

# Difficult Issue in Managing Epilepsy : Childhood

Apasri Lusawat M.D.

Pediatric Neurology

Prasat Neurological Institute



สถาบันประสาทวิทยา  
PRASAT NEUROLOGICAL INSTITUTE



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# What is this seizure type?

- Atonic seizure
- Myoclonic seizure
- Complex partial seizure
- Generalized tonic seizure

# **Atonic seizure** also called **drop seizures, akinetic seizures** or drop attacks

- “sudden diminution of muscle tone with which may be fragmentary, leading to head drop with slackening of the jaw, dropping of one limb or a loss of muscle tone leading to a slumping to the ground” (ILAE)
- The seizures are brief - usually < 15 seconds.
- Begin in childhood and may persist into adulthood.
- Seizure itself causes no damage, but the loss of muscle control can result in indirect damage from falling, so protection such as a helmet may be used

# เด็กหญิงอายุ 6 ปี

- อายุ 1 ปีมารดาให้ประวัติว่า ชักเกร็งทั้งตัวตาเหลือก เกร็งกระตุก 2-3 ชุดต่อวัน ชุดละ 6-7 ครั้ง แต่ละชุดนาน 1-2 นาที เรียกไม่รู้ตัว มักเป็นตอนตื่นนอนใหม่ๆหลังชักตัวอ่อน
- ได้รับการรักษาด้วยยากันชักหลายชนิด
  - Valproate , Clonazepam อายุ 1-4 ปี
  - Topiramate, Lamotrigine อายุ 4 ปีจนถึงปัจจุบัน
- ยังมีอาการชักทุกวันๆละ 2-3 ชุดๆละ 6-7 ครั้ง ไม่ดีขึ้นจึงมาสถาบันประสาทวิทยา



# เด็กหญิงอายุ 6 ปี

- พัฒนาการ ช้า ขณะนี้ยังไม่พูด ส่งเสียงอ้อแอ้ สบตายิ้ม เดินได้
- ประวัติครอบครัว เป็นบุตร 1/1 คลอดปกติแข็งแรงดี ไม่มีประวัติโรค  
ลมชักในครอบครัว
- PE: HC 51cm, Mental Retard , Mute.
  - Lateral squint Rt eye
  - CN intact
  - motor: normal
  - DTR 2+ all , no ataxia
  - General exam: no skin lesion, other WNL



# What is your first impression ?

- Multiple seizure types with mental retardation : atonic, absence, generalized tonic ->
  - Lennox Gastaut syndrome
- Atonic seizure -> refractory seizure
  - Corpus callosotomy may be helpful.

