

IMAGING

CASE REPORT: CLINICAL CASE

Acquired Coronary Cameral Fistula Mimicking Prosthetic Aortic Valve Regurgitation



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ABSTRACT

A coronary cameral fistula (CCF) is an abnormal communication between a coronary artery and any of the 4 cardiac chambers. Although congenital cases are more common, acquired CCFs, particularly after cardiac surgery, are rare. We present an unusual case of acquired CCF that occurred after bioprosthetic aortic valve replacement and septal myectomy and that mimics prosthetic regurgitation. (JACC Case Rep. 2024;29:102555) © 2024 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/>).

HISTORY OF PRESENTATION

A 57-year-old man presented with exertional dyspnea and was given a diagnosis of severe aortic stenosis and moderate aortic regurgitation (Figures 1A to 1C, Video 1). His blood pressure was 143/92 mm Hg on admission, and a 3/6 grade ejection murmur was noticed in the aortic valve auscultation area during physical examination. Preoperative transthoracic echocardiography (TTE) showed an abnormal hypertrophic septal base and systolic anterior motion (SAM)

of a mitral leaflet, which caused left ventricular outflow tract (LVOT) obstruction (Figures 2A and 2B). The preoperative coronary angiogram was normal. The surgical procedure involved septal myectomy and implantation of a Magna Ease (Edwards Lifesciences) 25-mm prosthesis. Postoperative TTE at discharge demonstrated normal prosthetic valve leaflet motion. The SAM disappeared after surgery, and there was no obvious blood flow obstruction in the cardiac cavity (Video 2). The presence of diastolic flow within the LVOT from the prosthesis was noted (Figures 3A and 3B, Video 3) and was initially interpreted as aortic regurgitation of unknown mechanism.

LEARNING OBJECTIVES

- To be able to make a differential diagnosis of acquired CCF after aortic valve replacement.
- To emphasize the role of TEE in the diagnosis of cardiac complications after aortic valve replacement.

PAST MEDICAL HISTORY

This patient had hypertension and a history of lumbar spine fracture surgery.

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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****CCF** = coronary cameral fistula**LV** = left ventricular**LVOT** = left ventricular outflow tract**SAM** = systolic anterior motion**TEE** = transesophageal echocardiography**TTE** = transthoracic echocardiography**DIFFERENTIAL DIAGNOSIS**

In this scenario, consideration should be given to the possibility of aortic valve bioprosthesis dysfunction and paravalvular leaks, both of which may necessitate subsequent surgical reintervention.

INVESTIGATIONS

Transesophageal echocardiography (TEE) was performed to elucidate the mechanism behind this diastolic retrograde flow across the aortic valve. We conducted a multiangle examination of the aortic valve at the midesophageal level. Ultimately, in the 5-chamber view, diastolic flow was identified originating from the basal segment of the interventricular septum, closely adjacent to the aortic valve. It ascended obliquely, causing an impact on the anterior leaflet of the aortic valve bioprosthesis, then folded back into the LVOT (**Figure 4, Video 4**). On meticulous examination of the aortic valve surroundings, no evidence of paravalvular regurgitation was observed. Because a septal myectomy was performed, this flow was concluded to be arising from a coronary cameral fistula (CCF).

MANAGEMENT

Given the absence of symptoms indicative of myocardial ischemia and corresponding electrocardiographic

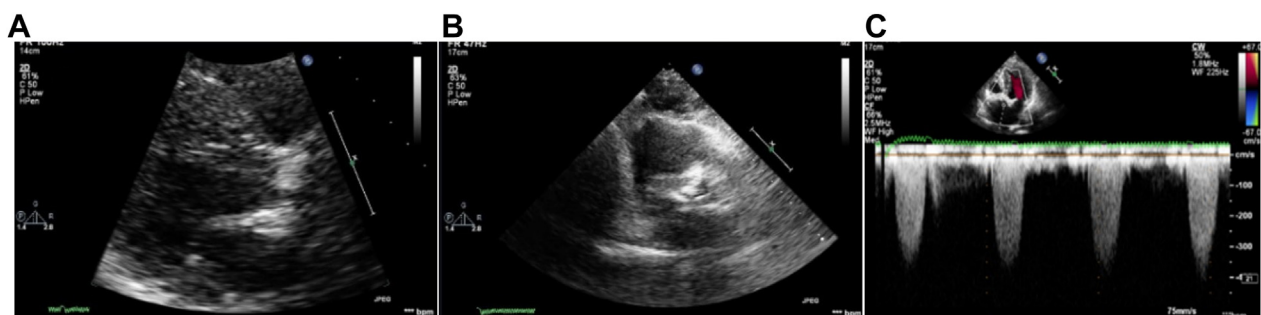
manifestations, further intervention was not performed, and the patient was discharged.

