BREATH-HOLDING SPELLS: AN ANALYSIS OF 43 CASES

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Abstract:
Objective:
Breath holding spells, very often misinterpreted as epileptic seizures, are most common in children aged 6 months to 6 years of age. In this investigation, we sought to prospectively document the natural history of breath holding spells (BHS) among children with cyanotic, pallid and mixed type BHS referred for neurological consultation.
Materials & Methods:
This was a cross-sectional study in which a total of 43 children (23 boys, 20 girls) with BHS, admitted to the out patient clinic of the Children's Hospital Medical Center, between Sept 1998 and June 1999, were enrolled. A structured interview was under taken at the time of initial consultation to confirm BHS and its type, associated phenomenon, family history, sex and age at initiation of spells. Laboratory, electroencephalographic and electrocardiographic tests were done.
Results:
Patients were between 1.7 and 42.8 months (mean age 18.4 months). In 76.8% of cases, BHS began during the first 12 months of age. Anger and pain were the most common triggering factors (65.1 %). A positive family history of BHS was identified in 51% and parental consanguinity was found in 30% of cases. The spells were cyanotic in 79.1% (34 children). 78% of cases were iron deficient and 53% of cases had iron deficiency anemia.
Conclusion:
The results of this study emphasize the role of genetic factors in BHS; measurement of hemoglobin and serum ferritin is recommended all such cases.
Key words: Breath-holding spells, Iron deficiency, Anemia, Serum ferritin

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Introduction

followed by seizure-like activity are defined as complicated BHS, and is seen in 15% of patients (4). BHS are classified as cyanotic, pallid and mixed types. Cyanotic BHS constitutes 60% of cases. After a cry or an attempt to cry, breathing is interrupted during expiration and the child becomes cyanotic, limp and loses

consciousness for a brief period of time. Pallid BHS constitutes 20% of cases; in whom after a little crying a significant bradycardia or asystole occurs, producing the pale color and loss of consciousness. These episodes are often frightening to parents and worrisome to physicians (1,2,3). BHS usually begins between 6-18 months of age (1,2,3,4,5). Approximately 7% of BHS begin during the first month of life (4). Based on several studies, BHS are self-limited and usually outgrown by school age (1,2,3,4,5). They, however, require careful evaluation to exclude more serious problems .In the past attempts have been made to implicate psychological factors in triggering BHS. Previous reports have linked BHS with iron-deficient states and iron therapy is effective in treatment (6,7,8,9,10). Therefore, in this 9 month cross-sectional study we assessed the clinical and epidemiologic aspects of BHS.

Materials & Methods

47 children with diagnosis of BHS, admitted to the out patient clinic of Children's Hospital Medical Center, between Sept 1998 and June 1999, were included in this cross- sectional study. Diagnosis of BHS was based on by medical history of apnea, unconsciousness and change in postural tone triggered by adverse stimuli.The patients were examined by a pediatric neurologist. Para-clinic examinations consisting of CBC, FBS, Ca, serum ferritin, EEG and ECG were done for all patients. Patients with previous history of epilepsy, developmental abnormalities and neurologic disorders were excluded from the study. FBS of less than 50 mg/dl, Ca of less than 8 mg/dl, corrected QT interval of more than 0.44 seconds and EEG with paroxysmal epileptiform discharges were para clinical exclusion criteria of this study. 4 cases were excluded because of abnormal EEG. According to history and color of the patient during the spells, cases classified as cyanotic, pallid and mixed type. In the cyanotic spells after a short period of crying, breathing is interrupted during expiration and the child becomes cyanotic, limp and then loses consciousness. In the pallid spells after a little or no crying, the patient loses consciousness with pallor. Patients with history of previous cyanotic and pallid type or both pallor and cyanosis in one spell, were classified as mixed type BHS. We analyzed data using SPSS statistical soft ware version 7.5.

Results

During a 9-month period, 47 patients with the diagnosis of BHS were enrolled in the study. 4 patients were excluded because of abnormal EEG, and 23 boys and 20 girls completed the study (1.15:1). Mean age of the children was 18.4 months (1.7-42 months) with 76.8 % presenting younger than 12 months of age (Table I).

Table I: Distribution of age in the patients

Age (months)	Number	Percent
0 – 6	14	32.6%
7 – 12	19	44.2%
13 – 24	8	18.6%
After 24	2	4.7%
Total	43	100

Parental consanguinity was found in 30.2 % of cases. 22 patients had family history of BHS (Table II).

Table II: Clinical and epidemiological findings

Parameters		Number	Percent
Sex	Boy	23	53.5
	Girl	20	46.5
Family history	Yes	22	51.2
	No	21	48.8
Parent consanguinity	Yes	13	30.2
	No	30	69.8
Type of attack	Cyanotic	34	79.1
	Pallid	5	11.6
	Mixed	4	9.3
Triggering factors	Anger	28	65.1
	Pain	28	65.1
	Head bump	20	46.5
	Frustration	18	4.1
	Fear	4	9.3

Serum calcium and glucose were found to be within normal limits for all patients. 78 % of patients had iron deficiency according to serum ferritin levels. 53 % of cases had iron deficiency anemia with mean hemoglobin value of 10.4±1.56g/dl. Normal electrocardiogram ruled out prolonged QT syndrome. All of the patients had normal EEG recording. Triggering factors of BHS are shown in Table 2. 34 cases had cyanotic episodes (79.1 %), 5 had pallid spells (11.6 %) and 4 had the mixed type of spells (9.3 %).

Discussion

BHS is a common non-epileptic paroxysmal event that mimics epilepsy and is very worrisome for parents. BHS were observed more frequently in boys than in girls (M/F1.15:1). Male to female ratios of 3:1, 1.7:1 and 1.3:1 have been reported in other studies (1-5). In a group of 384 children studied by Livingstone, the mean age at onset was 12 months (3 months to 4 years) (2). Age of our patients was between 1.7 to 42.8 months. BHS began during the first year of age in 76.8% of cases. In an Indian study as reported by Bhatia, 80% of cases experienced BHS during the first 18 months of age (12). Data from more detailed analyses suggest an autosomal dominant pattern of inheritance in some cases of severe BHS (1,2,3,11). Family histories of BHS were found in 51.2 % of our cases, in comparison with 47.5 % of the Daoud study (7). Parental consanguinity was found in 30% of our patients in comparison with 70% of Daoud study (7). BHS are provoked by frustration, anger, fear or pain (1-7). In our study anger and pain were the common triggering factors (65.1%). In agreement with previous reports, the most common type was cyanotic spells (1,2,3,4,5,7). Based on a number of recent studies it is probably worthwhile checking for iron deficiency or anemia, and to perhaps treat the child with an iron supplement (5-11). 78% of our cases were iron deficient and 53% had iron deficiency anemia. The limitation of this study, i.e. the small number of patients and the results could be confirmed in larger randomized controlled trials. The results suggest new data concerning the natural history of BHS have been determined. The information is important for family counseling and identifying intervention strategies, and could serve as baseline data to evaluate the efficacy of future treatment approaches. Iron deficiency and anemia should be checked in patients presenting with BHS. It appears that treating iron deficiency anemia is effective in reducing the frequency of BHS.

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