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Original article

Outcomes from anomalous origin of the left coronary artery from the pulmonary artery repair: Long-term complications in relation to residual myocardial abnormalities

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ABSTRACT

Background: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital coronary artery irregularity. This study aimed to clarify the long-term postoperative outcomes in ALCAPA patients, and to compare the infantile and adult types.
Methods: We retrospectively analyzed the clinical data from 33 patients with ALCAPA who underwent surgical repairs after 1980. The patients were grouped based on whether presentation occurred before (infantile type: $n = 14$) or after (adult type: $n = 19$) 1 year of age.
Results: The mean follow-up duration was 16 years. Preoperatively, the infantile type had greater impairment of the left ventricle ejection fraction (LVEF) ($45 \pm 15\%$) compared with the adult type ($59 \pm 10\%$) ($p < 0.01$). Coronary revascularization significantly improved the postoperative LVEF ($67 \pm 5\%$) ($p < 0.01$) in the patients with the infantile type. The postoperative LVEF did not change in the adult type. The mitral regurgitation (MR) severity improved postoperatively, but the between-group difference was not significant. Postoperatively, none of the patients with the infantile type and 37% of the patients with the adult type had left ventricular asynergy ($p = 0.01$), and both groups showed postoperative perfusion defects (79% vs 95%, $p = 0.29$). Compared with the infantile type, the adult type had a significant prognostic value for composite cardiovascular events that comprised cardiac death, arrhythmias, MR deterioration, and hospitalization as a consequence of heart failure ($p = 0.04$).
Conclusions: Most patients showed favorable clinical outcomes postoperatively, but myocardial damage remained long after surgery and cardiovascular events occurred postoperatively. Hence, meticulous long-term follow-up is warranted.

Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare malformation that occurs in one in 300,000 live births, and among all congenital heart diseases, it has an incidence of 0.25–0.50% [1]. In 1933, Edward Bland, Paul Dudley White, and Joseph Garland published the first clinical description of ALCAPA; consequently, ALCAPA is also known as Bland–White–Garland syndrome [2]. ALCAPA is largely asymptomatic in neonates because of the high pulmonary arterial pressure during the neonatal period. During the first months of life when the pulmonary vascular resistance and the oxygen content decline, the left coronary artery (LCA) changes to a retrograde flow with collaterals from the right coronary artery (RCA). Coronary steal from the LCA to the pulmonary artery (PA) leads to myocardial ischemia; consequently, symptoms develop. Collateral flow is an important factor in the development of myocardial ischemia. The nature and timing of the presentation of ALCAPA vary depending on the adequacy of the collateralized LCA circulation. ALCAPA can be classified into the infantile (early presentation) and adult (late presentation) types, based on the age at which the symptoms manifest themselves and the mode of presentation [3]. Currently, immediate surgical repair is the

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standard treatment for patients with ALCAPA. Left untreated, ALCAPA has a mortality rate of up to 90% in infancy [4]. However, a collateral circulation develops between the RCA and the LCA systems in the adult type, and patients may survive into adolescence or adulthood without any symptoms. The clinical course of ALCAPA differs according to the presentation type, but few studies' reports describe the long-term outcomes in the context of the presentation type. In this study, we sought to clarify the long-term postoperative outcomes in patients with ALCAPA and we compared the infantile and adult types.

Materials and methods

Forty-one consecutive patients with ALCAPA who underwent surgical coronary repairs at our institute after 1980 were enrolled to participate in the study. We retrospectively analyzed the clinical data from 33 patients with ALCAPA, comprising 9 males and 24 females. We excluded two patients who died perioperatively and six patients whose follow-up durations were <5 years (Fig. 1). At no less than 5 years after surgery, all of the patients underwent echocardiography and thallium cardiac perfusion scintigraphy under stress with adenosine or exercise.

We divided the patients into two groups based on the ages at which they presented with ALCAPA symptoms, namely, the group comprising the infantile type in which presentation occurred at <1 year of age ($n = 14$), and the group comprising the adult type in which presentation occurred at ≥1 year of age ($n = 19$). We retrospectively reviewed the patients' medical records and evaluated the preoperative and postoperative left ventricle ejection fractions (LVEFs) and the mitral regurgitation (MR) severities, the left ventricular asynergies that were determined using echocardiography, and the postoperative perfusion defects that were determined using cardiac perfusion scintigraphy. The MR severity was expressed as mild, moderate, or severe to categorize the regurgitation areas on the color Doppler images. The left ventricular asynergies were defined as segmental wall motion abnormalities in the anterior, high-lateral, or apical regions of the left ventricle. The patients' postoperative New York Heart Association (NYHA) classifications and the cardiovascular events, including cardiac death, arrhythmias, MR deterioration, and hospitalization as a consequence of heart failure, were recorded. The arrhythmias were defined according to the medical intervention required, namely, antiarrhythmic drugs, catheter ablation, device replacement, or cardioversion. MR deterioration was defined as an MR grade that had deteriorated during the long-term assessments to no less than moderate from mild or less soon after the operation.

