Bicuspid aortic valve–associated aortopathy: A slowly evolving picture

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The bicuspid aortic valve (BAV)—until a few decades ago considered a rather harmless anomaly affecting up to 2% of the general population—has been progressively recognized as a heterogeneous entity associated with a variety of severe aortic wall pathologies. The relationship between BAV and associated aortopathy is complex and has been the focus of numerous articles. In the current issue of the *Journal*, Wojnarski and colleagues¹ have shed more light on this topic by using machine-learning classification of computed tomography scans in patients with BAV undergoing thoracic aortic surgery. The authors identified 3 phenotypes of BAV aortopathy (ie, root, ascending, and arch) that are associated with various morphologic and clinical characteristics.

Several studies have focused on BAV phenotypes and morphology to gain insights into the cause of BAV aortopathy and to better define the risk of future aortic complications. Advanced imaging technologies, in particular 4-dimensional magnetic resonance imaging, have been used to support the hypothesis that altered blood flow architecture through the BAV triggers structural aortic wall pathology by increasing local wall stress in certain subgroups of patients. Studies focusing on functional BAV status and morphology have revealed several important findings, including the following.

1. Patients with BAV experience acute aortic dissection at a younger age and a larger diameter than their tricuspid peers.² In addition, patients with BAV with dissection present more frequently with aortic insufficiency and an entry tear in the aortic root.² This observation supports earlier and more aggressive intervention for the patient with “root phenotype” BAV.
2. BAV functional phenotype (ie, BAV stenosis vs aortic insufficiency) correlates significantly with the expression and severity of bicuspid aortopathy. Mild-to-moderate ascending aortic dilatation remains stable after aortic valve replacement in patients with BAV aortic stenosis.^{3,4} In contrast, patients with aortic root dilatation and a regurgitant valve (ie, root phenotype) are at

Four-dimensional magnetic resonance imaging of blood flow distal to a BAV.

Central Message

A slowly evolving picture of the various phenotypic expressions of BAV-associated aortopathy is emerging.

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significant risk for aortic events after isolated aortic valve replacement.^{3,4}

3. Aortic root replacement in young patients with BAV, particularly those with root phenotype, may cure BAV-associated aortic disease and achieve excellent long-term outcomes.⁵

Wojnarski and colleagues,¹ in the largest published cohort to date of patients with BAV with associated aortopathy, support these findings and further elucidate the heterogeneity of BAV aneurysm formation. They identified 3 distinct phenotype clusters of aortic dilation in patients with predominately dysfunctional aortic valves. The results supply more evidence supporting close follow-up of young male patients with regurgitant BAVs, who are more likely to have a genetic component that triggers structural aortic wall weakness. These patients are prone to develop proximal aortic root phenotypes at a young age and are likely to benefit from a more aggressive surgical strategy.

Whether individual phenotype-based risk stratification in patients with normal aortic valvular function is of prognostic value requires further evaluation and cannot be answered by the current report. In addition, further studies are required to determine whether the identified phenotypes are associated with distinct structural wall alterations. Such a finding would add further evidence to the theory that different etiologic mechanisms are involved in the various BAV phenotypes. The story of

BAV-associated aortopathy is complex, but the picture is gradually becoming more clear.

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