



Transcatheter Aortic Valve Replacement in Two Patients with Severe Aortic Insufficiency in the Setting Aortic Root and Valve Homografts for Congenital Aortic Disease: A Valve Planning Strategy for Congenital Heart Disease

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Abstract

The optimal therapeutic approach to severe aortic insufficiency in prior aortic root and valve homograft is not known and currently considered on an individual basis. We intervened in two individuals with severe aortic insufficiency in setting of an aortic root and valve homograft by placing a transcatheter aortic valve replacement (TAVR). Our first individual developed severe, symptomatic aortic insufficiency 18 years after a 20-mm homograft was placed for bicuspid aortic insufficiency and an ascending aortic aneurysm. Our second patient developed severe, symptomatic aortic insufficiency 13 years after a second repair requiring aortic root and valve replacement with a 20-mm aortic homograft for aortic insufficiency and ascending aortic ectasia. Both patients underwent successful TAVR with Edwards Sapien XT and 3 valves with excellent results and were discharged on post-operative day one. Both were symptomatically improved at follow-up with evidence of improvement in ventricular function on echocardiogram. Valve-in-valve TAVR within an aortic valve and root homograft can be considered as a less invasive method to treat homograft degeneration. Percutaneous valve therapies are attractive options in congenital heart disease and likely can be used in a valve planning strategy to manage the need and timing for repeat open heart surgery over a patient's lifetime.

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Introduction

Patients with congenital heart disease undergo multiple cardiac procedures in their lifetime with increasing risk with each operation. For patients with aortic valve and root disease, an aortic root homograft is sometimes preferable when infection or other concerns about a bioprostheses are present. Still, an aortic homograft is at risk for calcification and degeneration [1]. Almost 50% will require reoperation within 20 years though repair is often complicated by severe calcification of the coronary buttons and ascending aortic homograft [2]. The optimal approach to repair of a failing aortic root homograft is currently considered on an individual basis. Transcatheter aortic valve replacement (TAVR) has emerged as an option for treating aortic valve disease though is most commonly used in the elderly with severe calcific aortic valve stenosis [3-5]. We present two patients with severe aortic insufficiency in setting of an aortic root and valve homograft for congenital aortic disease who underwent TAVR. As valve-in-valve TAVR is utilized in patients high-risk for repeat sternotomy, TAVR in patients with failing aortic homografts has been considered before [6-11]. However, we present two patients with aortic valve and root homografts in the setting of congenital aortic disease and their follow-up to show that TAVR can be considered as part of a valve planning strategy in this patient group.

Case Presentation

Patient 1

At the age of 27, a woman with a history of a bicuspid aortic valve underwent aortic valve and root replacement with a 20-mm cryopreserved homograft with coronary reimplantation after developing severe aortic insufficiency accompanied by a dilation of ascending aortic measuring 6 cm. At the age of 43, she developed worsening dyspnea and evidence of valvular degeneration. On physical exam, she had a wide pulse pressure (blood pressure 129/45 mmHg), a 3/6 blowing diastolic



Figure 1: Echocardiogram in parasternal long axis view in diastole showing severe aortic insufficiency.

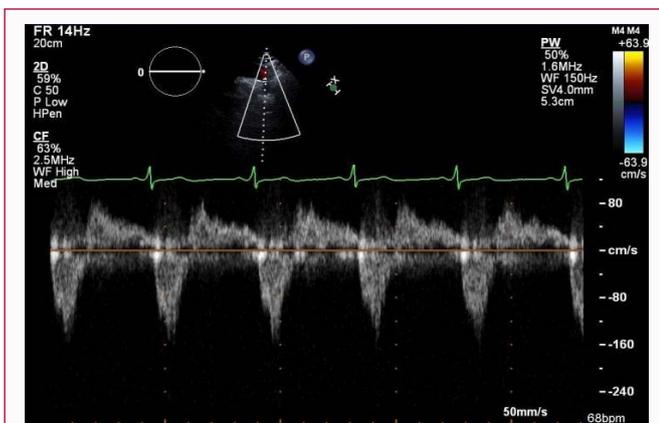


Figure 2: Pulse-wave Doppler in descending aorta showing systolic flow reversal consistent with severe aortic insufficiency.



