SEIZURE SEMIOLOGY & CLASSIFICATION

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SEMIIOLOGY

The branch of linguistics concerned with signs and symptoms


THE OFFICE VISIT

- Good seizure history is critical
  - Events surrounding seizure onset, precipitating factors, relationship to sleep/wakefulness
  - Auras
  - Lateralizing signs
  - Postictal phenomena
  - Ask about other seizure types
- Assess SUDEP risk
- General physical examination (abnormal facies, neurocutaneous stigmata, Wood’s lamp examination), chronic AED effects
- Neurological examination
LATERALIZING SIGNS IN SEIZURES

- Auras: somatosensory, visual
- Versive movements of the eyes and head
- Ipsilateral head turning
- Unilateral clonic movements
- Ictal vomiting
- Ictal speech and postictal dysphasia
- Unilateral automatisms
- Unilateral tonic posturing
- Unilateral dystonic posturing
- Lateralized ictal paresis
- Todd’s palsy

- Postictal nose wiping
- Automatisms with preserved responsiveness
- Unilateral eye blinking
- Asymmetrical limb posturing during GTC seizures
  (Figure 4 sign)
- Postictal headache
- Ictal crying
- Facial asymmetry
- Tongue biting
- Ictal spitting

AURAS OF LOCALIZING VALUE

- Somatosensory and elementary visual auras also have lateralizing value
- Epigastric sensation, psychical and olfactory auras are typical of temporal lobe epilepsy
- Cephalic and body sensations seen in frontal, temporal or parietal onset
- Auditory, vestibular and formed visual auras: posterior temporal, parietal
- Forced thinking - frontal lobe
- Gustatory, autonomic auras - perisylvian area
FOCAL CLONIC MOVEMENTS

• Indicate involvement of contralateral sensori-motor cortex
• Do not necessarily indicate seizure onset from this region, as seizures may spread from other areas
• Reliability is good
• Epilepsia Partialis Continua: focal motor status involving a small portion of the sensorimotor cortex

FOCAL CLONIC SEIZURE

SEIZURE VIDEO
LESION IN POST-CENTRAL GYRUS

MULTIPLE AURAS REFLECT SEIZURE SPREAD IN NON-DOMINANT HEMISPHERE


ICTAL SPEECH SUGGESTING SEIZURE ONSET IN THE NON LANGUAGE DOMINANT HEMISPHERE

SEIZURE VIDEO
### SPEECH MANIFESTATIONS IN TEMPORAL LOBE SEIZURES

26 patients, 14 left temporal and 12 right temporal
During postictal period, instructed to read aloud a test phrase, clearly and without errors

<table>
<thead>
<tr>
<th></th>
<th>LTLE</th>
<th>RTLE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean time to read phrase</td>
<td>321.9 sec</td>
<td>19.7 sec</td>
</tr>
<tr>
<td></td>
<td>(68-1276)</td>
<td>(0-106)</td>
</tr>
<tr>
<td>Seizures lateralized correctly</td>
<td>100%</td>
<td>96%</td>
</tr>
<tr>
<td>Using 60 sec cutoff</td>
<td>Privitera 1991</td>
<td></td>
</tr>
</tbody>
</table>

### PATIENT WITH LEFT MTLE

**SEIZURE VIDEO**

### SEQUENCE OF LATERALIZING SIGNS IN TEMPORAL LOBE EPILEPSY

![Diagram](image)
USEFULNESS OF VERSION DEPENDS ON HOW STRICTLY IT IS DEFINED

- **Version**: Clonic or tonic head and eye deviation, which is sustained, unquestionably forced and involuntary resulting in sustained unnatural lateral positioning of head and eyes. Version was always contralateral to side of seizure onset.

- **Non-Versive Movements**: lateral head and eye turning which is unsustained, wandering or seemingly voluntary. Non-verse movements have no lateralizing significance.

