Late presentation of an anomalous left coronary artery from the pulmonary artery treated with conservative surgical management with long-term cardiac magnetic resonance imaging follow-up

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Abstract
Anomalous origin of the left coronary artery from the pulmonary artery is rare congenital abnormality that most commonly presents in childhood and is associated with a high mortality. In the elderly, patients may present acutely with arrhythmias or signs of ischemia or with vague chronic presentations of shortness of breath and fatigue. In the high-risk elderly population, it is unclear as to whether conservative surgical management by means of suture ligation of the left coronary artery is associated with positive long-term outcomes. We present a case of a 69-year-old patient diagnosed with anomalous origin of the left coronary artery from the pulmonary artery, which was treated with conservative surgical management and followed up for 15 years with cardiovascular magnetic resonance imaging, with positive outcomes.

Keywords
Anomalous origin of the left coronary artery from the pulmonary artery, coronary vessel anomalies, coronary angiography

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Introduction
Congenital abnormal origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare (1/300,000) congenital defect with a high infancy mortality rate (85%).1 The pathophysiology of ALCAPA can be simplified into four phases: a neonatal phase associated with elevated pulmonary pressures and oxygenation providing anterograde perfusion from the pulmonary artery (PA), an early infancy phase with reduced pulmonary pressures and oxygenation leading to ischemia, an adult phase with the formation of collateral circulation from the right coronary artery (RCA), and a shunting phase where excessive left coronary artery (LCA) shunting to the RCA results in a coronary steal syndrome.2 It is uncommon for ALCAPA to be diagnosed later in life with late presentation hinging on how the ischemic infancy stage is tolerated and the extent of collateralization.3 In the elderly population, common presentations include chest pain, shortness of breath, palpitations, syncope or weakness.4 In the elderly population, it is unclear whether transplantation of the LCA to the aorta provides additional benefit to conservative management, ligation of the LCA. Developments in cardiovascular magnetic resonance imaging (CMRI) have allowed for patients with ALCAPA to be investigated and followed up non-invasively, identifying areas of fibrosis and reversible ischemia.5 The use of this imaging technology will assist clinicians to non-invasively follow-up patients with ALCAPA and document effects of aggressive versus conservative management options.

Case report
We present the case of a 69-year-old woman who presented with progressive chest discomfort and dyspnea. Her past medical history was not contributory and she had no cardiac risk

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factors. On physical examination, her jugular venous pressure was 10 cm above the sternal angle, apical impulse was displaced laterally, and a grade 2/6 holosystolic murmur was auscultated at the apex and second left intercostal space. Electrocardiogram demonstrated a left bundle branch block with moderate elevation of cardiac enzymes. Echocardiography demonstrated a dilated left ventricle (left ventricular internal dimension in diastole (LVID): 62 mm) and severely reduced ejection fraction of 20%.

Coronary angiography (Figure 1) was undertaken with RCA ectasia and inability to identify the LCA with standard techniques. Subsequently, power injection of the right coronary ostium demonstrated a large tortuous dilated RCA and right heart catheterization may show an oxygen saturation increase from the right ventricle to the PA. CMRI and CT can be helpful to clarify exact coronary ostial locations and CMRI perfusion imaging can be used to demonstrate retrograde flow from the RCA to the left coronary system through an extensive network of collaterals.

In pediatric cases, survival hinges upon the formation of collaterals from the RCA circulation, with subsequent surgically reimplantation of the LCA to the aorta. In the adult population, a two-coronary system repair remains the gold standard with ligation of the LCA along with coronary artery bypass graft (CABG), coronary button transfer or the Takeuchi procedure. However, in the elderly (>65 years) population with excellent collaterals, it is unclear whether the benefits of a two-coronary system repair outweigh the risks. A documented complication of a single-coronary system repair is anterior papillary muscle rupture and association mitral regurgitation, which did not occur in the presented case during follow-up. In the presented case, the patients’ coronary angiography demonstrated excellent collateral circulation, so it was felt that the benefits of re-implanting the anomalous artery or CABG to slightly improve collateral circulation did not outweigh the risks of cardiopulmonary bypass. Instead, the multi-disciplinary team elected a watchful waiting approach, where evidence of continued ischemia during follow-up would have initiated a surgical consultation for a two-coronary system repair.

ALCAPA in the elderly population is a rare diagnosis that is difficult to make prior to angiography. The majority of ALCAPA cases in the adult population presents with angina, dyspnea, palpitations, or fatigue. In the presence of excellent collateral circulation, it is unclear whether there is added benefit to re-implanting the anomalous artery or CABG to slightly improve collateral circulation. While further systematic research is needed, this case would support a growing body of literature.

**Figure 1.** Coronary angiography. Images were obtained using a 7 French multi-purpose diagnostic coronary catheter with power injection. (a) Right anterior oblique image: early after injection demonstrating the RCA ectasia and collaterals to the left anterior descending (LAD) and circumflex arteries. (b) Left anterior oblique image: demonstrating the prominent septal and posterior descending artery to LAD collaterals and (c) a late image demonstrating filling of the PA via prominent left to right shunt. (d) Direct injection of the LCA via a right heart catheterization with an Amplatzer left 2 diagnostic catheter in the PA.

**Discussion**

Physical examination and echocardiography can provide clues to the diagnosis; CMRI, computed tomography (CT) coronary angiography, and cardiac angiography remain the gold standard of diagnosis. In cases of ALCAPA presenting in adulthood, angiography typically demonstrates a tortuous dilated RCA and right heart catheterization may show an oxygen saturation increase from the right ventricle to the PA. CMRI and CT can be helpful to clarify exact coronary ostial locations and CMRI perfusion imaging can be used to demonstrate retrograde flow from the RCA to the left coronary system through an extensive network of collaterals.

Our case demonstrates good long-term outcome with a conservative strategy of LCA ligation in an elderly patients with excellent collaterals. While further systematic research is needed, this case would support a growing body of literature.
supporting less invasive approaches to the treatment of ALCAPA in the adult population.\textsuperscript{11}

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