



Phramongkutklao Comprehensive  
Pediatric Epilepsy Center of Excellence

*Integration • Passion • Wisdom*

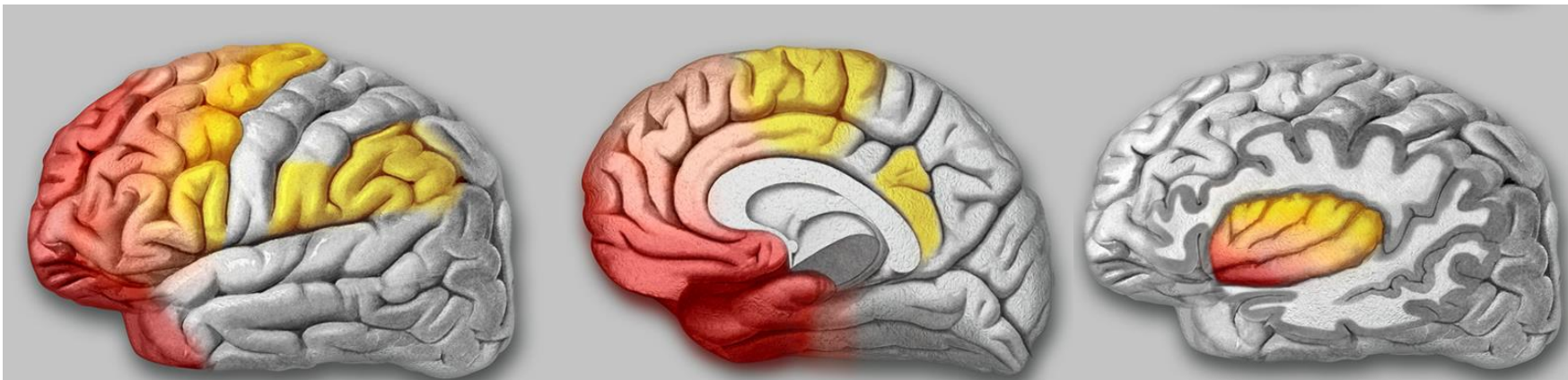
# Seizure semiology

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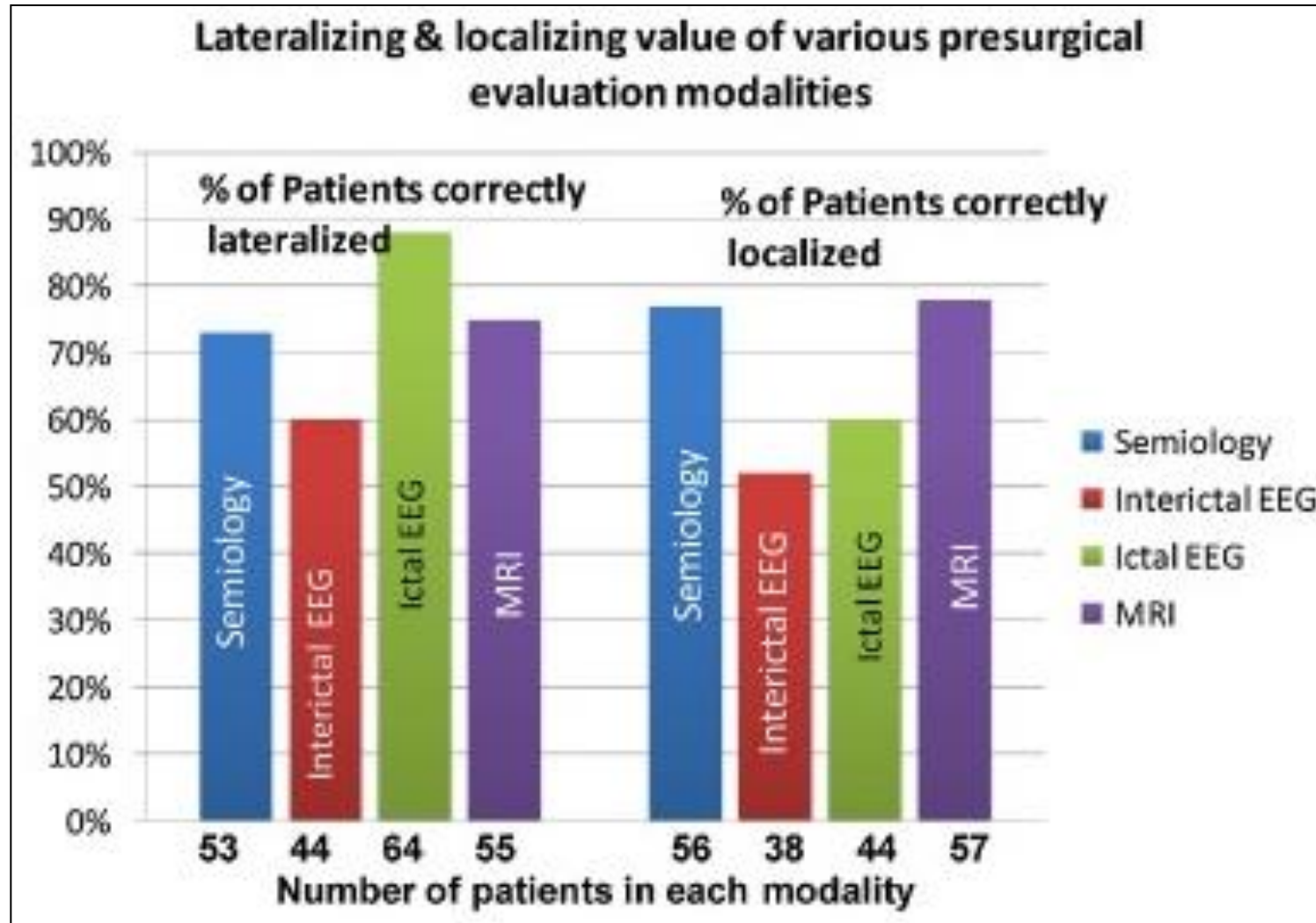
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# Ictal semiology

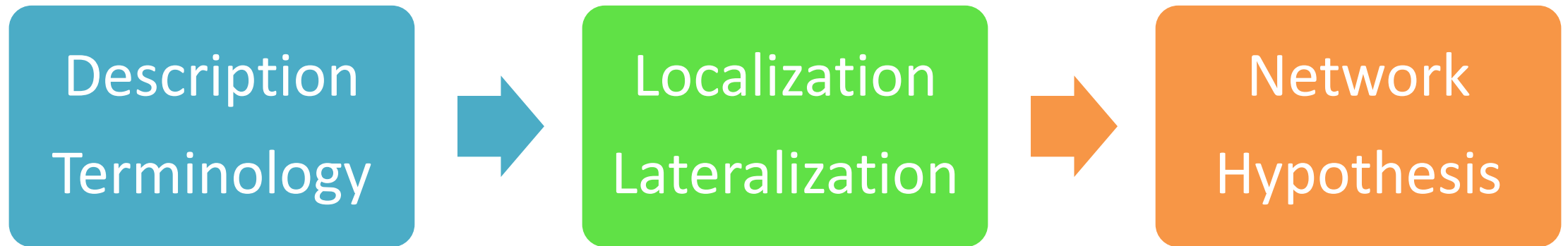
- **Semiology or Semiotics** : “The study of signs and sign-using behaviour”
- The semiology of epileptic seizures reflects activation, or dysfunction, of areas of brain (often termed the **symptomatogenic zone**)
- A simple and cost-effective tool that allows localization of the symptomatogenic zone which either **overlaps** or **close** proximity of “**the epileptogenic zone**”



