Anxiety in Patients With Arrhythmogenic Right Ventricular Cardiomyopathy and Implantable Cardioverter Defibrillators

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Diagnosis of a cardiac genetic disorder can be a life-changing event. Symptomatic status, uncertainty regarding disease progression, risk of sudden cardiac death (SCD), recommended lifestyle changes (ie, activity restrictions or career change), and genetic transmission to offspring all contribute to the potential psychological burden of disease. Most of the relevant literature on this topic focuses on 2 of the most common genetic cardiac diseases, hypertrophic cardiomyopathy and inherited long-QT syndrome. Studies on these disorders have consistently shown impairments in health status and quality of life, with an elevated risk for anxiety and depression.1–4 An implantable cardioverter defibrillator (ICD) may be indicated for primary or secondary prevention of SCD in these patients, and is likely to exacerbate psychological distress. Independent of underlying diagnosis, patients requiring ICDs commonly experience anxiety and/or depression related to the anticipation of a potential shock.5,6 Psychological implications of ICDs have been reported to be worse in patients with inherited diseases compared with ischemic or valvular heart disease.7 The experience of receiving 1 or more ICD shocks further negatively affects quality of life.7,8 Inappropriate shocks (ie, for sinus or supraventricular tachycardia or T-wave oversensing) can elicit similar psychological responses to appropriate shocks for life-threatening ventricular arrhythmias. This is a particularly important consideration for young patients because 1 in 4 may be expected to receive at least 1 inappropriate shock.9

Like hypertrophic cardiomyopathy and long-QT syndrome, a diagnosis of arrhythmogenic right ventricular cardiomyopathy (ARVC) carries the psychological burden of genetic transmission and risk of SCD, but patients and their health care providers face unique challenges that often begin with making the diagnosis itself. Although the modified task force criteria have improved the sensitivity without loss of specificity for diagnosis,10 the broad spectrum of disease, and prolonged concealed phase during which ventricular arrhythmias can occur in the absence of other definitive clinical findings, can cause a great deal of uncertainty. Further complicating the picture is the lack of systematic criteria for prophylactic ICD implantation in ARVC patients or asymptomatic gene mutation carriers. Recent data suggest high rates of appropriate ICD discharges in ARVC patients without prior ventricular tachycardia or fibrillation, with syncope being the only multivariate predictor of a life-saving ICD shock.11 Yet, too often, a patient’s first syncopal event results in death, making the absence of syncope an inadequate determinant of the need for ICD implantation. Unfortunately, despite the obvious value of an ICD in conferring protection against SCD in ARVC, the high rates of inappropriate shocks (19%) and device-related complications (17%) continue to be problematic.10 These complex issues surrounding the diagnosis and treatment algorithms for ARVC mean that extrapolation of data from the hypertrophic cardiomyopathy and long-QT syndrome literature relevant to the psychological impact of disease may underestimate its true prevalence and magnitude in this population. Only 1 recent study included ARVC patients in its assessment of health status and quality of life in those with cardiac genetic disease, and the number was small (n = 17), representing only 4% of the entire cohort.4

In this issue of Circulation: Cardiovascular Genetics, James et al12 examine psychosocial adjustment, anxiety, and depression in 86 adult ARVC patients with ICDs. They use 4 questionnaires that assess ICD-specific anxiety (Functional Self-Appraisal Scale), device acceptance (Florida Patient Acceptance Survey), generalized anxiety and depression (Hospital Anxiety and Depression Scale), and functional capacity (Duke Activity Status Index [DASI]). They analyze associations with demographic characteristics, including sex, age, family history of ARVC, time since ICD implantation, and history of ICD shocks. The population was high risk, with more than half having received ICDs for secondary prevention, and a similar fraction having experienced at least 1 shock (most were appropriate shocks). Despite this, the mean functional capacity was high, with a DASI score of 48.1 (ceiling value, 58.2), corresponding to an estimated peak oxygen uptake of 30.3 mL/kg per minute, which was more favorable than those of other ICD populations (mean DASI score, 17–21).13 This finding reflects the relative disconnect between arrhythmia risk and extent of structural and functional remodeling in ARVC.

Device acceptance scores in ARVC patients were in the range consistent with other ICD patients in previously published studies. Younger age was a significant multivariate...
Any intervention using an exercise component in ARVC patients will need to be carefully tailored, with direct input from the cardiologist, to ensure safe exercise practices.

James et al.\(^2\) should be commended on bringing to attention the risk for poor psychosocial adjustment in patients with ARVC and ICDs. This response is one that cardiologists often overlook and urgently needs further attention and direction.

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