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Talk overview

- Classification of seizure types and epilepsies by ILAE 2017
Milestones of ILAE classification

1981: Seizure types
1989: Epilepsy type/syndromes
2001: Seizure types (Ictal semiology)
2010: Seizure types and Epilepsy type/syndromes
2017: Seizure types and Epilepsy type/syndromes
Milestones of ILAE classification of seizure types

1981: Seizure types
Epileptic Seizures in Accordance with the ILAE Classification of 1981

- **Partial seizures**
  - simple partial seizure *(without)* impairment of consciousness
  - complex partial seizure *(with)* impairment of consciousness
  - focal seizure evolving to secondarily generalized seizures

- **Generalized seizures**
  (tonic, clonic or tonic-clonic, myoclonic and typical or atypical absences)

- **Unclassified epileptic seizures**

- **Prolonged or repetitive seizures (status epilepticus)**

Commission of Classification and Terminology of the International League Against Epilepsy; Epilepsia 1981
International Classification of Epileptic Seizures (ICES) 1981

- Electroclinical approach
- Focusing on the presence/absence of altered consciousness
  - Simple partial seizures
  - Complex partial seizures
Milestones of ILAE classification of seizure types

1981: Seizure types

2001: Seizure types (Ictal semiology)
Glossary of descriptive terminology for ictal semiology; 2001

- **Motor**
  - elementary
  - automatism

- **Non-motor**
  - aura
  - sensory
  - **dyscognitive**

- **Autonomic events**
  - autonomic aura
  - autonomic seizure

Describe seizure with unbundling from EEG, MRI or other data
Milestones of ILAE classification of seizure types

1981: Seizure types

2001: Seizure types (Ictal semiotics)

2010: Seizure types and Epilepsy type/syndromes
Revised terminology and concepts for organization of seizures and epilepsies 2010

Concept of mode of seizure onset

**Generalized seizures**: originating at some point within, and rapidly engaging, bilaterally distributed networks
- Although individual seizure onsets can appear localized, the location and lateralization are not consistent from one seizure to another.

**Focal seizures**: originating within networks limited to one hemisphere
- Ictal onset is consistent from one seizure to another

*Berg AT; Epilepsia 2010*
The distinction between **simple and complex partial seizures** is **eliminated**

Descriptors of seizure type: suggested to consult ILAE 2001

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**Table 1. Classification of seizures**

<table>
<thead>
<tr>
<th>Generalized seizures</th>
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</thead>
<tbody>
<tr>
<td>Tonic–clonic (in any combination)</td>
</tr>
<tr>
<td>Absence</td>
</tr>
<tr>
<td>Typical</td>
</tr>
<tr>
<td>Atypical</td>
</tr>
<tr>
<td>Absence with special features</td>
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<tr>
<td>Myoclonic absence</td>
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<tr>
<td>Eyelid myoclonia</td>
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</tr>
<tr>
<td>Myoclonic</td>
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<tr>
<td>Myoclonic atonic</td>
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<td>Myoclonic tonic</td>
</tr>
<tr>
<td>Clonic</td>
</tr>
<tr>
<td>Tonic</td>
</tr>
<tr>
<td>Atonic</td>
</tr>
<tr>
<td>Focal seizures</td>
</tr>
<tr>
<td>Unknown</td>
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<tr>
<td>Epileptic spasms</td>
</tr>
</tbody>
</table>

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*Seizure that cannot be clearly diagnosed into one of the preceding categories should be considered unclassified until further information allows their accurate diagnosis. This is not considered a classification category, however.

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Recognized and added

Recognized and added

( Previously called myoclonic astatic)

Recognized and added

(inadequate knowledge to make a firm decision regarding whether spasms should be classified as focal, generalized, or both)
Milestones of ILAE classification of seizure types

1981: Seizure types

2001: Seizure types (Ictal semiology)

2010: Seizure types and Epilepsy type/syndromes

2017: Seizure types and Epilepsy type/syndromes
Dyscognitive, simple partial, complex partial, psychic, and secondarily generalized seizure are eliminated.