Wyllie E et al. Neurology 1986;36:1212-7

ASYMMETRICAL LIMB POSTURING DURING SECONDARILY GENERALIZED SEIZURES

54 patients) seizure free > 1 year after resection (34 temporal, 14 frontal, 3 parietal, 3 occipital)

238 seizure videos analyzed by 3 observers

<table>
<thead>
<tr>
<th>Figure 4 sign</th>
<th>TLE</th>
<th>XTLE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Frequency</td>
<td>78.6%</td>
<td>87.5%</td>
</tr>
<tr>
<td>Predictive value</td>
<td>90.9%</td>
<td>87.5%</td>
</tr>
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</table>

<table>
<thead>
<tr>
<th>Version</th>
<th>TLE</th>
<th>XTLE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Frequency</td>
<td>64.3%</td>
<td>40%</td>
</tr>
<tr>
<td>Predictive value</td>
<td>100%</td>
<td>100%</td>
</tr>
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</table>

Kotagal P et al Epilepsia 2000;41:457-62
Bleasel A et al. Epilepsia 1997;38:168-74

INTER-OBSERVER AGREEMENT & PPV

<table>
<thead>
<tr>
<th>SIGN</th>
<th>TLE</th>
<th>XTLE</th>
<th>p value</th>
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</thead>
<tbody>
<tr>
<td>n=34</td>
<td>n=20</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dystonic Posturing</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Frequency</td>
<td>35%</td>
<td>20%</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Kappa</td>
<td>0.78</td>
<td>0.31</td>
<td></td>
</tr>
<tr>
<td>PPV</td>
<td>92%</td>
<td>100%</td>
<td></td>
</tr>
<tr>
<td>Tonic Posturing</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Frequency</td>
<td>17.7%</td>
<td>15%</td>
<td>0.032</td>
</tr>
<tr>
<td>Kappa</td>
<td>0.23</td>
<td>0.08</td>
<td></td>
</tr>
<tr>
<td>PPV</td>
<td>100%</td>
<td>-</td>
<td></td>
</tr>
<tr>
<td>Immobile Limb</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Frequency</td>
<td>12%</td>
<td>-</td>
<td>0.030</td>
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<tr>
<td>Kappa</td>
<td>0.23</td>
<td>0.06</td>
<td></td>
</tr>
<tr>
<td>PPV</td>
<td>100%</td>
<td>-</td>
<td></td>
</tr>
</tbody>
</table>

Bleasel A et al. Epilepsia 1997;38:168-74
VALUE OF LATERALIZING SIGNS IN TLE

<table>
<thead>
<tr>
<th>SIGN</th>
<th>FREQUENCY</th>
<th>PREDICTIVE VALUE</th>
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<tbody>
<tr>
<td>Focal clonic movements</td>
<td>11%</td>
<td>100% (p=0.05)</td>
</tr>
<tr>
<td>Version</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• &lt; 10sec before gen</td>
<td>27%</td>
<td>100% (p&lt;0.01)</td>
</tr>
<tr>
<td>• at any time during sz</td>
<td>45%</td>
<td>60% (NS)</td>
</tr>
<tr>
<td>Ipsilateral head tilt</td>
<td>9%</td>
<td>60% (NS)</td>
</tr>
<tr>
<td>Unilateral dystonia</td>
<td>18%</td>
<td>90% (p=0.05)</td>
</tr>
<tr>
<td>Unilateral tonic post.</td>
<td>13%</td>
<td>86% (NS)</td>
</tr>
<tr>
<td>Ipsilateral automatisms</td>
<td>9%</td>
<td>80% (NS)</td>
</tr>
<tr>
<td>Eye deviation</td>
<td>27%</td>
<td>57% (NS)</td>
</tr>
<tr>
<td>Face deviation</td>
<td>2%</td>
<td>100% (NS)</td>
</tr>
<tr>
<td>Postictal hemiparesis</td>
<td>2%</td>
<td>100% (NS)</td>
</tr>
</tbody>
</table>

N=55
Marks WJ et al. Epilepsia 1998; 39:721-8

SUPPLEMENTARY MOTOR AREA EPILEPSY

ICTAL ONSET

CORTICAL STIMULATION

GELASTIC SEIZURES CAN BE LOCALIZING!
LOCALIZING VALUE OF SEIZURES

- Escueta & Walsh described features of complex partial seizures and different types.
  - Type I began with motionless stare - temporal lobe onset
  - Type II without motionless stare - extratemporal onset

- Peter Williamson, Felipe Quesney and many others also described features of CPS arising from various locations.

