EPILEPSY IN CHILDREN

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WORLD FEDERATION
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Objectives

At the end of this lecture, the attendee should be able to

1. Define and classify seizures and epilepsies
2. Outline the causes of epilepsy with examples
3. Describe common epilepsy syndromes in infants, children and adolescents
4. Outline the general approach to the child with epilepsy

Definition: Seizure

- The term seizure is vaguely used to refer to anything that "seizes" or "takes hold" of a person.

- **An epileptic seizure is a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain.**

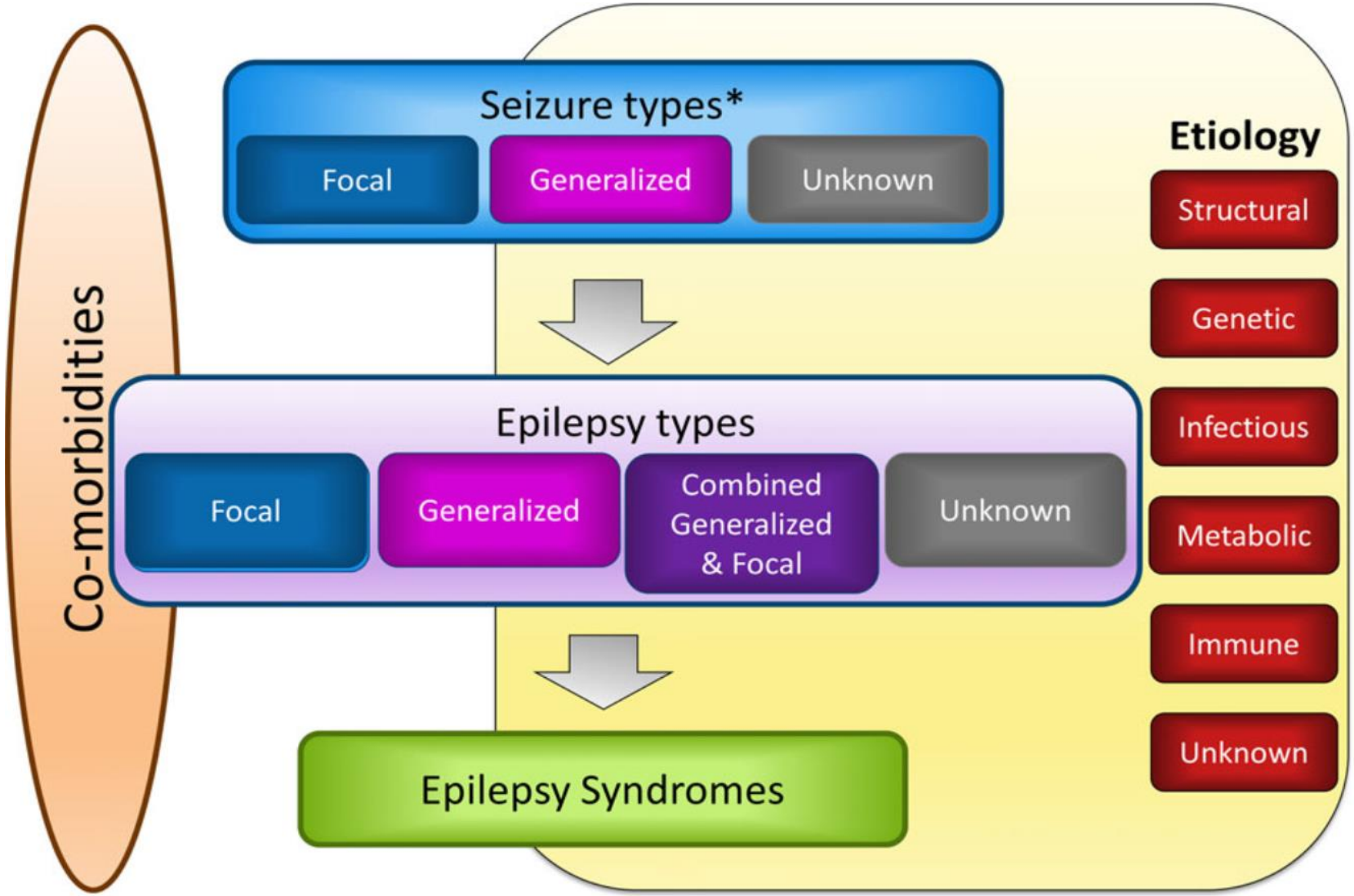
- The manifestations may be motor, sensory, autonomic or behavioral arrest, or as impaired consciousness.

