

A case series of left main coronary artery ostial atresia and a review of the literature

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Abstract

Left main coronary artery ostial atresia (LMCAOA) is a rare congenital anomaly of the coronary arteries. The published literature regarding the current diagnostic and management recommendations are limited. We present three case series of LMCAOA from our institution, including one with a unique association with anomalous origin of left coronary artery (LCA) from pulmonary artery. In addition, this report includes a review of 50 pediatric and 43 adult cases from literature. The majority of the patients were symptomatic. Sudden cardiac death occurred in 10% of pediatric patients and 7% of adult patients. Almost half of pediatric patients had additional cardiac lesions. At the time of diagnosis, 82% of patients had abnormal exercise stress test and 73% had abnormal myocardial perfusion imaging (MPI). The diagnosis of LMCAOA was suspected by echocardiography in 47% of pediatric patients, while 26% were initially misdiagnosed as anomalous origin of LCA from pulmonary artery. Coronary angiography confirmed the diagnosis in most cases and 70.5% of pediatric patients had small collaterals, while 80.5% of adult patients had large collaterals. Nine pediatric patients had no revascularization surgery with five deaths. Revascularization surgery was performed in 39 pediatric patients with four deaths. After 2005, there is a gradual shift toward performing coronary osteoplasty rather than coronary artery bypass grafting. Eighteen adult patients had revascularization surgery and all survived. Fifteen adult patients had no revascularization surgery, of which there were five deaths. In patients with LMCAOA, revascularization surgery is currently recommended in the presence of symptoms, ischemic changes on electrocardiogram or exercise stress test, myocardial perfusion defect on MPI, global left ventricular systolic dysfunction on echocardiogram, severe mitral regurgitation, or small-sized collaterals in coronary angiography. Short-term and mid-term outcomes are encouraging.

KEYWORDS

artery, atresia, coronary, left

1 | INTRODUCTION

Left main coronary artery ostial atresia (LMCAOA) is an extremely rare congenital coronary artery anomaly with an incidence of 0.1%–0.4% of “sole coronary” cases diagnosed on the basis of results of necropsy studies.¹ It is defined as the absence of a left main ostium with

the left anterior descending (LAD) and circumflex arteries (Cxs) connected as usual and ending blindly, having not originated from another vessel or cardiac chamber, and supplied by the right coronary artery (RCA) via collaterals.² The left main coronary artery (LMCA) trunk may be normal, hypoplastic, or atretic. The clinical presentation is non-specific and widely variable depending on the degree of collateral

circulation and myocardial perfusion. Some patients present early in life, mostly with congestive heart failure and syncope, whereas others remain asymptomatic into late adulthood, before presenting mostly with angina.³ Sudden cardiac death (SCD) was reported as the presenting symptom in some cases.³ Surgical revascularization either by coronary artery bypass grafting (CABG) or coronary reconstruction osteoplasty was performed in many cases in literature.^{3,4} Herein, we describe three cases of LMCAOA from our institution with variable clinical presentations, diagnostic investigations, and management strategies. In addition, a detailed account of the literature to date summarizing the cases of LMCAOA is also reported to help achieve a better understanding for this complex and rare cardiac lesion.

2 | CASE 1

A full-term 6-week-old female had multiple rhabdomyomas detected by a fetal echocardiogram. The brain magnetic resonance imaging

(MRI) was normal. At 2 weeks of age, her initial transthoracic echocardiogram (TTE) at referring hospital did not show any rhabdomyomas, however, it was suggestive for anomalous origin of left coronary artery (LCA) from right facing sinus. She was referred to our center for assessment at 7 weeks of age. At that time, she was growing well and had no cardiovascular-related symptoms. The resting electrocardiogram (ECG) was unremarkable. Repeated TTE revealed a suspected LMCAOA. The RCA was dilated (z-score 2.25) with normal ostial origin and antegrade flow (Figure 1A). The LAD and the Cx were both small and supplied retrograde from prominent collaterals arising anteriorly from the RCA. The ostial origin of the LMCA from the aorta was not seen (Figure 1B). There was no obvious anomalous origin of the LCA from the pulmonary artery (ALCAPA) and there was good biventricular systolic function with no regional wall motion abnormalities or mitral regurgitation (MR). The patient went on to have selective coronary angiography to confirm the diagnosis. Injection into the left coronary ostium demonstrated no antegrade flow (Figure 1C). Selective right

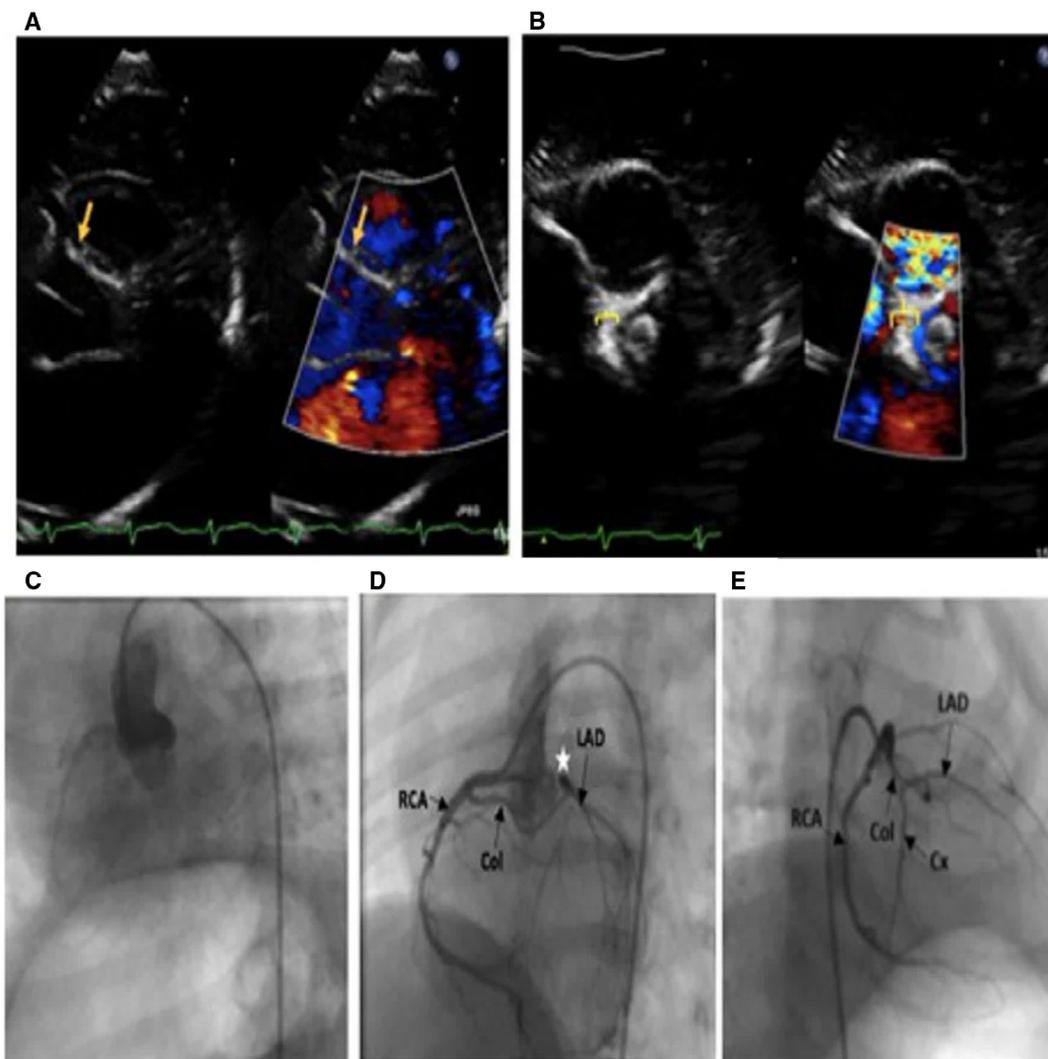


FIGURE 1 A, Prominent collateral branch (yellow arrow) arising from the dilated proximal RCA. B, The LMCA ostial origin is atretic with close proximity to aorta. Retrograde flow in the proximal portion of the LAD and antegrade flow in the Cx. C, Selective injection into the left coronary ostium showing no antegrade flow into the LMCA. D & E, Selective right coronary angiography showing a mildly dilated RCA, with a good-sized collateral artery arising from RCA as a first branch and connects to the proximal LAD. Both LAD and Cx forms a blind-ended pouch with close proximity to aorta (star). Cx, circumflex artery; LAD, left anterior descending artery; LMCA, left main coronary artery; RCA, right coronary artery

coronary angiography demonstrated an atretic ostial origin of LMCA with close proximity to the aortic sinus (Figure 1D,E). The RCA was dilated with a normal ostial origin and antegrade flow. The first branch arising from the RCA was a prominent large collateral artery that connected the proximal portion of the LAD (Figure 1D,E). The magnetic resonance myocardial perfusion imaging (MPI) was normal with no perfusion defects on either the rest or stress images and no evidence of scar as assessed by late gadolinium enhancement. As the patient was asymptomatic in the setting of normal MPI and a good-sized collateral supply of the left coronary system from RCA, no intervention was undertaken with routine follow up in the ambulatory clinic. At the last visit, she was over 1 year old with no concerns.

3 | CASE 2

A 5-years-old boy presented with a history of multiple episodes of exertional syncope and chest pain since 2 years of age. Cardiopulmonary resuscitation had been performed on multiple occasions. His initial TTE at the referring hospital was unremarkable except for near normal left ventricular systolic function and a dilated RCA. He had been

previously referred to neurology for the possibility of a seizure disorder and had a normal brain MRI and electroencephalogram. The resting ECG was unremarkable. On assessment at our center, TTE revealed a suspected LMCAOA. The RCA was prominent with normal ostial origin and antegrade flow. The ostial origin of the LMCA from the aorta was not seen with no antegrade flow (Figure 2A). There was retrograde flow in the LMCA and LAD. There were multiple prominent septal perforators from the posterior descending artery (PDA) (Figure 2B,C). There was no obvious ALCAPA. The left ventricle (LV) was mildly dilated. The biventricular systolic function was preserved with no regional wall motion abnormalities or MR. The patient went on to have selective coronary angiography to confirm the diagnosis. Injection into the left coronary ostium demonstrated no antegrade flow (Figure 2D). Selective right coronary angiography demonstrated a dilated RCA with a normal ostial origin and antegrade flow. There was retrograde filling of the entire left coronary system by multiple small septal perforators from the PDA which flowed into the distal LAD (Figure 2E,F). As the patient was symptomatic and in the setting of small collaterals, he underwent re-implantation of the LMCA to the aorta with left coronary osteoplasty. There was an atretic ostium of the LMCA within the left-right aortic commissure (Figure 3A).

