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# Prevalence and characteristics of patients with solitary coronary artery fistulas in 12,757 all-comer adult patients undergoing coronary angiography (RCD code: I-1C.4)

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## Abstract

**Introduction** Coronary artery fistula (CAF) is a rare anomaly, mostly congenital, defined as an abnormal direct connection between one of the coronary arteries and a heart chamber or major vessel. In case of coronary steal, CAF may manifest with symptoms of stable coronary disease (CAD). **Methods** We investigated medical records of all adult patients who underwent coronary angiography (CAG) from 1 July 2009 to 30 June 2019. Patients with solitary CAF were selected. Coronary artery-ventricular multiple microfistulas (MMFs) were recorded but were no object for further analysis. The CAFs were grouped depending on the origin and termination. Clinical symptoms, comorbidities and past medical history were analyzed. **Results** 26 solitary CAFs were found in 22 (0.17%) and 20 MMFs in 16 (0.13%) out of 12,757 patients who underwent CAG for any reason in that period. Indication for CAG varied among study group. Left coronary artery (LCA) gave origin to majority (57.69%) of fistulas. Pulmonary artery (PA) was the most common drainage site (69.23%). 17 patients had HA, 15 suffered from chest pain or dyspnoea, 13 had dyslipidemia, and half had CAD. **Conclusion** The incidence of solitary CAF in the sample of all-comer population of Polish patients undergoing CAG was 0.17%. LCA was the most common artery of origin and PA was the most frequent drainage site. The indication for CAG varied among patients. Majority of patients suffered from chest pain or dyspnea, had HA, dyslipidemia, and half had CAD. Moreover, the incidence of MMF was 0.13%. JRCD 2019; 4 (3): xx–xx

**Key words:** coronary artery fistula, coronary artery anomaly, rare cardiovascular disease, coronary angiography

## Background

Coronary artery fistula (CAF), first described by the Austrian anatomist Josef Hyrtl in 1841 [1], is a rare defect defined as an abnormal direct connection between one of the coronary arteries and a heart chamber or major vessel [2]. Two distinct congenital types are recognized: solitary CAFs and coronary artery-ventricular multiple microfistulas (MMFs) [3]. The majority of these fistulas are congenital and may occur along with other congenital heart diseases [1]. However, CAFs occasionally may be related to trauma, infection or iatrogenic techniques during cardiac surgery, myocardial biopsy or coronary angioplasty [4]. The incidence of CAF was reported to be high in transplant patients [5, 6]. Although majority of patients are asymptomatic, some may present with symptoms of dyspnea on exertion, increased fatigability, angina, palpitations, or signs of exercise-related coronary insufficiency. With increasing age, symptoms begin to appear, and the incidence of complications rises. [4]. The present study investigates incidence, clinical and angiographical characteristic of patients with solitary CAFs in all-comer population of Polish patients undergoing coronary angiography (CAG) in 10-year period in a large tertiary referral center in Poland.

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**Table 1. Angiographical characteristic of CAFs**

**Angiographical characteristic of CAFs with emphasis on drainage site**

| Artery of origin | Drainage site | | Number |
|---|---|---|---|
| Left coronary artery (15) | Heart chamber (3) | Right atrium | 0 |
| | | Right ventricle | 1 |
| | | Left atrium | 0 |
| | | Left ventricle | 2 |
| | Vessel (12) | Pulmonary artery | 12 |
| Right coronary artery (11) | Heart chamber (5) | Right atrium | 5 |
| | | Right ventricle | 0 |
| | | Left atrium | 0 |
| | | Left ventricle | 0 |
| | Vessel (6) | Pulmonary artery | 6 |

**Detailed angiographical characteristic of CAFs**

| Artery of origin | | Drainage site | Number |
|---|---|---|---|
| Left coronary artery | Left main coronary artery | Left ventricle | 1 |
| | Left anterior descending artery | Left ventricle | 1 |
| | | Right ventricle | 1 |
| | | Pulmonary artery | 11 |
| | Circumflex artery | Pulmonary artery | 1 |
| Right coronary artery Pulmonary artery | | Right atrium | 5 |
| | | | 6 |

# Methods

We investigated medical records of all adult patients who underwent CAG at the Jagiellonian University Department of Cardiac and Vascular Diseases, John Paul II Hospital, Kraków, Poland in 10-year period, from 1st July 2009 to 30th June 2019. Patients with CAF recognized during CAG were selected for future analysis. Anonymized angiograms were evaluated by the experienced core lab analyst.

