Diagnosis and treatment of abnormal left coronary artery originating from the pulmonary artery: A single-center experience

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ABSTRACT

Objective: We aimed to review symptoms, findings, surgical treatment options, short- and mid-term outcomes, and reoperation rate of patients diagnosed with left coronary artery from the pulmonary artery (ALCAPA) of an anomalous origin in our institution.

Methods: From May 2000 to March 2018, 33 patients who had left coronary artery originating from the pulmonary artery were retrospectively examined. The clinical features of patients, diagnostic tools and their efficacy, outcomes of surgical repair, and problems during follow-up were evaluated.

Results: Thirty-three patients (22 females, 11 males) were included in the study. At the time of surgery, the median age and weight of patients were 6 months (minimum/maximum, 1–166 months) and 6.5 kg (minimum/maximum, 3–38.5 kg), respectively. The mean follow-up was 5±3.5 years (range, 1–16 years). Dyspnea, tachypnea, diaphoresis, prolonged feeding time, and developmental delay were common presenting signs and symptoms. It was determined that all the patients who were diagnosed at another center reached our center for surgical treatment within 1 month. Twenty-three (69.7%) patients had pathologic Q wave with anterior and/or anterolateral myocardial infarction signs on an electrocardiogram (ECG), whereas 22 (66.6%) patients had ST-T segment changes. Twenty-one (63.6%) patients had cardiomegaly on the telecardiogram. A reimplantation surgery was performed to 22 patients and 10 patients underwent the Takeuchi procedure. In addition to ALCAPA repair, 5 patients needed mitral valve plasty. Atrial septal defect (ASD) and ventricular septal defect (VSD) were closed in one patient, and Tetralogy of Fallot was totally corrected in another. At discharge, there was a significant improvement in left ventricular (LV) systolic functions. At the last visit, all patients had normal LV systolic functions except four who had mild dysfunction. The mean follow-up of the four patients was 2.8 years. In the early postoperative period, complications were seen in 10 patients. Five patients died in the early postoperative period, while one patient died 9 months after the ALCAPA surgery because of low cardiac output syndrome that developed after mitral repair.

Conclusion: Patients with ALCAPA commonly present with congestive heart failure symptoms. When the diagnosis is confirmed in these patients, surgical treatment should not be delayed. The availability of surgical center and surgery outcomes for ALCAPA diagnosed patients are comparable with other countries, but the delay in the diagnosis of disease is still a problem in our country. (Anatol J Cardiol 2019; 22: 325-31)

Keywords: anomalous origin of coronary arteries, dilated cardiomyopathy, echocardiography, surgical treatment

Introduction

The anomalous origin of the left coronary artery from the pulmonary artery is a rare congenital heart disease that causes coronary artery steal, ischemia, left ventricular (LV) dysfunction, and mitral regurgitation. It represents 0.25%–0.5% of all congenital heart defects (1, 2). Since it was first described by Bland, White, and Garland in 1933, it is also known as the Bland-White-Garland Syndrome (3). Myocardial infarction, dilated cardiomyopathy, and mitral regurgitation are frequently seen due to impaired muscle perfusion. Clinical manifestations often occur in infancy when pulmonary vascular resistance and coronary artery blood flow drop. In the early infancy, myocardial ischemia leads to typical symptoms, such as poor feeding, developmental delay, diaphoresis, dyspnea, and pallor, which are rarely seen in infants younger than 2 months. Mitral regurgitation and LV dysfunction are common cardiac pathologies caused by myocardial ischemia. The mortality rate is high when ALCAPA is not repaired in the first year of life. The treatment and long-term problems of 33 patients who were diagnosed with ALCAPA in our clinic are reported.
Methods

