ILAE OFFICIAL REPORT

A practical clinical definition of epilepsy

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Epilepsia, 55(4):475–482, 2014
doi: 10.1111/epi.12550

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Summary

Epilepsy was defined conceptually in 2005 as a disorder of the brain characterized by an enduring predisposition to generate epileptic seizures. This definition is usually practically applied as having two unprovoked seizures >24 h apart. The International League Against Epilepsy (ILAE) accepted recommendations of a task force altering the practical definition for special circumstances that do not meet the two unprovoked seizures criteria. The task force proposed that epilepsy be considered to be a disease of the brain defined by any of the following conditions: (1) At least two unprovoked (or reflex) seizures occurring >24 h apart; (2) one unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years; (3) diagnosis of an epilepsy syndrome. Epilepsy is considered to be resolved for individuals who either had an age-dependent epilepsy syndrome but are now past the applicable age or who have remained seizure-free for the last 10 years and off antiseizure medicines for at least the last 5 years. "Resolved" is not necessarily identical to the conventional view of "remission or "cure." Different practical definitions may be formed and used for various specific purposes. This revised definition of epilepsy brings the term in concordance with common use.

KEY WORDS: **Epilepsy, Seizure, Definition, Unprovoked, Recurrence.**

Accepted January 3, 2014.

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In 2005, a Task Force of the International League Against Epilepsy (ILAE) formulated conceptual definitions of "seizure" and "epilepsy" (Table 1).[1] Conceptual definitions can be translated for specific purposes into operational (practical) definitions.

The ILAE commissioned a Task Force to formulate an operational definition of epilepsy for purposes of clinical diagnosis. This article summarizes the recommendations of the Task Force, including appended notes and case examples explaining the reasons for these recommendations and occasional dissenting views. In December of 2013, the ILAE Executive Committee adopted the recommendations as a position of the ILAE.

Why alter the definition of epilepsy? Doing so might cause confusion among patients who could be left uncertain as to whether they have or do not have epilepsy. Epidemiologists and other researchers would need to decide whether to use the new or old definition and how this might affect trends and comparisons. Rules and regulations might have to be changed. Arrayed against these potential negatives are positive aspects to reevaluation of the definition. The current definition requires two unprovoked seizures occurring at least 24 h apart.[2] Some epileptologists recognize and feel a need to address circumstances with high risk for future seizures after a first unprovoked seizure. For example, one Delphic study group in Spain[3] voted with high consensus in favor of treatment in five of seven hypothetical scenarios after a first seizure. A decision for treatment does not necessarily equate to a diagnosis of epilepsy, but it can be taken as a marker for belief in a strong enduring predisposition for further seizures. Conversely, a diagnosis of epilepsy does not necessarily require treatment. The current definition does not allow a patient to outgrow epilepsy, yet many older individuals have all but forgotten their two childhood seizures. A definition should conform to how clinicians and patients think, and usefully merge with other individual considerations in helping to make treatment decisions.

Practical Clinical Definition of Epilepsy

Conceptually, epilepsy exists after at least one unprovoked seizure, when there is high risk for another, although the actual required risk is subject to debate. After a single unprovoked seizure, risk for another is 40–52%.[4] With two unprovoked nonfebrile seizures, the chance by 4 years of having another is 73%, with a 95% confidence interval (CI) of 59–87%, subsequently herein portrayed as approximately 60–90%.[5]

Table 1. Conceptual definition of seizure and epilepsy – 2005 report

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 "two unprovoked seizure" definition of epilepsy has served us well, but it is inadequate in some clinical circumstances. A patient might present with a single unprovoked seizure after a remote brain insult, such as a stroke, central nervous system (CNS) infection, or trauma. A patient with such brain insults has a risk of a second unprovoked seizure that is comparable to the risk for further seizures after two unprovoked seizures.[6] When two individuals with a history of at least one unprovoked seizure have the same high risk for having another, an argument can be made that both have epilepsy. Under limits of the current definition, another patient might have photosensitive epilepsy, yet not be considered to have epilepsy because the seizures are provoked by lights. Another might be free of seizures and seizure medications for 50 years, yet still have epilepsy. In order to bring the practical (operational) clinical definition of epilepsy into concordance with how epileptologists think about epilepsy, the ILAE Task Force recommends broadening the definition of epilepsy to include the circumstances enumerated in Table 2. The Task Force also added a time limit to the definition.

