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ORIGINAL ARTICLE / ÖZGÜN MAKALE

Anomalous origin of left coronary artery from the pulmonary artery: Our 30 years of surgical experience and outcomes

Sol koroner arterin pulmoner arterden anormal çıkışı: 30 yıllık cerrahi deneyimimiz ve sonuçlar

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ABSTRACT

Background: The aim of this study was to evaluate our 30 years of surgical experience and outcomes in management of anomalous origin of left coronary artery from the pulmonary artery with various age groups.

Methods: Between March 1992 and August 2022, a total of 21 patients (10 males, 11 females; mean age: 16.3±15.9 years; range, 1 to 64 years) diagnosed with anomalous origin of left coronary artery from the pulmonary artery who underwent successful surgical repair were retrospectively analyzed. An initial diagnosis was made with two-dimensional echocardiography. Cardiac catheterization and angiography were performed in all our patients.

Results: The median follow-up was five (range, 1 to 14) years. The mean left ventricular ejection fraction was 43.47±14.30% with associated moderate-to-severe mitral regurgitation in seven (33.33%) patients. Coronary button transfer was performed in 15 (71.42%) patients, Takeuchi repair in four (19.04%) patients, and ligation of anomalous left main coronary artery and ligation with great saphenous venous graft in one (4.76%) patient. There was no mortality. However, two (9.52%) patients had prolonged intensive care unit stay (>7 days). At the final follow-up, the mean left ventricular ejection fraction improved to 57.47±4.97%. Regression of moderate mitral regurgitation was observed in four (66.6%) patients.

Conclusion: Preoperative left ventricular function is a major risk factor of perioperative mortality and morbidity. Mitral valve intervention is not warranted concomitantly in patients with moderate mitral regurgitation, if there are no structural lesions. Early diagnosis and meticulous surgical technique can yield excellent results and good long-term outcomes.

Keywords: Bland-White-Garland syndrome, congenital heart disease, coronary artery anomaly, echocardiography, heart failure, mitral regurgitation.

ÖZ

Amaç: Bu çalışmada çeşitli yaş gruplarında sol koroner arterin pulmoner arterden anormal çıkışının tedavisinde 30 yıllık cerrahi deneyimimiz ve sonuçlar değerlendirildi.

Çalışma planı: Mart 1992 and Ağustos 2022 tarihleri arasında sol koroner arterin pulmoner arterden anormal çıkışı tanısı konan ve başarılı cerrahi onarım yapılan toplam 21 hasta (10 erkek, 11 kadın; ort. yaş: 16.3±15.9 yıl; dağılım 1-64 yıl) retrospektif olarak incelendi. İlk tanı iki boyutlu ekokardiyografi ile kondu. Tüm hastalarımıza kardiyak kateterizasyon ve anjiyografi yapıldı.

Bulgular: Medyan takip süresi beş (dağılım, 1-14) yıl idi. Ortalama sol ventrikül ejeksiyon fraksiyonu %43.47±14.30 olup, yedi (%33.33) hastada orta ila şiddetli mitral yetersizlik ile ilişkili idi. On beş (%71.42) hastaya koroner buton transferi, dört (%19.04) hastaya Takeuchi onarımı ve bir (%4.76) hastaya anormal sol ana koroner arter ligasyonu ve büyük safen ven grefti ile ligasyon yapıldı. Mortalite izlenmedi. Ancak iki (%9.52) hastada yoğun bakım ünitesinde kalış süresi uzundu (>7 gün). Son takipte ortalama sol ventrikül ejeksiyon fraksiyonu %57.47±4.97'ye yükseldi. Dört (%66.6) hastada orta derecede mitral yetersizlikte gerileme görüldü.

Sonuç: Ameliyat öncesi sol ventrikül fonksiyonu, perioperatif mortalite ve morbidite için önemli bir risk faktörüdür. Orta derecede mitral yetersizliği olan hastalarda yapısal lezyon yoksa eş zamanlı olarak mitral kapak girişimi yapılmaz. Erken tanı ve titiz cerrahi teknik ile mükemmel sonuçlar ve uzun dönemde iyi sonuçlar elde edilebilir.

Anahtar sözcükler: Bland-White-Garland sendomu, doğuştan kalp hastalığı, koroner arter anomalisi, ekokardiyografi, kalp yetmezliği, mitral yetersizliği.

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Anomalous origin of left coronary artery from the pulmonary artery (ALCAPA) or Bland-White-Garland syndrome is a rare, but potentially life-threatening congenital coronary artery malformation. It has an incidence of 1 in 300,000 live births[1] and constitutes 0.25 to 0.5% of all congenital heart diseases.[2] Anomalous origin of left coronary artery from the pulmonary artery is one of the most common causes of myocardial ischemia/infarction in children, accounting for 90% of deaths within the first year of life, if left untreated.[3] Survival to adulthood is seen only in 10 to 15% of patients.[3] In the literature, average age of death in untreated adults with ALCAPA is 35 years.[4-6] Therefore, prompt surgical correction is recommended in all cases diagnosed with ALCAPA, irrespective of age and myocardial viability.

