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**Maarten Van Caenegem, Hans Vandekerckhove**

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**Case Report:** We report two cases, one between the right coronary artery (RCA) and the pulmonary artery, whereas the other between the circumflex artery (RCX) and the pulmonary artery. We describe our diagnostic methodology and analyze literature on the epidemiology, the diagnostic workout and the treatment possibilities.

**Conclusion:** Despite the role of non-invasive imaging for diagnosis and identification of the location of CAF including the origin and insertion of the recipient vessel, cardiac catheterization and coronary angiography remain the preliminary diagnostic tools for the precise allocation of coronary anatomy, for assessment of its hemodynamic importance, and to show other structural abnormalities.

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## INTRODUCTION

Coronary artery fistulae (CAF) are anomalous acquired or congenital terminations of the coronary arteries into other vascular structures, such as a cardiac chamber, vena cava, the pulmonary artery, or pulmonary veins. Most of the coronary anomalies are incidental findings during angiographic evaluation for coronary vascular disorders. Coronary artery fistulae are present in 0.002% of the overall population and are documented in almost 0.25% of the patients undergoing coronary angiography [1]. Majority of these fistulas originate from the right coronary artery or the left anterior descending artery. Patients are frequently asymptomatic, but angina due to coronary steal phenomenon or myocardial infarction and dyspnea due to heart failure and endocarditis have been reported in some cases [2, 3]. The management is complex, and recommendations are founded on anecdotal cases of very small retrospective series.

## CASE SERIES

### Case 1

A 64-year-old male presented to the hospital with dyspnea and angina during emotional stress. He was known with pulmonary sarcoïdosis in remission and obstructive sleep apnea syndrome. He did not take any medication. His general examination was unremarkable. An additional exercise test, however, was clinical suspicious with the reproduction of atypical thoracic complaints in the absence of electrocardiographic changes. Echocardiography showed a preserved left ventricular systolic function with absence of valvular disease. Coronary angiography confirmed slight coronary atheromatosis, however, there was a fistula between the right coronary artery and the pulmonary artery with the presence of minor pulmonary hypertension (Figure 1). A supplementary CT scan of the lungs identified agenesis of the right pulmonary artery with a dysplastic right lung. There was also collateral circulation form the iliac veins and vena cava inferior, an occlusion of the vena cava superior and an arteriovenous malformation of the right middle lobe originating from the right coronary artery (Figure 2). To exclude a vascular steal phenomenon form the right coronary artery, we organized a Thallium stress test, which ruled out stress induced ischemia. A pulmonary function test illustrated an unchanged restrictive pattern due to the dysplastic right lung and his obesity. Concerning the etiology of the absence of the right pulmonary artery and the presence of a dysplastic right lung, we could not differentiate between congenital versus acquired phenomenon (possibly due to external compression of calcified lymph nodes by sarcoidosis at young age). Considering the absence of stress induced ischemia, hemoptysis or pulmonary infections, we suggested a conservative approach with annual evaluation of progressive pulmonary hypertension. A calcium channel blocker was initiated experimentally with full symptom relief.

### Case 2

A 46-year-old male presented to the emergency room complaining of dyspnea, slight hemoptysis and atypical chest pain. Physical examination was unremarkable. Electrocardiography showed a normal sinus rhythm, normal QRS morphology and normal repolarization. Transthoracic echocardiography illustrated a normal left and right ventricular morphology with preserved left ventricular systolic function. Pulmonary artery pressure was within normal range. Based on his moderate cardiac risk profile and an inconclusive exercise test, a cardiac catheterization was planned. It revealed non-obstructive coronary artery disease and a large coronary pulmonary fistula communicating from the right circumflex (RCX) coronary artery to a network of collateral circulation to the right lung. An interruption of the right pulmonary artery and vascularization of the right lung by aberrant intercostal arteries and the right mammary artery (Figure 3) are illustrated as well by a supplementary angiography of the pulmonary arteries with the presence of a unique left pulmonary artery. A supplementary cardiac MRI could not detect any other site of an intracardiac of extracardiac shunt. A vascular steal phenomenon was ruled out by a normal Thallium stress test. Considering the absence of pulmonary hypertension, exercise induced ischemia or arrhythmia, a conservative approach was taken.

