A Concert in the Heart. Bilateral Melody Valve Implantation in the Branch Pulmonary Arteries

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ABSTRACT: Patients undergoing right ventricular outflow tract reconstruction are subject to valve and cusp degeneration later in life, requiring further intervention to alleviate the pulmonary regurgitation that ensues. In these cases, percutaneous pulmonary valve implantation can be an effective alternative to surgery; however, since the indications have been limited to dysfunctional valved conduits, only a small number of patients have access to this treatment option. We present the case of an 18-year-old male with a patch-enlarged right ventricle to pulmonary artery conduit who underwent pulmonary valve implantation using two Melody transcatheter pulmonary valves (Medtronic, Inc) into the proximal right and left branch pulmonary arteries.

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Key words: percutaneous pulmonary valve implantation (PPVI), pulmonary regurgitation, adult congenital heart disease (ACHD)

Patients undergoing right ventricular outflow tract (RVOT) reconstruction are subject to valve and cusp degeneration later in life, requiring further intervention to alleviate the pulmonary regurgitation that ensues. In these cases, percutaneous pulmonary valve implantation (PPVI) can be an effective alternative to surgery; however, since the indications have been limited to dysfunctional valved conduits, only a small number of patients have access to this treatment option.1 With the increasing experience and evolution of this technique, experts have been evaluating new indications and novel treatment strategies that could make PPVI available to a larger patient population.2 Recently, Robb and Gillespie demonstrated, both in an animal model and in a human patient, that bilateral branch deployment of two Melody valves (Medtronic, Inc) would be an effective alternative in alleviating pulmonary regurgitation in the presence of large patch-repaired RVOTs where PPVI may not be possible.3,4 We present a similar experience with an 18-year-old male with a patch-enlarged right ventricle to pulmonary artery (RV-PA) conduit who underwent pulmonary valve implantation using two Melody transcatheter pulmonary valves (TPV) into the proximal right (RPA) and left branch pulmonary arteries (LPAs).

Clinical History
An 18-year-old male with Tetralogy of Fallot, pulmonary atresia, confluent central pulmonary arteries (PA), and major aortopulmonary collateral arteries (MAPCAs), status post palliation with an RV-PA conduit was referred to our catheterization laboratory for PPVI. In 1993, at 3 months of age, he underwent staged surgical correction with a 13 mm pulmonary homograft as a RV-PA conduit and bilateral pulmonary artery enlargement. Subsequently, in 1994, he underwent ventricular septal defect (VSD) closure, RVOT enlargement, and ligation of the right MAPCAs. In 2003, because of progressive obstruction of his RVOT, he underwent a third surgical operation with patch enlargement of the RV-PA conduit. In 2011, because of the patient’s exercise intolerance and the echocardiographic demonstration of massive dilatation of the right ventricle (RV) secondary to free pulmonary regurgitation, a cardiac magnetic resonance imaging (MRI) scan was performed. The MRI showed severe dilatation and dysfunction of the RV (RV end diastolic volume of 478 mL/m² and RV ejection fraction of 19%), severe RVOT dilatation (RVOT measured 28 x 33 mm) with severe regurgitation (regurgitant fraction of 65%). There was associated diffuse narrowing of the RPA (Figure 1). Left ventricular (LV) function was also noted to be impaired due to compression by the RV (LV end diastolic volume of 105
mL/m² and ejection fraction of 48%). A lung scan was also performed, showing a relative perfusion of 14% to the right lung. After official discussion with our cardiac surgeons, the patient was deemed at extremely high risk for surgical intervention and was subsequently scheduled for PPVI.

Cardiac Catheterization
Baseline hemodynamic data are summarized in Table 1. At the beginning of the procedure, the RVOT was balloon sized with a 30 x 40 mm PTS-X balloon (NuMed) and no discernable waist was demonstrated, deeming it impossible to perform PPVI in the conventional fashion with placement of any commercially available transcatheter valve in the RVOT. The RPA stenosis and hypoplasia was relieved by primary stenting using a 28 mm CP Stent (NuMED) on a 15 x 40 mm Cristal Balloon (Balt) followed by high-pressure dilation with a 16 x 40 mm ATLAS balloon (Bard). The first Melody valve was then deployed in the RPA stent in a standard fashion, employing an 18 mm Ensemble delivery system. The balloon sizing of the LPA with an 18 x 40 mm Cristal Balloon demonstrated the presence of a moderate waist only at the take-off of the left lower PA, so we decided to implant a 43 mm XXL Andra Stent (Andramed) over a 20 x 45 mm Cristal Balloon, anchoring the stent to the left lower PA. An Andra stent was chosen due to its hybrid cell design, which would allow stenting of the entire LPA without complete jailing of the left upper lobe PA. A Melody valve, mounted on a 22 mm delivery system (Medtronic, Inc) was eventually delivered into the proximal portion of the stented LPA, leaving the origin of the left upper PA patent. Final angiographic assessment confirmed the good position of the valves and the patency of the left upper PA with no regurgitation across the valves (Figures 2A and Figure 2B). A new subset of hemodynamic data was obtained and is summarized in Table 1.

