



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

SEIZURE SEMIOLOGY

Piradee Suwanpakdee, MD

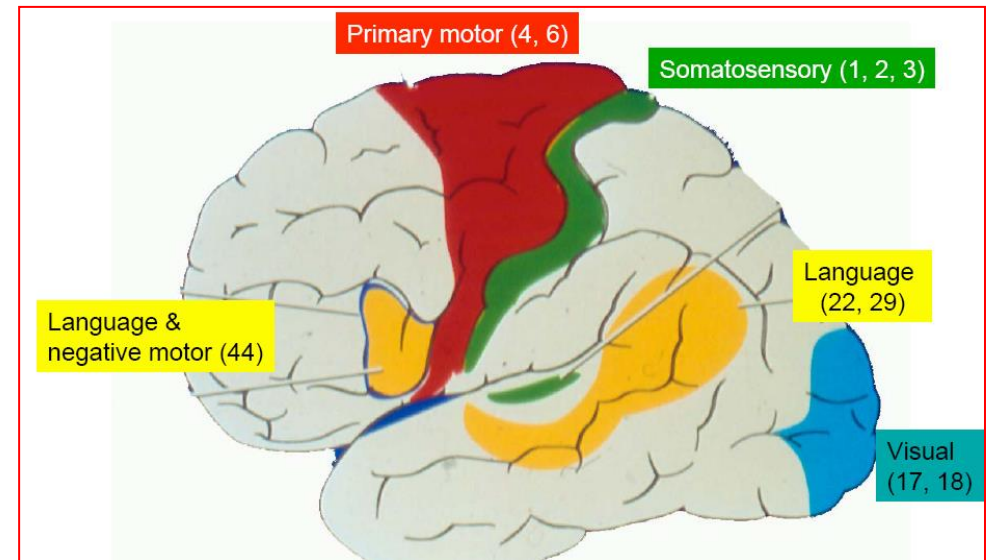
Division of Neurology

Department of Pediatrics

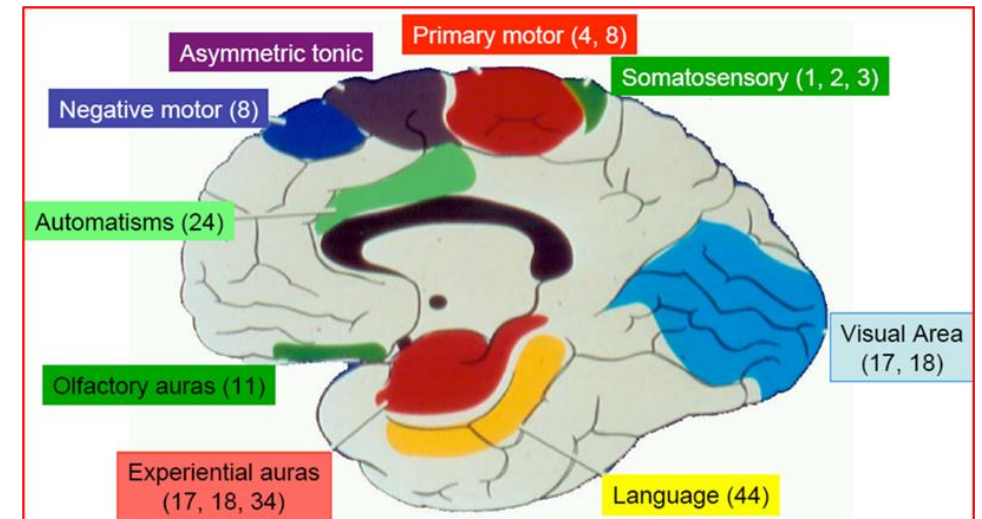
Phramongkutklao Hospital

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

Epileptic seizure semiology in different age groups

- Retrospective studied seizure semiology in all age groups (1 month - 90 years old), n = 270 patients
 - Group 1: >1 month to 3 years (n=36)
 - Group 2: > 3 years to 6 years (n=22)
 - Group 3: > 6 years to 10 years (n=33)
- (> 10 years old, the seizure semiology closely resembled of the adult population)
- Semiology is **related to age and cerebral maturation**

Phases of a seizure

Pre-Ictal (prodroma) →

includes the precipitating factors (fever, lack of sleep etc.) and prodromal symptoms (change in behavior, headache etc.);

Ictal (aura and progression) →

includes the ictal onset (aura, warning) and the ictal phase, which in case of focal seizures may have localizing characteristics;

