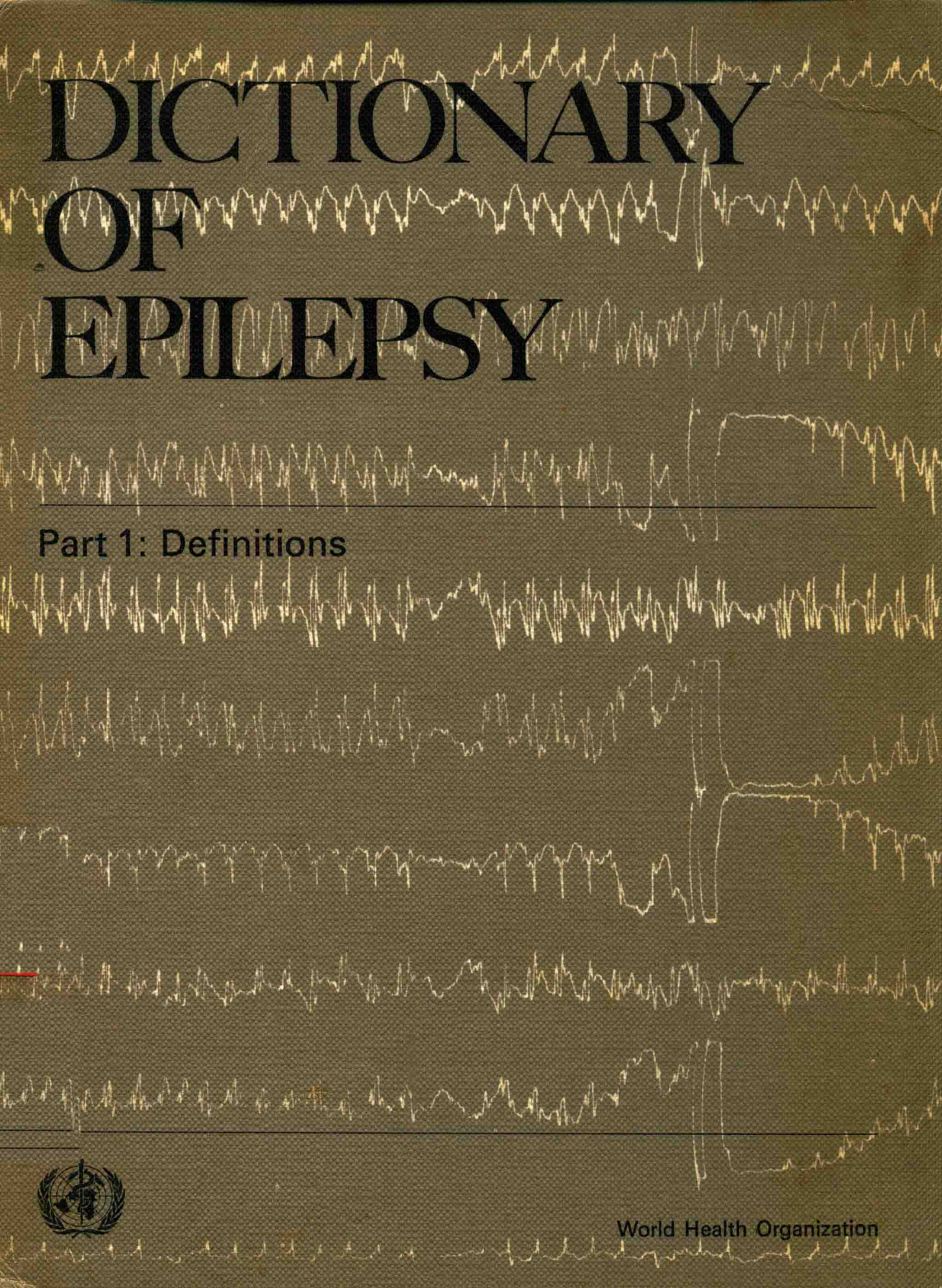
DICTIONARY OF EPILEPSY

Part 1: Definitions



World Health Organization

DICTIONARY OF EPILEPSY

PART I: DEFINITIONS

PROFESSOR H. GASTAUT
President, University of Aix-Marseilles, France

in collaboration with
an international group of experts

WORLD HEALTH ORGANIZATION
GENEVA
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INTRODUCTION

Although considerable advances have been made in recent years in understanding the etiology and pathogenesis of disorders of the nervous system, there are many areas in which knowledge is still lacking. Further progress requires critical re-examination of the available data and comparative research on a large scale, and here problems of terminology are of great significance. In the past almost every school of neurology or of psychiatry produced its own definitions, which were accepted by some and rejected by others. This has led to the same definition being used for different concepts, or to the same concept being defined in different ways.

The situation has deteriorated with the growth of published information ; terms used are frequently not defined and may have different meanings for author and reader. With present-day acceptance of the desirability of epidemiological investigations of neurological and mental disorders, the need for accuracy and comparability in reporting the primary data is therefore becoming increasingly urgent.

Although dictionaries already exist in some fields of neurology and psychiatry, as a rule they tend to reflect the personal ideas of the authors and ignore the terminology in use elsewhere by other specialists. An international group of experts convened in Paris by the Council for International Organizations of Medical Sciences (15-17 November 1965) stressed the urgent need for action to reduce the growing confusion in medical and scientific terminology. It was suggested that general agreement on terminology could be achieved through the efforts of international organizations because they can best assemble and coordinate the efforts of experts in any specific field. The present publication is WHO's response to this suggestion. Out of the vast field of neurology and psychiatry, epilepsy was chosen for the following reasons :

