Aorto-Right Ventricular Fistula Post-Transcatheter Aortic Valve Replacement: Multimodality Imaging of Successful Percutaneous Closure

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INTRODUCTION

Transcatheter aortic valve replacement (TAVR) has transformed aortic stenosis therapy, with greater than 200,000 procedures performed worldwide since the first case was performed by the French interventional cardiologist Alain Cribier in 2002.[1] One rare but dreaded complication is aortic periannular rupture, seen in less than 1% of all TAVR procedures.[2] Although TAVR is a safe procedure, with an average national in-hospital mortality of less than 4%, complications may still occur.[3]

Aortic periannular rupture post-TAVR refers to a spectrum of injuries that occur in the aortic root and left ventricular outflow tract (LVOT) region and portend a poor prognosis. It has been described as having four types: supra-annular, intra-annular, subannular, and combined.[4] These ruptures often lead to flow of blood into the pericardial space, which may lead to pericardial tamponade.

We describe a case of aorto-right ventricular (RV) fistula post-TAVR that is unique due to its flow pathway, Doppler findings, and lack of rupture into the pericardial space. Using a multimodality imaging approach for diagnosis and preprocedure planning, the rupture defect was successfully eliminated by percutaneous implantation of one Amplazter vascular plug (AVP) intravenous (IV) device (St. Jude Medical, St. Paul, MN). To our knowledge, this is the first reported case of the successful use of a plugging device for this type of disease.

CASE PRESENTATION

A 91-year-old man with severe symptomatic high-gradient aortic stenosis (American College of Cardiology/American Heart Association Stage D1) underwent successful percutaneous transfemoral implantation of a 29-mm CoreValve Evolut R (Medtronic, Minneapolis, MN) followed by postdilatation using single inflation of a 25-mm Z-Med balloon (B. Braun Medical, Bethlehem, PA).

Four months later, he developed progressive congestive heart failure with preserved left ventricular ejection fraction and no evidence for hemolytic anemia. Two-dimensional (2D) transthoracic echocardiography (TTE; Figure 1 and Video 1) and transesophageal echocardiography (TEE; Figure 2 and Video 2) revealed abnormal continuous color and spectral Doppler flow between the aortic root and the right ventricular outflow tract (RVOT) indicative of aorto-RV fistula. Upon review of the original procedural TTE, similar findings were also present immediately after postdilatation of the TAVR valve.

Electrocardiogram-gated computed tomographic angiography further characterized the fistulous tract originating at the right sinus of Valsalva (SOV) just above the aortic annulus, passing along the anterior portion of the CoreValve, and entering into the RV at the level of the RVOT (Figure 3).

He subsequently underwent successful percutaneous closure of the aorto-RV fistula using an 8-mm AVP IV device (St. Jude Medical) under fluoroscopic (Figure 4 and Video 3) and three-dimensional (3D) TEE guidance (Figure 5, Figure 6, and Video 4).

DISCUSSION

Although rare, aortic periannular rupture TAVR can be life threatening.[5] The diagnosis of periannular rupture is typically made by 2D and Doppler echocardiography at the time of implantation, which also can detect associated complications such as pericardial effusion with tamponade.

The current classification scheme of periannular rupture is based on its location relative to the aortic annulus: above (supra-annular), below (subannular), within (annular), or a combination (combined).[4] However, determining the exact location of rupture can be difficult and often requires advanced imaging such as cardiac computed tomography, magnetic resonance imaging, and 3D echocardiography to provide maximal spatial resolution for defect visualization.

