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SYNCOPE, ASYSTOLE AND ATRIOVENTRICULAR BLOCK IN A CHILD WITH BREATH-HOLDING SPELLS: A CASE REPORT

L.M.Makarov[1,2], V.N.Komolyatova[1,2], I.I.Kiseleva[1], A.V.Dmitrieva[1]

[1]FSCC Children and Adolescents of the FMBA of Russia, Russia, Moscow, 20 Moskvorechye str.;

[2]FSBEI of Additional Professional Education "Russian Medical Academy of Continuous Professional Education" of the MH RF, Russia, Moscow, 2/1 Barrikadnaya str., buil. 1.

A case of a child with the breath-holding spells (BHS), atrioventricular block and long pauses of heart rhythm till 12 sec is presented. The attacks began at 1 year and completely stopped at 3 years. A typical ECG pattern for BHS is identified. The issues of therapy and the need for implantation of pacemaker are discussed.

Key words: breath-holding spells; atrioventricular block; children; arrhythmia in children; syncope; asystole; pacing

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Corresponding author: Makarov Leonid, E-mail: dr.leonidmakarov@mail.ru

L.M.Makarov - ORCID ID 0000-0002-0111-3643, V.N.Komolyatova - ORCID ID 0000-0002-3691-7449, I.I.Kiseleva - ORCID ID 0000-0002-4867-0594, A.V.Dmitrieva - ORCID ID 0000-0002-0314-5235

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Syncope is a common occurrence in children [1]. According to the EPISODE study [2], up to 4% of Russian children have experienced episodes of syncope during their lifetime. The prognosis and management of a child with syncope are determined by identifying the specific mechanism underlying the event. We present a case involving a combination of syncope, prolonged asystole, and atrioventricular (AV) block in a young child.

