

*Coronary Anomalies*

# Surgical Reconstruction of the Anomalous Left Coronary Artery From the Pulmonary Artery

Slobodan Micovic, MD, PhD<sup>1,2</sup>; Petar Milacic, MD<sup>1,2</sup>; Miroslav Milicic, MD, PhD<sup>1,2</sup>; Milica Brkovic, MD<sup>3</sup>; Petar Vukovic, MD, PhD<sup>1,2</sup>; Igor Zivkovic, MD<sup>1,2</sup>

<sup>1</sup>Department of Cardiac Surgery, Dedinje Cardiovascular Institute, Belgrade, Serbia

<sup>2</sup>School of Medicine, University of Belgrade, Belgrade, Serbia

<sup>3</sup>Department of Radiology, Dedinje Cardiovascular Institute, Belgrade, Serbia

## Abstract

The anomalous left coronary artery from the pulmonary artery is a rare congenital disease. Early surgical reconstruction is mandatory to prevent adverse cardiac events. Direct coronary button transfer, vein graft interposition, ligation, and coronary artery bypass construction are the most commonly used techniques. This case report presents a modified technique of Dacron graft interposition and reimplantation anomalous left coronary artery from the pulmonary artery on the ascending aorta.

**Keywords:** Coronary vessel anomalies; Bland White Garland syndrome; ALCAPA syndrome; pulmonary artery; anomalous left coronary artery

## Introduction

The anomalous left coronary artery arising from the pulmonary artery (ALCAPA) is a rare congenital disease.<sup>1</sup> Clinical presentation depends on the extent of acquired collateral circulation between 2 coronary systems. The infant type (early expression of the disease) characterizes a lack of collateral coronary circulation. Abnormal left ventricular perfusion and myocardial ischemia lead to death in 90% of infants in the first year of life.<sup>2</sup> Approximately 10% to 15% of patients with ALCAPA develop adult-type clinical presentations in late life.<sup>3</sup> Surgical correction to reestablish the 2 coronary systems is the gold-standard treatment. Here, the authors describe the advantages and disadvantages of various surgical techniques and the use of a Dacron graft for surgical reconstruction of an anomalous left coronary artery (LCA) in an adult patient.<sup>4</sup>

## Case Report

A 39-year-old woman was admitted to the hospital for elective mitral valve surgery. The first manifestation—fatigue on exertion—was registered a year before surgical treatment; soon after experiencing the initial symptoms, the patient was hospitalized for cardiac decompensation. Ultrasound examination showed severe mitral valve regurgitation. Preoperative coronary angiography revealed an enlarged right coronary artery and the absence of aortic origin for the LCA. Additional computed tomography (CT) scanning showed an anomalous origin of the LCA from the pulmonary trunk (Fig. 1A). Transesophageal examination revealed a global reduction in systolic function (ejection fraction, 25%-30%) without segmental contractile disturbance. End-diastolic and end-systolic diameters were 62 and 43 mm, respectively. Moderate to severe mitral valve regurgitation resulting from posterior leaflet prolapse was diagnosed.

## Technique

After median sternotomy, cardiopulmonary bypass was established using bicaval venous cannulation. After cardioplegic arrest, the left atrium was opened, and mitral valve inspection revealed primary chord elongation of the posterior leaflet 2 segment. The reconstruction was performed by triangular resection of the flail segment and

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**Corresponding author:** Igor Zivkovic, MD, Department of Cardiac Surgery, Heroja Milana Tepica 1, 11000 Belgrade, Serbia (igor88zivkovic@gmail.com)

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placement of a 32-mm annuloplasty ring. Then, the main pulmonary artery (PA) was opened 3 cm above the pulmonary valve. The orifice of the ALCAPA was detected close to the commissure between the right and left leaflets. Detachment of the LCA from the PA and then termino-terminal anastomosis with a 6-mm Dacron graft was performed. Proximal anastomosis of the graft was constructed on the ascending aorta (Fig. 2). The main PA was reconstructed with an extracellular matrix (ProxiCor; Aziyo Biologics) patch. The perioperative period was uneventful. A control CT scan showed the correct position and patency of the vascular graft (Fig. 1B). The patient was discharge on the sixth postoperative day.