Postrupture, fistulous flow may occur along the following pathways:

1. From the aorta at the level of the sinuses to the lower pressure right atrium (RA) or RV, causing systolic and diastolic (continuous) flow tracings on spectral Doppler.[6]
2. From the LVOT to the RVOT, causing systolic-only flow on spectral Doppler similar to ventricular septal defects.[7]

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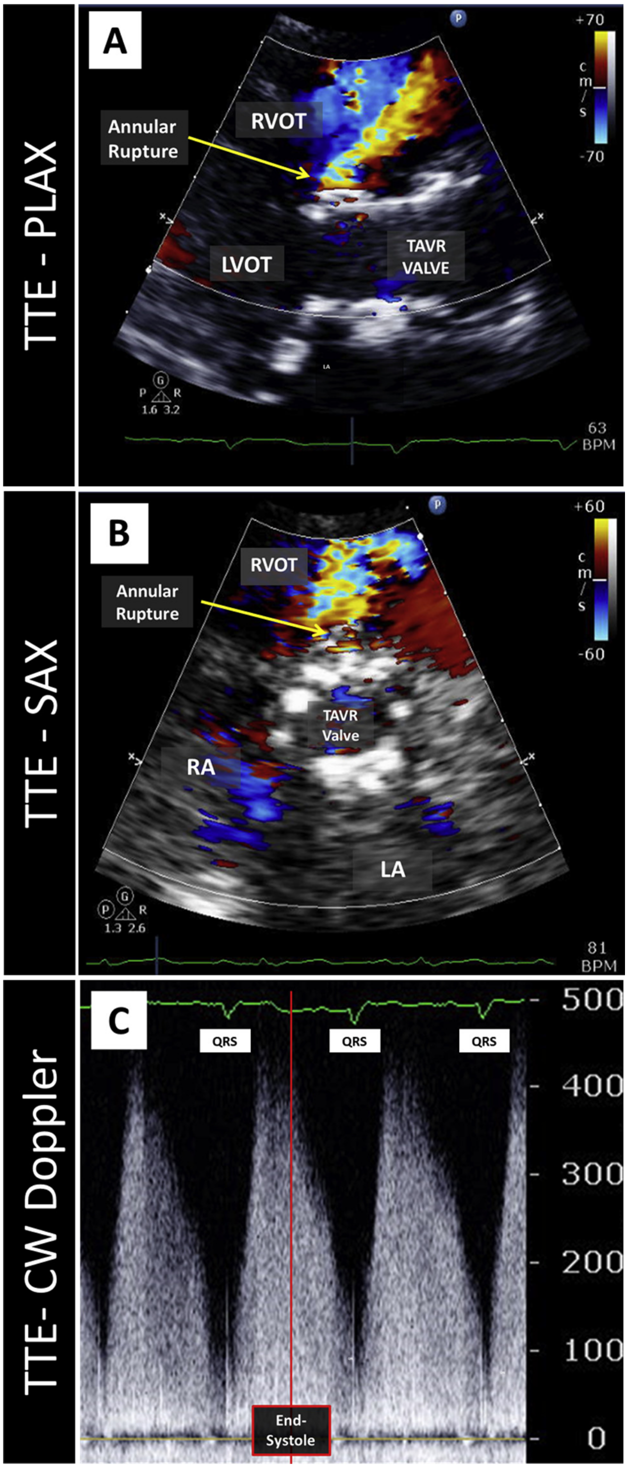

Figure 1 Aorto-RV fistula: TTE. **(A, B)** Post-TAVR TTE with color flow Doppler in the parasternal view demonstrates aorto-RV fistula with fistulous flow (*arrow*) from the aortic annular area to the RVOT in the long axis **(A)** and short axis **(B)**. **(C)** Continuous wave spectral Doppler demonstrates unusual, mostly high-velocity systolic and diastolic flow between the aortic annulus and RVOT. This flow pattern argues against a ventricular septal defect where the high velocity flow would have been in systole only. *CW*, Continuous wave; *LA*, left atrium; *PLAX*, parasternal long axis; *SAX*, short axis.

