JACC: CASE REPORTS
PUBLISHED BY ELSEVIER ON BEHALF OF THE AMERICAN COLLEGE OF CARDIOLOGY FOUNDATION.

VOL. 4, NO. 13, 2022

CASE REPORT

INTERMEDIATE

CLIICAL CASE: IMAGING AND GENERAL CARDIOLOGY

Indispensable Role of Multimodality Imaging in Diagnosis and Management of Coronary Arteriovenous Fistulas

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ABSTRACT

We describe a patient with right coronary artery to coronary sinus fistula requiring surgical elimination. The decision process in managing fistulas depends on the size, site of origin, and symptoms caused by the fistula. We highlight the pivotal role of multimodality cardiovascular imaging in the diagnosis and management of coronary fistulas. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2022;4:826–831) 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/).

HISTORY OF PRESENTATION

A 74-year-old man with a known coronary artery fistula (CAF) diagnosed in his 50s presented with progressive exertional dyspnea (New York Heart Association functional class III), reduced exercise tolerance, and abdominal discomfort. On initial evaluation, he was normotensive and his oxygen saturation was 97% on room air. Physical examination revealed a continuous murmur of grade 4/6 intensity, best appreciated at the left third and fourth intercostal spaces. Electrocardiography showed normal rate and rhythm with a QTc duration of 491 ms.

LEARNING OBJECTIVES

- To describe a unique case of CAF connecting the RCA to the coronary sinus, leading to a giant RCA aneurysm and biventricular heart failure.
- To recognize the role of multimodality imaging in planning complex structural heart interventions.
- To realize the indications and technical approaches to CAF closure and postoperative management.

MEDICAL HISTORY

His medical history included a right coronary artery (RCA) to coronary sinus (CS) fistula, hypertension, hyperlipidemia, and gout.

DIFFERENTIAL DIAGNOSIS

The differential diagnoses included acute decompensated heart failure due to hemodynamic sequela of RCA to CS fistula, ruptured sinus of Valsalva aneurysm, intracardiac shunts, unroofed coronary sinus, ventricular septal defect with aortic insufficiency, venous hums, aortopulmonary window, and pulmonary arteriovenous fistula.

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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.

Manuscript received December 21, 2021; revised manuscript received March 27, 2022, accepted April 13, 2022.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2022.04.005

