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MR Imaging of Arrhythmogenic Right Ventricular Cardiomyopathy: Morphologic Findings and Interobserver Reliability

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Key Words

Magnetic resonance imaging · Diagnosis · Cardiomyopathy · Right ventricle

Abstract

Background: Magnetic resonance (MR) imaging is frequently used to diagnose arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D). However, the reliability of various MR imaging features for diagnosing ARVC/D is unknown. The purpose of this study was to determine which morphologic MR imaging features have the greatest interobserver reliability for diagnosing ARVC/D. ***Methods:*** Forty-five sets of films of cardiac MR images were sent to 8 radiologists and 5 cardiologists with experience in this field. There were 7 cases of definite ARVC/D as defined by the Task Force criteria. Six cases were controls. The remaining 32 cases had MR imaging because of clinical suspicion of ARVC/D. Readers evaluated the images for the presence of (a) right ventricle (RV) enlargement, (b) RV abnormal morphology, (c) left ventricle enlargement, (d) presence of high T_1 signal (fat) in the myocardium, and (e) location of high T_1 signal (fat) on a Likert scale with formatted responses. ***Results:*** Readers indicated that the Task Force ARVC/D cases had significantly more (χ^2 = 119.93, d.f. = 10, $p < 0.0001$) RV chamber size enlargement (58%) than either the suspected ARVC/D (12%) or no ARVC/D (14%) cases. When readers reported the RV chamber size as enlarged they were significantly more likely to report the case as ARVC/D present (χ^2 = 33.98, d.f. = 1, $p < 0.0001$). When

[1] Readers of the MR studies.

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readers reported the morphology as abnormal they were more likely to diagnose the case as ARVC/D present (χ^2 = 78.4, d.f. = 1, $p < 0.0001$), and the Task Force ARVC/D (47%) cases received significantly more abnormal reports than either suspected ARVC/D (20%) or non-ARVC/D (15%) cases. There was no significant difference between patient groups in the reported presence of high signal intensity (fat) in the RV (χ^2 = 0.9, d.f. = 2, $p > 0.05$). ***Conclusions:*** Reviewers found that the size and shape of abnormalities in the RV are key MR imaging discriminates of ARVD. Subsequent protocol development and multicenter trials need to address these parameters. Essential steps in improving accuracy and reducing variability include a standardized acquisition protocol and standardized analysis with dynamic cine review of regional RV function and quantification of RV and left ventricle volumes.

Introduction

Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D) is a condition in which the right ventricular muscle undergoes replacement by fatty and/or fibrous tissue that is interspersed among the remaining muscular fibers [1, 2]. This forms a substrate for reentrant ventricular arrhythmias. ARVC/D presents most frequently in men between the ages of 15–40 with premature ventricular beats and nonsustained or sustained ventricular tachycardia of left bundle branch block morphology [3]. Though rare, sudden cardiac death may be the first manifestation of this disease. It must be included in the differential diagnosis of right ventricular outflow tract (RVOT) tachycardia, which is the most common cause of ventricular tachycardia in young people without obvious structural heart disease. It is important to establish the correct diagnosis since RVOT tachycardia is generally not a progressive condition, seldom a cause of arrhythmic death and not known to be transmitted genetically [4]. In contrast, ARVC/D may be progressive and may cause arrhythmic death. In addition, since ARVC/D may be familial (30–50% of cases) [3], it is important to correctly diagnose this disease in the proband in order to know if familial screening should be undertaken.

The diagnosis of ARVC/D is based on the identification of structural abnormalities, fatty replacement of the right ventricular myocardium, electrocardiographic changes, arrhythmias of right ventricular origin, and history of familial disease. Because of the difficulty in diagnosing ARVC/D with certainty, a task force was convened that proposed diagnostic criteria based on these abnormalities [6]. Individual criteria were listed as major or minor and combinations of these were judged to indicate the presence of this disease. Included among the major criteria were severe or marked structural abnormalities of the right ventricle (RV) and fibro-fatty replacement of the myocardium on endomyocardial biopsy. The Task Force stated that experience with MR imaging in the diagnosis of ARVC/D was limited and required further evaluation.

