Case Reports

Transcatheter Aortic Valve-in-Valve Replacement

Instead of a 4th Sternotomy in a 21-Year-Old Woman with Aortic Homograft Failure

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Transcatheter aortic valve replacement (TAVR) is a well-established method for replacing native aortic valves; however, it was conceived for elderly patients with aortic valve stenosis, and the lack of data on long-term durability has led practitioners to restrict the use of TAVR to patients who have short life expectancies. Here, we describe the case of a 21-year-old woman who had undergone 3 previous open aortic valve replacements and who presented with symptoms of recurrent valvular failure. Transthoracic echocardiograms and computed tomographic angiograms revealed a degenerating aortic root homograft with substantial calcification, moderate-to-severe aortic valve stenosis, and severe aortic valve regurgitation. Open surgical valve replacement posed substantial risk to our patient, so we decided to perform valve-in-valve TAVR with use of the Edwards SAPIEN XT Transcatheter Heart Valve. The patient's pulmonary artery pressure, valvular regurgitation, and symptoms improved substantially thereafter. We found that valve-in-valve TAVR into a failing aortic root homograft was less invasive than repeat surgical valve replacement in this young patient who had congenital vascular anomalies and a complex surgical history. ***(Tex Heart Inst J 2016;43(4):334-7)***

Key words: *Aortic valve insufficiency/therapy; cardiac catheterization/instrumentation/methods; heart valve prosthesis implantation/methods; reoperation; risk factors; treatment outcome*

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Aortic root homografts are a rarely used option for aortic valve replacement (AVR). Such use is typically reserved for highly complex repairs in which a tissue valve is desirable, but a bioprosthetic tissue valve is unsuitable because of infection or other factors. Like bioprosthetic tissue valves, the durability of a homograft is unclear; in time, these valves tend to degenerate and calcify. Repeat AVR surgery on a failed aortic root homograft is technically challenging and has many associated risks. Degenerative tissue failure and extensive calcification of the homograft well beyond the region of the aortic valve are thought to be responsible for the technical difficulties, because repair typically necessitates replacing a large portion of the proximal aorta. Aortic root homografts are rarely used; thus, few data are available to determine the ideal method for repeat homograft replacement.[1-4]

Transcatheter aortic valve replacement (TAVR) is a well-established treatment for elderly patients who have severe, symptomatic aortic stenosis and who are at high surgical risk.[5] Because this treatment's long-term results are unknown, some cardiac surgeons and interventional cardiologists think that TAVR should be reserved for older patients or those with short life expectancies. As TAVR use has become widespread, there has been great interest in using it for degenerated bioprosthetic tissue valves in order to avoid repeat open AVR; these valve-in-valve approaches are performed in a fashion similar to TAVR of the native aortic valve.[6] As a derivation of this approach, TAVR has been used in a few instances of degenerated homografts.[1,3,4] We report a case of TAVR in a young adult who had previously undergone 3 open AVRs and who presented with severe calcific homograft stenosis.

Case Report

In December 2013, a 21-year-old woman with New York Heart Association class III heart failure presented with increased fatigue, dyspnea on exertion, right-upper-quadrant abdominal pain, hepatomegaly, lower-extremity edema, and a history of congenital aortic valve stenosis. She had been born in China, where, at 2 years of age, she underwent repair of a ventricular septal defect and patent ductus arteriosus through a posterior left thoracotomy. At age 5 years, she underwent AVR through a median sternotomy, receiving a bileaflet mechanical valve. At age 6 years, she moved to the

United States. At age 10 years, she underwent repeat sternotomy and AVR by means of the Konno procedure with use of a 21-mm Carpentier-Edwards bioprosthetic tissue valve (Edwards Lifesciences Corporation; Irvine, Calif). At age 14 years, she again needed a sternotomy, this time for reimplantation of the coronary arteries, annulus débridement, and aortic root replacement with use of a 24-mm cryopreserved homograft.

