Idiopathic epilepsy syndromes

Kamornwan Katanuwong MD
Chiangmai University Hospital
1st Epilepsy Camp, Hua Hin
20th August 2010

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.

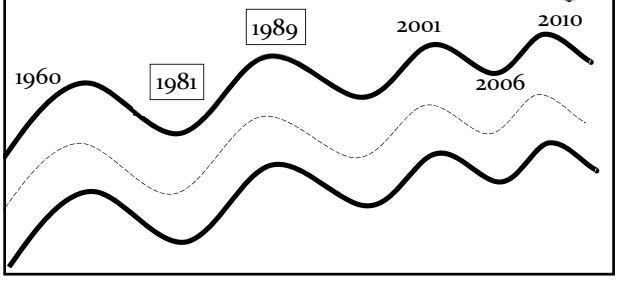
Idiopathic epilepsy syndromes

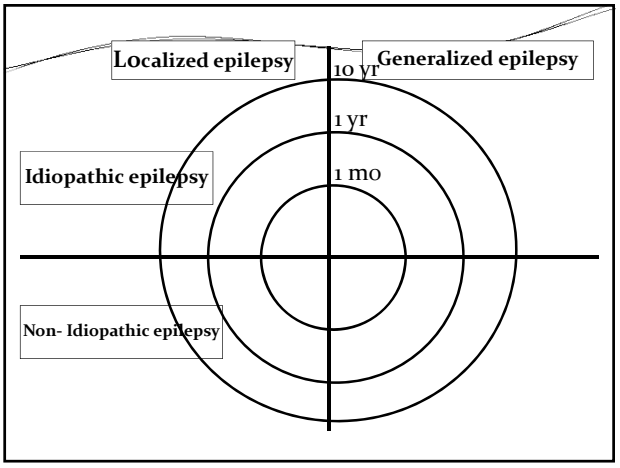
Outline
- Idiopathic partial epilepsy syndromes
- Idiopathic generalized epilepsy syndromes
- Idiopathic / symptomatic / cryptogenic
- Focal seizures / generalized seizures
- Modified concepts to replace the above...

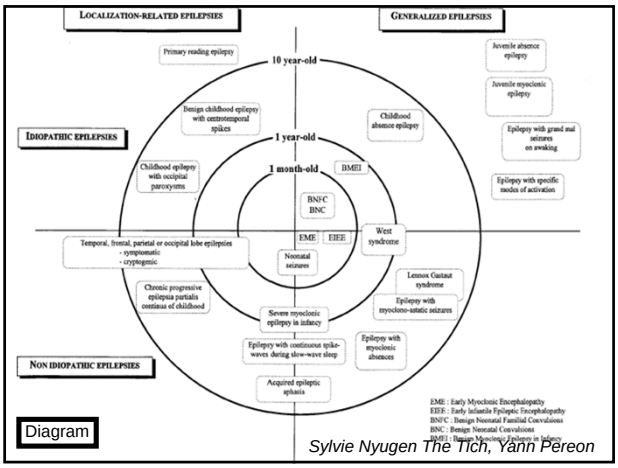
Idiopathic epileptic syndromes

- Presumed to be genetic and usually age dependent
- Idiopathic is not synonymous with benign

