

Presentation of Anomalous Origin of the Left Coronary Artery From the Pulmonary Artery with Left Sided Coronary Ostium in an Adult Patient

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An anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital coronary anomaly. It usually presents in infancy with intractable left sided heart failure. Most patients die in infancy, but survival into adulthood is possible. Patients may complain of dyspnea, syncope or effort angina. They may remain asymptomatic; or experience sudden death after exercise. A 56-year-old woman presented with a two-month history of exertional chest discomfort. Echocardiography showed a coronary anomaly with preserved systolic function and no resting regional wall motion abnormality. The coronary and CT (computed tomography) angiography studies revealed the anomalous origin of the left coronary artery. A review of ALCAPA studies is presented along with images from the echocardiogram, coronary angiogram and CT scan performed for this case.

Introduction

Coronary anomalies are observed in 1-5% of patients undergoing coronary angiography¹ and in 1% of the general population.² Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), also called Bland-White-Garland syndrome,³ is a rare congenital coronary entity that occurs in 1 of 300,000 births.²

The first report of the correlation between clinical and autopsy findings in ALCAPA was made by Bland et al. In the presence of extensive collaterals, patients may survive into adulthood. However, importantly, even asymptomatic patients may experience sudden death. Once detected, surgery is recommend-

ed because ALCAPA is associated with high mortality, cardiomyopathy and arrhythmia.⁴

Herein, we present a rare case of ALCAPA in a 56-year-old woman with a new onset of effort chest pain and no history of previous cardiac symptoms or acute events.

Case report

A 56-year-old woman was referred to the Echocardiography Department of Shaheed Rajaei Cardiovascular Research Center for evaluation of mitral insufficiency. She reported exertional chest discomfort induced by ordinary activities and was associated with diaphoresis that began 2 months earlier. She had no established coronary risk factors, no history of medication use, and had six previous uneventful pregnancies. After initial hospitalization at another center for evaluation of the new onset of symptoms and recurrent and

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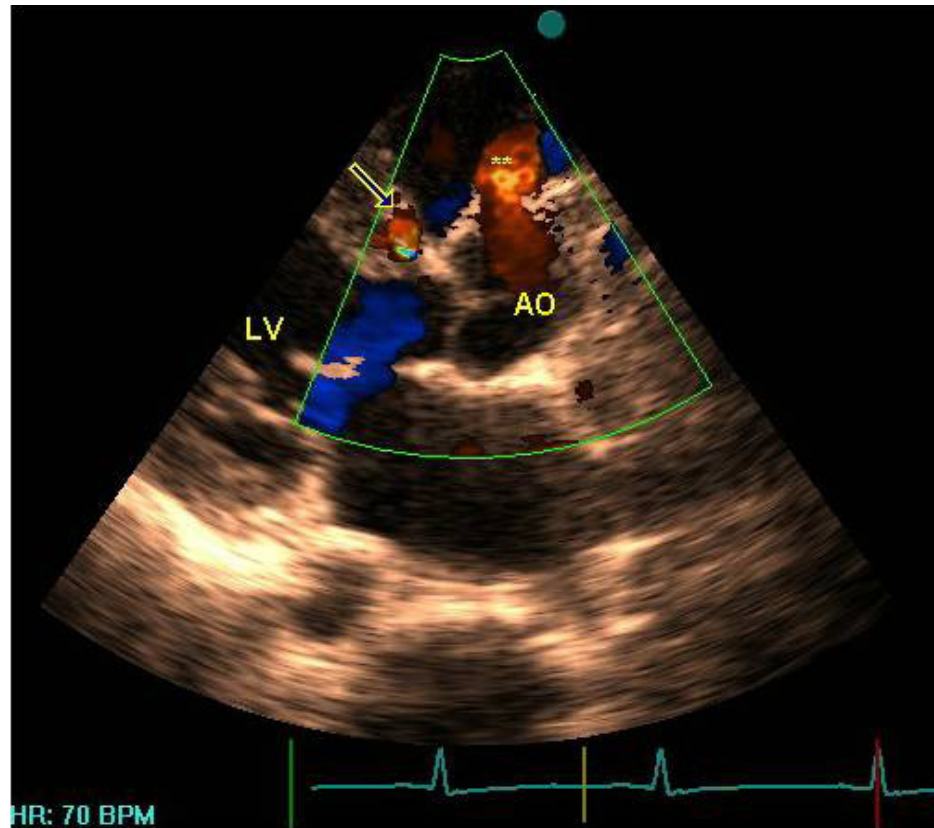


Figure 1. Parasternal long axis view in transthoracic echocardiography. The RCA is dilated at its ostium and at proximal part (asterisk) There is an abnormal color flow signal in the ventricular septum (arrow) due to the prominent development of inter-coronary collaterals.

intolerable chest pain, she was admitted to our center for additional work-up.

