CJC Open 6 (2024) 755–758

Case Report

# Acute Heart Failure Due to a Fistula Between the Anastomotic Pseudoaneurysm and the Right Atrium Following a Previous Aortic Dissection Repair

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**Intracardiac perforation of an aortic aneurysm is rare, whereas ruptures of it into the thoracic cavity or pericardial spaces are common fatal complications. Herein, we report the case of a patient with acute heart failure due to a fistula between the anastomotic pseudoaneurysm and the right atrium, following a previous aortic dissection repair. The cause of heart failure after cardiac surgery should be interrogated promptly, using multiple modalities, and the possibility of an aorto-cameral fistula should be considered. Right heart catheterization (RHC) may be an important adjunctive imaging modality to assess occult etiologies of heart failure by measuring right-sided cardiac oxygen saturations.**

## Case Presentation

A 73-year-old man with a history of diabetes mellitus presented to the emergency department with a 10-day history of diarrhea, abdominal distention, and dyspnea. Eighteen years prior, he had undergone an ascending aortic replacement for a Stanford type A aortic dissection. He had been followed-up at the previous institution for several years after surgery, but he self-interrupted the postoperative visits. On presentation, the patient was afebrile, with a blood pressure of 136/63 mm Hg, an irregular heart rate of 133 beats per minute, a respiratory rate of 32 breaths per minute, and an oxygen saturation of 98% on room air. Physical examination revealed decreased breath sounds in the lower chest, leg edema, and abdominal swelling. No cardiac murmurs were noted.

Chest radiography revealed dilation of the right first and second arches, obliterated costophrenic angle, and mild pulmonary congestion. Electrocardiography demonstrated atrial fibrillation with a Q wave in lead III, but no significant ST-segment changes were identified. Transthoracic echocardiography (TTE) revealed a preserved left ventricular ejection fraction, without a shunt or valvular disease, and dilation of the inferior vena cava (IVC). Laboratory examination indicated abnormalities in hepatic and kidney function (total bilirubin: 2.7 mg/dL; aspartate aminotransferase: 1558 U/L; alanine aminotransferase: 988 U/L; blood urea nitrogen: 53.8 mg/dL; and creatinine: 1.50 mg/dL), an elevated brain natriuretic peptide level (490.9 pg/mL), and lactic acidosis (9.4 mmol/L). Computed tomography (CT) showed a normal sinus of Valsalva but an aortic pseudoaneurysm with an 80-mm diameter at the proximal anastomotic site, with ascites and pleural effusion.

The patient was diagnosed with acute congestive heart failure with preserved ejection fraction, which responded poorly to furosemide, and the lactic acidosis progressed (13.9 mmol/L) despite dobutamine hydrochloride administration 6 hours after admission. Given that maintaining systemic circulation (blood pressure of 74/50 mm Hg; heart rate of 148 beats per minute; oxygen saturation of 94%; and lactate: 19.8 mmol/L) 12 hours after admission became difficult, RHC was performed simultaneously with the introduction of mechanical support. RHC revealed elevated right atrial (RA) pressure (a-wave/v-wave/mean pressure = 33/38/27 mm Hg) with a wide pulse pressure (Fig. 1A), and a step-up of oxygen saturation between the IVC and the right atrium (RA), suggesting the presence of a left-to-right shunt. Aortic root angiography

Received for publication August 14, 2023. Accepted January 19, 2024.

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See page 757 for disclosure information.

https://doi.org/10.1016/j.cjco.2024.01.005

**Novel Teaching Points**

- We encountered a case of acute heart failure due to an aorto-right atrium fistula following a previous aortic dissection repair.
- Clinicians should promptly interrogate the cause of heart failure after cardiac surgery using multimodalities, and consider the possibility of an aorto-cameral fistula.
- Right heart catheterization may be an important adjunctive imaging modality that can be used to assess occult etiologies of heart failure.

demonstrated direct inflow of contrast medium from the pseudoaneurysm into the RA (Fig. 1B; Video 1, view video online).

With the diagnosis of low cardiac output syndrome secondary to a fistula formation between the anastomotic pseudoaneurysm and the RA, emergency redo ascending aortic replacement surgery was indicated. After introducing veno-arterial extracorporeal membrane oxygenation (VA-ECMO), preoperative contrast-enhanced CT showed a break in the pseudoaneurysm (Fig. 2A), and transesophageal echocardiography (TEE) revealed continuous shunt flow (Fig. 2B; Video 2, view video online).