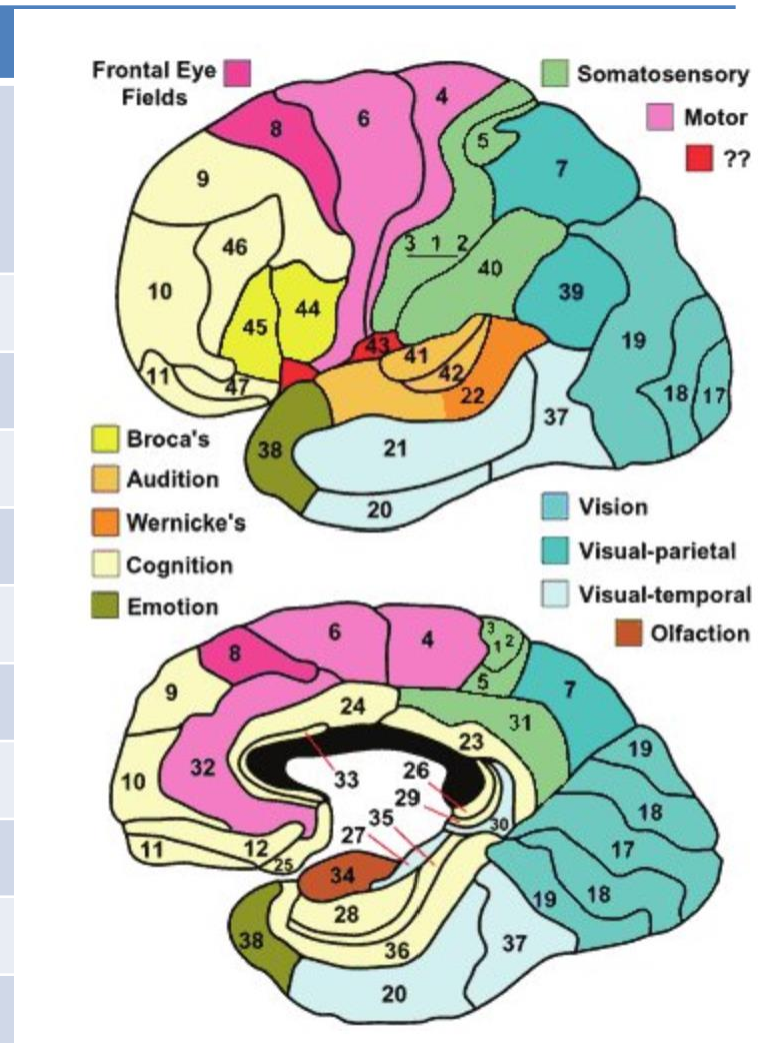
Post-Ictal

end of seizure usually more difficult to define than onset; may also demonstrate in focal seizures, localizing characteristics

