Definition and Classification of Epilepsy

Dr. Suthida Yenjun

Epilepsy Course for Neurological Residents
8 September 2018
Babylonian Cuneiform Tablet

- 1067-1046 B.C.
- The Sakikku (meaning All diseases)
- Many types of seizures were described
- Attributed to a certain demon or spirit

Definition

Epileptic seizure: a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain*

Epileptic seizure

Mode of onset – ‘transient’, demarcated in time, with a clear start and finish

Clinical manifestations – sensory, motor, and autonomic function; consciousness; emotional state; memory; cognition; and behavior

Ictogenesis - abnormal enhanced synchrony of neurones
Definition

Epilepsy: a disorder of the brain characterized by an enduring predisposition to generate epileptic seizures and by the neurobiological, cognitive, psychological and social consequences of this condition*, requires the occurrence of at least one epileptic seizure

Definition

Epileptic syndrome

- Group of clinical and EEG features common to patients with similar, but not necessarily identical etiologies
- Helps determine the appropriate therapy and the prognosis
A practical clinical definition of epilepsy

*Robert S. Fisher, †Carlos Acevedo, ‡Alexis Arzimanoglou, §Alicia Bogacz, ¶J. Helen Cross, #Christian E. Elger, **Jerome Engel Jr, ††Lars Forsgren, †††Jacqueline A. French, §§Mike Glynn, ¶¶Dale C. Hesdorffer, ###B.I. Lee, *****Gary W. Mathern, ††††Solomon L. Moshé, †††††Emilio Perucca, ††††††Ingrid E. Scheffer, †††††††Torbjörn Tomson, †††††Masako Watanabe, and ††††††††Samuel Wiebe

doi: 10.1111/epi.12550

Summary

Epilepsy was defined conceptually in 2005 as a disorder of the brain characterized by an enduring predisposition to generate epileptic seizures. This definition is usually practically applied as having two unprovoked seizures >24 h apart. The International League Against Epilepsy (ILAE) accepted recommendations of a task force altering the practical definition for special circumstances that do not meet the two unprovoked seizures criteria. The task force proposed that epilepsy be considered to be a disease of the brain defined by any of the following conditions: (1) At least two unprovoked (or reflex) seizures occurring >24 h apart; (2) one unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years; (3) diagnosis of an epilepsy syndrome. Epilepsy is considered to be resolved for individuals who either had an age-dependent epilepsy syndrome but are now past the applicable age or who have remained seizure-free for the last 10 years and off antiseizure medicines for at least the last 5 years. “Resolved” is not necessarily identical to the conventional view of “remission or “cure.” Different practical definitions may be formed and used for various specific purposes. This revised definition of epilepsy brings the term in concordance with common use.

KEY WORDS: Epilepsy, Seizure, Definition, Unprovoked, Recurrence.
Epilepsy is a disease of the brain defined by any of the following conditions:

1. At least **two unprovoked (or reflex) seizures** occurring >24 h apart

2. **One unprovoked (or reflex) seizure** and a **probability of further seizures similar to the general recurrence risk (at least 60%)** after two unprovoked seizures, occurring over the next 10 years

3. Diagnosis of an **epilepsy syndrome**

Epilepsy is considered to be **resolved** for individuals who had an age-dependent epilepsy syndrome but are now past the applicable age or those who have remained seizure-free for the last 10 years, with no seizure medicines for the last 5 years.
Epilepsy definition 2014

1. A least two unprovoked (or reflex) seizures occurring >24 h apart

- is the same as the old definition of epilepsy

- After a single unprovoked seizure, risk for another is 40-52%*. With two unprovoked non-febrile seizures, the chance by 4 years of having another seizure is 73%**.

