

## ALCAPA - The Choices of Repair

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### Abstract

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital abnormality that affects 1 of every 300,000 live births [1] and accounts for 0.25% - 0.5% of all congenital heart defects. Extensive collateralization of coronary flow may allow for survival of patients to adulthood but they still require surgical correction due to risk of sudden cardiac death. The gold standard for the ALCAPA treatment is the direct re-implantation of the abnormal coronary artery directly into the ascending aorta. Nevertheless, direct translocation of the anomalous left coronary artery is extremely difficult and dangerous in adult patients.

The Takeuchi repair based on the reconstruction with an intra-pulmonary baffle is an appropriate alternative in the adult patient population.

We present an interesting case of a patient treated with an intra-pulmonary baffle and selected images showing the tunnel between the ALCAPA and the ascending aorta.

**Keywords:** ALCAPA; Takeuchi Repair; Intra-Pulmonary Baffle

### Introduction

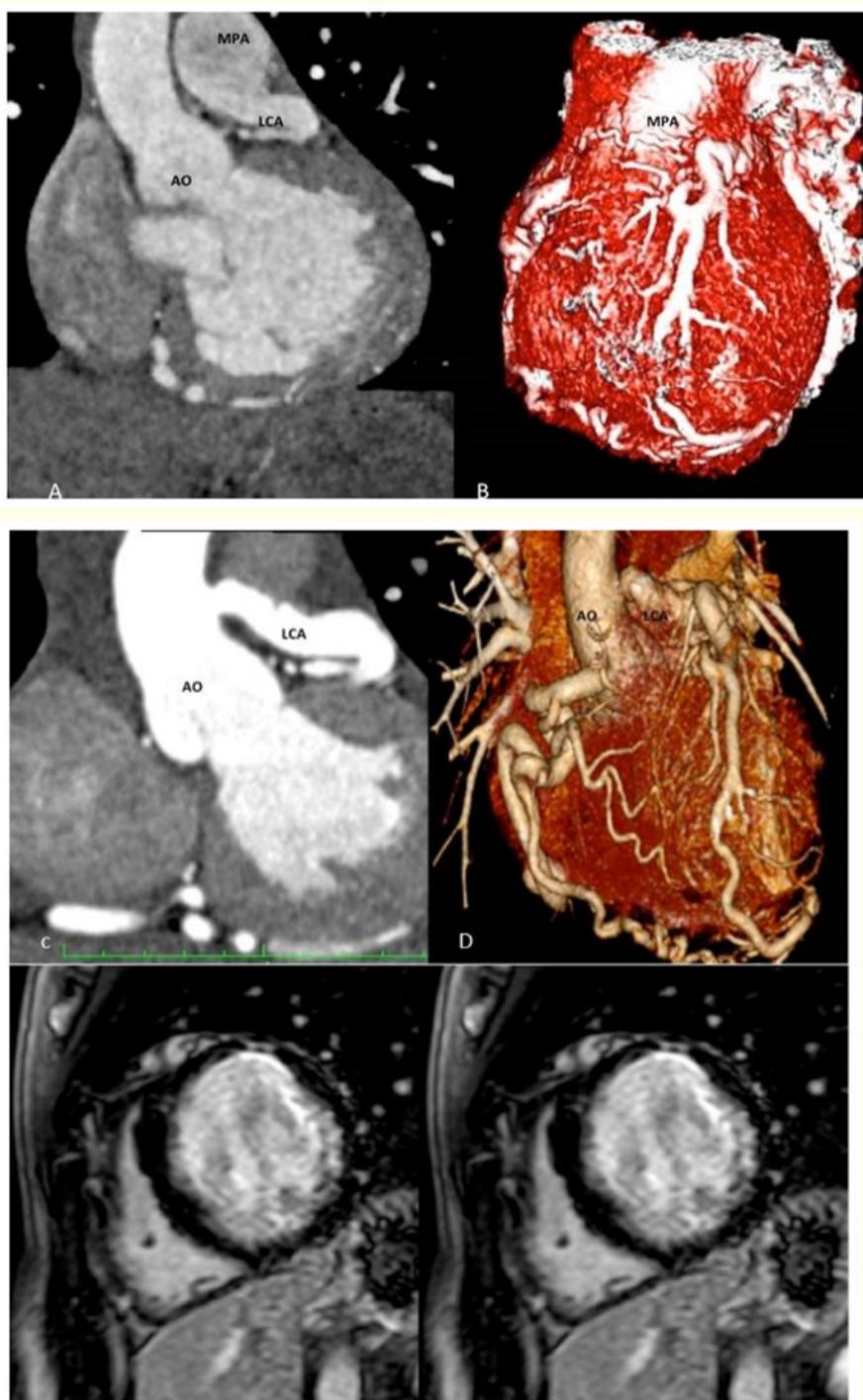
Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital abnormality that affects 1 of every 300,000 live births [1] and accounts for 0.25% - 0.5% of all congenital heart defects. Extensive collateralization of coronary flow may allow for survival of patients to adulthood but they still require surgical correction due to risk of sudden cardiac death. Surgical techniques to treat the ALCAPA syndrome include simple ligation of the coronary artery with coronary artery bypass grafting; end-to-end anastomosis of the left subclavian artery to the anomalous coronary artery [2,3]; creation of an intra-pulmonary baffle (Takeuchi repair) [1] and coronary artery re-implantation. Techniques that restore a two-artery anatomy are preferred although all patients require long-term follow-up due to potential development of complications [4].

