Preoperative Evaluation and Midterm Outcomes after the Surgical Correction of Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery in 50 Infants and Children

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Abstract

Background: Anomalous origin of the left coronary artery (LCA) from the pulmonary artery (ALCAPA) is a rather rare congenital anomaly that has a profound effect on heart function. This study aimed to retrospectively illustrate the perioperative clinical features, therapy experience, and midterm outcomes after surgical correction, and to determine the value of left ventricular ejection fraction (LVEF) and myocardial viability in differentiating critically ill patients among infants and children with ALCAPA.

Methods: From April 1999 to March 2013, infants and children patients diagnosed with ALCAPA in Beijing Fuwai Hospital were analyzed. Clinical data of patients were summarized and retrospectively analyzed. All patients were divided into two groups according to LVEF level (Group 1: LVEF >50%, or Group 2: LVEF ≤50%) to compare perioperative and follow-up variables. Effect of myocardial viability evaluated according to myocardial perfusion/^{18}F-fluorodeoxyglucose (FDG) imaging on the clinical variables was also analyzed.

Results: A total of 50 patients with ALCAPA (male/female: 29/21; median age: 3.1 years [range: 4 months to 18 years]) were included. Younger age, lower weight, intercoronary collaterals (ICC) dysplasia, ratio of the proximal right coronary artery diameter to the aortic root diameter <0.2, and larger cardiothoracic ratio (CTR) were more frequently found in Group 2 than those in Group 1. Forty-seven patients underwent cardiac surgery. The times of cross-clamp and cardiopulmonary bypass were not different between the two groups; however, the duration of mechanical ventilation and postoperative Intensive Care Unit stay were longer in Group 2 than those in Group 1. Follow-ups were possible in 38 patients (80.9%); median time: 84.5 months (range: 49 months to 216 months). There was one late sudden death with simple ligation of the LCA at 8 months after surgery. No severe complications and reoperation occurred. The relationship of the grades of myocardial viability and clinical features was analyzed in 15 patients with myocardial perfusion/^{18}F-FDG imaging, and the results showed that myocardial viability correlated well with LVEF, CTR, abnormal Q waves, and left ventricular end-diastolic dimension. It was not correlated with age, mitral regurgitation, and ICC. Heart implantation was decided in one patient with little viable myocardium; however, this patient died 2 months after the diagnosis while waiting for transplantation. Two patients with no viable myocardium in the area of aneurysm had aneurysmectomy concomitantly.

Conclusions: In infants and children with ALCAPA, heart function and myocardial viability are closely related to clinical features. LVEF and the grades of myocardial viability can differentiate high-risk patients before surgery and in the early stage of recovery after surgery. The area and extent of myocardial infarction are also crucial in making preoperative clinical decisions. However, even in patients with depressed ventricular function and severe myocardial infarction, the midterm follow-up showed satisfactory recovery of cardiac function after the successful restoration of a dual-coronary arterial system.

Key words: Follow-up Studies; Left Coronary Artery (LCA) from the Pulmonary Artery; Myocardial Ischemia; Surgical Procedures

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INTRODUCTION

As a rather rare congenital anomaly, anomalous origin of the left coronary artery (LCA) from the pulmonary

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artery (ALCAPA) occurs in approximately 1 per 300,000 live births and represents 0.5% of all congenital heart disease cases.[1] The presentation and onset of symptoms typically occur shortly after the neonatal period as the pulmonary vascular resistance decreases and left coronary blood flow diminishes. Decreased coronary blood flow results in myocardial ischemia, tissue infarction, mitral insufficiency, and dilated ischemic cardiomyopathy. If left uncorrected, the mortality rate of infants reaches 90%. Extensive collateral arteries may enable some patients to survive beyond infancy. However, chronic hypoperfusion causes subendocardial ischemia and later fibrosis, increasing the risk of sudden death secondary to ventricular arrhythmias.

