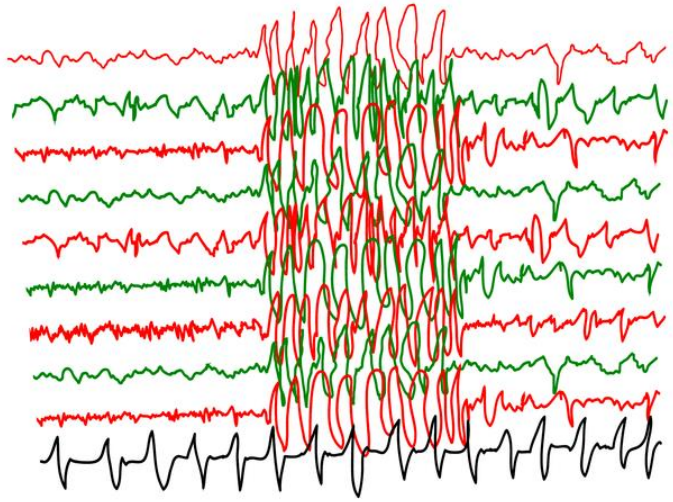
Pediatric Neurology

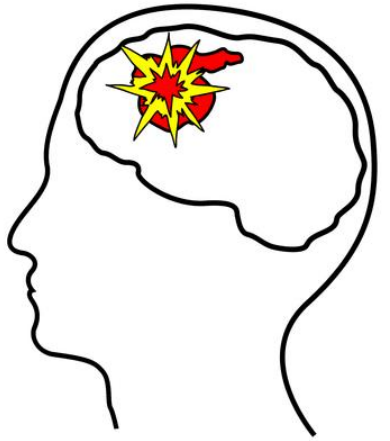
Amal Abu Libdeh, MBBS

Associate Professor
Pediatric Neurology

2023-2024

EPILEPSY AND EPILEPSY IMITATORS





Credit goes to:

Dr Abdelkarim A.Al-Qudah,MD ABCN

Professor, Pediatric neurology, JU, JUH

Outline

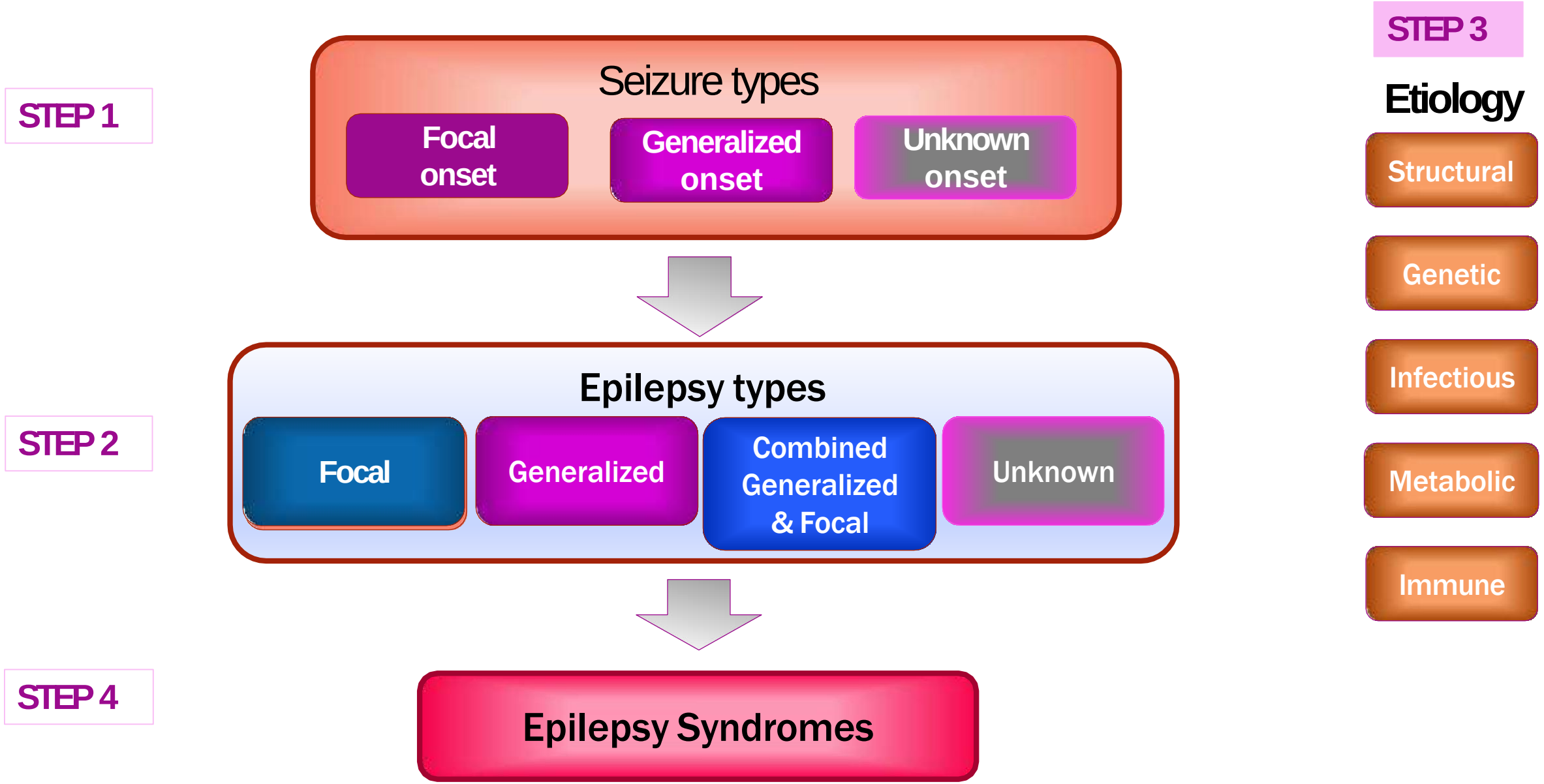
- **Epilepsy**
  - definition and new classification
- **Epilepsy imitators**
- **Headache**

DEFINITIONS

-Seizure: Is a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain.

-Epilepsy : is a disease of the brain defined by any of the following:

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


STEP 1
Seizure types
Focal onset
Generalized onset
Unknown onset
STEP 2
Epilepsy types
Focal
Generalized
Combined Generalized & Focal
Unknown
STEP 4
Epilepsy Syndromes
STEP 3
Etiology
Structural
Genetic
Infectious
Metabolic
Immune

ILAE 2017 Classification of Seizure Types Expanded Version [1]

Focal Onset

Aware | Impaired Awareness

Motor Onset
- automatisms
- atonic [2]
- clonic
- epileptic spasms [2]
- hyperkinetic
- myoclonic
- tonic

Nonmotor Onset
- autonomic
- behavior arrest
- cognitive
- emotional
- sensory

focal to bilateral tonic–clonic

Generalized Onset

Motor
- tonic–clonic
- clonic
- tonic
- myoclonic
- myoclonic–tonic–clonic
- myoclonic–atonic
- atonic
- epileptic spasms