Follow-up transthoracic echocardiography 1 month after the procedure showed appropriate function of the mitral valve without regurgitation and a left ventricular ejection fraction of 30%.

## Discussion

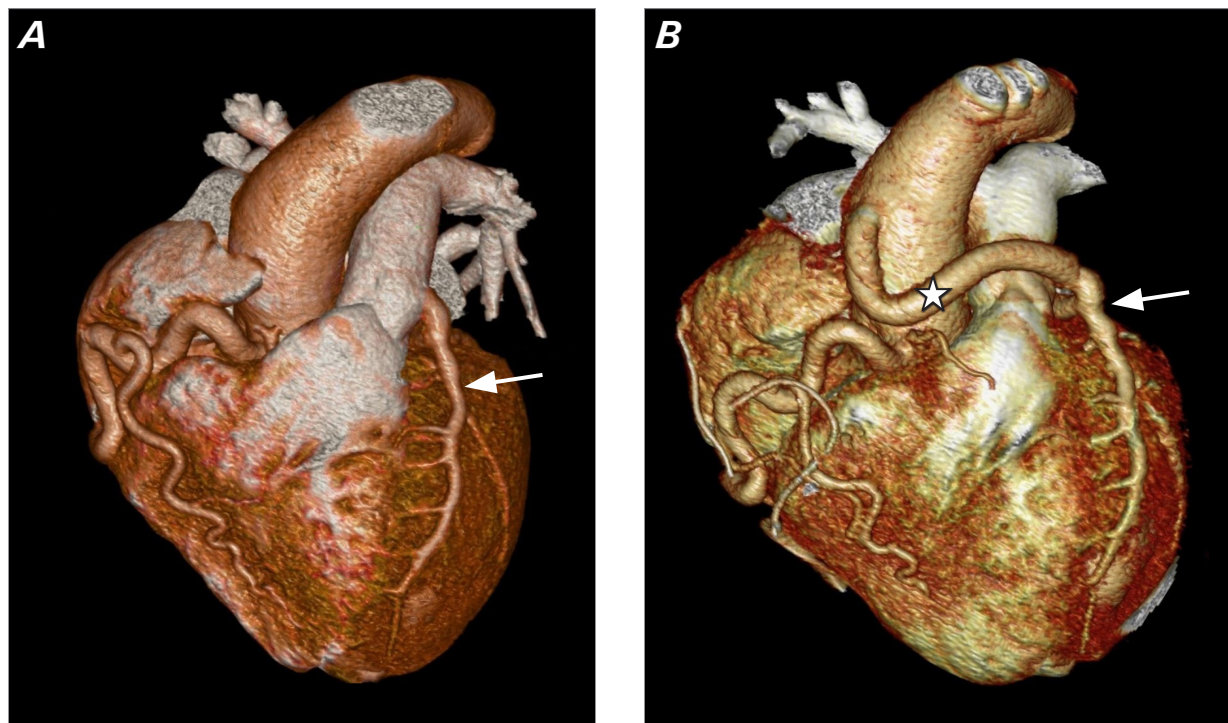
The ALCAPA supplies the left ventricle with low-oxygen-saturation blood. In addition, the left-to-right shunt and coronary “steal” phenomenon lead to abnormal left

## Abbreviations and Acronyms

ALCAPA	anomalous left coronary artery originating from the pulmonary artery
Ao	aorta
CT	computed tomography
LCA	left coronary artery
PA	pulmonary artery
RA	right atrium
RCA	right coronary artery
RV	right ventricle

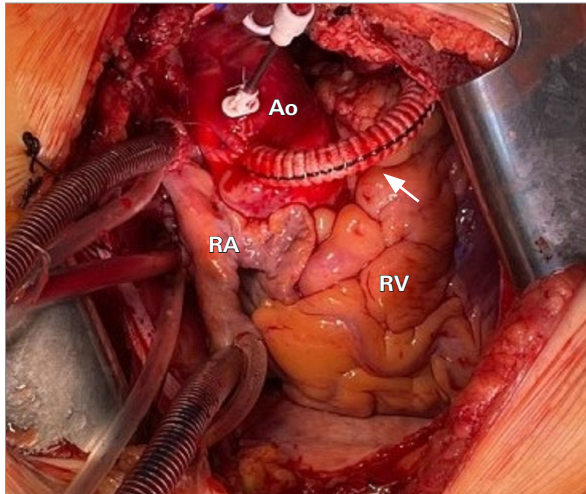
ventricular perfusion, which results in chronic heart ischemia, heart failure, and sudden death.<sup>2,4</sup> In most cases, myxomatous degeneration of the leaflets as well as ischemic dysfunction of the papillary muscles and adjacent myocardium may cause mitral insufficiency and prolapse.<sup>2</sup>

