

Idiopathic epilepsy syndromes

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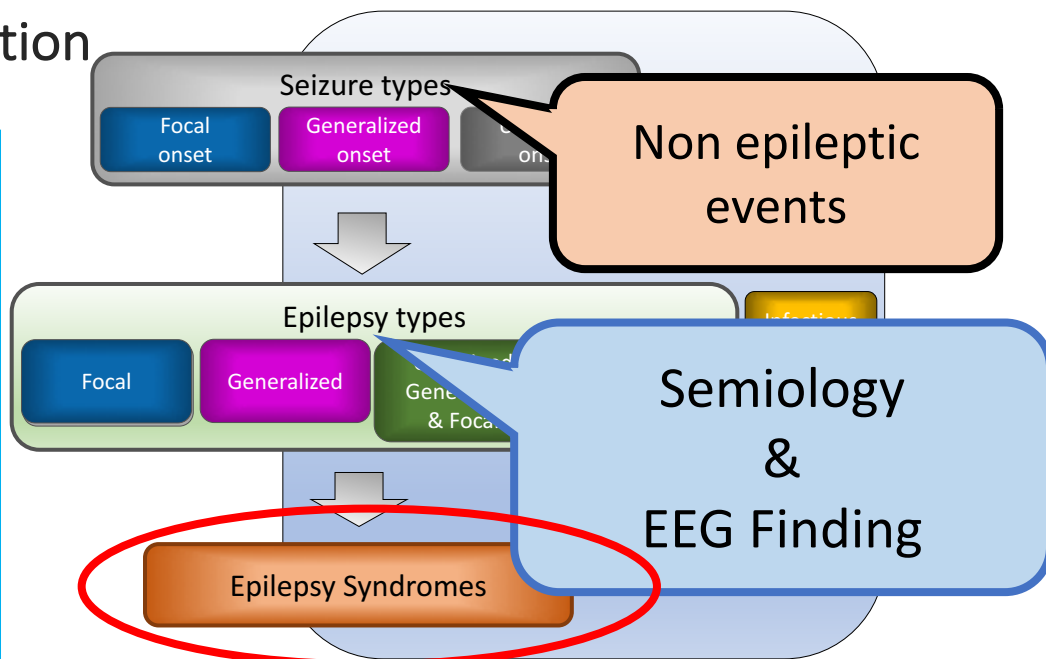
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Epilepsy course

26 August 2017

Classification

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



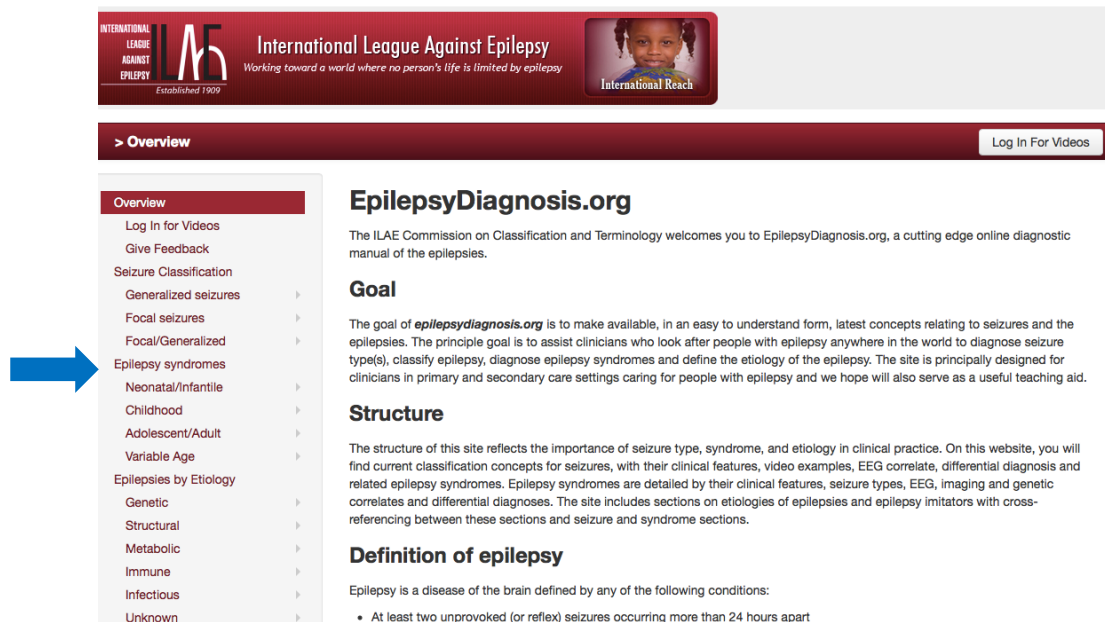
Identify of epilepsy syndromes based on:

- 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>



The screenshot shows the homepage of EpilepsyDiagnosis.org. The header includes the ILAE logo and the text 'International League Against Epilepsy' and 'Working toward a world where no person's life is limited by epilepsy'. Below the header is a navigation bar with a 'Log In For Videos' button. The main content area is divided into a left sidebar and a right main section. The sidebar has a 'Overview' section with a list of links: 'Log In for Videos', 'Give Feedback', 'Seizure Classification', 'Generalized seizures', 'Focal seizures', 'Focal/Generalized', 'Epilepsy syndromes', 'Neonatal/Infantile', 'Childhood', 'Adolescent/Adult', 'Variable Age', 'Epilepsies by Etiology', 'Genetic', 'Structural', 'Metabolic', 'Immune', 'Infectious', and 'Unknown'. A blue arrow points to the 'Epilepsy syndromes' link. The main section is titled 'EpilepsyDiagnosis.org' and contains a welcome message, a 'Goal' section, a 'Structure' section, and a 'Definition of epilepsy' section.

Overview

Log In for Videos

Give Feedback

Seizure Classification

Generalized seizures

Focal seizures

Focal/Generalized

Epilepsy syndromes

Neonatal/Infantile

Childhood

Adolescent/Adult

Variable Age

Epilepsies by Etiology

Genetic

Structural

Metabolic

Immune

Infectious

Unknown

EpilepsyDiagnosis.org

The ILAE Commission on Classification and Terminology welcomes you to EpilepsyDiagnosis.org, a cutting edge online diagnostic manual of the epilepsies.

Goal

The goal of **epilepsydiagnosis.org** is to make available, in an easy to understand form, latest concepts relating to seizures and the epilepsies. The principle goal is to assist clinicians who look after people with epilepsy anywhere in the world to diagnose seizure type(s), classify epilepsy, diagnose epilepsy syndromes and define the etiology of the epilepsy. The site is principally designed for clinicians in primary and secondary care settings caring for people with epilepsy and we hope will also serve as a useful teaching aid.

Structure

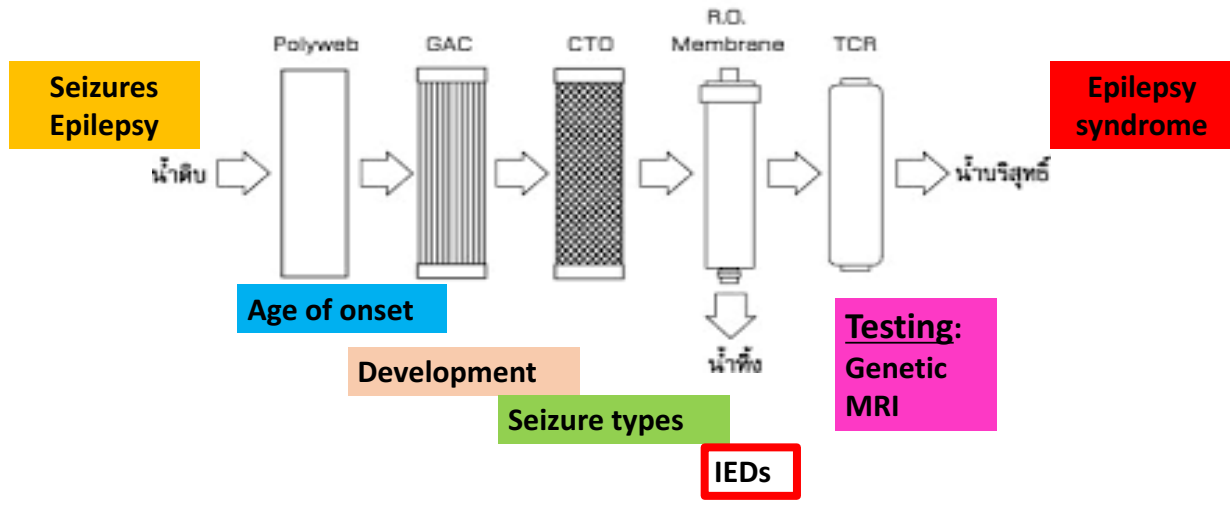
The structure of this site reflects the importance of seizure type, syndrome, and etiology in clinical practice. On this website, you will find current classification concepts for seizures, with their clinical features, video examples, EEG correlate, differential diagnosis and related epilepsy syndromes. Epilepsy syndromes are detailed by their clinical features, seizure types, EEG, imaging and genetic correlates and differential diagnoses. The site includes sections on etiologies of epilepsies and epilepsy imitators with cross-referencing between these sections and seizure and syndrome sections.