Definition: Epilepsy

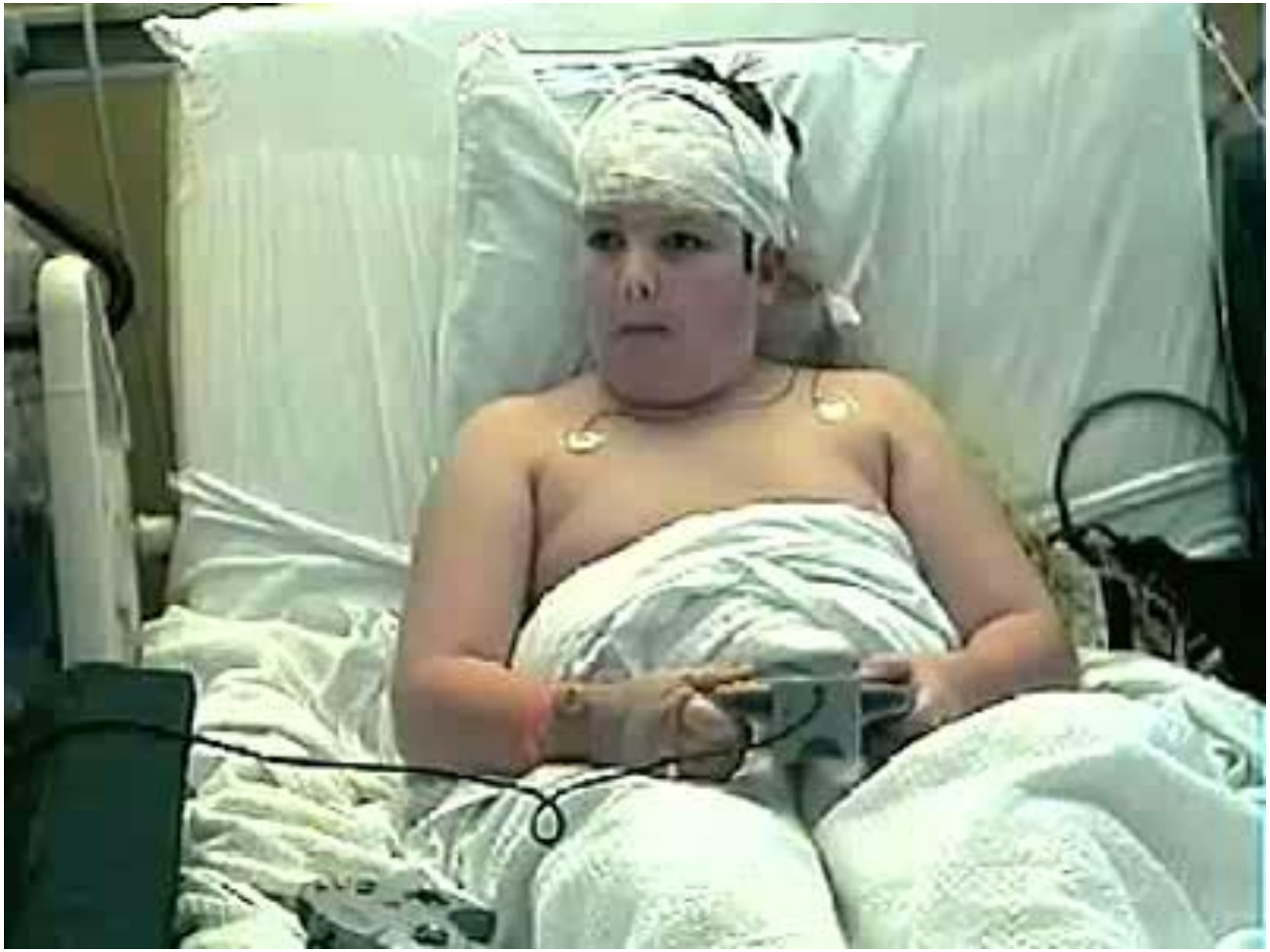
- **Epilepsy** is a disease of the brain defined by any of the following conditions:
  1. At least two unprovoked (or reflex) seizures occurring >24 h apart
  2. One unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years
  3. Diagnosis of an epilepsy syndrome
- Epilepsy is considered to be resolved for individuals who had an 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.
- Affects 50 million people worldwide and 1 in 200 children worldwide

