Pediatric Neurology

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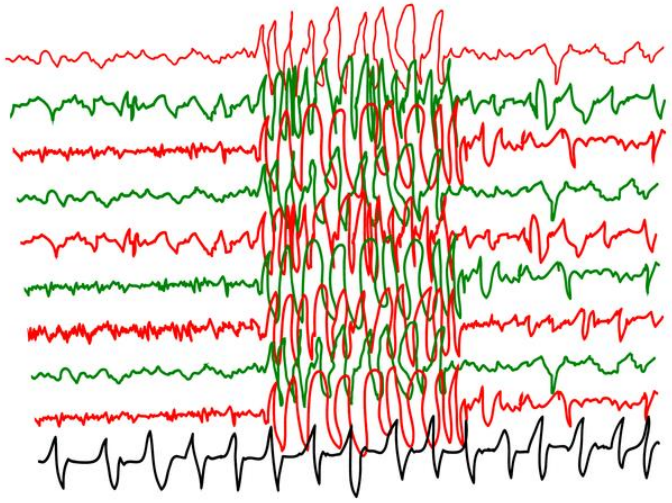
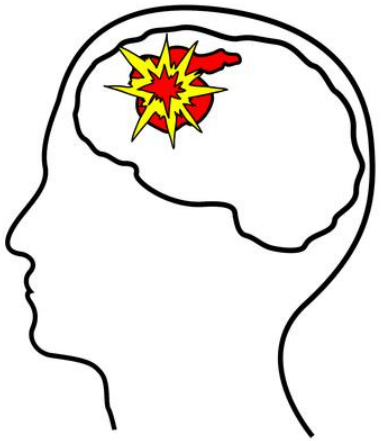
Outline

- **Epilepsy and related topics**
- **Epilepsy imitators**
- **Headache**





Spells

- Onset of spells.
- The spell description. Ask if there is a video.
- If not mentioned, we ask more specific questions about:
  - abnormal tone (going limp or tensing up).
  - abnormal eye movements.
  - change in color.
  - Altered level of awareness.
  - Can the spell be interrupted?
- Any specific symptoms before or after the spells, abrupt vs gradual onset and end.
- Duration of the spell (range if more than one spell).
- Frequency of the spells and do they cluster.
- Possible triggers (stress, crying, injuries, etc.).
- Specific times of occurrence (relationship to meals, position, sleep-wake transition).
- Do the spells happen during sleep.
- Any change in behavior, or developmental regression since the spells started.
- Recent events, trauma, serious illnesses, new medications, social changes.
- The rest of the history (perinatal history, developmental history, past medical and surgical history, family history, and social history).

Evaluation of seizures

- **History and physical exam**
- Look for an underlying (provoking) factor and treat accordingly: Hypoglycemia, hypocalcemia, hyponatremia, head injury, drug ingestion, infection, tumor, etc.

-Work up:

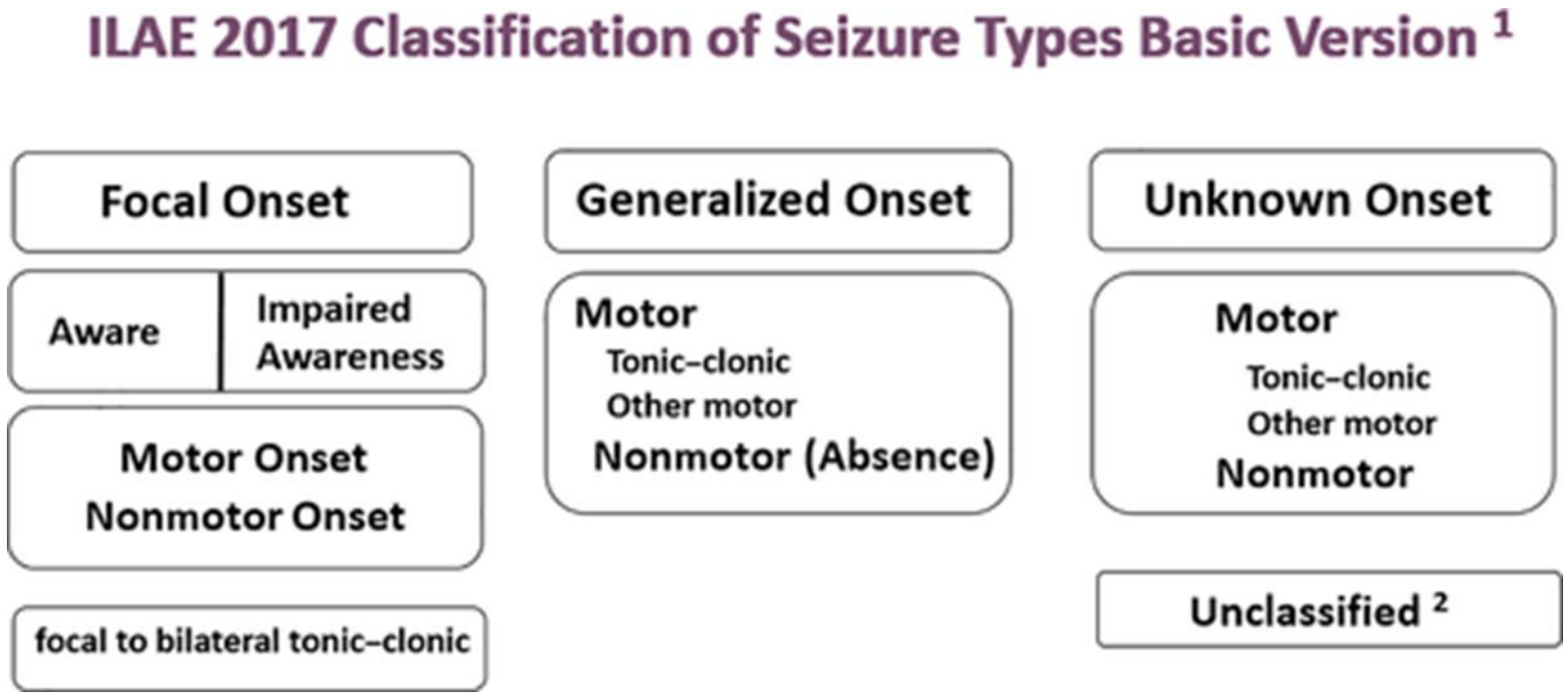
1) If unprovoked, will need an **EEG** for risk stratification.
   - One unprovoked seizure+ normal EEG: risk of recurrence around 40 %
   - One unprovoked seizure+ abnormal EEG: risk of recurrence around 60 %