Definition of epilepsy

Epilepsy is a disease of the brain defined by any of the following conditions:

- At least two unprovoked (or reflex) seizures occurring more than 24 hours apart

Easy guide for Epilepsy syndrome “Pattern diagnosis”



Epilepsy syndrome

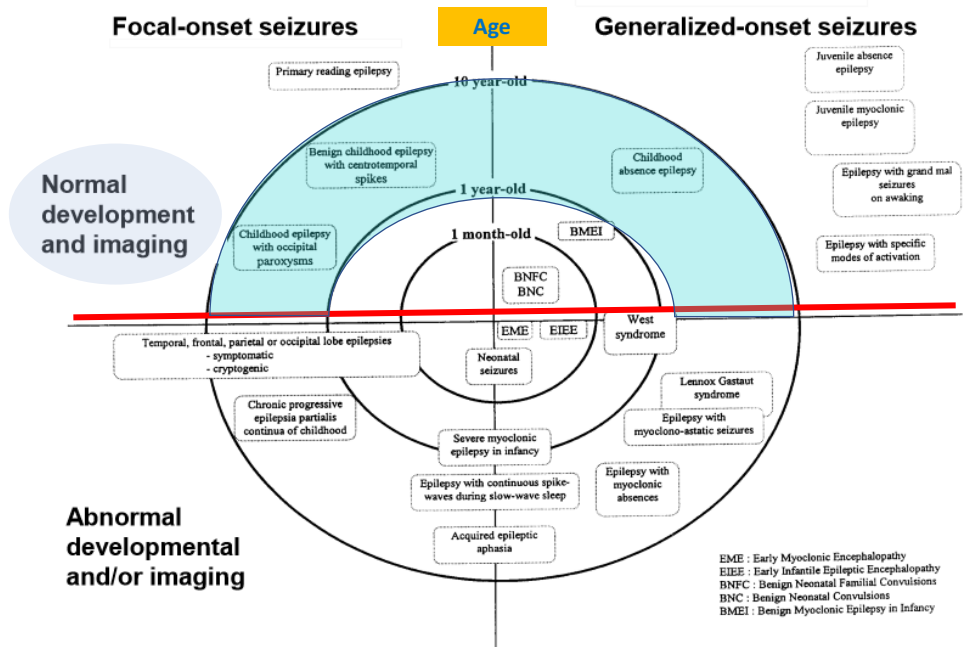


FIG. 1. Schematic diagram of the International Classification of Epilepsies and Epileptic Syndromes.

