



Seizure Disorders

objectives:

- To be able to differentiate seizure from epilepsy
- To know the Classification & Etiology of epilepsy in children
- To know common Epilepsy syndromes in Children.
- To recognize Seizure mimickers.
- Approach to history taking of a seizure.
- To understand basic investigations related to seizure

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Special thanks to team 437 & Faisal alsaif

Notes

Important

Book

Seizure VS Epilespy

Definitions:

Seizure:

➤ It is a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain.

Epilepsy:

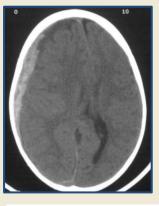
- ➤ It is a disorder of the brain characterized by an enduring predisposition to generate epileptic seizures.
- As a consequence, there are neurobiological, cognitive, psychological and social comorbidities.
- Seizures are an acute event and can be considered a symptom. Seizures don't necessarily mean epilepsy. Only 30-50% of patients with first unset unprovoked seizure progress to develop epilepsy

Practical definition of epilepsy

Epilepsy can be defined by any of the following:

- 1. At least two <u>unprovoked</u> (or reflex) seizure occurring > 24hrs apart.
- 2. One unprovoked (or reflex) seizure with a probability of further seizures similar to the general recurrence risk (at least 60%), occurring over the next 10 years.
- 3. A diagnosis of an epilepsy syndrome.

Examples of Provoked seizures:



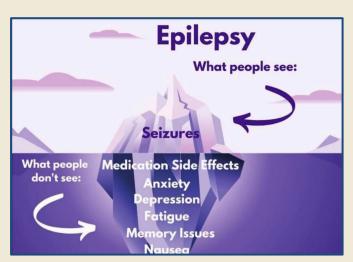
Subdural Hematoma



Epidural Hematoma

Epilepsy is "resolved" if one of the following is applicable:

- Age-dependant epilepsy syndrome and the individual past the applicable age.
- Seizure-free for the last 10 years a nd off anti-seizure medications for five years.



Seizure Classification

Epidemiology:

- Estimated incidence in the US and Europe is 48 per 100,000 per year.
- The prevalence in developed (high-income) countries is 5.8 per 1,000 population, whereas in resource-poor (lower-income) countries it was 10.3 per 1,000.
- The prevalence of epilepsy in Saudi children is 8.8 per 10,000.

Causes of New Cases of Epilepsy 4% CNS neoplasm 4% CNS defect at birth 11% Birth arrovla 5% Cerebrovacular 2% Other known causes 77% Idiopathic/Cryptogenic Hauser and Hesdorffer, 1990.

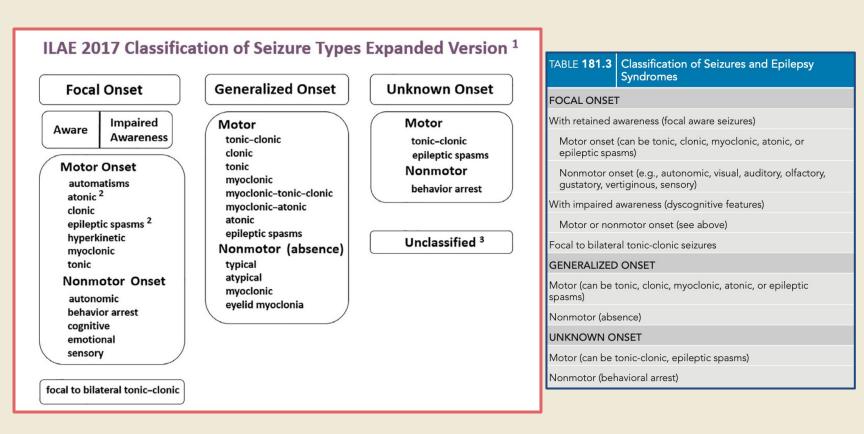
ILAE Classifications for Seizure and Epilepsy

Terms no longer in use:

- Complex partial
- Simple Partial
- Partial
- Psychic

- Dyscognitive
- Secondarily generalized tonic clonic
- Grand ma
- Petit mal

Classification:

