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The predictive value of normal CMR scans in patients with suspected ARVC - an outcomes study

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Objective

To assess the diagnostic accuracy of CMR scans in predicting future major cardiovascular adverse events in patients with suspected arrhythmogenic right ventricular cardiomyopathy (ARVC).

Background

ARVC is a genetic disease that affects young patients prone to sudden cardiac death. The incomplete penetrance of this disorder with variable age of onset means there is a large heterogeneity in the clinical expression of the disease with strikingly different phenotypes. It is therefore crucial to identify within a population with suspected ARVC, a subset who will definitely have a higher susceptibility of developing the disease, and thus be at an increased risk of major adverse cardio-vascular events (MACE).

Methods

We studied 379 consecutive patients referred for CMR examinations with suspected ARVC based on at least one minor criterion as defined by the Task Force Criteria, between 2002 and 2005. Patients were divided into 2 groups depending on their initial scan results: "normal" or "abnormal". The latter was defined as either increased RV end-diastolic volume, and/or decreased RV ejection fraction, and/or RV regional wall motion abnormality, and/or the presence of late gadolinium enhancement in the LV and/or RV. Cut-off values for normal versus abnormal volumes and ejection fraction were based on previ-

ously published reference ranges indexed for BSA and adjusted for age, from our institution. Follow-up data was obtained for all patients in 2009 and reviewed by two independent researchers. MACE were defined as all cause-mortality, occurrence of life-threatening arrhythmias, appropriate ICD discharge, and unplanned hospital admission for cardiovascular events. Positive and negative predictive values of abnormal and normal scans for MACE were calculated.

Results

223 patients (59%) had normal baseline scans. Both groups were well matched in their clinical presentations (VT episodes, unexplained syncope, family history including sudden cardiac death). Patients with abnormal scans were older (47.4 yrs \pm 14.5) and more often male (64.1%) than those with normal scans (40.9 yrs \pm 16.1, 44.8% males). The overall negative predictive value of a normal scan for MACE was 89%; the positive predictive value of an abnormal scan was 19%. This was statistically significant (p = 0.032), translating into an odds ratio for an abnormal scan of 1.89 (95% CI 1.05-3.40) in predicting MACE.

Conclusion

The data confirm the value of CMR in a "real-life" patient population referred for suspected ARVC. A normal CMR scan, regardless of whether the diagnosis of ARVC is con-

firmed or not, allows to reliably predict a long-term major cardiovascular event-free outcome.

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