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

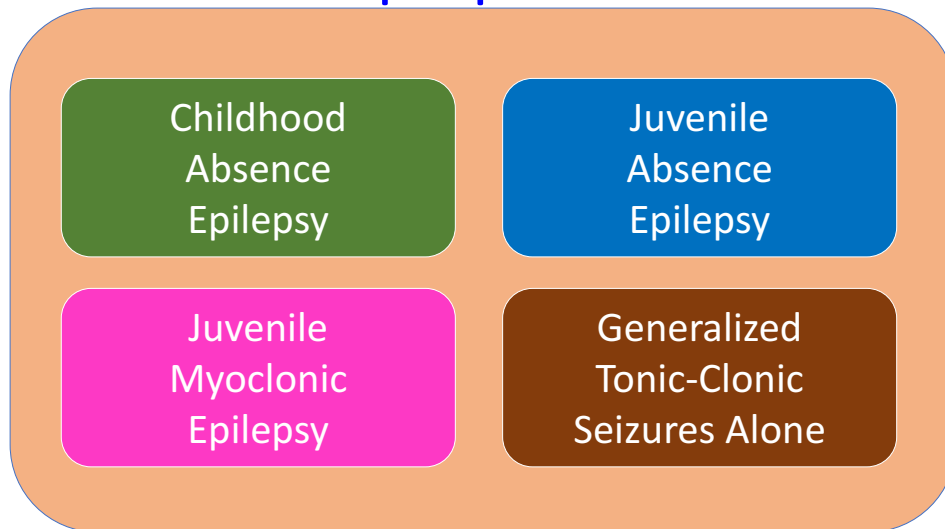
Idiopathic epilepsy syndromes

- 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

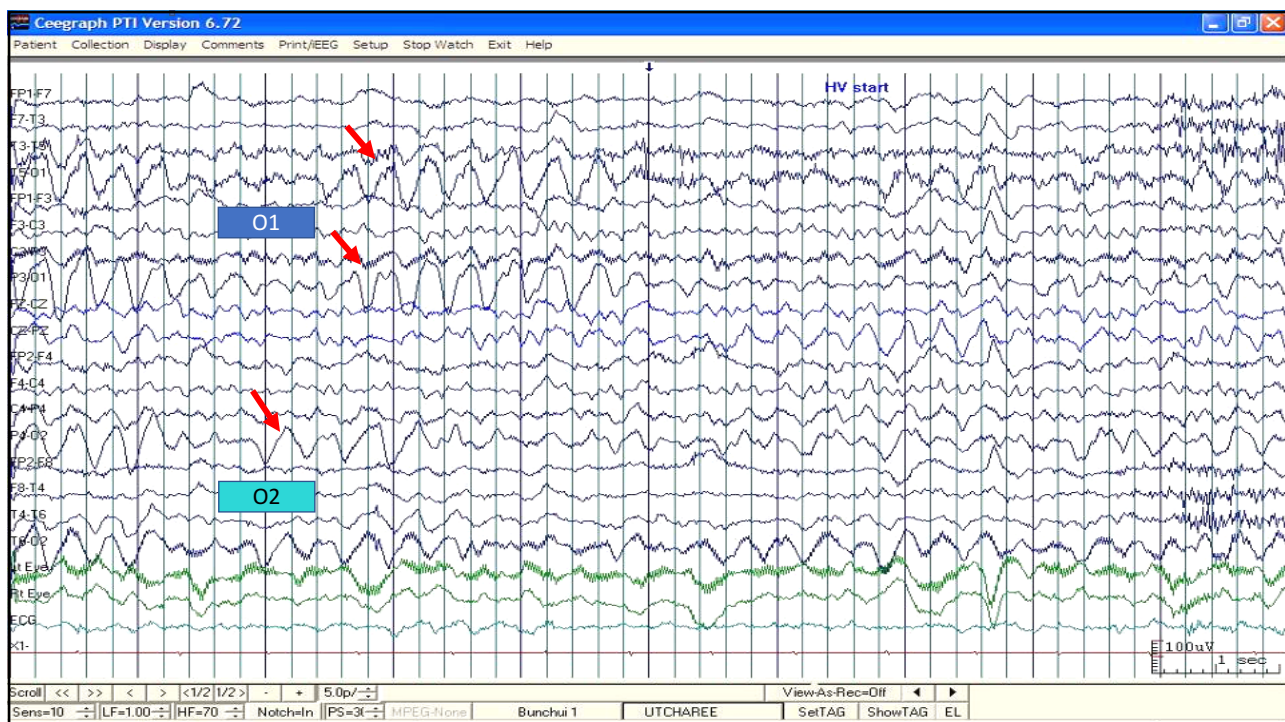
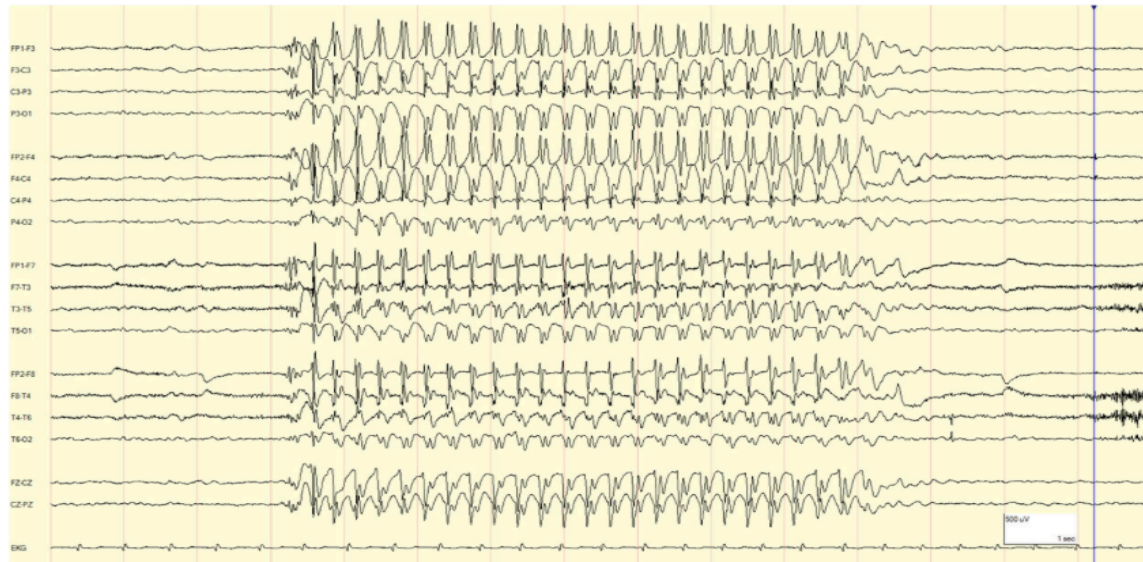
'Idiopathic/Genetic Generalized Epilepsies'



