Idiopathic epilepsy syndromes

Kamornwan Katanyuwong MD.
Chiangmai University Hospital
EST, July 2009

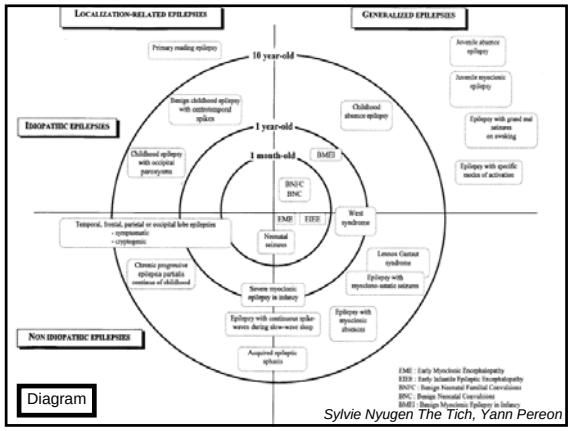
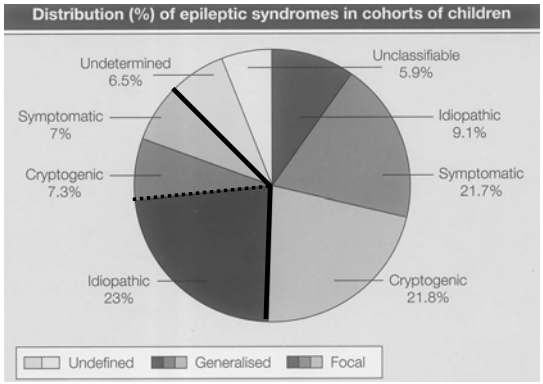
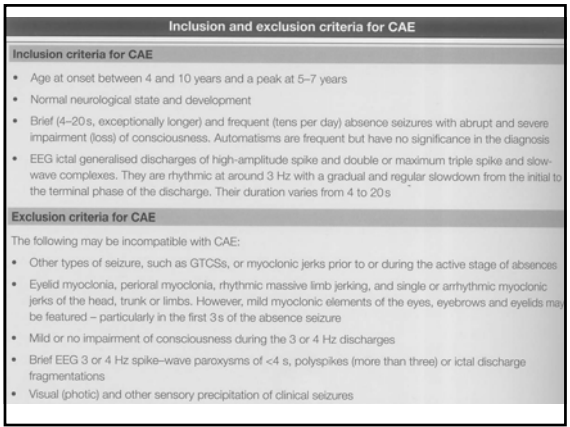


Diagram
Sylvie Nyugen The Tich, Yann Pereon

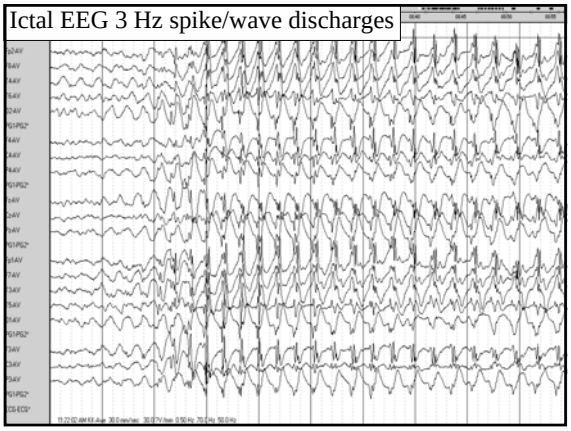


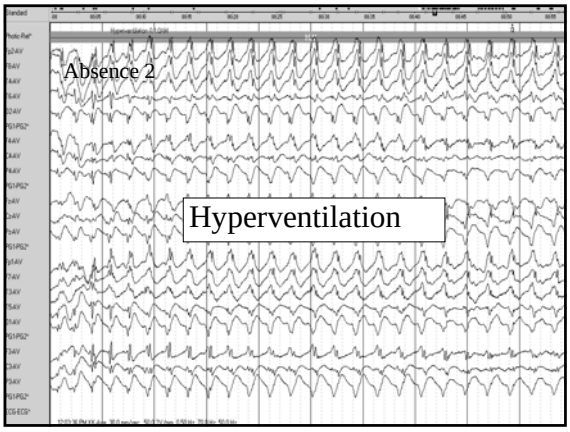
Childhood absence epilepsy (CAE)

