Percutaneous pulmonary valve replacement is one of the most important advancements in the field of interventional cardiology in the past decade. However, currently available technologies are not applicable to patients with oversized right ventricular outflow tracts (RVOTs), especially when there is concomitant proximal branch pulmonary artery (PA) stenosis. This combination is encountered in clinical practice. Our group has a longstanding interest in these complex patients, and we have recently published preclinical studies describing the feasibility and short-term effects of implanting 2 Melody valves—1 into each proximal branch PA—in an ovine model of postoperative pulmonary insufficiency and dilated RVOT. This report describes bilateral branch PA Melody valve implantation to treat complex RVOT dysfunction in a high-risk patient.

History
At presentation, the patient was a 27-year-old woman with a complex medical history. Her cardiac condition was double-outlet RV with pulmonary stenosis and interrupted inferior vena cava with azygos continuation to a left-sided superior vena cava. She was status post-Waterston shunt as an infant, with subsequent closure of the ventricular septal defect and patch augmentation of the RVOT. Three months before catheterization, the patient presented to her cardiologist with symptoms of progressive activity intolerance over the past year. She was oxygen dependent and on long-term continuous positive airway pressure therapy. Comorbidities included VATER and Klippel-Feil syndromes, renal agenesis, Müllerian agenesis, gout, and obesity. The patient had undergone multiple spinal fusion and Harrington rod procedures as well as repair of tracheoesophageal fistula. An echocardiogram was obtained but was nondiagnostic because of her body habitus. A limited cardiac CT scan obtained at an outside hospital revealed dilated RV and left ventricle (LV). The RVOT was large, and there were complex stenoses involving the origins of both the right and left branch PAs. Pulmonary function studies revealed severe restrictive and obstructive lung disease.

After discussion at our multidisciplinary rounds, we decided to bring the patient to our combined MRI-catheterization suite for a diagnostic cardiac MRI followed by invasive assessment. Because of her comorbidities, the patient was deemed at excessive risk for surgical intervention; thus we determined that if diagnostic data warranted, the patient was to be considered for possible bilateral branch PA Melody valve implantation.

Figure 1. A, A 3D volume-rendered image in the axial plane that shows the dilated proximal right ventricular outflow tract and the complex proximal branch pulmonary artery stenosis. There is long-segment narrowing of the proximal RPA. The proximal LPA rises rightward and then takes a sharp bend 180° to the left. Beyond the stenotic proximal regions, the distal RPA and LPA appear well developed. B, The same volume-rendered image seen from an extreme left anterior oblique cranial view. The acute angulation of the LPA is well demonstrated. LPA indicates left pulmonary artery; RPA, right pulmonary artery.
Cardiac MRI Methods and Results
The cardiac MRI was performed on a Siemens 1.5-T scanner. There was severe pulmonary regurgitation with a pulmonary regurgitant fraction of 45%. The RV was moderately dilated (RV end-diastolic volume, 139 mL/m2) with diminished ejection (RV ejection fraction, 45%). The RVOT measured 3 cm in diameter proximally, narrowing significantly at the PA bifurcation. There was complex stenosis of both proximal branch PAs as shown in Figure 1A and 1B. Flow distribution favored the right PA (right PA, 68%; left PA, 32%). The LV size was normal, and the LV ejection fraction of 65%. The cardiac index measured 2.5 L/min per m2. On completion of the MRI scan, the patient was transferred directly to the catheterization laboratory.

Cardiac Catheterization Methods and Results
Hemodynamic results are summarized in the Table. The most significant findings were near-systemic RV pressure, severe biventricular diastolic dysfunction, and moderate to severe bilateral proximal branch PA stenosis. Baseline angiography confirmed the MRI findings of a dilated RVOT and complex proximal branch PA stenosis (Figure 2).

Interventions
Based on the findings from MRI and diagnostic catheterization, we deemed that the patient required relief of both PA obstruction and PA insufficiency and decided to presten each PA before Melody valve implantation. We carried out the interventions sequentially, starting with the right PA.

The right PA was stented with a Genesis 3910B stent (Cordis) on an 18 mm balloon-in-balloon catheter (NuMed Inc; Hopington, NY). The Melody valve then was mounted on a 20-mm Ensemble delivery system deployed in standard fashion. Next, we addressed the left PA, which was slightly more complicated because of its convoluted nature along its proximal course. The vessel was prestented with a 3910B Genesis stent on an 18 mm balloon-in-balloon catheter, and a Melody valve was placed on an 18-mm Ensemble system.