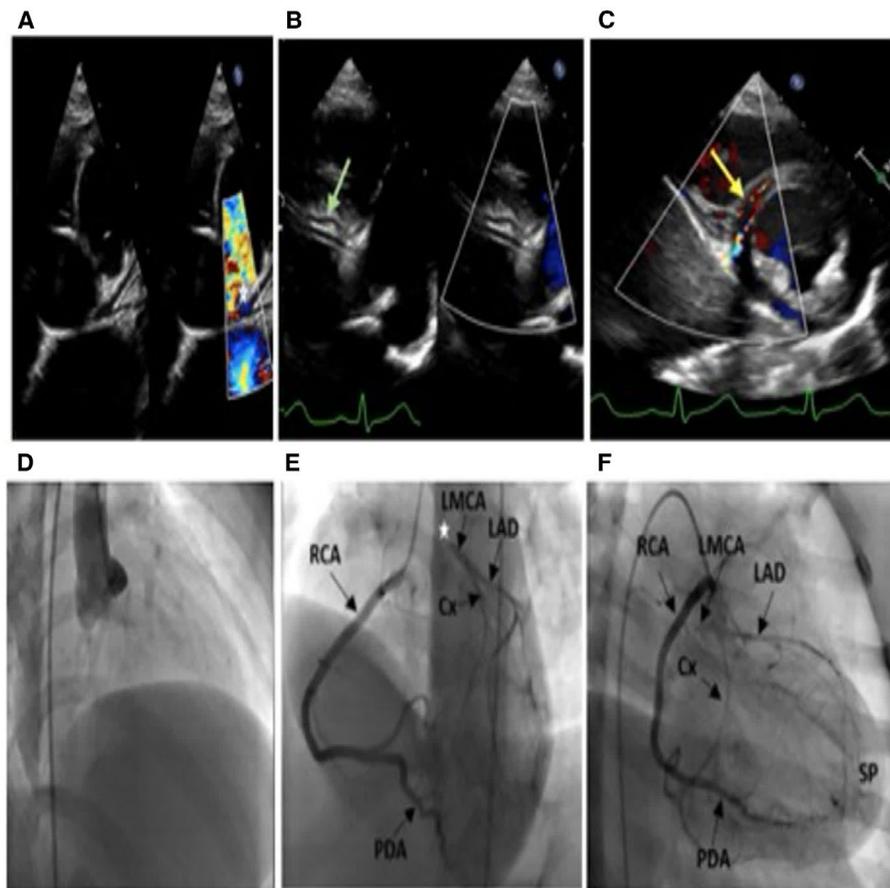


FIGURE 2 A, The LMCA ostial origin is atretic with retrograde flow in the LMCA (White star). B, Parasternal long axis view of the right ventricle inflow showing prominent PDA and septal perforators (Green arrow). C, Parasternal short axis view showing prominent septal perforators in the interventricular septum (Yellow arrow). D, Selective injection into the left coronary ostium showing no antegrade flow into the LMCA. E, Selective right coronary angiography showing a mildly dilated RCA with retrograde filling of the well-formed LMCA (White star). F, Multiple small septal perforators (SP) arising from the PDA and connect to LAD distally. Cx, circumflex artery; LAD, left anterior descending artery; LMCA, left main coronary artery; PDA, posterior descending artery; RCA, Right coronary artery; SP, septal perforators

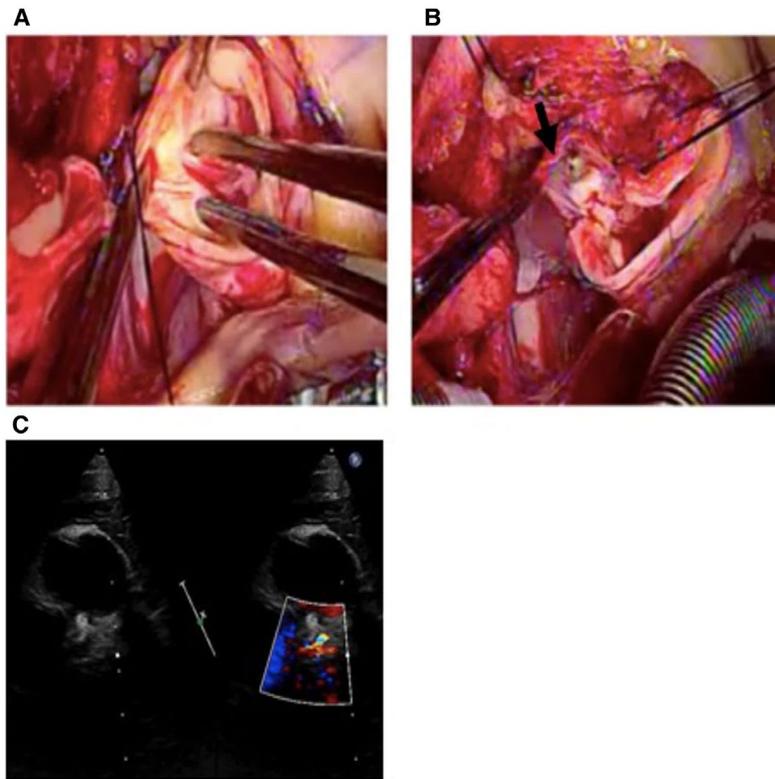


FIGURE 3 A, The aorta was transected above the sinotubular junction, we found atresia of the ostium of LMCA rightward to the left-right commissure. B, The posterior and lateral wall of the LCA and aortic wall were connected and then a triangle shaped patch with treated autologous pericardium was used to create an effective surface for coronary osteoplasty (Black arrow). C, Postoperative echocardiography showing normal antegrade flow in the LCA

The LMCA trunk was well developed and in close proximity to the aorta. The LMCA was incised until the bifurcation. The aorta was incised down to the top of the commissure to create an effective orifice. The posterior and lateral walls of the coronary artery were connected to the aortic wall using interrupted 7-0 sutures. A wide triangle shaped treated pericardial patch was used to enlarge the entire LMCA trunk (Figure 3B). He was discharged 4 days later after uneventful postoperative course and normal postoperative ECG. His TTE on discharge showed unobstructed antegrade flow in the LMCA, LAD, and Cx (Figure 3C) with good biventricular systolic function. He received prophylactic ASA for 3 months and is doing well 1 year later.

4 | CASE 3

A full-term 3-week-old female was referred to pediatric cardiology after a fetal echocardiogram showed multiple echogenic foci located adjacent to the interventricular septum, right ventricular moderator band, and left ventricular chordae. She was seen in ambulatory clinic for routine evaluation. At the time of clinic visit, she had appropriate weight gain, no cardiovascular-related symptoms and a normal ECG. The TTE revealed a prominent RCA (z-score 1.7) with normal ostial origin and antegrade flow, and a prominent branch arising anteriorly. Antegrade flow was seen in the usual location of the LAD; however, the ostial origin of the LMCA from the aorta was not seen, raising the possibility of anomalous origin of the LMCA (Figure 4A,B). Computed

tomography (CT) showed the LCA stump arising from the mid-part of the left posterior facing sinus of the pulmonary artery (Figure 4C). The RCA originated normally from the right aortic sinus and was only mildly dilated (Figure 4D). There was a collateral channel arising from the proximal RCA that coursed on the epicardial surface of the right ventricular outflow tract (RVOT) to connect to the LCA system below the left atrial appendage. The patient went on to have selective coronary angiography to confirm the diagnosis. Angiography demonstrated a mildly dilated RCA with retrograde filling of the LAD supplied by the collateral that was seen on CT along the RVOT and a separate collateral vessel from the RCA supplied the Cx. No LMCA connection to the aortic root was seen (Figure 5A). Pulmonary angiogram revealed an ostial "stump" of the LCA, which confirmed the diagnosis of ALCAPA with occlusion of the LMCA (Figure 5B). As the patient remained asymptomatic in the setting of adequate collateral supply of the LCA system from the right, the decision was made to follow her closely as an outpatient with ECG, echocardiogram, and Holter monitor, initially 3 months after presentation and subsequently annually, with periodic coronary artery imaging, and myocardial perfusion scanning. She is doing well 1 year later.

5 | REVIEW OF LITERATURE

All available literature reports in English on LMCAOA were collected by electronic search of the MEDLINE database. We have thus been

FIGURE 4 A, Prominent RCA with normal ostial origin and antegrade flow with a prominent branch arising anteriorly. B, Antegrade flow seen in the usual location of the LAD; however, ostial origin of the LMCA from the aorta not seen. C, LCA stump arising from the mid-part of the left posterior facing sinus of the pulmonary artery (arrow). The gray contrast of the LCA stump is clearly different from the bright contrast of the RCA-fed portion of the left coronary system. D, RCA arising normally from the right aortic sinus is bright in contrast and mildly dilated (arrow). LAD, left anterior descending artery; LCA, left coronary artery; LMCA, left main coronary artery; RCA, right coronary artery

