Knowledge, attitudes, and practice patterns in surgical management of bicuspid aortopathy: A survey of 100 cardiac surgeons

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Objective: Clinical practice guidelines have been established for surgical management of the aorta in bicuspid aortic valve disease. We hypothesized that surgeons’ knowledge of and attitudes toward bicuspid aortic valve aortopathy influence their surgical approaches.

Methods: We surveyed cardiac surgeons to probe the knowledge of, attitudes toward, and surgical management of bicuspid aortopathy. A total of 100 Canadian adult cardiac surgeons participated.

Results: Fifty-two percent of surgeons believed that the mechanism underlying aortic dilation in those with bicuspid aortic valve was due to an inherent genetic abnormality of the aorta, whereas only 2% believed that altered valve-related processes were involved in this process. Only a minority (15%) believed that bicuspid valve leaflet fusion type is associated with a unique pattern of aortic dilatation aortic phenotype. Sixty-five percent of surgeons recommended echocardiographic screening of first-degree relatives of patients with bicuspid aortic valve. Most surgeons (61%) elected to replace the aorta when the diameter is 45 mm or greater at the time of valve surgery. Fifty-five percent of surgeons surveyed suggested that in the absence of concomitant valvular disease, they would recommend ascending aortic replacement at a threshold of 50 mm or greater. Approximately one third of surgeons suggested that they would elect to replace a mildly dilated ascending aorta (40 mm) at the time of valve surgery. The most common surgical approach (61%) for combined valve and aortic surgery was aortic valve replacement and supracoronary replacement of the ascending aorta, and only a minority suggested the use of deep hypothermic circulatory arrest and open distal anastomosis. More aggressive approaches were favored with greater surgeon experience, and when circulatory arrest was chosen, the majority (68%) suggested they would use antegrade cerebral perfusion. In the setting of aortic insufficiency and a dilated aorta, 42% of surgeons suggested that they would perform valve-sparing surgery. Of note, 40% of respondents used an index measure of aortic size to body surface area in addition to absolute aortic diameter in assessing the threshold for intervention.

Conclusions: This large survey uncovered significant gaps in the knowledge and attitudes of surgeons toward the diagnosis and management of bicuspid aortopathy, many of which were at odds with current guideline recommendations. Efforts to promote knowledge translation in this area are strongly encouraged. (J Thorac Cardiovasc Surg 2013;146:1033-40)
recognized that 20% to 85% of patients with BAV also develop varying degrees of aortic dilatation (referred to as “bicuspid aortopathy”).

Three patterns of aortic dilatation have been suggested, including primary dilatation of the tubular/mid ascending aorta, involvement of the distal ascending aorta and proximal arch, and isolated aortic root dilatation, although no uniform classification scheme is endorsed.

Accumulating data emphasize that bicuspid aortopathy exhibits considerable heterogeneity with respect to molecular, rheological, and clinical characteristics. Controversy exists as to the mechanism through which bicuspid aortopathy develops. Proponents of the genetic theory argue that bicuspid aortopathy is due to an inherent molecular defect in the aorta and develops independently of valve function, valve morphology, or hemodynamics. On the other hand, emerging data now point toward a primary hemodynamic basis of bicuspid aortopathy. Interest in this has been fueled by recent data that suggest that even a normally functioning BAV can exhibit abnormal transvalvular flow patterns, resulting in regional increases in wall shear stress predicted by the morphologic fusion pattern of the valve. The type of fusion pattern may thus dictate the pattern of aortic dilatation, with a right-left leaflet fusion resulting in flow toward the right anterior wall (potentially resulting in isolated tubular ascending aortic dilatation) and the right nonleaflet fusion pattern resulting in a flow pattern toward the posterior aorta leading to arch dilatation.

Aortic aneurysm formation and aortic dissection are the 2 major complications of bicuspid aortopathy. Studies suggest that the rate of growth of ascending aortas is higher in patients with bicuspid versus tricuspid aortic valves. However, one of the most important determinants of aortic expansion is the baseline aortic dimensions (>40 mm). Although earlier reports suggested the incidence of aortic dissection to be as high as 5%, recent data suggest that the incidence is actually lower. In the Toronto series, a rate of 0.1% per patient year follow-up was noted, similar to other recent studies.