Nonmotor (absence)
- typical
- atypical
- myoclonic
- eyelid myoclonia

Unknown Onset

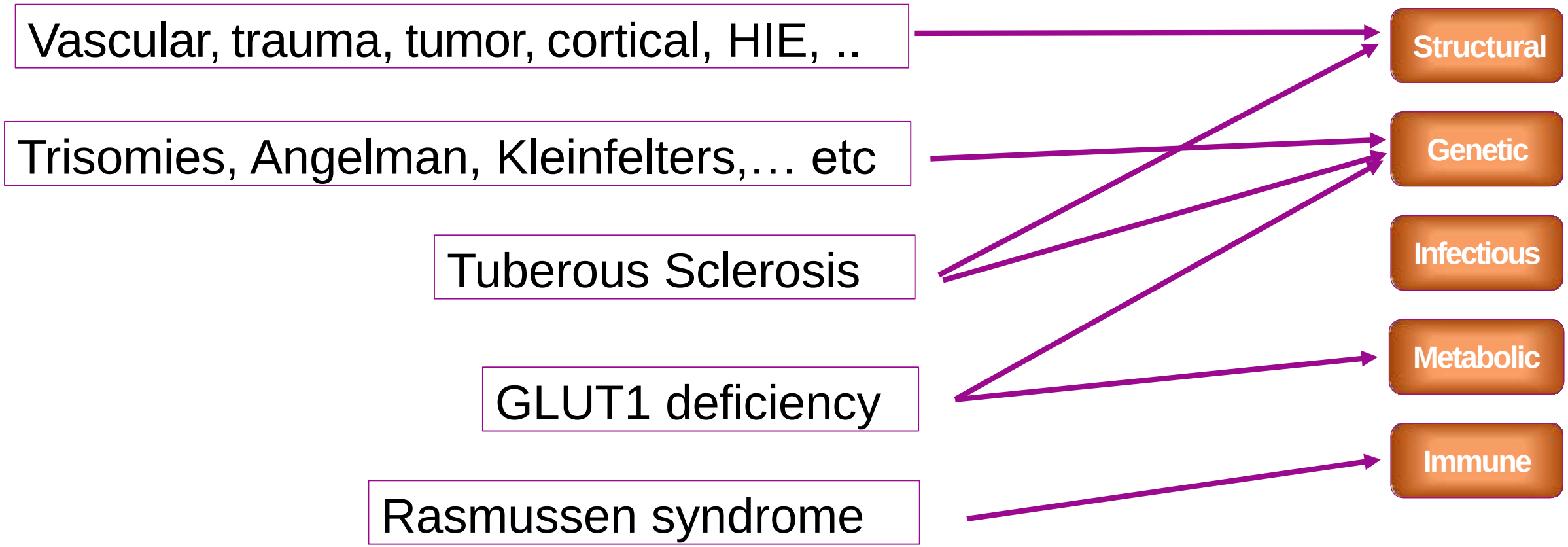
Motor
- tonic–clonic
- epileptic spasms

Nonmotor
- behavior arrest

Unclassified [3]

Absence seizures	Transient loss of consciousness, with an abrupt onset and termination, unaccompanied by motor phenomena except for some flickering of the eyelids and minor alteration in muscle tone. Absences may be typical (petit mal) or atypical and can often be precipitated by hyperventilation
Myoclonic seizures	Brief, often repetitive, jerking movements of the limbs, neck or trunk Non-epileptic myoclonic movements are also seen physiologically in hiccoughs (myoclonus of the diaphragm) or on passing through stage II sleep (sleep myoclonus)
Tonic seizures	Generalised increase in tone
Tonic–clonic seizures	Rhythmical contraction of muscle groups following the tonic phase. In the rigid tonic phase, children may fall to the ground, sometimes injuring themselves. They do not breathe and become cyanosed. This is followed by the clonic phase, with jerking of the limbs. Breathing is irregular, cyanosis persists and saliva may accumulate in the mouth. There may be biting of the tongue and incontinence of urine. The seizure usually lasts from a few seconds to minutes, followed by unconsciousness or deep sleep for up to several hours
Atonic seizures	Often combined with a myoclonic jerk, followed by a transient loss of muscle tone causing a sudden fall to the floor or drop of the head

Etiology


Vascular, trauma, tumor, cortical, HIE, ..
Trisomies, Angelman, Kleinfelters,… etc
Tuberous Sclerosis
GLUT1 deficiency
Rasmussen syndrome
Structural
Genetic
Infectious
Metabolic
Immune

Epilepsy Syndromes

Identified based on: *age at onset*, *seizure type(s)*, *EEG characteristics*, *etiology*, and other associated factors

Neonatal/infantile

- Benign familial
- Neonatal epilepsy,
- Early myoclonic
- Encephalopathy
- Ohtahara syndrome
- Dravet syndrome
- Myoclonic epilepsy of infancy,..

Childhood

- Febrile seizures plus
- Panayiotopoulos syndrome
- Epilepsy with myoclonic atonic (previously astatic) seizures
- Benign epilepsy with centrotemporal spikes
- Autosomal-dominant nocturnal frontal lobe epilepsy
- Epilepsy with myoclonic absences
- Lennox-Gastaut syndrome, …

Adolescent / adult

- Juvenile absence epilepsy
- Juvenile myoclonic epilepsy
- Epilepsy with generalized tonic- clonic seizures alone
- Progressive myoclonus epilepsies
- Autosomal dominant epilepsy with auditory features, etc

Variable age

- Familial focal epilepsy with variable foci.
- Reflex epilepsies.

ILAE 2017

- **Developmental encephalopathy**: where there is just developmental impairment without frequent epileptic activity associated with regression or further slowing of development.
- **Epileptic encephalopathy:** where there is no preexisting developmental delay, and the genetic mutation is not thought to cause slowing in its own right; and developmental and epileptic encephalopathy where both factors play a role.

- Sometimes it can be difficult to determine whether the epileptic or developmental component is more important in contributing to a patient's presentation.

ILAE 2017

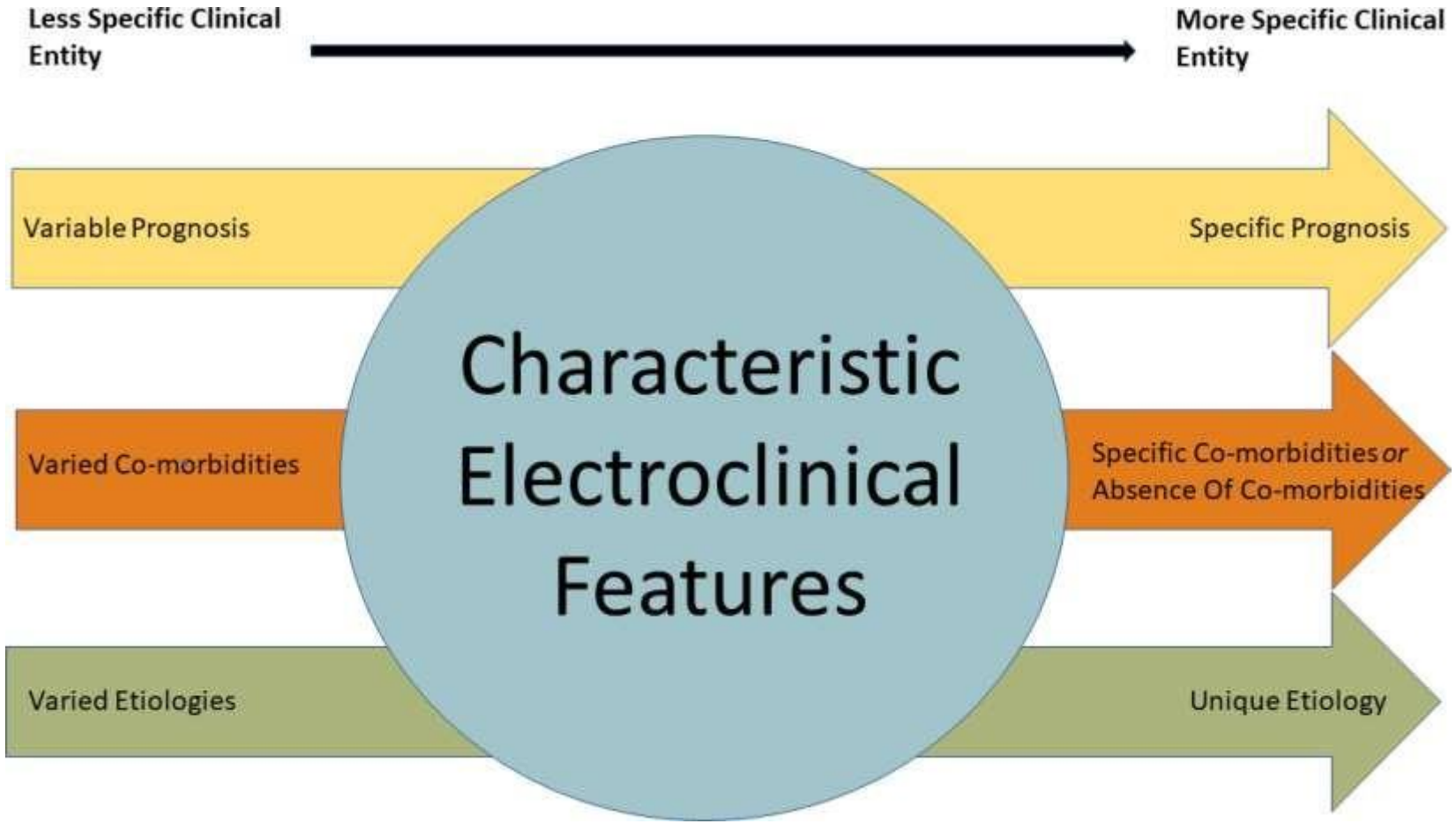
-Thus “benign,” as a descriptor for epilepsy, is replaced by both “self-limited” and “pharmacoresponsive,” each replacing different components of the meaning of benign.