In the present study, we aimed to evaluate our 30 years of surgical experience and outcomes in managing ALCAPA in patients with various age groups.

PATIENTS AND METHODS

This single-center, retrospective study was conducted at Sri Sathya Sai Institute of Higher Medical Sciences, Department of Cardiothoracic and Vascular Surgery between March 1992 and August 2022. A total of 21 patients (10 males, 11 females; mean age: 16.3±15.9 years; range, 1 to 64 years) diagnosed with ALCAPA who underwent successful surgical repair were included.

An initial diagnosis was made predominantly with two-dimensional echocardiography (2D-ECHO) using M mode and Doppler flow imaging (n=19, 90.47%). Diagnosis was doubtful in two (9.52%) patients and these patients underwent contrast-enhanced computed tomography (CECT). The mean left ventricular ejection fraction (LVEF) was 43.47±14.30% (range, 20 to 62%) and out of these eight (38.09%) patients had severe LVEF (≤35%). In our study, moderate mitral regurgitation (MR) was detected in six (28.57%) patients and severe MR associated with anterior mitral leaflet prolapse with elongated chordae in one (4.76%) patient. The mean left ventricle (LV) systolic and diastolic internal dimensions were 2.88±0.64 cm and 4.31±0.64 cm, respectively (Figure 1a, b, and c).

The CECT was performed in two (9.52%) patients to identify exact coronary ostial origin and course. Due to motion artefacts (tachycardia) at the time of study, exact origin of anomalous left main coronary artery (LMCA) and left circumflex artery were poorly visualized. Therefore, we preferred cardiac catheterization and angiography for diagnostic evaluation. It was performed in all our patients to confirm the diagnosis of ALCAPA (Figure 1d). It was helpful to detect origin and course of coronary arteries, inter-coronary collaterals, and direction of blood flow in coronaries.

Surgical technique

All patients underwent surgical repair through midline sternotomy. Amongst them, cardiopulmonary bypass (CPB) with moderate hypothermia was used in 19 (90.47%) patients. During opening pericardium, the heart surface was covered with dilated and tortuous coronary arteries were noted in all cases. The anomalous LMCA originated from the left posterior aspect of the main pulmonary artery (MPA) in all our cases. For better myocardial protection, the cannulation and cardioplegia technique was as follows: *(i)* aortic cannulation; *(ii)* superior vena cava cannulation; *(iii)* going on pump with single venous cannula; *(iv)* inferior vena cava cannulation; *(v)* CPB with two venous cannulas; *(vi)* cardioplegia cannulation over aorta proximal to aortic cannula; *(vii)* LV vent over left upper superior pulmonary vein; *(viii)* cross-clamping aorta; *(ix)* delivering cold blood Del Nido cardioplegia solution (20 mL/kg) at 4°C after clamping MPA/right pulmonary artery (RPA) and left pulmonary artery (LPA) based on the type of repair; and *(x)* topical ice slush was put to cool the heart further.

Coronary button transfer (n=15, 71.42%)

After establishing CPB, aorta and MPA were clamped separately. Aortic root antegrade cold blood cardioplegia was given. The LV was vented through right superior pulmonary vein. The LMCA was mobilized and dissected from MPA along with collar of native MPA tissue. A hole was made on left lateral aspect of aorta with a 4-mm punch. The LMCA button was anastomosed to lateral aspect of aorta with 7-0 polypropylene suture in a continuous fashion. The defect in MPA was augmented with autologous pericardial patch using 5-0 polypropylene suture (Figure 2).

Takeuchi repair (n=4, 19.04%)

After establishing CPB, LPAs and RPAs were clamped and snared separately. Aorta was cross-clamped and root antegrade cold blood cardioplegia was given. After satisfactory arrest, aorta and MPA were opened for assessment of anatomy. A hole was punched on left lateral wall of aorta corresponding to another made on right lateral wall of MPA. They were anastomosed to form an aortopulmonary (AP) window. The MPA was cut transversely above and below the level of AP window

creating a MPA flap. This flap was sutured all along posterior wall of MPA creating a MPA flap tunnel. It covers the opening of ALCAPA and AP window, thereby diverting aortic blood into ALCAPA. Aorta was closed and root cardioplegia was given to see satisfactory bulge of MPA tunnel and to confirm suture-line hemostasis. Residual defect in MPA was repaired with supra-annular autologous pericardial patch (Figure 3).

Ligation of ALCAPA followed by aorta-LMCA great saphenous venous graft (n=1, 4.76%)

In one (4.76%) patient, direct ligation of ALCAPA was done. After that, we performed anastomosis of reverse great saphenous venous graft between aorta (end to side) and anomalous LMCA end to end.

Ligation of ALCAPA (n=1, 4.76%)

Along with ALCAPA correction, mitral valve (MV) repair (chordal shortening with bilateral commissural plication) was done for severe MR in one (4.76%) patient. However, MV was not addressed in cases with moderate MR in our study. Along with ALCAPA correction, patent ductus arteriosus (PDA) was ligated in one (4.76%) patient. The mean CPB time was 139.05±24.79 (range, 82 to 191) min and the mean aortic cross-clamp time was 92±18.92 (range, 56 to 129) min. Perioperative and follow-up data of our patients are illustrated in Table 1.