There are conflicting reports regarding the fate of the ascending aorta after AVR. Although earlier studies suggested that moderate aortic dilatation (45–49 mm) was a risk factor for aortic complications after AVR, more recent data from a homogenous population of patients with aortic stenosis and mild-moderate aortic dilatation (40–50 mm) revealed that isolated AVR was associated with excellent 10- and 15-year freedom from aortic complications with no aortic dissection occurring in this cohort during follow-up.

Guidelines for the management of patients with valvular heart disease were established in 2006 by the American College of Cardiology and American Heart Association (AHA), with a focused update in 2008 that includes recommendations for the surgical management of bicuspid aortopathy. Elective replacement of the ascending aorta or aortic root is recommended for patients with BAV when the aortic diameter is 50 mm or greater whether or not concomitant valvarul or coronary indications are present. However, the most recent European guidelines recommend that the aorta be replaced when the aortic root or ascending aortic diameter is 50 mm or greater in the presence of risk factors, including coarctation of the aorta, systemic hypertension, family history of dissection, or increase in aortic diameter of 2 mm or greater per year. Although the European guidelines suggest that in other circumstances, surgery should be delayed until aortic dimensions are 55 mm or greater, they are in agreement with the American guidelines that recommend a lower threshold (45 mm) in patients undergoing AVR.

Although bicuspid aortopathy is a commonly encountered clinical scenario by the cardiac surgeon, the heterogeneous nature of the disease can lead to a variation in the threshold for aortic replacement. Furthermore, there is no consensus recommendations as to the extent or type of surgery that such individuals should be subjected to, and no randomized clinical trials are available to better inform clinicians as to whether an aggressive versus conservative approach is appropriate. Because the perioperative risk of complications must be balanced by the actual risk of aortic complications, it is essential to understand the contemporary knowledge and practice patterns of cardiac surgeons toward bicuspid aortopathy in an effort to develop knowledge translation platforms for improved clinical care.

**MATERIALS AND METHODS**

A Bicuspid Aortopathy Working Group (S.V., B.Y., M.R., M.D.P., P.W.M.F.) was established and used to develop and validate a questionnaire to survey cardiac surgeon knowledge and surgical decision-making regarding the management of patients with bicuspid aortopathy. Institutional ethics approval was obtained from St Michael’s Hospital, University of Toronto, Toronto, Canada. All Canadian adult cardiac surgeons were invited in person or by e-mail to participate in the study. Participation was voluntary, and informed consent was presumed for all participants. We defined an experienced surgeon as one who performs more than 5 cases per year of complex aortic procedures, such as valve-sparing aortic root replacement, modified Bentall (composite graft), and open aortic arch replacement.

**RESULTS**

**Surgeon Demographics**

We contacted 142 of the approximately 150 staff adult cardiac surgeons across Canada. Of these, 8 actively refused...
to participate and 34 did not respond. Our invitation therefore yielded a response rate of 70% (100/142) with representation from all 10 Canadian provinces (Alberta, British Columbia, Manitoba, New Brunswick, Newfoundland and Labrador, Nova Scotia, Ontario, Prince Edward Island, Quebec, and Saskatchewan) (Figure E1). The response rates by region were western Canada (50%), central Canada (73%), and eastern Canada (75%). Our population also was balanced with respect to years in practice from 0 to 5 to more than 25 years (Figure E2). The median overall annual case volume was 150 to 200 cases in 50% of surgeons (50/100), 200 to 250 cases in 25% of surgeons (25/100), and 100 to 150 cases in 15% of surgeons (15/100). Eighty-eight percent of participants described their practice as academic, and 12% of participants described their practice as community-based.

Knowledge of Bicuspid Aortopathy

Recent experimental and translational studies have provided evidence for a role of both genetics and hemodynamics in the pathogenesis of aortic dilation in patients with BAV.32-37 Our questionnaire demonstrated that 52% of cardiac surgeons (52/100) believed that the underlying cause of bicuspid aortopathy was due to genetics, and 40% believed both genetics and hemodynamics play a role (Figure 1). Only 2% of surgeons (2/100) believed that hemodynamics alone could be responsible for BAV aortopathy and progressive aortic dilatation.

Eighty-three percent of surgeons identified the most common leaflet fusion type as left-right, 6% incorrectly stated that fusion of the right and noncoronary cusps is the most common, only 2% selected fusion of the left and noncoronary cusps, and 9% of surgeons admitted that they did not know the most common fusion pattern. Only a minority of cardiac surgeons (15% [15/98]) believed that leaflet fusion type is associated with a unique aortic phenotype, whereas the majority did not know the answer (58% [57/98]).