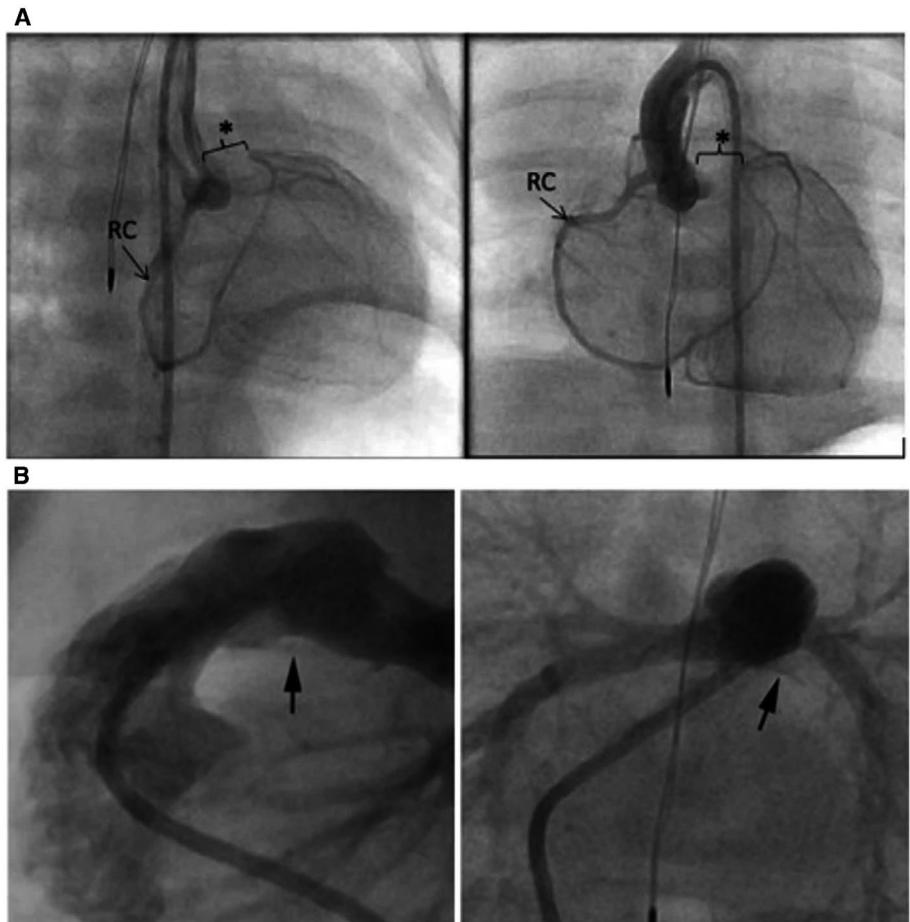
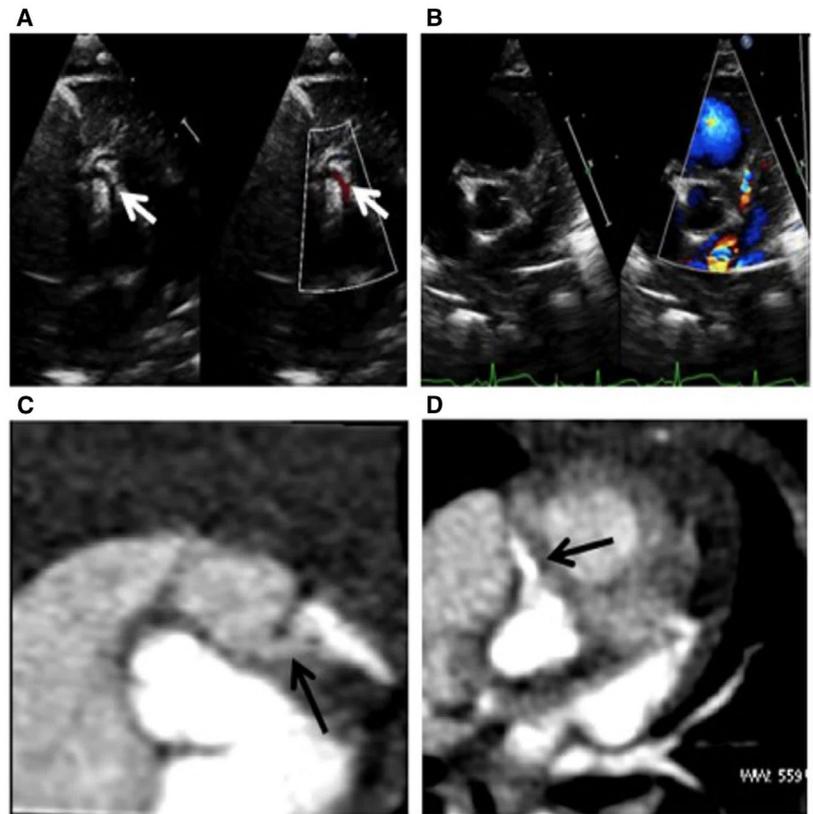


FIGURE 5 A, Angiography showing a mildly dilated RCA, with retrograde filling of the LAD and Cx. No LMCA is seen (*). B, Pulmonary angiogram revealing faint filling of the ostial "stump" (arrow) of the LCA. Cx, circumflex artery; LAD, left anterior descending artery; LMCA, left main coronary artery; RCA, right coronary artery

able to collect a total of 93 pediatric and adult cases of LMCAOA, summarized in Tables 1 and 2 respectively; of which 56% were male. Among these patients, there were 50 pediatric patients that ranged from the age of 3 to 17 years old, with more than one-third ($n = 18$) diagnosed in early infancy. There were also 43 adult patients that ranged from the age of 23 to 85 years old, and 84% of them were older than 40 years of age at the time of diagnosis.

5.1 | Presenting symptoms and associated cardiac lesions

Of the 50 pediatric patients, 88% ($n = 44$ cases) were symptomatic while only 10% ($n = 5$ cases) were asymptomatic, including three infants (Table 3). The most common symptoms in pediatric patients were heart failure in 44% ($n = 22$), syncope in 28% ($n = 14$), and chest pain in 24% ($n = 12$ patients). In the 18 reported cases occurring during early infancy at 1 year of age or less, they predominantly presented with heart failure ($n = 12$). Ten percent of pediatric patients ($n = 5$) were presented with sudden cardiac death (SCD), in that three of them were during early infancy. Of the 43 adult patients, 79% ($n = 34$) were symptomatic while only 9 patients were asymptomatic. Unlike pediatric patients, the most common presenting symptoms among adult patients was angina in 48.8% ($n = 21$) and exertional dyspnea in 14% ($n = 6$) patients. Heart failure was extremely rare and reported only in two adult patients. About 7% of adult patients ($n = 3$) presented with SCD, only one of them survived.

We found that 44% of pediatric patients ($n = 22$) had additional cardiac lesions (Table 3), most commonly mitral valve prolapse in seven cases and supra-aortic stenosis in four cases. Nearly half of the adult patients ($n = 22$) were associated with other cardiac lesions, most commonly coronary atherosclerosis in nine cases and severe aortic insufficiency in four cases. Unlike pediatric patients, only three adult patients had mitral valve prolapse.

5.2 | Diagnostic investigations

ECG at rest was available for 34 pediatric patients with evidence of ischemia in half of them (Table 4). However, only 16 adult patients had ECG at rest with evidence of ischemia in 37.2% ($n = 6$). Of the 50 pediatric patients, only 10 had an exercise stress test. Ischemic changes in the form of ST depression were reported in eight cases (Table 4). Additionally, 11 pediatric patients had MPI with perfusion defects reported in nearly 73% of them ($n = 8$). Exercise stress test (EST) was available for 13 adult patients, nearly 85% of them had ischemic changes ($n = 11$). MPI was performed in 15 adult patients, among which 73.3% ($n = 11$) had perfusion defects.

Echocardiography was suspicious for diagnosis of LMCAOA in 47% of the pediatric patients ($n = 16$) who had echocardiographic results available ($n = 34$) (Table 4). Nine pediatric patients were misdiagnosed as ALCAPA, while coronaries were reported to be normal in nine pediatric patients. As expected, of the 13 adult patients, LMCAOA was not suspected by echocardiography in any of them (Table 4).

5.3 | Left ventricular systolic function at time of diagnosis

The LV systolic function was known at the time of diagnosis in 37 of 50 cases in the pediatric population (Table 4). It was normal in 43.2% ($n = 16$) and severely reduced in 32.4% ($n = 12$). Nearly one-fifth of pediatric patients ($n = 7$) had wall motion abnormalities of the LV. Of the 43 adult patients, the LV systolic function was known at the time of diagnosis for 22 cases (Table 4). It was normal in 54.5% ($n = 12$) and severely reduced in 22.7% ($n = 5$). Unlike pediatric patients, only one adult patient had wall motion abnormalities of the LV.

The diagnosis of LMCAOA was confirmed by coronary angiography alone in about 72% of pediatric patients ($n = 36$) and by both coronary angiography and cardiac imaging (CT angiography or magnetic resonance angiography) in 12% ($n = 6$). Two pediatric cases were diagnosed by autopsy and six had intraoperative diagnosis. Nearly 42% ($n = 18$) of adult patients were diagnosed by coronary angiography alone, while 46.5% ($n = 20$) were diagnosed by both coronary angiography and cardiac imaging (CT angiography or magnetic resonance angiography). Five adult cases were diagnosed by autopsy.

The size of collaterals supplying the left coronary system from the RCA was described in 44 pediatric patients and 41 adult patients (Table 4). In pediatric patients, the collaterals were small-sized in about 70% ($n = 31$) and large-sized in only 9% ($n = 4$) compared to 80.5% of adult patients ($n = 33$). We also looked where the collaterals joined the left coronary system. This was described in 14 pediatric patients and 23 adult patients (Table 4). Among pediatric population, the collaterals joined the left coronary system at distal LAD and/or distal Cx in 14.3% ($n = 2$), mid and distal LAD and distal Cx in 43.8% ($n = 6$), mid LAD in 7.1% ($n = 1$), proximal LAD in 28.6% ($n = 4$) and first diagonal branch in 7.1% ($n = 1$). In adult patients, the collaterals joined the left coronary system at distal LAD and/or distal Cx in 30.4% ($n = 7$) cases, mid and distal LAD in 8.7% ($n = 2$), mid LAD in 39.1% ($n = 9$), and proximal LAD or Cx in 21.7% ($n = 5$).

5.4 | Management

Two pediatric patients died from SCD; while in two other patients the management was not mentioned (Table 5). Of the remaining 46 pediatric patients, 84.7% ($n = 39$) had myocardial revascularization surgery, CABG in 71.1% ($n = 27$) and coronary reconstruction osteoplasty in 28.9% ($n = 11$) cases. The surgery was not specified in one patient. In contrast, three adult patients died from SCD; two other patients were asymptomatic but diagnosed on autopsy after non-cardiac death. The management was unknown for seven adult patients. Of the remaining 31 patients, two-thirds ($n = 21$) had myocardial revascularization surgery as detailed in (Table 5). This included CABG in 17 cases and non-specified surgery in the other 4 patients. Interestingly, unlike the pediatric population, none of the adult patients had coronary reconstruction osteoplasty. Nearly one-third of adult patients ($n = 10$) and 15% of pediatric patients ($n = 7$) did not have coronary surgery and were treated medically.

