Midterm Surgical Outcomes for ALCAPA Repair in Infants and Children

Renjie Hu[1] Wen Zhang[1] Xiafeng Yu[1] Hongbin Zhu[1] Haibo Zhang[1] Jinfen Liu[1]

[1] Department of Cardiothoracic Surgery, Shanghai Children's Medical Center Affiliated to Shanghai Jiaotong University School of Medicine, Shanghai, People's Republic of China

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Address for correspondence Jinfen Liu, MD, Department of Cardiothoracic Surgery, Shanghai Children's Medical Center Affiliated to Shanghai Jiaotong University School of Medicine, Shanghai, People's Republic of China (e-mail: liujinfen2002@126.com).

Abstract

Background Surgical correction of an anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) has been associated with excellent survival during recent years. The purpose of this study was to evaluate the effectiveness of reimplantation of the coronary artery and to investigate the recovery of postoperative cardiac and mitral valve (MV) function.

Methods From 2005 to 2015, 80 patients who had ALCAPA received surgical correction. Among them, 49 were infants. The median patient age was 7.8 months. Operative strategies included reimplantation of the coronary artery in 71 patients, the Takeuchi procedure in another 7 patients, and coronary artery ligation in the remaining 2 patients.

Results There were 11 hospital deaths and 2 late deaths. Six patients required intraoperative or postoperative mechanical circulatory support. A significant improvement in the ejection fraction (EF) and shortening fraction (SF) was present in all surviving patients at discharge, at a 3-month follow-up and at a 1-year follow-up. MV function improved gradually after surgical repair with no late secondary intervention.

Conclusions The repair of ALCAPA can be accomplished by establishment of a dual-coronary system, which offers an acceptable mortality rate and will rarely require a second surgery. Left ventricular (LV) recovery is a progressive process, especially for infants with impaired LV function. Concomitant MV annuloplasty is safe and reliable and can be performed as necessary in patients with moderate or severe mitral valve regurgitation.

Keywords

- coronary artery disease
- cardiac function
- circulatory support devices
- anomalous origin of left coronary artery from pulmonary artery
- bland–White–Garland

Introduction

Anomalous origin of the left coronary artery (LCA) from the pulmonary artery (ALCAPA) is a rare congenital heart defect that was described by Edward Bland, Paul Dudley White, and Joseph Garland in 1933, which was also named the Bland–White–Garland syndrome.[1] The pathophysiology of this malformation results in the development of collateral circulation from the right coronary artery to the ALCAPA, which subsequently elicits myocardial ischemia and left ventricular (LV) dysfunction. Therefore, the onset of symptoms varies in patients, depending on the extent of the defect and pace at which LV dysfunction progresses. Some children will be asymptomatic for a long time and will not present with LV dysfunction until later childhood. In addition to LV dysfunction, mitral valve regurgitation (MR) can also be found in most patients, which is secondary to LV dilatation and papillary muscle ischemia.

Surgical treatments, such as reimplantation of the ALCAPA to the aorta or creation of an intrapulmonary baffle,

