

# A rare case of left main coronary artery atresia misdiagnosed as an anomalous left coronary artery from the pulmonary artery and presented as dilated cardiomyopathy

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## ABSTRACT

Left main coronary artery atresia is one of the rarest congenital anomalies characterized by the absence of the left coronary ostium and the left main trunk. This case report presents an extremely rare left main coronary artery atresia case in a 14-week-old female infant presenting with severe symptoms of dilated cardiomyopathy. The patient underwent surgery after successful weaning from extracorporeal membrane oxygenation (ECMO) support but died because of cranial complications after the second run of ECMO.

**Keywords:** Bland-White-Garland syndrome, cardiac surgery, congenital heart disease, extracorporeal membrane oxygenation, left main coronary artery disease.

Coronary artery anomalies are relatively uncommon congenital disorders of the coronary artery anatomy. The incidence of coronary artery anomalies has been reported to be 0.6-1.3%.<sup>[1]</sup> Left main coronary artery (LMCA) atresia (LMCAA) is the rarest form of congenital coronary malformations, in which the coronary ostium and the main trunk in the left coronary artery (LCA) system are absent.<sup>[2]</sup> Thus, blood flows through collateral vessels from the right coronary artery (RCA) to the LCA.<sup>[3]</sup> Clinical presentation, management, and prognosis of this disease depend on the characteristics of the collaterals and native vessels.<sup>[4]</sup> Herein, we report a case of congenital LMCAA, misdiagnosed as an anomalous LCA from the pulmonary artery (ALCAPA).

## CASE REPORT

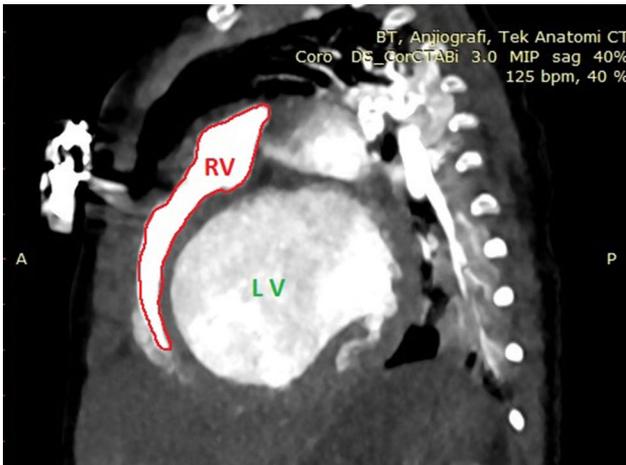
A 14-week-old and 7 kg female infant, who had respiratory distress worsened after bronchiolitis, was admitted to another hospital. According to the echocardiographic examination in this center, the left atrium and left ventricle were greatly enlarged and left ventricular contractions were markedly decreased (shortening fraction: 13%), indicating dilated cardiomyopathy. A moderate degree of regurgitation was observed in the mitral and tricuspid valves. Pulmonary arterial pressure was measured to be approximately 60 mmHg through the tricuspid valve

regurgitation. Since the patient had cardiogenic shock, intravenous immunoglobulin, hydrocortisone, and inotropic support drugs were started with the preliminary diagnosis of myocarditis. The patient was placed on venoarterial extracorporeal membrane oxygenation (ECMO) due to the low cardiac output leading to multiorgan failure using neck vessels. After 10 days of ECMO support, the patient was successfully weaned and separated from ECMO support. After three days of ECMO decannulation, both coronary multidetector computed tomography (MDCT) and cardiac catheterization was performed with a suspicion of ALCAPA syndrome. Figure 1 represents the right ventricle stuck between the sternum and the left ventricle. A hugely dilated left atrium and the left ventricle can be seen in Figure 2. Furthermore, Figure 2 shows the RCA and the absence of the left coronary ostium. The video of catheterization shows the retrograde flow of the LCA (Video 1). The patient was referred to our hospital for corrective surgery.

