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Coronary artery fistula: Review of 54 cases from single center experience*

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Abstract

Background: *Demographic and clinical characteristics and angiographic findings of Turkish patients with coronary artery fistula have been investigated in this study and diagnostic tests and treatment methods used in these patients have also been evaluated in detail.*

Methods: *We have examined the cardiac catheterization laboratory database retrospectively between March 2006 and July 2010. Among 49,567 patients, we have noted 60 patients diagnosed as coronary artery fistula. After coronary angiographic images were evaluated by two invasive cardiologists, 54 patients who had clear evidence of vessel of origin and drainage were included in the study.*

Results: *A total of 54 (0.1%) patients with coronary artery fistula were noted. Mean age was 56.7 ± 10.7 years; 42 out of 54 patients had accompanying cardiac disorders. Patients' complaints were directly associated with the presence of the fistula. Chest pain was the admission symptom in all of the patients with isolated coronary artery fistula. Six patients had coexistent congenital anomalies. Myocardial infarction with ST segment elevation occurred in 11 of the patients. In contrast to the previous reports, the most common artery of origin of the fistula was left anterior descending artery (50.8%) and pulmonary artery was found to be the most frequent region of the fistula drainage by 53.7%.*

Conclusions: *Our findings suggest that large fistulas originating from the proximal segments of coronary arteries may increase the likelihood of atherosclerosis and myocardial infarction even in asymptomatic patients with no evidence of ischemia in noninvasive tests and no dilatation of cardiac chambers, and should therefore be closed.* (Cardiol J 2012; 19, 3: 278–286)

Key words: congenital anomaly, coronary artery fistulas, congestive heart failure, coronary angiography, myocardial infarction

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Introduction

Coronary artery fistula has been described as a direct connection between a coronary artery and one of the cardiac chambers, large vessels or other vascular structures [1]. This abnormality occurs due to the persistence of intratrabecular gaps that are normally present in the intrauterine life. Majority of coronary artery fistulas are congenital; however, acquired fistulas may also occur following cardiac surgery [2]. Coronary artery fistulas are the most common hemodynamically significant congenital coronary artery abnormalities [3–5]. They constitute approximately 14% of all coronary artery abnormalities [6]. Coronary artery fistulas are observed in 0.002% of the general population. Its prevalence is around 0.08–0.4% among all congenital heart diseases and 0.3–0.8% in catheterization laboratories [7].

Clinical and angiographic characteristics of coronary artery fistulas in Turkish patients have not been previously investigated. Therefore, demographic and clinical characteristics and angiographic findings of our patients with coronary artery fistula have been investigated in this study and diagnostic tests and treatment methods used in these patients have also been evaluated in detail.

Methods

A total of 49,567 patients that had experienced coronary angiography in our institute for any reason between March 2006 and July 2010 were retrospectively reviewed for the presence of coronary fistula. Sixty patients with any type of coronary fistula were found in all 49,567 angiography reports and angiography CD's of these 60 patients were reviewed by two experts in coronary angiography. Fifty-four patients were diagnosed as coronary fistula, and these patient's past medical records were obtained to define demographic characteristics of patients, including age, gender, complaints, history of coronary artery disease, unstable angina or myocardial infarction, diabetes, hypertension, hypercholesterolemia, and cigarette smoking. To further define clinical characteristics of these patients, we attempted to contact 54 patients. Of 54 patients, 36 patients responded to invitation and gave their consent for further studies. Electrocardiography, telecardiography and echocardiography was performed to all contacted patients, while myocardial perfusion scintigraphy was performed in whom symptoms attributable to ischemia was present without presence of hemodynamically significant (more than 70%) atherosclerotic plaques in the region supplied by the fistulized artery. Coronary computed tomography (CT) angiography had been performed before our study in 10 out of 54 patients using Toshiba Aquilion 64-slice computerized tomography device. Three patients were additionally assessed by transesophageal echocardiography (TEE) indicated due to concomitant diseases.

Retrospective part of our study was approved by institutional scientific committee, while part of study that involved research on contacted subjects was approved by institutional ethics committee. All active participants gave their informed consent before study.