Data collection

Medical records of the patients were reviewed to evaluate clinical presentation, pathophysiological findings, imaging and surgical treatment with postoperative outcomes and follow-up data.

Follow-up

The median follow-up was five (range, 1 to 14) years. The patients were followed at our outpatient clinic

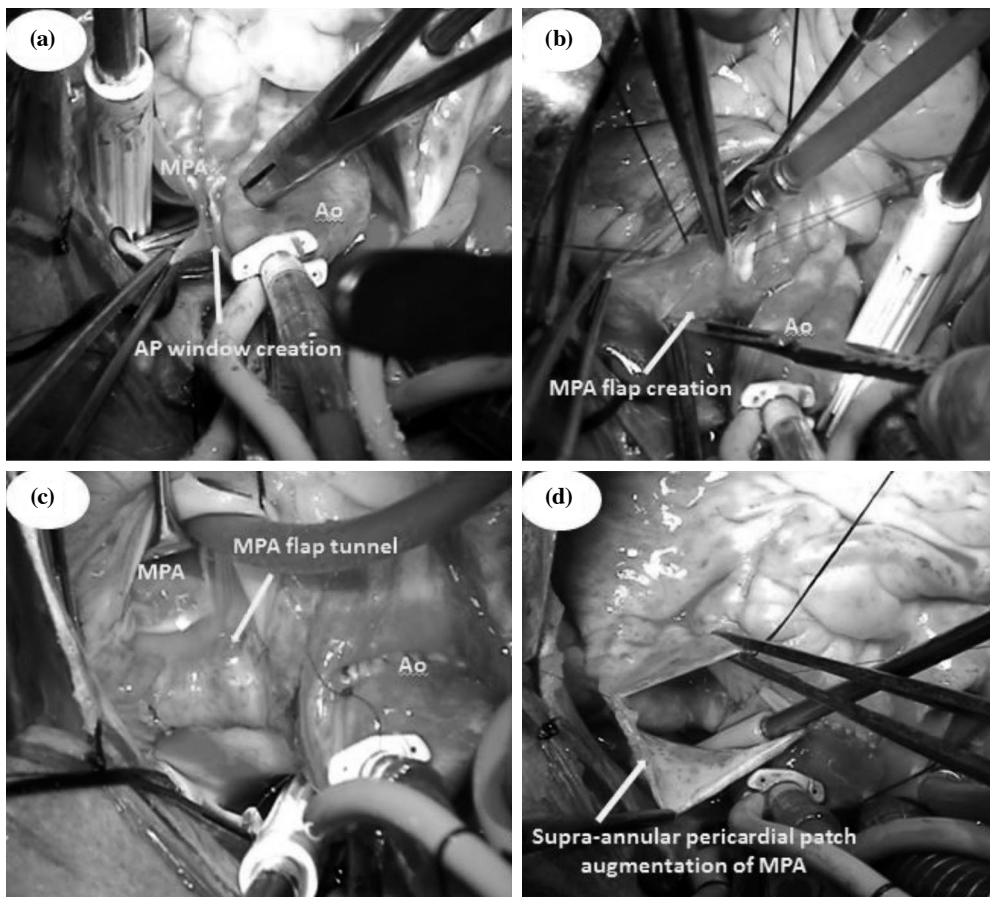

Figure 1. 2D ECHO and cardiac catheterization: **(a)** Modified 5-chambered view showing LMCA origin from MPA; **(b)** Parasternal short axis view showing retrograde flow of LMCA to MPA; **(c)** Apical four-chamber view showing dilated LV; **(d)** RCA shoot showing origin of LMCA from MPA with inter-coronary collaterals.

MPA: Main pulmonary artery; Ao: Aorta; AP: Aortopulmonary; 2D ECHO: Two-dimensional echocardiography; LMCA: Left main coronary artery; LV: Left ventricle; RCA: Right coronary artery.

and telephonic calls were made for data collection, if necessary. All patients underwent electrocardiogram (ECG) and 2D-ECHO at every follow-up visit.

Statistical analysis

Statistical analysis was performed using the IBM SPSS version 22.0 software (IBM Corp., Armonk, NY, USA). Descriptive data were expressed in mean ± standard deviation (SD), median (min-max) or number and frequency, where applicable. The Fisher exact test and two-sample t-test were used to calculate *p* value between two groups (Group 1: LVEF ≤35%, n=8 *vs*. Group 2: LVEF >35%, n=13). A *p* value of <0.05 was considered statistically significant.

RESULTS

Demographic and preoperative characteristics of the patients are shown in Table 2.

