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The Value of Cardiac Magnetic Resonance Imaging in Evaluation of Pediatric Patients for Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy

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In the March 17, 2015 issue of the *Journal*, Etoom et al. (¹) reported interesting findings about the role of cardiovascular magnetic resonance (CMR) for diagnosis of arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) in children. The results of Etoom et al. (¹) are unique in that there are few other studies on children with ARVD/C. For the revised Task Force Criteria (TFC), only 9 of 108 (8%) probands were diagnosed in subjects between 12 and 18 years of age (mean 38 ± 13 years of age) (^{2,3}). The study subjects of Etoom et al. (¹) averaged 13.8 ± 3.2 years of age, without a given age range. As such, the extrapolation of the revised TFC to children studied by Etoom et al. must be considered experimental.

A particularly vexing problem is trying to “validate” CMR for ARVD/C while also using CMR to diagnose ARVD/C. For Etoom et al. (¹), there was no independent anatomic comparator for CMR, because echocardiography had little or no contribution to the diagnosis (only 2 patients met major echocardiographic criteria). The children described by Etoom et al. (¹) had large right ventricles (mean right ventricular [RV] end-diastolic volume 118 mL/m^2), suggesting severe disease. Although CMR is well suited to identify RV abnormalities in that setting, it is surprising that echocardiography was unable to capture these findings.

There are a number of other challenges that the authors faced in applying the revised TFC. Their paper reports that minor diagnostic criteria were equally represented in all groups and therefore not included in the statistical model. However, T-wave inversions in the precordial leads cannot be counted in children ≤ 14 years of age, because this is a nonspecific finding at this age (³). The authors should specify an age cutoff in their analyses. Genetic testing was only performed in 49 of 142 study subjects. Less specific 1994 TFC were used for endomyocardial biopsy. Finally, the authors combined patients who met the TFC (4

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diagnostic points) with patients who had only 3 diagnostic points. We do not think that patients with 3 points should be classified as definite ARVD/C, especially in family members who receive a major criterion (2 points) merely by their family history (³).

In our own transatlantic series of >1,000 patients with ARVD/C, only 4 of 416 probands were present in patients <13 years of age (⁴). In our experience, children with ARVD/C typically have electrocardiographic and Holter abnormalities before CMR changes. Structural abnormalities in children with ARVD/C are often mild, and involve focal subtricuspid dyskinesia with preserved global function rather than severe RV enlargement. As such, we do not routinely advise CMR in children <10 years of age. In addition, a high percentage of false positive interpretation of CMRs from referring centers in the North American study suggests that misinterpretation of CMRs is common (²).

Nevertheless, Etoom et al. (¹) have raised an important question of applicability of revised TFC in children. Their patient population is worthy of more in-depth study due to the early age at which they have expressed disease, perhaps representing an aggressive form of disease that is otherwise not commonly seen.

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