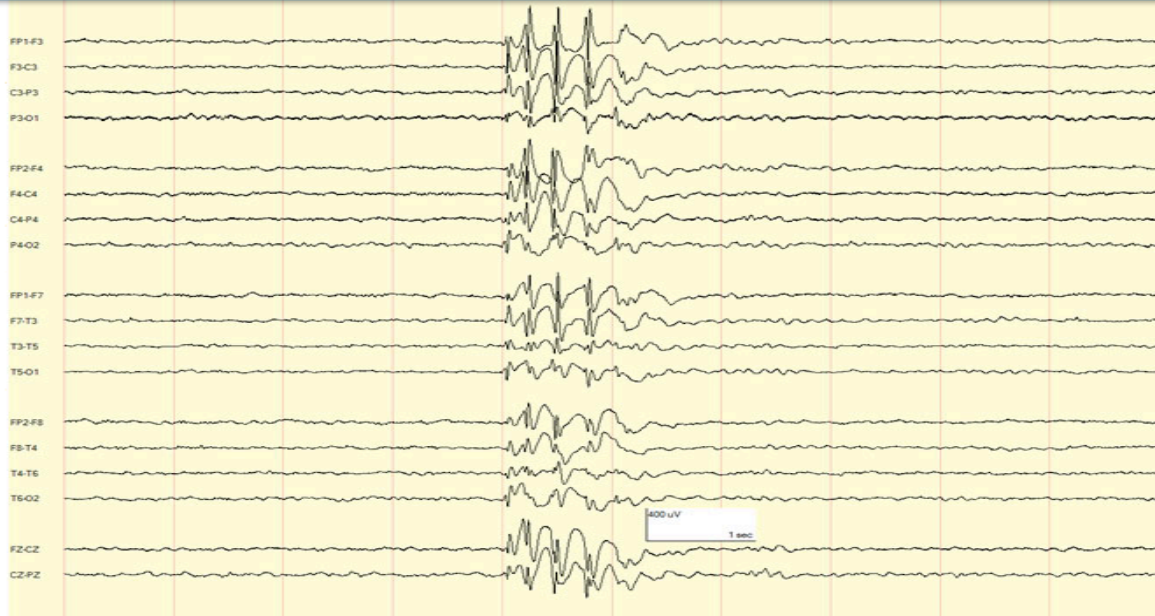
Childhood absence epilepsy

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



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 Mendelian CACNB4 , 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	<u>IEDS</u> : GSW/PSW (½ of cases seen only during sleep) fragmented, intermittent photoparoxysmal response, normal BG (no slowing) <u>Ictal</u> : GCs: Ictal EEG patterns
Tests: Genetic	complex inheritance, CLCN2 and others.

