What's in a name?

The International League Against Epilepsy 2017 classification of seizures and epilepsy

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Disclosures

Board of directors for the American Clinical Magnetoencephalography Society (ACMEGS)

Speakers bureau (honorarium)

- UCB - lacosamide (Vimpat) and brivaracetam (Briviact)
- Lundbeck - clobazam (Onfi)
- Eisai - peramapanel (Fycompa)

Objectives

Appreciate the history of seizures and epilepsy

Understand the pathophysiology of seizures and epilepsy

Know the definitions of seizures, epilepsy, and status epilepticus; specifically, the ILAE 2015 and 2017 classifications

The definition of name

Noun	A word or set of words by which a person, animal, place, or thing is known, addressed, or referred to A famous person
Verb	Give a name to Specify
Adjective	Having a name that is widely known

Epilepsy etymology

Greek origin

Same root as the verb, epilambanein

- "To seize" or "to attack"

Therefore, epilepsy means "seizure," "epileptic," or "seized"

However, over time….

The Falling Sickness; 1994.

Epilepsy, AKA…

Sacred disease or the Great Disease; Hippocrates, 400 BC

Morbus maior; generalized tonic-clonic seizure in Latin

Grand mal; Medieval French

Eclampsia infantum and eclampsia gravidarum were also called epilepsy

Caducus (synonym for epilepsy); Apuleius, Middle Ages

- Caducarii - "…it is common among us to call those whom epilepsy has smitten."

The Falling Sickness; 1994.

Epilepsy, AKA…

Passio caduca

Falling evil

Falling sickness

Fallendes Seichtum, Fallsucht (German)

Fallings Wai Padavica (southern Slavs)

“Lunacy” - the disease of the moon

The Falling Sickness; 1994.

Epilepsia

Absurd linguistic inventions and creations of the Middles Ages were gone

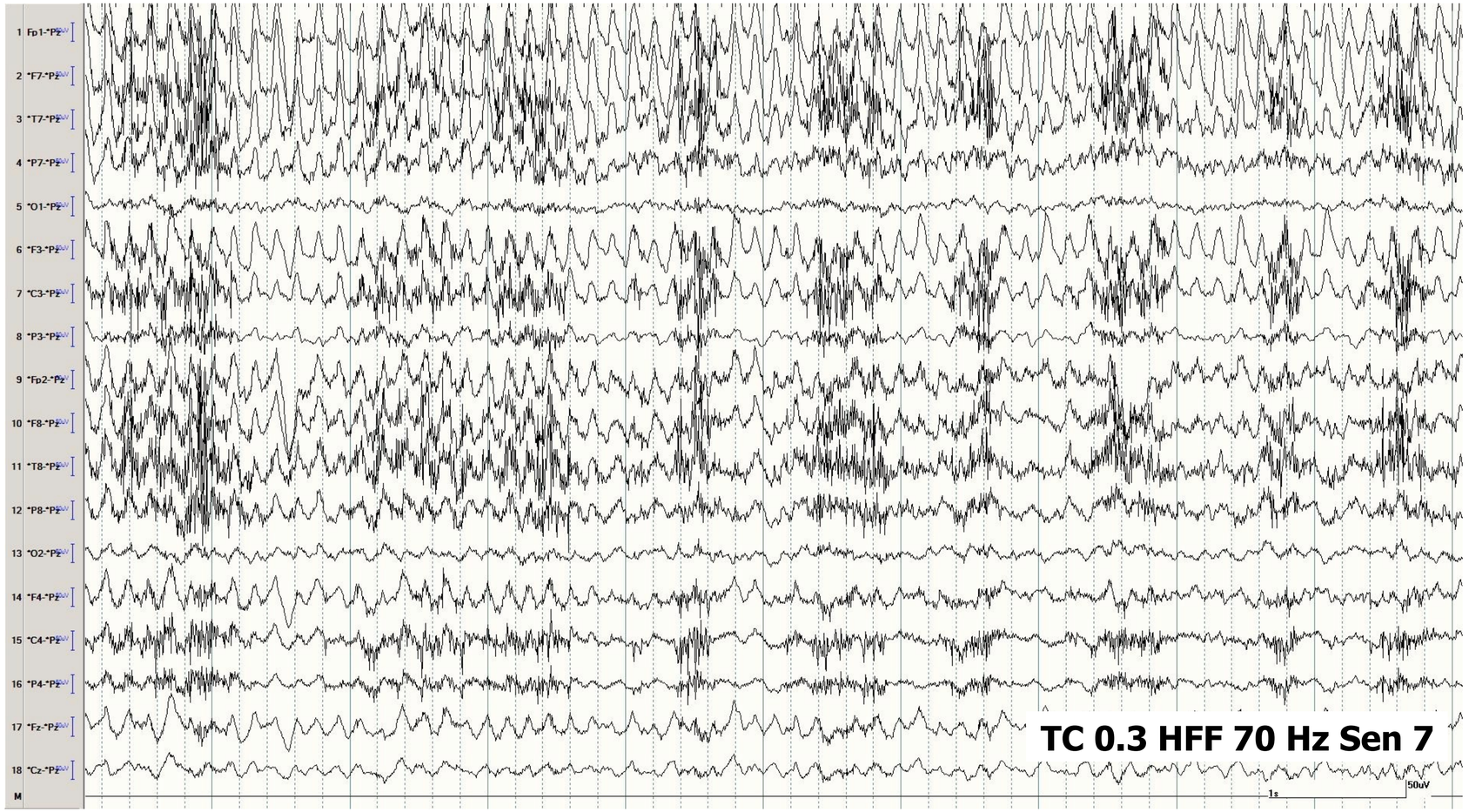
Antonius Guainerius

- **Epilepsia**
  - Epi - above
    Lesis - lesion
  - A lesion of the upper part, namely the head

The Falling Sickness; 1994.

What is a seizure?

A seizure is a transient occurrence of signs and/or symptoms (or lack thereof) due to abnormal excessive or synchronous neuronal activity in the brain



Wyllie's Treatment of Epilepsy: Principles and Practice, Fifth edition; 2011.

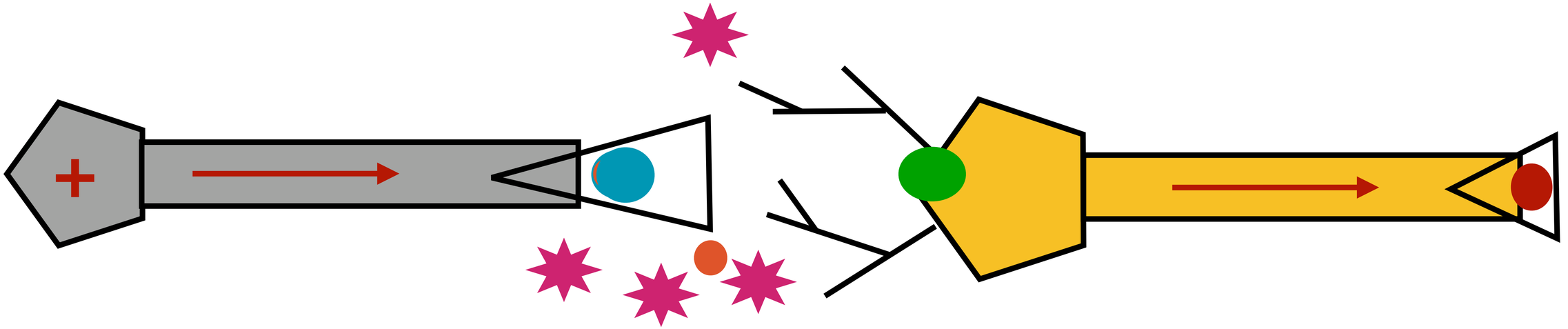
Mechanisms of seizures and pathophysiology of epilepsy

“What can be said at present about the fundamental mechanisms of different forms of epilepsy?” pg. 217

Complex interactions between genetics, neurodevelopment, environmental factors, cellular excitation/inhibition, inflammation, etc…

Epilepsy A Comprehensive Textbook, Second edition; 2008.