In infants and children with ALCAPA, a false initial diagnosis at different ages occurs frequently owing to the lack of specificity in the clinical manifestations, which range from asymptomatic to severe myocardial ischemia, heart failure, and sudden death.[2] The area and extent of myocardial infarction may decide the clinical manifestations and outcome. It has been proposed that myocardial viability evaluated by means of myocardial perfusion/^{18}F-fluorodeoxyglucose (FDG) imaging may be related to preoperative clinical manifestations and cardiac function.[3,4]

Multiple techniques have been introduced to establish a dual-coronary artery system, including coronary artery bypass grafting (CABG), coronary baffling procedures, and direct reimplantation of the anomalous coronary artery into the aorta (DIACA).

This study aimed to retrospectively illustrate the perioperative clinical features, therapy experience, and midterm outcomes after surgical correction, and to determine the value of left ventricular ejection fraction (LVEF) and myocardial viability in differentiating critically ill patients among infants and children with ALCAPA.

Methods
Ethical approval
The study was approved by the Ethical Committee of Fuwai Hospital. The patients or guardians of patients signed informed consent for the present study.

Patients and follow-up
A retrospective review of the medical charts of 50 infants and children with ALCAPA was performed at Beijing Fuwai Hospital, a large cardiac center in China, between April 1999 and March 2013. Data including patient demographics, preoperative clinical data, operative variables, early or late complications, and reoperations were collected. Clinical assessment at the most recent follow-up was conducted using electronic medical records, archived paper charts, and telephone calls.

The collected demographic variables included sex, age, and weight at surgery. The analyzed preoperative clinical variables included those from electrocardiography (ECG),

echocardiography, chest radiography, multidetector computed tomography (MDCT), angiocardiography, and myocardial perfusion/^{18}F-FDG imaging. The analyzed operative variables included the type of ALCAPA repair, mitral valve intervention, additional surgical procedures performed, cross-clamp time, cardiopulmonary bypass (CPB) time, duration of mechanical ventilation, duration of Intensive Care Unit (ICU) stay, postoperative complications, and discharge status. Delayed recovery was defined as a mechanical ventilation time of >3 days and postoperative ICU stay of >7 days. Ventricular function was assessed according to standard echocardiographic methods with the following parameters: LVEF, left ventricular end-diastolic dimension (LVED), and mitral regurgitation (MR). The MR severity was expressed as mild, moderate, or severe to categorize the regurgitation areas on the color Doppler images.

After discharge, patients were routinely followed up in the outpatient department and through telephone calls. Information about symptoms, ECG, and radiographic changes was acquired. LVEF, LVED, MR, supravalvular pulmonary stenosis (PS), and patency of the neoproximal LCA were evaluated through echocardiography. The composite endpoint was defined as death, severe complications such as severe supravalvular PS and deterioration of heart failure, or reoperation.

Patients were divided into two groups based on preoperative LVEF (Group 1: >50%; Group 2: ≤50%). The perioperative clinical variables and follow-up data were compared between the two groups.

Electrocardiography
Abnormal Q wave was defined when its duration was ≥0.03 s, its depth exceeded one-fourth of the height of the R wave in the same lead, or its depth was ≥3 mm in lead I or ≥2 mm in lead aVL (augmented voltage, left arm). An ST segment depression was defined as a depression of ≥0.05 mV.

Echocardiography
Routine two-dimensional echocardiography and Doppler flow imaging were performed to visualize the origin, course, and flow direction of the LCA and right coronary artery (RCA), as well as abnormal shunting into the pulmonary artery. LVEF, LVED, valvular function, ventricular wall motion, endocardial thickness, and the diameters of the RCA and the aortic root were recorded.

Multidetector computed tomography
Owing to its noninvasiveness and high spatial resolution, MDCT was used to show the origin of the LCA and RCA in most patients.[5]

Catheterization and angiography
Data were obtained during right heart catheterization. Retrograde aortic root angiography via the femoral artery was performed to detect the origin, course, and flow direction of each coronary artery, intercoronary collaterals (ICC), and abnormal shunting.

