Preoperative EKG Testing in Non-syndromic Children with Hand Syndactyly

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Objectives

**Purpose**
To examine the efficacy of an institutional policy requiring preoperative EKG screening for Timothy syndrome in all children referred for syndactyly release

**Timothy syndrome – syndactyly association**
- TS is caused by an autosomal dominant, Ca-channel mutation leading to prolonged QT on EKG
- Life expectancy of 2.5 years, due to fatal arrhythmia
- Most cases diagnosed in utero or as neonates
- Only 25 known Timothy syndrome cases (2011)
- 100% of TS cases have hand syndactyly

Methods

**Study subjects**
- Dx: non-syndromic hand syndactyly
- All patients underwent preoperative screening EKG for Timothy syndrome per institutional policy
- Treated by BCH hand surgeon between 2007–2013

**Medical and billing records review**
- Demographics, clinical presentation, cardiac testing results, and operative findings
- Charges for cardiac testing and interpretation

Results: Clinical Presentation

128 syndactyly patients identified
- 72% were boys
- 65% had bilateral involvement
- Median age at time of EKG and surgery was 1 year

Results: EKG Findings

<table>
<thead>
<tr>
<th>Syndrome (n = 128)</th>
<th>Normal EKG result (n)</th>
<th>Abnormal EKG result (n)</th>
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</thead>
<tbody>
<tr>
<td>Normal (92%)</td>
<td>118</td>
<td>10 (8%)</td>
</tr>
<tr>
<td>Prolonged QT interval (1%)</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Timothy syndrome (0%)</td>
<td>0</td>
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</tbody>
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Cleared for surgery (n) 128 (100%)

Downstream Effects

6% of patients had additional workup due to EKG
- Cardiology visit/consult (6)
- Repeat EKG (4)
- 2D EKG (2)
- Echocardiogram (2)
- Holter monitor (2)
- EKG for 1st degree relatives (prolonged QT patient)

Cost Analysis

**Cost of EKG Policy**
- Minimum cost of EKG policy is $183/patient
- Additional testing can lead to costs of $5000/patient

**By not adopting EKG policy**
- 10-year savings of $33k – $900k for institution
- 10-year savings of $2 – $58 million nationally

Conclusions

- Failed to yield a single instance of TS over 7 years
- False-positive EKG results led to additional testing and costs in 6% of patients
- 1 patient identified with long QT interval on EKG; normalized on subsequent workup; proceeded with syndactyly release without cardiac event
- Ineffective timing of EKG: hand surgeons unlikely to uncover TS as typical age of surgery is 1 year, while TS is usually identified in utero or as neonates
- EKG policy resulted in considerable cost and time of parents, staff, and providers

There is insufficient evidence for routine preop EKG screening for Timothy syndrome in children referred for surgery, and we have abandoned this policy