** Hauser WA et al. NEJM 1998;338:429-434.
2. One unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years.

- allows a condition to be considered epilepsy after one seizure if there is a high risk of having another seizure eg. the patient with a remote brain insult and an epileptiform EEG study.
3. Diagnosis of an epilepsy syndrome

- If evidence exists for an epilepsy syndrome, then epilepsy may be presumed to be present, even if the risk of subsequent seizures is low eg., BECTS
Epilepsy definition 2014

<table>
<thead>
<tr>
<th>Epilepsy is considered to be <strong>RESOLVED</strong> for individuals who had an age-dependent epilepsy syndrome but are now past the applicable age or those who have <strong>remained seizure-free for the last 10 years, with no seizure medicines for the last 5 years.</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>- Being resolved does not guarantee that epilepsy will not return, but it means the chances are small and the person has a right to consider that she or he is free from epilepsy. This is a big potential benefit of the new definition</td>
</tr>
</tbody>
</table>
Epilepsy definition 2014

The Treatment decision ≠ The diagnosis

: individualized depending upon the desires of the patient, the individual risk-benefit ratio and the available options
Epilepsy classification

1. **For seizures**: represent the symptoms and signs in epileptology, rarely provide clue as to the underlying aetiology and prognosis

2. **For epilepsies and epileptic syndromes**: defined by electroclinical characteristics, sometimes determined by therapeutic and/or prognostic implication
Epilepsy classification

- **1969** Gastaut proposed a classification of the epilepsies based on a combination of clinical and EEG data.

- **1970** ILAE: International Classification of the Epilepsies (ICE)
  - Generalized (primary, secondary, undetermined)
  - Partial (all were presumed to be symptomatic)
  - Unclassifiable
ILAE Classification (ICE) 1981

- **Partial-onset seizures**
  : begin in a focal area of the cerebral cortex

- **Generalized-onset seizures**
  : an onset recorded simultaneously in both cerebral hemispheres

- **Unclassified seizures**
  : seizures those are difficult to fit into a single class

ILAE Classification (ICE) 1981

Classification of epileptic seizures
(International League Against Epilepsy (ILAE), 1981)

1) Partial (focal, local) seizures
   - Simple partial seizures
     - With motor signs
     - With somatosensory or special sensory symptoms
     - With autonomic symptoms and signs
     - With psychic symptoms
   - Complex partial seizures
     - Simple partial onset followed by impairment of consciousness
     - With impairment of consciousness at onset
   - Partial seizures evolving to secondarily generalized seizures
     - Simple partial seizures evolving to generalized seizures
     - Complex partial seizures evolving to generalized seizures
     - Simple partial seizures evolving to complex partial seizures evolving to generalized seizures

2) Generalized (convulsive or nonconvulsive) seizures
   - Atonic seizures
     - Typical absence
     - Atypical absence
   - Myoclonic seizures
   - Clonic seizures
   - Tonic seizures
   - Tonic–clonic seizures
   - Atonic seizures

3) Unclassified seizures
**The clinical diagnosis is difficult!**

- Aura is a simple partial seizure

- About 20-40% of auras have ictal correlation on the scalp EEG, most are not recorded well on a routine EEG

- If aura > 30 minutes, considered as simple partial status epilepticus by definition >>> not in the classification
### ILAE Classification (ICE) 1981

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
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<tbody>
<tr>
<td>• In practice, assessing the patient's history whether consciousness was impaired is difficult</td>
<td></td>
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</tbody>
</table>

<p>| | |</p>
<table>
<thead>
<tr>
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</thead>
<tbody>
<tr>
<td>• Patients might be able to remember their aura but are unaware that they were briefly unable to respond to the environment &gt;&gt;&gt; ? SPS vs. CPS</td>
<td></td>
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</tbody>
</table>

<p>| | |</p>
<table>
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<tbody>
<tr>
<td>• CPS of frontal lobe origin may feature bizarre motor behaviors, might have a fast postictal recovery, and often appear in clusters &gt;&gt;&gt;</td>
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</tbody>
</table>

? Nonepileptic seizures
ILAE Classification (ICE) 1981

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<table>
<thead>
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<tbody>
<tr>
<td>• Clinically classifying a GTCs as being secondarily generalized (partial onset) or primarily generalized is difficult on the basis of the history alone</td>
<td></td>
</tr>
<tr>
<td>• the aura preceding the secondarily generalized seizure is often forgotten because of postictal amnesia &gt;&gt;&gt; ?</td>
<td></td>
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<tr>
<td>• ? Absence vs. CPS</td>
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</table>
ILAE Classification (ICEES) 1989

Syndromes were organized primarily according to

- **Mode of expression** (localization-related versus generalized)

- **Underlying cause** (idiopathic, symptomatic, and cryptogenic)

