Paediatric Seizures & Epileptic Syndromes

Amal Yousef Kentab
Associate Professor and Consultant Pediatric Neurologist
MRCP (UK), ABP, KSFP,KFHPN, MAAN
Pediatric Department, Neurology Division
College of Medicine & King Khalid University Hospital
King Saud University

Notes by: Jawaher Abanumy

Objectives

- Seizure is a symptom not a disease
- Clinical observation crucial for Dx , classification & treatment
- R/O other paroxysmal , non-epileptic disorders
- Acute management & prevention of recurrence of seizures.
- Evaluation & patient work-up
- Optimum use of anti-epileptic drugs (AED)
- Comprehensive patient management

This lecture is from your book “Illustrated”.

I want you to focus on **febrile seizures**, **infantile spasm, absence seizure.**

Childhood Epilepsies

When we talk about epilepsy it is like we are talking about seizure disorders.

- Synonyms for epilepsies
  - Seizure Disorders
    (Fits)
  - Paroxysmal Disorders*
    (Funny turns)
  - Convulsive Disorders

*Sometimes pts might have changes in the color and altered level of consciousness with abnormal movements.
Under this you can have epilepsy and nonepileptic disorders.

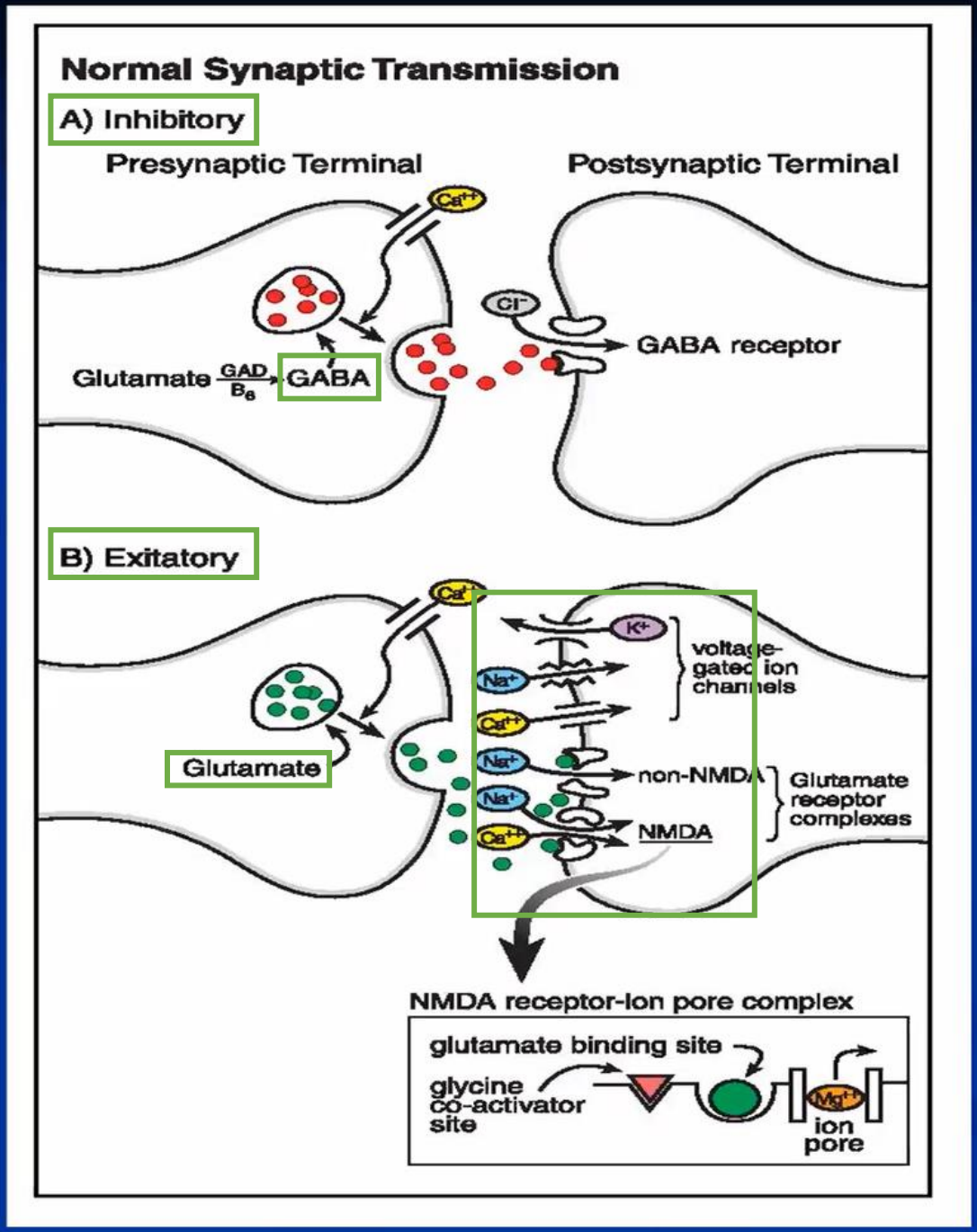
Definition

- **A Seizure :**

  is a clinical event in which there is a sudden disturbance of neurological function caused by an abnormal or excessive neuronal discharge.

This picture shows the synaptic transmission with the inhibitory (A) pathway and the excitatory (B) pathway.

The important thing in the inhibitory is **GABA** (inhibitory amino acid) while in the excitatory it is **glutamate** (excitatory amino acid)



The glutamate will go to the synaptic cleft and focus on multiple channels including sodium, calcium, non-NMDA and NMDA. It will act on them and produce neuronal excitation.

GABA is responsible for inhibition so once there is imbalance between the 2, the patient will have neuronal excitation.

Definition

- **Epilepsy :**

  Is a situation of chronic recurrent seizures (two or more unprovoked)other than febrile convulsions in the absence of an acute cerebral insult .

- **Convulsions :**

  Refer to seizures with motor manifestations.

Definition

In any patient having epilepsy you have to define:

- Provoking factors
- Aura (pre-ictal) If it is there.
- Ictus The event itself.
- Postictal
- Interictal What is happening to the child in between the seizures (inter-ictal)

Classification
of seizures

- Aetiology
- Clinical semiology
- Anatomical location

Aetiology of Seizures

Epilepsy

Non-epilepsy

- **Epilepsy**
- Primary /Idiopathic (70-80%) Cause is not known.
- Secondary / Symptomatic -> something is affecting the cerebral hemispheres
  - Cerebral dysgenesis /malformation
  - Cerebral vascular occlusion
  - Cerebral damage , e.g.
    . Congenital infection
    . Hypoxic-ischaemic encephalopathy
    . Intraventricular haemorrhage/ ischaemia
  - Cerebral tumour
  - Neurodegenerative disorders
  - Neurocutaneous syndromes -> Like Neurofibromatosis or Tuberous sclerosis

- **Non-epileptic**
  - Febrile convulsions
  - Metabolic
    . Hypoglycaemia
    . Hypocalcaemia/ hypomagnesaemia
    . Hypo/ hyprenatraemia
  - Head trauma
  - Meningitis/ encephalitis
  - Poisons/ toxins

The Epilepsies of Childhood

- Incidence : 20-50/100,000 General population
  0.05 % * USA 104/100,000
  (after 1st yr. / more common)

Most of these occur after the first year of life.

- Prevalence : 1.3-10% childhood population
  0.5 % * USA 6.8/100,000
  (6 children with childhood epilepsy/(per)
  large 2nd schools)
  1 in 200 children

Classification of seizures has passed through many stages over the years. This is an old one.

International Classification of Seizure Type

I. Partial seizures
   - A. Simple partial seizures
     1. With motor signs
     2. With somatosensory or special sensory hallucinations
     3. With autonomic symptoms
     4. With psychic symptoms
   - B. Complex partial seizures
     1. Simple partial followed by impairment of consciousness
     2. With impaired consciousness at onset
   - C. Partial seizures evolving to secondary generalized seizures
     1. Simple partial seizures evolving to generalized
     2. Complex partial seizures evolving to generalized
     3. Simple partial seizures evolving to complex partial seizures evolving to generalized

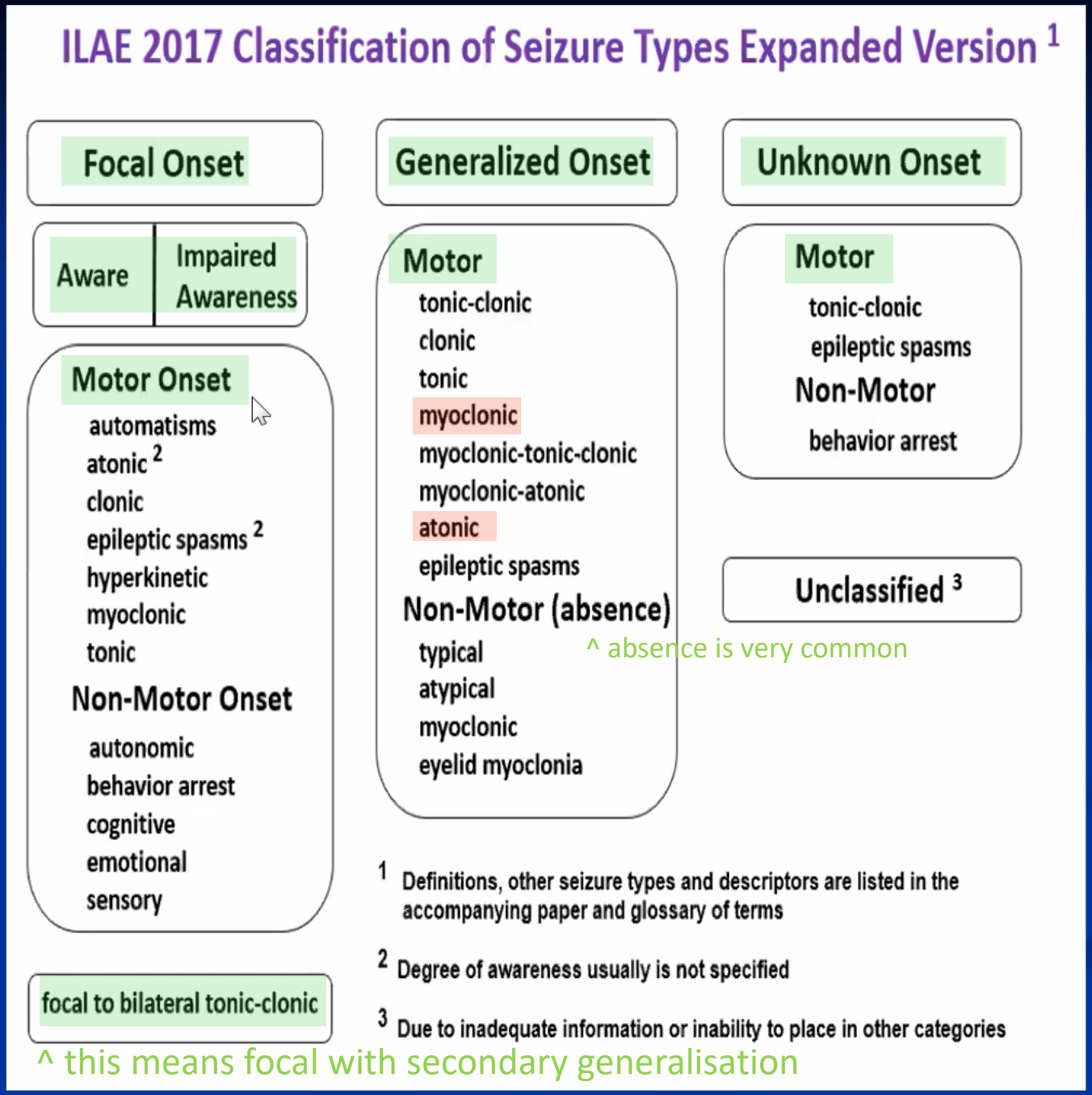
Simple vs complex = depends on level of awareness of child.

II. Generalized seizures
   - A. (1) Absence seizures
     (2) Atypical absence seizures
   - B. Myoclonic seizures
   - C. Clonic seizures
   - D. Tonic seizures
   - E. Tonic-clonic seizures
   - F. Atonic seizures

III. Unclassifiable epileptic seizures

Latest classification (done in 2017) divides seizures based on the onset: focal, generalized, unknown onset.

Then in each category we have the motor component and nonmotor component.



^ absence is very common

^ this means focal with secondary generalisation

From the start you have to decide if the child is aware or has impaired awareness.

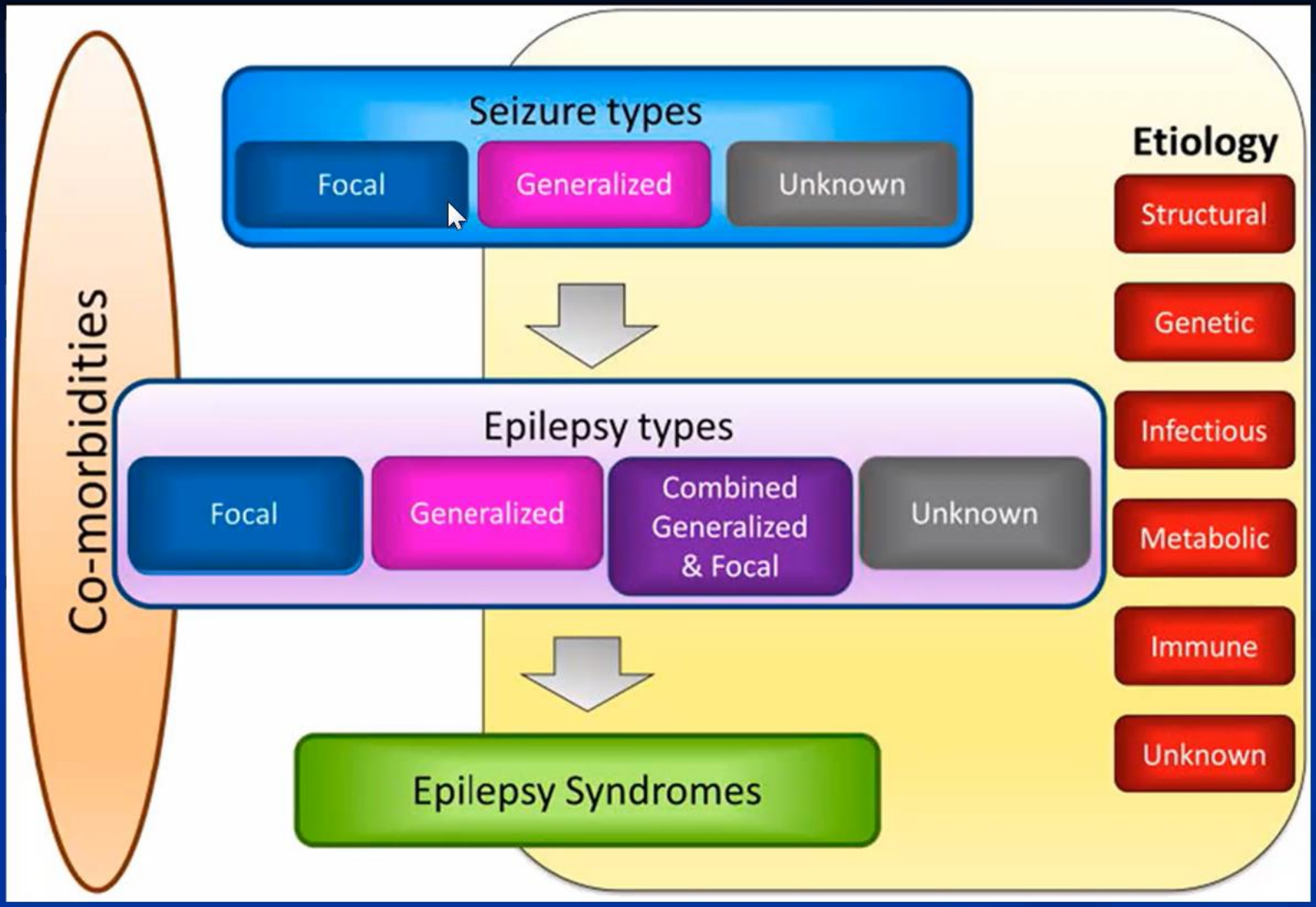


Figure 2. Schema for classification of epilepsy based on the International League Against Epilepsy. Reprinted with permission from Scheffer IE, Berkovic S, Capovilla G, et al. ILAE classification of the epilepsies: position paper

