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Short communication

## Treatment approaches to coronary artery fistulae: A single center trial

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## ABSTRACT

**Background:** A coronary artery fistula (CAF) is a sizable communication between a coronary artery, bypassing the myocardial capillary bed and entering either a chamber of the heart or any segment of the systemic or pulmonary circulation. It accounts for 0.27–0.40% of all cardiac defects.

**Patients and methods:** The study included 6 CAF patients hospitalized between January 2010 and June 2016. The mean patient age was 52.6. The sites of origin of the fistulae were from the left coronary artery in 4, from the right coronary artery in 1 and from right and left coronary arteries in 1 patient/patients. In all patients the site of termination was the pulmonary artery. The fistulae were closed surgically in 4 patients who also had accompanying cardiac pathology (3 patients with coronary artery disease and 1 patient with mitral stenosis) and in 2 patients without additional coronary pathology with coil embolization performed in the angiography unit. In the surgically treated group, with cardiopulmonary bypass the fistula opening was closed through pulmonary arteriotomy in 2 patients and fistula was ligated epicardially in 2 patients. Moreover, in the surgical group of 4 patients, coronary artery bypass grafting was performed on 3 patients with coronary artery disease, and mitral valve replacement on 1 patient with mitral stenosis.

**Results:** All patients had no pathological symptoms and findings in the follow-up controls one and three months after the surgery.

**Conclusion:** We suggest that CAF in patients with additional cardiac pathology should be treated surgically and others by performing coil embolization.

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## Introduction

A coronary artery fistula (CAF) may be described as a sizable communication between a coronary artery, bypassing the myocardial capillary bed and entering either a chamber of the heart or any segment of the systemic or pulmonary circulation. It accounts for 0.27–0.40% of all cardiac defects.<sup>1</sup> Coronary artery fistula was first described by Krause in 1865. In 1947, Bjork and Crafoord reported the first successful surgical intervention for CAF. The first successful transcatheter closure of CAF was reported in 1983.<sup>2</sup> Most babies and children are asymptomatic, but adults may show symptoms of fatigue, dyspnea, and chest pain which can also be associated with congestive heart failure occurring in young ages.<sup>3</sup>

## Patients and methods

The study included 6 CAF patients (3 males and 3 females) hospitalized between January 2010 and June 2016. The patients were diagnosed

by performing echocardiography (ECHO) and coronary angiography. The age range of the patients was between 42 and 57, with a mean of 52.6. The most frequent patient symptoms were fatigue (100%), effort angina (67%), and effort dyspnea (33%). In physical examination, continuous cardiac murmurs could be heard in 2 (33%) patients (Table 1). The radiographic examinations of the lungs showed cardiomegaly in 3 and increased pulmonary vasculature in 1 patient/patients. All patients had electrocardiographic (ECG) abnormalities which were left ventricular hypertrophy in 2, negative T wave in 3, and left axial deviation in 1 patient/patients. One patient had the results of a formerly made exercise test which showed inducible ischemia and negative T wave.

The definite diagnosis was reached by performing ECHO and coronary angiography. With ECHO, the origin and course of CAF could be determined in 2 patients as well as mitral stenosis in 1 patient (mitral valve area of 1.4–1.3 cm<sup>2</sup>) and 1–2° mitral valve regurgitation in 1 patient. Along with angiography, right and left heart catheterization procedure was performed on all patients. As additional pathologies, coronary artery disease requiring intervention was diagnosed in 3 patients and severe mitral stenosis in 1 patient. Angiographically, the origin and morphology of the fistulae were determined, and the coronary arteries were evaluated. Five patients (83%) had single fistulae; the sites of origin of the fistulae were from the left anterior descending (LAD) in

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**Table 1**  
Clinical characteristics of the patients.

