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Prevalence and characteristics of coronary artery fistula in adults: coronary angiographic analysis of 18,106 patients

Aim	Coronary artery fistula (CAF) is a rarely encountered anomaly that is characterized by an abnormal connection between a coronary artery and a cardiac chamber or a great thoracic vessel. Its incidence has not been precisely established due to the large number of undiagnosed cases and it shows heterogeneity in its anatomic configuration and clinical consequences. We aimed to assess the frequency, imaging findings, and clinical features of CAF among patients in our tertiary medical center.
Material and methods	The angiographic data of 18,106 consecutive adult patients who underwent coronary angiography between January 2011 and June 2013 were retrospectively analyzed.
Results	CAF was detected in 22 patients (0.14%). Of these, 5 patients had bilateral fistulas (23%). 65% of the fistulas originated from the left anterior descending coronary artery,and 53% drained into the pulmonary artery. The left ventricle and left atrium were the only drainage sites for left-sided coronary artery fistulas. One patient with a CAF presented with non-ST elevated myocardial infarction in the absence of an evident thrombosis.
Conclusion	Unlike previous reports, bilateral CAFs were more commonly encountered in this study. Contrary to most of the data in the literature, more than half of the CAFs originated from the left anterior descending coronary artery and most drained into the pulmonary artery. Rare anatomic types of CAFs were also detected.

Keywords	Fistula; coronary vessels; coronary vessel anomalies; coronary angiography
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Introduction

Coronary artery fistula (CAF) accounts for 14% of all coronary artery anomalies. It is defined as an abnormal connection between a coronary artery and any of the cardiac chambers or a great thoracic vessel [1–3]. CAF may develop as an acquired disease, but 20 to 45% of cases are congenital and are accompanied by other congenital heart defects [4]. It is noteworthy that some congenital fistulas may spontaneously regress during childhood, and that approximately half of all patients with a diagnosed CAF are asymptomatic [5, 6]. Therefore, it would be speculative to state the incidence of CAF in the general population. As to the site of the fistula, the right coronary artery (RCA) accounts for about 33–55% of cases, the left coronary artery (LCA) for about 35–49% of cases, and both coronary arteries for a few cases [7]. Most CAFs drain into the right ventricle (RV), the right atrium (RA), or the coronary sinus. Coronary to pulmonary artery (PA) fistulas are rare [7]. Here, we report the prevalence and characteristics of CAFs among patients who underwent coronary angiography at our tertiary medical center.

Material and methods

The medical records of 18,106 consecutive patients who underwent coronary angiography at a Ankara tertiary center between January 2011 and June 2013 were evaluated. Cine-angiograms and digital data of these patients were retrospectively examined with a picture archiving system by at least two experienced cardiologists. The patients with CAF were selected for further analysis. Their clinical and angiographic data were evaluated, and relevant demographic features, correlated symptoms, and coexisting pathologic conditions were recorded. The LCA was reviewed using at least four projections, and the RCA was reviewed using at least two projections. CAF was diagnosed if there was an anomalous connection between one or more coronary arteries and a cardiac chamber or a great thoracic vessel [1]. The origin and termination of the fistula, together with the coronary anatomy and neighboring structures were identified. The study was approved by the institutional ethics committee, and due to the retrospective design of the study, informed consent was waived.

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Statistical Analysis

Descriptive statistical analyses were performed. Categorical variables are presented as frequencies or percentages, and continuous variables are presented as means ± SD. Data were analyzed using Statistical Package for Social Sciences (SPSS) program version 20.0 for Windows. (SPSS Inc. Chicago, Illinois, USA)

Results

Of the 18,106 patients studied, 22 patients (0.14%; mean age 55.3± 9.6 yrs) had CAF. Clinical characteristics of these patients are listed in Table 1. Of these patients, 13 (59%) were male and 9 (41%) were female. All patients had symptoms of angina or dyspnea on clinical presentation. Angina was the most common symptom and was present in 16 (73%) cases; dyspnea was present in 6 (27%) cases. Coronary artery disease was evident in 11 patients. One patient had severe mitral regurgitation, and 1 patient had left ventricular global hypokinesis detected by echocardiography in the absence of atherosclerotic coronary artery disease. Myocardial bridge was present in 3 patients. One patient had non-ST elevation myocardial infarction in the absence of an evident thrombosis (Figure 1).

