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CLINICAL SYNDROMES RELATED TO FOCAL EPILEPSY IN NEONATES, INFANTS, CHILDREN AND YOUNG PEOPLE - LITERATURE REVIEW

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Abstract: Introduction: Convulsive crises are common events in childhood, affecting up to 10% of children. If we consider only epilepsy, we will see an incidence of 3% in the general population, with half of these cases starting in childhood. **Goal**: There are some clinical syndromes of focal epilepsy classified according to the age at which they appear, such as in neonates, infants, children and young people. This article aims to describe the syndromes according to their age group of presentation. **Method:** The search was carried out in the PubMed, BVS and Scielo databases, and was limited to articles between the period 2019 to 2023 that met the criteria of being literature reviews and case reports. **Result**: The clinical syndromes of epilepsy are divided into focal and generalized. We focus on benign neonatal seizures, benign familial neonatal seizures, benign partial epilepsy of infancy, benign partial childhood epilepsy with sharp centrotemporal waves, partial childhood epilepsy with sharp occipital waves, and continuous partial epilepsy.
Keywords: Epilepsy; Newborn; Infant; child; Adolescent;

INTRODUCTION

Epilepsy is a brain disorder characterized by a predisposition to generate seizures with possible consequent biological, cognitive, psychological and social repercussions. [1].

The clinical diagnosis of epilepsy is made by the presence of at least one seizure, with evidence of electroencephalogram or clinical history, such as abnormalities in the neurological examination and developmental delay, which suggest a risk of recurrence in the future. [2]. Another epidemiological definition is to consider epilepsy as the presence of two or more unprovoked seizures with an interval between them greater than 24 hours. [2].

The epileptic seizure is triggered by an abnormal electrical discharge in the

cerebral cortex, which can be restricted to a certain area, characterizing focal seizures; or disseminated/diffuse throughout the brain, characterizing generalized seizures [3]. The clinical expression of these abnormal electrical triggers will be compatible with the cortical area stimulated, therefore, motor, sensory, psychic or autonomic crises may occur. [3].

The classification of epilepsies according to the International League against Epilepsy (ILAE), 2009, proposes the definitions: focal epilepsy for electrical discharges located in one hemisphere and which may or may not be dyscognitive (with alteration of consciousness), and generalized for distributed discharges bilaterally; and according to the etiology, defines the causes as genetic, metabolicostructural and unknown [4].

Convulsive seizures are common events in childhood, affecting up to 10% of children, and this group includes febrile seizures, symptomatic seizures and epilepsy. [5]. If we consider only epilepsy, we will see an incidence of 3% in the general population, with half of these cases starting in childhood. [5].

Only 30% of children will have epilepsy after the first seizure and, if the neurological examination, electroencephalogram and neuroimaging are normal, this risk drops to 20%5. Epilepsy in childhood has a good prognosis in most cases, and 10-20% of children will develop seizures that are difficult to control with medication and neurological deficit [5].

One of the pathophysiological hypotheses that tries to explain why areas of neuronal death, for example, after hypoxia or infection, can become epileptogenic foci, argues that there would be a selective loss in these places, mainly of GABAergic inhibitory neurons, with excitatory synapses prevailing; other studies show that the sites of neuronal injury would form, over time, aberrant excitatory circuits [6].

There are some clinical syndromes of epilepsy classified according to the age at which they appear, such as in neonates, infants, children and young people. [7]. This article aims to describe the syndromes according to their age group of presentation.

The clinical syndromes of epilepsy are divided into focal and generalized [8]. We focus on benign neonatal seizures, benign familial neonatal seizures, benign partial epilepsy of infancy, benign partial epilepsy of childhood with sharp centrotemporal waves, partial childhood epilepsy with sharp occipital waves, and partial continuous epilepsy [8]. As generalized seizures, absence seizures, tonic-clonic, myoclonic and epileptic encephalopathies are observed. [8].

With regard to the benign neonatal crisis, it is also called the fifth day crisis, as it tends to start between the third and seventh day of life. It is characterized by unilateral clonic movements usually associated with apnea, which occur several times in 24 to 48 hours and disappear without leaving sequelae [9]. It is more frequent in males, has a good prognosis and neurological development is normal. [9]. Interictal EEG may be normal, focal, multifocal, or reveal alternating spiked theta pattern [9]. Ictal electroencephalogram usually shows spikes or slow waves in the centrotemporal region [9].

