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2020 Epilepsy Board Review Course

EPILEPSIES AND ELECTROCLINICAL SYNDROMES IN CHILDHOOD

Doose Syndrome

- Onset in early childhood
- Genetic with positive family history – 40%
- Multiple generalized seizure types, especially myoclonic atonic seizures with falling
- Generalized spike wave complexes and polyspikes on EEG

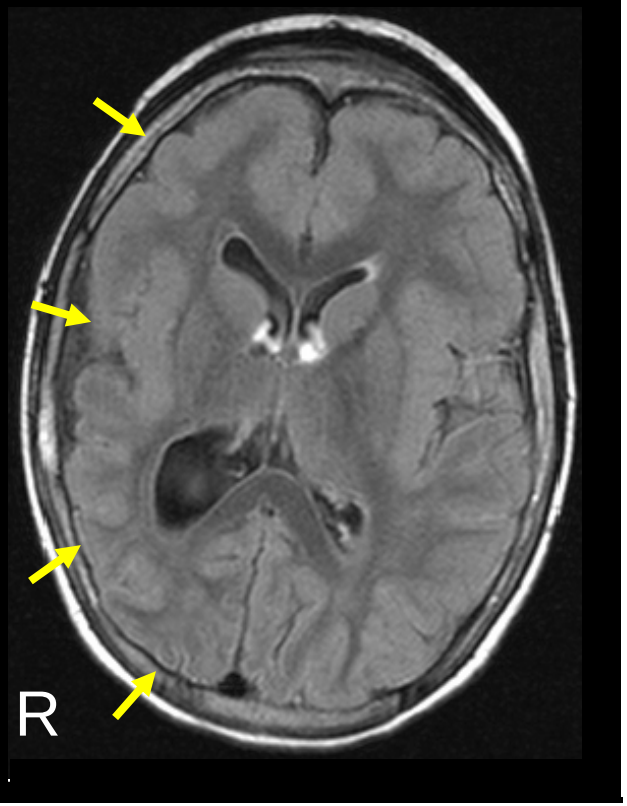
Doose Syndrome

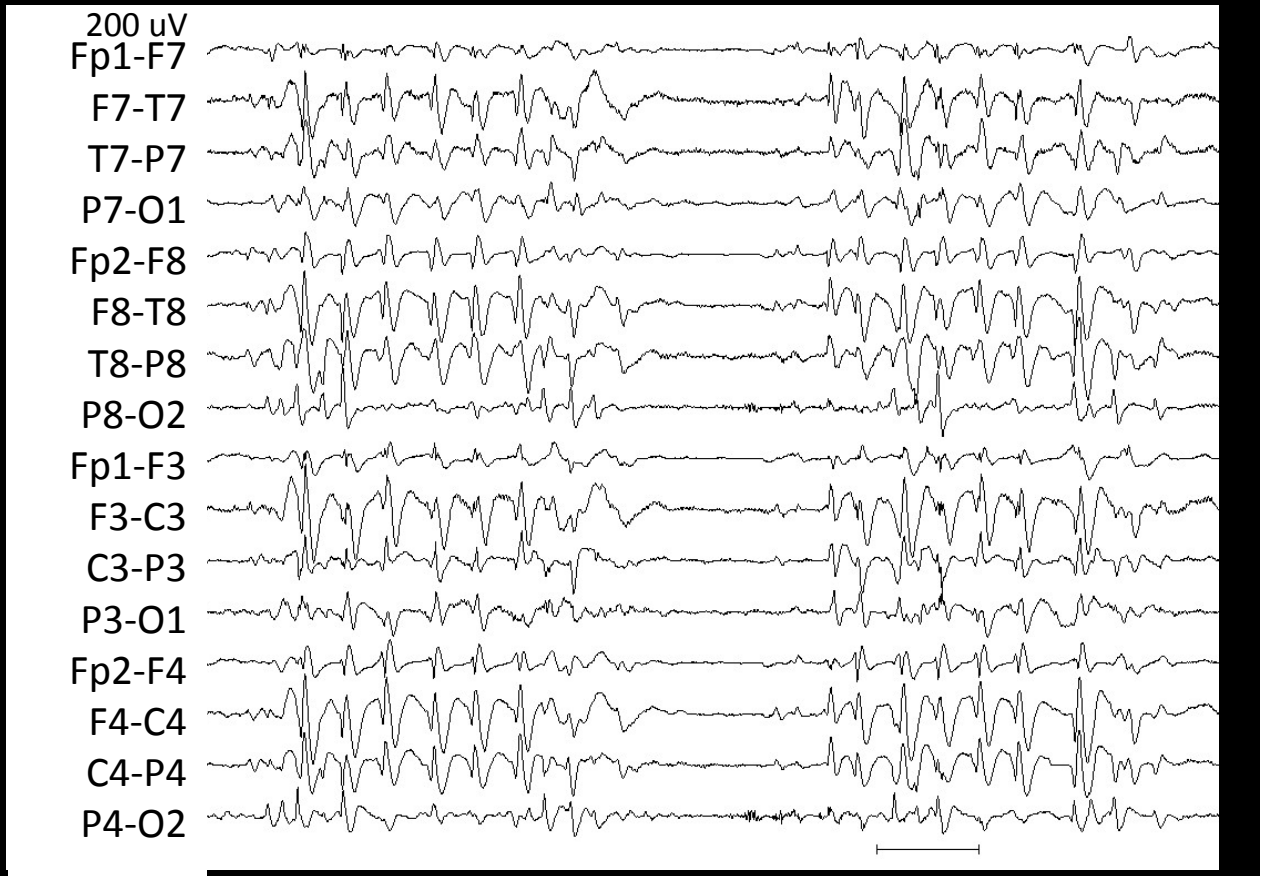
- Valproate, lamotrigine, ethosuximide, KG diet
- Avoid oxcarbazepine, phenytoin, vigabatrin
- Seizures controlled in > 50% of patients within 3 years, with good cognitive outcome

Lennox-Gastaut Syndrome

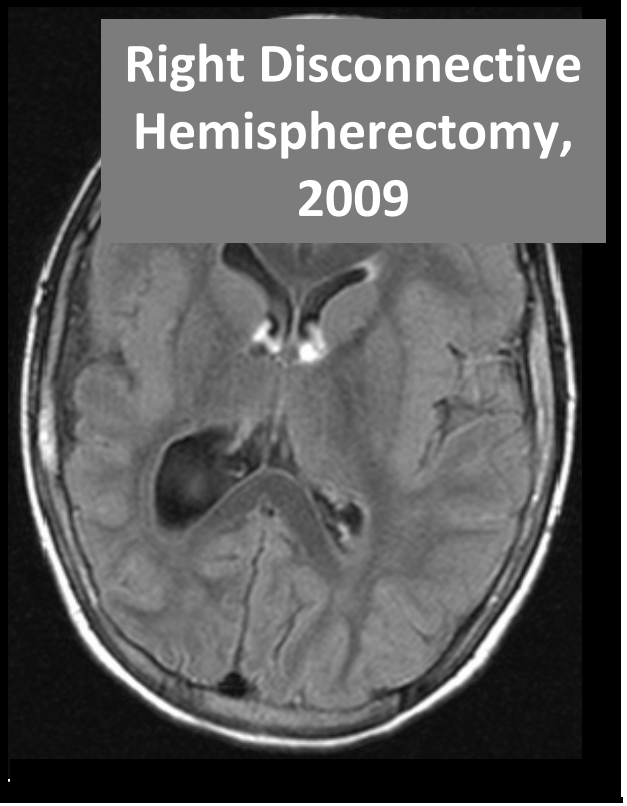
- Electroclinical features emerge at 2 to 6 years
- Multiple seizure types, high seizure frequency, episodes of status epilepticus
- Progressive intellectual deterioration
- Nonspecific response to various brain insults and conditions - infarction, hypoxia ischemia, infection, malformation, trauma, genetic disorders

