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CASE REPORT
Other

Aorto-right pulmonary venous fistula after mitral valve replacement for prosthetic mitral valve infective endocarditis: a case report

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Background Aorto-cavitary fistula is a rare condition, and the most common underlying aetiology is infective endocarditis (IE) of the native or the prosthetic aortic valve. We report a case of aorto-right inferior pulmonary venous fistula following redo mitral valve replacement (MVR) for prosthetic mitral valve IE.

Case summary A 74-year-old woman underwent urgent redo MVR for prosthetic mitral valve IE. The post-operative course was complicated with heart failure and mediastinal haematoma compressing the left atrium. The haematoma was surgically removed and laceration of the left atrial wall was suture ligated; this was attributed to the surgical trauma dissection of the adhesive tissues. One-week post-operatively, a continuous murmur emerged, which prompted an evaluation of the left to right shunt. Transthoracic echocardiography revealed an echolucent area posterior to the aorta, with continuous flow on colour Doppler. Three-dimensional computed tomography showed a fistula between the aorta and the right inferior pulmonary vein. There was a high risk involved in surgical management; therefore, she was managed medically. Fortunately, the continuous murmur and echolucent space disappeared after 6 months. The fistula was considered to be obstructed by spontaneous thrombus formation in the narrowed segment of the fistula tract.

Discussion The cause of fistula was attributed to possible surgical trauma in the presence of infection. The present case was unique, as it showed spontaneous healing of an aorto-cavitary fistula, which is very rare. The patient was alive with good health status, 6 years after the MVR.

Keywords Case report • Continuous murmur • Aorto-cavitary fistula • Infective endocarditis • Mitral valve replacement • Surgical trauma

Learning points

- An aorto-cavitary fistula is rare and is usually associated with prosthetic or native aortic valve infective endocarditis. An aorto-cavitary fistula can result from surgical trauma after complex surgical procedures such as redo mitral valve replacement.
- Aorto-cavitary fistulae usually require surgical repair. However, surgical and in-hospital mortality rates are high. Occasionally, these fistulae may resolve spontaneously.
- Careful clinical examination and the use of multimodality imaging in the assessment of post-operative complications are important.

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Introduction

Aorto-cavitary fistula is a rare condition. Its most common underlying aetiology is infective endocarditis (IE) of the native or prosthetic aortic valve.[1–3] Aortic dissection,[4] aneurysm of the ascending aorta,[5] and post-surgical trauma[6–8] may also lead to aorto-left atrial (LA) fistula. Recognition of this condition is becoming very important due to increase in complex aortic reconstructive surgery and redo valvular surgeries. Here, we have reported a case of aorto-pulmonary venous fistula following redo mitral valve replacement (MVR) for prosthetic mitral valve IE.

Timeline

At 42 years of age (32 years before)	Mitral valve replacement (MVR) with Hall–Kaster mechanical valve prosthesis.
At 68 years of age (6 years before)	Infective endocarditis with *Streptococcus agalactiae*. Medical treatment was successful.
At 73 years of age (1 year before)	Gradual development of congestive heart failure (CHF) and haemolytic anaemia due to perivalvular mitral regurgitation and tricuspid regurgitation.
At 74 years of age (Day 27)	Elective redo MVR was scheduled. However, she was admitted due to fever and shaking chill 2 days before the scheduled admission date. Blood cultures grew *Streptococcus constellatus*. Intravenous aminobenzyl penicillin was started at 8 g/day.
Twenty-seven days after admission (Day 0)	Urgent MVR and tricuspid valve replacement (TVR) were performed because of worsening of heart failure.
Day 0 after the surgery	The patient developed mediastinal haematoma that compressed the left atrium, leading to shock and CHF. The haematoma was successfully removed, and the patient's condition improved.
Day 7 after the surgery	A continuous murmur was noted. Three-dimensional computed tomography resulted in a diagnosis of aorto-right inferior pulmonary venous fistula. Medical treatment was performed due to high surgical risks.
Day 120 after the surgery	Discharged home.
Six months after the surgery	The continuous murmur disappeared. Previous echolucent structure posterior to the aortic wall became echodense and continuous flow on Doppler echo in the structure also disappeared. Thus, we concluded that the fistula healed spontaneously.
Six years after the surgery	The patient is alive and doing well.

