Original article

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Applicability and contribution of the new ILAE 2017 classification of epileptic seizures and epilepsies*

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ABSTRACT – *Aim*. The aim of the study was to evaluate the clinical applicability of the 2017 ILAE classification of seizures and epilepsies through the analysis of a sample of 100 outpatients with a diagnosis of epilepsy.
Methods. All clinical charts were reviewed applying both the 1981/1989 and 2017 classifications of seizures and epilepsies, respectively. For most focal seizures, descriptors were required to include all the relevant clinical information. The reclassification of complex partial seizures into focal seizures with impaired awareness with a motor / non-motor onset allowed the inclusion of features of topographic value, although the chronological sequence of awareness impairment was lacking.
Results. The use of the term "focal to bilateral tonic-clonic" reduced the number of seizures classified as generalized tonic-clonic seizures (GTCS) by 19%. A subset of GTCS (35%) and absence seizures (12.5%) were reclassified as seizures of unknown onset. Most focal symptomatic epilepsies (92%) were reclassified as focal structural epilepsies, while 27% of idiopathic generalized and 7% of focal cryptogenic epilepsies merged into the category of "epilepsies of unknown type".
Conclusion. Major strengths of the new classification are simplicity and the role of the category "unknown onset" to avoid forced categorization. A section assigned to uncertainty reinforces the need for further ancillary studies and periodic diagnostic re-evaluation.

Key words: seizures, epilepsy, classifications, applicability

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The classification of epileptic seizures and epilepsies has always been a subject of extensive debate given the complexity of ictal semiology and the aetiological heterogeneity. The most widely used classifications have been those proposed by the International League Against Epilepsy (ILAE) in 1981 for epileptic seizures (Commission on Classification and Terminology, 1981) and in 1989 for epilepsies (Commission on Classification and Terminology, 1989). An update of these classifications that could include new advances in the knowledge of electroclinical and aetiological correlations of seizures and epilepsies has been attempted for several years, including a stage of open debate within the international epileptological community. In 2017, the ILAE published a new classification (Fisher *et al*., 2017; Scheffer *et al*., 2017), which includes new categories, modifications in the nomenclature, and changes in aetiological systematization.
The aim of this study was to compare the new classification with those of 1981 and 1989 in terms of clinical applicability.

Methods

We included a cohort of 100 consecutive outpatients with an established diagnosis of epilepsy (Fisher *et al*., 2014) from the clinical database of patients treated at the Epilepsy Clinic of the Neurological Institute in Montevideo, a university tertiary centre. Only patients with at least one EEG and an imaging study were included.
Clinical data charts were reviewed. Based on 213 clinical descriptions of seizures provided by patients or witnesses, as stated in medical records, they were classified both according to the 1981 and 2017 ILAE classifications. Available EEG and imaging data were also considered. In the same subset of patients, the diagnosis of epilepsy type was ascertained, following the 1989 and 2017 ILAE classifications, according to published criteria.
As a first step, the variability across raters was evaluated in a sample of 10 patients. The nine raters (three senior epileptologists and six junior neurologists/residents) independently assessed these cases following the old and new classifications of seizures and epilepsies. A reliability analysis was performed using the Intraclass Correlation Coefficient (ICC, CI 95%, two-way, mixed effects model, consistency definition, average of multiple raters), using SPSS 11.5 version. The inter-rater agreement for 1989/2017 epilepsy and 1981/2017 seizure classifications is shown in *table 1* for the overall group of raters and according to level-of-training subgroups. The inter-rater agreement was excellent when applying the 1989 classification for epilepsies regardless of the raterĭs level of expertise. The ICC for the 2017 classification system for epilepsies was significantly lower, although it can be rated as good. In the overall group of raters, the ICC was good to excellent for both seizure classifications, without a statistically significant difference. Junior raters achieved a significantly lower ICC with a wider range of variation when applying the new seizure classification.
The remaining 90 cases were analysed by at least two researchers. Disagreements were resolved by consensus between the nine raters.