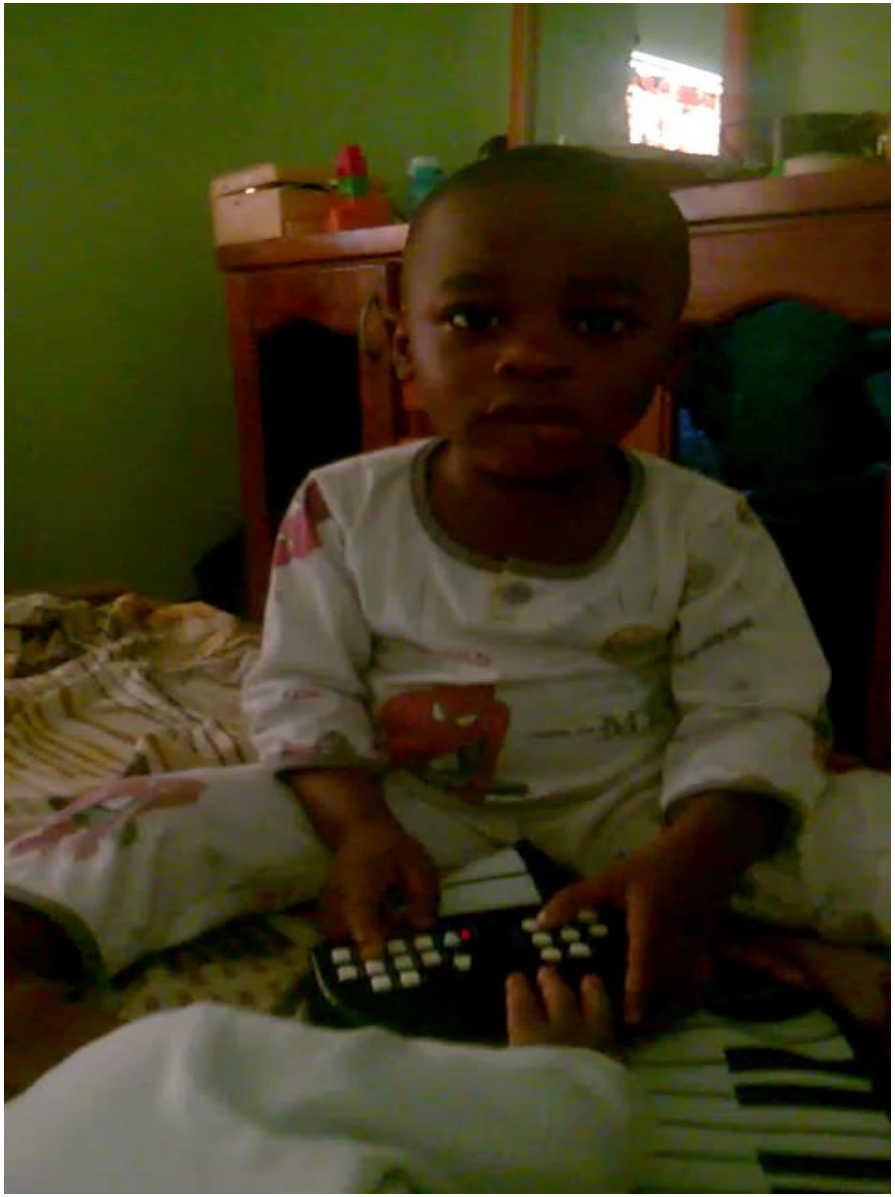
(Fisher et al. 2014)

