Cardiology in the Young

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Melody valve insertion for relief of "Tricuspid" stenosis in an unbalanced atrioventricular septal defect

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Brief Report

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Abstract

A Melody valve was successfully placed across a very stenotic right-sided component of a common atrioventricular valve because of ongoing troublesome arrhythmias in a young woman with an unbalanced atrioventricular septal defect, a very dilated right atrium and a hypoplastic right ventricle. Four years later, she remains well.

The management of a stenotic atrioventricular congenital heart abnormality remains problematic, more so if part of a congenital heart abnormality is previously subjected to surgery.

Case report

"A" now aged 33, with a non-trisomic unbalanced atrioventricular septal defect, had a total repair at the age of 29 months. At the time, the "Anderson's cleft" was sutured to reduce the moderate left-sided atrioventricular valve regurgitation. The large atrial defect and a smaller ventricular component of the atrioventricular septal defect were closed by a two-patch technique. A small right ventricle was enlarged by extensive resection of trabeculations following which the right-sided atrioventricular valve measured 19 mm, only 1 mm smaller than that expected at her age thereby allowing a two pump repair. A single stitch was placed between the edge of the superior common leaflet and the Dacron patch to reduce the mild-to-moderate right-sided atrioventricular valve regurgitation. Off bypass, the mean right atrial pressure was 10 mmHg when compared to 9 mmHg mean left atrial pressure. Despite that she developed increasing systemic venous congestion. At catheteriation during the age of 5, her right atrial pressure was 28 mmHg. Further surgery was undertaken. Her superior caval vein was connected to her right pulmonary artery. In addition, a 1 mm valvotomy on either side of the single commissure of her right-sided atrioventricular valve enlarged it to 13 mm with a gradient of 14 mmHg with dysplastic regurgitation. The right ventricle remained small and heavily trabeculated and the right atrium very dilated. The superior caval vein pressure dropped to 13–14 mmHg post-bypass when compared to 6–8 mmHg in the left atrium.

Considerable clinical improvement was subsequently observed, but with ongoing evidence of right atrioventricular valve stenosis (Fig 1). At the age of 24, she started having atrial arrhythmias – atrial flutter/atrial tachycardia/fibrillation. Each episode required a transoesophageal echocardiogram followed by direct current cardioversion, maintenance of anti-arrhythmic therapy, and ongoing anticoagulation. Three electrophysiology studies were undertaken with multiple flutter pathways ablated. Throughout, her right atrium remained very dilated with a mean Doppler gradient of 12–14 mmHg across the atrioventricular valve.

Further surgery was not considered as a good option, neither was balloon dilating the valve that was previously tried with rheumatic tricuspid stenosis.[1] The decision was made to endovascularly rehabilitate the right atrioventricular valve connection. At the age of 29, she underwent an uncomplicated procedure with balloon interrogation of that valve, ensuring that the right coronary artery was not compressed and with pre-stenting the stenotic connection. A Melody valve was deployed to 20 mm (Fig 2). That halved the mean Doppler gradient across the valve to 6–7 mmHg. Two further episodes of atrial flutter occurred 3 and 5 months later despite anti-arrhythmic medication. She had further dilation of the Melody valve to 22 mm 8 months after its insertion, with a reduction in the gradient down to 5 mmHg but with the development of mild incompetence, which gradually improved with time.

Four years later, she has remained well and fully employed. She has a mean Doppler gradient of 6 mmHg across the valve during inspiration dropping to 4 mmHg during expiration with minimal evidence on colour Doppler of incompetence. The "lumen" of the valve measured 13 mm as estimated on cross-sectional imaging. Her liver subjected to prolonged venous congestion has improved considerably when last checked a year ago with a fall in the liver stiffness