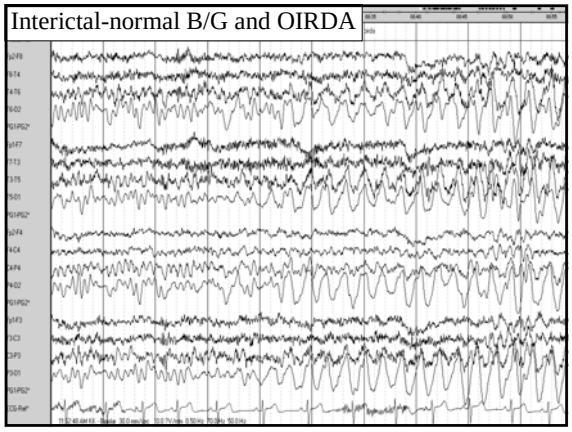
- Age : onset between 4-10 yrs (peak 5-6)
  (range 2-13 /1-14 yrs, peak 6-7 yrs)
- Sex : G>B (66%)
- Development : normal
- Genetic : unknown but ? Multifactorial
- FHx of epilepsy ~ 15-45% of cases



Ictal EEG 3 Hz spike/wave discharges







Factors influencing clinical features of absence seizure

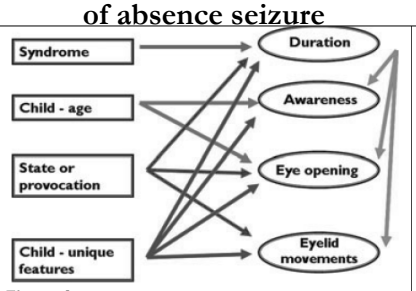


Figure 6.
Summary of associations found between the child, their seizures' characteristics, and the clinical features of absence seizures.

Epilepsia © ILAE *Epilepsia, 49(12): 2100–2107,2008*

Main differences between atypical and typical absence seizures

Clinical and EEG features	Atypical absences	Typical absences
Onset and termination	Usually gradual	Abrupt
Responsiveness	Decreased but not abolished	Varies from mild to severe
Changes in tone	Usually pronounced	Usually mild
Duration	Usually long sometimes for minutes	Usually brief; never >30–40 s
Post-ictal recovery	Cognitive impairment may persist	Immediately
Inter-ictal EEG	Background often abnormal with frequent discharges of various types and combinations	Background usually normal sometimes with typical IGE discharges
Ictal EEG	Slow (<2.5 Hz) spike and wave	**Fast**->2.5 Hz) spike and slow wave
Normal neurological and mental state	Exceptional	As a rule
Other types of seizure	Commonly atonic and tonic seizures of symptomatic generalised epilepsies	Depend on IGE syndrome (myoclonic jerks, GTCS or both)
Prognosis	Commonly bad	Commonly good

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

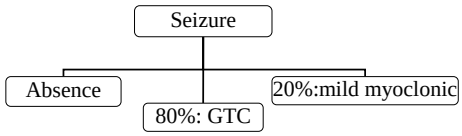
Differential diagnosis CAE

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

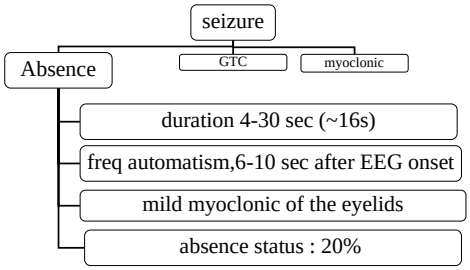
Juvenile absence epilepsy (JAE)

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

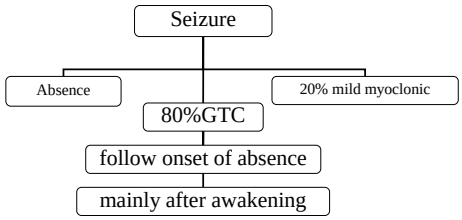
JAE



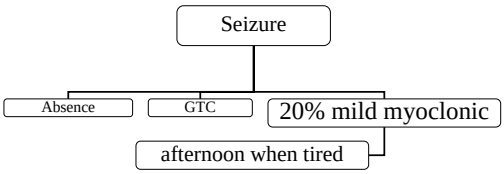
JAE



JAE



JAE



Main inclusion and exclusion criteria for JAE

Inclusion criteria for JAE

- Unequivocal clinical evidence of absence seizures with severe impairment of consciousness. Nearly all patients may have GTCSs. A fifth have myoclonic jerks, but these are mild and do not show the circadian distribution of JME
- Documentation of ictal 3–4 Hz GPSWD, >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