Fourteen hours after his admission, his operation was started under VA-ECMO. Intraoperatively, managing the bleeding was difficult due to severe adhesions. Therefore, a cardiopulmonary bypass was established urgently by the femoral artery and the vena cava cannulation. An aperture of approximately 10 mm in diameter was detected between the aortic pseudoaneurysm and the RA (Supplemental Fig. S1), which was ligated with suture. Given that the size and adhesion of the pseudoaneurysm necessitated a new artificial graft, and the presence of a suture cite at the aortic root, an ascending aortic replacement was performed instead of the Bentall procedure. The patient was weaned from cardiopulmonary bypass back to VA-ECMO and transported back to the intensive care unit in critical stable condition. However, his lactic acidosis did not improve, and he died due to multiple organ failure on the day after admission.

## Discussion

Intracardiac perforation of an aortic aneurysm is rare, whereas ruptures of it into the thoracic cavity or pericardial spaces are common fatal complications.[1] More than one-third of reported patients have a history of cardiac surgery, and postoperative adhesions are suggested to contribute to the aorto-cameral fistula.[2] Also, anatomically, it is known to perforate the RA more often than the left atrium, causing congestive heart failure and low cardiac output syndrome arising from a left-to-right shunt.[3] This mechanism is similar to a rupture of the sinus of Valsalva aneurysm.

A continuous murmur is the hallmark of an aorta-cameral fistula. However, this condition is observed infrequently in patients with a large shunt hole, and following cardiac surgery. We believe that the fistula had already formed 10 days prior to his visit and that the shunt volume gradually increased as the aperture expanded. Therefore, possibly, we could not hear its continuous murmur on admission. Although TTE is an essential initial test for screening shunt diseases, it possesses limited utility for evaluating the aorta.[4] TEE is more sensitive than TTE for aortic disease detection, but it is not tolerated

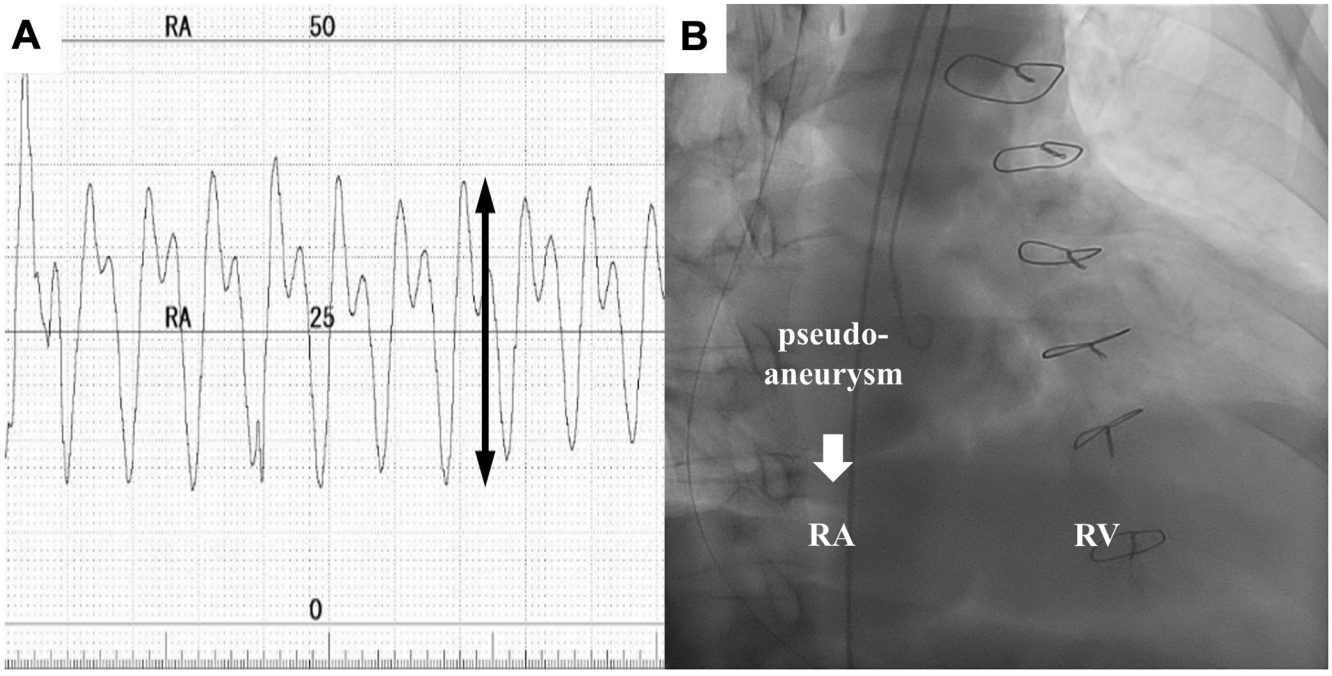


**Figure 1.** (**A**) Right heart catheterization showing elevated right atrial pressure with a wide pulse pressure (**arrow to arrow**). (**B**) An aortic root angiography image of direct inflow of the contrast medium from the aortic pseudoaneurysm into the right atrium (RA; **arrow**). RV, right ventricle.

