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Bicuspid Aortic Valve Disease – Evolving Concepts with Clinical Relevance

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The congenital anomaly, bicuspid aortic valve (BAV), was mentioned only briefly during my medical school and residency training. I was taught later, during cardiology fellowship, that this abnormality was usually not thought to be problematic during early adult life but might become stenotic or regurgitant in middle age or later. I was not informed about abnormalities in the ascending aorta although it was frequently pointed out that patients with coarctation of the aorta invariably had a bicuspid aortic valve. Since that time, a great deal of clinical, pathological, and genetic information has been reported concerning this entity and its associated ascending aortic pathology.[1-3]

Bicuspid aortic valve is the commonest congenital cardiac defect in the United States occurring in 1-2% of live births. It is now known that as many as 50% of patients with bicuspid aortic valve have associated ascending aortic pathology including aneurysm and a propensity to develop dissection. Pathological study of resected ascending aortic tissue from these patients has demonstrated degenerative changes that are either primary to the syndrome or secondary to abnormal hemodynamic flow patterns resulting from the deformed aortic valve.[4-6] This syndrome is no longer referred to as bicuspid aortic valve but rather bicuspid aortopathy because of the associated ascending aortic abnormalities.[1]

Regardless of the exact etiology of the changes in the aorta, these alterations can lead to dilation of the aorta with subsequent aneurysm formation. The weakened aortic wall structure

also places these individuals at increased risk for potentially fatal aortic dissection. In middle life (40-60 years of age) bicuspid aortic valves can become stenotic or regurgitant and often require aortic valve replacement possibly combined with an ascending aortic graft. Moreover, Wedin and co-workers observed that bicuspid aortic valve patients with aortic stenosis who underwent aortic valve replacement had decreased postoperative left ventricular function and an increased risk for heart failure hospitalization compared with non-BAV patients with aortic stenosis.[7] Patients with bicuspid aortic valve are also at increased risk to develop infective endocarditis on the bicuspid aortic valve.

A bicuspid aortic valve can often be diagnosed during a careful cardiac examination when a prominent early systolic click is followed by a short to medium length aortic flow murmur. Confirmation of the diagnosis is usually made by transthoracic echocardiography although in some patients with challenging echocardiographic windows, CT or MRI may be needed to confirm the diagnosis.

There is no specific therapy advised for patients with bicuspid aortic valve. In the recent past, prophylactic antibiotic administration was advised for dental procedures including tooth cleaning as well as other operative interventions. This is no longer recommended unless the bicuspid aortic valve patient has had an earlier episode of infective endocarditis. Patients with bicuspid aortic valve who require valve replacement because of stenosis or regurgitation usually require a surgical approach. The transcutaneous catheter valve replacement so popular with elderly patients and calcific aortic stenosis can be challenging in bicuspid aortic valve patients because of the distorted anatomy associated with this condition.

The take home message for physicians caring for a patient with a bicuspid aortic valve is as follows:

1. Careful dental prophylaxis is important to reduce the chance for infective endocarditis involving the bicuspid aortic valve.
2. Prophylactic antibiotics for dental work or other surgery are not indicated unless the patient has had a previous episode of infective endocarditis.
3. The presence of systemic hypertension increases the risk for aneurysm formation and progressive dilation. Therefore, systemic hypertension should be aggressively treated.
4. An initial transthoracic echocardiogram should be obtained to assess valve morphology and any degree of stenosis or regurgitation.
5. Repeat echo studies should be done yearly if more than mild aortic stenosis, regurgitation, or ascending aortic pathology is noted.
6. An initial thorax CT scan or MRI should be performed to assess possible ascending aortic abnormality. If dilation of the ascending aorta is noted, then regular CT scans/MRIs should be repeated at 3-5 year intervals. Once ascending aortic diameter exceeds 4.0 cm, CT/MRI should be repeated yearly.

7. Cardiology and thoracic surgical consultation should be obtained if progressive ascending aortic dilation is observed. Aneurysms equal to or exceeding 4.5-5 cm diameter should be considered for graft replacement. Grafts with a prosthetic aortic valve are usually chosen for this surgical intervention.
8. In general, strenuous isometric exercise should be avoided in bicuspid aortic valve patients because of the marked increase in arterial blood pressure with such exercise. Repeated episodes of markedly increased blood pressure could contribute to accelerated valve or aortic degenerative changes.

I hope that this review has been helpful for clinicians who follow these patients. As always, I enjoy hearing from readers at jalpert@arizona.edu.

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