Seizure disorders In children

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Objectives

- Introduction
- What Questions should ask ?
- Classification & Etiology
- Important investigations
- Tips in treatments
- Common Epilepsies in Children
- Seizure mimikers

Introduction

- 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.

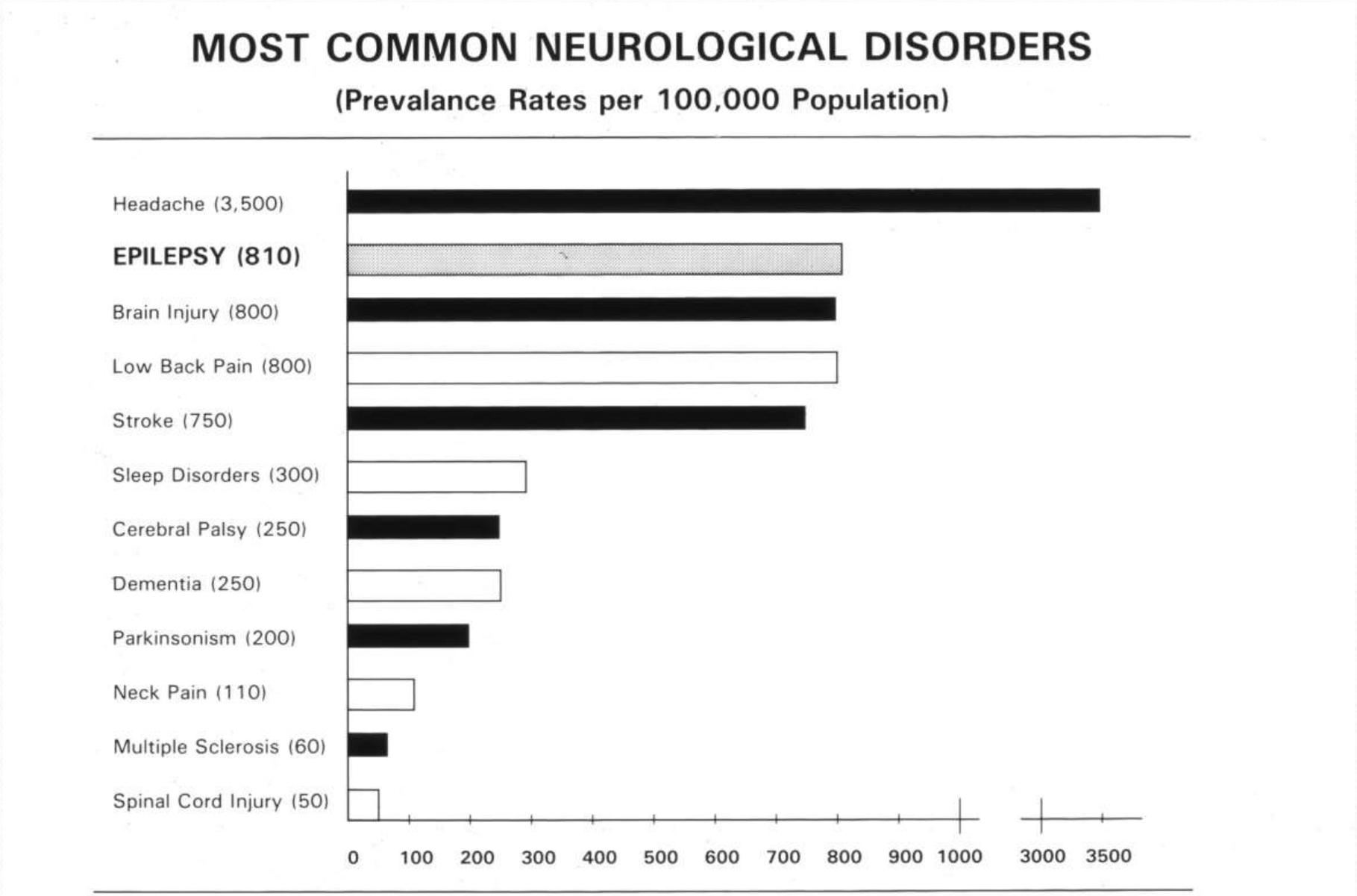
Introduction

- Epileptic Syndrome: a complex of symptoms and signs that define a unique epilepsy condition. (e.g L.G.S)
- Epileptic Disorder: A chronic neurological condition characterized by recurrent epileptic seizures

Introduction

- 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.

Introduction



Introduction

- 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%.
- Every year, an estimated 25,000 to 40,000 US children experience their first nonfebrile seizure
- The prevalence of epilepsy in Saudi children is 8.8 per 10 000

Al Salloum et al ,J Child Neurol. 2011 Jan;26(1):21-4.

Epilepsy is a clinical diagnosis

What Questions To ask ???

What Questions To ask ???

