

The Classification of Seizures and Epilepsy Syndromes

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1. Introduction

Understanding the classification of epileptic seizures is the first step towards the correct diagnosis, treatment and prognostication of the condition. The initial management of a patient with seizures begins with an understanding of the patient's seizure type and, if pertinent, epilepsy syndrome. Specific seizure types or syndromes often respond better to specific medications or surgical approaches. Some seizure types or syndromes carry a benign prognosis or high likelihood of seizure remission by a certain age. Other seizure syndromes may carry a far poorer prognosis, and early knowledge of this allows focused treatment and lifestyle modifications for patients and families.

The classification of epileptic seizures is still largely based on clinical observation and expert opinions. The International League Against Epilepsy (ILAE) first published a classification system in 1960. The last official update for seizures was published in 1981, and the last official update for the epilepsies was in 1989. By definition, epilepsy is diagnosed after a patient has two or more unprovoked seizures. The 1981 and 1989 updates form the officially accepted classification system, although there continues to be efforts to develop a clinically meaningful revision to the current system. A report in 2010 by the ILAE Commission on Classification and Terminology recommended that changes be made in the current conceptualization, terminology, and definitions of seizures and epilepsy. This chapter will focus primarily on the currently accepted standard based on the 1981 and 1989 reports, and discuss the recommendations of the 2010 ILAE report.

2. The classification of epileptic seizures

2.1 Partial seizures

Partial or focal seizures comprise one of the two main classes of epileptic seizures, with generalized seizures being the other. Partial seizures are subdivided between simple and complex partial seizures, which are distinguished by the presence or absence of impairment of consciousness. Simple partial seizures are defined as seizures without impairment of consciousness while complex partial seizures are defined as seizures with impairment of consciousness. Consciousness is defined as the "degree of awareness and/or responsiveness of the patient to externally applied stimuli". Responsiveness refers to the ability of the patient to respond to external stimuli, and awareness refers to the recall of events occurring

during the ictal period. These two features of consciousness are usually tested during and after a seizure in an epilepsy monitoring unit. A patient may be able to follow commands during a seizure, but may not be able to recall portions of the event afterwards, which indicates intact responsiveness but impaired awareness.

Partial seizures manifest themselves in many different forms, depending on which area of the cortex is involved in the onset and spread of the ictal discharge. Partial seizures originate from a focal area of cerebral cortex and may spread to other cortical regions either unilaterally or bilaterally. A partial seizure may manifest with motor signs, autonomic symptoms, somatosensory or special sensory symptoms, or psychic symptoms. The term aura comes from the Latin word "breeze" and is synonymous with a simple partial sensory or psychic seizure. An aura often reflects the location of the seizure onset zone, although there are exceptions.

2.1.1 Simple partial seizures

Focal motor seizures can originate in the precentral gyrus or spread to the precentral gyrus from neighboring cortical regions. They can remain focal, causing right hand clonic activity for example, or can spread or "march" along the motor strip involving different areas of the motor homunculus. This type of seizure is known as a "Jacksonian seizure" and often clinically manifests as clonic activity originating in the hand and then marching up the ipsilateral arm, shoulder, face, and leg. After a focal motor seizure, post-ictal weakness (Todd's paralysis) can last for minutes to hours. The mechanism of Todd's paralysis is thought to be either from "neuronal exhaustion due to the increased metabolic activity of the discharging focus" or from "increased inhibition in the region of the focus." Epilepsia Partialis Continua (EPC) is defined as a continuous focal motor seizure which remains confined to a specific body part and usually consists of clonic movements which can persist for up to months with preserved consciousness (1981). EPC can be seen in Rasmussen Syndrome, focal lesions (cortical dysplasia, vascular lesions, or tumors), nonketotic hyperglycemia, and some inborn errors of metabolism (MERRF) (Engel, 2006).

In a series of 14 patients with focal motor seizures who underwent epilepsy surgery at Mayo Clinic, 11 patients were seizure-free post operatively (Sandok & Cascino, 1998). Other types of focal motor seizures originating from the language area include those with a motor speech arrest or vocalization. Versive seizures originating from the dorsolateral frontal cortex (frontal eye fields) involve contralateral head, eye, or trunk deviation. Tonic seizures originating from the SMA (supplementary motor area) involve abrupt bilateral or asymmetric posturing usually of the contralateral arm, where sometimes the contralateral arm is abducted, externally rotated, and elevated and the head is also deviated contralaterally. This has been termed the "fencing posture" or M2e sign. Consciousness is usually preserved.

Simple partial seizures can also have autonomic symptoms such as vomiting, sweating, piloerection, pupil dilation, pallor, flushing, borborygmi, and incontinence. Simple partial seizures with somatosensory symptoms originating from the post central gyrus may include feelings of focal paresthesias ("pins and needles"), numbness, warmth, or electrical shock-like sensations which can also spread like Jacksonian seizures (a sensory Jacksonian march). Simple partial seizures with somatosensory symptoms can also originate from the secondary sensory area which lies above the Sylvian fissure anterior to the precentral gyrus. Secondary sensory seizures are characterized by more widespread involvement of the sensation (contralateral, ipsilateral, and bilateral involvement) and may include symptoms of feeling cold, pain, or the desire to move (Penfield W et. al., 1950). Sensory seizures can also originate from the supplementary sensory area, which is just posterior to the supplementary motor area, and involve tingling, the desire for movement, feeling stiff, pulling, pulsing, and heaviness (Lim et al.,

1994). Finally, sensory seizures can also originate from the insular cortex. The symptoms often involve the naso-oropharyngeal-laryngeal regions and consist of throat paresthesias, warmth, tightening, or a sense of strangulation or suffocation (Nguyen et al., 2009).