-“Self- limited” refers to the likely spontaneous resolution of a syndrome.

-“Pharmacoresponsive” means that the epilepsy syndrome will be likely to be controlled with appropriate antiepileptic therapy.

-Some individuals with these syndromes may still not be pharmacoresponsive.

ILAE 2022 epilepsy syndrome



ILAE 2022 epilepsy syndromes

- **Epilepsy syndrome:** a characteristic cluster of clinical and EEG features, often supported by specific etiological findings (structural, genetic,metabolic, immune, and infectious).
- This designation frequently carries prognostic and treatment implications. Syndromes often have age-dependent presentations (they typically start at specific ages, and in some cases may also remitat certain ages).
- Many syndromes are strongly correlated with a range of specific intellectual, psychiatric, and other comorbidities, whereas these are absent in other types.
- Epilepsy syndromes have traditionally been grouped according to age at onset. Accordingly, the ILAE position papers describe separately syndromes with onset in neonates and infants (up to age 2 years), syndromes with onset in childhood, and syndromes that may begin at variable ages (in both pediatric and adult patients).
- The syndromes are further subdivided into generalized,focal, or generalized and focal, based on seizure type(s), with a separate category for syndromes with developmental and epileptic encephalopathy (DEE) or progressive neurological deterioration.

ILAE 2022 epilepsy syndromes in neonates and infants

Self-limited epilepsies

- Self-limited neonatal epilepsy (SeLNE)
- Self-limited familial neonatal-infantile epilepsy (SeLFNIE)
- Self-limited infantile epilepsy (SeLIE)
- Genetic epilepsy with febrile seizures plus (GEFS+)
- Myoclonic epilepsy in infancy (MEI)

Developmental and epileptic encephalopathies (DEE)

- Ealy infantile developmental and epileptic encephalopathy (EIDEE)
- Epilepsy in infancy with migrating focal seizures (EIMFS)
- Infantile epileptic spasms syndrome (IESS)
- Dravet syndrome (DS)

Etiology-specific syndromes

- *KCNQ2*-DEE
- Pyridoxine-dependent (*ALDH7A1*)-DEE (PD-DEE)
- Pyridox(am)ine 5'-Phosphate Deficiency (PNPO)-DEE (P5PD-DEE)
- *CDKL5*-DEE
- *PCDH19* clustering epilepsy
- Glucose Transporter 1 Deficiency Syndrome (GLUT1DS)
- Sturge Weber syndrome (SWS)
- Gelastic seizures with hypothalamic hamartoma (GS-HH)

ILAE 2022 epilepsy syndromes

- The childhood onset epilepsy syndromes, most of which have both mandatory seizure type(s) and interictal electroencephalographic (EEG) features.
- Based on the 2017 Classification of Seizures and Epilepsies, some syndrome names have been updated using terms directly describing the seizure semiology. Epilepsy syndromes beginning in childhood have been divided into three categories:
- (1) Self-limited focal epilepsies: 4 syndromes: self-limited epilepsy with centrotemporal spikes, self-limited epilepsy with autonomic seizures, childhood occipital visual epilepsy, and photosensitive occipital lobe epilepsy;
- (2) Generalized epilepsies: 3 syndromes: childhood absence epilepsy, epilepsy with myoclonic absence, and epilepsy with eyelid myoclonia;
- (3) Developmental and/or epileptic encephalopathies: 5 five syndromes: epilepsy with myoclonic–atonic seizures, Lennox–Gastautsyndrome, developmental and/or epileptic encephalopathy with spike-and-wave activation in sleep, hemiconvulsion–hemiplegia Epilepsy syndrome, and febrile infection-related epilepsy syndrome.

Epilepsy syndromes

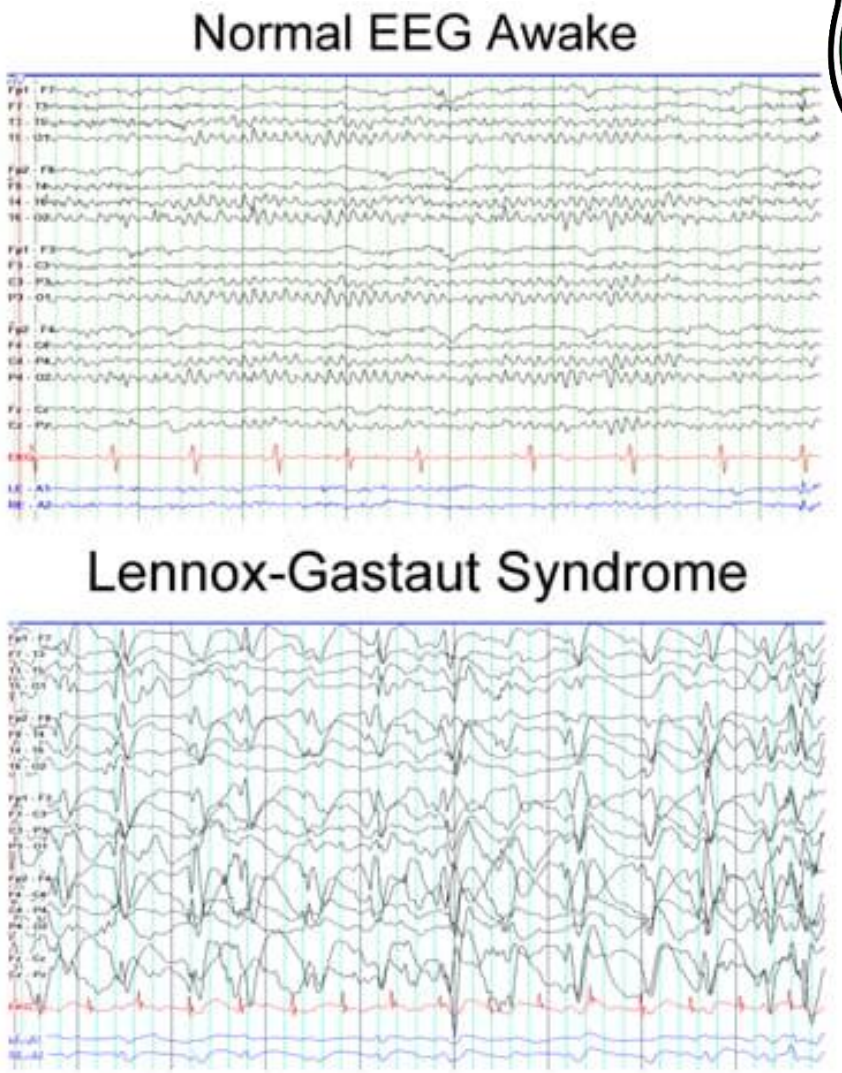
Age + seizure semiology + EEG pattern.

<u>Lennox Gastaut syndrome:</u>

- (Age < 8 yr. Multiple seizure types including tonic, myoclonic, and atypical absence. EEG very abnormal).

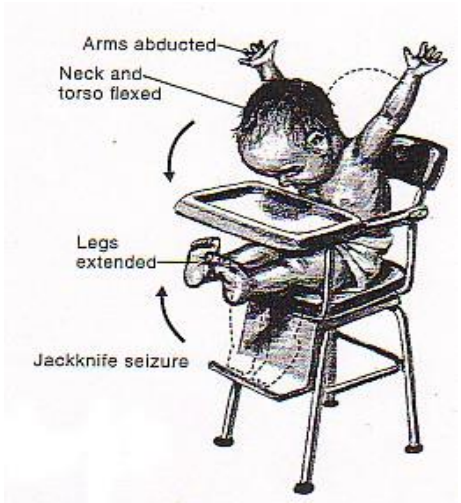
- **Treatment : V**alproate, Clobazam, Lamotrigine.

