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CASE REPORT
Heart failure

Percutaneous closure of an iatrogenic aorto-right atrial fistula of the sinus of Valsalva through total arm approach: a case report

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Background Creation of an iatrogenic aorto-right atrial fistula is a rare but clinically relevant complication of cardiac surgery. Transfemoral percutaneous closure is an attractive alternative to surgical repair, but there are no reports about transcatheter repair using a complete arm access.

Case summary We present the case of a 44-year-old woman with heart failure (NewYork Heart Association Class III) due to a longstanding iatrogenic fistula from the non-coronary aortic cusp to the right atrium (RA) with aorta to RA shunting and severe tricuspid regurgitation (TR) caused by mitral valve replacement 15 years ago. The patient was successfully treated by percutaneous closure with an Amplatzer Vascular Plug II using complete brachial access. Following the procedure right heart chambers and TR decreased and symptoms resolved.

Discussion To the best of our knowledge this is the first report of percutaneous repair of an aorto-right atrial fistula using total arm accesses (radial artery and basilic vein). In appropriately selected patients, this approach is an attractive alternative to femoral access.

Keywords Percutaneous closure • Aorto-right atrial fistula of non-coronary cusp • Sinus of Valsalva • Total arm approach • Case report

Learning points

- Iatrogenic fistula of the non-coronary cusp during mitral valve replacement is a rare, but clinically relevant complication.[1]
- Percutaneous closure of an aorta to right atrium shunting of the sinus of Valsalva through total arm access is an attractive alternative approach to femoral access.

Introduction

Development of an iatrogenic aorto-right atrial fistula is a rare but clinically relevant complication of cardiac surgery.[1] Transfemoral percutaneous closure is considered a suitable alternative to surgical repair.[2–5] This is the first report to cover percutaneous closure using complete arm access.

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Timeline

Time	Events
15 years ago	Mitral valve replacement (mechanical prosthesis) in 2004
Day 1	The patient was referred by the cardiologist for evaluation of a high-pitched murmur on the left parasternal border
Day 2	A transthoracic echocardiographic examination revealed an enlarged right atrium (RA), enlarged right ventricle, severe tricuspid regurgitation (TR), and mild aortic regurgitation
Day 7	Transoesophageal echocardiography demonstrated a fistula of 6 mm in width from the non-coronary cusp (NCC) with colour jet directing towards the RA
Day 12	Aortography confirmed a torrential flow across the NCC towards the RA
Day 19	Percutaneous closure of the aorta to RA shunting of the sinus of Valsalva through complete arm access
Day 19 and 3 months after discharge	Significant improvement of right heart dimensions with residual mild TR and mild aortic regurgitation

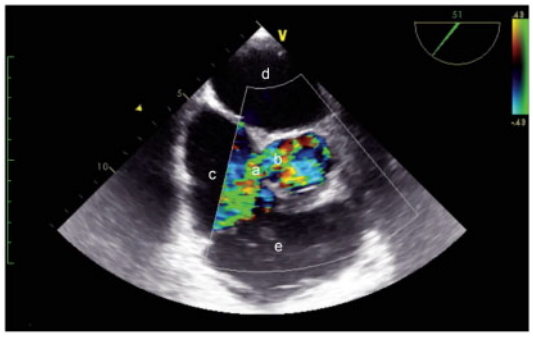

Figure 1 Transoesophageal echocardiography showing a 6 mm colour jet (a) originating from the non-coronary cusp of the aortic root (b) towards the right atrium (c). Left atrium (d). Right ventricular outflow tract (e).

