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EPILEPSY IS NOT A DISEASE

PROPOSAL FOR A NEW APPROCH IN THE INTERNATIONAL CLASSIFICATION OF DISEASES (ICD)

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SUMMARY - What is epilepsy? a disease? a symptom? a syndrome? is it important to define it? Basing his study on an extensive bibliography, the author shows that epilepsy is not a disease. He emphasizes the significance of that definition and suggest a new approach in the International Classification of Diseases (ICD).

KEY WORDS: epilepsy, disease, symptom, syndrome, psychoses, new classification.

Epilepsia não é uma doença: proposta para uma nova estrutura da epilepsia dentro da classifcação internacional das doenças (CID)

RESUMO - O que é epilepsia? uma doença? um sintoma? uma síndrome? é importante defini-la? Baseando seu estudo em extensa bibliografia, o autor mostra que epilepsia não é uma doença. Ele enfatiza o significado dessa definição e propõe nova abordagem na Classificação Internacional das Doenças (CID).

PALAVRAS-CHAVE: epilepsia, doença, sintoma, síndrome, psicoses, nova classificação.

First of all, we must conceptualize what disease, symptom, syndrome and epilepsy are. According to the International Dictionary of Medicine and Biology[19] - "DISEASE [Middle English disease sickness, discomfort; from Old French desaise (from des - DIS + aise ease, from L adjacens Lying Close) Discomfort]: A condition which alters or interferes with the normal state of an organism and is usually characterized by the abnormal functioning of one or more of the host's systems parts or organs; ... a given disease is often manifested by a characteristic set of signs and symptoms; ... Disease is usually distinguished from ... and often from syndrome, a complex of symptoms descriptive of a disorder[21]. According to the dictionaries: Disease is more specifically a definite morbid process having a characteristic train of symptoms[6]. "An impairment of the normal state of the living animal ..."[33]. **Symptom** [Gk symptom (gen: symptomatos; from sympiptein to fall together, meet violently, fall in with, from SYM + Gk piptein to fall) anything that befalls one ...]: Any evidence of disease which is experienced by the patient and often reported as a subjective observation, such as pain. If the symptom is demostrable to an observer upon examination, it is an objetive symptom, or more generally, a sign[21]. **Syndrome** [Gk syndromé (from SYN + Gk dromos a running, course from dramein 2nd aorist inf. of trechein to run) a running together ...][6]:The aggregate of signs and symptoms considered to constitute the characteristics of a morbid entity: used especially when the cause of the condition is unknown[21]. **Epilepsia** [LATE L (from Gk epilepsies a laying hold of, seizure, from EPI ± Gk lepsomai, fut of lambanein to take hold of, seize ± IA) a seizure]. **Epilepsy:** A neurologic disorder characterized by the tendency to suffer recurrent seizures or fits, where minor or major"[21].

The Greek believed in something coming from the top making the patient fall to the ground on account of a sudden attack. Considering the etymological aspect of the word Epilepsy (a seizure), it gives us reason to believe that Epilepsy is a symptom. Another aspects relates to the word seizure, which means a disturbance that happens in the course of a disease. We all know the essencial characteristic of Epilepsy, which is recurrent seizures. Consenquently

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the word Epilepsy basically means epileptic seizures. Thefore, Epilepsy = seizures = symptom. Moreover, disease is a "definite morbid process"[6] and Epilepsy is a process which has not been properly defined.

According to Trimble, "In Clinical Medicine there are fundamental kinds of data. Symptoms are the complaints that the patient tells us about or are elicited by clinical history. Signs are observed by the physician, the patient, or a friend or relative of the patient, which indicate the presence of abnormal functioning of one or more bodily systems. A syndrome is a constellation of signs and symptoms which may coalesce to provide what seems to be a recognizable entity with definiting characteristics. Syndromes then become classified, and are the clinical representatives of ilness. The latter infers some biological change or variation of the organism and it is the task of Medicinal Science to explore this (...) Syndromes are not synonymous with diseases, since the same syndrome may have differing aetiologies and pathogeneses. As a consequence, classification of disease in Medicine generally represents a variety of notions, and Neurology is no exception. Epilepsy is a syndrome or group of syndromes identified by a key symptom, namely recurrent seizures (...)[31]. Adams (1966) wrote: "Epilepsy is an intermittent disorder of the nervous system due presumably to a sudden, excessive, disordely discharge of cerebral neurones. This was the postulation of Huglinghs Jackson (...). Sometimes it is an obvious symptom of a brain disease that also manifests itself in other ways, and at times it is the solitary expression of deranged cerebral function in an individual who otherwise maintains perfect health. The latter is the more frequent circunstances ans explains why the convulsive state has for so long been looked upon as a disease entity"[1].

