

Children's Mercy Kansas City

**SHARE @ Children's Mercy**

---

Research Days

---

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

Anmol Goyal

Follow this and additional works at: <https://scholarlyexchange.childrensmercy.org/researchdays>



Part of the [Cardiology Commons](#), [Cardiovascular Diseases Commons](#), and the [Pediatrics Commons](#)

---

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

**Authors:** Anmol Goyal, Lindsey Malloy-Walton, Christopher Follansbee

**Background:** Atrial standstill (AS) is a rare arrhythmia characterized by absence of electrical and mechanical atrial activity 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 done (Fig 1a). Nonsustained multifocal atrial tachycardia was induced without ablative targets (Fig 1b). Procainamide challenge was negative for Brugada, however induced AS (Fig 1c-d). 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 1e). AS resolved in <24 hours with resultant functioning of the atrial lead.

**Decision-Making:** SCN5A disease can have a variable phenotype ranging from 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.

**Conclusion:** SCN5A channelopathy can result in a unique phenotype that requires careful and serial evaluation by an electrophysiologist. As progressive AS can occur, a detailed EPS with high density atrial mapping should be considered when pacemaker implantation is required.