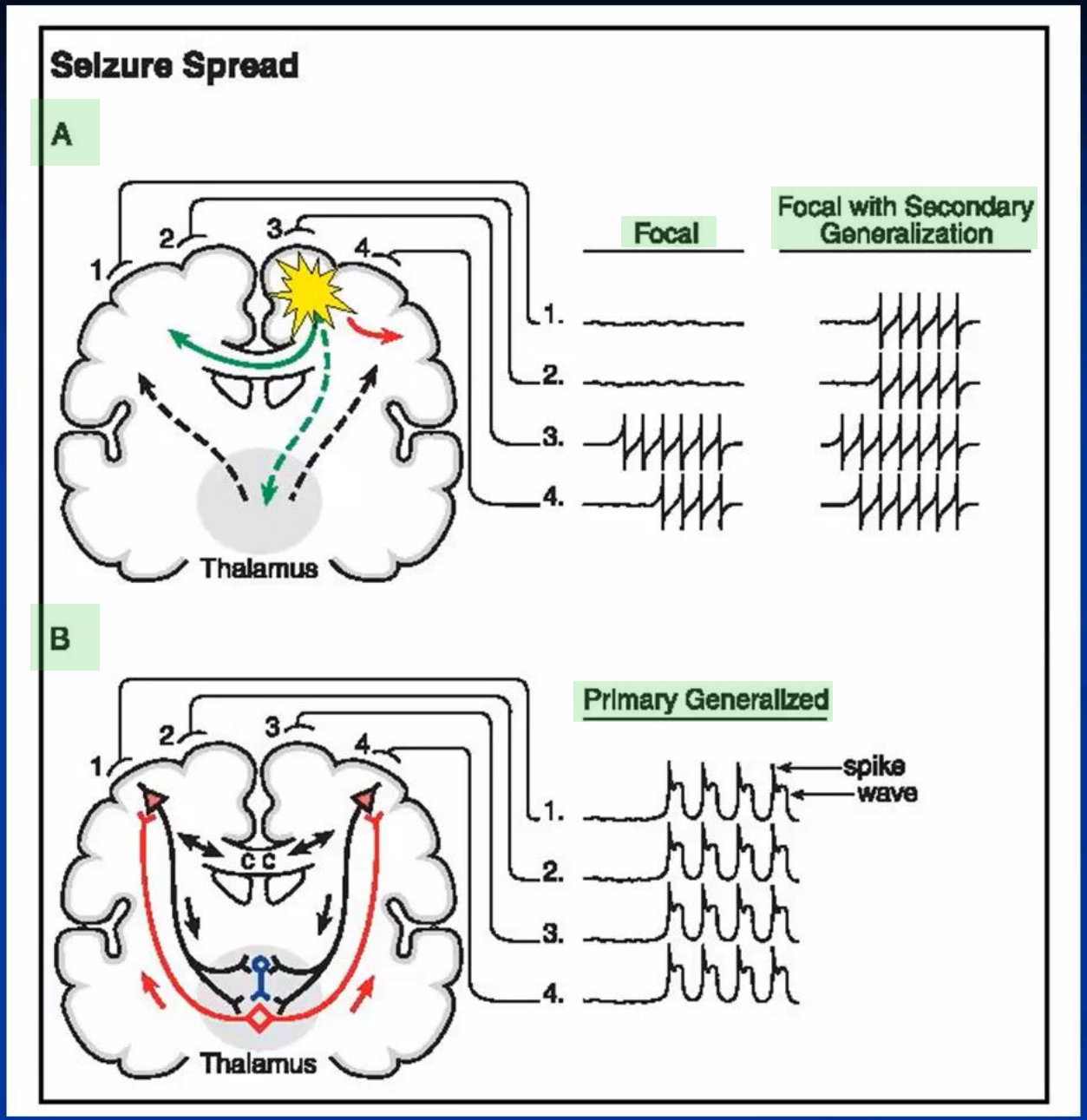
Epilepsy is also divided into categories like seizures then we have the epileptic syndromes. On the side we see the different causes or etiologies (in red).

An International Classification of Epilepsy

- Generalized epilepsies
- Focal epilepsies (partial seizures)
- Epilepsy syndromes

In focal (A) the electrical discharge will come from **one specific part of the hemisphere** and this will be reflected in the electrodes. As you can see in number 3 and 4 only.

These discharges might **spread** through corpus callosum (green line) or through the thalamus (dotted lines) and the child will have focal with secondary generalization (so all the areas will show activity: 1, 2, 3, and 4)



In generalized (B) the electrical discharge is coming from **both hemispheres simultaneously** , so the patient will have all electrodes showing an alert of the electrical discharge.
The child will be have **loss of consciousness** and there is **no warning** signs or aura.
The pt will have bilateral symmetrical seizures and the EEG will show **synchronous discharge**.

In focal the child will have an **aura** which reflects the site of origin.
The level of consciousness might be impaired but **not completely lost**.

Focal Epilepsies (Partial seizures)

- Arise from one or part of one hemisphere
- Begin in a small group of dysfunctional neurones in one of the cerebral hemispheres
- May be heralded by an aura which reflects the site of origin
- May or may not be associated with change in consciousness or more generalised motor jerking

Epilepsy Syndromes

- **Generalized Epilepsies**
  - Infantile spasms
  - Childhood absence epilepsy
  - Lennox-Gastaut syndrome
  - Juvenile myoclonic epilepsy

- **Partial Epilepsies**
  - Benign rolandic epilepsy (BCECTS)
  - Occipital epilepsy

^These are the common known ones.

Absence Seizures (Petit mal)

- Account for only 5-10% of childhood epilepsy
- Age : 4-12 years , 2/3 are Female
- Characteristic :
  - Transient loss of consciousness
  - An abrupt onset & termination
  - Unaccompanied by motor manifestation/ only flickering of eyelids , twitch of hand minimally & minor alteration of muscle
  - Last few sec – 30 sec
  - Child look puzzled / missed something
  - Interfere with school performance

<- There is no motor component just staring, and sometimes flickering of eyelid or twitching of the hand.

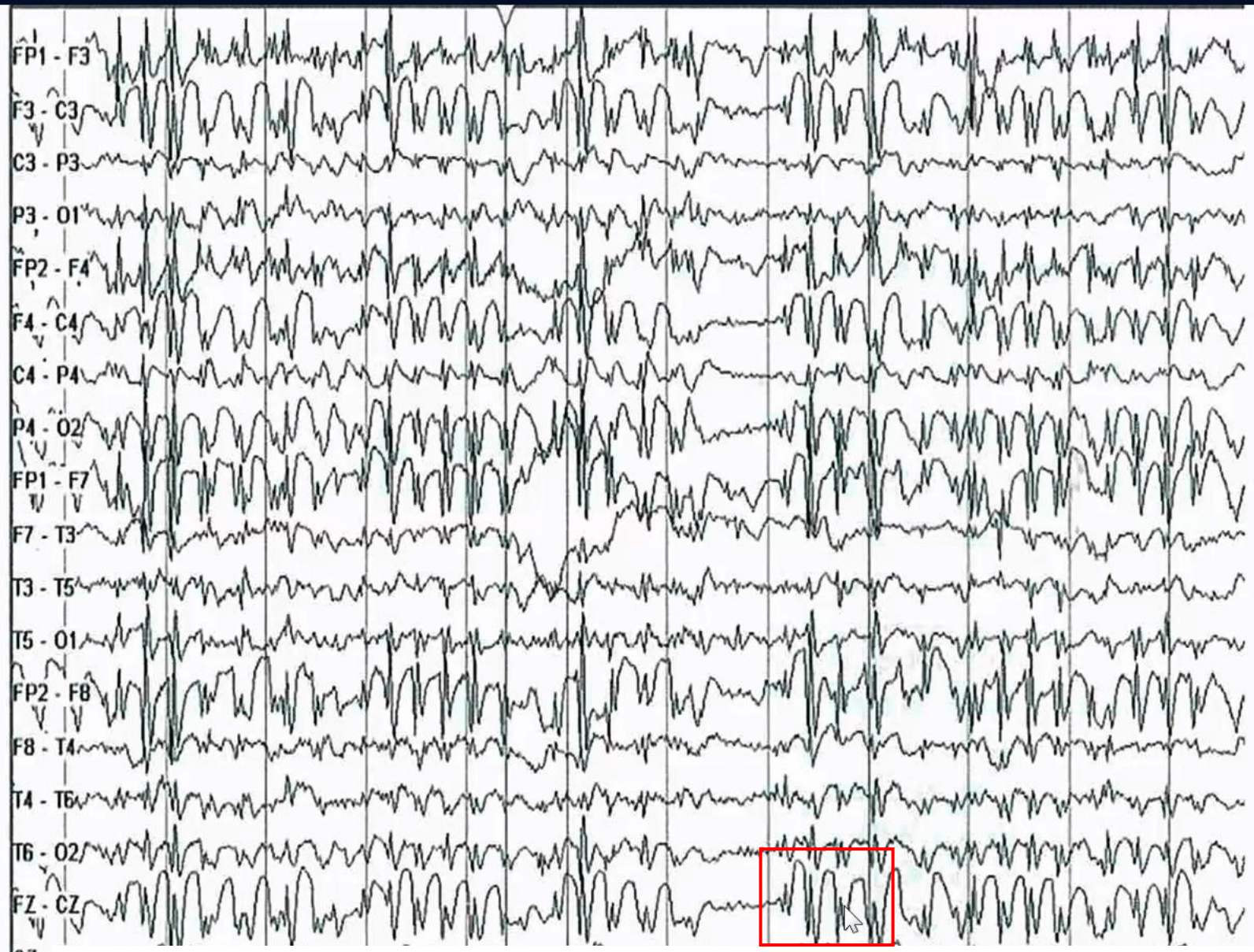
because it can happen several times (up to 100 times) a day.

Absence Seizures(Petit mal)

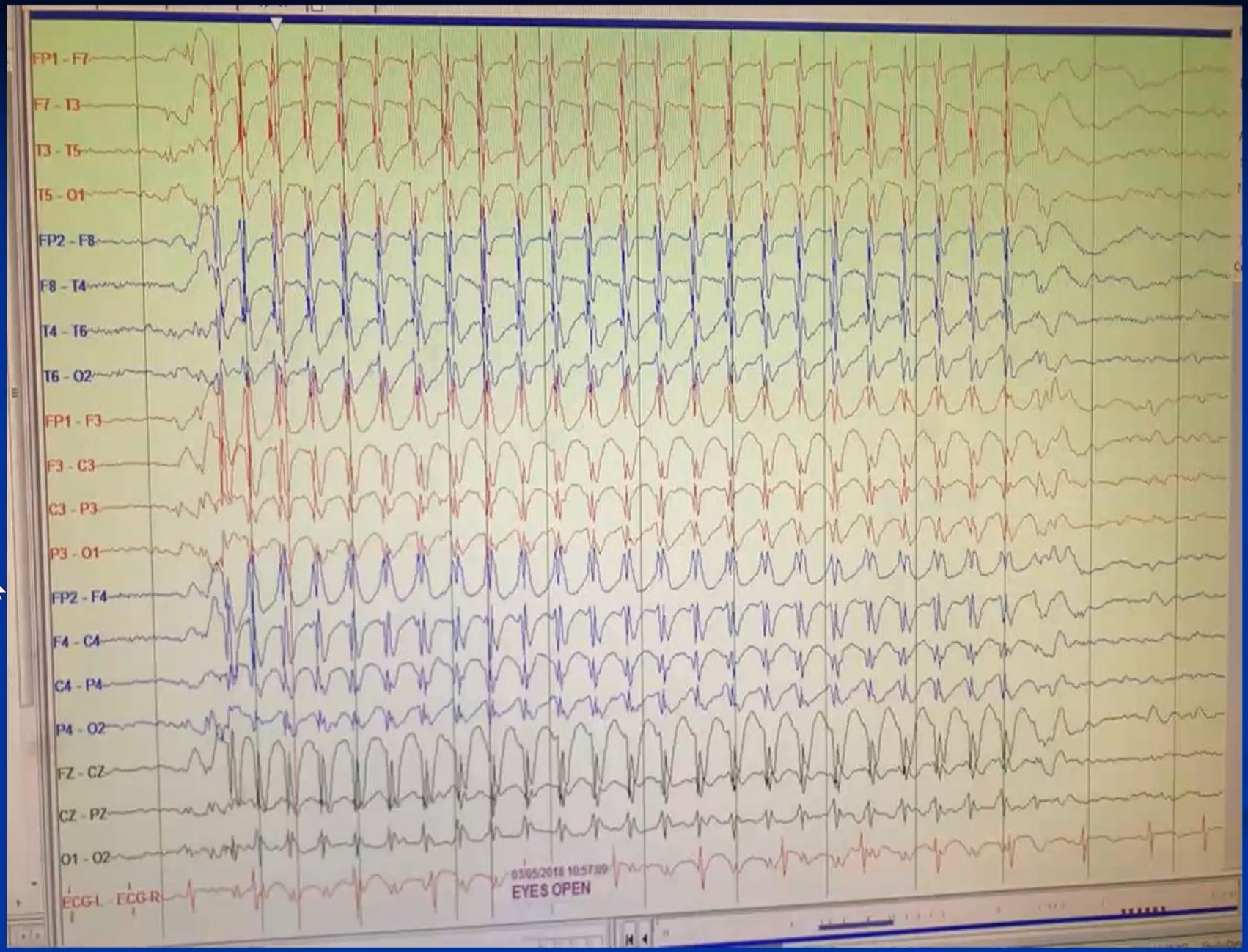
- Often provoked by hyperventilation (2-3 min) You can ask a child to **hyperventilate**: blow on a paper or a wheel for 2 – 3 mins and this will **provoke** the seizure.
- EEG :

  Generalized bilateral synchronous 3/ sec. spike & wave discharge during / between attacks

- Rx : 1st line : Ethosuximide ,Valproate
  2nd line : Lamotrigine
- Prognosis : good Prognosis for these pts is excellent.
  95% remission / adolescence
  5-10% may develop / GTC seizure in adult life



As you can see here, the pt will have spike then slow wave, spike then slow wave, spike then slow wave. There will be **3 per second** and this is **characteristic** for absence. The outlined part is one second and the whole paper is 10 seconds.


FP1 - F7
F7 - T3
T3 - T5
T5 - O1
FP2 - F8
F8 - T4
T4 - T6
T6 - O2
FP1 - F3
F3 - C3
C3 - P3
P3 - O1
FP2 - F4
F4 - C4
C4 - P4
P4 - O2
FZ - CZ
CZ - PZ
O1 - O2
ECG-L
ECG-R
EYES OPEN

Juvenile myoclonic epilepsy

- Age : Adolescence- adulthood — Starts in adolescence meaning from age 12-13 and above.
- Characteristic : throwing drinks in the morning 2nd to myoclonus
- Seizure type : 1- Myoclonic (mostly)
  2- GTC
  3- Absences
- EEG : Characteristic
- Hereditary : A genetic linkage / identified
- Rx : 1st line Valproate
  2nd line Lamotrigine
- Prognosis : good / no learning impairment
  respond to Rx / usually lifelong

They have no learning impairment but they will need to take their medication lifelong.

The scenario will be:
The mom will come to you saying my child is **having weird behaviour**. He is **throwing** his cup of milk in the morning. This throwing of the drink in the morning is secondary to **myoclonus**.
She will also say "I'm calling to him and he is **not responding** to me, he's just **staring**" -> so he is having **absence**.
Then she'll say I discovered yesterday that he was wet his bed -> so he is having **nocturnal enuresis** which reflects **generalized tonic-clonic seizure.**

Tonic-Clonic Seizures

- Generalized ↑ in tone/ muscle contraction
- Rigid tonic phase/ fall/injury
- No breathing /cyanosis
- Clonic phase /jerking of limbs
- Breathing is irregular/ excessive saliva
- Biting of tongue / incontinence of urine
- Seizure last few sec.-min
- Followed by unconsciousness or deep sleep for several hrs.
- Rx : 1st line : Valproate , Carbamazepine
  2nd line : Lamotrigine ,Topiramate

نفس الكلام اللي مكتوب بس مرتبته بشرح الدكتوره

First the patient will have increase in tone -> become rigid -> fall to the ground -> there is no breathing -> then he will enter into clonic phase -> jerking of the limbs -> breathing is irregular -> he may have excessive salivation and might bite his tongue -> then there will be incontinence of urine.

