



## Anomalous left coronary artery from pulmonary artery (ALCAPA) in asymptomatic 11-year-old case

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### Summary

Origin of the left coronary artery from the pulmonary trunk [Anomalous origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA)] is a rare congenital heart defect. ALCAPA is one of the common causes of myocardial ischemia or infarction in infants and children. In this report, we present an eleven-years-old patient presenting with murmur and accompanied by echocardiographic findings of dilated cardiomyopathy and angiocardiographic investigation that revealed left coronary artery originating from the pulmonary artery. (*Turk Arch Ped* 2012; 47: 294-297)

**Key words:** ALCAPA, coronary artery anomalies, echocardiography

### Introduction

“Anomalous origin of the Left Coronary Artery from the Pulmonary Artery” (ALCAPA) constitutes 0.023% of congenital cardiac anomalies and occurs with an incidence of 1/300 000 live births (1). Since the pulmonary arterial pressure in the abnormal left coronary artery originating from the pulmonary artery is low, it can not supply the myocardium sufficiently. These myocardium areas which are not sufficiently supplied try to be supplied by the collaterals which develop from the right coronary artery. Clinical findings occur according to the severity of myocardial dysfunction. If signs of congestive heart failure occur in the infancy, it is named as “Bland-White-Garland Syndrome” and it constitutes 83% of all these patients (2). Abnormal left coronary artery originating from the pulmonary artery is one of the leading causes of myocardial ischemia and/or infarction in infancy (3). If untreated, its prognosis is poor and the mortality rate is above 90% in the first year (4).

The clinical prognosis depends mainly on the size of the collaterals which are present between the right and left coronary artery. In symptomatic patients, heart failure, cardiogenic shock, valve failures and infarction may be observed in relation with impaired myocardial blood supply

(5,6). Only 15% of the patients develop sufficient collaterals and reach adolescence or adulthood, but these patients have a high risk of sudden death (7,8). In this article, a patient who reached the age of 11 without symptoms, referred because of a murmur heard on physical examination and diagnosed as ALCAPA as a result of catheterization-angiography performed upon findings on echocardiogram was presented to emphasize that patients with this anomaly can reach advanced ages without complaints.

### Case

An 11-year-old male patient who had no complaints and who was found to have a cardiac murmur on physical examination was referred to our Pediatric Cardiology Outpatient Clinic. On physical examination, body weight was found to be 40 kg (50-75<sup>th</sup> percentile), height was found to be 145 cm (50-75<sup>th</sup> percentile), pulse rate was found to be 90/min, respiratory rate was found to be 20/min and blood pressure was found to be 110/70 mmHg. Peripheral pulses were normal. The first and second heart sounds were natural. No additional sound was heard. A 2-3/6 systolic ejection murmur was heard in the 3-4<sup>th</sup> intercostal space in the left side of the sternum. Telecardiography revealed a

cardiothoracic ratio of 0.48 and bronchovascular appearance was normal (Figure 1). Electrocardiographic findings were as follows: sinus rhythm, heart rate 90/min, QRS axis +85° and all findings were normal. There was no finding related to ischemia, myocardial damage or infarction (Figure 2).

Echocardiography revealed enlarged left cardiac chambers and mild-moderate mitral failure. The right coronary artery originating from the aorta was visualized as tortuous and enlarged and the left coronary artery could not be visualized completely (Figure 3). Colored Doppler echocardiography revealed enlarged, reverse flow into the pulmonary artery and two dimensional examination afterwards revealed that this was the left coronary artery and was divided into two branches as left anterior artery and "circumflex" artery. Colored Doppler revealed flows which might be compatible with the collateral circulation between the left coronary artery and right coronary artery on the interventricular septum (Figure 4). Left ventricular function was normal (Ejection fraction: 65%, shortening fraction: 35%). Catheterization-angiography was decided to be performed considering ALCAPA as a result of echocardiographic examination. On angiographic examination, it was observed that the right coronary artery was tortuous and enlarged, the left coronary artery was supplied by the

collaterals originating from the right coronary artery and originated from the pulmonary artery (Figure 5). Aortic root injection revealed that the left coronary artery was not originating from the aorta (Figure 6). The patient was referred to the department of cardio-thoracic surgery for operation.

### Discussion

Anomalous origin of the left coronary artery from the pulmonary artery is a significant condition which is observed rarely, but which may lead to myocardial ischemia, myocardial infarction and sudden death in childhood. Currently, surgical intervention is recommended to be performed as soon as possible after the diagnosis is made.



Figure 1. Normal telecardiogram showing no cardiomegaly

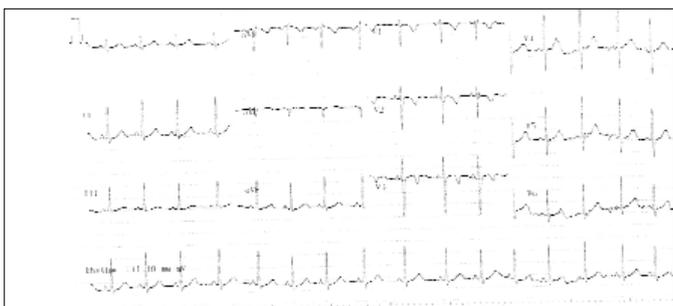


Figure 2. Electrocardiogram with normal sinus rhythm without ischemic findings



Figure 3. On parasternal short-axis imaging, enlarged and tortuous right coronary artery was observed, while the left coronary artery could not be visualized