DISCUSSION

An acquired CCF may occur after a diagnostic or therapeutic endovascular coronary intervention or subsequent to thoracic trauma, permanent pacing, endomyocardial biopsy, or cardiac surgery.¹ These fistulas arise from the right coronary artery in approximately 55% of patients, whereas 35% of CCFs arise from the left side. Septal myectomy, when performed in conjunction with an aortic valve replacement, portends a heightened risk for CCF development.²

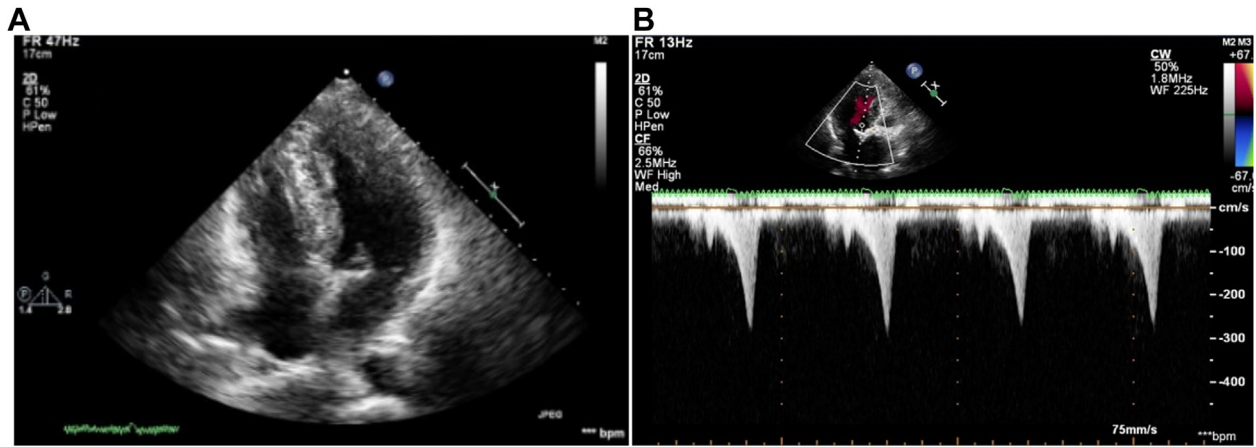
Diastolic flow arising from the vicinity of a prosthetic valve is most likely to be prosthetic regurgitation or paravalvular leaks. In the present case, because of the CCF's originating very close to the aortic valve and forming a retrograde pathway on the aortic valve, it became challenging to differentiate it from aortic valve regurgitation and paravalvular leaks. Furthermore, all 3 conditions involve diastolic blood flow, thus further complicating the discernment process. Paravalvular leak jets generally are seen to track along the left ventricular (LV) wall, unlike in the present case, where the jet was directed to and along the aortic annular plane (**Figures 5A to 5C**).

Large CCFs, particularly those draining into the right ventricle or atria, can lead to hemodynamic

FIGURE 1 Preoperative Transthoracic Echocardiography Images Indicate Aortic Valve Stenosis

The (A) parasternal long-axis and (B) short-axis views revealed prominent aortic valve sclerosis with restricted opening. (C) Continuous-wave (CW) Doppler imaging estimated a peak systolic velocity across the aortic valve of approximately 4 m/s, with a mean pressure gradient of 39 mm Hg. Using the continuity equation method, the calculated effective valve orifice area was approximately 1.0 cm². bpm = beats/min; FR = frequency; 2D = 2-dimensional.