2) If concern for focality per history or exam, will need brain imaging, specifically a brain MRI.

3) Other tests to consider: genetic, metabolic, CSF studies ….etc

Seizure

-Definition: a transient occurrence of signs or symptoms resulting from abnormal excessive or synchronous neuronal activity in the brain.



Epilepsy

-Definition: 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

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

STEP 1

Seizure type

- Focal onset
- Generalized onset
- Unknown onset

STEP 2

Epilepsy type

- Focal
- Generalized
- Combined Generalized & Focal
- Unknown

Etiology

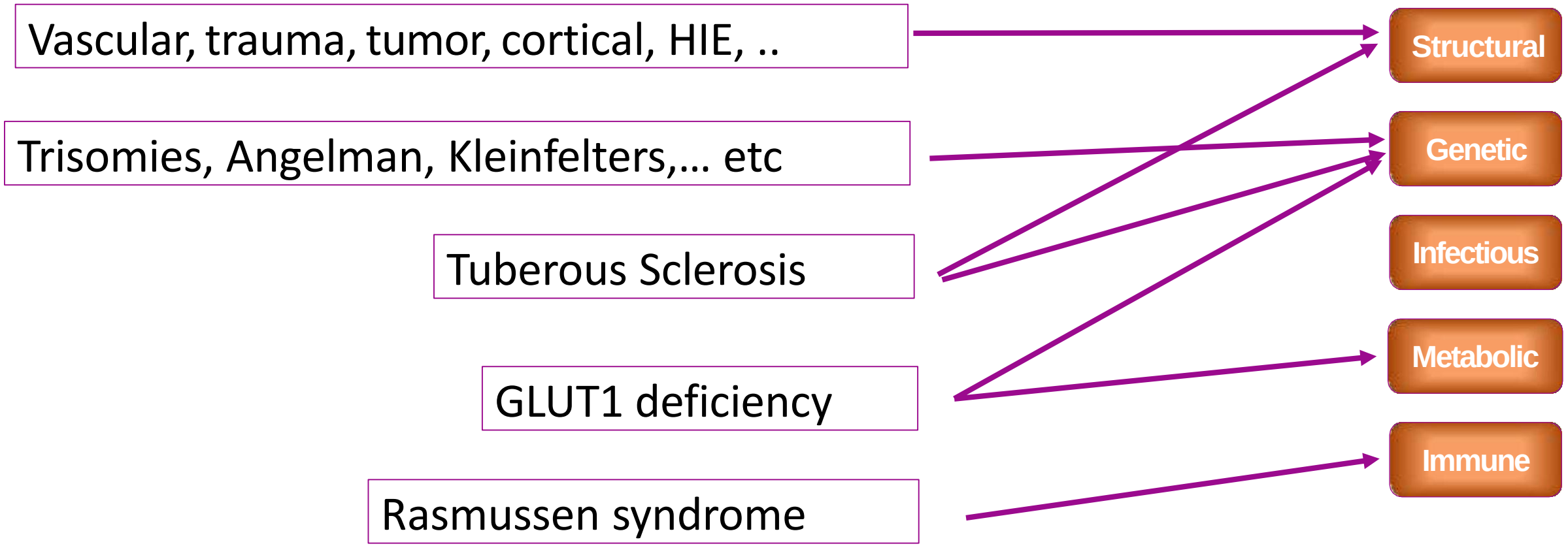
STEP 3

- Structural
- Genetic
- Infectious
- Metabolic
- Immune

STEP 4

Epilepsy Syndromes

Etiology


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

Epilepsy Syndromes

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
- Autosomal-dominant nocturnal frontal lobe epilepsy
- Benign epilepsy with centrotemporal spikes
- Panayiotopoulos syndrome
- Epilepsy with myoclonic atonic seizures
- Epilepsy with myoclonic absences
- Lennox-Gastaut syndrome

Adolescent / adult

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

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.