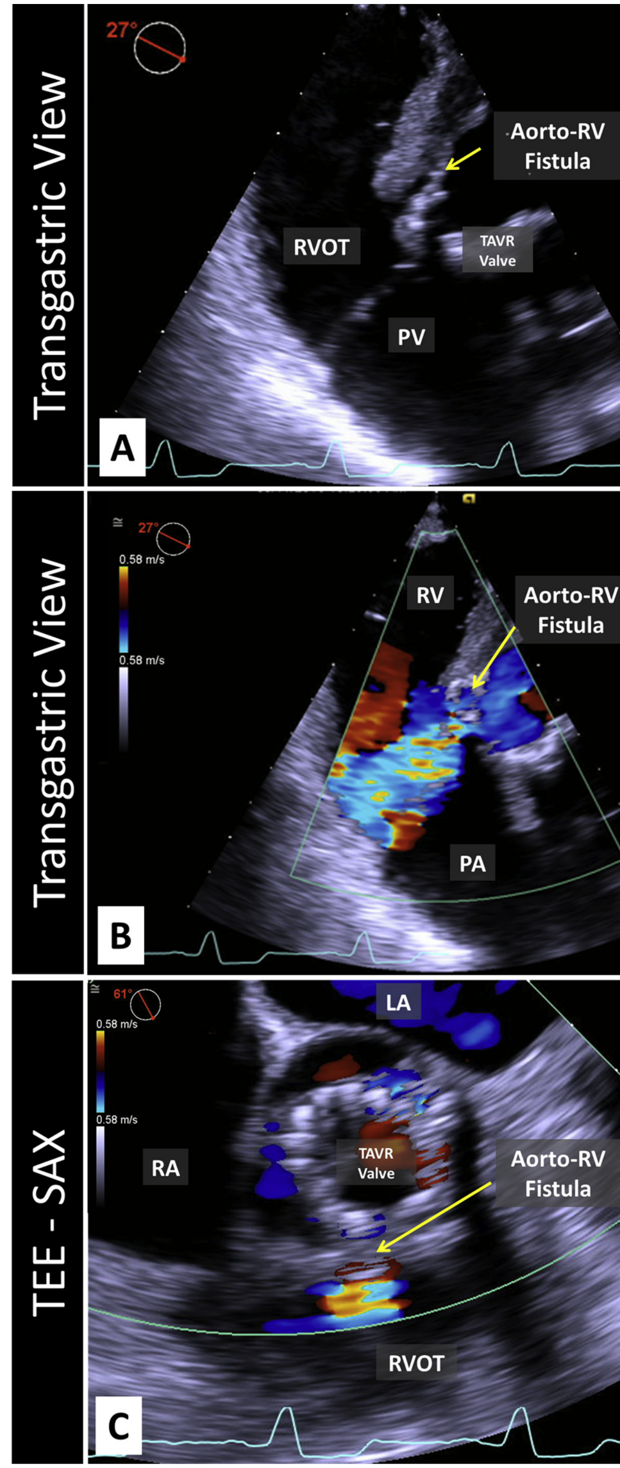

Figure 2 Aorto-RV fistula: TEE. **(A, B)** Post-TAVR TEE in the deep transgastric view demonstrates aorto-RV fistula represented by an echolucent channel (*arrow*) between the TAVR prosthesis and the RVOT on B mode **(A)** and color Doppler imaging **(B)**. **(C)** Mid-esophageal short-axis view with color Doppler demonstrates fistulous flow from the aortic root into the RVOT. Note that on TEE, short-axis visualization of the defect may be more difficult than on TTE since the anteriorly located annular rupture is partly shadowed by the posteriorly located TAVR valve on TEE. This added to the challenge of TEE guidance of percutaneous closure. *LA*, Left atrium; *PA*, pulmonary artery; *PV*, pulmonic valve; *SAX*, short axis.