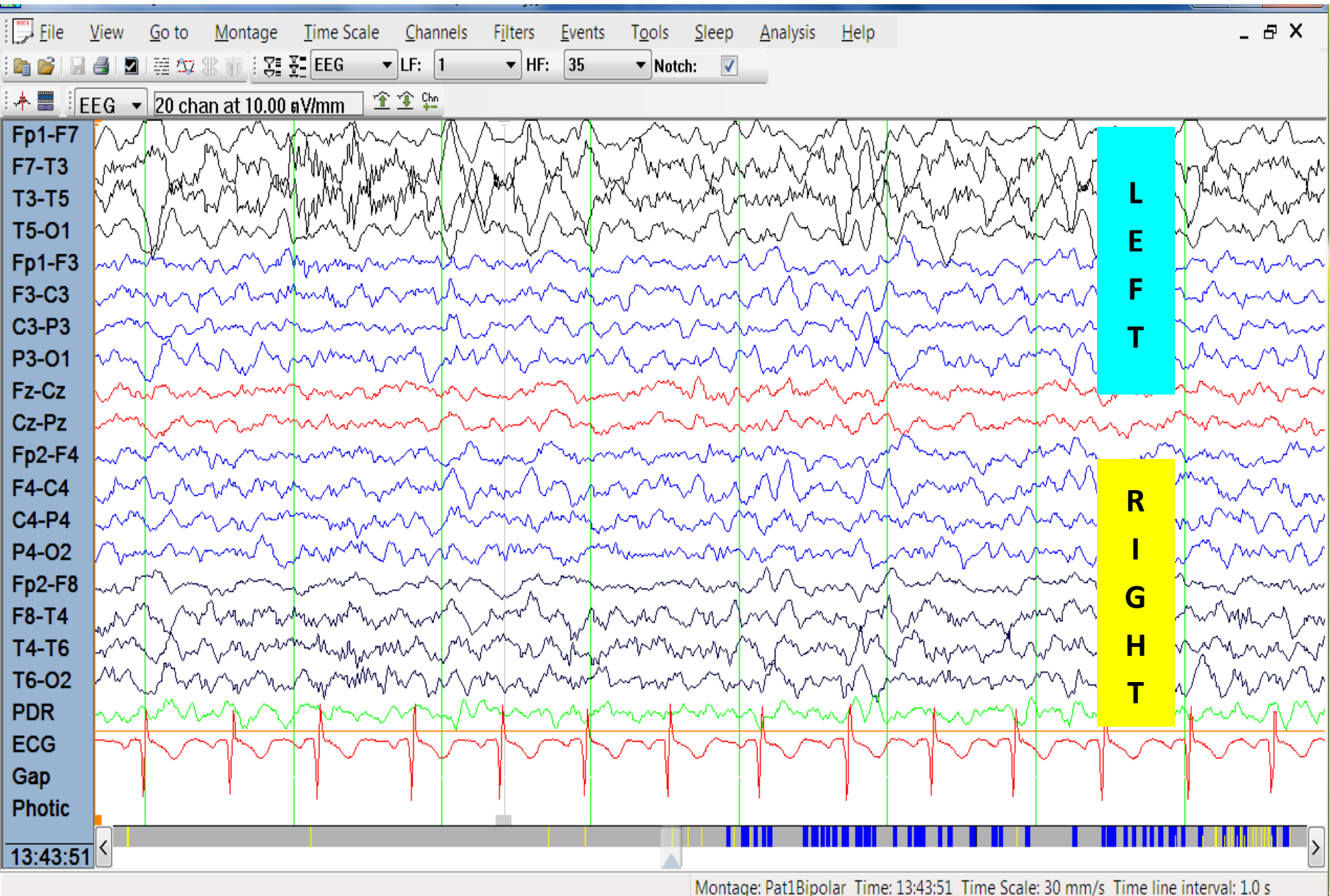
Yes

No



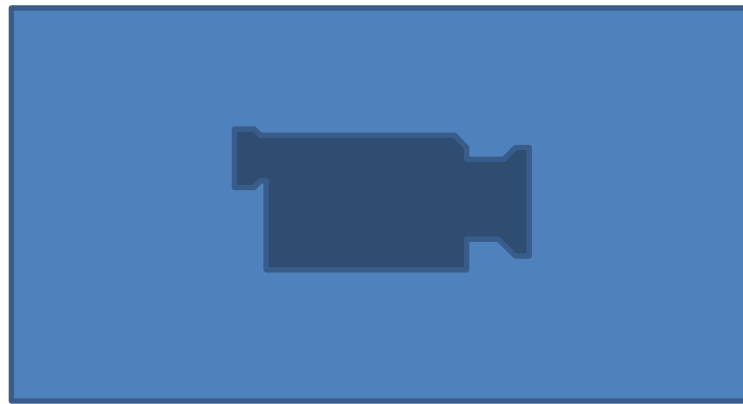
**Not yet making decision !!!**

# Interictal EEG





# VDO-EEG ictal



# Question: what is this EEG pattern?

- A. Generalized fast spike-wave
- B. Generalized slow spike-wave
- C. Focal sharp-slow activity
- D. Focal dipolar spike-wave

A

B

C

D

# Most likely diagnosis in this patient?

- A. Lennox-Gastaut syndrome
- B. Myoclonic-astatic syndrome (Doose)
- C. Severe myoclonic epilepsy of infancy (Dravet)
- D. Focal with secondarily generalized epilepsy



# Epilepsy syndrome

	Seizure type	EEG
<b>LGS</b>	Generalized tonic, Atypical absence Atonic seizure	Generalized Slow spike-wave < 3 Hz
<b>Doose</b>	Myoclonic Astatic / atonic	Generalized spike- or polyspike-wave 2-3 Hz, without focal spike
<b>Dravet</b>	GTC / focal (early) Myoclonic (later)	Generalized or multifocal asynchronous fast spikes or PSW, esp. the F-C, C-T and vertex

# Most likely diagnosis in this patient?

- ~~A. Lennox-Gastaut syndrome~~
- ~~B. Myoclonic-astatic syndrome (Doose)~~
- ~~C. Severe myoclonic epilepsy of infancy (Dravet)~~
- ~~D. Focal with secondarily generalized epilepsy~~