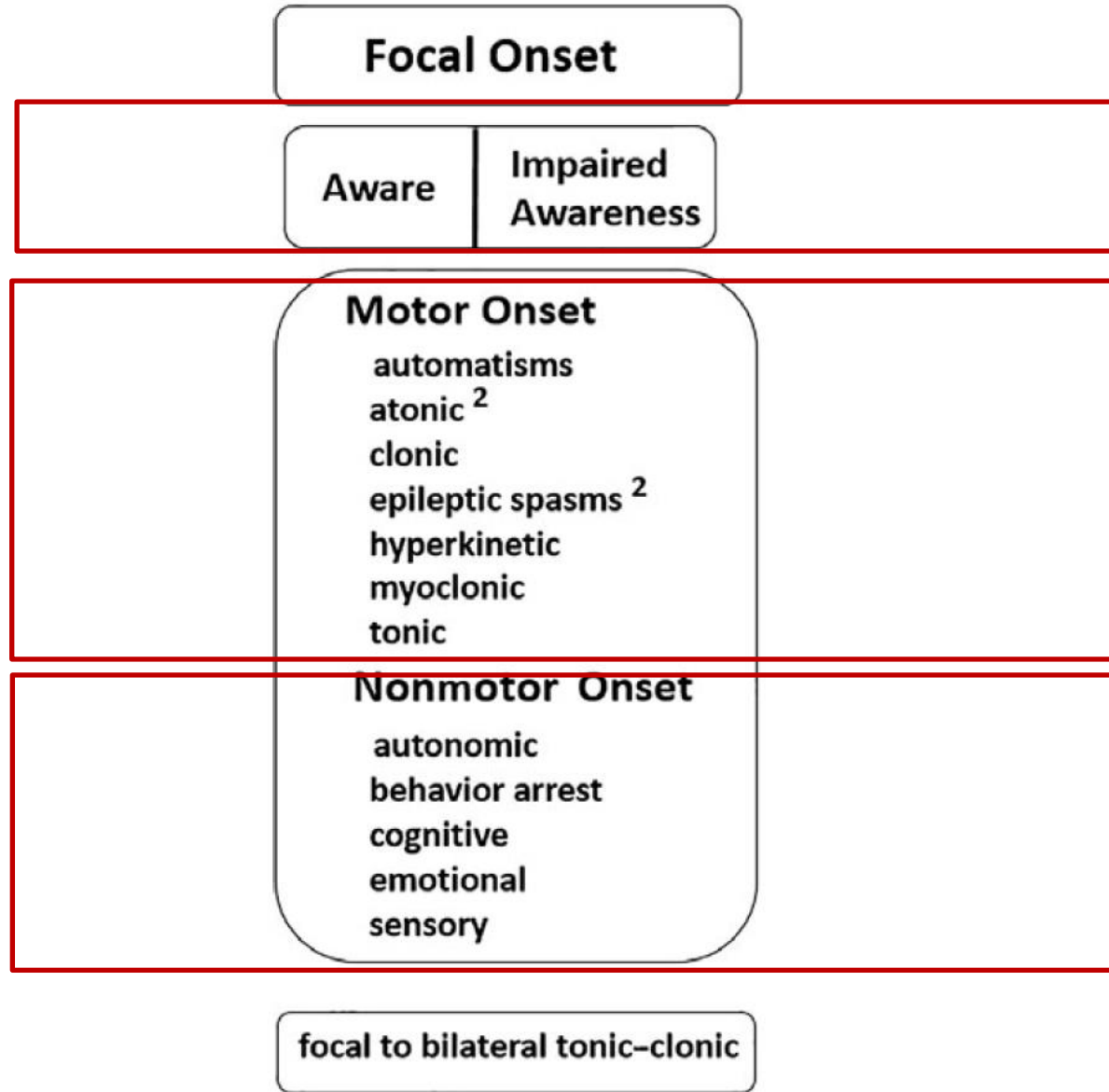
Aura

is the first ictal symptoms, auras can provide important localizing value

Type of aura	Symptomatogenic zone
Somatosensory	Primary somatosensory cortex (areas 1,2, and 3b) Secondary somatosensory areas (parietal operculum/SSII) SSMA
Simple visual	Primary visual cortex (areas 17, 18, and 19)
Complex visual	Temporo-occipital junction and basal temporal cortex
Simple auditory	Primary auditory cortex (area 41)
Complex auditory	Auditory association cortex (areas 42 and 22)
Vertiginous	Temporo-occipital junction
Olfactory	Orbitofrontal region, amygdala, and insula
Gustatory	Parietal operculum and basal temporal cortex
Autonomic	Insula, amygdala, anterior cingulum, and SSMA
Fear	Amygdala, hippocampus, and mesial frontal lobe
Déjà vu/jamais vu	Uncus, entorhinal cortex, and temporal neocortex
Cephalic/whole body	Amygdala, entorhinal cortex, and temporal neocortex/SSII and SSMA

