Case Report

Severely Regurgitant Left Ventricle to Ascending Aorta Conduit in a Failing Fontan Patient Treated With a Vascular Endograft and Melody Transcatheter Pulmonary Valve via Hybrid Approach

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A 28-year-old male with single ventricular heart disease status post Fontan palliation and subsequent placement of left ventricle to ascending aorta (LV-AAo) valved conduit developed ascites and edema. Diagnostic catheterization revealed elevated ventricular end diastolic pressures (EDP) secondary to severe LV-AAo conduit regurgitation. Given the unique anatomy, surgical access via the right axillary artery provided optimal route for transcatheter valve implantation within the conduit. The procedure resulted in significant hemodynamic improvement with no complications. © 2016 Wiley Periodicals, Inc.

INTRODUCTION

Single ventricle patients with declining or failing Fontan circulations often have limited interventional options for improving overall hemodynamics and functional status [1,2]. The off-label use of FDA-approved transcatheter valves provides a relatively new therapeutic option for patients with palliated complex congenital heart disease [3,4]. We describe the innovative use of transcatheter valve implantation (TVI) in a failing Fontan patient using a hybrid surgical vascular access approach.

CASE HISTORY

A 28-year-old male with d-transposition of the great arteries, hypoplastic right ventricle (RV), ventricular septal defect (VSD), and straddling tricuspid valve underwent pulmonary artery (PA) banding at one year of age followed by a classic Fontan with pulmonary artery ligation at age 2. Four years following his Fontan operation, the patient underwent placement of a 20 mm valved homograft conduit from the left ventricle to ascending aorta (LV-AAo) due to progressive severe restriction of the VSD and outflow to the aorta. At 22 years of age, he developed multiple atrial arrhythmias and underwent a revision of his Fontan with a 24 mm

intra-atrial conduit, Maze procedure, and placement of an epicardial pacemaker system. Preoperative catheterization prior to Fontan revision demonstrated no conduit regurgitation and minimal gradient (peak systolic ejection gradient 5 mm Hg). As a result, the conduit was not replaced at that time. Six years following his Fontan revision, he developed symptoms of abdominal ascites and lower extremity edema. Given his new and

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progressive symptoms, the patient was referred to the catheterization laboratory. Hemodynamic evaluation revealed mean Fontan pressures of 25 mm Hg secondary to elevated RV and LV end diastolic pressures (EDP) of 22 and 21 mm Hg, respectively. The peak systolic ejection gradient across the conduit was
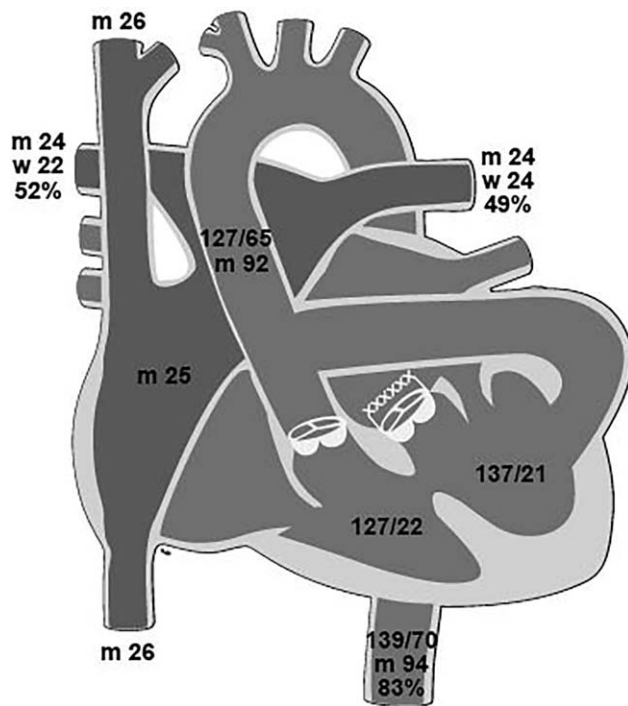

Fig. 1. Illustrative diagram depicting the patient's anatomy and hemodynamic measurements obtained during diagnostic catheterization. Ventricular pressures are noted as systolic/ end diastolic pressure. m = mean pressure, w = mean wedge pressure.

