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Platypnea–Orthodeoxia Syndrome Following Transcatheter Aortic Valve Replacement

CASE REPORT

INTRODUCTION

Transcatheter aortic valve replacement (TAVI) is a well-established treatment modality in patients with aortic valve stenosis who require valve replacement.[1] However, as for each interventional procedure, this novel treatment procedure has different types of complication risks, including cerebrovascular events, vascular complications, bleeding, coronary obstruction, myocardial infarction, valve regurgitation, valve malpositioning or migration, conduction disturbances, and acute kidney injury, which may occur during and/or after the procedure.[2,3] These complications could be seen in up to one-third of the patients, and some of them may need urgent surgical intervention and may have a higher risk of death.[1-3] We herein described this rare case of a 79-year-old woman presenting with worsening shortness of breath following the TAVI procedure, and we discussed the diagnostic process and therapeutic course of the case.

CASE REPORT

A 79-year-old lady with severe aortic stenosis judged at high risk for surgery was refrerred for TAVI to our center. Symptoms included presyncope episodes leading to frequent falls, orthopnea, and exertional chest pain refractory to medical therapy. Past medical history included hypertension and dyslipidemia. Transthoracic echocardiography (TTE) confirmed severe aortic stenosis (Vmax 4.16 m/s, mean gradient 54 mm Hg, maximum gradient 77 mm Hg) with a calcified trileaflet valve, and additionally, the systolic function was normal. There was a interatrial septum (IAS) aneurysm without shunt using bubble test or color Doppler. Then, she underwent transesophageal echocardiography (TEE) imaging to see the baseline morphological and hemodynamic assessment of the calcific aortic valve; at the same time, IAS was also assessed (Figure 1A). Results were similar with TTE regarding IAS, and it did not show any shunt (Figure 1B). The Society of Thoracic Surgeons (STS) Score was 3%; however, our heart team recommended TAVI rather than open surgery. The pre-operative multi-slice computed tomography (MSCT) evaluation of the aorta and its branches confirmed the patency of femoral arteries that allowed a transfemoral approach.

A Sapien 3 Ultra (Edwards Lifesciences, Irvine, Calif, USA) 26 mm was directly implanted without post-dilatation and paravalvular leak. Post-procedural TTE confirmed an excellent result without any complications. The patient was scheduled for discharge; however, she had an episode of partial amnesia just before leaving. She was consulted by neurologists, and then she underwent a cranial MSCT scan, which did not show any acute hemorrhagic or ischemic lesions. Her postoperative course was unremarkable, and she was discharged well with a neurology follow-up after 7 days. Less than 3 months after following the TAVI procedure, she was admitted to the emergency room with an inability to feed herself and dyspnea which worsened when standing upright with reduced oxygen saturation. On admission, the patient's oxygen saturation was 79% on room air in an upright position, which did improve in a supine position. Oxygen saturation slightly improved to 86% with an inspired oxygen concentration of 100%. A physical exam revealed a Grade 3/6 holo systolic murmur. An arterial blood gas

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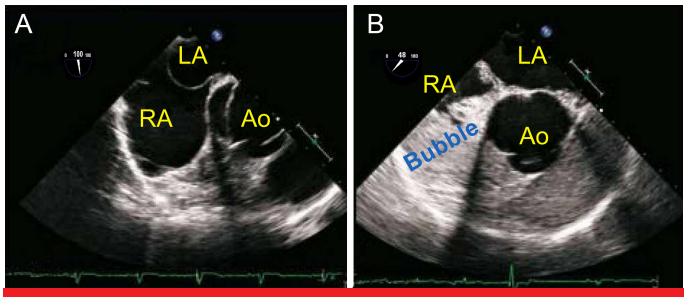

Figure 1. Transesophageal echocardiography imaging before the TAVI showed IAS aneurysm (A), whereas no bubbles are present on the LA side during a bubble study, even with the Valsalva maneuver (B). IAS, interatrial septum; LA, left atrium; TEE, transesophageal echocardiography.

analysis was significant for marked hypoxemia with oxygen tension of 45 mm Hg. Chest radiography was within normal limits. Electrocardiogram showed normal sinus rhythm without any ischemia changes. CT angiography was negative for pulmonary embolism and showed no partial filling defect. Since the patient had an increasing O_2% amount to the average level by lying flat, TTE was performed. It did not reveal any issue with the prosthesis but showed moderate left atrial dilatation (44 mm, 108 mL, 41 cm^2), in addition to interatrial septal aneurysm, but the right chambers were within normal limits. The mean transaortic gradient was 3 mm Hg, similar to the measured following TAVI. Transesophageal echocardiography revealed a bulging interatrial septum along with a significant right-to-left interatrial shunt due to patent foramen ovale (PFO) (Figure 2) that was not present when compared to previous TEE before TAVI (Figure 1A and 1B).