- Get a detailed account of the event or spell from a witness
- Was the spell in question a seizure?
- What type of seizure?
- Cause of seizure?

What Questions To ask ???

- Start with asking the witness to describe the spell from beginning to end

What Questions To ask ???

- Questions regarding the possible seizure can be divided into :

✓ pre-ictal

✓ Ictal

✓ post-ictal

Pre-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?

Ictal

- Was the child responding during the spell or was consciousness impaired?
- Did the child remember anything that occurred during the spell?
- Were there any repetitive behaviors during the episode, such as lip smacking, pulling at clothing, and constant rubbing of objects.

Ictal

- Did any body movements occur?
- Was there any perioral cyanosis?
- Did the patient lose continence during the spell?
- How long did the spell last?
- How many episodes?
- How often do the spells occur?

post-ictal

- 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:

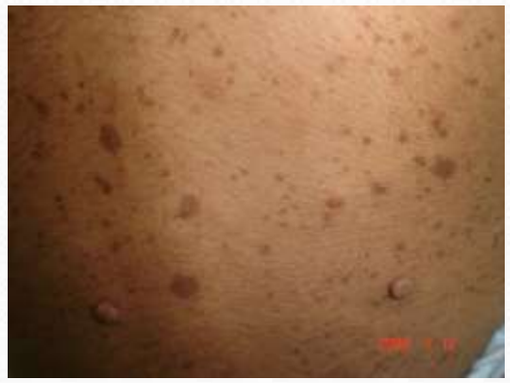
- Has the child ever had any seizures before?
- Is there any history of febrile seizures?
- Ask about past medical history
- Pregnancy and birth history
- Developmental history
- Current medications
- Is there any family history of seizures?

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

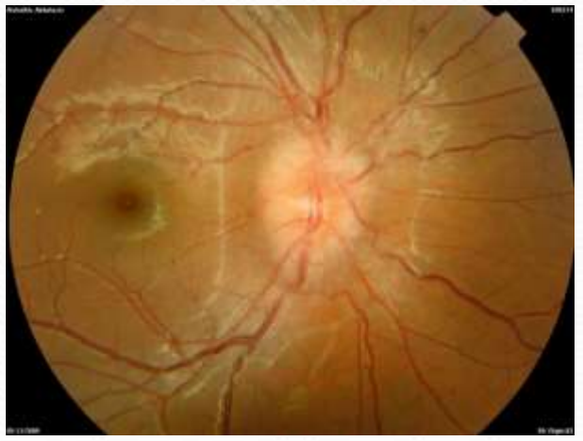
Physical Examination

- Signs of trauma.
- Signs of increased intracranial pressure
- Skin lesions may suggest a neurocutaneous diseases



Physical Examination

- 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

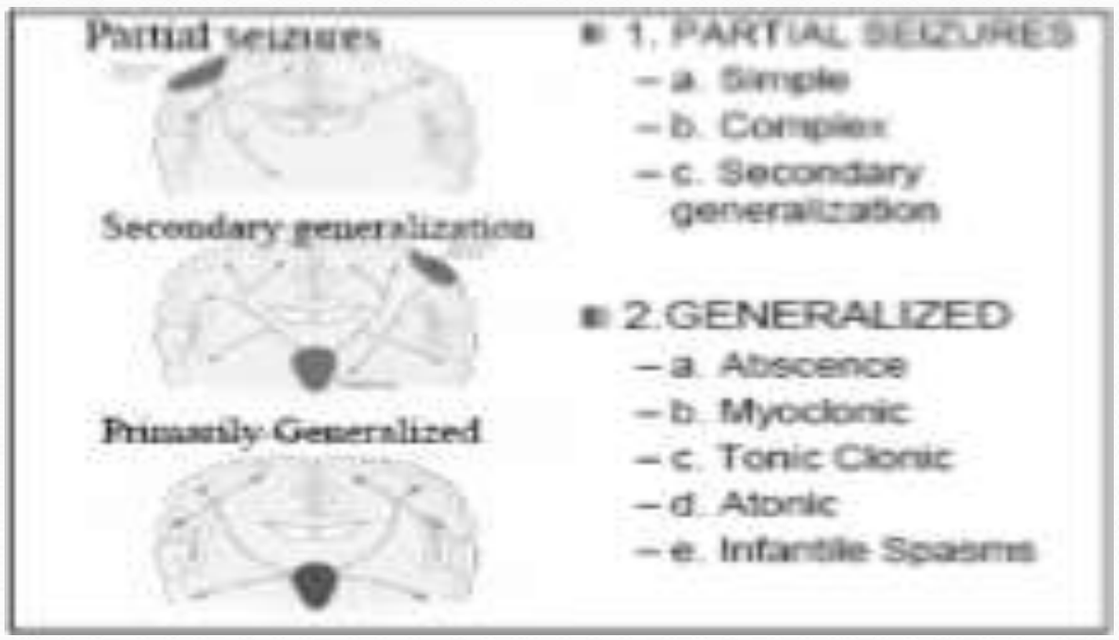
Seizure Categorization

- 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

- I. Focal Seizures
- A. Simple partial seizures (motor, sensory, hallucinations, autonomic psychic symptoms)
- B. Complex Partial Seizures (impaired consciousness)
- C. Partial seizures with secondary generalized.
- II. Generalized seizures
- A. Absence seizures (typical and atypical)
- B. Myoclonic seizures
- C. Clonic Seizures
- D. Tonic seizures
- E. Tonic-clonic seizures
- F. Atonic seizures
- III. Unclassified seizures