Remain Aware of Surgical Options

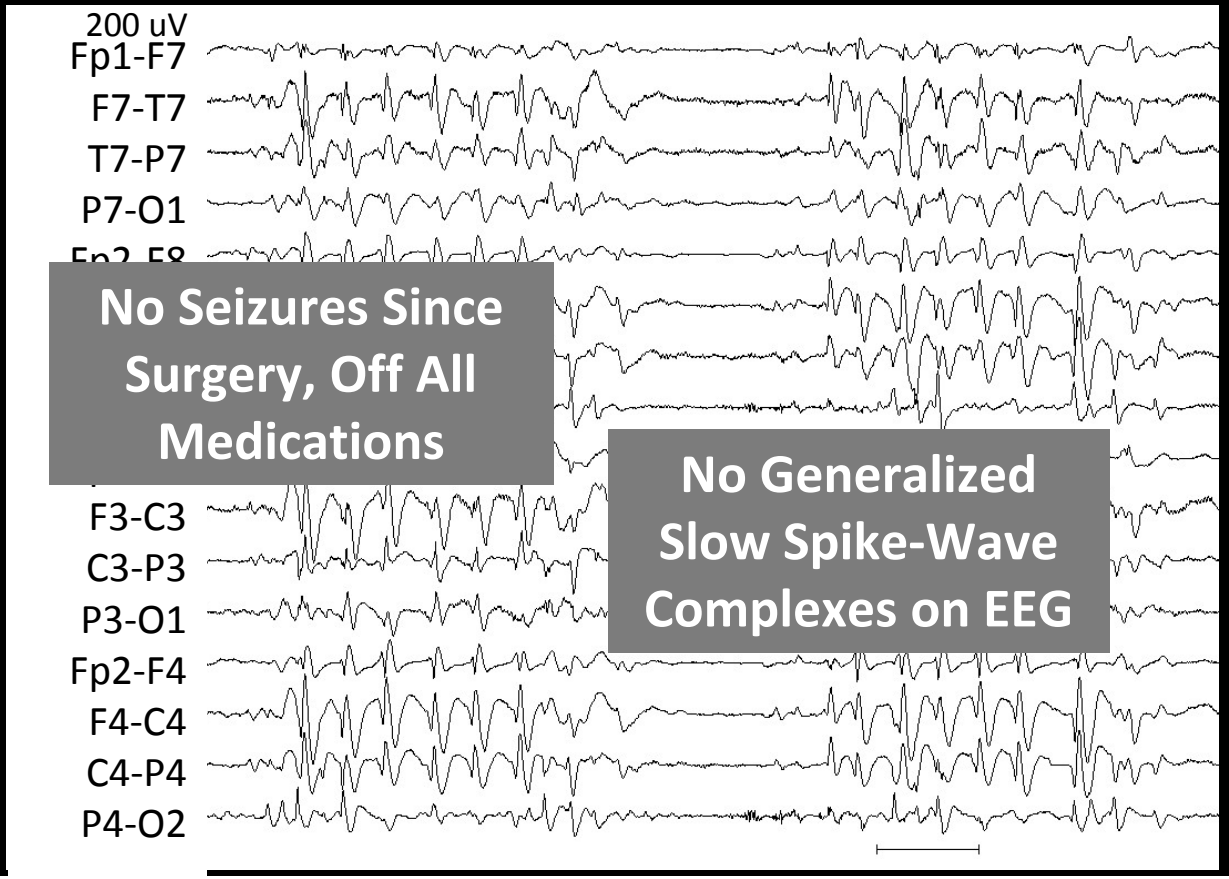




Remain Aware of Surgical Options



Right Disconnective Hemispherectomy, 2009



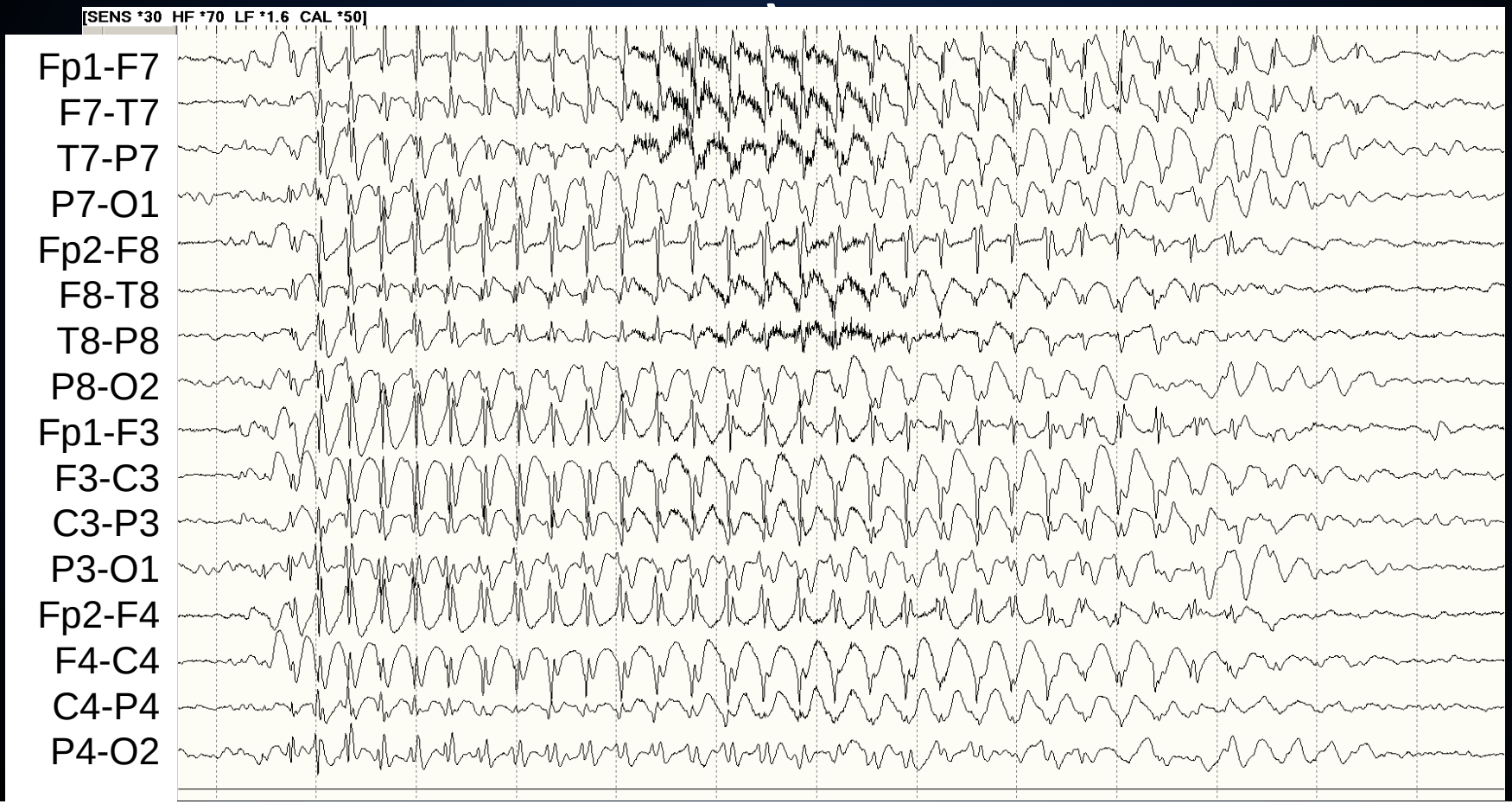
No Seizures Since Surgery, Off All Medications

No Generalized Slow Spike-Wave Complexes on EEG

Childhood Absence Epilepsy

- Peak onset 4 to 8 years
- About 1/3rd also have generalized tonic clonic seizures
- Remission by 10 to 12 years in > 50% of children

During Hyperventilation



Childhood Absence Epilepsy

Favorable Options

- Ethosuxmide (if no GTCs)
- Valproate
- Lamotrigine
- Levetiracetam
- Zonisamide

To Be Avoided

- Carbamazepine
- Oxcarbazepine
- Phenytoin
- Tiagabine
- Vigabatrin

Febrile Seizures

- Age 3 months to 5 years
- Source of fever is outside the CNS
- Recurrence in 1/3rd of children
- Provide rescue medication for prolonged seizures

Febrile Seizures

- Risk for subsequent epilepsy is increased after febrile status epilepticus
- May cause hippocampal sclerosis or be due to pre-existing hippocampal abnormality

Genetic Epilepsy with Febrile Seizures Plus

- Genetically and phenotypically heterogeneous
- Usually autosomal dominant with low penetrance
- Most frequent mutations - sodium channel and GABAa receptor subunit genes – SCN1B, SCN1A, STX1B, SCN9A, GABRG2, GABRD, CACNA1H, HCN2

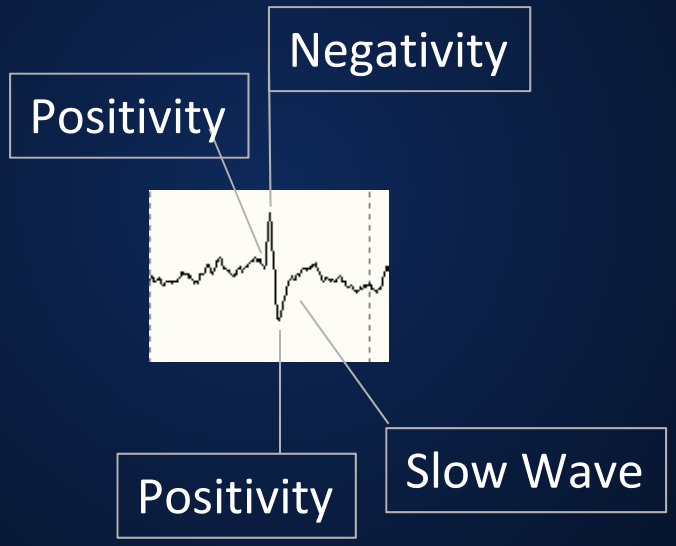
GEFS + Phenotypic Variability

- Simple febrile seizures, 3 mo to 6 yrs old – 44%
- Afebrile seizures or seizures after 6 yr – 27%
- Generalized or focal epilepsy with or without febrile seizures, Doose syndrome, Dravet syndrome – less common