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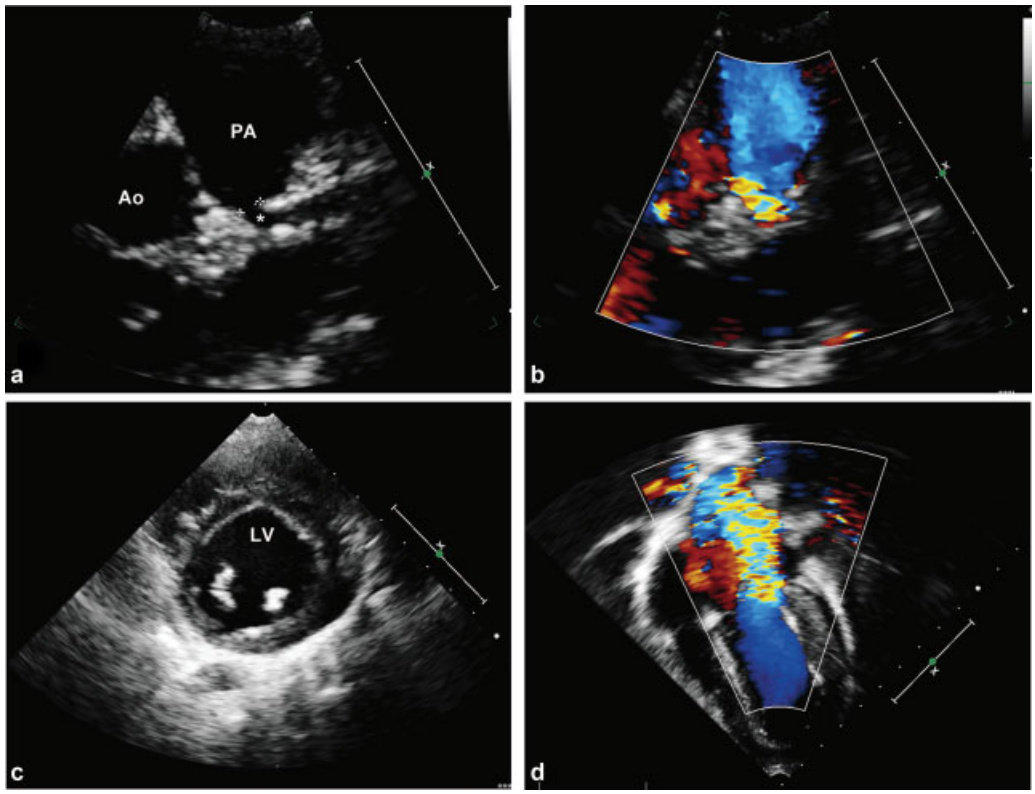

Fig. 1 Preoperative echocardiography examination of one patient. (**a**) The orifice of the left coronary artery from pulmonary artery. (**b**) Blood flow of the anomalous coronary artery. (**c**) Calcification of papillary muscle of mitral valve. (**d**) Severe mitral valve regurgitation (Ao, aorta; LV, left ventricular; PA, pulmonary artery).

have dramatically dropped the mortality to less than 20% during recent years.[2,3] Reestablishment of a dual-coronary system, as soon as possible, is an ideal procedure to preserve LV function. At the same time, several problems regarding mitral valve (MV) intervention at the time of reimplantation and the use of postoperative mechanical circulatory assistance remain unresolved.

The present study summarizes the results of our recent surgical outcomes for patients undergoing ALCAPA repair. Our goal was to evaluate the effectiveness of reimplantation of the coronary artery and to investigate the recovery of postoperative cardiac and MV function.

Patients and Methods

Patient Characteristics

With approval of the Ethics Committee of Medical Center, we retrospectively reviewed 80 patients with ALCAPA from 2005 to 2015. Among them, 49 were infants and the remaining 31 patients presented with ALCAPA after 1 year of age. Ten infants were younger than 6 months and 4 were younger than 3 months. Eight infants were admitted to our hospital as emergent patients with severe LV dysfunction and heart failure. Two of them required preoperative mechanical ventilation.

All patients were examined by two-dimensional and Doppler echocardiography to evaluate the severity of the LV dysfunction and the degree of MR (►**Fig. 1**). Thirty-three patients also underwent cardiac catheterization for a more precise diagnosis. LV function was based on the calculations of the ejection fraction (EF), shortening fraction (SF), and the left ventricular end diastolic diameter (LVEDD). LVEDDs were indexed to normal values for body surface areas to calculate the Z-score. These details, in addition to demographic data, are shown in ►**Table 1**.

Surgical Techniques

All patients underwent surgical correction through a median sternotomy. The procedure was performed with a distal aortic and bicaval venous cannulation under normothermia in 7 patients, mild hypothermia in 48 patients, and moderate hypothermia in 25 patients. Cardioplegia administration was performed via both the ascending aorta and the main pulmonary artery with cold crystalloid. The main pulmonary artery was incised at a high level and the orifice of the LCA was identified.

