Update in Epilepsy Classification

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Classification

What

- A system for representing knowledge about items
  - Reflects key characteristics

Why

- Communicates among the clinicians
- Established a taxonomy for the research on epilepsy

Types of classification

In general

- Biology - species: plant, animal, Immunology
- Etiology - 1st Primary or 2nd Secondary
- Pathology - Cancer
- Imaging - Cortical dysplasia
- Clinical criteria - e.g. age of onset, disease course, distribution of symptoms: HA
- Mixed - Movement disorder
Classification

What
- A system for representing knowledge about items
- Reflects key characteristics

When
- 2001, 2006
- 2010, 2013

Problem
- Knowledge ≠ Update

New knowledge

Plus neurochemistry, developmental neurobiology, computational neuroscience, electrophysiology, epidermiology

ILAE classification working group

2005-2009


### ILAE: Epileptic seizures 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 2nd 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)

### What has been changed for ILAE 2010

1. **Partial (focal/local seizure)**
   - 1981
     - Simple partial seizure
     - Complex partial seizure (consciousness)
   - 2010
     - Focal seizure
     - Originating within networks limited to one hemisphere
   - One foci, One hemisphere

2. **Partial (focal/local seizure)**
   - 1981
     - Simple partial seizure
     - Complex partial seizure (consciousness)
   - 2010
     - Focal seizure
     - Characterized according to
       - Aura
       - Motor
       - Autonomic
       - Awareness/Responsiveness
         : altered (dyscognitive)
         : retained
   - One foci, One hemisphere
Partial (focal/local seizure)
- Partial seizure evolving to secondary generalized seizure

Focal seizure evolves to bilateral convulsive seizure
: eg. tonic, clonic, T-C

Generalized seizures
(convulsive and non convulsive)
: absence, myoclonic, clonic, tonic, tonic-clonic, atonic

Generalized seizures
: absence
- typical
- atypical
- absence with special features: myoclonic absence, eyelid myoclonia
: myoclonic, myoclonic, myoclonic- atonic, myoclonic-tonic
: other = the same
What has been changed for ILAE 2010

<table>
<thead>
<tr>
<th>1989</th>
<th>2010</th>
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<tbody>
<tr>
<td>Localization-related</td>
<td>Localization-related</td>
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<tr>
<td>1.1 idiopathic</td>
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<tr>
<td>Specific syndromes</td>
<td>Specific syndromes</td>
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<tr>
<td>1.2 symptomatic</td>
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<td>Specific syndromes</td>
<td>Specific syndromes</td>
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<tr>
<td>1.3 cryptogenic</td>
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<td>Specific syndromes</td>
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<tr>
<td>2. Generalized</td>
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<tr>
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<tr>
<td>3. Undetermined whether focal or generalized</td>
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<td>e.g. FC, reflex epilepsy</td>
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1989: Mixed of two dichotomies
Onset and Etiology

### Etiology

#### Idiopathic
- Presumed genetic,
- No underlying cause other than a possible hereditary predisposition

#### Genetic
- Direct result of a known or presumed genetic defect in which seizures are the core symptom of the disorder
- e.g. Channelopathies, Glut1 def
**What has been changed for ILAE 2010**

### 6.

**1989**

Symptomatic:
Consequence of a known or suspected disorder of CNS

**2010**

Structural/metabolic:
- Proven to be associated with an increased risk of developing epilepsy, may be acquired or genetic in origin
  - eg. stroke, trauma, infection, cortical malformation or may be genetic origin (TSC)

### 7.

**1989**

Cryptogenic:
- Cause is hidden or occult,
- Presumed to be symptomatic

**2010**

Unknown:
- Not yet identified
  - Genetic vs Structural defect (metabolic)

**Terms no longer recommended**

- Benign
  - self-limited: tendency to resolve spontaneously with time
  - pharmacoresponsive: highly likely to be controlled with medication

- Catastrophic
  - pharmacoresistant
1. Localization
   1.1 idiopathic: specific syndromes
   1.2 symptomatic: specific syndromes
   1.3 cryptogenic: specific syndromes

2. Generalized
   2.1...

3. Undetermined whether focal or generalized
   3.1 specific syndromes SMEI, LKS, CSWS

4. Special syndromes
   4.1 PC, reflex epilepsy

Epileptic syndrome 2001, 2006

<table>
<thead>
<tr>
<th>Table 4</th>
<th>Epilepsy, syndromes and related conditions</th>
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<tbody>
<tr>
<td></td>
<td>Bizarre functional neurologic disorders</td>
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<td></td>
<td>Nervous system malformations</td>
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<td>Cerebral palsy</td>
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<td>Epilepsy in infancy</td>
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<td>Benign idiopathic epilepsy</td>
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<td>Benign familial infantile epilepsy</td>
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<td>Benign Roland's syndrome</td>
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<td>Benign Lahey's syndrome</td>
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<td>Lennox-Gastaut syndrome</td>
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<td>Landau-Kleffner syndrome</td>
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<td>Epilepsy with myoclonic seizures, ataxia, and encephalopathy</td>
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<td>Progressive myoclonic-astasia-encephalopathy</td>
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Epilepsia 2001, Epilepsy research 2006

- Infantile spasms (IS) with seizures evolving into Lennox syndrome
- Infantile spasms (IS) with atypical absence EEG
- Infantile spasms (IS) with atypical absences

Age at onset

Special group, surgical syndrome

Non syndromic
Debate on new organization of seizures and epilepsies 2010

- Many comments
  - Disagree; in all, in some details
  - Agree; in all, in some details
  - Suggest new options;
    1. compromised ideas with ways to approach,
    2. new classification,
    3. their own classification (their new update)
- No option

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

Thank you for your attention