1. Recent studies have shown that epilepsy is of unexpectedly high frequency and severity in many parts of the world. In developing countries, where medical care has been insufficient, the total prevalence may reach 8 per 1000 of the population, and frequent severe seizures have been reported in 3 per 1000. Epilepsy presents an important mental health and public health problem, not only because of its serious economic implications but also through its social impact on the family and the community. Furthermore, severe epilepsy causes serious disability and has a high mortality. For various reasons, including superstition, many patients are not brought for treatment, although with modern methods the disease can probably be controlled in 75 % of patients. An unnecessary burden of disability therefore exists.

2. Epilepsy is a problem encountered not only by neurologists and psychiatrists but also by general practitioners and public health personnel.

3. Significant advances have been made in recent years in disciplines particularly relevant to research in epilepsy, notably in the fields of neurophysiology and electroencephalography, but it is impossible to take full advantage of these advances until terminology has become more standardized. At the same time, the new knowledge that has accrued makes it easier to agree on definitions.

The first draft of the dictionary was prepared in French by Professor H. Gastaut and was sent to a panel of specialists from 16 countries (see page 5). Their comments and suggestions were discussed at a WHO meeting held in Geneva in February 1967.[1] *On the basis of the agreed amendments, a second draft was prepared and circulated for further comments. These were discussed at a second WHO meeting held in Geneva in July 1968,*[2] *after which a further revision was undertaken. From the beginning, the preparation of this* Dictionary of Epilepsy *has been actively supported by the International League Against Epilepsy. The present version thus represents the consensus of experts in the field of epilepsy from many parts of the world.*

The dictionary covers mainly terms pertaining to the clinical aspects of epilepsy. Terms used in neurophysiology and electroencephalography and clinical terms for conditions related to epilepsy, such as hysteria and narcolepsy, have not been included, with the exception of a few terms considered important for the understanding of certain clinical aspects of epilepsy, e.g., neuronal discharge, electroencephalographic paroxysm, and convulsive syncope.

