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Brief Report

Treatment of severe refractory valvar pulmonary stenosis with primary transcatheter pulmonary valve implantation

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Abstract Some patients with pulmonary valve stenosis do not respond to balloon valvuloplasty and must undergo surgical repair. We report the case of a 12-year-old child with pulmonary valve stenosis and Noonan syndrome in whom we performed transcatheter Melody pulmonary valve implantation after balloon dilation failed. The result was excellent. This technique can be proposed as an alternative to surgery in such cases.

Keywords: Pulmonary valve disease; percutaneous intervention; percutaneous valve therapy; CHD; paediatrics; valvar pulmonary stenosis

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Transcatheter pulmonary valve implantation is a widely accepted treatment for right ventricular outflow tract conduit dysfunction. Short- and medium-term results are well known, and long-term outcomes are promising.[1]

More recently, transcatheter pulmonary valve implantation has been successfully applied in dysfunctional post-surgical native right ventricular outflows that are mainly regurgitant.[2] Transcatheter pulmonary valve implantation in a native unoperated pulmonary valve stenosis, however, has not been described. Balloon pulmonary valvuloplasty is usually successful in these patients;[3] nevertheless, in some cases with particularly dysplastic valve leaflets, transcatheter pulmonary valvuloplasty may fail. Surgical therapy is then performed with valvulotomy or a transannular patch in very dysplastic valves, such as in patients with Noonan syndrome.[4] Direct pulmonary valve replacement with a conduit may be considered as an alternative in clinical practice.[5] We report the case of a child with isolated severe pulmonary valve stenosis and a history of two previous balloon valvuloplasties who underwent transcatheter pulmonary valve implantation as first-line primary treatment without preceding surgical intervention.

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Case report

A 12-year-old girl (weight 40 kg, height 145 cm) with Noonan syndrome presented with severe recurrent pulmonary valve stenosis after a 3-year lapse in follow-up. She was asymptomatic. Her past medical history was significant for balloon pulmonary valvuloplasty at 2 and 7 months of age. A residual peak-to-peak systolic pulmonary valve pressure gradient of 45 mmHg was noted following the second procedure. She had been stable during follow-up until March 2010 with a mean pulmonary valve gradient of 35 mmHg by continuous Doppler flow. Her physical examination showed typical Noonan morphology. Cardiac examination revealed a long, harsh ejection systolic murmur. A 12-lead electrocardiogram showed right ventricular hypertrophy. Severe stenosis of the pulmonary valve was confirmed by continuous Doppler flow, with a peak and mean gradient of 102 and 65 mmHg, respectively. Her estimated right ventricular systolic pressure was 95 mmHg (cuff blood pressure 120/70 mmHg). The pulmonary valve was dysplastic, thickened, and doming with a 16-mm annulus diameter. An additional non-obstructive left ventricular hypertrophic

cardiomyopathy was also documented. Biventricular function was normal. Cardiac MRI showed normal right ventricular volume (56 ml/m^2) and ejection fraction (72%), without pulmonary regurgitation. Surgical repair was not expected to preserve pulmonary valve function. Therefore, the consensus was in favour of transcatheter treatment with pulmonary valve implantation if the patient failed to respond adequately to balloon valvuloplasty.

Cardiac catheterisation was performed under general anaesthesia following informed consent. Haemodynamic data confirmed severe pulmonary valve stenosis. Systemic right ventricular hypertension was confirmed (systolic right ventricular pressure 95 mmHg, aorta 97 mmHg). A right ventricular angiogram revealed a thick, doming pulmonary valve (Fig 1, Supplementary movie 1). Balloon pulmonary valvuloplasty was performed with a 20-mm × 4-cm Z-Med balloon (NuMED Inc., Hopkinton, New York, United States of America) without an adequate response. An aortic root angiogram during right ventricular outflow tract balloon inflation ruled out coronary compression. Pre-stenting was performed to prepare a landing zone with deployment of CP8Z34 and CP8Z39 stents (NuMED Inc., Hopkinton, New York, United States of America) in the right ventricular outflow tract over a 20-mm BIB balloon (NuMED Inc., Hopkinton, New York, United States of America). A 22-mm Melody valve (Medtronic, Minneapolis, Minnesota, United States of America) was implanted within the landing zone without complication. Post-implantation cardiac catheterisation angiogram in the main pulmonary artery revealed no residual obstruction and absence of significant pulmonary regurgitation (Fig 2, Supplementary movie 2). Post-implantation systolic right ventricular pressure was normal at 30 mmHg. There were no procedural complications, and the patient was discharged home the following day. A follow-up cardiac MRI performed 7 months later confirmed an unobstructed right ventricular outflow tract with a well-functioning Melody valve with no regurgitation.