- Weiser was first to apply statistical methods (cluster analysis) to study CPS and proposed 5 subtypes.

SEIZURE VERSUS EPILEPSY

- An epileptic seizure is a transient occurrence and/or symptoms due to a normal excessive or synchronous neuronal activity in the brain.
- Epilepsy is a disorder of the brain characterized by an enduring predisposition to generate epileptic seizures, and by the neurobiologic, cognitive, psychological and social consequences of this condition. The definition of epilepsy requires the occurrence of at least one epileptic seizure.


OPERATIONAL (PRACTICAL) DEFINITION OF EPILEPSY

- At least 2 unprovoked (or reflex) seizures occurring > 24 hours apart
- One unprovoked (or reflex) seizure and > 60% risk of recurrence over the next 10 years. This may be suggested by the finding of epileptiform discharges on EEG, a brain scan showing an epileptogenic lesion, etc.
- Diagnosis of an epilepsy syndrome

EARLY CLASSIFICATION EFFORTS

- 1936 Frederic Gibbs and William Lennox classified psychomotor seizures as separate from petit mal and grand mal
- 1941 Jasper & Kershman proposed a classification of epilepsy based on EEG patterns, location of activity and clinical characteristics of the fits
- 1948 Gibbs concluded that psychomotor seizures originated from the anterior and lateral temporal lobe

CLASSIFICATION OF SEIZURES, EPILEPSIES & EPILEPTIC SYNDROMES

- In 1965, Henri Gastaut started efforts to develop a seizure classification based on EEG and clinical features, principally consciousness
- 1971 ILAE adopted Gastaut’s proposal for Classification of Epileptic Seizures, revised in 1981
- Followed by the ILAE Classification of Epilepsies and Epileptic Syndromes in 1989

1981 ILAE CLASSIFICATION OF EPILEPTIC SEIZURES

PARTIAL
- Simple Partial
  - motor signs
  - sensory, special sensory symptoms
  - autonomic symptoms
- Complex Partial
  - Simple Partial -> Complex Partial
  - Complex Partial from onset
  - Partial -> GTC

GENERALIZED
- Absence
- Myoclonic absences
- Atonic
- Tonic
- Tonic-Clonic
- Atonic

UNCLASSIFIED
- Neonatal

Additional Factors
- Medication
- Stress
- Sleep

*Epilepsy 1981;22:489-501*
### 1989 ILAE Classification of Epilepsies & Epileptic Syndromes

<table>
<thead>
<tr>
<th>Localization-Related</th>
<th>Generalized</th>
<th>Special Syndromes</th>
<th>Undetermined</th>
</tr>
</thead>
<tbody>
<tr>
<td>Idiopathic</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Benign childhood epilepsy w/ C-T spikes</td>
<td>Idiopathic</td>
<td>- BFNC, BNC</td>
<td>- Neonatal</td>
</tr>
<tr>
<td>- Childhood epilepsy w/ occipital paroxysms</td>
<td></td>
<td>- Childhood Absence</td>
<td>- Severe Myoclonic Epilepsy of Infancy (CSWS)</td>
</tr>
<tr>
<td>Symptomatic</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Chronic EPC</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Epilepses w specific modes of precipitation</td>
<td></td>
<td>- Juvenile Absence</td>
<td></td>
</tr>
<tr>
<td>- Epilepses by lobe of origin</td>
<td></td>
<td>- Juvenile Myoclonic Epilepsy w/ GTCs on awakening, etc.</td>
<td></td>
</tr>
<tr>
<td>- Temporal (mesial, lateral)</td>
<td></td>
<td>Symptomatic or Cryptogenic</td>
<td></td>
</tr>
<tr>
<td>- Frontal (SMA, cingulate, frontopolar, orbitofrontal, dorsolateral, opercular, motor cortex)</td>
<td></td>
<td>West Syndrome</td>
<td></td>
</tr>
<tr>
<td>- Parietal</td>
<td></td>
<td>Lennox-Gastaut Syndrome</td>
<td></td>
</tr>
<tr>
<td>- Occipital</td>
<td></td>
<td>Epilepsy w Myoclonic Absences</td>
<td></td>
</tr>
</tbody>
</table>