Evaluation of myocardial viability through myocardial perfusion/^{18}F-fluorodeoxyglucose imaging

On the basis of the standard 17-segment model, both perfusion and ^{18}F-FDG images were visually evaluated by two nuclear physicians blinded to the clinical data. Hypoperfused segments were scored on a four-point scale, as follows: 0, normal (normal perfusion and normal ^{18}F-FDG uptake); 1, mismatch (abnormal perfusion but normal ^{18}F-FDG uptake); 2, partial mismatch (abnormal perfusion and abnormal ^{18}F-FDG uptake, but ^{18}F-FDG uptake greater than the perfusion uptake); and 3, match (concordant degree of perfusion and ^{18}F-FDG defects).[3,4]

Cardiac surgery

The major surgical methods included DIACA, Takeuchi operation, and ligation of the LCA at its origin with or without CABG.

Statistical analysis

Continuous variables were reported as median (range), or mean ± standard deviations (SD). Independent continuous variables were compared using unpaired Student's test for normally distributed data, and Mann-Whitney U-test was used for the comparison of parameters that did not exhibit a normal distribution. Paired-samples t-test was used for the comparison of variables before and after surgery. Categorical variables were reported as frequencies with percentages, and the Chi-square test or Fisher's exact tests were used. Relationship between rank variables was calculated using Spearman rank correlation. A two-tailed $P < 0.05$ was considered statistically significant. All statistical analyses were performed with SPSS software, version 19.0 (IBM Inc., Somers, NY, USA).

RESULTS

A total of 50 patients with ALCAPA (male/female: 29/21; median age: 3.1 years [range: 4 months to 18 years]) were included in this study. Thirty-nine patients had concomitant MR, two had left ventricular aneurysm, one had Tetralogy of Fallot (TOF)/atrial septal defect (ASD), one had severe PS/partial anomalous pulmonary venous connection (PAPVC), and one had ASD. Forty-seven patients underwent cardiac surgery, and their definitive diagnoses were confirmed with surgery. In three patients without surgery, the definitive diagnosis was established with angiography, and/or MDCT scan. All patients had MDCT and/or angiocardiography (28 MDCT, 19 angiocardiography, and 3 MDCT + angiocardiography) and 15 patients had myocardial perfusion/^{18}F-FDG imaging preoperatively.

Perioperative clinical variables between two groups with different left ventricular ejection fraction

Perioperative clinical variables and outcomes were compared between the two groups (Group 1: LVEF >50%, $n = 34$; Group 2: LVEF ≤50%, $n = 16$) and shown in Table 1. Younger age, lower weight, ICC dysplasia, ratio of the proximal RCA

Table 1: Comparison of perioperative clinical features between two groups with different LVEF level in 50 infants and children with ALCAPA

Clinical data	LVEF		χ^2 or t or Z	P
	>50% ($n = 34$)	≤50% ($n = 16$)		
Age (year), median (range)	4.9 (0.5-16.0)	0.9 (0.3-5.2)	−3.80*	<0.001
Weight (kg), mean ± SD	20.0 ± 13.0	8.8 ± 3.1	3.04†	0.004
Abnormal Q-waves, n (%)	26 (76.5)	14 (87.5)	0.87	0.363
Preoperative LVED (mm), mean ± SD	43.0 ± 6.6	46.9 ± 6.9	1.91†	0.055
Preoperative LVEF (%), mean ± SD	64.6 ± 7.3	29.1 ± 6.4	16.34†	<0.001
RCA/AO <0.2, n (%)	2 (7.1)	8 (53.3)	11.68	<0.001
ICC dysplasia, n (%)	2 (6.7)	7 (43.8)	9.12	0.005
Preoperative MR (moderate or severe), n (%)	19 (55.9)	9 (56.3)	0.001	0.950
Preoperative CTR	0.59 ± 0.06	0.68 ± 0.04	5.42†	<0.001
Operation‡				
DIACA, n (%)	22 (66.7)	13 (92.9)	3.55	0.050
Takeuchi, n (%)	9 (27.3)	0		
Other procedures§, n (%)	2 (6.1)	1 (7.1)		
Cross-clamp time (min), mean ± SD	82.4 ± 22.3	82.2 ± 24.3	0.02†	0.982
CPB time (min), mean ± SD	120.1 ± 31.5	134.6 ± 51.5	1.10†	0.277
Postoperative ventilation time (h), median (range)	14.5 (3.0-336.0)	49.5 (10.0-504.0)	−3.47*	0.001
Postoperative ICU stay (h), median (range)	48 (15-624)	96 (45-720)	−2.90*	0.004
LVED 1-week postoperation (mm), mean ± SD	34.2 ± 7.2	36.9 ± 6.4	1.20†	0.294
LVEF 1-week postoperation (%), mean ± SD	63.7 ± 8.6	32.1 ± 8.6	11.25†	<0.001