The records of 33 patients who were diagnosed with ALCAPA in our center along with the long-term surgical outcomes and reoperations were retrospectively reviewed. The Ethics Committee of the University approved the study. Informed consent was obtained from all individual participants included in the study. Further, 12-lead ECG, transthoracic echocardiographic examination, telecardiogram, and cardiac catheterizations were performed for all patients. Also, computed coronary artery angiography (CCA) was used in selected patients. When the ALCAPA diagnosis was confirmed, patients were treated surgically without delay. Ejection fraction (EF) and shortening fraction (SF) were employed to classify LV systolic dysfunction. According to this, LV systolic function was accepted as normal when LV-EF ≥60%, and there were mild, moderate, and severe systolic dysfunction if LV-EF was 50–59%, 35–49%, and <35%, respectively. Mitral regurgitation and endocardial fibroelastosis were demonstrated by transthoracic echocardiography. Mitral regurgitation was graded as I (none/trivial), II (mild), III (moderate), and IV (severe). Cardiomegaly was defined as cardiothoracic ratio greater than 0.55 as seen on telecardiogram.

Surgical technique

The Takeuchi technique was to be used initially. In this technique, an intrapulmonary tunnel was created using a bovine pericardial patch. The intrapulmonary baffle can be created using a flap tissue from the pulmonary artery wall, autologous pericardial tissue, or synthetic material. The bovine pericardial patch was a readily available substitute with perfect handling and was used as the personal choice of the surgical team. Direct pulmonary repair (8 patients) or patch augmentation (2 patients) of the pulmonary artery following intrapulmonary reconstruction of the baffle was done depending on each patient’s specifications, mainly on the size of the patient. This technique was used in the beginning of the series when it was popular and adopted by most centers. However, it was abandoned because it was seen to be associated with a high rate of pulmonary stenosis development and reoperation. Direct coronary implantation was the second technique to be chosen, and this procedure was performed in the last 22 patients. Following sternotomy, patients were placed on cardiopulmonary bypass (CPB) by means of aorto-bicaval cannulation. The pulmonary artery branches were controlled using snare. HTK Custodiol solution was infused through the aortic root cannula and diastolic cardiac arrest was achieved. In patients with left coronary artery originating from adjacent sinus left coronary artery was directly re-implanted to the aorta with a button of the pulmonary artery. When the left coronary artery originated from the remote sinus, the pulmonary artery was transected distal to the origin of the left coronary artery, the posterior pulmonary commissure was taken down using fine scissors. If the ostium was located deep in the sinus, a generous flap of posterior pulmonary wall was tailored, folded, and sutured using 7/0 propylene suture as a tube, and the coronary artery was extended and anastomosed to the aorta. A pulmonary artery defect was reconstructed with fresh autologous pericardial patch.

Three of 10 patients who had undergone the Takeuchi procedure and 2 of 22 patients who had undergone direct coronary re-implantation had concomitant mitral valve repair for severe mitral regurgitation. These interventions mostly addressed structural valvular defects, such as mitral clefts. Simple mitral valve repair techniques, such as commissural sutures and simple annular reduction techniques were preferred. More comprehensive mitral repair was reserved for late interventions in older children.

Statistical analysis

Statistical analysis was performed using the PASW version 17.0 software (SPSS™ Inc., Chicago, IL, USA). Descriptive statistics were expressed in mean±standard deviation, and frequency (%). A p value less than 0.05 was considered statistically significant.