Fifty-nine percent of surgeons surveyed indicated that they believed the most common aortic phenotype encountered in practice was isolated dilation of the ascending aorta, whereas 30% of respondents indicated that the most common pattern involved combined aortic root and ascending aortic dilatation.

Preoperative Perspectives

Seventy-three percent of cardiac surgeons (73/100) were aware of current AHA/American College of Cardiology and ESC guidelines for the surgical management of BAV. Awareness of guidelines did not differ significantly between surgeons in academic compared with community practices or years in surgical practice (data not shown). In contrast to the recommendations provided within clinical practice guidelines, only 65% of cardiac surgeons (65/100) recommended echocardiographic screening of first-degree relatives of patients with BAV (Figure 2). Overall, 97% of cardiac surgeons (96/99) used preoperative computed tomography for determination of maximal aortic dimensions. We found that 42% of surgeons (42/100) indexed the maximal aortic dimension to body surface area, 21% of surgeons (21/100) indexed to the size of the descending aorta, 9% of surgeons (9/100) indexed to patient height, and 41% of surgeons (41/100) did not use any size indexing approach (responses were not mutually exclusive).

Surgical Management of the Bicuspid Aorta

A hypothetical 50-year-old male patient with isolated ascending aortic dilation and a normal functioning BAV was presented as a case example. Fifty-five percent of surgeons (54/99; Figure 3) reported an aortic diameter threshold for replacement at 50 mm for this patient. In contrast, 23% of surgeons (23/99) favored a more aggressive approach and recommended replacement of the aorta at 45 mm. As suggested by the recent European guidelines, only 19% of surgeons (19/99) responded that they would not replace the aorta unless 55 mm in size, and 2% of surgeons (2/99) recommended a threshold greater than 60 mm.
for this patient. When changing the patient scenario to include a stenotic BAV requiring AVR, 61% of surgeons (61/100; Figure 3) reported a threshold diameter for aortic replacement at 45 mm. In the setting of AVR, 29% of surgeons (29/100) indicated that they would replace the aorta at 40 mm and 2% of surgeons (2/100) indicated that they would replace the aorta at 35 mm. Despite the indication for AVR and at odds with current guidelines, 7% of surgeons (7/100) would not replace the aorta unless it was 50 mm or more, and 1% of surgeons (1/100) would not replace the aorta unless it was 55 mm or more. The nature of the surgeon’s practice as academic or community-based, the number of cases performed per year, or awareness of current guidelines did not influence the threshold for aortic replacement. Surgeons with fewer years in surgical practice tended toward size thresholds consistent with current guidelines (Figure 4), perhaps reflecting more recent training and education in this area.

We then presented a hypothetical 35-year-old patient with stenotic BAV, a 53-mm ascending aorta with normal aortic root and arch dimensions. We found that 61% of surgeons (60/99; Figure 5) recommended AVR with supracoronary replacement of the ascending aorta. Patient age did not significantly influence the decision to replace the aorta.

There was a cohort of surgeons who would perform AVR, replacement of ascending aorta and hemiarch (15% [15/99]), Bentall (12% [12/99]), or Bentall and hemiarch (7% [7/99]). When grouped, this cohort (34% [34/99]) performed more than 10 Bentall procedures (42% [14/33] vs 10% [6/64]) and more than 10 arch replacements with circulatory arrest (33% [10/33] vs 6% [4/64]) per year. In addition, surgeons in this cohort were more likely to be referred patients with a dilated root, ascending aorta, and arch (18% [6/33] vs 2% [1/64]) as the most common presentation. This cohort did not differ significantly from the remaining respondents with respect to years in practice, overall cases per year, academic or community setting, and aortic replacement thresholds. With respect to knowledge, more surgeons in this cohort believed that an association between leaflet fusion configuration and patterns of aortic dilation exists (24% [8/33] vs 11% [7/62]).

