DEFINITION AND CLASSIFICATION OF EPILEPSY

DEFINITION OF EPILEPSY

SEIZURE

• Greek : meaning *to take hold*
• Modern : sudden and severe event
• Seizure = epileptic seizure
• Cardiology : heart seizure

OUTLINE

• Definition of epilepsy
• Definition of seizure
• Epilepsy classification
• Test your recent memory....


Warren T. Illes—Chet, Reo O. Liem, Eli Mendel, Carla Turrion, Victor von Engel, and Jerome Engel, Jr., Editors

Special Article

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

Epilepsia 2001

An Operational Clinical Definition of Epilepsy


Epilepsia 2005

ILAE website 2013
2005: EPILEPSY

A disorder of the brain characterized by enduring predisposition to generate epileptic seizure

• Usually practically applied as having 2 unprovoked seizures more than 24 hours apart

2013: 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)

2013: EPILEPSY

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

2005

<table>
<thead>
<tr>
<th>Table 1: Conceptual Definition of Seizure and Epilepsy – 2005 Report</th>
</tr>
</thead>
<tbody>
<tr>
<td>An epileptic seizure is a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain.</td>
</tr>
<tr>
<td>Epilepsy 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>
</tr>
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</table>

2005: ELEMENTS OF A DEFINITION OF EPILEPSY

• History of at least one seizure

• Enduring alteration in the brain that increase the likelihood of future seizures

• Associated neurobiologic, cognitive, psychological and social disturbances
2013

- Epilepsy presents after 2 unprovoked seizures occurring at least 24 hours apart (Hauser et al 1991)

- 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

Q: Why 24 hours apart?
A: 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 patient with a single unprovoked seizure in a circumstance of an epilepsy syndrome ⇒ high risk of recurrence ⇒ epilepsy

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

TYPES OF CLASSIFICATION

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

- Mixed:

OVER THE PAST 150 YEARS

1860 1910 1960 2010

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

COMMISSION ON CLASSIFICATION AND TERMINOLOGY OF ILAE

ILAE CLASSIFICATION WORKING GROUP

2005-2009

SPECIAL REPORT

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

*Anne T. Berg, ‡Walter van Emde Boas, §Jonne Engel, †Jacqueline French, ¶Tracy A. Glauser, **Gary W. Mathern, ***Simone L. Moshe, ‡‡Douglas Nordli, §§Perrine Proux, and ‡§§Ingrid B. Scheffer
THE CLASSIFICATION CRITERIA OF EPILEPSIES

**Clinical criteria**
- Seizures
- Neurologic status
- Age of onset
- Etiology

**EEG criteria**
- Interictal EEG
- Ictal EEG

**Epileptic seizures**
- Simple partial seizure
  - with motor signs
  - with somatosensory symptoms
  - with autonomic symptoms and signs
  - with psychic symptoms
- Complex partial seizure
  - start with SPS followed by impairment of consciousness
  - with impairment of consciousness at onset
- Partial seizure evolving to generalized seizure

1. **Partial (focal, local) seizures**
   - Simple partial seizure
     - with motor signs
     - with somatosensory symptoms
     - with autonomic symptoms and signs
     - with psychic symptoms
   - Complex partial seizure
     - start with SPS followed by impairment of consciousness
     - with impairment of consciousness at onset
   - Partial seizure evolving to generalized seizure
     - SPS → GTC
     - CPS → GTC
     - SPS → CPS → GTC

2. **Generalized seizures (convulsive and non-convulsive)**
   - Absence, Myoclonic, Tonic, Tonic-clonic, Atonic

3. **Unclassified epileptic seizures**

4. **Prolonged or repetitive seizure (status epilepticus)**

ILAE 1981

<table>
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<tr>
<th>Clinical seizure type</th>
<th>EEG sz type</th>
<th>EEG interictal expression</th>
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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

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 with grand mal (GTCS) sz on awakening
      - etc.
   2.2 Cryptogenic or symptomatic (in order to age)
      - Lennox-Gastaut syndrome
      - Epilepsy with myoclonic-astatic sz
      - Epilepsy with myoclonic absences

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

ILAE 1989

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TWO DICHOTOMIES, A 4-PART CLASSIFICATION

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<th>Generalized</th>
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<td>SPE</td>
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ILAE 1989

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The use of generalized and partial (focal) to refer to the underlying was abandoned.

But the terms were retained in reference to mode of seizure initiation and presentation.

focal seizure should be described according to their manifestation.

2001

Axis 1: ictal phenomenology
Axis 2: Seizure type
Axis 3: Epileptic syndromes
Axis 4: Etiology
Axis 5: Impairment
Structural/metabolic: proven to be associated with an increased risk of developing epilepsy, include stroke, trauma, infection or may be genetic origin (TSC).

### Table 1. Comparison of major changes between old and proposed Terminology and the newly proposed Classification (Convocration 1981,1985: Berg et al., 2010)

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<td>CAE: IGE (old)</td>
<td>Generalized epilepsy, absence, genetic cause (new)</td>
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**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)**
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<th>Gene Function</th>
<th>Associated Neurological Syndromes</th>
<th>Brief Description of Syndrome</th>
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<td>SCN 14A-1 subunit of the neuronal voltage-gated sodium channel</td>
<td>Onset: infancy or early childhood, progressive cognitive impairment, EEG shows normal to background slowing with absence of spike-wave discharges</td>
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<td>SCN 14A-1 subunit of the neuronal voltage-gated sodium channel</td>
<td>Early onset absence seizures</td>
<td>Onset: infancy, absence seizures, EEG shows generalized spike-wave discharges.</td>
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<tr>
<td>SCN 14A-1 subunit of the neuronal voltage-gated sodium channel</td>
<td>Doose syndrome</td>
<td>Onset: early childhood, multiple seizure types, generalized tonic-clonic seizures, EEG shows generalized spike-wave discharges.</td>
</tr>
<tr>
<td>SCN 14A-1 subunit of the neuronal voltage-gated sodium channel</td>
<td>Non-syndromic focal epilepsy</td>
<td>Onset: early childhood, multiple seizure types, EEG shows generalized spike-wave discharges.</td>
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<td>SCN 14A-1 subunit of the neuronal voltage-gated sodium channel</td>
<td>KCNQ2 neuronal sodium channel</td>
<td>Onset: early childhood, multiple seizure types, EEG shows generalized spike-wave discharges.</td>
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