

MINI-FOCUS ISSUE: HEART FAILURE

BEGINNER

CASE REPORT: CLINICAL CASE

Acute Heart Failure From Coronary Cameral Fistula



Diagnostic Challenge and Therapeutic Opportunity

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ABSTRACT

A 40-year-old African American woman presented with dyspnea, orthopnea, weight gain, and ankle edema. She was admitted for acute decompensated heart failure. Coronary angiography revealed coronary cameral fistula. We used multiple imaging modalities to confirm the fistula's pathway. Her left ventricular systolic function improved after repair of coronary cameral fistula. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2020;2:2368-71) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

A 40-year-old African American woman without prior cardiac history presented with a 2-week history of increasing dyspnea, orthopnea, weight gain, and ankle edema. On initial assessment, her blood pressure was elevated at 160/115 mm Hg, heart rate of 87 beats/min, respiratory

rate was 18 breaths/min, temperature was 97.6°F, and oxygen saturation was 98% on room air. Physical examination was significant for decreased breath sounds at both lung bases with crackles superiorly, +2 bilateral lower limb pitting edema, but surprisingly normal heart sounds, diffuse point of maximal impulse, and no murmur.

LEARNING OBJECTIVES

- To be familiar with coronary cameral fistula as a rare cardiac anomaly that can rarely result in acute decompensated heart failure.
- To recognize the multiple imaging modalities that can be used in patients with coronary cameral fistula to diagnose and confirm the fistula's pathway.
- To evaluate treatment options for patients with symptomatic coronary cameral fistula including surgical correction, which might provide an opportunity to improve heart failure.

DIFFERENTIAL DIAGNOSIS

Acute decompensated heart failure (ADHF) secondary to ischemic or nonischemic cardiomyopathy.

INVESTIGATIONS

Diagnostic data included an elevated N-terminal pro-B-type natriuretic peptide value (3,110 pg/ml; normal range, <450 pg/ml). Electrocardiogram showed normal sinus rhythm without ischemic changes, and a chest radiograph demonstrated findings suggestive of pulmonary edema, cardiac silhouette enlargement, and small bilateral pleural effusions.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the [Author Center](#).

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FIGURE 1 Coronary Angiogram With Coronary Cameral Fistula



Coronary angiogram showed a large and tortuous anomalous vessel arising from the mid left circumflex artery.

MANAGEMENT

The patient was initially treated for ADHF with intravenous diuretics with marked symptomatic improvement. An echocardiogram revealed mild left

ventricular dilation with reduced systolic function, with an estimated ejection fraction of 42% (Video 1).

Coronary angiography revealed a right-dominant circulation with a large and tortuous vessel arising from the mid left circumflex just distal to the first obtuse marginal, and emptying into a difficult to define space (Figure 1). Brisk clearance of contrast opacification suggested significant shunting, but the full course could not be identified (Video 2). Coronary anatomy and hemodynamic significance were further delineated with cardiac computed tomographic angiography (Figures 2A, 2B, and 3, Videos 3A and 3B) and magnetic resonance imaging (Figures 4A, 4B, and 5, Video 4) A large fistulous connection appeared to terminate in the right atrium. The cardiac magnetic resonance imaging pulmonary blood flow/systemic blood flow measurement was 1.4:1.

A right heart catheterization was performed to further confirm the shunt route using oxygen saturation. Oxygen saturations in the superior vena cava (69%), high right atrium (81%), middle right atrium (75%), and low right atrium (71%) were consistent with a left-to-right shunt emptying into the right atrium. The tortuosity and large caliber of the fistula warranted closure through open cardiothoracic surgery (Central Illustration).

