G94 Breath Holding Spells Associated With Unexpected Sudden Childhood Death

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After attending this presentation, attendees will be able to recognize a medical condition that may account for sudden death in childhood (greater than 12 months old).

Breath holding spells (BHS) have traditionally been thought to be benign–"something that the patient will outgrow." However, clinical monitoring during some of these "spells" has resulted in the documentation of both severe bradycardia and dispersed QT intervals. A recent study reported ten patients with significant bradycardia that required permanent pacemaker placement.

The authors report the case of a 20-month-old Caucasian male, born by induction via membrane rupture and the use of pitocin at the gestational age of 38 weeks with APGAR scores of 7/8. He was noted to be a "slow starter," requiring vigorous sternal rubs to facilitate normal vital signs. In addition to this he had temperature instability that required his transfer to the Special Care Nursery for monitoring and isolette placement to maintain his temperature. He remained in the hospital for six days after his birth. After discharge he had what his parents described, and his doctor related, as "breath holding spells," occurring two to three times a week. These did not alarm his parents because his older sister also suffered from them. His spells were provoked by crying or coughing, and after those activities he occasionally had a quiet period with apparent apnea. These spells were associated with both pallor or cyanosis that could last for five to ten seconds. Multiple 911 calls were made when the apneic spells progressed into seizure activity lasting more than 30 seconds. At the age of seven weeks, following a coughing episode with an emergency room evaluation, he was found to have oxygen saturation of 88 to 89 percent. While the phlebotomist was drawing his blood he became apneic. He was given supplemental oxygen and became more alert. A follow up chest x-ray revealed a slightly enlarged cardiac silhouette. A subsequent echocardiogram was normal. He was worked up for gastroesophageal reflux disease and placed on omeprazole. In the last four months of his life he had a marked decrease in these episodes.

On the day of his death, while under the care of his aunt, he was placed in his crib after his lunch meal. No articles of potential respiratory compromise were in the crib. He awoke with a cry after his nap, and when his caregiver checked on him 20 minutes later, he was found unresponsive and a 911 was called. Attempts at resuscitation were unsuccessful.

Postmortem exam revealed no anatomic cause of death. Toxicology, blood cultures, histology, postmortem radiographs, and vitreous electrolytes were unremarkable. Detailed cardiac pathology was normal.

Breath holding spells are a frequently observed event in infancy and early childhood. Their association with sudden and unexpected death is rare. Typical cases begin between six and twelve months of age, and rarely last past age four. Breath holding spells have been associated with pallor and/or cyanosis, and severe cases involving convulsions have been described. Several causes and explanations have been proposed, but proof of etiology is not found in most cases. Autonomic dysfunction and paroxysmal vagal overactivity have been felt to play a significant role.

The death of this child represents a case of paroxysmal vagal overactivity with a fatal outcome. While rare, when the history is consistent with this premorbid diagnosis, and no alternative explanation is found, this cause of death should be a consideration.

Breath Holding Spells, Paroxysmal Vagal Overactivity, Sudden Unexpected Death in Childhood

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