

# Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery in Adulthood: Challenges and Outcomes

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Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is an extremely rare, potentially fatal, congenital anomaly with a high mortality rate in the first year of life. It occurs rarely in adulthood and may appear with malignant ventricular arrhythmia or sudden death. We report a case of a 49-year-old woman with ALCAPA who presented with dyspnea on exertion. Management was coronary artery bypass grafting to the left anterior descending artery and obtuse marginal arteries, closure of the left main coronary artery ostium, and reestablishment of the dual coronary artery system.

*Key words:* 1. Coronary artery bypass  
2. Cardiopulmonary bypass

## Case report

A 49-year-old woman presented with breathlessness on exertion with gradual onset over the previous one month; there was no other significant past medical history. Cardiac and respiratory examinations were normal, and the 12-lead electrocardiogram (ECG) was inconspicuous. Two-dimensional echocardiography showed an anomalous left main coronary artery (LMCA) arising from the main pulmonary artery (MPA), moderate left ventricular dysfunction, an ejection fraction of 40%, and mild mitral regurgitation. The coronary angiogram revealed anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). Multislice computed tomography (CT) coronary angiography revealed an ALCAPA from the posterior wall of the MPA, which bifurcated into the left anterior descending (LAD) artery and the left circumflex artery. The right coronary artery (RCA) was tortuous,

dilated, and arising from the aorta (Fig. 1). The patient was otherwise healthy and had an uneventful life history (from childhood to adulthood), without any evidence of arrhythmic or syncopal attacks.

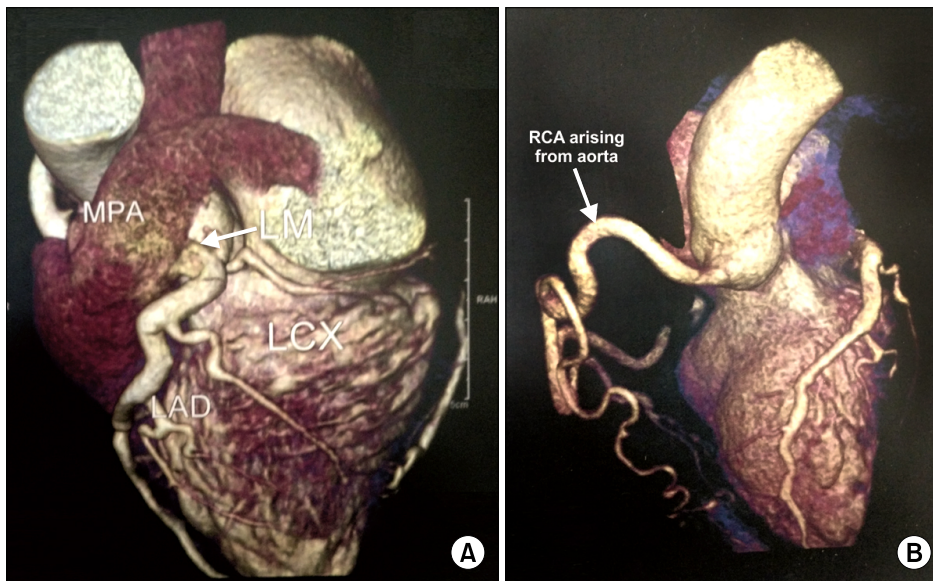
The patient underwent coronary artery bypass grafting (CABG) with closure of the LMCA opening from inside the MPA. Surgery was performed with both invasive arterial and pulmonary artery pressure monitoring. After sternotomy and opening of the pericardium, we observed the RCA arising from the aorta, which was hugely dilated, tortuous, and the LMCA was arising from the lateral aspect of the MPA (Fig. 2). It was not possible to mobilize the LMCA for reimplantation to the aorta, therefore we planned a CABG with closure of the left main orifice of the LMCA. The left internal mammary artery (LIMA) was harvested and cardiopulmonary bypass (CPB) was established with right atrial and aortic cannulation. After aortic cross clamping, cardioplegia was given

Received: March 3, 2016, Revised: July 27, 2016, Accepted: July 29, 2016, Published online: October 5, 2016