The CAFs were described depending on the origin and termination. Unilateral CAF was recognized if one coronary artery was the origin of CAF, bilateral if two, and multilateral if all three. Multiple fistulas were recognized when single patient was affected by two or more CAFs. Coronary artery-ventricular MMFs were recorded but were no object for further analysis.

Indication for CAG was recorded in all patients of the study group. Clinical symptoms, comorbidities and past medical history were analyzed. Coronary artery disease (CAD) was defined by presence of at least 50% diameter stenosis in any major epicardial vessel or presence of coronary stent or bypass graft (CABG).

# Results

26 CAFs were found in 22 (0.17%) out of 12,757 patients who underwent CAG for any reason in that period. The age of the patients with CAF ranged from 20 to 81 years, with a mean age of 63 ±14 years. 12 (54.55%) patients were female, with an age range from 46 to 79 and a mean of 65 ±12 years, and 10 (45.45%) patients were male with an age range from 20 to 81 and a mean of 59 ±18 years. 20 MFFs were present in 16 (0.13%) out of all patients who underwent CAG, 15 draining into left ventricle (LV) and 5 into right ventricle (RV) (3:1).

Indication for CAG varied among study group. 11 (50%) patients underwent CAG due to chronic coronary syndromes (CCS), 2 (9.09%) in acute coronary syndrome (ACS), 4 (18.18%) in the course pulmonary hypertension, 3 (13.64%) due to structural heart defect, 1 (4.54%) exacerbation of heart failure and 1 (4.54%) before planned surgical procedure.

Table 1 presents CAFs grouped by artery of origin and drainage site. All CAFs were unilateral. Left coronary artery (LCA) gave origin to 57.69% of fistulas, of those left anterior descending (LAD) was 'parent' artery of 50%, left main coronary artery (LMCA) or circumflex (Cx) of 3.85% CAFs. Right coronary artery (RCA) gave origin to 42.31% of CAFs. Pulmonary artery (PA) was the most common drainage site (69.23%), followed by right atrium (RA) (19.23%), RV (7.69%) and LV (3.85%). 4 (18.18%) patients had multiple CAFs, in all these patients one CAF originated from LAD, while the other one from RCA. All were draining to PA.

Table 2 presents symptoms, comorbidities and past medical history of each referral diagnosis and the whole study group. 11 (50%) patients had CAD, of those stents in coronary arteries were present in 4 (18.18%) and CABG in 2 (9.09%) patients (1 patient had both).

1 (4.55%) patient had history of open-heart surgery due to atrial myxoma removal. Congenital heart disease was present in 4 (18.18%) patients, 2 atrial septal defect and 2 patent foramen ovale.

Among 11 patients referred for CAG for CCS indication, CAD was detected in 5. Of 4 patients with marvelous myocardial ischemia in the fistulas index territory, interventional management was performed in 3 (2 no CAD, 2 CAD; 2 – surgical closure, 1 – coil occlusion).

# Discussion

The principal finding of this study is determination of the prevalence of solitary CAF in a sample of population of Polish patients undergoing CAG that was 0.17%. Further, LCA is the most common artery of origin and PA is the most common drainage site. Moreover, the incidence of MMF was 0.13%.

Due to high resolution and dynamic imagining, CAG remains the best diagnostic tool for CAFs diagnosis [1]. Most CAFs are

**Table 2. Clinical symptoms, comorbidities and past medical history of each CAG indication group and the whole study population**