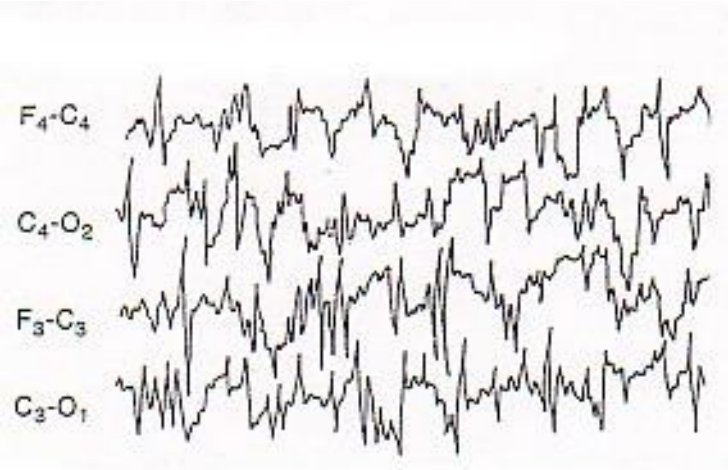
- Refractory to treatment. Typically associated with significant intellectual dysfunction.



Infantile spasms (West syndrome)

- <u>Infantile age onset.</u>
- (Age: infancy + sz: spasms + EEG: Hypsarrhythmia).
- Prognosis variable depending on underlying etiology. Frequently leads to dev delay.
- Severely abnormal EEG pattern: disorganized, discontinuous, high amplitude, multifocal spikes called hypsarrhythmia.
- **Treatment:** is Steroids (ACTH, prednisone) or Vigabatrin.





Childhood absence

-Childhood age onset.

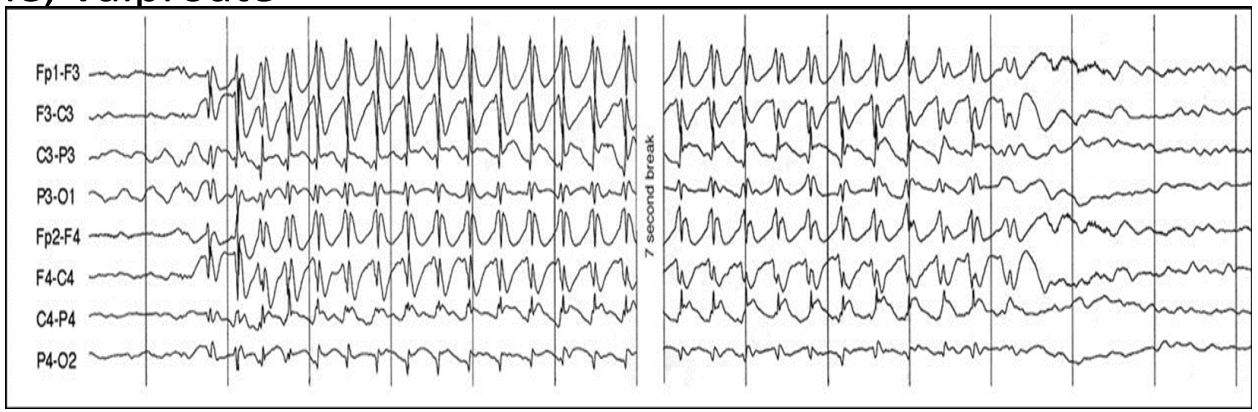
-(Age: 4-8 yr + Sz: absence + EEG: generalized 3 Hz spike wave discharges)

-Sudden onset of staring, interrupting speech or activity. Occurs multiple times per day. Short duration (seconds).

- Hyperventilation may provoke a seizure.

-Good prognosis, typically resolves by adolescence.

-**Treatment :** Ethosuximide, Lamotrigine, Valproate

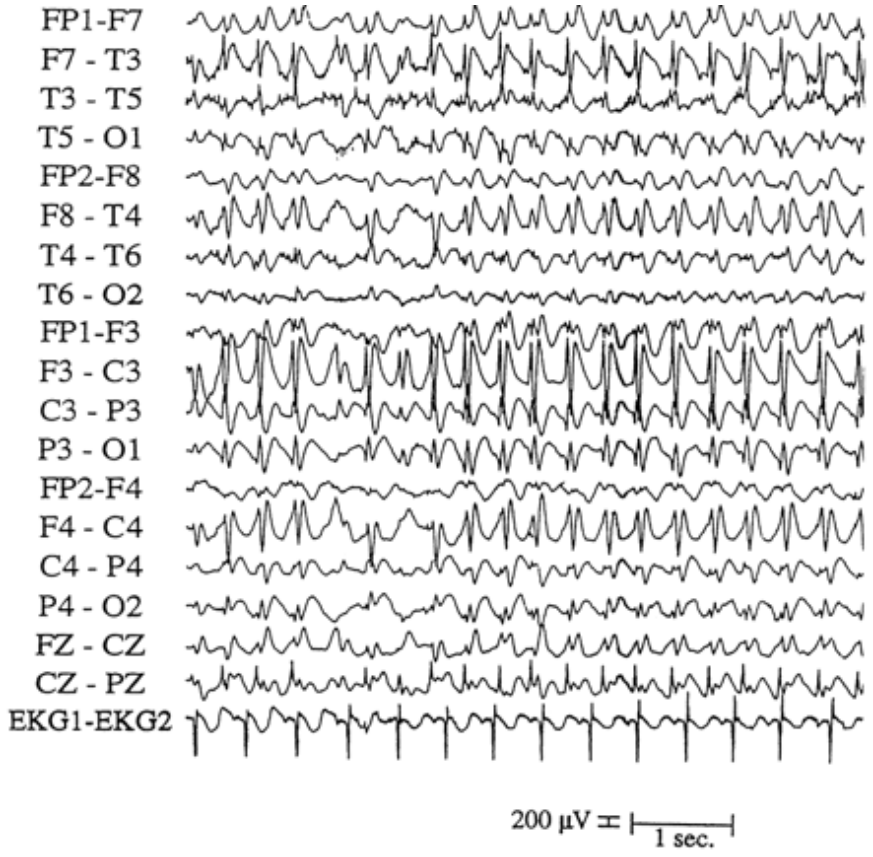


Benign Rolandic Epilepsy (BECTS)

-Childhood age onset.
- (Age: 4-11 yr + Sz: focal + EEG: centrotemporal spikes).

- Seizures: brief, infrequent, when awake or upon arousal from sleep, paresthesia on one side of the tongue or mouth, followed by dysarthria or gagging, jerking of the ipsilateral face, and excessive drooling.
- Good prognosis, resolves by puberty.

- Treatment: Carbamazepine, oxcarbazepine.



Juvenile Myoclonic epilepsy (JME)

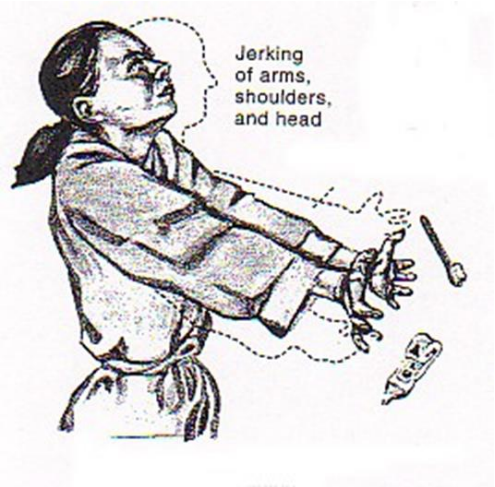
- Adolescent age onset.

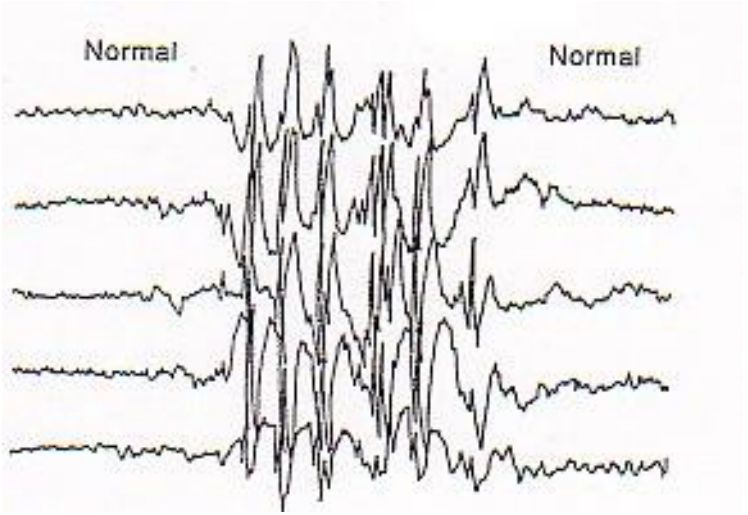
- (Age >12 yr + Sz: myoclonic sz (Kelog's sz)+ EEG: fast spike wave 4-6 hz).

