FOCAL EPILEPSY AND SEIZURE SEMIOLOGY

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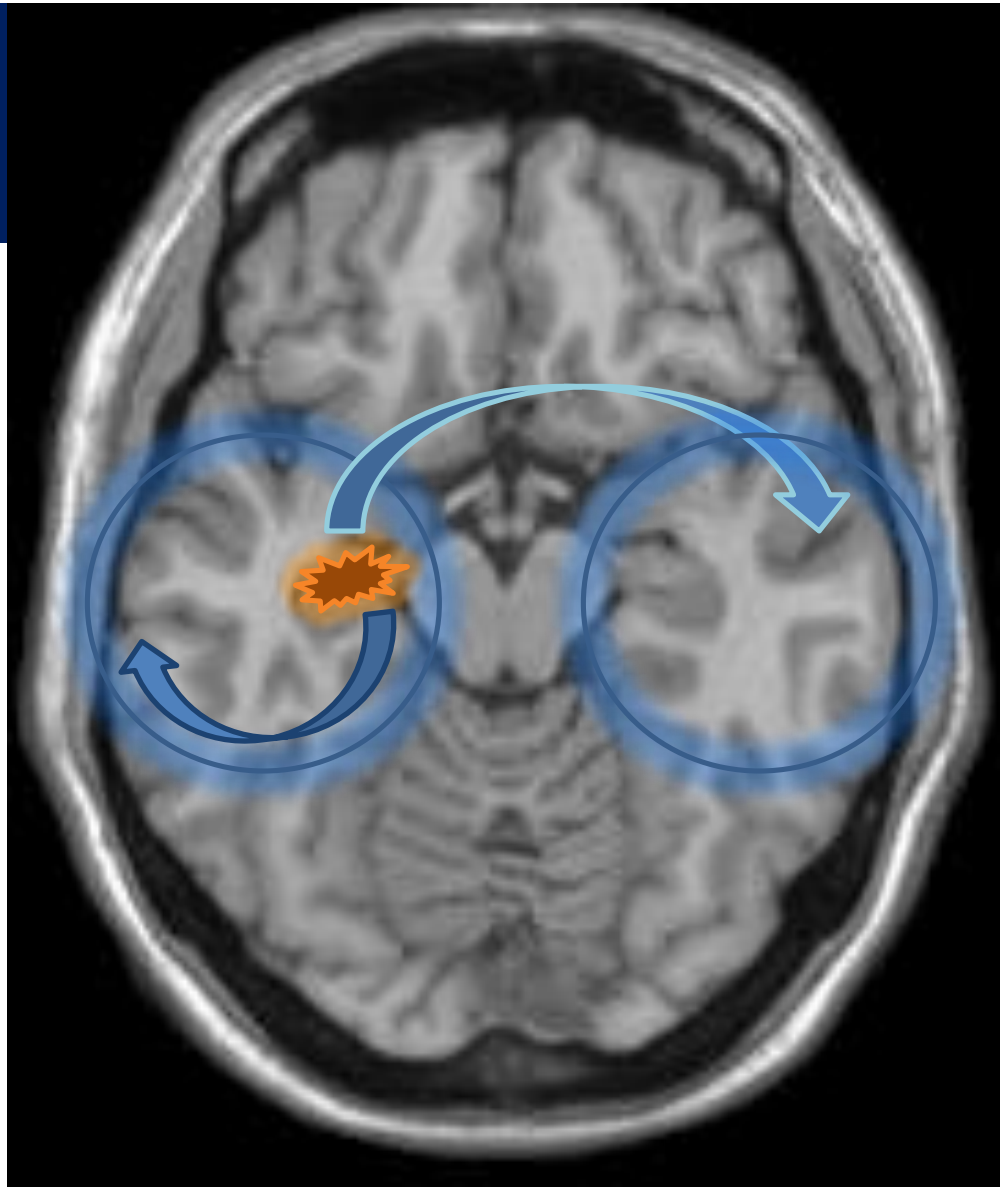
OUTLINE

- **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

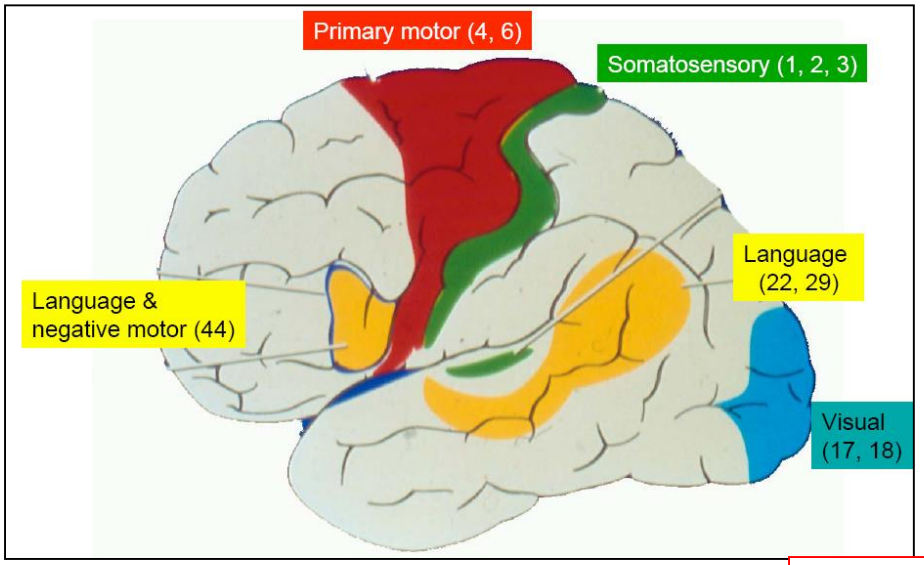
From *Fisher et al. Instruction manual for the ILAE 2017 operational classification of seizure types. Epilepsia doi: 10.1111/epi.13671*