Results

Classification of epileptic seizures

For simple partial seizures with motor signs, the correlation between the different types (1981) and more frequent subtypes (2017) is shown in *table 2*. While in the previous classification, the subtypes of motor seizures alluded to body topographies, in the new classification they refer to types of movements (*e.g.* tonic, clonic, *etc*.). From those 21 patients with simple partial motor seizures, 14 were reclassified as focal aware clonic seizures. The focal seizures reclassified as tonic mostly correspond to postural and versive seizures under the previous classification. In *table 3,* we summarize the data of simple partial seizures with psychic symptoms (1981), as an example of the denomination variants for focal seizures with predominant non-motor symptomatology. Within this group, the nomenclature of affective seizures was changed to

Table 1. Inter-rater agreement for 1989/2017 epilepsy and 1981/2017 seizure classifications (ICC, 95% CI, two-way, mixed effects model, consistency definition).

CLASSIFICATION	Year	ICC Global	95% CI	ICC Senior	95% CI	ICC Junior	95% CI
Epileptic seizures	1981	0.92	0.807-0.979	0.911	0.783-0.969	0.847	0.632-0.956
	2017	0.867	0.669-0.969	0.998	0.996-0.999	0.527	-0.265-0.891
Epilepsies	1989	0.976	0.940-0.994	0.951	0.857-0.987	0.968	0.916-0.993
	2017	0.873	0.679-0.970	0.658	0-0.908	0.796	0.456-0.953

Table 2. Correlative distribution of simple partial seizures with motor signs (1981), relative to the ILAE 2017 classification.

1981 SEIZURE CLASSIFICATION	2017 SEIZURE CLASSIFICATION
SIMPLE PARTIAL SEIZURES WITH MOTOR SIGNS (1981)	**FOCAL AWARE WITH MOTOR ONSET (2017)**
UNSPECIFIED (9)	CLONIC (14)
WITHOUT JACKSONIAN MARCH (5)	
WITH JACKSONIAN MARCH (2)	TO BILATERAL TONIC CLONIC (3)
PHONATORY (1)	
VERSIVE (3)	TONIC (4)
POSTURAL (1)	

Table 3. Reclassification of simple partial seizures with psychic symptoms (1981 classification) in the 2017 seizure classification.

1981 CLASSIFICATION	2017 CLASSIFICATION
COMPLEX PARTIAL SEIZURES	FOCAL-ONSET IMPAIRED AWARENESS
IMPAIRMENT OF CONSCIOUSNESS AT ONSET (15)	MOTOR (6)
	NON-MOTOR (9) -Cognitive (2) -Behaviour arrest (2)
SIMPLE PARTIAL ONSET FOLLOWED BY IMPAIRMENT OF CONSCIOUSNESS (9)	MOTOR (1)
	NON-MOTOR (8) -Automatisms (3)
WITHOUT INFORMATION ABOUT CONSCIOUSNESS AT ONSET (15)	MOTOR (6)
	NON-MOTOR (7)
	UNCLASSIFIABLE (2)

focal aware non-motor emotional seizures. On the other hand, the remaining types of seizures had to be merged into the category of "cognitive seizures", with the subsequent use of the corresponding descriptors. Moreover, simple partial seizures with somatosensory or special sensory symptoms were reclassified as focal aware sensory seizures. The application of the expanded version of the new classification required the use of suggested descriptors (visual, auditory, *etc.*), equivalent to the subtype nomenclature of the 1981 classification. Autonomic simple partial seizures were renamed as focal aware non-motor autonomic seizures, maintaining the use of similar descriptors used in the previous classification.

The reclassification of complex partial seizures is shown in *table 4*. Among 15 complex partial seizures with an impairment of consciousness at onset, 10 were now assigned to different subtypes according to their initial semiological features: six with motor onset and, out of the nine cases with non-motor onset, two could be classified as cognitive and two as behaviour arrest. On the other hand, simple partial seizures followed by an impairment of consciousness turned to focal impaired awareness non-motor onset seizures in eight out of nine cases. Among complex partial seizures without accurate data about timing of consciousness impairment, only two out of 15 cases remained without motor/non motor subtype classification.