Results

Thirty-three patients (22 females, 11 males) were included in this study. At the time of diagnosis, the median age of patients was 6 months (minimum/maximum, 1–164 months), whereas at the time of surgery, the median of age and weight was 6 months (minimum/maximum, 1–166 months) and 6.5 kg (minimum/maximum, 3–38.5 kg), respectively. The follow-up period varied from 1 to 16 years averaging 5±3.5 years. At the time of diagnosis, 18 patients were younger than 6 months of age, and 5 of them were 6–12 months of age; 10 patients were older than 1 year. Dyspnea, tachypnea, diaphoresis, early fatigue during feeding, and developmental delay were frequent presenting signs and symptoms. ALCAPA was associated with congenital heart disease in two patients and with cleft palate in one (Table 1). In 29 (87.9%) patients, echocardiography (Video 1) and cardiac catheterization (Fig. 1) were sufficient to diagnose the disease. CCA was needed in three (9.1%) patients to definitely diagnose ALCAPA in other centers before they were admitted to our clinic. ALCAPA was identified in one patient during complete repair of Tetralogy of Fallot (TOF). Echocardiographic examination showed coronary artery collaterals on the interventricular septum in 26 patients (78.8%) (Video 2), coronary retrograde flow into the pulmonary artery in 30 patients (90.9%), and right coronary artery (RCA) dilation in 18 (54.5%) patients. Fourteen (42.4%) patients were diagnosed with dilated cardiomyopathy previously in other centers. One of these patients underwent mitral valve replacement (MVR) at 3 years of age due to severe mitral regurgitation. When this patient was 5.5 years old, control echocardiographic examination revealed a collateral artery flow on the interventricular septum, and the diagnosis of ALCAPA was confirmed by catheter angiography. ALCAPA could be shown only when the second angiocardiography performed in other two patients with
dilated cardiomyopathy. Ten (30.3%) patients were diagnosed after 1 year of age. Twenty-three patients (69.7%) had pathological Q wave that indicated anterior/anterolateral myocardial infarction, and 22 patients (66.6%) had ST-T segment changes on ECG. Cardiomegaly was defined in 21 patients (63.6%) by a telecardiogram. The mean cardiothoracic rate was 61.3%±6.9% (range, 50 to 76%). Reimplantation was performed in 22 patients, whereas 10 patients underwent the Takeuchi procedure. Besides ALCAPA repair, five patients needed mitral valve repair, VSD and ASD were closed in one patient, and TOF was completely corrected in one (Table 1). One patient could not be treated surgically because the family refused treatment. This patient now has severe systolic dysfunction (EF: 33%, FS: 15%), and he has been followed for seven years. The mean CPB time was 125±19.7 minutes (range, 91–172 minutes), whereas the average cross clamping time was 85±17.6 minutes (range, 50–125 minutes). Although the mean positive inotropic support duration was 5.6±3.3 days, it varied from 1 to 14 days in different patients. The mean duration of intubation was 2.9±2.3 days (range, 1–14 days). The mean duration of intensive care unit stay was 6.2±3.3 days (range, 1–14 days), and the mean length of hospital stay was 11.7±3.4 days (range, 6–18 days). EF and FS averaged 44.6±1 6.7% (range, 21–76%) and 22.3±9.8% (range, 9%–42%), respectively in the preoperative period, whereas in the postoperative period, at discharge, these measurements increased when their mean values were 49.8±12% (range, 24%–70%) and 25.2±6.9% (range, 11–39%), respectively. LV systolic function was normal in seven patients, whereas LV systolic function was depressed mildly in seven, moderately in seven, and severely in 11 patients before surgery. After surgery, at discharge, seven patients had normal LV systolic function, whereas there was mild left ventricular (LV) systolic dysfunction in eight, moderate in nine, and severe in three patients (Table 2). A considerable improvement in LV systolic function was seen at the time of discharge (p<0.05). At the last visit, mild systolic dysfunction was observed in four patients, and the mean follow-up of these patients was 2.8 years. In contrast, LV function returned to normal in the rest of patients. There was a big improvement in LV systolic function at the most recent clinic visit (p<0.05).

Mitral regurgitation was none/trivial in seven patients while it was mild in nine, moderate in 13, and severe in two patients prior to treatment. Also, mitral valve had been replaced because of severe mitral regurgitation in one patient before he was diagnosed with ALCAPA. There was none/trivial mitral regurgitation in seven patients, whereas mild regurgitation in 13, moderate regurgitation in two patients, and severe regurgitation in one patient were observed in the postoperative period at the last visit (Table 2). As mentioned earlier, one patient had undergone MVR 2.5 years before the diagnosis of ALCAPA. Also, another patient developed severe mitral regurgitation after ALCAPA repair and MVR was performed to this patient. Finally, the mitral valve was replaced in the two patients. Echocardiographic examination showed marked decrease in the degree of mitral regurgitation at the most recent visit (p<0.05).

