

Special Article

Epileptic Seizures and Epilepsy: Definitions Proposed by the International League Against Epilepsy (ILAE) and the International Bureau for Epilepsy (IBE)

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Summary: The International League Against Epilepsy (ILAE) and the International Bureau for Epilepsy (IBE) have come to consensus definitions for the terms *epileptic seizure* and *epilepsy*. An epileptic seizure is a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain. Epilepsy is a disorder of the brain characterized by an enduring predisposition to generate

epileptic seizures and by the neurobiologic, cognitive, psychological, and social consequences of this condition. The definition of epilepsy requires the occurrence of at least one epileptic seizure. **Key Words:** Epilepsy—Seizure—Definition—Classification—Epidemiology—International League Against Epilepsy.

Epilepsy is the name of a brain disorder characterized predominantly by recurrent and unpredictable interruptions of normal brain function, called epileptic seizures. Epilepsy is not a singular disease entity but a variety of disorders reflecting underlying brain dysfunction that may result from many different causes. Little common agreement exists on the definition of the terms *seizure* and *epilepsy*. Such definitions are important for communication among medical professionals and also for communication others involved in legislation, disability pensions, driving regulations, workplace safety, education, and for many other purposes. The definitions in this article are directed to a diverse group of physicians, educators, researchers, public officials, and people with epilepsy and their families.

The current proposal reflects consensus discussions held by representatives of the International League Against Epilepsy (ILAE) and the International Bureau for

Epilepsy. It presents practical and operational definitions applicable both in medical and nonmedical settings.

Although the definitions have been formulated in English because of the prevalence of English in scientific communications, the intent is to express the essential features and meaning of these terms in a way applicable to translations into all languages. Preliminary definitions of *epileptic seizure* and *epilepsy* can be found in the ILAE 2001 Glossary of Descriptive Terminology for Ictal Semiology (1), and in a commonly cited epidemiologic review from 1991 (2). The current work is an expansion of the two central terms and should be considered to supersede the prior definitions.

ELEMENTS OF A DEFINITION OF SEIZURE

A definition (Table 1) is a useful tool for communication, diagnosis, and differential diagnosis. However, a description of the essential nature or boundaries of a condition may not lead to easily applicable rules in a practical setting. With respect to epileptic seizures, a statement concerning the electrical activity of the brain during

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TABLE 1. *Definitions*

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

Epilepsy is a disorder of the brain characterized by an enduring predisposition to generate epileptic seizures and by the neurobiologic, cognitive, psychological, and social consequences of this condition. The definition of epilepsy requires the occurrence of at least one epileptic seizure.

the episode appears to be necessary in the definition, but difficult to apply in clinical practice. A definition ideally would include assertions about pathophysiology and the metabolic consequences of the seizure, but the understanding of pathophysiology changes with each research advance.

As a word, *seizure* derives from the Greek meaning *to take hold*. Modern popular terminology uses the word *seizure* for any sudden and severe event (for example, “he had a heart seizure”). Many physical or psychological sudden events, some of them not even pathological, all resemble epileptic seizures in some ways. To emphasize this usage, typically, we will refer to an *epileptic seizure*.

Different definitions emphasize different features of an epileptic seizure (Table 2): nature of onset and termination, clinical manifestations, and enhanced neuronal synchrony. Some previous definitions also considered issues of etiology, classification, and diagnosis, although none of these strictly fits within the task of a definition, and these aspects are not reviewed here.

Mode of onset and termination

An epileptic seizure is “transient,” demarcated in time, with a clear start and finish. Termination of an epileptic seizure often is less evident than is the onset, because symptoms of the postictal state can blur the end of the seizure. Status epilepticus is a special circumstance with prolonged or recurrent seizures; however, clinical and EEG analysis usually allows diagnosis of status epilepticus. The start and finish of an epileptic seizure can be determined on behavioral or EEG grounds, but we must recognize that these two operational criteria do not always coincide.

Clinical manifestations

An epileptic seizure is a clinical event; therefore signs and symptoms must feature prominently in the definition. Detailed specification of subjective and objective clinical phenomena during an epileptic seizure is difficult, because

of the wide range of possible manifestations. Seizure presentation depends on location of onset in the brain, patterns of propagation, maturity of the brain, confounding disease processes, sleep–wake cycle, medications, and a variety of other factors. Seizures can affect sensory, motor, and autonomic function; consciousness; emotional state; memory; cognition; or behavior. Not all seizures affect all of these factors, but all influence at least one. In this context, sensory manifestations are taken to include somatosensory, auditory, visual, olfactory, gustatory, and vestibular senses, and also more complex internal sensations consisting of complex perceptual distortions. In previous definitions, these complex internal sensations were referred to as “psychic” manifestations of seizures.

