

## Comments about the Revised terminology and concepts for organization of the epilepsies: Report of the Commission on Classification and Terminology

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Certainly, the new modifications made in the classification and terminology since the first proposal, are nearer approximations to a system of rules that attempt an explaining theory of epilepsy. But, for a further approximation I think there is a limiting barrier imposed by the model of the brain in use for a very long time. Even more, the current model of disease, the same that explains gastritis pretty well, is not enough to explain a so complex pathological process as epilepsy.

As everybody knows, the model of the brain we use to explain human diseases is the generic mold of the mammals' brain; consciousness is nothing more than sensitivity to a stimulus, awareness of the world, the body, the self. But, all these properties seem to be common to every living creature. Instead, doesn't it sound anything different if I say that only a person is able to be aware by means of his/her consciousness? ¿Can a primate take account of its own image in a mirror through *its* consciousness? If the answer were affirmative, it does mean that *it has* a consciousness? I invite you to think about some fundamental principles like these: a) We, people, are the only human beings born with a neocortical neural network ready to be transformed in a consciousness after they learn and codify in that structure the kinds of information created by society in the course of thousands of years. b) During infancy, childhood and adolescence all this "consolidated" now psychic information –as feelings, knowledge and motivations– becomes a material consciousness that when active and integrated through subcortical networks (hippocampus, amygdala, hypothalamus, thalamus) takes the form of percepts, images, concepts and actions. Finally, c) The whole of this new psychic structure transforms a human being in a *conscious personality*.

On the other hand, wouldn't you expect some changes in the actual theory of epilepsy if I argue that the biomedical model of disease we use since Hippocrates, Virchow, and others, is not enough to explain, not only mental disorders, but neurological defects and morbid disorders of a *personal* brain and nervous system? Doesn't it prompt us to change in some way the explanation and classification of the different and complex processes of any brain or neurological disease, among them epilepsy and its actual manifestation, name it a seizure?

Even though it is not my intention to change your mind with this paradigm of "personal brain" and disease we are developing since 1994, the question is that we have an alternative model of both, brain and disease, to explain in a novel way diseases that affect our unique personal psychic activity. Therefore, I'll try to introduce you a model of the cortex of man, and a model of disease, that might call your attention. My intention is to put in your hands a sort of test to compare the ever used Gall's model with that of ours.

Compare, for example, table 1 and table 2. The first one says "types of cerebral cortex", in general. In this case, localization has to be referred to cortical vs. subcortical, psychic vs. sensory/motor, and so forth. In table 2 we say "types of personal cerebral cortex". In this case we have to talk about two kinds of psychic activity: unconscious (paleocortical: affective and cognitive sensations), subconscious (neocortical: feelings, knowledge and motivations), and a personal

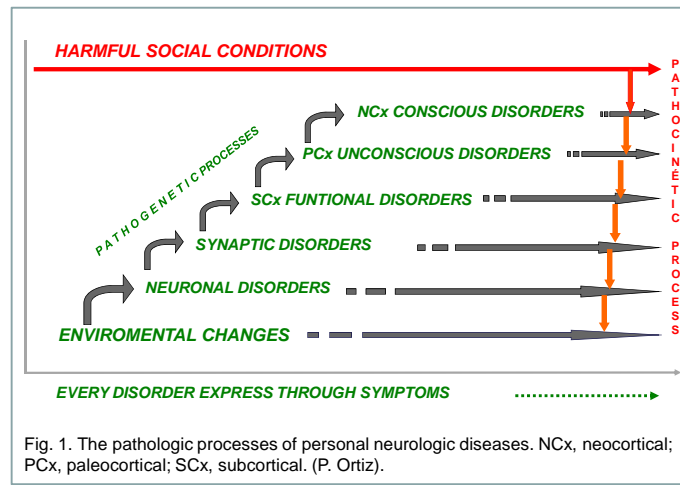
*epiconscious* (a whole brain, nervous system and individual: perception, imagination, thinking and personal behavior, performance and conduct) activity.

