SEIZURE DISORDER

Objectives:

- To know that seizures (ZT's) a symptom NOT a disease
- To know clinical observation crucial for Dx, classification, and Rx.
- R/O other paroxysmal, non-epileptic disorders.
- To know acute management & prevention of recurrence.
- To know thoughtful & rational patient work-up
- To know optimum use of anti-epileptic drugs (AED's)
- To know comprehensive patient (not SZ's) management.

Sources: Dr.Albashiri's slides and notes, illustrator

Definitions:

- **Seizures**: are the clinical manifestation of aberrant, abnormal electrical activity in the cortical neurons.
    - It is a symptom of cerebral pathology, not a disease

- **Epilepsy:** is a chronic disorder characterized by the tendency for spontaneous, recurrent seizures and requires at least **two unprovoked** seizures to be considered as a diagnosis.
- **Epileptic Syndrome:** a complex of symptoms and signs that define a unique epilepsy condition. (e.g L.G.S Lennox-Gastaut syndrome)
- **Epileptic Disorder:** A chronic neurological condition characterized by recurrent epileptic seizures.
- **Non-epileptic seizures (NES):** is a descriptive term for a group of disorders that refer to paroxysmal events that can be mistaken for epilepsy, but are not due to an epileptic disorder.

Epidemiology:

- Seizure is the most common neurological disorders
- Seizures occur in 3% to 5% of all children
- Febrile seizures occur in 2% to 4% of the pediatric population .
- Epilepsy occurs in approximately 1%.

History Taking:

Questions regarding the possible seizure can be divided into :

Pre-ictal	Ictal	Post-ictal
• Was there any warning before the spell? If so, what was the warning? • Did the child complain of abdominal discomfort, fear or any other unpleasant sensations before the spell? • What was the child doing before the spell? • Was the child asleep or awake prior to the event? • Was the child sleep deprived prior to the spell? • Were there any triggers for the spell? • Was the child well before the spell or was there a fever or illness?	• Did any body movements occur? • Was there any perioral cyanosis? • Did the patient lose consciousness during the spell? • How long did the spell last? • How many episodes? • How often do the spells occur?	• How did the patient feel after the spell? • Did the child seem confused and tired after the spell? • How long did it take for the child to get back to baseline condition? • Did the child suffer from a headache after the spell? • Was there any weakness noticed ?

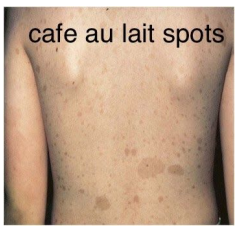
Other questions to ask:

- Has the child ever had any seizures before?
- Is there any history of febrile seizures?
- Ask about past medical history
- Pregnancy and birth history
- Developmental history
- Current medications
- Is there any family history of seizures?

Physical Examination:

A complete pediatric exam.

- Pay attention to the following elements :
    - Vitals, including temperature
    - Height, weight and head circumference - plot on a growth chart to determine percentiles
    - Developmental stage of child in gross motor, fine motor, language and social domains
- Signs of trauma.
- Signs of increased intracranial pressure
- Skin lesions may suggest a neurocutaneous disease
- Special tests:
    - Fundoscopy
    - Neurologic exam: looking for focal deficits indicates symptomatic seizure.



Seizure Categorization:

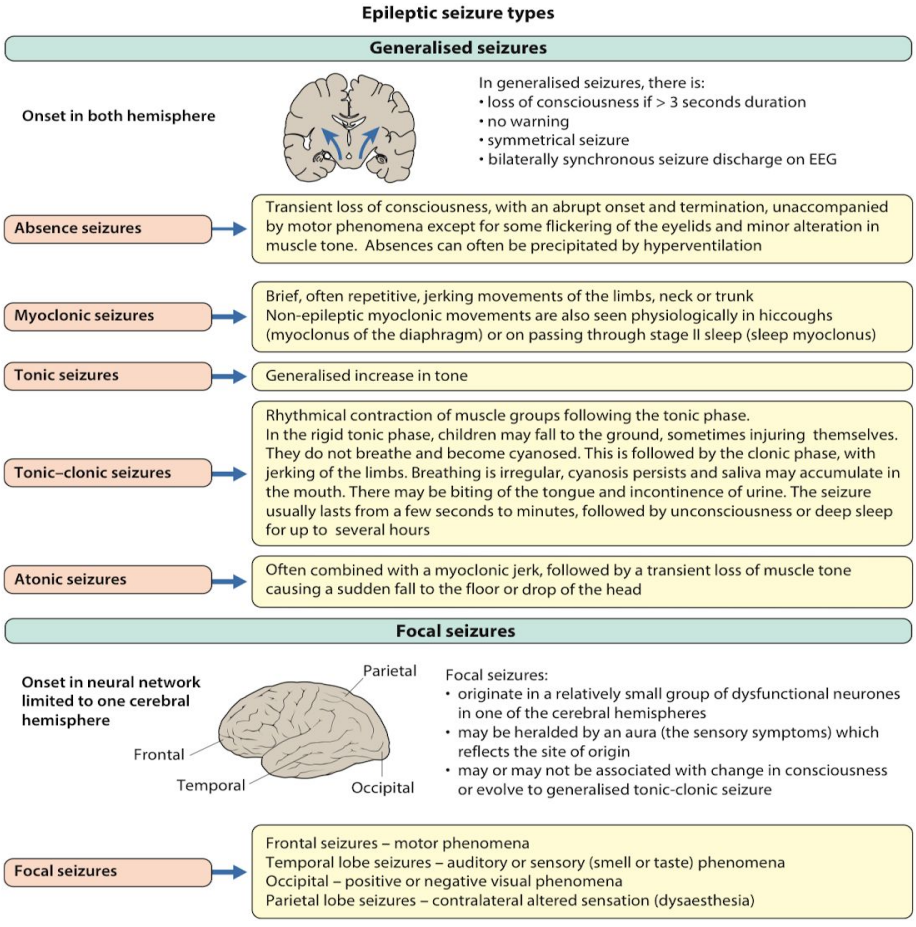
★ After taking history and exam, you will be able to categorize the seizure which is important for determining treatment and prognosis.

We need to answer two main questions:

1. Is the seizure focal or generalized?
2. Is the seizure simple or complex? Is there impairment in consciousness?

Classification of Epileptic Seizures: old classification but we still use it very often

1. **Focal(partial) Seizures: NO loss of consciousness**
   a. Simple partial seizures (motor, sensory, hallucinations, autonomic psychic symptoms)
   b. Complex Partial Seizures (impaired consciousness)
   c. Partial seizures with secondary generalized.
2. **Generalized seizures: Complete LOSS of consciousness**
   a. Absence seizures (typical and atypical)
   b. Myoclonic seizures
   c. Clonic Seizures
   d. Tonic seizures
   e. Tonic-clonic seizures
   f. Atonic seizures
3. **Unclassified seizures**



Auro with temporal epilepsy, while frontal lobe epilepsy never have aura

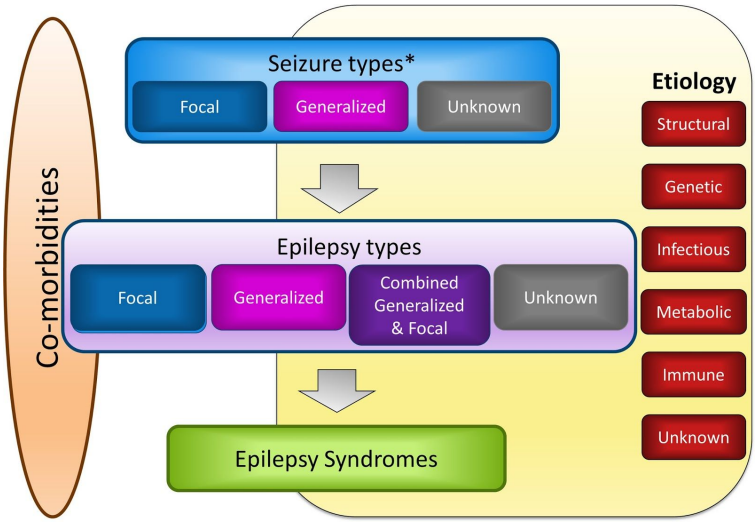
International classification of epilepsies ,epileptic syndrome and related seizure disorders New classification