- Mostly affects arms, typically upon awakening.

- Sleep deprivation and flashing lights may provoke seizures.

- **Life-long treatment** : Lamotrigine, Valproate.





Diagnosis

- History and physical exam

- Work up depends on initial impression:
  - EEG
  - Imaging (brain MRI), genetic, metabolic,, CSF study ….etc

Epilepsy Demographic data in Jordan:

Neurosciences (Riyadh). 2017 Oct; 22(4): 267–273.
doi: 10.17712/nsj.2017.4.20170164

PMCID: PMC5946375
PMID: 29057851

Type and etiology of pediatric epilepsy in Jordan

a multi-center study

Abdelkarim A. Al-Qudah, MD, ABCN, Abla Albsoul-Younes, PhD, Amira T. Masri, MD, Samah K. AbuRahmah, MD, Ibrahim A. Alabadi, PhD, Omar A. Nafi, MD, Lubna F. Gharaibeh, MSc, Amer A. Murtaja, Bsc, Lina H. Al-Sakran, MSc, Haya A. Arabiat, MD, and Abdallah A. Al-Shorman, MD

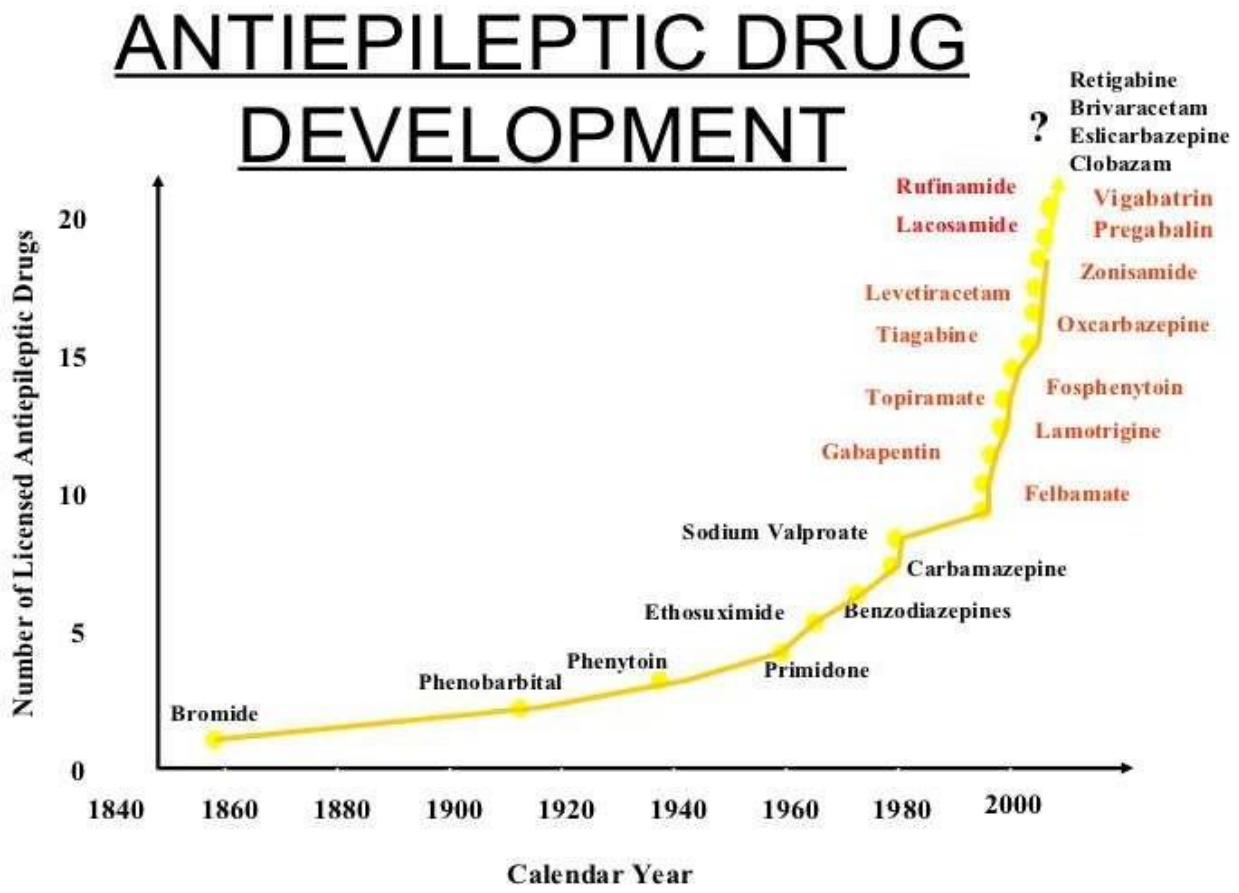
Out Of the 663 patients included in the study, (90.2%) had one seizure type, (53%) of this type were focal seizures followed by generalized seizures (41.5%) and spasms (5.5%). Distinctive constellations were found in 11/663 (1.7%) patients. Benign epilepsies with centrotemporal spikes were the most common electro clinical syndromes 60/221 (27.1%). Epilepsies attributed to structural-metabolic causes were documented in 278/663 (41.9%) patients, unknown causes 268/663(40.4%) and genetic causes in 117/663(17.7%). Most common causes of structural-metabolic group were due to perinatal insults (32%) and most common causes of the genetic group were the presumed genetic electro clinical syndromes (93.1%).

TREATMENT

- Antiseizure medications
- Other medications
  - Immune therapy (Rasmussen ….)
  - Treatable metabolic disorders (Pyridoxine, Folinic acid …..)

- Epilepsy surgery (resection, callosotomy, etc)
- Ketogenic diet
- Vagal nerve stimulation

Antiseizure medications



Antiseizure medications

- DRUGS THAT AFFECT VOLTAGE-DEPENDENT SODIUM CHANNELS
  - Old : Carbamazepine Phenytoin
  - New : Lamotrigine Oxcarbazepine Zonisamide Lacosamide Rufinamide
- DRUGS THAT AFFECT CALCIUM CURRENTS
  - Ethosuximide
- DRUGS THAT AFFECT GABA ACTIVITY
  - Benzodiazepines, barbiturates, gapabentine
- DRUGS THAT AFFECT GLUTAMATE RECEPTERS
  - Topiramate
- DRUGS WITH MULTIPLE MECHANISMS OF ACTION
  - Topiramate, valproic acid
- Unknown
  - Levetiracetam

Drug Resistant Epilepsy (DRE)

- It is defined as failure of seizure control after adequate medical therapy with **two** or more **appropriate** anti-epileptic drugs.

- Epilepsy surgery can be indicated earlier when drug-resistance is highly expected such as in the mesial temporal lobe epilepsy with hippocampal sclerosis or when adverse effect of poor seizure control is expected on patient's development in young children.

1) K wan P, et al. Definition of drug resistant epilepsy:consensus proposal by the ad hoc task force of the ILAE Commission on therapeutic strategies.Epilepsia, 2010 .
2) K wan P, et al. Drug-resistant epilepsy. N Engl J Med 365: 919–926, 2011

Non-invasive pre-surgical evaluations for epilepsy

1. Detailed Clinical history and exam
2. EEG and video EEG
3. MRI (for epileptogenic lesions)
4. FDG-PET
5. Ictal SPECT, inter ictal SPECT
6. Magnetoencephalography (MEG) and Functional MEG
7. Functional MRI
8. Neuropsychological evaluation

Masaki et al 2016

Status epilepticus

- SE is a condition resulting either from the failure of the mechanisms responsible for seizure termination or from the initiation of mechanisms which lead to abnormally prolonged seizures.

- It is a condition that can have long-term consequences, including neuronal death, neuronal injury, and alteration of neuronal networks, depending on the type and duration of seizures.

