

Successful percutaneous Transcatheter closure of coronary artery fistulas (CAFs)

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Abstract

Background-Coronary artery fistulas (CAFs) are vascular anomalies which in rare cases can cause hemodynamic problems with indication for intervention. Congenital CAFs is a very rare malformation that may involve any or all coronary artery branches and any cardiac chamber. Elective closure of coronary artery fistulas by percutaneous transcatheter techniques is generally accepted in the presence of symptoms, but controversies exist in the management of asymptomatic patients.

Methods-We described two patients: A 49 years old female developed episodes of increasing exertional chest pain and dyspnea over 3 years, another 32 years old was referred for coronary angiograph because of dyspnea on exertion with-increasing its severity since one year age.

Results- Although non invasive imaging may be helpful, we demonstrated that cardiac catheterization and coronary angiography is necessary for the precise delineation of coronary anatomy and CAF

Conclusion-As mentioned above treatment is advocated for symptomatic patients and for those asymptomatic patients who are at risk for future complication. Form available data and our results Transcatheter closure (TCC) of coronary artery fistula (CAF) is an acceptable alternative to surgery in most patients(*Iranian Heart Journal 2011; 12 (2):59-65*).

Coronary artery fistula was first described in 1865 and is a relatively rare anomaly with a reported incidence between 0.01 to 0.2 percent.^{1,2}

Said and Landman found an incidence about 1 in 1000 diagnostic angiograms, only few cases were clinically apparent.^{3,4}

A coronary artery fistula (CAF) involves a sizable communication between a coronary artery, bypasses the myocardial capillary bed, and enters either a chamber of the heart or segment of the systemic to pulmonary circulation (coronary arterio venous fistula).⁵

A coronary artery connection to the pulmonary may also be considered under this grouping.⁶

CAFs originate from the right coronary artery (RCA) in 55% of cases, from the left coronary artery (LCA) in 35% of cases, and from both in 5% of cases.¹¹

These fistulas have a propensity to drain into low pressure sites, including the RV (42.5%), the RA (34%) and the pulmonary artery (15%).^{12, 14}

Only a minority drain in to left sided chambers, with 5% and 3.5% of lesions draining into the LA and LV respectively.^{12, 14}

The most common etiology of CAFs is congenital. These fistulas are thought to arise as a persistence of sinusoidal connections between the lumens of the primitive tubular heart that supply myocardial blood flow in the early embryologic period. A coronary fistula rarely may arise as a consequence of surgical

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is due to increased risk of complications. Including heart failure, myocardial ischemia, infective endocarditis, arrhythmias and rupture.⁹

Embryology

Coronary artery fistulae are thought to arise as a persistence of sinusoidal connections between the lumens of the primitive tubular heart that supply myocardial blood flow in the early embryologic period. Coronary fistulae occur in the absence of any outflow obstruction.

Another explanation may be faulty development of the distal branches of the coronary artery rectiform vascular network.

Discussion

Although CAFs represent only 0.2% to 0.47% of all congenital cardiac abnormalities, they are the most common congenital defects of coronary arteries.¹⁰

Coronary artery fistula may present in patients at any age but is usually suspected early in childhood when a murmur is detected in an asymptomatic child.

In a multicenter review, appreciably note problems related to operative risks and postoperative complications occurred after age 20 years.

The pathophysiologic mechanism of CAF is myocardial stealing or reduction on myocardial blood flow distal to the site of CAF connection if the fistula is large, the intracoronary diastolic perfusion progressively diminished.¹⁶

The coronary vessel attempts to compensate by progressive enlargement of the ostia and feeding artery, eventually, myocardium beyond the site of the fistulas origin is at risk for ischemia. With time, the coronary arteries leading to the fistulous tract progressively dilates which, in turn may progress to frank aneurysm formation, intimal ulceration, medial degeneration, intimal rupture, atherosclerotic deposition, calcification, side

branch obstruction. Mural thrombosis, and rarely, rupture.

The factors that determine the hemodynamic significance of the fistulous connection include the size of communication. The resistance of the recipient chamber and the potential for development of myocardial ischemia occasionally high output congestive heart failure has been described.

