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Original Article / Özgün Makale

Anomalous left coronary artery arising from the pulmonary artery repair with pulmonary artery reconstruction

Pulmoner arter çıkışlı sol koroner arterin pulmoner arter rekonstrüksiyonu ile düzeltilmesi

M. Fatih Ayık,[1] Emrah Oğuz,[1] Pelin Öztürk,[1] Yüksel Atay,[1] Naim Ceylan,[2] Ertürk Levent,[3] Ruhi Özyürek,[3] Emin Alp Alayunt[1]

[1]Department of Cardiovascular Surgery, Medical Faculty of Ege University, İzmir, Turkey;
[2]Department of Radiology, Medical Faculty of Ege University, İzmir, Turkey;
[3]Department of Pediatric Cardiology, Medical Faculty of Ege University, İzmir, Turkey

Background: This study aims to report the clinical features, surgical management and outcome of patients with anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) who underwent Takeuchi operation combined with pulmonary artery reconstruction.

Methods: We retrospectively reviewed the charts of seven patients (2 females, 5 males, mean age 53.3±52.1 months; range 6 to 151 months) with the diagnosis of ALCAPA who underwent Takeuchi operation combined with pulmonary artery reconstruction between January 2007 and June 2010 in our clinic. Detailed cardiovascular examination including electrocardiography, chest X-ray, echocardiography, computed tomography coronary angiography and cardiac catheterization were performed preoperatively on all subjects. Demographic characteristics as well as pre- peri- and postoperative data were documented and evaluated.

Results: The most common symptom was periodic dyspnea (57.1%), however 28.6% of the patients were asymptomatic. The mean cardiopulmonary bypass duration and mean aortic cross-clamp time were 105.8±33.0 and 69.4±35.1 min, respectively. Postoperative durations of mechanical ventilation, Intensive Care Unit stay, and normal ward stay were 15.0±14.9 hours, 1.8±0.5 days, and 6.0±2.7 days, respectively. Malignant arrhythmia was the only complication of surgery which was identified in one patient who died at the 36 hour of post cardiac surgery. Six of seven patients who underwent Takeuchi operation with pulmonary artery reconstruction (85.7%) survived and among all of the surviving patients were symptom-free within a mean follow-up of 11.7±10.3 months.

Conclusion: ALCAPA is a kind of pathology which can be corrected by surgery and an early diagnosis and intervention are of utmost importance for the long-term prognosis.

Key words: ALCAPA; pulmonary artery reconstruction; Takeuchi operation.

Amaç: Bu çalışmada kliniğimizde pulmoner arter çıkışlı sol koroner arter anomalisi (ALCAPA) tanısı konmuş olan hastaların klinik özellikleri, cerrahi tedavisi ve Takeuchi ameliyatı ile birlikte pulmoner arter rekonstrüksiyonu sonuçları bildirildi.

Çalışma planı: Ocak 2007 - Haziran 2010 tarihleri arasında kliniğimizde Takeuchi ameliyatı ile birlikte pulmoner arter rekonstrüksiyonu yapılan yedi hastanın (2 kız, 5 erkek; ort. yaş 53.3±52.1 ay; dağılım 6-151 ay) kayıtları retrospektif olarak değerlendirildi. Tüm hastalara ameliyat öncesi detaylı kardiyovasküler muayene, elektrokardiyografi, direkt göğüs filmi, ekokardiyografi, bilgisayarlı tomografi koroner anjiyografi ve kardiyak kateterizasyon yapıldı. Demografik özellikler, ameliyat öncesi, esnası ve sonrası veriler kaydedilerek değerlendirildi.

Bulgular: En sık görülen semptom, periyodik dispne (%57.1) idi, ancak hastaların %28.6'sı asemptomatik idi. Ortalama kardiyopulmoner baypas süresi ve ortalama aort kros-klemp süresi, sırasıyla 105.8±33.0 ve 69.4±35.1 dakika idi. Ameliyat sonrası mekanik solunum süresi ortalama 15.0±14.9 saat, yoğun bakımda kalış süresi 1.8±0.5 gün ve hastanede kalış süresi 6.0±2.7 gün olarak bulundu. Ameliyatın tek komplikasyonu olan malign aritmilere bağlı olarak 36. saatte bir hasta kaybedildi. Takeuchi ameliyatı ile birlikte pulmoner arter rekonstrüksiyonu yapıyan yedi hastanın altısı (%85.7) ortalama 11.7±10.3 ay takip süresince semptomsuz izlendi.