Figure 3: Cardiac CT showing severe calcification of the ascending aortic homograft.

murmur, a pulsatile precordium, and bounding carotid upstrokes. Her echocardiogram demonstrated severe aortic insufficiency with pressure half-time 250 milliseconds, peak aortic velocity of 3.14 m/s, mean gradient of 22 mmHg, diastolic flow reversal in the proximal abdominal aorta, preserved left ventricular ejection fraction, and an increase in her left ventricular end-systolic diameter as well as mild-moderate mitral regurgitation and mildly elevated right ventricular

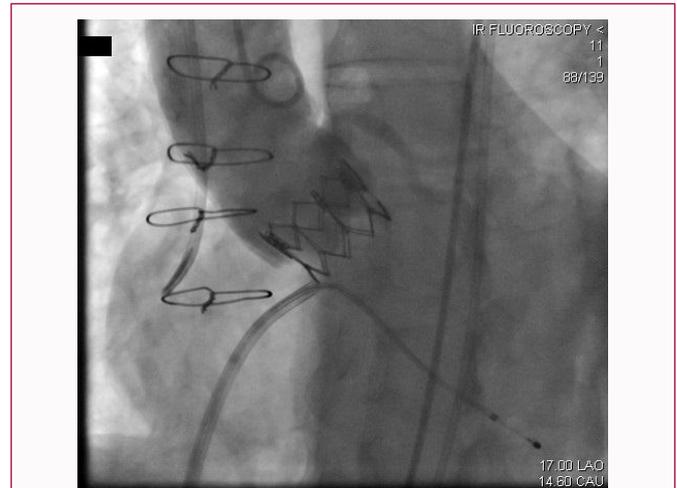


Figure 4: Angiographic view after delivery of 26 mm Edwards Sapien XT TAVR.

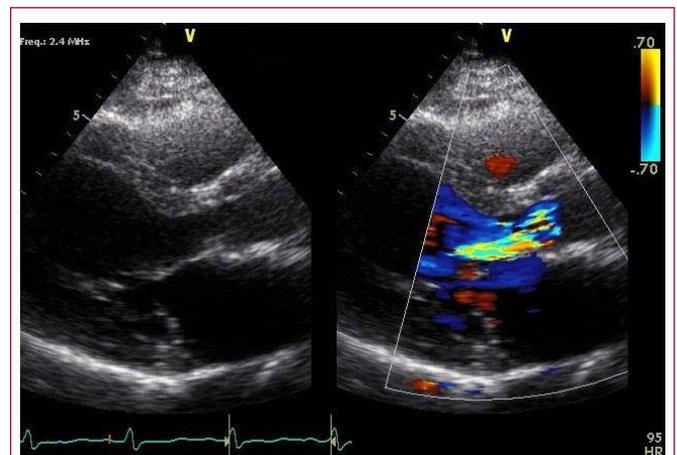


Figure 5: Echocardiogram in parasternal long axis view with and without color Doppler in diastole showing severe aortic insufficiency.

systolic pressure of 39 mmHg (Figure 1,2). As her aortic root homograft had become severely calcified with severe calcification of her coronary buttons, open surgical repair was high risk (Figure 3). TAVR was considered the best option.

She underwent TAVR via the transfemoral approach with the placement of a 26 mm Edwards Sapien XT valve under general anesthesia with transesophageal echocardiographic guidance (Figure 4). Immediately following valve deployment, the aortic diastolic pressure improved from 30 mmHg to 80 mmHg and the left ventricular end-diastolic pressure decreased from 25 mmHg to 12 mmHg. Only trace anterior paravalvular leak was demonstrated by echocardiography and angiography. She tolerated the procedure well and was discharged home on her first post-operative day. Her echocardiogram prior to discharge demonstrated no valvular aortic insufficiency, a mean gradient across the aortic valve of 12 mmHg, trace anterior paravalvular leak, preserved left ventricular ejection fraction, and trace mitral regurgitation. Two weeks after her procedure, her symptoms were markedly improved and she had returned to work. Her blood pressure was 136/96 mmHg and her femoral access sites were well healed. Her mild hypertension persistent and she was started on 3.125 mg carvedilol twice daily. At 13 months, she returned for follow-up. Her follow-up echocardiogram showed



Figure 6: Live 3D echocardiogram in parasternal long axis view demonstrating prolapse of aortic valve homograft leaflet resulting in severe aortic insufficiency.



Figure 7: 3D volume rendering CT of aortic root and arch, coronary arteries, and outline of left ventricular endocardium. Within the ascending aortic homograft, there is severe calcification.

the valve remained well-positioned without evidence of paravalvular leak. Her exercise capacity continued to improve. At this point, she was walking daily and working normal hours. Her need for further valvular therapy in the future will be considered weighing the options of open heart procedures versus percutaneous approaches.