Different surgical corrections have been performed including reimplantation of the coronary artery in 71 patients, the Takeuchi procedure in 7 patients, and coronary artery ligation in 2 patients. Coronary artery ligation and the Takeuchi procedure were used early on, but have since been largely replaced. Reimplantation of the coronary artery into the aorta to create a dual-coronary system is now our standard procedure, which was developed using direct reimplantation with some modifications, such as using the aortic and pulmonary flaps reported by Sese and Imoto[4] or the pulmonary wall tubular extension techniques described by Turley et al.[5]

Table 1 Demographic data of patients

Characteristics	Number (80 patients)
Age, mo (median, range)	7.8 (1.4–141.2)
Weight, kg (median, range)	6.9 (4.0–51.0)
BSA (median, range)	0.3 (0.2–1.6)
Preoperative LVEF, % (mean ± SD), *N*	48.0 ± 15.3
Normal	27
Mildly impaired	24
Moderately impaired	20
Severely impaired	9
Preoperative LVFS, % (mean ± SD)	24.4 ± 9.8
Preoperative LVEDD Z-score (mean ± SD)	2.2 ± 0.9
Preoperative MR, *N*	
None	9
Mild	33
Moderate	27
Severe	11
Preoperative ECG characteristics, *N*	
Deep Q waves	35
ST-T changes	37
Preoperative inotrope support, *N*	39
LCA origin, *N*	
Sinus 1	50
Sinus 2	4
Nonfacing sinus	21
High takeoff from left/right pulmonary artery	5
Associated cardiovascular anomalies, *N*	
PDA	11
ASD	18
VSD	2
PAPVC	1

Abbreviations: ASD, atrial septal defect; BSA, body surface area; ECG, electrocardiogram; LCA, left coronary artery; LVEDD, left ventricular end diastolic diameter; LVEF, left ventricular ejection fraction; LVFS, left ventricular fraction of shortening; MR, mitral regurgitation; PAPVC, partial anomalous of pulmonary vein connection; PDA, patent ductus arteriosus; VSD, ventricular septal defect.

Direct reimplantation was performed in 18 patients. The orifice of the LCA was harvested with a generous cuff of the pulmonary sinus wall and mobilized upward to be directly reimplanted into the aortic wall. To obtain a tension-free anastomosis, a two-flap technique was performed. An aortic flap was also resected, and the coronary artery flap was anastomosed to the aortic flap to form a planned tunnel. If there was any suspicion of tension of the anastomosis, an autologous pericardial patch was employed for augmentation. This modified technique was performed in 37 patients. If the distance between the coronary artery and the aorta was too long for transfer, a tubular extension technique by suturing together the edges of the large LCA cuff was used to lengthen the coronary artery, so that it would reach the aorta. This procedure was performed in 16 patients. After the LCA was reimplanted, the pulmonary artery was reconstructed with autologous pericardium.

MV structural abnormalities were detected in four patients by intraoperative inspection, including a double orifice MV in one patient, and an MV cleft in three patients. All these patients underwent concomitant MV valvuloplasty. An ATS mechanical MV of 29 mm (ATS Medical, Inc., Minneapolis, Minnesota, United States) was implanted in a 9-year-old patient for severe calcification and infarction of the papillary muscles, which we were unable to repair. Twenty-four patients also received MV annuloplasty at the commissure along the posterior leaflet to treat MV regurgitation concomitantly. Additionally, patent ductus arteriosus (PDA) ligation and septal defect closure were performed in patients with associated anomalies.

Statistical Analysis

Data were analyzed with SPSS software, version 20.0 (SPSS Inc., Chicago, Illinois, United States). Continuous data were expressed as a median value for non-normally distributed variables and as the mean ± standard deviation (SD) for normally distributed variables. Group comparisons were assessed with a Student *t*-test for continuous variables that were normally distributed and with a Mann–Whitney *U* test for skewed variables. Fisher's exact test was applied for categorical data. Rates for survival were determined as time-related events using a Kaplan–Meier curve. Logistic regression analysis was used to evaluate several variables as potential risk factors for death and left ventricular assistant device (LVAD) implantation. A receiver operating characteristic (ROC) curve was established to evaluate the reliability of preoperative EF and LVEDD Z-scores as sensitive predictors of death. A p-value < 0.05 was considered statistically significant.

Results

All patients were successfully weaned from cardiopulmonary bypass (CPB), except two who required mechanical circulatory support. Median CPB and aortic cross-clamp time were 113 (range: 44–271) and 69 (range: 22–-165) minutes, respectively. Delayed sternal closure became necessary in 16 patients. The median duration of the intensive care unit (ICU) stay and of mechanical ventilation was 8 (range: 2–45) days and 94 (range: 4–336) hours, respectively.