The fistulas were unilateral in 17 (77%) patients and bilateral in 5 (23%). The origin of the unilateral fistulas was from the left anterior descending artery (LAD) in 11 (65%), from the circumflex artery (Cx) in 3 (17.5%), and from the RCA in 3 (17.5%) cases. The termination site of the unilateral fistulas was the PA in 9 (53%), RV in 4 (23.5%), left ventricle (LV) in 2 (11.7%), RA in 1 (5.9%), and left atrium (LA) in 1 (5.9%) cases. Overall, the PA was the most frequent drainage site for unilateral fistulas; 7 of 9 fistulas originating from the LAD terminated in the PA. The LV and the LA were the only drainage sites for left-sided

Table 1. Clinical characteristics of patients with coronary artery fistulas

Age (yrs)	Gender	Presenting Symptom	Origin of CAF	Termination of CAF
47	Male	Angina	LAD	PA
72	Female	Angina	LAD	LV
61	Male	Dyspnea	LAD	RV
52	Male	Angina	RCA, Cx	PA, PA
41	Female	Dyspnea	LAD	PA
61	Female	Angina	LAD, RCA	PA, PA
58	Male	Dyspnea	LAD	PA
48	Female	Dyspnea	LAD	LA
59	Male	Dyspnea	RCA	RV
44	Female	Angina	LAD	PA
45	Male	Angina	LAD	RV
42	Male	Dyspnea	Cx	RA
65	Male	Angina	Cx	PA
54	Male	Angina	LAD	PA
55	Female	Angina	RCA	PA
73	Female	Angina	RCA, Cx	RA, RA
59	Male	Angina	Cx	LV
42	Female	Angina	LAD	PA
70	Male	Angina	RCA	RV
58	Male	Angina	LAD	PA
55	Female	Angina	LAD, Cx	LV, LV
55	Male	Angina	LAD, RCA	PA, PA

LAD, left anterior descending coronary artery; Cx, circumflex coronary artery; RCA, right coronary artery; PA, pulmonary artery; RV, right ventricle; RA, right atrium; LA, left atrium.

Figure 1. Left panel: Angiographic view of the LAD and Cx showing no thrombosis. Right panel: Angiographic view of the RCA showing a CAF originating from it and draining into the PA. A thrombus is not evident

