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Midterm outcome after surgical correction of an anomaly of the left coronary artery from the pulmonary artery

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Abstract

Objective This study aims to present the midterm outcomes of surgical correction of the anomalous left coronary artery from the pulmonary artery (ALCAPA).

Methods This is a retrospective study of patients undergoing anomalous origin of the LCA from the pulmonary artery repair between 2010 and 2019.

Results Forty-nine patients (20 boys and 29 girls) underwent ALCAPA repair. Patients were divided into two groups based on their age at ALCAPA repair: infant (< 1 year of age: $n = 24$) and non-infant (≧ 1 year of age: $n = 25$). Median age at time of repair was 23 months(7-60months). LCA reimplantation was performed in 47 patients, and Takeuchi repair was performed in 2 patients. Hospital mortality in the infant group was 8.2% (4 of 49). Infant group had significantly lower LVEF in pre-operation ($p < 0.05$), but there was not significantly different between the two groups about LVEF at discharge. The median follow-up duration was 43(18–85)months. The freedom from reoperation was not significantly different between two groups (infants vs. non-infants: 68.8% vs. 87.5% at 10 years; $p = 0.096$).

Conclusions Surgical treatment of ALCAPA had an excellent early and midterm outcomes. Left ventricular dysfunction in pre-operation was the main risk of mortality in-hospital. The freedom from reoperation did not differ significantly between infant group and non-infant group.

Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital heart disease in children, with an estimated incidence of 1 in 300,000 live births [1]. Myocardial ischemia is the main pathological manifestation, often leading to left ventricular hypofunction and mitral regurgitation [2]. Early establishment of dual coronary circulation is currently the main surgical approach. Approximately 90% of patients who are left untreated die within the first year of life [3]. Proper evaluation and management, including diagnostic techniques and medical treatment, can result in satisfactory clinical outcomes.

Left ventricular function and mitral regurgitation were major factors associated with hospital mortality and reoperation. Because infants lack collateral vessels, left heart function and mitral regurgitation tend to be severe while operation. However, postoperative recovery is unclear compared with that in the non-infants. Many studies have explored the need for concomitant mitral valve (MV) repair to reduce the incidence of reoperation

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[4–7]. However, few studies have compared left ventricular function and reoperation between different age groups.

The purpose of this study was to review the midterm surgical results of ALCAPA repair, evaluate the recovery of left ventricular function and rates of reoperation.

Patients and methods

Patients

This retrospective study included patients who underwent surgery for ALCAPA at a single center from June 2010 to June 2019. Medical records included patient demographics, preoperative data, surgical management data, perioperative complications, and postoperative outcomes. In-hospital mortality was defined as death occurring within 30 days of surgery or before hospital discharge. The follow-up data included mortality and complications. The echocardiographic parameters included ejection fraction (EF) and Fractional shortening (SF), and the degree of MV regurgitation was graded as none or trivial, mild, moderate or severe.

Ethical approval

Given the retrospective design of the study, which is based on data collected for routine clinical care, administrative, and audit purposes, individual informed consent was not required. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. This study has approval by the institutional research committee at Beijing Anzhen hospital, who permitted the collection of data for audit and research purposes. The approval number is 202417X. We are permitted to collect data for audit and research purposes.

Surgical technique

Left coronary artery (LCA) reimplantation was the first choice for re-establishing the dual coronary system in this study. A standard cardiopulmonary bypass (CPB) was performed for this procedure. Before the initiation of CPB, temporary blockade of the right and left pulmonary arteries to avoid coronary mal-perfusion. Surgery was carried out using conventional cardiopulmonary bypass at 28–32 °C.

Coronary button transfer is the preferred method for LCA reimplantation. When the length of the LCA was not sufficient, coronary elongation with an autologous pericardium tube was used to increase the length of the LCA. Due to the different choices of surgical methods made by different operators, twelve patients underwent coronary elongation with an autologous pericardium tube. Takeuchi repair can connect the anomalous left coronary artery and aorta by creating an intrapulmonary baffle. Then, the pulmonary artery was reconstructed with autologous pericardium.

Some patients with moderate or severe mitral regurgitation detected by echocardiography underwent concomitant mitral annuloplasty. Surgical strategy was to annul the posterior mitral junction to reduce mitral regurgitation.

Statistical analysis

All the data were analyzed by using SPSS 24.0 software. Quantitative data are presented as the mean, standard deviation or median (range). Qualitative values are expressed as percentages. Patients were divided into two groups: infant group and non-infant group. For comparisons of normally distributed variables between two groups, we used the independent sample t test; for comparisons of categorical variables between groups, the chi-square test was used. K–M survival curves were generated and compared with the log-rank test according to the clinical subgroups. One-, 5-, and 10-year survival were estimated for each of the groups. And $p<0.05$ was considered to indicate statistical significance.

