Case Reports

Transcatheter Pulmonary Valve Replacement in a Carcinoid Heart

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Carcinoid heart disease presents as right-sided heart failure attributable to the dysfunction of the tricuspid and pulmonary valves. Although surgical valve replacement is the mainstay of treatment when patients become symptomatic, it is associated with substantial perioperative mortality rates.

We present a case of severe pulmonary valve stenosis secondary to carcinoid heart disease, treated successfully with percutaneous valve replacement. A 67-year-old man with severe pulmonary valve stenosis was referred to our center for pulmonary valve replacement. The patient had a history of metastatic neuroendocrine tumor of the small bowel with carcinoid syndrome, carcinoid heart disease, and tricuspid valve regurgitation previously treated with surgical valve replacement.

Because of the patient's severe chronic obstructive pulmonary disease and hostile chest anatomy seen on a computed tomographic scan dating from previous cardiothoracic surgery, we considered off-label percutaneous valve replacement a viable alternative to open-heart surgery. A 29-mm Edwards SAPIEN XT valve was successfully deployed over the native pulmonary valve. There were no adverse sequelae after the procedure, and the patient was discharged from the hospital the next day. This case report shows that percutaneous valve replacement can be a valid option in carcinoid heart disease patients who are not amenable to surgical valve replacement. ***(Tex Heart Inst J 2016;43(4):341-4)***

Key words: *Carcinoid heart disease/therapy; heart valve prosthesis implantation, percutaneous; treatment outcome*

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Carcinoid heart disease (CHD) presents as right-sided heart failure attributable to dysfunction of the tricuspid valve and pulmonary valve (PV). Although surgical valve replacement is the mainstay of treatment when patients become symptomatic, it is associated with substantial perioperative mortality rates. In the transcatheter valve-replacement era, the percutaneous option has become more appealing for application to poor surgical candidates. We present a case of severe PV stenosis secondary to CHD that was treated successfully with percutaneous valve replacement.

Case Report

A 67-year-old man with severe PV stenosis was referred to our center for PV evaluation and replacement. The patient had a medical history of metastatic neuroendocrine tumor of the small bowel with carcinoid syndrome, CHD, and tricuspid valve regurgitation treated by means of surgical valve replacement with a 31-mm Mosaic® valve (Medtronic, Inc.; Minneapolis, Minn) 4 years prior. He reported increased fatigue and shortness of breath over the past several months. The patient was already oxygen dependent because of severe chronic obstructive pulmonary disease (FEV1, 35% by pulmonary function test).

Echocardiographic examination revealed severe PV stenosis (peak gradient, 55 mmHg; and peak velocity, 3.71 m/s), moderate pulmonary regurgitation, mild-to-moderate tricuspid regurgitation, and increased right ventricular (RV) systolic pressure with preserved left ventricular ejection fraction.

Poor lung function and hostile chest anatomy, revealed by a computed tomogram (CT) from prior tricuspid valve replacement, allowed the patient to undergo implantation of an Edwards SAPIEN XT transcatheter heart-tissue valve (Edwards Lifesciences LLC; Irvine, Calif) on an off-label compassionate indication. Lone pulmonary valvuloplasty was considered the second-best treatment option because of the baseline presence of moderate pulmonary regurgitation.

We chose off-label use of the SAPIEN XT valve deployed over a Palmaz® XL stent (Palmaz Scientific; Fremont, Calif), as previously described in the medical literature.

The Palmaz XL stent would be used to provide the anchor for the SAPIEN XT valve over the native PV. The initial sizing of the valve and stent was performed with the aid of cardiac CT angiography. These images confirmed severe stenosis of the PV, with a pulmonary valve orifice area of 127 mm^2 (Fig. 1). In addition, as another consideration in sizing, we measured diameters at 5 mm and 10 mm (cranially and caudally) from the pulmonary annulus into the RV outflow tract (RVOT) caudally and from the pulmonary annulus into the main pulmonary artery (PA) cranially (Fig. 2). The PA areas at 5 and 10 mm were 436 and 618 mm^2, respectively. Areas in the RVOT at 5 and 10 mm were 529 and 637 mm^2, respectively. On the basis of these measurements, the combination of the SAPIEN XT 26-mm valve and the 10 × 39-mm Palmaz XL stent was estimated to be appropriate.

