Twenty-Year Outcome of Anomalous Origin of Left Coronary Artery From Pulmonary Artery: Management of Mitral Regurgitation

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Background. **This study is a single-center experience with surgical repair of anomalous origin of left coronary artery from pulmonary artery (ALCAPA) with focus on the management of associated mitral regurgitation (MR).**

Methods. **We performed a retrospective analysis of cases presenting to a quaternary referral center between November 1990 and October 2011.**

Results. **In all, 25 patients (18 female) presented with a diagnosis of ALCAPA at a median age of 5 months (range, 1.5 to 102). Twenty-one patients (84%) had moderate to severe impairment of left ventricular function with median fractional shortening of 14% (range, 2% to 33%), and 19 patients (76%) had moderate to severe MR. Surgery was performed with direct coronary reimplantation in 16 patients (64%) and intrapulmonary tunnel (Takeuchi repair) in 9 (36%). Four patients had mitral valve repair at time of surgery, all for structural anomalies. Functional MR with a structurally normal mitral valve was not repaired. The median duration of postoperative follow-up was 93 months (range, 9 to 240). There were no early or late deaths, and no patient required mechanical support. Four patients (16%) required surgical or catheter reintervention. At last follow-up, 24 of 25 patients were asymptomatic; the left ventricular function was normal in 22 patients. Moderate MR was present in 4 patients. There was significant improvement in left ventricular function and MR ($p < 0.01$) during follow-up.**

Conclusions. **Surgical repair of ALCAPA has good long-term results with low mortality and reintervention rates. The majority of MR is functional and will improve with reperfusion, but structural mitral valve abnormalities should be repaired at the time of surgery.**

(Ann Thorac Surg 2014;97:938–44)

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital cardiac lesion with an incidence of 1 in 300,000 live births [1]. It constitutes 0.25% to 0.5% of congenital heart disease [2]. It causes myocardial ischemia with left ventricular (LV) dysfunction and mitral regurgitation (MR). If untreated, survival beyond infancy is rare [2, 3]. The first clinical description of ALCAPA was reported by Edward Bland, Paul Dudley White, and Joseph Garland in 1933, so the anomaly is also called Bland-White-Garland syndrome [4]. ALCAPA was classified into infantile (early presentation) and adult (late presentation) types based on the age and mode of presentation [5]. Neonates are usually asymptomatic, as high pulmonary arterial pressure in the neonatal period maintains the antegrade flow into the left coronary artery (LCA). As the pulmonary vascular resistance drops, there is retrograde flow of blood from the LCA into the pulmonary artery, resulting in coronary steal and myocardial ischemia. If there is insufficient collateral blood supply to the left ventricle, patients present during early infancy with cardiac failure. The patients with extensive collateral blood supply can survive into adulthood. However, these patients are likely to present in later life with arrhythmias, chest pain, cardiac failure, and sudden death.

Surgical repair for establishing a two coronary artery system has evolved over a period of time with direct implantation being now the procedure of choice [9–15]. The early presentation group are usually patients in poor condition with impaired LV function. The late presentation group have preserved LV function with MR. Concomitant mitral valve (MV) surgery with ALCAPA repair has been the area of debate because the majority of MR is functional with a structurally normal MV. There are limited studies describing the long-term outcomes of ALCAPA [6–8, 16, 17], and the importance of addressing the MR remains unclear.

Accepted for publication Nov 11, 2013.

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Published by Elsevier Inc

0003-4975/$36.00
http://dx.doi.org/10.1016/j.athoracsur.2013.11.042

Abbreviations and Acronyms

ALCAPA = anomalous origin of the left coronary artery from the pulmonary artery
DCM = dilated cardiomyopathy
FS = fractional shortening
LCA = left coronary artery
LV = left ventricular
MPA = main pulmonary artery
MR = mitral regurgitation
MV = mitral valve

The purpose of this study was to evaluate a single pediatric cardiac surgical center's experience in the diagnosis, management, and long-term outcome of ALCAPA with focus on the management of MR.

Material and Methods

A retrospective case record review was made of all patients with the diagnosis of ALCAPA presenting to our department over the period from November 1990 to October 2011 as identified by our departmental database. The study was reviewed and approved by the local research and clinical audit departments. The need for individual consent was waived.

