

Generalized Epilepsy



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Generalized Epilepsy

Generalized epilepsies and syndromes?

- ▶ Generalized Sz's with initial involvement of both hemisphere
- ▶ Idiopathic
 - ▶ No underlying cause (normal EEG and neuroimaging) other than a possible hereditary predisposition
- ▶ Cryptogenic
 - ▶ a hidden or occult cause, but unknown etiology
 - ▶ presumed to be symptomatic
- ▶ Symptomatic
 - ▶ the consequence of a known or suspected CNS disorder

International Classification of Epilepsies & Epilepsy Syndromes (ILAE 1989)

1. Localization-related

- 1.1. Idiopathic: BECT, CEOP, PRE
- 1.2. Symptomatic: Kojewnikow's syndrome, Sz by SMP
Lobar epilepsies: localized vs. unlocalized*
- 1.3. Cryptogenic: Lobar epilepsies: localized vs. unlocalized*

2. Generalized

- 2.1. Idiopathic: BME in infancy, CAE, JAE, JME,
GTC at awakening, Sz by SMP, others*
- 2.2. cryptogenic or symptomatic :WS, LGS, EMAS, MAE
- 2.3. symptomatic: nonspecific etiology*, specific diseases

3. Undetermined*

- 3.1. with both focal and generalized features
- 3.2. without unequivocal generalized or focal features

4. Special Syndromes

Semiology is a keystone of epilepsy diagnosis

Diagnosis of Sz or Epilepsy is based on clinical judgment

History is the most essential for the diagnosis

Epilepsy is a minority diagnosis among paroxysmal events

EEG and MRI are important but provide only supportive evidence to clinical judgment

Semiological Seizure Classification (Lüders et al., 1998)

❖ Aura

- somatosensory
- gustatory
- abdominal
- visual
- olfactory
- psychic
- auditory
- autonomic

❖ Autonomic Sz (documentation of autonomic dysfunction)

- ❖ Dialeptic Sz (predominant Sx is altered consciousness)
 - typical dialeptic Sz (consistent with typical absence Sz)

❖ Motor Sz

- simple motor: - myoclonic, - tonic, - clonic, - spasm
- tonic-clonic, - versive
- complex motor: - hypermotor, - gelastic, - automotor

❖ Special Sz (Szs difficult to classify into one of above 4 types)

- atonic
- akinetic
- astatic
- negative
- hypomotor
- aphasic

Semiological Seizure Classification (Lüders et al., 1998)

♣ **Epileptic syndrome:**
Defined by considering all clinical information

- Semiologic seizure type
- Interictal EEG
- Ictal EEG
- Functional & anatomic neuroimaging

<p>1. Left Mesial Temporal Lobe Epilepsy Seizures: Abdominal aura → automotor seizure Cause: Left mesial temporal sclerosis Related medical conditions: Febrile convulsions, Memory deficit, Cerebellar atrophy</p> <p>2. Absence Epilepsy Seizures: Typical dialeptic seizure → generalized tonic-clonic seizure Cause: Genetic Related medical conditions: None</p> <p>3. Lennox-Gastaut syndrome Seizures: Generalized tonic seizure, Dialeptic seizure, Astatic seizure Cause: Tuberos sclerosis Related medical conditions: Mental retardation, Depression</p>	
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Diagnostic Scheme for People with Epileptic Seizures and Epilepsies (Engel J. ILAE commission, 2001)

This diagnostic scheme is divided into five parts, or Axes, organized to facilitate a logical clinical approach to the development of hypotheses necessary to determine the diagnostic studies and therapeutic strategies to be undertaken in individual patients:

Axis 1: Ictal phenomenology, from the Glossary of Descriptive Ictal Terminology, can be used to describe ictal events with any degree of detail needed.

Axis 2: Seizure type, from the List of Epileptic Seizures. Localization within the brain and precipitating stimuli for reflex seizures should be specified when appropriate.

Axis 3: Syndrome, from the List of Epilepsy Syndromes, with the understanding that a syndromic diagnosis may not always be possible.

Axis 4: Etiology, from a Classification of Diseases Frequently Associated with Epileptic Seizures or Epilepsy Syndromes when possible, genetic defects, or specific pathologic substrates for symptomatic focal epilepsies.

Axis 5: Impairment, this optional, but often useful, additional diagnostic parameter can be derived from an impairment classification adapted from the WHO ICDH-2.

♣ **Axis-dependent**

Axis 1. ictal phenomenology
= In The Glossary of Descriptive Ictal Terminology (Blume et al. 2001)

Axis 2. seizure type
Use the International Classification of Epileptic Seizure (ILAE, 1981)

Axis 3. epilepsy syndrome
Use the International Classification of Epileptic Syndromes (ILAE, 1989)

Axis 4. etiology
Use Classification of Diseases Frequently Associated with Epileptic Seizures or Epilepsy Syndromes (Engel, 2001) - Malformation, tumors, neurocutaneous disorders, genetic factors, metabolic disorders...