## ILAE Classification (ICEES) 1989

### Localization-related (local, focal, partial epilepsies and syndromes)

<table>
<thead>
<tr>
<th>Idiopathic (with age-related onset)</th>
<th>Symptomatic or Cryptogenic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign childhood epilepsy with centrotemporal spikes (benign rolandic epilepsy)</td>
<td>Chronic progressive epilepsy partialis continua</td>
</tr>
<tr>
<td>Childhood epilepsy with occipital paroxysms (Panyiotopoulos syndrome and Gastauf syndrome)</td>
<td>Syndromes characterized by seizures with specific modes of precipitation</td>
</tr>
<tr>
<td>Primary reading epilepsy</td>
<td>Temporal lobe epilepsies</td>
</tr>
<tr>
<td></td>
<td>Frontal lobe epilepsies</td>
</tr>
<tr>
<td></td>
<td>Parietal lobe epilepsies</td>
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<td></td>
<td>Occipital lobe epilepsies</td>
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</tbody>
</table>

### Generalized epilepsies and syndromes

<table>
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<tr>
<th>Idiopathic (with age-related onset)</th>
<th>Symptomatic or Cryptogenic</th>
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<tbody>
<tr>
<td>Benign familial neonatal seizures</td>
<td>West syndrome</td>
</tr>
<tr>
<td>Benign neonatal convulsions</td>
<td>Lennox-Gastaut syndrome</td>
</tr>
<tr>
<td>Benign myoclonic epilepsy in infancy</td>
<td>Epilepsy with myoclonic-astatic seizures</td>
</tr>
<tr>
<td>Childhood absence epilepsy</td>
<td>Epilepsy with myoclonic seizures</td>
</tr>
<tr>
<td>Juvenile myoclonic epilepsy</td>
<td>Early infantile epileptic encephalopathy with suppression burst</td>
</tr>
<tr>
<td>Epilepsy with grand mal seizures on awakening</td>
<td>Early myoclonic encephalopathy</td>
</tr>
<tr>
<td>Juvenile absence epilepsy</td>
<td>Other symptomatic generalized epilepsies</td>
</tr>
<tr>
<td>Syndromes characterized by seizures with specific modes of precipitation</td>
<td></td>
</tr>
<tr>
<td>Other generalized idiopathic epilepsies</td>
<td></td>
</tr>
</tbody>
</table>

### Specific Syndromes

<table>
<thead>
<tr>
<th>Epileptic seizures complicating with other disease states</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epilepsies and syndromes undetermined whether focal or generalized with both generalized and focal seizures</td>
</tr>
<tr>
<td>Neonatal seizure</td>
</tr>
<tr>
<td>Severe myoclonic epilepsy of infancy</td>
</tr>
<tr>
<td>Epilepsy with continuous spike waves during slow-wave sleep</td>
</tr>
<tr>
<td>Acquired epileptic epilepsies (Landau Kleffner syndrome)</td>
</tr>
</tbody>
</table>

### Other determined epilepsies without unequivocal generalized or focal features

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<th>Epilepsies and syndromes undetermined whether focal or generalized with both generalized and focal seizures</th>
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<td>Acquired epileptic epilepsies (Landau Kleffner syndrome)</td>
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</table>

### Special syndromes

<table>
<thead>
<tr>
<th>Situation–related seizures</th>
<th>Febrile convulsions</th>
</tr>
</thead>
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<td>Situation–related seizures</td>
<td>Febrile convulsions</td>
</tr>
</tbody>
</table>
Epileptic syndrome

- “benign” refers to prognosis, but some patients have subtle cognitive and behavioral disorders

- “idiopathic” refers to aetiology

- “generalized” : generalized seizure types and bilateral EEG abnormalities, but some patients have focal seizure clinical and EEG*

Epileptic syndrome

- Value in children greater than adults
- Some syndromes have specific pathology, aetiology
- Some specific pathologies result in a variety of syndromes
- Knowledge advances, syndromes reclassified, and distinction from disease becomes blurred – e.g. molecular genetics
The Task Force on Classification and Terminology of ILAE 2001

• **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.
The Task Force on Classification and Terminology of ILAE 2001

- **Axis 3: Syndrome**: from the List of Epilepsy Syndromes, not always possible

- **Axis 4: Etiology**, including specific genetic defects or pathologic substrates

