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Permanent cardiac pacing in a 2.5 month-old infant with severe cyanotic breath-holding spells and prolonged asystole

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Abstract

We report the case of a 2.5 month-old infant with cyanotic breath-holding spells, loss of consciousness and seizures. Prolonged asystole up to 70 s despite cardiopulmonary resuscitation was documented by 24 hour Holter monitoring. An epicardial pacemaker was implanted followed by no further loss of consciousness and seizures during spells in a six month follow-up period. (Cardiol J 2011; 18, 6: 704–706)

Key words: breath-holding spells, pacemaker, child

Introduction

Breath-holding spells (BHS) are common non-epileptic paroxysmal disorders in infancy. It is rarely seen in newborns but typically occurs between the ages of six months and four years. Clinically it can be pallid, cyanotic or mixed type [1–3]. The attack is usually triggered by physical or emotional stimuli and has a benign prognosis in the long term. However, severe pallid spells, and less commonly cyanotic spells, may be complicated by loss of consciousness, posturing, seizures and asystole. It is an infantile form of neurally mediated syncope with exaggerated vagally cardioinhibitory response, prolonged cerebral hypoxia and other complex mechanisms [1–4]. Iron therapy for anemia [5], anticholinergic treatment with atropine [6], clonidine and levetiracetam [7] are multiple medications which are tried with variable success in complicated and common forms, but a permanent cardiac pacemaker implantation may be required for spells with prolonged bradycardia and asystole [4, 8, 9]. In this study, we report a patient with severe cyanotic BHS and asystole who was successfully treated with pacemaker implantation.

Case report

A 2.5 month-old boy was referred to our hospital for evaluation of frequent cyanotic BHS once or twice daily over the previous 20 days. Some spells were complicated by loss of consciousness and seizures. His initial cardiologic and neurologic examinations were found to be normal. He started to cry in the examination room and became cyanotic after 15 s. Loss of consciousness was followed by myoclonic seizures. He was resuscitated due to respiratory and cardiac arrest with positive pressure ventilation and cardiac massage. Consciousness returned rapidly after his cardiac function normalized. He was otherwise healthy. His seven year-old brother had a similar clinical history and underwent a permanent pacemaker implantation at 2.5 years of age. But he has already been followed up by the neurology department because of epileptic attacks and mental retardation.

The infant was admitted for further investigation. His electrocardiography (ECG), electroencephalography (EEG), echocardiography, cranial ultrasonography, cranial magnetic resonance imaging and metabolic tests were all found to be nor-

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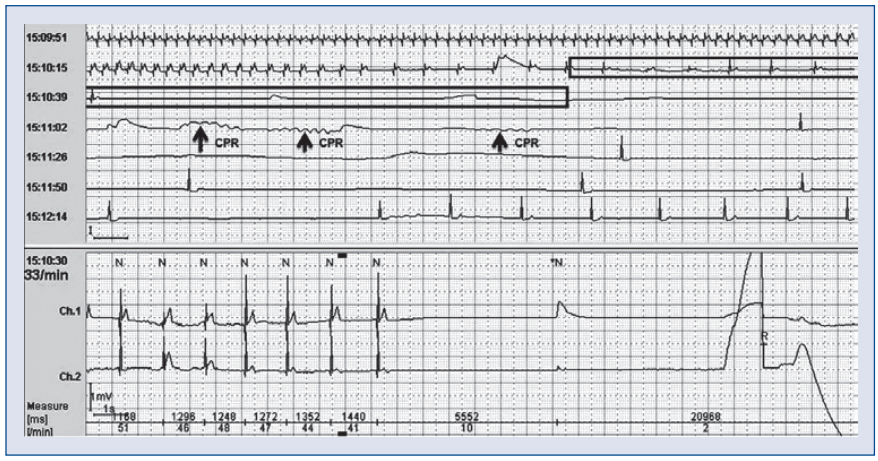

Figure 1. Holter monitoring records showing prolonged asystole of about 70 s despite cardiopulmonary resuscitation (CPR) during breath-holding spells.

mal. Ambulatory ECG monitoring demonstrated a sudden drop in his heart rate with the spell and bradycardia which was followed by asystole for 70 s. A junctional rhythm appeared following resuscitation and sinus rhythm with ST elevation returned after 60 s (Fig. 1). A permanent pacemaker was implanted epicardially immediately after the attack. He was observed to have two short episodes of cyanosis without loss of consciousness during the follow-up.

Discussion

Breath-holding spells are relatively common and generally benign paroxysmal attacks in infancy, and often disappear with aging. The spells are cyanotic in most patients (54–62%). Other patients have either pallid (19–22%) or mixed attacks (19––24%). Some patients have a family history [1–3]. Although the pathophysiology of the pallid and cyanotic types is different, loss of consciousness, seizures, sphincter incontinence, bradycardia and asystole are similar clinical features of both types [1–4]. Pallid type BHS is an infantile form of cardioinhibitory neurocardiogenic syncope. On the other hand, the pathogenesis is more complex in the cyanotic BHS which occurs at full expiration. Hyperventilation, expiratory apnea, valsalva maneuver, and intrinsic pulmonary interactions are the proposed mechanisms in these patients [1, 9].

Breath-holding spells are rather dramatic events for parents, and reassuring families is a mainstay of therapy. However, BHS with asystole has been documented to result in sudden death. Southall et al. [10] reported sudden death in eight of 51 infants and young children with recurrent episodes of cyanosis and loss of consciousness precipitated by crying. Autopsies suggested other causes of death in six of the children, but sudden death could have been precipitated by primary BHS in the other two children. There has been a relationship between frequent BHS and the development of epilepsy and neuromotor retardation as a consequence of repetitive ischemic cerebral trauma. The incidence of epilepsy has been suggested at between 0.5% and 11% of these patients [1–4]. We believe that frequent BHS treated with relatively late implantation of a permanent pacemaker was responsible for the developmental delay and epileptic attacks of our patient's brother.

Once the diagnosis of BHS is established by excluding all other reasons for loss of consciousness and seizures, detailed explanation and reassurance of families are the mainstays of therapy. The minority of patients require specific therapy. Although BHS attacks are generally benign, in some infants they are recurrent, prolonged and accompanied by seizures as a result of cerebral hypoperfusion. Children experiencing prolonged asystole during BHS have been treated with permanent pacemaker implantation. In the study by Kelly et al. [8], ten patients (aged between 12 months and five years) with BHS associated with bradycardia were treated with permanent ventricular pacemaker implantation at the Mayo Clinic between 1985 and 1995. The mean asystolic pause was 11.9 s and pacemaker implantations were found to be safe and efficacious. In a similar study, Legge et al. [4] reported successful treatment of cyanotic BHS complicated by prolonged asystole and seizures by ventricular pace-

maker implantation. The patients remained free of syncope although they continued to demonstrate breath-holding behavior.

Breath-holding spells with unconsciousness and seizures need a cardiologic evaluation and pacemaker implantation. As in our patient's brother, BHS may precipitate epileptic seizures and neurodevelopmental delay.

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