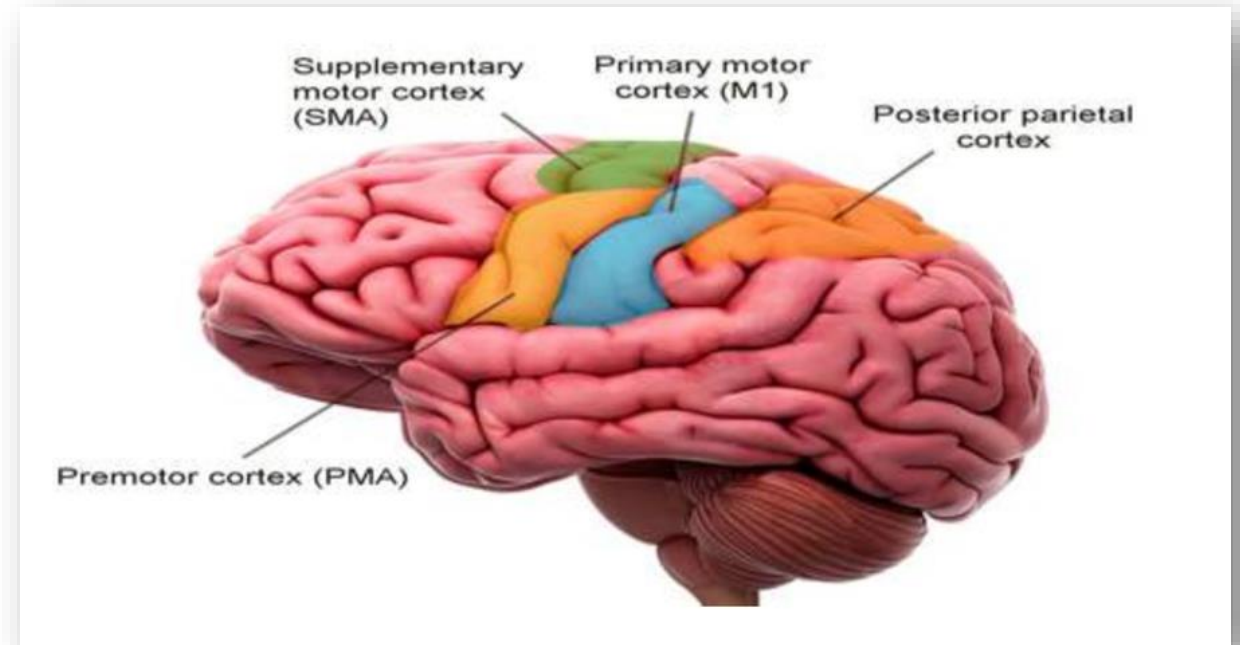


ILAE 2017 Classification of Seizure Types Expanded Version ¹



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 (EPC)

- Spontaneous regular or irregular clonic muscular twitching affecting a limited part of the body, occurring for a minimum of one hour, and recurring at intervals of no more than ten seconds¹
- Localization: involving a small portion of the contralat. **sensorimotor cortex**
- The pathologies that underlie EPC are heterogeneous
- The main diagnoses of EPC in children²:
 - Rasmussen's encephalitis
 - Mitochondrial disease
 - MRI lesion-positive focal epilepsy
 - MRI lesion-negative EPC (inflammatory, neurometabolic, genetic)

¹Bien CG et al. Epileptic Disord. 2008

²Surana et al. Epilepsia 2020

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

- 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
- Pathophysiology:
 - generalized seizures (LGS) resulting from a sudden cortically-mediated activation of inhibitory brain stem centers via fast corticoreticulospinal tracts

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)

Focal hyperkinetic seizure

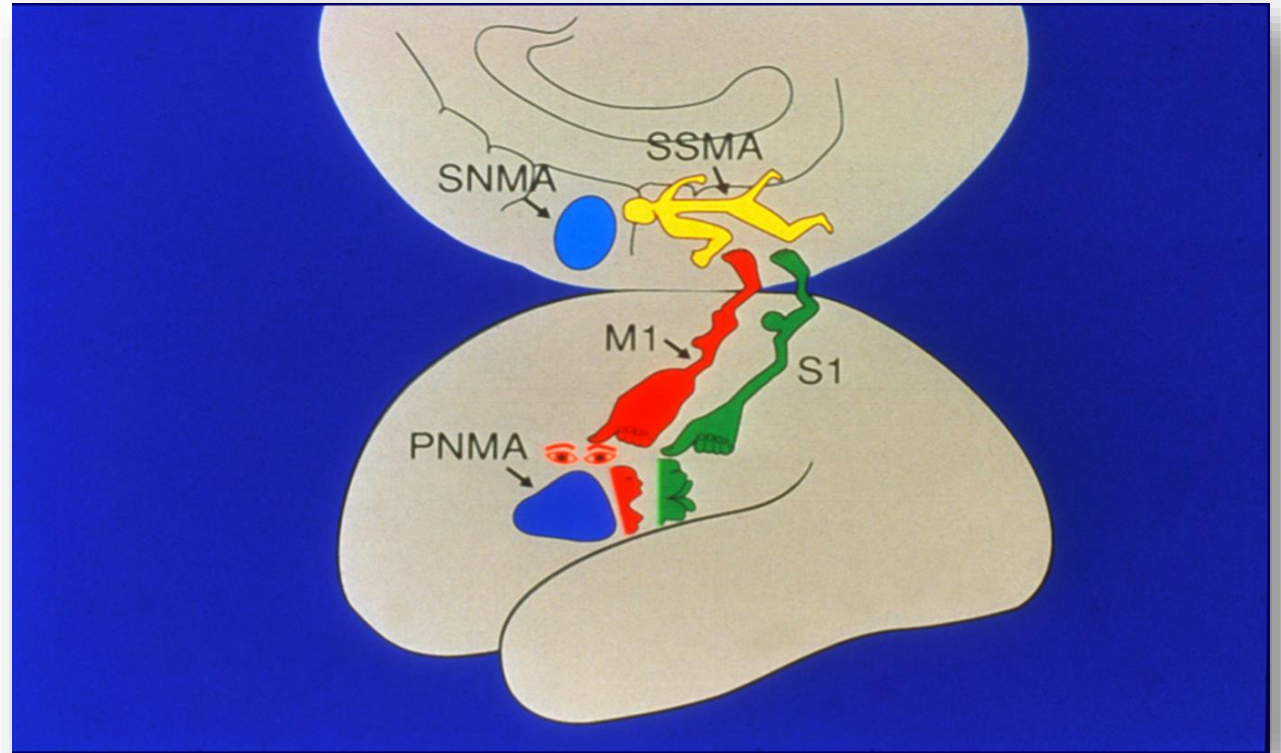
- This seizure type involves movements of proximal limb or axial muscles, producing irregular large amplitude movements, such as pedaling, pelvic thrusting, jumping, thrashing and/or rocking movements
- Consciousness may be preserved
- Occur mostly during sleep
- Pathophysiology:
 - Primarily an expression 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