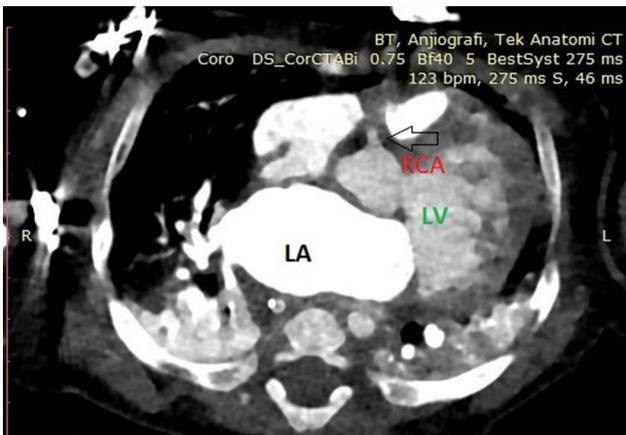
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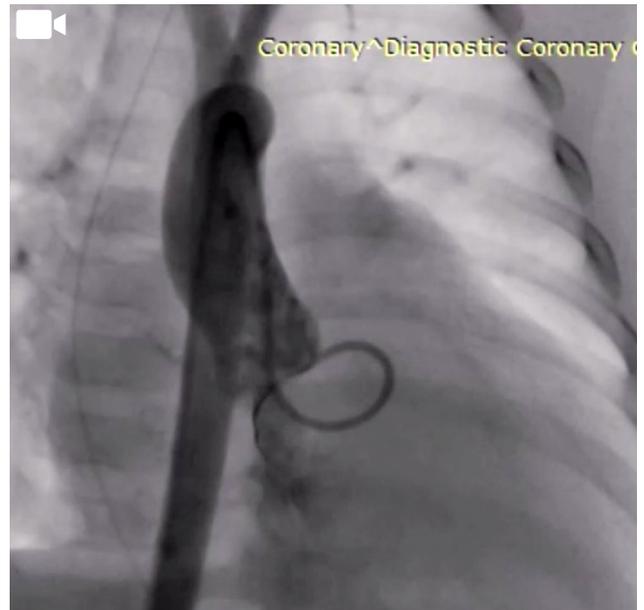


**Figure 1.** The right ventricle stuck between the sternum and the left ventricle.



**Figure 2.** Hugely dilated left atrium and the left ventricle, the right coronary artery, and the absence of the left coronary ostium.

The patient had small ischemic lesions on the left frontoparietal, left thalamus, and basal ganglia on a computed tomography scan, which was evaluated by a pediatric neurologist. The patient underwent surgery under anticonvulsant treatment according to the recommendations of the pediatric neurologist. The patient could be referred for left ventricular assist device implantation or the transplantation list because of dilated cardiomyopathy. However, the patient was not in a position to wait long on the transplantation list. In addition, the need for lifelong anticoagulation treatment under ventricular assist device support could increase the risk of cerebral events. Therefore, we preferred surgical correction.



**Video 1.** The retrograde flow of the left coronary artery.

Although the origin of the LCA from the aorta or retrograde blood flow from the left anterior descending artery (LAD) to the pulmonary artery could not be visualized, the patient underwent an operation for LCA revascularization, with the possible preoperative diagnosis of ALCAPA. After a midline sternotomy and heparinization, aortic and bicaval cannulation was performed to initiate cardiopulmonary bypass. Diastolic cardiac arrest was provided with 30°C systemic hypothermia and antegrade tepid blood cardioplegia after cross-clamping. Tepid blood cardioplegia was repeated every 20 min until releasing the cross-clamp. The left side of the heart was vented via the patent foramen ovale. Pulmonary arteriotomy and aortotomy were performed. There were no left coronary ostia in the pulmonary artery or in the aorta.

The circumflex artery and LAD were visible but small in diameter. They had merged into a small blind pouch 2 cm away from the left coronary sinus of the aorta. There was no possibility for preparation and anastomosis to the aorta due to the tiny diameter and fragile vessel. The left internal mammary artery (LIMA) was harvested. The LIMA was not spastic, and its flow was sufficient. The LAD, which is located posterior to the pulmonary artery, just distal to the atretic ostium of LMCA, was opened, and a LIMA-LAD anastomosis was performed with 8-0 prolene. Pulmonary artery and aortotomy were

closed. The cross-clamp was removed after deairing. The patient was weaned from cardiopulmonary bypass with moderate inotropic support doses, including adrenaline, noradrenaline, and milrinone. The sternum was left open due to hemodynamic instability. Cardiopulmonary bypass time was 141 min, and cross-clamp time was 101 min.

According to the echocardiographic examination on the first postoperative day, the left ventricular shortening fraction was 15%, and there was a moderate degree of mitral regurgitation. The patient needed ECMO support due to the low cardiac output on the second postoperative day. The patient died of a massive cranial hemorrhage under ECMO support, documented with both head ultrasonography and a computed tomography scan, despite improving ventricular function (last measurement of shortening fraction: 20%) on the 10<sup>th</sup> postoperative day.

