

CONGENITAL HEART DISEASE

IMAGING VIGNETTE: CLINICAL VIGNETTE

Coronary Artery Fistula Involving the RCA, LAD, and the Main Pulmonary Artery



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ABSTRACT

We present a case of a complex congenital coronary artery fistula between the right coronary artery, left anterior descending artery, and the main pulmonary artery complicated by massive aneurysms and a left-to-right shunt. We highlight the multimodality approach to assessment and the importance of individualized management of complex coronary fistulas. (J Am Coll Cardiol Case Rep 2024;29:102256) Crown Copyright © 2024 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 68-year-old woman presented with intermittent chest pain for 20 years but had recently become exertional with associated dyspnea and presyncope. She reported a history of a congenital cardiac anomaly diagnosed over 20 years ago. Background otherwise included hypertension and dyslipidemia. Clinical examination showed dual heart sounds with a 3/6 continuous murmur. She was hemodynamically stable with no peripheral signs of heart failure.

The 12-lead electrocardiogram was normal and transthoracic echocardiogram showed normal anatomical alignment, biventricular size and function, and valvular anatomy. Blood test results showed normal Troponin-I of 4 ng/L. Computed tomography coronary angiography revealed a coronary artery fistula (CAF) arising from the proximal right coronary artery (RCA) and passing antero-superiorly over the right ventricular outflow tract before passing laterally around to abut the left anterior descending (LAD) coronary artery before inserting into the left lateral wall of the main pulmonary artery (MPA) (Figure 1). The fistula contained 2 large aneurysmal components: the first sat in the superior portion of the right atrioventricular groove and the second abutted the superior aspect of the LAD. A leash of vessels surrounded the distal fistula arising from a septal branch of the LAD. The computed tomography coronary angiography otherwise showed diffuse mild nonobstructive coronary artery plaque. Invasive coronary angiography confirmed the CAF and the feeding vessels from both the RCA and LAD. Cardiac magnetic resonance imaging showed a left-to-right shunt with Qp/Qs of 1.2. There were no other congenital abnormalities identified.

Given the large fistula size and tortuosity and the multiple vessels feeding the CAF, the multidisciplinary heart team recommended surgical resection. Following cardiopulmonary bypass and antegrade cardioplegia, the fistula was transected, and the RCA stump was oversewn. The fistula was further dissected free of the adventitial tissues and then transected at the point of entry into the MPA. The MPA was oversewn. The

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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](#).

