Bicuspid aortic valve aortopathy: One size fits all?

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Central Message

Individualized decision making is advisable in determining treatment for patients with bicuspid aortic valve aortopathy.

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It well documented that a relevant proportion of patients with a bicuspid aortic valve (BAV) will have development of aneurysmal dilatation of the ascending aorta.¹ It is increasingly recognized that this aortopathy not only varies in its extent or phenotype² but also includes a spectrum of clinical scenarios, including different forms of aortic valve dysfunction.

Older guidelines recommended an aortic diameter threshold of 5 cm as trigger for surgical therapy in patients with BAV.³ In the past 10 years, it has become more apparent that the risk of aortic dissection is actually lower than previously thought for the majority of patients with BAV,⁴ and guidelines have consequently been modified. The threshold for surgery has been increased to 5.5 cm, and only in the presence of risk factors is the previous threshold of 5 cm maintained.⁵

Nonetheless, actual clinical decision making has not always followed current guidelines,⁶ and it is worth reconsidering the background of the guidelines to improve counseling of patients and surgical decision making. In this context, the review by Braverman⁷ in this issue of the *Journal* provides valuable information that should be read and considered carefully.

Braverman⁷ points out that the prognosis of aortic aneurysm in the presence of a BAV is not uniformly good. In fact, young patients with a root aneurysm of 5 cm are at an increased risk for development of dissection or for size increase of the aneurysm in the first 10 years after diagnosis. Equally important is the type of aortic valve dysfunction, with aortic regurgitation exposing the patient to a significantly higher chance of progressive dilatation than that seen with aortic stenosis.

As common sense indicates, patient size should also be considered, even though indexing aortic size to body surface area has not yet found entry into guidelines. Although z scores are accepted criteria in pediatric cardiac surgery, the adult cardiology and cardiac surgery fields have been reluctant to use them in the context of aneurysm, an omission that should be reconsidered. Finally, age and comorbidity of the patient must be taken into consideration,

because they have an effect on life expectancy as well as on the morbidity and mortality of the operation.⁸ The extent of surgery should be carefully planned. Aneurysm of the tubular aorta does not necessarily warrant aggressive root replacement, because in the vast majority of patients, the root will remain stable for a long time.⁹

The key message that treatment should be carefully considered on an individual basis is thus a very important one. Although an aortic diameter of more than 5.5 cm clearly triggers surgery adequately in many cases (always considering the anticipated risk of the procedure related to the patient's risk profile and experience of the surgeon), patients with lower aortic diameters should not automatically be followed up with observation alone but rather should be evaluated on an individualized basis. A high probability of valve preservation at the time of surgery may play an additional role in this process.¹⁰

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