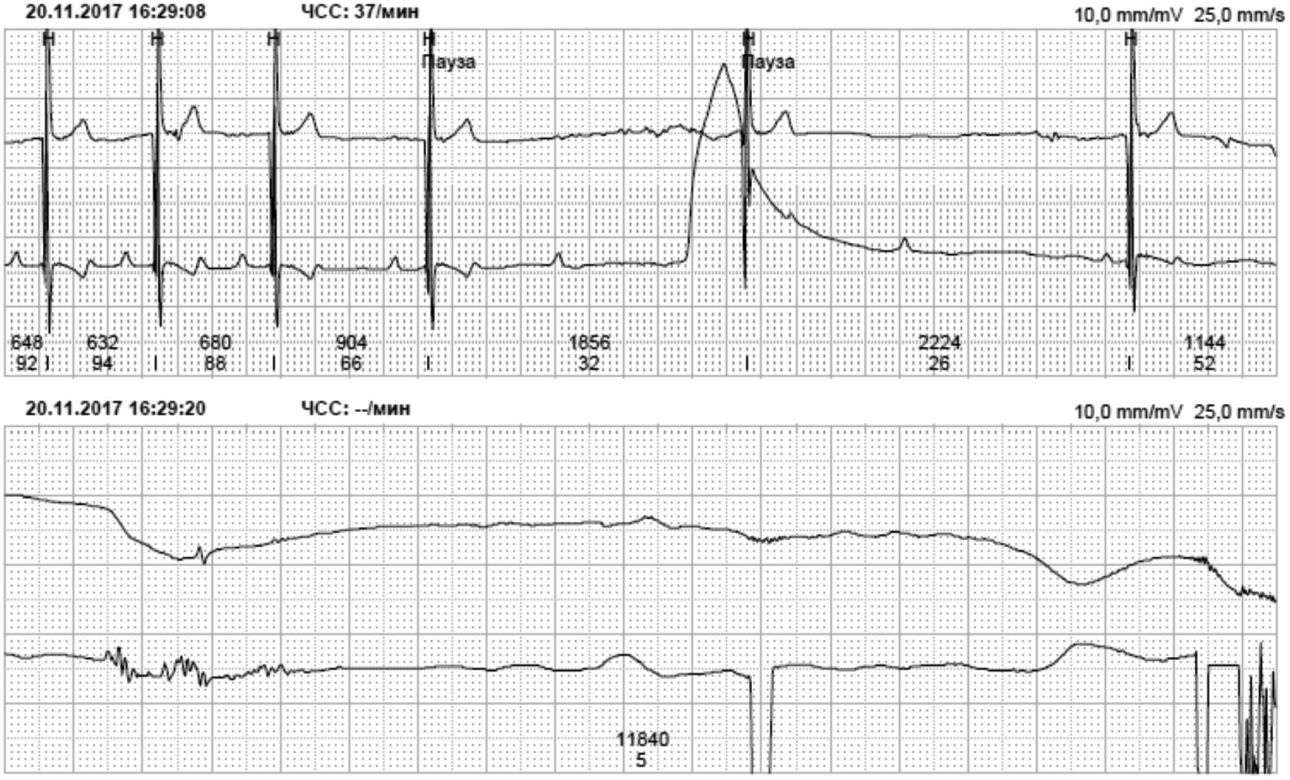

Figure 1. Child M., 1 year 6 months old (20.11.2017), at 16:29, during agitation and heart rate of 45 bpm, a sudden rhythm pause of 11,840 ms occurs, accompanied by loss of consciousness. During the pause, transient first-degree AV block with PR prolongation to 180 ms is observed, progressing to advanced AV block (top panel) and subsequent absence of atrial contractions as asystole prolongs (bottom panel).

Clinical case description

We observed a six-year-old child, M.D. The child was born at term from the third pregnancy and third full-term delivery, with two healthy siblings. The pregnancy and delivery were uncomplicated, and the child's development proceeded according to age. At the age of 1 year and 1 month, episodes of loss of consciousness began. These episodes occurred during the day and were consistently triggered by negative emotions or painful stimuli (e.g., a fall during play). During heightened emotional distress, the child would suddenly pale and "collapse" during inhalation. The syncope lasted up to one minute, with a frequency ranging from weekly to several times a day. There were no seizures during the episodes, although occasional involuntary urination was noted. The child recovered independently, with clear consciousness, recognizing the mother and surroundings immediately afterward, without drowsiness. Skin color returned from pale to normal pink.

The child was evaluated by a neurologist, and epilepsy was ruled out. Blood tests revealed no abnormalities, including no anemia. Holter monitoring (HM) conducted earlier recorded prolonged rhythm pauses of up to 8 seconds during these episodes. At the age of 1 year and 6 months, the child was referred to a cardiac surgery center, where hospitalization for pacemaker implantation was recommended. The following day (13 November 2017), the child's mother sought our consultation.

At the consultation, the child weighed 11.5 kg and measured 83 cm in height, with a proportional body structure. A primary physical examination revealed no abnormalities across systems, and there were no dysmorphic features. The heart's borders were normal, with no pathological murmurs. Blood pressure was 85/45 mmHg. A 12-lead electrocardiogram (ECG) showed a heart rate (HR) of 134 bpm, an electrical axis of 75°, a PR interval of 0.12 seconds, and a QT interval of 280 ms (QTc 395 ms). All ECG parameters were within the normal range for age [3]. Echocardiography showed no pathology, including no heart defects, cardiomyopathies, chamber dilation, or valvular abnormalities.

During HM (20 November 2017), two typical episodes were recorded while the child was awake and experiencing negative emotional reactions. These episodes began with increasing sinus tachycardia (150-160 bpm), progressing to brief rhythm slowing (62-34 bpm), first-degree AV block, and sinus node arrest lasting up to 11,840 ms (Fig. 1). Three additional clinical episodes without loss of consciousness were recorded, displaying similar ECG patterns with rhythm pauses of up to three seconds. Several episodes of transient first-degree AV block with a maximum PR interval of 0.3 seconds were also noted.

Based on the typical clinical presentation, the diagnosis was established as pallid-type breath-holding spells (BHS), cardioinhibitory variant. A comprehensive therapeutic regimen recommended for children with BHS was implemented [4-7], including piracetam (30-50 mg/kg/day), iron supplements (3 mg/kg/day) as advised regardless of anemia presence, belladonna, and beta-blockers (propranolol, 1 mg/kg/day) to prevent reflex syncope triggered by increasing sinus tachycardia [8]. However, clinical improvement was not achieved, leading to a decision to discontinue medication.

