New Horizon in Epilepsy

Update in Classification for Seizure and Epilepsy

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Why classification is needed ?

Classification :

- A universal vocabulary that facilitated communication among clinicians
- Also established a taxonomy foundation for the research on epilepsy

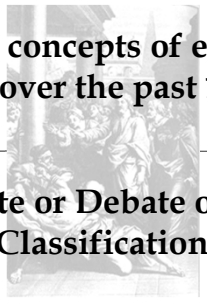
In general

Types of Classification

- Biology:
- Etiology: 1^0 (idiopathic) or 2^0 (symptomatic)
- Pathology: Cancer
- Imaging: Cortical dysplasia
- Clinical criteria e.g. age onset, disease course, distribution of symptoms: HA

- Mixed:

Changing concepts of etiology of epilepsy over the past 150 years

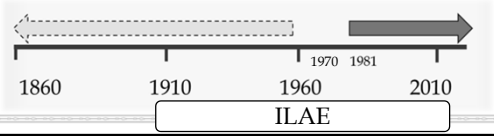


Update or Debate on the Classification

ILAE

International League Against Epilepsy

100^{th} anniversary of ILAE in 2009



Commission on Classification and Terminology of ILAE

- Classification of Epileptic Seizures in 1981
- Classification of Epilepsies and Epileptic syndromes in 1989

- A proposed diagnostic scheme for people with epileptic seizures and with epilepsy : Report of the ILAE Task Force on Classification and Terminology in 2001
- ….2006,……2010, …..2011,…..

At present

Terminology

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

Seizure, Greek meaning, to take hold

At present

Terminology

- **Epilepsy**: a disorder of the brain characterized by an enduring predisposition to generate epileptic seizures, and by neurobiologic, cognitive, psychological and social consequences of the condition.
- The definition of epilepsy requires the occurrence of at least "*one epileptic seizure*"

The classification criteria of

Epilepsies (Merlis 1970)	Epileptic seizures (Gastuat 1970)
Clinical criteria	Clinical seizure type
Seizures	EEG seizure type
Neurologic status	EEG interictal expression
Age of onset	Anatomical substrate
Etiology	Etiology
EEG criteria	Age
Interictal EEG	
Ictal EEG	

Human EEG 1929, Epileptic spike 1934, 3 Hz SW 1935, 1st EEG lab 1936

ILAE 1981

Clinical seizure type | EEG sz type | EEG interictal expression

1. Partial (focal, local) seizures
   - Simple partial sz
     - with motor signs
     - with somatosensory symptoms
     - with autonomic symptoms and signs
     - with psychic symptoms
   - Complex partial sz
     - start with SPS followed by impairment of consciousness
     - with impairment of consciousness at onset
   - Partial sz evolving to 2^0 gen sz
     - SPS → GTC
     - CPS → GTC
     - SPS → CPS → GTC
2. Generalized sz (convulsive and non-convulsive)
   Absence, Myoclonic, Clonic, Tonic, Tonic-clonic, Atonic
3. Unclassified epileptic sz
4. Prolonged or repetitive seizure (status epilepticus)

Classification of Epilepsies and Epileptic syndromes in 1989

- The term partial is replaced with " localization-related " but after 2001 returns to use the same word-focal
- Term : ***Idiopathic, Cryptogenic, Symptomatic*** is introduced

Definition

- ***Idiopathic*** = a disorder unto itself, sui generis, and not etiology unknown
- ***Cryptogenic*** = designate conditions that are not idiopathic, or are presumed to be symptomatic, when the etiology has not been determined, some conditions that not known whether they are idiopathic or symptomatic

ILAE 1989

1. Localization-related epilepsies and syndromes

1.1 Idiopathic
- benign childhood epilepsy with centro-temporal spike
- childhood epilepsy with occipital paroxysms
- primary reading epilepsy

1.2 Symptomatic e.g. TLE, FLE, PLE, OLE

1.3 Cryptogenic

ILAE 1989

2. Generalized epilepsies and syndromes

2.1 Idiopathic (with age-related onset, listed in order to age)
- Benign neonatal familial convulsions
- Benign neonatal convulsions
- Benign myoclonic epilepsy of infancy
- Childhood absence epilepsy (pyknolepsy)
- Juvenile absence epilepsy
- Juvenile myoclonic epilepsy
- Epilepsy w grand mal (GTCS) sz on awakening
- etc.