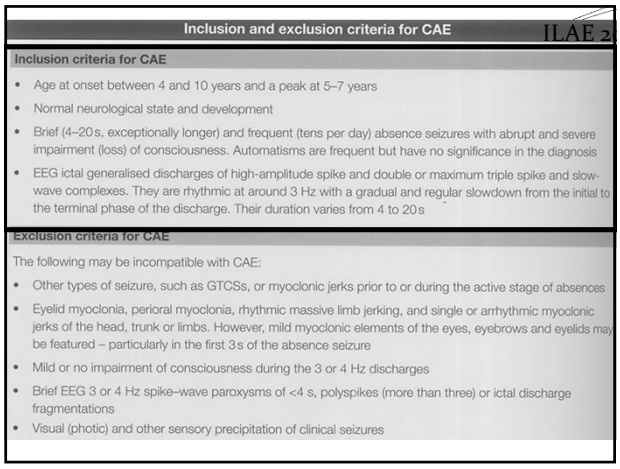
Terminology and classification





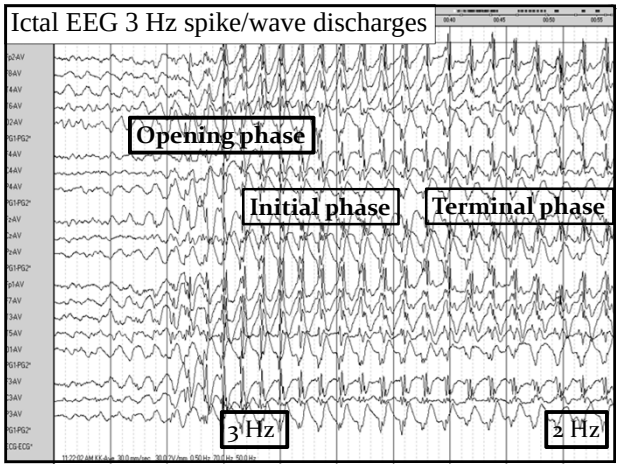


Sylvie Nyugen The Tich, Yann Pereon



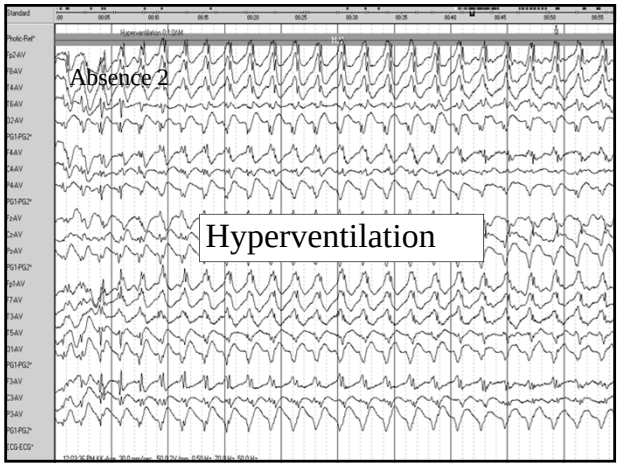
Childhood absence epilepsy (CAE)

- 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

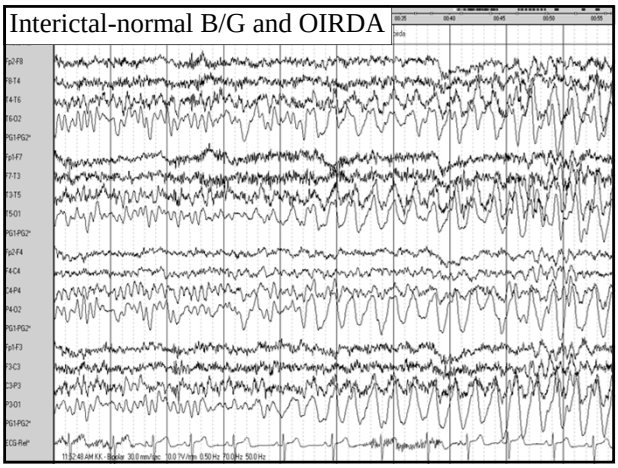


4 Major types of Absences

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

Inclusion and exclusion criteria for CAE

Inclusion criteria for CAE

- Age at onset between 4 and 10 years and a peak at 5–7 years
- Normal neurological state and development
- Brief (4–20 s, exceptionally longer) and frequent (tens per day) absence seizures with abrupt and severe impairment (loss) of consciousness. Automatisms are frequent but have no significance in the diagnosis
- EEG ictal generalised discharges of high-amplitude spike and double or maximum triple spike and slow-wave complexes. They are rhythmic at around 3 Hz with a gradual and regular slowdown from the initial to the terminal phase of the discharge. Their duration varies from 4 to 20 s

Exclusion criteria for CAE

The following may be incompatible with CAE:

- Other types of seizure, such as GTCSs, or myoclonic jerks prior to or during the active stage of absences
- Eyelid myoclonia, perioral myoclonia, rhythmic massive limb jerking, and single or arrhythmic myoclonic jerks of the head, trunk or limbs. However, mild myoclonic elements of the eyes, eyebrows and eyelids may be featured – particularly in the first 3 s of the absence seizure
- Mild or no impairment of consciousness during the 3 or 4 Hz discharges
- Brief EEG 3 or 4 Hz spike–wave paroxysms of <4 s, polyspikes (more than three) or ictal discharge fragmentations
- Visual (photic) and other sensory precipitation of clinical seizures

