Updated classification of epileptic seizures: Position paper of the International League Against Epilepsy


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Abstract

The International League Against Epilepsy (ILAE) has updated the operational classification of epileptic seizures, building upon the framework established in 2017. This revision, informed by the implementation experience, involved a working group appointed by the ILAE Executive Committee. Comprising 37 members from all ILAE regions, the group utilized a modified Delphi process, requiring a consensus threshold of over two-thirds for any proposal. The updated classification maintains four main seizure classes: Focal, Generalized, Unknown (whether focal or generalized) and Unclassified. Taxonomic rules distinguish between classifiers, directly impacting clinical management, and descriptors, indicating other clinical seizure characteristics. Focal seizures and those of unknown origin are further classified according to the patient’s state of consciousness during the seizure: impaired, preserved, or unknown state of consciousness, which is defined operationally by clinical assessment of awareness and responsiveness. Generalized seizures are grouped into absence seizures and generalized motor seizures, now including recognition of negative myoclonus as a seizure type. Seizures are described in the basic version as with or without observable manifestations, while an expanded version utilizes the chronological sequence of seizure semiology. This updated classification comprises a total of 20 seizure types. Special emphasis was placed on ensuring translatability into languages beyond English. Its aim is to establish a common language for all healthcare professionals involved in epilepsy care, from resource-limited areas to highly specialized centers, and to provide accessible terms for patients and caregivers.

Key Points

- The ILAE has updated the operational classification of epileptic seizures.
- Adjustments were based on experience with the clinical implementation of the classification established in 2017.
- The four main classes are: Focal, Generalized, Unknown whether focal or generalized and Unclassified.
- Consciousness is a classifier, and it is operationally defined by awareness and responsiveness.
- Seizures are described as with or without observable manifestations (basic) or by the chronological sequence of semiology (expanded).
Introduction

The ILAE operational classification of seizure types was published in 2017\(^1\). The paper concluded with a statement suggesting that the application of this classification in the field for a few years will prompt minor revisions and clarifications. The ILAE actively promoted the implementation of the 2017 classification, sparking intense debates within the international epilepsy community\(^2\)-\(^6\).

In 2023, the ILAE's Executive Committee appointed a working group assigned to assessing the real-world application of the 2017 seizure classification and recommending adjustments while preserving the fundamental framework of the 2017 classification. The basic organization of the 2017 classification evolved from the original 1981 version\(^7\) through subsequent modifications. The primary objective remains the establishment of a common language and standardized definitions for clinical practice. Emphasizing flexibility, the classification aims to accommodate diverse clinical settings, including resource-limited areas and highly specialized centers. Simultaneously, it seeks to offer a clear and robust structure for implementation in research databases and clinical trials.

This seizure classification does not encompass neonatal seizures, which are addressed in a separate position paper\(^8\). Additionally, a new definition of acute symptomatic seizures\(^9\) and the nosology of status epilepticus\(^10\) have been allocated to other working groups. Notably, this classification specifically encompasses clinical seizures, omitting those events solely identified by electrographic activity.

The working group, appointed by the ILAE's Executive Committee, comprised a diverse and inclusive international representation. The methodology employed three successive steps: firstly, the identification of strengths and weaknesses within the 2017 classification; secondly, proposing adjustments and updates; and finally, engaging in an iterative Delphi process to attain a broad consensus. The updated version will be made available on the ILAE website for a two-month period to receive public comments, subsequently undergoing successive revisions. The final version will be submitted for approval from the ILAE's Executive Committee.

Methods

The working group

In January 2023, the Executive Committee appointed a working group comprising 37 experts, with a balanced representation of 19 women and 18 men. The group intentionally mirrored the diverse composition of the ILAE, incorporating members from all ILAE Regions: North America (7), Latin America (5), Europe (11), Eastern Mediterranean (2), Asia Oceania (9), and Africa (3). Within the group, 23 experts specialized in adult epileptology, while 13 primarily focused on pediatric epileptology. Additionally, one member brought expertise as a neurosurgeon. To ensure continuity, four members were selected from the task force involved in developing the 2017 version.

The working group conducted three workshop meetings: two were conducted entirely online in April and May 2023, while one meeting adopted a hybrid approach, combining face-to-face and online elements, held in September 2023 in Dublin, Ireland. Communication primarily occurred electronically, utilizing emails and an online work management platform (Monday). Video recordings and comprehensive documentation of the entire process were electronically archived and made accessible to all members throughout the duration of the process. The ILAE office provided technical assistance with the process.

Systematic review
We conducted a systematic review to evaluate the strengths and weaknesses of the 2017 ILAE seizure classification. We searched PubMed and Embase databases for articles addressing the implementation of the 2017 ILAE seizure classification. Eligibility criteria were: 1) research papers investigating applicability and feasibility of the 2017 seizure classification; 2) review and opinion papers. For the first criterion, we included congress abstracts too, if they provided sufficient details for evaluation. For the second criterion, we excluded congress abstracts and reviews by the authors of the 2017 classification.