Upon the 21-year-old patient's presentation at our institution, a transthoracic echocardiogram showed a degenerating aortic homograft, moderate-to-severe stenosis, severe transvalvular regurgitation (peak gradient, 64 mmHg), and mild concentric left ventricular (LV) hypertrophy. The noncoronary cusp of the homograft appeared to be flail, with an associated 2.3-cm, hyperechoic, mobile mass. In addition, the patient had a moderately enlarged left atrium, moderate-to-severe mitral regurgitation, and moderate tricuspid regurgitation. Her estimated pulmonary artery pressure (PAP) was 55 to 60 mmHg. Computed tomographic angiograms of the chest showed diffuse aortic root calcification (Fig. 1A) and a 22.4 × 17.9-mm aortic annulus (Fig. 1B); 3-dimensional reconstruction revealed the congenital absence of the abdominal aorta just below

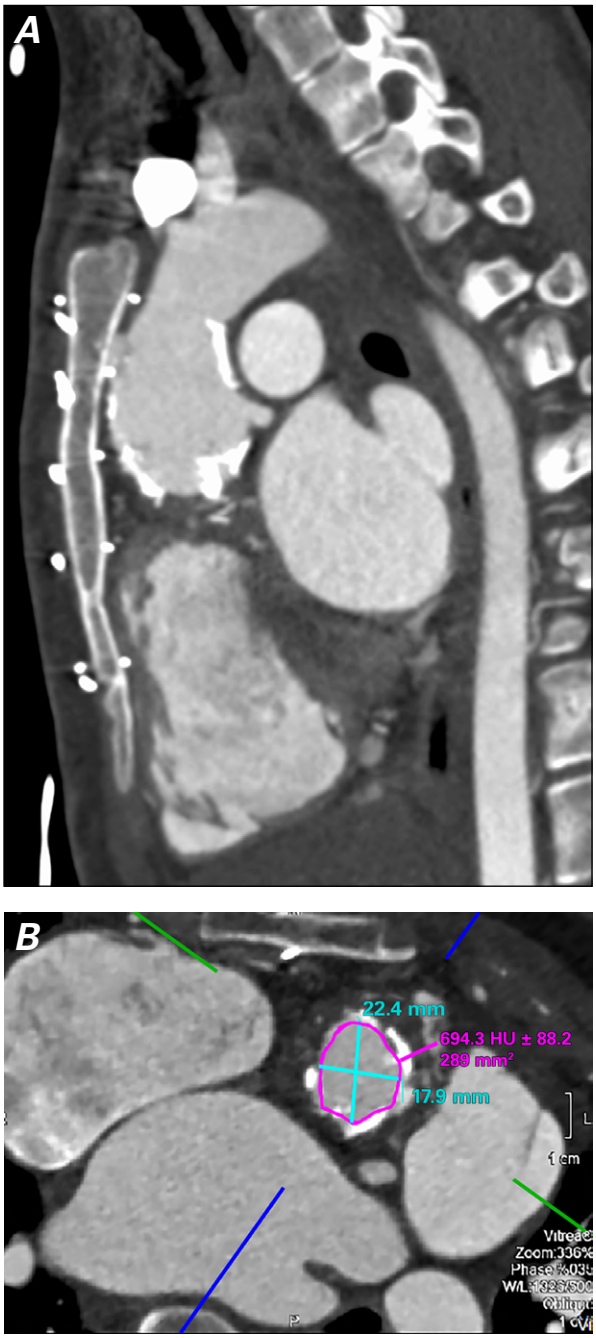

Fig. 1 *Preprocedural computed tomographic angiograms show* ***A****) a diffusely calcified homograft near the ostium of the left coronary artery and* ***B****) a 22.4 × 17.9-mm aortic annulus.* ***C****) Three-dimensional reconstruction image of the abdomen and pelvis shows the absence of the infrarenal aorta, and large, bridging, collateral-vessel reconstitution of the distal common iliac arteries, just proximal to the bifurcation of the external iliac arteries.*

the renal arteries (Fig. 1C). Both common iliac arteries had reconstituted themselves distally through superior mesenteric collateralization. The patient's logistic EuroSCORE was 14.8%. Left-sided selective coronary angiography revealed no angiographic evidence of coronary artery disease. The patient's LV end-systolic and end-diastolic pressures were 144 and 32 mmHg, respectively. Her aortic pressure was 84/40 mmHg. Because of the high surgical risk of a 4th sternotomy and the congenital absence of the infrarenal aorta, we decided to perform TAVR through a minithoracotomy from a transapical approach.

