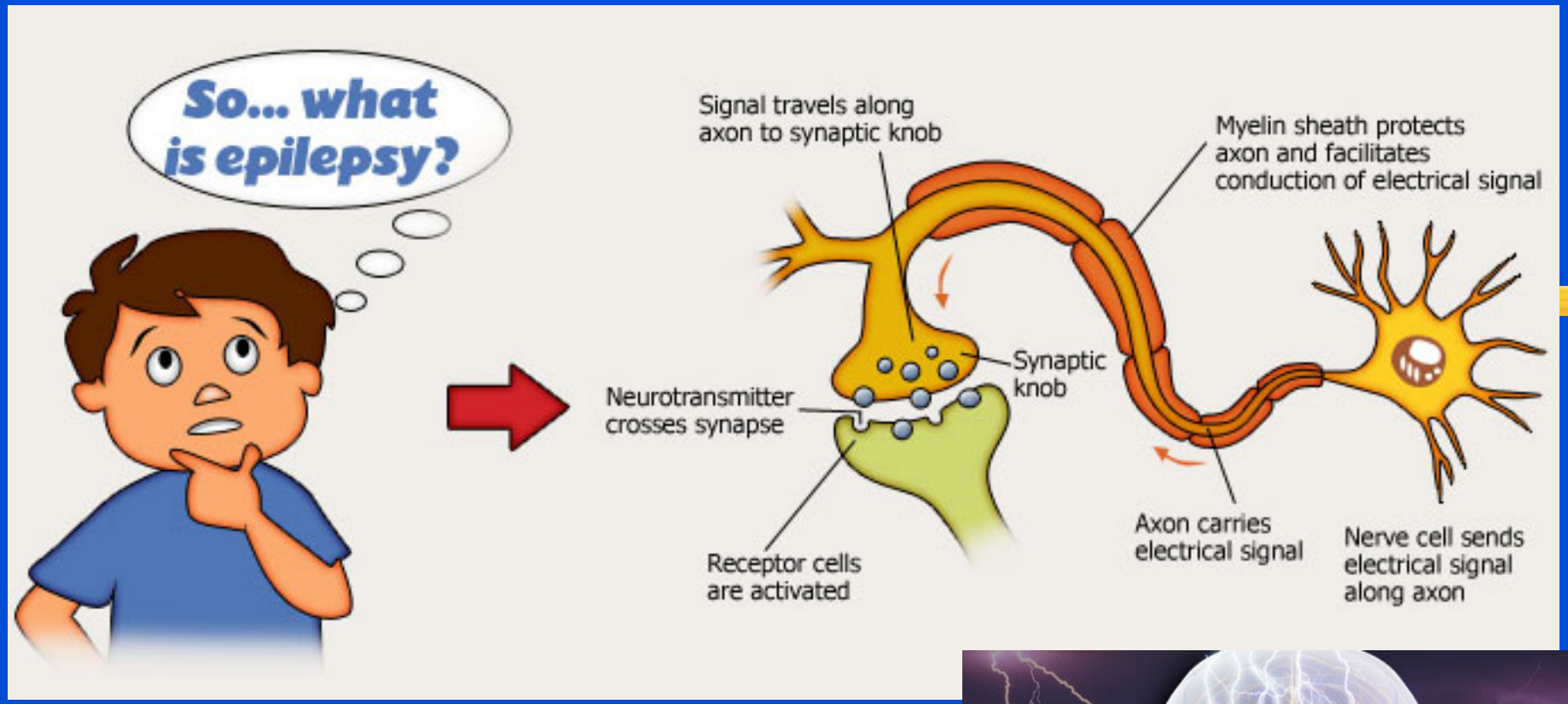
Epilepsy: diagnosis and treatment

Sergiusz Jóźwiak

Klinika Neurologii Dziecięcej WUM

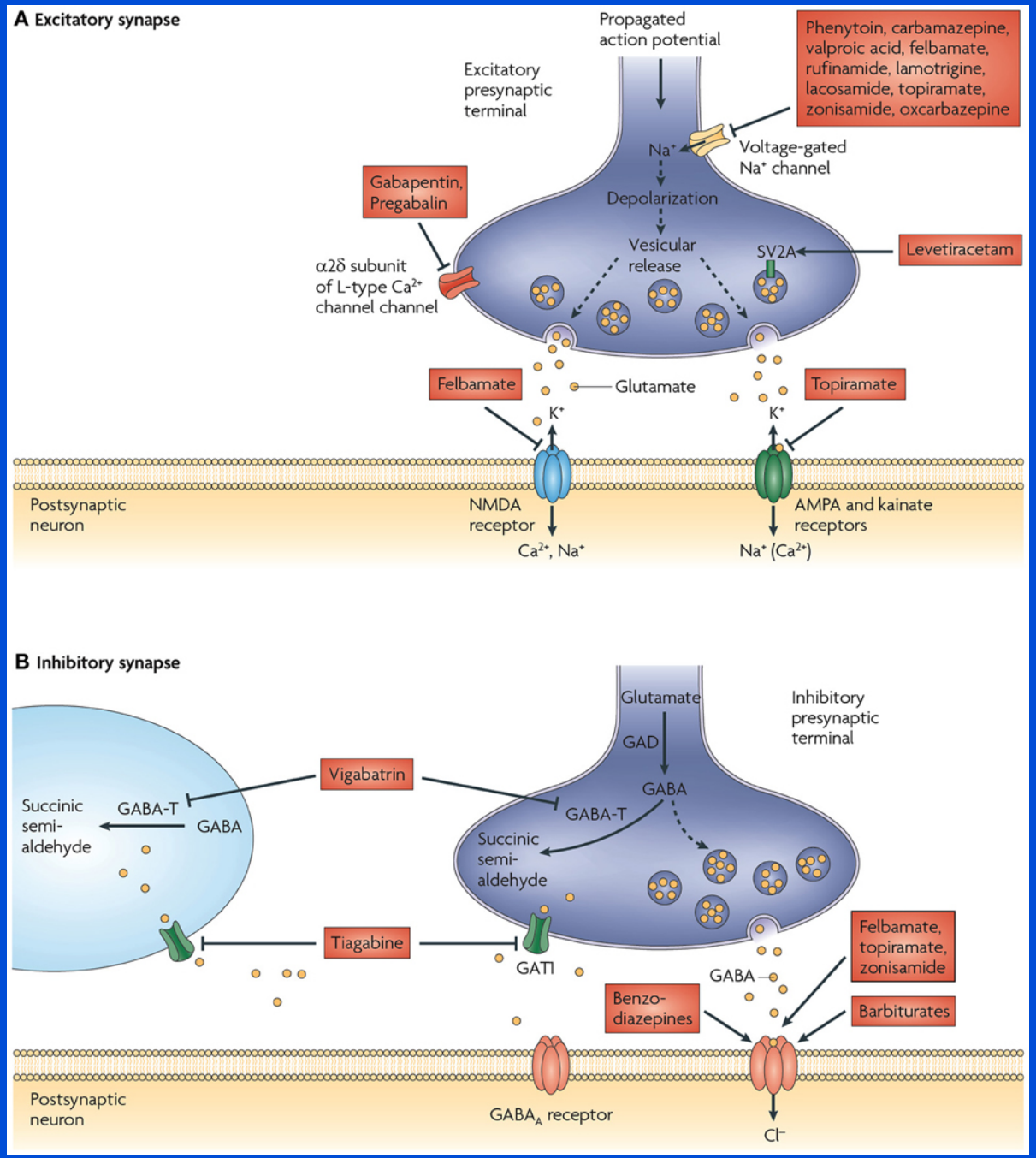


Definition:

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

Neurotransmitters:

GABA vs Glutamate



Seizure

What are Seizures?

- Clinical Definition of Seizures
  - "Paroxysmal episodes of brain dysfunction manifested by stereotyped alteration in behavior"
    - Clinical manifestations of a seizure based on anatomy of the brain that is seizing
      - Symptoms: sensory, motor, autonomic with or without loss of consciousness
  - Epilepsy is recurrent and unprovoked seizures

Operational definition of epilepsy

Operational (Practical) Clinical Definition of Epilepsy

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

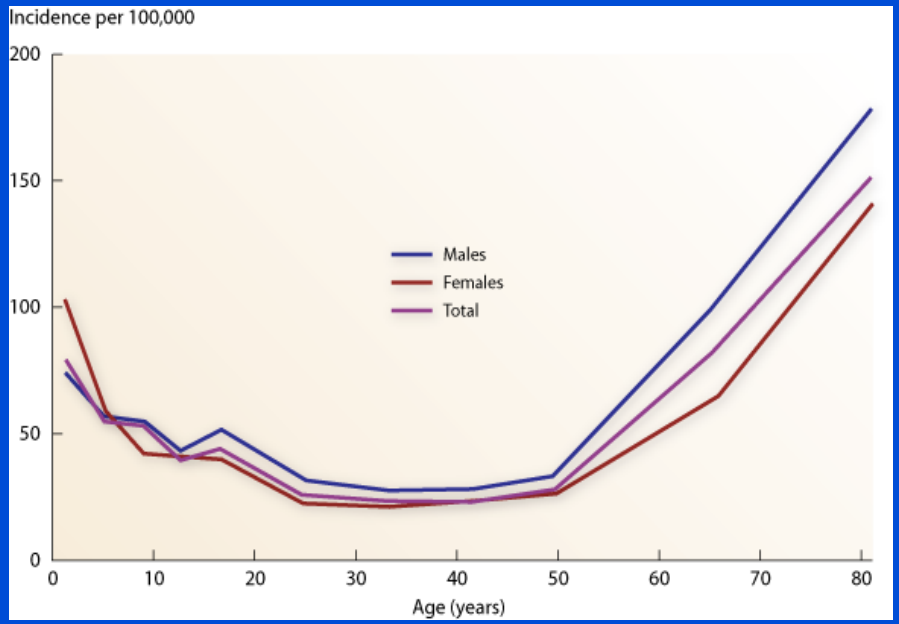
2. One unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years;

