FOCAL EPILEPSY AND SEIZURE SEMIOLOGY

Piradee Suwanpakdee, MD.

Division of Neurology

Department of Pediatrics

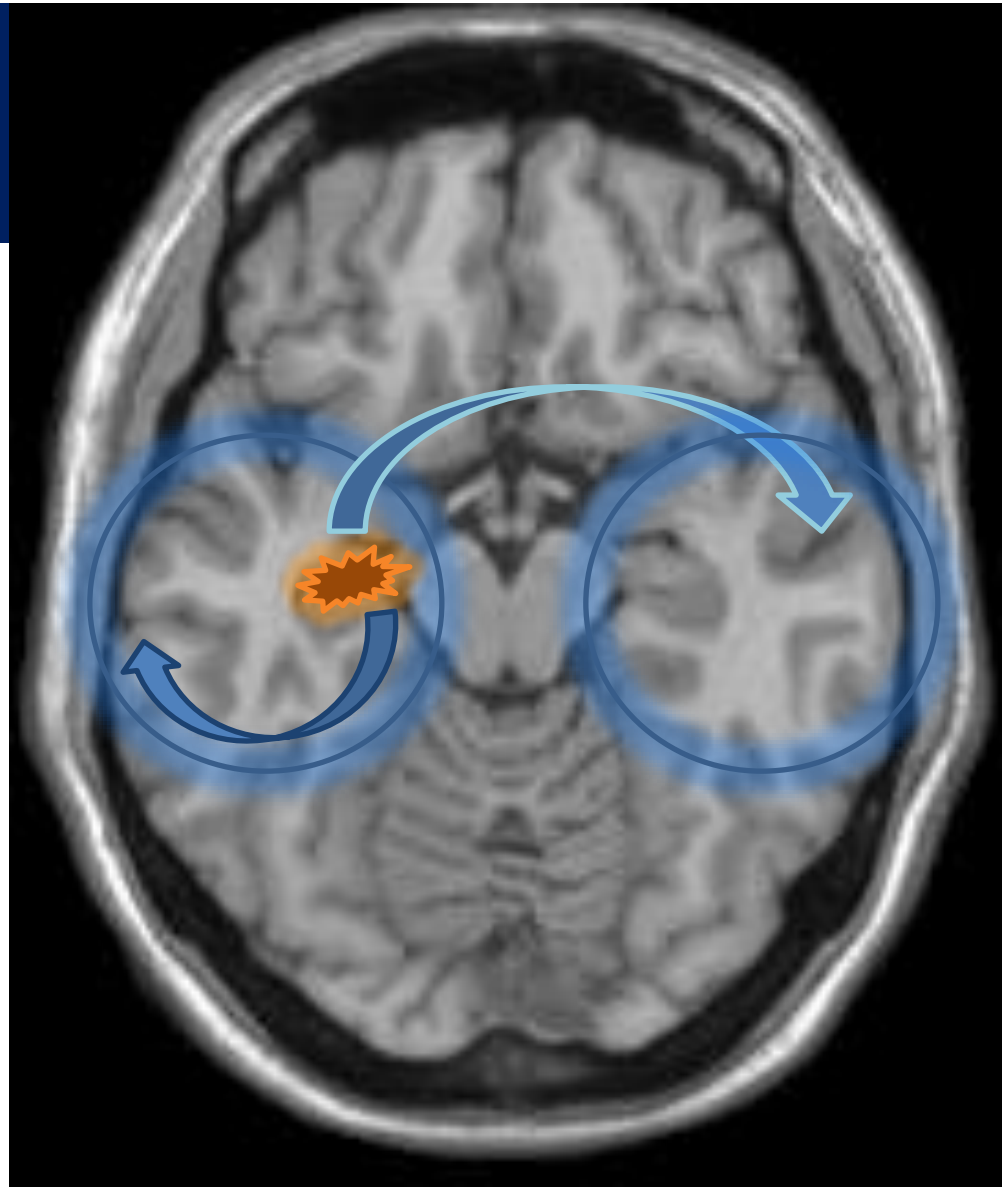
Phramongkutklao Hospital

OUTLINE

- **Focal onset seizure classification**
- **The definition of semiology**
- **How can we get the elements of semiology?**

Focal seizures

- Originate within networks limited to one hemisphere
- May be discretely localized or more widely distributed….



www.ilae.org

ILAE 2017 Classification of Seizure Types Expanded Version[1]

Focal Onset

Aware	Impaired Awareness

Motor Onset
- automatisms
- atonic[2]
- clonic
- epileptic spasms[2]
- hyperkinetic
- myoclonic
- tonic

Non-Motor Onset
- autonomic
- behavior arrest
- cognitive
- emotional
- sensory

focal to bilateral tonic-clonic

Generalized Onset

Motor
- tonic-clonic
- clonic
- tonic
- myoclonic
- myoclonic-tonic-clonic
- myoclonic-atonic
- atonic
- epileptic spasms[2]

Non-Motor (absence)
- typical
- atypical
- myoclonic
- eyelid myoclonia

Unknown Onset

Motor
- tonic-clonic
- epileptic spasms

Non-Motor
- behavior arrest

Unclassified[3]

1 Definitions, other seizure types and descriptors are listed in the accompanying paper and glossary of terms.

2 These could be focal or generalized, with or without alteration of awareness

3 Due to inadequate information or inability to place in other categories

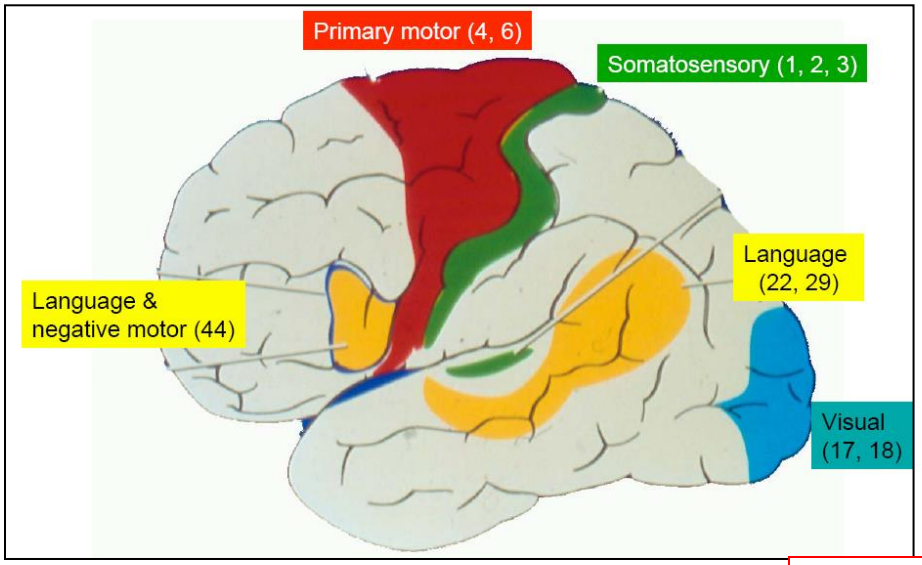
From *Fisher et al. Instruction manual for the ILAE 2017 operational classification of seizure types. Epilepsia doi: 10.1111/epi.13671*

What is the seizure semiology?

- **Seizure semiology is an expression of activation and disinhibition of cerebral areas**

- **It thus provides some information what cerebral areas are “involved” during a seizure**