Chemical neurotransmission



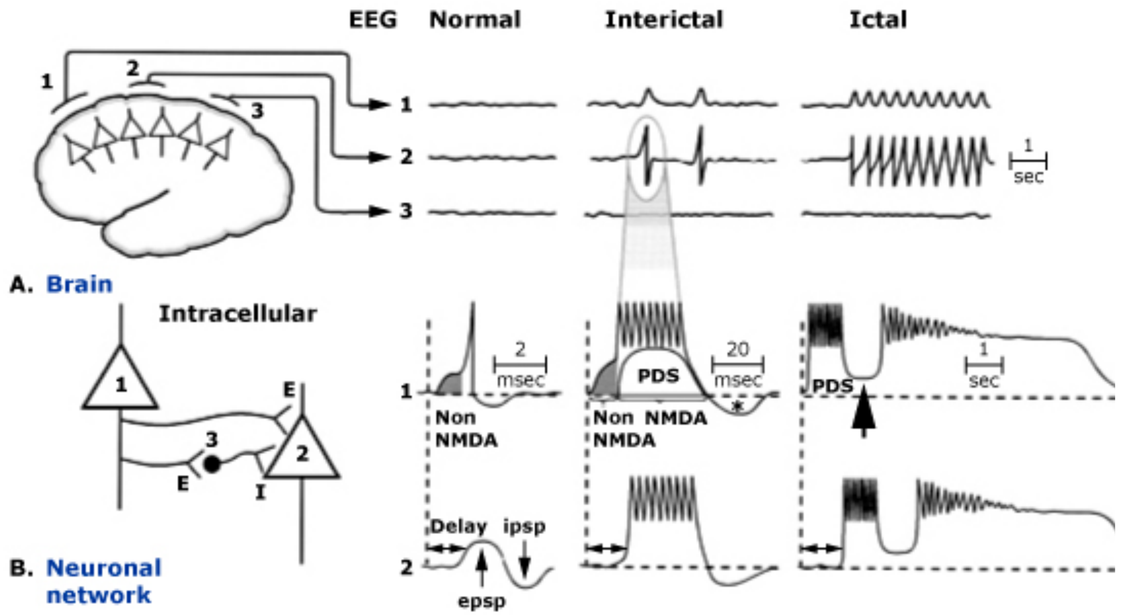
Steps

1. Secretory vesicle synthesis and transport to the synaptic terminal
2. Small-molecule NTs are loaded into the vesicle; neuropeptide synthesis occurs during vesicle synthesis
3. Depolarization of the presynaptic terminal
4. Vesicle docking with the presynaptic membrane, exocytosis, and trans-synaptic diffusion of the NT
5. Binding of the NT and activation of the post-synaptic neuron
6. Signal transduction and post-synaptic response
7. Active reuptake of the NT by the pre-synaptic neuron
8. Enzymatic degradation of the transmitter in the synaptic cleft

Fundamental Neuroscience, Second edition; 2008.

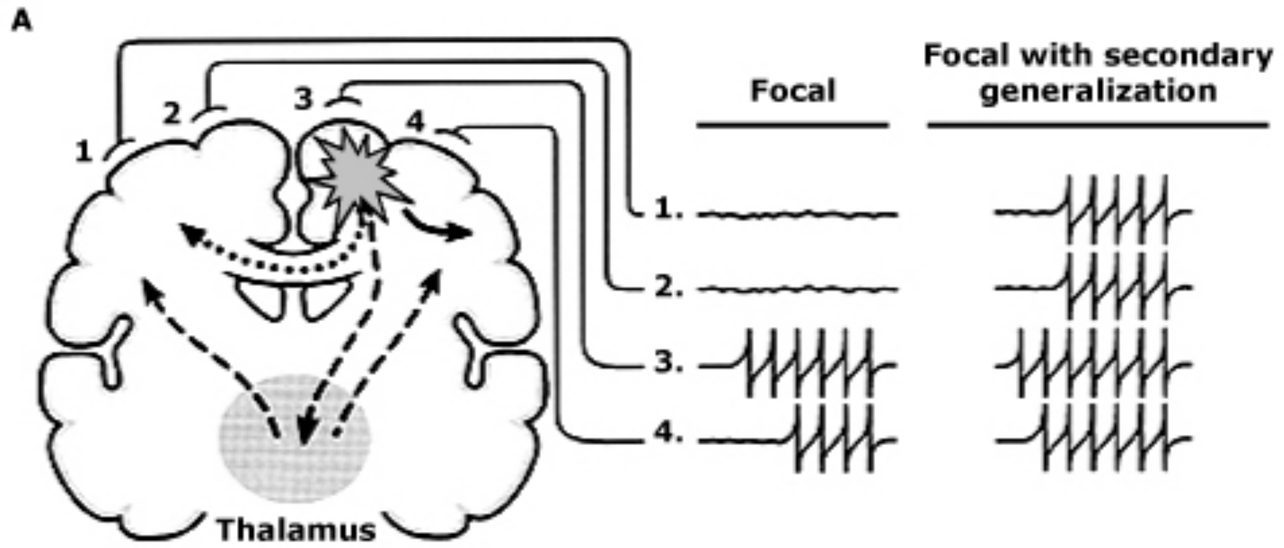
Pathophysiology of focal seizures

Abnormal neuronal firing in epilepsy



Neuronal synchronization is likely mediated by defective inhibition/ excitation at the margin of the lesion and adjacent white matter

Seizure types and potential routes of spread



PDS consists of an initial rapid and prolonged depolarization followed by a burst of repetitive action potentials; mediated by AMPA receptors, but sustained depolarization by NMDA activation

Up To Date.

Epilepsy defined

Epilepsy is a disease of the brain defined by any of the following conditions:

- 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 (at least 60%) after two unprovoked seizures, occurring over the next 10 years
- 3. Diagnosis of an epilepsy syndrome

Epilepsy is considered to be resolved for individuals who had an age-dependent epilepsy syndrome, but are now past the applicable age or those who have remained seizure-free for the last 10 years with no seizure medication for the last 5 years

Epilepsia. 2014;55(4):475-482.

~ 300 BC	Theophrastus, "When he sees a madman or an epileptic, he shudders and spits in his bosom."
~ 23-79 AD	Pliny the Elder, "In cases of epilepsy we spit, that is, we throw back contagion."
Medieval	Epilepsy was the result of demonic possession; people with epilepsy were treated as witches and warlocks
Renaissance	Andreas Caesalpinus - Epilepsy differentiated from demonism; Daemonum investigatio peripatetica (Peripatetic investigation of demons)
19th century 20th century	Theodore Herpin (1799-1865) - 300 cases of people with epilepsy; likely first description of JME William Richard Gowers (1845-1915) - 3000 cases covering every possible clinical feature of epilepsy John Hughlings Jackson (1835-1911) - localized clinical semiology; "Jacksonian March"
2006 study	Children and adolescents view physical impact of epilepsy similar to Down syndrome and more severe than asthma, diabetes, arthritis, migraine, leukemia, and HIV; 1% thought epilepsy was contagious
Present day	Trump, Fox News, Netflix…

Why classify?

Provides a framework for diagnosis

Diagnosis in turn directs management and allows to outline prognosis

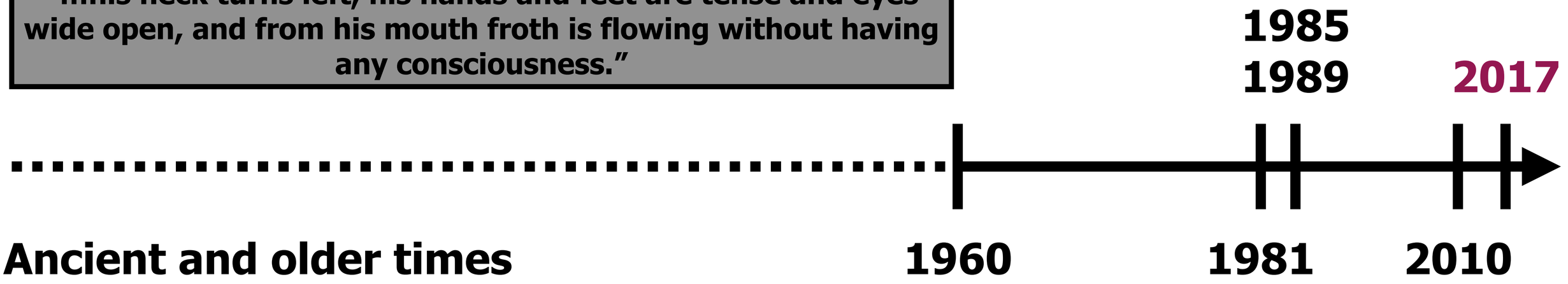
"If names be not correct, language is not in accordance with the truth of things. If language be not in accordance with the truth of things, affairs cannot be carried on to success," Confucius, Analects, 6th century BC.

