Follow-Up of Electrocardiographic Findings and Arrhythmias in Patients With Anomalously Arising Left Coronary Artery from the Pulmonary Trunk

Judson A. Moore, MD[a], Antonio G. Cabrera, MD[a,b], Jeffrey J. Kim, MD[a,b], Santiago O. Valdés, MD[a,b], Caridad de la Uz, MD[a,b], and Christina Y. Miyake, MD, MS[a,b,*]

Follow-up data and correlation of arrhythmias, electrocardiogram (ECG) changes, and cardiac function in anomalous left coronary artery from the pulmonary trunk or artery have not been previously studied. This is a retrospective single-center review of 44 anomalous left coronary artery from the pulmonary trunk or artery patients diagnosed between 1992 and 2014, at a median age of 3 months (3 days to 13 years). Clinical history, ECG, Holter, and echocardiogram data were reviewed. ECGs were reviewed for contiguous Q-or T-wave inversions, hypertrophy, bundle branch block, and axis deviation. High-grade ventricular ectopy, supraventricular tachycardia (SVT), and ventricular tachycardia (VT) were recorded. Patients with <6 months of clinical follow-up were excluded from longitudinal analysis. At diagnosis, 43 (98%) were noted to have electrocardiographic changes. During hospitalization, arrhythmias were seen in 13 patients (30%): 2 (5%) with sustained VT or ventricular fibrillation, 6 (17%) with high-grade ventricular ectopy, and 4 (9%) with SVT. Seven patients (16%) required antiarrhythmic treatment. During outpatient follow-up, arrhythmias were seen in 11 patients. New arrhythmias were documented in 6 without a history of in-hospital arrhythmias. Of 34 patients with at least 6 months follow-up (median 6 years, 0.5 to 20 years), 20 had left ventricular (LV) dysfunction before surgery. Normalization of function occurred in 94% (median 1 year, 5 days to 4 years). Electrocardiographic changes persisted in 94% at the time of LV function recovery. In conclusion, electrocardiographic changes and arrhythmias may persist despite recovery of ventricular function. Therefore, prolonged myocardial remodeling may continue even after resolution of LV dysfunction during which time arrhythmias may occur. **(Am J Cardiol 2016;118:1563−1567)**

Anomalous left coronary artery from the pulmonary trunk, also commonly referred as anomalous left coronary artery from the pulmonary artery (ALCAPA), is a rare condition making up approximately 0.5% of all congenital heart disease.[1] Coronary reimplantation, now the standard of care, typically results in excellent outcomes when undertaken in the neonatal period.[2,3] Although ALCAPA is commonly associated with electrocardiographic changes, data from the only 2 major studies evaluating electrocardiographic changes in this population differ in reported frequency of electrocardiographic abnormalities and lack follow-up evaluation of electrocardiographic changes. In addition, there have been no studies evaluating arrhythmia, electrocardiographic changes, and cardiac function in the context of the overall clinical picture over time.[4,5] The objective of this study was to provide information regarding the correlation of electrocardiographic changes, incidence of arrhythmias, and left ventricular (LV) function over time in ALCAPA patients.

Methods

This was a retrospective single-center review of patients at Texas Children's Hospital diagnosed with ALCAPA between 1992 and 2014. Patients were excluded if they did not have an available presurgical electrocardiogram (ECG) or if they had significant congenital heart disease in addition to ALCAPA. All patients with less than 6 months of outpatient cardiology follow-up were omitted from analysis of longitudinal trends but remained a component of the presurgical ECG and arrhythmia data analysis. To evaluate trends over time, we focused on 3 major categories: electrocardiographic changes, LV function, and incidence of supraventricular and ventricular arrhythmias.

Preoperative and postoperative ECGs were reviewed for previously published changes associated with ALCAPA[4,5]: (1) pathologic deep Q waves >3 mm in leads I, aVL and >5 mm in V5, V6, or V7; (2) LV hypertrophy (R-wave amplitude in V6 >98th percentile for age); (3) left axis deviation (QRS axis >98th percentile for age); (4) ST-segment elevation or 4 depression in leads I, aVL, V5, V6, or V7; (5) T-wave inversion in leads I, aVL, V5, V6, or V7; and (6) QT pattern (deep Q wave with T-wave inversion) or QR pattern (prominent Q and R wave representing the QRS complex) in leads I, aVL, and V5, V6, or V7. A pediatric electrophysiologist (CM) reviewed all ECGs. Although in previous studies, ECGs were considered abnormal if they displayed

[a]Department of Pediatrics and [b]Lillie Frank Abercrombie Section of Pediatric Cardiology, Department of Pediatrics, Baylor College of Medicine, Texas Children's Hospital, Houston, Texas. Manuscript received December 20, 2015; revised manuscript received and accepted August 2, 2016.

See page 1567 for disclosure information.

*Corresponding author: Tel: (832) 826-5650; fax: (832) 825-0237.
E-mail address: cymiyake@texaschildrens.org (C.Y. Miyake).

