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Surgical Ligation of a Large Coronary-Cameral Fistula Suspected of Causing Coronary Steal Phenomenon



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Coronary artery fistulas are rare anomalies that are categorized by site of termination: coronary-cameral (into a heart chamber) and coronary arterio-venous (into systemic or pulmonary vessels). Alternatively, they are classified by segment of origin: Sakakibara type A originates in the proximal third of the native vessel and Sakakibara type B originates beyond the proximal third or as a termination of the native vessel.¹ Twenty percent of patients will have 2 or more fistulas. Most are diagnosed incidentally on computed tomography coronary angiography (CTCA) or invasive coronary angiography (with CTCA being more sensitive).² When identified, a thorough evaluation of the clinical relevance (steal, degree of shunt, risk of endocarditis and thrombosis) is required. CTCA may be utilized to accurately define origin and termination, where invasive coronary angiography is equivocal. Closure is indicated when symptomatic, with little evidence to support medical management given the natural history of fistulas to enlarge. Closure is also recommended for large fistulas (≥ 2 times the diameter of the coronary artery of origin), irrespective of symptoms.³ Closure can be surgical or percutaneous. A heart team decision is advocated by society guidelines as neither technique has been demonstrated to be superior.

A 58-year-old male presented with unstable angina with inferior limb-lead T wave inversion (Supplemental Figure S1A). Coronary angiography demonstrated severe atherosclerotic stenosis in the proximal left anterior descending artery (LAD), moderate right coronary artery (RCA) stenoses, and 2 coronary-cameral fistulas: a large-caliber RCA to the right ventricle (RCA-RV) (Figure 1A, Supplemental Video 1) and a small-caliber septal perforator to the right ventricle (Figure 1B, Supplemental Video 2). CTCA confirmed the origin of the RCA-RV fistula as the first branch of the RCA (hence deemed to be a conus artery fistula; Figure 1C). CTCA also confirmed the course and termination of these fistulas (Figure 1D, E). Given the inferior limb-lead T wave inversion, it was suspected that the RCA-RV fistula was resulting in a coronary steal

phenomenon, that was exacerbated by the moderate stenoses of the RCA. Provocative testing was not completed to confirm the coronary steal phenomenon given the large size of the RCA-RV fistula warranted closure irrespective of symptoms. Heart team meeting favored surgical ligation with concurrent LAD and RCA coronary artery bypass due to the patient's low surgical risk, the large size of RCA-RV conus fistula making percutaneous closure more challenging, and concurrent double vessel coronary artery disease. Successful surgical ligation was confirmed with intraoperative epicardial ultrasound color Doppler (Supplemental Figure S2). Postoperative electrocardiogram demonstrated resolution of inferior limb-lead T wave inversion (Supplemental Figure S1B), supporting the role of the RCA-RV fistula and moderate RCA stenoses as the culprit in this presentation. New anterolateral T wave inversion is noted postoperatively, with a subsequent CTCA demonstrating patent left internal mammary artery graft to the LAD. Benign repolarization abnormalities are recognized after coronary artery bypass surgery.⁴

After closure, clinical monitoring is imperative as fistula recurrence is common, either through failure of closure device or neo-fistula (although given that 20% of patients have multiple fistulas, some recurrence is suspected to be secondary to increasing size of pre-existing subclinical fistulas). If closure is not initially indicated, monitoring for complications such as pulmonary hypertension and development of endocarditis or ischemia is warranted. This case demonstrates a rare situation of a symptomatic coronary-cameral fistula with identification of a further nonculprit fistula and severe nonculprit atherosclerotic disease. Careful consideration of the clinical features is required to treat the appropriate target, as fistulas can be nonculprit bystanders that may not require intervention (the septal fistula in this case). In this case, features that make coronary steal likely (large fistula caliber, Sakakibara type A, coronary-cameral to a low-pressure chamber, and concurrent moderate RCA stenoses) and electrocardiogram supportive of

Keywords: coronary artery fistula; decision-making; fistula ligation.

