

## NON-EPILEPTIC PAROXYSTIC DISORDERS AS DIFFERENTIAL DIAGNOSIS OF SEIZURES IN PEDIATRICS - LITERATURE REVIEW

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**Abstract:** **Introduction:** Seizures are common events in childhood, affecting up to 10% of children. <sup>1</sup>. Non-epileptic paroxysmal disorders are conditions that begin abruptly, with a stereotyped course and spontaneous termination, often leading to altered consciousness and clonic/tonic motor manifestations that mimic seizures. <sup>3</sup>. **Goal:** Such conditions need to be known by the pediatrician and neuropaediatrician, as their treatment is absolutely different from epilepsy and, therefore, this article aims to describe them. **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:** It is essential to be able to understand the difference between paroxysmal non-epileptic disorders (NPPE) and epileptic seizures for an adequate treatment for children<sup>3</sup>. For neonates, we have as DPNE concussions and benign sleep myoclonus. For infants, shortness of breath, shivering, paroxysmal torticollis, spasmus nutans and jactatio capitis. Finally, in preschoolers and schoolchildren, we have vasovagal syncope, syncope associated with cough, and long QT syndrome.

**Keywords:** Differential diagnosis; Convulsions; Pediatrics.

## INTRODUCTION

Convulsive seizures are common events in childhood, affecting up to 10% of children, and this group includes febrile seizures, symptomatic seizures and epilepsy. <sup>1</sup>. If we consider only epilepsy, we will see an incidence of 3% in the general population, with half of these cases starting in childhood. <sup>1</sup>.

Epilepsy is a brain disorder characterized by a predisposition to generate seizures with possible consequent biological, cognitive, psychological and social repercussions. <sup>2</sup>.

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.<sup>2</sup> 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.<sup>2</sup>

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<sup>2</sup>.

Non-epileptic paroxysmal disorders are conditions that begin abruptly, with a stereotyped course and spontaneous termination, often leading to altered consciousness and clonic/tonic motor manifestations that mimic convulsive seizures, but are not caused by abnormal cortical electrical discharges.<sup>3</sup>

Such conditions need to be known by the pediatrician and neuropsychiatrician, as their treatment is absolutely different from epilepsy, with no improvement with the use of anticonvulsant drugs.<sup>3</sup> The neurological examination, neuropsychomotor development and electroencephalogram of these children are completely normal<sup>3</sup>.

In neonates, we can observe convulsions, which are tremors of the limbs often observed in healthy full-term babies or in those with polycythemia, children of diabetic mothers, withdrawal syndrome or in those with some degree of hypoxia at birth.<sup>4</sup> Tremors differ from convulsive clonic tremors in that they always occur when the neonate is awake, usually after tactile stimulation, cease after repositioning the limbs, are faster and have less amplitude than clonic seizures, and are not associated with abnormal ocular/orobuccolingual movements or alterations in

color/apnea/heart rate<sup>4</sup>.

There are also, in sleep, myoclonic jerks similar to startles, which occur naturally at the onset of sleep or close to awakening. They do not have an epileptiform representation on the electroencephalogram<sup>5</sup>.

In infants, we can observe breathlessness crises, which are characterized by an involuntary apnea at the end of expiration, and can be divided into cyanotic form and pale form<sup>6</sup>. Shortness of breath attacks are common paroxysmal disorders that affect infants between six and eighteen months and have an incidence of 5%<sup>6</sup>. They are unrelated to the child's temperament or behavioral problems and a positive family history can be found in 20 to 35% of cases.<sup>6</sup>

The cyanotic form is the most common<sup>6</sup>. The episode is always preceded by anger or frustration imposed on the baby, followed by high-pitched crying and, subsequently, a forced expiration, apnea and cyanosis, lasting approximately one minute<sup>6</sup>. If prolonged, the infant loses consciousness and may experience tonic stiffening of the limbs, assuming an opisthotonos posture.<sup>6</sup> In this form, there is a spasm of the glottis and expiratory muscles at the end of expiration, increasing intrathoracic pressure; with this, cardiac output is reduced and cerebral oxygenation is reduced<sup>6</sup>.

The pale form is due to an exacerbated vasovagal reflex triggered by an episode of sudden fear or pain (e.g. bumping the head, falling)<sup>6</sup>. The child begins to cry, and quickly becomes pale, loses consciousness, and subsequently may have a hypoxic reflex crisis, characterized by clonic jerks and urinary incontinence.<sup>6</sup> Some episodes may have an associated cyanotic component<sup>6</sup>. In this form, increased vagal reflex leads to bradycardia, low cardiac output and reduced cerebral perfusion.<sup>6</sup> Ocular compression can trigger a similar episode, as such a measure leads to a vagal discharge with bradycardia

through the oculocardiac reflex.<sup>6</sup> However, it is not recommended to perform this test by an inexperienced physician or without the presence of equipment or medication for the treatment of asystole.<sup>6</sup> The presence of iron deficiency anemia intensifies shortness of breath in the pale form<sup>6</sup>.

