

CASE REPORT

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Modified Cabrol technique for the treatment of adult anomalous left coronary artery from the pulmonary artery

James E. Davies¹, Gagandip Singh^{2*} and Panos N. Vardas¹

Abstract

Background: Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly that typically presents within the first year of life but can also sporadically present in adults. ALCAPA, if left untreated, can lead to coronary ischemia and sudden death.

Case presentation: We present the case of a 32-year-old male with ALCAPA with ischemic cardiomyopathy and the anomalous left main coronary originating from the non-facing sinus of the pulmonary artery (PA). A posterior reimplantation of the anomalous coronary artery was accomplished with a modified Cabrol technique using synthetic polytetrafluoroethylene (PTFE) graft.

Conclusions: A posterior re-implantation technique using a modified Cabrol graft, in cases where length of the native button is challenging, has excellent outcomes for this rather rare and challenging clinical entity.

Keywords: Adult-type ALCAPA, Cabrol, ALCAPA, Congenital heart defect

Background

Our patient is a 32-year-old male with a history of anomalous left main coronary artery from the pulmonary artery (ALCAPA) who presented with heart failure symptoms. Due to anatomic characteristics of the ALCAPA with the left main coronary artery (LMCA) originating from the non-facing sinus of the pulmonary artery (PA), a direct coronary reimplantation technique was not feasible. We present a unique case of surgical correction using a modified Cabrol technique using polytetrafluoroethylene (PTFE) graft instead of direct coronary reimplantation.

Case presentation

A 32-year-old male with a history of chronic systolic heart failure, New York Heart Association (NYHA) class III, presented with worsening shortness of breath and was found to have an ALCAPA. His past medical history was significant for ischemic cardiomyopathy and obesity. A perfusion nuclear study a few months prior to presentation showed a partially reversible distal anterior wall defect suggesting ischemia in the left anterior descending (LAD) territory. A computed tomographic angiography (CTA) indicated an anomalous origin of the LMCA from the PA with retrograde flow from the coronary artery into the PA, indicating coronary steal. The LMCA divided into normal-appearing left LAD and left circumflex (LCx) arteries. This was confirmed with a coronary angiogram (Figs. 1 and 2). Mild compensatory dilation of the right coronary artery along with left ventricular dilation with suspected subendocardial hypoperfusion in the LAD territory was also noted. Ultimately, the patient was diagnosed with an ALCAPA, and it was advised that he

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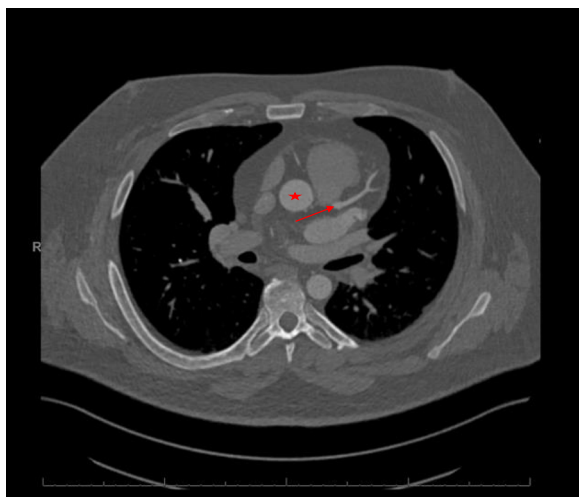


Fig. 1 Anomalous left coronary from the non-facing sinus of the pulmonary artery (red arrow), bifurcating into the left anterior descending and the circumflex artery. The ALCAPA is opacified at the same phase as the aorta, reflecting a retrograde flow into the pulmonary artery. The red star represents the ascending aorta