Patient 2

A 56-year-old woman with hypertension, asthma, and a history of aortic coarctation presented with worsening dyspnea and orthopnea. She has a history of aortic coarctation and underwent repair at age 17 though subsequently developed ectasia of the ascending aortic and required aortic valve and root replacement with a 20-mm aortic homograft with coronary reimplantation at age 43. Her internist noticed a loud diastolic murmur and her echocardiogram demonstrated valvular degeneration and severe aortic insufficiency. Specifically, she had degeneration of the aortic valve with a leaflet prolapsing into the left ventricular outflow tract resulting in severe aortic insufficiency (Figure 5,6). The mean gradient across the valve



Figure 8: Angiographic view during delivery of 23 mm Edwards Sapien 3 TAVR.

was 35 mmHg and the left ventricular end-diastolic diameter was 5.5 cm. There was also mild left atrial dilation with a left atrial volume index of 43 mL/m² and mild-moderate mitral regurgitation though her left ventricular systolic function was hyperdynamic. Her two prior sternotomies made a reoperation high risk especially in the setting of a calcified ascending aorta. TAVR was thought to be the best approach.

She underwent TAVR via the transfemoral approach under moderate sedation. Her homograft had significant calcification and an area of focal kinking at the level of annulus (Figure 7). The left ventricular outflow tract was larger than the annulus however such that a 23 mm Edwards Sapien 3 valve was selected for implantation. After deployment, the valve was post-dilated with a 20 mm True balloon (Figure 8). Immediately following valve deployment, the left ventricular end-diastolic pressure decreased from 40 mmHg to 15 mmHg. She tolerated the procedure well and was discharged the next day. At 30 days, she felt remarkably improved and returned to work. She was no longer taking furosemide. Her follow-up echocardiogram showed no valvular aortic regurgitation, a mean gradient 15 mmHg, no paravalvular leak, a low-normal left ventricular ejection fraction of 50%, and an improved left ventricular end-diastolic diameter of 5.0 cm.

Discussion

The management of congenital aortic disease later in life is currently considered on an individual basis. The need for high risk procedures is growing as medical therapy for chronic diseases has improved. We present two individuals with at least one prior sternotomy for congenital aortic disease and prior aortic valve and root homografts who were successfully managed with TAVR when the risk of an open surgical repair outweighed the benefit. Both patients had uneventful procedures via the transfemoral approach and were discharged home the following day. Both exhibited substantial hemodynamic improvement on echocardiography. Both had improvement in exercise capacity and went back to work shortly after their procedures.

Failure of an aortic root and valve homograft is an inevitable consequence in young individuals. Nearly half of patients with an aortic root and valve homograft will face a repeat operation within 20

years [5]. Failure of an aortic homograft is often associated with severe calcification of the coronary buttons and ascending aortic homograft and requiring replacement of a large segment of ascending aorta [12]. In the prior cases, access was variable demonstrating the differing clinical circumstances in this population. Four used the transfemoral approach; one transaortic, one subclavian, and one transapical. The earliest discharge was on post-operative day two though most required three to five days in the hospital after their procedure. TAVR also has been implored in the setting of a failing Bentall procedure [13]. Two patients with Marfan syndrome underwent successful TAVR after prior ascending aortic dissections requiring Bio-Bentall procedures with a bioprosthetic aortic valves and Dacron aortic root grafts. Our patients had less co-morbidity outside of these aortic disease and prior surgery. In the setting of congenital aortic disease for coarctation, Marfan syndrome, or bicuspid aortic disease, patients undergo aortic surgery at a younger age and often encounter a subsequent valvular dysfunction later in life.

The commonly used cardiac surgical risk scores (Euro SCORE, STS score, etc.) often underestimate surgical risk in the congenital heart disease population [14]. Furthermore, in the subset of patients with a failed aortic root homograft, standard risk scores do not take into account potential adverse consequences involved in homograft replacement. TAVR has been described successfully in this population and represents an alternative to repeat open surgery for a failure homograft. Our case series demonstrates two patients who tolerated TAVR well via the transfemoral approach with short hospital stays. Additionally, for the first time, we present TAVR for a failed homograft using the minimalist approach. In this challenging population, minimally invasive therapies are especially needed. As the congenital heart disease population increases, percutaneous valve treatments are attractive options and likely can be used in a valve planning strategy to manage the need and timing for repeat open heart surgery. For example, patients with congenital heart disease may now opt to use a combination of surgical and percutaneous valvular therapy to minimize the need for successive sternotomies and plan the optimal timing of surgery when needed over a patient's lifetime. Information is forthcoming regarding the long-term durability of percutaneously deployed valves.

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