Transcatheter closure of a rare coronary artery fistula using a modified mother–child technique

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ABSTRACT

Coronary artery fistulas (CAFs) are rare abnormal communications between a normal coronary artery and a cardiac chamber or great vessel, such as the pulmonary artery, bypassing the myocardial capillary network. We report the case of a 17-year-old male with a medical history of pulmonary valve stenosis, who presented with progressive dyspnea and fatigue. Transthoracic Doppler echocardiography showed multiple continuous flows both on the apical interventricular septum and entering the left atrium. A tortuous CAF arising from the left main coronary artery to the left atrium was revealed by coronary angiography. The lesion was successfully closed percutaneously using an off-label Amplatzer™ Duct Occluder II Additional Sizes with a backup support of a modified "mother–child" system. This case highlights the effort of both pediatric and adult cardiology teams to come up with new potential strategies and combined techniques to overcome the difficulties of managing complicated CAFs, such as the use of percutaneous coronary intervention techniques and the selection of the most adequate occlusion devices, even when used off-label.

Keywords: Cardiac catheterization, coronary artery fistula, coronary vessel anomalies, pediatric

INTRODUCTION

Coronary artery fistulas (CAFs) are rare vascular connections from a coronary artery to a cardiac chamber (coronary cameral) or major central vessel (coronary arteriovenous) without an intervening capillary bed.[1,2] They are frequently congenital and have been reported in 0.2%–0.6% of patients, but the true incidence is not known because many may be small and asymptomatic.[1,2] The majority of CAFs arise from the right coronary artery or the left anterior descending artery.[3] The right-sided cardiac chambers and the pulmonary artery are the most common sites of drainage.[3] Although several cardiac imaging modalities can be used, selective coronary angiography remains the gold standard for the diagnosis.[3] While it is recommended to close all symptomatic and large fistulas, the best approach for smaller and asymptomatic cases is still controversial.[1] Treatment can be performed either by surgery or by percutaneous techniques depending on the anatomical and technical issues.[3] Percutaneous closure is a feasible and safe procedure that can be performed with a variety of techniques and devices.[3,4]

CASE REPORT

A 17-year-old male presented with progressive dyspnea and fatigue (New York Heart Association Functional Class II). He had a medical history of critical pulmonary valve stenosis with two previous percutaneous balloon pulmonary valvuloplasties and a surgical intervention for subpulmonary stenosis and ostium secundum atrial septal defect closure. There was no history of genetic disorders such as Noonan syndrome. On examination,

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he had a harsh systolic ejection murmur and a mild diastolic murmur in the left upper sternal border. The electrocardiogram showed right axis deviation and complete right bundle brunch block. The exercise stress test showed a low-level exercise tolerance, but no signs of ischemia. Transthoracic echocardiography showed biventricular hypertrophy with more evident right ventricular hypertrophy and no left chamber dilation (left atrium, 34 mm – *Z*-Score of +1.58 and left ventricle, 47 mm - Z-score of -0.43). There was also evidence of multiple continuous flows on the left atrium and the apical interventricular septum into the right ventricle. Aortography revealed the presence of multiple CAFs. Taking into account the patient was symptomatic, it was decided to attempt CAF percutaneous closure. After acquiring informed consent, the procedure was performed by a team of pediatric and adult cardiologists.