Several elements of this definition require clarification.

Disease

Epilepsy has traditionally been referred to as a disorder or a family of disorders, rather than a disease, to emphasize that it is comprised of many different diseases and conditions. The term disorder implies a functional disturbance, not necessarily lasting; whereas, the term disease may (but not always) convey a more lasting derangement of normal function. Many heterogeneous health problems, for example, cancer or diabetes, comprise numerous subdisorders and are still considered to be diseases. The term "disorder" is poorly understood by the public and minimizes the serious nature of epilepsy. The ILAE and the International Bureau for Epilepsy (IBE) have recently agreed that epilepsy is best considered to be a disease.

Two unprovoked seizures

Epilepsy exists in a patient who has had a seizure and whose brain, for whatever reason, demonstrates a pathologic and enduring tendency to have recurrent seizures. This tendency can be imagined as a pathologic lowering of the seizure threshold, when compared to persons without the condition. Table 2, item 1, represents the current commonly employed definition of epilepsy as at least two unprovoked seizures occurring >24 h apart. A seizure that is provoked by a transient factor acting on an otherwise normal brain to temporarily lower the seizure threshold does not count toward a diagnosis of epilepsy. The term "provoked sei-

Table 2. Operational (practical) clinical definition of epilepsy

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

1. At least two unprovoked (or reflex) seizures occurring >24 h apart
2. One unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years
3. Diagnosis of an epilepsy syndrome

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

zure" can be considered as being synonymous with a "reactive seizure" or an "acute symptomatic seizure."[7] Etiology should not be confused with provocative factors, as some etiologies will produce an enduring tendency to have seizures. A brain tumor, for example, might cause a person to have an epileptic seizure, but not as a transient insult.

The condition of recurrent reflex seizures, for instance in response to photic stimuli, represents provoked seizures that are defined as epilepsy. Even though the seizures are provoked,[8] the tendency to respond repeatedly to such stimuli with seizures meets the conceptual definition of epilepsy, in that reflex epilepsies are associated with an enduring abnormal predisposition to have such seizures.

A seizure after a concussion, with fever, or in association with alcohol-withdrawal, each would exemplify a provoked seizure that would not lead to a diagnosis of epilepsy. The term "unprovoked" implies absence of a temporary or reversible factor lowering the threshold and producing a seizure at that point in time. Unprovoked is, however, an imprecise term because we can never be sure that there was no provocative factor. Conversely, identification of a provocative factor does not necessarily contradict the presence of an enduring epileptogenic abnormality. In an individual with an enduring predisposition to have seizures, a borderline provocation might trigger a seizure, whereas in a non-predisposed individual, it might not. The Definitions Task Force recognizes the imprecise borders of provoked and unprovoked seizures, but defers discussion to another venue.

High recurrence risk

Table 2, item 2 defines another path for diagnosing epilepsy. Its intent is to encompass circumstances for which some practitioners[9] and expert epileptologists[3] manage patients as if epilepsy is present after a single unprovoked seizure, because of a very high risk of recurrence. Such examples may include patients with a single seizure occurring at least a month after a stroke[6] or a child with a single seizure conjoined with a structural or remote symptomatic etiology and an epileptiform electroencephalography (EEG) study.[10] Another example is a patient in whom diagnosis of a specific epilepsy syndrome associated with persistent threshold alteration can be made after the occurrence of a single seizure. A first seizure might present as status epilepticus,[11,12] but this does not in itself imply epilepsy. Recurrence risks are not known for the majority of individual cases. However, if a treating physician is aware that the lesion has generated an enduring predisposition for unprovoked seizures with a risk comparable to those who have had two unprovoked seizures (which we all agree is epilepsy), then that person too should be considered to have epilepsy. Choosing a specific threshold risk number might be excessively precise, but for general comparison, this risk is about 60–90% after two unprovoked seizures.[I] A threshold level of 60% appropriately exceeds the 50% level of recurrence risk found at 5 years after a single seizure in the United Kingdom multicentre study of early epilepsy and single seizures (MESS) study.[13]