Complications were seen in 10 (31.2%) patients in the early postoperative period. Supravalvular pulmonary stenosis developed in four patients. The other complications were left diaphragmatic paralysis in two patients, mild mitral valve stenosis in one, and sepsis in one. Coronary tunnel leaked in one patient while another one had junctional ectopic tachycardia.

Mortality was observed in six (18.7%) patients included in the study (Table 1). Low cardiac output syndrome caused death in four patients in the early postoperative period. A 13-day-old baby died on the fifth postoperative day due to sepsis, and this was the only death seen in the last 24 patients. One patient developed progressive mitral regurgitation following ALCAPA repair. She
Table 1. Clinical characteristics of the patients and cardiac examination

| Diagnoses | Sex | Surgery- age | Surgery | Surg-Wt (kg) | Mid-term complications | EF/FS before surgery | Diagn methods | Follow-up time |
|-----------|-----|--------------|---------|--------------|------------------------|----------------------|---------------|---------------|
| ALCAPA    | M   | 96 M         | Takeuchi+MVP | 23           | Supravalvar PS (moderate)+PR (mild) | 56/28                | Echo, Cath, CCA | -             |
| ALCAPA    | F   | 11 M         | Takeuchi   | 8            | Supravalvar PS (moderate)+PR (mild) | 34/17                | Echo, Cath    | 8 years       |
| ALCAPA    | F   | 10 M         | Takeuchi+MVP | 6.5          | Supravalvar PS (moderate)+ coronary artery tunnel leak | 72/41                | Echo, Cath    | 14 years      |
| ALCAPA    | F   | 10 M         | Takeuchi   | 8.5          | Supravalvar PS (severe)+PR (mild) | 47/23                | Echo, Cath    | 6 years       |
| ALCAPA    | F   | 6 M          | Takeuchi   | 5.5          | Supravalvar PS (moderate)+ coronary artery tunnel leak | 27/12                | Echo, Cath    | 16 years      |
| ALCAPA    | F   | 25 M         | Takeuchi+MVP | 12          | Supravalvar PS (moderate)+PR (mild) | 46/22                | Echo, Cath    | 5 years       |
| ALCAPA, TOF | F  | 30 M        | Takeuchi,TOF surg | 14        | - | 64/34 | Echo, Cath, Surg | 7 years |
| ALCAPA    | F   | 3 M          | Takeuchi+MVP | 4.5          | - | 51/25 | Echo, Cath | 5.5 years |
| ALCAPA    | M   | 3 M          | Takeuchi   | 5            | - | 23/10 | Echo, Cath | 5.4 years |
| ALCAPA    | F   | 2 M          | Takeuchi   | 4.5          | - | 36/16 | Echo, Cath | - |
| ALCAPA    | F   | 3 M          | Reimplant  | 4.5          | - | 25/12 | Echo, Cath | 5 years |
| ALCAPA    | F   | 104 M        | Reimplant   | 24           | - | 62/32 | Echo, Cath | 3 years |
| ALCAPA    | F   | 38 M         | Reimplant  | 12           | Supravalvar PS (mild) | 55/28                | Echo, Cath    | 3 years |
| ALCAPA    | F   | 5 M          | Reimplant  | 5.5          | - | 64/33 | Echo, Cath | 2.5 years |
| ALCAPA    | M   | 4 M          | Reimplant  | 4.7          | Left main coronary artery stenosis | 23/10                | Echo, Cath    | 7 years |
| ALCAPA    | M   | 2 M          | Reimplant  | 3.4          | PR (moderate) | 56/27                | Echo, Cath    | 4 years |
| ALCAPA    | F   | 5 M          | Reimplant  | 6            | - | 36/18 | Echo, Cath | - |
| ALCAPA    | F   | 3 M          | Reimplant  | 5            | - | 49/20 | Echo, Cath | - |
| ALCAPA    | F   | 3 M          | Reimplant  | 5.5          | - | 52/26 | Echo, Cath | 6 years |
| ALCAPA, VSD, ASD | F | 1 M | Reimplant,VSD,ASD | 3.5 | - | 34/16 | Echo, Cath | - |
| ALCAPA    | F   | 6 M          | Reimplant  | 6.5          | - | 44/24 | Echo, Cath, CCA | 5 years |
| ALCAPA    | M   | 165 M        | Reimplant   | 38           | - | 70/40 | Echo, Cath | 3 years |
| ALCAPA    | F   | 66 M         | Reimplant  | 16           | - | 27/13 | Echo, Cath | 4 years |
| ALCAPA, MVR (36 M) | M | 66 M | Reimplant | 16 | - | 54/27 | Echo, Cath | 10 years |
| ALCAPA    | M   | 166 M        | Reimplant+MVP | 38.5      | - | 64/35 | Echo, Cath | 2 years |
| ALCAPA    | F   | 5 M          | Reimplant  | 5.5          | - | 76/42 | Echo, Cath | 3 years |
| ALCAPA    | F   | 3 M          | Reimplant  | 4.5          | - | 25/12 | Echo, Cath | 5 years |
| ALCAPA    | M   | 5 M          | Reimplant  | 6.2          | - | 22/11 | Echo, Cath | 4.5 years |
| ALCAPA    | F   | 7 M          | Reimplant  | 6.6          | - | 22/10 | Echo, Cath | 1.5 years |
| ALCAPA    | F   | 3 M          | Reimplant  | 4.6          | - | 46/22 | Echo, Cath, CCA | 4.5 years |
| ALCAPA    | F   | 9 M          | Reimplant  | 7.3          | - | 58/29 | Echo, Cath | 1 year |
| ALCAPA    | M   | 6 M          | Reimplant  | 6.5          | - | 21/10 | Echo, Cath | - |
| ALCAPA    | M   | -            | No surgery | -            | - | 33/15 | Echo, Cath | 7 years |