It lasts for a few minutes then the patient will enter the postictal phase during which he might go into deep sleep for several hours

Atonic Seizures

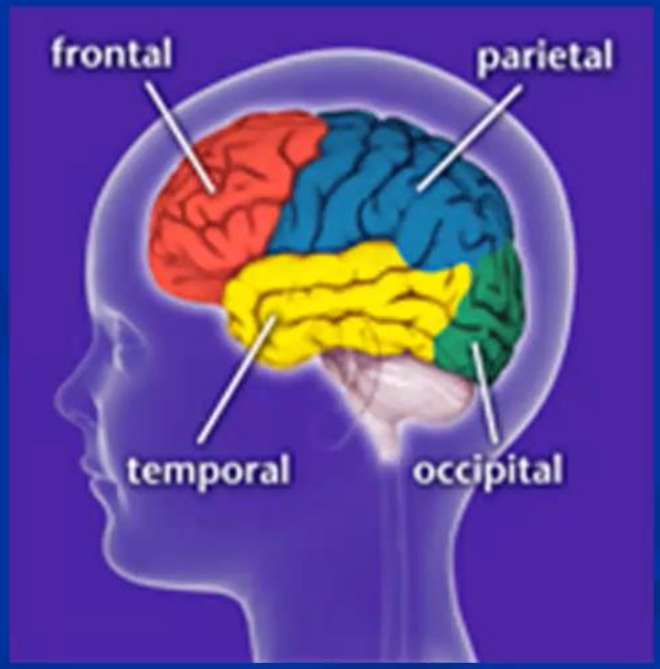
- Transient loss of muscle tone / fall to the floor or droop of head
  Loss of muscle tone so if he is standing he will fall to the floor and if he is sitting he will have droop of the head.
- Differential Dx :
  non – epileptic myoclonic movements
  - Physiologically – hiccoughs اللي يسمونها الحادوقه او الشرقه
    (myoclonus of the diaphragm)
  - Passing through stage II sleep
    (sleep myoclonus)
    <- Sometimes when you are sleeping and you feel like you are passing through a hole and you just pull your leg, this happens when you go in the second stage of sleep and it is normal.
- Rx : Valproate

You have to differentiate atonic from myoclonic seizure, so we usually ask the mom: "لما ولدك طاح، طاح زي الورقة ولا زي الشجره؟" . What is the difference? The one that falls like a paper will not hurt himself while the one that falls like a tree will injure his head. So both of these are minor motor symptoms, and both of these can result in drop attack, and both of these respond very well to valproate.

Focal Seizures

- The manifestations will depend on the part of the brain where the discharge originates

- **Types :**
  - Frontal seizures
  - Temporal seizures
  - Occipital seizures
  - Parietal lobe seizures



Frontal seizures

- Involve the motor cortex
- Lead to clonic movements that travel proximally (Jacksonian march)
- Bizarre , hyperkinetic , asymmetrical tonic seizures
- Differential Dx : non-epileptic events

The most important differential diagnosis is **parasomnia** which we will discuss later.

انا بس اعطيكم فكره عن الـchildhood seizures.
You don't have to know everything

Temporal lobe seizures

- The most common of all epilepsies (15%)
- Result in aura (strange warning feelings)/ 2nd to spread to the pre-motor cortex
  - Smell /taste abnormalities
  - Distortions of sound /shape
  - Lip-smacking
  - Plucking at one´s clothing يعني يشد على ملابسه
  - Walking in a non-purposeful manner (automatisms)
  - Deja-vu and jamais-vu : intense feeling of having been, or never having been, in the same situation before
- Consciousness is impaired
- Duration : longer than a typical absence.

The child will say to his mom: "I am smelling something" or "I am tasting something abnormal".

For example he will enter the clinic totally fine, then suddenly he will start walking in a non purposeful manner and he is not responding to the call of his name.

^It lasts for minutes unlike absence which is only for seconds

Benign epilepsy

For occipital: It Is a benign form of seizures.
Older age group will have visual aura, the child will have hallucination so he will say he see's something.
Then after that he start to have tonic clonic seizures followed by migraine like headache and nausea.
Very characteristic: this happens when the child is in the transition zone from wake to sleep.
Induced by photic/video games so the mom will say he is having frequent attacks b/c he has been playing more video games

For Rolandic:
Mom will tell you that the child was in the room and suddenly I heard gargling from the child's room. I enter the room and find my child sitting in the bed staring at me, having speech arrest, he might also have some secretions, twitching of the face and tonic movement involving one of the arms.
Most of the seizures last for **seconds to minutes** then **half** will go into **generalised tonic clonic.**
They are usually **infrequent** and may come just once a year or once every 4-6 months. So this type of seizure because it is infrequent **we don't treat.**

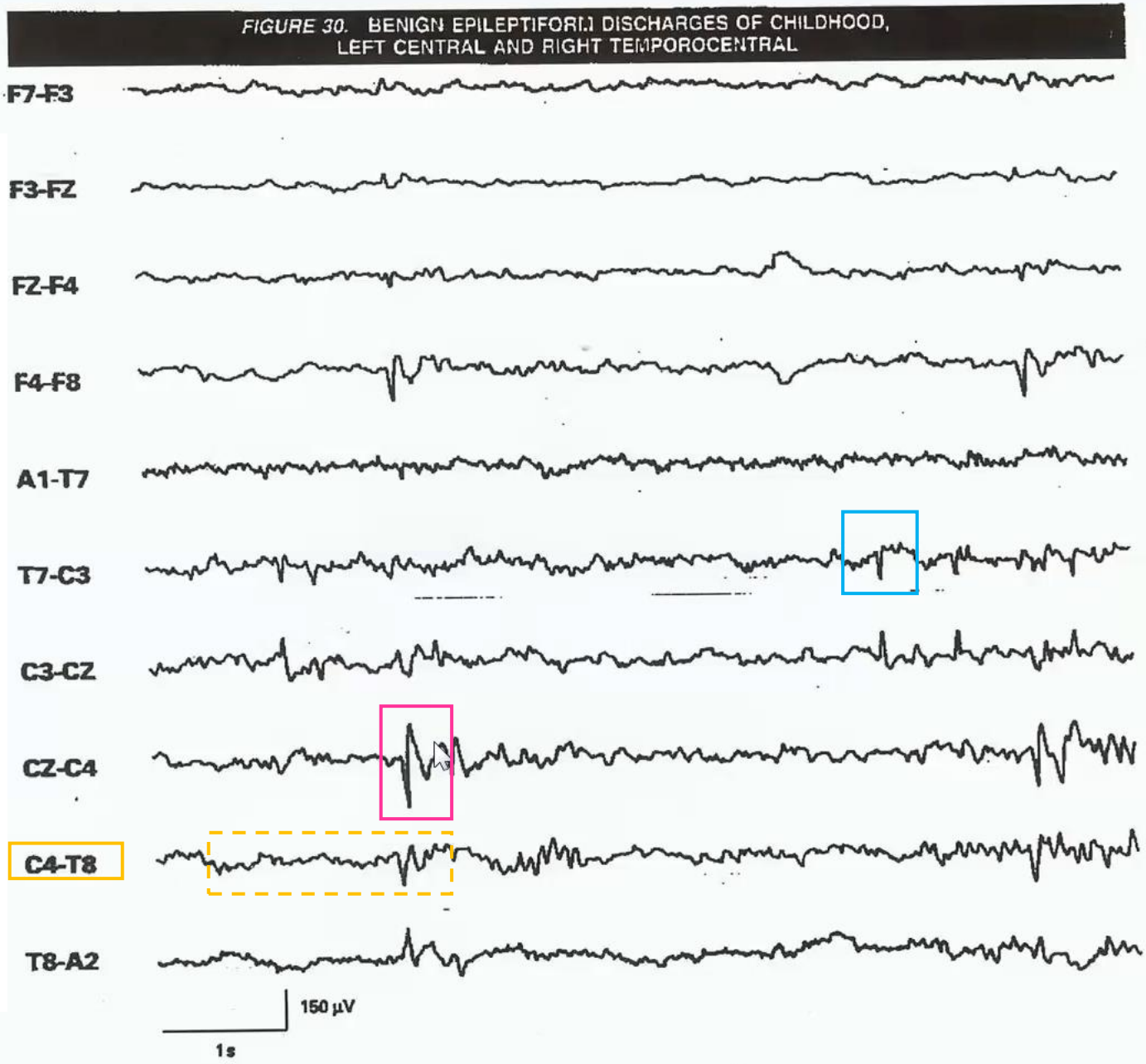
	Rolandic	Occipital
Age	3-13y(Peak 5-10y) M>F	15mon-17y (Peak 4-8y) Younger age
Genetic	autosomal dominant -> AD 40% close relative with febrile/epilepsy	AD 1/3 of Patient +ve family Hx of Rolandic seizure.
Mentality	Normal	Normal
Clinical Manifestation	Unilatral parasthesia of the face (lip tongue & cheek) twitching of the face mouth & Pharynx, speech arrest, Dysarthria, ½ develop GTC (Nocturnal), consciousness preserved.	Visual Aura (Hallucinations, blindness & illusion) followed by hemiclonic seizure (CPS, GTC or automatism) migraine like headache & nausea.
Frequency	70 % during sleep, 15% during awake, 15% both. (10%-one seizure, 70% infrequent, 20% frequent)	More during transition from wake to sleep, induced by photic/Video games
Duration	1-2 min	6-10 sec
EEG	Interictal: Unilateral/bilateral high voltage spike discharge at cetrotemporal region, activated by sleep & drowsiness	Ictal spike at occipital lobes Interictal: Unilateral/bilateral high voltage spike or sharp discharge at occipital or posterotemporal regions of both hemispheres, (More with eye closure)
Outcome	Stop spontaneously by age 14y even without treatment, normal IQ	Ceases spontaneously by12y (if persistent/structural abnormality)DDx Basiler Migraine.

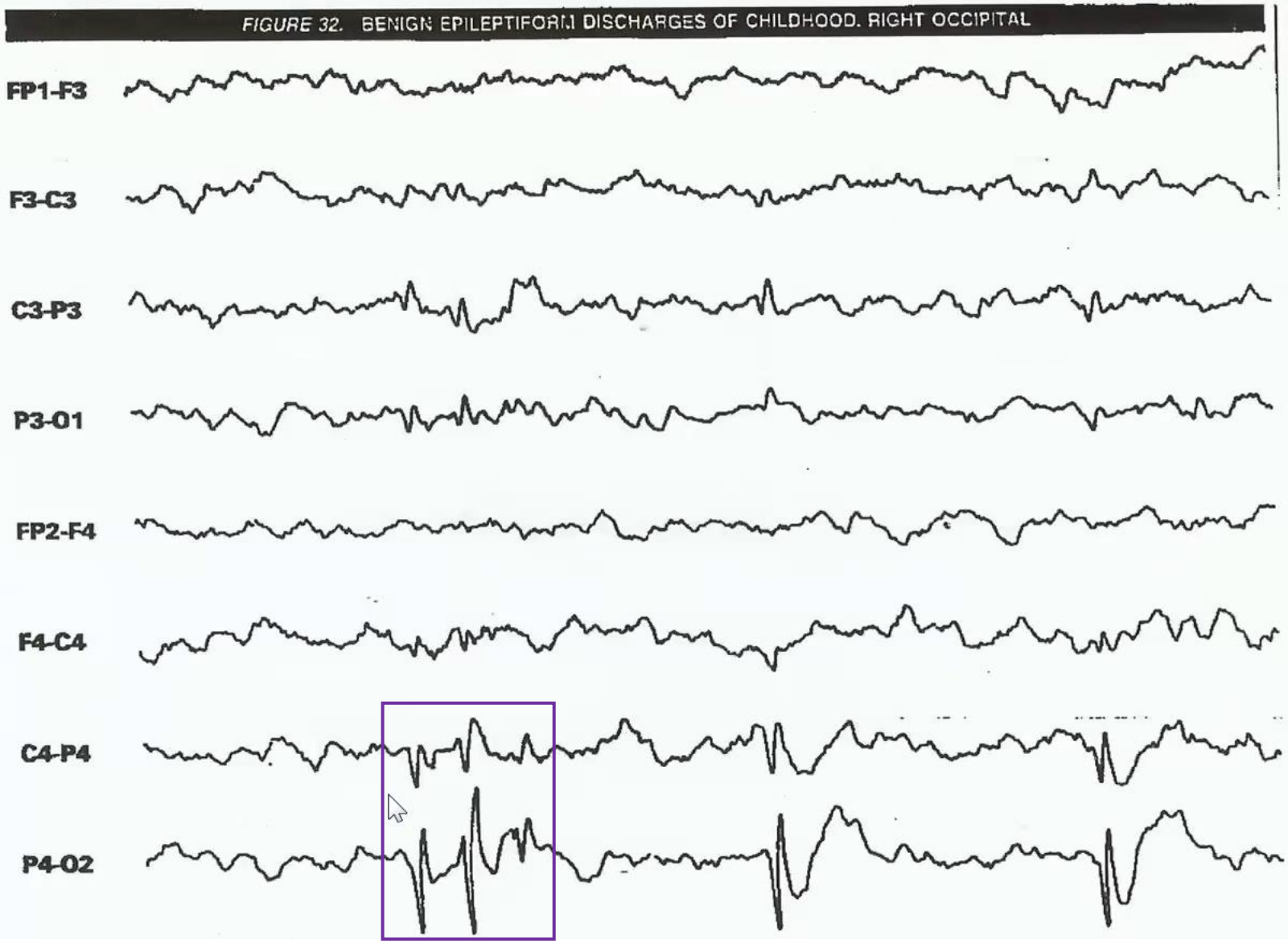
Both will disappear completely by adulthood. If it is persistent in the occipital you have to rule out basilar migraine. As for treatment both of them respond very well to **carbamazepine** (first line)

The Rolandic comes from centro-parietal / from centro-temporal.

Odd numbers (like 3) indicate the left and even numbers indicate the right.

Here the pt has central affection on the left (blue box) and central affections on the right (pink box). There is also temporal affection on the right (C4-T8) **I'm not sure where the doctor pointed**





The occipital, from its name, comes from the parieto-occipital. So we can see the discharges here (purple box) and once we ask the patient to close their eyes they will be more pronounced, this is photic type of seizure.

Visual Aura

These are other conditions that can come with visual aura:
(anything that affects the visual cortex can cause aura)

- Hypoglycaemia
- Toxins ie Cyclosporine
- Hysteria
- ? Psychosis
- Benign occipital epilepsy
- ? Basilar migraine
- TIAS
- Demyelination ie MS , ADEM
- Occipital metastic lesions

These are other conditions that can come with visual aura:
(anything that affects the visual cortex can cause aura)

- Photosensitive Epilepsy : Avoid visual stimulation e.g. video games , flash lights , T.V , Computers

Aura

The aura depends on the site.

- Mesial temporal lobe
  Epigatric sensation
- Amygdala of temporal lobe
  Psychic symptoms ie fear
- Visual association cortex in the posterior temporal cortex
  Visual hallucinations
  (person , animal , object)
- Uncus of the temporal lobe
  Olfactory aura / smelling foul odors (uncal fits)
- Temporal neocortex
  Auditory aura

- Occipital lobe
  Epileptic visual aura
  Positive : flickering lights , spots , lines
  Negative : blindness

Migraine aura
Develop over 5 min
Last for longer time
No disruption of /or LOC
Headaches occur during / or within 60 min of visual aura

<- I want you to know this b/c it is one of the ddx of occipital lobe epilepsy

- Parietal lobe
  Sensory aura
  Positive : pins & needles
  Negative : numbness

Focal epilepsies

- **First line Rx**
  - Carbamazepine
  - Valproate
- **Second line Rx**
  - Topiramate
  - Lamotrigine
  - Levetiracetam
  - Oxcarbazepine
  - Gabapentin
  - Tiagabine
  - Vigabatrin

Difference between primary generalized and partial epilepsy.

	1st Generalized	Partial
1. Aetiology	Idiopathic 25% genetic base	Focal lesion
2. Onset	Usually > 3 years of age	Variable
3. Seizure Pattern	Generalized	Focal
4. Neurological Sign	Normal, rarely focal	Common (focal)
5. EEG	Generalized	Focal onset or focal with 2nd generalization
6. Brain CT Scan	Normal	May be abnormal
7. Reason to RX	Good	Variable, some-refractory

Epileptic Syndromes

Let's review:

If you have a patient having a **staring like look** what is the ddx?

It might be **epileptic** or **nonepileptic**.