It is important to note that a single seizure plus a lesion or a single seizure plus epileptiform EEG spikes does not automatically satisfy criteria for this operational definition of epilepsy, because data may vary among different studies and specific clinical circumstances. In the Dutch Epilepsy Study,[10] children with epileptiform EEG patterns after their first seizure had a 2-year risk for recurrence of 71%, but in the study by Shinnar et al.,[12] children with a first idiopathic seizure and abnormal EEG patterns had recurrence risk of 56% at 3 years. No formula can be applied for additive risks, since data are lacking on how such risks combine; such cases will have to be decided by individualized considerations. Recurrence risk is a function of time, such that the longer the time since the last seizure, the lower the risk.[14]

The revised definition places *no burden* on the treating physician to specify recurrence risk in a particular circumstance. In the absence of clear information about recurrence risk, or even knowledge of such information, the default definition of epilepsy originates at the second unprovoked seizure. On the other hand, if information is available to indicate that risk for a second seizure exceeds that which is usually considered to be epilepsy (about 60%), then epilepsy can be considered to be present.

Epilepsy syndrome

It makes little sense to say that someone has an epilepsy syndrome[15] but not epilepsy. If evidence exists for an epilepsy syndrome, then epilepsy may be presumed to be present, even if the risk of subsequent seizures is low. This is the case with benign epilepsy with centro-temporal spikes (BECTS).

Exceptional syndromic cases may exist in which obvious behavioral seizures may not occur at all, as can be the case with continuous spike and waves during sleep and the Landau-Kleffner Syndrome.[16]

Implications for treatment

Diagnosing epilepsy after a single unprovoked seizure when there is high risk for recurrence may or may not lead to a decision to initiate treatment. The proposed practical definition may provide support to a physician who wishes to treat a patient with high recurrence risk after a single unprovoked seizure. However, a treatment decision is distinct from a diagnosis, and should be individualized depending upon the desires of the patient, the individual risk-benefit ratio and the available options. The physician should weigh the possible avoidance of a second seizure with associated risks against the risk for drug-related side effects and costs for the patients.

To be clear, the diagnosis of epilepsy and a decision to treat are two related but different issues. Many epileptologists treat for a time after an acute symptomatic seizure (for example, with Herpes encephalitis), with no implication of epilepsy. In contrast, patients with mild seizures, with seizures at very long intervals, or those declining therapy might go untreated even when a diagnosis of epilepsy is beyond dispute.

Unprovoked seizures separated in time

The time span between two unprovoked seizures that together qualify as epilepsy is subject to ambiguity. Seizures clustering within 24 h confer approximately the same risk for later seizures as does a single seizure.[17] The Task Force retained the current thinking that unprovoked seizures clustering in a 24 h period be considered to be a single unprovoked seizure for purposes of predicting recurrence risk.

Some authorities[17] consider epilepsy to be present, but in remission, after 5 years of seizure freedom. However, the definition of epilepsy does not specify an outer time limit for occurrence of the second unprovoked seizure to mark the onset of epilepsy. Therefore, epilepsy could be considered present if an unprovoked seizure occurred at age 1 and at age 80, a condition sometimes referred to as oligoepilepsy.[18] The Task Force acknowledges that, in such circumstances, the causes of the seizures occurring at the two time points might be different, and if so then epilepsy would not be present.[II] Otherwise, the Task Force did not agree on a specific interval of time between seizures that would "reset the clock" for counting an event as a second seizure. A rationale for setting such an interval might emerge from future research.