Risks and benefits of the procedure were explained to the patient in detail, and informed consent was ob-

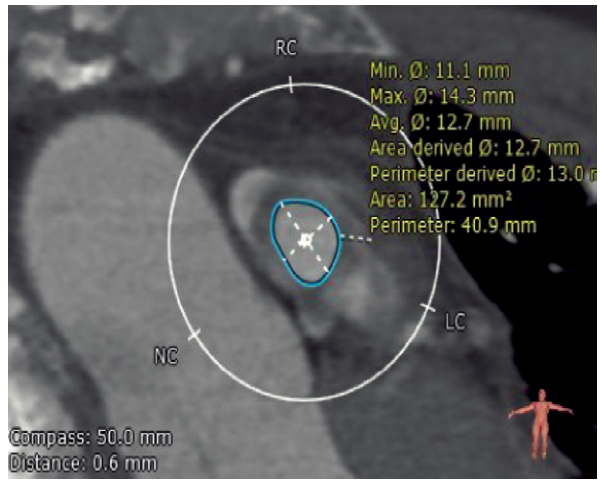

Fig. 1 *Computed tomographic angiogram shows the sizing of the pulmonary valve.*

LC = left coronary cusp; NC = noncoronary cusp; RC = right coronary cusp

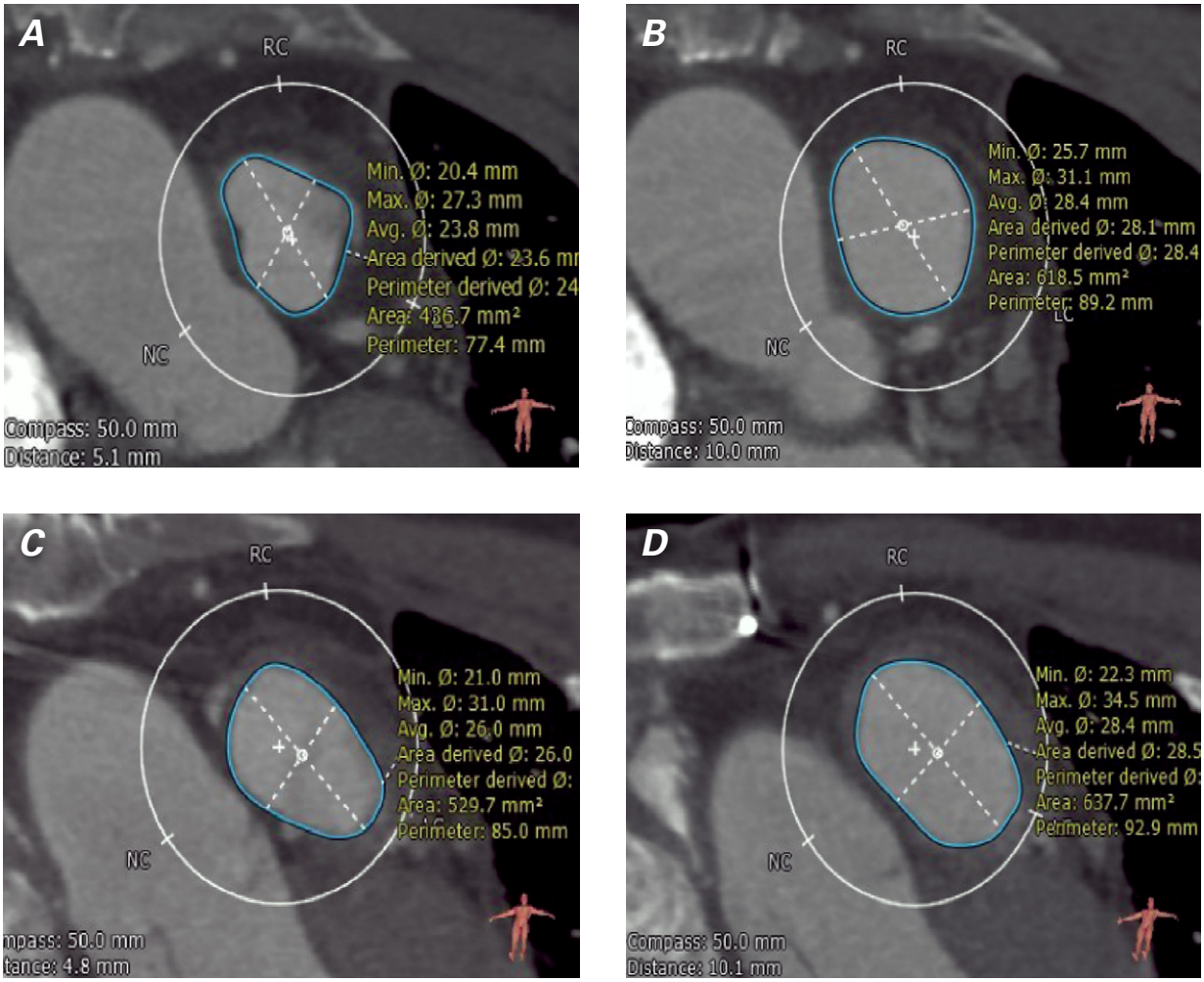

Fig. 2 *Computed tomographic angiograms show the sizing of the pulmonary valve, with pulmonary artery diameters at **A**) 5 and **B**) 10 mm, and right ventricular outflow tract diameters at **C**) 5 and **D**) 10 mm.*

LC = left coronary cusp; NC = noncoronary cusp; RC = right coronary cusp

tained. With the patient under general anesthesia, the procedure was conducted via transesophageal echocardiographic (TEE) guidance. Access was gained bilaterally through the common femoral arteries (5F sheaths) and the common femoral veins (7F sheaths). Selective left coronary angiography, performed first, showed no angiographic disease of substance in the left main, left anterior descending, or left circumflex coronary arteries. Right-sided pressures were as follows: pulmonary capillary wedge pressure, 13 mmHg, RV systolic pressure, 82 mmHg, and mean PA pressure, 30 mmHg. The Swan-Ganz catheter was exchanged, over