Mortality

There were 11 hospital deaths and 2 late deaths. One patient died on the second day after LVAD implantation as a result of multi-organ failure. Five patients died of sudden cardiac arrest secondary to ventricular fibrillation. Five patients died in the early period of the reported experience because

of severe LV dysfunction and it was not possible to support them with LVAD. Six patients received a two-flap technique and three patients underwent direct reimplantation and the other two patients underwent the Takeuchi procedure.

The median follow-up time was 32 (range: 5–126) months. Late deaths occurred in two patients, one at 4 months and one at 1 year after the initial surgery. The first patient was readmitted to the hospital for severe LV dysfunction and heart failure. Postoperative transthoracic echocardiography (TTE) indicated that EF was less than 25% and that the left ventricle was markedly enlarged. Unfortunately, this patient failed to survive, even with inotropic support. The other patient suffered a sudden death at home during the follow-up period. The last TTE showed greater than moderate MR in this patient. A combination of diuretic and angiotensin-converting enzyme inhibitors was recommended. Death is likely attributable to prolonged congestive heart failure.

The Kaplan–Meier survival rate was estimated at 85.0% for 6 months and 83.5% for 1 year postsurgery (►**Fig. 2**).

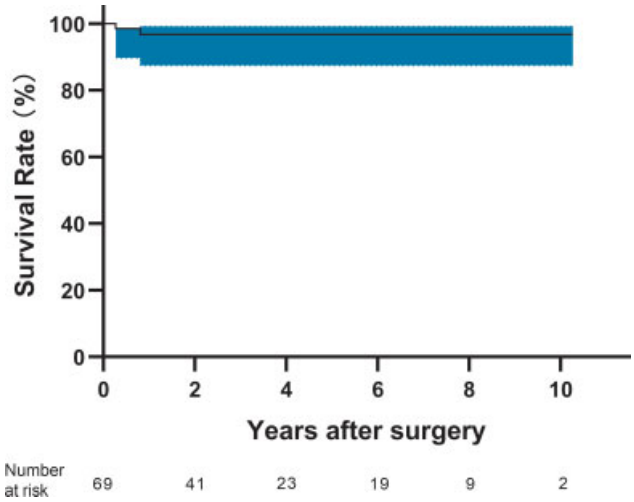

Fig. 2 The Kaplan–Meier survival rate after surgical correction of anomalous origin of the left coronary artery from pulmonary artery.

Multiple preoperative and intraoperative variables were used to identify risk factors for death. Preoperative EF (odds ratio = 0.946; 95% confidence interval [CI] = 0.901–0.994; $p = 0.027$) and LVEDD Z-score (odds ratio = 2.966; 95%CI = 1.278–6.884; $p = 0.011$) were regarded as significant predictors associated with mortality. However, age at operation, different surgical techniques of LCA transplantation, and MV intervention were not supposed to be risk factors. In terms of multivariable regression analysis, the LVEDD Z-score was considered to be an independent predictor of death (odds ratio = 3.382; 95%CI = 1.189–9.619; $p = 0.022$; ►**Table 2**). Furthermore, the ROC curve was used to assess the reliability of these two indicators. The area under the curve of preoperative EF and LVEDD Z-score was 77.3 and 75.9%, respectively. The optimal cutoff point for EF was 43.1%, which was associated with a sensitivity and specificity of 65.2 and 83.3%, respectively. Meanwhile, the optimal cutoff point for LVEDD Z-score was 2.5 with a sensitivity and specificity of 81.8 and 70.8%, respectively.

Mechanical Circulatory Support

Six patients required intraoperative or postoperative mechanical circulatory support. All of these patients were infants. Two patients received LVAD during the operation to manage severe LV dysfunction on weaning from CPB with a left atrial pressure over 15 mm Hg. The other four patients received LVAD implantation on postoperative day 1. Two of these patients were unable to maintain marginal arterial pressure, even with inotropic support, whereas the other two patients suffered from malignant ventricular tachycardia. These patients were supported with LVAD to allow for LV recovery. With the exception of one patient who died after LVAD support, all the other patients were successfully weaned from the assist device after 3 to 6 days.