Supplementary Document 1 displays the PRISMA flow diagram depicting the review process. Two authors (SB and ET) independently reviewed and rated the records, resolving any disagreements through consensus discussions. Subsequently, the working group further reviewed and edited the outcomes. The review encompassed a total of 41 articles, as detailed in Supplementary Document 2. Among these, 22 research articles evaluated the applicability and feasibility of the 2017 classification: nine studies supported its feasibility, 11 studies found it partially feasible, and two studies deemed it unfeasible. Additionally, 19 articles comprised reviews and opinions: 10 papers expressed negative critiques, six held neutral positions with an optimistic outlook for future implementations, while three presented mixed opinions—supportive and critical.

Strengths and weaknesses

We clustered strengths and weaknesses extracted from the systematic review (Supplementary Document 2) alongside additional input provided by the working group members.

Overall, the 2017 seizure classification's strengths lie in its operational approach and basic organization of seizure types, divided into four main classes. It offers flexibility for classification at varying levels of complexity, making it more practical for real-world clinical use. The addition of the "unknown" class was perceived as an improvement, enhancing the feasibility and applicability of the classification system. There were differing opinions on the introduction of the term "focal to bilateral tonic-clonic seizure." However, a prospective study demonstrated that this term facilitated more accurate classification of seizures compared to its synonym in the older version (1981) of the classification system. The inclusion of more descriptors was seen as a strength, particularly for implementation in databases. A study validated the usefulness of distinguishing focal epileptic spasms from generalized ones.

A robust debate has occurred regarding the suitability of the term "awareness" to describe seizure semiology, rather than using the term consciousness. Several papers pointed out the disadvantages of using awareness as a surrogate marker for consciousness. Impaired consciousness is a term broadly used in clinical neurology, and it is operationally defined by awareness and responsiveness. For general neurologists, epileptic seizure is a differential diagnosis within conditions of transient loss or impairment of consciousness. For medical students and similarly to lay persons, consciousness is simply explained as the ability to respond and to remember. The debate against using responsiveness as a classification criterion revolves around its dependence on intact motor functions and its difficulty in outpatient settings, although studies indicate that impaired responsiveness is often reported during patient history-taking. In epilepsy monitoring units, responsiveness is frequently evaluated over awareness. Some clinicians have adopted the term "impaired awareness" to denote impaired responsiveness, believing it aligns with ILAE position paper, despite this interpretation being incorrect. In children under 4-5 years, assessing awareness is often challenging or impossible, whereas responsiveness can be evaluated using age-appropriate methods. A crucial consideration lies in the translatability of these terms: "awareness" faces challenges in translation across languages like Spanish, French, Portuguese, and
German, while "consciousness" is more translatable and already a universally accepted medical term.

The clinical relevance of categorizing seizures into 'motor vs. nonmotor' and utilizing the first observed phenomenon as a classifier have been questioned. In contexts such as clinical trials or resource-limited settings, a more practical dichotomy, “with vs. without observable manifestations”, has been considered more beneficial. Notably, nonmotor seizures may exhibit observable manifestations such as aphasia or flushing. The use of the first semiology phenomenon as a classifier has shown limited clinical relevance. It does not influence critical factors such as the selection of antiseizure medication, prognosis, or the localization of seizures for surgical therapy. A more clinically relevant approach for characterizing the epileptic network, especially in the context of presurgical evaluation and clinic-anatomic correlation, involves describing the seizure evolution, specifically, the chronological sequence of semiology phenomena.

The 2017 classification categorized absences as nonmotor seizures, which is misleading. Typical absence seizures often present observable motor phenomena such as discrete automatisms, head retropropulsion, and eye blinks, while atypical absences may involve atonic phenomena. Notably, motor manifestations are characteristic features of specific seizure types, such as eyelid myoclonia with absence and myoclonic absences.

Epileptic negative myoclonus is a well-documented phenomenon acknowledged in both the earlier and the revised version of the ILAE semiology-glossary. It is important to note that epileptic negative myoclonus differs from asterixis found in toxic-metabolic encephalopathies. While discussed in prior works, negative myoclonus was not included in the 2017 classification. Experimental studies in animal models and humans demonstrated the focal onset in generalized seizures, and this has been incorporated in the current ILAE definitions. The term “generalized onset seizure” seems to be in contradiction with this, and it may be misleading in the clinical practice, since focal onset of generalized seizures was well-documented in large survey studies and video-EEG recordings.

Epileptic seizures can be classified using various principles, potentially resulting in numerous seizure types, some of which might be redundant and lack clinical relevance. Establishing clear taxonomic rules is essential to precisely define and differentiate classifiers (used to identify seizure types) from descriptors (used to characterize specific features within a seizure type).

Proposals for adjustments

Building upon the strengths and weaknesses discussed and clustered in the previous section, we formulated proposals for adjustments.

