

82. Percutaneous Closure Through Coiling Embolization of Coronary Artery Fistula Presenting as Acute Coronary Syndrome

Richardson Lorilla, Vhina Therese Sison, Jessore Isidro, Manila Doctors Hospital, Manila, Philippines

Body

Background: Coronary artery fistulas are rare coronary anomalies accounting for 0.4% and are seen in only 0.002% of the general population. They are usually discovered incidentally via patients undergoing coronary angiography ranging between 0.1 to 0.2%.

Case: A 51-year-old Filipino woman with newly diagnosed hypertension stage 2 presented with 3 months history of intermittent chest heaviness accompanied by occasional dizziness and episodes of exertional dyspnea. Persistence of symptoms sought consults and was subsequently admitted. On admission patient was hemodynamically stable with unremarkable physical examination. An electrocardiogram was requested which revealed Sinus rhythm with non-specific ST-T wave changes and an elevated HS-Troponin I. Transthoracic echocardiography revealed a preserved ejection fraction of 61%, concentric left ventricular hypertrophy with normal contractility and adequate systolic function, mild tricuspid regurgitation and normal pulmonary artery pressure with mild pulmonic regurgitation. She was managed as a case of Non ST elevation myocardial infarction and was subsequently transferred to our institution for further investigation due to persistence of symptoms despite maximal medical management.

She underwent left heart cardiac catheterization which revealed normal coronary arteries with an incidental finding of fistula was noted upon selective cannulation of the conus artery showed an anomalous vessel originating at right conus artery just behind right coronary artery ostium and draining into the right atrium with noted visible contrast blush appearing after dye injection consistent with a coronary artery fistula. (Figure 1) Medical management was continued and she was discharged stable. In the interim, she continued to experience intermittent chest heaviness with episodes of exertional dyspnea hence was offered percutaneous intervention. Our patient is symptomatic on admission and diagnosis of MI was made and coronary steal phenomenon was considered the cause. The main indications for intervention are clinical symptoms, especially of heart failure and myocardial ischemia due to coronary steal phenomenon.

Patient underwent percutaneous closure of coronary artery fistula through coiling embolization, where a target 3 x 10 mm coil was delivered through the microcatheter and implanted at the proximal segment of the conus artery with noted successful coil embolization of the coronary artery fistula. (Figure 2) Post procedure patient was markedly improved and was discharged well. At 3 weeks follow up, the patient did not report recurrence of symptoms.

Discussion: Coronary artery fistulas (CAF) are rare cardiac abnormalities that account for 0.3% of congenital heart diseases. (4) Different origins and drainage sites account for several morphologies that explain the highly variable presentations. Coronary artery fistulas represent the most hemodynamically significant congenital defect of the coronary arteries. (3) Clinical manifestations are determined mainly by factors such as the site and size of the shunt, the pressure gradients between the origin and drainage site, and the presence of underlying valvular or coronary pathologies. (6–8)

The drainage site, more than the origin of coronary artery fistulas, largely contributes to symptoms. Over 90% of the fistulas drain to low pressure structures, such as right-sided chambers (right ventricle 41%, right atrium 26%), pulmonary artery (17%), left ventricle (15%), left atrium (9%) and superior vena cava and coronary sinus (7%). (4,9)

Fistulas that drain to the right side of heart, with left to right shunting, can result in volume overload to the right heart chambers or to the pulmonary vascular bed which can primarily lead to pulmonary hypertension or high cardiac output failure. (3,1) On the other hand, when CAFs drains into the left cardiac chambers or the pulmonary vein, there is a risk of left heart volume overload increases, leading to hypertrophy or dilation of cardiac chambers as well. (1) Although majority of affected individuals remain asymptomatic, coronary fistulas may grow with age causing significant symptoms such as dyspnea, angina, and if unaddressed, may create large shunts leading to development of complications such as heart failure, endocarditis, arrhythmias, stroke, myocardial ischemia or myocardial infarction. Dyspnea and chest pain represented a frequent (71%) clinical symptom in CAFs in adults. (10) Pathophysiology of larger fistulas can lead to a coronary steal phenomenon, which is the reduction of myocardial blood flow distal to the site of the fistula, bypassing the myocardium, precipitating myocardial ischemia.(11) At this point, patients develop chronic dyspnea and angina and congestive heart failure symptoms, aggravated by exercise or other activities that increase the oxygen demand. (12) (1) These pressure differences between the coronary artery and the chamber into which the fistula drains are also responsible for echocardiographic appearances of dilation of different cardiac chambers. (13) CT coronary angiogram, the non-invasive modality of choice, has the advantage of higher temporal and spatial resolution with multiplanar reconstruction and 3D volume-rendered images, hence supply detailed anatomical information on the origin, course, and drainage site of CAFs. (4) Transthoracic and transesophageal echocardiography are particularly useful in the evaluation of the hemodynamic effects of the fistula on cardiac chambers. (4) Drainage site can be identified with color flow imaging, used to identify the origin and drainage site of coronary arterial fistulas and assess their functional consequences. 2D echocardiography is useful in evaluating right and left ventricular size, mean pulmonary artery pressure in left to right shunt, and evaluation of regional or global left ventricular systolic function. (1)