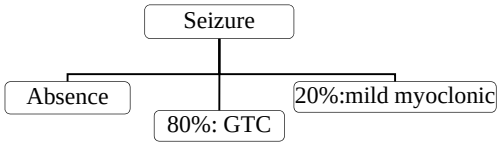
Juvenile absence epilepsy (JAE)

- 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

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

JAE



JAE

- seizure
  - Absence
    - duration 4-30 sec (~16s)
    - freq automatism,6-10 sec after EEG onset
    - mild myoclonic of the eyelids
    - absence status : 20%
  - GTC
  - myoclonic

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

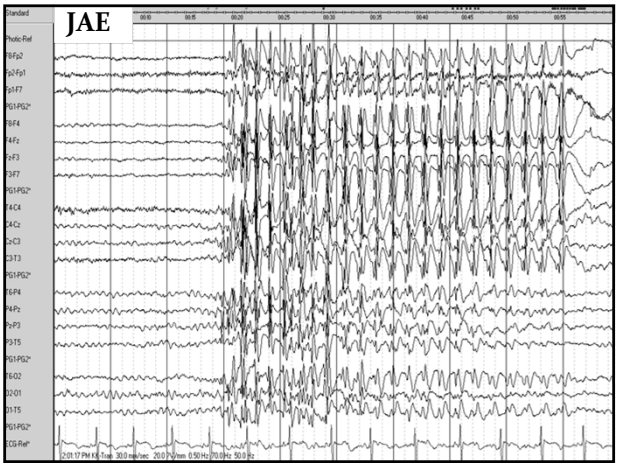
EEG exclusion criteria:

- 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)

JAE

- Seizure
  - Absence
    - follow onset of absence
    - mainly after awakening
  - 80%GTC
  - 20% mild myoclonic

JAE



JAE

- Seizure
  - Absence
    - afternoon when tired
  - GTC
  - 20% mild myoclonic

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

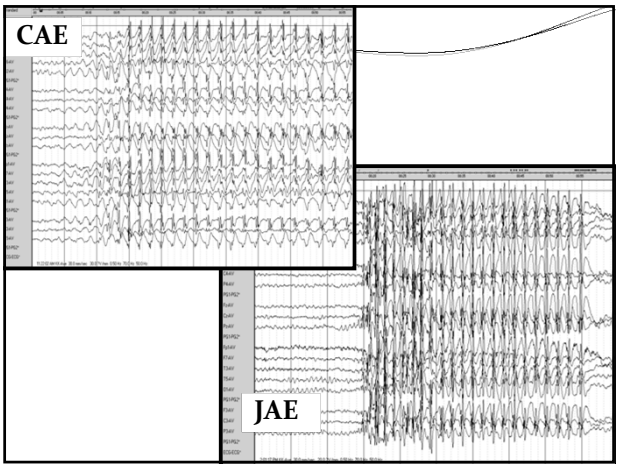
The following may be incompatible with JAE

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)



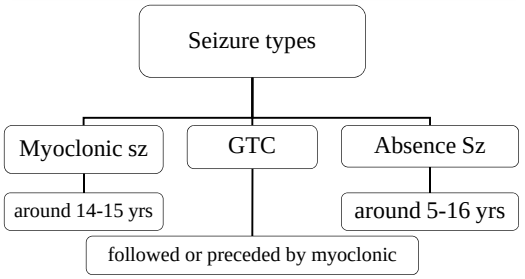
Juvenile myoclonic epilepsy (JME)

- 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

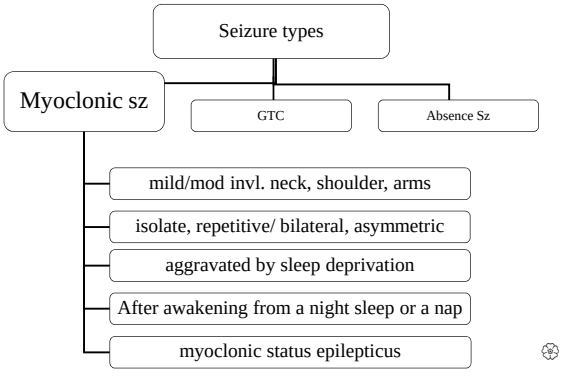
JME



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, fatique and alcohol consumption
- Myoclonic jerks are not problematic