All previous angiograms of identified coronary fistula patients were reviewed by two experts in invasive cardiology using a commercial analysis software (Axiom Artis FC; Siemens AG Healthcare Sector, Forchheim, Germany). Patients whose coronary angiography remained doubtful for presence for fistula were withdrawn from further examination. In those that presence of fistula was confirmed, fistulas were classified according to their angiographic characteristics. Singular fistulas between a coronary artery and cardiac chamber or great vessel were termed as type I fistulas, while multiple small fistulas between coronary artery and left ventricle was termed as type II fistulas, as classified by Chiu et al. [8]. Type I fistulas were further classified as large (inner diameter > 1.5 mm) and small (inner diameter < 1.5 mm) fistulae. Origin and distal connection of all fistulae were recorded when clearly identifiable.

Twelve channel electrocardiography (ECG), telecardiography and echocardiography were performed to all patients that accepted and gave consent for further research. Echocardiography was performed by a cardiologist with an echocardiography device (Vivid 3,GE Vingmed Ultrasound, Horten, Norway) using a phased-array transducer with a maximal frequency of 2.5 GHz. To minimize artifacts, 2D gain was set to 70% and Doppler gain was set to 60%. Left ventricular wall thickness, systolic and diastolic left and right ventricular diameters, left and right atrial dimensions were measured in M-mode and B-mode as appropriate and recorded thereafter. Pulsed wave, continuous wave, color and tissue Doppler were used to determine maximal flow velocities, presence of stenosis and regurgitation, and diastolic functions, according to the recommended methods of American Society of Echocardiography.

For 23 patients that had less than < 70% stenosis in fistulized coronary artery myocardial perfusion scintigraphy using Tc-99m sestamibi was performed to determine fistula-related ischemia. To

Table 1. Admission symptoms and clinical comorbidities of patients and comparison of these findings with existence of significant coronary stenosis.

	Total (n = 54)	Significant stenosis		P
		Yes (n = 26)	No (n = 28)	
Chest pain[1]	32 (59.3%)	22(84.6%)	10 (35.7%)	0.001**
Shortness of breath[1]	17 (31.5%)	2 (7.7%)	15 (53.6%)	0.001**
Palpitation[+]	4 (7.4%)	1 (3.8%)	3 (10.7%)	0.612
Hemoptysis[+]	1 (1.9%)	1 (3.8%)	0 (0%)	0.481
Hypertension[1]	28 (51.9%)	21 (80.8%)	7 (25.0%)	0.001**
Diabetes mellitus[+]	11(20.4%)	8 (30.8%)	3 (10.7%)	0.068
Dyslipidemia[+]	12 (22.2%)	11 (42.3%)	1 (3.6%)	0.001**
Smoking[1]	14 (25.9%)	10 (38.5%)	4 (14.3%)	0.043*
Ischemic heart disease	25 (46.3%)	25 (96.2%)	0 (0.0%)	0.001**
Congenital heart disease[+]	6 (11.1%)	1 (3.8%)	5 (17.9%)	0.194
Valvular heart disease[1]	12 (22.2%)	2 (7.7%)	10 (35.7%)	0.013*
Cardiomyopathy[1]	2 (3.7%)	0 (0.0%)	2 (7.1%)	0.491
STEMI[1]	11 (20.4%)	11 (42.3%)	0 (.0.0%)	0.001**
NSTEMI/USAP[+]	8 (14.8%)	8 (30.8%)	0 (0.0%)	0.002**
CABG[+]	3 (5.6%)	3 (11.5%)	0 (0.0%)	0.105
Valve replacement[+]	8 (14.8%)	0 (0.0%)	8 (28.6%)	0.004**

[1]Chi square test;[+]Fisher's exact test; **p < 0.01; *p < 0.05; CABG — coronary artery by-pass graft surgery; STEMI — ST elevation myocardial infarction; NSTEMI — non-ST elevation myocardial infarction; USAP — unstable angina pectoris

patients who are eligible for exercise testing, treadmill test was used and 85% of age corrected maximal heart rate was accepted as adequate threshold for ischemia induction. For the rest of patients, dipyridamole with a dose of 0.56 mg/kg was administered. Stress images were acquired immediately after exercise or dipyridamole administration, and test images were obtained 4 h after. An expert in cardiac nuclear medicine reviewed these images and areas with stress-induced ischemia were recorded according to distribution of coronary arteries.