- **Localization related** (focal ,local, partial)
- **Generalized epilepsies and syndromes**
- **Undetermined epilepsies:**
    - With both generalized and focal seizure:
        - Neonatal seizure
        - Severe Myoclonic Epilepsy of Infancy (SMEI)
        - Epilepsy with continuous Slow – wave during sleep
        - Landau – Kleffner Syndrome (LKS)
    - Without unequivocal generalized/focal features

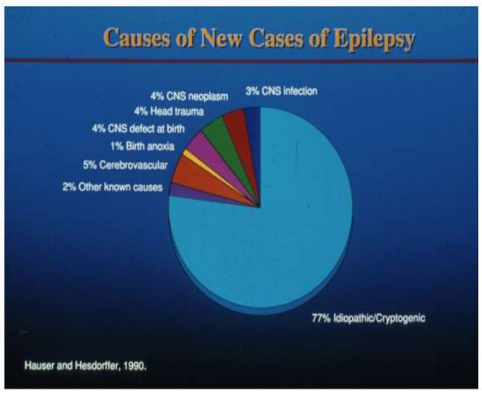
- **Special syndromes:**
    - Conditions with epileptic seizure that do not require a diagnosis of epilepsy
    - Situation related seizure:
        - febrile convulsion
        - isolated seizure or status epilepticus
        - seizure occurring only with acute /toxic event

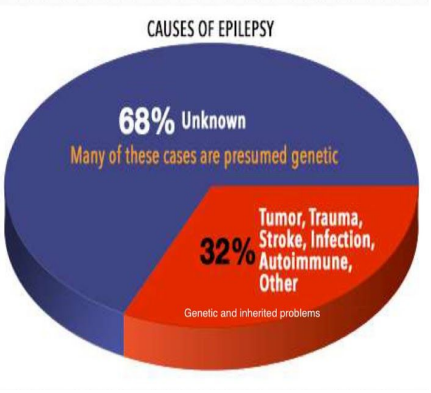
Epilepsy Classification framework: ***IMPORTANT***

You have to take the child as a whole



Causes of Epilepsy:





Differential Diagnosis:

- Syncope
- Breath holding spell
- GERD
- Panic attack
- Daydreaming
- Conversion or Non Epileptic seizure
- Benign sleep myoclonus
- Benign paroxysmal vertigo
- Complicated migraine
- Motor tics Investigations

Investigations: **Epilepsy is a clinical diagnosis**

- Blood tests: In general, all patients should have acute symptomatic causes of seizures ruled out:
    - CBC and differential
    - Blood glucose level
    - Electrolytes
    - Sodium
    - Calcium
    - Magnesium
    - Phosphorus Investigations

- Additional tests need to be considered :
    - Hemorrhagic basis : INR, PTT
    - Toxic basis : blood levels of suspected drugs and metabolites
    - Genetic disease : possible karyotype and other tests specific to illness
    - Metabolic disease :tests specific to disease, may include: Ammonia - Lactate Pyruvate Amino acids - Urine organic acids

- Lumbar puncture : AAP Recommendation:
    - If the child has fever :
        - Less than 12 months Strongly considered (because their meningeal signs very subtle and it's difficult to identify it)
        - Less than 18 months considered
    - ✓ Any child with meningeal signs Investigations

- Neuro - Imaging:
    - CT scan - indicated if head trauma is present/suspected
    - MRI - indicated if the child has focal seizure or focal neurological deficits , Signs of high ICP Investigations
- **The EEG** is recommended as part of the neurodiagnostic evaluation of the child with an apparent first unprovoked seizure. However, An EEG abnormality by itself is not sufficient to make a diagnosis of epileptic seizure nor its absence rule out a seizure.

