A Sharp Right Turn: A Case of Aortic to Right Atrial Fistula During Transcatheter Aortic Valve Replacement

Benjamin M. Kristobak
Theodore J. Cios

Corresponding Author: Theodore J. Cios, e-mail: tcios@pennstatehealth.psu.edu

Financial support: This report was supported with a grant from the authors’ clinical department

Conflict of interest: None declared

Patient: Female, 91-year-old
Final Diagnosis: Fistula
Symptoms: None
Medication: —
Clinical Procedure: Transcatheter aortic valve replacement
Specialty: Anesthesiology

Objective: Rare disease
Background: Aorto-atrial fistulas (AAF) are rare lesions typically associated with paravalvular abscesses or aortic aneurysms. Iatrogenic AAFs have been described after cardiac surgery. While these lesions are often asymptomatic, they can cause shunting and volume overload. Diagnosis of AAFs can be challenging. Transesophageal echocardiography plays a critical role in their diagnosis.

Case Report: A 91-year-old man undergoing transcatheter aortic valve replacement (TAVR) for severe aortic stenosis had extreme tortuosity of the aorta and iliofemoral vessels. The patient developed a fistula from the non-sinus of Valsalva to the right atrium during the procedure. After the procedure, the patient developed stroke and retroperitoneal hematoma.

Conclusions: This case represents the first full report of an aorta to right atrial fistula after TAVR. The anatomy of the aortic root in relation to the right atrium and ventricle may make aorta to right ventricle fistulas more common than aorta to right atrial fistulas. This patient’s vascular tortuosity may have played a role in the development of this lesion. Blood flow in an aorta to right atrial fistula occurs during both systole and diastole, making both right and left ventricle overload possible. Echocardiography is essential to the diagnosis of these lesions. Both vascular injury and landing zone rupture are possible during TAVR, although the observed timing and anatomy of this lesion suggest that it was caused during retrograde access of the left ventricular outflow tract via the ascending aorta.

Keywords: Anatomy • Aorto-Atrial Tunnel • Transcatheter Aortic Valve Replacement

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/936749
Background

Aorto-atrial fistulas (AAF) are rare lesions connecting the aorta to the cardiac atria. Reports are limited to cases and postmortem series, making their true incidence unknown [1]. In adults, AAFs are most often reported as a complication of endocarditis with paravalvular abscess, or as a complication of aortic aneurysm and dissection. Fistulas can form between the aorta and either atrium, although reports are more frequent with the right atrium (RA) [2]. Iatrogenic causes of AAF are most often associated with cardiac surgery, including valve replacement, saphenous vein graft aneurysm after coronary bypass graft surgery, and atrial septal defect closure [2-4]. AAFs are often asymptomatic, and once AAFs are identified, the patients are observed for clinical deterioration. Larger AAFs can lead to left- to right-sided shunting and volume overload [1,5,6].

The diagnosis and treatment of AAFs can be challenging. AAFs are typically small, and the direction of the shunt can change direction throughout its course, making it difficult to observe the entire course of the lesion via standard echocardiography views [1,2]. The rarity of this lesion could lead to it being misclassified as a more common lesion associated with cardiac surgery, such as a paravalvular leak. Transesophageal echocardiography (TEE) plays a key role in the detection of AAFs and has a high sensitivity, owing to the proximity of the esophagus to the atria [1,7-9]. AAFs are typically repaired surgically, although transcatheter interventions have also been described [1,5,6]. We report an iatrogenic aorta to right atrial fistula that developed during transcatheter aortic valve repair (TAVR).