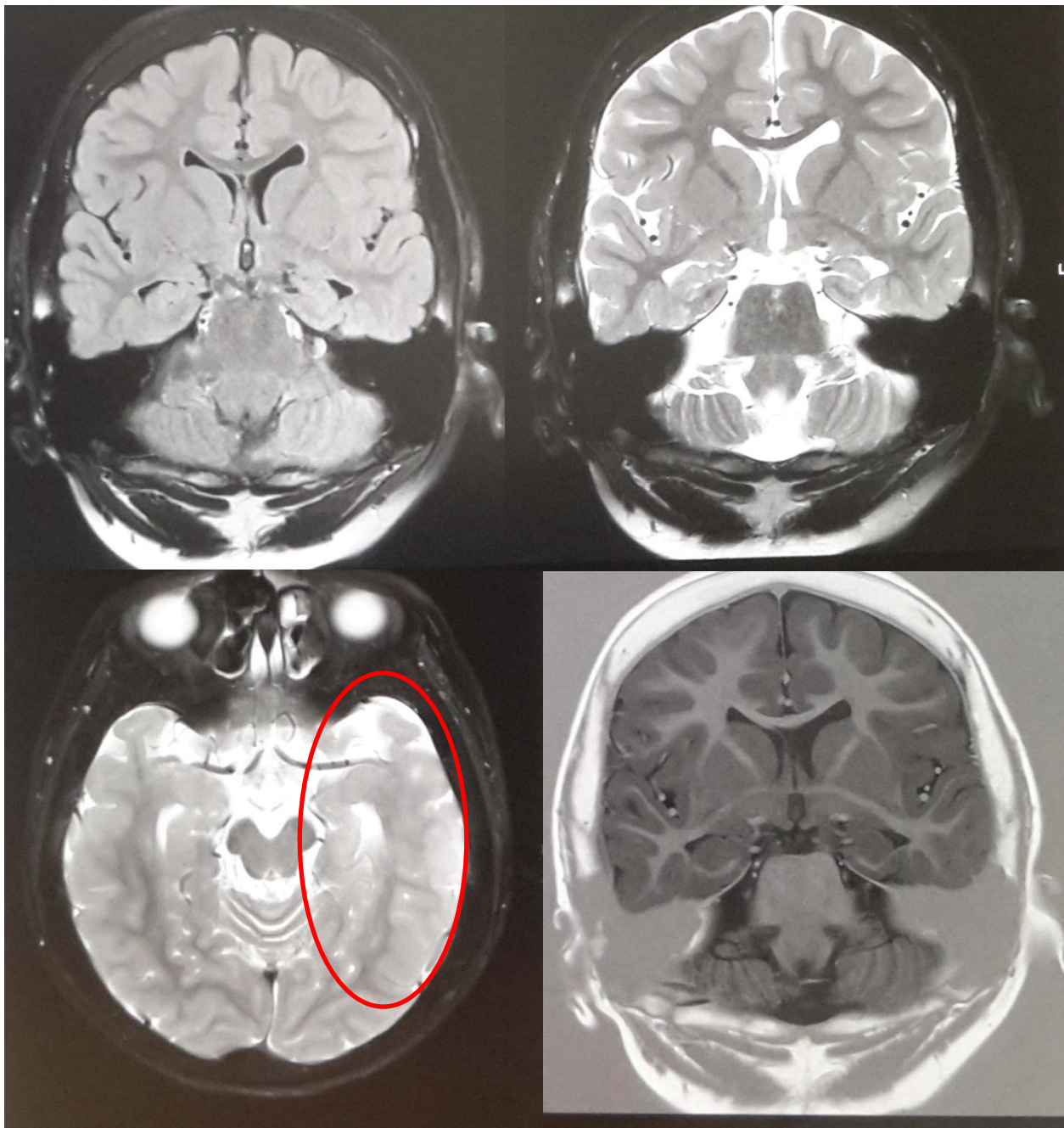
- **Localization-related symptomatic epilepsy**

