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The new classification of epilepsy and seizures of the International League Against Epilepsy (ILAE) 2017

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Summary

In 2017, the International League Against Epilepsy (ILAE) published "position papers" on a new classification of epilepsies [1] and seizures [2], as well as an "instruction manual" for use in the operational classification of seizures [3]. An ad hoc taskforce of the German speaking Leagues against Epilepsy translated these papers into German with authorisation of the ILAE (see appendix 1).

Keywords: *classification, epilepsy, seizures, 2017, International League Against Epilepsy*

The classification of epilepsy and seizures undergoes continual evaluation by the ILAE. Given the dramatic increase of our knowledge in neuroscience and computional network sciences over recent decades, the last, highly influential and broadly used classification system for epilepsy (published in 1985 and 1989) and seizures (published in 1981) were revised. The classification of epilepsy is the most important tool for evaluating a patient suffering from seizures. It guides all clinical assessments and therapeutic decisions. Beyond its clinical impact, it influences any basic and clinical research, public health and epilepsy care policies, as well as insurance issues.

Classification systems are almost always fiercely debated, as they have to be easy to use but also precise, which are often difficult to reconcile. Thus, the current classification was established after extensive consultations with the eminent experts in clinical epileptology and basic science, as well as with patients, organisations and epilepsy leagues across the world.

The new classification system suggests: firstly, to ascertain if the paroxysmal event was truly an epileptic seizure; secondly, to determine whether the seizure was focal, generalised or unknown regarding onset; and thirdly, to classify epilepsy according to the seizure type (focal, generalised, combined focal and generalised, or unclassified). At first presentation, the aetiology/aetiologies (structural, genetic, infectious, metabolic, immune, or unknown) of epilepsy should be determined and comorbidities, such as depression, anxiety disorder, etc. should be sought. All this information may allow identification of a specific epilepsy syndrome or a non-syndromal epilepsy (fig. 1).

What is new regarding the classification of epilepsy?

Four changes have to be mentioned. First, epilepsy cannot be just focal or generalised, but also combined focal AND generalised. Second, epilepsies previously called "benign" are now termed "self-limiting" or "pharmacoresponsive" to highlight that epilepsy is never "benign", but associated with a burden. Epilepsy, however, can spontaneously resolve or respond to therapy and can therefore be classified as self-limiting and/or pharmacoresponsive. Third, patients with epilepsy and encephalopathy may suffer from an "epileptic", "developmental" or "epileptic and developmental" encephalopathy. This takes into account that the epileptic process and seizure burden may lead to encephalopathy in some cases (e.g., epileptic status epilepticus in sleep), whereas in others a severe developmental delay is associated with epilepsy, but the encephalopathy results mainly from the metabolic, genetic or structural disorder (such as in autism). Or there is an early developmental component that is further aggravated by the additional effect of severe seizures (such as in Dravet syndrome). Fourth, psychosocial and other comorbidities, which can fundamentally influence the course and treatability of epilepsies have been included into the characterisation and the conceptual definition of epilepsy [4] (fig. 1).

What is new regarding the classification of seizures?

Some characterising terms used to classify seizures were eliminated or replaced by new ones (table 1). The most important change concerns the term "partial", which becomes "focal" underscoring the fundamental fact that the seizure is neither "incomplete" nor a "part" of a seizure, but has a spatially located onset and symptomatogenic zone. In addition, awareness has become a classifier and thus the terms "simple" and "complex" were replaced by the terms "aware" and "impaired awareness". This change will improve understanding, especially by lay people who can better follow the concept that awareness is fully present in some seizures and not in others, instead of wondering whether they have "simple" or "complex" (with regard to what?) seizures. Of note, it was difficult to translate the expression "awareness" into German as the term "Bewusstsein" means rather "consciousness" than "awareness". The taskforce decided (after extensive discussion) to use the terms "bewusst erlebt" and "nicht-bewusst erlebt".

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Figure 1: The multi-dimensional classification scheme of epilepsy according to the ILAE 2017 operational classification of epilepsy (with permission from *Epilepsia,* the Journal of the ILAE).