The terms epilepsy and epileptic seizure occur frequently in the text. Epilepsies are conditions of diverse etiology characterized by recurrent seizures caused by varied mechanisms, and a given condition may often be described as either epilepsy or an epileptic seizure. Although the distinction is sometimes arbitrary, the two terms are used in the following ways:

epilepsy *when referring to the etiology or the site of the lesion, e.g., genetic epilepsy, temporal lobe epilepsy;*

epileptic seizure *when referring to the nature of the phenomenon or the frequency or circumstances of occurrence, e.g., oropharyngeal epileptic seizure, automatic epileptic seizure, hallucinatory epileptic seizure, and evoked epileptic seizure.*

Each definition is followed by synonyms and by additional linguistic or explanatory notes where necessary. A special effort has been made to include all known synonyms of the defined terms.

The dictionary is divided into two parts: Part I contains the definitions discussed above and is being published simultaneously in English, French, and Spanish. It is hoped that a Russian edition will also be available shortly. Part II, which is still in

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[1] Participants: Professor H. Gastaut, Dr R. L. Masland, Professor R. J. Broughton, Professor P. M. Saradžišvili, and members of the WHO Secretariat.

[2] Participants: Professor H. Collomb, Professor H. Gastaut, Dr R. L. Masland, Professor D. A. Pond, Professor P. M. Saradžišvili, Dr M. M. Velasco Suárez, Professor T. Wada, and members of the WHO Secretariat.

preparation, will be a multilingual index to all four versions, giving the equivalent terms in the different languages.

In spite of the collaboration of eminent specialists in its preparation and although it represents the outcome of years of work, the dictionary is still regarded as provisional. Terms that epileptologists would like to see included may have been omitted and some of the definitions may be found not to be entirely satisfactory. Only when the dictionary has been put to practical use in the field will such defects become apparent. Comments and suggestions will be welcome and should be addressed to: Chief, Mental Health, World Health Organization, 1211 Geneva 27, Switzerland.

The World Health Organization expresses its gratitude to all those specialists who have assisted in compiling this work—especially to Professor H. Gastaut—and without whose enthusiasm, energy, and profound knowledge of epilepsy this venture would not have been possible. Special acknowledgement should also be made of the valuable assistance provided by Professor R. J. Broughton in the preparation of the English translation.

NOTES ON THE USE OF THE DICTIONARY

The terms are listed in alphabetical order, the noun taking precedence in compound terms, e.g.,

epilepsy, centrencephalic

If more than one adjective is used, the most specific adjective is placed first, immediately after the noun, e.g.,

seizure, contraversive epileptic
seizure, ecmnesic hallucinatory epileptic

If the term is composed of two or more nouns, with or without adjectives, the noun describing the manifestation takes precedence, e.g.,

hallucination, epileptic mirror

Where a compound term is used in the text of a definition, the usual order of words is employed.

As far as possible, all the synonyms that could be found in the literature have been included in the dictionary, but wherever more than one term has been used for the same concept only a single term is recommended. If a term is followed by a definition, this indicates that it is the recommended term for that concept, unless otherwise specified in the definition. All terms that are not followed by definitions are therefore obsolete, incorrect, or not recommended for some other reason.

If a term used in a definition is printed in italics and followed by an asterisk, this indicates that the user of the dictionary should look up the definition of that term for further information.

Several abbreviations are used in the text; these are:

Adj.	Adjective
Ant.	Antonym
Expl.	Explanatory
Ling.	Linguistic note
Syn.	Synonym

The use of proper names has been avoided as far as possible and there are very few terms where it has seemed necessary to use an author's name for a syndrome, e.g., Jacksonian. They have been used only when intensive usage and practical convenience so dictated; in all fourteen have been retained. In all other instances the reference to a proper name, if there is one, is given only in the text.

