Sudden Cardiac Death in Mitral Valve Prolapse

To the Editor:

I read with interest the excellent report on sudden cardiac death with apparently normal heart by Chugh et al.1 Of note was the finding that 28 of 270 hearts (10%) had pathological criteria for mitral valve prolapse. In addressing the association between mitral valve prolapse and sudden cardiac death, the authors classified these hearts as having a nonspecific structural abnormality. Such an assumption is open to question.

Evidence to date suggests that the predominant mechanism of sudden cardiac death in mitral valve prolapse is fatal ventricular arrhythmias.2 Therefore, it would be extremely difficult for a pathological study such as that by Chugh et al1 to prove or disprove conclusively the causal relation between mitral valve prolapse and sudden cardiac death.

Whether mitral valve prolapse claimed the life of Shylock in the Merchant of Venice, who claimed the heart of Portia’s lover,3 or Antonia in the Tales of Hoffman, who sang herself to death,4 there is no question that although generally a benign condition, mitral valve prolapse may be a deadly disease at times.5 Admittedly, mitral valve prolapse is an uncommon cause of sudden cardiac death,2 and its propensity to cause sudden death has been of major concern mainly because of the high frequency of this abnormality in the general population.5

Tsung O. Cheng, MD
George Washington University
Washington, DC

5. Cheng TO. Mitral valve prolapse: the Merchant of Venus or the Tales of Hoffman? Eur Heart J. In press.

Response

In his insightful letter, Professor Cheng appropriately questions our assignment of cases of sudden cardiac death with mitral valve prolapse to the subgroup of “structural abnormality, nonspecific findings.”

Two factors influenced this categorization. Although mitral valve prolapse has been associated with sudden death, the causal relationship between the 2 entities remains controversial.1–3 The significant incidence of mitral valve prolapse in the general population and extremely low risk of sudden death have led to the recommendation of reassurance as a major part of medical management for this condition.4 Another factor for our cautious approach was that although abnormalities of the atrioventricular nodal artery have been reported in association with mitral valve prolapse and sudden cardiac death,5 we did not complete detailed morphological studies of this vessel as a part of our investigation.

We are grateful to Dr Cheng for his comments and pleased that he found our article of interest. However, as far as we can tell, the conclusion of the Merchant of Venice finds Shylock outwitted, but alive and well. If he were to die suddenly, may we venture the opinion that the aging Shylock is most likely to succumb to coronary artery disease? For the 19-year old Antonia’s familial malady in the Tales of Hoffman, should we not consider hypertrophic cardiomyopathy, long-QT syndrome, or familial dilated cardiomyopathy among the differential diagnoses?

Sumeet S. Chugh, MD
Oregon Health Sciences University
Portland, Oregon
Karen L. Kelly, MD
Jack L. Titus, MD, PhD
Jesse E. Edwards Registry of Cardiovascular Disease
St Paul, Minnesota

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Tsung O. Cheng

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