Congenital sick sinus syndrome with breath holding and severe syncope episodes during infancy A case report

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SUMMARY: Karagöz T, Çeliker A, Özer S, Özme Ş, Saraçlar M. Congenital sick sinus syndrome with breath holding and severe syncope episodes during infancy: a case report Turk J Pediatr 2000; 42: 338-340.

Sick sinus syndrome is a rare cause of bradycardia in children without structural heart disease. A case of profound sinus bradycardia, sinus arrest with junctional escape, and pauses in a two-year-old infant with breath-holding and syncope episodes is presented. As a result of these clinical symptoms and elecrocardiographic findings, the patient with sick sinus syndrome underwent implantation of transvenous ventricular pacemaker. He has been well and asymptomatic since the insertion of the pacemaker. In the differential diagnosis of an infant with breath-holding and syncope episodes, when these symptoms in particular cannot be explained by other common reasons, sick sinus syndrome should be kept in mind. This case also illustrates the importance of electrocardiographic studies for the diagnosis.

Key words: sick sinus syndrome, breath holding, syncope, infancy.

Although sick sinus syndrome (SSS) is usually a disease of the elderly produced by idiopathic degeneration of the sinoatrial node, it can be seen in children after extensive cardiac surgery, particularly involving the atria, such as the Mustard or Senning procedure, and after operations for tetralogy of Fallot or tricuspid atresia[1]. SSS occurs infrequently in children who have not undergone cardiac surgery, otherwise involving a normal heart without structural defect[1]. The reason sometimes can be a focal myocarditis or arteritis[1].

Its initial manifestations range from asymptomatic to nonspecific symptoms including palpitation, fatigue, dizziness, chest pain, syncope, congestive heart failure and even sudden death[1]. Although SSS rarely occurs, considering its serious consequences, for example severe syncopes and sudden death, it should be borne in mind in the differential diagnosis of an infant with breath holding and severe syncope episodes.

Case Reports

A two-year-old male infant was referred for evaluation of breath holding and syncope. He was born at term, the first pregnancy of a 23-year-old healthy mother. His birth weight was 3000 g. The parents were first cousins. A slow fetal heart rate was detected by a Doppler recording at the prenatal routine examination at 32 weeks of gestation. Then fetal echocardiography was performed which revealed bradycardia with normal anatomy. Because of the poor cultural and economic level, the woman gave birth in another center without informing us and did not bring her baby for a check-up after the delivery for two years. His medical history included four breath-holding episodes lasting about 20 seconds and three syncope episodes lasting about 3-10 minutes, noticed by the parents over the two years.

Physical examination revealed only bradycardia (45 beats per minute). His weight was 12,000 g (25^{th}-50^{th} percentile); and head circumference, 49 cm (50^{th} percentile). Otherwise the physical examination was normal.

Electrocardiography (ECG), ambulatory ECG analysis and echocardiographic study were performed. A standard 12-lead electrocardiogram revealed profound sinus bradycardia less than

50 beats per minute (Fig. 1). Echocardiographic study showed normal cardiac anatomy. On 24-hour electrocardiography, his rate ranged from 33 to 77 (average 45) beats per minute (bpm). Also, it represented frequently occurring sinus arrest with junctional escape and pauses above two seconds (Fig. 2).

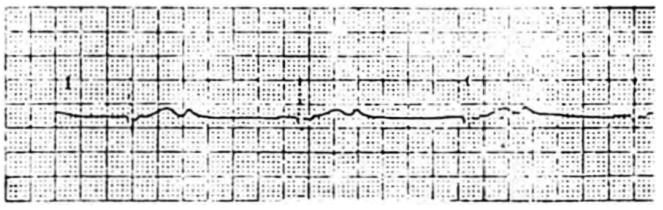
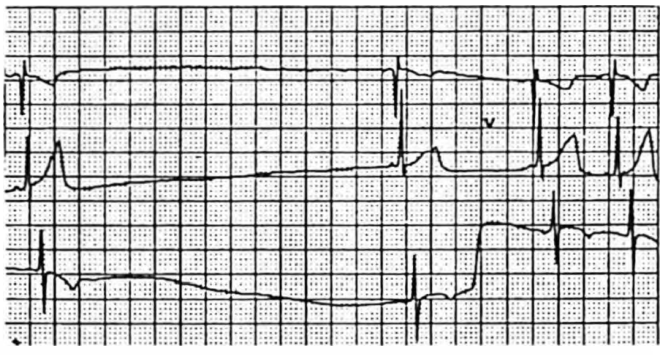


Fig. 1. Standard electrocardiogram of patient with profound sinus bradycardia.