Table 2 Univariate and Multivariate Analysis of Risk Factors for Mortality

	Univariate analysis			Multivariate analysis		
	Odds ratio	95% CI	*p* value	Odds ratio	95% CI	*p* value
Age	0.999	0.998–1.000	0.230			
Weight	0.942	0.832–1.066	0.341			
BSA	0.215	0.006–7.467	0.396			
LVEF	0.946	0.901–0.994	0.027	0.971	0.917–1.028	0.307
LVEDD *Z*-score	2.966	1.278–6.884	0.011	3.382	1.189–9.619	0.022
Moderate or greater MR	1.937	0.543–6.913	0.308			
Preoperative inotrope support	1.276	0.388–4.199	0.688			
Mitral valve intervention	1.339	0.373–4.805	0.654			
CPB time	0.999	0.985–1.013	0.849			
Aortic cross-clamp time	0.997	0.970–1.024	0.802			
LCA reimplantation technique	1.051	0.444–2.486	0.910			

Abbreviations: BSA, body surface area; CI, confidence interval; CPB, cardiopulmonary bypass; LCA, left coronary artery; LVEDD, left ventricular end diastolic diameter; LVEF, left ventricular ejection fraction; MR, mitral regurgitation.

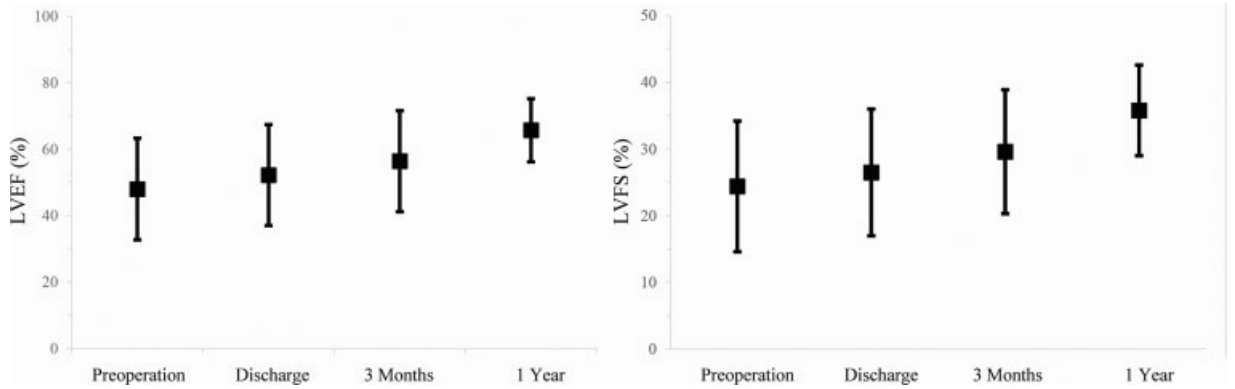

Fig. 3 The recovery of left ventricular ejection fraction and shortening fraction ($p < 0.01$ vs. preoperation; $p < 0.01$ vs. discharge; $p < 0.01$ vs. 3 mo).

Weight at the time of operation (odds ratio = 0.125; 95% CI = 0.025–0.629; $p = 0.012$) and preoperative EF (odds ratio = 0.872; 95%CI = 0.772–0.985; $p = 0.028$) were determined to be univariate factors for LVAD implantation. However, no variable was found to be significant by multivariable logistic analysis.

Cardiac Function

Postoperative TTE measures were required for all patients at discharge and follow-up. The median follow-up time was 32 (range = 3–124) months. A significant improvement in EF was present in all surviving patients at discharge (52.2 ± 15.2 vs. 48.0 ± 15.3 preoperatively, $p = 0.007$), at a 3-month follow-up (56.4 ± 15.2 vs. 52.2 ± 15.2 at discharge, $p = 0.002$) and at a 1-year follow-up (65.7 ± 9.5 vs. 56.4 ± 15.2 at a 3-month follow-up, $p = 0.003$; ►**Fig. 3**). Statistical analysis revealed an increasing SF of 26.5 ± 9.5 at discharge compared with 24.4 ± 9.8 preoperatively ($p = 0.025$), which was 29.6 ± 9.3 at the 3-month follow-up ($p = 0.002$) and 35.8 ± 6.8 at the 1-year follow-up ($p = 0.002$). The median duration of LV recovery to normal conditions (EF $\geq$ 55%) was 4 (range = 0–19.1) months.