Cryptogenic

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### Semiological Seizure Classification

**Recommended classifying seizures by semiology alone (independent of EEG, MRI)**

- Introduced new seizure terms: hypermotor, hypomotor, automotor, complex motor, dialeptic, and B/L asymmetric tonic
- Showed how to describe the seizure evolution:
  - Abdominal aura -> automotor seizure -> right versive seizure -> GTC seizure


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### Dialeptic Seizures

**Characterized primarily by alteration of consciousness or relative unresponsiveness.** Subtle motor manifestations may be observed but the alteration of consciousness is always the predominant symptom.

Dialeptic comes from the Greek verb *dialeipin* which means to *stop*, to *interrupt* or to *seize*. It is a synonym of *epileipin*, the Greek root of *epilepsy*.
SEIZURE VIDEO
Abdominal aura -> Automotor Seizure -> Right Versive Seizure -> GTC Seizure
Lateralizing signs: Dystonic posturing, ipsilateral Automatisms, Version, Figure 4

GLOSSARY OF DESCRIPTIVE TERMINOLOGY FOR Ictal SEMIOLOGY:
ILAE TASK FORCE ON CLASSIFICATION & TERMINOLOGY
• Seizure phenomena were categorized and defined:
  – Motor phenomena
  – Auras
  – Autonomic Events
  – Laterality
  – Modifiers and Descriptors of Seizure timing, Precipitating factors
  – Status Epilepticus
  – Severity
  – Prodrome
  – Potencial phenomena
• Introduced new terms: hyperkinetic, hypokinetic,
dyscognitive seizure and somatotopic modifiers

Blume WT, Lüders HO, Mizrahi E et al. Epilepsia 2001;42:1212-8

DYSCOGNITIVE SEIZURE
When disturbance of cognition is the predominant or most apparent feature, typically with 2 or more of these components:-
  • Perception
  • Attention
  • Emotion
  • Memory
  • Executive function

Blume WT, Lüders HO, Mizrahi E et al. Epilepsia 2001;42:1212-8
PROPOSAL FOR DIAGNOSTIC SCHEME FOR EPILEPTICS:
ILAE TASK FORCE REPORT

Axis I: Ictal Phenomenology

Axis II: Seizure Type including localization, precipitating factors

Axis III: Epilepsy Syndrome

Axis IV: Etiology

Axis V: Impairment as per WHO ICIDH-2 scale

Engel J, Jr. Epilepsia 2001;42:796-803
Manual for WHO Disability Assessment Schedule – WHODAS 2.0 (WHO, 2010)

REVISED ILAE TERMINOLOGY & CONCEPTS FOR ORGANIZATION OF SEIZURES AND EPILEPSIES

• New definitions for Generalized, Focal Seizures
• Infantile Spasms included under Epileptic Spasms
• Encourage Use of Descriptive Words (Glossary of Seizures)
• Etiologies: Unknown replaces Cryptogenic
• Categorization of Electroclinical Syndromes by Age of Onset, Constellations and Epilepsies due to Structural/Metabolic causes
• Avoid terms such as “catastrophic” or benign
• Concept of Epileptic Encephalopathy: epileptic activity itself may contribute to severe cognitive and behavioral impairments above and beyond what might be expected from the underlying pathology alone

ILAE Commission on Classification & Terminology. Epilepsia 2010;51:676-85

REVISED DEFINITIONS

SEIZURE
Transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain (Fisher 2005)

