Response to Letters Regarding Article, “Arrhythmic Mitral Valve Prolapse and Sudden Cardiac Death”

Our article on “arrhythmic” mitral valve prolapse (MVP) triggered a debate among clinical cardiologists about the implications in terms of diagnosis, risk stratification, and therapy. Thus, the letters by Sheppard et al and Providencia and Lambiase are welcome and give us the opportunity to be back on this hot topic.

Sheppard et al, by studying a series of sudden cardiac death (SCD) victims with classic myxoid mitral valve degeneration, found similar features, in terms of myocardial fibrosis in one or both papillary muscles and adjacent left ventricular free wall, although they did not specifically assess the inferobasal wall. Their findings further strengthen our hypothesis that left ventricular scarring in targeted areas subjected to higher mechanical stress is the substrate of electric instability in arrhythmic MVP and support a role for contrast-enhanced cardiac magnetic resonance for risk stratification in a selected subgroup of patients.

Furthermore, the authors did not find a female preponderance and report a lower prevalence of MVP in their SCD cohort compared with our North East Italy SCD series (1.7% versus 7%). To explain the difference, they hypothesize an effect of the Italian preparticipation screening program in identifying people with ECG-detectable heart diseases at risk of SCD such as cardiomyopathies and ion channel diseases, with the consequent “relative” preponderance of ECG-silent cardiovascular causes of SCD, including MVP, in our series. As additional explanation, we should point to the fact that our center is not a tertiary referral center like the Cardiac Risk in the Young but is a registry set up to study prospectively all autopsied cases of SCD occurring in the young adult population 40 years of age according to a homogeneous morphological protocol by a team of cardiovascular pathologists.

Concerning the letter by Providencia and Lambiase, our study does not pretend to provide epidemiological data on the real incidence of arrhythmic MVP prolapse. However, their estimation of the annual incidence of SCD resulting from MVP seems unreliable. First, data on annual incidence of SCD in the young are obtained by retrospective analysis of death certificates and public media reports on fatal events in Denmark, which are quite different from those gathered in Italy according to a prospective study design with systematic investigation of young people who died suddenly and underwent a pathological heart investigation. Second, epidemiological data of MVP in the general population (2%-3%) are based on the simple echocardiographic diagnosis of MVP (ie, single-leaflet or bileaflet prolapse of at least 2 mm beyond the long-axis annular plane), which is not representative of either a cardiac disease or the nosographic entity we are referring to with the term arrhythmic MVP.

We are well aware that the risk of SCD in people with MVP, as identified by simple echocardiographic criteria, is low. For this reason, 2 major tasks should be accomplished in the next future: identifying diagnostic criteria capable of recognizing patients with an MVP corresponding to a real disease entity and recognizing among patients with true MVP those at higher risk of severe ventricular arrhythmias.

The arrhythmic MVP profile is that of a patient, usually female, with not just an echocardiographic diagnosis of MVP but also bileaflet or posterior myxoid degeneration, ECG repolarization abnormalities, and polymorphic/RBBB morphology complex ventricular arrhythmias. Only these patients would deserve further investigation, including contrast-enhanced cardiac magnetic resonance and a strict arrhythmias surveillance for proper management and SCD prevention, thus avoiding the risk of exponential increase in costs, referral, and false-positive results.

Finally, we agree that the fight against SCD should always combine primary and secondary prevention strategies. In fact, there are cardiovascular causes of SCD that either escape identification or, if identified, do not yet have precise guidelines for management. Thus, dissemination of automated external defibrillators and promotion of the culture of resuscitation to professionals or even laypeople trained to use the device can make the difference. However, we should be aware that most patients with MVP die suddenly at rest or during sleep at home.

Disclosures

None.

References

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