

# Congenital Combined Atresia of the Left Main Coronary with Supravalvar Aortic Stenosis

## Abstract

Congenital atresia of the left main coronary artery (LMCA) is an exceedingly rare phenomenon, and in the most of them, coronary artery bypass graft is required. We here describe a rare case of this anomaly that concomitantly was associated with supravalvar aortic stenosis and coronary–pulmonary fistula without the presence of conventional collateral circulation in a 16-year-old boy. The patient was admitted to our center with chest pain and dyspnea. Echocardiographic examinations showed supravalvar aortic stenosis with normal function of the aortic valve. Coronary angiography revealed atresia of LMCA with poorly developed left anterior descending coronary artery and well-developed circumflex coronary artery and diagonal artery that perfused by dominant and lengthy right coronary artery. The patient underwent coronary artery bypass grafting with repair of supravalvar aortic stenosis. The postoperative course was uneventful. The 6-month follow-up revealed normal diameter of the ascending aorta with symptomatic relief of preoperative chest complaint.

**Keywords:** Congenital heart disease, coronary artery bypass, supravalvar aortic stenosis

## Introduction

Congenital absence of the left main coronary artery (LMCA) or its ostium is an exceedingly rare congenital heart anomaly. The incidence of all types of coronary artery anomaly varies between 0.01% and 0.04% in forensic medicine following autopsy formation.<sup>[1-3]</sup> Only a few patients with a combination of LMCA atresia and other cardiac anomaly were reported, and indeed, the concomitant presence of supravalvar aortic stenosis and left coronary artery ostial stenosis is an exceedingly rare phenomenon.<sup>[4-6]</sup> Sometimes, atresia of the ostium or the left main trunk can be associated with congenital aortic valve stenosis, bicuspid aortic valve, mitral valve prolapse, and rarely supravalvar aortic stenosis.<sup>[7]</sup> Our patient has four unique properties that rarely reported in the medical literature.<sup>[8]</sup> The first special characteristic was large and well-developed right coronary artery (RCA) that running from distal posterior descending artery to the apex and then to interventricular septum and connected to the large diagonal artery. The second characteristic was the absence of collateral circulation as seen in the most cases of LMCA. The third property has

referenced to the presence of supra-aortic valve stenosis (SVAS). The fort features were the presence of some collateral vessels from RCA to main pulmonary artery as a fistula.<sup>[9]</sup>

## Case Report

A 16-year-old male admitted to our center, with typical chest pain and electrocardiography (ECG) finding of anterolateral wall ischemia. He had an exercise-induced respiratory problem and dyspnea from 3 years earlier that his previous diagnostic workup as transesophageal echocardiography (TEE) was unremarkable; however, ECG was not found in his document that obtained in his record. Physical examination was unremarkable. ECG showed ST elevation in lateral leads and in V1 to V6 and aVL at the anterolateral wall. Chest radiography showed moderate enlargement of the cardiac silhouette with a normal cardiothoracic ratio. The blood gas examination was normal. Echocardiography revealed a moderately dilated left ventricular dimension with 50 mm, with no evidence of mitral regurgitation, hypokinesia of anterolateral wall, and reduced left ventricular ejection fraction (ejection fraction, 40%). TEE