10 mm Hg (Fig. 1). The heavily and circumferentially calcified LV-AAo conduit had severe regurgitation (Fig. 2). The patient was an extremely high-risk surgical candidate secondary to his numerous prior sternotomies, high Fontan pressures, severe diastolic dysfunction, and calcified conduit with close proximity to the sternum. As a result he was referred back to the catheterization laboratory two months later for TVI within the LV-AAo conduit.

The LV-AAo conduit inserted on the leftward aspect of the ascending aorta following the lesser curve of the transverse arch. This created an almost 180 degree turn from the transverse arch to the distal insertion point of the LV-AAo conduit and precluded transcatheter valve delivery from a femoral arterial approach. In addition, the proximal LV-AAo conduit takeoff was from the LV apex with an oblique orientation to the ventricle, which precluded a transapical delivery approach. Vascular surgery was consulted to provide access via the right axillary artery. Consent for off-label use of a humanitarian device was obtained.

An 8 mm Dacron® graft was anastomosed at an oblique angle directly to the right axillary artery, which provided the most direct approach to the LV-AAo conduit via the right innominate artery (Fig. 3). The distal end of the tube graft was clamped with a vascular clamp for hemostasis and the catheterization was performed via direct puncture through the anterior aspect of the Dacron® graft. Once the conduit was accessed, a 20 mm × 82 mm Endurant II™ stent graft (Medtronic, Minneapolis, MN) was inserted using the 16 Fr Sentrant sheath (Medtronic, Minneapolis, MN). The stent graft was placed within the heavily calcified LV-AAo conduit prior to angioplasty to protect from potential
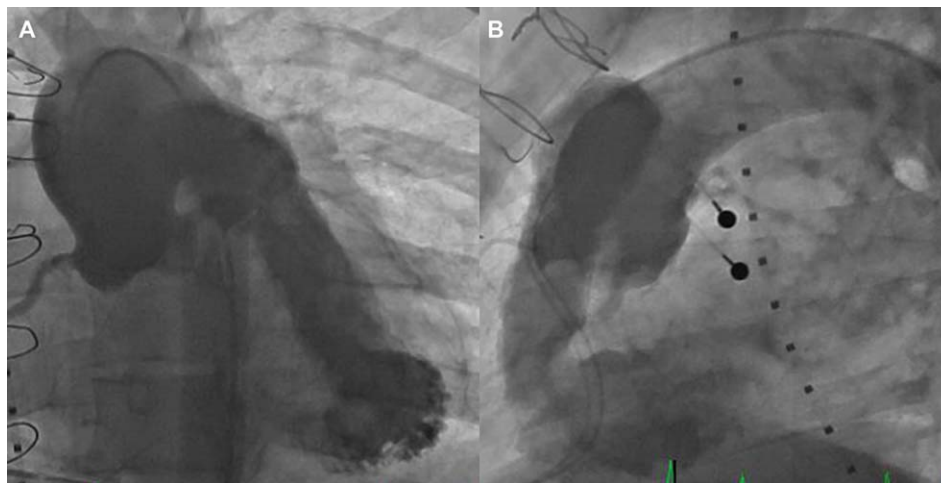
Fig. 2. The anteroposterior (A) and lateral (B) projections of an aortic root angiogram showing severe regurgitation of the calcified LV-AAo conduit. [Color figure can be viewed in the online issue, which is available at wileyonlinelibrary.com.]