The results of the physical examination were unremarkable. The heart sounds were normal. A holosystolic murmur with a 2/6 intensity was heard in the left lower sternal border. The electrocardiogram showed secondary ST-T changes in the inferolateral leads and voltage criteria for possible left ventricular hypertrophy.

Both transthoracic and transesophageal echocardiograms showed the left ventricle to be moderately enlarged, but its systolic function was preserved. The ejection fraction was found to be 50%, as determined by visual estimation and according to Simpson's method.

No abnormality in the regional wall motion was detected. Mitral regurgitation of mild to moderate severity was present. The estimated pulmonary artery pressure was 32 mmHg based on the tricuspid regurgitation.

The ostium of the RCA (diameter=6.5 mm) and its proximal portion (diameter=1.4 cm) were dilated (Fig. 1). Furthermore, the RCA appeared to be tortuous in its course. A 4 mm-diameter vessel was found to originate from a coronary ostium that was detected in the left sinus of the valsalva. Bifurcation of the vessel was not found in the echocardiographic study.

Multi-colored flow signals that looked like scattered candle flames appeared all over the

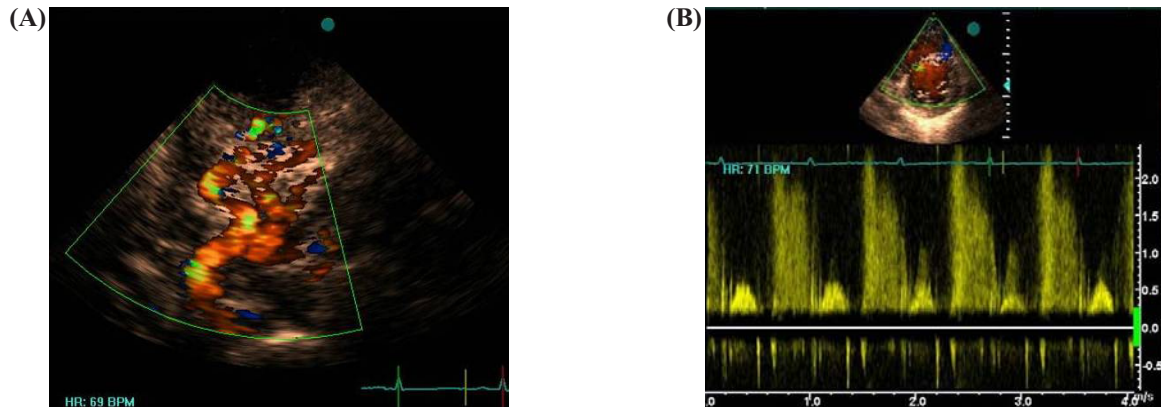


Figure 2. Extensive formation of intercoronary steal collaterals: A. An off-axis view of the apical four chambers in TTE showed prominent coronary plexuses in the interventricular septum, which were dilated septal branches. B. Pulsed Doppler study of the septal coronary collateral with a prominent Doppler signal in diastole.

free wall of the RV. The signals were found anterior to the RA, at the apex and in the interventricular septum. Their flow was prominent in diastole, which is suggestive of an extensive network of coronary vessels that interconnect the LCA and RCA. Of note, enlarged septal branches and their tortuous courses were clearly visible (Fig. 2). A Doppler interrogation of the septal collateral showed a systolodiastolic signal that was prominent in diastole. This signal indicated that the diastolic velocity increased by more than 2 m/sec; the increased velocity resulted from markedly increased collateral flow.

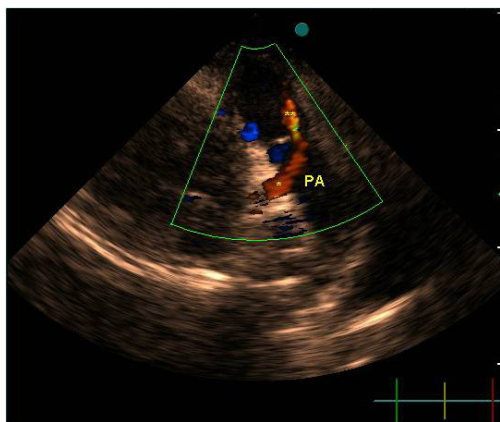


Figure 3. Abnormal large vessel (LCA) entering the PA trunk at its posterior aspect (*). ** denotes a PI jet.