Mitral Valve Regurgitation

During the follow-up period, MV function gradually improved in most patients. At discharge, no or mild MR was present in 57 patients, and moderate MR was present in 12 patients. No surviving patients were found to have severe MR. An increase in MR was found in only one patient, which went from moderate to severe. This patient died at home, as mentioned earlier in the section on mortality. The MV function of the other patients remained relatively stable after discharge.

We also compared the postoperative MV function between patients with concomitant MV repair and those without MV intervention, if they presented with more than moderate MR before surgery. Among the 17 patients with MV repair, no or mild MR was observed in 13 patients and moderate MR was observed in 4 at discharge. Among the 18 patients with more than moderate MR who did not undergo any MV repair, 6 patients exhibited moderate MR at discharge. There were no significant differences in MV function between the two groups at discharge and at the final follow-up.

Reoperation

A second operation was performed in two patients for main pulmonary artery stenosis at 1 and 8 years after the initial operation. These patients underwent transannular patching for pulmonary angioplasty. No patients received late MV repairs or replacements.

Infants versus Children

Differences in age at presentation may result in different surgical outcomes. When infant and child ALCAPA patient characteristics were directly compared, we found that the infant group had a lower preoperative EF and SF, higher LVEDD Z-score, and a higher incidence of inotropic support before surgery (►**Table 3**). Intraoperative and postoperative variables showed that a longer CPB time, longer duration of intubation time, and longer ICU stay were common in the infant group, and a greater number of infant patients required LVAD implantation, peritoneal dialysis, and delayed sternal closure. The infant patients showed a significant improvement in EF after surgical correction, whereas the older patients tended to show a slight decrease in EF after surgery (►**Fig. 4**). However, no differences were found in the surgical mortality and the severity of MR between these two groups.

Discussion

Surgical correction of ALCAPA has evolved during the past decades, evolving from LCA ligation and the Takeuchi procedure to the translocation of the LCA into the aorta.[6,7] The main purpose of surgical correction today is to establish a dual-coronary artery system soon after diagnosis of ALCAPA. Direct reimplantation of the LCA into the aorta is widely used.[8] This procedure is simple to perform, but it is influenced by several limiting factors, especially issues concerning the origin of the anomalous coronary artery from different pulmonary sinuses.

Reimplantation of the LCA to the aorta using the two-flap technique (aortic and pulmonary flaps) is now the favored

Table 3 Comparable analysis between infants and children

	Infant (49)	Children (31)	*p* value
Age (mo)	5.1 (1.4–11.8)	30.0 (12.4–141.3)	0.000
Weight (kg)	6.0 (4.0–9.5)	12.2 (6.9–51.0)	0.000
Preoperative LVEF (%)	41.7 ± 12.8	58.0 ± 13.7	0.000
Preoperative LVFS (%)	20.3 ± 7.9	31.0 ± 8.9	0.000
Preoperative LVEDD *Z*-score	2.4 ± 0.9	1.9 ± 0.9	0.012
Preoperative MR more than moderate (*N*)	24	14	0.739
Preoperative inotrope support (*N*)	32	7	0.000
CPB time (min)	118 (68–271)	93 (44–230)	0.027
Aortic cross-clamp time (min)	70 (30–131)	67 (22–165)	0.426
LVAD implantation (*N*)	5	0	0.183
Peritoneal dialysis (*N*)	11	1	0.048
Delayed sternal closure (*N*)	17	0	0.000
Postoperative ventilation times (h)	140 (46–336)	24 (4–238)	0.000
Postoperative ICU stays (d)	10.5 (4–45)	5 (2–14)	0.000
Duration of LVEF return to normal (mo)	0.8 (0–19.1)	0.1 (0–15.9)	0.070
Surgical mortality, *N* (%)	8(16.3%)	5(16.1%)	0.981

Abbreviations: CPB, cardiopulmonary bypass; ICU, intensive care unit; LVAD, left ventricular assistant device; LVEDD, left ventricular end diastolic diameter; LVEF, left ventricular ejection fraction; LVFS, left ventricular fraction of shortening; MR, mitral regurgitation.

