

Anomalous Left Coronary Artery from the Pulmonary Artery in an Adult

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ABSTRACT

Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital heart defect that usually presents before the age of 1 year. Several surgical options exist for the correction of ALCAPA; however, debate continues regarding the optimal repair technique in adult populations. We report the case of successful surgical repair of ALCAPA with a direct aortic implantation technique in a 44-year-old mother of 4 children.

INTRODUCTION

Anomalous left coronary artery (LCA) originating from the pulmonary artery (ALCAPA) is a rare congenital defect found in approximately 1 in 300,000 children. Untreated, only 15% of such children will survive into adult life [Wesselhoeft 1968]. In adults, ALCAPA may present as severe mitral regurgitation or sudden cardiac death [Wesselhoeft 1968]. Surgical correction of ALCAPA in adults can be accomplished via several techniques, ranging in complexity from simple ligation of the anomalous LCA to complex direct aortic implantation with one of the various forms of LCA extension [Dodge-Khatami 2002]. We describe a case of late presentation of ALCAPA accompanied by severe mitral valve insufficiency, for which direct aortic implantation of the LCA was successfully performed.

CASE REPORT

A 44-year-old mother of 4 children from Honduras was referred to our institution for evaluation of her mitral valve insufficiency. For several months before presentation, she had complained of severely limiting dyspnea on exertion, which carried a diagnosis of rheumatic mitral valve disease.

A transthoracic echocardiogram revealed a rheumatic-appearing mitral valve with severe mitral insufficiency, mild

mitral stenosis, and an ejection fraction of 45% to 50%. A cardiac catheterization evaluation revealed a dilated right coronary artery that supplied most of the left ventricle. The LCA arose from the main pulmonary artery immediately adjacent to the left sinus of Valsalva. Injection into the right coronary artery was followed by retrograde filling of the left anterior descending coronary artery (LAD) via significant collateral vessels, with drainage into the pulmonary artery. A cardiac computed tomographic angiography (CCTA) scan confirmed ALCAPA.

Given the symptoms, as well as the objective findings of ALCAPA and severe mitral insufficiency, the patient was referred to the operating room for surgical intervention. Surgery was performed via a median sternotomy. Prior to heparin administration and initiating cardiopulmonary bypass, the left internal mammary artery (LIMA) was harvested. An intraoperative transesophageal echocardiogram revealed that the LCA originated from the pulmonary artery and showed a reversal of flow in the LCA with a diastolic jet of coronary flow entering the pulmonary artery (Figure 1). Total cardiopulmonary bypass was established between the ascending aorta and both vena cavae. Myocardial protection was achieved with moderate systemic hypothermia (29°C) and retrograde cold blood cardioplegia, which was repeated every 20 minutes. Surgical inspection revealed that the LCA originated from the posterior pulmonary sinus, which was adjacent to the left sinus of Valsalva. The pulmonary artery was transected immediately distal to the ostium of the LCA. The LCA was excised as a coronary button, which was widely mobilized to allow a tension-free anastomosis. A punch aortotomy was performed in the ascending aorta. The LCA was then anastomosed in an end-to-side fashion to the ascending aorta with running 5-0 Prolene suture. The defect in the pulmonary artery was repaired with a piece of bovine pericardium. The pulmonary artery was anastomosed in an end-to-end fashion with running 5-0 Prolene suture. The mitral valve was exposed via a superior septal approach. The valve appeared rheumatic in nature and was deemed not amenable to repair. The valve was replaced with a mechanical prosthesis per the patient's preference, and the atrial septum and the right atriotomy were closed. Finally, the LIMA was grafted onto the LAD. The remainder of the operation proceeded without complication. The patient's postoperative course

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was uneventful, and she was discharged from the hospital 10 days postoperatively. A CCTA examination performed at her 5-week follow-up revealed widely patent LCA, LAD, left circumflex, and LIMA vessels (Figure 2).