Flow across the PFO was significantly higher in the upright than the supine position on TTE imaging, consistent with the lower oxygen saturation during the upright position. Right and left heart catheterizations were performed on the second day of the presentation to measure shunt fraction and left atrial oxygen saturation. Cardiac catheterization revealed the following hemodynamic measurements: pressures of right atrial 6 mm Hg; right ventricular 25/3; mean pulmonary artery pressure 15/2 (12); pulmonary artery capillary wedge pressure 7; and peripheral vascular resistance, 3.9 woods (Fick) and cardiac output, 4.8 L/min. Additionally, the hyperoxia test showed a pathological shunt rate of about 30%-35%. Given these findings, the patient with a diagnosis of platypnea–orthodeoxia syndrome (POS) was referred for PFO closure percutaneously. The patient's symptoms resolved following successful percutaneous PFO closure using a 35 mm Cardia Ultrasept (Cardia Inc., MN, USA) PFO occlusion device (Figure 3). The patient's dyspnea immediately improved. At the time of discharge, the patient had an oxygen saturation of 95% on room air, and she was able to maintain it with moderate exercise. The patient has given her consent for her images and other clinical information to be reported.

DISCUSSION

Transcatheter aortic valve has become the treatment of choice for inoperable patients, nevertheless, specific periprocedural and late cardiac (5%-61%) and noncardiac (5%-84%) complications may occur.[1] However, the occurrence of intracardiac shunts post-TAVI due to changes in cardiac physiology and anatomy is poorly described. The current case was admitted with increased shortness of breath, like POS following a successful TAVI procedure. Due to the pathophysiology of orthodeoxia, paradoxical embolism is a frequent coexisting condition, which may explain a brief episode of amnesia.

Platypnea–orthodeoxia syndrome is an uncommon clinical cause of dyspnea and hypoxemia, and the primary mechanisms can be broadly classified as intracardiac or extracardiac abnormalities and miscellaneous etiologies.[4] Platypnea–orthodeoxia syndrome has also been described in a patient who had undergone recent surgical aortic valve replacement, attributed to the presence of significant aortic dilatation.[5] Of note, symptoms may occur due to pulmonary-based, such as embolism and pulmonary hypertension (PHT); however, the current case had a CT angio which showed no filling defect. Besides, this mentioned incidence of a PFO in the PHT population increases with more dilated and dysfunctional RVs, suggesting that the PFO may be stretched

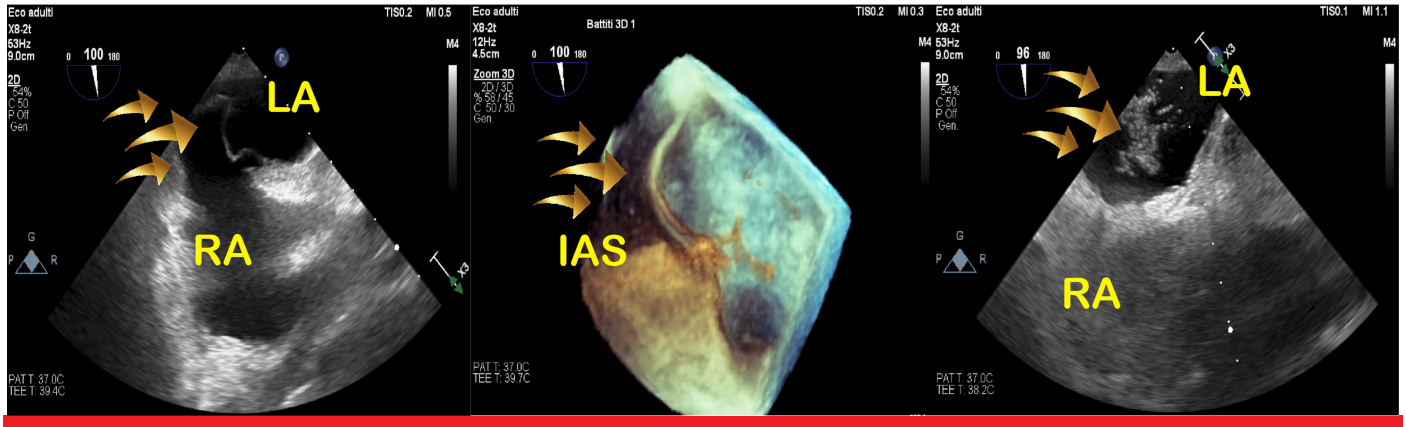

Figure 2. Left and middle images: 2D and 3D TEE images show severe IAS aneurysm. Right: Arrows showing the bubbles in the RA just after injection via the right femoral vein. IAS, interatrial septum; RA, right atrium; TEE, transesophageal echocardiography.

