ILAE Revised Terminology for Organization of Seizures and Epilepsies 2011 - 2013

Classification of Seizures

Generalized seizures
Arising within and rapidly engaging bilaterally distributed networks

Focal seizures
Originating within networks limited to one hemisphere

May evolve to
Bilateral convulsive seizure

Electroclinical Syndromes and Other Epilepsies Grouped by Specificity of Diagnosis

Electroclinical syndromes

One example of how syndromes can be organized: Arranged by typical age at onset*
(Syndromes unchanged except for minor changes in terminology)

Neonatal period
- Self limited neonatal seizures *
- Self limited familial neonatal epilepsy
- Ohtahara syndrome
- Early Myoclonic encephalopathy (EME)

Infancy
- Febrile seizures*, Febrile seizures plus (FS+)
- Self limited infantile epilepsy
- Self limited familial infantile epilepsy
- West syndrome
- Dravet syndrome
- Myoclonic epilepsy in infancy (MEI)
- Myoclonic encephalopathy in nonprogressive disorders
- Epilepsy of infancy with migrating focal seizures

Childhood
- Febrile seizures*, Febrile seizures plus (FS+)
- Early onset childhood occipital epilepsy (Panayiotopoulos syndrome)
- Epilepsy with myoclonic atonic (previously astatic) seizures
- Childhood absence epilepsy (CAE)
- Self limited epilepsy with centrotemporal spikes (ECTS)
- 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)

Adolescence - Adult
- Juvenile absence epilepsy (JAE)
- Juvenile myoclonic epilepsy (JME)
- Epilepsy with generalized tonic-clonic seizures alone (GTCA)
- Autosomal dominant epilepsy with auditory features (ADEAF)
- Other familial temporal lobe epilepsies

Familial Epilepsy Syndromes
- Familial focal epilepsy with variable foci (childhood to adult)
- Reflex epilepsies
- Genetic epilepsy with febrile seizures plus (GEFS+)

* The arrangement of electroclinical syndromes does not reflect etiology
* Sometimes referred to as Electrical Status Epilepticus during Slow Sleep (ESES)
* Not traditionally diagnosed as epilepsy

Unknown
Insufficient evidence to classify as focal, generalized or both

- Epileptic Spasms
- Other

- Tonic-Clonic
- Absence
- Clonic
- Tonic
- Atonic

Mycloonic
- Myoclonic
- Myoclonic-atonic
- Myoclonic-tonic

Awareness/Responsiveness:
altered (dyscognitive) or retained

- Aura
- Motor
- Autonomic

Electroclinical Syndromes
- Other

Bilateral convulsive seizure

- Typical
- Absence with special features
  - Myoclonic absence
  - Eyelid Myoclonia

- Atypical

- Typical
## ILAE Revised Terminology for Organization of Seizures and Epilepsies 2011 - 2013

### Major changes in terminology and concepts

<table>
<thead>
<tr>
<th>New Term and Concept</th>
<th>Examples</th>
<th>Old Term and Concept</th>
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</thead>
<tbody>
<tr>
<td><strong>Etiology (an individual may fit into more than one group)</strong></td>
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<td><strong>Genetic:</strong> genetic defect directly contributes to the epilepsy and seizures are the core symptom of the disorder</td>
<td>Channelopathies, GLUT1 deficiency, etc</td>
<td><strong>Idiopathic:</strong> presumed genetic</td>
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<tr>
<td><strong>Structural:</strong> caused by a structural disorder of the brain</td>
<td>Tuberous sclerosis, cortical malformations, mesial temporal lobe epilepsy with hippocampal sclerosis (MTLE with HS), gelastic seizures with hypothalamic hamartoma</td>
<td><strong>Symptomatic:</strong> secondary to a known or presumed disorder of the brain</td>
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<td><strong>Metabolic:</strong> caused by a metabolic disorder of the brain</td>
<td>Pyridoxine deficiency, GLUT1 deficiency, etc</td>
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<td><strong>Immune:</strong> epilepsy with evidence of autoimmune mediated CNS inflammation</td>
<td>NMDA receptor antibody encephalitis, voltage gated potassium channel antibody encephalitis</td>
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<td><strong>Infectious:</strong> an infectious etiology refers to a patient with epilepsy, rather than seizures occurring in the setting of acute infection such as meningitis or encephalitis. These infections sometimes have a structural correlate.</td>
<td>Tuberculosis, HIV, cerebral malaria, neurocysticercosis, subacute sclerosing panencephalitis, cerebral toxoplasmosis</td>
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<td><strong>Unknown:</strong> the cause of epilepsy is unknown</td>
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<td><strong>Cryptogenic:</strong> presumed symptomatic</td>
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</tbody>
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### Terms no longer recommended

- **Self-limited:** tendency to resolve spontaneously over time
- **Pharmacoresponsive:** highly likely to be controlled with medication
- **Focal seizures:** seizure semiology described according to specific subjective (auras), motor, autonomic, and dyscognitive features
- **Evolving to a bilateral convulsive seizure**

**Benign**

**Catastrophic**

**Complex Partial**

**Simple Partial**

**Secondary generalized**

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*We would welcome your thoughts on this proposal. Please visit the “Request for Comments” page on the ILAE website to read the full document and register your comments.*

[http://www.ilae.org/Visitors/Center/Organization.cfm](http://www.ilae.org/Visitors/Center/Organization.cfm)

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**References:**