### New seizure types
- **Focal**: automatisms, autonomic, behavior arrest, cognitive, emotional, hyperkinetic, sensory, and focal to bilateral tonic-clonic seizures. Atonic, clonic, epileptic spasms, myoclonic, and tonic seizures can be either focal or generalized.
- **Generalized**: eyelid myoclonia, myoclonic absence, myoclonic-tonic-clonic, myoclonic-tonic, myoclonic atonic, and epileptic spasm.

### Table 1. Changes in seizure type classification from 1981 to 2017

1. Change of “partial” to “focal”
2. Certain seizure types can be either of focal, generalized, or unknown onset.
3. Seizures of unknown onset may have features that can still be classified.
4. Awareness is used as a classifier of focal seizures.
5. The terms dyscognitive, simple partial, complex partial, psychic, and secondarily generalized were eliminated.
6. New focal seizure types include automatisms, autonomic, behavior arrest, cognitive, emotional, hyperkinetic, sensory, and focal to bilateral tonic-clonic seizures. Atonic, clonic, epileptic spasms, myoclonic, and tonic seizures can be either focal or generalized.
**Awareness** is initially used for classifying focal seizures.

**Motor activity**

If further develop bilateral tonic-clonic seizure.

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**ILAE 2017 Classification of Seizure Types Expanded Version**

**Focal Onset**

- **Aware**
- **Impaired Awareness**

**Generalized Onset**

- **Motor**
  - tonic-clonic
  - clonic
  - tonic
  - myoclonic
  - myoclonic-tonic-clonic
  - myoclonic-atonic
  - atonic
  - epileptic spasms

- **Nonmotor (absence)**
  - typical
  - atypical
  - myoclonic
  - eyelid myoclonia

- **Unclassified**

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Fisher RS et.al., Epilepsia 2017
Optional further descriptions

- **Focal aware or impaired awareness** seizures optionally may be further characterized by one of the listed motor onset or nonmotor onset symptoms, reflecting the first prominent sign or symptom in the seizure, for example, focal impaired awareness automatism seizure

- A focal seizure name **can omit mention of awareness** when awareness is not applicable or unknown, thereby classifying the seizure directly by motor onset or nonmotor onset characteristics
The classification of an individual seizure can stop at any level: a “focal onset” or “generalized onset” seizure, with no other elaboration, or a “focal sensory seizure,” “focal motor seizure,” “focal tonic seizure,” or “focal automatism seizure,” and so on.

The Task Force recommends classifying a seizure as having focal or generalized onset only when there is a high degree of confidence (e.g., ≥80% arbitrarily chosen to parallel the usual allowable beta error) in the accuracy of the determination.
Example

- A seizure beginning with sudden inability to understand language followed by impaired awareness and clonic left arm jerks would be classified as a

  "Focal impaired awareness (nonmotor onset) cognitive seizure" (progressing to clonic left arm jerks)

- The terms in parentheses are optional
Milestones of ILAE classification of epilepsy type, epilepsy syndromes and epilepsy etiology

1989: Epilepsy type/syndromes
Epileptic Syndromes in Accordance with Currently Valid ILAE Classification of 1989

- Localization-related (focal, local and partial) epilepsies and syndromes
  idiopathic, symptomatic, cryptogenic

- Generalized epilepsies and syndromes
  idiopathic, cryptogenic or symptomatic,
  symptomatic (EIEE, EMA)

- Epilepsies and syndromes undetermined as to whether they are focal or generalized

- Special syndromes
  FS, isolated seizure, acute symptomatic seizure
**Etiology**

- **Idiopathic (primary) epileptic syndrome**
  - no underlying structural brain lesion or other neurological signs or symptoms
  - presumed to be genetic and are usually age-dependent

- **Symptomatic**
  - result of one or more identifiable structural lesions of the brain

- **Cryptogenic (Probably symptomatic)**
  - believed to be symptomatic, but no etiology has been identified e.g. Lennox-Gastaut syndrome, West syndrome
Milestones of ILAE classification

