Case Reports

Transcatheter Aortic Valve Implantation in a Patient with Unicuspid Aortic Valve

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Transcatheter aortic valve replacement (TAVR) in patients with bicuspid aortic valves has been successfully performed, but there is a lack of published experience in percutaneous treatment of patients with unicuspid valves and severe aortic stenosis. We describe a case of TAVR in such a patient.

A 31-year-old woman with Turner syndrome—who had undergone coarctation repair via subclavian flap at age 7 days and an aortic valvotomy at age 6 weeks—presented with severe symptomatic aortic stenosis. She was deemed inoperable because of her severe pulmonary hypertension and numerous comorbidities; consequently, a 20-mm Edwards SAPIEN 3 Transcatheter Heart Valve was offered for compassionate use. Postdeployment angiography and transesophageal echocardiography and aortography revealed no aortic insufficiency.

Transcatheter aortic valve replacement for unicuspid aortic valve stenosis is technically feasible. Before implantation, particular attention should be paid to the interplay between the large single leaflet, coronary ostia, and stented valve, to select the correct size and position of the device. Some degree of intraoperative aortic migration should be anticipated.
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The concept of a malformed unicuspid aortic valve was first put forth by Edwards in 1958.[1] Congenitally malformed aortic valves are present in 1% of live births in the United States; of these malformed valves, the frequency of bicuspid-to-unicuspid is 85% to 15% in males and 95% to 5% in females. In a 2012 case series,[2] none of the examined unicuspid valves had normal function. A strong prevalence of severe stenosis was found in association with aortic isthmic coarctation, congenitally bicuspid pulmonic valve, quadricuspid pulmonic valve, parachute mitral valve or single papillary muscle syndrome (Shone syndrome), Turner syndrome, and congenital coronary anomaly.[2]

Although the published experience with transcatheter aortic valve replacement (TAVR) in patients with bicuspid aortic valves has increased,[3] there currently appears to be no published experience on percutaneous treatment for severe aortic stenosis (AS) in patients with unicuspid valves. Here we document the case of a patient with a unicuspid aortic valve who presented with severe AS and was treated with TAVR.

Case Report

In September 2014, a 31-year-old woman affected by Turner syndrome presented with severe symptomatic AS. Because multiple comorbidities—including severe pulmonary hypertension, frailty, hepatitis C-related liver cirrhosis (Child-Pugh score B), hypothyroidism, diastolic dysfunction, and chronic atrial flutter—rendered her ineligible for surgical intervention, she had been referred to our outpatient clinic for consideration of TAVR. The patient's medical history included, at 7 days after birth, surgical correction of aortic coarctation with a subclavian flap; and, at 6 weeks of age, an aortic valvotomy.

Baseline echocardiograms revealed a unicuspid aortic valve with a preserved ejection fraction of approximately 0.50, severe AS, and severe pulmonary hypertension (Fig. 1). The aortic valve area was estimated to be 0.45 cm^2 with a mean gradient of 46 mmHg, peak transaortic velocity of 4.09 m/s, and estimated right ventricular systolic pressure of 84 mmHg.

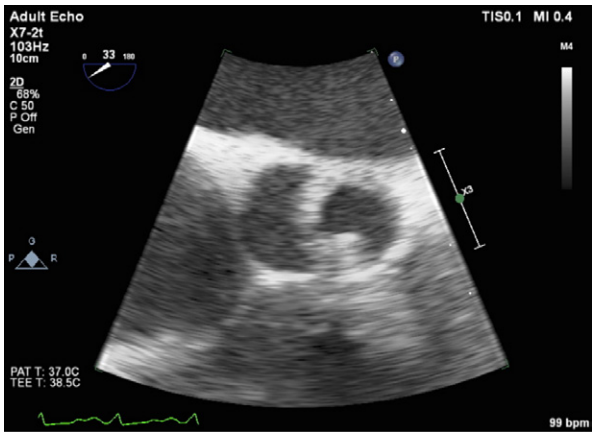

Fig. 1 *Transesophageal echocardiogram shows severe aortic stenosis in the presence of a unicuspid aortic valve.*

A computed tomogram of the chest revealed that the aortic annulus was 22.3 × 17.3 mm with an annular area of 307 mm^2, in the absence of any substantial calcification. The left ventricular outflow tract (LVOT) was 16.9 × 22.2 mm in size. The right and left iliofemoral arteries showed minimal luminal diameters: 5.3 mm on the right and 5.8 mm on the left. No disease was noted in the coronary arteries.

