Idiopathic epilepsy syndromes

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Epilepsy course

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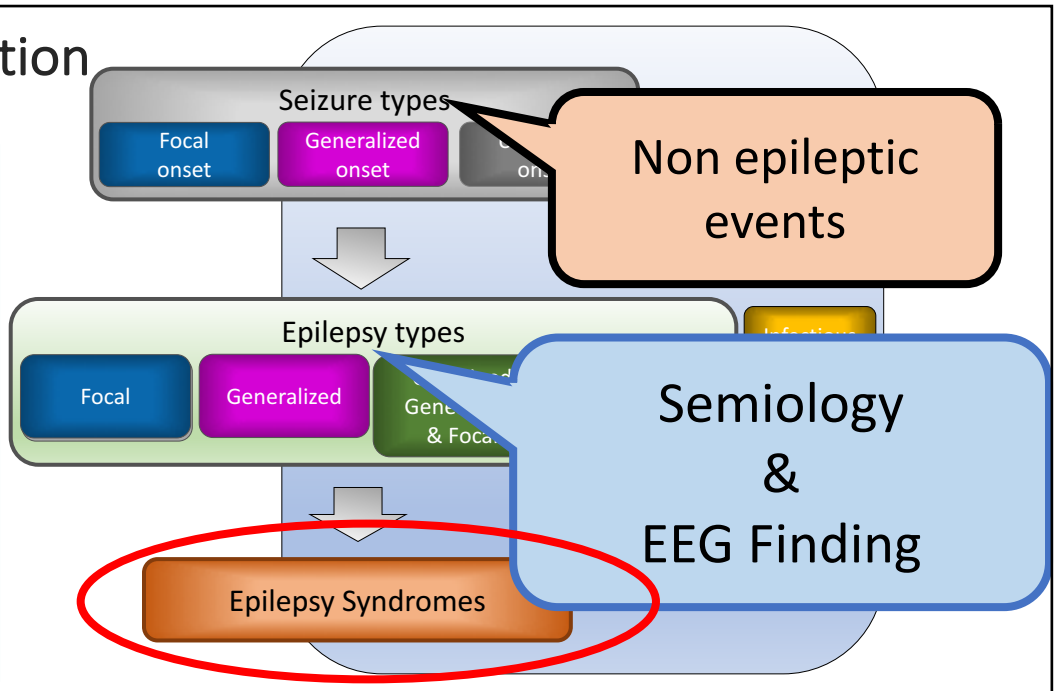
Classification

- 1964
- 1970
- 1981
- 1989
- 2001
- 2006
- 2010
- 2013
- 2017



Identify of epilepsy syndromes based on:

- Typical age onset
- Seizure types
- specific EEG characteristics
- Other features or related symptoms
- **Implications for** treatment, management, and prognosis

no approved ILAE epilepsy syndromes

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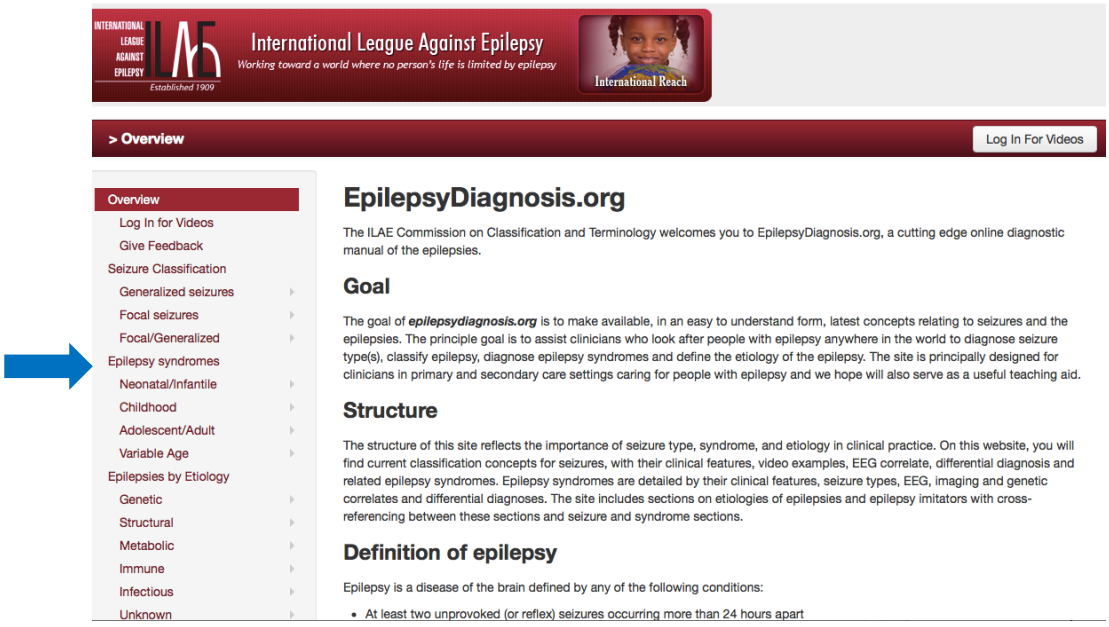
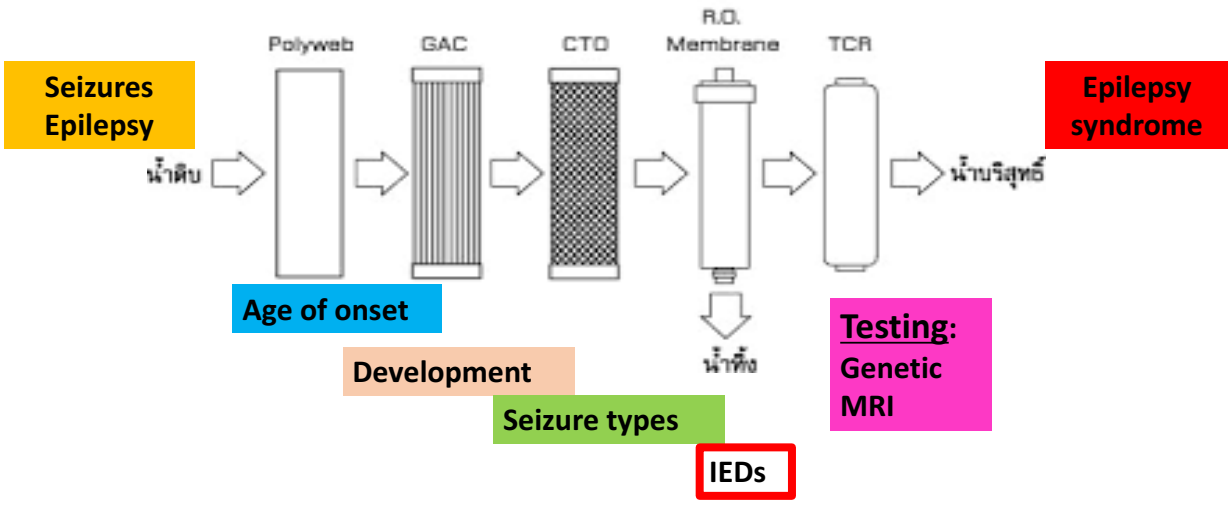
https://www.epilepsydiagnosis.org