Epilepsy resolved

Is epilepsy, once diagnosed, always present? The traditional definition does not allow for its disappearance. Should a person who has been seizure-free and off medication for decades after absence seizures as a child still be considered to have epilepsy? Likewise, are patients with mesial temporal lobe epilepsy who have been seizure-free off medications for 10 years after resection of their hippocampal sclerosis considered to still have epilepsy? Seizure freedom for long intervals of time can result from one of several different underlying circumstances and treatments. An abnormal tendency to have unprovoked seizures may remain, but the seizures are successfully controlled by therapy. Children can outgrow their epilepsy, as with BECTS. Some persons might have had a definitive treatment, such as brain surgery, rendering them permanently seizure-free.

The Task Force sought a definition that would allow a possible end to the burden of having epilepsy. Medical literature uses the term "remission" to imply an abeyance of a disease, but this term is not well-understood by the public, and remission does not convey absence of the disease. "Cure" implies a risk for future seizures no greater than that of the baseline unaffected population, but after a history of epilepsy such a low risk is never achieved. The Task Force therefore adopted the phrase "resolved."[III] When epilepsy is resolved, it implies that the person no longer has epilepsy, although it does not guarantee that it will not return.

What time intervals and circumstances should characterize resolved epilepsy?[IV] Recurrence risk depends on the type of epilepsy, age, syndrome, etiology, treatment, and many other factors. Juvenile myoclonic epilepsy is known to be subject to an elevated risk of seizures for several decades,[19] but remissions do still occur. Structural brain lesions, such as malformations of cortical development,[20] may elevate risk of seizures long term. Seizures may recur at variable intervals after remission due to removal of an epileptogenic lesion, such as a cavernous malformation.[21] A study[22] of 347 children achieving at least 5-year "complete remission" including at least 5-years free of antiseizure drugs identified late seizure relapses in 6%. One occurred as long as 8 years after the prior seizure. Data were not given for those remaining free of seizures after a 10-year complete remission, but the number would be <6%. After temporal lobe epilepsy surgery,[23] 54.2% of patients relapse within 6 months; whereas, only 1.9% relapse 4 years after surgery. Similar results were seen in another study,[24] with only 0.6% having seizures in the last year of follow-up, provided that they had been seizure-free for 3 years after surgery.

The risk of seizure recurrence after unprovoked seizures diminishes with time, although the risk may never reach levels for normal individuals who have not had a prior seizure. Most relapses are early. After a single unprovoked seizure, 80%[14,17] to 90%[25] of those who had a second did so within 2 years. In one study,[5] after a second unprovoked seizure, subsequent seizures occurred within 4 years, but none in the ensuing 3 years, suggesting that the risk may not be zero but is low. The National General Practice Study of Epilepsy in the United Kingdom[14] identified a 3-year recurrence risk of 44% after a seizure-free period of 6 months, 32% after 12 months,

and 17% after 18 months. No adequate data are available on seizure recurrence risk after being seizure-free and off medication for extended periods of time. Delayed relapses are rare after 5 years.[26] By 10 years off antiseizure medicines, the annual risk for seizures probably is very low.[27,V]

Clinicians will have to individualize a determination of whether epilepsy is resolved. The Task Force chose to define epilepsy as being resolved for individuals who had an age-dependent epilepsy syndrome but are now past the applicable age or those who have remained seizure-free for the last 10 years, with no seizure medicines for the last 5 years. Delineation of circumstances in which epilepsy is definitively cured is beyond the scope of this paper.

Imperfect information

From the clinician's perspective, the new practical definition linking epilepsy to a predefined probability of seizure recurrence brings greater clarity and clinical relevance to the diagnostic process. However, optimal application of this definition often requires specialized diagnostic and interpretative skills—specifically, in assessing recurrence risks, or in diagnosing syndromes—which may not be broadly available in all settings, particularly at the primary care level. Even more important is the inevitable uncertainty in many situations about the potential epileptogenicity of an magnetic resonance imaging (MRI)–demonstrated lesion. For instance, one or more brain cysts in an individual with neurocysticercosis[28] may be incidental findings with no epileptogenic activity in a particular individual. Risk does not equate with causation. When in doubt, practitioners should consider referring a patient to a specialized epilepsy center with experience in diagnosis.

