DEFINITION AND CLASSIFICATION OF EPILEPSY

KAMORNWAN KATANYUWONG MD.

6th epilepsy camp 2015: Nakomprathom
OUTLINE

• **Definition** of epilepsy
  Definition of seizure
  Definition of epilepsy

• Epilepsy **classification**
DEFINITION OF EPILEPSY

Epilepsia 2001

Special Article

Epileptic Seizures and Epilepsy: Definitions Proposed by the International League Against Epilepsy (ILAE) and the International Bureau for Epilepsy (IBE)

Epilepsia 2005

An Operational Clinical Definition of Epilepsy

ILAE website 2013

A practical clinical definition of epilepsy

Epilepsia 2014
SEIZURE

- **Greek**: meaning *to take hold*

- **Modern**: sudden and severe event

- **Seizure = epileptic seizure**

- **Cardiology**: heart seizure
I GENERAL TERMS

1.0 SEMIOLOGY
That branch of linguistics concerned with signs and symptoms.

2.0 EPILEPTIC SEIZURE
Manifestation(s) of epileptic (excessive and/or hypersynchronous), usually self-limited activity of neurons in the brain.

3.0 ICTUS
A sudden neurologic occurrence such as a stroke or an epileptic seizure.

4.0 EPILEPSY
a) Epileptic Disorder: A chronic neurologic condition characterized by recurrent epileptic seizures.

b) Epilepsies: Those conditions involving chronic recurrent epileptic seizures that can be considered epileptic disorders.

Neuron excitation + Hypersynchrony
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<th>Table 1: Conceptual Definition of Seizure and Epilepsy – 2005 Report</th>
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<tr>
<td><strong>An epileptic seizure</strong> is a <strong>transient</strong> occurrence of <strong>signs</strong> and/or <strong>symptoms</strong> due to abnormal excessive or synchronous neuronal activity in the brain.</td>
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<td><strong>Epilepsy</strong> is a disorder of the brain characterized by an enduring predisposition to generate epileptic seizures, and by the neurobiological, cognitive, psychological, and social consequences of this condition. The definition of epilepsy requires the occurrence of at least one epileptic seizure.</td>
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An epileptic seizure is a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain.

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<td>• Associated neurobiologic, cognitive, psychological and social disturbances</td>
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2013: DEFINITION OF EPILEPSY

A **disease** of the brain defined by any of the following conditions:

1. At least 2 **unprovoked** seizures occurring more than 24 hours apart

2. **One** unprovoked seizure and a probability of further seizures similar to the general recurrence risk after two unprovoked seizures (approx 75% or more)
• After 2 unprovoked non-febrile seizure, the chance of having another is 73% (Hauser et al 1998) at 4 years

• After a single unprovoked seizure, the chance of having another is 40-52% (Berg&Shinnar 1991)
2013: WHY 24 HOURS APART?

Answer:

if seizures clustering within 24 hours
⇒ risk factor for later seizures
⇒ = risk after a single seizure
2013

• Stroke, CNS infection and trauma is important

• If the patient has a single unprovoked seizure after a remote brain insult ⇒ risk of a second unprovoked seizure = risk for further seizures after two unprovoked seizures
2013

- Some patients with a single unprovoked seizure in a circumstance of an epilepsy syndrome
  - high risk of recurrence
  - epilepsy
2013

• Definition/ Diagnosis

• Prognosis/outcome
Epilepsy is considered to be no longer present for:

- individuals who had an age-dependent epilepsy syndrome but are now past the applicable age or
- those who have remained seizure-free for at least 10 years off AEDs

- No known risk factors associated with a high probability (≥ 75%) of future seizure
2013

- Cure = disappearance
- Remission = abeyance of a disease

“no longer present” = the person no longer has epilepsy, although it does not guarantee that it will not return
2014: DEFINITION OF EPILEPSY

1. At least 2 unprovoked (or reflex) seizures occurring > 24 hours apart
2. One unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (>60%) after 2 unprovoked sz, occurring over the next 10 years
3. Diagnosis of a epilepsy syndrome

Epilepsia 2014
Resolved

- Had age-dependent epilepsy syndrome but are now past the applicable age
- Seizure free for at least 10 years and off AEDs for at least the last 5 years

Resolved is not identical to remission or cure
2014

• A decision for treatment does not equate to a diagnosis of epilepsy

• A diagnosis of epilepsy does not require treatment
NEW DEFINITION

• ? Affect prevalence of epilepsy

• Making the clinicians more comfortable in initiating treatment after some unprovoked seizures

• Required specialized diagnostic and interpretative skills—esp in assessing recurrence risks or in diagnosing syndromes
CLASSIFICATION
WHY CLASSIFICATION IS NEEDED?

