FIVE THINGS TO KNOW ABOUT ...

Bicuspid aortic valve disease

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1 Bicuspid aortic valve disease is the most common congenital heart disease in developed nations

Bicuspid aortic valves are present in 1%–2% of the population, with men being 3 times more commonly affected than women. This disease is more than just anatomic variation and may be associated with other heart and aorta abnormalities as a result of molecular and connective tissue derangements.[1]

2 Bicuspid aortic valves are heritable

Although the exact cause is unknown, familial clustering of bicuspid aortic valves and associated aortopathy has revealed genetic components. Clinical and echocardiographic screening of first-degree relatives of affected patients is warranted to rule out bicuspid aortic valves and thoracic aortic disease.[2]

3 Valvular and aortic complications are common among patients with bicuspid aortic valves

The clinical presentation is heterogeneous. Many patients with bicuspid aortic valves are asymptomatic and have minimal valvular dysfunction. However, with increasing age, aortic stenosis (Figure 1) and insufficiency (Appendix 1, available at www.cmaj.ca/lookup/suppl/doi:10.1503/cmaj.121875/-/DC1) are common, with over one-third of initially asymptomatic patients experiencing a major cardiovascular event or requiring cardiac surgery over 20 years.[3] An estimated 15%–45% of patients may also have ascending aortic aneurysms;[3,4] however, the risk of aortic dissection is relatively low (1%–6%).[3–5]

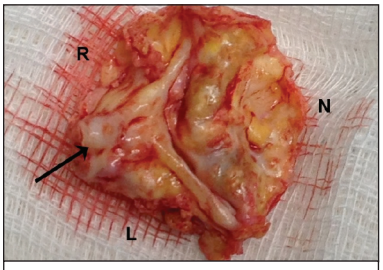

Figure 1: Severely calcified bicuspid aortic valve showing the most common pattern in bicuspid aortic valve disease: fusion of the left (L) and right (R) cusps with a prominent calcified raphe (arrow), opposed to a calcified noncoronary cusp (N).

4 Lifelong clinical follow-up and serial echocardiography are required

Patients with aortic stenosis and a mean/peak gradient of more than 30/50 mm Hg or aortic root diameter of more than 40 mm should have annual follow-up. For those with less severe asymptomatic disease, clinical follow-up may be performed every 3 years.[2]

5 Aortic valve surgery is needed after symptoms develop (angina, dyspnea, syncope)

Other indications for surgery include left ventricular dysfunction and concomitant need for heart surgery in patients with severe aortic stenosis or insufficiency. Surgical outcomes are excellent, and most patients return to a normal quality of life. Surgery for an associated aneurysm is recommended if the diameter of the ascending aorta is more than 50 mm or if there is more dilation than 5 mm/yr, regardless of symptoms.[2]

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