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CORRELATION BETWEEN BICUSPID AORTIC VALVE STENOSIS AND ASCENDING AORTIC ANEURYSMS: A SHORT REVIEW

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ABSTRACT
Bicuspid aortic valve (BAV) is the most common congenital cardiac valve abnormality and is frequently associated with progressive aortic valve stenosis and ascending aortic aneurysm. The coexistence of these conditions represents a significant clinical challenge because both altered hemodynamic forces and intrinsic abnormalities of the aortic wall contribute to disease progression. This mini-review summarizes the current understanding of the relationship between bicuspid aortic valve stenosis and ascending aortic aneurysms, focusing on epidemiology, pathophysiological mechanisms, clinical presentation, diagnostic evaluation, and contemporary management strategies. Evidence suggests that abnormal extracellular matrix remodeling, genetic susceptibility, and increased wall shear stress generated by the bicuspid valve all play important roles in the development of ascending aortic dilation. Although the severity of aortic stenosis does not consistently correlate with aneurysm size, patients with BAV remain at increased risk of progressive aortic enlargement and, less commonly, aortic dissection. Echocardiography, computed tomography, and magnetic resonance imaging are essential for diagnosis and long-term surveillance. Current management emphasizes individualized risk assessment, optimal timing of surgical intervention, and lifelong imaging follow-up. Continued advances in cardiovascular imaging and molecular research are expected to improve risk stratification and guide future therapeutic approaches for patients with bicuspid aortic valve disease.

KEYWORDS: Bicuspid aortic valve; Aortic stenosis; Ascending aortic aneurysm; Bicuspid aortopathy; Thoracic aortic aneurysm; Aortic dilation; Hemodynamics; Valve disease; Aortic dissection; Echocardiography.

INTRODUCTION

Bicuspid aortic valve (BAV) is the most common congenital cardiac valve abnormality, affecting approximately 0.5–2% of the general population. It is characterized by the presence of two functional leaflets instead of the normal three and demonstrates a marked male predominance. Although many individuals remain asymptomatic during early life, BAV is associated with several cardiovascular complications, including aortic valve stenosis, aortic regurgitation, infective endocarditis, and progressive dilation of the ascending aorta, which may eventually lead to aneurysm formation or aortic dissection (1,2).

The coexistence of bicuspid aortic valve stenosis and ascending aortic aneurysm has attracted considerable attention because of its implications for patient surveillance, surgical timing, and long-term prognosis. While valve degeneration contributes to hemodynamic abnormalities, intrinsic abnormalities of the aortic wall also appear to play a critical role in aneurysm development. This review summarizes the relationship between BAV stenosis and ascending aortic aneurysms, focusing on epidemiology, pathophysiology, clinical presentation, diagnostic evaluation, and current management strategies.

Epidemiology

Bicuspid aortic valve affects approximately 1–2% of the population and accounts for nearly half of all patients undergoing surgery for severe aortic stenosis before the age of 70 years (1). Ascending aortic dilation develops in approximately 30–60% of patients with BAV during their lifetime, although the prevalence varies according to age, valve phenotype, and imaging modality used (3).

Compared with individuals with tricuspid aortic valves, patients with BAV have a significantly increased lifetime risk of developing thoracic aortic aneurysms and acute aortic dissection. However, the absolute incidence of dissection remains relatively low, particularly among patients who undergo regular surveillance and timely surgical intervention (4).

The coexistence of severe aortic stenosis and ascending aortic aneurysm is clinically important because both conditions frequently require simultaneous surgical correction, reducing the need for repeat operations and minimizing future cardiovascular risk.

Pathophysiological Mechanisms

The relationship between bicuspid aortic valve stenosis and ascending aortic aneurysm is multifactorial, involving both genetic predisposition and altered hemodynamic forces.

Genetic and Structural Abnormalities

Numerous studies suggest that BAV is not merely a valvular abnormality but rather part of a generalized connective tissue disorder affecting the entire aortic root and ascending aorta. Histopathological examination frequently demonstrates cystic medial degeneration, fragmentation of elastic fibers, increased collagen remodeling, smooth muscle cell apoptosis, and abnormalities in extracellular matrix organization (5).

Abnormal expression of matrix metalloproteinases (MMP-2 and MMP-9) contributes to degradation of elastin and collagen within the aortic wall, promoting progressive weakening and dilation (6). Several genetic pathways have also been implicated, including mutations involving the NOTCH1 gene and alterations in transforming growth factor-beta (TGF-β) signaling, although the genetic basis remains incompletely understood (7).

Hemodynamic Factors

In addition to intrinsic aortopathy, abnormal blood flow generated by the bicuspid valve plays an important role. Fusion of the valve cusps creates eccentric systolic flow jets that increase wall shear stress within specific regions of the ascending aorta (8).

Four-dimensional flow magnetic resonance imaging has demonstrated that patients with BAV exhibit abnormal flow patterns even before significant valvular dysfunction develops. These abnormal flow characteristics correlate closely with the location and severity of aortic dilation (9).

When progressive calcification results in aortic stenosis, the increased transvalvular velocity further augments wall shear stress, potentially accelerating aneurysm formation. Therefore, both congenital weakness of the aortic media and abnormal hemodynamic stress contribute synergistically to disease progression.