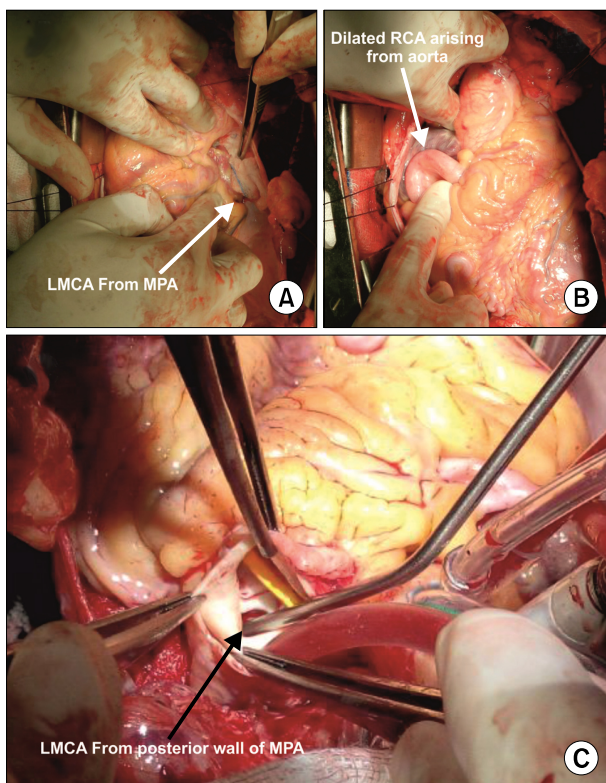
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**Fig. 1.** Preoperative computed tomography coronary angiogram. (A) Left main coronary artery arising from the MPA, bifurcating into the LAD artery and the LCx artery. (B) Dilated and tortuous RCA arising from the aorta. MPA, main pulmonary artery; LAD, left anterior descending; LCx, left circumflex; RCA, right coronary artery.

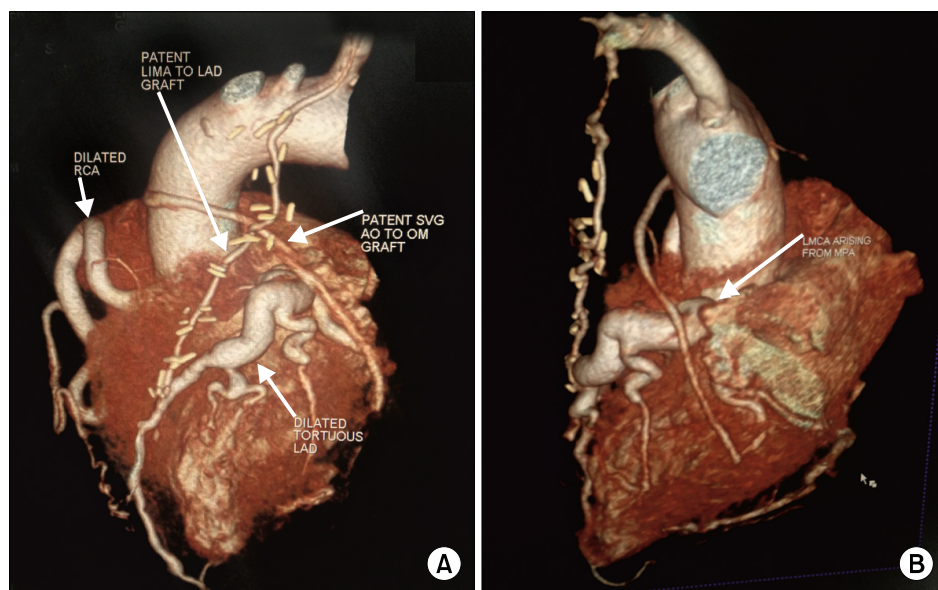


**Fig. 2.** Intraoperative view. (A) Origin of the LMCA from the posterior aspect of the MPA. (B) Hugely dilated and tortuous RCA arising from the aorta. (C) Slit-like opening of the LMCA from the posterolateral wall of the MPA. LMCA, left main coronary artery; MPA, main pulmonary artery; RCA, right coronary artery.

by two methods: root cardioplegia was given in the aorta for the RCA and followed by left main ostial cardioplegia after opening the MPA (Fig. 2). The LIMA was grafted to the LAD and the reversed saphenous vein was grafted to the second obtuse marginal artery. The left main ostium was closed with 6-0 Prolene sutures from inside the MPA. Aortic cross clamp was released, CABG completed, the patient was weaned from CPB, and the chest was closed. The patient was transferred to the intensive care unit with minimal inotropes, stable hemodynamics, and an isoelectric ECG. Extubation occurred on the morning of the next day (postoperative day 1). On postoperative day 2, the patient had atrial fibrillation with a fast ventricular rate, which required amiodarone infusion, oral beta-blockers, and oral digoxin for rate control. After 72 hours (i.e., postoperative day 5), the patient converted to normal sinus rhythm and was maintained with oral amiodarone and beta-blockers. On postoperative day 8, a CT coronary angiogram was performed, which showed blood flowing through the bypass grafts and no communication between the MPA and LMCA (Fig. 3).