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**

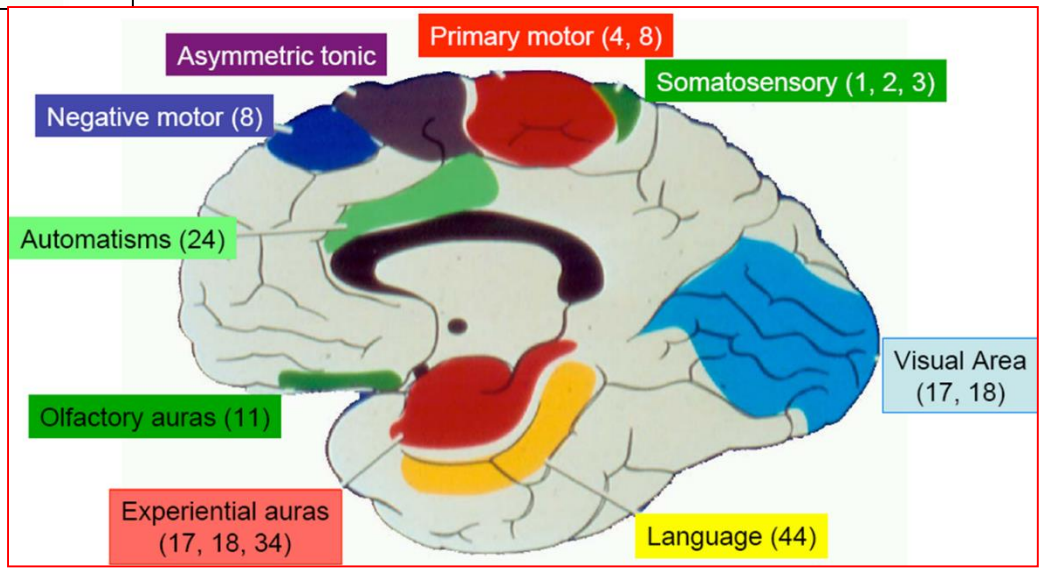
Symptomatogenic areas



Left hemisphere

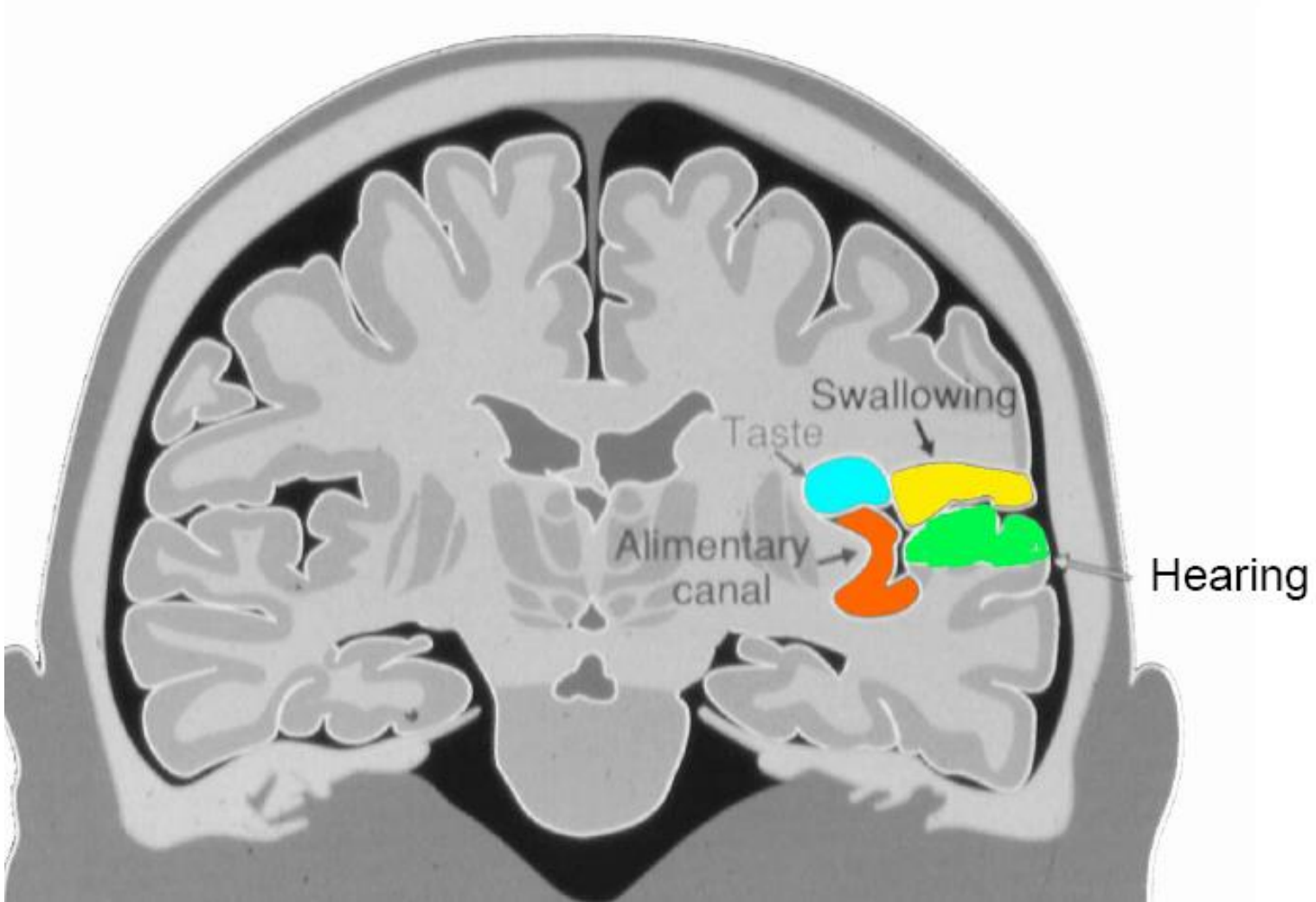
lateral aspect

Mesial aspect



Symptomatogenic areas

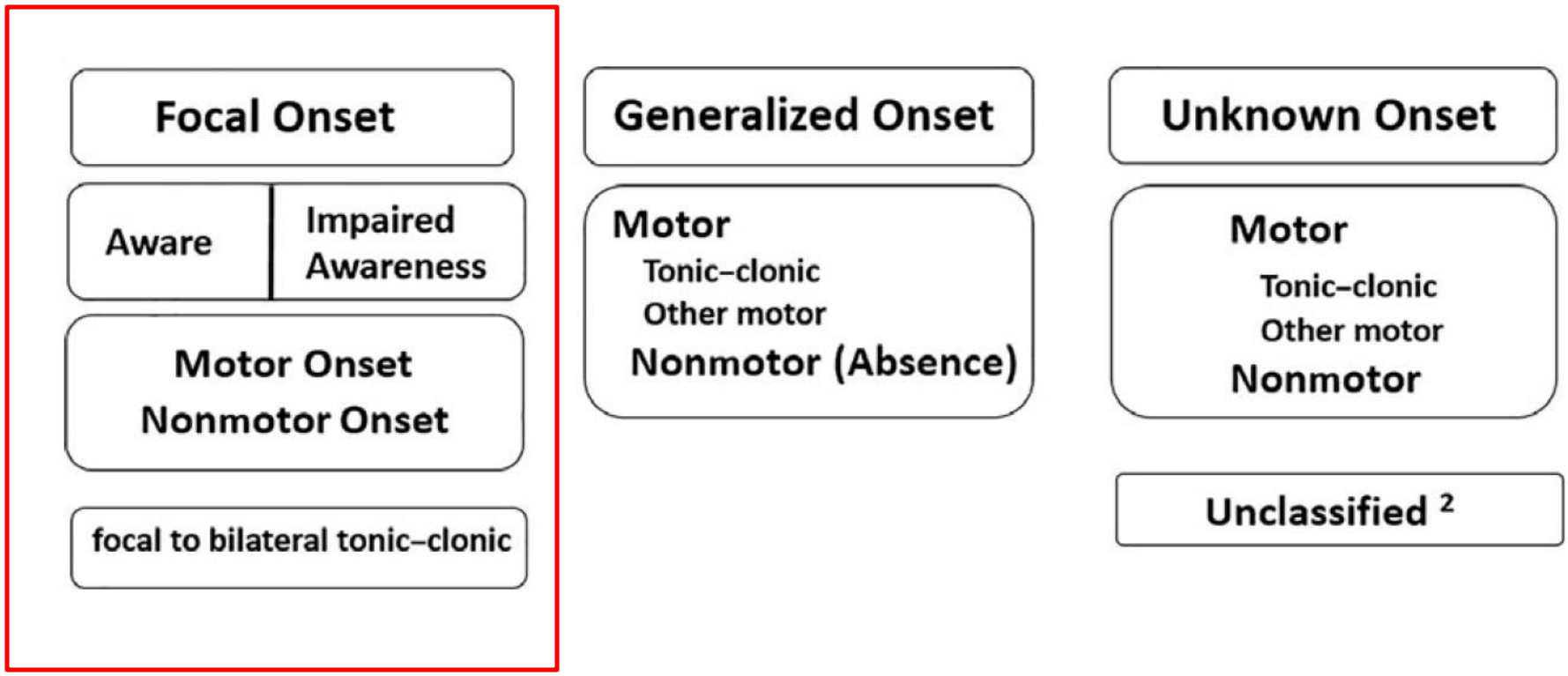
Left Insula



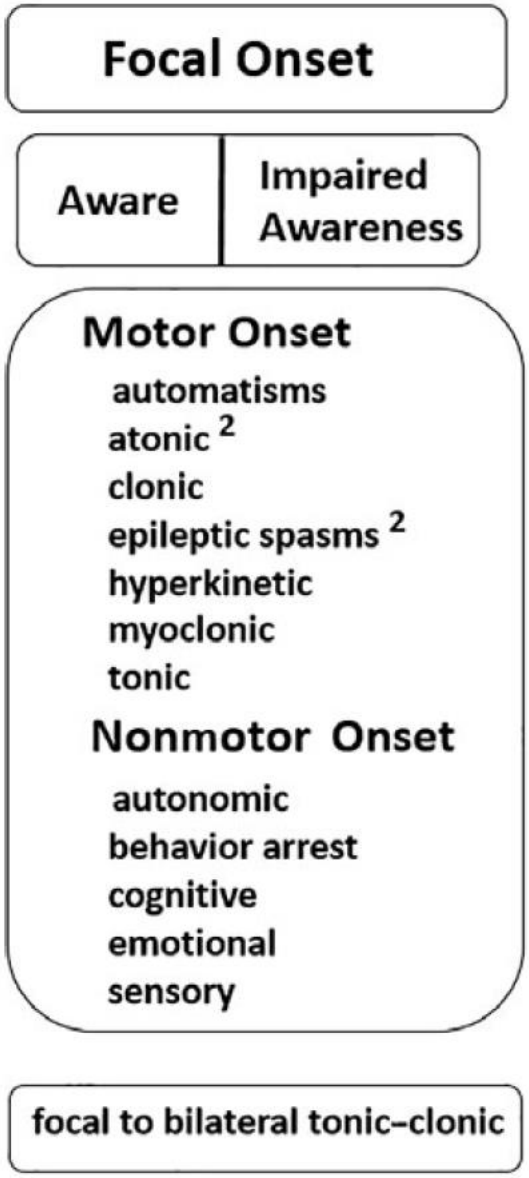