International classification of epilepsies ,epileptic syndrome and related seizure disorders

- Localization related (focal ,local, partial)
- Generalized epilepsies and syndromes
- Undetermined epilepsies
- Special syndromes

Undetermined epilepsies

- With both generalized and focal seizure

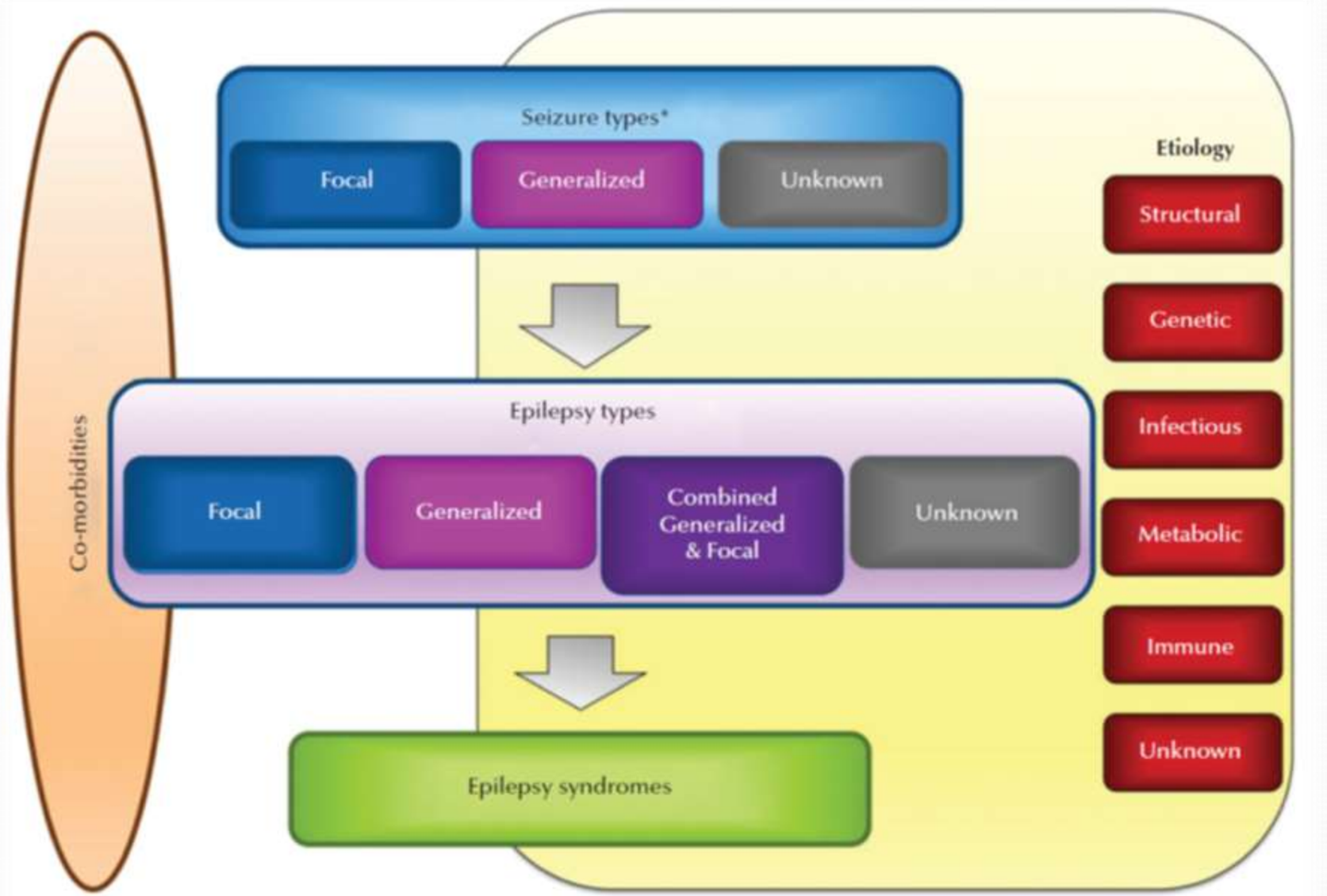
  - Neonatal seizure
  - Sever Myoclonic Epilepsy of Infancy (SMEI)
  - Epilepsy with continuous Slow – wave during sleep
  - Landau – Kleffner Syndrome (LKS)