Pupo (1971) has defined: "Epilepsy is not disease, it is a syndrome that is a group of various clinical manifestations that occurs as the result of several morbid brain process which varies considerably, according to the patient's age-group"[25]. Matthes (1976), "Epilepsy is not a nosological concept of disease, but at the same time, it is different from some paroxystic symptom such as cough or kidney colonic"[22]. Drapper (1979), "Epilepsy is a symptom not necessarily a disease. It may be a symptom of: 1) congenital neurologic dysfunction; 2) systemic metabolic disease; 3) structural disease of the brain"[5]. Goldsohn et al. (1984), "The word Epilepsy does not apply to a specific disease but to a group of complex symptoms of different aetilogies due to processes sometimes motionless, sometimes evolutionary"[15]. Jeavons and Aspinall (1985), "Epilepsy is an established tendency to recurrent seizures"[20]. Rothner (1985), "The original definition of Epilepsy by Jackson still holds good today although some improvement has been added to it. Epilepsy for example is not regarded as a disease; on the contrary it has been considered a symptom proceding from a disorded electrical activity of the brain as a consequence of a wide variety of disturbances"[26]. Huck (1983), "Epilepsy is not a disease as a specific defined morbid entity. In fact, the Epilepsies are syndromes, that is, a varied constellation of signs and symptoms from several aetilogies, showing a unity in its basic mechanisms"[17]. Porter (1987), "Epilepsy is a chronic disturbance. It is a group of syndromes rather than a disease"[24]. Shorvon (1988), "Epilepsy is best viewed as a symptom and the aetiology is varied and multifactorial"[27]. Dichter (1988). "The Epilepsy are a group of disturbances charatcterized by chronical and recurrent paroxystic changes in the neurological function caused by abnormalities in the activity of the brain"[7].

The last proposal for reviewing the Classification of Epilepsies and Epiletic Syndromes (1988/89), by the Commision on Classification and Terminology from ILAE, practically solved that question, refering to Epilepsy specifically as a disturbance or a syndrome[3]. Sunami (1989) says that "it is well known that Epilepsies have many pathogeneses and their symptoms are manifold. That is the reason why Epilepsy is regarded not as a single disease but as a syndrome"[30]. Fisher (1989): "Epilepsy is not a disease, but a collection of diverse syndromes, some of wich are secondary to other derangements, and some of which are seemingly primary"[12]. Niedermeyer (1990), "Is Epilepsy a disease? The answer is an emphatic 'no' "[23]. Engel (1990), "Epilepsy is a word employed to define a group of disturbances sometimes called Epilepsies which are characterized by transitory and spontaneous recurrent paroxysms of the hyperactive function of the brain, resulting in epileptic seizures"[9]. Simon et al. (1991), "Epilepsy, a group of disturbances characterized by recurrent seizures, is a common cause of episodic loss of consciousness"[29].

After considering so many definitions proposed by welknown scientists one should be led to believed that such a question has already been solved, but that is not so. Gastaut (1973) defines it: "Epilepsy is a chronic disease of various aetiologies, characterized by recurrent seizures proceeding from an excessive discharge of the cerebral neurons, regardless the clinical or paraclinical symptoms to which it should be associated"[13]. For Cambier et al. (1983), "Epilepsy is a long-lasting disease whose clinical manifestations are intermittent"[4]. Shorvon (1988) wrote: "Epilepsy in one of the commonets serious diseases in school aged children"[27]. Buchpieguel et al. (1992), "Epilepsy is a chronic neurologic disease characterized by recurrent ictal episodes derived from the synchronous hyperactivity of cortical neuronal populations"[2]. Gilman said recently that "Remarkable advances have clarified the pathogenesis of illnesses such as migraine and Epilepsy leading to specific and effective therapies for them"[14]. Shorvon refered to the incidence and prevalence of Epilepsy: "The importance of the disease definition in the evaluation of the prevalence index (...)"[28]. Guerreiro (1993) conceptualized: "There is not a complete and satisfactory definitions for epilepsy.

Epilepsy is a chronic condition, or a set of diseases which have in common epiletic seizures that recur in the absence of toxic-metabolic and febrile disease"[16]. Some authors define it as a symptom at the beginning of their papers, only to refer to it again in the course of their works as a disease[22,27].

