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Percutaneous Closure of an Aorta to Left Atrium Fistula with an Amplatzer Duct Occluder

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We describe percutaneous closure of an unusual paravalvular leak, with a communication from the noncoronary cusp of the aortic valve to the left atrium adjacent to a St. Jude mitral valve prosthesis, in the absence of an infection. The patient presented with worsening dyspnea and edema. The anatomic location of the valve annulus adjacent to the noncoronary cusp underlies the etiology of this communication. Outcomes from surgical repair are associated with high mortality. We performed percutaneous closure of the defect, using general anesthesia and transesophageal echo guidance. An Amplatzer duct occluder was placed in the fistula, with immediate and complete closure of the shunt. While no devices specific for closure of paravalvular leaks have been designed, growing experience with a variety of devices and the use of echo guidance have allowed successful therapy in many complex cases.

Key words: paravalvular leak; intracardiac shunt; prosthetic valve

INTRODUCTION

Clinically significant paravalvular periprosthetic leaks are relatively uncommon, occurring in <5% of patients with prosthetic valves [1–4]. They are more common with mechanical valve implants, and small leaks may be found in over 40% of the patients, if searched for carefully [5,6]. We describe percutaneous closure of a particularly unusual lesion associated with a paravalvular leak in which there was a fistulous communication from the noncoronary cusp of the aortic valve to the left atrium, adjacent to a St. Jude mitral valve prosthesis [7]. A fistula connection from the aorta to the left atrium has usually been associated with the sequelae of infective endocarditis. The appearance of this lesion in association with a mechanical valve in the absence of an infection is highly unusual. The anatomic location of the valve annulus adjacent to the noncoronary cusp underlies the etiology of this communication. Heart failure is a significant complication of paraprosthetic leaks, and outcomes from surgical repair are associated with high mortality [7,8].

CASE DESCRIPTION

A 76-year-old woman presented with a history of St. Jude mitral valve replacement in 1994. In 2001, she had a transesophageal echocardiogram that showed normal prosthetic function and no aortic insufficiency. She had been well until about 1 year prior to her current presentation, when she began developing progressive exertional dyspnea and lower extremity edema. Ultimately, she was dyspneic, walking only 15–20 feet despite therapy with captopril, furosemide, aldactone, metolazone, and digoxin. She had not required hospitalizations for congestive heart failure. She had no history of endocarditis. She had murmurs that were thought to be due to pulmonic insufficiency or tricuspid regurgitation. During evaluation for the etiology of her shortness of breath, she underwent transthoracic and transesophageal echocardiography, and cardiac catheterization. Echocardiography demonstrated left ventricular hypertrophy with an ejection fraction of 70%. There was right ventricular hypokinesis with pulmonic and tricuspid insufficiency. The estimated pulmonary artery systolic pressure was 70 mm Hg. Transesophageal echo demonstrated the mitral prosthetic leaflets to function normally. Spontaneous echo contrast was seen in the left atrium. Doppler interrogation demonstrated an abnormal, eccentric jet in the perivalvular area.

Cardiac catheterization included aortography. This demonstrated a fistula from the noncoronary cusp of the aorta into the left atrium. The left atrium was

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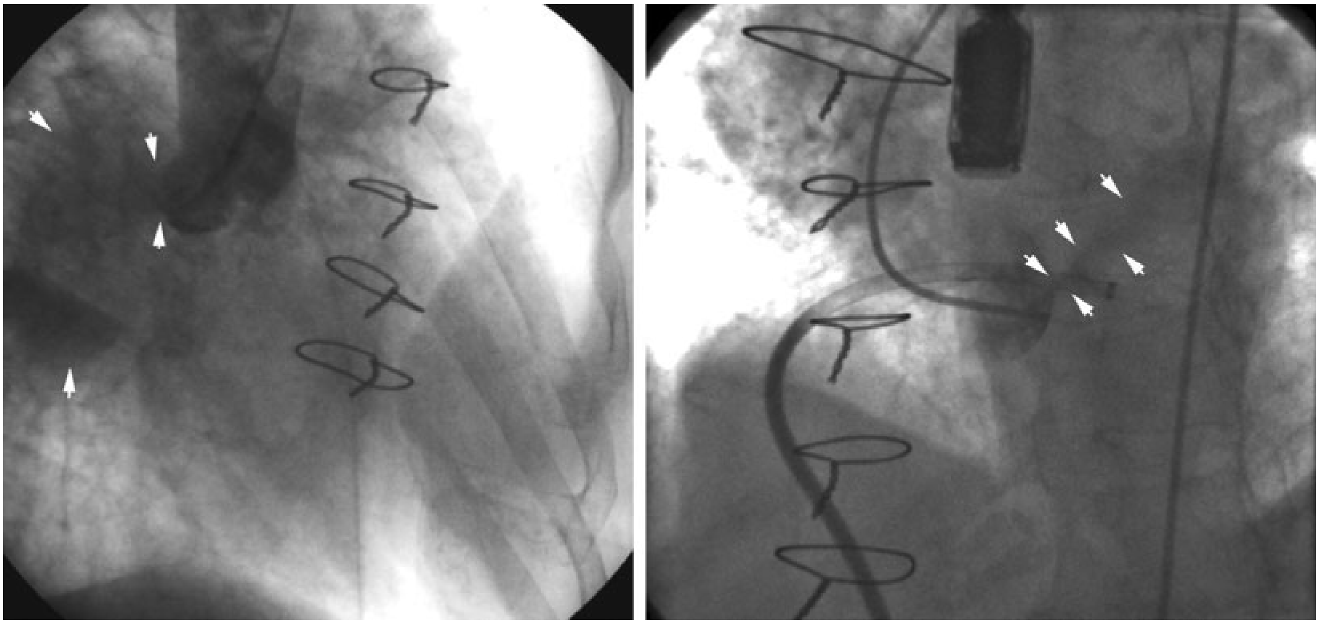

