Outcomes of Anomalous Left Coronary Artery From Pulmonary Artery Repair: Beyond Normal Function

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Background. **Anomalous left coronary artery from pulmonary artery (ALCAPA) is associated with high mortality if left uncorrected. We describe long-term outcomes after surgical repair and ventricular function changes on long-term follow-up.**

Methods. **A retrospective review of patients who had ALCAPA repair from January 1996 to December 2011 was completed. Ventricular function was assessed by shortening fraction and ejection fraction, left ventricular end-diastolic dimension, and severity of mitral regurgitation. Speckle tracking echocardiography served as a marker of early myocardial dysfunction.**

Results. **In total, 34 patients underwent ALCAPA repair at median age of 5 months (range, 3 days to 39 years). Surgical interventions included coronary translocation in 31 patients (91%), Takeuchi repair in 2 (6%), and coronary ligation in 1 patient (3%). Concomitant mitral valve repair was performed in 5 patients (15%). No patient required mechanical circulatory support postoperatively. There was no early mortality. At median follow-up of 6 years (range, 1 month to 14 years), there were 5 reoperations: 2 heart transplants (6%), 1 mitral valve replacement (3%), 1 coronary artery bypass graft (3%), and 1 ventricular septal defect closure (3%); there was 1 death 2 years after surgery. Ejection fraction improved from 21% ± 6% to 60% ± 7% ($p = 0.008$), mean shortening fraction from 25% ± 14% to 38% ± 5% ($p = 0.01$), mean left ventricular end-diastolic dimension Z-score from 6.0 ± 3.8 to 0.9 ± 0.7 ($p < 0.001$), and number of patients with moderate-severe mitral regurgitation from 44% (15 of 34) to 5% (1 of 21). All patients had normal ejection fraction and shortening fraction at last follow-up. Speckle tracking echocardiography showed decreased global longitudinal ($p = 0.01$) and circumferential strain ($p = 0.03$) for 11 of 14 patients (79%).**

Conclusions. **Excellent outcomes are achieved with repair of ALCAPA without mechanical circulatory support and with low reintervention rates. Normal ejection fraction and shortening fraction do not accurately convey myocardial dysfunction in these patients.**

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Anomalous left coronary artery from pulmonary artery (ALCAPA) is a congenital coronary abnormality associated with high infant mortality and adult sudden cardiac death [1]. In the absence of adequate collateralization, a coronary steal phenomenon occurs after postnatal changes in cardiopulmonary circulation, resulting in severe myocardial ischemia and dysfunction. After birth, decreased pulmonary vascular resistance causes left to right shunting; blood in the anomalous coronary artery also reverses its flow owing to higher resistance in the coronary system. If left uncorrected, the mortality rate of infants reaches 90% [2]. Extensive collateral arteries may enable some patients to survive beyond infancy. However, chronic hypoperfusion causes subendocardial ischemia and later fibrosis, increasing risk of sudden death secondary to ventricular arrhythmias [1–3].

The standard treatment options for ALCAPA are direct aortic reimplantation or creation of intrapulmonary baffle when coronary translocation is not feasible [4, 5]. Primary ligation has mostly been abandoned. Preservation of the two-coronary system leads to rapid recovery of left ventricular (LV) function, and long-term survival rates are more than 80% [5–7]. In addition, mitral valve regurgitation (MR) from valvular annular dilation or papillary muscle ischemia and secondary dysfunction may also occur in ALCAPA patients. Despite excellent left ventricle recovery and long-term survival rates after ALCAPA repair, follow-up complications such as persistent MR, late-onset congestive heart failure, and arterial stenosis may necessitate reinterventions, including heart transplantation [4–6].

Outpatient management of patients after ALCAPA repair requires ongoing assessment of systolic and diastolic ventricular function. Conventional methods have largely relied on echocardiographic measurements such

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as shortening fraction (SF) or ejection fraction (EF). In addition to routine two-dimensional echocardiographic variables of function, myocardial strain imaging, or deformation imaging, is frequently used for assessing regional myocardial wall motion [8]. Myocardial strain has been an increasingly prevalent adjunct in detecting myocardial dysfunction for congenital heart diseases because of its ability to quantify regional ventricular abnormalities [9, 10]. This technique measures regional ventricular deformation in several directions, and it can identify subclinical dysfunction at earlier stages than traditional methods [11, 12].

