The Terrible T’s: T-wave Alternans, Timothy Syndrome, and a Two-year-old

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Background
- **T-wave alternans** is an uncommon electrocardiographic (ECG) finding of beat-to-beat alternation in T-wave shape or amplitude.
- T-wave alternans is associated with a prolonged QT interval and long QT syndrome (LQTS).
- T-wave alternans may degenerate to torsades de pointes.¹

Case Description
- A 2-year-old, 15.7-kg male with bilateral hand and foot syndactyly presented for skin graft revision of prior syndactyly repair.
- Medical history included developmental delay.
- The patient had two uneventful anesthetics at 6 and 9 months of age.
- Induction was via mask with sevoflurane 8% in N₂O and O₂, followed by a propofol 2mg/kg bolus prior to a smooth tracheal intubation.
- Midway through the case, ephedrine (10mg total) was dosed intermittently for hypotension.
- The BP improved and vital signs stabilized, yet the ECG demonstrated T-wave alternans (Figure 1).

Outcome
- The patient was admitted to the cardiology service after the procedure.
- He was started on a beta-blocker and had an AICD implanted.
- Genetic testing revealed a heterozygous variant in the CACNA1C gene, consistent with Timothy Syndrome (LQTS type 8).

Discussion
- There are 13 known genotypes of LQTS (overall incidence 1 : 2,500).
- **Timothy syndrome** is associated with arrhythmias during anesthesia, syndactyly, and craniofacial and cognitive abnormalities.
- Anesthetic management includes avoidance of QT-prolonging medications (sevoflurane, ondansetron, ephedrine, phenylephrine, and albuterol) and premedication and pain control to minimize sympathetic stimulation.²³
- Prompt recognition and treatment of T-wave alternans is crucial to prevent progression to a lethal arrhythmia.


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**Figure 1:** T-wave alternans on (A) OR monitor and (B) intraoperative rhythm

**Intraoperative Management**
- Immediate cardiology consult to assist with management.
- Intravenous magnesium (20mg/kg) was given.
- T-waves normalized; patient remained stable and was extubated without incident.
- A basic metabolic panel was sent and was normal.
- A 12-lead ECG (Figure 2) showed a QTc interval of 585 ms.

**Figure 2:** 12-lead EKG rhythm strip (QTc = 585 ms; normal QTc is <450 ms)