Easy guide for Epilepsy syndrome "Pattern diagnosis"



Epilepsy syndrome

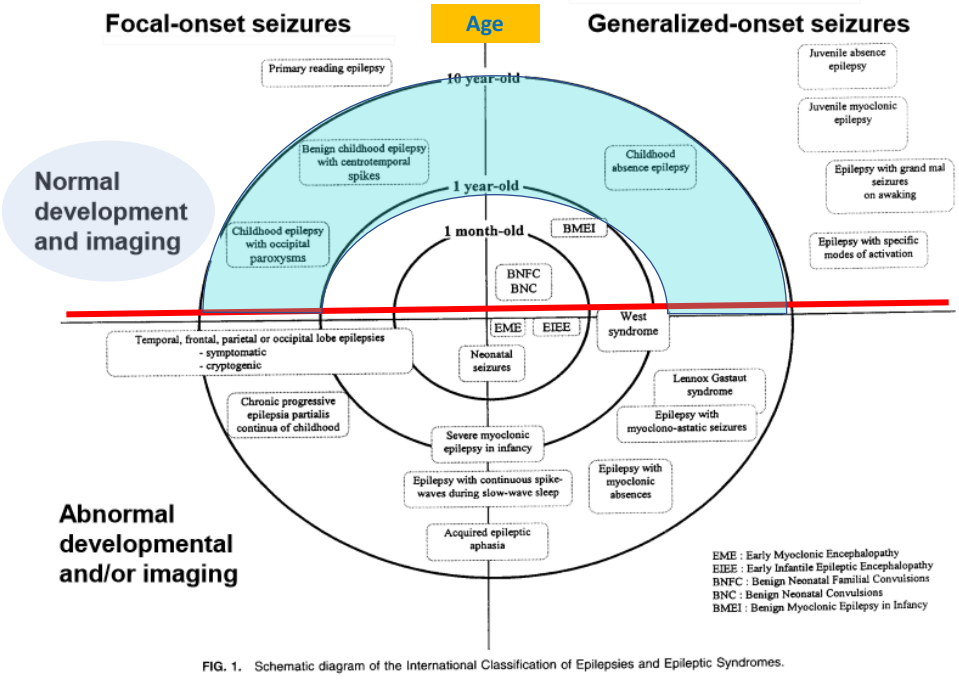


FIG. 1. Schematic diagram of the International Classification of Epilepsies and Epileptic Syndromes.

Nguyen The Tich S, Pereon Y. Epilepsia 1999;40:531

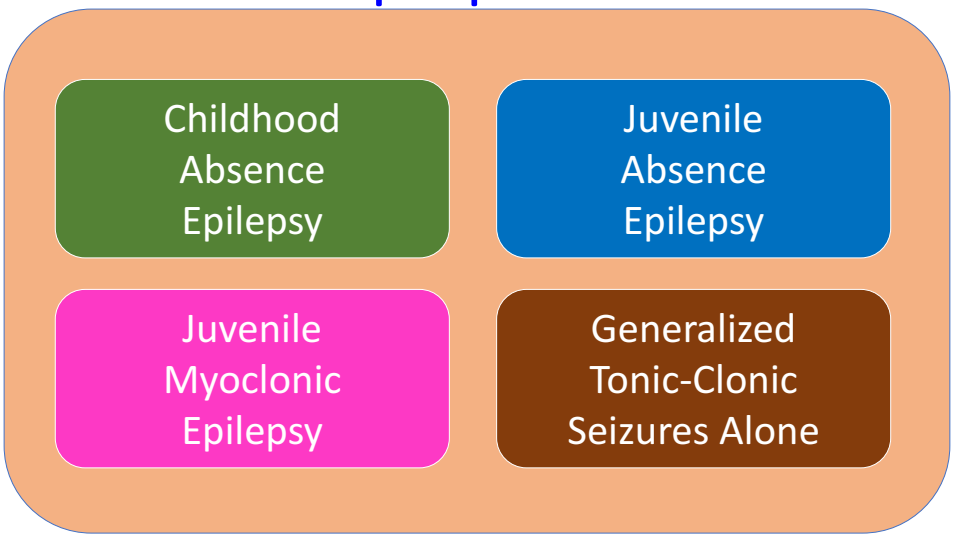
Idiopathic epilepsy syndromes

- A syndromic that is only epilepsy, with no underlying structural brain lesion or other neurological signs or symptoms. These are presumed to be genetic and are usually age-dependent.