	Age/sex	Symptoms	Murmur	ECHO	Origin	Surgery	Coil
1	42/M	Fatigue, effort angina, palpitation	Thrill +		LAD		+
2	49/F	Fatigue, effort dyspnea			LAD	+ ACBG	
3	57/M	Fatigue, palpitation, effort angina			LAD	+ ACBG	
4	52/M	Fatigue, effort angina			RCA	+ ACBG	
5	68/F	Fatigue, effort angina		1–2 MR	LAD		+
6	48/F	Fatigue, palpitation, effort dyspnea	Thrill +	MVA 1.4 cm <sup>2</sup>	RCA and LAD	+ MVR	

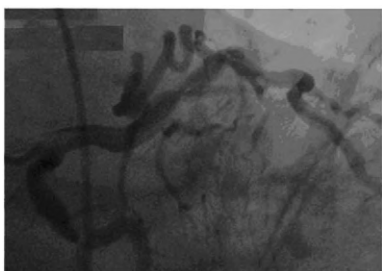
4 (67%) and from the right coronary artery (RCA) in 1 (17%) patient/patients. One patient had fistula originating from right and left coronary arteries (17%) (Figs. 1 and 2). In all patients the site of fistula termination was the pulmonary artery.

The appropriate modes of therapy were decided by the council formed by members from the Clinics of Cardiology and Cardiovascular Surgery, after analyzing hemodynamic and angiographic findings of the patients.

### Surgical intervention

The surgery was approached via a median sternotomy on all 4 patients. Following standard aorta-bicaval cannulation, the patients were put on cardiopulmonary bypass (CPB). In 2 patients, following application of moderate hypothermia and topical cold and placing a cross-clamp, cardiac arrest was achieved by antegrade cardiac cardioplegia. The arteriotomy of the pulmonary artery was performed with a longitudinal incision. The site of drainage of the fistula coming from the coronary artery was identified and closed with 6/0 prolene suture. Following, the pulmonary arteriotomy incision was appropriately closed and accompanying cardiac pathologies were corrected, thus ending the surgical intervention (Fig. 3). In 1 patient the fistula originated from RCA and in 1 patient from RCA and LAD. Since the coronary fistulae in these patients could not be epicardially determined, pulmonary arteriotomy was preferred (Figs. 4 and 5). External ligation of the fistula was performed on 2 patients. The fistulae of these 2 patients were of LAD-origin, and epicardial ECHO localized the artery feeding the fistula. In these 2 patients, CAF was ligated and cardiac pathologies were corrected before and after placing a cross-clamp, respectively.

Additionally, 3 patients underwent coronary artery bypass graft (CABG) and 1 patient underwent placement of valve prosthesis with MVR operation. The operations of all patients ended without problems. Following the appropriate closure of median sternotomy, all 4 patients were held under observation in the Intensive Care Unit until postoperative day 1.



**Fig. 1.** Fistula from left coronary artery.



**Fig. 2.** Fistula from right coronary artery.

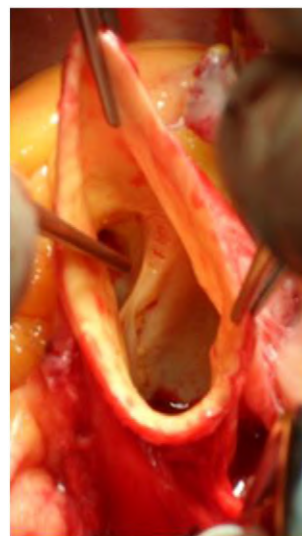
Coil embolization of the opening of coronary fistula under angiographic guidance was performed on 2 patients by the Cardiology Clinic.

### Results

All patients undergoing surgery were closely followed up. There was no case of morbidity or mortality. The period of hospitalization ranged between 5 and 7 days with an average of 5.5 days. In the ECHO controls 1 and 3 months after surgery no pathology was detected, and the preoperative symptoms were resolved in all of the patients.