- **Axis 5: Impairment**, optional but useful parameter can be derived from the WHO ICIDH-2 impairment classification
The 2006 ILAE report

Epilepsy Syndromes by Age of Onset and Related Conditions

<table>
<thead>
<tr>
<th>Neonatal period</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Benign familial neonatal seizures (BFNS)</td>
</tr>
<tr>
<td>• Early myoclonic encephalopathy (EME)</td>
</tr>
<tr>
<td>• Ohtahara syndrome</td>
</tr>
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<table>
<thead>
<tr>
<th>Infancy</th>
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</thead>
<tbody>
<tr>
<td>• Migrating partial seizures of infancy</td>
</tr>
<tr>
<td>• West syndrome</td>
</tr>
<tr>
<td>• Myoclonic epilepsy in infancy (MEI)</td>
</tr>
<tr>
<td>• Benign infantile seizures</td>
</tr>
<tr>
<td>• Dravet syndrome</td>
</tr>
<tr>
<td>• Myoclonic encephalopathy in nonprogressive disorders</td>
</tr>
</tbody>
</table>

Engel J Jr. Epilepsia 2006;47:1558-68.
The 2006 ILAE report

Epilepsy Syndromes by Age of Onset and Related Conditions

**Childhood**
- Early onset benign childhood occipital epilepsy (Panayiotopoulos type)
- Epilepsy with myoclonic astatic seizure
- Benign childhood epilepsy with centrotemporal spikes (BCECTS)
- Late-onset childhood occipital epilepsy (Gastaut type)
- Epilepsy with myoclonic absences
- Lennox-Gastaut syndrome
- Epileptic encephalopathy with continuous spike-and-wave during sleep (CSWS) including:
  - Landau-Kleffner syndrome (LKS)
  - Childhood absence epilepsy (CAE)

Engel J Jr. Epilepsia 2006;47:1558-68.
The 2006 ILAE report

### Epilepsy Syndromes by Age of Onset and Related Conditions

<table>
<thead>
<tr>
<th>Adolescence</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Juvenile absence epilepsy (JAE)</td>
</tr>
<tr>
<td>• Juvenile myoclonic epilepsy (JME)</td>
</tr>
<tr>
<td>• Progressive myoclonus epilepsies (PME)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Less-specific age relationship</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Autosomal-dominant nocturnal frontal lobe epilepsy (ADNFLE)</td>
</tr>
<tr>
<td>• Familial temporal lobe epilepsies</td>
</tr>
<tr>
<td>• Mesial temporal lobe epilepsy with hippocampal sclerosis (MTLE with HS)</td>
</tr>
<tr>
<td>• Rasmussen syndrome</td>
</tr>
<tr>
<td>• Gelastic seizures with hypothalamic hamartoma</td>
</tr>
</tbody>
</table>

Engel J Jr. Epilepsia 2006;47:1558-68.
The 2006 ILAE report

Epilepsy Syndromes by Age of Onset and Related Conditions

Special epilepsy conditions
- Symptomatic focal epilepsies not otherwise specified
- Epilepsy with generalized tonic-clonic seizures only
- Reflex epilepsies
- Febrile seizures plus (FS+)
- Familial focal epilepsy with variable foci

Conditions with epileptic seizures that do not require a diagnosis of epilepsy
- Benign neonatal seizures (BNS)
- Febrile seizures (FS)

Engel J Jr. Epilepsia 2006;47:1558-68.
Epileptic syndrome

Debate!

• “Splitters”
• “Lumpers”

? Different and separate syndromes or different facets of a broader neurobiological spectrum of epilepsy
SPECIAL REPORT


*†Anne T. Berg, ‡Samuel F. Berkovic, §Martin J. Brodie, ¶Jeffrey Buchhalter, #**J. Helen Cross, ††Walter van Emde Boas, †‡Jerome Engel, §§Jacqueline French, ¶¶Tracy A. Glauser, #Gary W. Mathern, ***Solomon L. Moshé, †Douglas Nordli, †††Perrine Plouin, and ‡Ingrid E. Scheffer

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2010 International League Against Epilepsy (ILAE) classification of seizure types

