An unexpected finding in a symptomatic athlete with congenital heart disease and an epicardial pacemaker

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Introduction

This case report highlights the complexities of evaluating a competitive high school athlete with repaired congenital heart disease, an epicardial dual-chamber pacemaker, and exertional symptoms. Intrinsic conduction properties can change over time in patients with congenital heart disease. Reassessment and surveillance is recommended.

Case report

An 18-year-old presented to an emergency department (ED) following an episode of near-syncope while running the 400-meter race during a track meet. Symptoms were described as abrupt loss of power coming out of the starting blocks followed by fatigue, disorientation, and near syncope. Bystanders reported pallor and change in mental status. Patient was transported to the ED for evaluation. Patient history included D-transposition of the great arteries and a ventricular septal defect (VSD), status post atrial septostomy repair and arterial switch with pericardial patch closure of large inlet VSD, and ligation and division of a patent ductus arteriosus. An epicardial dual-chamber pacemaker was implanted for sinus node dysfunction 6 years post surgical repair.

Prior to this event, the patient had a several-year history of exertional symptoms of chest pain, shortness of breath, and brief disorientation occurring 1–2 times a year. Over the years, these symptoms were evaluated with echocardiograms, pacemaker interrogations, Holter monitors, 30-day looping event monitors, cardiopulmonary exercise stress tests (CPET), stress echocardiograms, and computed tomography angiograms (CTA). Cardiac catheterization with angiography was not completed owing to prior anaphylaxis with contrast during CTA. CTA showed normal coronary ostia and lack of obvious coronary compression by epicardial leads. The patient had a remote history of palpitations with associated high-rate episodes on pacemaker interrogations, concerning for atrial tachycardia, that was treated with digoxin. These episodes did not correlate with the patient's exertional symptoms. An etiology for the patient's symptoms was not determined from this testing. Of note, on prior CPETs the patient achieved peak sinus rates of 190 beats per minute (bpm) with intact conduction to the ventricle.

The evaluation in the ED included an electrocardiogram, chest radiography, and blood work, which included a normal troponin. The electrocardiogram showed normal sinus rhythm with intact conduction without ST- or T-wave changes. The chest radiograph was without evidence of atrial or ventricular lead fractures. A Medtronic CareLink Express (Medtronic, Minneapolis, MN) transmission showed stable lead characteristics and no monitored arrhythmia events. The noncardiac evaluation was unremarkable. During ED observation, vital signs and rhythm were within normal limits. The patient was discharged home from the ED and was restricted from exercise until further evaluation was completed.

KEY TEACHING POINTS

- Ambulatory cardiac monitoring may not be adequate for symptom rhythm correlation in competitive athletes.
- Pacemaker programming can be challenging in competitive athletes and requires understanding of a patient's intrinsic conduction both at rest and with exercise, which may change over time.
- A comprehensive pacemaker assessment with pacemaker monitoring during cardiopulmonary exercise stress testing may provide further insight on potential causes of symptoms in addition to evaluation for stored arrhythmia events on the device.

KEYWORDS Epicardial pacemaker; Congenital heart disease; Transposition of the great arteries; Exertional symptoms; Athletes; Cardiopulmonary exercise test; Sinus node dysfunction; Atrioventricular block
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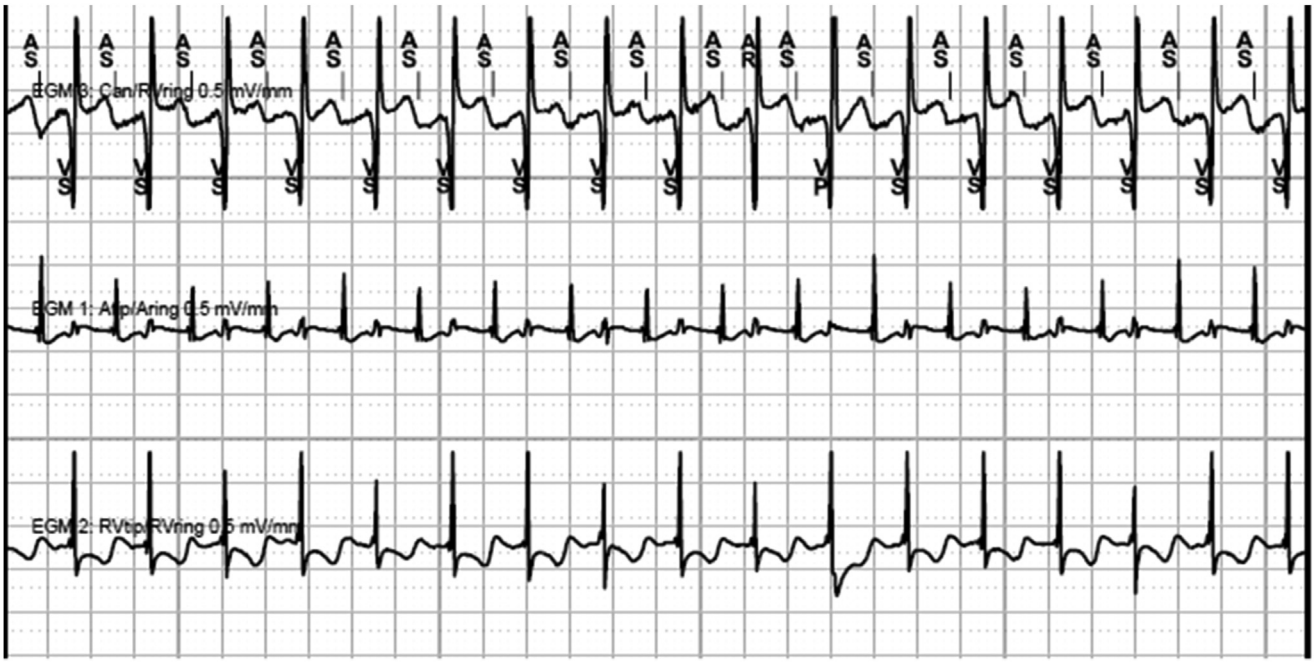

