

Bland-White-Garland Syndrome in an 8-Year-Old Child

M. H. Nezafati MD, M. Abbasi MD and G. Soltani MD

Abstract

The anomalous origin of the left coronary artery from pulmonary artery (ALCAPA) or Bland-White-Garland syndrome is a rare congenital lesion. It is the most common congenital coronary artery anomaly (0.26% of patients with congenital heart disease) that results in myocardial ischemia and infarction in children.¹ ALCAPA usually presents in infancy when the pulmonary vascular resistance drops in the first few weeks of life.²

Depending on the degree of collateral development, myocardial ischemia or infarction may occur. Since 90% of such patients die during the first year of life,³ ALCAPA is rarely seen and reported in children (*Iranian Heart Journal 2007; 8 (4): 69-71*).

Key words: Bland-White- Garland syndrome ■ anomalous coronary artery

Case report

We report the case of an 8-year-old girl who had mild exertional fatigue and atypical chest pain. During routine physical examination, a grade II-III/VI systolic murmur was discovered at the lower left sternal border by the health care team at her school. She was referred to a pediatric cardiologist for further evaluation. An ECG showed pathologic Q waves in leads I and aVL and diffuse ST-T wave changes consistent with anterolateral ischemia. Transthoracic echocardiography demonstrated a huge tortuous right coronary artery originating from the pulmonary trunk with normal left ventricular (LV) function (ejection fraction {EF}=70%) and trivial mitral valve regurgitation (MR).

Coronary angiography demonstrated a tortuous right coronary artery, collateral circulation between the right and left coronary systems and entry of contrast from the left main trunk into the pulmonary artery (Figs. 1, 2).

Right heart catheterization showed an increase in blood oxygen content at the pulmonary artery (PA) level (left-to-right shunt).

The patient underwent surgery with the diagnosis of ALCAPA with direct implantation of the left main coronary artery (LMCA) into the ascending aorta under standard cardiopulmonary bypass (CPB, Fig. 3). The PA defect, which occurred after the excision of the left coronary artery ostium from the pulmonary trunk, was reconstructed with an autologous pericardial patch. The LMCA was extensively mobilized and, with a large button, was reimplanted to the aortic root. The postoperative course was uneventful. Postoperative echocardiography showed improved LV function with good EF. ECG did not show any changes. Two months later, exercise thallium scan was done and there was no evidence of myocardial ischemia or mitral valve regurgitation.

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From the Cardiac Surgery Department, Imam Reza (A.S.) Hospital, Mashhad University of Medical Sciences, Mashhad, Iran.

Address for correspondence: M. H. Nezafati MD, Department of Cardiac Surgery, Imam Reza (A.S.) Hospital, Mashhad, Iran.

Email: m-h-nezafati@mums.ac.ir

Discussion

ALCAPA was first described in 1866. The first clinical description in conjunction with autopsy findings was reported by Bland and colleagues in 1933, hence the anomaly is also called Bland-White-Garland syndrome.⁴

During fetal life, ALCAPA does not cause hemodynamic problems because systemic and pulmonary pressures are equal and provide antegrade flow in the right and left coronary arteries. After birth, PA pressure gradually diminishes and flow reverses in the LMCA to one-third, so symptoms develop progressively.^{5,6} Today, although definitive diagnosis is often made by echocardiography, current practice requires cardiac catheterization and cineangiography.² Without surgical correction, about 80-90% of patients die from heart failure or sudden death during the first year of life.⁷ Insufficient collateral coronary flow and also a coronary steal from the left coronary artery into the PA trunk result in malperfusion of the left ventricular myocardium and ischemia. In the remaining patients, the RCA becomes large and tortuous and rapid development of collateral vessels between the right and left coronary system prevents ischemia. Therefore, these children survive to adolescence or adulthood.⁶ At the present time, prognosis for patients with ALCAPA is markedly improved due to early echocardiography and improvement in surgical techniques.

Our patient did not have symptoms until 8 years of age, and it was discovered during a routine physical examination. The symptoms were not discovered until this age because of a balance between sufficient collateral flow from the RCA to LCA and a minimum coronary steal syndrome.

The possibility of sudden death in ALCAPA patients is very high; therefore, surgical intervention is recommended.³ Whenever possible, a two-artery coronary system will be the choice technique.³ The other surgical options for treatment include ligation of the left main coronary ostium combined with

coronary bypass using saphenous vein graft, LIMA or radial artery and Takeuchi procedure.²



Fig 1. Preoperative angiography showing retrograde filling of the left main trunk and the main pulmonary artery.

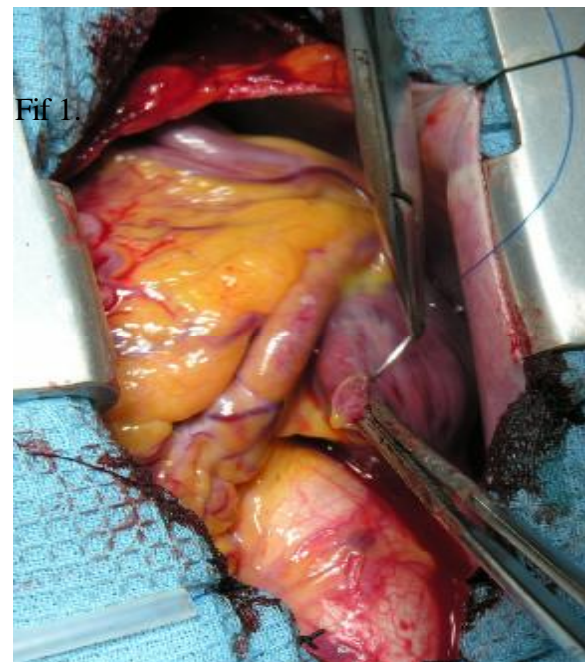


Fig. 2. Intraoperative view showing collateral network and tortuosity of RCA.



Fig. 3. Intraoperative view showing anastomosis of LMCA to ascending aorta.

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