### Conclusions

Coronary artery fistula is rare, but may present in patients at any age. Its incidence in angiographical series is 0.3–0.8%.<sup>4</sup> It may originate from any coronary artery including the left main trunk.<sup>5</sup> Major sites of origin of the fistulae are from RCA and LAD. CX is rarely affected.<sup>6</sup> In 55% of the cases, the site of origin is RCA or its branches. Fistulae originate from LAD in 35% and from both coronary arteries in 5% of the cases. In our study; fistulae originated from LAD in 4 (67%), from RCA in 1 (17%) and from both RCA and LAD in 1 (17%) of our patients. About 90% of the fistulae drain to venous circulation. Fistulae most commonly drain to low-pressure receiving cavities which are right cardiac chambers,



**Fig. 3.** Pulmonary arteriotomy and closure of the fistula.

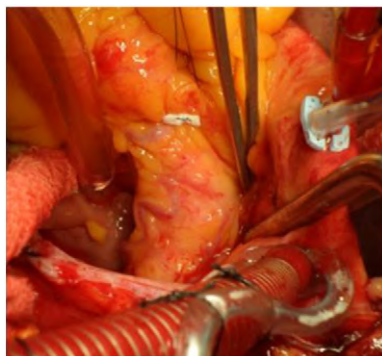


Fig. 4. External ligation to the proximal of the fistula.

pulmonary artery, superior vena cava, and coronary sinus.<sup>7</sup> Fistula draining to left cardiac chambers is very rare.<sup>8</sup> Fistulae drain to right ventricle in 41%, to right atrium in 26%, to pulmonary artery in 17%, to left ventricle in 3%, and to superior vena cava in 1% of the cases. There is only one single case in the literature draining to and causing hematoma in the pericardium.<sup>9</sup> Dilatation of the coronary artery may occur, but is not related to the size of the fistula.<sup>4</sup> Fistulous opening into a chamber or the drainage is mostly single or, rarely, multiple.<sup>10</sup> The probability of shunt formation is usually low if the drainage is to pulmonary artery. A left-to-right shunt exists in 90% of the cases. In all of patients, the coronary fistula drained to the pulmonary artery.

The clinical symptoms vary with the degree of left-to-right shunt.<sup>11</sup> Young patients are usually asymptomatic. The presenting symptoms of symptomatic cases may include fatigue, dyspnea, angina, arrhythmias, paralysis, and myocardial ischemia or infarction.<sup>11–12</sup> All of the patients in our study were symptomatic cases; there was the complaint of fatigue in all of the patients, effort angina in 4, palpitations in 2, and effort dyspnea in 2. Myocardial ischemia or infarction may occur in the absence of atherosclerotic coronary lesion, caused by decreased coronary flow at the distal of the fistula. Rarely, pericardial effusion or sudden death may also occur.<sup>13</sup> Continuous crescendo-decrescendo type systolodiastolic murmur is detected, and the murmur may have an unusual diastolic accentuation. Murmur was detected in 2 of our patients. In the presence of a major left-to-right shunt, pulmonary hypertension and congestive cardiac failure may develop as complications. Thrombosis of the ruptured fistula or fistula may be due to arterial aneurysm and coronary artery steal.

In the differential diagnosis, the disorders to be considered include persistent ductus arteriosus, pulmonary arteriovenous fistula, ruptured sinus of valsalva aneurysm, aortopulmonary window, prismatic ventricular septal defect with severe right coronary cusp prolapse, fistula originating from internal mammary artery draining into pulmonary artery, and systemic arteriovenous fistula.<sup>14</sup>

In ECHO, dilated coronary artery is frequently detected. Contrast-ECHO may give an idea about the site and size of the fistula.<sup>15</sup> Color Doppler may not reveal the flow at the distal of CAF. In such a case, transesophageal ECHO provides high quality images of the origin,



Fig. 5. External ligation of the fistula.

course, and draining site of CAF.<sup>4–16,17</sup> The coronary-pulmonary artery fistulae in our 2 patients were detected by ECHO.

Selective coronary angiography, Standard aortography, and angiography made by balloon closure of aortic root supply information about the course and anatomic features of the fistulae.<sup>18</sup> Contrast computed tomography (CT) or contrast magnetic resonance imaging (MRI) may be performed as alternatives to coronary angiography.<sup>19</sup>

Spontaneously thrombolized secondary spontaneous closure is rarely reported.<sup>10</sup> Antiplatelet treatment is recommended for patients with distal coronary artery fistula or with abnormally dilated coronary artery.<sup>12</sup> Also prophylactic treatment for subacute bacterial endocarditis should be made.