In the case of a patient with mild-moderate aortic insufficiency secondary to a BAV and an aortic root size of 50 mm with normal ascending and arch dimensions, 41% of
surgeons (41/100) indicated that they would perform a valve-sparing root replacement (David) procedure. Surgeons who perform complex aortic surgeries more frequently (>5 procedures per year) were more likely to perform valve-sparing procedures for aortic root dilatation compared with others (71% [12/17] vs 35% [29/81]; Figure E3). In regard to intraoperative strategy for hypothermic circulatory arrest to replace the arch/hemi-arch, 58% of surgeons would cool the patient to 18°C to 20°C, 28% of surgeons would cool the patient to 25°C, and 10% of surgeons would cool the patient to 28°C to 30°C. Also, 68% of surgeons would use selective antegrade perfusion for cerebral protection, 9% of surgeons would use selective retrograde, and 22% of surgeons would not use any type of cerebral perfusion.

DISCUSSION

In this report, we probed cardiac surgeon knowledge of, attitudes toward, and approaches to the surgical management of bicuspid aortopathy. Our data highlight a significant variability in practice patterns and disparity between guidelines and proposed strategies for bicuspid aortopathy. The majority of cardiac surgeons were knowledgeable of BAV aortopathy, leaflet fusion patterns, and predisposition to aortic dilatation. Few surgeons acknowledged an association between leaflet fusion configuration and pattern of aortic dilatation—an emerging theme in bicuspid aortopathy. Surgeons strongly favored genetics over hemodynamics as the underlying mechanism for bicuspid aortopathy. However, the relative contribution of genetics or an inherited aortopathy compared with acquired effects of altered blood flow and wall stresses from the abnormal valve remains unclear. Although theories supporting an inherited aortopathy have been popular, more recent data support a critical role of hemodynamic influences. BAV is a heterogeneous condition with different anatomic and clinical phenotypes. Phenotypic classification is somewhat challenged by the referral bias to surgeons and to aortic surgeons in particular, who may see more patients with arch or root dilatation. The emergence of specialized clinics for patients with BAV may be capable of registering patients with BAV before surgery and may shed light on phenotypes and their progression to requiring surgical interventions. Further research to correlate tissue changes with regional hemodynamic stresses may shed light on the cellular and molecular mechanisms of this elusive clinical condition. As reviewed by Verma and Fedak, elegant work by Ikonomidou and colleagues has demonstrated a unique fingerprint of bicuspid aortopathy correlating leaflet fusion morphology with the extent of aortic dilatation.

A significant number of cardiac surgeons do not support echocardiographic screening of first-degree relatives, which is indicated in the 2010 American College of Cardiology Foundation/AHA/American Association for Thoracic Surgery/American College of Radiology/American Stroke Association/Society of Cardiovascular Anesthesiologists/Society for Cardiovascular Angiography and Interventions/Society of Interventional Radiology/Society of Thoracic Surgeons/Society for Vascular Medicine guidelines. Studies have documented an increased prevalence of BAV in first-degree relatives, supporting a hereditary component for BAV. An echocardiographic study of 30 families with BAV found that 37% of families had more than 1 affected first-degree relative. This is a clear opportunity to identify a vulnerable population and should be a focus for knowledge dissemination efforts. Enrollment of patients with BAV into specialized valve clinics may increase the screening efforts and improved adherence to current guidelines.

Although the majority of surgeons follow recommended aortic size thresholds for replacement of the ascending aorta, it was notable that those who had been more recently trained made choices more consistent with the guidelines, suggesting greater familiarity with recent guidelines. However, other surgeons are considerably more aggressive or conservative in their approach. It was surprising that some surgeons advocated resection of the aorta with normal dimensions and no indication of dilatation. Although concomitant replacement likely adds only a small incremental operative risk, such an aggressive approach is not

FIGURE 5. Survey results of the effect of age on surgical management. Surgical management for a stenotic BAV with dilated ascending aorta in 35-, 50-, and 75-year-old patients with 53-mm ascending aorta and normal aortic root and arch. AVR, Aortic valve replacement.
supported by current evidence. The true additive risks of concomitant procedures require further investigation. In contrast, others treated patients with BAV as they would any other patient with aortic dilatation, perhaps reflecting a lack of knowledge of current recommendations and the threat of BAV aortopathy. The strength of the evidence used in the development of the current guidelines may be questionable, resulting in variable surgeon practices and poor adherence to expert recommendations. The surgical guidelines for the bicuspid aorta recommend thresholds for aortic replacement primarily on the basis of nonrandomized data and expert opinion. The current observations suggest that a randomized control trial of surgical intervention (at a specific aortic threshold) versus watchful waiting should be considered. For now, therefore, it would seem that continuing medical education efforts focused on appropriate management of patients with BAV with aortopathy should be considered.