## Discussion

ALCAPA is a rare and potentially fatal congenital anomaly. First described by Brooks [1] in 1886, ALCAPA has an incidence of 1 in 300,000 live births (0.25% to 0.5%) [2]. The anomaly, if left untreated,



**Fig. 3.** Postoperative computed tomography coronary angiogram. (A) LIMA with good blood flow to LAD and RSVG → OM2 artery grafts (arrows). (B) Closed opening of the LMCA from the MPA without any communication between the two vessels (arrow). LIMA, left internal mammary artery; LAD, left anterior descending; RSVG, reversed saphenous vein; OM2, second obtuse marginal; LMCA, left main coronary artery; MPA, main pulmonary artery; RCA, right coronary artery.

has a mortality rate approaching 90% in the first year of life [3]. Due to extensive intercoronary collaterals with the large dominant RCA, only 10% to 15% of patients with this ALCAPA survive into adulthood [4].

ALCAPA occurs from either an abnormal septation of the conotruncus or from the persistence of the pulmonary buds combined with involution of the aortic buds that form the coronary arteries. It usually occurs as an isolated entity, but may be associated with atrial septal defect, ventricular septal defect, and coarctation of the aorta. ALCAPA is well tolerated in utero and in infancy because the pulmonary arterial pressure equals the systemic pressure, which leads to antegrade flow in the anomalous left coronary artery (LCA) and the normal RCA. Both circulations have negligible saturation differences.

Development of the extensive collateral circulation between the RCA and LCA during this time period determines the extent of myocardial injury and the outcome. Patients with ALCAPA who survive into adulthood have an enormous enlargement of the RCA and extensive intercoronary collaterals [2].

Variable clinical presentation of ALCAPA is seen in this adult cohort, as patients may be asymptomatic or present with mitral insufficiency, ischemic cardiomyopathy, and cardiac failure or with malignant ventricular dysrhythmias. Sudden cardiac death occurs in 80% to 90% of patients with malignant ventricular dysrhythmias. Our patient presented with dyspnea on exertion, but was otherwise asymptomatic.

Coronary angiography is the traditional standard method used for diagnosis; however, cardiac CT and cardiac magnetic resonance imaging have been increasingly employed for the diagnosis of ALCAPA. Echocardiography serves as an important tool to aid in diagnosis, because it visualizes the abnormal origin of the LCA, dilated RCA, retrograde filling of the anomalous coronary, abnormal diastolic flow in the MPA, and abnormal septal or epicardial color flow signals from the collateral vessels.

Surgical correction is standard in the therapeutic treatment of ALCAPA. Examples of surgical correction methods include direct reimplantation of the left coronary into the aorta [5], baffle repair [6], interposition of the free subclavian artery [7], and use of the internal thoracic artery as a bypass graft [8]. Establishment of a dual coronary system is a widely used corrective surgery to repair ALCAPA in adult patients. All variants of CABG incorporate ligation of the ALCAPA at the pulmonary origin in order to avoid competitive flow.

Administration of cardioplegia for myocardial protection is a crucial element common to all surgical techniques. In this case, we gave cardioplegia in the aortic root and direct ostial cardioplegia for the left coronary system.

Management of ALCAPA in adults is controversial and may consist of ligation of the anomalous left coronary artery, CABG, or direct reimplantation.

A high index of suspicion and appropriate diagnostic

modalities should allow rapid diagnosis of ALCAPA in any patient seen with dilated congestive cardiac failure. Recently, operations that reestablish two coronary arterial circulations have dramatically improved surgical results for an otherwise fatal disease. Close and long-term follow-up of this patient is necessary to better understand the corrected natural history after surgical treatment of ALCAPA.

We report a case of a 49-year-old woman with ALCAPA who presented with dyspnea on exertion. Management involved CABG to the LAD and obtuse marginal arteries, closure of the LMCA ostium, and reestablishment of the dual coronary artery system.

### Conflict of interest

No potential conflict of interest relevant to this article was reported.

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