Myocarditis as a Cause of Sudden Death

To the Editor:

We appreciated the outstanding review by Drs Zipes and Wellens on sudden cardiac death. However, the authors did not mention myocarditis as a cause of sudden death. In our experience at the Forensic Institute of Paris, myocarditis accounts for 5% of sudden, out-of-hospital cardiac deaths. Two patterns are common. Acute myocarditis, the first type, occurs most frequently in children or young adults. The diagnosis is likely to be overlooked because of the frequent lack of symptoms. When present, symptoms are not specific; they suggest a bad cold in most instances. The heart is grossly normal. In particular, it is not enlarged. Myocyte necrosis and inflammatory infiltrates, predominantly composed of lymphocytes, are present, but they are most frequently patchy rather than diffuse (American Academy of Forensic Sciences, Orlando, Fla. personal communication, 1999). Therefore, multiple myocardial samples are required for the diagnosis.

The second pattern is chronic myocarditis, which also involves young adults. The heart is again grossly normal. Histological examination of both ventricles shows patchy areas of fibrosis. In some sections, small clusters of lymphocytes may be present. These victims have no history of drug abuse, and toxicological analyses are negative. Viral infection is likely to be involved in most cases of both acute and chronic myocarditis. Idiopathic giant cell myocarditis, granulomatous myocarditis, and eosinophilic myocarditis are exceptional in our experience.

Fibrosis is known to cause slow conduction in cardiomyocytes, resulting in arrhythmogenic substrate reentry circuits and subsequent ventricular desynchronization.

Even more important is the possible role of acute inflammation of the myocardium, which could, by itself, promote ventricular arrhythmias or act as a trigger if this inflammation occurs in patients with heart disease who are prone to arrhythmia, such as those with concealed forms of cardiomyopathies. Hoffman et al studied reperfusion arrhythmia and demonstrated, for the first time, the role played by the activation of neutrophils in producing early after-depolarization. These results demonstrate a direct link between acute inflammation and an electrophysiological phenomenon that is a possible trigger of arrhythmias. From a personal correspondence with Dr Brian Hoffman (October 1997), we learned that this mechanism is also likely to be involved in patients with advanced congestive heart failure in which tumor necrosis factor-α is present and that it is also an important cause of early after-depolarizations.

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Response

Fontaine et al correctly called our attention to the role of myocarditis as a cause of sudden death. Indeed, in our article, we did not comment on inflammatory and infiltrative causes of sudden arrhythmic death for 2 reasons: restrictions regarding the size of the article and difficulty in obtaining reliable information on the incidence of these abnormalities as causes of sudden death.

Fontaine et al refer to their findings at the Forensic Institute of Paris, where myocarditis was found in 5% of sudden, out-of-hospital cardiac deaths. However, we would like to know the true incidence of myocarditis in an unselected population of sudden death victims. Myocarditis continues to be a very difficult diagnosis to make when the patient is still alive, both in the acute and the chronic state. We hope that new molecular biological and genetic techniques will provide better insight into the incidence of myocarditis and its contribution to sudden cardiac death. It will be quite difficult to unravel the arrhythmogenic mechanism(s) in this setting.

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