# Epilepsy: diagnosis and treatment

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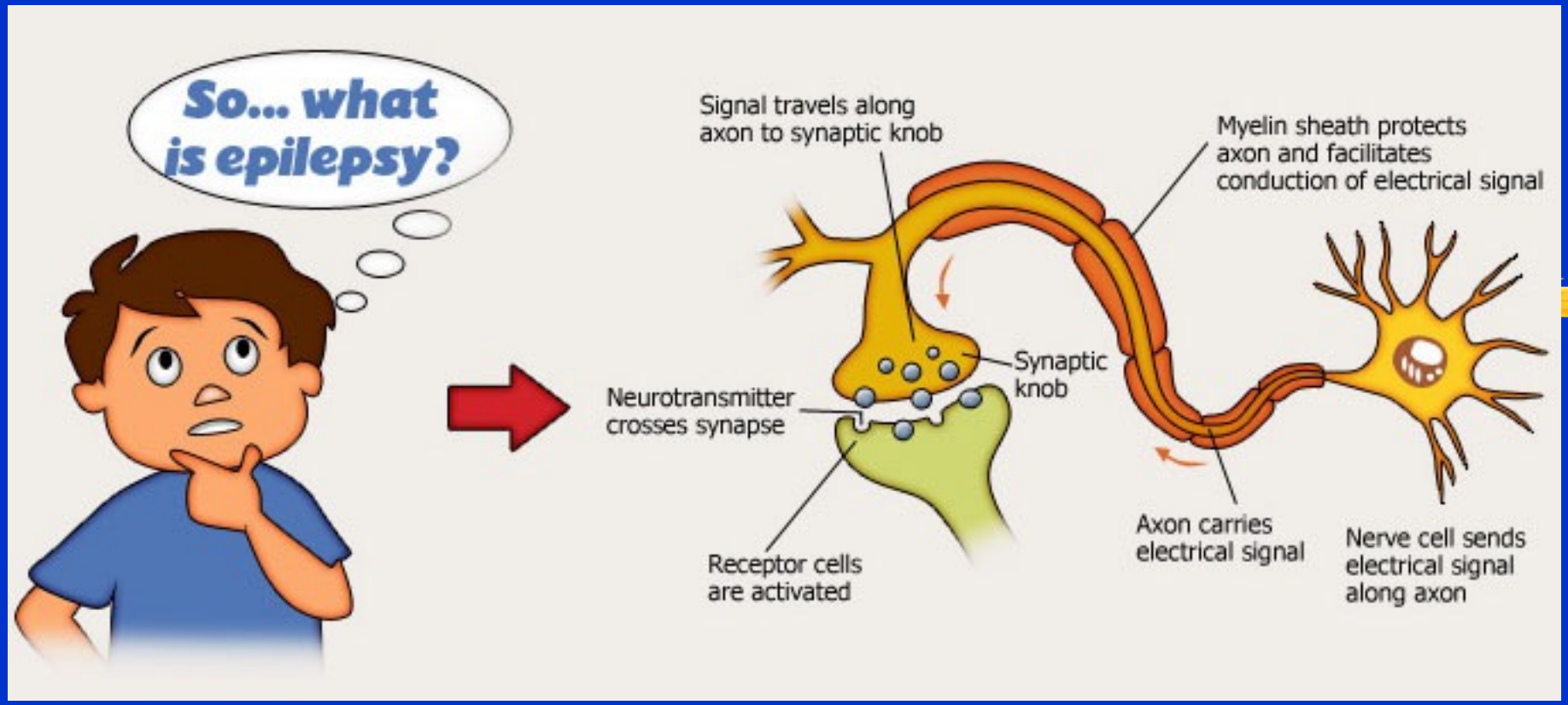
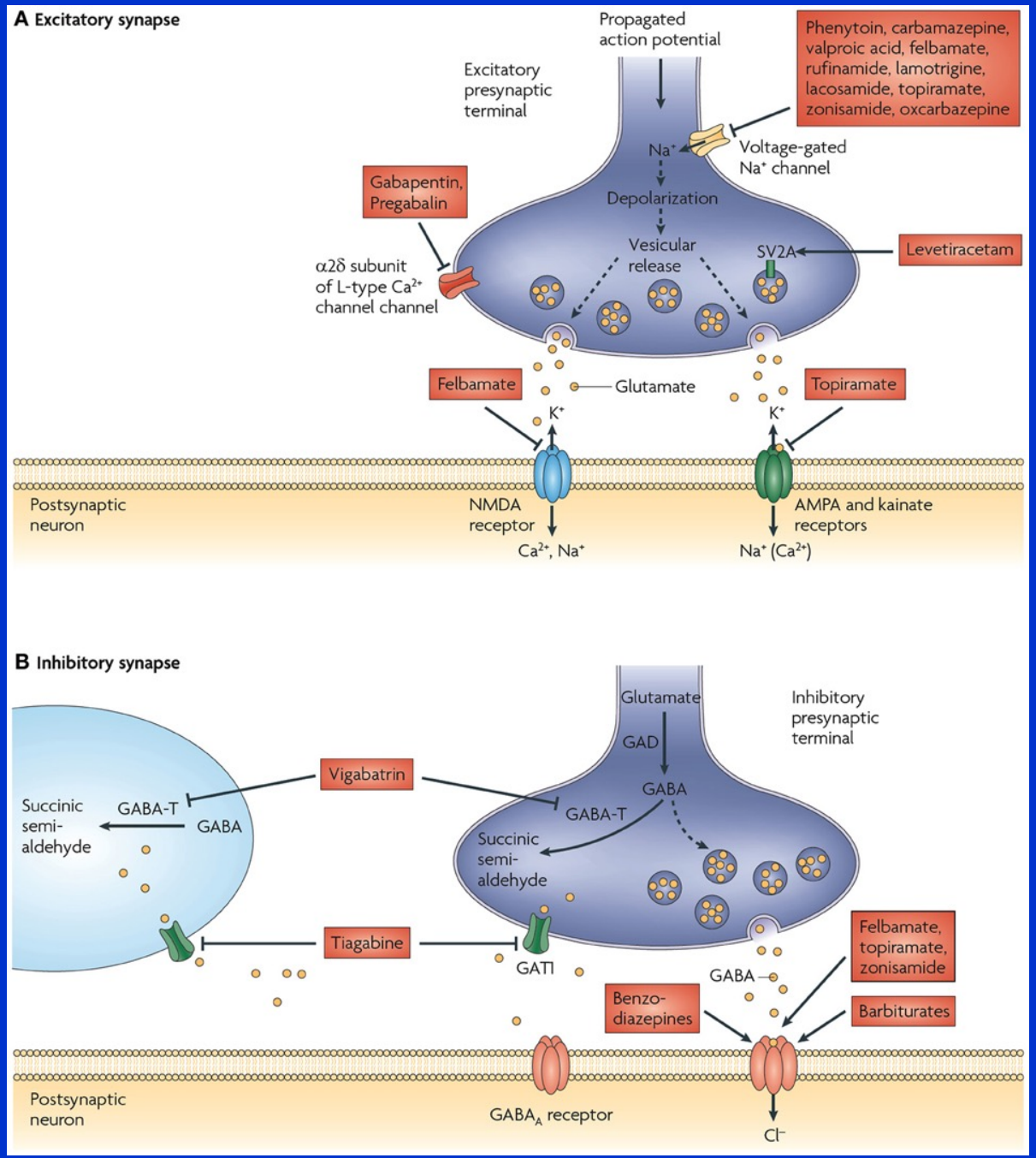



**Definition:**

**the clinical manifestation of an excessive excitation of a population of cortical neurons**

# Neurotransmitters:

## GABA vs Glutaminian



# Seizure and Epilepsy

**Seizure** is a paroxysmal episode of brain dysfunction manifested by stereotyped alteration in behaviour

Clinical manifestations of a seizure based on:

- anatomy of the brain that is seizing,
- age of patient and
- maturity of the brain

# Epilepsy

**Epilepsy:**

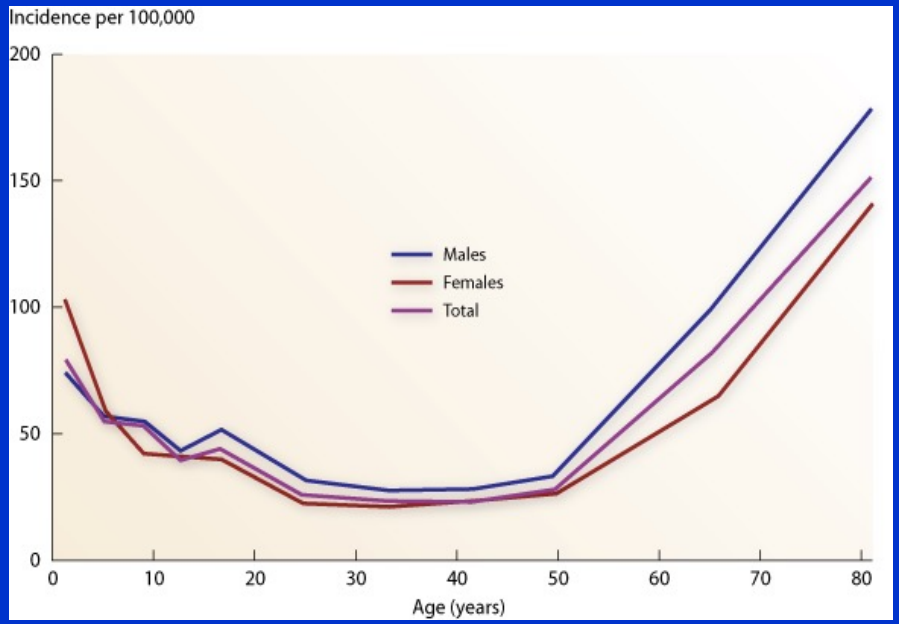
A group of long-term neurological symptoms characterized by presence of epileptic seizures

Epileptic seizure represents a transient condition characterized by excessive and repetitive bioelectrical discharges in neurons.

*Sudden, recurrent and unprovoked*

# Epilepsy: epidemiology

- Epilepsy affects 1-2% of the population
- Seizures including febrile seizures affect about 4-5% of pediatric population
- Epilepsy refractory to AEDs: 20-30%



# Operational definition of epilepsy

## Operational (Practical) Clinical Definition of Epilepsy

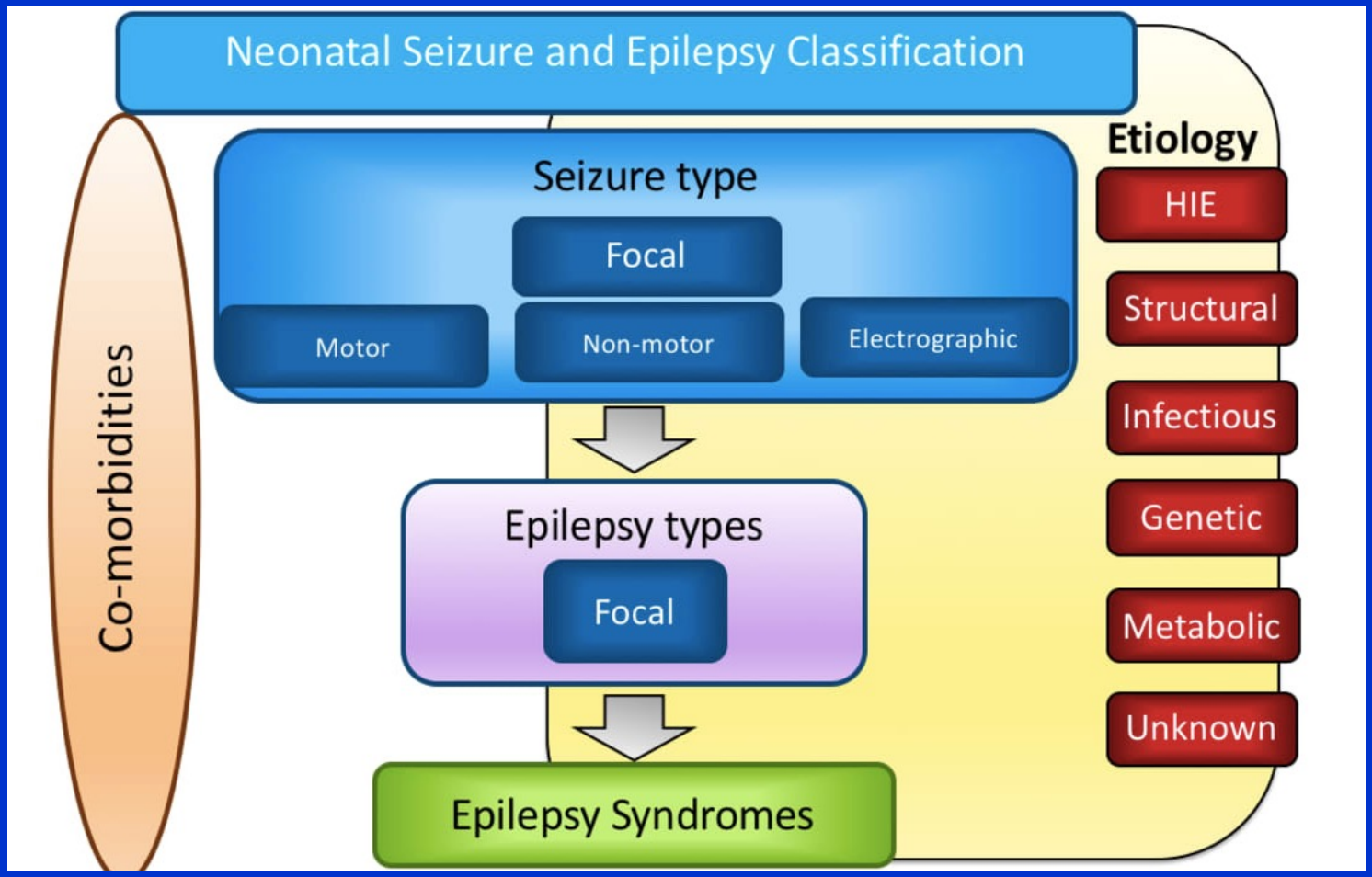
1. At least two unprovoked (or reflex) seizures occurring more than 24 hours apart;