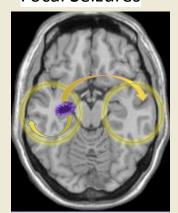


- 1. First determine if the symptomes are focal and can be localised or are generalized.
- 2 If focal:
 - a. Is the awareness intact or impaired?
 - b. Is there motor involvement?
- 3. If generalized:
 - a. Generalized seizures always impair the awareness
 - b. Generalized non motor seizures present with the complaint of loss of awareness

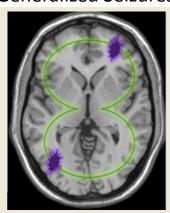
Seizure Classification

ILAE Classifications for Seizure and Epilepsy

Focal Seizures

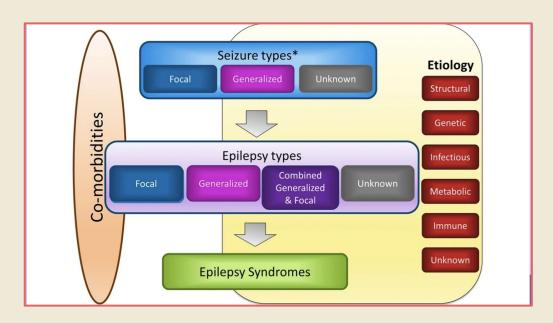


Generalized Seizures



Focal seizures usually have a closed propagation circuit with the potential to cross over

Generalized seizures can have multiple originations with a propagation circuit that involves both hemispheres



Examples: try to classify the seizure!

An 8-year old male has a seizures during which he remains fully aware. He has clonic movement of his left arm for 1 minute.

Focal Aware motor (clonic) seizure

❖ A 2-year old boy developed a seizure characterized by generalized stiffness, followed by a clonic movement of all 4 extremities, during which he was unresponsive.

Generalized motor (Tonic-clonic)

Epilepsy Syndromes:

- It is a cluster of features incorporating seizure types, EEG, and imaging features that tend to occur together.
- It often has age-dependant features such as age at onset and remission (where applicable)

Approach to a seizure patient

DIfferential:

- Before approaching the patient as a seizure patient. Make sure its a seizure!
- Mimickers include:
 - > Syncope
 - Breath holding spell
 - ➤ GERD
 - Panic attack
 - Daydreaming

- Conversion or Non Epileptic Seizure
- Benign sleep myoclonus
- Benign paroxysmal vertigo
- Complicated migraine
- Motor tics

History taking:

- Always try to ask a witness
- Questions are divided into:
 - Pre-Ictal (before):
 - Was there any warning before the spell? If so, what was the warning?
 (Abdominal discomfort, fear or any other unpleasant sensations)
 - What was the child doing before?
 - Was the child just fed?
 - Was the child asleep or awake?
 - Was the child sleep deprived?
 - Were there any triggers?
 - Did the child have fever or illness?
 - > Ictal (during):
 - Was the child responsive or unresponsive?
 - Did the child remember anything that occurred during the spell?
 - Were there any repetitive behaviors during the episode, such as lip smacking, or pulling at clothing?
 - Describe the movement and the distribution.
 - Was there any perioral cyanosis?
 - Did the patient lose bowel or bladder control?
 - How long did it last?
 - How many episodes?
 - How often does it happen?
 - Post-Ictal (after):
 - What happen immediately after?
 - Confused, tired, or asleep
 - 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?

Approach to a seizure patient

Risk Factors to ask about:

- 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 examination should be done
- Pay attention to:
 - 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 (Especially head)
- Signs of increased intracranial pressure. LOC, Pupils, posture, check fontanelles if applicable
- Skin lesions suggesting neurocutaneous diseases



Hypomelanotic macules
Seen in Tuberous sclerosis



Cutaneous neurofibroma
Seen in Neurofibromatosis 1

- Focused neurological exam to look for focal deficits
- Fundoscopy



Cherry red spot Seen in Sphingolipidoses

Investigations

Initial blood work:

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

Lumbar puncture

- ❖ AAP Recommendation:
- If the child has fever::
 - Less than 12 months. LP is strongly considered
 - Less than 18 months can be considered.
 - > Any child with meningeal signs