Repeated HM consistently demonstrated the same ECG changes during episodes, which we termed the "clinical-electrocardiographic pattern of BHS" (Fig. 2). Rhythm pauses were characterized by either a clean isoelectric line or a rhythm with episodes of atrial activity (second-degree AV block with 3:1 conduction, as seen in Fig. 1). Throughout the observation period, transient first- and occasionally second-degree AV block episodes were regularly recorded on HM. However, the duration of pauses during AV block was not associated with clinical manifestations of BHS.

Since no effect was achieved with the administered medical therapy, the necessity of implanting a pacemaker (PM) was repeatedly considered. However, the primary clinical symptom remained breath-holding spells (BHS), occurring at a typical age, leading us to conclude that the child's prognosis was generally favourable. The diagnosis of sick sinus syndrome (SSS) was ruled out based on the typical clinical presentation, asystole occurring only during wakefulness, the absence of nocturnal bradycardia, and high heart rates during the day.

Evaluating the opinions of leading experts on this issue, we noted that the question of pacemaker implantation regularly arises in children with BHS and prolonged asystole. This decision is made on an individual basis, depending on the treatment protocols adopted in a particular clinic, as well as the preferences of the parents and the

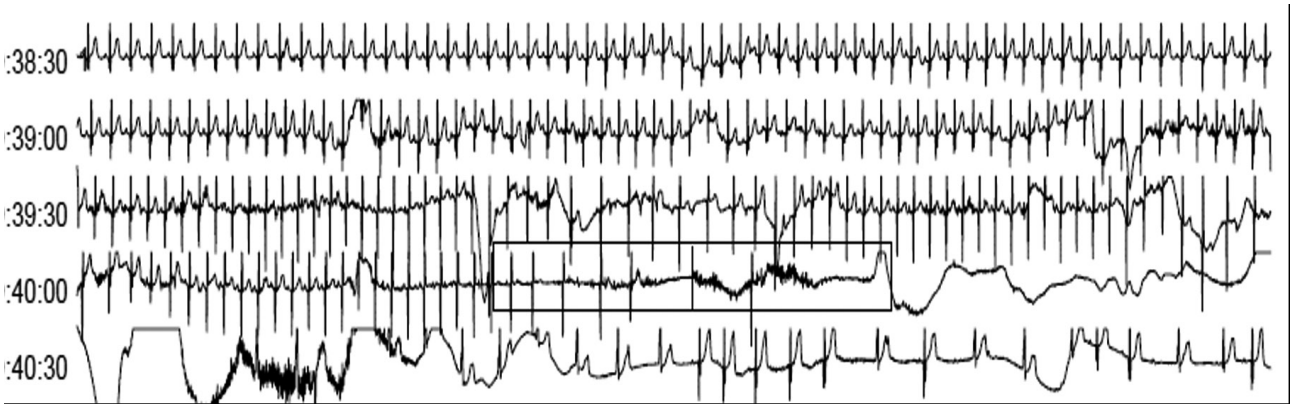

Figure 2. "Clinical and Electrocardiographic Pattern of BHS" in child M.D., 2 years and 3 months old, during Holter monitoring. A progressive sinus tachycardia develops in response to the child's negative emotional reaction, transitioning to a sharp rhythm deceleration during breath-holding and a prolonged pause (highlighted in rectangle), leading to syncope and subsequent gradual spontaneous rhythm recovery.

attending physician [9-13]. Based on our own experience with children with BHS and the recommendations of the European Society of Cardiology on syncope management [1], we opted against PM implantation.

At the age of 3 years, the patient's episodes completely ceased. During the latest examination (17 July 2024), when the child was 6 years old, the following ECG parameters were recorded: heart rate (HR) of 110 bpm (normal up to 105 bpm), an electrical axis of 78°, PR interval of 0.18 seconds (normal up to 0.14 seconds), and a QTc interval of 435 ms using Bazett's formula (normal up to 440 ms) and 393 ms using Fridericia's formula (normal up to 430 ms) [3]. Holter monitoring (17 July 2024) showed a mean 24-hour HR of 90 bpm (normal 79-91 bpm) with a normal circadian rhythm profile (circadian index of 1.31). Almost continuously (except during sinus tachycardia with an HR above 125-130 bpm), first-degree AV block was recorded, with a maximum PR interval of 0.3 seconds during the night.