The patient was placed under general anesthesia. A transesophageal probe was inserted for continuous monitoring. A full cardiopulmonary bypass system was primed in case rapid surgical conversion became necessary. A 5F, balloon-tipped, bipolar pacing catheter was advanced through a 6F jugular vein sheath and positioned in the right ventricle, and pacing thresholds were tested. Another 6F sheath was placed percutaneously in the right radial artery, a 6F angled pigtail catheter was advanced over a 0.035-in J-wire and positioned in the ascending aorta, and aortography was performed. Access to the left radial artery was attained by using a micropuncture introducer set and a 6F sheath. Heparin was administered to achieve an activated clotting time of 300 s. Because the left main coronary artery was so close to the homograft, a Judkins left 4 guiding catheter was advanced over a 0.035-in J-wire under fluoroscopic guidance and inserted into the left coronary artery (Fig. 2A). A 0.014-in guidewire and a 3.5 × 16-mm drug-eluting stent were positioned in the mid-left anterior descending coronary artery; the guiding catheter was then retracted from the ostium into the ascending aorta (Fig. 2B).

Next, a 5-cm thoracotomy incision was made in the left intercostal space, and pledgets were sewn around the apex of the LV. A 24F RetroFlex® 3 introducer sheath (Edwards Lifesciences) was inserted into the LV apex over a 0.035-in guidewire. A 5F, 65-cm internal mammary artery catheter (Cordis, a Johnson & Johnson company; Miami Lakes, Fla) was passed over a 0.035-in J-wire through the prosthetic valve, around the arch, and into the descending thoracic aorta. The guidewire was then exchanged for a 0.035-in Amplatz Super Stiff™ guidewire (Boston Scientific Corporation; Natick, Mass). Predilation was not performed because of the severe regurgitation. Because of the compliant nature of the degenerated donor tissue in the previous 24-mm homograft, a 26-mm transapical Edwards SAPIEN® 9000TFX delivery system (Edwards Lifesciences) was aligned and positioned under fluoroscopic and echocardiographic guidance. During rapid ventricular pacing (rate, 160 beats/min), the 26-mm SAPIEN® XT Transcatheter Heart Valve was deployed and expanded into position with use of a 26-mm Edwards

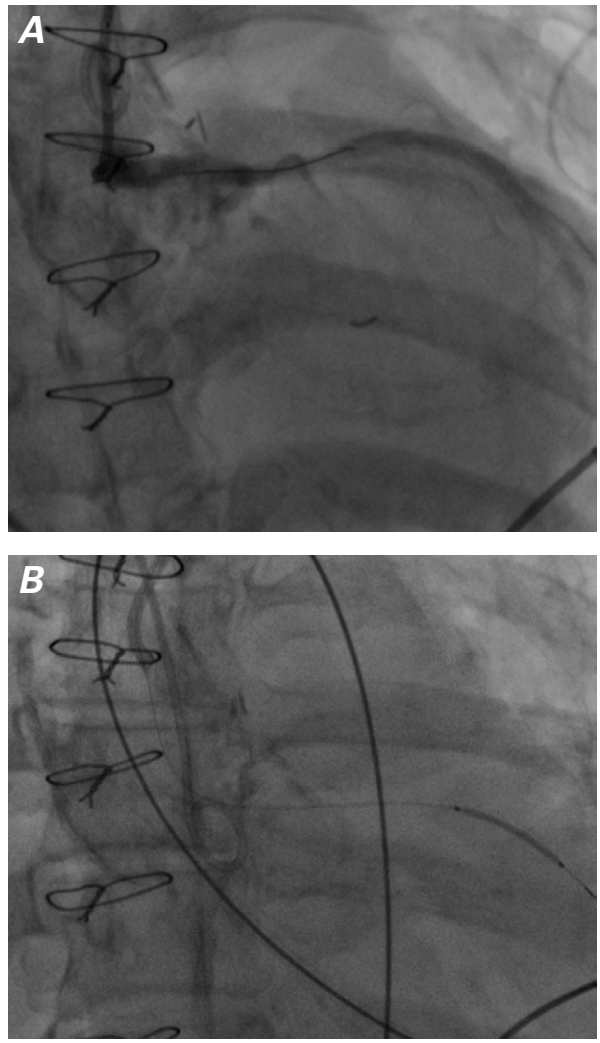

Fig. 2 *Selective intraprocedural angiograms of the left coronary artery.* ***A****) The distal tip of a 0.014-in guidewire is inserted into the lumen of the mid-left anterior descending coronary artery.* ***B****) A 3.5 × 16-mm drug-eluting stent is safely positioned over that guidewire. The guiding catheter was then retracted from the left main coronary artery ostium and kept in the ascending aorta for immediate recanalization, if necessary.*

Ascendra® Balloon Aortic Valvuloplasty Catheter (Edwards Lifesciences) (Fig. 3). Accurate valve placement was confirmed angiographically.