Imaging

- CT head: If head trauma is suspected
- MRI: If the child has focal seizures or a focal neurological deficit, or if there are signs of high ICP
- **EEG:** recommended as a part of the neurodiagnostic evaluation of the child with an apparent first unprovoked seizure.
 - An EEG abnormality by itself is not sufficient to make a diagnosis of epileptic seizure nor its absences rule out a seizure
 - Epilepsy is a clinical diagnosis

Case-based investigations:

- 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
 - Serum Amino acids
 - Urine organic acids

Febrile Seizures:

- It is a seizure occurring in association with a febrile illness:
 - In the absence of a central nervous system infection or acute electrolyte imbalance
 - o In children older than 1 month, without prior afebrile seizures.
- Prevalence: 2-5%
- Age: 3 month –5 years, peak at 18 months.
- Body temperature should be > 38.4°

Classification of Febrile seizures

	Simple	Complex	Febrile Status Epilepticus
Focality	Generalized tonic- clonic	Focal	Seizure lasting > 30 minutes
Duration	<15 minutes	>15 minutes	Brief serial seizures
Frequency	One	Multiple during same febrile illness	without regain of consciousness in between

Risk of development of Epilepsy

Recurrent FS	Epilepsy	
Family history of febrile seizures	Neurodevelopmental abnormality	
Age less than 18 months	Complex febrile seizures	
Height of peak temperature	Family history of epilepsy	
Duration of fever (<1hr before onset of seizure)	Duration of fever	

Risk for epilepsy after simple FS is 1-2%, similar to general population

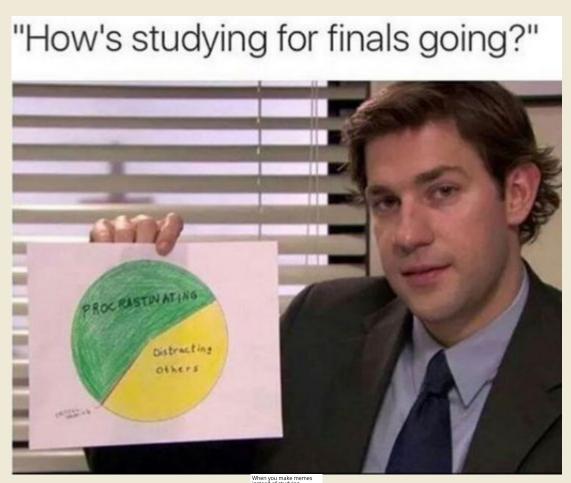
Focal, unilateral, status seizures all increase the risk of developing epilepsy

Investigations for Febrile Seizures:

- A serum glucose level should be assessed.
- Routine laboratory studies are not necessary because electrolyte abnormalities are rare.
- 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.

Treatment of Febrile Seizures:

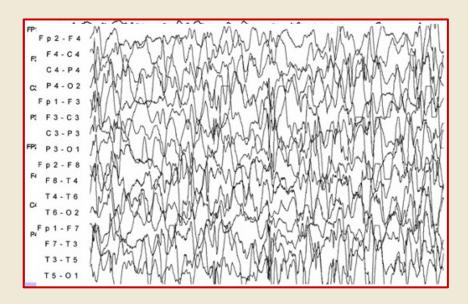
- Reassurance and education for caregivers
- Antipyretic agents (Control the fever)
- Long-term anti-seizure medications are not required
- Abortive treatment with **rectal Diazepam**; It's the preferred modality for prolonged repetitive seizures.





Infantile Spasms:

- It is an age-dependant epilepsy syndrome.
- Age: < 1 year with peak at 4-7 months
- Spasms: flexor, extensor or mixed
- Occurs in clusters, usually before or after sleeping.
- EEG: hypsarrhythmia (MCQ)
- West Syndrome is a triad of:
 - 1. Infantile spasms
 - 2. Developmental arrest or regression
 - 3. EEG: hypsarrhythmia





This is Hypsarrhythmia on EEG
It can come in the exam

Infantile spasm (IS):

- Identified causes in 75% of the cases.
- Tuberous Sclerosis accounts for a 20% of IS.
- Brain malformation and HIE are the most common causes
- Idiopathic IS has the best prognosis
- Treatment for IS: Stepwise approach.
 - ACTH, or Prednisone.
 - Vigabatrin For infants with underlying tuberous sclerosis, vigabatrin is the treatment of choice.

