

# Introduction to Seizure Semiology: Focal Epilepsy

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# 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

### 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<sup>2</sup>

### Non-Motor (absence)

typical  
atypical  
myoclonic  
eyelid myoclonia

## Unknown Onset

### Motor

tonic-clonic  
epileptic spasms  
**Non-Motor**  
behavior arrest

### Unclassified<sup>3</sup>

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

<sup>2</sup> These could be focal or generalized, with or without alteration of awareness

<sup>3</sup> Due to inadequate information or inability to place in other categories

# Seizure Semiology

- Seizure semiology: a simple and cost effective tool
  - allows localization of the **symptomatogenic zone** which either overlaps or is in close proximity of the **epileptogenic zone**.
  - particularly important in cases of MRI negative focal epilepsy.

# Signs for Localization

Ideally the signs used for localization should fulfill these criteria;

- 1. Easy to identify and have a high inter-rater reliability**
- 2. It has to be the first or one of the earlier components of the seizure in order to have localizing value**
  - Later symptoms or signs are more likely to be due to ictal spread and may have only a lateralizing value.
- 3. The symptomatogenic zone corresponding to the recorded ictal symptom has to be clearly defined and well documented.**
  - Reproducibility of the initial ictal symptoms with cortical stimulation identifies the corresponding symptomatogenic zone.

# Localization

- Hemispheric lateralization: Left or Right
- Lobar localization: Temporal, frontal, occipital, parietal

# Aura



- Somatosensory
- Visual
- Auditory
- Olfactory
- Gustatory
- Autonomic
- Abdominal
- Psychic

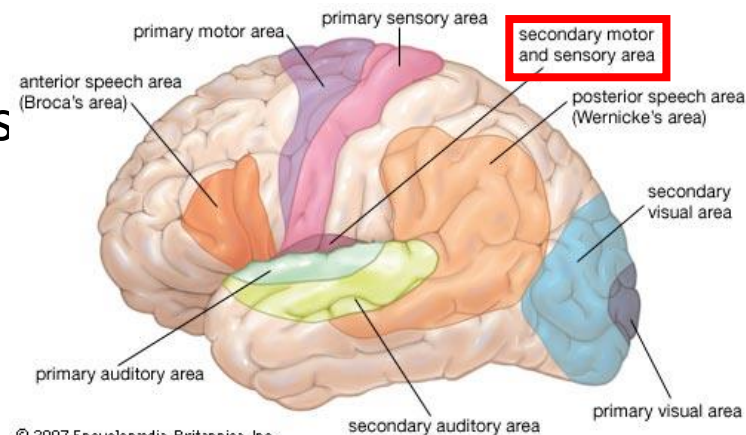
# Somatosensory auras

## Primary sensory cortex

- Abnormal sensations: tingling, numbness
- Limited to clearly defined somatosensory region
- Unilateral, distal distribution

## Supplementary sensorymotor area (SSMA, 2<sup>nd</sup> somatosensory area)

- Superior bank of the Sylvian fissure, the posterior insula.
- Bilateral and widespread sensations
- Stimulation → unpleasant sensations of heat or pain.
- ❖ "all body sensations": **no** localizing



# Visual auras

## **Visual hallucinations: Brodmann's area 17 and 18**

- Flashing lights of different colors, blink and move in the visual field.
- In front of both eyes >> lateralized to one visual field
- Amaurosis

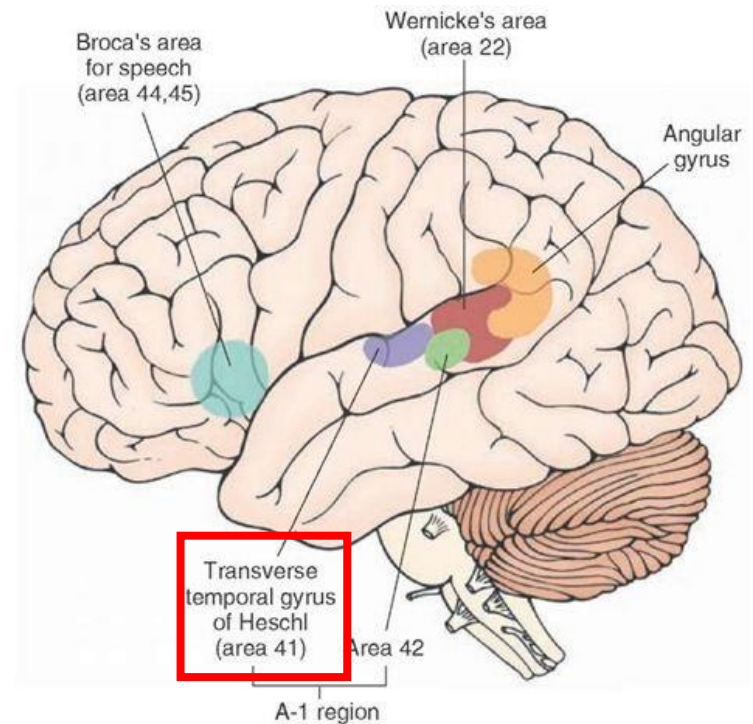
## **Complex visual hallucinations and visual illusions:**

