

Multi-vessel Coronary Cameral Fistulas: The Role of Multimodality Imaging in Guiding Conservative Management

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Abstract

Coronary cameral fistulas (CCF) are rare anomalies that can lead to significant morbidity due to abnormal coronary shunting, which may result in coronary steal, volume overload, and aneurysmal dilatation of the affected segments. Current guidelines recommend that the management strategy—whether conservative follow-up, percutaneous intervention, or surgical closure—be individualized by a multidisciplinary team with expertise in congenital heart disease, while carefully considering the fistula's origin, drainage anatomy, haemodynamic impact, patient age, and comorbidities. We report a rare case of a 66-year-old woman with multiple CCFs involving all three major coronary arteries, which were incidentally detected during elective coronary angiography. To evaluate potential myocardial ischemia secondary to coronary steal, cardiac magnetic resonance imaging was performed, revealing no pathological signal abnormalities. Following a multidisciplinary heart team evaluation, a conservative management strategy was adopted due to the absence of significant functional impairment. The patient has remained asymptomatic throughout a two-year follow-up period. This case underscores the critical importance of functional assessment over anatomical complexity in guiding clinical management, highlighting that multimodality imaging is essential for identifying patients who can safely avoid unnecessary invasive interventions.

Keywords: Coronary cameral fistula, right ventricular drainage, multimodality imaging, conservative management

Introduction

Krause first described the concept of coronary artery fistula (CAF) in 1865 (1). In contemporary series, the prevalence of coronary fistulas is reported to range from 0.08% to 0.3% among patients undergoing coronary angiography. This rate increases to approximately 0.9% with the use of computed tomography (CT) or magnetic resonance angiography (2). In contrast, the incidence of coronary cameral fistulas (CCF) in the general population is exceedingly low, estimated at approximately 0.0002% (3). CAFs may be either congenital or acquired, with approximately 90% of cases considered to be congenital (4).

CCF is defined as a rare congenital anomaly characterised by an abnormal connection between a coronary artery and a cardiac chamber. Under normal conditions, coronary arteries originate

from the aorta and terminate in the myocardial bed; however, in the presence of CCF, blood flow deviates from its physiological course and shunts into a cardiac chamber. The right ventricle is reported to be the cardiac cavity most commonly drained by CCFs (5). In contrast, coronary artery-left ventricle fistulas are extremely rare, accounting for only approximately 2-3% of all CCF cases (6). This extremely low incidence of CCF means that evidence is largely limited to case reports; and consequently, each case requires a personalised, meticulous assessment and management strategy to address potential haemodynamic burden and ischaemic complications.

Current guidelines recommend that the choice between conservative management, percutaneous intervention, or surgical closure should be individualized by a multidisciplinary team comprising cardiologists and cardiac surgeons experienced



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in congenital heart disease, taking into account the anatomical characteristics of the fistula, its haemodynamic significance, and the patient's clinical profile (4,7). We report a rare case of multiple CCF involving all three major coronary arteries that were incidentally detected during elective coronary angiography.

Case Report

A 66-year-old woman with no prior history of cardiac disease but with known diagnoses of left adrenal adenoma and primary hyperaldosteronism (Conn's syndrome), presented to our cardiology outpatient clinic with typical angina. Her medical history was notable for hypertension, hyperlipidemia, and active smoking (50-pack-year history). She was receiving spironolactone and dexamethasone for the adrenal adenoma and telmisartan for hypertension.

On presentation, the electrocardiogram demonstrated a normal sinus rhythm at a rate of 72 beats per minute. Transthoracic echocardiography revealed a normal left ventricular ejection fraction and mild mitral, tricuspid, and aortic regurgitation. The maximum aortic valve velocity (V_{max}) was 1.3 m/s, and the estimated pulmonary artery pressure was 25 mmHg. The estimated Qp/Qs ratio was 1.1, with preserved right ventricular size and function, and no right-sided chamber enlargement or volume overload. Given her family history of coronary artery disease, comorbidities, and typical angina, she underwent coronary computed tomography (CCT) angiography. CCT angiography demonstrated an abnormal vascular course with focal contrast opacification in the distal extension of the acute marginal branch of the right coronary artery (RCA), adjacent to

