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EEG Findings in Pediatric Epilepsy.

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Objectives

- Normal Pediatric Findings (infancy >)

1. Background
2. Sleep
3. State changes / sleep transients.

- Age Specific Features
- Electroencephalographic patterns commonly associated with the “named” epilepsy syndromes.

Approach To Epilepsy Syndromes

1. Developmental course.
2. Prototypical Seizures.
3. Electroencephalographic Features

- Ictal
- Interictal / background.

EEG in the diagnosis of epilepsy

- Sensitivity: single recording, interictal epileptiform activity 50%.
- Specificity: 98% (if conservatively read; most “false positives” are misread, some are in the 1st degree relatives of patients with epilepsy)
- Increasing yield: 3 records, 70% sleep/sleep deprivation another 5-10%
- AED does no abolish interictal epileptiform discharges (exception: GGE – e.g., absence epilepsy.)
- A normal EEG does not rule out the presence of epilepsy.

Pediatric Epilepsy

- Approximately 75% of all pediatric epilepsy appears within the first 21 years of life.
- The majority of the named epilepsy syndromes (e.g., Lennox-Gastaut syndrome) have their onset during childhood, with remaining occurring during early adulthood (e.g., JME).
- Pediatric epilepsy syndromes are commonly organized based on age of presentation (i.e., neonatal, infantile, childhood or adolescent onset).
- The importance of accurately classifying an epilepsy syndrome stems on the importance of therapeutic and prognostic implications.

Pediatric Epilepsy Syndrome

- Epilepsy syndrome is a constellation of signs and symptoms occurring commonly in a specific disease state.
- When referring to epilepsy syndromes this means the patients electroclinical syndrome (specific EEG findings in association with specific clinical findings)
- Typically consist of a clinical triad:

1. Development (normal or abnormal)
2. EEG findings (ictal and interictal)
3. Seizure type(s).

Pediatric Epilepsy Syndrome Characteristics

- Define the patient's developmental status
- Define the patient's seizures types.
- Age of onset
- Seizure type(s)
- Interictal EEG
- Ictal EEG.
- Genetics
- Response to EEG.

Indications for an EEG in Pediatric Epilepsy

- Initial diagnosis
- Change in seizure semiology
- Consideration of withdrawing ASD/AEDs.

Epileptic Encephalopathy

- Denotes the concept that the epileptic activity itself is likely directly (and independently) contributing to the patients cognitive and developmental abnormalities, over those expected from the underlying disorder alone.
- Some authors theorize that suppression of the interictal epileptiform activity might minimize additional impairment.

Epilepsy Syndromes – Age Groups

- Neonatal / Infantile Syndromes:
  - Early Myoclonic Encephalopathy (EME)
  - Early Infantile Epileptic Encephalopathy (Ohtahara syndrome).
  - West Syndrome (Epileptic Spams – Infantile spasms)
  - Dravet Syndrome (“Severe Myoclonic Epilepsy of infancy”)
  - Myoclonic Epilepsy of Infancy.
  - Epilepsy of Infancy with Migrating Focal Seizures

- Self-limited neonatal / infantile familial and nonfamilial epilepsy/seizures
- Myoclonic encephalopathy in non-progressive disorders
- Genetic Generalized Epilepsy with Febrile Seizures Plus (GEFS+)

Posterior Dominant Rhythm Frequency

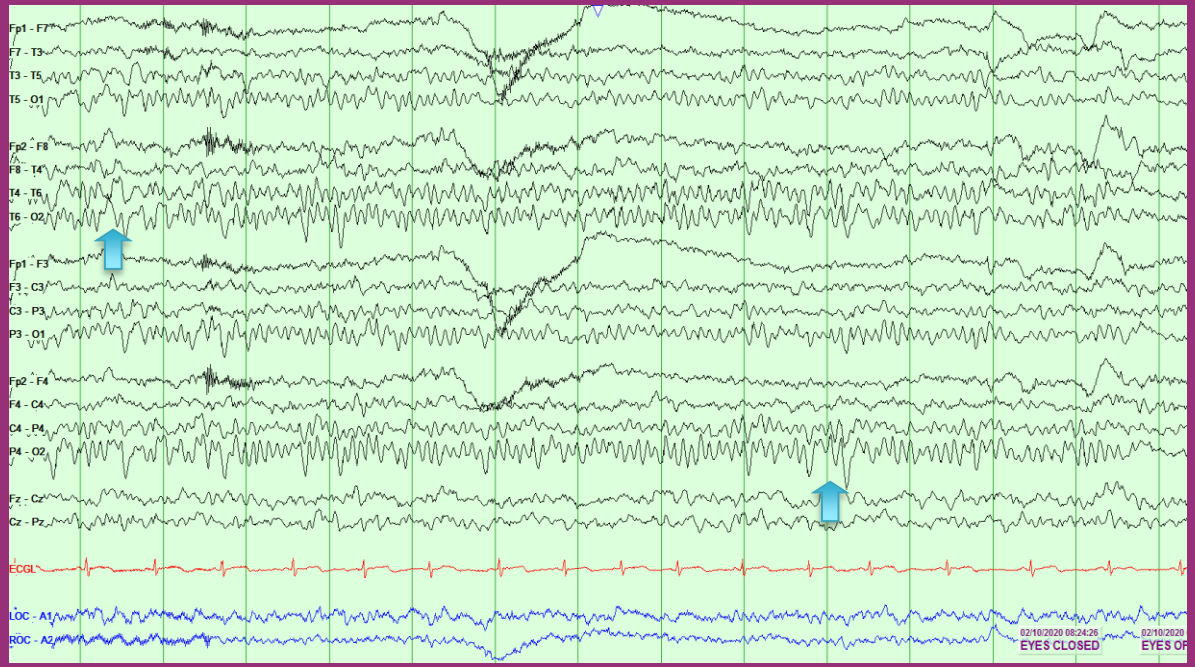
Average PDR Frequencies

- 4 months – 4 Hz
- 6 months – 5 Hz
- 12 months – 6 Hz
- 24 months – 7 Hz
- 36 months – 8 Hz
- 8 years – 9 Hz
- 10 years – 10 Hz

Abnormal PDR

- 1-year-old: <4 Hz
- 4 year-old: <6 Hz
- 5-year-old: <7 Hz
- ≥ 8-year-old: <8 Hz

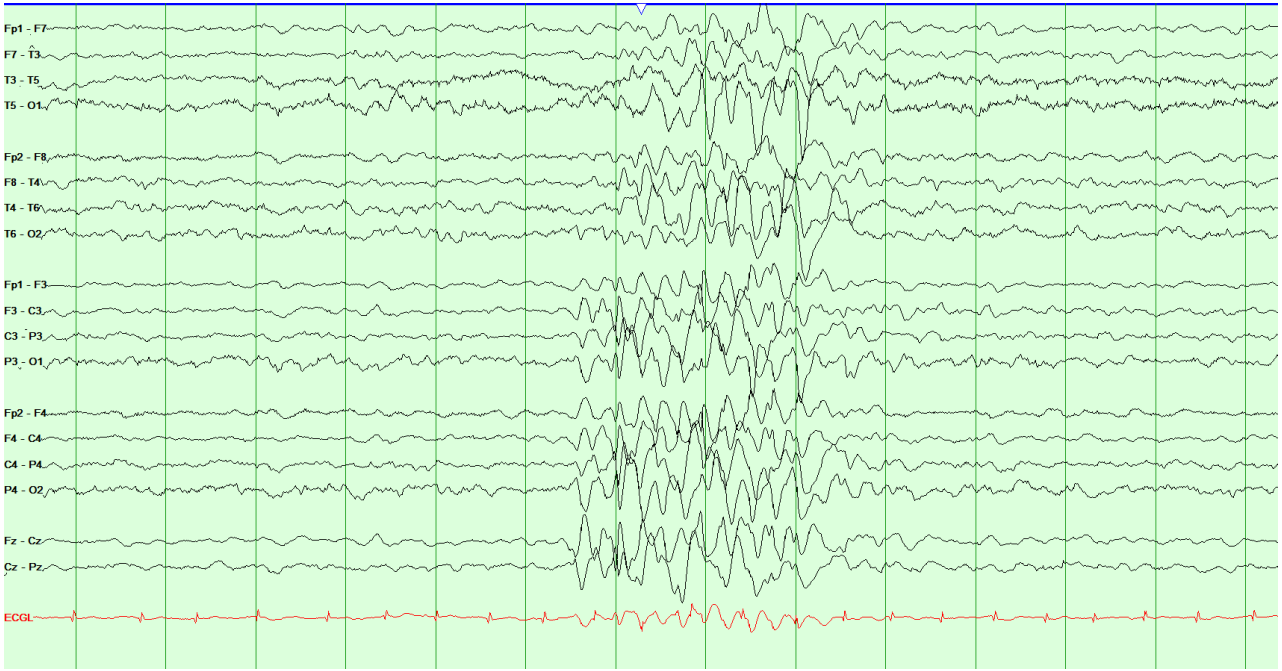
Posterior Slow Waves of Youth (PSWY)



6-year-old male

- Delta-theta admixed with the patients PDR.
- Best appreciated during relaxed wakefulness.
- Commonly seen in children between the age of 8-15.
- Uncommon in children <2-years of age.
- Infrequent (~15% incidence) between the age of 16-20.
- Rare in adults (>21-years)
- Follows the same rules as the patients normal PDR.

Hypnagogic Hypersynchrony



- Appearing during state transitions.
- Characterized by bilateral synchronous, high voltage, rhythmic 3-5 Hz activity.
- Can appear as early as 2-3 months of age.
- At 9-months, more prominent and continuous.
- Few infants do not show this pattern bit show occipital or widespread rhythmic and synchronous 4-5 Hz waves.
- Rare in older children (some authors still consider normal up to age 12-13 years).
- Occasional small sharps or spike discharges may be seen admixed with the waveforms.

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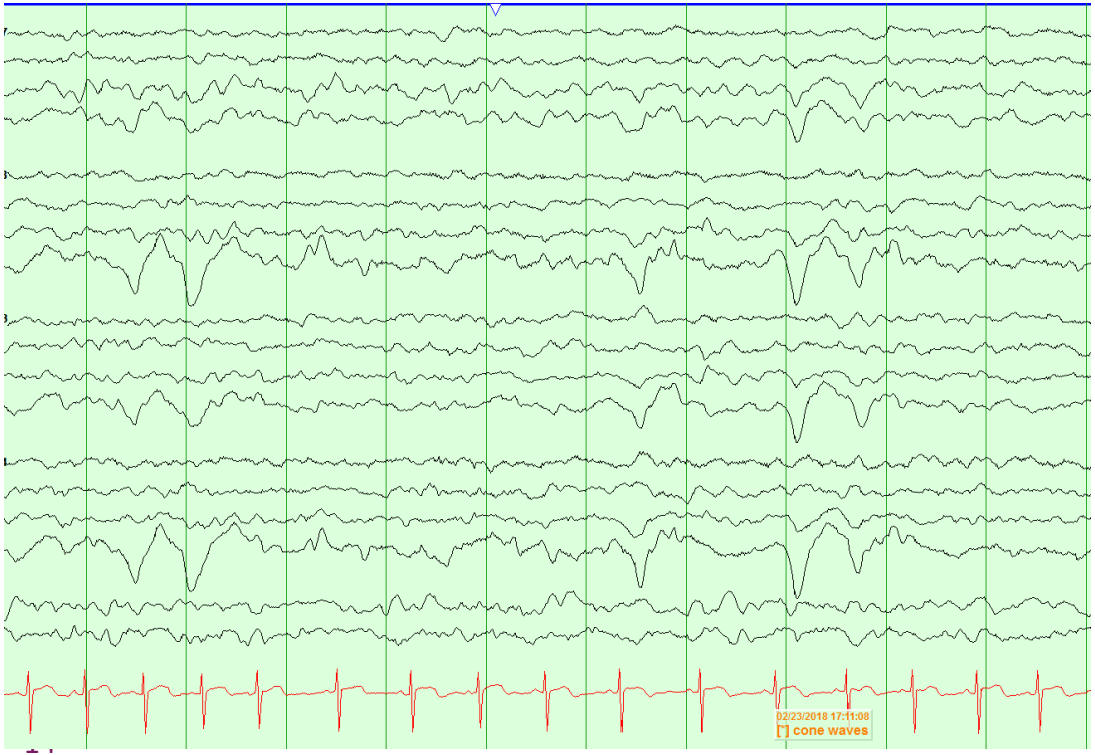
Occipital Slow Transients of Sleep (Cone Waves).