Clinical features

The majority of adult patients are usually asymptomatic. A small percentage of pediatric patients tend to be symptomatic.¹⁷

The clinical presentations include fatigue, dyspnea, orthopnea, angina, endocarditis, arrhythmias, stroke, myocardial ischemia or myocardial infarction.^{17,18}

Myocardial ischemia /infarction can occur from decreased coronary blood flow distal to the fistula and has been documented in patients with coronary fistulas with no evidence of coronary atherosclerosis. Rarely pericardial effusion and sudden death can be a presenting feature across all ages, 55% to 60% of congenital CAFs are asymptomatic.^{21,22}

Congestive heart failure (CHF) from large left to right shunts is the most common presentation in symptomatic neonates and small infants.^{23,24}

Both CHF and coronary steal angina can occur in adulthood with shunt augmentation from progressive enlargement of the fistula.

Diagnosis

Many fistula are small and found incidentally during coronary arteriography. Coronary angiography remains the gold standard for imaging the coronary arteries .but the relation of coronary artery fistulas to other structures, their origin and course may not be apparent with coronary angiography it is difficult to measure abnormal tortuous blood vessels in one section. The course of blood vessels

could be evaluated quite well by rearrangement serial cross sectional magnetic resonance angiography images using multiplanar reconstruction. multidetector row computed tomography (MDCT) cardiac imaging has provided excellent distal coronary artery and side branch imaging. Imaging of an entire 3 dimensional volume and the heart can be acquired within 20 seconds. With better temporal and spatial resolution than MR. several authors now advocate consideration of MDCT in imaging of coronary anomalies (as you can see such images in our cases).²⁵

Treatment

Most fistulas are small and do not need therapeutic intervention. however. surgical or catheter intervention is indicated .when the patient is symptomatic or when a shunt is particularly large, and thus associated with risk of complications (premature atherosclerosis due to shear induced intimal damage from the turbulent blood flow at the origin of the fistulas²⁶. Arrhythmias, rupture, endocarditis.

Surgical intervention for coronary fistula that performed in later adult life carries higher morbidity and mortality than in young adults.^{27,28}

Spontaneous closure secondary to spontaneous thrombosis although uncommon /has been reported^{29,30}

In asymptomatic patient with large fistulas the decision between interventional and conservative treatment has to be taken individually.

The main indications for closure are clinical symptoms especially of heart failure and myocardial ischemia and in asymptomatic patients with high flow shunting to prevent occurrence of symptoms or complications especially in pediatric population¹⁸

Catheter based closure of the fistulous connection is the nonsurgical treatment option for closure of coronary fistulas .with good

reported success. Since its introduction in early 1980s, transcatheter closure of coronary artery fistulas is now widely available and is being as an effective and safe treatment for CAFs.³¹

Catheter closure can be performed with a variety of techniques; including detachable balloons, stainless steel coils, controlled release coils, controlled release patent ductus arteriosus coils, patent ductus arteriosus plug, regular and covered stents and various chemicals.^{31,32,33,34,35,36,37}

Various other methods such as the use of vascular occluders or double umbrella devices has been reported^{38, 39}. they typically require large and non tortuous feeding vessels but can achieve effective closure.

Stent grafts such as expanded poly tetrafluoro ethylene (PTFE) covered stents may be used in more proximal lesions where both atherosclerosis and fistula coexist. The potential for occlusion of branch vessel is a disadvantage of the use of these devices.

Mavaroudis and coauthors recommended effective coil occlusion in patients who satisfy the following criteria; absence of multiple fistula, a single narrow drainage site, absence of large branch vessels. And safe accessibility to the coronary artery supplying the fistula.⁴⁰

The most serious complications of CAF is embolization stem from either proximal or distal coil migration. Proximal migration is usually due to device recoil during deposition and may cause thrombosis of the parent epicardial vessel / distal migration is typically due to under sizing of the coil in a high flow shunt. If this occurs the patient should be fully anticoagulated immediately and coil retrieval attempted.

