



## JSCAI Case Report

# Transcatheter Treatment for Double-Chambered Right Ventricle in Patient Presenting Severe Chronic Heart Failure



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## ABSTRACT

We report a case involving a 9-year-old girl with severe biventricular dysfunction due to anomalous right ventricle muscle bands. After a thorough discussion, she underwent a transcatheter stenting procedure for the double-chambered right ventricle. During her follow-up, she regained biventricular function, while remaining asymptomatic, resulting in complete recovery.

First described in the 19th century, obstruction of the right ventricle (RV) by an anomalous muscle bundle is a rare congenital heart disease (CHD), usually associated with a ventricular septal defect (VSD). Depending on the degree of obstruction, the clinical course varies significantly, and the current treatment of choice is surgical resection.<sup>1,2</sup>

## Case

A 9-year-old girl presented with a subaortic VSD and anomalous RV muscle bands. She was first seen as an outpatient at 5 years old, 4 years after a corrective VSD surgery. She presented exertional shortness of breath and cyanosis triggered by crying, with an echocardiogram showing a 97 mm Hg gradient due to RV muscle bands. Resection was indicated, but the patient missed her appointments for the next 3 years. At the age of 5 years old, she already presented severe RV systolic dysfunction and severe tricuspid regurgitation (TR).

At the age of 9 years, she was referred from another quaternary facility for a heart transplant evaluation, which was prompted by concerns about her symptomatology and findings on echocardiogram. Despite a prior course of levosimendan, she presented with severe functional impairment of the RV and diastolic dysfunction of the left ventricle (LV) (Figure 1). Surgical repair was felt to be high risk and was not considered an option. Consequently, a heart transplant evaluation was the most appropriate course of action. Subsequent echos revealed a greater gradient of 116 mm Hg in the RV, while cardiac MRI confirmed severe dysfunction of the RV (right ventricular

ejection fraction [RVEF] 14%) and moderate impairment of the LV (left ventricular ejection fraction [LVEF] 37%).

After discussion with the heart team, transcatheter stenting of the RV seemed like a reasonable palliative option in order to alleviate RV pressure, thereby improving its function and potentially recovering LV function due to the concept of interventricular interdependence.

The procedure was performed in a hybrid room with the surgical team on standby via the right femoral vein. RV angiogram revealed a double-chambered RV with severe contractile dysfunction and intraventricular pressure at 2/3 of systemic levels. Prior to the intervention, a 32-mm Hg gradient was observed across the RV outflow tract, with good mobility of the pulmonary valve. Predilation was performed using a 12-mm × 40-mm balloon (POWERFLEX Pro, Cordis) over an Amplatz super stiff wire placed across the RV outflow tract. A 12F sheath was advanced to the RV, followed by the deployment of a Palmaz P4014 stent (Cordis) using a 20-mm × 40-mm balloon (MAXI LD, Cordis), guided by both echocardiography and fluoroscopy. The stent was implanted successfully across the obstructive muscle bundle. Follow-up angiography demonstrated improved anterograde flow. RV pressure reduced to 40/10 mm Hg and the interventricular gradient decreased from 32 to 7 mm Hg. No signs of dissections were observed, and the pulmonary valve remained intact with trivial regurgitation. The echocardiogram showed improvement in TR from severe to mild, and the interventricular gradient was reduced to 6 mm Hg. The patient was transferred to the cardiac intensive care unit, where she was extubated immediately. Vasoactive drugs were weaned off and discontinued