This classification excludes epileptic spasms

**Focal seizure**

- Describe the seizure in detail and specifically to include one or more of the following features:
  - aura
  - motor
  - autonomic
  - awareness/responsiveness: altered/dyscognitive / retained

**Generalised seizure**

- **Tonic-clonic**
  - Absence
    - Typical
    - Absence with special features
      - myoclonic absence
      - eyelid myoclonia
    - Atypical
- **Clonic**
- **Atonic**
- **Myoclonic**
  - myoclonic
  - myoclonic-atonic
  - myoclonic-tonic
The 2010 ILAE report

Revised terminology and concepts for organization of seizures and epilepsies

**Focal seizures**

- “originating within networks limited to one hemisphere
- may be discretely localized or more widely distributed and may originate in subcortical structures

Focal seizures

- For each seizure type, ictal onset is consistent from one seizure to another, with preferential propagation patterns that can involve the contralateral hemisphere.

- In some cases, more than one network, and more than one seizure type, but each individual seizure type has a consistent site of onset.

Generalized seizures

• “originating at some point within, and rapidly engaging, bilaterally distributed networks which include cortical and subcortical structures, but not necessarily include the entire cortex.

• Generalized seizures can be asymmetric

The 2010 ILAE report

<table>
<thead>
<tr>
<th>Generalized seizures</th>
<th>Focal seizures</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Tonic-clonic (in any combination)</td>
<td>• Epileptic spasms</td>
</tr>
<tr>
<td>• Absence</td>
<td></td>
</tr>
<tr>
<td>• Typical</td>
<td></td>
</tr>
<tr>
<td>• Atypical</td>
<td></td>
</tr>
<tr>
<td>• Absence with special features</td>
<td></td>
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<tr>
<td>• Myoclonic absence</td>
<td></td>
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<tr>
<td>• Eyelid myoclonia</td>
<td></td>
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<tr>
<td>• Myoclonic</td>
<td></td>
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<tr>
<td>• Myoclonic atonic</td>
<td></td>
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<tr>
<td>• Myoclonic tonic</td>
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<tr>
<td>• Clonic</td>
<td></td>
</tr>
<tr>
<td>• Tonic</td>
<td></td>
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<tr>
<td>• Atonic</td>
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</tbody>
</table>

*Seizures that cannot be clearly diagnosed into one of the preceding categories should be considered unclassified until further information allows their accurate diagnosis.

*This is not considered a classification category, however.
The 2010 ILAE report

Replaced the terms “idiopathic,” “symptomatic,” and “cryptogenic” with the following

• Genetic: “the direct result of a known or presumed genetic defect(s) in which seizures are the core symptom of the disorder, but the possibility that environmental factors contribute to expression of the disease is not excluded.”

• Structural-metabolic: “there is a distinct other structural or metabolic condition or disease that has been demonstrated to be associated with a substantially increased risk of developing epilepsy in appropriately designed studies.” These disturbances can be acquired or genetic (e.g., tuberous sclerosis).

• Unknown cause
<table>
<thead>
<tr>
<th>1981 ILAE Seizure types</th>
<th>2010 ILAE Seizure types</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Partial or Focal Seizures</strong></td>
<td><strong>Focal Seizures</strong></td>
</tr>
<tr>
<td>Simple partial seizures</td>
<td>Without impairment of consciousness or awareness with observable motor or autonomic components</td>
</tr>
<tr>
<td>Complex partial seizures</td>
<td>With impairment of consciousness or awareness. (“Dyscognitive”)</td>
</tr>
<tr>
<td>Partial with secondary generalization</td>
<td>Evolving to a bilateral, convulsive seizure</td>
</tr>
<tr>
<td><strong>Generalized Seizures</strong></td>
<td><strong>Generalized seizures</strong></td>
</tr>
<tr>
<td>Tonic clonic</td>
<td>Tonic clonic</td>
</tr>
<tr>
<td>Absence typical, Absence atypical</td>
<td>Absence typical, absence atypical</td>
</tr>
<tr>
<td></td>
<td>Absence with special features</td>
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<tr>
<td></td>
<td>Myoclonic absence</td>
</tr>
<tr>
<td></td>
<td>Absence with eyelid myoclonus</td>
</tr>
<tr>
<td><strong>1981 ILAE Seizure types (continued)</strong></td>
<td><strong>2010 ILAE Seizure types (continued)</strong></td>
</tr>
<tr>
<td>Myoclonus</td>
<td>Myoclonic</td>
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<tr>
<td></td>
<td>Myoclonic atonic</td>
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<td></td>
<td>Myoclonic tonic</td>
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<tr>
<td>Atonic/akinetict</td>
<td>Atonic</td>
</tr>
<tr>
<td>Tonic</td>
<td>Tonic</td>
</tr>
<tr>
<td>Spasms</td>
<td>Clonic</td>
</tr>
<tr>
<td><strong>Not clearly focal or generalized</strong></td>
<td>Spasms</td>
</tr>
</tbody>
</table>
# The 2010 ILAE report