TABLE 1 Pediatrics patients with LMCAOA

Reference, year	Sex	Age	Presentation	ECG	Stress test	Diagnosis by Echo	LV systolic function	Diagnosis tool	Collaterals	Associated findings	Treatment	Outcome
Goormaghtigh et al ⁵ 1955	F	9 y	Syncope Chest pain	LVH. No ischemia	N/A	N/A	N/A	Autopsy	Not specified	RCA stenosis	Died	Died
Mullins et al ⁶ 1972	M	10 y	Syncope Chest pain	Antero septal ischemia	EST: ST depression	N/A	N/A	Coronary angiography	Multiple small collaterals from mid RCA to mid LAD, PDA and PLV to distal LAD and Cx	RCA stenosis	CABG (SVG to LAD)	Survived (2 mo FU)
Allen et al ⁷ 1974	M	9 y	Syncope	N/A	EST: ST depression	N/A	N/A	Coronary angiography	Multiple small collaterals from PDA to distal LAD and Cx	Supravalvar aortic stenosis	CABG (SVG to LAD)	Survived (Unknown FU)
Levin et al ⁸ 1978	M	17 y	Asymptomatic	N/A	N/A	N/A	N/A	Coronary angiography	Multiple small and moderate collaterals from RCA to LAD	Supravalvar aortic stenosis	N/A	N/A
Vidne et al ⁹ 1979	F	12 y	Decreased exercise tolerance	RVH. No ischemia	N/A	N/A	N/A	Coronary angiography	Multiple small and moderate collaterals from RCA to LAD	VSD, pulmonary stenosis	No coronary surgery	Survived (6 y FU)
Byrum et al ¹⁰ 1980	F	6 mo	Heart failure	Antero lateral infarction	N/A	N/A	Akinesia of apex, anterior & lateral LV	Surgery	Multiple small collaterals from RCA to small LAD	None	Non-specified Surgery	Died after surgery
Van der Hauwaert et al ¹¹ 1982	F	2 y	Heart failure	Antero lateral infarction	N/A	N/A	Severe LV dysfunction	Coronary angiography	Multiple small collaterals from PDA and PLV	None	No coronary surgery	Died (at age 11 y)
Fortune et al ¹² 1987	F	17 mo	Heart failure	N/A	N/A	N/A	N/A	Coronary angiography	Multiple small collaterals from PDA and PLV to distal LAD and Cx	Mitral valve prolapse and severe MR	CABG (LIMA to LAD) + MV repair	Survived MV replacement after few year
Leitz et al ¹³ 1987	F	5 y	Heart failure	Normal	N/A	Normal coronaries	Anterior wall hypokinesia	Coronary angiography	Multiple collaterals from RCA to LAD	Mild MR	CABG (LIMA to LAD)	Survived (1 y FU)
Debich et al ¹⁴ 1989	M	13 y	SCD	N/A	N/A	N/A	N/A	Autopsy	Small collaterals from conal and marginal RCA branches to LAD	None	Died	Died
Gay et al ¹⁵ 1989	F	2 mo	N/A	N/A	N/A	N/A	N/A	Coronary angiography	N/A	None	Reconstruction of LCA with aortic wall	Survive (UnknownFU)
Koh et al ¹⁶ 1989	F	11 y	Syncope	Normal	EST: ST depression MPI (SPECT) perfusion defect in anterior wall	Suspected LMCAOA	Normal	Coronary angiography	Multiple small collaterals from RCA, PDA and PLV to mid and distal LAD and distal Cx	None	CABG (LIMA to LAD)	Survived (2 y FU) MPI SPECT: no exercise induced perfusion defect

(Continues)

TABLE 1 (Continued)

Reference, year	Sex	Age	Presentation	ECG	Stress test	Diagnosis by Echo	LV systolic function	Diagnosis tool	Collaterals	Associated findings	Treatment	Outcome
Sato et al ¹⁷ 1990	M	17 y	Palpitations Ventricular tachycardia	N/A	N/A	N/A	Akinesia of LV apex	Coronary angiography	N/A	None	CABG (LIMA to LAD, SVG to OM)	Survived (8 mo FU)
Bedogni et al ¹⁸ 1992	M	16 y	Chest pain Syncope	Ischemia: ST depressed in V1-V6	N/A	N/A	Akinesia of LV apex	Coronary angiography	Multiple small and moderate collaterals from PLV to LAD & Cx	None	CABG (LIMA to LAD, SVG to OM)	Survived (46 mo FU)
Rosenkranz et al ¹⁹ 1992	F	5 y	Chest pain	LVI: No ischemia	EST: Severe ST depression in anterior precordial leads	Normal coronaries	Normal	Surgery	Multiple small and moderate collaterals from PDA, PLV to LAD and Cx	Supravalvar aortic stenosis, quadri-cuspid aortic valve	CABG (LIMA to LAD) Ascending aorta patch plasty 15 mo before	Survived (UnknownFU) Normal ECG
Amaral et al ¹ 2000	M	9 mo	Heart failure	Antero lateral infarction	N/A	ALCAPA	Severe LV dysfunction, lateral wall hypokinesia	Coronary angiography	Multiple small and moderate collaterals from PDA, PLV to LAD and Cx	None	CABG (LIMA to LAD)	Survived (11 mo FU) LV function improved
Amaral et al ¹ 2000	M	11 y	Asymptomatic	Antero lateral infarction	EST: Antero apical ischemia MPI (Tc-99) diffuse moderately decreased contractility of LV	ALCAPA	Severe LV dysfunction	Surgery	Not specified	Antero apical aneurysm	CABG (LIMA to LAD)	Survived (3.5 y FU) Persistent anterolateral infarct on ECG
Gerlis et al ²⁰ 2002	M	3 mo	Heart failure	BVH. Ischemic changes: inverted T waves	N/A	ALCAPA	Severe LV dysfunction	Surgery	Small Collaterals from distal portion of the PLV of the RCA to the LAD	None	CABG (LIMA to LAD)	Died POD 3
Shah et al ²¹ 2004	M	4 y	Syncope	Normal	EST: normal	Suspected LMCAOA	Normal	Coronary angiography	Multiple small collaterals from RCA, PDA and PLV to mid and distal LAD and distal Cx	None	CABG (LIMA to LAD)	Survived (FU till Discharge)
Lin et al ²² 2005	F	7 y	Heart failure	N/A	N/A	Suspected LMCAOA	Moderate LV dysfunction	Coronary angiography	Multiple collaterals from RCA to LAD	Moderate MR	Aortic reimplantation No MV surgery	Survived (3 y FU) LV function + MR improved
Sunagawa et al ²³ 2005	M	1 y	Heart failure	No ischemia	N/A	Normal coronaries	N/A	Coronary angiography	Multiple small and moderate collaterals from PDA, PLV to LAD	MV prolapse with severe MR	CABG (LIMA to LAD) + MV repair	Survived (30 mo FU)

(Continues)

TABLE 1 (Continued)

Reference, year	Sex	Age	Presentation	ECG	Stress test	Diagnosis by Echo	LV systolic function	Diagnosis tool	Collaterals	Associated findings	Treatment	Outcome
Betrian Blasco et al ²⁴ 2006	F	11 y	Chest pain Syncope	Normal	EST: ST depression MPI: normal	Normal coronaries	Normal	Coronary angiography	Multiple small collaterals from RCA, PDA and PLV to mid and distal LAD and distal Cx	MV prolapse with severe MR	CABG (LIMA to LAD) No MV repair	Survived (6 mo FU) MR improved to mild
Varghese et al ²⁵ 2007	M	8 mo	Heart failure, Cardiac arrest	RBBB; No ischemia.	N/A	ALCAPA	Severe LV dysfunction	Coronary angiography	Multiple small and moderate collaterals from proximal RCA and PDA to proximal LAD and distal Cx	None	Coronary reconstruction with autologous pericardial patch	Survived (3 mo FU)
Gebauer et al ²⁶ 2008	F	3 y	Chest pain	Ischemic changes	N/A	ALCAPA	Severe LV dysfunction	Coronary angiography	Small Collaterals from RCA to left coronary system	None	CABG: (LIMA to LAD and SVG to marginal branch of Cx)	Survived (10 mo FU) LV function improved
Gebauer et al ²⁶ 2008	F	3 mo	Cardiac arrest	Ischemic changes	N/A	ALCAPA	Normal	Coronary angiography	Small collaterals from RCA to LCA	Severe MR	CABG (LIMA to LAD)	Survived (5 mo FU) Moderate MR
Takeuchi et al ²⁷ 2009	M	30 mo	Heart failure	LA enlarged. No ischemia	MPI (SPECT): diffuse dissociation between perfusion and lipid metabolism	Normal coronaries	Normal	Coronary angiography	Multiple small collaterals from RCA, PDA and PLV to mid and distal LAD and distal Cx	None	Coronary reconstruction using azygous vein graft	Survived (6 y FU) Severe LCA stenosis pericardial patch osteoplasty at 7 mo + PTCA at 36 mo
Sohn et al ²⁸ 2010	M	10 mo	Heart failure	N/A	N/A	Suspected LMCAOA	Normal	Coronary angiography	Multiple small and moderate collaterals from PDA, PLV to LAD and Cx	MV prolapse with severe MR, EFE	No coronary surgery No MV repair	Died 4 h after Cath.
Jatene et al ²⁹ 2012	M	7 y	Syncope	N/A	N/A	N/A	N/A	Coronary angiography	Multiple small collateral from proximal RCA to proximal LAD	CoA and VSD	CABG (LIMA to LAD), COA repair and VSD closure	Survived (FU till discharge)
Tanawuttiwat et al ³ 2013	M	11 y	Chest pain Syncope	Antero-lateral ischemia	MPI: Infarction in lateral, anterior & septal walls of LV	Suspected LMCAOA	Mild LV dysfunction	Coronary angiography, cardiac CT, CMR	Multiple small & moderate collaterals from PDA, PLV to Cx. Large collateral from proximal RCA to mid LAD	Myocardial bridging	CABG (LIMA to LAD)	Survived (7 d FU)
Kaczorowski et al ² 2012	M	10 mo	Asymptomatic	Ischemic changes: Antero lateral Q wave	N/A	ALCAPA	Low normal LV function	Surgery	Multiple small collaterals from RCA, PDA to LAD	Severe MR, EFE	Homograft patch osteoplasty No MV repair	Survived (2 y FU) MR improved to mild

