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
- Olmsted syndrome
- Infantile spasms
- Infantile neuronal migration defects
- Benign myoclonic epilepsy in infancy
- Benign familial and non-familial infantile seizures
- Dravet's syndrome
- HH-syndrome
- Hydantoin status in nonprogressive encephalopathies
- Benign childhood epilepsy with centrotemporal spikes
- Early onset benign childhood occipital epilepsy (Phenylketonuria type)
- Late onset childhood occipital epilepsy (Gauchet type)
- Epilepsy with myoclonic absences
- Epilepsy with myoclonic-atatic seizures
- Lennox-Gastaut syndrome
- Landau-Kleffner syndrome
- Epilepsy with continuous spike-wave during slow-wave sleep
- Childhood absence epilepsy
- Progressive myoclonic epilepsies
- Idiopathic generalized epilepsies with variable phenotypes
  - Juvenile absence epilepsy
  - Juvenile myoclonic epilepsy
  - Epilepsy with generalized tonic-clonic seizures only
- Febrile seizures
  - Infraorbital photosensitive occipital lobe epilepsy
  - Visual sensitive epilepsies
  - Primary reading epilepsy
  - Stutter 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 syndromes
    - Other forms defined by location and childhood
### Interictal EEG & Clinical seizures

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### 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

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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 counselling

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
    Covaris 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
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

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% of 124 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 activity
- Burst of spikes from 10 – 16 Hz during ongoing jerking movement
- Slow waves after polyspike-and-wave between 2 – 2.5 Hz

Focal semiologic 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

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<tr>
<th></th>
<th>Childhood</th>
<th>Juvenile</th>
<th>JME</th>
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<td><strong>Age of onset</strong></td>
<td>2 - 12 yrs</td>
<td>puberty</td>
<td>puberty</td>
</tr>
<tr>
<td><strong>Frequency</strong></td>
<td>multiple/D</td>
<td>rarely/D</td>
<td>variable</td>
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<tr>
<td><strong>EEG</strong></td>
<td>3 Hz.S+W</td>
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<td><strong>GTC</strong></td>
<td>40-60%</td>
<td>80%</td>
<td>80 - 85 %</td>
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**Comparison of Absence and JME**

- **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
- **Prognosis**: favorable
Benign Epilepsy of Childhood with Centrottemporal 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

- 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 attenuation
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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