Results

Perioperative data

During the past 10 years in our center, 49 patients (20 boys and 29 girls) underwent ALCAPA repair. The baseline characteristics are described in Table 1. Patients were divided into two groups based on their age at ALCAPA repair: infants (<1 year of age: $n=24$) and noninfants (≥1 year of age: $n=25$). Seventeen patients (34.7%) had symptoms of respiratory failure or chest tightness, and the remaining symptoms were detected via examination. There were 24 patients with the Infancy subtype in this study. The median age at the time of repair was 23 months (7–60 months). Left coronary artery anomalies originating in the right sinus of the pulmonary artery in thirty-three patients. Sixteen patients had left coronary artery anomalies originating from the left sinus of the pulmonary artery. LCA reimplantation was performed in 47 patients, and Takeuchi repair was performed in 2 patients. Twelve patients received autologous pericardium to increase coronary length during LCA reimplantation. And four patients underwent ventricular aneurysm resection, two patients had 2 cm ventricular aneurysm, another had a ventricular aneurysm of 4 cm, and the fourth patient, who had a ventricular aneurysm of 6 cm, was treated with ECMO after operation, and died on 8th. LVEF was significantly lower in infant group than non-infant group($p<0.05$), but there was no significantly of LVEF at discharge between two groups. Thirty-four patients experienced MR before the operation. Thirty patients underwent concurrent surgical repair of

Table 1 Patients' characteristics and outcomes between infant and non-infant groups ($n=49$)

Characteristics	Infant($n=24$)	Non-Infant($n=25$)	*p*-Value
Males	8(33.3%)	12(48%)	0.226
Age(years)	7(4–9)	58(32-119.2)	0.000
Weight(Kg)	6.9 ± 2.0	22.4 ± 2.7	0.000
Height(cm)	68.1 ± 8.5	103.5 ± 39.7	0.000
Symptom	9(37.5%)	8(32%)	0.458
Infancy subtype	14(58.3%)	4(16%)	**0.024**
Associated lesions			
ventricular septal defect	1(4%)	0	
patent ductus arteriosus	0	1(4%)	
patent foramen ovale	4(16.7%)	1(4%)	
Mitral regurgitation, n (%)	18(75%)	16(64%)	0.300
None/trivia	1(4.2%)	3(12%)	
Mild	6(25%)	6(24%)	
Moderate	6(25%)	5(20%)	
Severe	5(20.8%)	2(8%)	
Surgery type			
Reimplantation	15(62.5%)	20(80%)	
Coronary elongation	7(29.2%)	5(20%)	
Takeuchi repair	2(8.3%)	0	
CPB-time(min)	184.9 ± 66.8	156.4 ± 39.4	0.075
ACC-time (min)	92.0 ± 25.8	87.2 ± 17.1	0.449
Mechanical ventilation time (hour)	82.5(52.5–168)	21.5(17.0–65.0)	**0.000**
ICU-stay(days)	18(7.3–23)	10(7–15)	0.056
LVEF at preoperation, %	47.7 ± 14.1	59.0 ± 15.5	**0.018**
LVEF at discharge, %	55.2 ± 14.0	63.3 ± 8.5	0.052
LVEF at follow-up, %	67.8 ± 5.1	64.0 ± 4.5	0.026
Ventricular aneurysm	4(17.4%)	2(7.7%)	0.314
Delayed sternal closure	3(12.5%)	1(4.0%)	0.289
ECMO	3(12.5%)	0	0.110
Death	4(16,7%)	0	0.050
Reoperation	3	1	

mitral regurgitation (MR) at initial coronary reconstruction. There was no significant difference in MR between the groups. The duration of mechanical ventilation in the infant group was significantly longer than that in the noninfant group ($p<0.05$). Three patients (6.1%) needed extracorporeal membrane oxygenation (ECMO), and four (8.2%) infant patients without ECMO underwent delayed sternal closure.

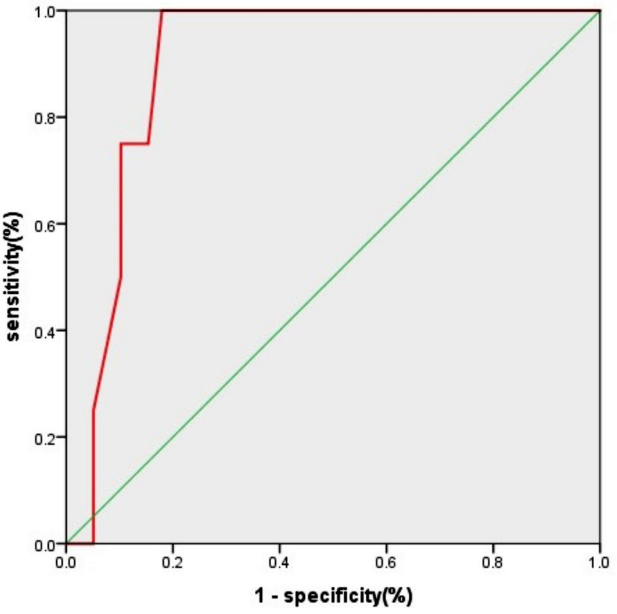

Fig. 1 ROC curve analysis demonstrating the predictive power of the preoperative ejection fraction. The area under the ROC curve and the cutoff values for EF were 0.901 and 39%, respectively