How can we get the elements of seizure semiology?

- **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

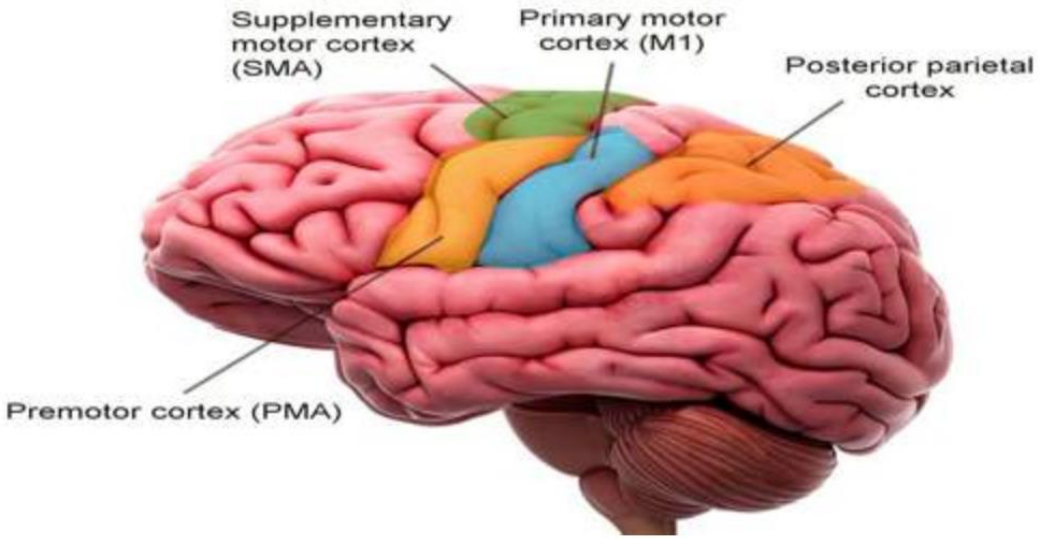
- Atonic seizures and epileptic spasms would *not* have level of awareness specified
- Pedalling grouped in hyperkinetic rather than automatisms (arbitrary)
- Cognitive seizures
  - impaired language
  - other cognitive domains
  - positive features eg déjà vu, hallucinations, perceptual distortions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

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

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

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)

