**Idiopathic epilepsy syndromes**

Kamornwan Katanuwong MD  
Chiangmai University Hospital  
1st Epilepsy Camp, Hua Hin  
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**Idiopathic Epileptic Syndromes**

- Greek words "idios" = self, own and personal  
  "pathic" = suffer

- Is a syndrome that is only epilepsy, with no underlying structural brain lesion or other neurological signs or symptoms.

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**Outline**

- Idiopathic partial epilepsy syndromes  
- Idiopathic generalized epilepsy syndromes

- Idiopathic / symptomatic / cryptogenic  
- Focal seizures / generalized seizures

- Modified concepts to replace the above...

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**Idiopathic epilepsy syndromes**

- Presumed to be genetic and usually age dependent

- Idiopathic is not synonymous with benign

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**Terminology and classification**

![Diagram showing the classification of localized and generalized epilepsy with time lines from 1960 to 2010.]
**Childhood absence epilepsy (CAE)**

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

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**4 Major types of Absences**

1. Typical absence seizure (TAS)
2. Atypical absence (more in SGE; LGS)
3. Epilepsy with myoclonic absences (MAE) (myoclonic absence epilepsy)
4. Eyelid myoclonia with absence (EMA)
Interictal-normal B/G and OIRDA

CAE: prognosis

- 1/3 of typical absence sz may have absence status epilepticus
- 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

Juvenile absence epilepsy (JAE)

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

Main differences between atypical and typical absence seizures

<table>
<thead>
<tr>
<th>Clinical and EEG features</th>
<th>Atypical absence</th>
<th>Typical absence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset and termination</td>
<td>Usually gradual</td>
<td>Abrupt</td>
</tr>
<tr>
<td>Responsiveness</td>
<td>Decreased but not abolished</td>
<td>Varies from mild to severe</td>
</tr>
<tr>
<td>Duration</td>
<td>Usually prolonged</td>
<td>Usually brief; never &gt;30-40 s</td>
</tr>
<tr>
<td>Post-ictal recovery</td>
<td>Cognitive impairment may persist</td>
<td>Immediately</td>
</tr>
<tr>
<td>Inter-ictal EEG</td>
<td>Background often abnormal with thalamic discharges of various types and combinations</td>
<td>Background rapidly normal sometimes with typical IGE discharges</td>
</tr>
<tr>
<td>IGE EEG</td>
<td>High (&lt;2.5 Hz) spike and wave</td>
<td>Low (&lt;2.5 Hz) spike and slow waves</td>
</tr>
<tr>
<td>Normal neurological and mental state</td>
<td>Exceptional</td>
<td>As a rule</td>
</tr>
<tr>
<td>Other types of seizure</td>
<td>Commonly atomic and tonic seizures of symptomatic generalized epilepsies</td>
<td>Depend on IGE syndrome (myoclonic jerks, GTCS or both)</td>
</tr>
<tr>
<td>Prognosis</td>
<td>Commonly bad</td>
<td>Commonly good</td>
</tr>
</tbody>
</table>

JAE

Seizure

Absence

80%: GTC

20%: mild myoclonic
JAE

Seizure

Absence

GTC

myoclonic

duration 4-30 sec (~16s)

freq automatism, 6-10 sec after EEG onset

mild myoclonic of the eyelids

absence status: 20%

Main inclusion and exclusion criteria for JAE

Inclusion criteria for JAE:
- Unilateral 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 clinical distribution of JAE.
- Documentation of at least 2-4 Hz GPSIAW, >4 s, that are associated with severe impairment of consciousness and often with automatisms. Normal EEG in treated patients are common.

Exclusion criteria for JAE:
- The following may be incompatibility with JAE:
  - Clinical exclusion criteria:
    - Absolotes with marked eyelid or palpebral myoclonus or marked single or rhythmic limb and trunk myoclonic jerks
    - Absolotes with exclusively mild or definitely undetectable impairment of consciousness
    - Consistent visual, photomotor, and other sensory precipitant of clinical absences is probably against the diagnosis of JAE. However, on the EEG, intermittent photic stimulation often facilitates generalized discharges and absences.
    - EED exclusion criteria:
      - Irregular, arrhythmic GPSIAW with marked variations of the interdischarge frequency
      - Significant variations between the spiky polyspike and slow wave relations in GPSIAW
      - Preponderantly focal discharges (>4)

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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

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

JAE: prognosis

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

Seizure types

- Myoclonic sz
- GTC
- Absence Sz

- Majority, the onset precede by myoclonic
- Shortly after awakening

40% of patient associated with GTC after awakening or...
if begin before 10 yrs-more severe

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
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)

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 5-15 yr
  : 12% of persons whom EEG abnormal have clinical seizure.
Interictal EEG in BRE

- Spike/wave discharges
  - triphasic follow by after coming slow wave
  - the complex lasts for 80-120 m-seconds
  - unilateral discharges 70%
  - bilateral discharges in 30% 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

Ictal EEG in BRE
Benign childhood focal epilepsies

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<thead>
<tr>
<th>BRE</th>
<th>PS</th>
<th>ICOE-G</th>
</tr>
</thead>
<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>
</tbody>
</table>

Prevalence amongst children age 1-15 yrs

<table>
<thead>
<tr>
<th></th>
<th>BRE</th>
<th>PS</th>
<th>ICOE-G</th>
</tr>
</thead>
<tbody>
<tr>
<td>Single sz only</td>
<td>10-20%</td>
<td>30%</td>
<td>exceptional</td>
</tr>
<tr>
<td>Frequent sz</td>
<td>10%</td>
<td>10%</td>
<td>90%</td>
</tr>
<tr>
<td>Nocturnal (sleep only)</td>
<td>70%</td>
<td>64%</td>
<td>exceptional</td>
</tr>
<tr>
<td>Sz after age of 13</td>
<td>rare</td>
<td>exceptional</td>
<td>common</td>
</tr>
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Typical onset

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<thead>
<tr>
<th>BRE</th>
<th>PS</th>
<th>ICOE-G</th>
</tr>
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<tbody>
<tr>
<td>Hemifacial sensory-motor or Oro-pharyngolaryngeal symptoms</td>
<td>Autonomic symptoms mainly emesis</td>
<td>Visual symptoms mainly with elementary visual hallucination</td>
</tr>
</tbody>
</table>

EEG

<table>
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<th>BRE</th>
<th>PS</th>
<th>ICOE-G</th>
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<tbody>
<tr>
<td>CTS alone Occipital spikes</td>
<td>Yes</td>
<td>Rare</td>
<td>65%</td>
</tr>
<tr>
<td>Spikes in other location</td>
<td>Uncommon</td>
<td>Frequent</td>
<td>Exceptional</td>
</tr>
<tr>
<td>Photosensitivity</td>
<td>No</td>
<td>Exceptional</td>
<td>20-30%</td>
</tr>
<tr>
<td>Ictal onset Rolandic regions</td>
<td>Ant' /Post' regions</td>
<td>Occipital regions</td>
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New concept
- Focal vs Generalized seizure
Old
New concept

• Focal vs Generalized seizure

There is no absolute distinction between generalized and focal epilepsies, but rather a continuum.

However

The terminology of focal and generalized seizure should be continued due to the direct impact on management decisions:
Pt with focal epilepsy are good candidates for epilepsy surgery

New concept

• Idiopathic/ symptomatic/cryptogenic
• Genetic
• Structural / metabolic diseases
• Unknown cause

Terminology and classification

Thank you for your attention