Idiopathic Epilepsy Syndromes

- Idiopathic/Genetic Generalized Epilepsy
- Idiopathic/Self-Limited Focal Epilepsy

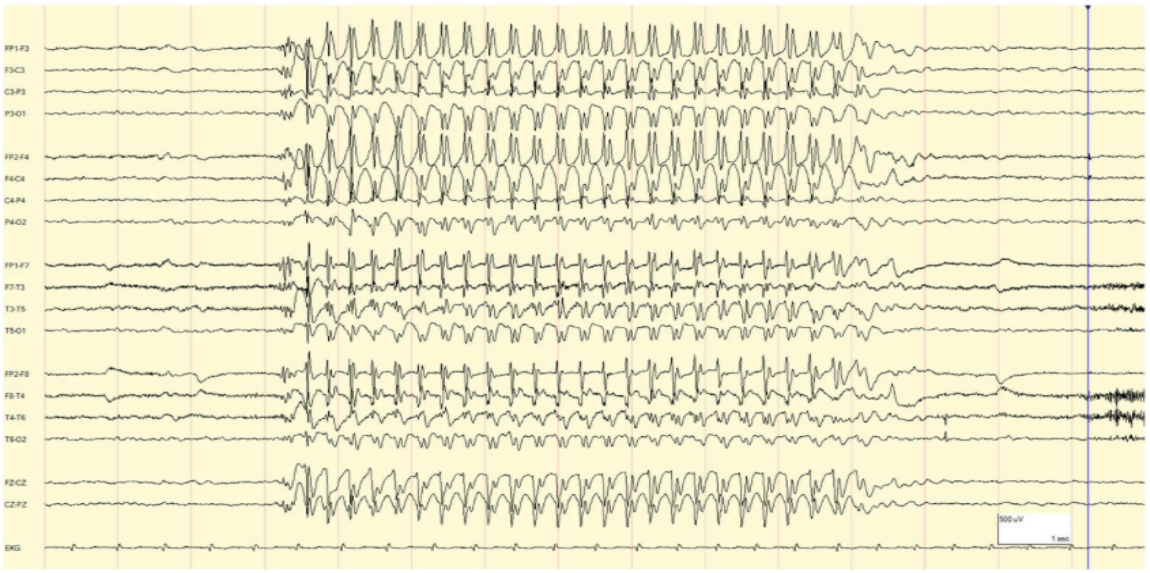
'Idiopathic/Genetic Generalized Epilepsies'

