## **SAS Journal of Medicine**

Abbreviated Key Title: SAS J Med ISSN 2454-5112 Journal homepage: <u>https://saspublishers.com</u> **∂** OPEN ACCESS

Cardiology

# Fortuitous Discovery of a Coronarocameral Fistula in a Patient with a Acute Coronary Syndrome

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**DOI:** https://doi.org/10.36347/sasjm.2025.v11i01.015

| Received: 12.12.2024 | Accepted: 20.01.2025 | Published: 29.01.2025

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#### Abstract

Case Report

Coronary-cameral fistulae are rare congenital anomalous communications that may be single or multiple and occur between 1 or more coronary arteries and a cardiac chamber. We report the case of a 78-year-old patient, without modifiable cardiovascular risk factors admitted for an inferior Post-MI with incidental discovery of a coronarocameral fistula during angioplasty. Coronarocameral fistula is reported to be found in approximately 0.08–0.3% of unselected patients undergoing diagnostic coronary angiography. The imaging models allow the diagnosis to be evoked, but angiography remains the gold standard for the diagnosis. Surgical or interventional treatment is reserved only for symptomatic patients with large or multiple fistulas.

Keywords: Coronary-cameral fistulae; Angiography; Acute Coronary Syndrome.

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## **INTRODUCTION**

Coronary-cameral fistulae are rare congenital anomalous communications that may be single or multiple and occur between 1 or more coronary arteries and a cardiac chamber. Their discovery is often fortuitous and it can interest the right or left coronary arteries with a clear predominance for the right network. Given its rare character and also the silent character of its symptomatology, there is no well-established consensus for their diagnosis and management. We will try, through a clinical case and literature review, to shed light on this rather rare diagnosis and to codify the means of diagnosis and management.

## **CASE REPORT**

We report the case of a 78-year-old patient, without modifiable cardiovascular risk factors, operated

12 years ago for an inguinal hernia, admitted to the cardiovascular emergency unit for management of acute chest pain. The patient presented with typical infarction pain for 3 days, neglected by the patient, associated with NYHA stage III dyspnea. Given the persistence of the pain, the patient consulted 48 hours after the presumed start of the pain.

The examination on admission finds a conscious patient supporting the dorsal decubitus keeping a painful background, BP: 120/69mmhf, HR: 76, with a normal cardiovascular and pleuropulmonary examination.

The 18-lead ECG shows a Q wave in the anterosepto apical territory and in the inferior territory with negative T waves in the basal lateral septal and in the inferior territory.

**Citation:** Youssef Fihri, Zakaria Lahlafi, Driss Britel, Hicham Faliouni, Soumia Faid, Aatef Benyass, Zouhair Lakhal. Fortuitous Discovery of a Coronarocameral Fistula in a Patient with a Acute Coronary Syndrome. SAS J Med, 2025 Jan 11(1): 81-84.

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Figure 1: 18-lead ECG showing Q wave in the antero-septo apical territory and in the inferior territory with negative T waves in the basal lateral septal and in the inferior territory

Echocardiography demonstrates an aspect of ischemic heart disease in LV dysfunction with LVEF estimated at 30% and akinesia of the apex and adjacent segments, hypokinesia of the middle segment of the anterior, septal and lateral wall and moderate Mitral regurgitation.

The biological assessment finds a HB at 14.5g/dl, GB: 8500/mm3, Platelets: 145000/mm3, K+:

3.7mmol/l, Creat: 7mg/l, Troponin: 4679ng/ml which went to 15498 ng/ml.

The coronary artery showed a severe stenosis of the first marginal, a chronic occlusion of the middle segment of LAD and an acute occlusion of the middle right coronary with an incidental discovery of a coronarocameral fistula.



Figure 2: An RAO 30 degree incidence showing a coronarocameral fistula

The patient underwent angioplasty for middle right coronary artery with placement of an active stent.

#### DISCUSSION

Coronary-cameral fistulae are rare congenital anomalous communications that may be single or

multiple and occur between 1 or more coronary arteries and a cardiac chamber [1]. It is reported to be found in approximately 0.08–0.3% of unselected patients undergoing diagnostic coronary angiography [2].

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There are two types of coronarocameral fistulas: arterioluminal, where there is a direct and localized communication with the heart chamber, or arterio-sinusoidal, as in this case where the arterial blood communicates with the heart chambers via a sinusoidal network.

His anomaly more frequently arises from the right coronary system ( $\approx$ 55%) but can originate from the left side (35%) or bilaterally (5%) [1].