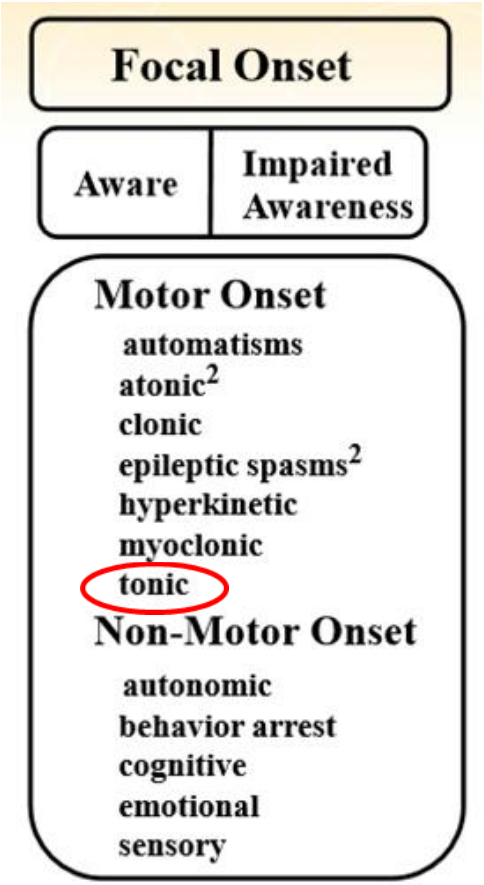
Focal hyperkinetic seizure

- 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



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

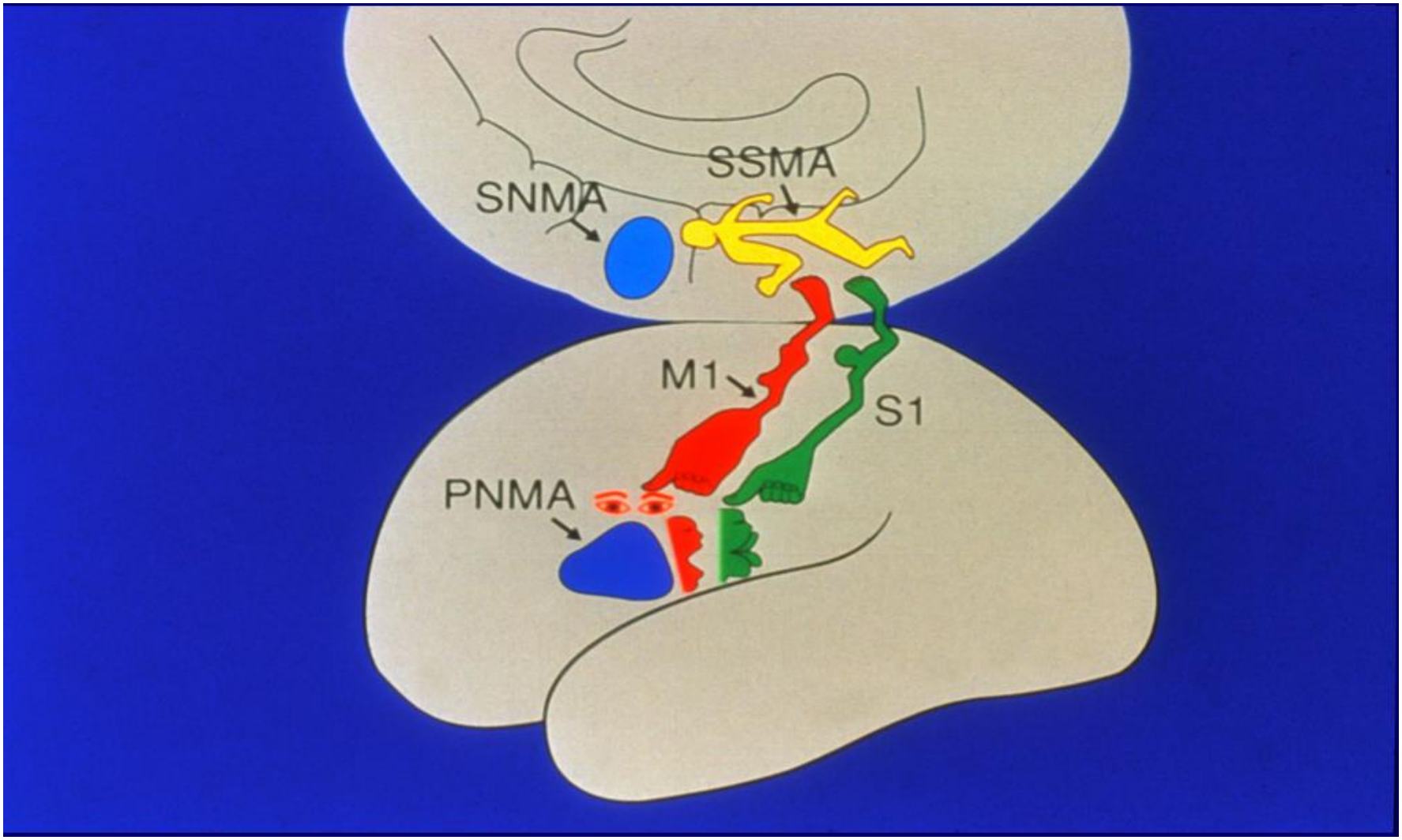
Asymmetrical tonic seizure

- 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

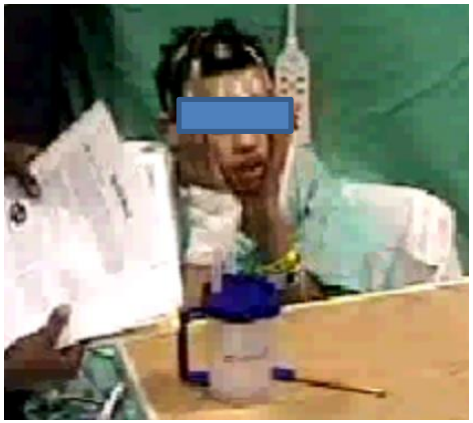
- Location: Supplementary sensorimotor area (SSMA)

Supplementary sensorimotor area (SSMA)