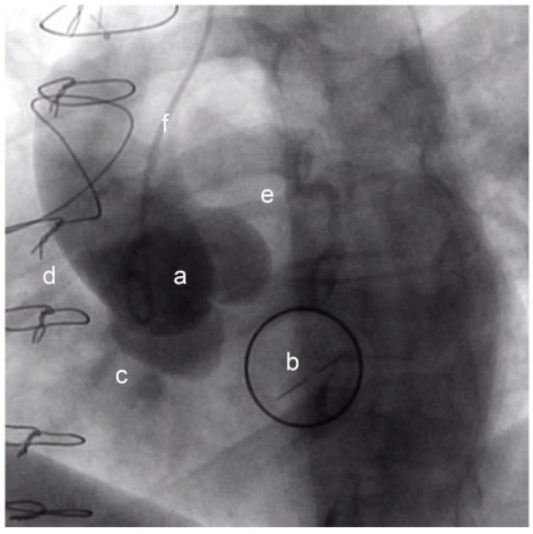

Figure 2 Aortography. Ascending aorta (a). Mitral valve prosthesis (b). Torrential flow across the non-coronary cusp (c). Right atrium (d). Left main coronary artery (e). 5 Fr diagnostic catheter (f).

Case presentation

A 44-year-old Asian woman was referred to our hospital with 15-year history of chronic heart failure. She was New York Heart Association functional classification Class III. She had undergone surgical mitral valve replacement (MVR) using a 29 mm St. Jude Medical bileaflet mechanical prosthesis at the age of 29 to address rheumatic mitral stenosis. She had been complaining of exertional dyspnoea. On physical examination, a high-pitched, grade 5/6 systolic and 3/6 diastolic murmur was detected at the left parasternal border. There were no clinical signs or symptoms of endocarditis or haemolysis. The patient was on Vitamin K anticoagulant (phenprocoumon).

Transthoracic echocardiography (TTE) revealed a normal left ventricular, end-diastolic diameter of 48 mm and systolic function (ejection fraction 70%), abnormal left atrial dimensions, volume index of 75 mL/m^2 (22–52 mL) after MVR, a dilated right ventricle with an end-diastolic diameter (RVEDD) at the base of 57 mm (≤41 mm) with normal right ventricular systolic function, a dilated right atrium (RA) with end-systolic area 32 cm^2 (≤18 cm^2), mild aortic regurgitation, and severe tricuspid regurgitation (TR) with an estimated pulmonary artery systolic pressure (PASP) of 58 mmHg (≤36 mmHg). Transoesophageal echocardiography (TOE) with colour Doppler flow revealed a continuous shunting from the non-coronary cusp (NCC) through a fistula into the enlarged RA without aneurysm formation (*Figure 1*, Supplementary material online, *Video S1*).

Further evaluation and both left and right heart catheterization were performed with our preferred access sites, the right radial artery and the right basilic vein. There was a 13% step-up in the oxygen saturation in the RA, from 65% in the superior vena cava (SVC) to 78%. Aortography revealed a torrential flow from the NCC of sinus of Valsalva towards the RA (*Figure 2*, Supplementary material online, *Video S2*). The final calculated ratio of pulmonary to systemic circulation (Qp:Qs) was 1.6:1. Due to heart failure caused predominantly by relevant left-to-right shunting and right heart enlargement the patient was advised to opt between percutaneous closure or surgical repair.

After detailed discussion with the patient and her family, the patient opted for percutaneous closure. The patient further requested the procedure be performed through the same access route as the diagnostic aortography. We decided to use a complete arm access.

