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MINI-FOCUS ISSUE ON VALVULAR HEART DISEASE

ADVANCED

CASE REPORT: CLINICAL CASE

Percutaneous Transcatheter Valve-in-Valve Pulmonary and Tricuspid Replacement in Carcinoid Heart Disease

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ABSTRACT

Surgical valve replacement is the most effective treatment for carcinoid heart disease; however, reoperation for prosthetic valve failure is burdened by high risk. We report the first described percutaneous transcatheter pulmonary and tricuspid valve-in-valve replacement for bioprosthesis degeneration for any reason in a patient with carcinoid heart disease. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2020;2:533–6) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 57-year-old woman with congestive heart failure secondary to carcinoid heart disease from ileum carcinoid tumor with liver metastasis and peritoneal carcinomatosis presented with 3 months of progressive breathlessness, marked jugular venous distension, hepatomegaly, and peripheral edema. Blood pressure was 105/58 mm Hg, HR was 103 beats/min, sat on air was 90%.

PAST MEDICAL HISTORY

She had tricuspid and pulmonary valve replacements with 27-mm and 25-mm CE Perimount valves (Edwards Lifesciences, Irvine, California) 9 years earlier for severe tricuspid and pulmonary regurgitation. She had dilated right heart with severe pulmonary hypertension. She had multiple transarterial chemoembolizations and somatostatin analogue peptide receptor radionucleotide therapy for metastatic control. She underwent further resections for unrelated carcinoma of the breast and bowel.

LEARNING OBJECTIVES

- To make a differential diagnosis of heart failure with consideration of the past medical history.
- To understand the safety and feasibility of the percutaneous valve-in-valve approach to patients with failed surgical valves while considering the option of further future interventions.

DIFFERENTIAL DIAGNOSIS

Prosthetic valve dysfunction, chronic pulmonary thromboembolism, chronic pulmonary disease with cor pulmonale, and acute coronary syndrome were considered.

INVESTIGATIONS

Pre-operative transthoracic echocardiography showed severe tricuspid and pulmonary prosthetic valve stenosis with a mean gradient of 11 and 18 mm Hg, respectively (**Figure 1** and **2**).

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, or patient consent where appropriate. For more information, visit the *JACC: Case Reports* author instructions page.

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ABBREVIATIONS AND ACRONYMS

ViV = transcatheter valve-in-valve

MANAGEMENT (MEDICAL/INTERVENTIONS)

Combined transcatheter valve-in-valve (ViV) pulmonary and tricuspid valve replacement was performed with perioperative octreotide infusion to reduce the risk of carcinoid crisis. Right heart catheterization was performed from the right jugular vein with a 10-F sheath. A 26-F Gore DrySeal sheath (Gore Medical, Flagstaff, Arizona) was then passed into the right pulmonary artery over a Meier wire. The stent frames of the bioprosthesis were used for reference. A balloon-mounted 26-mm Sapien 3

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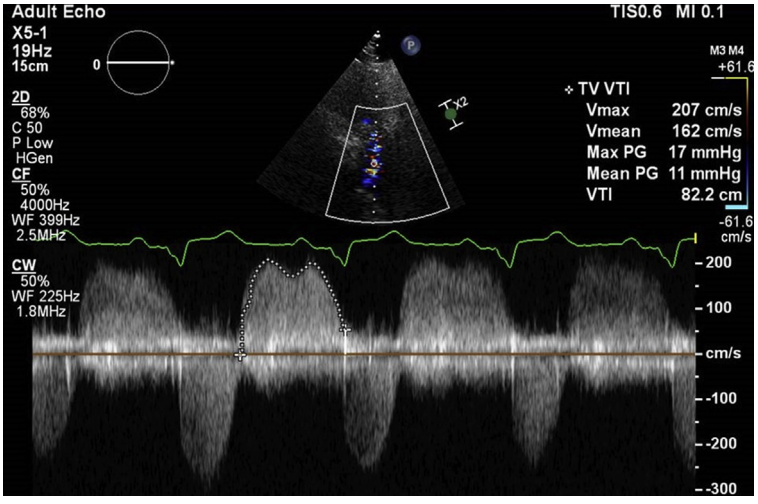
(Edwards Lifesciences) transcatheter heart valve was successfully delivered and deployed in the pulmonary position with rapid ventricular pacing (Video 2). A 29-mm Sapien 3 transcatheter valve was then successfully delivered and deployed in a tricuspid position through a 29-mm Commander delivery system (Edwards Lifesciences) from the inferior vena cava under fluoroscopic guidance with rapid pacing (Video 1). Mean gradients reduced from 11 mm to 3 mm (tricuspid) and from 18 mm to 11 mm (pulmonary), with modest reduction in right atrial, right ventricular, and pulmonary artery pressures. Post-deployment fluoroscopy and echocardiogram confirmed well-seated valves and no paravalvular regurgitation (**Figure 3**).