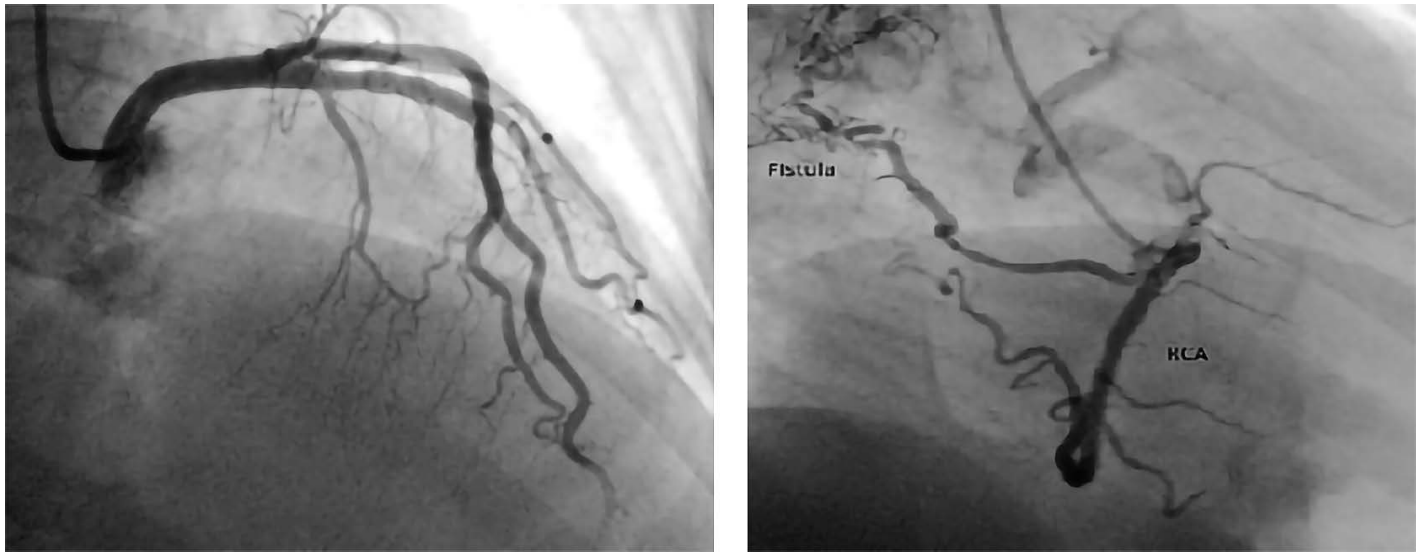


Figure 2. Angiographic view of a CAF originating from the LAD and draining into the LA

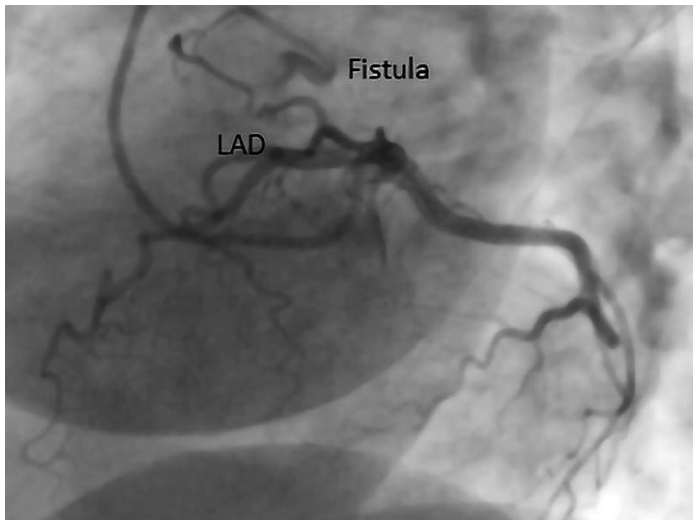
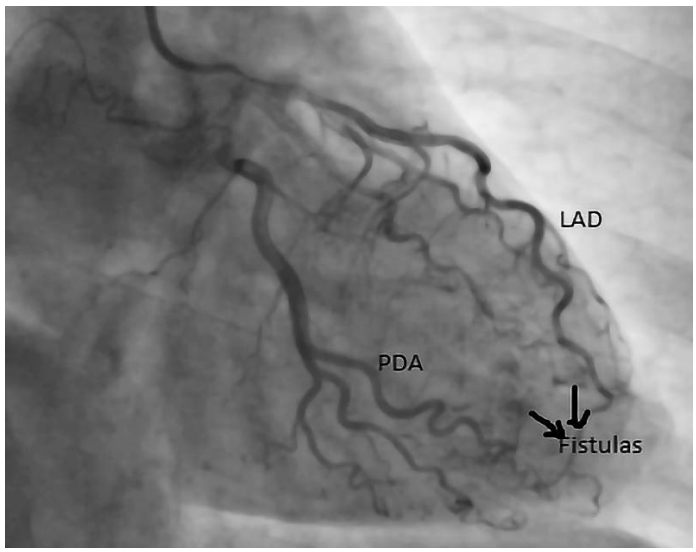


Figure 3. Angiographic view of CAFs originating from the LAD and from the RCA and draining into the PA