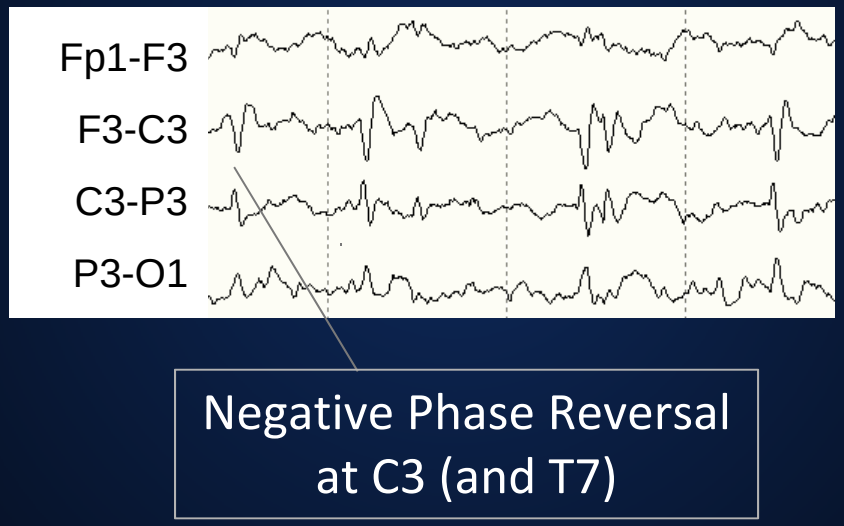
Benign Rolandic Epilepsy

SELF-LIMITED CHILDHOOD EPILEPSY WITH CENTROTEMPORAL SPIKES

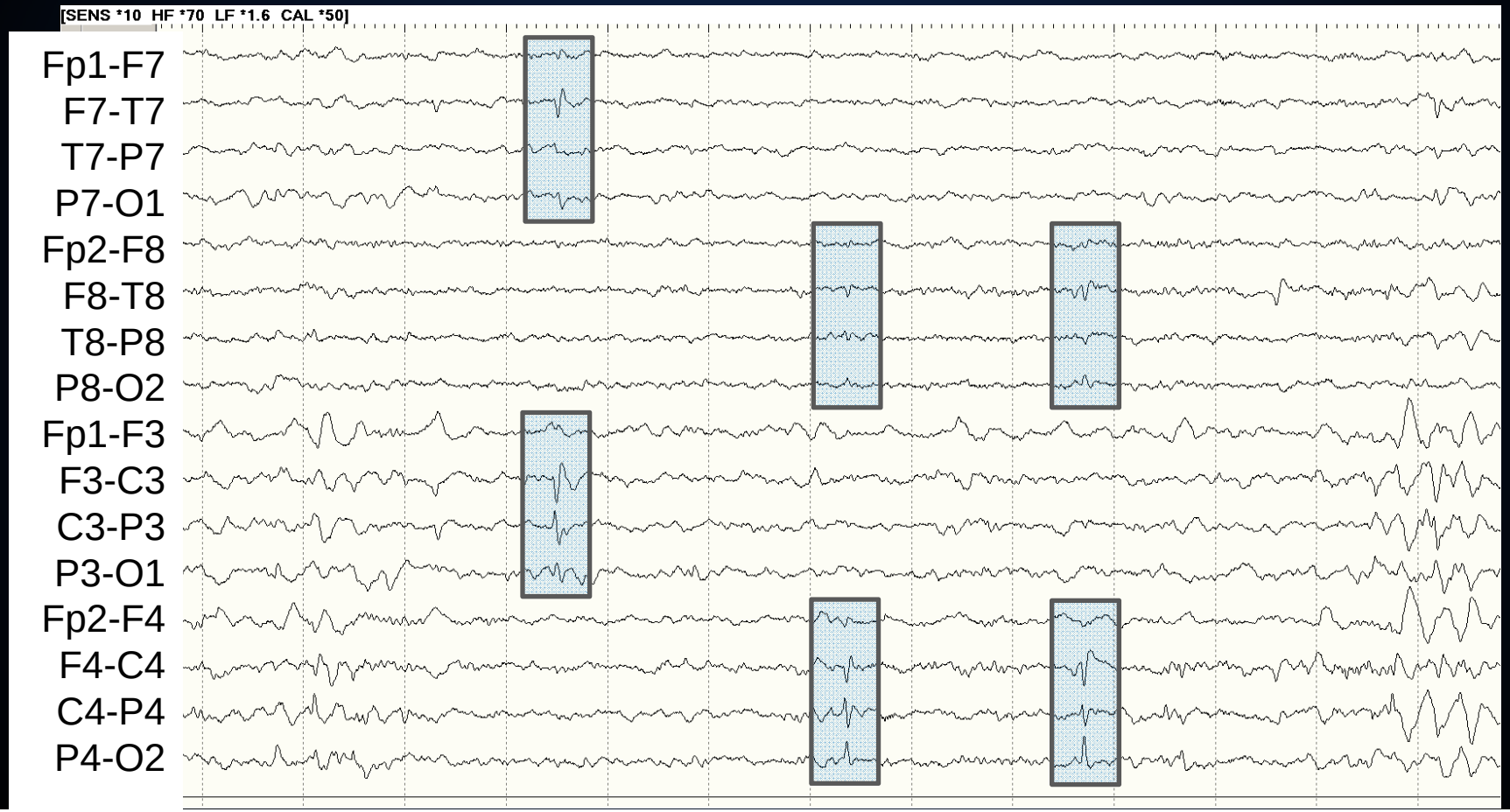
EEG Features - Stereotyped Morphology



Stereotyped Distribution



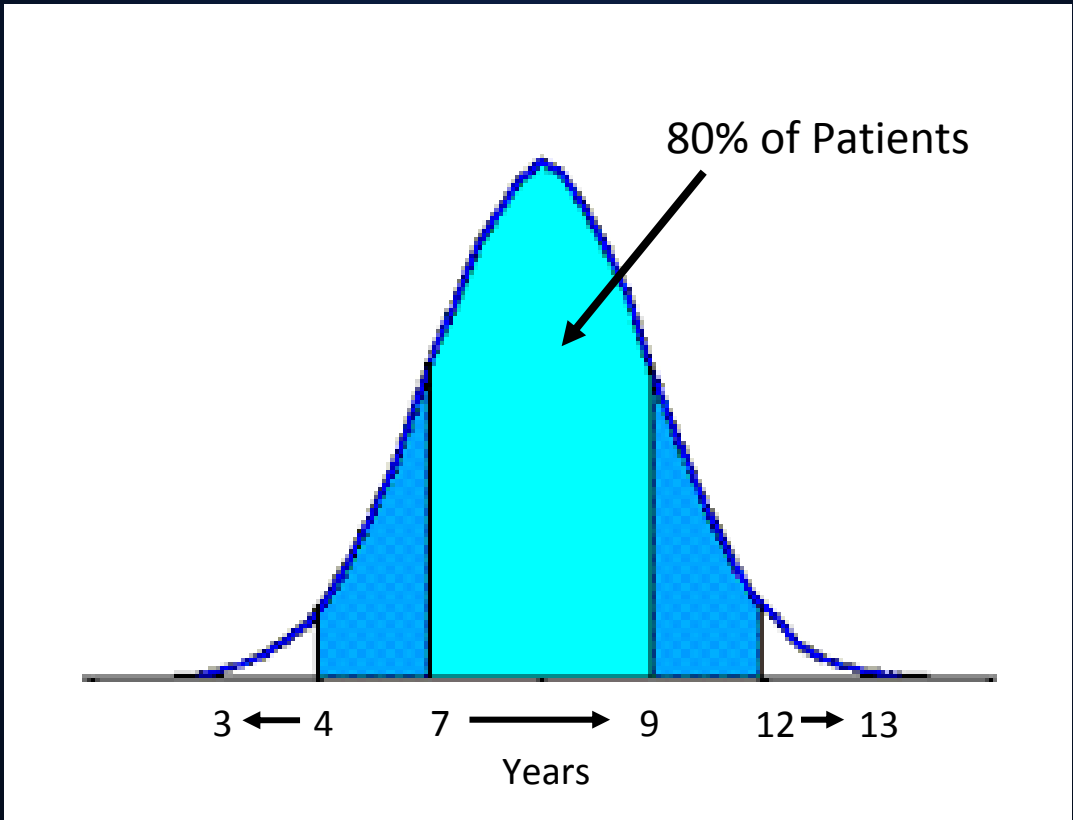
Sleep Activation



Correlate Carefully with Episode Type

MAY BE AN INCIDENTAL FINDING

Age at Seizure Onset – Tight Peak



Seizure Type at Presentation

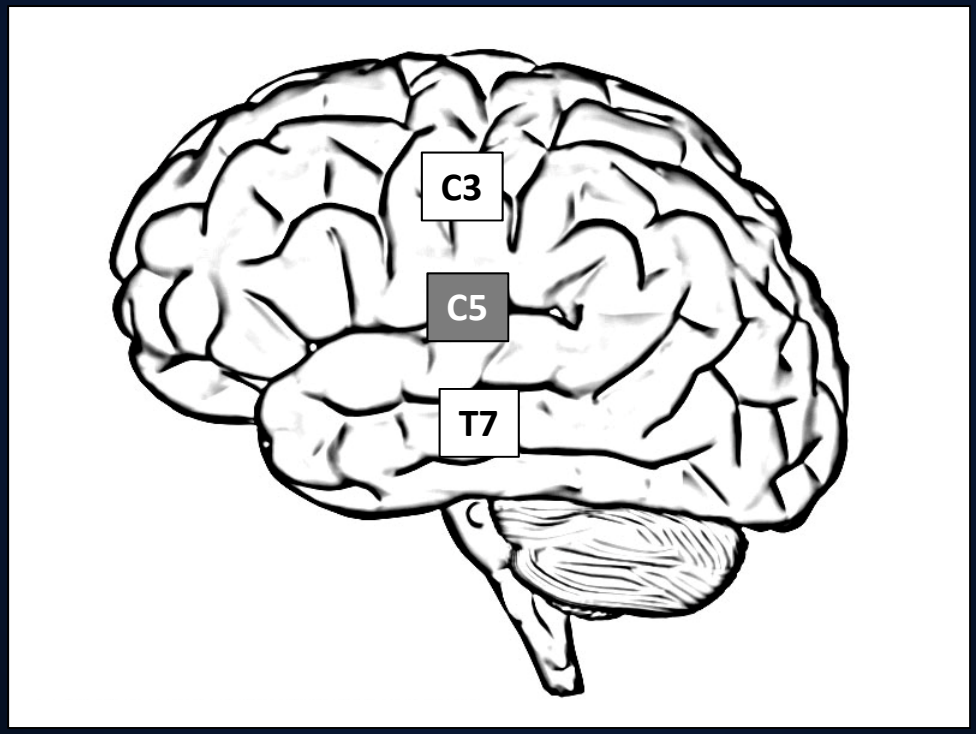
Most Common
80% of Patients

Generalized
Tonic Clonic
in Sleep

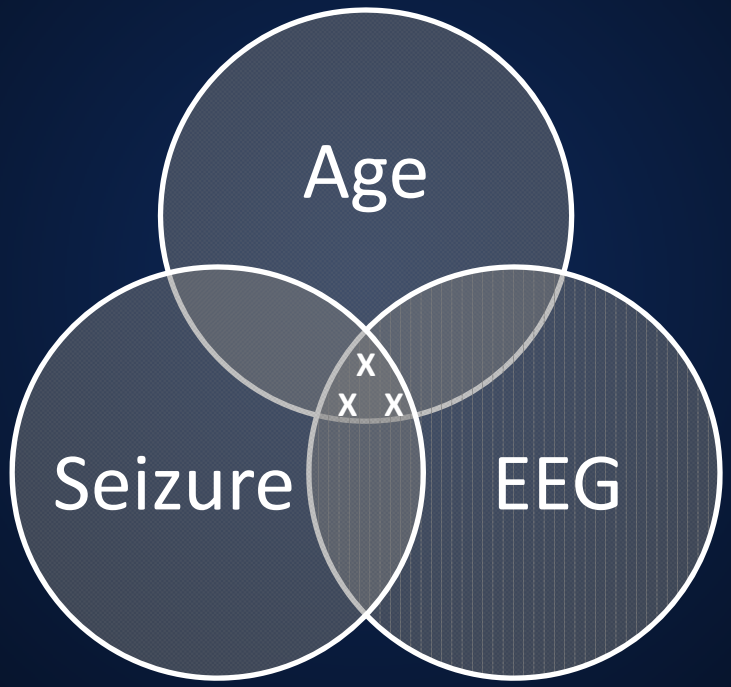
2nd Most Common

Facial
Sensorimotor

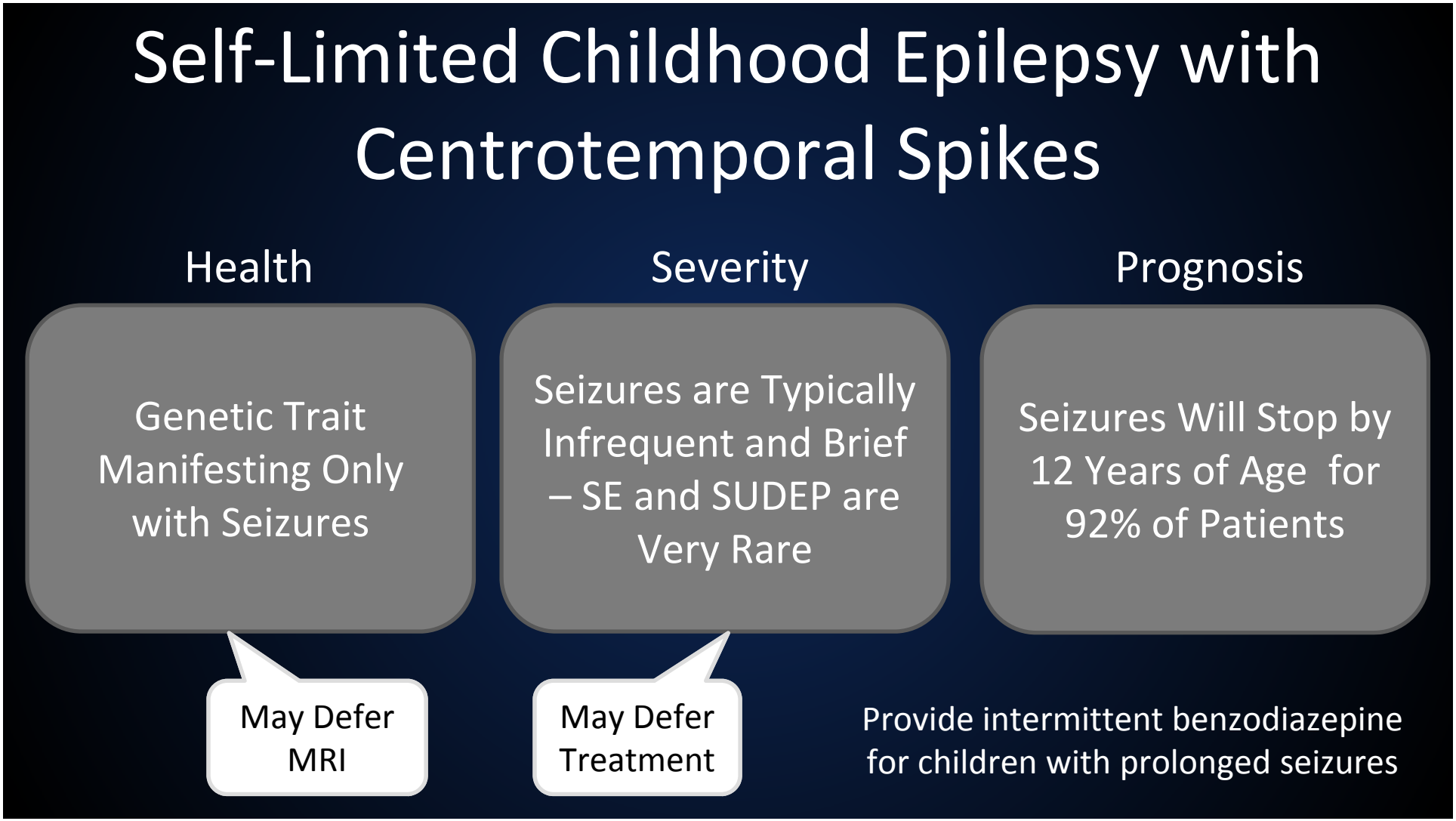
Perisylvian / Peri-rolandic



Typical Features - Clinical Confidence



Self-Limited Childhood Epilepsy with Centrotemporal Spikes



Consider Treating…

- If seizures are more frequent
