

Transcatheter Closure of Congenital Left Main Coronary Artery Fistula to Right Atrium

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Abstract

Coronary cameral fistula is a rare congenital anomaly with an incidence of 0.002% in the general population. Most of the coronary artery fistula arising from right coronary artery, only a small percentage of case has been reported with coronary fistula arising from left coronary artery. It can be either asymptomatic or symptomatic. But prompt diagnosis and treatment is needed to prevent fetal complications.

Even though the gold standard management of congenital coronary artery fistula is the surgical closure with the development of interventional cardiac catheterization has been considered an alternative to surgery.

However, only a few cases of successful catheter intervention for congenital CAFs have been reported. Therefore, we present a rare form of coronary cameral fistula of LMCA to right atrium which is managed successfully with transcatheter intervention.

Keywords: Coronary Cameral Fistula; Congenital Anomaly; Coronary Artery Fistula; LMCA

Introduction

Coronary cameral fistula can present as a rare congenital anomaly with an incidence of 0.002% in the general population [1]. Cameral fistula is characterized by an abnormal communication between coronary artery and a cardiac chamber. It can be either congenital or acquired. Acquired form can be following chest trauma, infective endocarditis or iatrogenic following repeated coronary angiography, endomyocardial biopsy. Left main coronary artery fistula (LMCA fistula) is even more rarer than right coronary artery fistula (RCA fistula). Patients can be symptomatic or asymptomatic which need prompt diagnosis and treatment of this condition since this is associated with serious complications and poor prognosis. We share a rare form of coronary cameral fistula of LMCA to right atrium (RA) which was managed with transcatheter intervention.

Case Presentation

A fourteen year old patient second born to non-consanguineous healthy parents from an average socioeconomic background was referred to cardiology unit at Lady Ridgeway Hospital for Children with a diagnosis of Left main coronary artery to right atrium fistula (Figure 1).

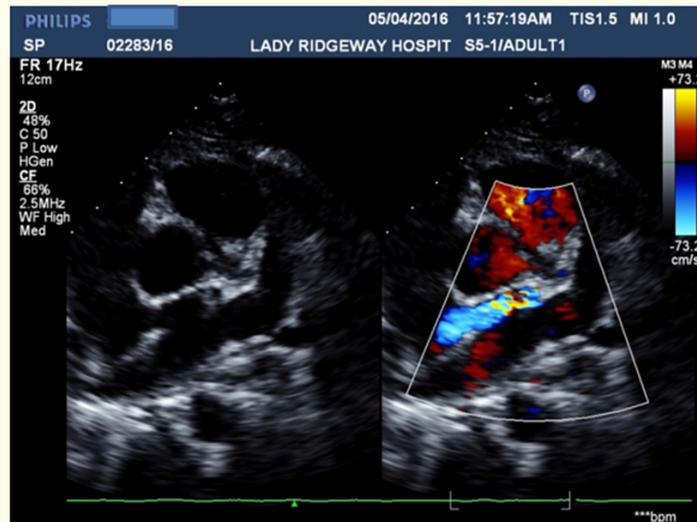


Figure 1: Arrow-fistulous tract.

He was apparently well and was incidentally diagnosed during a febrile illness at the age of 6 years and remained asymptomatic throughout.

Cardiac catheterization done in 2016, at the age of 10 years, found to have LMCA to right atrium (RA) fistula with a partially restrictive opening of fistula into RA (Figure 2). Selective angiograms of LMCA showed left anterior descending (LAD) and circumflex (LCX) arteries arising proximal to the origin of fistulous tract (Figure 3). It was then planned to close the fistula closer to its origin just distal to the origin of left LAD and circumflex artery using an Amplatzer Vascular Plug 1V device (AVP 1V).

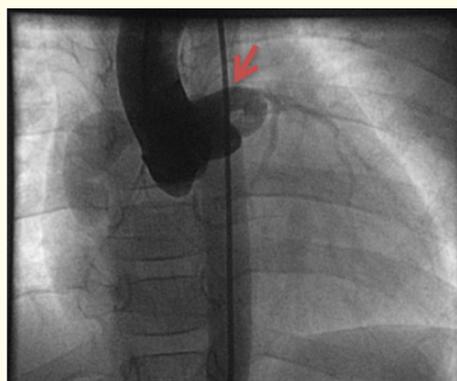


Figure 2: Arrow-aortic end of fistula.

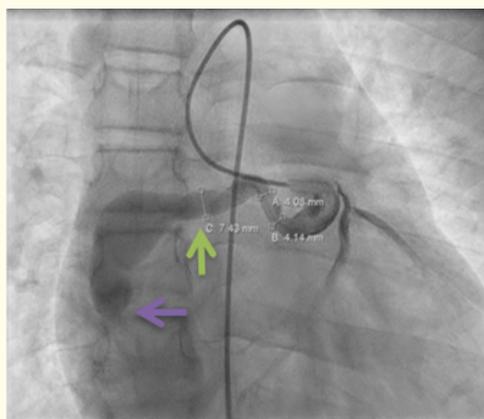


Figure 3: Green arrow - Fistulous tract. Purple arrow - RA end of fistula.

Initial attempt for device occlusion of fistula was made in 2019. Fistula was engaged from aortic end with a 4F JL catheter and a 0.032 Terumo angle tip guide wire which was advanced across the fistula in to RA and then to PA. The terumo wire was then snared in PA using a 10 mm Gooze neck snare and an arterio-venous loop was created. Multiple attempts taken to negotiate the fistulous tract using 5F ADO 11 delivery system and 5F guiding catheter both antegradely and retrogradely failed due to repeated kinking of sheath. As the delivery sheath could not be positioned well within the fistulous tract for optimal deployment of the device it was decided to re attempt the procedure at a later date using a smaller size AVP 11 vascular plug (not available in the lab at the time) which could be passed through a smaller 4F delivery sheath which is more likely to negotiate the tortuous fistulous tract.