- Association cortex (parieto-temporal) or the adjacent lobes
- Frequently are part of psychic auras.



# Auditory auras

- Simple auditory hallucinations, like hearing a "buzz" or a "noise"
- Heschl's gyrus  
the superior temporal gyrus.
- Not lateralizing



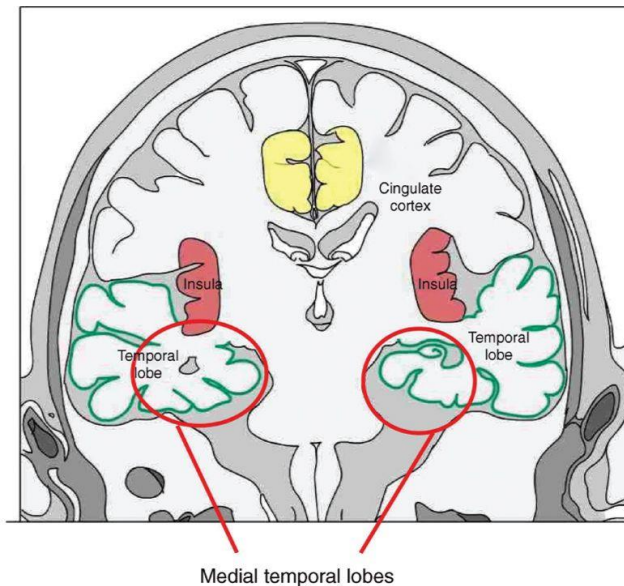
# Olfactory and Gustatory auras

## Olfactory auras

- Unpleasant smells.
- Most frequently seen in mesial temporal lobe epilepsy
- Not lateralizing

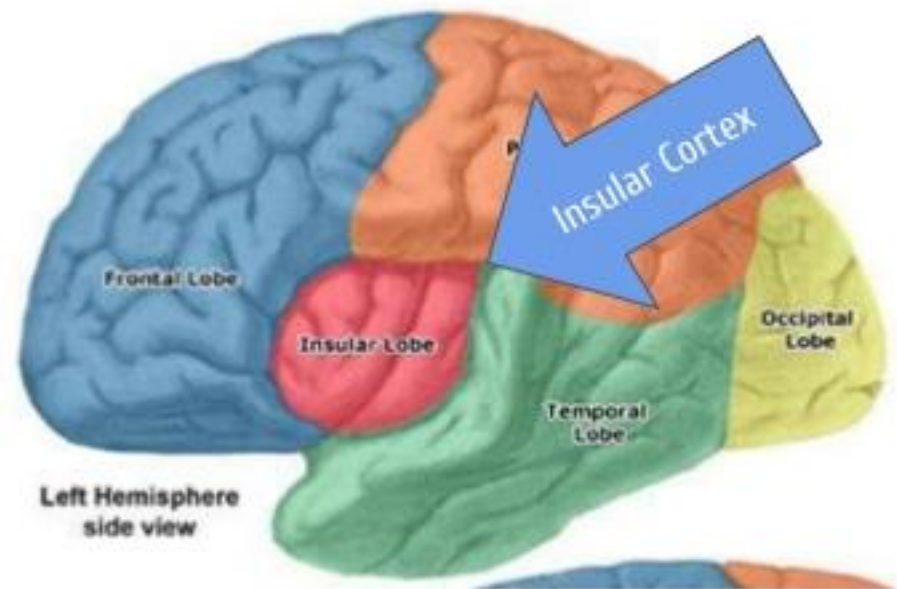
## Gustatory auras

- Unpleasant taste.
- Insula
- Not lateralizing.



