Bilateral Percutaneous Pulmonary Valves for Severe Pulmonary Regurgitation in a Patient with Prior Valvotomy

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INTRODUCTION

Pulmonary regurgitation (PR) is a known long-term complication after surgical repair of congenital pulmonary stenosis (PS). It is most commonly seen two to three decades after correction of congenital tetralogy of Fallot.1 According to the 2014 American Heart Association/American College of Cardiology valvular heart disease guidelines, interventions for PR should be considered only if there are signs or symptoms of right ventricular (RV) dysfunction in the setting of severe PR.2 Although surgical pulmonary valve replacement remains the gold standard treatment for PR, transcatheter pulmonary valve implantation has evolved as a safe alternative to minimize the need for repeat surgical interventions in these patients. Available transcatheter valves, approved for implantation in the RV outflow tract (RVOT), have been shown to result in good short- and long-term outcomes.3 However, there are limited data regarding outcomes among patients in whom deployment of the valve in the outflow tract is contraindicated. Only a few case reports and small case series have been reported. In this case report, we present the unusual management of a patient with severe PR after valvotomy in whom a primary intervention was contraindicated because of severe RVOT dilation.

CASE PRESENTATION

A 32-year-old man with a medical history of congenital ventricular septal defect and PS status post patch repair and surgical valvotomy, respectively, was diagnosed with heart failure. Transthoracic echocardiography (TTE) showed a left ventricular ejection fraction of 20% and an RV end-diastolic volume of 379 mL (indexed value 223 mL/m²) and an RV end-systolic volume of 246 mL (indexed value 145 mL/m²) and an RV ejection fraction of 35% (Videos 2 and 3).

At 4-month follow-up, TTE showed worsening RV dysfunction (tricuspid annular plane systolic excursion of 1.0 cm; Figure 1), at which point it was decided to proceed with an intervention. Initial catheterization showed a severely dilated pulmonary valve annulus of 32.39 × 34.25 mm. The diameters of the right and left pulmonary arteries (PAs) were 15 and 14 mm, respectively. Given the size of the pulmonary valve annulus and the patient’s multiple comorbidities, he was deemed a high-risk surgical candidate, and as such a transcatheter approach with Melody valves (Medtronic, Minneapolis, MN) was planned. Bilateral stents 2 cm in length were placed initially in the main PAs, with final diameters of 19.6 mm on the left and 17.9 mm on the right. Valve deployment at the time was delayed, allowing stent epithelization for a more secure landing zone (Videos 4 and 5).

Five months after the stents were placed, hemodynamics were notable for ventricularization of PA pressures secondary to severe PR (PA diastolic pressure = RV end-diastolic pressure). There was no gradient from the right or left PA to the main PA, and there was no gradient across the pulmonary valve. At that time, 22-mm Melody valves were deployed, overlapping the prior PA stents. No regurgitation of the valves was noted on angiography (Video 6). RV end-diastolic pressure remained at about 6 to 8 mm Hg. Detailed pressures are outlined in Table 1. The patient tolerated the procedure without complications and was safely discharged home days later. Postoperative TTE with direct view of the RVOT demonstrated no PR on continuous-wave Doppler (Video 7, Figure 2).

Figure 1 TTE demonstrating tricuspid annular plane systolic excursion of 1.0 cm.

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Outpatient TTE 3 weeks after the procedure showed severely depressed RV function but no significant PR. The peak velocity of the proximal PA branches was normal at 1.5 m/sec (Video 8).

**DISCUSSION**

Severe PR is uncommon and is mostly seen after surgery for tetralogy of Fallot or congenital lesions such as PS, as in the case of our patient. In a longitudinal study of patients who underwent surgical repair of PS before the age of 15 years, up to 21% had severe PR at a follow-up period of 33 years. In that study, 15% required reintervention and 9% underwent homograft implantation because of late PR. Interestingly, Nielsen et al. showed that freedom from reintervention decreases drastically with each decade after surgical repair of PS. Freedom from reintervention was 91% at 20 years post-procedure but decreased drastically to 26% after 50 years. This highlights the need for long-term follow-up of these patients by a cardiovascular team.

One of several challenges in the management of our patient was his late referral. By the time he was evaluated in the clinic there was already evidence of severe RV dysfunction. As mentioned before, current guidelines recommend intervention in cases of signs or symptoms of severe RV dysfunction. In the case of our patient, his intellectual disability was a barrier for communicating specific symptoms. Cardiac magnetic resonance imaging was essential for further evaluation, aiding in assessing the degree of RV dilation. At the time cardiac magnetic resonance imaging was done, the patient already exceeded the recommended thresholds for surgical intervention: RV EDV index > 150 mL/m² or RV end-systolic volume index > 80 mL/m². It is important to point out that severe PR, regardless of symptoms, also contributes to an increased incidence of arrhythmias and an increased risk for sudden death.

Percutaneous pulmonary valve implantation was preferred because of high surgical risk, as previously mentioned. This approach was first introduced in the early 2000s, and since then, improvements in devices and techniques have been made. Currently there are two available valves: the Melody valve and the SAPIEN XT pulmonary valve (Edwards Lifesciences, Irvine, CA). The Melody valve can be expanded only up to 24 mm, and the largest available diameter of the SAPIEN XT valve is 29 mm, thus implantation in larger RVOTs is not feasible.

In our patient, the RVOT was severely dilated to 34 mm in diameter, so bilateral implantation of Melody valves in the main PA branches was the only practical option.

The patient underwent bilateral implantation of Melody valves without any complications. However, it is important to point out potential complications of this approach. Stent fracture resulting from dynamic recoil of the RVOT has been reported to occur in up to 35% of implanted patients at a median follow-up of 5 years. Coronary compression, which should be assessed during the procedure, is seen in approximately 5% of patients. Last, pulmonary valve–related endocarditis can also occur. A meta-analysis by Chatterjee et al. showed a pooled incidence of 0.6 per 100 person-years.

Outpatient TTE 3 weeks after the procedure showed severely depressed RV function but no significant PR. The peak velocity of the proximal PA branches was normal at 1.5 m/sec (Video 8).
important, 50% of patients had improvement in RV size after implantation at 2-year follow-up. This highlights the need for long-term follow-up of patients undergoing percutaneous pulmonary valve implantation.

CONCLUSION

In this case we present the feasibility and good short-term outcomes of bilateral pulmonary valves implanted in the main PA branches. This was the only option in a patient with history of valvotomy with higher than standard surgical risk and whose anatomy was not suitable for implantation in the RVOT. Further studies are needed to evaluate long-term outcomes of patients with bilateral transcatheter implantation of Melody valves.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2020.10.005.

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