3. Diagnosis of an epilepsy syndrome.

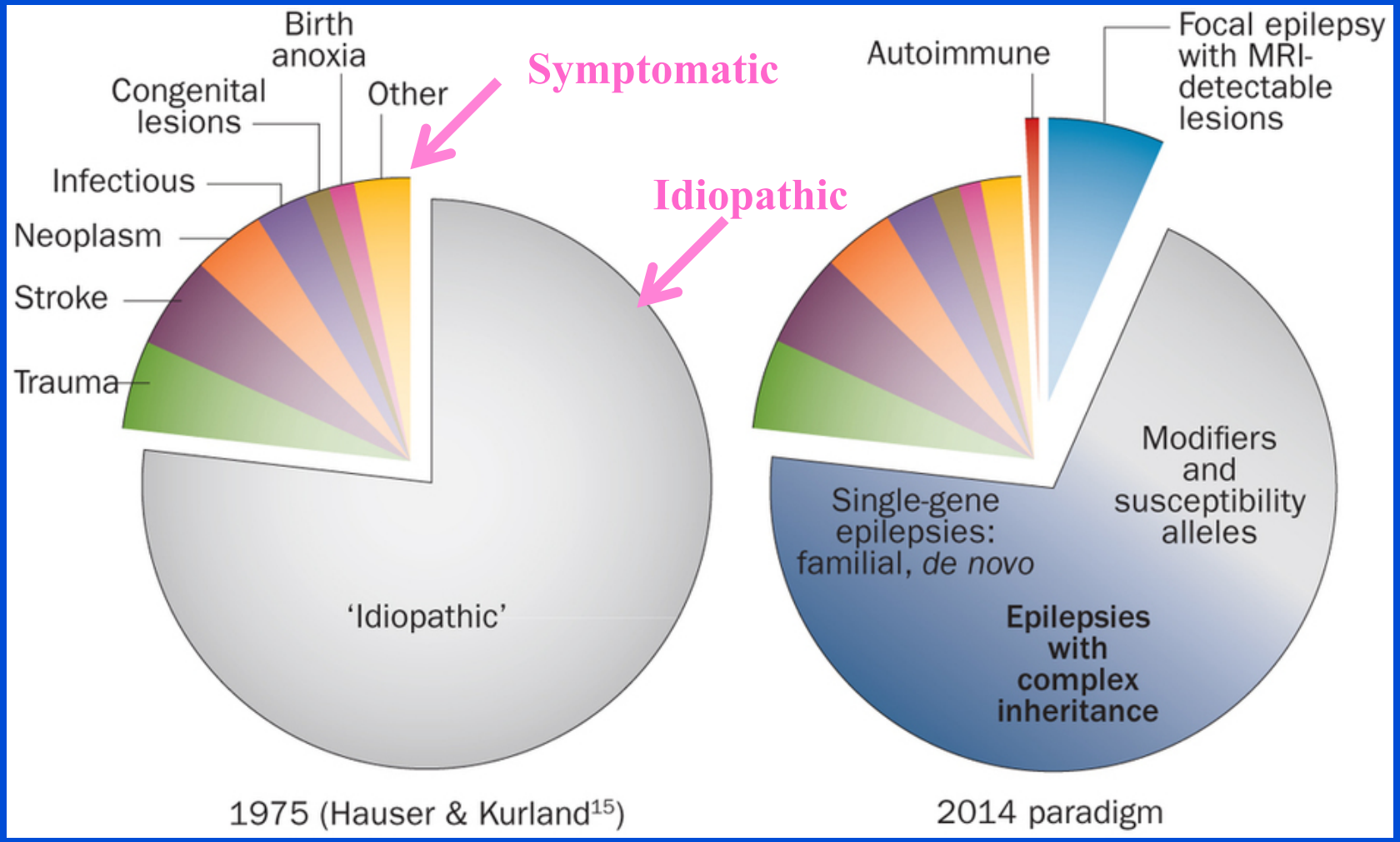
Epilepsy is considered to be resolved for individuals who had age-dependent epilepsy syndrome but are now past the applicable age or those who have remained seizure-free for the last 10 years, with no seizure medicines for the last 5 years.

Epilepsy: epidemiology

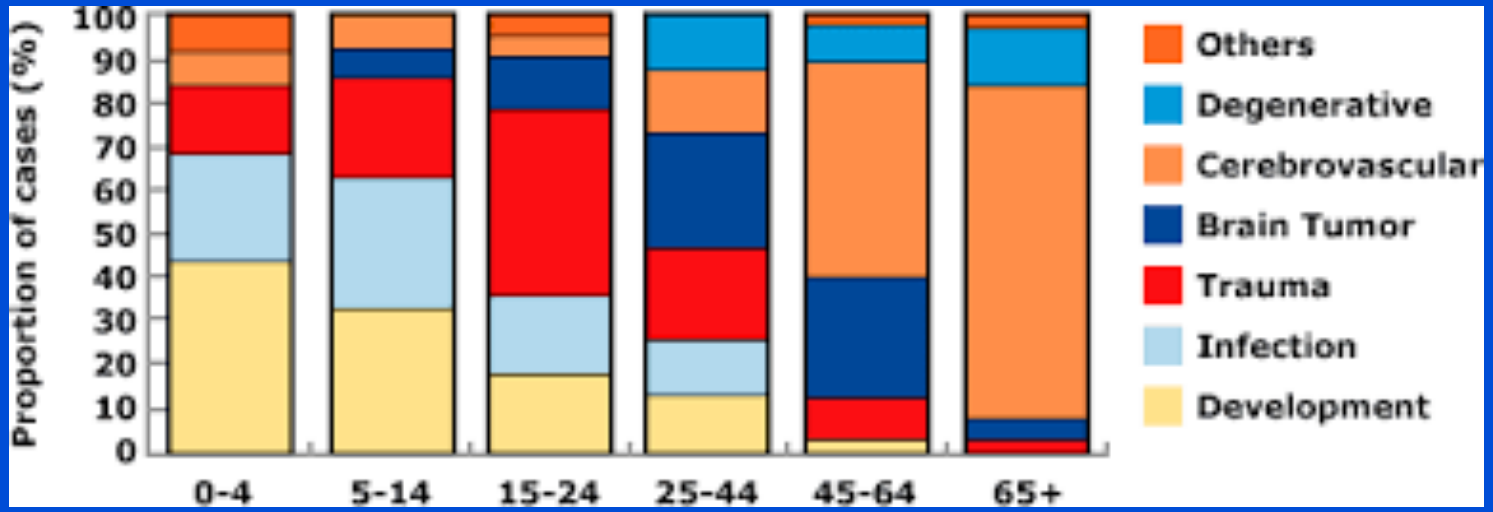
- Epilepsy affects 1-2% of the population
- Seizures including febrile seizures affect about 4-5% of pediatric population
- Lifetime prevalence: 9%
- Epilepsy refractory to AEDs: 20-30%