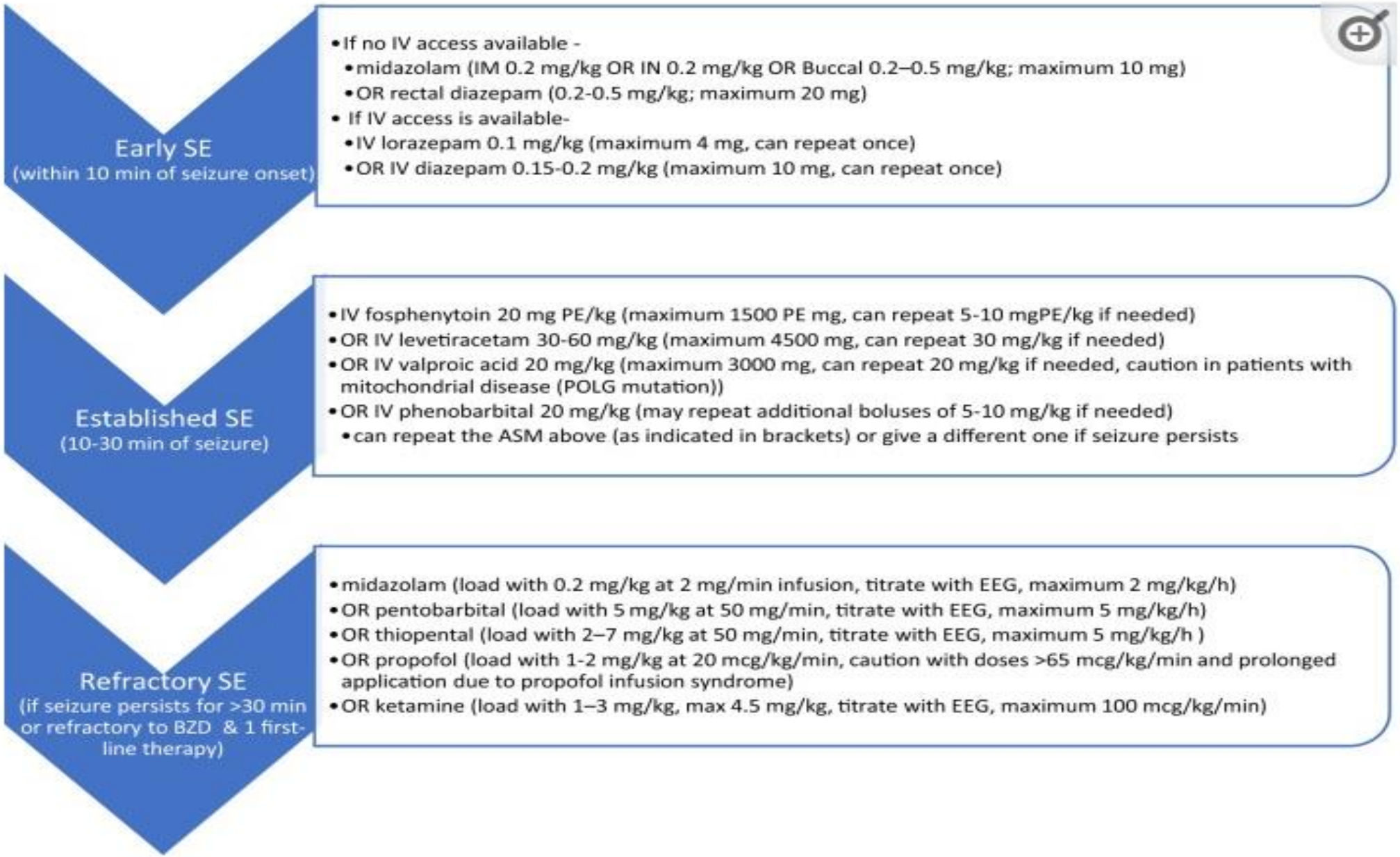
Operational dimensions with t_1 indicating the time that emergency treatment of SE should be started and t_2 indicating the time at which long-term consequences may be expected

Type of SE	T 1	T 2
Tonic–clonic SE	5 min	30 min
Focal SE with impaired consciousness	10 min	>60 min
Absence status epilepticus	10–15 min	Unknown

ILAE org 2017

Treatment of SE :

- Assessment (ABC)
- Supportive care
- Anticonvulsant :
  - 0 to 5 minutes : Benzodiazepine : (Diazepam. Midazolam …)
  - 5 to 10 minutes : Benzodiazepine (second dose)
  - 10 to 15 minutes : Fosphenytoin (second line), Phenobarbital.
  - 15 to 30 minutes : Phenobarbital, valproic acid, pyridoxine)
  - After 30 minutes : obtain anesthesiology consult


Early SE
(within 10 min of seizure onset)
• If no IV access available -
• midazolam (IM 0.2 mg/kg OR IN 0.2 mg/kg OR Buccal 0.2–0.5 mg/kg; maximum 10 mg)
• OR rectal diazepam (0.2-0.5 mg/kg; maximum 20 mg)
• If IV access is available-
• IV lorazepam 0.1 mg/kg (maximum 4 mg, can repeat once)
• OR IV diazepam 0.15-0.2 mg/kg (maximum 10 mg, can repeat once)
Established SE
(10-30 min of seizure)
• IV fosphenytoin 20 mg PE/kg (maximum 1500 PE mg, can repeat 5-10 mgPE/kg if needed)
• OR IV levetiracetam 30-60 mg/kg (maximum 4500 mg, can repeat 30 mg/kg if needed)
• OR IV valproic acid 20 mg/kg (maximum 3000 mg, can repeat 20 mg/kg if needed, caution in patients with mitochondrial disease (POLG mutation))
• OR IV phenobarbital 20 mg/kg (may repeat additional boluses of 5-10 mg/kg if needed)
• can repeat the ASM above (as indicated in brackets) or give a different one if seizure persists
Refractory SE
(if seizure persists for >30 min or refractory to BZD & 1 first-line therapy)
• midazolam (load with 0.2 mg/kg at 2 mg/min infusion, titrate with EEG, maximum 2 mg/kg/h)
• OR pentobarbital (load with 5 mg/kg at 50 mg/min, titrate with EEG, maximum 5 mg/kg/h)
• OR thiopental (load with 2–7 mg/kg at 50 mg/min, titrate with EEG, maximum 5 mg/kg/h)
• OR propofol (load with 1-2 mg/kg at 20 mcg/kg/min, caution with doses >65 mcg/kg/min and prolonged application due to propofol infusion syndrome)
• OR ketamine (load with 1–3 mg/kg, max 4.5 mg/kg, titrate with EEG, maximum 100 mcg/kg/min)

Febrile seizures

- A seizure occurring in childhood after 6 months of age associated with a febrile illness not caused by an infection of the central nervous system (CNS), without previous neonatal seizures or a previous unprovoked seizure, and not meeting the criteria for other acute symptomatic seizures.
  - Simple: lasted less than 15 minutes. Generalized. Did not recur within 24 hours.
  - Complex: last more than 15 minutes, and/or focal, and/or recurred within 24 hours.

- Age: 6 months to 5 years.
- Evaluation after first time febrile seizure: should be directed towards the etiology of the fever. No tests are done routinely.
- Not treated with daily prophylactic anti seizure medication.
- Antipyretics (both as needed and scheduled) have not been shown to prevent seizures.
- Nasal or Rectal diazepam (Valium gel, Diastat, Valtoco) may be used as a rescue medication for prolonged seizures lasting more than 4 minutes.

Neonatal Seizures

- Incidence is higher during this period than in any other period in life:
  - 60/1,000 in infants with birth weights <1.5 kg.
  - 3/1,000 in infants weighing between 2.5 to 4kg.

- There are 5 main neonatal seizure types :
  - Subtle seizures: (transient eye deviations, nystagmus, blinking, mouthing, abnormal extremity movements, fluctuations in heart rate, hypertension episodes, and apnea. Subtle seizures occur more commonly in premature than in full-term infants).
  - Clonic seizures (can be focal or multifocal).
  - Tonic seizures (can be focal or generalized).
  - Spasms (sudden generalized jerks lasting 1-2 sec)
  - Myoclonic seizures (can be focal, multifocal, and generalized).