1. **Taxonomic rules**: Seizure types directly impact patient management, influencing therapeutic decisions, prognosis, or imply a syndrome diagnosis (*utilitarian justification*). Seizure types to the best of the current knowledge represent biological classes (*conceptual justification*). Meanwhile, descriptors represent key seizure characteristics that, along with other clinical data and modalities, indirectly contribute to patient management.

2. **Terminology of the main seizure classes**: change “generalized onset seizure” to “generalized seizure”, change “focal onset seizure” to “focal seizure”, and change “unknown onset seizure” to “unknown whether focal or generalized”.

3. **Sub-classifier (1)**: Substitute awareness (aware or impaired awareness) with consciousness (preserved or impaired), operationally defined based on awareness and responsiveness.
4. **Sub-classifier (2):** Replace the motor versus nonmotor sub-classification within focal seizures and within seizures unknown whether focal or generalized, with a sub-classification distinguishing between seizures with observable manifestations and those without. Describe seizure semiology in chronological sequence, depicting the sequence of seizure phenomena.

5. **Negative myoclonus:** Include the recognition of negative myoclonus within the seizure classification.

6. **Generalized seizures:** Remove “nonmotor” when categorizing absence seizures.

7. **Epileptic spasms:** Incorporate epileptic spasm as a semiology descriptor for focal seizures and for seizures unknown whether focal or generalized. Retain epileptic spasms as a seizure type for generalized seizures.

**Delphi method**

We employed a modified Delphi method\(^4\)\(^9\) to achieve consensus regarding the proposed adjustments and the update of the seizure classification. For a proposal to pass, it required at least a 2/3 majority vote from the group. Acting as moderators, two authors (SB and ET) facilitated the process. They gathered and summarized the votes, incorporating comments, and returned them for the subsequent round, refraining from voting themselves. Throughout the process, thirty-five members of the working group participated in voting. Individual responses were anonymized to other participants, but after each round, they received a summary of results, along with incoming comments and suggestions.

Consensus was achieved after seven rounds. The first three Delphi rounds focused on addressing the proposals, while the subsequent four rounds were dedicated to the entire updated classification system. All implemented proposals garnered more than 2/3 of the votes, and the final version received unanimous approval from all members of the working group.

**Results**

The fundamental framework for classifying epileptic seizures is maintained\(^1\)\(^-\)\(^7\). The main seizure classes include **Focal, Generalized, Unknown whether focal or generalized, and Unclassified**. Figures 1 and 2 illustrate the basic and expanded seizure classifications, and Table 1 presents the taxonomic hierarchy of seizure classification. **Classifiers** define the seizure types, with direct influence on patient management by guiding syndrome diagnosis, therapeutic decisions and prognosis. **Descriptors**, on the other hand, are significant seizure characteristics that, along with other clinical data and modalities, indirectly contribute to shaping patient management.

Focal seizures are defined as originating within networks limited to one hemisphere\(^1\)\(^-\)\(^4\). They may be discretely localized or more widely distributed, may originate in cortical or subcortical structures. For each seizure type, ictal onset is consistent from one seizure to another, with preferential propagation patterns that may involve the contralateral hemisphere. In some cases, however, there is more than one network, and more than one seizure type, but each individual seizure type has a consistent site of onset\(^4\). Focal-to-bilateral tonic-clonic seizures\(^1\) are focal seizures in which the ictal activity propagates to both hemispheres, while the semiology evolves to impairment of consciousness and bilateral tonic muscle activation, followed by a clonic phase with progressive decrease in frequency, due to a gradual increase in the duration of the silent periods interrupting the tonic muscle activity\(^3\)\(^7\).
Generalized seizures are defined as originating at some point within, and rapidly engaging, bilaterally distributed networks, which can include cortical and subcortical structures, but not the entire cortex\(^1\). Seizure onset can appear localized, and seizures can be asymmetric.

When there is information available to characterize certain aspects of seizures, but it is insufficient for a clear classification as focal or generalized, they are categorized as “Unknown whether focal or generalized”. In cases where there is no available information to characterize the seizure, but the clinician is confident that the event is an epileptic seizure, it is labeled as “Unclassified”.

Subsequently, as more information becomes available to the clinician, these seizures can be reclassified as either focal or generalized.

Figure 1. The basic version of the updated seizure classification

Focal seizures and seizures unknown whether focal or generalized are further classified according to the patient’s state of consciousness during the seizure: impaired, preserved or unknown state of consciousness. Consciousness is operationally defined by establishing awareness and responsiveness, relying on information obtained from medical history\(^2\) or through behavioral testing by medical personnel\(^6\). These operational terms are explained to the patients and caregivers as the ability to remember and to respond appropriately and normally during the seizure. Rather than asking patients and caregivers about consciousness, it is advisable to ask specifically about recall of the events (awareness) and degree of responsiveness during the seizure. Please note that an inadequate response or a significantly longer response latency compared to the interictal (baseline) state qualifies as impaired responsiveness\(^26,37\). Patients and caregivers may need to be reminded that consciousness can still be impaired though the eyes are open, and the patient attempts to interact. In real-world scenarios, information may be available only about one of these characteristics (awareness or responsiveness). If either is impaired in any way, the seizure is classified as impaired consciousness. It is important to exercise caution and consider isolated epileptic amnesia as a potential cause for the lack of recall of ictal experiences, and to rule out ictal paresis or ictal receptive aphasia as potential causes of unresponsiveness, whenever possible.
Figure 2. The expanded version of the updated seizure classification