Sonuç: ALCAPA cerrahi olarak düzeltilebilir bir patoloji olup, erken tanı ve tedavi uzun dönem prognoz açısından önemlidir.

Anahtar sözcükler: ALCAPA; pulmoner arter rekonstrüksiyonu; Takeuchi ameliyatı.

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Correspondence: M. Fatih Ayık, M.D. Ege Üniversitesi Tıp Fakültesi Kalp ve Damar Cerrahisi Anabilim Dalı, 35100 Bornova, İzmir, Turkey.
Tel: +90 232 - 390 40 61 e-mail: fayik35@yahoo.com

Coronary artery anomalies occur in approximately 1% of the general population,[1] and most of the cases are incidentally diagnosed during coronary angiography. Symptoms such as arrhythmias, syncope, chest pain, myocardial infarction, and sudden cardiac death are predominantly related to myocardial ischemia and dilated cardiomyopathy. Cardiomyopathies associated with coronary artery anomalies can be successfully treated via surgical interventions; hence, it is very important to diagnose the disease as early as possible. Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital coronary artery anomaly with an incidence rate of approximately 0.2-0.5% of all congenital heart disease. It usually occurs in isolation but can be associated with other congenital cardiac malformations. Bland, White, and Garland described this anomaly in 1933, and that is why ALCAPA bears the eponym Bland-White-Garland syndrome.[2-4]

Anomalous left coronary artery from the pulmonary artery presents in infancy with varying symptoms of myocardial ischemia, left ventricular dysfunction, mitral regurgitation, cardiomyopathy, and progressive heart failure. Approximately 15% of patients can survive past childhood depending on the development of collateral circulation; however, sudden cardiac death may be seen as the first clinical presentation.[4-6] Urgent surgical intervention is mandatory after the confirmation of diagnosis, and the Takeuchi operation is most often preferred.[7,8] This operation involves the creation of an aortopulmonary window and an intrapulmonary tunnel extending from the anomalous ostium to the window. In this retrospective study, we examined patients with a confirmed diagnosis of ALCAPA who underwent the Takeuchi operation combined with pulmonary artery reconstruction in the Cardiovascular Surgery department of our clinic with the goal of reporting the clinical features, surgical management, and outcomes of these patients.

PATIENTS AND METHODS

We retrospectively reviewed the charts of seven patients (2 females, 5 males; mean age 42.3±56.8 months; range 6 to 151 months) who had been diagnosed with ALCAPA and had been referred to our clinic between January 2007 and June 2010. None of them had undergone any previous surgical procedure. All of the patients had a detailed preoperative cardiovascular examination that included electrocardiography, a chest roentgenogram, echocardiography (ECG), computed tomographic (CT) coronary angiography, and cardiac catheterization. Axial CT was performed by using a 128-section multi-detector CT scanner (Somatom Definition AS-plus, Siemens, Erlangen, Germany) with the following scanning parameters: 250 mAs, 120 kV, pitch 0.2, and 0.6 mm section thickness. The scanning direction was craniocaudal, and it extended from the arcus aorta to the diaphragm during a single breath hold (Figure 1). Iodinated contrast material was injected through a catheter in the antecubital vein at a rate of 5.5 mL/sec with an automatic injector.

All of the seven patients underwent the Takeuchi operation combined with pulmonary artery reconstruction in our clinic,, and informed consent was obtained from the parents of the patients before surgery. In this procedure, a cardiopulmonary bypass (CPB) was established by aorto-bicaval cannulation. After the replacement of both the aortic root vent and the right superior pulmonary vein vent, blood cardioplegia was initiated. Under CPB, a pulmonary arteriotomy was performed, and the ostium of the left main coronary artery was observed. Then an aortopulmonary window was made, and an intrapulmonary tunnel extending from the anomalous ostium to the window was created via pericardial patch (Figures 2 and 3). At the end of the anastomosis, the pulmonary artery was enlarged using a Dacron patch. Stenosis of the left main coronary artery ostium was present in one patient (patient number 1); hence, coronary artery bypass grafting (CABG) was combined along with the surgical intervention. Ligation of patent ductus arteriosus (PDA) was performed during the surgery in another patient (patient number 4). Detailed postoperative cardiovascular examinations comprised