Axis 5. impairment

Glossary of Descriptive Terminology for Ictal Semiology (Blume et al. Epilepsia 2001)

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ILAE Commission Report

Glossary of Descriptive Terminology for Ictal Semiology: Report of the ILAE Task Force on Classification and Terminology

Warren T. Blume—Chair, Hans O. Lüders, Eli Mizrahi, Carlo Tassinari, Walter van Emde Boas, and Jerome Engel, Jr., Ex-officio

Glossary of Descriptive Terminology for Ictal Semiology (Blume et al. Epilepsia 2001)

1.0 MOTOR

<p>1.1 ELEMENTARY MOTOR</p> <p>1.1.1 TONIC</p> <p>1.1.1.1 EPILEPTIC SPASM</p> <p>1.1.1.2 POSTURAL</p> <p>1.1.1.2.1 VERSIVE</p> <p>1.1.1.2.2 DYSTONIC</p> <p>1.1.2 MYOCLONIC</p> <p>1.1.2.1 NEGATIVE MYOCLONIC</p> <p>1.1.2.2 CLONIC</p> <p>1.1.2.2.1 JACKSONIAN MARCH</p> <p>1.1.3 TONIC-CLONIC</p> <p>1.1.3.1 GENERALIZED TONIC-CLONIC SEIZURE</p> <p>1.1.4 ATONIC</p> <p>1.1.5 ASTATIC</p> <p>1.1.6 SYNCHRONOUS</p>	<p>1.2 AUTOMATISM</p> <p>1.2.1 ORALIMENTARY</p> <p>1.2.2 MIMETIC</p> <p>1.2.3 MANUAL OR PEDAL</p> <p>1.2.4 GESTURAL</p> <p>1.2.5 HYPERKINETIC</p> <p>1.2.6 HYPOKINETIC</p> <p>1.2.7 DYSPHASIC</p> <p>1.2.8 DYSPRAXIC</p> <p>1.2.9 GELASTIC</p> <p>1.2.10 DACRYSTIC</p> <p>1.2.11 VOCAL</p> <p>1.2.12 VERBAL</p> <p>1.2.13 SPONTANEOUS</p> <p>1.2.14 INTERACTIVE</p>
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Glossary of Descriptive Terminology for Ictal Semiology (Blume et al. Epilepsia 2001)

2.0 NON-MOTOR

2.1 AURA

2.2 SENSORY

2.2.1 ELEMENTARY

2.2.1.1 SOMATOSENSORY

2.2.1.2 VISUAL

2.2.1.3 AUDITORY

2.2.1.4 OLFACTORY

2.2.1.5 GUSTATORY

2.2.1.6 EPIGASTRIC

2.2.1.7 CEPHALIC

2.2.1.8 AUTONOMIC

2.2.2 EXPERIENTIAL

2.2.2.1 AFFECTIVE

2.2.2.2 MNEMONIC

2.2.2.3 HALLUCINATORY

2.2.2.4 ILLUSORY

2.3 DYSCOGNITIVE

3.0 AUTONOMIC EVENTS

3.1 AUTONOMIC AURA

3.2 AUTONOMIC SEIZURE

Seizures in Generalized Epilepsy

What kind of seizures in Gen. Epilepsy?

Gen. Seizures (Blume, 2001)

Epileptic spasm (formerly infantile spasm)

- sudden flexion, extension, flexion-extension of predominantly prox. muscles (longer than myoclonus, ~1s)

Myoclonic seizure

- sudden, brief (<100 ms) involuntary single or multiple contractions of muscle or muscle group.

Tonic seizure

- sustained muscle contraction lasting a few seconds to min.

Atonic seizure

- sudden loss or diminution of muscle tone without apparent preceding myoclonus or tonic event lasting 1-2 s.

Astatic seizure

- loss of erect posture that results from an atonic, myoclonic, or tonic mechanism (synonym: drop attack).

Revised Classification of Epileptic Seizures (ILAE, 2010)

Classification of Seizures	Descriptors of focal seizures according to degree of impairment during seizure
<p>Generalized seizures: Szs originating and rapidly engaging <u>bilaterally distributed networks</u></p> <p>Tonic-clonic (in any combination)</p> <p>Absence</p> <ul style="list-style-type: none"> Typical Atypical <p>Absence with special features</p> <ul style="list-style-type: none"> Myoclonic absence Eyelid myoclonia <p>Myoclonic</p> <ul style="list-style-type: none"> Myoclonic Myoclonic atonic Myoclonic tonic <p>Clonic</p> <p>Tonic</p> <p>Atonic</p> <p>Focal seizures: Szs originating within <u>networks limited to one hemisphere</u></p> <p>Unknown</p> <p>Epileptic spasms</p>	<p>Without impairment of consciousness or awareness</p> <p>With observable motor or autonomic components. This roughly corresponds to the concept of "simple partial seizure".</p> <p>"Focal motor" and "autonomic" are terms that may adequately convey this concept depending on the seizure manifestations).</p> <p>Involving subjective sensory or psychic phenomena only. This corresponds to the concept of an aura, a term endorsed in the 2001 Glossary.</p> <p>With impairment of consciousness or awareness.</p> <p>This roughly corresponds to the concept of complex partial seizure. 'Dyscognitive' is a term that has been proposed for this concept (Blume et al., 2001).</p> <p>Evolving to a bilateral, convulsive seizure (involving tonic, clonic, or tonic and clonic components).</p> <p>This expression replaces the term "secondarily generalized seizure."</p>

Seizures in Generalized Epilepsy

What kind of seizures in Gen. Epilepsy?

Cases