# Question: what to do next?

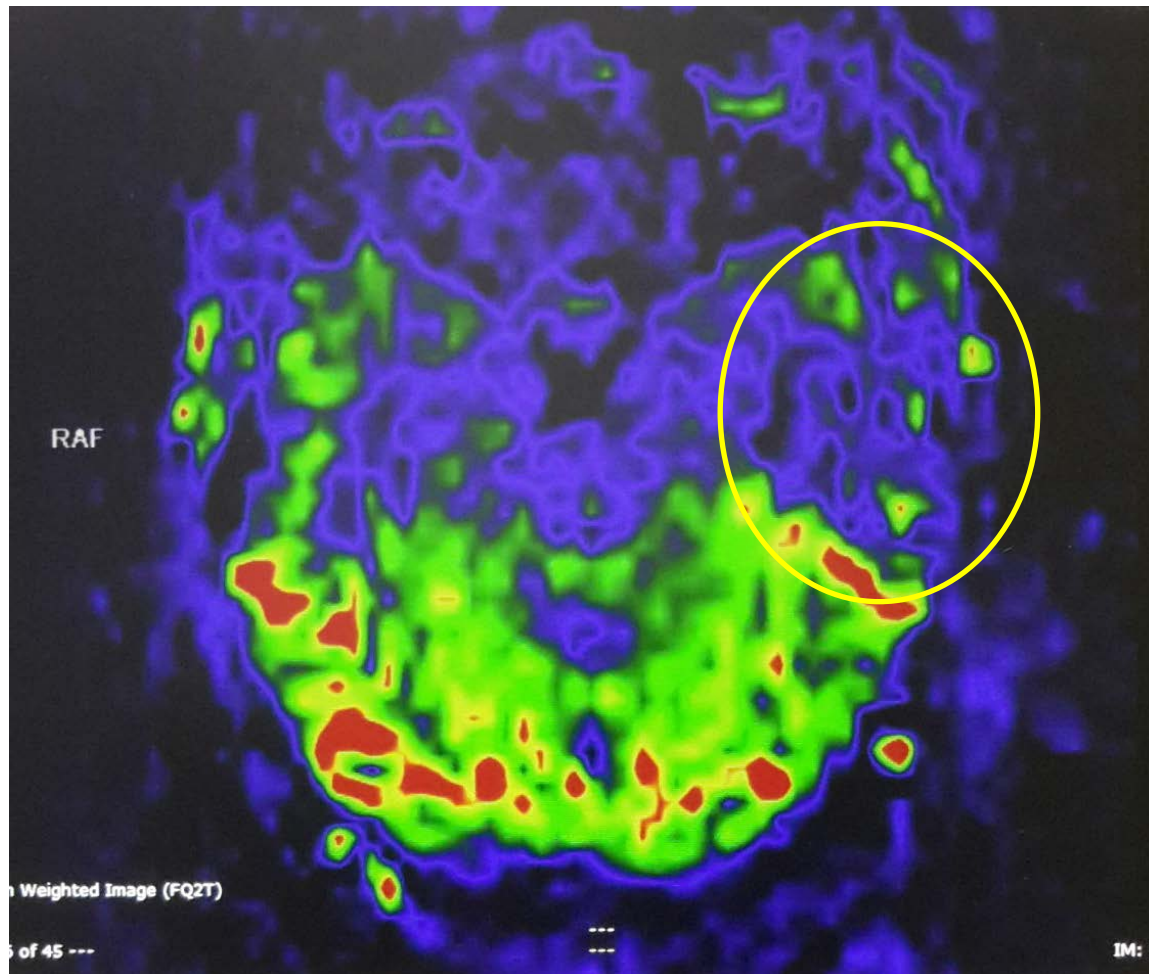
- A. Continue medical treatment
- B. Add ketogenic diet
- C. Work up for epilepsy surgery