In the absence of a seizure documented by video-EEG recording and typical for a person's recurrent unprovoked seizures, there will be situations where a diagnosis of epilepsy remains uncertain. One approach to these ambiguities would be to define a condition called "probable (or possible) epilepsy."[VI] Such an approach has been adopted with other diseases, such as multiple sclerosis with the McDonald criteria,[29] amyotrophic lateral sclerosis with the El Escorial criteria,[30] migraine,[31] and vascular dementia.[32] The ILAE Task Force recognized the subtle, but important, difference between telling a patient that "you have probable epilepsy" versus "you probably have epilepsy." In the absence of secure information, the latter statement, or another statement simply expressing uncertainty, seemed a more straightforward assertion. Therefore, the Task Force has not defined probable epilepsy as a specific entity, but has left that possibility open for the future.

Consequences of the Practical Definition

Definitions have consequences. From the viewpoint of the patient, epilepsy is associated with stigma and psychological, social, cognitive, and economic repercussions so important as to be built into the conceptual definition of epilepsy.[1] The new practical definition could improve outcomes by sensitizing clinicians about the need to give greater consideration to the risk of recurrence after a single unprovoked seizure, and making the clinicians more comfortable in initiating treatment after some initial unprovoked seizures. This must be individualized, since a diagnosis of epilepsy does not necessarily require prescription of an antiseizure drug, and treatment might be justified in some patients for whom a definitive diagnosis of epilepsy has not been made. A practical definition allowing earlier diagnosis will be especially useful for prevention of unnecessary risks of physical injuries or social consequences resulting from recurrent seizures in patients deemed to be susceptible to a high risk for recurrence. The revised definition also provides an expanded opportunity for disease-modifying interventions that prevent the progression of epilepsy and onset of comorbidities.

How revision of the definition of epilepsy will affect the measured prevalence of epilepsy is unpredictable. Future epidemiologic studies may choose to use the older operational definition for consistency. If the revised definition is used, some patients previously considered to have epilepsy will no longer carry an epilepsy diagnosis because of the provisions for epilepsy being resolved. Other individuals who meet the "single seizure with high risk for another" criteria might be added to the epilepsy group.

The definition of epilepsy will affect diagnosis and treatment in both resource-rich and resource-poor societies. The Task Force has been careful to define epilepsy in a way that can be applied in general with or without expensive technology that may not be universally available.

The correct diagnosis of epilepsy in people who might not have been diagnosed previously may have both negative and positive consequences. For example, economic consequences might include reimbursement by a national health service for medications whose cost otherwise would have to be covered by the affected person. On the other hand, many people with epilepsy have difficulty in obtaining life or medical insurance. Some cannot purchase a first home without a life insurance policy secured at the time of home purchase. Stigma could profoundly affect some people not previously considered to have epilepsy, with serious and misguided consequences such as loss of access to education or marriage bans. Allowing epilepsy to be declared "resolved" may lift the stigma from some who should no longer be considered to have epilepsy. Positive economic and health consequences will accrue when more accurate diagnosis results in appropriate preventative treatment before a second seizure occurs.

People with reflex epilepsies previously have been disenfranchised by the requirement that seizures be unprovoked. The inclusion of reflex epilepsy syndromes in a practical clinical definition of epilepsy now brings these individuals into the epilepsy community.

The revised practical definition described in this report is intended for clinical diagnosis, and might not be suitable for all research studies. Different operational definitions will be used depending on specific purposes, and comparisons could still be made using the traditional "two-unprovoked-seizure" definition of epilepsy whenever appropriate. Investigators must clearly identify the definition used in any study or publication.

A revised definition has implications for legislation and health economics. Regulations affecting individual life activities, such as driving restrictions, relate more to seizure frequency or to risk of seizure recurrence than to a diagnosis of epilepsy, but this is not always the case. In some countries a diagnosis of epilepsy per se limits the period of validity of a driving permit, or the type of permit that can be acquired. Guidelines about participation in certain sports may stipulate restrictions for people with a diagnosis of epilepsy, irrespective of seizure history. Insurance coverage and social benefits might also be affected by the diagnostic label. To the extent that a revised practical definition might affect the number of people diagnosed with epilepsy, there could be cost repercussions for the individual and for the society. Costs to society may not necessarily be higher, however, particularly if the new operational diagnosis codifies the current approach of epileptologists and leads to improved management of individuals who are likely or unlikely to have future seizures.