The most common cause of coronary cameral fistulae is abnormal embryogenesis. Other important causes include Trauma (stab injury or gunshot), Invasive procedures (coronary angiography, pacemaker implantation, or endomyocardial biopsy) or Cardiac surgery during a septal myomectomy [3].

Generally, the coronary cameral fistula is asymptomatic, but it can be clinically significant depending on the hemodynamic significance of the anomaly. Coronary fistulas draining into the left ventricle can produce a diastolic murmur and diastolic volume overload, mimicking aortic valve regurgitation [4]. Anginal symptoms may be the presenting feature, particularly in patients with multiple fistulae as a result of left-to-left shunting causing a coronary steal phenomenon and diastolic overload and in those patients with a single fistula, exertional dyspnea is more likely to predominate [5]. Patients with coronary cameral fistulas may present with myocardial infarction, congestive heart failure, arrhythmias [6].

Clinical examination findings in patients with coronary cameral fistulae include a Collapsing pulse, Wide pulse pressure, Diffuse apex beat Palpable third heart sound (S3), Loud continuous murmur on auscultation that peaks in mid to late diastole heard best at the mid to lower sternal border depending on the site of drainage of the fistula and Signs of heart failure [3].

Chest X-ray and electrocardiogram are usually not helpful in the diagnosis of coronary-cameral fistulas. Although chest X-ray may show cardiomegaly in the presence of significant shunt flow and electrocardiogram may reveal the effects of volume overload in larger fistulas, these findings are nonspecific [7].

Transthoracic and transesophageal echocardiography is useful especially in the evaluation of the hemodynamic effects of the fistula on cardiac chambers [8]. Inspection of the color Doppler imaging clearly may reveal the dilated coronary artery and sustained flow of blood from the epicardial surface to the Chamber cavity throughout diastole more suggestive of a coronary fistula [5-7].

Coronary cameral fistulae can be detected noninvasively using 64 slices multidetector CT scanner.

It provides high-quality three-dimensional images of the distal coronary artery and side branches [3].

Although gated magnetic resonance imaging allows detailed delineation of cardiac anatomy and blood flow, and may be helpful in resolving discrepancies raised by other investigations, coronary angiography remains the gold standard for imaging the coronary arteries and defining the pattern of structure and flow [7-9].

Common complications associated with coronary cameral fistulae include a Cardiac ischemia, Congestive heart failure, Cardiac arrhythmia, Infective endocarditis and a Rupture of coronary cameral fistula [3].

Hemodynamically insignificant fistulae, which are clinically silent and not associated with other abnormal findings, may not require further treatment. Large, hemodynamically significant fistulas should be closed by ligation that includes surgical or catheter closure [7]. The choice of approach for closure of these fistulae depends on the expertise of the team involved in taking care of the patient. Furthermore, the surgical approach would be more appropriate in patients with large fistulae, fistulae with multiple openings, aneurysmal dilatation, or acute angulations that are not amenable to catheterization [3].

Surgical methods of closure are associated with low mortality and morbidity; however, there is a risk of myocardial infarction postoperatively and also a risk of recurrence of the fistula. Risks of fistula closure with devices include myocardial infarction and migration of closure devices to extra coronary vascular structures or elsewhere within the coronary artery branches [1].

Antiplatelet therapy should be initiated after closure and continued for at least one year [3], the 2020 European guidelines state that small fistulas have a good prognosis without treatment. Medium or large fistulas are associated with long-term complications (angina, myocardial infarction, arrhythmias, heart failure and endocarditis). The presence of symptoms, complications and a shunt are the main indications for percutaneous or surgical closure. CCT is the preferred technique for the evaluation of high-risk anatomy, including features such as an intramural path and orifice abnormalities (slit-like orifice, acute angle lift-off, orifice > 1 cm above the Sino tubular junction) [10].

On the other side, the 2018 US guidelines highlight the importance of a competent team that may include congenital or non-congenital cardiologists and surgeons to determine the role of medical treatment and/or percutaneous or surgical closure [11].

Life expectancy for patients with a coronary cameral fistula is normal [3].

## CONCLUSION

Although the majority of patients are asymptomatic, nevertheless the risk of complications is present and stipulates the need for adequate and multidisciplinary care. Patients with coronary cameral fistulae should be managed by an interprofessional team that includes an interventional cardiologist, a cardiac surgeon, an echocardiographer, a radiologist. The imaging models allow the diagnosis to be evoked, but angiography remains the gold standard for the diagnosis. Surgical or interventional treatment is reserved only for symptomatic patients with large or multiple fistulas; however, close and close monitoring is necessary given the evolving nature of these types of anomalies.

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