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Transcatheter aortic valve replacement for bicuspid aortic valve stenosis

BICUSPID AORTIC VALVE is the most common congenital cardiac abnormality in humans and is a significant risk factor for premature aortic valve dysfunction due to accelerated leaflet deterioration and calcification from altered hemodynamics. From 20% to 50% of patients with bicuspid aortic valve need aortic valve replacement during their lifetime, mostly for aortic stenosis.^{1,2}

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While 0.5% to 2% of the general population are born with a bicuspid aortic valve, more than 40% of patients (mainly younger patients) who undergo surgical or transcatheter intervention for aortic valve disease in some cohorts have this abnormality, suggesting that its true prevalence may be underreported.³

In the past decade, transcatheter aortic valve replacement (TAVR) has cemented its place as an option for patients with severe tricuspid aortic stenosis who cannot undergo surgery because their surgical risk is intermediate or high.⁴ However, most of the studies of balloon-expandable and self-expanding TAVR devices have excluded patients with bicuspid aortic valve.

■ BICUSPID AORTIC VALVE POSES CHALLENGES FOR TAVR

As TAVR is explored in younger and lower-risk populations, in which the prevalence of bicuspid aortic valve is presumably higher, the discussion of feasibility, safety, and efficacy of TAVR in patients with bicuspid aortic valve is both important and timely.

doi:10.3949/ccjm.85a.18101

Bicuspid aortic valve is commonly categorized according to the Sievers classification,⁵ which describes 3 main morphologic types (designated types 0, 1, and 2) according to the number of raphe connecting the leaflets. Unique anatomic features of bicuspid aortic valve render the TAVR procedure challenging in these patients and merit consideration. These include, but are not limited to:

- Asymmetric calcification of the valve leaflets and calcified raphe. This results in asymmetric and incomplete expansion of the prosthesis, leading to incomplete sealing and paravalvular leak.
- A larger and more elliptically shaped aortic annulus, leading to challenges with proper sizing and apposition of the prosthesis
- Concomitant aortopathy, posing a higher risk of aortic rupture, dissection, paravalvular leak, and other complications during implantation.

Thus, compared with patients with tricuspid degenerative aortic stenosis, patients undergoing TAVR who have bicuspid aortic valve have a higher short-term risk of death and a higher risk of residual aortic regurgitation, and are more likely to need implantation of a second valve.

■ PARAVALVULAR LEAK

Paravalvular leak, arguably an independent marker of higher morbidity and mortality risk after TAVR, is more common in patients with bicuspid aortic valve undergoing TAVR than in those with tricuspid aortic valve. Earlier studies reported rates of moderate or severe paravalvular leak between 16% and 32%.^{6,7}

The newer-generation balloon-expandable Sapien 3 valve (Edwards Lifesciences, Irvine, CA) is associated with a lower incidence

Though younger, patients with bicuspid aortic valve face higher risks during TAVR

of moderate or severe paravalvular leak than earlier devices, mainly attributable to its outer skirt, which allows more complete sealing.⁸ There are also reports of successful treatment of bicuspid aortic valve stenosis using the Lotus device (Boston Scientific, Marlborough, MA).⁹ This device features adaptive sealing along with retrievability and repositioning ability, which may facilitate optimal positioning and prevent paravalvular leak.

■ SIZING OF THE PROSTHESIS

Sizing of the prosthesis in patients with bicuspid aortic valve stenosis remains a challenge: some experts advocate the usual practice of measuring the perimeter and area at the level of the annulus, while others advocate measuring at the level of the commissures, 4 to 8 mm above the annulus. Balloon valvuloplasty may be a useful sizing tool, though it carries the hazards of severe aortic regurgitation and periprocedural stroke.

Angiography of the ascending aorta during balloon valvuloplasty can help verify whether an adequate seal is achievable and aid in selecting an appropriately sized prosthesis. Liu et al¹⁰ performed sequential balloon aortic valvuloplasty before TAVR with a self-expanding valve in 12 patients. Of these, 11 (91.7%) received a smaller device than they would have with multidetector computed tomography-guided annulus measurement.

Given that a larger valve may increase the risk of annular rupture, implantation of a smaller valve is always reasonable in bicuspid aortic valve as long as it achieves appropriate sealing with no paravalvular leak.

■ THE NEED FOR A PACEMAKER

After undergoing TAVR, more patients who have a bicuspid aortic valve need a permanent pacemaker than those who have a tricuspid aortic valve. This group appears to be more vulnerable to conduction abnormalities after TAVR, and rates of new pacemaker implantation as high as 25% have been reported with the newer-generation devices. Perlman et al⁸ observed that even when the Sapien 3 valve was implanted high in the left ventricular outflow tract, nearly 10% of patients needed a new pacemaker.

This is an important issue, as most patients with bicuspid aortic valve with severe aortic stenosis are relatively young and may endure deleterious effects from long-term pacing.

■ LONG-TERM OUTCOMES

The data on long-term outcomes of patients with bicuspid aortic valve who undergo TAVR are limited, and the available studies were small, with relatively short-term follow-up. However, Yoon et al compared TAVR outcomes between bicuspid and tricuspid aortic stenosis patients using propensity-score matching and demonstrated comparable all-cause mortality rates at 2 years (17.2% vs 19.4%, $P = .28$).⁶

Given the relatively long life expectancy of patients with bicuspid aortic valve undergoing TAVR, who tend to be younger than those with tricuspid aortic valve stenosis, longer-term data are needed to draw meaningful conclusions about the durability of transcatheter valves in this population. The bicuspid aortic valve is asymmetric, so that during TAVR the stent may not expand completely, and this in theory may result in more strain on the prosthesis and accelerate its structural deterioration.

In a recent meta-analysis, Reddy et al¹¹ analyzed 13 observational studies in 758 patients with severe bicuspid aortic valve stenosis undergoing TAVR with older and newer devices. The mean Society of Thoracic Surgeons Predicted Risk of Mortality score, which predicts the risk of death within 30 days, was 5.0%, but the actual rate was 3.7% (95% confidence interval [CI] 2.1%–5.6%). A high procedural success rate of 95% (95% CI 90.2%–98.5%) was also noted, but the rates of new permanent pacemaker implantation (17.9%, 95% CI 14.2%–22%) and severe perivalvular leak (12.2%, 95% CI 3.1%–24.8%) were somewhat elevated.¹¹

■ NOT FOR ALL, BUT AN EMERGING, VIABLE OPTION

As implanted prostheses and TAVR techniques undergo continuous improvement and as the experience of operators and institutions advances, procedural outcomes will likely improve.

The available data suggest that TAVR with the newer devices, when performed by experienced hands, is a viable option across most

Outcomes will likely improve with experience and newer devices

TRANSCATHETER AORTIC VALVE REPLACEMENT

of the risk spectrum in patients with severe tricuspid aortic stenosis, including low-risk patients,¹² and selectively in patients with bicuspid aortic valve stenosis. However, for patients with bicuspid aortic valve with severe aortic

stenosis and associated aortopathy, surgery remains the standard of care.

More study is needed to identify patients with bicuspid aortic valve who can be safely and effectively treated with TAVR. ■

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