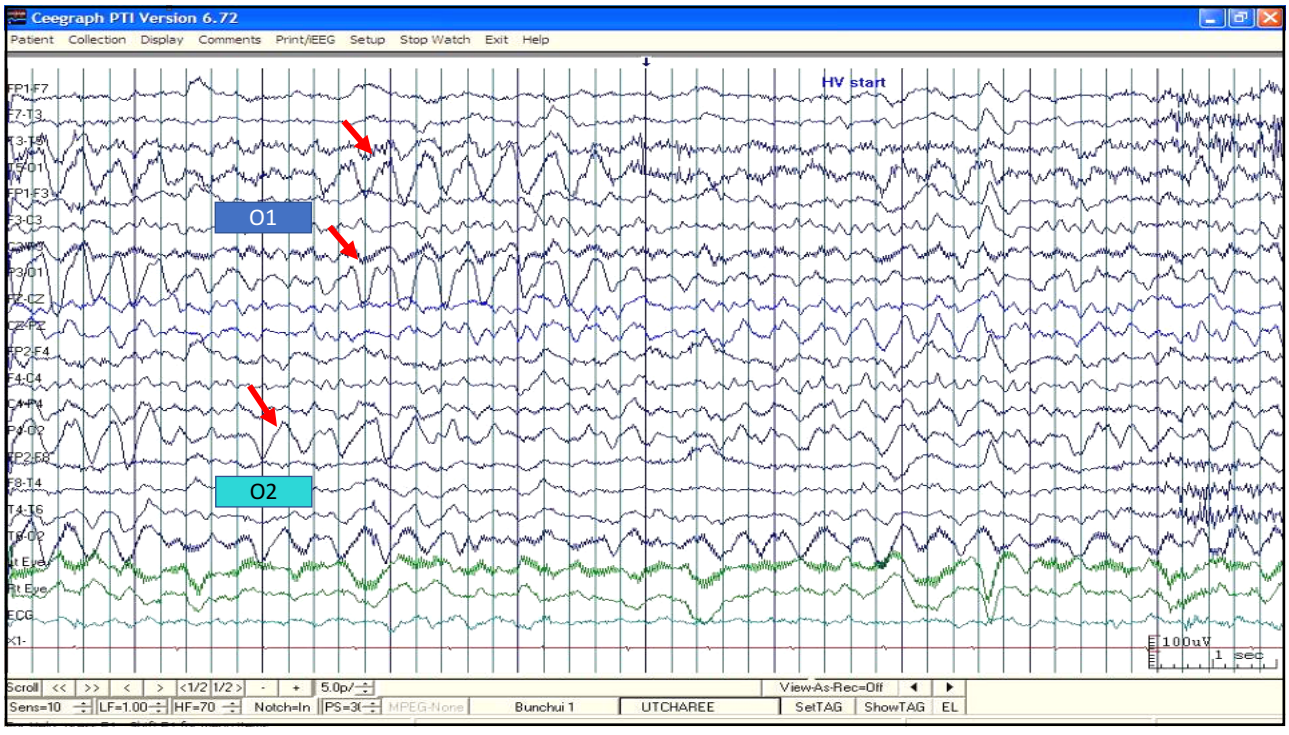


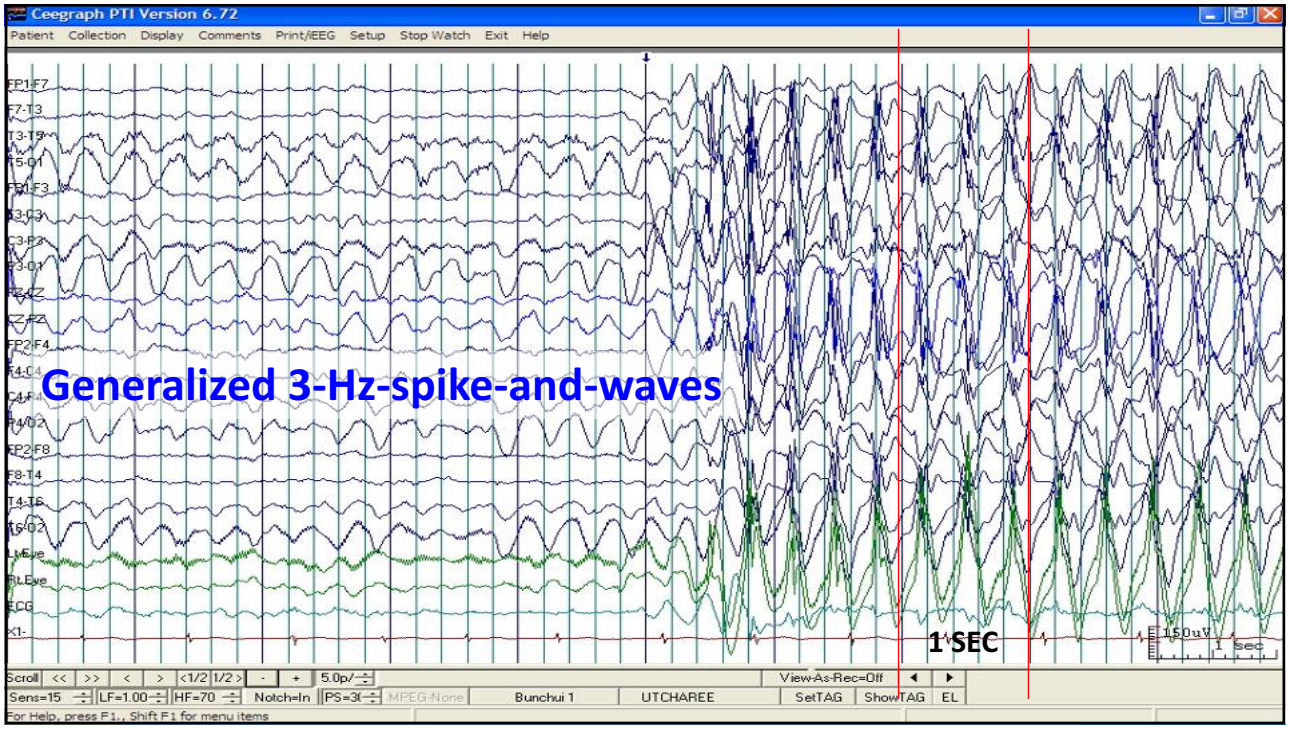
Childhood absence epilepsy

Age of onset	2-12 years (peak 5-6 years)
Seizure type	Absence only (multiple daily, brief, LOA)
EEG	IEDS: 3 Hz Generalized spikes and waves; Normal background, OIRDA
Tests: Genetic	SLC2A1, GABRG2 and CACNA1A