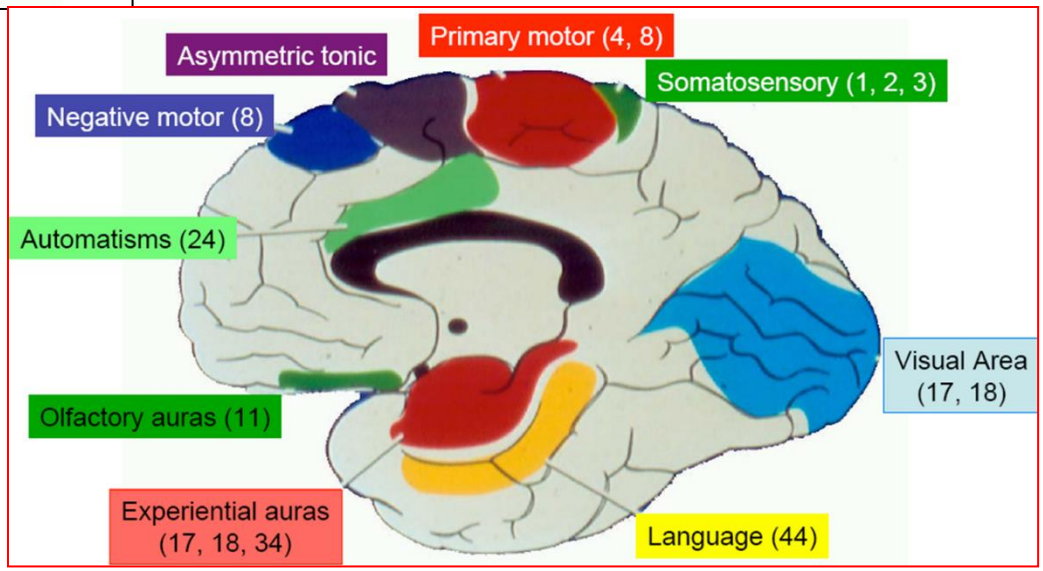
Symptomatogenic areas



Left hemisphere

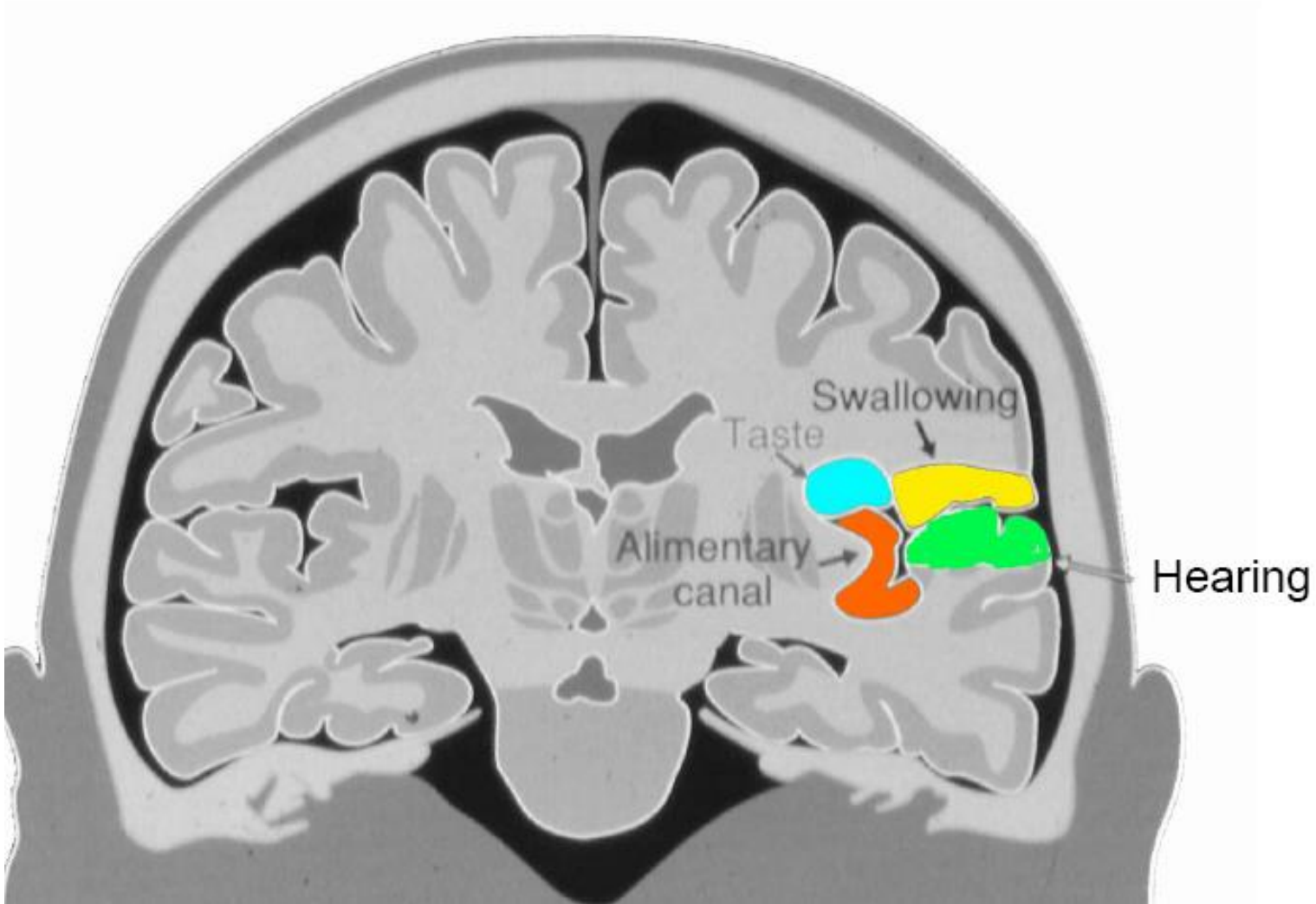
lateral aspect

Mesial aspect



Symptomatogenic areas

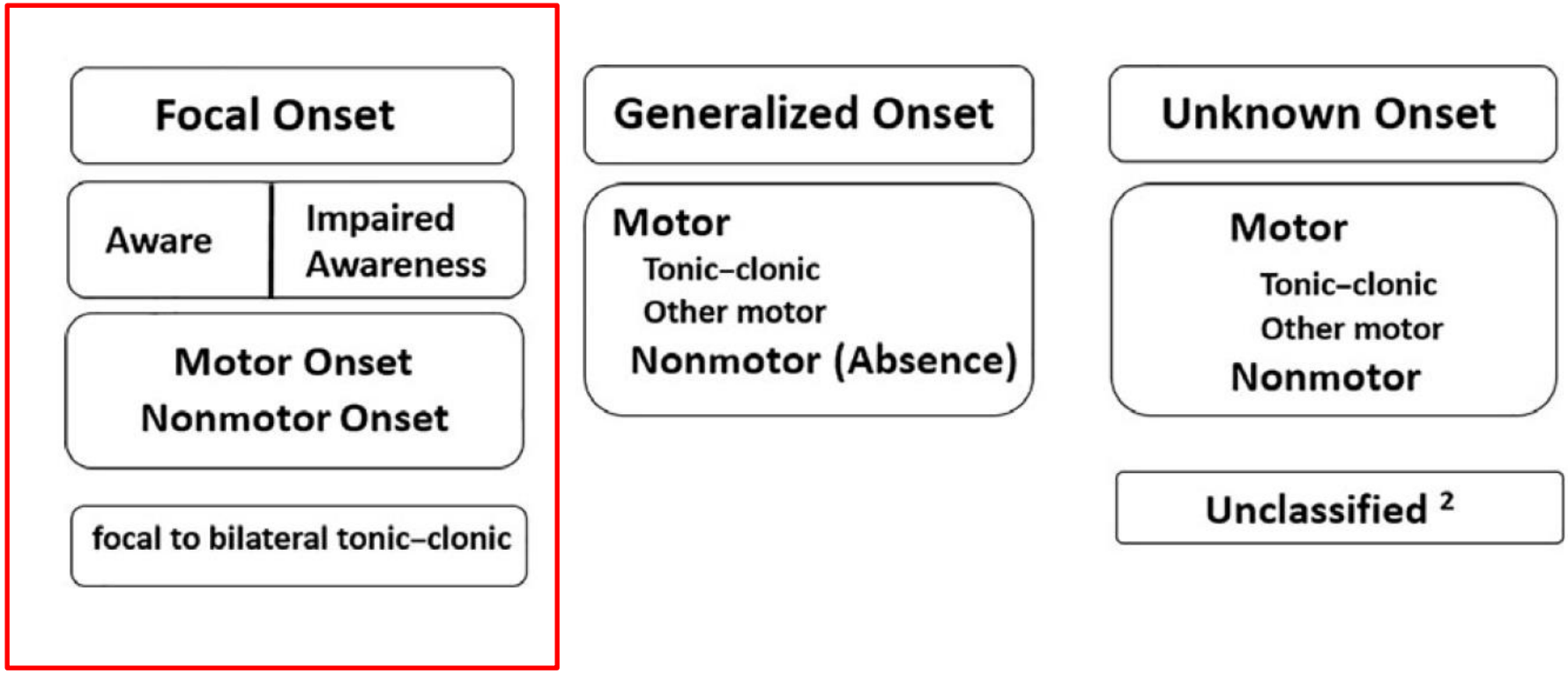
Left Insula



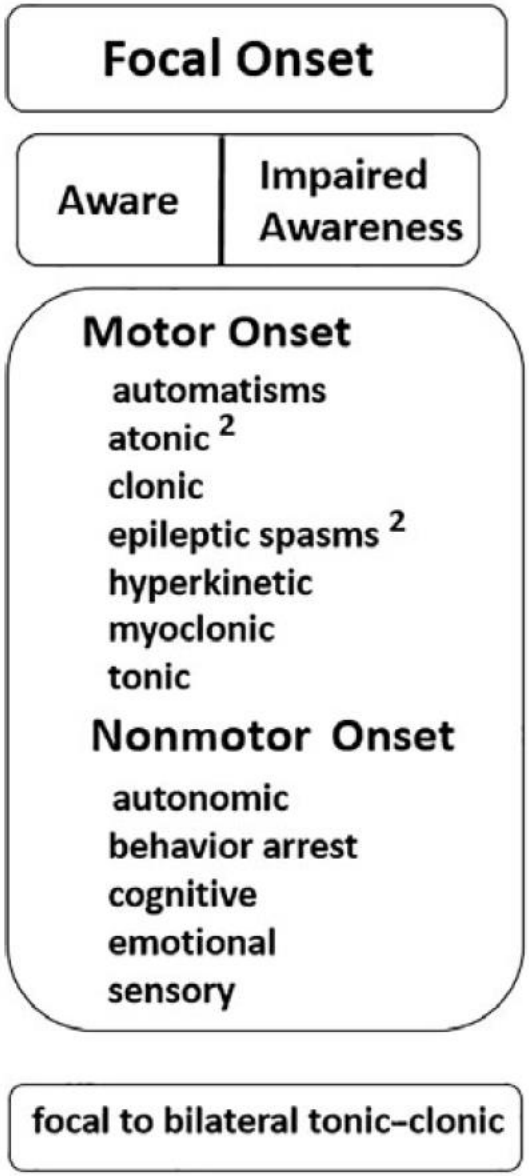
How can we get the elements of seizure semiology?

- **The information on semiology comes from patient's and witness' history**
- ***Video EEG* provides objective data on seizure semiology**
- ***Seizure classification* aims to intellectually organise and summarise information about seizure semiology**

ILAE 2017 Classification of Seizure Types Basic Version [1]



ILAE 2017 Classification of Seizure Types Expanded Version [1]



Notes

