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

Tevfik Karagoz, Alpay Celiker, Sema Ozer, Sencan Ozme, Muhsin Saraclar
Cardiology Unit, Department of Pediatrics, Hacettepe University Faculty of Medicine, Ankara, Turkey


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 electrocardiographic 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 atresia1. SSS occurs infrequently in children who have not undergone cardiac surgery, otherwise involving a normal heart without structural defect1. The reason sometimes can be a focal myocarditis or arteritis1.

Its initial manifestations range from asymptomatic to nonspecific symptoms including palpitation, fatigue, dizziness, chest pain, syncope, congestive heart failure and even sudden death1. Although SSS rarely occurs, considering its serious consequences, for example severe synapses 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 (25th-50th percentile); and head circumference, 49 cm (50th 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
with fibrosis and bile stasis. Iron deposition in the other parenchymatous organs of the body is a constant feature. Three cases of IMF associated with iron deposition in the liver are reported in the literature (one is dense, the other is moderate, and the third is slightly higher than normal range). These findings are considered coincidental and not related to IMF. Excess iron intake by the mother is a suspected etiologic agent in the development of neonatal hemochromatosis. However, various experimental studies showed that the placenta controls iron transportation and that excess iron in the mother does not affect the fetus in utero. Therefore, maternal iron overload alone is not responsible for the abnormalities seen in NHC. In our case, due to secondary changes like hemorrhage and necrosis of the tumors, excessive free iron and its compounds from the degenerated cells caused hemochromatosis.

REFERENCES