EXPANDED VERSION

**Focal**

- Consciousness:
  - Preserved
  - Impaired
  - Unknown state

- With vs. Without observable manifestations

- Semiology descriptors in chronological sequence (including epileptic spasms)

- Focal to bilateral tonic-clonic seizure

**Generalized**

- Typical absence
- Atypical absence
- Myoclonic absence
- Eyelid myoclonia with / without absence
- Myoclonic
- Clonic
- Negative myoclonic
- Epileptic spasms
- Tonic
- Atonic
- Myoclonic-atonic
- Tonic-clonic
  - Tonic-clonic
  - Myoclonic-tonic-clonic
  - Absence-to-tonic-clonic

**Unknown whether focal or generalized**

- Consciousness:
  - Preserved
  - Impaired
  - Unknown state

- With vs. Without observable manifestations

- Semiology descriptors in chronological sequence (including epileptic spasms)

- Tonic-clonic seizure unknown whether focal or generalized

**Unclassified**

1. Operationally defined by awareness and responsiveness.
2. Observable manifestations are readily recognized by an eyewitness. These may be motor, aphasic, autonomic or other (see Table 2). Impaired consciousness qualifies as an observable manifestation.
3. Described using the terms in the ILAE semiology glossary incl. observable and not observable semiological features (see table).
4. These phenomena may occur also in focal seizures (usually unilaterally or asymmetrically) as part of the semiology of a focal seizure.

Descriptors can be employed to provide additional characterization of seizures. In the basic version, a straightforward dichotomy is utilized: seizures are described as either having observable manifestations or not. Observable manifestations are easily identified by eyewitnesses\(^\text{2}\), are non-volitional and can include motor, aphasic, autonomic, or other features (see Table 2). Impaired consciousness is considered an observable manifestation. In the expanded version, seizures are described in detail, by listing in chronological order the semiology features (see Table 2) that occur during the seizure\(^\text{37,56}\). The sequence is indicated by arrows pointing in the direction of seizure evolution (for example: epigastric aura → right hand automatism → impaired responsiveness + impaired awareness). All items in the table outlining the semiology features (Table 2) are defined and their significance is explained in detail in the ILAE glossary of seizure semiology\(^\text{37}\). Additionally, video examples are available for each item\(^\text{37}\). The ictal evolution offers crucial insights, as it can identify specific conditions, such as epilepsy of infancy with migrating focal seizures\(^\text{54}\), and aid in the localization of the cortical areas generating the seizures\(^\text{37}\).

**Table 1. The taxonomic hierarchy of epileptic seizure classification.** Classifiers are shown in black, while descriptors are in blue color. Main classes are indicated in bold font, seizure types are underlined. The
hyphen in the numbering separates classifiers (to the left) from descriptors (to the right); the basic version uses descriptors numbered 1 and 2, while the expanded version uses descriptors numbered 3.

1. **Focal**
   1.1. **Focal Preserved Consciousness seizure (FPC)**
   1.1. – 1. With observable manifestations
   1.1. – 2. Without observable manifestations
   1.1. – 3. Semiology descriptors in chronological sequence: Semiology (glossary***) + Somatotopic modifiers
   1.2. **Focal Impaired Consciousness seizure (FIC)**
   1.2. – 1. With additional* observable manifestations
   1.2. – 2. Without additional* observable manifestations
   1.2. – 3. Semiology descriptors in chronological sequence: Semiology (glossary***) + Somatotopic modifiers
   1.3. **Focal Unknown State of Consciousness seizure (FUSC)**
   1.3. – 1. With observable manifestations
   1.3. – 2. Without observable manifestations
   1.3. – 3. Semiology descriptors in chronological sequence: Semiology (glossary***) + Somatotopic modifiers
   1.4. **Focal-to-bilateral tonic-clonic seizure (FBTC)**
   1.4. – 3. Semiology descriptors in chronological sequence: Semiology (glossary***) + Somatotopic modifiers

2. **Generalized**
   2.1. **Absence seizures**
   2.1.1. **Typical absence seizure (TA)**
   2.1.2. **Atypical absence seizure (AA)**
   2.1.3. **Myoclonic absence seizure (MA)**
   2.1.4. **Eyelid myoclonia with / without absence (EMA)**
   2.2. **Generalized motor seizures**
   2.2.1. **Generalized motor seizures – other than tonic-clonic**
   2.2.1.1. **Generalized myoclonic seizure (GM)**
   2.2.1.2. **Generalized clonic seizure (GC)**
   2.2.1.3. **Generalized negative myoclonic seizure (GNM)**
   2.2.1.4. **Generalized epileptic spasm (GES)**
   2.2.1.5. **Generalized tonic seizure (GT)**
   2.2.1.6. **Generalized atonic seizure (GA)**
   2.2.1.7. **Generalized myoclonic-atonic seizure (GMA)**
   2.2.2. **Generalized tonic-clonic seizure (GTC)**
   2.2.2.1. **Generalized tonic-clonic seizure**
   2.2.2.2. **Myoclonic tonic-clonic seizure**
   2.2.2.3. **Absence-to-tonic-clonic seizure**

