

Evolving treatment options for valve and aortic disease with bicuspid aortic valve

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Treatment decisions for patients with bicuspid aortic valves (BAVs) are unique for two principal reasons: (I) aortopathy with dilation of the proximal aorta is seen about 50% of cases, and frequently requires a combined repair; (II) patients are significantly younger than those undergoing treatment for acquired aortic valve disease, making the durability and lifestyle implications of treatment choices increasingly important (1). The traditional treatment for valve and aortic disease with BAV is composite or separate replacement of the valve and ascending aorta. While mechanical aortic valves are more durable, they require life-long anticoagulation. Bioprosthetic valves may be an even less attractive option for younger patients as primary valve failure has been reported in 26% of cases at 15 years (2). In this context, an alternative treatment approach with repair rather than replacement of the valve and aortic root, as proposed by Cosgrove *et al.* and later refined by Schäfers *et al.*, is very appealing (3,4).

Recently Schneider *et al.* reported excellent 10- and 15-year results for valve repair with root remodeling in a cohort of 357 younger patients with BAV and aortic root dilation (5). The majority of the patients included had both aortic aneurysm and significant aortic regurgitation. They discuss three key advances in their two decades of experience with this operation: (I) emphasis on effective cusp height, or the difference between the central free margins and the aortic insertion lines, being at least 8 mm after repair (6); (II) alteration of commissural orientation

with root replacement to create near symmetric tongues (approaching 180°) in appropriate cases (7); and (III) adding suture annuloplasty to root replacement in order to improve valvular competency (5). The common theme for all three of these surgical advances is the approximation of normal, symmetric aortic valve function. Schneider *et al.* report a cumulative incidence of reoperation of 22% at 15 years, which is better than data reported for bioprosthetic aortic valves (2).

While root dilation and aortic insufficiency are important manifestations of BAV disease, aortic stenosis and aneurysm of the mid-ascending aorta are more common. Data suggests that the majority of patients with BAV eventually develop aortic stenosis, and that dilation of the tubular ascending aorta is more than twice as common as isolated root dilation (8,9). Given the marked thickening, calcification and restriction of leaflet motion seen with BAV stenosis, the valve repair discussed by Schneider *et al.* is not a viable option for most cases of stenotic BAV. In fact, they caution that calcified plaques are a strong predictor for development of aortic stenosis, and that this should be factored into patient selection for their valve-sparing operation. So, what can be offered to patients with stenotic BAV aside from traditional surgical replacement?

One option that has gained ground in recent years is transcatheter aortic valve replacement (TAVR). It is now a more common procedure in Germany that surgical aortic valve replacement (SAVR), has a role for the treatment of

intermediate-risk patients, and has been used for stenotic BAV with good short- and intermediate-term results (10-13). The role of TAVR in low-risk populations is currently being investigated, and while early results have been promising, the approach remains controversial (14). The long-term results of TAVR are unknown, and questions of durability are particularly important for younger patients with BAV that will likely require more than one valve intervention. Having said this, TAVR in younger patients with stenotic BAV provides a strategy to improve quality of life by avoiding life-long anticoagulation and delaying the need for open surgery. Recent studies have shown that repeat TAVR is feasible and associated with favorable early and mid-term outcomes, suggesting that the transcatheter approach may be a durable option for younger patients with BAV in the future (15).

Another potential advantage of TAVR for stenotic BAV is more favorable systolic flow patterns in the ascending aorta. Bicuspid aortic valves produce distinctive helical blood flow patterns within the aorta, a finding that supports the importance of hemodynamics in the progressive aortopathy seen with BAV (16,17). While valve replacement improves transvalvular gradients and relieves patient symptoms, studies using advanced three-dimensional MRI blood flow imaging (“4D Flow”) have shown that ascending aortic flow patterns remain markedly abnormal despite aortic valve replacement (18). Furthermore, when comparing SAVR and TAVR techniques, the degree of helical and vertical flow in the ascending aorta is less with TAVR, despite both prostheses being constructed around a stented frame. The underlying cause of these differences remains unclear, but considering that eccentric flow has been proposed as a driver of ascending aortic growth, these findings suggest that TAVR may reduce rates of progressive BAV aortopathy via reduced flow abnormality (19). If future studies demonstrate long-term valve durability and function to be equal between SAVR and TAVR, the technique that minimizes abnormal blood flow in the ascending aorta may be best suited for a younger BAV population that is subject to high rates of ascending aortic disease. Similarly, among patients with insufficient BAVs for whom valve repair and root remodeling are undertaken, the techniques described by Schneider *et al.* may reduce the hemodynamic impact on the ascending aorta, given their emphasis on stabilizing the aortic root as well as increasing cusp and commissural symmetry.

Aortic valve repair and replacement techniques that improve hemodynamics in the ascending aorta may

decrease the need for future aortic repair among patients who undergo surgery for isolated valve disease. However, surgeons are often presented with the challenge of patients with dysfunctional BAV and a mildly dilated ascending aorta. Standard surgical thresholds for determining the need for concomitant aortic repair rely on diameter, but have shifted considerably over the last two decades (20). The aggressive threshold of 42–45 mm used by Schneider *et al.* for concomitant aortic root remodeling in patients with regurgitant BAV is supported by the excellent stability they report. For stenotic BAV, however, pre-operative flow imaging may help inform this decision in borderline cases by better revealing the hemodynamic stress placed on the aortic wall.

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Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

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