Case Report

A 91-year-old man with a history of hypertension and former heavy tobacco use presented with shortness of breath and inability to perform independent activities of daily living. He had no prior cardiac surgeries or history of anesthetic complications. The physical examination revealed a 3/6 systolic murmur that was loudest at the left upper sternal boarder, and the patient was referred for evaluation by transthoracic echocardiography (TTE). The patient was found to have aortic stenosis and insufficiency. A TEE was obtained to better characterize the aortic stenosis prior to the procedure. The patient was found to have severe aortic stenosis, with a calculated aortic valve area of 0.55 cm², a mean gradient of 30 mm Hg, and a dimensionless index of 0.18. The patient also had aortic insufficiency, with a deceleration slope of 248 cm/s². No aortic insufficiency vena contracta was reported in this study, which was performed at an outside hospital. The patient had a left ventricle (LV) ejection fraction of 60% to 65%, no regional wall motion abnormalities, moderate concentric LV hypertrophy, grade 1 LV diastolic dysfunction, and normal right ventricular (RV) systolic function. Preoperative heart catheterization was challenging owing to extreme tortuosity of the aorta and iliofemoral vessels. It was not possible to engage the right coronary artery at that time, and it was only reported that the circulation was right dominant and that the right coronary artery was grossly patent. Left coronary artery disease was less than 50% occlusive. A preoperative computed tomography angiogram noted tortuous common and external iliac arteries.

The patient underwent TAVR under monitored anesthesia care. Left femoral and left subclavian access was obtained after we discussed that the previously demonstrated tortuosity of the aorta and iliofemoral arteries could preclude access to the LV outflow tract (LVOT) by femoral access. Tortuosity of the aorta and iliofemoral vessels was indeed noted on angiography (Figure 1). Initial attempts to access to the LVOT from wires and catheters placed in the left subclavian artery revealed a waveform characteristic of the RV. Unusual friction was also noted with wire advancement in this position. General endotracheal anesthesia was induced at this time to allow a TEE to be performed to guide the wire and catheter placement. Immediately, a periaortic root hematoma was noted (Figure 2). The patient was observed for 1 h, and the hematoma did not expand. In consideration of the patient’s age and frailty, the procedure continued to attempt to stabilize any aortic root injury and to avoid surgical intervention on the atrioventricular. Left axillary access was obtained and used to place a 23-mm Edwards Sapien Ultra valve (Edwards Lifescience, Irvine, CA, USA). No paravalvular leak was noted. After deployment of the valve, color Doppler assessment revealed flow from the non-coronary sinus of Valsalva to the RA (Figure 3). Flow through the fistula was noted to be primarily during systole by continuous wave Doppler, although some diastolic flow was also likely (Figure 4). The patient was observed for 2 h after deployment of the valve. No other complications arose, and the patient was taken to the Intensive Care Unit (ICU) intubated for recovery.

Figure 1. Iliofemoral arteriogram demonstrates extreme tortuosity of the patient’s vasculature.
Figure 2. Periaortic hematoma located at the aortic root. The hematoma was noted after many unsuccessful attempts to access the left ventricular outflow tract with wires and catheters via the ascending aorta and prior to balloon valvuloplasty.

Figure 3. Color flow Doppler demonstrating the angulated course of the aorto-atrial fistula.

Figure 4. Continuous wave spectral Doppler profile of the blood flow through the aorta to right atrial fistula.
After arrival in the ICU and weaning of sedation, the patient was noted to have right facial droop and right hemiplegia. A grade 3 systolic murmur at the left lower sternal boarder, with no rubs, gallsops, or thrills, was noted on physical examination at this time. A noncontrast computed tomography scan of the head revealed left middle cerebral artery stroke. The patient was not a candidate for thrombolytic therapy owing to the aortic root hematoma. On post-procedural day 1, the patient became hypotensive. TEE was negative for tamponade, and the patient was treated with vasoactive infusions. On post-procedural day 2, the patient developed anemia and thrombocytopenia. A computed tomography scan revealed a large, bilateral retroperitoneal hematoma extending from the level of the kidneys into the pelvis. The patient remained lethargic, without improvement of his neurologic symptoms. The patient was extubated in accordance with his wishes on post-procedural day 7 and died shortly after.

