Phramongkutklao Comprehensive
Pediatric Epilepsy Center of Excellence
Integration • Passion • Wisdom

FOCAL EPILEPSY AND SEIZURE SEMIOLOGY

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OUTLINE

Focal onset seizure classification

What is the semiology?

Lateralizing/localizing value of semiology

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

Nonmotor 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

Nonmotor (absence)

- typical
- atypical
- myoclonic
- eyelid myoclonia

Unknown Onset

Motor

- tonic–clonic
- epileptic spasms

Nonmotor

- behavior arrest

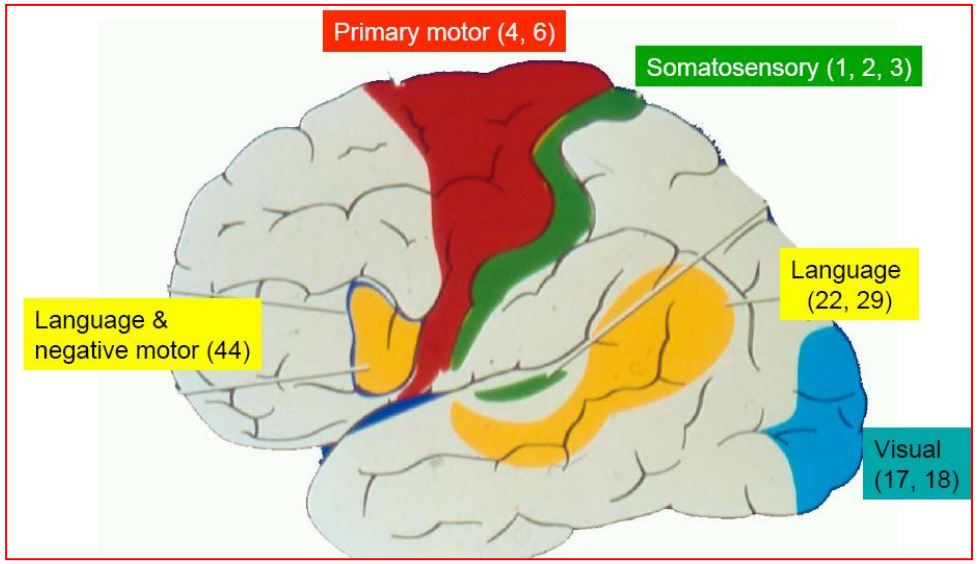
Unclassified [3]

What is the seizure semiology?

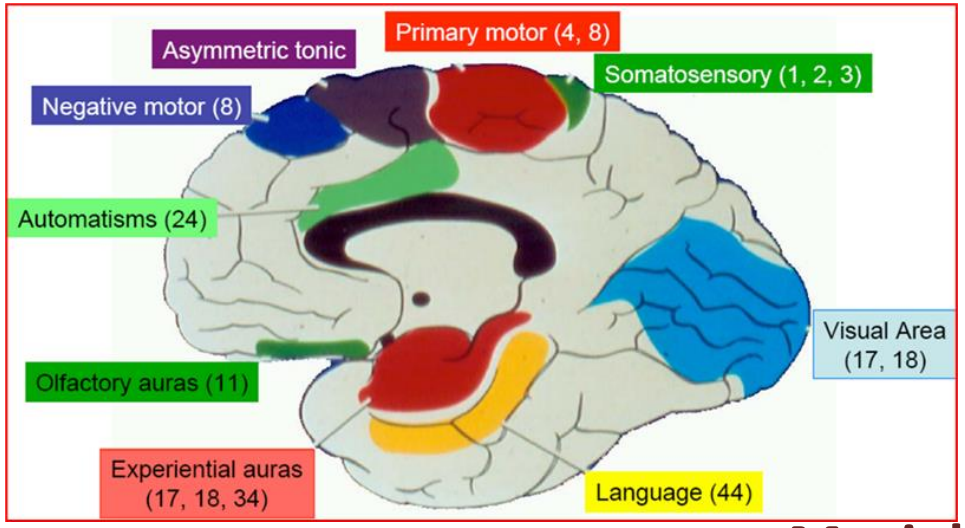
- Seizure semiology is the manifestation of the activation of the **symptomatogenic zone**

- *A simple and cost-effective tool* that allows localization of the symptomatogenic zone which either **overlaps** or **close** proximity of