See this video

Childhood Absence Epilepsy:

- Age of onset: 5-10 years.
- Female are affected more than boys.
- Brief starring, subtle eye blinking, with loss of awareness.
- Seizure can be triggered by hyperventilation.
- Duration: 5-20 seconds.
- 10-50% of patients will have develop generalized tonic-clonic seizure at least once in their lives.
- Largely disappear by teen age, 90% by the age of 20.

EEG:

Generalized 3Hz spike and wave (MCQ)

Can be provoked by hyperventilation

Doctor: a trick for your exam:

If the EEG is of an infant → Infantile

Spasm

If the EEG is of a child → Absence



Absence Epilepsy Treatment:

• First line: Ethosuximide, Valproic Acid.

• Second line: Lamotrigine, Topiramate

See this video

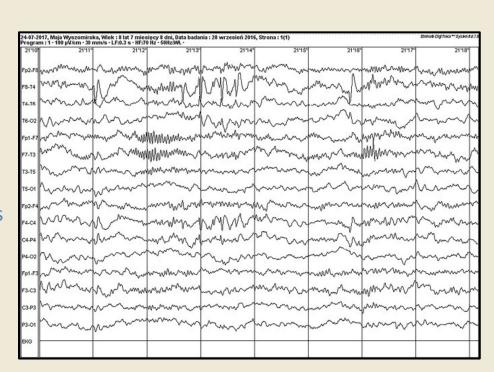
TABLE 181.4 Differentiating Absence Seizures and Focal Seizures with Impaired Awareness		
FEATURE	ABSENCE SEIZURE	FOCAL SEIZURE WITH IMPAIRED AWARENESS
Duration	Seconds	Minutes
Provoking maneuver	Hyperventilation Photic stimulation	Variable, but often none
Postictal phase	None (return immediately to baseline)	Confusion, sleepiness
Number of seizures	Many per day	Infrequent (rarely >1/day)
EEG features	Interictal: normal except bursts of generalized spike wave and sometimes occipital intermittent rhythmic delta activity Ictal: 3-Hz generalized spike-wave	Interictal: normal or focal slowing, sharp waves, or spikes Ictal: focal discharges (with or without spread to contiguous regions or the contralateral hemisphere)
Neurologic examination	Normal	Normal, or focal deficits
Neuroimaging	Normal*	Normal, or focal abnormalities (mesial temporal sclerosis, focal cortical dysplasia, neoplasm, encephalomalacia)
First-line treatment	Ethosuximide or valproic acid [†]	Often levetiracetam, oxcarbazepine, or lamotrigine are used

Self Limited Epilepsy with Centro-temproal spikes (SeLECTS):

- Previous names: **Rolandic epilepsy**, benign childhood epilepsy with centrotemporal spikes.
- Age: 5-9 years of age.
- Remission: in 2-4 years, almost 100% by the age of 16 years.
- Seizure semiology: Nocturnal, focal seizure with unilateral facial sensori-motor symptoms, oro-pharyngeal-laryngeal presentation
 - Gurgling sounds, drooling, speech arrest (wake up unable to speak with drooling, possibly progressing to a tonic clonic fit.)
- Intact awareness in 60%

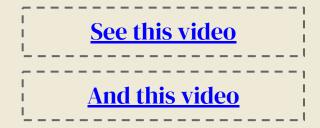
EEG:

classically demonstrates independent bilateral centrotemporal sharp waves but is otherwise normal (نفس الاسم)



Treatment:

- **Reassurance** and education about the natural course of the disease.
- Conventional anti-seizure medication might not be necessary
- If the parents are worried or It is nocturnal and severe, we can give BZDs or Anti epileptics



Seizure Mimickers

With altered Level of consciousness	Without altered Level of consciousness	
Syncope	Sandiffer (GERD)	
Apnea	Daydreaming	
Breath-Holding spells	Migraine	
Psychogenic	Movement Disorders (tics, stereotypes)	
Sleep-related disorders.	Self gratification	
-	Jitteriness	

Syncope:

- A transient episode of altered level of consciousness due to decrease cerebral blood flow.
 - Vasovagal mechanism → Bradycardia and hypotension.
- It occurs in ≤ 15% of teenagers.
- Preictal: Lightheadedness, dizziness, appears clammy and pale, fading of vision and/or hearing.
- Ictal: falling to ground.
- Post-ictal: Recovery is complete within a few minutes with no post-ictal confusion.