Childhood absence epilepsy



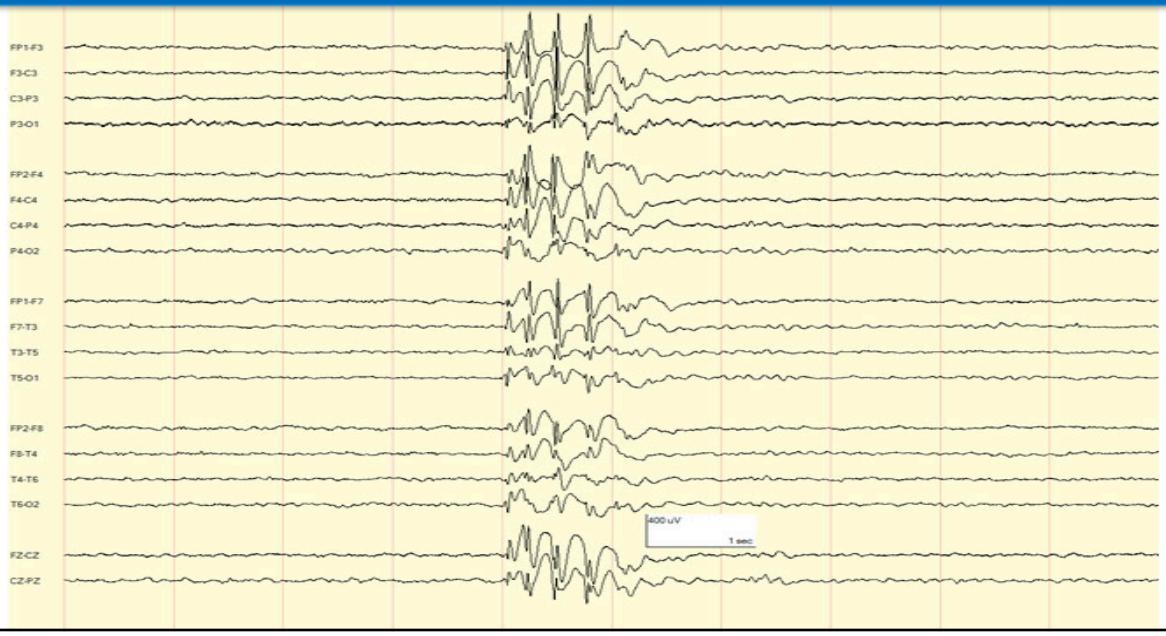




Juvenile absence epilepsy

Age of onset	8-20 years (peak 9-13 years)
Seizure type	Absence (not frequent, not severe, awareness) GTC at onset-> Absence in adolescent GCs (80% of cases, upon awakening)
EEG	IEDS: **3-6 Hz** Generalized spikes/polyspikes and waves, normal background, OIRDA(may) Ictal: Absence: Regular 3-6 Hz GSW or PSW GCs : EEG obscure by artifact, generalized fast rhythmic spikes-tonic phase, spike and slow waves and postictal period slowing
Tests: Genetic	GABRG2, CACNA1A and others

Juvenile absence epilepsy



Juvenile myoclonic epilepsy

Age of onset	8-25 years (peak 9-13 years) **5% of cases from CAE**
Seizure type	Myoclonic (mandatory), especially on awakening (within 30min-1hr) GTCs (>90%) preceded by series of myoclonic , Absence (1/3 of cases, briefer<3 seconds)
EEG	IEDS: **3.5-6 Hz** GSW/P SW,normal background, fragments hyperventilation may provoked absence, <10% sz induced by visual stimuli Ictal: single generalized PSW correlates with myoclonic seizures
Tests: Genetic	Complex or MendelianCACNB4, GABRA1, CLCN2, GABRD and EFHC1, Microdeletions, such as the 15q13.3 microdeletion and others

Epilepsy with GTCs alone

Age of onset	5-40 years (peak 11-23 years)
Seizure type	GCs especially on awakening (within 1-2 hr of wakening) infrequent, typically provoked by sleep deprivation, PH of childhood absence epilepsy
EEG	IEDS: GSW/PSW (½ of cases seen only during sleep) fragmented, intermittent photoparoxysmal response, normal BG (no slowing) Ictal: GCs: Ictal EEG patterns
Tests: Genetic	complex inheritance, CLCN2 and others.

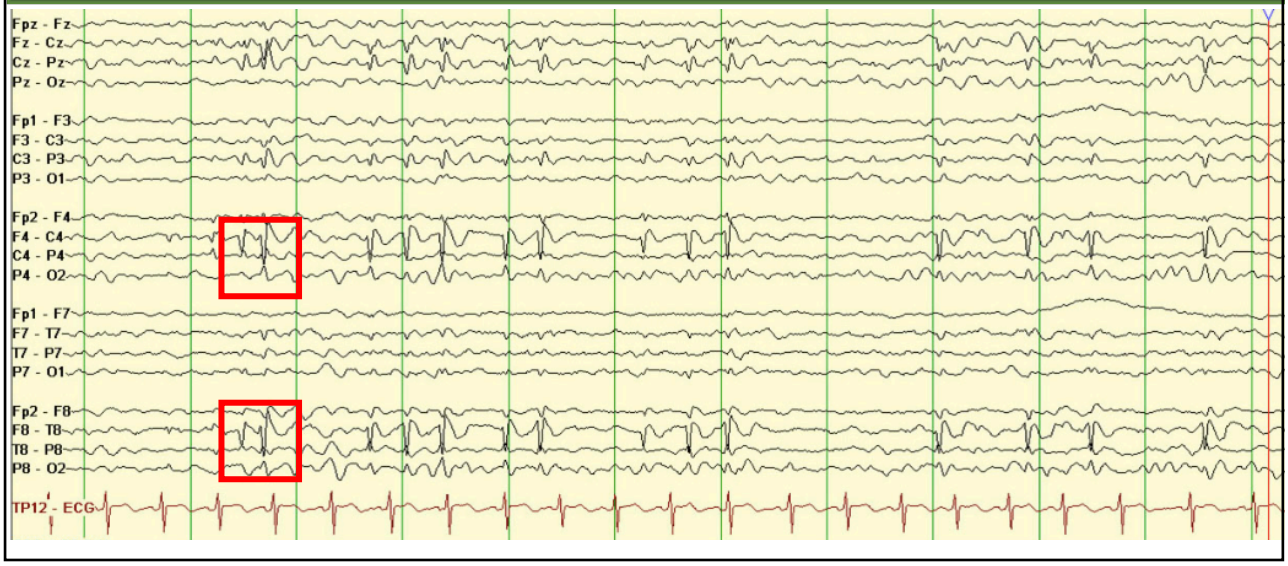
'Self-Limited Focal Epilepsies'

- Benign Epilepsy with CentroTemporal Spikes (BECTS)
- Self-Limited Occipital Epilepsy of Childhood: Panayiotopoulos type (early-onset):PS
- COE-G Gastaut type (late onset)
- Self-limited frontal /temporal/parietal lobe epilepsies