Simple partial seizures with special sensory symptoms include visual, auditory, gustatory, olfactory, and vertiginous symptoms. Visual seizures can originate from primary visual cortex and consist of primary visual hallucinations such as flashing lights, spots, stars, or circles of colored light which can appear in the contralateral visual field or directly ahead. More complex visual hallucinations originate from visual association cortex and can include seeing persons or scenes. One patient described seeing Fred Flintstone and The Gingerbread Man at the onset of seizures. Post-ictal darkness or blindness can follow simple visual seizures. Auditory seizures which arise from the lateral temporal region, specifically the superior temporal gyrus and Heschl's gyrus, can include the clinical symptoms of buzzing, ringing, hearing a rushing sound, hyper- or hypoacusis, sound distortion, or hearing words or music. Olfactory seizures originating from the uncinate gyrus or mesial temporal region typically involve smelling unpleasant odors such as burning rubber, smoke, or sulfur. Gustatory sensations originating from the temporal lobe, insula, or parietal operculum can be pleasant or unpleasant and usually are described as a metallic taste but can also be bitter or sweet. On rare occasions, vertiginous symptoms may also be a type of simple partial seizure which originates from the lateral temporal region.

Simple partial seizures with psychic symptoms indicate a disturbance of higher cortical function. For example, dysphasic symptoms include expressive or receptive language disturbances and may involve repetition of a word or phrase (epileptic palilalia). Dysmnesic symptoms involve a distortion of memory and include déjà vu, jamais-vu, déjà-entendu, jamais-entendu, autoscopy, or panoramic vision (see Table 1). Other cognitive disturbances such as dreamy states, distorted time sense, derealization, or a sense of unreality may be present. Emotional symptoms include pleasure, fear, or anger which occurs in paroxysms lasting seconds to minutes. Illusions may be present which result in distorted perceptions of the person him or herself or objects around him or her. Structured hallucinations can take the form of music or scenes and may affect multiple sensory modalities (somatosensory, visual, olfactory, or gustatory). Primitive hallucinations originate from the corresponding primary sensory area whereas more complex and elaborate hallucinations originate from the corresponding association cortices. Psychic auras often originate from the temporal lobe.

Psychic Aura	Definition
Déjà vu	An illusion of a familiar memory
Jamais vu	When what should be a familiar visual experience becomes unfamiliar
Déjà entendu	An auditory illusion of something familiar
Jamais entendu	When what should be a familiar auditory experience becomes unfamiliar
Autoscopy	Seeing oneself in external space, as if the mind has left the body
Derealization	A feeling of unreality of the outside world; the world seems strange and unreal
Depersonalization	A feeling of unreality in one's sense of self; feeling as if in a dream or watching oneself act
Macro-/Micropsia	Objects appear larger or smaller than usual
Macr-/Micracusia	Sounds are louder or softer than usual

Table 1. Psychic Auras

2.1.2 Complex partial seizures

Complex partial seizures are partial seizures with impairment of consciousness. They may start as simple partial seizures (auras) and progress to complex partial seizures or may begin as complex partial seizures with impairment of consciousness at the onset of the seizure. They may or may not involve automatisms. The clinical features of the complex partial seizure depend on the region affected by abnormal electrical activity. Complex partial seizures usually originate in the frontal or temporal lobes but can occur in the parietal or occipital lobes.

3. Generalized seizures

Absence Seizures are characterized by a sudden onset behavioral arrest, a blank stare, unresponsiveness, and sometimes a brief upward rotation of the eyes. The duration is typically a few seconds to half a minute. There is little to no post-ictal confusion, and the patient typically resumes the activity he/she was doing prior to the seizures. This seizure type is also referred to as simple absence. The ILAE's 1981 classification also recognizes five subtypes of absence seizures: absence with impairment of consciousness only, with mild clonic components, with atonic components, with tonic components, and with automatisms. In absence with mild clonic components, there are subtle clonic movements of the eyelids, corner of the mouth, or upper extremities sometimes at a frequency of 3 Hz. In absence with atonic components, there is a loss of postural tone causing the head to drop, the trunk to slump forward, the arms to drop, or the grip to relax. Falls are rare. In absence with tonic components, tonic muscle contraction of the trunk and neck extensors may cause the head to extend and the trunk to arch, thus causing retropulsion. Tonic contraction of the neck muscles may cause the head or trunk to deviate to one side. In absence with automatisms, the patient engages in purposeful or semipurposeful repetitive movements while consciousness is impaired. Examples of automatisms may include lip licking, chewing, lip smacking, swallowing, grimacing, smiling, yawning, fumbling with the hands, picking, scratching, rubbing, or aimless walking. Absence with autonomic phenomena is another subtype with signs of tachycardia, pallor, flushing, piloerection, salivation, or urinary incontinence. A mixture of clonic, atonic, tonic, automatic, and autonomic features may occur which is also referred to as complex absence. The ictal EEG pattern in absence seizures is a 3 Hz generalized monomorphic spike and wave with abrupt onset and termination. Absence seizures in the EEG laboratory can be precipitated by hyperventilation and less commonly photic stimulation.

Atypical absence seizures are usually seen in patients with symptomatic generalized epilepsy. They are similar to absence seizures in that they have both simple and complex presentations. One distinguishing feature is that they are less abrupt in onset clinically. The seizures are usually less than 10 seconds but may be prolonged and result in absence status. They also are not usually induced by hyperventilation or photic stimulation. The ictal EEG pattern consists of a less monomorphic slow spike and wave discharge characterized by a blunt sharp wave and occurs at a frequency of < 2.5 Hz.

Tonic-clonic seizures, also known as grand mal seizures, are characterized by abrupt loss of consciousness followed by tonic contraction of the muscles. This leads to the ictal cry, where air is forcefully expired against a closed glottis. The mouth is forcefully closed which can result in a tongue bite. The pupils become dilated and the eyes deviate upwards. The upper extremities often symmetrically abduct and flex at the elbows while the lower extremities

may briefly flex and then extend and adduct with the toes pointed. Clonic activity then ensues which is initially rapid and then slows. Gasping respirations occur as the respiratory muscles are involved in the clonic activity. The patient may become cyanotic. Urinary incontinence may occur. At the end of the seizure, the patient is unconscious for a brief period of time and then gradually recovers. Patients typically report generalized muscle soreness and sometimes a headache post-ictally.

Tonic-clonic seizures may occur independently, may arise from other generalized seizures, or may occur during secondary generalization of a partial onset seizure. The semiologic features of tonic-clonic seizures in primary generalized epilepsy may be bilaterally symmetric or may involve a forced head deviation to either side (Ochs et al., 1984). During secondarily generalized partial onset seizures, patients often assume a figure-4 posture where the contralateral arm extends, and the ipsilateral arm flexes at the elbow. This posture can occur with the legs as well. Tonic-clonic seizures may lead to injuries such as burns, head injuries, vertebral compression fractures, shoulder dislocations, and tongue and cheek lacerations.

