SEIZURE SEMIOLOGY & CLASSIFICATION

Prakash Kotagal, M.D.

SEMIOLOGY

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
- Determine Etiology
- Assess SUDEP risk
- General physical examination (abnormal facies, neurocutaneous stigmata, Wood’s lamp examination), chronic AED effects
- Neurological examination
FACIAL ASYMMETRY IN PATIENT WITH LEFT TLE

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

LATERALIZING SIGNS IN SEIZURES

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

VIDEO OF FOCAL CLONIC SEIZURE
SENSORY AURA LEFT FACE -> LEFT FACE & ARM CLONIC

VIDEO

LESION IN POST-CENTRAL GyrUS
A 10 year old boy’s seizures begin with staring, unresponsiveness, lip-smacking and twitching of left eyelid lasting around 1 minute. His seizures likely arise in the:

A. Left temporal lobe  
B. Right temporal lobe  
C. Right parietal lobe  
D. Left occipital lobe  
E. Right occipital lobe

IPSILATERAL (LEFT EYE) BLINKING WITH CONTRALATERAL (RIGHT HAND) TONIC POSTURING IN BOY WITH LEFT TEMPORAL LOBE TUMOR
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>LTLE</th>
<th>RTLE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean time to read phrase</td>
<td>321.9 sec</td>
</tr>
<tr>
<td>(68-1276)</td>
<td>(0-106)</td>
</tr>
<tr>
<td>Seizures lateralized correctly</td>
<td>100%</td>
</tr>
</tbody>
</table>

Using 60 sec cutoff

Privitera 1991

VIDEO OF PATIENT WITH LEFT MTLE

EPIGASTRIC AURA

AUTOMOTOR PHASE (staring, lip-smacking, right hand dystonic posturing with left hand automatisms)

RIGHT VERSION

FIGURE 4 POSTURING (right arm extended, left arm flexed during tonic phase of GTC seizure)

GTC SEIZURE

SEQUENCE OF LATERALIZING SIGNS IN TEMPORAL LOBE EPILEPSY

BILATERAL UPPER LIMB AUTOMATISMS

CONTRALATERAL DYSTONIC POSTURING & IPSILATERAL HAND AUTOMATISMS

VERSION

SECONDARY GENERALIZATION

Kotagal P, Arch Neurol 1999;56:912-3
SUBDURAL EEG RECORDING DURING DYSTONIC POSTURING


HYPERPERFUSION OF CONTRALATERAL BASAL GANGLIA DURING DYSTONIC POSTURING

Newton 1992

RIGHT TEMPORAL & BASAL GANGLIA HYPOMETABOLISM IN PATIENT WITH LEFT ARM DYSTONIC POSTURING

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-versive movements have no lateralizing significance.

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

EEG DURING LATE IPSIVERSION TO RIGHT

After bilateral spread, seizure ended in the Left hemisphere

Seizure started in Right hemisphere

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

VIDEO

ASYMMETRIC LIMB POSTURING DURING SECONDARILY GTC SEIZURE (FIGURE 4 SIGN)
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 1

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

Bleasel A et al. Epilepsia 1997;38:168-74

INTER-OBSERVER AGREEMENT & PPV

Table 2

<table>
<thead>
<tr>
<th>SIGN</th>
<th>TLE</th>
<th>XTL</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dystonic Posturing</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Frequency</td>
<td>35%</td>
<td>20%</td>
<td></td>
</tr>
<tr>
<td>Kappa</td>
<td>0.78</td>
<td>0.31</td>
<td>&lt; 0.001</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></td>
</tr>
<tr>
<td>Kappa</td>
<td>0.23</td>
<td>0.08</td>
<td>0.032</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></td>
</tr>
<tr>
<td>Kappa</td>
<td>0.23</td>
<td>0.06</td>
<td>0.030</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 3

<table>
<thead>
<tr>
<th>SIGN</th>
<th>FREQUENCY</th>
<th>PREDICTIVE VALUE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Focal clonic movements</td>
<td>11%</td>
<td>100% (p&lt;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&lt;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>

Marks WJ et al. Epilepsia 1998;39:721-6
VIDEO OF SEIZURES ARISING FROM LEFT SUPPLEMENTARY MOTOR AREA

BILATERAL ASYMMETRIC TONIC SEIZURE
NOTICE RIGHT ARM EXTENSION

SUPPLEMENTARY MOTOR AREA EPILEPSY

ICTAL ONSET  CORTICAL STIMULATION

GELASTIC SEIZURES CAN BE LOCALIZING!

VIDEO
QUESTION
A patient's seizures begin with awareness of his eyes pulling to the left, followed by loss of awareness, rapid eye blinking and then abrupt stiffening of all extremities. You suspect his seizures to start in the:
A. Right Frontal lobe
B. Right Temporal lobe
C. Left Parietal lobe
D. Left Occipital lobe
E. Right Occipital lobe

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 Querney 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.
NEOCORTICAL TEMPORAL EPILEPSY
STATISTICAL ANALYSIS OF SEIZURES

<table>
<thead>
<tr>
<th>SYMPTOM</th>
<th>NTLE</th>
<th>NTLE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Auras</td>
<td>Epigastric</td>
<td>Auditory</td>
</tr>
<tr>
<td></td>
<td>Fear</td>
<td>Cephalic</td>
</tr>
<tr>
<td></td>
<td>Olfactory</td>
<td>Indescribable sens.</td>
</tr>
<tr>
<td>Ictal symptoms</td>
<td>Oralimentary</td>
<td>Early Loss of Contact</td>
</tr>
<tr>
<td></td>
<td>Dystonic posturing</td>
<td>Complex gestures</td>
</tr>
<tr>
<td></td>
<td>Ictal vomiting</td>
<td>Anxiety/agitation</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Ictal speech</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Earlier generalization</td>
</tr>
</tbody>
</table>

