

# Right Coronary Artery Fistulae Draining into Right Atrium: A Case Report

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## Abstract

We report the case of a 16-year-old young girl seen for precordial pain and stage II NYHA dyspnea. Clinical examination found a maximal systolodiastolic murmur in the left subclavicular. Lateral Q waves were noted on the electrocardiogram and echocardiography revealed a right coronary fistula draining into right atrium. CT scan confirmed the diagnosis. Patient was addressed to a specialized center for percutaneous closure of the fistula. In case of signs indicating coronary artery disease, the search for coronary anomalies should be systematic in children by using EKG and transthoracic echocardiography. In case of doubt, a second imaging such as the multislice Ct scan is performed. Coronary angiography has a dual diagnostic and therapeutic interest. The indications for treatment depend on the symptoms and the results of ischemia tests.

## Keywords

Right Coronary Fistula, Congenital Heart Disease, Sub-Saharan Africa

## 1. Introduction

Coronary fistulas are abnormal communications between a coronary artery and a heart chamber or large base vessel, thereby bypassing the myocardial capillary bed [1] [2]. These are rare defect and usually occur in isolation. These rare anomalies were first described by Krauss in 1965 [3]. The exact incidence is unknown. The majority of these fistulas are congenital, although they may occasionally be detected after cardiac surgery. Most are often well tolerated, however, complications can arise. Complications include “steal” from the adjacent myocardium, thrombosis and embolism, cardiac failure, atrial fibrilla-

tion, rupture, endocarditis/endarteritis and arrhythmias. Thrombosis within the fistula is rare but may cause acute myocardial infarction, paroxysmal atrial fibrillation and ventricular arrhythmias [2]. We report a case of coronary cardiac fistula discovered during the exploration of exertion dyspnea in a 16-year-old girl.

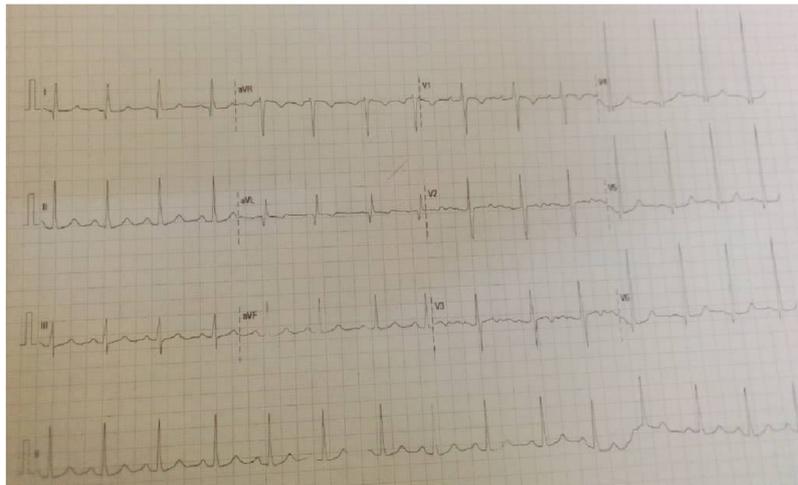
## 2. Observation

A 16-year-old girl consulted for precordial pain and NYHA stage II dyspnea.

On clinical examination, the blood pressure was 119/49 mmHg, the heart rate was 92 beats/min, the temperature was 36.5°C and the ambient air saturation was 99%. Cardiac auscultation finds a regular rhythm and systolo-diastolic murmur throughout the maximum precordium in the left subclavicular and along the left edge of the sternum. The vascular exam was normal. The peripheral pulses were well perceived. There were no signs of cardiac failure.

The electrocardiogram (EKG) showed a regular sinus rhythm at 83 cycles/min, a normal QRS axis and lateral qR waves (**Figure 1**).

The chest x-ray shows cardiomegaly with a cardio-thoracic index of 0.58 (**Figure 2**).