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also showed supra-valvar aortic stenosis (gradient of 67.5 mmHg) that confirmed by angiography. ECG showed ST elevation and Q wave in V1 to V6 and aVL at the anterolateral wall. TEE and ECG suggested coronary artery disease, and therefore, angiography was planned. Thallium 201 scintigraphy during exercise revealed a persistent ischemia in anteroseptal and lateral segments; angiography revealed the lengthy and dilated RCA, originated from normal site of the right coronary sinus. The origin of the LMCA was atretic and was not found in the left coronary sinus, and the left circumflex artery (LCX) and diagonal branches of the left coronary artery had normal size, especially diagonal artery was a well-developed artery and connected to RCA but left anterior descending (LAD) was small, thin with <1 mm diameter [Figure 1]. The LMCA, poorly developed LAD, normal LCX, and well-developed D artery were perfused retrogradely by abnormally dilated and tortuous RCA, in the absence of collaterals and looped branches as reported in the other studies [Figure 2]. In opposed to others, our case had an abnormal collateral flow into the pulmonary artery as a small fistula. The patient was scheduled for coronary artery bypass grafting (CABG) and repair of supra-valvar stenosis. Due to ischemia in anterolateral wall, the induction was induced by sufentanil and cisatracurium. The patient was scheduled first for off-pump coronary artery bypass (OPCAB) surgery. Anesthesia was induced with sufentanil and atracurium and maintained by propofol midazolam, and sufentanil. The lungs ventilation was continued with 60% oxygen and air, so the arterial saturation was maintained between 97% and 98%. A central venous line through the right subclavian vein was inserted without any problem. OPCAB was performed through midline sternotomy and LIMA harvesting. Then, CABG was performed with saphenous venous graft and LIMA graft was put on LCX and LAD, respectively [Figure 3]. During OPCAB, the mean blood pressure was maintained at a level of 80 mmHg or more, by using alpha receptor stimulation by phenylephrine; however, a short time of hypotension (3 min, 60/40 mmHg) was documented during bypass of the left circumflex coronary artery. Blood gas analysis, such as Pao<sub>2</sub> and Paco<sub>2</sub>, was maintained to a normal level. On admission to the Intensive Care Unit, the anesthetic agents were discontinued, and with the absence of hemodynamic instability or active bleeding or serious arrhythmias and when the patient became awake, he weaned from the mechanical ventilator. During the weaning, his hemodynamic status was stable, and he softly adapted the weaning process. The amount of postoperative drainage during the first, second, and third day of operation was 700 ml, 400 ml, and 200 ml subsequently. The volume of transfusion was 1200 mL of packed blood cells. The patient was conscious at the following morning, without any complaint of motor and sensory deficit in his lower extremities. No any wall motion abnormality detected during induction, during surgery,

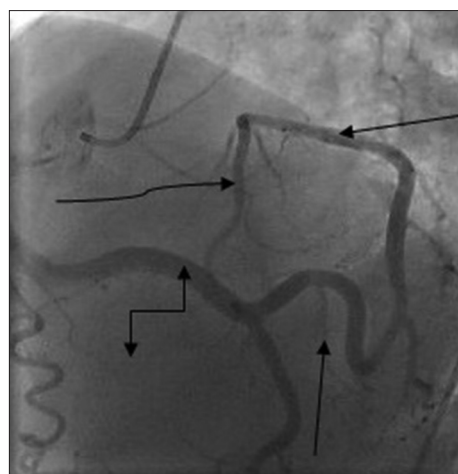


Figure 1: Diminutive left anterior descending (vertical arrow) and left circumflex artery (curve arrow) and well-developed right coronary artery (bi-headed arrow) connected to D1 (transverse arrow)

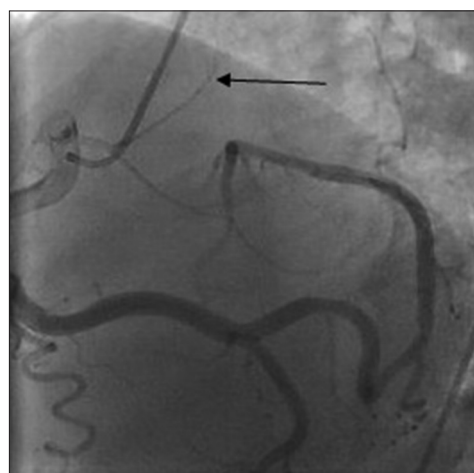


Figure 2: Collateral to pulmonary artery (black arrow)