The main indications for fistula closure are cardiac insufficiency, myocardial ischemia, and high-volume flow rate of fistula in asymptomatic patients.<sup>16</sup> Surgical and direct epicardial or endocardial ligation are the primary methods used in the management of CAF.<sup>11</sup> Surgical intervention is safe and effective, providing good results.<sup>11–20,21</sup> The management of young and asymptomatic patients with no significant shunt is still disputable. Some authors claim that small, asymptomatic CAF may be followed up for long term, requiring no surgery.<sup>22</sup> On the other hand, some authors claim that appropriate therapeutic approaches should be made as soon as CAF is diagnosed. According to the latter authors, the early closure of asymptomatic CAF might prevent the cardiac problems to occur in the future.<sup>22</sup>

The catheter-technique for closure of coronary fistulae is a nonsurgical alternative technique reported to provide success.<sup>23</sup> The transcatheter closure, started in the early 1980s, is an effective and safe method used in the management of CAF.<sup>23</sup> Multiple different closure devices are used including balloons, stainless steel coils, patent ductus arteriosus coils with controlled delivery systems, and plain or covered stents.<sup>21–24</sup> Following transcatheterization, the feeding artery is occluded distal to all normal branches to the myocardium minimizing the left-to-right shunt, which results in normalization of myocardial perfusion.<sup>23</sup> This technique may effectively and safely be used in the closure of significantly symptomatic CAF in children. Four of our patients underwent coil embolization in the Clinic of Cardiology.

The results of both transcatheter and surgical approaches indicate a good prognosis. Close clinical follow-up in all patients should be considered after CAF closure.<sup>25</sup>

Our approach to the management of CAF includes surgical closure of symptomatic fistulae that cannot be closed by coil embolization and/or fistulae with accompanying cardiac pathologies requiring correction; epicardial ligation in cases of fistula track that can be epicardially detected; and in case of no detection of epicardial track, detection and closure of the draining end of fistula by pulmonary arteriotomy and/or correction of the accompanying cardiac pathology. In cases of CAF with no accompanying cardiac pathology and requiring only epicardial ligation, we do not use CPB; on the other hand, in all other cases we perform bicaval cannulation and use CPB with intermediate hypothermia and cold blood cardioplegia. We think that median sternotomy is an appropriate technique for all patients. All patients who have undergone coronary surgical interventions should be provided with follow-up care after hospital discharge to check for evidence of ischemia or recurrence of CAF. Patients should be checked for symptoms and other findings and should have ongoing cardiac follow-up monitoring that may include ECHO and repeat angiography as needed. We are of the opinion that the surgical management of CAF is a safe and effective method with no morbidity and mortality. Asymptomatic CAF should carefully be evaluated in view of potential cardiac risks and closely followed up.

## References

1. Angelini P. Normal and anomalous coronary arteries: definitions and classification. *Am Heart J* 1989;117:418–434.
2. Reidy JF, Sowton E, Ross DN. Transcatheter occlusion of coronary to bronchial anastomosis by detachable balloon combined with coronary angioplasty at same procedure. *Br Heart J* 1983;49:284–287.