In children, the transition from light to deep sleep may be associated with bilateral high-voltage slow transients in the occipital regions.

These waves vary from a cone-shaped configuration to a biphasic slow transient. These transients occur every 3-6 seconds during light sleep and more frequently during deeper stages of sleep.

Cones or O waves (arrow) are physiologic waves presenting during non-REM sleep from infancy until 5 years of age.

They are isolated, medium-to-high amplitude, monomorphic triangular shaped, delta waves with a typical duration > 250 msec that occur over the occipital region

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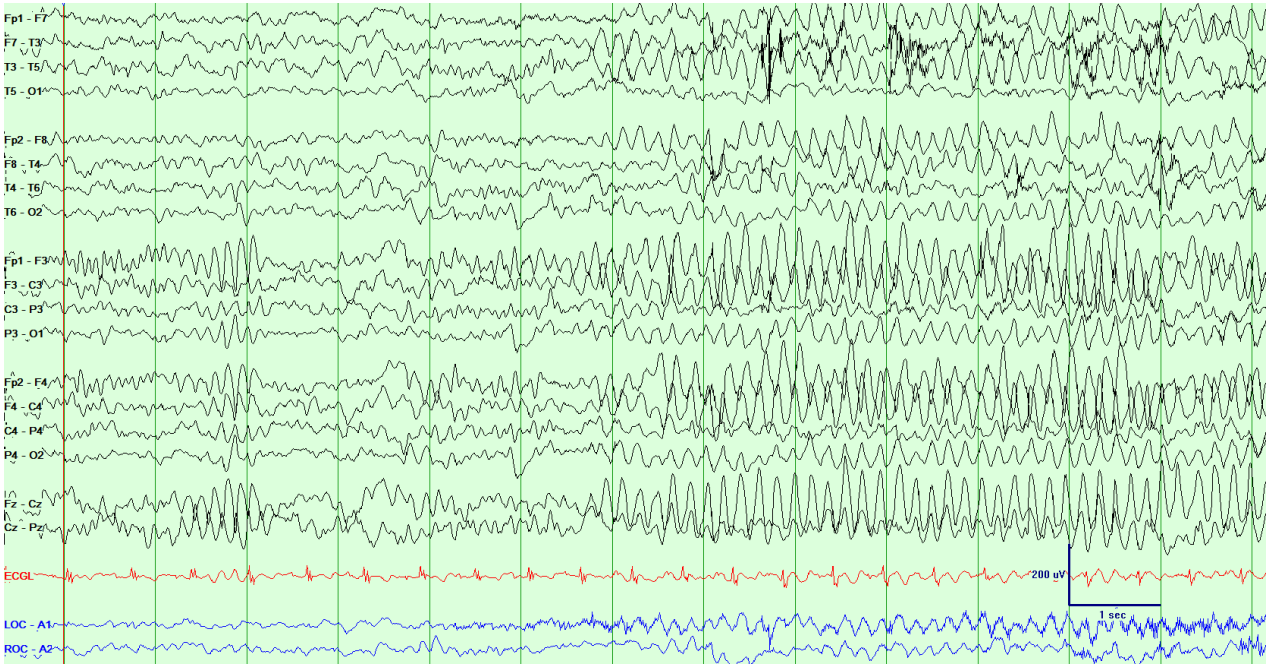
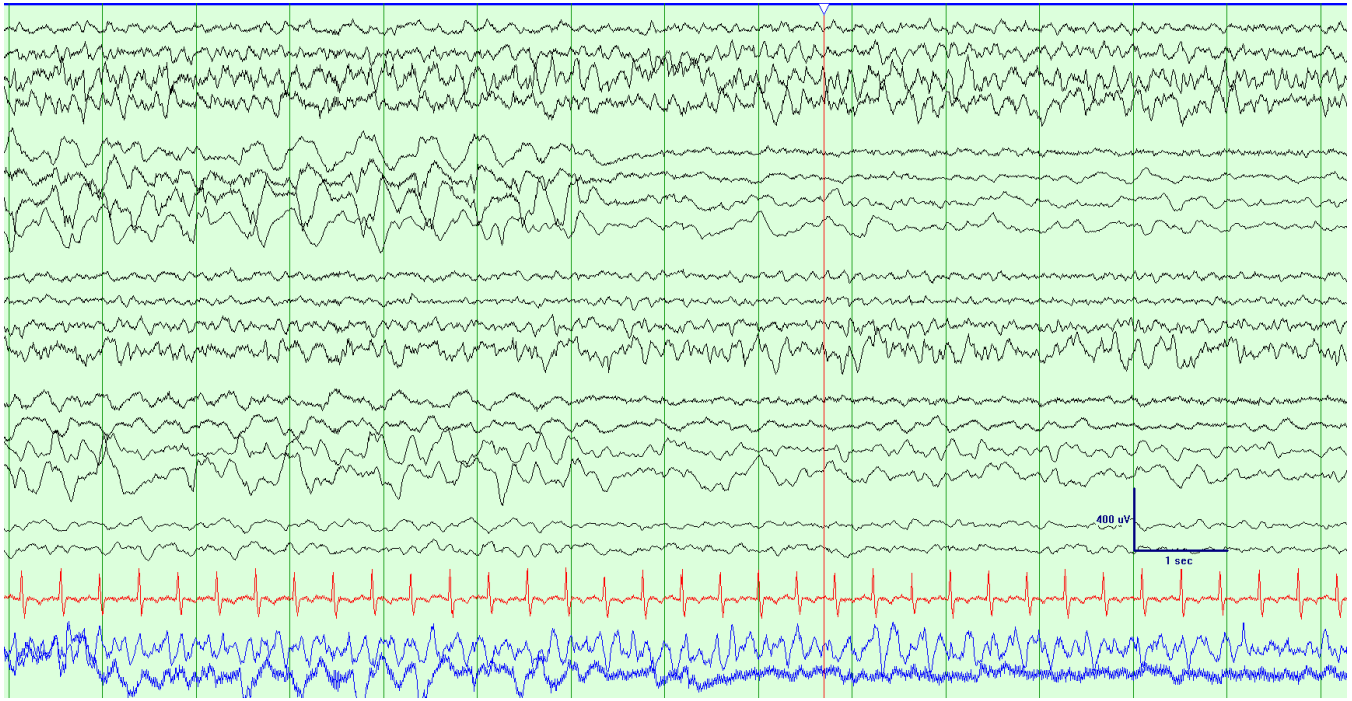
Frontal Arousal Rhythm (FAR)



- Rare nonspecific EEG pattern of no clinical significance.
- Frontal regions (F3-F4) electrodes with minimal spread to nearby scalp areas) during arousal from sleep in children.
- Characterized by 30-50 uV, predominantly monophasic negative waves, occurring in bursts or runs lasting up to 13 seconds (usually ~5 sec) with a characteristic notching of the ascending or descending phase of each wave that may represent harmonics of the waveforms.
- The waxing and waning of the amplitude often leads to a spindle-like morphology.

Neonatal / Infantile Onset Epileptic Encephalopathies.

Epilepsy of Infancy with Migrating Focal Seizures



1) Genetic etiology; KCNT1, KCNQ2, SCN1A, SCN2A.

2) Multiple seizure foci with ictal discharges migrating randomly, but sequentially from one region to another during the same ictal discharge.

3) Background is normal prior to the onset of seizures, followed by progressive slowing

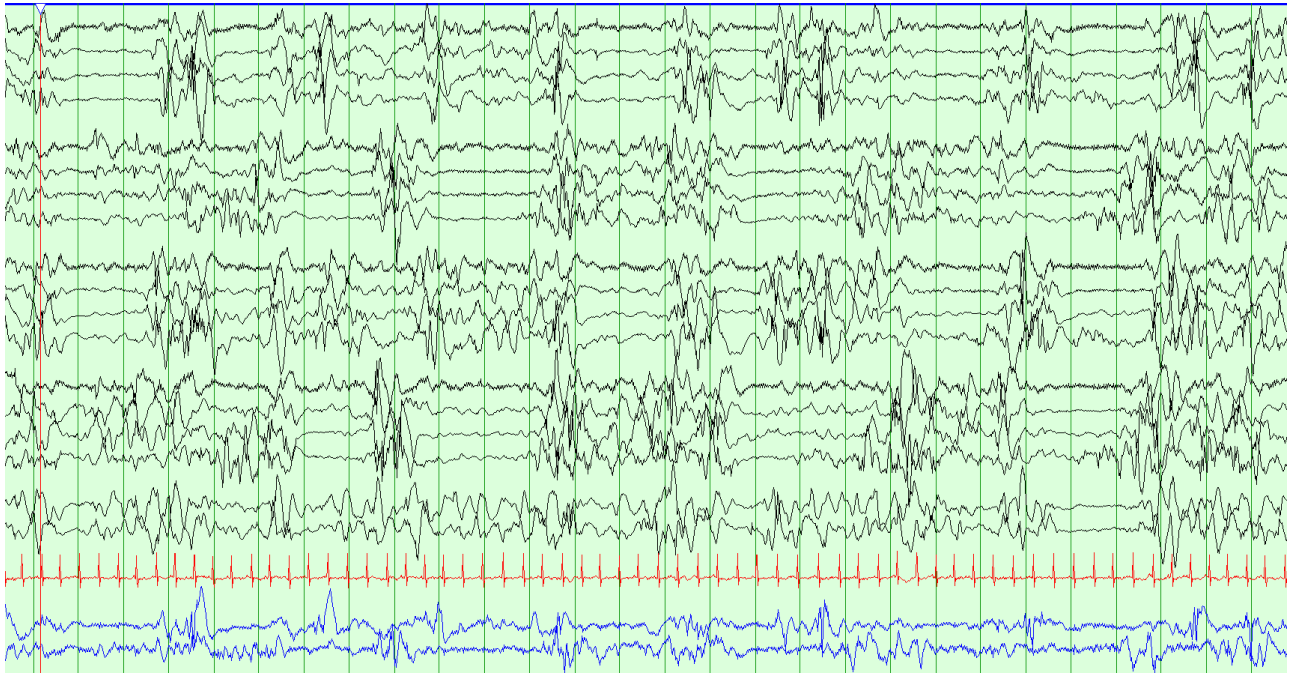
4) Seizure are characterized by prominent autonomic features.

5) Considered an epileptic encephalopathy.

6) Seizures are refractory and prognosis is poor.

Neonatal / Infantile Onset Epileptic Encephalopathies.

Early Infantile Epileptic Encephalopathy – EIEE (Ohtaha Syndrome)



- Early myoclonic encephalopathy (EME) and early infantile epileptic encephalopathy (EIEE – Ohtahara syndrome)
- Can also bee seen in patients with MCD, Aicardi syndrome, glycine encephalopathy and metabolic disorders.
- The EEG shows bursts, lasting several seconds of polyspikes alternating with very low-voltage activity, this combination being called suppression-burst pattern (S-B)
- For Ohtahara the S-B pattern appears consistently throughout the awake and asleep tracing.
- For early myoclonic encephalopathy (EME) the S-B pattern may only be seen during sleep.