MR imaging is often used in the diagnostic evaluation of ARVC/D because it is noninvasive and readily depicts *morphologic* abnormalities of the RV, including right ventricular chamber size or aneurysm, as well as *functional* abnormalities of the RV using MR cine techniques [7–12]. MR cine images, however, lack standardization and many MR centers do not have the capability to transmit or display cine images for purposes of referral to tertiary medical centers. Morphologic MR imaging using spin-echo technique is widely available and offers excellent soft-tissue contrast resolution. In particular, fat is depicted with high signal intensity on T_1-weighted MR images, whereas the myocardium has an intermediate signal. The presence of myocardial fat identified by MR imaging has been reported to be strongly associated with ARVC/D [5, 7–12]. The reliability and interobserver agreement of these MR imaging features, however, has never been evaluated in a multi-center trial. The purpose of this study was to determine the reliability of various *morphologic* MR imaging features for diagnosing ARVC/D and to determine interobserver variability for these MR features.

Methods

Hardcopy films of 39 patients were obtained from the ARVC/D registry at the University of Arizona Health Sciences Center. Only 'static' MR images were evaluated since cine 'functional' MR images were not available from most of the referring hospitals. Twelve of the MR examinations were performed at university medical centers, 21 at private hospitals and 6 at imaging centers. The MR studies were performed in 17 states and in Canada between the years 1994 and 2000 (median 1998). In addition, hardcopy films of 6 cardiac MR examinations that were *not* performed for suspected ARVC/D were also randomly selected from the film library at the University of Arizona Medical Center. These cases were designated as control cases for the blinded readings. The indications for performing these MR studies included anomalous coronary artery, cardiac arrest, aortic dissection, hypertension, stroke, and pectus excavatum. The MR scanners were 1.5 T General Electric scanners (34 imagers), a 1.5 T Philips scanner (1), 1.5 T Siemens scanners (6), and 0.5 T General

Table 1. Questions and possible answers on the pre-formatted response sheet

1 *Is the study diagnostic for ARVC/D?*					
0	1	2	3	4	5
Definitely no	Probably no	Possibly no	Possibly yes	Probably yes	Definitely yes
2 *Is there evidence of fat in the myocardium?*					
0	1	2	3	4	5
Definitely no fat	Probably no fat	Possibly no fat	Possibly fat	Probably fat	Definitely fat
3 *Circle areas where fat is located.*					
0	1	2	3	4	
Anterior wall	RV apex	RV RVOT	RV septum	LV myocardium	
4 *RV chamber size?*					
0	1	2	3	4	5
Definitely normal	Probably normal	Possibly normal	Possibly enlarged	Probably enlarged	Definitely enlarged
5 *RV configuration, morphology, shape (aneurysm, muscle bundles, scalloping, trabeculation)?*					
0	1	2	3	4	5
Definitely normal	Probably normal	Possibly normal	Possibly abnormal	Probably abnormal	Definitely abnormal
6 *LV chamber size?*					
0	1	2	3	4	5
Definitely normal	Probably normal	Possibly normal	Possibly enlarged	Probably enlarged	Definitely enlarge
7 *How is overall image quality?*					
0	1	2	3		
Poor	Fair	Good	Excellent		
8 *Are motion artifacts present?*					
0	1	2	3		
None present	Present: mild	Present: moderate	Present: severe		
9 *Are there other artifacts?*					
0	1	2	3		
No artifacts	Yes: mild	Yes: moderate	Yes: severe		

Electric scanners (4). All MR scanners had ECG gating capability. None of the MR scanners were 'dedicated' cardiovascular MR scanners.