A dilated vessel was found to drain into the main PA from its posterior region, just above the pulmonic valve. In a color Doppler study, a continuous flow was observed to enter the PA trunk, which was prominent in the diastolic phase (Fig. 3).

Coronary angiography was performed for delicate evaluation of coronary anatomy. During selective left coronary angiography, a coronary ostium was found in the left sinus of the valsalva and following dye injection, an accessory vessel with a tortuous course was observed that eventually drained into the left main vessel and subsequently, pulmonary artery (PA) showed retrograde opacification (Fig. 4). These observations were suggestive of a fistula that originated from the left sinus of the valsalva to the left main coronary vessel, which had retrograde flow toward the PA.

A large RCA was apparent in the right coronary angiogram (Fig. 5). The angiogram showed opacified patulous LAD and LCX branches being filled by means of an extensive and remarkably well-developed collateral circulation from the RCA branches. Later in

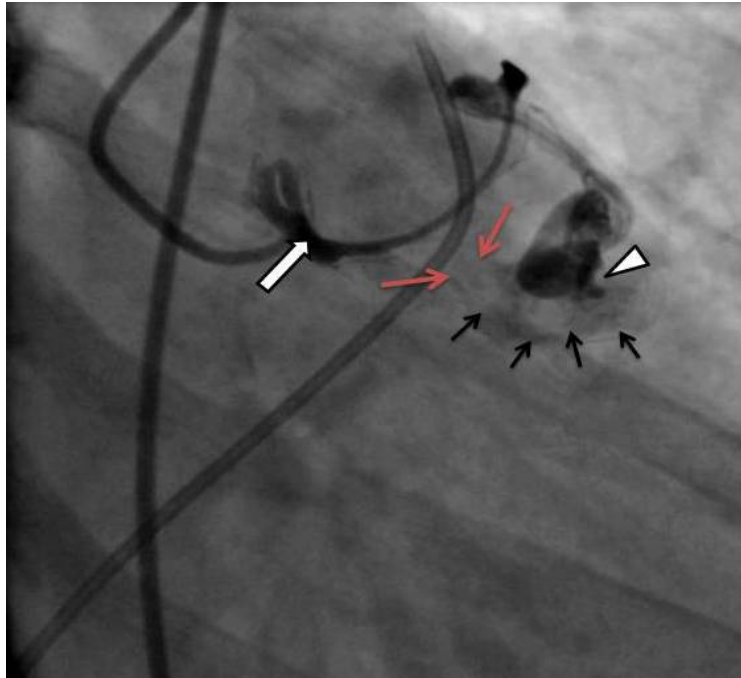


Figure 4. Catheter engagement in the ostium (white arrow) located in the left sinus of Valsalva (RAO caudal). This image shows an accessory coronary artery with an elongated and tortuous course terminating in the left main coronary artery (arrow head). Subsequently, the body of the Left main coronary artery (LMCA) appeared (black arrows). The red arrows denote the insertion site of the LMCA into the pulmonary artery.

the filming sequence, retrograde flow from the LAD and LCX arteries opacified the LMCA and its origin from the main PA.

The images obtained from a 10-slice MDCT

coronary angiography showed a left main coronary artery with an anomalous origin from the left posterior side of the main pulmonary artery.