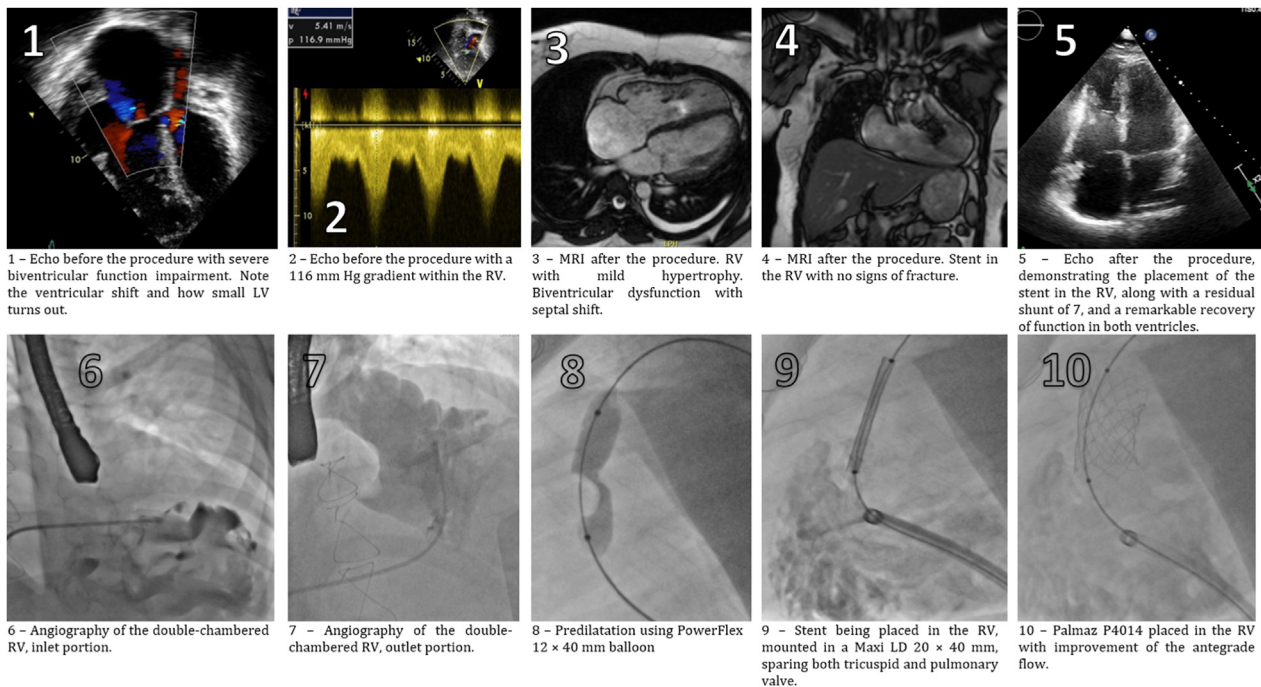
**Keywords:** biventricular dysfunction; double-chambered right ventricle; heart failure; heart transplant; right ventricular obstruction; stenting in the right ventricle.

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**Figure 1.**

**Preprocedural, periprocedural, and postprocedural imaging.** LV, left ventricle; MRI, magnetic resonance imaging; RV, right ventricle.

after 14 days. An echocardiogram before discharge revealed mild TR, improved biventricular function with an LVEF of 58%, and a residual RV intraventricular gradient of 12 mm Hg.

During her follow-up in the clinic, 7 years after the procedure, she remained asymptomatic, actively participating in exercise. Her latest echocardiogram and MRI revealed no TR, preserved biventricular function (LVEF 58%), and a gradient of 17 mm Hg in the RV. She has been regularly monitored for arrhythmias with electrocardiogram, with no changes noted in this regard.

## Discussion

A recurring concept in CHD is ventricular interdependence, where one ventricle impacts the other's function. In cases of significant RV outflow obstruction, a septal shift might occur, leading to a subsequent decrease in LV diastolic compliance.<sup>3</sup> The natural history of our patient illustrates this concept effectively. As a result of RV pressure overload, LV compliance initially decreases while preserving its ejection fraction. However, over time, this leads to LV atrophic remodeling and altered systolic function.<sup>3</sup>

This case involves a 9-year-old girl with severe biventricular dysfunction and functional limitations caused by a mechanical lesion obstructing her RV. Initially, stenting the RV was considered a palliative measure due to the impairment of her ventricular function and clinical condition, with the intention of serving as a bridge to surgery. The procedure proved highly successful, leading to the recovery of biventricular function, and she has remained asymptomatic for the past 7 years.

She will continue her follow-up with regular echocardiograms. If, at any time, there is a significant increase in gradients, any ventricular function impairment, or signs of stent fracture, she should undergo a new catheterization for evaluation.

## Conclusion

The presented case underscores the importance of innovative and timely interventions in the management of complex CHD. This patient's remarkable 7-year asymptomatic course, with preserved biventricular function, exemplifies the sustained benefits of such interventions, providing a bridge to sustained recovery beyond short-term palliative relief. However, long-term follow-up and vigilance are essential in managing these patients.

## Declaration of competing interest

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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## Ethics statement and patient consent

The research adhered to ethical guidelines, and patient consent was obtained as necessary.

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