2.2 Cryptogenic or symptomatic (in order to age)
- West syndrome
- Lennox-Gastaut syndrome
- Epilepsy w myoclonic-astatic sz
- Epilepsy w myoclonic absences

ILAE 1989

2. Generalized epilepsies and syndromes

2.3 Symptomatic
2.3.1 Non specific etiology
- EME
- EIEE w supression burst
- other symptomatic generalised epilepsies not defined above

2.3.2 Specific syndromes/etiologies
- Cerebral malformation
- IBEM

3. Epilepsies and syndromes undetermined whether focal or generalized e.g. SMEI, LKS, CSWS, neonatal sz

4. Special syndromes e.g. FC, reflex epilepsy, isolated sz

Epileptic syndromes

- a cluster of S&S occurring together
- including :
  - : Type of seizures
  - : Age of onset
  - : Etiology
  - : Anatomy
  - : Precipitating factors
  - : Severity
  - : Chronicity
  - : Prognosis
  - : Diurnal and circadian cycling

1989

Two dichotomies, a 4-part classification

Cryptogenic
Special syndromes

	Localization-related	**Generalized**
Idiopathic	Localization-related Idiopathic e.g. BRE	Generalized Idiopathic e.g. CAE, JAE, JME
Symptomatic	Localization-related Symptomatic e.g. TLE, FLE	Generalized Symptomatic e.g. LGS, West synd

The reason for 2001 diagnostic scheme

- Using the term of complex partial seizure, in the past , sometime confuses with temporal lobe seizures
- The division in the past created that impairment of consciousness had certain mechanism (limbic system)
- Replaced the term <u>febrile convulsion</u> by <u>febrile seizures</u>

The reason for 2001 diagnostic scheme

- Misunderstanding of the correct definition of idiopathic, cryptogenic.
- But the Task Force believes that most epileptologists have now learned to use the term correctly and that there is value in maintaining continuity.
- Great believe that in the near future, genetic classifications of certain epilepsy syndromes will become possible.

Diagnostic scheme for people with epileptic sz and with epilepsy: 2001

- ***Axis 1*** : **Ictal phenomenology**-describe ictal events
- ***Axis 2*** : **Seizure type**, from the list of epileptic seizures.
- ***Axis 3*** : **Syndrome**, from the list of epilepsy syndromes
- ***Axis 4*** : **Etiology**
- ***Axis 5*** : **Impairment**

Epilepsia 2001; 42:796-803

Table 4
Epilepsy syndromes and related conditions

Benign familial neonatal seizures
Early myoclonic encephalopathy
Ohtahara syndrome
[a]Migrating partial seizures of infancy
West syndrome
Benign myoclonic epilepsy in infancy
Benign familial infantile seizures
Benign infantile seizures (non-familial)
Dravet's syndrome
HHE syndrome
[a]Myoclonic status in non-progressive encephalopathies
Benign childhood epilepsy with centrotemporal spikes
Early onset benign childhood occipital epilepsy (Panayiotopoulos type)
Late onset childhood occipital epilepsy (Gastaut type)
Epilepsy with myoclonic absences
Epilepsy with myoclonic-astatic seizures
Lennox–Gastaut syndrome
Landau–Kleffner syndrome
Epilepsy with continuous spike-and-waves during slow-wave sleep (other than LKS)
Childhood absence epilepsy
Progressive myoclonus epilepsies

Idiopathic generalized epilepsies with variable phenotypes
  Juvenile absence epilepsy
  Juvenile myoclonic epilepsy
  Epilepsy with generalized tonic-clonic seizures only
Reflex epilepsies
  Idiopathic photosensitive occipital lobe epilepsy
  Other visual sensitive epilepsies
  Primary reading epilepsy
  Startle epilepsy
Autosomal dominant nocturnal frontal lobe epilepsy
Familial temporal lobe epilepsies
[a]Generalized epilepsies with febrile seizures plus
[a]Familial focal epilepsy with variable foci
Symptomatic (or probably symptomatic) focal epilepsies
  Limbic epilepsies
    Mesial temporal lobe epilepsy with hippocampal sclerosis
    Mesial temporal lobe epilepsy defined by specific etiologies
    Other types defined by location and etiology
  Neocortical epilepsies
    Rasmussen syndrome
    Other types defined by location and etiology
Conditions with epileptic seizures that do not require a diagnosis of epilepsy
  Benign neonatal seizures
  Febrile seizures
  Reflex seizures
  Alcohol-withdrawal seizures
  Drug or other chemically induced seizures
  Immediate and early post cerebral insult seizures
  Single seizures or isolated clusters of seizures
  Rarely repeated seizures (oligoepilepsy)