Case presentation

A 74-year-old female patient was admitted to our hospital due to fever, shaking chills, and positive blood cultures.

Her medical history was remarkable for MVR with Hall–Kaster single tilting disc mechanical valve for rheumatic mitral valve disease at the age of 42. Twenty-six years after the MVR (at age 68), she experienced an episode of IE caused by *Streptococcus agalactiae* that was successfully treated by medical therapy. Her daily medications included 0.125 mg digoxin, 40 mg furosemide, 25 mg spironolactone, 2 mg warfarin, and 50 mg ferrous citrate, once a day. She gradually developed dyspnoea on exertion 1 year prior to admission (31 years after the MVR). One month prior to admission, transthoracic echocardiography (TTE) revealed moderate to severe perivalvular mitral regurgitation (MR), pulmonary hypertension, and moderate tricuspid regurgitation (*Figure 1A* and *B*). Transtricuspid pressure gradient was 37 mmHg. Left ventricular ejection fraction was 60%. Laboratory data revealed low haemoglobin, elevated lactic dehydrogenase (LDH) level, and low haptoglobin level, which were consistent with mechanical haemolytic anaemia due to perivalvular MR. Consequently, she was scheduled for elective surgery. Two days prior to the scheduled date of admission, however, she developed fever with rigours and was thus admitted to the emergency room. Her height and weight were 140 cm and 32 kg, respectively; blood pressure was 130/80 mmHg; heart rate was 93 b.p.m. (chronic atrial fibrillation); respiratory rate was 24 breaths per minute; and body temperature was 38.1°C. Her initial blood tests were remarkable for haemolytic anaemia [haemoglobin: 7.3 g/dL (11.8–14.8 g/dL), mean corpuscular volume: 101 fL (84–98 fL), LDH: 715 IU/L (124–222 IU/L), haptoglobin: 2 mg/dL (58–160 mg/dL)]; renal failure [creatinine level: 1.40 mg/dL (0.46–0.76 mg/dL) and estimated glomerular filtration rate: 28.9 mL/min/1.73 m^2 (>90 mL/min/1.73 m^2)]; signs of inflammation [C-reactive protein: 8.31 mg/dL (<0.39 mg/dL) and increased white blood cell: 10 200/μL with a left shift (3300–8600/μL)]. Multiple blood cultures grew *Streptococcus constellatus*. Therefore, 8 g/day intravenous aminobenzyl penicillin was initiated under the diagnosis of IE. Although repeated TTE (performed on the day of admission and repeated weekly thereafter) did not reveal vegetation or perivalvular abscess, congestive heart failure (CHF) progressively worsened due to increased perivalvular MR. Transtricuspid pressure gradient increased to 70 mmHg, and her chest roentgenogram revealed pulmonary oedema. A transoesophageal echocardiogram (TOE) was attempted but the patient could not tolerate the procedure. Because of refractory CHF, she underwent urgent MVR and tricuspid valve replacement (TVR) surgery 27 days after admission to the hospital. The mitral valve was partially detached at the posterior portion of the sewing ring. Vegetation and