Although our patient's surgical risk was estimated to be 3.1% (Society of Thoracic Surgeons score), she was deemed inoperable by our cardiovascular surgeons because of her frailty, severe pulmonary hypertension, hepatitis C-related liver cirrhosis, hypothyroidism, diastolic dysfunction, and chronic atrial flutter. Therefore, she was offered transcatheter implantation of a 20-mm Edwards SAPIEN 3 valve (Edwards Lifesciences Corporation; Irvine, Calif) on a compassionate-use basis.

A 20-mm SAPIEN 3 valve (with 6% annular oversizing) was selected to match our patient's annular size. The main vascular access was obtained at the left common femoral artery, then the artery was "pre-closed" with use of 2 Perclose ProGlide® Suture-Mediated Closure Systems (Abbott Vascular, part of St. Jude Medical, Inc.; Redwood City, Calif), deployed in cross fashion. A 14F Edwards eSheath (Edwards Lifesciences) was inserted without sequelae and was sutured in place. The aortic valve was easily crossed with a generic AL1 catheter. An Amplatz Super Stiff™ Guidewire (Boston Scientific Corporation; Natick, Mass) was used for added support (helpful in the presence of a horizontal aorta), and we performed the aortic valvuloplasty with use of an 18-mm × 4-cm Z-MED™ balloon (Numed, Inc.; Hopkinton, NY). An aortogram helped us determine if the native leaflet would obstruct flow into the coronary vessels (Fig. 2). The valvuloplasty revealed Thrombolysis In Myocardial Infarction (TIMI)-3 coronary flow, with the lucency of the native leaflet just below the coronary ostia. The Edwards SAPIEN 3 Commander delivery system (Edwards Lifesciences) was inserted, and valve alignment was performed in the descending aorta without difficulty. The aortic annulus was easily crossed. We canted the valve within the annulus and adjusted it to a more coaxial position by applying the maximum distal flex of the Commander delivery system, modifying the distal loop, and adjusting the tension on the 0.035-in Amplatz wire. Because of the small LVOT diameter measured earlier, the primary operator decided to position the center marker at the insertion of the leaflets. This was done in anticipation of a substantial migration to a more aortic position due to the lack of annular calcium and to our reliance on the external skirt to prevent paravalvular leaks (Figs. 2 and 3).

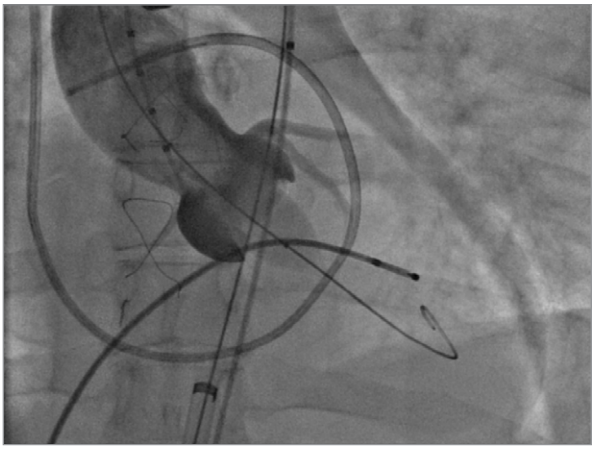

Fig. 2 *Aortogram shows the inflated balloon with concomitant coronary flow.*

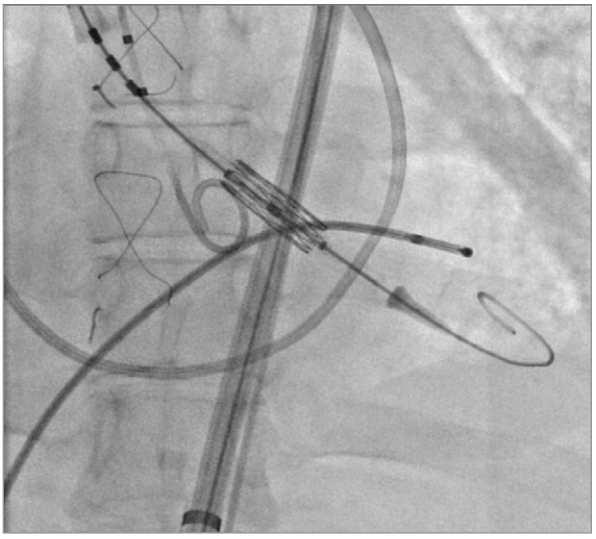

Fig. 3 *Postdeployment angiogram shows the valve in relation to the sinotubular junction and the aortic annulus. The presence of the aortic annulus can be inferred from the position of the pigtail catheter in the noncoronary cusp (in the absence of substantial calcification).*