A medium-sized fistula with acute takeoff angle was visualized by selective coronary angiography, arising from the left main coronary artery to the left atrium. It had a tortuous course, with a stenotic origin (3.1 mm), a fusiform poststenotic dilation (6.7 mm), and a maximum length of 11.8 mm [Figure 1]. To overcome the difficult approach to the CAF, we used a modified "mother–child technique," to provide support for safe device advancement and deployment. Unfractionated heparin 100 UI/Kg was administered before the procedure, and the activated clotting time was monitored every 30 min thereafter. We accessed the right femoral artery to place a 7Fr Judkins Left 3.5 guiding catheter (Medtronic Inc., Minneapolis, MN, USA) - "mother" catheter - in the left coronary ostium. A 5Fr Judkins Right 3.5 guiding catheter (Terumo Corporation, Tokyo, Japan) - "child" catheter - was passed distally through the "mother" guiding catheter to get into the fistula. The 7Fr Judkins Left guiding catheter was 90 cm long, whereas the 5Fr Judkins Right inner guiding catheter was 100 cm. The CAF was then selectively catheterized with a 0.014″ BMW guidewire (Abbott Laboratories, IL, USA). The guidewire distal extremity was passed to the distal end of the fistula into the left atrium and then it was placed in the right upper pulmonary vein. The left femoral artery access was used to selectively catheterize the left anterior descending artery and the circumflex coronary artery with two 0.014″ BMW guidewires (Abbott Laboratories, IL, USA), in order to protect both coronary arteries [Figure 2].

We tried to advance the "child" guiding catheter along the fistula with no success. As such, a 1.8Fr microcatheter (Terumo Corporation, Tokyo, Japan) was used to support the "child" guiding catheter over the guidewire; however, this was also unsuccessful. At last, a coronary angioplasty balloon 2 mm × 15 mm (Cordis Co., USA) was used to taper the "child" guiding catheter distal extremity, by moving it away from the stenotic
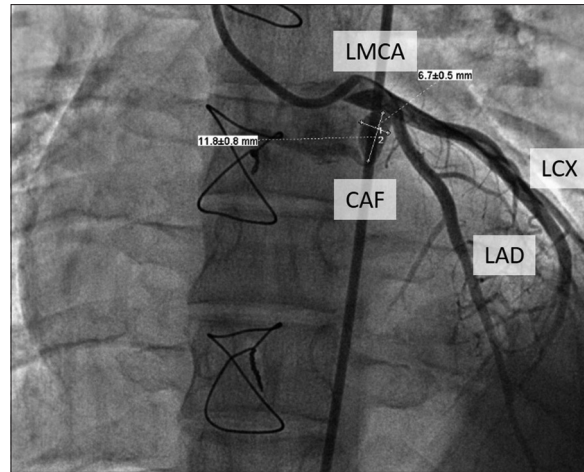

Figure 1: Coronary angiogram showing the coronary artery fistula originating from the left main coronary artery. LAD: Left anterior descending artery, LCX: Left circumflex artery
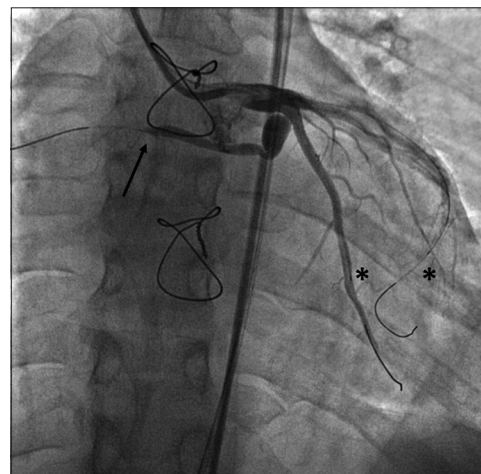

Figure 2: Coronary angiogram showing a guidewire in the coronary artery fistula (arrow) and two guidewires protecting both LAD and LCX (*). LAD: Left anterior descending artery, LCX: Left circumflex artery
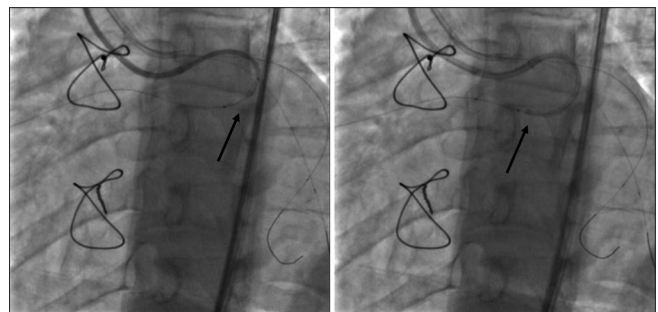

Figure 3: Triple coaxial catheter system (modified "mother–child technique") advancing along the coronary artery fistula (arrow)

border of fistula entrance, and finally advancing it over the guidewire into the fistula [Figure 3].