TABLE 1 CYTOARCHITECTONIC TYPES OF CEREBRAL CORTEX <i>Bailey y Bonin (1951); Daly (1974)</i>					
	HETEROGENÉTIC		HOMOGENÉTIC		
<b>STRUCTURE</b>	Always with less than 6 layers		With 6 layer at embryonic and/or mature states		
<b>SYNONIM</b>	ALLOCÓRTEX		NEOCÓRTEX		
<b>SUBTYPES</b>	Paleocortex	Archycortex	Homotypic with 6 layers	Heterotypic with less of 6 layers	
	Olfactory area	Hippocampus Dentate gyrus	Eulaminar	Granular	Agranular

TABLE 2 HISTOLOGIC TYPES OF CEREBRAL CORTEX OF PERSONS <i>(Ortiz, 1999, 2004)</i>					
<b>GENESIS</b>	HETEROGENÉTIC		HOMOGENÉTIC		
<b>DESCRIPTION</b>	ALLOCÓRTEX		ISOCÓRTEX		
<b>PHYLLOGENETICS</b>	PALEOCÓRTEX, PALEOPALLIUM		NEOCÓRTEX, NEOPALLIUM		
<b>STRUCTURE</b>	Always with less than 6 layers (at any stage)		Eulaminar cortex with de 6 layers		
<b>CODIFIED INFORMATION IN CORTEX</b>	PSYCHIC UNCONSCIOUS		PSYCHIC CONSCIOUS		
<b>SUBTYPES</b>	LÍMBIC PALEOCÓRTEX	HETEROTYPIC PALEOCÓRTEX	PARALÍMBIC NEOCÓRTEX	POSTERIOR NEOCÓRTEX	ANTERIOR NEOCÓRTEX

From a pathological point of view, instead of a quasi static biomedical Virkowian we prefer a multilevel schema which explains disease in both senses pathogenetic and pathokinetic, from the cellular to the conscious level of activity, of any person (fig. 1). Explaining disease in this way, it is possible to identify many more possibilities to discover and explain about epilepsy: are epilepsies determined by genetic, synaptic, functional, paleocortical o neocortical defects or disorders? Are this defects or disorders intrinsic to the cortex o extrinsic to it (cortical vs. functional subcortical, metabolic cerebral, genetic cerebral, or metabolic systemic and genetic systemic)? Are the processes of epilepsy *pathogenetically* or *pathokinetically* determined? The explanation of seizures would go through the similar line, but this has not to be exactly the same. The reason is that a seizure (the symptom) proceeds in a lineal way in seconds from a starting point in the epileptogenic lesion, while epilepsy (the disease) runs in months or even years. It means that one

has to classify seizures in terms of neocortical-conscious vs. paleocortical-unconscious, as well as cortical vs. subcortical-functional, synaptic-metabolic, and neuronal-genetic.



From a clinical point of view, in medical practice, even though not all practitioners (specialized or not) master explanatory theories, they need a multiple entry classification to remember all the well known diagnostic possibilities in order to decide the best diagnostic test, the best treatment and a prognosis. So, we suggest for them (and possibly for researchers):

- 1) A classification of seizures (the symptoms; i.e., table 3)
- 2) A classification of epilepsy (the disease), that is, of different clinical forms of epilepsy, or of different epilepsies, accounting data from:
  - a. Anamnestic examination (i.e. table 4)
  - b. Current examination (i.e. table 5)
  - c. Ancillary examinations (i.e. table 6)
- 3) A classification of epilepsy (or epilepsies) according to prognosis

**TABLE 3. Classification of epileptic seizures**

- I. Archicortical (hippocampal)
- II. Paleocortical
  - a. Límbic (olfactory, ...)
  - b. Heterotypic (tactile, visual, motor, ...)
- III. Neocortical
  - a. Paralimbic (affective)
  - b. Temporo-parieto-occipital (hallucinations)
  - c. Prefrontal dorsolateral (automatisms)
- IV. Multimodal (absence, convulsive, ...)

In relation to seizures, the aim is to localize the anatomical origin of the symptom complex of each seizure, in which case the clinician needs a simple rule of correlation (between the symptom and the possible anatomical focus (deduced from theory per se, an EEG, an image, or any other evidence). Then, it is necessary to reconsider a practical clinic-anatomical classification. At this point, seizures, always considered of cortical origin (even when they are determined by a subcortical or systemic causes), should be classified as it is shown in table 3.