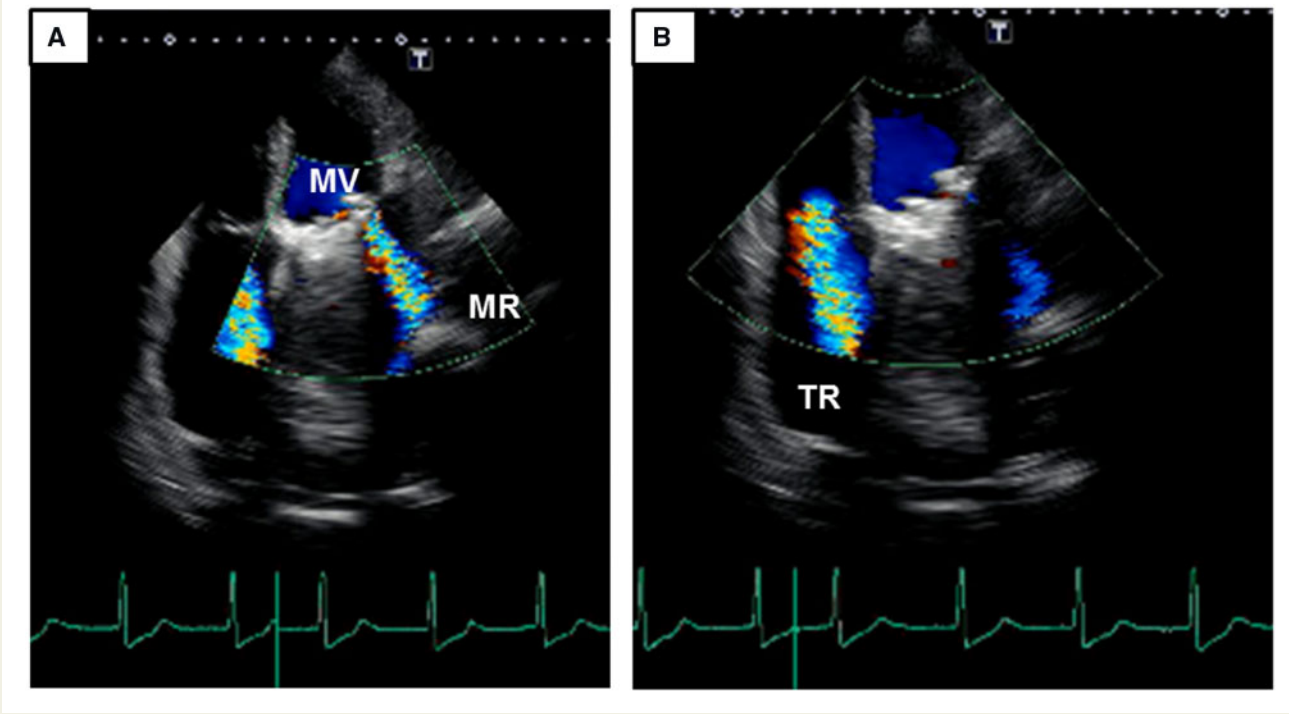

Figure 1 (*A* and *B*) A colour Doppler in 4-chamber view showing perivalvular mitral regurgitation (a) and tricuspid regurgitation (b). MR, mitral regurgitation; MV, mechanical mitral valve; TR, tricuspid regurgitation.

perivalvular abscess were not identified. Furthermore, because the tricuspid annulus was extremely dilated, valve repair was technically challenging. Thus, TVR with St. Jude tissue valve (31 mm) was performed in addition to MVR with Carpentier-Edwards Magna bioprosthesis (25 mm). Because there was extremely severe adhesion of the tissues from the previous MVR, the LA wall was damaged during manipulation, thus requiring suture haemostasis of the posterior LA wall. Mild bleeding from the posterior wall of the ascending aorta was successfully managed with a tissue sealing sheet containing fibrinogen and thrombin.

Shortly after the patient was transferred to the intensive care unit (Day 0 of the surgery), she developed refractory heart failure and shock. Transthoracic echocardiography revealed possible thrombus occupying the LA (*Figure 2A*). Therefore, the patient was taken back to the operating room. At the time of surgery, the apparent thrombus in the LA (*Figure 2B*) turned out to be a haematoma on the exterior surface of the posterior LA wall, which caused a compression of the cavity (*Figure 2A and B*). A tear in the posterior LA wall, attributed to surgical trauma secondary to severe adhesion, was repaired with a simple suture.

After surgery, compression of the LA improved, and the haemodynamic condition of the patient improved gradually (Fig 2C). However, CHF symptoms persisted 1-week post-operatively, and a *de novo* continuous murmur on the third sternal border was noted, suggesting possible left–right shunt. Transthoracic echocardiography revealed small tubular echolucent structures posterior to the aortic wall, wherein a continuous flow was detected by colour Doppler (*Figure 3B and C*, Supplementary material online, *Video S2*), which was not observed preoperatively (*Figure 3A*; Supplementary material online, *Video S1*). Determining the detailed anatomy, with respect to the location of the origin of the left-right shunt and the location of the shunt flow drainage, was a challenge. Contrast-enhanced computed tomography (CT) revealed that the fistula originated from the posterior wall of the aorta and travelled between the aorta and the left atrium up to its ceiling, and thereafter drained into the right inferior pulmonary vein (RIPV) (*Figure 4A–4F*). Given the patient's high surgical risk, the heart team decided on medical management for the patient. Her CHF was successfully managed with dobutamine (~10 μg/kg/min), carperitide (0.1 μg/kg/min), and furosemide (40–60 mg/day). She was discharged on the 120th hospital day with New York Heart Association (NYHA) Class II. Discharge medications were: 40 mg of furosemide, 50 mg of spironolactone, 100 mg of amiodarone, and 1.5 mg of warfarin. Six months post-operation, the continuous murmur had disappeared and the echo-free space with continuous flow posterior to the aortic wall had likewise spontaneously disappeared (*Figure 3D*), and her condition has been stable in NYHA functional Class II.