A

absence. Very brief clouding or loss of consciousness (lasting usually 2–15 seconds) accompanying certain *generalized epileptic discharges**. When impairment of consciousness is the only detectable clinical sign, the term *simple absence** is used. When other signs are also present, the term used is *complex absence**, several varieties of which can be distinguished. An absence may be accompanied by the following types of electroencephalographic discharge: (1) A discharge of the typical rhythmic 3/sec spike-and-wave type characterizes *typical absence**, easily recognized by its very sudden onset and end and by a slight flickering of the eyelids that may occur synchronously with each spike. (2) *A recruiting epileptic rhythm**, or a pseudorhythmic succession of slow spike-and-wave complexes, characterizes *atypical absence**, which begins and ends less abruptly than the typical absence. The characteristic brief losses of consciousness accompanying certain *partial epileptic seizures** (principally of temporal lobe origin) present clinically as absences. However, for various etiological, therapeutic, and other reasons, they are not usually considered to be absences as such. Nevertheless, some authors continue to refer to them as "absences", qualified by an adjective denoting the region of origin (usually *temporal lobe absences**). This usage is not recommended. Syn.: *absentia epileptica ; epileptic absence ; epileptic lapse* (incorrect and obsolete); *epileptic vertigo* (incorrect and obsolete).

Expl.: The problem of defining the term "absence", or rather "absences", is particularly difficult and is far from having been solved. Neither the Commission on Terminology of the International League against Epilepsy, which drew up the International Classification of Epileptic Seizures, nor the World Health Organization experts who acted as advisers in the preparation of this dictionary, were able to devise a satisfactory definition. The problem should therefore be considered in detail. In French, where the term originated, "absence" meant "absence of mind" and could refer just as well to a fit of absent-mindedness as to a loss of consciousness. Gradually, the term acquired a purely medical connotation and came to be applied exclusively to loss of consciousness of any kind, but mainly to the type characteristic of syncope. In his doctoral thesis Calmeil (1824) introduced the concept of "epileptic absence" to designate the short periods of loss of consciousness that occur in epileptics and that, up to that point, had been mingled with other types of epileptic seizure in the categories of *epileptic equivalents**, *minor epileptic seizures**, *larval epilepsy**, and *petit mal**. Delasiauve, in his *Traité de l'épilepsie* (1854), included absence among the various types of epileptic attack and defined it as a sudden loss of consciousness lasting a few seconds and apparent to the observer only from the patient's vacant expression, facial pallor, and immobility. The introduction of electroencephalography in the United States of America was immediately followed by the discovery of the rhythmic discharge of bilateral, symmetrical, and synchronous 3/sec spike-and-wave complexes that accompany absences, as defined by the French authors of the nineteenth century. Unfortunately, instead of keeping the term "absence" for the overall clinical and EEG picture, American authors employed the term "petit mal", introduced by Esquirol in 1815, for all minor epileptic seizures, including absences. They even used the word "petit", not as an abbreviation for "petit mal" but through the misinterpretation of Tissot (1769), who had referred to "large attacks and small ones [des petits]", the latter undoubtedly referring to absences, with their highly characteristic "slight flickering of the eyes". With the progress of epileptology that followed the introduction of electroencephalography, it became clear that there existed other forms of epileptic attacks with loss of consciousness of brief duration, and this gave rise to terminological difficulties. First of all, it was realized that certain *partial epileptic seizures**, particularly of temporal lobe origin, could take the form of a loss of consciousness a few seconds in duration that was clinically almost indistinguishable from an absence. Some authors, using mainly clinical criteria, proposed logically that these too should be called "absences", qualified as "temporal". However, in view of the clear differences in symptomatology, etiology, pathogenesis, and therapy between temporal lobe seizures and true absences, other authors rejected any common term. Still others proposed the term "pseudo-absences", qualified by the adjective "temporal". It also became clear that some *generalized epileptic seizures** too could be clinically almost identical with absences but without displaying the typical 3/sec spike-and-wave discharge. On etiologic and therapeutic grounds such seizures could be clearly distinguished from absences; but in terms of