- Atonic seizures and epileptic spasms would *not* have level of awareness specified
- Pedalling grouped in hyperkinetic rather than automatisms (arbitrary)
- Cognitive seizures
  - impaired language
  - other cognitive domains
  - positive features eg déjà vu, hallucinations, perceptual distortions
- Emotional seizures: anxiety, fear, joy, etc

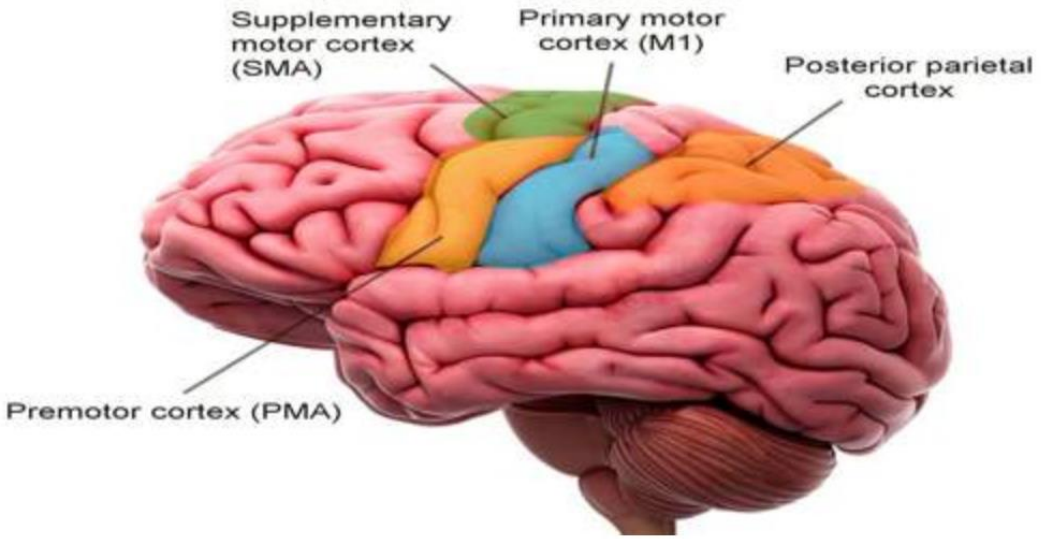
Case

Focal onset aware seizure

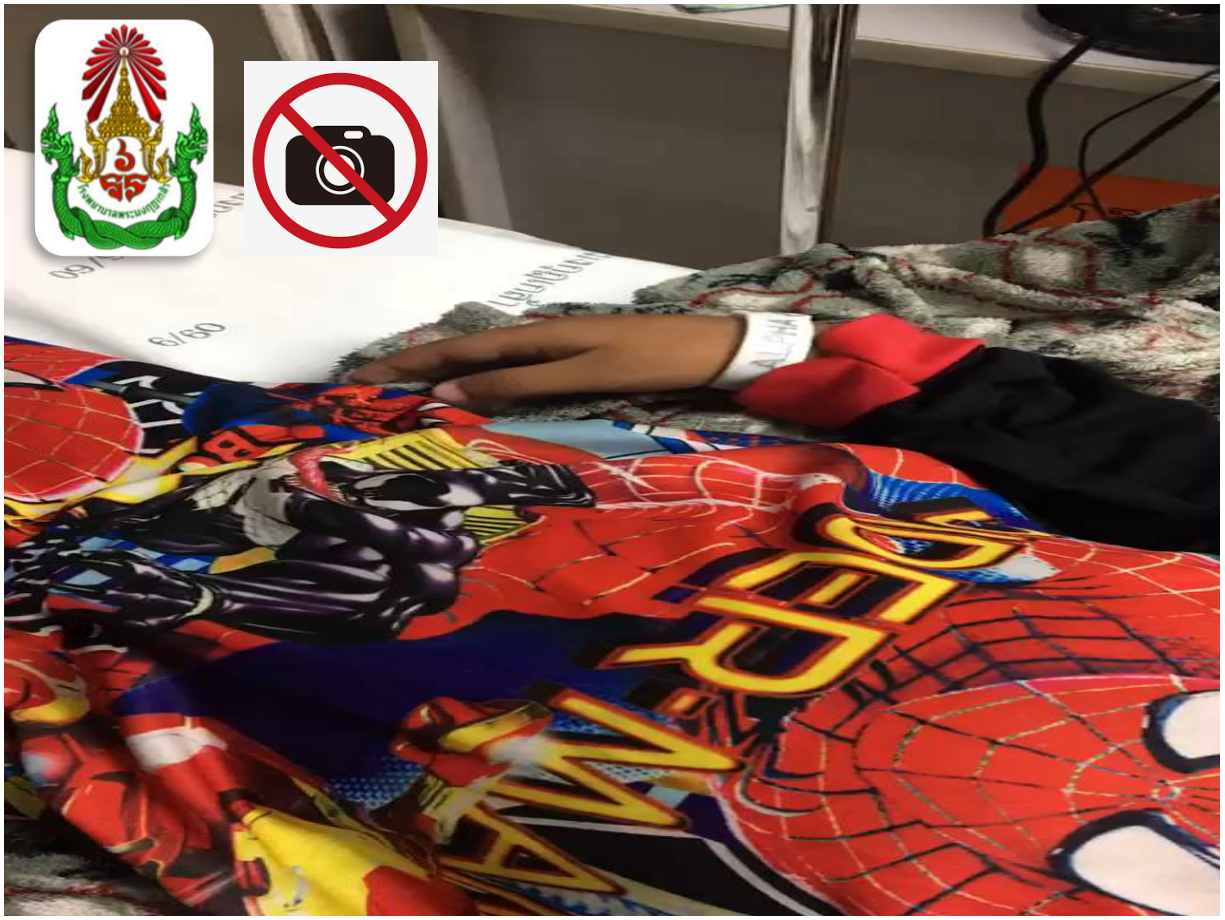
- This term replaces simple partial seizure
- A seizure that starts in one area of the brain and the person remains alert and able to interact is called a focal onset aware seizure.
- These seizures are brief, lasting seconds to less than 2 minutes.