Clinical Correlation Between Aortic Stenosis and Ascending Aortic Aneurysm

Calcific degeneration develops earlier in bicuspid valves than in tricuspid valves because abnormal leaflet mechanics increase mechanical stress throughout life. Progressive fibrosis and calcification eventually produce clinically significant aortic stenosis, characterized by obstruction of left ventricular outflow (10).

At the same time, many patients develop progressive enlargement of the ascending aorta. Importantly, the severity of valve stenosis does not always correlate with the degree of aortic dilation. Some individuals with only mild valvular dysfunction develop marked aneurysmal enlargement, whereas others with severe stenosis exhibit relatively normal aortic dimensions (3).

Different BAV morphologies may influence the pattern of aortic enlargement. Right-left coronary cusp fusion is more commonly associated with dilation of the ascending tubular aorta, whereas right-noncoronary fusion has been linked to enlargement of the aortic root in some studies (11).

Clinically, patients may remain asymptomatic for years. Symptoms usually reflect progressive aortic stenosis and include exertional dyspnea, angina, syncope, fatigue, and reduced exercise tolerance. Ascending aortic aneurysms themselves are often silent until they become large or complicated by dissection or rupture.

Diagnostic Evaluation

Echocardiography remains the first-line imaging modality for evaluating BAV morphology, determining the severity of aortic stenosis, and measuring proximal aortic dimensions (12). Doppler echocardiography provides accurate assessment of transvalvular gradients and valve area, facilitating disease staging.

Computed tomography (CT) angiography and magnetic resonance imaging (MRI) offer more accurate assessment of the entire thoracic aorta, particularly when the ascending aorta cannot be adequately visualized by echocardiography. MRI additionally permits evaluation of blood flow patterns through four-dimensional flow imaging, improving understanding of regional wall shear stress (9).

Current guidelines recommend serial imaging of the ascending aorta in all patients with BAV because aneurysm progression varies considerably among individuals. Surveillance intervals depend on baseline aortic diameter and the rate of enlargement (12,13).

Management Strategies

Management aims to treat both valvular dysfunction and progressive aortic disease while preventing catastrophic complications such as dissection.

Medical Management

No medical therapy has been conclusively shown to prevent aneurysm progression in BAV-associated aortopathy. Nevertheless, aggressive control of hypertension is recommended because elevated blood pressure increases wall stress. Beta-blockers and angiotensin receptor blockers are commonly prescribed, although evidence supporting their superiority remains limited (13).

Patients should also receive comprehensive cardiovascular risk reduction, including smoking cessation, lipid management, weight control, and regular follow-up. Participation in high-intensity isometric exercise may be restricted in patients with significant aortic dilation.

Surgical Treatment

Surgical aortic valve replacement remains the definitive treatment for severe symptomatic bicuspid aortic valve stenosis. Depending on patient age and anatomy, either surgical valve replacement or transcatheter approaches may be considered, although transcatheter aortic valve implantation (TAVI) presents unique technical challenges in bicuspid anatomy (14).

When ascending aortic aneurysm coexists, replacement of the ascending aorta is frequently performed during valve surgery. Current international guidelines generally recommend concomitant ascending aortic replacement when the diameter reaches 45 mm in patients already undergoing aortic valve surgery. In isolated aneurysm disease without an indication for valve replacement, surgery is usually considered at diameters of 50–55 mm depending on individual risk factors, family history, growth rate, and associated conditions (12,13).
Combined procedures have demonstrated excellent long-term outcomes and substantially reduce the future risk of dissection or reoperation.

Prognosis
The prognosis of patients with BAV has improved substantially owing to advances in imaging, surveillance protocols, and surgical techniques. Most patients who undergo timely intervention experience excellent long-term survival comparable to that of the general population.
However, lifelong surveillance remains essential because residual segments of the aorta may continue to enlarge even after valve replacement. Aortic complications have been reported years after isolated valve surgery, emphasizing that correction of valvular stenosis does not eliminate the underlying aortopathy (15).
Individual risk stratification based on valve morphology, genetic background, family history, aortic growth rate, and imaging findings is becoming increasingly important for optimizing long-term management.

Future Perspectives
Recent research is focused on identifying biomarkers capable of predicting aneurysm progression before significant dilation occurs. Advances in molecular genetics may improve identification of high-risk patients who require closer surveillance or earlier intervention.
Four-dimensional flow MRI has emerged as a promising technique for quantifying abnormal wall shear stress and may eventually become part of routine clinical assessment. Artificial intelligence-based imaging analysis may further enhance individualized prediction of aneurysm growth and surgical timing.
Continued investigation into the molecular pathways responsible for BAV-associated aortopathy may also identify novel pharmacological targets capable of slowing aneurysm progression, an area where effective medical therapies remain lacking.

CONCLUSION
Bicuspid aortic valve stenosis and ascending aortic aneurysm represent closely related manifestations of a complex cardiovascular disorder involving both congenital abnormalities of the aortic wall and altered hemodynamic forces. Although progressive calcific stenosis increases mechanical stress on the ascending aorta, intrinsic abnormalities of connective tissue significantly contribute to aneurysm formation independently of valve dysfunction. Comprehensive imaging surveillance is therefore essential in all patients with bicuspid aortic valves, regardless of stenosis severity. Early recognition, individualized risk assessment, and timely surgical intervention have markedly improved patient outcomes while reducing the incidence of life-threatening complications such as aortic dissection. Ongoing advances in molecular biology and cardiovascular imaging are expected to further refine management strategies and improve long-term prognosis.

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