Each set of 45 MR films was sent to 8 radiologists and 5 cardiologists with expertise in cardiovascular MR imaging. No clinical information was provided, other than that the reader was to evaluate the films for the MR imaging findings of ARVC/D. All readings were conducted independently. A preformatted response sheet accompanied each case. There were nine questions with Likert-scale formatted responses to choose from (table 1). The readers circled the appropriate response based on their interpretation of the images. Five morphologic descriptors previously described to be associated with ARVC/D were evaluated by the readers: (a) enlargement of the RV; (b) morphologic abnormality of the RV (e.g., aneurysm, scalloping, increased trabeculation); (c) enlargement of the left ventricle (LV); (d) presence of high T_1 signal (fat) in the myocardium, and (e) location of high T_1 signal (fat). An overall impression as to the presence or absence of ARVC/D was recorded. Overall image quality and the presence or absence of image artifacts was also recorded.

Adjudication of Patient Diagnoses

All patients referred for the evaluation of ARVC/D were evaluated according to ARVC/D Task Force guidelines by a single reviewer (F.M.) [7, 13, 14]. Patients were then classified as either 'suspected ARVC/D' or 'definite' ARVC/D patients. Suspected ARVC/D patients had been referred for possible ARVC/D but these patients did not meet Task Force guidelines and ultimately were not diagnosed as ARVC/D. Definite ARVC/D patients met Task Force guidelines for the diagnosis. Finally, a 'control' group of patients who had undergone MR imaging for reasons other than ARVC/D was included. MR imaging results were not used for adjudication to classify patients into any of these categories.

Of the 45 patients evaluated, 6 (13%) were control cases, 32 (71%) were suspected ARVC/D cases and 7 (16%) were definite ARVC/D cases. Since there were 13 readers evaluating the 45 cases (585 total reads), 78 reads were of the control cases, 416 reads were of the suspected ARVC/D cases and 91 reads were of the definite ARVC/D cases. Readers were allowed to not score MR findings if they felt they were unable to assess that specific finding (e.g., due to image artifacts). Thus, in the Results section below, the total number of reads in each category is less than above totals.

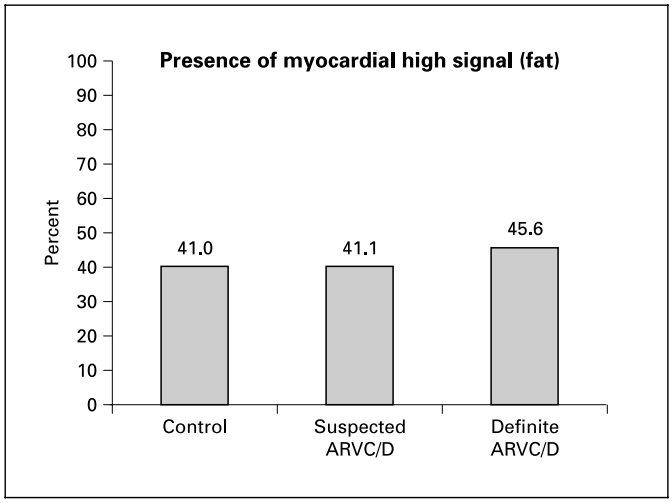

Fig. 1. Percentage of readings that identified increased T_1-signal intensity (corresponding to fat signal) in the RV in the control group, the suspected and the definite ARVC/D group. There is no significant difference between the groups ($p > 0.05$).

Statistical Analysis

A z-test for proportions [15] was used to compare the proportions of ARVC/D image findings present versus ARVC/D findings absent for the control cases, suspected ARVC/D cases and definite ARVC/D cases. χ^2 analysis [15] was used to compare the presence or absence of morphologic features between the three groups of cases. p values less than 0.05 were defined as statistically significant. κ analysis was performed to measure the overall rate of diagnostic agreement between the 13 readers on all 45 cases [16]. All statistical analyses were carried out using StatView version 5.0 software (SAS Institute Inc., Cary, N.C., USA).

Results

MR Imaging Findings

The presence of high signal (equal to fat signal intensity) on T_1-weighted images in the RV has been reported to be associated with ARVC/D [5, 8–13]. Readers reported no significant difference between patient groups in the reported presence of high signal intensity in the RV (fig. 1; 32/78 control cases, 159/397 suspected ARVC/D cases, 41/90 definite ARVC/D cases; $\chi^2 = 0.9$, d.f. = 2, $p > 0.05$). The location of the reported high T_1 signal (e.g., anterior wall, right ventricular outflow tract, apex) was not related to whether the case was reported as positive or negative for ARVC/D ($\chi^2 = 22.98$, d.f. = 14, $p > 0.05$). When a reader detected high T_1 signal in the RV, this was highly associated with that reader's final diagnosis of ARVC/D present (188/217 reads, 87%) rather than ARVC/D absent (44/348 reads, 13%; $\chi^2 = 304$, d.f. = 1, $p < 0.0001$).