Statistical analysis

Statistical analyses were performed using SPSS (Statistical Package for Social Sciences) for Windows 17.0 software. Quantitative variables were expressed as mean, qualitative variables were expressed as percentage (%) values. Univariate comparison between continuous variables was performed with the Student T test or Mann-Whitney U test, and for categorical data, comparison was performed with the χ^2 test or Fisher exact test. A p value < 0.05 was considered statistically significant.

Results

Demographic variables, admission complaints and clinical comorbidities of patients were given in Table 1, while angiographic characteristics were summarized in Table 2. A total of 54 (0.1%) patients with coronary artery fistula were noted among 49,567 coronary angiographies performed between 2006 and 2010. Mean age was 56.7 ± 10.7 years (range: 34–80). Among the study population, 31 (57.4%) patients were men. While coronary artery fistula was thought to be congenital in 53 patients, one patient had acquired fistula between left internal mammary artery (LIMA) and the pulmonary artery due to previous open coronary artery bypass graft surgery (CABG). There was a total of 59 fistulas in 54 patients. Two (3.7%) patients had concomitant right coronary artery (RCA) and circumflex artery (Cx) originated fistulas, while 3 (5.6%) patients had concomitant left anterior descending artery (LAD) and RCA originated fistulas. In 12 out of 54 (22.2%) patients fistula was the sole final diagnosis that remained as the unique possible cause of admission symptoms. Presenting symptom was chest pain in all these patients with "isolated" coronary fistula. The existence of accompanying coroner artery stenosis (> 50%) were found to be significantly higher in coronary fistula patients with chest pain complaints and non critical coronary artery disease were found to be significantly higher in patients referring with dyspnea. Depending on the results of risk factor analysis in coronary fistu-

Table 2. Angiographic characteristics of coronary fistulas and comparison of these findings with existence of significant coronary stenosis.

	Total	Significant stenosis		P
		Yes	No	
Fistula's artery of origin	**N = 59**			
Left anterior descending artery	30 (50.8%)	12 (42.9%)	18 (58.1%)	0.243
Right coronary artery	20 (33.9%)	13 (46.4%)	7 (22.6%)	0.170
Circumflex artery+	7 (11.9%)	2 (7.1%)	5 (16.1%)	0.240
Left internal mammary artery+	1 (1.7%)	1 (3.6%)	0 (0.0%)	1.000
Intermediary artery+	1 (1.7%)	0 (0.0%)	1 (3.2%)	0.475
Drainage chamber or vessel	**N = 54**			
Pulmonary artery	29 (53.7%)	12 (46.2%)	17 (60.7%)	0.540
Left ventricle+	9 (16.7%)	3 (11.5%)	6 (21.4%)	0.487
Right ventricle+	8 (14.8%)	5 (19.2%)	3 (10.7%)	0.442
Right atrium+	5 (9.3%)	3 (11.5%)	2 (7.1%)	0.647
Superior vena cava+	1 (1.9%)	1 (3.8%)	0 (0.0%)	0.451
Left atrium+	1 (1.9%)	1 (3.8%)	0 (0.0%)	0.451
Left upper lung+	1 (1.9%)	1 (3.8%)	0 (0.0%)	0.451

Chi square test; +Fisher's exact test; **p < 0.01; *p < 0.05

la patients, hypertension, smoking and dyslipidemia were found to be significantly higher in accompanying critical coronary artery disease.