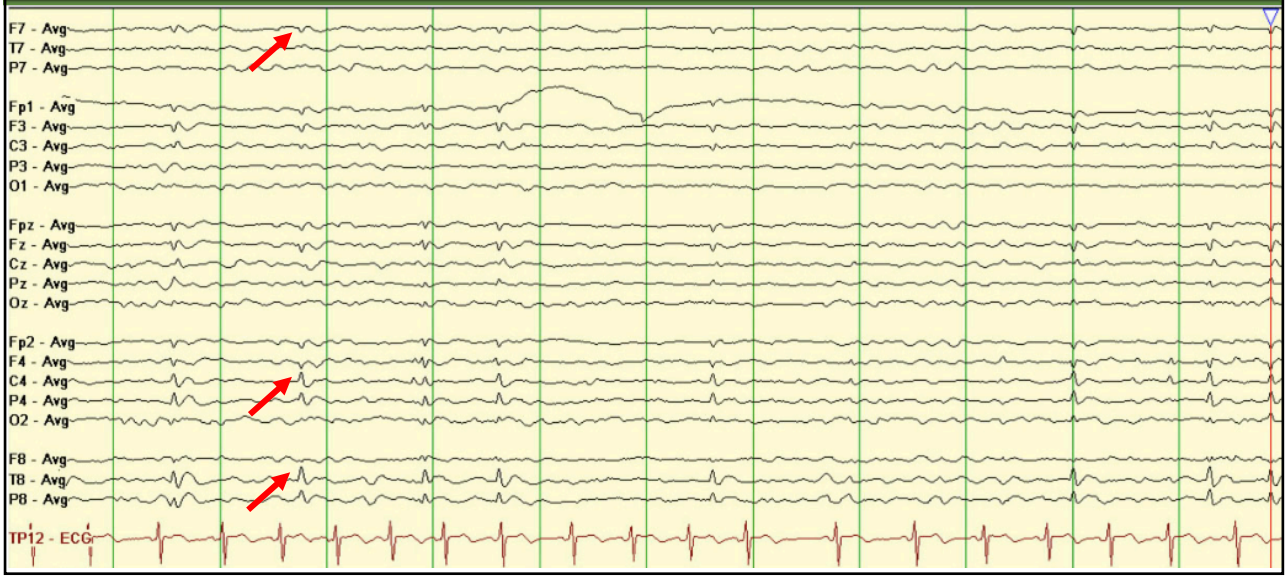
Benign Childhood Epilepsy with Centrotemporal Spikes	
Age of onset	3-14 years (peak 8-9 years)
Seizure type	**Fronto-parietal opercular features** –hemifacial (lip,mouth and tongue),clonic movements (with may be unilateral), laryngeal symptoms, articular difficulty (aphasia), swallowing or chewing movements and hypersalivation, brief (<5 minutes), Few, (may) secondarily generalize (typically nocturnal events) (not GTC during awake) **Self-limited** usually resolved by age 13 years (occasionally occur up to age 18 years)

Benign Childhood Epilepsy with Centrotemporal Spikes	
EEG	IEDs: High amp. Centrotemporal Spikes or Sharp-and-slow wave complexes, max. negativity in CT (C3/C4 and T3/T4) and max. positivity F, increased during drowsiness and sleep, unilat or bilat, (may) SPK outside CT region (midline, parietal, frontal and occipital), (may) photoresponsive (age.10 yrs), 10-20%-by sensory stimuli of fingers or toes Ictal: rare to obtained ictal recording
Tests: Genetic	Complex inheritance, GRIN2A gene

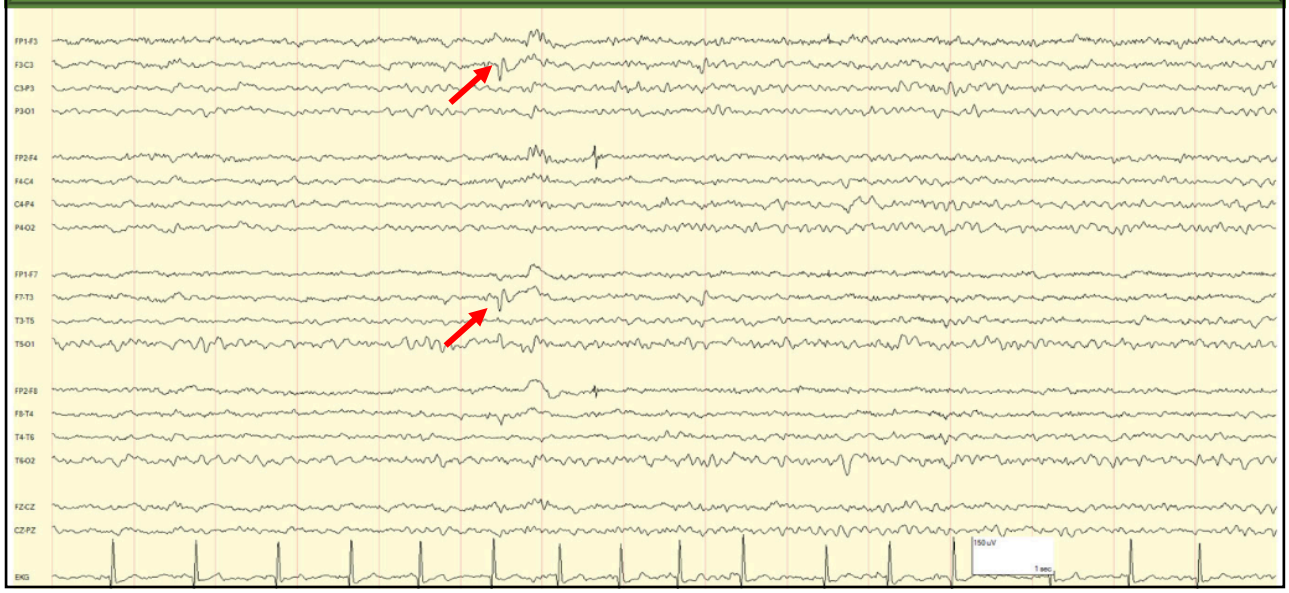
Benign Childhood Epilepsy with Centrotemporal Spikes



