Postgraduate Medicine

Breath-holding spells in children

How to distinguish the benign type from serious conditions

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Breath-holding spells (BHS) are dramatic, involuntary episodes that occur in otherwise healthy children. These episodes are often frightening to parents and worrisome to physicians. Because they can mimic serious or even life-threatening conditions, these generally benign spells require careful evaluation. In this article, Dr Jennette reviews how to differentiate conditions that require treatment from harmless breath-holding episodes that a child will outgrow.

Childhood breath-holding conjures up an image of a stubborn toddler willfully holding his breath until he gets what he wants. The reality is quite different, however. The typical breath-holding episode begins when a child becomes upset, is startled, or suffers a minor injury, and then begins to cry. Crying may be brief or prolonged, but typically, after a few cries, the child becomes silent and apneic in what is described as noiseless expiration. This stage quickly is followed by a dramatic change in skin color. The skin becomes cyanotic or pallid or has a mixed-color appearance.

In simple BHS, the event resolves with no associated syncope or postural change. In severe BHS, however, subsequent loss of consciousness and change in postural tone do occur. Usually the child falls limp, and occasionally a few myoclonic jerks may be observed. In some cases, a brief period of increased muscle tone, or opisthotonos, may be seen after or instead of limpness.

The entire episode, which lasts from several seconds to more than a minute, may end with a sudden, deep inspiration or with the return of normal breathing. Especially with severe BHS, the child may be drowsy for a few moments before recovering completely and resuming normal activities.

Epidemiologic factors

BHS are a fairly common pediatric problem. Simple spells occur in 27% or more of healthy children, and severe episodes may be seen in as many as 4.6% (1,2). In most children who have BHS, spells begin by age 12 months, although some children begin experiencing them as early as 2 months of age. Usually, breath-holding events with cyanotic skin color change begin between a child's neonate period and 18 months of age. For spells with pallid skin color, the age at onset is 12 to 24 months. By 24 months, almost all children who experience BHS have had their first episode. Among 384 children studied by Livingston, the mean age at onset was 12 months (range, 3 months to 4 years) (2).

Spells can occur as often as several times a day or as rarely as once a year. More commonly, patients have several episodes a week, and overall, occurrence ranges from daily to monthly. The greatest frequency of events tends to be in the second year of life. By the time patients are 4 years old, about half of breath-holding cases have spontaneously resolved; by age 6, about 90% have done so; and by age 7 or 8, virtually all have resolved (1,2).

Possible explanations

Because they occurred in the setting of anger, agitation, or frustration, BHS were thought for many years to derive from emotional or behavior problems. In the past, children who had them were described as stubborn, disobedient, and aggressive. In 1993, however, DiMario and Burleson3 found no significant differences between the behavior profiles of breath-holders and those of other children.

Instead, several different physiologic mechanisms have been proposed. For some children with BHS, especially those with the pallid type, noxious stimuli may lead to centrally mediated cardiac inhibition through the vagus nerve. In turn, this mechanism may induce bradycardia or brief asystole and subsequent spells. This cardiac phenomenon has been demonstrated repeatedly in patients through the use of ocular compression (1,4). In children with the cyanotic type of BHS, about 25% have a positive reaction to ocular compression; in those with the pallid type, the percentage increases to 61% to 78% (2). Similarly, cyanotic episodes may be caused by central inhibition of respiratory movements, again mediated through the vagus nerve (2,4). In both situations, cerebral hypoxia results.

In other patients the problem may be genetic, resulting in a more generalized dysregulation of the autonomic nervous system. The role of reduced central nervous system (CNS) sensitivity to hypoxia and hypercapnia, as well as abnormalities in pulmonary reflexes and lung mechanics, also has been studied (5).

Differential diagnosis

Because potentially life-threatening conditions may present similarly to BHS, diagnosis of the benign condition can be difficult. Therefore, evaluation of breath-holding episodes requires the consideration and elimination of other, more worrisome explanations.

Seizure disorders often are the first entity to exclude because BHS can closely resemble them, particularly when myoclonic jerks are observed. Likewise, in episodes with vivid

color change and, in some cases, sudden loss of consciousness, cardiac problems and rhythm disturbances (including congenital QT syndrome) must be considered (table 1). Orthostatic syncope and apnea are also differential diagnostic considerations.

Table 1. Differentiation of severe BHS from generalized seizures and cardiac disturbances

Feature	Severe BHS	Generalized seizures	Cardiac disturbances
Age at onset	Often infancy	Rarely infancy	Variable
Family history of BHS	Often positive	None	None
Precipitating event	Usually present	Usually absent	Usually absent
Sleep state	Always awake	Asleep or awake	Awake, often with stressor
Pallor or cyanosis	Always; before syncope	Variable; after syncope	Variable
Myoclonic jerks	Variable; few beats	Usually	Absent
Incontinence	Uncommon	Common	Absent

BHS, breath-holding spells.