According to their coronary angiographic images, 54 patients diagnosed as coronary artery fistula were grouped as type I and type II coronary fistula. Type I fistulas were further grouped as large (diameter ≥ 1.5 mm) and small (diameter < 1.5 mm). According to this classification, 31 (57.4%) patients had large fistulas, 18 (33.3%) had small fistulas, and 5 (9.3%) had microfistulas. Two different fistulas were simultaneously present in different arteries of the same patient in 5 patients and all of these were large fistulas. When accompanying vessel lesions of the patients were evaluated considering ≥ 50% stenosis as significant, 18 (33.3%) LAD, 7 (13%) Cx, and 15 (27.8%) RCA lesions were noted. No lesion causing significant stenosis in coronary arteries was noted in 28 (51.9%) patients. Aneurysmatic dilatation of the feeding vessel (RCA) was observed in only 1 (1.85%) patient. Coronary fistula was accompanied by a myocardial bridge (LAD) in one patient.

Primary percutaneous intervention (PCI) was performed in four patients admitting with myocardial infarction with ST elevation (STEMI). The fistulae were located in the proximal LAD and drained into the pulmonary artery in all of these cases (Fig. 1). PCI was performed in all of these four vessels which were occluded just distal to the fistula with no distal flow. TIMI III flow was established in two of these cases and fistular flow was noted to be significantly reduced after the occlusion beyond the fistula was eliminated. This so-called reverse steal phenomenon describes that fistula flow which was previously towards the pulmonary artery, hence-

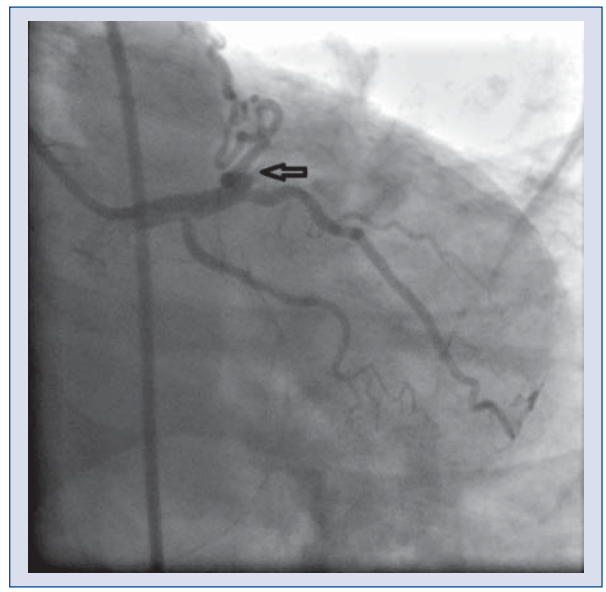

Figure 1. Fistula originating from proximal left anterior descending artery (LAD) (arrow) and complete occlusion of LAD just below the fistula. Slow coronary flow developed after primary percutaneous coronary intervention and fistula flow persisted.

Table 3. Echocardiographic findings of the patients (n = 36, 18 with significant stenosis) and indicators of right ventricule systolic functions (n = 23, 15 with significant stenosis) and comparison of these findings with existence of significant coronary stenosis.

	Total (n = 36)	Significant stenosis		P
		Yes (n = 18)	No (n = 18)	
Right ventricular expansion[1]	10 (27.7%)	2 (11.1%)	8 (44.4%)	0.026*
Left ventricular wall motion abnormality[1]	13 (36.1%)	12 (66.7%)	1 (5.6%)	0.001**
Left atrial expansion[1]	14 (38.9%)	4 (22.2%)	10 (55.6%)	0.040*
Ejection fraction[2]	51.8 ± 11	49.72 ± 10.21	53.89 ± 11.83	0.266
Left ventricular systolic diameter[2]	34.5 ± 8.8	34.67 ± 8.19	34.33 ± 9.74	0.912
Left ventricular diastolic diameter[2]	50.7 ± 6.7	50.06 ± 5.86	51.50 ± 7.65	0.529
Estimated pulmonary artery pressure[2]	33.3 ± 17.7	26.17 ± 6.43	40.55 ± 22.31	0.016*
	Total (n = 23)	**Significant stenosis**		**P**
		Yes (n = 15)	**No (n = 8)**	
TAPSE (median)[3]	2.25 ± 0.36 (2.20)	2.13 ± 0.30 (2.10)	2.50 ± 0.37 (2.50)	0.027*
IVA (median)[3]	3.4 ± 0.98 (3.10)	3.16 ± 0.76 (3.00)	3.85 ± 1.25 (3.95)	0.081
Sm wave (median)[3]	15.7 ± 3.1 (16.0)	15.16 ± 2.33 (15.00)	16.75 ± 4.23 (17.00)	0.037*