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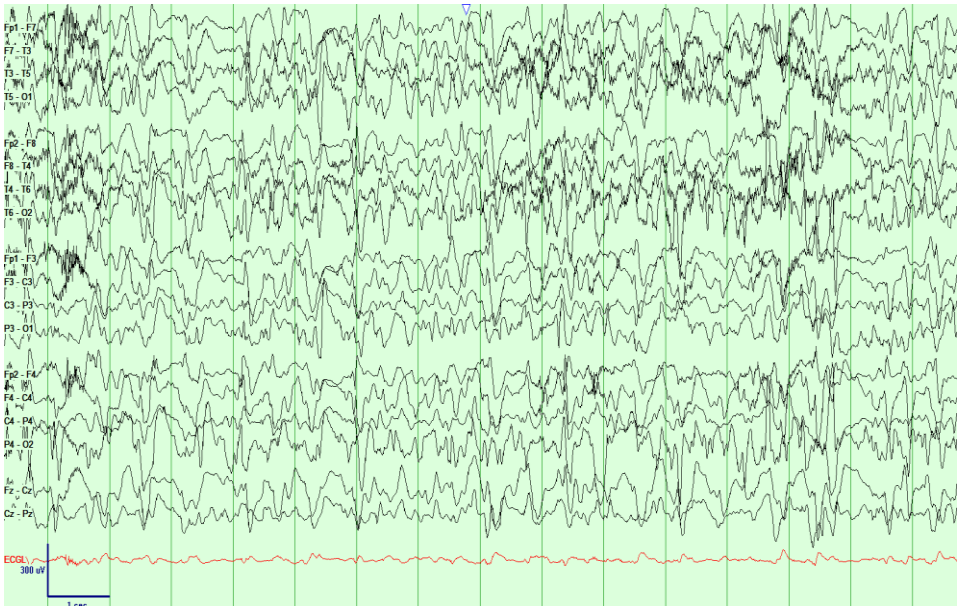
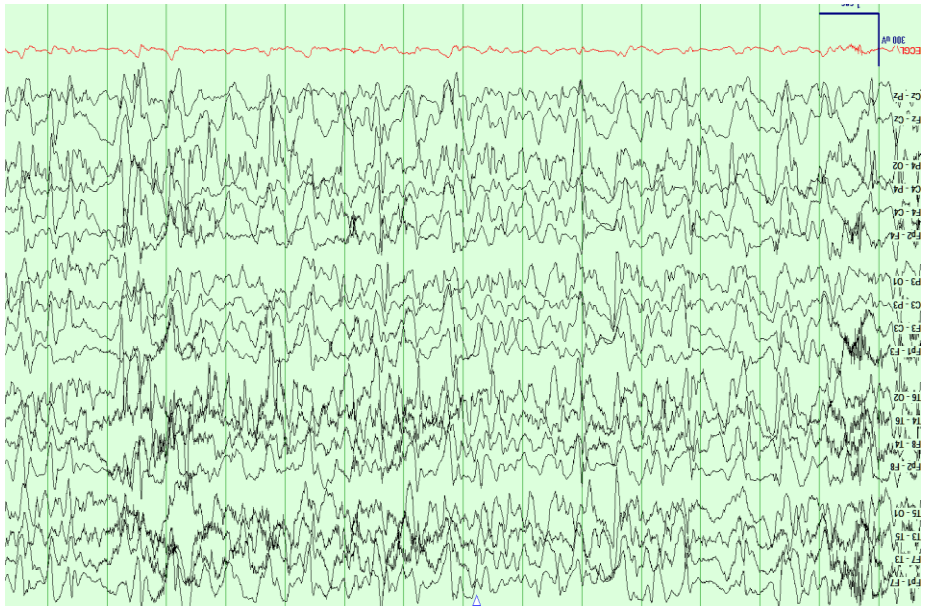
West Syndrome

- Heterogenous etiology (structural, metabolic, genetic, etch.)
- Typical age of onset range 3-12 months.
- An electroclinical syndrome characterized by the triad of:
- Developmental delay/regression (often normal prior to the onset)
- Interictal EEG pattern of hypsarrhythmia (can be absent at the onset)
- Seizure type of epileptic spams (mandatory)
- Often evolve into other epilepsy syndromes (e.g., LGS)
- Often other epilepsy syndrome evolve into West syndrome (e.g., EIEE)
- Treatment: neuro-hormonal therapies (e.g., ACTH).

West Syndrome – Hypsarrhythmia





LFF 1 Hz, HFF 70 Hz, Notch 60 Hz, Sensitivity 15 uV/mm, Timebase 30 mm.sec

Reversed

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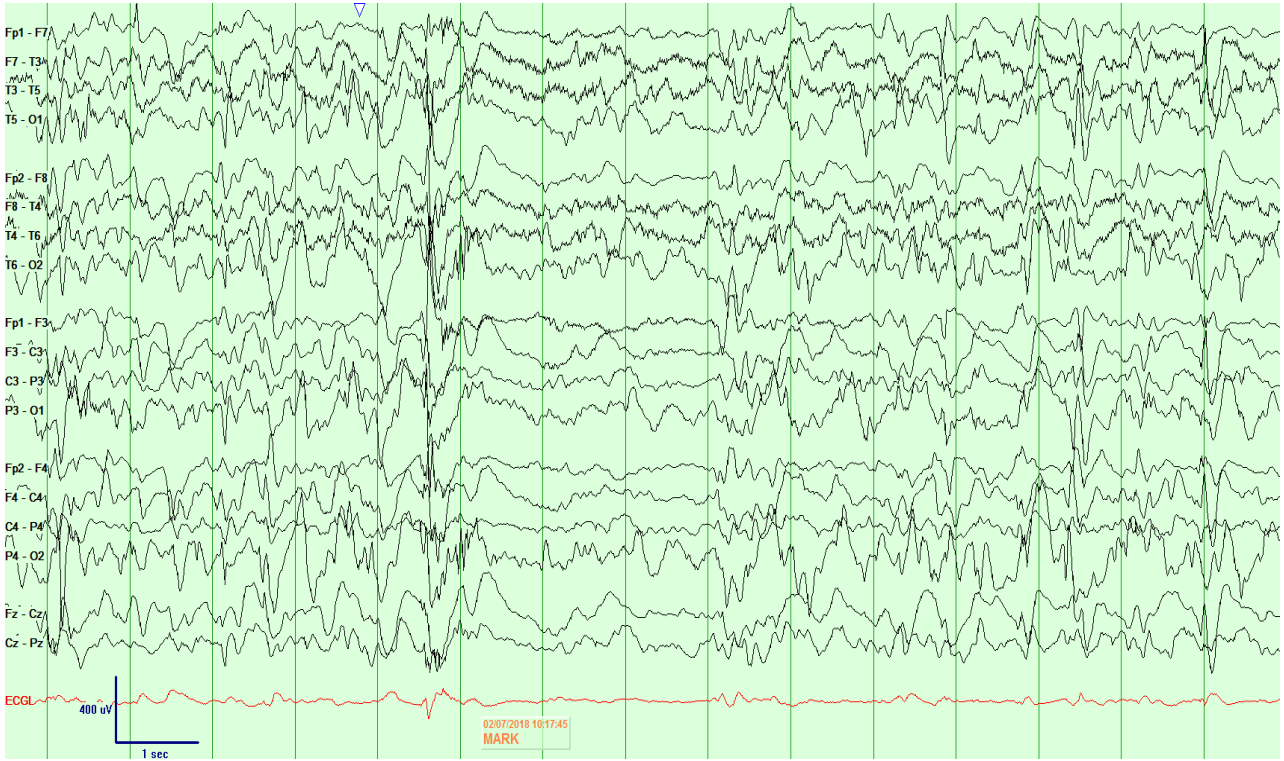
West Syndrome – Hypsarrhythmia

- Definition coined by Gibbs and Gibbs (1952)
- Derived from the Greek word *hypselos*, meaning high.
- Characterized by chaotic (disorganized), high-amplitude (>300 µV)
- Admixed, abundant, multifocal sharp(spike/polyspike)-wave discharges and low amplitude paroxysmal fast activity.
- At the onset findings only (more commonly) seen during non-REM sleep.
- Variants exist (“modified hypsarrhythmia), characterized by asymmetry, interhemispheric synchronization, focal IED.
- The prognosis does not differ with hypsarrhythmia variants.

West Syndrome – Epileptic Spasms



- Electrodecremental response lasting several seconds.
- Four components:
- High-amplitude diffuse (vertex maximal) positive slow-wave.
- Admixed multifocal epileptiform discharges riding the underlying slow wave.
- Diffuse attenuation of the background rhythms.
- Overriding low-voltage fast rhythms.

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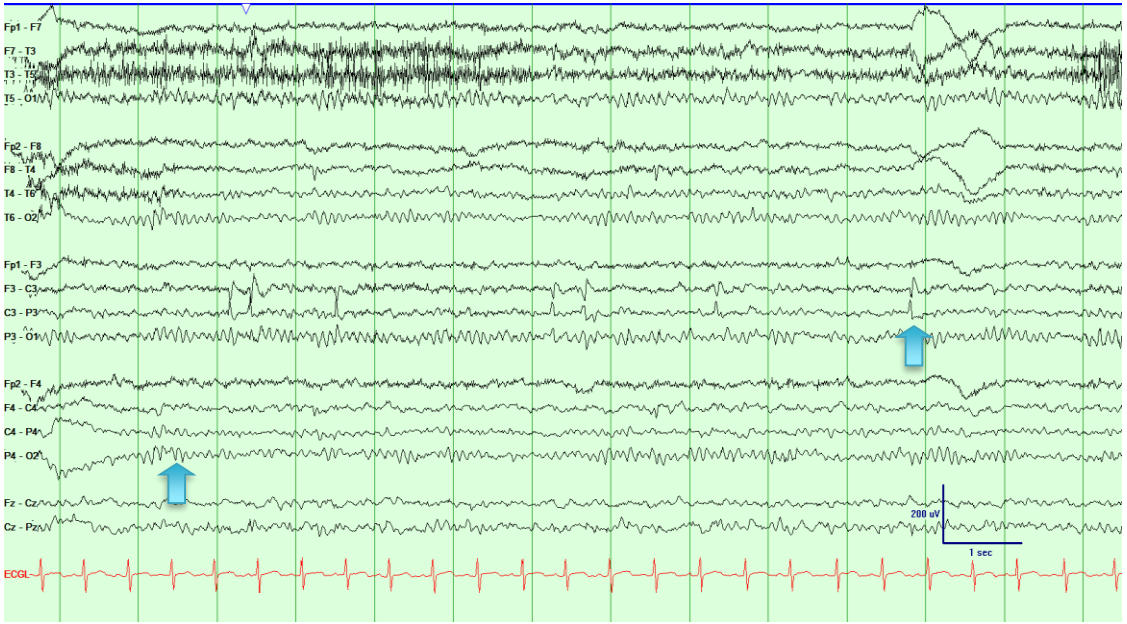
Self-Limited Focal Epilepsies of Childhood.

Childhood Epilepsy with Centro-Temporal Spikes (CECTS) - BRE



7-year-old male presenting with stroke-like episode

1) ~10-25 of new onset pediatric epilepsies.

2) Typically normal background.

3) Stereotyped, high-amplitude, diphasic spike/sharp-wave discharges.

4) Commonly located over the central (~40%) or centro-temporal (~30%) regions.

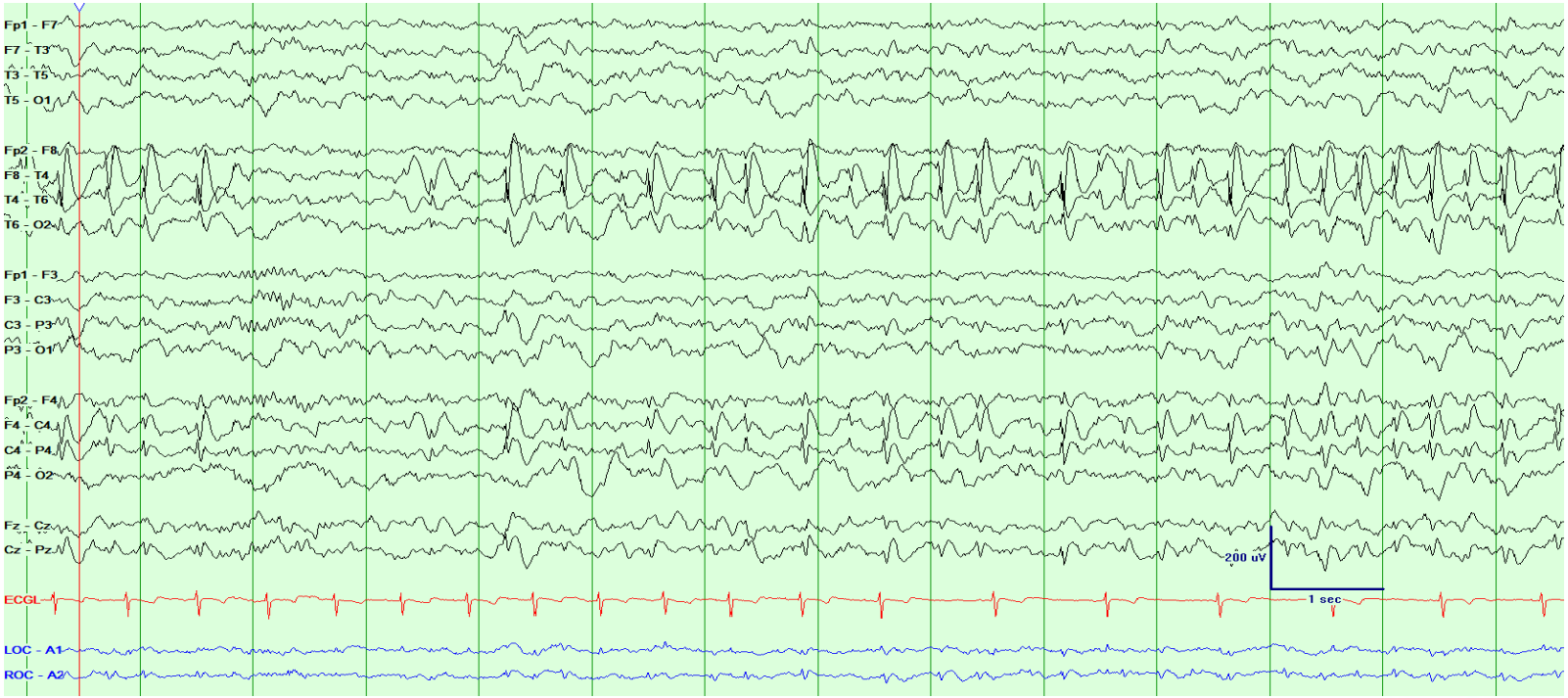
5) Associated with a horizontal dipole with maximal negativity over C3/T5 or C4/T6 and frontal positivity. Only found ~50% of patients.