After the decision to replace the aorta, there is a wide variation in the extent and type of aortic repair used. The majority of surgeons supported a conservative approach to both aortic root and aortic arch replacement at the time of ascending aorta replacement. Among surgeons who more frequently perform complex aortic procedures, the optimal surgical approach differed widely but generally favored a more aggressive extent of aortic resection. These surgeons presumably are more aggressive in an attempt to prevent future complications or reoperations, and perhaps their increased experience with complex aortic procedures justifies this approach by a low operative morbidity and mortality. In selected patients, a complex root or arch intervention in experienced hands has a low incremental surgical risk, and such an approach may be rational. For example, a single-center series of 206 patients with BAV stenosis and aortic aneurysm underwent a Bentall procedure with low surgical mortality (2.9%) and no short-term reoperations for aortic complications. However, even in this series, the surgical approach varied widely for Bentall, hemi-arch, or some combination. This suggests that the optimal strategy is yet unidentified and highlights an unmet need in terms of surgical management.

A less aggressive approach, despite the ability to perform complex procedures with a low risk, may be warranted. The rationale for replacement of the dilated aorta is to prevent dissection, aneurysm formation, aortic rupture, and sudden cardiac death. McKellar and colleagues reviewed 1286 patients with BAV at the Mayo Clinic who underwent AVR and found 89% 15-year freedom from aortic dissection, aortic replacement, or aortic dilatation. Multivariable predictors included repeat AVR, concomitant coronary artery bypass grafting, smoking, and aortic enlargement at the time of operation. Girdauskas and colleagues followed 153 patients with BAV and 40- to 50-mm ascending aortas who underwent AVR. They reported a 93% 15-year freedom from adverse aortic events, including 3% aortic replacement and no incidence of dissection. A recent long-term follow-up study demonstrated that aortic dissection occurred in only 2 of 416 patients with BAV over a mean follow-up of 16 ± 7 years, an incidence of 3.1 cases per 10,000 patient years with no dissections with aortic diameter less than 45 mm or normal functioning aortic valves. Some surgeons advocate for aggressive resection of the distal aorta and perform an “open distal” using short periods of circulatory arrest in an effort to resect as much aorta as possible. More recent data do not support this practice. A less aggressive approach as described in this study by surgeons who would not replace a concomitant dilated aorta until 50 or 55 mm may put the patient at risk of future aortic complications or interventions. There were no differences between practice settings, but surgeons who were in earlier years of practice tended to adhere closer to current guidelines.

Although the guideline recommendations are largely based on aortic root or ascending aortic diameter thresholds, it was noteworthy that 63% of respondents indicated that in addition to these absolute measurements they would consider alternative indices of aortic size (aortic diameter indexed to body surface area or ratio of ascending to descending diameter) in decision-making.

**Study Limitations**

These data represent the results of voluntary participation and may not be reflective of the entire cardiac surgeon population, particularly outside of Canada where practice patterns may differ. Answers were not verified for accuracy. We were unable to determine the rationale behind the cardiac surgeons’ decision-making. Our surgical scenarios were rather simple, and the surgical decision-making for individual patients is reality more complex. We did not consider other factors, such as patient preference for a bioprosthetic or mechanical valve, symptoms, comorbidities, or other factors that determine the decision to perform surgery and the type of repair performed. We did not specifically ask whether the surgeons’ approach to bicuspid aortopathy would change if coincident replacement of the aortic valve was performed with a mechanical versus tissue prosthesis. It is possible that surgeons would be more aggressive at addressing the aorta when implanting a mechanical versus tissue valve. Finally, we did not ask surgeons about their recommendations regarding antihypertensive therapies, exercise prescription, and management of bicuspid aortopathy in pregnancy, which are all important clinical questions.

**CONCLUSIONS**

Through a survey of 100 Canadian cardiac surgeons, we determined that the majority were knowledgeable of bicuspid aortopathy and the current guidelines regarding its surgical management. Seemingly at odds with these
findings, the surgical management of patients with BAV was highly variable, particularly in size thresholds for aortic replacement and the type and extent of surgical repair for the aortic root and arch. To our surprise, approximately one third of surveyed cardiac surgeons would replace the ascending aorta at a threshold of 40 mm in patients with BAV. Emerging data suggest that the actual rates of aortic dissection are lower than previously believed, and that only unique phenotypes (eg, the root phenotype) may be at so-called higher risk. Thus, it is imperative that surgeons be educated about the natural history of bicuspid aortopathy and the types of dilatation that are more likely to progress to ensure that an exceedingly aggressive approach is not adopted unless necessary. There is an urgent need for a bio-markers of aortic progression (biochemical or rheological) to help better define, within this heterogeneous population, the individuals with bicuspid aortopathy in whom aggressive intervention should be begun. The highly variable surgical approaches and departures from current guidelines represent potential gaps in knowledge translation and the need for evidence-based decision-making for this population of patients.