Focal clonic seizure

- **Indicate involvement of contralateral primary motor cortex**
- **Reliability is good**



Epilepsia partialis continua



focal motor status involving a small portion of the sensorimotor cortex

Case

Focal Onset Impaired Awareness Seizures

- A seizure that starts in one area of the brain and the person is not aware of their surroundings
- Focal impaired awareness seizures typically last 1 to 2 minutes.
- These seizures include automatisms (such as lip smacking, picking at clothes), becoming unaware of surroundings, and wandering.
- Not localized or lateralized
- Duration of seizures has a localizing value
  - Mesial temporal seizure -> longer duration than frontal lobe seizure

Automotor seizures

- Repetitive, stereotyped, semipurposeful motor behaviors, involving primarily distal limbs, mouth, and tongue
- 95% associated with altered consciousness

  - Preservation of consciousness -> non-dominant mesial temporal epilepsy
- Temporal lobe > Frontal lobe epilepsy (shorter duration)
- Unilateral automatisms: ipsilateral epileptogenic zone

Atonic seizure

Sudden loss or diminution of muscle tone lasting ~1–2 s involving head, trunk, jaw, or limb musculature

Atonic seizure

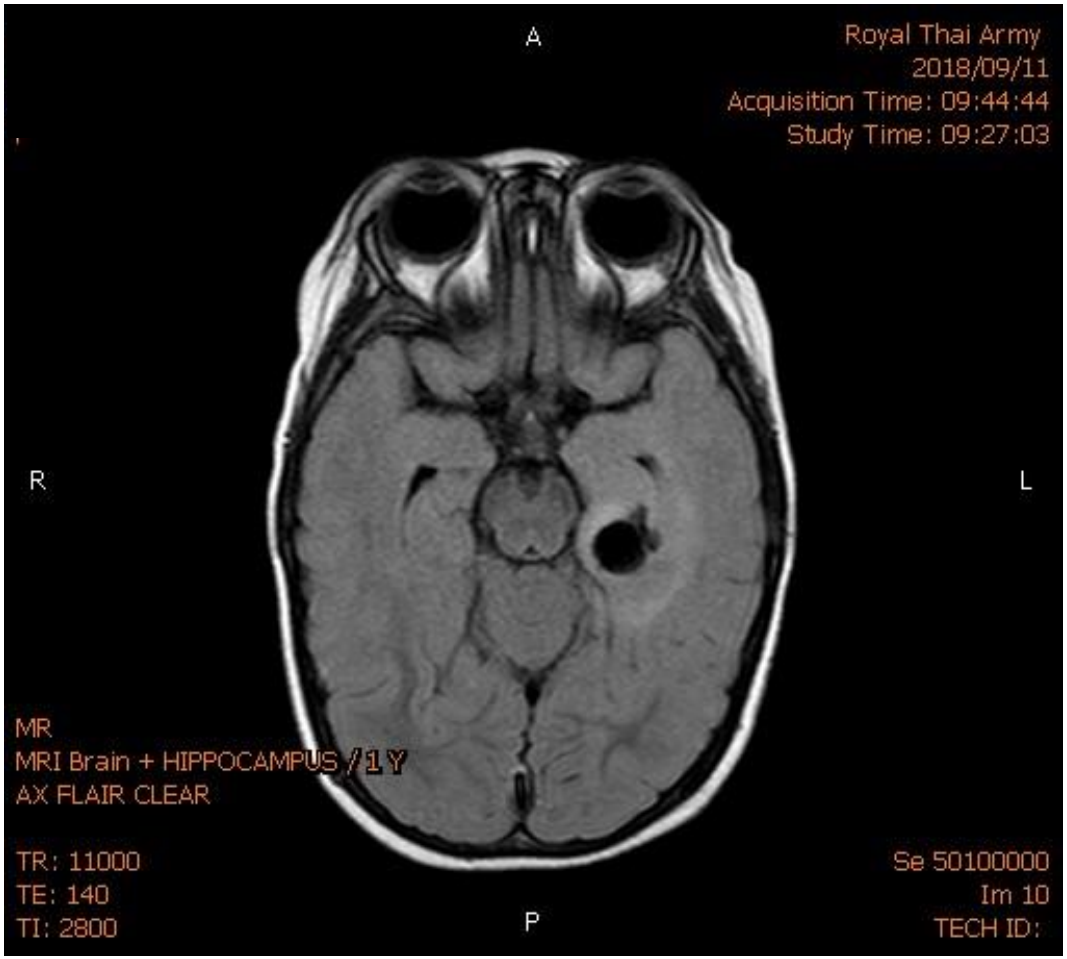
- Atonic means a loss of muscle tone
- They are also known as drop attacks
- Atonic seizures can begin in one area or side of the brain (focal onset) or both sides of the brain (generalized onset)
- Often seen in syndromes like Lennox-Gastaut or Dravet syndrome

Case

Epileptic spasms

- Sudden flexion, extension or mixed flexion-extension of proximal and truncal muscles, lasting 1-2 seconds
- Spasms typically occur in a series, usually on wakening
- CAUTION Epileptic spasms usually occur in a series (several in a cluster) if singular, consider other seizure types
- Generalized epilepsies > focal epilepsy (parieto-occipital)

MRI brain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Case

Focal hyperkinetic seizure

- This seizure type involves movements of proximal limb or axial muscles, producing irregular large amplitude movements, such as pedaling, pelvic thrusting, jumping, thrashing and/or rocking movements
- Consciousness may be preserved
- Occur mostly during sleep
- Pathophysiology:
  - Primarily an expression of the epileptic activation of orbitofrontal or mesial frontal lobe structures, but may also be the result of a propagation from other structures (TL, insula)

Case

Myoclonic seizure

A myoclonic seizure is a single or series of jerks
Each jerk is typically milliseconds in duration

Myoclonic seizure

- Sudden muscle jerks of variable topography (distal, proximal, axial): uni- or bilateral, focal, multifocal or generalised
- Prominently affecting shoulders and proximal arms
- Consciousness likely preserved
- 100-400 msec in duration
- Unilateral myoclonic seizures -> contralateral primary motor area or premotor cortex

Asymmetrical tonic seizure

Focal Onset

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- emotional
- sensory

If consistent focal features ->consider focal seizure involving the frontal lobe (SMA)

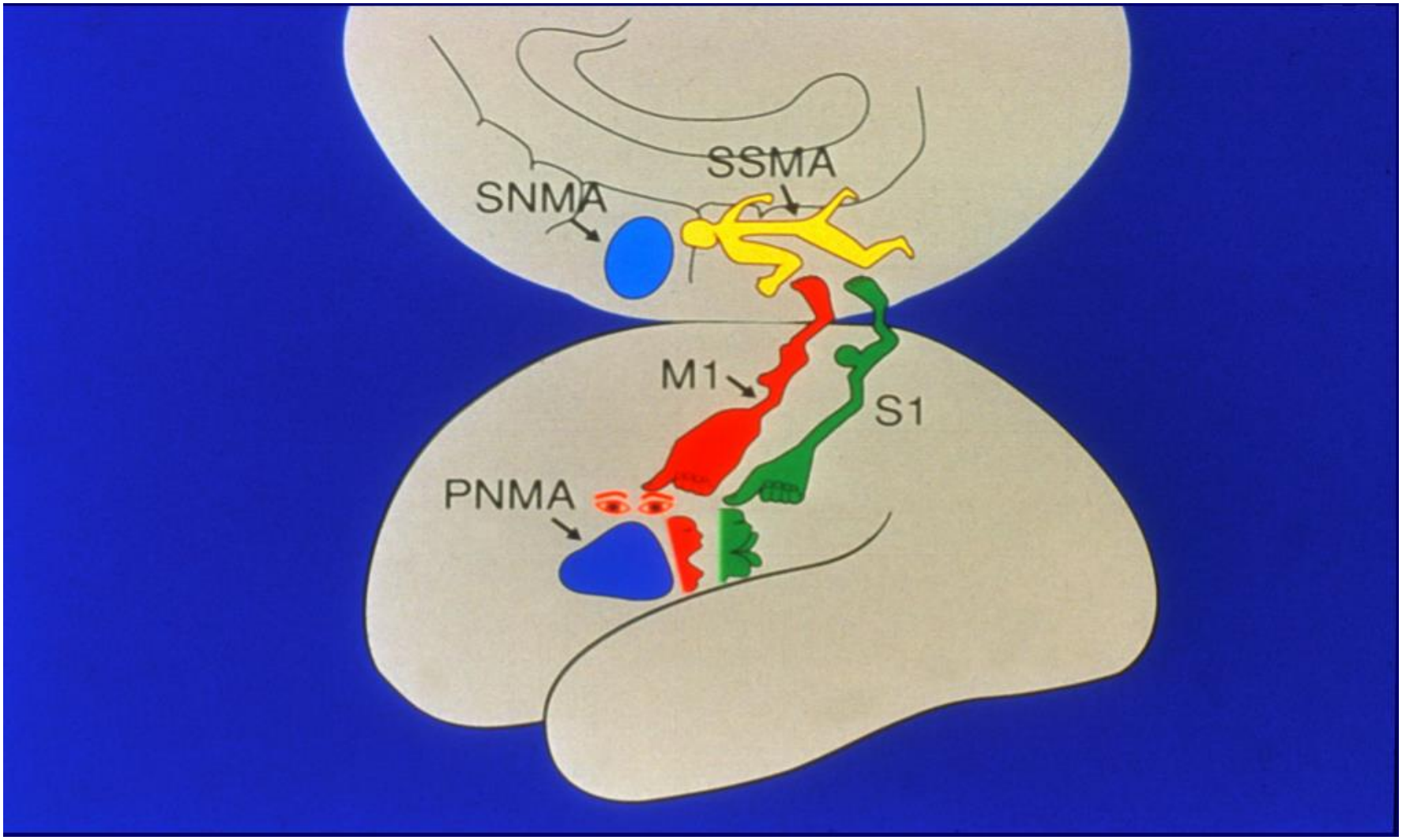
Asymmetrical tonic seizure

- Preferentially affect proximal muscle on both sides, but more prominent over the contralateral side
- Conscious is intact in most patients at least at the seizure onset
- Asymmetric tonic limb posturing “sign of four”