Myoclonic seizures are generalized seizures characterized by brief, irregular, shock-like jerks of the head, trunk, or limbs. They can be symmetric or asymmetric and involve the whole body, regions of the body, or focal areas. They tend to occur close to sleep onset and upon awakening from sleep. Myoclonic seizures can be a feature of some idiopathic generalized epilepsies (Juvenile Myoclonic Epilepsy), symptomatic generalized epilepsies (Myoclonic-Astatic Epilepsy), the progressive myoclonic epilepsies (Lafora Disease), and infantile spasms. Myoclonus can be positive or negative. Negative myoclonus refers to the brief loss of postural tone when the body part is held against gravity. Consciousness is not impaired and there is no post-ictal confusion with single myoclonic jerks. Myoclonic seizures can occur in clusters and evolve into clonic-tonic-clonic seizures, with resultant loss of consciousness and postictal confusion. The ictal EEG pattern is characterized by brief generalized polyspike or polyspike and wave discharges which corresponds to the myoclonic jerk.

Tonic seizures are seizures which involve tonic contraction of the face, neck, axial, or appendicular musculature lasting from 10 seconds to one minute. They can involve extension or flexion of the muscles and often lead to falls and head injuries. They may be more subtle and involve only upward eye deviation. They often occur out of NREM sleep. They are usually seen in patients with symptomatic generalized epilepsy and are one of the common seizure types in patients with Lennox-Gastaut syndrome. They can also occur in epilepsy with myoclonic-astatic seizures. The ictal EEG usually shows a brief generalized attenuation of cerebral activity followed by generalized paroxysmal fast activity in the beta frequency range.

Clonic seizures are generalized seizures that are characterized by repetitive rhythmic clonic jerks (1-2 Hz) with impairment of consciousness and a short post-ictal phase. They can lead into a clonic-tonic-clonic seizure. It is thought that the repetitive discharges are due to rhythmic excitatory discharges from the cortex (1981; Engel, 2006). The ictal EEG demonstrates generalized polyspike and wave discharges or generalized fast activity.

Atonic seizures are characterized by a sudden loss of muscle tone which can lead to a head drop, a limb drop, or a drop of the whole body (a.k.a. – a drop attack). There is a brief loss of consciousness and injuries, particularly to the face, may occur (1981). Atonic seizures last less than 5 seconds, and there is minimal post-ictal confusion. Atonic seizures may be preceded by

a brief myoclonic jerk or tonic component. Atypical absence seizures may have an atonic component. The criteria distinguishing between negative myoclonus, atonic seizures, and some atypical absences still needs to be developed (Engel 2006). Atonic seizures are usually seen in the symptomatic generalized epilepsies such as Lennox-Gastaut syndrome. The ictal EEG typically shows a high voltage spike and wave or slow wave followed by a generalized attenuation of cerebral activity or low voltage paroxysmal fast activity.

4. Unclassified epileptic seizures

This category listed in the ILAE's Classification of Epileptic Seizures (1981) includes all seizures that defy classification due to incomplete data. An example is seizure in infancy, which may involve chewing, swimming movements, eye movements, jittering, and apnea, and have not yet been subtyped.

5. The classification of epilepsies and epileptic syndromes

An epileptic disorder can be symptomatic, idiopathic, or cryptogenic. Symptomatic is a term that means the etiology is known—usually a structural lesion within the brain. Idiopathic is a term that refers to an epilepsy of presumed genetic etiology without a structural brain lesion or other neurological signs or symptoms. Cryptogenic is a term that refers to an epilepsy that is presumed to be symptomatic but the etiology is unknown (1989). The term cryptogenic has been replaced by “probably symptomatic” (Engel, 2001). The 1989 classification system is divided into four main categories: localization-related (focal, local, or partial), generalized, epilepsies and syndromes undetermined whether focal or generalized, and special syndromes (see Table 2).

1. Localization-related epilepsies and syndromes

1.1 Idiopathic

- Benign childhood epilepsy with centrotemporal spikes
- Childhood epilepsy with occipital paroxysms
- Primary reading epilepsy

1.2 Symptomatic

- Chronic progressive epilepsia partialis continua of childhood (Kojewnikow's syndrome)
- Syndromes characterized by seizures with specific modes of precipitation
- Temporal lobe epilepsies
- Frontal lobe epilepsies
- Parietal lobe epilepsies
- Occipital lobe epilepsies

1.3 Cryptogenic

2. Generalized epilepsies and syndromes

2.1 Idiopathic

- Benign neonatal familial convulsions
- Benign neonatal convulsions
- Benign myoclonic epilepsy in infancy
- Childhood absence epilepsy

- Juvenile absence epilepsy
 - Juvenile myoclonic epilepsy
 - Epilepsy with GTCS on awakening
 - Other generalized idiopathic epilepsies not defined above
 - Epilepsies with seizures precipitated by specific modes of activation
- 2.2 Cryptogenic or symptomatic
- West syndrome
 - Lennox-Gastaut syndrome
 - Epilepsy with myoclonic-astatic seizures
 - Epilepsy with myoclonic absences
- 2.3 Symptomatic
- 2.3.1 Non-specific etiology
- Early myoclonic encephalopathy
 - Early infantile epileptic encephalopathy with suppression burst
 - Other symptomatic generalized epilepsies not defined above
- 2.3.2 Specific syndromes
- Diseases in which seizures are a presenting or predominant feature
3. Epilepsies and syndromes undetermined whether focal or generalized
- 3.1 With both generalized and focal seizures
- Neonatal seizures
 - Severe myoclonic epilepsy in infancy
 - Epilepsy with continuous spike-waves during slow wave sleep
 - Acquired epileptic aphasia (Landau-Kleffner syndrome)
 - Other undetermined epilepsies not defined above
- 3.2 Without unequivocal generalized or focal features (i.e. – Sleep related GTCS; when the EEG shows both focal and generalized ictal or interictal discharges, and when focal or generalized onset cannot be determined clinically)
4. Special syndromes
- 4.1 Situation-related seizures
- Febrile convulsions
 - Isolated seizures or isolated status epilepticus
 - Seizures occurring only when there is an acute metabolic or toxic event (alcohol, drugs, eclampsia, nonketotic hyperglycemia)