## DISCUSSION

Coronary artery fistulae are uncommon congenital abnormalities of the coronary arteries or more seldom

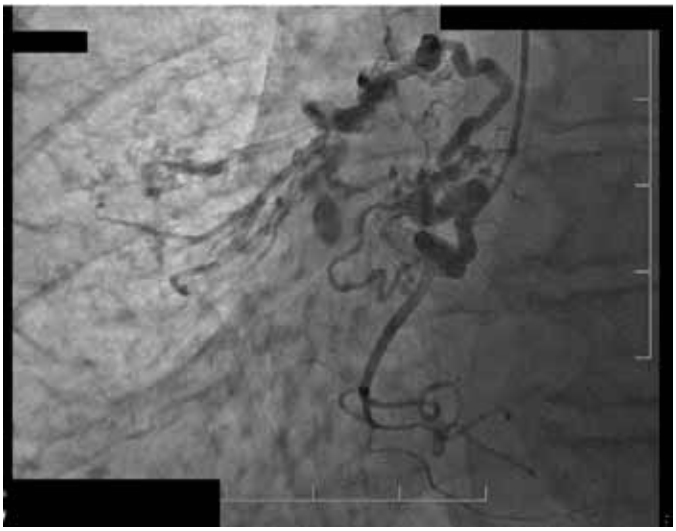

Figure 1: Cardiac catheterization: A coronary pulmonary fistula originating from the proximal right coronary artery (Case 1).

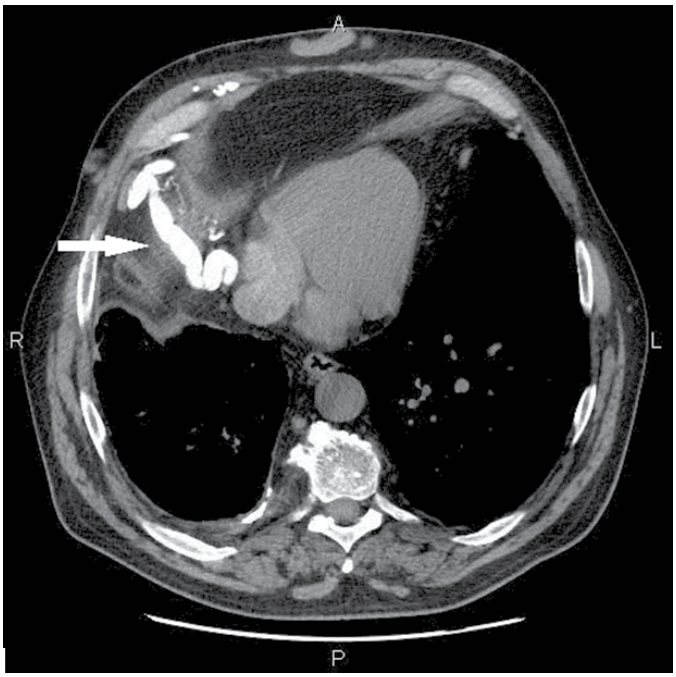


Figure 2: An arteriovenous malformation of the right middle lobe originating from the right coronary artery (Case 1).

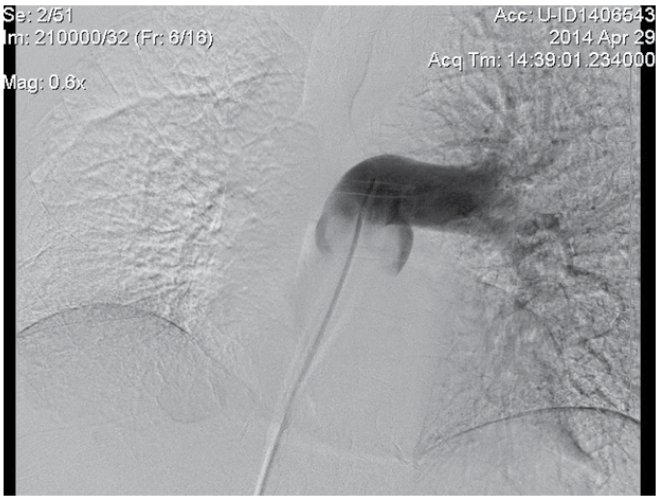


Figure 3: Agenesis of the right pulmonary artery by pulmonary angiography (Case 2).

acquired conditions and can occur iatrogenic after cardiac surgery like coronary artery bypass surgery or cardiac transplantation, or as a complication of coronary angioplasty and myocardial biopsy [4].