Fg. 2. Ambulatory electrocardiography of patient with sinus pauses above two seconds.

Because of severe syncope episodes and profound bradycardia, we decided to implant a permanent pacemaker, and immediately inserted a "Pacesetter Microny SR+ 2425T S/N: 732501994 VVIR" pacemaker (ventricular). Since the insertion of the pacemaker, he has been well and asymptomatic. He is now three-years-old and his growth and development are normal.

We investigated the other members of the family for SSS and found no evidence of sinus node disorders.

Discussion

The underlying pathologic abnormality in patients who have undergone extensive cardiac surgery is probably sinus node damage because of hemorrhage, necrosis, and injury during operation with placemant of sutures through the sinus node[1]. However, the underlying pathologic process and causes in young patients with normal cardiac anatomy are unknown. There is not enough information about pathologic features of sinus node dysfunction in the medical literature. Thus, to describe SSS when no other cause is found, the terms "idiopathic, primary, and congenital" are commonly used. Familial occurrences have been described in a few reports but are probably rare, and these reports revealed an autosomal dominant mode of inheritance[1,4]. For this reason we investigated the parents of our case, but they showed no evidence of sinus node disorders.

The diagnosis of SSS in children often depends on the clinical setting. The clinical manifestation of SSS is related directly to age, the function of the remaining conduction system and the underlying hemodynamic state. In infants, poor feedings, lethargy or signs and symptoms of congestive heart failure may be associated with severe bradycardia. In older children bradycardia may manifest as general fatigue, and increased sleep requirement. Dizziness and syncope are difficult to detect in infants and young children. In this age group, seizures may be secondary to unwitnessed syncope. Furthermore, unless these young patients have noticeable syncope episodes and seizures, the disease is unlikely to be detected. Some reports are indicating that SSS may frequently be undiagnosed in young patients who have this syndrome and a normal cardiac anatomy[1]. The incidence of such dysfunction in children without associated cardiac defects may be higher than the reported values in the literature. We thus believe that any complaints of "breath-holding spells", syncopes, dizziness or light-headedness, unexplained seizures, or any observation of palpitation with a fast or slow heart rate should be investigated further by ambulatory or exercise electrocardiography, or both. In some cases, an electrophysiologic study may be used for the diagnosis.

The diagnosis of sick sinus syndrome is based on the following electrocardiographic findings: sinus arrest (pauses) for more than two seconds, sinus bradycardia (less than 60 beats/min during sleeping), marked sinus arrhythmia, slow escape rhythms, sinoatrial exit blocks, bradyarrhythmias, tachyarrhythmias, sinus node re-entry tachycardia, or atrial muscle re-entry tachycardia (atrial flutter or fibrillation)[1,5]. Patients may have any one or a combination of these arrhythmias[1]. But atrial muscle re-entry tachycardia may also exist without accompanying sinus node

dysfunction[1]. We diagnosed the sinus node dysfunction in our patient who met at least three surface ECG criteria for sinus node dysfunction.

When severe sinus or junctional bradycardia and frequent sinus arrest result in loss of consciousness, acute medical treatment is indicated. Regardless of causes, intravenous atropine 0.04 mg/kg and/or isoproterenol 0.05-0.5 ug/kg/min usually increases the heart rate[1]. But chronic medical treatment is not accepted as standard treatment for SSS. Treatment in asymptomatic patients with SSS is controversial. When there are life-threatening symptoms such as near-syncope or syncope episodes associated with arrhythmias, like in our patient, the accepted mode of treatment is permanent pacemaker implantation[1,5,6]. The prognosis in patients with congenital SSS is good. To our knowledge, sudden death from sinus node dysfunction after pacemaker implantation has not been reported[1,5].

In order to obtain a diagnosis in infants with breath-holding and syncope episodes, the following are important: detailed investigation of medical history, performance of complete physical examination and electrocardiographic studies. Finally, due to life-threatening symptoms and sudden death, early detection and treatment are valuable for symptomatic children.

Thus, in the differential diagnosis of an infant with breath-holding and syncope episodes, particularly when these symptoms cannot be explained by other common reasons, sick sinus syndrome should be kept in mind.

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