



An Eligible Surgical Repair Technique for Bland-White-Garland Syndrome

Bland-White-Garland Sendromlu Olguda Seçilebilir Bir Cerrahi Teknik


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ABSTRACT

Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) is an uncommon but major congenital cardiac anomaly, which was firstly defined in 1866. Multiple surgical techniques intend to compose 2-coronary artery system, including direct reimplantation of left coronary artery to aorta and coronary artery bypass grafting procedures. Left coronary artery seldomly rises from non-facing pulmonary sinus, where direct reimplantation is technically quite difficult for surgical correction because of long distance from native aorta. In this case report, a surgical technique option of reconstructing left coronary artery, arising from non-facing sinus which we turned to aorta with a tunnel created from autologous pulmonary artery wall, was presented.

Keywords: ALCAPA; coronary artery anomaly; congenital heart disease.

ÖZ

Sol koroner arterin pulmoner arterden orijin alması (ALCAPA) nadir olmakla birlikte, ilk olarak 1866'da tarif edilen bir major konjenital kardiyak anomalidir. Sol koroner arterin direkt aortaya reimplantasyonu ve koroner arter baypas greftleme prosedürleri de dahil olmak üzere çok sayıda cerrahi teknik ikili koroner arteriyel sistem oluşturmayı amaçlamaktadır. Sol koroner arter nadir olarak non-facing pulmoner sinüsten köken alır ve bu nativ aortaya uzak mesafede bir bölge olduğu için, direkt reimplantasyonun teknik olarak oldukça zor olduğu cerrahi onarımdır. Bu vaka sunumunda, non-facing pulmoner sinus kökenli sol koroner arterin, otolog pulmoner arter duvarından tünel oluşturularak aortaya rekonstrüksiyonu cerrahi tekniği bir seçenek olarak sunulmuştur.

Anahtar kelimeler: ALCAPA; koroner arter anomalisi; konjenital kalp hastalığı.

INTRODUCTION

Anomalous origin of left coronary artery from pulmonary artery (ALCAPA), also known as Bland-White-Garland syndrome, is an uncommon congenital abnormality that affects 300000 live births per year and approximately 0.25-0.5% of all congenital heart defects (1). There are two varieties of ALCAPA: The infant and the adult type. They have different appearance and outcomes. If not interfered, approximately 90% of infant type patients die in the first year (2). Although, certain number of the patients with adult type do not have symptoms at early periods in life. Elder patients with ALCAPA generally have symptoms as mitral insufficiency, cardiomyopathy, dysrhythmias, and moreover death (3). As a result, presumptive identification of ALCAPA indicates prompt certain diagnosis and early surgical intervention regardless of the age except for the newborn.

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Received / Geliş Tarihi : 12.02.2020

Accepted / Kabul Tarihi : 25.05.2020

Available Online /

Çevrimiçi Yayın Tarihi : 25.08.2020

CASE REPORT

A 4 month-old and 5 kilograms weighing infant presented with paroxysmal tachycardia, pulmonary infection and mild dyspnea with feeding. Transthoracic echocardiography demonstrated a dilated left ventricle and significantly reduced ejection fraction with measurements under 20%. Angiography determined the way that ALCAPA arises from non-facing pulmonary sinus. Patient was identified as ALCAPA and taken to surgical procedure.

A median sternotomy incision was employed, autologous pericardium was harvested, and glutaraldehyde preserved autologous pericardium was prepared for reconstruction. Bicaval cannulation was used for venous drainage, and distal aortic cannulation was performed. Antegrade crystalloid cardioplegia (30 mL/kg body weight) was introduced under moderate hypothermia of 28°C. After horizontal incision of main pulmonary artery, left coronary artery (LCA) was identified in the non-facing sinus of valsalva.