[1]Chi Square Test; [2]Student t Test; [3]Mann-Whitney U Test; **p < 0.01; *p < 0.05; TAPSE — tricuspid annular plane systolic excursion, IVA — myocardial acceleration during isovolumic contraction [m/s^2], Sm wave — right ventricular peak myocardial systolic velocity

forth redirected to jeopardized myocardium. No reduction in fistula flow was observed in 2 patients in whom significantly slow flow in infarct related artery noted after PCI.

ECG evaluation of the 36 patients revealed normal sinus rhythm in 30 patients and atrial fibrillation in 6 patients. ECG was completely normal in 16 (44.4%) patients. On telecardiography, cardiothoracic ratio was more than > 0.5 in 10 (27.7%) patients. ECG findings and telecardiography was normal in all patients with "isolated" coronary fistula.

Echocardiography was performed in all 36 patients included in the study. Additionally, in 23 patients that had no accompanying disorders which may impair pulmonary circulation, right ventricular systolic functions were evaluated. Echocardiographic findings of the patients and indicators of right ventricle systolic functions were shown in Table 3. The existence of non significant coronary artery disease were higher in patients with right ventricular chamber enlargement whereas mean TAPSE and Sm wave mean values were found to be lower in patients having critical coronary artery stenoses. Transthoracic echocardiography confirmed the turbulent flow at the distal end of the fistula in 2 patients, out of 36 (5.6%) during transthoracic echocardiography (TTE). Qp/Qs ratio was found as < 1.5 in all except one patient who had atrial septal defect and dilated right heart chambers. One (2.7%) patient had PDA, one patient had VSD (2.7%), and three patients had ASD (8.3%) accompanying fistula.

Ten patients who had diagnosed as coronary fistula by conventional coronary angiography were subjected to CT angiography. In 9 of 10 patients, origin of coronary fistula was found in concordance with conventional angiography findings. CT angiography failed to confirm only 1 patient with Cx to pulmonary artery type I large fistula which was clearly demonstrated with conventional coronary angiography. In 5 patients, coronary CT angiography clearly showed distal anastomosis of fistula, while in the rest of patients distal anastomosis could not be demonstrated with CT angiography. Coronary CT angiography findings in patients were given in Table 4.

Stress TI-201 Myocardial Perfusion Gated Spect Scintigraphy was performed to investigate the presence of ischemia in 23 patients with no significant stenosis noted in the vessel of origin. While perfusion defect was observed in 7 (30.4%) patients, no evidence of ischemia was found in 16 (69.6%). In 7 patients with evidence of ischemia, small fistulas were present in 2 and large fistulas were present in 5 patients. In 2 patients with type II fistula, ischemia could not be demonstrated .

Table 4. Coronary computed tomography (CT) angiography findings of ten patients.

Patient	Origin	Drainage	Type	CT origin	CT drainage
1	CX	PA	I large	–	–
2	LAD	PA	I large	+	–
3	RCA	SVC	I large	+	+
4	LAD	LA	I large	+	+
5	LAD	PA	I small	+	–
6	RCA	RV	I small	+	–
7	CX	LV	II	+	+
8	LAD	PA	I large	+	+
9	LAD-RCA	PA	I large	+	+
10	IM	PA	I large	+	–

CX — circumflex artery; PA — pulmonary artery; LAD — left anterior descending artery; RCA — right coronary artery; SVC — superior vena cava; LA — left atrium; LV — left ventricle; RV — right ventricle; IM — intermediary artery

Table 5. Characteristics of patients treated by coil embolization.