| | Stable unaccompanied CAD (n=11) | Pulmonary hypertension (n=4) | Structural heart defect (n=3) | Acute coronary syndrome (n=2) | Exacerbation of heart failure (n=1) | Before planned procedure (n=1) | Overall (n=22) |
|---|---|---|---|---|---|---|---|
| Arterial hypertension | 10 (90.91%) | 3 (75%) | 1 (33.33%) | 2 (100%) | - | 1 (100%) | 17 (77.27%) |
| Chest pain or dyspnoea | 8 (72.73%) | 2 (50%) | 2 (67.67%) | 2 (100%) | 1 (100%) | - | 15 (68.18%) |
| Chest pain | 6 (54.55%) | - | 1 (33.33%) | 2 (100%) | - | - | 9 (40.91%) |
| Dyspnoea | 4 (36.36%) | 2 (50%) | 2 (66.67%) | - | 1 (100%) | - | 9 (40.91%) |
| Dyslipidemia | 9 (81.82%) | 1 (25%) | 1 (33.33%) | 1 (50%) | - | 1 (100%) | 13 (59.09%) |
| CAD | 5 (45.45%) | 1 (25%) | 2 (67.67%) | 2 (100%) | 1 (100%) | - | 11 (50%) |
| Moderate/severe valvular hear disease | 3 (27.27%) | 3 (75%) | 2 (66.67%) | - | 1 (100%) | - | 9 (40.91%) |
| Type 2 diabetes | 4 (36.36%) | - | 1 (33.33%) | 2 (100%) | - | 1 (100%) | 8 (36.36%) |
| Any arrythmia | 4 (36.36%) | 2 (50%) | 1 (33.33%) | - | 1 (100%) | - | 8 (36.36%) |
| Atrial fibrillation | 3 (27.27%) | 2 (50%) | 1 (33.33%) | - | 1 (100%) | - | 7 (31.82%) |
| Other arrythmia | 2 (18.18%) | - | - | - | - | - | 2 (9.09%) |
| Myocardial infarction | 2 (18.18%) | 1 (25%) | 1 (33.33%) | 1 (50%) | - | - | 5 (22.73%) |
| Pulmonary hypertension | 1 (9.09%) | 3 (75%) | 1 (33.33%) | - | - | - | 5 (22.73%) |
| Palpitation | 3 (27.27%) | - | 1 (33.33%) | - | - | - | 4 (18.18%) |
| Congenital heart defect | 1 (9.09%) | 1 (25%) | 2 (66.67%) | - | - | - | 4 (18.18%) |
| Stroke | - | 1 (25%) | 1 (33.33%) | 1 (50%) | - | 1 (100%) | 4 (18.18%) |
| Vertigo | 3 (27.27%) | - | - | - | - | - | 3 (13.64%) |
| Heart failure | 1 (9.09%) | - | 1 (33.33%) | - | 1 (100%) | - | 3 (13.64%) |
| Previous cardiac surgery | 1 (9.09%) | 1 (25%) | - | - | 1 (100%) | - | 3 (13.64%) |
| Pulmonary embolism | 1 (9.09%) | 2 (25%) | - | - | - | - | 2 (9.09%) |
| Fainting | - | 1 (25%) | - | - | - | - | 1 (4.55%) |