Conclusion

Epilepsy previously has been defined as at least two unprovoked seizures >24 h apart. The revised practical definition implies that epilepsy also can be considered to be present after one unprovoked seizure in individuals who have other factors that are associated with a high likelihood of a persistently lowered seizure threshold and therefore a high recurrence risk. Such risk should be equivalent to the recurrence risk of a third seizure in those with two unprovoked seizures, approximately at least 60%. The latter risk level occurs with remote structural lesions, such as stroke, CNS infection, certain types of traumatic brain injury, diagnosis of a specific epilepsy syndrome, or in some circumstances with the presence of other risk factors. Those with recurrent reflex seizures, for example, photosensitive seizures, are also considered to have epilepsy. This definition of epilepsy brings the term in concordance with common use by most epileptologists.[VII] Epilepsy is not necessarily life-long, and is considered to be resolved if a person has been seizure-free for the last 10 years, with at least the last 5 year off antiseizure medicines, or when that person has passed the age of an age-dependent epilepsy syndrome. The new definition is more complicated than is the old definition. Studies providing detailed knowledge of seizure recurrence risk are few, so most diagnoses of epilepsy will of necessity still be made by documentation of two unprovoked seizures. As more knowledge of recurrence risks is accrued for specific etiologies, application of the epilepsy definitions will become more precise and more useful.

Case Examples[VIII]

1. *Two seizures*. A 25-year-old woman has two unprovoked seizures, 1 year apart. *Comment*: This person has epilepsy, according to both the old and new definitions.
2. *Stroke and seizure*. A 65-year-old man had a left middle cerebral artery stroke 6 weeks ago and now presented with an unprovoked seizure. *Comment*: With a seizure in this time relation to a stroke (or brain infection or brain trauma) the literature[6] suggests a high (>70%) risk of another unprovoked seizure. Therefore, in the new (but not the old) definition, this man would have epilepsy.
3. *Photic seizures.* A 6-year-old boy has had two seizures 3 days apart while playing a videogame involving flashing lights. There have been no other seizures. EEG shows an abnormal photoparoxysmal response. *Comment*: This boy has epilepsy according to the new definition (but not the old), even though the seizures are provoked by lights, since there is an abnormal enduring predisposition to have seizures with light flashes.
4. *Benign Epilepsy with Centrotemporal Spikes (BECTS).* A 22-year-old man had seizures with face twitching when falling asleep at ages 9, 10, and 14 years; he has had none since. EEG at age 9 years demonstrated centrotemporal spikes. Medications were discontinued at age 16. *Comment*: For this young man, epilepsy is resolved, because of passing the relevant age range of an age-dependent syndrome. The old definition has no provision for considering epilepsy to be resolved.
5. *Single seizure and dysplasia*. A 40-year-old man had a focal seizure characterized by left hand twitching that progressed to a tonic–clonic seizure. This was his only seizure. Magnetic resonance imaging (MRI) shows a probable transmantle dysplasia in the right frontal lobe and EEG shows right frontotemporal interictal spikes. *Comment*: Although many clinicians would reasonably treat this man with antiseizure medications, the recurrence risk for seizures is not precisely known, and therefore epilepsy cannot yet be said to be present according to either definition. Future epidemiologic studies might clarify this situation.
6. *Two seizures long ago*. An 85-year-old man had a focal seizure at age 6 and another at age 8 years. EEG, MRI, blood tests, and family history were all unrevealing. He received antiseizure drugs from age 8 to age 10 years, when they were discontinued. There have been no further seizures. *Comment*: According to the new definition, epilepsy is resolved, since he has been seizure-free for >10 years and off seizure medication for at least the last 5 years. This is not a guarantee against future

seizures, but he has a right to be viewed as someone who does not currently have epilepsy.

