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Reply

We appreciate Fontaine's interest in our recent article (1). His comments provide an opportunity to clarify certain aspects concerning the pathophysiology of the syndrome of right bundle branch block (RBBB), right precordial ST segment elevation and sudden death. Our study (1) thoroughly investigated a family affected by the syndrome. As Fontaine points out, the proband's clinical findings were anticipated in a previous report from our group that addressed ventricular fibrillation without apparent heart disease (2). Compared with the previous electrocardiogram (ECG), the one shown in the more recent report was recorded later, during the clinical follow-up period, and showed a more accentuated right precordial ST segment elevation. Of note, it was recorded while the patient was taking beta-antiadrenergic drugs. Differences with time in the ST segment elevation pattern, spontaneous or induced by pharmacologic interventions, have been previously reported in this syndrome (3,4). ST segment elevation is characteristically enhanced by vagal maneuvers or class I antiarrhythmic agents and reduced after beta-adrenergic stimulation. These responses have been explained by postulating a local right ventricular (RV) "depolarized" area, which results in changes of ST segment elevation after the above interventions (4). This is even consistent with our hypothesis that RBBB and ST segment elevation are due to a double RV conduction defect, both "septal" and "parietal," which may be modulated by autonomic influences and antiarrhythmic drugs.

With regard to the structural changes of the RV wall, they resembled those observed in the "fatty pattern" of RV cardiomyopathy, with predominant fatty replacement and without inflammatory infiltrates or significant left ventricular lesions (5). However, the coexistence of a severe and progressive (as suggested by the lengthening of the PR interval in the more recent ECG) disease of the specialized conduction system with RBBB interruption, raises some concerns about the nosography of the condition. As recently reported, the conduction system is substantially spared by the dystrophic process of the RV cardiomyopathy but is affected at the advanced stage of the disease, secondary to the septal involvement (6). In our family, there was evidence that the structural changes of RV myocardium and conducting tissue may have been inherited together in the setting of a "heritable cardiac conduction and myocardial disease" (7). Further studies of linkage analysis and gene mapping are needed to better characterize the genetic background of the syndrome.

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