



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

### Yetişkin Bir Hastada Pulmoner Arterden Orjin Alan Sol Ana Koroner Arter Çıkış Anomalisi

Sol Ana Koroner Arter Çıkış Anomalisi / Anomalous Origin of the Left Main Coronary Artery

Yunus Nazli<sup>1</sup>, Omer Nuri Aksoy<sup>1</sup>, Kemal Korkmaz<sup>2</sup>, Ismail Olgun Akkaya<sup>1</sup>, Necmettin Colak<sup>1</sup>  
<sup>1</sup>University of Turgut Ozal, Faculty of Medicine, <sup>2</sup>Numune Research and Training Hospital, Department of Cardiovascular Surgery, Ankara, Turkey

#### Özet

Garland-Bland-White olarak bilinen sol ana koroner arterin anormal olarak pulmoner arterden çıkması, oldukça nadir fakat ölümcül bir konjenital kardiyovasküler anomalidir ve sıklıkla izole bir durumdur. Biz göğüs ağrısı ve nefes darlığı şikayeti nedeniyle cerrahi düzeltme gerektiren pulmoner arterden köken alan sol ana koroner arter çıkış anomalili 32 yaşında alışılmadık bir hastayı sunduk. Bu anomali basitçe sol ana koroner arterin bağlanması ve koroner arter bypass greft operasyonu ile onarıldı.

#### Anahtar Kelimeler

Konjenital Kalp Anomalisi; Koroner Arter Bypass Greftleme

#### Abstract

Anomalous origin of the left main coronary artery from the pulmonary artery (ALCAPA), also known as Garland-Bland-White syndrome, is an extremely rare but potentially fatal congenital cardiovascular anomaly and it often exists as an isolated condition. We report an unusual case of a 32 years-old patient with ALCAPA presenting with chest pain and dyspnea who underwent surgical correction of this rare anomaly. This anomaly was simply repaired by the combination of LMCA ligation and coronary artery bypass grafting.

#### Keywords

Congenital Heart Defect; Coronary Artery Bypass Grafting

DOI: 10.4328/JCAM.2501

Received: 18.04.2014 Accepted: 30.04.2014 Printed: 01.08.2013

J Clin Anal Med 2013;4(suppl 4): 446-8

Corresponding Author: Yunus Nazli, Alparslan Turkes Cad. No: 57 Emek, Ankara, 06510, Turkey.

T: +903122035186 E-Mail: yunusnazli@gmail.com

## Introduction

The anomalous origin of the left main 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 as the Bland-White- Garland syndrome [1]. ALCAPA is a rare congenital cardiovascular defect with an incidence of 1 in 300,000 live births. It is the most common anomaly of the coronary vasculature, with a frequency of 0-5 % of all congenital cardiac defects [2]. An embryological defect during fetal cardiac development leads to the left coronary artery arising from the pulmonary artery instead of the aorta. In patients with ALCAPA the pulmonary vascular resistance and pulmonary arterial pressure decrease shortly after birth, along with oxygen content of the pulmonary artery [3]. This causes a drop in antegrade flow and oxygen content of the anomalous left coronary artery, leading to myocardial ischemia. This may progress to myocardial infarction during periods of increased myocardial oxygen consumption. Collateral circulation between the right and left coronary systems ensues and left coronary artery flow reverses and enters in the pulmonary trunk due to the low pulmonary arterial pressure (coronary steal phenomena). Consequently, the myocardium remains inadequately perfused (fixed ischemia).

We report an unusual case of a 32 years-old patient with ALCAPA presenting with chest pain and dyspnea who underwent surgical correction of this rare anomaly.

## Case Report

A 32-year-old woman referred to our center with complaints of chest pain and dyspnea on exertion for three months, which was evaluated as in New York Heart Association (NYHA) functional class II. She had almost been normal during her life, carrying out her ordinary daily activities without limitation. There was no history of systemic hypertension, diabetes, or dyslipidemia. At presentation, blood pressure was normal. There was no murmur. A twelve-lead electrocardiogram showed normal sinus rhythm and without any Q wave or ST-T changes. Chest X-ray showed marked pulmonary venous congestion. Laboratory data were normal. Transthoracic echocardiography demonstrated left ventricular hypertrophy; left atrial dilatation (46 mm), slightly decreased left ventricular ejection fraction [LVEF = 55%] and mild mitral insufficiency and pulmonary artery pressure was 35 mmHg. The patient was transferred to the cardiac catheterization laboratory for coronary angiography and further evaluation. Coronary angiography showed an anomalous left coronary artery arising from the posterolateral of common pulmonary artery with retrograde filling through collaterals from a highly developed apparent right coronary artery (Figure 1). Multi-detector computed tomographic (CT) angiography revealed ALCAPA. Left main coronary artery originated from main pulmonary trunk (figure 1). Calibration of the main pulmonary artery was measured as 28 mm. The left main coronary artery ends in the form of trifurcation. Calibration of the LAD was increased and measured as 20 mm. Calibration of the left circumflex artery was within the normal range of 4 mm wide. Intermediate artery was patent. The left atrium was dilated and measured as 47 mm. Right coronary artery was dilated.