Table 2. ILAE's 1989 International Classification of Epilepsies and Epileptic Syndromes, from (1989). Proposal for revised classification of epilepsies and epileptic syndromes. Commission on Classification and Terminology of the International League Against Epilepsy. *Epilepsia*, Vol. 30, No. 4, (August 1989), pp. 389-99, ISSN 1528-1167

An epilepsy syndrome is defined as “a complex of signs and symptoms that define a unique epilepsy condition” (Engel, 2001). The groups of syndromes are: idiopathic focal epilepsies of infancy and childhood, familial (autosomal dominant) focal epilepsies, symptomatic (or probably symptomatic) focal epilepsies, idiopathic generalized epilepsies, reflex epilepsies, epileptic encephalopathies, progressive myoclonus epilepsies, and seizures not necessarily requiring a diagnosis of epilepsy (see Table 3). There are over 25 specific syndromes in the 1989 ILAE report. A discussion regarding a few of the more common syndromes affecting adolescents and adults follows.

Groups of syndromes	Specific syndromes
Idiopathic focal epilepsies of infancy and childhood	Benign infantile seizures <ul style="list-style-type: none"> • Benign childhood epilepsy with centrotemporal spikes • Early-onset benign childhood occipital epilepsy • Late-onset childhood occipital epilepsy
Familial (autosomal dominant) focal epilepsies	Benign familial neonatal seizures Benign familial infantile seizures Autosomal dominant nocturnal frontal lobe epilepsy Familial temporal lobe epilepsy Familial focal epilepsy with variable foci*
Symptomatic (or probably symptomatic) focal epilepsies	Limbic epilepsies <ul style="list-style-type: none"> • Mesial temporal lobe epilepsy with hippocampal sclerosis • Mesial temporal lobe epilepsy defined by specific etiologies • Other types defined by location and etiology Neocortical epilepsies <ul style="list-style-type: none"> • Rasmussen syndrome • Hemiconvulsion-hemiplegia syndrome • Other types defined by location and etiology • Migrating partial seizures of early infancy*
Idiopathic generalized epilepsies	Benign myoclonic epilepsy in infancy Epilepsy with myoclonic astatic seizures Childhood absence epilepsy Epilepsy with myoclonic absences Idiopathic generalized epilepsies with variable phenotypes <ul style="list-style-type: none"> • Juvenile absence epilepsy • Juvenile myoclonic epilepsy • Epilepsy with GTCS only Generalized epilepsies with febrile seizures plus*
Reflex epilepsies	Idiopathic photosensitive occipital lobe epilepsy Other visual sensitive epilepsies Primary reading epilepsy Startle epilepsy

Groups of syndromes	Specific syndromes
Epileptic encephalopathies	Early myoclonic encephalopathy Ohtahara syndrome West syndrome Dravet syndrome Lennox-Gastaut syndrome Landau-Kleffner syndrome Epilepsy with continuous spike-waves during slow-wave sleep
Progressive myoclonic epilepsies	Ceroid lipofuscinosis Sialidosis Lafora disease Unverricht-Lundborg disease Neuroaxonal dystrophy MERRF Dentatorubropallidoluysian atrophy
Seizures not necessarily requiring a diagnosis of epilepsy	Benign neonatal seizures Febrile seizures Reflex seizures Alcohol-withdrawal seizures Drug or other chemically induced seizures Immediate and early posttraumatic seizures Single seizures or isolated clusters of seizures Rarely repeated seizures (oligoepilepsy)

* Syndromes in development

Table 3. An Example of a Classification of Epilepsy Syndromes, from Engel, J., Jr. (2001). A proposed diagnostic scheme for People with epileptic seizures and with epilepsy: report of the ILAE Task Force on Classification and Terminology. *Epilepsia*, Vol.42, No. 6, (June 2001), pp. 796-803, ISSN 1528-1167

5.1 Temporal lobe epilepsies

Temporal lobe seizures are the most common type of partial epilepsy. Temporal lobe seizures often begin with an aura (Quesney, 1986). Auras may include viscerosensory symptoms (epigastric sensation, thoracic sensation, and warm ascending sensation) or sensory illusions or hallucinations. The ictal event is usually characterized by a blank stare, loss of contact with the environment, oroalimentary or vocal automatisms, hand automatisms, upper limb tonic or dystonic posturing, early head or eye deviation, and dysphasia. Oroalimentary automatisms are defined as stereotyped, repetitive movements of the mouth, tongue, lips, or jaw which have the appearance of chewing or lip-smacking. They may also involve gulping, swallowing, or spitting. Hand automatisms are repetitive, purposeless movements of the hands including grasping, fumbling, and searching movements. Both oroalimentary and hand automatisms often localize to the mesial temporal lobe. Table 4 lists other ictal signs with localizing and lateralizing value. In terms

of lateralization (determining right vs. left hemispheric involvement), unilateral automatisms and post-ictal dysphasia were determined to have the highest predicative value. Unilateral automatisms are typically ipsilateral to region of seizure onset, and post-ictal dysphasia lateralized to the dominant hemisphere (Chee et al., 1993). Nondominant temporal lobe seizures can have preservation of language and responsiveness with minimal post-ictal confusion. Patients with right temporal lobe epilepsy due to MTS (mesial temporal

Clinical Event	Localization/Lateralization
Head turn	
• Early non-forced	Ipsilateral temporal
• Forced	
Early forced	Frontal
Late forced	Contralateral temporal (in process of generalizing)
Ocular version	Contralateral occipital
Focal clonic	Contralateral peri-rolandic or temporal
Dystonic limb	Contralateral temporal > frontal
Unilateral tonic limb	Contralateral hemisphere
M2e sign (fencing posture)	Contralateral frontal > temporal
Figure 4	Contralateral hemisphere (to extended arm)
Ictal paresis	Contralateral hemisphere
Todd's paresis	Contralateral hemisphere (extratemporal > temporal)
Unilateral blinking	Ipsilateral hemisphere
Unilateral limb automatism	Ipsilateral hemisphere
Postictal nose rubbing	Ipsilateral temporal > frontal (to hand used)
Postictal cough	Temporal
Bipedal automatisms	Frontal > temporal
Hypermotor	SMA (supplementary motor area)
Ictal spitting	Right temporal
Automatisms with preserved responsiveness	Right temporal
Gelastic	Hypothalamic, mesial temporal
Ictal vomiting/retching	Right temporal
Ictal urinary urge	Non-dominant temporal
Loud vocalization	Frontal > temporal
Ictal speech arrest	Temporal
Ictal speech preservation	Non-dominant hemisphere
Post-ictal aphasia	Language-dominant hemisphere