Mortality

Hospital mortality of this study was 8.2% (4 of 49) (Table 2). They were in the infant group. Four patients had a low LVEF < 35% and symptoms of heart failure. Three patients underwent LCA reimplantation, and one patient underwent Takeuchi repair. Two patients died after postoperative ECMO support, including one patient who received autologous pericardium to increase the length of the LCA and another one who underwent LCA reimplantation and resection of the ventricular aneurysm. The other two patients, both with abnormal deep and wide Q waves, died within 1 day after surgery due to sudden cardiac arrest.

Early mortality was associated with left ventricular dysfunction. ROC curve analysis (Fig. 1) revealed that the area under the ROC curve and the cutoff values for EF were 0.901 and 39%, respectively.

Table 2 Characteristics of dead patients

Patients	Age (months)	MI (preoperation)	Other malformation	LVEF% (preoperation)	Surgery type	ECMO
1	2	moderate		34	Reimplantation autologous pericardium tube	yes
2	6	moderate	Abnormal Q waves	35	Takeuchi repair	no
3	9	moderate	Abnormal Q waves	33	reimplantation	no
4	12	moderate	Ventricular aneurysm	37	reimplantation	yes

Follow-up

The median follow-up duration was 43 (18–85) months. There was no late mortality in either group. On the follow-up echocardiogram, there was a satisfactory EF for left ventricular function in both groups. Compared with the noninfant group, the infant group had a significantly greater EF at follow-up ($p<0.05$). Four patients underwent reoperation during the follow-up period. The estimated rates of freedom from reoperation after surgery at 1, 3, and 5 years were 97.4%, 94.2%, and 94.2%, respectively (Fig. 2). In the infant group, one patient who underwent the Takeuchi procedure underwent reoperation due to baffle leakage resulting in a residual left to right shunt in the pulmonary artery, and another patient experienced main pulmonary artery stenosis 1.5 years after the operation. Additionally, two patients with severe MR underwent reoperation.

Discussion

Anomalous origin of the left main coronary artery from the pulmonary artery is a rare congenital heart disease. Most patients develop signs and symptoms in the first few months of life, and some of them die of myocardial ischemia and infarction after the LCA flow decreases. For infants, surgical treatment is indicated at the time of diagnosis, but in the non-infant group, it is controversial. Left heart function and mitral regurgitation are major risk factors of postoperative mortality. For infants, left heart function and mitral regurgitation tend to be severe while operation, however, postoperative recovery is unclear compared with that in the non-infants. Surgical repair is currently the mainstay of treatment for this disease. However, this procedure is still challenging and carries a significant risk of early mortality and reoperation at follow-up. Our study suggested that surgical treatment had excellent early and midterm outcomes.

Several surgical techniques have been used to treat diseases, including direct implantation, Takeuchi repair, coronary artery bypass grafting (CABG) and pericardial tube interposition. The use of Takeuchi repair is associated with a high rate of complications in infants [8, 9]. In our study, two patients underwent Takeuchi repair. However, one patient died in the hospital, and one underwent reoperation. Currently, coronary artery transposition is a widely used surgical technique. When there is unfavorable coronary anatomy or unforeseen coronary tension, coronary elongation with a pericardial tube is feasible [4, 10]. Two patients required postoperative ECMO in this study. This was related to the immaturity of early coronary artery transplantation techniques.

Early mortality was associated with left ventricular dysfunction. Retrograde flow from the left coronary artery to the pulmonary artery after pulmonary vascular resistance decreases can result in complications, including myocardial ischemia, mitral regurgitation, resultant ventricular dysfunction or death [11, 12]. Although improvements in surgical techniques and medical therapy have reduced the impact on left ventricular function, many studies have shown that preoperative LV dysfunction is an independent predictor of early mortality [4, 13]. Our study also confirmed EF to be a risk factor for mortality. This study showed that the preoperative EF in the infant group was lower than that in the noninfant group, but there was no significant difference in the EF at discharge