If it is **epileptic** it might be **absence** seizure or **complex partial seizure**.

How to **differentiate** between these 2?

- In absence the staring occurs up to 100 times per day while in complex partial it is 3-4 times per day.
- The staring of complex is preceded by aura while there is no aura in absence.
- Absence recover instantly while complex-partial will go into postictal drowsiness with confusion, or slurred speech.
- Doing EEG will show generalised in absence and focal in complex partial.
- Treatment: for absence is ethosuximide or valproate while the other we give carbamazepine.

If it is **non epileptic** it might be **daydreaming** or **depression**, **ADHD** or **autism**.

Infantile Spasm

The most important epileptic syndrome is infantile spasm.

- Age : 4-6 months
- Clinical Types :

  * Flexor
  * Extensor
  * Mixed flexor & extensor
  * Arrest / akinetic

Infantile Spasm

- Characteristic :

  * ´Salaam spasms` : flexor spasms of head , trunk & limbs followed by extension of arms
  * Flexor spasm last for 1-2 sec.
  * Cluster of 20-30 spasms
  * On awakening
  * Repeated many times / day

- Differential Dx : Infantile colic

- Developmental Hx : deterioration in social interaction

The mom will say the baby is having flexion of the upper extremities and drawing his lower limbs to his abdomen like he's having an infantile colic so how will you differentiate? Ask her was you child interacting normally, smiling and interacting with the surrounding and after having this movement he started having deterioration in social interaction? Because this is characteristic for infantile spasm.

The name of infantile spasm or salam spasm came from the Japanese performers who go to the theatre and greet people by bending their neck and chest then bring both hands on his chest and say salam.

Infantile Spasm

- Aetiology :

  * Idiopathic / cryptogenic / non-symptomatic

    . 1/3 cases
    . Normal development / Neuro. Exam
    . No cause (neuroimaging / metabolic)
    . Rapid & good response to Rx
    . Prognosis : good

  * Secondary/ Symptomatic

    . 2/3 cases
    . Abnormal development / Neuro. Exam
    . A cause identified
    (neuroimaging / metabolic)
    . ????Refractory / relapse
    . Prognosis : poor <- because they are refractory and do not respond to therapy.

Table 5: Etiological factors associated with infantile spasms

Before birth it is mostly congenital causes

After birth the metabolic causes start to appear

Prenatal Disorders	Perinatal Disorders	Postnatal Disorders
Hydrocephalus	Hypoxic-ischemic encephalopathies	Pyridoxine dependency *
Microcephaly	Meningitis	Nonketotic hyperglycinemia
Hydranencephaly	Encephalitis	Maple syrup urine disease
Schizencephaly	Trauma	Phenylketonuria
Polymicrogyria	Intracranial hemorrhages	Mitochondrial encephalopathies
Sturge-Weber		Meningitis
Incontinential pigmenti		Encephalitis
Tuberous sclerosis Down Syndrome		Degenerative disease
Aicardi's syndrome		Biotinidase deficiency *
Hypoxic-ischemic encephalopathies		Trauma
Congenital infections		
Trauma		

Modified from Holmes GL and Vigevanoi F. Infantile spasms, In: Engel J Jr. Pedley TA, eds. Epilepsy: a comprehensive textbook. Philadelphia: Lippincott-Raven, 1997.

*2 important causes which may produce infantile spasm and the treatment is just vitamin.

^ the pt will have multiple type of seizure, including infantile spasm, and alopecia, hepatosplenomegaly, sensorineural hearing loss. we diagnose by biotinidase activity which will be low. They respond very well to biotin.

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Prenatal Disorders	Perinatal Disorders	Postnatal Disorders
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Microcephaly	Meningitis	Nc
Hydranencephaly	Encephalitis	Ma
Schizencephaly	Trauma	Ph
Polymicrogyria	Intracranial hemorrhages	Mi
Sturge-Weber		Me
Incontinential pigmenti		En
Tuberous sclerosis Down Syndrome		Degenerative disease
Aicardi's syndrome		Biotinidase deficiency *
Hypoxic-ischemic encephalopathies		Trauma
Congenital infections		
Trauma		

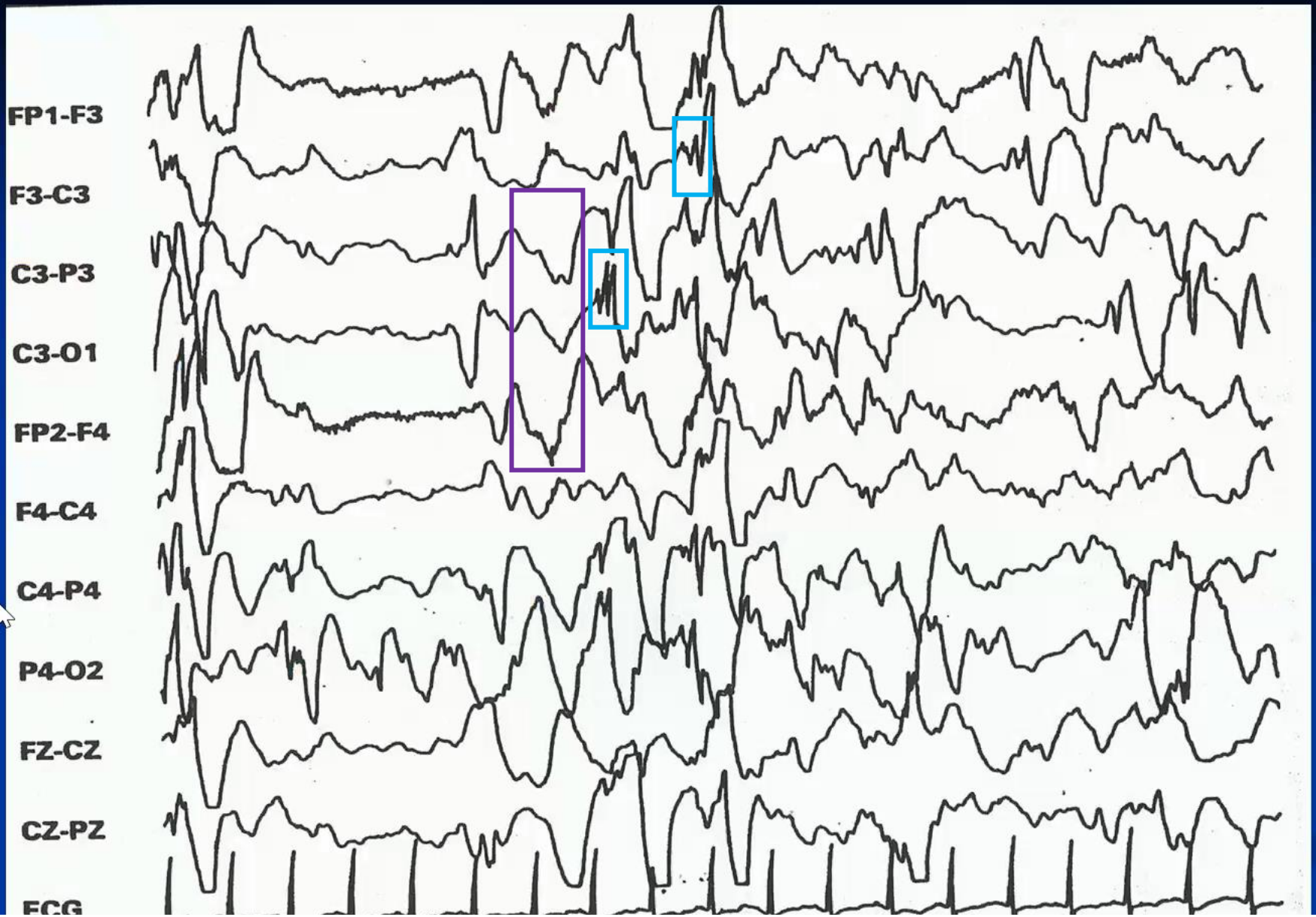
Modified from Holmes GL and Vigevanoi F. Infantile spasms, In: Engel J Jr. Pedley TA, eds. Epilepsy: a comprehensive textbook. Philadelphia: Lippincott-Raven, 1997.

^ the mom will say the child is having increased movement in my tummy compared to his siblings (which means he is convulsing inside the mom), then he will be delivered and will act like someone who is hypoxic ischemic = having apnea and frequent seizures and when we do EEG, we will see infantile spasm so we give antiepileptic but the pt is not responding , then we give pyridoxine and he will respond immediately and should continue this treatment for life..

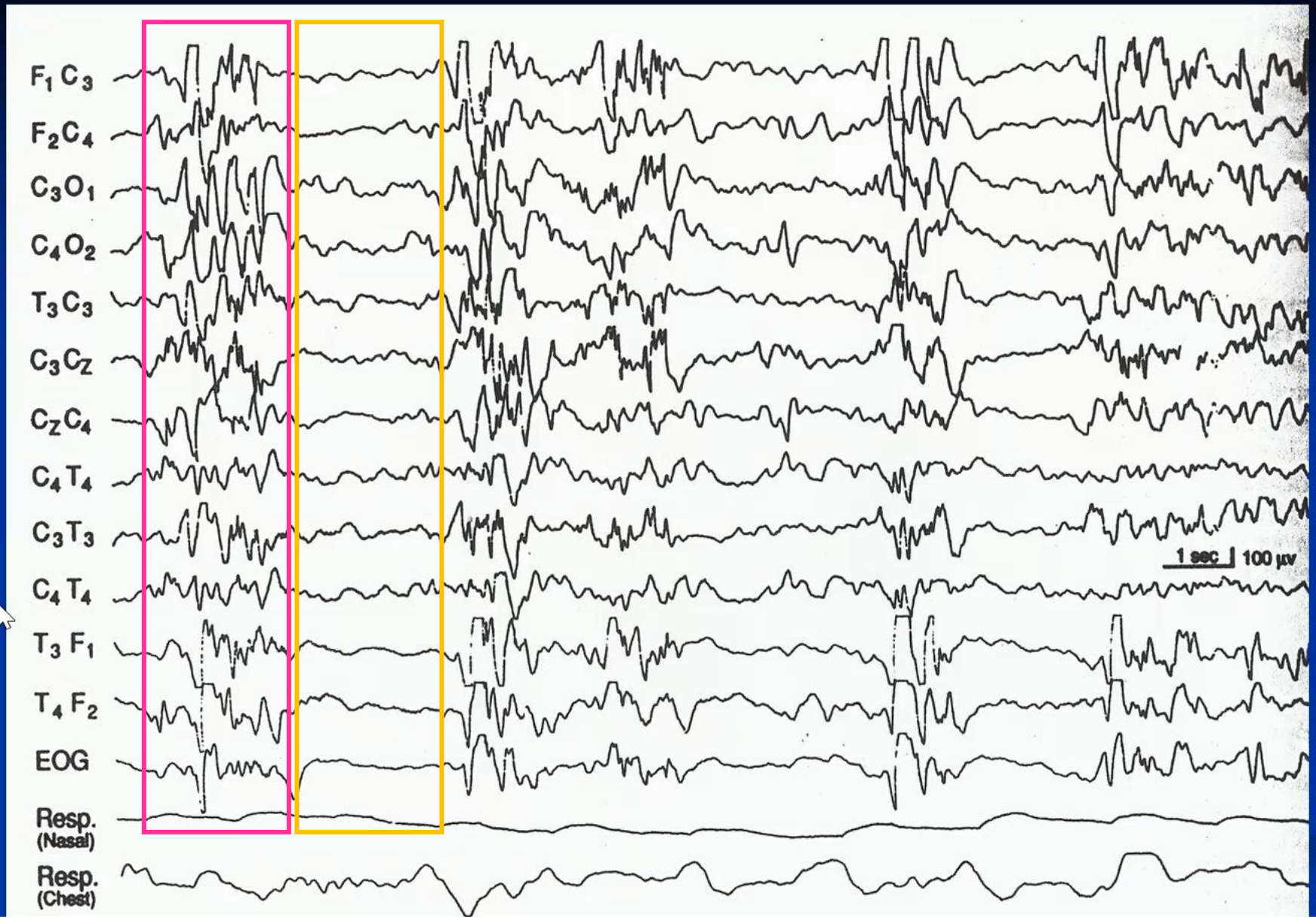
*2 important causes which may produce infantile spasm and the treatment is just vitamin.

Infantile Spasm

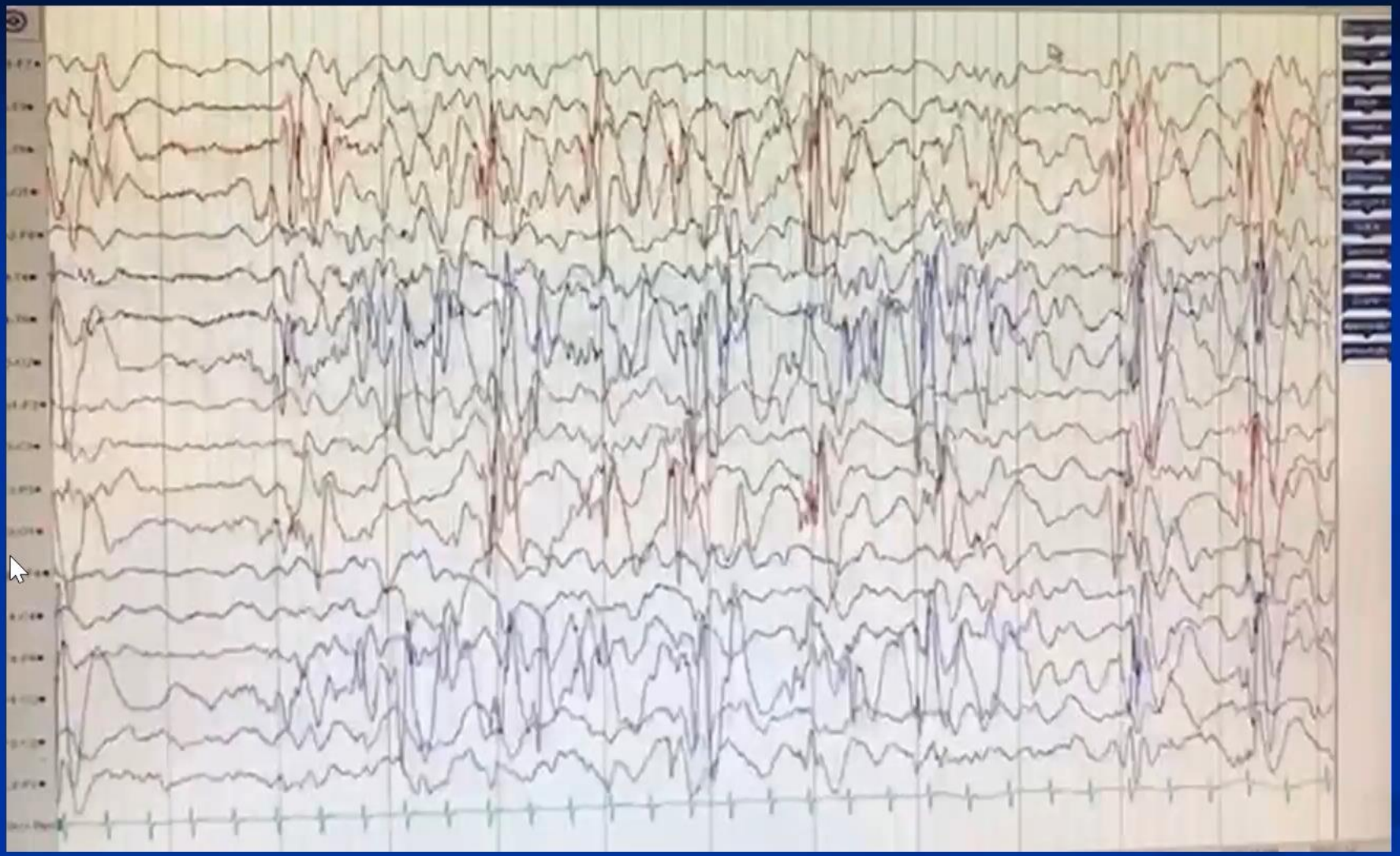
- EEG :
  - Hypsarrhythmia : a chaotic pattern of high-voltage slow waves & multi-focal sharp wave discharges
  - Modified-hypsarrhyyhmia : hypsarrhythmia followed / Burst-suppression pattern
- **West syndrome:**
  1- MR or developmental regression
  MR = Mental retardation
  2- Infantile spasm / myoclonic seizure
  3- Hypsarrhythmia (EEG)



This is an EEG of infantile spasm: he will have **hypsarrhythmia** which means (كأن واحد اخذ المرسام ويشخبط) the background is completely disorganised and the patient is having hypophage slow wave activity and it is intermixed with spikes. It is all mixed with each other and this is hypsarrhytmia.