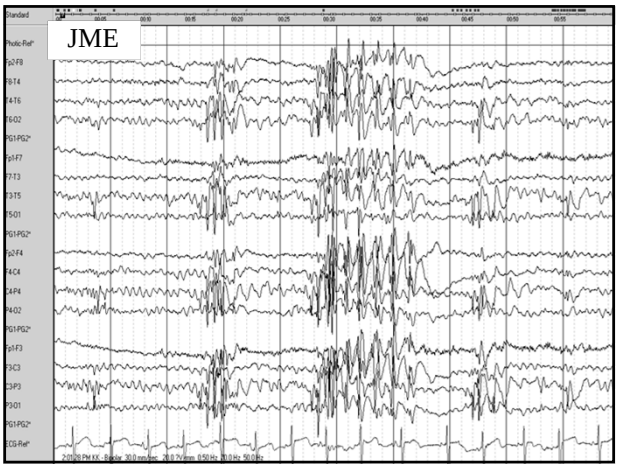
JME



JME

Seizure types

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

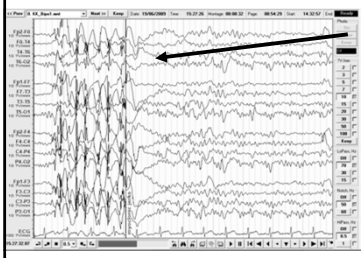


JME

Seizure types

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

JME EEG



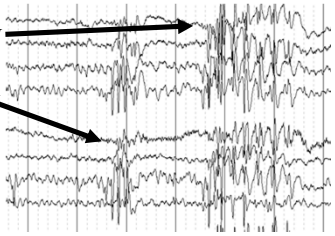
Ictal EEG

- myoclonic sz
  - fast 10-16 Hz spikes followed by irregular slow waves (PSW)
- absence sz
  - multiple spikes preceding on slow wave, last 1-4 sec

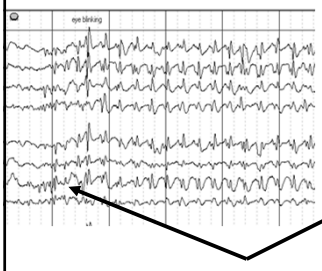
JME EEG

Interictal EEG

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



JME EEG



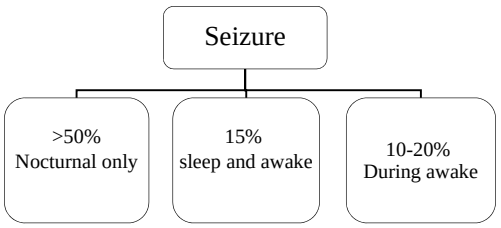
Ictal EEG

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

Key differences between JME and JAE

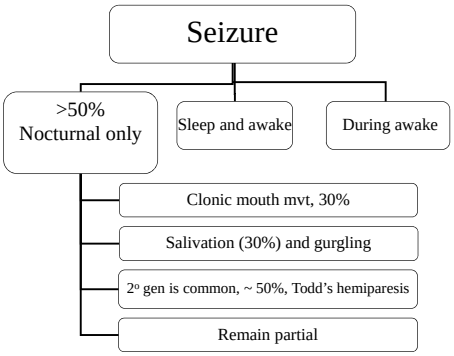
	JME	JAE
Main type of seizures	Myoclonic jerks	Typical absences
Circadian distribution	Mainly on awakening	Any time during the day
Typical absences	Mild and often imperceptible; they occur in a third of patients	Defining seizure type; they are very severe and occur in all patients
Myoclonic jerks	Defining 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 (1–3 s) 3–6 Hz GPSWD, which are usually asymptomatic	Lengthy (8–30 s) 3–4 Hz GPSWD, which are usually associated with severe impairment of consciousnes