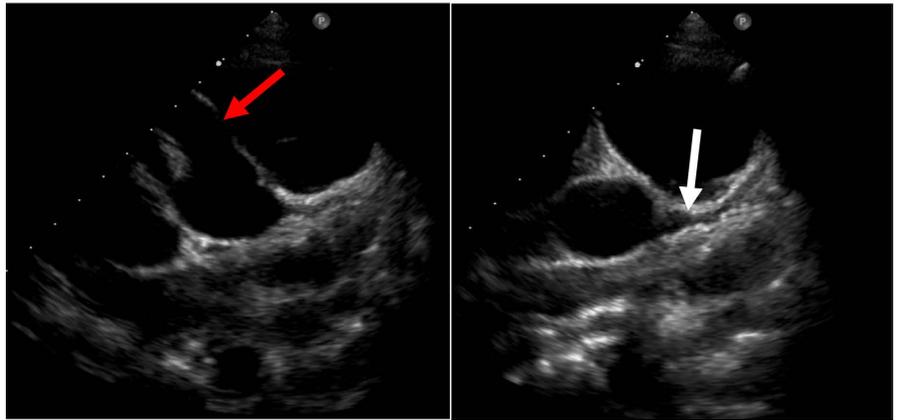


**Figure 1.** EKG showing abnormal Q waves in V5 V6 leads.

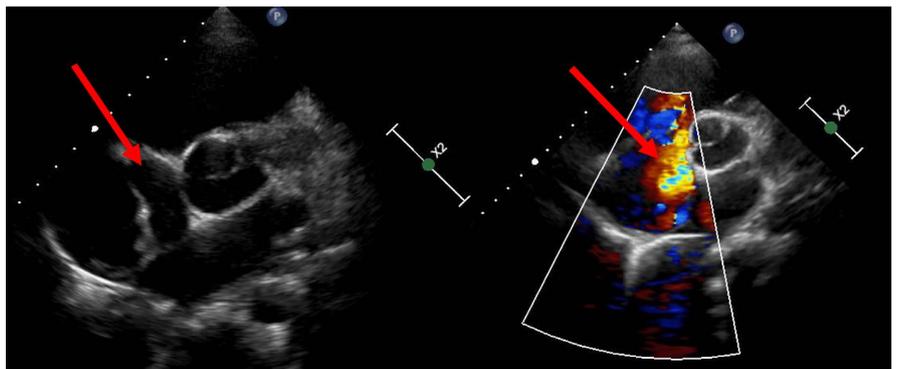


**Figure 2.** Cardiomegaly at the chest X ray.

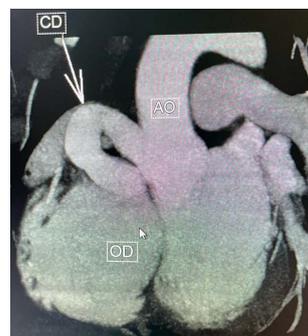
Transthoracic echocardiography shows normokinetic and dilated left ventricle with good systolic function (73% ejection fraction), dilatation of both atria, aneurysmal right coronary artery and normal left coronary artery (**Figure 3**). Color Doppler notes a high velocity systolo-diastolic flow between the right coronary artery and the right atrium, suggesting a coronaro-cameral fistula between the right coronary artery and the right atrium (**Figure 4**), there was no PAH. Multislice CT scan (**Figure 5**) confirm the diagnosis of right coronary



**Figure 3.** 2D transthoracic echocardiography, short axis view: Dilated right coronary artery (Red arrow) and normal left coronary artery (White arrow).



**Figure 4.** Transthoracic echocardiography, short axis view, 2D and color: Dilated right coronary artery draining in right atrium (Red arrow).



**Figure 5.** CT Scan right coronary artery fistula draining in right atrium (Ao = Aorta, OD = Right atrium, CD = Right coronary artery).

artery draining into the right atrium.

Patient was addressed to a specialized center for percutaneous closure of the fistula.

### 3. Discussion

Coronary fistula is an anomaly making a coronary artery communicated with a heart cavity or a large base vessel, thus bypassing the myocardial capillary bed [1] [2].

This malformation is rare and affects 0.002% of the population. It represents 0.4% of congenital heart disease [4].

Its incidence is estimated between 0.1% and 0.2% of all patients who had coronary angiography [5] [6].

In the congenital form, the fistulas are secondary to the persistence of intratrabecular sinusoids between the coronary arteries and the coronary sinus and vary greatly in terms of morphological appearance [7].

