

Isolated Sinus Node Dysfunction in an Infant with Developmental Delay

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Abstract We present a case of an infant with developmental delay, a structurally normal heart, and prolonged sinus pauses that suffered an unmonitored cardiac arrest while in the hospital. An investigation into the etiology of her sinus pauses did not reveal a definitive cause, and she continued to have pauses up to 15 s in duration. Her history of the cardiac arrest and the duration of the pauses guided the physicians' recommendations and the family's decision to undergo pacemaker implantation prior to discharge from the hospital.

Keywords Sinus pause · Cardiac arrest ·
Pacemaker implantation · Developmental delay

Sinus pauses are a common finding in healthy adults, occurring in 1–11% of control subjects [3, 8], usually during sleep and times of high vagal tone. They occur less frequently in children [2, 9]. Typically, these pauses do not cause symptoms, although patients might experience dizziness, syncope, or sudden death when the pauses are prolonged. Symptomatic sinus node dysfunction is referred to as sick sinus syndrome. The majority of patients with sick sinus syndrome have underlying structural heart disease or systemic diseases. The decision to implant a pacemaker in patients with sinus bradycardia or prolonged sinus pauses is largely dependent on symptomatology. We report a case of an infant with a structurally normal heart and prolonged sinus pauses who suffered an unmonitored

cardiac arrest while in the hospital and subsequently underwent pacemaker implantation prior to discharge from the hospital.

Case Report

SJ is a 10-month-old girl who was the product of a full-term, uncomplicated pregnancy in Puerto Rico. She remained hospitalized for 8 days after birth for hypotonia and poor feeding. Her parents are first cousins and there is a strong family history of developmental delay but no family history of cardiac anomalies or arrhythmias. At 2 months of age, she was hospitalized in Puerto Rico for viral meningitis. At that time, her mother reports that she had “funny heart beats” that were evaluated and the family was told there was nothing further to be done for the arrhythmia. She has always had poor oral intake, slow weight gain, nystagmus, and severe developmental delay. The patient's family moved to the United States approximately 5 months prior to admission.

In March of 2005 she was admitted to our institution with acute gastroenteritis and dehydration. On hospital day 2, her mother reported that she suddenly became blue and began shaking. Her nurse initiated CPR after noting the patient to be apneic and pulseless. The patient was not being monitored at the time of arrest, and by the time monitors were connected, the patient had regained a pulse and was in sinus rhythm on the monitor. She did, however, have intermittent pauses on the monitor that lasted several seconds, but her perfusion was not compromised during these pauses. During the pauses, she was given Epinephrine and Atropine, both of which transiently increased her heart rate from the 50s to greater than 100, but several minutes later, she would have recurrence of the sinus pauses. The

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patient was intubated and transferred to the pediatric intensive care unit (ICU).

In the ICU, the patient was empirically started on antibiotics after cultures were drawn, but she required no other pharmacological support. She was weaned from the ventilator and extubated on hospital day 4. She continued to have sinus pauses, with a maximal duration of 15 s (Fig. 1). Of note, these pauses would resolve spontaneously, and the patient never had any documented hemodynamic changes during the pauses, although an arterial line was never placed for invasive monitoring of her blood pressure. Although some episodes were followed by junctional escape beats, others resolved with a sinus beat. The majority of these pauses occurred when the patient was asleep, but she did have episodes while awake as well.