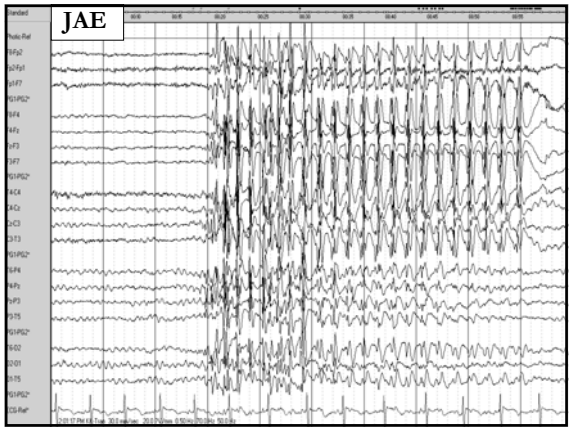
The following may be incompatible with JAE

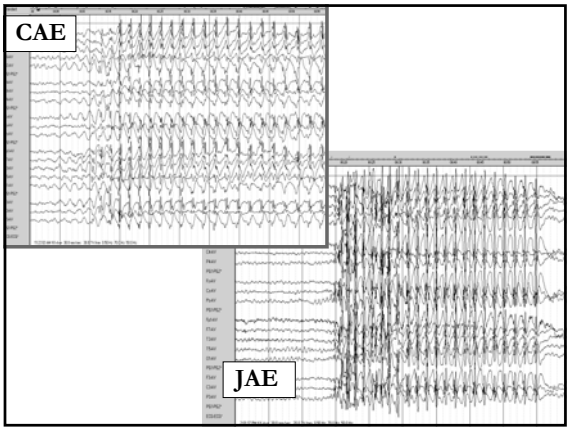
Clinical exclusion criteria:

- Absences with marked eyelid or perioral myoclonus or marked single or rhythmic limb and trunk myoclonic jerks
- Absences with exclusively mild or clinically undetectable impairment of consciousness
- Consistent visual, photosensitive and other sensory precipitation of clinical absences is probably against the diagnosis of JAE. However, on the EEG, intermittent photic stimulation often facilitates generalised discharges and absences

EEG exclusion criteria:

- Irregular, arrhythmic GPSWD with marked variations of the intradischarge frequency
- Significant variations between the spike/polyspike and slow wave relations in GPSWD
- Predominantly brief discharges (<4 s)





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 Fq with age
- GTC: infreq but precipitated by sleep deprivation, fatique and alcohol consumtion
- Myoclonic jerks are not problematic

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

JME

Seizure types

- Myoclonic sz
  - around 14-15 yrs
- GTC
  - followed or preceded by myoclonic
- Absence Sz
  - around 5-16 yrs

JME

Seizure types

- Myoclonic 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
- GTC
- Absence Sz

JME

Seizure types

- Myoclonic sz
- GTC
  - majority, the onset precede by myoclonic
  - Shortly after awakening
- Absence Sz

JME

Seizure types

- Myoclonic sz
- GTC
- Absence Sz
  - 40% of patient
  - associated with GTC
  - after awakening
  - if begin before 10 yrs-more severe

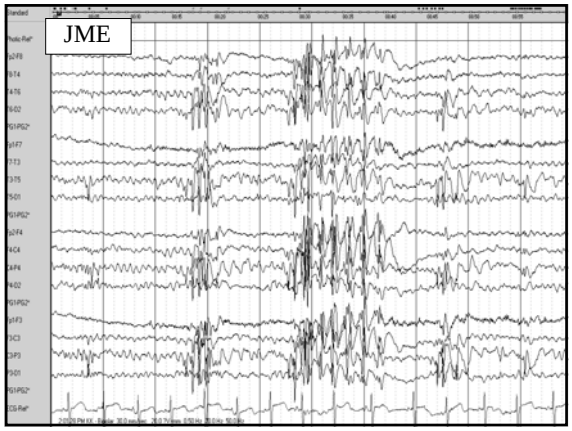
JME EEG

Interictal EEG

- Irregular fast 3.5-6 Hz SW
- GPSW: ant predominant
- intra-discharge fragment^{n}
- 1/3 : focal abn, spike, sw, slow waves
- 1/3 PPR