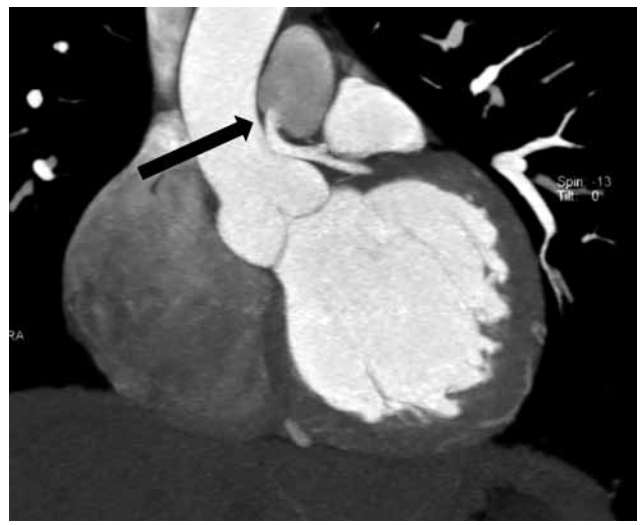

Figure 1. Preoperative coronal maximum intensity projection showing the computed tomography angiography image which revealed the left main coronary artery originating from the pulmonary artery (arrow).

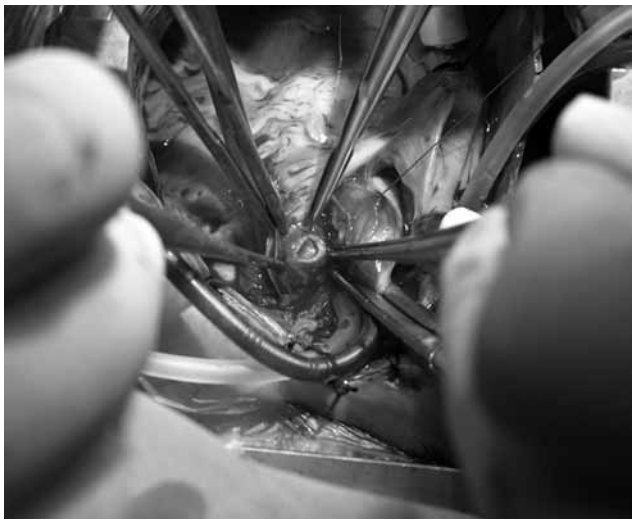

Figure 2. Creation of the aortopulmonary window.

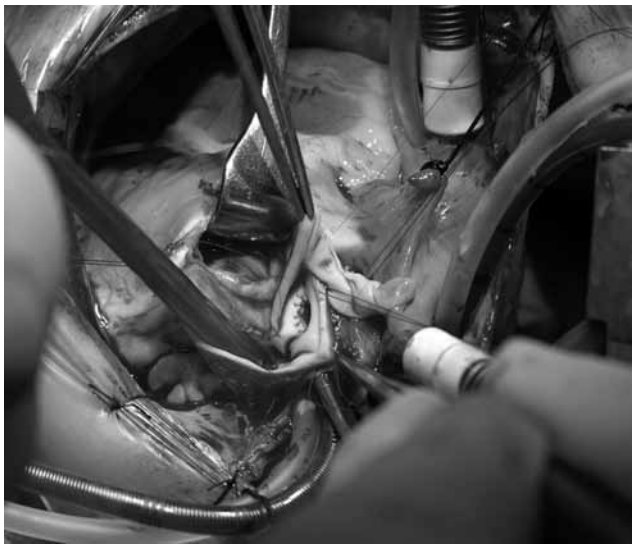

Figure 3. Creation of the intrapulmonary tunnel extending from the anomalous ostium to the window via pericardial patch.

of electrocardiography, a chest roentgenogram, ECG, and CT coronary angiography (Figure 4) were then performed and recorded for statistical analysis at postoperative six-month intervals by pediatric cardiologists.

The data was stored on a computed database and analyzed using the Statistical Package for the Social Sciences 15.0 version software program for Windows (SPSS Inc., Chicago, Illinois, USA). Wilcoxon signed rank tests were used in the statistical analysis, and a *p* value less than 0.05 was considered to be significant.

RESULTS

Symptoms at referral were periodic dyspnea in four patients (57.1%) and cardiac arrest in another (14.3%). However, two of these (28.6%) were asymptomatic. Concomitant systemic pathologies were thrombocytosis in one patient (14.3%), hydronephrosis in one (14.3%) and sepsis in another (14.3%). Five patients had ALCAPA that occurred in isolation (71.4%). One of these (14.3%) had PDA and one (14.3%) had pulmonary hypertension. Due to the lack of specificity in the clinical manifestations, five patients in our study population (71.4%) had a false initial diagnosis. The diagnoses at referral were dilated cardiomyopathy in three patients (42.9%), atrial septal defect in one (14.3%), and coronary arteriovenous fistula with mitral valve prolapse in another (14.3%). On preoperative ECG, the mean left ventricular ejection fraction (LVEF) and the left ventricular end-diastolic diameter (LVEDD) were measured as 47.3±11.3% and 36.4±6.0 mm, respectively. The preoperative mean pulmonary artery diameter (PAD) was measured as 15.7±1.0 mm by axial CT scans. The demographics of all patients are shown in Table 1.