- If seizures interfere with QOL
  - School absences for morning postictal tiredness
  - Frightening symptoms such as pharyngeal contraction with intact awareness

C-T Spikes and Generalized Epilepsy

- Focal and generalized “idiopathic” phenotypes may co-exist
- Either or both may be clinically active
- Most often, the generalized seizures are the bigger problem
- Tailor the drug choice accordingly

C-T Spikes and Symptomatic Epilepsy

- C-T spikes may be a “distractor” in surgical cases
  - Hippocampal sclerosis
  - Malformation of cortical development
  - Low-grade tumor
- If video EEG also shows findings concordant with MRI…
- Do not let the C-T spikes derail the surgical plan

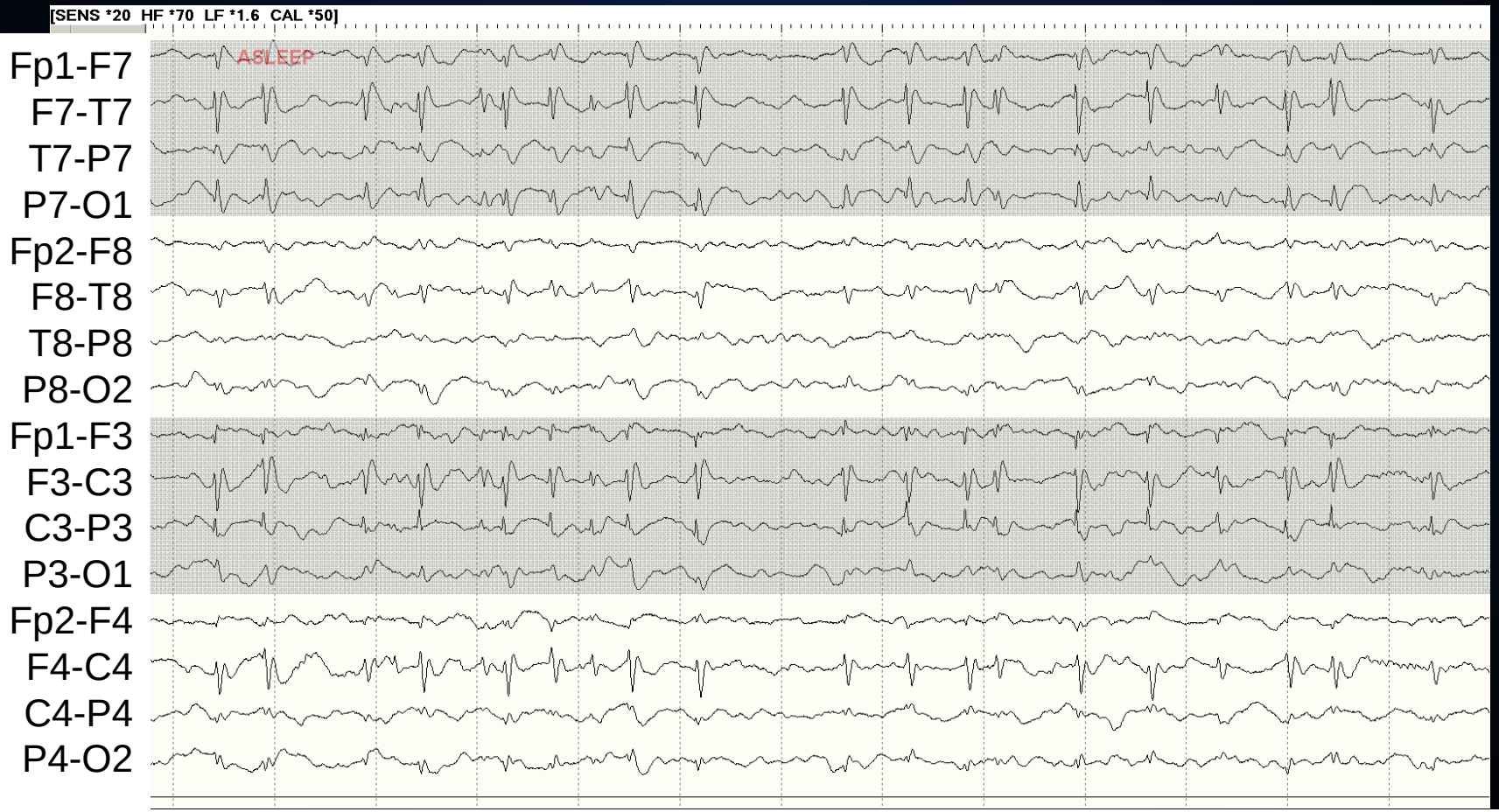
Self-Limited Childhood Epilepsy with Centrotemporal Spikes

“EXCESSIVE SPIKING” SYNDROMES

Acquired Epileptic Aphasia (Landau-Kleffner Syndrome)