6) EEG features are accentuated by sleep.

7) Can be unilateral (60%) or bilateral (40%).

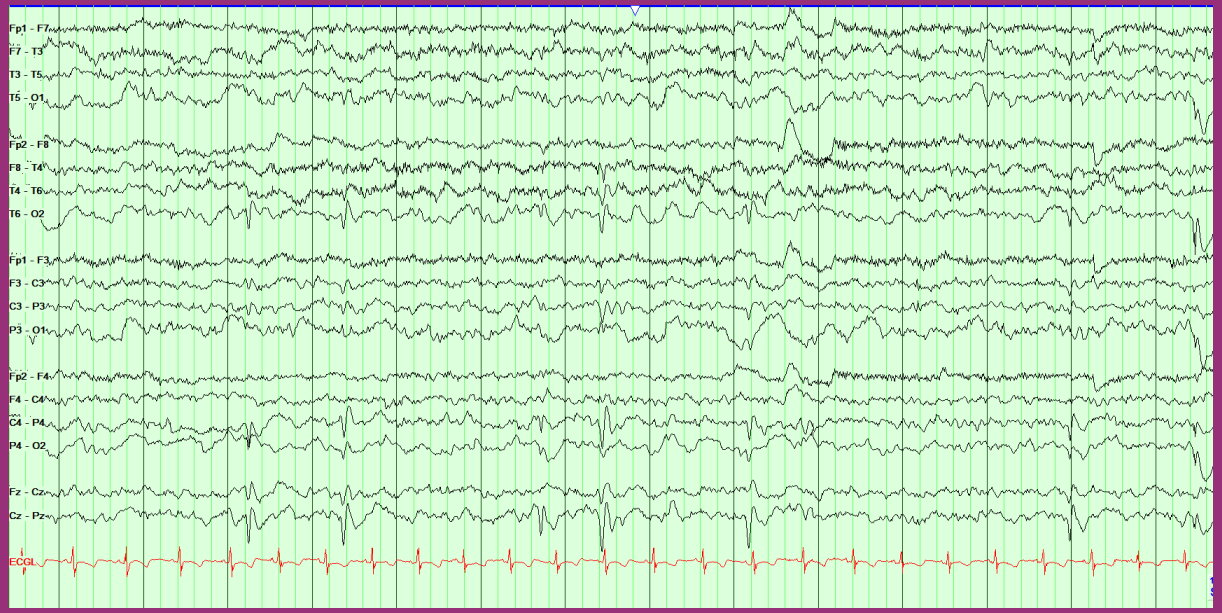
8) ~30% of patients only have abnormalities during sleep.

Childhood Epilepsy with Centro-Temporal Spikes (CECTS) - BRE



Self-Limited Focal Epilepsies of Childhood

Panayiotopoulos Syndrome (Early Type Occipital Epilepsy)



2-year-old male presenting with episodes of nocturnal emesis and confusion

1) ~5-10% of childhood onset epilepsies.

2) Interictal findings demonstrate multiple IED foci, at shifting, with an occipital predominance.

3) Marked variability, ranging from normal to multifocal, varying greatly within the same patients in sequential EEGs.

4) Small percentage of patients (<10%) have consistently normal awake and asleep interictal EEG.

5) Occipital spikes are common, but not necessary for the diagnosis. Frontal and centro-temporal spikes are common.

6) Seizure burden is minimal with most patients experiencing infrequent seizures (2-5 on average).

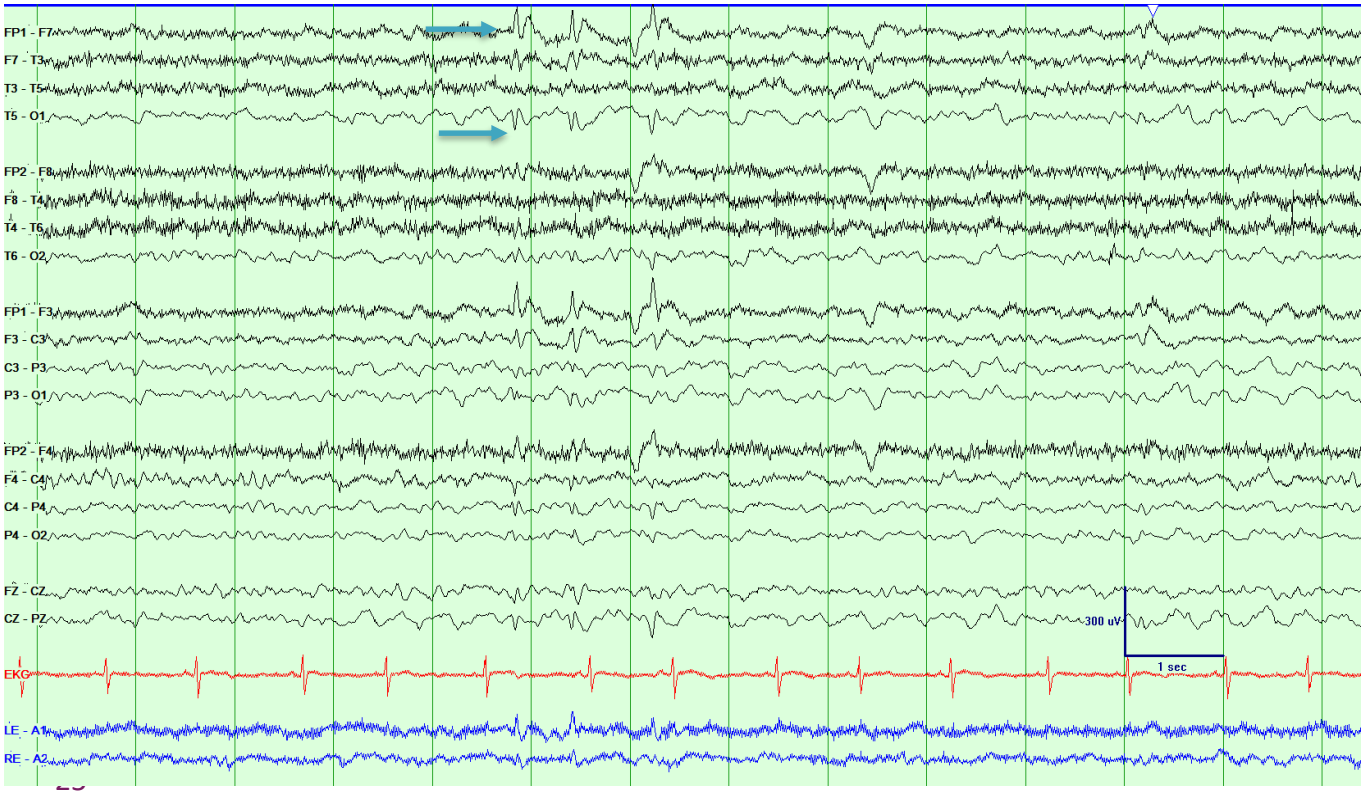
7) Normal background.

8) Stereotyped, diphasic, moderate to high amplitude discharges.

9) EEG abnormalities accentuated by sleep.

Fronto-polar / Occipital Spike-Waves (Fp-O)

Panayiotopoulos Syndrome



Shifting pattern occipital sharps to the frontal-polar regions, resulting in secondary fronto-polar synchrony.

Age dependent phenomenon

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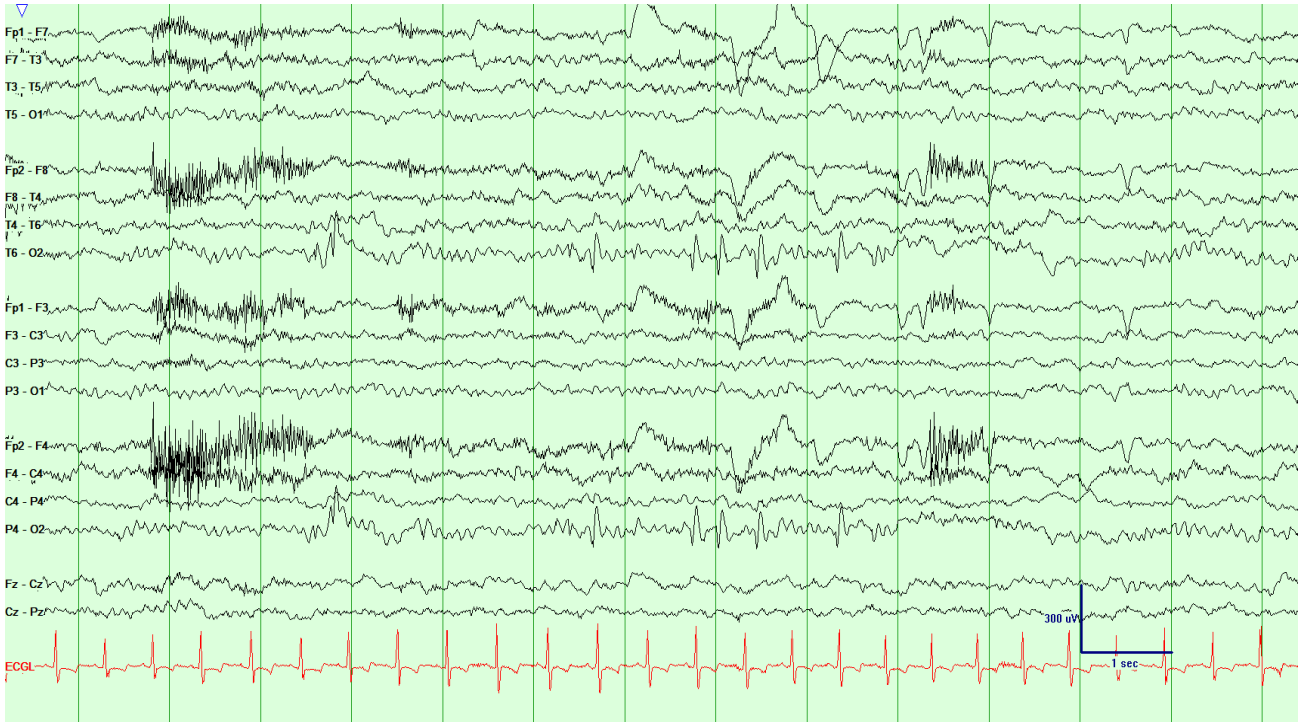
Self-Limited Focal Epilepsies of Childhood

Childhood Occipital Epilepsy - Late-Onset (Gastaut Type)



10-year-old female presenting with a diagnosis of “migraine with aura”

1) ~2-7% of new-onset childhood epilepsy.

2) High amplitude stereotyped occipital spikes or paroxysms demonstrating fixation-off sensitivity (FOS).

3) FOS refers to IED or seizures elicited by elimination of central vision and fixation, commonly seen in idiopathic occipital lobe epilepsies, but not pathognomonic.

