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Repair of anomalous coronary artery from the pulmonary artery: A-signal center 20-year experience

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ABSTRACT

Background: Anomalous origin of coronary artery from the pulmonary artery (ACAPA) is a rare congenital coronary malformation with a high mortality whether in infants or adult patients. This study reviews 20 years of surgical treatment in a single center and aims to establish the optimal surgical strategies for this rare pathology.
Methods and results: From April 1994 to March 2015, 96 consecutive patients aged from 3 months to 60 years underwent coronary repair surgery. The surgical procedures included ligation (3 cases), ligation along with CABG (6 cases), transpulmonary baffling (Takeuchi Procedure, 14 cases) and directly implantation of the anomalous coronary artery (ACA) into the aorta (73 cases). Postoperative extracorporeal mechanical circulatory support (ECMO) was necessary in 4 cases. Mitral valve repair was performed in 40 patients with moderate or severe mitral regurgitation (MR). Mitral replacement was performed in one patient with severe MR.
There were one early and two late deaths. One patient underwent a second operation because Baffle leaks. During mean 10.45 ± 8.96 year follow-up (1 month–18 years), both early and late improvement of left ventricular function was observed in most patients (8 patients lost of follow-up).
Conclusions: The establishment of a two-coronary system is the main goal of surgical therapy today. In different procedures, the direct implantation of the ACA into the ascending aorta is the best method and has good long-term results. ECMO as a bridge to recovery that will play an integral part in moderns' surgical treatment.

1. Introduction

Coronary artery anomalies are not uncommon, occurring in 1.3% (range = 0.3–5.6%) of the population [1]. Anomalous origin of the coronary artery from the pulmonary artery (ACAPA) is one part of coronary artery anomalies. ACAPA can be one of 4 types [2]. More than 90% of the encountered and studied anomalies are anomalous origins of the left coronary artery from the pulmonary artery (ALCAPA) [2].

ALCAPA is a rare congenital anomaly which is usually seen as an isolated lesion and is present in 1/300,000 live births (0.25 to 0.5%) [3,4], and is also known as Bland-White-Garland syndrome [5]. Anomalous origin of right coronary artery from pulmonary artery (ARCAPA) is even rare compared to ALCAPA, accounting for only 0.002% of cases of congenital heart disease [6].

Different techniques such as ligation of the anomalous coronary artery, ligation or closure of the anomalous coronary artery with CABG, creation of an intrapulmonary tunnel with an aortopulmonary window, creation of an extrapulmonary composite tunnel, and direct implantation have been used to treat these anomalies.

Nowadays successful surgical repair basically depends upon the re-establishment of a two-coronary system [7]. Direct implantation of the anomalous coronary artery (ACA) into the aorta has become the procedure of choice whenever possible [8,9].

In the last 20 years, the cardiac technique developed very quickly in our hospital. Our surgical technique has evolved. We use the previously described techniques to repair ACAPA, mostly with good results. We are reporting our experience with these different techniques and assess the postoperative cardiac recovery of cardiac function and long-term outcomes.

2. Material and methods

The surgical database of our hospital was retrospectively reviewed for surgical correction cases performed for anomalous origin of the coronary artery from the pulmonary artery, between April 1994 and March 2015. Clinical, operative, and outcome data of the patients were recorded. Approval of this study was obtained from the Research Ethics Board at our hospital.

3. Patient demographics

Ninety-six patients (54 male and 42 female) with a diagnosis of ACAPA were identified during the study period. This includes

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80(83.3%) ALCAPA patients and 16(16.7%) ARCAPA patients. The median age at presentation was 14.2 ± 17.4 years (range, 3 months to 60 years). The median weight was 29.1 ± 24.4 kg (range, 4 to 86 kg). Thirty two patients (33.3%) presented in infancy (early presentation group), and 64 patients (66.7%) presented after 1 year of life (late presentation group). Twenty three patients presented with an asymptomatic murmur, 32 patients had effort angina, and 44 patients had complaints of fatigue, dyspnea or palpitation. 12 infant patients presented with poor feeding and low weight, 8 with persistent tachypnea, 7 patients had myocardial ischemia or heart infarction, and 67 patients presented with heart failure to thrive and clinical features of cardiac failure.

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

4. Surgery

Surgery was performed for all 96 patients after confirming the diagnosis of ARCAPA. Three patients (3.1%) had ligation, six patients (6.3%) had ligation along with CABG, fourteen patients (14.6%) had intrapulmonary tunnel (Takeuchi procedure), and seventy three patients (76%) had direct implantation of the anomalous coronary artery. In early presentation group, all 32 cases underwent direct implantation of the anomalous coronary artery. The patients included a seven-year-old girl who was diagnosed with MR and had mitral valve repair at the age of three at another hospital. This patient continued to catch colds, and she was diagnosed with ALCAPA which was confirmed by CT in our hospital. Then she successfully underwent direct implantation of LCA into the aorta.