Fig. 1. In the left panel, the arrows outline contrast filling the left atrium, arising from the posterior or noncoronary aortic cusp during aortography. The right panel shows a multipurpose catheter in the noncoronary cusp, with a discrete jet of contrast streaming into the left atrium, also noted by arrows. A transseptal sheath is seen in the left atrium, and a transesophageal echo probe is seen at the top of the right panel.

noted to largely fill during a single cardiac cycle and contrast entered the pulmonary veins. The pulmonary artery pressure at that time was noted to be 84/20 mm Hg, with a mean of 45 mm Hg and a pulmonary capillary wedge mean pressure of 25 mm Hg. Angiography showed mild coronary artery disease, and ventriculography demonstrated an ejection fraction of 55% with no angiographic evidence for mitral regurgitation.

Following review of the diagnostic images, percutaneous closure of the fistula was planned. After general anesthesia, and using transesophageal echo guidance, an 8F sheath was placed in the right femoral vein, and a 6F sheath was placed in the right femoral artery. Transseptal puncture was accomplished without incident, using an 8F Mullins transseptal sheath. Anticoagulation with heparin was administered. Aortography once again demonstrated the fistulous connection between the noncoronary cusp of the aorta and the base of the left atrium (Fig. 1). Transesophagel echo clearly demonstrated the lesion adjacent to the mitral annulus and just above the aortic leaflets (Fig. 2). Continuous flow was noted through the fistula. The systolic Doppler-estimated gradient from the aorta to the left atrium was 90 mm Hg, and the diastolic gradient was 50 mm Hg. A second, small paravalvular leak was noted adjacent to the mitral prosthetic annulus, with a narrow-based, systolic jet from the left ventricle to the left atrium. The pulmonary artery peak systolic pressure was 42 mm Hg, reflecting recent intensive therapy for the patient's lower extremity edema and dyspnea.

Via the arterial access, a 5F multipurpose catheter was placed in the noncoronary cusp, and contrast injection demonstrated close proximity to the origin of the fistula (Fig. 1). A hydrophilic 0.035 in. angle-tip wire was used to probe the sinus of Valsalva, until it crossed the fistula into the left atrium. The 5F multipurpose catheter was easily passed across the defect without any feeling of resistance into the left atrium (Fig. 3).

A 0.035 in. $\times$ 300 cm long wire was passed through the 8F transseptal sheath into the left atrium, and snared with a 10-mm gooseneck snare placed directly through the 5F multipurpose catheter (Fig. 3). The wire was pulled across the fistula from the left atrium into the aorta and exteriorized through the arterial sheath, using a 7F arterial sheath (Fig. 4). With the wire exiting both the venous and arterial sheaths, the transseptal sheath was exchanged for a 7F AGA medical ASO delivery sheath. The dilator of the sheath tracked through the left atrium, across the defect, and into the aorta with only minimal resistance. The distal portion of the sheath was placed in the top of the aortic arch, and the dilator and the wire were removed. This left the sheath in the aorta, from the venous side, for delivery of a duct occluder (Fig. 5). Based on the feeling of some resistance around the sheath and crossing the defect, and a diminution of color jet flow with