The main measure in managing these cases is to reassure the parents about the benignity and transience of the episodes. Guide them not to wet the child, or shake it or perform mouth-to-mouth resuscitation. Just lay her down on a flat surface or turn her on her side.<sup>6</sup>

Besides, in infants, we have attacks of tremors or shivering.<sup>7</sup> They are characterized by sudden flexion of the head, neck and trunk followed by shudders and tremors resembling chills, lasting less than ten seconds, and up to one hundred attacks a day may occur.<sup>7</sup> The peak incidence is between four and six months of age, and may remain for up to seven years.<sup>7</sup> There are no EEG abnormalities and some children will develop essential tremor. No specific treatment is needed<sup>7</sup>.

Paroxysmal torticollis occurs from two to eight months of life, and resolves spontaneously around two to three years.<sup>8</sup> It is characterized by a tonic deviation of the neck, associated with pallor, restlessness, vomiting, without loss of consciousness<sup>8</sup>. In the preschool phase, it can evolve into benign paroxysmal vertigo, and in the school/adolescent phase, migraine can be observed.<sup>8</sup> Such cases may be associated with calcium channel mutation<sup>8</sup>. Infants with permanent torticollis must be investigated with imaging tests to identify abnormalities in the cervical vertebrae and spinal cord and posterior fossa tumors<sup>8</sup>.

In spasmus nutans, we have typical episodes, characterized by the triad: pendular nystagmus, head nods and torticollis<sup>9</sup>. It starts between four months and one year<sup>9</sup>. In most cases it is a benign and transient condition,

but at other times it may be the manifestation of a hypothalamic tumor or optic chiasm.<sup>9</sup>

Finally, in infants, there is jactatio capitis<sup>10</sup>. It is a rhythmic and prolonged shaking of the head at the beginning of and during sleep.<sup>10</sup> It seems to be a comforting move for the child. There is no change in tone and other abnormal movements. Most often seen around one year<sup>10</sup>.

For preschoolers and schoolchildren, we have the well-known vasovagal syndrome, which are episodes characterized by loss of consciousness, loss of muscle tone, followed or not by brief tonic contractions of the face, trunk and extremities, and some children may present upward gaze deviation.<sup>11</sup> Syncope can be differentiated from seizures by being shorter, lasting less than 20 seconds, having a feeling of “empty” head, nausea, sweating, associated visual blurring and weakness, and complete recovery of the memory of the episode<sup>11</sup>. They are more frequent among girls, usually after ten to twelve years.<sup>11</sup>

Syncope occurs due to a reduction in cerebral blood flow due to arterial hypotension.<sup>11</sup> Generally, stimuli such as pain, fear, excitement or prolonged standing in the standing position during hot weather trigger a vasovagal reflex, leading to bradycardia and arterial hypotension.<sup>11</sup> With ischemia, the cortical centers release inhibition on the ascending reticular formation of the trunk and the individual loses the conscience<sup>11</sup>. In most cases, no treatment is necessary, in others, vasopressin can be tried.<sup>11</sup>

On the other hand, syncope associated with cough is more common in asthmatic children, usually triggered by intense coughing shortly after falling asleep.<sup>12</sup> The child coughs, producing an intense increase in intrathoracic pressure, with reduced venous return and reduced cardiac output, leading to cerebral ischemia<sup>12</sup>. Thus, there is loss of consciousness, loss of muscle tone, upward gaze deviation,

and associated clonic movements in cases of prolonged cerebral ischemia.<sup>12</sup> The treatment is done with control of asthma attacks<sup>12</sup>.

We also have long QT syndrome, which is an underlying heart condition that must be suspected whenever there is exercise-related syncope.<sup>13</sup> Long QT syndrome is an arrhythmia characterized by a QT interval  $\geq$  0.46 milliseconds<sup>13</sup>. It affects about 1:10,000 to 1:15,000 people, either due to acquired heart disease, such as viral myocarditis, mitral valve prolapse or congenital, with recessive or dominant autosomal inheritance<sup>13</sup>.

## 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 13 articles were selected for full reading.

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## DISCUSSION

It is essential to be able to understand the difference between non-epileptic paroxysmal disorders and epileptic seizures. The differential diagnosis in convulsive crises is of paramount importance for an adequate treatment of children.<sup>3</sup>

## CONCLUSION

It is essential to be able to understand the difference between paroxysmal non-epileptic disorders (NPPE) and epileptic seizures for adequate treatment of children<sup>3</sup>. For neonates, we have as DPNE concussions and benign sleep myoclonus. For infants, shortness of breath, shivering, paroxysmal torticollis, spasmus nutans and jactatio capitis. Finally, in preschoolers and schoolchildren, we have vasovagal syncope, syncope associated with cough, and long QT syndrome.