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# BREATH HOLDING SPELLS IN EARLY INFANCY

**Masahiko KIMURA, Yoshiko RYOKE, Satomi MIYAMOTO, Makoto INOUE, Hitoshi SEJIMA and Seiji YAMAGUCHI**

*Department of Pediatrics, Shimane Medical University, Izumo 693, Japan*

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**We report two patients of breath holding spells (BHS) within the first two months of life presenting with apparent life threatening events, characterized by respiratory suppression and bradycardia. Although breath holding spells have been generally considered a benign phenomenon, they could cause a critical condition in early infancy due to the severe respiratory suppression. The pathophysiologic mechanism of BHS has remained unknown, respiratory suppression recognized in our patients might be related to immaturity of the autonomic function. We conclude that breath holding spells in early infancy should be taken account as a cause of apparent life threatening or even sudden infant death syndrome.**

Key words : Breath holding spell / Autonomic dysfunction / Apparent life threatening event / Sudden infant death syndrome

Breath holding spells (BHS) are generally regarded as a common and benign phenomenon in childhood (1). DiMario (1) noted that the term breath holding is a misnomer that implies a voluntary action resulting in a prolonged inspiration, and in actuality these episodes are involuntary and reflexive and occurring during active or full expiration. BHS are relatively rare, but present within the first two months of life. Apparent life threatening event (ALTE) is defined as an episode that is frightening to the observer and is characterized by some combination of apnea (central or occasionally obstructive), color change (usually cyanotic or pallid but occasionally erythematous or plethoric), marked change in muscle tone (usually marked limpness), choking or gagging (2). ALTE has heterogeneous causes, but there have been few reports on the relationship between BHS and ALTE or sudden infant death syndrome (SIDS). We hereby report two cases of BHS and discuss whether BHS cause ALTE or SIDS in early infancy.

## CASE REPORT (Table 1)

Patient 1, a 28-day-old girl, with unremarkable prenatal and perinatal histories, began to cry abruptly while she was asleep, and became cyanotic and flaccid under the condition of apnea. She was brought to a hospital where her respiratory condition recovered spontaneously. After drawing blood sample for laboratory tests, another attack occurred with vigorous crying. When she was referred to our hospital 5 hrs after the onset, she showed the generalized hypotonia and intermittent apnea followed by bradycardia. These conditions lasted for 3 hrs. Then, the respiration became stable and she could start feeding 5 hrs after the admission.

Patient 2 was a 39-day-old boy with no special problem in delivery and neonatal period. He stopped breathing and became cyanotic when he woke up with vigorous crying. Soon after that, he was brought to a private doctor and an intratracheal intubation was performed because of his bradycardia and weak respiration. No obstruction such as milk to the larynx was found. When referred to our hospital 2 hrs later, he began to move actively and could be extubated 4 hrs later. Bottle feeding was started 6 hrs after the onset.

Both cases had no preceding infection or the history of apnea. The routine laboratory tests including complete blood cell counts, biochemical tests, chest X-ray and electrocardiogram were evaluated to be normal as shown in Table 1. The second chest X-ray taken after the extubation in Patient 2 showed atelectasis of the right upper lobe without any clinical symptoms. The echocardiogram (ECG), brain magnetic resonance imaging (MRI) and electroencephalogram (EEG) exhibited no abnormalities. The urine organic acid analysis, and amino acid analysis in blood and urine were also normal in both patients. The 24-hr holter electrocardiogram recording performed after the complete recovery in Patient 1 was uneventful. Barium esophagography was normal in Patient 2.

Table 1. Clinical findings of the 2 cases

| | Patient 1 | Patient 2 |
|---|---|---|
| onset age (days) | 28 | 39 |
| prenatal and neonatal history | unremarkable | unremarkable |
| preceding infection | none | none |
| conditions during and after BHS | unstable respiration and hypotonia lasting for 2 hours | intratracheal intubation, extubated 3hours later |
| timing of oral feeding after BHS | 5 hours later | 5 hours later |
| noxious stimuli | BHS induced | BHS not induced |
| recurrence of BHS | once at the age of 2 months none after that | none |
| development at the age of 1 year | normal | normal |
| laboratory findings | | |
| hemoblobin (g/dl) | 13.7 | 10.5 |
| CRP (mg/dl) | <0.3 | <0.3 |
| biochemistry | normal | normal |
| metabolic screening tests* | unremarkable | unremarkable |
| chest X-ray, EKG | normal | normal |
| brain CT, MRI | normal | normal |

BHS: breath holding spells
* including amino acid analysis, urinary organic acid analysis, blood lactate and pyruvate, and free thyroxine and thyroid stimulating hormone

Correspondence : Masahiko Kimura, M.D. Department of Pediatrics, Shimane Medical University, 89-1 Enya, Izumo, Shimane 693, Japan. Tel+81.853.23.2111(ext.2715) Fax+81.853.23.4009.

Patient 1 had another episodes of BHS provoked by drawing blood sample at the age of two months. Both patients showed a normal physical examination and development at the age of 1 year.

## DISCUSSION

The diagnosis of BHS was most likely in both the patients based upon the clinical course and findings of stopping breathing and cyanosis after crying. In particular, Patient 1 had a typical history of breath holding which was induced by noxious stimuli of drawing blood sample. BHS generally occur in the first 12 months, and considered a benign and transient phenomenon. It was reported that 5% of BHS patients had spells in the neonatal period (3) and 8% during the first 3 months of life (4). In both our patients, the onset age was around one month. Their BHS were characterized by the irregular respiration associated with bradycardia in Patient 1, and the bradycardia and respiratory suppression in Patient 2. Although the clinical condition had been critical a few hrs after the spell, the oral feeding could be started 6 hrs after the onset.

BHS rarely have been reported to be fatal (5,6). On the other hand, Southall *et al*. (5) reported that 276 of their 9856 cohort patients suffered from cyanotic breath holding, and 2 of 276 or 0.7% of children with cyanotic breath holding were likely to have a fatal outcome. They mentioned that the risk of sudden death in patients with BHS appeared to be very low, but the BHS episodes in infants less than 6 months of age or in those with frequent loss of consciousness should be concerned to be a risk factor in sudden death (7). The pathophysiologic mechanism of BHS has remained unknown. There is a hypothesis that BHS are provoked by such as noxious stimuli, and then an initially excessive sympathetic response is followed by an exaggerated rebound parasympathetic (vagal) response (8). The exaggerated parasympathetic response could suppress the respiration and sympathetic activity of the heart. DiMario *et al*. (8) reported that children with severe cyanotic BHS had underlying autonomic nervous system dysregulation, and showed a significant decrease of diastolic blood pressure and abnormal R-R interval ratio during changing posture from lying to standing. In our patients, who were around one month of age, the respiratory suppression after BHS would relate to immaturity of the autonomic function. BHS are usually no longer present after the age of 6 (1). Therefore, BHS patients underlying immaturation of the nervous system including both the autonomic and central nervous system, especially brainstem, should be considered to give rise to a severe condition such as ALTE or SIDS.

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