(Continues)

TABLE 1 (Continued)

Reference, year	Sex	Age	Presentation	ECG	Stress test	Diagnosis by Echo	LV systolic function	Diagnosis tool	Collaterals	Associated findings	Treatment	Outcome
Kaczorowski et al ² 2012	F	7 wk	Heart failure	Ischemic changes: ST elevation	N/A	ALCAPA	Severe LV dysfunction	Surgery	Minimal Collaterals from RCA	Mild MR	Homograft patch osteoplasty	Died POD3 LV necrosis
Kaczorowski et al ² 2012	F	31 mo	Heart failure	Ischemic changes: Lateral T wave inversion	N/A	Suspected LMCAOA	Severe LV dysfunction	Coronary angiography	Extensive small collaterals	None	Homograft patch osteoplasty	Survived (1 y FU) LV function normalized
Patil et al ³⁰ 2012	F	2 y	Cyanotic spells, Heart failure	Ischemic changes: Diffuse ST elevation	N/A	Suspected LMCAOA	Severe LV dysfunction	Coronary angiography	Multiple small collaterals from RCA	Tetralogy of Fallot	Left coronary angio-plasty + BT shunt	Survived (3 mo FU) LV function improved
Laux et al ³¹ 2013	F	3 d	Cardiac arrest	N/A	N/A	Suspected LMCAOA	Severe LV dysfunction	Cardiac CT, Coronary angiography	N/A	None	CABG (LIMA to LAD)	Died in OR
Laux et al ³¹ 2013	F	7 d	Heart failure	N/A	N/A	Normal coronaries	Severe LV dysfunction	Coronary angiography	Multiple small collaterals from RCA, PDA to the LAD	Bicuspid aortic valve, Critical AS	No coronary surgery; Balloon dilation of aortic valve	Died few hours after Cath.
Weigeldt et al ³² 2017	M	4 y	Cardiac arrest, Ventricular fibrillation	No ischemia	N/A	Normal coronaries	Normal	Cardiac MRI, Coronary angiography	Small collaterals from RCA	Mild MR, EFE	CABG (LIMA to LAD)	Survived (1 y FU)
Luo et al ³³ 2013	M	1 y	Asymptomatic	Ischemic changes: Q wave in I,AVL, V3-V5	N/A	Suspected LMCAOA vs ALCAPA	Moderate LV dysfunction	Coronary angiography	One large collateral from proximal RCA to proximal LAD + small collaterals	MV prolapse with severe MR	No coronary surgery No MV repair	Survived (UnknownFU)
Ohba et al ³⁴ 2015	F	9 y	Chest pain	Ischemic changes: ST depressed in V4-6, Q wave in I and AVL	MPI (CMR): reduced perfusion in LAD and Cx areas	N/A	Hypokinesia of LV anterior wall	Cardiac CT, Coronary angiography	Small collaterals from RCA and PDA to LAD	None	Coronary angioplasty by autologous pericardial patch	Survived (4 mo FU) MPI: improved perfusion
Sugimoto et al ³⁵ 2016	M	13 y	Syncope Shortness of breath Chest pain	No ischemia	MPI: reduced perfusion in LCA region	Suspected LMCAOA and anomalous origin from Non-coronary cusp	N/A	Cardiac CT, Coronary angiography	Small collaterals from RCA to left coronary system	None	Coronary osteoplasty autologous pericardial patch	Survived (8 mo FU) MPI: no ischemia

(Continues)

TABLE 1 (Continued)

Reference, year	Sex	Age	Presentation	ECG	Stress test	Diagnosis by Echo	LV systolic function	Diagnosis tool	Collaterals	Associated findings	Treatment	Outcome
D'Souza et al ³⁶ 2016	M	7 wk	Heart failure	N/A	N/A	Normal coronaries	Normal	Coronary angiography	Small Collaterals from RCA to LAD	MV prolapse Ruptured MV chordae, severe MR, ASD	CABG (LIMA to LAD) + MV repair	Survived (1 y FU) Required second MV repair after 4 wk
Karayiannis et al ³⁷ 2016	M	12 y	Syncope palpitations	Normal	EST: ST depression MPI (CMR) Normal	Suspected LMCAOA	Normal	Coronary angiography	Extensive multiple small collaterals from RCA to the left coronary system.	None	CABG (LIMA to LAD)	Survived (FU till discharge)
Pizzuto and Zampieri ³⁸ 2016	F	6 mo	Heart failure	N/A	N/A	Suspected LMCAOA	Severe LV dysfunction	Coronary angiography	Large collateral from RCA to LMCA	Shone's complex	No coronary surgery, only Ross/Konno	Survived (1 y FU) LV Function normalized
Tominaga et al ³⁹ 2016	F	5 mo	Heart failure Shock	N/A	N/A	N/A	N/A	Coronary angiography	Multiple small collaterals from RCA to left coronary system	Severe MR	At 2 y age: Coronary angioplasty using azygous vein patch + MV repair	Survived (2 y FU) MV replacement after 2 y
Yajima et al ⁴⁰ 2017	F	6 y	Chest pain	Normal	N/A	N/A	N/A	Coronary angiography	N/A	None	CABG (LIMA to second diagonal branch, SVG to obtuse marginal branch)	Survived (27 y FU). Required re do CABG at age 33 y
Fujita et al ⁴¹ 2017	M	13 y	Syncope seizure	No ischemia	EST + MPI normal	Suspected LMCAOA	Normal	Coronary angiography, cardiac CT	Small collaterals from PDA to Cx	None	CABG (LIMA to LAD)	Survived (FU till discharge)
Sathanandam et al ⁴² 2017	M	9 y	Syncope	Normal	N/A	Suspected LMCAOA	Normal	Coronary angiography	Prominent conal branch off the RCA that supplies the LAD	Anomalous origin of RCA: origin superior and leftwards more than usual	CABG (LIMA to LAD)	Survived (8 wk FU) LV function normal
Hayashi et al ⁴³ 2017	M	13 y	Syncope Chest pain	Normal	EST: ST depression V4-V6 MPI: ischemia and hypokinesia at the area of the lateral branch in the early phase	Suspected LMCAOA	Normal	Coronary angiography	Small Collaterals from RCA and PDA to left coronary artery	Mitral valve prolapse with mild MR	CABG (LIMA to LAD)	Survived (3 y FU) EST: normal

(Continues)

TABLE 1 (Continued)

Reference, year	Sex	Age	Presentation	ECG	Stress test	Diagnosis by Echo	LV systolic function	Diagnosis tool	Collaterals	Associated findings	Treatment	Outcome
Weigand et al ⁴⁴ 2017	M	3 wk	Apparent life threatening event	N/A	N/A	Suspected LMCAOA	Normal	Coronary angiography	Extensive collaterals from RCA to proximal LAD	None	N/A	N/A
Sabzi et al ⁴⁵ 2018	M	1.6 y	Chest pain Exertional dyspnea	Antero lateral ischemia	MPI (Th 201): persistent ischemia in antero-septal and lateral segments	Normal coronaries	Moderate LV dysfunction Hypokinesia of antero lateral wall	Coronary angiography	Large collateral from RCA to first diagonal branch	Supravalvar aortic stenosis, small coronary fistula to main pulmonary artery	CABG (LIMA to LAD and SVG to Cx) + supravalvar aortic stenosis repair	Survived (1 y FU)
Veerappan et al ⁴⁶ 2018	M	10 d	Asymptomatic	N/A	N/A	Suspected LMCAOA	Normal	Coronary angiography	extensive collaterals from the RCA	PDA	No coronary surgery	Survived (4 y FU) Normal LV function on Echo

Abbreviations: AD, atrial septal defect; ALCAPA, anomalous origin of left coronary artery from pulmonary artery; BVH, biventricular hypertrophy; CABG, coronary artery bypass grafting; CMR, cardiac magnetic resonance imaging; CoA, coarctation of aorta; CT, computed tomography; Cx, circumflex artery; EFE, endocardial fibroelastosis; EST, exercise stress test; FU, follow up; LAD, left anterior descending; LCA, left coronary artery; LIMA, left internal mammary artery; LMCAOA, left main coronary artery ostial atresia; LVH, left ventricular hypertrophy; MPI, myocardial perfusion imaging; MRI, magnetic resonance imaging; N/A, not available; OM, obtuse marginal artery; PDA, posterior descending artery; PLV, posterolateral ventricular branch; PTCA, percutaneous transluminal coronary angioplasty; RBBB, right bundle branch block; RCA, right coronary artery; RVH, right ventricular hypertrophy; SCD, sudden cardiac death; SPECT, single-photon emission computed tomography; SVG, saphenous vein graft; VSD, ventricular septal defect.

