



# SADS FOUNDATION

SUDDEN ARRHYTHMIA DEATH SYNDROMES

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## Timothy Syndrome

### What is Timothy Syndrome (TS)?

TS is a rare and serious genetic disorder characterized by a spectrum of health concerns including the following **warning signs:**

- Abnormally prolonged QTc interval (>480ms but usually much longer).
- Syndactyly (webbing between or apparent fusing of fingers and toes)
- Slow heart rate in fetus or newborn
- 2:1 AV Block at birth or in utero
- Congenital heart defects (ASD, VSD, PDA, PFO, TOF), facial anomalies (flat bridge nose, low set ears, thin upper lip or very round fattish face) and/or weakened immune system

### Major Complications of Timothy Syndrome:

**Anesthesia is a known trigger of arrhythmias in TS.** Secondary complications include severe infections (bronchial and sinus) with reported deaths despite aggressive antibiotic therapy. Also death due to intractable hypoglycemia has been anecdotally reported. Close monitoring of glucose levels is prudent.

**An ECG should be performed on all children with syndactyly prior to surgery.** Most cases of TS have been identified this way in the past 2 years.

It is imperative that anesthesiologists, pediatric orthopedic surgeons and pediatricians, among other healthcare providers, are aware of this condition prior to scheduling a surgery on a child with possible TS to avoid serious complications.

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