- 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

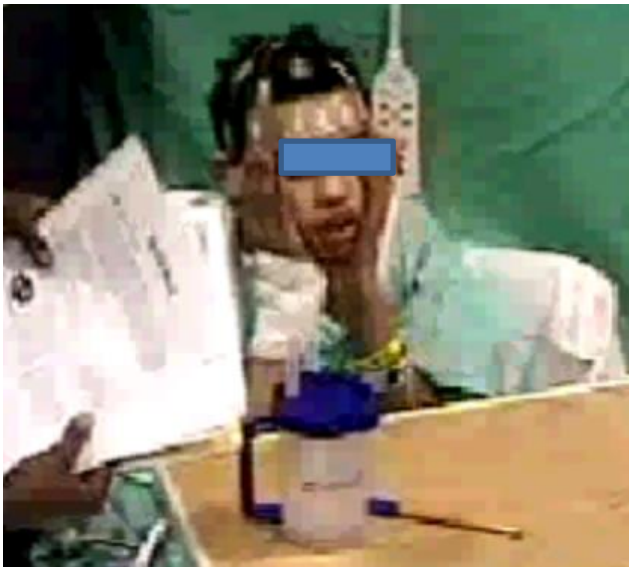
- Preferentially affect proximal muscle both sides, but more prominent over the contralateral side
- Conscious is intact in most patients
- Asymmetric tonic limb posturing “sign of four”
 - > Hemisphere *contralateral* to extended arm



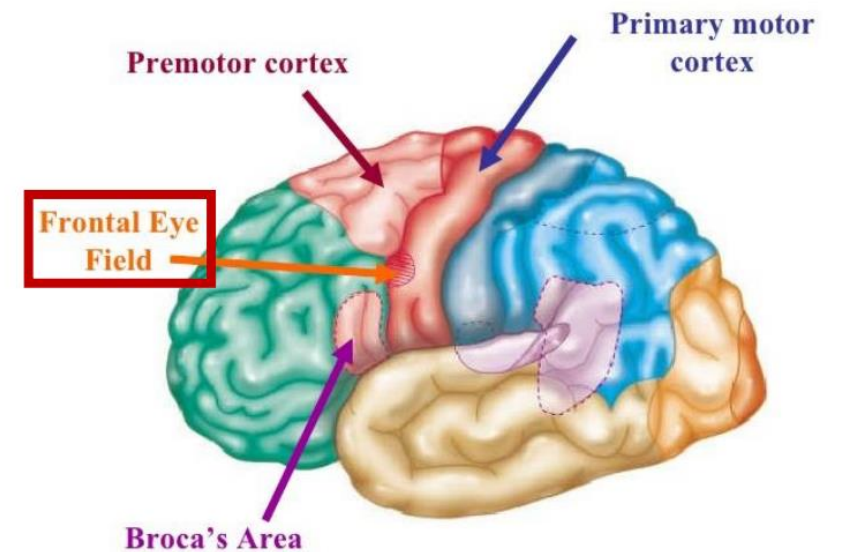
Location: Supplementary sensorimotor area (SSMA)