Classification of Epilepsy



(ILAE, 2017)





Etiology of Epilepsy

1. **Structural:** e.g. calcifications, post-stroke, tuberous sclerosis complex, post-infection structural changes, etc.
2. **Metabolic:** e.g. inborn errors of metabolism, vitamin deficiency, etc.
3. **Genetic:** e.g. familial neonatal seizures, Dravet syndrome, GEFS+, etc.
4. **Infectious:** e.g. neurocysticercosis, HIV, CMV, cerebral toxoplasmosis, etc.
5. **Immune-mediated:** autoimmune encephalitis, antibody-mediated limbic encephalitis.
6. **Unknown** etiology

(ILAE, 2017)

Definition: Epilepsy Syndrome

- An epileptic syndrome is defined as a characteristic cluster of clinical and EEG features, often supported by specific etiological findings (structural, genetic, metabolic, immune, and infectious).
- The diagnosis of a syndrome in an individual with epilepsy frequently carries prognostic and treatment implications.
- Epilepsy syndromes often have age-dependent presentations and a range of specific comorbidities.

(Zuberi *et al*, 2022)

Epilepsy Syndrome

- Epilepsy syndromes can be divided in two groups:
  - self-limited epilepsy syndromes and
  - developmental and epileptic encephalopathies.

- Epilepsy syndromes can also be divided based on the age at onset:
  - Epilepsy syndromes with onset in the neonatal period and infancy
  - Epilepsy syndromes with onset in childhood
  - Epilepsy syndromes with onset in adolescents
  - Epilepsy syndromes that may begin at variable age

A Few Examples of Common Epilepsy Syndromes with Onset in Neonatal Period and Infancy

- **Neonatal period**
  - Self-limiting (benign) neonatal seizures
  - Self-limiting (benign) familial neonatal epilepsy
  - Early myoclonic encephalopathy (EME)
  - Ohtahara syndrome

- **Infancy (onset under 2 years)**
  - Genetic Epilepsy with Febrile seizures plus (GEFS+)
  - Epilepsy of infancy with migrating focal seizures
  - West syndrome
  - Dravet syndrome (Severe myoclonic epilepsy of infancy)

(Hammond and Wilmshurst, 2018)

A Few Examples of Common Epilepsy Syndromes with Onset in Childhood and Adolescence

- **Childhood onset epilepsy syndromes**
  - Genetic Epilepsy with Febrile Seizures plus (GEFS+)
  - Early-onset childhood occipital epilepsy (Panayiotopoulos type)
  - Doose syndrome) Epilepsy with myoclonic atonic (previously astatic) seizures
  - Lennox-Gastaut syndrome (LGS)
  - Self-limiting (Benign) epilepsy with centro-temporal spikes (BECTS)
  - Autosomal-dominant nocturnal frontal lobe epilepsy (ADNFLE)
  - Late-onset childhood occipital epilepsy (Gastaut type)
  - Childhood absence epilepsy (CAE)
  - Landau-Kleffner syndrome (LKS)

- **Adolescent onset epilepsy syndrome**
  - Juvenile absence epilepsy (JAE)
  - Juvenile myoclonic epilepsy (JME)