2. One unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years;

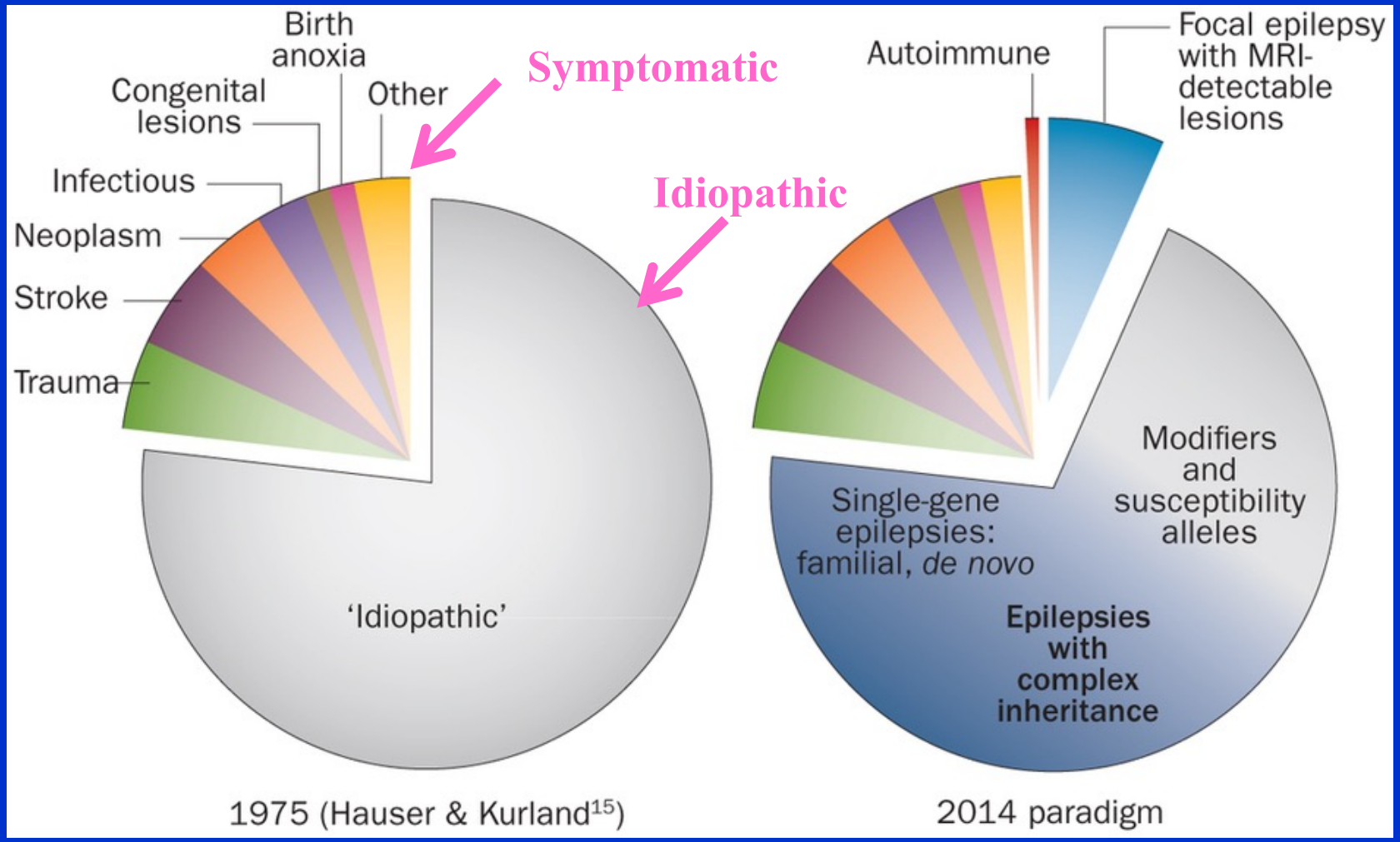
3. Diagnosis of an epilepsy syndrome.

Epilepsy is considered to be resolved for individuals who had age-dependent epilepsy syndrome but are now past the applicable age or those who have remained seizure-free for the last 10 years, with no seizure medicines for the last 5 years.