# The important of ictal semiology



# Ictal Semiology approach



# Description and Terminology

<b>Symptoms</b>	<b>Medical Term</b>
<b>automatic behaviors</b>	automatisms
<b>emotions or appearance of emotions</b>	emotions
<b>extension or flexion postures</b>	tonic
<b>flushing/sweating/piloerection</b>	autonomic
<b>jerking arrhythmically</b>	myoclonus
<b>jerking rhythmically</b>	clonus
<b>language or thinking problems, deja vu</b>	cognitive
<b>lid jerks</b>	eyelid myoclonia
<b>limp</b>	atonic
<b>numb/tingling, sounds, smells, tastes visions, vertigo</b>	sensations
<b>pausing, freezing, activity arrest</b>	behavior arrest
<b>thrashing/pedaling</b>	hyperkinetic
<b>trunk flexion</b>	spasm

# ILAE 2017 Classification of Seizure Types Expanded Version <sup>1</sup>

## Focal Onset

Aware

Impaired  
Awareness

### Motor Onset

automatisms  
atonic <sup>2</sup>  
clonic  
epileptic spasms <sup>2</sup>  
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

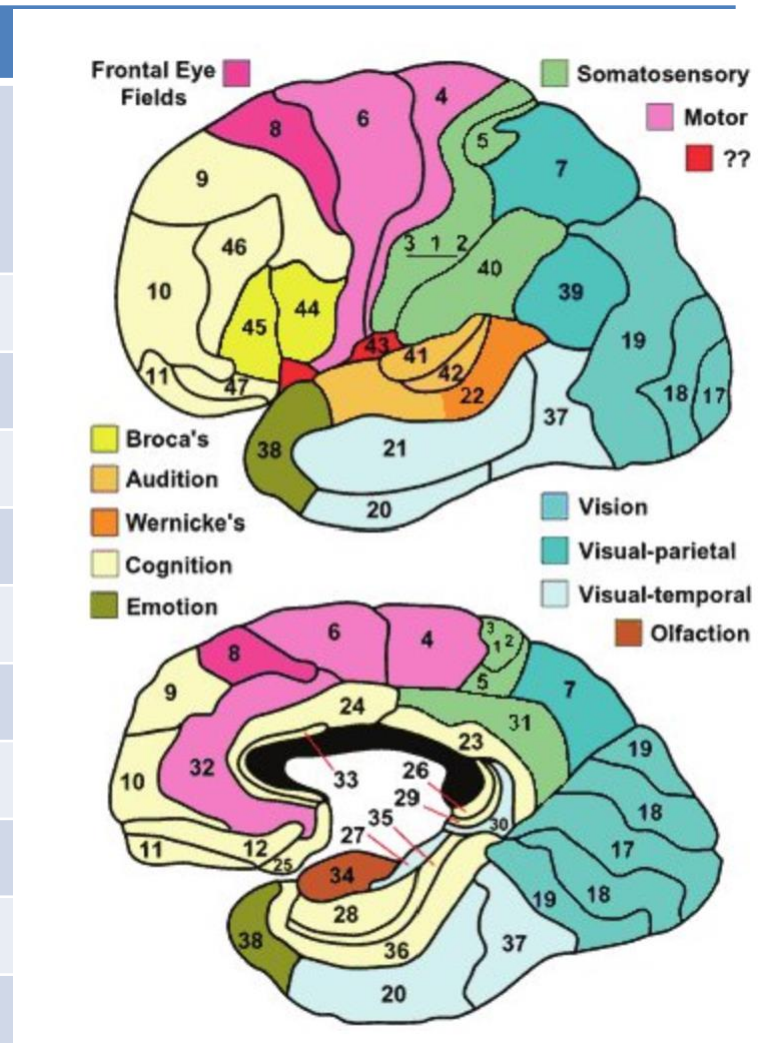
behavior arrest

## Unclassified <sup>3</sup>

# Aura

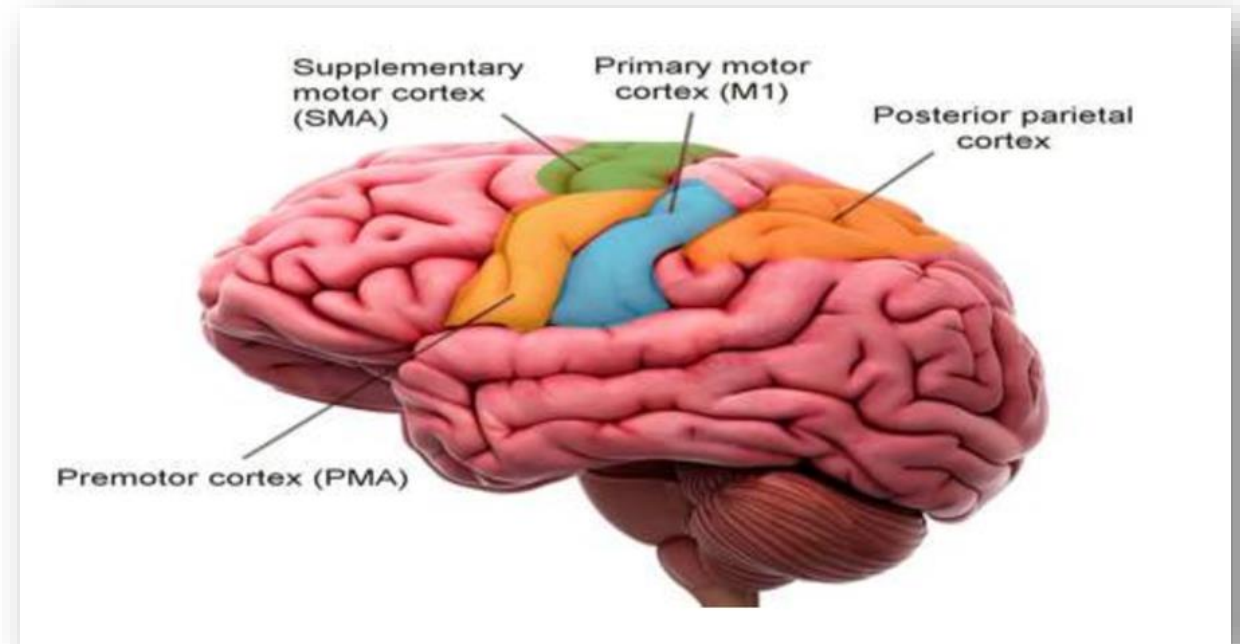