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References


APPENDIX E1. Canadian Bicuspid Aortopathy Survey
(Version 1.1)

Question 1: How many years have you been in practice as a staff cardiac surgeon? (Check one)
☐ A: 0-5 years
☐ B: 5-10 years
☐ C: 10-15 years
☐ D: 15-20 years
☐ E: 20-25 years
☐ F: >25 years

Question 2: How would you describe your cardiac surgical practice? (Check one)
☐ A: Academic setting
☐ B: Community setting

Question 3: In what province is your cardiac surgical practice? (Check one)
☐ A: British Columbia
☐ B: Alberta
☐ C: Saskatchewan
☐ D: Manitoba
☐ E: Ontario
☐ F: Quebec
☐ G: Nova Scotia
☐ H: New Brunswick
☐ I: Newfoundland

Question 4: How many cardiac cases do you perform per year? (Check one)
☐ A: 0-50
☐ B: 50-100
☐ C: 100-150
☐ D: 150-200
☐ E: 200-250
☐ F: >250

Question 5: What is the proportion of aortic valve replacements/repairs that you perform each year? (Check one)
☐ A: 1%-10%
☐ B: 10%-20%
☐ C: 20%-30%
☐ D: >40%

Question 6: Approximately how many of the following procedures do you perform per year in your current practice? (Check one)

I Ascending aortic replacements (supracoronary graft, closed distal)
☐ A: None
☐ B: 0-5 cases
☐ C: 5-10 cases

II Bentall procedure
☐ A: None
☐ B: 0-5 cases
☐ C: 5-10 cases
☐ D: 10-20 cases
☐ E: >20 cases

III Hemiarch and arch replacements with circulatory arrest
☐ A: None
☐ B: 0-5 cases
☐ C: 5-10 cases
☐ D: >10 cases

IV Aortic valve repair (valve-sparing operation)
☐ A: None
☐ B: 0-5 cases
☐ C: 5-10 cases
☐ D: >10 cases

Question 7: Do you think that aortic dilation associated with bicuspid aortic valve is a unique pathological condition? (Check one)
☐ A: Yes
☐ B: No
☐ C: Don’t know

Question 8: In your opinion, what is the primary driver of bicuspid aortopathy? (Check one)
☐ A: Genetics
☐ B: Hemodynamics
☐ C: Both
☐ D: Don’t know

Question 9: In your practice, what is the most common form of aortic dilatation that you see in patients with bicuspid aortic valve? (Check one)
☐ A: Dilation of the ascending aorta only
☐ B: Dilation of the aortic root only
☐ C: Dilation of the transverse aorta only
☐ D: Dilation of the ascending aorta and aortic root
☐ E: Dilation of the ascending aorta and transverse aortic arch
☐ F: Dilation of the aortic root, ascending aorta and transverse aortic arch
☐ G: Don’t know

Question 10: In your opinion, what is the most common type of leaflet fusion pattern associated with bicuspid aortic valve? (Check one)
☐ A: Left-Right
☐ B: Left-Non
Question 11: In your opinion, is the type of leaflet fusion pattern associated with a unique aortic dilation phenotype? (Check one)

- A: Yes
- B: No
- C: Don’t know

Question 12: Consider a 50-year-old patient with bicuspid aortic valve that is neither stenotic nor regurgitant. In your practice, what is your size threshold to replace the ascending aorta? (Check one)

- A: 35 mm
- B: 40 mm
- C: 45 mm
- D: 50 mm
- E: 55 mm
- F: ≥60 mm
- G: Don’t know

Question 13: Consider a 50-year-old patient with bicuspid aortic valve undergoing aortic valve replacement for aortic stenosis. In your practice, what is your size threshold to replace the ascending aorta? (Check one)

- A: 35 mm
- B: 40 mm
- C: 45 mm
- D: 50 mm
- E: 55 mm
- F: ≥60 mm
- G: Don’t know