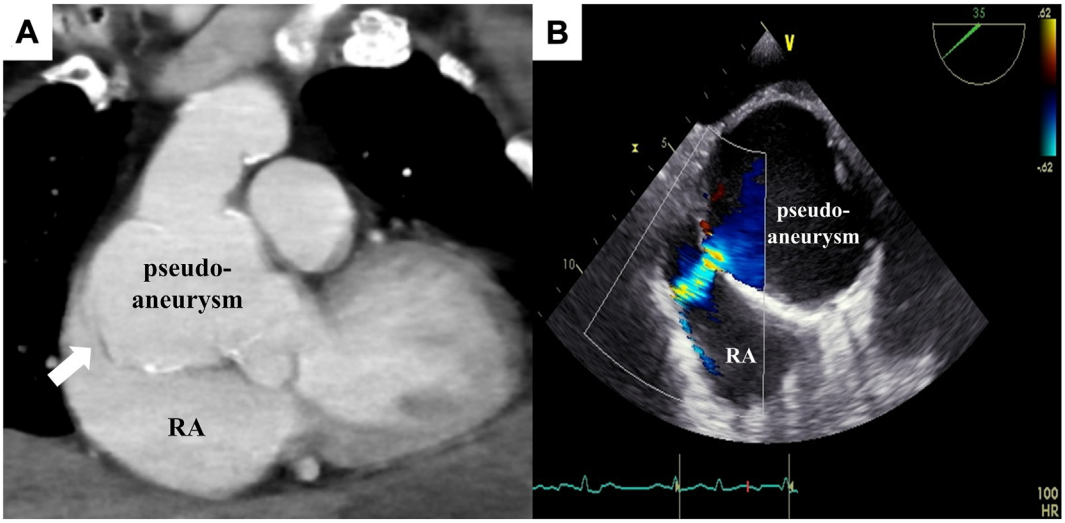


**Figure 2.** (**A**) Contrast-enhanced computed tomography showing a break in the aortic pseudoaneurysm (**arrow**). (**B**) Transesophageal echocardiography demonstrating the aorto-right atrial fistula with continuous shunt flow on color Doppler. RA, right atrium.

well, due to its invasiveness, especially in a patient who is hemodynamically unstable without intubation. Contrast-enhanced CT requires a short acquisition time, yields a high spatial resolution, and can provide accurate anatomic information about the morphology of aortic aneurysms or fistulas; however, performing it in patients with poor renal function often is challenging. Magnetic resonance imaging possesses superior temporal resolution and enables quantification of cardiac function, including the degree of shunting if the patient's condition is stable.[5]

In retrospect, we believe that prioritizing TEE or contrast-enhanced CT could have identified the underlying pathology more easily in this case. However, on admission, recognizing the lactic acidosis as a sign of cardiogenic shock was difficult because of the preceding abdominal symptoms and preserved cardiac function. Hence, RHC was the key examination that revealed the left-to-right shunt, which was not clarified by noninvasive examinations. Previous studies have demonstrated a reduction of in-hospital mortality with RHC, in patients with cardiogenic shock, before initiation of mechanical support, and a significant reduction in low cardiac output syndrome in an early RHC group ($<$ 2 days), compared to that in a late RHC group ($\geq$ 2 days).[6] Although RHC is also safer to perform than left heart catheterization in patients with aortic aneurysms, it cannot evaluate coronary arteries in the setting of cardiogenic shock. In fact, we initially planned to conduct only RHC, because echocardiographic findings indicated that ischemic heart disease was unlikely. As a result, aortic root angiography was added for suspected aorto-cameral fistula.

The 2022 American College of Cardiology/American Heart Association Guideline for the Diagnosis and Management of Aortic Disease also recommends that long-term surveillance imaging with CT be conducted annually if the patient is stable and has undergone open repair for acute aortic dissection.[7] If our patient had not been lost to follow-up, the aortic pseudoaneurysm might have been detected earlier, preventing its rupture.

## Conclusions

This case highlights the need for clinicians to promptly clarify the cause of heart failure after cardiac surgery using multimodalities, keeping the possibility of aorto-cameral fistulas in mind. RHC may be an important adjunctive imaging modality to assess occult etiologies of heart failure.

## Ethics Statement

This study was approved by the Omihachiman Community Medical Center Ethics Committee, Omihachiman, Japan.

## Patient Consent

The authors confirm that written informed consent was obtained from the patient's family.

## Funding Sources

The authors have no funding sources to declare.

## Disclosures

The authors have no conflicts of interest to disclose.

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## Supplementary Material

To access the supplementary material accompanying this article, visit *CJC Open* at https://www.cjcopen.ca/ and at https://doi.org/10.1016/j.cjco.2024.01.005.