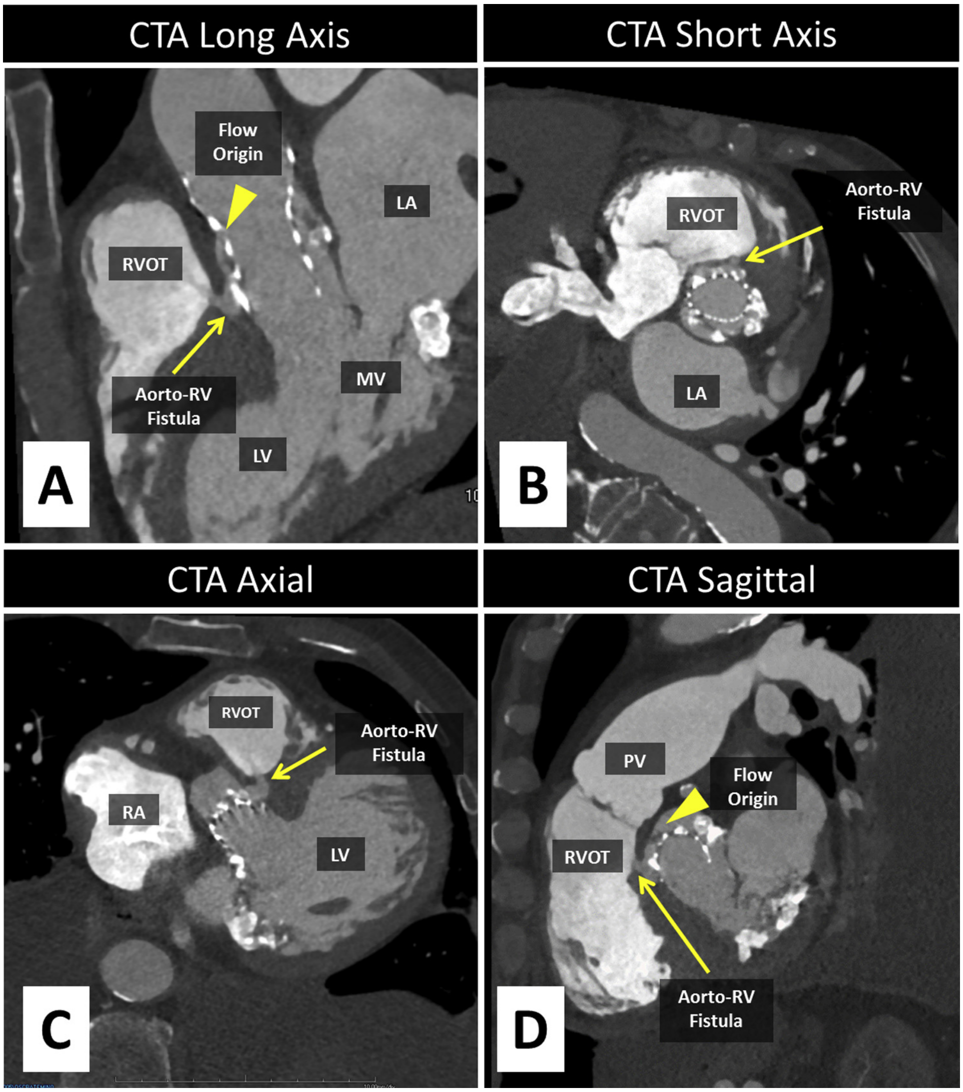

Figure 3 Aorto-RV fistula: computed tomographic angiography (CTA). **(A-D)** Electrocardiogram-gated CTA with multiplane image reconstruction demonstrates the aorto-RV fistula. The flow originates from the right SOV (*arrowhead*), passes along the anterior portion of the TAVR stent, and ultimately enters the RVOT through the aorto-RV fistula defect (*arrow*). *LA*, Left atrium; *LV*, left ventricle; *MPA*, main pulmonary artery; *MV*, mitral valve; *PV*, pulmonic valve.

3. From the aorta at the level of the sinuses to the pericardium.
4. Some combination of the above three types.

In this case, fistulous flow from the aorta at the SOV to the RVOT is demonstrated by the presence of systolic and diastolic spectral Doppler signals. Although conceptually this defect is similar to aneurysm-associated SOV rupture, the spectral Doppler tracings appear unique. In SOV rupture, spectral velocities typically do not drop to baseline, while in our case there was distinct cessation of velocities in late diastole (Figure 1C). A perimembranous ventricular septal defect was also in the differential diagnosis. However, with a ventricular septal defect, the high flow velocities would have been seen in systole only.

High-resolution computed tomography with multiplanar reconstruction points to an entry point defect into the RV at the level of the aortic annulus. Two possibilities may explain this—either the defect is slightly above the aortic annulus or the defect is at or below the aortic annulus but the TAVR skirt creates a seal between the LVOT and the aorta, permitting flow only between the SOV and RVOT.