BRE



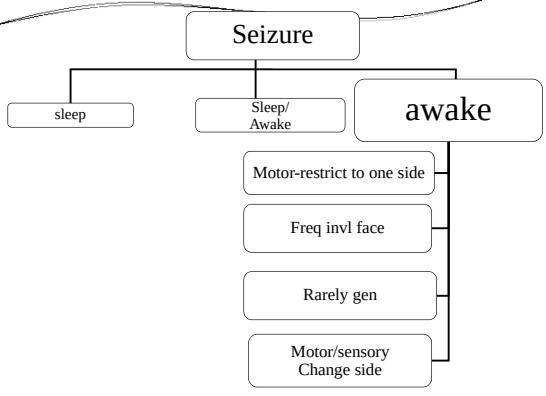
Benign childhood focal epilepsies

- 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)



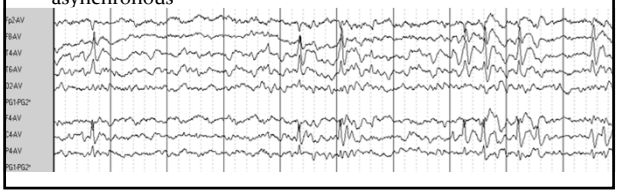
Benign rolandic epilepsy

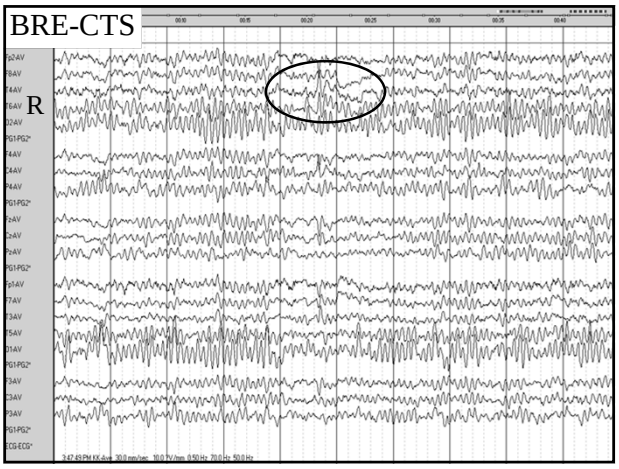
- Age: 3-13 years (peak 7-8yrs of age)
- Sex: Boys > Girls
- Development: normal
- Genetic: familial, linked to Chromosome 15 q
  : 50% of close relatives have EEG abnormalities between the ages of 5-15 yr
  : 12% of persons whom EEG abnormal have clinical seizure.



Interictal EEG in BRE

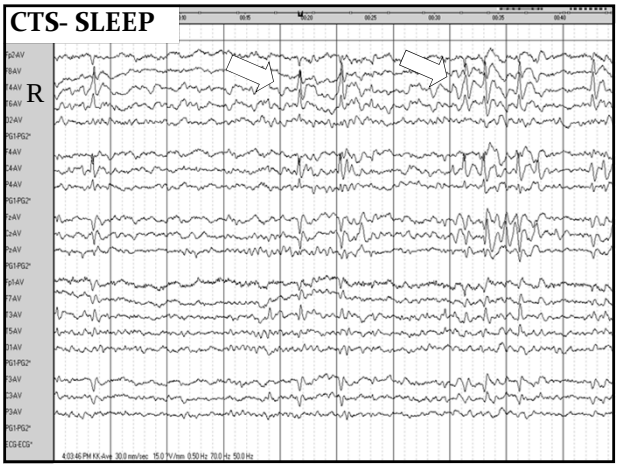
- Spike/wave discharges
  - triphasic follow by after coming slow wave
  - the complex lasts for 80-120 m-seconds
  - unilateral discharges 70 %
  - bilateral discharges in 30 % of patients, independent& asynchronous

