Letter by Barison et al Regarding Article, “Familial Clustering of Mitral Valve Prolapse in the Community”

To the Editor:

Delling et al1 wrote an interesting article on the familial clustering of mitral valve prolapse (MVP), using a well-characterized population cohort with 3-generation data. This article paves the way to further studies on the genetic determinants of MVP, but also on its clinical significance once survival data will be analyzed.

As the authors acknowledge, this work is partly retrospective, because echocardiography was not available in all of the original (generation 1) participants, and was not performed at the same age in the offspring (generation 2) and the generation 3 populations. On the one hand, little is known about the progression of MVP over time, its age-related prevalence, or its long-term prognostic significance. On the other hand, this could explain why the prevalence of MVP was 1.3% (49 of 3679) in generation 3, whereas its prevalence was higher in the offspring population: according to the 5.1% prevalence of parental MVP (186 of 3679 participants had at least 1 parent with MVP), the prevalence (p) of MVP in the offspring population could be estimated at \( \approx 2.6\% \) (because the probability that at least 1 parent presents MVP \( \approx 1 - \left(1 - \frac{1}{p}\right)^2 \) is 0.051, then \( p=0.026 \)). We agree with the authors that longitudinal prospective studies will be needed to address these points.

Moreover, the authors found no difference in left atrial size, left ventricular size, or systolic function between patients with or without parental MVP, suggesting it is a primary valvular disease. Nevertheless, previous studies correlated MVP to other morphological abnormalities, involving papillary muscles,2 the right ventricle, and the tricuspid valve,3 and to electrophysiological disorders, such as ion-channel diseases,4 supraventricular and ventricular arrhythmias, and sudden death.5 In particular, it has been hypothesized that the same genes may control the myocardial, valvular, and conduct system formation during early embryonic heart development,6 explaining why several different pathological phenotypes are often associated. Other pathological and cardiovascular magnetic resonance studies have described perivalvular ventricular fibrosis and papillary muscle fibrosis,7 suggesting that MVP might be an acquired disease progressing over time owing to a tight relationship between the prolapsing valve and ventricular structure, with possible myocardial remodeling and enhanced arrhythmic susceptibility. It would be probably worth investigating these aspects in this well-characterized 3-generation population to confirm whether MVP presents broader morphological, functional, and arrhythmic correlations and whether they progress over time.

Overall, the time has come to consider MVP as much more than an isolated valvular disease. We agree with the authors that further studies are needed to elucidate not only the genetic determinants of MVP and the potential role of family screening, but also its broader mechanical, electrophysiological, and prognostic implications.

Disclosure

None.

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