Figure 1 Conduction at peak exercise on cardiopulmonary exercise stress test as recorded by the Medtronic programmer, showing 1:1 conduction and rare oversensing on the atrial lead.

In the electrophysiology clinic, an echocardiogram showed flow in right and left coronary origins, no outflow tract obstruction, and normal biventricular size and function. Of note, the patient had a previous coronary CTA notable for normal coronary ostia and normal appearance of distal coronary arteries. Interrogation of patient's Medtronic Azure pacemaker showed stable capture thresholds, no monitored arrhythmia events, atrial pacing of 15.3%, and ventricular pacing <0.1%. There was a Managed Ventricular Pacing (MVP) mode switch event on the date of and at approximately the same time as the patient's symptoms. The median ventricular rate was 182 bpm at the time of the MVP mode switch. A limited noninvasive programmed stimulation (NIPS) demonstrated a 2:1 block at heart rates greater than 162 bpm (370 ms). Digoxin was discontinued and a Holter was unremarkable except for rare atrial and ventricular ectopy, occasional atrial pacing, and rare isolated ventricular paced beats.

A CPET was performed. The patient completed 14.63 minutes of the Bruce Protocol with a respiratory exchange ratio of 1.15 and a peak oxygen consumption of 51.1 mL/kg/min. The peak heart rate was 179 bpm with 1:1 conduction (Figure 1). There were no ST-T wave abnormalities. Isolated premature atrial contractions were noted. Importantly, the patient did not develop clinical symptoms. A modified protocol with sustained running at 11.1 mph at an incline of 1% was performed in an attempt to elicit the symptoms. Neither atrioventricular (AV) block nor symptoms were elicited. A second NIPS was performed immediately following CPET that showed 2:1 AV block at 200 bpm (300 ms) without associated symptoms (Figure 2). The patient's pacemaker was reprogrammed with an increased upper tracking rate (200 bpm) and the sensed AV delay was decreased to 100 ms (Table 1). Mode switch was turned on owing to the remote history of atrial tachycardia and recent discontinuation of digoxin. In the 9 months since pacemaker reprogramming, the patient resumed competitive running at the collegiate level and is asymptomatic.

Discussion

This case report highlights the complexity of symptom evaluation in competitive athletes with congenital heart disease and implantable cardiac rhythm devices. For patients who had an arterial switch operation for D-transposition of the great arteries, careful evaluation of coronary arteries, ventricular function, and outflow tracts is warranted when exertional symptoms arise.[1] Coronary artery compression by pacing leads should be included in the differential for patients with epicardial leads. When hemodynamic and structural evaluations are unremarkable, the evaluation should focus on arrhythmia and, if an implantable cardiac rhythm device is present, on device programming. AV block is the most common arrhythmia following arterial switch, with VSD closure as an additional risk factor.[2]

When arrhythmias are suspected, obtaining symptom rhythm correlation is paramount. Ambulatory monitors may be difficult for athletes to wear during training or competition. CPET can be a useful evaluation tool for patients with exertional symptoms; however, traditional protocols may not capture symptoms. Modification of the traditional protocols may be required to elicit symptoms.[3,4] For this competitive sprinter, traditional treadmill protocols did not replicate the explosive acceleration or the maximum speed the patient achieves in a race. Despite our best efforts, we were unable