catastrophic rupture secondary to its position within the high pressure systemic circulation. The stent graft also prevented potential calcific embolization/stroke as a result of anticipated aggressive conduit dilation. The calcified conduit created a very rigid tube which measured 18–20 mm along the entire course with little to no pulsatility. Therefore, we did not feel additional presenting was necessary and proceeded directly to TVI. A Melody Transcatheter Pulmonary Valve®
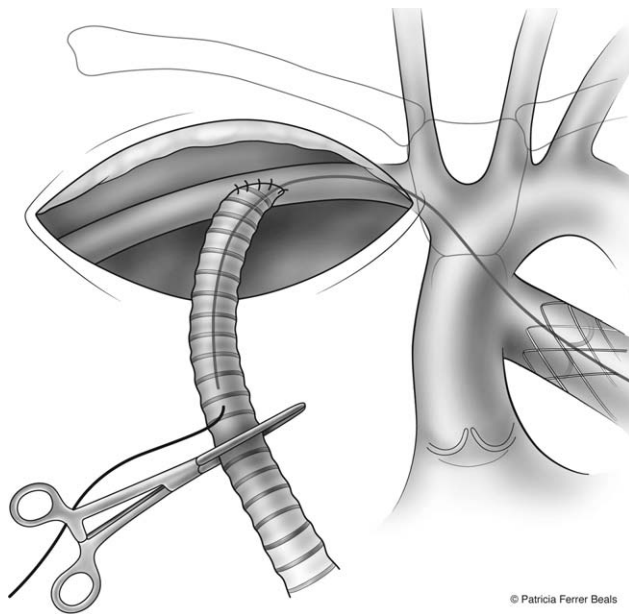

Fig. 3. Artistic illustration depicting the hybrid surgical approach used to obtain procedural access. The proximal Dacron® tube graft is sutured at an oblique angle to the axillary artery to facilitate procedural access. The distal Dacron tube graft was clamped and the anterior aspect of the graft was directly punctured for the catheterization.

(TPV) (Medtronic, Minneapolis, MN) was implanted within the LV-AAo conduit on a 20-mm Ensemble® Transcatheter Valve Delivery System (Medtronic, Minneapolis, MN) entirely within the distal end of the stent graft. Due to the significant residual narrowing within the Melody TPV following delivery, the entire length of the stent graft and Melody valve was dilated using a 20 mm × 2 cm Atlas Gold balloon (BARD Peripheral Vascular, Tempe, AZ) with effective relief of the stent narrowing (Fig. 4). Post-procedural LV EDP dropped considerably to 10 mm Hg and the LV-AAo conduit peak systolic ejection gradient was unchanged at 11 mm Hg. Angiography demonstrated no residual LV-AAo conduit insufficiency and no perivalvar leak (Fig. 5). The axillary artery access site was repaired and wound site closed. The patient tolerated the procedure well without complication and was discharged to home the following day.