Epilepsia 2001, Epilepsy research 2006

Table 1
Definitions of key terms

Epilepsy syndrome: A complex of signs and symptoms that define a unique epilepsy condition. This must involve more than just the seizure type: thus frontal lobe seizures *per se*, for instance, do not constitute a syndrome. (changed concept)

sequelae (clarified concept).
Reflex epilepsy syndrome: A syndrome in which all epileptic seizures are precipitated by sensory stimuli. Reflex seizures that occur in focal and generalized epilepsy syndromes that are also associated with spontaneous seizures are listed as seizure types. 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).
Focal seizures and syndromes: Replaces the terms partial seizures and localization-related syndromes (changed terms).
Simple and complex partial epileptic seizures: These terms are no longer recommended, nor will they be replaced. Ictal impairment of consciousness will be described when appropriate for individual seizures, but will not be used to classify specific seizure types (new concept)
Idiopathic epilepsy syndromes: A syndrome that is only epilepsy, with no underlying structural brain lesion or other neurological signs or symptoms. These are presumed to be genetic and are usually age-dependent (unchanged term).
Symptomatic epilepsy syndrome: A syndrome in which the epileptic seizures are the result of one or more identifiable structural lesions of the brain (unchanged term).
Probably symptomatic epilepsy syndrome: Synonymous with, but preferred to, the term cryptogenic, used to define syndromes that are believed to be symptomatic, but no etiology has been identified (new term).

Epilepsia 2001, Epilepsy research 2006

Comment for 2001 diagnostic scheme

- A syndromic diagnosis cannot be made in many pts with epilepsy
- But the percentage of pts for whom a syndromic diagnosis is not possible has varied from one study to another
- Syndromic diagnoses are most useful in pediatric epileptology esp in children with idiopathic epilepsies but in focal symptomatic epilepsy is in doubt
- Difficult to make syndromic diagnoses at the time they appear with new onset epilepsy

Difference between the approach : Cleveland Clinic and ILAE diagnostic scheme

Cleveland 2005 (a patient-oriented approach)		**ILAE** 2001 (a category-based approach)	
Dimension 1	Epilepsy localization (EZ)	*Axis 1*	Ictal phenomenology
Dimension 2	Seizure semiology	*Axis 2*	Seizure type
Dimension 3	Etiology	*Axis 3*	Syndrome
Dimension 4	Seizure frequency	*Axis 4*	Etiology
Dimension 5	Related medical condition if applicable	*Axis 5*	Impairment

comment

Epilepsy ??

Genotypic heterogenity	Phenotypic heterogenity
Several genes – one syndrome	genetic factors
several epileptic syndromes – can occur same family	environment factor

Syndromes are man-made constructs develops to help consolidate large amounts of information into a form that can be used diagnostically and as a guide to Mx and prognosis

There is no right or wrong, apart from establishing what is the most useful system for the job in hand

Changes in classification

Subject to social and political circumstance

Lumper **Splitter**

Published paper **2009**

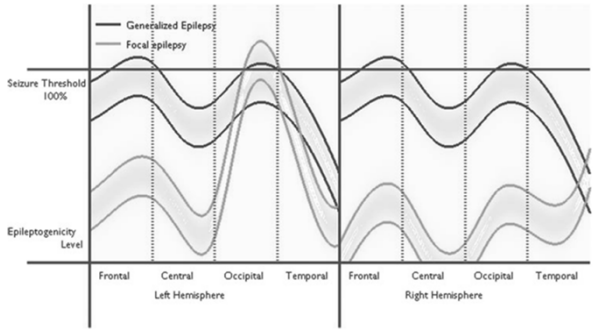


Figure 2.
Epileptogenicity level of a patient with left occipital epilepsy and a patient with generalized epilepsy.
Epilepsia © ILAE