# Autonomic auras

- Subjective sensations suggesting possible autonomic alterations such as palpitations, sweating, "goose bumps", etc.
- Insular cortex



# Abdominal auras (1)

- Temporal lobe >> frontal lobe, insula
  - ❖ Sensation produced by increased peristalsis
  - ❖ Direct activation of sensory cortical areas of abdominal viscera.
- Nausea, tenseness, knot, external weight or squeezing, rolling, turning or whirling movement in the abdomen, tickling, tingling or electric shock sensation, pain, vibrating, fluttering or **butterflies sensation**, gas or pressure within the abdomen, an empty, hungry feeling, sensation of warmth, sensation of sudden descent in an elevator, burning or heartburn

# Abdominal auras (2)

- The sensation begins usually in the epigastrium or stomach in the midline and can remain localized there but not infrequently rises to the chest, throat, head or even face.
- Insula, mesial temporal structures, basal ganglia, supplementary motor area, pallidum and centrum medianum of the thalamus.

# Psychic auras

- Complex hallucinations/illusions, usually affect different senses.
- Fear, elation, déjà vu and jamais vu.
- Autoscopic phenomena:
  - seeing a double of the whole or part of the body
  - feeling of presence without optical image
  - out of body experience with observing the self from an elevated position
  - failure to perceive one's own body.
- Temporal lobe (mesial > posterior temporal)
- Not lateralizing

# Aura



Aura	Lateralizing	Localizing
Somatosensory	CL	PLE
Visual	±	OLE
Auditory	No	TLE
Olfactory	No	TLE
Gustatory	No	TLE (Insula)
Autonomic	No	TLE (Insula)
Abdominal	No	TLE > FLE, Insula
Psychic	No	TLE

# Autonomic seizures

- Ictal tachycardia
- Pilomotor seizures; a somatotopic distribution and may spread in a "Jacksonian march" like pattern
  - Ipsilateral to seizure onset zone, not localization
- Ictal vomiting and ictal retching:
  - Temporal lobe (insula) seizures, not lateralizing value
- Ictal spitting:
  - Temporal lobe epilepsy, no lateralizing value
- Ictal hypersalivation:
  - Mesial temporal lobe (non-dominant)



# Dialeptic seizures

- Alteration of consciousness: unresponsiveness during the seizure and amnesia post-ictally.
- At a distance from the primary or supplementary motor areas.
- Duration of dialeptic seizures
  - Seizures arising from mesial temporal structures longer duration than the ones arising from the frontal lobe.

# Motor seizures

## **Simple motor seizures:**

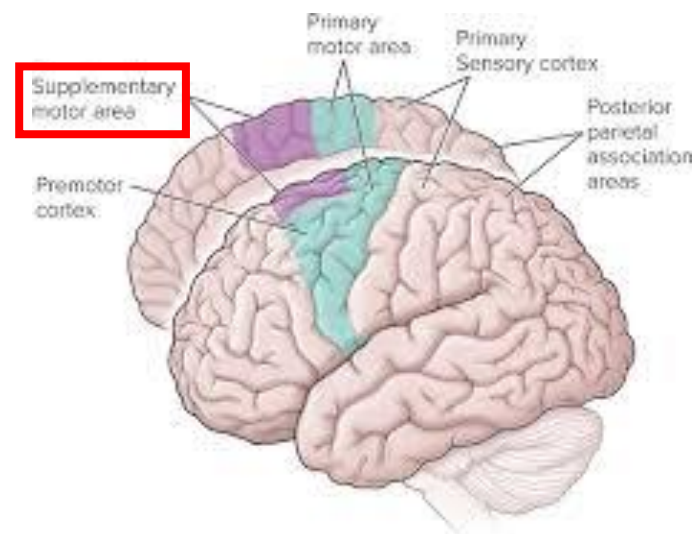
- unnatural but simple movements, usually involving only one articulation in one plane.

## **Complex motor seizures:**

- movements that imitate natural movements, involving several articulations in different planes, and tend to be repetitive.
  - These movements cannot be elicited by electrical cortical stimulation unless a seizure discharge is triggered.