the interventricular septum (IVS) at the mid-ventricular level, with drainage into the right ventricle (Figure 1). In addition, a thin septal branch originating from the distal left anterior descending (LAD) artery was observed coursing within the myocardium and was associated with focal contrast opacification and communication with the right ventricular cavity, consistent with a coronary-cameral fistula (Figure 1). Furthermore, one of the distal branches of the circumflex (CX) artery was found to course within the inferior wall at the apical level and drain into the right ventricle via the IVS. Collectively, these findings indicated the presence of multiple CCF involving all three major coronary arteries that drained into the right ventricle.

In addition, focal mid-myocardial contrast enhancement consistent with adipose tissue was identified at the left ventricular apex. A cardiac magnetic resonance (CMR) viability study was recommended to evaluate the possibility of ischemia related to coronary steal and was subsequently performed. Invasive coronary angiography demonstrated that the LAD and CX arteries were tortuous and ectatic, whereas the RCA was tortuous, non-dominant, and exhibited 20-30% proximal segment stenosis. CCF were identified in all three coronary arteries (Figures 2,3). Acetylsalicylic acid, metoprolol, and atorvastatin were added to the patient's existing medical therapy. CMR imaging was performed mainly to assess viability and scarring. Stress perfusion CMR was not performed. Late gadolinium enhancement imaging demonstrated no evidence of ischemic myocardial scarring in the sagittal, axial, and coronal planes, corresponding to Figures 4A, 4B, and 4C, respectively.

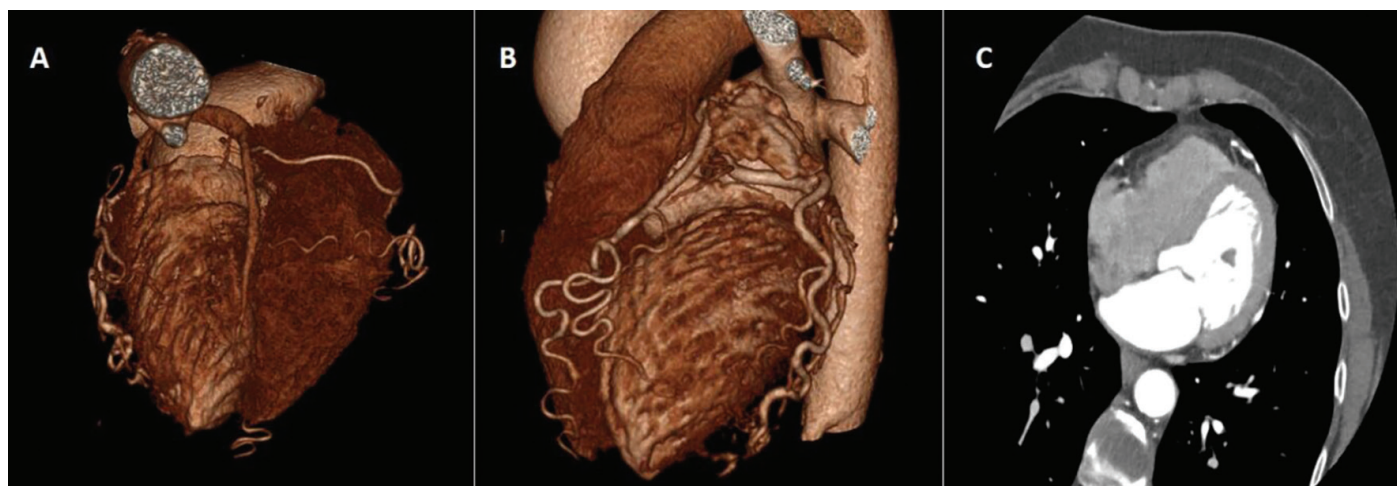


Figure 1. Coronary cameral fistulas on 3D computed tomography (CT) coronary angiography

Three-dimensional computed tomography (CT) coronary angiography demonstrating coronary cameral fistulas