3. **Unknown whether focal or generalized**
   3.1. **Unknown whether focal or generalized - Preserved Consciousness seizure (UPC)**
   3.1. – 1. With observable manifestations
   3.1. – 2. Without observable manifestations
   3.1. – 3. Semiology descriptors in chronological sequence: Semiology (glossary***) + Somatotopic modifiers
   3.2. **Unknown whether focal or generalized - Impaired Consciousness seizure (UIC)**
   3.2. – 1. With additional* observable manifestations
   3.2. – 2. Without additional* observable manifestations
   3.2. – 3. Semiology descriptors in chronological sequence: Semiology (glossary***) + Somatotopic modifiers
   3.3. **Unknown whether focal or generalized – Unknown State of Consciousness seizure (UUSC)**
   3.3. – 1. With observable manifestations
   3.3. – 2. Without observable manifestations
   3.3. – 3. Semiology descriptors in chronological sequence: Semiology (glossary***) + Somatotopic modifiers
   3.4. **Unknown whether focal or generalized - tonic-clonic seizure (UTC)**
   3.4. – 3. Semiology descriptors in chronological sequence: Semiology (glossary***) + Somatotopic modifiers

4. **Unclassified**
   *In addition to impaired consciousness which is an observable manifestation. **See table with the semiology phenomena.

Table 2. Semiology features
### Somatotopic modifiers
Side (left, right, bilateral-symmetric, bilateral-asymmetric) + Body part

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<tr>
<th>1. Elementary motor phenomena*</th>
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<tbody>
<tr>
<td>Akinetic</td>
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<td>Astatic</td>
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<td>Atonic</td>
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<td>Clonic</td>
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<td>Dystonic</td>
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<td>Epileptic nystagmus</td>
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<td>Epileptic spasm</td>
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<td>Eye blinking</td>
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<td>Eye deviation</td>
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<td>Gyratory</td>
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<td>Head orientation</td>
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<td>Ictal paresis</td>
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<td>Myoclonic</td>
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<td>Myoclonic-atonic</td>
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<td>Negative myoclonus</td>
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<td>Tonic (focal tonic, chapeau de gendarme, fencing posture)</td>
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<td>Tonic-clonic (Figure-of-four)</td>
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<td>Versive</td>
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<tr>
<th>2. Complex motor phenomena*</th>
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<td>Automatisms</td>
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<td>- Gestural automatisms-distal</td>
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<td>- Gestural automatisms-genital</td>
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<td>- Gestural automatisms-proximal</td>
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<td>- Ictal grasping</td>
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<td>- Mimic automatisms (Gelastic, dacrystic)</td>
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<td>- Oro-alimentary automatisms</td>
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<td>- Verbal automatisms</td>
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<td>- Vocal automatisms</td>
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<td>Hyperkinetic behavior</td>
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<th>3. Sensory phenomena**</th>
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<td>Auditory</td>
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<td>Body-perception illusion</td>
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<td>Depersonalization</td>
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<td>Gustatory</td>
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<td>Olfactory</td>
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<td>Somatosensory</td>
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<td>- painful</td>
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<td>- non-painful</td>
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<td>Vestibular / Dizziness</td>
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<td>Visual</td>
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<th>4. Cognitive &amp; language phenomena*</th>
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<td>Aphasia</td>
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<td>Dysmnesia</td>
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<td>- Amnesia</td>
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<td>- Dérà vu/ jamais vu/ dreamy state/reminiscence</td>
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<td>Forced thinking</td>
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<th>5. Autonomic phenomena*</th>
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<td>- Ictal asystole</td>
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<td>Epigastric</td>
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<td>Gastrointestinal</td>
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<td>- Flatulence</td>
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<td>- Hypersalivation</td>
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<td>- Nausea / Vomiting</td>
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<td>- Sialorrhea</td>
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<td>- Spitting</td>
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<td>Pupillary</td>
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<td>- Miosis</td>
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<td>- Mydriasis</td>
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<td>Respiratory</td>
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<td>- Apnea</td>
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<td>- Choking</td>
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<td>- Hyperventilation</td>
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<td>- Hypoventilation</td>
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<td>Urinary</td>
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<tr>
<td>- Incontinence</td>
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<td>- Urinary urge</td>
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<table>
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<th>6. Affective (emotional) phenomena*</th>
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<tbody>
<tr>
<td>Anger</td>
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<td>Anxiety</td>
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<td>Ecstatic/bliss</td>
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<td>Fear</td>
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<td>Guilt</td>
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<td>Mystic</td>
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<td>Sadness</td>
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<td>Sexual</td>
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<th>7. Indescribable aura**</th>
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### Postictal phenomena
- Autonomic signs
- Blindness (hemianopsia or amaurosis)
- Confusion
- Headache
- Language dysfunction
- Nose-wiping
- Palinacousis
- Paresis (Todd’s paresis)
- Psychiatric signs
- Unresponsiveness