symptomatology and pathogenesis the two types of seizure were sufficiently similar to justify bracketing them together. Some authors therefore proposed referring to the ordinary absences accompanied by a 3/sec discharge as *petit mal absences* and using the term *variant of petit mal absence* for the attacks unaccompanied by this discharge. Other authors, however, have strongly opposed these terms and have proposed replacing them by *typical absences** and *atypical absences**. A further complication arose when it became evident that the symptomatology of absences could be either "simple", i.e., when restricted to the impairment of consciousness, or "complex", when accompanied by other signs. This led to the introduction of the terms *simple absence* and *complex absence*, applicable to both typical absences and atypical absences, and to the definition of a wide variety of complex absences in terms of the signs accompanying them (atonic absence, automatic absence, myoclonic absence, etc.). As things now stand, the problem of the terminology of absences has not been satisfactorily solved. It seems difficult, however, to find an ideal solution in view of the impossibility of using clinical or EEG features alone to define complex manifestations that are also characterized by their etiology, pathogenesis, and response to drugs.

absence, atonic. A *complex absence** characterized by a loss of postural tone sufficient to cause the subject to slump to the ground. In a *typical absence**, with a rhythmic 3/sec spike-and-wave discharge, the loss of tone may be rhythmic and produce jerky falling movements synchronous with each spike-and-wave complex. On the other hand, in absences with an atypical EEG (*atypical absences**), the loss of tone is always continuous and may be sufficiently sudden to cause the subject to collapse and injure himself. Even when brief, an atonic absence must be distinguished from an *epileptic drop attack**. Syn.: *cataplectic epileptic seizure* (incorrect and obsolete); *inhibitory epileptic seizure; atonic petit mal.*

absence, atypical. A type of simple or complex *absence** characterized by EEG features that differ from those of *typical absences**. There are at least three varieties of EEG discharge peculiar to atypical absences: (1) a low-voltage *recruiting epileptic rhythm** with a very high frequency of about 20/sec; (2) a higher-voltage recruiting epileptic rhythm with a slower frequency of about 10/sec; (3) bilateral, roughly synchronous and symmetrical spike-and-wave complexes, repeated more or less rhythmically at a slow frequency of about 2/sec. Electroencephalographically, atypical absences are easily distinguished from *typical absences* by their lack of bilateral, synchronous, and symmetrical 3/sec spike-and-wave complexes. Moreover, unlike typical absences, they are very difficult to activate by hyperventilation or intermittent photic stimulation. Finally, atypical absences do not belong to the benign group of *primary generalized epilepsies** but are part of the severe symptom complex that constitutes the *Lennox-Gastaut syndrome**. Syn.: *petit mal variant absence ; petit mal variant* (incorrect).

Ling.: Considered out of context and disregarding some clinical features that are often too minor to be easily assessed, in particular a less abrupt onset and termination, this variety of absence can be distinguished from the *typical absence* (or "petit mal absence") only by electroencephalography. This is why it has also been called "petit mal variant absence", the word "variant" here signifying a mode of expression that deviates from the standard. See *absence* (Expl.).

absence, automatic. A *complex absence**, typical or atypical, characterized by usually simple *epileptic automatisms**, for example, involuntary movements of the lips or tongue, or behaviour such as rubbing the hands together or adjusting the clothes. Such attacks must be differentiated from partial seizures of temporal lobe origin that have similar psychomotor symptoms and that, when brief, are sometimes referred to incorrectly as *temporal lobe absences**.

absence, autonomic. A *complex absence** characterized by marked autonomic phenomena.

