Sobbing spasm

DEFINITION

Sobbing spasm is the most common non-epileptic paroxystic phenomenon in infants and preschool age children; it is observed in 5 to 7% of infants and preschool age children. It is secondary to episodes of acute cerebral hypoxia precipitated by crying, pain, surprise, or frustration. Following a deep inspiration a spasm is produced, which stops respiration and conditions apnea. The patient goes rigid, with cyanosis of lips and fingers and toes, or flaccid and pallid.

It often begins between 6 and 12 months of life; in the large majority of cases it disappears before 6 years of age. The greatest frequency occurs between one year and two years of age.²,³

Classification

Two types of sobbing spasm are recognized:

A. Cyanotic: episodes that begin in the course of crying due to frustration, pain, or anger. After one or several respiratory movements during crying, it is interrupted, the child goes into apnea, and after a few seconds turns cyanotic, and may even lose consciousness. Loss of consciousness may be associated with generalized hypotonia or hypertonia with opisthotonos, and subsequently short seizures. This sequence may be complete or incomplete. In any case, recuperation is immediate. After a few seconds the child regains full wakefulness. The total duration is 1-4 minutes.⁴,⁵

Four grades are distinguished:

Grade 1. Crying with prolonged inspiration and brief apnea.

Grade 2. Crying, prolonged inspiration, apnea and acrocyanosis.

Grade 3. Crying, prolonged inspiration, apnea, cyanosis, and hypotonia or hypertonia.
Grade 4. Crying, prolonged inspiration, apnea, cyanosis, hypertonia and generalized clonic seizures.

**B. Pallid or reflex anoxic seizure**: following minor trauma (especially to the head) or a situation of fear or surprise, the patient starts crying and loses consciousness; the patient is pallid with generalized hypotonia and may also present clonic seizures of extremities. This is due to an increased cardioinhibitory response, secondary to an increase in vagal tone.

In some cases, the two forms alternate in the same patient. Some children with pallid spasms will subsequently present infant juvenile syncope as a hypervagotonic reflex reaction to situations of stress.2,3

**C. Mixed.** Pallid and cyanotic sobbing spasms may coexist in a patient, although it is uncommon.5

**Pathophysiology**

Sobbing spasm has been related to a primitive childhood respiratory reflex; it has a certain degree of family aggregation, because one in every four children with sobbing spasm has a direct family member who suffered it in infancy.6 Some authors have related iron deficiency anemia to the genesis of sobbing spasm and hemoglobin below 8 g/100 mL has been reported in up to 23.5% of children with sobbing spasms. Iron deficiency may play a role in the pathophysiology of sobbing spasm because iron is important for catecholamine metabolism and the functioning of neurotransmitters.7

It has been suggested that a delay in maturation of brainstem myelination, measured through brainstem evoked potentials, may influence the development of sobbing spasm.6,8

**Recommended medical evaluation**

1. Complete clinical history.
2. Physical examination.
3. **Blood biometry** in case of suspected anemia.
4. An **electroencephalogram** should be performed if epilepsy is suspected, when there is no evident triggering phenomenon, in patients under 6 months or over 6 years of age.
5. **Electrocardiogram** in case of pallid sobbing spasm. To rule out arrhythmias or long QT syndrome.

**Management of sobbing spasm**

1. Keep calm.
2. Remove objects the child has in his or her mouth.
3. Place the patient on his or her side and remove any objects with which he or she could hit himself/herself.
4. Maintain a well-ventilated environment, loosen the patient’s clothing.
5. Do not attempt to stop the spasm.
7. When the spasm ends let the patient take a short nap.
8. If the spasm is caused by pain offer consolation and relief.

If a child has several sobbing spasms a day behavioral management is probably inappropriate.
Some children use spasms as an inappropriate form of communication.

**What should be avoided?**

1. Resuscitation maneuvers.
2. Striking or bathing the child with cold water. There is a risk of bronchoaspiration and pulmonary complications.
3. Do not place objects in the child’s mouth, as they can cause injury or suffocation.
4. It is very important not to confuse sobbing spasm with other convulsive disorders; avoid administering antiepileptic drugs as much as possible.

**When is medical reevaluation needed?**

1. When episodes occur without a triggering factor or during sleep.
2. In case of convulsive movements.
3. In case of onset before 5 months or after 6 years of age.
4. When there is diagnostic doubt.

The prognosis is good, sobbing spasm is not associated with subsequent development of epilepsy or cognitive alterations.

**Treatment**

There is no specific medication; it is recommended to provide extensive information to the family and use educational strategies for control and inhibition of sobbing spasms.

If iron deficiency anemia is confirmed management with diet and ferrous sulfate is suggested.5,7 Some authors recommend the use of piracetam in patients with recurrent sobbing spasm.9

Antiepileptic treatment is not justified.

Long-term follow-up suggests that sobbing spasms are alleviated spontaneously. Patients present a higher incidence of attention problems, and in the case of pallid spasms, may present a higher incidence of syncope.10

**REFERENCES**