DISCUSSION

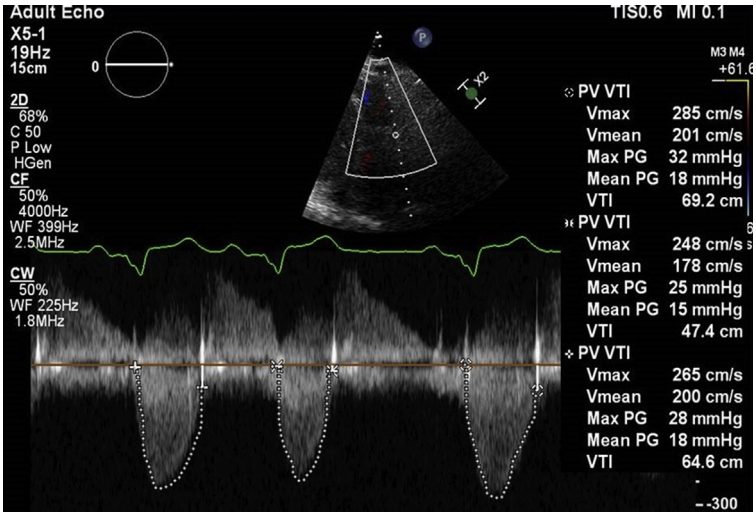
Carcinoid tumors are rare neuroendocrine tumors arising most commonly from the amine precursor uptake and decarboxylation cells in the gastrointestinal tract. Overall, 50% of patients develop carcinoid syndrome (facial flushing, intractable secretory diarrhea, and bronchoconstriction). Of these, 50% develop right heart disease (valvar, subvalvar, and endomyocardial plaques and fibrosis) due to the paraneoplastic effects of vasoactive 5-hydroxytryptamine (serotonin), histamine, tachykinins, and prostaglandins released by the malignant cells. Right heart disease develops only with hepatic metastasis because the vasoactive substances are degraded in the liver, lung, and brain. These manifestations include tricuspid and pulmonary stenosis/regurgitation and right heart dilatation and failure, necessitating valve replacements. The high burden of metastasis may be associated with early bioprosthetic failure due to circulating vasoactive substances requiring high-risk reoperation (1,2). Transcatheter SAPIEN valves (Edwards, Irvine, California) in inferior and superior vena cava for tricuspid regurgitation and transventricular Melody® valves (Medtronic, Minneapolis, Minnesota) in the right ventricular outflow tract for pulmonary stenosis/regurgitation have been used to avoid repeat sternotomy (3-6). The phase 4 HOVER (Heterotopic Implantation Of the Edwards-Sapien XT Transcatheter Valve in the Inferior VEna cava for the treatment of severe Tricuspid

FIGURE 1 Severe Tricuspid Bioprosthetic Stenosis



Pre-operative echocardiogram showing severe tricuspid bioprosthetic stenosis.

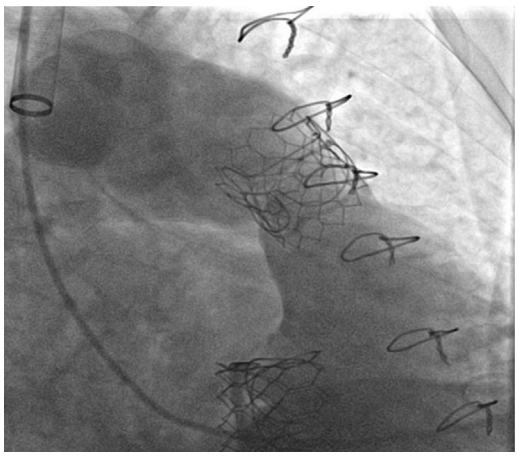
FIGURE 2 Severe Pulmonary Bioprosthetic Stenosis



Pre-operative echocardiogram showing severe pulmonary bioprosthetic stenosis.

Regurgitation) trial has shown 30 days of procedural success and 1 year of safety for the heterotropic implantation of Sapien XT for severe tricuspid regurgitation (7). Because of lack of a rigid annulus in native pulmonary and tricuspid valves, direct implantation of percutaneous valves risks dislodgement. A rigid frame of the conventional bioprosthesis provides the scaffold for secure seating. The Melody valve (Medtronic) received the Conformité Européenne mark for pulmonary position in Canada and Europe in 2006 and was approved by the U.S. Food and Drug Administration in 2010. The Sapien XT valve was approved by the U.S. Food and Drug Administration for pulmonic position in 2016. The transcatheter valves approved originally for the aortic position were used for ViV for failed bioprostheses in the right heart mostly as off-label indications. The VIVID (Valve-in-Valve International Data) registry has since shown the safety and efficacy of ViV for the tricuspid position, at least in the midterm (8). The median follow-up was 15.9 months after implantation, with 64% of patients estimated to be alive without reintervention or valve-related event at 3 years. The cumulative 3-year incidences of death, reintervention, and valve-related adverse outcomes (endocarditis, thrombosis, or significant dysfunction) were 17%, 12%, and 8%, respectively. Although rare, leaflet thrombosis was an important adverse outcome.

FIGURE 3 Valve-in-Valve Edwards Sapien 3 Valves



Valve-in-valve Edwards Sapien 3 valves within the struts of Carpentier-Edwards Perimount valves after successful deployment in the tricuspid and pulmonary positions.

FOLLOW-UP

Follow-up transthoracic echocardiography 4 months later showed well-seated tricuspid and pulmonary prostheses with no paravalvular leaks and stable gradients (Videos 3 and 4).

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CONCLUSIONS

Ours is the first reported concomitant tricuspid and pulmonary ViV implantation with Sapien 3 for bioprosthetic failure in metastatic carcinoid heart disease. Percutaneous ViV presents distinct advantages over conventional surgery with re-sternotomy and is associated with reduced procedural risk and length of stay. Durability and longevity in midterm data have been encouraging.

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KEY WORDS carcinoid heart disease, prosthetic degeneration, transcatheter valve in valve

APPENDIX For supplemental videos, please see the online version of this paper.