This case not only demonstrates a unique pathophysiologic mechanism but also a unique treatment approach. Although percutaneous valve-in-valve implantation has reportedly been used to seal post-TAVR annular rupture,[8] ours appears to be the first reported case of a successful elimination of a post-TAVR aorto-RV fistula using a vascular plug. Risk factors for post-TAVR aorto-RV fistula in our patient included heavy calcifications of the native aortic valve and LVOT as well as postdilatation of a self-expanding TAVR valve.

CONCLUSION

As the use of TAVR as a treatment for severe aortic stenosis grows, the likelihood of encountering rare but life-threating complications such as aortic annular rupture becomes greater. As such,

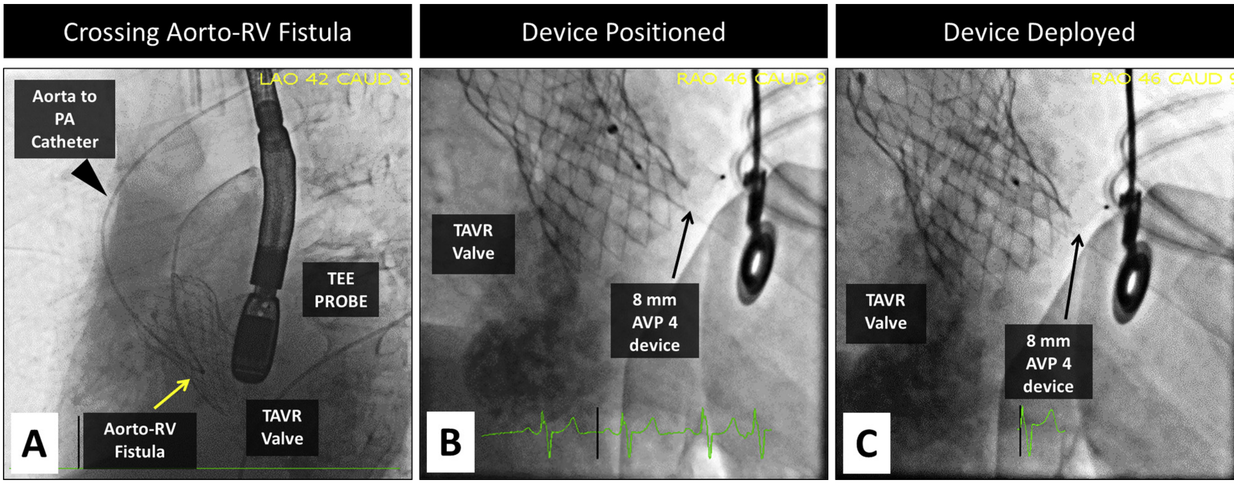

Figure 4 Cine angiography during percutaneous closure of aorto-RV fistula. **(A)** The aorto-RV fistula was initially crossed with a multipurpose catheter and an angled wire. Subsequently, a 4-French shuttle sheath was placed through the fistula; this was confirmed with cine angiography and 3D TEE. **(B)** Subsequently, an 8-mm AVP 4 device was placed across the fistula. **(C)** Once the proper position was verified, the cable was detached and the device was deployed.

