Simultaneous Use of Double-IR and Triple-IR FSE Imaging Techniques Is a Sensitive Approach for Diagnosis and Early Detection of Arrhythmogenic Right Ventricular Cardiomyopathy

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Introduction: Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC) is a familial and sporadic cardiomyopathy with unknown causes. Pathologically, the disease is characterized by the replacement of the right ventricular (RV) muscle by adipose and fibrous tissue, leading to an enlargement of the RV chamber, RV muscle dysplasia, regional RV bulging, and decrease of RV function. At present, establishment of ARVC diagnosis requires a demonstration of RV enlargement, compromised RV function, RV bulging/aneurysm, and ECG abnormalities. Pathological diagnosis of the disease requires confirmation of the replacement of RV muscle by adipose and fibrous tissue. The diagnostic tools presently available for *in vivo* assessment lack sufficient sensitivity and specificity to ARVC. As such, it is clinically significant to develop a reliable method for diagnosis and early detection of the disease. It has been shown that double inversion-recovery fast spin echo (DIR-FSE) imaging clearly delineates anatomic structure of the RV while triple inversion-recovery fast spin echo (TIR-FSE) technique highlights the ventricular muscle, leaving the areas of fibroadipose tissue as dark regions. Simultaneous use of the two imaging techniques is therefore able to provide information about anatomic structure and tissue components of the RV wall. The present study was to determine whether simultaneous use of the two imaging techniques help in diagnosis and early detection of ARVC.

Materials and Methods: Ten ARVC patients and 12 symptom-free familial members (with only occasional ECG abnormalities) underwent MRI studies. Diagnosis of ARVC was established based on patient history, clinical symptoms, and the findings of 24-hr ambulatory electrocardiography (ECG), ventricular late potential, ventriculography, nuclide myocardial visualization, and echocardiography. MR imaging was performed on GE SIGNA Horizon CV/I 1.5 Tesla system. DIR-FSE, TIR-FSE, and FastCine images were acquired in the short cardiac-axis plane, LV outflow-tract long cardiac-axis plane, and four-chamber plane. Some of our imaging parameter are: slice thickness, 8 mm; field of view, 34 x 34 cm; imaging matrix, 256 x 224; TR, 2 R-R intervals for DIR- and TIR-FSE; TE, 4.0 ms and 60-80 ms for DIR-FSE and TIR-FSE, respectively.

Results: We found on DIR-FSE images that the RV wall was much brighter (high fat signal) than the LV wall only in three patients (Figure 1-A). In the remaining patients the signal intensities of the RV walls were similar to those of the LV wall. On TIR-FSE images, on the other hand, an irregular and discontinuous RV wall was observed in nine patients (Figure 1-B). The RV wall appeared significantly thinner on the TIR-FSE images than on the DIR-FSE images. Both DIR- and TIR-FSE images show an enlarged RV chamber in six patient patients (diastolic diameter, 5.9 ± 1.2 cm x 8.5 ± 0.8 cm). Decreased RV ejection fraction (EF) was found in six patients. Three patients showed regional RV bulging on FastCine images. The incidences of the MRI findings are listed in Table 1. In addition, among the 12 "healthy" familial members, eight of them showed a thin and discontinuous RV wall and only four members exhibited an enlarged RV chamber.

Discussion and Conclusions: Although ARVC is pathologically due to the replacement of the RV muscle by adipose and fibrous tissue, we found that only small percent of patients (3/10) showed high fat signal on DIR-FSE images. This suggests that normal signal intensity of the RV wall cannot exclude the possibility of ARVC. As mentioned above, six patients (6/10) showed an enlarged RV chamber, indicating that RV enlargement may not be a prerequisite criterion for establishment of the diagnosis either. However, majority of the patients (9/10) and their "healthy" familial members (8/12) exhibited thin and irregular RV wall on the TIR-FSE images. This suggests that thin and irregular RV wall may be the most sensitive parameter for diagnosis and early detection of the disease. We also found in this study that the muscular layer of the RV wall could be readily delineated by comparative analysis of both DIR- and TIR-FSE images. Thus, we conclude that simultaneous use of the two imaging techniques is a logical and sensitive approach for diagnosis and early detection of the disease.

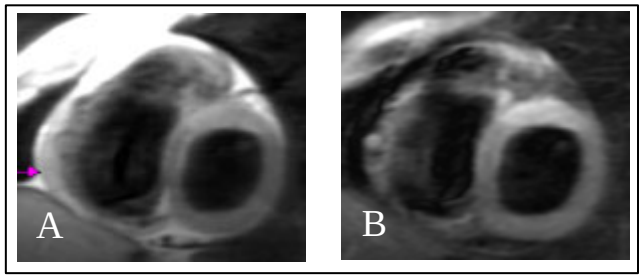

Table 1. Incidence of the MRI findings

MRI findings	# of 10	MR Methods
Thin and irregular RV wall	9	TIR-FSE
RV enlargement	6	DIR- & TIR-FSE
Reduced RV EF	6	FastCine
High fat signal in RV	3	DIR-FSE
RV bulging	2	FastCine

Figure 1. Short-axis MR images acquired with DIR-FSE (A) and TIR-FSE (B) from an ARVC patient. Both images show an enlarged RV chamber. On the TIR-FSE image the RV wall appears discontinuous with rough edges on both sides.