



Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) in an Old Adult

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Abstract

The anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital cardiac malformation. It presents predominantly in infancy and its main presenting feature is myocardial ischemia or heart failure. Survival to adulthood is quite uncommon. If untreated, mortality from ALCAPA approaches 90% in infancy; early recognition and surgical correction are, therefore, essential. With early surgical correction, the prognosis is good. There are two types of ALCAPA syndrome: the infant type and the adult type, each of which has different manifestations and outcomes. Infants experience myocardial infarction and congestive heart failure, and approximately 90% die within the first year of life. A literature review regarding this anomaly in teenagers and adults show that only 25 cases have been diagnosed during life and 18 additional cases of ALCAPA in these age groups have been diagnosed post mortem. We present a rare case of a 60-year-old man, who referred to our center due to dyspnea on exertion from the previous year without any history of chest pain and diagnosed as ALCAPA. Given the absence of ischemia and the patient's age, only medical therapy was recommended.

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Introduction

The anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) was first described in 1866. The first clinical description, in conjunction with autopsy findings, was described by Bland and colleagues in 1933, so the anomaly is also called the Bland-White-Garland syndrome.¹ In 1962, Fontana and Edwards reported a series of 58 postmortem specimens and demonstrated that most patients had died at a young age.² An embryological defect during fetal cardiac development leads to the left

coronary artery arising from the pulmonary artery instead of the aorta. At birth, the infant is asymptomatic but as the pulmonary artery pressure decreases during the neonatal period, desaturated blood flows under low pressure from the pulmonary artery via the left coronary artery to the left ventricle.³ This predisposes one to myocardial ischemia. Collateral vessels develop between the right and left coronary arteries and may provide adequate perfusion of the left ventricular myocardium. Subsequently, as the pulmonary resistance decreases, a retrograde flow from the high-pressure coronary arteries to the pulmonary trunk

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results in myocardial steal and contributes to myocardial ischemia.^{3, 4} Over time, there is anterolateral myocardial infarction, mitral valve dysfunction, and congestive cardiac failure. The presenting features are paroxysms of irritability, which correlate with episodes of angina pectoris and symptoms of heart failure. ALCAPA can occur in isolation or in tandem with other congenital cardiac lesions. Dilated cardiomyopathy is an important differential diagnosis and may also arise as a result of ALCAPA. Although ALCAPA presents predominantly in infancy, there are several case reports in adolescents and adults,^{5, 6} with the oldest reported patient being 72 years at diagnosis.⁷

Case Report

A 60-year-old man referred to our center due to dyspnea on exertion from one year previously, which was in New York Heart Association (NYHA) functional class II, without any history of chest pain. He had almost been normal during his life, carrying out his ordinary activities without limitation. He was a past smoker for several years and also was a current opium user. There was no history of systemic hypertension, diabetes, or dyslipidemia. At presentation, blood pressure was normal and a holosystolic murmur of grade III/VI at the apex and another diastolic murmur at the left sternal border were detected. A twelve-lead electrocardiogram showed normal sinus rhythm with left-axis deviation and no Q wave or ST-T changes. Chest X-ray showed marked cardiomegaly (cardiothoracic ratio = 60%) and pulmonary venous congestion. Lab data were normal. Transthoracic echocardiography demonstrated severe left ventricular enlargement with moderate dysfunction [left ventricular end-diastolic dimension (LVEDD) = 7 cm, left ventricular end-systolic dimension (LVESD) = 4.5 cm, left ventricular ejection fraction (LVEF) = 40%] and mild mitral and tricuspid insufficiency along with mildly 35 (mmHg) increased systolic pulmonary artery pressure. There was regional wall motion abnormality in the inferior wall and multiple dilated coronary branches through the inferior wall extending toward the apex (Figures 1 & 2). Transesophageal echocardiography showed dilated coronary branches in both right and left territories. The origin of the right coronary artery from the right sinus of Valsalva was seen; however, in spite of dilated left coronary branches, its connection to the aorta could not be visualized (Figure 3).