Early results (<30 days)

There was no early mortality in our study. In the majority of the patients (90.47%), postoperative period was uneventful. However, two (9.52%) patients had prolonged intensive care unit (ICU) stay (>7 days). The first case was a four-year and one-month female with query dilated ischemic cardiomyopathy admitted in cardiac care unit. On further evaluation, she was diagnosed with ALCAPA with moderate MR. Based on a low LVEF (20%), the patient was stabilized with dobutamine (5 µg/kg/min) injection for six days and underwent corrective surgery (direct re-implantation). Postoperatively, the patient was electively ventilated for five days due to poor cardiopulmonary reserve (LVEF: 15%, low partial pressure of oxygen [PO_2] and high partial pressure of carbon dioxide [PCO_2]). Trial of extubation was given on postoperative Day 5 (POD) and the patient was re-intubated due to tachypnea with high PCO_2 on the same day. Then, she was gradually weaned and extubated on POD 10. Non-invasive continuous positive airway pressure mode of ventilation was continued for two more days. On POD 12, the patient was re-intubated again due to further worsening with thick airway secretions and high PCO_2. She was gradually improved and

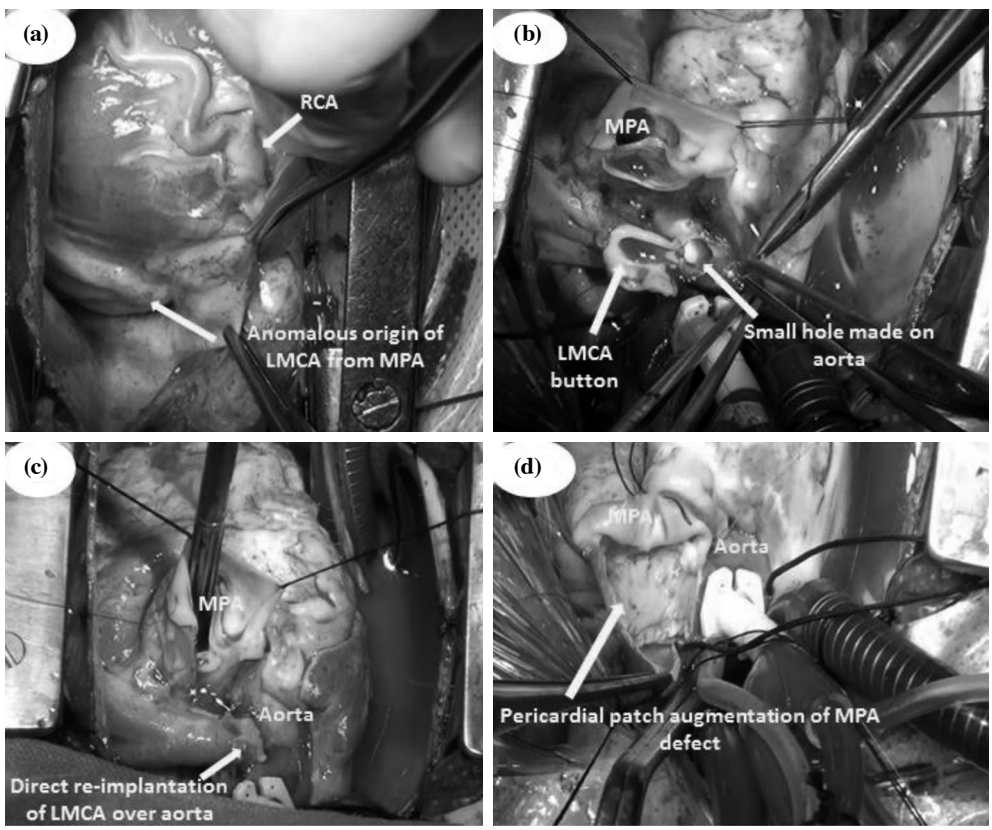

Figure 2. (a) Anomalous origin of LMCA from MPA with dilated and tortuous RCA; **(b)** Mobilization of anomalous LMCA button; **(c)** Anastomosis of anomalous LMCA button with aorta; **(d)** Pericardial patch augmentation of MPA defect.

RCA: Right coronary artery; MPA: Main pulmonary artery; LMCA: Left main coronary artery.

