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Echocardiographic Findings in Patients With Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia

We read with interest the study by Yoerger et al. (1) presenting echocardiographic data of 29 probands affected by arrhythmogenic right ventricular cardiomyopathy (ARVC) diagnosed with the International Society and Federation of Cardiology task force criteria. In all cases the echocardiogram showed typical features of the disease. It is well known that ARVC diagnosis is made considering different parameters. The purpose of their study was to assess echocardiographic abnormalities in a series of patients affected by ARVC, in whom the echocardiogram has not been considered for the diagnosis.

Thus, we can assume that all these patients showed a clear form of the disease, fulfilling the diagnostic criteria even in the absence of the echocardiographic tool. However, the real problem is to understand whether the typical ARVC echocardiographic alterations have a diagnostic value per se, even when the task force criteria (2 major, 1 major plus 2 minor, 4 minor) are not satisfied. This point is critical because in the presymptomatic phase of the disease the lack of diagnosis can lead to heavy consequences for the patients. One cannot assess the utility of a diagnostic method without previous without the data. This was an intrinsic design of our study members, in a position that enhances and complements the vital role of “cognitive, electrician, or plumber” HF cardiologists.

an echocardiogram is a noninvasive technique that can be repeated in time, can we consider this diagnostic tool as the “gold standard” of ARVC diagnosis in asymptomatic patients or in patients with minor forms of the disease? Hence, the real significance of echocardiographic abnormalities in this group of “uncertain subjects” constitutes the major challenge in echocardiographic evaluation of ARVC.

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REPLY

Dr. Bause and colleagues raise several issues with regard to our investigation of the echocardiographic findings in arrhythmogenic right ventricular dysplasia (ARVD). The first is to indicate that our patients all met the task force criteria for ARVD/C without the echocardiographic-derived data. Therefore, the diagnosis was obvious without the data. This was an intrinsic design of our study (1), as one cannot assess the utility of a diagnostic method without having a firm diagnosis of the disease excluding the test that is being evaluated because there is no “gold standard.”

The purpose of our report was to identify the best quantitative echocardiographic markers of ARVD in individuals meeting task force criteria for ARVD (2). The data presented compare the measurements of different right ventricular (RV) dimensions in ARVD probands versus controls. We found that the right ventricular outflow tract was the most commonly enlarged dimension in ARVD probands. We provided a numerical value that separated the two groups. At present, it is not uncommon for echocardiographic laboratories to report only qualitative RV size. In order to discriminate the normal RV from the abnormal, as is required in the assessment for ARVD, quantitative measurements should be used.

Additionally, Dr. Bause and colleagues correctly point out the difficulty detecting ARVD in its preclinical or “presymptomatic” phase. We agree that there is an important need to be able to detect minimal structural abnormalities of the RV in patients suspected of ARVD. Upon its completion, the Multidisciplinary Registry of ARVD will have quantitative data from echocardiographic...