According to the CAF anatomy, the Amplatzer™ Duct Occluder II Additional Sizes (ADO II

AS) (9-PDA2AS-05-06-L) (St. Jude Medical, MN, USA) was implanted and successfully occluded the fistula with no impaired flow to normal coronary arteries [Figure 4]. The patient was started on antiplatelet therapy for 6 months. On follow-up, he reported complete resolution of his previous complaints of fatigue and exertional dyspnea and has been asymptomatic in a sustained form since the procedure.

DISCUSSION

The CAFs are anomalies of myocardial/coronary artery interaction, and the exact cellular and molecular mechanisms of CAFs' embryogenesis are not yet clear.[5] They are usually congenital, but postoperative acquired forms can occur, although less frequently.[3] In this case, the fistula could be iatrogenic, but we cannot rule out the possibility of a congenital defect, even more with the medical history of critical pulmonary valve stenosis. Congenital CAFs may coexist with other congenital heart diseases in 20%–45%. Pulmonary atresia with intact ventricular septum seems to have the highest incidence of CAFs, but other defects such as tetralogy of Fallot, patent ductus arteriosus, and atrial septal defect are quite common as well.[2,6]

The importance of this case relates to the rarity of proximal CAF arising from the left main coronary artery (LMCA) and the drainage site to a left-sided chamber.[7] Less than 10% of fistulae drain into the left heart structures, with the left atrium accounting for 5%–6% of cases. On the other hand, only 0.7% of fistulas arise from the LMCA.[6]

The large CAFs are usually symptomatic and should be closed to prevent complications such as myocardial ischemia and heart failure.[3] Transcatheter closure, although technically challenging, is an effective and a safe procedure for anatomically favorable CAFs.[3,8] There are several devices such as coils, detachable balloons, Amplatzer™ devices, covered stents, and chemicals that can be used to occlude the CAFs.[9] The antegrade or retrograde delivery approach is selected according to the operator's experience and the selected device. The retrograde method is the most used, although the antegrade approach has the advantage of avoiding femoral arterial access, despite the risk of device embolization.[9]

The presence of both adult and pediatric cardiologists working alongside in the catheterization laboratory was essential for the success of this case. "Mother–child technique" is a widely used procedure by adult cardiologists for stent delivery on percutaneous coronary intervention, as well as the guidewire protection of the other coronary arteries.[10] On the other hand, the experience and know-how of pediatric cardiologists were vital to assure a successful delivery of an appropriately chosen device to occlude the CAF.

After assessing the CAF anatomy in detail, we tried to choose the device that best fitted its characteristics (tortuosity and size) from those available in our catheterization laboratory. Several options were considered, and our choice for the ADO II AS was based on its size and length combination, malleability, softness, and ease of push through a difficult catheter course, allowing a secure placement of a single device with a high rate of complete occlusion.

The ADO II devices are ideal for vascular occlusion due to the availability of a wide range of diameters, short length, flexibility, and low profile delivery sheaths.[8] Although designed for duct occlusion, there have been reports of off-label use in other vessels, namely CAFs.[11] To the best of our knowledge, there is only one published manuscript describing the use of the ADO II AS for CAF closure in two children.[12] The present case reports a tortuous fistula successfully closed using ADO II AS, thus confirming its effectiveness, particularly when coronary side branches are adjacent to the fistulous orifice.[12]