FIGURE 2 Preoperative Transthoracic Echocardiography Images Indicate Left Ventricular Outflow Tract Obstruction

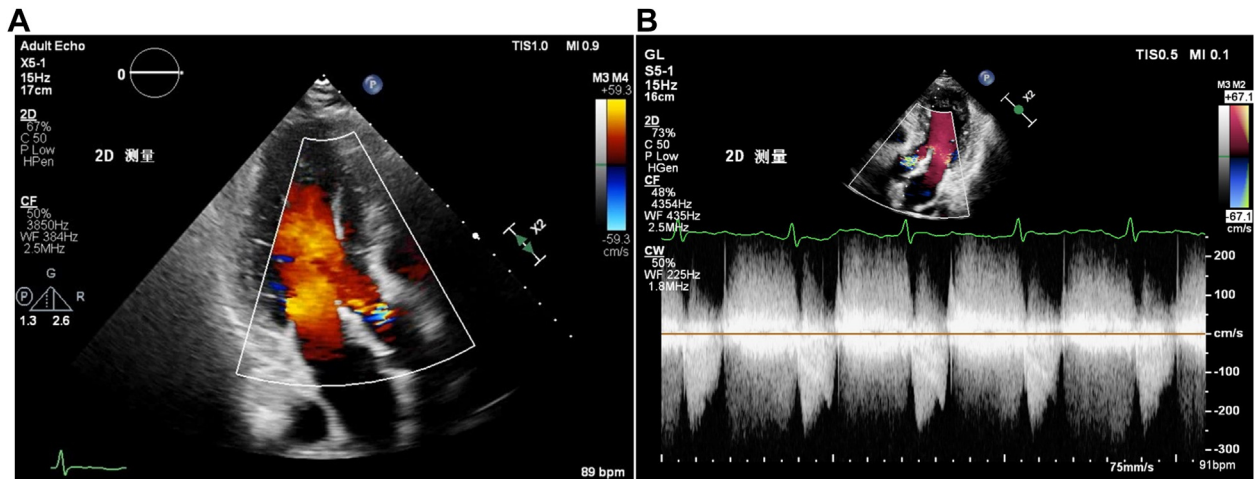


(A) The apical 4-chamber view revealed pronounced thickening of the interventricular septum, particularly at the basal segment. (B) The blood flow spectrum in the left ventricular outflow tract demonstrated a dagger-shaped pattern on continuous-wave (CW) Doppler, with a peak pressure gradient of 31 mm Hg. C = contrast; CF = color flow; HPen = high penetration; P = persistence; WF = wall filtering; other abbreviations as in Figure 1.

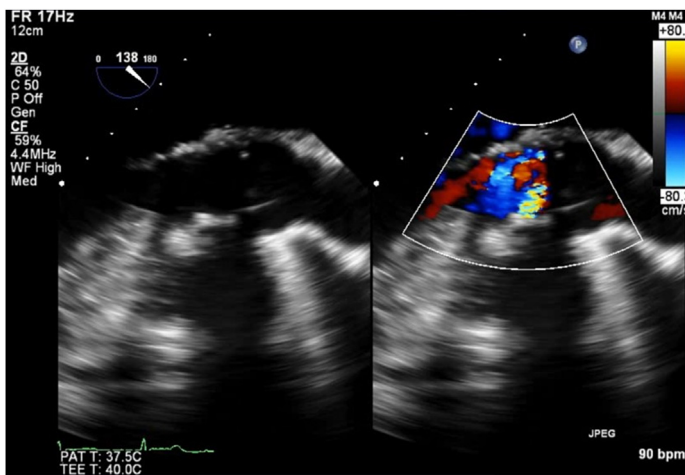
consequences such as congestive heart failure, coronary steal, and arrhythmias. Conversely, CCFs draining into the left ventricle generally impose a limited hemodynamic burden. In our case, the small size of the CCF and the absence of coronary steal, indicated by the lack of new onset regional wall motion

abnormalities or ischemic symptoms, justified a conservative approach without intervention. However, because restricted effort after surgery may mask ischemia-related symptoms, a noninvasive assessment such as strain imaging or treadmill exercise testing is recommended. Large CCFs causing

FIGURE 3 Postoperative Transthoracic Echocardiography Images



(A) The color flow (CF) image revealed an abnormal diastolic blood flow originating from the aortic valve, resembling aortic valve regurgitation. (B) Continuous-wave (CW) Doppler estimated a diastolic flow velocity of >2 m/s. Abbreviations as in Figures 1 and 2.