Epileptic Disord. 2018;20(2):77-87.

Seizure and epilepsy classification

2,000 BC*
Akkadian text (language used in Mesopotamia)

"…his neck turns left, his hands and feet are tense and eyes wide open, and from his mouth froth is flowing without having any consciousness."



2017**
English text
(language used by many people)

"…recognize and classify seizures based on clinical characteristics as opposed to the pathogenesis."
Focal to bilateral tonic-clonic seizure

* Epilepsy Behav. 2010;17(1):103-108.
**Epilepsia. 2017;58(4):531-542.

Aura

Greek origin

Originally meant a “breeze”

A patient told Galen:

- Symptoms began in his lower leg and “from here it climbed upwards in a straight line through the thigh an further through the flank and see to the neck and as far as the head; but as soon as it had touched the latter he was no longer able to follow.”

- The patient could not exactly rose up to the head, but another observer (child) said, **“that it was like a cold breeze.”**

The Falling Sickness; 1994.

First (modern) classifications

Henri Gastaut proposed a classification of seizures in 1964

International League Against Epilepsy (ILAE) adopted this proposal in 1969

Primary focus was on distinguishing partial onset seizures from generalized onset seizures

Gastaut's multidimensional classification focused on:

- Clinical and EEG manifestations, interictal EEG, age of onset, neuropsychiatric phenomena, treatment response, cause, and known or hypothesized pathophysiology

Curr Opin Neurol. 2013;26:163-167.

International classification of epileptic seizures (ICES) - 1981

Has been the cornerstone of seizure and epilepsy classification

Primary goal - dichotomize seizures and epilepsy classification

Partial seizures

"The first clinical and EEG changes indicate initial activation of a system of neurons limited to part of one cerebral hemisphere."

Simple

1. Focal motor with/without Jacksonian March
2. Somatosensory or special symptoms
3. Autonomic features
4. Psychic features

Complex

Loss of awareness/impairment of consciousness

Generalized seizures

"First clinical changes indicate initial involvement of both hemispheres."

Absence
Myoclonic
Tonic-clonic
Tonic
Atonic
Clonic

Chang et al. J Formos Med Assoc. 2017;116:736-741.

International classification of epilepsies and epileptic seizures (ICE) - 1985-1989

Dichotomization of seizures; now **FOCAL and GENERALIZED**
Further divisions into: Idiopathic or Symptomatic (known/identifiable lesion)
Divisions into anatomical localizations: Temporal lobe v Frontal lobe v Parietal lobe v Occipital lobe

Epilepsy Type	Classification
Localization-related (partial/focal/local) epilepsies and syndromes	**Idiopathic Symptomatic Cryptogenic**
Generalized epilepsies and syndromes	**Idiopathic Cryptogenic or symptomatic Symptomatic**
Epilepsies and syndromes undetermined to be generalized or focal	**With both generalized and focal seizures Without equivocal generalized or focal features**
Special syndromes	

Chang et al. J Formos Med Assoc. 2017 [Epub Ahead of Print].

New(er) terminology

Epilepsia, 51(4):676–685, 2010
doi: 10.1111/j.1528-1167.2010.02522.x

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, ‡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

*Department of Biology, Northern Illinois University, DeKalb, Illinois, U.S.A.; †Department of Neurology, Epilepsy Center, Northwestern Children's Memorial Hospital, Chicago, Illinois, U.S.A.; ‡Epilepsy Research Centre, University of Melbourne (Austin Health), West Heidelberg, Victoria, Australia; §Epilepsy Unit, Western Infirmary, Glasgow, Scotland; ¶Department of Neurology, Phoenix Children's Hospital, Phoenix, Arizona, U.S.A.; #Neurosciences Unit, UCL-Institute of Child Health, Great Ormond Street Hospital, London, United Kingdom; **National Centre for Young People with Epilepsy, Lingfield, United Kingdom; ††Department of EEG/EMU, Epilepsy Clinic "Meer & Bosch," Heemstede; Stichting Epilepsie Instellingen Nederland, Hoofddorp, The Netherlands; ‡‡Department of Neurology, UCLA, Los Angeles, California, U.S.A.; §§Department of Neurology, New York University, New York, New York, U.S.A.; ¶¶Department of Neurology, Cincinnati Children's Hospital, Cincinnati, Ohio, U.S.A.; ##Department of Neurosurgery, UCLA, Los Angeles, California, U.S.A.; ***Department of Neurology, Albert Einstein College of Medicine, Bronx, New York, U.S.A.; and†††Department of Neurology, Hôpital Necker Enfant Malades, Paris, France

Epilepsia. 2010;51(4):676-685.

Seizure

Focal seizures

Begin in a localized network of neurons limited to one hemisphere of the brain

Focal seizures without impairment of awareness (**simple partial seizures**, auras)
Focal dyscognitive seizures (**complex partial seizures**)

Evolution to bilateral convulsive seizures (**secondary generalized tonic-clonic seizures**)

Generalized seizures

Begin in a network of neurons involving both hemispheres

Tonic-clonic seizures
Absence seizures
Myoclonic seizures
Tonic seizures
Atonic seizures
Clonic seizures

Electroclinical syndromes

Characterized by age of onset, seizure types, EEG features, and response to seizure medications

Epilepsia. 2010;51(4):676-685.

Revised terminology of seizures and epilepsies 2005-2009; published in 2010

Introduced the concept of "network"

- Focal seizures originated in a localized network within 1 hemisphere of the brain, while generalized seizures involved and rapidly engaged networks within both cerebral hemispheres

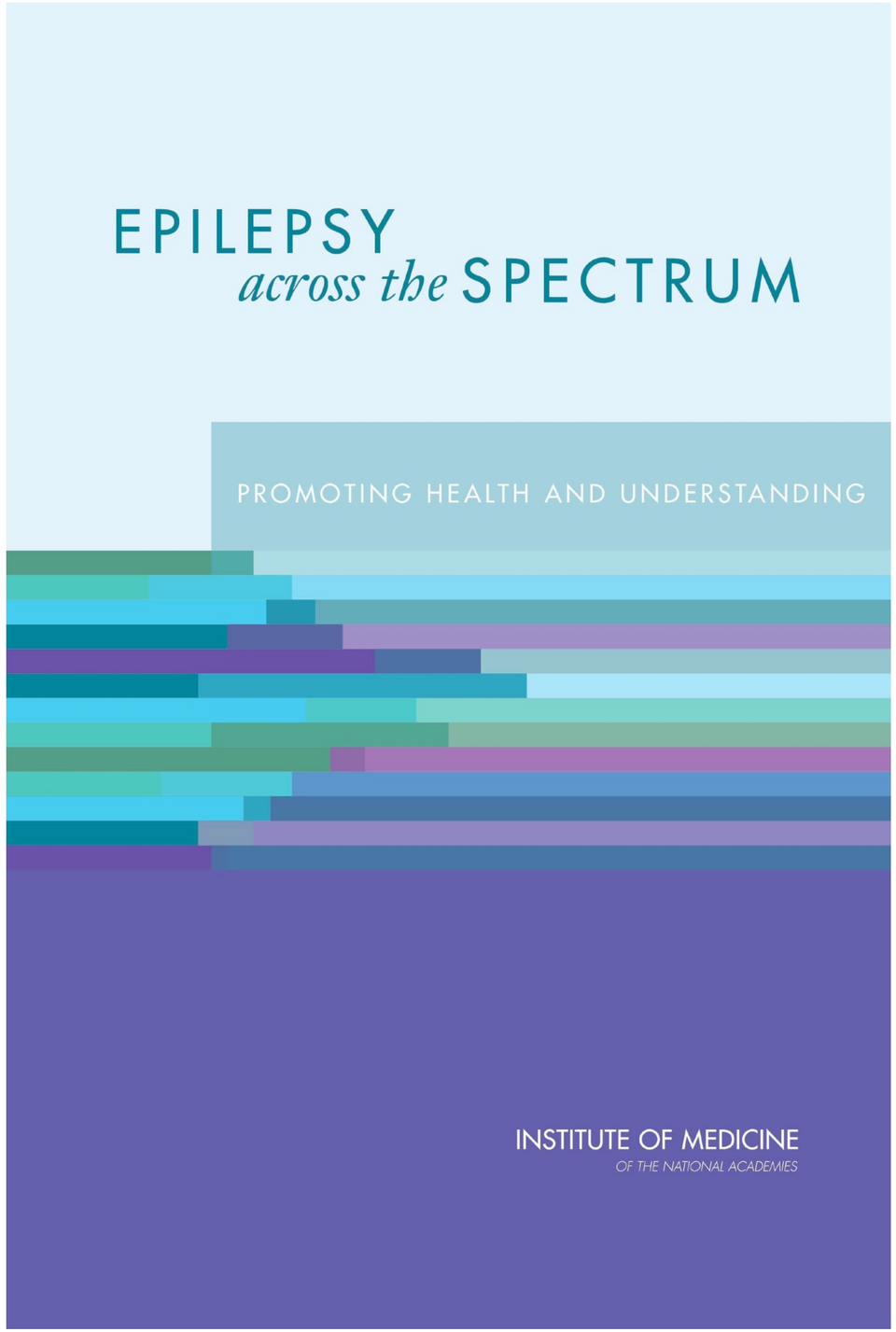
Partial seizures now referred to as focal seizures