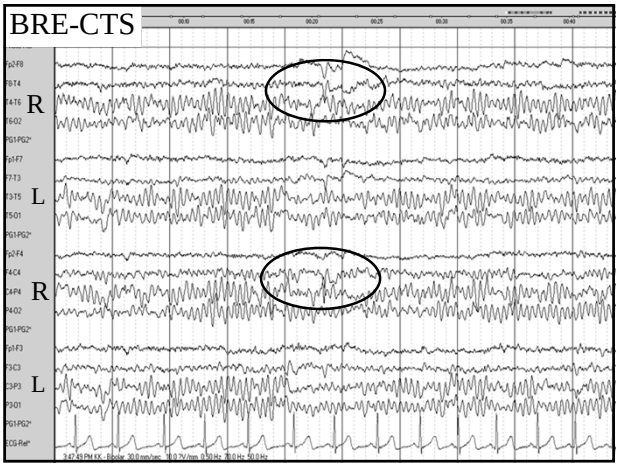


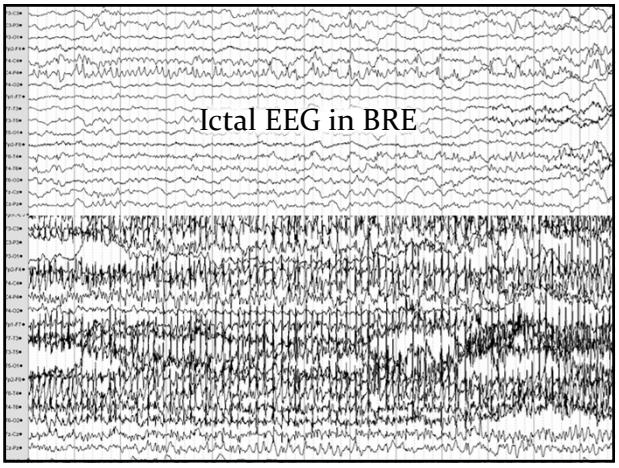


CTS are not specific to Rolandic sz

- 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







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

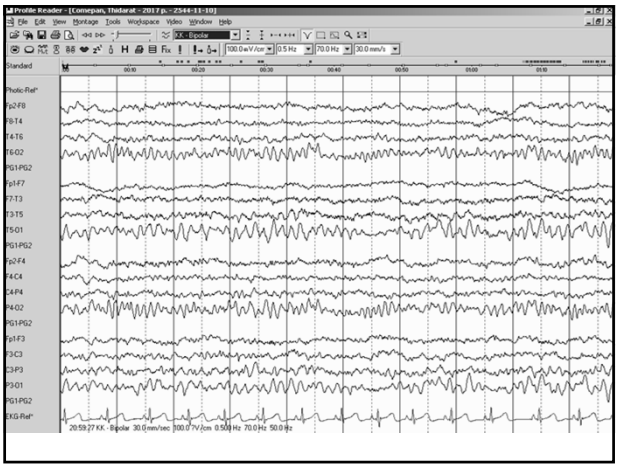
	BRE	PS	ICOE-G
Prev amongst children age 1-15 yrs	15%	6%	0.5-1%
Range of age(yrs)	1-14	1-14	3-15
Peak age at onset (yrs)	7-10	3-6	8-11

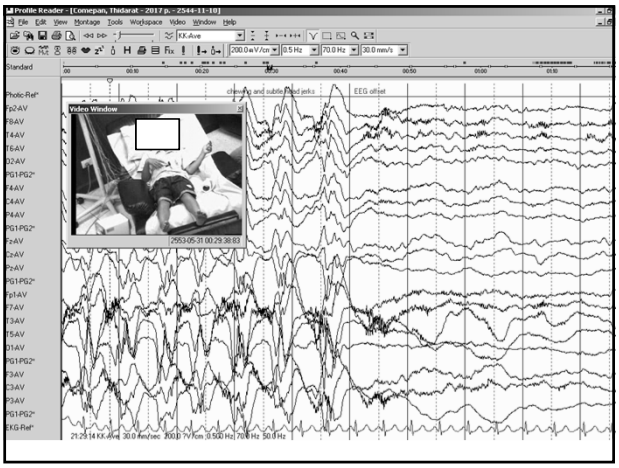
Brain,131:2264-86, 2008

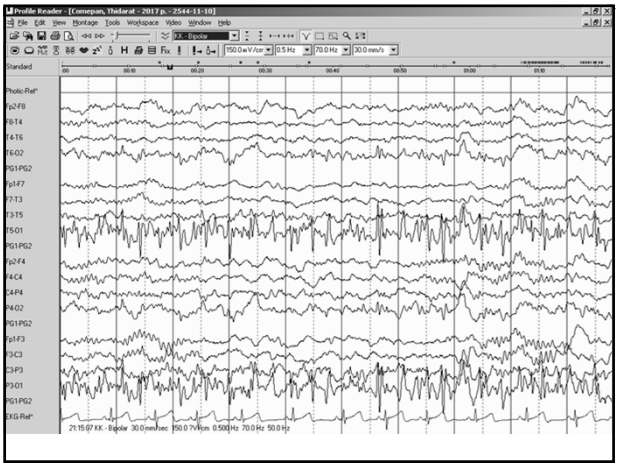
	BRE	PS	ICOE-G
Single sz only	10-20%	30%	exceptional
Frequent sz	10%	10%	90%
Nocturnal (sleep only)	70%	64%	exceptional
Sz after age of 13	rare	exceptional	common