Electrocardiographically-gated multi-detector computed tomographic (CT) angiography revealed ALCAPA, with a retrograde flow from the left coronary artery to the pulmonary artery and extensive collateral vessels at the left ventricle apex (Figure 4).

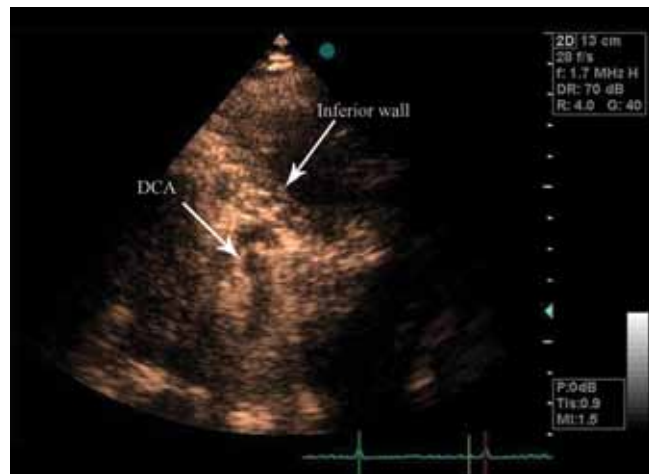
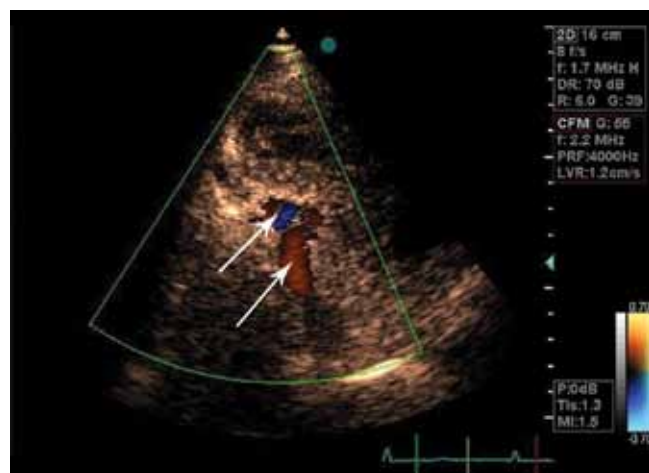
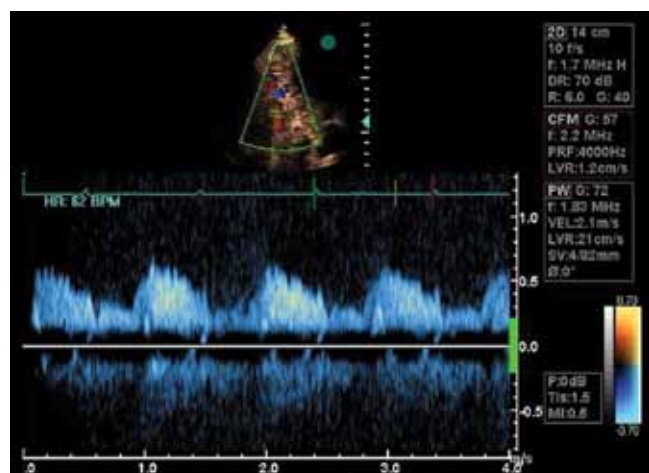


Figure 1. Apical two-chamber off axis echocardiography view shows dilated coronary artery (DCA) along the inferior wall