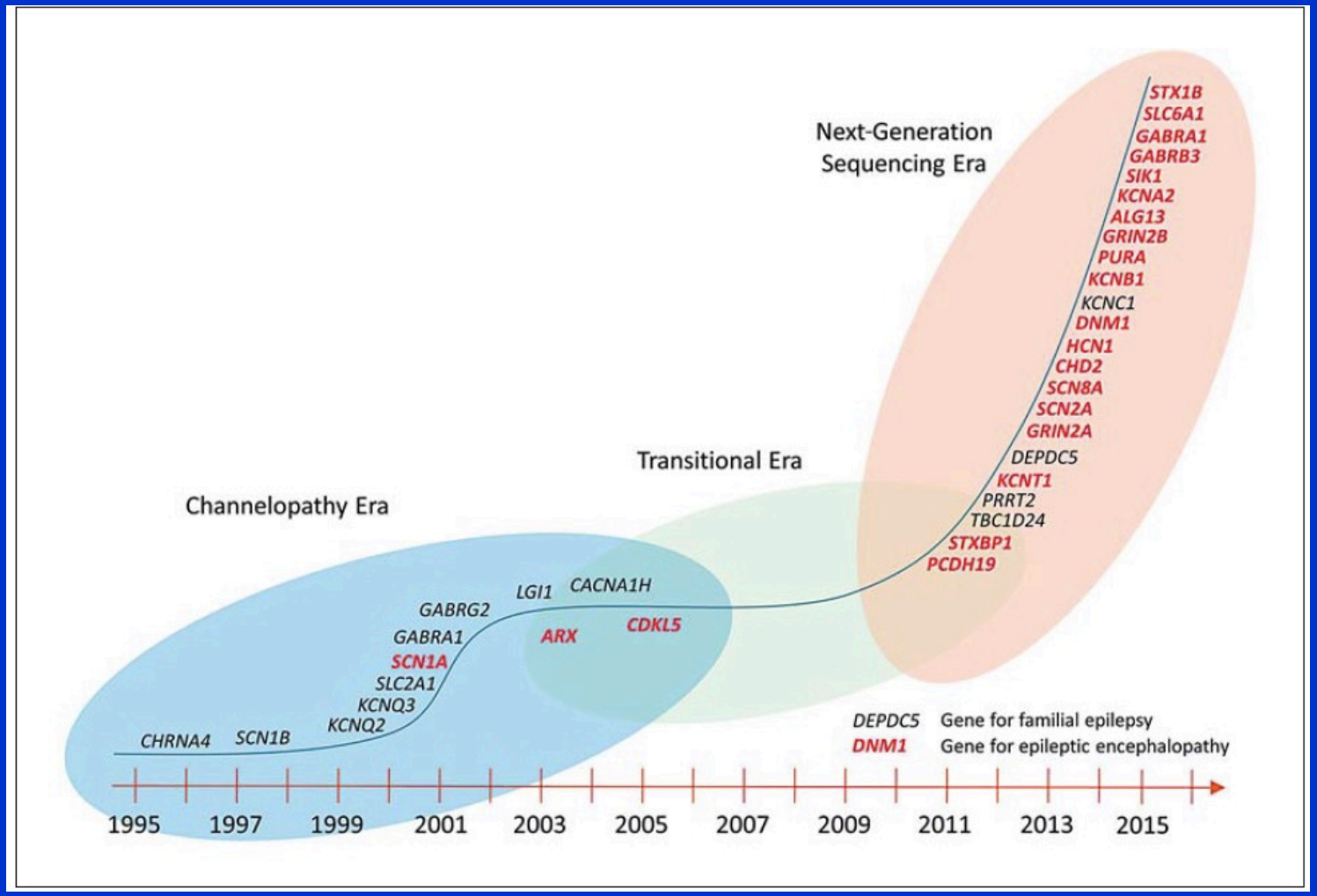
# Present classification of epilepsies



# Epilepsy: etiology

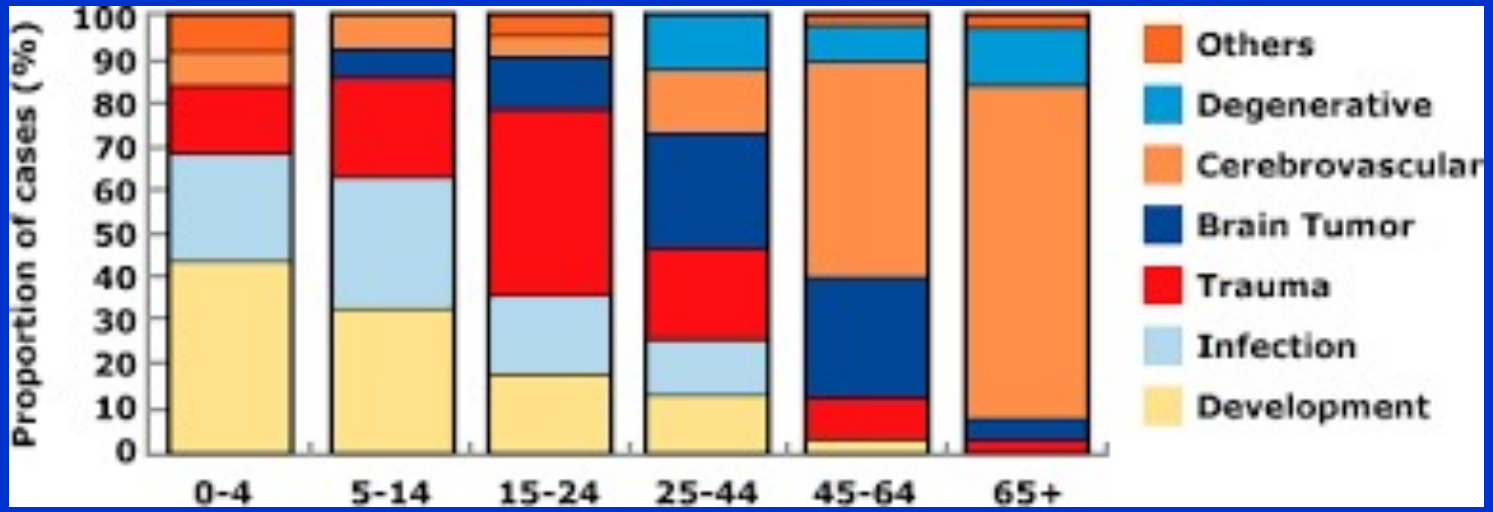


# Accelerated discovery of new epilepsy genes

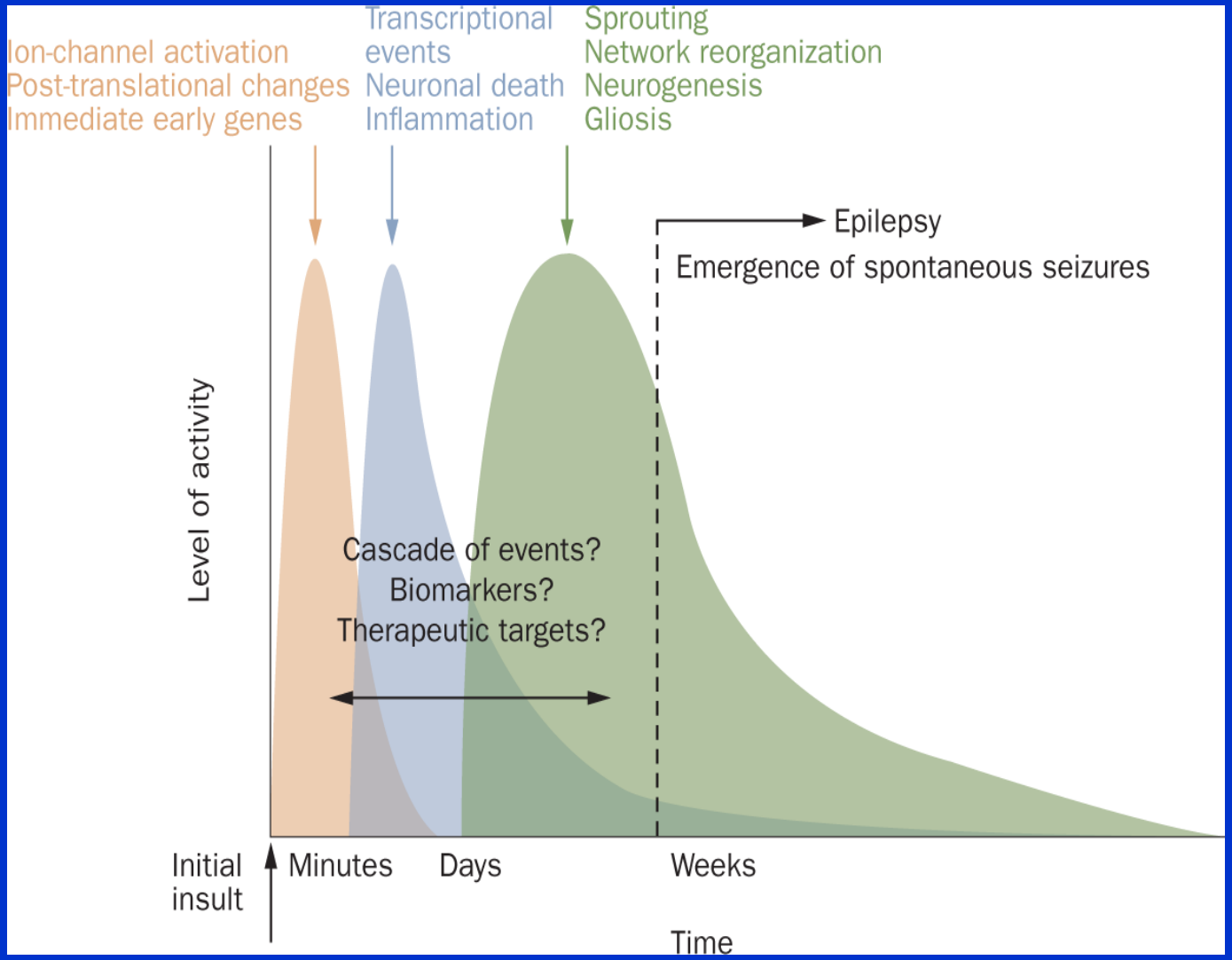



Helbig and Tayoun. Mol Syndromology 2016

# Epilepsy: etiology according to age

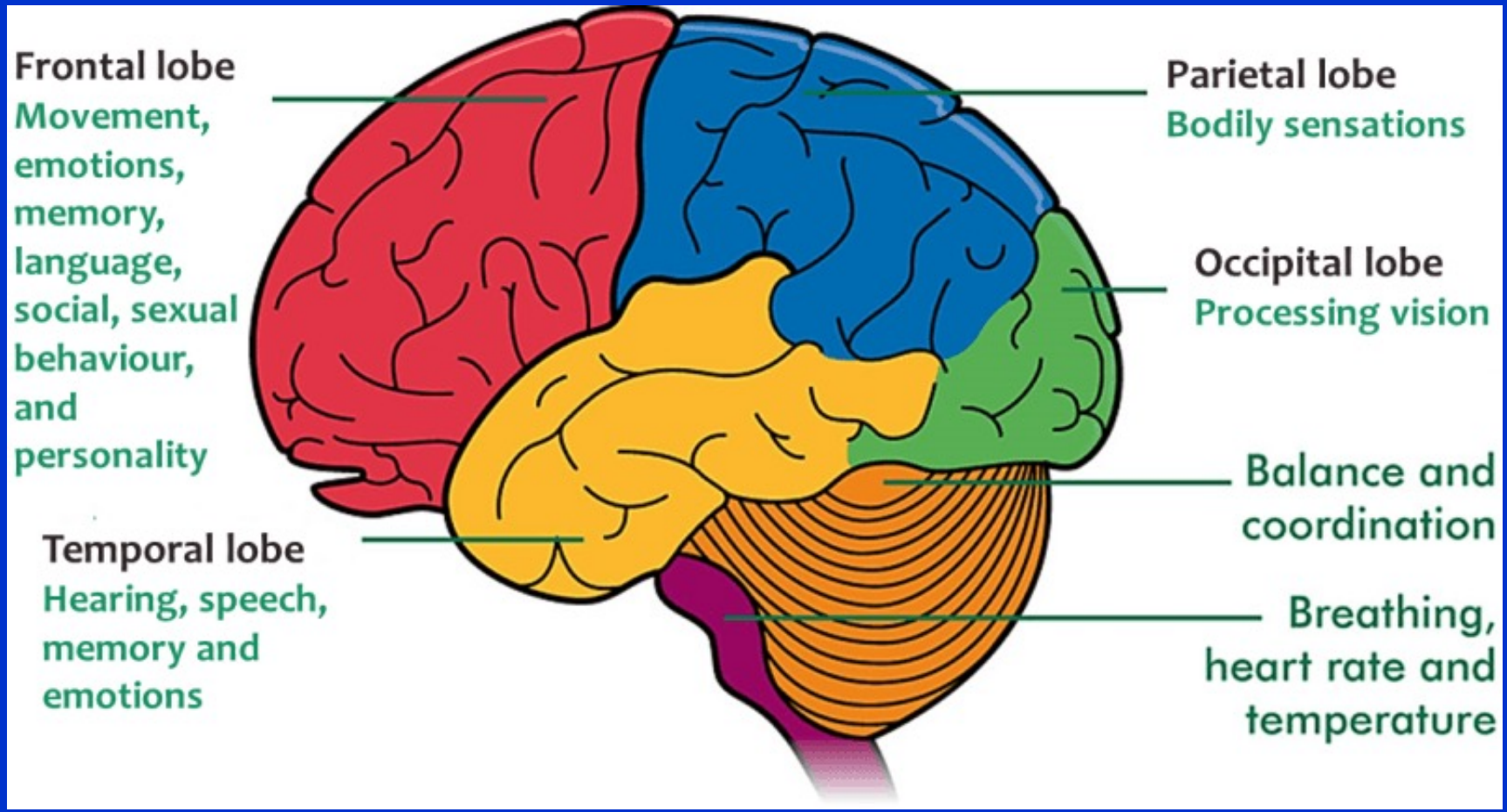


# Epileptogenesis as a process

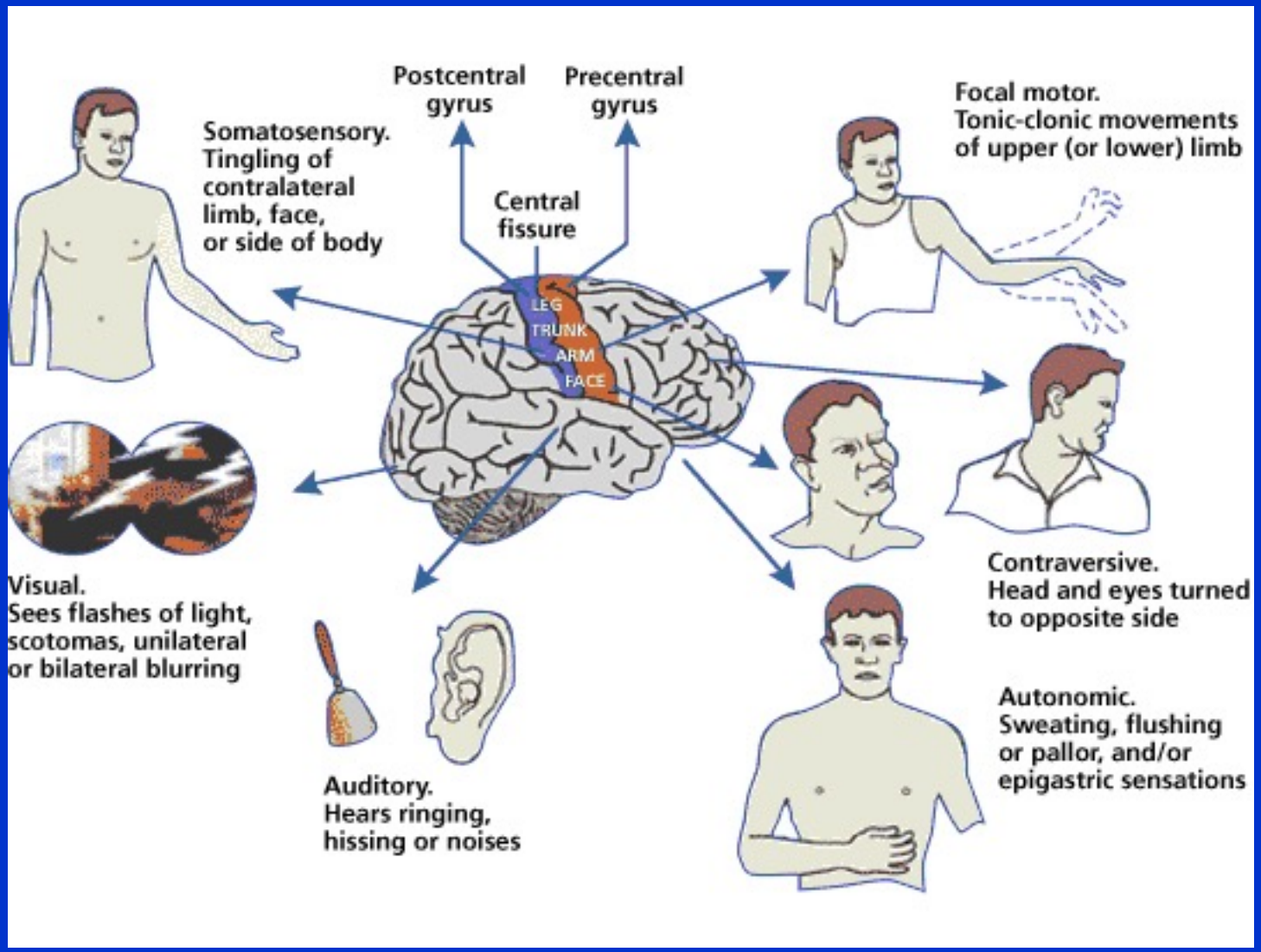



Rakhade et al. 2010

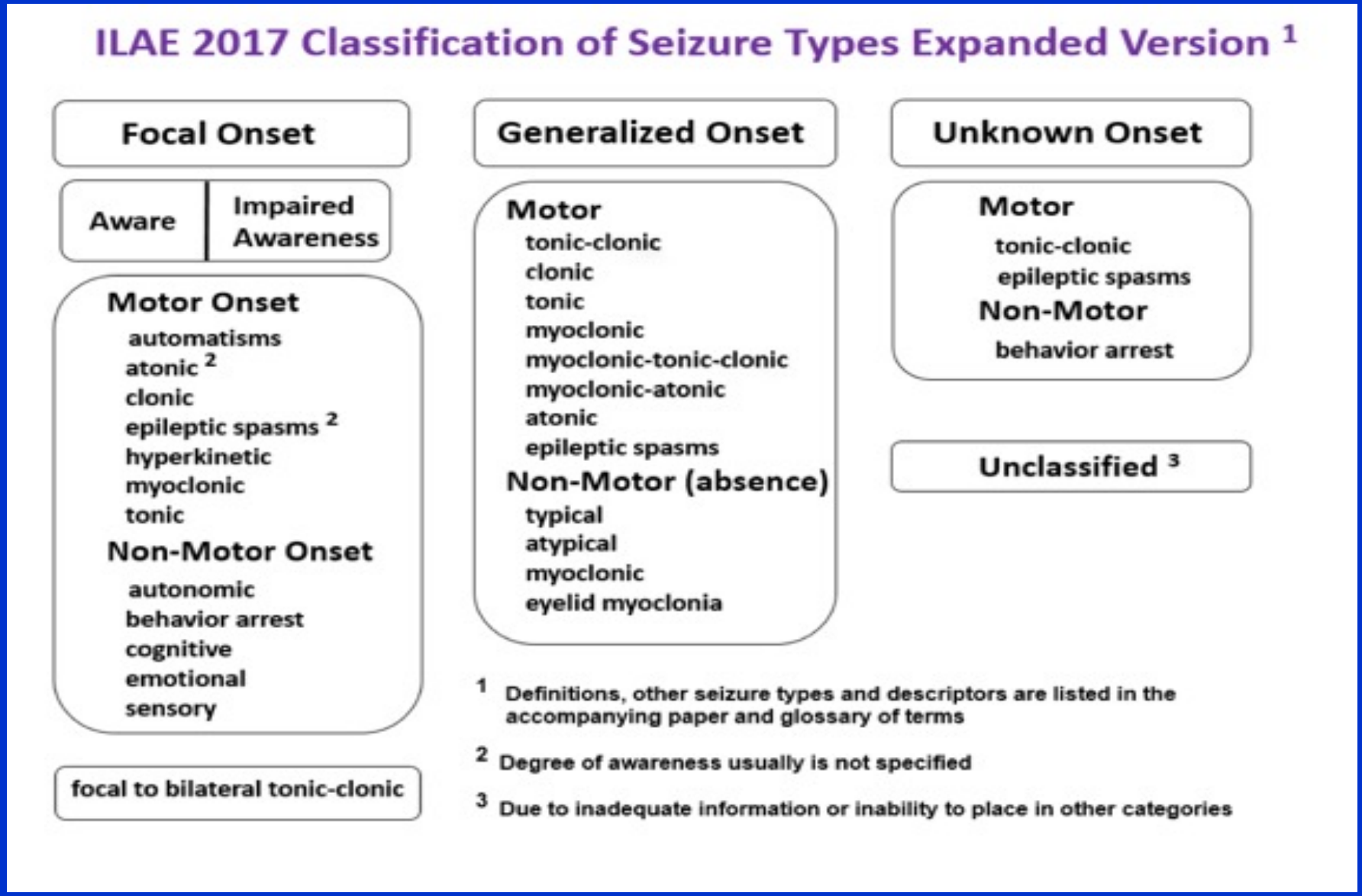
# Functional organisation of the brain



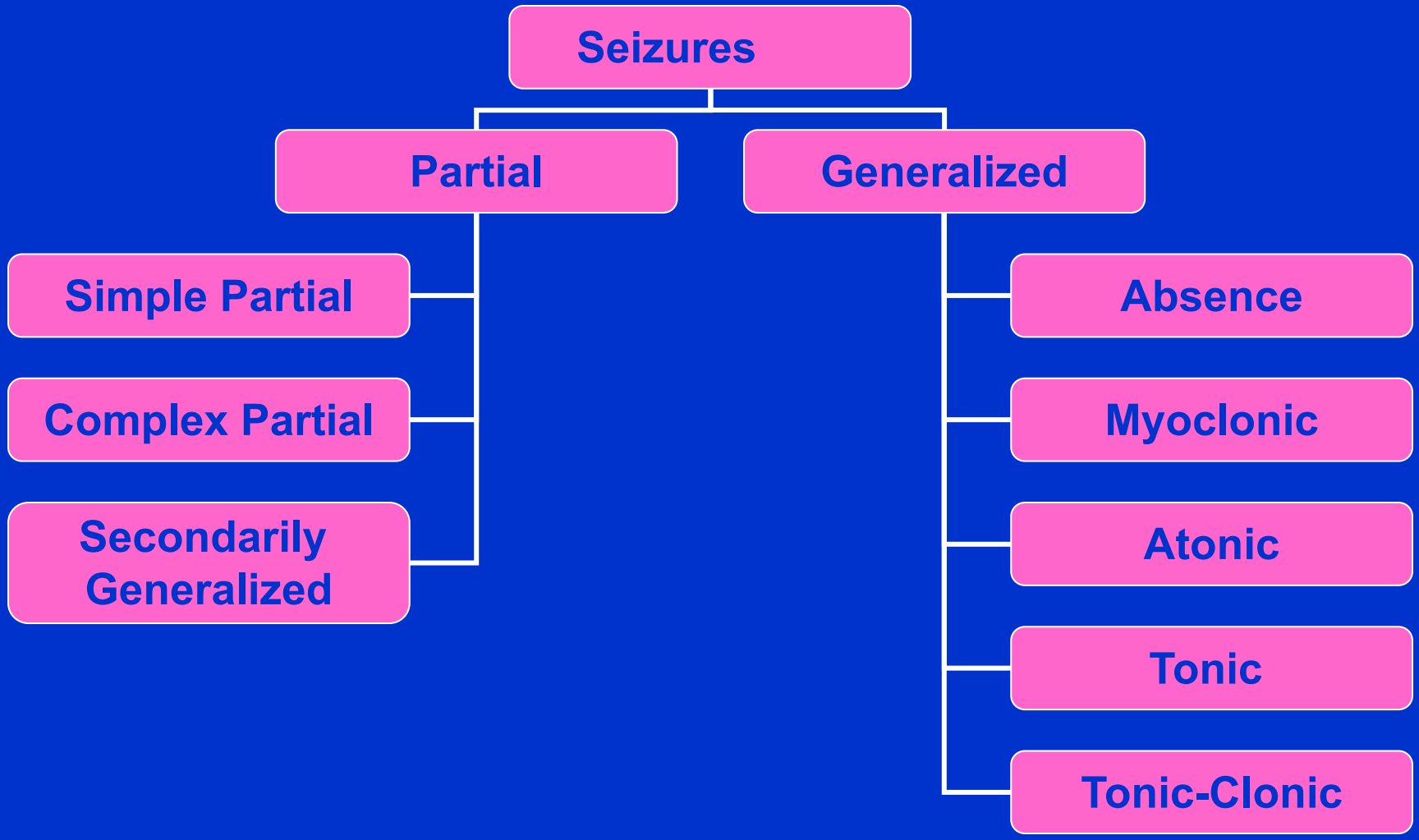
# Epileptic foci and morphology of seizure