	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

EEG	BRE	PS	ICOE-G
CTS alone	Yes	Rare	No
Occipital spikes	No	65%	90%
Spikes in other location	Uncommon	Frequent	Exceptional
Photo-sensitivity	No	Exceptional	20-30%
Ictal onset	Rolandic regions	Ant^{r} /Post^{r} regions	Occipital regions







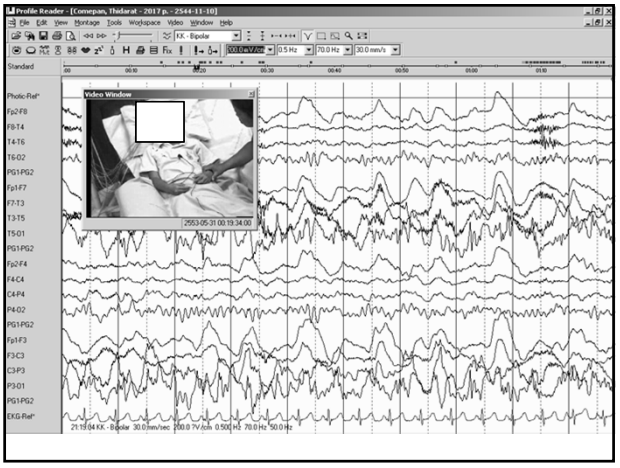
Idiopathic epilepsy syndromes

Outline

- Idiopathic partial epilepsy syndromes
- Idiopathic generalized epilepsy syndromes

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

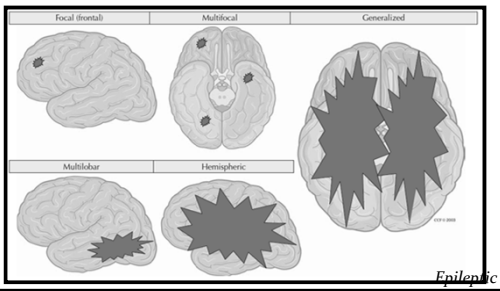
- Modified concepts to replace the above…



New concept

- Focal vs Generalized seizure

Old



Epileptic disorder 2005

New concept

- Focal vs Generalized seizure

New

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

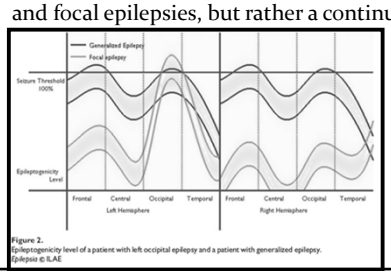


Figure 2.
Epileptogenicity level of a patient with left occipital epilepsy and a patient with generalized epilepsy.
Epilepsia © ILAE

Epilepsia 2009

New concept

- Idiopathic/ symptomatic/cryptogenic

New

- Genetic
- Structural / metabolic diseases
- Unknown cause

New concept

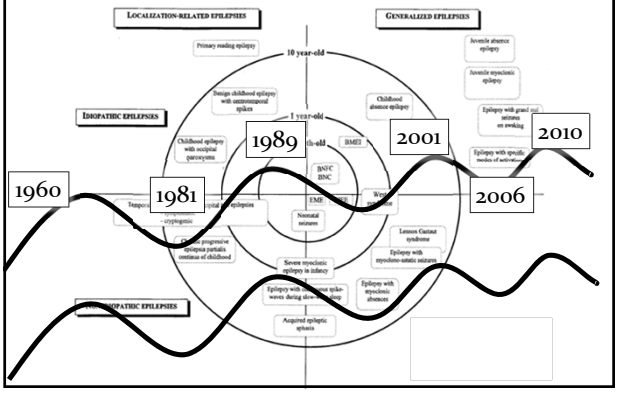
- Focal vs Generalized seizure

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

Terminology and classification



New concept

- Idiopathic/ symptomatic/cryptogenic

Old