Discussion

Aorto-cavitary fistula is a relatively rare condition, and the most common underlying mechanism is IE of the native or prosthetic aortic valve.[1] Notably, aorto-cavitary fistula following MVR is extremely

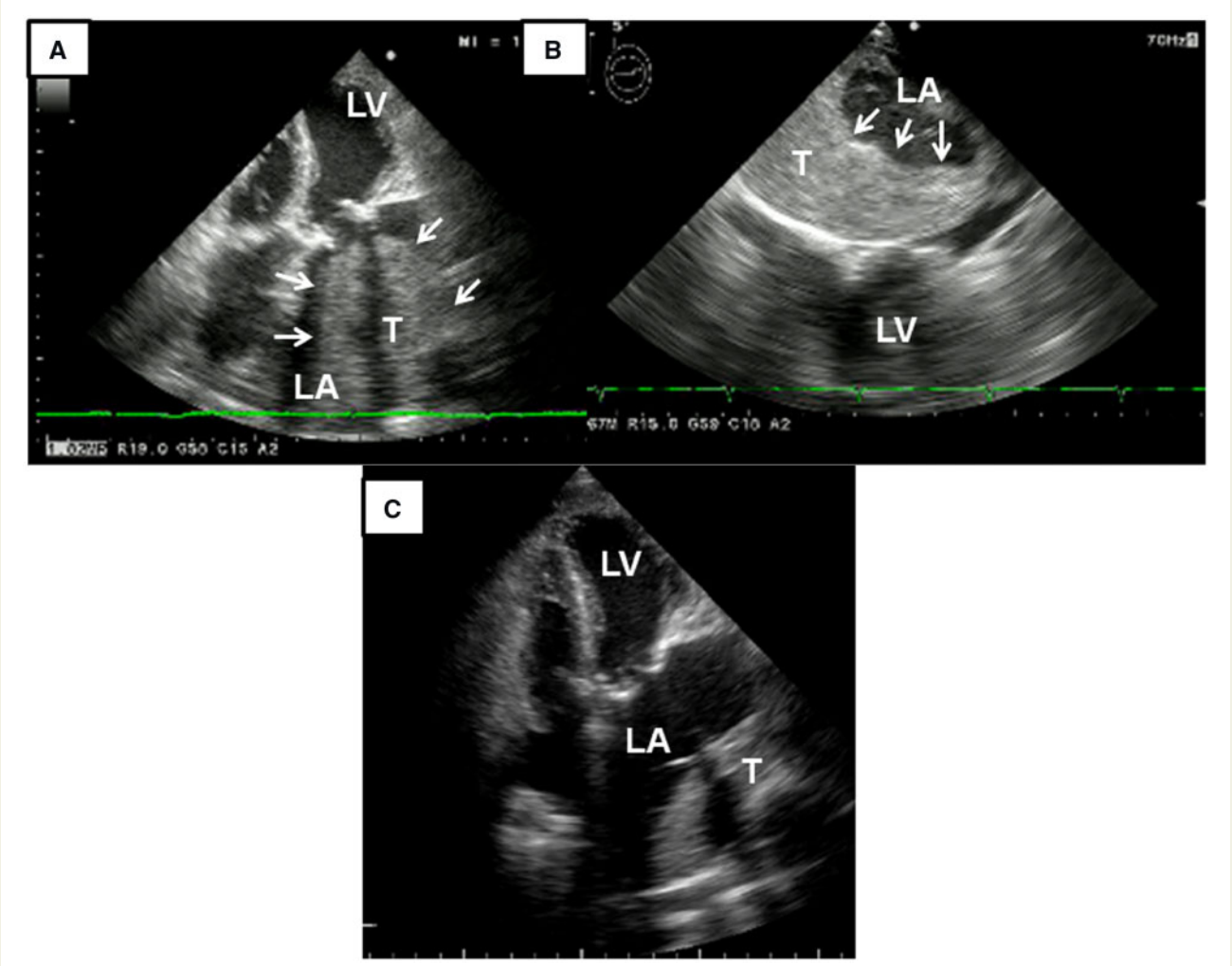

Figure 2 (*A*) An apical four-chamber view of the transthoracic echocardiogram showing possible thrombus (white arrows) in the left atrium that was actually a mediastinal haematoma compressing the left atrium from the outside. (*B*) A mid-oesophageal transverse view of the transoesophageal echocardiogram at the left atrial level showing possible thrombus (white arrows). The right ventricle and atrium collapsed and were not well-visualized because this image was taken during the extra-corporeal circulation at the time of repeat surgery. (*C*) After removal of the haematoma, the left atrium expanded, but residual haematoma was noted. LA, left atrium; LV, left ventricle; T, possible thrombus that was actually a mediastinal haematoma.

rare. Anguera *et al.*[1] did not observe any case of aorto-cavitary fistula following mitral valve IE (0 of 1256 cases). Usually, the fistula is closely associated with periannular abscesses and directly drains into the adjacent cardiac cavities. Among 76 cases of aorto-cavitary fistula, 19 (25.0%) had aorto-LA fistula.[1] On the other hand, Raut *et al.*[7] and Yesin *et al.*[8] reported cases wherein aorto-LA fistula was detected late following mechanical MVR. Their cases were not related to IE, and the authors had hypothesized that the traumatic damage to the aortic wall was caused by the previous surgery. Although our case developed aorto-RIPV fistula after redo surgery for prosthetic IE, the clinical course, intra-operative findings, and the morphology of the long tract of the fistula were not consistent with IE as the aetiology. Rather, traumatic damage to the aortic, LA, and pulmonary venous walls during surgical manipulation of the adhesive tissue, may have resulted in a haematoma in the posterior space of the left atrium, and from which a fistula possibly formed, between the posterior wall of the ascending aorta and the RIPV. To the best of our knowledge, aorto-RIPV fistula has not yet been reported.

Diagnosis of aorto-cavitary fistula is usually made by TTE or TOE.[1] Sometimes, angiography or CT scan is required.[7] In this present case, evaluation with multimodality imaging, prompted by the continuous murmur, was the key to establishing the final diagnosis. Because of the long and tortuous fistula tract, it was difficult to visualize the whole fistula by TTE and TOE. Therefore, it required the use of 3D CT.