- Simple partial seizure —> Focal seizure without impairment of awareness
  Complex partial seizure —> Focal seizure with impairment of awareness
  Secondary generalized tonic-clonic seizure —> Focal evolving to bilateral convulsive seizure

Idiopathic and cryptogenic/symptomatic replaced with genetic and structural/metabolic

Concept of electroclinical syndrome introduced

Epilepsia. 2010;51(4):676-685.



The IOM committee prepared a report to recommend priorities in public health, health care and human services, and health literacy and public awareness for the epilepsies as well as provide strategies to achieve these recommendations

Report was released on March 30, 2012

Epilepsy across the spectrum, IOM; 2012.

New epilepsy terminology

~~Epileptic seizures~~

Epilepsy seizures

~~AEDs~~

Seizure medications

~~Seizure disorder~~

Epilepsy or epilepsies

~~Psychogenic nonepileptic seizures or pseudoseizures~~

Seizure-like events with a psychological basis

Epilepsy across the spectrum, IOM; 2012.

Even newer terminology????

CONTROVERSY IN EPILEPSY

Table 1. Neurological characteristics of the different types of alterations/loss of consciousness observed during or after epileptic seizures

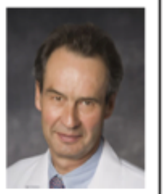
Alteration of consciousness	Alert system	Postural muscle tone	Fine distal movements	Eyes	Memory	Responsiveness to external stimuli
1. Aura[a]	Normal	Normal	Normal	Open	Normal	Normal
2. Dyscognitive seizure	Normal	Normal	Normal	Open	Normal except with amnestic seizures	Altered selectively by the specific cognitive deficit (aphasia, apraxia, etc.)
3. Ictal delirium	Normal or hyperactive (insomnia)	Normal	Normal	Open	Mainly abnormal	Hyperactive Reacts to illusions and hallucinations as real perceptions
4. Dialeptic seizure	Normal	Normal	Normal	Open	Amnesia	Unresponsive or markedly reduced
5. Coma during epileptic seizures	Impaired	Impaired	Impaired	Open during the seizure Closed after the seizure	Amnesia	Totally unresponsive

[a]Includes only auras consisting of hallucinations or illusions. Auras consisting of sensations produced by autonomic or motor seizures that cannot be detected by an observer (example an abdominal aura produced by ictal gastric peristalsis) should not be included as seizures that produce alterations of consciousness.

Proposal: Different types of alteration and loss of consciousness in epilepsy

[1]Hans Lüders, [1]Shahram Amina, [1]Christopher Bailey, [2]Christoph Baumgartner, [3]Selim Benbadis, [4]Adriana Bermeo, [5]Maria Carreño, [1]Michael Devereaux, [6]Beate Diehl, [1]Matthew Eccher, [7]Jonathan Edwards, [1]Philip Fastenau, [1]Guadalupe Fernandez Baca-Vaca, [8]Jaime Godoy, [9]Hajo Hamer, [10]Seung Bong Hong, [11]Akio Ikeda, [12]Philippe Kahane, [1]Kitti Kaiboriboon, [13]Giridhar Kalamangalam, [14]David Lardizabal, [1]Samden Lhatoo, [15]Jürgen Lüders, [16]Jayanti Mani, [17]Carlos Mayor, [18]Tomas Mesa Latorre, [1]Jonathan Miller, [19]Harold H. Morris, [20]Soheyl Noachtar, [21]Cormac O'Donovan, [1]Jun Park, [22]Maria Angeles Perez-Jimenez, [23]Sabine Rona, [24]Felix Rosenow, [1]Asim Shahid, [25]Stephan Schuele, [26]Christopher Skidmore, [27]Bernhard Steinhoff, [28]Charles Á. Szabó, [1]Jennifer Sweet, [29]Nitin Tandon, [15]Adriana Tanner, and [30]Sadatoshi Tsuji

Epilepsia, 55(8):1140–1144, 2014
doi: 10.1111/epi.12595



Hans Lüders, specializes in Neurology at Case Medical Center, Cleveland, Ohio.

SUMMARY

There are at least five types of alterations of consciousness that occur during epileptic seizures: auras with illusions or hallucinations, dyscognitive seizures, epileptic delirium, dialeptic seizures, and epileptic coma. Each of these types of alterations of consciousness has a specific semiology and a distinct pathophysiologic mechanism. In this proposal we emphasize the need to clearly define each of these alterations/loss of consciousness and to apply this terminology in semiologic descriptions and classifications of epileptic seizures. The proposal is a consensus opinion of experienced epileptologists, and it is hoped that it will lead to systematic studies that will allow a scientific characterization of the different types of alterations/loss of consciousness described in this article.

KEY WORDS: Consciousness in epilepsy, Dialepsis, Dyscognitive seizures.

Dyscognitive - interference of higher cortical functioning (aphasia, apraxia, amnesia) with fully intact consciousness

Dialepsis - Dialepein is a synonym of epilepein (to seize or to stop) and consists of unresponsiveness or markedly decreased responsiveness to external stimuli

Epilepsia. 2014;55(8):1140-1144.

Controversy from 2010 classification

Insufficient scientific understanding in epileptogenesis to support novel classification scheme

No ground to replace existing time-proven systems

Too complicated

And…

- Dys is of Greek origin, while cognitive is of Latin origin

Chang et al. J Formos Med Assoc. 2017;116:736-741.

ILAE 2017 Classification of seizures and epilepsy

ILAE POSITION PAPER

Operational classification of seizure types by the International League Against Epilepsy: Position Paper of the ILAE Commission for Classification and Terminology

*Robert S. Fisher, †J. Helen Cross, ‡Jacqueline A. French, §Norimichi Higurashi, ¶Edouard Hirsch, #Floor E. Jansen, **Lieven Lagae, ††Solomon L. Moshé, ‡‡Jukka Peltola, §§Eliane Roulet Perez, ¶¶Ingrid E. Scheffer, and ##***Sameer M. Zuberi

Epilepsia, 58(4):522–530, 2017
doi: 10.1111/epi.13670

Dr. Robert S. Fisher, past president of American Epilepsy Society and editor of *Epilepsia* and epilepsy.com, led the Seizure Classification Task Force.