# Tonic seizures

- Sustained muscle contractions, usually lasting several seconds which lead to "posturing"
- Preferentially affect proximal muscle groups
- Occur commonly in frontal lobe epilepsy (62.2%) and very rarely in temporal lobe epilepsy (1.7%).
- High lateralizing significance, pointing to a contralateral seizure onset.
- Preservation of consciousness during bilateral motor activity: seizure focus to the supplementary motor area



# Clonic seizures

- Myoclonic contractions, recur regularly at a rate of 0.2-5/sec
- Primary motor strip
  - temporal lobe epilepsies the face, the frontal eye field and hand areas tend to be affected earlier than legs
- Unilateral clonic seizures: highly reliable lateralizing

## **"End of seizure paradoxical clonus"**

- In 2<sup>o</sup>GTC, clonic activity may persist longer on the side ipsilateral to the epileptogenic focus
- Probably related to "exhausted" of epileptogenic hemisphere.

# Asymmetrical tonic seizures

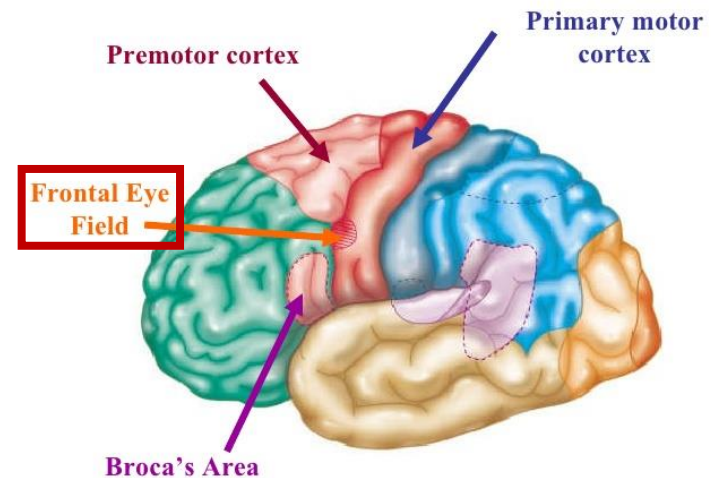
A typical "motor sequence", lateralizing, not localizing

- **Version and pulling of the face (tonic) to the contralateral side → M2e position → asymmetric tonic limb posturing called "sign of four"**.
- Epileptogenic zone can be lateralized with confidence when  $\geq 2$  of these lateralizing signs, (not localizing):
  - Tonic face seizure and the versive seizure lateralize to the contralateral side.
  - Fencing position (M2e) lateralizes to the Contralateral side to the raised arm
  - Asymmetric tonic limb posturing "sign of four" lateralizes to the Contralateral side to the extended arm.



# 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 of both.
- Frontal eye fields, highly lateralizing to the contralateral hemisphere
- Non-versive head turnings: resemble natural movements



