

## Case Report

# Transcatheter closure of right coronary artery fistula to the right ventricle

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### Abstract

Coronary artery fistula (CAF) is an uncommon anomaly usually congenital but can be acquired. Although, most of the patients are asymptomatic, some may present with Congested Heart Failure (CHF), infective endocarditis, myocardial ischemia or rupture. In the past, surgical ligation was the only option in the management of CAF, but since 1983, transcatheter closure of CAF has been increasing as an alternative to surgery.

We report a 3 year old boy, presented in Queen Alia Heart institute, who underwent successful transcatheter closure of a large fistula communicating the distal part of the right coronary artery (RCA) to the right ventricle (RV). Our case differs from other CAF in that the fistula was communicating the RCA itself to the RV.

Keyword: coronary artery fistula, right ventricle, vascular plug.

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### Introduction

CAF is a direct communication between a coronary artery and a cardiac chamber, great vessels as well as vascular structure[1]. It is a rare disease of coronary arteries, which can cause a significant hemodynamic problems depending on the size, exit chamber and it's relationship to the native coronary artery. It occurs in around 0.002% of general population[2].

Around 50% of CAF patients are asymptomatic, picked up with an incidental heart murmur whereas others may present with acute myocardial ischemia, angina pectoris, and infective endocarditis [3].

In this case report, we present a percutaneous closure of right coronary artery fistula to the right ventricle in a 3 year old male patient.

## Case presentation

A 3 year old boy, born at full term, one of identical twins following an uneventful pregnancy and cesarean section delivery. He presented in neonatal period with attack of tachypnea and feeding difficulties with normal O<sub>2</sub> saturation. Cross sectional echocardiogram at the neonatal period showed RCAF to the RV. The patient was asymptomatic during the regular follow up until the age of 3 year; he started complaining of effort intolerance with profuse sweating post minimal activity. The echo showed the same CAF with mild dilatation of the right heart. The patient was started on furosemide and planned for cardiac catheter.

On clinical examination: patient looked well, not in failure, no visible impulse and with soft S1 and S2 and continuous murmur grade 3/6, at the 3<sup>rd</sup> intercostal space. ECG showed sinus tachycardia with no evidence of myocardial ischemia (Figure 1). Coronary arteries CT angiogram showed RCA ectasia and RCA fistula to the RV.

Under general anesthesia, cardiac catheterization was done using right femoral vein and artery. Firstly, hemodynamic data was collected, pressures was taken in left ventricle, aortic root and RV. The pulmonary to systemic shunt ratio was 2:1. Aortic root angio showed a patent non-dominant left coronary artery (LCA) with normal branches and the RCA was dilated, gave rise to RCA branches but continued as fistula which exited in the RV. The anatomy and dimension of coronary arteries and fistula were determined using selective coronary angiograms. Having created an arteriovenous circuit through the fistula balloon occlusion was applied firstly to precisely identify all distal coronary branches (figure 2) and, secondly to get accurate details and dimensions of the fistula in order to decide on a landing zone and the appropriate type and size of device for occlusion, preserving all the native coronary branches to the myocardium. Based on the anatomy of the fistulae we used the AMPLATZER vascular plug type II (AVP2-010) 10mm diameter and 7mm length, deployed by Gutken catheter 6 French. The device deployment was performed retrograde via the femoral artery placing the AVPII distally close to the exit point in the RV. An Angiogram was done post device deployment and no immediate complications were noted (figure 2C&D). The patient was extubated immediately post procedure and started on heparin continuous infusion and warfarin. No ECG changes (figure 1), and no leaking through the device have been noted during follow up. The

patient was discharged home after 3 days with INR: 1.8 on warfarin and aspirin.