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## DISCUSSION

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Left main coronary artery atresia is a rare condition with unclear etiology. In LMCAA, the left coronary system receives blood only from collateral arteries from the RCA. Thus, the heart may eventually be unable to cope with collateral circulations and develop myocardial ischemia. Patients mostly present with nonspecific symptoms depending on their age group and the formation of collateral vessels from the RCA to the LCA. Adult patients usually present with angina pectoris at an advanced age when collateral flow cannot keep pace with myocardial demands. Children and adolescents often present with chest pain, dyspnea, syncope, tachyarrhythmia, and sudden cardiac death.<sup>[2,3]</sup> Infants mostly present with growth retardation and myocardial infarction.<sup>[2]</sup> Catastrophic situations like sudden cardiac arrest, low cardiac output, and cardiomyopathy, as seen in our case, can also be the first symptoms of infants with LMCAA.<sup>[5,6]</sup> Since these symptoms are not specific to LMCAA, the clinical diagnosis might be neglected. Thus, other coronary anomalies should be excluded to reach a correct diagnosis. As stated in a retrospective study by Yildiz et al.,<sup>[1]</sup> LMCA was the most common anomalous vessel. Separate origins of LAD and circumflex artery from the left coronary sinus of Valsalva were the most common anomaly and should be excluded in patients presenting with symptoms of myocardial infarction. An ALCAPA is one situation that LMCAA can be confused with and should be differentiated from.<sup>[2,7]</sup>

Although congenital atresia of the LMCA usually occurs as an isolated cardiac lesion, concomitant anomalies including bicuspid aorta, supra-avalvular aortic stenosis, right coronary ostial stenosis, pulmonary stenosis, ventricular septal defect, and mitral valve prolapsus secondary to myocardial ischemia can be encountered.<sup>[4,8,9]</sup>

The diagnosis of LMCAA can be done by coronary angiographic findings, which usually show no left coronary ostium and LCA filled in a retrograde manner via the RCA instead of antegrade blood flow. In recent years, MDCT has also played an essential role in diagnosing LMCAA in older children and adults and can be used in patients suspected of congenital coronary artery abnormalities.<sup>[3,10]</sup> According to some researchers, MDCT provides more precise details in a less invasive way than coronary angiography and is thus recommended to evaluate congenital coronary abnormalities.<sup>[11]</sup> Multidetector computed tomography not only defines the anatomic course and the ostium shape but also has no complications as coronary spasm than conventional coronary angiography. However, there is no reliable research reporting the use of MDCT in infants with coronary anomalies.<sup>[9]</sup> The physician may merge findings that are revealed by transthoracic echocardiography examination and MDCT or cardiac catheterization. The absence of retrograde filling from LAD to the pulmonary artery may support the possible diagnosis of LMCAA.

The prognosis of LMCAA is poor.<sup>[2]</sup> Due to the symptomatic nature of LMCAA and the risk of sudden cardiac death, patients in the pediatric population with LMCAA should undergo surgical intervention to restore the antegrade flow to the left coronary system. Various surgical interventions have been described, and coronary artery bypass grafting using the internal mammary artery or the saphenous vein has been identified as the treatment of choice, regardless of the caliber of left-sided vessels.<sup>[4,8,12]</sup> A LIMA graft for LMCAA appears to be a reasonable early interventional approach with successful results one year postoperatively. Nevertheless, due to the rarity of this disease, long-term postoperative outcomes have not been reported.<sup>[2,4,5,8,9,11]</sup> Alternative to bypass grafting, direct surgical reimplantation or reconstruction of the LMCA using the azygos vein has been described to provide the shortest and most efficient way for blood to the myocardium.<sup>[6,12,13]</sup> The advantages of direct surgical reimplantation relative to bypass grafting is to provide antegrade

flow without a bypass material. However, direct surgical reimplantation could not be performed in small infants with LMCAA. The long-term results of bypass grafting in pediatric patients are reasonable and good results have been reported before.<sup>[12,14]</sup>

In conclusion, LMCAA is an extremely rare congenital coronary anomaly in which the left coronary ostium and the left main trunk in the LCA system are absent. Considering the severe symptoms, such as myocardial infarction or sudden cardiac death, surgical revascularization should most likely be the treatment choice.

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

**Author Contributions:** Idea/concept, writing the article: S.B., B.K., E.E.; Design, data collection and/or processing, analysis and/or interpretation, literature review, references and fundings; materials: S.B., B.K.; Control/supervision, critical review: E.E.

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