At operation, aortic cannulation and venous cannulation through right atrium was performed following median sternotomy. Then we started cardiopulmonary bypass. Patient was cooled at 28°C degree and aorta was cross-clamped. Cardiac arrest was obtained by antegrade and retrograde cardioplegia. The anomalous origin of the left main coronary artery was sutured with the pledgeted polypropylene sutures from the pulmonary artery. Then, the left internal mammary artery was grafted to the LAD and saphenous vein was grafted to the Circumflex artery (figure 2).

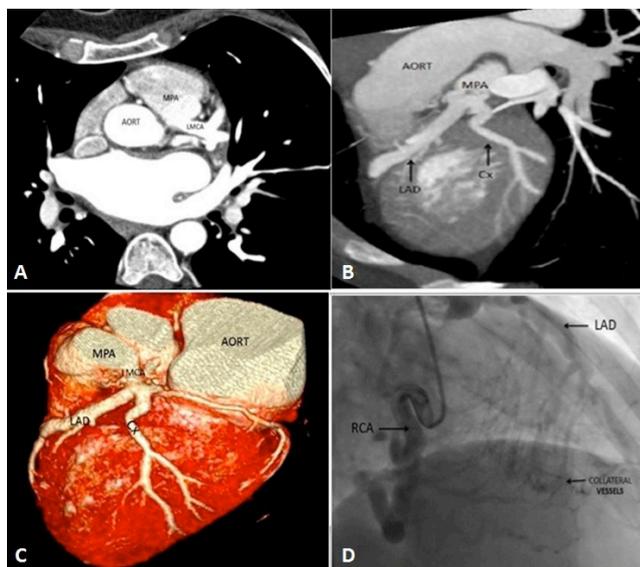


Figure 1. (A) Coronary CT-angiography showing an anomalous origin of the left main coronary artery (LMCA) from the main pulmonary artery (MPA). Axial (A), curved multiplanar reconstruction (B) and Volume-rendered reformation (C). (D) Coronary angiography of the right coronary artery showing collateral filling of the left coronary vascular territory, which connects directly to the pulmonary trunk.

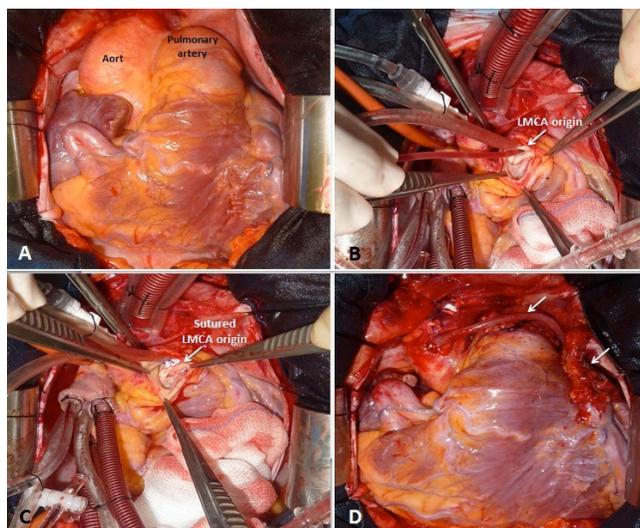


Figure 2. Operative photographs showing (A) extended right coronary artery, left anterior descending artery, (B) left main coronary artery origin in the pulmonary artery, (C) sutured LMCA origin, (D) coronary artery bypass grafts (with left internal thoracic artery and saphenous vein).

## Discussion

Anomalous connection of left coronary artery to pulmonary trunk is a rare condition, occurring in 0.26% of patients with congenital heart disease undergoing cardiac catheterization [4]. The anomalous left main coronary artery (LMCA) connects most often to the sinus of Valsalva immediately above the left

or posterior cusp of the pulmonary trunk and rarely from that above the right cusp [5,6]. The left main coronary artery is of variable length but usually divides into anterior descending and circumflex branches within 5 or 6 mm of its origin. Collateral communications between right and left coronary arteries are always present but vary in extent and are grossly visible in only a few cases, mainly in adults. Uncommonly, only the circumflex branch connects anomalously to the pulmonary trunk, and rarely only the left anterior descending branch connects anomalously [7,8]. The left ventricle is always hypertrophied and usually greatly dilated, with dilatation often involving primarily the left ventricular apex [9]. Several pathologic features may result in mitral valve regurgitation. The chest radiograph may be normal or may show cardiac enlargement.

