

# Uncommon Anomaly and Uncommon Errors: Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA) in 60 year Old Women

Inga Botchorishvili. MD.PHD<sup>1\*</sup>, Rusudan Agladze. Md. PHD<sup>2</sup>, Konstantine Kipiani.Md.PHD<sup>3</sup>, Zviad Kereselidze. MD<sup>4</sup>, Archil Chukhrukidze. MD.PHD<sup>5</sup>, Tamar Gaprindashvili. MD<sup>6</sup>, Lika Injia. MD<sup>7</sup>, Sergo Khajalia. MD<sup>8</sup>, Nikoloz Lekiasvili. MD<sup>9</sup>, Tornike Sologashvili. MD.PHD<sup>10</sup>, Zurab Pagava. MD.PHD<sup>11</sup>

<sup>1</sup>Interventional cardiologist, Bokhua Memorial Cardiovascular Center. Endovascular surgery department. Georgia. Tbilisi.

<sup>2</sup>Cardiologist., Bokhua Memorial Cardiovascular Center. Cardiology department. Georgia. Tbilisi.

<sup>3</sup>Vascular Surgeon, Bokhua Memorial Cardiovascular Center. Cardiology department. Georgia. Tbilisi

<sup>4</sup>Interventional Cardiologist, Tbilisi Heart Center. Cardiology department. Georgia. Tbilisi.

<sup>5</sup>Cardiologist, Tbilisi Heart Center. Cardiology department. Georgia. Tbilisi.

<sup>6,7,8</sup>Cardiologist, Bokhua Memorial Cardiovascular Center. Cardiology department. Georgia. Tbilisi.

<sup>9</sup>Radiologist, Bokhua Memorial Cardiovascular Center. Radiology department. Georgia. Tbilisi.

<sup>10</sup>Cardiac Surgeon, Geneva University Hospital. Pediatric cardiac surgery unit. Geneva. Switzerland.

<sup>11</sup>Cardiologist, Bokhua Memorial Cardiovascular Center. Cardiology department. Georgia. Tbilisi

\*Corresponding Author: Inga Botchorishvili. MD.PHD, Interventional cardiologist, Bokhua Memorial Cardiovascular Center. Endovascular surgery department. Georgia. Tbilisi.

## ABSTRACT

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is rare congenital cardiac anomaly which results high mortality during infantile period and only 10% to 15% of the individuals with this defect reach adulthood. We describe a 60 year-old women case with ALCAPA who was misdiagnosed with LM occlusion that lead to unnecessary coronary artery bypass grafting (CABG). After surgery a patient's symptoms deteriorated until visit at our clinic definitive diagnosis was not established.

This clinical case highlights the importance of recognizing the risks of misdiagnosis, which can lead to life-threatening conditions. Invasive coronary angiography (ICA) was the standard for ALCAPA diagnosis as it depicted the course of the anomalous coronary artery; however, it has been largely replaced by noninvasive diagnostic testing. Thorough examination is essential, particularly when atypical alterations in the vascular bed are identified.

**Keywords:** Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA); Invasive Coronary Angiography (ICA). Computed tomography coronary angiography (CTCA). Coronary artery bypass grafting (CABG). Takeuchi technique.

## ARTICLE INFORMATION

Received: 04 May 2025

Accepted: 14 May 2025

Published: 21 May 2025

**Cite this article as:**

**Inga Botchorishvili, Agladze, Konstantine Kipiani, et al.** Uncommon Anomaly and Uncommon Errors: Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA) in 60 year Old Women. Open Journal of Medical Images and Case Reports. 2025;2(1); 14-18.

<https://doi.org/10.71123/3067-1078.020104>

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## INTRODUCTION

Coronary artery anomalies are rare condition but their impact on cardiac morbidity and mortality among newborns and young adults is not trivial. Impaired myocardial perfusion

is affected by certain types of coronary anomalies, which can lead myocardial ischemia and death [1].

Anomalous origin of the left coronary artery from the pulmonary artery – ALCAPA is very rare anomaly. The

prevalence of ALCAPA is 1/300,000 live births (between 0.24% and 0.46% of all congenital cardiac anomalies) [2]. During embryogenesis anomalous septation of the truncus arteriosus into aorta and pulmonary artery, followed by anomalous development of coronary ostia is believed to be the underlying cause of this anomaly [3].

