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

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

**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 3-5).
- 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.

**DISCLOSURE INFORMATION**

- Authors have no disclosures to report

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