DISCUSSION

We report the use of a transcatheter valve to treat a failing Fontan patient secondary to LV-AAo conduit insufficiency via a hybrid surgical approach. There are few publications describing transcatheter valve placement within the systemic semilunar valve position in patients with complex congenital heart disease [5–7]. The approaches reported include hybrid transapical, cutdown on the innominate artery, and percutaneously via femoral or internal jugular veins. Given the patient's unique anatomy, hybrid surgical access via a vascular graft provided an ideal approach for intervention in our patient. This approach has been utilized in adult patients to facilitate the implantation of transcatheter aortic valves and catheter-based Impella® left
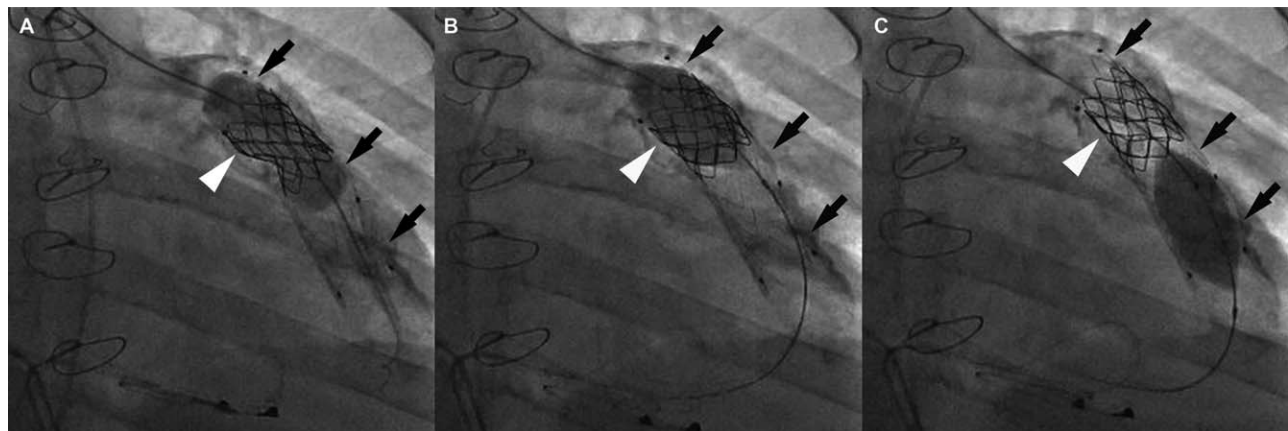
Fig. 4.   Intraprocedural images showing the delivery of the Melody TPV® within the Endurant II™ stent graft (A), balloon dilation of the Melody valve (B), and proximal Endurant II™ stent graft (C) with a 20 mm balloon. The Endurant II™ stent graft is noted by the black arrows and Melody TPV® is noted by the white arrowheads.

Fig. 5. Final angiogram of the aortic root demonstrating no valvar regurgitation or peri-valvar leak.

ventricular assist devices (ABIOMED, Inc. Danvers, MA) [8,9]. In addition to limiting the injury of the axillary artery, the tube graft provided additional vascular length for manipulation of the 100 cm long delivery system.

The Melody TPV is made from a bovine jugular venous valve which has naturally deep commissures allowing for competency in non-circular outflow tracts. The valve is supported by a platinum/iridium frame which is highly malleable and can conform to irregular conduits [10]. We were uncertain of the final configuration and long-term diameter of the heavily calcified LV-AAo conduit in this patient. As a result, we felt the Melody TPV would function better than a Sapien Valve (Edwards Lifesciences Corporation, Irvine, CA) in a possible elliptical configuration and in a generally smaller outflow diameter. Conduit tear is a known risk for TVI and was the most common serious adverse event reported in the one-year follow-up of the Melody TPV multicenter Post-Approval Study [11]. Heavily calcified conduits are at risk for rupture and have been successfully treated with covered stents such as the covered Cheatham Platinum stent (NuMED Inc., Hopkinton, NY) [11–13]. The calcium deposits within the LV-AAo conduit circumscribed its entire length and would have extended beyond a single covered stent. Deployment of the stent graft within the patient's conduit prior to any intervention mitigated the bleeding risks from conduit rupture in the high pressure systemic circulation and protected against possible calcific embolization from the heavily calcified conduit.

The stent graft was not used for typical Melody "prestenting" as is standardly performed in RV-PA conduits. The conduit stenosis provided an optimal landing zone for the transcatheter valve remote from the coronary arteries. The hybrid procedural approach coupled with the off-label use of the aortic stent graft and Melody TPV resulted in successful and uncomplicated treatment in this high-risk surgical patient.

Although the Melody TPV has performed well in relatively high pressure circulations, the long-term durability is not known and remains a concern in these off-label uses [5,14]. With the development of smaller diameter and more easily steerable transcatheter delivery systems, future re-intervention from the percutaneous femoral arterial approach may be possible. However in the absence of this technology, a similar well-planned hybrid approach remains a viable option for TVI in patients with complex congenital heart disease.

CONCLUSION

Innovative off-label use of transcatheter valve therapy can be used to treat high-risk patients, including failing Fontan patients that have limited medical and surgical options. Hybrid surgical approaches should be considered to provide the safest and most direct routes for TVI.

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