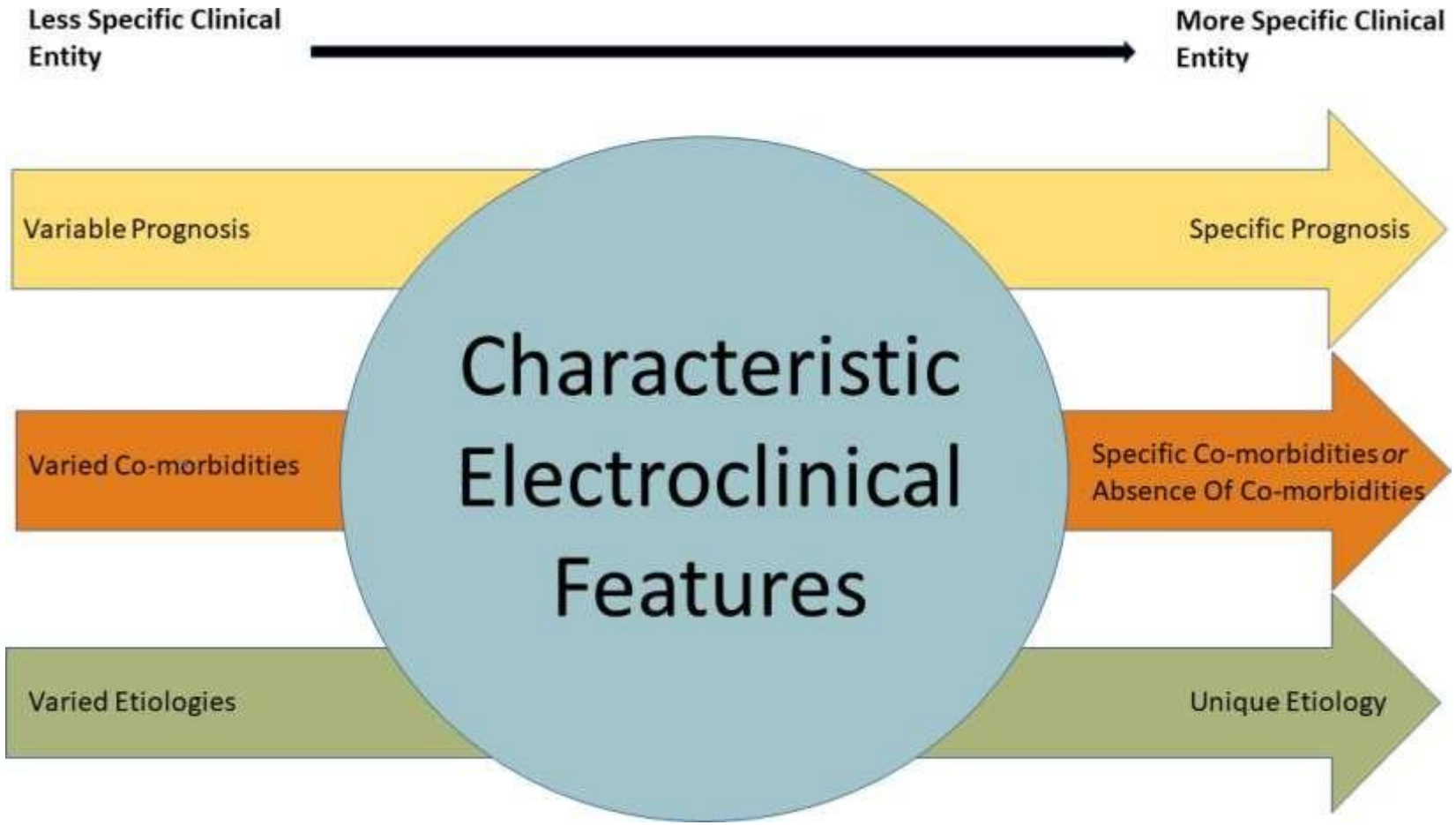
ILAE 2017

-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.

ILAE 2022 epilepsy syndrome



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

-Based on the 2017 Classification of Seizures and Epilepsies, some syndrome names have been updated using terms directly describing the seizure semiology.

-This designation frequently carries prognostic and treatment implications.

-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 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

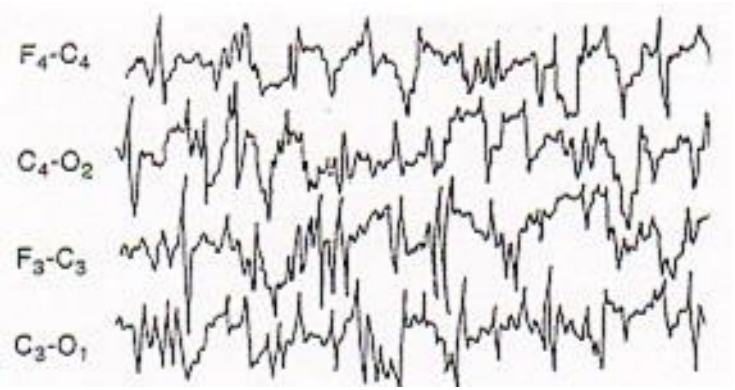
Age + seizure semiology + EEG pattern

1) Infantile spasms (IS)
2) Lennox Gastaut syndrome (LGS)
3) Childhood Absence Epilepsy (CAE)
4) Benign epilepsy with centrotemporal spikes (BECTS)
5) Juvenile Myoclonic epilepsy (JME)

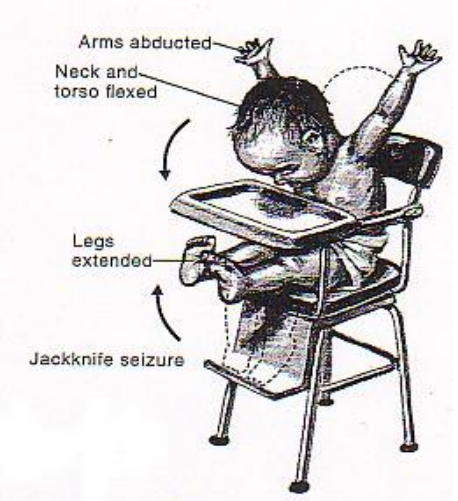
Infantile spasms (West syndrome)

-Infantile age onset

- (Age: infancy + sz: spasms + EEG: Hypsarrhythmia). Severely abnormal EEG pattern: disorganized, discontinuous, high amplitude, multifocal spikes called hypsarrhythmia.