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# MRI

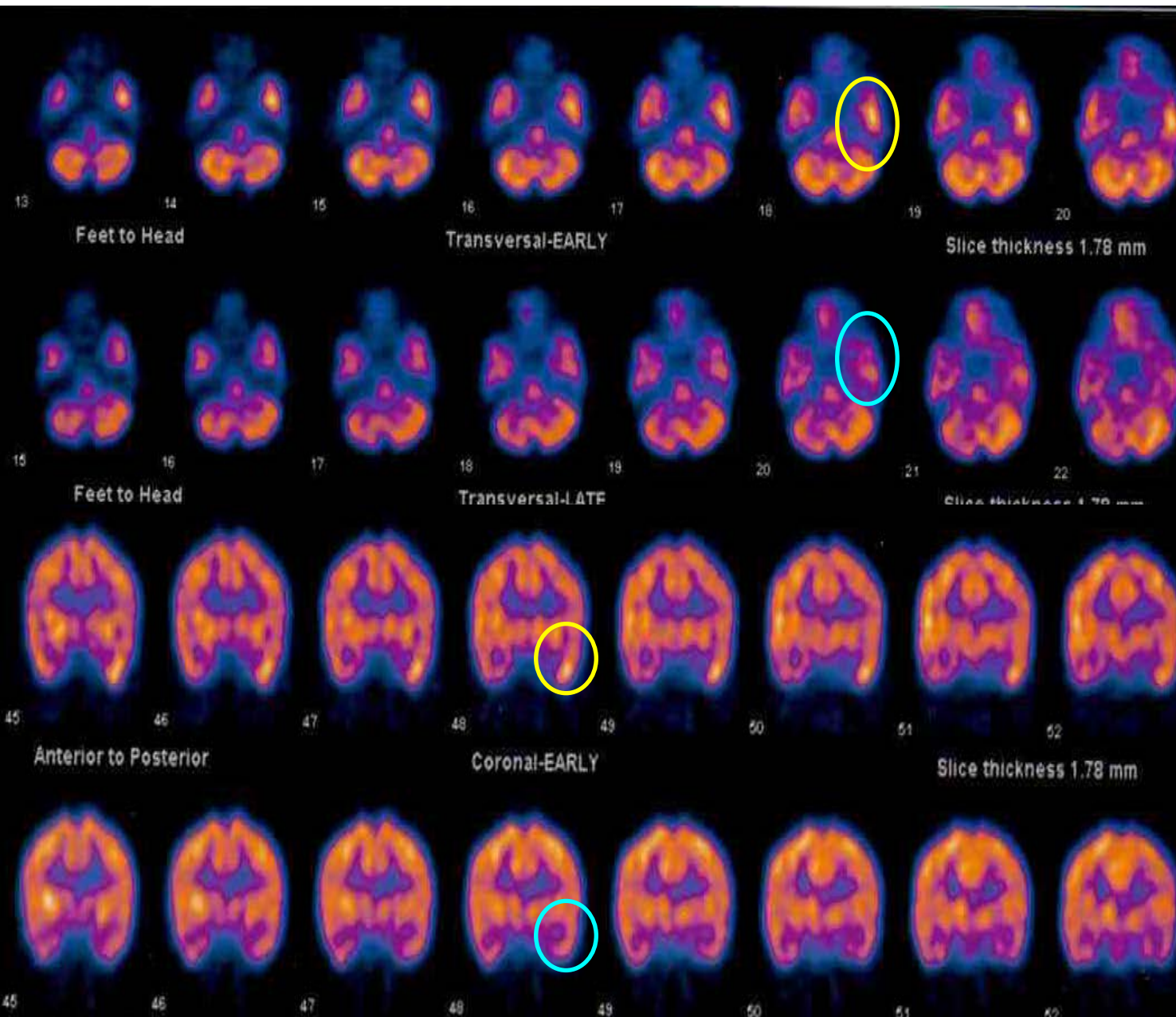


# ASL (Arterial spin labeling : perfusion)





# SPECT



Ictal

Inter-ictal

Ictal

Inter-ictal



# Concordance

- Semiology : clinical CPS + atonic
- Ictal EEG : onset Lt temporal
- MRI : focal abnormality at Lt > Rt temporal
- ASL : focal low perfusion at left temporal
- Ictal SPECT : hyperperfusion at left temporal

**What is your decision now ?**



# What to do next?

- A. Epilepsy Sx : resection left temporal
- B. Epilepsy Sx : total corpus callosotomy
- C. Further investigation : invasive EEG

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# Treatment in this girl

- Left temporal lobectomy extending to posterior part under ECoG.

## Pathology

**Focal cortical dysplasia**



# Outcome after surgery

- Seizure free for 10 Months
- Continue AED :
- Development : improved
  - Can understand but cannot speak
  - Response to one step command



# Atonic seizure

- Traditionally described in **generalized epilepsies**

## Rx

AEDs : VPA, LTG, TPX, LEV, CLZ, CLB

Surgery: Corpus callosotomy, VNS

Ketogenic diet

## Prognosis

Resistant to drug therapy : LGS(100%) , MAE (18%)

Callosotomy : improve seizure in 78%, seizure free 38%

(Tanriverdi T, et al, 2009)

**VNS** : improve sz in 27-64% (Rosenfield WE, 2009)

- Ictal atonia is increasingly recognized as a phenomenon of **focal seizures**.