- Onset 3 to 8 years
- Auditory verbal agnosia with language regression
- Behavioral and cognitive decline
- Continuous left centrotemporal sharp waves

Acquired Epileptic Aphasia (LKS)



Continuous Spike and Waves in Sleep (CSWS or ESES)

- Global epileptic encephalopathy with cognitive and behavioral regression
- Continuous bilateral centrotemporal sharp waves in sleep
- Spike wave index > 60% to 80%

Treatment – LKS and CSWS

Least Effective	More Effective	Most Effective
Valproate, Ethosuximide, Levetiracetam, Lamotrigine, Sulthiame	Nighttime Benzodiazepine, 0.3 to 0.6 mg/kg	Corticosteroids (Adverse Effects are Limiting)

C-T Spikes and Myoclonia

- Facial twitches, positive or negative limb or axial myoclonia
- Due to frequent interictal spiking
- Not to be confused with EPC of severe symptomatic epilepsies
  - Rasmussen syndrome
  - MRI-occult cortical dysplasia

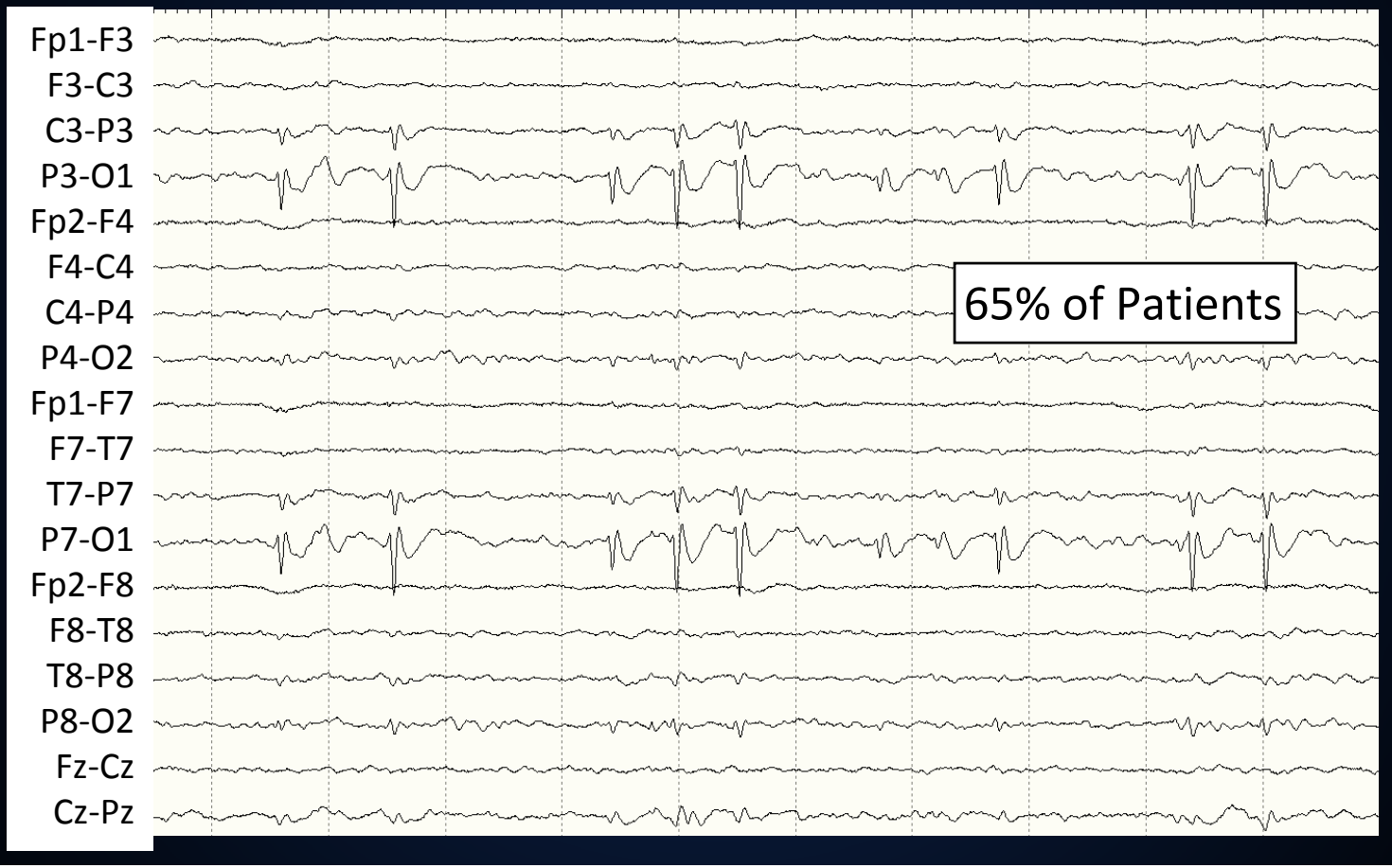
Panayiotopoulos Syndrome

SELF-LIMITED CHILDHOOD OCCIPITAL EPILEPSY WITH AUTONOMIC SEIZURES

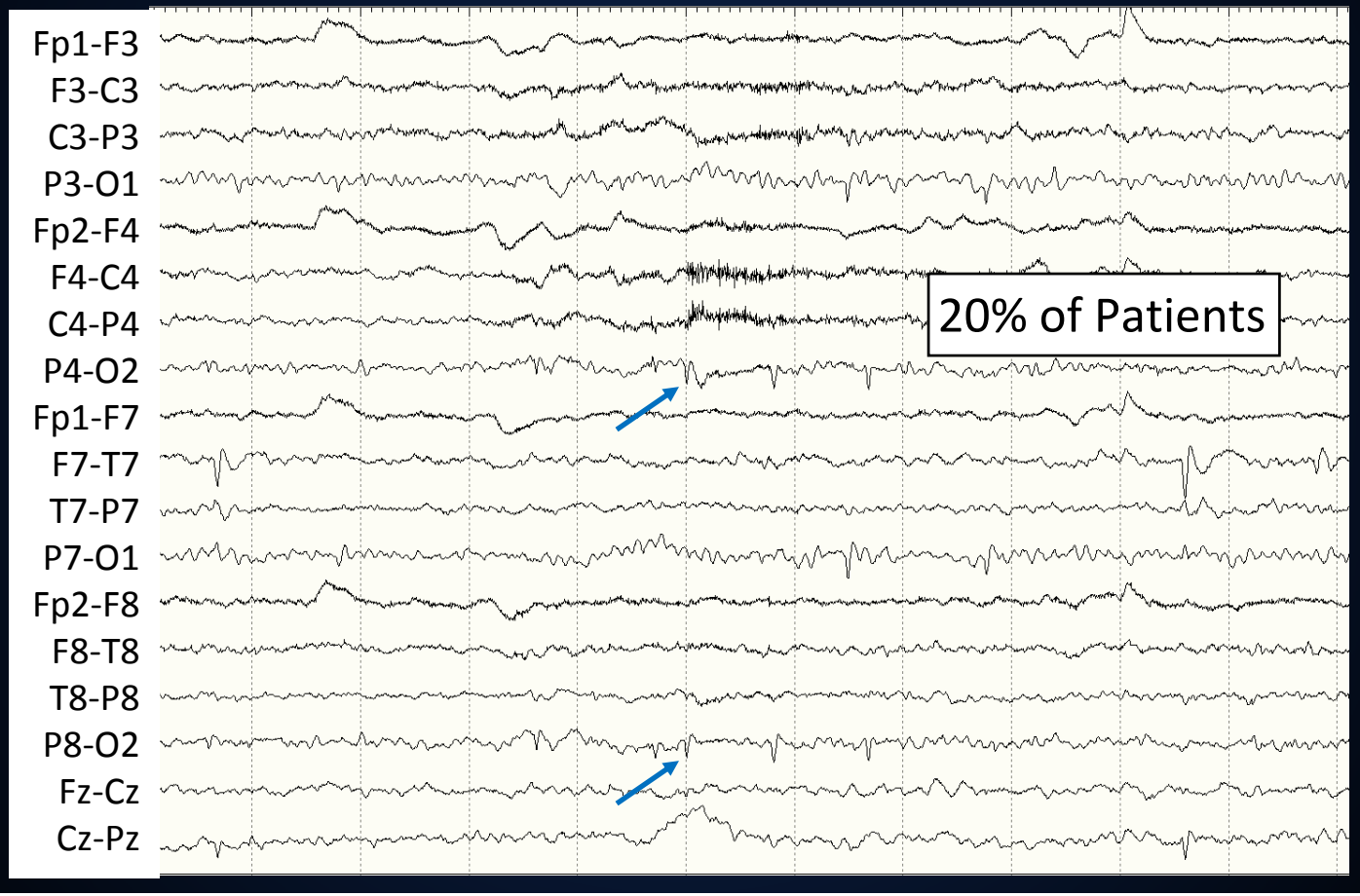
Panayiotopoulos Syndrome

- Healthy children, peak age 4 to 5 years
- Prolonged seizures, usually in sleep
- Altered awareness, limp unresponsiveness, autonomic features (especially vomiting)
- Infrequent seizures, spontaneous remission by 12 years – may defer treatment