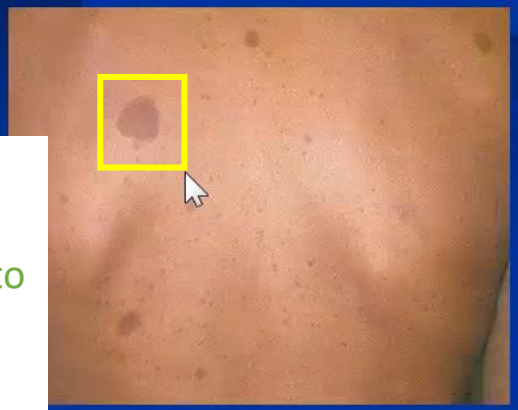
Sometimes a child will have a chaotic pattern followed by partial suppression. This is called modified hypsarrhythmia

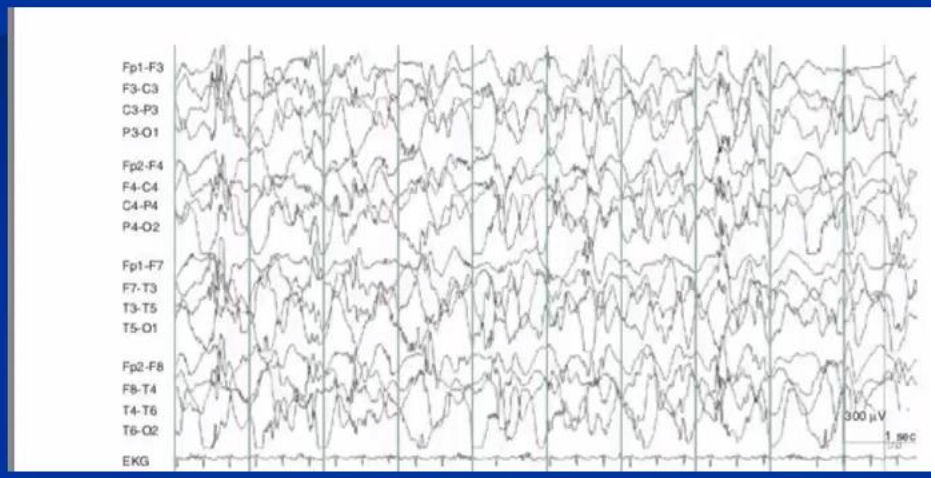


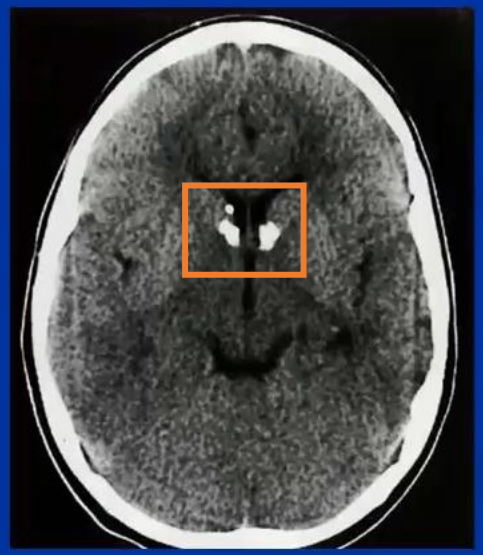
Here the pt is having chaotic pattern which is hypsarrhythmia.

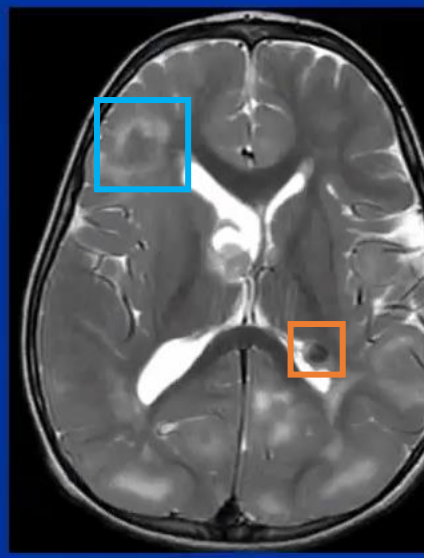
Neurocutaneous syndrome

Tuberos sclerosis complex (TSC)









This is a patient coming to you having infantile spasm and on examination you find multiple hypopigmented patches and cafe au lait spots

And you do an EEG and see the chaotic pattern, and on CT scan there is periventricular calcification. All this goes with tuberos sclerosis (a neurocutaneous syndrome) and on MRI you will see (حبات البطاطا) which means tuberous. The only treatment that can control the infantile spasm here is **vigabatrin**.

Infantile Spasm

- Treatment : **Neurological emergency****

^ You have to start medication within a month of onset to save the mentality of the child.

- Vigabatrin / Pyridoxine
- ACTH , corticosteroid
- Valproate
- Clonazepam
- Topiramate
- Levetiracetam
- Lamotrigine

Vigabatrin needs monitoring with visual field assessment because it can produce toxicity.

We do an EEG and if the child is having infantile spasm (chaotic pattern, hypsarrhythmia) we give **pyridoxine** – if he gets better and he reverts to normal, that's good. If not we give vigabatrin for 2 weeks, if he is not improving give ACTH.
The **idiopathic** type usually respond to **ACTH** or corticosteroid therapy.
Those who have **symptomatic** we may try **topiramate** or the other conventional AED.

MCQ:
Drug of choice for treatment of idiopathic infantile spasm? ACTH therapy
Drug of choice for treatment of infantile spasm with tuberous sclerosis? Vigabatrin.

Infantile Spasm

- Prognosis :
  - Good response to therapy /idiopathic & early Rx , but side-effects are common (40%)
  - Most will have : (60%)
    - Developmental regression
    - Learning disability
    - Epilepsy
    - LGS (20%)
      Lennox Gastaut Syndrome

Those with idiopathic have good prognosis and they only worry about the side effects of rx. But in general infantile spasm is bad (نسميها من التشنجات الصعبه). Because 60% may develop those complications listed.

Lennox-Gastaut Syndrome

- **Triad**

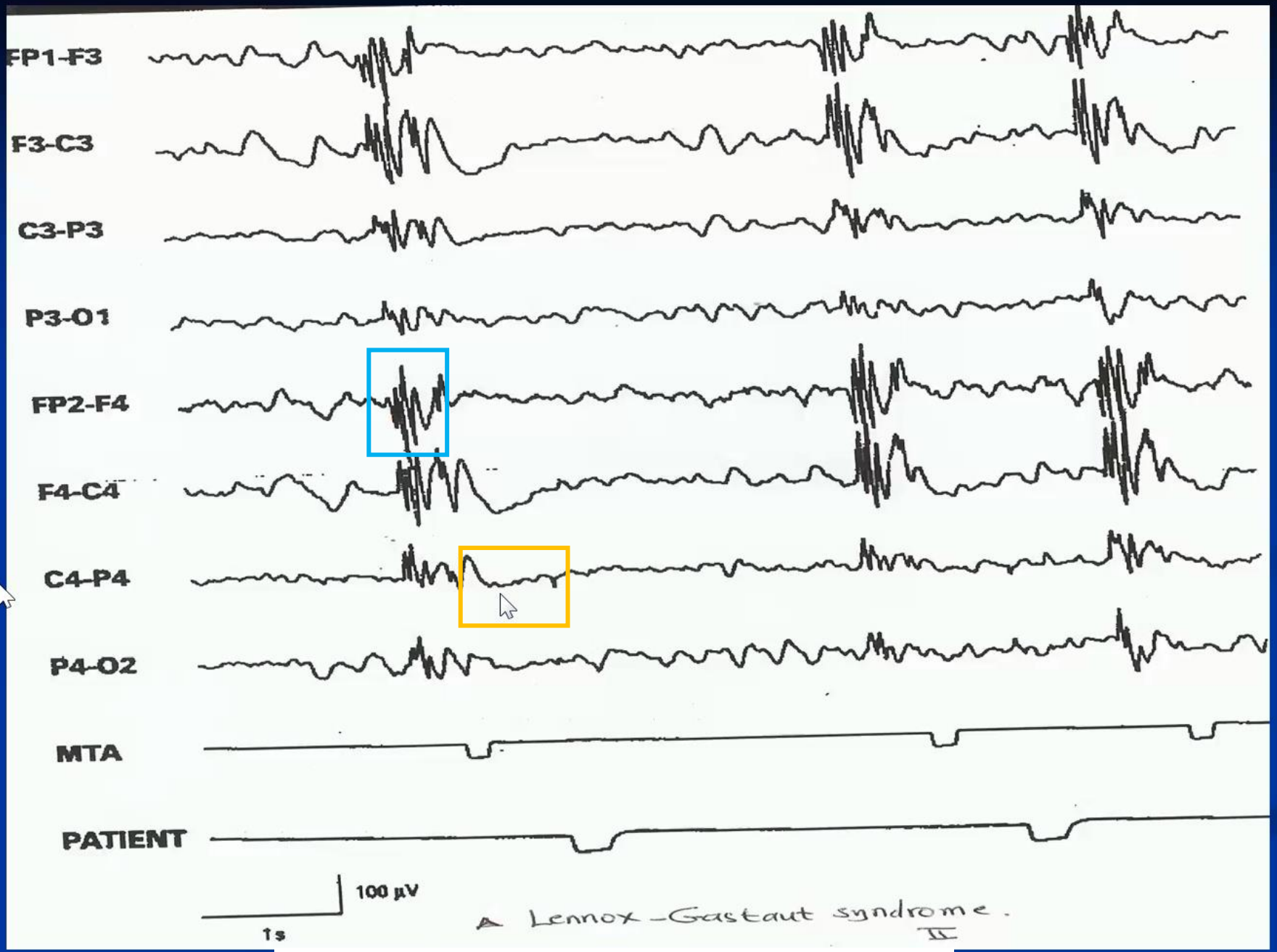
(1) Epileptic seizure. The pt can have all types of seizures:

- Atonic (astatic)/ drop attacks
- Atypical absence
- GTC
- Myoclonic
- Partial seizure

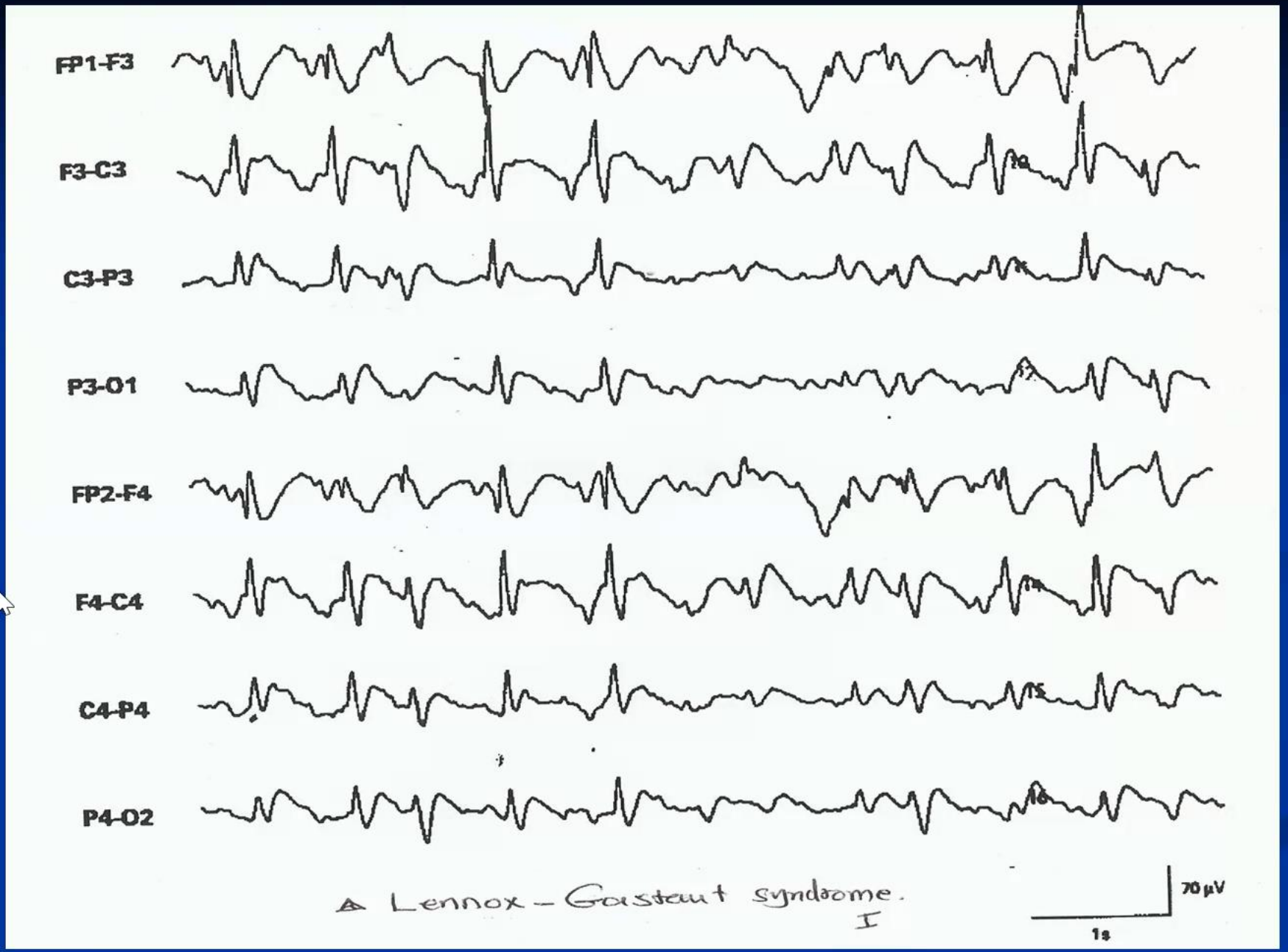
(2) MR (arrest or regression) + Behavioural problem (up to 90% mod-sever) MR = mental retardation

(3) EEG (characteristic)

- Polyspikes
- Spike-slow wave discharges (1.5-2.5 Hz / during sleep)



The will have Poly-spikes and slow wave complexes


FP1-F3
F3-C3
C3-P3
P3-O1
FP2-F4
F4-C4
C4-P4
P4-O2
Lennox-Gastaut syndrome.
I
70 µV
1s

Lennox-Gastaut Syndrome

- Treatment :

  !!!!! Refractory / Prognosis is poor

  Old : Valproic acid
  Benzodiazapin (Clonazepam)
  ACTH
  Corticosteroid

  New : Felbamate
  Lamatrigine
  Toprimate
  Vigabatrin
  IgG IV

  Surgery : VNS
  Corpus callostomy
  (stop atonic seizure & ↓ frequency of GTC)

Because in generalised the seizure spreads to the other hemisphere through the corpus callosum so we cut it and this will reduce the atonic seizures and improve the status of the child.

Diagnosis & Evaluation of 1st Unprovoked Seizure

If you have a child that presented to you with the 1st unprovoked seizure, what will you do?

We start with history.

Medical History

- **History of acute event**
  - Age
  - Timing, duration, frequency
  - Warning symptom (Preictal/aura) before onset of seizure
  - Onset & progression of symptoms
  - Description of episode (ictal)
  - Post ictal occurrence & duration

Note : above need eye witness (reliable observer)

- Precipitating events

<- Don't forget: having a post-ictal phase of drowsiness or sleepiness for more than 4-6 hours always think of something else. For example meningitis, encephalitis, or he is having continuous discharge in his brain. We usually need a reliable eye witness like the mother.

Precipitating Factors

Things that can provoke seizures.

- Fever.
- Irregular or overdose of AED medication. AED= anti-epileptic drug
- Sudden discontinuation of AED.
- Sleep deprivation. الحرمان من النوم
- Fatigue.
- Metabolic derangements. Ex: hypoglycemia, hypocalcemia
- Concomitant use of other medications
  - . Theophyline
  - . Isoniazid
  - . Amphitamine
  - .Tricyclic antidepressent
- Hyperventilation. Can provoke absence.
- Intermittent photic stimulation. Can provoke absence benign occipital epilepsy and juvenile myoclonic epilepsy.