- Without unequivocal generalized/focal features

Special Syndromes

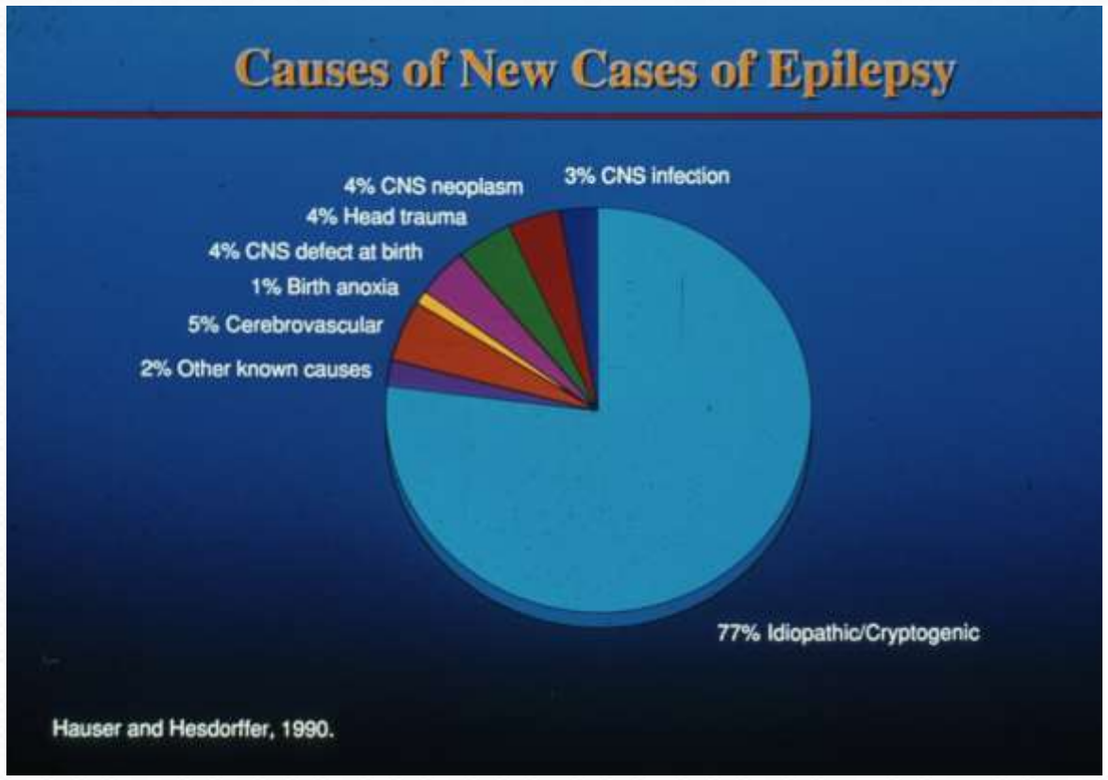
- Conditions with epileptic seizure that do not require a diagnosis of epilepsy
- Situation related seizure
  - febrile convulsion
  - isolated seizure or status epilepticus
  - seizure occurring only with acute /toxic event

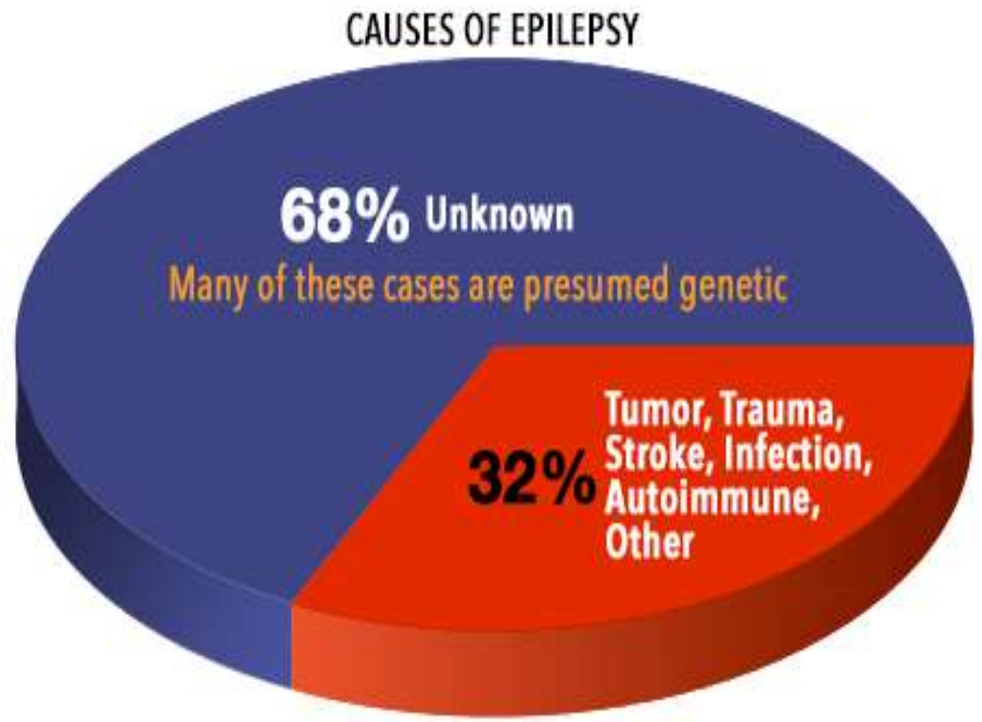
Epilepsy Classification framework:



(*) Denotes onset of seizure

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

- <u>Blood tests:</u> 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

Investigations

- <u>Lumbar puncture :</u> AAP Recommendation

✓ If the child has fever :

- Less than 12 months Strongly considered
- Less than 18 months considered

✓ Any child with meningeal signs

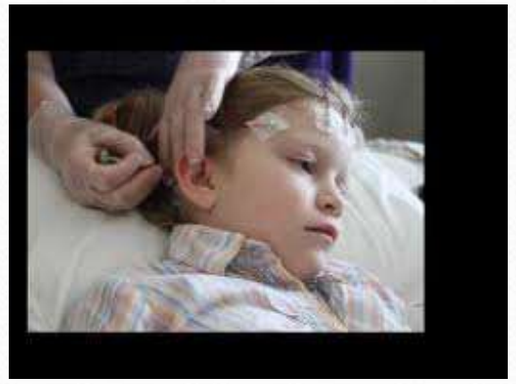
Investigations

- <u>Neuro - Imaging:</u>

✓ 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

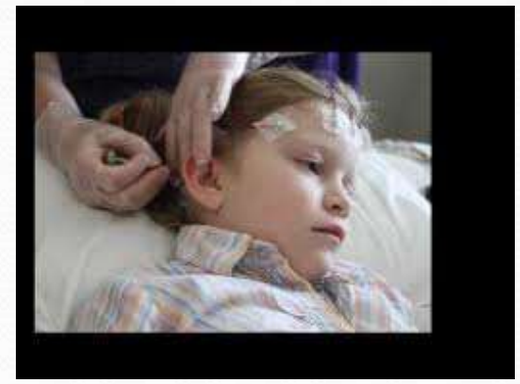
Investigations



- The EEG is recommended as part of the neurodiagnostic evaluation of the child with an apparent first unprovoked seizure.

NEUROLOGY 2000;55:616–623

Investigations



- However ,
- An EEG abnormality by itself is not sufficient to make a diagnosis of epileptic seizure nor its absence rule out a seizure

NEUROLOGY 2000;55:616–623

Epilepsy is a clinical diagnosis

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 1 Drug options by seizure type

Seizure type	First-line drugs	Second-line drugs	Other drugs that may be considered	Drugs to be avoided (may worsen seizures)
Generalised tonic–clonic	Carbamazepine* Lamotrigine[b] Sodium valproate Topiramate*[,b]	Clobazam Levetiracetam Oxcarbazepine*	Acetazolamide Clonazepam Phenobarbital* Phenytoin* Primidone*[,c]	Tiagabine Vigabatrin
Absence	Ethosuximide Lamotrigine[b] Sodium valproate	Clobazam Clonazepam Topiramate*		Carbamazepine* Gabapentin Oxcarbazepine* Tiagabine Vigabatrin
Myoclonic	Sodium valproate (Topiramate*[,d])	Clobazam Clonazepam Lamotrigine Levetiracetam Piracetam Topiramate*		Carbamazepine* Gabapentin Oxcarbazepine* Tiagabine Vigabatrin
Tonic	Lamotrigine[b] Sodium valproate	Clobazam Clonazepam Levetiracetam Topiramate*	Acetazolamide Phenobarbital* Phenytoin* Primidone*[,c]	Carbamazepine* Oxcarbazepine*
Atonic	Lamotrigine[b] Sodium valproate	Clobazam Clonazepam Levetiracetam Topiramate*	Acetazolamide Phenobarbital* Primidone*[,c]	Carbamazepine* Oxcarbazepine* Phenytoin*
Focal with/without secondary generalisation	Carbamazepine* Lamotrigine[b] Oxcarbazepine*[,b] Sodium valproate Topiramate*[,b]	Clobazam Gabapentin Levetiracetam Phenytoin* Tiagabine	Acetazolamide Clonazepam Phenobarbital* Primidone*[,c]	