-Treatment: steroids (ACTH, Prednisone) or Vigabatrin.



- Prognosis variable depending on underlying etiology. Frequently leads to dev delay. Earlier treatment leads to better outcome.

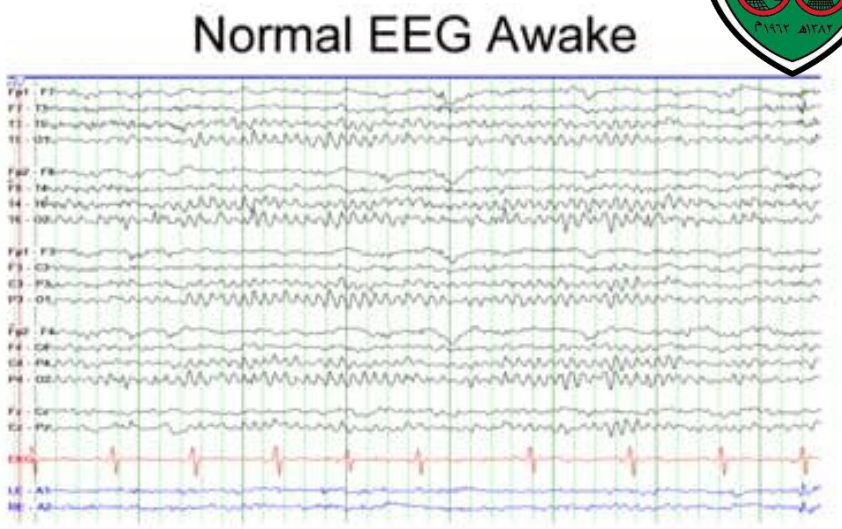
Lennox Gastaut syndrome:

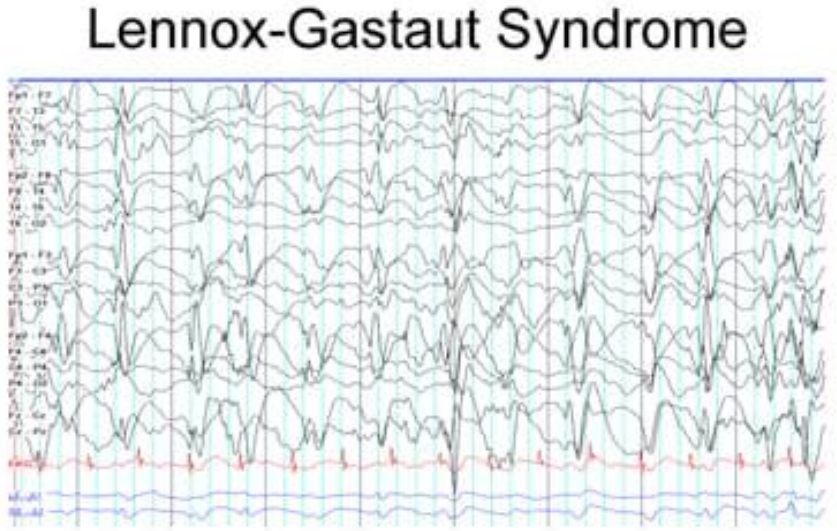
<u>-Childhood age onset.</u>

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

- **Treatment:** Valproate, Clobazam, Lamotrigine.

- Refractory to treatment. Typically associated with significant intellectual dysfunction (epileptic encephalopathy).

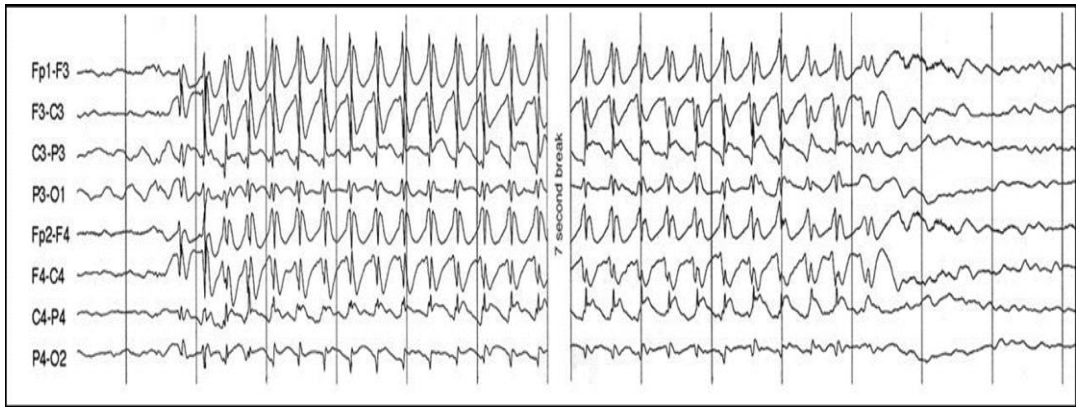




Childhood Absence Epilepsy (CAE)

-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.