2005-2009

ILAE classification working group

Epilepsia, 51(4):676-685, 2010
doi: 10.1111/j.1528-1167.2010.02522.x

SPECIAL REPORT

Revised terminology and concepts for organization of seizures and epilepsies: Report of the ILAE Commission on Classification and Terminology, 2005–2009

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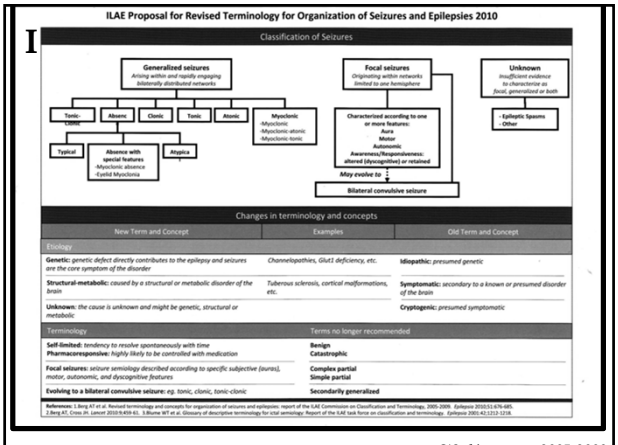
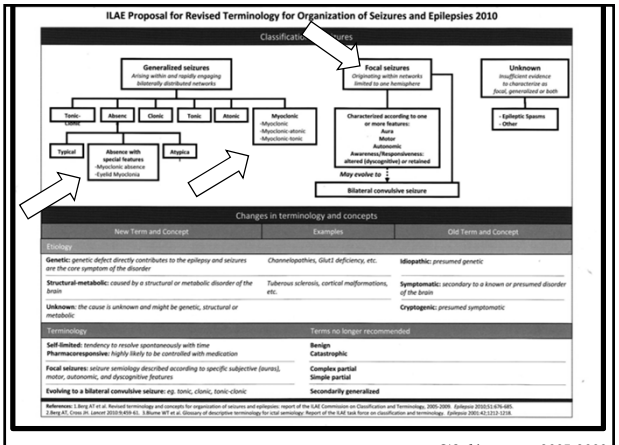


Working group 2005-2009



Working group 2005-2009

old New

Table 1. Comparison of major changes between the 1989 and 1981 Classification and Terminology and the newly proposed Terminology and Concepts (Commission 1981, 1989; Berg et al., 2010)

Old terminology and concepts	Recommended new terminology and concepts
Focal and generalized	
For seizures	
Focal (previously "partial"): the first clinical and electroencephalographic changes indicate initial activation of a system of neurons limited to a part of one cerebral hemisphere	Focal seizures are conceptualized as originating at some point within networks limited to one hemisphere
Generalized: the first clinical changes indicate initial involvement of both hemispheres	Generalized seizures are conceptualized as originating at some point within and rapidly engaging bilaterally distributed networks
For epilepsies	
Localization-related (focal, partial): epilepsies with focal seizures	These terms were abandoned as overarching categories for classifying epilepsies per se, as many syndromes include both seizure types; they may still apply in some but not all instances
Generalized: epilepsies with generalized seizures	
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**

focal seizure should be described according to their manifestation

old New

Table 1. Comparison of major changes between the 1989 and 1981 Classification and Terminology and the newly proposed Terminology and Concepts (Commission 1981, 1989; Berg et al., 2010)

Old terminology and concepts	Recommended new terminology and concepts
	Etiology
Idiopathic: there is no underlying cause other than a possible hereditary predisposition	Genetic: the epilepsy is, as best as understood, the direct result of a known or presumed genetic defect(s) in which seizures are the core symptom of the disorder. This attribution must be supported by specific forms of evidence
Symptomatic: the epilepsy is the consequence of a known or suspected disorder of the central nervous system	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. These disorders may be of acquired or genetic origin. When of genetic origin, there is a separate disorder interposed between the gene defect and the epilepsy
Cryptogenic: this refers to a disorder whose cause is hidden or occult. Cryptogenic epilepsies are presumed to be symptomatic	Unknown: the nature of the underlying cause is unknown; it may have a fundamental genetic basis (e.g., a previously unrecognized channelopathy) or it may be the consequence of an unrecognized structural or metabolic disorder not yet identified

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*

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, Absenc, Clonic, Tonic, Atonic, Myoclonic (-Myoclonic, -Myoclonic-atonic, -Myoclonic-tonic); Typical, Absence with, Atypica

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

ILAE 1989

2. Generalized epilepsies and syndromes

2.1 Idiopathic (with age-related onset, listed in order to age)
- Benign neonatal familial convulsions
- Benign neonatal convulsions
- Benign myoclonic epilepsy of infancy
- Childhood absence epilepsy (pyknolepsy)
- Juvenile absence epilepsy
- Juvenile myoclonic epilepsy
- Epilepsy w grand mal (GTCS) sz on awakening
- etc.

