Aortic Implantation of Anomalous Origin of the Left Coronary Artery From the Pulmonary Artery: Long-Term Outcomes

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Background. **Since 1989 all patients with anomalous origin of the left coronary artery from the pulmonary artery at our institution have been treated with aortic implantation. The purpose of this review was to assess the late outcomes of these patients, especially regarding left ventricular (LV) function and mitral valve insufficiency.**

Methods. **Between 1989 and 2014, 36 patients had aortic implantation of anomalous origin of the left coronary artery from the pulmonary artery. Mean age at surgery was 2.5 ± 5.1 years (median, 0.5 years). Operative strategy included antegrade cold-blood cardioplegia, main pulmonary artery transection, aortic implantation with a large button of pulmonary artery, pulmonary reconstruction with fresh autologous pericardium, and prolonged postoperative inotropic and ventilator support. Mitral regurgitation and LV dysfunction were graded as 0 to 4 (0 = none, 1 = trivial, 1.5 = trivial-mild, 2 = mild, 2.5 = mild-moderate, 3 = moderate, 3.5 = moderate-severe, and 4 = severe).**

Results. **Mean mitral regurgitation grade preoperatively was 2.95 ± 0.95. Mean LV dysfunction grade was 3.14 ± 1.27. Mean cross-clamp and cardiopulmonary bypass times were 49.1 ± 18 minutes (median, 48.5 minutes) and 147.5 ± 45 minutes (median, 139 minutes), respectively. There was no operative or late mortality. Four patients had delayed sternal closure. Mean duration of ventilator support was 11 ± 6.6 days (median, 9 days). Two patients required 3 and 6 days of postoperative extracorporeal mechanical circulatory support. Mean length of stay was 25 ± 18 days (median, 19 days). No patient has required reoperation for supravalvar pulmonary stenosis, coronary stenosis, or mitral valve repair or replacement. Late echocardiographic follow-up shows a mean mitral regurgitation grade of 1.67 ± 1.05 and a mean LV dysfunction grade of 0.23 ± 0.68.**

Conclusions. **Aortic implantation is our procedure of choice for patients with anomalous origin of the left coronary artery from the pulmonary artery. No patient required mitral valve repair or transplant. There was marked improvement of mitral regurgitation grade, return to essentially normal LV function, and no mortality during a 25-year period.**

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Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital defect that causes coronary artery steal with ischemia-induced left ventricular (LV) dysfunction and mitral regurgitation (MR). Bland and colleagues [1] proposed the pathophysiologic effects of coronary steal and ensuing myocardial ischemia in 1933. If left uncorrected, there is a high rate (92% to 100%) of mortality during infancy [2, 3].

Several techniques [4–6] for the surgical management of ALCAPA have been reported since Sabiston and associates [7] described ligation of the left coronary artery at the pulmonary artery origin in 1960. In 1974, Neches and colleagues [8] reported the direct implantation of the left coronary artery into the aorta with good results, and that is now accepted as the procedure of choice [9]. Recovery of LV function after creation of a dual coronary system is well documented [10–14]. However, controversy still exists over the initial management of MR, with some surgeons recommending mitral valve repair at the time of ALCAPA repair [15, 16].

Since 1989, all patients with ALCAPA at our institution have been treated with direct aortic implantation without a concomitant mitral valve procedure. The purpose of this review was to analyze our surgical strategy and to assess the late outcomes, particularly regarding LV function and the evolution of mitral valve regurgitation.

Material and Methods

Permission to review the health records and follow-up information was obtained from Ann & Robert H. Lurie

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Abbreviations and Acronyms

ALCAPA = anomalous origin of the left coronary artery from the pulmonary artery
BSA = body surface area
CPB = cardiopulmonary bypass
ECMO = extracorporeal membrane oxygenation
LV = left ventricular
LVEDD = left ventricular end-diastolic dimension
MR = mitral regurgitation
TTE = transthoracic echocardiography

Children's Hospital of Chicago Institutional Review Board committee. The need for consent from the individuals or parents was waived. A retrospective chart review was performed.