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

-Good prognosis, typically resolves by adolescence.

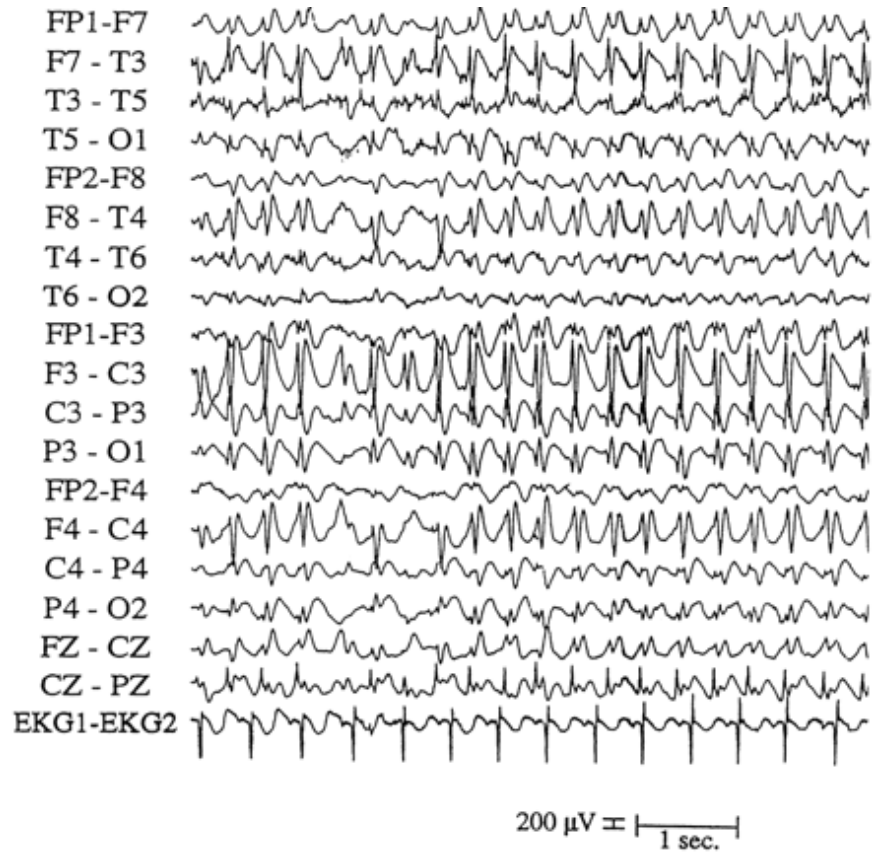
Benign Rolandic Epilepsy (BECTS)

-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.

- Treatment: Carbamazepine, oxcarbazepine.

- Good prognosis, resolves by puberty.



Juvenile Myoclonic epilepsy (JME)

- Adolescent age onset.

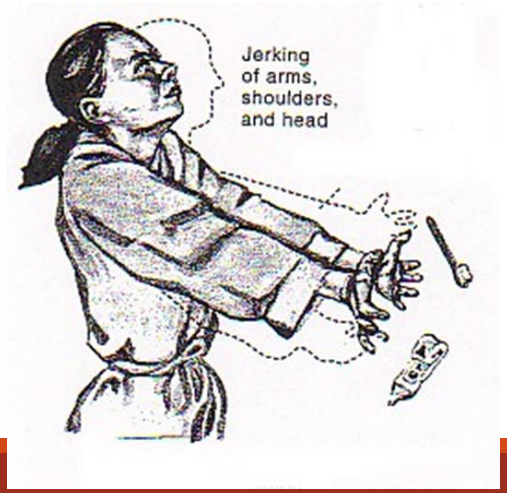
- (Age >12 yr + Sz: myoclonic sz (Kelog's sz)+ EEG: fast spike wave 4-6 hz).

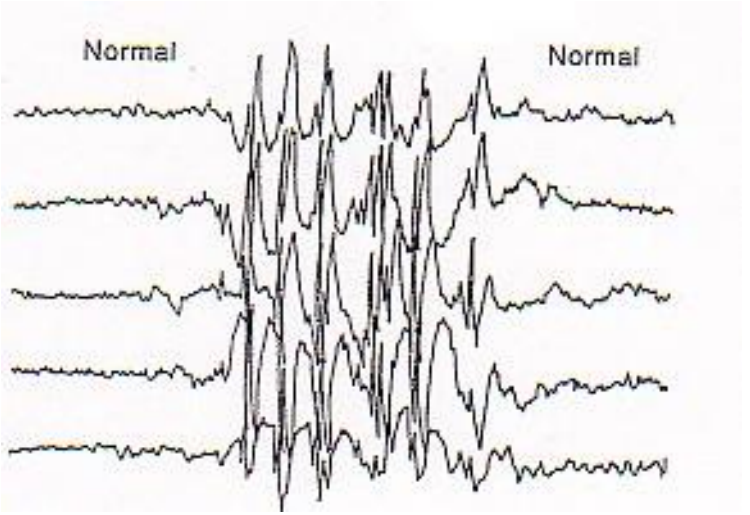
- Mostly affects arms, typically upon awakening.

- Sleep deprivation and flashing lights may provoke seizures.

- **Treatment**: Lamotrigine, Valproate.

- Life-long treatment .