During nocturnal sleep, there were 209 episodes of second-degree AV block, Mobitz type I, with a maximum rhythm pause of 1981 ms (Fig. 3). At peak heart rates, AV conduction was intact, with a PR interval of 0.12-0.13 seconds. The average daily QTc interval was within normal limits (429 ms). Heart rate variability was moderately reduced.

We consider the prognosis for this child regarding the development of life-threatening bradyarrhythmias to be favourable without PM implantation. However, continued monitoring is essential given the persistence and moderate progression of the AV block. During follow-up, it is necessary to exclude conditions associated with progressive conduction system disorders (e.g., by analysing family history and performing serial ECG assessments of the patient and family members). No medication is currently prescribed.

DISCUSSION

Breath-holding spells, classified under ICD-10 code R06, occur in early childhood and are characterised by sudden cessation of breathing, often accompanied by loss of consciousness and, occasionally, seizures. BHS typically develop in response to negative emotional stimuli or painful irritation. In neurology, BHS are categorised as "generalised tonic or tonic-clonic paroxysms of a non-epileptic nature." The term has numerous synonyms, including reflex anoxic seizures, non-epileptic vagal attacks, anoxo-asphyxial seizures, and affective-respiratory paroxysms [1, 4, 9, 14, 15].

The prevalence of BHS in the population varies, but most sources cite a frequency of up to 5% [14, 15]. Episodes typically begin between 6 and 18 months of age [15]. Less than 10% of cases develop after the age of 2 years. The frequency of episodes ranges from daily to once a year, but most children experience one to six episodes per week [9, 15]. The incidence of BHS tends to decrease with age: by 4 years, 50% of children no longer experience BHS, and episodes generally cease by the age of 8 [9, 10, 14, 15].

BHS are classified into "blue" and "pale" types based on skin colour changes during episodes. Cyanotic episodes ("blue" BHS) are more common, occurring in 52% of cases, while 28% of children are diagnosed with the "pale" type, and the remainder exhibit mixed features [9]. Prolonged rhythm pauses are characteristic of the "pale" type, usually triggered by sudden fear, pain, falls, or minor injuries [9, 15]. The occurrence of asystole in this variant is associated with a reflexive increase in the vagal sensitivity of the sinus node [14, 15]. Nearly all children with the pale variant of BHS exhibited asystole lasting over 20 seconds during the Aschner test, an effect absent in the control group and eliminated with atropine administration [15].

Long-term follow-up of 70 children with BHS by D.D. Korostovtsev [15] revealed no association between BHS and sudden death. Thirty-five children were monitored into adolescence (ages 7-12 years). While prolonged asystole of 20 seconds or more was noted in cases of the pale variant, none of the children experienced sudden death. Intellectual and psychological development of children with BHS did not differ from that of the control group, indicating that frequent syncopal episodes and asystole do not result in chronic cerebral hypoxia.

Long-term follow-up of children with early-life BHS showed that 60-75% may develop asthenic syndrome, 10-15% suffer from hysterical neuroses and sleep disturbances, and up to 10% experience reflex syncope and migraines. Only 2.4% of children with epilepsy had a history of BHS [15]. We monitored 14 children with a full clinical presentation of BHS and another 13 with a preschool history of BHS. None required pacemaker implantation, and all exhibited a favourable prognosis, with symptoms resolving by 6-7 years of age [16].

The cornerstone of BHS therapy is parental education, ensuring parents understand the benign nature of these episodes and the normal intellectual development of