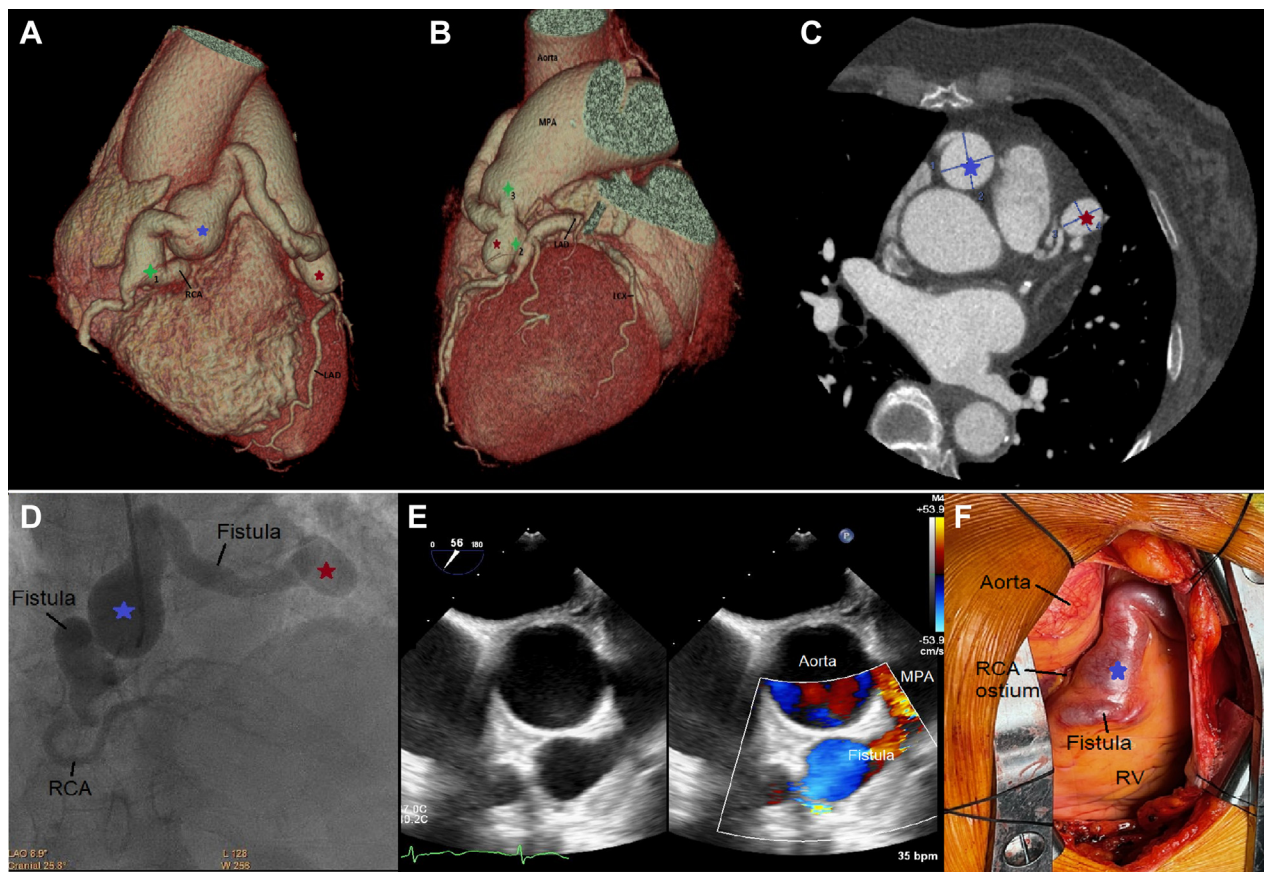
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**ABBREVIATIONS
AND ACRONYMS**

CAF = coronary artery fistula
LAD = left anterior descending
MPA = main pulmonary artery
RCA = right coronary artery

abnormal feeding vessel from the LAD to the fistula was clipped. Postoperative recovery was uneventful. At 1 month follow-up, her exercise tolerance improved, and the chest pain had resolved.

CAF is a rare coronary artery anomaly. Because of the heterogeneity of CAFs, optimal management strategies vary and should be determined by a multidisciplinary team. The 2018 American Heart Association guidelines recommend consideration of patient age, ischemic symptomatology, heart failure and/or arrhythmia, the anatomical structure of the anomaly, and the risk of sudden cardiac death.¹ Percutaneous closure is feasible and successful for selected patients with smaller and less tortuous CAFs. Highly tortuous vessels are technically difficult to wire and require arteriovenous wire loop for support.² Surgical resection has traditionally been first-line management, particularly with giant and tortuous CAFs not suitable for percutaneous intervention.³ Overall, symptomatic CAFs and large fistulas should be closed promptly. Multimodality imaging can guide management and improve outcomes.

FIGURE 1 Multimodality Imaging of the Coronary Artery Fistula

(A) Computed tomography (CT) angiographic 3-dimensional (3D) reconstructed view of the coronary artery fistula (CAF) from the right coronary artery (RCA) (green star 1) with 2 aneurysmal segments (blue and red stars). (B) CT angiographic 3D reconstructed view of the CAF with a second aneurysmal segment (red star) and communication with the left anterior descending (LAD) coronary artery (green star 2) before terminating in the main pulmonary artery (MPA) (green star 3). (C) CT angiographic axial view of the aneurysmal segments adjacent to the RCA, measuring 24 mm × 23 mm (blue star), and the LAD, measuring 18 mm × 14 mm (red star). (D) Invasive coronary angiographic view of the RCA, complex fistula with 2 aneurysmal segments adjacent to the RCA (blue star), and the LAD (red star). (E) Intraoperative transesophageal echocardiogram mid-esophageal view at 56° showing the communication between the CAF and the MPA. (F) Surgical view showing the CAF and the aneurysmal segment (blue star). RV = right ventricle.

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KEY WORDS chest pain, coronary angiography, coronary vessel anomaly