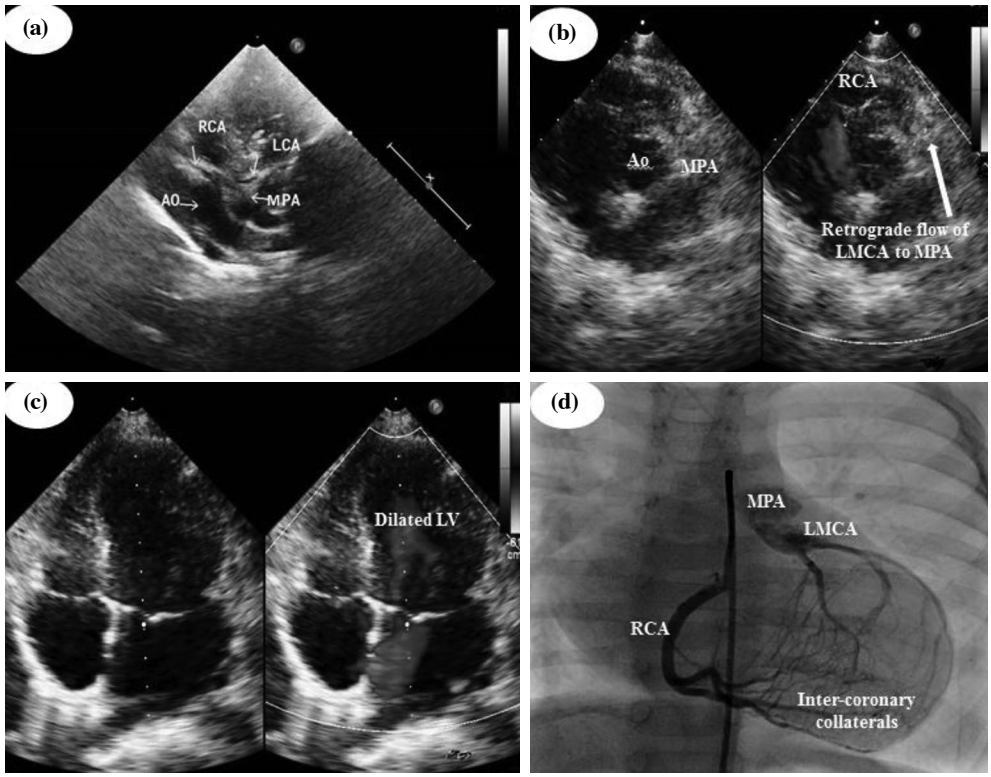

Figure 3. **(a)** Aortopulmonary window creation. **(b)** MPA dissection with flap creation. **(c)** Creation of MPA flap tunnel along posterior wall of MPA. **(d)** Supra-annular autologous pericardial patch augmentation of MPA.

RCA: Right coronary artery; LCA: Left coronary artery; Ao: Aorta; MPA: Main pulmonary artery; LMCA: Left main coronary artery.

extubated on POD 16. The Patient was clinically asymptomatic and, thus, discharged on POD 28 with moderate MR and LVEF of 15%. At three months of follow-up, LVEF improved to 35% and at one-year follow-up, it significantly improved to 55%. Based on the improvement in LVEF, moderate MR regressed to mild MR at three months of follow-up and to trivial MR at one year of follow-up.

The second case was a one-year male diagnosed with ALCAPA with moderate MR and low LVEF (35%). He underwent direct re-implantation. Postoperatively, he required prolonged ventilator and inotropic support due to low LVEF (30%) and low PO_2. He was weaned off ventilator and inotropic support gradually and extubated on POD 8. During follow-up, LVEF improved to 45% at three months and 55% at five years. Unlike the first case, moderate MR persisted even at five years of follow-up, but he was clinically asymptomatic with preserved LV dimensions.

The mean duration of mechanical ventilation was 19.42±30.56 (range, 4 to 120) h. The mean length of ICU stay was 5.28±4.7 (range, 2 to 24) days. The mean length of hospital stay was 7.52±5.28 (range, 4 to 28) days. All patients were clinically asymptomatic at the time of discharge.

Late results (>30 days)

No late mortality was detected in our study during follow-up. There was an improvement of LVEF during follow-up in all patients. At the last follow-up, the mean LVEF improved to 57.47±4.97% (range, 45 to 62%) compared to the mean preoperative LVEF of 43.47±14.30% (range, 20 to 62%). Regression of moderate MR to mild MR was observed in four patients (66.6%). However, moderate MR persisted in one (16.6%) patient and it progressed to severe MR in another patient, but both patients were clinically asymptomatic at their last follow-up with preserved LV dimensions.

Perioperative clinical variables and outcomes were compared between the two groups (Group 1: LVEF ≤35%, n=8 ***vs.*** Group 2: LVEF >35%, n=13) (Table 3).

Table 1. Perioperative and follow-up data of patients

Case No.	Surgery performed	CPB time (min)	Aortic cross clamp time (min)	Ventilator time (h)	Inotropic supports used (h)	ICU stay (Days)	Hospital stay (Days)	2D ECHO at the time of hospital discharge		2D ECHO at last follow-up		Last follow-up (years)
								LVEF (%)	MR	LVEF (%)	MR	
1	Coronary button transfer	135	95	6	Dobutamine (66)	4	6	45	Mild	60	Mild	11
2	Takeuchi repair	130	90	5	Dobutamine (56)	6	9	40	Trivial	45	Trivial	1
3	Ligation + venous graft to LMCA	124	100	6	Dopamine (73)	5	6	55	Mild	60	Mild	12
4	Ligation	-	-	5	Dopamine (34)	4	6	45	Moderate	45	Mild	10
5	Takeuchi repair	154	102	6	Dobutamine (35)	4	7	55	Trivial	60	Trivial	3
6	Coronary button transfer	135	98	7	Dopamine (25)	4	5	55	Mild	60	Mild	6
7	Coronary button transfer	147	100	9	Dopamine (26)	7	10	35	Moderate	60	Mild	14
8	Takeuchi repair	170	120	5	Dopamine (55)	5	7	60	Mild	60	Mild	7
9	Takeuchi repair	120	86	4	Dopamine (35)	3	5	55	Mild	60	Mild	14
10	Coronary button transfer	118	68	23	Dobutamine (96)	5	8	35	Moderate	55	Mild	5
11	Coronary button transfer	82	63	4	Dobutamine (48)	3	5	55	Trivial	60	Trivial	6
12	Coronary button transfer	122	90	5	Dopamine (43)	4	6	60	Mild	60	Mild	7
13	Coronary button transfer	120	93	7	No supports (0)	2	4	55	Mild	62	Mild	3
14	Coronary button transfer	155	98	8	Dobutamine (28)	2	4	60	Trivial	60	Mild	3
15	Coronary button transfer	161	90	56	Milrinone (24) and Dobutamine (26)	7	9	25	Mild	60	Mild	5
16	Coronary button transfer	122	56	86	Milrinone (160) and Dobutamine (168)	10	14	30	Moderate	55	Moderate	5
17	Coronary button transfer + PDA ligation	125	84	21	Milrinone (45)	4	6	25	Moderate	60	Severe	6
18	Coronary button transfer	138	66	6	Dopamine (46)	2	4	55	Trivial	60	Mild	5
19	Coronary button transfer	168	121	7	Dopamine (23)	3	4	55	Mild	60	Mild	1
20	Coronary button transfer	191	129	120	Milrinone (168) and Adrenaline (150)	24	28	15	Moderate	55	Mild	1
21	Coronary button transfer	164	91	12	Milrinone (36)	3	5	60	Trivial	60	Trivial	1