is the first ictal symptoms, auras can provide important localizing value

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



# 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 seconds<sup>1</sup>
- Localization: involving a small portion of the contralat. **sensorimotor cortex**
- The pathologies that underlie EPC are heterogeneous
- The main diagnoses of EPC in children<sup>2</sup>:
  - **Rasmussen's encephalitis**
  - Mitochondrial disease
  - MRI lesion-positive focal epilepsy
  - MRI lesion-negative EPC (inflammatory, neurometabolic, genetic)

<sup>1</sup>Bien CG et al. Epileptic Disord. 2008

<sup>2</sup>Surana 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

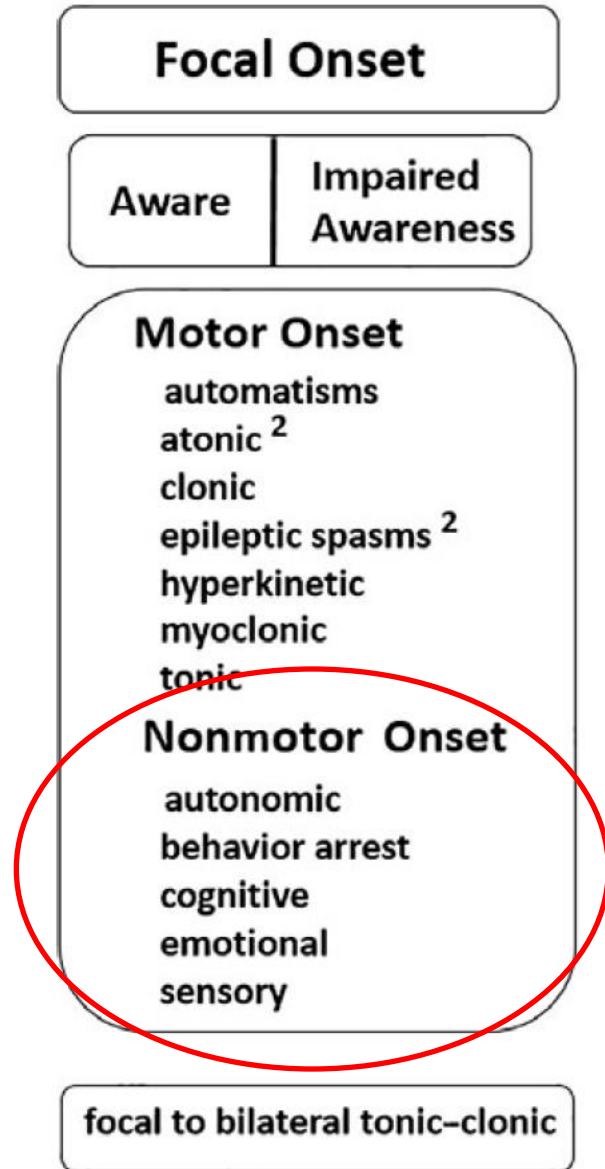
- Sudden muscle jerks of variable topography (distal, proximal, axial): uni- or bilateral, focal, multifocal or generalised
- Prominently affecting shoulders and proximal arms
- Consciousness likely preserved
- 100-400 msec in duration
- Unilateral myoclonic seizures -> contralateral primary motor area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