# Hypermotor seizures

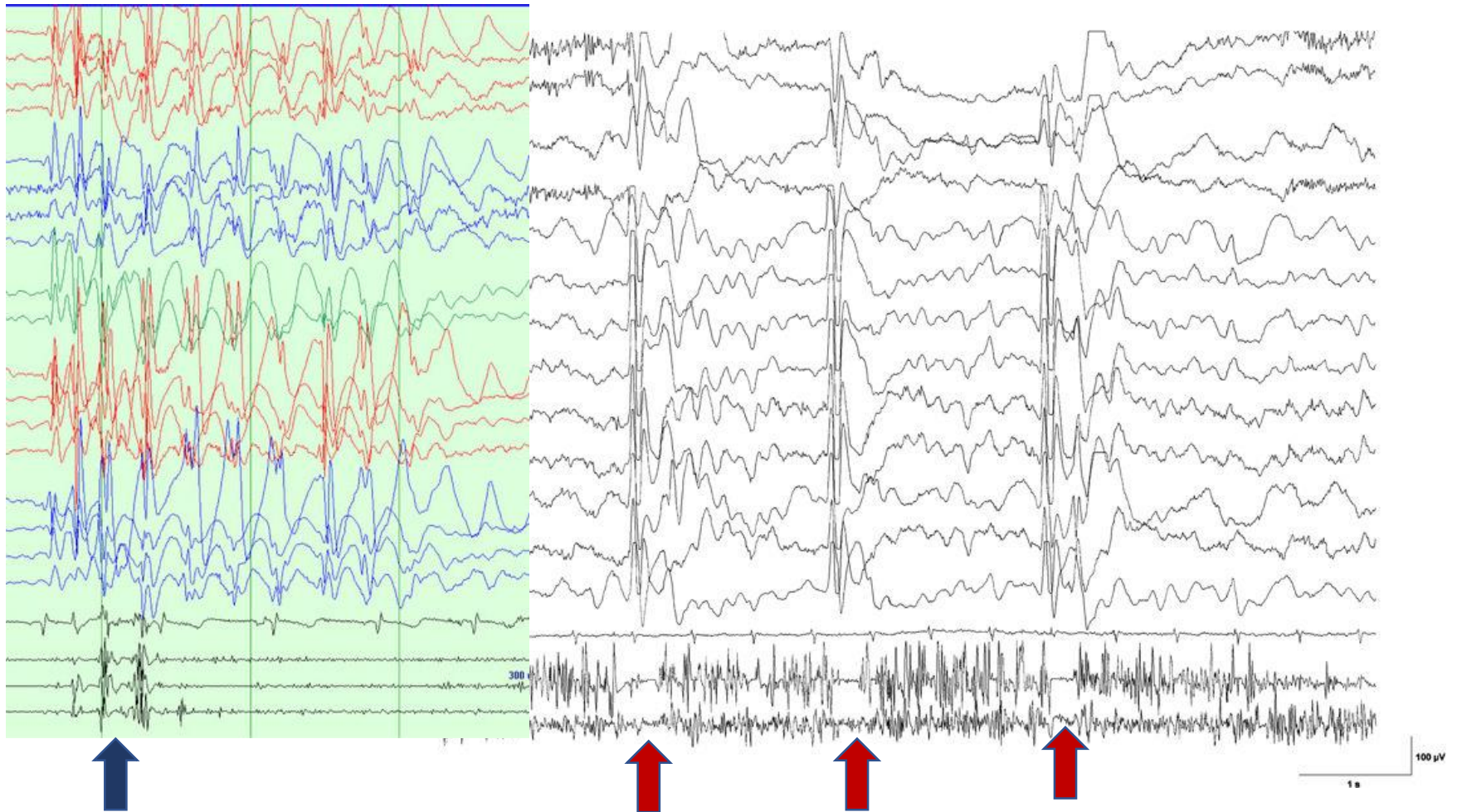
- Complex movements involving trunk and proximal segments of limbs; pedaling, running, etc.
- Automatism resemble sexual activity; violent writhing, thrusting and rhythmic movements of pelvis, arms and legs.
- Sometimes associated with picking and rhythmic manipulation of the groin or genitalia and exhibitionism.
- Consciousness may be preserved
- Occur mostly during sleep.
- Orbital or mesial frontal regions, temporal lobe and insular epilepsies.

# Hypomotor seizures

- Decrease or total absence of motor activity.
- Only used in patients in whom consciousness cannot be tested; children < 3 years, mentally retarded
- In focal epilepsy, hypomotor seizures seen most frequently in temporal and parietal lobe epilepsy.



# Myoclonus and Negative myoclonus

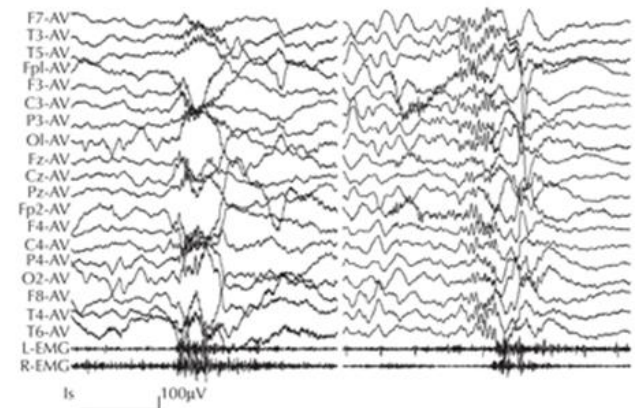


# Negative myoclonic seizures

- Brief movement is produced by a loss of muscle tone of less than 400 msec duration.
- Postcentral cerebral cortex

# Epileptic spasms

- Muscle contractions of relatively symmetric, affect predominantly proximal axial muscles.
- Flexion of the trunk and an extension and abduction of the arms in a "salaam position".
- Infrequently opisthotonic posturing is seen.
- Generalized >> focal epilepsies (parieto-occipital epilepsies)