In newborns at birth symptoms are rarely presented due to relatively higher pulmonary artery resistance [4], which allows for maintaining an antegrade flow into the anomalous left coronary artery. Symptoms mostly develop at 2-3 months of age, when resistance in the pulmonary artery drops, causing an increase retrograde flow through the anomalous connection of left coronary artery, stealing blood flow into pulmonary artery and which reduces myocardial perfusion, especially if the intracollateral blood supply is not sufficiently developed [5]. In most cases survival is low without surgical treatment, but in recent decades several cases of ALCAPA in adults have been reported.

We present a case of an elder women in whom ALCAPA was misdiagnosed.

## CASE PRESENTATION

In December 2023 a 60 year-old women patient was referred to our clinic. She had been experiencing intense, sharp, precordial pain during physical exertion accompanied by fatigue. Prior to visiting our clinic she reported several hospitalizations at another clinics due to ischemic heart disease from February 2023.

We present a timeline of her medical history and procedures according her medical reports.

### First Hospitalization - Feb.2025

In February 2023 a patient was hospitalized in one of

the hospitals of Tbilisi in cardiology department with diagnosis :

Acute coronary syndrome (ACS) and Dyslipidemia. By echocardiography wall motion abnormalities were not found; There were no changes on ECG and no elevation of cardiac biomarkers. According her past history she didn't mentioned any symptoms of angina pectoris until last five months. No family history of ischemic heart disease. She was a current smoker. Last period before hospitalization she experienced exercise intolerance and palpitations during physical activity. She had two pregnancies without complications.

The patient presented with recurrent chest pain despite appropriate medical therapy which prompted further evaluation with invasive coronary angiography (ICA) at the same time on first hospitalization. Selective cannulation of left coronary artery (LM) was unsuccessful with standard technique. ICA revealed dilated and tortuous right coronary artery (RCA) with multiple collaterals to the left coronary system. (Fig 1). There were atherosclerotic changes in the left circumflex artery (CX) distal segment was occluded. According to the specialist's opinion LM was considered to be occluded as well. According these findings coronary artery bypass grafting (CABG) was suggested to the patient.

Upon reviewing initial coronary angiogram examination showed absence of left coronary artery originating from aorta, dilated and tortuous right coronary artery (RCA) and abundant Rentrop grade 3 coronary collaterals communicating with left coronary artery originating from pulmonary trunk. But interestingly, very minor communication between LM and PA (Fig.1 B), which caused the main error and subsequently triggered other issues.



Figure 1(A). ACX distal segment is occluded



**Figure 1(B).** Coronarography shows there is minor communication between LM and PA.

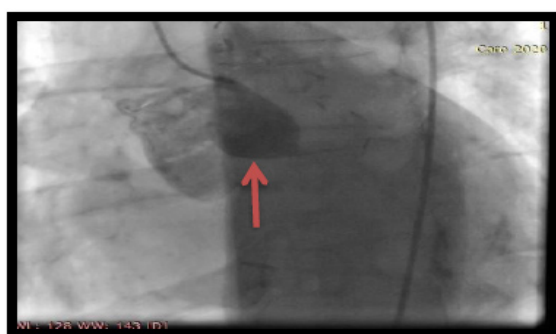
After several days CABG was performed with mini thoracotomy. Left Internal mammary artery (IMA) graft had been applied to LAD. A patient was discharged with stable condition and appropriate prescription.

### Outpatient Visit - Oct.2023

Until October 2023 a patient remained symptom free. In November she was admitted at the ambulatory unit of the same hospital with the same symptoms she experienced before CABG. Patient underwent echocardiography and laboratory tests were performed. Examinations didn't revealed any sign of cardiac damage. Presented symptoms were considered to be an extra cardiac. Conservative treatment was continued and additional examinations of gastrointestinal system were suggested.