2.2 Cryptogenic or symptomatic (in order to age)
- West syndrome
- Lennox-Gastaut syndrome
- Epilepsy w myoclonic-astatic sz
- Epilepsy w myoclonic absences

ogy and concepts

Examples	Old Term and Concept
, Glut1 deficiency, etc.	**Idiopathic:** *presumed genetic*
ii, cortical malformations,	**Symptomatic:** *secondary to a known or presumed disorder of the brain*
	Cryptogenic: *presumed symptomatic*

Terms no longer recommended

Benign Catastrophic

Complex partial Simple partial

Secondarily generalized

Working group 2005-2009

II

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

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 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 Discussion Group at: http://community.ilae-epilepsy.org/home/ to login and register your comments.

Working group 2005-2009

Comment against proposal 2010

- Clarifying their meaning rather than replacing them would be sufficient : *Ferrie 2010*
- New terminology and concepts instead of proposing a new classification : *Panayiotopoulos 2011*
- The change of concepts is not reflected by any change in terminology: *Wolf 2010*

Comment against proposal 2010

- Free text descriptions of the numerous types of focal epileptic seizures are fine in a manual of differential diagnosis but should not be used as a classification system and makes futures research, epidermiology and comparisons difficult : *Fisher 2010*
- Dose not agree with " focal seizure that be described according to their manifestation" : *Panayiotopoulos 2011*

Comment against proposal 2010

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*

- Genetic epilepsy : some genetic also made structural brain defect → TSC linked with chromosome abnormalitis therefor TSC should be in genetic category or Structural-metabolic category ??
- Structural-Metabolic: ??

2011

Simon D Shorvon has published

" the etiology classification of epilepsy "

Why more ?

The diagnostic process in clinical practice comprises 2 stages

1. The classification of seizure type/syndrome
2. The assessment of cause

Aim: a framework for a database of etiology
: etiology dimension is only one of the aspects of a classification

The etiology classification of epilepsy

Table 1. Suggested scheme for an etiological classification of epilepsy

Main category	Subcategory	Examples[a]
Idiopathic epilepsy (1)	Pure epilepsies due to single gene disorders	Benign familial neonatal convulsions; autosomal dominant nocturnal frontal lobe epilepsy; generalized epilepsy with febrile seizures plus; severe myoclonic epilepsy of childhood; benign adult familial myoclonic epilepsy
	Pure epilepsies with complex inheritance	Idiopathic generalized epilepsy (and its subtypes); benign partial epilepsies of childhood
Symptomatic epilepsy		
Predominately genetic or developmental causation (2)	Childhood epilepsy syndromes	West syndrome; Lennox-Gastaut syndrome
	Progressive myoclonic epilepsies	Unverricht-Lundborg disease; Dentato-rubro-pallido-luysian atrophy; Lafora body disease; mitochondrial cytopathy; sialidosis; neuronal ceroid lipofuscinosis; myoclonus renal failure syndrome
	Neurocutaneous syndromes (2.1)	Tuberous sclerosis; neurofibromatosis; Sturge-Weber syndrome
	Other neurologic single gene disorders (2.1)	Angelman syndrome; lysosomal disorders; neuroacanthocytosis; organic acidurias and peroxisomal disorders; prophyria; pyridoxine-dependent epilepsy; Rett syndrome; Urea cycle disorders; Wilson disease; disorders of cobalamin and folate metabolism
	Disorders of chromosome function	Down syndrome; Fragile X syndrome; 4p-syndrome; isodicentric chromosome 15; ring chromosome 20
	Developmental anomalies of cerebral structure	Hemimegalencephaly; focal cortical dysplasia; agyria-pachygyria-band spectrum; agenesis of corpus callosum; polymicrogyria; schizencephaly; periventricular nodular heterotopia; microcephaly; arachnoid cyst

