Classification and concepts of generalized seizures and epilepsies: what is new?

Sameer M Zuberi

Chair, ILAE Commission on Classification & Terminology 2013-17

Royal Hospital for Children &
University of Glasgow
United Kingdom

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The 2017 Classification of Seizures and the Epilepsies

Will be published in Epilepsia very shortly

First official / ILAE Position Papers on Classification since 1989

New concepts are presented & explained in 3 companion papers

Work of many Commissions, Task Forces & Individuals
ILAE Classification of the Epilepsies

Position Paper of the ILAE Commission for Classification and Terminology

Ingrid E Scheffer¹, Samuel Berkovic², Giuseppe Capovilla³, Mary B Connolly³,
Jacqueline French⁴, Laura Guilhoto⁵, Edouard Hirsch⁶, Satish Jain⁷, Gary W. Mathern⁸,
Solomon L Moshe⁹, Douglas Nordli¹⁰, Emilio Perucca¹¹, Torbjorn Tomson¹², Samuel
Wiebe¹³, Yue-Hua Zhang¹⁴, Sameer M Zuberi¹⁵

ILAE Position Papers - in press in Epilepsia
Operational Classification of Seizure Types by the International League Against Epilepsy

Robert S. Fisher¹, J. Helen Cross², Jacqueline A. French³, Norimichi Higurashi⁴, Edouard Hirsch⁵, Floor E. Jansen⁶, Lieven Lagae⁷, Solomon L. Moshé⁸, Jukka Peltola⁹, Eliane Roulet Perez¹⁰, Ingrid E. Scheffer¹¹, Sameer M. Zuberi¹²

Instruction manual for the ILAE 2017 Operational Classification of Seizure Types

Robert S. Fisher¹, J. Helen Cross², Carol D’Souza³, Jacqueline A. French⁴, Sheryl Haut⁵, Norimichi Higurashi⁶, Edouard Hirsch⁷, Floor E. Jansen⁸, Lieven Lagae⁹, Solomon L. Moshé¹⁰, Jukka Peltola¹¹, Eliane Roulet Perez¹², Ingrid E. Scheffer¹³, Andreas Schulze-Bonhage¹⁴, Ernest Somerville¹⁵, Carol D’Souza¹⁶ Michael Sperling¹⁷, Elza Marcia Yacubian¹⁸, Sameer M. Zuberi¹⁹
The 2017 Classification of Seizures and the Epilepsies

A seizure is the transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain.

A seizure type is a useful grouping of seizure characteristics for purposes of clinical care, teaching & research.

The principal aim of the 2017 classification is to provide a communication framework for clinical use.
Generalized seizures - 2017

• For seizures
  – Generalized epileptic seizures are conceptualized as originating at some point within, and rapidly engaging, bilaterally distributed networks. …can include cortical and subcortical structures, but not necessarily include the entire cortex.

Previous (1989) definition of Generalised
  – seizures in which the first clinical changes indicate initial involvement of both hemispheres
Focal reconceptualized

• For seizures:
  – *Focal epileptic seizures are* conceptualized *as originating within networks limited to one hemisphere. These may be discretely localized or more widely distributed.*

ILAE 2017 Classification of Seizure Types Basic Version

- **Focal Onset**
  - Aware
  - Impaired Awareness
- **Generalized Onset**
  - Motor
    - Tonic-clonic
    - Other motor
  - Non-Motor (Absence)
- **Unknown Onset**
  - Motor
    - Tonic-clonic
    - Other motor
  - Non-Motor
  - Unclassified

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1. Definitions, other seizure types and descriptors are listed in the accompanying paper & glossary of terms.
2. Due to inadequate information or inability to place in other categories.
ILAE 2017 Classification of Seizure Types Expanded Version

**Focal Onset**
- Aware
- Impaired Awareness

**Motor Onset**
- automatisms
- atonic
- clonic
- myoclonic
- tonic
- epileptic spasms
- hyperkinetic
- myoclonic-tonic-clonic
- myoclonic-atonic
- tonic
- epileptic spasms