FIGURE 4 Postoperative Transesophageal Echocardiography Images

The transesophageal echocardiographic (TEE) examination revealed abnormal diastolic blood flow originating from the basal segment of the interventricular septum, closely adjacent to the subaortic valve. It propelled forward, causing an impact on the anterior leaflet of the aortic valve bioprosthesis, then folded back into the left ventricular outflow tract. Abbreviations as in [Figures 1 and 2](#).

myocardial steal, hemodynamic instability, or LV volume overload warrant evaluation by coronary angiography and may require surgical or percutaneous repair.

FOLLOW-UP

At the 6-month follow-up, the patient remained asymptomatic, with satisfactory ventricular and prosthetic valve function. The CCF persisted on TTE.

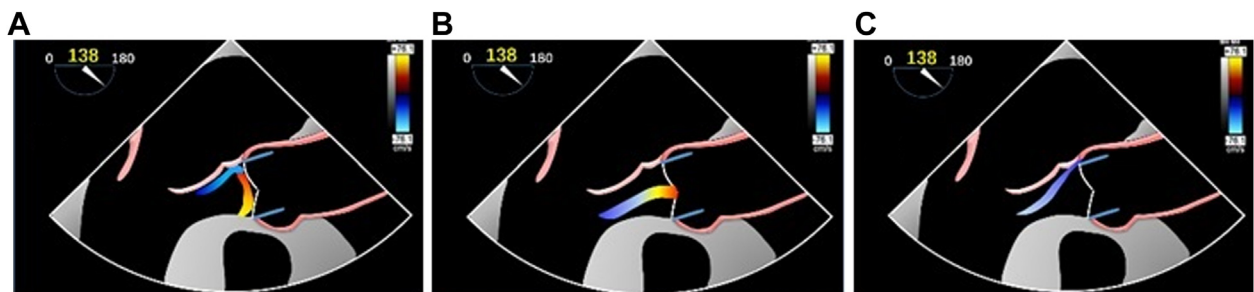
CONCLUSIONS

In summary, we report a case of CCF in a patient who underwent an uneventful aortic valve replacement with septal myectomy. The timeline of the case is illustrated as in [Table 1](#). The TTE diagnosis of this entity is challenging and depends on operator awareness. CCF can be mistaken for regurgitation, paravalvular leaks, or ventricular septal defects, sometimes leading to unnecessary reinterventions. TEE provides more confirmative information, and coronary angiography is rarely needed for small fistulas.

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

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FIGURE 5 Illustrative Diagrams of Transesophageal Echocardiography

The mid-esophageal long-axis view demonstrating (A) an acquired coronary cameral fistula (the present case), (B) aortic valve bioprosthesis regurgitation, and (C) paravalvular regurgitation.

TABLE 1 Timeline of the Case

Date	Events
June 22, 2022	A 57-year-old man presented with exertional dyspnea. TTE showed severe aortic stenosis and moderate aortic regurgitation. LVOT obstruction caused by a hypertrophic septal base and SAM of a mitral leaflet was also noted.
June 23, 2022	The patient underwent septal myectomy and aortic prosthesis implantation.
July 1, 2022	Diastolic flow within the LVOT from the prosthesis was noticed on TTE.
July 2, 2022	TEE showed the presence of diastolic flow originating from the basal segment of interventricular septum, closely adjacent to the aortic prosthesis.
December 18, 2022	The patient remained asymptomatic with satisfactory ventricular and prosthetic valve function. The CCF persisted on TTE.

CCF = coronary cameral fistula; LVOT = left ventricular outflow tract; SAM = systolic anterior motion; TEE = transesophageal echocardiography; TTE = transthoracic echocardiography.

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KEY WORDS acquired coronary cameral fistula, aortic valve replacement, echocardiography, septal myectomy

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