The notion that now previals after reading papers and books about Epilepsy is that scientists have been worrying so much about the physiopathogenesis, classification and therapies of Epilepsy that they have overlooked considering a basic question: What is Epilepsy? On the other hand, we have noticed that conservative sectors, especially from Psychiatry and Psychological Medicine, a strong resistance in solving that question. Worse yet, some neurologists and psychiatrists are bringing back into use the old concept of Epileptic psychoses[31,32]. Even the ICD-10-NA, contributes to maintain the prejudice (see: FO6.8)[19]. If Epilepsy is not a disease, how can we accept it as a psychosis or an Epiletic Mental disease?[10,11,31,32]. Concerning this, Engel has said that "Although the evidences have been questionable, it was reported that aberrant features of personality, affective disturbances and psychoses, including Late Paranoid Schizophrenia, are most common in patients with Epilepsy, specially those with complex partial seizures from limbic system. It is uncertain to what extent those disturbances result from underlying pathological lesions or specific convulsive activities and to how much can be attributed to the effect of antiepileptic drugs therapy on long-term treatment and to what extent can be related to limitations imposed daily and to the stigma of being epileptic"[9]. By regarding Epilepsy as a symptom one is by no means underestimating it. It does not mean to disregard everything that has been done in the past or at present. Epilepsy is undoubtedly the most important and perhaps the only symptom that is exclusively related to the brain, and really deserves the whole attention of researches anywhere in the world. It is urgent to reach an agreement, to stop that indecision and prejudice.

The purpose of this paper is to provoke a discussion on the problem and to exclude Epilepsy from the International Classification of Disease as it appears in the ICD-10-NA[17]. My proposal is a classification based on aetiologic factors, without any changes in the present code. Epilepsy would remain then in its proper place as a single symptom or as the main symptom or one of the symptoms with a known or unknown aetilogy. It is certain that in the years to come the ICD-10-NA will come into use, but my suggestion remain open to future consideration.

CLASSIFICATION OF EPILEPSIES, BASED ON AETIOLOGY*

G40. Structural lesions of the brain, infectious diseases, systemic diseases, toxic and metabolic disturbances, genetic disorders. Other causes including unknown aetiologies, in which epilepsy is the only symptom or the major one. The type of epiletic seizure or syndrome can be mentioned along side the codification.

G40.0. Genetic disturbances with autosomal dominant, recessive or polygenic inheritance, liable for epiletic syndromes. Mention the type of syndrome.

G40.1. Genetic diseases and syndromes with autosomal dominant, recessive or X-linked. Mention the sort of malformation, and/or epileptic and/or metabolic disorders.

G40.2. Cerebral hypoxia and anoxia (in utero, during delivery, in the neonatal period). Others.

G40.3. Head trauma in all groups, including in utero, and perinatal (intracraneal hemorrhage, cerebral contusion, laceration and hematoma). Others.

G40.4. Systemic diseases (cardiovascular, endocrine, connective tissue and inflammatory gastrointestinal, metabolic, hematologic diseases, water electrolyte and acid-base imbalance). Others.

G40.5. Strokes. Cerebrovascular lesion in utero (porencephalic cyst formation), prenatal and perinatal arterial occlusion (infantile hemiplegia, the syndrome of Gastaut), vascular malformations (including Sturge-Weber syndrome), aneurysms, subarachnoidal hemorrhage, vasopasm, venous thrombosis, hypertensive encephalopathy (including toxemia of pregnancy). Others.

G40.6. Brain tumors (mengioma, arachnoid cysts, gliomas, gangliomas, hamartomas, tubers, epidermoids and dermoids, hemimegalencephaly), cleidocranial dysostosis. Others.

G40.7. Infections: prenatal and perinatal infections (toxoplasmosis, cytomegalic inclusion disease, rubella, herpes and syphilis), late progressive encephalitis (congenital rubella and rubella virus), chronic encephalitis of Rasmussen's syndrome, meningitis, encephalitis, abscesses, granulomas, parasitic infections (cysticercosis, malaria, shistosomosis, etc.) AIDS (opportunistic infections, lymphomas, encephalopathy due HIV). Others.

G40.8. Acquired degenetative diseases: multiple sclerosis, presenile and senile dementia of the Alzheimer's type. Others.

G40.9. Others cases (miscellanies). Dysplasias, heterotopias, toxic disturbances (lead poisoning, organic and inorganic compounds, proconvulsant drugs), toxic-metabolic processes (uremia and hyperammonemia) hippocampal sclerosis.

G41. Unknown and controversial aetiologies.

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