

The role of Magnetic Resonance Imaging in the morpho-functional study of the right ventricle and in the correct diagnosis of a case of Arrhythmogenic Cardiomyopathy: a case study

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KEYWORDS:

Arrhythmogenic right ventricle dysplasia, ARVD, CMR. End diastolic volume, ejection fraction

ABSTRACT

Arrhythmogenic Right Ventricular dysplasia (ARVD) is an inherited disease characterized by fibroadipose replacement of cardiac tissue, which can lead to severe arrhythmias and sudden cardiac death, especially in young athletes. This experimental study analyzes the efficacy of Magnetic Resonance Imaging (MRI) in the early diagnosis and monitoring of ARVD, highlighting how this diagnostic technique represents the gold standard for the morpho-functional evaluation of the right ventricle. 43 patients were examined, one of whom tested positive for ARVD, thanks to the diagnostic parameters obtained (RVEDVi, RVESVi and FE) and Delayed enhancement sequences by MRI. It was found that the reference parameters of the TASK FORCE 2010 were exceeded with values of RVEDVi equal to 110 ml/m² and FE equal to 36% and in the DE sequences we had an accumulation of contrast medium in the free wall of the right ventricle.

INTRODUCTION

Arrhythmogenic Right Ventricular Cardiomyopathy (ARVD) is an inherited disorder that can cause ventricular dysfunction, severe arrhythmias, and sudden cardiac death [1-3]. Early diagnosis is crucial to avoid serious clinical consequences. ARVD is found in up to 20% of individuals who experience sudden cardiac death (SCM) before the age of 35 and is even more common among athletes who die suddenly. It has been reported that the disease has a prevalence of 1 in 2000-5000 people, although according to some studies the true prevalence may be 1 in 1000 in some regions of the world [4]. It has been observed that the incidence of sudden cardiac death among young competitive athletes has decreased significantly in the Veneto region of Italy following the introduction of a nationwide systematic screening program. This program, which aims to identify potentially life-threatening cardiac conditions early, has been a major contributor to the prevention of fatal events. The reduction in mortality has been attributed primarily to a lower frequency of sudden death related to cardiomyopathies, often silent conditions that, thanks to extensive monitoring, have been increasingly detected in athletes during preparticipation screening. This early identification has made it possible to intervene with therapies or preventive measures, thus reducing the risk of dramatic events during sports competition

[10]. Cardiac MRI represents the gold standard for the evaluation of the morphology and function of the right ventricle, as it allows a detailed study of ventricular volumes and ejection fraction as well as the analysis of DE sequences that allow to evaluate the accumulation or not of the contrast medium, fundamental parameters for the diagnosis of ARVD [5]. Ventricular volumes and ejection fraction are compared with the reference values of the 'Task Force 2010' [6]. The objective of this study is to refine the study protocol of right cardiac sections, and to validate the normal values, contained in the 2010 Task Force, of right ventricular volume and function to increase the diagnostic sensitivity of right ventricular diseases such as ARVD

MATERIALS AND METHODS

The study was conducted on 43 randomly selected patients on the basis of those who were to have cardiac MRI, divided by sex, aged 18 to 69 years (Fig. 1), at the ASL BT P.T.A. "San Nicola Pellegrino" in Trani, using a "SIEMENS MAGNETOM Aera 1.5 Tesla" MRI equipment, during a period between March and September 2024. A specific protocol with fixed parameters of TR, TE and layer thickness was used for each patient (Table 1). The parameters obtained from the volumetric and functional sequences of the right ventricle (end-diastolic volume, end-systolic



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PATHY"

HTTPS://DOI.
ORG/10.36017/
JAHC202571394

RECEIVED: 2024-11-10
REVISED: 2025-01-17
ACCEPTED: 2025-01-20
PUBLISHED: 2025-01-21



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volume, and ejection fraction) were analyzed, with particular attention to the presence of areas of fibrosis detected by Delayed Enhancement technique, injecting a Gadolinium-based contrast agent with a

value of 0.2 ml/kg. [The results were compared with the normal values imposed by the 2010 Task Force [6].