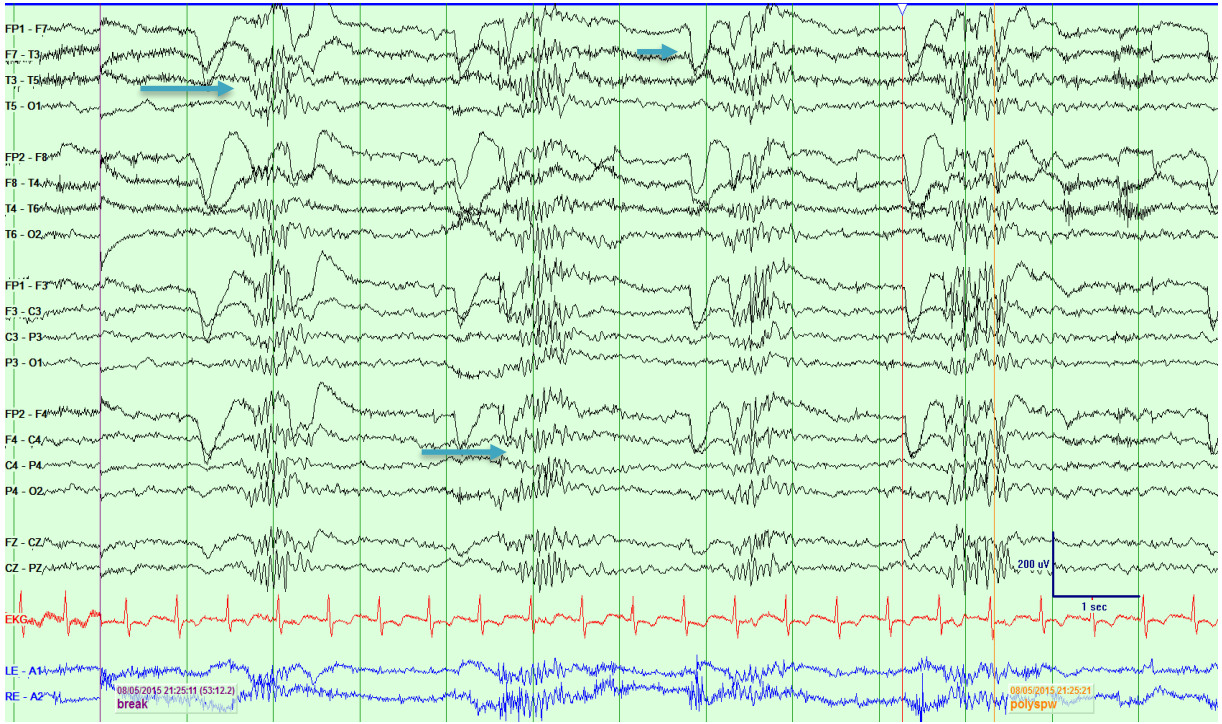
4) EEG abnormalities are accentuated by sleep.

5) Seizure are manifested by complex and elementary visual hallucination, eye deviation, forced eye closure, ictal blindness and ictal/postictal headaches.

6) Seizure burden is high and treatment is often necessary.

Genetic/Idiopathic Generalized Epilepsies

Epilepsy with Eyelid Myoclonias (Jeavons Syndrome)



1) Ictal discharge of 3-6 Hz generalized polyspike-wave following eye closure.

2) Fixation off sensitivity / eye closure will often elicit a seizure.

3) Photosensitivity is mandatory

4) Normal background.

5) EEG remains abnormal, despite seizure control.

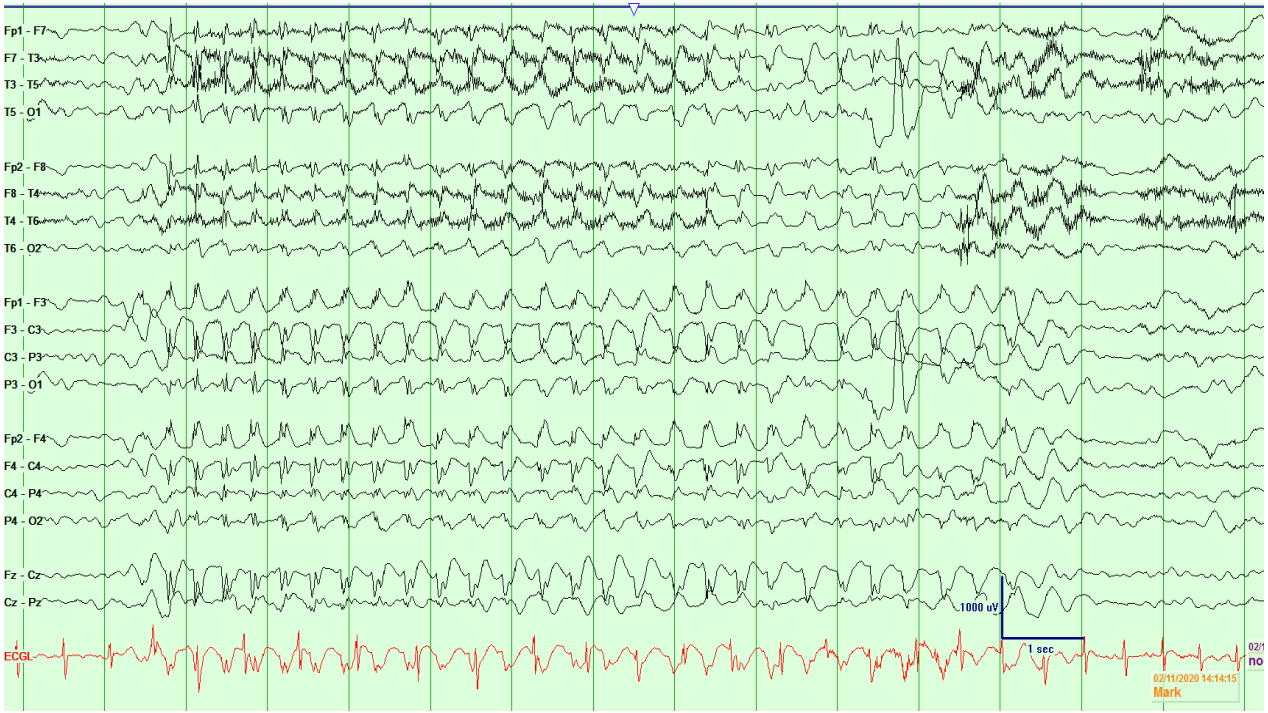
6) Ictal discharges can often be eliminated by elimination of light.

7) Self-induction seen in ~10% of patients, often in patients with ID.

9-year-old female presenting with episodes excessive eye blinking

Genetic/Idiopathic Generalized Epilepsies

Childhood Absence Epilepsy - CAE



7-year-old male

1) ~8-15% of childhood epilepsies

2) Ictal discharge: stereotype high-amplitude, generalized (frontal predominant) 3-Hz spike-and-wave paroxysms.

3) Typical duration 10-20 seconds, but shorter seizures (~3 sec.) are often associated with alteration of awareness.

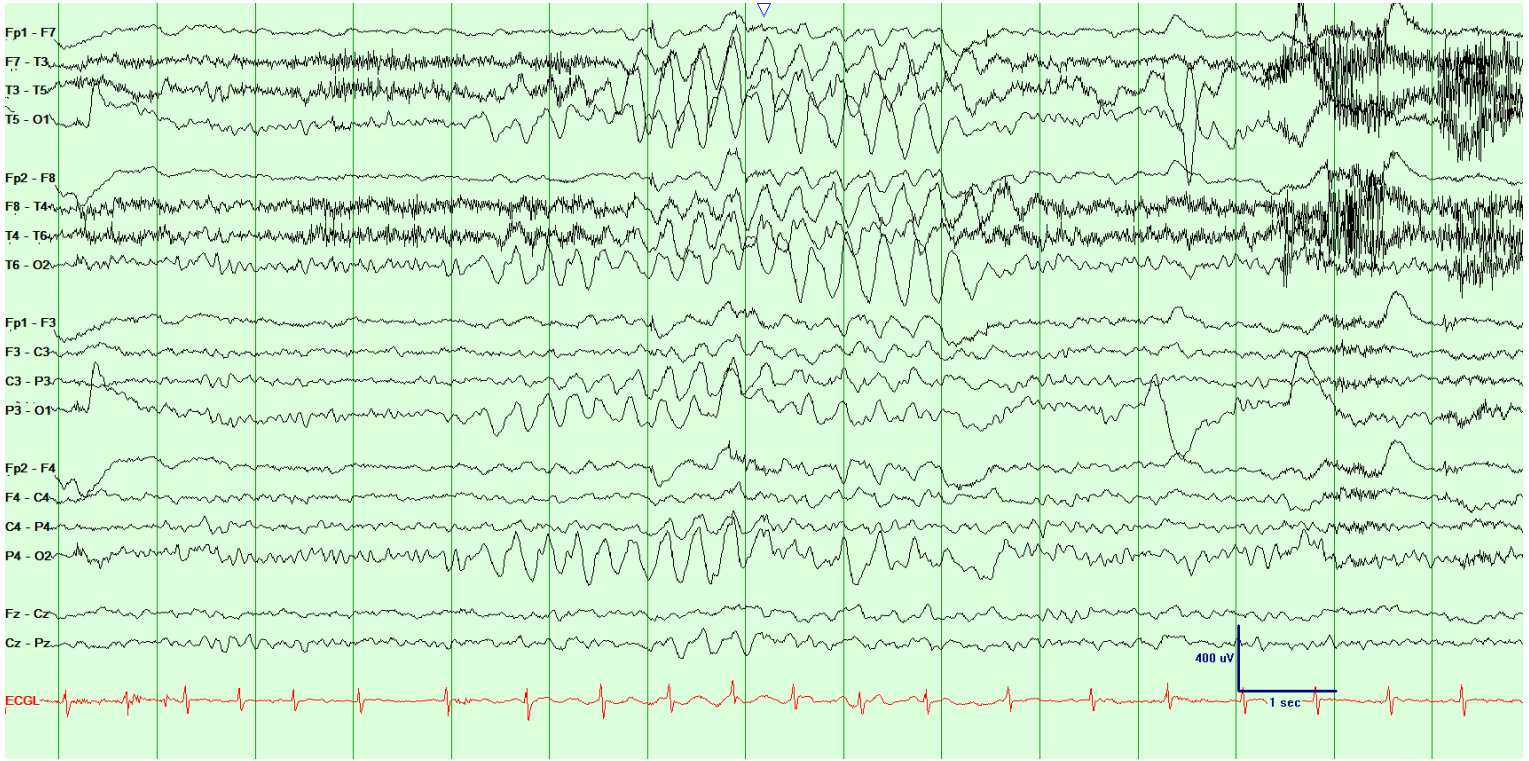
4) Occipital intermittent rhythmic activity (OIRDA), often with a notched appearance can be seen in up to 70% of patients.

5) OIRDA thought to be predictive of elicitation of GSW during HV and favorable response to ASD.

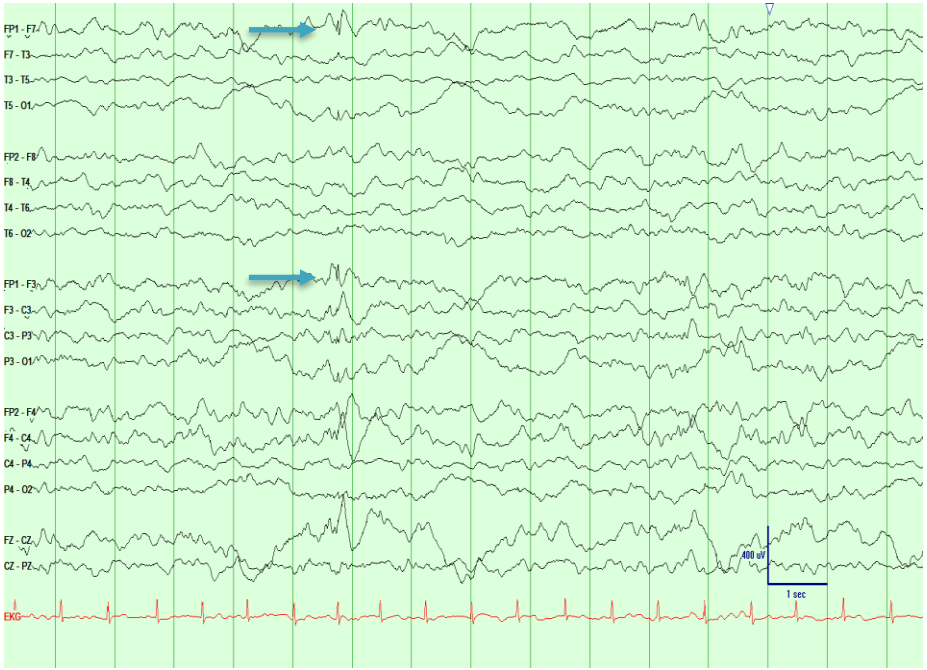
6) Normal background.