As you can see, it is not necessary to say that the seizure is psychic or sensorial, since they both are psychic (either paleocortical or neocortical); even the motor ones, because they disrupt the psychological structure of voluntary actions, as a visual or tactile does. The sequence, extension and/or amplitude of the cerebral processes in between the epileptic focus discharge and the subjective or objective final symptomatic complex of the seizure, would be filled out theoretically, or deduced logically from the clinical evidence. For example, the role of the hippocampus, amygdala hypothalamus, thalamus, the various brain stem nuclei, or even the cerebellum, are less relevant from the clinical point of view.

In tables 4-6, I reproduce a proposed a multiple entry classification of epilepsy for use in clinical practice (in 1998), to show you the kind of schemas medical practitioners in general, we thought, need to attend epileptic patients.

**TABLE 4. Causes of epilepsy according to anamnesic examination:**

A. Clonic, tonic, myoclonic and other attacks during the neonatal period, usually to signs of diffuse cerebral dysfunction:

a) During the first week

- Perinatal hypoxia
- Perinatal trauma (with intracranial hemorrhage)
- CNS infection
- Congenital brain malformations
- Metabolic disorders: hypocalcemia, hypoglycemia, etc
- Sequel of any of above lesions (cerebral palsy, arterial or venous occlusion etc)
- Congenital metabolic disorders
- Neurocutaneous syndromes

b) During the second week

- CNS infection
- Electrolyte imbalance; infection, nutritional hypocalcemia
- Kernicterus
- Congenital brain malformations

c) Between third week and third month

- CNS infection
- Subdural suffusion due to intracranial infection
- Congenital brain malformations

B. Massive spasm partial and tonic-clonic attacks of infancy

- a) Infantile spasms, usually with neurological deficit and mental retardation (West syndrome, hypsarrhythmia); of unknown cause, rarely due to congenital brain malformations, sequel of traumatic or vascular disease; metabolic disorders
- b) Febrile seizures, usually without any other neurologic disorder: acute fever is provoking factor, cause is genetic.
- c) Tonic-clonic and partial seizures with or without secondary generalization.
  - Infections
  - Cerebral palsy, cerebral infarction or hemorrhage
  - Errors of metabolism
  - Neurocutaneous syndromes

C. Tonic-clonic, absence, myoclonic, clonic, tonic, atonic and mixed seizures, partial seizures of childhood and adolescence

- a) Primary generalized epilepsy: expressed with typical absences and/or tonic-clonic seizures with no associated neurological disorder: hereditary
- b) Lennox-Gastaut syndrome: generalized seizures in several combinations, with other signs of neurological deficit or mental retardation usually of unknown cause; a few ones as sequel of trauma, infection or early vascular brain disease, or metabolic disorders and other progressive hereditary metabolic disorders
- c) Focal seizures (rolandic, temporal, occipital, parietal, usually expressed by tonic-clonic or partial attacks related to focal lesion): usually benign forms of unknown cause; less frequently due to trauma, granuloma and other CNS infections, residual lesion of early disease, arteriovenous malformation, tumor.

D. Partial and tonic-clonic seizures of the adult, usually with other neurologic symptoms of focal or generalized dysfunction

- a) From 20 to 60 years
    - Cerebral cysticercosis
    - Brain tumors, primary or metastatic
    - Cranioencephalic trauma
    - Granuloma (particularly tuberculous), abscess
    - Alcoholism
    - Arteritis
    - Residual lesion or early disease
    - Arteriovenous malformation
  - b) After 60 years
    - Sequel of cerebral infarct or hemorrhage
    - Brain tumors, primary or metastatic
    - Cerebral cysticercosis
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**TABLE 5. Causes of epilepsy according to current examination**

A. Patients who have only epileptic seizures with no other signs of brain dysfunction:

- Febrile seizures (“benign”)
- Primary generalized epilepsy
- Benign rolandic epilepsy and similar seizures
- Focal non progressive nor residual, asymptomatic lesions

B. Patients who have epileptic seizures and headache, papilledema in some patients and/or signs of diffuse brain dysfunction deficit:

- Cerebral cysticercosis
- Brain tumors, primary or metastatic
- Arteritis

C. Patients who have epileptic seizures and signs of focal or diffuse brain functional deficit or diffuse residual (sequel)

- Cerebral palsy; mental retardation
- Hemiplegia-hemiclonic syndrome
- CNS infections: meningitis, encephalitis, cysticercosis, abscess, granuloma
- Cranioencephalic trauma: early posttraumatic epilepsy, late posttraumatic epilepsy.
- Cerebrovascular disease: brain infarct or hemorrhage due to diverse vascular diseases