Question 14: You are performing aortic valve replacement on a 35-year-old patient with bicuspid aortic stenosis. The ascending aorta is 53 mm and the root and arch are normal. What operation would you perform? (Check one)

- A: AVR only
- B: AVR and replacement of ascending aorta (crossclamp on)
- C: AVR and aortoplasty
- D: AVR, replacement of ascending aorta and hemiarch (open distal anastomosis)
- E: Bentall
- F: Bentall and hemiarch
- G: Aortic valve sparing procedure
- H: Aortic valve sparing procedure and hemiarch
- I: Replacement of ascending aorta and hemiarch
- J: Don’t know

14a: If the aforementioned patient was aged 50 years, which of the following would you perform? (Check one)

- A: AVR only
- B: AVR and replacement of ascending aorta (crossclamp on)
- C: AVR and aortoplasty
- D: AVR, replacement of ascending aorta and hemiarch (open distal anastomosis)
- E: Bentall
- F: Bentall and hemiarch
- G: Aortic valve sparing procedure
- H: Aortic valve sparing procedure and hemiarch
- I: Replacement of ascending aorta and hemiarch
- J: Don’t know

14b: If the aforementioned patient was aged 75 years, which of the following would you perform? (Check one)

- A: AVR only
- B: AVR and replacement of ascending aorta (with crossclamp)
- C: AVR and aortoplasty
- D: AVR, replacement of ascending aorta and hemiarch (open distal anastomosis)
- E: Bentall
- F: Bentall and hemiarch
- G: Aortic valve sparing procedure
- H: Aortic valve sparing procedure and hemiarch
- I: Replacement of ascending aorta and hemiarch
- J: Don’t know

Question 15: In a patient with mild-moderate aortic insufficiency secondary to a bicuspid aortic valve and a aortic root of 50 mm, with a normal ascending aorta and normal aortic arch, would you perform a valve sparing root replacement? (Check one)

- A: Yes
- B: No
- C: Don’t know

Question 16: In patients with a bicuspid aortic valve when replacing the ascending aorta electively how often do you perform an open distal anastomosis? (Check one)

- A: Never
- B: 0%-25%
- C: 25%-50%
- D: 50%-75%
- E: <75%
16a: If you are planning an open distal anastomosis, what cerebral protection strategy would you use during deep hypothermic circulatory arrest? (Check one)

☐ A: Antegrade selective cerebral perfusion
☐ B: Retrograde selective cerebral perfusion
☐ C: No cerebral perfusion
☐ D: Don’t know
☐ E: Not applicable

16b: If you are planning on performing an open distal anastomosis or limited hemiarch with circulatory arrest, to what temperature do you cool the patient? (Check one)

☐ A: 18°C-20°C
☐ B: 25°C
☐ C: 28°C-30°C
☐ D: 32°C
☐ E: 34°C
☐ F: Not applicable

Question 17: In your practice what modality do you use to best assess the size of the ascending aorta? (Check one)

☐ A: Echocardiography
☐ B: Computed tomography
☐ C: Magnetic resonance imaging

Question 18: In your practice which of the following alternative measures of aortic size do you employ? (Check all that apply)

☐ A: Ratio of ascending aorta to descending aorta
☐ B: Ascending aorta indexed to body surface area
☐ C: Ascending aorta indexed to patient height
☐ D: I do not use other measures
☐ E: Other_________________________

Question 19: In your practice, do you recommend that first degree relatives of patients with bicuspid aortic valve undergo echocardiographic screening? (Check one)

☐ A: Yes
☐ B: No
☐ C: Don’t know

Question 20: Are you aware of the current AHA/ESC guidelines regarding the management of the ascending aorta in patients with bicuspid aortic valves? (Check one)

☐ A: Yes
☐ B: No

AHA, American Heart Association; AVR, aortic valve replacement; ESC, European Society of Cardiology.
FIGURE E3. Cardiac surgeon demographics. Top: Proportion of Bentall procedures by surgeons who perform 0 to 5 versus more than 5 Bentall procedures per year. Middle: Proportion of aortic arch replacement procedures by surgeons who perform 0 to 5 versus more than 5 aortic arch replacement procedures per year. Bottom: Proportion of valve-sparing root replacement procedures by surgeons who perform 0 to 5 versus more than 5 valve-sparing root replacement procedures per year.