The 20-mm SAPIEN 3 valve was deployed with a slow continuous inflation, which revealed a slight parallax. Once the delivery-system balloon was deflated, the valve shifted to a more horizontal position; the noncoronary cusp side was at an 80/20 aortic position, and the left coronary cusp side was at a 60/40 aortic position. A pigtail catheter was advanced into the left ventricle to document a transvalvular gradient of 8.5 mmHg. Post-deployment transesophageal echocardiography and aortography revealed no aortic insufficiency (Figs. 4 and 5).

The 14F eSheath was removed and the arteriotomy site was closed percutaneously with success. The patient was extubated and transferred to our institution's intensive care unit in stable condition. Despite a straightforward procedure, she needed long-term acute care and a lengthier recovery than might have been anticipated after a successful and relatively short procedure. In retrospect, any alternative surgical option might not have been tolerated at all by this patient.

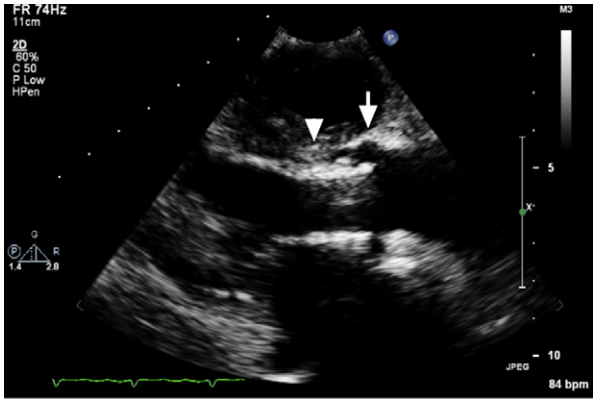

Fig. 4 *Transesophageal echocardiogram shows the deployed valve in relation to the sinotubular junction (arrow) and the aortic annulus (arrowhead).*

Fig. 5 *Final angiogram shows the deployed valve in relation to the sinotubular junction and the aortic annulus. Note the absence of significant aortic regurgitation (suggested by the absence of contrast medium in the left ventricle).*

Discussion

This case illustrates the feasibility of TAVR in patients with unicuspid aortic valves and appears to be the first reported case in the scientific literature. The particular nature of the native valve posed several challenges in device selection and implant strategy. The choice of an Edwards SAPIEN 3 valve was deemed an advantage over an Edwards XT Transcatheter Heart Valve because of the extensive external skirt, which we hoped would reduce or eliminate paravalvular leaks that might be caused by the irregular "reverse funnel" shape of a unicuspid valve. The absence of calcification and the funnel-shaped geometry of the LVOT might have prevented successful anchoring of the valve at the annulus, so we paid particular attention to the size of the LVOT and annular diameters with the aid of computed tomography and transesophageal echocardiography. The predeployment position was slightly lower (purposefully) than that of a conventional SAPIEN 3.

The valve was implanted only after careful examination of the degree of coronary flow during the balloon aortic valvuloplasty, to rule out the possibility that a large single leaflet could have occluded one of the ostia after valve deployment. We proceeded with the valve implantation only after observing good contrast flow in the native coronary arteries during the valvuloplasty step. Some degree of uncertainty was anticipated in the final position of the implanted valve after deployment because of the interplay between a fully deployed stented valve and a single large native leaflet. We in fact observed a substantial degree of aortic migration during deployment and a slight horizontal tilt toward the left coronary cusp once the deployment device was removed.

We thought that the percutaneous option for this patient was the only viable one because of our patient's advanced clinical presentation and severe, disabling symptoms. Given our center's surgical and cardiologic expertise and our extensive experience in the performance of TAVR procedures (>800 in the last 4 yr), we deemed it appropriate to offer the percutaneous option to this patient on a compassionate, off-label indication.

We found that the Edwards SAPIEN 3 valve was particularly suitable in dealing with this patient's extremely difficult anatomy because of the valve's low profile, ease of deployment, and skirt design. Device selection, together with careful radiologic, angiographic and anatomic planning, remains the mainstay for procedural success as we progress in our experience with TAVR in extreme conditions.

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