# Nomenclature

- **Focal atonic seizures** are partial seizures where the ictal manifestation consists of paresis or paralysis of one or more parts of the body
- **Epileptic drop attacks** imply predominant atonia of trunk muscles
- **“Drop attack”** has been proposed for both tonic and atonic seizures used in many studies looking at therapeutic outcomes in this type of epilepsy.

Reutens DC,1993, Maehara T,2001, Oguni H,2001

# Ictal atonia with focal epilepsy : case reports

Author	Age (Y) Sex	Ictal features	Sz duration	Etio-logy	Imaging	Ictal scalp EEG
Satow et al., 2002	12; M	LOC - followed by:(1) Neck atonia – atonia in axial and limb muscles ( <b>R&gt;L</b> )(2) Neck atonia – atonia in axial and limb muscles (R>L) – rt hand clonic - GTC	3–5 s atonia	FCD	MRI: <b>rt</b> parietal FCD and rt hippocampal atrophy PET: bilateral parietal hypometabolism; rt mesial temporal hypometabolism	Ictal spikes <b>lt</b> central – gradually involving <b>rt</b> central region
Calarese et al., 2008	11; M	LOC – Staring <b>rt</b> head turn bilateral mydriasis – flush <b>lt</b> hemiparesis; at times unresponsive	3 min	Un-known	MRI: normal	<b>Rt</b> fronto-temporal PLEDs
Zhao et al 2010	38; F	LOC - facial grimacing – <b>neck atonia</b> – <b>generalized atonia</b>	2–3 s atonia	Un-known	MRI: normal	Generalized discharge at onset of atonia; <b>rt</b> frontal seizure pattern



# Atonic seizure

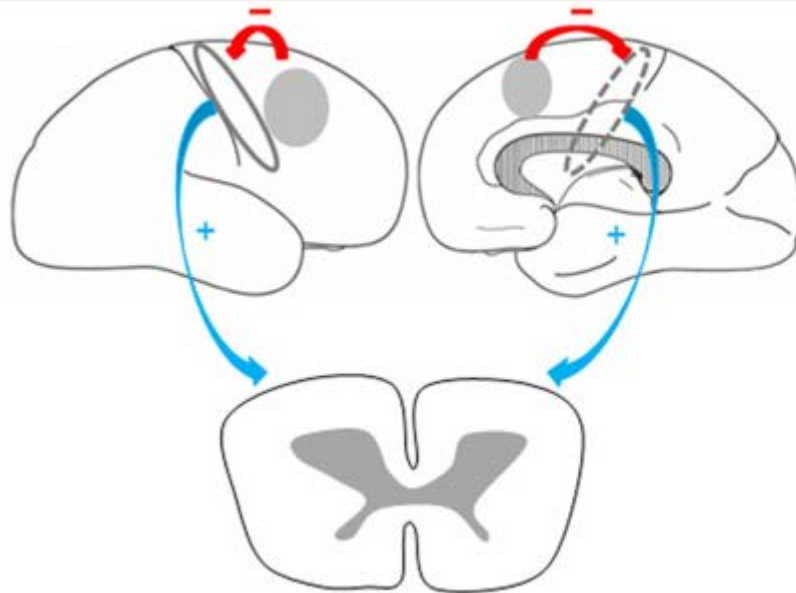
- The gold standard in diagnosing **ictal atonia** : simultaneous recording of the **EEG** and preferably also the **EMG** to demonstrate absence of myographic activity although the diagnosis can be made based on clinical observation.

So NK,1995, Noachtar, 2000

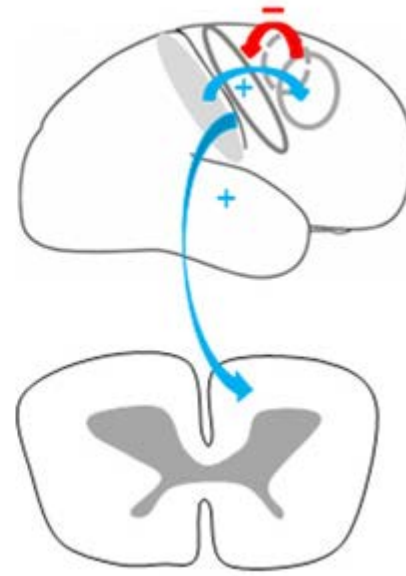
# Proposed pathophysiology and clinical characteristic of atonic seizures