The use of the term "focal to bilateral tonic-clonic" (2017) for secondary generalized seizures (1981) solved the need to add a new type of seizure in patients with focal epilepsy. Therefore, from 43 seizures classified as GTCS under the 1981 classification, eight were reclassified as focal to bilateral tonic-clonic seizures (19%) (*table 5*).

A significant number of generalized seizures, including 12.5% of the episodes previously classified as absences and 35% of former generalized tonic-clonic seizures, were reclassified as unknown-onset seizures (*table 5*). In one case, typical absences with a myoclonic component were reclassified as myoclonic absences.

Classification of epilepsies (*figure 1*)

Most symptomatic focal epilepsies (92%) were reclassified as structural focal epilepsies. Cryptogenic focal epilepsies were renamed as focal epilepsies of unknown aetiology (93%) or reclassified as epilepsies of unknown type (7%). For those previously categorized as generalized idiopathic epilepsies, 27% were considered to correspond to the group of epilepsies of unknown type. A case in which a concomitant diagnosis of two types of epilepsy was established (focal and generalized, thus included in the former "undetermined epilepsy" category) was included in the new category of "combined epilepsy".

Discussion

The aim of this study was to compare old and new classifications, recognised as tools for clinical data synthesis, to simplify medical records as well as facilitate research and communication. Therefore, the analysis

Table 4. Reclassification of complex partial seizures (1981) in the 2017 ILAE classification.

1981 SEIZURE CLASSIFICATION		**2017 SEIZURE CLASSIFICATION**		
		ONSET		**TYPE**
ABSENCE (9)	TYPICAL (8)	GENERALIZED (7)	NON-MOTOR	ABSENCE (6)
				MYOCLONIC ABSENCE (1)
		UNKNOWN (1)		BEHAVIOUR ARREST (1)
	ATYPICAL (1)	GENERALIZED		ATYPICAL ABSENCE (1)
MYOCLONIC (6)		GENERALIZED (6)	MOTOR	MYOCLONIC (6)
TONIC (6)		GENERALIZED (4)		TONIC (6)
		UNKNOWN (2)		
ATONIC (5)		GENERALIZED (5)		ATONIC (5)
INFANTILE SPASMS (1)		GENERALIZED (1)		SPASMS (1)
TONIC-CLONIC (43)		GENERALIZED (20)		TONIC-CLONIC (20)
		FOCAL (8)		TO BILATERAL TONIC-CLONIC (8)
		UNKNOWN (15)		TONIC-CLONIC (15)

was centred on the identification of strengths and limitations in their clinical application, and not on the evaluation of classifications as diagnostic tools that could impact the accuracy of individual clinical diagnoses. This study was focused on the evaluation of information in the outpatient clinic and the findings may not always be applicable to other settings. Including only patients who had EEG and neuroimaging may have helped to move patients from the "unknown onset" category to either "focal" or "generalized onset" category. Therefore, if we had included patients without such paraclinical studies, there would have probably been more unknown cases. However, since our goal was not to estimate the prevalence of each seizure type but rather to compare both classification systems, which were applied in similar conditions, we did not consider that this selection bias would have any significant impact on our core results. Although the number of included individuals is limited (100 patients; 213 clinical descriptions of seizures), we believe it is sufficient to encompass most categories of both classifications, enabling the analysis of correlations between them. In fact, a saturation effect regarding not only epilepsy, but also seizure types, was identified by the researchers even before achieving the initial goal of 100 individuals. The inclusion of nine researchers with different levels of expertise (from senior epileptologists to neurology residents) could be considered a weakness of this study, introducing significant heterogeneity. However, since the classifications are supposed to be widely used and applied by the medical community, this diversity makes our results more applicable to real clinical practice. In any case, we highlight that an ICC of 87% was found for the new classification of both seizures and epilepsies. The inter-rater agreement is considered good to very good (in the upper 75-90% range) for the application of the new classification, and excellent (more than 90%) using the previous classification (Koo and Li, 2016). It should be noted that junior raters exhibited a significantly lower ICC for the new

Table 5. Correlative distribution of generalized seizures according to the ILAE classifications of 1981 and 2017.