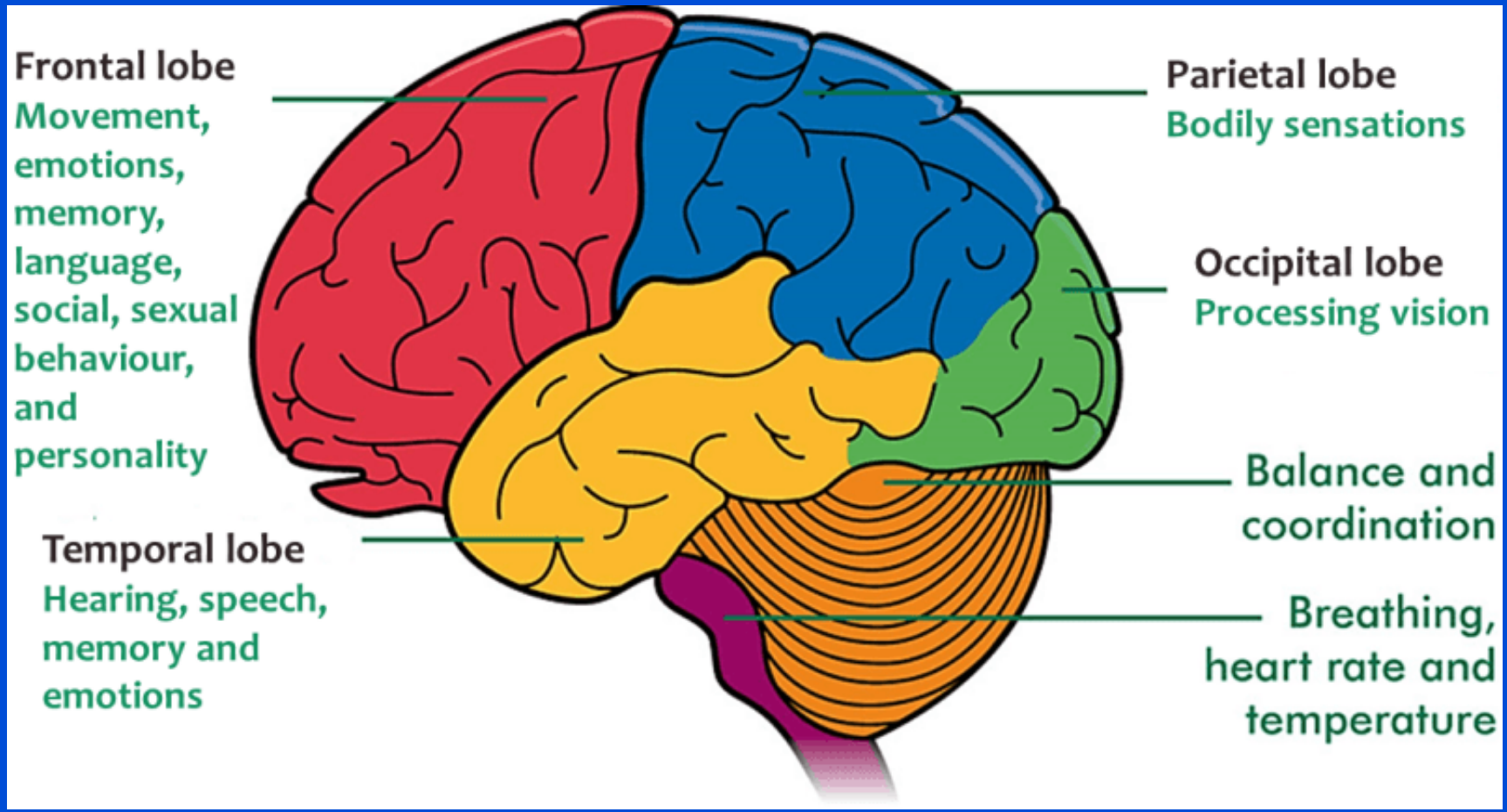
Epilepsy: etiology



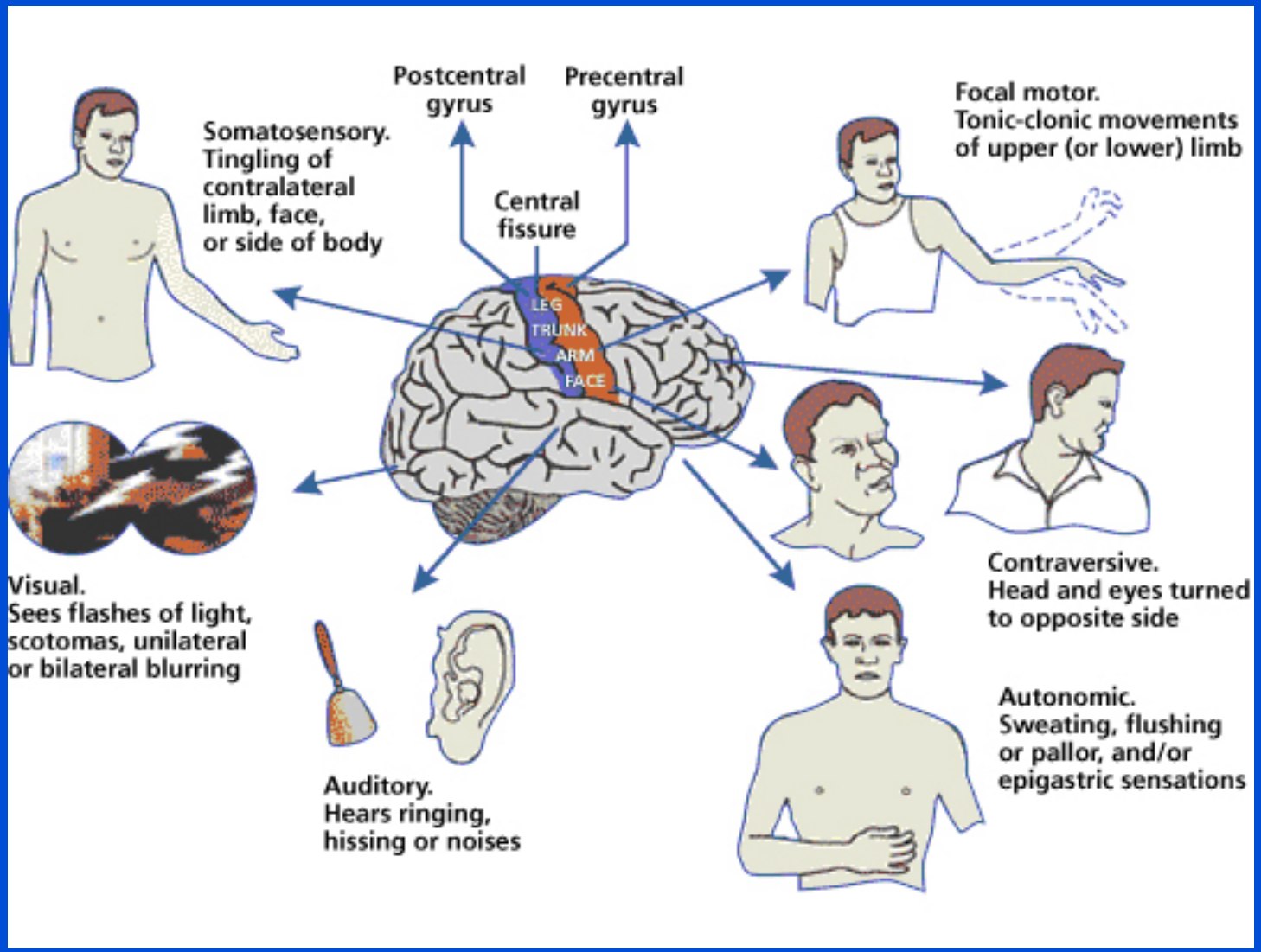
Epilepsy: etiology according to age



Functional organisation of the brain

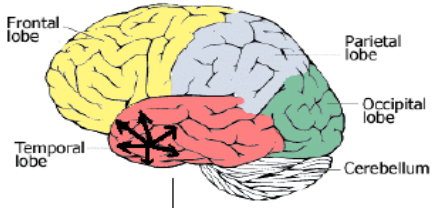


Location of locus and type of seizures

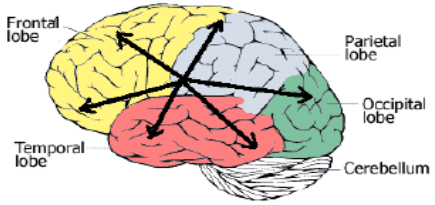


Seizure Classification

Partial
Seizure activity starts in one area of the brain



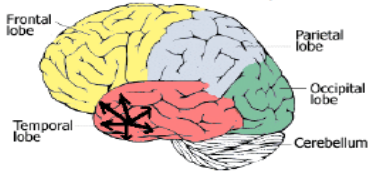
Generalized
Seizure involves whole brain & consciousness is affected



Simple
Retains awareness

Complex
Altered awareness and behavior

Secondary generalisation (spreading from one area to the whole brain)



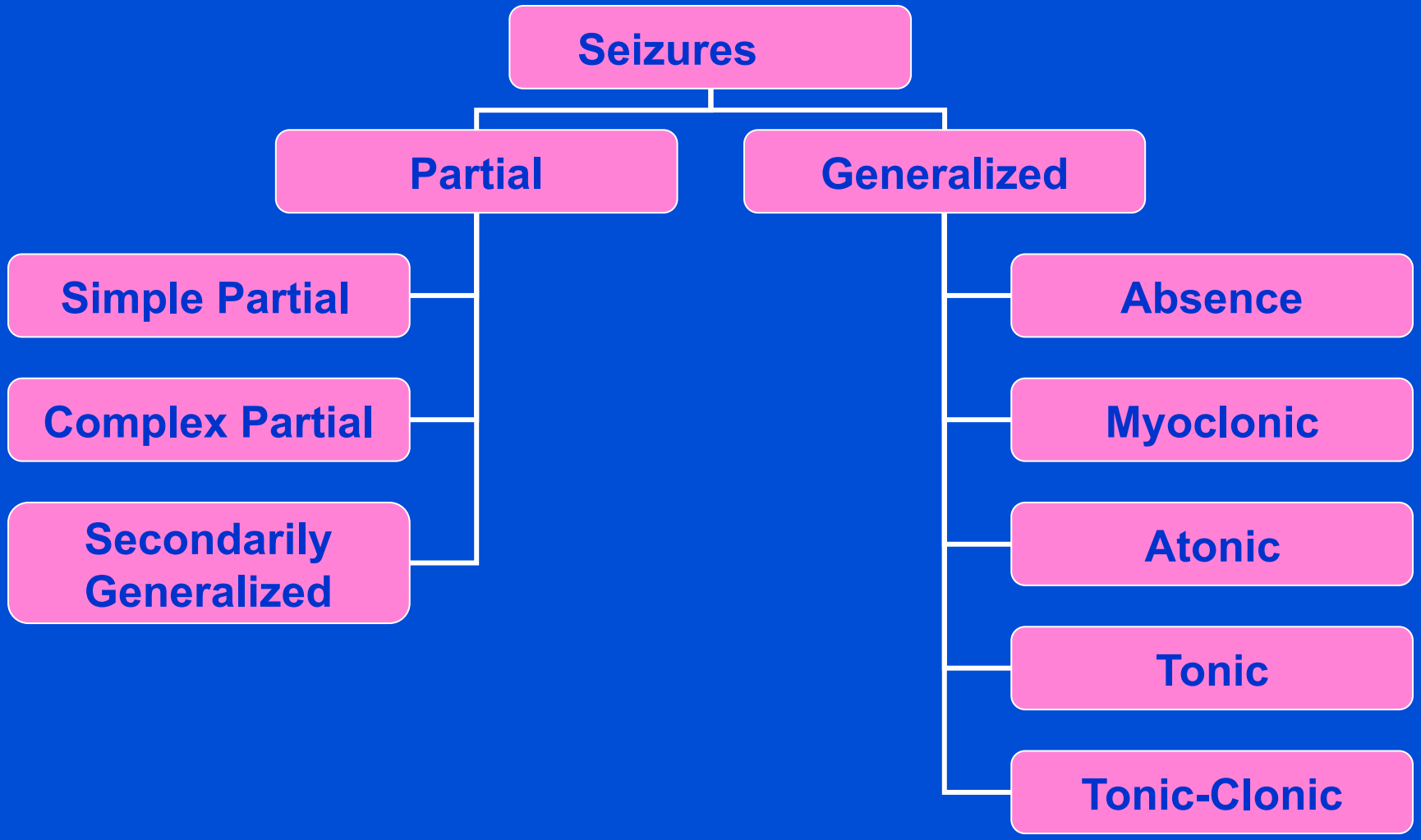
Tonic Clonic
"grand-mal" or convulsion
Loss of consciousness, stiffening of body then jerking of limbs

