

INSIGHTS IN CLASSIFICATION OF SEIZURES AND EPILEPSIES – REVIEW OF KEY CONCEPTS THROUGHOUT HISTORY

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ABSTRACT

Since the Babylonian era up to nowadays, the large complexity of epileptic disorders made the attempt of classifying and organizing concepts one of the most challenging.

The 1981 classification was a revision of the one devised by Henri Gastaut for the ILAE and published in 1970. The distinction between simple and complex partial seizures represented a significant difference.

In 1989 the report of ILAE classification and terminology task force introduced the concept of epileptic syndrome. An epileptic syndrome is defined by the sum of signs and symptoms that tend to occur together, not reflecting a particular etiology and prognosis, these latter features being the hallmark not of a syndrome, but of a disease condition. Firstly, any epileptic disorder can be described as either idiopathic (primary, whose etiology is probably genetic), symptomatic (with a known or presumed cerebral pathology) or cryptogenic (with an occult cause). The epileptic diagnostic tree ramifies into four main branches in the 1989 ILAE Classification: localization-related (focal, local, or partial) epilepsies and syndromes, generalized epilepsies and syndromes, epilepsies and syndromes undetermined whether focal or generalized and special syndromes.

The next important milestone in epilepsy classification was settled by ILAE in 2001, when a glossary for ictal semiology was developed and multiaxial approach of diagnosing epilepsy was proposed.

The necessity of processing the new insights in epilepsy was answered by the latest ILAE Report released in february 2010, which brings awareness of and appropriately integrates the currently available data. It presents a modular approach of diagnosing epilepsy and recommends to keep a flexible vision on this topic, as every individual case requires a different prioritizing scheme.

Key words: seizure, epileptic syndrome, epilepsy, classification

INTRODUCTION

The vast complexity of epilepsy spectrum makes the task of organizing epileptic disorders a challenging one. Periodically, International League against Epilepsy reviews the growing body of evidence in the epilepsy field and makes suggestions regarding a more accurate approach in understanding the epilepsy subtypes. For instance, as new genetic data became available through the years, particular nosologic entities gained the signature of a particular gene defect, which consequently imposed their withdrawal from former vague descriptive

categories. Also, since 1970, when the first ILAE harmonized attempt of creating taxonomy for epilepsy was made, a careful long-term observation of electroclinical evolutive patterns has enabled a better perspective for some epileptic disorders. More refined imaging tools were developed over the decades, they being invaluable in identifying some structural lesions as the cause of seizures, fact that claimed sometimes a lesion-tailored diagnosis in the first place. The necessity of processing the new insights in epilepsy was answered by the latest ILAE Report released in February 2010, which brings awareness of and appropriately integrates

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the currently available data. It presents a modular approach of diagnosing epilepsy, precludes the use of some obsolete terms, and emphasizes the necessity of keeping a flexible vision on this topic, as every individual case requires a different prioritizing scheme.

BRIEF HISTORY OF EPILEPSY CLASSIFICATION

The most ancient classification of seizures has been mentioned in Babylon during the reign of the Babylonian king of the Second Dynasty of Isin (1067-1046 BC.), where the medical knowledge was carved into stone tablets known as the *Sakikku* (meaning *All Diseases*). Many types of seizures were described, each attributed to a certain demon or spirit and a given prognosis was presumed for most of them (1).

There are several relevant milestones that deserve a special attention in the development of epilepsy classification.

The beginning of international efforts of organizing epilepsy may be traced back in 1970, when ILAE issued the first international classification of epileptic seizures (2). The seizures were divided into:

- partial seizures or seizures beginning locally: partial seizures with elementary symptomatology (generally without altered consciousness), partial seizures with complex symptomatology (with impaired consciousness) and partial seizures secondarily generalized
- generalized seizures, bilateral symmetrical seizures or seizures without local onset: absences, bilateral massive epileptic myoclonus, infantile spasms, clonic seizures, tonic seizures, tonic-clonic seizures, atonic seizures, akinetic seizures
- unilateral or predominantly unilateral seizures
- unclassified seizures

The above taxonomy was reviewed several times before 1981, when another important step was taken by the ILAE Commission on Classification and Terminology, which then published a renewed classification of epileptic seizures based on clinical and electrographic grounds (3). This gained a worldwide acceptance, being the foundation of epileptic seizure description as we understand today.