# Common lateralizing signs of focal seizures

Subtype	Symptomatogenic zone	Lateralization
Dystonic limb posturing	Activation of basal ganglia	Contralateral
Tonic posturing	Activation of SSMA, basal ganglia, cingulum, and primary motor cortex	Contralateral
Eye version	Frontal eye fields (area 8) and extrastriate cortex (area 19)	Contralateral
Head version	Premotor area and Frontal eye fields (areas 6 and 8)	Contralateral
Asymmetric tonic limb posturing	SSMA and precentral area	Contralateral



# ILAE 2017 Classification of Seizure Types Expanded Version <sup>1</sup>



**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

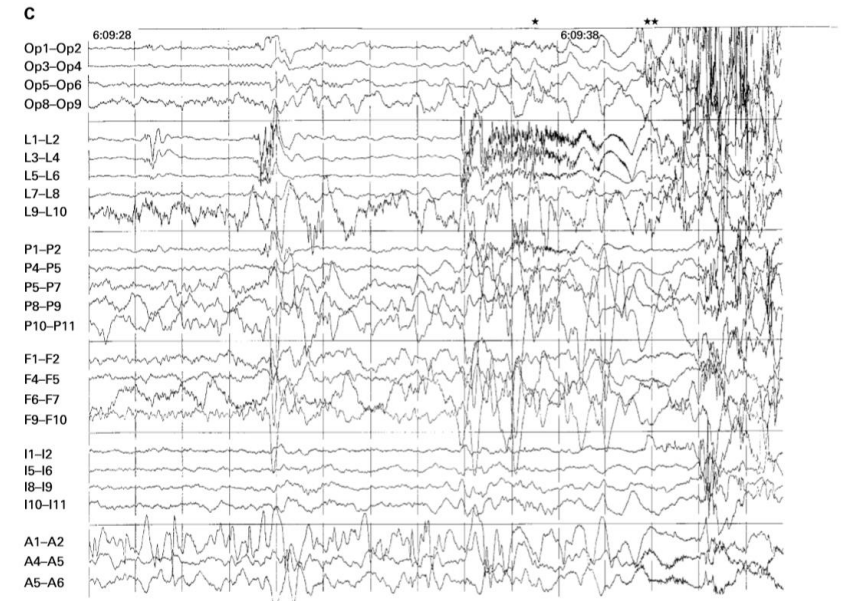
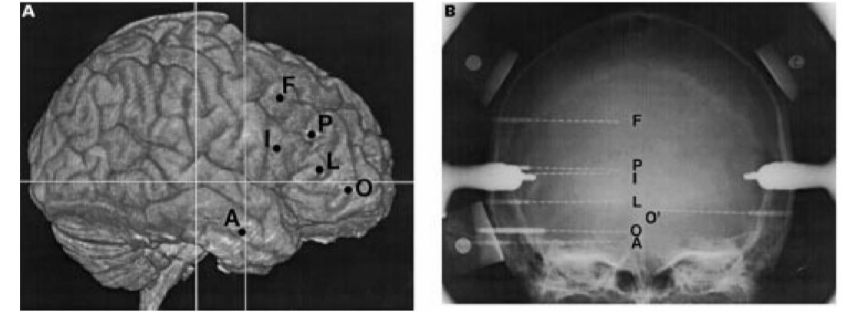
- Characterized by alterations in mood or emotion, or the appearance of altered emotion without the subjective emotion, at seizure onset
- Described as:
  - Focal emotional seizure with fear/anxiety/panic
  - Focal emotional seizure with laughing (gelastic)
  - Focal emotional seizure with crying (dacrystic)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