Medical History

- **Past medical history**
  - Headache ,changes in vision, Wt. loss, gait disturbance, speech changes
  - Perinatal complications
  - Head trauma
  - Encephalitis , meningitis
  - Febrile convulsion
  - Developmental history (any regression)
  - Deterioration in school performance
  - Drug history
  - Family history

<- You have to rule out the possibility of tumor so you ask these details.

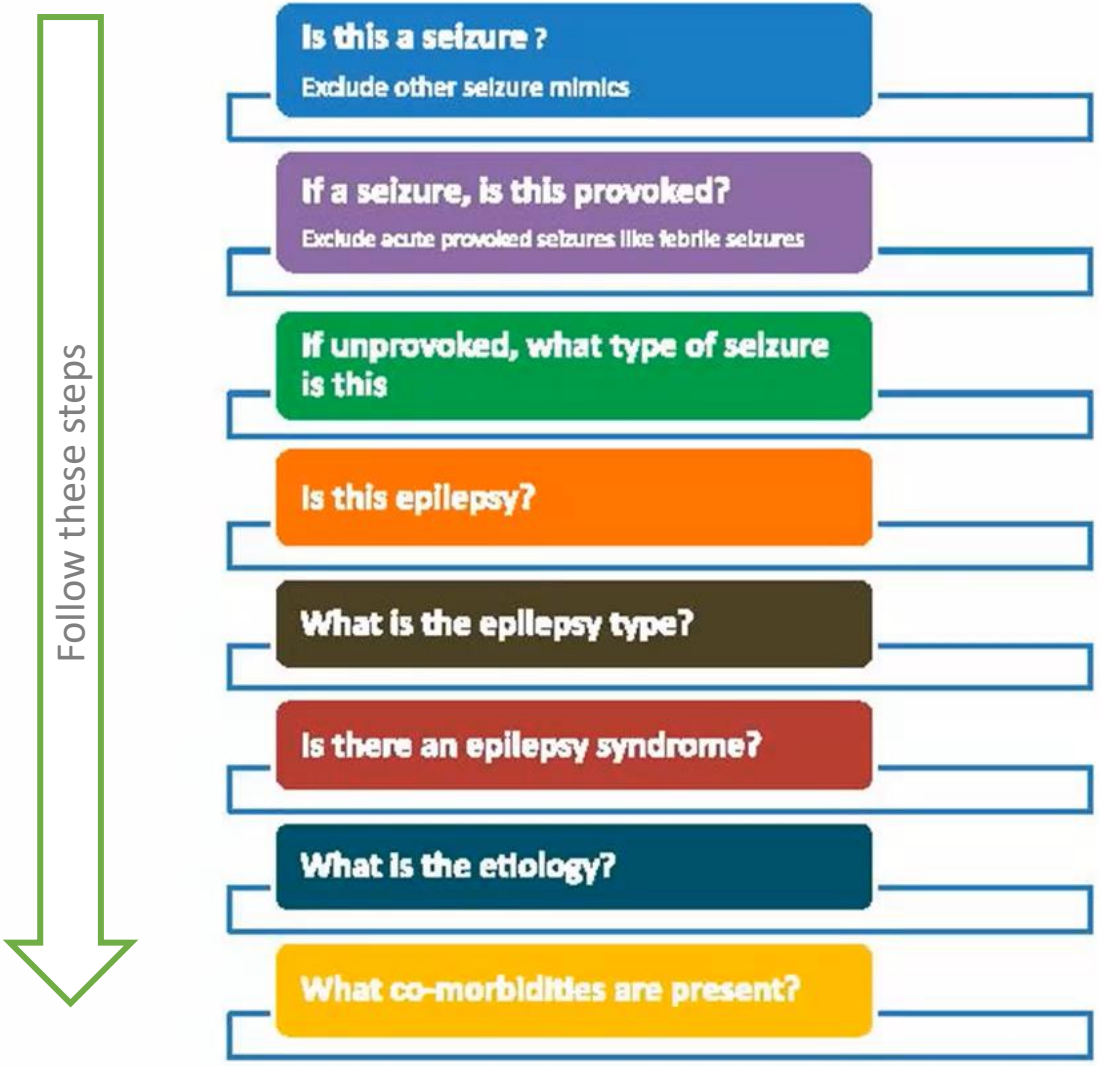


Figure 1. A clinical approach to a child presenting with possible new-onset seizures.

Examination

- Vital signs : Temperature , B.P , Pulse
- Growth parameters : WT , HT, OFC
- Dysmorphic features
- Neurocutaneous stigmata
- For cranial bruit (AVM)
- Organomegaly
- Ophthalmological evaluation
- CNS exam.

  * Mental status / alertness
  * Cranial nerves
  * Focal deficit

- Hyperventilation test (3X min if indicated)
- Developmental assessment

Those with high ICP might have Cushing's triad: hypertension, bradycardia, respiratory problem .

Growth parameters are imp b/c pt with macrocephaly or certain syndromes are prone to have seizures.

Like we mentioned before: hypopigmented patches and cafe au lait spots

Put the stethoscope behind the ear or on the carotid to listen for AVM or aneurysm.

Laboratory Testing

- Complete blood count <- Important as a basic (baseline) because when you start antiepileptic it might cause bone marrow suppression
- Blood sugar
- Blood calcium, magnesium, phosphorus
- Blood electrolytes , BUN ,Cr & CO2
- Liver function test
- Serum & Urine toxicology screen (if indicated) <- We do this for toddler who has somebody at home taking psychotic medication, the toddler might have ingested it.
  Or it could be an adolescent who took street drug medication and developed seizure for the first time.
- AEDs serum levels

Liver function test and AED are done for those who are on AED: valproic acid, carbamazepine, phenytoin, phenobarbital.

Laboratory Testing

These tests are not done for everybody only when indicated:

- Lumbar puncture – Guided by clinical circumstances
  - Persistent fever
  - Very young age
  - Signs of meningitis
  - Persistent drowsiness
  - Neurodegenerative disorders

- Metabolic work-up / developmental regression / seizures related to food or fasting مثلا يقولون لك كل ما صام معنا جته تشنجات وتغيرت ريحة البول (تكرمون)

- Genetic studies
  looking for genetic deletions causing the channelopathies

EEG

- Useful diagnostic tool / not emergency**
- Identify : It helps us identify the seizure type and epilepsy syndrome by telling us about:
  - Asymmetry
  - Focal abnormalities ie sharp waves or slowing
- Suggest underlying abnormalities
- Classify both seizure type & epilepsy syndrome
- Positive : supportive evidence*
  Negative : does not R/o as many children may have normal EEG
  may need record during sleep/ sleep-deprived <- The best maneuver to provoke it is sleep deprivation. We ask the mom to wake the child from 4 am then he sleeps in the EEG and at that time we may record epileptic discharge
- Others : 24-Ambulatory EEG
  forms: Video-telemetry
  Subdural electrodes
  (invasive technique / for surgery)

We don't do it in emergencies unless the child is in the ICU and is having status epilepticus then we can do it.

Neuroimaging

- Structural
  - CT scan (contrast)
  - MRI scan (Greater sensitivity) / flair for MTS

    MRI is better than CT for epilepsy we order epilepsy protocol: especially the flair.
- CT not indicated as emergency* but only

  These are the conditions in which we can order a CT in emergency
  - History of trauma
  - The presence of VP shunt <- Child might have malfunction of the shunt.
  - The presence of bleeding disorders
  - The activity doesn't return to base line post seizure
  - Signs of increased ICP
  - Focal neurological sign

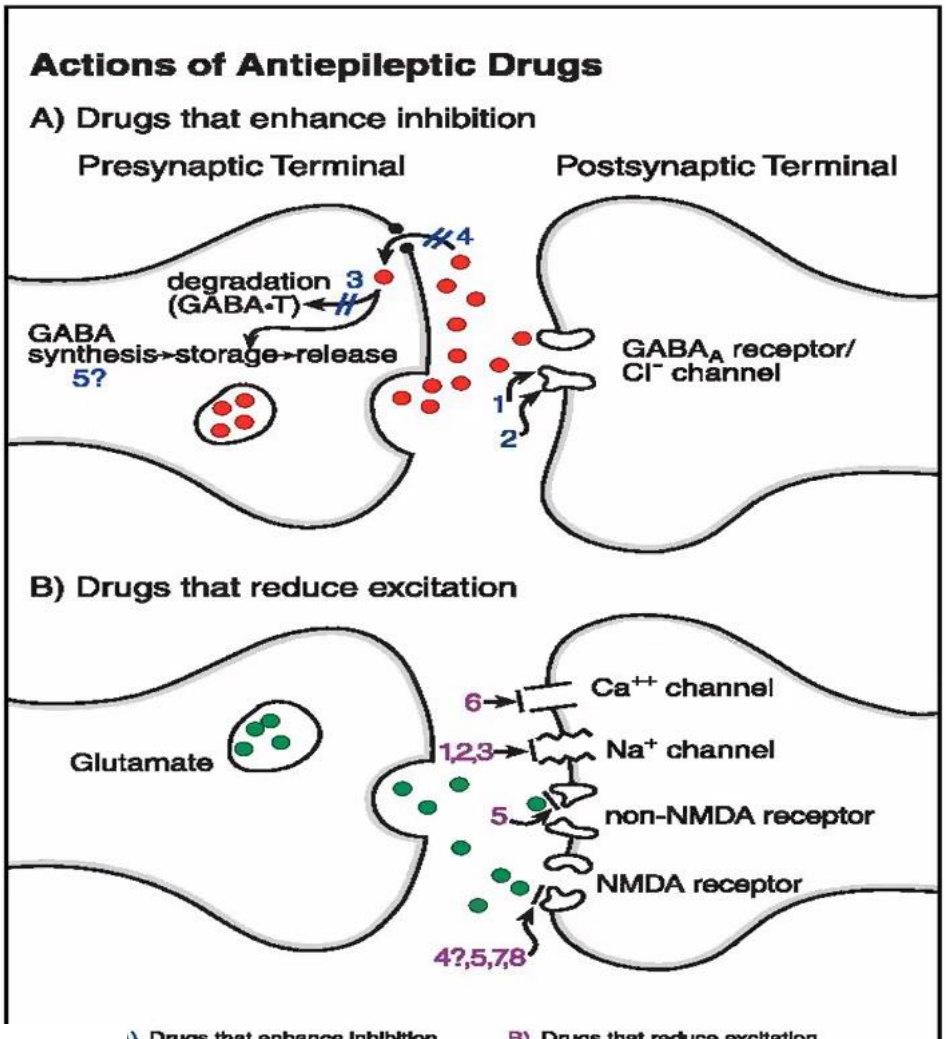
Neuroimaging

- **MRI indicated**
  - Infants (up to 1 yr of age)
  - Loss of developmental milestones/cognitive or motor impairment
  - Focal seizures (do not fit age related epileptic synd.)
  - Focal signs : tumour , vascular lesion, area of sclerosis
  - Abnormal EEG
    - (1) persistent focal epileptiform discharges
    - (2) Focal slowing
- Functional neuroimaging (localization)
  - SPECT
  - PET See the notes on the side →
- Cerebral angiography/ MRA

^Only use when you suspect the child is having a vascular lesion: aneurysm / AVM

SPET: single-photon emission computerized tomogram: they will give contrast and it will show the perfusion, it's expected that neuronal excitability will take too much blood during the seizure. So during the ictal phase there will be hyper-perfusion and between the attack the pt will have hypoperfusion from the site where the seizure is coming.

PET: Positron emission tomogram: very effective functional neuroimaging where we give glucose and those with neuronal excitability will take too much glucose, the same as those with tumors. So areas of high uptake are the abnormal areas.



These are drugs that increase inhibition by increasing GABA →

← These are drugs that reduce excitation by reducing glutamate

Seizure type	First line	Second line
Generalized epilepsies		
Tonic-clonic	Valproate , Carbamazepine	Lamotrigine, Topiramate
Absence	Ethosuximide, Valproate,	Lamotrigine
Myoclonic	Valproate	Lamotrigine
Focal Epilepsies	Carbamazepine, valproate	Topiramate, lamotrigine, levetiracetam, Oxcarbazepine, Gabapentin, Tiagabine, Vigabatrin

Management Principles

- The diagnosis of epilepsy requires two or recurrent unprovoked seizures
- Common practice not to start Rx after 1st single unprovoked seizure
- Explain to family a 50% of further seizure
- The decision of starting AED depends on :

  * Seizure`s type
  * Frequency
  * Social & educational consequence of seizure against unwanted side effects of drugs

Ex: Rolandic seizure is infrequent and if it comes only at night we don't need to start treatment. But if it is coming in the morning we have to treat.

Risk of AED Therapy versus Consequences of Seizure Recurrences*

Risk of AED Therapy	Consequences of Recurrence
Psychosocial	Physical injury
Self-consciousness about medication	Status epilepticus
Labeling as chronically ill	Embarrassment
	Driving restrictions
Adverse Effects	Activity restritions
Idiosyncratic	Labeling as epileptic
Systemic toxicity	Parental overprotectiveness
Behavioral toxicity	
Cognitive effects	
Teratogenicity	
Cost	
Doctor's visits	
Blood levels of AEDs	
Other laboratory tests	
Medication	

Management Principles

- Choose the appropriate drug for seizure, inappropriate AED may worsen the seizure e.g. Carbamazepine & phenytoin, both worsen myoclonic & absence seizures.
- The desired goal is monotherapy at the minimum dosage, but you may need to ↑

  * Until seizures are controlled
  * Until adequate (therapeutic) serum level is obtained
  * Until toxicity prohibit further increase

Management Principles

- In practice polytherapy may be required & one anticipate problems
  - * Drugs compete w.each other for protein binding sites
  - * One drug ↑ the rate/ pathway of catabolism of 2nd drug
  - * Drugs cumulative toxicity
  - * Compliance is more difficult

- All AEDs have potential side effect ,need regular follow-up / explanation for the child & parent

Management Principles

- Follow – up by measurement of drug levels is useful to asses the compliance & for some drugs w. erratic pharmacokinetic e.g phenytion
- Patients w. prolonged seizures are given rescue therapy to have it with them,
  Benzodiazepine e.g . Rectal diazepam or Buccal midazolam
- Duration of therapy : seizure free interval for 2 yrs , for refractory epilepsy ,it may be prolonged or even life long for others

We give treatment for 2 years and it should be seizure free. Some cases if refractory we give longer or if juvenile myoclonic we give for life long.

Management Principles

- Withdrawal of medications

  * Decision to stop AED should be individualized to child & cause of epilepsy

  * In general should be very gradual ie Gradually over 6 or 12 weeks (6/52- 12/52)especially for phenobarbitone / phenytoin

  * An EEG to be done before stopping AED / risk of withdrawal seizures

Comorbidities

- Intellectual disability
- Learning disability
- ADHD
- Behavioural problems
- Anxiety
- Depression
- Sleep problems

Side-effect of AEDs

Focus on the ones in yellow color

- Carbamazepine (Tegretol)
  - . Headache
  - . Dizziness
  - . Leukopenia
  - . Arrhythmia
  - . Rash b/c of allergic reaction
  - . Lethargy
  - . Drowsiness
  - . Diplopia
  - . Hepatotoxicity
  - . Ataxia
  - . Aplastic anaemia
  - . Vertigo
- Benzodiazepine (Clonopin)
  - . Drowsiness
  - . Secretions excessive
  - . Behavioral problems
  - . Tolerance to effect
  - . Ataxia
  - . Hypotonia
- Ethosuximide (Zarontin)
  - . GIT distress
  - . Drowsiness
  - . SLE
  - . Leukopenia
  - . Rash
  - . Dizziness / vertigo
  - . WT gain
  - . Liver damage

Side-effect of AEDs

Focus on the ones in yellow color

- Vigabatrin (Sabril)
  - . Rash
  - . Irritability
  - . W.M abnormality in animal !!!
  - . Behavioral problem
  - . Restriction of visual field
  - . WT gain
- Lamotrigine (lamictal)
  - . Rash (SJS)
  - . GIT upset
  - . Diplopia
  - . Rash
  - . Hepatic toxicity
  - . Headache
- Gabapentin (Neurontion)
  - . Insomnia
  - . Dizziness
  - . Diarrhea
  - . Fatigue
  - . Ataxia

Side-effect of AEDs

- Phenobarbital (Gardinal) We don't like this medication we it for status epilepticus but not beyond that
  - . Drowsiness
  - . Rash
  - . Behavioural problem/ irritability / hyperactivity
  - . Cognitive problem/ learning difficulties
  - . Ataxia

- Phenytoin (Dilantine) We don't give this to females b/c of hum hyperplasia and hirsutism
  - . Drowsiness
  - . Rash
  - . Ataxia
  - . Folate deficiency
  - . Hepatic toxicity
  - . Defective hematopoiesis
  - . Gum hyperplasia
  - . Anaemia
  - . Hirsutism
  - . Pseudolymphoma
  - . Nystagmus / diplopia
  - . SJS

- Valproic acid (Depakene) We don't give it to females b/c of weight gain and alopecia
  - . GIT distress
  - . Alopecia
  - . Ataxia
  - . Pancreatitis
  - . Hepatitis
  - . Drowsiness
  - . Tremors
  - . Thrombocytopenia
  - . WT gain

Side-effect of AEDs

- Topiramate (Topamax)

  . Abnormal taste . Parasthesia
  . Poor appetite / Wt. loss
  . Drowsiness
  . Cognitive impairment
  . Hematuria / renal stones

  In the first few months then we assure the mom that he will gain it again

- Levetiracetam (keppra)

  . Sedation
  . Drowsiness
  . Irritability
  . Cognitive impairment

Intractable seizures

- Ketogenic (fat based / CHO restricted) diet
  Modified Atkins diet

  * Mechanism : create a ketotic state ,breakdown of fat → Intracellular acidosis → alter brain neurochemistry → ↓ seizure frequency
  * Precaution : metabolic disorder / SE* Don't do it for metabolic disorders b/c they will have side effects : hyperuricemia, dehydration, weight loss, hypoglycemia

- Vagal nerve stimulation

  * external programmable of a wire implanted around vagal nerve Usually on the left side
  * 50-70% reduction of seizure frequency 3-6 mon. post implantion & max effect at 18 mon .