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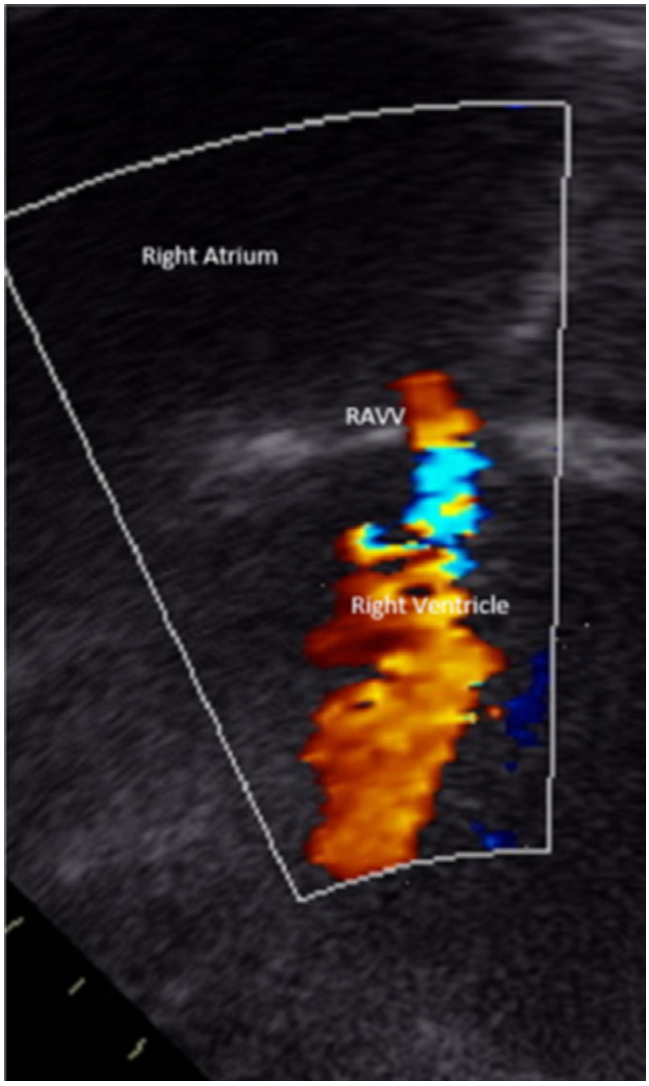

Figure 1. Stenotic right-sided atrioventricular valve. RAVV = right-sided atrioventricular valve.

from a high of 30 KPA to 7.2KPA by FibroScan. Her liver is described as smooth on ultrasound, not suggestive of cirrhosis. There has been a halving of her slightly elevated liver enzymes. With the onset of symptomatic bradycardia, she recently had a dual-chamber pacemaker inserted with epicardial leads for atrial pacing. As the right atrium remains enlarged (see supplementary Figure 2), it was thought prudent to maintain her on Apixaban.

Discussion

The management of tricuspid stenosis remains problematic. Despite her earlier surgery followed subsequently by a surgical valvotomy of the right-sided component of the common atrioventricular valve and the creation of a superior cavopulmonary connection, the right atrium remained dilated. The right ventricle failed to grow with the persistence of a substantial and increasing gradient across the valve. The development of recurrent arrhythmias as a young adult with the need for ongoing medication, repeated cardioversions, and ablation procedures raised the need for further intervention. Surgery was considered, but thought inappropriate as well as balloon dilatation of the valve orifice. Previous

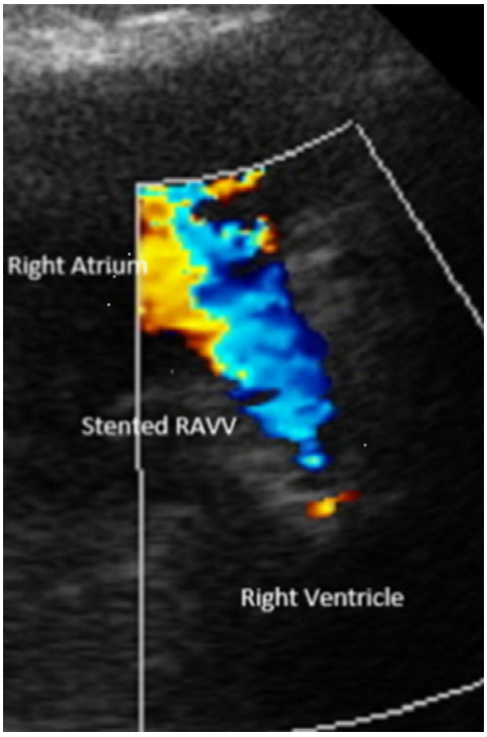

Figure 2. Colour flow through the Melody valve at the right atrioventricular valve site. RAVV = right-sided atrioventricular valve.

experience with the insertion of a Melody valve[2,3] at the tricuspid site raised the possibility as to whether that might be an option in this patient. However, most of that experience arose from dilatation and/or insertion of the Melody valve in previously surgically inserted bioprosthetic valves, while multiple intervention procedures have evolved for the treatment of significant tricuspid incompetence.[4] Nitta et al[5] reported ballooning of a bioprosthetic valve, which is placed at the right atrioventricular valve site for severe right-sided regurgitation following earlier repair of an atrioventricular septal defect with a large primum atrial component. Previous reports have included patients with CHD, but without clarification of the underlying anomaly (except for Ebstein anomaly), or of the previous surgery undertaken.[6] Our report describes a novel approach with medium-term relief of severe right-sided atrioventricular stenosis in an unbalanced atrioventricular septal defect when further surgery and/or balloon valvuloplasty were not considered as viable options.

Conclusion

This report suggests percutaneous rehabilitation of the stenotic right atrioventricular connection with a Melody or other percutaneous valve may be a reasonable option to consider in patients with persistent or troublesome stenosis. Further experience and improvement in the devices available may broaden its applications.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951121002419.

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Conflict of interest. None.

Ethical standard. Not applicable.

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