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Congenital coronary artery-left ventricle direct micro-fistulas may cause effort angina and positive stress tests in Western adults

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Coronary artery anomalies (CAAs) occur in 0.64 % to 5.6% of patients undergoing coronary angiography.[1–4] Most pathophysiology and clinical histories involving CCAs have been fully clarified in the past 30 years. Isolated congenital coronary artery-left ventricle direct microfistulas (CVmF) have been recently described,[5,6] but the clinical significance and anatomical characteristics associated with this rare type of CAA in a western population are still unclear. The aim of our retrospective, single center study is to assess the clinical significance and anatomical features associated with this anomaly in a western population.

We retrospectively reviewed the medical records of 17,892 patients (mean age 74.8 ± 22.3 years, 56.8% males) who underwent coronary angiography in our secondary referral center during the past 12 years from July 2003 to July 2015. Patients were analyzed for medical history, classical risk factors, medical instrumental examinations prior to coronary angiography. Coronary angiographies were reviewed by two independent and blinded observers with < 10 years expertise in coronary angiography (inter-observer agreement was 99.8%). The population with the targeted anomaly was compared with a population with normal coronary arteries and no epicardial coronary atherosclerosis by coronary angiography during a period of three months in 2015. Normal coronary artery was define as a vessel with < 10% of lumen narrowing.

Chi-square, Fisher exact test and Student-t tests were used to compare frequencies and continuous variables between the groups. Statistical analysis was performed using a statistical software package (SAS for Windows, version 8.2; SAS Institute; Cary, NC USA). A probability value of \( P < 0.05 \) was considered to be statistically significant.

Sixty-five patients (male/female: 34/31) had angiographic appearance of micro-fistula between the coronary arteries and left ventricle. Indications prior to coronary angiography were recurrent effort angina in 56/65, effort dyspnea in 5/65 patients associated in 52/65 patients to positive ergometric stress test (37/65 patients) or congruent nuclear stress test (15/65 patients). Coronary angiography was performed in four patients for acute coronary syndrome non ST–elevated and mild troponin increase (> 2-time normal value). In 43/65 patients, preoperative echocardiography showed a mild hypokinesis of the anterior wall (56.9%) or apex (9.1%).

The majority of patients (58/65, 89.2%) showed a CVmF invariably appearing as a net of small vessels filling the left ventricle with no recognizable single vessel. CVmF arising from the left anterior descending (LAD) in 100% of cases in the mid-distal portion of the LAD, whereas six patients (9.2%) had a CVmF arising from the right coronary artery (RCA), and 100% in the distal portion of RCA. Two patients had CVmF arising from both LAD and RCA (3.1%). Risk profile and cardiac co-morbidities did not differ significantly from a control population with normal coronary artery and no coronary atherosclerosis (Table 1 & 2).

Table 1. Demographic and clinical features of patients with CVmF.

|                      | CVmF, \( n = 65 \) | Control, \( n = 60 \) | \( P \) |
|----------------------|-------------------|----------------------|-------|
| Age, yrs             | 70.4 ± 11.8       | 78.4 ± 7.8           | < 0.03|
| Male/Female          | 34/31             | 28/32                | NS    |
| High blood hypertension| 51 (78.4%)       | 49 (81.6%)           | NS    |
| Dyslipidemia         | 32 (49.2%)        | 29 (48.3%)           | NS    |
| Smoking habits       | 32 (49.2%)        | 30 (50%)             | NS    |
| Diabetes             | 10 (15.3%)        | 7 (11.6%)            | NS    |

Data are presented as mean ± SD or n (%). CVmF: coronary artery-left ventricle direct microfistula; NS: not significant.

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Table 2. Related cardiac comorbidities comparison with normal coronary artery population.

|                                | CVmF, n = 65 | Control, n = 60 | P       |
|--------------------------------|--------------|-----------------|---------|
| Superimposed coronary atherosclerosis | 2/65 (3.1)   | 0/60 (0)        | NS      |
| Cardiac valve disease           | 12/65 (18.4%)| 11/60 (18.2%)   | NS      |
| Dilated cardiomyopathy          | 15/65 (23.1%)| 16/60 (26.6%)   | NS      |
| Congenital heart disease (PFO, ASD) | 2/65 (3.1%)  | 0/60 (0)        | NS      |

Data are presented as n (%). ASD: atrial septal defect; CVmF: coronary artery-left ventricle direct microfistula; NS: not significant, PFO: patent foramen ovale.

Our study suggests that at least in a western population, isolated congenital CVmF is involved in the vast majority of cases of the mid-distal segments of LAD. Western patients had a relatively young age, the same prevalence of classical risk factors of the control population, and usually showed a benign presentation with symptoms of effort angina and signs of reduced coronary reserve during an ergometric or nuclear stress test.

Usually most of the congenital fistulas originate from the first segment of the involved coronary artery, and those located in the middle or distal segment of a coronary artery usually drain into the ventricular chambers. CVmF seemed to follow the same pattern, and as matter of fact no direct micro-fistula between the coronary vessel and atrial chamber have been detected during the study. Except for the only Western report of this anomaly found in Claeys, et al.[7] in the early 90s, congenital CVmF has been well described by eastern authors: Doğan, et al.[5] that found a prevalence of 0.11% in the Turkish population with chest pain presentation, dividing them in: (1) direct filling of the heart cavity during selective coronary injection without interposing “capillary” phase or venous filling, and (2) visualization of small vessels interposed between the epicardial coronary vessels and the heart cavity and emptying into the heart. We believe that only the second subtype can effectively described as micro-fistula, and accordingly in our series the appearance was in 100% of case a net of small vessels.

Figure 1. Typical angiographic appearance of CVmF arising from the LAD during coronary angiography. (A&B): opacification of the CVmF (arrow); (C&D): spontaneous partial left ventriculography (arrow). CVmF: coronary artery-left ventricle direct microfistula; LAD: left anterior descending.
filling the left ventricle. Our findings are consistent with the study of Chiu, et al.\cite{8} in far eastern literature who found in 26 patients out of more than 20,000 coronary angiography, a coronary-left ventricle multiple fistula with the same appearance found in our patients.

The peculiar angiographic appearance might explain both effort symptoms and stress tests as mild positivity: CVmF is likely to act by means of a mild diversion of the blood flow stream from the myocardium to the left ventricle under exercise or effort.

Although a more structured, long term, follow up study has not been conducted for the multiple confounding factors that are likely to impact the natural history of this anomaly, such as the development of vascular atherosclerosis or other morbidities, CVmF seemed to be a benign entity quite rarely discovered during coronary angiography.

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