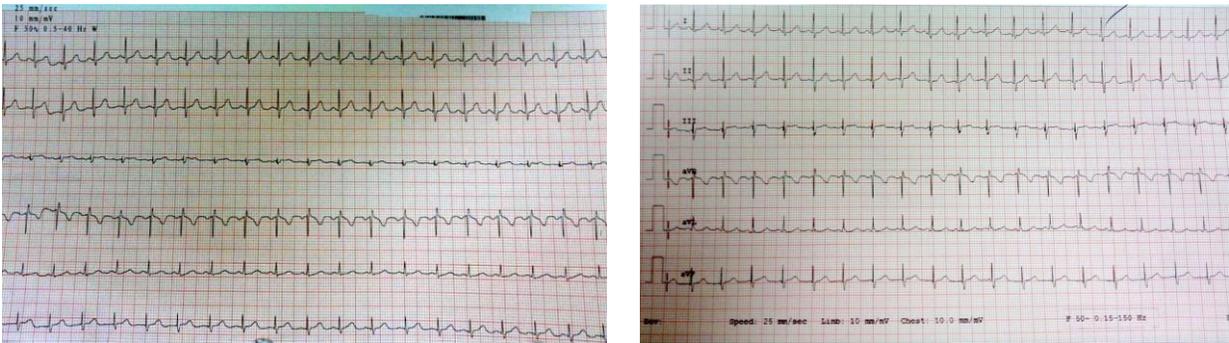


Figure 1: A: ECG before CAF closure

B: ECG after CAF closure, shows no changes in ST and T waves

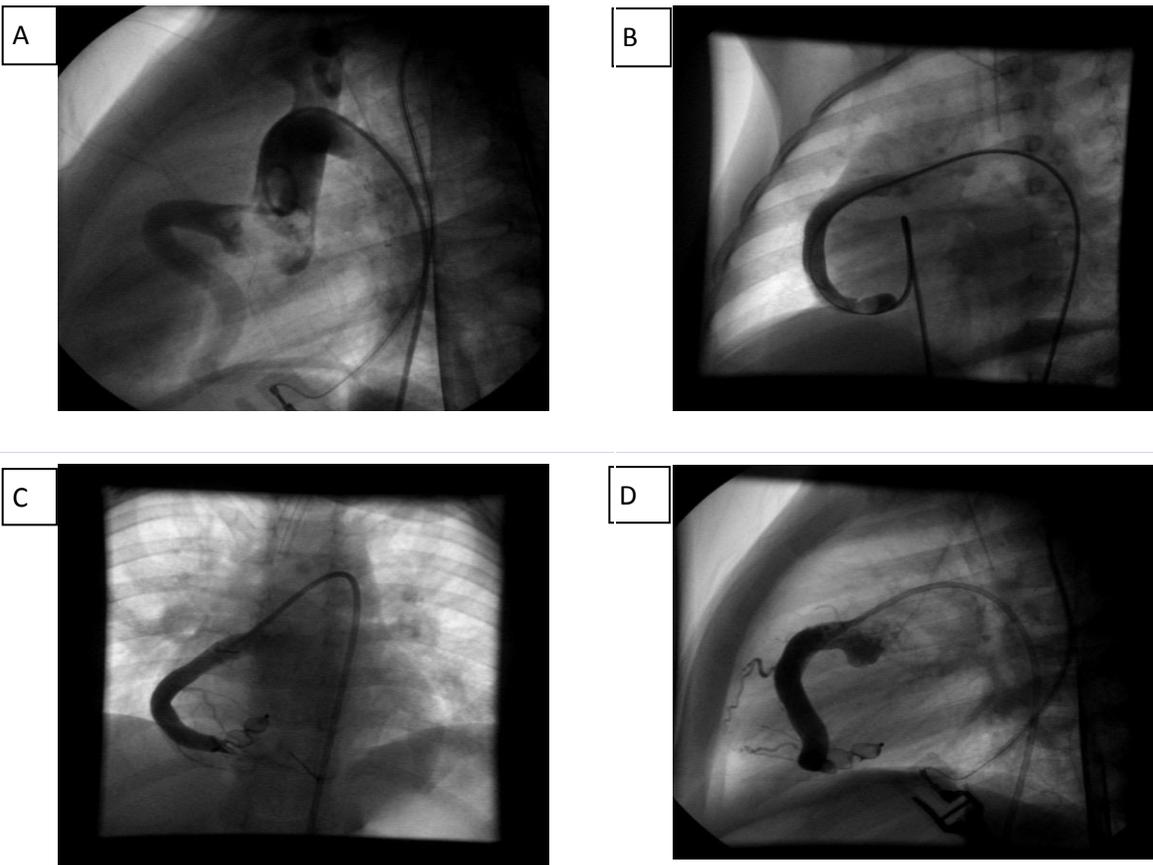


Figure 2: A: Aortic Root Angio with RCAF. B: Balloon occlusion test. C: Angio post device deployment A-P view. D: Angio post device deployment lateral view

## Discussion

CAF is a rare disease, accounting for 0.2-0.4% of congenital cardiac anomalies[4]. The major sites of origin is from RCA in 55% of patients while 35% from the LCA and less common from the circumflex artery. Very rarely these can be bilateral. The major receiving chamber is the RV in 45%, followed by the right atrium in 25%, pulmonary artery 15%, and less commonly the coronary sinus 7%. Complications related to the CAF are present in 11% of patients younger than 20 years and in 35% of patients older than 20 years. For this reason, elective closure of a significant CAF at a young age is recommended [5],[6].

CAF are thought to be formed as result from persistence of sinusoidal connection between the lumens of primitive tubular heart that supply myocardial blood flow in the early embryonic period[1].

A small fistula does not cause any hemodynamics compromise, while large fistula can cause coronary artery steal phenomenon, which lead to ischemia; the latter can give rise to chest pain, arrhythmias or mitral valve insufficiency[5]. The pathophysiologic mechanism of CAF is reduction in myocardial blood flow, which is related to the diastolic pressure gradient and runoff from the coronary vascular to the low pressure receiving cavity. In large fistulas the intra coronary diastolic perfusion pressure becomes progressively diminished. As a compensation, the coronary artery enlarges leading to increase the risk of myocardial ischemia beyond the origin of fistula[4].

The management of CAF in children is still controversial, spontaneous closure has been reported in one patient secondary to thrombus [3], otherwise surgical closure was traditionally viewed as the main therapeutic method. Since 1983 transcatheter closure of CAF has widely used with a good results [2], [4], [5],[7] , [8], [9],[10].

Our case is different because the RCA was the fistula itself which gave the branches of the RCA. We follow the same technique using right femoral vein and artery, the anatomy and dimension of coronary arteries and fistula were determined using selective coronary angiograms. Balloon occlusion test within the fistula was applied mainly to precisely identify all distal coronary branches. Using the AMPLATZER vascular plug type 2, the fistula was successfully closed and the device deployment was performed retrograde via the femoral artery. Angiogram was done post device deployment, and no immediate complication has been reported.

Medical follow up with daily ECG showed no ST-T wave changes and no leaking was notice on follow up 2D echo.

## Conclusion

The transcatheter closure of coronary fistula in children is a safe alternative to surgical closure. We reported a successful transcatheter closure of fistula communicating the distal part of the right coronary artery to the right ventricle in a 3 year old boy with no complications ,and no scar as well as low cost in comparing to surgical closure.

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