1989: Epilepsy type/syndromes

2010: Seizure types and Epilepsy type/syndromes
Revised terminology and concepts for organization of seizures and epilepsies 2010

- The dichotomy of focal versus generalized is abandoned

- Electroclinical syndromes arranged by age at onset
  - Neonatal period
  - Infancy
  - Childhood
  - Adolescence-adult
  - Less specific age relationship

- Distinctive constellations

- Epilepsies attributed to and organized by structural-metabolic causes

- Epilepsies of unknown cause

- Conditions with epileptic seizures that are traditionally not diagnosed as a form of epilepsy per se
## Epilepsy etiology

<table>
<thead>
<tr>
<th>1989</th>
<th>2010</th>
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<tr>
<td>Idiopathic</td>
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</tr>
<tr>
<td>Symptomatic</td>
<td>Structural-metabolic</td>
</tr>
<tr>
<td>Cryptogenic</td>
<td>Unknown</td>
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</table>
Milestones of ILAE classification

1989: Epilepsy type/syndromes

2010: Seizure types and Epilepsy type/syndromes

2017: Seizure types and Epilepsy type/syndromes
The diagnosis is made on clinical grounds (seizure types), supported by EEG findings.

**Combined generalized & Focal**: both types of seizures occur e.g. Dravet syndrome and Lennox-Gastaut syndrome.

**Unknown**: the clinician is unable to determine if the Epilepsy Type is focal or generalized because there is insufficient information available.

The Epilepsy type may also be the final level of diagnosis achievable where the clinician is unable to make an Epilepsy Syndrome diagnosis.
## Epilepsy etiology

<table>
<thead>
<tr>
<th>Year</th>
<th>Classification</th>
<th>1989</th>
<th>2010</th>
<th>2017</th>
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</table>
Genetic etiology

- **Diagnosis of genetic etiology** based on
  - solely on a family history of an autosomal dominant disorder
  - may be suggested by clinical research in populations (familial aggregation studies) with the same syndrome such as Childhood Absence Epilepsy or Juvenile Myoclonic Epilepsy
  - a molecular basis may have been identified and may implicate a single gene or copy number variant of major effect e.g. Dravet’s syndrome in which >80% of patients have a pathogenic variant of SCN1A
Epilepsy syndromes

- Epilepsy syndrome refers to a cluster of features incorporating seizure types, EEG, and imaging features that tend to occur together.

- Retain the term “**Idiopathic generalized epilepsy (IGE)**”
  - Childhood Absence Epilepsy (CAE)
  - Juvenile Absence Epilepsy (JAE)
  - Juvenile Myoclonic Epilepsy (JME)
  - Generalized Tonic–Clonic Seizures Alone
Developmental and Epileptic encephalopathies

- The concept of the epileptic encephalopathy may be applicable to epilepsies at all ages and should be utilized more widely than just for the severe epilepsies with onset in infancy and childhood.

- A key component of the concept is that amelioration of the epileptiform activity may have the potential to improve the developmental consequences of the disorder.
Developmental and Epileptic encephalopathies (cont.)

“symptomatic generalized epilepsies” will no longer be used as it was applied to a highly heterogeneous group of patients

- **Developmental encephalopathy**: where there is just developmental impairment without frequent epileptic activity associated with regression or further slowing of development

- **Epileptic encephalopathy**: where there is no preexisting developmental delay and the genetic mutation is not thought to cause slowing in its own right

- **Developmental and epileptic encephalopathy**: where both factors play a role e.g. SCN1A causes Dravet’s syndrome
Self-limited and pharmacoresponsive

- “**Benign**” as a descriptor for epilepsy, is replaced by both “self-limited” and “pharmacoresponsive”
- “**Self-limited**” refers to the likely spontaneous resolution of a syndrome
- “**Pharmacoresponsive**” means that the epilepsy syndrome will be likely to be controlled with appropriate antiepileptic therapy
Thank you for your attention