  “the epileptogenic zone”



lateral



Mesial

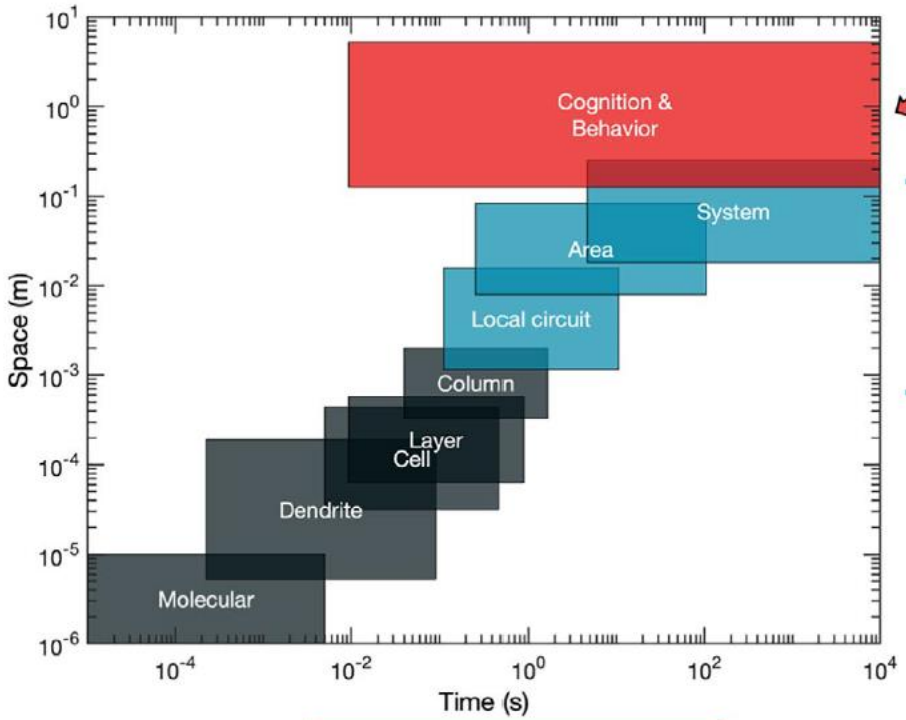
A *multi-scale framework* to think about spatial and temporal features in epileptic seizure expression

Spatial features of discharge
e.g., anatomical localization (primary cortex versus associative cortex; functional specificities); focal or widespread onset; propagation

Other factors modulating electroclinical expression

Bottom up effects on network, e.g. sleep versus wake, arousal, antiepileptic drugs; stimulation versus spontaneous seizure; specific neurotransmitter systems, genetic profile?

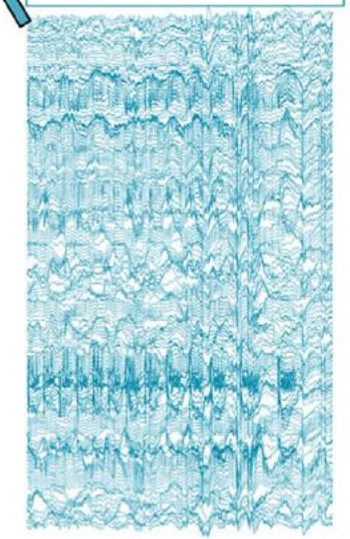
Top down effects on network, e.g interaction with person/objects, emotional state, specific seizure triggers (e.g; noise, stress)



Temporal features of discharge
e.g., seizure onset pattern; discharge frequency (fast versus slower); relations between activity in different structures (phase lag, synchrony)