Management

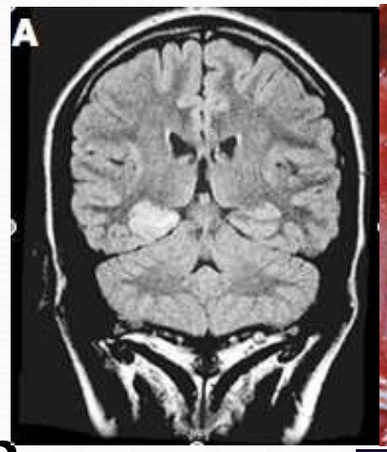
- Anti-epileptic drugs may <u>worsen</u> specific seizures

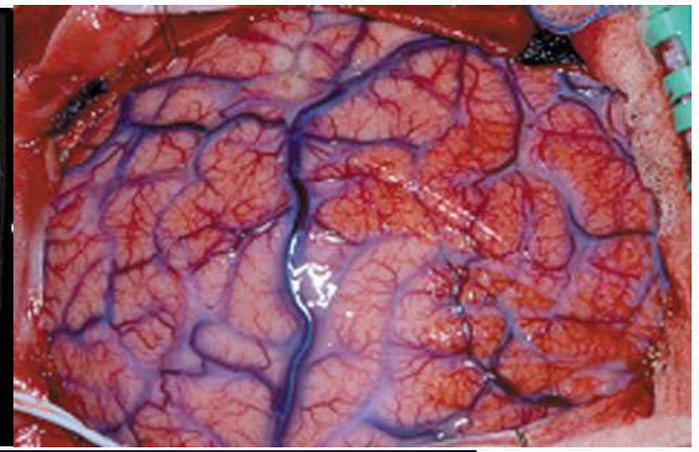
Antiepileptic drug	Epileptic syndrome/seizure type
carbamazepine, vigabatrin, tiagabine, phenytoin	childhood absence epilepsy, juvenile absence epilepsy, juvenile myoclonic epilepsy[134]
vigabatrin	absences and absence status[134]
clonazepam	generalised tonic-status in Lennox-Gastaut Syndrome[135]
lamotrigine	Dravet's syndrome[132] juvenile myoclonic epilepsy[136,137]

Epilepsy NICE guideline October 2004

Is there any other modality of treatments?

- Epilepsy surgery





- Vagal Nerve stimulation

- Ketogenic diet

When do you discontinue AEDs ?

Practice parameter by AAN (1996)

- Seizure-free 2-5 years on AEDs
- Single type of seizures
- Normal neurologic exam and normal IQ
- EEG normalized with treatment

Discontinuing AEDs .....Do Not Rush!

- Abrupt cessation of AEDs increase the risk of relapse and status epilepticus
- Withdrawal period: several weeks to several months; take in account the duration of AED therapy, specific AED in question, serum levels, degree of abnormal EEG findings
- The chance for regaining control after seizures relapse is still high


MAID OF THE MIST
Since 1846, millions of visitors have
voyaged on Maid of the Mist I through
VII to feel the roar of the Falls.

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 <u>prolonged focal seizure</u> 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
- A shorter duration of temperature before the first seizure
- A lower temperature at the time of the first seizure