-> Hemisphere *contralateral* to extended arm

- Location: Supplementary sensorimotor area (SSMA)

Supplementary sensorimotor area (SSMA)

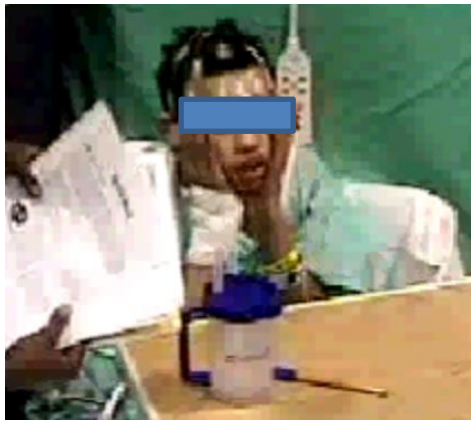


Tonic/dystonic seizure

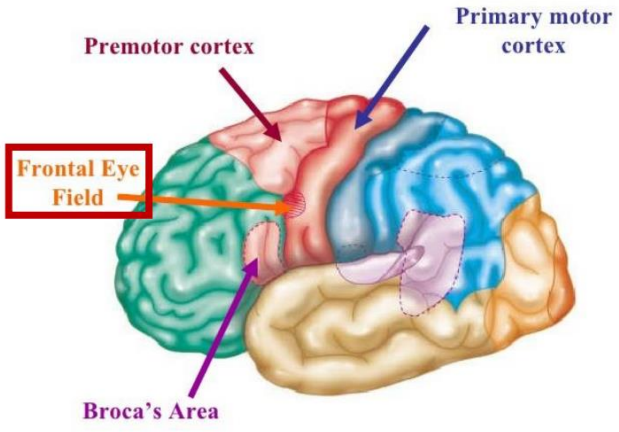
right hand dystonic-tonic-> fall down-> right face clonic-> right arm/leg clonic->GTC

Versive seizures

- **Forced and involuntary** turning of the head and eyes in one direction with an associated neck extension resulting in a sustained unnatural position
- Symptomatogenic zone-> Frontal eye fields, highly lateralizing to the contralateral hemisphere



Right head version



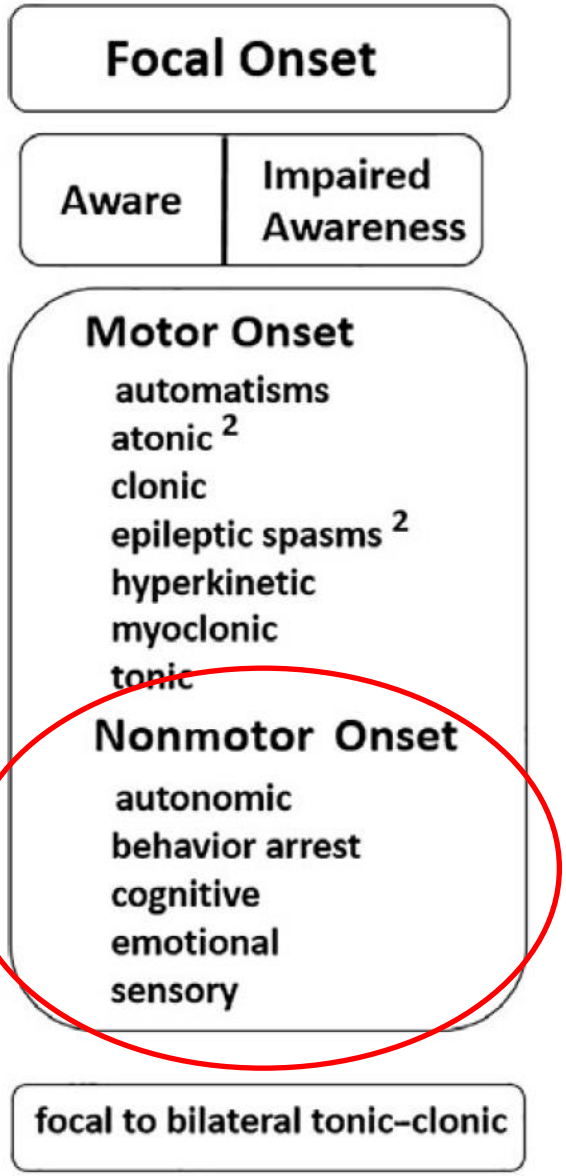
Common lateralising seizure manifestations

Symptom	Localisation	Specificity	Frequency*
Forced head turn ("version")	Contralateral	>90%	35-40%
Unilateral dystonic posturing	Contralateral	>90%	20-35%
"Figure of Four"	Contralateral	90%	65% (sGTCS)
Postictal nose wiping	Ipsilateral	>70%	10-50%
Ictal speech	Nondominant	>80%	10-20%
Ictal automatisms with preserved awareness	Nondominant	100%	5%
(Post)ictal dysphasia	Dominant	>80%	20%

*In patients referred for presurgical video telemetry

Courtesy: Dr.Prakash kotagal

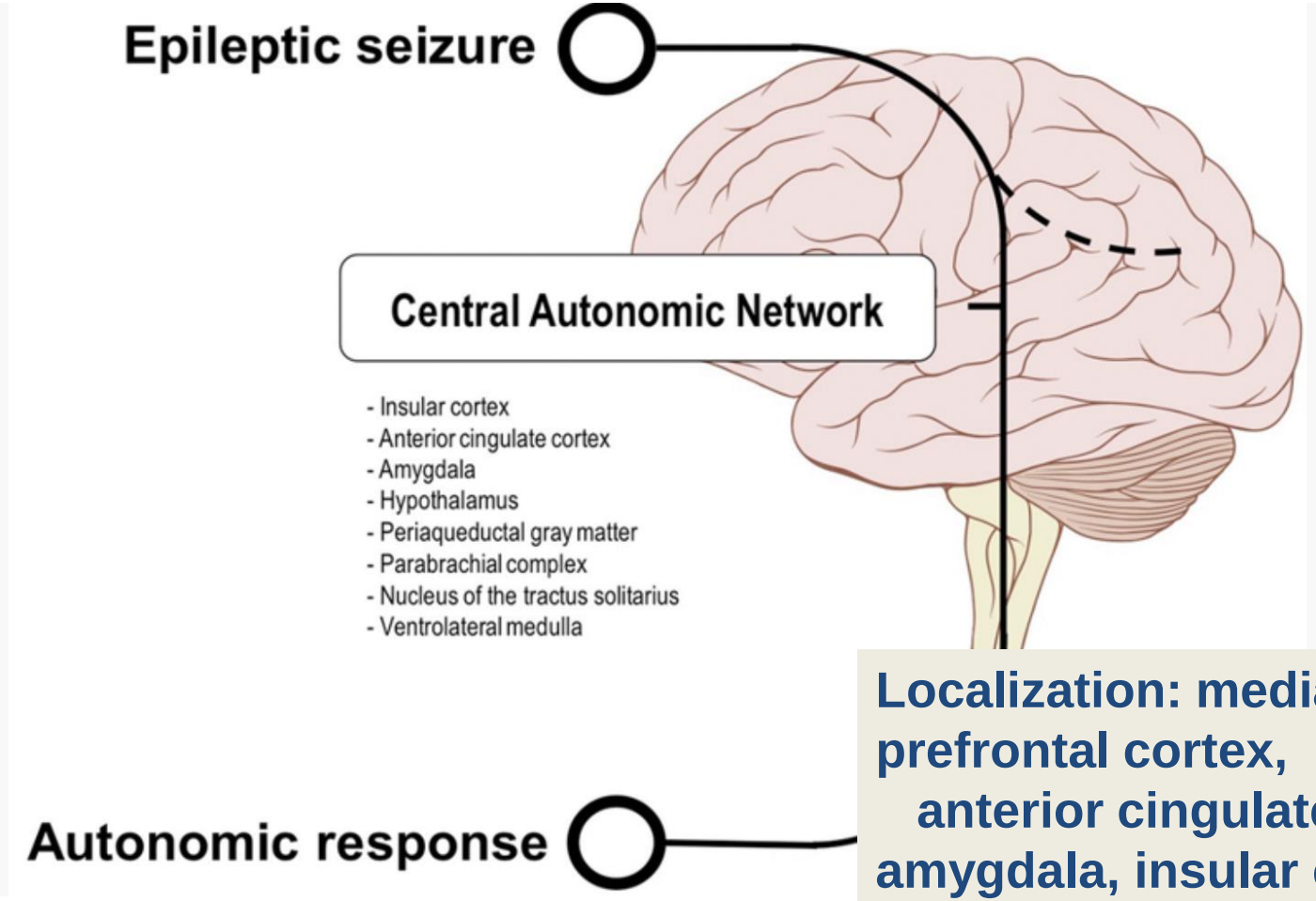
ILAE 2017 Classification of Seizure Types Expanded Version [1]