Summary

The International League Against Epilepsy (ILAE) presents a revised operational classification of seizure types. The purpose of such a revision is to recognize that some seizure types can have either a focal or generalized onset, to allow classification when the onset is unobserved, to include some missing seizure types, and to adopt more transparent names. Because current knowledge is insufficient to form a scientifically based classification, the 2017 Classification is operational (practical) and based on the 1981 Classification, extended in 2010. Changes include the following: (1) "partial" becomes "focal"; (2) awareness is used as a classifier of focal seizures; (3) the terms dyscognitive, simple partial, complex partial, psychic, and secondarily generalized are eliminated; (4) new focal seizure types include automatisms, behavior arrest, hyperkinetic, autonomic, cognitive, and emotional; (5) atonic, clonic, epileptic spasms, myoclonic, and tonic seizures can be of either focal or generalized onset; (6) focal to bilateral tonic–clonic seizure replaces secondarily generalized seizure; (7) new generalized seizure types are absence with eyelid myoclonia, myoclonic absence, myoclonic–atonic, myoclonic–tonic–clonic; and (8) seizures of unknown onset may have features that can still be classified. The new classification does not represent a fundamental change, but allows greater flexibility and transparency in naming seizure types.

KEY WORDS: Classification, Seizures, Focal, Generalized, Epilepsy, Taxonomy.

ILAE POSITION PAPER

ILAE classification of the epilepsies: Position paper of the ILAE Commission for Classification and Terminology

[1,2,3]Ingrid E. Scheffer, [1]Samuel Berkovic, [4]Giuseppe Capovilla, [5]Mary B. Connolly, [6]Jacqueline French, [7]Laura Guilhoto, [8,9]Edouard Hirsch, [10]Satish Jain, [11]Gary W. Mathern, [12]Solomon L. Moshé, [13]Douglas R. Nordli, [14]Emilio Perucca, [15]Torbjörn Tomson, [16]Samuel Wiebe, [17]Yue-Hua Zhang, and [18,19]Sameer M. Zuberi

Epilepsia, 58(4):512–521, 2017
doi: 10.1111/epi.13709

Dr. Ingrid E. Scheffer chairs the ILAE Task Force on the Classification of the Epilepsies.

Summary

The International League Against Epilepsy (ILAE) Classification of the Epilepsies has been updated to reflect our gain in understanding of the epilepsies and their underlying mechanisms following the major scientific advances that have taken place since the last ratified classification in 1989. As a critical tool for the practicing clinician, epilepsy classification must be relevant and dynamic to changes in thinking, yet robust and translatable to all areas of the globe. Its primary purpose is for diagnosis of patients, but it is also critical for epilepsy research, development of antiepileptic therapies, and communication around the world. The new classification originates from a draft document submitted for public comments in 2013, which was revised to incorporate extensive feedback from the international epilepsy community over several rounds of consultation. It presents three levels, starting with *seizure type*, where it assumes that the patient is having epileptic seizures as defined by the new 2017 ILAE Seizure Classification. After diagnosis of the *seizure type*, the next step is diagnosis of *epilepsy type*, including focal epilepsy, generalized epilepsy, combined generalized, and focal epilepsy, and also an unknown epilepsy group. The third level is that of *epilepsy syndrome*, where a specific syndromic diagnosis can be made. The new classification incorporates *etiology* along each stage, emphasizing the need to consider etiology at each step of diagnosis, as it often carries significant treatment implications. Etiology is broken into six subgroups, selected because of their potential therapeutic consequences. New terminology is introduced such as developmental and epileptic encephalopathy. The term benign is replaced by the terms self-limited and

Motivation for change in terminology

1. Some seizure types can have either a focal or generalized onset

2. Lack of knowledge about the onset makes a seizure unclassifiable and difficult to discuss with the 1981 system

3. Retrospective seizure descriptions often do not specify a level of consciousness, and altered consciousness, although central to many seizures, is a complicated concept

4. Some terms in current use do not have high levels of community acceptance or public understanding, such as, “psychic,” “partial,” “simple partial,” “complex partial,” and “dyscognitive”

5. Some important seizure types are not included

Epilepsia. 2017;58(4):531-542.

ILAE 2017 classification

Proposed to recognize and classify seizures **based on clinical characteristics** as opposed to the pathogenesis

Focal seizures - Focal aware seizures (FAS) or Focal impaired awareness seizures (FIAS); subdivided into motor/nonmotor

Generalized seizures - Motor or Nonmotor (absence)

Epilepsy syndromes defined further, e.g., mesial temporal lobe epilepsy (mTLE) and juvenile myoclonic epilepsy (JME)

Epilepsia. 2017;58(4):531-542.

Changes in seizure classification from 1981-2017

1. Change of “partial” to “focal”

   - Partial conveys a sense of part of a seizure, rather than a location or anatomic system; focal is more understandable in terms of seizure-onset location

2. Certain seizure types can be either of focal, generalized, or unknown onset

3. Seizure of unknown onset may have features that can still be classified

4. Awareness is used as a classifier of focal seizures

Epilepsia. 2017;58(4):531-542.

Changes in seizure classification from 1981-2017

5. The terms dyscognitive, simple partial, complex partial, psychic, and secondary generalized were eliminated

6. New focal seizure types include automatisms, autonomic, behavioral arrest, cognitive, emotional, hyperkinetic, sensory, and focal to bilateral tonic-clonic seizures. Atonic, clonic, epileptic spams, myoclonic, and tonic seizures can be either focal or generalized

7. New generalized seizure types include absence with eyelid myoclonia, myoclonic absence, myoclonic-tonic-clonic, myoclonic-atonic, and epileptic spasms

Epilepsia. 2017;58(4):531-542.

ILAE 2017 Classification of Seizure Types Expanded Version [1]

Focal Onset

Aware | Impaired Awareness

Motor Onset
- automatisms
- atonic [2]
- clonic
- epileptic spasms [2]
- hyperkinetic
- myoclonic
- tonic

Nonmotor 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

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

Unknown Onset

Motor
- tonic–clonic
- epileptic spasms

Nonmotor
- behavior arrest

Unclassified [3]

Epilepsia. 2017;58(4):531-542.

ILAE 2017 classification

Focal seizures

- "Originating within networks limited to one hemisphere. They may be discretely localized or more widely distributed. Focal seizures may originate in subcortical structures."

  - Focal is preferred over partial, because focal is more understandable in terms of seizure-onset localization

Generalized seizures

- "Originating at some point within, and rapidly engaging, bilaterally distributed networks."

Epilepsia. 2017;58(4):531-542.

ILAE 2017 classification

Consciousness and awareness

- Retained awareness
  - "Seizure with no impairment of consciousness;" retain awareness as knowledge of self and environment
  - Dyscognitive no longer included in the classification of impaired awareness (complex partial) seizures, because of lack of clarity and negative public and professional feedback
- Not included as a classifier for generalized seizures, because the majority of generalized seizures present with impaired awareness

Epilepsia. 2017;58(4):531-542.

ILAE 2017 classification

Discontinued terms

- Simple/complex partial
- Convulsion

Epilepsia. 2017;58(4):531-542.

ILAE 2017 classification

Added terms

- Hyperkinetic
  - Hypermotor is a combination of Greek and Latin and hyperkinetic is more historically consistent
- Cognitive replaces psychic
  - Cognitive seizure can also comprise positive cognitive phenomena, such as deja vu or hallucinations
- Emotional, e.g., fear or joy

Epilepsia. 2017;58(4):531-542.

ILAE 2017 classification

Added terms

- New focal seizure types, e.g., epileptic spasms, tonic, clonic, atonic, and myoclonic seizures

- New generalized seizure types, e.g., absence with eyelid myoclonia, myoclonic-atonic, and myoclonic-tonic-clonic (clonic-tonic-clonic was mentioned in the 1981 publication)

- Epileptic is implied with every seizure type, but now specifically used for epileptic spasms (spams was too vague)

Epilepsia. 2017;58(4):531-542.

OLD

Onset localization

- Partial
- Generalized

Level of awareness

- Simple
- Complex

- Secondary generalization

Disorder

NEW

- Focal
- Generalized
- Unknown

- Aware
- Impaired awareness

- Focal to bilateral tonic-clonic seizures

Disease

ILAE 2017 Classification of seizures and epilepsy

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Dr. Robert S. Fisher, past president of American Epilepsy Society and editor of *Epilepsia* and epilepsy.com, led the Seizure Classification Task Force.

