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# Up-to-date Critical Review of the Classification of Epilepsies and Epileptic Seizures

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The classification of epileptic seizures and epilepsies is a subject of interest in various medical disciplines (such as neurology, pediatric neurology, molecular biology and genetics, neurosurgery, pharmacology, radiology, histopathology), and each of them requires a different approach in their practice. In last 15 years, enormous amount of debate in which irrelevant to actual level of knowledge, were ongoing in the literature. Epilepsy classification is a fundamental tool that impacts not only daily clinical practice but also research era and education. The current lack of consensus in this field causes a serious obstacle in patient management, student and resident education, and information sharing among different scientific interest groups. The comparison of different classification proposals by means of positive and negative aspects is beyond the scope of discussion in this article; therefore, I will try to give a brief summary of our current level of understanding.

Main issues regarding the classifications proposal are as follows:

1. Concepts of epileptic seizure/epilepsy/syndrome
2. Focal & generalized epilepsy concept
   a. Idiopathic, genetic, cryptogenic, and symptomatic (structural/metabolic) concepts

**Main Differences Between Epileptic Seizure/Epilepsy/Syndrome Classification**

**Epileptic seizure** is a transient occurrence of signs and/or symptoms because of abnormal, excessive, and synchronous discharges of brain neurons. The clinical signs depend not only on the origin of the electrical activity but also on the amplitude, speed, and spreading pathways of the discharges. The diagnosis of the seizure relies on observing the event or watching a video recording of the event or noting down the history of an eye witness.

**Epilepsy** is a disorder of the brain, characterized by an enduring predisposition to generate epileptic seizures and by the neurobiologic, cognitive, psychological, and social consequences of this condition. The underlying etiology (such as genetic, tumoral, and vascular diseases; immune mechanisms trauma; environmental toxins; infections; neurodegenerative disease) is highly heterogeneous and multifactorial, and the prognosis highly depends on the underlying etiology.

**Syndrome** is a collection of signs and symptoms that occur together. These include items such as seizure type, etiology, anatomy, precipitating factors, age of onset, severity, chronicity, diurnal and circadian cycling, and sometimes prognosis. In contrast to a disease, there is no single etiology and/or pathology. Therefore, contrary to epileptic seizure, the diagnosis of epilepsy or syndrome is not possible by only observing and/or watching the video but requires information on the age of onset, family history, predisposing factors, frequency of the attack, electroencephalography (EEG) and neuroimaging data, etc.

**Why Syndromic Diagnosis is Important?**

Syndromic diagnosis is necessary for prognostication. For example, while evaluating a patient who has right-sided hemiparesis, a neurologist should spend all his/her effort to determine the underlying etiology. For instance, the right-sided hemiparesis could be due to a brain infarction, hemorrhage, or a tumor. The treatment should not be targeted to hemiparesis but to the underlying etiology, and in this case it is obviously quite different than each other. Thus, by adopting this practice, the doctor can provide the necessary intervention and determine the prognosis of the situation.

**Why Seizure Diagnosis is Important?**

**1. Drug Choice**

As an example of routine daily practice, consider the case of a patient presenting with a generalized tonic clonic seizure (GTCS) and/or dialeptic seizure, in which the only manifestation is a loss of awareness and unresponsiveness. This particular patient could have

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**Table 1.** Seizure/syndrome/drug choice

| Seizure-aggravated drugs | Seizure-specific drugs |
|---|---|
| 1. "Dialeptic" seizure in absence epilepsy → CBZ, OXC, PTH, VGB, GBP, PGB | 1. Epileptic spasm → VGB, ACTH |
| 2. "Myoclonic" seizure in generalized epilepsy syndromes → CBZ, PTH | 2. "Dialeptic" seizure in absence epilepsy → ESM, VPA, LTG |
| 3. Lennox–Gastaut Syndrome;<br>"Tonic" seizures → BZD, PB<br>"Atypical absence" → CBZ, GBP, PB | 3. Myoclonic" seizure in generalized epilepsy syndromes → VPA, LEV, BZD, ZNS |
| | 4. Lennox–Gastaut syndrome "Atonic" seizures → vagal nerve stimulation, RFN |

CBZ: carbamazepine; OXC: oxcarbazepine; PTH: phenytoin; VGB: vigabatrin; GBP: gabapentin; PGB: pregabalin; BZD: benzodiazepine; PB: phenobarbital; ESM: ethosuximide; ZNS: zonisamide; LEV: levetiracetam; RFN: rufinamide

been suffering from three completely different epileptic syndromes, such as juvenile myoclonic epilepsy (JME), frontal lobe epilepsy (FLE), or mesial temporal lobe epilepsy with hippocampal sclerosis (MTLE). Obviously, each of them has its own peculiar EEG findings or concomitant other seizure type or types, genetic tendency, and finally a prognosis. The drug of first choice could also be completely different. All patients with focal epilepsy are treated with more or less the same type of antiepileptic drugs. Patients with generalized epilepsy, however, respond differently to different antiepileptic drugs, mainly because of the type of the seizures (for example, patients with generalized myoclonic seizures respond differently to different antiepileptic drugs than patients with absence seizures). A brief summary of drug choice is given in Table 1.

### 2. Presurgical Evaluation

One of the important steps of presurgical evaluation is video-EEG monitoring, by which lateralizing and/or localizing features of epileptic seizures could be recorded; thus, it helps in defining the "symptomatogenic zone."

### Focal and Generalized Epilepsy Concepts

### Idiopathic, Genetic, Cryptogenic, and Symptomatic (Structural/Metabolic)

#### Pitfalls of Electroclinical Diagnosis

Recent advances in neuroimaging techniques, widespread use of video-EEG monitoring, and micro-neurosurgical interventions have led to dramatic changes to our understanding on electroclinical syndromes. For example, the seizure type of the West syndrome, which was considered a generalized epilepsy syndrome, was infantile spasms and EEGs showed generalized spike and wave discharges; however, many infants had a causative focal lesion and their epilepsy had been cured by focal resections. Similarly, patients with generalized epilepsies such as JME exhibited bilateral asymmetric tonic seizures or those with JAE exhibited automotor seizures and vice versa.

#### Pitfalls of Genotype–Phenotype Heterogeneity

Recently characterized genetic disorders challenge the distinction between the "pure" genetic epilepsies and structural/metabolic disorders. As a typical example, ARX phenotypes can be responsible either in the West syndrome or in lissencephaly. In contrast, we do not have any particular causative gene or genes in well known, classic electroclinical syndromes such as JAE or JME.

Therefore, abandoning the term of idiopathic and refer to that group in unknown or structural/metabolic frame is not compensate our lack of information.

## CONCLUSION

Modern epileptology has stated that epileptic seizures are the consequences of a basic epileptogenic tendency (genes), various more or less obvious triggering factors, and major causative factors. For any given patient, it is essential that we clearly identify these factors. Unfortunately, today we are far more than that to understand completely to the whole pathogenetic mechanisms for all epilepsies.

In general, classification proposals describe seizures as an epilepsy, in line with our current understanding. By its very nature, epilepsy classification will always be subject to revision as our understanding grows. Interested readers can refer to the relevant literature (1,2).

## REFERENCES

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