Absence
"petit mal" or staring fit or trance like state

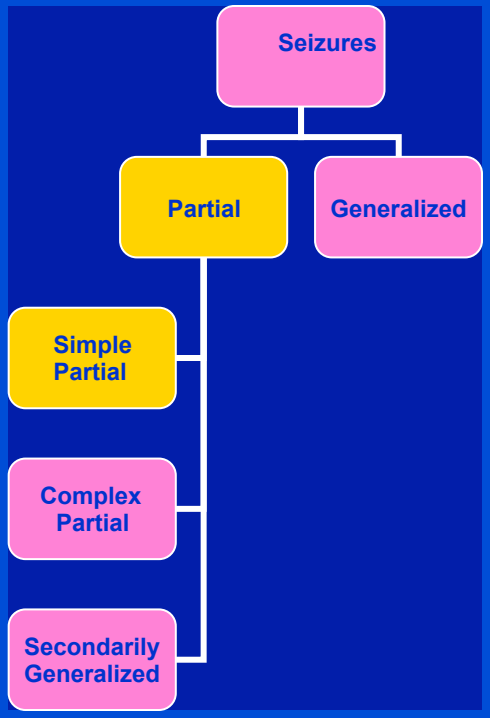
Tonic or Atonic
"drop attack"
Abrupt fall, either with stiffening (tonic) or with loss of muscle tone (atonic or "astatic" attacks)

Myoclonic
Sudden muscle jerks

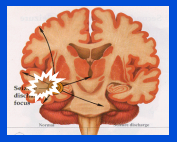
ILAE Classification of Seizures

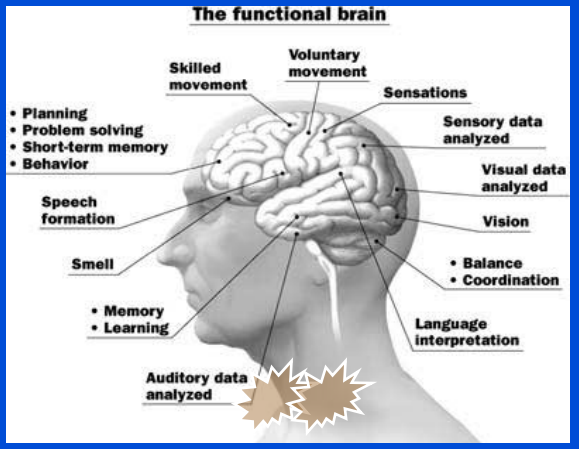


Partial (focal) Seizures



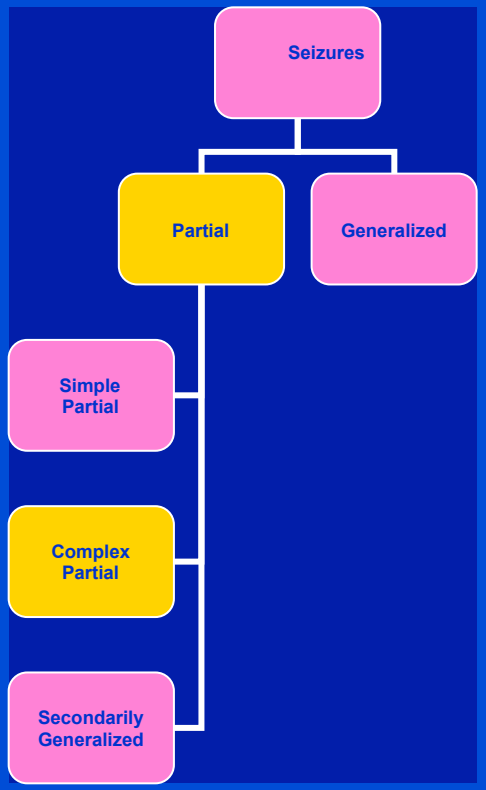
- **Simple Partial Seizure**
  - no loss of awareness
  - Auras
    - Temporal lobe:
      - Smell (uncus)
      - Epigastric sensation
      - déjà vu (hippocampus)
      - Fear/anxiety (amygdala)
    - Parietal lobe: Sensory
    - Occipital lobe: visual
  - Focal motor clonic movement



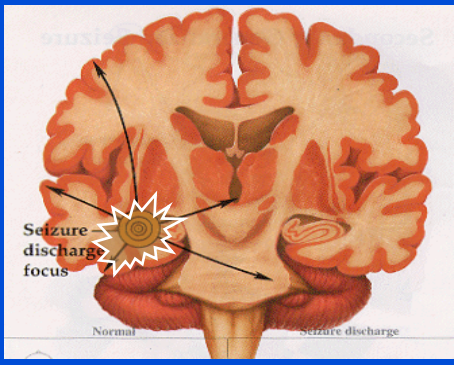


- **Supplementary Motor Seizure**
  - dystonic posturing
    - upper extremities (fencing)
    - lower extremities
  - Bicycling
  - Short duration 10-30 sec

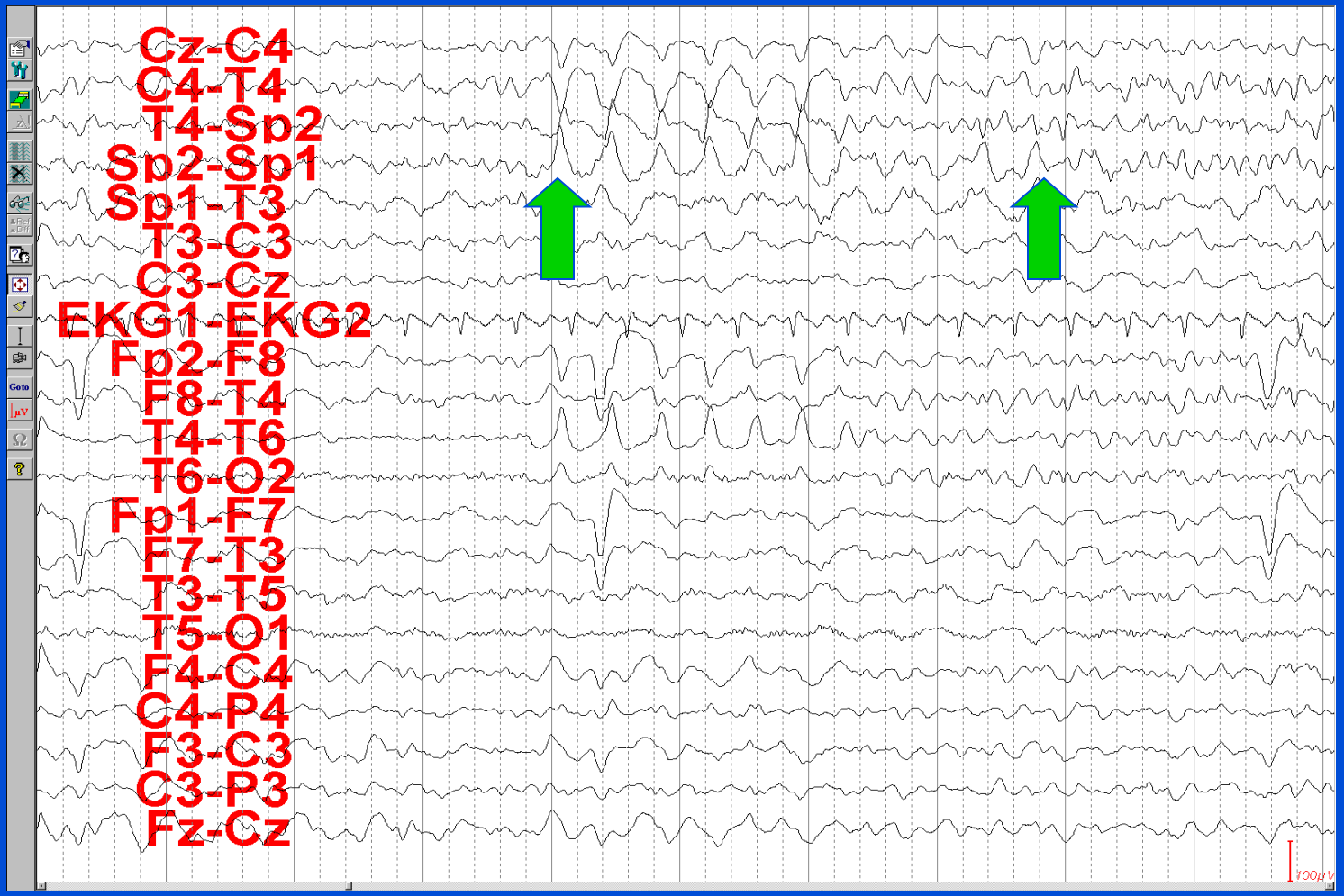
Partial (focal) Seizures



- **Complex Partial Seizure**
  - Impaired consciousness/ level of awareness (staring)
  - Clinical manifestations vary with origin & degree of spread
  - Presence and nature of aura
    - Temporal lobe: smell, epigastric sensation, deja vu
  - Automatisms (manual, oral)
  - Other motor activity
    - Frontal: bicycling and fencing posture
  - Duration (typically 30 seconds to 3 minutes)
  - Amnesia for event and confusion often after event