Treatment :

- Choices of anti-epileptic medications
- Start with small dose and build up slowly
- Explain to the parents possible side effects
- Educate the parent about how to manage the attack of seizure

Table 29.2 Choice of antiepileptic drugs (NICE guidelines 2014)

Seizure type	First-line	Second-line
Generalised		
Tonic-clonic	Valproate, carbamazepine*, lamotrigine	Clobazam, levetiracetam, topiramate
Absence	Valproate, ethosuximide, lamotrigine	Clobazam, levetiracetam, topiramate
Myoclonic	Valproate, levetiracitam, topiramate	Clobazam, piracitam
Focal seizures	Carbamazepine, valproate, levetiracetam, lamotrigine	Clobazam, topiramate, gabapentin*, tiagabine*

*Avoid with absence seizures, myoclonic seizures or juvenile myoclonic epilepsy.

Generalized think about valproate
Focal think about carbamazepine
If you understand this that's enough for your level

Anti-epileptic drugs may worsen specific seizures	
Antiepileptic drug	**Epileptic syndrome/seizure type**
Carbamazepine, vigabatrin, tiagabine, phenytoin	• childhood absence epilepsy • Juvenile absence epilepsy • Juvenile myoclonic epilepsy
Vigabatrin	• Absence and absence status
clonazepam	• Generalised tonic status in lennox gastaut syndrome
Lamotrigine	• Dravet's syndrome, • Juvenile myoclonic epilepsy

- **Is there any other modality of treatments?**
  - Epilepsy surgery
  - Vagal Nerve stimulation
  - Ketogenic diet

- **When do you discontinue AEDs ?**
  - Practice parameter by AAN American academy of neurology (1996)
  - Seizure-free 2-5 years on AEDs
  - Single type of seizures
  - Normal neurologic exam and normal IQ
  - EEG normalized with treatment

FEBRILE SEIZURE

Febrile Seizures:

- Affects 4% of children
- Age: 5 month to 6 years
- Usually generalized convulsion
- Duration: < 15 min
- Fever: >38.4
- No previous neonatal seizures or unprovoked seizures

Complex Febrile Seizures:

- Prolonged >15 min Partial onset
- Multiple recurrences within 24 hours
- **Febrile status epilepticus** is a seizure that lasts more than 30 min or series of seizures without full recovery in between

AAP Guidelines for seizures associated with fever:

- **Routine Serum Electrolytes**, Ca, Phos., Mg, CBC or glucose
    - limited value in the absence of suspicious history, or abnormal physical exam in infants older than 6 months
- **CT/MRI:** Not helpful. It might be considered in prolonged focal seizure with no clear etiology
- **EEG :** limited value in the evaluation of febrile seizures.

Recurrent Febrile Seizures:

- F/H of febrile seizure
- First febrile seizure before age of 18 months
- Low grade fever
- Brief interval(<1 hr) between onset of recognized fever and seizure

Febrile Seizures and subsequent Epilepsy:

- Preexisting neurodevelopmental abnormality
- Complex febrile seizures
- F/H of epilepsy is the strongest factor in developing epilepsy later on in life.
- A shorter duration of temperature before the first seizure
- A lower temperature at the time of the first seizure

Treatment of Febrile Seizures:

- Usually brief and self-limited
- Assurance and education
- Antipyretic agents can be used to reduce the fever however it will not affect the seizure recurrence or duration.

- It's a rule: febrile seizure is not an indication to give antiepileptic drugs but for prolonged seizure more than 5 minutes give PR diazepam
- Intermittent Diazepam PO or PR; for prolonged repetitive seizures; sedation could mask signs of meningitis
- Abortive treatment with rectal Diazepam; for prolonged repetitive seizures; more preferred modality of treatment

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Common Epilepsies in Children

Infantile Spasms (I.S):

- Age: < 1 year with peak at 4-7 months
- Spasms: flexor , extensor or mixed it came in cluster followed by irritability
- Usually they will have Developmental arrest or regression
- EEG: hypsarrhythmia characteristic
- **Identified causes in 75% of the cases:** Tuberous Sclerosis in 20%. Brain malformation and Hemorrhagic ischemic encephalopathy are the most common causes
- Idiopathic I.S has the best prognosis
- Treatment:
    - ACTH, Prednisone in idiopathic I.S
    - vigabatrin with tuberous sclerosis
    - Carpamizabine, valproate, pyridoxine

Childhood Absence Epilepsy:

- Age: 5-10 years
- Girls more than boys
- 30% of pt develop GTCs
- EEG : 3 Hz generalized spike and wave
- Remission: 80% after age of 10 years
- Treatment: First choice: **Ethosuximide** ياويلكم مني اذا نسيتوها, valproate
  Second choice: Lamotrigate,Topiramate

Benign Childhood Epilepsy with Centrotemporal Spikes (Rolandic epilepsy):

- Age: 5-9 years
- Remission: By 16 years of age
- Nocturnal seizures with sensory motor symptoms involving the face and oropharynx (majority during sleep however in minority they can have it during awake)
- EEG: has characteristic spikes coming from central and temporal
- Treatment: Benign they will grow ou not necessarily needs meds because it's not frequent just happens once or twice per year
    - **indication for Rx:** affects school performance, family anxiety, EEG abnormal, increased in frequency

شوفوا صراحة الجدول الي تحت موش بالسلايد بس بكتاب الليستريتد فحطيته بس عشان يرتاح ضميري فاذا ماتبون سووله سكيب

Table 29.1 Some epilepsy syndromes – arranged by age of onset

Name	Age of onset	Seizure pattern	Comments
Infantile spasms (West syndrome)	3–12 months	Violent flexor spasms of the head, trunk, and limbs followed by extension of the arms, last 1–2 s, often multiple bursts of 20–30, often on waking or many times a day. May be misinterpreted as colic. Social interaction often deteriorates – a useful marker in the history	Most have underlying neurological cause. EEG – hypsarrhythmia (Fig. 29.3). Treatment is vigabatrin and/or corticosteroids; good initial response in 60–70%, but unwanted side effects of therapy, and relapses common. Most will lose skills and develop learning disability and continuing epilepsy.
Lennox–Gastaut syndrome	1–3 years	Multiple seizure types, but mostly atonic, atypical (subtle) absences, and tonic seizures in sleep. Also neurodevelopmental arrest or regression and behaviour disorder.	Many causes, and often other complex neurological problems or history of infantile spasms. EEG shows slow generalized spike and wave (1–3 Hz). Prognosis is poor.
Childhood absence epilepsy	4–12 years	Momentary unresponsive stare with motor arrest, may twitch their eyelids or a hand or mouth minimally. Sudden onset, lasts only a few seconds (<30 s). Child has no recall except realises they have missed something and may look puzzled or say 'pardon' on regaining consciousness. Developmentally normal but can interfere with schooling. Accounts for only 2% of childhood epilepsy.	Two-thirds are female. Episodes can be induced by hyperventilation blowing on a piece of paper or windmill for 2–3 min; useful during EEG. The EEG shows fast generalised spike and wave (3–4 Hz) discharges, bilaterally synchronous during and sometimes between absences (Fig. 29.4). Prognosis good with 80% remission in adolescence; a few evolve into juvenile absence or juvenile myoclonic epilepsy.
Benign rolandic epilepsy (Benign epilepsy with centro-temporal spikes)	4–10 years	Tonic-clonic seizures in sleep, or simple focal seizures with awareness of abnormal feelings in the tongue and distortion of the face (supplied by the rolandic (centro-temporal) area of the brain).	15% of all childhood epilepsies. EEG shows focal sharp waves from the rolandic area. Important to recognise as relatively benign and may not require AEDs. Remits in adolescence.
Panayiotopoulos syndrome (Early-onset benign occipital epilepsy)	1–5 years	Autonomic features with vomiting and unresponsive staring in sleep, with head and eye deviation, progressing sometimes to a convulsive seizure.	Comprises 5% of childhood epilepsies. EEG shows posterior focal sharp waves and occipital discharges when eyes are shut. Remits in childhood. Some have specific learning difficulties.
Juvenile absence epilepsy	10–20 years	Absences, and generalised tonic-clonic seizures, often with photosensitivity. Learning is unimpaired.	Characteristic EEG. Response to treatment is usually good but lifelong. Remission unlikely.
Juvenile myoclonic epilepsy	10–20 years	Myoclonic seizures, generalized tonic-clonic seizures, and absences may occur, mostly shortly after waking. A typical history is throwing drinks or cereal about in the morning as myoclonus occurs at this time. Learning is unimpaired.	Characteristic EEG. Response to treatment is usually good but lifelong. Remission unlikely