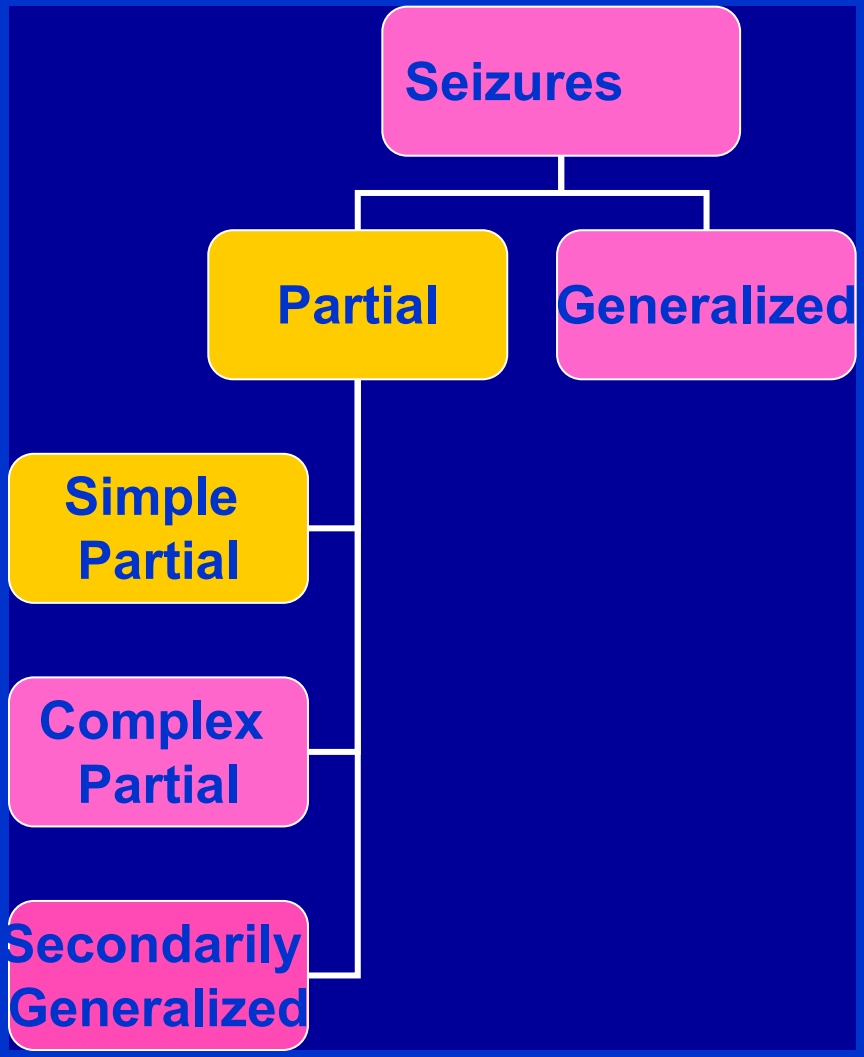
# Classification of seizures according to ILAE



# ILAE Classification of Seizures

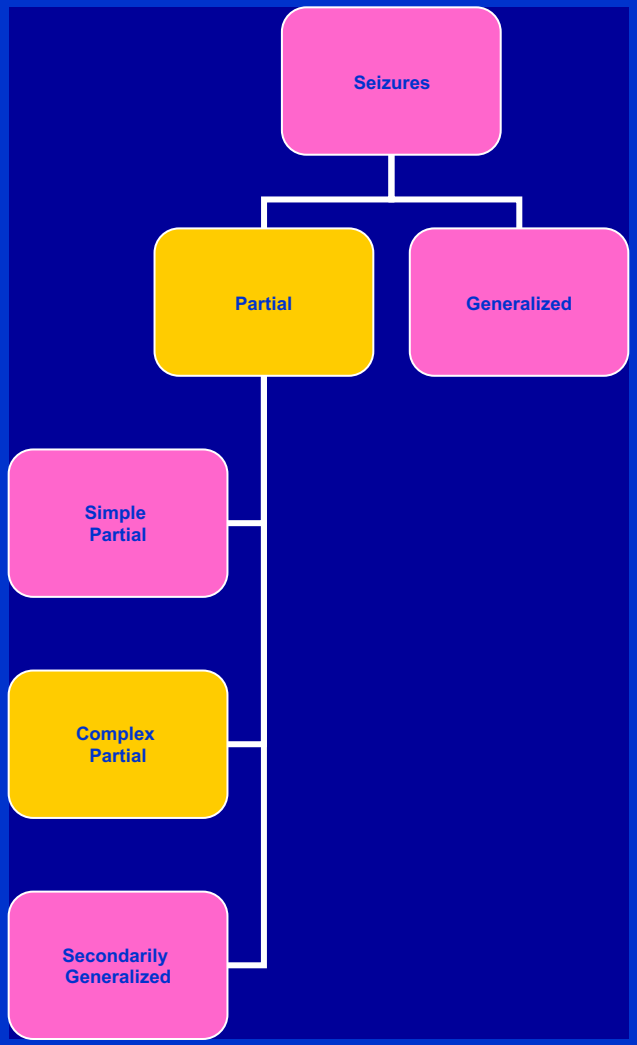


# Partial (focal) Seizures

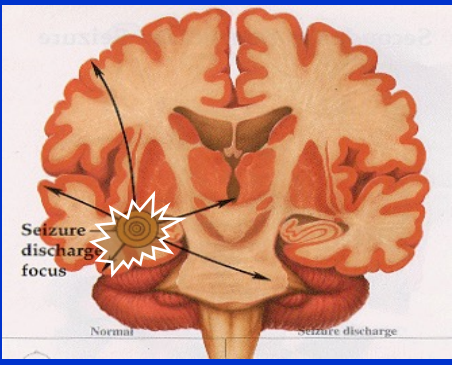



- **Simple Partial Seizure**
  - Preserved consciousness
  - Aura
  - Short duration 10-30 sec

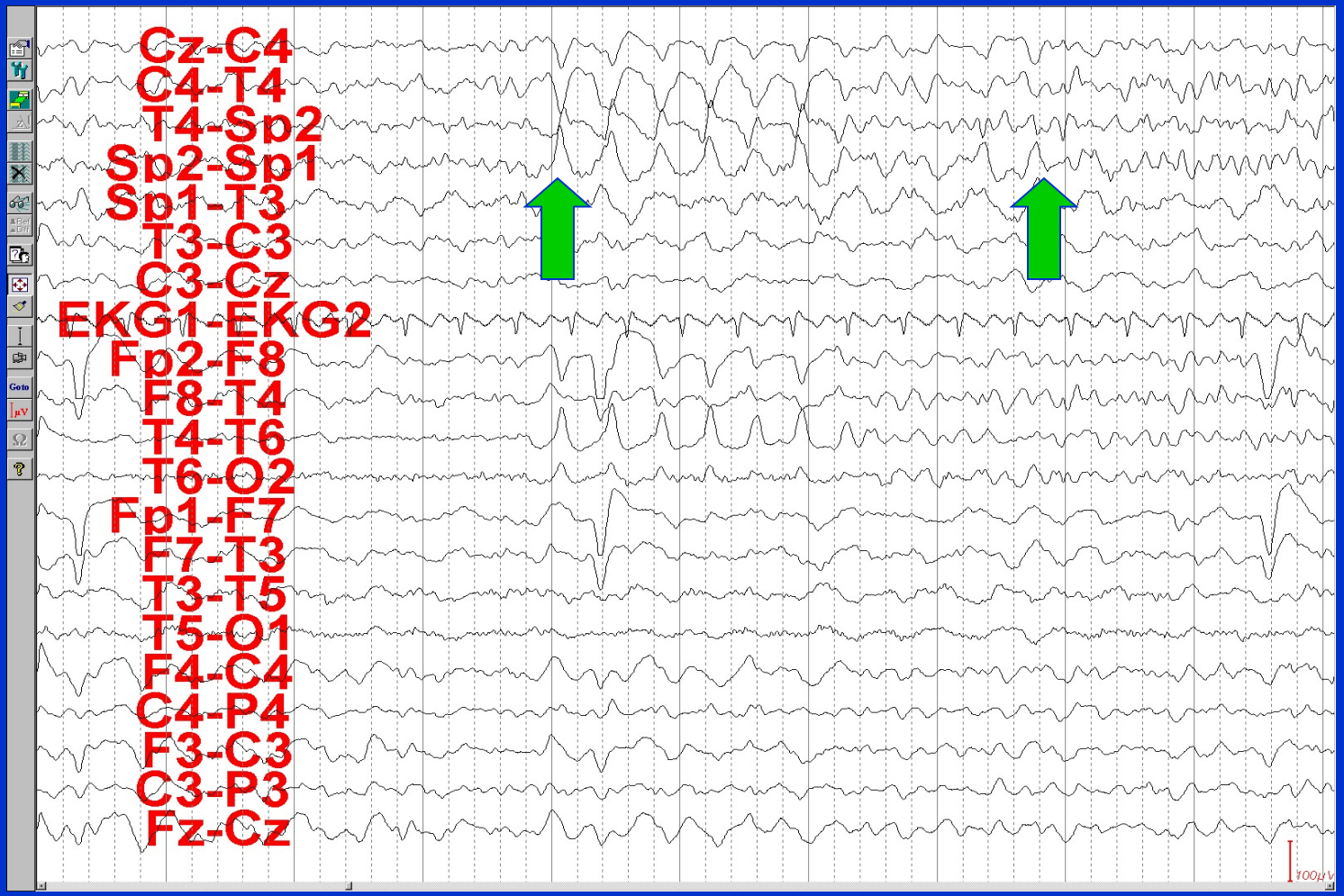
# Partial (focal) Seizures




- **Complex Partial Seizure**
  - Impaired consciousness/ level of awareness (staring)
  - Clinical manifestations vary with origin & degree of spread
  - Presence of aura
    - Temporal lobe: smell, epigastric sensation, déjà vu
  - Duration (typically 30 seconds to 3 minutes)
  - Amnesia and confusion often after event