Summary

The International League Against Epilepsy (ILAE) presents a revised operational classification of seizure types. The purpose of such a revision is to recognize that some seizure types can have either a focal or generalized onset, to allow classification when the onset is unobserved, to include some missing seizure types, and to adopt more transparent names. Because current knowledge is insufficient to form a scientifically based classification, the 2017 Classification is operational (practical) and based on the 1981 Classification, extended in 2010. Changes include the following: (1) "partial" becomes "focal"; (2) awareness is used as a classifier of focal seizures; (3) the terms dyscognitive, simple partial, complex partial, psychic, and secondarily generalized are eliminated; (4) new focal seizure types include automatisms, behavior arrest, hyperkinetic, autonomic, cognitive, and emotional; (5) atonic, clonic, epileptic spasms, myoclonic, and tonic seizures can be of either focal or generalized onset; (6) focal to bilateral tonic–clonic seizure replaces secondarily generalized seizure; (7) new generalized seizure types are absence with eyelid myoclonia, myoclonic absence, myoclonic–atonic, myoclonic–tonic–clonic; and (8) seizures of unknown onset may have features that can still be classified. The new classification does not represent a fundamental change, but allows greater flexibility and transparency in naming seizure types.

KEY WORDS: Classification, Seizures, Focal, Generalized, Epilepsy, Taxonomy.

ILAE POSITION PAPER

ILAE classification of the epilepsies: Position paper of the ILAE Commission for Classification and Terminology

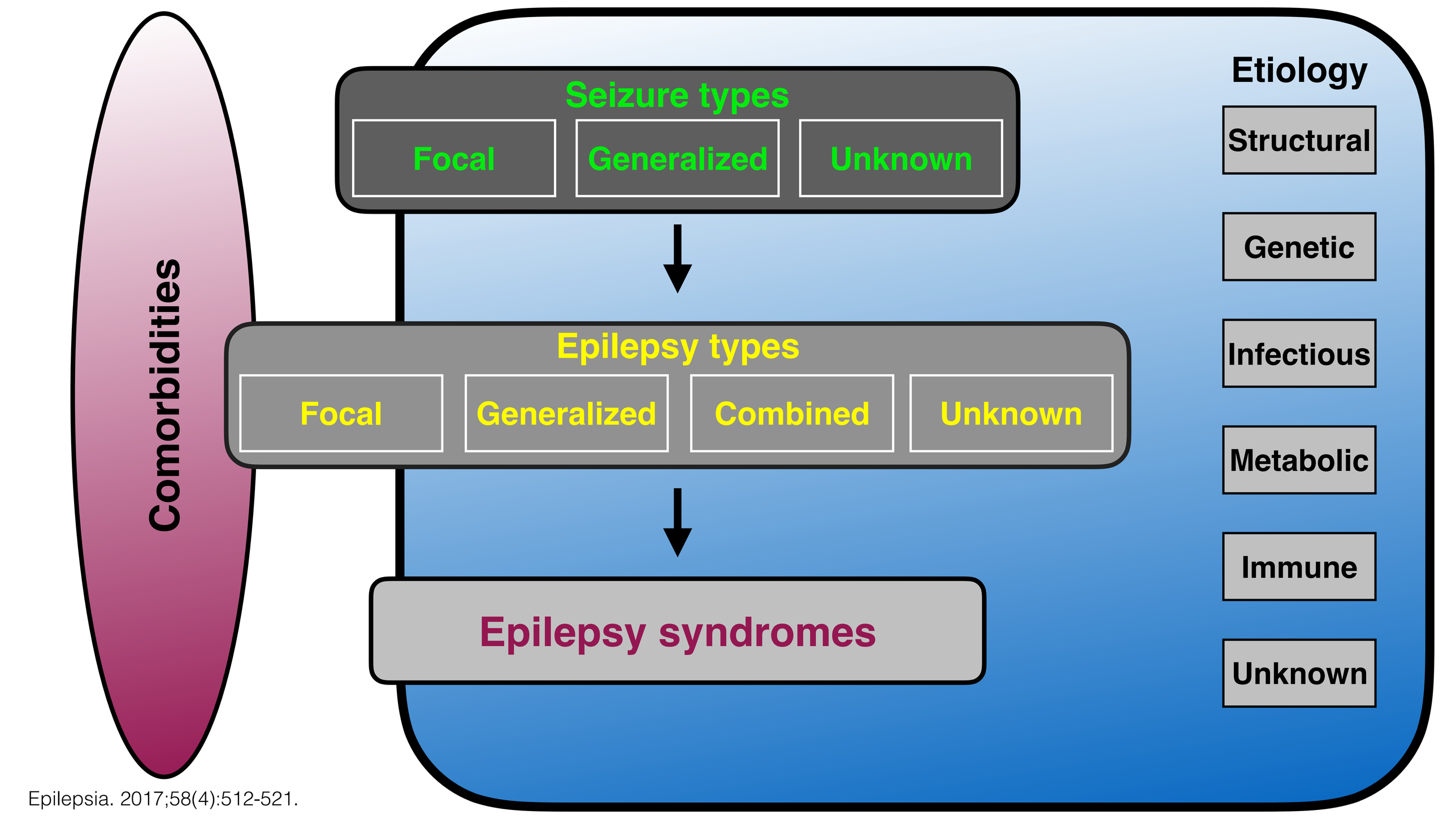
[1,2,3]Ingrid E. Scheffer, [1]Samuel Berkovic, [4]Giuseppe Capovilla, [5]Mary B. Connolly, [6]Jacqueline French, [7]Laura Guilhoto, [8,9]Edouard Hirsch, [10]Satish Jain, [11]Gary W. Mathern, [12]Solomon L. Moshé, [13]Douglas R. Nordli, [14]Emilio Perucca, [15]Torbjörn Tomson, [16]Samuel Wiebe, [17]Yue-Hua Zhang, and [18,19]Sameer M. Zuberi

Epilepsia, 58(4):512–521, 2017
doi: 10.1111/epi.13709

Dr. Ingrid E. Scheffer chairs the ILAE Task Force on the Classification of the Epilepsies.

Summary

The International League Against Epilepsy (ILAE) Classification of the Epilepsies has been updated to reflect our gain in understanding of the epilepsies and their underlying mechanisms following the major scientific advances that have taken place since the last ratified classification in 1989. As a critical tool for the practicing clinician, epilepsy classification must be relevant and dynamic to changes in thinking, yet robust and translatable to all areas of the globe. Its primary purpose is for diagnosis of patients, but it is also critical for epilepsy research, development of antiepileptic therapies, and communication around the world. The new classification originates from a draft document submitted for public comments in 2013, which was revised to incorporate extensive feedback from the international epilepsy community over several rounds of consultation. It presents three levels, starting with *seizure type*, where it assumes that the patient is having epileptic seizures as defined by the new 2017 ILAE Seizure Classification. After diagnosis of the *seizure type*, the next step is diagnosis of *epilepsy type*, including focal epilepsy, generalized epilepsy, combined generalized, and focal epilepsy, and also an unknown epilepsy group. The third level is that of *epilepsy syndrome*, where a specific syndromic diagnosis can be made. The new classification incorporates *etiology* along each stage, emphasizing the need to consider etiology at each step of diagnosis, as it often carries significant treatment implications. Etiology is broken into six subgroups, selected because of their potential therapeutic consequences. New terminology is introduced such as developmental and epileptic encephalopathy. The term benign is replaced by the terms self-limited and



Epilepsia. 2017;58(4):512-521.

Epilepsia, 51(4):676–685, 2010
doi: 10.1111/j.1528-1167.2010.02522.x

SPECIAL REPORT

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

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Genetic generalized epilepsy

ILAE POSITION PAPER

ILAE classification of the epilepsies: Position paper of the ILAE Commission for Classification and Terminology

[1,2,3]Ingrid E. Scheffer, [1]Samuel Berkovic, [4]Giuseppe Capovilla, [5]Mary B. Connolly, [6]Jacqueline French, [7]Laura Guilhoto, [8,9]Edouard Hirsch, [10]Satish Jain, [11]Gary W. Mathern, [12]Solomon L. Moshé, [13]Douglas R. Nordli, [14]Emilio Perucca, [15]Torbjörn Tomson, [16]Samuel Wiebe, [17]Yue-Hua Zhang, and [18,19]Sameer M. Zuberi

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Dr. Ingrid E. Scheffer chairs the ILAE Task Force on the Classification of the Epilepsies.