All of the seven patients underwent the Takeuchi operation combined with pulmonary artery reconstruction. The mean CPB duration and mean aortic cross-clamp time were 95.9±31.2 and 65.0±29.8 minutes, respectively. Only one patient (14.3%), who had been diagnosed with sepsis preoperatively (patient number 5), required inotropic support. Postoperative nitrate was ordered in all patients. Postoperative durations of mechanical ventilation (hours), intensive care unit

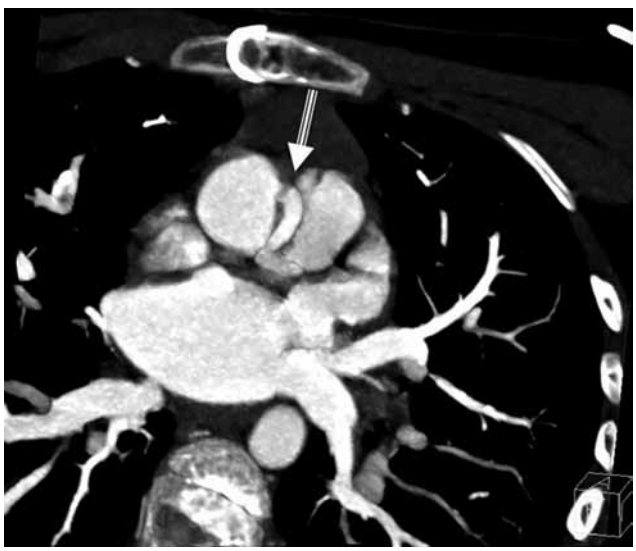

Figure 4. Axial maximum intensity projection coronary computed tomographic angiography image shows the anastomotic site between left mean coronary artery and aorta with pulmonary tunnel after Takeuchi operation (arrow).

Table 1. Demographics of the patients

Patient	Age (months) gender	Concomitant cardiac pathology	Concomitant systemic pathology	Preoperative LVEF (%)	Preoperative LVEDD (mm)	PAD (mm)	CPB time (minutes)	Cross-clamp time (minutes)
1	92/M	No	No	66	43	16	152	130
2	151/M	PHT	Hydronephrosis	55	46	17	60	39
3	12/M	No	No	44	36	16	92	64
4	6/F	PDA	Thrombocytosis	48	34	14	111	61
5	9/F	No	Sepsis	30	29	16	109	56
6	15/M	No	No	47	35	16	75	55
7	11/M	No	No	41	32	15	72	50

LVEF: Left ventricular ejection fraction; LVEDD: Left ventricular end-diastolic diameter; PAD: Pulmonary artery diameter; CPB: Cardiopulmonary bypass; Cross-clamp: Aortic cross-clamp; PHT: Pulmonary hypertension; PDA: Patent ductus arteriosus.

(ICU) stay (days), and normal ward stay (days) were 11.6±11.4, 1.4±0.5, and 5.3±0.8, respectively. Malignant arrhythmia related to hyperkalemia secondary to severe metabolic acidosis was the only complication that was identified in one patient (patient number 5) who died at the postoperative 36th hour. Sepsis and metabolic acidosis were preoperatively present in this patient owing to a prolonged pediatric ICU stay due to cardiac failure and pneumonia.

On ECG performed at the last follow-up, the mean LVEF and LVEDD were measured as 67.3±4.5% and 33.7±4.8 mm, respectively. The postoperative mean PAD was measured as 16.0±1.3 mm via axial CT scan. Six of the seven patients who underwent the Takeuchi operation combined with pulmonary artery reconstruction (85.7%) are still alive, and among these surviving patients, 100% have improved or are currently symptom-free (mean follow-up period 15.3±9.2 months). Both the improvement in LVEF and the decrease in LVEDD of the surviving patients were statistically significant (p=0.027 and p=0.027, respectively), but no statistical significance was found between pre- and postoperative PADs (p=0.157). The postoperative patient data is documented in Table 2.