The objective of this study was to investigate long-term clinical outcomes of patients after ALCAPA repair and to determine the incidence of early ventricular dysfunction by speckle tracking echocardiography (STE).

Patients and Methods

Data Collection

A retrospective review of charts for patients who underwent surgery for ALCAPA at Texas Children's Hospital from January 1996 to April 2011 was performed. Patients whose primary ALCAPA repair was performed at another hospital or whose ALCAPA repair was not the primary operation were excluded. Data were collected from electronic databases (congenital heart surgery, pediatric cardiology and echocardiography databases) and clinical records. Institutional Review Board approval was obtained before starting this study.

Demographic variables included sex, age, and weight at surgery, ethnicity, previous ALCAPA interventions, preoperative arrhythmias, use of mechanical ventilation, and inotropic support. Operative variables analyzed include type of ALCAPA repair, mitral valve intervention, additional surgical procedures performed, duration of mechanical ventilation, duration of intensive care unit and hospital stay, postoperative complications, and discharge status. Ventricular function was assessed by standard echocardiographic methods: SF and EF, left ventricular end-diastolic dimension (LVEDD), and MR. The composite endpoint was defined as death, transplantation, or reoperation.

Surgical Technique

CORONARY ARTERY TRANSLOCATION. After standard cardiopulmonary bypass and institution of cardioplegia, the left coronary artery was harvested with a large button of pulmonary arterial wall and widely mobilized without injuring any branches. After inspection of the aortic valve (usually through a separate aortotomy), an aortic flap was performed to minimize torsion of the vessel. The coronary button was anastomosed to the aortic wall with fine polypropylene suture. The pulmonary arterial trunk was reconstructed with a patch of autologous pericardium.

INTRAPULMONARY BAFFLE (TAKEUCHI) REPAIR TECHNIQUE. After initiation of cardiopulmonary bypass and diastolic cardiac arrest, a pulmonary arteriotomy was performed, creating a transverse flap of pulmonary artery tissue. An aortopulmonary window was created, and the pulmonary artery flap was used to baffle the left coronary artery into the aorta. The pulmonary artery was then reconstructed with autologous pericardium.

Myocardial Strain

Prospective examination of myocardial strain was performed on patients at 1 year or after surgery. The technique aims to differentiate between active and passive movement of myocardial segments, to measure intraventricular dyssynchrony and to evaluate components of myocardial function, such as longitudinal or circumferential myocardial shortening, that are not assessable by standard echocardiographic techniques. The degree of left ventricle deformation is reported as percentages of global longitudinal and circumferential peak systolic strain. Decreased myocardial shortening (impaired ventricular function) is represented by a higher (less negative) percentage number. Normal values vary by age. That provides potential for determining changes in function before EF or SF have been affected [13]. Global two-dimensional strain measurements by STE have been found to detect myocardial dysfunction before it is manifested by decreased EF or SF [14].

Of the 34 patients in the study, 14 (41%) agreed to participate in the STE data collection. The technique of STE was applied to obtain an average global peak systolic longitudinal strain and peak systolic longitudinal strain rate, with systole manually defined by aortic valve closure. After initial tracing of the endocardial border and software processing, the operator confirmed adequate tissue tracking. Segments unable to be adequately tracked were excluded. Peak radial strain and strain rate were also assessed from the parasternal short-axis view at the basal level. All measures were made by a single observer.

Statistical Analysis

Continuous variables were reported as medians with minimum and maximum or means with standard deviations. Categoric variables were reported as frequencies with percentages. Mortality, transplantation, and reoperations were modeled as time-related events using the Kaplan-Meier method and are reported as freedom from the composite endpoint. Myocardial strain values were compared with reference benchmarks by age, as described by Marcus and colleagues [15]. For every patient, the strain percentage was normalized using the mean and standard deviation for the corresponding age group according to the published benchmarks. Normalized values were compared with corresponding benchmarks using Students *t* test.

Results

Demographics and Preoperative and Postoperative Course

There were 34 patients (44% female) who underwent surgery for ALCAPA. Median age at time of repair was 5