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Surgical Repair of Congenital Atresia of the Left Coronary Ostium

Jacob Hartz, MD, MPH,[a] Meena Nathan, MD,[b] Jane W. Newburger, MD, MPH,[a] Luis Quinonez, MD[b]

ABSTRACT

Congenital left main coronary artery atresia is an exceedingly rare condition with potentially fatal consequences if not diagnosed in a timely fashion. We present a case series in children and adolescents, including surgical repair and outcomes. We describe the presenting symptoms and subsequent management of each patient, including surgical repair and outcomes. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:198–201) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Congenital left main coronary artery atresia (CLMCAA) occurs when the left main coronary artery (LMCA) does not have a patent connection to any cardiac chamber, systemic vessel, or pulmonary vessel and only receives its blood supply from collateral vessels arising from the right coronary artery (RCA). Although CLMCAA can occur in aortopathies, such as William's Syndrome (1), isolated CLMCAA is an exceedingly rare condition (2,3). The largest case series describes 5 patients, but only 2 had true ostial atresia and 3 had severe ostial stenosis (4). Previous reports suggest that infants usually present with ischemia and congestive heart failure (2,3), whereas ischemia and syncope are more common presenting symptoms in adolescents and adults (3,5). Although it can be demonstrated using echocardiography, CLMCAA is best delineated on cardiac computed tomography angiography (cCTA) (6) or coronary angiography (5).

LEARNING OBJECTIVES

- To understand that CLMCAA is a rare, potentially lethal anomaly that requires a high index of suspicion.
- To understand the role of cCTA or coronary angiography in defining the coronary artery anatomy.
- To determine the optimal surgical approach based on the length of the atretic segment.

Whereas timely diagnosis and initial medical management are key to the stabilization of patients with CLMCAA, surgical intervention to establish unobstructed antegrade flow remains the gold standard (1). A variety of approaches have been proposed to create continuity between the LMCA and a systemic vessel, including: 1) using a saphenous vein or left internal mammary artery (LIMA) bypass graft; to the left anterior descending artery (LAD), or circumflex; 2) reconstructing LMCA using the arterial wall; 3) coronary ostial plasty; or 4) a combination of these approaches (3). We describe the treatment and outcomes of this rare condition in 5 patients at a pediatric tertiary center. Our single-center, retrospective review included all patients with CLMCAA seen at Boston Children's Hospital between 2008 and 2019. Patients were excluded if they had aortopathies or had a previous cardiac surgical procedure in which

From the [a]Department of Cardiology, Boston Children's Hospital, Boston, Massachusetts, USA; and the [b]Department of Cardiovascular Surgery, Boston Children's Hospital, Boston, Massachusetts, USA.

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the coronaries were manipulated, such as repair of transposition of the great arteries or palliated single ventricle. Institutional review approval with waiver of consent was obtained before review of departmental databases and medical records for data on clinical presentation, imaging, treatment, and outcome.

HISTORY OF PRESENTATION

Five children were identified with a diagnosis of CLMCAA who met our inclusion criteria. Their demographic information, presenting symptoms, imaging findings, and outcomes are presented in **Table 1**. Patient #1 was a 20-month-old female who presented for evaluation of a murmur appreciated at a previous well-child evaluation. Patient #2 was an 18-year-old male with trisomy 21 from the United Arab Emirates who presented with dyspnea on exertion, syncope, and weight loss. Patient #3 was a 9-year-old girl who presented to an outside hospital with syncope, hypotension, and diffuse ST-segment changes. During fluid resuscitation, she developed severe pulmonary edema. Patient #4 was a 9-year-old boy who also presented with syncope that first occurred at age 5 years. Patient #4 was referred to a pediatric cardiologist shortly thereafter for evaluation of a murmur. Over the next 4 years, he had 3 additional syncopal episodes. Patient #5 was a 6-month-old male who was admitted during an acute viral illness with persistent tachypnea and suspected viral myocarditis.

MEDICAL HISTORY

Except for Patient #2, who had a diagnosis of trisomy 21, the past medical history was unremarkable, except for that described in the History of Presentation.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for our cohort included a mitral valve abnormality (Patient #1), hyperthyroidism (Patient #2), myocarditis (Patients #3 and #5), arrhythmia (Patient #4), and anomalous left coronary artery from the pulmonary artery (Patient #5).