These fistulas are most often congenital and linked to the persistence of embryonic structures: intertrabecular and sinusoidal spaces [8].

They can also be acquired and occur during trauma, infection (Kawasaki disease) but can also be iatrogenic during heart surgery, myocardial biopsy or coronary angioplasty [9] [10].

Coronary fistulas can be isolated or associated with another congenital heart disease such as pulmonary atresias with intact septum, tetralogies of Fallot, interventricular or interatrial communications.

The most common origin is the right coronary artery (50% of cases), but the fistula can also come from the left coronary artery (42% of cases) or from its branches, or even from both coronaries (5% of cases) [11]. The circumflex artery is rarely involved [7] [11] [12]. Most often, the fistula results from a single communication (1 entry point), but several entry points are possible [7] [11] [12] [13]. Drainage from the artery is most often done directly into the right ventricle (41% of cases), the right atrium (26% of cases) or the pulmonary artery (17%) [6] [10] [12]. One of the less common drainage sites is the coronary sinus (1.6%) [7] [11]. More rarely, fistulas emptying into the left chambers of the heart are described.

The abnormal communication between a coronary artery and a venous structure causes a left-to-right shunt, the size of which depends on the size of the fistula and the pressure difference between the artery and the venous structure.

The clinical presentation of coronary fistulas depends mainly on the size of the left – right shunt [14]. The majority of patients are asymptomatic. They are usually referred for a heart murmur discovered incidentally.

In older patients, certain signs and sometimes clinical pictures may be seen: exertional dyspnea, exertional angina, arrhythmia, endocarditis, stroke [14] [15].

In case of wide left – right shunt, complications can occur: pulmonary arterial hypertension, congestive heart failure, aneurysmal dilation and rupture of the

fistula or ischemia, or even myocardial infarction due to the phenomenon of “arterial theft” [16].

The EKG and chest X-ray are routine tests that are not helpful in the diagnosis. However, in the event of a high-flow left-to-right shunt, the chest x-ray may show an increase in the cardiothoracic ratio and, on the ECG, signs of myocardial ischemia may be seen, reflecting a phenomenon of “arterial blood loss” [16].

Echocardiography, including transesophageal ultrasound, is a useful test in the diagnosis of large left-right shunt fistulas. Indeed, a dilated coronary artery as well as its drainage site can be easily detected especially in children in whom an optimal acoustic window can be obtained. However, the fistula path is difficult to see, especially when it is tortuous [17] [18] [19] [20].

In some cases a disturbance of the Doppler signal is demonstrated at the drainage site, especially the right ventricular one.

The cardiac CT scan provides morphological information: coronary circulation involved, caliber of the artery concerned, diagnosis of possible complications.

The gold standard is coronary angiography, which can confirm the diagnosis and study the exact anatomy of the coronary fistula by showing the affected artery, which is often dilated, the fistula path and the drainage site.

Surgical or percutaneous treatment aims to obliterate the fistula, allowing to preserve blood flow in the coronary arteries. Surgical treatment consists of ligation, by suture, of the fistula while the percutaneous technique allows closure of the fistula by embolization [7] [21]. The percutaneous method is preferred when the anatomy is favorable [7] [13]. For very tortuous lesions or in the presence of an aneurysm, surgery is still preferable [13].

The therapeutic indications remain controversial [11]. The onset of fistula-related symptoms and complications are indications for surgery [7] [12]. In asymptomatic patients, the invasive strategy should be cautious and should be considered in the event of a ratio of pulmonary flow to systemic flow (QP/QS) > 1.5 or in the presence of an aneurysm that could lead to thrombosis, rupture or obstruction [13]. For other patients, simple monitoring is recommended [7] [13].

#### 4. Conclusion

Coronary fistulas are rare but easily detected conditions thanks to advances in ultrasound imaging. Angiography and catheterization can confirm the diagnosis and are essential if percutaneous closure is planned in the patient. The therapeutic approach will depend on the anatomical situation. While the surgical approach remains valid, percutaneous occlusion currently appears to be the most appropriate therapeutic choice.

#### Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this

paper.

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