Guidelines for management of bicuspid aortic valve aneurysms: what’s the clinician to do?

Alan C. Braverman

The timing of prophylactic ascending aortic aneurysm surgery in the setting of bicuspid aortic valve disease is complex, with multiple factors influencing the decision. The 2014 ACC/AHA Valve guidelines recommend prophylactic replacement of the aortic root and/or ascending aorta once the aortic diameter exceeds 5.5 cm. This aortic size threshold for surgery is at a larger diameter than had been recommended by the 2010 Thoracic Aortic Disease guidelines, the 2013 Society of Thoracic Surgeons Clinical Practice Guidelines, or the 2006 ACC/AHA Valve guidelines. Five recent societies or committees recently published their guidelines to assist with managing these cases. Making the decision regarding the timing of bicuspid aortic valve aneurysm surgery even more difficult are the small, but important, differences in recommendations provided among recent guidelines addressing this issue.

Keywords
aortic aneurysm, bicuspid aortic valve, guidelines

The 2014 ACC/AHA Valve Disease guidelines have given a class I indication (level of evidence C) for prophylactic replacement of the aortic root and/or ascending aorta in adults with bicuspid aortic valve disease when the aortic dimension exceeds 5.5 cm [1]. Surgery is suggested (class IIa) to replace the aortic root and/or ascending aorta when the aorta is more than 5.0 cm if there are risk factors for aortic dissection, such as a family history of aortic dissection or rapid growth of the aorta (>0.5 cm/year). These recommendations differ from those reported in the 2010 Thoracic Aortic Disease guidelines (TAD), which recommended that patients with Marfan syndrome or other genetically mediated disorders (vascular Ehlers–Danlos syndrome, Turner syndrome, bicuspid aortic valve, or familial thoracic aortic aneurysm and dissection) should undergo elective operation at smaller diameters (4.0 to 5.0 cm depending on the condition) [2]. For many of us caring for patients with bicuspid aortic valve aneurysm disease, the 2010 Thoracic Aortic Disease guideline translated into recommending prophylactic aortic surgery for appropriate candidates once the aorta reached or exceeded 5.0 cm, the size threshold also recommended in the 2006 ACC/AHA Valve guidelines [3].

There are now five recent documents carefully produced by experienced, thoughtful, and seasoned clinicians and surgeons that provide differing recommendations about the timing of prophylactic root and/or ascending aortic surgery in the setting of bicuspid aortic aneurysm disease (Table 1) [1,2,4–6]. These recommendations are for aneurysm surgery alone (i.e., when the aortic valve does not require valve surgery).

The timing of prophylactic aortic root or ascending aortic replacement in the setting of bicuspid aortic valve disease is complex. The risk of aortic events (such as dissection and rupture) is not predicted by aortic diameter alone. Issues to consider include the absolute size of the aorta, condition of the bicuspid aortic valve, and rate of expansion of the aortic aneurysm. The patient’s age, sex, body surface area, family history, underlying condition, and general health and operative risk are also important to consider [2,5–7]. The pattern of aortic dilatation may also be of prognostic significance, with the aortic root phenotype associated with more rapid growth [8]. Abnormal aortic flow hemodynamics, related to helical flow patterns, bicuspid aortic valve leaflet orientation, and subsequent aortic wall stress, may also predict risk [9]. The presence of coexistent aortic stenosis or...
regurgitation also correlates with aortic wall medial abnormalities [10]. Writing succinct guidelines that encompass all of the many variables that influence the decision for timing of aortic surgery in bicuspid aortic valve aortopathy is very difficult. One would not expect any guideline to be able to envision all circumstances.

The risk of aortic surgery must be weighed against the risk of subsequent aortic complications, such as aortic dissection and rupture. Are the short-term and long-term risk of an aortic event for the 40-year old with a bicuspid aortic valve and a 5 cm aneurysm with the ‘aortic root phenotype’ the same as the risk for a 70-year old with a 5 cm mid-ascending aortic aneurysm and mild bicuspid aortic valve sclerosis? There is very little information about the natural history of bicuspid aortic valve patients with aortic aneurysms more than 5 cm followed expectantly to provide insight into the absolute or yearly risk of aortic catastrophes, such as dissection or rupture. Although bicuspid aortic valve patients without aneurysms have a low absolute risk of acute aortic events, those with aneurysms have a much worse fate. Among patients with ascending aortic aneurysms more than 45 mm, the incidence of acute aortic dissection was 45 cases per 10 000 person-years [11]. The cumulative risk of aortic dissection or rupture was 8.6% among bicuspid aortic valve patients with aortic aneurysm followed in the Yale Center for Thoracic Aortic Disease [12]. In this series, 90% of patients had an initial aortic size less than 54 mm and 67% of patients had a final aortic aneurysm diameter of less than 54 mm [12].

I do not believe ‘one size fits all’ when it comes to decision making about the timing of aortic aneurysm surgery. One has to individualize management based on each patient’s characteristics. Certain patients will be at higher risk for aortic dissection at a given aortic size, yet we do not have all the clinical tools to identify these patients well. Other patients will be at higher risk for prophylactic surgery. The Canadian Cardiovascular Society recommendations provide a range of aortic dimension (50–55 mm) for which to proceed with prophylactic aortic root replacement in the presence of a bicuspid aortic valve [6]. They further state that patient characteristics such as rapid aortic growth, concomitant valve disease, genetic syndromes, or connective tissue disorders trigger the decision for surgery at the smaller size, as long as this procedure is done by an experienced surgeon and team leading to a very low mortality rate [6]. The expertise and experience of the cardiac surgeon are very important. Most cardiac surgeons perform very few aortic root and/or ascending aortic replacements, and a 1% mortality rate will likely be achieved in only the most experienced centers.

However, I am concerned that for some bicuspid aortic valve patients the strategy of observing the dilated aorta (>5 cm) while waiting for the diameter to exceed 5.5 cm before recommending surgery will be associated with an unacceptable risk of aortic dissection. Although the 2014 ACC/AHA Valve guidelines do suggest surgery at more than 5.0 cm if the aorta grows very rapidly (>0.5 cm/year) or if there is a family history of aortic dissection, the guidelines do not address other important variables, notably age, small body size, and gender. It is likely that ongoing research will better delineate patients with aortic aneurysm at increased risk of aortic dissection. In discussions with cardiologists and surgeons who have expertise in managing the aortopathy of bicuspid aortic valve disease, it is my impression that they will use many factors other than size alone to inform the decision as to when to go forward with aortic surgery. We all should!
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REFERENCES