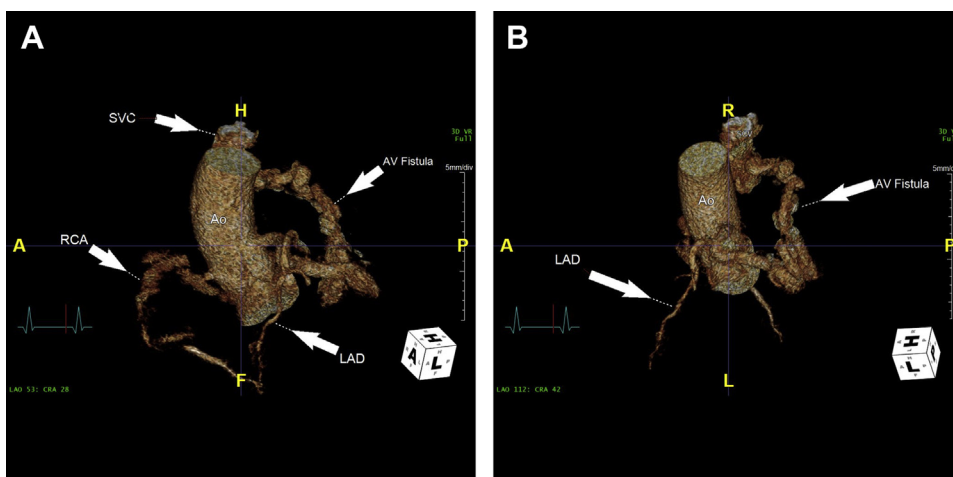
During surgery, the left circumflex to right atrium shunt was identified and ligated. In addition, a

ABBREVIATIONS AND ACRONYMS

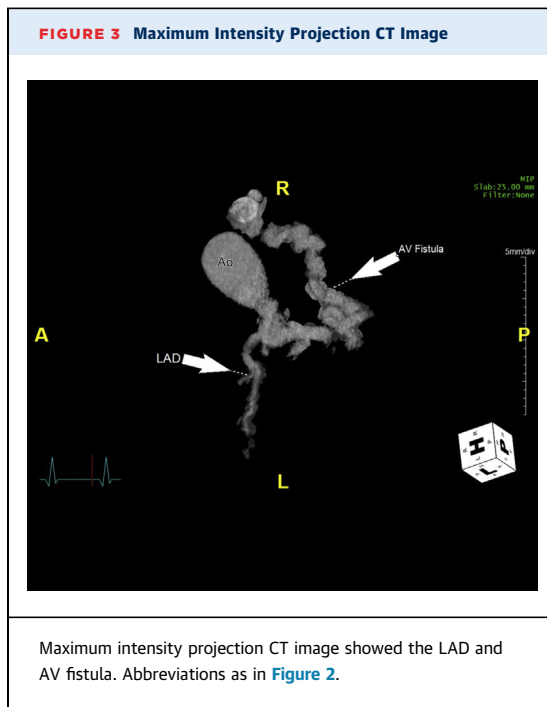
ADHF = acute decompensated heart failure

CCF = coronary cameral fistula

FIGURE 2 Volume-Rendered CT Scans



(A) Left-lateral view volume-rendered CT scan image showed the aorta, superior vena cava, left anterior descending, right coronary artery, and arteriovenous fistula. (B) Superolateral view volume-rendered CT scan image showed the aorta, left anterior descending, and arteriovenous fistula. Ao = aorta; AV = arteriovenous; CT = computed tomography; LAD = left anterior descending; RCA = right coronary artery; SVC = superior vena cava.