Summary

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Idiopathic generalized epilepsy

Epilepsia. 2017;58(4):512-521.

Idiopathic generalized epilepsy

1. Childhood absence epilepsy

2. Juvenile absence epilepsy

3. Juvenile myoclonic epilepsy

4. Generalized tonic-clonic seizures

May be reasonable to refer to this group as genetic generalized epilepsies (GGE) when there is sufficient evidence for this classification

Epilepsia. 2017;58(4):512-521.

Epileptic encephalopathies

Epileptic activity contributes to severe cognitive and behavioral impairments above and beyond what might be expected from the underlying pathology alone

Term developmental and epileptic encephalopathy may be used when appropriate

1. West syndrome

2. Dravet syndrome

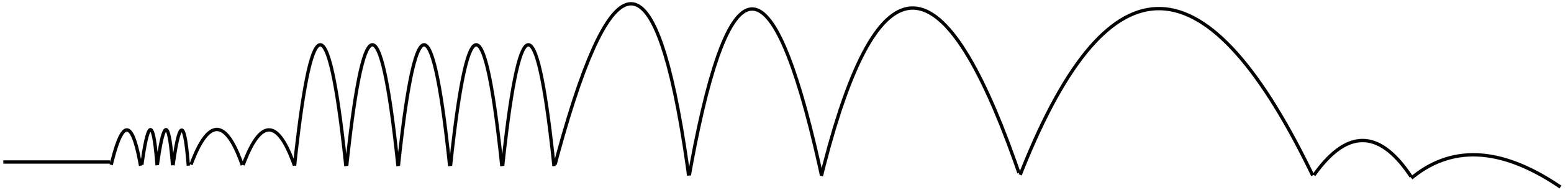
3. Lennox-Gastaut syndrome

Epilepsia. 2017;58(4):512-521.

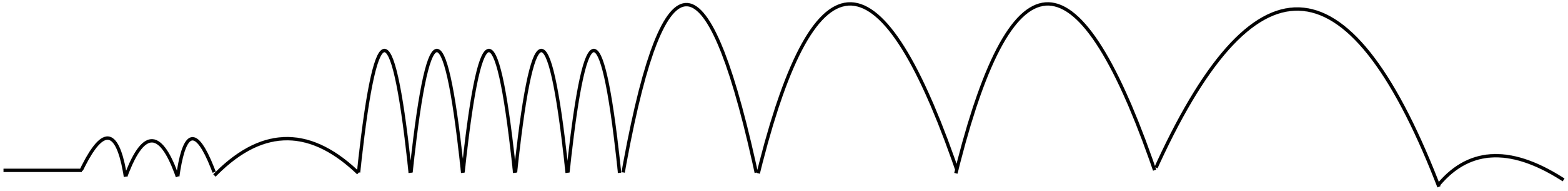
Discontinued and new terms

1. Benign
   - Underestimates disease burden
   - Self-limited and pharmacoresponsive
2. Malignant and catastrophic
   - Serious and devastating connotations

Epilepsia. 2017;58(4):512-521.



Status epilepticus definitions

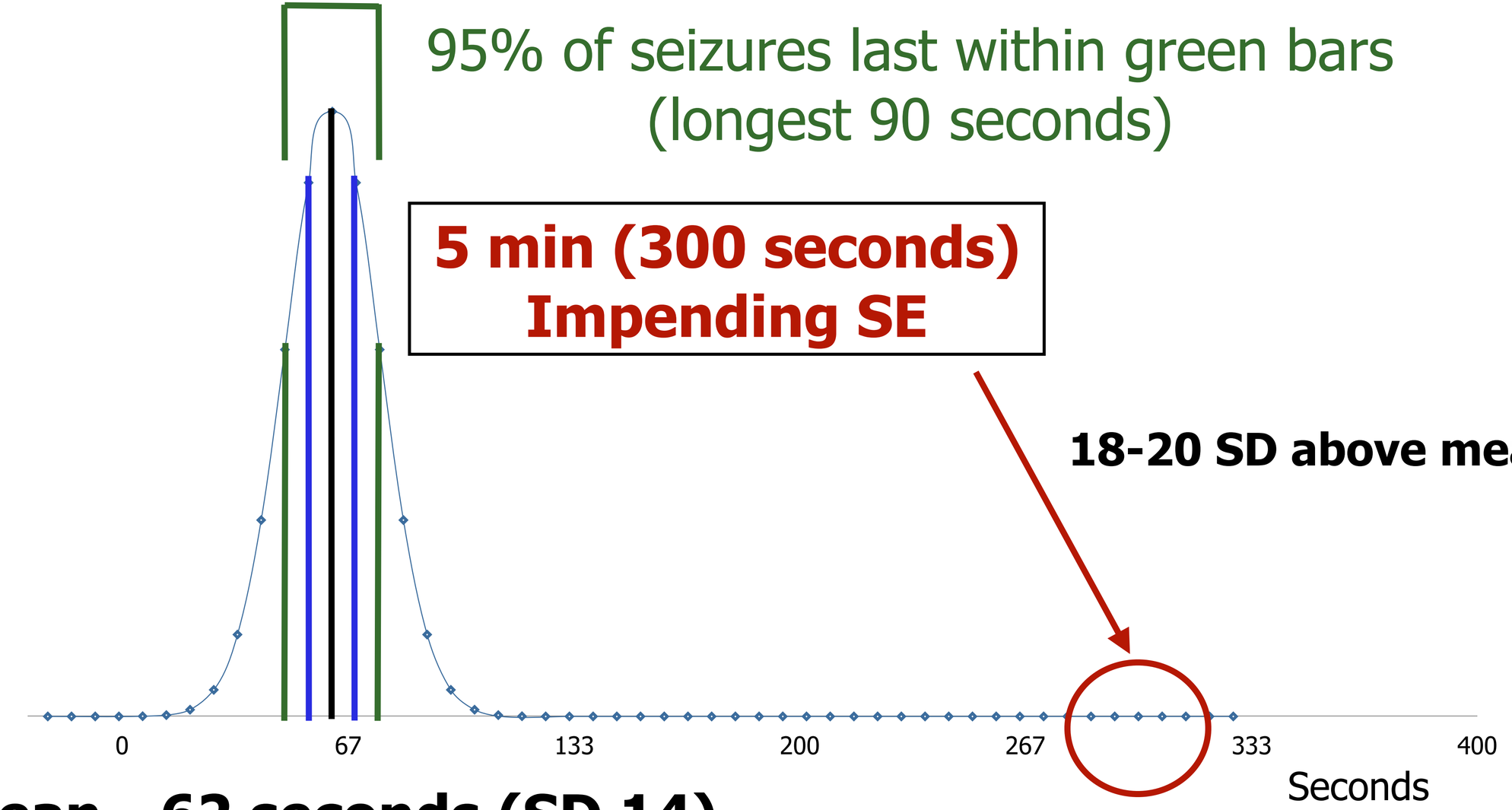


Beaumont

Seizure duration

95% of seizures last within green bars
(longest 90 seconds)

5 min (300 seconds)
Impending SE



Mean - 62 seconds (SD 14)

Lancet Neurol. 2006;5:246-256.

Status epilepticus defined

SE is 5 minutes or more of:

1. Continuous clinical and/or electrographic seizure activity
2. Recurrent seizure activity without recovery (return to baseline)

Most seizures last less than 5 minutes and seizures more than 5 minutes often do not stop spontaneously
Animal data suggests there may be permanent neurological injury and drug resistance before 30 minutes
Experts have revised the definition

Impending SE (early SE or early heralds of status) for seizures lasting > 5 minutes, but eventually resolve

Lancet Neurol. 2006;5:246-256.
Neurocrit Care. 2012;17(1):3-23.