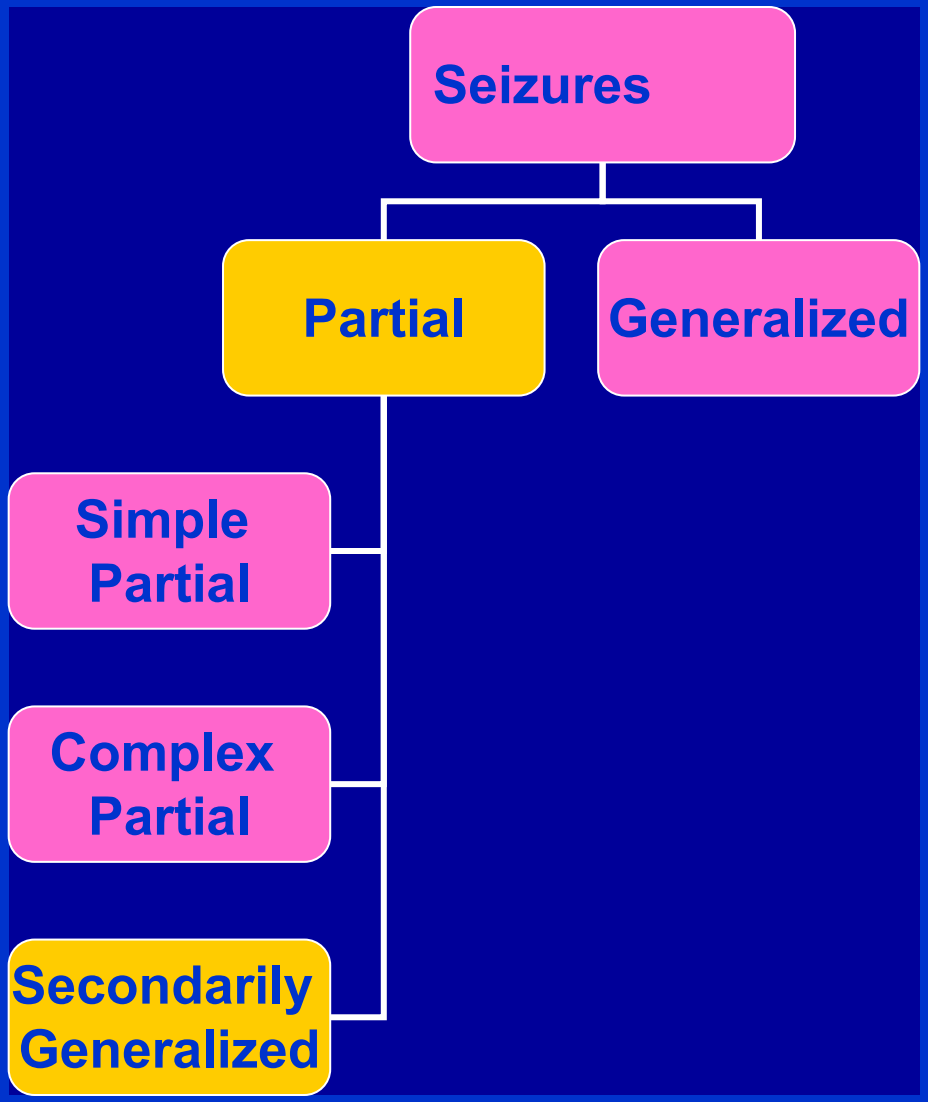


# EEG: Partial Seizure



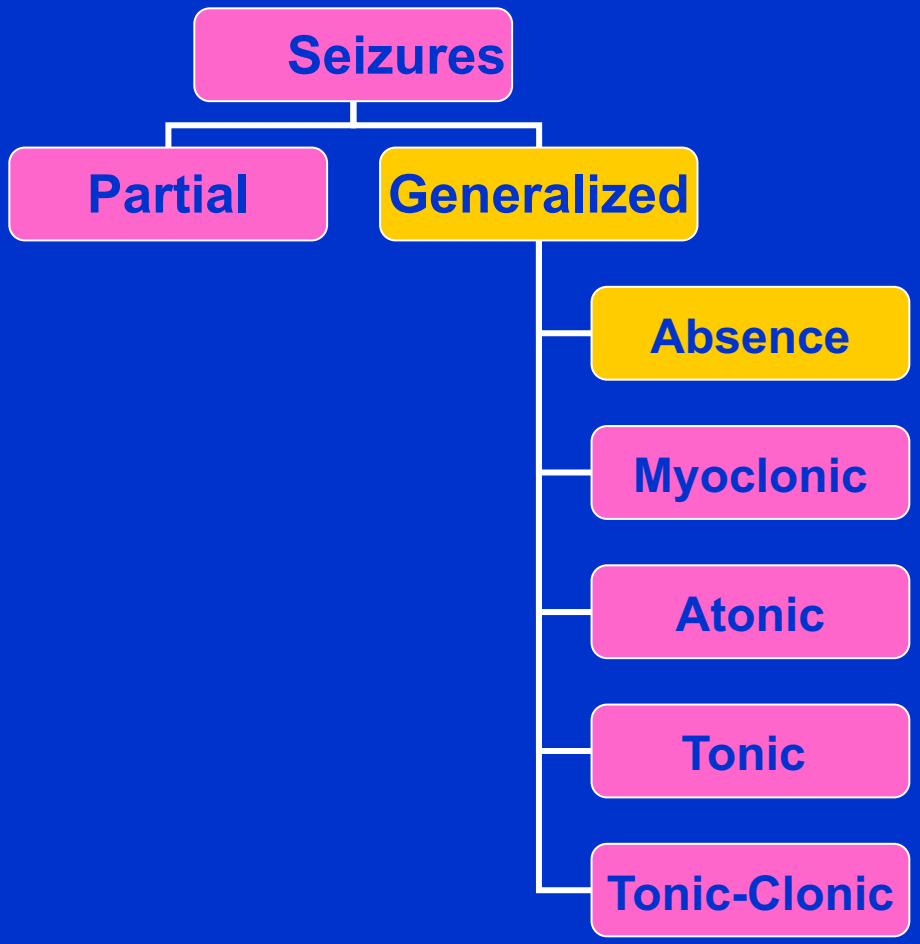

**Right temporal seizure with maximal phase reversal in the right temporal lobe**

# Secondarily Generalized Seizures



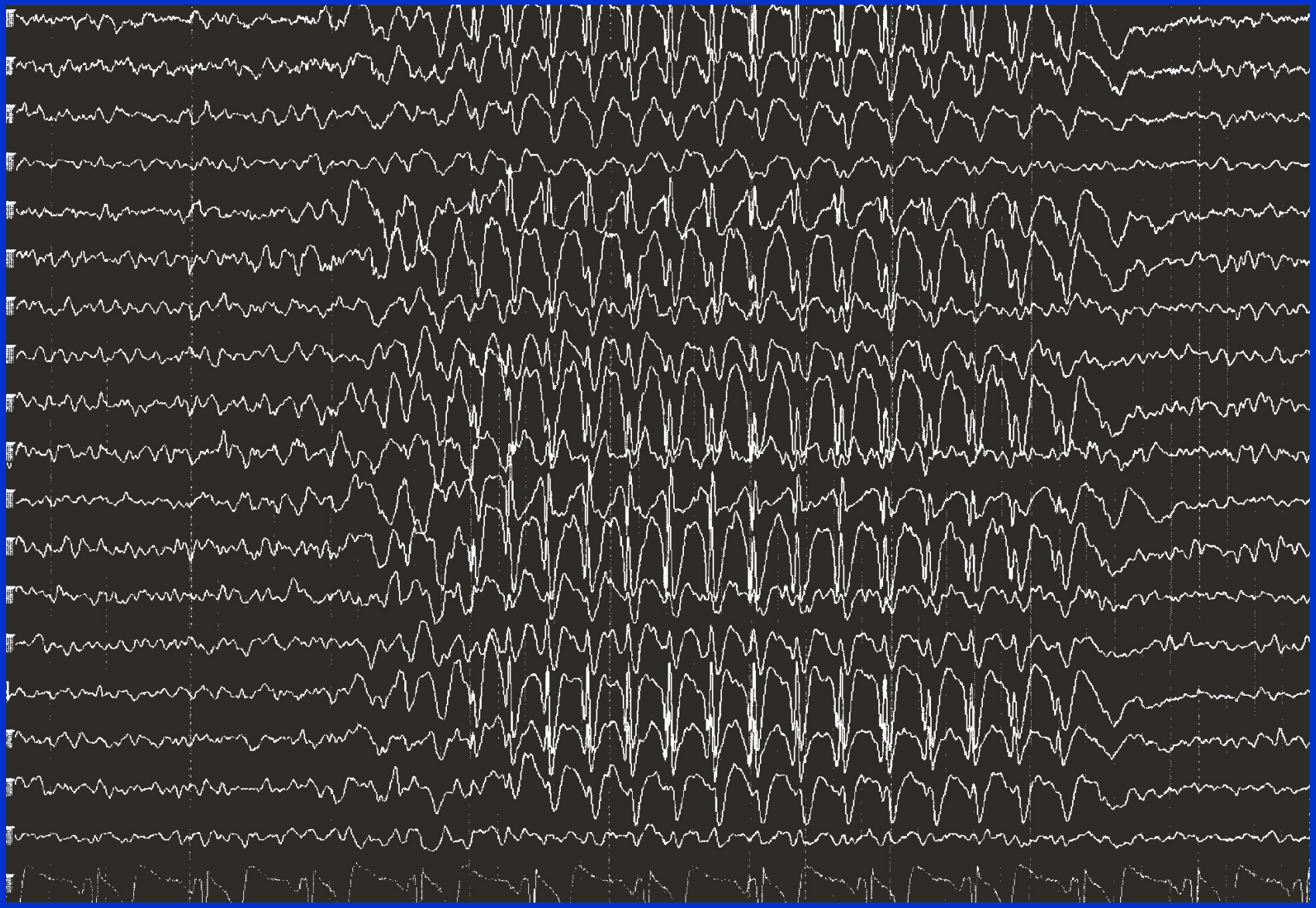

- Begins focally, with or without focal neurological symptoms
- Variable symmetry, intensity, and duration of tonic (stiffening) and clonic (jerking) phases
- Typical duration 1-3 minutes
- Postictal confusion, somnolence, with or without transient focal deficit

# Childhood Absence Seizures




- Brief staring spells ("petit mal") with impairment of awareness
  - 3-20 seconds
  - Sudden onset and sudden resolution
  - Often provoked by hyperventilation
  - Onset typically between 4 and 7 years of age
  - Often resolve by 18 years of age
- Normal development and intelligence
- EEG: Generalized 3 Hz spike-wave discharges

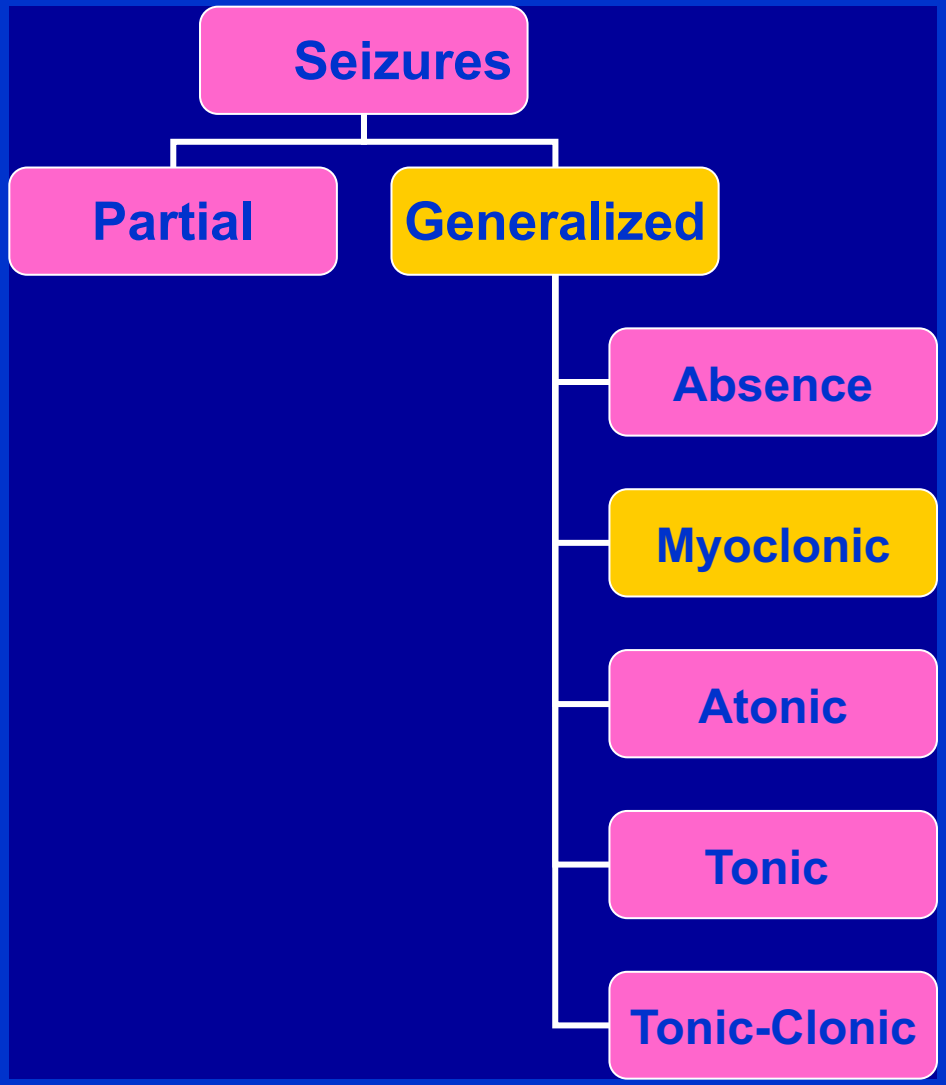
# EEG: Typical Absence Seizure



# Juvenile Absence Seizures

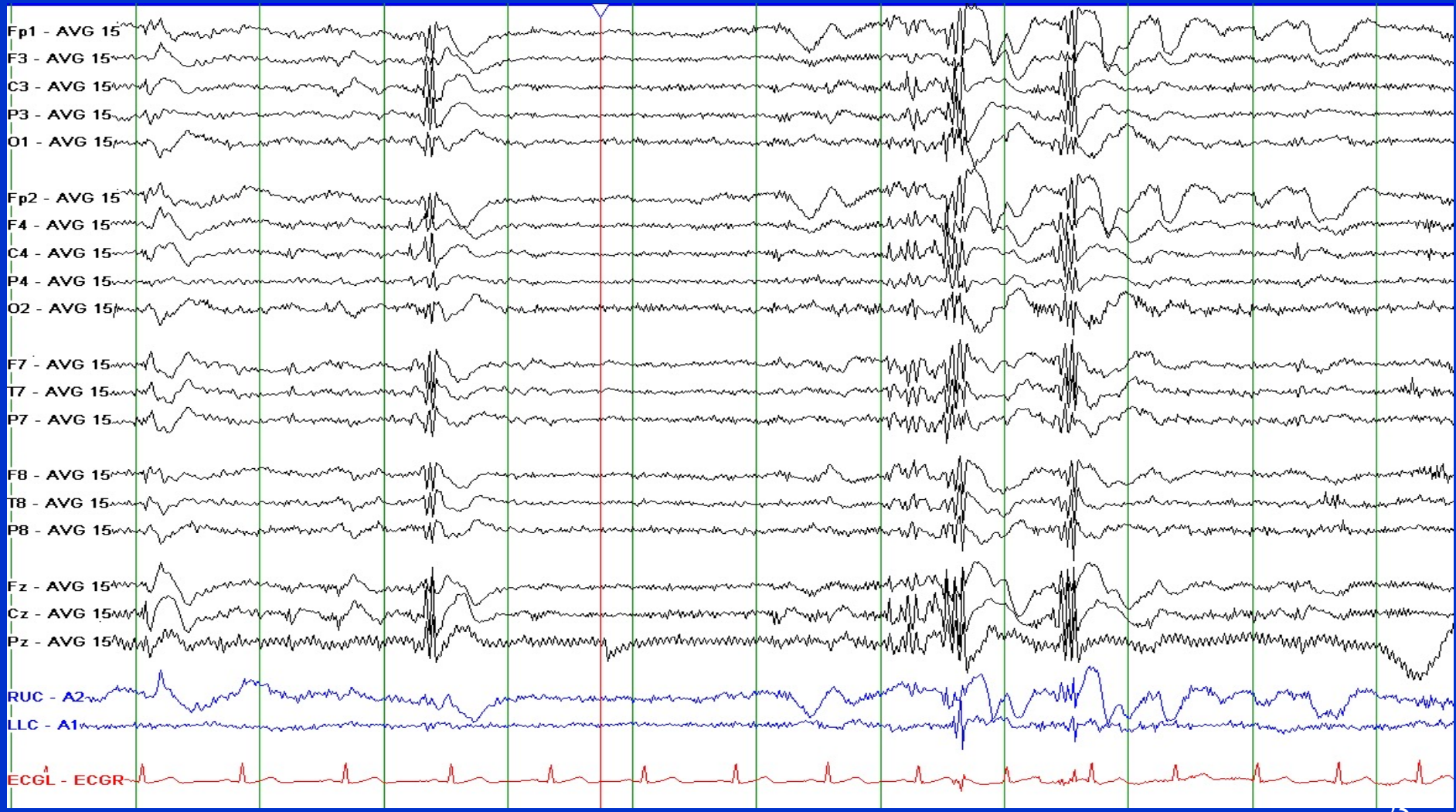
- Brief staring spells with variably reduced responsiveness
  - 5-30 seconds
  - Gradual (seconds) onset and resolution
  - Generally not provoked by hyperventilation
  - Onset typically after 7-8 years of age
  - Absence seizures are far less frequent than in childhood onset absence seizures
- Often evolve into myoclonic and generalized tonic-clonic seizures
- Patients continue to have seizures lifelong