The crucial dichotomy between partial (focal, local) and generalized (convulsive or nonconvulsive) seizures was again highlighted in the 1981

ILAE Report. Partial seizures, defined by the initial activation of neurons limited to one cerebral hemisphere, are further subdivided based on the state of consciousness in simple, when consciousness is preserved, and complex, when there is an alteration of the consciousness state. The partial seizures evolving to generalized tonic-clonic convulsions are delineated as a separate subcategory. The simple partial seizures can take the following forms, based on the clinical picture: motor, somatosensory and special sensory, autonomic or psychic. The complex partial seizures may be either with a simple partial onset, or starting with impairment of consciousness. Generalized seizures are defined by the involvement of both cerebral hemispheres from the beginning, as shown by the bilateralism of clinical manifestations and ictal electroencephalographic patterns. They are further subdivided into: absence, myoclonic, clonic, tonic, tonic-clonic and atonic seizures. Besides partial and generalized seizures, a third category is mentioned – “unclassified seizures” – for those that cannot be included in other categories due to insufficient data.

Another important step in the development of epilepsy classification was taken in 1989, when ILAE launched another proposal for classification of epilepsies and epilepsy syndromes (4). An epileptic syndrome is defined by the sum of signs and symptoms that tend to occur together, not reflecting a particular etiology and prognosis – these latter features being the hallmark not of a syndrome, but of a disease condition. Firstly, any epileptic disorder can be described as either idiopathic (primary, whose etiology is probably genetic), symptomatic (with a known or presumed cerebral pathology) or cryptogenic (with an occult cause).

The epileptic diagnostic tree ramifies into four main branches in the 1989 ILAE Classification:

- localization-related (focal, local, or partial) epilepsies and syndromes
- generalized epilepsies and syndromes
- epilepsies and syndromes undetermined whether focal or generalized
- special syndromes

Localization-related epilepsies can be either idiopathic (benign childhood epilepsy with centrotemporal spikes, childhood epilepsy with occipital paroxysms, primary reading epilepsy), 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 and occipital lobe epilepsies) or cryptogenic.

Generalized epilepsies and syndromes are also subdivided into 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, epilepsies with seizures precipitated by specific modes of activation), symptomatic (with a non-specific etiology – early myoclonic encephalopathy, early infantile epileptic encephalopathy with suppression burst, other symptomatic generalized and specific syndromes – diseases in which seizures are a presenting or predominant feature) and cryptogenic/symptomatic (West syndrome, Lennox-Gastaut syndrome, epilepsy with myoclonic-astatic seizures, epilepsy with myoclonic absences).

Epilepsies and syndromes undetermined whether focal or generalized have two main subcategories: 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) and without unequivocal generalized or focal features (sleep related GTCS, when focal or generalized onset cannot be determined by EEG or clinical findings).

Special syndromes refer to 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).

The next important milestone in epilepsy classification was settled by ILAE in 2001, when a glossary for ictal semiology was developed (5) and multiaxial approach of diagnosing epilepsy was proposed (6). The previous classifications of epileptic seizures and syndromes issued in 1981 and 1989 were generally kept as still valid, but changes were made when more recent data imposed more accuracy (e.g. creating a separate category for autosomal dominant epilepsies).

The categories of epileptic syndromes recognized in the 2001 ILAE Report 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.

