

Melody valve-in-valve implantation in the tricuspid position through a Fontan conduit fenestration

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ABSTRACT

Patients with single right ventricle and tricuspid regurgitation have increased morbidity and mortality rates. The outcomes of surgical tricuspid valve repair and replacement are dismal. Although some centers prefer valve replacement with a bioprosthetic valve in the tricuspid position for the pediatric population, these valves have limited longevity and will eventually need to be replaced. Unfortunately, single ventricle patients are fragile and unlikely to tolerate a second replacement. Percutaneous valve-in-valve (ViV) implantation is an acceptable alternative with low morbidity and mortality. Studies have also shown acceptable short- and mid-term outcomes. We report a case of Melody ViV placement in the tricuspid position in a patient with hypoplastic left heart syndrome through the fenestration of a Fontan conduit.

Keywords: Hypoplastic left heart syndrome, single ventricle, transcatheter valve replacement, valve-in-valve

INTRODUCTION

In patients with a functional single right ventricle, tricuspid valve regurgitation is a well-described risk factor for morbidity and mortality. However, in this population, surgical repair and replacement have very poor outcomes, and in those who survive, reoperation carries a significant risk.^[1-3] Since the advent of transcatheter valve replacements, the Valve-in-Valve International Database (VIVID) Registry has demonstrated the feasibility, safety and positive outcomes of transcatheter valve-in-valve (ViV) placement in the tricuspid position.^[4]

Transcatheter ViV in the atrioventricular valve position after the Fontan operation has only been reported using a hybrid approach.^[5,6] We describe percutaneous transcatheter ViV in the tricuspid position through a Fontan fenestration in a patient with hypoplastic left heart syndrome (HLHS). To the best of our knowledge, this has never been reported.

CASE REPORT

The patient is an 11-years-old, 28 kg, male born with HLHS (mitral stenosis and aortic atresia) and mild tricuspid regurgitation. At 1 week of life, he had a Norwood procedure, atrial septectomy, and a 3.5 mm right-sided modified Blalock-Thomas-Taussig shunt followed by bidirectional cavopulmonary anastomosis when he was 3-month-old. Over time, his tricuspid valve regurgitation progressed, and at 4 years of age, tricuspid valvuloplasty was performed at the time of the extracardiac fenestrated Fontan, with suboptimal results. Three years later, he underwent tricuspid valve replacement with a 25-mm Mosaic valve (Medtronic, Inc., Minneapolis, MN, USA). Over the next 4 years, the inflow gradient across the bioprosthetic valve progressively worsened to a mean of 10 mmHg, leading to protein losing enteropathy and recurrent atrial flutter which

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required multiple ablations. Given the risk of another surgical intervention, it was decided to attempt a transcatheter valve replacement.

At the time of the intervention, this patient was in sinus rhythm. Given the more inferior location of the fenestration, access in the femoral veins and artery was obtained. His baseline hemodynamics showed a low systemic saturation of 74%, as well as low cardiac index of 2 L/min/m² and Qp/Qs of 0.6. A simultaneous pressure measurement in the Fontan conduit, right atrium and ventricle showed severely elevated Fontan pressures at 23 mmHg with a mean right atrial pressure of 19 mmHg and bioprosthetic tricuspid valve inflow gradient of 13 mmHg which correlated with a mean gradient by transesophageal echocardiography (TEE) of 11 mmHg [Figure 1]. A 0.035" Amplatzer Super Stiff wire (Boston Scientific, Washington, DC, USA) was placed prograde across the fenestration, tricuspid valve and neo-aortic valve, and anchored in the descending aorta via a 7 Fr balloon wedge catheter. Balloon sizing of the tricuspid valve revealed a waist diameter of approximately 15 mm. A Melody valve mounted on a 22 mm Ensemble delivery system (Medtronic, Inc. Minneapolis, MN, USA) was easily advanced over the wire across the 4 mm fenestration into the tricuspid valve to the level of the radio-opaque markers of the Mosaic valve spanning the ring, and once in position, the Melody valve was deployed. Postdilation was performed with a 22 mm × 4 cm Atlas Gold balloon (Bard Peripheral Vascular, Inc. Tempe, AZ, USA) to a final valve diameter

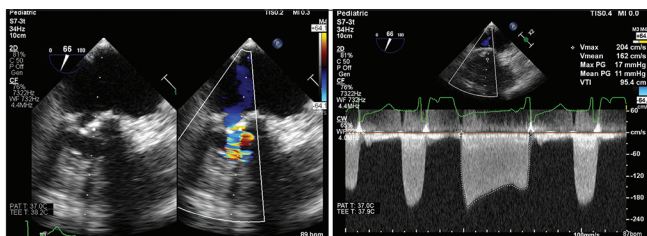


Figure 1: Preimplantation transesophageal echocardiography shows a tricuspid bioprosthetic valve mean inflow gradient of 11 mmHg

of 19 mm [Figure 2]. A TEE showed excellent valve placement with mild tricuspid regurgitation and an inflow gradient of 2 mmHg [Figure 3].