Focal autonomic seizures

- Characterized by alterations in systems controlled by the autonomic nervous system at seizure onset.
- Ictal tachycardia is the most common ictal autonomic manifestation
- Ictal vomiting: nondominant TLE
- Ictal spitting: nondominant TLE
- Ictal hypersalivation: nondominant TLE

Focal autonomic seizures



Localization: medial prefrontal cortex, anterior cingulate, amygdala, insular cortex

Focal emotional seizure

- Characterized by alterations in mood or emotion, or the appearance of altered emotion without the subjective emotion, at seizure onset
- Described as:
  - Focal emotional seizure with fear/anxiety/panic
  - Focal emotional seizure with laughing (gelastic)
  - Focal emotional seizure with crying (dacrystic)
  - Focal emotional seizure with pleasure
  - Focal emotional seizure with anger

Case a 15 years old female with intractable epilepsy

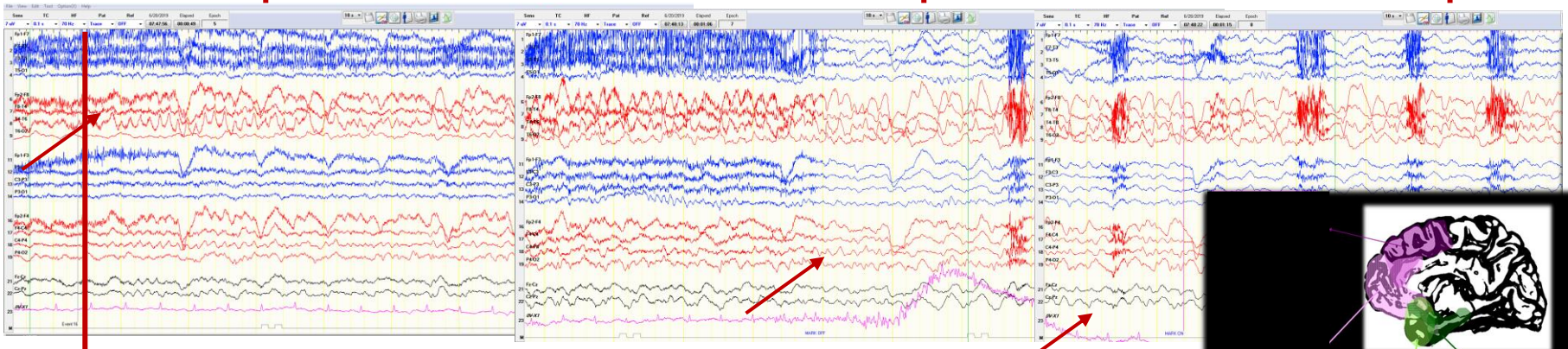
- Seizure semiology described as most of seizure **started with fear** then left arm stiffeness followed shortly by numbness or pain that going down form shoulder toward leg, with postictal left sided weakness. Sometimes oroalimentary automatisms were noted.

Courtesy of General Charcrin Nabangchang, MD

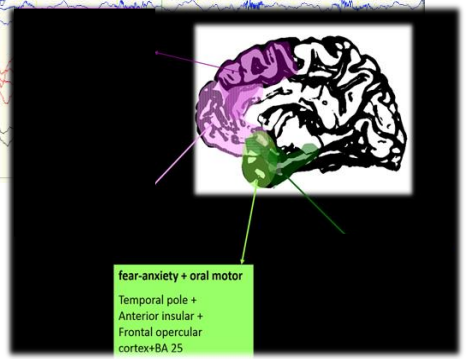
Fear

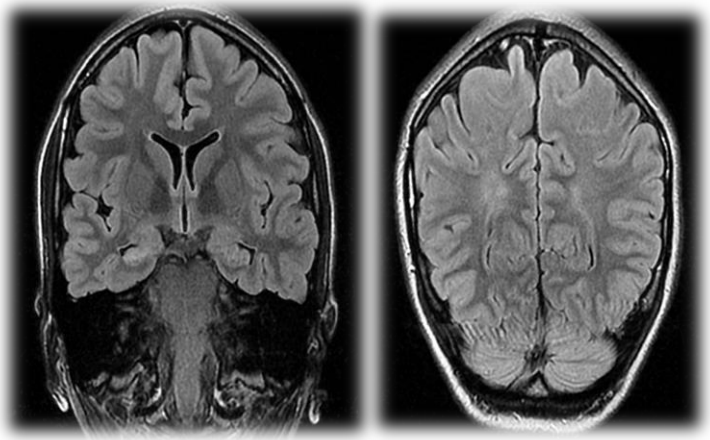
Left arm stiffed/ felt numb

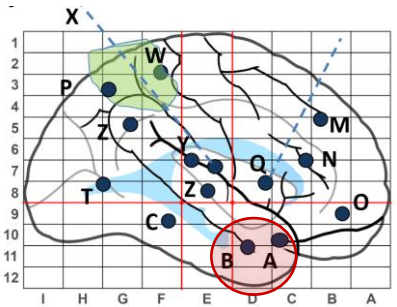
Mouth automatisms



EEG onset
R temporal







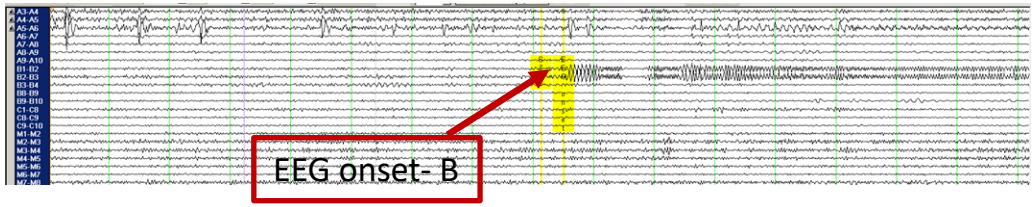
- A 15 years old female with intractable epilepsy
- EEG: 2 Ictal onset: F8T4, T4P4
- MRI brain: 2 lesions

  (R hippocampal sclerosis,

  ulegyria of R precuneus)
- Ictal SPECT:Increased perfusion at right

  frontal cortex (Injection time 10 s)