## A. PNMA/SNMA activation

PNMA primary negative motor area , SNMA supplementary negative motor area



## B. S1 activation -> PNMA/SNMA activation



Duration of atonia in a single sz : **min**

Muscle atonia typically **not** preceded by a positive phenomenon

Predominantly **contralateral** but also **ipsilateral** distal muscles

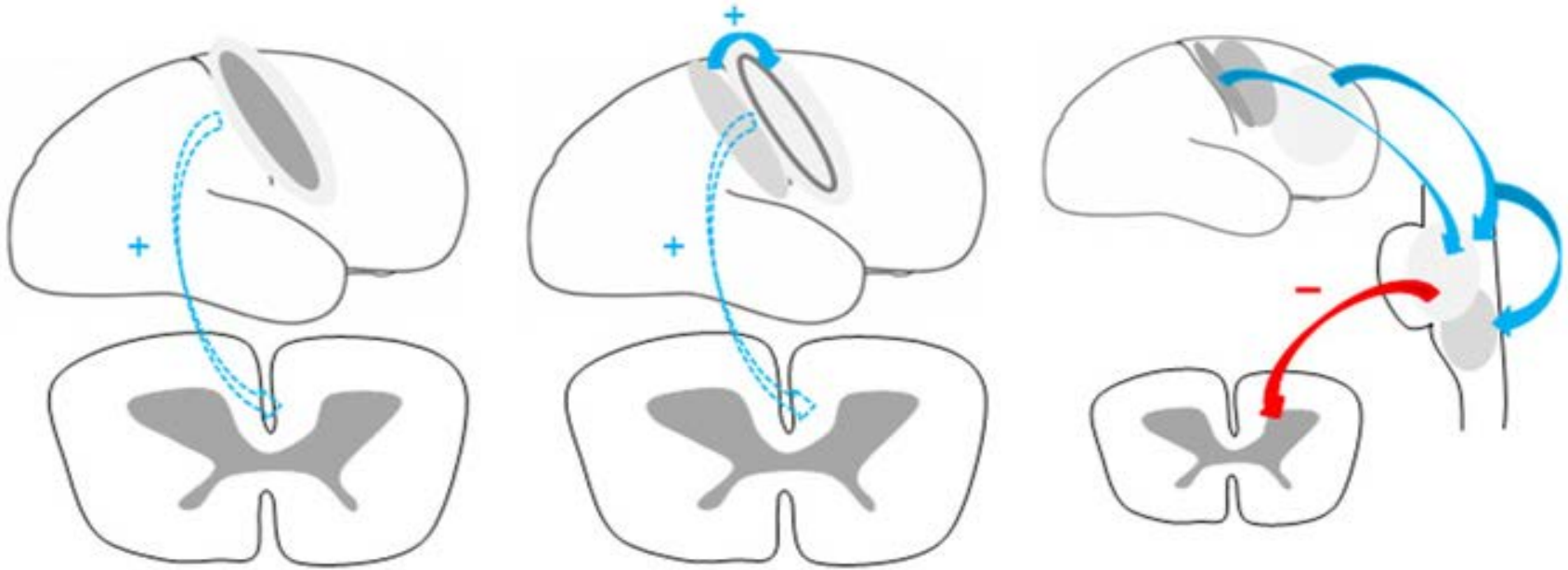
# Proposed pathophysiology and clinical characteristic of atonic seizures

**C. M1 activation**  
Silent period

**D. S1 activation -> M1 activation**  
Silent period

**E. Cortico-reticulo-spinal pathway**

silent period is characterized by a decreased output of M1



Duration of atonia in a single sz : **msec-sec**

Muscle atonia typically preceded by **a positive phenomenon**

**Contralateral, distal muscles**

Duration of atonia

in a single sz : **msec-sec**

Muscle atonia typically preceded by

**a positive phenomenon**

**Bilateral trunk muscles**

# Conclusion

- Atonic seizure is difficult to treat: syndrome
- Atonic seizure : phenomena of both generalized and focal epilepsy
- Atonic seizure : danger from falling should be prevented
- Intractable atonic seizure : palliative epilepsy surgery ; callosotomy may be helpful
- Atonic seizure : focal lesion can be treated by resective epilepsy surgery

**Thank you for your attention**