Breath-holding Spells:

- Common frightening event for children 6-month to 4-years.
- They are classified based on the skin color change during the event:
 - Cyanotic: Triggered by anger and frustration
 - Pallid (less common): triggered by minor trauma.
- It can provoke a reflex generalized tonic-clonic seizure. Secondary to the spell

See this video

Seizure Mimickers

Jitteriness:

- Common in the neonatal period.
- Recurrent, low amplitude tremor.
- Exaggerated response to touch and or loud noise.

Apnea:

- Unexplained cessation of breathing for at least 20 seconds
- Or shorter respiratory pause associated with bradycardia, cyanosis, pallor, and/or marked hypotonia

Benign neonatal sleep myoclonus:

- Repetitive myoclonic jerks during non-REM sleep.
- Suppressible when the baby is aroused.
- Resolved by 2-3 months.
- Otherwise normal development.

Sandifer Syndrome (GERD):

- Affects children less than 2 years of age.
- Intermittent events characterized by: back arching, stiffening of the neck or arms, and resolve within a few minutes.
- Associated with feeding (between 1—30 minutes after meals).
- Can be accompanied by apnea, starring and jerking of the extremities.
- Frequency: Multiple times per day.

Gratification Disorder:

- Also known as known as "benign idiopathic infantile dyskinesia".
- Age: 3 months to 5 years.
- Can be considered as stereotype:
 - Dystonic posturing, grunting, rocking, and sweating.
- Self-limited and can resolve with distraction.
- Its a form of masturbatory behaviour. Also known as Infantile masturbation.... yep.

Seizure Mimickers

Tics:

- Brief, sudden, rapid, and intermittent movements (motor tics) or sounds (vocal tics).
- Repetitive and non-rhythmic.
- Usually associated with urge.
- Disappears in sleep

Stereotypes:

- It is a coordinated, patterned, repetitive, rhythmic and purposeless movement.
- It could be involuntary or in response to internal stimuli.
 - Excitement or stress.
- Examples; head nodding, hand flapping or clapping, finger wiggling, and facial grimacing.

Psychogenic Nonepileptic Seizures (PNES):

- Sudden, involuntary behavioral, motor, cognitive changes that resemble epileptic seizure
- It is a dissociative psychological response to aversive stimuli
- It is not under voluntary control (NOT fake)
- Mean age at presentation is 10-14 years.
- Females more than males.
- Comorbid epilepsy is common

PNES VS ES

Table 1 Signs and symptoms distinguishing psychogenic nonepileptic seizures (PNES) and epileptic seizures (ES)			
	Signs and symptoms favoring PNES	Signs and symptoms favoring ES	
Differences between PNES and ES irrespective of types of events	Duration of longer than 10 min		
	Vocalization during or after seizures, complex, with affective content	Vocalization at the beginning, primitive, has no emotional expression	
	Ictal crying		
	Occurrence from preictal pseudosleep	Occurrence from sleep	
	Ictal eye closure		
Signs and symptoms discriminating Hypermotor and Focal Motor PNES from ES	Side-to-side head or body movement (compared with generalised tonic clonic seizures only)	Postictal confusion	
	Pelvic thrusting (frontal lobe partial seizures excluded)	Postictal stertorous (generalised tonic clonic seizures only)	
	Fluctuating course		
	Asynchronous jerks (frontal lobe partial seizures excluded)		
Signs and symptoms discriminating Akinetic and Subjective Symptoms	Memory recall		



Status Epilepticus:

- Status epilepticus is a neurologic emergency defined as
 - Ongoing seizure activity for **5 or more minutes** or
 - Repetitive seizures without recovery of consciousness
- Etiologies include:
 - New-onset epilepsy
 - drug intoxication
 - O drug withdrawal (missed anticonvulsant doses),
 - hypoglycemia
 - Electrolyte imbalance
 - Others: acute head trauma, infection, ischemic stroke, intracranial hemorrhage, metabolic disorders, and hypoxia

