## Case Report

# *Rare Form of Coronary Artery Anomalies in Adults*

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## ABSTRACT

The congenital atresia of the left main coronary artery is a very rare form of coronary anomalies with poor clinical outcomes if left untreated. Patients require surgical correction with coronary artery bypass grafting after the diagnosis. Here, we report a case of the congenital left main atresia in a 36-year-old woman who had a previous heart surgery with this anomaly having gone undetected. ***(Iranian heart Journal 2018; 19(3): 74- 76)***

**KEYWORDS:** Congenital heart disease, Left main atresia, Coronary abnormality

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The left main coronary artery atresia (LMCAA) is a rare form of coronary anomalies in which the left coronary ostium is absent and has a blind aortic end. The blood flow to the left heart chambers is provided by collaterals from the right coronary artery. The prognosis is poor due to the occurrence of myocardial ischemia and ensuing myocardial dysfunction and arrhythmias. [1,2] The management in adults consists of surgical revascularization with arterial and venous bypass grafts placed on the left anterior descending (LAD) and left circumflex ($LC_X$) arteries. In the present case, we report a patient with the congenital LMCAA accompanied by subvalvular aortic stenosis not detected at the time of her first operation. [3-5]

## CASE REPORT

A 36-year-old woman referred to the adult congenital heart disease clinic for follow-up. The patient complained of atypical chest pain episodes. She had a history of the surgical repair of subvalvular aortic stenosis at childhood, and there was no record of coronary anomalies detected during the previous surgery. On physical examination, there was a grade III/VI systolic murmur at the left sternal border. Echocardiographic evaluations revealed the recurrence of severe subvalvular stenosis and severe aortic regurgitation. The patient underwent cardiac catheterization, during which an ectatic and tortuous right coronary artery filling the left system retrogradely was visualized. Attempts to engage the left main coronary artery failed (Fig. 1). Coronary computed tomography angiography (CTA) with a 3D reconstruction was performed, and it showed that the left main ostium was atretic, confirming the diagnosis of the congenital LMCAA (Fig. 2).

The patient was scheduled for cardiac surgery, during which she received a saphenous vein graft on the $LC_X$ and an internal mammary artery graft on the LAD. The subvalvular stenosis was also repaired, and the aortic valve was replaced. She had an uneventful recovery.

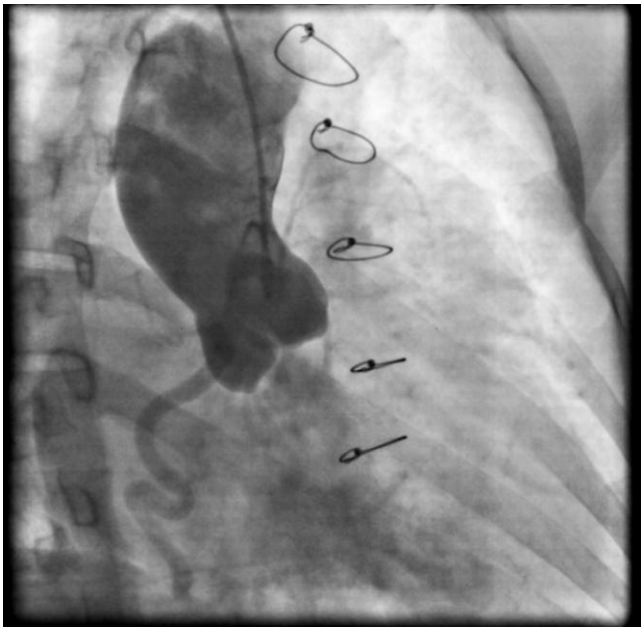

**Figure 1.** No filling of the left system during the aortic root injection

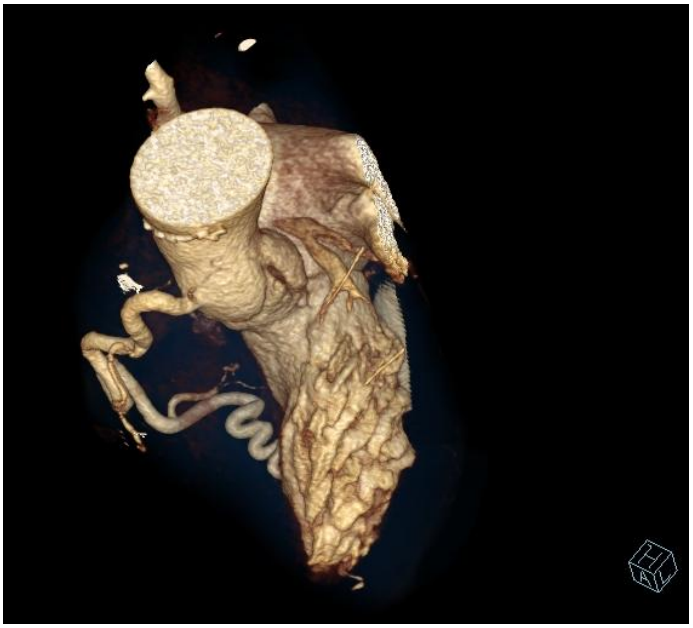

**Figure 2.** Tortuous right coronary artery and a blind left main coronary artery ostium

## DISCUSSION

The clinical diagnosis of the LMCAA might be neglected due to the wide range of nonspecific presenting symptoms—including chest pain in a young patient, dyspnea, and syncope—and aborted sudden cardiac death. [2,6-8] Patients could remain asymptomatic for a long time due to a well-developed collateral system. The LMCAA is often an isolated anomaly; however, associations with other congenital heart defects such as pulmonary stenoses, bicuspid aortic valves, supravalvular aortic stenoses, and ventricular septal defects have been described. [2,6] Selective coronary angiography is helpful, but multislice CTA can provide more precise details in a less invasive manner and is, therefore, recommended for the evaluation of congenital coronary abnormalities.[9]

Patients with the LMCAA should undergo surgical correction with the restoration of the antegrade flow to the left coronary system. Nevertheless, there are no guidelines on the management due to the scarcity of the condition. [10]

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