2D ECHO: Two-dimensional echocardiography; CPB: Cardiopulmonary bypass; ICU: Intensive care unit; LVEF: Left ventricular ejection fraction; MR: Mitral regurgitation.

Table 2. Demographic and preoperative characteristics of patients

Case No.	Age/sex (year)	Weight (kg)	Height (cm)	Year of surgery	Complaints		Chest X-ray (CTR)	ECG		Preoperative 2D ECHO			CATH study
					DOE (NYHA class)	Other complaints		Rhythm	ALMI	LVEF (%)	MR	LVid (S/D) (cm)	
1	37/M	55	150	1992	II	-	0.55	SR	No	45	Mild	4.3/2.8	Yes
2	11/M	30	125	1994	II	AOE-II	0.62	SR	Yes	20	Trivial	4/2.4	Yes
3	64/M	66	176	1995	II	AOE-II	0.57	SR	Yes	55	Mild	4.8/3	Yes
4	26/M	52	140	1996	III	AOE-II	0.56	SR	Yes	30	Moderate	5.5/4.2	Yes
5	35/F	48	143	1998	II	-	0.55	SR	Yes	50	Trivial	4.2/2.8	Yes
6	19/F	55	157	1998	II	-	0.56	SR	Yes	48	Mild	3.8/2.9	Yes
7	4.4/M	14	100	2000	II	RRTI	0.55	SR	Yes	30	Moderate	3.4/2.8	Yes
8	8/M	18	122	2005	II	RRTI	0.6	SR	Yes	55	Severe (AML prolapse	5.5/3.1	Yes
9	24/F	46	150	2006	II	-	0.55	SR	Yes	50	Mild	4.1/2.4	Yes
10	2/F	10	80	2007	III	RRTI and feeding difficulty	0.53	SR	Yes	30	Moderate	5.9/4.6	Yes
11	7/F	18	115	2009	II	RRTI	0.54	SR	Yes	62	Trivial	4.3/2.5	Yes
12	14/M	52	164	2009	II	-	0.55	SR	No	50	Mild	4.1/2.5	Yes
13	37/M	75	166	2012	II	AOE-II	0.45	SR	No	55	Mild	4.2/2.9	Yes
14	13/F	49	157	2012	II	-	0.51	Sinus tachycardia	Yes	60	Trivial	4.1/2.2	Yes
15	2.3/M	12	91	2013	II	RRTI	0.81	SR	Yes	20	Mild	4/2	Yes
16	1/M	10	78	2014	II	RRTI and feeding difficulty	0.54	SR	Yes	35	Moderate	4.4/3.1	Yes
17	2.1/F	8.2	80	2016	II	Palpitations	0.65	SR	Yes	20	Moderate	3.7/3	Yes
18	7.2/F	18	121	2016	II	RRTI	0.55	SR	No	58	Mild	3.9/2.6	Yes
19	8.2/F	14.4	105	2017	II	RRTI	0.52	SR	No	55	Trivial	3.6/2	Yes
20	4.1/F	4.7	59	2018	III	Feeding difficulty	0.65	SR	Yes	30	Moderate	4.8/3.6	Yes
21	17/F	38.8	145	2018	II	-	0.55	SR	No	55	Mild	4.1/3.2	Yes

ECG: Electrocardiography; 2D ECHO: Two-dimensional echocardiography; DOE: Dyspnea on exertion; NYHA: New York Heart Association; CTR: Cardiothoracic ratio; ALMI: Anterolateral wall myocardial ischemia / infarction; LVEF: Left ventricular ejection fraction; MR: Mitral regurgitation; LVid: Left ventricular internal dimensions; S/D; Systolic/diastolic; CATH: Cardiac catheterization; SR: Sinus rhythm; AOE: Angina on exertion; RRTI: Recurrent respiratory tract infections.

