# Congenital Coronary Artery Fistulas, a Polish single-center computed tomographic registry

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***Related article***
by Michałowska et al.

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DOI: 10.33963/v.kp.98517

**Received:**
December 10, 2023

**Accepted:**
December 10, 2023

**Early publication date:**
December 12, 2023

Congenital coronary artery fistulas (CAF) are aberrant connections between the coronary arteries and contiguous structures like non-coronary vessels or cardiac chambers. In patients undergoing coronary computed tomography (CT) angiography, the prevalence of CAFs ranges from 0.2 to 0.9% [1–3].

CAF is often found incidentally during a study for other reasons [4], however depending on the anatomy (donor vessel, recipient vessel or chamber, size and length) and patient characteristics (concomitant heart disease), it may cause symptoms and lead to life-threatening complications like acute ischemia, congestive heart failure, pulmonary hypertension, endarteritis and rupture [5, 6]. Usually small incidentally found CAFs are asymptomatic and do not require intervention [1, 4, 7]. Some CAF may remain clinically silent for decades, however once it becomes hemodynamic significant (myocardial ischemia through a steal phenomenon, left ventricle volume overload or significant left to right shunt leading to congestive heart failure or pulmonary hypertension) treatment must be considered [8]. Percutaneous treatment is currently the mainstay of therapy when technically feasible and there are no other indications for heart surgery [4, 8]. Patients that underwent intervention should be followed up more frequently early after closure to recognize the possible recurrence of the fistula, persistent dilatation of the coronary artery, thrombus formation, calcification, arrhythmias, and myocardial infarction [8]. Regarding medical therapy, antiplatelet therapy is recommended empirically to prevent thrombosis when there is coronary artery or fistula dilatation, anti-anginal drugs should be used to alleviate angina, and most authors recommend endocarditis prophylaxis.

In this issue of the *Polish Heart Journal*, Michałowska et al. [9] reported the prevalence, anatomic characteristics, and clinical significance of congenital CAFs, of 42 patients diagnosed as having at least one CAF among 39 066 unselected adults, that had a cardiac CT assessment over a period of 12 year in a tertiary single center. It is the largest, CT diagnosed congenital CAF series, ever reported. Overall, the findings regarding anatomy and clinical manifestations are in line with other international series, however some points deserve a thoughtful reflection. The prevalence of CAF in the present report was 0.11%, which is within the range of invasive angiography registries, but bellow most of the largest CT series (0.19%–0.9%) [1–3]. This may be due to: 1) the retrospective nature of the study and the methodology that the authors used to identify CAF cases; 2) exclusion of CAFs suspected to be acquired and non-diagnostic CTs, (nevertheless, it accounted only for 10 patients, and the prevalence would have been otherwise 0.13%, still bellow most of the CT series); and 3) finally a possible difference in CAFs prevalence among distinct populations, with higher prevalence in eastern population, where the largest CT reports were derived from [1–3]. There was an association between CAF size and drainage site (namely, low pressure right sided vessels/chambers — superior vena cava, right atrium, coronary sinus) with fistula calcification, infective endocarditis, pulmonary hypertension, and clinical significance (as defined by the authors). Clinical significant

CAF were more common in younger and male patients. This results should be interpreted with caution, given the small sample size, prone to type II error. Also, mortality at a median follow up of 22.5 months was very high, 16.7% (7/42) in the overall CAF population, 43% (3/7) in patients with clinically significant CAFs, and 11% (4/35) in patients with non-significant CAF, the mean ages of these groups were 57.5, 47.1 and 59.5 years old, respectively.

The present results add up to the evidence that already exists, reinforcing that CAF is a rare entity, and clinically significant CAF is even rarer; in adults the diagnosis is usually made "incidentally" by CT as an investigation for other causes of chest pain, and heart failure; and that anatomical factors influence the clinical presentation. In addition the current findings, raise some hypothesis, namely: 1) the possibility that CAF prevalence may vary in different populations; 2) that CAFs draining into right sided structures may increase in size and progress to larger shunts with clinical repercussion at younger ages, and 3) that mortality in CAF patients is high, even in patients with non-clinically significant CAFs. This issues deserve further investigation, to clarify if there are differences in the prevalence and presentation in different populations; if CAFs draining into right sided structures deserve a different approach, namely and more preemptive follow up and treatment; and if CAF presence is associated with higher risk of mortality. Furthermore, the role of other diagnostic modalities, namely functional cardiac magnetic resonance and invasive hemodynamic assessment, in establishing a mechanistic correlation between the anatomical characteristics and clinical repercussion, needs to be clarified. Finally, given the high anatomical and clinical variability, therapy should always be individually tailored, however it is not known when is the sweet spot for intervention nor what is the best therapeutic option for incidentally found CAFs. This kind of reports highlight the need for larger prospective multicenter international registries. An effort should be made to incite physicians collaboration in reporting the natural history and management of all types of CAFs, so that a more comprehensive characterization of this rare and highly heterogeneous condition can be made.

## *Article information*

**Conflict of interest:** None declared.

**Funding:** None.



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