The next attempt was made in 2020 where a similar arterio-venous loop was created as before. A 4F ADOII delivery sheath was then advanced over wire from venous end and an AVP II 8mm device (Figure 5) was deployed in fistula just proximal to origin of LMCA. Prior to release of the device check angiograms at aortic end of fistula showed unobstructed flow in LMCA with occlusion of fistulous tract (Figure 4). Also no ECG changes suggestive of ischemia were noted.

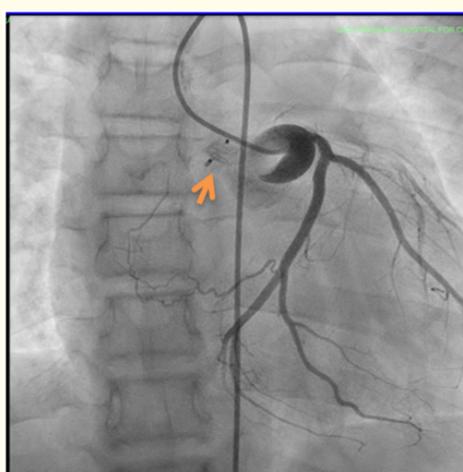


Figure 4: Arrow – AVP II device.

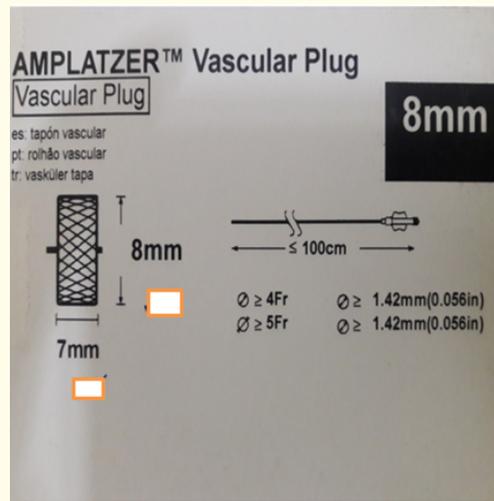


Figure 5

Post procedure echocardiogram showed occlusion of fistulous tract with unobstructed flow in LMCA and a normal ECG. He was prescribed a daily dose of Clopidogrel for 3 months and lifelong anti platelet dose of Aspirin.

He is being followed up at cardiology clinic without any complications.

Discussion

Coronary cameral fistula is an abnormal communication between coronary artery and a cardiac chamber. It is a rare entity. It can be either congenital or acquired but majority of cases are congenital. Most of the patients are asymptomatic.

The failure of obliteration of embryonic vascular sinusoids in the myocardium is the most possible explanation for the pathogenesis for the congenital form of fistula [2]. The coronary artery fistula (CAF) arising from right coronary artery accounts for 55% of cases while 35% arise from left coronary artery and 5% involve both coronary arteries [3]. More than 90% of the fistulas drain into the venous circulation which includes pulmonary artery, superior vena cava, right atrium, and right ventricle. Fistula drainage into RV accounts for 41%, RA 26%, pulmonary artery 17%, LV 3% and SVC 1% [4]. Even though most fistulas are single communications there can be multiple communications as well.

Only a smaller proportion of pediatric patients are asymptomatic but majority of adult patients are usually asymptomatic [5]. The steal phenomena where the blood in CAF bypasses the myocardial capillary network results in a wide range of symptoms ranging from mild shortness of breath to myocardial ischemia, congestive heart failure, cardiac arrhythmia, rupture or dissection of fistula with or without cardiac tamponade and endocarditis [6].

Even though 90% of cases are congenital, there are acquired coronary fistulas due to infective endocarditis, takayasu arteritis, iatrogenic and trauma. The selective coronary angiography is capable of identifying the CAF, but it is difficult to clarify the relation of the distal site of the CAF to other cardiac chambers because of dilution of a contrast medium or overlapping of adjacent structure. Conventional and

CT angiography, Magnetic resonance (MR) angiography are emerging noninvasive imaging modalities which enable precise visualization of the anatomy [7].

It is very important that all symptomatic patients should be managed with a surgical option because the surgical risk appears to be considerably less in most of the cases than the potential development of serious and fatal complication.

The gold standard treatment for congenital CAF closure is the surgical closure but during past decade transcatheter intervention has been considered an alternative to surgery due to less post-operative hospital stay, avoidance of sternotomy and cardiopulmonary bypass [8]. However, only a few cases of successful catheter intervention for congenital CAFs have been reported and very few data available for long-term prognosis. Therefore, we reported our experience with transcatheter closure of congenital CAFs in a pediatric population for better understanding of prognosis and complications encountered in this population.

Conclusion

Coronary cameral fistula is an abnormal communication between coronary artery and a cardiac chamber which is a rare entity. Only a smaller proportion of pediatric patients are asymptomatic but majority of adult patients are usually asymptomatic. The gold standard treatment for congenital CAF closure is the surgical closure but transcatheter intervention has been considered an alternative to surgery due to less post-operative complications. Only a few cases of successful catheter intervention for congenital CAFs have been reported, therefore we reported our experience with transcatheter closure of congenital CAFs in a pediatric population.

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