*Z; †t; ‡47-operation, 2-quit, 1-waiting for heart transplantation. §1-LCA ligation, 1-LCA ligation + CABG + MVR, 1-CABG + MVP + aneurysmectomy - at 18 months after LCA ligation. SD: Standard deviation; LVED: Left ventricular end-diastolic dimension; LVEF: Left ventricular ejection fraction; ICC: Intercoronary collaterals; RCA/AO: Ratio of the proximal right coronary artery diameter to the aortic root diameter; MR: Mitral regurgitation; CTR: Cardiothoracic ratio; DIACA: Direct reimplantation of the anomalous coronary artery into the aorta; Takeuchi: Intrapulmonary baffle reconstruction, which is also known as the "Takeuchi procedure"; LCA: Left coronary artery; CABG: Coronary artery bypass grafting; MVR: Mitral valve replacement; MVP: Mitral valve plasty; CPB: Cardiopulmonary bypass; ALCAPA: Anomalous left coronary artery from the pulmonary artery.

diameter to the aortic root diameter (RCA/AO) <0.2, and larger cardiothoracic ratio (CTR) were more frequently found in Group 2 than in Group 1. One infant with profoundly depressed ventricular function and very little viable myocardium died 2 months after the diagnosis while waiting for heart transplantation. Forty-seven patients underwent cardiac surgery including one case of LCA ligation (2.1%), two cases of LCA ligation plus CABG (4.3%), nine cases of Takeuchi operation (19.1%), and thirty-five cases of DIACA (74.5%). Meanwhile, 20 patients had concomitant mitral valve repair, 1 patient had concomitant mitral valve replacement, 2 patients had concomitant aneurysm resection, and 3 patients had concomitant repairs of TOF/ASD, PS/PAPVC, and ASD, respectively. After surgery, there were no early surgical deaths.

More patients underwent DIACA in Group 2; however, no significant difference was found between the two groups. The times of cross-clamp and CPB were not significantly different between the two groups; however, the duration of mechanical ventilation and postoperative ICU stay were longer in Group 2 than in Group 1. Except for one patient, six of the seven patients with delayed recovery were under 1 year old. Among them, five patients had LVEF ≤50%, two had aneurysm, and one underwent extracorporeal membrane oxygenation (ECMO) after surgery. Echocardiographic evaluation revealed a significant improvement in terms of LVED after surgery (preoperative vs. 1 week after surgery: 43.0 ± 6.6 mm vs. 34.2 ± 7.2 mm in Group 1, and 46.9 ± 6.9 mm vs. 36.9 ± 6.4 mm in Group 2, $P < 0.001$, respectively), whereas LVEF recovered at a slower rate (preoperative vs. 1 week after surgery: $64.6 \pm 7.3\%$ vs. $63.7 \pm 8.6\%$ in Group 1, and $29.1 \pm 6.4\%$ vs. $32.1 \pm 8.6\%$ in Group 2, $P > 0.05$, respectively).

Twenty-one patients with moderate or severe MR preoperatively (9 severe MR and 12 moderate MR) underwent concomitant mitral valve intervention, and mild MR was found in 19 patients at midterm follow-up (12 patients had previous mitral valve intervention). As shown in Table 1, the severity and incidence of MR were not significantly different between the two groups before and after surgery.