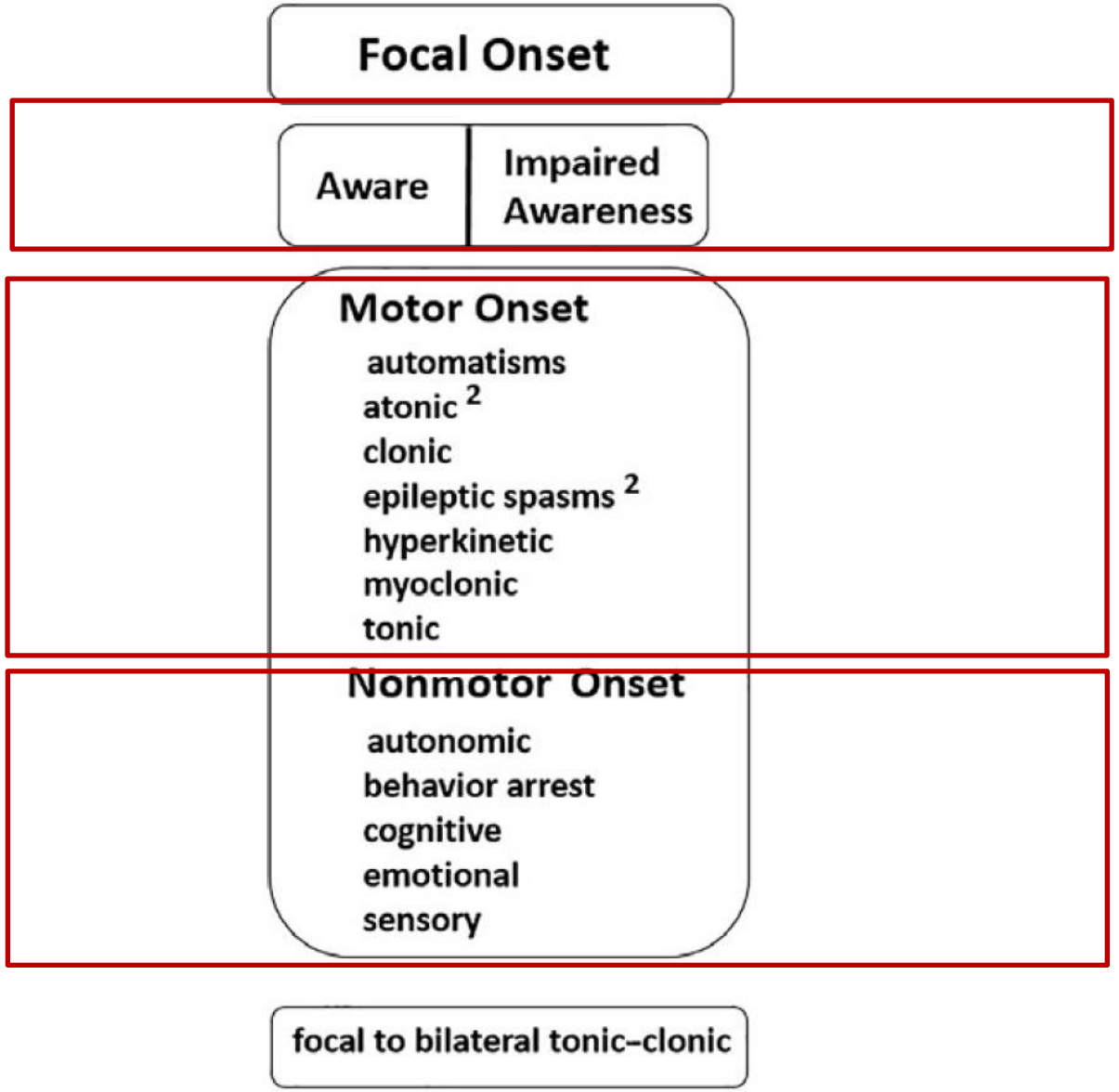
Seizure semiology

Neural activity



McGonigal A, et al. Onseizure semiology. Epilepsia 2021

ILAE 2017 Classification of Seizure Types Expanded Version [1]