Patient Demographics

Twenty-five patients (7 male and 18 female) with a diagnosis of ALCAPA were identified during the study period. The median age at presentation was 5 months (range, 1.5 to 102). Sixteen patients (64%) presented in infancy (early presentation group), and 9 (36%) presented after 1 year of life (late presentation group). Three patients presented with an asymptomatic murmur, 2 patients presented with episodes of screaming and sweating with feeds, and 20 patients presented with failure to thrive and clinical features of cardiac failure.

Echocardiography with color Doppler was used to make the diagnosis of ALCAPA and associated lesions. The LV systolic function was assessed and expressed as fractional shortening (FS). The degree of MR was expressed as mild, moderate, or severe on color Doppler. Cardiac catheterization was performed if necessary to confirm the diagnosis.

Surgery

Surgery was performed for all 25 patients immediately after confirming the diagnosis of ALCAPA. The median age of surgery was 5.5 months (range, 1.80 to 102), with a median weight of 5.81 kg (range, 3.99 to 30.8 kg). Sixteen patients (64%) had direct implantation of LCA into aorta, and 9 patients (36%) had intrapulmonary tunnel (Takeuchi procedure). The patients included a 20-month-old girl who was diagnosed as having ALCAPA at 10 months of age and had a left internal mammary graft placed to the LCA in a different cardiac center, as the ostium of LCA was thought to be too far away to be reimplanted into the aorta. This patient continued to have moderate to severe MR, and cardiac catheter study had shown blocked internal mammary graft. She successfully underwent a Takeuchi procedure and MV cleft repair in our institution.

Five patients had associated MV structural abnormalities (three MV clefts, one MV prolapse, and one dysplastic double orifice MV), 1 patient had ventricular septal defect, 1 patient had atrial septal defect, and 1 patient had significant branch pulmonary artery stenosis. All the patients who had structural MV problems were in the late presentation group. Four patients had MV repair (all for structural anomalies), and 1 patient had pulmonary arterioplasty at the time of first surgery. Patients with functional MR but a structurally normal MV did not undergo any attempt at MV repair. The coronary anatomy of patients was as follows: arising from the left and posterior sinus of the main pulmonary artery (MPA) in 15 (60%; Figs 1A, 1B); left and lateral side of MPA in 8 (32%; Figs 1C, 1D); at the junction of the MPA and right pulmonary artery in 1 (4%; Figs 1E, 1F); and high up laterally in the ascending part of the MPA in 1 (4%). The median bypass time was 94 minutes (range, 38 to 351), and the median cross-clamp time was 50 minutes (range, 33 to 97). There were no operative deaths. The median bypass time in the direct implantation group was 92 minutes (range, 38 to 351), and in the Takeuchi procedure group, it was 95 minutes (range, 64 to 141; $p = 0.28$). The median cross-clamp time in the direct implantation group was 54 minutes (range, 33 to 88), and in the Takeuchi procedure group, it was 47 minutes (range, 38 to 97; $p = 0.45$).

The data on follow-up with last clinical and echocardiographic evaluation and any further surgical or catheter intervention were noted for the purpose of study. The data were analyzed with the Mann-Whitney U test, Fisher's exact test, and χ^2 test with Yates' correction for the statistical significance of variables. A p value of less than 0.05 was taken as the level of significance. Kaplan-Meier curves were used to analyze the reintervention rates.

Results

Nineteen of 25 patients with a median age of 4.5 months (range, 1.5 to 102) were diagnosed on initial presentation. Six of 25 patients (24%) with a median age of 40 months (range, 15 to 66) had a delay in diagnosis by 9 to 34 months (median 11.5). The latter were followed up for mitral regurgitation in 5 and presumed dilated cardiomyopathy (DCM) in 1.

At the time of presentation, 21 patients had moderate to severe impairment of LV systolic function with a median FS of 14% (range, 2% to 33%), and 19 patients (76%) had moderate to severe MR (Fig 2). The early presentation group of patients, however, were much sicker with poor LV systolic function at the time of presentation ($p < 0.001$) and a median FS of 10% (range, 5% to 33%) when compared with the late presentation group with a median FS of 26% (range, 18% to 31%). There was no