These fistulas originate more frequently from the right coronary artery and commonly shunt into one of the right heart chambers. Clinical presentation is dependent on the magnitude of the fistulous connection and could seldom result in a significant left-to-right shunt with congestive heart failure and cardiomegaly in infancy if a large fistula is present [5].

Nevertheless the growing potential of several non-invasive techniques like transthoracic and transesophageal echocardiography or cardiac imaging with magnetic resonance or computed tomography for identification and follow-up, coronary angiography still is the gold standard at present because of its accuracy in defining the artery of origin, as well as the recipient vascular structure.

Coronary artery fistulae cause myocardial ischemia in only in a small number of patients [6]. In planning therapy, evaluation of the hemodynamic importance of the fistulae is crucial. Next to a stress electrocardiography, a stress/rest 99mTc sestamibi single photon emission tomography is trustworthy for assessing the functionality of the anomalies detected by coronary angiography.

The natural history of CAF is unpredictable within one reported case an uncommon spontaneous closure due to spontaneous thrombosis [7]. For that reason, there is still some controversy about the management and follow-up, which is generally based on small retrospective series or anecdotal cases.

Antiplatelet therapy with at least one antiplatelet agent is recommended, especially in patients with distal coronary artery fistulas and abnormally dilated coronary arteries [8]. Based on limited experience and anecdotal cases bacterial endocarditis is a known complication and for that reason prophylactic precautions against subacute bacterial endocarditis are suggested.

The presence of heart failure and myocardial ischemia are predominant clinical symptoms to consider closure of the CAF. In addition to prevent occurrence of symptoms or complications, closure of CAF must also be considered in asymptomatic patients with high-flow shunting, especially in pediatric population [9]. Nevertheless, treatment of non-significant shunting in asymptomatic adult patients is still doubtful. There is no sufficient medical treatment for CAF. The choice between trans-catheter closure of the fistula and surgical intervention is still controversial. However, trans-catheter closure may be indicated if the anatomy is favorable (e.g. non-tortuous vessel) and the distal portion of the fistula is accessible with the closure device and should be narrow to avoid embolization to the drainage site. Catheter closure can be performed with a variety of techniques, including detachable balloons, stainless steel coils, regular and covered stents, and various chemicals. The main goal is to restore the myocardial perfusion by reduction in left to right shunt after occlusion of the treated vessel to the level of first branch. The basic surgical technique is ligation of the fistula and may be performed with or without cardiopulmonary bypass, when there is a simple and easily accessible fistula [10]. Results from the transcatheter and surgical literature show percutaneous closure were associated with lower procedural risk and therefore becomes the preferred method of treatment [11].

There was also reported that recanalization of the treated coronary fistulae can occur, and they suggested follow-up angiography or other imaging modality, like stress electrocardiography or cardiac MRI scan, to be performed annualy in the beginning of follow-up in these patients and decreasing the frequency in case of asymptomatic stable condition [12].

## CONCLUSION

Coronary artery fistulae (CAF) are exceptional, isolated abnormalities that are usually asymptomatic. However, certain forms are associated with myocardial ischemia, congestive heart failure, and sudden cardiac death. Identification of signs and symptoms should lead to supplementary testing, especially thorough initial evaluation of coronary artery anatomy using echocardiography to detect shunting, cardiac MRI scan or coronary computed tomography. However, coronary angiography remains the gold standard for diagnosis. Clinically significant CAF has to be considered for elective closure based on the current safety and efficacy of both transcatheter and surgical closure of CAF. The type of intervention will depend on the anatomy and origin of the fistula, the magnitude of the vascular malformation and possible associated defects and certainly the experience of the surgeons and interventional cardiologists.

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## Author Contributions

Maarten Van Caenegem – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, revising it critically for important intellectual content, Final approval of the version to be published

Hans Vandekerckhove – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

## Guarantor

The corresponding author is the guarantor of submission.

## Conflict of Interest

Authors declare no conflict of interest.

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