The main concern is the risk of thromboembolic complications post procedure after reducing the high flow velocity in extremely dilated coronary arteries.⁴¹

Therefore in immediate postembolization period, the reduction in flow predisposes the

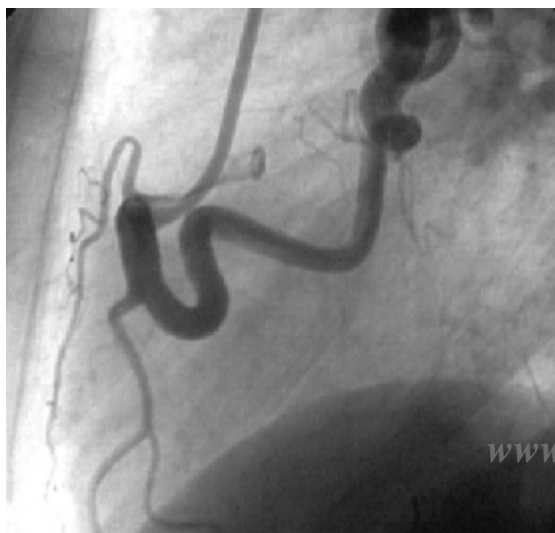
parent coronary artery to thrombosis, especially in the arteries with distal fistula. Conversely, occlusion of more proximal fistula may cause dilatation of distal circulation. In both situations, antiplatelet therapy seems prudent until normalization of the coronary arteries occurs. additional anticoagulation may become necessary if there is progressive dilation over time.

Case Report:

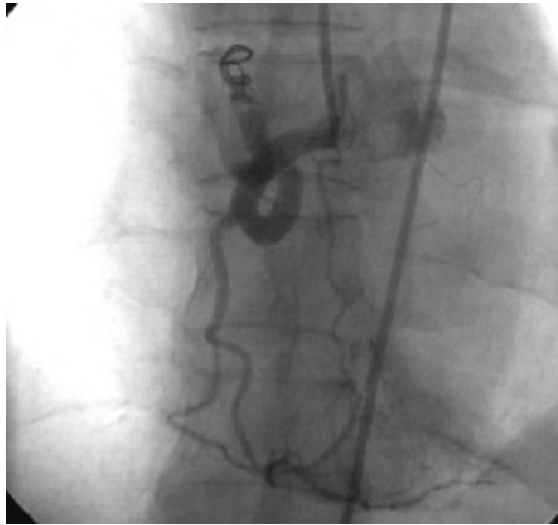
Case 1:

A49 year's old female developed episodes of increasing exertional chest pain and dyspnea over 3 years, there were no cardiac risk factors. P/E was unremarkable and no murmur was heard on auscultation .her ECG showed a sinus rhythm with a LBBB. Echocardiogram demonstrated globally reduced LV function with an EF of45%and moderate enlargement of LA (LA 40 mm) the mitral valve showed regurgitation grade2, no thrombus was seen. Radionuclide study showed reversible defect in inferior wall. Cardiac cath was performed to rule out coronary artery disease.

A selective coronary angiogram showed a right dominant coronary circulation. Two coronary artery fistula from RCA to pulmonary artery were found. The large fistula originating immediately after origin of the right coronary system and connected to the right pulmonary artery .procedure was done on this fistula. First by using guiding catheter JR4 that was placed in the proximal RCA and through it was passed with a micro



catheter and a 0.010 inch guide wire manipulated through the fistula at its most distal portion with use of two detachable trufill coil try to close the fistula. We have to use one Nit-occludcoil system (6 ×5mm) ecause of residual leak. After ten minutes selective coronary angiography indicated completely occlusion of the fistulae.