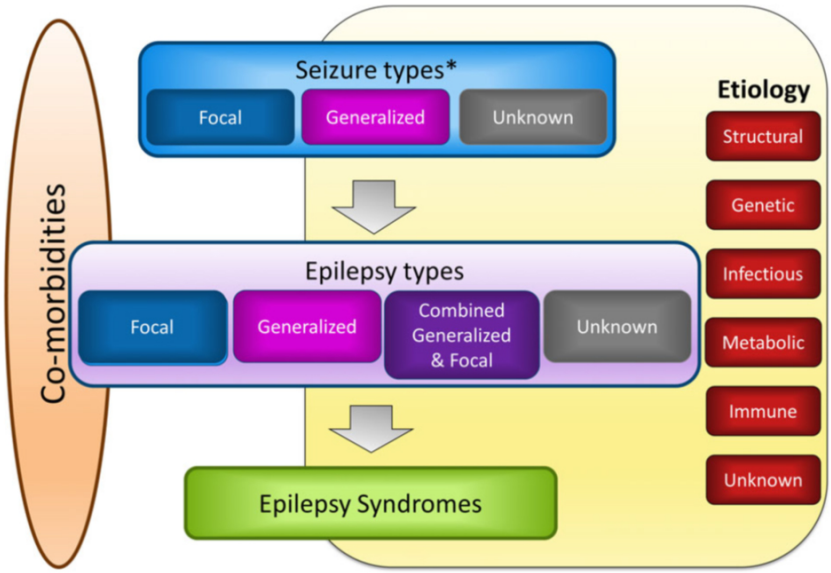

Table 1: The most important changes of the 2017 classification of epilepsy and seizures.

Epilepsy	Eliminated terms	Malignant/catastrophic epilepsy	
		Symptomatic generalised epilepsy	
	New terms	Combined focal and generalised epilepsy	
		Epileptic encephalopathy (seizure burden by itself leads to encephalopathy)	
		Developmental and epileptic encephalopathy (both seizure burden AND metabolic, structural or genetic alterations lead to encephalopathy)	
	Changed terms	*Old*	*New*
		Genetic generalised epilepsy (GGE) may be still used when a genetic background is suspected or evident)	"New-old" term: idiopathic generalised epilepsies (IGE)
		Benign	Self-limiting (spontaneously resolving)
			Pharmacoresponsive (easily controlled by appropriate medication)
Seizures	Eliminated terms	Dyscognitive	
		Convulsion	
	New term	Emotional (fear, joy, orgasmic, specific individual emotions)	
	New seizure types	Focal	Automatisms
			Autonomic
			With behaviour arrest
			Sensory
			Atonic
			Clonic
			Epileptic spasms
			Myoclonic
			Tonic
		Generalised	Absence with eyelid myoclonia (Jeavons syndrome)
			Myoclonic absence
			Myoclonic-tonic-clonic
			Myoclonic-atonic (Doose syndrome)
			Epileptic spasms (West syndrome, infantile spasms, Blitz-Nick-Salaam seizures)
	Changed terms	*Old*	*New*
		Partial seizure	Focal (onset) seizure
		Secondarily generalised clonic	Focal to bilateral tonic-clonic
		Simple	Aware
		Complex	Impaired awareness
		Hypermotor	Hyperkinetic
		Psychic/experiential	Cognitive

The ILAE 2017 classifications have already been the topic of numerous reviews [5–9]. Furthermore, they were evaluated in a few studies, which showed that the new classifications indeed increase the precision of diagnoses, but at the cost of leaving more epilepsies classifiable at the mode of onset level only [10, 11]. The new classifications will hopefully be gradually adopted and those changes regarded helpful may be incorporated into daily practice in order to improve diagnosis and care, and eventually the lives of our patients we care for – the main goal and motivation of every epilepsy professional [6, 12].

Key points

- The ILAE presents a revised classification of the epilepsies, complemented by a classification of the different forms of seizures.
- Diagnosis of epilepsy includes a definition of the seizure form, type of epilepsy (focal, generalised, combined focal and generalised, unclassified) and epilepsy syndrome.
- The aetiology of epilepsy should be established at the first neurological assessment and further refined with each diagnostic step. Epilepsy can be based on more than one aetiological category.
- The term "benign" is replaced by the terms "self-limiting" and "pharmaco-responsive", each to be used when appropriate.
- The term "developmental and epileptic encephalopathy" can be used to describe both entities occurring together, or can be adapted to describe either of the two entities separately.
- Several terms describing seizures have been eliminated or replaced (see table 1).

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Disclosure statement
The author declares no conflict of interest regarding this manuscript, except for his position as the president of the Swiss League against Epilepsy.

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Appendix 1
Ad-hoc taskforce of the Deutschen Gesellschaft für Epileptologie (DGfE), the Oesterreichischen Gesellschaft für Epileptologie (ÖGfE) and Schweizerischen Epilepsie-Liga (SEL) (in alphabetical order):

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Univ. Prof. Dr. Mag. Eugen Trinka, FRCP, Salzburg, Austria

Links:

https://www.ilae.org/files/ilaeGuideline/Classification-Scheffer-2017-GERMAN.pdf (classification of epilepsy)

https://link.springer.com/article/10.1007/s10309-018-0218-6 (classification of epilepsy)

https://link.springer.com/article/10.1007/s10309-018-0216-8 (classification of seizures)

https://link.springer.com/article/10.1007/s10309-018-0217-7 (instruction manual)