*Observable manifestations; **Not observable manifestations; # Possibly observable manifestations. If phenomena not listed above occur during the seizure, they are added in free text. Awareness and responsiveness define consciousness and hence are classifiers. All items in this table are defined in the glossary of semiology.
In the basic version of seizure classification, generalized seizures are further classified as absences and generalized motor seizures. The latter includes generalized seizures with prominent motor phenomena, and a distinction is made between generalized tonic-clonic seizures and other generalized motor seizures. In the figures illustrating seizure classification and in the taxonomic hierarchy table, tonic-clonic seizures are positioned at the end of each main class: focal-to-bilateral tonic-clonic seizures, generalized tonic-clonic seizures, and tonic-clonic seizures unknown whether focal or generalized. This placement aims to emphasize these seizure types, which carry the highest associated morbidity and mortality\textsuperscript{2}–\textsuperscript{5}. In the expanded seizure classification, all generalized seizure types are listed (Figure 2 and Table 1). Generalized negative myoclonus is now recognized as a distinct seizure type, while the other generalized seizure types remain consistent with the 2017 classification\textsuperscript{1}. It is acknowledged that generalized tonic-clonic seizures may be heralded by myoclonic jerks or an absence seizure, a distinction reflected in the subtypes of this seizure\textsuperscript{4,5,6,7}.

Epileptic spasms can be generalized, focal or unknown whether generalized or focal. Distinguishing between them may pose challenges and often requires a multimodal diagnostic work-up. Within the generalized main class, epileptic spasms are considered a distinct seizure type. In the other two main classes, epileptic spasms are described within the seizure semiology (such as focal epileptic spasm). It is important to note that certain motor phenomena may occur in both generalized seizures (where they define the seizure type) and in focal seizures (usually appearing unilaterally or asymmetrically) as part of the semiology of the focal seizure (Figure 2).

Definitions of all generalized seizure types are provided in Supplementary Document 3.

Epileptic seizures are classified along the taxonomic hierarchy, consisting of main classes, sub-classes and seizure-types (Table 1). It is important to explicitly outline the specific list of seizure types, following the principles illustrated in the figures and detailed in this paper. The purpose of the table is to offer clear guidance for electronic databases. The revised seizure classification now comprises 20 seizure types, grouped into four main classes and two sub-classes, representing a simplification compared to the 2017 edition, which included 63 seizure types\textsuperscript{5,8}. The updated classification retains the flexibility of the 2017 edition. The classification of an individual seizure can halt at any level on the hierarchical tree, and seizures initially labeled as unknown or unclassified can be later reclassified, as new information about the seizure becomes available.

While the revised seizure classification places significant emphasis on seizure semiology and can be applied in resource-limited settings, similar to the 2017 edition, it remains interpretative. This allows for the incorporation of supplementary data to identify the seizure types\textsuperscript{1}. In alignment with clinical practice, it is recommended to classify seizures by considering all available information, encompassing semiology and supportive data such as EEG, neuroimaging, laboratory results, and genetics.

In the following section, we illustrate the implementation of the updated seizure classification, utilizing examples from the previous edition and from the articles which criticized it\textsuperscript{4,5,9–60}.

A young woman awakens to find her 20-year old boyfriend having a seizure in bed. The onset is not witnessed, but she is able to describe bilateral stiffening followed by bilateral “shaking”. EEG and magnetic resonance imaging (MRI) findings are normal. This seizure is classified as Tonic-clonic seizure - unknown whether focal or generalized (UTC; 3.4.).

In an alternate scenario of the previous case, the EEG shows a clear right parietal slow-wave focus. The MRI shows a right parietal region of cortical dysplasia. In this circumstance, the seizure is classified as Focal-to-bilateral tonic–clonic seizure (FBTC; 1.4).
A 25-year-old woman describes seizures beginning with 30 s of an intense feeling that “familiar music is playing.” She can hear other people talking, but afterwards realizes that she could not determine what they were saying. Eyewitnesses report that the patient does not respond to external stimuli during the seizure – neither verbal nor tactile (touching the patient). After an episode, she is mildly confused, and has to “reorient herself.” The seizure is classified as Focal Impaired Consciousness seizure (FIC; 1.2. – 1) with additional observable manifestations: auditory aura → receptive aphasia → impaired responsiveness → postictal confusion.

A 22-year-old man has seizures during which he remains fully aware, with the “hair on my arms standing on edge” and a feeling of being flushed. These are classified as Focal Preserved Consciousness seizure (FPC; 1.1. – 1) with observable manifestations: piloerection + flushing.