Table 4. Localization and Lateralization of Ictal Seizure Semiology

sclerosis) may be able to speak normally during the seizure. Patients with left temporal lobe seizures due to MTS often make prominent paraphasic errors during and after seizures. Specific semiological features are also helpful in distinguishing between mesial, mesial-lateral, and lateral temporal lobe epilepsy. Mesial temporal lobe seizures are often characterized by an initial epigastric sensation or viscerosensory sensation, fear, a dreamy state, longer seizure duration, delayed loss of contact, and delayed oroalimentary and upper limb automatisms while lateral temporal lobe seizures are characterized by an initial sensory illusion or hallucination (mainly auditory), an initial loss of contact, a shorter duration (< 1 minute), and frequent secondary generalizations. The mesial-lateral temporal lobe seizures were similar to the mesial temporal seizures but had an earlier loss of contact and earlier oroalimentary, verbal, and vocal automatisms (Maillard et al., 2004). It is important to distinguish between mesial (limbic) and lateral (neocortical) temporal lobe epilepsy if one is considering a surgical option such as a temporal lobectomy, as postsurgical seizure-freedom and complication rates differ.

5.2 Frontal lobe epilepsies

Frontal lobe seizures are the second most common type of focal epilepsy and occur in approximately 30% of patients with partial epilepsy (Bancaud & Talairach, 1992). Frontal lobe seizures are often confused with pseudoseizures due to the bizarre clinical semiology. Frontal lobe seizures are usually brief (less than 30 seconds), tend to occur in clusters, can occur multiple times per day, and often have minimal or no post-ictal confusion. The clinical semiology includes an abrupt onset of stereotyped hypermotor behavior and may include vocalizations, gestural or sexual automatisms, and bilateral leg automatisms consisting of pedaling or bicycling movements. The seizure semiology of frontal lobe seizures varies depending on what region of the frontal lobe is involved. The patient may have asymmetric tonic extension of the proximal extremities, as in SMA seizures, or clonic activity of the contralateral limb, as in seizures from the lateral convexity. Seizures originating from the mesial frontal region or SMA are characterized by vocalizations and abrupt tonic extension of the proximal extremities which may be bilateral and is often asymmetric. There is minimal impairment of consciousness or post-ictal confusion. Lateral dorsal frontal lobe seizures are characterized by speech arrest, forced thinking, contraversive head and eye deviation, and automatisms such as laughing, crying, sniffing, chewing, or kicking. Orbitofrontal seizures are characterized by prominent autonomic symptoms (flushing, mydriasis, tachycardia), automatisms, and loud vocalizations. They can also appear similar to mesial temporal lobe seizures due to rapid spread to this region. Cingulate gyrus seizures are similar to SMA seizures but also involve behavioral arrest, oroalimentary automatisms, gestural or sexual automatisms, mood changes, and sometimes urinary incontinence. Because of the extensive inter-regional connectivity within the frontal lobe and rapid seizure propagation, frontal lobe seizures are difficult to localize on the basis of clinical semiology.

5.3 Parietal lobe epilepsies

Parietal lobe seizures account for < 10% of focal seizures. Often they arise from clinically silent areas and only manifest symptoms when the seizure spreads to other functional cortical regions. They can spread to the occipital, temporal, or frontal regions. Clinically,

patients may report somatosensory symptoms, most commonly in the face and hand, contralateral to the seizure focus. In nondominant parietal lobe seizures, patients can have spatial neglect of the contralateral body or environment. In dominant parietal lobe seizures, patients may have language dysfunction.

5.4 Occipital lobe epilepsies

Occipital lobe seizures also account for < 10% of focal seizures. They are characterized by elementary visual hallucinations of fixed or moving flashing white or colored lights which start in the contralateral visual field and can spread to the entire visual field. Patients may also report a “whiting out” or “blacking out” of their vision. The eyes may deviate contralaterally, and the eyelids may rapidly blink. The remainder of the seizure is characterized by where the seizure discharge spreads. If the seizure spreads to the posterior temporal region (area of visual association cortex), complex visual hallucinations may occur. Occipital seizures may also spread to the mesial temporal, parietal, and perirolandic regions and mimic seizures of those regions.

5.5 Autosomal dominant nocturnal frontal lobe epilepsy

Autosomal Dominant Nocturnal Frontal Lobe Epilepsy (ADNFLE) is a familial autosomal dominant focal epilepsy characterized by clusters of brief seizures (5-30 seconds) during NREM sleep (stages N2 and N3). They are often initially misdiagnosed as nightmares or parasomnias. The mean age of onset is around 12 years of age (range 1-30). The seizures themselves are characterized by brief motor attacks, usually with a dystonic or dyskinetic component. During a seizure patients may have complex and bizarre behaviors, shouting, bimanual and bipedal automatisms, mumbling, urinary incontinence, and rarely violent behavior. The ictal EEG may demonstrate a frontally predominant ictal discharge in approximately 30% of patients and focal background attenuation or focal rhythmic slowing over the anterior head regions in about 55% of patients (Oldani et al., 1998). The *CHRNA4* gene on chromosome 20, which encodes the neuronal nicotinic acetylcholine receptor (nAChR) alpha 4 subunit, is mutated in patients with ADNFLE type 1. The *CHRN2* gene on chromosome 1 is mutated in patients with ADNFLE type 3. The molecular pathogenesis of how these mutations cause ADNFLE is unknown (Hirose et al., 2005).