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<https://doi.org/10.1016/j.jscai.2024.101300>

Received 20 December 2023; Received in revised form 9 January 2024; Accepted 9 January 2024

Available online 26 March 2024

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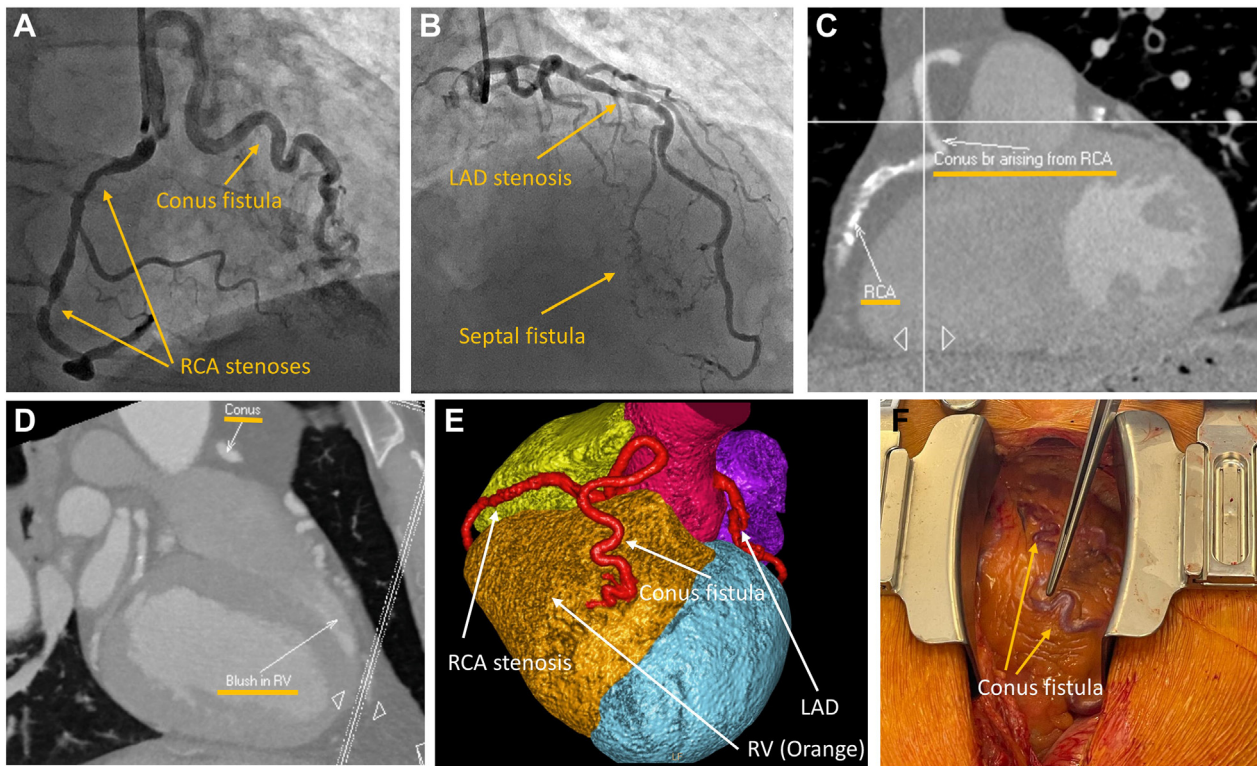


Figure 1.

(A) Coronary angiogram: right coronary artery-right ventricle (RCA-RV) conus artery coronary-cameral fistula. Moderate stenoses in proximal to mid RCA were demonstrated. (B) Coronary angiogram: septal artery to right ventricle coronary-cameral fistula and severe proximal left anterior descending artery (LAD) stenosis. (C) Computed tomography coronary angiography (CTCA) demonstrating the origin of the RCA-RV conus fistula and dense calcification of the proximal to mid-right coronary artery. (D) CTCA contrast blush confirming right ventricular termination of RCA-RV fistula. (E) CTCA 3D reconstruction of the RCA-RV coronary-cameral fistula. (F) Surgical image of the RCA-RV fistula.

territory involvement indicated RCA-RV fistula closure with an excellent clinical result.

Declaration of competing interest

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding sources

This work was not supported by funding agencies in the public, commercial, or non-for-profit sectors.

Ethics statement and patient consent

This work adhered to the relevant ethical guidelines, and patient consent was obtained.

Supplementary material

To access the supplementary material accompanying this article, visit the online version of the *Journal of the Society for Cardiovascular Angiography & Interventions* at [10.1016/j.jscai.2024.101300](https://doi.org/10.1016/j.jscai.2024.101300).

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