7. *Long-interval seizures.* A 70-year-old woman had unprovoked seizures at ages 15 and 70. EEG, MRI, and family history are unremarkable. *Comment*: Both old and new definitions consider this woman to have epilepsy. Despite the diagnosis, many clinicians would not treat because of the low frequency of seizures. Should investigations somehow show that the causes of the two seizures were different, then epilepsy would not be considered to be present.
8. *Questionable information*. A 20-year-old man has had three unobserved episodes over 6 months consisting of sudden fear, difficulty talking, and a need to walk around. He is not aware of any memory loss during the episodes. There are no other symptoms. He has no risk factors for epilepsy and no prior known seizures. Routine EEG and MRI are normal. *Comment*: Declaring this man to have epilepsy is impossible by either the old or new definition. Focal seizures are on the differential diagnosis of his episodes, but both definitions of epilepsy require confidence that the person has had at least one seizure, rather than one of the imitators of seizures. Future discussions may define the boundaries of "possible and probable epilepsy."

Acknowledgments

The Task Force would like to thank a group appointed by the ILAE to review revision of the article specifically in response to the public comments. This group consisted of Lars Forsgren Umeå University Hospital Sweden; Angelina Kakoozaa, Makerere University College of Health Sciences, Kampala, Uganda; and Akio Ikeda, University of Kyoto.

Disclosure or Conflict of Interest

Robert S. Fisher has received support from, and/or has served as a paid consultant for, the Maslah Saul MD Chair, the Anderson fund for Epilepsy Research, The Susan Horngren Fund, SmartMonitor, and ICVRx, and has done consulting for Cyberonix, Oracle, and UCB. Alexis Arzimanoglou has received support from, and/or has served as a paid consultant for, Cyberonics, Eisai, GlaxoSmithKline, UCB Pharma, and Viropharma. J. Helen Cross has received support from, and/or has served as a paid consultant for, Eisai, Viropharma, and GlaxoSmithKline. Christian E. Elger has received support from, and/or has served as a paid consultant for, Bial, Eisai, Novartis, Desitin, and UCB. He also received support from the DFG (Deutsche Forschungsgemeinschaft). Jerome Engel, Jr., receives support from the Jonathan Sinay Chair. Lars Forsgren has received support from, and/or has served as a paid consultant for, GSK, UCB, Eisai, and Orion Pharma. Jacqueline A. French has received support from, and/or has served as a paid consultant via the Epilepsy Study Consortium or the HEP project for, Eisai Medical Research, GlaxoSmithKline, Impax, Johnson & Johnson, Mapp Pharmaceuticals, Novartis, Lundbeck, Pfizer, Sepracor, Sunovion, SK Life Science, Supernus Pharmaceuticals, UCB Inc/Schwarz Pharma, Upsher Smith, Vertex, Eisai Medical Research, LCGH, Impax, Mapp Pharmaceuticals, Novartis, UCB, UCB Inc/Schwarz Pharma, Upsher Smith, and Lundbeck. Dale C. Hesdorffer has received support from, and/or has served as a paid consultant for, UCB, Esai, and UpsherSmith. B.-I. Lee has received support from UCB and GlaxoSmithKline and has done consulting for UCB. Solomon L. Moshé has received support from the Charles Frost Chair In Neurosurgery and Neurology, and has served as a paid consultant for Lundbeck and UCB. Emilio Perucca has received support from, and/or has served as a paid consultant for, Bial, Eisai, GlaxoSmithKline, Lundbeck, Medichem, Pfizer, Sun Pharma, Supernus, UCB Pharma, Viropharma, and Vertex. Ingrid E. Scheffer has received support from, and/or has served as a paid consultant for, UCB, Athena Diagnostics, GlaxoSmithKline, and Janssen-Cilag EMEA. Torbjörn Tomson has received support from, and/or has served as a paid consultant for, GlaxoSmithKline, UCB, Eisai, Sun Pharma, and Bial. S. Wiebe has received support from, and/or has served as a paid consultant for, the Hopewell Professorship in Clinical Neurosciences Research, University of Calgary, and ElectroCore. The remaining authors have no potential conflicts of interest to disclose. We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

Notes

I. Specifying a level of risk for recurrence to quantify the concept of "enduring predisposition" was difficult for the Task Force. All agreed that an individual with two unprovoked seizures had epilepsy. The risk for a third seizure in such an individual is about 3 in 4, but the 95% confidence intervals are about 60–90%. Therefore, the Task Force agreed that an individual having a similar risk after one unprovoked seizure should logically be considered also to have epilepsy. The number >60% is intended to be an approximate guideline, rather than a sharp cutoff.