About neonatal seizures beingnas familial, it is characterized as epileptic syndrome of autosomal transmission, determined by a mutation in the long arm of chromosome 20 (20q 13,2)[10]. They are characterized by unilateral tonic movements, associated with tachycardia and apnea, which begin between the second and third day of life and persist for a long period. [10]. They tend to disappear without leaving sequelae, but the risk of secondary epilepsy is 11%, slightly higher than for the general population. [10]. Interictal EEG may be normal, focal, multifocal, or

reveal alternating spiked theta pattern [10]. Ictal electroencephalogram usually shows attenuation of background activity followed by spikes or focal or generalized slow waves [10].

With regard to infants, there is Watanabe syndrome or benign partial epilepsy in infants, which is characterized by the onset of complex partial or partial seizures with secondary generalization in infants aged between three and twenty months, with normal neuropsychomotor development before and after onset of seizures, with good response to antiepileptic drugs and normal interictal electroencephalogram [11]. Ictal EEG may reveal focus in temporal, central, parietal, or occipital regions [11]. Complex partial seizures can occur in clusters of up to 10 times a day, and loss of contact with the environment, ocular and head deviation, mild automatisms and clonic movements of the face and one of the limbs are observed. [11].

As far as childhood is concerned, we have benign partial childhood epilepsy with sharp centrotemporal waves or rolandic epilepsy [12]. This epilepsy has an entirely benign character. [12]. In the vast majority of patients, abnormal EEG findings disappear with age12. Manifestations are most common between two and fourteen years of age, with a peak incidence between nine and ten years of age. [12]. It affects boys more often and has a strong familial predisposition. [12].

Clinical findings consist of motor seizures that begin on the hemiface [13]. Oropharyngeal symptoms are also present and include tonic contractions and paresthesias of the tongue, tingling of the cheek, dysphagia, and excessive salivation.[13]. Tonic-clonic involvement of the ipsilateral extremities can be found [13]. Consciousness may or may not be preserved and, eventually, secondary generalization occurs. [13]. Symptoms affect children more commonly at night, reaching 75% of cases13. Electroencephalogram demonstrates spike waves in the centrotemporal region or rolandic area [13].

Still regarding childhood, we have benign partial childhood epilepsy with acute occipital waves (Gaustat type)[14]. The average age is 8.5 years. Begins with visual symptoms such as shimmering scotomas, sudden loss of vision, rapid visual hallucinations or illusions, followed by hemiclonic jerks, complex partial seizures, or generalized tonic-clonic seizures (secondary generalization)[14].

Post-ictal symptoms are marked: severe headache, nausea and vomiting, similar to a migraine [14]. The baseline EEG activity is normal; With eyes closed, high amplitude wave spikes are observed in the occipital region. [14]. Spontaneous sleep can trigger these paroxysms and eye opening suppresses the paroxysms [14].

With regard to childhood partial epilepsy with acute occipital waves (Panayotopoulos type), the average age of onset is 4.5 years [15]. Always begins with vomiting, followed by ocular and head version, clonic jerks or secondarily generalized tonic-clonic seizures [15]. The electroencephalogram also reveals sharp waves with occipital predominance, predominant during spontaneous sleep. [15]. They are not reactive to eyelid opening and closing [15].

Last in childhood we have Rasmussen's encephalitis or continuous partial epilepsy [16]. It is characterized by unilateral continuous motor partial seizures, starting around the age of ten [16]. It usually manifests after a febrile illness and is characterized by poor response to anticonvulsant treatment and progressive neurological deficit. [16]. Over time, hemiparesis/plegia, cognitive impairment, hemianopia and aphasia are established [16].

Some studies suggest an autoimmune etiology with the formation of antibodies that bind to glutamatergic receptors, activating them. Neuroimaging performed in later stages

of the disease reveals atrophy of only one of the cerebral hemispheres [16].

MATERIAL AND METHODS

The search was carried out in the PubMed, BVS and Scielo databases, and was limited to articles between the period 2019 to 2023 that met the criteria of being literature reviews and case reports.

Then, the keywords of the titles of the articles were analyzed and those whose theme best fits our objective were selected.

A total of sixteen articles were selected for full reading.

DISCUSSION

Often, any type of seizure is seen as epilepsy. There are dozens of differential diagnoses for seizures that must be properly investigated by a professional. [7].

CONCLUSION

The clinical syndromes of epilepsy are divided into focal and generalized. We focus on benign neonatal seizures, benign familial neonatal seizures, benign partial epilepsy of infancy, benign partial childhood epilepsy with sharp centrotemporal waves, partial childhood epilepsy with sharp occipital waves, and continuous partial epilepsy. As for generalized seizures, there are absence seizures, tonic-clonic, myoclonic and epileptic encephalopathies.

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