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. 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. Psychological implications of ICDs have been reported to be worse in patients with inherited diseases compared with ischemic or valvular heart disease. The experience of receiving 1 or more ICD shocks further negatively affects quality of life. 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.

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, 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. 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. 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.

In this issue of Circulation: Cardiovascular Genetics, James et al 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). 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
predictor of poor device acceptance, driven by significantly lower scores on the body image and device-related distress subscales. Similarly, younger age was an independent predictor of higher ICD-specific anxiety, for which overall scores were worse than for other published ICD populations. Other multivariate predictors of higher device-related anxiety in ARVC patients included previous ICD shocks, shorter duration from device implantation, and lower functional capacity. The same factors predicted clinically significant anxiety and depression, which were evident in 31% and 9% of respondents, respectively. Surprisingly, female sex was not associated with higher levels of anxiety or depression.

This is a timely and meaningful study that addresses a frequently neglected aspect of clinical care for ARVC patients. Although recommendations for ICD implantation in ARVC patients obviously need to be driven by clinical data and perceived SCD risk, a balanced discussion that incorporates an assessment of likelihood of poor psychological adjustment to a device is extremely important. Recognition of risk predictors for poor adjustment and anxiety found in this study will greatly facilitate identification of patients who might benefit from more extensive counseling.

This study raises several questions that deserve further attention in future studies. Perhaps most important will be trying to dissect out to what extent the presence of anxiety or depression is attributable to the diagnosis of ARVC itself, as opposed to the necessity or consequences of a device. This will be challenging, particularly in the Unites States, where rates of device placement in ARVC patients (particularly probands) are high, and indications obviously correlate with higher risk of SCD. Given the strong association between younger age and anxiety scores in this study, assessment of psychological distress in children with ARVC and their parents will be critical. It will also be important to assess the psychological impact of device complications and inappropriate shocks in this population. The influence of other variables on anxiety, such as genetic status and participation and/or restriction from athletic participation, would also be worth exploring.

The data presented herein provide a cogent argument that we should be considering structured psychosocial interventions for patients deemed to be at particularly high risk for disease- or device-related anxiety or for those experiencing postshock traumatic stress. The most successful strategies for reducing anxiety in ICD patients involve cognitive-behavioral therapy to teach techniques to improve coping skills and illness appraisal, such as developing a shock plan and strategies to reduce negative thoughts. Other important components of psychological intervention include ICD-specific education, symptom and stress management training, and group therapy. Finally, exercise training as part of a comprehensive cardiac rehabilitation program has reduced anxiety and depression and improved quality of life in ICD patients. However, these studies enrolled older adults (mean age, late 50s), and the largest one specifically excluded patients with genetic arrhythmias, so it remains to be seen whether similar interventions will be equally feasible and successful at reducing anxiety and improving quality of life in ICD patients with ARVC and other inherited cardiac diseases.

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. 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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References


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