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

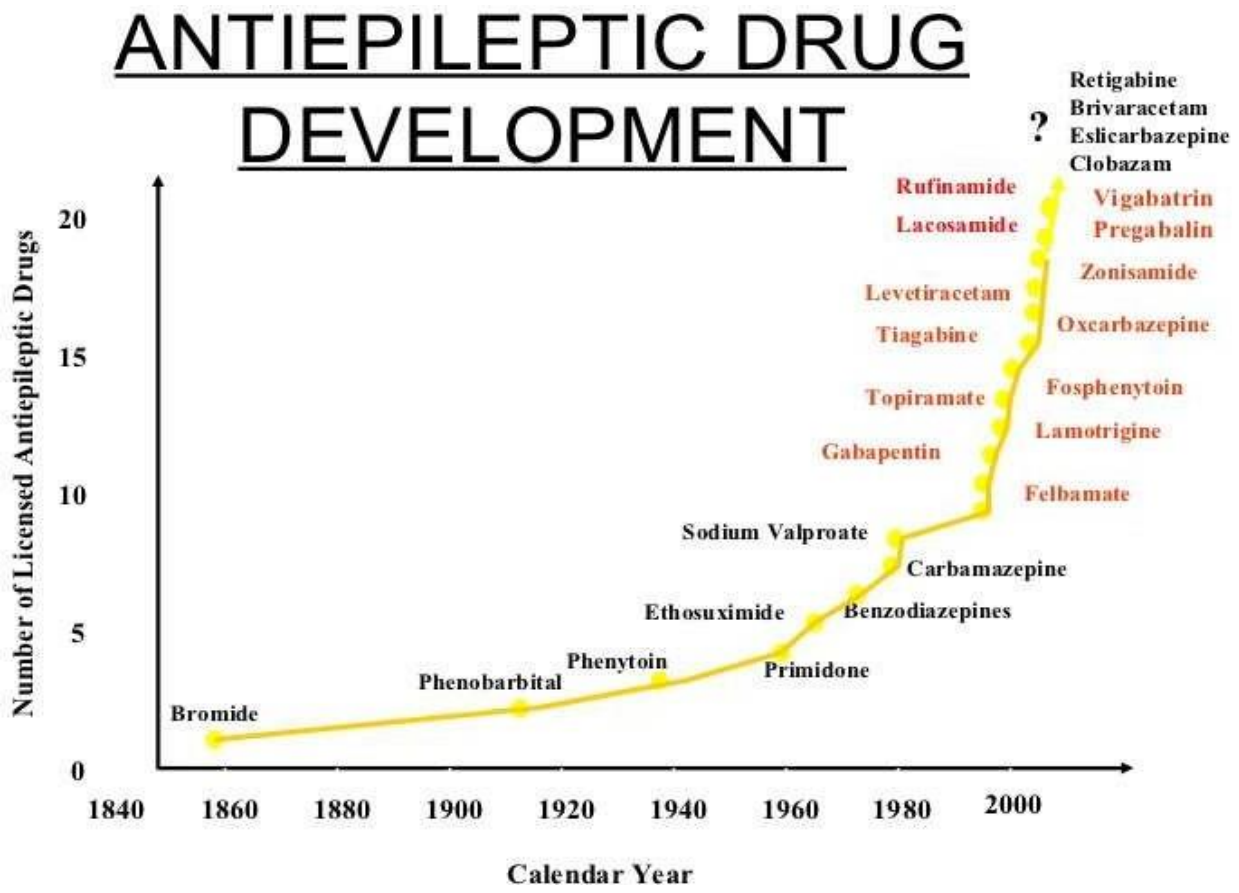
1) Antiseizure medications:

- Anti-seizure medication: decide based on type of epilepsy, and associated co-morbidities.
- Aim for the least number of medications, and lowest effective dose, to minimize side effects.
  - - 70% become seizure-free on monotherapy
  - - an additional 15% become seizure free on polypharmacy
  - -15% remain intractable (fail 2 or more medications) .

2) Other medications

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

Antiseizure medications



Antiseizure medications

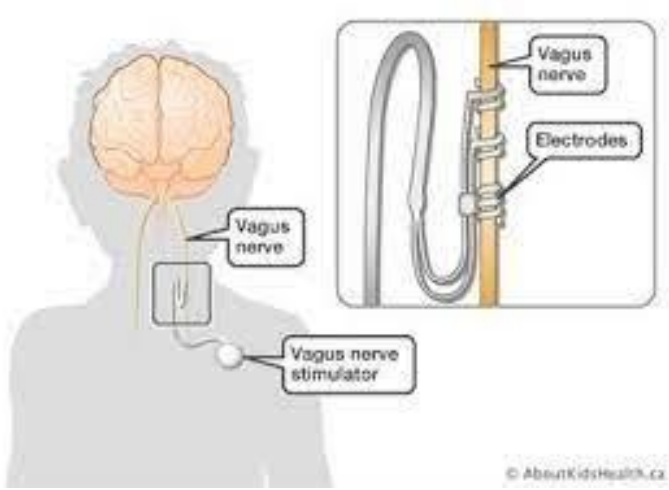
- DRUGS THAT AFFECT VOLTAGE-DEPENDENT SODIUM CHANNELS
  - Old : Carbamazepine, Phenytoin.
  - New : Lamotrigine, Oxcarbazepine, Zonisamide, Lacosamide, Rufinamide.
- DRUGS THAT AFFECT CALCIUM CURRENTS
  - Ethosuximide.
- DRUGS THAT AFFECT GABA ACTIVITY
  - Benzodiazepines, Barbiturates, Gabapentin.
- 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.