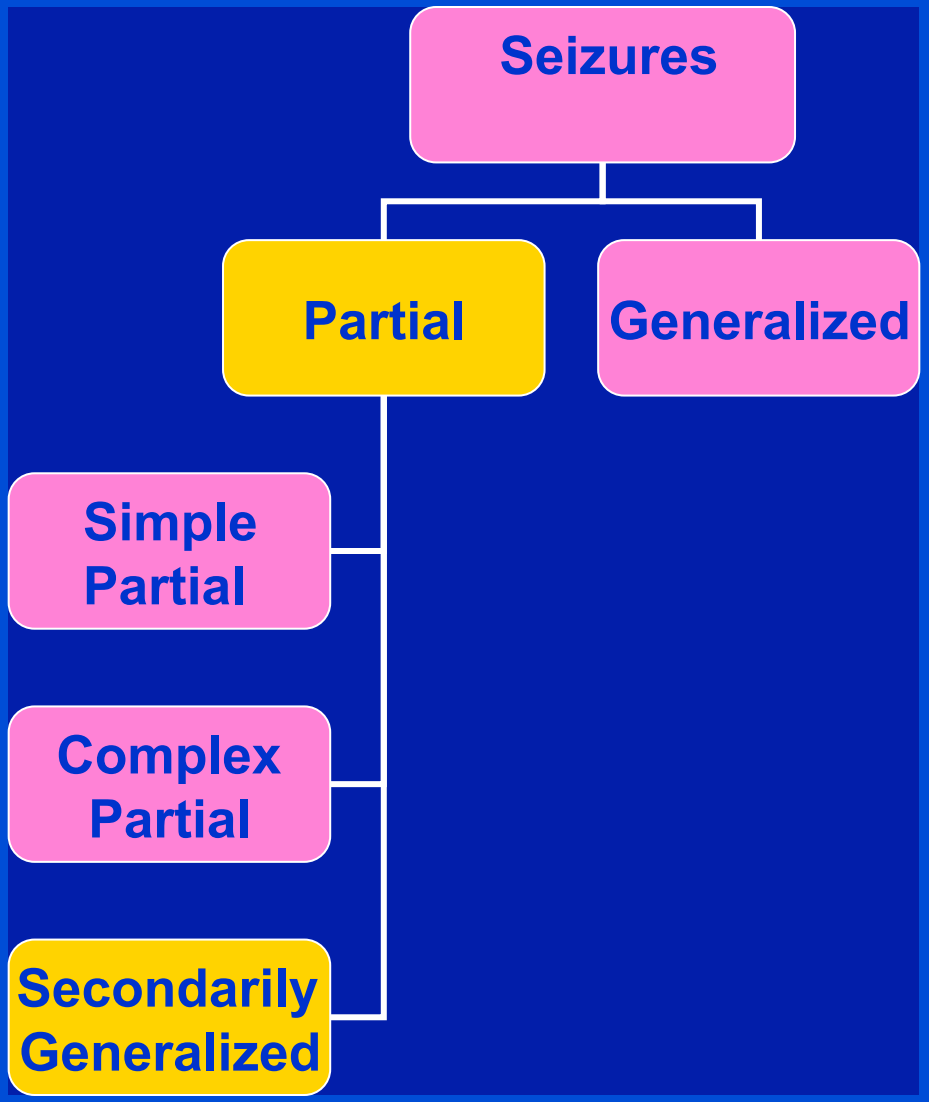
EEG: Partial Seizure



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

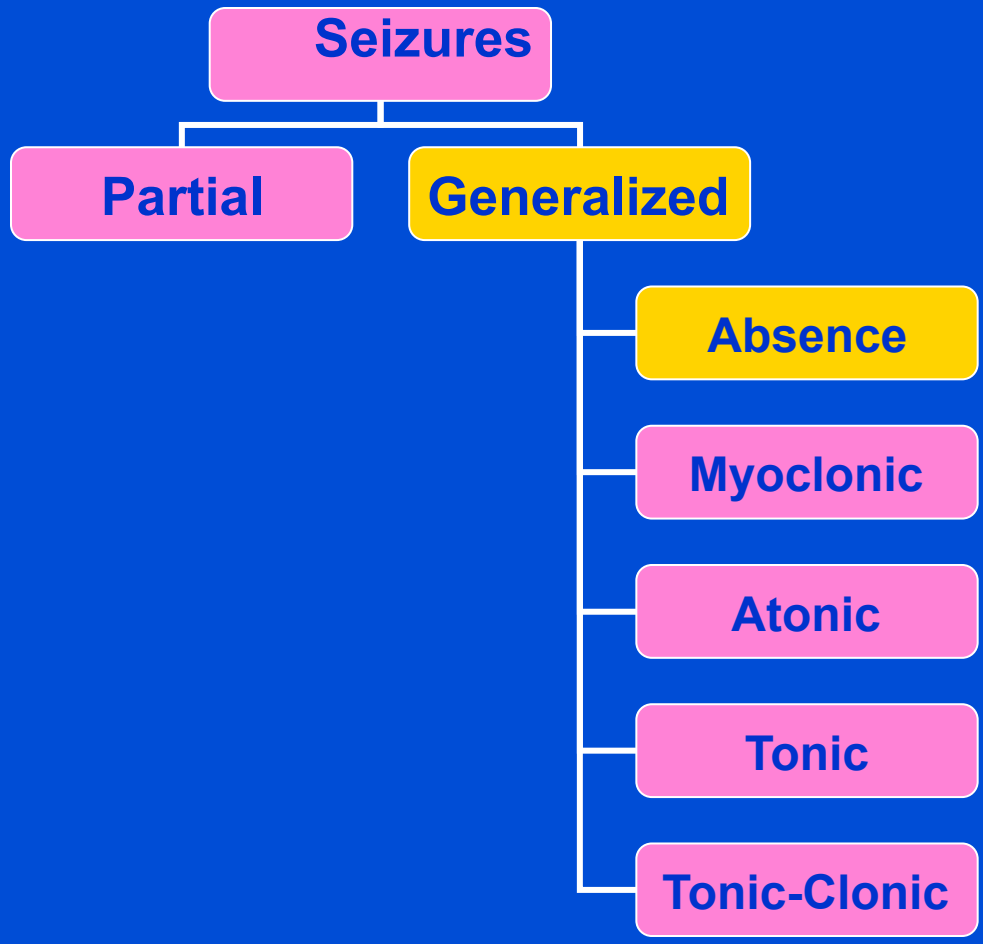
Secondarily Generalized Seizures

- 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

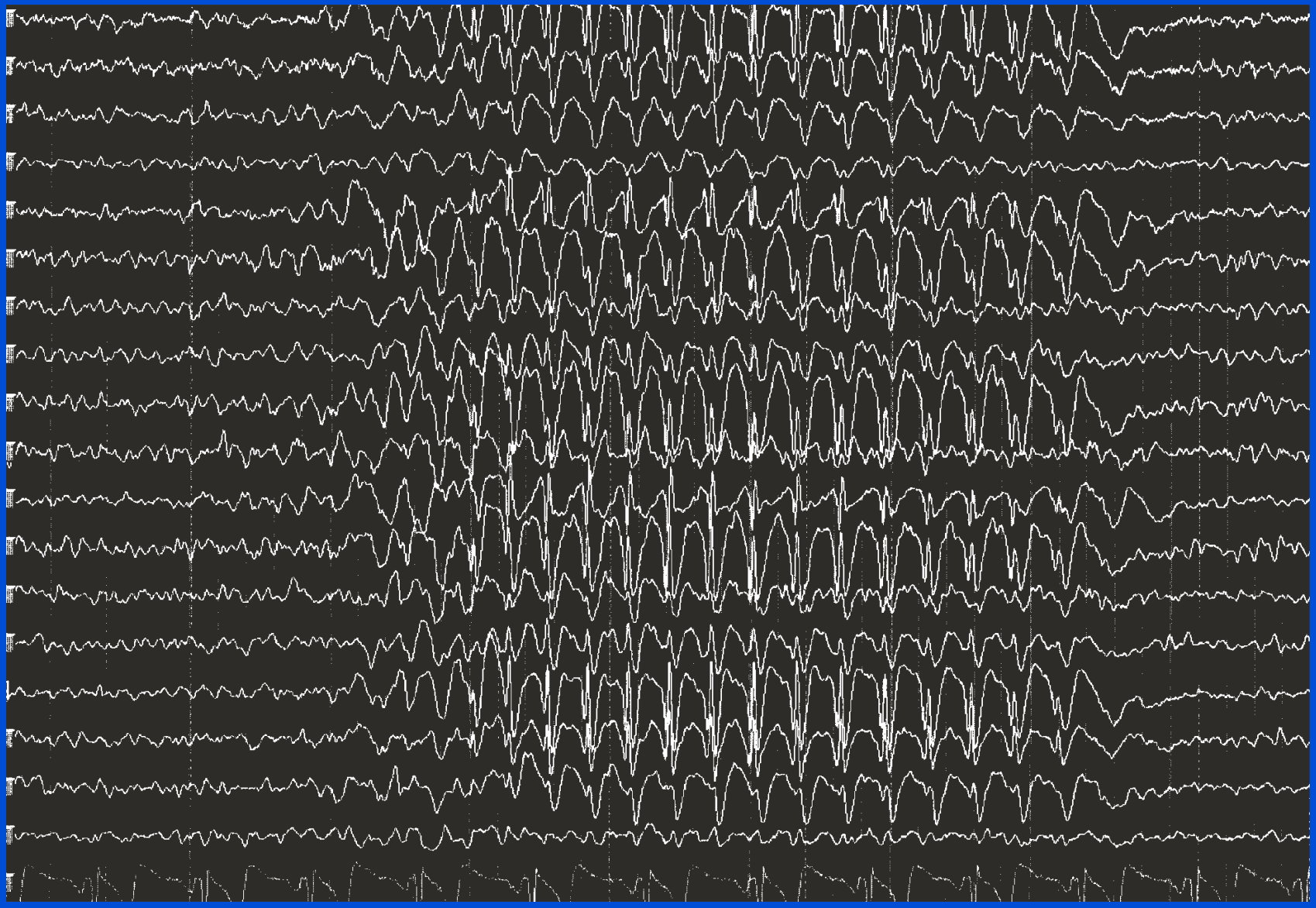


Childhood Absence Seizures

- 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

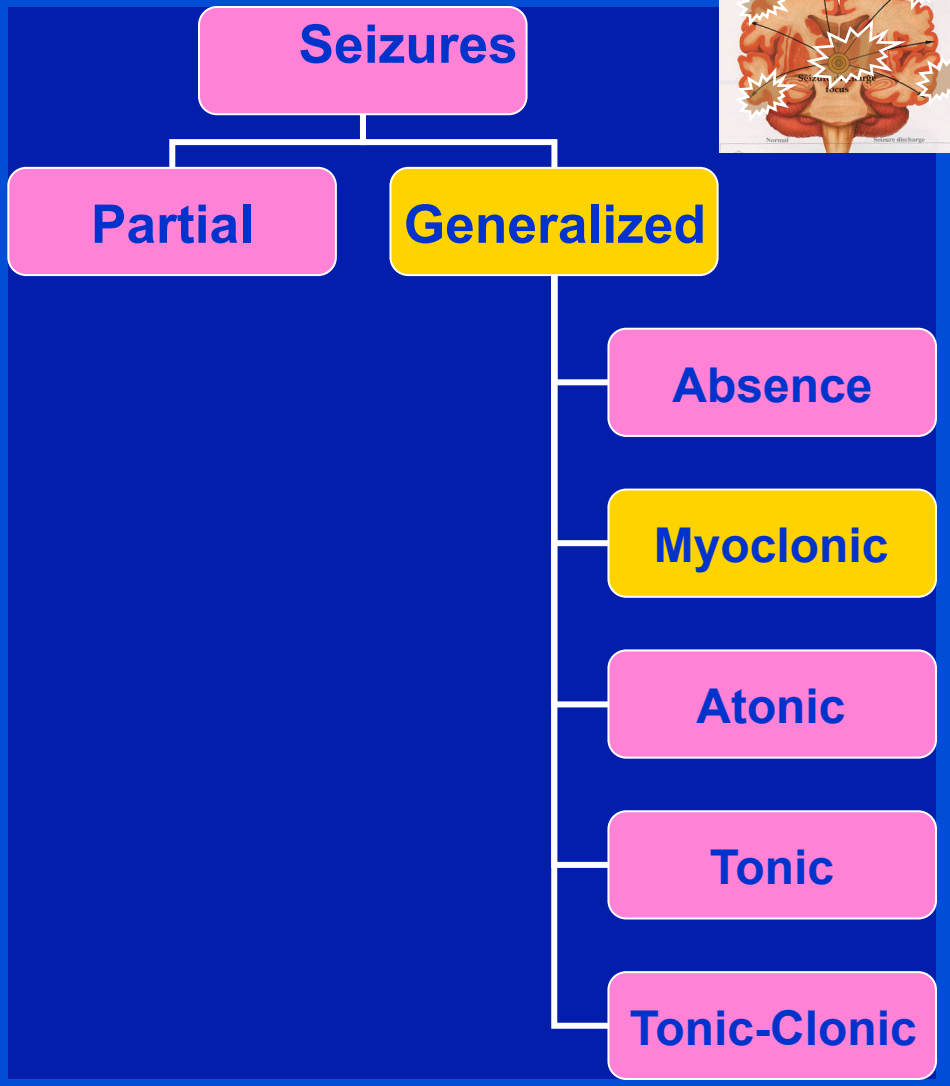


Juvenile Absence Seizures

- 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

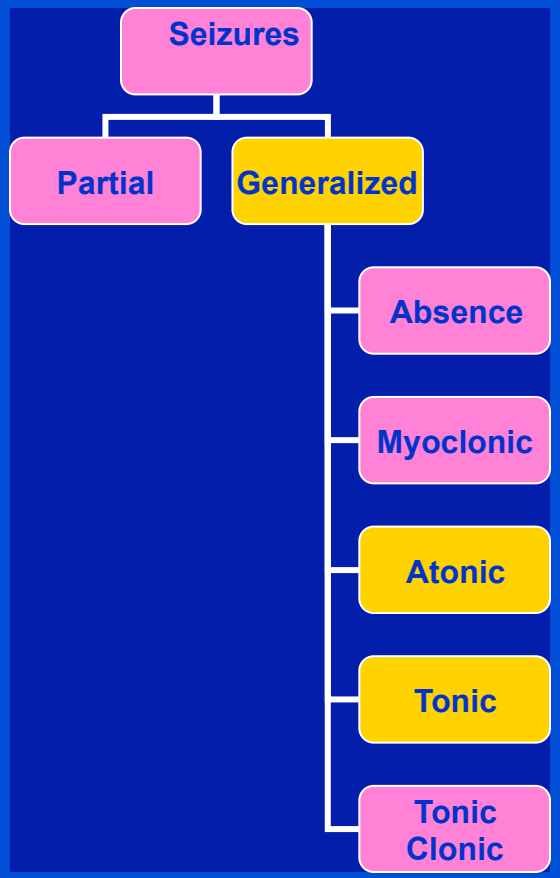
Myoclonic Seizures

- Brief, shock-like jerk of a muscle or group of muscles
