Editorial

Diameter-based decision making in bicuspid aortic valve aortopathy: is it time to expand our perspective?

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Bicuspid aortic valve (BAV) and the associated aortopathy (BAVA) are remarkably commonplace; the former is the most common congenital heart anomaly occurring in 1–2% of the general population and aortic dilatation to a greater or lesser degree is present in 30–60% if not more (1). Guidelines for ascending aortic intervention (AAI) to prevent acute aortic syndromes (AAS) have been established and recommend aortic replacement when aortic diameter exceeds 55 mm (Class I), at 50 mm when other factors such as family history, expansion rate are present, (Class IIa), or when the patient is low operative risk at a center with established expertise (Class IIa), or at 45 mm if concomitant aortic valve replacement is planned (Class IIa) (2). Despite these *size-based* guidelines, still there is controversy in the literature and, more importantly, in practice, with intervention occurring over one-third of the time in BAVA patients with diameters less than 45 mm (1).

This practice is likely driven by Pape's observation that 40% of patients in the International Registry of Acute Aortic Dissection (IRAD) presenting with type A dissection had ascending aorta diameter less than 5.0 cm and one-third of BAV patients had aortic diameters less than 5.5 cm (3). Dare we neglect the risk of AAS to those in the "gray zone" between 4.5 and 5.5 cm? At the same time, if we operate on everyone, surely we will do some harm. Clearly, we need a more nuanced approach.

The data concerning size seem little help to us in determining who needs an operation; perhaps we should acknowledge what the data do tell us, which is that diameter alone is woefully inadequate. To be fair, the focus on valve morphology—BAV or tricuspid—is itself an effort in this direction. But even here the message is uncertain. Michelena and colleagues found the age-adjusted relative risk of aortic dissection in patients with BAVA to be eight times greater than those with tricuspid aortic valves (TAV) (4). Still, the absolute risk is low, with only two dissections (0.5%) among 416 consecutive BAVA patients followed for 16 years. In Elbadawi's query of the NIS database yielding over 38,000 hospitalizations for AAI, the incidence of BAV was similar to the general population incidence at only 1.1%, challenging the notion that BAV is itself a risk factor for dissection (5).

What about the other established risk factors for aortic dissection? Hypertension is clearly an important predisposing factor for acute aortic dissection (3), as are other risk factors include age, smoking, chronic obstructive pulmonary disease (COPD), pregnancy/delivery, cocaine usage and fluoroquinolone use (6). And yet none of these factors are addressed in our guidelines. Evidence is accumulating for other parameters such as the presence of "root type" phenotype, in which the sinuses of Valsalva are dilated to a greater extent than the tubular portion, as an independent predictor of rapid progression (>0.9 mm/year) with a striking odds ratio of 14 (7). In the same study, functional aortic regurgitation was also predictive of rapid growth (OR 2.3). Others have focused on indexing size to body surface area (8) or height (9), arguing that one should not expect a small person's aorta to be the same size as a larger person's. Despite the logic, however, the data are sparse. Advanced imaging modalities such as 4D-flow MRI have shown early promise to categorize specific phenotypes of BAVA and can map patient-specific wall stress in the

ascending aorta (10). Non-image-based predictors are being explored as well, such as the circulating TGF-β-1 to soluble endoglin ratio and others (11).

In the 17th century, John Graunt transformed demography by summarizing key elements of Bills of Mortality in numerical tabular form. This permitted the application of mathematical methods of analysis to understand trends in the frequency of human disease and was the beginning of the field of epidemiology. As powerful as such an approach can be, its danger and weakness lie in the information left off the table—the individual characteristics that might serve to nuance our understanding of the disease if only we were to attend to them. Perhaps there is a lesson for us here to broaden our perspective. Guidelines are carefully constructed based on the best possible information and are intended to establish a foundation or common ground for practicing clinicians. As such they should neither be ignored nor canonized. The following century, Thomas Bayes would introduce a statistical approach that invites nuance in estimating probability as additional information accumulates. Perhaps we should accept aortic diameter as informing our "prior probability" and move on to seek new information and novel parameters, particularly for patients in the "gray zone" to inform our judgements. Accepting the limitations of our knowledge, perhaps cardiologists and surgeons should grow more comfortable in observing BAV patients with moderate-sized aortic dilation who do not have concomitant additive risk factors. Finally, let's acknowledge that aortic diameter is the first data point but not the only data point, and as such should be the beginning of the conversation but not the end.

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Footnote

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