Seizure mimickers

Paroxysmal Non-Epileptic Events (PNE)	
PNE With Alterations in Consciousness	PNEC Without Alterations in Consciousness
• Breath-holding Spells. • Syncope. • Hyperventilation. • Sleep-related phenomena. Apnea. • Somatoform disorders. • Munchausen syndrome by proxy.	• Stereotypy. • Gratification disorder. • Movement Disorders. • GERD. • Complicated Headaches. • Hyperekplexia. • Alternating Hemiplegia of Childhood. • Diaphragmatic Flutter. • Episodic Behavioral Syndromes. • Paroxysmal Extraocular Gaze Deviations. • Familial rectal pain syndrome.

Jitteriness:

- Common in the neonatal period.
- Observed as an excessive response to stimulation such as touch or loud noise.
- Baby is typically awake during the events and no associated autonomic disturbance.
- Can be lessened by removing the stimulus or relaxing the affected limb.
- Causes:
    - Mild jitteriness are common in healthy newborn.
    - Drug withdrawal.
    - Hypocalcemia.
    - Hypoglycemia.
    - Hypoxic ischemic encephalopathy

Apnea:

- Pause in breathing for more than 15 seconds.
- Non-epileptic apnea are associated with bradycardia.(No increase in HR, BP, temperature)
- Few jerks may occur.
- When to suspect Epileptic seizure?
    - If accompanied by eye closure or opening.
    - Eye deviation, mouth movement.
    - High BP or tachycardia.

Benign neonatal sleep myoclonus:

- Repetitive myoclonic jerks that occur during non-REM sleep in the first few weeks of life.
- Jerks are typically bilateral, symmetric movements of the arms and/or legs.
- Stops when the baby is aroused.
- Resolve spontaneously by 2-3 months.
- Absence of autonomic disturbances, myoclonic jerks occurring only while asleep.
- Normal development.
- Normal neurological exam.

Breath-holding spells:

- Occur in children 6 months to 6 years of age. will resolve spontaneously
- Pathogenesis is not clear.
- Iron deficiency is more prevalent in children with breath-holding spells.

Cyanotic breath-holding spells	Pallid breath-holding spells
• Child becomes angry or upset. • Brief period of crying, followed by breath-holding in forced expiration with apnea and cyanosis,followed by limpness and LOC. • Few children have generalized motor seizures with prolonged postictal unconsciousness.	• Less common than the cyanotic. • Child typically loses consciousness after a minor trauma to the head or upper body. • Child then stops breathing and becomes pale, diaphoretic and limp.

Gratification disorder:

- known as “benign idiopathic infantile dyskinesia”.
- 3 months to 3 years.
- Exact mechanism is poorly understood.
- Associated with self-tension, boredom, excitement, genital infection, and lack of stimulation.
- Stereotyped episodes of variable duration.
- Pressure on the perineum with characteristic posturing of the lower extremities video
- Vocalizations with quiet grunting.
- Facial flushing with diaphoresis.
- Cessation with distraction.

Tics:

- Brief, sudden, rapid, and intermittent movements (motor tics) or sounds (vocal tics).
- They may be repetitive and stereotypic.
- Tics are usually abrupt in onset and brief (clonic tics) but may be slow and sustained (dystonic tics).
- Usually associated with urge.

GERD (Sandifer syndrome):

- Intermittent paroxysmal opisthotonic posturing that are caused by GERD in infants video
- Reflects a pain response to acidic reflux.
- Could be associated with apnea, staring and minimal jerking of the extremities Associated with feedings. (30 min following a meal)

Stereotypies:

- Stereotypies are patterned, repetitive, purposeless, involuntary movements.
- Head nodding, hand flapping or clapping, finger wiggling, and facial grimacing.
- Occur with certain stimuli, such as excitement, participation in a favorite activity or boredom.
- Commonly associated with autism but it can happen in normal children.

Psychogenic nonepileptic seizures:

- Dramatic behavioral events in a conscious individual.
- Comorbid epilepsy is common.
- Present as a prolonged episode with generalized,atypical-appearing motor activity and a prompt return of consciousness.
- During the episode, patients often close their eyes tightly and resist their opening.

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