Idiopathic epilepsy syndromes

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Childhood absence epilepsy (CAE)

- Age: onset between 4-10 yrs (peak 5-6)
  (range 2-13 /1-14 yrs, peak 6-7 yrs)
- Sex: G>B (66%)
- Development: normal
- Genetic: unknown but ? Multifactorial
- FHx of epilepsy ~ 15-45% of cases

Ictal EEG 3 Hz spike/wave discharges
Absence

Hyperventilation

Factors influencing clinical features of absence seizure

**4 Major types of Absences**

1. Typical absence
2. Atypical absence
3. Myoclonic absence
4. Eyelid myoclonia with (and) absence (EMA)

**CAE: prognosis**

- Excellent prognosis, remission before age of 12 years
- <10% may develop infrequent GTC in the adult life: poor adjustment behaviour
- Better select proper antiepileptic medication
Differential diagnosis CAE

1. Complex partial seizure
2. Juvenile absence epilepsy
3. Juvenile myoclonic epilepsy
4. Eyelid myoclonia with absence
5. Myoclonic absence epilepsy
6. Non-epileptic manifestation; day-dreaming, attention disturbance

Juvenile absence epilepsy (JAE)

- Age: 9-13 yrs (range 5-20 yrs)
- Sex: F=M
- Development: normal
- Genetic: may linked to 8, 21, 18, 5

JAE

Seizure

Absence

80% GTC: 20% mild myoclonic

JAE

Seizure

Absence

80% GTC: 20% mild myoclonic

mainly after awakening

JAE

Seizure

Absence

80% GTC: 20% mild myoclonic

follow onset of absence

JAE

Seizure

Absence

80% GTC

20% mild myoclonic

afternoon when tired
**JAE**: prognosis

- Sz can be controlled in 70-80% of patients
- Absences become less severe in terms of impairment of cognition, duration and Fq with age
- GTC: infreq but precipitated by sleep deprivation, fatigue and alcohol consumption
- Myoclonic jerks are not problematic

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**Juvenile myoclonic epilepsy (JME)**

- Age: 2nd decade of life (range 8-24 yrs)
- Sex: equal but female has less Sz threshold
- Development: mentally and neurologically normal
- Genetic: familial; polygenic/? chro 6

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**DDx of JAE**

- Vs. CAE
  - overlap, age in JAE is later and less frequent, less severe impairment of cognition.
  - Automatism is equal. No myoclonic and GTC in CAE
- Vs. EMA
- Vs. JME

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**Main inclusion and exclusion criteria for JAE**

- Inclusion criteria for JAE:
  - Unprovoked clinical evidence of absence seizures with severe impairment of consciousness. Nearly all patients may have GTCs. A few have myoclonic jerks, but these are mild and do not show the circadian distribution of JAE.
  - Documentation of ictal EEG: GTCs, or loss that are associated with severe impairment of consciousness and often with automatisms. Normal EEG is treated patients are common

- Exclusion criteria for JAE:
  - The following may be incompatibility with JAE:
    - Absence with readily recoverable or rarely myoclonic or normal single or rhythmic tonic and tonic myoclonic jerks
    - Absence with exclusively mild or slightly automatized impairment of consciousness
    - Consistent visual, phosphenie or other sensory perception of seizures (absences) is probably against the diagnosis of JAE. However, in many cases intermittent photoelectric or other facilitation-related discharge and absences
  - EEG-elimination criteria:
    - Irregular aminotonic (SPWDM) with marked variations of the intradischARGE frequency
    - Significant relations between the spikes/complex and slow waves in SPWDM
    - Reappearance of mild discharges (≤ 4 /s)
JME

Seizure types

Myoclonic sz  GTC  Absence Sz

around 14-15 yrs  around 5-16 yrs

followed or preceded by myoclonic

JME

Seizure types

Myoclonic sz  GTC  Absence Sz

mild/mod invl. neck, shoulder, arms

isolate, repetitive/bilateral, asymmetric

aggravated by sleep deprivation

After awakening from a night sleep or a nap

myoclonic status epilepticus

JME

Seizure types

Myoclonic sz  GTC  Absence Sz

majority, the onset precede by myoclonic

Shortly after awakening

JME

Seizure types

Myoclonic sz  GTC  Absence Sz

40% of patient

associated with GTC

after awakening

if begin before 10 yrs-more severe

JME EEG

Interictal EEG
- Irregular fast 3.5-6 Hz SW
- GPSW: ant predominant
- intra-discharge fragment
- 1/3: focal abn, spike, sv, slow waves
- 1/3 PPR

Ictal EEG
- Myoclonic sz
  - fast 10-16 Hz spikes followed by irregular slow waves (PSW), - 0.5-2 sec
- Absence sz
  - multiple spikes preceding on slow wave, last 1-4 sec
**JME EEG**