# Myoclonic Seizures

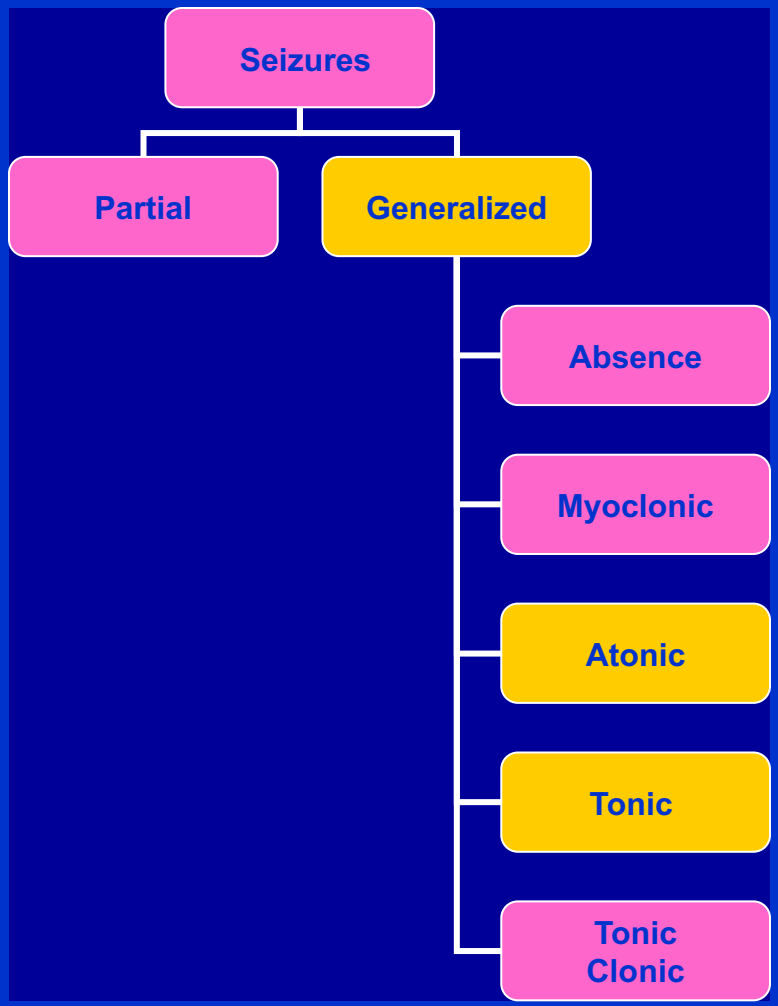



- Brief, shock-like jerk of a muscle or group of muscles
- Epileptic myoclonus
  - Typically bilaterally synchronous
  - Impairment of consciousness is difficult to assess (seizures <1 second)
  - Clonic seizure – repeated myoclonic seizures (may have impaired awareness)
- Differentiate from benign, nonepileptic myoclonus (e.g., while falling asleep)
- EEG: Generalized 4-6 Hz polyspike-wave discharges

# Myoclonic Seizures



# Tonic and Atonic Seizures



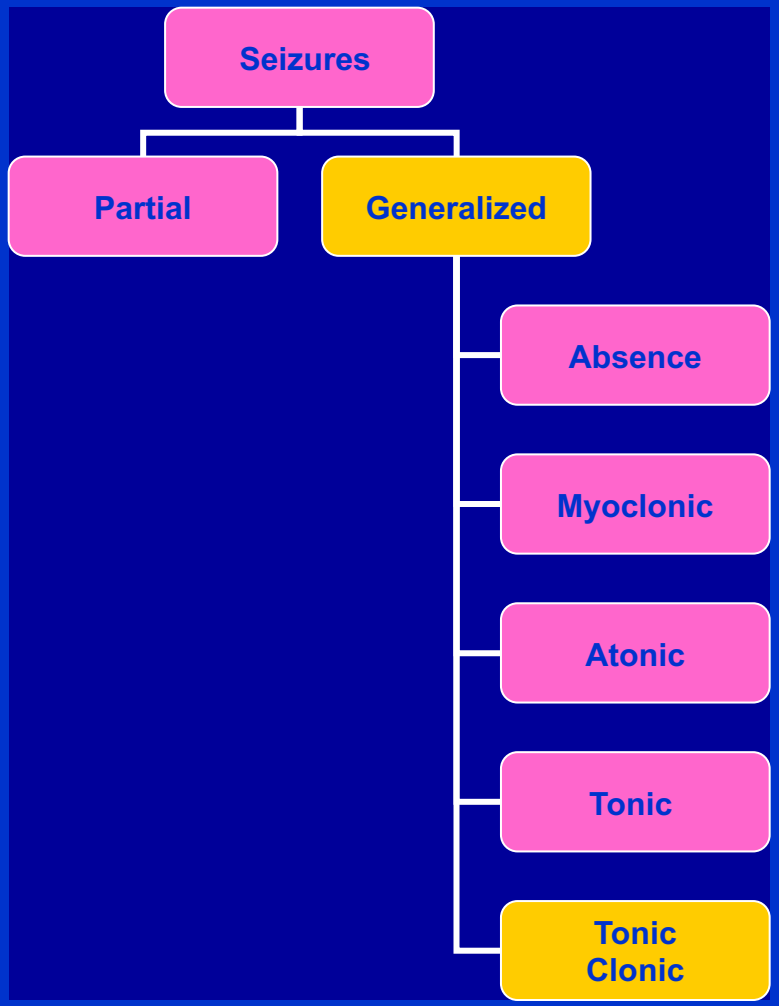

## Tonic seizures

- Symmetric, tonic muscle contraction of extremities with tonic flexion of waist and neck
- Duration - 2-20 seconds.
- EEG – Sudden attenuation with generalized, low-voltage fast activity (most common) or generalized polyspike-wave.

## Atonic seizures

- Sudden loss of postural tone
  - When severe often results in falls
  - When milder produces head nods or jaw drops.
- Consciousness usually impaired
- Duration - usually seconds, rarely more than 1 minute
- EEG – sudden diffuse attenuation or generalized polyspike-wave

# Generalized tonic-clonic seizures



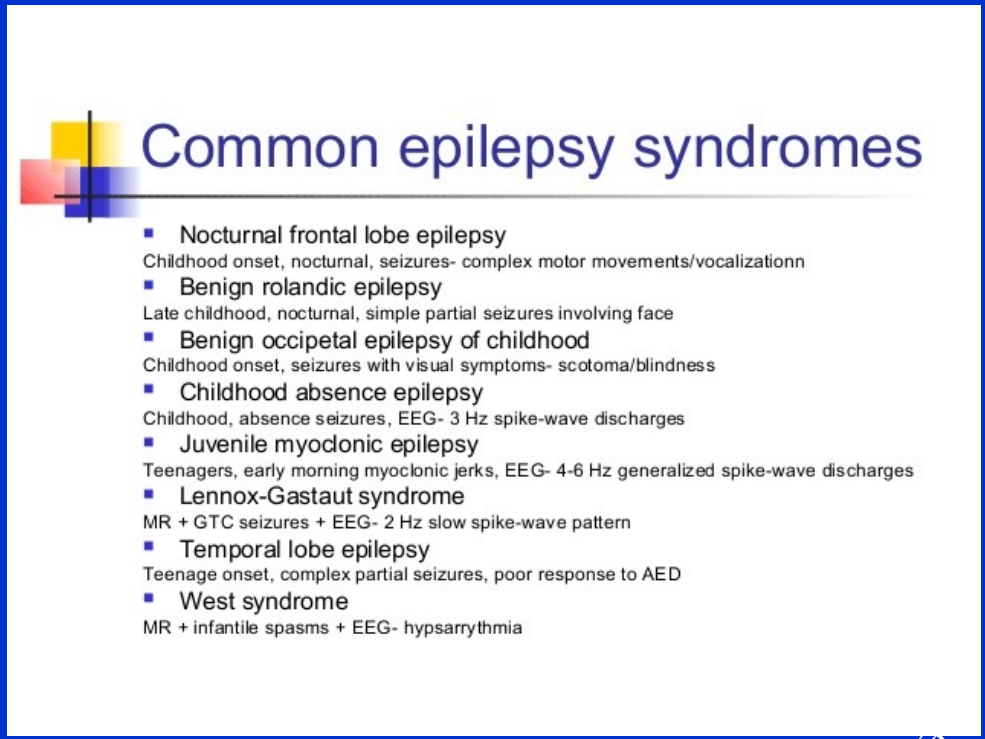

- Loss of consciousness
- First phase: tonic
  - Lasts about 1 minute
  - Tonic spasm of large muscle groups leading to abnormal breathing, cyanosis, tongue bites, and urination
- Second phase: Clonic
  - Lasts about 1 minute
  - Starts with deep breath and multiple clonic jerks
- During event – tachycardia, salivation, wide pupils.

# Epilepsy Syndromes

**Epilepsy Syndrome**

***Grouping of patients that share similar:***

- **Seizure type(s)**
- **Age of onset**
- **Natural history/Prognosis**
- **EEG patterns**
- **Genetics**
- **Response to treatment**



# Infantile spasms
# West syndrome

- Onset ages 3-12 months
- Brief axial contractions
  - usually bilateral, may be asymmetrical
  - typically flexor, may be extensor
  - usually in clusters, less likely random
  - typically on awakening, or when drowsy
- EEG shows hypsarrhythmia
  - multifocal spikes
  - high voltage, chaotic background

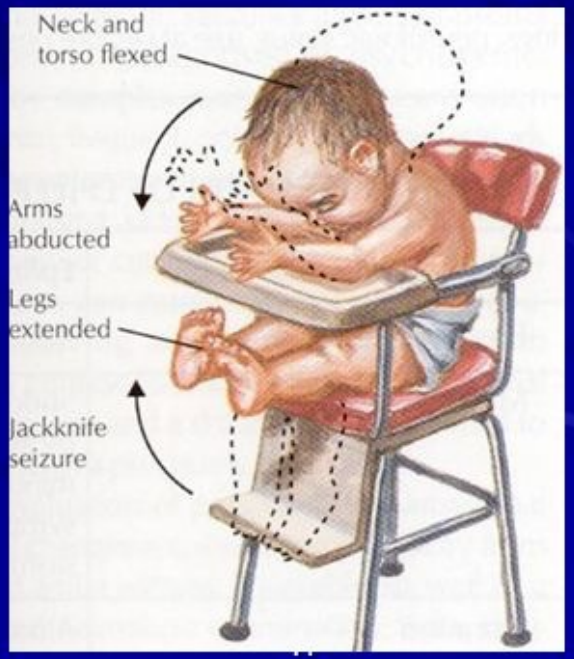
ON A PECULIAR FORM OF INFANTILE CONVULSIONS.

*To the Editor of* THE LANCET.

W. J. WEST.

Tunbridge, Jan. 26, 1841.

P.S.—In my own child's case, the bowing convulsions continued every day, without intermission, for seven months; he had then an interval of three days free; but, on the fourth day, the convulsions returned, with this difference, instead of bowing, he stretched out his arms, looked wild, seem to lose all animation, and appeared quite exhausted.