RV chamber enlargement and morphology were assessed by the readers. Readers found both of these morphologic features to be present more frequently in definite ARVC/D cases than in control or suspected ARVC/D groups (fig. 2, 3). RV enlargement was present in 11/78 (14%) control reads, 50/394 (13%) suspected and 52/89 (58%) definite ARVC/D reads ($\chi^2 = 96.4$, d.f. = 2, $p < 0.0001$). Abnormal right ventricular morphology was present in 12/76 (16%) control reads, 84/395 (21%) suspected and 42/89 (47%) definite ARVC/D reads ($\chi^2 = 29.83$, d.f. = 2, $p < 0.0001$).

When a reader diagnosed right ventricular enlargement, this was associated with that reader's final diagnosis of ARVC/D present (70/218 reads, 32%) rather than ARVC/D absent (43/343 reads, 13%; $\chi^2 = 31.60$, d.f. = 1, $p < 0.0001$). Similarly, abnormal right ventricular morphology detected by the reader was also associated with that reader's final diagnosis of ARVC/D present (105/218 reads, 48%) rather than ARVC/D absent (33/342, 10%; $\chi^2 = 105.1$, d.f. = 1, $p < 0.0001$).

Readers also assessed the size of the LV. Left ventricular size was judged by the readers to be similar in all groups (fig. 4). Normal LV size was present in 63/76 (83%) control reads, 356/393 (90%) suspected and 80/87 (92%) definite ARVC/D reads ($\chi^2 = 4.22$, d.f. = 2, $p > 0.05$).

Reader Agreement: Final ARVC/D Diagnoses

Based on all of the morphologic MR findings (functional cine images were not available), readers were asked for a final classification of ARVC/D as present or absent (fig. 5). For control cases, readers correctly classified 52/78 (67%) of cases as ARVC/D absent versus 26/78 (33%) as ARVC/D present ($z = 2.86$, $p < 0.01$). For suspected ARVC/D cases, readers classified 157/416 (38%) of cases as ARVC/D absent versus 259/416 (62%) as ARVC/D present ($z = 4.75$, $p < 0.01$). For definite ARVC/D cases, readers classified 41/91 (47%) of cases as ARVC/D absent versus 50/91 (53%) as ARVC/D present ($z = 0.57$, $p > 0.05$). Overall reader agreement in the final classification of ARVC/D present or absent was very poor ($\kappa = 0.017$).

Reader agreement for individual MR imaging features was evaluated (fig. 6–9). In these figures, the minimum and maximum rates of agreement with the final clinical diagnosis of each case are shown within each category (control, suspected ARVC/D, definite ARVC/D). For example, for high T_1 signal in the myocardium, figure 6 shows that for the control group, 12/13 (92%) readers

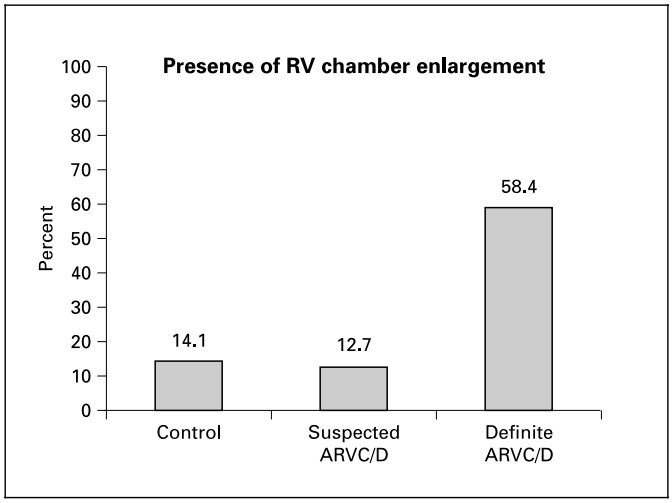

Fig. 2. Percentage of readings that identified enlargement of the RV in the control group, the suspected and the definite ARVC/D group. The differences between the groups was statistically significant ($p < 0.0001$).