### Second Hospitalization - Nov.2023

One month later a patient was admitted at another hospital. Since the previous visit patient's condition has worsened. She reported reduced exercise tolerance dyspnea and fatigue during minimal physical activity. Echocardiography demonstrated apical wall motion abnormalities. Cardiac biomarkers were in normal range. ICA was performed and examination revealed IMA anastomosis dysfunction. Part of collateral flow was diminished. (Fig.2.B). Distal to the anastomosis apical segment didn't visualized.(Fig.2.C). Invasive coronary intervention was not considered at that time. Optimal medical treatment (OMT) was continued. And again uncommon coronarography findings were left out of focus.



**Figure 2. (A)** Aortic root without LM origin. **(B).** Diminished collateral flow **(C).** Occlusion distal to IMA anastomosis

It was obvious that OMT was ineffective as well as clinical symptoms were progressing.

### Outpatient Visit – 12.2023

In December 2023 a patient referred to our clinic. As

mentioned above during examinations according previously performed coronary angiographies non atherosclerotic LAD and ectasic RCA raised suspicion that a rare anomaly was present. Computed tomography coronary angiography (CTCA) confirmed diagnosis of ALCAPA. (Fig.3)

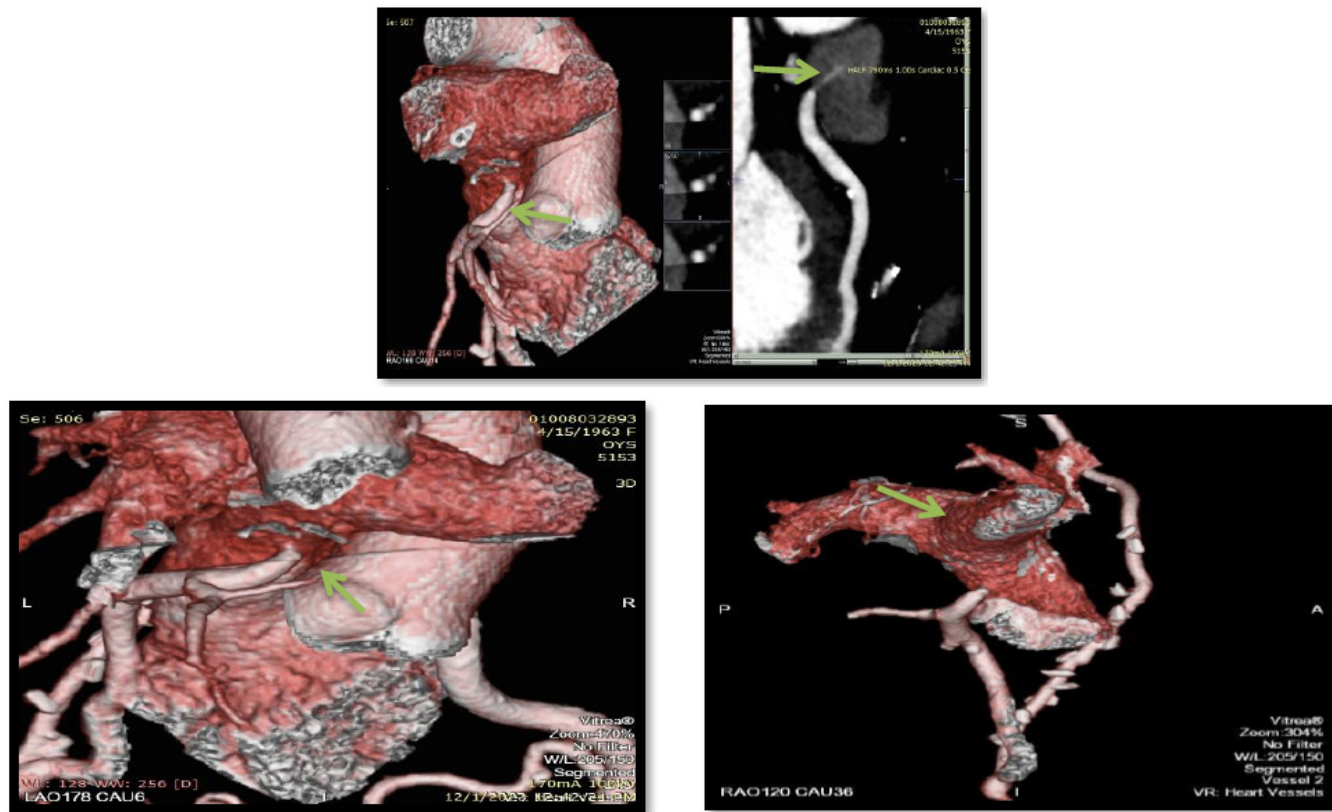


Figure 3. CTCA .Green arrows indicate the origin of LM from PA

After this finding, the pathogenesis of ischemia became clear.

