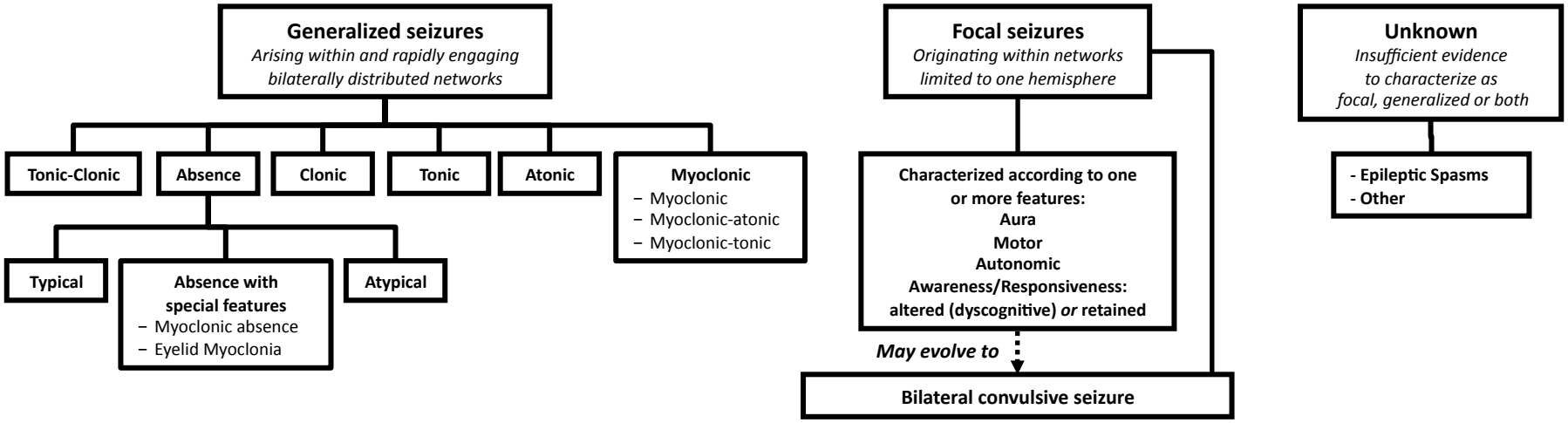
ILAE Proposal for Revised Terminology for Organization of Seizures and Epilepsies 2010

Classification of Seizures



Changes in terminology and concepts

New Term and Concept	Examples	Old Term and Concept
Etiology		
Genetic: *genetic defect directly contributes to the epilepsy and seizures are the core symptom of the disorder*	*Channelopathies, Glut1 deficiency, etc.*	**Idiopathic:** *presumed genetic*
Structural-metabolic: *caused by a structural or metabolic disorder of the brain*	*Tuberous sclerosis, cortical malformations, etc.*	**Symptomatic:** *secondary to a known or presumed disorder of the brain*
Unknown: *the cause is unknown and might be genetic, structural or metabolic*		**Cryptogenic:** *presumed symptomatic*
Terminology		**Terms no longer recommended**
Self-limited: *tendency to resolve spontaneously with time* **Pharmacoresponsive:** *highly likely to be controlled with medication*		**Benign** **Catastrophic**
Focal seizures: *seizure semiology described according to specific subjective (auras), motor, autonomic, and dyscognitive features*		**Complex partial** **Simple partial**
Evolving to a bilateral convulsive seizure: *eg. tonic, clonic, tonic-clonic*		**Secondarily generalized**

References: 1.Berg AT et al. Revised terminology and concepts for organization of seizures and epilepsies: report of the ILAE Commission on Classification and Terminology, 2005-2009. *Epilepsia* 2010;51:676-685. 2.Berg AT, Cross JH. *Lancet* 2010:9;459-61. 3.Blume WT et al. Glossary of descriptive terminology for ictal semiology: Report of the ILAE task force on classification and terminology. *Epilepsia* 2001:42;1212-1218.

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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 astatic) seizures
- Childhood absence epilepsy (CAE)
- 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 (LGS)
- 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
- 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

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 eletroclinical 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/ to login and register your comments.