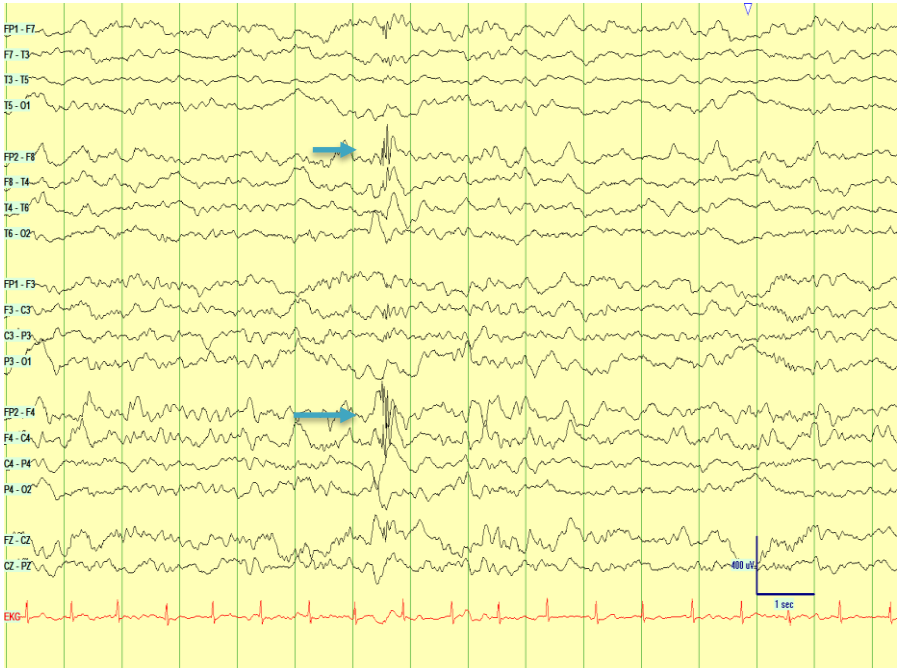
7) Interictal frequent burst of 3-Hz paroxysm, GSW and fragmentation during sleep.

Occipital Intermittent Rhythmic Delta Activity (OIRDA)



Fragmentation of Generalized Spike-Waves.

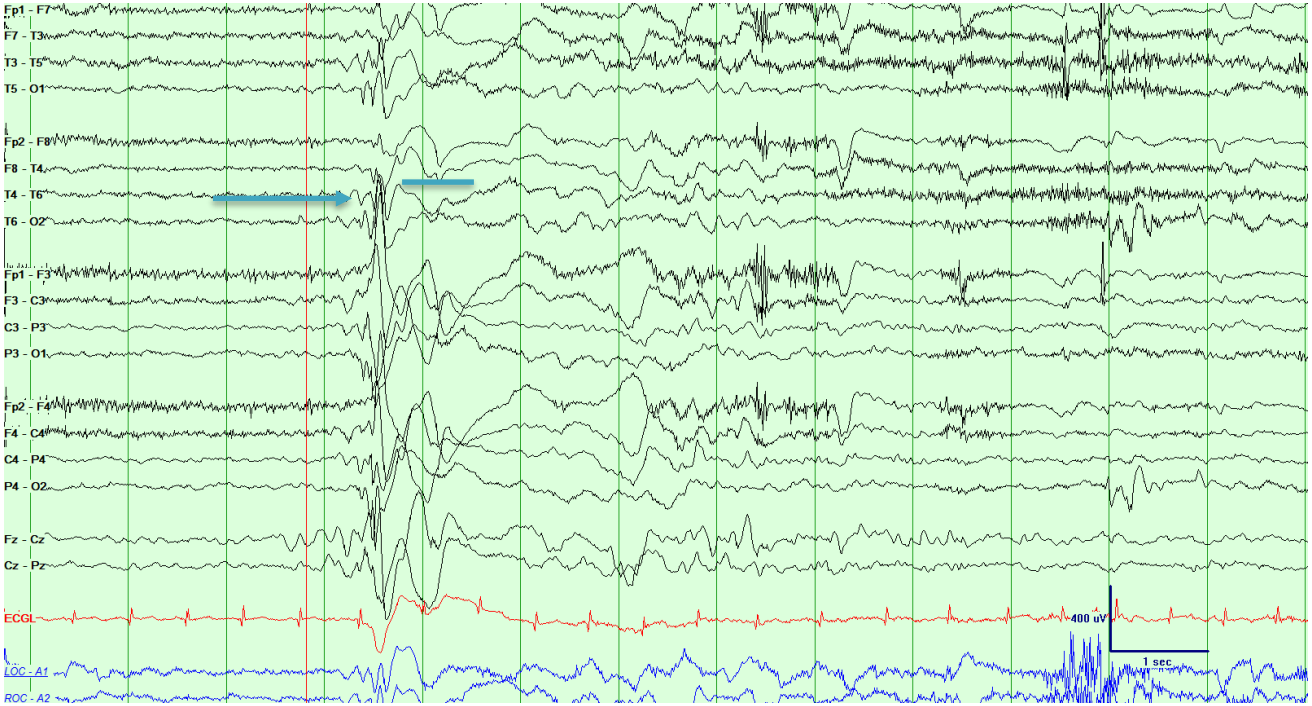




Fragmentation of the generalized interictal epileptiform discharges during wakefulness and sleep (more commonly) are common features of genetic generalized epilepsy. Are often characterized by polyspike with shifting laterality (often anteriorly distributed). Often described as forme fruste of generalized spike-and-wave discharges. Often precipitated by sleep deprivation.

Genetic/Idiopathic Generalized Epilepsies

Epilepsy with Myoclonic-Atonic Seizures (Doose Syndrome)*



1) ~1-2% of childhood epilepsies.

2) Considered an epileptic encephalopathy.

3) Myoclonic-atonic seizures are mandatory seizures for this epilepsy syndrome.

4) The myoclonic component is associated with generalized spike/polyspike-waves and the atonic component with the high amplitude after-slowing.

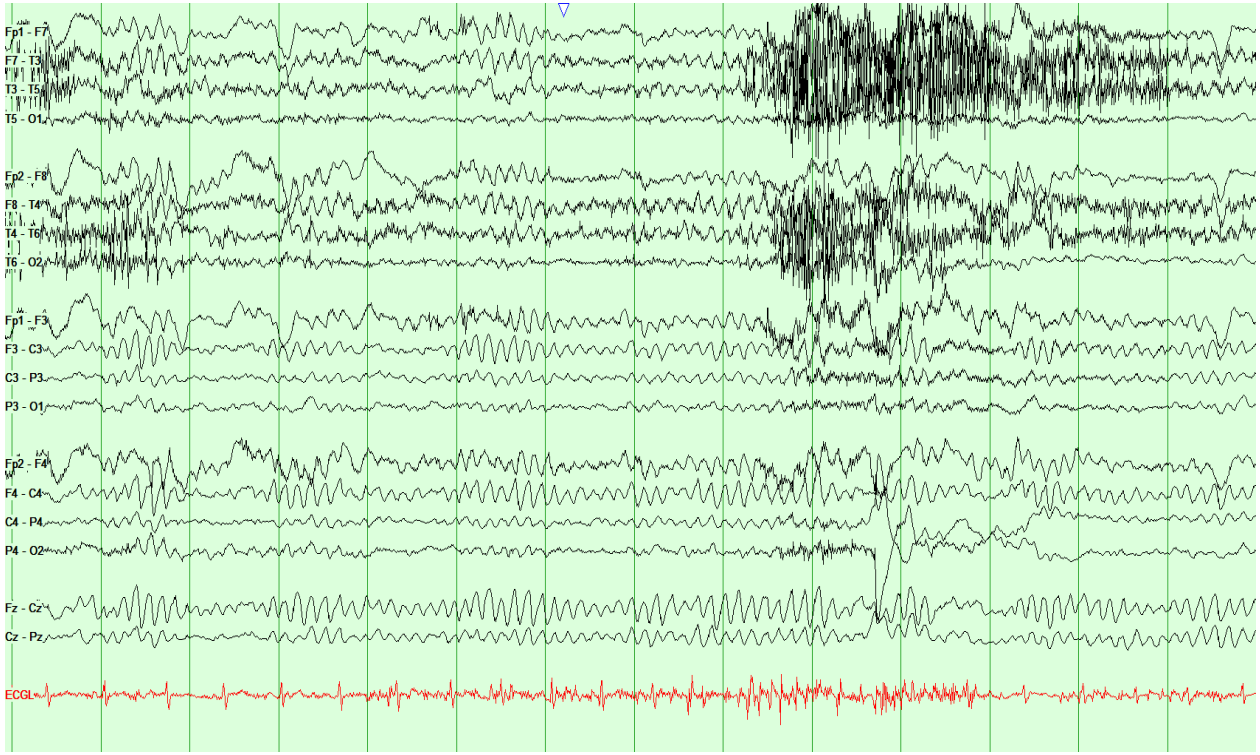
5) Background initially normal, but as the disorder progresses bi-parietal theta activity emerges.

6) Photosensitivity has been reported.

7) Variability in the prognosis, ~50% of patients with normal or mild cognitive deficits with resolution of epilepsy.

4-year-old with history of febrile seizures now presenting with episodes abrupt falls.

Bi-parietal (parasagittal) Theta Activity



4-year-old with history of febrile seizures now presenting with episodes abrupt falls

- Nonspecific Pattern:
- Rett syndrome
- Doose syndrome
- Dravet syndrome
- Angelman Syndrome

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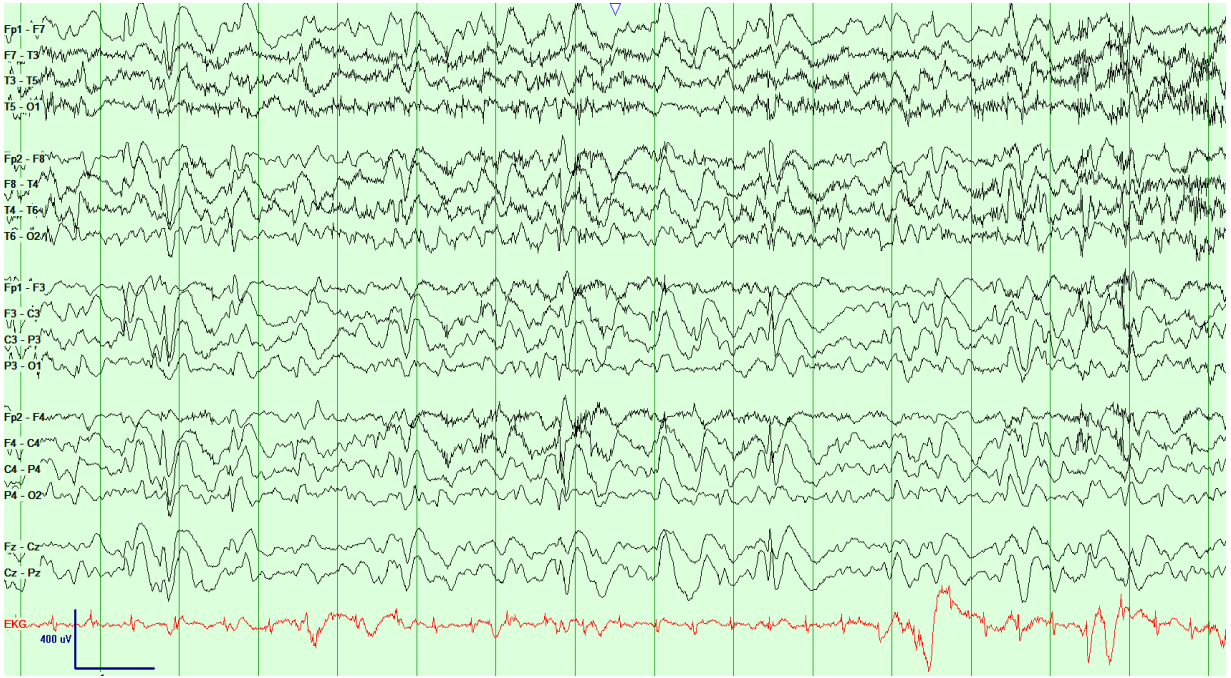
Dravet Syndrome (Severe Myoclonic Epilepsy of infancy)

- ~75% of patients carry a mutation in the SCN1A gene.
- Age of onset ~6-months.
- Temperature sensitive epilepsy (both environmental and fever).
- Experience hemiconvulsive febrile seizures, and develop febrile and febrile seizures.
- Typical seizures include atypical absence, myoclonic and atonic seizures.
- Medically intractable seizures, associated with developmental delay (considered an epileptic encephalopathy).
- Interictal EEG: initially normal, background slowing seen later on. Generalized spike-and-wave discharges and multifocal IED.
- Ictal EEG: depends on the EEG.