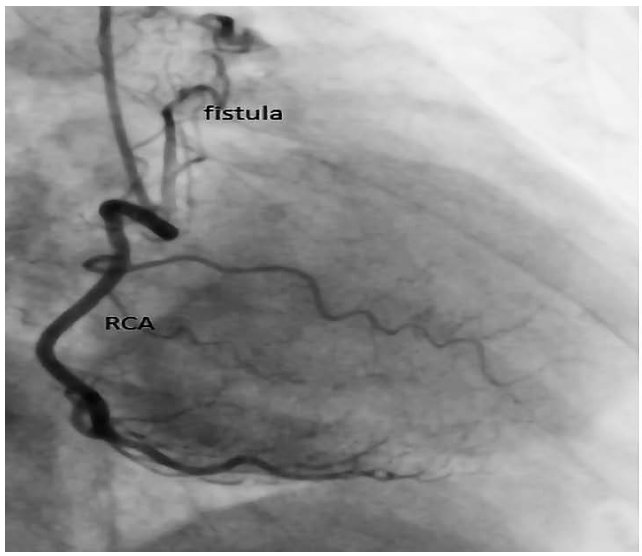
coronary arteries (Figure 2). Among patients with bilateral fistulas, these originated from the LAD and the Cx in 1, from the LAD and the RCA in 2, and from the RCA and the Cx in 2 cases (Figures 3 and 4). Six (60%) of the bilateral fistulas drained into the PA. Of the remaining fistulas, 2 (20%) terminated in the RA and 2 (20%) in the LV. Proximal CAF were usually large, whereas distal fistulas were usually smaller and more tortuous. Additionally, one patient had a giant fistula originating from the proximal RCA and terminating in the PA. This fistula caused a massive enlargement of the RCA (Figure 5)

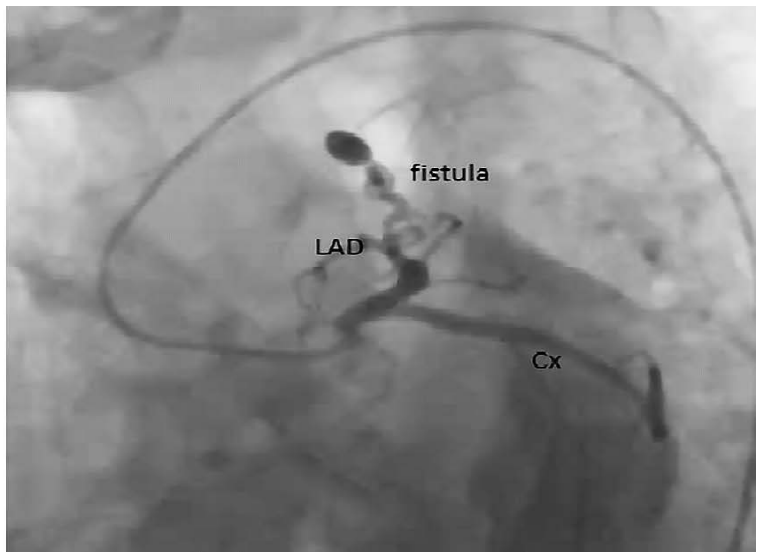
Discussion

Although the exact incidence of CAF is as yet unpredictable due to the unknown rate of undiagnosed cases, the number of persons with CAF in the general population has been estimated to be 0.002% [1, 4, 8]. Among patients who had undergone coronary angiography, previous studies reported the prevalence of CAF to range from 0.06 to 0.37% [4, 8–13]. In the present study, we found a rate of 0.14% among a total of 18,106 patients, which is within the reported range. The largest reported serial study, which was comprised of 298,558 Polish patients, found a CAF prevalence of 0.087% [11]. The highest CAF prevalence, 0.37%, was found in an earlier Turkish study [12]. Although no racial predilection for CAF is known, CAF prevalence varied among different populations. We presume that this variation was related to differences in criteria for performing coronary angiography, or to the success in detecting fistulas due to the difficulty in cannulating some of the anomalous arteries.

Regarding the origin and termination sites of CAFs, it has been historically accepted that most CAFs originate from the RCA, and most terminate in the RV, RA, or coronary

Figure 4. Angiographic view of CAFs originating from the LAD and from the RCA and draining into the PA