A	B	C	E	F	G
RVEDVi ml/m ² (M)	RVESVi ml/m ² (M)	Frazione Eiezione (M)	RVEDVi ml/m ² (F)	RVESVi ml/m ² (F)	Frazione Eiezione (F)
38	17	56%	40	18	56%
35	14	61%	71	25	64%
57	30	47%	77	29	62%
36	18	48%	63	22	65%
60	29	52%	83	27	68%
47	13	72%	37	15	59%
88	28	69%	43	13	69%
88	29	67%	71	17	77%
51	19	63%	58	25	57%
91	35	61%	54	25	54%
88	38	57%	54	20	63%
98	43	56%	64	23	64%
62	25	60%	64	26	60%
47	18	62%	42	13	68%
50	22	57%	26	13	50%
78	33	57%			
64	33	48%			
61	23	62%			
61	26	58%			
67	25	63%			
54	20	63%			
67	27	65%			
53	24	55%			
40	12	70%			
53	23	56%			
46	22	51%			
71	23	68%			
110	70	36%			

Figure 1. Data collection by gender

Table 1. Study Protocol sequences and parameters

Sequence	TR (ms)	TE (ms)	Slice thickness (mm)	TI (ms)
CINE_2 CHAMBERS	36,66	1,17	6.0	/
TRUFL_SHORTAXIS_IPAT	275,34	1,12	8.0	/
CINE_4 CHAMBERS	35,36	1,15	6.0	/
CINE_3 CHAMBERS	35,36	1,15	6.0	/
CINE_AFFL_EFFL	35,36	1,15	6.0	/
SOMM. MDC	/	/	/	/
CINE_ASSE CORTO_CINE 10SL FA 70	50,40	1,17	8.0	/
AFTER 8' FROM SOMM. MDC	/	/	/	/
TI-SCOUT (SHORT AXIS)	23,58	1,11	8.0	/
DE_OVEVIEW_TFL_PSIR_10SL_I PAT	700	1	8.0	chosen by radiologist and cardiologist
DE_OVERVIEW_TFL_PSIR_SIS TOLIC	lowest possible	1	6.0	chosen by radiologist and cardiologist
DE HIGH-RES_PSIR_2 CHAMBERS	700	1,15	8.0	chosen by radiologist and cardiologist +5 ms
DE HIGH-RES_PSIR_4 CHAMBERS	700	1,15	8.0	chosen by radiologist and cardiologist +5 ms
DE HIGH-RES_PSIR_3 CHAMBERS	700	1,15	8.0	chosen by radiologist and cardiologist +5 ms
DE HIGH- RES_PSIR _AFFL_EFFL	700	1,15	8.0	chosen by radiologist and cardiologist +5 ms



RESULTS

Out of 43 patients examined, one case of ARVD was found in a 50-year-old male patient (highlighted in red in Fig. 1). The volume values, obtained on the basis of cine (Fig. 2), end-diastolic (RVEDVi) and end-systolic (RVESVi) images indexed to the body surface area exceed the normal limits established by the 2010 Task, with an ejection fraction (EF) of

less than 45%. The images obtained by MRI also showed the presence of fibrotic areas through the use of gadolinium (Fig 3). The data obtained were then mathematically represented through scatter plots (Fig. 4). The graphs confirm that all values respect a certain relationship except for an outlier which represents the positive case.

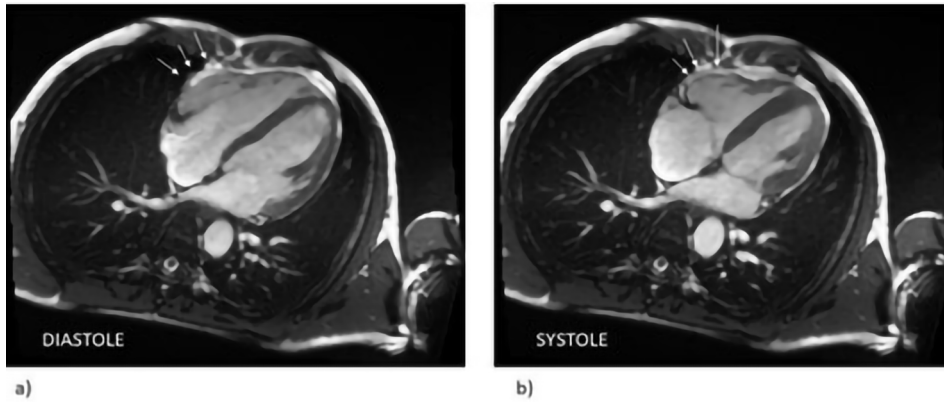


Figure 2. Cine-SSFP 4-chamber sequences in CMR show significant dilatation and moderate dysfunction of the right ventricle due to abnormalities mainly involving the basal part of the free wall of the right ventricle (white arrows) in both diastolic (a) and systolic (b).