(A) Fistula originating from the right coronary artery (B) fistula originating from the left anterior descending artery (LAD) (C) axial contrast-enhanced CT image showing an abnormal vascular structure consistent with an LAD-originating fistula

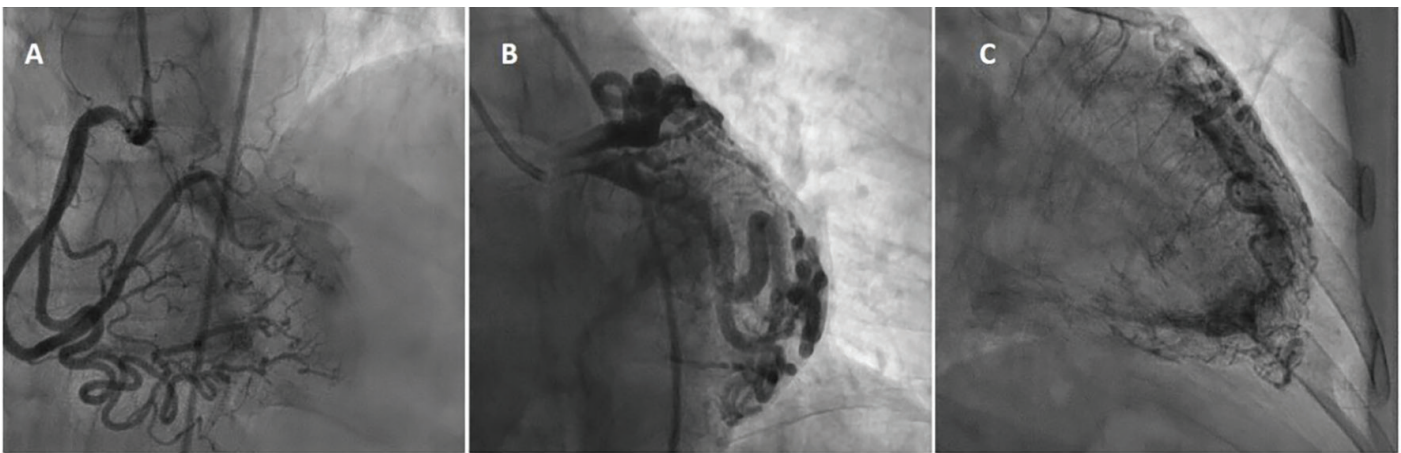


Figure 2. Early ventricular filling in conventional coronary angiography
In the coronary angiography images, early filling consistent with coronary artery fistulas is observed in the right, circumflex, and left coronary arteries, respectively (A, B, and C)

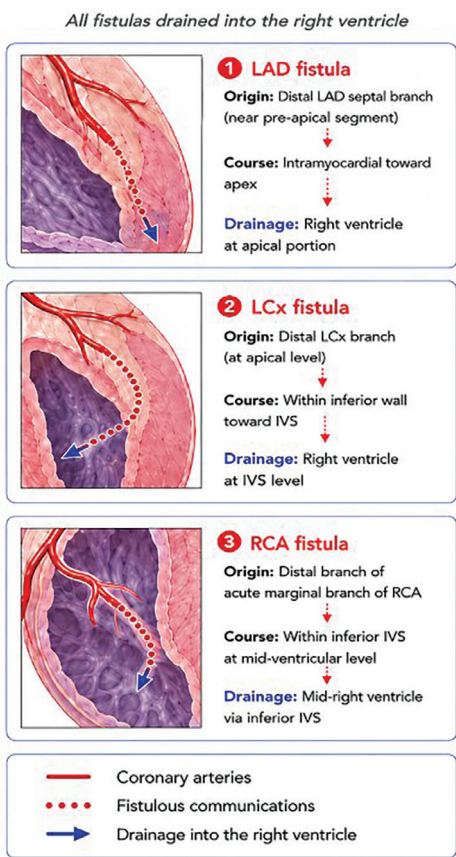


Figure 3. Schematic illustration of triple-vessel coronary-cameral fistulas
The illustration summarizes fistulous communications arising from the left anterior descending artery, circumflex artery, and right coronary artery, all draining into the right ventricle
LAD: Left anterior descending artery, LCx: Left circumflex artery, RCA: Right coronary artery

T1 mapping was obtained, demonstrating myocardial T1 values within normal limits. T2-weighted short-axis images showed no evidence of myocardial edema. Overall, left ventricular myocardial viability was preserved, with no evidence of ischemic scar, edema, or regional wall motion abnormality. Exercise treadmill testing was negative for inducible ischemia; the patient completed stage III of the Bruce protocol and achieved 10 METs without exercise-induced angina or ischemic ST-segment changes.