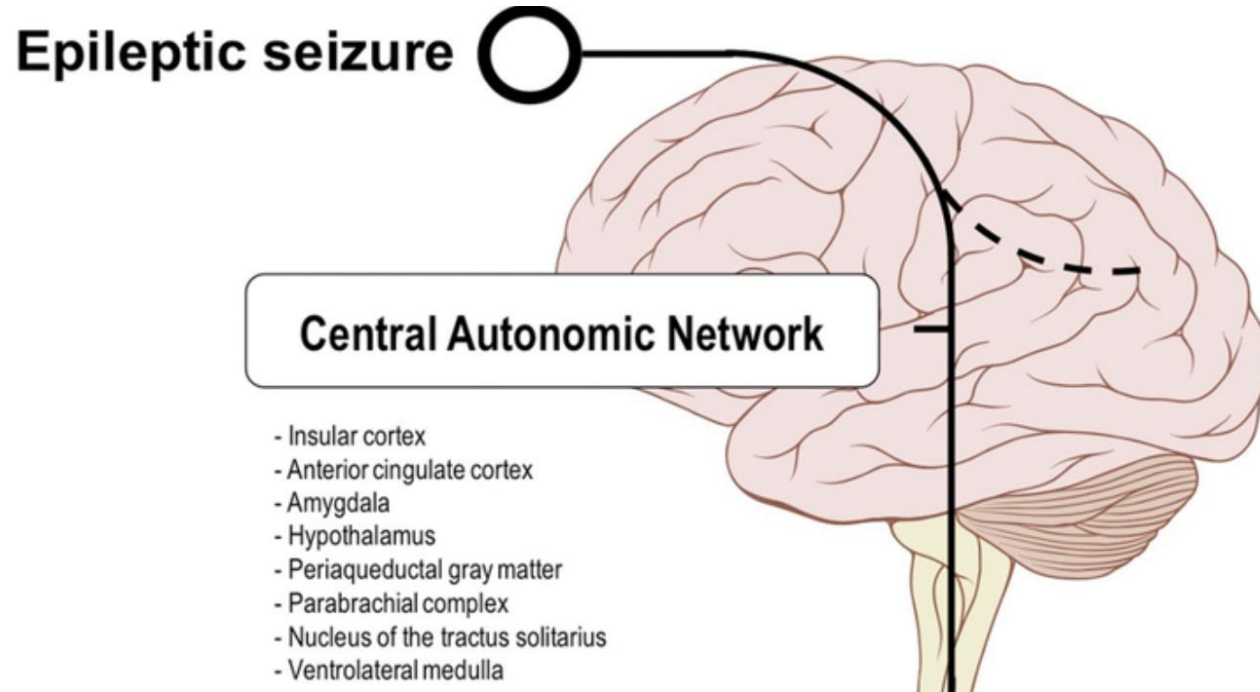


# Gelastic seizure



- 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



# Summary

- The elements of ictal semiology strongly suggests the seizure onset and propagation
- Carefully analyze semiology step by step from the initial symptom/sign until the end
  - Describe the event
  - Terminology
  - Localization/ Lateralization
  - Correlate with other investigations (EEG, MRI)
- Then you will understand epilepsy the underlying epileptic network

**Transparent language: use words that mean what they say**



***Thank you for your attention***