Upon diagnosis, all patients were taken for urgent operation. Our surgical technique has been previously described [17]. Aortobicaval cannulation is accomplished. An LV vent is placed by means of the right superior pulmonary vein. After administering cardioplegia with the pulmonary arteries snared, the main pulmonary artery is transected. A large coronary ostial button is harvested and the left coronary artery mobilized. An opening, 30% to 40% smaller than the ostial button, is created on the left posterolateral aspect of the aorta, allowing the enlarged ostial button to act as a funnel for elongation of the left coronary artery. Direct implantation is then performed. The defect in the sinus of the pulmonary artery is reconstructed with an autologous pericardial patch. Left atrial pressure is monitored through the vent site during weaning from cardiopulmonary bypass (CPB). Initiation of mechanical assistance is considered if the left atrial pressure is greater than 20 mm Hg, with associated hypotension, suggestive of severe LV dysfunction.

Patients were supported with inotropic therapy (dopamine, 3–10 $\mu g \cdot kg^{-1} \cdot min^{-1}$; epinephrine, 30–100 $\mu g \cdot kg^{-1} \cdot min^{-1}$; and milrinone, 0.3–0.5 $\mu g \cdot kg^{-1} \cdot min^{-1}$) while undergoing mechanical ventilation until improvement in LV function was observed. Several patients required prolonged (>1 month) management with inotropic therapy. Patients were discharged home on a combination of a diuretic (furosemide, spironolactone, or diuril), angiotensin-converting enzyme inhibitor (captopril, enalapril), and digoxin, the duration of which was left to the discretion of the patient's cardiologist.

Standard clinical echocardiographic reports, obtained from an Intersocietal Accreditation Commission Echocardiography–certified laboratory, were reviewed. As this was a retrospective chart review, the cardiologist reading the examination was unaware of the patient's participation in this study. The preoperative, discharge, and most recent late (>6 months postoperative) echocardiogram reports were included. Descriptions of MR and LV dysfunction were given a numeric grade: 0 = none, 1 = trivial, 1.5 = trivial-mild, 2 = mild, 2.5 = mild-moderate, 3 = moderate, 3.5 = moderate-severe, and 4 = severe. In patients not followed up at our institution, attempts were made to obtain the echocardiographic reports from the patient's cardiologist.

Results

Between 1989 and 2014, 36 consecutive patients underwent aortic implantation for ALCAPA. There were 18 females and 18 males. Mean age at surgery was 2.5 ± 5.1 years, (median, 0.5 years; range, 14 days to 18.4 years). Twenty patients were younger than 6 months, 7 were between 6 and 12 months, and 9 were older than 1 year. Weight ranged from 2.9 to 54 kg (median, 6.1 kg). Two patients presented in extremis, whereas 34 presented with signs of congestive heart failure. Diagnosis was made by both transthoracic echocardiography (TTE) and cardiac catheterization in 18 patients, but solely by TTE in 18 patients. Twenty-eight patients had evidence of an anterior or anterolateral myocardial infarction pattern (Q waves in leads I, aVL, and V4–V6) by electrocardiogram. Cardiomegaly (cardiothoracic ratio > 0.5) was observed in 29 patients.

In 26 patients, the left coronary artery originated from sinus I (posterior facing sinus; Fig 1). In 1 patient the left coronary artery originated from the nonfacing sinus. The anomalous coronary artery arose from the main pulmonary artery in 3 patients, the right pulmonary artery in 2, and at the junction of the right and main pulmonary arteries in 2. The origin of the ALCAPA was not specified in 1 patient.

The mean CPB time was 147.5 ± 45 minutes (median, 139 minutes). The mean aortic cross-clamp time was 49.1 ± 18 minutes (median, 48.5 minutes). Four patients underwent delayed sternal closure. Mean duration of ventilator support was 11 ± 6.6 days (median, 9 days). The mean length of hospital stay was 25 ± 18 days (median, 19 days).

Two patients required mechanical assistance for a duration of 3 and 6 days. The first child was a 2-month-old boy who presented with severe LV dysfunction with a shortening fraction of less than 15%. On weaning from CPB, the patient had severe LV dysfunction with a left atrial pressure greater than 25 mm Hg. The patient was supported with extracorporeal membrane oxygenation (ECMO) for 6 days to allow LV recovery. The second child was a 5-week-old girl who presented in extremis. After repair of ALCAPA, the patient exhibited severe LV dysfunction with moderate to severe MR. Although the patient was able to remain off CPB for 2 hours, the child was placed on ECMO because of the high inotropic support required to maintain marginal ventricular function. The patient subsequently had her cannulas removed on the second postoperative day. At last follow-up, the patient had trivial MR and normal LV function.

Preoperative and discharge TTE reports were available for 34 patients, whereas 22 had late follow-up echocardiographic information (>6 months after repair; Fig 2). Preoperative MR was trivial in 1 patient, mild in 8,