CASE 2:

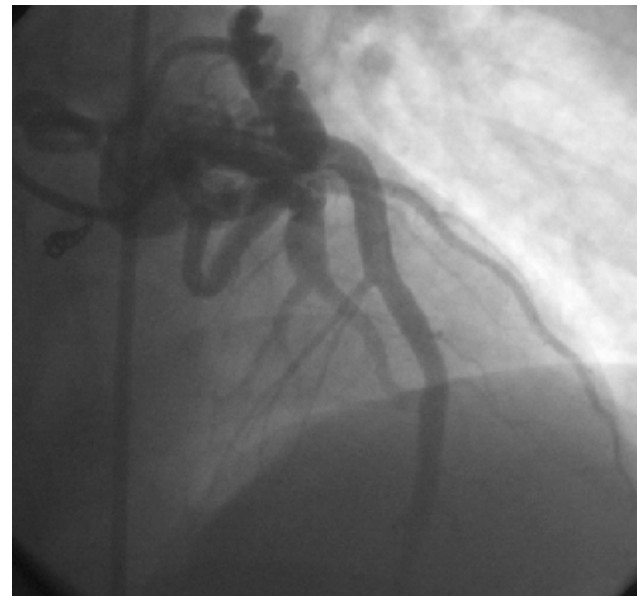
A 32 years old male was referred for coronary angiography because of dyspnea on exertion which had increased during one year. Cardiac risk factors were hypertension and hypercholesterolemia. There were no signs of

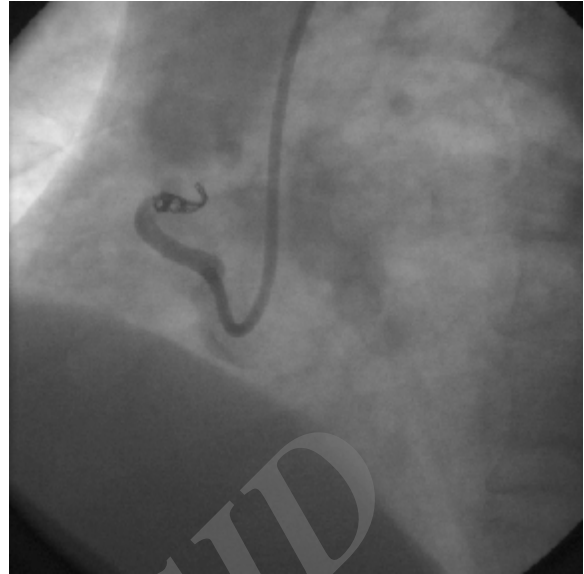
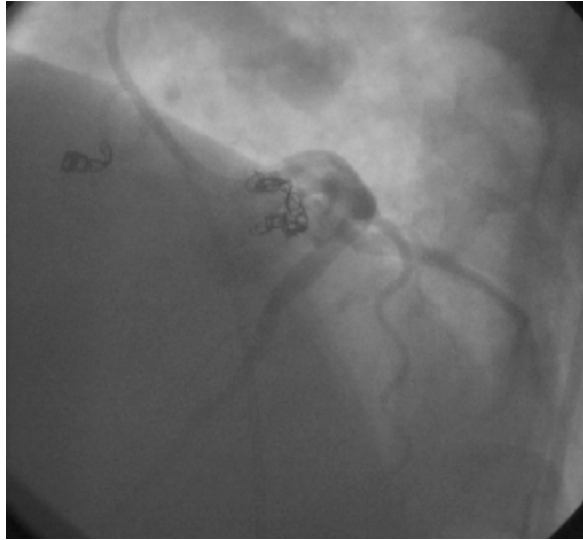
cardiopulmonary insufficiency .ECG was normal and TTE showed a hypertrophic septum (16mm). Selective coronary angiogram which was done in another center revealed a prominent fistula originating from two vessels out of proximal parts of LAD & RCA draining to the pulmonary artery. No significant stenosis was seen in coronary arteries .

Selective coronary angiogram and oximetry revealed existence of right and left coronary fistula with considerable shunting was proceed to close them with coil.

First of all with left guiding catheter judkins-4 and AL2 introduced into them and with using 0.01inch guide wire and micro catheter enter into the fistula and embolized 5-detachable trufill coil with 6-mm diameter and 15 mm length for each fistula.

Selective coronary angiogram was done 12 minutes after procedure indicated completely occlusion of the fistulas.





Conclusion

Management of CAF depends on the experience of each medical center, primary surgical treatment or transcatheter closure are valid options, the latter procedure may be performed with coil embolization, chemical substances or different devices depending on the size of the fistula. From available data and our results transcatheter closure (TCC) of coronary artery fistula (CAF) is an acceptable alternative to surgery in most patients.

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