Preoperative Electrocardiograms for Nonsyndromic Children With Hand Syndactyly

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Purpose To examine the efficacy of preoperative electrocardiogram (EKG) screening for Timothy syndrome, a rare and fatal condition characterized by prolonged QT, in children referred for syndactyly release.

Methods We reviewed the records of nonsyndromic syndactyly patients seen by a hand surgeon at our institution between 2007 and 2013. All underwent a preoperative screening EKG for Timothy syndrome. We reviewed the medical records for demographics, presentation, EKG results, and operative findings, and calculated median age at the time of EKG and surgery and frequency distributions for sex, side affected, EKG result, and clinical finding. The mean patient charge for EKG and interpretation was calculated.

Results We identified 128 syndactyly patients, 72% of which were boys. Median age at the time of EKG testing and syndactyly release was 1 year. A total of 92% of patients had normal EKG results; one patient exhibited a prolonged QT. Ten patients (8%) had further cardiac evaluation because of the EKG result and were found to be normal on repeat testing. No patient met QT threshold for Timothy syndrome and all patients were cleared for surgery. The minimum patient charge for EKG testing was $183.

Conclusions To improve patient safety, some have advocated preoperative EKG testing for all children undergoing syndactyly release to rule out Timothy syndrome. Analysis of our experience failed to yield an instance of Timothy syndrome over a 7-year period. Although EKG charges were relatively low, costs resulting from additional testing, cardiology consultation, and provider and parent time should be considered. Our study does not support routine EKG testing for children referred for syndactyly release, and we have abandoned this practice.

Type of study/level of evidence Prognostic IV.

Key words EKG, pediatric, syndactyly, timothy syndrome.

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SYNDACTYLY IS THE MOST COMMON congenital hand anomaly, occurring in roughly 0.03% of live births.[1,2] Syndactyly can be familial or sporadic and in some instances syndromic (eg, Apert syndrome).[3] The extremely rare Timothy syndrome is characterized by hand syndactyly and a prolonged QT interval on an electrocardiogram (EKG), often resulting in a lethal cardiac arrhythmia.[4] The risk of sudden cardiac events in this subset of syndactyly patients has led to recommendations in the literature

for preoperative EKGs for all patients undergoing surgical treatment.[5,6] In 2007 our institution adopted a policy at the recommendation of our anesthesia and cardiology services, based on a review of the literature and similar to that proposed by Gettys and Gaston,[2] requiring a preoperative EKG for all patients undergoing syndactyly release.

Although early clinical descriptions appeared in the 1990s, Splawski et al[4] established the cause of Timothy syndrome in 2004. A de novo missense mutation in the Cav1.2 L-type calcium channel *CACNA1C* gene produces a clinical syndrome characterized by a prolonged QT interval of 480 to 700 ms on EKG,[7] a number of other cardiac defects, syndactyly of the hands and feet, facial deformities, low-set ears, small teeth, global developmental delays, and autism.[5] As a result, the overwhelming majority of these patients have been diagnosed as neonates during routine testing and evaluation.[7] Although early identification and intervention can prolong life with medication, patients with Timothy syndrome typically have a life expectancy of 2.5 years, with lethal arrhythmia as the primary cause of death.[8] As of 2011, only 25 cases of Timothy syndrome have been reported.[9]

A prolonged QT interval can predispose children to a variety of arrhythmias under general anesthesia. These include ventricular tachycardia, ventricular fibrillation, and bradyarrhythmias that may lead to cardiac arrest or sudden death.[10] For these patients, the anesthesia plan must include minimizing adrenergic stimulation that may be experienced during the induction and emergence phases of an anesthetic, because there is the potential for many anesthetic agents to prolong the QT interval and increase the risk of fatal arrhythmia.[11,12]

The purpose of this study was to examine the evidence supporting a policy regarding preoperative EKG screening of nonsyndromic patients undergoing syndactyly release. We report the prevalence of abnormal EKG findings in this patient population. Charge calculations were performed and recommendations for management are made.

MATERIALS AND METHODS

After we obtained approval from our institution's committee on clinical investigation with a waiver of informed consent, we reviewed the medical records of patients with hand syndactyly treated from 2007 through 2013. Patients were identified by the International Classification of Diseases−revision 9 diagnosis codes: 755.11 (syndactyly of fingers, no fusion) and 755.12 (syndactyly of fingers, with fusion) and using the following Current Procedural Terminology codes: 26560, 26561, and 26562 (formal syndactyly release with skin flap, with skin flaps and grafts, and complex syndactyly, respectively). Inclusion criteria included nonsyndromic syndactyly patients who were in good health, were examined by a pediatric hand surgeon, and had preoperative screening EKG results. We excluded syndromic patients because they often carry cardiac diagnoses and frequently undergo EKG testing. Medical records were reviewed for demographics, clinical presentation, EKG results, and operative findings.

We calculated the median age at time of EKG testing and surgery, as well as frequency distributions for sex, side affected, syndactyly release status, EKG result, and clinical finding.

A charge-savings analysis was conducted in conjunction with the Division of Cardiology at our institution. From our cohort, we conducted an analysis of patients requiring a cardiology office visit or further workup as a result of the original EKG interpretation. The mean patient charges for EKG screening and subsequent evaluation were generated. Charges were then extrapolated to yield savings for our institution and nationally over a 10-year period.

RESULTS

A total of 128 nonsyndromic syndactyly patients with available EKG results were seen between 2007 and 2013. Most were boys (92; 72%). The distribution of unilateral and bilateral syndactylies was even (49% vs 51%, respectively). Age at time of EKG and surgery was unevenly distributed, skewed right by older outliers, with a median age of 0.8 years (IQR, 0.7−1.7) and 1.1 years (IQR, 0.8−1.8), respectively.

Most patients (118; 92%) had normal EKG results. Ten patients (8%) had results suggestive of an abnormality without a specific comment of normal, including borderline criteria for left ventricular enlargement (n = 6), right ventricular conduction delays suggestive of right ventricular enlargement (n = 3), and prolonged QT interval (477 ms) below threshold for Timothy syndrome (n = 1).

Ten patients (8%) underwent further cardiac evaluation. Preexisting cardiac conditions or symptoms confirmed by EKG (eg, Kawasaki disease, muscular ventricular septal defect, syncope) and parent concern motivated 5 families to have cardiology visits. The remaining 5 patients had cardiac evaluations because of EKG findings; some received repeated EKGs and echocardiograms, all of which were found to be normal. In addition, 4 patients were managed with discussion by a cardiologist because of EKG findings. Three patients