CHAPTER 55

BREATH-HOLDING SPELLS

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Breath-holding spells consist of brief apneic episodes provoked by some acute emotional stimulus, such as anger or pain. The child then displays cyanosis or pallor, loss of consciousness, rigidity, and, sometimes, seizure-like twitching. Breath-holding spells usually begin spontaneously in otherwise normal children between the ages of 6 and 24 months and usually stop by the age of 5 years. Every primary-care physician will encounter patients with these common spells.

Pathophysiology

Breath-holding spells consist of a stereotypical sequence of pulmonary and cardiovascular responses to strong, usually adverse, emotions. The cyanotic type of breath-holding spells prevails over the pallid type in a ratio of about 3:1. Vagally mediated cardiac inhibition generally occurs in the pallid type of breath-holding spells, whereas reflex-induced forceful and prolonged expiration causes the cyanotic type. Cerebral anoxia, whether produced by asystole or prolonged apnea, apparently causes the loss of consciousness. Some breath-holding spells may terminate with a true epileptic seizure.

Clinical Features

Breath-holding spells most commonly occur in children who appear physically and developmentally normal, but they can also affect retarded children. Breath-holding spells usually begin between the ages of 6 and 24 months, peaking in frequency by around 2 to 3 years. They may appear numerous times per day, but usually disappear by 5 to 6 years of age. Breath-holding spells affect boys more than girls in a ratio of about 1.3:1. Most children with breath-holding spells seem to have urgent, demanding personalities, although studies have not shown a diagnostic personality profile.

Typically, breath-holding spells display four phases: (1) provocation, (2) expiratory apnea and cyanosis, (3) opisthotonic rigidity (backward arching), and (4) stupor. The provocation consists of some strong physical or emotional stimulus: a fall, anger, frustration, or pain. A breath-holding spell typically starts with crying, lasting 15 seconds or less. If crying lasts much longer, the breath-holding spell may not occur. Abortive breath-holding spells occur, as well as the full tetrad. Sometimes the second phase occurs without the expiratory phase. The second phase consists of sustained, forced expiration, followed by progressive cyanosis in the majority of patients. The child remains aware during the first few seconds of this phase. During this interval, a counter-stimulus or diversion of the patient’s attention may abort an attack. At the onset of the third stage, the rigid stage, the patient becomes restless and then opisthotonic, with strongly extended back, arms, and legs. Cyanosis has become prominent by this time, and the patient has lost consciousness. Some clonic twitches and, sometimes, incontinence may occur. The apneic stage of the attack ends with a gasp or the resumption of quiet breathing. The normal skin color promptly returns, and the patient lies motionless. The patient typically remains stuporous or drowsy for minutes to hours after an attack. Although the whole sequence to the stage of stupor lasts only a few minutes, anxious parents may estimate the time to be much longer, a matter to consider when eliciting the history. If in doubt, ask the parents to time an episode with a watch. A family history of breath-holding spells in 25 to 35% suggests an autosomal dominant pattern of inheritance with reduced penetrance. Whether this means a genetic predisposition to autonomic instability remains unknown. Because the child’s parents often do not know whether they themselves have had breath-holding spells, questioning of the child’s grandparents may lead to more accurate information.
Diagnosis and Differential Diagnosis

The diagnosis, as in migraine or any such episodic disorder, rests squarely on the quality of the history. In particular, the examiner has to determine that an emotional event has triggered each attack. Table 55-1 outlines the specific criteria for the diagnosis of breath-holding spells.

The common differential diagnoses include epilepsy, congenital heart disease, vasovagal syncope or hyperactive carotid sinus, prolonged QT interval, and posterior fossa or brainstem lesions. Breath-holding spells generally feature arrest of breathing after prolonged expiration, which distinguishes them from voluntary breath-holding, in which the person arrests breathing after inspiration. The fact that breath-holding spells rarely appear before 6 months of age serves to differentiate them from neonatal apnea and other apneic disorders of early infancy. The normal history and physical findings should readily differentiate congenital heart disease, with syncope or cyanosis precipitated by exercise. Syncopal attacks from ordinary causes in older patients rarely occur in the range of onset of breath-holding spells (6 to 24 months). Vasovagal syncope or carotid sinus hypersensitivity should be investigated by combined electroencephalographic-electrocardiographic monitoring, including eyeball compression and carotid sinus massage.