Ictal EEG

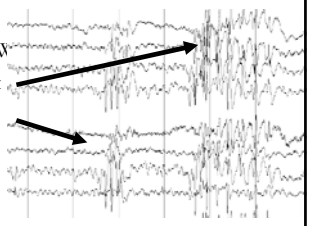
- myoclonic sz
  - fast 10-16 Hz spikes followed by irregular slow waves (PSW),
  - 0.5-2 sec
- absence sz
  - multiple spikes preceding on slow wave, last1-4 sec



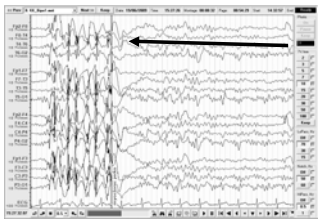
JME EEG

Interictal EEG

- Irregular fast 3.5-6 Hz SW
- GPSW: ant predominant
- intra-discharge fragment[n]
- 1/3 focal abn, spike, sw, slow waves
- 1/3 PPR



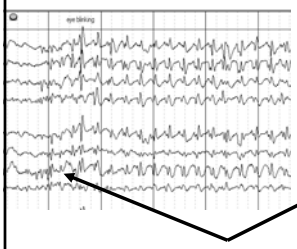
JME EEG



Ictal EEG

- myoclonic sz
  - fast 10-16 Hz spikes followed by irregular slow waves (PSW)
- absence sz
  - multiple spikes preceding on slow wave, last1-4 sec

JME EEG



Ictal EEG

- myoclonic sz
  - fast 10-16 Hz spikes followed by irregular slow waves (PSW)
- absence sz
  - multiple spikes preceding on slow wave, last1-4 sec

Key differences between JME and JAE

	JME	JAE
Main type of seizures	Myoclonic jerks	Typical absences
Circadian distribution	Mainly on awakening	Any time during the day
Typical absences	Mild and often imperceptible; they occur in a third of patients	Defining seizure type; they are very severe and occur in all patients
Myoclonic jerks	Defining seizure type; they occur in all patients and mainly on awakening	Mild; they occur in a fifth of patients and are random
GTCS	They mainly occur after a series of myoclonic jerks on awakening	They mainly occur independently or less commonly after a series of absence seizures
EEG	Brief (1–3s) 3–6 Hz GPSWD, which are usually asymptomatic	Lengthy (8–30s) 3–4 Hz GPSWD, which are usuall associated with severe impairment of consciousne

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)

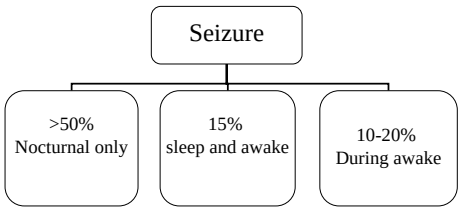
Benign rolandic epilepsy

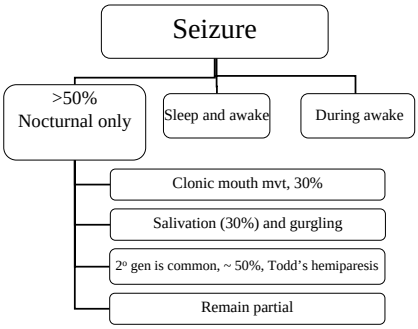
- Age: 3-13 years (peak 7-8yrs of age)
- Sex: Boys > Girls
- Development: normal
- Genetic: familial, linked to Chromosome 15 q

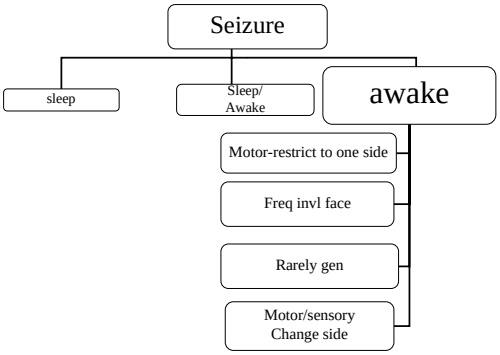
  : 50% of close relatives have EEG abnormalities between the ages of 5-15 yr

  : 12% of persons whom EEG abnormal have clinical seizure.