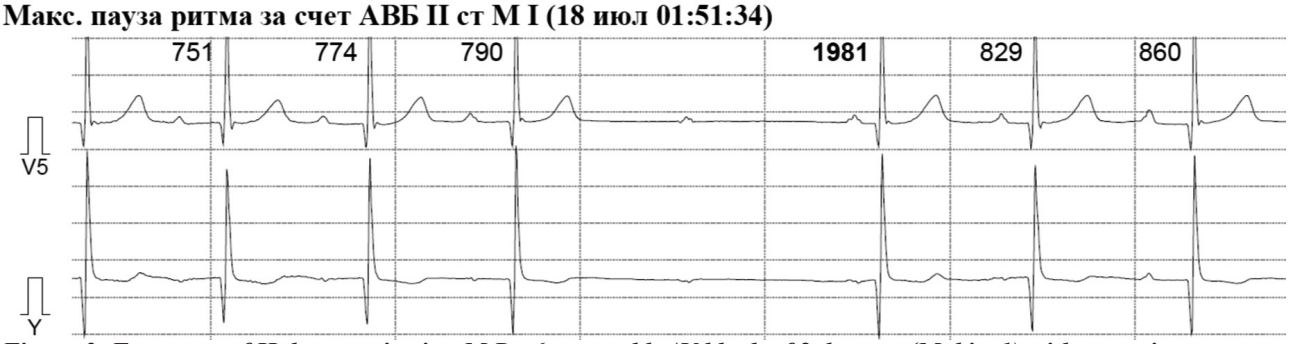

Figure 3. Fragment of Holter monitoring M.D., 6 years old. AV block of 2 degrees (Mobitz 1) with a maximum rhythm pause of 1981 ms.

their child [15, 17]. There is no standardised pharmacological treatment for BHS; however, several studies and guidelines suggest the effectiveness of certain methods. Commonly recommended treatments include piracetam, belladonna preparations, iron supplements (regardless of serum iron levels), and vagolytics [4-7]. Children with BHS are consistently monitored by neurologists and rarely raise concerns about life-threatening risks, as syncopal episodes always resolve spontaneously, even in cases of prolonged asystole and apnoea.

With the increasing use of HM in the evaluation of children with BHS, cardiologists have questioned whether prolonged pauses in heart rhythm in BHS increase the risk of sudden death and whether pacemaker implantation (PI) is necessary. From a traditional arrhythmology perspective, symptomatic asystole associated with loss of consciousness is an unequivocal indication for PI as a means to prevent sudden death in children [18]. However, this approach is justified in cases of true sinus node dysfunction or AV block of organic origin. In children with BHS, the "positive" effects of such therapy have been periodically reported, including a reduction in the frequency and duration of syncope [19-21].

On the other hand, S. Sartori [22], analysing 47 publications on the effectiveness of PI in children with BHS, demonstrated that while PI significantly reduces the duration of asystole in such cases, it is associated with technical issues in 25.7% of cases and medical complications in 11.4%. Importantly, no evidence was found linking BHS to sudden death. Although sudden deaths in children with BHS have been reported, they were attributable to other causes, including long QT syndrome, postoperative tracheoesophageal fistula, spindle-shaped dilation of the upper oesophagus, bronchopneumonia, progressive cerebral atrophy, brain glioma, and craniofacial malformations such as cleft palate [23, 24].

Recommendations to implant a pacemaker may appeal to parents and family members distressed by the frequent and seemingly "endless" severe paroxysms their child experiences. However, the European Guidelines on the Management of Syncope specify that BHS are classified as reflex syncope specific to infancy and childhood. Even in the presence of prolonged asystole, pacemaker implantation should be avoided due to the transient nature of these episodes and the favourable prognosis [1]. Similarly, we believe it is unjustifiable to implant a permanent pacing system in a child aged 1-3 years, given the high likelihood of resolution of non-life-threatening episodes within a few years and the potential complications associated with PI in this age group. We found no documented cases of lead extraction in children after the resolution of BHS.

The complexity of our case lies in the presence of a mildly progressive AV block, which is atypical for isolated BHS. While we cannot rule out its progression to symptomatic stages requiring PI in the future, this underscores the necessity for dynamic follow-up and further investigations to exclude conditions associated with progressive conduction system disease. Nevertheless, we firmly believe that PI was not indicated during the manifestation period of BHS in this child.

CONCLUSION

1. Asystole occurring during "pale" type breath-holding spells is not indicative of sinus node dysfunction. It is transient, prognostically benign, and does not warrant pacemaker implantation, regardless of the duration of asystole or the presence of syncope.
2. The clinical and electrocardiographic pattern of "pale" type BHS includes progressive sinus tachycardia triggered by the child's negative emotions or painful stimuli, abruptly interrupted by bradycardia transitioning into asystole, leading to syncope and sudden pallor of the skin.

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