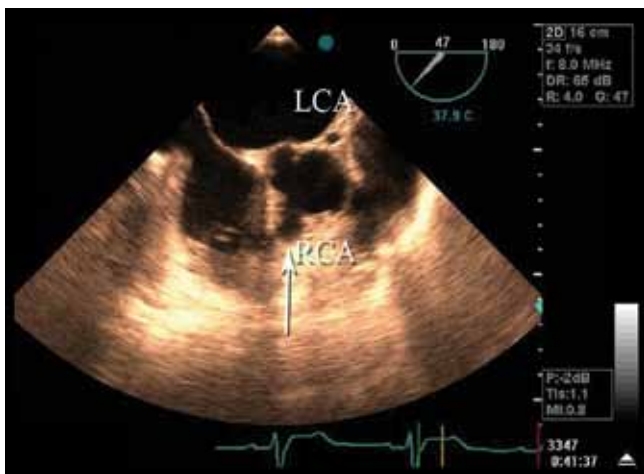


A



B

Figure 2. Color Doppler flow (A) shows diastolic flow in dilated coronary artery branch (arrows). Pulsed wave Doppler (B) revealed low velocity diastolic flow in coronary artery lumen

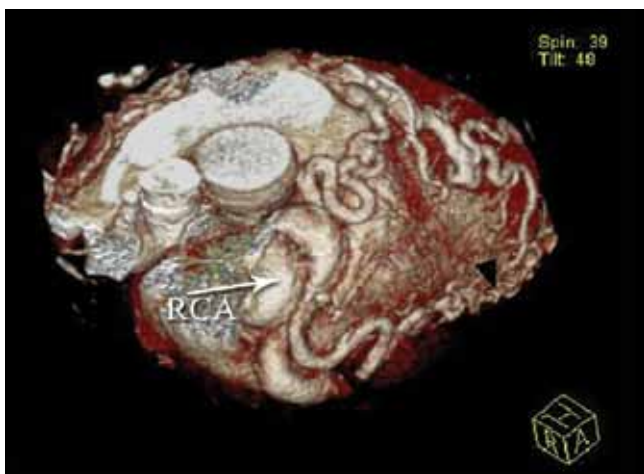


A

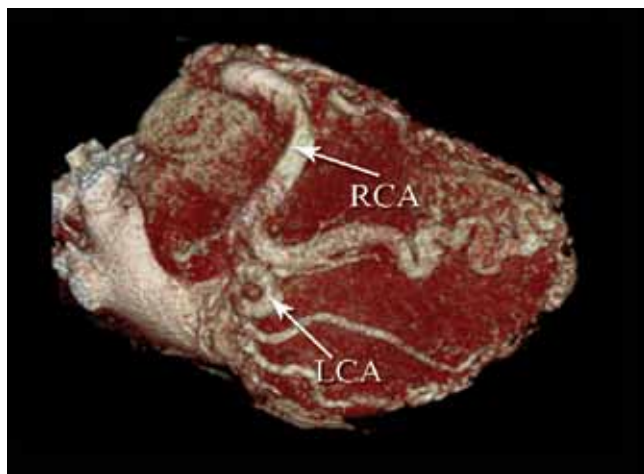


B

Figure 3. Transesophageal short-axis view (A) shows that right coronary artery (RCA) ostium is severely dilated (arrow) but left coronary artery (LCA) is not seen. The other Transesophageal view (B) shows dilated left coronary branches between aorta and left atrial appendage (LAA)



A



B

Figure 4. Coronary CT angiography shows: severely dilated right coronary artery (RCA) along with extensive collateral vessels at apical portion while left coronary artery (LCA) ostium was not seen (A). Dilated left coronary branches, are connected to pulmonary artery (B)

Discussion

ALCAPA is a rare but serious congenital anomaly. Presently, the prognosis for patients with ALCAPA is dramatically improved as a result of both early diagnosis using echocardiography with color flow mapping and improvements in surgical techniques, including myocardial preservation.

ALCAPA anomaly may result from abnormal septation of the conotruncus into the aorta and pulmonary artery,¹ or from persistence of the pulmonary buds together with involution of the aortic buds that eventually form the coronary arteries.² There are two types: the infant type and the adult type, each of which has different clinical manifestations and carries a different prognosis.

Nowadays, the prognosis for patients with ALCAPA is dramatically improved as a result of both early diagnosis using echocardiography with color flow mapping and electrocardiographically-gated multi-detector CT angiography and improvements in surgical techniques.

In asymptomatic adults, the recent literature suggests that if only moderate chronic ischemia and limited necrosis are present, survival without surgical correction is possible.

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