<table>
<thead>
<tr>
<th>Terminology</th>
<th>Terms no longer recommended</th>
</tr>
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<tbody>
<tr>
<td>• Self-limited</td>
<td>• Benign</td>
</tr>
<tr>
<td>• Pharmacoresponsive</td>
<td>• Catastrophic</td>
</tr>
<tr>
<td>• Focal seizures</td>
<td>• Complex partial, Simple partial</td>
</tr>
<tr>
<td>• Evolving to a bilateral convulsive seizure</td>
<td>• Secondarily generalized</td>
</tr>
<tr>
<td>– eg. Tonic, clonic, tonic-clonic</td>
<td></td>
</tr>
</tbody>
</table>
The 2010 ILAE report
Electroclinical syndromes and other epilepsies

**Electroclinical syndromes arranged by age at onset**

**Neonatal period**
- Benign familial neonatal epilepsy (BFNE)
- Early myoclonic encephalopathy (EME)
- Ohtahara syndrome

**Infancy**
- Epilepsy of infancy with migrating focal seizures
- West syndrome
- Myoclonic epilepsy in infancy (MEI)
- Benign infantile epilepsy
- Benign familial infantile epilepsy
- Dravet syndrome
- Myoclonic encephalopathy in nonprogressive disorders
The 2010 ILAE report
Electroclinical syndromes and other epilepsies

**Childhood**
- Febrile seizures plus (FS+) (can start in infancy)
- Panayiotopoulos syndrome
- Epilepsy with myoclonic atonic (previously astatic) seizures
- Benign epilepsy with centrotemporal spikes (BECTS)
- Autosomal-dominant nocturnal frontal lobe epilepsy (ADNFLE)
- Late-onset childhood occipital epilepsy (Gastaut type)
- Epilepsy with myoclonic absences
- Lennox-Gastaut syndrome
- Epileptic encephalopathy with continuous spike-and-wave during sleep (CSWS)**
- Landau-Kleffner syndrome (LKS)
- Childhood absence epilepsy (CAE)

** Sometimes referred to as electrical status epilepticus during slow sleep (ESES)**
The 2010 ILAE report
Electroclinical syndromes and other epilepsies

**Distinctive constellations**
- Mesial temporal lobe epilepsy with hippocampal sclerosis (MTLE with HS)
- Rasmussen syndrome
- Gelastic seizures with hypothalamic hamartoma
- Hemiconvulsion-hemiplegia-epilepsy

Epilepsies that do not fit into any of these diagnostic categories can be distinguished firstly on the basis of the presence or absence of a known structural or metabolic condition (presumed cause), and then on the basis of the primary mode of seizure onset (generalized vs. focal)
The 2010 ILAE report
Electroclinical syndromes and other epilepsies

**Epilepsies attributed to and organized by structural-metabolic causes**
- Malformations of cortical development (hemimegalencephaly, heterotopias, etc.)
- Neurocutaneous syndromes (tuberous sclerosis complex, Sturge-Weber, etc.)
- Tumor
- Infection
- Trauma
- Angioma
- Perinatal insults
- Stroke
- Etc.

**Epilepsies of unknown cause**
Conditions with epileptic seizures that are traditionally not diagnosed as a form of epilepsy per se.
- Benign neonatal seizures (BNS)
- Febrile seizures (FS)
Definition

Epileptic seizure type

An ictal event believed to represent a unique pathophysiological mechanism and anatomical substrate.

This is a diagnostic entity with etiologic, therapeutic, and prognostic implications. (*new concept*)
Epilepsy syndrome

A complex of signs and symptoms that define a unique epilepsy condition with different etiologies.