Anomalous coronary artery's direct reimplantation into aorta was technically challenging due to its remote site. So, coronary artery reimplantation was performed by method of transecting pulmonary artery wall and releasing LCA by means of an extended incision to the coronary ostium in the non-facing pulmonary sinus. We diverted LCA and non-facing sinus with a tunnel created from autologous pulmonary artery wall (Figure 1). We think that this technique provides the most well agreed and adapted anastomosis with proper linear measure, appropriate angling, and the best course of the coronary artery. Pulmonary artery reconstruction was performed with autologous pericardium which was trimmed to the appropriate size. After de-airing and aortic unclamping, inotropic support was achieved with infusions of adrenalin by dosing of 0.05 mcg/kg/min and milrinone by 0.5 mcg/kg/min before weaning from cardiopulmonary bypass (CPB). The patient was transferred to the intensive care unit in an uneventful manner.

DISCUSSION

ALCAPA is a rare condition that requires early surgical repair. Even so, nearly 15% of patients reach adulthood (4). In adults, this can be a cause of sudden death. Sudden cardiac death occurs in most of the cases due to exercise, mostly in males and young athletes (5).

Surgical techniques necessitate qualifications for ALCAPA. Direct reimplantation of the anomalous coronary artery to aorta is the most commonly accepted repair modality that has provided the most successful results at long-term follow-up. Other techniques are subclavian and LCA anastomosis, Takeuchi repair, LCA with autologous pulmonary artery wall tubular extension and coronary artery bypass grafting with vein grafts or left internal thoracic artery (1).

In our patient, direct implantation of ALCAPA was not convenient, due to the non-facing sinus origin of coronary artery. We believe that, our technique of diversion of LCA and non-facing sinus with a tunnel created from autologous pulmonary artery wall, verified a new point of view. Also in this technique, we have preferred autologous pericardium for pulmonary arterial repair because of advantages: it provides sufficient integrity and strength, it

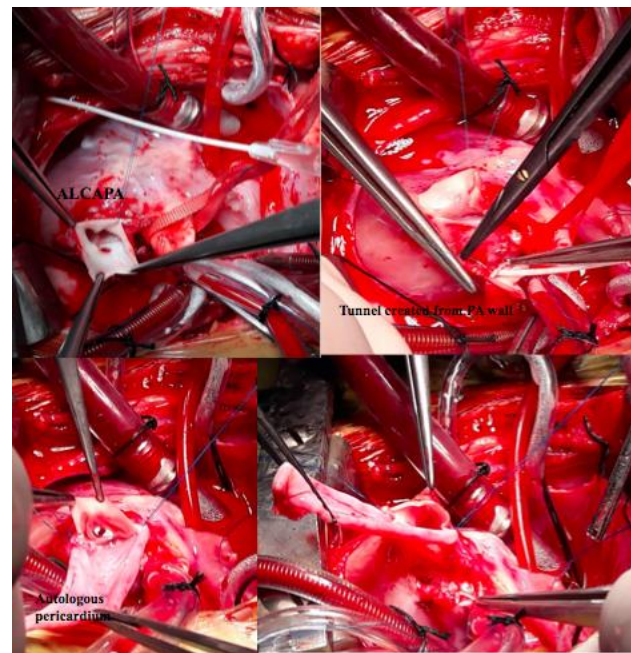


Figure 1. Coronary artery reimplantation was performed by method of transecting the pulmonary artery and releasing the left coronary artery through an extended incision to the coronary ostium in the non-facing pulmonary sinus and with a tunnel created from autologous pulmonary artery wall.

has a potency of growth and also remodeling, it is free of cost, it has more biocompatibility than bovine pericardium, the tissue is adequate for procedure and we do not need a separate operation for harvesting.

If compared with Takeuchi procedure which we can identify as an intrapulmonary baffle from LCA to ascending aorta, this procedure has best patency rates initially, but it has long-term complications, such as pulmonary artery and baffle obstruction, leak, and aortic valvar insufficiency. Reintervention was necessary in 30% of these patients to resolve the complications (6).

As conclusion, anomalous coronary artery that rises from non-facing sinus of valsalva of pulmonary artery or from a greater distance from the ascending aorta may necessitate different approaches. We describe our technique to be instructive for this challenging group of patients. However, further postoperative outcomes and follow-up parameters is necessary to evaluate whether this technique can contribute to the improvement of current surgical management of ALCAPA.

Informed consent: Written informed consent was obtained from the patient's parents.

Conflict of interest: All the authors declare that there is no conflict of interest.

Financial Disclosure: The authors declared that there is no financial support.

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