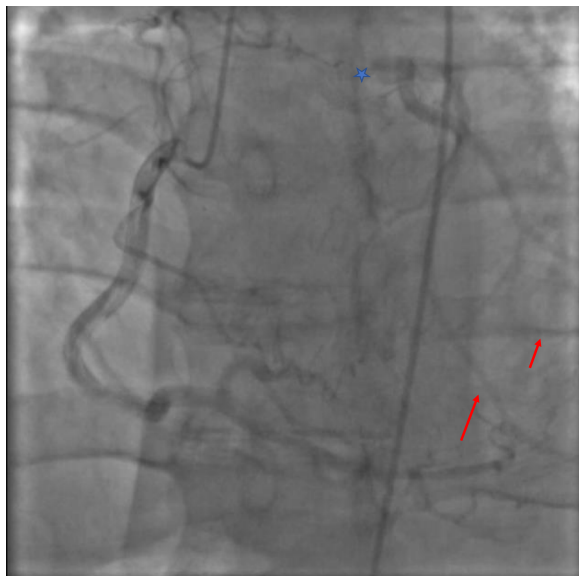


Fig. 2 Image from the preoperative angiogram of the ALCAPA. A right coronary injection fills with contrast into the left circumflex and left anterior descending artery through right-to-left collaterals (red arrows). The left main artery empties into the pulmonary artery (blue star)

undergo surgery. Preoperative transesophageal echocardiography (TEE) indicated a dilated left ventricle, with moderate to severe anterolateral hypokinesia, a depressed left ventricular ejection fraction (approximately 35%), and no significant valvular abnormalities.

The patient was therefore offered surgical correction of the ALCAPA.

After median sternotomy and establishment of cardiopulmonary bypass, the heart was arrested (for a total of 83 min) with cold blood cardioplegia. The PA was transected, and the anomalous LMCA was identified to come off of the non-facing sinus of the PA. The LMCA was harvested with a button technique and mobilized for about 25 mm distally. Despite the mobilization, the rare finding of the LMCA arising from the non-facing sinus precluded direct end-to-side anastomosis of the button to the aorta, since it was impossible to reach and prone to kinking. As such, the LMCA was anastomosed in an end-to-end fashion with a 10-mm Gelweave Terumo™ graft, which was brought posterior to the aorta and anastomosed end-to-side to the right side of the ascending aorta (modified Cabrol technique) [1]. The anastomosis was tested with direct cardioplegia administration into the graft and pressurization, which was deemed hemostatic. Finally, an autologous pericardial patch was harvested and used to reconstruct the PA. The patient was weaned from cardiopulmonary bypass after 95 min, and he had an uneventful recovery. Ultimately, the patient was discharged in good condition on the third postoperative day. One year after surgery, the patient was in good health, NYHA class I, with a patent Cabrol conduit (Fig. 3), without kinking or flow limiting external compression. On routine postoperative TTE, the ejection fraction appeared improved (approximately 45%) with improved anterolateral wall motion and no valvular abnormalities.

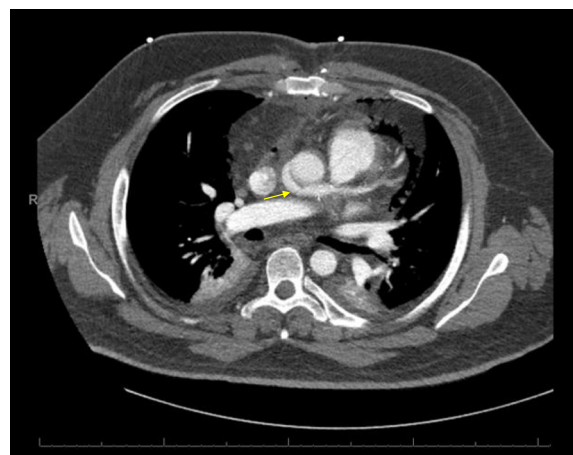


Fig. 3 The patent Cabrol bypass is shown (yellow arrow) with its course posterior to the aorta in a CTA 1 year later