Shorvon, Epilepsia 2011

The etiology classification of epilepsy

Table 1. Suggested scheme for an etiological classification of epilepsy

Main category	Subcategory	Examples[a]
Predominately acquired causation (2.2)	Hippocampal sclerosis	Hippocampal sclerosis
	Perinatal and infantile causes	Neonatal seizures; postneonatal seizures; cerebral palsy; vaccination and immunization
	Cerebral trauma	Open head injury; closed head injury; neurosurgery; epilepsy after epilepsy surgery; nonaccidental head injury in infants
	Cerebral tumor	Glioma; ganglioglioma and hamartoma; DNET; hypothalamic hamartoma; meningioma; secondary tumors
	Cerebral infection	Viral meningitis and encephalitis; bacterial meningitis and abscess; malaria; neurocysticercosis, tuberculosis; HIV
	Cerebrovascular disorders	Cerebral hemorrhage; cerebral infarction; degenerative vascular disease; arteriovenous malformation; cavernous hemangioma
	Cerebral immunologic disorders	Rasmussen encephalitis; SLE and collagen vascular disorders; inflammatory and immunologic disorders
	Degenerative and other neurologic conditions	Alzheimer disease and other dementing disorders; multiple sclerosis and demyelinating disorders; hydrocephalus and porencephaly
Provoked epilepsy (3)	Provoking factors	Fever; menstrual cycle and catamenial epilepsy; sleep-wake cycle; metabolic and endocrine-induced seizures; drug-induced seizures; alcohol and toxin-induced seizures
	Reflex epilepsies	Photosensitive epilepsies; startle-induced epilepsies; reading epilepsy; auditory-induced epilepsy; eating epilepsy; hot-water epilepsy
Cryptogenic epilepsies[b] (4)		

Shorvon, Epilepsia 2011

Some comments

J Engel agree with Shorvon

1. about the seperation symptomatic cause to genetic/acquired therefore can get rid of structural-metabolic term
2. not agree to include provoked/cryptogenic here, should describe seperately
3. suggest category of surgical remediable

- Berg said that Shorvon's table creates confusion in identifying clinical syndrome as cause

Compromised Approaches

Two-tiered classification *M Wong 2011*

- Categorized according to both semiology/syndrome and causes
- **First Tier**: Describe the most specific syndrome/sz
- **Second Tier**: Identified etiologies or unknown is specified

Epilepsy syndrome X secondary to Cause Y

- e.g. JME secondary to *GABRA1* mutation

JME secondary to unknown cause

JME secondary to a malformation of a cortical development

Compromised Approaches

Two-tiered classification *M Wong 2011*

- Nonsyndromic epilepsy→ Symptom based classification

e.g. Epilepsy with partial/focal seizure secondary to cause Y

- Multifactorial causes

e.g. Epilepsy syndrome X secondary to traumatic brain injury and SCNA1 mutation

Update Comment: March 2012

Panayiotopoulos CP **Disagree that…...**

1. 2010 ILAE commission abandoned the disease-syndrome distinction
2. 2010 ILAE commission indicated the concepts of generalized and focal were no longer applicable with respect to electroclinical syndromes

? Epilepsy with GTC only

? IGE: CAE

? Familial (AD) focal epilepsy

Update Comment: March 2012

Panayiotopoulos CP **Disagree that….**

3. 2010 ILAE commission proposed new terminology and concepts for underlying cause:

Idiopathic → Genetic

Symptomatic → Structural/-metabolic

All cryptogenic and many idiopathic(BRE, PS, ICOE-G) → Unknown

4. 2010 ILAE commission recommended abandoning the designation of benign epilepsy
5. 2010 ILAE commission proposed that age at onset be used as a primary dimension for organizing the epilepsy

Update Comment March 2012

Luder et al. : The commission should specify the purpose of the classification

1. Suggest to use the Four dimensions

Location

Seizures symptomatology

Etiology

Related medical conditions

2. Suggest ILAE to abandon the current approach of defining hundreds of epileptic syndromes that difficult to memorize

The Near Future

- Commission on Classification (2009-2013) is preparing the final draft, to be submitted for approval by ILAE general assembly at 2013 International Epilepsy Congress