Treatment options for Drug Resistant Epilepsy (DRE):

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



-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 to affect patient's development in young children.

Status epilepticus

- It is a condition resulting either from the failure of the mechanisms responsible for seizure termination or from the initiation of mechanisms that 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

Status epilepticus

-Definition: ongoing seizure activity for more than 5 minutes, or recurrent seizures with no return to baseline for more than 30 minutes.

-Evaluation and treatment:

- Initial assessment: ABC's
- Check serum glucose.
- Vital signs (temperature,..)
- Treat with IV meds:
  - 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
- Determine if other tests are needed based on clinical picture (electrolytes , imaging, etc.)
- Possible etiologies: medication noncompliance, infection, meningitis, drug toxicity, head injury, etc.

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). More common in preterm babies.
  - 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
- (must be considered at any age)

Any Age

- Pyridoxine deficiency

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 - Treatment

- Treat the underlying cause (infection, electrolyte imbalance, etc)
- Anti-seizure medications:
  - Phenobarbital (1st line)
  - Phenytoin and Fosphenytoin
  - Levetiracetam
  - Lorazepam
  - Diazepam, midazolam

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.
- 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

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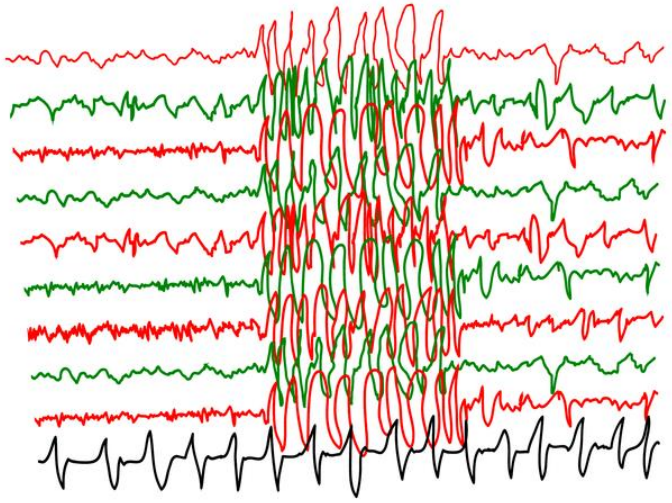
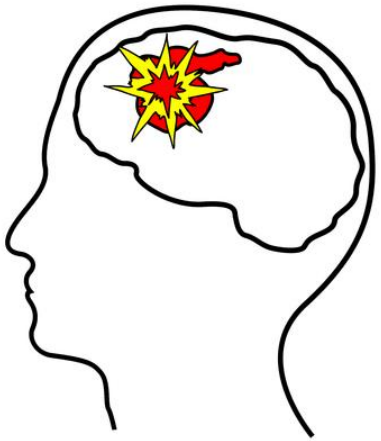




EPILEPSY IMITATORS

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 to minutes
- Preceded by triggers
- Patient is upright, lightheadedness, pallor, sweating, etc
- Convulsive movements occur in 50%
- Positive Fm. Hx is common

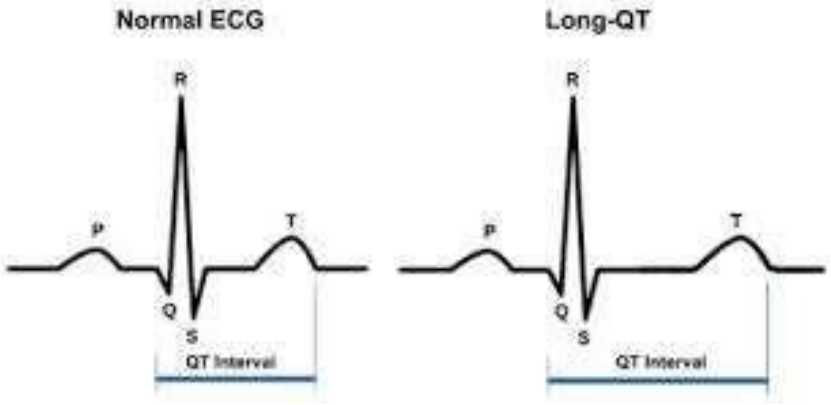
SYNCOPE AND ANOXIC SEIZURES

- 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

- Affects pre-school children
- Start with crying then stop breathing in expiration.
- Becomes blue with deep cyanosis, or become pale
- They breath in or go to transient syncope, tonic posturing is possible
- Worse if child has iron deficiency anemia. Check Ferritin.

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



- 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 cardiac syncope.

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 dis
- Fabricated / factitious illness

- 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

-Healthy newborn, normal exam.
-myoclonic jerks happen only during sleep.
- Resolve without intervention

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 tends to wax and wane in frequency over time.
- Urge to perform the tic, and an ability to suppress the tic support the diagnosis.

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- Semi-voluntary, non-rhythmic, mostly involves arms and legs more than the face.
- Starts in early childhood and tends to persist.
- No urge. Can be interrupted by distraction.
- Common in patients with autism and developmental delay.

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- Characterized by an exaggeration of the normal startle response.
- Evident from the neonatal period or early infancy.
- Responds to benzodiazepines
- Can be familial.