This must involve more than just the seizure type; thus, frontal lobe seizures, for instance, do not constitute a syndrome. *(changed concept)*
Definition

Epilepsy disease

A pathologic condition with a single, specific, well-defined etiology. Thus, progressive myoclonus epilepsy is a syndrome, but Unverricht-Lundborg is a disease. (new concept)
Definition

Epileptic encephalopathy

A condition in which the epileptiform abnormalities themselves are believed to contribute to the progressive disturbance in cerebral function. (new concept)
Benign epilepsy syndrome

A syndrome characterized by epileptic seizures that are easily treated, or require no treatment, and remit without sequelae.

However, the use of this term is discouraged in the 2010 report because of new data revealing cognitive deficits in some of these conditions. (clarified concept)
**Definition**

**Reflex epilepsy syndrome**

A syndrome in which all epileptic seizures are precipitated by sensory stimuli.

Isolated reflex seizures can also occur in situations that do not necessarily require a diagnosis of epilepsy.

Seizures precipitated by other special circumstances, such as fever or alcohol withdrawal, are not reflex seizures. *(changed concept)*
Instruction manual for the ILAE 2017 operational classification of seizure types

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Epilepsia, **(*):1–12, 2017
doi: 10.1111/epi.13671

**SUMMARY**

This companion paper to the introduction of the International League Against Epilepsy (ILAE) 2017 classification of seizure types provides guidance on how to employ the classification. Illustration of the classification is enacted by tables, a glossary of relevant terms, mapping of old to new terms, suggested abbreviations, and examples. Basic and extended versions of the classification are available, depending on the desired degree of detail. Key signs and symptoms of seizures (semiology) are used as a basis for categories of seizures that are focal or generalized from onset or with unknown onset. Any focal seizure can further be optionally characterized by whether awareness is retained or impaired. Impaired awareness during any segment of the seizure renders it a focal impaired awareness seizure. Focal seizures are further optionally characterized by motor onset signs and symptoms: atonic, automatisms, clonic, epileptic spasms, or hyperkinetic, myoclonic, or tonic activity. Nonmotor-onset seizures can manifest as autonomic, behavior arrest, cognitive, emotional, or sensory dysfunction. The earliest prominent manifestation defines the seizure type, which might then progress to other signs and symptoms. Focal seizures can become bilateral tonic–clonic. Generalized seizures engage bilateral networks from onset. Generalized motor seizure characteristics comprise atonic, clonic, epileptic spasms, myoclonic, myoclonic–tonic, myoclonic–tonic–clonic, tonic, or tonic–clonic. Nonmotor (absence) seizures are typical or atypical, or seizures that present prominent myoclonic activity or eyelid myoclonia. Seizures of unknown onset may have features that can still be classified as motor, nonmotor, tonic–clonic, epileptic spasms, or behavior arrest. This “users’ manual” for the ILAE 2017 seizure classification will assist the adoption of the new system.

**KEY WORDS:** Classification, Seizures, Focal, Generalized, Epilepsy (taxonomy).
The 2017 ILAE Classification

- Level 1: Seizure type
- Level 2: Epilepsy based on seizure type
- Level 3: Epileptic syndrome
- Level 4: Epilepsy with etiology
# The New Basic Classification

Based on 3 key features

- Where seizures begin in the brain
- Level of awareness during a seizure
- Other features of seizures
The type of seizure onset affects choice of seizure medication, possibilities for epilepsy surgery, outlook, and possible causes.

1. Focal seizures: Previously called partial seizures, these start in an area or network of cells on one side of the brain.
Defining Where Seizures Begin

2. Generalized seizures: Previously called primary generalized
   - Engage or involve networks on both sides of the brain at the onset.

3. Unknown onset
   - Later on, the seizures type can be changed if the beginning of a person’s seizures becomes clear.
### Defining Where Seizures Begin

<table>
<thead>
<tr>
<th>4. Focal to bilateral seizure: Previously called partial seizures with secondary generalized seizures</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Now the term generalized refers only to the start of a seizure</td>
</tr>
<tr>
<td>- The new term for secondary generalized seizure would be a focal to bilateral seizure.</td>
</tr>
</tbody>
</table>
One of the main factors affecting a person’s safety during a seizure. Awareness is used instead of consciousness, because it is simpler to evaluate.