Lennox-Gastaut

- Heterogenous etiology (common final pathway for many epileptic encephalopathies).
- Age of onset/diagnosis is prior to 8 years of age (peak 3-5 years)
- Clinical triad of multiple generalized seizure types (tonic > atypical absence > atonic > GTC seizures > myoclonic), developmental delay and signature EEG pattern.
- Interictal EEG: slowing of the background cerebral rhythms for age. Slow (<3 Hz) spike-and-wave discharges. GPFA activity during sleep.
- Ictal EEG: tonic > atypical absence > atonic > GTC seizures > myoclonic

Lennox-Gastaut Syndrome (LGS)



- Slow and poorly modulated background rhythms.
- Focal or multifocal spike(sharp)-and-wave discharges.
- So called slow spike-and-wave discharges (<2.5 Hz) with an anterior predominance.

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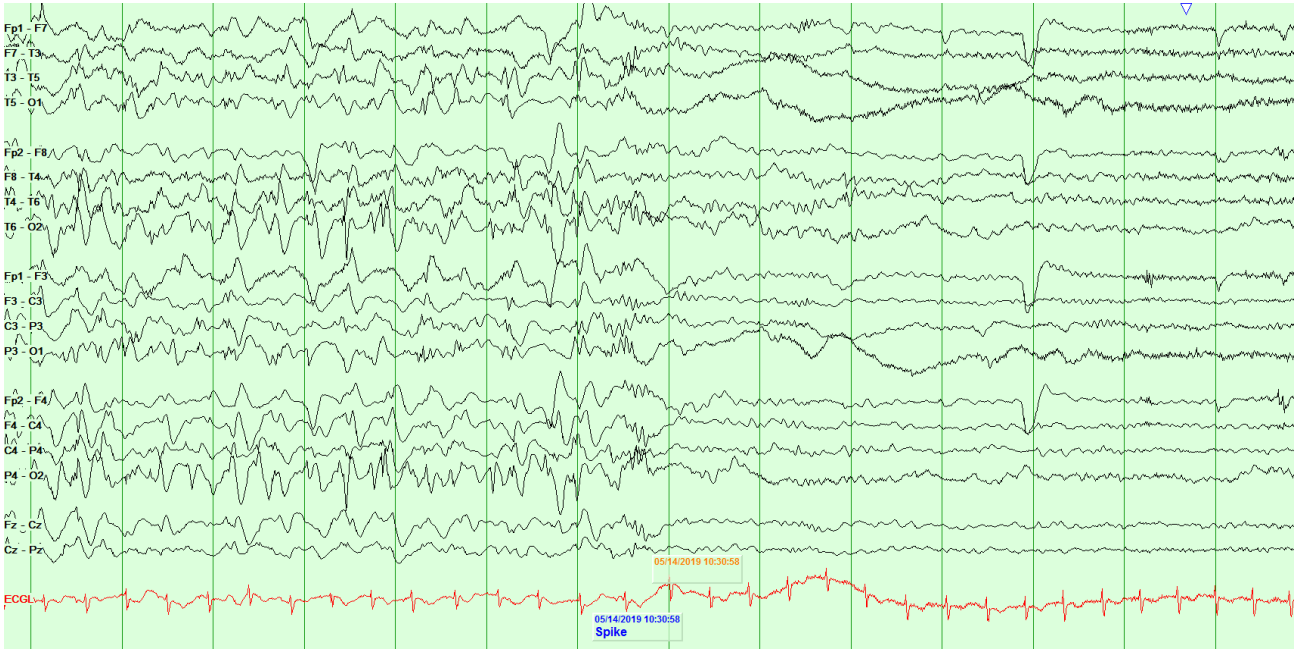
Tonic Seizure – Ictal Discharge.



- Typically arise from sleep.
- Typical duration <10 sec
- Broad-based transients, diffuse distributed, followed electrodecrement with superimposed low amplitude fast activity.

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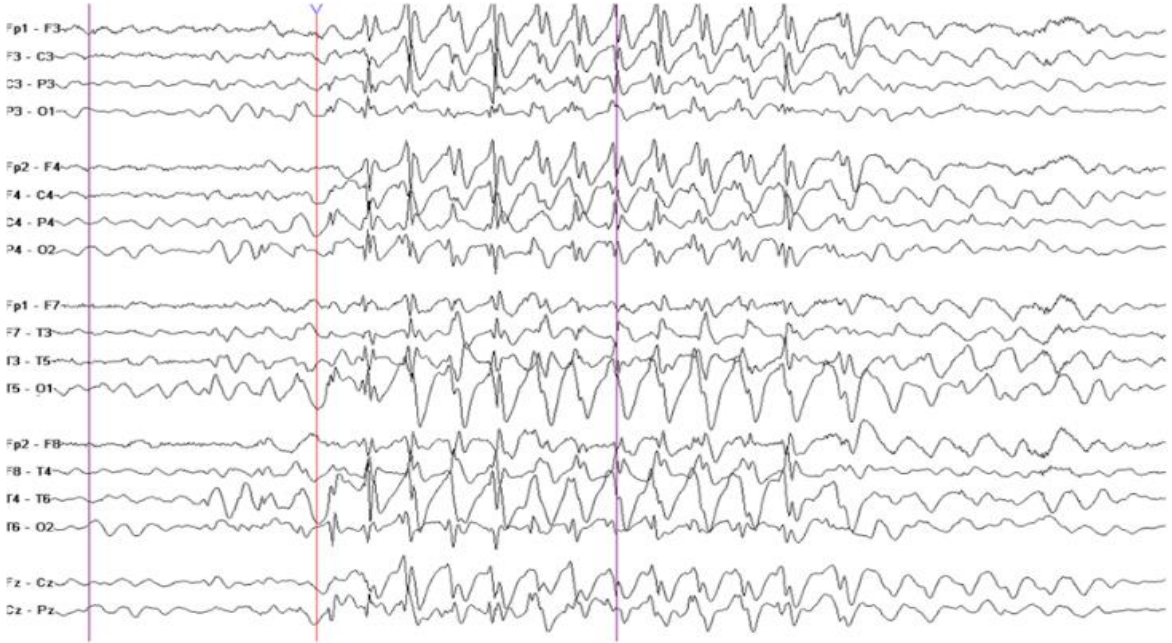
Atypical Absence Seizures



- Characterized by gradual onset and offset.
- Slower frequency (1.5 t0 2.5 Hz).
- Longer duration.
- Associated with epileptic encephalopathies.

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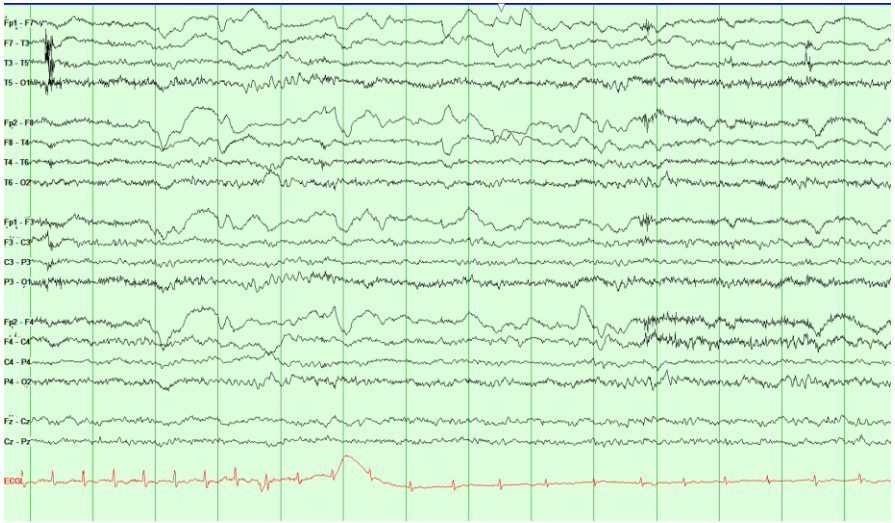
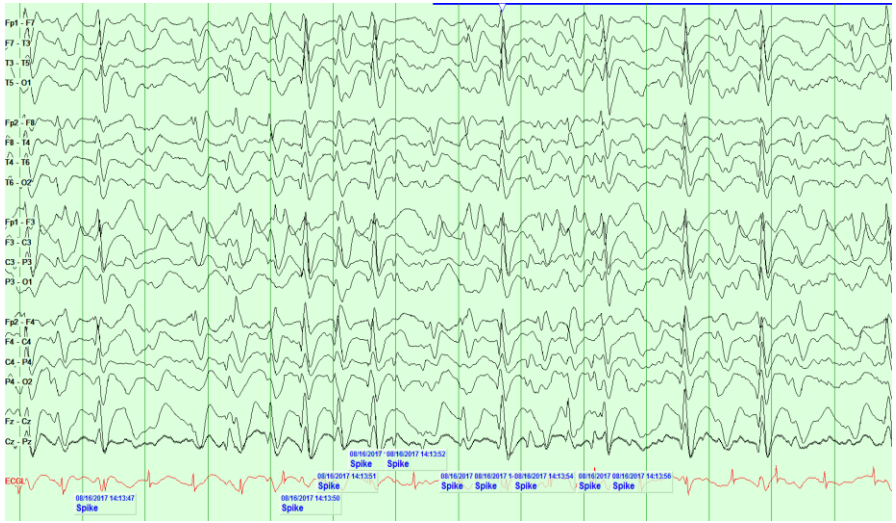
Electrical Status Epilepticus of Sleep (ESES)

- Clinical syndrome of epileptic encephalopathy with continuous spike-and-wave complexes during sleep.
- Epileptic encephalopathy consisting of the triad of cognitive decline, mixed generalized seizures and the EEG pattern of electrical status epilepticus of sleep (ESES).
- Traditional definition of ESES specified >85% of spike-and-wave complexes occupying slow-wave sleep.
- Heterogenous etiology (e.g., MCD, remote symptomatic, thalamic lesions).
- EEG can improve, but neurocognitive sequela persist.