There are five recommended axes to follow when diagnosing an epileptic condition:

- axis 1: a concrete presentation of the ictal clinical picture, based on the Glossary of Descriptive Ictal Terminology issued in 2001
- axis 2: type of seizure
- axis 3: the syndrome diagnosis, when possible
- axis 4: etiology, when possible (e.g. genetic, structural lesions)
- axis 5: the degree of impairment

Of note, in 2001 the concept of a flexible approach when diagnosing epilepsy is introduced – it should be kept in mind that a detailed picture of seizures is not always needed, the type of seizures may change when new data is obtained and not all cases can be integrated in a particular syndrome. Also, the diagnostic scheme may be adjusted according to a specific utility: teaching, clinical trials, basic research, genetic studies or epilepsy surgery.

Several important novelties are also brought by the 2006 ILAE Report (7). An updated definition for epileptic seizures is presented – seizure is “a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain”. An epileptic seizure has to be seen as an entity delineated by a particular pathophysiology (for instance, the increased neuronal excitability and decreased inhibition for generalized tonic-clonic seizures) and anatomical substrate (neocortex, thalamic reticular nucleus, and brainstem).

Different ways of categorizing epileptic seizures were proposed in 2006. Firstly, they were divided into self-limited seizure types and status epilepticus.

Self-limited seizure types have the following categories: with generalized onset (with tonic and/or clonic components, absences, myoclonic seizure types, epileptic spasms, atonic seizures), with focal onset (local – neocortical or hippocampal & parahippocampal; with ipsilateral spread to neocortex or limbic areas; with contralateral spread to neocortex or limbic areas; secondarily generalized) and neonatal seizures.

The recognized subtypes of status epilepticus are: *epilepsia partialis continua* of Kojevnikov, supplementary motor area status epilepticus, *aura continua*, *dyscognitive focal (psychomotor, complex partial) status epilepticus*, *tonic-clonic status epilepticus*, *absence status epilepticus*, *myoclonic status epilepticus*, *tonic status epilepticus* and *subtle status epilepticus*.

In the 2006 ILAE Report, it is recommended for the epileptic syndromes (listed in the previous ILAE Report in 2001) to be evaluated by covering several facets: epileptic seizure type, age of onset, progressivity of the epileptic disorder, relevant interictal EEG, interictal clinical manifestations, pathophysiology, anatomical substrate, etiology and genetic component.

From the above possibilities, age of onset was selected for a new ordering, and the list of epileptic syndromes and related conditions issued in the 2006 ILAE Report is presented as follows:

- in the neonatal period: benign familial neonatal seizures, early myoclonic encephalopathy, Ohtahara syndrome
- in infancy: migrating partial seizures of infancy, West syndrome, myoclonic epilepsy in infancy, benign infantile seizures, Dravet syndrome, myoclonic encephalopathy in nonprogressive disorders
- in childhood: early onset benign childhood occipital epilepsy (Panayiotopoulos type), epilepsy with myoclonic astatic seizures, benign childhood epilepsy with centrotemporal spikes, late onset childhood occipital epilepsy (Gastaut type), epilepsy with myoclonic absences, Lennox-Gastaut syndrome, epileptic encephalopathy with continuous spike-and-wave during sleep including Landau-Kleffner syndrome, childhood absence epilepsy
- in adolescence: juvenile absence epilepsy, juvenile myoclonic epilepsy, progressive myoclonus epilepsies
- less specific age relationship: autosomal-dominant nocturnal frontal lobe epilepsy, familial temporal lobe epilepsies, mesial temporal lobe epilepsy with hippocampal sclerosis, Rasmussen syndrome, gelastic seizures with hypothalamic hamartoma
- special epilepsy conditions: symptomatic focal epilepsies not otherwise specified, epilepsy with generalized tonic-clonic seizures only, reflex epilepsies, febrile seizures plus, familial focal epilepsy with variable foci
- conditions with epileptic seizures that do not require a diagnosis of epilepsy: benign neonatal seizures, febrile seizures.

In the 2006 ILAE Report, it is emphasized again that the presented scheme is not intended to be an autocratic classification, but a discussion opened to future epilepsy classifications.