1. Focal aware seizure:
   - Intact awareness, even if unable to talk or respond during a seizure
   - This replaces the term simple partial.
## Describing Awareness

### 2. Focal impaired awareness seizure

- This replaces the term complex partial seizure.

### 3. Awareness unknown

- Sometimes it’s not possible to know if a person is aware or not
- For example if a person lives alone or has seizures only at night.
4. Generalized seizures:

- All presumed to affect a person’s awareness or consciousness in some way
- Thus no special terms are needed to describe awareness in generalized seizures.
ILAE 2017 Classification of Seizure Types Basic Version

Focal Onset
- Aware
- Impaired Awareness
- Motor Onset
- Nonmotor Onset
- focal to bilateral tonic–clonic

Generalized Onset
- Motor
  - Tonic–clonic
  - Other motor
  - Nonmotor (Absence)

Unknown Onset
- Motor
  - Tonic–clonic
  - Other motor
  - Nonmotor
- Unclassified

Epilepsia, 58(4):522–530, 2017
doi: 10.1111/epi.13670
Describing Motor and Other Symptoms in Focal Seizures

1. Focal motor seizure

- Some type of movement occurs during the event eg., twitching, jerking, or stiffening movements of a body part or automatisms (automatic movements such as licking lips, rubbing hands, walking, or running)
Describing Motor and Other Symptoms in Focal Seizures

2. Focal non-motor seizure

- Has other symptoms that occur first, such as changes in sensation, emotions, thinking, or experiences.

- Also possible for a focal aware or impaired awareness seizure to be sub-classified as motor or non-motor onset.
Describing Motor and Other Symptoms in Focal Seizures

3. Auras

- The term aura to describe symptoms a person may feel in the beginning of a seizure is not in the new classification.

- These early symptoms may be the start of a seizure.
Describing Generalized Onset Seizures

1. Generalized motor seizure

- The generalized tonic-clonic seizure term is still used to describe seizures with stiffening (tonic) and jerking (clonic)

- This loosely corresponds to “grand mal.”

- Many of these terms have not changed and a few new terms have been added
Describing Generalized Onset Seizures

2. Generalized non-motor seizure

- These are primarily absence seizures and the term corresponds to the old term “petit mal.”
- Involve brief changes in awareness, staring, and some may have automatic or repeated movements like lip smacking.
### Operational Classification of Seizure Types

#### ILAE 2017 Classification of Seizure Types Expanded Version

<table>
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<tr>
<th></th>
<th>Focal Onset</th>
<th>Generalized Onset</th>
<th>Unknown Onset</th>
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<tbody>
<tr>
<td></td>
<td>Aware</td>
<td>Impaired Awareness</td>
<td>Motor</td>
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<td>Motor Onset</td>
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<td>epileptic spasms</td>
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<td>focal to bilateral</td>
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Figure 1.
Epilepsia © ILAE
Some other points

- The new classification is designed to have some flexibility. Use of other descriptive terms or even free text is encouraged.

- Most seizures can be classified by signs and symptoms that happen during a seizure. However, other information is useful when available: phone videos, EEG, MRI, and other brain imaging, blood tests, or gene tests.
Definition and Classification

The ILAE strives to provide definitions for key concepts and classification schemes that will help the world epilepsy community in developing a common language to communicate effectively regarding the many facets of epilepsy. It is also hoped that the clear terminology, based on available evidence, will help develop treatments adapted to each condition.

Definition of Epilepsy 2014 (Final)

- PowerPoint presentation illustrating the 2014 Definition of Epilepsy

Operational Classification of Seizure Types by the International League Against Epilepsy (2017): Position paper of the ILAE Commission for Classification and Terminology

- Instruction manual for the ILAE 2017 Operational Classification of Seizure Types
- PowerPoint presentation illustrating the 2017 Classification of Seizure Types

ILAE Classification of the Epilepsies (2017): Position paper of the ILAE Commission for Classification and Terminology

- Commentary: A new classification is born, Sameer M. Zuberi and Emilio Perucca (2017)
- PowerPoint Presentation on the 2017 Classification of the Epilepsies

Other classification documents

Thank You for your attention