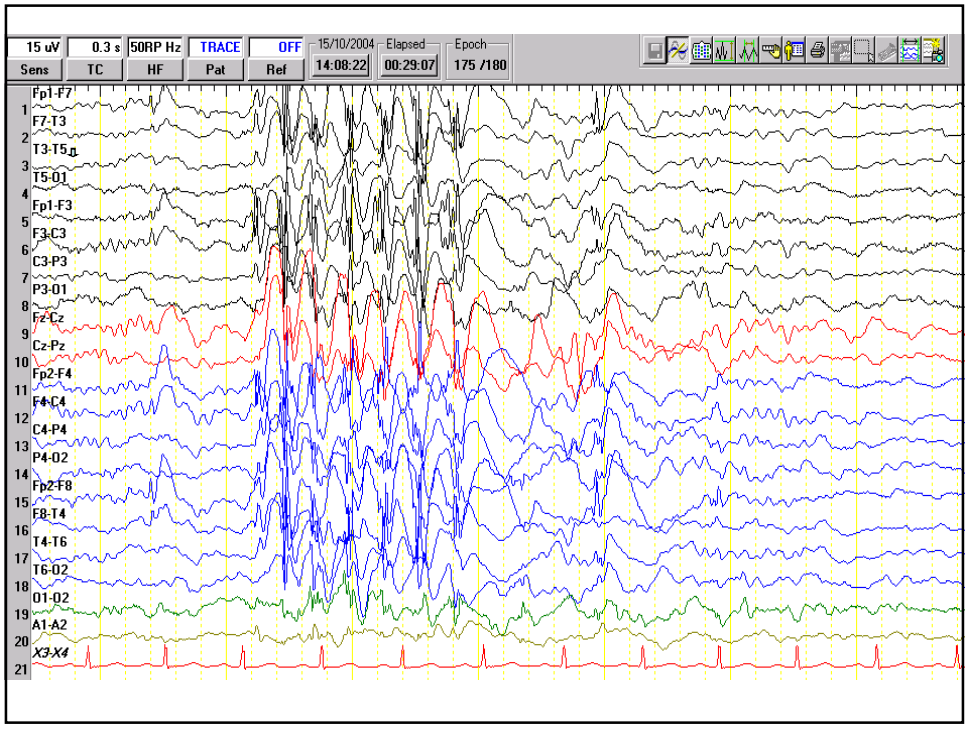
- Clusters of 3.5 - 4 Hz generalized spike-wave-complexes with preponderance to the frontal region
- Aftercoming slow waves after spike-wave complexes
- Normal background
- Photoparoxysmal response 30 %

Juvenile Myoclonic Epilepsy Ictal EEG Findings

- Myoclonic jerk in association with a burst of 3-4 Hz polyspike-and-wave acitivity
- Burst of spikes from 10 – 16 Hz during ongoing jerking movement
- Slow waves after polyspike-and-wave between 2 – 2.5 Hz