The left main vessel bifurcated to the large and

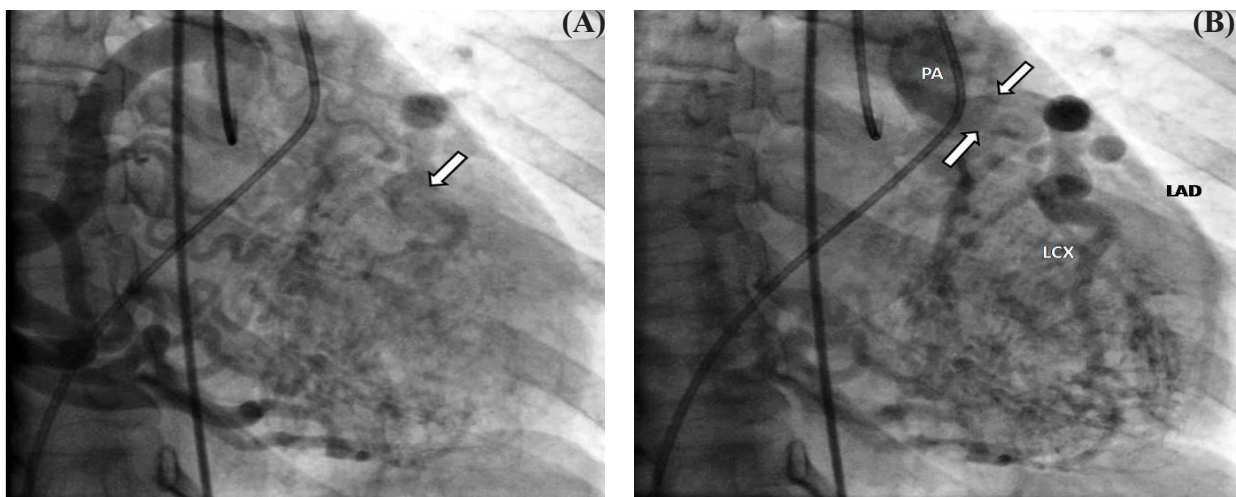


Figure 5. Right coronary angiogram (RAO caudal) showed a greatly dilated RCA with development of extensive intercoronary collaterals. The left coronary system became opacified retrogradely via collaterals (arrow in A). In the late phase, the left main coronary artery appeared and eventually, further along the course of the LMCA, the pulmonary artery appeared as well. In B, white arrows denote the anomalous site of origin of the LMCA from the PA.

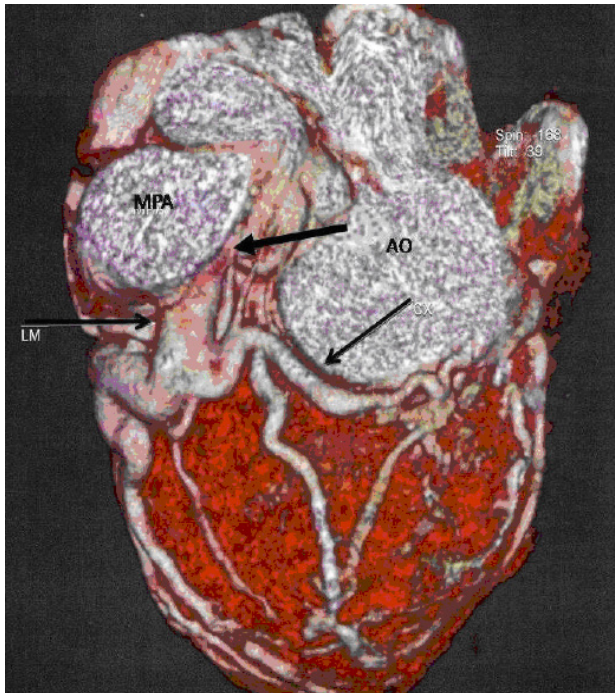


Figure 6. The 10-slice MDCT coronary angiography confirmed the presence of ALCAPA. The thicker black arrow denotes the insertion site of the LMCA in the PA.

tortuous LAD and LCX branches (Fig. 6). The RCA originated from the right coronary sinus and had a normal course; however, the course was tortuous and the vessel was obviously dilated. There were multiple branches from the right and left coronary arteries that connected to the anterior aspect of the RA wall, the septum and the anterior part of the RV chamber.

A high resolution, ECG-gated computed tomography of the heart, with special attention focused on the coronary arteries, was performed using Siemens calcium scoring software. According to the Agatston's scoring system, the calcium score was zero, indicating that the patient was at low risk of developing coronary artery disease.

The patient was referred to cardiac operation but refused to undergo the surgery.

Discussion

An anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital coronary anomaly that accounts for 0.2-0.5% of all congenital heart diseases.⁵ Only a small minority of undiagnosed and untreated patients reach adulthood. The prevalence of this disease in adults, as determined by coronary angiography studies, is estimated to be 1–5 in 10,000.⁶ More than 90% of untreated adult patients will die at a mean age of 35 years.⁷

At first, the ischemia is transient and occurs only with exertion; however, when the myocardial demand increases, an infarction of the anterolateral wall occurs. Heart failure is a prominent clinical sequel in ALCAPA. Another outcome is mitral regurgitation, which may occur through one of several mechanisms including dysfunction of an ischemic anterolateral papillary muscle or dilation of the ring caused by an enlarged dysfunctional left ventricle.