Anand & Kotagal 1997

PROGRESSION OF SEMIOLOGY IN TEMPORAL LOBE
vs FRONTAL LOBE COMPLEX PARTIAL SEIZURES

TEMPORAL LOBE SEIZURES
- Behavioral Anomalies
- Cognitive Anomalies
- Auditory Anomalies
- Speech Anomalies
- Motor Anomalies
- Olfactory Anomalies
- Visual Anomalies
- Auditory/Visual Anomalies
- Irritability
- Agitation
- Partial LOC

FRONTAL LOBE SEIZURES
- Complete LOC
- Complex Motor Activity
- Hyperactivity
- Vocalizations
- Olfactory Anomalies
- Visual Anomalies
- Speech Anomalies
- Anxious/agitated
- Partial LOC

QUESTION

A 8 year old girl presents with 5 episodes in the past 2 months consisting of left face and left arm jerking without loss of awareness. These occurred 1-2 hours after going to bed. Her development and examination are normal. Her EEG is most likely to show:

A. Generalized spikes
B. Focal sharp waves in the Right anterior temporal lobe
C. Focal sharp waves in the Right occipital region
D. Focal sharp waves in the Right centro-temporal region
E. Bifrontal spikes and sharp waves

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


QUESTION

A 17 year old girl has a convulsion at school witnessed by her teacher, described as body stiffening, arching of the trunk and shaking of the whole body lasting 1 minute with tongue biting and urinary incontinence. She had not slept the previous night. Her outpatient EEG showed generalized spike wave complexes at 6Hz. You conclude that she has:

A. Isolated seizure due to sleep deprivation
B. Spike-wave discharges are an incidental finding
C. Non-epileptic event
D. Juvenile Myoclonic Epilepsy

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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
  - autonomic or special sensory symptoms
  - psychic symptoms
- Complex Partial
  - Simple Partial -> Complex Partial
  - Complex Partial from onset
  - Partial -> GTC
SPS -> GTC
CPS -> GTC
SPS -> CPS -> GTC

GENERALIZED
- Absence
- Myoclonic absence
- Myoclonic
- Tonic
- Tonic-Clonic
- Atonic

UNCLASSIFIED
- Status Epilepticus

**1989 ILAE CLASSIFICATION OF EPILEPSIES & EPILEPTIC SYNDROMES**

**LOCALIZATION-RELATED**
- Idiopathic
  - Benign childhood epilepsy w/ C/T spikes
  - Childhood epilepsy w/ occipital paroxysms
- Symptomatic
  - Chronic EPC
  - Epilepsies w/ specific modes of precipitation
  - Epilepsies by lobe of origin
    - Temporal (mesial, lateral)
    - Frontal (SMA, cingulate, frontopolar, orbitofrontal, dorsolateral, opercular, motor cortex)
    - Parietal
    - Occipital

- Cryptogenic

**GENERALIZED**
- Idiopathic
  - BFNC, BNC
  - Childhood Absence
  - Juvenile Absence
  - Juvenile Myoclonic
  - Epilepsy w/ GTCs on awakening, etc.

- Symptomatic or Cryptogenic
  - West Syndrome
  - Lennox-Gastaut Syndrome
  - Epilepsy w/ Myoclonic-Astatic seizures
  - Epilepsy w/ Myoclonic Absences

**SPECIAL SYNDROMES**
- Situation related
  - Isolated seizures/status
  - Metabolic / toxic

**UNDETERMINED**
- Neonatal
  - Severe Myoclonic Epilepsy of Infancy (CSWS)

**SEMILOGICAL 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

**VIDEO OF HYPERMOTOR (HYPERKINETIC) SEIZURE**
VIDEO OF HYPOMOTOR SEIZURE

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*.

Glossary of Descriptive Terminology for Ictal Semiology:

- Seizure phenomena were categorized and defined:
  - Motor phenomena
  - Auras
  - Autonomic Events
  - Laterality
  - Modifiers and Descriptors of Seizure timing, Precipitating factors
  - Status Epilepticus
  - Severity
  - Prodrome
  - Postictal phenomena
- Introduced new terms: hyperkinetic, hypokinetic, *dyscognitive seizure* and somatotopic modifiers

Blume WT, Lüders HO, Mozah E et al. Epilepsia 2001;42:1212-8
DYSOCGNITIVE 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
- Infants seizures 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

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
- Epileptic spasms

UNKNOWN

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 Structural-Metabolic Causes
- MCD, Neurocutaneous Syndromes, Tumor, Infection, Trauma, Angioma, Perinatal Insults, Stroke etc.

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)

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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
QUESTION

A 22 year old female with recurrent uncontrolled seizures who failed high dose Oxcarbazepine is admitted to the Epilepsy Monitoring Unit. Four typical episodes were recorded on the first night showing vigorous movements of all extremities, bizarre posturing, pelvic thrusting and screaming. EEG did not reveal any interictal spikes and no seizure pattern could be seen during these episodes. You conclude that these are:

A. Psychogenic seizures
B. Frontal lobe seizures
C. Seizures arising from the mesial-basal temporal lobe
D. Occipital lobe seizures
E. Needs additional VEEG monitoring

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E. Needs additional VEEG monitoring

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)