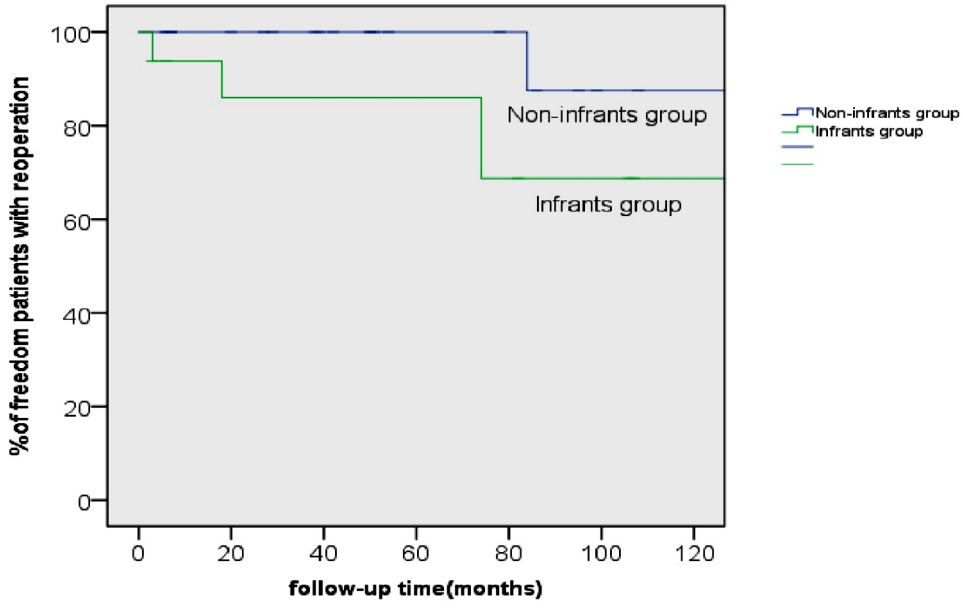

Fig. 2 Freedom from reoperation. The estimated rates of freedom from reoperation after surgery at 1, 3, and 5 years were 97.4%, 94.2%, and 94.2%, respectively

between the two groups. Early reconstruction of the dual coronary system and restoration of coronary blood can promote recovery of left ventricular function. Muna Ismail's [14] study showed that LV systolic function and dimensions could improve after operation. However, the time of operation depends on the time of diagnosis, yet some patients are not diagnosed until they present with symptoms of heart failure caused by myocardial ischemia. It also increased the risk of complications and mortality. It is important to promote earlier diagnosis and intervention in infants and prevent further deterioration of cardiac function.

MR is a common complication of papillary muscle damage caused by myocardial ischemia. However, whether to surgically intervene in the mitral valve during the initial surgical repair of ALCAPA remains controversial. Some studies [15, 16] have suggested that an increase in cross-clamp time due to additional MR repair could increase complications. Our study showed that patients with MR who underwent concurrent surgical repair at the time of initial coronary reconstruction did not show that MR was associated with an increased risk of in-hospital mortality. Persistent MR can cause congestive heart failure and decrease in-hospital survival, especially moderate or more severe MR. Therefore, concomitant repair is safe for this approach. Amanda S's^2 study showed that the degree of MR at the time of ALCAPA surgery did not correlate with early or late survival, and concomitant MV surgery improved in-hospital survival.

Reoperation is not a common complication after surgical repair. MR and pulmonary artery stenosis were the main reasons for reoperation. Some studies [5, 17] have shown that reoperation of the mitral valve is associated with moderate or severe MR at discharge, and the size of the pericardial patch, which is prone to shrinkage, may lead to pulmonary stenosis (PS). The reasons for reoperation in our study included MR, PS and residual left-to-right shunt after Takeuchi. The use of the Takeuchi technique carries a greater risk of reoperation than other surgical methods. Other studies [9, 18] have suggested that significant PS is more common in Takeuchi repair. Our study suggested that freedom from reoperation is not significantly different between infants and noninfants at 10 years. The specific risk factors might not influence the decision to operate age.

The main limitations of this study were its retrospective and single-institution nature. There was inadequate systematic midterm follow-up and assessment of the mitral valve. There was limited information on the description of MV pathology and preoperative LVEDD (Z score).

Conclusions

ALCAPA is a serious condition, but its early and midterm outcomes are excellent. Early reconstruction of the dual coronary system and restoration of coronary blood yield satisfactory results, with improvements in left ventricular function in survivors. Lower left ventricular dysfunction of pre-operation was the major risk of mortality in-hospital. Concomitant mitral valve repair did not increase perioperative risk, but mitral regurgitation needs to be observed during follow-up, although the incidence of reoperation is not high. The rate of freedom from reoperation did not differ significantly between the infant group and the noninfant group.

Author contributions

Cao wrote the main manuscript text, and made substantial contributions to the conception, design of the work and interpretation of data. Wang had substantively revised this paper.

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Data availability

No datasets were generated or analysed during the current study.

Declarations

Consent to participate

Every person agreed to participate in the study.

Competing interests

The authors declare no competing interests.

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