Similarly, the modified "mother–child technique," with a coronary angioplasty balloon to taper the "child" catheter distal end and help its navigation into a tortuous vessel, proved to be an important strategy for the success of the procedure. To the best of our knowledge, the classic "mother–child technique" has been reported in a single previous publication in CAF closure, but with coil embolization.[13] We are the first to report the use of this "modified" technique to advance and deliver a duct occluder device in order to close a congenital CAF. Our case highlights the importance of a combined team of pediatric and adult interventionists to address complex congenital coronary malformations.
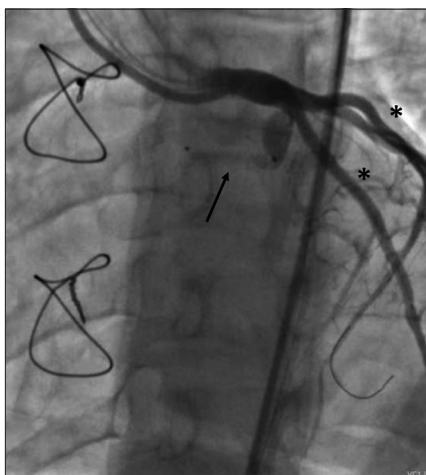

Figure 4: Coronary angiogram after Amplatzer™ Duct Occluder II Additional Sizes placement. Coronary artery fistula successfully occluded (arrow) with no impaired flow to LAD and LCX (*). LAD: Left anterior descending artery, LCX: Left circumflex artery

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Jama A, Barsoum M, Bjarnason H, Holmes DR Jr., Rihal CS. Percutaneous closure of congenital coronary artery fistulae: Results and angiographic follow-up. JACC Cardiovasc Interv 2011;4:814-21.

2. Latson LA. Coronary artery fistulas: How to manage them. Catheter Cardiovasc Interv 2007;70:110-6.

3. Gowda RM, Vasavada BC, Khan IA. Coronary artery fistulas: Clinical and therapeutic considerations. Int J Cardiol 2006;107:7-10.

4. Patted SV, Halkati PC, Dixit M, Patil R. A case of symptomatic double coronary artery-pulmonary artery fistulae. Ann Pediatr Cardiol 2010;3:171-3.

5. Pérez-Pomares JM, de la Pompa JL, Franco D, Henderson D, Ho SY, Houyel L, *et al.* Congenital coronary artery anomalies: A bridge from embryology to anatomy and pathophysiology – A position statement of the development, anatomy, and pathology ESC working group. Cardiovasc Res 2016;109:204-16.

6. Mangukia CV. Coronary artery fistula. Ann Thorac Surg 2012;93:2084-92.

7. Bagul PK, Phadke MS, Kerkar PG. Percutaneous closure of left main coronary artery fistula using AMPLATZER vascular plug II and AMPLATZER duct occluder I. JACC Cardiovasc Interv 2016;9:e67-70.

8. Bruckheimer E, Harris M, Kornowski R, Dagan T, Birk E. Transcatheter closure of large congenital coronary-cameral fistulae with amplatzer devices. Catheter Cardiovasc Interv 2010;75:850-4.

9. Oto MA, Yorgun H, Aytemir K. Percutaneous approaches to closure of coronary artery fistulas. Interv. Cardiol 2011;3:79-89.

10. Takeshita S, Takagi A, Saito S. Backup support of the mother-child technique: Technical considerations for the size of the mother guiding catheter. Catheter Cardiovasc Interv 2012;80:292-7.

11. Vijayalakshmi IB, Narasimhan C, Agarwal A. Transcatheter closure of left coronary cameral fistula with amplatzer duct occluder II. J Invasive Cardiol 2013;25:265-7.

12. Marini D, Ferraro G, Agnoletti G. Off-label use of the amplatzer ductal occluder II additional size for percutaneous closure of congenital and acquired coronary fistulae. Catheter Cardiovasc Interv 2016;87:E261-7.

13. Justo RN, Slaughter RE, Whight CM, Radford DJ. Transcatheter embolisation of coronary artery fistulae. Heart Lung Circ 2001;10:53-7.