• 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:
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......2013......2015

*†Anne T. Berg, ‡Samuel F. Berkovic, §Martin J. Brodie, ¶Jeffrey Buchhalter, #**J. Helen Cross, ††Walter van Emde Boas, ‡‡Jerome Engel, §§Jacqueline French, ¶¶Tracy A. Glauser, ##Gary W. Mathern, ***Solomon L. Moshé, †Douglas Nordli, †††Perrine Plouin, and ‡Ingrid E. Scheffer
# The Classification Criteria of Epilepsies

## Epilepsies (Merlis 1970)

- Clinical criteria
  - Seizures
  - Neurologic status
  - Age of onset
  - Etiology
- EEG criteria
  - Interictal EEG
  - Ictal EEG

## Epileptic seizures (Gastaut 1970)

- Clinical seizure type
- EEG seizure type
- EEG interictal expression
- Anatomical substrate
- Etiology
- Age
# ILAE 1981

## Clinical seizure type

### 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 $\rightarrow$ GTC
  - CPS $\rightarrow$ GTC
  - SPS $\rightarrow$ CPS $\rightarrow$ 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)
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

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

TWO DICHOTOMIES, A 4-PART CLASSIFICATION

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<td>Localization-related Symptomatic</td>
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Cryptogenic Special syndromes
2001

Axis 1: Ictal phenomenology
Axis 2: Seizure type: Partial Sz or Gen Sz
Axis 3: Epileptic syndromes:
Axis 4: Etiology: Idiopathic, Cryptogenic, Symptomatic
Axis 5: Impairment

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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
  - Typical
  - Absence with special features
    - Myoclonic absence
    - Eyelid Myodonia
  - Atypical

Focal seizures
- Originating within networks limited to one hemisphere
  - Myoclonic
    - Myoclonic
    - Myoclonic-ataxic
    - Myoclonic-tonic
  - 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

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<td>With motor signs</td>
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<td>With autonomic symptoms</td>
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<td>With psychic symptoms (but no impaired consciousness)</td>
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<td>Atonic (astatic)</td>
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Focal seizures
Originating within networks limited to one hemisphere

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  - Myoclonic-atonic
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focal seizure should be described according to their manifestation
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ILAE 1989

2. Generalized epilepsies and syndromes

2.1 Idiopathic (with age-related onset, listed in order to age)
  - Benign neonatal familial convulsions
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  Juvenile absence epilepsy
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  2.2 Epilepsies or symptomatic (in order to age)
  - West syndrome
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<td>Symptomatic: the epilepsy is the consequence of a known or suspected disorder of the central nervous system</td>
<td>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</td>
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<td>Cryptogenic: this refers to a disorder whose cause is hidden or occult. Cryptogenic epilepsies are presumed to be symptomatic</td>
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**2015:** Structural/metabolic...... include immune and infectious cause
## 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)

<table>
<thead>
<tr>
<th>Old terminology and concepts</th>
<th>Recommended new terminology and concepts</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Etiology</strong></td>
<td></td>
</tr>
<tr>
<td>Idiopathic: there is no underlying cause other than a possible hereditary predisposition</td>
<td>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</td>
</tr>
<tr>
<td>Symptomatic: the epilepsy is the consequence of a known or suspected disorder of the central nervous system</td>
<td>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</td>
</tr>
<tr>
<td>Cryptogenic: this refers to a disorder whose cause is hidden or occult. Cryptogenic epilepsies are presumed to be symptomatic</td>
<td>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</td>
</tr>
</tbody>
</table>

### New Term and Concept

- **Genetic:** genetic defect directly contributes to the epilepsy and seizures are the core symptom of the disorder
  - Channelopathies, Glut1 deficiency, etc.

### Old Term and Concept

- **Idiopathic:** presumed genetic
  - Symptomatic: secondary to a known or presumed disorder of the brain
  - Cryptogenic: presumed symptomatic
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)

**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
  - Hemi-convulsion-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 (ESSE)
**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.
2010

• Classification system versus diagnosis

• 2010 entail little or no change in what health care providers do in daily practice (diagnose and treat individual patients)

• CAE : IGE (old)
  : generalized epilepsy, absence, genetic cause (new)
Thank you for your time