ALCAPA - anomalous left coronary artery originating from the pulmonary artery, ASD - atrial septal defect, Cath - catheter angiography, CCA - computed coronary angiography, Echo - echocardiography, EF - ejection fraction, MVP - mitral valve plasty, MVR - mitral valve replacement, PS - pulmonary stenosis, PR - pulmonary regurgitation, FS - shortening fraction, Surg - surgery, VSD - ventricular septal defect, TOF - tetralogy of Fallot, Wt - weight
had had moderate mitral regurgitation before ALCAPA repair, and hence a concomitant intervention for the mitral valve was not considered during the initial surgery. She had undergone a mitral valve plasty procedure 9 months after the ALCAPA repair. The weaning from CPB was uneventful. The early postoperative course was as expected; however, this patient died in the intensive care unit 6 days after the surgery because of low cardiac output syndrome. Her postoperative LV function was not worse than preoperative measurements; therefore, an extracorporeal membrane oxygenation (ECMO) run was not considered. Even though ECMO was readily available for patients with LV systolic dysfunction it was not employed because all patients could be weaned successfully and easily from CPB.

In the mid-term postoperative period, 50% patients palliated with the Takeuchi procedure experienced complications, while this ratio was 13.6% in the reimplantation group (Table 1). The reoperation rate was 30% among patients who underwent the Takeuchi procedure, and it was 4.5% in reimplantation patients. Supravalvar pulmonary stenosis was repaired in three patients who were in the Takeuchi performed group, and coronary tunnel was repaired in two of these patients. The left main coronary artery was reconstructed in a patient who was in the reimplantation group. In addition to this MVR in one patient and mitral valve repair was performed in another one. At discharge, 27 patients were on aspirin and one on warfarin therapy, and 22 patients were receiving angiotensin-converting enzyme inhibitors. Also, diuretics were prescribed to 15 and digoxin to 14 patients.