INVESTIGATIONS

All patients underwent an echocardiogram. For Patient #1, initial echocardiograms demonstrated a trileaflet mitral valve with severe mitral insufficiency and moderate mitral stenosis. There was mild-to-moderate dilation of the left ventricle and left atrium, but function was preserved. The coronaries were not described. It was only after a post-operative arrest after mitral valve repair was the diagnosis of CLMCAA made using cardiac catheterization. In a review of the pre-operative echocardiograms, including transesophageal echocardiograms, the coronary anomaly could not be identified. The initial echocardiograms for Patient 2 demonstrated a bicuspid aortic valve, ventricular dilation, and dysfunction, which were attributed to a thyroid disorder. A small coronary fistula was suspected as well. Patient #2 underwent a cardiac catheterization because his function did not improve after treatment with carbimazole, which suggested CLMAA. The diagnosis of CLMCAA subsequently was confirmed using cCTA. The patient also underwent a dobutamine stress test, which demonstrated dyskinesia in the territories of the LAD and left circumflex arteries. For Patient #3, an echocardiogram obtained after her presentation to an outside facility was consistent with acute congestive cardiomyopathy, which led to placement of an Impella left ventricular assist device (Abiomed, Danvers, Massachusetts) before being transferred to our facility for further management. A repeat cardiac catheterization at our facility to evaluate the function of the Impella demonstrated CLMCAA (Video 1).

Patient #4 had an echocardiogram at the time of his first syncopal episode and was found to have a mildly abnormal mitral valve with mild mitral insufficiency. He also had a LINQ device (Medtronic, Minneapolis, Minnesota) placed, which revealed ST-segment changes and biphasic T waves consistent with ischemia. A repeat echocardiogram could not identify the LCA origin and demonstrated retrograde flow into the LAD with the RCA size at the upper limits of normal. A cCTA demonstrated CLMCAA. Patient #5 initially had an echocardiogram that demonstrated severe mitral regurgitation and left ventricular dysfunction. Because an echocardiogram could not demonstrate a LMCA, he underwent cardiac catheterization and a cCTA, both of which demonstrated CLMCAA.

MANAGEMENT

The diagnosis of CLMCAA led to prompt surgical correction in all cases. Patients #1, #2, #3, and #4 were repaired with a LIMA to LAD bypass graft. Patient #5 underwent repair with a left main coronary ostial plasty and mitral valve repair.

DISCUSSION

We describe the largest case series of 5 patients with CLMCAA, who ranged in age from 20 months to 18 years at presentation. In each patient, the diagnosis

ABBREVIATIONS AND ACRONYMS

cCTA = cardiac computed tomography angiography

CLMCAA = congenital left main coronary artery atresia

LAD = left anterior descending artery

LIMA = left internal mammary artery

LMCA = left main coronary artery

RCA = right coronary artery

TABLE 1 Presenting Characteristics, Imaging, and Operations of Patients With Left Main Coronary Artery Ostial Atresia

Patient #	Age, Sex	Presentation and Initial Symptoms	Initial Echocardiogram	Catheterization/cCTA	Operative Findings Surgical Repair	Outcomes
1	20 months, female	Murmur	Severe MR and moderate MV stenosis, trileaflet MV, mild-to-moderate dilation of LA and LV, normal function; coronaries not described	Cath (after cardiac arrest): n antegrade filling of LCA, large RCA, multiple small collaterals from RCA, and severe biventricular dysfunction.	LIMA to LAD, MV plasty; MV noted to have 3 papillary muscles, cleft in posterior leaflet, and prolapse of anterior leaflet	Death
2	18 yrs, male	Dyspnea on exertion, weight loss, possible syncope; transferred from outside hospital for surgical evaluation	Ejection fraction of 50%	Cath: ostial atresia, bidirectional flow in distal RCA, hypokinesis, decreased function, 1.7 cm from occlusion to aorta on cardiac magnetic resonance imaging	LIMA to LAD	Normal LV size and function 8 months after repair
3	9 yrs, female	Syncope (possibly recurrent), mental status changes; concern for myocarditis; cardiac arrest	Decreased function with concern for myocarditis; initial reports from echocardiogram not available, but no discussion of concerns for coronary anomalies in transfer notes	Cath: no LCA; collateral vessels arising from large RCA	LIMA to LAD Dimple at LCA ostia with no evidence of inflammation	Low-normal LV function 2 yrs after event; remains on β-blocker and aspirin
4	9 yrs, male	Recurrent syncope	Mildly abnormal MV and mild MR, RCA size is upper limits of normal; repeat echo suggested LCA artery arising from right sinus	Cath: LCA ostial occlusion, collaterals; normal LVEDp; cCTA: LMCA atresia; mildly dilated RCA	LIMA to LAD Occlusion approximately 1.3–1.5 cm from aorta	Normal LV size and function
5	6 months, male	Viral illness with cardiomegaly on CXR, concern for myocarditis	Several views suggestive of LCA arising from pulmonary artery, severe MR, bright PM, normal RCA dimensions, severe LA dilation	Cath: near atresia of ostia; 6 mm distance from aorta; robust collateral circulation	LCA osteoplasty with thin-walled pulmonary artery homograft patch, MV repair, cryoablation for ectopic atrial tachycardia	Required reoperation for recurrent MR 3 weeks postoperatively; normal LV size and function 2 weeks after repair

