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Case Report

# A case of anomalous origin of the left coronary artery from the pulmonary artery presenting with sudden cardiac arrest due to coronary artery steal generated by excessive exercise

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ABSTRACT

This case report describes a 43-year-old man who temporarily survived cardiac arrest that was prospectively related to ventricular fibrillation due to the anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). Prior to admission to our hospital, he was asymptomatic for ALCAPA syndrome. Emergent coronary angiography revealed that the dilated right coronary artery was connected with extensive collateral vessels to the left coronary artery. The origin of the latter was in the pulmonary artery. Moreover, coronary steal phenomenon was identified by examining the pulmonary arterial blood oxygen saturation. The patient later died of acute decompensated acidosis.

<**Learning objective:** It is common that oxygen saturation exams may be normal or show a slight increase within the pulmonary artery in this anomaly. However, moderate to severe left to right shunt was remarkably detected in this patient, which may have contributed to acute ischemia in addition to inadequate collateral flow.>



## Introduction

Coronary artery anomalies (CAA) occur in less than 1% of the general population. CAA may involve an abnormal origin (number, location, size, angle of origination), course (intramural aortic), termination, or structure (fistula) of the coronary arteries [1]. Most CAAs are clinically insignificant and are discovered as incidental findings during coronary angiography. The most common CAA in adults is the coronary artery fistula which could cause ischemia due to the coronary steal phenomenon [2]. Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) syndrome, also known as Bland–White–Garland (BWG) syndrome, is a rare type of congenital heart disease with an estimated incidence of 1 in every 300,000 live births [3]. In patients with BWG syndrome, only 10% survive beyond childhood, and often have varying symptoms of myocardial ischemia, impaired left ventricular function, mitral regurgitation, and progressive heart failure, depending on the development of collateral vessel circulation. Sudden cardiac death is another major clinical manifestation, and is sometimes the first indication of BWG Syndrome in patients [3,4]. We report herein a case of BWG syndrome which survived sudden cardiac arrest diagnosed by coronary angiography and right-sided blood sampling.

## Case presentation

In September 2015, a 43-year-old man experienced cardiac arrest while excessively exercising by riding a bicycle up a steep hill in a road race. His friend, who was awakened by this abnormal state, found him unconscious and pulseless and immediately called emergency medical services, and initiated cardiopulmonary resuscitation. Emergency medical services arrived 35 min after receiving the emergency call and found that he had ventricular fibrillation by automatic analyzing system of automated external defibrillator (AED). He was successfully converted to sinus rhythm

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with an AED, and was then intubated and transported to the emergency room.

The patient was admitted to our hospital after experiencing sudden cardiac arrest with a history of syncope but no family history of sudden death. On physical examination, a 2/6–3/6 systolic murmur was heard at the second intercostal space along the left sternal border. In addition, mild coarse crackle was also heard in the lung zones. The electrocardiogram (ECG) administered upon admission showed ST elevation in leads $aV_R$, $V_1$, and ST depression in the inferolateral leads (Fig. 1A). Heart enlargement and pulmonary congestion were observed in the chest X-ray (Fig. 1B). Echocardiography indicated mild hypokinesia of the left ventricular anterior wall with an ejection fraction of 53% and normal heart valve function. The patient was transferred to the cardiac catheterization laboratory for coronary angiography and further evaluation. Coronary angiography showed an ALCAPA with retrograde filling through collateral vessels from an enlarged right coronary artery (Fig. 2). Next, measurements of the pressures and oxygen saturations in the right atrium, right ventricle, and right pulmonary artery were obtained with standard Swan–Ganz catheters using standard methods during the heart catheterizations. This examination identified the oxygen step-up, in which the saturation of blood in the pulmonary artery was 72.2%, which was higher than that of the blood in the right atrium and ventricle (Table 1). In addition, the left to right shunt ratio was calculated as 27.9%. In consideration of each examination performed, the diagnosis of ALCAPA syndrome was confirmed. The patient died of uncontrolled acidemia and unstable hemodynamic status a few hours following hospital admittance.