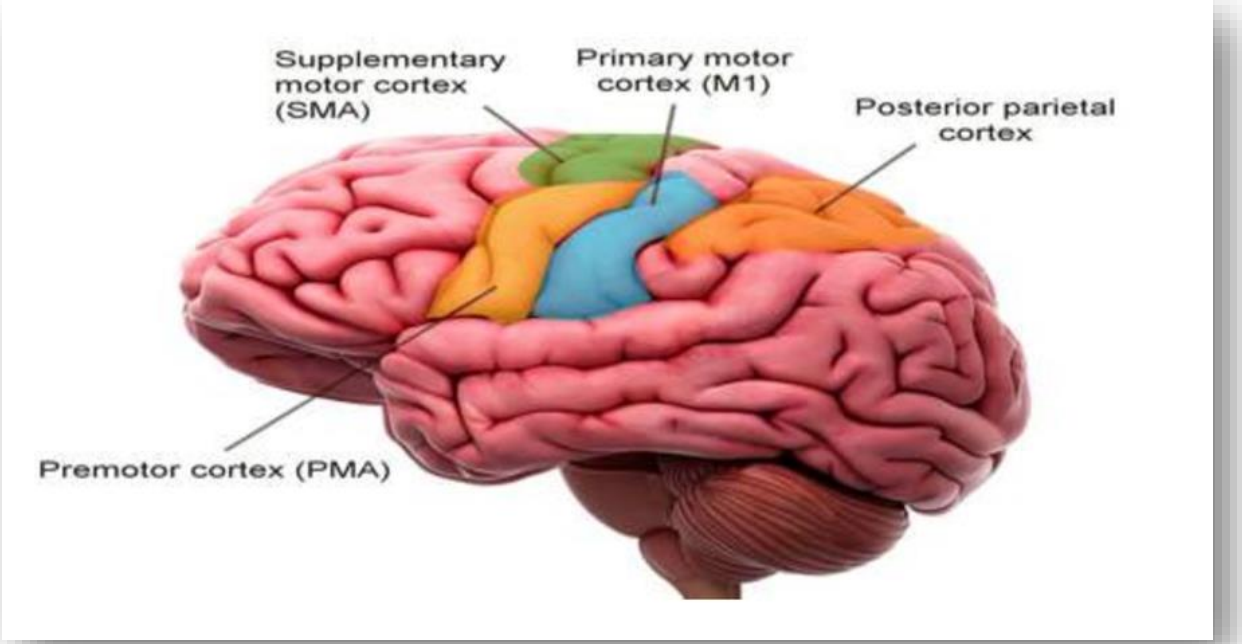


Case

*ใช้เพื่อการศึกษาเท่านั้น

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
- **Right arm clonic- indicate involvement of contralateral primary motor cortex**



Epilepsia partialis continua

focal motor status involving a small portion of the motor cortex

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Case

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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

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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

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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)

Case

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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)

Myoclonic seizure

Myoclonic seizure is a single or series of jerks

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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