Patient	Origin-drainage	Size	Symptom	Indication	Treatment — follow-up
1	LAD-PA	Large fistula	Chest pain	Anterior ischemia on MPS	Closed by 2 coils. Control MPS normal
2	RCA-SVC	Large fistula	Chest pain, variant angina	CT angiography Aneurysmatic dilatation of RCA with 8 mm of lumen diameter at the widest	Closed by 4 coils. On control residual fistula noted on control CAG
3	RCA-left upper lung	Large fistula	Massive hemoptysis	MPS normal. Life-threatening massive hemoptysis as a rare symptom associated with the fistula	Closed by 5 coils. Complete symptom remission

CX — circumflex artery; PA — pulmonary artery; LAD — left anterior descending artery; RCA — right coronary artery; SVC — superior vena cava; LA — left atrium; LV — left ventricle; RV — right ventricle; CT — computed tomography; MPS — myocardial perfusion scintigraphy; CAG — coronary angiography

Among 54 patients who had been found to have coronary fistulas on angiography, 3 patients had been treated surgically (ligation), and 2 patients had been treated percutaneously by coil embolization method. One patient was treated by coil embolization method during our study. Among patients treated surgically, 2 patients had been operated for mitral valve surgery, and 1 for aortic valve surgery. Conservative approach had been utilized in 48 patients. Important clinical features of the 3 patients treated by coil embolization are presented in Table 5.

Discussion

Coronary artery fistulas are very rare congenital coronary termination abnormalities. Up until now, several reports about coronary artery fistulas have been published from the US, Europe and Far East countries. No previous reports reaching our sample size have been made from our country. We have evaluated 54 patients diagnosed as coronary artery fistula in our catheter laboratory within the last 4 years in terms of demographic and clinical features, diagnostic methods, morphological features of the fistulas and treatment methods. This is the first large-scale single center retrospective study investigating coronary artery fistulas in Turkish patients.

The 4-year coronary artery fistula incidence was found as 0.1% in our hospital's catheter laboratory. The general incidence of fistula has been reported within the range of 0.3% to 0.8% in several previous studies [7]. Those fistulas between a coronary artery and a cardiac chamber or large artery were defined as type I fistulas, while multiple microfistulas between a coronary artery and left ventricle were defined as type II fistulas. Type I

fistulas were further grouped as large (diameter ≥ 1.5 mm) and small (diameter < 1.5 mm). According to this classification, 31 (57.4%) patients had large fistulas, 18 (33.3%) had small fistulas, and 5 (9.3%) had microfistulas. RCA or its branches have been reported as the most frequent artery of origin of the fistula in many studies by 55%. Coronary artery fistula originates from the left coronary system in 35% of the patients, and from both coronary arteries at the same time in 5% [9]. Vessels of origin were noted as LAD in 50.8%, RCA in 33.8%, and Cx in 11.8% in this study. In parallel with our study, both Carrel et al. [10] and Abdelmoneim et al. [11] have found the most common artery of origin of the fistula as LAD. More than 90% of the fistulas drain into venous structures. Low pressure chambers are the most regions of drainage for the fistulas. These can be listed in descending order of frequency as right ventricle by 41%, right atrium by 26%, pulmonary artery by 17%, left ventricle by 3%, superior vena cava by 1% [3]. Left ventricle has been reported as one of the rarest drainage regions in nearly all studies. In contrast to previous reports, pulmonary artery was found to be the most frequent drainage region of the fistula by 53.7% in our study, followed by left ventricle in 16.7%, right ventricle in 14.8%, right atrium in 9.3% and superior vena cava, left atrium and left upper lung parenchyma in very few patients. Number of fistula opening into the left ventricle was significantly higher compared to other reports. Abdelmoneim et al. [11] have also reported the most common drainage region as pulmonary artery by 50%; however, there were no fistula cases opening into the left chambers in this study of 30 patients. There was no significant difference in the origin and drainage sites of fistulas between the patients with critical or non critical coronary artery disease.