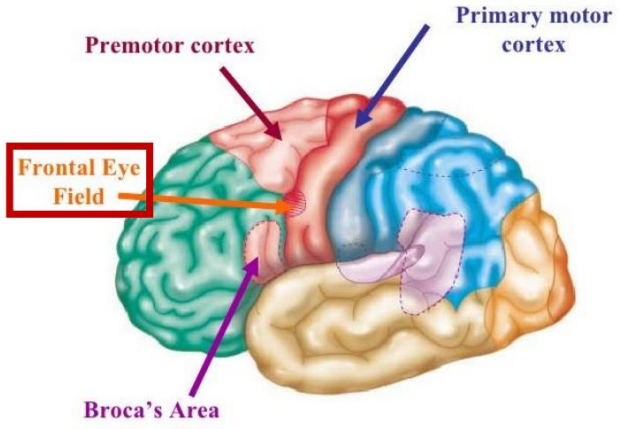


Versive seizures

- **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



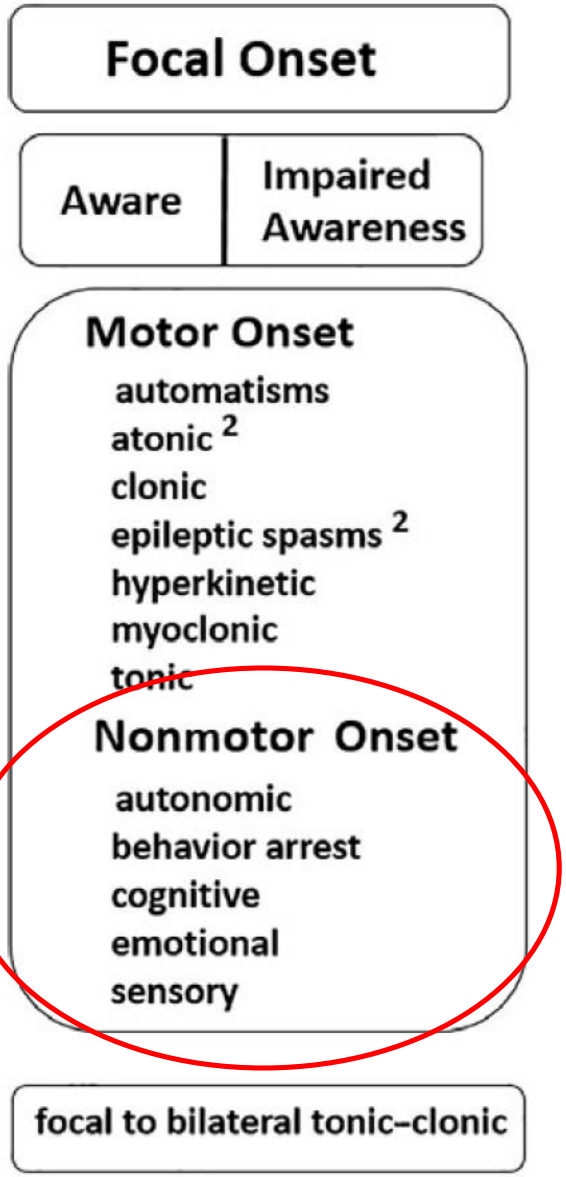
Common lateralising seizure manifestations

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

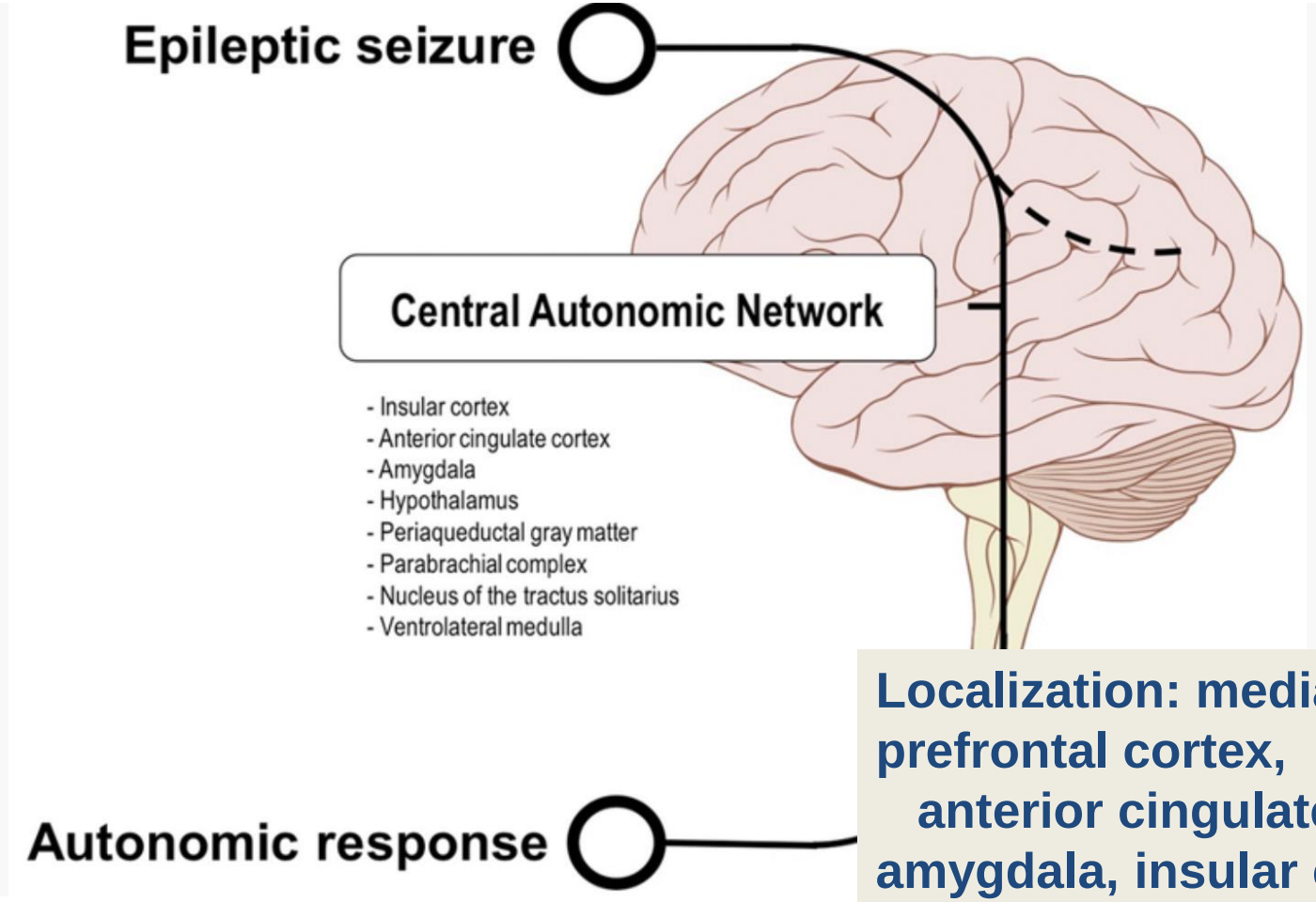
ILAE 2017 Classification of Seizure Types Expanded Version [1]