Discussion

Anomalous left coronary artery from the pulmonary artery is a rare congenital defect in which the LMCA arises from the PA, rather than the aorta. There is an infant type and much more uncommon adult type of ALCAPA syndrome. Within the first months of life, as the PA diastolic pressure drops leading to a decreased perfusion pressure, infants with this condition typically manifest with left ventricular dysfunction and ischemia, with failure to thrive and irritability [2]. In addition, perfusion to the left coronary bed becomes retrograde from the right collaterals, causing a steal phenomenon that worsens myocardial ischemia [2]. Occasionally, patients may develop enough collateral vessels between the right coronary artery (RCA) and the LMCA, allowing them to present with the adult form of ALCAPA syndrome. The collateral vessels allow for sufficient perfusion for patients to survive into adulthood, although they can suffer from sequela of ischemic cardiomyopathy, as exhibited by our patient's presentation. Overall, adult ALCAPA patients can have variable clinical presentations, ranging from myocardial infarction to ischemic cardiomyopathy and congestive heart failure. Ultimately, adult ALCAPA patients occasionally may be asymptomatic, but they may eventually suffer a sudden cardiac death [3]. Thus, it is crucial to correct this defect once it is identified.

There are several surgical options for ALCAPA correction, all with the goal of achieving physiological coronary flow by establishing a dual coronary artery system. Simple ligation of the ALCAPA with an end result of a single coronary system has been associated with subendocardial ischemia and sudden cardiac death. The superiority of the dual coronary system in this clinical entity has been established by Backer et al. [4]. One surgical option is the Takeuchi procedure, which involves creating a transpulmonary baffle to connect the LMCA to the aorta [5]. This is more common in the infant population than the adult type correction. Also, adult ALCAPA ligation of the anomalous LMCA, followed by a coronary artery bypass graft (CABG) procedure has been described [6]. Although this is an easier technique, there is concern for the maturation of an arterial graft to the distally dilated vessel with major collaterals from the right and the rate of long-term patency of vein grafts in this relatively young patient population. The re-implantation technique involves the direct anastomosis of the LMCA to the aorta by transferring the LMCA with a button from the PA wall. In our case, the LMCA originated from the non-facing sinus of the PA. As such, the distance that had to be covered was rather long to reach, despite the meticulous dissection to free it up. There was also concern for tension on the anastomosis and possible kinking with respect to long-term patency. It has been proposed

by Turley et al. [7] to perform a posterior re-implantation technique with a lengthening procedure using a PA flap, but we felt that might result in stretching of the reconstructed tube posteriorly to the PA with risk of kinking, considering the distance to reach the aorta. The use of a PTFE graft implying a modified Cabrol technique with posterior reimplantation has not been previously reported. Traditionally, the Cabrol technique is used to repair ascending aortic aneurysms and dissections, when direct implantation of the coronaries to the graft is not feasible. The ostia of the coronary arteries are connected by a graft in an end-to-end fashion, and then the graft is anastomosed itself to the ascending aorta in a side-to-side fashion [1]. We prefer a PTFE graft versus a saphenous vein graft due to the less pliable wall of the synthetic material which also can contribute to protection from potential kinking behind the distended ascending aorta.

Conclusions

To our knowledge, a Cabrol bypass technique has not been previously described as a method for adult type ALCAPA correction. Based on our experience, the posterior re-implantation technique using a modified Cabrol graft, in cases where length of the native button is challenging, has a satisfactory outcome. However, a long-term follow-up to document patency is necessary.

Abbreviations

ALCAPA: Anomalous coronary artery from the pulmonary artery; CABG: Coronary artery bypass graft; CTA: Computed tomographic angiography; LAD: Left anterior descending; LCx: Left circumflex; LMCA: Left main coronary artery; NYHA: New York Heart Association; PA: Pulmonary artery; PTFE: Polytetrafluoroethylene; RCA: Right coronary artery; TEE: Transesophageal echocardiography.

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Authors' contributions

PV was the primary author of the manuscript and collected data. JD performed the case described. GS assisted in writing the manuscript. The authors read and approved the final manuscript.

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Availability of data and materials

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Consent for publication was obtained.

Competing interests

The authors declare that they have no competing interests.

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