

CLINICAL IMAGE

Abnormal left and right coronary-to-aortic arch and main and right pulmonary artery fistulas in a 63-year-old patient

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A 63-year-old woman was admitted to the hospital with typical chest pain at rest and 1-mm ST depression in leads V₃ through V₆. The symptoms were relieved by intravenous administration of nitroglycerine. The results of the troponin test were negative. Prodromal symptoms occurred 2 weeks earlier. Mild limitation of exercise tolerance was

present since childhood. A physical examination was unremarkable except for 2/6 left parasternal murmur. An electrocardiogram showed sinus rhythm and incomplete right bundle branch block (RBBB). Urgent coronary angiography showed smooth nonobstructed epicardial arteries and the presence of an arterial small vessel network

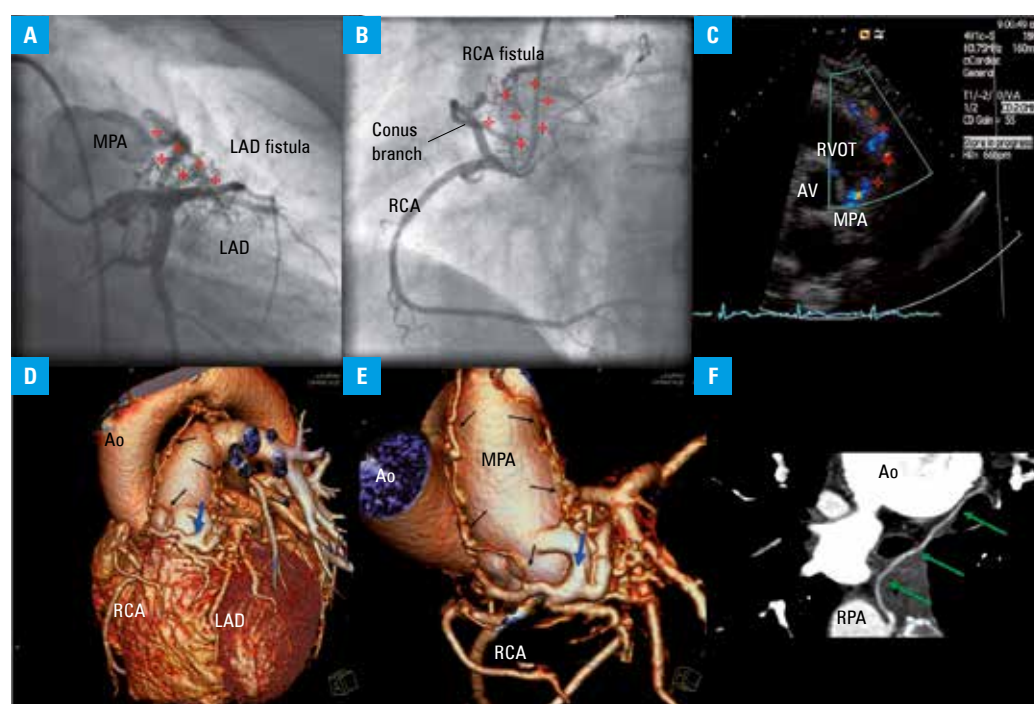


FIGURE 1 **A** – left anterior descending (LAD) artery-to-main pulmonary artery (MPA) fistula on coronary angiography (red asterisks); **B** – right coronary artery (RCA)-to-MPA fistula on coronary angiography (red asterisks); **C** – transthoracic echocardiography: abnormal flow with a mosaic pattern adjacent to the right ventricular outflow tract (RVOT) entering the MPA; **D, E,** and **F** – computed tomography: abnormal vessel (blue arrows) originating from the MPA connecting with the LAD and RCA, right pulmonary artery (green arrows) and 2 abnormal arteries arising from the aortic (Ao) arch (black arrows)
Abbreviations: AV – aortic valve

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Received: May 27, 2013.

Revision accepted: June 3, 2013.

Conflict of interest: none declared.

Pol Arch Med Wewn. 2013;

123 (9): 498-499

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Kraków 2013

connecting the proximal left anterior descending (LAD) artery and the main pulmonary artery (MPA; **FIGURE 1A**) as well as the proximal right coronary artery (RCA) and the MPA (**FIGURE 1B**). Mean pulmonary artery pressure was normal (22 mmHg) and systemic-to-pulmonary shunt (Qp:Qs) was 1.4 on right heart catheterization. On echocardiography, abnormal flow with a mosaic pattern was observed adjacent to the right ventricular outflow tract and entering the MPA (**FIGURE 1C**). Right and left ventricular function and dimensions were normal. A saline contrast study (left and right median cubital vein) did not show any abnormalities. Computed tomography revealed an abnormal vessel (**FIGURE 1DE**), originating from the MPA and giving rise to a small vessel network connecting with the LAD and RCA in the right pulmonary artery (**FIGURE 1F**), and another 2 abnormal arteries arising from the aortic arch (**FIGURE 1DE**). The MPA was enlarged (31 mm).

A coronary artery fistula is defined as a communication between a coronary artery and either a chamber of the heart (coronary-cameral fistula)¹ or any segment of the systemic or pulmonary circulation. Most fistulas are congenital. In a series of over 126,000 angiograms, congenital coronary-to-MPA fistulas were found in 0.17%.² Bicornary fistulas are rare (0.004% to 0.001% of the cases).² Small fistulas are often an incidental finding; on the other hand, atypical symptoms are common. An exercise test is sometimes of limited value³; in selected patients, scintigraphy can be used to diagnose ischemia and quantify myocardium at risk.⁴ Possible, but uncommon, complications are endocarditis, myocardial infarction, sudden death, and fistula rupture. Surgical or transcatheter closure is usually indicated in patients with angina, heart failure, large shunt, or pulmonary hypertension. Patients with large fistulas, multiple openings, or significant aneurysms may not be candidates for transcatheter closure.⁵

The presented case is unique in terms of anatomy; the multiple connections with both left and right coronary arteries, main and right pulmonary artery, and aortic arch through the network of small vessel is a challenge for surgical or interventional treatment. The patient had new-onset angina and mild-to-moderate dyspnea (class II of the New York Heart Association Functional Classification). The systemic-to-pulmonary flow at rest was borderline. The pulmonary artery pressure and RV function were normal, RBBB was incomplete, and the MPA was enlarged. There were no malignant arrhythmias in Holter monitoring. The patient was started on diltiazem (120 mg orally) and an exercise test showed satisfactory exercise tolerance (6.8 metabolic equivalents and 70% of the maximal heart rate) without anginal symptoms. Balancing operative risk and projected immediate benefits of surgical intervention, conservative treatment was deemed optimal.

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