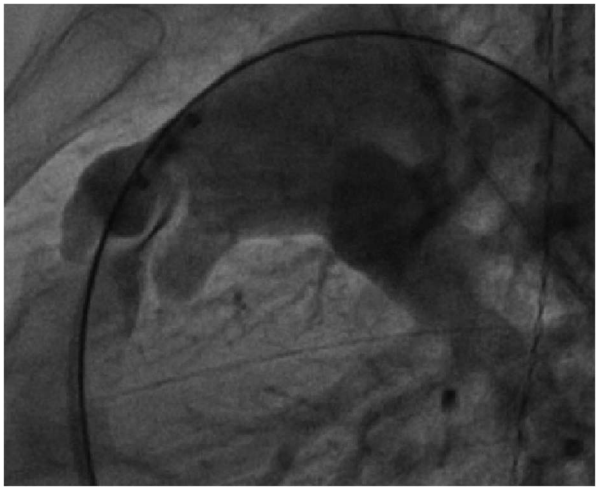

Figure 1.
Frontal right ventricular outflow tract angiogram showing a thickened and dysplastic pulmonary valve. There is mild-to-moderate regurgitation that is mainly functional due to the position of the 5-French Multitrack catheter through the pulmonary valve during the contrast injection.

Discussion

Surgical repair for pulmonary valve stenosis is known to be followed by pulmonary regurgitation. In a recent study, moderate-to-severe pulmonary regurgitation occurred in as many as 45% during long-term follow-up. Up to 20% of patients required at least one re-intervention, which was pulmonary valve replacement for severe regurgitation 67% of times.[6] Patients with Noonan syndrome typically have dysplastic pulmonary valves and are known to be poor responders to balloon pulmonary valvuloplasty.[5] These patients are therefore more likely to develop chronic right ventricular pressure and/or volume overload after repeated balloon valvuloplasties and/or surgical repair of the pulmonary valve. Moreover, the coexistence of hypertrophic cardiomyopathy in Noonan syndrome is an additional risk factor for post-operative cardiac failure;[7] hence, application of an alternate non-surgical technique is an attractive consideration. Transcatheter pulmonary valve implantation can be considered in these patients as it provides relief of right ventricular outflow tract obstruction without the incumbent risk of pulmonary regurgitation.

There are no specific reports describing the application of transcatheter pulmonary valve implantation

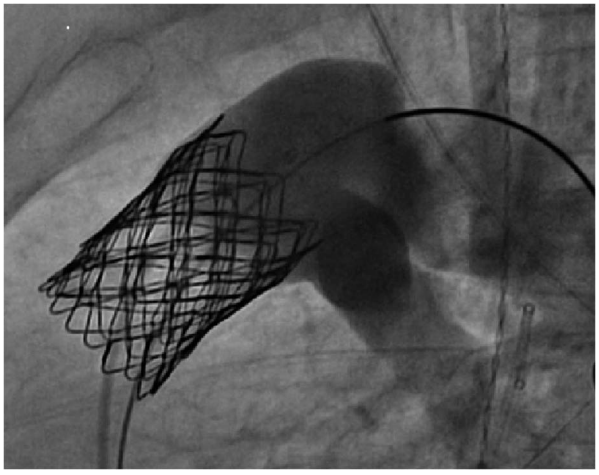

Figure 2.
Frontal right ventricular outflow tract angiogram after implantation of a 20-mm Melody valve showing no residual obstruction and no regurgitation.

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in patients with native pulmonary valve stenosis. Meadows et al presented a retrospective review of Melody valve implantation in non-conduit right ventricular outflow. Of the 31 patients in the study, three patients had isolated pulmonary valve stenosis; however, it is not clearly stated whether the patients were primarily treated at the time of Melody valve implantation without preceding history of surgical intervention.[2] The few other studies that reported transcatheter pulmonary valve implantation in native right ventricular outflow included patients who were previously operated.[8,9] Therefore, our case report is the first to fully describe transcatheter pulmonary valve implantation in a patient with isolated pulmonary valve stenosis without a preceding history of surgical treatment.

In conclusion, transcatheter pulmonary valve implantation was offered directly to a 12-year-old patient with Noonan syndrome for recurrent severe pulmonary valve stenosis after the patient failed to respond to conventional balloon pulmonary valvuloplasty. Resolution of right ventricle outflow tract obstruction was obtained without the risk of pulmonary regurgitation with a satisfactory result at follow-up. Such an application of this technique should be further studied in a prospective manner with a larger, multicentre cohort.

Acknowledgement

None

Conflicts of Interest

Alain Fraisse acts as a proctor for Medtronic (Melody valve implantation).

Supplementary material

To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951117000191

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