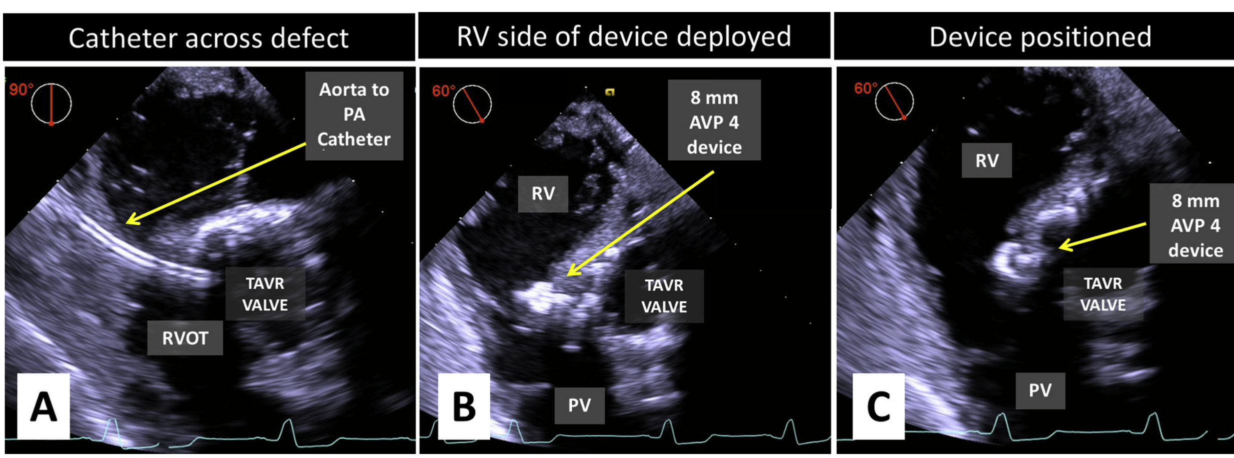

Figure 5 Two-dimensional TEE of aorto-RV fistula closure. **(A)** Initially, 2D TEE in a deep gastric view confirmed crossing of a 4-French shuttle sheath through the aorto-RV fistula defect. **(B)** Subsequently, the RV side of AVP 4 device was deployed. **(C)** The AVP 4 was then pulled back, seating the device in the defect. *PV*, Pulmonic valve.

multimodality imaging is critical in diagnosing aortic root injury and its complications, as well as in preprocedure planning for percutaneous therapies.

In summary, this case demonstrates the use of multimodality imaging in the diagnosis of an aorta-RV fistula post-TAVR, a unique disease on the aortic annular rupture spectrum that allowed for an innovative and effective therapeutic approach. The fistula likely developed immediately following postdilatation of the TAVR valve, and its clinical significance was fully recognized 4 months later when the patient presented with congestive heart failure. Risk factors for post-TAVR annular rupture in our patient included heavy calcifications of the native aortic valve and LVOT as well as postdilatation of a self-expanding TAVR valve.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.case.2017.02.002.

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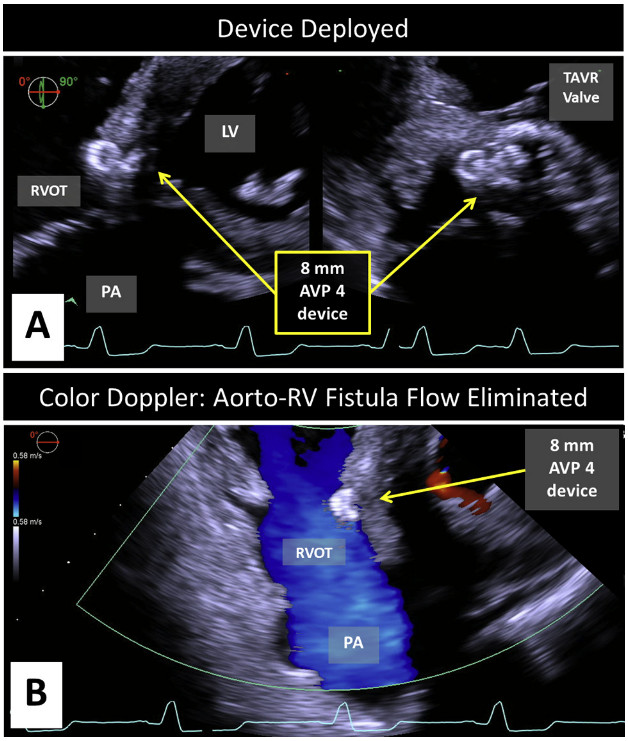

Figure 6 Two-dimensional TEE of aorto-RV fistula closure: final result. **(A)** Biplane TEE demonstrating a well-seated AVP 4 device on both sides of the aorto-RV fistula defect. **(B)** Color flow Doppler confirms no residual flow across the prior site of aorto-RV fistula. *LV*, Left ventricle; *PA*, pulmonary artery.

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