MAIN CHANGES INTRODUCED BY THE NEWEST ILAE REPORT

Modern advances in neuroimaging and genomic research and application in clinical practice in the last decades changed the way we understand some of the fundamental concepts regarding classification of seizures and epilepsies. After repeated, controversial attempts of ILAE Committee or personalities in the field (9), a recent, comprehensive review and update of classification and terminology for seizures and epilepsy syndromes has been issued in 2010. In fact, the new ILAE report is not actually a classification scheme but rather a discussion of novel concepts that impact our understanding of clinical features, prognostic implication, and eligibility for surgery, standardization of data in clinical trials and for research purpose.

One of the key concepts that have been reviewed was the definition of generalized epileptic seizures “as originating at some point within, and rapidly engaging, bilaterally distributed networks”. The bilateral networks involved “can include cortical and subcortical structures, but do not necessarily include the entire cortex” as well as “individual seizure onsets can appear localized but the location and lateralization are not consistent from one seizure to another. Generalized seizures can be asymmetric.”

Regarding the focal seizures, their description is recommended to take into account clinical features that are considered relevant for a given specific purpose. For example: when we talk about driving permission, the most relevant clinical feature is the impairment of consciousness but if we discuss the eligibility for surgery, aura and the detailed sequence of symptoms and signs are at utmost importance. The 2001 version of the glossary of ictal semiology⁵ (see Table 1) represents the recommended reference for seizure’s clinical descriptors. Instead of classifying focal seizures in simple or complex, the ILAE report encourages to mention whether the consciousness or awareness are impaired or not and to use the term “dyscognitive” instead of complex partial seizure. (Table 2)

In the **seizure’s classification** are now recognized some categories that have been disregarded or controversial in the previous reports.

Seizures occurring in the neonatal period are classified similar to the seizures occurring in any other period of life. There are no reasons to advocate for a separate entity.

The classification of absence seizures now recognizes myoclonic absence seizures and eyelid myoclonia.

Table 1. Recommended terminology for seizure's description (Blume et al., 2001)

Motor	Elementary motor	Tonic A sustained increase in muscle contraction	Epileptic spasm – a sudden flexion, extension or mixed extension-flexion of predominantly proximal and truncal muscles, that is usually more sustained than a myoclonic movement, but not so sustained as a tonic seizure. Postural – adoption of a posture that may be bilaterally symmetric or asymmetric.
		Myoclonic Sudden, brief (<100 ms) involuntary single or multiple contractions of muscles or muscle groups of variable topography.	Negative myoclonic – an interruption of tonic muscular activity for <500 ms without evidence of preceding myoclonia. Clonic – myoclonus that is regularly repetitive, involves the same muscle groups, at a frequency of ~2–3 c/s, and is prolonged. Synonym: rhythmic myoclonus.
		Tonic-clonic	A sequence consisting of a tonic followed by a clonic phase.
		Atonic	Sudden loss or diminution of muscle tone without apparent preceding myoclonic or tonic event lasting ≥1 to 2 s, involving head, trunk, jaw, or limb musculature.
		Astatic	Loss of erect posture that results from an atonic, myoclonic, or tonic mechanism. Synonym: drop attack.
Automatism	Oroalimentary	Lip smacking, lip pursing, chewing, licking or swallowing.	
	Mimetic	Facial expression suggesting an emotional state, often fear.	
	Manual or pedal	1 Indicates principally distal components, bilateral or unilateral 2 Fumbling, tapping, manipulating movements.	
	Gestural	Fumbling or exploratory movements with the hand, directed toward self or environment.	
	Hyperkinetic	1 Involves predominantly proximal limb or axial muscles producing irregular sequential ballistic movements, such as pedaling, pelvic thrusting, thrashing, rocking movements. 2 Increase in rate of ongoing movements or inappropriately rapid performance of a movement.	
	Hypokinetic	A decrease in amplitude and/or rate or arrest of ongoing motor activity.	
	Dysphasic	Impaired communication involving language without dysfunction of relevant primary motor or sensory pathways, manifested as impaired comprehension, anomia, paraphasic errors, or a combination of these	
	Dyspraxic	Inability to perform learned movements spontaneously or on command or imitation despite intact relevant motor and sensory systems and adequate comprehension and cooperation.	
	Gelastic	Bursts of laughter or giggling, usually without an appropriate affective tone.	
	Dacrystic	Bursts of crying.	
	Vocal	Single or repetitive utterances consisting of sounds such as grunts or shrieks.	
	Verbal	Single or repetitive utterances consisting of words, phrases, or brief sentences.	
	Spontaneous	Stereotyped, involve only self, virtually independent of environmental influences.	
Interactive	Not stereotyped, involve more than self, environmentally influenced.		