  Especially in pts with Lennox Gastaut Syndrome

Intractable seizures

- Surgery

Used for pts with:

* Focal seizures w. localization on

(1) EEG
(2) Functional imaging

* Procedures

(1) Temporal lobectomy/ MTS <- Mesotemporal sclerosis
(2) Hemispherectomy /
Hemispherotomy /LGS <- Lennox Gastaut Syndrome
(3) Focal resection / CD

Pt should be transferred to epilepsy centre for:

* Detailed pre & intraoperative assessment is required to ensure benefit outweigh the risks

Habitation & Psychosocial Support

- Parent & Patient Education

  - What is epilepsy ?
  - Is epilepsy common?
  - What is the duration of treatment ?
  - What is the follow-up ?
  - What is the effect on schooling ?
  - What restriction are necessary & what are safety consideration ?
  - What to do if fit occur ?
  - How it is treated ?
  - What is the future ?
  - Avoid over protection
  - Psychological illness.

Management Outlines

- Safety consideration
  - No swimming alone
  - No bathing alone for young children
  - No bathing in older children with locked bathroom
  - No climbing ladders or heights
  - No bicycle riding on busy roads
  - Use of helmets when going for cycling
  - Use of helmets specially during drop attacks

Acute Management of Seizure Episode

Status Epileptics

Status Epileptics

- Single seizure that last for at least 30 minutes or recurrent seizures lasting for more than 30 minutes without the patient fully regaining consciousness in between.

- Recently, changed to seizure activity longer than 5 minutes or two or more seizures, without a return to consciousness between seizures.

Classification of Status Epileptics

- Generalized Convulsive SE (GCSE)
  - Primary generalized
  - Tonic-Clonic
  - Myoclonic
  - Clonic/Tonic
- Partial
  - Simple partial (includes epilepsia partialis continua)
  - Complex partial SE (CPSE)
- Secondary generalized SE
  - Partial seizures with 2nd generalization
  - Tonic seizures
- Non-Convlsive SE (NCSE) No motor component
  - Absence status (Petit Mal)
  - Atypical absence status
  - Atonic
  - NCSE due to partially treated GCSE
- Neonatal SE

Table 2 - Complications of status epilepticus.[3]

Cardiovascular	***Respiratory***
Tachycardia	Apnea
Bradycardia	Anoxia
Dysrhythmia	Hypoxia
Cardiac arrest	CO_2 narcosis
Conduction disturbance	DIC
Congestive heart failure	Metabolic acidosis
Hypertension	Respiratory acidosis
Hypotension	Altered respiratory pattern
	Pulmonary edema
Renal	Pneumonia
Oliguria	
Uremia	***Endocrine***
Renal tubular acidosis	Endocrine failure
Lower nephron nephrosis	Altered pituitary function
Myoglobinuria	Elevated prolactin
	Elevated vasopressin
Other	Hyperglycemia
Fever	Hypoglycemia
Increased CPK	Increased plasma cortisol
Autonomic dysfunction	

CPK - creatine phosphokinase, DIC - disseminated intravascular coagulation, CO_2 - carbon dioxide

- **Complication**

  Neuronal death can occur under certain circumstances after as little as 30 to 6o min of continuous seizure activity.

  So we have to control the seizure before it reaches 30 or 60 minutes

Etiology

- The commonest cause for acute seizure / SE varies with the age of the child

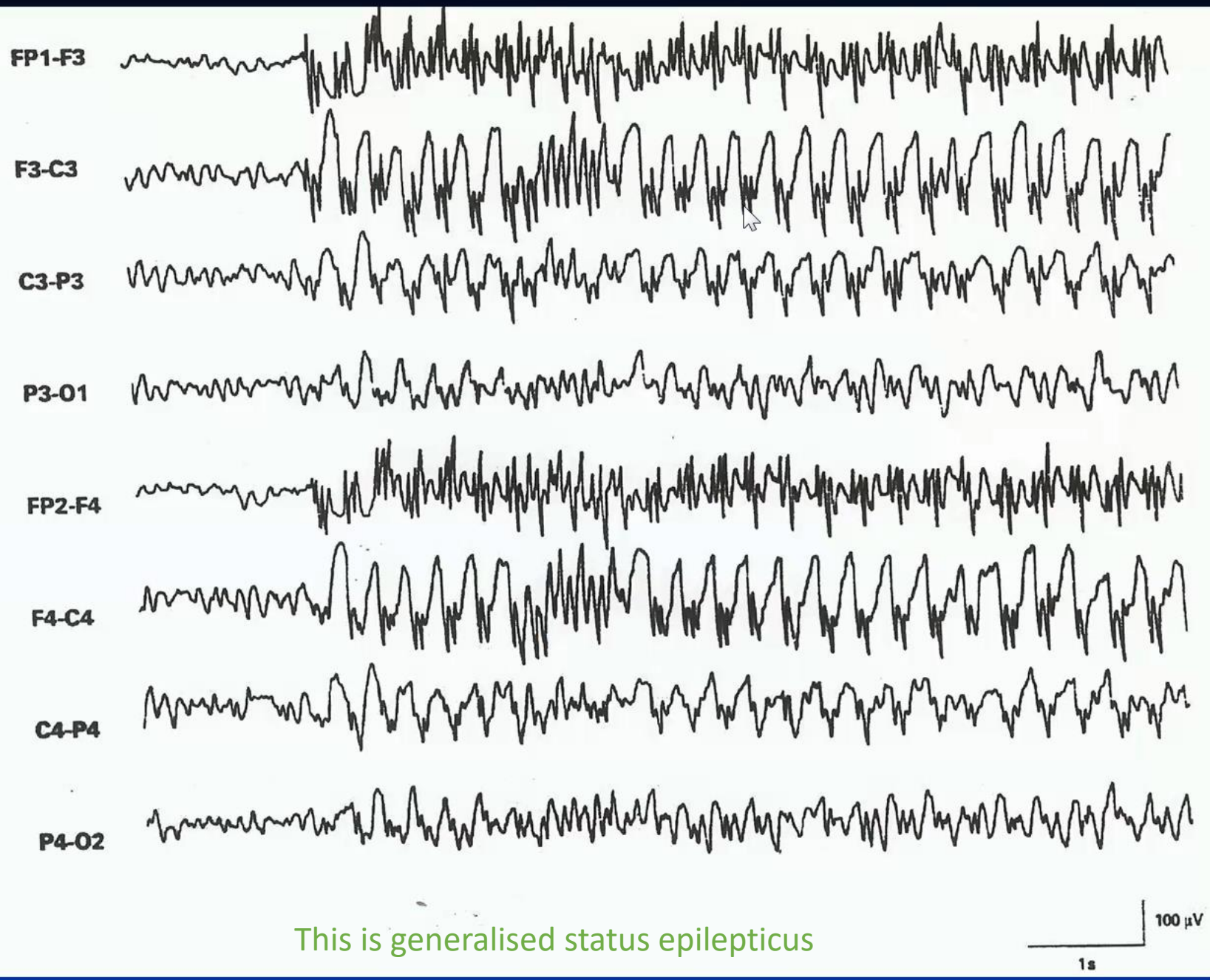
A- HIE , IEM (newborn).

HIE: Hypoxic ischemic encephalopathy
IEM: Inborn error of metabolism

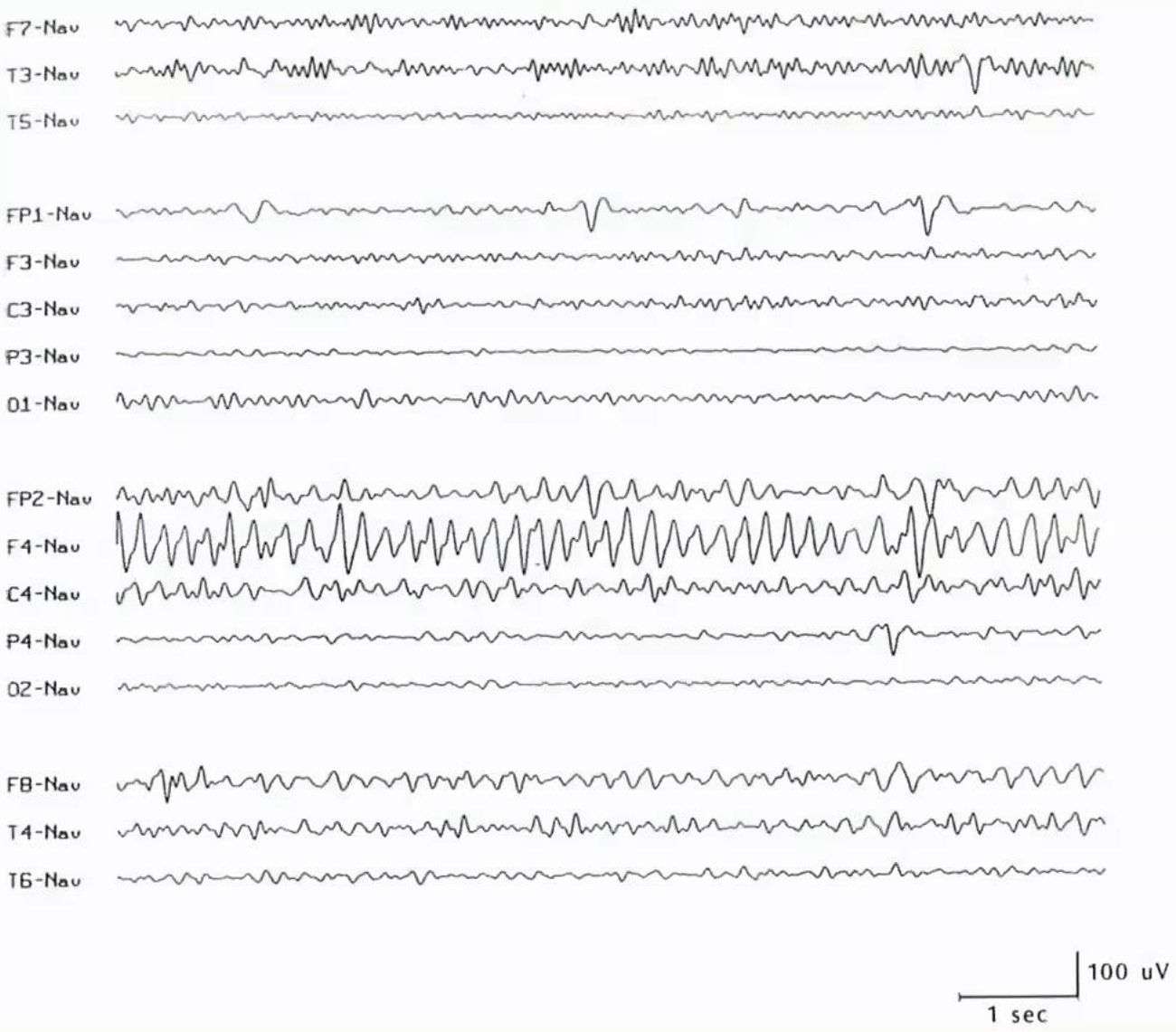
B- Febrile SE (common < 5 yrs).

SE: status epilepticus

C- Trauma & Infection (older children).



This is generalised status epilepticus


F7-Nav
T3-Nav
T5-Nav
FP1-Nav
F3-Nav
C3-Nav
P3-Nav
O1-Nav
FP2-Nav
F4-Nav
C4-Nav
P4-Nav
O2-Nav
F8-Nav
T4-Nav
T6-Nav
100 uV
1 sec

Treatment Options

- A- Out- of-hospital or No IV Access Available.

  . Buccal midazolam (0.2mg/kg)
  . Intranasal midazolam (0.2mg/kg)
  . Rectal diazepam 0.5 mg/kg (2-5y),
  0.3 mg/kg (6-11y), 0.2 mg/kg (>12 y).

This is how we give buccal midazolam

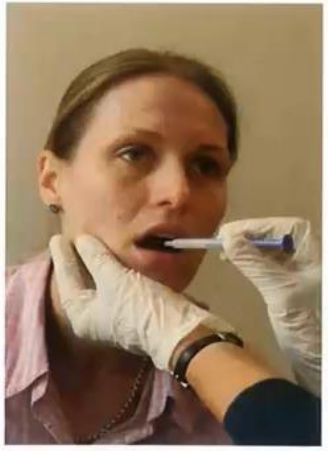
To give the midazolam

1. Support the person's head and chin

Be careful that you do not press on their throat.

Supporting their head helps to stop the liquid leaking out of their mouth when you give it.

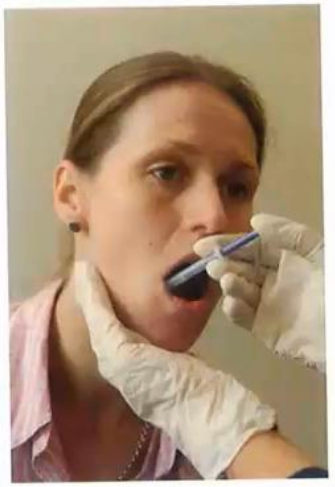
2. Insert the dispenser



Gently slide the dispenser horizontally into the person's mouth, between their cheek and lower gum on one side.

Do this carefully to avoid damaging their gums and teeth.

3. Put the dispenser into the buccal cavity



Tilting the plunger-end of the dispenser upwards, (so that the tip points downwards) put it into the buccal cavity.

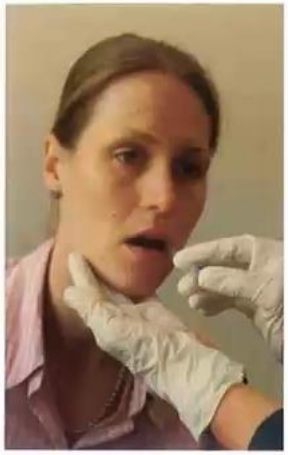
Make sure that you don't place the dispenser below their tongue because they may clamp their teeth shut and break it.

4. Give half of the dose

Very slowly push on the dispenser plunger to give half of the midazolam into the buccal cavity. This can take 30 seconds to a minute to do.

Doing this slowly means that the drug is absorbed into the body more effectively. If it is given too quickly the person may swallow or choke on it (although if a small amount is swallowed it will not cause any harm).

5. Follow instructions 2 to 5 on the other side of the mouth



To help the midazolam get absorbed quickly, if possible put half of the dose into one side of the mouth and half into the other side.

However, if this is not possible (for example, if you can't reach the other side of their mouth because they are lying on the floor), put the whole dose into one side of their mouth.

If they are lying on their side, put it into the side of the mouth that is nearest the ground.

6. Support the person

Gently close their mouth and continue to gently support their chin for a minute or two. This helps to make sure that the liquid does not leak out of their mouth after you have given it.