**Interictal EEG**
- Irregular fast 3.5-6 Hz SW
- GPSW: ant predominant
- Intra-discharge fragment
- 1/3 focal abn, spike, SW, slow waves
- 1/3 PPR

**Ictal EEG**
- Myoclonic sz
  - Fast 10-16 Hz spikes followed by irregular slow waves (PSW)
- Absence sz
  - Multiple spikes preceding on slow wave, last 1-4 sec

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**Key differences between JME and JAE**

<table>
<thead>
<tr>
<th>JME</th>
<th>JAE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Main type of seizure</td>
<td>Myoclonic sz</td>
</tr>
<tr>
<td>Circadian distribution</td>
<td>Mainly on awakening</td>
</tr>
<tr>
<td>Typical absences</td>
<td>Unilateral and often inaudible, they occur in a third of patients and occur in all patients</td>
</tr>
<tr>
<td>Myoclonic absences</td>
<td>Myoclonic absences usually occur in a third of patients and are usually not seen in all patients</td>
</tr>
<tr>
<td>Spikes</td>
<td>Absence seizures usually occur after a series of myoclonic seizures on awakening</td>
</tr>
<tr>
<td>ECG</td>
<td>Bilateral 3-5 Hz PSW, which are usually asymptomatic</td>
</tr>
</tbody>
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**Benign childhood focal epilepsies**
- Rolandic epilepsy (BRE)
  : Benign childhood epilepsy c centro-temporal spikes (BECTS)
  : Benign focal epilepsy of childhood (BFEC)
- Panayiotopoulos syndrome (PS)
- Idiopathic childhood occipital epilepsy of Gastaut (ICOE-G)

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**Benign rolandic epilepsy**
- Age: 3-13 years (peak 7-8yrs of age)
- Sex: Boys > Girls
- Development: normal
- Genetic: familial, linked to Chromosome 15 q
  : 50% of close relatives have EEG abnormalities between the ages of 3-15 yr
  : 12% of persons whom EEG abnormal have clinical seizure.
**BRE**

**Seizure**

- >50% Nocturnal only
- 15% Sleep and awake
- 10-20% During awake

**Interictal EEG in BRE**

- Spike/wave discharges
- Triphasic follow by after coming slow wave
- The complex lasts for 80-120 seconds
- Unilateral discharges in 70% of patients, independent & asynchronous

**CTS are not specific to Rolandic sz**

- 2-3% of normal school-aged children (< 10% develop rolandic sz)
- Non-epileptic children with various symp eg. headache, speech and learning difficulty
- Occur in a variety of organic brain diseases with or without sz eg. tumors, Rett’s synd, focal cortical dysplasia
- Common among relatives
Benign childhood focal epilepsies

- Rolandic epilepsy (BRE)
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- Panayiotopoulos syndrome (PS)
- Idiopathic childhood occipital epilepsy of Gastaut (ICOE-G)

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<thead>
<tr>
<th>BRE</th>
<th>Ps</th>
<th>ICOE-G</th>
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<tbody>
<tr>
<td>Duration for 1-3 min</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Duration &gt; 5 mins</td>
<td>Rare</td>
<td>Common</td>
</tr>
<tr>
<td>Partial status</td>
<td>no</td>
<td>40%</td>
</tr>
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<tr>
<th>BRE</th>
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<th>ICOE-G</th>
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<tr>
<td>Prev amongst children age 1-15 yrs</td>
<td>15%</td>
<td>6%</td>
</tr>
<tr>
<td>Range of age (yrs)</td>
<td>1-14</td>
<td>1-14</td>
</tr>
<tr>
<td>Peak age at onset (yrs)</td>
<td>7-10</td>
<td>3-6</td>
</tr>
<tr>
<td>Event Description</td>
<td>BRE</td>
<td>PS</td>
</tr>
<tr>
<td>-------------------</td>
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<td>----</td>
</tr>
<tr>
<td>Single sp only</td>
<td>10-12%</td>
<td>30%</td>
</tr>
<tr>
<td>Frequent sp</td>
<td>10%</td>
<td>10%</td>
</tr>
<tr>
<td>Nocturnal (sleep only)</td>
<td>70%</td>
<td>64%</td>
</tr>
<tr>
<td>Spikes after age of 13</td>
<td>rare</td>
<td>exceptional</td>
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<th>Event Description</th>
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| Occipital spikes  | Yes | Rare | 65% | Not reported 90%
| Spikes in other location | Not reported | Exceptional | 20-30% |
| Photopsisensitivity | Nocturnal region | Rolandic region | Am and post regions | Occipital region |