Time point 1 (t1)
Seizure is abnormally prolonged (5 minutes)

Time point 2 (t2)
Ongoing seizure beyond which there is increased risk for long-term consequences (30 minutes)

SPECIAL REPORT

A definition and classification of status epilepticus – Report of the ILAE Task Force on Classification of Status Epilepticus

*†‡Eugen Trinka, §Hannah Cock, ¶Dale Hesdorffer, #Andrea O. Rossetti, **Ingrid E. Scheffer, ††Shlomo Shinnar, ‡‡Simon Shorvon, and §§Daniel H. Lowenstein

Epilepsia, 56(10):1515–1523, 2015
doi: 10.1111/epi.13121

Eugen Trinka is professor and chairman of Department of Neurology, Paracelsus Medical University Salzburg Austria.

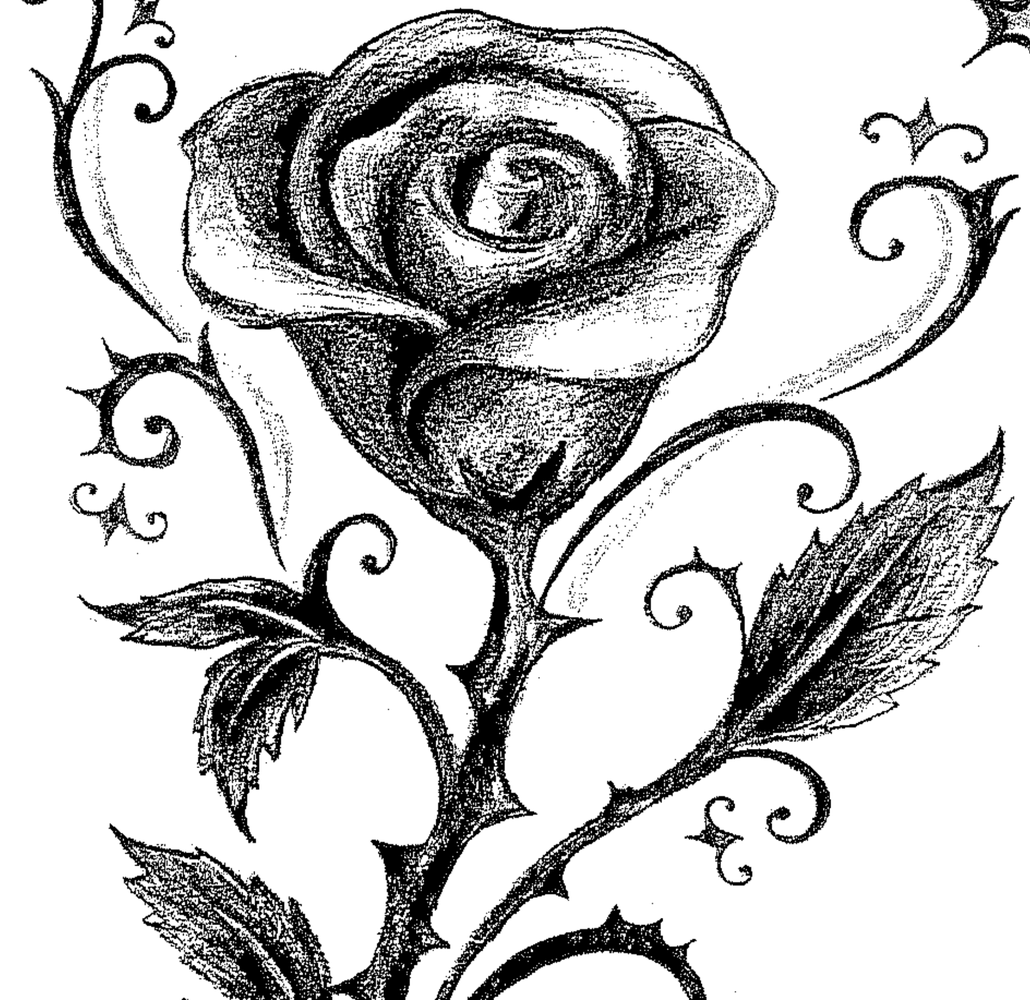
Summary

The Commission on Classification and Terminology and the Commission on Epidemiology of the International League Against Epilepsy (ILAE) have charged a Task Force to revise concepts, definition, and classification of status epilepticus (SE). The proposed new definition of SE is as follows: *Status epilepticus is a condition resulting either from the failure of the mechanisms responsible for seizure termination or from the initiation of mechanisms, which lead to abnormally, prolonged seizures (after time point t_1). It is a condition, which can have long-term consequences (after time point t_2), including neuronal death, neuronal injury, and alteration of neuronal networks, depending on the type and duration of seizures.* This definition is conceptual, with two operational dimensions: the first is the length of the seizure and the time point (t_1) beyond which the seizure should be regarded as "continuous seizure activity." The second time point (t_2) is the time of ongoing seizure activity after which there is a risk of long-term consequences. In the case of convulsive (tonic–clonic) SE, both time points (t_1 at 5 min and t_2 at 30 min) are based on animal experiments and clinical research. This evidence is incomplete, and there is furthermore considerable variation, so these time points should be considered as the best estimates currently available. Data are not yet available for other forms of SE, but as knowledge and understanding increase, time points can be defined for specific forms of SE based on scientific evidence and incorporated into the definition, without changing the underlying concepts. A *new diagnostic classification system* of SE is proposed, which will provide a framework for clinical diagnosis, investigation, and therapeutic approaches for each patient. There are four axes: (1) semiology; (2) etiology; (3) electroencephalography (EEG) correlates; and (4) age. Axis 1 (semiology) lists different forms of SE divided into those with prominent motor systems, those without prominent motor systems, and currently indeterminate conditions (such as acute confusional states with epileptiform EEG patterns). Axis 2 (etiology) is divided into subcategories of known and unknown causes. Axis 3 (EEG correlates) adopts the latest recommendations by consensus panels to use the following descriptors for the EEG: name of pattern, morphology, location, time-related features, modulation, and effect of intervention. Finally, axis 4 divides age groups into neonatal, infancy, childhood, adolescent and adulthood, and elderly.

KEY WORDS: Status epilepticus, Seizure, Definition, Classification, Seizure duration.

Romeo and Juliet

BY WILLIAM SHAKESPEARE



'Tis but thy name that is mine enemy:
What's Montague? Is it not hand nor foot,
Nor arm, nor face, nor any other part.
What's in a name? That which we call a rose,
By any other name would smell as sweet.

Because it is my name! Because I cannot have another in my life. Because I am not worth the dust on the feet of them that hang! How may I live without my name? I have given you my soul, leave my name!

Arthur Miller's

The Crucible

Conclusions

"If names be not correct, language is not in accordance with the truth of things. If language be not in accordance with the truth of things, affairs cannot be carried on to success," Confucius, Analects, 6th century BC.

Old terminology	ILAE 2017 New terminology
Grand mal	Generalized tonic-clonic seizure

Epilepsia. 2017;58(4):531-542.

Old terminology	ILAE 2017 New terminology
Complex partial seizure	Focal impaired awareness seizure

Epilepsia. 2017;58(4):531-542.

Old terminology	ILAE 2017 New terminology
Grand mal or clonic-tonic-clonic seizure	Generalized tonic-clonic seizure or myoclonic-tonic-clonic seizure

Epilepsia. 2017;58(4):531-542.

Old terminology	ILAE 2017 New terminology
Complex partial seizure	Focal motor (hyperkinetic) impaired awareness seizure

Epilepsia. 2017;58(4):531-542.

Old terminology	ILAE 2017 New terminology
Simple partial seizure or aura	Focal aware seizure or focal nonmotor sensory aware seizure

Epilepsia. 2017;58(4):531-542.

Old terminology	ILAE 2017 New terminology
Complex partial seizure	Focal (automatisms) impaired awareness seizure

Epilepsia. 2017;58(4):531-542.

Old terminology	ILAE 2017 New terminology
Complex partial seizure	Focal motor (hyperkinetic) impaired awareness seizure

Epilepsia. 2017;58(4):531-542.

Old terminology	ILAE 2017 New terminology
Tonic seizure	Generalized motor tonic seizure

Epilepsia. 2017;58(4):531-542.

Old terminology	ILAE 2017 New terminology
Typical absence seizure	Generalized nonmotor typical absence seizure

Epilepsia. 2017;58(4):531-542.

Old terminology	ILAE 2017 New terminology
Simple partial seizure	Focal aware seizure

Epilepsia. 2017;58(4):531-542.