Twenty patients had associated MV structural abnormalities (five MV clef, fifteen MV prolapse), 3 patients had atrial septal defects, 2 patients had tetralogy of Fallot, 2 patients had ventricular septal defects, and 1 patient had patent ductus arteriosus. 12 patients who had structural MV problems were in the early presentation group. All 20 patients with structural MV problems had MV repair, and other 20 patients (in the late presentation group) with moderate or severe functional MR underwent MV repair at the time of the first surgery. Among the patients who were diagnosed of ALCAPA, the site of origin of the left coronary artery was, in order of frequency, posterior pulmonary trunk wall (43.8%, 35/80), inner wall (33.7%,27/80), and lateral wall (22.5%,18/80). The right coronary anatomy of patients was as follows: arising from posterior pulmonary trunk wall in 5(31.2%), inner wall in 9(56.3%), and lateral wall in 2(12.5%). Operations were performed with cardiopulmonary bypass in 92 patients (95.8%). The median bypass time was 148 ± 86 min (range, 59 to 648), the median cross-clamp time was 89 ± 35 min (range, 33 to 205). The median bypass time in the early presentation group was 160 ± 114 min (range, 72 to 648), and in the late presentation group was 135 ± 72 min (range, 59 to 465, $p = 0.27$). The median cross-clamp time in the early presentation group was 82 ± 32 min (range, 33 to 159), and in the late presentation group was 89 ± 41 min (range, 39 to 205, $p = 0.35$).

The follow-up data with the last clinical and echocardiographic evaluation and any further surgical interventions were noted for the purpose of the study. Ventricular function was classified as normal or mildly impaired (ejection fraction [EF] > 0.5), moderately impaired (EF 0.35 to 0.5), and severely impaired (EF < 0.35). Mitral regurgitation (MR) was evaluated with Doppler echocardiography (0 = no MR; 1 = mild MR; 2 = moderate MR; 3 = severe MR).

The composite endpoint was defined as left ventricular EF (LVEF) less than 0.5, MR grade 2 or more, or death after hospital discharge after initial operation.

5. Statistics

Descriptive data for continuous variables are presented as mean ± standard deviation or as medians with range; categoric variables are presented as relative frequencies. The Fischer exact test was performed to detect significant differences between groups. For comparison of continuous variables between two groups, the *t*-test was used. The probability of freedom from events was estimated according to the Kaplan–Meier method. Freedom-from-events curves were compared by means of the log-rank test. The Wilcoxon signed-rank test for related data was used to analyze the differences between preoperative EFs values and postoperative EFs values at different time points. Values of p less than 0.05 were considered as statistically significant. Analyses were performed with SPSS 19.0 for Windows (SPSS Inc., Chicago, IL).

6. Results

6.1. Early postoperative results

The median duration of intensive care unit stay was 4.3 days (range, 1 to 32), with the early presentation group staying for a median of 7.97 days (range, 1 to 32) compared with patients in the late presentation group staying for a median of 2.84 days (range, 1 to 8, $p = 0.002$) (Table 1). Compared with the late presentation group, patients in the early presentation group had a lower preoperative EF ($p < 0.001$). The degree of preoperative MR in ALCAPA was more serious than ARCAPA patients, with significant differences ($p = 0.002$) (Table 2). As mentioned above, simultaneous mitral annuloplasty at the anterolateral commissure was performed in 40 patients with polytetrafluoroethylene sutures. There were several plasty techniques, including Kay–Reed type annuloplasty, folding of the unsupported component and commissure plasty approximated with several interrupted sutures. Four patients required extracorporeal membrane oxygenation support. The duration of ECMO support was 2, 7, 8 and 16 days, respectively. ECMO support was successfully weaned in all 4 patients with improvement of hemodynamics and cardiac function, with all 4 patients being finally discharged without significant ECMO-related complications.

There was only one early death in an infant who was diagnosed with ARCAPA. He had an atrial septal defect repaired at the same setting as reimplantation from the coronary artery. The patient died of severe pulmonary infection after staying in ICU for 32 days. The other patients had uneventful recoveries.

Table 1
Preoperative variables of 96 patients with ACAPA.

Characteristics	Early presentation group ($n = 32$)	Late presentation group ($n = 64$)	p-Value
Gender: male, n (%)	18 (56.25%)	36 (56.25%)	>0.99
Mean age at operation (years)	0.73 ± 0.28	19.48 ± 17.75	<0.001
Mean weight at operation [kg]	7.77 ± 2.25	39.74 ± 23.01	<0.001
Preoperative MR > I, n (%)	14 (43.75%)	26 (40.63%)	0.828
Preoperative EF (%)	42.52 ± 17.93	59.44 ± 8.64	<0.001
Anomalous origin of the coronary			0.311
Posterior	17 (53.12%)	27 (42.19%)	
Inner	11 (34.38%)	22 (34.37%)	
lateral	4 (12.50%)	15 (23.44%)	
Mean cardiopulmonary bypass time [minutes]	159.69 ± 114.27	134.86 ± 72.17	0.260
Mean aortic cross-clamp time [minutes]	81.84 ± 32.18	89.11 ± 40.83	0.345
Mean ICU time [days]	7.97 ± 9.20	2.84 ± 1.44	0.002

ACAPA = Anomalous origin of the coronary artery from the pulmonary artery; EF = Ejection fraction; MR = Mitral regurgitation; ICU = Intensive Care Unit.