‘Self-Limited Focal Epilepsies’

Benign **E**pilepsy with
Centro**T**emporal
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

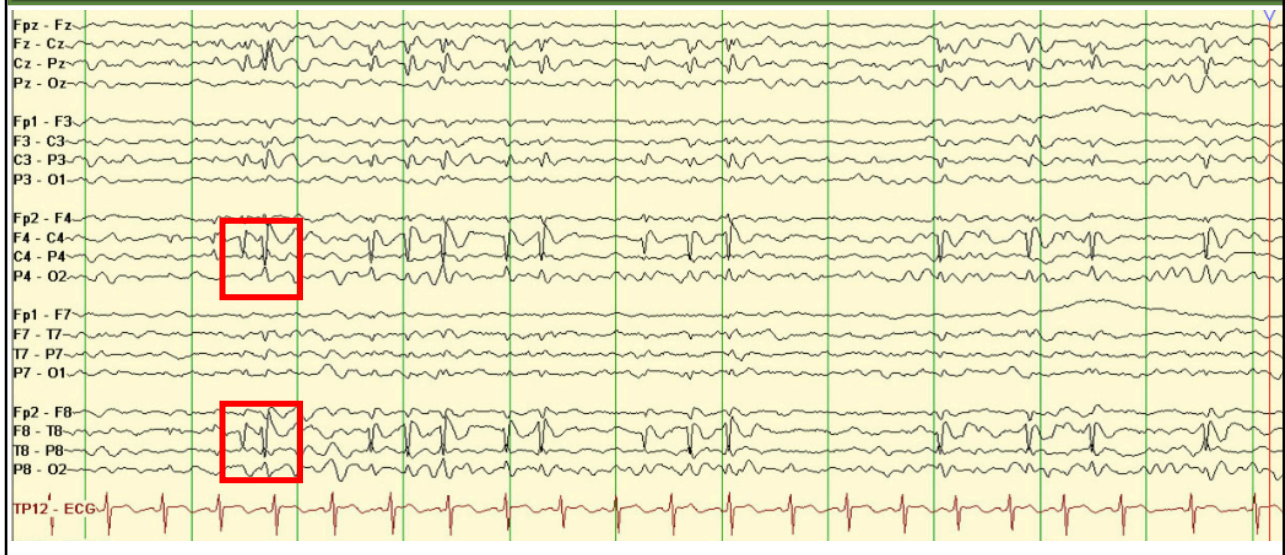
EEG

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