Neonatal Seizures – etiology

AGES 1-4 DAYS

- Hypoxic-ischemic encephalopathy
- Drug withdrawal, maternal drug use of narcotic or barbiturates
- Drug toxicity: lidocaine, penicillin
- Intraventricular hemorrhage
- Acute metabolic disorders
- Inborn errors of metabolism
- Pyridoxine deficiency (must be considered at any age)

AGES 4-14 DAYS

- Infection
- Metabolic disorders
- Drug withdrawal, maternal drug use of narcotics or barbiturates
- Benign neonatal convulsions, familial and nonfamilial
- Kernicterus, hyperbilirubinemia

AGES 2-8 WK

- Infection
- Head injury
- Inherited disorders of metabolism
- Malformations of cortical development
- Lissencephaly
- Tuberous sclerosis

Neonatal Seizures

Diagnosis :

- Taking a detailed prenatal and postnatal history and performing an adequate physical examination.
- EEG is considered the main tool for diagnosis.
- Blood test : glucose, Ca, Mg, other electrolytes, and BUN.
- Lumber puncture.
- Metabolic work up
- Imaging (brain MRI)

Neonatal Seizures - Prognosis

- Prognosis of neonatal seizures has become better owing to improvement and advancement of obstetric care and NICU care.
- Mortality from neonatal seizures has decreased from 40 to 20%.
- The underlying etiology of the seizures is the main determinant of outcome.
- EEG is highly associated with the outcome in premature and full-term infants.
- Predictors of less-favorable later outcome
  - Abnormal EEG background.
  - Prolonged electrographic seizures (>10 min/hour).
  - Multifocal periodic electrographic discharges.
  - Spread of the electrographic seizures to the contralateral side

Neonatal Seizures - Treatment

- Treat the underlying cause (infection, electrolyte imbalance, etc)
- Anti seizure medications:
  - Phenobarbital
  - Phenytoin and Fosphenytoin
  - Levetiracetam
  - Lorazepam
  - Diazepam, midazolam

Outline

- **Epilepsy**
  - definition and new classification
- **Epilepsy imitators**
- **Headache**

EPILEPSY IMITATORS

SYNCOPE AND ANOXIC SEIZURES

BEHAVIORAL, PSYCHOLOGICAL AND PSYCHIATRIC DISORDERS

SLEEP RELATED CONDITIONS

PAROXYSMAL MOVEMENT DISORDERS

MIGRAINE ASSOCIATED DISORDERs

MISCELLANEOUS EVENTS

SYNCOPE AND ANOXIC SEIZURES

- Vasovagal syncope
- Reflex anoxic seizures
- Breath-holding attacks
- Hyperventilation syncope
- Compulsive valsalva
- Neurological syncope
- Imposed upper airways obstruction
- Orthostatic intolerance
- Long QT and cardiac syncope
- Hyper-cyanotic spells

- Affects all ages
- Brief, lasting seconds
- Preceded by triggers
- Convulsive movements occur in 50%
- Positive F. Hx is common

SYNCOPE AND ANOXIC SEIZURES

- Vasovagal syncope
- Reflex anoxic seizures
- Breath-holding attacks
- Hyperventilation syncope
- Compulsive valsalva
- Neurological syncope
- Imposed upper airways obstruction
- Orthostatic intolerance
- Long QT and cardiac syncope
- Hyper-cyanotic spells

- Occurs from early infancy onwards, either remit or evolve into vasovagal
- Preceded by sudden stimulus such as bump or knock, result in profound vagal stimulus, transient asystole
- Child become exceedingly pale and loss of consciousness, tonic posturing is possible.

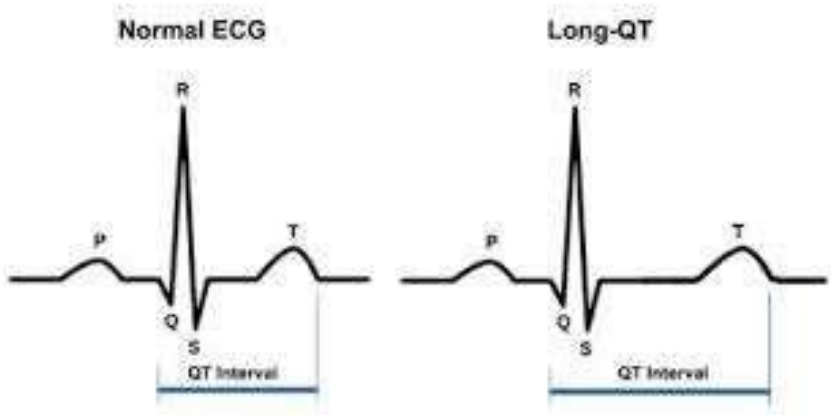
SYNCOPE AND ANOXIC SEIZURES

- Vasovagal syncope
- Reflex anoxic seizures
- Breath-holding attacks
- Hyperventilation syncope
- Compulsive valsalva
- Neurological syncope
- Imposed upper airways obstruction
- Orthostatic intolerance
- Long QT and cardiac syncope
- Hyper-cyanotic spells

- Affect pre-school children
- Start with crying then stop breathing in expiration.
- Becomes blue with deep cyanosis
- They breath in or go to transient syncope, tonic posturing is possible
- More common if the child has iron deficiency anemia.

SYNCOPE AND ANOXIC SEIZURES

- Vasovagal syncope
- Reflex anoxic seizures
- Breath-holding attacks
- Hyperventilation syncope
- Compulsive valsalva
- Neurological syncope
- Imposed upper airways obstruction
- Orthostatic intolerance
- Long QT and cardiac syncope
- Hyper-cyanotic spells



- In long QT syndrome a ventricular tachyarrhythmia may be spontaneous or triggered by fright, exercise, surprise, and immersion in water.
- Syncope in sleep, a strong family history of syncope and a history of sudden death or drowning should raise suspicions of a cardiac syncope.
- Sensorineural deafness is associated with some types of long QT syndrome

BEHAVIORAL, PSYCHOLOGICAL AND PSYCHIATRIC DISORDERS

- Daydreaming /inattention
- Infantile gratification
- Eidetic imagery
- Tantrums and rage reactions
- Out of body experiences
- Panic attacks
- Dissociative states
- Non-epileptic seizures
- Hallucinations in psychiatric disorders
- Fabricated / factitious illness

- Self-stimulation includes behavior which may be seen from infancy onwards, more so in pre-school girls.
- Rhythmic hip flexion and adduction may be accompanied by a distant expression, a flushed face and sometimes followed by sleepiness.

SLEEP RELATED CONDITIONS

- Sleep related rhythmic movement disorders
- Hypnogogic jerks
- Parasomnias
- REM sleep disorders
- Benign neonatal sleep myoclonus
- Periodic leg movements
- Narcolepsy-cataplexy

PAROXYSMAL MOVEMENT DISORDERS

- Tics
- Stereotypies
- Paroxysmal kinesigenic dyskinesia
- Paroxysmal nonkinesigenic dyskinesia
- Paroxysmal exercise induced dyskinesia
- Benign paroxysmal tonic upgaze
- Episodic ataxias
- Alternating hemiplegia
- Hyperekplexia
- Opsoclonus-myoclonus syndrome

- Involuntary, sudden, rapid, repetitive, non-rhythmic, simple or complex movements or vocalizations.
- Common in childhood and tend to wax and wane in frequency over time
- Urge or compulsion to perform the tic, and an ability to suppress the tic (to some degree) are important features on history that support the diagnosis.

PAROXYSMAL MOVEMENT DISORDERS

- Tics
- Stereotypies
- Paroxysmal kinesigenic dyskinesia
- Paroxysmal nonkinesigenic dyskinesia
- Paroxysmal exercise induced dyskinesia
- Benign paroxysmal tonic upgaze
- Episodic ataxias
- Alternating hemiplegia
- Hyperekplexia
- Opsoclonus-myoclonus syndrome

Genetically determined movement disorders

PAROXYSMAL MOVEMENT DISORDERS

- Tics
- Stereotypies
- Paroxysmal kinesigenic dyskinesia
- Paroxysmal nonkinesigenic dyskinesia
- Paroxysmal exercise induced dyskinesia
- Benign paroxysmal tonic upgaze
- Episodic ataxias
- Alternating hemiplegia
- Hyperekplexia
- Opsoclonus-myoclonus syndrome

- Characterized by an exaggeration of the normal startle response and has several genetic linked to dysfunction of the inhibitory glycinergic pathway in the nervous system.
- Evident from the neonatal period or early infancy.
- Die of apnea.

PAROXYSMAL MOVEMENT DISORDERS

- Tics
- Stereotypies
- Paroxysmal kinesigenic dyskinesia
- Paroxysmal nonkinesigenic dyskinesia
- Paroxysmal exercise induced dyskinesia
- Benign paroxysmal tonic upgaze
- Episodic ataxias
- Alternating hemiplegia
- Hyperekplexia
- Opsoclonus-myoclonus syndrome

- Autoimmune neurological disorder that may be seen in association with neuroblastoma, following viral infections
- The earliest feature is often ataxia followed by opsoclonus, followed by myoclonus.