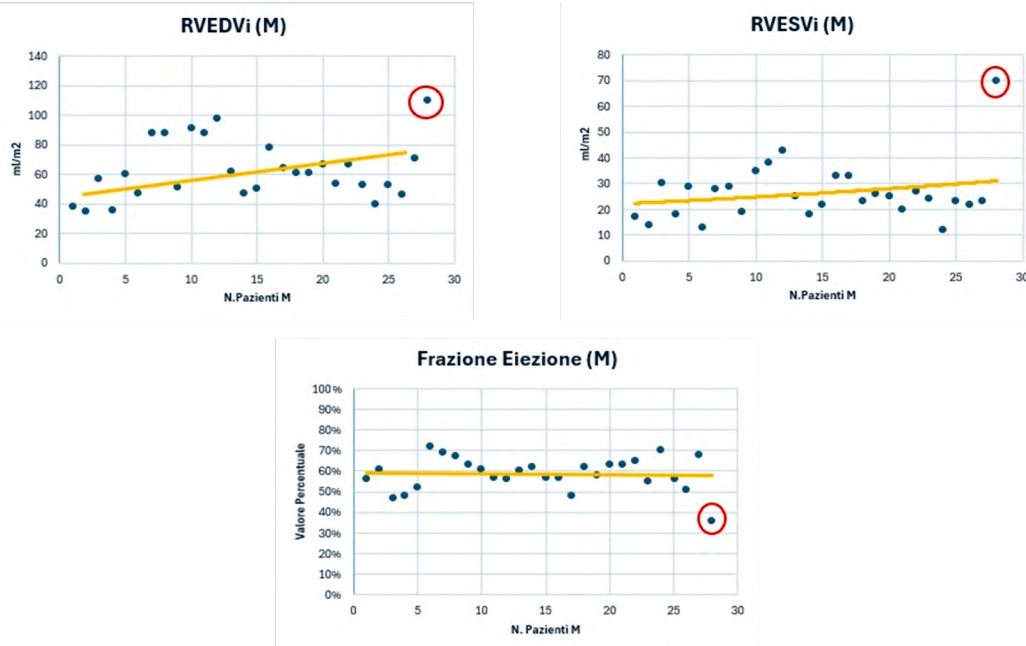


Figure 3. Scatter charts

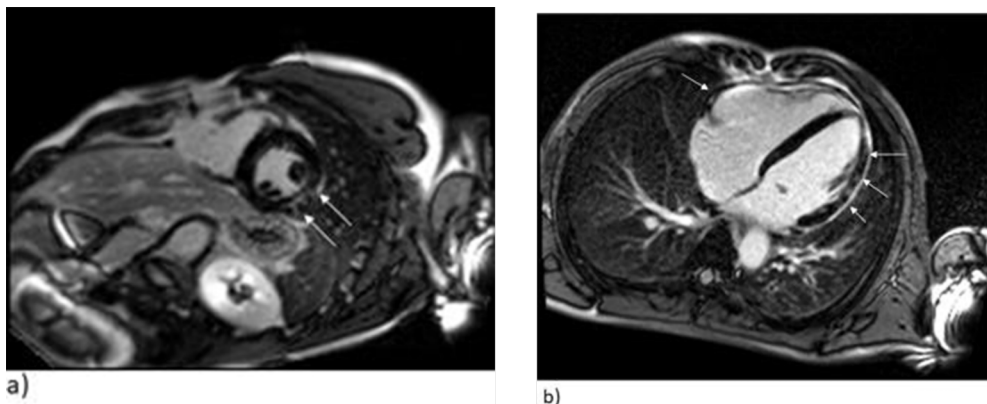


Figure 4. LGE sequences showing enhancement with a “patchy” distribution in the lateral wall of left ventricle (a) and right ventricular free wall (b) indicated by white arrows.



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DISCUSSION

The results obtained confirm the usefulness of cardiac MRI as a method of first choice for the diagnosis and monitoring of ARVD. The ssfp-cine sequences, which allow the detailed analysis of the morphology and kinetics of the right ventricle [9], were essential to quantify the ejection fraction in addition to the end-diastolic and end-systolic volume that detected segmental abnormalities of contractility at the level of the free wall of the right ventricle. The delayed enhancement sequences showed an accumulation of gadolinium in the lateral wall of the left ventricle, while in the right ventricle a pattern of "patchy" distribution was observed at the level of the middle and sub-epicardial wall, and in the free wall at the basal level, corresponding to dyskinesia. Compared to echocardiography, MRI offers a higher level of detail

in the evaluation of the right ventricle, a complex structure that is often difficult to assess with other imaging techniques. The results of the study show that MRI is able to detect alterations that escape other diagnostic methods, making it an essential tool for the early and accurate identification of ARVD. This is particularly important in young and asymptomatic patients, in whom timely diagnosis can prevent disease progression and reduce the risk of potentially fatal arrhythmic events.

CONCLUSIONS

Our study made it possible to refine the examination protocol in cardio-RM of the right ventricle by confirming normal parameters of size and function thus enhancing the sensitivity in the diagnosis of rare cardiomyopathies such as ARVD.

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