**Non-Motor Onset**
- autonomic
- behavior arrest
- cognitive
- emotional
- sensory
- focal to bilateral tonic-clonic

**Generalized Onset**
- Motor
  - tonic-clonic
  - clonic
  - tonic
  - myoclonic
  - myoclonic-tonic-clonic
  - myoclonic-atonic
  - atonic
  - epileptic spasms
- Non-Motor (absence)
  - typical
  - atypical
  - myoclonic
  - eyelid myoclonia

**Unknown Onset**
- Motor
  - tonic-clonic
  - epileptic spasms
  - Non-Motor
  - behavior arrest

**Unclassified**

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1. Definitions, other seizure types and descriptors are listed in the accompanying paper and glossary of terms.
2. Degree of awareness usually is not specified.
3. Due to inadequate information or inability to place in other categories.
Epilepsy Types 2017

Diagnosis made on clinical grounds supported by EEG features

To have a generalised epilepsy type the patient should have a generalised seizure type(s) and generalised spike wave on the EEG.

If the EEG is normal you must be cautious about making a diagnosis of generalised epilepsy

A new category of combined generalised and focal epilepsy
Co-morbidities

Seizure types
- Focal
- Generalized
- Unknown

Epilepsy types
- Focal
- Generalized
- Combined Generalized & Focal
- Unknown

Epilepsy Syndromes

Etiology
- Structural
- Genetic
- Infectious
- Metabolic
- Immune
- Unknown
Genetic and Idiopathic Generalised Epilepsies

Idiopathic in 1989 Classification – an epilepsy of predominantly genetic origin in which there is no gross neuroanatomical or neuropathological abnormality.

Genetic in the 2017 Classification - the epilepsy results from a known or presumed genetic cause (gene identified, family history or family studies). Genetic is not always inherited.

Should we keep or discard the term idiopathic in 2017?
Co-morbidities

Seizure types
- Focal
- Generalized
- Unknown

Epilepsy types
- Focal
- Generalized
- Combined Generalized & Focal
- Unknown

Epilepsy Syndromes

Etiology
- Structural
- Genetic
- Infectious
- Metabolic
- Immune
- Unknown
- Unknown
Genetic generalised epilepsies

Always best to classify to syndrome level if you can.

If you don’t want to use genetic for an individual with generalised epilepsy or you feel there isn’t evidence to use the term genetic use “generalised epilepsy of unknown cause”

Childhood absence epilepsy, juvenile absence epilepsy, juvenile myoclonic epilepsy & generalised tonic clonic seizures alone are genetic generalised epilepsies

For these syndromes alone you can also use the term “Idiopathic Generalised Epilepsies”. Otherwise the term idiopathic is not used in epilepsy classification.
Co-morbidities

Seizure types
- Focal
- Generalized
- Unknown

Epilepsy types
- Focal
- Generalized
- Combined Generalized & Focal
- Unknown

Epilepsy Syndromes

Etiology
- Structural
- Genetic
- Infectious
- Metabolic
- Immune
- Unknown
- Unknown
Look out for 3 companion papers in

Epilepsia
Official Journal of the International League Against Epilepsy

www.epilepsydiagnosis.org

sameer.zuberi@nhs.net
Previous classifications were all *proposals* – but became established

- **Generalised** – seizures in which the first clinical changes indicate initial involvement of both hemispheres

- **Focal** – seizure semiology or findings at investigation disclose a localised origin of the seizures.

- **Idiopathic** – an epilepsy of predominantly genetic origin in which there is no gross neuroanatomical or neuropathological abnormality

- **Symptomatic** – an epilepsy of an acquired or genetic cause associated with neuroanatomical or neuropathological abnormalities indicative of underlying disease or condition

- **Cryptogenic** – an epilepsy of presumed symptomatic nature in which the cause has not been identified