A 13-year-old with juvenile myoclonic epilepsy has seizures beginning with a few jerks, followed by stiffening of all limbs and then rhythmic jerking of all limbs. These are classified as Generalized myoclonic-tonic-clonic seizures (GTC; 2.2.2.2.)

A 3-month-old boy has clusters of short seizures with flexion in the neck and hips, and abduction in the shoulders of short duration (up to 2 s). The patient has 3-15 clusters per day. The child was encephalopathic, without developmental progression. Seizures were resistant to multiple antiseizure medications, including ACTH. Repeated MRI was unrevealing. Video-EEG showed epileptic spasms associated with a generalized burst suppression pattern on EEG. The seizure is classified as Generalized epileptic spasm (GES; 2.2.1.4).

A 14-month-old girl has sudden extension of both arms and flexion of the trunk for about 2 s. These seizures repeat in clusters. EEG shows hypsarrhythmia with bilateral spikes, most prominent over the left parietal region. MRI shows left parietal cortical dysplasia. Because of the ancillary information, the seizure is classified as Focal Unknown State of Consciousness seizure (FUSC; 1.3. – 1) with observable manifestations: epileptic spasms (brief version: Focal seizure with epileptic spasms).

During long-term video-EEG monitoring, a 28-year-old female patient experiences an ascending sensation from the stomach and then starts chewing and manipulating nearby objects using the right hand. The patient can recall what happens during these episodes and is able to respond. The seizure is classified as Focal Preserved Consciousness seizure (FPC; 1.1) with observable manifestations: epigastric aura → oroalimentary automatisms + gestural automatisms with the right hand + preserved awareness and responsiveness.

The patient reports episodes starting with seeing colored dots and stripes on the left side. The patient cannot recall what happened after that, but eyewitnesses report that the patient does not respond to verbal and tactile stimuli, turns the head to the left, becomes stiff and then has jerks in all limbs. The seizure is classified as Focal-to-bilateral tonic-clonic seizure (FBTC; 1.4) with: elementary visual aura on the left side → versive to left + loss of awareness & responsiveness → bilateral tonic-clonic.

A 33-year-old, right-handed man experienced febrile seizures in infancy. Habitual, unprovoked seizures started at the age of 15 years and were accompanied by a feeling of abdominal discomfort followed by loss of awareness. His wife reported that about once a month he displays episodes of lip smacking, fumbling hand movements and occasional right-hand posturing. The seizure is classified as Focal Impaired Consciousness seizure (FIC; 1.2 - 1) with additional observable manifestations: epigastric aura → impaired awareness → oroalimentary automatisms + gestural automatisms + dystonic posturing in the right hand.
Discussion

The revised seizure classification adheres to the same framework as the 2017 version, maintaining the four main classes. In addition to the archetypical classes of focal and generalized seizures, two more main classes have been introduced for practical reasons: “unknown” (for cases where the distinction cannot be made) and “unclassified” (a temporary class, when no further information is available about the seizure). The impetus for the update arose from the collective experiences after applying the 2017 seizure classification and an iterative discourse of the international epilepsy community. The 2017 version was anticipated to require adjustments based on the insights gained during its implementation in clinical practice.

The working group employed a robust, yet conservative methodology, based on a systematic analysis of the strengths and weaknesses of the 2017 version. Proposals for updates were only considered if they addressed a problem documented in the literature. Approval of any proposal required more than two-thirds of the votes in the Delphi process. The large working group represented the diversity of the ILAE, encompassing broad representation from all regions and various sub-specialties allowing for a broad discussion on the ontological relativity of the terms used in the 2017 classification and widely varying conceptual schemes in different languages. Much like the 2017 edition, the primary objective was to establish a common language and framework for clinical practice. With a focus on flexibility, the classification aims to accommodate diverse settings, ranging from resource-limited areas to highly specialized centers. Simultaneously, it strives to offer a well-defined and clear structure, suitable for implementation in research databases and clinical trials.

Special emphasis was placed on ensuring the coherence and internal consistency of the classification. Following traditional principles employed in scientific classification systems, we established clear taxonomic rules derived from clinical and conceptual reasoning. Features directly impacting patient management were designated as classifiers, while other seizure characteristics served as descriptors. These were organized within the taxonomic hierarchy, resulting in four main classes, two sub-classes (within generalized seizures), and a total of 20 seizure types. The descriptors were structured into two layers: in the basic version, based on the dichotomy of observable ictal manifestations or the lack thereof, and in the expanded version, organized according to the chronological sequence of seizure semiology. The numbering in the taxonomic hierarchy list is designed to ensure consistency across databases and languages, mitigating any potential ambiguity.

To keep the classification system as simple as possible, we refrained from introducing neologisms. Instead, we utilized established medical terminology commonly found in literature and ensured translatability into languages beyond English. The classification has been translated into ten languages, providing a broad, global coverage (Supplementary document 4). We aimed to create a system that is easily communicable to both patients and caregivers.