The patient underwent an extensive workup. A 15-lead electrocardiogram showed normal sinus rhythm with a nonspecific T-wave abnormality, a QRS duration of 60 ms, a QT interval of 284 ms, and a QTc interval of 424 ms. A 24-h Holter monitor showed normal sinus rhythm with occasional sinus bradycardia and sinus pauses up to 3 s in duration with junctional escape. On the Holter monitor, the heart rate ranged from 40 to 190 beats per minute, with an average heart rate of 130 beats per minute. A transthoracic echocardiogram revealed normal intracardiac anatomy and good biventricular function, with a shortening fraction of 38%. A head magnetic resonance image (MRI) showed diffuse volume loss above the tentorium and a watershed pattern of injury consistent with a history of hypotension. An EEG showed slow background activity, but no lateralized, epileptiform, or electrographic seizures. A pH probe revealed only minimal gastroesophageal reflux, for which she was treated with ranitidine. A barium swallow/upper gastrointestinal tract radiography revealed normal intestinal anatomy. Additionally, she has received an extensive metabolic and genetic workup both in the past and during the current hospitalization that included a chromosomal analysis, DNA methylation studies, mitochondrial DNA, serum amino acids, urine organic acids, carnitine profile, long-chain fatty acid analysis, ammonia level, and a muscle biopsy, all of which were nondiagnostic.

After extensive discussions with the family, the decision was made to place an epicardial single-chambered

pacemaker, which she underwent on hospital day 21. The patient tolerated the procedure well and was discharged from the hospital 14 days later. She had significant oral aversion and feeding issues both preoperatively and postoperatively, which necessitated the prolonged postoperative course; no active cardiac issues were present in the postoperative period. She was discharged home with the pacemaker programmed VVI with a rate of 90 beats per minute and hysteresis set at 60 beats per minute. At her follow-up clinic visit, the patient's mother reports that her daughter remains asymptomatic from a cardiovascular standpoint, and her pacemaker interrogation revealed that the device was working properly with low thresholds and pacing approximately 30% of the time.

Discussion

When sinus node dysfunction occurs to the extent that the atrial rate is insufficient to provide adequate cardiac output for physiologic requirements, clinical symptomatology occurs and the patient is diagnosed with sick sinus syndrome. In adults, sick sinus syndrome can arise from a variety of both intrinsic and extrinsic causes, and it is seen with increasing frequency above age 50 [1, 3]. In pediatric patients, however, sinus node dysfunction, whether symptomatic or not, is very uncommon and is usually associated with increased vagal tone or structural heart disease [2, 9, 10]. Several genetic and metabolic diseases have known associations with sinus node dysfunction, such as Rett and Emery-Dreifuss syndromes [6, 11].

As detailed earlier, an extensive workup was performed to search for the cause of this patient's sinus pauses, but no definitive diagnosis has been made. Although the cause of her sinus pauses remains unclear, the unmonitored cardiac arrest on the floor caused us to classify her as symptomatic, and she thus met criteria for sick sinus syndrome.

A review of the literature revealed that the longest reported "asymptomatic" sinus pause in an adult patient was 35 s [7], and pauses up to 24 s have been reported in children with breath-holding spells [5]. There is no accepted upper limit for the duration of "safe" sinus pauses in asymptomatic children, and the ACC/AHA/NASPE

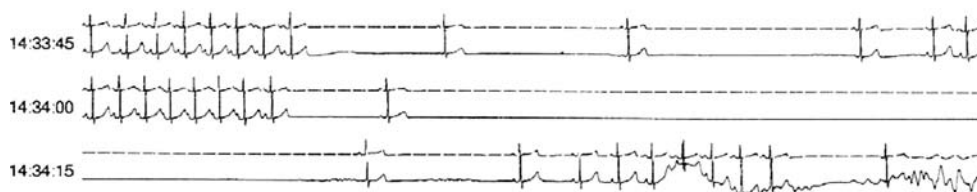


Fig. 1 Telemetry tracing showing a sinus pause of ~15 s followed by a sinus beat

guidelines for pacemaker insertion for patients with sinus pauses or bradycardia base the decision solely on symptomatology [4].

This patient's severe developmental delay impaired our ability to assess whether her sinus pauses limited her functional status. Additionally, because the cause of her developmental delay remains unclear but is presumed to be due to a metabolic or genetic disorder, it is unknown if placement of a pacemaker will alter her life expectancy. The decision to place an epicardial pacemaker in this infant was made after a joint meeting with cardiology, neurology, genetics, and the patient's family based largely on the length of the sinus pauses and the prior cardiac arrest. Future patients similar to SJ will continue to require individualized therapeutic decisions.

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