BICUSPID AORTOPATHY OR BICUSPID AORTOPATHIES? THE RISK IN GENERALIZING To the Editor:

We read with interest the article by Fazel and coworkers,¹ which highlighted the previously underreported issue of transverse aortic arch involvement in aortopathy related to the bicuspid aortic valve (BAV). We totally agree with the authors' claim that the surgical approach to BAV aortopathy should be custom-tailored, inasmuch as different morphologic patterns can be encountered: this emerged also in our recent study on 280 BAV subjects.² However, their conclusion in favor of pre-emptive concomitant total arch replacement for the majority of BAV patients undergoing proximal aortic operations may raise concerns.

In a recent large series,³ the dilatation/aneurysm involved the arch in only 4.3% of BAV patients, versus 7.3% without BAV. The anticipation by Fazel's group¹ that a proximal arch measuring 3.6 cm (as in "cluster IV," including 45% of the study patients) will dilate to 5 cm within 15 years after ascending aortic replacement is not confirmed by clinical evidence. The 1.9 mm/y growth rate that they mention actually refers to the mid-ascending tract,³ with the rate of arch size progression currently being unknown.

Unfortunately, this study lacks a comparison with matched subjects with a tricuspid aortic valve¹: thus

the observed patterns may not be unique to the BAV setting, and the definition itself of arch dilatation was not done by comparison with normal values. Moreover, as evident from Figure 3,¹ only cluster III (ascending+arch pattern) was quite homogeneous in terms of transverse arch involvement, whereas many patients in cluster IV (root+ascending±arch pattern) actually had small arch dimensions, in all comparable with those in cluster I (root pattern) or II (ascending pattern). However, the authors omitted a statistical comparison of aortic dimensions between clusters.

The BAV function and BAV morphology factors should be considered when trying to better discriminate subgroups of BAV aortopathy. In the Westhoff-Bleck magnetic resonance imaging study,⁴ only BAV patients with moderate-severe aortic regurgitation had significantly greater transverse arch size compared with matched controls, whereas in normally functioning BAV patients only root and ascending diameters were larger than in controls. Schaefer and colleagues⁵ recently found right-left orientation of the BAV cusps (right-noncoronary fusion) to be associated with a significantly larger proximal arch compared with anteroposterior orientation (right-left coronary fusion). Interestingly, in Fazel's report, the highest prevalence of the right-noncoronary pattern and the highest mean grade of aortic insufficiency were observed in cluster III. In our ongoing magnetic resonance imaging study, 8 nonstenotic/nonregurgitant BAV subjects with right-left coronary fusion have been compared so far with matched controls, showing no significant difference in proximal (24.3 ± 2.8 cm vs 25.9 ± 4.1 ; $P = .36$) and transverse arch size (20.5 ± 2.3 cm vs 22 ± 3.1 ; $P = .29$).

Different aortopathies may subtend the various possible patterns of BAV-related aortic dilatation: probably it is not possible to issue general principles and recommendations on such a com-

plex and multifaceted matter. The identification of a definite BAV phenotype representing a more "malignant" form of disease, amenable to more extensive surgical resections, still requires special research efforts.

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Reply to the Editor

We thank Della Corte and Cotrufo from Naples for their interest in our observations and for raising several critical points concerning the "aortopathy" associated with bicuspid aortic valve (BAV) disease. Owing to space limitations, we can only discuss the important question of extent of aortic resection at the time of operation, particularly in light of the recent condemnation of the more aggressive American College of Cardiology/American Heart Association guidelines by Guntheroth.¹

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