Non-motor	Aura	A subjective ictal phenomenon that, in a given patient, may precede an observable seizure; if alone, constitutes a sensory seizure.	
	Sensory	Elementary	Somatosensory – Tingling, numbness, electric-shock sensation, pain, sense of movement, or desire to move.
			Visual – Flashing or flickering lights, spots, simple patterns, scotoma, or amaurosis.
			Auditory – Buzzing, drumming sounds or single tones.
			Olfactory – Odor, usually disagreeable.
			Gustatory – Taste sensations including acidic, bitter, salty, sweet, or metallic.
			Epigastric – Abdominal discomfort including nausea, emptiness, tightness, churning, butterflies, malaise, pain, and hunger; sensation may rise to chest or throat. Some phenomena may reflect ictal autonomic dysfunction.
			Cephalic – Sensation in the head such as light-headedness, tingling or headache.
			Autonomic – A sensation consistent with involvement of the autonomic nervous system, including cardiovascular, gastrointestinal, sudomotor, vasomotor, and thermoregulatory functions.
	Experiential		Affective – Components include fear, depression, joy, and (rarely) anger.
			Mnemonic – Components that reflect ictal dysmnesia such as feelings of familiarity (déjà-vu) and unfamiliarity (jamais-vu).
			Hallucinatory – A creation of composite perceptions without corresponding external stimuli involving visual, auditory, somatosensory, olfactory, and/ or gustatory phenomena. Example: “hearing” and “seeing” people talking.
			Illusory – An alteration of actual percepts involving the visual, auditory, somatosensory, olfactory, or gustatory systems.
			perception: symbolic conception of sensory information attention: appropriate selection of a principal perception or task emotion: appropriate affective significance of a perception memory: ability to store and retrieve percepts or concepts executive function: anticipation, selection, monitoring of consequences, and initiation of motor activity including praxis, speech
Autonomic events	Autonomic aura	A sensation consistent with involvement of the autonomic nervous system, including cardiovascular, gastrointestinal, sudomotor, vasomotor, and thermoregulatory functions	
	Autonomic seizure	A documented and distinct alteration of autonomic nervous system function involving cardiovascular, pupillary, gastrointestinal, sudomotor and vasomotor functions.	

Table 2. Classification of seizures 1970-2010

1970	Partial seizures with elementary symptomatology (generally without altered consciousness), with complex symptomatology (with impaired consciousness) secondarily generalized	Generalized seizures , bilateral symmetrical seizures (or seizures without local onset): absences, bilateral massive epileptic myoclonus, infantile spasms, clonic, tonic, tonic-clonic seizures, atonic seizures, akinetic seizures	Unilateral or predominantly unilateral seizures	Unclassified seizures
1981	Partial seizures (initial activation of neurons limited to one cerebral hemisphere) simple (when consciousness is preserved): motor, somatosensory and special sensory, autonomic or psychic complex (when there is an alteration of the consciousness state) – with a simple partial onset or starting with impairment of consciousness. partial seizures evolving to generalized tonic-clonic convulsions	Generalized seizures (involvement of both cerebral hemispheres from the beginning, as shown by the bilaterality of clinical manifestations and ictal electroencephalographic patterns): – absence, – myoclonic, – clonic, – tonic, – tonic-clonic – atonic seizures.		Unclassified seizures Insufficient data
2010	Focal seizures <i>Without impairment of consciousness or awareness</i> With observable motor or autonomic components. Involving subjective sensory or psychic phenomena only. <i>With impairment of consciousness or awareness.</i> <i>Evolving to a bilateral, convulsive seizure</i> (involving tonic, clonic, or tonic and clonic components).	Generalized seizures (originating at some point within, and rapidly engaging, bilaterally distributed networks). Tonic-clonic (in any combination) Absence Typical Atypical Absence with special features Myoclonic absence Eyelid myoclonia Myoclonic Myoclonic Myoclonic atonic Myoclonic tonic Clonic Tonic Atonic	–	Unknown Epileptic spasms