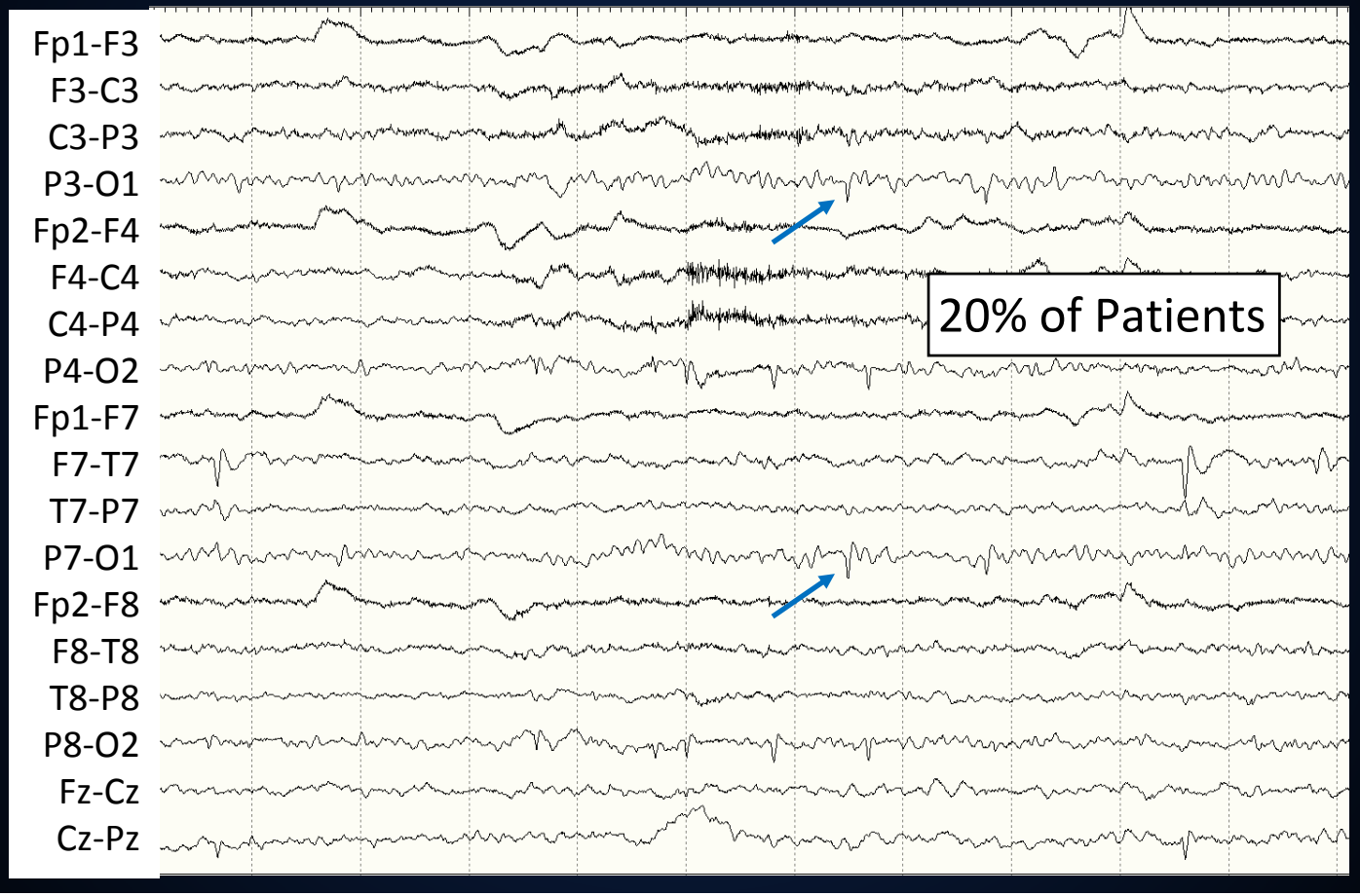
Occipital Sharp Waves



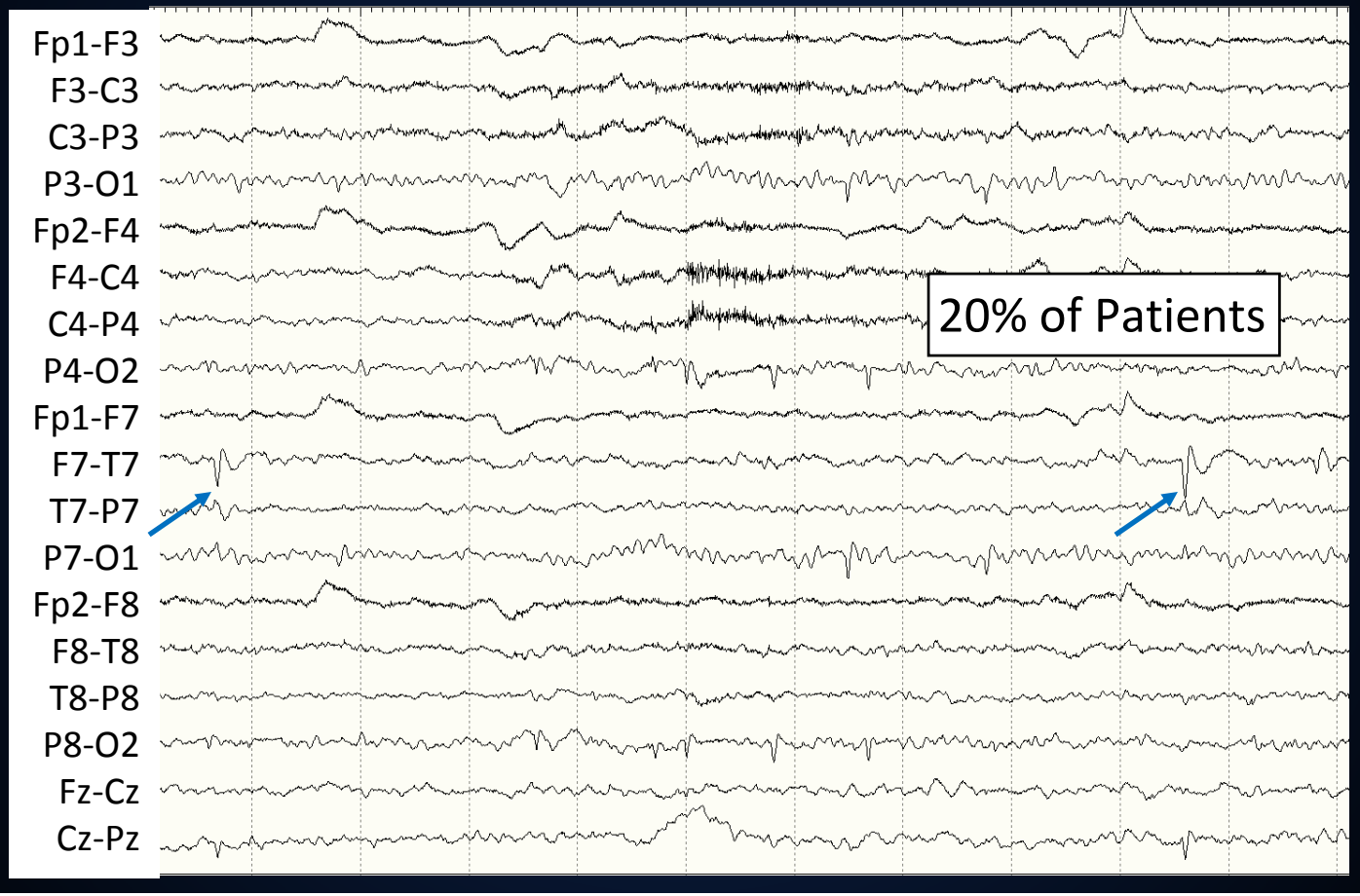
Occipital and Rolandic Sharp Waves



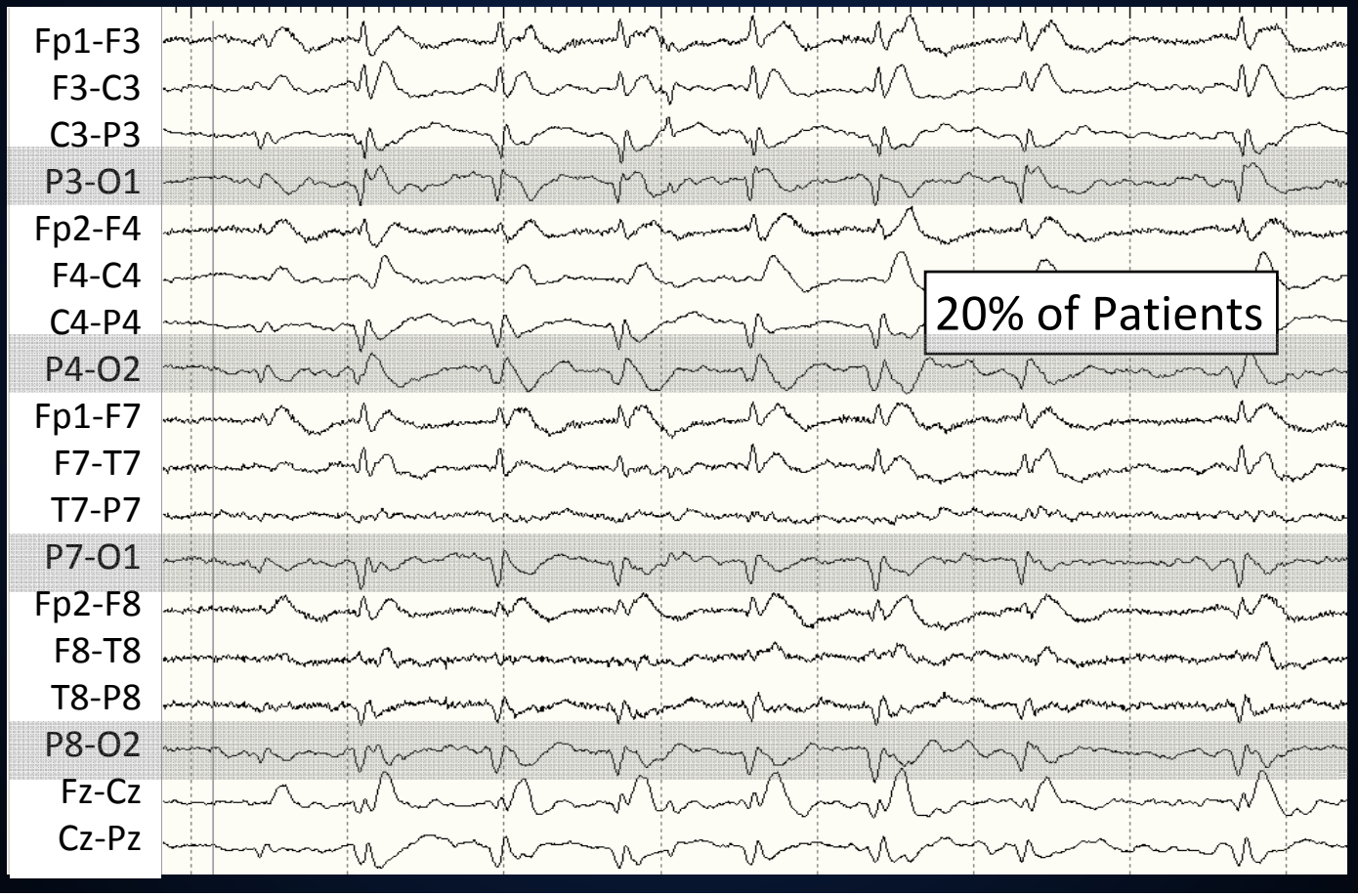
Occipital and Rolandic Sharp Waves

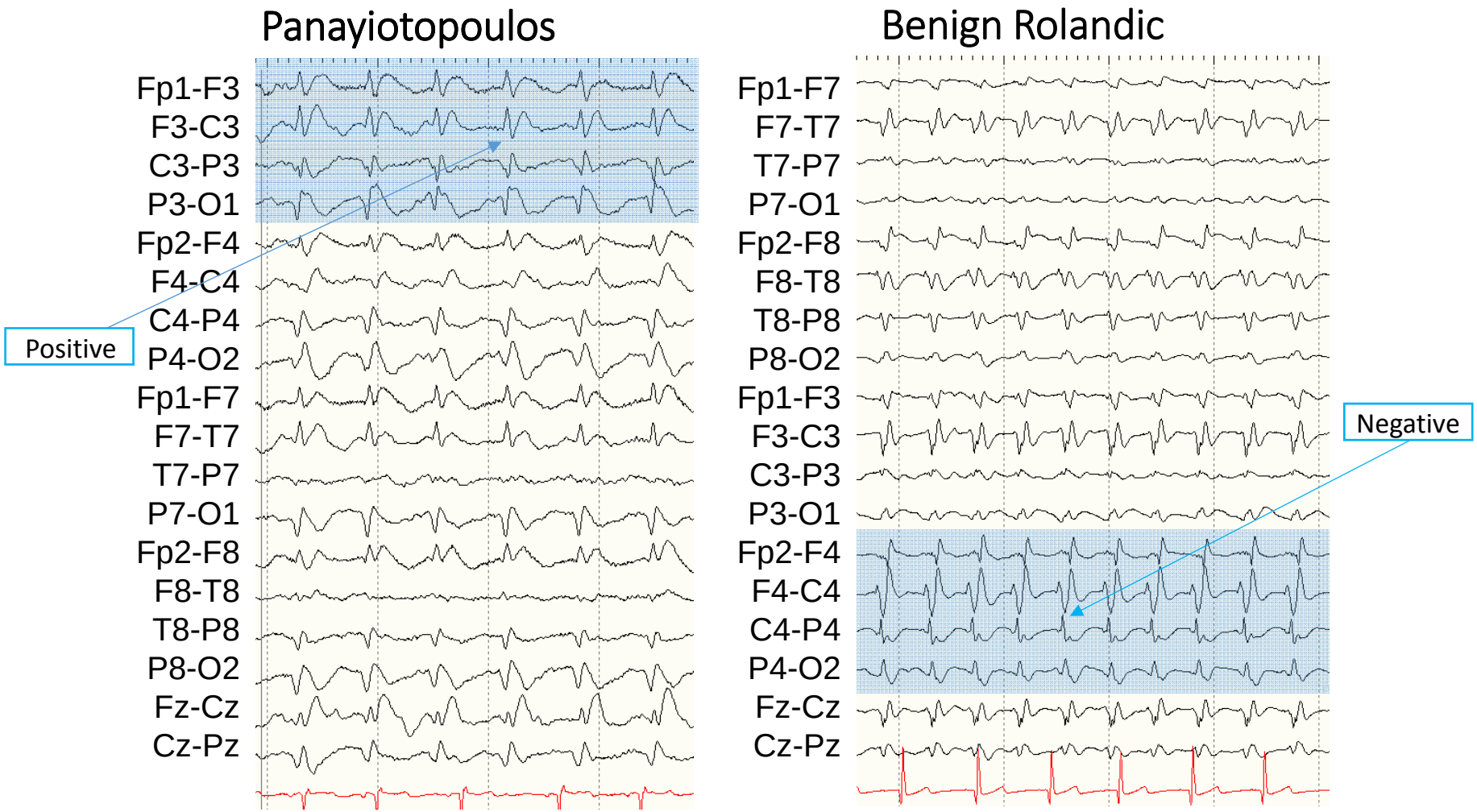


Occipital and Rolandic Sharp Waves



Bilateral Occipito-Frontal Sharp Waves

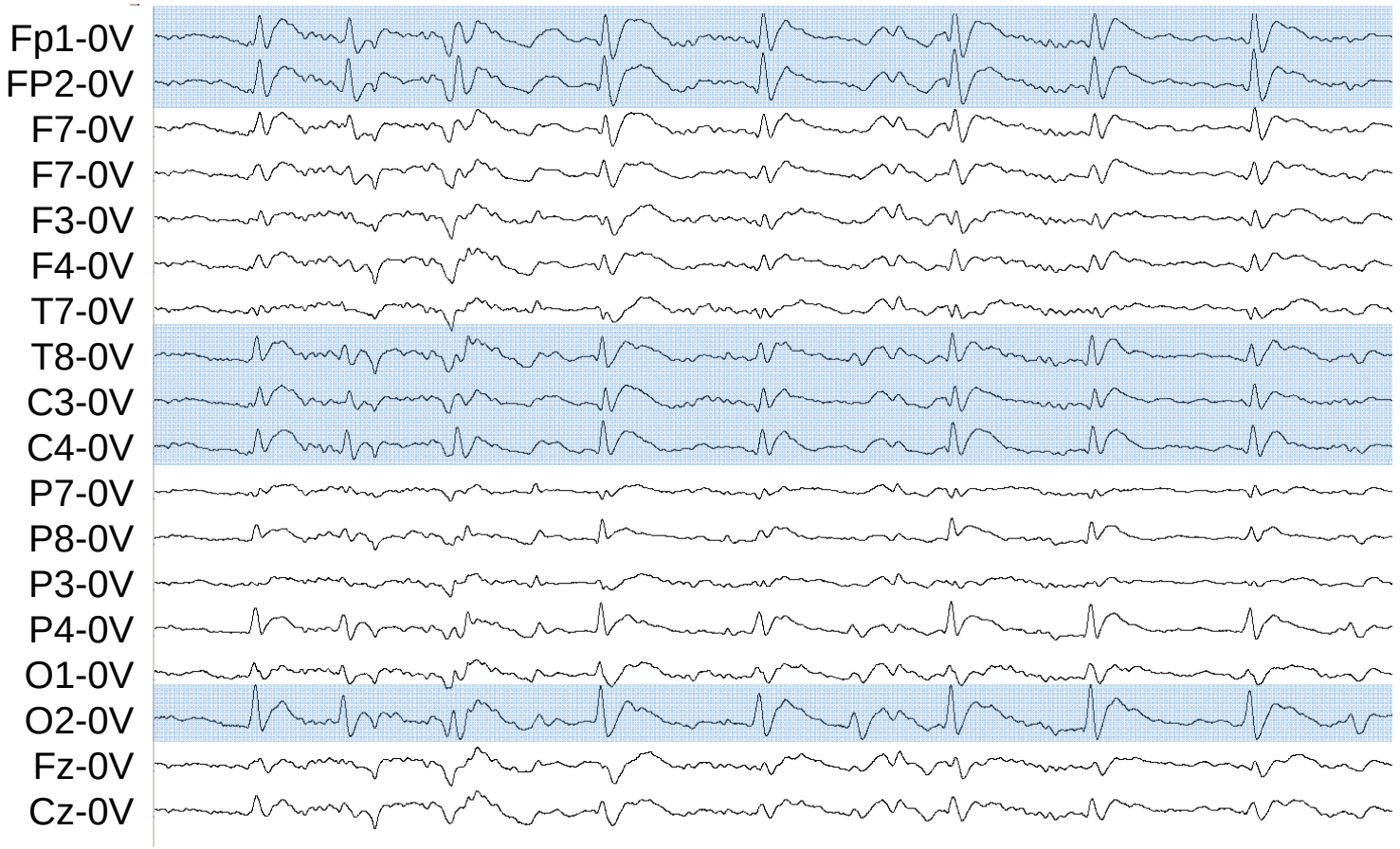


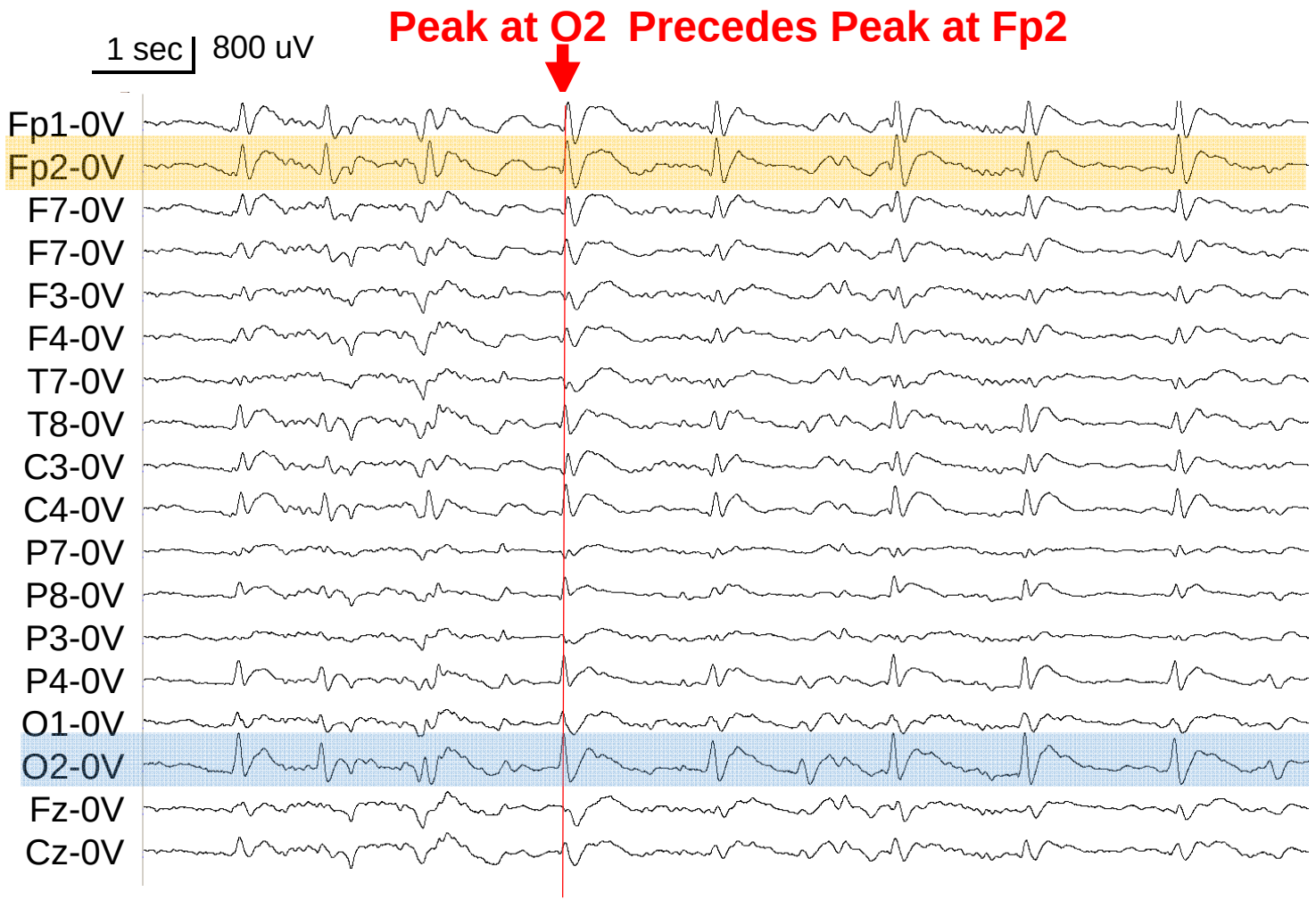

Panayiotopoulos
Fp1-F3
F3-C3
C3-P3
P3-O1
Fp2-F4
F4-C4
C4-P4
P4-O2
Fp1-F7
F7-T7
T7-P7
P7-O1
Fp2-F8
F8-T8
T8-P8
P8-O2
Fz-Cz
Cz-Pz
Positive
Benign Rolandic
Fp1-F7
F7-T7
T7-P7
P7-O1
Fp2-F8
F8-T8
T8-P8
P8-O2