### Table 2. Left ventricular and mitral valve functions before and after surgical treatment

| LV dysfunction  | Before surgery number of the patients | Hospital discharge number of the patients | Last follow-up number of the patients |
|-----------------|---------------------------------------|------------------------------------------|---------------------------------------|
| Absent          | 7                                     | 7                                        | 22                                    |
| Mild            | 7                                     | 8                                        | 4                                     |
| Moderate        | 7                                     | 9                                        | Absent                                |
| Severe          | 11                                    | 3                                        | Absent                                |
| Mitral regurgitation grade | | | |
| Absent/Trivial  | 7                                     |                                         | 8                                     |
| Mild regurgitation | 9                                   |                                         | 13                                    |
| Moderate regurgitation | 13                                  |                                         | 2                                     |
| Severe regurgitation | 2                                    |                                         | 1                                     |
| MVR             | 1                                     |                                         | 2                                     |

MVR - mitral valve replacement, LV - left ventricular

Discussion

The anomalous origin of left coronary artery from the pulmonary artery is a rare congenital cardiac malformation. Since the pressure and oxygen saturation of the aorta and pulmonary artery are equal in the fetus, there is no overt evidence of disease during fetal life and collateral do not occur in this period because myocardial perfusion is normal. However, after birth, the pulmonary vascular resistance falls, resulting in developing coronary steal phenomenon, which causes ischemia. Also, the onset of symptoms depends on the number of collaterals between right and left coronary arteries and pulmonary artery pressure (2). It is known that some patients are surviving to adulthood, as they have a narrowed slit like the left coronary artery ostium that limits coronary steal. Congestive heart failure symptoms, which are caused by myocardial ischemia are presenting symptoms in early infancy in a majority of patients. Similarly, congestive heart failure signs and symptoms were seen commonly in our patients.

ALCAPA may cause anterolateral myocardial infarction that manifests as pathologic Q waves in DI, aVL, and V4-V6 on ECG. ECG changes may be nonspecific in patients with intense collateral circulation (4). Retrograde flow from left coronary artery to pulmonary artery, RCA dilation, and collateral arteries on the interventricular septum were demonstrated in an echocardiographic examination in the large number of patients, and the presence of these echocardiographic features was considered valuable markers to identify ALCAPA in this study. Although echocardiography and color Doppler flow give useful information about the disease, cardiac catheterization should be performed in all patients suspected to have ALCAPA. CCA and cardiovascular magnetic resonance imaging are very useful alternative diagnostic tools, which help to evaluate the origin of coronary artery, ventricle function, and valve regurgitation not only in the preoperative period but also in postoperative period (5). In our study CCA was obtained at other centers and it confirmed ALCAPA diagnosis in three patients. Also, in our series common ECG findings in patients with LV dysfunction were pathologic Q wave and ST-T segment changes.
Although ALCAPA is usually isolated cardiac anomaly, cases associated with patent ductus arteriosus, VSD, TOF, and aorta coarctation were reported (4, 6). In our series, ALCAPA was accompanied by other congenital heart diseases in a small number of patients similar to literature.

The majority of patients with ALCAPA are initially misdiagnosed as dilated cardiomyopathy or endocardial fibroelastosis (4, 7). Zheng et al. (4) reported that 78.3% of patients were initially misdiagnosed in their study. Also, 66.6% of patients in Ramirez et al. (7) study had false initial diagnosis. We found that 42.4% of patients were misdiagnosed as dilated cardiomyopathy, and this was lower than the rate reported in literature. Also, one patient after MVR and two patients in the second catheter angiography could be diagnosed in our institution. ALCAPA should be definitely investigated in patients with idiopathic dilated cardiomyopathy or unexplained mitral regurgitation.