Electrical Status Epilepticus of Sleep (ESES)



Awake



At the onset of NREM sleep

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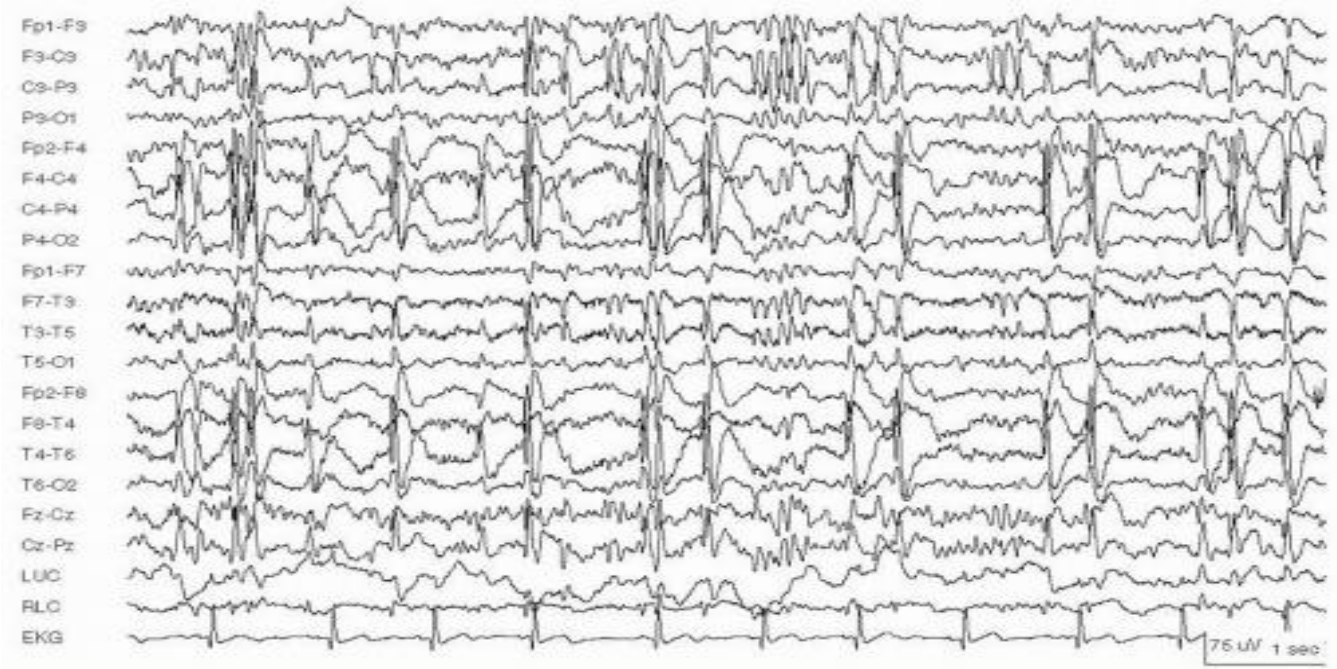
Acquired Epileptic Aphasia Landau-Kleffner Syndrome

- Age of onset between 2 to 14 years of age (typically 3-7 years)
- Characterized by loss of previously acquired language skills (acquired auditory agnosia).
- Clinical course can be rapid or insidious.
- Patient have multiple seizures types or no reported clinical seizures prior to diagnosis.
- Interictal EEG: background can be normal initially, followed by diffuse and focal slowing (temporal-parietal). IED (high-amplitude) observed over the temporal-parietal regions.
- Sleep EEG: characterized by ESES, but with a predilection for the perisylvian and posterior temporal regions.

Landau-Kleffner Syndrome



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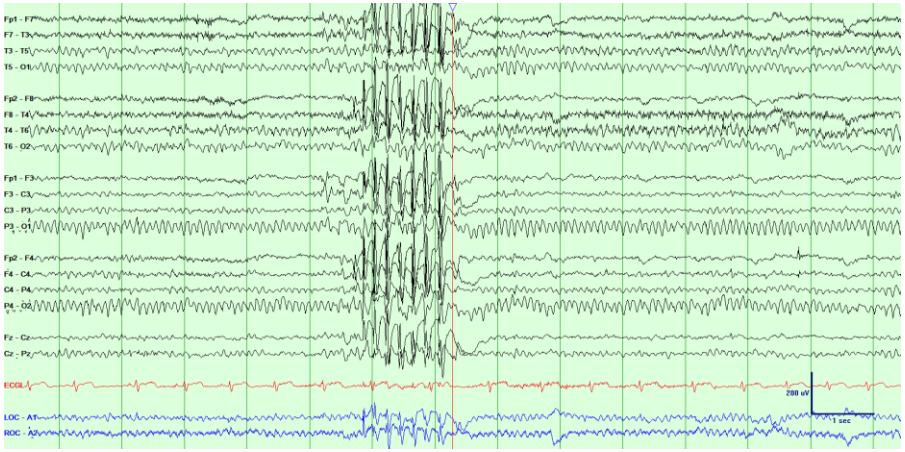
Epilepsy Syndromes – Age Groups

- Adolescent / Adult Onset Syndrome:
- Juvenile Absence Epilepsy (JAE)
- Juvenile Myoclonic Epilepsy (JME)
- Epilepsy with generalized tonic-clonic seizures alone.

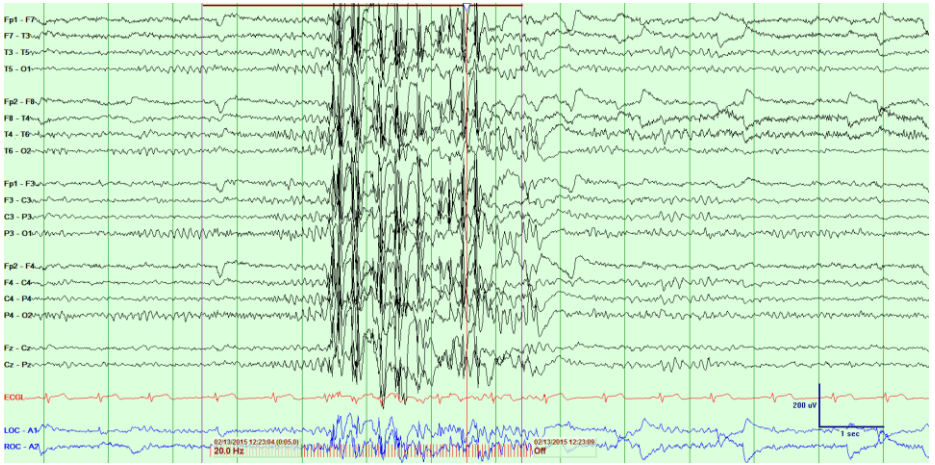
Juvenile Myoclonic Epilepsy (JME)

- Comprises 5-10% of all epilepsy syndromes.
- Onset between 12-18 years of age.
- Normal developmental, normal examination and normal neuro imaging.
- Predominant seizure type is myoclonic seizure (especially upon awakening), GTC seizure and ~1/3 experience absence seizures. Myoclonic-Tonic-Clonic seizure can occur.
- Interictal EEG: normal background/PDR for age. Generalized (“fast”) 4-6 Hz spike(polyspike)-and-wave discharges, accentuated in sleep (or with sleep deprivation). Fragmentation can occur (giving the false appearance of focal IEDs).
- Ictal EEG: myoclonic seizures are characterized by brief generalized paroxysms of 10-16 Hz poly-spikes.

Juvenile Myoclonic Epilepsy - JME



Generalized S/W



PPR

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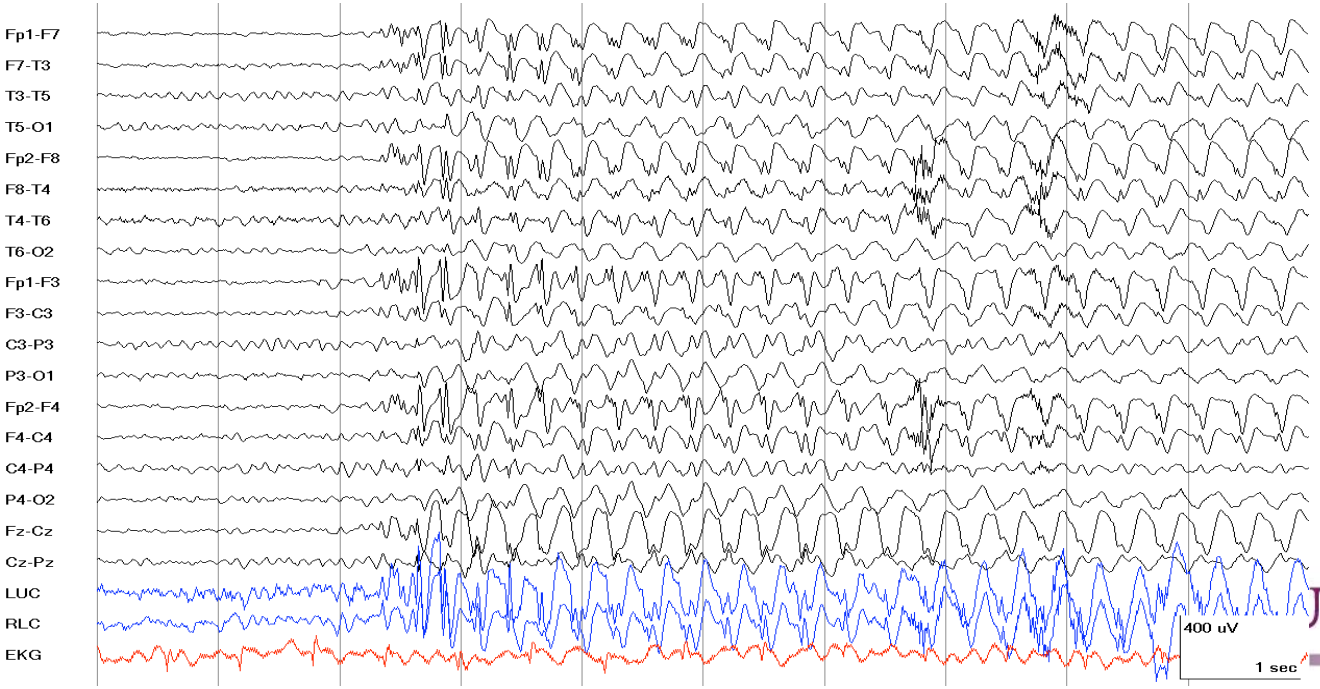
Juvenile Absence Epilepsy - JAE

- Age of onset 10-19 years of age (peak ~15 years)
- Seizures are less frequent, don’t cluster, but typically last longer and are associated more commonly with automatisms, speech arrest and loss of awareness.
- Normal exam, normal development and normal neuroimaging

Juvenile Absence Epilepsy - TAS



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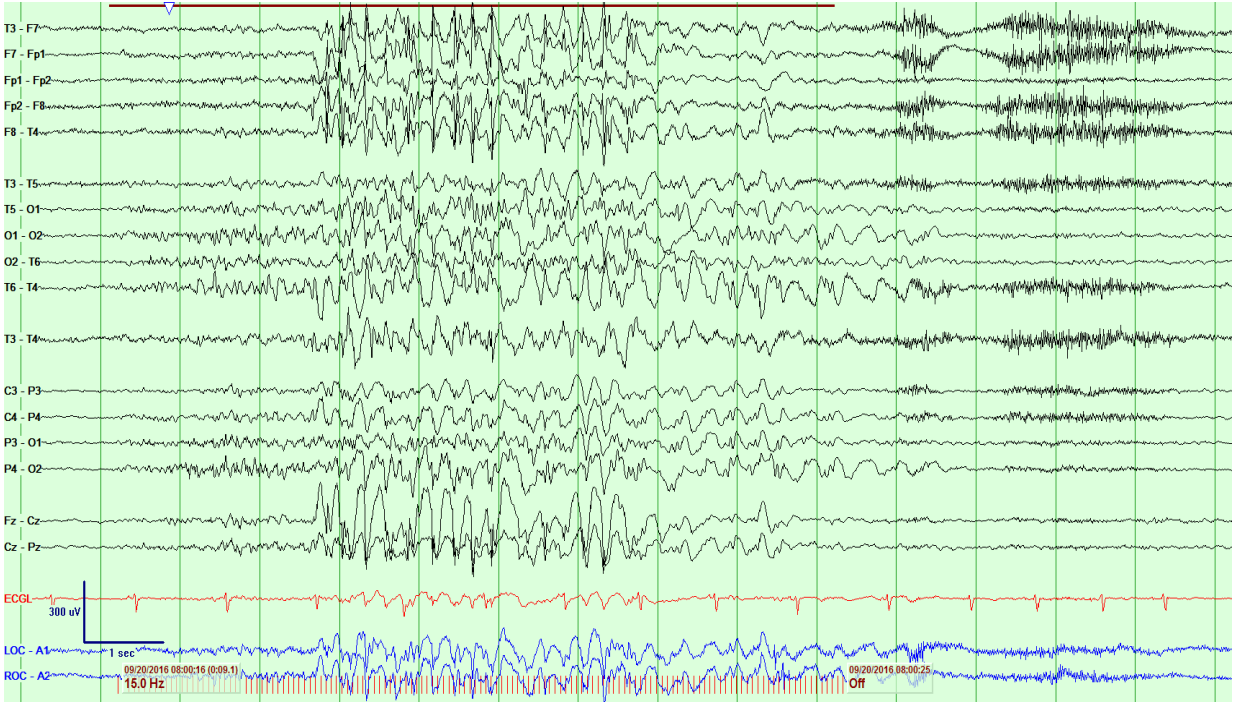
Common Findings Associated with GGE/IGE



Photoparoxysmal Response (PPR)

- Photoparoxysmal Response:
- Epileptiform discharge that occurs in response to a photic stimulus.
- Typical consist of generalized spike (polyspike)-and-wave discharges.
- Bilateral, symmetric, synchronous and typically generated between 15-18 Hz frequencies.
- It may occur independently of seizures and exist as an inherited trait.

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