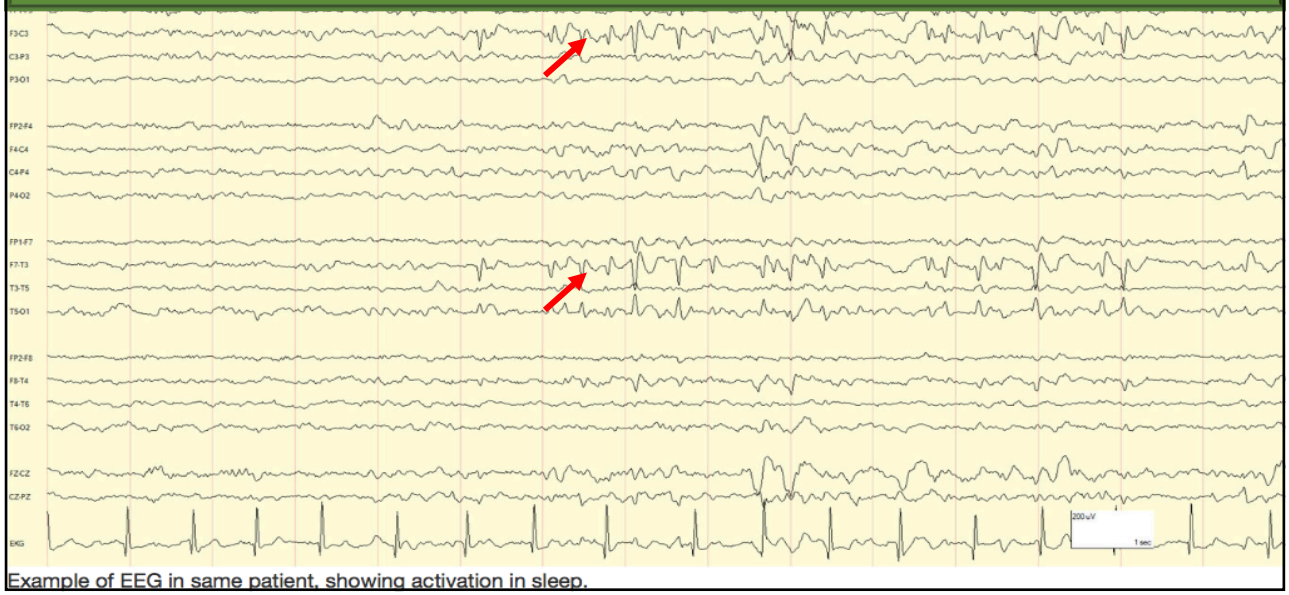
Benign Childhood Epilepsy with Centrotemporal Spikes



Benign Childhood Epilepsy with Centrotemporal Spikes



Benign Childhood Epilepsy with Centrotemporal Spikes



Example of EEG in same patient, showing activation in sleep.

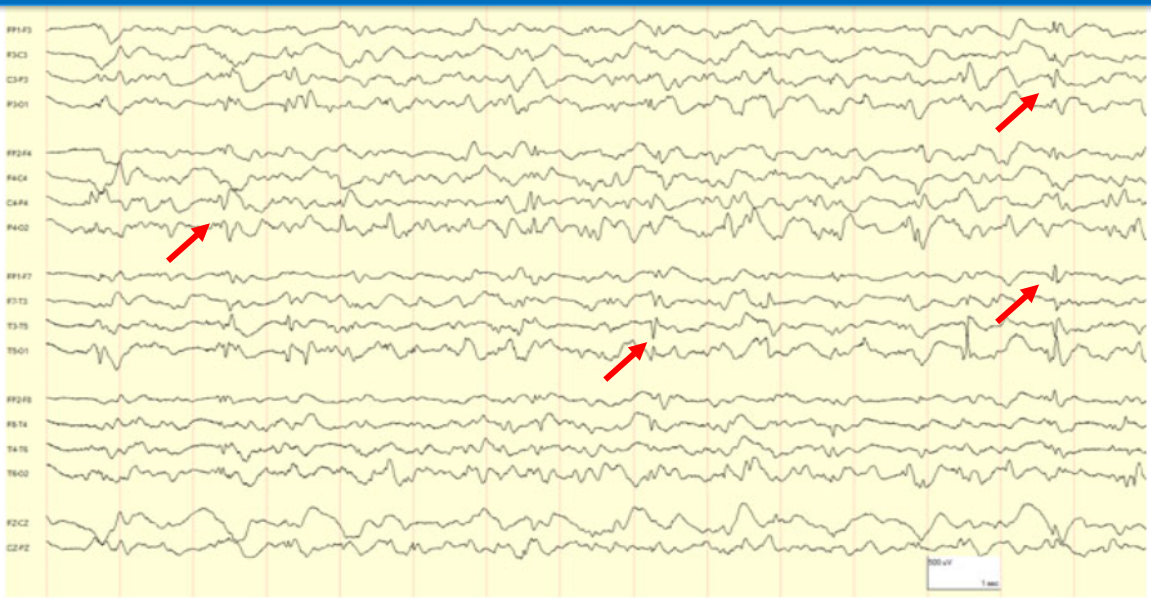
Panayiotopoulos syndrome

Age of onset	1-14 years (peak 3-6 years) **Self-limiting, resolve by age 11-13 years**
Seizure type	**Autonomic features** mainly emetic (nausea, retching, vomiting), pupillary (mydriasis), circulatory (pallor, cyanosis), heart and respiratory change. Apnea and asystole can occur (severe case). Prolong duration, but without residual neuro deficit, some of case- fronto-parietal opercular (25%may autonomic SE), infrequent
Tests: Genetic	unknown gene, complex (report in sibling)