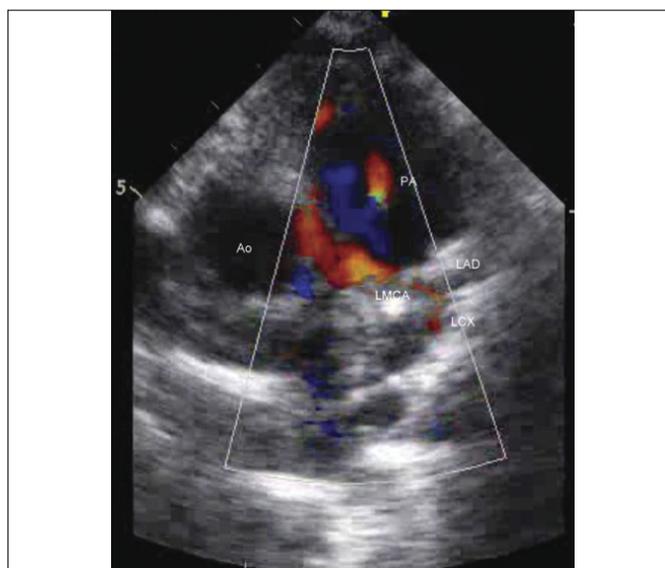
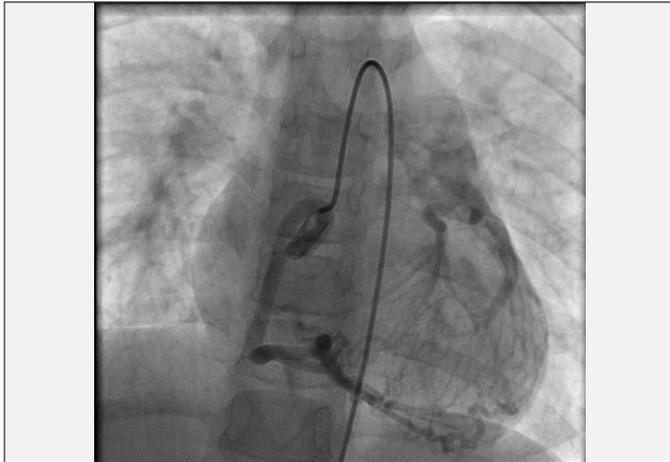


Figure 4. On parasternal short-axis imaging, origin of the left coronary artery from the pulmonary artery



**Figure 5.** Angiography reveals that the right coronary artery is dilated and tortuous, the left coronary artery is supplied by the collateral arteries originating from the right coronary artery and the left coronary artery originates from the pulmonary artery



**Figure 6.** Aortic root injection reveals that the right main coronary artery originates from the aorta, while the left main coronary artery does not originate from the aorta

In patients with anomalous left coronary artery originating from the pulmonary artery, the time of clinical signs and symptoms varies. The time of occurrence of signs and symptoms is related to the degree of myocardial blood supply by the collaterals which develop as the pulmonary pressure decreases. In symptomatic patients, clinical findings of heart failure are present. In infants, symptoms may appear with sudden crying spells accompanied by feeding difficulties, sweating and pallor. At more advanced ages, growth failure, tachycardia due to left ventricular contraction failure and progressive enlargement; dyspnea due to pulmonary venous congestion and hepatomegaly due to systemic venous congestion may be found (9). While a small proportion of the patients reach adolescence and adulthood due to sufficient development of collateral arteries, 85% are lost in the first year of life because of insufficient collateral arteries (10,11,12,13). In our patient, none of these clinical findings were observed.

Clinical findings alone are not sufficient in the diagnosis of anomalous origin of the left coronary artery from the pulmonary artery. On electrocardiogram, findings of acute or chronic anterior-lateral infarction are usually present. Q wave and changes of ST segment and T wave are observed in DI, aV Land V4-6 (3,4,9). Cardiomegaly and pulmonary venous congestion may be observed on telecardiogram. In our patient, electrocardiogram and telecardiogram were found to be normal. Two-dimensional echocardiographic examination may demonstrate that the left coronary artery originates from the pulmonary artery. Robinson et al. (14) reported that they false negatively observed the origin of the left coronary artery to be normal in their series of three patients and the origin of the left coronary artery should be demonstrated echocardiographically or angiographic evaluation should be done in patients in whom ALCAPA is suspected (14).

When enlarged right coronary artery, left ventricular enlargement and contraction failure are observed on echocardiogram, ALCAPA should be suspected. In these patients, demonstration of multiple flows related to collateral circulation between the left and right coronary arteries on the interventricular septum by colored Doppler and presence of reverse flow from the coronary artery into the pulmonary artery on colored Doppler echocardiogram suggest this anomaly. In the differential diagnosis of ALCAPA, dilated cardiomyopathy should be considered.

In our patient, enlargement in the diameter of the right coronary artery, enlargement in the chambers of the left heart and a flow compatible with multiple collateral circulation on the interventricular septum on colored Doppler were found. While some centers think that echocardiography is sufficient for the diagnosis, some others believe that enlarged and tortuous right coronary artery, the collateral arteries originating from the right coronary artery and transfer of the injected contrast material into the pulmonary artery should be demonstrated by injection of contrast material into the aortic root (1).

Definite treatment is performed surgically. After surgical treatment restructure of the left ventricle occurs and a progressive improvement is observed in left ventricular functions related to sufficient blood supply to the myocardium (1,15,16). Surgical treatment consists of addition of the coronary artery which originates from the pulmonary artery to the aorta or association of the coronary artery with the aorta by a tunnel through the pulmonary artery (Takeuchi method) (15).

Conclusively, ALCAPA can be treated surgically and can present asymptotically at advanced ages, if well developed collateral arteries are present as in our patient.

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