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Atrial standstill in a pediatric patient with SCN5A mutation following procainamide challenge

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Atrial standstill in a pediatric patient with SCN5A mutation following procainamide challenge

Anmol Goyal, MBBS; Lindsey Malloy-Walton, DO; Christopher Follansbee, MD

Children's Mercy Kansas City

BACKGROUND

- Atrial standstill (AS) is a rare arrhythmia characterized by absence of electrical and mechanical atrial activity which can be associated with SCN5A channelopathy.

CASE

- An 18-year-old male with structurally normal heart, frequent sinus pauses, nonsustained atrial tachycardia and high-grade block was found to have SCN5A mutation c.3823G>A (p.Asp1275Asn).
- An electrophysiology study (EPS) with high density voltage mapping of the right atrium was performed (Fig 1).
- Nonsustained multifocal atrial tachycardia was induced without ablative targets (Fig 2).
- Procainamide challenge was performed which was negative for Brugada, however induced AS (Fig 3-4).
- No atrial capture could be achieved at maximal output. Empiric atrial lead positioning in the right atrial appendage was utilized based on prior atrial mapping (Fig 5).
- There was recovery of atrial activity in <24 hours and atrial threshold improved to 1.25 V at 0.4 ms with impedance of 318 ohms.

DECISION MAKING

- SCN5A disease can have a variable phenotype ranging from being asymptomatic to progressive AS.
- A detailed EP study with high density mapping should be considered to assess for viable atrial tissue prior to pacemaker implantation.
- Progressive disease may result in high thresholds, failure to capture or AS, and patients should be followed closely.

A detailed electrophysiology study with high density atrial mapping should be considered prior to pacemaker implantation in patients with SCN5A channelopathy.

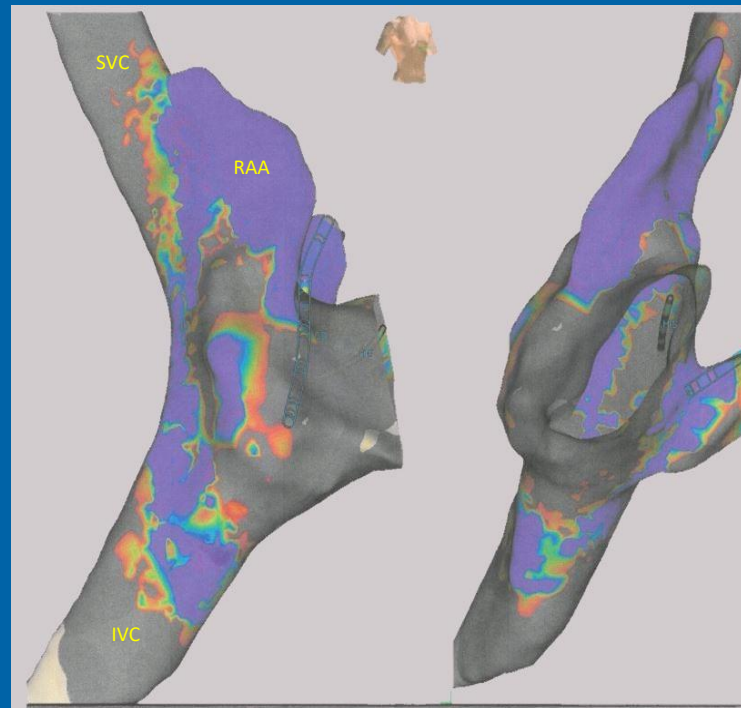


Fig 1. High density voltage mapping of the right atrium. Diffuse low voltage areas (grey) were identified along the anterior, lateral and posterior walls of the right atrium. The area of the sinus node was found to be along the border of high and low voltage regions. High voltage signals (purple) were identified throughout the right atrial appendage (RAA), as well as a thin band extending vertically along the lateral wall.



Fig 2. Non-sustained multi-focal atrial tachycardia with spontaneous termination to normal sinus rhythm.

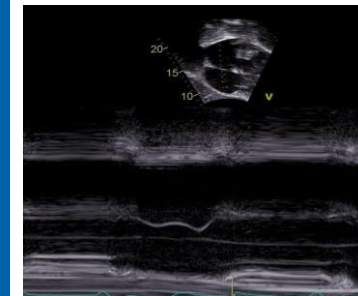


Fig 3. M-mode across the atrium showing no mechanical atrial activity.



Fig 4. 12 lead rhythm strip with absence of P waves with junctional rhythm.

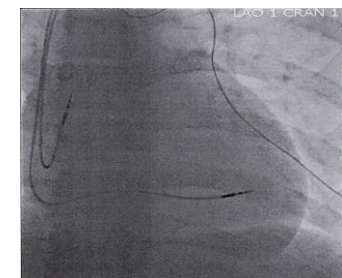


Fig 5. Successful position of the atrial lead in the right atrial appendage.