According to the 2001 ILAE Glossary of Descriptive Terminology for Ictal Semiology (1), cognitive deficits during seizures can appear as problems with perception, attention, emotion, memory, execution, praxis, or speech. Memory distortions can be either negative or positive, in the sense of interruption of memory formation or retrieval as a negative symptom, or intrusion of inappropriate memories as a positive symptom. Positive memory symptoms give rise to *déjà vu* and other forced memories during seizures. Some of the distorted memories previously were classified as psychic symptoms, which is a potentially ambiguous term. Emotional state is difficult to specify but must be considered in the definition, because some seizures manifest as fear, elation, satisfaction, anxiety, or other subjective sensations that cannot be ascribed to the primary senses.

EEG patterns that resemble those seen during some epileptic seizures, but are perceived neither by the patient nor by the observer (sometimes inappropriately called “subclinical seizures”), are not defined as epileptic seizures. However, the clinician should be aware that that more careful observation or interaction may reveal previously missed subtle behavioral correlates of the EEG changes.

Abnormal enhanced synchrony

Hughlings Jackson in 1870 provided a now classic definition of an epileptic seizure as a “symptom . . . an occasional, an excessive and a disorderly discharge of nerve tissue.” This is the most difficult feature of the definition to apply in practice, because the electrical discharge is visible only under some circumstances of testing. Patients with recurrent seizures may have a normal scalp EEG between seizures and even during seizures. Nevertheless, the definition assumes that such an abnormal electrical discharge could be ascertained under ideal circumstances. Inclusion of the requirement for an uncontrolled electrical discharge as a feature of the definition of an epileptic seizure is an issue separate from that of how best to determine whether such an electrical discharge is or is not present. Without the electrical discharge criteria, many other clinical events

TABLE 2. *Elements of a definition of epileptic seizure*

Mode of onset and termination
Clinical manifestations
Abnormal enhanced synchrony

that are not epileptic seizures would meet the other definition criteria.

By “disorderly,” Jackson probably meant “capable of producing dysfunction,” which is certainly accurate. However, EEG discharges during epileptic seizures are orderly and relatively stereotyped. Firing of neurons may involve inhibition as well as excitation, so it is not always the case that an epileptic seizure involves an excess of excitation over inhibition. A feature more common to epileptic seizures is abnormal enhanced synchrony of neurons.

Discussion of systems giving rise to epileptic seizures properly falls more within the realm of seizure pathophysiology than within that of seizure definition. Definition of an epileptic seizure becomes operationally difficult without ascribing it to the brain. Trigeminal neuralgia, for example, can result from an abnormal enhanced synchrony of neurons in the trigeminal ganglion or the fifth cranial nerve, but would not be considered an epileptic seizure. Neither would hyperactive spinal reflexes resulting in excessive discharge of anterior horns cells and tonic stiffening of a limb. Cerebral cortex is the primary element in the generation of epileptic seizures, but it is not the only one. In some circumstances, epileptic seizures can originate in thalamocortical interactive systems or in the brainstem. Consideration of this issue is beyond the scope of this proposal for a definition of an epileptic seizure.

DEFINITION OF EPILEPSY

Epilepsy is not one condition, but is a diverse family of disorders, having in common an abnormally increased predisposition to seizures. Some writers prefer the plural term, “the epilepsies,” but we will use the singular phrase while recognizing this diversity.

History of at least one seizure

At least one seizure is required to establish the presence of epilepsy; a predisposition, as determined, for example, by a family history, or by the presence of epileptiform EEG changes, is not sufficient to determine epilepsy. The definition does not include a requirement that the seizure be “unprovoked,” a feature of several prior individual definitions. Instead, the definition requires, in addition to at least one seizure, the presence of an enduring alteration in the brain, as described in the next paragraph.

Enduring alteration in the brain

The central concept in our definition of epilepsy is an enduring alteration in the brain that increases the likeli-

TABLE 3. *Elements of a definition of epilepsy*

History of at least one seizure
Enduring alteration in the brain that increases the likelihood of future seizures
Associated neurobiologic, cognitive, psychological, and social disturbances

hood of future seizures. The diagnosis of epilepsy, under this concept, would not require two seizures; it would require only one epileptic seizure in association with an enduring disturbance of the brain capable of giving rise to other seizures. Multiple epileptic seizures due to multiple different causes in the same patient would not be considered to be epilepsy. A single epileptic seizure due to an enduring epileptogenic abnormality would indicate epilepsy, and a single epileptic seizure in a normal brain would not.

Associated conditions

At times, epilepsy must be defined by more than just the recurrence, or a potential for recurrence, of seizures. For some people with epilepsy, behavioral disturbances, such as interictal and postictal cognitive problems, can be part of the epileptic condition. Patients with epilepsy may suffer from stigma, exclusion, restrictions, overprotection, and isolation, which also become part of the epileptic condition. Seizures and the potential for recurrence of seizures also often have psychological consequences for the patient and for the family.

The elements of the definition of epilepsy are summarized in Table 3.

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