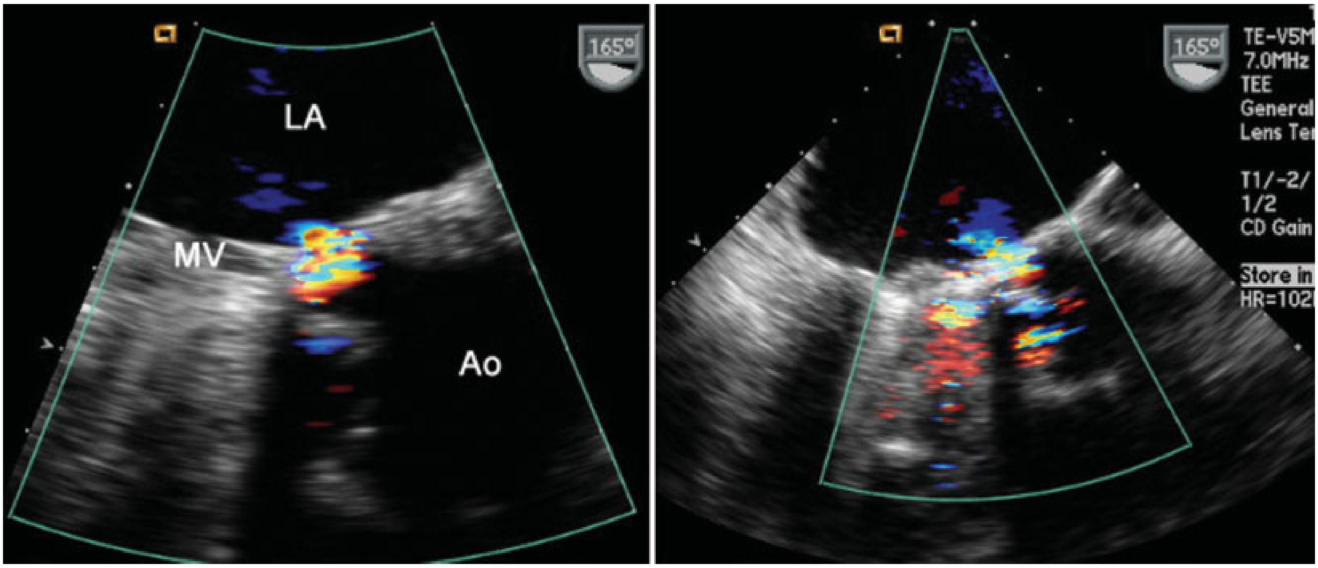

Fig. 2. Transesophageal echo images obtained during the procedure. The color flow can clearly be seen between the aorta and left atrium. In the left panel, there is acoustic shadowing from the St. Jude prosthetic mitral valve. AO, aorta; LA, left atrium; MV, mitral valve. [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