Table 3. Changes in seizure classification from 2017 to 2024

| 1. | “Onset” is removed from the names of the main seizure classes. |
| 2. | A distinction is made between classifiers and descriptors, based on taxonomic rule. |
| 3. | Consciousness is used as a classifier instead of awareness, with consciousness operationally defined by awareness and responsiveness. |
| 4. | The motor vs. non-motor dichotomy is replaced by observable vs. non-observable manifestations. |
5. The chronological sequence of seizure semiology is used to describe seizures, rather than relying solely on the first sign.

6. Negative myoclonus is recognized as a seizure type.

The changes included in the updated seizure classification are summarized in Table 3. The term "onset" has been omitted from the names of the main seizure classes, as there is compelling evidence suggesting focal onset in generalized seizures as well. The names of these classes now align with their definitions in the ILAE position papers.

Both awareness and responsiveness are used to characterize consciousness, which is now the classifier. The motor vs. non-motor dichotomy was extended to observable vs. non-observable manifestations, which is deemed advantageous for clinical trials. This is now considered a descriptor in the basic version of the seizure classification. In the expanded version, the entire chronological sequence of seizure semiology is utilized for describing the seizure, rather than just the initial sign. This approach was considered more suitable for advanced settings, such as long-term video-EEG monitoring and presurgical evaluation.

The term 'non-motor' has been removed from absence seizures due to the presence of motor phenomena that may be observed during them, some of which are characteristic of certain types of absence seizures (e.g., myoclonic absence, eyelid myoclonia with absence). Negative myoclonus is now recognized as a seizure type. Within generalized seizures, epileptic spasm is considered a seizure type, while within focal seizures and seizures of unknown origin, epileptic spasm is described as part of the seizure semiology (e.g., focal epileptic spasm). Similarly, motor phenomena defining generalized seizure types (myoclonic, tonic, atonic) may also be part of the semiology of a focal seizure.

The updated classification maintains the continuity with the 2017 edition, so that seizures already classified with the previous version can easily be converted. For example, impaired awareness translates to impaired consciousness, a motor seizure is an observable manifestation.

These adjustments of the updated seizure classification were based on experience with the application of the 2017 version. They are relatively minor modifications that preserve the fundamental framework of seizure classification. The aim is to enhance broad clinical applicability across diverse settings.
Conflicts of interest:

The authors do not report conflicts of interest directly related to this paper.

BJ serves as Associate Editor of the Journal Neurology. She receives research support from NIH, CDC and Neuropace, Inc.

DC received educational grant from UCB, Astra Zeneca, Desitin and is a member of the advisory board of Astra Zeneca and UCB. Dr Craiu is a Chair of the Education and Training Committee and of the Guidelines Committee of the EPNS (European Pediatric Neurology Society). No COIs related to this article.


JHC has acted as an investigator for studies with GW Pharma/Jazz Pharmaceuticals, Zogenix/UCB, Vitafla, Stoke Therapeutics, Ultrygenyx and Marinus. She has been a speaker and on advisory boards for Jazz Pharmaceuticals, UCB, Biocodex and Nutricia; all remuneration has been paid to her department. She holds an endowed chair at UCL Great Ormond Street Institute of Child Health; she holds grants from NIHR, EPSRC, GOSH Charity, LifeARC and the National Institute of Health Research (NIHR) Biomedical Research Centre at Great Ormond Street Hospital. She is President of the ILAE 2021-2025.

JMW: National (South African) advisory board for Novartis and Sanofi. Associate Editor Epilepsia (honorarium for work covered).

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NS has served on scientific advisory boards for GW Pharma, BioMarin, Arvelle, Marinus and Takeda; has received speaker honoraria from Eisai, Biomain, Livanova, Sanofi; has served as an investigator for Zogenix, Marinus, Biomain, UCB, Roche. He was supported by #NEXTGENERATIONEU (NGEU) and funded by the Ministry of University and Research (MUR), National Recovery and Resilience Plan (NRRP), project MNESYS (PE000006) – A Multiscale integrated approach to the study of the nervous system in health and disease (DN. 1553 11.10.2022). He was supported also by the Italian Ministry of Health with Current Research Funds.

SA is Deputy Editor for Epilepsia; has served as a consultant or received honoraria for lectures from Angelini Pharma, Biocodex, Eisai, Encoded, Jazz Pharmaceutics, Grintherapeutics, Neuraxpharm, Nutricia, Orion, Proveca, Stoke, Takeda, UCB Pharma, and Xenon; and has been an investigator for clinical trials for Eisai, Marinus, UCB Pharma, Proveca, and Takeda

SB serves as Editor-in-Chief of Epileptic Disorders. He received compensation for speaking at CME programs from Lundbeck, Eisai, UCB and GSK. He received research support from: Independent Research Fund Denmark; Innovation Fund Denmark; European Union: Eurostars Programme / EUREKA; European Union: Horizon Europe Framework Programme (HORIZON); Danish Agency for Higher Education and Science: International Network Programme.

SW received educational grants on behalf of his institution from UCB Pharma, Jazz Pharma, Paladin Labs, and served on the advisory board of Paladin Labs.
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