Table 3. Comparison of perioperative clinical variables and outcomes between two groups with different LVEF values

	Group 1 (LVEF <35%) (n=8)		Group 2 (LVEF >35%) (n=13)		
Clinical variables	%	Mean±SD	%	Mean±SD	*p*
Age in years		6.6±8.4		22.3±16.8	0.02*
Weight (kg)		17.6±15.8		42.6±19.8	0.007*
Height (cm)		94.1±26.7		143.9±21.8	<0.001*
DOE-NYHA Class-III	37.5		0		0.04*
Cardiothoracic ratio		0.6±0.1		0.5±0.0	0.02*
ALMI	100		53.8		0.04*
LVEF		26.9±5.9		53.7±4.9	<0.001*
Moderate MR	75.0		0		0.001*
CPB time (min)		142±26.5		137.5±24.9	0.71 (NS)
Aortic cross clamp time (min)		88.1±23.4		94.1±16.8	0.52 (NS)
Ventilator support (h)		40.6±42.8		6.4±2.1	0.009*
Inotropic support (h)		77.8±60.3		39.5±19.2	0.04*
ICU stay (days)		8.4±6.6		3.4±1.0	0.01*
Hospital stay (days)		11.3±7.2		5.2±1.1	0.007*
LVEF at discharge		31.3±9.5		55.8±4.0	<0.001*
Moderate MR at discharge	83.33		0		0.001*
LVEF at last follow-up		54.4±6.2		59.4±2.9	0.02*
Moderate MR at last follow-up	33.33		0		<0.03*

LVEF: Left ventricular ejection fraction; SD: Standard deviation; DOE: Dyspnea on exertion; NYHA: New York Heart Association; ALMI: Anterolateral wall myocardial ischemia/infarction; MR: Mitral Regurgitation; CPB: Cardiopulmonary bypass; ICU: Intensive care unit.

DISCUSSION

In fetal and early neonatal life, pulmonary artery (PA) pressure is equal to aortic pressure and it allows enough myocardial perfusion through the anomalous LMCA arising from PA.[7] Soon after birth, the PA resistance decreases below systemic arterial pressure. Therefore, oxygenated blood from the right coronary artery becomes shunted to PA system via collaterals and anomalous LMCA causing coronary artery steal phenomenon.[8] This phenomenon affects the physiological coronary blood flow, thereby leading to myocardial ischemia/infarction worsened during physiological activities such as feeding and crying.[7]

Based on collateral circulation, ALCAPA is classified into two main groups as the infantile group with little or no collateral circulation and the adult group with extensive inter coronary collateral circulation. The manifestations and outcomes of these two groups are completely different.[9] In the infantile group, diagnosis of ALCAPA is possible within the first year of life, when infants present with irritability, fatigue, inability to feed, sweating and tachypnea. Due to extensive inter-coronary collateral circulation, the adult group patients are asymptomatic and, thus, diagnosis is usually delayed. However, these patients may suffer from MR due to anterolateral papillary muscle ischemia/infarction and sudden cardiac death in the future.[10] Bland et al.[11] and Boutsikou et al.[12] reported that survival of adult group mainly depended upon right coronary artery dominance with extensive inter-coronary collaterals and restrictive opening between ALCAPA and PA. In our study, most of the patients had late presentation.

Anomalous origin of left coronary artery from the pulmonary artery is often an isolated anomaly, but rarely associated with anomalies such as PDA, ventricular septal defect, tetralogy of Fallot, pulmonary atresia, hemi-truncus and coarctation of aorta.[7] In our

study, only one (4.96%) patient had PDA associated with ALCAPA.

The vast majority of patients have cardiomegaly on chest X ray. Electrocardiography may yield normal results or show features suggestive of anterolateral myocardial ischemia/infarction (ALWMI) (i.e., inverted T wave in leads V4 to V6 and pathological Q waves in lead I and aVL).[13,14] In our study, 15 patients (71.42%) had ALWMI, consistent with the literature.[13,15] The 2D ECHO also plays a major role in the diagnosis of ALCAPA.[16] It is useful to detect anomalous origin of coronary arteries, enlargement of left heart chambers, decreased LV systolic function, quantification of MR, aneurysmatic expansion of coronary arteries, and retrograde diastolic blood flow from coronary artery to PA.[16] Consistent with the literature, the majority of our patients (90.47%) were diagnosed with ALCAPA using 2D ECHO in our study. All patients underwent cardiac catheterization with coronary angiography to confirm the diagnosis and to delineate coronary anatomy and inter-coronary collaterals. The CECT and cardiac magnetic resonance imaging are also useful tools for diagnostic examinations.[17] However, our experience with CECT, cardiac magnetic resonance imaging, and thallium perfusion scan in the management of ALCAPA is limited.