5.6 Autosomal dominant partial epilepsy with auditory features

Autosomal Dominant Partial Epilepsy with Auditory Features (ADPEAF) or Autosomal Dominant Lateral Temporal Epilepsy (ADLTE) is also a familial autosomal dominant focal epilepsy characterized by lateral temporal lobe epilepsy and auditory aura. It is due to a mutation in the *LGI1* gene (leucine-rich glioma-inactivated 1 gene) which is expressed in neurons in the neocortex and limbic regions (Hirose et al., 2005). Mutations in the *LGI1* gene have been found in 50% of families with this type of epilepsy (Ottman et al., 2004). The age of onset is between 1 and 60 years with a mean of 18 years. The seizures are characterized by auditory auras (64%), complex visual (17%), psychic (16%), autonomic (12%), vertiginous (9%), other sensory (13%), and aphasia (17%). The majority of auditory auras are simple in nature (humming, buzzing, ringing). A minority of patients report complex hallucinations such as music or voices. The MRI of the brain is normal, and patients typically have a good response to treatment with antiepileptic drugs (Michelucci, 2003, 2009).

5.7 Mesial temporal lobe epilepsy with hippocampal sclerosis

Mesial temporal lobe epilepsy with hippocampal sclerosis (MTLE-HS) is a symptomatic focal epilepsy and subcategorized as a limbic epilepsy (vs. neocortical epilepsy). Mesial temporal lobe epilepsy is one of the most common types of epilepsy referred for epilepsy surgery and is often refractory to AEDs. The age of onset is between late childhood to mid adolescence. Patients often had febrile convulsions in infancy or early childhood. Most patients report an aura. Common auras include an epigastric sensation (a rising sensation, butterflies, nausea), fear, olfactory hallucinations, lightheadedness, and déjà vu (French et al., 1993). Complex partial seizure semiology may also consist of ipsilateral upper extremity automatism and ipsilateral early non-forced head turn. Contralateral dystonic posturing, Todd's paralysis, and late forced head turn prior to secondary generalization can also be seen.

Hippocampal sclerosis is the most common pathological substrate found in patients with medial temporal lobe epilepsy who undergo surgical resection. Hippocampal sclerosis is strongly associated with prolonged febrile seizures in childhood, but the cause is still unknown. The majority of patients who undergo surgical resection for MTLE-HS become seizure free (Ozkara et al., 2008). Is an example of a patient with right mesial temporal sclerosis who was rendered seizure-free after a right temporal lobectomy.

5.8 Juvenile absence epilepsy

Juvenile absence epilepsy (JAE) is classified as an idiopathic generalized epilepsy. The age of onset is typically at or after puberty between the ages of 10-17. Unlike in childhood absence epilepsy (CAE) where absence seizures can occur hundreds of times per day, absence seizures in JAE may only occur sporadically. There is less impairment of consciousness with absence seizures in JAE compared to absences in CAE. Patients with JAE can have generalized tonic-clonic seizures (usually upon awakening), myoclonic seizures, and even absence status epilepticus. The ictal EEG pattern resembles that of CAE (3 Hz spike and wave) but the discharges tend to vary slightly in frequency (usually > 3 Hz), are more irregular, and include more polyspike discharges. There is a strong genetic component with linkage to chromosomes 5, 8, 18, and 21. The response to antiepileptic medication is usually excellent (Beghi et al., 2006).

5.9 Juvenile myoclonic epilepsy

Juvenile Myoclonic Epilepsy (JME) is also classified as an idiopathic generalized epilepsy. The age of onset is in the mid-teens between the ages of 12-18. Patients may present with myoclonic jerks upon awakening in the morning. Patients may first ignore the myoclonic jerks, often attributing them to clumsiness. Sometimes the diagnosis is not made until the patient has a generalized tonic-clonic seizure. The myoclonus usually involves the neck, shoulders, arms, or legs with the upper extremities being more frequently affected. Consciousness is usually not impaired during the myoclonic seizures. Generalized tonic-clonic and absence seizures are also seen. Generalized tonic-clonic seizures may also occur in the morning upon awakening and can be triggered by sleep deprivation, alcohol, and stress. Often, several myoclonic jerks may precede a generalized tonic-clonic seizure, which is known as a clonic-tonic-clonic seizure. Approximately 50% of patients can be photosensitive. The ictal EEG consists of generalized polyspike and wave discharges > 3 Hz. There is a strong genetic component with linkage to chromosomes 2, 3, 5, 6, and 15. The response to AED treatment is excellent but needs to be continued lifelong in most patients due to a high rate of relapse (Beghi et al., 2006).

5.10 Epilepsy with generalized tonic clonic seizures (GTCS) on awakening

This syndrome is also known as Epilepsy with generalized tonic-clonic seizures only and is classified as one of the idiopathic generalized epilepsies. The age of onset is the second decade of life. GTCS occur > 90% of the time, with absence and myoclonic seizures occurring less frequently. Seizures occur 1-2 hours after awakening from sleep or during periods of relaxation in the evening. Sleep deprivation, alcohol, and photic stimulation can be precipitating factors. The ictal EEG demonstrates frontally predominant fast rhythmic spiking. The prognosis is good if the patient is adequately treated with AED's and avoids provoking factors (1989; Beghi et al., 2006).

5.11 Lennox-Gastaut syndrome

Lennox-Gastaut Syndrome (LGS) is classified as an epileptic encephalopathy. The age of onset is usually before age 8 with a peak age of onset between 3-5 years of age. Rarely, the disorder can present in early adulthood. The syndrome is characterized by a triad of multiple seizure types (tonic and atypical absence are the most common), slow spike and wave on EEG (1-2.5 Hz), and some degree of mental retardation. The etiology can be symptomatic or cryptogenic. It may evolve from West syndrome. Tonic seizures are considered a prerequisite for the diagnosis. Atypical absence and atonic seizures are also common. Myoclonic, generalized tonic-clonic, unilateral clonic, and partial seizures can occur less frequently. Non-convulsive status epilepticus can occur in > 50% of patients and involves near continuous atypical absence seizures interrupted by brief tonic seizures. The interictal EEG is characterized by slow spike and wave complexes (< 2.5 Hz) and activation of generalized paroxysmal fast activity during sleep. The diagnosis may be difficult to make at first because not all features of the syndrome may be present. The seizures in LGS are typically refractory to medical treatment (Arzimanoglou et al., 2009).