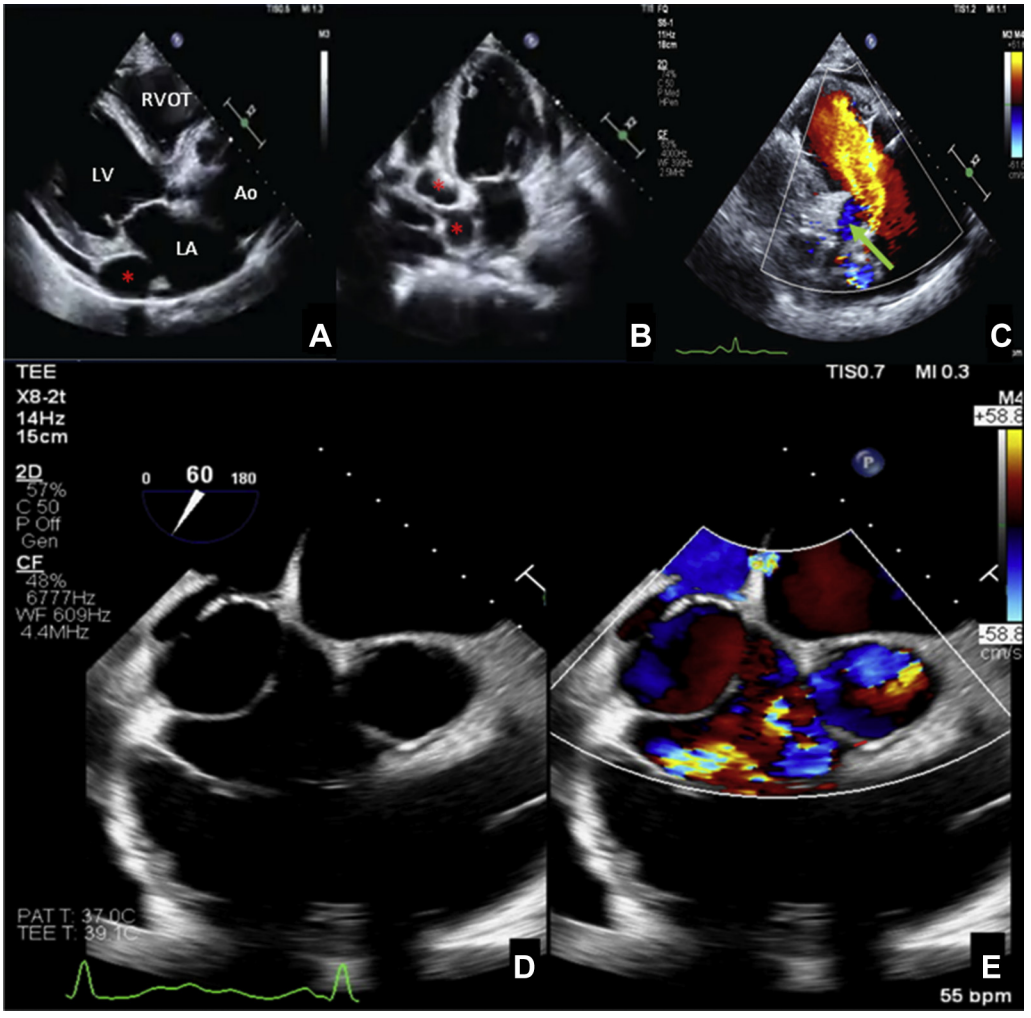
INVESTIGATIONS

A transthoracic echocardiogram showed a dilated CS and a massively enlarged and convoluted RCA. The left ventricle (LV) was moderately dilated, with a mildly reduced ejection fraction (45%). The right ventricle (RV) was moderately dilated, with reduced systolic function (fractional area change 19%) (**Figures 1A and 1B**, Videos 1 and 2). Color Doppler images showed a continuous high-velocity turbulent flow signal from the RCA fistula into the right atrium (RA) (**Figure 1C,** Video 3) with evidence of severe eccentric functional mitral regurgitation (peak mitral E velocity of 1.8 m/s). Transesophageal echocardiography redemonstrated the fistula, connecting the giant diffuse aneurysmal RCA to the CS with high-turbulent jet across the CS into the right atrium, with approximate diameter of 6 to 8 mm (**Figures 1D and 1E,** Video 4). Multidetector cardiac computed tomography (CT) confirmed the presence of a diffusely dilatated giant RCA aneurysm