Follow-ups were possible in 38 patients (80.9%, 38/47) and the median length of follow-up after surgery was 84.5 months (range: 49–216 months). The rate of loss to follow-up was 19.1%. There was one late sudden death with simple ligation of the LCA at 8 months after surgery. No severe complications and reoperation occurred. Nineteen patients had mild MR and three had mild supravalvular PS at midterm follow-up. All patients with preoperative LVEF ≤50% reached normal values at 0.5–15 months after surgery. At the most recent follow-up, all patients were asymptomatic and had New York Heart Association (NYHA) Class I status.

Preoperative evaluation of the relationship of myocardial viability and clinical variables in 15 patients with myocardial perfusion/18F-fluorodeoxyglucose imaging

Myocardial perfusion/18F-FDG imaging remains the gold standard for the diagnosis of the extent of myocardial viability. The clinical features and the grades of myocardial viability in 15 patients with myocardial perfusion/18F-FDG imaging are shown in Table 2. Except for 1 patient (Grade 0), the other 14 patients had decreased myocardial perfusion that involved the anterior, lateral, and apical walls, which were the blood supplementation areas of the LCA. Among them, 4 patients had viable myocardium (Grade 1) and 10 had different severities of myocardial infarction (Grade 2–3), including two with ventricular aneurysm who showed no viable myocardium in the area of aneurysm.

The relationship of the grades of myocardial viability and clinical features was analyzed in the 15 patients with myocardial perfusion/18F-FDG imaging, and the results showed that myocardial viability correlated well with LVEF ($r = -0.783$, $P < 0.001$), CTR ($r = 0.770$, $P < 0.002$), abnormal Q waves ($r = 0.667$, $P < 0.01$), and LVED ($r = 0.637$, $P < 0.02$). It was not correlated with age, MR, and ICC ($r = -0.206$, -0.268, -0.342, respectively; $P > 0.05$).

As shown in Table 2, most patients with myocardial infarction (Grade 2–3 in myocardial perfusion/18F-FDG imaging) had abnormal Q waves (9/10), depressed ventricular function (LVEF ≤50%, 9/10), ICC dysplasia (6/10), and CTR >0.65 (6/10). In one infant (case 15) with extensive myocardial infarction and very little viable myocardium, heart transplantation was decided rather than other surgical procedures after a preoperative discussion; however, this patient died 2 months after the diagnosis before heart transplantation. The angiocardiography and echocardiography of this patient showed that the left ventricular wall motion almost disappeared, the left ventricular end-diastolic pressure remarkably increased, and the ventricular function profoundly depressed. Of three patients (cases 11, 13, and 14) with apical ventricular aneurysm, two patients (cases 11 and 14) with no viable myocardium in the area of aneurysm had concomitant aneurysmectomy and one patient (case 13) with some viable myocardium in the area of aneurysm recovered well without aneurysmectomy.

Of seven patients with severe myocardial infarction (Grade 3 in myocardial perfusion/18F-FDG imaging in at least one segment of the standard 17-segment model), six had LVEF <30%, one died preoperatively, two had an aneurysm, one underwent ECMO after surgery, and three had delayed recovery. Among them, no late death, severe complications, and reoperation occurred.

DISCUSSION

An increasing number of ALCAPA cases has been reported recently; however, the sample size of each study has been relatively small.[6,7] The origin of the LCA from the pulmonary artery is well tolerated in fetal and early neonatal periods because the pulmonary arterial pressure is the same as the systemic pressure, which leads to antegrade flow in both the anomalous LCA and the normal RCA. Soon after

Table 2: Clinical features and grades of myocardial viability of 15 patients with myocardial perfusion/^{18}F-FDG imaging

