Sandifer Syndrome

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Sandifer syndrome, named after neurologist Paul Sandifer, was first reported by Marcel Kinsbourne in 1962, who noted an upper gastrointestinal disorder that occurs in children and adolescents with neurological manifestations. Sandifer syndrome is a neurobehavioral disorder that causes a series of paroxysmal dystonic movements in association with gastroesophageal reflux and, in some cases, with hiatal hernia. It is characterized by esophagitis, iron deficiency anemia, and is often mistaken for a seizure of epileptic origin [1].

The incidence and prevalence of Sandifer syndrome are not known for sure, and the epidemiology has not been well studied. However, there are authors who estimate that 7% of babies may present typical GERD symptoms, such as regurgitation, and 1% of them may present as associated muscular dystonia [2].

Its appearance generally occurs during childhood. Dystonic movements are characterized by an abnormal posture of the head and neck (torticollis) and a severe arch of the spine. The episodes usually last between 1 and 3 minutes and can occur up to 10 times a day, although they are usually associated with the ingestion of food, it is also common to find abnormal involuntary eye movements, decreased mobility of the cervical spine, and hematemesis [2].

Although its pathophysiological mechanism remains uncertain, several investigators have indicated that dystonic posture is due to a pathological reflex in response to abdominal pain caused by gastroesophageal reflux and esophagitis. Although conflicting results have been obtained, some authors have suggested that dystonic posture provides relief from abdominal pain [3].

The signs and symptoms of this condition are usually mistakenly attributed to neurological problems, leading to the exposure of affected children to unnecessary, often expensive, and invasive procedures and often to the prescription of ineffective and ineffective medications. free of significant side effects [3].

Sandifer syndrome is diagnosed on the basis of the association of gastroesophageal reflux with the characteristic movement disorder. The neurological examination is usually normal. This syndrome can affect the child by generating a psychomotor delay, so the early detection and identification of this delay is
essential for their well-being and their environment [4].

In patients whose presentation is not as clear, research has shown that simple video-EEG monitoring can clarify the diagnosis, as the EEG will provide valuable clinical information to rule out seizure disorders and metabolic conditions. Scintigraphy and upper gastrointestinal tract studies are left to evaluate anatomical abnormalities such as hiatal hernia [4].

Differential diagnoses for Sandifer syndrome should include all diseases that produce paroxysmal events in childhood, including epileptic syndromes, congenital muscular torticollis, metabolic conditions, CNS pathologies, trauma, and inflammatory or infectious conditions of the neck and head [5].

Its main complications include irritation of the respiratory tract, including cough, wheezing and stridor, apnea, and/or bradycardia [5].

In terms of treatment, lifestyle changes are the first step in treating Sandifer syndrome. About 90% of babies will improve their symptoms in 2 weeks with lifestyle changes alone. Parents of healthy, full-term infants should be advised to thicken expressed breast milk or formula with 1 tablespoon of infant rice cereal for every 2 to 4 oz. Of breast milk or formula, or to use commercially prepared pre-thickened formula. Thickened formula should be used with caution with premature babies due to the increased risk of necrotizing enterocolitis. The child should be placed fully upright after feedings, as recent research reports appear to be beneficial, as does placing the child in the prone position. Babies can be placed on their tummies for only short periods and only while awake and supervised. They should not be allowed to sleep in the prone position due to the increased known risk of sudden infant death syndrome. An elevated or supine position, such as leaning on a carrier, does not provide any benefit [6].

If all of these lifestyle changes fail, drug therapy with antireflux medications should be started. Proton pump inhibitors and H2 blockers are first-line therapy. In most cases, paroxysmal events will resolve or dramatically decrease in frequency over time [7].

Surgical interventions for a possible hiatal hernia should be considered if pharmacological and non-pharmacological measures fail. Surgical correction with fundoplication shows almost complete relief of symptoms in 70% to 90% of children. Fundoplication surgery increases the length of the intra-abdominal portion of the esophagus, accentuates the angle of His, and corrects the hiatal hernia if present. This is an important procedure and is considered safe, but as already mentioned, it should be reserved for those who fail other therapies [7].

On the other hand, early stimulation of deficiencies in psychomotor development will be key to inhibiting inappropriate patterns acquired. The applied treatment consists of stimulating the child through active play to achieve the most appropriate psychomotor progression, avoiding pain and discomfort generated by perceptual-sensory alterations and by the syndrome itself [8].

The prognosis for these infants is excellent; prompt and appropriate treatment decreases dystonic episodes, and over time they resolve in most patients [8].

Much is still unknown about Sandifer Syndrome, so we must continue researching and paying special attention to affected patients since it is a digestive disorder with great influence on psychomotor development.

Competing Interests
The author has read and approved the final version of the manuscript. The author has no conflicts of interest to declare.

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