Follow-up
At 3-month follow-up visit, a lung scan demonstrated symmetric perfusion of the lungs. An MRI study was also performed (Figure 3). There was dramatic improvement in the RV size, function, and regurgitant fraction (Table 2). Since implantation of the two valves, the patient experienced a dramatic improvement in his exercise tolerance and functional status (New York Heart Association [NYHA] class 3 pre-PPVI to NYHA class 2 at latest follow-up post-PPVI).

Discussion
Patients suffering from complex congenital heart disease are subject to numerous palliative surgeries and reinterventions with incremental difficulties and associated risks. PPVI provides a non-surgical alternative to treat dysfunctional RV-PA conduits thus possibly reducing the number of open-heart surgical reinterventions. Although current indications remain restrictive, recent studies and case reports have already demonstrated the feasibility of PPVI in native as well as patch-enlarged RVOTs in the presence of a suitable waist. In this case report, we have taken a novel approach, first described in 2011 by Robb and Gillespie, in order to treat a patient with a patch-enlarged RVOT in the absence of a waist. We postulated that implanting a valve in each pulmonary artery would produce the same outcome as the conventional single valve implanted in the main pulmonary artery. The procedure was carried out without complications and with excellent results, restoring normal pressure values. In a very short-term follow-up, the patient showed both a dramatic improvement in his exercise tolerance and clinical status as well as an improvement in his cardiac function evaluated by MRI. This case shows that bilateral branch PPVI is feasible, may provide a viable alternative for some patients at high risk for surgery, and is best suited to patients with branch PA stenosis. New techniques such as this could, in the near future, extend PPVI to patients formerly considered unsuitable, thereby allowing physicians to evaluate treatment strategies based on clinical conditions rather than lack of alternatives.

Conclusion
In selected patients, bilateral branch PPVI is feasible and may provide a viable alternative for patients at high risk for surgery and it is best suited to patients with branch PA stenosis.
References

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**Figure 1.** (A) A three-dimensional volume-rendered image in the frontal plane that shows the dilated proximal right ventricular outflow tract and the severe stenosis and hypoplasia of the right pulmonary artery (arrow). (B) The same volume-rendered image seen from behind. Asterisk shows the patch-enlarged right ventricle to pulmonary artery conduit; Double asterisk shows the left pulmonary artery.
Figure 2. (A) Right pulmonary artery (RPA) angiography after implantation of the RPA Melody valve. There is no insufficiency. (B) Left pulmonary artery (LPA) angiography after implantation of the LPA Melody valve. There is no insufficiency and the left upper pulmonary artery is patent.

Figure 3. (A) Magnetic resonance image (MRI) before implantation of the Melody valves. Four-chamber cine SSP showing a markedly dilated right ventricle (RV) with diastolic interventricular septum deviation to the left due to RV volume overload (arrow). (B) MRI after implantation of the Melody valves. Four-chamber cine SSP showing a reduction of the RV volume and a normalized diastolic interventricular septum position (arrow).
Table 1. Hemodynamic findings before and after two Melody valves were implanted into the bilateral proximal branch pulmonary arteries.

<table>
<thead>
<tr>
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<th>Preprocedure Valve Pressures (mm Hg)</th>
<th>Postprocedure Valve Pressures (mm Hg)</th>
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<tbody>
<tr>
<td>Right atrium</td>
<td>12 / 12 (mean, 11)</td>
<td>10 / 8 (mean, 8)</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>64 / 12 (mean, 15)</td>
<td>35 / 8 (mean, 28)</td>
</tr>
<tr>
<td>Right pulmonary artery</td>
<td>23 / 10 (mean, 25)</td>
<td>31 / 16 (mean, 28)</td>
</tr>
<tr>
<td>Left pulmonary artery</td>
<td>64 / 10 (mean, 25)</td>
<td>31 / 16 (mean, 28)</td>
</tr>
<tr>
<td>Descending aorta</td>
<td>82 / 52 (mean, 65)</td>
<td>130 / 65 (mean, 96)</td>
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Table 2. Comparison of the right ventricular and left ventricular volumes and function before and after three months since bilateral pulmonary valves were implanted.

<table>
<thead>
<tr>
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<th>MRI Before Dual Valve Implantation</th>
<th>MRI After Dual Valve Implantation</th>
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<tbody>
<tr>
<td></td>
<td>RV</td>
<td>LV</td>
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<tr>
<td>EDV</td>
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<tr>
<td>ESV</td>
<td></td>
<td></td>
</tr>
<tr>
<td>EF</td>
<td>19%</td>
<td>48%</td>
</tr>
<tr>
<td>RF</td>
<td>—</td>
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RV = right ventricle; LV = left ventricle; MPA = main pulmonary artery; LPA = left pulmonary artery; RPA = right pulmonary artery; EDV = end-diastolic volume; ESV = end-systolic volume; EF = ejection fraction.