The conundrum of aortic dissection in patients with bicuspid aortic valve: the tissue, the mechanics and the mathematics

Alessandro Della Corte*

Department of Cardiothoracic Sciences, Second University of Naples, V Monaldi Hospital, Naples, Italy

* Corresponding author. Department of Cardiothoracic Sciences, Second University of Naples, Cardiovascular Surgery and Transplant, V. Monaldi Hospital, via L Bianchi, 80131 Naples, Italy. Tel: +39-081-7064020; fax: +39-081-5464594; e-mail: aledellacorte@libero.it (A. Della Corte).

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Studies addressing the features and outcomes of type A aortic dissection (TAAD) in the bicuspid aortic valve (BAV) patient population can importantly feed into our knowledge of BAV aortopathy, increasing the level of evidence underpinning official recommendations [1].

Etz et al. presented a retrospective comparison between tricuspid aortic valve (TAV) and BAV patients operated on for acute TAAD [2]. Two findings were particularly striking, although not statistically significant: (i) the mean ascending aortic diameter at the time of dissection was greater with BAV than with TAV (by more than 7 mm); (ii) notwithstanding the significantly younger patient age at dissection, BAV implied a higher hospital mortality. Both findings confirm previous independent observations [3−5] and call for in-depth reflections.

Regarding the greater aortic diameter at dissection, this might seem unexpected, given the common notion that BAV patients’ aorta is prone to ‘earlier’ failure. Of note, in a recent study, the same difference in mean diameters was found [4], reaching statistical significance there, probably due to the greater numbers. What is the possible explanation for this evidence? Firstly, the fact that ‘post-dissection’ diameters were reported in both studies [2, 4] could allow the hypothesis that when TAAD occurs, the BAV aorta undergoes a greater acute enlargement than the TAV aorta, possibly due to intrinsically worse aortic wall cohesion. In a recent investigation, a mean diameter increment of ~30% was calculated; unfortunately, BAV patients were excluded from such analysis [6]. However, in another report [3] predissection diameters (when available) were also significantly greater in the BAV group, which disproves that hypothesis. So, if the BAV aorta really dissects at a greater diameter than the TAV one, then why has prophylactic elective surgery of BAV aortopathy been traditionally indicated for smaller diameters? Indeed, the latest version of the guidelines recommend surgery for the same cut-off of 55 mm for both non-syndromic TAV- and BAV-associated aortopathies, in the absence of risk factors [1].

Secondly, considering this matter also in mathematical terms as previously suggested [7], the a priori risk of TAAD at a given aortic diameter is a ratio between the number of events occurring (numerator) and the number of patients at risk (denominator), i.e. those presenting with the given diameter. Studies having TAAD as their referral [2−5] can only estimate the numerator; thus, the absolute number of TAADs occurring in dilated aortas could be high with BAV because in the general BAV population aortic dilatation is frequent. The denominator question also relates to why indexing methods are not a solution: as Etz et al. correctly argued, a policy of pre-emptive operation on all patients with a Svensson index >9−10 could have led to a number of unnecessary operations [2]. None of the proposed indexing methods has been
adequately tested so far, in terms of sensitivity and, equally important, specificity [1].

Thirdly, the mechanics: both hypertension and aortic enlargement increase wall tension, but the mechanical failure of the diseased intimal and medial layers, prompting TAAD development, can be demonstrably reached even without dilatation. The mechanical stimulus that is currently believed to be causative in intimal tear formation is abnormal shear stress: normally, wall shear stress decreases exponentially with increasing aortic diameter; however, this does not happen in the BAV aorta [8], owing to the altered flow patterns prompted by the eccentricity of valve opening. Thus, with TAV, local haemodynamics would favour TAAD onset more likely when the diameter is not aneurysmal, whereas, in the BAV aorta, the triggering biomechanical cue would stand even with dilatation, a hypothesis that fits with the observation by Etz et al. [2].

There is a clear need for further improvement of our knowledge of the mechanisms and risk factors for TAAD: they might be only in part shared with aortic dilatation and at least in part different between BAV and TAV patients. Rather than searching for a cut-off diameter for universal surgical indication, future efforts should be aimed at discovering which are the measurable factors determining TAAD risk, along with (or instead of) aortic diameter [9]. Unfortunately, Etz’s study failed to report on the ‘pattern’ of proximal aortic dimensions [2], i.e. distinguishing dilatations prevailing at the sinuses of Valsalva (root phenotype) from the more common phenotype with maximal dilatation at the supracoronary tract [10]. Notably, the site of intimal tear was more frequently at the root level in BAV than TAV patients [2], and the rate of >grade II aortic regurgitation (typically associated with root phenotype [10]) was particularly high [2]. Compared with the usually reported proportions—20–30% of the BAV-associated dilatations [10]—the root phenotype seems consistently over-represented in BAV-TAAD series [2–4], thus confirming our suggestion that it might represent a higher risk phenotype [10].

The comparable hospital mortality in BAV- and TAV-TAAD patients [2, 4], mirroring the report from the International Registry of Aortic Dissection of similar mortality in young and older TAAD patients [5], shows that the catastrophic nature of the disease implies a high perioperative risk quite independent of the patient’s age. In the BAV group [2], the higher prevalence of entry tear location at the sinuses (possible coronary ostial involvement), the more frequent need for resuscitation at presentation (possible rupture, tamponade, shock or infarction) is, and the higher postoperative rates of both assist device implant and cardiac deaths may all indicate that aortic wall fragility increases the severity of TAAD in some BAV patients. In addition, the most complicated anatomic-clinical forms of TAAD may exert an ‘age-selection’, in that the older patients (more frequently TAV) may die even before hospital admission, whereas the younger, otherwise healthier ones (more often BAVs) may reach the operative room even in the most compromised conditions, implying worse outcomes.

No study has ever compared long-term outcomes of supracoronary replacement versus composite root replacement specifically in BAV-associated dissection. Therefore, even though late survival was excellent with an aggressive approach with regard to the BAV root (94% composite root replacements vs 46% with TAV) [2], we do not know whether a more individualized management, including selective indication to root replacement (with its higher inherent operative risk) in BAV patients with the root phenotype and/or entry tear at the sinus level, would have entailed a better early outcome too.

The general perspective on BAV aortopathy pathogenesis should change from the dichotomy between haemodynamic and genetic theories to a new unifying vision whereby variable combinations of flow disturbance and genetic defects produce diverse phenotypes with different risks of TAAD [9]. This new vision calls for the development of risk stratification criteria and personalized methods of surveillance and treatment: the disease is multiform; any generalization would be misleading.

REFERENCES