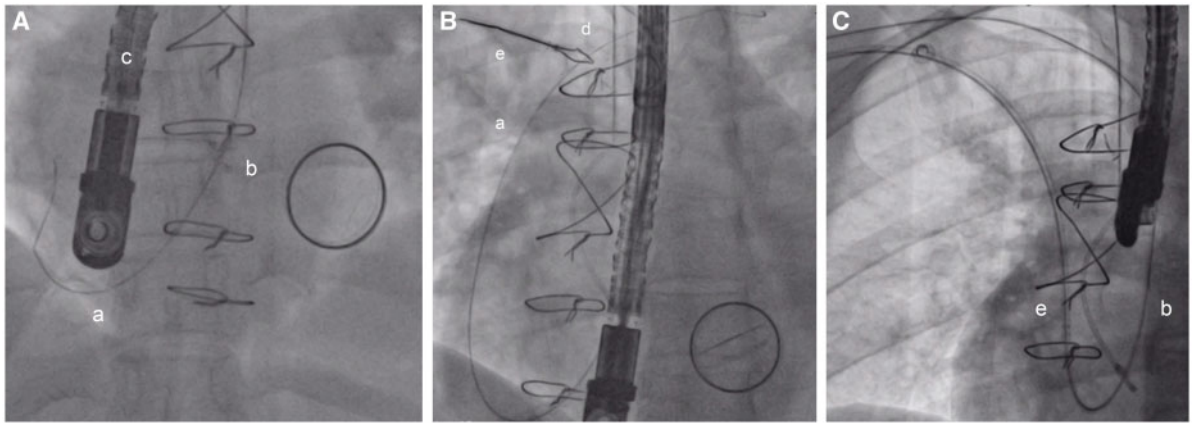

Figure 3 Probing of the fistula and creation of an arteriovenous loop. (*A*) Retrograde crossing of the aorto-atrial fistula using a glidewire (a) and a 5 Fr MP-II guiding catheter (b) under fluoroscopic and transoesophageal echocardiography (c) guidance. (*B*) Snaring (d) of the retrograde wire (a) via right basilic vein into a venous 6 Fr sheath (e). (*C*) Mother–child technique: the retrograde MP-II-guide catheter (b) is negotiated into the venous 6 Fr sheath (e).

The procedure was performed under general anaesthesia. Both radial arteries were accessed with a 5 Fr Glidesheath slender sheath (Terumo Corporation, Somerset, NJ, USA). The right basilic vein was punctured under ultrasound guidance and exchanged for a 6 Fr 90 cm angulated tip destination sheath (Terumo Corporation). A 5 Fr pigtail catheter was negotiated from the left radial artery into the ascending aorta to perform aortography. A 5 Fr MP-II-guide catheter (Medtronic, Minneapolis, MN, USA) was negotiated from the right radial artery, which was advanced under TOE and fluoroscopic guidance through the fistula towards the RA (*Figure 3A*). An angled hydrophilic 0.035″ Glidewire (Terumo Corporation) was advanced from the aorta into the SVC. The glidewire was snared with a 20 mm Gooseneck snare (eV3, Plymouth, MN, USA) (*Figure 3B*) and externalized through the right basilic vein access site making a complete arteriovenous loop. The MP-II-guide catheter was negotiated towards the inner lumen of the destination sheath, which facilitated the advancement of the sheath through the fistula into the right subclavian artery (mother–child technique, *Figure 3C*).

The glidewire and the guidecatheter were removed from the right radial artery. Guided by fluoroscopy and TOE, the left lobe of an 10 mm Amplatzer Vascular Plug (AVP) II (Abbott, Abbott Park, IL, USA) was partially deployed in the right subclavian artery. The whole assembly was retracted to the NCC level. We excluded residual shunting along the fistula using TOE and aortography and then performed a tug test. The middle lobe and the right disc were released with an excellent coaxial alignment of the device to the NCC and the RA (*Figure 4A–C*, Supplementary material online, *Video S3*). Phenprocoumon was continued. Dual antiplatelet therapy (DAPT) was given for 4 weeks in combination with a proton pump inhibitor.

The patient was discharged without any further complications. TTE at 3 months revealed significant improvement of right heart dimensions under residual mild TR (PASP 31 mmHg, RVEDD 45 mm, RA 19 cm^2) and no increase in aortic regurgitation. The patient remained asymptomatic as of a 3-month follow-up.

Discussion

After discussion among the heart team, we strongly considered an iatrogenic cause of the fistula of the NCC. Because the symptoms occurred just after MVR, the fistula was probably created iatrogenically during suturing of the mitral valve prothesis, which presumably pierced the NCC. The development of an iatrogenic defect of the NCC during MVR is a rare though clinically relevant complicaton.

Registry data and case reports consider surgical repair of the first-line treatment for ruptured sinus of Valsalva aneurysm (RSVA) with low mortality rate.