ABBREVIATIONS AND ACRONYMS

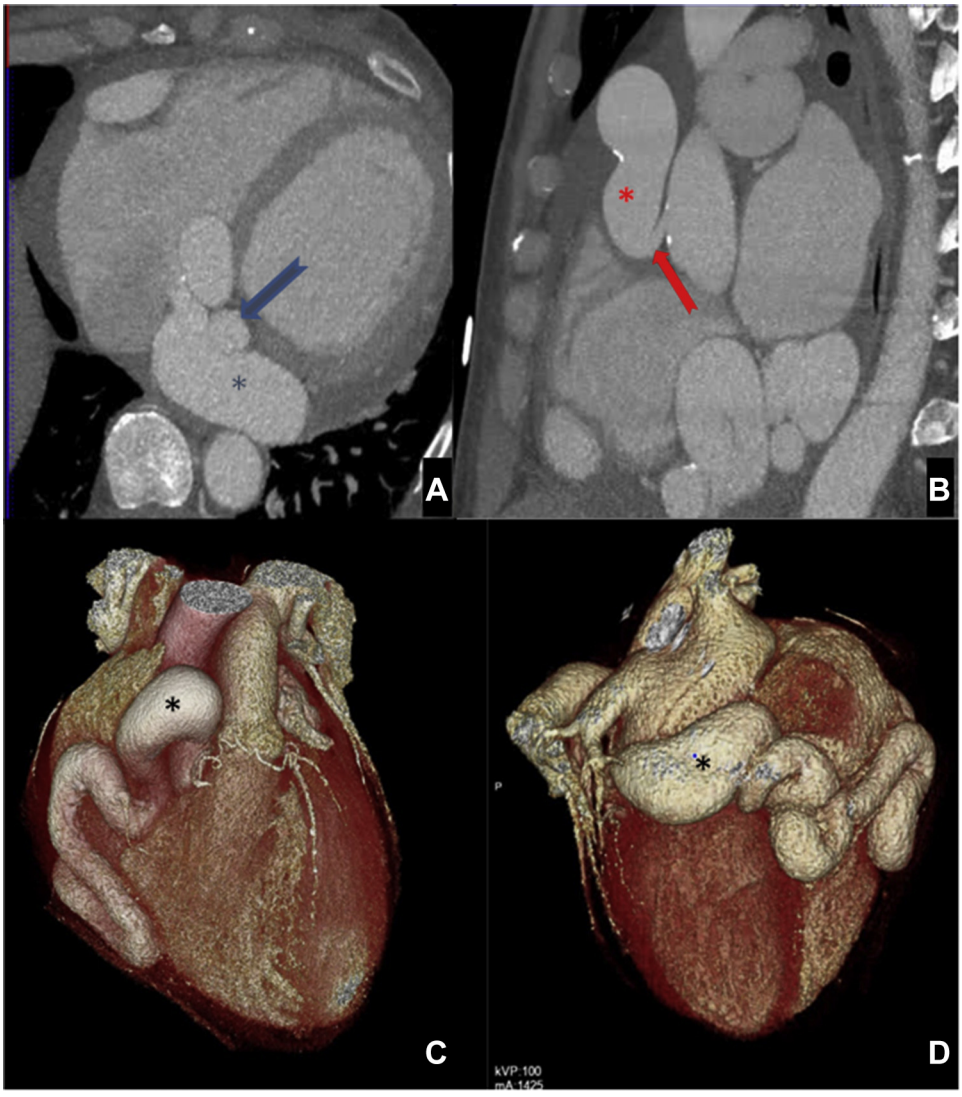
CAF = coronary artery fistula
CS = coronary sinus
CT = computed tomography
RCA = right coronary artery
RV = right ventricle

FIGURE 1 Preoperative Transthoracic Echocardiographic and Intraoperative Transesophageal Echocardiographic Images



Transthoracic echocardiographic images: **(A)** Parasternal long-axis projection showing a severely dilated coronary sinus **(red asterisk)** and left atrium (LA). **(B)** Apical 4-chamber view demonstrating the severely enlarged and tortuous right coronary artery **(red asterisks)**. **(C)** Right ventricular inflow tract (RVOT) view with color Doppler shows flashing jet of coronary sinus **(green arrow)** with high-velocity turbulence in the right atrium. **(D)** Transesophageal echocardiographic images are shown in midesophageal short-axis projection, demonstrating the coronary sinus, aorta, and tricuspid valve **(D)** and high-velocity flow entering the right atrium through the coronary sinus **(E)**. Ao = aorta; LV = left ventricle.