SIMPLE PARTIAL SEIZURE WITH PSYCHIC SYMPTOMS (1981)	**FOCAL AWARE WITH NON-MOTOR ONSET (2017)**
PSYCHIC (3)	COGNITIVE (9)
DYSPHASIC (3)	
DYSMNESIC (2)	
STRUCTURED HALLUCINATIONS (1)	
AFFECTIVE (3)	EMOTIONAL (3)

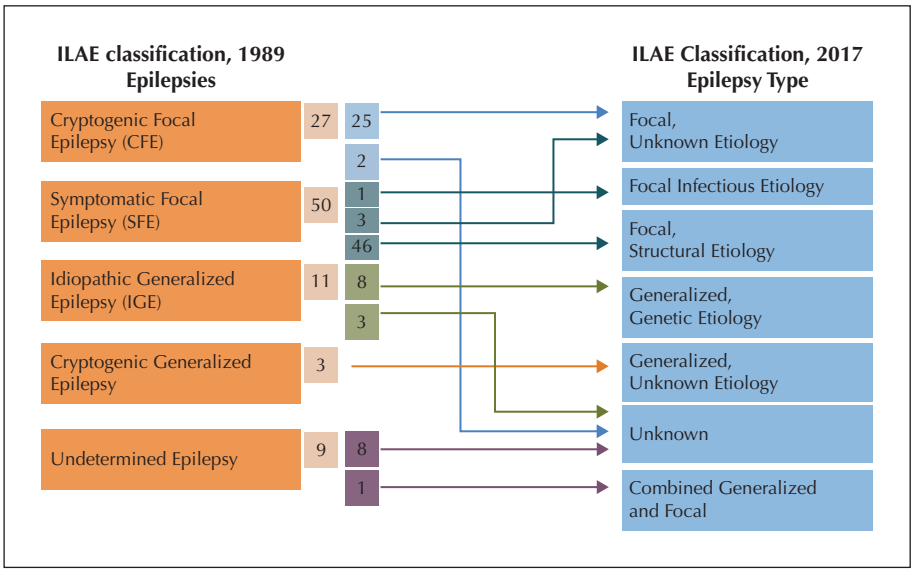

Figure 1. Differential distribution of patients assigned to different categories in the 1989 and 2017 ILAE classifications of epilepsies.

seizure classification, supporting a role of epilepsy training for a more accurate use of the classification. This can be viewed as a limitation in terms of reliability potentially achieved by general physicians but, on the other hand, is an opportunity to obtain excellent rates through training, thus leading to a significant increase in the reliability of this system after years of use.

A key feature of the 2017 classification is the importance given to the onset of seizures, including the category of "unknown onset", not considered in the 1981 classification. Seizures formerly included in the category "unclassified seizure" under the 1981 classification are now discriminated between the groups "unknown onset" and "unclassified" seizures.

Regarding epileptic seizures, it is noteworthy that no seizure previously classified as focal is now classified as unknown onset. In both focal aware motor and non-motor-onset seizures, it was easy to classify the ictal events according to clinical data, although descriptors were frequently needed, particularly in the case of focal non-motor, sensory, and emotional seizures.

Concerning seizures previously classified as generalized, absence seizures, there was one single case without conclusive clinical and EEG features, which was reclassified as an unknown-onset non-motor behaviour arrest seizure under the new classification. This category for uncertainty among these seizure types allows for a conservative approach, thus avoiding clinically significant diagnostic and therapeutic errors. Moreover, it may help to avoid the excessive use of the term "absence", as it exists both in the medical and lay community. On this basis, it is not surprising that idiopathic generalized epilepsies (ultimately registered as "probable" in medical records) have been one of the main sources of the unknown type epilepsy group.