The first percutaneous closure of RSVA was reported by Cullen *et al.*[2] in 1994 using a Rashkind umbrella device. Since then, few case reports or case series covering this technique have been reported with encouraging follow-up results, and percutaneous closure of RSVA as well as aortocardiac fistula via femoral approach has been shown to be a suitable alternative to surgery in appropriately selected patients.[3–5]

Considering the longstanding nature of the disease, specifically the low risk of rupture of the NCC due to well ingrown sutures of the mitral valve prosthesis, the percutaneous approach via total arm access is a suitable therapeutic option as compared to femoral access.[1] For coronary interventions, radial access is associated with fewer complications, early ambulation, reduced costs, reduced morbidity, and improved clinical outcomes.[6] A technical advantage of deploying the device through the externalized wire via arm access provides enhanced support via the AV loop. The relative low profile of the AVP II allows for delivery by the basilic vein route. Although, in most cases, the 5–7 Fr delivery sheath is sufficient, larger defects necessitating larger devices that require an 8 Fr delivery sheath may limit device delivery by the basilic vein. However, selective venography could be helpful for guide sizing of the vein.

Device-related thrombosis is rare after a percutaneous procedure, but it can involve serious complications. To prevent thrombembolism, the preprocedural regimen of phenprocoumon was continued

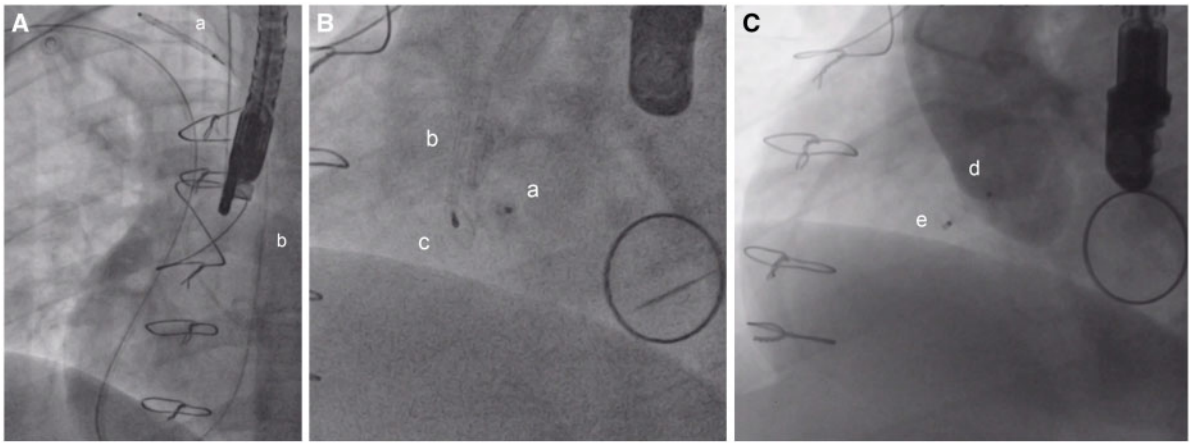

Figure 4 Deployment of the Amplatzer Vascular Plug. (*A*) Partial release of the left lobe (a) of the Amplatzer Vascular Plug from the 6 Fr sheath (b), which is advanced into the right subclavian artery. (*B*) After the sheath (b) and the Amplatzer Vascular Plug are withdrawn back to the aortic root, the left lobe (a) of the Amplatzer Vascular Plug is completely released at the entry site of the fistula in the non-coronary aortic cusp, while the right lobe (c) of the Amplatzer Vascular Plug is released at the right atrial exit site. (*C*) Final angiography demonstrating complete sealing of the fistula in the non-coronary cusp (d) by the Amplatzer Vascular Plug (e) with no increase in mild aortic regurgitation.

without interruption with a target international normalized ratio (INR) of 2–2.5. During the procedure the patient received additional unfractionated heparin. After the procedure, she was treated with DAPT with aspirin (100 mg per day), clopidogrel (75 mg per day), and coadministration of a proton pump inhibitor. DAPT was discontinued on clinical and echocardiographic follow-up after 4 weeks, and oral vitamin K antagonist therapy was continued with a target INR of 3.0. We acknowledge that optimum pharmacological treatment after shunt device implantation in patients requiring anticoagulation is not fully established.

In some patients, this approach is an attractive alternative to femoral access with high technical success and good short-term outcome; however, long-term follow-up is absolutely necessary.

Lead author biography

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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 author/s 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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