Transthoracic echocardiography is the most effective noninvasive method for diagnosing patients with ALCAPA. It can describe the origin of coronary arteries, their courses, hemodynamic changes, and other malformations. Magnetic resonance imaging is an excellent tool in the therapeutic decision algorithm in asymptomatic adults. When extensive delayed subendocardial en-



**Fig. 1** Multislice CT scan with contrast of the heart and great vessels. **A)** Presurgical CT scan shows anomalous left coronary artery (arrow) originating from the pulmonary artery. **B)** Postsurgical CT scan shows reimplanted anomalous left coronary artery (arrow) from the pulmonary artery in the systemic circulation. The star indicates the right coronary artery.

CT, computed tomography.



**Fig. 2** Surgical view of the interposed Dacron vascular graft (arrow).

Ao, aorta; RA, right atrium; RV, right ventricle.

hancement caused by infarction is seen on magnetic resonance images, surgery should be performed.<sup>2</sup> Immediate surgical reconstruction after diagnosis of ALCAPA is the gold-standard treatment. Establishing a dual coronary artery system is a safer treatment option than is ALCAPA ligation (single coronary system) because it lessens the chance of subendocardial ischemia and sudden death.<sup>2,5</sup>

The literature describes various surgical techniques. On December 23, 1963, Dr Denton Cooley performed the first successful coronary bypass reconstruction using a Dacron graft interposed between the aorta and a coronary artery.<sup>6</sup> Coronary button transfer is preferred in infants and children, but it is more difficult in adults because of the friability of the vessels and a loss of tissue elasticity.<sup>4</sup> The Takeuchi procedure (transpulmonary baffle) is used, but its most significant disadvantages are supralvalvular pulmonary stenosis, aortic valve insufficiency, baffle obstruction, and leak. The alternative method is reconstruction with vein graft interposition. A disadvantage of this technique is the potential risk of graft stenosis or occlusion after implanting a vein in the high-pressure arterial system. Midterm follow-up reported 80% patency after saphenous vein use in patients with ALCAPA.<sup>5</sup> The vein graft is also prone to kinking. Coronary artery bypass after ALCAPA ligation could be a good option. Excellent patency was registered with the single or bilateral left internal artery as a conduit for coronary artery bypass grafting. Data on the long-term patency rate of the internal thoracic artery after ALCAPA reconstruction are seemingly unavailable in the literature.<sup>2,4,5</sup> Concerns about this strategy exist based on competitive flow from the extensive collaterals and

consequent arterial graft failure. The natural course of the atherosclerotic process of used grafts could produce adverse events across the lifespan. Some surgeons claim that direct aortic reimplantation should be considered as a primary treatment option for adults.<sup>5</sup>

Treatment for mitral valve regurgitation remains controversial and depends on the degree of the regurgitation and its etiology (functional or structural). Ischemic mitral valve regurgitation could be treated in the case of severe mitral regurgitation. In most cases, significant improvement in mitral valve function was recognized 6 months after the procedure because of the resolution of myocardial hibernation. In contrast, correction of structural mitral valve dysfunction would be recommended for more than mild regurgitation, unless prolongation of cross-clamp time is deemed undesirable for the patient.<sup>7</sup>

In the described case, mobilization and direct implantation of the ALCAPA were not feasible. We used the interposition of the Dacron graft to reduce adverse events. The advantage of this approach is easy positioning, with a small risk of graft kinking. Ziganshin et al<sup>8</sup> showed excellent results during the 5 years of follow-up after coronary artery reconstruction using a Dacron graft (Cabrol procedure).

The ALCAPA demands immediate surgical reconstruction. This technique should be considered a viable treatment option, but it necessitates long-term follow-up to confirm appropriate benefit.

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