Exclusion of syncope leaves epilepsy as the major differential diagnosis. The typical full tetrad (provocation by emotion, expiratory apnea and cyanosis, opisthotonic rigidity, and stupor) readily differentiates breath-holding spells from typical epileptic attacks. Unfortunately, not all patients with epilepsy or breath-holding spells have typical attacks. The most difficulty arises with rigid or akinetic seizures, in which the patient may exhibit no overt convulsive movements. These attacks sometimes seem to follow provocations of the type that induce breath-holding spells. The correct diagnosis may require recording a spell by continuous video electroencephalogram (EEG) monitoring. Table 55-2 summarizes the differential diagnosis of breath-holding spells and epilepsy.

Course and Prognosis

Patients 2 to 3 years of age usually display the most frequent breath-holding spells. The attacks then gradually grow fewer and usually disappear by 5 to 6 years of age. Despite having many cyanotic attacks per day for several years, the patients do not seem to suffer overt or cumulative brain damage. No specific, long-term psychiatric sequelae are known.

Management

During an attack, parents should place the patient in a lateral recumbent position but should generally avoid cardiopulmonary resuscitation. Usually the history suffices to secure the diagnosis, but some authors recommend routine electrocardiograms (ECGs) to check for a prolonged QT interval. Resolution of the diagnosis in a few patients may require combined ECG and video EEG monitoring, with ocular compression or other autonomic tests.

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Breath-Holding Spells</th>
<th>Epilepsy</th>
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<tbody>
<tr>
<td>Pregnancy, birth, and physical examination</td>
<td>Usually normal</td>
<td>Frequently abnormal, particularly with soft or hard neurologic signs</td>
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<tr>
<td>Family history</td>
<td>No history of epilepsy but may have history of breath-holding spells</td>
<td>Epilepsy common in near relatives</td>
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<tr>
<td>Trigging</td>
<td>Always triggered by an emotionally provocative event</td>
<td>May be triggered by fever, sound, or light or occur spontaneously</td>
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<td>Evolution of events</td>
<td>Follows typical tetrad of provocation, opisthotonic rigidity, and stupor; cyanosis precedes loss of consciousness</td>
<td>Usually has aura and early loss of consciousness; often shows clear tonic-clonic phase or other specific seizure pattern</td>
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<tr>
<td>Electroencephalogram</td>
<td>Nonepileptiform</td>
<td>Usually epileptiform</td>
</tr>
<tr>
<td>Response to antiepileptic medication</td>
<td>None</td>
<td>Frequent response</td>
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ECG = electrocardiogram; EEG = electroencephalogram; MRI = magnetic resonance imaging.
After having secured the diagnosis of breath-holding spells, the physician should thoroughly explain their benign nature to the parents. First of all, the physician should realize that the oft-recited advice merely to ignore the spells does not suffice. The spells frighten the parents, make them very anxious, and negatively affect their relationship with their child. The physician should educate the parents as to the involuntary, reflexive or autonomic nature of breath-holding spells, that the patient does not willfully and by malicious intent induce a spell to seek attention, and that the spells do not signify a flawed child with a serious physical or mental illness. After understanding the benign nature of breath-holding spells, the parents can usually cope with them and can learn to abort some spells by distracting the patient when provocative emotional stimuli arise. No known behavioral technique controls breath-holding spells, certainly not punishment.

Although the vast majority of children do not require any medication, severely affected patients may respond to piracetam. Antiepileptic drugs are not effective. Pallid breath-holding spells may respond to atropine when the ocular compression test shows significant asystole during ECG monitoring. Very rarely pacemaker implantation is indicated. Reduction of the breath-holding spells may follow correction of any underlying systemic illnesses. Evidence now suggests that iron-deficiency anemia occurs in greater frequency in patients with breath-holding spells than in normal control subjects, and correction may eliminate the breath-holding spells.

**Suggested Readings**