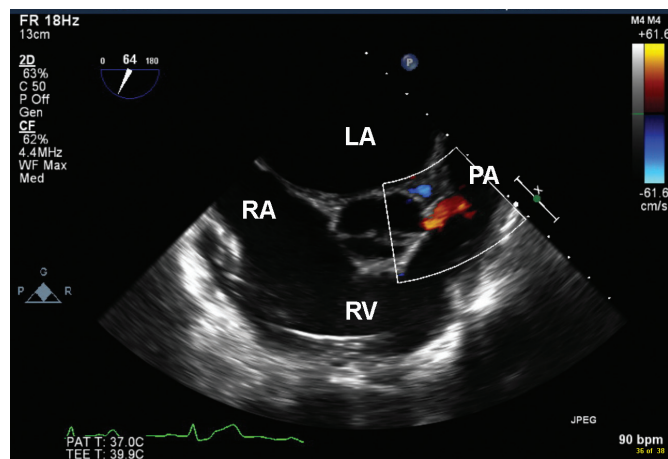


Figure 1. Intraoperative transesophageal echocardiogram demonstrating flow from the left coronary artery (red diastolic jet) entering the pulmonary artery adjacent to the aortic sinus of Valsalva. In addition, Doppler color flow demonstrates diastolic flow reversal (blue jet) in the left coronary artery, a hallmark finding of anomalous left coronary artery from the pulmonary artery. LA indicates left atrium; RA, right atrium; RV, right ventricle; PA, pulmonary artery.

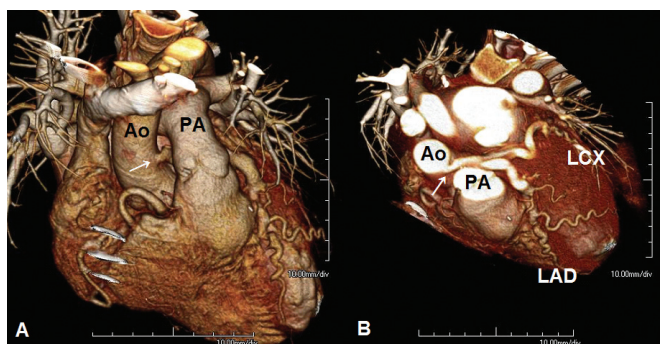


Figure 2. Postoperative cardiac computed tomographic angiography images showing a widely patent anastomosis (arrow, A and B) at the site of direct aortic implantation. Ao indicates aorta; PA, pulmonary artery; LAD, left anterior descending coronary artery; LCX, left circumflex coronary artery.

DISCUSSION

The literature has described several different techniques for the surgical management of ALCAPA to achieve a dual coronary system. The majority of the studies regarding ALCAPA repair have focused on pediatric patients. Pediatric

patients have been shown to have better long-term survival and better cardiac function with a dual-coronary system repair than with simple ligation of the anomalous LCA [Dodge-Khatami 2002]. Management of ALCAPA in adults is controversial, however.

The techniques for surgical correction for ALCAPA vary greatly and include surgical ligation at its pulmonary origin [Sabiston 1960], closure with a percutaneous device [Collins 2007], and a combined approach with LIMA-to-LAD grafting and closure with a transpulmonary pericardial patch [Murala 2006]. In contrast to these approaches, direct aortic implantation provides a more physiological repair and has shown good results to date, even in the adult population [Kottayil 2011]. Additionally, given that the 5-year patency rates for LIMA grafts are approximately 95%, a LIMA-to-LAD graft repair would therefore be expected to fail in 5% of patients, thus leaving these patients with no arterial supply to their left coronary system [Hayward 2007]. We have described a challenging case of ALCAPA in a 44-year-old woman, whose diagnosis was masked by the presence of rheumatic mitral valve disease. In our case, preoperative imaging studies revealed that the anomalous LCA originated from the posterior pulmonary sinus, adjacent to the left sinus of Valsalva. Therefore, we performed direct aortic implantation of the coronary button and achieved excellent results, despite the late age of presentation for this lesion.

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