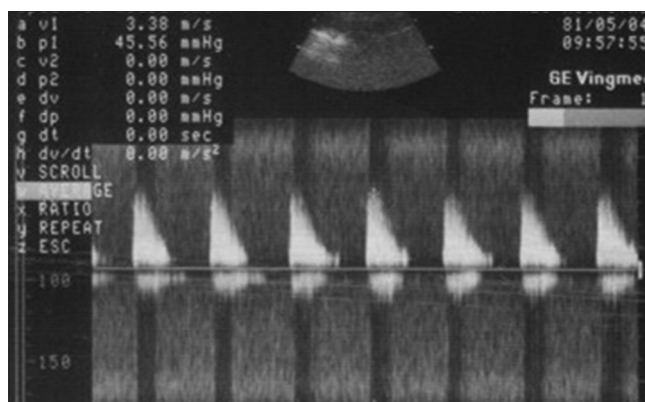
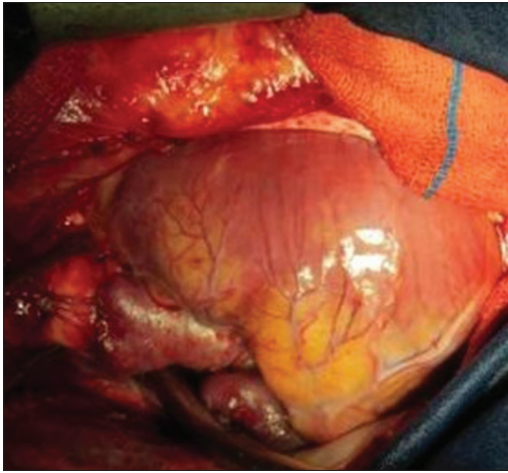


Figure 3: Gradient of 67.4 mmHg across the supra-valvar aortic stenosis

and postbypass period. After OPCAB, cardiopulmonary bypass was instituted with aortic and bi-caval cannulation. Following the aortic cross-clamping, transverse aortotomy with extension to both right and noncoronary sinuses was performed, and pantaloon-like defect was repaired with fresh pericardium [Figures 4]. Then, CABG was performed



**Figure 4: Repair of supra-valvar aortic stenosis by fresh pericardial patch (black arrow)**

with saphenous venous graft and LIMA graft was put on LCX and LAD, respectively [Figure 3]. Postoperative course was uneventful, and the patient was discharged on the 7<sup>th</sup> day of operation. One-year follow-up showed relief of his preoperative symptoms, and TEE revealed a normal size of ascending aorta with normal function of the aortic valve. To the best of our knowledge, concomitant diagnosis of LMCA with congenital stenosis of the ascending aorta and survival without collateral arteries with the presence of only dominant RCA has not been previously reported.

## Discussion

Any genetic defect that contributes to poor formation of primitive coronary artery may be associated with a defect in the muscular wall of the aorta as concentric stenosis revealed as a sand hour defect.<sup>[8]</sup> The combination of supra-valvar aortic stenosis and coronary-pulmonary fistula aggravates symptoms of congestive heart failure or ischemic change in the left main atresia. Despite the presence of these combinations, confession of signs and symptoms was delayed due to the presence of abnormal and lengthy and dominant RCA that connected directly to diagonal artery. However, the clinical presentation of infantile type is stormy and is associated with congestive heart failure that in many cases, it has been diagnosed as a congenital cardiomyopathy. The clinical suspicion in infantile type based on sole clinical finding is difficult because the symptoms are similar and nonspecific to the others cause of heart failure.<sup>[9]</sup> On the other hand, simple ECG examinations and ischemic change or infarction pattern in the anterolateral segments of the heart play an important role in the diagnosis of this anomaly. Chest X-ray is normal in many cases of the adult type of this anomaly however in fulminant congestive heart failure reveals enlargement of cardiac silhouette and signs of lung congestion.<sup>[10]</sup> An interesting finding that may be detected in TEE is the absence of LMCA. Another differential diagnosis is the similarity of the clinical

finding observed in the case with the abnormal source of the LMCA of the main pulmonary trunk. Hypothetically, in spite of the variability in the patient anatomy or presence of collateral, the diagnosis of this anomaly may be made by the ECG.<sup>[11]</sup> Angiography is the main and gold standard for definitive diagnosis of atresia of the LMCA. In our case, congenital atresia of the LMCA was detected first by ECG followed by other diagnostic methods.<sup>[12,13]</sup>

## Conclusion

Congenital abnormalities of the coronary artery orifice such as single coronary artery (SCA) atresia and acute angle takeoff with tangential course are rare congenital heart diseases; we report a rare case of symptomatic SCA anomaly with combination of SVAS that undergone bypass and repair of SVAS.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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## Conflict of interest

There are no conflicts of interest

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