D. Patients who have epileptic seizures associated to systemic metabolic disorder (usually acute disease)

- Hypoglycemia, hyperglycemia
  - Hypocalcemia, hypercalcemia
  - Hyponatremia
  - Intermittent porphyria
  - Kidney or liver failure; hypertensive encephalopathy
  - Eclampsia
  - Hypothyroidism, hyperthyroidism
  - Intoxications: lead, manganese, methylalcohol, organophosphate compounds
  - Alcoholism: abstinence, subacute alcoholic encephalopathy
  - Addiction to hypnotic and sedatives : barbiturates or benzodiazepines abstinence
  - Medical drugs: phenothiazines, tricyclics, penicillin, isoniazide
  - Extensive burning
  - Prolonged electroconvulsive therapy
  - Anoxic encephalopathy
  - Congenital metabolic errors
- 

**TABLE 6. Causes of epilepsy according to the ancillary examinations:**

A. Patients with epilepsy of unknown cause: these are patients with generalized seizures either convulsive or no convulsive (absence attacks, myoclonic attacks) , negative clinical examination, “specific” epileptic EEG activity (slow spike-wave 3Hz complex), other negative ancillary examinations.

They are usually disorders of a possible hereditary etiology, like primary generalized epilepsy and primary partial epilepsy (rolandic, occipital) both benign

**B.** Patients with epilepsy of undetermined cause: with tonic-clonic crisis, or simple or complex partial, negative clinical examination, focal “specific” epileptic EEG activity or normal EEG, and other negative ancillary examinations. Epileptogenic lesions are “scars” or residual without metabolic expression in CSF or blood, as shown by some radiologic findings (although there could be inespecific lesions which do no change on CAT scann through the years). Heredodegenerative diseases may be included in this group

**C.** Patients with epilepsy of determined cause: with any clinical type of seizures (except typical absences) usually with positive clinical examination, focal slow or generalized EEG activity and other ancillary examinations with enough data for the diagnosis of underlying pathology.

The most important diseases that may be confirmed by the available diagnostic methods are: cerebral cysticercosis, tumors, granulomas, arteriovenous malformations, and the sequel of previously treated diseases particularly trauma, infections, brain infarct or hemorrhage, cerebral involvement of some systemic diseases (i.e.: SLE)

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I wonder whether these classifications must be elaborated by the International Commission. Probably not, but it is necessary to let you know what I mean by a multiple entry classification with a clinical aim. (At the time we proposed those classifications –1998–, we did not offer a prognostic classification, in terms, for example, of benign vs. malignant, treatable vs. non-treatable, spontaneous remitting vs. non-remitting)

Another different situation is the theory or theories that explain each form of epilepsy or the different kinds of epilepsies; theory or theories that must explain the whole process of a disease or the various diseases. But, some people would argue that we need a classification of those theories, or perhaps better, a systematization of them. Let us suppose that we assume a splitter attitude, and prefer a systematization of the different diseases called epilepsies; a systematization of this kind requires of various entries, each from a specific point of view:

1. As a systemic disease: genetic, metabolic
2. As a nervous disease: genetic, metabolic, functional
3. As a cerebral disease: genetic, metabolic, functional, psychic
4. As a cortical disease: genetic, metabolic, functional, psychic unconscious (paleocortex involvement) , psychic conscious (neocortex involvement)

It should be noticed that, although an epileptic seizure starts from a cortical network, the disease might be started everywhere in the body. But the farther the etiologic process from the cortex, the lesser the probability of having a seizure (as is the case of a tumor or a parasite). Even more, the same processes we found in the body and may be found in the proper location of the cerebral cortex.

A systematization of this kind avoids at least two disadvantages of the classical approaches: we avoid the simple statistical correlation between syndrome and lesion/etiology; the model demands to fulfill the gaps along the pathologic process. Furthermore, the model reminds us that we need a double explanation: pathogenetic from cells to consciousness (neocortex with codified psychic information), and pathokinetic, from social kinds of information (and neocórtex) to cells (neurons). It also shows that the greater the complexity of a scientific explanation, the greater the possibility that it is really progressing. And, contradictorily, having solved up to a point the brain-mind dualism, the model seems to simplify any explanation.