Furthermore, it is necessary to rule out dilated cardiomyopathy/endocardial fibroelastosis before diagnosis of ALCAPA.[18] Medical management of ALCAPA is associated with high morbidity and mortality.[19] Therefore, it is recommended to tailor surgery as soon as diagnosis is confirmed to prevent further progression of myocardial ischemia/infarctioh.[20]

Although several surgical methods have been developed to date, establishment of dual-coronary system has become the standard surgical approach for repair. This has a positive survival effect in all age groups. Currently, Takeuchi repair and direct re-implantation of coronary artery to ascending aorta are two commonly used surgical options.[20] In Takeuchi repair, creation of AP window is followed by redirecting blood from ascending aorta to left coronary ostia via an intra-pulmonary tunnel. This surgical technique is associated with intraoperative coronary insufficiency, supravalvular pulmonary stenosis, coronary-PA fistula, and pulmonary regurgitation.[21] However, none of these complications were encountered and four (19.04%) patients underwent Takeuchi repair in our study. Consistent with the recent literature, Takeuchi repair is preferred over coronary re-implantation in cases where coronaries are difficult to mobilize. In 1974, Neches et al.[22] described re-implantation of LMCA to ascending aorta. This technique can increase survival and decrease coronary stenosis and re-operation rate.[23,24] In our study, 15 (71.42%) patients underwent direct re-implantation of LMCA to ascending aorta, as the anatomy was conducive and associated with fewer complications. The mean age of the patients who underwent Takeuchi repair and direct re-implantation of LMCA to ascending aorta was 19.5±12.4 years and 11.6±11.7 years, respectively in our study. We preferred Takeuchi repair/ligation/ligation with reverse great saphenous vein graft in patients where anomalous LMCA was unable to mobilize and direct re-implantation of LMCA was preferred in patients where anomalous LMCA was able to mobilize.

Coronary ischemia can cause subendocardial infarcts and ischemia of anterolateral papillary muscle. This, in turn, causes MR. The need for MV intervention in cases with moderate MR during initial ALCAPA repair is still controversial. However, consistent with the literature,[13,14,24] there was an improvement of MR and LV function after establishment of physiological dual coronary circulation without MV repair, regardless of age in our study. However, in some patients, MR can persist postoperatively due to irrepressible ischemic damage to papillary muscle and may require additional intervention. In our study, moderate-to-severe MR was observed in seven (33.33%) patients preoperatively, out of which only one patient with severe MR underwent MV repair (anterior mitral leaflet chordal shortening with commissural plication). No additional intervention was done to the remaining six patients with moderate MR without any structural abnormality. During follow-up, MR improved in four (66.66%) patients over a short period of time (range, 6 months to 2 years) after the establishment of dual physiological coronary circulation. In the remaining two (33.33%) patients, moderate MR persisted in one and progressed to severe MR in the other. Surgical intervention was not warranted, as they were clinically asymptomatic with preserved LV dimensions.

In general, operative mortality of ALCAPA with dual coronary repair ranged from 0 to 16%.[25] Left ventricular dysfunction is a major risk factor for perioperative mortality and morbidity.[4] However, in our study, no mortality was observed, although 38.09% of the patients presented with LVEF ≤35%. The LV function recovery may vary according to the efficiency of surgical repair and medical treatment. It may take several months, even up to one year after repair.[25]

Our patients (n=8) with a preoperative LVEF of ≤35% reached normal range over a span ranging from 0.5 to 15 months postoperatively.

The role of extracorporeal membrane oxygenation (ECMO) after ALCAPA repair is still under debate. Proponents of ECMO advocate its advantage in cases where there is unstable hemodynamics while weaning from the CPB machine. However, some authors are against ECMO use due to the fact that it worsens cardiac ischemia by decompressing the PA and increasing steal from right coronary system, thereby compromising the chances of recovery. Also, pediatric cardiac assist devices still carry a high risk for mortality and morbidity. Our knowledge with cardiac assist devices is still limited, as all our cases were weaned from CPB smoothly. As a result, the need for cardiac assistance can be case-dependent and it can be avoided in most cases with successful perioperative myocardial support strategies.

The single-center, retrospective design of the study with a relatively small sample size is the main limitation.

In conclusion, anomalous origin of left coronary artery from the pulmonary artery should be suspected in patients with dilated cardiomyopathy and/or with unexplained mitral regurgitation and electrocardiographic changes suggestive of anterolateral myocardial ischemia/infarction. Two-dimensional echocardiography with color flow Doppler is helpful to establish the early diagnosis. However, cardiac catheterization with angiography is the gold-standard diagnostic modality. An aggressive surgical approach is warranted, irrespective of age to halt the progress of myocardial ischemia, ventricular arrhythmias, and sudden death. Among several surgical techniques for anomalous origin of left coronary artery from the pulmonary artery repair, re-implantation of anomalous coronary artery to establish dual coronary circulation is the most preferred technique. We suggest that mitral valve intervention is not warranted concomitantly in patients with moderate mitral regurgitation, if there are no structural lesions. Preoperative left ventricular dysfunction is a major risk factor of perioperative mortality and morbidity.

Ethics Committee Approval: Ethical approval was waived off being a retrospective study of a standard surgery. The study was conducted in accordance with the principles of the Declaration of Helsinki.

Patient Consent for Publication: A written informed consent was obtained from each patient and/or their legal parents and guardians.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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