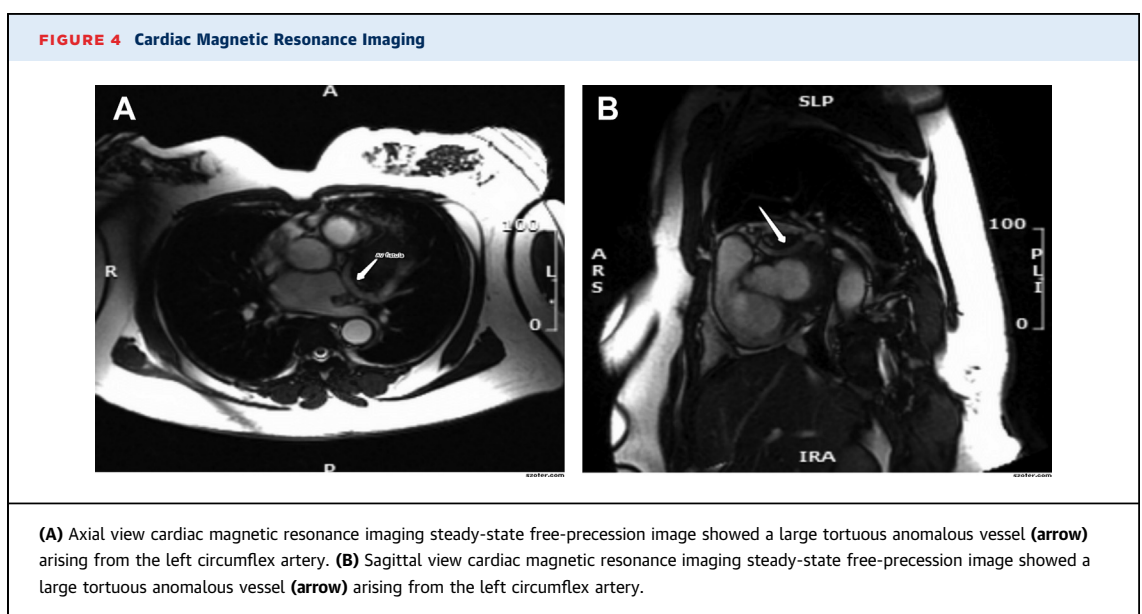


branch from the right coronary artery emptying into the right atrium was identified and ligated. The patient was treated with appropriate heart failure medical therapy and discharged.

DISCUSSION

Coronary cameral fistula (CCF) is a rare cardiac anomaly described as an abnormal connection

between a coronary artery with any of the 4 chambers of the heart (1). The term “camera” in Latin means a vault; hence a chamber. Muir (2) was first to suggest the name “coronary arterio-cameral fistula” to describe communication between a coronary artery and a cardiac chamber. This term was suggested to differentiate this anomaly from other arteriovenous communications of the heart. Usually CCF is congenital, but it can also occur secondary to invasive cardiac procedures, such as pacemaker placement, coronary artery bypass grafting, or endomyocardial biopsy (3). Yuksel *et al.* (4) reported that most CCFs originate from the left anterior descending coronary artery (46.6%). Most CCFs drain into the right heart including the right ventricle (40%), the right atrium (26%), and the pulmonary arteries (17%) (5). Patients with CCF are usually asymptomatic. Adults with symptomatic CCF usually present in their third or fourth decades of life (6). Common presenting symptoms for CCF include angina, dyspnea on exertion, syncope, and palpitations (7). The most common reported finding on physical examination is a continuous murmur (8). Patients with CCF presenting with new-onset ADHF are rare and very few cases have been reported in the literature. Cardiac catheterization is considered the gold standard for diagnosing CCF, although in some cases, transthoracic echocardiography can be used to make the diagnosis. However, it is usually difficult to determine the course of the fistula with transthoracic echocardiography alone (9). Electrocardiogram-gated coronary computed tomography angiography plays an important role in identifying the anatomy of CCF (10). In



addition, cardiac magnetic resonance imaging can provide more detailed evaluation of the fistula's anatomy, blood flow within the fistula, hemodynamic significance of any shunt (pulmonary blood flow/systemic blood flow), and myocardial wall viability.

Our case is unusual in several ways. First, our patient presented with ADHF and did not have any cardiac or valvular anomalies other than the CCF, nor was there a murmur. Second, this case required multiple imaging modalities to confirm the fistula's pathway, which was difficult to delineate using cardiac catheterization alone. Finally, the patient's left ventricular systolic function improved after repair of CCF.

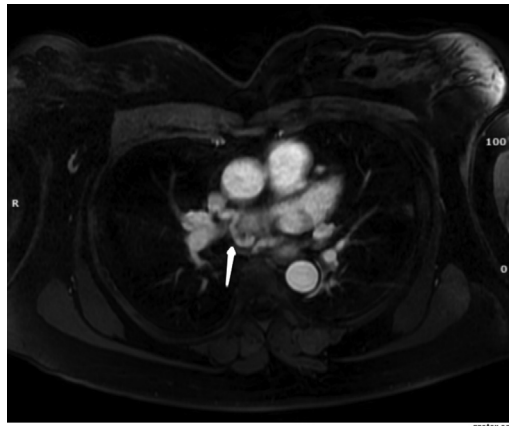
FOLLOW-UP

On follow-up in the cardiology clinic she was doing well. Repeat echocardiogram showed normal left ventricular systolic function, with an increase of the left ventricular ejection fraction from 42% to 65% (Video 5).

CONCLUSIONS

CCF is a rare cardiac anomaly described as an abnormal connection between a coronary artery with any of the 4 chambers of the heart. Patients with CCF are usually asymptomatic; however, they can rarely present with ADHF. The diagnosis of CCF can be challenging requiring multiple imaging modalities. In addition, surgical correction can provide an opportunity to improve heart failure symptoms and systolic function.