5.12 Progressive myoclonic epilepsies (PME)

These diseases are characterized by myoclonic jerks, seizures (GTC, absence, clonic, partial) and dementia caused by cerebral and cerebellar atrophy. The myoclonus is termed "massive myoclonus" which can cause falls and lead into generalized tonic-clonic seizures. Patients may exhibit cerebellar dysfunction, action myoclonus, or extrapyramidal dysfunction. Childhood development is normal until the age of onset. The autosomal recessive forms include Lafora disease, Unverricht-Lundborg disease, the neuronal ceroid lipofuscinoses, sialidosis, Action Myoclonus-Renal Failure Syndrome (AMRF), and Gaucher disease. The autosomal dominant form is dentatorubropallidoluyisan atrophy. PME is also seen in some mitochondrial cytopathies such as myoclonic epilepsy with ragged-red fibers (MERRF).

6. 2010 ILAE commission on classification and terminology report

The 1981 and 1989 ILAE classifications are based on concepts formulated prior to modern neuroimaging and genomic research. The 1989 classification was not a true scientific classification but rather an organized list built on concepts which no longer correspond to or accurately describe our increasing knowledge of seizures and the epilepsies. Numerous attempts have been made by the ILAE Committee (Engel, 2001, 2006) and individual investigators (Luders et al., 1998) to revise the current classification. These attempts have generated controversy, and the lack of consensus has blocked any formal revision until 2010 (Berg et al., 2010). The most recent ILAE Commission on Classification and Terminology

report is an update and revision to the classification schemes which are now several decades old. It was devised to simplify the classification of seizures, as new concepts have emerged broadening our understanding of seizures and epilepsy syndromes. The 2010 ILAE report on revised terminology and concepts for organization of seizures and epilepsies is not a new classification of epilepsies but rather a reflection of new terminology and concepts that lead to a better understanding of the current neurobiology, clinical features, prognostic implications, and features relevant to clinical practice and research. The ILAE did not feel that there was adequate knowledge at this time to propose a new classification of seizures and the epilepsies. The following paragraphs are a summary of the ILAE's report on classification and terminology from 2010 (Berg et al., 2010).

6.1 Generalized and focal redefined

The new definition of generalized epileptic seizures is "originating at some point within, and rapidly engaging, bilaterally distributed networks" which include both cortical and subcortical structures, can appear localized but have inconsistent localization and lateralization, and can be asymmetric. Focal epileptic seizures originate "within networks limited to one hemisphere" and can originate in subcortical structures, can be localized or widely distributed, and have a consistent site of ictal onset (Berg et al., 2010).

6.2 Changes to the ILAE's 1981 classification of seizures

There have been a few changes to the 1981 ILAE classification of seizures. First, neonatal seizures are no longer classified as a separate entity. Secondly, the subclassification of absence seizures was simplified to either typical, atypical, or absence with special features which now included myoclonic absence and eyelid myoclonia. Thirdly, epileptic spasms which were not previously acknowledged in the 1981 seizure classification are now included. It is still unknown whether epileptic spasms are focal in onset, generalized in onset, or both so they are classified as "unknown." Fourth, the distinction between simple partial and complex partial focal seizures was eliminated, however, the concept of impairment of consciousness/awareness is still recognized. Lastly, myoclonic astatic seizures are now termed "myoclonic atonic" seizures. (see Table 5).

6.3 Focal seizures should be described

Focal seizures should be described according to their specific elemental features and sequence of occurrence. The glossary of ictal semiology (Blume, et al., 2001) should be consulted for clearly defined and recommended descriptors. For example, the term "dyscognitive" corresponds to the old term complex partial seizure. (see Table 6).

6.4 Replacing idiopathic, symptomatic, and cryptogenic

The term idiopathic is replaced by the term "genetic." The epilepsy must be a direct result of a known or presumed genetic defect and the seizures a core symptom of the disorder. An example would be Dravet syndrome due to a mutation in the SCN1A mutation. Classifying an epilepsy as genetic does not exclude the possibility that environmental factors may contribute to expression of the disease.

The term symptomatic is replaced by the term "structural/metabolic." To be classified in this category, the epilepsy has to be associated with a structural lesion or metabolic disease that has been shown in previous studies to substantially increase the risk of developing

epilepsy. Examples of structural lesions include stroke, trauma, infection, tuberous sclerosis, and malformations of cortical development.

The term cryptogenic is replaced by the term “unknown cause.” The epilepsies of unknown cause constitute over one-third of all epilepsies. These epilepsies are an area of active current research in the fields of genetics, immunology, and neuroimaging.

Classification of Seizures
Generalized seizures
Tonic-clonic
Absence
Typical
Atypical
Absence with special features
Myoclonic absence
Eyelid myoclonia
Myoclonic
Myoclonic
Myoclonic atonic
Myoclonic tonic
Clonic
Tonic
Atonic
Focal seizures
Unknown
Epileptic spasms

Table 5. The Classification of Seizures, from Berg, A.T., Berkovic, S.F, et. al. (2010). Revised terminology and concepts for organization of seizures and epilepsies: Report of the ILAE Commission on Classification and Terminology, 2005-2009. *Epilepsia*, Vol. 51, No. 4, (April 2010), pp. 676-685, ISSN 1528-1167

Descriptors of focal seizures according to degree of impairment during seizure	Replaces the term
Without impairment of consciousness or awareness	
With observable motor or autonomic components	Simple partial seizure
Involving subjective sensory or psychic phenomena only	Aura
With impairment of consciousness or awareness	Complex partial seizure
Evolving to a bilateral, convulsive seizure	Secondarily generalized seizure

Table 6. Replacement of the terms simple partial, complex partial, and secondarily generalized seizures, from Berg, A.T., Berkovic, S.F, et. al. (2010). Revised terminology and concepts for organization of seizures and epilepsies: Report of the ILAE Commission on Classification and Terminology, 2005-2009. *Epilepsia*, Vol. 51, No. 4, (April 2010), pp. 676-685, ISSN 1528-1167

6.5 Disease-syndrome groupings

Instead of using the terms “disease” or “syndrome” to classify the epilepsies, four distinct groupings were developed: electroclinical syndromes, constellations, structural/metabolic epilepsies, and epilepsies of unknown cause (previously termed cryptogenic). The electroclinical syndromes are defined as “a complex of clinical features, signs, and symptoms that together define a distinctive, recognizable clinical disorder.” They are defined on the basis of age of onset, specific EEG findings, seizure types, and other features which combine to form a specific diagnosis. Constellations are groups of epilepsies defined by diagnostically meaningful specific lesions or other causes which often have treatment implications. Examples of constellations are: mesial temporal epilepsy with hippocampal sclerosis, gelastic seizures and hypothalamic hamartoma, epilepsy with hemiconvulsion and hemiplegia, and Rasmussen syndrome. Regarding epilepsies due to structural or metabolic causes, more emphasis should be given to the seizure etiology rather than localization because the structural or metabolic etiology lends to a better understanding of prognosis. Based on these new concepts, instead of defining an epilepsy as “symptomatic temporal lobe epilepsy” the new terminology would read “epilepsy with focal seizures secondary to a cavernous angioma in the temporal lobe.” (See Table 7).