## Discussion

In 1886, Brooks first described ALCAPA. This anomaly, also known as BWG syndrome, accounts for approximately 0.25% to 0.5% of congenital heart defects [5]. The clinical presentation in infants is well defined since the symptoms usually occur in the first few months. In the absence of surgical intervention, death occurs in 93% of patients with BWG syndrome. Late presentation of BWG syndrome in the adult is extremely rare. There are three hemodynamic phases in the evolution of this anomaly that have

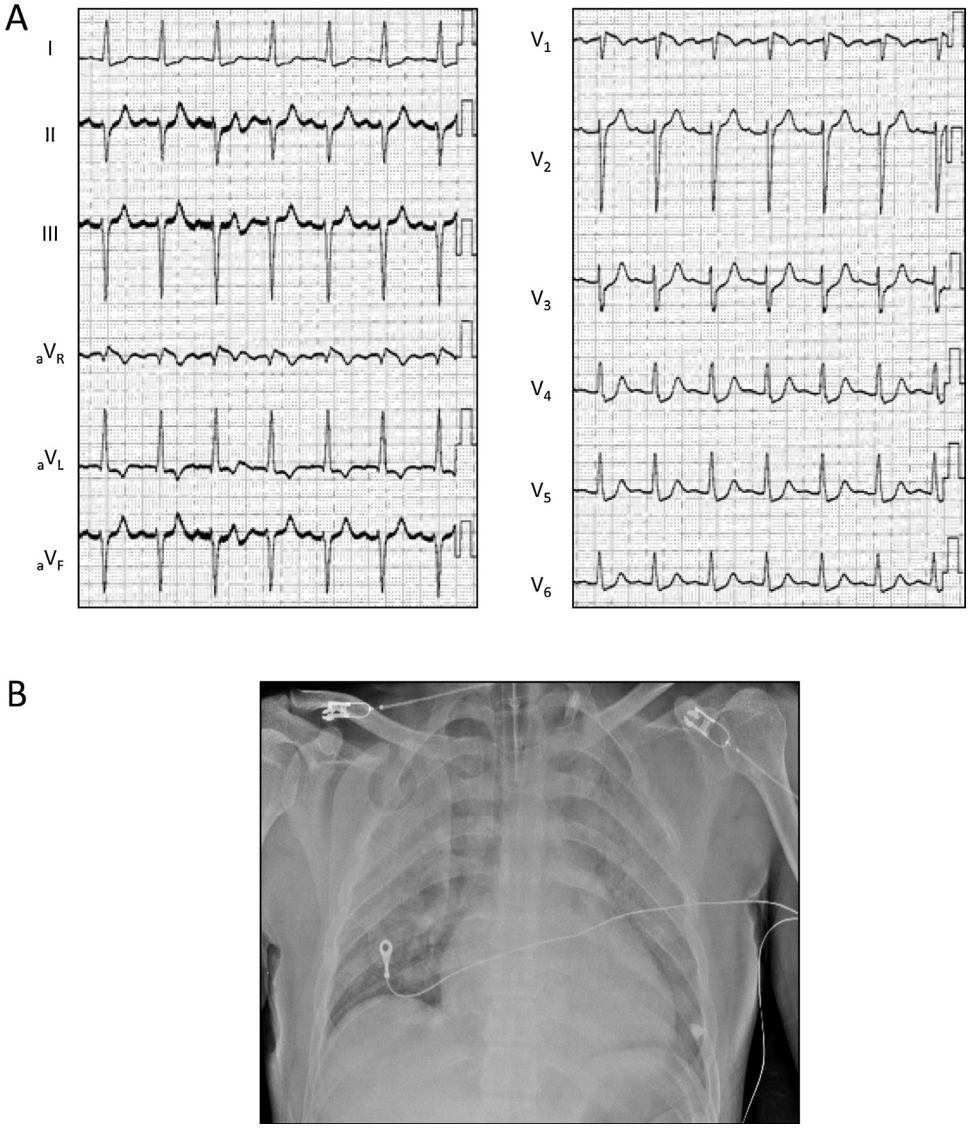


**Fig. 1.** (A) The 12-lead electrocardiogram upon admission. ST segment was elevated in leads, $aV_R$, $V_1$, and was depressed in the inferolateral leads. (B) The chest X-ray upon admission. Heart enlargement and pulmonary congestion was observed in the chest X-ray.

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