About one in five formerly classified GTCS was reclassified as focal onset and one third as unknown onset. Seizures now classified as unknown-onset tonic-clonic were previously GTCS occurring as the only seizure type and often exclusively during sleep, raising suspicion of an underlying focal epilepsy. This is the semiological substrate for the reclassification of a considerable group of cryptogenic focal epilepsies into epilepsies of unknown type. It is remarkable that by just applying new terminology, a diagnostic uncertainty may be highlighted which may therefore encourage further investigation and follow-up.

About half of the seizures classified as GTCS in 1981 remained in this category under the 2017 classification, mostly based on the presence of either generalized EEG patterns, bilateral myoclonus or typical absence seizures, a strong family history of generalized epilepsy, and/or seizures upon awakening. Allowing the inclusion of all clinical data and ancillary tests in the 2017 classification of seizures would limit the potential overuse of the category "unknown-onset tonic-clonic seizures", supporting the allocation as truly GTCS.

The differentiation of true generalized seizures from those of unknown onset may also be of use for patients' phenotyping, contributing to a more appropriate selection of candidates for both clinical studies and pharmacological trials.

While in a recent publication (Gao *et al.*, 2018), the authors highlight the relevance of the new seizure classification in reducing the number of previously unclassified cases, we consider that one of

the most relevant aspects of the 2017 classification is a section allocated to uncertainty under the term "unknown", applicable to both seizures and epilepsies, and regarding onset, type, or aetiology. Recognition of uncertainty avoids forced categorization and reinforces the consideration of further ancillary studies and/or periodic re-evaluation. Moreover, we emphasize that this may lead to more accurate identification of the diagnostic stage in which uncertainty lies.

Another important aspect of the 2017 classification is the relevance given to the lesional basis and the new approach that contributes to topographical analysis. The new scheme also involves ancillary studies, which confers greater certainty when classifying seizures in patients with focal epilepsies. The reclassification of complex partial seizures to focal seizures with impaired awareness of motor or non-motor onset allows the inclusion of features of topographic value, however, the chronological sequence of disturbance of consciousness is overlooked. At the same time, we consider that the "focal non-motor-onset sensorial" category in the new seizure classification is rather broad, and consistently requires the use of descriptors of the type of sensation to which it refers, in correlation with recognized anatomical landmarks. In any case, additional semiological data beyond seizure classification has always been necessary in order not to overlook information of localizing value, which is particularly critical for epilepsy surgery evaluation. Concerning the classification of epilepsies, most symptomatic focal epilepsies were reclassified as structural focal epilepsies. While the 1989 classification highlighted the cause of focal epilepsy, in 2017, relevance was given to the presence of an identifiable lesion based on neuroimaging studies regardless of its aetiology, which has implications for epilepsy surgery and other therapeutic approaches, including neurostimulation.

Finally, since this study shows that the correlation between the old and new terminology is not "one-to-one", it would be advisable to maintain the original nomenclature when referring to a prior study. □

Supplementary data.
Summary didactic slides are available on the www.epilepticdisorders.com website.

Disclosures.
None of the authors have any conflict of interest to declare.

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TEST YOURSELF

(1) What is gained and what is missing in the reclassification of a seizure described as a forced deviation of the head to the left without disturbance of consciousness, according to the 2017 versus 1981 seizure classification system?

(2) Why do the authors state that the application of the 2017 classification would lead to better management regarding diagnostic uncertainty?

(3) What is the main consequence of the addition of "structural" as an option among the aetiological categories offered in the 2017 classification of epilepsies?

Note: Reading the manuscript provides an answer to all questions. Correct answers may be accessed on the website, www.epilepticdisorders.com, under the section "The EpiCentre".