Fp1-F3
F3-C3
C3-P3
P3-O1
Fp2-F4
F4-C4
C4-P4
P4-O2
Fz-Cz
Cz-Pz
Negative

Refential Montage

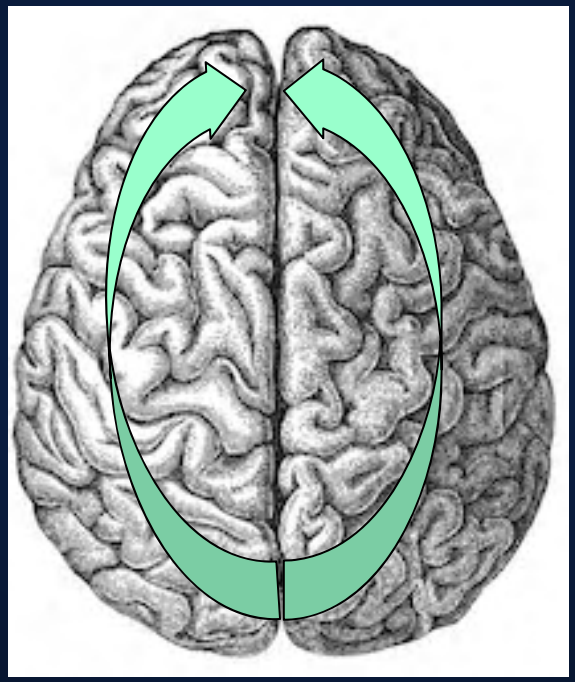
1 sec | 800 uV

Frontal & Occipital > Centrotemporal




Peak at O2 Precedes Peak at Fp2
1 sec
800 uV
Fp1-0V
Fp2-0V
F7-0V
F7-0V
F3-0V
F4-0V
T7-0V
T8-0V
C3-0V
C4-0V
P7-0V
P8-0V
P3-0V
P4-0V
O1-0V
O2-0V
Fz-0V
Cz-0V

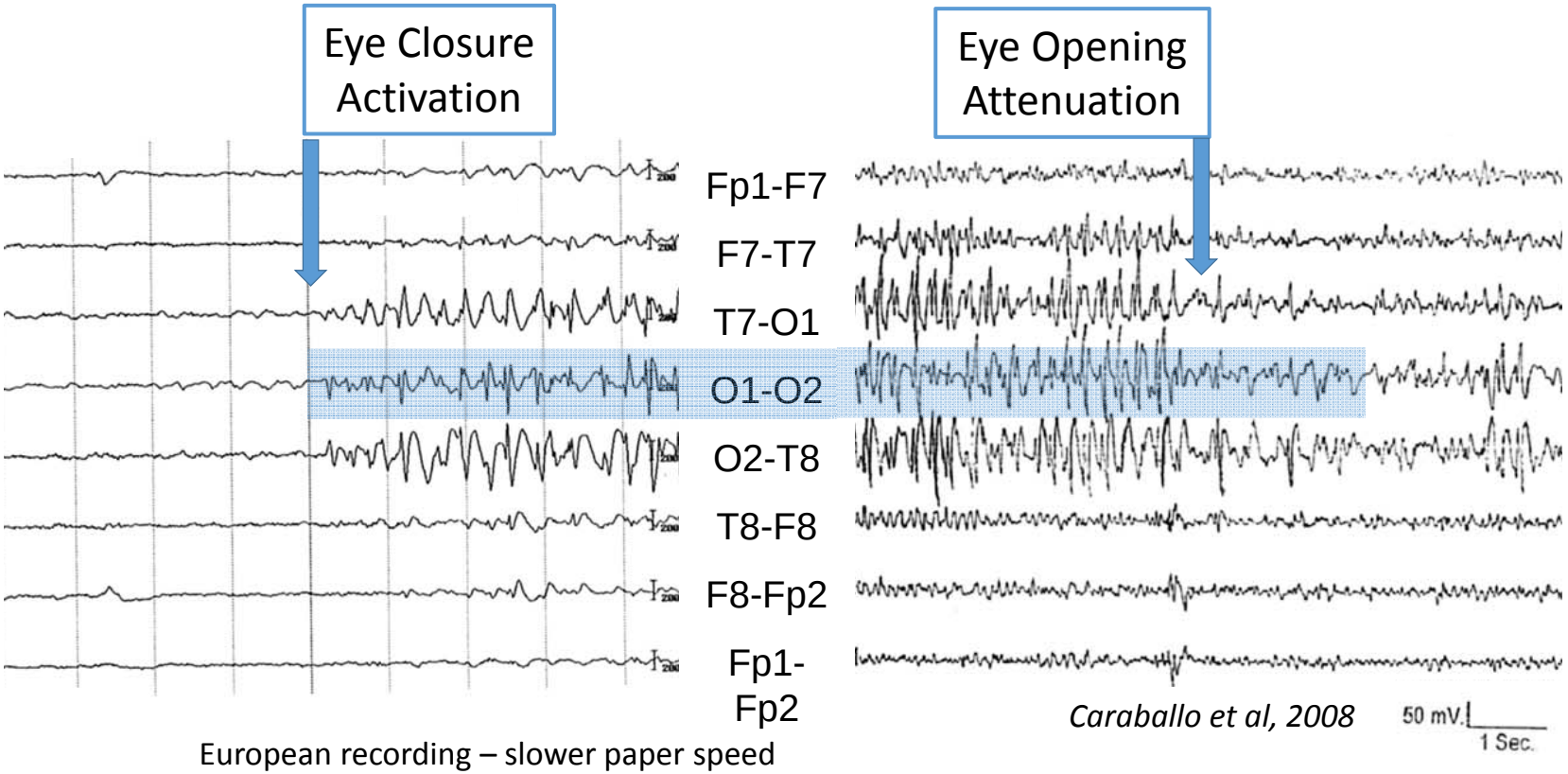
Bilateral Mesial Occipital – Frontal Projection



Late Onset Benign Occipital Epilepsy (Gastaut Type)

- Peak age at onset – 8 to 9 years
- Visual auras – short and frequent
- May involve tonic eye deviation, eyelid closure, or evolution to GTC

Occipital Spikes with “Fixation-Off Sensitivity”



Caraballo et al, 2008

European recording – slower paper speed

Gastaut Phenotype - Various Etiologies

- Presumed genetic – normal MRI, healthy child
- May be symptomatic – occipital pathologies
  - Cortical dysplasia
  - Mitochondrial disease
  - Lafora disease
  - Celiac disease

Prognosis - Gastaut's Series – Presumed Genetic Cases

- Most cases require treatment for frequent seizures
- 60% of patients achieve complete seizure control
- 95% have spontaneous remission before adulthood

Good Luck on Your Board Examination!

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