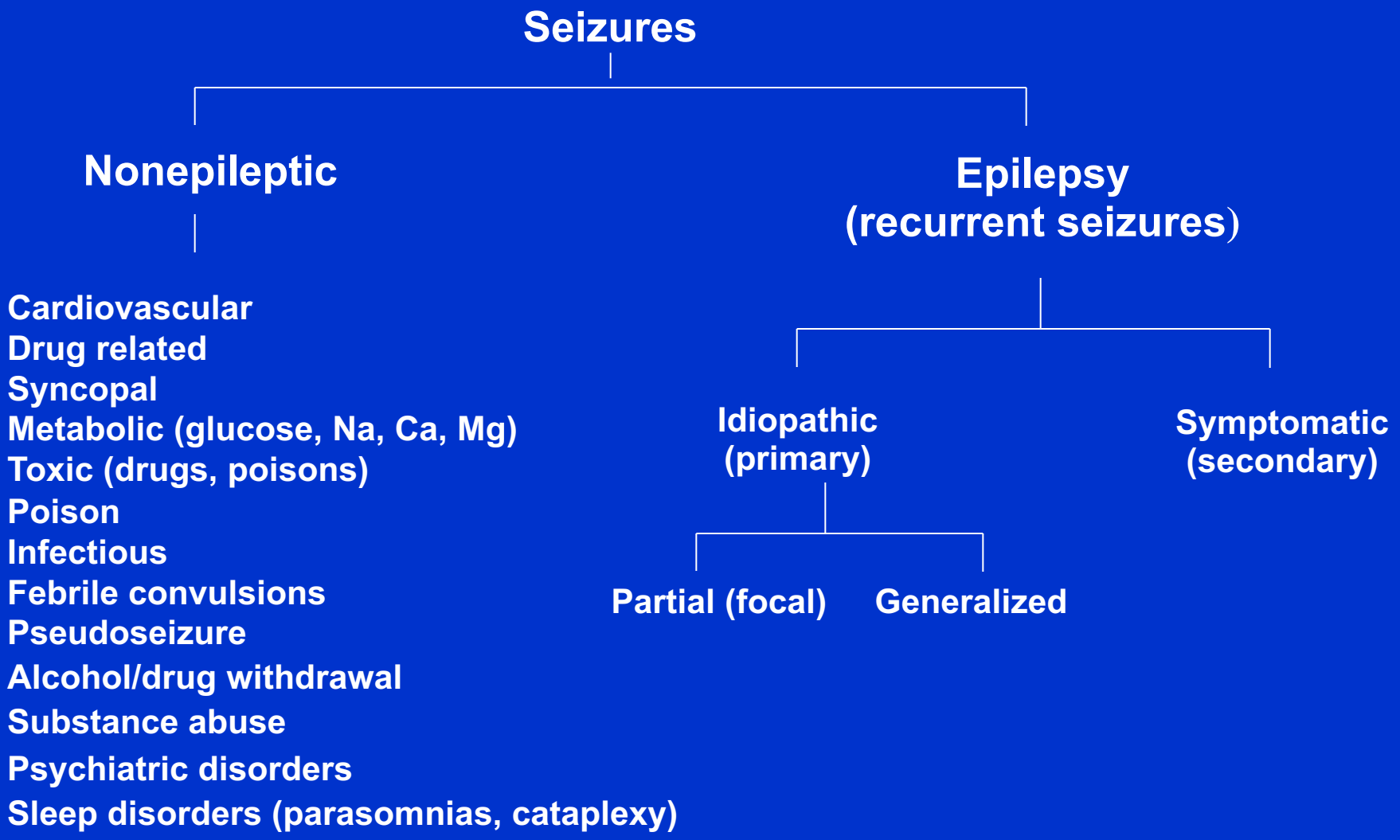
# Psychogenic/Non-epileptic Events

- pseudoseizures
- Represent genuine psychiatric disease
- 10-45% of refractory epilepsy at tertiary referral centers
- Females > males
- Psychiatric mechanism:
  dissociation, conversion, most unconscious (unlike malingering)
- Association with physical, sexual abuse
- Epileptic and nonepileptic seizures may co-exist
- Video-EEG monitoring often helps clarify the diagnosis
- Once recognized, approximately 50% respond well to specific psychiatric treatment

# Febrile seizures in 5% of pediatric population

| Simple FS | Complex FS |
|---|---|
| >90% of FS, | |
| Usually between 6m and 5 ys | |
| Generalized | Focal |
| Lasting <15 minutes | Lasting > 15 minutes |
| Does not reoccur within 24 hours | Reoccuring in 24 hours |

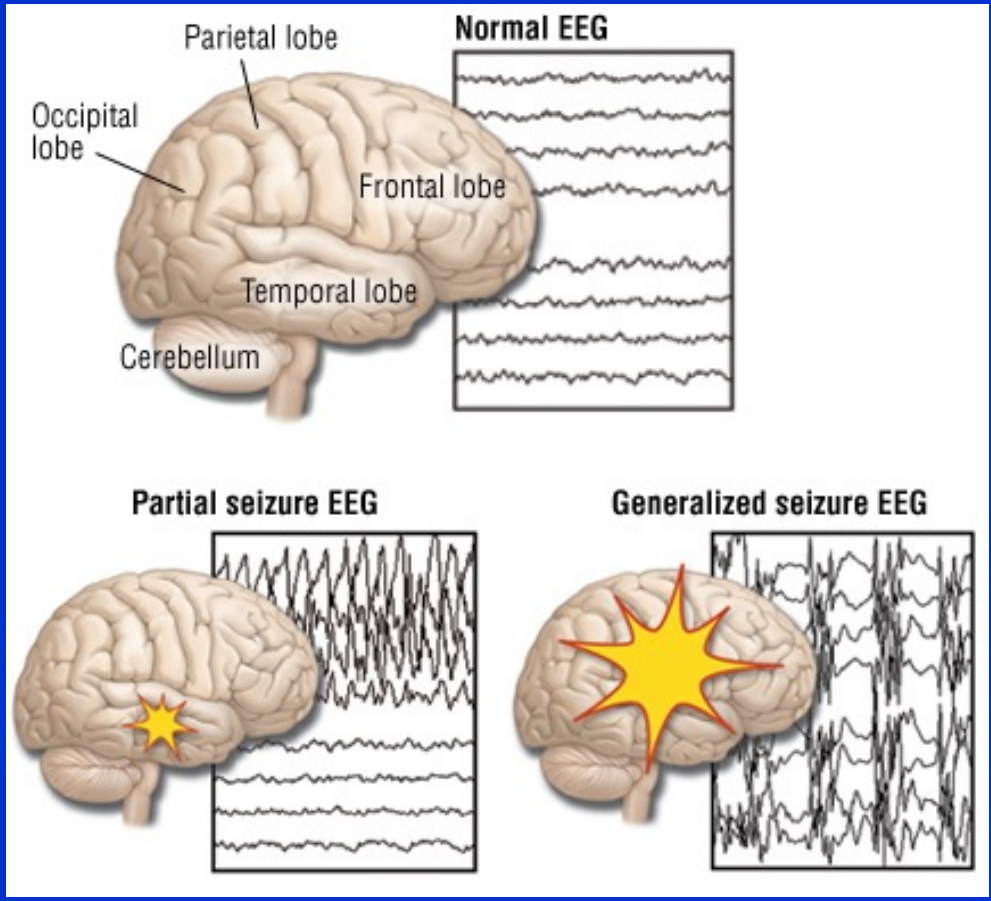
# Differential Diagnosis of Seizures



# Resources for the Diagnosis of Epilepsy

## EEG (ElectroEncephaloGraphy)

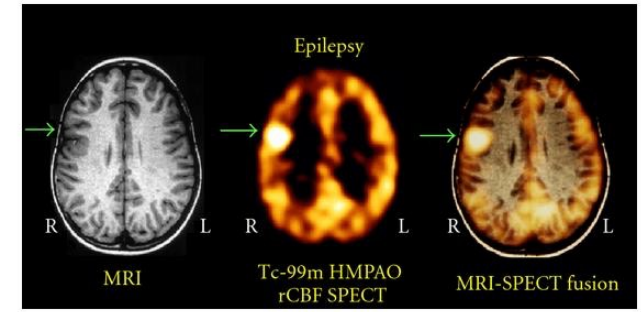
Electroencephalography is a measurement of the electrical activity of the brain by recording from electrodes placed on the scalp. Includes video EEGs and sleep EEG.



# Resources for the Diagnosis of Epilepsy

- **MRI (Magnetic Resonance Imaging)**

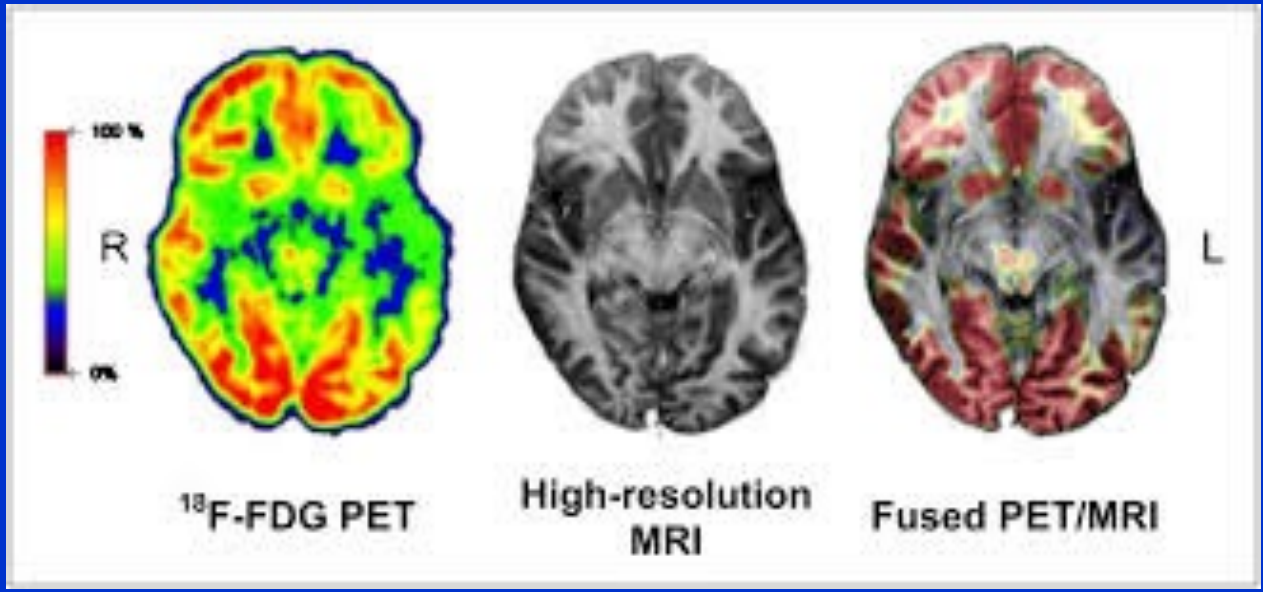
A method of creating images of the structure and contents of the brain using a powerful, uniform magnetic field.




# Resources for the Diagnosis of Epilepsy

- **PET (Positron Emission Tomography)**

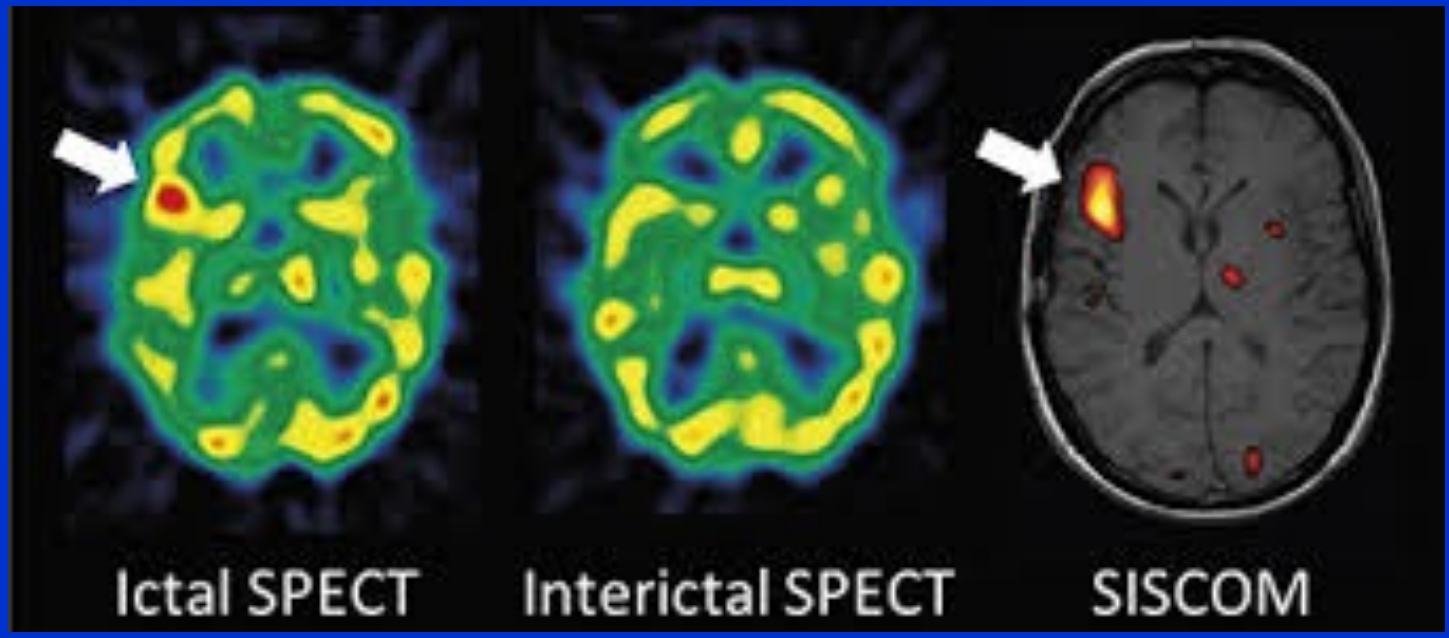
An advanced imaging technique that involves the acquisition of images of the brain based on the detection of radiation from the emission of positrons. Positrons are tiny particles emitted from a radioactive substance administered to the patient.



# Resources for the Diagnosis of Epilepsy

- **Ictal SPECT (Single Photon Emission Computed Tomography)**

SPECT scans show brain function (what the brain is doing). SPECT involves an intravenous injection of substances that are given during or immediately following a seizure (Ictal SPECT).