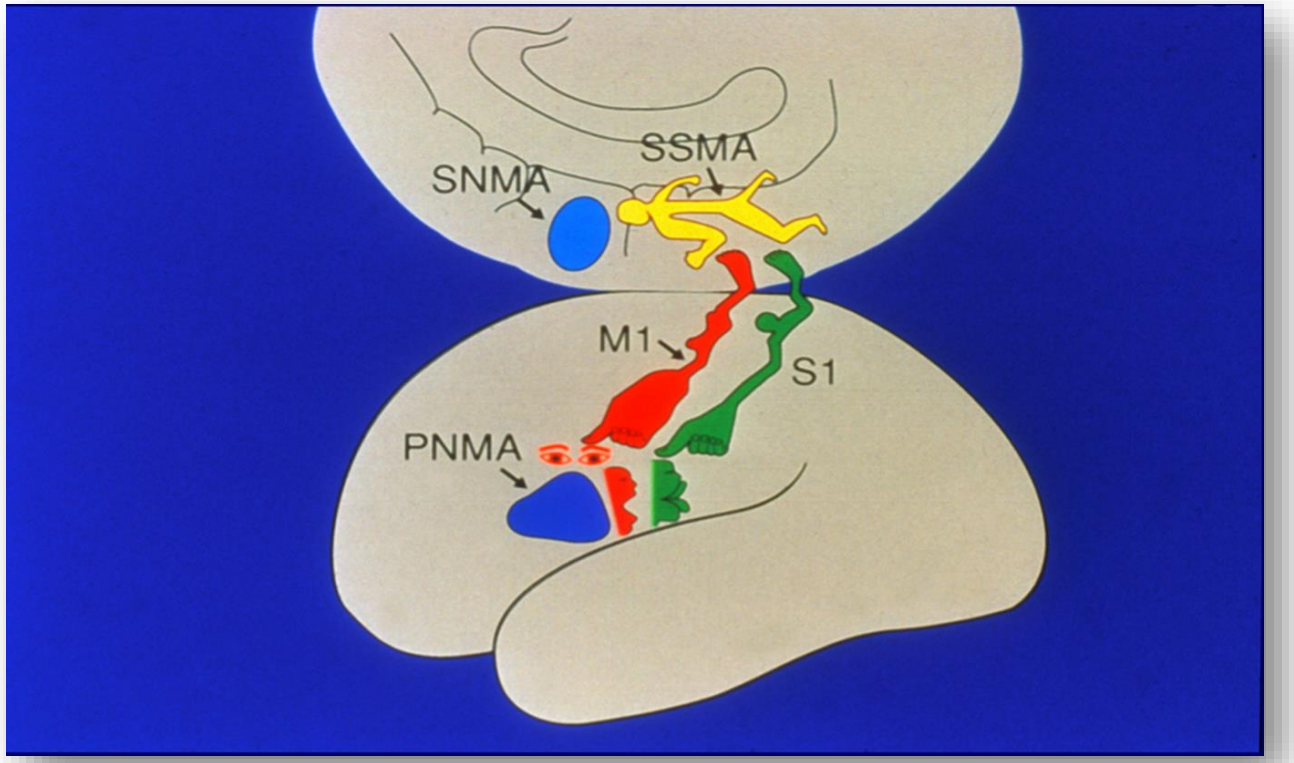
Asymmetrical tonic seizure

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

Asymmetrical tonic seizure

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

-> Hemisphere *contralateral* to extended arm



Location: Supplementary sensorimotor area (SSMA)

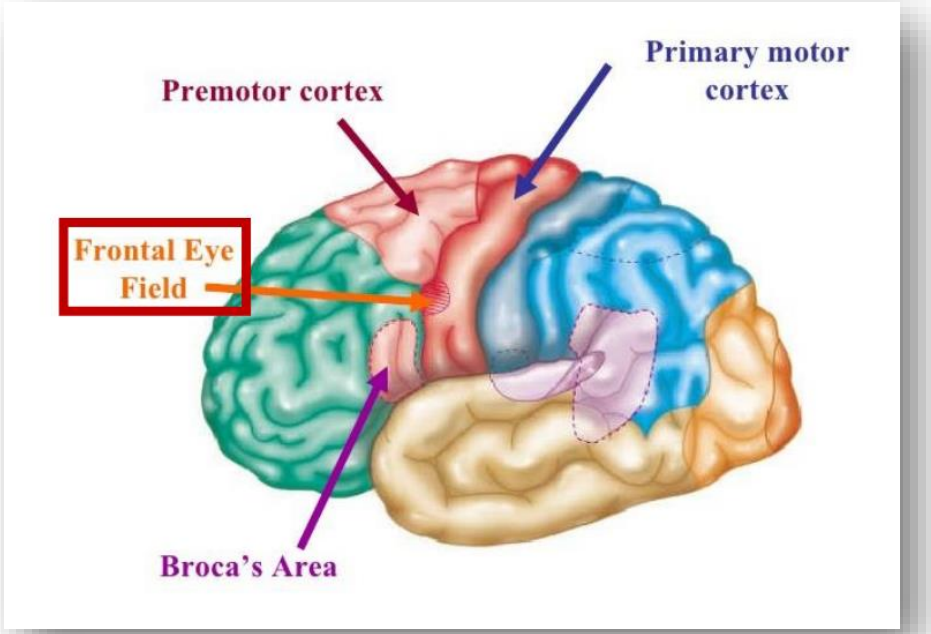
Tonic/dystonic seizure

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



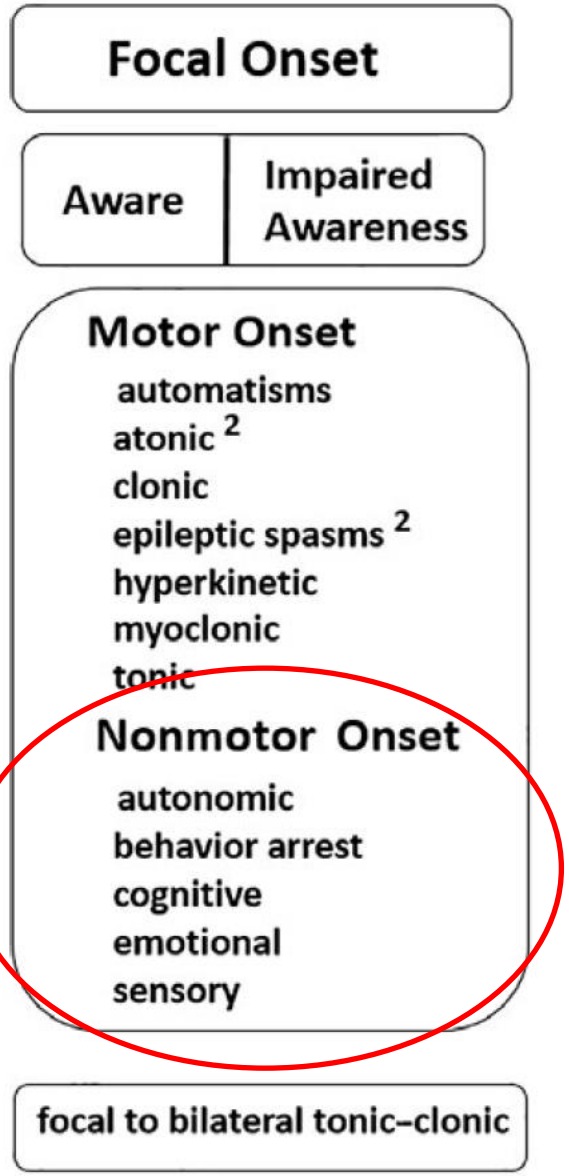
Common lateralising seizure manifestations