absence, complex. An *absence** in which the impairment of consciousness is accompanied by other symptoms, which tend to dominate the clinical picture. See the following types of *absence: atonic; automatic; autonomic; enuretic; hypertonic; myoclonic; retrocursive; retropulsive; tussive*; and *vasomotor*. In exceptional cases, atony or myoclonus accompanying a complex absence may predominate on one side of the body or even be completely unilateral. See *seizure, unilateral epileptic.*

absence, enuretic. A *complex absence** characterized by urinary incontinence. Such absences must be differentiated from partial seizures of temporal lobe

origin, which may be accompanied by urinary incontinence or, more frequently, by automatisms of micturition. See *seizure, enuretic epileptic.*

absence, epileptic. Syn. for *absence.*

absence, hypertonic. A *complex absence** characterized by an increase in postural tone producing backward extension of the head and conjugate upward deviation of the eyes, and sometimes backward extension of the trunk (*retropulsive absence**); the latter may force the subject to walk backwards in order to keep his balance (*retrocursive absence**).

absence, mnesic. An obsolete term describing an *absence** characterized mainly by an arrest of ideation; consciousness is maintained more or less intact and the subject remains capable of memorization and recall. See *absence, subclinical; seizure, mnesic epileptic.*

absence, myoclonic. A *complex absence** characterized by bilateral and rhythmic *myoclonus** predominating in the cephalic region and the upper limbs. The attack is almost always a *typical absence** with a rhythmic 3/sec spike-and-wave discharge. The myoclonus occurs at the same frequency and is synchronous with the spikes on the EEG.

absence, petit mal. Syn. for *typical absence.*

absence, petit mal variant. Syn. for *atypical absence.*

absence, pure. Syn. for *simple absence.*

absence, retrocursive. A type of *hypertonic absence** in which the subject walks backwards as a result of the backward extension of his body produced by an increase in postural tone.

absence, retropulsive. A type of *hypertonic absence** characterized by a backward extension of the body due to an increase in postural tone.

absence, simple. An *absence** characterized mainly or exclusively by clouding or loss of consciousness. Syn.: *pure absence.*

absence, sternutatory. A *complex absence** characterized by sneezing.

absence, subclinical. An *absence** that is easily recognizable from the EEG but that clinically is limited to a slight decrease of alertness (or perhaps merely a slight loss of efficiency or of decision-making ability); it can be assessed only by appropriate psychometric tests. See *seizure, subclinical epileptic* (Ling.).

absence, temporal lobe. An incorrect term occasionally used to designate a *non-convulsive epileptic seizure** resulting from a discharge in the temporal lobe and characterized by clouding or loss of consciousness.

Ling.: The expression *"temporal lobe partial seizure* with clouding of consciousness" is preferable to the term "absence", which must be reserved for brief episodes of loss of consciousness occurring as an expression of generalized epilepsy (see *absence,* Expl.). Some authors, however, consider that the problem can be resolved by using the term *temporal lobe pseudo-absence**.

absence, tussive. A *complex absence** characterized by bouts of coughing. See *seizure, tussive epileptic.*

absence, typical. A type of simple or complex *absence** associated by definition with a bilateral, synchronous, and symmetrical EEG discharge of 3/sec spike-and-wave complexes. Clinical features often include a sudden onset and termination, and flickering of the eyelids, or other bilateral clonic phenomena, occurring synchronously with each spike on the EEG. Such absences generally occur in subjects who are apparently free from organic brain disease but who have a familial *epileptic predisposition** (see *epilepsy, primary generalized*). The pathogenesis of these attacks is highly specific (see *seizure, centrencephalic epileptic*); they are easily induced by hyperventilation and are particularly responsive to the so-called "anti-petit mal" drugs (diones, succinimides, benzodiazepines, etc.). All these features make it easy to distinguish typical absences from *atypical absences**. Syn.: *petit mal absence.*

absence, vasomotor. A *complex absence** characterized by marked vasomotor phenomena.