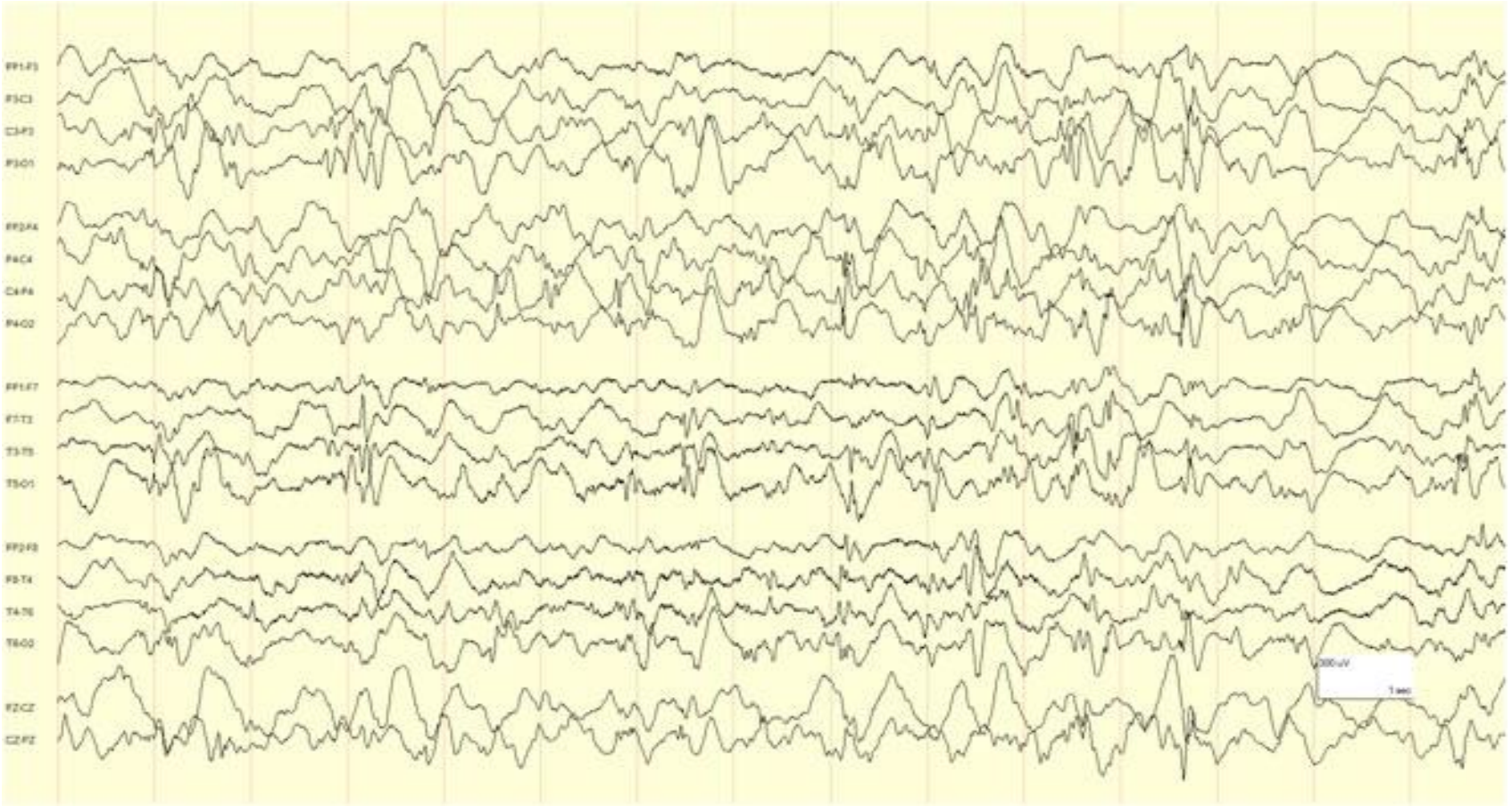
(Hammond and Wilmshurst, 2018)

Case 1

- A 7 month old baby presents to the clinic with abnormal movements described as repetitive extensor jerks of the limbs and trunk, mainly on awakening. The jerks occur in clusters, and she has 6-8 clusters in a day.
- The jerks started at the age of 6 months. She has lost neck control, become less interactive and has stopped babbling.