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]



semiology may be less well localizing

-> arising from associative cortex

- More wide-spread networks
- Complex dynamics

McGonigal, 2020

Knowing epileptic network helps

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

Ictal Fear

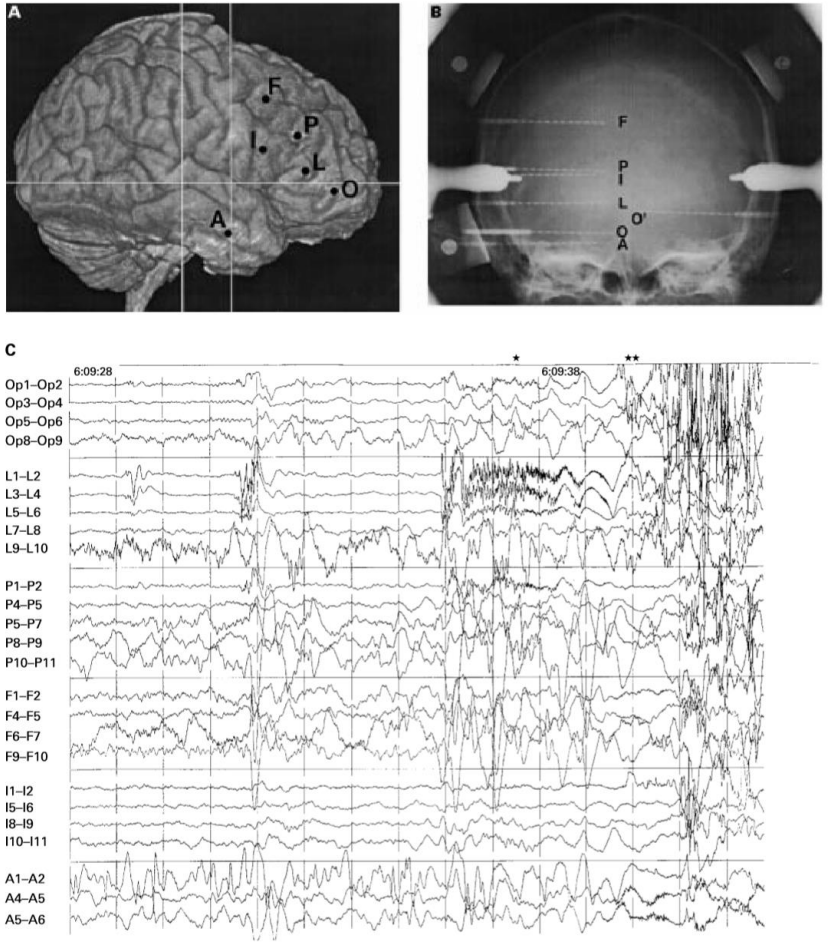
- Case a 10 years old female with epilepsy.
- During daytime, She **presents with fear** followed by screaming and tachycardia lasted 20 sec.
- During nighttime, she arose from sleep and looked scary followed by screaming and vigorous movements.