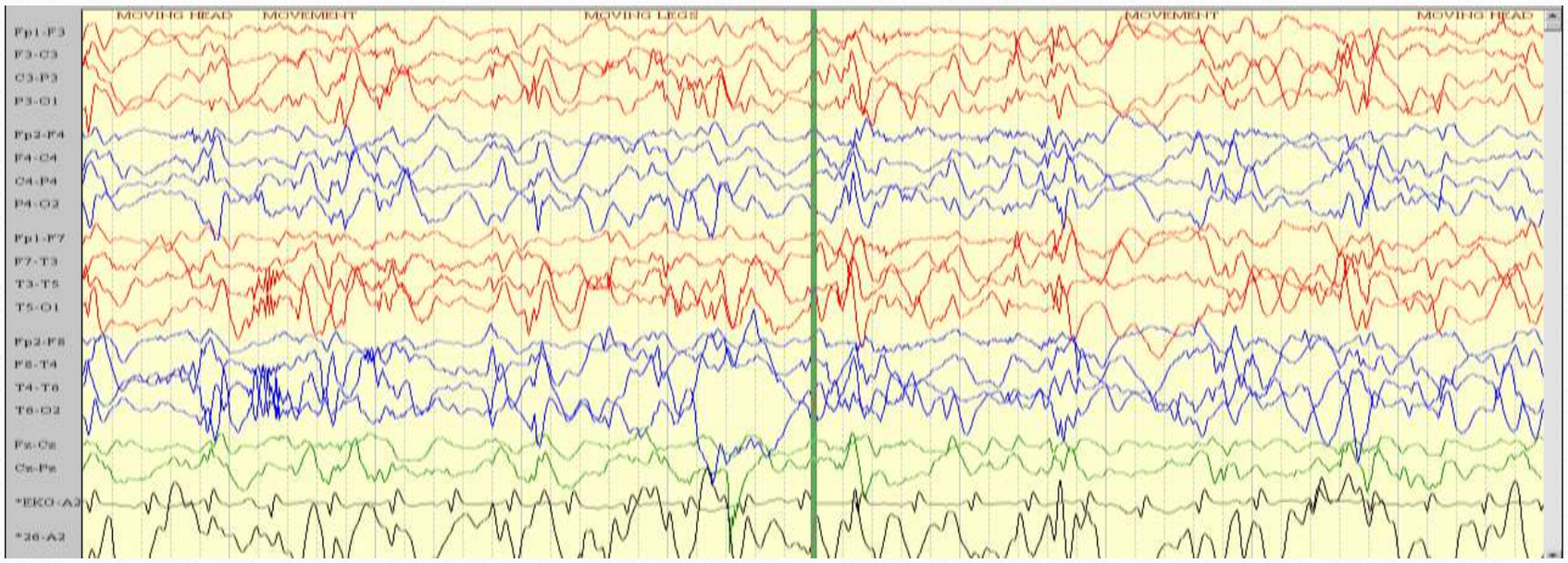
Treatment of Febrile Seizures

- Usually brief and self-limited
- Assurance and education
- Antipyretic agentsContinuous AED treatment: phenobarb, VPA; effective for prolonged repetitive seizures; not recommended due to side effect profile
- 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

Common Epilepsies in Children

Infantile Spasms (I.S)

- Age: < 1 year with peak at 4-7 months
- Spasms: flexor , extensor or mixed
- Developmental arrest or regression
- EEG: hypsarrhythmia



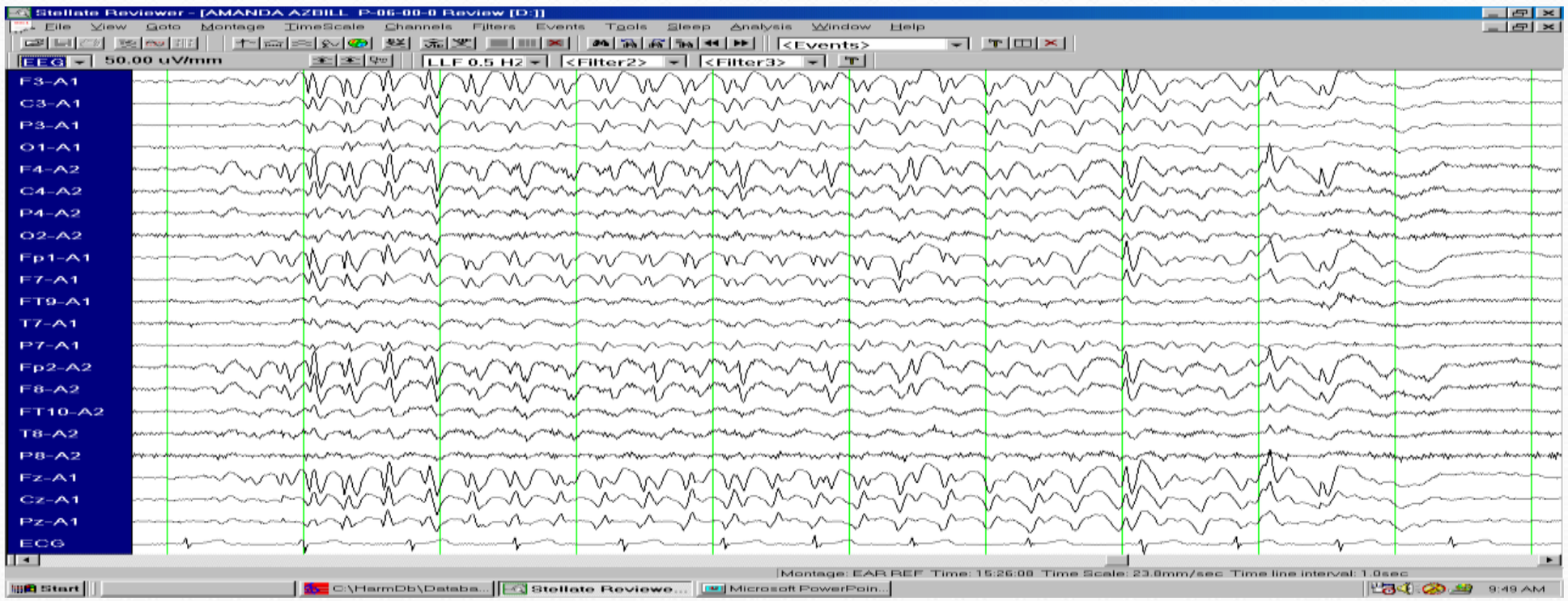
Infantile Spasms (I.S)

- Identified causes in 75% of the cases. TS in 20%. Brain malformation and HIE are the most common causes
- Idiopathic I.S has the best prognosis
- Treatment: ACTH, Prednisone in idiopathic I.S