The gold standard however for diagnosing coronary artery fistulas mainly is through coronary angiography. It provides finer details such as blood flow patterns, device landing zones, or surgical ligation sites which may not be fully evident on CT or MR images, thus allows for complete procedural planning. (4,14)

Controversies in management of Coronary artery fistulas remain yet to be explored. Management strategies for coronary artery fistulas have not been full defined in both the 2018 American Heart Association and American College of Cardiology and the 2020 European Society of Cardiology guidelines for the management of adults with congenital heart disease, and were proposed mainly based of review of published reports. An overall premise, as with review of literature, is that when clinical symptoms do arise or hemodynamically significant shunts are present, patients are recommended to undergo closure of the fistula. The proposed indications for closures of coronary arterial fistulas include the following: presence of a large or increasing left-to-right shunt, left ventricular volume overload, myocardial ischemia, left ventricular dysfunction, congestive cardiac failure and for prevention of endocarditis/endarteritis.(1) (15) In a single center experience by Cebi, N. Et al (2008), 10 of the patients were discovered to have coronary artery fistulas via cardiac catheterization. The primary indications for

intervention include patient's age with persistent angina (CCS class II–III) and dyspnea (NYHA class II–III). (7)

On the other hand, intervention among asymptomatic adult patients with coronary artery fistulas is still a matter of debate. In the presence of a high flow shunting however, it can be justified, to evade development of debilitating symptoms, or complications. (7,16)

Two treatment strategies are offered for closure of coronary artery fistulas, surgical or via transcatheter. Indications for endocardial or epicardial surgical ligation are as follows: large fistula with high blood flow, tortuous and aneurysmal fistula, multiple communications and drainage sites, and need for a simultaneous distal bypass. (14) (3,13,16). Surgery is reserved for cases of multiple fistulae, those affecting large branches during embolization of coils, or when the fistulous connection is narrow or restrictive. (9)

Currently, percutaneous treatment is proposed as the first choice because it is less radical and entails a shorter period of hospitalization³; surgery is reserved for cases of multiple fistulae, those affecting large branches during embolization of coils, or when the fistulous connection is narrow, restrictive and draining into a cardiac chamber. Percutaneous procedures are preferred in patients whose anatomy is favorable, including those with a single narrow drainage site, a proximal fistula origin, an absence of multiple fistulas or large branch vessels, and/or an absence of concomitant cardiac disorders warranting surgical approach. (4). It is ideal for older patients who are at risk for perioperative complications. (10,17) Interestingly, prognosis from the transcatheter and surgical techniques in literature show that both these approaches are similar in effectiveness, morbidity, and mortality. (2,6,18) Recently, Transcatheter interventions are considered favorable as the treatment of choice and has been proposed as the primary option for coronary artery fistula closures. Deemed less radical, advantages over surgical techniques include, lower morbidity and shorter period of hospitalization. (2,9). Transcatheter closure of CAFs with microcoils and hydrocoils is feasible and safe in the anatomically suitable vessels. The technique is simple, with low risk of complications. Among fistula embolization products are polyvinyl alcohol foam, balloons, and coated stents but the most widely used devices are undoubtedly coils.

Evidence base for medical therapy are limited and none of which has proven to mitigate progression of coronary fistulas. Antiplatelet therapy has been recommended in patients with distal coronary artery fistulas and abnormally dilated coronary arteries while prophylaxis was offered as precautions against subacute bacterial endocarditis. (4,18)

Conclusion: Coronary artery fistula are rare anomaly which is important to recognize in symptomatic patients. In adult patients who are symptomatic, unaddressed fistula can lead to complications such as myocardial ischemia/infarction, heart failure, arrhythmias, endocarditis and pulmonary hypertension. Treatment approach to coronary artery fistulas are still a matter of debate, as in asymptomatic fistulas that may carry risk for development of complications. Closure of coronary fistulas should be offered when patients become symptomatic or when complications do arise. Our case demonstrated that in patient with coronary artery fistula can be close via percutaneous through coiling embolization given that such patient meets the criteria and indications.