TABLE 2 Adult patients with LMCAOA

Reference, year	Sex	Age	Presentation	ECG	Stress test	Echo	Diagnosis by	LV systolic function	Diagnosis tool	Collaterals	Associated findings	Treatment	Outcome
Murphy et al ⁴⁷ 1967	M	71 y	Angina	N/A	N/A	N/A	N/A	N/A	Autopsy	Three moderate to large collaterals to LAD	Non-obstructive coronary atherosclerosis	Died	Died
Fortuin and Roberts ⁴⁸ 1971	M	60 y	Angina SCD	A Fib with non-specific ST-T wave changes	N/A	N/A	N/A	N/A	Autopsy	Large conus branch to mid LAD and large marginal branch to distal LAD	Ruptured atherosclerotic plaque with stenosis of conus branch	Died	Died
Vidne et al ⁹ 1979	M	52 y	Angina	No ischemia	N/A	N/A	Normal	Normal	Coronary angiography	Large conus branch & minor collaterals from RCA to LAD and Cx	None	CABG (SVG to LAD and Cx)	Survived (6 wk FU)
Vidne et al ⁹ 1979	F	50 y	Angina	Complete RBBB. No ischemia	N/A	N/A	N/A	N/A	Coronary angiography	Multiple small collaterals from mid RCA to LAD & Cx & large collateral from PDA to distal LAD	None	CABG (SVG to LAD)	Survived (6 wk FU)
Dymond et al ⁴⁹ 1980	M	38 y	Angina	Normal	EST: ST depression MPI: SPECT anterior ischemia	Normal coronaries	Anterior hypokinesia of LV	Normal	Coronary angiography	Large collateral branch continuing from PDA to distal LAD	None	CABG (SVG to LAD)	Survived (2 mo FU) SPECT: normal
Leivo and Laurila ⁵⁰ 1987	F	76 y	Asymptomatic Non-cardiac death	N/A	N/A	N/A	N/A	N/A	Autopsy	Large intra myocardial collateral branch from proximal RCA to proximal LAD	Myocardial bridging	Died	Died
Beretta et al ⁵¹ 1990	F	49 y	Angina	N/A	N/A	N/A	N/A	N/A	Coronary angiograph	Collateral vessels from RCA to LAD (not defined)	None	CABG (LIMA to LAD)	Survived (22 mo FU)
Ruiz and Lau ⁵² 1991	F	48 y	Angina	Ischemic changes: Inverted T wave V1-V4	MPI: SPECT anterior, inferior and postero lateral wall ischemia	N/A	Normal	Normal	Coronary angiography	Large collateral from PDA and PLV to distal LAD and Cx	None	CABG (SVG)	Survived (5 y FU) SPECT: normal
Carosio et al ⁵³ 1991	F	64 y	Angina	N/A	N/A	N/A	N/A	N/A	Coronary angiography	Large collateral from PDA and left auricular branch	None	Non-specified Surgery	N/A
Bedogni et al ¹⁸ 1992	F	43 y	Angina	N/A	EST critical ST depression MPI: SPECT lateral ischemia	N/A	Normal	Normal	Coronary angiography	Large collaterals from PDA and PLV to distal LAD and Cx	None	CABG (LIMA to LAD), 50% distal stenosis of the graft, PCI at 2 mo later	Survived (2 mo FU). SPECT: unchanged
Ghosh et al ⁵⁴ 1993	M	56 y	Angina	Normal	N/A	N/A	Normal	Normal	Coronary angiography	Large conus branch of RCA to mid LAD	Obstructive coronary Atherosclerosis	CABG (SVG to LAD and OM)	Survived (FU till discharge)

(Continues)

TABLE 2 (Continued)

Reference, year	Sex	Age	Presentation	ECG	Stress test	Diagnosis by Echo	Diagnosis by LV systolic function	Diagnosis tool	Collaterals	Associated findings	Treatment	Outcome
Musiani et al ⁵⁵ 1993	M	44 y	Angina	N/A	N/A	N/A	N/A	Coronary angiography	Large single collateral from the ostium of the RCA to LAD	Coronary artery atherosclerosis	CABG (LIMA to LAD, SVG to OM, diagonal and PDA)	Survived (16 mo FU)
Musiani et al ⁵⁶ 1995	M	68 y	Angina	N/A	MPI: SPECT Antero apical ischemia	N/A	Severe LV dysfunction	Coronary angiography	Large conus branches of RCA to mid LAD. Hypoplastic segment at mid LAD is noted	None	CABG (LIMA to proximal LAD, SVG to distal LAD)	Survived (1 y FU) SPECT: normal. LV function: unchanged
Elian et al ⁵⁷ 2003	M	42 y	Asymptomatic	N/A	MPI: SPECT anterolateral and septal ischemia	N/A	N/A	Coronary angiography	Large collaterals from PDA and PLV branch to proximal LAD and Cx, respectively	None	No coronary surgery	Survived (18 mo FU) SPECT: unchanged
Elian et al ⁵⁷ 2003	M	23 y	Syncope	N/A	EST: ST depression	N/A	Normal	Coronary angiography	Multiple small collaterals from RCA to LAD, Cx	Mitral valve prolapse. No MR	CABG (LIMA to LAD)	Survived (30 mo FU)
Nishida et al ⁵⁸ 2005	F	82 y	Asymptomatic Non-cardiac death	N/A	EST: ST depression	N/A	N/A	Autopsy	Large collateral from RCA sinus to proximal LAD	Non-obstructive coronary atherosclerosis	Died	Died
Nishida et al ⁵⁸ 2005	M	34 y	SCD	N/A	N/A	N/A	N/A	Autopsy	Large collateral from RCA sinus to distal LAD	None	Died	Died
Kapetanopoulos et al ⁵⁹ 2007	F	53 y	Angina	N/A	EST: abnormal (not specified)	N/A	N/A	Cardiac CT, coronary angiography	Large V marginal branch to mid LAD. Several collaterals from PLV to Cx	None	N/A	N/A
Nicol et al ⁶⁰ 2007	M	72 y	Angina	N/A	N/A	N/A	N/A	Cardiac CT, coronary angiography	Large collateral from marginal branch to mid LAD	None	N/A	N/A
Nicol et al ⁶⁰ 2007	M	76 y	Angina	N/A	N/A	N/A	hypokinetic lateral LV wall	Coronary angiography, cardiac CT, CMR	Large collateral from conus branch and mid RCA to mid LAD	Malignant path of the collateral vessels	N/A	N/A
Nicol et al ⁶⁰ 2007	M	62 y	Asymptomatic	N/A	EST: ST depression in anterior leads	N/A	N/A	Coronary angiography, cardiac CT	Large collateral from conus branch to mid LAD	Malignant path of the collateral vessels	N/A	N/A
Levisman et al ⁶¹ 2009	M	53 y	Atypical chest Pain	N/A	EST: Mild anterior and inferior ischemia	N/A	N/A	Cardiac CT, coronary angiography	Large acute marginal branch to distal LAD	None	N/A	N/A

(Continues)

TABLE 2 (Continued)

Reference, year	Sex	Age	Presentation	ECG	Stress test	Echo	Diagnosis by	LV systolic function	Diagnosis tool	Collaterals	Associated findings	Treatment	Outcome
Saito et al ⁶² 2009	M	48 y	Asymptomatic	Ischemic changes: ST elevation V1-V3, Q wave in V1,V2	N/A	Normal coronaries	Normal	Severe LV dysfunction	Coronary angiography, cardiac CT	Small collaterals from PDA and PLV to Cx and hypoplastic LAD	None	CABG (LIMA to SVG, SVG to RI)	Survived (22 d FU) LV function normalized
Chou et al ⁶³ 2009	F	32 y	Heart failure	N/A	MPI: SPECT anterior ischemia	Anomalous origin of LCA between the non-coronary and left coronary cusps	Coronary angiography, cardiac CT	Severe LV dysfunction	Coronary angiography, cardiac CT	Two large marginal branches from RCA to mid LAD and Cx. Collaterals from PDA to septal branches of LAD	Patent ductus arteriosus, severe AI, moderate pulmonary hypertension	No coronary surgery. PDA ligation and aortic valve repair.	Survived (30 mo FU) LV function improved, Pulmonary HTN + AI resolved. SPECT: improved
Duarte et al ⁶⁴ 2010	F	61 y	Angina	N/A	EST: equivocal	N/A	Coronary angiography, cardiac CT	N/A	Coronary angiography, cardiac CT	Moderate size conus branch of RCA to mid LAD	None	N/A	N/A
Veronese et al ⁶⁵ 2011	F	85 y	Atypical chest pain	Ischemic changes: Inverted T wave V3-V6, I, AVL	EST: ST depression	Normal coronaries	Coronary angiography, cardiac CT	Normal	Coronary angiography, cardiac CT	Multiple moderate and large collaterals from conus, RCA, PDA and PLV to LAD and Cx	Non-obstructive coronary atherosclerosis	No coronary surgery	Survived (FU till discharge)
Srinivas et al ⁶⁶ 2011	M	55 y	Angina	N/A	EST: equivocal MPI: normal	N/A	Coronary angiography, cardiac CT	N/A	Coronary angiography, cardiac CT	Large collaterals from PDA and PLV to distal LAD, Cx	None	No coronary surgery	Survived (FU till discharge)
Saremi et al ⁶⁷ 2011	F	67 y	Angina Exertional dyspnea	Normal	Stress Echo: mild global left ventricle hypokinesis	N/A	Coronary angiography, cardiac CT	Mild LV dysfunction	Coronary angiography, cardiac CT	Large collateral from the RCA to proximal Cx. Smaller collateral from RCA to LAD	None	N/A	N/A
Shen et al ⁶⁸ 2012	F	37 y	Angina Exertional dyspnea	Lateral wall ischemia, atrial flutter	MPI: SPECT ischemia in the LCA territories	N/A	Coronary angiography, cardiac CT	Moderate LV dysfunction	Coronary angiography, cardiac CT	Two large conal branches from RCA	Patent ductus arteriosus, severe AI, ascending aorta aneurysm	CABG (SVG to LMCA trunk) + Aortic valve & ascending aorta replacement	Survived (1 y FU)
Tanawuttiwat et al ⁹ 2013	M	59 y	Angina	N/A	SPECT: normal	N/A	Coronary angiography, cardiac CT	N/A	Coronary angiography, cardiac CT	Large collateral from conus branch and PDA to LAD and Cx, respectively	Obstructive coronary atherosclerosis	CABG (SVG to LAD, diagonal, OM, PLV and PDA)	Survived (FU till discharge)
Tanawuttiwat et al ² 2013	M	77 y	Asymptomatic	N/A	EST: ST depression MPI: SPECT equivocal (artifact vs mild inferior ischemia)	N/A	Coronary angiography, cardiac CT	N/A	Coronary angiography, cardiac CT	Large collaterals from proximal RCA and small collaterals from PDA to mid and distal LAD, respectively	Non-obstructive coronary atherosclerosis	No coronary surgery	Survived (FU till discharge)

(Continues)

TABLE 2 (Continued)