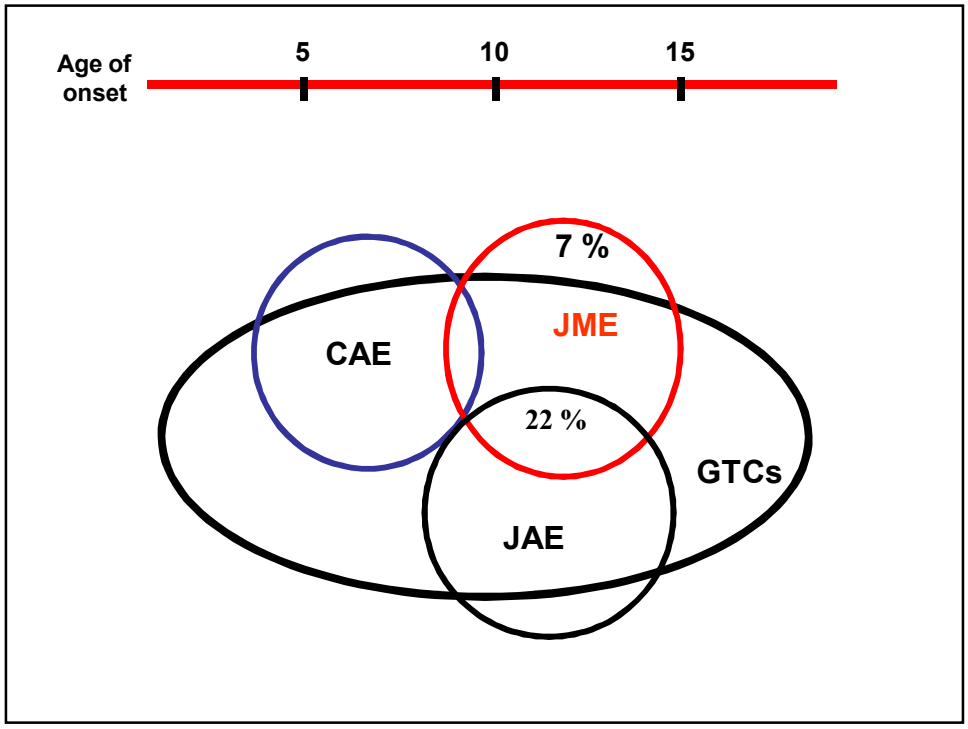
Focal semiolgic or EEG features in JME

- Not uncommon
- 38 - 54%
  - Montalenti E, et al. J Neurological Sc 2001
  - Usni N, et al. Epilepsia 2005
- 56%
  - A cohort study of mixed primary generalized epilepsies. Lombroso CT. Epilepsia 1997



Comparison of Absence and JME

	Childhood	Juvenile	JME
Age of onset	2 -12 yrs	puberty	puberty
Frequency	multiple/D	rarely/D	variable
EEG	3 Hz.S+W	3.5-4 Hz S+W	3.5-6 Hz.S+W
GTC	40-60%	80%	80 - 85 %
AED	ETH, VPA	VPA	VPA
Prognosis	favorable	favorable	favorable



Benign Epilepsy of Childhood with Centrotemporal Spikes

Benign partial childhood epilepsy with centrotemporal discharges/spikes

- **Characteristics:**
  1. onset between 2 and 14 years (3 - 10)
  2. simple partial motor seizure
  3. characteristic EEG foci over rolandic (centrotemporal region) with normal posterior dominant rhythm
- **Most common partial epilepsy**
  - 15.7 % of epilepsy before 15 years old
  - 24 % of epilepsy with onset 5 - 14 years

Infantile Spasms & Hypsarrhythmia

Infantile Spasms

- Symmetric, salaam-like contractions of trunk, with extension and elevation of arms, and tonic extension of legs
- Initial brief phasic contraction followed by gradually relaxing tonic component
- Lasts for less than 1- 5 seconds, with clusters of 3-more than 100 spasms
- Occurring several clusters per day

Infantile Spasms Movie

- Rarely occur during sleep
- Activated after arousal from sleep
- Occasionally triggered by loud noises with associated arousal from drowsiness
- Not sensitive to photic stimulation
- 1/3 – ½ have other seizure types preceding or accompanying the onset of spasms

Infantile Spasms: EEG features

Interictal pattern: hypsarhythmia

"Complete chaotic and disorganized background pattern consisting of high amplitude slow waves and spikes that are asynchronous, non-rhythmic, and variable in duration and topography (focal, multifocal, generalized) ''

Infantile Spasms: EEG features

Interictal pattern: hypsarhythmia

- Most pronounced in slow- wave sleep
- Diminished or completely suppressed during REM
- May absent during awake
- Disappear on arousal from sleep
- Disappear during spasms

Infantile Spasms: EEG features

Ictal pattern

- Generalized sharp or slow waves
- Generalized voltage attenuation (electrodecremental discharges, most common ~70%)
- Fast burst activity

Variants or atypical Hypsarrhythmia

1. Hyparrhtymia with increase interhemispheric synchronization
2. Asymmetrical hypsarrhythmia
3. Hypsrrhythmia with episodes of generalized or lateralized voltage attentuation
4. Hypsarrhythmia with a consistent focus of spike or sharp wave activity
5. Hypsarrhythmia with little or no spike or sharp wave

Hrachovy RA, Frost JD. J Clin Neurophysiol 2006;23:312-32

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