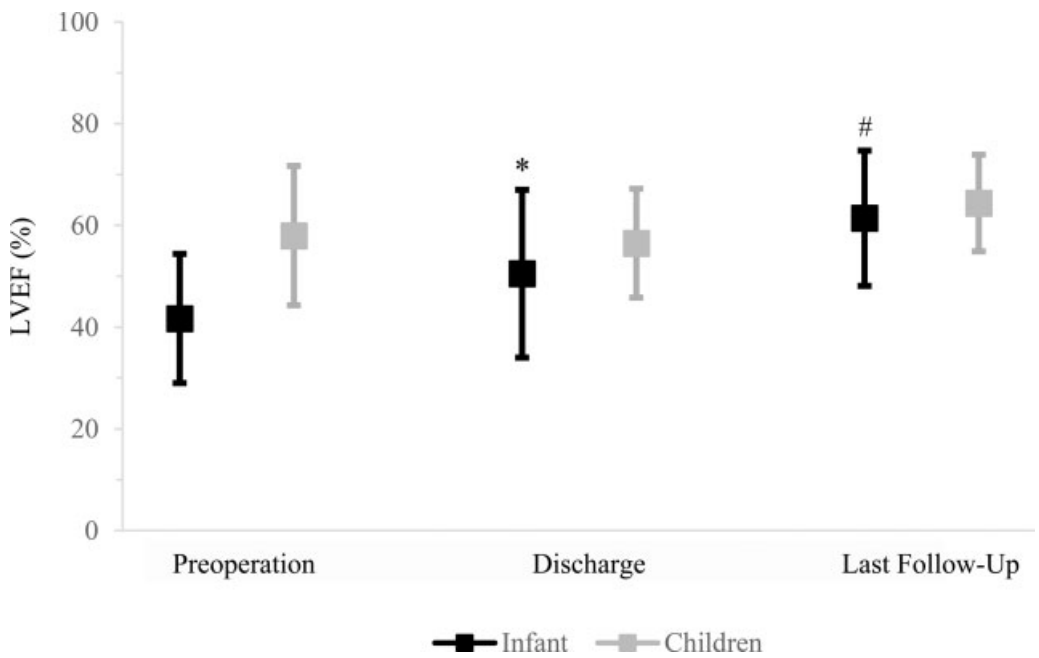

Fig. 4 The comparison of left ventricular ejection fraction between infants and children before and after surgery ($^{*}p < 0.01$ vs. preoperation; $^{\#}p < 0.01$ vs. discharge).

surgical choice, which can be recommended in all patients with ALCAPA, regardless of the origin of the anomalous coronary artery. The biggest benefit of this procedure is that it can be used to perform a tension-free anastomosis, thus preventing LCA stenosis and distortion postoperatively. When performing this procedure, the aortic flap is not supposed to be too large to prevent a long and blindly ending tunnel after anastomosis, which could result in LCA orifice thrombosis and occlusion. However, we found that regardless of the methods used, there was no difference in patient survival.

The optimal time for surgical correction of ALCAPA is supposed to be soon after clinical diagnosis regardless of the patients' age and cardiac function. We believe that it is true for most elderly patients with stable cardiac function. However, infants with ALCAPA are more likely to encounter acute

myocardial ischemia due to inadequate coronary collateral development after the pulmonary vascular resistance decreased and the coronary perfusion becomes compromised. For them, it is better to receive a short period of drug therapy such as digoxin and diuretic to extenuate myocardial cellular edema and recover the cardiac function as far as possible. If the preoperative inotropic therapy is found to be useless, surgical correction has to be performed immediately, and the mortality of these patients is definitely higher than other patients secondary to the close relationship between preoperative cardiac function and postoperative recovery.

At present, due to successful translocation techniques and the wide use of mechanical circulatory support, mortality and morbidity after ALCAPA repair have dramatically decreased. Many centers report relatively low hospital deaths and often no late deaths. At our institution, the mortality after surgical correction was 16.3%, which was slightly higher than some recent reports.[9–11] As far as we know, this is the largest ALCAPA patient series published with the largest number of infants included. The higher mortality is probably related to the higher severity of ALCAPA in infants before surgery and the limited surgical experience in developing countries during the early surgical period.

At the same time, poor EF and large LVEDD Z-scores were major predictors of mortality. To determine which indicators are more reliable in evaluating the correlation between LV function and mortality, an ROC curve was used. The areas under the curve for preoperative EF and LVEDD Z-scores were quite similar, but there was a higher sensitivity for LVEDD Z-scores and a higher specificity for EF, which could be explained by the fact that the LV dimension is supposed to change earlier than the LV systolic function.