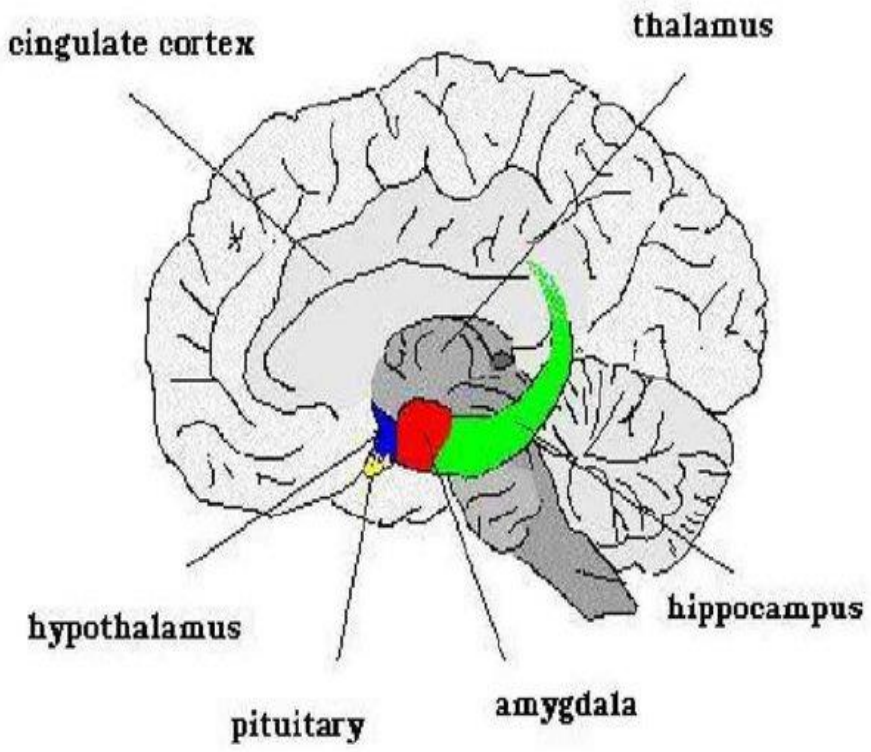
Plan: SEEG exploration



Seizure outcome: Engel I

Ictal Fear

- Ictal Fear (IF) with coordinated behavior and autonomic features may be part of or interfere with a complex information processing network involving orbito-prefrontal, anterior cingulate and temporal limbic cortices

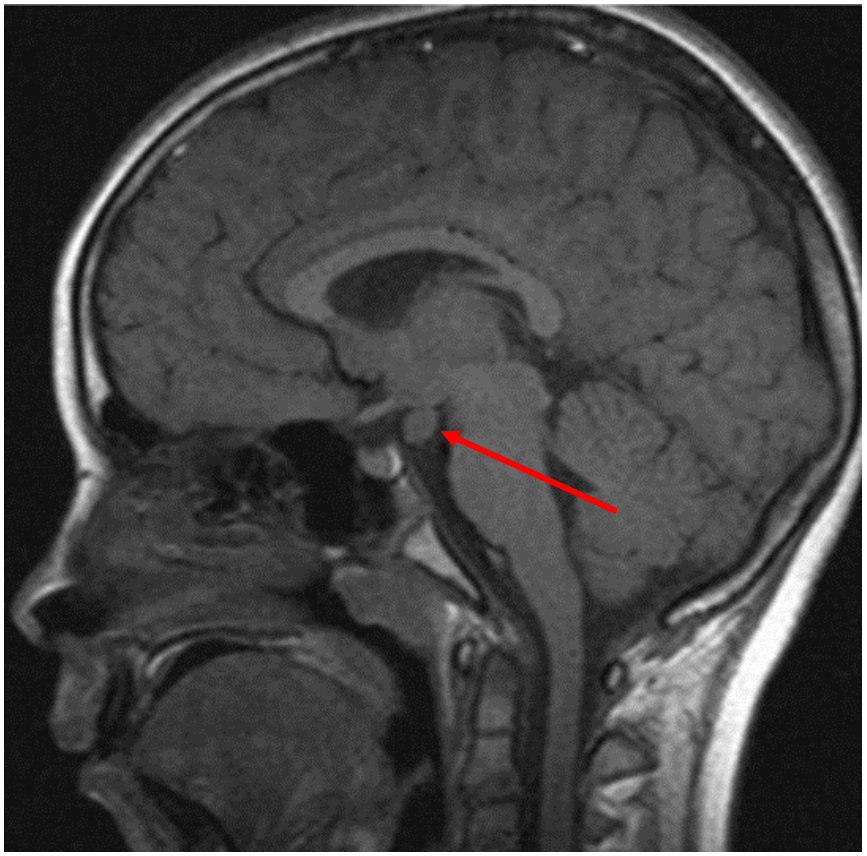


Limbic network

Gelastic seizure

- Bursts of laughter or giggling, usually without appropriate related emotion of happiness

Gelastic seizure

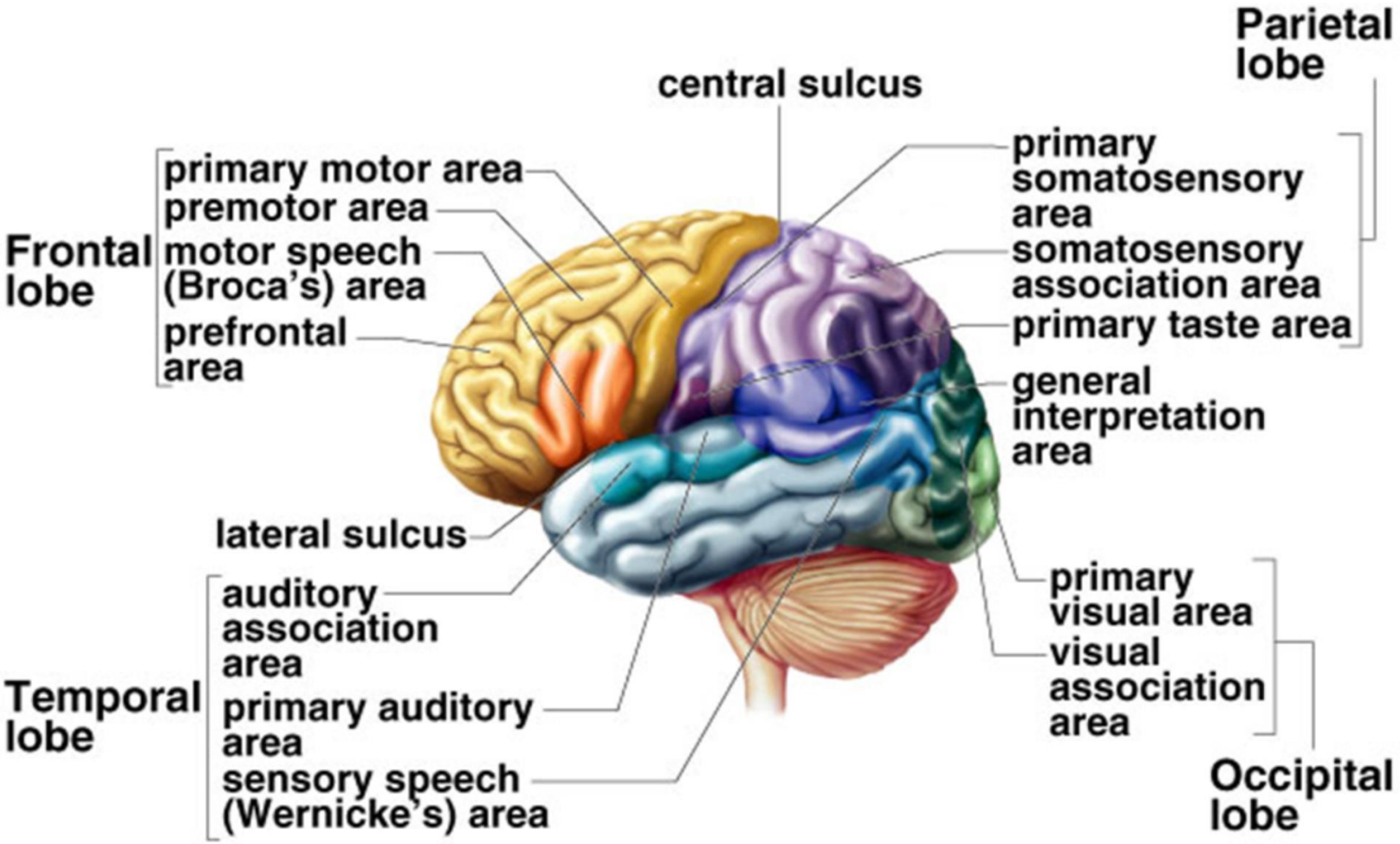


- This seizure type is characteristic of seizures arising in the hypothalamus (*Hypothalamic hamartoma*)
- But can occur in seizures arising in the *frontal* or *temporal* lobes.