Treatment:

- ABCs
- Give O2 if needed and secure IV access
- O Give IV Benzos.
- If no access, give Rectal Diazepam
- No resolution after two doses? Anticonvulsants
- No resolution? Infusion of Midazolam
- No resolution? Anesthetise the patient

TABLE 181.5 Mar	nagement of Status Epilepticus
STABILIZATION	
ABCs (airway, breath	ing, circulation)
Cardiac monitoring	
Oxygen and pulse or	kimetry
Intravenous access	
Immediate laborator	y tests
Glucose	
Basic metabolic pa	anel—sodium, calcium, magnesium
Anticonvulsant dru	ıg levels
Toxicology studies	as appropriate
Complete blood c	ounts, platelets, and differential
PHARMACOLOGIC	MANAGEMENT
Benzodiazepine	
Lorazepam IV, IN	
Diazepam IV Rectal diazepam	
Midazolam IV, IM,	IN, buccal
Standard anticonvuls	ant medications
Fosphenytoin IV, IN	М
Phenobarbital IV	
Levetiracetam IV	
Valproic acid IV	
Continuous infusions	
Pentobarbital	
Midazolam	
General anesthesia	

EXTRA

Status Epilepticus:

- **JME** is the **most common generalized epilepsy** among adolescents and young adults.
 - Onset is typically in early adolescence with myoclonic jerks (exacerbated in the morning)
 - o **levetiracetam is now favored**, particularly for girls and women since treatment of JME is usually lifelong.
- Lennox-Gastaut syndrome is a severe epilepsy syndrome
 - o most children present before age 5 years.
 - Frequent, multiple seizure types including atonic, focal, atypical absence, and generalized tonic, clonic, or tonic-clonic semiologies characterize the disorder.
 - Many children have underlying brain injury, malformations, or genetic etiologies.
 - The seizures are typically difficult to control, and most patients have **significant intellectual disability**.
- Benign (self-limited) neonatal convulsions are an autosomal dominant genetic disorder linked to abnormal neuronal potassium channels.
- Otherwise well newborns present with focal seizures toward the end of the first week of life, leading to the colloquial term *fifth-day fits*.
- Response to treatment is generally excellent, and the long-term outcome is typically favorable.
- Acquired epileptic aphasia (Landau-Kleffner syndrome)
 - is characterized by the **abrupt loss of previously acquired language** in young children.
 - The language disability is an **acquired cortical auditory deficit (auditory agnosia).**
 - The **EEG** is highly epileptiform in sleep, the peak area of abnormality often being in the dominant perisylvian region (language areas).

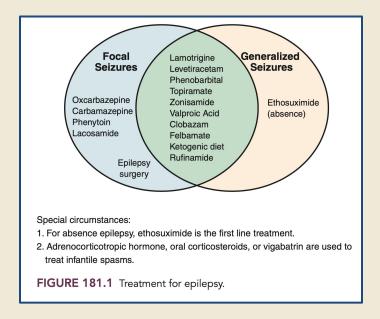


Atypical absence, Myoclonus, Atonic seizures

- **Atypical absence:** episodes of impaired consciousness with automatisms, autonomic phenomena, and motor manifestations, such as eye opening, eye deviation, and body stiffening. They are associated with **slower EEG discharges (2 Hz)**
- Myoclonus is a sudden jerk of all or part of the body; not all myoclonus is due to epilepsy.
 - Non Epileptic myoclonus may be benign, as in sleep myoclonus
 - Myoclonic epilepsy usually is associated with multiple seizure types. The
 underlying disorder producing myoclonic epilepsy may be static (e.g., juvenile
 myoclonic epilepsy [JME]) or progressive and associated with neurologic
 deterioration (e.g., neuronal ceroid lipofuscinosis).
 - Myoclonic absence refers to body jerks that commonly accompany typical or atypical absence seizures.
- Atonic seizures: involve a sudden loss of postural tone that often results in falls and injuries; though typically brief (lasting 1–2 seconds), they are very disabling.