absentia epileptica. Syn. for *absence.*

activation of the epilepsies. A technique designed to induce either clinical and EEG manifestations of an

epileptic attack or *interictal (electroencephalographic) epileptic discharges**. Many different methods are employed, but the most common are hyperventilation, intermittent photic stimulation, natural or induced sleep, sleep deprivation, and the cautious injection of convulsant drugs (pentetrazol, bemegride, etc.). Activation methods must be used with care, since those that are the most effective (injections of convulsants) are liable to produce a nonspecific *occasional epileptic seizure** in any subject, epileptic or not. See *predisposition, epileptic; threshold, epileptic.*

affect-epilepsy. Syn. for *affective epilepsy.*

alcohol-epilepsy. Syn. for *alcohol-induced epilepsy.*

amaurosis, epileptic. Transient amaurosis accompanying or following an attack of *occipital epilepsy**. In such cases the amaurosis is the main symptom of a *visual elementary epileptic seizure**.

anticonvulsant. 1. (Adj.) Preventing or arresting convulsions. 2. (Noun) A drug that prevents or stops convulsions, primarily those of an epileptic nature.

antiepileptic. 1. (Adj.) Preventing or arresting *epileptic seizures**, or used to treat epilepsy. 2. (Noun) A drug that prevents or stops convulsive or non-convulsive epileptic seizures.

aphasia, paroxysmal. Incorrect syn. for *aphasic epileptic seizure.*

Ling.: It is well established that almost all aphasic episodes of very brief duration (less than one minute) are the expression of an epileptic seizure of temporal or frontal lobe origin. It is an equally well known fact that most aphasic episodes of longer duration (a few minutes) reflect a non-epileptic *seizure**, particularly ischaemic seizures of the Sylvian region of the dominant hemisphere.

arrest, epileptic speech. See *speech arrest, epileptic.*

attack. A sudden episode affecting a person in apparently good health, or a sudden worsening of a chronic condition (e.g., attack of appendicitis, heart attack, apoplectic attack).

attack, cataplectic. A sudden and very brief loss of postural tone affecting part or all of the body musculature and occurring in certain narcoleptic subjects independently of any epileptic mechanism.

Ling.: This term should never be used in reference to epilepsy. See *seizure, atonic epileptic.*

attack, cerebellar. Syn. for *cerebellar seizure.*

attack, cerebral. Syn. for *seizure.*

attack, epileptic drop. An *atonic epileptic seizure** in which the decrease or abolition of postural tone is of very brief duration (generally a fraction of a second). Depending on whether the loss of tone involves all the postural muscles or only those of the head and neck, the subject either slumps to the ground or his head suddenly falls onto his chest (*epilepsia nutans**). He gets up again immediately after the fall, which may be violent enough to cause injury, particularly when his head strikes an object in its path. Such epileptic drop attacks coincide with the slow waves of polyspike-wave complexes on the EEG and are very typical in young people, in whom they may occur in association, and sometimes in combination, with *epileptic myoclonus** (in which case they are called *myoclono-atonic epileptic seizures**). Epileptic drop attacks are sometimes described as *akinetic petit mal** because they are accompanied by a polyspike-wave discharge, but this view is untenable since drop attacks never occur in subjects with *primary generalized epilepsy** in association with *petit mal myoclonus** or *typical absences**. They are only observed, alone or in association with either myoclonus or tonic seizures, in children with chronic encephalopathy and some degree of mental retardation, especially the *Lennox-Gastaut syndrome**. See *petit mal; petit mal, atonic.* Syn. (all incorrect): *epileptic collapse; akinetic epileptic seizure* (established by usage); *akinetic petit mal; astatic epileptic seizure; epileptic fall; static epileptic seizure* (obsolete).

attack, hysterical. See *seizure.*

attack, laughing. A brief and unmotivated attack of laughter constituting the essential manifestation of some *affective epileptic seizures**, usually of temporal lobe origin. Syn.: *explosive laughter; ictus ridendi.*

attack, salaam. See *salaam attack.*

attack, tetanoid. 1. Obsolete syn. for *decerebrate seizure.* 2. Obsolete syn. for *tonic epileptic seizure.*