# Recovery position

Topical Review Article

# Treatment of Infantile Spasms: Emerging Insights From Clinical and Basic Science Perspectives

Journal of Child Neurology
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DOI: 10.1177/0883073811413129
http://jcn.sagepub.com
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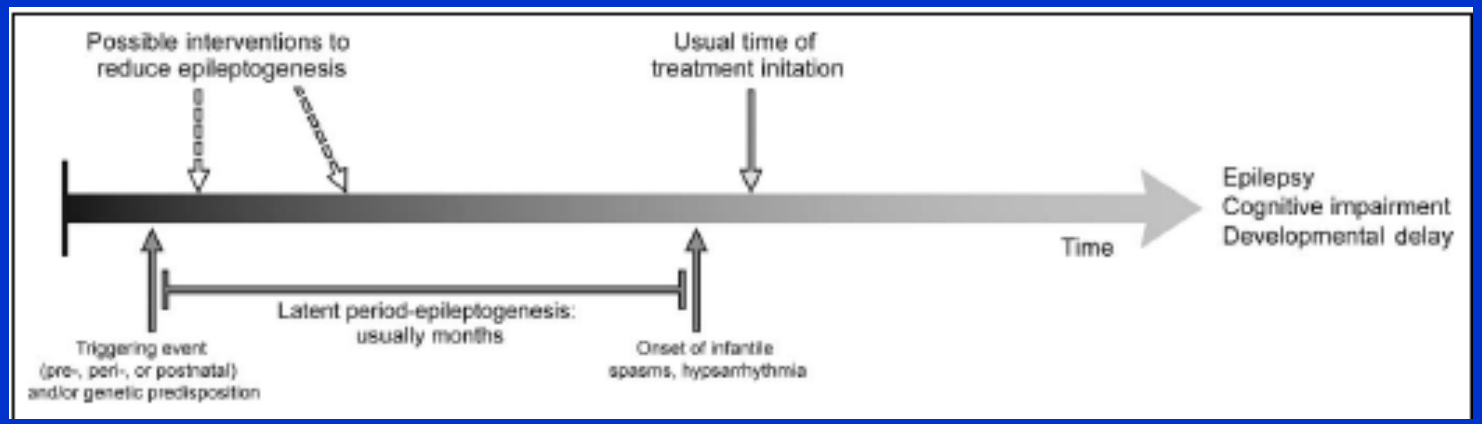
Carl E. Stafstrom, MD, PhD[1], Barry G. W. Arnason, MD[2], Tallie Z. Baram, MD, PhD[3], Anna Catania, MD[4], Miguel A. Cortez, MD[5], Tracy A. Glauser, MD[6], Michael R. Pranzatelli, MD[7], Raili Riikonen, MD, PhD[8], Michael A. Rogawski, MD, PhD[9], Shlomo Shinnar, MD, PhD[10], and John W. Swann, PhD[11]




**Antiepileptogenic treatment !**

J.Child Neurology 2011, Nov; 26(11): 1411-21.

# Treatment Sequence for Pharmacoresistent Epilepsy

1st Monotherapy AED Trial

2nd Monotherapy AED Trial

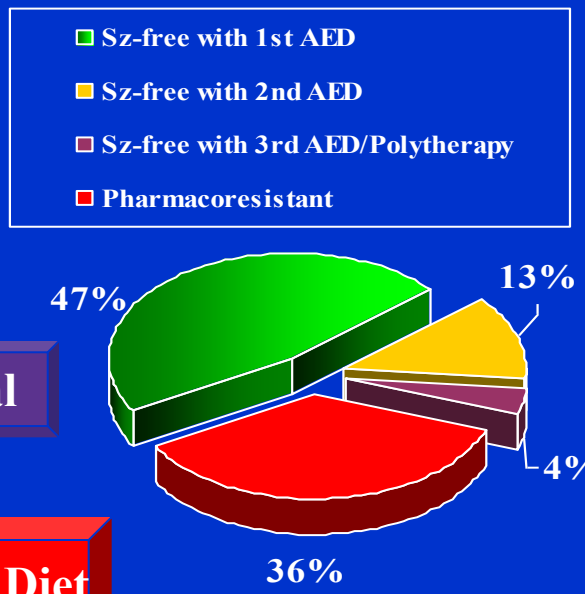
3rd Monotherapy/Polytherapy AED Trial

Epilepsy Surgery/VNS Therapy/Ketogenic Diet Evaluation with videoEEG

Resective Surgery

VNS Therapy

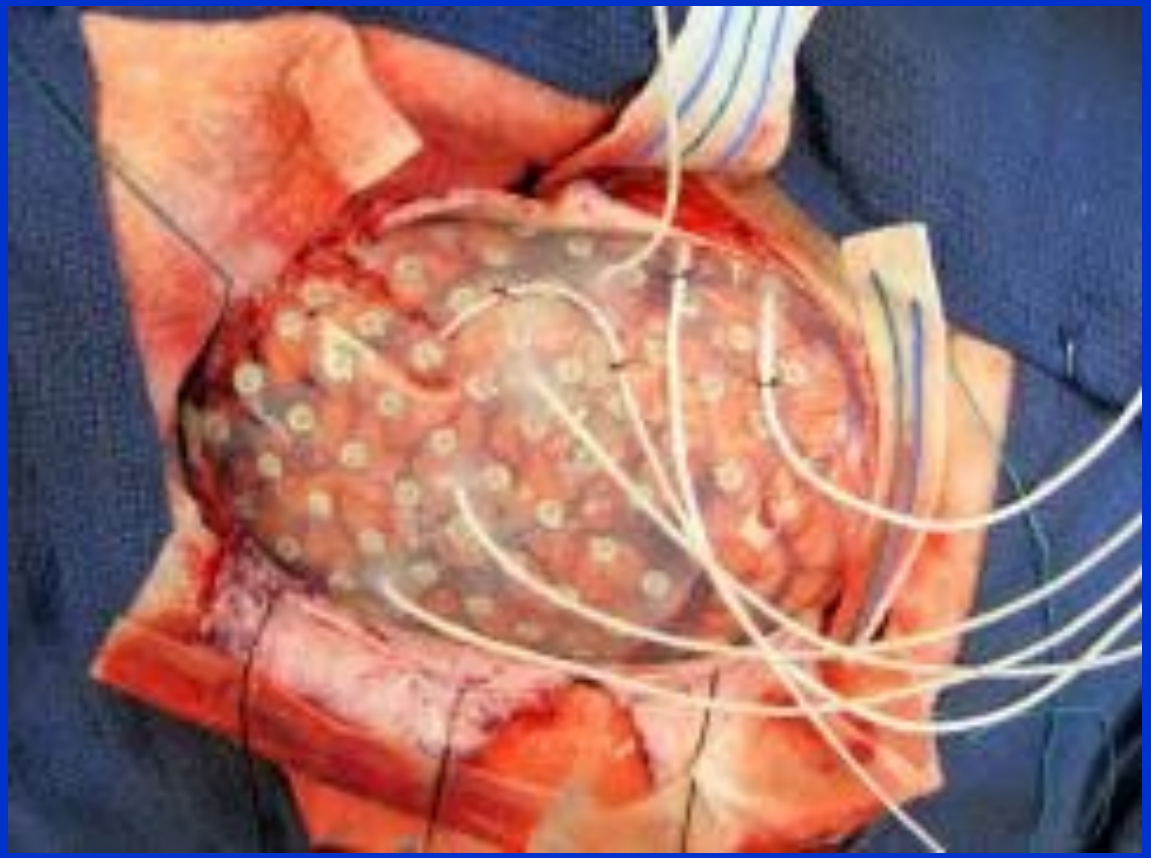
Polytherapy AED Trials




Kwan P, Brodie MJ. *NEJM*;342:314-319.

# Drug-resistant epilepsy treatment

- Surgery
- Nerve vagus stimulator
- Ketogenic diet



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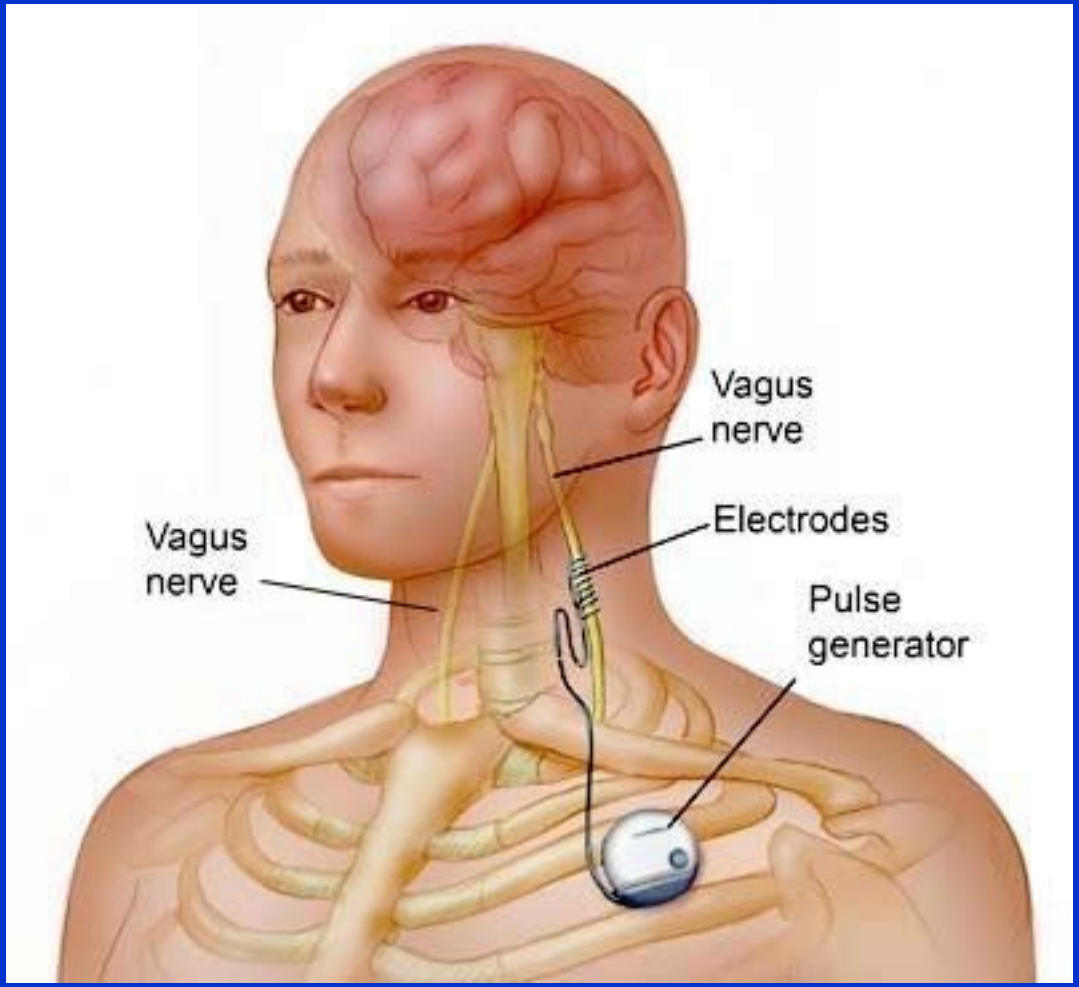

# Drug-resistant epilepsy treatment

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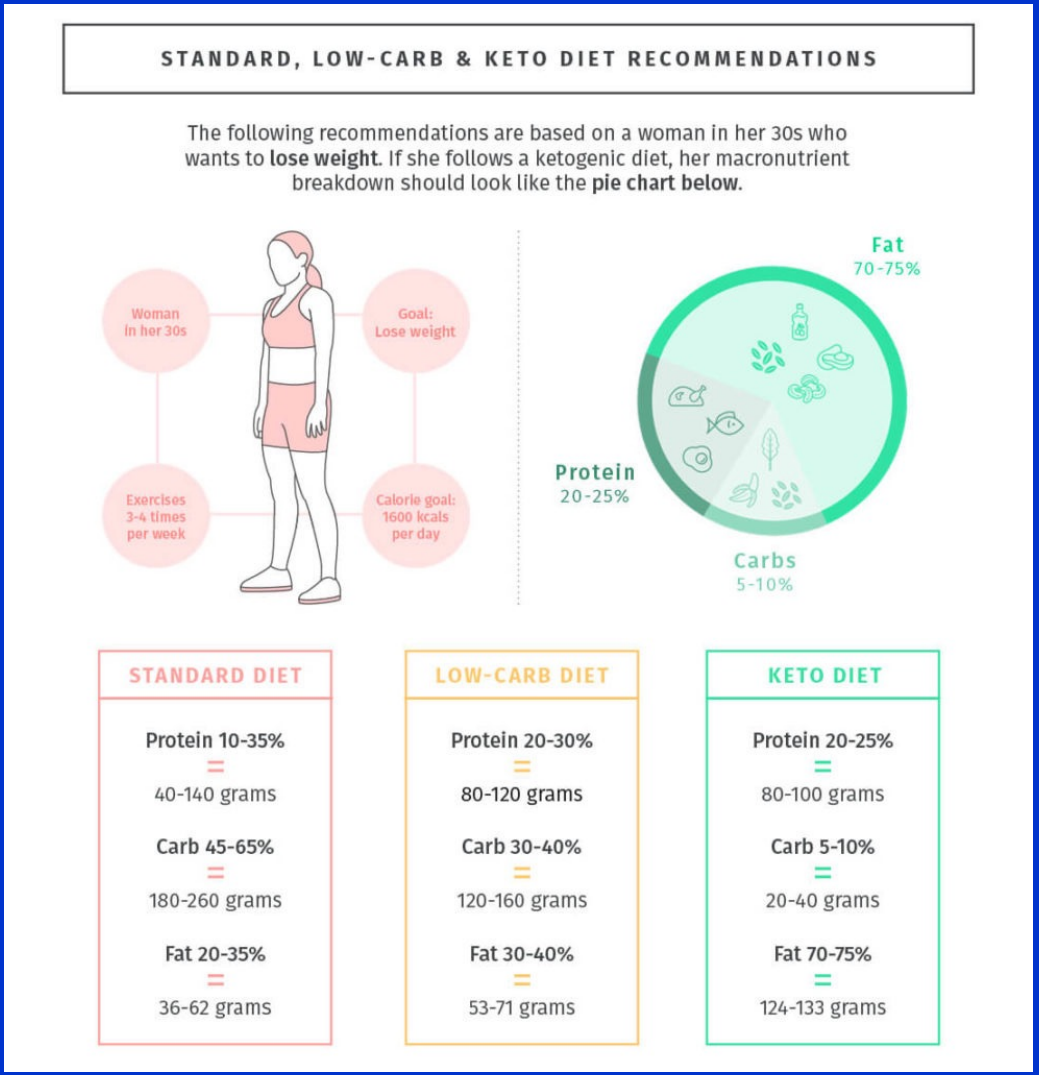

# Thank you