Following the initiation of medical therapy with acetylsalicylic acid, atorvastatin, and metoprolol, the patient's anginal symptoms completely resolved. The multidisciplinary heart team favored conservative follow-up with medical therapy in this individual patient, given the absence of heart failure, inducible ischemia on exercise testing, haemodynamically significant left-to-right shunting, right-sided volume overload, and pulmonary hypertension. The patient has been followed regularly for two years and remains asymptomatic on medical therapy.

Discussion

A CAF is a vascular malformation characterized by an abnormal communication between a coronary artery and a cardiac chamber or major vessel and may be either congenital or acquired. The fundamental pathophysiological mechanism involves the diversion of high-pressure coronary arterial blood away from the physiological low-pressure arteriolar and capillary network supplying the myocardium toward a low-resistance venous compartment (8).

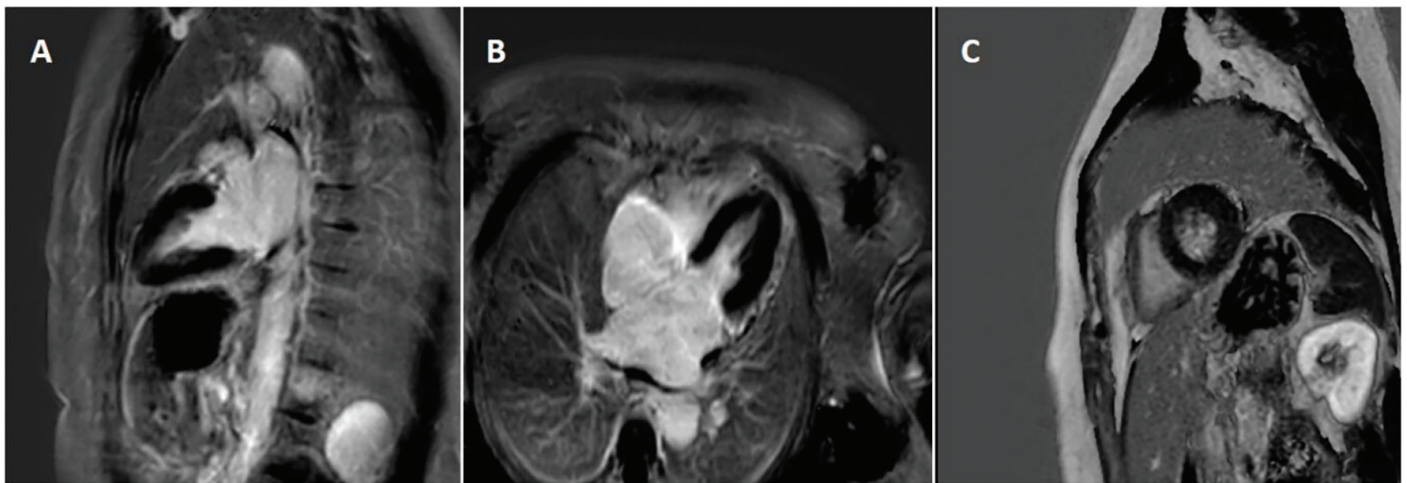


Figure 4. Cardiac magnetic resonance viability imaging

Late gadolinium enhancement images showed no evidence of ischemic myocardial scar. Sagittal (A), axial/transverse (B), and coronal (C) planes are shown

Although many coronary fistulas remain asymptomatic, symptomatic cases may present with heart failure, endocarditis, conduction abnormalities, or myocardial ischemia due to the coronary steal phenomenon, most commonly manifesting as angina and dyspnea (9). The haemodynamic impact of the fistula is primarily determined by its origin, course, and drainage site. Therefore, accurate assessment of the anatomical origin and termination of the fistula is essential to identify perfusion abnormalities distal to the fistula related to coronary steal (8).