Episodic syndromes that may be associated with migraine

- Recurrent gastrointestinal disturbance
  - Cyclical vomiting syndrome
  - Abdominal migraine
- Benign paroxysmal vertigo
- Benign paroxysmal torticollis

- Recurrent episodic attacks of intense nausea and vomiting, usually stereotypical in the individual and with predictable timing of episodes.
- Attacks may be associated with pallor and lethargy.
- There is complete resolution of symptoms between attacks.
- Boys and girls are equally affected
- The usual age of onset is 5 years
- Typically a self-limiting episodic condition occurring in childhood, children will “outgrow” these attacks by age 10

Episodic syndromes that may be associated with migraine

- Recurrent gastrointestinal disturbance
  - Cyclical vomiting syndrome
  - Abdominal migraine
- Benign paroxysmal vertigo
- Benign paroxysmal torticollis

- Recurrent brief attacks of vertigo, occurring without warning and resolving spontaneously, after minutes to hours without loss of consciousness, in otherwise healthy children.
- Associated with at least one of the following:
  - nystagmus
  - ataxia
  - vomiting
  - pallor
  - fearfulness

Episodic syndromes that may be associated with migraine

- Recurrent gastrointestinal disturbance
  - Cyclical vomiting syndrome
  - Abdominal migraine
- Benign paroxysmal vertigo
- Benign paroxysmal torticollis

- Recurrent episodes of head tilt to one side, perhaps with slight rotation, which remit spontaneously.
- The condition occurs in infants and small children, with onset in the first year.
- Associated with at least one of the following:
  - pallor
  - irritability
  - malaise
  - vomiting
  - ataxia

MISCELLANEOUS EVENTS

- Benign myoclonus of infancy and shuddering attacks
- Jitteriness
- Sandifer syndrome
- Non-epileptic head drops
- Spasmus nutans
- Paroxysmal extreme pain disorder
- Spinal myoclonus

- This syndrome is seen in young children with gastro-oesophageal reflux (with or without vomiting).
- Events are often seen with or after feeding.
- Typically, there is arching of the back, dystonic posturing of the limbs and turning/tilting of the head.

Outline

- **Epilepsy**
  - definition and new classification
- **Epilepsy imitators**
- **Headache**

HEADACHES

ICHD – 3 classification

Part ONE : Primary headaches

1. Migraine

1. Migraine without aura
2. Migraine with aura
3. Chronic migraine
4. Complications of migraine
5. Probable migraine
6. Episodic syndromes that may be associated with migraine

2. Tension-type headache

1. Infrequent episodic tension-type headache
2. Frequent episodic tension-type headache
3. Chronic tension-type headache
4. Probable tension-type headache

3. Trigeminal autonomic cephalalgias

1. Cluster headache
2. Paroxysmal hemicrania
3. Short-lasting unilateral neuralgiform headache attacks
4. Hemicrania continua
5. Probable trigeminal autonomic cephalalgia

4. Other primary headache disorders

1. Primary cough headache
2. Primary exercise headache
3. Primary headache associated with sexual activity
4. Primary thunderclap headache
5. Cold-stimulus headache
6. External-pressure headache
7. Primary stabbing headache
8. Nummular headache
9. Hypnic headache
10. New daily persistent headache (NDPH)

Part two: the secondary headaches

5. Headache attributed to trauma or injury to the head and/or neck
6. Headache attributed to cranial or cervical vascular disorder
7. Headache attributed to non-vascular intracranial disorder
8. Headache attributed to a substance or its withdrawal
9. Headache attributed to infection
10. Headache attributed to disorder of homoeostasis
11. Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure
12. Headache attributed to psychiatric disorder

Part three: painful cranial neuropathies, other facial pains and other headaches

13. Painful cranial neuropathies and other facial pains
14. Other headache disorders

Headache in children

- Headache is the most common reason that children are referred to child neurology practices
- The prevalence of headache ranges from 35-50 % in 7-yo children, gradually rising to 60-80% by age 15
- The prevalence of migraine headache steadily increases through childhood and the male: female ratio shifts during adolescence.

Migraine in children :

- Recurrent headache disorder manifesting in attacks lasting 2-72 hr.
- Typical characteristics of the headache are unilateral location, pulsating quality, moderate or severe intensity, aggravation by routine physical activity and association with nausea and/or photophobia and phonophobia.
- The prevalence of migraine rises from 3% at age 3–7 years to 4-11% by age 7–11, and up to 8–23 % during adolescence.
- The mean age of onset of migraine is 7 yrs for boys and 11 yrs for girls.

Migraine in children :

Migraine

1. Migraine without aura
2. Migraine with aura
3. Chronic migraine
4. Complications of migraine
5. Probable migraine

1.2.1 Migraine with typical aura (visual, sensory, motor ....)

2. **Migraine with brainstem aura (vertigo, nausea, diplopia ....)**
3. **Hemiplegic migraine**
4. **Retinal migraine**

1.6 Episodic syndromes that may be associated with migraine

Migraine in children :

Migraine

1. Migraine without aura
2. Migraine with aura
3. Chronic migraine
4. Complications of migraine
5. Probable migraine

1.2.1 Migraine with typical aura (visual, sensory, motor ....)

2. **Migraine with brainstem aura (vertigo, nausea, diplopia ....)**
3. **Hemiplegic migraine**
4. **Retinal migraine**

1.6 Episodic syndromes that may be associated with migraine

Migraine in children :

Migraine

1. Migraine without aura
2. Migraine with aura
3. Chronic migraine
4. Complications of migraine
5. Probable migraine

Headache occurring on 15 or more days per month for more than three months, which, on at least 8 days per month, has the features of migraine headache

1.6 Episodic syndromes that may be associated with migraine

Migraine in children :

Migraine

1. Migraine without aura
2. Migraine with aura
3. Chronic migraine
4. Complications of migraine
5. Probable migraine

1. Status migrainosus (> 72 HR)
2. Persistent aura without infarction
3. Migrainous infarction
4. Migraine aura-triggered seizure

1.6 Episodic syndromes that may be associated with migraine

Migraine in Children- management

Medical treatment (abortive and preventive)

- Complementary and Alternative Treatments

The abortive (symptomatic) therapy

- Analgesics (Acetaminophen, Ibuprofen …)
- Triptans
- Ergotamine drugs

Migraine in Children- management

- ANTIDEPRESSANT AGENTS (Amitriptyline ……)
- ANTIEPILEPTIC AGENTS (Topiramate, Valproic acid, Levetiracetam ….)
- ANTIHYPERTENSIVE AGENTS (Propranolol, Clonidine …..)
- CALCIUM CHANNEL BLOCKERS (Nimodipine ….)
- ANTIHISTAMINES (Cyproheptadine …..)

Tension-type headache

Typically bilateral, pressing or tightening in quality and of mild to moderate intensity, lasting minutes to days.

The pain does not worsen with routine physical activity and is not associated with nausea, but photophobia or phonophobia may be present.

Tension-type headache

2. Tension-type headache

1. Infrequent episodic tension-type headache → At least 10 episodes of headache occurring on <1 day per month
2. Frequent episodic tension-type headache → At least 10 episodes of headache occurring on 1-14 days per month
3. Chronic tension-type headache → ≥15 days per month
4. Probable tension-type headache

Headache in children

Red flags (possible secondary headache)

-Side locked headache

-Headache getting worse with laying down, with Valsalva, with cough or exercise.

-Headache getting worse with standing upright

- Headache waking the patient from sleep.

-New-onset headaches with accompanying features suggestive of meningitis or encephalitis

-Focal neurologic symptoms (eg, seizure, weakness, altered mental status, visual field defect).

-Abnormal exam: focal neurological deficits, papilledema, hypertension, etc.

-Immune compromised patients.

Secondary Headache

-Brain Tumor.

-Brain bleeding: subdual hematoma, subarachnoid hemorrhage.

-Idiopathic Intracranial Hypertension (IIH).

-Cerebral venous sinus thrombosis.

-Concussion.

-Infections: meningitis, encephalitis.

-TMJ problem, temporal arteritis, sinusitis.