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symptomatic than that observed in the community. The observational nature of the study prevents one from drawing causal conclusions about the relations between predictors and particular outcomes.

Limitations notwithstanding, the present study represents an important contribution to our understanding of the age-dependent penetrance of ARVC and characteristics of early-onset disease. The findings dovetail with a growing body of research implicating repeated cardiac stress as a factor underlying early-onset and more severe presentations of ARVC. For example, cardiac dysfunction and markers of arrhythmia have been demonstrated in plakoglobin-deficient mice exposed to exercise (10). Human data have correlated endurance or competitive athletics with earlier onset disease and increased disease severity (11,12). In addition to exercise, the present report also suggests that desmosomal mutations are enriched in early-onset disease. Although not observed in this study, others have also related the presence of multiple mutations to early-onset ventricular arrhythmias (13) in patients with ARVC. Thus, both exercise-related and genetic factors likely interact to promote early-onset ARVC, and earlier presentations can be particularly severe.

A number of important clinical questions remain, however. What proportion of the variability in age of disease onset is explained by genetic factors and exercise? Are there mutation-specific forms of ARVC that are more likely to present in childhood or increase disease susceptibility with exercise? What are the optimal risk stratification techniques for pediatric patients with ARVC? Can the long-term course of ARVC in pediatric patients be modified by early identification and interventions?

References


Key Words: arrhythmogenic right ventricular cardiomyopathy, genetics, heart failure, inherited arrhythmia, sudden death.