Although significant improvement was found in patients before and after surgery, some patients still suffered from impaired LV function at discharge and follow-up. The median duration of LV recovery was 4 months. Some patients required closer to a year for LV function to recover. With effective inotropic support, LV function in these patients did not continue to deteriorate, with the exception of one patent who died of severe LV dysfunction 4 months after the initial surgery. Late death and additional surgical procedures were seldom necessary in patients with ALCAPA, unless coronary artery stenosis or severe MR was identified during the short-term follow-up.

No consensus has been reached yet about the criteria for MV repair. The majority of MR cases in ALCAPA were functional with a structurally normal MV, but a largely dilated annulus secondary to papillary muscle dysfunction and LV dilatation; in most cases, this will improve after ALCAPA repair. As a result, many centers have suggested avoidance of any MV interventions at the time of ALCAPA repair,[12] whereas others are in favor of concomitant MV repair in patients with severe MR.[8,13,14]

At our institution, we performed MV valvuloplasty in four patients with structural abnormalities and MV annuloplasty in nearly half of the patients with a normal MV structure. There were no significant differences in postoperative MR between patients with and without MV repair. Nevertheless, we found slightly fewer patients with moderate MR in the early postoperative period after MV intervention.

For patients with moderate or even severe MR, MV annuloplasty at the time of ALCAPA repair can decrease the severity of MR shortly after surgery, which might be beneficial for LV recovery and will help support cardiac output during the ICU stay. At the same time, it is reliable and safe to perform MV annuloplasty in cases where MV leaflets are unlikely to be damaged. The cross-clamp time should not increase very much. During follow-up, only one patient suffered from an increase in MR. The MV function of the other patients remained relatively stable. No secondary operation for severe postoperative MR or MV stenosis was required. Meanwhile, Azakie et al also found that an improvement in MR was related to the normalization of ventricular function.[15] As mentioned earlier, the median duration of LV recovery was 4 months. It has been speculated that some patients would encounter consistent postoperative MR if they presented unsatisfactory LV recovery after surgery. Some centers have even performed late MV interventions without any evidence of LV dysfunction.[2,15,16]

Some authors have also demonstrated that MV intervention should be performed in older patients due to their reduced probability of MV recovery secondary to irreversible myocardial damage or papillary muscle infarction.[12] However, we did not find any correlation between age at the time of repair and MV intervention; there were no significant differences in postoperative MR between infants and children. Therefore, our current policy is to repair a structurally abnormal MV and to perform MV annuloplasty in patients with moderate or severe MR, regardless of their age at the time of operation.

Mechanical circulatory support is frequently required in patients with ALCAPA to allow the LV to recover. Considering the high cost and morbidity after extracorporeal membrane oxygenation support, LVAD is our first choice for these patients, unless there is evidence of biventricular dysfunction. The indications for mechanical circulatory support are mainly based on our prior surgical experience. If the patients present with low cardiac output syndrome even with inotropic support or with malignant ventricular tachycardia after surgery, LVAD is recommended. The decision to use LVAD implantation should be made as soon as inotropic support is deemed ineffective. At our institution, 6 of 80 patients (7.5%) had an LVAD implanted after ALCAPA repair. All of these patients were younger than 6 months, which was similar to other reports, indicating that LVAD is rarely required in older patients.[17]

Limitations

There are several limitations of the present study. This is a retrospective analysis of a single center; some results may be affected by selection bias. Another limitation is that our average follow-up time is 32 months, with the longest duration being 10 years. Longer follow-up data are required to evaluate cardiac function and MR. We also lack magnetic resonance imaging facilities necessary to measure myocardial perfusion of LV.

Conclusion

In conclusion, this study demonstrates favorable outcomes of ALCAPA repair with an acceptable mortality rate and a very rare occurrence of secondary surgery. The LVEDD Z-score appears to be a sensitive predictor of death. LV recovery is a long-term process after surgical correction, especially for infants with impaired LV function. Concomitant MV annuloplasty is safe and reliable and can be performed as indicated in patients with moderate or severe MR.

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Conflict of Interest

None declared.

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