Reference, year	Sex	Age	Presentation	ECG	Stress test	Echo	Diagnosis by	LV systolic function	Diagnosis tool	Collaterals	Associated findings	Treatment	Outcome
Graidis et al ⁶⁹ 2012	M	65 y	Atypical chest Pain	Normal	EST & MPI (SPECT) small defect in anterior and posterior lateral wall	Normal coronaries	Normal	Normal	Coronary angiography	Large collateral from sinus node branch and posterolateral branch to LAD and Cx	None	No coronary surgery	Survived (3 y FU)
Numasawa et al ⁷⁰ 2013	F	65 y	Heart failure	N/A	Stress MPI: normal	Normal coronaries	Severe LV dysfunction	Severe LV dysfunction	Coronary angiography, cardiac CT	Collateral circulation from posterior descending	Moderate MR	No coronary surgery	Survived (1 y FU)
Sayin et al ⁷¹ 2013	M	65 y	Exertional dyspnea	N/A	Positive stress test	N/A	N/A	N/A	Coronary angiography	Single large artery originating from the proximal portion of the RCA to LAD. A short LMCA trunk was visible in the proximal segment	None	Non-specified surgery	N/A
Nicolini et al ⁷² 2014	M	51 y	Asymptomatic	N/A	N/A	N/A	N/A	N/A	Coronary angiography	Large conal branch from RCA	Supra-aortic membrane covering LMCA origin. Severe AI	Non-specified surgery	Survived (Unknown FU)
Rubio Alonso et al ⁷³ 2015	M	72 y	Asymptomatic	N/A	N/A	N/A	Moderate LV dysfunction	Moderate LV dysfunction	Coronary angiography, cardiac CT	Large conal branch with independent origin and anterior path to proximal LAD	Aortic valve stenosis	No coronary surgery	Survived (1 y FU)
Unzué et al ⁷⁴ 2015	M	67 y	Exertional dyspnea	A Fib, RBBB, No ischemia	MPI: ischemia in anterior and lateral territories	Normal coronaries	Severe LV dysfunction	Severe LV dysfunction	Coronary angiography	Large collateral from conal branch to middle of LAD	None	No coronary surgery. VAD then heart transplantation 2 mo later	Survived. Required heart transplant 2 mo later
Tapuz et al ⁷⁵ 2015	F	76 y	Asymptomatic	LBBB, No ischemia	MPI, SPECT Small defects in anterior and posterior lateral walls	Normal coronaries	Normal	Normal	Coronary angiography, cardiac CT	Significant moderate collaterals from conal branch, sinus node artery and PDA to LAD and Cx	None	Non-specified surgery	Survived (FU till discharge)
Kashou et al ⁷⁶ 2017	M	76 y	Status epilepticus	Ischemic changes: ST elevated in I, AVL & depressed in inferior leads	N/A	Normal coronaries	Normal	Normal	Coronary angiography	Large collaterals from distal RCA to LAD and Cx	LAD stenosis by 80%-90%	No coronary surgery	Survived (1 y FU) Normal LV function

(Continues)

TABLE 2 (Continued)

Reference, year	Sex	Age	Presentation	ECG	Stress test	Diagnosis by Echo	LV systolic function	Diagnosis tool	Collaterals	Associated findings	Treatment	Outcome
Tian et al ⁷⁷	F	48 y	Exertional dyspnea	Ischemic changes: ST depressed in I, AVL V3-V6	N/A	Normal coronaries	Normal	Coronary angiography, cardiac CT	Multiple small collaterals from RCA to left coronary system	MV prolapse with severe MR, ruptured chordae	CABG (LIMA to proximal LAD) + MV repair	Survived (3 mo FU) Normal LV function
Saedi et al ⁷⁸	F	36 y	Angina	N/A	N/A	Normal coronaries	Normal	Coronary angiography, cardiac CT	Multiple small collaterals from RCA to left coronary system	Subvalvar stenosis. Severe AI (had surgery as a child)	CABG (LIMA to LAD, SVG to LCx)	Survived (FU till discharge)
Cortes et al ⁷⁹	F	28 y	Cardiac arrest (SCD)	Normal	N/A	Normal coronaries	Moderate LV dysfunction	Coronary angiography, cardiac CT	moderate distal RCA collaterals supplying left coronary artery via septal collateral system	None	CABG (LIMA to LAD, Left radial artery to obtuse marginal)	Survived (1 y FU) LV function normalized
Tolia et al ⁸⁰	F	55 y	Exertional dyspnea	N/A	Myocardial perfusion scan anterior wall ischemia	Normal coronaries	Normal	Coronary angiography	Large collaterals from RCA to left coronary system	MV prolapse with severe MR	CABG (Non-specified) No MV repair	N/A

Abbreviations: A fib, atrial fibrillation; AI, aortic insufficiency; CABG, coronary artery bypass grafting; CMR, cardiac magnetic resonance imaging; CT, computed tomography; Cx, circumflex artery; EST, exercise stress test; FU, follow up; LBBB, left bundle branch block; LAD, left anterior descending; LCA, left coronary artery; LIMA, left internal mammary artery; LMCA, left main coronary artery; MPI, myocardial perfusion imaging; MR, mitral regurgitation; N/A, not available; OM, obtuse marginal artery; PCI, percutaneous coronary intervention; PDA, posterior descending artery; PLV, posterolateral ventricular branch; RBBB, right bundle branch block; RCA, right coronary artery; RI, ramus intermedius; SCD, sudden cardiac death; SPECT, single-photon emission computed tomography; SVG, saphenous vein graft; VAD, ventricular assist device.

TABLE 3 Characteristics of pediatric and adult patients with LMCAOA

	Pediatric patients	Adult patients
Total number	50	43
Associated symptoms		
Asymptomatic	5 (10%)	9 (20.9%)
Symptomatic	44 (88%)	34 (79.1%)
Heart failure	22	2
Syncope	14	1
Chest pain (angina)	12	21
Atypical chest pain		3
Palpitations	2	0
Cardiac arrest	5	3
Exertional dyspnea		6
Isolated	28 (56%)	21 (48.8%)
Associated with other lesions	22 (44%)	22 (51.2%)
Mitral valve prolapse	7	3
Supravalvar aortic stenosis or membrane	4	1
Bicuspid aortic valve with critical aortic stenosis	1	0
Valvar aortic stenosis	0	1
Subvalvar aortic stenosis	0	1
Severe aortic insufficiency	0	4
Right coronary artery stenosis	2	0
Anomalous origin from right coronary artery	1	0
Shone complex	1	0
Tetralogy of Fallot	1	0
Valvar pulmonary stenosis	1	0
Ventricular septal defect	2	0
Coarctation of aorta	1	0
Coronary atherosclerosis	0	9
Myocardial bridging	1	1
Patent ductus arteriosus	1	2
Apical aneurysm	1	0
Malignant pathway of collaterals	0	2
Ascending aorta aneurysm	0	1

5.5 | Survival

Two pediatric patients had unknown outcome. Of the remaining 48 patients, 81% ($n = 39$) survived and 19% ($n = 9$) died (Table 5). Forty-three percent ($n = 5$) of pediatric patients who did not have myocardial revascularization surgery ($n = 9$) died, including two with SCD. On the contrary, only 10% ($n = 4$) of pediatric patients who had revascularization surgery ($n = 39$) died. All of them had their surgery at the age of 6 months or less, three had preoperative severe LV systolic dysfunction but none of them had more than mild MR. The first patient

presented with heart failure at 3 months of age with severe LV systolic dysfunction and died in the third postoperative day after CABG. The second patient also presented with heart failure at 7 weeks of age with severe LV systolic dysfunction and died on the third postoperative day after CABG. The third patient presented with sudden cardiac arrest at the age of 3 days with severe LV systolic dysfunction and died immediately after CABG. The last patient presented with heart failure at 6 months of age and with akinesia of the anterior and lateral LV walls and died immediately after non-specified coronary surgery. Ten adult patients had unknown outcomes. Of the remaining 33 patients, 84.8% ($n = 28$) survived. Five adult patients died, three of them presented with SCD, and the other two were diagnosed by autopsy after non-cardiac death. About 66.7% ($n = 10$) of adult patients who had no revascularization surgery ($n = 15$) survived, but one of them required heart transplantation after 2 months. Interestingly, all of 18 adult patients who had revascularization surgery survived, even those with preoperative moderate or severe LV systolic dysfunction.

5.6 | Management and outcome of MR

Of the 34 pediatric patients with available echocardiographic results, 41% ($n = 14$) had MR at the time diagnosis. The MR was graded as severe in nine cases (six with mitral prolapse), moderate in one case and mild in four cases. Four of nine pediatric patients with severe MR had mitral valve repair in the same procedure of myocardial revascularization surgery, two of them required mitral valve replacement later, and one required a second mitral valve repair for ruptured chordae. The fate of MR in the fourth patient was unknown. Three of nine pediatric patients with severe MR had myocardial revascularization surgery without mitral valve repair. All the three patients survived with improved MR to mild degree in two and to moderate degree in one patient. Two of nine pediatric patients with severe MR had no revascularization surgery or mitral valve repair. One of them died 4 hours after coronary angiography was performed for diagnosis. The other one survived but the fate of MR was unknown. Unlike the pediatric population, only 3 of 13 adult patients had MR. It was graded as severe in two and moderate in one. Both patients with severe MR required myocardial revascularization surgery, and only one had mitral valve repair in the same procedure.

5.7 | Left ventricular systolic dysfunction and mortality

The LV systolic function was severely reduced in 12 pediatric patients, of which 9 had revascularization surgery. Post-surgery, the LV systolic function improved in four patients only. Regarding the association between the baseline LV systolic function and post-surgical mortality, three of nine pediatric patients with preoperative severe LV dysfunction died after their revascularization surgery and a fourth was noted to have regional wall motion abnormalities prior to surgery. The remaining three pediatric patients with severe LV systolic dysfunction did not have revascularization surgery. The first patient was diagnosed at the age of 2 years after presentation

TABLE 4 Investigation results in pediatric and adult patients with LMCAOA

	Pediatric patients	Adult patients
Patients with ECG at rest	34	16
Ischemic changes	17 (50%)	6 (37.5%)
No ischemic changes	17 (50%)	10 (62.5%)
Patients with available EST	10	13
Abnormal (ST depression)	8 (80%)	11 (84.6%)
Normal	2 (20%)	0
Equivocal	0	2 (15.4%)
Patients with available MPI	11	15
Perfusion defect(s)	8 (72.7%)	11 (73.3%)
Normal	3 (27.3%)	3 (20%)
Equivocal	0	1 (6.7%)
Coronary in echocardiography	34	13
Suspected LMCAOA	16 (47%)	0
Misdiagnosed as ALCAPA	9 (26.5%)	0
Misdiagnosed as anomalous LCA origin between RCC and NCC	0	1 (7.7%)
Reported as normal coronaries or not seen	9 (26.5%)	12 (92.3%)
Patients with known Left ventricular systolic function at diagnosis	37	22
Normal global LV function	16 (43.2%)	12 (54.5%)
Severe global LV dysfunction	12 (32.4%)	5 (22.7%)
Moderate global LV dysfunction	3 (8.1%)	3 (13.6%)
Mild global LV dysfunction	1 (2.7%)	1 (4.5%)
LV wall motion abnormalities	7 (18.9%)	1 (4.5%)
Patients with described size of collaterals in angiography	44	41
Multiple small-sized collaterals	31 (70.5%)	4 (9.8%)
Multiple moderate-sized collaterals	9 (20.5%)	4 (9.8%)
Large collateral(s)	4 (9%)	33 (80.5%)
Patients with described insertion of collaterals into LCA	14	23
To distal LAD or/and distal Cx	2 (14.3%)	7 (30.4%)
To mid and distal LAD ± distal Cx	6 (43.8%)	2 (8.7%)
To mid LAD	1 (7.1%)	9 (39.1%)
To proximal LAD or/and proximal Cx	4 (28.6%)	5 (21.7%)
To first diagonal branch	1 (7.1%)	0