BRE

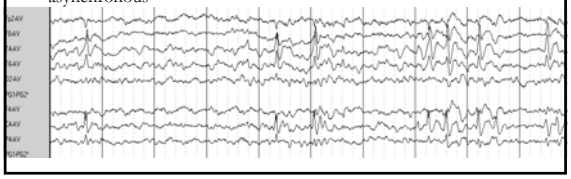






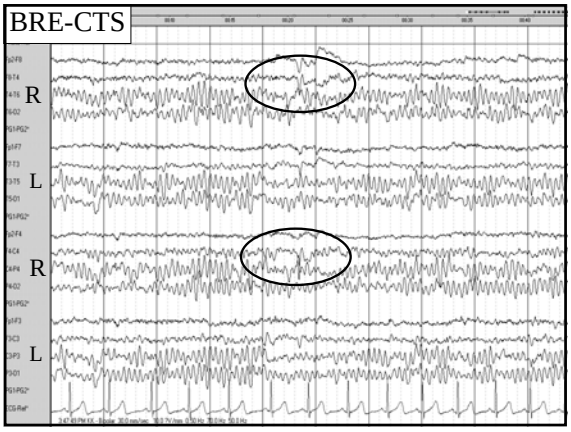
Interictal EEG in BRE

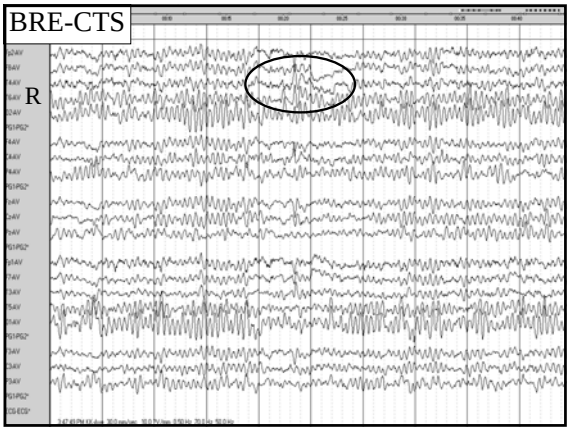
- Spike/wave discharges
  - triphasic follow by after coming slow wave
  - the complex lasts for 80-120 seconds
  - unilateral discharges 70 %
  - bilateral discharges in 30 % of patients, independent& asynchronous

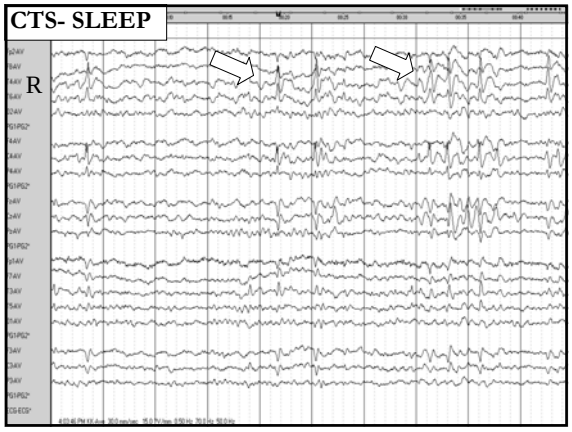


CTS are not specific to Rolandic sz

- 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







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)

	BRE	PS	ICOE-G
Prev amongst children age 1-15 yrs	15%	6%	0.5-1%
Range of age (yrs)	1-14	1-14	3-15
Peak age at onset (yrs)	7-10	3-6	8-11

Brain,131:2264-86, 2008

	BRE	PS	ICOE-G
Typical onset	Hemifacial sensory-motor or oro-pharyngo-laryngeal symptoms	Autonomic symptoms mainly emesis	Visual symptoms mainly with elementary visual hallucination

	BRE	PS	ICOE-G
Duration for 1-3 min	Yes	No	Yes
Duration > 5 mins	Rare	Common	Rare
Partial status	no	40%	no

	BRE	PS	ICOE-G
Single sz only	10-12%	30%	exceptional
Frequent sz	10%	10%	90%
Nocturnal (sleep only)	70%	64%	exceptional
Sz after age of 13	rare	exceptional	common

EEG	BRE	PS	ICOE-G
CTS alone	Yes	Rare	Not reported
Occipital spikes	Not reported	65%	90%
Spikes in other location	Uncommon	Frequent	Exceptional
Photo-sensitivity	Not reported	Exceptional	20-30%
Ictal onset	Rolandic region	Ant^{r} and post^{r} regions	Occipital region