

Cardiac Magnetic resonance in clinical diagnosis of left dominant phenotypes of arrhythmogenic cardiomyopathy

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Recently, the spectrum of arrhythmogenic cardiomyopathy (AC) has been expanded with newly described biventricular (BVAC) and left-dominant (LDAC) forms. Cardiac magnetic resonance (CMR) with its superb tissue discrimination abilities, and high reproducibility for ventricular volume calculation, offers potentially relevant information for their diagnosis.

Objective: To analyze the diagnostic performance of different CMR parameters in the diagnosis of AC.

Methods: Patients included in this study come from a prospective protocol evaluating cases of familiar cardiac sudden death (SD), with a clinical or anatomopathological diagnosis of AC in the index case. All patients underwent the following diagnostic tests: EKG, cycloergometry, Holter study, echocardiography, CMR, and genetic study specific for AC. Diagnosis was based on Task Force criteria in patients without the mutation, while in carriers, the presence of findings suggestive of AC in at least two diagnostic tests of different categories was considered as diagnostic.

The diagnostic value of the following CMR parameters was evaluated: 1- presence of late gadolinium enhancement (LGE) in left ventricle (LV) and right ventricle (RV); 2- LV ejection fraction (LVEF) $\leq 55\%$ and RV EF $\leq 45\%$, and 3- biventricular dilatation (LVEDVi $\geq 98\text{ml/m}^2$ and RVEDVi $\geq 100\text{ml/m}^2$ in male or 90ml/m^2 in female)..

Results: The group comprised 59 patients: 49% male. There were 5 index cases (3 resuscitated, and 2 with cardiogenic syncope and no SD), and 54 first-degree relatives. AC was diagnosed in 10 pts (17%), 6 (60%) with LDAC and 4 (40%) with BVAC. The diagnostic value of LGE was, in the LV (Sensitivity 100%, Specificity 94%), and in the RV (sensitivity 20%, specificity 97%); for LVEF (Sensitivity 60%, specificity 100%), for RVEF (sensitivity: 30%; specificity: 97%), and for RV dilatation (Sensitivity: 10%, Specificity: 94%). No patient showed LV dilatation

Conclusion: 1- In this clinical setting, with high prevalence of left and biventricular forms of AC, left ventricular delayed gadolinium enhancement is the parameter with the highest diagnostic performance. 2- The other parameters are very specific, though poorly sensitive for diagnosis.

LEFT VENTRICLE	S	E	PPV	NPV
LVEDVi $> 98\text{ml/m}^2$	0	0	0	83%
LGE LV	90 %	93%	75%	97%
EF $< 55\%$	60 %	100%	60%	90%
IMPAIRED SEGMENTAL CONTRACTILITY	10%	97%	10%	80%
RIGHT VENTRICLE	S	E	PPV	NPV
RVEDVi $> 100\text{ml/m}^2$	10 %	94 %	33 %	80 %
LGE RV	20 %	97 %	66 %	81%
EF $< 45\%$	30 %	97%	40 %	100 %
IMPAIRED SEGMENTAL CONTRACTILITY	60 %	91 %	66 %	82 %