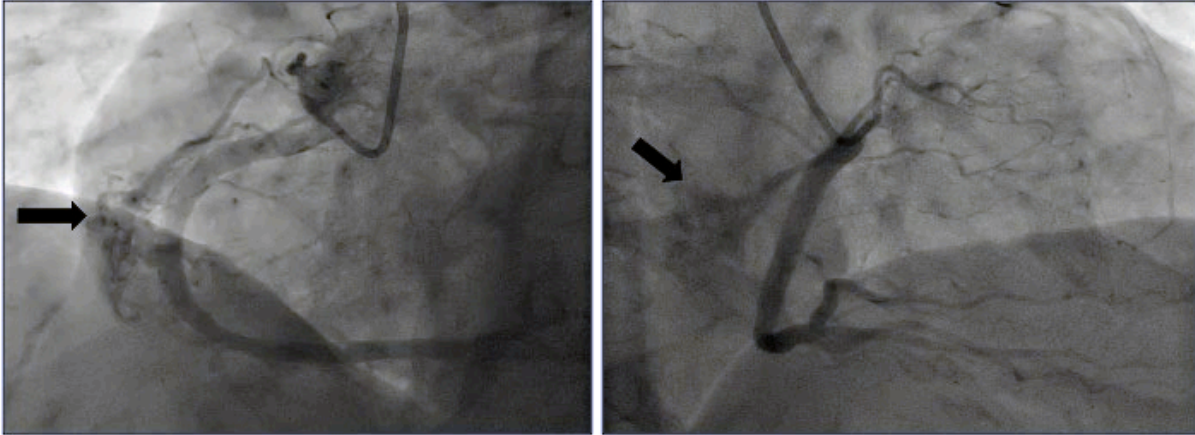


Figure 1. In RAO and LAO views showing an anomalous vessel originating at right conus artery, just behind right coronary artery ostium, and draining into the right atrium. (black arrow)

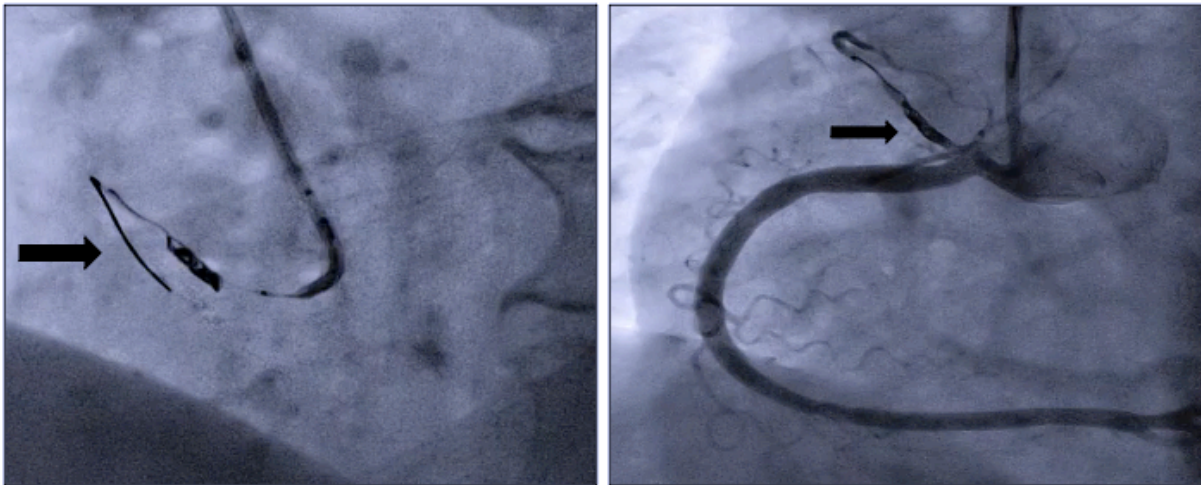


Figure 2. The embolization was performed with a Target helical coil with a diameter of 3mm and length of 10 mm was delivered and deployed into the fistula at its proximal portion through the micro catheter (Excelsior 150 x 6 cm) and a guide wire (Transend Ex 0.014 x 205 cm).

Figure 2.1 After coil embolization the fistula was sealed completely at the proximal segment by the coil. Final angiography revealed a substantial reduction in flow through the fistula with a small residual leak, and no obstruction of flow in the right coronary artery.