- Epileptic myoclonus
  - Typically bilaterally synchronous
  - Impairment of consciousness 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

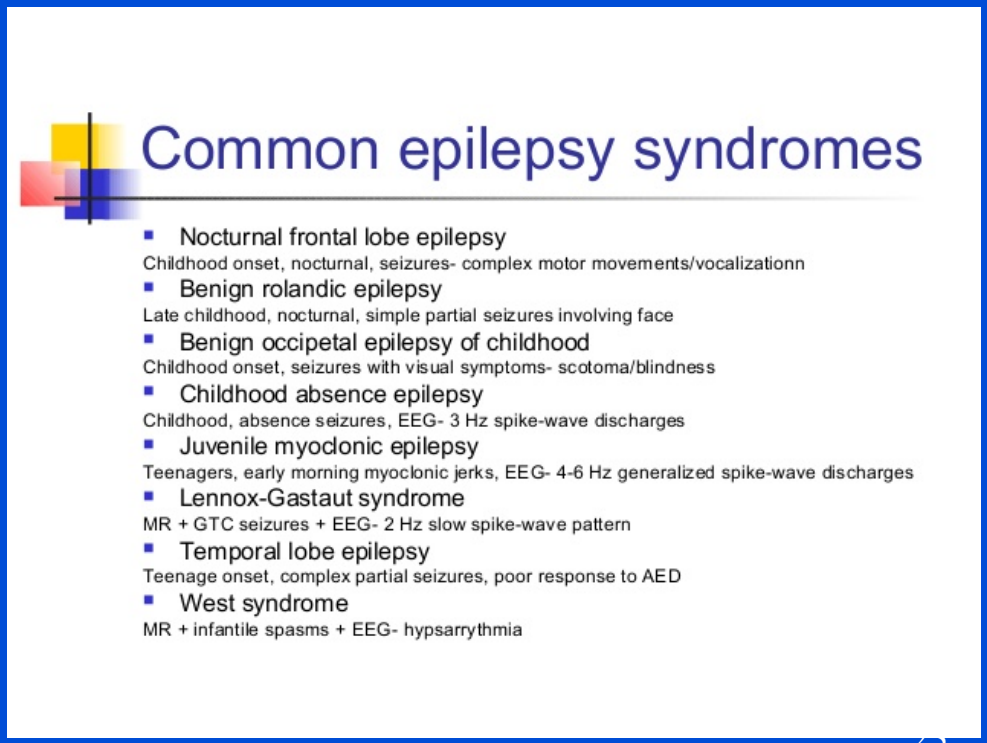
- Sudden loss of postural tone
  - When severe often results in falls
  - When milder produces head nods or jaw drops.
- Consciousness usually impaired
- Duration - usually seconds, rarely more than 1 minute
- EEG – sudden diffuse attenuation or generalized polyspike-wave

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

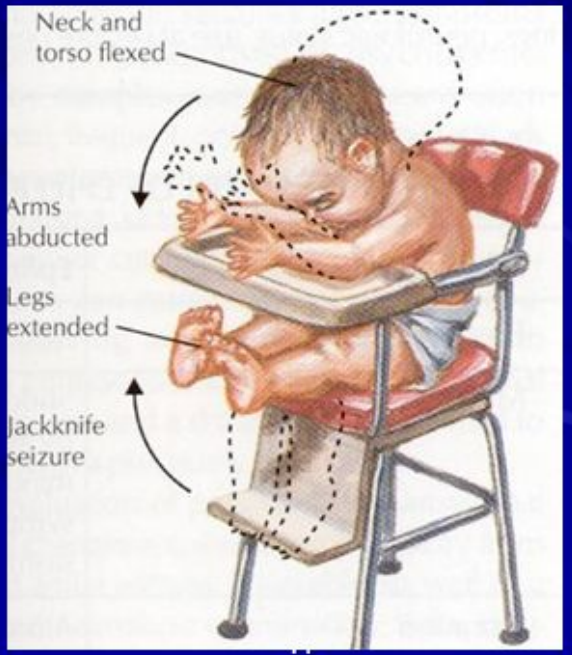
ON A PECULIAR FORM OF INFANTILE CONVULSIONS.