II. Some suggested a time limit within which the two spontaneous seizures must occur to diagnose epilepsy. In the absence of consensus and evidence on which to base a specific time, lifetime occurrence was retained as the default.

III. The motivation for this aspect of the definition was twofold. First, many clinicians, patients, and families consider epilepsy to be in the past when seizures no longer occur and no antiseizure medications are employed. Second, the Task Force desired to remove lasting stigma associated with a lifetime diagnosis of epilepsy. Other terms considered included remission, terminal remission, complete remission, inactive epilepsy, epilepsy absent, epilepsy not present, epilepsy no longer present, and cure. Many of these did not convey the concept that epilepsy was gone. Cure implied complete success of some treatment or passage of time, such that risk was that of the baseline population.

IV. Evidence to guide a specific required seizure-free number of years is limited, and existing risk functions show a continuous decline over time, rather than a natural breakpoint. Some argued for 5 years, but as many as 5% annually may have a seizure after a 5-year seizure-free interval. Being seizure-free for the most recent 10 years and off medications for the most recent 5 years predicts future freedom from seizures in a high percentage of cases.

V. Although evidence exists for a (low) relapse rate after 5 years of seizure freedom, no evidence was available at time of writing for relapse rates after being seizure-free for 10 years, which therefore was selected to be a time longer than 5 years, for which relapse rate would be consider likely very low.

VI. Whether to define a condition called "probable epilepsy," "possible epilepsy," or both, generated the most debate in the deliberations, and ultimately the issue was settled by majority view rather than full consensus. Probable epilepsy was considered for two different circumstances: the first in which one seizure had occurred and risks were high but not very high for having another. The second circumstance encompassed limited information in cases that seemed to be epilepsy, but reliable seizure descriptions or other key data were lacking. Allowing a diagnosis of probable epilepsy in the second circumstance could harmfully short-cut necessary diagnostics to clarify the diagnosis. The Task Force did see value in defining probable epilepsy, but believed that extensive future consideration would be needed in order to make its definition operationally consistent and useful.

VII. An earlier draft of the manuscript was posted for comments on the ILAE website. A total of 315 comments, some very extensive, were received. The majority of opinions were positive, but there also were some very thoughtful and strongly felt disagreements. It was considered

unreasonable to place a burden on a treating physician for knowing the precise risk for a subsequent seizure. The authors agreed with this criticism. Many commenters were for and many others against calling epilepsy a disease, rather than a disorder. This decision was commandeered by the respective IBE and ILAE Executive Committees in favor of the term "disease." The phrase "no longer present" was not embraced by those responding to comments, and it was changed to "resolved." Many commenters wished for epilepsy to be resolved at 5 years of seizure freedom, on or off antiseizure drugs. The Task Force wanted resolved to mean a risk sufficiently low that epilepsy could be put aside, and achieving that requires a more stringent time interval, so we settled on 10 years of seizure freedom, 5 years off medicines. Several commenters wanted to eliminate the slippery concept of provoked versus unprovoked seizures. Such a change would have been quite fundamental, altering our view of acute symptomatic seizures, now comprising 40% of all seizures. We left that discussion for another venue. In general, the authors believed that the "wisdom of the crowd" strengthened and clarified the arguments and, more importantly, moved the definition closer to how working clinicians think of epilepsy.

VIII. These examples were presented on June 24, 2013, to the audience of the ILAE Congress Presidential symposium, with >1,000 epileptologists in attendance. Audience votes on whether epilepsy was present in these cases correlated very strongly with the terms of the revised definition. Although not a scientifically valid survey, the responses indicated that epileptologists thought of epilepsy in ways consistent with the revised definition.

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