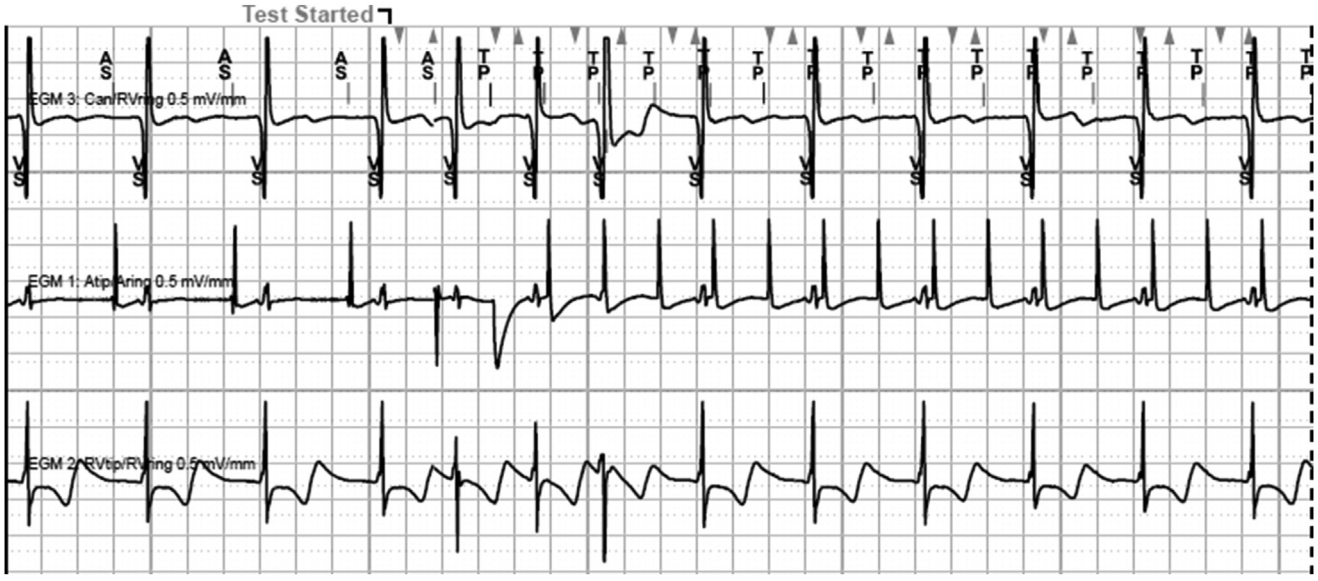

Figure 2 Atrial bust pacing at a rate of 200 beats/min (300 ms) revealing 2:1 block.

to replicate the patient's symptoms during the stress test even with modifications of the testing protocol.

Our hypothesis is that this patient experienced a period of 2:1 block immediately out of the starting blocks, prior to the MVP mode switch, and/or they were subsequently limited by the programmed upper pacing rate during sprinting. We were unable to prove this with the testing performed. The patient's MVP mode switch event supports the assumption of inadequate AV nodal conduction during exercise. The patient developed 2:1 block on a limited NIPS performed at rest but was able to conduct 1:1 with exercise at rates faster than the cycle length at which 2:1 conduction occurred. Of note, NIPS immediately following exercise did show 2:1 conduction at a heart rate of 200 bpm, supporting the possible development of AV conduction abnormalities at a heart rate not obtained on a stress test, but possibly a rate the patient might have during competition. The shorter cycle length at which 2:1 conduction occurred may be due to catecholamine status immediately following CPET and/or the discontinuation of digoxin a couple of weeks prior to the CPET. Pacemaker programming was adjusted to allow for a higher maximum tracking rate. Although we could not replicate the patient's symptoms, in the last 9 months following pacemaker reprogramming, symptoms have not recurred during competitive sprinting.

Conclusion

This case report emphasizes the complexity of symptom evaluation in a competitive high school athlete with repaired congenital heart disease and a pacemaker that was initially implanted for symptomatic sinus node dysfunction. A CPET 3 years prior to this evaluation suggested normal AV nodal function; however, further evaluation showed new AV nodal dysfunction. Available ambulatory cardiac monitors may not be adequate for symptom rhythm correlation in competitive athletes. Finally, pacemaker programming for athletes should consider both resting and active conduction properties of the individual patient.

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Table 1 Comparison of pacemaker settings programmed prior to the cardiopulmonary exercise stress test and changes made based on stress test and noninvasive programmed stimulation findings

	Initial programming	Final programming
Mode	MVP (AAI <=> DDD)	MVP (AAI <=> DDD)
Upper tracking rate	175 bpm	200 bpm
Mode switch	Off	>214 bpm
Paced AV/Sensed AV delay	160 ms/160 ms	120 ms/100 ms
Rate adaptive AV delay	On	Off
PVARP	250 ms	300 ms
Atrial sensitivity	0.45 mV	0.6 mV

AV = atrioventricular; bpm = beats per minute; MVP = managed ventricular pacing; PVARP = postventricular atrial refractory period.

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