Case 1

Her EEG is as shown:



Case 1: Approach to the Child with Epileptic Spasms

- History: video is important
- Physical examination: look for dysmorphic features, features of TSC, etc.
- Investigation: EEG, brain MRI, metabolic workup, etc.
- Treatment: ACTH, oral prednisolone, vigabatrin

History

- Eye-witness account
- Patient's baseline neurological status
- Age at onset
- Seizure semiology
  - Ictal event (detailed description of all seizure types, duration, etc)
  - Postictal events
- Family history
- Developmental/cognitive outcome and co-morbidities
- Any videos to show?

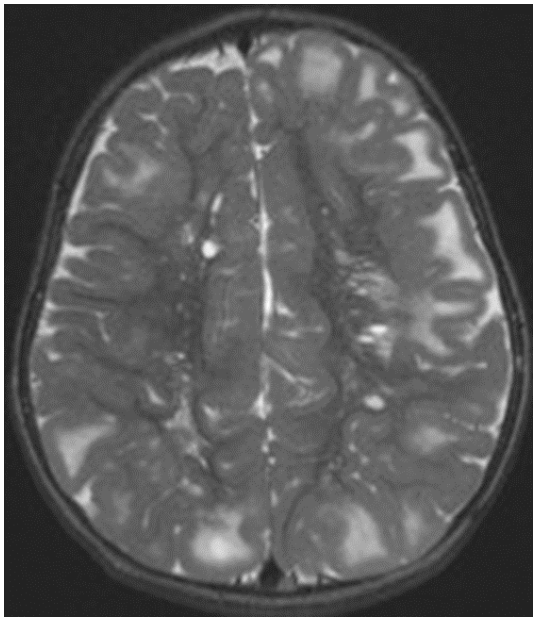
Physical examination

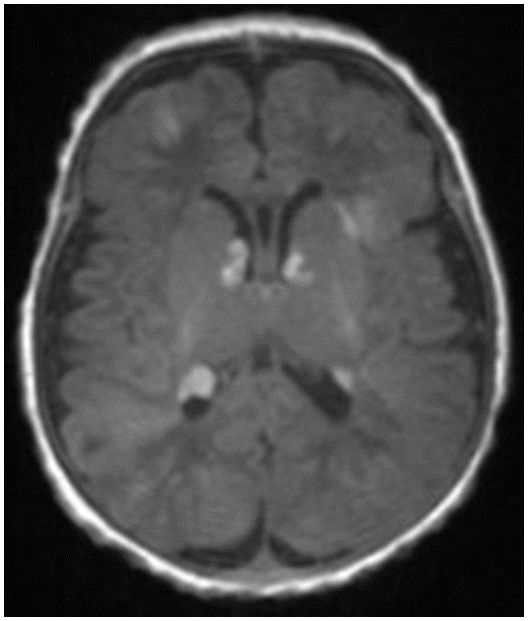
- General Examination (including skin)
- Detailed Neurological Examination
  - Cranial nerves
  - Motor examination (including cerebellar and extra-pyramidal manifestations)
  - Sensation
  - Developmental assessment
- Systemic Examination