Long-term Epilepsy therapy:

- based on the likelihood of recurrence balanced against the risk of long-term drug therapy.
- Absence seizures, infantile spasms, atypical absence seizures, and atonic seizures are universally recurrent at the time of diagnosis, so treatment should be initiated immediately.
- For most children, medications can be weaned off after 2 years without seizures.



Questions

1- Angelo, a 15-month-old boy, had been unwell with a runny nose and cough for a day when his father brings him to the Emergency Department.

At lunch he suddenly became stiff, his eyes rolled upwards and both his arms and legs started jerking for 2 minutes. He felt very hot at the time. When examined 2 hours later, he has recovered fully. This is the first time this has happened. He has a normal neurological examination and is acquiring his developmental milestones normally. He has no other medical problems. The triage nurse performed a blood glucose test, which indicated a glucose level of 4.2 mmol/L (within normal range).

What would be the most appropriate investigation?

- A. CT scan of the brain
- B. ECG
- C. EEG (electroencephalography)
- D. No investigation required
- E. Oral glucose tolerance test
- 2- Pamela is an 8-year-old girl with recurrent seizures. She has three or four seizures a month, where she lets out a cry, her arms and legs become stiff, her eyes roll upwards and then she jerks her arms and legs. This lasts about 3 minutes. Afterwards she sleeps for 2 hours and is then back to normal. She is doing well at school but is sometimes missing school because of her seizures. She is currently not on any medication and has no other medical problems.

What would be the best intervention for this child?

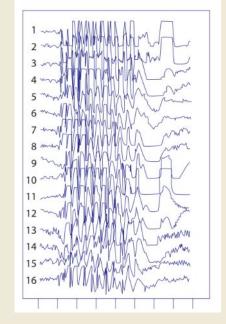
- A. Anti-epileptic drug therapy
- **B.** Home schooling
- C. Ketogenic diet
- D. No intervention required
- E. Vagal nerve stimulation
- 3- Antonia, a 5-year-old girl, is seen by her general practitioner. Her mother and schoolteacher have noticed she has episodes where she stops her activity for a few seconds, stares blankly ahead and then resumes the activity as if she had never stopped.

These episodes happen many times a day.

She has no other medical problems and there is no family history of note.

The EEG during an episode is shown

- A. Childhood Rolandic epilepsy (benign epilepsy with centro-temporal spikes)
- B. Childhood absence epilepsy
- C. Juvenile myoclonic epilepsy
- D. Lennox-Gastaut syndrome
- E. Infantile spasms (West syndrome)



4- Vijay is a 5-month-old infant who has been seen repeatedly by his general practitioner because of colic. His mother brings him to the Accident and Emergency department as he is having episodes of suddenly throwing his head and arms forward. These episodes occur in repetitive

bursts. His mother thinks they may be something more than

just colic, as he is now not smiling or supporting

his head as well as he did previously.

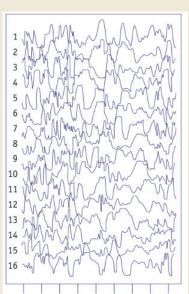
He was born at term by normal vaginal delivery and has no other medical problems.

His EEG is shown. Choose the most likely diagnosis.

- A. Juvenile myoclonic epilepsy
- **B.** Lennox-Gastaut syndrome
- C. Childhood Rolandic epilepsy (benign

epilepsy with centro-temporal spikes)

- D. Childhood absence epilepsy
- E. Infantile spasms (West syndrome)



Answers

1- D. No investigation required

Angelo had a simple febrile seizure, secondary to a respiratory tract infection. A febrile seizure is a clinical diagnosis and does not require investigation. Indeed, there is no confirmatory test.

2- A. Anti-epileptic drug therapy

This child should be started on an anti-epileptic drug. Pamela has recurrent generalised tonic-clonic seizures which are affecting her quality of life as she is missing school.

3- **B.** Childhood absence epilepsy

The history suggests this diagnosis. In addition, the EEG corresponds with it, showing 3/s spike and wave discharge, which is bilaterally synchronous during, and sometimes between attacks.

4- E. Infantile spasms (West syndrome)

Vijay has violent flexor spasms of the head, trunk and limbs followed by extension. These are known as infantile spasms. He also has developmental regression. The EEG shown is characteristic, called hypsarrhythmia.