To the Editor of THE LANCET.

W. J. WEST.

Tunbridge, Jan. 26, 1841.

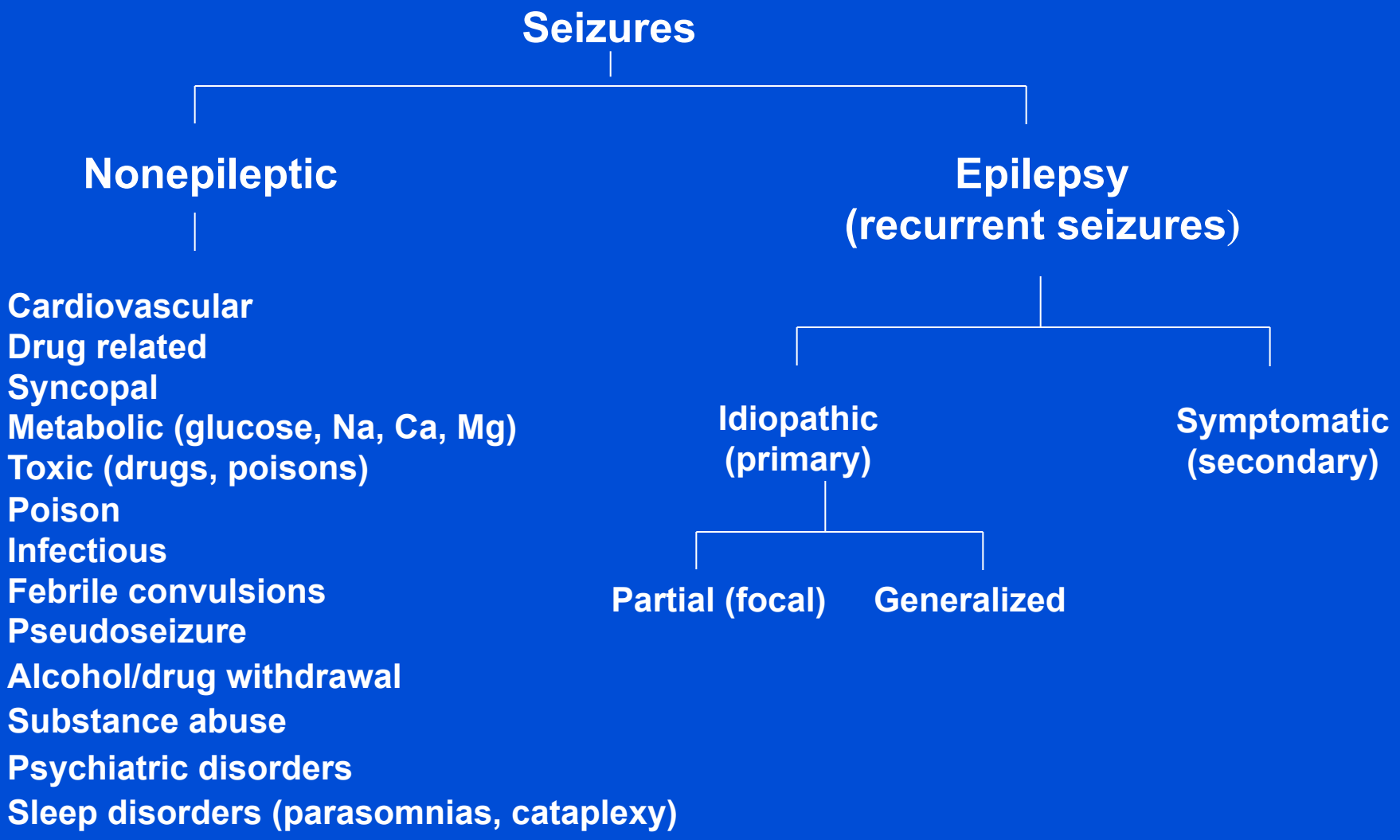
P.S.—In my own child's case, the bowing convulsions continued every day, without intermission, for seven months; he had then an interval of three days free; but, on the fourth day, the convulsions returned, with this difference, instead of bowing, he stretched out his arms, looked wild, seem to lose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



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



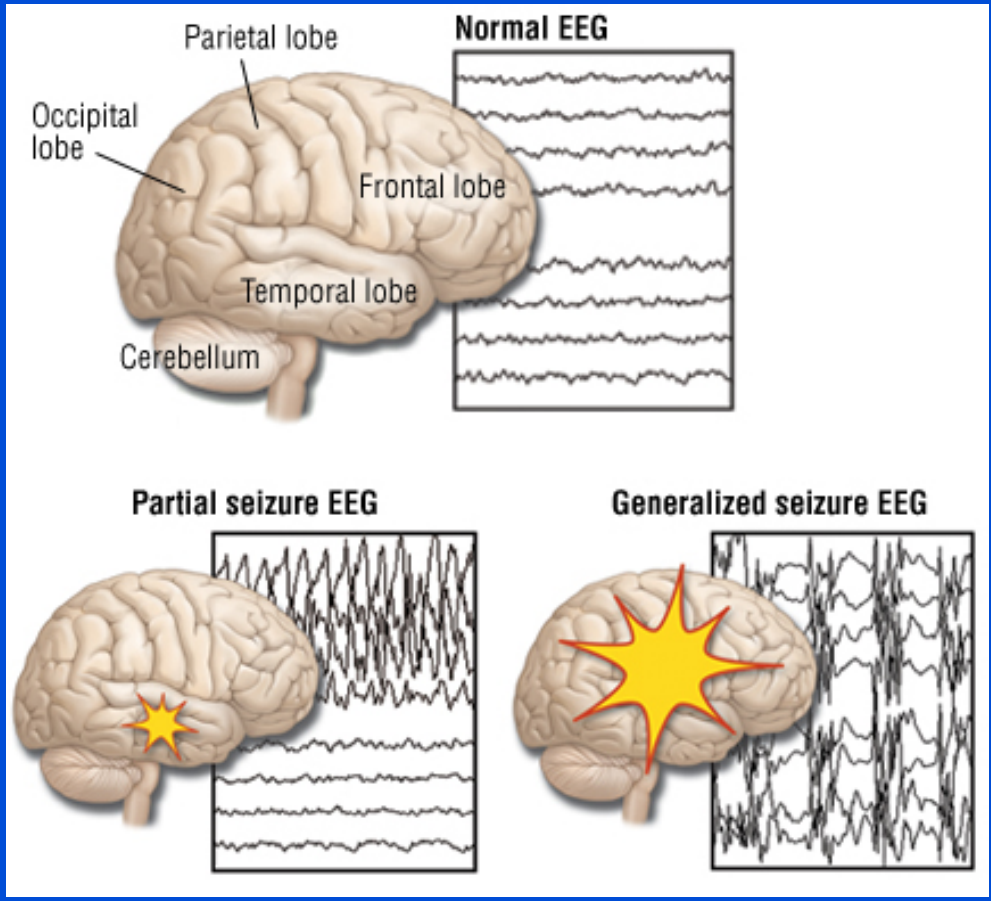
Psychogenic/Non-epileptic Events

- 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

Resources for the Diagnosis of Epilepsy

EEG (ElectroEncephaloGraphy)

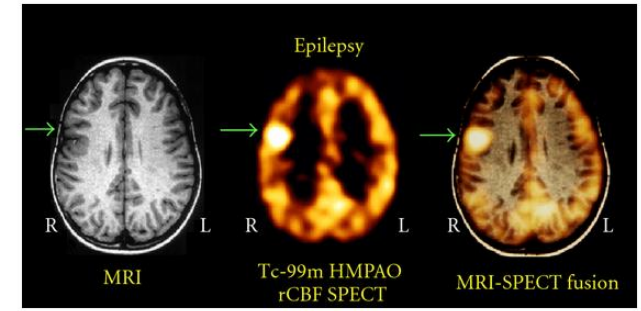
Electroencephalography is a measurement of the electrical activity of the brain by recording from electrodes placed on the scalp. Includes video EEGs and sleep EEG.