SENSORY SEIZURE

- Focal sensory seizures are one type of epileptic 'aura’
- The 'aura' reflects the initial seizure discharge in the brain
- Types:
  - Somatosensory (S1, S2, SMA)
  - Visual (visual cortex, temporal asso. cortex)
  - Auditory (Heschl’s gyrus, temporal asso. cortex)
  - Olfactory (amygdala, OF cortex (gyrus rectus))
  - Gustatory (S2 and rolandic operculum, insula)
  - Vestibular (insular-parietal-temporal)

SENSORY SEIZURE

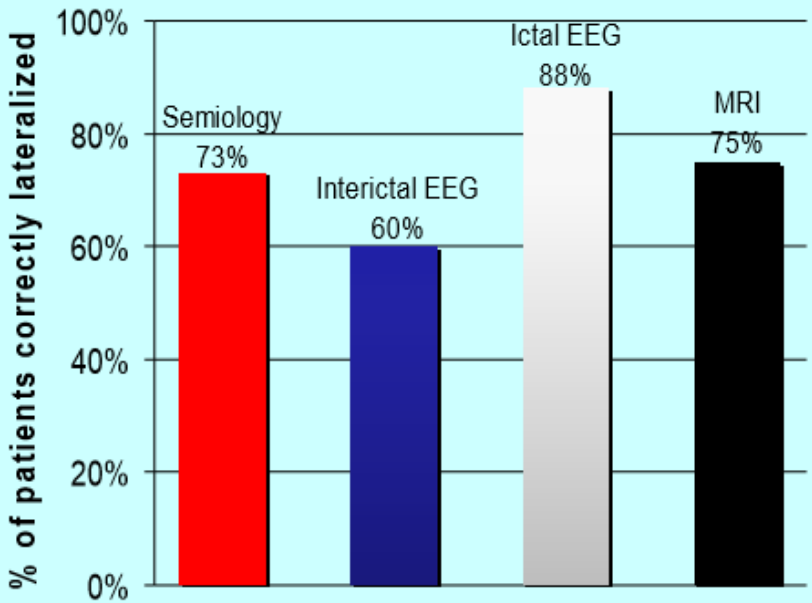


Why is the seizure semiology important?

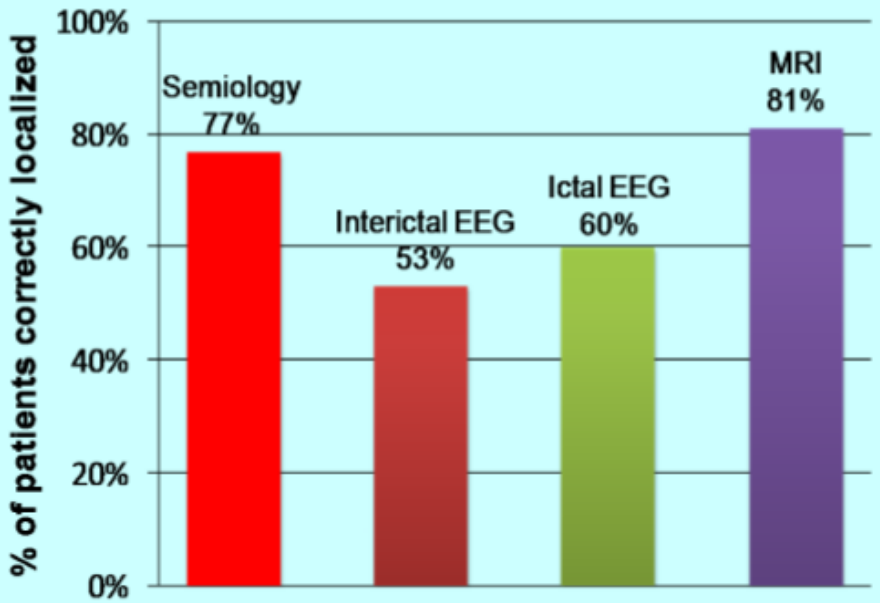
- **Semiological features strongly suggest lobe or origin and spread pattern**
- **Semiology is a unique way to study mechanisms of pathways of ictal spread**
- **Excellent for seizure lateralization; certain auras also useful for localization**
- **Give a clue for epilepsy surgery**

Seizure Semiology may be as good as EEG, MRI

LATERALIZING VALUE



LOCALIZING VALUE



Elwan S, Alexopolous A, Silveria D, Kotagal P (in preparation)

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*In patients referred for presurgical video telemetry

Courtesy: Dr.Prakash kotagal

Less common lateralising or localising seizure manifestations

Symptom	Localisation	Specificity	Frequency
Elementary visual aura	Contralateral occipital	>90%	?
Acoustic aura	Temporal, if unilateral then contralateral	>90%	?
Olfactory aura	Mesiotemporal	>70%	?
Abdominal aura ➔ Automotor sz	Temporal Temporal	90% 98%	Common
Ictal aphasia	Dominant	>80%	?
Ictal nystagmus	contralateral	>95%	?
Hyperkinetic movements	Frontal/frontomesial	>80%	>10%

A 16-year-old girl with intractable seizure

- **Seizure description**
- **Prior to seizures she may feel anxiety or panic, or she may feel "over-heated“**
- **At seizure onset she has blank staring with unresponsiveness**
- **And version of head and eyes to the right with clonic twitching of the head to the right for 10 or 15 seconds, followed by evolution to generalized tonic clonic convulsion.**

How can we classify?

A 16-year-old girl with intractable seizure

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Psychic/autonomic aura

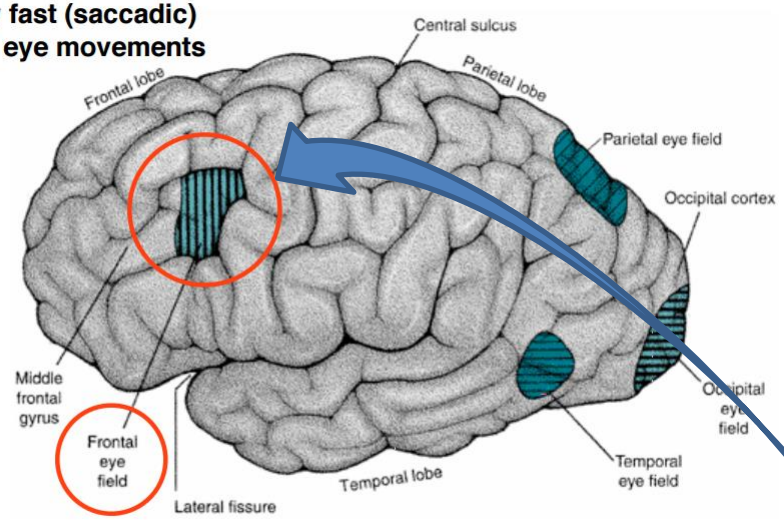
Focal impaired awareness

Right head version

Focal to bilateral tonic clonic

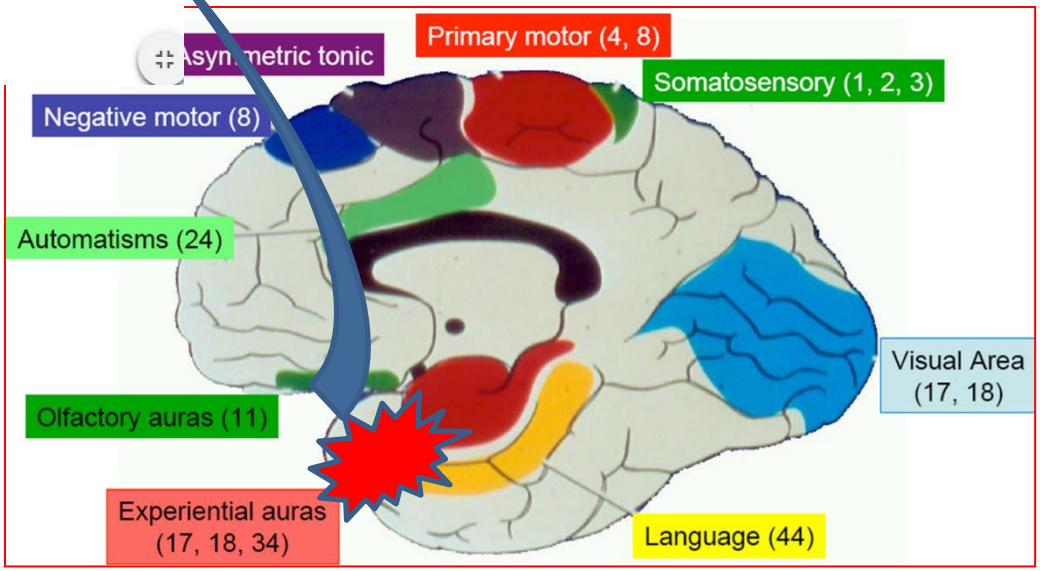
Symptomatogenic areas

Voluntary fast (saccadic) horizontal eye movements



Young, Young, and Tolbert Fig. 10-4A

- Psychic/autonomic aura
- Focal impaired awareness
- Right head version
- Focal to bilateral tonic clonic



Take home message

- **The elements of semiology strongly suggests the seizure onset and spread pattern**
- **Detailed of semiology from history taking is very important**
- **Video EEG provides objective data**
- **Don’t forget to analyze semiology from the first symptom/sign until the end**
- **Then you will understand epilepsy the underlying epileptic network**

Transparent language: use words that mean what they say

Thank you for your attention