Episodic syndromes that may be associated with migraine

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

- Recurrent episodes of intense nausea and vomiting, usually stereotypical with predictable timing.
- Episodes may be associated with pallor and lethargy.
- Boys and girls are equally affected
- Usual age of onset is 5 years
- Typically a self-limiting condition, that resolves by age 10

Episodic syndromes that may be associated with migraine

- Recurrent gastrointestinal disturbance
  - Cyclical vomiting
  - 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: nystagmus, ataxia, vomiting, pallor, fearfulness

MISCELLANEOUS EVENTS

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

- Typically seen in young children with gastro-esophageal reflux (GERD).
- 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 and related topics
- Epilepsy imitators
- Headache

ICHD – 3 Headache 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)

ICHD – 3 Headache classification

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.

Primary Headache

1) Migraine:

- affects 5% of children. Prevalence increases steadily through childhood. Mean age is 7 yrs for boys and 11 yrs for girls.

- Affects males and females equally in young age, then shifts to female predominance during adolescence.

- **Pathophysiology** in children is similar to adults. Calcitonin gene-related peptide (CGRP) levels are elevated during migraine attacks.

- **Symptoms:** throbbing headache lasting hours to days, nausea, vomiting, photophobia, phonophobia, anorexia, fatigue, irritability/mood change. May be preceded by an aura. Not necessarily unilateral.

Primary Headache

1) Migraine:

- Management:

- **- Lifestyle modifications:** Regular sleep. Regular healthy diet. Regular exercise. Good hydration. Avoid caffeine.
- **- Cognitive behavioral therapy**
- **- Medications:**
  - Rescue medications: Acetaminophen, NSAIDs, Prochlorperazine, Triptans.
  - Prophylactic medications: Propranolol, Topiramate, Amitriptyline, Riboflavin.

Primary Headache

2) Tension headache :

- affects 5-10% of children.

- Affects males and females equally in young age.

- Generally, not as disabling as migraine.

- **Symptoms:** headache lasting hours to days, typically bilateral, pressing or tightening in quality and of mild to moderate intensity, lasting minutes to days. Otherwise “featureless”.

Primary Headache

2) Tension headache :

- **Management:**
  - - Lifestyle modifications.
  - - Cognitive behavioral therapy
  - - Medications:
    - Rescue medications: Acetaminophen, NSAIDs.
    - Prophylactic medications: usually not needed.

Secondary Headache

Red flags

-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.

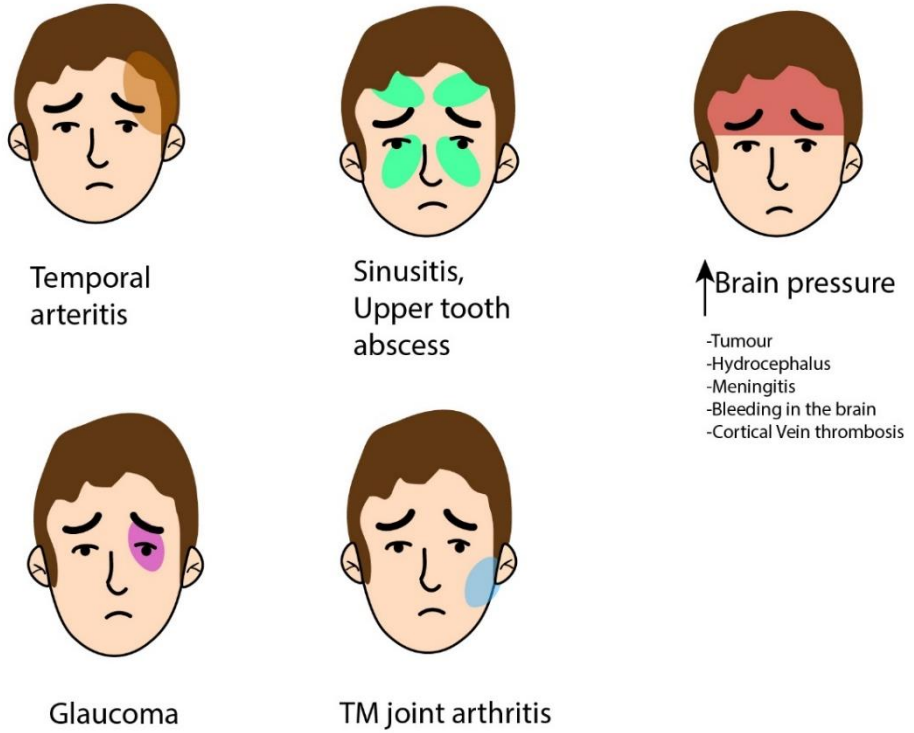
-Idiopathic Intracranial Hypertension (IIH).

-Cerebral venous sinus thrombosis.

-Concussion.

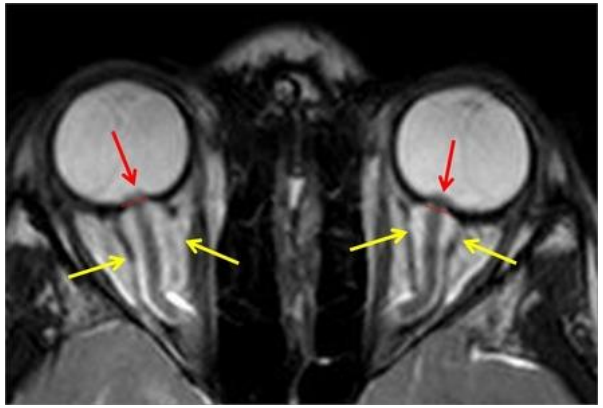
-Infections: meningitis, encephalitis.

-TMJ problem, temporal arteritis, sinusitis.



IIH imaging findings

papilledema,



Empty sella,

slit-like ventricles