Electroclinical syndromes and other epilepsies
Electroclinical syndromes arranged by age at onset
Neonatal period
Benign familial neonatal epilepsy
Early myoclonic encephalopathy
Ohtahara syndrome
Infancy
Epilepsy of infancy with migrating focal seizures
West syndrome
Myoclonic epilepsy in infancy
Benign infantile epilepsy
Benign familial infantile epilepsy
Dravet syndrome
Myoclonic encephalopathy in nonprogressive disorders
Childhood
Febrile seizures plus
Panayiotopoulos syndrome
Epilepsy with myoclonic atonic (previously astatic) seizures
Benign epilepsy with centrotemporal spikes
Autosomal-dominant nocturnal frontal lobe epilepsy
Late onset childhood occipital epilepsy (Gastaut type)
Epilepsy with myoclonic absences
Lennox-Gastaut syndrome
Epileptic Encephalopathy with continuous spike-and-wave during sleep (CSWS)
Landau-Kleffner syndrome
Childhood absence epilepsy
Adolescence-Adult
Juvenile absence epilepsy

Juvenile myoclonic epilepsy
Epilepsy with generalized tonic-clonic seizures alone
Progressive myoclonus epilepsies
Autosomal dominant epilepsy with auditory features
Other familial temporal lobe epilepsies
Less specific age relationship
Familial focal epilepsy with variable foci
Reflex epilepsies
Distinctive constellations
Mesial temporal lobe epilepsy with hippocampal sclerosis
Rasmussen syndrome
Gelastic seizures with hypothalamic hamartoma
Hemiconvulsion-hemiplegia-epilepsy
Epilepsies attributed to and organized by structural-metabolic causes
Malformations of cortical development
Neurocutaneous syndromes (tuberous sclerosis complex, Sturge-Weber, etc.)
Tumor
Infection
Trauma
Angioma
Perinatal insults
Stroke
Etc.
Epilepsies of unknown cause
Conditions with epileptic seizures that are traditionally not diagnosed as a form of epilepsy per se
Benign neonatal seizures
Febrile seizures

Table 7. Electroclinical syndromes and other epilepsies, adapted from Berg, A.T., Berkovic, S.F, et. al. (2010). Revised terminology and concepts for organization of seizures and epilepsies: Report of the ILAE Commission on Classification and Terminology, 2005-2009. *Epilepsia*, Vol. 51, No. 4, (April 2010), pp. 676-685, ISSN 1528-1167

6.6 Pros and cons of the new ILAE classification (2010)

The new classification of seizures and epilepsy syndromes is both an update of the old and a radical restructuring of it. It is a work in progress and it remains to be seen whether this new classification will come to acceptance or require further revision. The older classifications were simplified and specific seizure types were added. The replacement of the term "idiopathic" by the term "genetic" may be problematic. First, genetic etiologies due to a single gene mutations are still rare. Secondly, many of the epilepsies may be defined by multiple gene mutations and not due to a specific genetic defect. Additionally, some of the epilepsies don't fit into a single category in the new classification system. An example would be the age-specific epilepsies, such as the primarily generalized epilepsies. Overall however, the changes to the classifications needed to be made. Although it may be difficult to come to a consensus and there will be disagreement in the future, revisiting the way we classify seizures and epilepsy is a step forward for the field. (Shinnar, 2010).

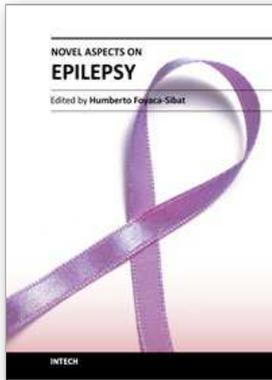
7. Conclusion

The current ILAE Classification System for seizures and the epilepsies has formed the basis for a worldwide standardized approach to diagnosing, treating, and studying seizure disorders. The seizure classification system is primarily based on clinical semiology and EEG correlation, with a major distinction made between focal and generalized seizures. Focal seizures are further subdivided into simple and complex partial seizures, with the presence or absence of impairment of consciousness distinguishing the two. Generalized seizures are divided into absence, tonic, tonic-clonic, myoclonic, or atonic seizures. The epilepsy classification system highlights specific syndromes defined from anatomic-pathological bases (mesial temporal lobe epilepsy with hippocampal sclerosis) to electroclinical bases (Lennox-Gastaut Syndrome). This system has been useful for both clinicians and researchers over the past 30 years, but new data from modern neuroimaging techniques, molecular biology studies, and genetics research has revealed the limitations of the 1981 and 1989 Classification systems. The 2010 ILAE report sets forth new concepts and terminology, with an emphasis on reducing the dichotomy between focal and generalized epilepsies. The 2010 ILAE report did not propose a new classification system, but noted its recommendations will likely be a precursor to a substantive revision of the current classification system in the near future.

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This book covers novel aspects of epilepsy without ignoring its foundation and therefore, apart from the classic issues that cannot be missing in any book about epilepsy, we introduced novel aspects related with epilepsy and neurocysticercosis as a leading cause of epilepsy in developing countries. We are looking forward with confidence and pride in the vital role that this book has to play for a new vision and mission. Therefore, we introduce novel aspects of epilepsy related to its impact on reproductive functions, oral health and epilepsy secondary to tuberous sclerosis, mitochondrial disorders and lisosomal storage disorders.

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