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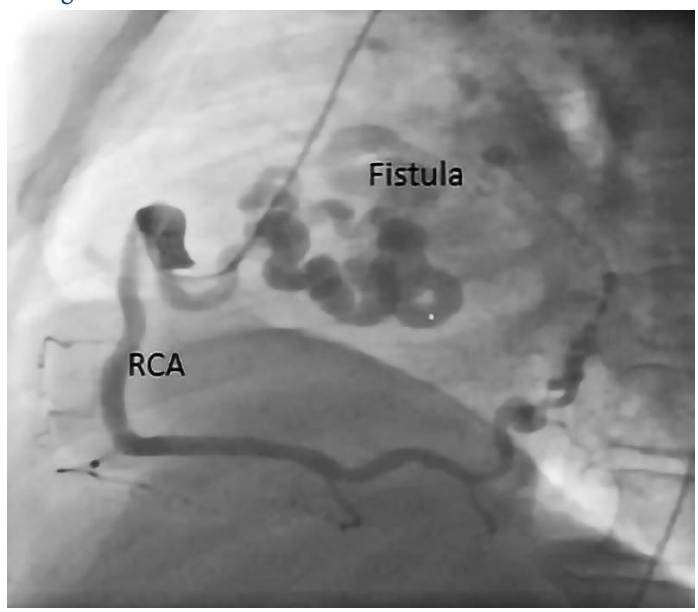
sinus [1, 4, 7, 14–17]. However, in the current study, the LAD was the most common site of origin, and the PA was the most common site of termination. These current findings agree with some prior reports [7, 12, 18–22], and they are consistent with the findings of the largest reported study of CAFs, which found that the LAD was the most frequent site of CAF origin, and that the PA was the most frequent site of CAF termination [12].

According to a detailed analysis of CAFs conducted by Said et al., some types of CAFs are very rarely mentioned in the literature [20]. These include fistulas originating from the LAD or the Cx, which account for 3% and 4% of all CAFs, respectively, and which terminate in the LV [20]. Interestingly, in our study, there were two of each type of these rare fistulas. According to the number of fistulas in a single patient, CAF can also be classified as unilateral, bilateral or multiple. Unilateral fistulas are the most common type, occurring in 74–90% of cases; bilateral fistulas are very rare and account for 5% of all fistulas [20]. Intriguingly, in our series, bilateral fistula were encountered in 5 patients, accounting for 23% of the cases. We also detected a rare type of bilateral fistula that originated from the LAD and from the Cx and terminated in the LV. Of the bilateral fistulas described in the literature, only 2% were of this type [20].

CAF can manifest in a diverse clinical spectrum ranging from an asymptomatic state to symptoms related with the degree of shunting. Angina, arrhythmia, congestive heart failure, pulmonary hypertension and myocardial infarction are the possible clinical conditions that can occur in the setting of CAF. Coronary steal phenomenon involves a pressure gradient from a coronary artery to a lower pressure vein or chamber that "steals" collateral flow from under perfused myocardium and is the underlying pathophysiological mechanism attributed to the development of angina and myocardial infarction. A CAF would be expected to have a similar effect, Thus, it is surprising that myocardial infarction was present in only 2% of CAF cases [11, 13, 20, 21]. However, there was a CAF patient in our study that presented with a non-ST elevation myocardial infarction in the absence of an evident thrombosis on coronary angiography.

The size of the CAF and the symptoms are the main factors for determining the treatment modality. Since most fistulas are small and hemodynamically insignificant, they could be managed by clinical follow-up, including echocardiography every 3 to 5 years. Closure of mild to moderate fistulas is recommended in the presence of symptoms caused by documented myocardial infarction, arrhythmias, endocarditis, ventricular dilation or dysfunction of uncertain origin. On the other hand, a large CAF should be closed regardless of symptomatology [22–24].

Figure 5. Angiographic view of a CAF originating from the RCA and draining into the PA. Massive enlargement of the RCA is also evident



We acknowledge that this study has some limitations. Firstly, it is a single-center, retrospective study. Additionally, all patient data regarding the demographic features and correlated symptoms were based on the medical records and no laboratory tests could be performed. Finally, long term monitoring of the patients was absent due to the retrospective character of the study protocol.

Conclusion

In agreement with previous studies, CAF was rarely present. However, extremely rare types of unilateral and bilateral CAF were found. Contrary to the majority of previous reports, the LAD was the most frequent site of CAF origin, and the PA was the most frequent site of termination. Also, bilateral fistulas were much more commonly encountered than in previous studies. In one CAF case, the patient had non-ST elevation myocardial infarction with no evidence of a coronary thrombosis.

No conflict of interest is reported.

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