cath = cardiac catheterization; cCTA = cardiac computed tomography angiography; CXR = chest radiograph; LAD = left anterior descending artery; LA = left atrium; LCA = left coronary artery; LIMA = left internal mammary artery; LV = left ventricle; LVEDp = left ventricular end diastolic pressure; MR = mitral regurgitation; MV = mitral valve; RCA = right coronary artery.

was made using either cCTA or coronary angiography, rather than on echocardiography. In 3 of the 5 cases, there was suspicion for a coronary anomaly, but not for CLMCAA. In the remaining 2 cases, the coronary anatomy was not diagnosed until after coronary angiography. After surgical correction, all but 1 patient survived, despite ventricular dysfunction in all at presentation, including 1 patient who required the use of a ventricular assist.

In each of these cases, the patient's management was aided by advanced imaging modalities that expose the patient to ionizing radiation. Although echocardiography eliminates the exposure to ionizing radiation, it may not sufficiently delineate the coronary anatomy and delay diagnosis. Although a cCTA and cardiac catheterization expose patients to ionizing radiation, recent recommendations on the management of coronary anomalies acknowledge that these tests may be necessary and should not be delayed in suspected cases (7).

Surgical correction remains the gold standard and not only improves/prevents symptoms, but more importantly prevents sudden death and preserves myocardial viability and mitral valve function (3). Despite the presence of collaterals from the RCA, myocardial demand will eventually outstrip the ability of collaterals to provide adequate myocardial perfusion in the left coronary distribution, as evidenced by the variable age of presentation in our series and other reports (1,3,4,6).

FOLLOW-UP

Patient #1 died of multisystem organ failure despite successful revascularization. Patient #2 has limited follow-up information available because he does not live domestically. He was discharged with mildly decreased but improving left ventricular function, but long-term follow-up is unavailable. Follow-up for Patient #3 included a cardiac catheterization 2 years after the procedure, which found an unobstructed graft with antegrade filling of the left circumflex and distal LAD (Video 2). At 3 years, she had complete resolution of symptoms. However,

she remained on aspirin and carvedilol because of low-normal function with mild hypokinesis of the left ventricular apex and mild mitral insufficiency. Patient #4 also was asymptomatic and a postoperative stress echocardiogram demonstrated normal left ventricular function with excellent augmentation with physical activity and no wall motion abnormalities. Only short-term follow-up is available for Patient #5 because of his recent presentation, but echocardiograms before discharge demonstrated mild mitral stenosis, moderate mitral insufficiency, and normal left ventricular size and function.

In 4 patients a coronary artery bypass was performed using the LIMA to LAD coronary artery, whereas the youngest patient had a LMCA ostial plasty. Among the 4 patients in whom coronary artery bypass grafting was performed, the atretic main coronary artery segment was long and precluded an ostial procedure. Intraoperative assessment of the length of the atretic segment is important because contrast may not adequately fill the LMCA through collaterals. In severe cases, the LMCA may be completely atretic extending onto the bifurcation of the LAD and circumflex artery. Ostial plasty is the procedure of choice when the length of the atretic segment is relatively short (8), whereas bypass graft with an arterial conduit should be performed for long-segment atresia. It is particularly important to have periodic imaging of the coronary arteries for early detection and management of postoperative coronary occlusions and/or bypass graft failures.

CONCLUSIONS

Despite the small number of cases with CLMCAA in this single-center series, several important lessons can be drawn. First, CLMCAA presents at a range of ages and with a variety of signs and symptoms. Second, the inability to consistently define the coronaries with echocardiography warrants further investigation with either a cCTA or coronary angiography. Third, although an ostial procedure is our preferred surgical treatment, it may not be possible if the length of the atretic segment is too long. In the remainder, a LIMA to LAD graft has resulted in excellent short-term outcomes.

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ADDRESS FOR CORRESPONDENCE: Dr. Luis Quinonez, Department of Cardiac Surgery, Boston Children's Hospital, 300 Longwood Avenue, Boston, Massachusetts 02115, USA. E-mail: Luis.Quinonez@cardio.chboston.org.

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APPENDIX For supplemental videos, please see the online version of this paper.