85% of all cases of ALCAPA present within the first two months of life. About 65% of infants born with it die during the first year from intractable left ventricular failure [6]. If death does not occur during the first year, the hazard lessens considerably and the chronic phase of natural history is reached. Survival to this stage may be related to presence of rich interarterial collaterals, possibly associated with a slightly restrictive opening between left coronary artery and pulmonary trunk. Many such patients are in good health, and a few have normal ECGs. Survival beyond the first year may also be related to marked right coronary dominance, with this vessel supplying not only the diaphragmatic portion of the left ventricle but also much of the septum and lateral wall [6]. When severe symptoms do not occur in infancy, presentation is often delayed to beyond age 20 years. In our case, Collateral circulation from the right coronary artery is apparently adequate to prevent massive infarction [10]. Nowadays, the prognosis for patients with ALCAPA is dramatically improved as a result of both early diagnosis using echocardiography with color flow mapping, electrocardiographically gated multi-detector CT angiography and Coronary angiography.

Several surgical techniques have been tried, but each has a drawback. Direct re-implantation of the left main coronary artery into the aorta is often technically difficult especially in adults, due to the distance between the aorta and the anomalous orifice. The combination of LMCA ligation and coronary artery bypass grafting (CABG) is the best technique in this era. The other surgical technique is a creation of a baffle through the pulmonary artery (Takeuchi procedure). Although re-implantation of the LMCA to the aorta remains the most physiological correction for this anomaly, the combination of LMCA ligation and CABG provides a dual coronary flow system and is preferable when re-implantation is impossible. In our case, re-implantation of the LMCA to the aorta was considered unfeasible because of the distance between the insertion site of the LMCA on the pulmonary artery and the aorta. Therefore, the combination of LMCA ligation and CABG was preferred because of technically simple technique in this case. The left internal mammary artery was used to graft the anomalous left coronary artery in our patient. It has satisfactorily established antegrade flow into the left coronary artery and should maintain patency. Restoration of a dual coronary system will prevent further ischemia and arrhythmias of acute ischemic origin, but the anatomical substrate for ventricular arrhythmias in patients with old MI will

not be altered after revascularization.

### Competing interests

The authors declare that they have no competing interests.

### References

1. Brooks HS. Two Cases of an Abnormal Coronary Artery of the Heart Arising from the Pulmonary Artery: With some Remarks upon the Effect of this Anomaly in producing Cirroid Dilatation of the Vessels. *J Anat Physiol* 1885;20(1):26-9.
2. Bland EF, White PD, Garland J: Congenital anomalies of the coronary arteries: Report of an unusual case associated with cardiac hypertrophy. *American Heart Journal* 1933;8:787-801.
3. Riedel M, Hall RJC, Haworth SG. Disorders of the pulmonary circulation. In: Julian DG, Camm AJ, Fox KM, Hall RJC, Poole-Wilson PA, editors. *Diseases of the Heart*. London: WB Saunders; 1996. p.1237-63.
4. Askenazi J, Nadas AS. Anomalous left coronary artery originating from the pulmonary artery. Report on 15 cases. *Circulation* 1975;51(6):976-87.
5. Smith A, Arnold R, Anderson RH, Wilkinson JL, Qureshi SA, Gerlis LM, et al. Anomalous origin of the left coronary artery from the pulmonary trunk. Anatomic findings in relation to pathophysiology and surgical repair. *J Thorac Cardiovasc Surg* 1989;98(1):16-24.
6. Wesselhoeft H, Fawcett JS, Johnson AL. Anomalous origin of the left coronary artery from the pulmonary trunk. Its clinical spectrum, pathology, and pathophysiology, based on a review of 140 cases with seven further cases. *Circulation* 1968;38(2):403-25.
7. el Habbal MM, de Leval M, Somerville J. Anomalous origin of the left anterior descending coronary artery from the pulmonary trunk: recognition in life and successful surgical treatment. *Br Heart J* 1988;60(1):90-2.
8. Roberts WC, Robinowitz M. Anomalous origin of the left anterior descending coronary artery from the pulmonary trunk with origin of the right and left circumflex coronary arteries from the aorta. *Am J Cardiol* 1984;54(10):1381-3.
9. Rittenhouse EA, Doty DB, Ehrenhaft JL. Congenital coronary artery cardiac chamber fistula. *Ann Thorac Surg* 1975;20(4):468-85.
10. Moodie DS, Fyfe D, Gill CC, Cook SA, Lytle BW, Taylor PC, et al. Anomalous origin of the left coronary artery from the pulmonary artery (Bland-White-Garland syndrome) in adult patients: long-term follow-up after surgery. *Am Heart J* 1983;106(2):381-8.

### How to cite this article:

Nazli Y, Aksoy ON, Korkmaz K, Akkaya İO, Colak N. Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) in an Adult. *J Clin Anal Med* 2013;4(suppl 4): 446-8.