Epileptic spasms are now acknowledged as a group of their own, as there is evidence they may continue from past infancy, or even occur “de novo”. Still there is not enough evidence accumulated to help classify them as generalized or focal.

The new classification also recognized myoclonic atonic seizures, previously named myoclonic atonic.

Concerning the **etiology**, the terms used in the old classification (idiopathic, symptomatic and cryptogenic) were replaced by three other terms:

- **Genetic:** The term refers to an epileptic disorder that is the direct result of a known or presumed genetic defect and seizures are the main clinical manifestation. The basis for labeling epilepsy as being genetic may come either from molecular studies (e.g. Dravet syndrome), or from appropriately designed family studies. Environmental factors might

influence the clinical expression of a certain disorder.

- **Structural/metabolic:** In this case, the underlying cause of the epileptic disorder is a distinct structural or metabolic disease or condition, proven to be associated with an increased risk of developing epilepsy in appropriately designed studies. Structural lesions include stroke, trauma, and infection or may be of genetic origin (tuberous sclerosis or malformations of cortical development). Localization is no longer considered a primordial factor for treatment options and outcome prediction. As a consequence, the diagnostic formulation will emphasize the type of lesional substrate more than the seizure focus localization.
- **Unknown cause:** this category is meant to designate the cause leading to seizures as yet

unknown. Unlike cryptogenic (presumed symptomatic), it makes no presumptions and requires no explanation or reinterpretation.

Even though the concepts of disease and syndrome are still recognized, it was decided not to insist on the disease-syndrome distinction when talking about epilepsies. Instead, it is recommended to use the categories described below.

Electroclinical syndrome: consist in a group of clinical entities that can be reliably identified by a number of electroclinical characteristics “a complex of clinical features, signs, and symptoms that together define a distinctive, recognizable clinical disorder.” The distinction between different electroclinical syndromes is based on age of onset, EEG findings, seizure types, effect on cognition and behavior and other features that help treatment indication and prognostic prediction. The distinction between focal and generalized could not be applied for this category.

Constellations: This refers to a number of entities that are not exactly electroclinical syndromes, but are distinctive on the basis of specific lesions or other causes. The main implication is in epilepsy surgery. Examples of constellations include mesial temporal lobe epilepsy with hippocampal sclerosis (10), hypothalamic hamartoma with gelastic seizures, and epilepsy with hemiconvulsion and hemiplegia and Rasmussen syndrome.

Structural/metabolic epilepsies: The group includes epilepsies secondary to specific structural or metabolic lesions or conditions, but which do not, given our current understanding, fit a specific electroclinical pattern. They represent a lower level of specificity than the two previous groups.

Epilepsies of unknown cause: This is the new terminology describing epilepsies of unknown cause, instead of cryptogenic.

According to the purpose we use these categories, flexibility in further organization within these particular divisions is recommended in the new report. For example, epileptic encephalopathy is a concept recognized in the previous classification as well and is now defined as an electroclinical syndrome with cognitive and behavioral impairments aggravated by epileptic activity, beyond what might be expected from the underlying pathology alone. The impairments may be global or more selective. The encephalopathic effects of seizures can occur in association with any form of epilepsy. The label of “encephalopathic course” for an epileptic disorder is mainly based on documenting the cognitive decline or hampered development of these abilities relative to same-aged peers. This has considerable

implications for encouraging seizure control as early as possible and for recognizing the cognitive impact of seizures and epileptic activity regardless patient’s age.