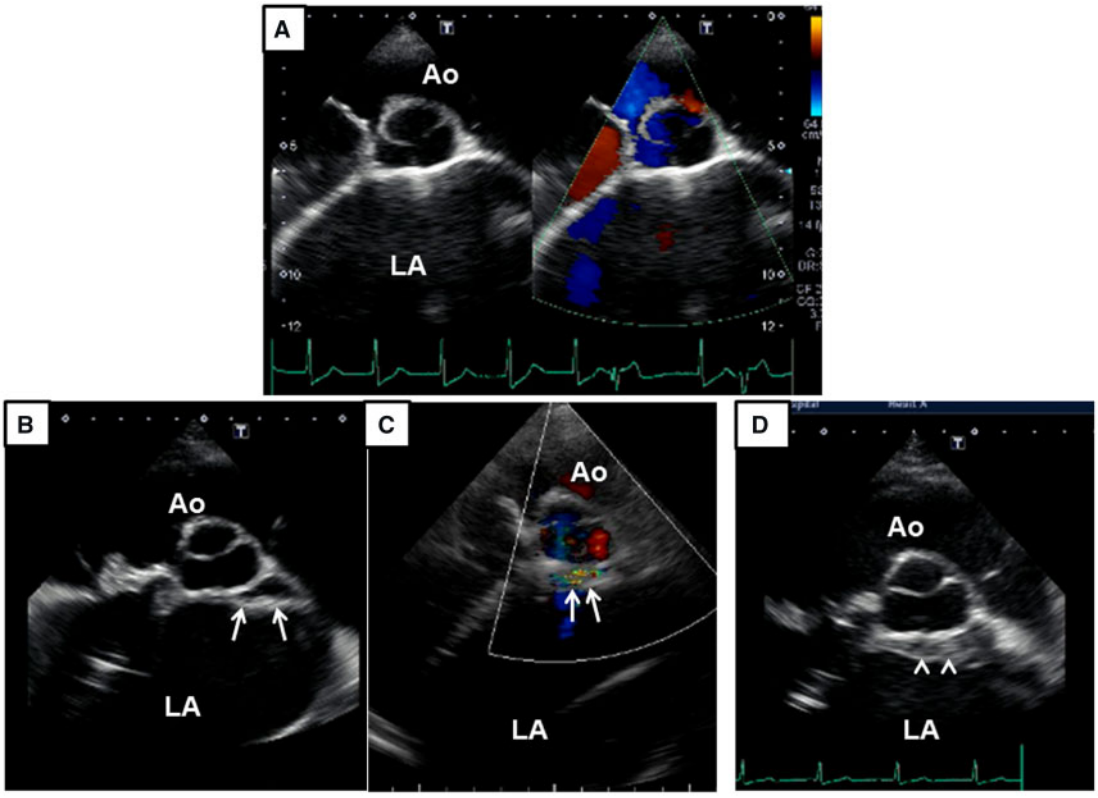

Figure 3 (*A*) A short-axis view of pre-operative transthoracic echocardiography of the aortic valve level (left). There was no echolucent space posterior to the aorta or abnormal continuous flow by colour Doppler (right panel; Supplementary material online, *Video S1*). (*B* and *C*) One-week post-operatively, an echolucent space posterior to the aorta (white arrows in *B*) was noted, wherein a colour Doppler revealed continuous flow (white arrows in *C*; Supplementary material online, *Video S2*). (*D*) Six months post-operation. The previous echolucent space posterior to the aortic wall was occupied by echogenic material (possible organized thrombus, white arrow heads) and there was no continuous flow by colour Doppler (Supplementary material online, *Video S3*). Ao, aorta at the aortic valve level; LA, left atrium.

Also noteworthy was the clinical course of our patient. As per the study of Anguera *et al.*,[1] 66 of 76 patients with aorto-cavitary fistula required surgery. However, the mortality rate in their cases was as high as 42%. Among the 10 patients who did not undergo surgery, three died during admission. Among the seven who survived without surgery, 80% of patients developed major complications during the 2-year follow-up period.[1] Fortunately, in our case, our patient was successfully managed medically without requiring surgery. Moreover, the fistula spontaneously healed with possible thrombus formation at the narrow segment of the fistula tract (*Figure 3D*; Supplementary material online, *Video S3*). The patient is alive and doing well 6 years after discharge.

Conclusions

In conclusion, post-operative aorto-RIPV fistula following MVR is rare. Careful clinical examination and assessment of post-operative complications using multimodality imaging are very important in achieving a favourable clinical outcome.

Lead author biography

Kazuhito Hirata graduated from Kumamoto University Medical School in 1982. During 1982–86, he completed post-graduate medical education program at Okinawa Chubu Hospital, Okinawa, Japan. During 1988–90, he served as a Fellow in Cardiology, Division of Cardiology, The Ohio State University, USA. In 1990, he served as Staff Cardiologist in the Division of Cardiology, Okinawa Chubu Hospital. During 2003–14, he served as Chief Division of Cardiology, Okinawa Chubu Hospital. In 2008, he served as a Fellow of the American College of Cardiology. In 2016, he served as Chief of the Department of Internal Medicine.