Case	Age (years)	Myocardial imaging-scores	Abnormal Q-waves	LVEF (%)	LVED (mm)	MR	ICC	CTR	Operation and follow-up
1	1.7	Normal myocardial perfusion and glucose metabolism	-	68	39	+-++	+	0.63	DIACA + MVP, no MR
2	6.7	Anterior-1	-	61	47	+	+	0.56	DIACA
3	0.5	Anterior, lateral-1	-	74	38	+++	+	0.64	DIACA + MVP, mild MR
4	1.8	Anterior, lateral-1	+ (I aVL)	61	41	+++	+	0.64	DIACA + MVP, no MR
5	6.7	Anterior, apical-1	+ (I aVL)	65	48	+++	+	0.63	Takeuchi + MVP, no MR
6	3.4	Apical, antero-apical-2, antero-mid-1	+ (I aVL V_5)	65	42	+	+	0.60	DIACA
7	1.6	Antero, anterolateral-2	+ (IaVL)	28*	42	++	+	0.61	DIACA + MVP, LVEF >50% 12 months after surgery, mild MR
8	4.0	Lateral-2	+ (aVL)	35*	47	++	+	0.63	DIACA + MVP, LVEF >50% 2 months after surgery, no MR
9	0.3	Antero-mid + basal, lateral-basal-3, apical, antero-apical, anterolateral-apical + mid-1	+ (I aVL V_3–V_5)	26*	45	-	-	0.68	DIACA, LVEF >50% 4 months after surgery
10	2.8	Antero-3	+ (I aVL)	23*	63	++	-	0.77	DIACA + MVP, LVEF >50% 6 months after surgery, no MR
11	2.8	Apical, anterior, anterolateral-3	+ (I aVL)	40*	42	+++	-	0.68	CABG + MVP + aneurysmectomy, delayed recovery, LVEF >50% 0.5 month after surgery, mild MR
12	0.3	Antero-apical-3, apical, antero-basal + mid-1, lateral-1	-	25*	52	++	-	0.71	DIACA, delayed recovery, ECMO for 7 days, LVEF >50% 13 months after surgery, mild MR
13	5.2	Apical, antero-apical-3, antero-mid + basal, lateral, inferior-1	+ (I aVL)	24*	61	+	-	0.66	Mild apical paradoxical movement, DIACA, LVEF >50% 13 months after surgery
14	0.6	Apical-3, anteroseptal-3, antero-mid + apical-2, lateral-1	+ (I aVL V3–V_5)	23*	50	+-++	+	0.64	DIACA + MVP + aneurysmectomy, delayed recovery, LVEF >50% 3 months after surgery, no MR
15	0.7	Apical-3, anterior-3, lateral, inferior-2, septal-1	+ (I aVL V_5)	25*	52	-	-	0.71	Died preoperatively

*LVEF <50%. LVED: Left ventricular end-diastolic dimension; LVEF: Left ventricular ejection fraction; MR: Mitral regurgitation (+: Mild; ++: Moderate; +++: Severe); ICC: Intercoronary collaterals (+: Well-developed ICC; -: ICC dysplasia); CTR: Cardiothoracic ratio; ECG: Electrocardiography; LV: Left ventricular; RV: Right ventricular; ECMO: Extracorporeal membrane oxygenation; DIACA: Direct reimplantation of the anomalous coronary artery into the aorta; Takeuchi: Intrapulmonary baffle reconstruction, which is also known as the "Takeuchi procedure"; CABG: Coronary artery bypass grafting; ^{18}F-FDG: ^{18}F-fluorodeoxyglucose; aVL: Augmented voltage, left arm.

birth, when the pulmonary arterial pressure decreases, the flow in the LCA decreases and then reverses, which leads to myocardial ischemia and infarction.[8]

As usually described, the electrocardiogram of patients with ALCAPA presents the pattern of left anterolateral myocardial infarction, typified, among other findings, by abnormal Q waves in leads V_4–V_6, I, and aVL. However, unfortunately, these typical changes do not always occur in ALCAPA.[9,10] Similar changes may occur in acute myocarditis or dilated cardiomyopathy, which decrease the value for predicting myocardial infarction. Although the reasons for the absence of abnormal Q waves are not fully understood, they are associated with subendocardial rather than transmural infarction, inferior rather than anterior infarcts, and smaller rather than larger infarcts in adults.[10,11]

Between the two groups based on LVEF, there were statistically significant differences in many preoperative clinical variables including age of onset, CTR, and ICC formation; however, the severity and incidence of MR were not significantly different between the two groups.