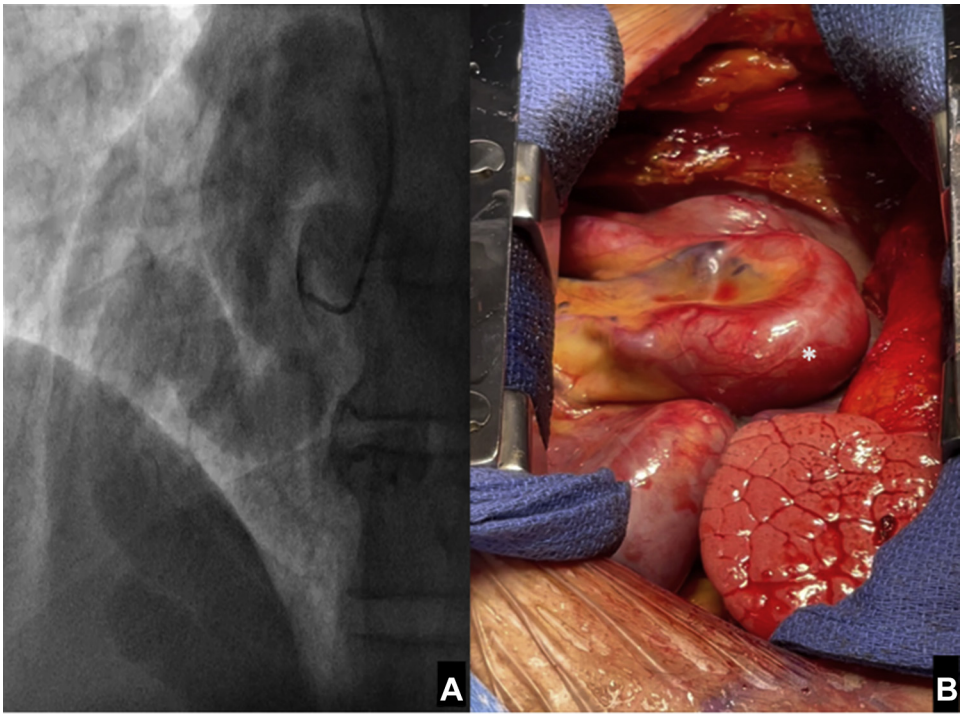
FIGURE 2 Preoperative Enhanced Computed Tomography Imaging



(A) Multidetector cardiac computed tomography showing the aneurysmal right coronary artery **(blue arrow)** entering the dilated coronary sinus **(blue asterisk)**. **(B)** Right coronary artery is redemonstrated, originating from the right sinus of Valsalva with a normal caliber **(red arrow)** and an aneurysmal distal segment **(red asterisk)**. **(C, D)** Volume-rendered 3D images show **(C)** a dilated right coronary artery arising from the sinus of Valsalva **(black asterisk)** and **(D)** a dilated right coronary artery entering into the coronary sinus **(black asterisk)**.

with a fistula to an enormously enlarged CS (Figures 2A and 2B). The largest segment of the aneurysm within the distal RCA measured 34 mm and within the CS measured 29 mm. The 3-dimensional volume-rendered CT images were used to define the anatomical course of the fistula showing the sinus of Valsalva as the origin of the large fistula and the coronary sinus as the termination site (Figures 2C and 2D). Cardiac catheterization showed an aneurysmal RCA with a significant pressure drop: 70/40 mm Hg in the proximal RCA and 138/70 mm Hg in the ascending aorta (Figure 3A, Video 5). Biventricular heart failure was suspected with RV pressure of 60/25 mm Hg, mean pulmonary artery pressure of 40 mm Hg, and mean pulmonary capillary wedge pressure of 20 mm Hg. Furthermore, significant coronary artery disease was noted in the first obtuse marginal (proximal 85%-90% stenosis).

FIGURE 3 **Preclosure Cardiac Catheterization and Intraoperative Image**



(A) Still images of selective coronary angiography in right anterior oblique 90° straight projection, showing a massively aneurysmal right coronary artery with a fistula to a giant coronary sinus. **(B)** Intraoperative image redemonstrating enlarged and tortuous right coronary artery **(white asterisk)**.

MANAGEMENT

A definitive surgical exclusion was planned, given the symptoms and concurrent need for revascularization. Intraoperatively, the giant tortuous RCA was noted to have multiple branches feeding the RV myocardium and overlying the aorta and most of the right side of the heart (Figure 3B). The patient underwent 2-vessel bypass grafting (saphenous vein grafts to the posterior descending artery and the first obtuse marginal) along with surgical exclusion of the RCA-to-CS fistula. Intraoperative transesophageal echocardiography revealed adequacy of the fistula exclusion with no residual fistula tract visible. Also, a perforated aortic valve leaflet was found during the surgery, requiring a concomitant aortic valve replacement.