GENERALIZED SEIZURE
Seizures arising in and rapidly engaging bilaterally distributed networks. They can include cortical and subcortical structures, but do not necessarily include the entire cortex; can be asymmetric

FOCAL SEIZURE
Seizures originating within networks limited to one hemisphere. For each type, ictal onset is consistent from one seizure to another with preferential propagation patterns that can involve the contralateral hemisphere

ILAE Commission on Classification & Terminology. Epilepsia 2010;51:676-85
2010 ILAE SEIZURE CLASSIFICATION

GENERALIZED SEIZURES
- Tonic-clonic (in any combination)
  - Absence
    - Typical
    - Atypical
      - Absence with special features (myoclonic absence, eyelid myoclonia)
  - Myoclonic
    - Myoclonic
    - Myoclonic atonic
    - Myoclonic tonic
  - Clonic
  - Tonic
  - Atonic

FOCAL SEIZURES
- Unknown
  - Epileptic spasms

DESCRIPTORS OF FOCAL SEIZURES
Without impairment of consciousness or awareness
- With observable motor or autonomic components (Simple Partial seizure).
  Terms like focal motor and autonomic may be used instead.
- Involving subjective sensory or psychic phenomena only (Aura)

With impairment of consciousness or awareness
- Corresponds to Complex Partial Seizure / Dyscognitive Seizure
- Evolving to a Bilateral convulsive seizure
  - May involve tonic, clonic or tonic and clonic components. Replaces secondarily generalized seizure.

Descriptive Terms from Glossary of Ictal Semiology suggested
ILAE Commission on Classification & Terminology. Epilepsia 2010;51:676-85

ELECTROCLINICAL SYNDROMES & OTHER EPILEPSIES

Electroclinical Syndromes by Age of Onset
- Neonatal (<1 month), Infancy (<1 year), Childhood (1-12 years), Adolescence (12-18 years), Adult (18+), ? Elderly (>65 yrs)

Distinctive Constellations
- MTLE with HS, Rasmussen’s, Gelastic Seizures with HH, HHE

Epilepsies due to organized by Structural-Metabolic Causes
- MCD, Neurocutaneous Syndromes, Tumor, Infection, Trauma, Angioma, Perinatal Insults, Stroke etc. includes Genetic causes

Epilepsies of Unknown Cause
- Conditions with epileptic seizures not traditionally diagnosed as epilepsy (benign neonatal seizures, febrile seizures)
2010 ILAE CLASSIFICATION

Without impairment of consciousness or awareness
- With observable motor or autonomic components (Simple Partial seizure). Terms like focal motor and autonomic may be used instead.
- Involving subjective sensory or psychic phenomena only (Aura)

With impairment of consciousness or awareness
Corresponds to Complex Partial Seizure / Dyscognitive Seizure

Evolving to a Bilateral convulsive seizure
May involve tonic, clonic or tonic and clonic components. Replaces secondarily generalized seizure

ILAE Commission on Classification & Terminology Epilepsia 2010;51:676-85

HOW DOES SEIZURE SEMIOLOGY COMPARE TO OTHER MODALITIES?

- Excellent for seizure lateralization; some auras useful for localization
- Semiological features strongly suggest lobe or origin and spread pattern; sublobar localization possible
- Different seizure types with different ictal EEG patterns likely indicate >1 epileptogenic focus
- Discordance of presurgical data should be resolved before placing invasive electrodes or performing resection
- A unique way to study mechanisms, pathways of ictal spread

ILAE TASK FORCE REPORT: CLASSIFICATION OF STATUS EPILEPTICUS

- Operational definition
  Tonic clonic status should be treated if sz is > 5 min (10 min for non-convulsive sz)
- Seizure type: convulsive, non-convulsive
- Etiology: known vs unknown, acute/remote/progressive or unknown
- EEG correlates: generalized or lateralized
- Age and/or electroclinical syndrome

Trinka E et al. Epilepsia 2015;56:1515-23
Seizure Semiology may be as good as EEG, MRI

Elwan S, Alexopolous A, Silveria D, Kotagal P (in preparation)