

Right Bundle-Branch Block, ST-Segment Elevation, and Sudden Death

To the Editor:

The review article of Alings and Wilde¹ on the syndrome of right bundle-branch block ST-segment elevation and sudden death, ignores many of the published data on this disease. It is not true that most of the published cases affected by the disease show no heart abnormalities.² Since the first detailed description of the syndrome in 1988–1989 by our group,^{3,4} a number of patients with similar ECG patterns (including some in the Brugada series) have been recognized as having structural heart disease, particularly of the conduction system and/or right ventricle, by both invasive and noninvasive techniques. Right ventricular abnormalities (at angiography, 2D echo, or MRI), a positive late potential study, a prolonged HV interval (indicative of an organic conduction disturbance), an abnormal right ventricular biopsy, and fibro-fatty replacement of the right ventricle have all been documented.^{1,5} From our data, it is evident that the right bundle-branch block pattern and ST-segment elevation (according to electrophysiological and necropsy data) are related to a diseased conduction system both at the septal and intramyocardial levels.⁵ The ST-segment elevation in the right precordial leads, especially when associated with minor forms of right bundle-branch block,³ has been explained in some cases by involvement of genetically isolated defective ion channels, which results in voltage gradient of repolarization between an abnormal epicardial action potential and a normal endocardial one.¹ This transmural dispersion of repolarization may either be acquired or enhanced in the setting of ischemia, inflammation, and degenerative heart disease.

In this regard, myocyte degeneration and necrosis in right ventricular cardiomyopathy characteristically progresses from the epicardium, which is always involved, to the endocardium, which can be spared from the disease. A definitive diagnosis of functional electrical disease, underlying a “distinct syndrome,” cannot be advanced before a detailed clinical and morphological investigation, including necropsy in fatal cases, rules out a concealed heart muscle disease.

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Response

The issue of whether structural or functional abnormalities underlie the syndrome of right bundle-branch block, right precordial ST-segment elevation, and sudden death raised by Drs Martini et al is obviously of great importance. As indicated in our article,¹ it is no surprise that the authors stress the importance of the causal presence of (concealed) heart muscle disease. Indeed, their thoroughly studied families (family?) show evidence of right ventricular cardiomyopathy.² We therefore excluded (and did not ignore) these patients from additional analysis. However, the available clinical data of the remaining 163 patients (echocardiogram, left and right ventricular angiography, ergometry, and biopsy samples available in 96%, 49%, 40%, and 20% of the patients, respectively) show no evidence of structural heart disease. By definition, concealed heart muscle disease is not excluded.

For the diagnosis of Brugada syndrome, we emphasized the presence of a dynamic ST segment that seems not to be present in the northern Italian family. Indeed, on the basis of inflammatory or degenerative heart disease, such dynamicity is not to be expected. Genetically altered ionic currents may, however, be more sensitive to modulating factors, such as the autonomic nervous system, and explain day-to-day variability. Also, the (predictable) response to various drugs seems a more logical consequence of a functional rather than of a structural defect. For the growing number of patients with drug-induced J-point elevation, this hypothesis remains to be proven.³

Without doubt, before the diagnosis is made, extensive clinical investigation should be directed to exclude any structural disorder, even in patients with established SCN5A mutations.¹ However, we hope that Drs Martini et al no longer ignore the existence of a functional disorder underlying the syndrome of right bundle-branch block, right precordial ST-segment elevation, and sudden death.

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