ILAE Proposal for Revised Terminology for Organization of Seizures and Epilepsies 2010

**Classification of Seizures**

**Generalized seizures**
- Arising within and rapidly engaging bilaterally distributed networks

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

**Myoclonic**
- Myoclonic
- Myoclonic-atonic
- Myoclonic-tonic

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

**Atypical**

**Focal seizures**
- Originating within networks limited to one hemisphere

**Characterized according to one or more features:**
- Aura
- Motor
- Autonomic
- Awareness/Responsiveness: altered (dyscognitive) or retained

**May evolve to:**
- Bilateral convulsive seizure

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

**Changes in terminology and concepts**

<table>
<thead>
<tr>
<th>Etiology</th>
<th>New Term and Concept</th>
<th>Examples</th>
<th>Old Term and Concept</th>
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</thead>
<tbody>
<tr>
<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>Idiopathic: presumed genetic</td>
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<td><strong>Structural-metabolic:</strong> caused by a structural or metabolic disorder of the brain</td>
<td>Tuberous sclerosis, cortical malformations, etc.</td>
<td>Symptomatic: secondary to a known or presumed disorder of the brain</td>
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<tr>
<td><strong>Unknown:</strong> the cause is unknown and might be genetic, structural or metabolic</td>
<td>Cryptogenic: presumed symptomatic</td>
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<thead>
<tr>
<th>Terminology</th>
<th>Terms no longer recommended</th>
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<tbody>
<tr>
<td><strong>Self-limited:</strong> tendency to resolve spontaneously with time</td>
<td>Benign</td>
</tr>
<tr>
<td><strong>Pharmacoresponsive:</strong> highly likely to be controlled with medication</td>
<td>Catastrophic</td>
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<tr>
<td><strong>Focal seizures:</strong> seizure semiology described according to specific subjective (auras), motor, autonomic, and dyscognitive features</td>
<td>Complex partial</td>
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<tr>
<td><strong>Evolving to a bilateral convulsive seizure:</strong> eg. tonic, clonic, tonic-clonic</td>
<td>Simple partial</td>
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<tr>
<td><strong>Secondarily generalized</strong></td>
<td>Secondary generalized</td>
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**References:**
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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

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

- **Infancy**
  - Febrile seizures, Febrile seizures plus (FS+)
  - Benign infantile epilepsy
  - Benign familial infantile epilepsy (BFIE)
  - 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 atactic) seizures
  - Childhood absence epilepsy (CAE)
  - Benign epilepsy with centrotemporal spikes (BECS)
  - Autosomal dominant nocturnal frontal lobe epilepsy (ADNFLE)
  - Late onset childhood occipital epilepsy (Gastaut type)
  - Epilepsy with myoclonic absences
  - Lennox-Gastaut syndrome (LGS)
  - Epileptics 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
  - Autosomal dominant epilepsy with auditory features (ADEAF)
  - Other familial temporal lobe epilepsies

- **Variable age at onset**
  - Familial focal epilepsy with variable foci (childhood to adult)
  - Progressive myoclonus epilepsies (PME)
  - Reflex epilepsies

**Distinctive constellations/surgical syndromes**

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

**Nonsyndromic 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, antenatal and perinatal insults, stroke, etc

**Epilepsies of unknown cause**

* The arrangement of electroclinical syndromes does not reflect etiology,
^ Not traditionally diagnosed as epilepsy
^+ Sometimes referred to as Electrical Status Epilepticus during Slow Sleep (ESES)
** Forms of epilepsies not meeting criteria for specific syndromes or constellations

This Proposal is a work in progress.....
We welcome your thoughts on this proposal. Please visit our Classification & Terminology Discussion Group at: [http://community.ilae-epilepsy.org/home/](http://community.ilae-epilepsy.org/home/) to login and register your comments.