

Congenital Atrial Standstill: A Rare Case of Atrial Inexcitability and Mechanical Standstill

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Introduction

Atrial standstill (AS) is characterized by lack of electrical and mechanical activity that is often seen in older adults with scarred atria post ablation or post cardiac surgery.

We report a unique case of a young patient who presented with symptomatic junctional bradycardia and was found to have atrial electrical inactivity (absence of p waves) and mechanical standstill.

A 43-year-old female presented with dyspnea and lightheadedness with a junctional rhythm at a rate of 32 bpm. Transthoracic Echo demonstrated EF of 65%.with severe biatrial enlargement with no A wave on tissue doppler. With exercise, junctional tachycardia was noted. Holter monitor demonstrated junctional rhythm with heart rates ranging from 26 bpm to 65 bpm. There was no evidence of sinus activity on EKG. Cardiac MR demonstrated global cardiomegaly and small subendocardial delayed enhancement in anterior wall. Left and right heart catheterization revealed normal coronary arteries, pulmonary artery systolic pressure of 59 mmHg, and wedge of 17 mmHg. Symptoms persisted despite diuresis and decision was made to implant a pacemaker. During pacemaker implantation, It was noted that the right atrium was inexcitable with lack of atrial capture and sensing. A catheter in coronary sinus also revealed only ventricular far field signals with absence of atrial electrical activity in left atrium as well. A single lead was implanted In the right ventricle. No retrograde atrial activity was noted with ventricular pacing. She was placed on apixaban for anticoagulation. Genetic testing was negative and only revealed a RYR2 gene variant c.3026G>A of unknown significance.

This case demonstrates lack of atrial sensing and capture due to congenital atrial electrical absence.

Key Words

Atrial Fibrillation, Atrial standstill, Atrial Inexcitability

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Congenital atrial standstill is rare and usually associated with SCN5A mutation and Connexin 40 polymorphisms.

Management includes pacemaker implantation and anticoagulation.

Conflict of Interest

Nothing to declare.



Figure A: Junctional tachycardia on prior treadmill stress test.

Figure C: Echo with absence of A wave on mitral valve inflow Doppler.

Figure E: Pacemaker stimulus from atrial pacemaker lead not capturing atria.

Figure B: EKG with junctional rhythm.

Figure D: Echo showing biatrial dilation.