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



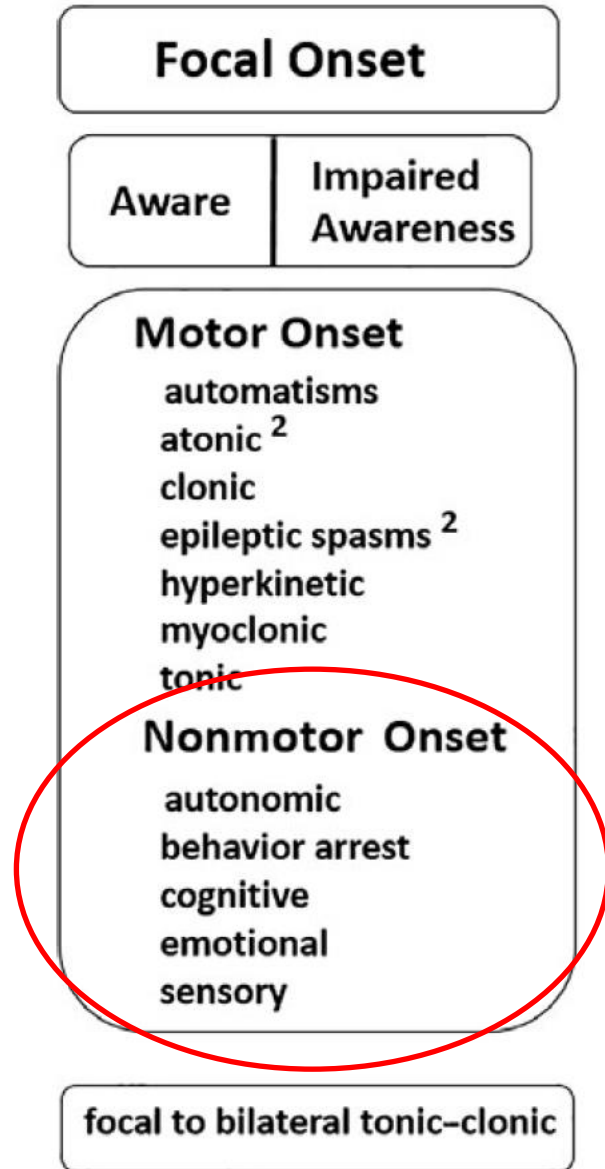
Common lateralising seizure manifestations

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

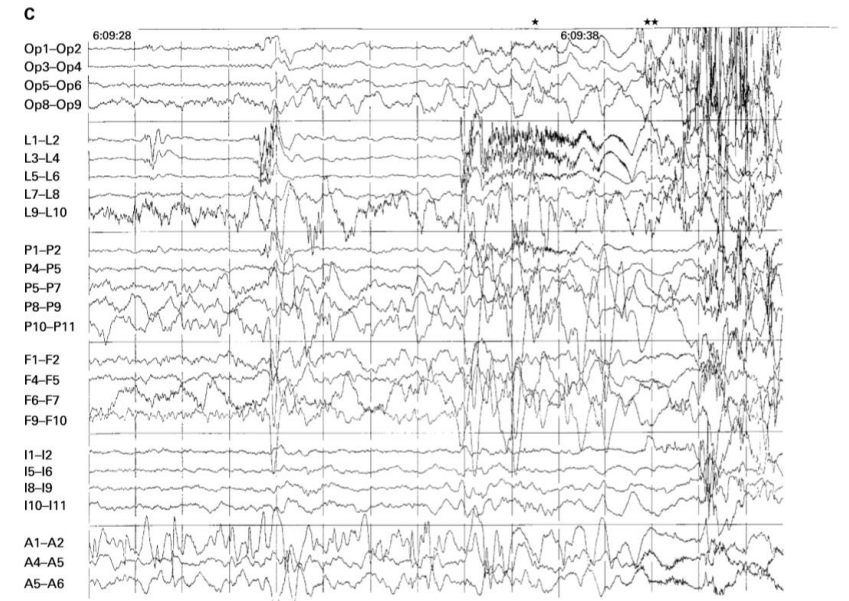
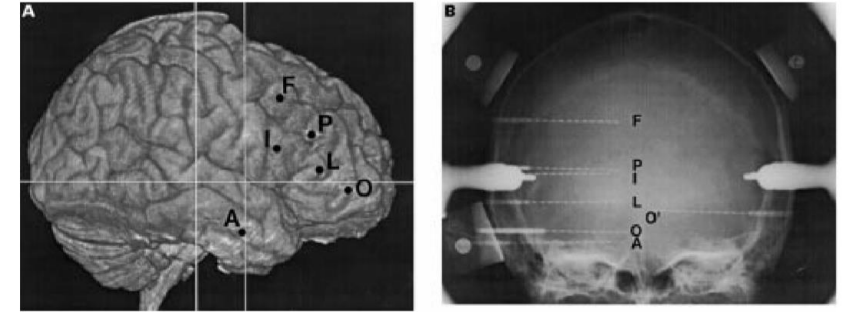
Fear as the main feature of epileptic seizures