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



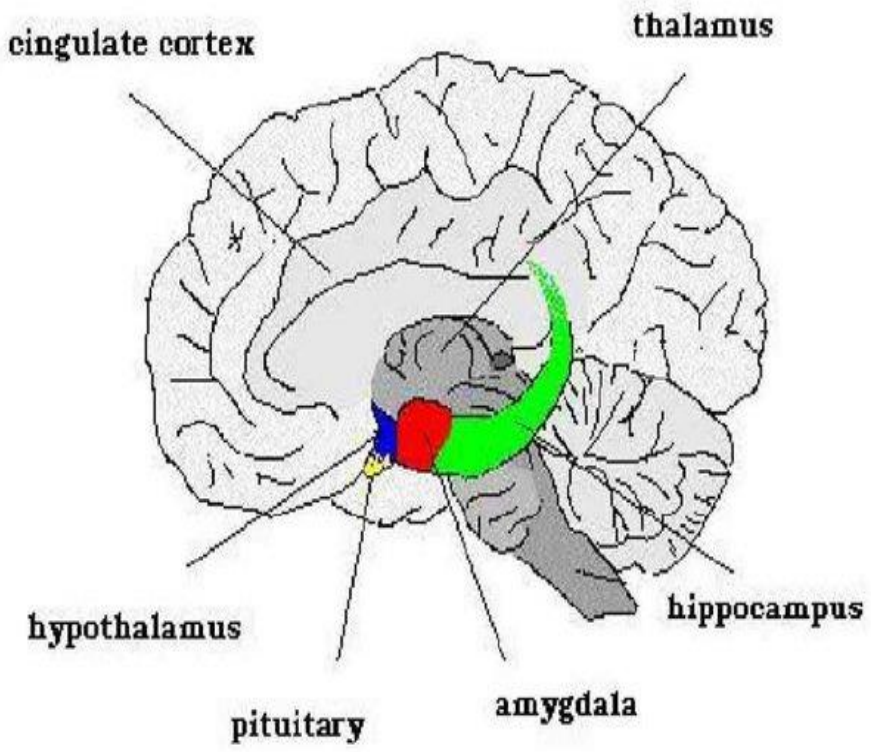
Localization: medial prefrontal cortex, anterior cingulate, amygdala, insular cortex