Several surgical methods have been developed for the treatment of ALCAPA, and techniques for establishing a dual-coronary system have become the standard approach for repair. These techniques include DIACA, CABG, and the Takeuchi procedure. In our series, there were more patients

undergoing DIACA in Group 2; however, no significant difference was found between the two groups. The Takeuchi procedure was usually conducted before 2006 and in older children. DIACA has been recommended in recent years.[12]

The main procedures of Takeuchi include performance of a pulmonary arteriotomy and creation of a transverse flap of pulmonary artery tissue and an aortopulmonary window. The pulmonary artery flap was used to baffle the LCA into the aorta. The pulmonary artery was then reconstructed with autologous pericardium. Patients with Takeuchi repair have a higher incidence of PS and baffle leaks on long-term follow-up.[13] In our series, nine patients had Takeuchi procedures and only three had mild supravalvular PS at midterm follow-up.

LCA ligation has been described as a therapeutic option with the aim of interrupting the steal of coronary flow from the pulmonary artery and increasing the perfusion pressure in the coronary territory, and although this leads to an improvement in the ventricular function and to a decrease in MR, the risk for sudden death in the midterm and short-term does not decrease.[14] In our study, a 10-year-old child with normal cardiac function preoperatively, who underwent LCA ligation without CABG, died suddenly at 8 months after surgery. Another patient underwent LCA ligation without CABG in other hospital. This patient had low cardiac function and left ventricular aneurysm 1.5 years after the first surgery, and underwent secondary procedures consisting of CABG + MVP + aneurysmectomy at our hospital and recovered well.

Mitral valve intervention during the initial surgical repair of ALCAPA remains controversial. In general, mitral valve repair or replacement is not necessary at the time of ALCAPA repair; however, if MR remains persistent, and depending on the severity, it can be managed surgically at a later date.[15,16] In our series, 78% (39/50) patients had concomitant MR. Mitral valve intervention was performed in most patients (75%, 21/28) with moderate or severe MR. Only 19 patients had mild MR in a comparably longer follow-up of 4–18 years, and no patient required reintervention.

The times of cross-clamp and CPB were not different between the two groups; however, the postoperative ventilation time and postoperative ICU stay were significantly longer in Group 2 than in Group 1. The LVEF of one patient who received postoperative ECMO assistance was 25%. Patients with younger age and/or lower LVEF had higher risks in the early period after surgery. In patients undergoing ALCAPA repair, LV function recovery may vary according to the efficiency of the surgical and medical treatment, and may take several months, even up to 1 year, after repair.[17] However, we believe that the period between the intervention and recovery is difficult to determine and depends on multiple factors, including the preoperative status of the patient and the intraoperative approaches used. In our study, LVED rapidly decreased, whereas LVEF recovered at a slower rate after a successful repair. All 16 patients with LVEF ≤50% preoperatively reached normal values at 0.5–15 months after surgery. At the most recent follow-up, all patients were asymptomatic and had NYHA Class I status.

Evaluating the area and severity of myocardial infarction before coronary revascularization is critical in adults with coronary artery disease, but is seldom reported in children.[18] Myocardial perfusion/^{18}F-FDG imaging remains the gold standard for the diagnosis of the extent of myocardial viability.[19] Consistent with previous reports, myocardial viability correlated well with some clinical features such as abnormal Q wave, LVED, LVEF, and CTR in our study. Children with less viable myocardium had more severe clinical symptoms. In infants and children with ALCAPA, especially with regional wall motion abnormalities and depressed ventricular function, evaluation of the area and severity of myocardial infarction preoperatively is also crucial for the choice of the operation method. Patients with severe myocardial infarction may experience delayed recovery in the early stage after surgery; however, the midterm follow-up showed satisfactory recovery of cardiac function after the successful restoration of a dual-coronary arterial system.

In infants and children with ALCAPA, heart function and myocardial viability are closely related to clinical features. LVEF and the grades of myocardial viability can differentiate high-risk patients before surgery and in the early stage of recovery after surgery. The area and extent of myocardial infarction are also crucial in making preoperative clinical decisions. However, even in patients with depressed ventricular function and severe myocardial infarction, the midterm follow-up showed satisfactory recovery of cardiac function after the successful restoration of a dual-coronary arterial system.

Conflicts of interest
There are no conflicts of interest.

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