*ใช้เพื่อการศึกษาเท่านั้น

J Neurol Neurosurg Psychiatry 2001;**70**:186–191

Fear as the main feature of epileptic seizures

A Biraben, D Taussig, P Thomas, C Even, J P Vignal, J M Scarabin, P Chauvel



This limbic network involve- Orbitoprefrontal
- Anterior cingulate
- Temporal limbic cortices

Biraben et al, 2001

Gelastic seizure

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

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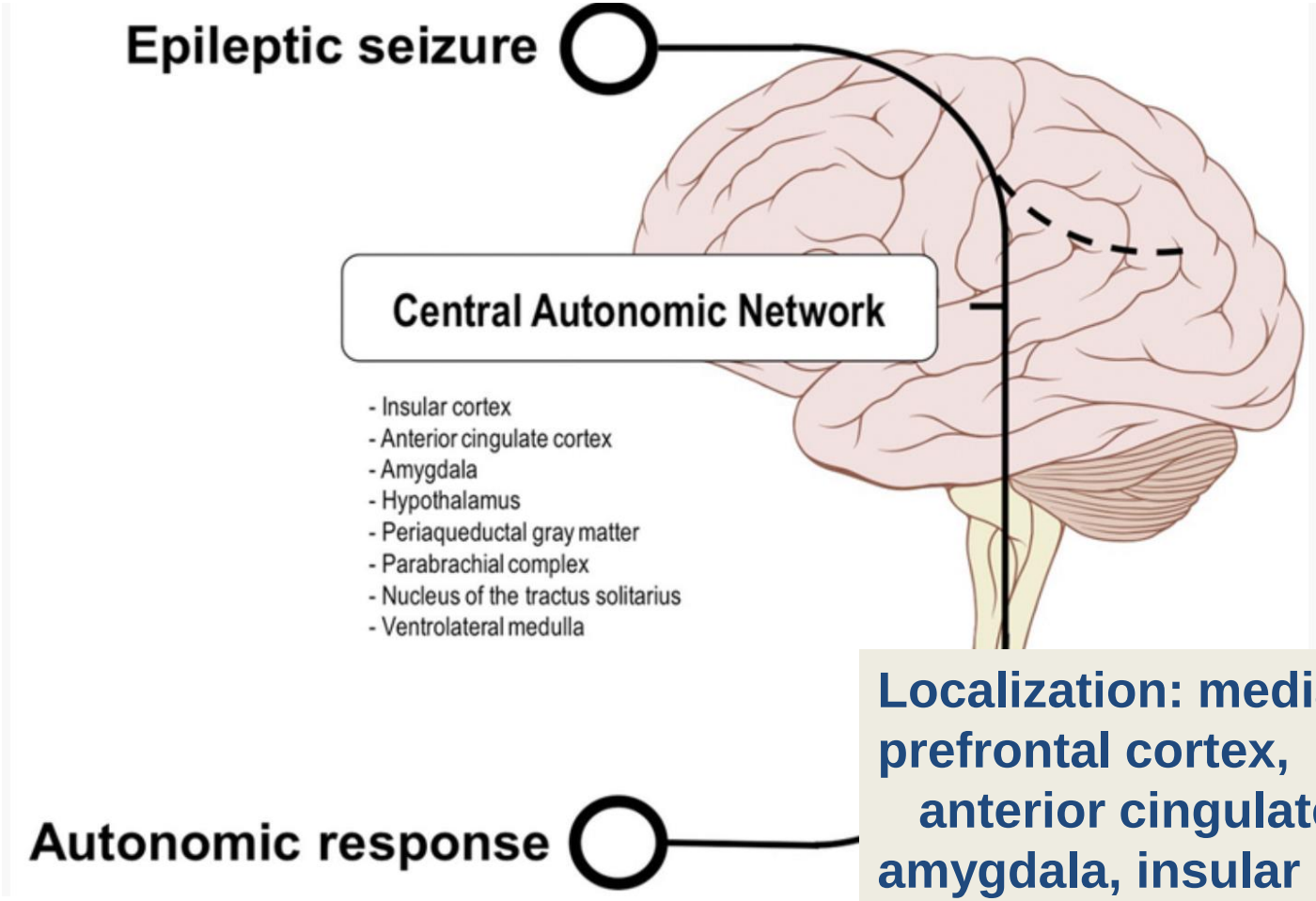
Gelastic seizure

- 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.

Focal autonomic seizures



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

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

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)

Take home points regarding semiology

- Analyze semiology in order is important- early signs more reliable
- Record sufficient number of seizures
- Look for consistency between seizures
- Identifying features in common is the key to categorization
- Think of epileptic networks could be involved according to electroclinical correlation!

Transparent language: use words that mean what they say

Thank you for your attention