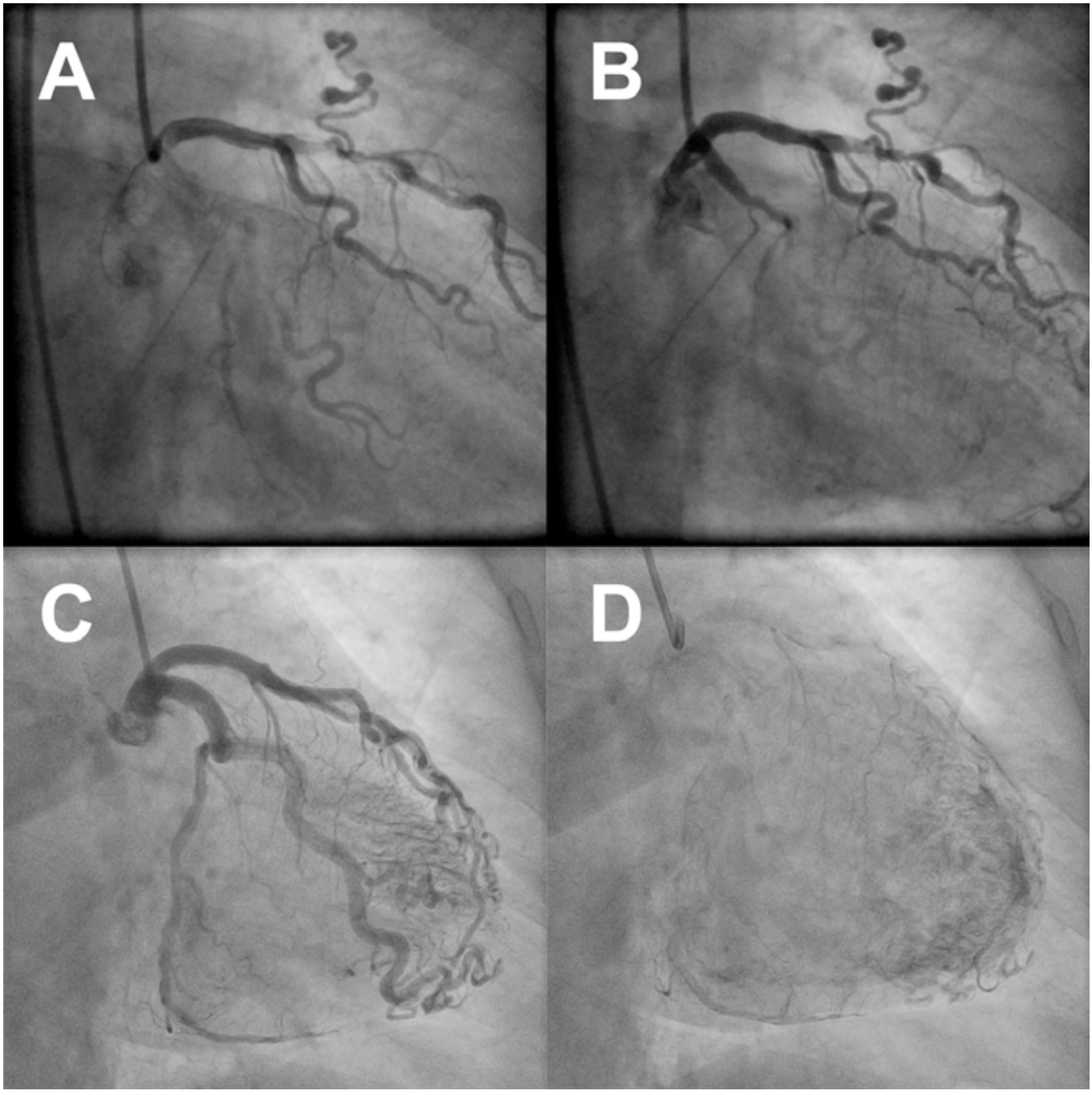


**Figure 1.** CAFs diagnosed during CAG. A, B – Solitary CAF originating from LAD and draining into PA. C, D – MMFs originating from LAD and draining into LV

diagnosed incidentally during cardiac catheterization [3]. Prevalence of solitary CAFs in overall population is estimated to be about 0.002% [1], while in patients undergoing CAG it fluctuates around 0.1–0.2% [2, 7–11]. In our study, the incidence was 0.17%. Our result is consistent with the outcome of the recent Dutch Registry (DR) study [2]. However, in another study based on computed tomography angiography the incidence was found to be as high as 0.9% [12]. There is no striking difference in CAF distribution among sexes [3] (54.45% females and 45.55% males in the current study).

Single fistulas are overwhelmingly more common, occurring in up to 90% of cases [13]. Multiple fistulas constitute in about 10% and both coronary arteries are affected in 5%. [4]. In our study, 18.18% of patients had multiple CAFs and in all of them both LCA and RCA were affected.

Anatomic characteristics of CAFs vary between studies. Earlier studies report RCA as the most frequent site of origin, while more recent suggest that LCA is more commonly affected [3]. Accordingly, we found that LCA was the 'parent' artery of 57.69% CAFs.

Our result is once again consistent with DR [2], in which LCA was origin of 66.67% CAFs.

PA constitutes the most common drainage site (69.23%). The term "coronary-cameral fistula" (31%) refers to a communication between the coronary arteries and a cardiac chamber. Right side of the heart is reported to be more frequently affected than the left (23.07% vs 7.69%, respectively) [3].

Majority of patients suffered from chest pain or dyspnoea, had HA, dyslipidemia, and half had CAD. 41% of patients had moderate or severe valvular heart disease. Every third patient had diabetes mellitus type 2 and arrhythmia. A congenital heart defect is reported to be associated with about 20% of CAFs [1, 3], in our study they coexisted in 9%. Our study reports high prevalence of pulmonary hypertension (22.73%) among patients with CAF (only 4% of patients were affected in DR) [2], which most probably represents a selection bias due to the fact, that our center is the only referral site for pulmonary hypertension in a large area.

Clinical presentation of CAFs depends on a number of factors including age of the patients [4] and is often clouded, because of the presence of other underlying conditions such as CAD (50%) or moderate/severe valvular disease (41%). The majority of patients are asymptomatic, the lesion is detected as an incidental finding during CAG for unrelated symptoms, however a soft continues murmur, that tends to be crescendo-decrescendo may be present during physical examination [3]. Chest pain and dyspnea are the most common presenting symptoms [3, 14]. In our study, only 4 solitary CAFs were remarked symptomatic.

## Conclusion

In our sample the incidence of solitary CAF in all-comer population of Polish patients undergoing CAG was 0.17%. LCA was the most common artery of origin and PA was the most frequent drainage site. The indication for CAG varied among patients. Majority of patients suffered from chest pain or dyspnea, had HA, dyslipidemia, and half had CAD. Solitary CAF rarely requires interventional management. Moreover, the incidence of MMF was 0.13%.

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