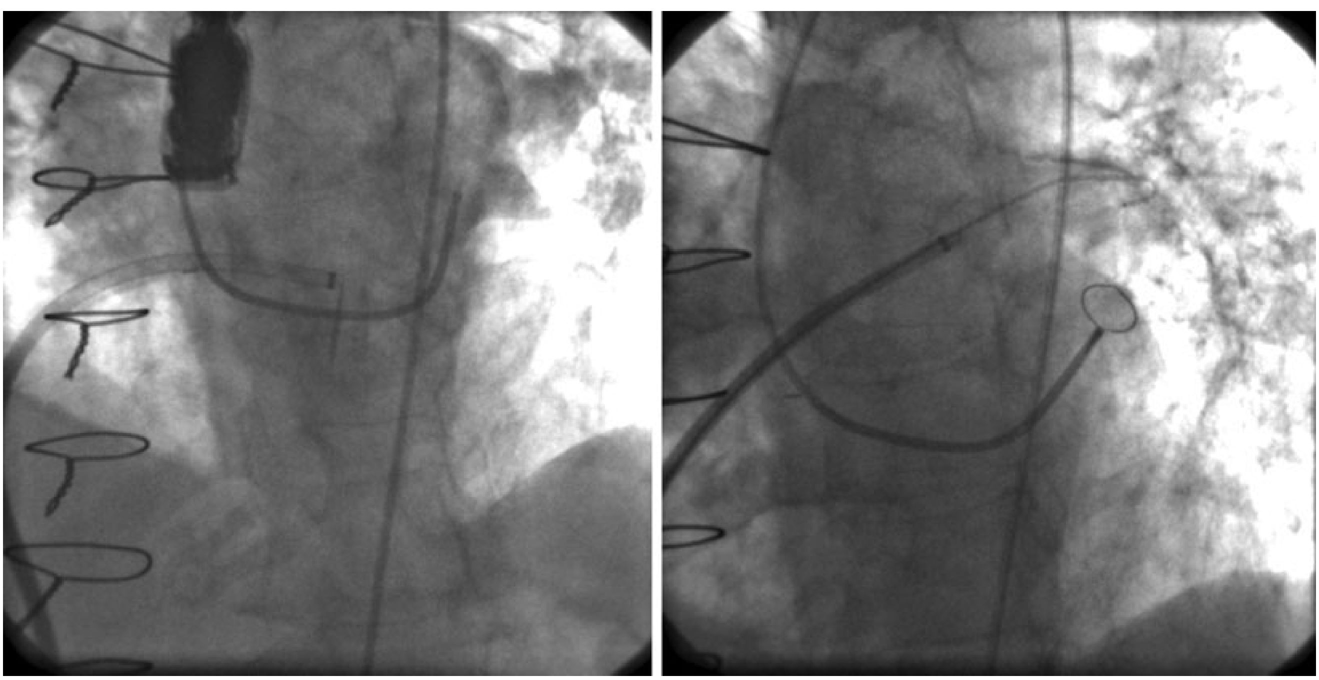

Fig. 3. In the left panel, a multipurpose catheter is seen traversing the aorta and entering the left atrium above the aortic valve. In the right panel, a snare has been passed through the multipurpose catheter into the left atrium. A guide wire has been passed through the Mullins sheath into the left atrium as well.

the 7F sheath (2.3 mm diameter) across the defect, a 6-4 mm AGA duct occluder was chosen. The sheath appeared to have kinked in the fistula tract after removal of the guidewire, but advancement of the duct occluder on its delivery cable required no force at the point of the apparent kink (Fig. 5). The distal retention flange of the duct occluder was opened in the aorta, and the sheath and the device were withdrawn until the flange abutted the sinus of Valsalva on the aortic side (Fig. 6). At this point, TEE imaging showed that aortic leaflet motion was unimpeded, and there was no evidence for aortic insufficiency on color Doppler

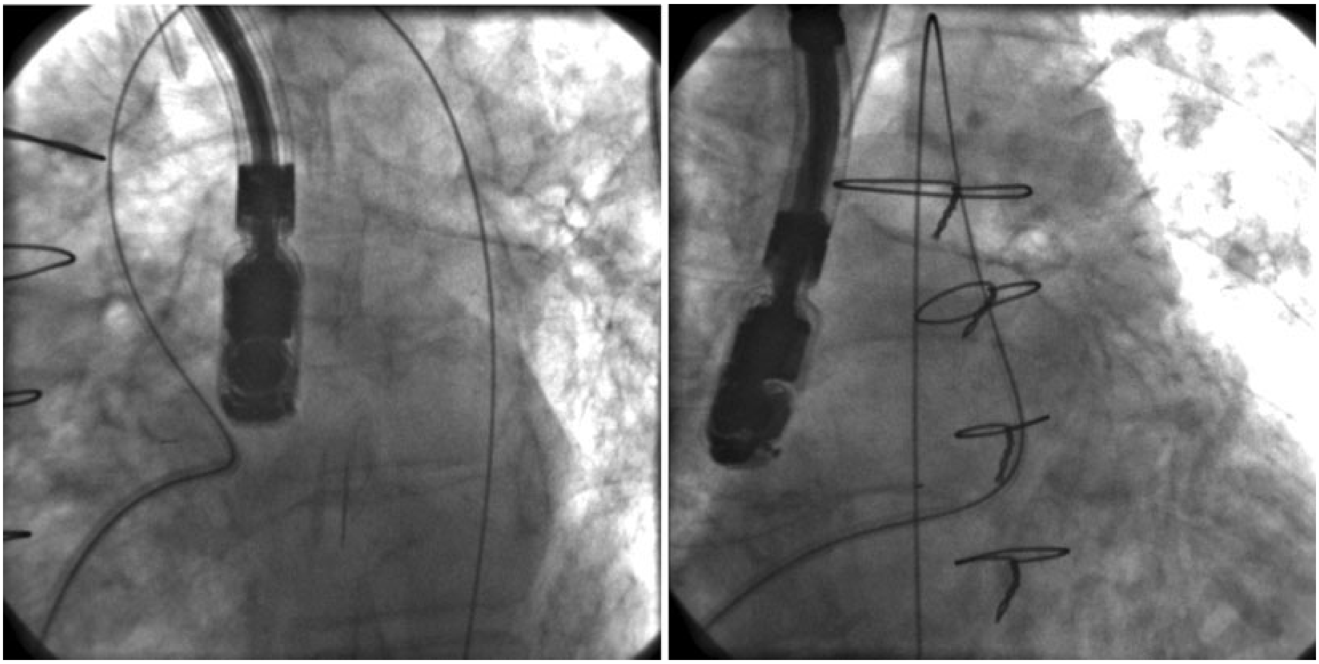

Fig. 4. Left panel shows an anteroposterior view of a guide wire looped through the circulation. From the lower left, the wire traverses the Mullins sheath, crosses from the left atrium into the aortic root just above the aortic valve, and goes through the aortic arch and then into the descending aorta. In the right panel, a right anterior oblique view shows the course of the wire from posterior in the left atrium, coursing anteriorly into the aorta.