Follow-up angiography revealed unobstructed and competent Melody valves bilaterally (Figures 3 and 4). The postintervention hemodynamic data are shown in the Table.

**Table. Hemodynamic Findings Before and After Melody Valve Implantation Into the Bilateral Proximal Branch Pulmonary Arteries**

<table>
<thead>
<tr>
<th>Site</th>
<th>Before Melody Valves, mm Hg</th>
<th>After Melody Valves, mm Hg</th>
</tr>
</thead>
<tbody>
<tr>
<td>RA</td>
<td>22/23</td>
<td>22/20</td>
</tr>
<tr>
<td>RV</td>
<td>75/20</td>
<td>60/20</td>
</tr>
<tr>
<td>DAO</td>
<td>95/65</td>
<td>130/65</td>
</tr>
<tr>
<td>MPA</td>
<td>65/20</td>
<td>55/20</td>
</tr>
<tr>
<td>Distal RPA</td>
<td>45/15</td>
<td>50/25</td>
</tr>
<tr>
<td>Distal LPA</td>
<td>47/20</td>
<td>49/25</td>
</tr>
<tr>
<td>Mean RPCW and LPCW</td>
<td>23</td>
<td>22</td>
</tr>
<tr>
<td>LV</td>
<td>96/22</td>
<td>...</td>
</tr>
<tr>
<td>Cardiac index (MRI), L/min per m2</td>
<td>2.5</td>
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</tbody>
</table>

DAO indicates descending aorta; LPA, left pulmonary artery; LPCW, left pulmonary capillary wedge pressure; LV, left ventricle; MPA, main pulmonary artery; RA, right atrium; RPA, right pulmonary artery; RPCW, right pulmonary capillary wedge pressure; RV, right ventricle.

Figure 2. A right pulmonary arteriogram seen from the right anterior oblique cranial view. There is long-segment stenosis of the proximal RPA. The distal RPA measured 20 mm in diameter. The RVOT measured 28 to 33 mm in various views and was therefore too large for Melody valve implantation into the orthotopic position. The origin and acute angulation of the stenotic LPA also is shown. RPA indicates right pulmonary artery; RVOT, right ventricular outflow tract; LPA, left pulmonary artery.

Discussion
In the past decade, percutaneous pulmonary valve implantation has become well-established therapy mostly for patients with dysfunctional RV-to-PA conduits. Unfortunately, patients with conduits represent only a small fraction of those with clinically significant postoperative RVOT dysfunction, with most patients having dilated and distorted RVOTs often with branch PA stenosis, as was the case in the present patient. Consequently, there is worldwide interest in developing minimally invasive strategies to address the needs of patients who currently are underserved by existing techniques and technologies. The present patient manifested symptoms of right-sided (and left-sided) heart failure with complex pulmonary stenosis and insufficiency. She was deemed a high-risk surgical candidate because of her significant comorbidities, and existing percutaneous devices were too small for implantation into the orthotopic position. Thus, an alternative approach was pursued.

Our recent preclinical experiments have demonstrated the feasibility and potential benefits of bilateral branch PA Melody valves, and this report describes our first successful
attempt in a human patient. The patient’s hemodynamic measures were significantly improved acutely by our intervention, and we are hopeful that this will translate into clinical improvement over time. Admittedly, there are many questions yet to be answered regarding the risks, benefits, costs, durability, and so forth of branch PA valve implantation in the long term, but based on this early experience, we remain optimistic that in a select group of patients, this technique will prove advantageous.

Disclosures
None.

References

KEY WORDS: stents heart defects congenital magnetic resonance imaging heart valve prosthesis implantation
Bilateral Branch Pulmonary Artery Melody Valve Implantation for Treatment of Complex Right Ventricular Outflow Tract Dysfunction in a High-Risk Patient
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Circ Cardiovasc Interv. 2011;4:e21-e23
doi: 10.1161/CIRCINTERVENTIONS.111.962373

Circulation: Cardiovascular Interventions is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 1941-7640. Online ISSN: 1941-7632

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