3. Mavroudis C, Backer CL, Rocchini AP, Muster AJ, Gevitz M. Coronary artery fistulas in infants and children: a surgical review and discussion of coil embolization. *Ann Thorac Surg* 1997;**63**:1235–1242.
4. Vitarelli A, De Curtis G, Conde Y, et al. Assessment of congenital coronary artery fistulas by transesophageal color Doppler echocardiography. *Am J Med* 2002;**113**:127–133.
5. Iadanza A, del Pasqua A, Fineschi M, Pierli C. Three-vessel leftventricular microfistulization syndrome: a rare case of angina. *Int J Cardiol* 2004;**96**:109–111.
6. Umana E, Massey CV, Painter JA. Myocardial ischemia secondary to a large coronary–pulmonary fistula. *Angiology* 2002;**53**:353–357.
7. Fujimoto N, Onishi K, Tanabe M, et al. Two cases of giant aneurysm in coronary pulmonary artery fistula associated with atherosclerotic change. *Int J Cardiol* 2004;**97**:577–578.
8. Papazoglou PD, Mitsibounas D, Nanas JN. Left anterior descending coronary artery–left ventricular fistula presenting as unstable angina and syncope. *Int J Cardiol* 2004;**96**:121–122.
9. Mutlu H, Kucukoglu M, Ozhan H, Kansiz E, Oztürk S, Uner S. A case of coronary artery fistula draining into the pericardium causing hematoma. *Cardiovasc Surg* 2001;**9**:201–203.
10. Sapin P, Frantz E, Jain A, Nichols TC, Dehmer GJ. Coronary artery fistula: an abnormality affecting all age groups. *Medicine* 1990;**69**:101–103.
11. Balanescu S, Sangiorgi G, Castelvechio S, Medda M, Inglese L. Coronary artery fistulas: clinical consequences and methods of closure: a literature review. *Ital Heart J* 2001;**2**:669–676.
12. Wang NK, Hsieh LY, Shen CT, Lin YM. Coronary arteriovenous fistula in pediatric patients: a 17-year institutional experience. *J Formos Med Assoc* 2002;**101**:177–182.
13. Ozeki S, Utsunomiya T, Kishi T, et al. Coronary arteriovenous fistula presenting as chronic pericardial effusion. *Circ J* 2002;**66**:779–782.
14. Guray U, Guray Y, Ozbakir C, Yilmaz MB, Sasmaz H, Korkmaz S. Fistulous connection between internal mammary graft and pulmonary vasculature after coronary artery bypass grafting: a rare cause of continuous murmur. *Int J Cardiol* 2004;**96**:489–492.
15. Goswami NJ, Zabalgoitia M. Localization of a coronary artery fistula using contrast transesophageal echocardiography. *J Am Soc Echocardiogr* 2002;**15**:839–840.
16. Cox ID, Heald SC, Murday AJ. Value of transesophageal echocardiography in surgical ligation of coronary artery fistulas. *Heart* 1996;**76**:181.
17. Krishnamoorthy KM, Rao S. Transesophageal echocardiography for the diagnosis of coronary arteriovenous fistula. *Int J Cardiol* 2004;**96**:281–283.
18. Hofbeck M, Wild F, Singer H. Improved visualization of a coronary artery fistula by the laid-back aortogram. *Br Heart J* 1993;**70**:272.
19. Ropers D, Moshage W, Daniel WG, Jessi J, Gottwik M, Achenbach S. Visualization of coronary artery anomalies and their anatomic course by contrast-enhanced electron beam tomography and three-dimensional reconstruction. *Am J Cardiol* 2001;**15**(87):193–197.
20. Kamiya H, Yasuda T, Nagamine H, et al. Surgical treatment of congenital coronary artery fistulas: 27 years' experience and a review of the literature. *J Card Surg* 2002;**17**:173–177.
21. Wang S, Wu Q, Hu S, et al. Surgical treatment of 52 patients with congenital coronary artery fistulas. *Chin Med J (Engl)* 2001;**114**:752–755.
22. Urrutia-S CO, Falaschi G, Ott DA, Cooley DA. Surgical management of 56 patients with congenital coronary artery fistulas. *Ann Thorac Surg* 1983;**35**:300–307.
23. Armsby LR, Keane JF, Sherwood MC, Forbess JB, Pery SB, Lock JE. Management of coronary artery fistulae. Patient selection and results of transcatheter closure. *J Am Coll Cardiol* 2002;**39**:1026–1032.
24. Sreedharan M, Prasad G, Barooah B, Dash PK. Vortex coil embolisation of coronary artery fistula. *Int J Cardiol* 2004;**94**:323–324.
25. Sunder KR, Balakrishnan KG, Tharakan JA, et al. Coronary artery fistula in children and adults: a review of 25 cases with long-term observations. *Int J Cardiol* 1997;**58**:47–53.