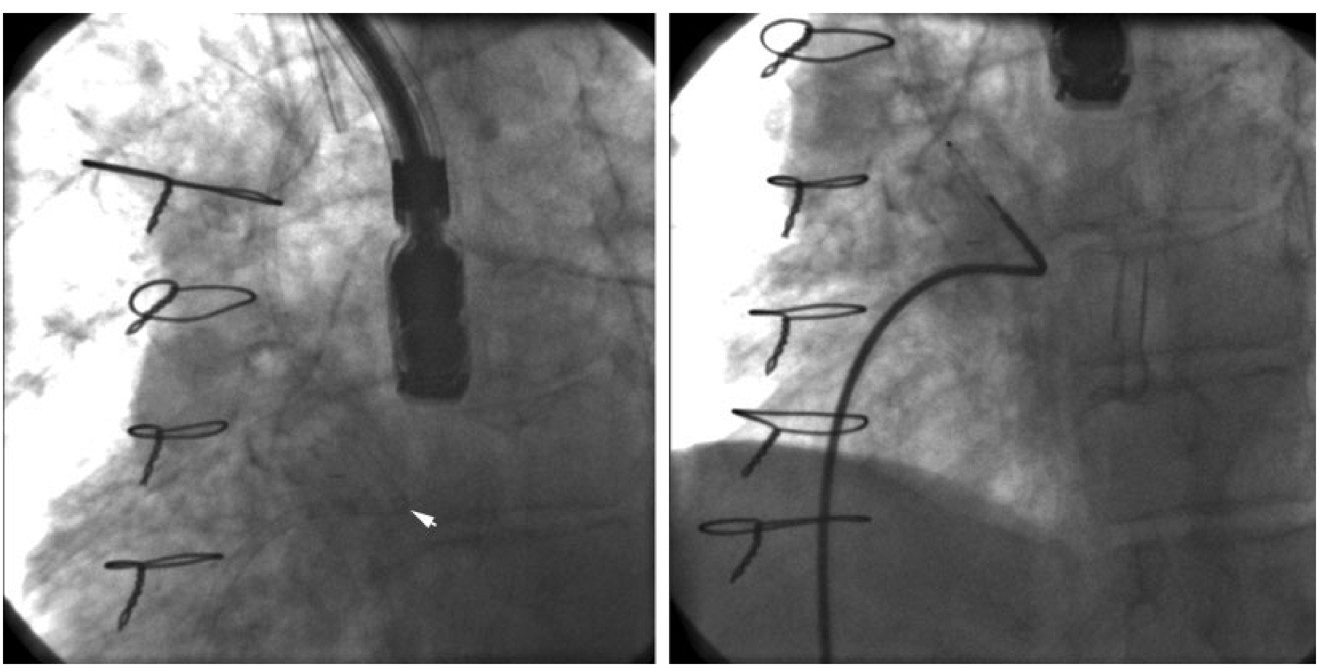

Fig. 5. In the left panel, a 7F delivery sheath has been passed through the defect from the venous side, and extends into the ascending aorta. The arrow shows the point at which the catheter is kinked in the fistula. In the right panel, the Amplatzer duct occluder has been passed through the delivery sheath into the aorta. The delivery cable is opaque, and the device can be seen to be compressed within the sheath. The tip of the device is marked by a small radiopaque marker.