### Case Report

We describe the case of a 27-year-old lady presenting with a history of progressive exertional dyspnoea and angina. A systolic murmur was heard at the left upper sternal edge on clinical examination. A 12-lead ECG showed anterior ischaemic changes. Transthoracic echocardiography showed hypokinesia of the anterior wall with echobright endocardium antero-laterally. A 256 multi-slice CT confirmed the diagnosis of ALCAPA with the left coronary artery arising from the posterolateral aspect of the main pulmonary artery (Figure A and B) with large collaterals and dilated RCA. Finally, a cardiac MRI with gadolinium enhancement showed a mildly dilated ventricle with moderately impaired left ventricular function (EF = 42%). A small segment of the anterior wall had late gadolinium enhancement consistent with subendocardial infarction (Figure E and F).

The case was discussed at our multidisciplinary team (MDT) meeting and the decision was made to offer surgery. Following median sternotomy and heparinization, cardiopulmonary bypass (CPB) was established through a cannula in the distal ascending aorta and a 2-stage venous cannula in the right atrium. Decompression of the pulmonary artery (PA) on establishing CPB may result in coronary 'steal' from the right coronary system via the collateral arteries causing enhanced myocardial ischaemia. With this in mind, the aorta was cross-clamped and cold blood cardioplegia infused into the right coronary artery through the aortic root. The main PA was opened longitudinally and the orifice of the left main stem was identified in the postero-lateral wall of the PA. The orifice of the anomalous artery was digitally occluded to prevent drainage of cardioplegia out of the orifice and achieve satisfactory myocardial protection. Active cooling was not performed.

A 5mm aorto-pulmonary window was created side-to-side along the adjacent walls of the two great arteries. Autologous pericardium was then sutured along the superior margin and along the sides of this window on the pulmonary aspect along the posterior endothelium of the PA up to and including the anomalous coronary ostium (Figure C and D). The cross-clamp was then released to allow visualization of the flow from the ascending aorta through the fistula and into the baffle that bulged into the pulmonary artery without any leak from the edges. The PA was then closed directly with prolene sutures and the patient weaned off CPB. Trans-oesophageal echo confirmed satisfactory repair with Doppler flow across the baffle into the coronary ostium. A coronary CT angiogram confirmed successful repair (Figures C and D).



**Figure 1:** CT Scan images - Image A LVOT view Showing the aorta and main pulmonary artery. LCA arises from the inferior surface of the pulmonary artery - confirming the diagnosis of ALCAPA. Image B - 3D reconstructed image showing LCA arising from the MPA. Image C showing the LCA has been connected to aorta via aortopulmonary window via baffle created in the pulmonary artery by using pericardium. Image D - 3D created image showing patent LCA. Image E - shows a SAX late gadolinium image prior to surgery, which has shown a small area of subendocardial scar in the anterior wall - as shown by the blue arrow. Image F - Post repair late gadolinium image SAX image that shows no change, reassuring a successful outcome from surgery.

## Discussion

85% with ALCAPA present in the first two months, in some cases symptoms may be absent and they may present in adulthood with angina, heart failure, arrhythmias or even mitral valve insufficiency.

Various surgical techniques have been described to repair an ALCAPA. One technique involves simply ligating the artery at its origin or occluding the ostium with a pericardial patch. This prevents perfusion of the myocardium with deoxygenated blood with its attendant consequences for myocardial function. Support for this technique comes from the presence of adequate collateral flow from the normally arising coronary artery.

The preferred technique in creating a two coronary system involves re-implantation of the anomalous origin into the lateral wall of the ascending aorta in instances where it arises from an adjacent position and kinking of the proximal artery can be avoided. However translocation of the coronary artery may be challenging when an insufficient length of vessel cannot be mobilized due to its origin from the non-facing anterior pulmonary sinus or from the left lateral position on the PA.

In this case, we used autologous pericardium to create a baffle [5] while the Takeuchi repair was originally described with the use of the anterior wall of the PA to create the baffle with autologous pericardium to close the defect in the PA [1]. When performing the Takeuchi repair, it is important to ensure that the baffle is placed without distorting the anatomy of the pulmonary root since this can cause regurgitation; equally, if too large a baffle is fashioned there could be obstruction to pulmonary blood flow; this potential risk can be minimized by the use of a patch pulmonary aortoplasty to maintain the caliber of the pulmonary root if required [5]. Creation of a baffle is more challenging if the LCA arises from the anterior-facing sinus of the PA and in such cases, one could consider reconstruction of a channel with either a segment of the aortic wall [6] or a length of saphenous vein [7] or simply by direct translocation of the anomalous ostium to the aortic root.

We have discussed a case who presented in adulthood and underwent successful repair with an alternative technique, which may be appropriate for other patients [8].

## Conclusions

Intra-pulmonary baffle known as Takeuchi repair is an alternative and excellent solution for the treatment of ALCAPA in the adult congenital patient.

Adult patients with ALCAPA normally develop an important collateral coronary system to supply the left coronary artery. The right coronary artery is dilated and there is often a shunt between the right and the left coronary artery. The left coronary artery normally drain retrograde into the pulmonary artery stealing coronary blood flow. For this reason several surgeons prefer to close the left coronary artery at the origin of the pulmonary artery to prevent the coronary blood flow steal. In the majority of cases a direct translocation is dangerous and difficult because the left coronary artery is not anymore so elastic like children arteries and the way become too long to be translocated. Intra-pulmonary baffle described by Takeuchi in 1979 offers an appropriate antegrade coronary blood flow to the left coronary artery. It is a reproducible procedure and preserves the anatomy of the patient. We advocate the use of the Takeuchi procedure as a valid alternative for the treatment of patients with ALCAPA.

## Conflict of Interest

None.

## Funding Source

None.

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