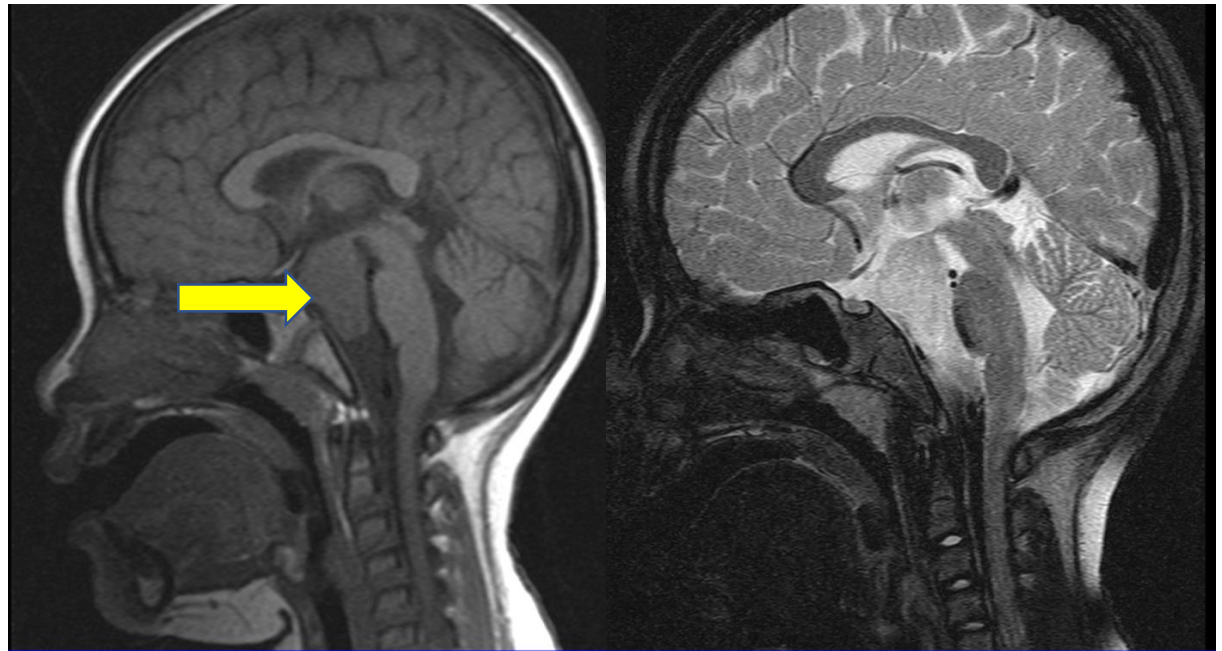


# Automotor seizures

- Automatisms involving distal segments of hands, feet, mouth and tongue.
- 95% associated with altered consciousness.
  - Preservation of consciousness during automotor seizures: almost exclusively in non-dominant mesial temporal epilepsy
- Temporal lobe >> Frontal lobe epilepsy (shorter duration)
- Unilateral automatisms: ipsilateral epileptogenic zone

# Gelastic seizures

- "laughing or giggling"
- Hypothalamic hamartomas (50%), anterior cingulate, frontal, parietal and temporal lobes.



# Akinetic seizures

- Inability to perform voluntary movements.
- Only Dx in patients who are conscious and cooperative, i.e., they try to perform a movement but are unable to do so (apraxia).
- Possibly arise from activation of the negative motor areas in the mesial frontal and inferior frontal gyri.

# Aphasic seizures

- Aphasic despite preserved awareness and memory.
- Can present as status epilepticus.
- Dominant hemisphere

# Lateralizing signs

Semiology	Ipsilateral	Contralateral	PPV(%)
Non forced head turn	X		
Forced head turn		X	94
Eye deviation		X	
Focal clonic		X	95
Asymmetrical clonic ending	X		83
Dystonic limb		X	93
Tonic limb		X	85
M2e, fencing		X	90
Figure of 4		X*	89
Ictal paresis		X	100
Tad paresis		X	80-100

\* To extended limb

So EL, J Clin Neurophysiol 2006.