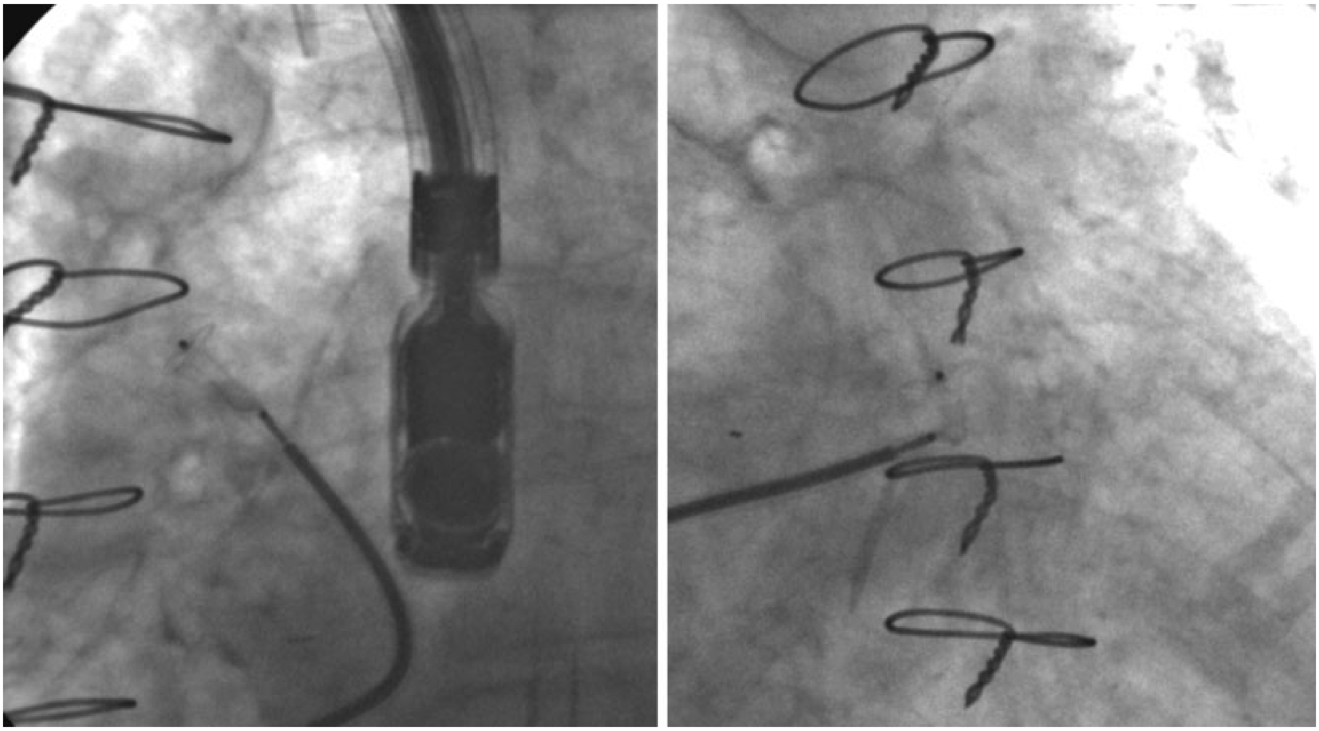

Fig. 6. In the left panel, the device can be seen with the retention phlange pushed out of the delivery sheath and opened in the aorta. In the right panel, the device has been pulled back so that the delivery cable is now fully within the left atrium and the device is partially deployed within the defect.

examination. The duct occluder was pulled back to seat well into the aortic side of the fistula opening, and the remainder of the occluder exposed as the delivery sheath was pulled back into the left atrium (Fig. 7). Echocardiographic, Doppler, and angiographic assessments demonstrated complete occlusion of the fistula without aortic insufficiency or impingement on the mitral leaflets. The small left ventricle to left atrium paravalvular leak was unchanged compared to preprocedure.

The device was unscrewed from the delivery cable and released. A final aortogram demonstrated complete closure of the shunt without aortic insufficiency (Fig. 8). This was confirmed on Doppler echocardiographic examination (Fig. 9).

Less then 48 hr after the procedure, she was able to walk the length of the hospital corridor (200 feet) without dyspnea. By 2 weeks post procedure, her edema was resolved, and her diastolic murmur (initially thought to be from pulmonic or tricuspid insufficiency) was no longer detectable.

DISCUSSION

A communication between the aorta and the left atrium is unusual. Most reported cases are related to infective endocarditis. This patient clearly had no history of endocarditis or clinical signs or findings to suggest an infective etiology. The communication in this patient is paravalvular. The proximity of the posterior sinus of Valsalva to the mitral annulus is easily appreciated in anatomic drawings (Fig. 10). The long delay between mitral valve replacement and the appearance of clinical symptoms related to this fistula cannot be clearly explained. It is possible that a very small leak existed in the early postoperative time frame, and enlarged very slowly. It is also possible that erosion of suture occurred much later in the course after valve replacement. It is also possible that repair of the intra-atrial septum at the time of surgery might have involved the posterior sinus.

A wide variety of devices have been used to close paravalvular leaks. Various atrial septal, ventricular septal occluders, and coils have been reported [9–17]. None of these devices are specifically designed or ideally suited for closure of paravalvular leaks [15,18]. More recently, the AGA medical Amplatzer PDA occluder has been reported as useful in this clinical setting [17]. The PDA occluder is especially well suited, because it has a retention flange to help maintain its position in a narrow-necked, relatively short defect such as these paravalvular leaks.