The most common cause of death is intractable heart failure. In the first year of life, usually after 2 months of age, 65% to 85% of infants die.⁶ About 15% of patients survive into adulthood. These are the patients who have sustained myocardial blood flow at rest or, in some cases, during exercise.

The presence of extensive intercoronary collaterals, as found in our case, or restrictive opening between the origin of the LCA and the pulmonary trunk is believed to permit some ALCAPA-affected persons to remain asymptomatic.³ Another reason for an asymptomatic course could be the existing systemic collateral supply. Rare cases have been reported in which collaterals join the bronchial arteries

or RITA to the LCA.⁸

Importantly, adult patients, even those who are asymptomatic, are still at an increasing risk of sudden death.⁶

An echocardiographic diagnosis of the anomaly is difficult. In adult patients, direct visualization of the anomalous connection is often a challenge. More specifically, a deficiency in the lateral resolution in the left aortic sinus wall in 2D studies is found which mimics LCA arising from left sinus of the valsalva. The identification of intercoronary collaterals in the ventricular septum may be the only echocardiographic finding that can alert the examiner to a case of ALCAPA.⁹

In our case, we suspected a coronary anomaly from the presence of the dilated right coronary and its tortuous course. In addition, in a 2D and color Doppler study, there were large and abundant lacunae present in the RV free wall, in the apex and in the anterior interventricular septum with turbulent systodiastolic flow. This was more prominent in the diastolic phase and a dilated vessel, with a prominent diastolic flow, that drained into the main pulmonary artery. A 4mm-diameter coronary artery originating from a coronary ostium was detected in the aortic root (the left sinus of valsalva). An angiographic study revealed that the artery was an accessory vessel that drained to the left main artery. Based upon the echocardiographic findings, a right coronary artery fistula to the main PA was considered. The pitfall in the echocardiographically directed diagnosis was due to finding this accessory coronary artery originating from the coronary ostium at the left sinus of valsalva. Notably, we could not find a bifurcation of that vessel in the echocardiog-

raphy study, which is contrary to its nature of being the left coronary artery. We initially attributed this observation to the technical difficulty of having a limited echocardiography window, which is known to occur in some adult echocardiographic studies. Therefore, observation of an ostium in the left coronary sinus does not rule out ALCAPA.

In our patient, the Doppler signals in the dilated RCA, the anomalous LCA and the intercoronary vessels in the septum were more prominent in the diastolic phase. In contrast, a previous report of ALCAPA indicated that the coronary flow intensified in the systolic phase; the presence of such an increase in systolic coronary flow had been offered as a diagnostic clue for differentiating ALCAPA from other coronary anomalies.¹⁰ As a practical point, in the presence of a dilated RCA or diastolic flow in the main PA, ALCAPA should be strongly considered as a possibility, regardless of the patient's age or severity of symptoms. In connection with a simple dilation of coronary arteries, other diseases to be considered in the differential diagnoses include atherosclerosis, vasculitis (Takayasu's arteritis), Scleroderma, Ehlers-Danlos syndrome, Kawasaki's disease and hemorrhagic telangiectasia.¹¹ In this patient, the left to right shunt was significant, based on the echocardiographic data, but the pulmonary artery pressure was within the normal range.

In ALCAPA, other signs may be present, such as LV enlargement, a regional wall motion abnormality in the anterolateral region, increased echogenicity of the papillary muscle and an adjacent endocardium due to fibrosis or mitral regurgitation. In our case, no regional

wall motion abnormalities was found in the resting echocardiographic study and myocardial function was preserved. Once an ALCAPA diagnosis is confirmed, surgical repair is recommended.

Currently, the preferred surgical method is to establish a dual coronary artery system. Other methods include ligation of LCA at its origin and reconstitution of the flow with either a subclavian arterial or saphenous venous graft; direct reimplantation of the coronary artery into the aorta; or the Takeuchi technique (creation of an AP window and a tunnel with a direction of flow from the aorta to the left coronary ostium). The best result is achieved with direct implantation of the LCA into the ascending

aorta and simultaneous mitral valve repair, if needed.¹²

The present observation is unique in that it describes a case of ALCAPA in an adult patient who had a long asymptomatic course and presented with preservation of the myocardium with no evidence of infarction or regional wall motion abnormalities in a resting echocardiographic study.

Acknowledgements

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