Most of the fistula patients are generally asymptomatic. However, dyspnea, fatigue, palpitation, chest pain, congestive heart failure, arrhythmia, sudden death, infective endocarditis and rupture may be present or the patients may present symptoms and signs of concomitant atherosclerotic heart disease and/or valvular heart disease [12]. The reasons for clinical admission of patients with coronary artery fistulas depend on the severity of left-to-right shunt [13]. In our study all patients that we examined were symptomatic. This might be due to the fact that our clinic is a tertiary center since 42 out of 54 patients had accompanying cardiac disorders and their fistulas were detected coincidentally while they were being investigated for accompanying disorders. The most frequent complaint in our patients was chest pain (typical or atypical) by 59.3%. Chest pain complaint was more frequent in patients with concomitant significant vessel disease as compared with non-significant. So it's difficult to attribute this symptom to coronary fistula in patients with accompanying coronary artery disease. The other symptoms in our fistula patients were dyspnea (31.5%), palpitation (7.4%) and hemoptysis (1.9%), respectively. We noticed 12 fistula patients without coronary and valvular heart disease in whom symptoms were attributable to the fistula. Chest pain was the main complaint of all our 12 patients with isolated fistula and atypical angina was present in most of these patients (75%). Our findings suggest that coronary artery fistulas may cause angina without the presence of accompanying occlusive vessel lesions. This can be explained as follows: as some of the coronary blood flow is directed towards another chamber via the fistula, the resting coronary flow is continuously kept at a high rate in order to compensate for this stolen blood flow. When exercising, myocardial perfusion may be inadequate due to inability of coronary flow reserve to be augmented [14]. Therefore, ischemic symptoms may occur when the fistula is accompanied by uncritical atherosclerotic stenosis or even in individuals with normal coronary arteries especially during exercise.

Coronary angiography is the gold standard method for the diagnosis of coronary fistula. However, the relationship of the fistula with other structures, its vessel of origin and drainage regions may not always be clearly delineated by coronary angiography. In such instances, multi-slice CT coronary angiography and magnetic resonance imaging may be used as complementary methods. Sensitivity of CT coronary angiography for the detection of coronary fistula was also evaluated based on coronary angiography in our study. While multi-slice CT confirmed the presence of fistula in 9 patients, it failed to demonstrate the fistula in only 1 patient. While structures of fistula origin and drainage were clearly demonstrated in 5 out of 9 patients who were confirmed to have fistula, drainage structures could not be accurately visualized in 4 patients, which were clearly demonstrated with coronary angiography. Although coronary CT angiography is a non-invasive method, which can visualize cardiac anatomy in full detail and diagnose the fistula with a 90% success rate, it was able to demonstrate the course and drainage region of the fistula in 50% of our cases. In conclusion, it is clear that coronary CT angiography cannot replace conventional coronary angiography particularly when determining the treatment approach but may be used as a complementary test.

Qp/Qs values of patients (excluding the one patient with a large ASD) in whom the amount of left-to-right shunt was assessed by echocardiography were found to range between 1.0 and 1.5. Although further studies evaluating more number of patients are needed to confirm these findings, we may state that the extent of shunts caused by coronary fistulas are rather less compared to shunts caused by other congenital heart diseases causing left-to-right shunt such as ASD, PDA, VSD. Qp/Qs was measured with right heart cardiac catheterization and > 1.5 was noted in 3 patients and 2 cases were related to coronary artery fistulas in a clinical study of 125 patients from China [8]. Reasonable association between right ventricular functions and coronary artery fistulas couldn't be found in this study and further studies will be needed to demonstrate this issue.

In a TI-201 SPECT study performed in 23 patients with no accompanying vessel lesions, perfusion defect in the region supplied by the vessel of origin of the fistula was noted only in 7 (30.4%) patients. Perfusion defect was noted in 11 out of 20 patients (55%) evaluated by myocardial perfusion scintigraphy in a Dutch study [15]. In another study from China, perfusion defect was found in all of the patient group consisting of 33 patients [8]. Myocardial perfusion scintigraphy which was performed to demonstrate the prevalence of coronary steal syndrome has revealed that fistula related ischemia was less prevalent in our study compared to the mentioned studies.