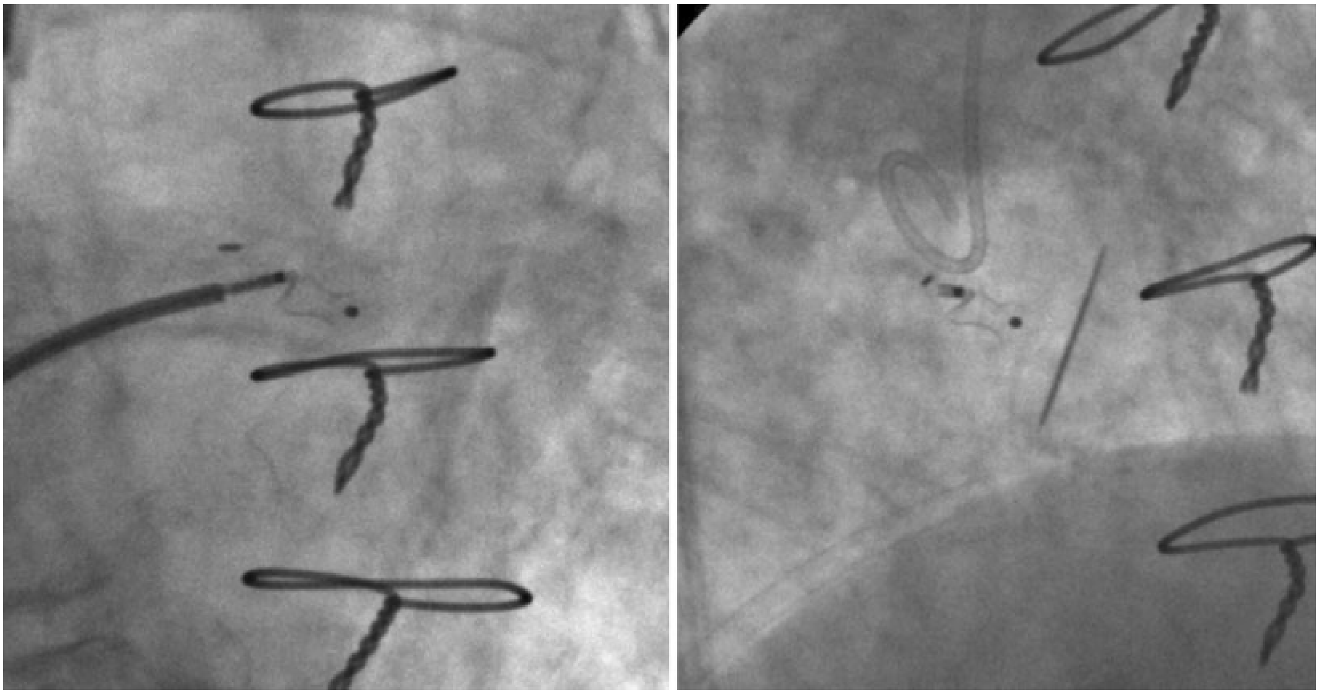

Fig. 7. The left panel shows the device well implanted into the fistula, with the delivery cable attached in the left atrium. The right panel shows the device after release.

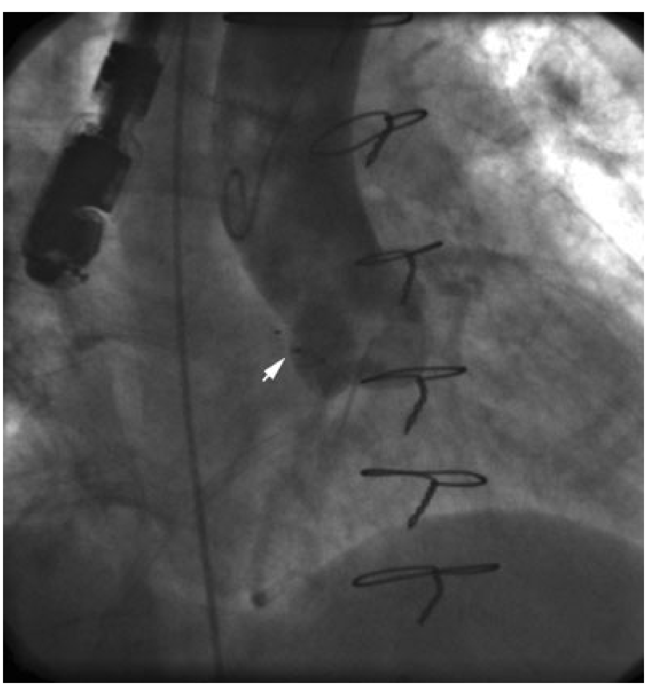

Fig. 8. Aortography after device placement shows no further evidence for contrast leaking into the left atrium. The arrow shows the location of the device in the noncoronary cusp.