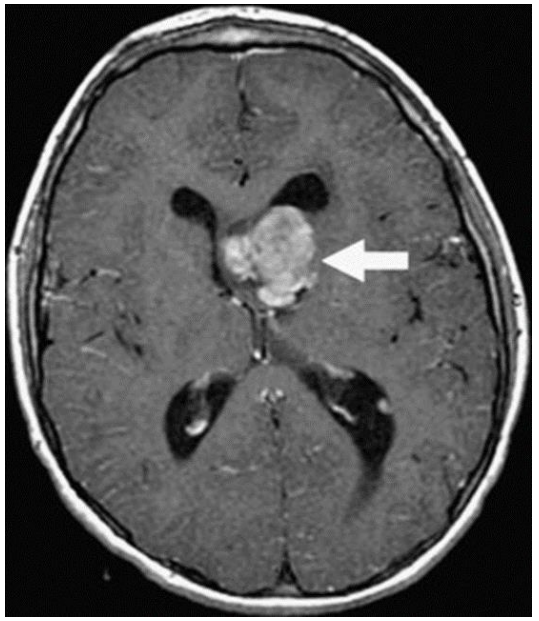
Investigations

- Basic investigations:
  - Blood counts
  - Serum electrolytes
  - LFTs, renal function, etc.
- Advanced investigations
  - Electroencephalogram (EEG)
  - Neuro-imaging (CT, MRI, SPECT, PET)
  - Metabolic screening
  - Auto-immune assays
  - Etc.

Brain MRI in a child with TSC







Treatment

- Medical treatment – traditional and newer ASMs
- Hormonal treatment – ACTH for epileptic spasms
- Steroids – Prednisolone for epileptic spasms
- Vitamins – pyridoxine, folinic acid and biotin for vitamin-responsive seizures
- Surgical treatment
- Dietary treatment (ketogenic diet)

Medical treatment

- Medical treatment uses traditional or newer ASMs
- Not all children with epilepsy require ASM therapy
- ASM selection is based on seizure type, epilepsy syndrome, and potential side effects
- Monotherapy is always preferred
- A few patients will require rational polypharmarcy
- Select ASM at the minimum dosage that provides good seizure control with minimal adverse effects

Traditional ASMs

Phenobarbitone
- Focal and generalized seizures
- Status epilepticus
- Neonatal seizures

Phenytoin
- Focal and generalized seizures
- Status epilepticus
- Neonatal seizures