### Third Hospitalization and Redo Surgery - 02.2024

In February 2024 redo surgery was performed successfully. Ligation of the left coronary artery (LCA) at its origin from the pulmonary artery was performed. Surgical angioplasty was performed at occluded distal anastomosis site. LIMA anastomosis to LAD was re-implanted.

Restoration of blood flow and perfusion pressure contributed to the normalization of oxygen supply and demand.

After several days she was discharged. The patient remained symptom-free since last surgery. During follow up period cardiac MRI was performed. MRI didn't revealed any myocardial scare or fibrosis.

### DISCUSSION

ALCAPA syndrome was described in 1866 and 1908 [6,7]. However, the first clinical description was published in 1933 by Bland, White and Garland [8], and the condition is

also known as Bland-White-Garland syndrome. ALCAPA is isolated anomaly but in 5% cases coarctation of aorta and atrial or ventricular septal defects were described [9].

As mentioned above due to hemodynamic changes – particularly the drop in pulmonary artery resistance at 3-4 months of age - myocardial ischemia, angina, mitral insufficiency, life threatening arrhythmias and sudden cardiac death may develop. Symptoms can be manifested at any decade of life and are driven by hemodynamic alterations in apical/subendocardial vessels as well as collateral circulation. ALCAPA is associated with high infant mortality and 80%-90% of individuals with this anomaly die suddenly with a mean age around 35 y [10,11,12].

Previous publications described 'infantile' and 'adult' forms of ALCAPA classified according the age at clinical presentation [2].

In our case the most intriguing aspect is the patient's age - 60 year. ALCAPA is exceptionally rare diagnosis especially in elderly individuals. Remarkably, this patient didn't develop any clinical signs of ischemia until the age of 60.

Survival at any age depends on several factors including extent of collateral flow from RCA, diameter of LM/PA shunt and although myocardial area supplied by anomalous left coronary artery (ALCA). In current case large portion of anterior wall was supplied by LAD, however significant stenosis was observed at the site of connection of LM/PA. LAD was filled retrogradely from RCA and drained into pulmonary artery (PA), but diameter of this connection was insufficient to cause coronary steal phenomenon. It likely helped maintain perfusion pressure and preserved myocardial viability, until the development of atherosclerosis in CX which compromised myocardial blood supply and we believe that this atherosclerotic changes on CX was a reason to suspect that atherosclerotic changes were reasonable for LM occlusion. As well the presence of abundant Rentrop grade 3 intercoronary collaterals between RCA and LAD was likely the main factor that supported the patient's survival over six decades.

After CABG competitive flow caused diminishing collateral flow. It is our opinion that flow distal to anastomoses was not sufficient because of stealing minor part of blood from LM into PA. Despite LM ostial stenosis probably this anomaly connection impaired perfusion pressure maintenance which caused apical segment thrombosis - distal to anastomoses.

There are several types of surgical techniques for correction this anomaly. The surgical technique is reimplantation with translocation of the left coronary artery from the pulmonary trunk to the aorta, which is possible most of the time. If this technique cannot be applied, "tunneling" or the Takeuchi technique is recommended; Other surgical options are the simple ligation of the left coronary artery and the performance of venous or arterial grafts [9].

## CONCLUSION

This case is highly unusual due to factors which preserved myocardium perfusion during 60 years : well established collateral net from RCA and LAD ostial stenosis at the site of communication with PA, that more or less maintained perfusion pressure in LAD. Moreover this case is unusual due to errors during all 'treatment' period. Performing CABG without ligating LAD lead to perfusion imbalance and myocardial ischemia.

Despite all factors discussed above, the mechanism that developed rapidly in infancy and protected the myocardium from life-threatening complications for six decades remains unclear which keeps this case unique.

## Conflict of Interest

We do not declare any conflict of interests.

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