a J-wire, for a pigtail catheter, which was then advanced to the RV. The multipurpose catheter was advanced (together with a long J-wire) to the right PA, and the J-wire was exchanged for an Amplatz Super Stiff™ wire (Boston Scientific Corporation; Natick, Mass). The 7F sheath was replaced with an 18F Edwards delivery sheath over an Amplatz guidewire in the right PA. The pulmonic valvuloplasty proceeded with deployment of a 28 × 40-mm Z-MED™ balloon (B. Braun Interventional Systems Inc.; Bethlehem, Pa). The Amplatz guidewire was exchanged for a Lunderquist® Extra Stiff Wire Guide (Cook Medical Inc.; Bloomington, Ind) to obtain additional support. The 10 × 39-mm Palmaz XL stent was advanced across the PV and deployed in an effort to provide an anchor for the SAPIEN valve.

As the stent was being deployed, it migrated forward as we pulled back the balloon, which ultimately resulted in only partial deployment of the stent. A 26 × 45-mm True Dilatation™ Balloon (Bard Peripheral Vascular, Inc.; Tempe, Ariz) was followed by final postdilation of the migrated Palmaz stent with a 40-mm Coda® balloon (Cook Medical) against the main PA wall. However, RV angiography confirmed that the stent had migrated distally in the main PA and was not across the PV.