Panayiotopoulos syndrome

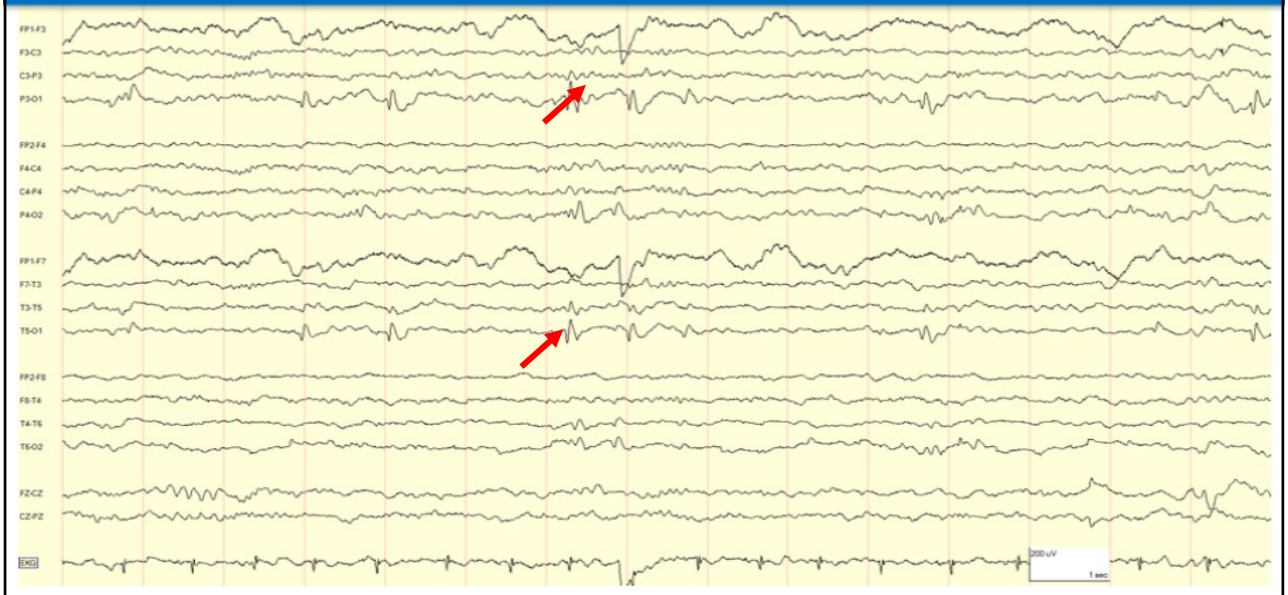
EEG	**IEDs: Multifocal SPK/SW 90%** **Normal single EEG 10%** **Occipital spikes 60% of patients** **Low voltage SPK and Gen d/c minority of cases.** **Activation: Eye closure (elimination of central vision and fixation off sensitivity) may activate occipital spikes. EEG abnormality is enhanced by sleep deprivation and by sleep** **Ictal: Unilateral, often posterior onset, with rhythmic slow (theta or delta) activity intermixed with small spikes**

Panayiotopoulos syndrome



Example of multifocal spikes in Panayiotopoulos syndrome.

Panayiotopoulos syndrome



Example of occipital (left) spikes of high amplitude in Panayiotopoulos syndrome.

Late onset childhood occipital epilepsy(Gastaut type)	
Age of onset	5 months-19 years (peak 8-9 years)
Seizure type	Seizures with **visual aura** occur from awake states, brief (typical seconds, most < 3 minutes, rarely up to 20minutes Visual aura; multi-colored circles in peripheral vision increased involved and moving horizontally to the other side, these may be followed by deviation of eyes or head turning (ipsilateral) May Other occipital features; ictal blindness, complex visual hallucinations, visual illusions, orbital pain, eyelid fluttering or repetitive eye closure, ictal headache or N/V May spread outside the occipital lobe resulting in hemiparesthesia, dyscognitive features, hemiclonic

Late onset childhood occipital epilepsy(Gastaut type)	
EEG	**IEDs: Occipital spikes or SW (may) only during sleep, 20% of cases may co-exist with CT, frontal or GSW, BG normal** **Activation by sleep deprivation and by sleep, 20-90% of cases –induced by fixation-off sensitivity (elimination of central vision)** **Ictal: during oculo-clonic seizure or ictal blindness : BG activity reduction and then occipital faster rhythms with spikes of low amplitude, these may be slower SW**
Tests: Genetic	**Unknown**
Prognosis	**Self-limiting** Easily controlled (50-60% remission in 2-4 years after onset) 90% dramatic response to carbamazepine

Summary of IGE

IGE	CAE	JAE	JME	GTCSA
Age onset	childhood	Juvenile	Juvenile	Juvenile
Seizure type	Absence	Absence GTCs	Myoclonic GTCs, Absence	GTCs
EEG	3 Hz GSW	3-6 Hz GSW	3.5-6 Hz GSW	GSW/PSW

Summary of SFE

SFE	PS	BECTS	COE-G
Age onset	Infantile 1-14 (3-6) yrs	Childhood	Childhood
Seizure type	Autonomic (Emetic)	Perisylvian	Occipital
EEG	Multifocal 90% Occipital 60%	Centrotemporal	Occipital