Introduction to Seizure Semiology: Focal Epilepsy

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ILAE 2017 Classification of Seizure Types Expanded Version

Focal Onset
- Aware
- Impaired Awareness

Motor Onset
- automatisms
- atonic
- clonic
- epileptic spasms
- hyperkinetic
- myoclonic
- tonic

Non-Motor Onset
- autonomic
- behavior arrest
- cognitive
- emotional
- sensory

Generalized Onset
- Motor
  - tonic-clonic
  - clonic
  - tonic
  - myoclonic
  - myoclonic-tonic-clonic
  - myoclonic-atonic
  - atonic
  - epileptic spasms
- Non-Motor (absence)
  - typical
  - atypical
  - myoclonic
  - eyelid myoclonia

Unknown Onset
- Motor
  - tonic-clonic
  - epileptic spasms
- Non-Motor
  - behavior arrest

Unclassified

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.

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.

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, 2nd 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: Broadmann'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"
• Heschell'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
<table>
<thead>
<tr>
<th>Aura</th>
<th>Lateralizing</th>
<th>Localizing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Somatosensory</td>
<td>CL</td>
<td>PLE</td>
</tr>
<tr>
<td>Visual</td>
<td>±</td>
<td>OLE</td>
</tr>
<tr>
<td>Auditory</td>
<td>No</td>
<td>TLE</td>
</tr>
<tr>
<td>Olfactory</td>
<td>No</td>
<td>TLE</td>
</tr>
<tr>
<td>Gustatory</td>
<td>No</td>
<td>TLE (Insula)</td>
</tr>
<tr>
<td>Autonomic</td>
<td>No</td>
<td>TLE (Insula)</td>
</tr>
<tr>
<td>Abdominal</td>
<td>No</td>
<td>TLE &gt; FLE, Insula</td>
</tr>
<tr>
<td>Psychic</td>
<td>No</td>
<td>TLE</td>
</tr>
</tbody>
</table>
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 2ºGTC, 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.
• Automatisms 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 opistothonic 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

<table>
<thead>
<tr>
<th>Semiology</th>
<th>Ipsilateral</th>
<th>Contralateral</th>
<th>PPV(%)</th>
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<tr>
<td>Non forced head turn</td>
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<tr>
<td>Forced head turn</td>
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<td>X</td>
<td>94</td>
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<tr>
<td>Eye deviation</td>
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<td>X</td>
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<tr>
<td>Focal clonic</td>
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<td>X</td>
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<tr>
<td>Asymmetrical clonic ending</td>
<td>X</td>
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<tr>
<td>Dystonic limb</td>
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<tr>
<td>Tonic limb</td>
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<tr>
<td>M2e, fencing</td>
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<tr>
<td>Figure of 4</td>
<td>X*</td>
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<tr>
<td>Ictal paresis</td>
<td>X</td>
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<tr>
<td>Tad paresis</td>
<td>X</td>
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<td>80-100</td>
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</table>

* To extended limb

## Peri-ictal speech

<table>
<thead>
<tr>
<th>Semiology</th>
<th>Ipsi</th>
<th>Cont</th>
<th>Localization</th>
<th>PPV (%)</th>
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<tr>
<td><strong>Speech</strong></td>
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<td>Ictal speech arrest</td>
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<td>TLE, dominant</td>
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<td>Ictal speech preservation</td>
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<td>Non-dominant</td>
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<td>Postictal dysphasia</td>
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<td>Dominant</td>
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### Automatisms and Autonomic

<table>
<thead>
<tr>
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<th>Ipsi</th>
<th>Cont</th>
<th>Localization</th>
<th>PPV (%)</th>
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<tbody>
<tr>
<td><strong>Automatism</strong></td>
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<tr>
<td>Unilateral limb automatism</td>
<td>X</td>
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<td>90</td>
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<td>Unilateral eye blink</td>
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<td>Oral automatism</td>
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<td>TLE</td>
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<td>Postictal cough</td>
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<td>TLE (50%)</td>
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<td>TLE (50%), FLE (10%)</td>
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<td>Rt TLE</td>
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<tr>
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<td>Urinary urge</td>
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<tr>
<td>Pilo-erection</td>
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<td>Lt TLE</td>
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Seizure characteristics change with age

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