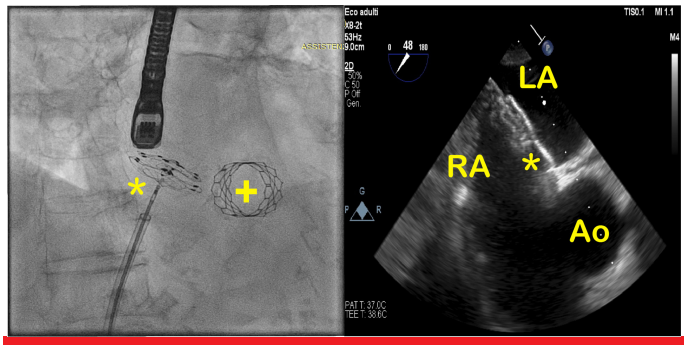

Figure 3. Left: Fluoroscopy imaging showing 35 mm septal occlusion device *in situ* before deployment, with Edwards S3 Ultra 26 mm transcatheter aortic valve device (+) and transesophageal probe visible. Right: TEE image showing the postprocedure negative bubble study with PFO occlusion device (*) *in situ*. PFO, patent foramen ovale; TEE, transesophageal echocardiography.

open rather than post-TAVI complication and congenital;[6] however, there was no remarkable dilatation on the right chambers. There are limited reports and conference papers in the literature regarding this post-TAVI complication, and the chief complaint for hospital admissions is mostly similar.[7] It is obvious that there are physiological, anatomical, and electrical changes following TAVI, and many hypotheses have been put forward to explain orthodeoxia in the presence of these secondary cardiopulmonary factors. There is also a pathophysiological explanation of post-TAVI PFO, like an anatomic modification of the aorta (dilatation, aneurysm, or distortion), that leads to right atrial compression.[5] Regarding another explanatory factor, aortic valve stenosis leads to raised left ventricular filling pressures, which results in increased left atrial pressure overload and dilatation. Cardiac MRI studies demonstrated reverse left ventricular remodeling after TAVI in a short time period, with reductions in cardiac volume and mass and improved left ventricular ejection fraction. These changes may improve left ventricular compliance and diastolic pressure, together with distortion of the right atrium by the aortic root elongation, contributing to the clinical manifestation of PFO as a POS.[8]

Interatrial shunts following TAVI are uncommon; however, these findings raise attention to consider the relevance of aortic root anatomy in the associations between atrial septal characteristics; besides, aortic aneurysms have been described to increase atrial septal mobility and potentiate PFO shunting.[9] So, the reverse structural remodeling of the atrium may increase PFO shunting during post-TAVI follow-up and may cause POS. Therefore, clinicians should consider this potentially reversible condition in patients with unexplained dyspnea and associated with high mortality if left untreated. Although data is limited for this rare syndrome, percutaneous closure has thus far proven safe and effective.

Informed Consent: The patient has given her consent for her images and other clinical information to be reported.

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REFERENCES

1. Leon MB, Smith CR, Mack M, et al. Transcatheter aortic-valve implantation for aortic stenosis in patients who cannot undergo surgery. *N Engl J Med*. 2010;363(17):1597-1607. [CrossRef]
2. Winter M-P, Bartko P, Hofer F, et al. Evolution of outcome and complications in TAVR: a meta-analysis of observational and randomized studies. *Sci Rep*. 2020;10:1-9.
3. Neragi-Miandoab S, Michler RE. A review of most relevant complications of transcatheter aortic valve implantation. *ISRN Cardiol*. 2013;2013:956252. [CrossRef]
4. Chen GP-W, Goldberg SL, Gill EAJ. Patent foramen ovale and the platypnea-orthodeoxia syndrome. *Cardiol Clin*. 2005;23(1):85-89. [CrossRef]
5. Medina A, Lezo JS de, Caballero E, Ortega JR. Platypnea-orthodeoxia due to aortic elongation. *Circulation*. 2001;104(6):741. [CrossRef]
6. Sharan L, Stackhouse K, Awerbach JD, Bashore TM, Krasuski RA. Effect of patent foramen ovale in patients with pulmonary hypertension. *Am J Cardiol*. 2018;122(3):505-510. [CrossRef]
7. Kokroo A, Hou L, Cunningham TF, et al. Platypnea-orthodeoxia syndrome after transcatheter aortic valve replacement and concomitant aortic root elongation. *J Am Coll Cardiol*. 2022;79(9):2790. [CrossRef]
8. Mehdipoor G, Chen S, Chatterjee S, et al. Cardiac structural changes after transcatheter aortic valve replacement: systematic review and meta-analysis of cardiovascular magnetic resonance studies. *J Cardiovasc Magn Reson*. 2020;22(1):41. [CrossRef]
9. Heidemann A, Dall'Oglio L, Bertoldi EG, Foppa M. Increased mobility of the atrial septum in aortic root dilation: an observational study on transesophageal echocardiography. *Front Physiol*. 2021;12:701399. [CrossRef]