Abbreviations: ALCAPA, anomalous origin of left coronary artery from pulmonary artery; Cx, circumflex artery; ECG, electrocardiogram; EST, exercise stress test; LAD, left anterior descending; LCA, left coronary artery; LMCAOA, left main coronary artery ostial atresia; LV, left ventricle; MPI, myocardial perfusion imaging; NCC, non-coronary cusp; RCC, right coronary cusp.

with heart failure and died 9 years later. The second one had Shone's complex repair at the age of 2 weeks and Ross-Konno at 6 months of age. This patient survived and his LV systolic function recovered. The third patient died few hours after balloon dilation of his critical aortic stenosis at 7 days of age.

In the adult population, the LV systolic function was moderately or severely impaired in eight patients, and half of them had revascularization surgery. Post-surgery, the LV systolic function improved in three patients and was unknown in one patient. Unlike the pediatric population, all adult patients who had revascularization surgery survived, even those with preoperative moderate or severe LV systolic function suggesting the possibility of a more robust collateral

circulation simply by the factor of having survived until adulthood. Regarding the four other patients who did not have revascularization surgery, three patients had improvement of LV function although two required aortic valve repair/replacement. The fourth patient had persistent severe LV systolic dysfunction that required a left ventricular assist device and heart transplantation.

6 | DISCUSSION

LMCAOA is a very rare congenital coronary artery anomaly with less than 100 cases reported in the literature to date. The overall

	Pediatric patients	Adult patients
Management		
Unknown	2	7
Sudden death	2	5
Medical management	7 (15.2%)	10 (32.3%)
Surgery for myocardial revascularization	39 (84.7%)	21 (67.6%)
CABG		
CABG using LIMA	20	5
CABG using SVG	2	7
CABG using both LIMA and SVG	5	4
Non-specified CABG	0	1
Coronary reconstruction surgery		
Using autologous Pericardial patch	3	
Using homograft patch osteoplasty	3	
Using azygous vein graft	2	
Non-specified coronary surgery	3	4
Outcomes		
Unknown	2	10
Patients who do not have surgery for myocardial revascularization		
Survived	4 (44.4%)	10 (66.7%)
Died	5 (55.6%)	5 (33.3%)
Patients who had surgery for myocardial revascularization		
Survived	35 (89.7%)	18 (100%)
Died	4 (10.3%)	0 (0%)

Abbreviations: CABG, coronary artery bypass grafting; LIMA, left internal mammary artery; SVG, saphenous vein graft.

mortality for this lesion is not insignificant with at least 18% in pediatric and 11% in adult published cases. Despite this, the optimal management strategy for this lesion is not clear. LMCAOA usually occurs in isolation, however, concomitant anomalies have been described in more than one-third of pediatric patients, with the most common lesion being mitral valve prolapse and supravalvular aortic stenosis. The most common presenting symptom in infants was congestive heart failure, whereas children older than 2 years of age presented primarily with syncope and chest pain. Unlike pediatric patients, adult patients presented predominantly with angina, which occurred in more than half of cases. The majority of patients in our review (~85%) developed symptoms as a consequence of myocardial ischemia and inability of the collateral circulation to cope with myocardial oxygen demands. Although the type and timing of these symptoms was variable, patients could be grouped into two broad categories—the “infantile-childhood type” where patients became symptomatic early in life, or the “adult type” where the patients remained asymptomatic, or at least survived into old age before developing symptoms.

There has been general agreement from previous literature reviews that myocardial revascularization is recommended for patients

TABLE 5 Management and outcomes in pediatric and adult patients with LMCAOA

with LMCAOA if they have symptoms.³ However, waiting for symptoms to develop does incur the risk for a SCD episode which occurred in 10% of pediatric patients and 7% of adult patients in our review. This needs to be balanced with the risks of surgical revascularization overall, and potential complications that will occur in some patients who may have had a benign course. With respect to our first case, the clinical decision not to intervene was challenging. One important factor that was considered was the presence of large collateral vessels. Tanawuttiwat and his colleagues³ have previously reported some differences in angiographic findings between “infantile-childhood” type of patients and those with “adult type.” About 70% of pediatric patients from our review had small-sized collaterals. In contrast, large collateral vessels, particularly conal and marginal branches from the RCA to LAD, were found in 80% of adult patients compared to 9% of pediatric patients. Additionally, in the pediatric population, the collaterals arising from the RCA tend to insert more distally in the left coronary system (54%) rather than in the proximal or mid LAD (35%). However, in the adult population, the collaterals tend to join the mid or proximal LAD (61%) more than the distal segments (39%).

Myocardial ischemia was present in most cases at time of diagnosis. Diagnostic confirmation has been made through selective

coronary angiography with or without non-invasive cardiac imaging in the majority of cases in both children and adults.

Nearly one-fourth of the pediatric patients were initially misdiagnosed as ALCAPA. In both situations, blood flow in the LCA is reversed. Association with ALCAPA has not been described and is in fact usually considered an important differential diagnosis as the RCA supplies the entire coronary circulation. ALCAPA typically features a dilated RCA, collateral channels between the right and left coronary arterial systems, and retrograde flow through the LCA reaching the pulmonary artery. In the third case of our series, with occlusion of the LMCA ostium, the RCA was only mildly dilated. The collateral circulation between the right and left coronary arterial systems were not as striking as usually observed, and the proximal LCA was very faintly visualized on the contrast enhanced CT scan and angiography. Taken together, the atypical findings seen on multiple imaging modalities are explainable and can be regarded as telltale signs for this unusual association of ALCAPA and LMCAOA. The diagnosis of LMCAOA requires a high index of suspicion because identification of this condition may provide preoperative clarity and alleviate the need for division of the pulmonary artery to look for ALCAPA.²

Giving that the majority of pediatric patients in published reports were symptomatic and with small collaterals, myocardial revascularization was performed in 85% of them. An interesting finding from our review was that prior to 2005, among the 13 pediatric patients who had myocardial revascularization, the surgeon's preference was for CABG (92%) rather than coronary reconstruction osteoplasty (8%, one case). However, this started to change after 2005 with the propensity toward coronary reconstruction osteoplasty, which was done in 40% of cases. Some previous case reports^{2,39} considered coronary reconstruction osteoplasty superior to CABG as it restores physiologic antegrade coronary flow pattern and is believed to carry more sufficient blood flow reserve to the myocardium of the LCA area. Furthermore, it avoids the potential drawbacks of coronary artery bypass grafting, which restores only retrograde perfusion of the myocardium proximal to the graft anastomosis and can result in competitive flow when the LMCA is stenotic. In addition, coronary reconstruction osteoplasty does not preclude percutaneous coronary intervention for the distal coronary tree if it is necessary at a later stage. Lastly, coronary osteoplasty preserves the internal mammary artery or saphenous vein graft for possible future use in the event of later development of atherosclerotic coronary artery disease.³⁵ Therefore, coronary osteoplasty is considered to be an excellent option for LMCAOA in pediatric age group. It is worth knowing that coronary osteoplasty is technically more challenging and requires presence of well-developed LMCA trunk compared to CABG.

Regarding the outcomes for LMCAOA, the survival for both children and adults after myocardial revascularization surgery is good.^{3,39} Nearly 45% of pediatric patients who were managed medically died compared to 10% of those who had revascularization surgery. The beneficial effect of revascularization surgery was clearer in the adult population with no deaths among those who had the surgery compared to 33% among those who had not. Interestingly, we noticed that a third of the nine pediatric patients with preoperative severe LV systolic dysfunction died after their revascularization surgery, however, the

function has improved or even normalized in all of those who survived the surgery. In addition, all pediatric patients whose preoperative LV systolic function was normal, mildly reduced or moderately reduced survived after revascularization surgery. Unlike the pediatric population, all adult patients who had revascularization surgery survived, even those with preoperative moderate or severe LV systolic dysfunction.

7 | CONCLUSION

A high index of suspicion is needed to diagnose LMCAOA because of its rarity and non-specific clinical presentation across the entire lifespan. It is important to consider LMCAOA as a differential diagnosis in patients with suspected ALCAPA. Myocardial revascularization can significantly reduce mortality in selected patients. Based on the currently available data, we recommend that it should be strongly considered in patients with at least one of the following findings: symptoms, evidence of ischemia on ECG or EST, myocardial perfusion defects on MPI, wall motion abnormalities or global LV systolic dysfunction on echocardiogram, severe MR or small-sized distal collaterals in coronary angiography. Conservative management may be preferred for those with no symptoms or evidence of ischemia and in the presence of good-sized proximal collaterals. There has been a shift toward performing coronary reconstruction osteoplasty rather than CABG over the last 15 years, as it restores a physiological antegrade coronary flow pattern. More studies are needed to assess the long-term outcomes of patients after myocardial revascularization and to aid in further clarifying which patients are best served by surgical revascularization vs conservative management.

CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest with the contents of the article.

AUTHOR CONTRIBUTIONS

Mahmoud Alsalehi contributed in drafting the first and second cases and prepared the literature review. Aamir Jeewa was involved in critical revision and review of the article. Andrea Wan was involved in critical revision and review of the article. Juan Contreras contributed in drafting the second case. Shi-Joon Yoo was involved in critical revision and review of the article. Jessica A. Laks contributed in drafting the third case and was involved in critical revision and review of the article.

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