It is now known that historical ligation of abnormal coronary artery does not change survival. In fact, anatomic repair that consists two coronary circulation has a positive effect on survival at all ages, and it is now the preferred surgical treatment. The Takeuchi procedure and reimplantation of the coronary artery in the ascending aorta are two commonly used surgical treatment options (4, 7). In the Takeuchi procedure, creating aortopulmonary window is followed by redirecting blood flow from the ascending aorta into the left coronary artery ostium directly by an intrapulmonary tunnel (8). Neches et al. (9) described left main coronary artery reimplantation in 1974. Jin et al. (10) reported good results and improvement in the LV functions with the reimplantation of the left coronary artery into the ascending aorta in 11 patients. Early mortality rate was reported as 9.5% by Dehaki et al. (6) in a reimplantation series with 10 patients. It has been shown that the coronary implantation technique increases survival and decreases coronary stenosis and the reoperation rate (11-13). No patient developed coronary/pulmonary stenosis or was reoperated in the study by Mongé et al. (11), which consisted 36 patients who underwent reimplantation. Ben Ali et al. (13) reported that 2 of 61 patients who underwent reimplantation were reoperated because of coronary artery stenosis. The operative mortality depends on the surgical technique, and it varies as 0%–23% (4, 6, 7, 10, 14-16). In a study by Zheng et al. (4), 6 of 21 patients died in the early postoperative period. There was no early or late death in the Mongé et al. (11) study on 36 patients with reimplantation and in the Ramirez et al. (7) study on 15 patients with reimplantation. Although the Takeuchi surgery was preferred initially, it has not been used in the last 10 years in our clinic, as a high complication rate was seen in patients who underwent this procedure. One patient who was in the reimplantation group developed reconstruction requiring left coronary artery stenosis. Coronary reimplantation technique is recently the procedure of choice in our center since it has a lower reoperation and complication rate. The early mortality rate was low in our series, as it was reported in literature.

The mortality rate is very high in untreated ALCAPA patients. Even though some patients with collaterals can survive until adulthood, they are at risk of heart failure and sudden death. One patient with ALCAPA who had dilated cardiomyopathy and severe LV dysfunction could not be operated because his family approval for procedure could not be obtained. This patient has been receiving medical treatment for 7 years and remains to have severe LV systolic dysfunction (EF, 33%; FS, 15%). In addition, 30.3% of patients were older than 1 year when they were diagnosed with ALCAPA. Interestingly, two patients were diagnosed at 13 and 14 years of age, but they had no evidence of mitral regurgitation or LV dysfunction. LV function was not impaired in these two patients due to good collateral circulation.

LV dysfunction has been reported as the main risk factor for perioperative mortality in a number of studies (12, 17, 18). In our series, five patients died in the early postoperative period. Two of these patients were in the Takeuchi performed group, and the other three patients were in the reimplantation group. We did not find a statistically significant relationship between the type of surgical technique and early mortality. All the patients had moderate to severe LV dysfunction before surgical treatment. In this study, moderate to severe LV dysfunction was the major risk factor of early postoperative mortality, as it was reported in literature.

**Study limitations**

This was a retrospective study and the main limitation is the lack of information about the patients who were lost to follow-up.

**Conclusion**

ALCAPA should be considered in all infants with dilated cardiomyopathy and/or unexplained mitral regurgitation. Reimplantation surgery has become applicable with a high success and low complication rate. LV dysfunction is the main risk factor of surgical mortality. Moderate to severe mitral regurgitation can also be an important risk factor for mortality in the early postoperative period. It should be known that LV systolic function becomes normal in all patients who can survive in the early postoperative period.

**Conflict of interest:** None declared.

**Peer-review:** Externally peer-reviewed.

**Authorship contributions:** Concept – N.K.T., B.V.; Design – K.Y., M.Ö.; Supervision – K.Y., M.Ö.; Funding – None; Materials – K.Y., N.K.T.; Data collection and/or processing – M.Ö., I.E.; Analysis and/or interpretation – K.Y., N.K.T.; Literature search – B.V.; Writing – K.Y., N.K.T.; Critical review – N.K.T., B.V., M.S.A.

**Video 1.** Echocardiographic examination shows a left coronary artery originating from the pulmonary artery

**Video 2.** Collateral arteries flow on the interventricular septum are demonstrated in an echocardiographic examination
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