VGB, Cpz, VPA, pyridoxine

Childhood Absence Epilepsy

- Age: 5-10 years
- Girls more than boys
- 30% of pt develop GTCs
- EEG : 3 Hz generalized spike and wave

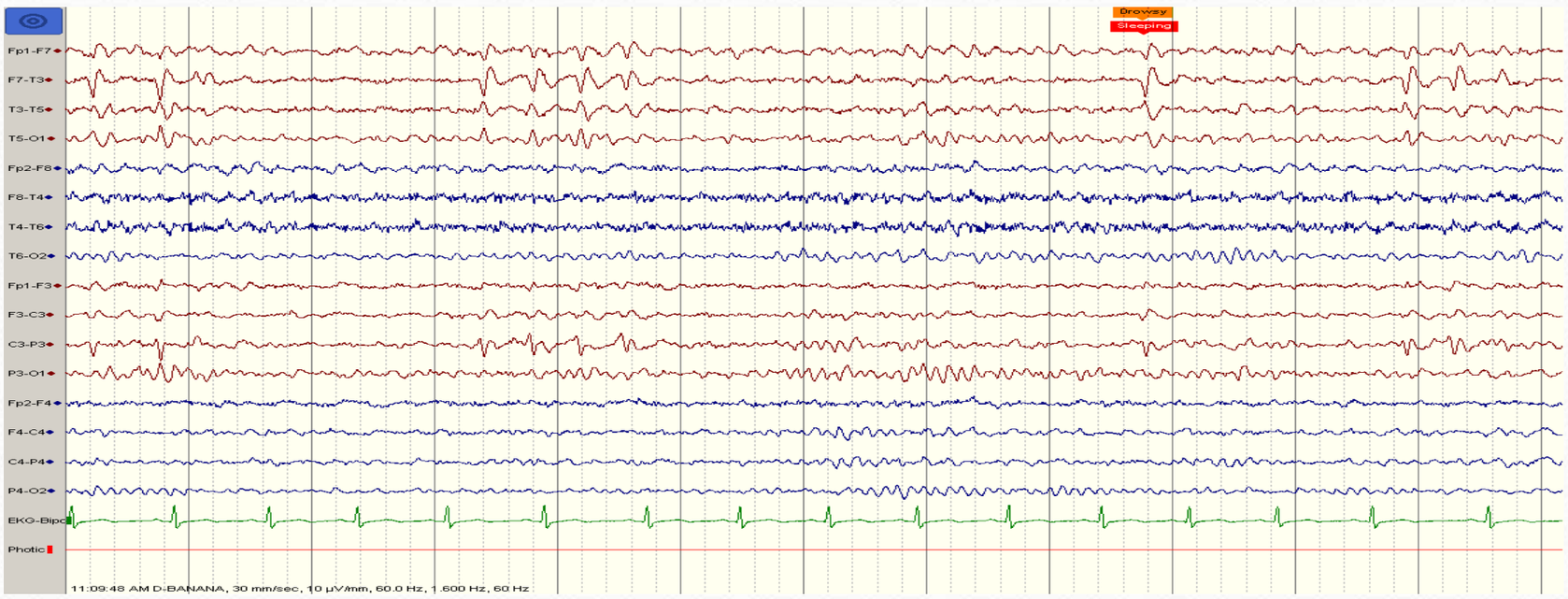


Childhood Absence Epilepsy

- Remission: 80% after age of 10 years
- Treatment: First choice: ETX, VPA
  Second choice: LTG,TPM

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
- EEG:



- Treatment:

Seizure mimikers

Paroxysmal Non-Epileptic Events

PNE With Alterations in Consciousness

- Breath-holding Spells.
- Syncope.
- Hyperventilation.
- Sleep-related phenomena.
- Apnea.
- Somatoform disorders.
- Munchausen syndrome by proxy.

PNEC Without Alterations in Consciousness

- 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.

Jitteriness

- Can be lessened by removing the stimulus or relaxing the affected limb.
- Causes:
  - Mild jitteriness are common in healthy newborn.
  - Drug withdrawal.
  - Hypocalcemia.
  - Hypoglycemia.
  - HIE.

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.

Apnea

- **<u>When to suspect Epileptic seizure ?</u>**
  - 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.

Benign neonatal sleep myoclonus

- 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.
- Pathogenesis is not clear.
- Iron deficiency is more prevalent in children with breath-holding spells.

Cyanotic 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.

Pallid breath-holding spells

- 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.

Gratification disorder

- Stereotyped episodes of variable duration.
- Pressure on the perineum with characteristic posturing of the lower extremities.
- 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.
- 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.

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.

- Childhood epilepsies: What should a pediatrician know? Bashiri FA. Neurosciences (Riyadh). 2017 Jan;22(1):14-19. doi: 10.17712/nsj.2017.1.20160244.

Q &A

Thank You

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