7. Wipe away any spit and put them in the recovery position

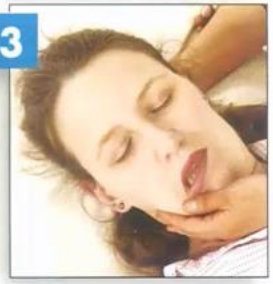
Use a tissue to wipe away any saliva (spit) from the person's face and put them into the recovery position if appropriate.

This is how we give intranasal

Epistatus is generally administered after some minutes of convulsive activity or for repeated seizures or as directed by your doctor or epilepsy nurse.

1 Open the bottle by pressing down on the child resistant cap and turning it anti-clockwise. Insert the syringe firmly into the bung on the top of the bottle, with the plunger pressed to the bottom of the syringe

2 Supporting the bottle, tip the bottle upside down and slowly pull the plunger on the syringe, until you have withdrawn the prescribed amount



3 Support the head. Locate the buccal area by holding the chin and gently applying downward pressure on the lower lip as shown in this picture

4 Insert the syringe into the buccal cavity. Do not place the syringe between the patient's teeth as the syringe may be bitten

5 Slowly administer half the liquid in the syringe by pushing the plunger downwards. Then repeat on the other side

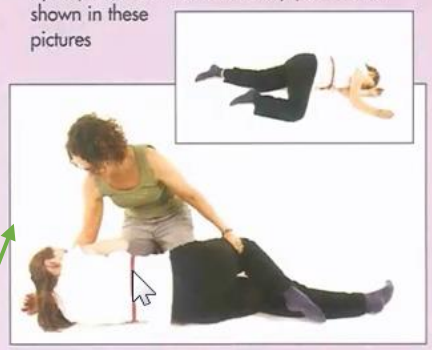
6 Epistatus can also be given intra-nasally with half of each dose being administered into each nostril

7 Screw the cap back on the bottle immediately after use

Convulsive Seizures

If you see someone having a convulsive seizure you should:

- Keep calm
- Stay with the person
- Move things like chairs and other objects away from them so that they don't hurt themselves
- If you can, put something soft such as a cushion under their head
- Reassure the person throughout the seizure and when it is over
- **DO NOT** try to physically stop the seizure or try to restrain the person
- **DO NOT** put anything in the mouth
- Stay with the person until the seizure is over and try to place them in the recovery position as shown in these pictures



There is usually no need to call an ambulance for medical assistance unless:

- The seizure lasts longer than usual for the individual
- The person has repeated seizures
- The person has sustained an injury
- The person has had a seizure in the sea or a swimming pool

It's important to make sure the child is in the right position

Acute Management of the SZ.

- What to do first : the ABC :
  . Stabilize & Maintain the airway.
  .Establish breathing (ie, ventilation).
  .Maintain the circulation.
- Monitor vital signs:
  pulse oximetry, respiratory rate, blood pressure, temperature.
- Maintain Oxygenation.
- Position head to prevent/relieve airway obstruction.
- Early intubation to protect airway ,provide adequate oxygenation &ventilation.
- Establish vascular access
- Establish the presence of SE
- Evaluate

The above measures are essential to provide oxygen to the brain & effective cerebral perfusion pressure (CPP).

Acute Management of the SZ.

- Establish vascular accesses
  IV , Intraosseous , CVL

- Blood : Glucose
  Ca , Mg
  Na , BNU
  ABG
  LFT
  Drug level
  CBC/ Blood C/S If the child has fever

- Benzodiazepine to abort
- Correct : Hypoglycaemia / electrolyte imbalance
- Rx hyperthermia / passive cool + antipyretics
- Correct : metabolic acidosis (If sever)
- Pyridoxine (refractory in < 3 yrs age)

Acute Management of the SZ.

- May need other abortive medications / refractory
- Long – acting AED to prevent recurrence
- ????? What aetiological diagnosis & manage accordingly

Drug Rx For SE

- 1st - (Seizure 0-5 min) Stabilization phase
- 2nd - (Seizure 5-30min) Early SE

  Benzodiazepines

  * Lorazepam , Diazepam , Midazolam

  We usually give 2 doses of **lorazepam** b/c it lasts for 1-2 hours
- 3rd - (Seizure 30-60 min) Established SE

  Phenytoin, Fosphenytoin , Phenobarbital

  * Levetiracetam , Valproic acid
- 4th - (Seizure >60 min) Refractory SE

  Midazolam , Thiopental , Pentobarbital

  In refractory pt should be admitted to ICU, intubated, with continuous EEG monitoring.

What parenteral medications can be given if no IV access is available?

Drugs to use when you don't have IV

	Access Dose	Route	Side Effect
Diazepam	0.5 – 1 mg/kg make 20 mg	Rectal	Sedation respiratory depression, hypotension
	0.2-0.5 mg/kg	IO	
Lorazepam	0.05 – 0.1 mg Max 4 mg	Rectal IO	Similar but less in comparison to Dizepam
Midazolam	0.05 – 0.1 mg/kg	?IN IM	
Phenobarbitone	15-20 mg/kg	IM IO	Hypotension, respiratory depression, bradycardia
Fosphenytoin	15-20 mg/kg of PE	IM	Lightheadedness, nystagmus, dizziness
Phenytoin	15-20 mg/kg	IO	Hypotension, respiratory depression, cardiac arrhythmia.
Paraldehyde	0.3-0.5 ml/kg 1:1 with mineral oil	Rectal	Respiratory depression, pulmonary edema, RF, hepatitis, metabolic acidosis, tissue necrosis, nerve damage, HF
Valoprate	40-60 mg/kg diluted 1:1 with tap water	Rectal	Liver failure, ataxia, tremor, GIT irritation, pancreatitis.
	20-60 mg/kg	PO (NG)	

- *Note:* As per routes noted above:
  - Diazepam onset 5 min, peak concentration within 15 min
  - Phenobarbitone onset 30 min, peak concentration within 1-2 hr.
  - Fosphenytoin onset 30 min.

Don't forge that these medication cause hypotension and respiratory depression so put the pt of cardiac monitor

Prognosis

- Outcome most dependent on :

  * Age.
  * Etiology.
  * Duration of SE.

- Worse outcome :

  * Young age
  * Female sex
  * Long duration SE / continuous
  * Symptomatic CSE

CSE Neurologic Sequelae

- Sequelae

  * 2nd epilepsy / hemicovulsion-hemiplegia syndrome
  * cognitive deterioration.
  * behavioral problems.
  * focal neurologic deficits.

- The neurologic syndrome

  * diplegia.
  * extrapyramidal syndromes.
  * cerebellar syndromes.
  * decorticate rigidity.

- Permanent sequelae : 5-10 %

Febrile Convulsion

- Definition :

Is a seizure associated with fever in the absence of other cause & not due to intracranial infection (meningitis or encephalitis)

It is just a fever follow by a seizure

Febrile Convulsion

- Occur in 3% of children .
- Have a genetic predisposition ,
  10% risk (1st degree relative with febrile seizures).
- Age : 6 months - 6 years.
- Occurring with rapid rise in temperature
  (> 38.5c)

Febrile Convulsion

- Simple (Typical)

  . Generalized mostly TC or T.
  . Last <15 min.
  . No recurrence within the acute event.
  . No postictal abnormality

- Complex (Atypical)

  . Focal.
  . Prolonged > 15 min. Or 20 minutes
  . Multiple within 24 hr. (at least two).

Febrile Convulsion

- Evaluation :

  * Examination : Look for the site of the infection

    * Cause of fever .
      ? Viral
      ? Bacterial / meningitis vs encephalitis

    * Signs of meningeal irritation.
      Neck stiffness , photophobia , etc.
      Absent < 18 months. This age group may not have any signs so you may due lumbar puncture to make sure if they have meningitis.

Febrile Convulsion

- Evaluation :

  * Infection screen

    * CBC (dif .) , ESR
    * Blood C/S
    * Chest x-ray
    * Throat swab
    * Urine C/S
    * Lumbar puncture (CSF)
      (cell count & diff., gram stain , latex agglutination , biochemistry ,eg. Glucose ,protein)

Febrile Convulsion

- Lumbar puncture :

  * Is contraindicated

    * Child with altered level of consciousness
    * Unconsciousness (Glasgow Coma Scale < 8)
    * Continuous seizure ie SE
    * Focal neurological signs / ↑ ICP

  * LP may be deferred until neuroimaging is done
  * Antibiotic should be started empirically.

Febrile Convulsion

- EEG

  * Is not indicated

    * Does not serve as a guide for treatment

    * Does not predict seizure recurrence

Febrile Convulsion

- Management :

  * reassurance.

  * Advice sheet : We explain these things to the mom

    * Temperature control : tepid sponges, antipyretics (paractamol , ibuprofen)

    * 1st aid of seizure management

    If (seizure > 3-5 min)/ cyanosis at onset.

    Give Rectal diazepam or
    Buccal midazolam

Febrile Convulsion

- Management:

  * Prophylaxis

    * Rectal dizepam PRN - convulsion during febrile

    * Oral AED (phenobarbition, valporic acid)
      not recommended

      because

      Do not ↓ risk of recurrence
      Do not ↓ risk of epilepsy
      Associated with side effect
      (hyperactivity, cognitive impairment ,
      hepatic dysfunction especially < 2 yrs of age)

Febrile Convulsion

- **Facts :** We explain these to the mom

Simple febrile seizure :

* Do not cause brain damage.
* Normal intellectual performance.
* Risk of epilepsy : 1-2 %
  (as for all children).

Complex febrile seizure :

* Risk of epilepsy : 4-12%.

Febrile Convulsion

- Recurrence :

34%
Most recurrence ~ 75% occur within 1 yr.

* Younger age.
* Short duration of illness before the seizure.
* Low temperature at the time of seizure.
* Positive family history of FS

Febrile Convulsion

- Risk factors for development of epilepsy following FS

. Abnormal development / Abnormal neurological examination before first seizure
. Family history of afebrile seizures. Or epilepsy
. Prolonged febrile convulsion.
. Complex type

NB: No risk found with multiple episodes of FS within the acute illness.

Paroxysmal disorders (Funny turns)

- These are conditions that can mimic childhood epilepsy.
- Epilepsy is a clinical diagnosis based on:

  * Eye witnesses
  * Child´s own story
  * Videos of the events

- The key to diagnosis

  * Detailed history
  * Clinical examination
  * ??? EEG

Paroxysmal disorders (Funny turns)

- Decrease cerebral blood flow (CBF)
- Sleep disorders
- Movement disorders
- Psychological disorders

cyanotic

Breath-holding attacks

- Occur in 3% children (toddlers & up to 4 yrs)
- Precipitated by anger/ upset
- The child cries ,hold his breath & goes blue, then limp , rapid recovery
- Attacks resolve spontaneously
- Management :

  * Drug therapy is unhelpful
  * Iron therapy/ Pirectam containing drugs may ↓ the frequency of episodes
  * Behaviour modification/ Avoidance of confrontation

The best -> thing to do for them

Characteristic	Breath-holding spells	Seizure disorder
Age at onset	6-18 months	Rare in infant with normal neurologic exam Onset usually 3-18 months
Precipitated by event	Usually	Rarely
Crying at onset of episode	Always in cyanotic spell, usually in pallid spell	Rarely
Cyanosis	Nearly always at oneset of cyanotic spell	Rare at onset episode; may occur later
Heart rate	Asystole or bradycardia	Rapid
Seizure activity visible	in 20% -25% of episodes	Almost always
Incontinence	Never	Sometimes
Postictal confusion	Occasionally, of short duration	Common; sleepiness
Family history of breath-holding spells	In 30% of children	In 11% of children

Reflex anoxic seizures

- Occur in 1 % of children (infants or toddlers)
- Many with a history of faints in a 1st degree relatives
- Precipitated by:

  * Pain/discomfort e.g.minor head trauma
  * Cold food e.g.ice-cream or cold drinks
  * Fright
  * Fever
    (some children with febrile convulsion
    may have similar episodes)

- The child stop breathing, goes very pale ,fall to the floor , brief GTC seizure 2nd to hypoxia , rapid recovery

Reflex anoxic seizures

- The episodes are due to cardiac asystole from ↓ threshold to vagal inhibitory reflex
- Drug therapy is unhelpful
- Ocular compression under controlled conditions leads :

  * Asystole
  * EEG : Paroxysmal slow-wave discharge
  * ECG : should be done It is mandatory

Differential Diagnosis-Pallid spells

- Seizures with autonomic symptoms
- Vasovagal syncope
- Cardiac arrhythmia
- Prolonged QT syndrome
- Hypothermia
- Familial dysautonomia

Syncope

- The most common seizures mimicker in school age children
- Faint occur 2nd to :
  * Hot & stuffy environment
  * Standing for long periods
  * Fear
- Precipitant → vasovagal response → venous pooling → decrease CBF , preceded by lightheadness, blurred vision , ringing in the ears , pallor , diaphoresis , abdominal discomfort (CBF: cerebral blood flow)
- Non-epileptic tonic-clonic seizure may occur 2nd to anoxia
- Differential Dx : cardiac dysrhythmias
- Management : avoid the precipitatory events

Cardiac arrhythmia

- Prolonged QT interval / Sick sinus syndromes

  Produce

  * Syncope
    → sudden LOC with atonic / tonic posture
    → prolonged confusion
  * Triggered by fright , surprise , immersion in water
  * Exercise-induced seizures
  * Relatives (epileptic or sudden death)
  * Extensive cardiac investigation is mandatory

Characteristic: mom tells you always when he is playing football he will have a seizure

Ex: his uncle died at age of 15, or his uncle has epilepsy and impaired hearing and we found he has cardiac problems which is prolonged QT syndrome.

Migraine Can mimic seizures

- Paroxysmal headache
- Children may have special episodes without headache
- Unsteadiness / light-headedness
- Visual disturbance which are like the ones that have occipital epilepsy
- GIT disturbance Similar to those with temporal epilepsy
- Hemiplegic migraine ; Aura of focal weakness +/- speech disturbance, visual symptoms, and paresthesia onsets before typical migraine-like headache
- Often family history is positive For migraines

Sleep Disorders

- Narcolepsy & Cataplexy
- Parasomina ; Night terrors, sleepwalking (somnambulism), confusional arousals
- Obstructive sleep apnoea
- Bruxism , noct.enuresis , noct.myoclonus

Narcolepsy: excessive daytime sleepiness.
Cataplexy: immediately will lose tone b/c of strong emotion + sleep paralysis and hypnogogic hallucination.

Occurs in the first few hours of sleep during NREM, lasts for 3 – 5 minutes and may recur. Most imp ddx: nocturnal frontal epilepsy

Obstructive sleep apnea:
The child might develop seizure 2ry to obstruction of airway (adenoid or tonsil) and develop seizure b/c of anoxia. Rx here is not AED but adenectomy (treat underlying cause)

Bruxism:
الناس اللي تعض على اسنانها ويطلع منهم صرير
This may happen to anxious personality types during sleep and its fine,
But it may also happen in those with seizures.

Nocturnal enuresis:
This can happen because of seizures or can be in normal boys.

Psychological Disorders

- Panic attacks
- Day dreaming
- Psychogenic nonepileptic spells
- Conversion reactions
- Fictitious epilepsy
- Hyperventilation syndrome

Panic attack:
The child will have feeling of doom then develop tachycardia, diaphoresis, abdominal pain, and it is provoked by something. It aborts by itself, this is not seizure and no loss of consciousness.

Day dreaming:
A person who day dreams is staring but it is not a seizure it can be interrupted by simple simulation.

Psychogenic nonepileptic spells:
The child pretending to have a seizure and we can tell because the post ictal phase is very short and they will recover immediately so this does not fit a seizure.

Conversion reactions:
The child will again pretend to have a seizure if he wants to get something.

Hyperventilation syndrome:
An adolescent gets upset at home or at school and he develops this attack after hyperventilation -> b/c hyperventilation will cause respiratory alkalosis which leads to electrolyte disturbance -> reduced ionized calcium resulting into a tetany like picture (pt is aware) Rx: breath into a closed bag / NO AED drugs

Fictitious epilepsy:
Ex: there was a pt admitted for seizures and he was on 3 AED but they could not control it and then they found out the mother was giving her child insulin which induced hypoglycemia and seizures. The mother did not like her husband and did not want to go home.

At the end of the lecture, the doctor showed us some videos of **infantile spasm, absence seizure, and complex partial seizure.**

She said to focus on them (EEG finding and management) because they are important (احفظوها صم)