Although it is a much less likely scenario, BHS may be secondary to CNS lesions and malformations, such as Arnold-Chiari malformation. Spells may be associated with developmental disorders, such as Rett syndrome, or with Riley-Day syndrome, a familial dysautonomia (2). However, these uncommon entities are themselves associated with multiple other findings and can be excluded readily on the basis of history and physical examination results.

not with Pmith

Lastly, sporadic reports have linked BHS with underlying hematologic abnormalities, including transient erythroblastopenia of childhood and iron-deficient states (6,7). In the latter case, treatment with iron supplements was found to significantly reduce the number of spells (7).

Evaluation

Thorough history taking, including a detailed description of the episode, is an essential starting point in the evaluation. In particular, close attention should be paid to the circumstances and sequence of events at the time of the spell. This information can offer important diagnostic clues. For example, most BHS are preceded by agitation and crying, in contrast to seizures, cardiac disturbances, and orthostatic syncope, which often occur with no emotional provocation. Also, without exception, cyanosis or pallor is seen in BHS before other manifestations, such as syncope or postural changes; color change with seizures, if noted at all, tends to occur after loss of consciousness.

In an older child with breath-holding episodes, additional diagnostic clues from the patient's history may include urinary incontinence, which often occurs with certain types of seizures but is uncommon with benign BHS. Also, caregivers report that these spells occur with the child fully awake and alert. This characteristic differentiates BHS from apneic conditions, which generally are linked to sleep states.

Physicians need to elicit information about whether a child has apparent distress with eating or other physical activities or experiences chest pain or other associated physical symptoms. These findings suggest something other than benign BHS, such as an underlying cardiopulmonary problem.

Family history is another key element in patient assessment. Previous observations have noted that perhaps 20% to 30% of children with benign BHS have family members who were similarly affected during childhood (2,8). In addition, data from more detailed analyses suggest an autosomal dominant pattern of inheritance in some cases of severe BHS (8). Also noteworthy, family histories of these children often reveal an increased incidence of syncope in adult family members.

Inquiry should include family history of neurologic disorders as well as cardiac diseases--especially events that occurred early in life or were associated with sudden death.

A complete physical examination should include assessment of the child's general appearance, behavior, growth, and development. The cardiovascular system requires particular attention, including careful auscultation of the heart for murmurs and rhythm irregularities. Likewise, a thorough neurologic evaluation to identify focal deficits is important. A complete blood cell count can identify hematologic abnormalities, and an electrocardiogram can rule out prolonged QT syndrome.

Dmitri has normal exam

If assessment findings are significant or if the diagnosis is still uncertain, additional evaluation may be needed. If history taking and physical examination suggest seizures or other CNS disease, electroencephalographic testing is recommended, and referral to a pediatric neurologist should be considered. If a cardiovascular problem is suspected, it would be appropriate to use a Holter monitor and consult a pediatric cardiologist.

Management

It is most important to assure parents that although BHS are frightening to observe, they are benign and children will outgrow them. Also, parents should be told that evidence suggests no serious long-term effects of benign breath-holding episodes in otherwise healthy children; these patients do not have increased risk of epilepsy or other neurologic problems. The only significant finding on subsequent follow-up of children with BHS was

a mildly increased incidence of syncope later in life, especially in childhood or adolescence (1). Later syncope is rare in children with the cyanotic type of BHS, whereas it occurs in 17% of those with the pallid type (2).

Adequate education of caregivers about BHS and alleviation of their anxiety about these episodes are essential. Because of fear, parents may try to prevent every conflict or minor mishap in a child's life. Such efforts are neither practical nor possible and may lead parents to overindulge their child or to forego appropriate discipline to pacify him or her. Behavior problems may ensue. A thorough understanding of the benign nature of this problem can help parents avoid these pitfalls and improve how they deal with the episodes.

Nevertheless, when a child with BHS becomes upset and cries, reasonable efforts to calm the child should be made. If an episode occurs despite these measures, observation of the child and prevention of injury are generally all that is required. In cases in which a child loses consciousness, the child should be placed in a lateral supine position to help avoid injury and possible aspiration. If a spell occurs while eating and food occludes the oral airway, the airway obviously should be cleared. Other resuscitative efforts are not necessary. Once an episode has resolved, the child should be reassured. However, drawing excessive attention to the event or expressing extreme worry to the child should be avoided.

Medication

Although some patients receive medication for BHS, drugs are not generally indicated. In severe cases with associated bradycardia or asystole or in patients with multiple daily episodes, a 0.1-mg dose of oral atropine three times daily has been found to be effective in preventing BHS (2). Oral theophylline, transdermal scopolamine, and pacemakers also have been used. Anticonvulsants have not proved useful in prophylaxis of benign BHS (2,9). Referral to a pediatrician is recommended in all cases in which pharmacologic therapy is being considered.

Summary

BHS are benign and self-limited. However, they require careful evaluation to exclude more serious problems. Thorough history taking, physical examination, and limited screening studies can establish the diagnosis. Consultation should be considered if the clinical situation warrants. Treatment includes parental reassurance and education about the condition. Patient care involves attempts to calm the child, but if such care proves unsuccessful, observation and protection from injury are generally all that is required during an episode. Medication is not indicated except in severe cases.

→ our practice has excluded more serious problems.

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