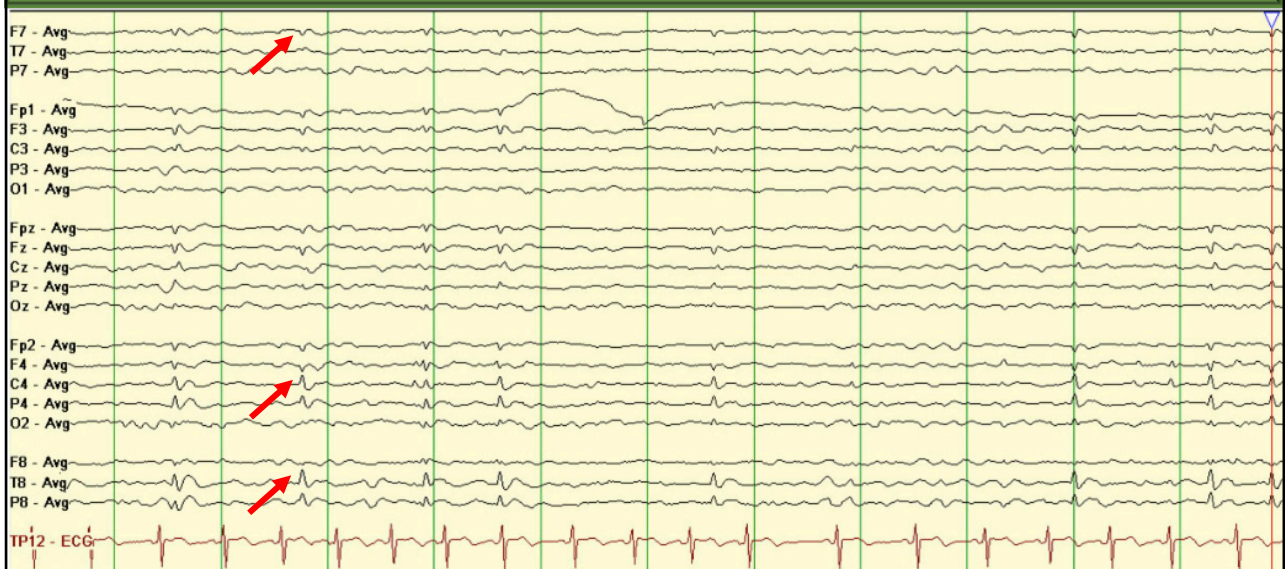
Tests: Genetic

Complex inheritance, GRIN2A gene

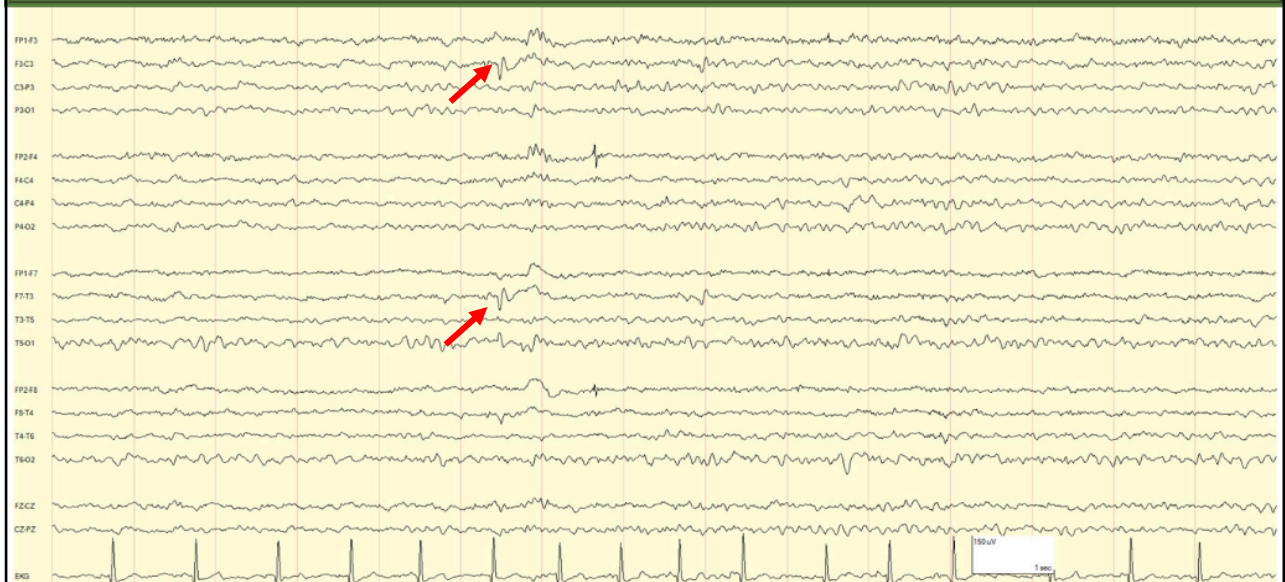
Benign Childhood Epilepsy with Centrotemporal Spikes