**Discussion**

There is only one other brief report of aorta to right atrial fistula after TAVR that we identified in the literature [10]. Descriptions of ventricular septal defects [11] and aorta to RV fistulas [12-15] seem to be more common. The limitations of using case reports to determine the incidence of a lesion makes it difficult to draw conclusions whether aorta to RV fistulas are more common than aorta to RA fistulas. It may be true that aorta to RA fistulas are less likely to be clinically significant and require surgical or percutaneous intervention, making reports less common. There has been no observational study that describes the incidence of these lesions [1,2], let alone after TAVR. Because these lesions are rare, a very large number of patients would need to be observed to accurately determine incidences. However, given that there were almost 73,000 TAVRs performed in the United States in 2019 [16], it seems unlikely that repair of these lesions would go unreported if they are in fact as common as aorta to RV fistula after TAVR.

The ascending aorta, aortic root, RA, and RV are all in close proximity at the base of the heart. The RV is close to the right coronary sinus of Valsalva, while the RA is located basally, seemingly next to rather than above the aortic root. This orientation could make aorta to RV fistula formation more likely in TAVR than aorta to RA fistula formation. A wire traversing the ascending aorta in a retrograde manner would likely be oriented toward the more apically oriented RV rather than the basally oriented RA. Figure 5 shows a white wire placed through the right coronary cusp into the right ventricular outflow tract, and a red wire placed through the non-coronary cusp into the RA of a pig heart. The red wire requires a much more angulated course to reach the RA than the white wire requires to reach the RV. The angle taken by the red wire requires a sharp bend if the more proximal end of the wire is to remain inside of the ascending aorta, which was resected for this dissection. This bend would be unlikely to occur with a wire being pushed from its opposite end. Our particular patient’s vascular tortuosity could have allowed an unusual orientation of the wire in the ascending aorta that made a straight orientation from the non-coronary sinus of Valsalva to the RA more likely. These anatomic variations and the inability to detect them on the procedural fluoroscopy highlight the importance of TEE in the detection and evaluation of AAF in TAVR, similar to previous reports of AAF [7-9].

The timing of blood flow in aorta to RA fistulas is important to the clinical manifestations in aorta to RA fistulas. Aortic pressure is higher than right atrial pressure in both systole and diastole. Thus, it would be expected that blood flow through such a fistula would be continuous, with greater flow during systole [17]. This pattern was seen in the present case (Figure 4). Systolic flow occurs when the pulmonic valve is open. Thus, it may be expected that left ventricular overload would be more likely to occur. However, the presence of diastolic flow may mean that right-sided overload is also possible in patients with compromised RV systolic function. The presence or absence of shunting and volume overload was key to the decision of whether to intervene once the fistula was discovered.

The presence of a periaortic hematoma prior to valvuloplasty suggested that an injury occurred during placement of wires and catheters rather than during deployment of the valve. It
is possible, however unlikely, that this AAF represented a fracture of the aortic annulus or aortic root. Vascular injury and device landing zone rupture are well-known complications of TAVR. Acute uncontained rupture can be fatal and often requires emergent surgical intervention with cardiopulmonary bypass. Contained ruptures are more likely to manifest as a paravalvular leak from the aorta into the LVOT. Oversizing the final prosthetic atrioventricular size greater than 120% of the initial annulus and a high level of calcification in the LVOT are risk factors for device landing zone rupture [18].