Sodium valproate
- Broad spectrum
- Generalized seizures
- Absence seizures

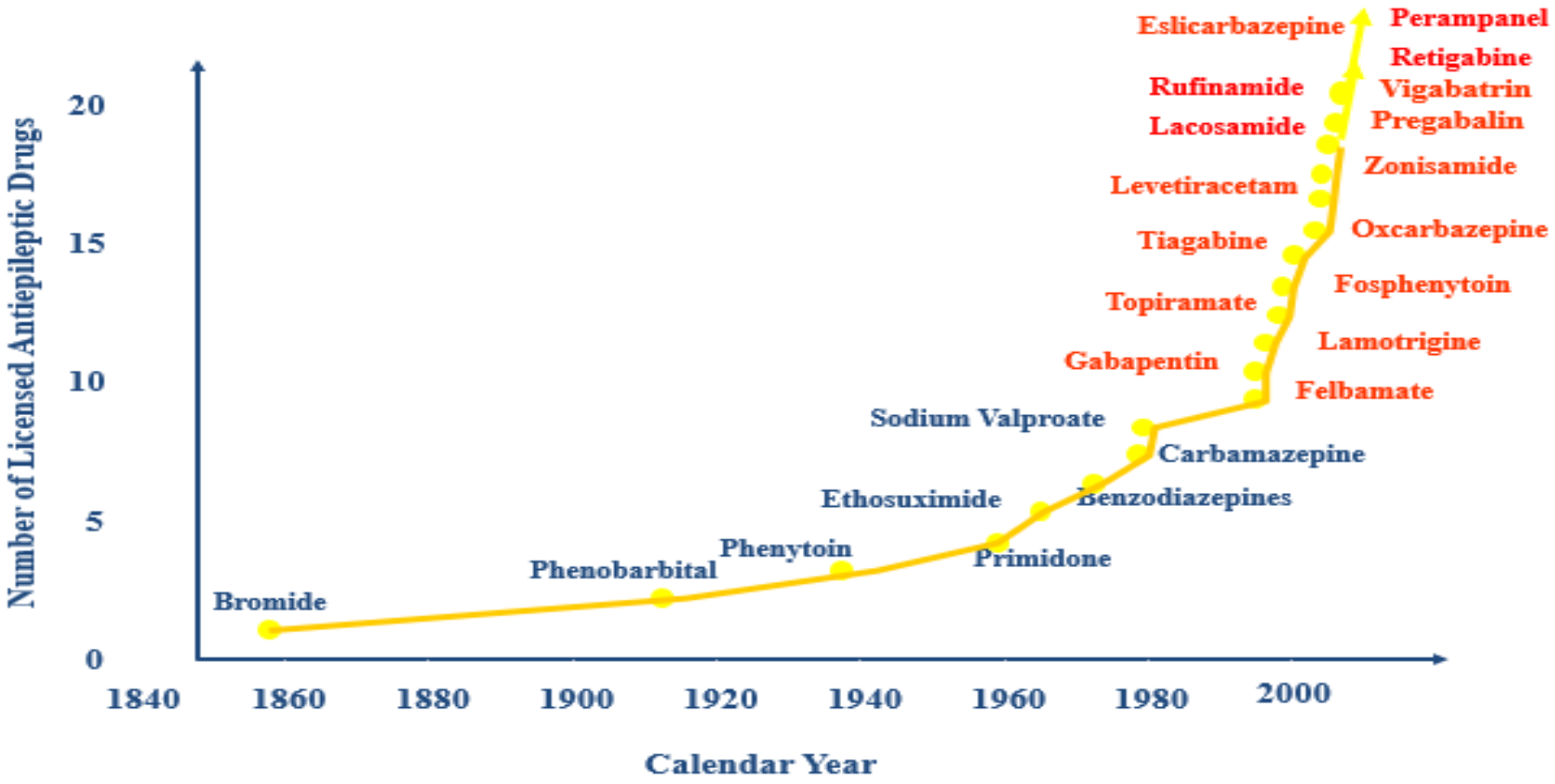
Carbamazepine
- Focal seizures
- Avoid in absences and myoclonic seizures

Ethosuxamide
- Absence seizures

Newer ASMs

- Lamotrigine
- Topiramate
- Levetiracetam
- Clonazepam
- Clobazam
- Vigabatrine
- Rufinamide
- Lacosamide
- Oxcarbazepine
- Perampanel
- Fosphenytoin
- Zonisamide
- Others including
  - Adrenocorticotropic hormone (ACTH) – for epileptic spasms
  - Steroids – for epileptic spasms
  - Vitamins (pyridoxine, folinic acid, biotin)

ASM Development



Surgical treatment

- Focal resection
- Lobectomy
- Hemispherectomy
- Corpus callostomy
- Vagus nerve stimulation

Ketogenic diet

- High fat, low carbohydrate, low protein
- Patient assumes a fasting state
- Brain relies on fatty acids instead of glucose
- Exact mechanism of action not known but few hypotheses proposed
- Effective for all seizure types

Summary

- Currently, epilepsy is classified at three levels, namely seizure type, epilepsy type and epilepsy syndrome, if possible.
- The etiology of epilepsy include structural, metabolic, genetic, infection, immune and unknown causes
- Epilepsy syndromes have age-dependent presentations, distinct clinical and EEG features, specific etiologies, response to treatment, comorbidities and prognosis
- Modalities for treatment of epilepsy include medications, surgery and ketogenic diet

ICNC 2022
17th INTERNATIONAL CHILD NEUROLOGY CONGRESS

International Child Neurology Congress 2024, (ICNC)
Cape Town, South Africa

The International Child Neurology Association
promoting education & research in child neurology worldwide

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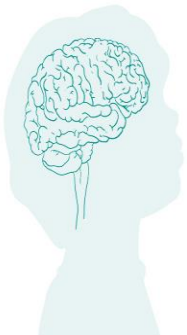
Footprints to the Future

SAVE THE DATE!!!!!



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Thank you