Following implantation of the valve, the cardiac index increased significantly to 3.1 L/min/m² without changes in the systemic saturation. Impressively, the mean right atrial and Fontan pressures decreased to 13 and 15 mmHg, respectively. There was a 1 mmHg inflow gradient across the Melody valve.

His postcatheterization course was remarkable for a low-grade fever of 101°F within the first 24 h with no changes in inflammatory markers. The fever resolved spontaneously. He was started on warfarin and aspirin and discharged 48 h after. The gradient on day 1 after implantation was 6 mmHg and remains unchanged on his most recent follow-up 11 months after the procedure. Remarkably, his symptoms have improved significantly with no clinical signs of PLE, or further episodes of atrial flutter.

DISCUSSION

Surgical techniques and long-term management in single-ventricle patients have improved significantly in the past 10 years, leading to an excellent 30-year survival rate.^[7] However, atrioventricular valve regurgitation is a major risk factor for mortality in this population,^[8,9] and regardless of repair techniques, the outcomes remain poor with a reported mortality close to 34%.^[1] On this basis, some have advocated for a “low threshold to replace the valve,”^[3] yet data on valve replacement in single ventricle patients are limited. In a study in Japan involving 82 centers, the mortality following valve replacement in patients with a single ventricle was 34%, and of those who survived, 20% required a second valve intervention.^[2] Most of these valves were used as a last resource when the clinical status of the patients had deteriorated significantly.

The VIVID Registry has shown the feasibility and excellent acute and mid-term outcomes of transcatheter



Figure 2: (a) Balloon sizing of the bioprosthetic valve with a 25 mm × 3 cm sizing balloon, demonstrating a minimum balloon diameter of 14.6 mm. The guide wire is advanced through the Fontan fenestration, across the bioprosthetic tricuspid valve and neo-aortic valve, and placed in the descending aorta. (b) The Melody valve mounted on a 22 mm Ensemble delivery system implanted in the tricuspid position. A right ventriculogram after Melody valve implantation and postdilatation with a 22 mm Atlas Gold balloon (c) shows that the Melody valve is proximal to the bioprosthetic valve ring and extends slightly beyond the 22 mm radioopaque markers with mild tricuspid regurgitation

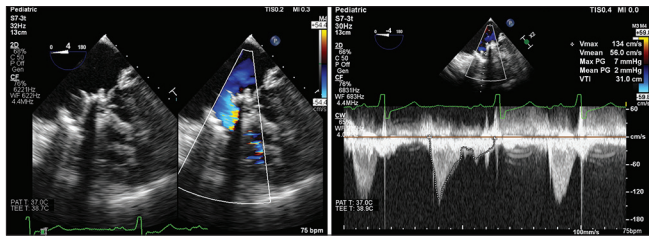


Figure 3: Postimplantation transesophageal echocardiography shows a low mean inflow gradient of 2 mmHg across the Melody valve

ViV implantation in the tricuspid position with minimal procedural complications and only 10% re-intervention rate. In addition, mortality was low and was mostly related to older age and patients who were critically ill at the time of the intervention.^[4] Patients with a single ventricle are subject to multiple surgeries and unlikely to tolerate a second valve replacement.

Our patient had multiple sternotomies and he had been admitted several times for a failing Fontan physiology making the surgical approach for valve replacement “too risky.” Therefore, it was decided for the least invasive approach with a percutaneous implantation taking advantage of the fenestration. Surprisingly, advancing the Ensemble delivery system across the 4 mm fenestration into the bioprosthetic valve was relatively easy, and no predilation was required.

Despite antiplatelet therapy with or without anticoagulant, the risk of thrombosis is small but not negligible.^[4,10] In patients with two functional ventricles, a small thrombus in the tricuspid valve may result in pulmonary microembolism and is generally well tolerated. In single-ventricle patients, thrombosis can lead to devastating outcomes. In addition, thrombus serves as a nidus for endocarditis, and as in other valve replacements, endocarditis remains an ongoing concern. Our patient was previously on warfarin, but due to a mild complication (bruising), it was discontinued. Interestingly, after discontinuation of anticoagulation, the gradient across the Mosaic valve increased significantly over the following year. Therefore, he was started on both warfarin and aspirin after the Melody valve was placed; thus far, he has had no reported complications.

CONCLUSION

Transcatheter ViV placement in the tricuspid position is feasible in patients with extracardiac Fontan through a fenestration and should be considered as an early alternative in symptomatic patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the

patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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