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

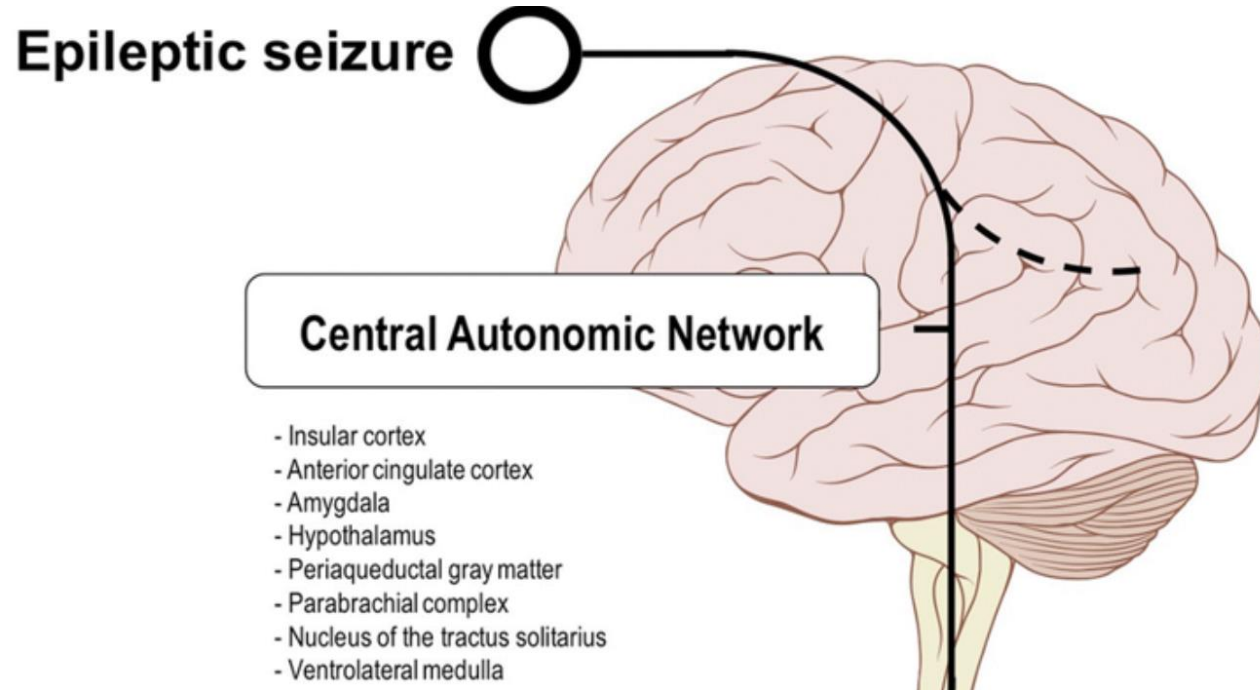


Gelastic seizure



- 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



Autonomic response

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

“Chapeau de gendarme” or Ictal pouting

Anterior cingulate region plays the important role



Fig. 2 Facial expression of patients with ictal pouting (IP). Open eyes in patients 1, 3, 4, 5, and 6 give them a frightened or menacing air, whereas patients 2 and 7 seem disgusted. Patients 8, 9, 10 and 11 have closed eyes, given them 9, and 10 have clos...

Clinical Manifestations of Insular Lobe Seizures



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

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