DISCUSSION

CAF is an anomalous communication between the coronary artery(s) and a cardiac chamber or major vessels. The prevalence of CAFs by CT angiography is estimated to be 0.9% of coronary anomalies, notably higher than the previously presumed 0.002% in invasive angiography.[1] CAFs may be congenital or acquired after cardiac surgery or direct trauma. An estimated 20% to 45% of CAFs are associated with other congenital heart disease, including atrial septal defect, tetralogy of Fallot, patent ductus arteriosus, ventricular septal defect, and pulmonary atresia.[2] Most fistulas arise from the RCA (52%) and drain into a low-pressure structure, most commonly the RV (41%).[3] The ensuing physiological derangements of CAFs depend on the anatomical course of the aberrant connection (origin and termination sites) and the size of the fistula. CAFs bypass the myocardial capillary bed, resulting in a low flow resistance leading to dilation of the coronary arteries proximal to the site of termination. Whereas small CAFs close spontaneously, large fistulas often lead to marked shunting with progressively worsening symptoms. As the fistula increases in size, the left-to-right shunting can

occur, resulting in chamber dilation, ventricular dysfunction, and myocardial ischemia due to steal phenomenon.[4]

Accurate delineation of the coronary anatomy, fistulous connection, and extracardiac structures remains the cornerstone of preprocedural planning. As highlighted in this case, a multimodality imaging approach is paramount in the diagnosis and management of CAFs. Transthoracic echocardiography plays an important role in assessing the hemodynamics and the severity and direction of the shunt. Coronary angiography is the most commonly used imaging modality, allowing clear delineation of the origin and proximal course of the fistula. However, delayed antegrade contrast filling and contrast dilution at the distal drainage site makes it a suboptimal tool. Cardiac CT is particularly helpful in patients with distal fistulas because it allows for more accurate evaluation of the size, anatomical course, and origin and termination sites of the fistula. A transesophageal echocardiogram, similarly to cardiac CT, can also precisely delineate the anatomy of the fistula, and it has the unparalleled advantage of intraoperative localization and guidance during surgical closure.

In the 2008 guidelines for the management of congenital heart disease in adults from the American College of Cardiology/American Heart Association (ACC/AHA), surgical or transcatheter exclusion is a Class I recommendation (Level of Evidence: C) for symptomatic small-to-moderate-size fistulas and for large fistulas irrespective of symptoms.[5] Other indications include evidence of ischemia, endarteritis, vessel rupture, chamber enlargement, or ventricular dysfunction.[4] The updated 2018 ACC/AHA guidelines emphasize the importance of a multidisciplinary approach to the elimination of CAFs, depending on the expertise of the physicians in transcatheter and surgical exclusion techniques.[6] Therefore, the management strategy for these patients is variable and is largely driven by their anatomical suitability for either transcatheter or surgical closure.[4] CAFs originating from the distal coronary segments, as in our case, are particularly challenging because of the high risk of postclosure myocardial infarction. In such cases, surgical closure with concomitant bypass of the distal branches is favored to preserve the distal coronary flow. However, fistula closure, by either a percutaneous or a surgical approach, may result in stagnation of flow at or near the termination site, leading to thrombus formation.[4] Therefore, indefinite anticoagulation therapy is recommended for patients with at least moderate persisting aneurysmal dilation after closure.[4,7]

FOLLOW-UP

Postoperatively, the patient experienced episodes of symptomatic bradyarrhythmia, ensuing in the placement of a permanent pacemaker. Currently, the patient is asymptomatic 9 months after the procedure and has returned to an active lifestyle. A transthoracic echocardiogram 8 months after surgery showed no evidence of recanalization and marked improvement of the overall function.

CONCLUSIONS

CAF is a rare cause of heart failure, and closure is indicated when symptoms develop. A multimodality imaging approach is imperative in the evaluation and management of CAFs. More data are needed to evaluate postoperative outcomes and determine the optimal approach for fistula closure.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

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

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KEY WORDS computed tomography, coronary artery aneurysm, coronary fistula, echocardiography, multimodality imaging

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