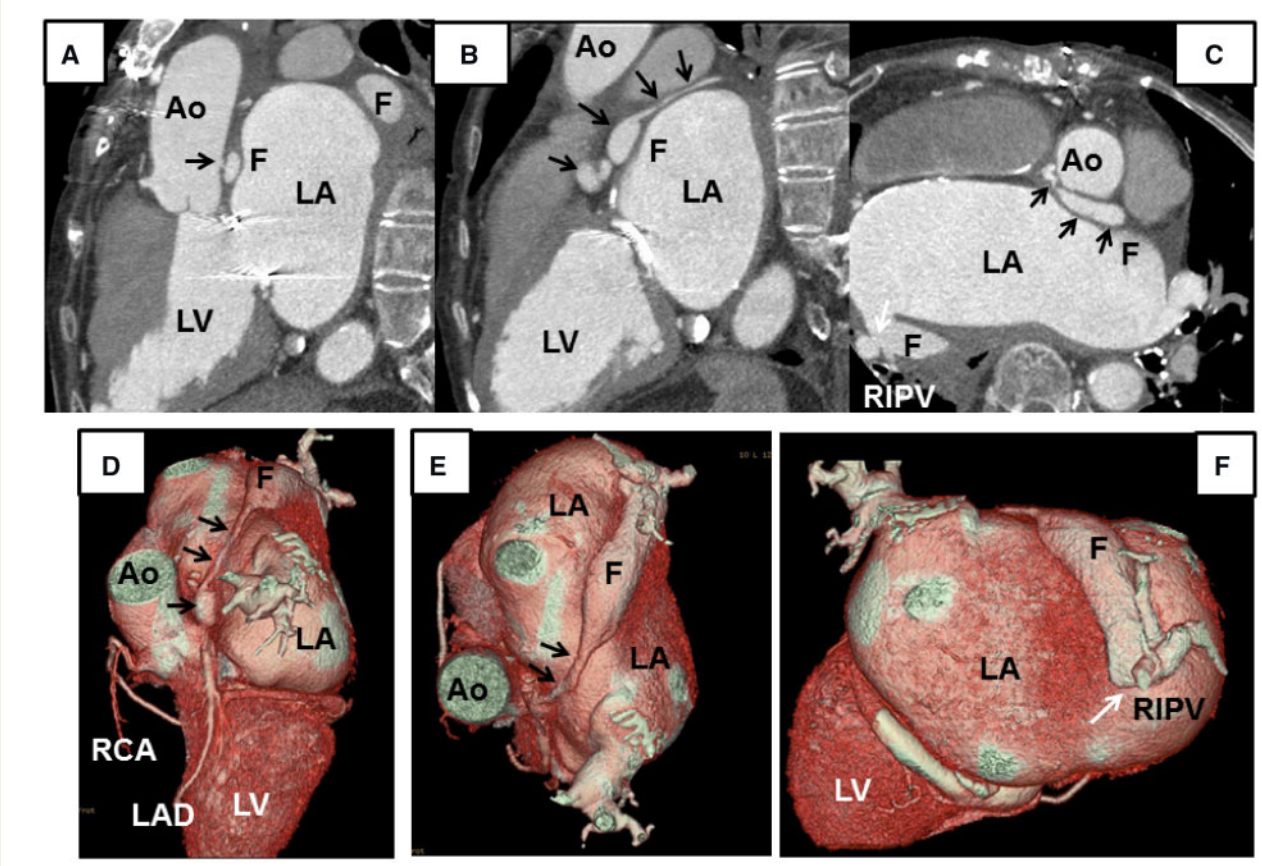

Figure 4 Computed tomography images. The fistula (F) originates from the posterior wall of the ascending aorta with a narrow neck (*A*: black arrow), just above the ostium of the left coronary artery and travels upward to form an irregular sac (black arrows in *B–D*); then, it forms a narrow tract towards the ceiling of the left atrium (black arrows in *B* and *D*). Thereafter, the diameter of the fistula tract suddenly enlarges (F in *D–F*) and fistula tract finally drains into the right inferior pulmonary vein (white arrow in *C* and *F*). *A* and *B* are sagittal views, and *C* is a transverse view. *D–F* are volume rendering images. Because the fistula tract is extremely long and complex, a single image cannot depict the whole tract; therefore, multiple images are shown to visualize the whole fistula tract. (*D*) A left-anterior oblique and cranial view, (*E*) an image from the top, and (*F*) an image from the back. Ao, aorta; F: fistula tract; LA, left atrium; LV, left ventricle; RIPV, right inferior pulmonary vein.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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