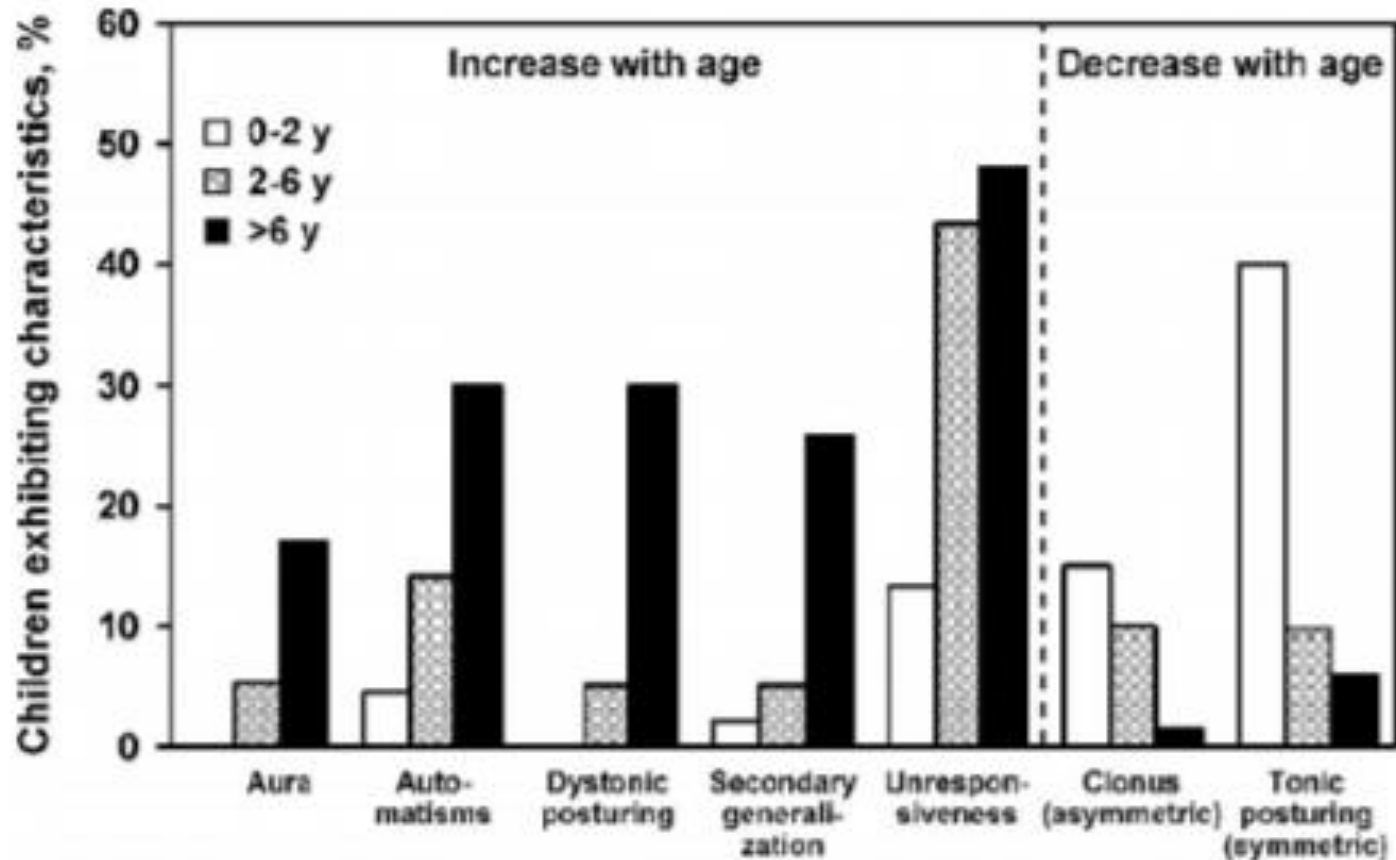
# Peri-ictal speech

Semiology	Ipsi	Cont	Localization	PPV (%)
<b>Speech</b>				
Ictal speech arrest			TLE, dominant	67
Ictal speech preservation			Non-dominant	83
Postictal dysphasia			Dominant	

# Automatisms and Autonomic

Semiology	Ipsi	Cont	Localization	PPV (%)
<b>Automatism</b>				
Unilateral limb automatism	X			90
Unilateral eye blink	X			83
Oral automatism			TLE	
Postictal cough			TLE (50%)	
Postictal nose wiping	X		TLE (50%), FLE (10%)	
Ictal splitting			Rt TLE	
<b>Autonomic</b>				
Ictal emitus			Rt TLE	
Urinary urge			Rt TLE	
Pilo-erection			Lt TLE	

# Seizure characteristics change with age



**FIGURE 2.** Seizure characteristics that change with age. (Modified from Nordli et al., 2001. Used with permission.)