It is very well-known that bifurcations and ostiums of side branches are highly susceptible for the development of atherosclerosis due to the changes in flow dynamics. We want to emphasize the impact of the fistula on local wall shear stress which causes a sudden change in direction of flow with a serious pressure difference. Such flow disturbances influence the behaviour of endothelial cells, including morphological adaptations, and expression of inflammatory cell attractors. Thus, we may hypothesize that the stenosis neighbouring the fistula in 4 of our 11 STEMI patients may have developed secondary to the flow changes caused by the large fistula (≥ 1.5 mm) in addition to coronary risk factors. In contrast to the findings of Sherwood et al. [16] who have suggested that asymptomatic fistulas were not associated with a negative prognosis, our findings suggest that large fistulas originating from the proximal segments of coronary arteries may increase the likelihood of atherosclerosis and myocardial infarction even in asymptomatic patients with no evidence of ischemia in non-invasive tests and no dilatation of cardiac chambers.

Reverse steal phenomenon is another issue that will be emphasized. It has been mentioned in two different cases until today. In patients with fistulas originating before critical stenosis, when occlusion in the coronary artery is relieved and distal flow is re-established, blood flow directed towards the fistula is re-directed towards the native vessel and fistula flow is significantly reduced [17, 18]. Significant reduction in fistula flow was observed in two of our patients with total lesions who were successfully treated and TIMI III flow was established. No reduction in fistula flow was observed although distal flow was achieved in 2 other patients developing considerably slow flow following the intervention. This can be attributed to the fact that resistance to coronary blood flow continues at the microvascular level and blood flow is directed towards the low pressure fistula region. In the light of these findings, it can be suggested that primarily the coronary lesion should be aimed and decline or even disappearance of fistula flow can be anticipated after intervention in patients with critical stenosis beyond the fistula, instead of treating coronary fistula and critical lesion separately.

Treatment decision should be based on the location, size and the amount of shunt in a coronary artery fistula. Coronary fistulas should be treated in the presence of large fistulas and progressive left-to-right shunt, left ventricle volume loading findings, myocardial ischemia, left ventricle dysfunction, congestive heart failure and in order to prevent endocarditis/endarteritis. Generally clinical follow-up is recommended in fistulas that do not cause significant shunt. There are two treatment alternatives; surgery or closure by catheter. It has been reported that transcatheter methods and surgical methods had similar efficacy, mortality and morbidity rates [19]. When treatment methods used for the management of coronary artery fistulas were analysed, medical treatment was preferred most frequently in our hospital. 48 of the 54 patients were treated medically, 3 of the patients were treated surgically and the other three patient were treated by coil embolization. The most important reasons might be that symptoms usually are not directly attributed to the fistula due to accompanying disorders and symptoms in patients with isolated fistula are often not quite serious. As demonstrated in our study, absence of a large left-to-right shunt (Qp/Qs < 1.5), absence of any significant accompanying echocardiographic right and left ventricle loading findings, low prevalence of fistula related ischemia might have played role in the preference of medical treatment.

Limitations of the study

Our study has certain limitations due to the retrospective nature of the database used. Coronary fistulas are very rare congenital anomalies therefore small number of patients could be collected. We searched fistula word in around 50.000 coronary angiography reports and found 54 patients with fistula. Certainly, we are aware that some small fistulas are often overlooked which might lead to underestimation of the prevalence of coronary artery fistula. Demographic and angiographic data were obtained from all 54 patients who were studied retrospectively for the presence of coronary artery fistula; however, 36 patients who gave informed consent were studied for further analysis. As a retrospective study, selection of treatment modality was made by the clinician at the time of case evaluation. Thus, we couldn't compare alternative treatment methods.

Conclusions

In conclusion, we believe that all large fistulas (≥ 1.5 mm) specially, which are originated from proximal LAD, open into a single chamber with a single outflow point that do not cause aneurysmatic changes and are not very tortuous in nature should be closed percutaneously without considering the presence of symptoms.

Conflict of interest: none declared

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