0002-9149/16/$ - see front matter
http://dx.doi.org/10.1016/j.amjcard.2016.08.022

Table 1
Patient demographics and characteristics

Characteristic	N=44
Male	21 (48%)
Age at presentation	3 months (3 days − 13 years)
Presenting symptom:	
Respiratory	22 (50%)
Feeding Difficulties	9 (21%)
Murmur	8 (18%)
Other	5 (11%)
Time between diagnosis and surgery	2 days (1 − 119 days)
Median length of hospitalization	18 days (5 − 143 days)
Median duration of follow up	2 years (24 days − 20 years)

any of the aforementioned criteria in at least one of the pertinent leads, for the purposes of this study, we considered electrocardiographic changes to be abnormal only if demonstrated in 2 or more contiguous leads.[4,5] Thresholds for abnormal, such as maximal Q wave depth and hypertrophy, were determined by previously published large scale data sets for available leads.[6,7] Rate of complete ECG normalization was defined as no longer displaying any previously mentioned electrocardiographic changes.

LV dysfunction was defined as a shortening fraction with a Z-score ≤ 2 or ejection fraction $<55\%$ on echocardiography. Data were reviewed preoperatively, in the immediate postoperative period, and during annual follow-up visits for the first 5 years after surgical correction, as well as data available beyond 5 years. Timing of restoration of function was then compared with normalization of electrocardiographic findings and occurrence of arrhythmias.

Evidence for rhythm abnormalities on all ECGs, Holters, and exercise stress tests was reviewed for existence of complete bundle branch block, ventricular ectopy, supraventricular tachycardia (SVT), ventricular tachycardia (VT), and ventricular fibrillation (VF). All arrhythmias were divided into preoperative, postoperative hospitalization, and outpatient follow-up periods. For the purposes of this study, ventricular ectopy was defined as frequent premature ventricular contractions (PVCs; present on ECG or >10/hour) and ventricular couplets (recorded as rare, occasional, or frequent). Supraventricular (narrow complex) and ventricular (wide complex) tachycardias (≥ 3 beats) were recorded as either sustained (≥ 30 seconds) or nonsustained (<30 seconds). Chart review for any history of antiarrhythmic medications, cardiac arrests, or deaths related to arrhythmia was performed. The timing of arrhythmias from date of reimplantation was compared with progression of LV function and normalization of electrocardiographic findings. Last follow-up was the most recent record with cardiac evaluation.

Results

A total of 44 patients with ALCAPA (21 men, 48%) with available preoperative ECGs were diagnosed at a median age of 3 months (3 days to 13 years). Table 1 summarizes patient demographics and presentation. Nearly all patients (98%) had some electrocardiographic findings, although changes were not consistently seen among all patients. The most common electrocardiographic finding at presentation was deep Q waves and T-wave inversions (Table 2).

Electrocardiographic follow-up was available in 34 of 44 patients (77%). Median follow-up was 6 years, extending to 20 years. Of the 34 patients, 9 (26%) demonstrated complete normalization of their ECG during the follow-up period at a median time of 2 years from the date of surgery. Deep Q waves in lead I, aVL, V5 to V7 tended to resolve before T-wave resolution. Of the 29 of 34 patients with deep Q waves, 82% had resolution in at least 1 lead by their most recent ECG. Abnormal T waves were often the single persistent electrocardiographic abnormality during late follow-up. Most commonly, T-wave inversion persisted in lead I and aVL. Twelve patients in the longitudinal data cohort had LV hypertrophy on ECG before coronary reimplantation with 9 (75%) returning to normal after correction.

In addition to commonly associated findings, 9 patients (20%) demonstrated complete bundle branch block on electrocardiography (4 left bundle, 4 right, and 1 nonspecific). Four were evident preoperatively, with the other 5 developing in the postoperative period. Based on the most recent electrocardiographic data available for each patient, resolution of the bundle branch block occurred in 4 of 9 patients (44%).

Thirty-one patients had preoperative echocardiographic functional measurements and were included in the LV function arm of the study. Preoperative echo data showed.

LV systolic dysfunction in 20 of 31 patients (65%) with 80% of those persisting in the immediate postoperative period. By 1-year follow-up, LV dysfunction was still present in 26% of patients and 16% by 2 years. Complete resolution of LV dysfunction occurred in 94% of patients by their last echocardiogram, at a median time of 1 year (5 days to 4 years) and tended to precede electrocardiographic normalization. Of the 9 patients with complete normalization of electrocardiographic findings, 7 had resolution of LV dysfunction before electrocardiographic normalization. Therefore, 29 of 31 patients (94%) still had electrocardiographic changes at the time of return of LV function.

Arrhythmias occurred in a total of 19 ALCAPA patients (43%). Thirteen patients (30%) developed arrhythmias during their hospital course, 5 occurring before surgical intervention. PVCs or VT documented by ECG almost uniformly demonstrated ectopy or VT arising from the left ventricle. In-hospital arrhythmias (summarized in Table 2) included sustained VF leading to cardiac arrest at presentation in 1 patient, sustained VT in 1 patient, and 4 patients with SVT, 1 of whom required cardioversion. The remaining patients demonstrated high-grade ventricular ectopy. Seven patients were discharged from the hospital on antiarrhythmic medication (6 beta adrenergic blocker, 1 amiodarone) all for antiarrhythmia purposes.

During outpatient follow-up, a total of 11 patients had arrhythmias during outpatient monitoring up to 13 years after surgical correction. Six of these patients had no previous history of in-hospital arrhythmias. In the outpatient setting, arrhythmias most commonly consisted of frequent ventricular ectopy, however in 2 patients, new onset nonsustained VT was documented by Holter (at 2 years and 7 years after surgery), both of whom had a history of