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

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

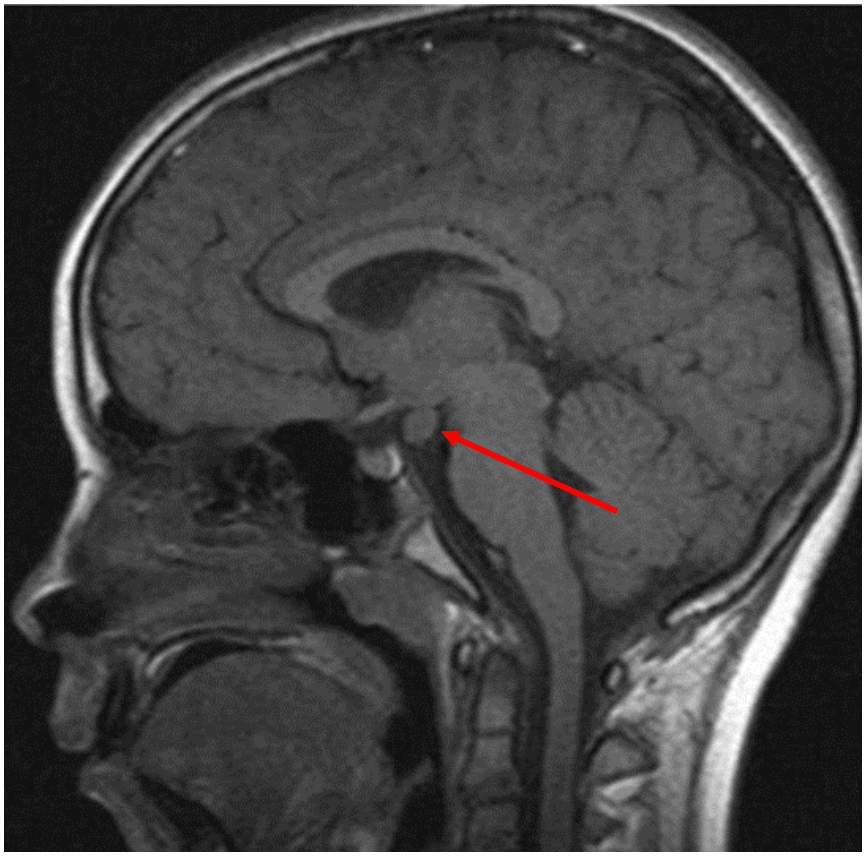


Limbic network

Gelastic seizure

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

Gelastic seizure

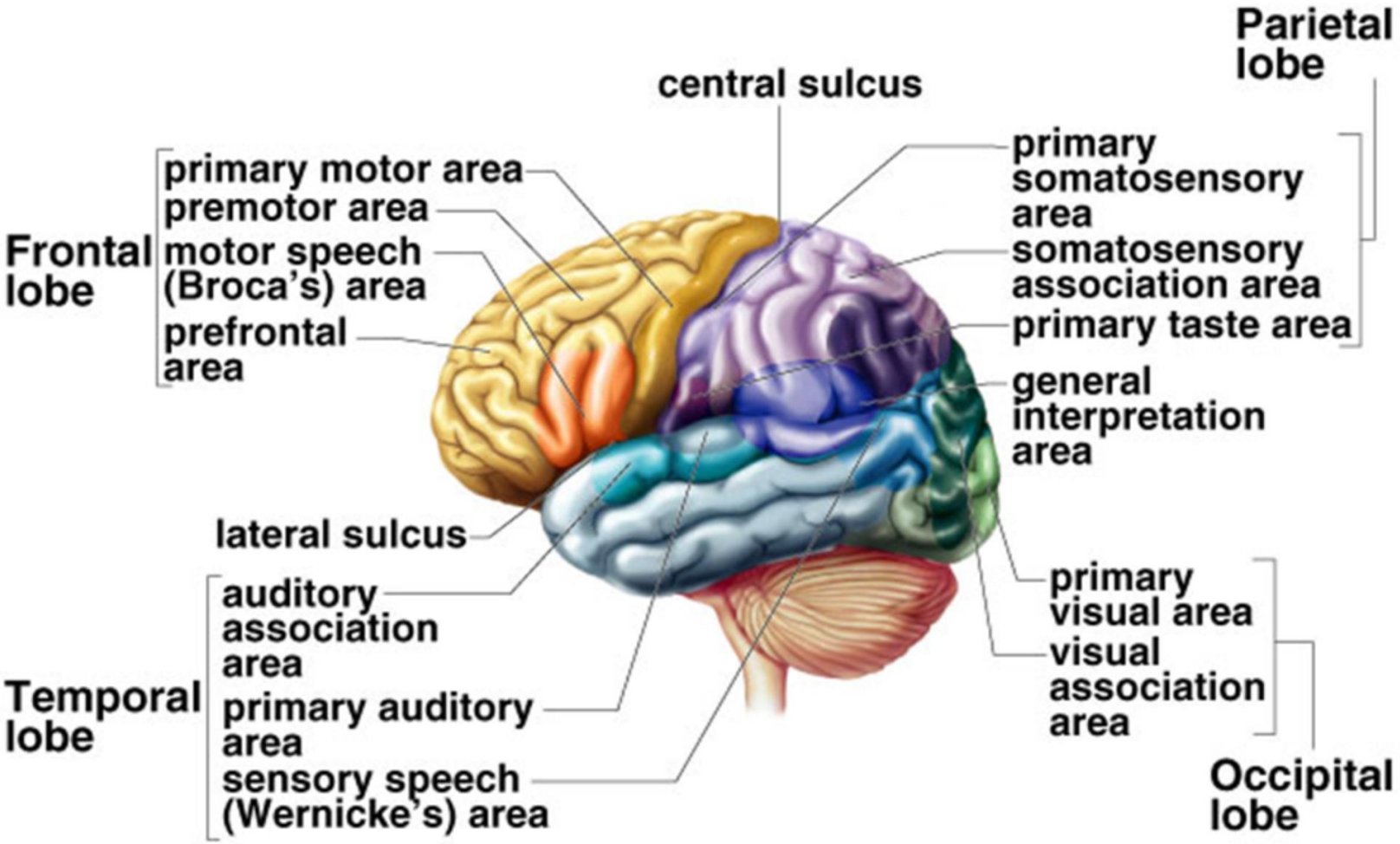


- This seizure type is characteristic of seizures arising in the hypothalamus (*Hypothalamic hamartoma*)
- But can occur in seizures arising in the *frontal* or *temporal* lobes.

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



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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%

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