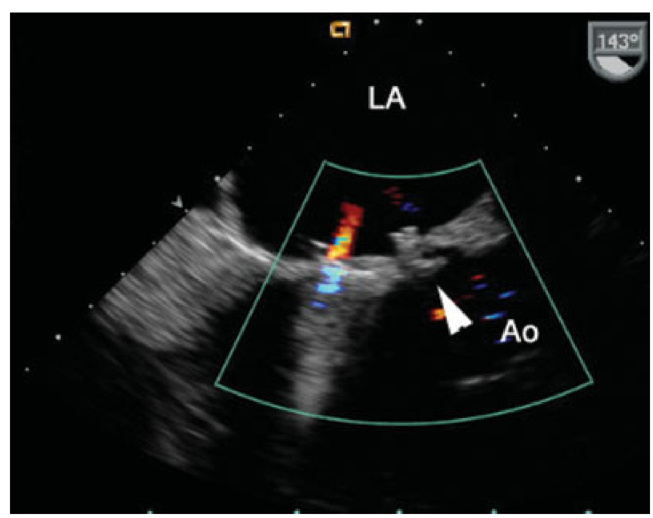

Fig. 9. Transesophageal echo imaging after device placement. There is no longer flow between the aorta and left atrium. The arrow points at the device in the wall of the aorta, adjacent to the left atrium. AO, aorta; LA, left atrium. [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

A number of technical challenges are presented by the use of vascular plug devices such as this for paravalvular leak closure. In this case, crossing the lesion was not difficult. After a multipurpose catheter was positioned in the noncoronary cusp, a glide wire was easily passed into the left atrium. Snaring the wire in the left atrium required considerable maneuvering because of the large size of the left atrium. We were best able to get the wire and snare into close proximity by maneuvering into a common pulmonary vein in which there was less room for the two devices.

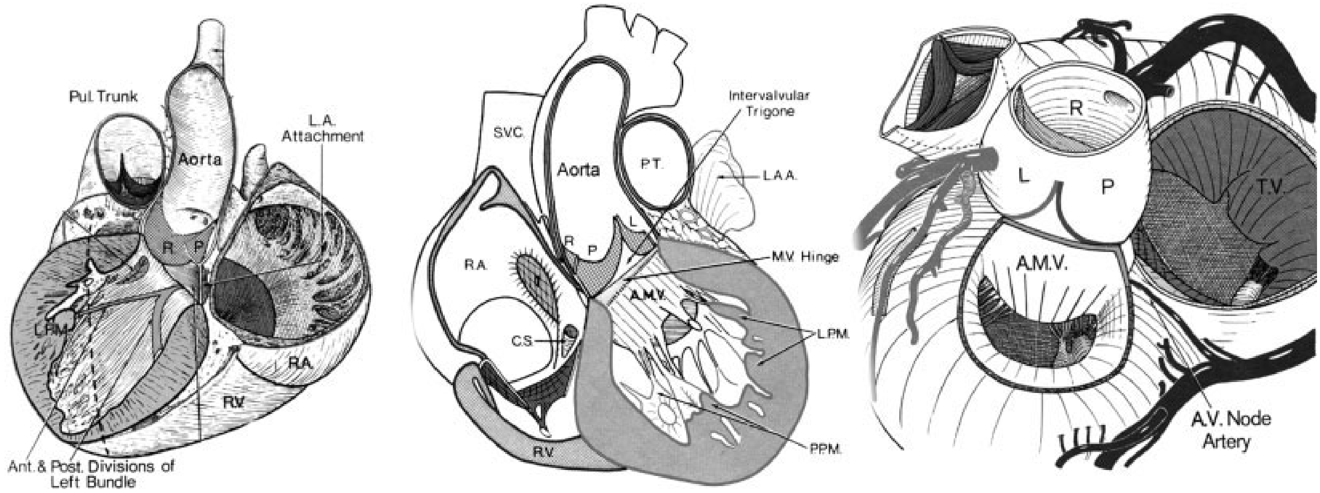

Fig. 10. Anatomic drawings showing the relationship between the posterior or noncoronary cusp of the aortic root and the left atrium. It is clear that the mitral annulus abuts the noncoronary cusp. Reproduced with permission from McAlpin WA. Heart and coronary arteries: an anatomical atlas for clinical diagnosis, radiological investigation, and surgical treatment. New York: Springer-Verlag; 1975. p 19, 161.

The duct occluder delivery sheath passed across the defect with the support of a wire that had been exteriorized on the arterial side and snared. It is not clear whether it would have been possible to advance the sheath through the defect without that degree of support. The sheath kinked in the defect, after the wire and the dilator were removed; but it was surprisingly easy to pass the occluder across the kink with minimal retraction of the sheath.

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