The data for the normally distributed continuous variables are presented as the means ± standard deviations (SD), and the non-normally distributed variables are presented as the medians with the interquartile ranges. The data for the categorical variables are presented as numbers and relative frequencies. The differences in the continuous variables were assessed using Student's t-test, and the differences in the categorical variables were analyzed using the chi-square test. The cumulative cardiovascular event incidence was estimated using the Kaplan–Meier method, and the freedom from cardiovascular events was compared using the log-rank test. A value of $p < 0.05$ was considered statistically significant. All of the statistical analyses were performed using JMP Pro version 11.2.0 (SAS Institute Inc., Cary, NC, USA).

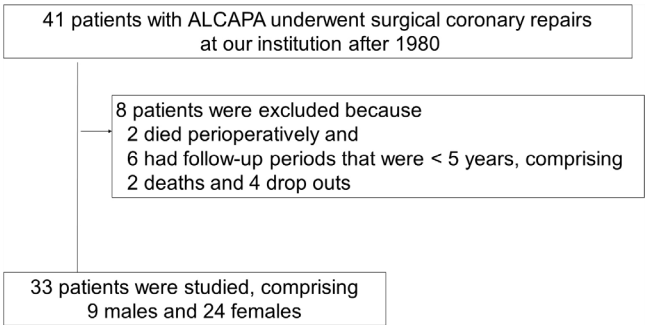

Fig. 1. Patient population. Forty-one consecutive patients with anomalous origin of the left coronary artery from the pulmonary artery who underwent surgical coronary repair at our institute after 1980 were enrolled to participate in the study, and eight patients were excluded because two died perioperatively and six had been followed up for fewer than 5 years. Finally, the data from 33 patients comprising nine males and 24 females were analyzed. ALCAPA, anomalous origin of the left coronary artery from the pulmonary artery.

Results

Table 1 presents the patients' characteristics. At surgery, the median age of the group of patients with infantile-type ALCAPA was 0.5 (range: 0.4–1.2) years, which was significantly lower than that of the group of patients with adult-type ALCAPA [11 (range: 5–18) years] ($p < 0.01$). The LCAs arose from the main PAs and no associated malformations were detected in any of the patients. The surgical method is selected based on the anatomical LCA origin. Hence, reimplantation is favored in cases where the LCA originates from the left posterior sinus of the main PA or the junction of the main PA and right PA, while the Takeuchi procedure is favored in cases where the LCA originates from the left and lateral sides of the main PA. Concomitant mitral annuloplasty was undertaken in approximately 80%of the patients with MR that was no less than moderate or visible ischemic changes of papillary muscle during operation regardless of MR grade in the two groups. There were no significant differences between the groups in relation to the surgical methods used. The age at which the patients were studied was significantly lower in the group with infantile-type ALCAPA [23 (range: 17–30) years] compared with the group with adult-type ALCAPA [34 (range: 27–44) years] ($p < 0.01$). The median follow-up durations were similar for the infantile [13 (range: 8–23) years] and the adult [18 (range: 9–22) years] types ($p = 0.99$).

Fig. 2 shows the preoperative and postoperative LVEFs. Preoperatively, the left ventricular systolic function in the patients with infantile-type ALCAPA was more severely impaired compared with that in the patients with adult-type ALCAPA (LVEFs: 45 ± 15% and 59 ± 10%, respectively, $p < 0.01$). After coronary revascularization, the LVEF in the patients with infantile-type ALCAPA improved significantly (66 ± 5%) compared with the preoperative LVEF ($p < 0.01$) and compared with the postoperative LVEF in the patients with adult-type ALCAPA (57 ± 11%) ($p = 0.01$). The postoperative LVEF did not change significantly compared with the preoperative

Table 1
Patients' characteristics.

	Infantile type ($n = 14$)	Adult type ($n = 19$)	p value
Sex, male, n (%)	2 (14)	7 (37)	0.24
Age (years)[a]	23 (17–3)	34 (27–44)	<0.01
Age at operation (years)[a]	0.5 (0.4–1.2)	11 (5–18)	<0.01
Operative method, n (%)			
Reimplantation	6 (43)	14 (74)	0.07
Takeuchi	8 (57)	5 (26)	
Concomitant mitral annuloplasty, n (%)	11 (79)	15 (79)	1.00
Postoperative follow-up duration (years)[a]	13 (8–23)	18 (9–22)	0.99

[a] The data presented are the medians (interquartile ranges).

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