FIGURE 5 Axial Spoiled Gradient Echo Magnetic Resonance Imaging Sequence



An axial spoiled gradient echo magnetic resonance imaging sequence with fat suppression showed a large tortuous anomalous vessel (arrow) draining in right atrium.

AUTHOR DISCLOSURES

All authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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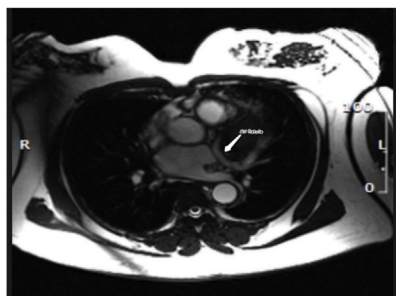
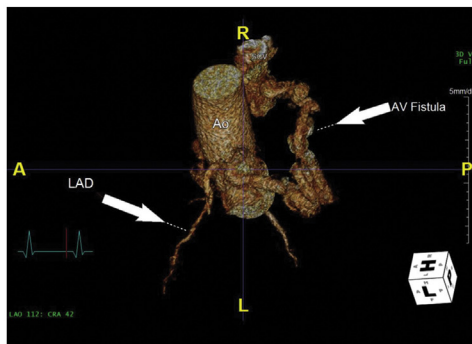
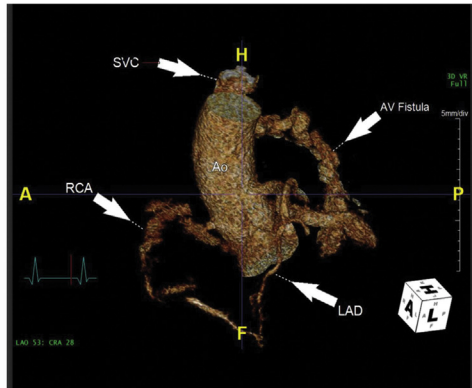
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KEY WORDS acute heart failure, cardiac magnetic resonance, computed tomography, coronary artery fistula, coronary cameral fistula, coronary vessel anomaly

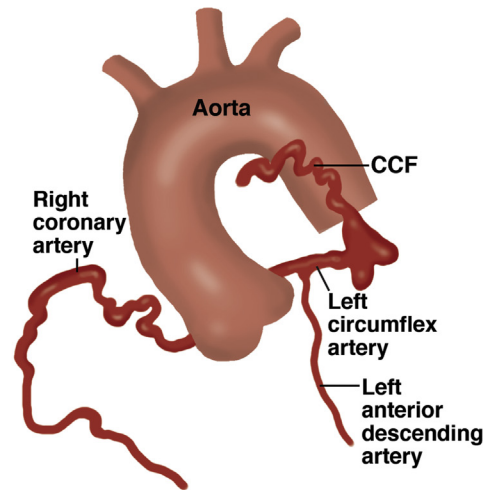
APPENDIX For supplemental videos, please see the online version of this paper.

CENTRAL ILLUSTRATION Treatment of Symptomatic Coronary Cameral Fistula

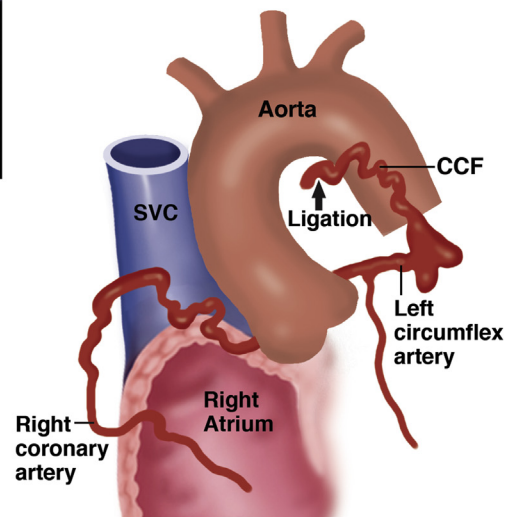
Coronary Cameral Fistula (CCF)



Anatomy



Surgery



Treatment Options for Symptomatic CCF

- Surgical closure
- Transcatheter closure

Multiple factors play a role in choosing a method of closure including: origin, size, and anatomy of fistula. In our case the tortuosity and large caliber of the fistula warranted surgical closure.