Given that a 10 × 39-mm Palmaz XL stent had migrated forward, we decided to increase the size of the SAPIEN XT valve to 29 mm. An 18F Edwards introducer was exchanged for a 20F Edwards introducer in order to accommodate the 29-mm NovaFlex delivery system (Edwards). The valve was advanced over the Lunderquist wire and was successfully deployed across the PV (Fig. 3). Despite the off-target deployment of the anchoring Palmaz XL stent and direct stenting of the native PV with a 29-mm SAPIEN XT, an intraoperative TEE after the deployment indicated accurate positioning of the deployed valve, a transpulmonary gradient of 8 mmHg, and trivial pulmonary regurgitation across the valve. There were no complications after the procedure and the patient was discharged from the hospital the next day with instructions to take dual antiplatelet therapy of aspirin and clopidogrel.

At a follow-up clinical appointment, 30 days after the procedure, transthoracic echocardiography revealed preserved RV systolic function; the bioprosthetic PV appeared to be well seated and functioning properly. The patient reported that his respiratory status had been gradually improving. Clopidogrel was discontinued, and the patient remained clinically stable at his 5-month follow-up appointment.

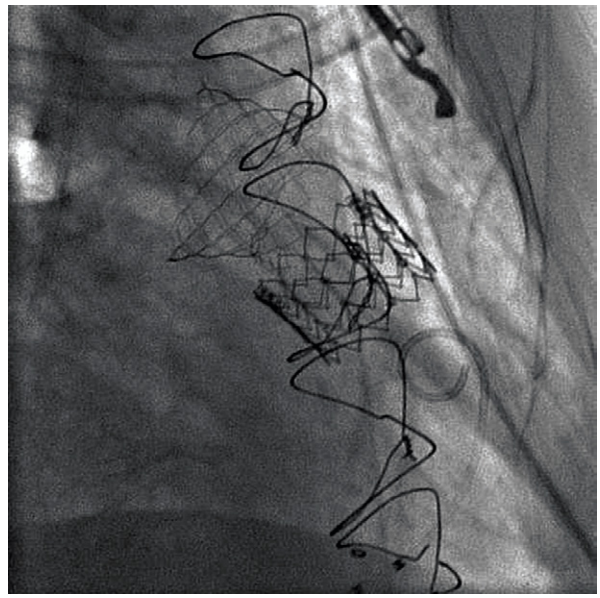

Fig. 3 *Right ventricular angiogram (right anterior oblique view) shows the Palmaz stent dilated against the main pulmonary artery and the SAPIEN XT valve deployed successfully across the pulmonary valve.*

Discussion

Carcinoid tumors are rare neuroendocrine tumors derived from the enterochromaffin cells, affecting 1.2 to 2.1 per 100,000 members of the general population.[1] First described in the 1950s,[2] CHD occurs in 50% to 60% of patients with carcinoid syndrome, thereby accounting for substantial illness and death.[3] The time period between the onset of symptoms and the diagnosis of CHD is about 2 years.[4] The cause of valve injury in CHD is not completely understood, but the high level of circulating serotonin is most likely the major contributor.[3,4] Typically, fibrous plaque-like endocardial thickening causes retraction and fixation of the right-sided heart valves, thereby leading to right-sided heart failure.

Surgical valve replacement is associated with a high perioperative mortality rate: 18% to 20%.[3-5] Valve replacement should be considered in patients who have severely dysfunctional valves or are refractory to medical therapy. In our case of severe PV stenosis not amenable to surgical replacement because of the patient's high perioperative risk, we showed that percutaneous valve replacement could be a valid option. It should be noted that CT sizing of the annulus can result in underestima-

tion of the annular dimension necessary for anchoring the stent-valve system. The pulmonary annulus is more compliant than is the aortic annulus; therefore, angiographic and noninvasive methods are routinely used to confirm sizing[6,7] before valve deployment. Because commercially available valves are mounted on balloon-expandable stents designed for the aortic annulus and the aortic wall, a rigid landing zone is necessary and pre-stenting is widely used to facilitate pulmonary valve anchoring.[6,7] As a result, the 26-mm stent and 26-mm Palmaz XL that we at first used were grossly undersized and could not anchor, which ultimately required our use of a 29-mm XT as an effective alternative.

In conclusion, our report emphasizes an excellent outcome in a patient with CHD and native PV stenosis, after treatment with percutaneous valve replacement.

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