Resources for the Diagnosis of Epilepsy

- **MRI (Magnetic Resonance Imaging)**

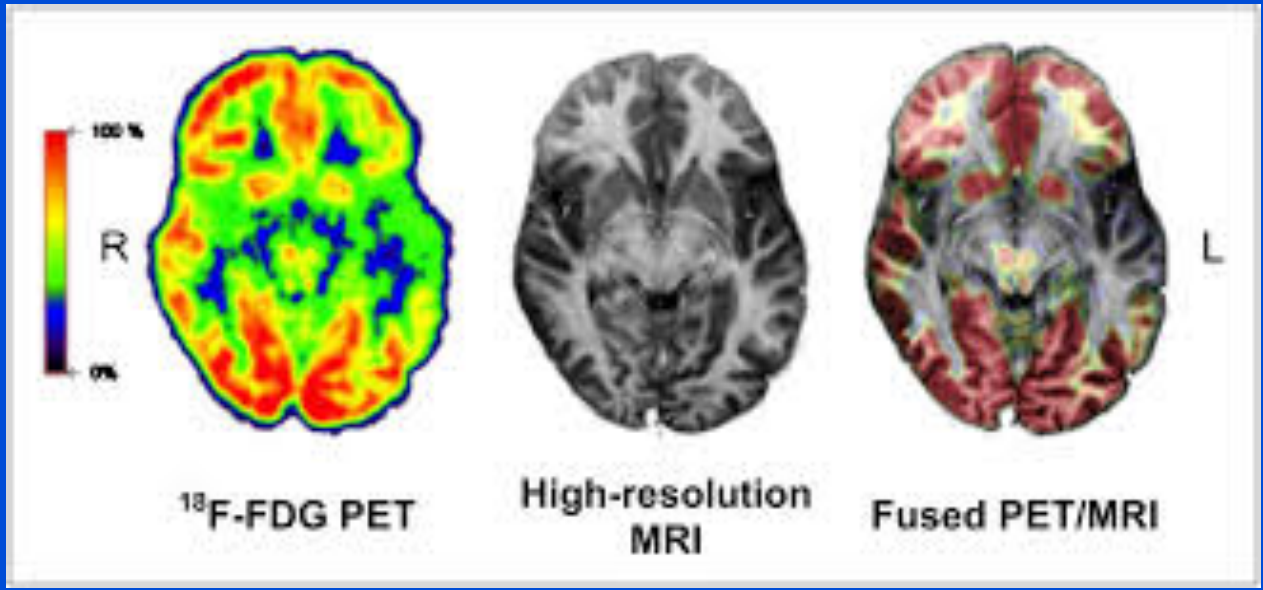
A method of creating images of the structure and contents of the brain using a powerful, uniform magnetic field.



Resources for the Diagnosis of Epilepsy

- **PET (Positron Emission Tomography)**

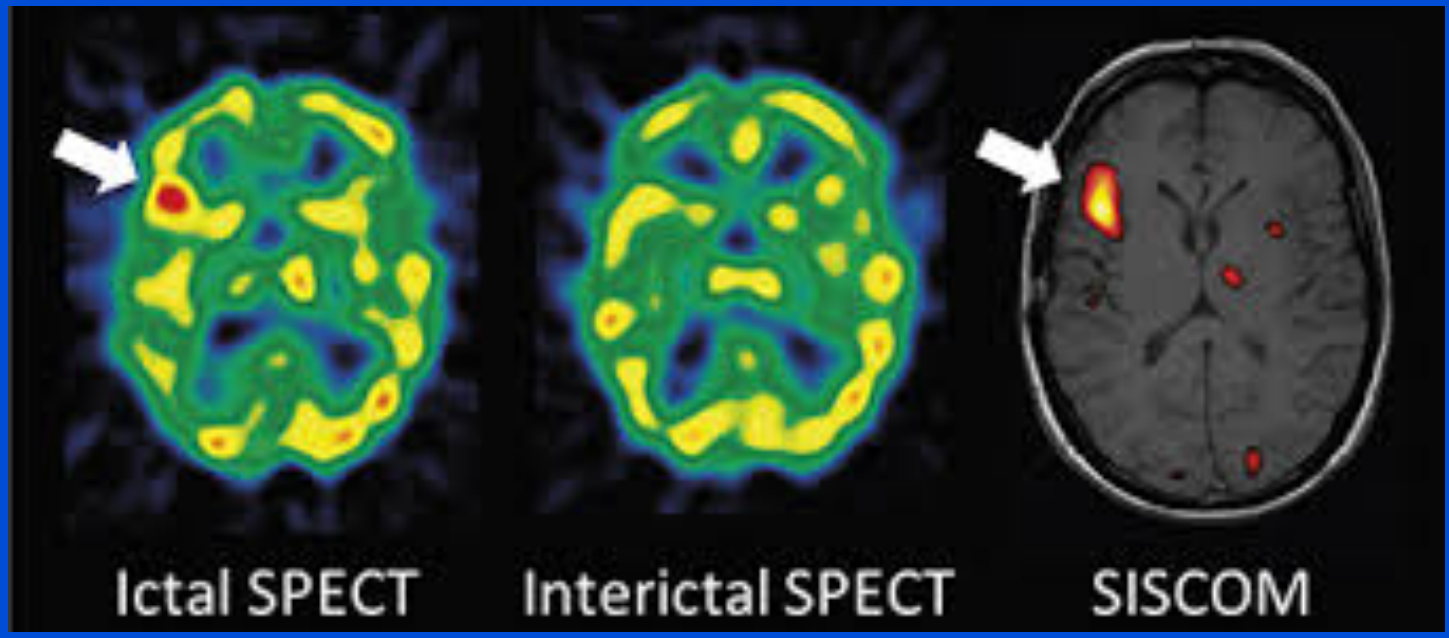
An advanced imaging technique that involves the acquisition of images of the brain based on the detection of radiation from the emission of positrons. Positrons are tiny particles emitted from a radioactive substance administered to the patient.



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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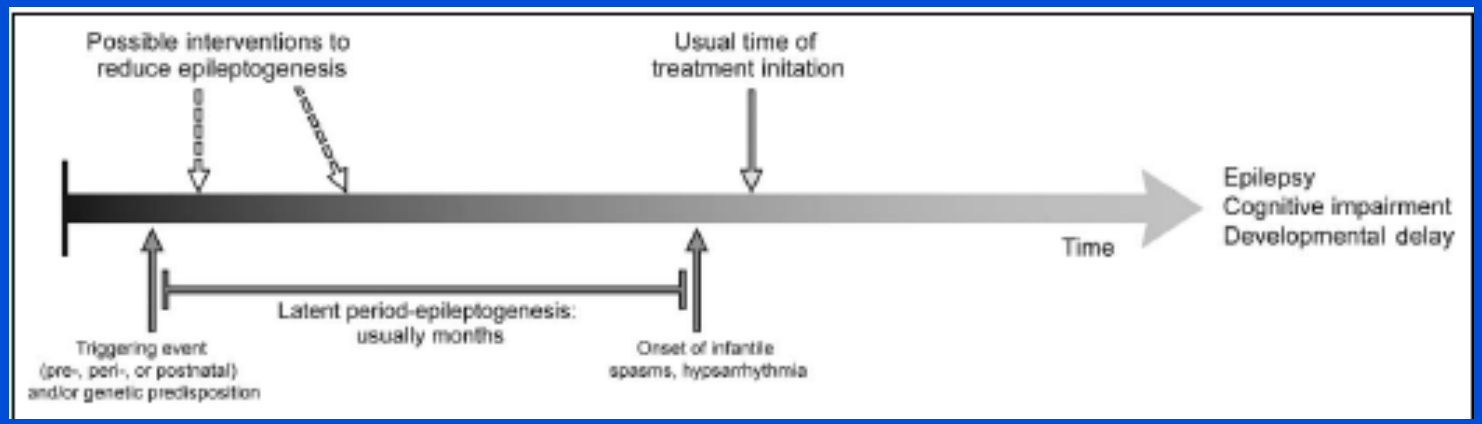
Carl E. Stafstrom, MD, PhD[1], Barry G. W. Arnason, MD[2], Tallie Z. Baram, MD, PhD[3], Anna Catania, MD[4], Miguel A. Cortez, MD[5], Tracy A. Glauser, MD[6], Michael R. Pranzatelli, MD[7], Raili Riikonen, MD, PhD[8], Michael A. Rogawski, MD, PhD[9], Shlomo Shinnar, MD, PhD[10], and John W. Swann, PhD[11]



Antiepileptogenic treatment !

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

Treatment Sequence for Pharmacoresistent Epilepsy

1st Monotherapy AED Trial

2nd Monotherapy AED Trial

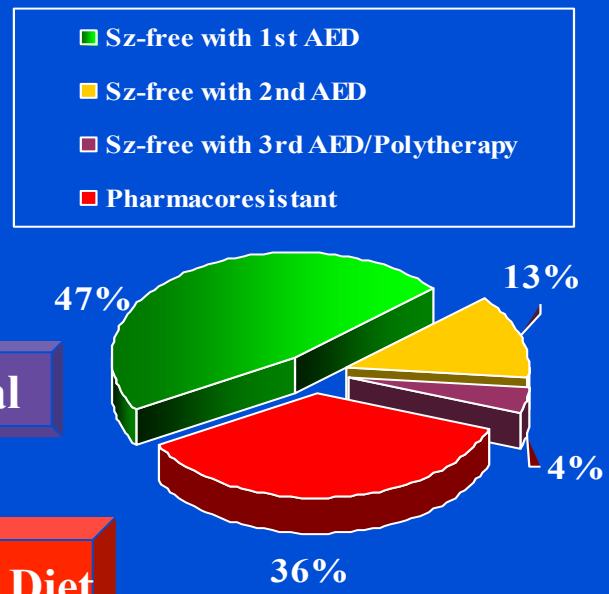
3rd Monotherapy/Polytherapy AED Trial

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

Thank you