Another concept reconsidered in the new report refers to the group of so called “benign epilepsies”. The term benign is no longer appropriate to describe the course and nature of an epileptic disorder and is replaced by “self limited” emphasizing the high probability to get into remission of seizures at a certain age.

RELEVANT CLINICAL EXAMPLES ILLUSTRATING THE APPLICATION OF DIAGNOSTIC CHANGES

Case 1

Right handed young women, 28 years of age. History of seizures since 11 years of age.

No significant personal antecedents or hereditary. Normal birth and development.

Seizure’s description:

Somatosensory aura with tingling over the left hand on the dorsal aspect and sometimes perioral as well followed by repetitive blinking, speech arrest, loss of contact, dystonic posture in the left upper limb, hyperkinetic automatisms in the right upper and lower limbs.

Postictal: rapid recovery, no functional deficits.

Seizure’s duration: 30-40 seconds

Neurological and neuropsychological examination interictal with normal findings.

Cerebral MRI 1.5 T showing a dysplastic lesion in the right suprasylvian operculum centro-parietal.





Figure 1. Patient does not answer to the examiner's questions

Seizure's classification:

Focal seizures with impairment of consciousness and observable motor component.

Diagnostic conclusion:

Epilepsy with focal seizures secondary to cortical dysplasia in the suprasylvian operculum.

Comments: some of the clinical features are acknowledged only if the patient is appropriately tested during the ictal period (see Figure 2).

Case 2

Right handed woman, 37 years of age. Febrile seizures during early childhood.



Figure 2. Patient not following 5 commands

Seizure's description:

Epigastric aura then she is experiencing a complex visual hallucination, manifested as a familiar scene from a movie. The patient is still able to talk and interact with the examiner during the whole ictal event. Appropriate testing does not reveal language or contact impairment during the seizure. If the patient had been involved in some activity before the seizure's onset than ictal activity arrest would have been obvious. Otherwise we can only say that the seizure is evolving without observable motor component.

Neuropsychological examination reveals a moderate deficit in completion of nonverbal memory tasks.



Figure 3. 1.5 TMRI shows right hippocampal sclerosis

Seizure's classification:

Focal seizures without impairment of consciousness or awareness involving only subjective and psychic phenomena.



Figure 4. Patient naming correctly the object that is presented by the technician

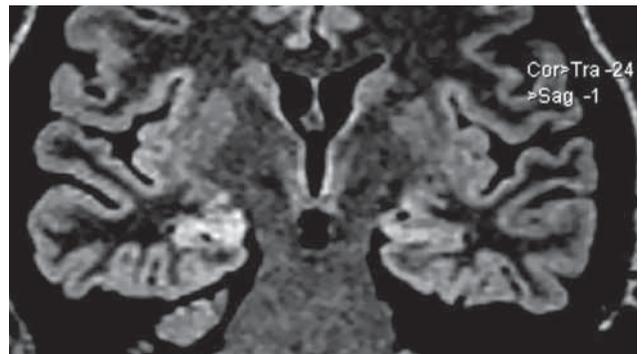


Figure 5. Right hippocampal sclerosis (MRI 1.5T)

Diagnosis of epilepsy:

Epilepsy with focal seizures secondary to right mesiotemporal sclerosis

CONCLUSION

Epileptology is a dynamic field with concepts adapted periodically according to the progress of knowledge in basic neuroscience and clinical practice.

The major changes in the new ILAE report issued in 2010 concern:

Definition of generalized seizures is revised and classification is simplified.

Classification of focal seizures is replaced with the recommendation to use specific terminology that reflect the clinical manifestations.

Diagnostic formulation that emphasize the presumed cause: genetic, structural/metabolic and unknown replaced idiopathic, symptomatic and respectively cryptogenic.

The label of “electroclinical syndrome” was restricted to certain entities and no longer applied to all the epilepsies.

No changes were applied to the list of “syndromes” already recognized and in use since 2006.

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