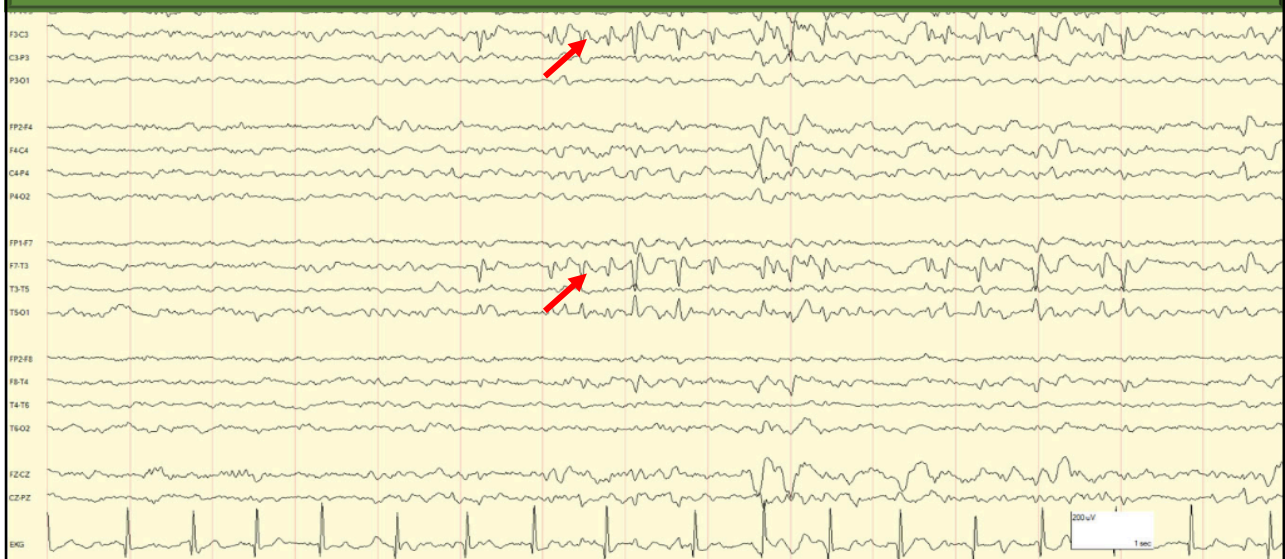
Benign Childhood Epilepsy with Centrotemporal Spikes



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

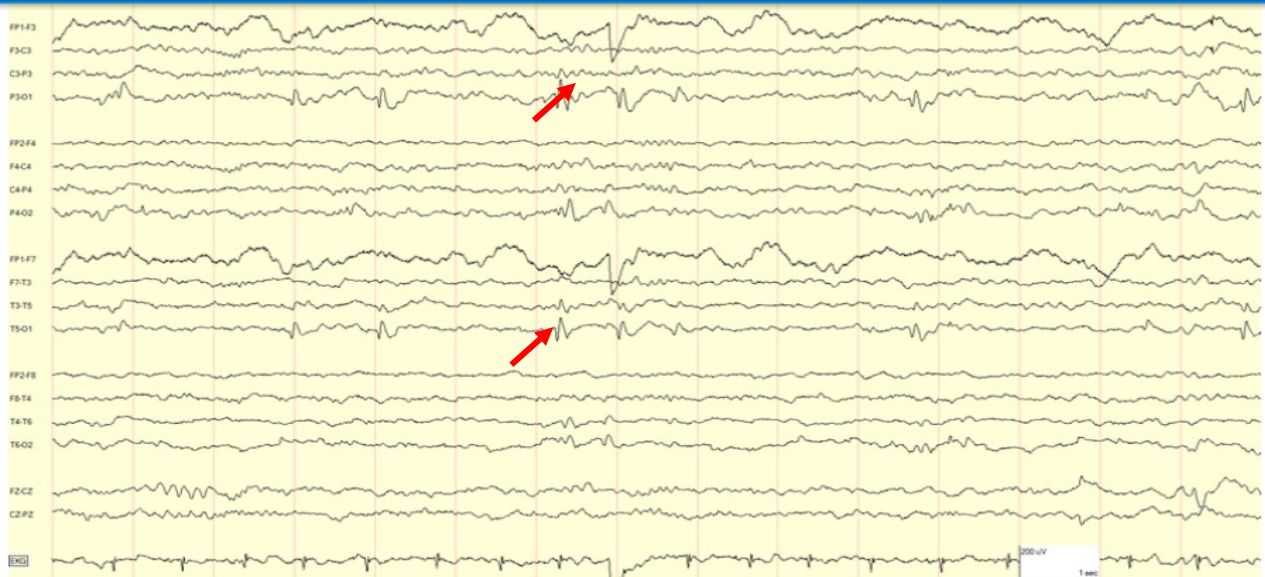
EEG	<p>IEDs: Multifocal SPK/SW 90% Normal single EEG 10% Occipital spikes 60% of patients Low voltage SPK and Gen d/c minority of cases.</p> <p>Activation: Eye closure (elimination of central vision and fixation off sensitivity) may activate occipital spikes. EEG abnormality is enhanced by sleep deprivation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