The transfemoral approach for TAVR is the access route of choice and is associated with superior results [19,20]. Tortuous iliofemoral anatomy can preclude the use of the route. TAVR can also be considered via the transaortic, transapical, transcatheter, and transsubclavian routes [21]. Transapical and transapical TAVRs require surgical approaches and are associated with higher mortality than transfemoral approaches [21-23]. Transcarotid and transapical approaches avoid the need for surgical access to the thorax [21]. Transaxillary approach appears to have improved outcomes to surgical nonfemoral approaches [24,25] and similar outcomes to transfemoral TAVR [26], with the possible exceptions of small differences in midterm all-cause mortality [25] and stroke [27]. These considerations made the transapical approach an attractive choice for our frail patient with multiple comorbidities who wished for intubation and a long hospital stay. Femoral access was also obtained to facilitate monitoring and to allow for access, should that route be feasible. It was clear from the initial angiograms that the tortuosity of the aorta and iliofemoral vessels precluded the use of the transfemoral route for placement of the TAVR valve. Still, the transapical approach is subject to tortuosity in the proximal aorta. The transapical approach may have avoided the possibility of malalignment of the catheters in the ascending aorta if this was indeed the cause of the fistula. However, it would have exposed the patient to other complications associated with a surgical approach. It may be reasonable to use TEE to guide wire and catheter placement in patients with tortuous aortas when a transsubclavian approach is selected. This would require utilizing general anesthesia from the start of the case.

Conclusions

Aorta to right atrial fistulas are now known to be a rare complication of TAVR, even among the already rare iatrogenic intracardiac fistulas. Anatomic relationships likely make aorta to RA fistula uncommon, but aortic tortuosity could make it more likely. The challenges in diagnosing and treating these lesions are likely similar to those of AAF associated with endocarditis and surgery. This means that echocardiography likely plays an important role in the detection and evaluation of these lesions, both during and after TAVR. It may be important to be aware of these rare lesions when diagnosing new abnormalities in color flow Doppler after TAVR.

Institution and Department Where Work Was Done

Department of Anesthesiology and Perioperative Medicine, Penn State Hershey Medical Center, Hershey, PA, USA.

Statement

This case report was supported by a grant from the authors clinical department. The authors have no financial or other relationships with the commercial devices discussed in this report.

Declaration of Figures’ Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

References:

1. Fierro EA, Sikachi RR, Agrawal A, et al. Aorto-atrial fistulas: A contemporary review. Cardiol Rev. 2018;26(3):137-44
2. Jainandunsing JS, Linnemann R, Bouma W, et al. Aorto-atrial fistula formation and closure: A systematic review. J Thorac Dis. 2019;11(3):1031-46
3. Ahmad T, Chithiraichelvan S, Patil TA, Jawali V. Aortic root to left-atrial fistula after aortic valve replacement: A rare complication and its intraoperative management. Ann Card Anaesth. 2014;17(2):155-56
4. Alkhouri M, Almustafa A, Kawara T, Tarabishy A. Transcatheter closure of an aortoatrial fistula following a surgical aortic valve replacement. J Card Surg. 2017;32(3):186-89
5. Alkhouri M, Almustafa A, Kawara T, Tarabishy A. Transcatheter closure of an aortoatrial fistula following a surgical aortic valve replacement. J Card Surg. 2017;32(3):186-89
6. Aoyagi S, Akashi H, Kawara T, et al. False aneurysm of the ascending aorta with fistula to the right atrium. Noninvasive diagnosis by computed tomographic scan and two-dimensional echocardiography with successful repair. Thorac Cardiovasc Surg. 1994;42(1):58-60
7. Hill EE, Herijgers P, Claus P, et al. Abscess in infective endocarditis: The value of transthoracic echocardiography and outcome: A 5-year study. Am Heart J. 2007;154:923-28
8. Ananthasubramaniam K. Clinical and echocardiographic features of aortoatrial fistulas. Cardiovasc Ultrasound. 2005;3:1
9. Patel V, Fountain A, Guglin M, Nanda NC. Three-dimensional transthoracic echocardiography in identification of aorto-right atrial fistula and aorto-right ventricular fistulas. Echocardiography. 2010;27(9):E105-8
10. Musa, A Suboc, M, Marinescu K, et al. Aorto-right atrial fistula following percutaneous transcatheter aortic valve replacement. J Am Coll Cardiol. 2020;75(11 Supplement. 1):2357
11. Ando T, Holmes AA, Taub CC, et al. Iatrogenic ventricular septal defect following transcatheter aortic valve replacement: A systematic review. Heart Lung Circ. 2016;25(10):968-74
12. Alabady AM, Sattur S, Bauch TD, Harjai KJ. Aorto-right ventricular fistula and paravalvular leak after transcatheter aortic valve implantation. JACC Case Rep. 2019;1(5):859-64
13. Almanfi A, Qurie A, Strickman N. Aorto-right ventricular shunt after TAVR: Rare complication of common procedure. Case Rep Cardiol. 2017;2017:1834394
14. Konda MK, Kalavakunta JK, Pratt JW, et al. Aorto-right ventricular fistula following percutaneous transcatheter aortic valve replacement: Case report and literature review. Heart Views. 2017;18(4):133-36
15. Shakoor MT, Islam AM, Ayub S. Acquired aorto-right ventricular fistula following transcatheter aortic valve replacement. Case Rep Cardiol. 2015;2015:608539
16. Glenn K. Report finds TAVR is dominant form of aortic valve replacement, outcomes steadily improving in the United States. American College of Cardiology Press Release. Published November 16, 2020. Accessed March 4, 2022. [https://www.acc.org/about-acc/press-releases/2020/11/16/18/53/report-finds-tavr-is-dominant-form-of-aortic-valve-replacement-outcomes-steadily-improving-in-the-us#](https://www.acc.org/about-acc/press-releases/2020/11/16/18/53/report-finds-tavr-is-dominant-form-of-aortic-valve-replacement-outcomes-steadily-improving-in-the-us#)
17. Ananthasubramaniam K, Karthikeyan V. Aortic ring abscess and aortoatrial fistula complicating fulminant prosthetic valve endocarditis due to Proteus mirabilis. J Ultrasound Med. 2000;19(1):63-66
18. Scarsini R, De Maria GL, Joseph J, et al. Impact of complications during transfemoral transcatheter aortic valve replacement: How can they be avoided and managed? J Am Heart Assoc. 2019;8(18):e013801
19. Zhan Y, Saadat S, Soin A, et al. A meta-analysis comparing transaxillary and transfemoral transcatheter aortic valve replacement. J Thorac Dis. 2019;11(12):5140-51
20. Madigan M, Atoui R. Non-transfemoral access sites for transcatheter aortic valve replacement. J Thorac Dis. 2018;10(7):4505-15
21. Overchouk P, Modine T. Alternate access for TAVI: Stay clear of the chest. Interv Cardiol. 2018;13(3):145-50
22. Kodali S, Thurani VH, White J, et al. Early clinical and echocardiographic outcomes after SAPIEN 3 transcatheter aortic valve replacement in inoperable, high-risk and intermediate-risk patients with aortic stenosis. Eur Heart J. 2016;37(28):2252-62
23. Aral T, Romano M, Lefèvre T, et al. Direct comparison of feasibility and safety of transfemoral versus transaortic versus transapical transcatheter aortic valve replacement. JACC Cardiovasc Interv. 2016;9(22):2320-25
24. Price J, Bob-Manuel T, Tafur J, et al. Transaxillary TAVR leads to shorter ventilator duration and hospital length of stay compared to transapical TAVR. Curr Probl Cardiol. 2021;46(3):100624
25. Takagi H, Hari Y, Nakashima K, et al; ALICE (All-Literature Investigation of Cardiovascular Evidence) Group. Comparison of early and midterm outcomes after transsubclavian/axillary versus transfemoral, transapical, or transaortic transcatheter aortic valve implantation. Heart Lung. 2019;48(6):519-29
26. Zhan Y, Toomey N, Ortoleva J, et al. Safety and efficacy of transcatheter aortic valve replacement using a current-generation balloon-expandable valve. J Cardiothorac Surg. 2020;15(1):244
27. Faroux L, Junquera L, Mohammadi S, et al. Femoral versus nonfemoral subclavian/carotid arterial access route for transcatheter aortic valve replacement: A systematic review and meta-analysis. J Am Heart Assoc. 2020;9(19):e017460