DISCUSSION

Anomalous left coronary artery from the pulmonary artery is a rather rare congenital anomaly with an incidence of only 0.2-0.5% of all congenital heart disease and occurs approximately once per 300.000 live births. The embryological etiology is speculated to be abnormal septation of the conotruncus into the aorta and pulmonary artery or persistence of the pulmonary endothelial buds together with involution of the aortic buds that eventually form the coronary arteries. It usually occurs in isolation but can be associated with other congenital cardiac malformations such as patent ductus arteriosus (PDA), ventricular septal defect, tetralogy of Fallot, and coarctation of the aorta[1-4]

The equivalence of the pulmonary artery pressure and the left ventricular end-diastolic pressure secondary to PDA is the main factor for the lack of ALCAPA symptoms in prenatals. After birth, both the pressure and the resistance of the pulmonary artery start to decrease, and perfusion of the left ventricular myocardium fails. This leads to myocardial ischemia and triggers formation of collateral circulation between the two coronary artery systems. Because of the low pulmonary vascular

Table 2. Postoperative patient data

Patient	Inotropic support	Nitrate	Mechanical ventilation (hours)	ICU stay (days)	Ward stay (days)	Complication	LVEF (%)	LVEDD (mm)	PAD (mm)	Currently surviving	Follow-up (months)
1	No	Yes	6	2	6	–	72	36	16	Yes	15
2	No	Yes	3	1	6	–	60	42	17	Yes	12
3	No	Yes	8	1	6	–	64	32	16	Yes	30
4	No	Yes	15	2	5	–	68	29	14	Yes	6
5	Yes	Yes	36	2	–	Arrythmia*	–	–	–	No	–
6	No	Yes	6	1	4	–	71	33	16	Yes	7
7	No	Yes	7	1	5	–	69	30	15	Yes	22

ICU: Intensive care unit; LVEF: Left ventricular ejection fraction; LVEDD: Left ventricular end-diastolic diameter; PAD: Pulmonary artery diameter; * Malignant arrythmia related to hyperkalemia secondary to severe metabolic acidosis.

resistance, the left coronary artery flow misdirects into the pulmonary trunk, which is known as the coronary steal phenomenon. As a result of the perfusion deficit in the left ventricular myocardium, left ventricular dysfunction, mitral valve insufficiency, cardiomyopathy, and congestive heart failure symptoms may emerge in the young infant.[9] Within the first two months of life, approximately 85% of patients present with clinical symptoms of congestive heart failure, for example tachypnea, tachycardia, diaphoresis after feeding, and poor weight gain.[4,5,9,10] Electrocardiography, chest roentgenogram, ECG, and CT coronary angiography help make the differential diagnosis; however, cardiac catheterization is the gold standard for this anomaly.

Due to the nonspecific symptoms of ALCAPA, 71.4% of the patients had a false initial diagnosis in our study, and this rate was consistent with the literature.[3,11] However, Zheng et al.[12] established a correct diagnosis by ECG in most of their cases and also suggested angiography as a posssible gold standard for confirming the diagnosis. Screening the left-to-right shunting blood flow and measuring the pulmonary artery pressure via right heart catheterization can also successfully help to establish the correct diagnosis.

The treatment of choice for ALCAPA is urgent surgical intervention that mainly targets the correction of the coronary steal phenomenon. Although favorable prognosis was reported in several case reports, ligation of the left coronary artery (LCA) at its origin from the pulmonary artery or transcatheter occlusion of LCA are abandoned after high mortality rates were reported by most of the studies.[2,13-16] Transcatheter occlusion might be an alternative treatment of choice for ALCAPA patients who were planning to undergo LCA ligation surgery.[17] The most popular surgical methods create two coronary artery systems via LCA ligation plus CABG, the Takeuchi operation, or LCA reimplantation.[3,4,7,11,18-21] We preferred to perform the Takeuchi operation combined with pulmonary artery reconstruction in our study population. While the major complications of the Takeuchi operation are occlusion of the intrapulmonary tunnel and supravalvular pulmonary stenosis, such postoperative complications were not seen in our patients owing to the dilatation of the pulmonary artery using a Dacron patch.[14,22-24] The literature documents excellent prognoses after early surgical interventions in cases who did not develop postoperative ventricular tachyarrhythmia or cardiac arrest related with left ventricular dysfunction.[3,7,25-27] Although the results of surgical treatment for ALCAPA are favorable, an early diagnosis and urgent surgical intervention are crucial for long-term prognosis.

In conclusion, enlargement of the pulmonary artery in order to avoid postoperative complications of the Takeuchi procedure, such as supravalvular pulmonary artery stenosis and occlusion of the intrapulmonary tunnel, was found to be safe and effective in patients with ALCAPA. However, similar trials are needed to conclusively identify the best surgical procedure for this group of patients.

Declaration of conflicting interests

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