Coronary steal is recognized as a key mechanism underlying exertional angina, particularly in conditions associated with increased myocardial oxygen demand, such as physical exertion. Small-diameter fistulas generally follow a benign natural course and often do not require treatment. In contrast, medium and large fistulas are associated with clinically significant long-term complications, including angina, myocardial infarction, arrhythmias, heart failure, and endocarditis. Accordingly, the presence of symptoms, fistula-related complications, or a haemodynamically significant shunt constitutes the main indications for percutaneous closure or surgical repair in patients with CAF (4,10). Current guidelines recommend individualized assessment by a multidisciplinary team experienced in congenital and acquired heart disease to determine the relative merits of medical therapy, percutaneous closure, or surgical repair in patients with CAF. However, because no clear consensus exists regarding the optimal timing or modality of closure, management should be guided by a case-specific risk–benefit assessment that balances anatomical complexity and procedural risk against the anticipated natural history and haemodynamic significance of the fistula (4,7).

In a surgical series of 46 patients, the predominant preoperative symptoms were angina and heart failure. In the same study, the incidence of postoperative myocardial infarction, attributed to reduced flow in the dilated coronary artery proximal to the site of fistula closure, reached 11%. Furthermore, long-term survival was significantly lower compared with an age-matched reference population (8). In another reported case, a 31-year-old patient with a giant aneurysmal fistula originating from the sinus node artery of the RCA and draining into the superior vena cava underwent successful surgical management, including aneurysm resection, aortocoronary bypass using a saphenous vein graft, and fistula closure. The postoperative course was uneventful, and the patient was discharged in good clinical condition (11).

In our case, the preserved left ventricular systolic function, the absence of significant volume overload or pulmonary hypertension on echocardiography, and the lack of obstructive coronary artery disease on invasive coronary angiography suggest a limited haemodynamic impact of the fistulas. CMR imaging showed no myocardial scar or regional wall motion abnormality. Exercise treadmill testing demonstrated no objective evidence of inducible ischemia: no anginal symptoms or ischemic ST-segment changes were observed.

Although a complex network of fistulas involving all three major coronary arteries was present, the potential morbidity and mortality associated with surgical or transcatheter closure were considered high relative to the patient's current symptom burden and clinical stability. Following a multidisciplinary heart-team evaluation, conservative follow-up with medical therapy was favored for this patient in the absence of symptomatic heart

failure, inducible ischemia on exercise testing, haemodynamically significant left-to-right shunting, right-sided volume overload, or pulmonary hypertension. This case highlights that extensive anatomical involvement does not necessarily translate into haemodynamic significance, emphasizing the importance of functional assessment in guiding management decisions.

Unlike previous reports of triple-vessel CCF, which have mainly described left ventricular drainage or symptomatic presentations, the present case involved fistulous drainage from all three major coronary arteries into the right ventricle. Importantly, this anatomical complexity was not associated with significant shunt burden, right-sided volume overload, pulmonary hypertension, or adverse events during two years of conservative follow-up.

Conclusion

Congenital CCF are rare malformations of the coronary arteries and are infrequently detected on coronary angiography. Management strategies vary widely, ranging from close clinical follow-up in asymptomatic patients with haemodynamically insignificant lesions to transcatheter closure in anatomically suitable cases and to surgical repair in selected patients.

Ethic

Informed Consent: Written informed consent was obtained from the patient.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

Footnotes

Author Contributions

Surgical and Medical Practices: M.K., Concept: M.K., Ç.O., Design: M.K., M.M.B., Data Collection or Processing: M.K., Ç.O., M.M.B., Analysis or Interpretation: M.K., Ç.O., M.M.B., Literature Search: M.K., Ç.O., M.M.B., Writing: M.K., Ç.O.

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