Post-deployment echocardiograms showed a well-seated valve and minimal paravalvular regurgitation. The mean gradient had improved from 39 to 17 mmHg. During rapid pacing, the delivery system and sheath were removed from the LV, and the apex was surgically closed with the previously placed pledgets. A chest tube was placed for drainage, and the minithoracotomy incision was closed in standard fashion. The guiding catheter, guidewire, and stent were removed, along with the transvenous pacemaker. Both radial artery sheaths were then removed. Heparinization was reversed, and the patient was transferred to the cardiovascular recov-

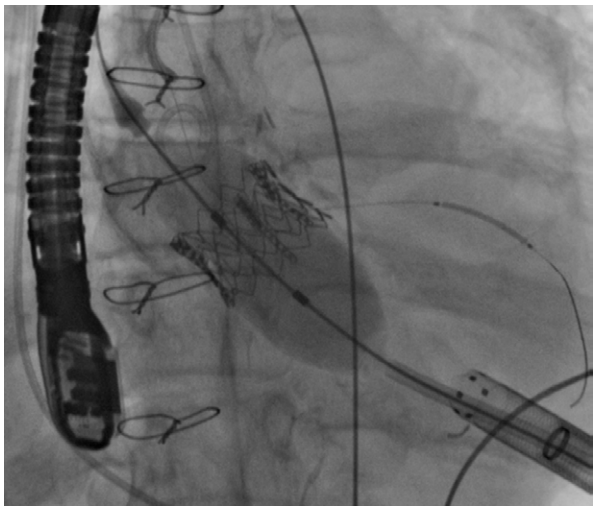

Fig. 3 *Angiogram shows the 26-mm Edwards SAPIEN XT Transcatheter Heart Valve deployed and expanded into position with use of a 26-mm Edwards Ascendra Balloon Aortic Valvuloplasty Catheter.*

ery room in hemodynamically stable condition. She was mobilized the next morning and reported immediate improvement in her dyspnea and fatigue. Echocardiograms showed trace paravalvular regurgitation and improved mitral regurgitation. The PAP had decreased from 55 to 45 mmHg. The patient was discharged from the hospital in stable condition on postoperative day 3.

One year later, the patient reported that she had remained symptomatically and functionally improved. She had upper-quadrant abdominal distention but had resumed normal activity, including walking 1.5 miles daily (without dyspnea). Echocardiograms showed a mild-to-moderate posterior paravalvular leak with a mean aortic valve gradient of 20 mmHg. The patient's LV ejection fraction was normal, and she had mild-to-moderate mitral regurgitation with a PAP of 45 to 50 mmHg.

Discussion

Repeat AVR is associated with significantly higher morbidity and mortality rates than is first-time valve-replacement surgery.[7] The number of patients referred for valve-in-valve replacement of degenerated bioprosthetic aortic valves is rapidly increasing.[4,6] The feasibility of percutaneous or minimally invasive valve-in-valve placement of the Edwards SAPIEN valve into failing aortic root homografts—an off-label use—has been discussed in a few case reports.[1,3,4] With the use of the radiopaque Edwards SAPIEN bioprosthesis, valve-in-valve replacement can be comparatively simple. We used the transapical approach because the direct access enabled optimal coaxial alignment and positioning of the new valve within the degenerated homograft; this transapical approach was first reported by Attia and colleagues[8] to treat a patient who had a degenerated bioprosthesis. Although radiopaque markers are typically not available in cryopreserved homografts, it is possible that these markers will soon be added to bioprosthetic aortic valves. Upon the addition of these markers, a more traditional TAVR approach could be used.

Preliminary global registry data show that gradients, competency, and functional class are maintained with use of the Edwards SAPIEN valve at one-year follow-up evaluation.[6] However, clinical experience is still comparatively minimal, and long-term follow-up data are needed to establish the clinical usefulness of valve-in-valve implantation for treating homograft degeneration.[1,3,4]

In our patient, a 4th open AVR would have been prohibitively risky. Pending the results of long-term follow-up studies and more experience, we think that similar young patients who have undergone multiple valve replacements might benefit from TAVR.

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