Electroclinical Syndromes and Other Epilepsies Grouped by Specificity of Diagnosis

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
- Epileptiform 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+)**

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

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Electroclinical Syndromes

**Generalized seizures**
- Tonic-Clonic
- Absence
- Clonic
- Tonic
- Atonic

**Focal seizures**
- Myoclonic
  - Myoclonic
  - Myoclonic-atonic
  - Myoclonic-tonic

**Unknown**
- Insufficient evidence to classify as focal, generalized or both
  - Epileptic Spasms
  - Other

**Bilateral convulsive seizure**

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Classification of Seizures
**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</strong> (an individual may fit into more than one group)</td>
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<tr>
<td>Genetic: genetic defect directly contributes to the epilepsy and seizures are the core symptom of the disorder</td>
<td>Channelopathies, GLUT1 deficiency, etc</td>
<td>Idiopathic: presumed genetic</td>
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<tr>
<td>Structural: 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>Symptomatic: secondary to a known or presumed disorder of the brain</td>
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<tr>
<td>Metabolic: caused by a metabolic disorder of the brain</td>
<td>Pyridoxine deficiency, GLUT1 deficiency, etc</td>
<td>Symptomatic</td>
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<tr>
<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>
<td><strong>Cryptogenic</strong>: presumed symptomatic</td>
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<td><strong>Unknown</strong>: the cause of epilepsy is unknown</td>
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**Terms no longer recommended**

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

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**References:**


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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)