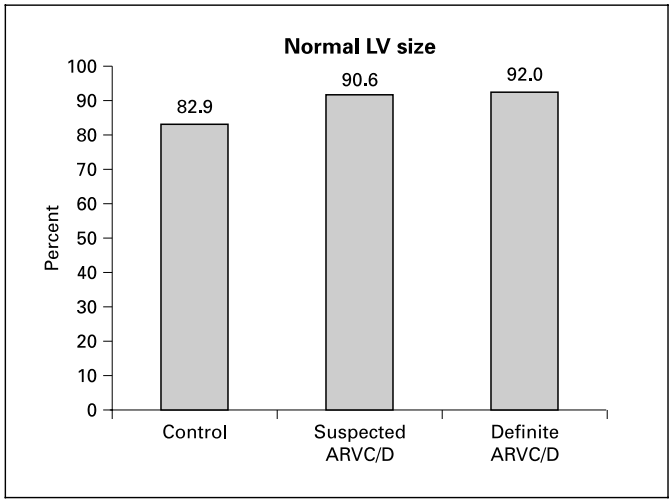

Fig. 4. Percentage of readings that identified normal left ventricular (LV) size in the control group, the suspected and the definite ARVC/D group. The differences between the groups was not statistically significant ($p > 0.05$).

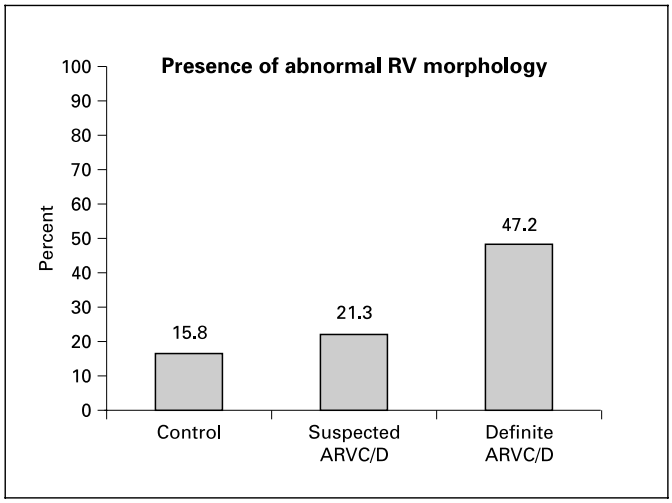

Fig. 3. Percentage of readings that identified abnormal morphology of the RV in the control group, the suspected and the definite ARVC/D group. The differences between the groups was statistically significant ($p < 0.0001$).

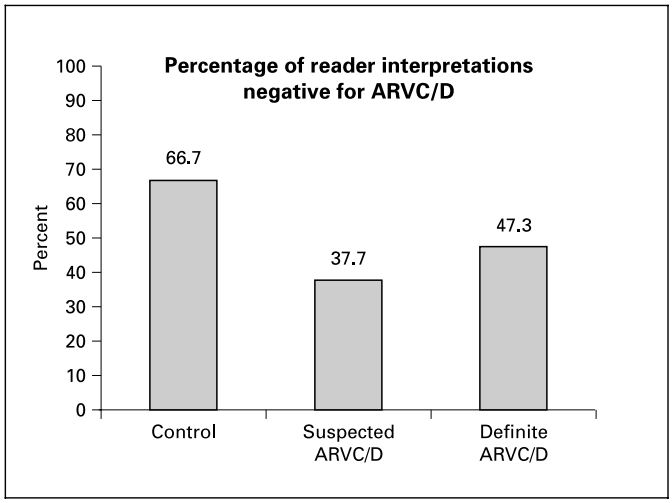

Fig. 5. Percentage of readings that defined ARVC/D as absent, in the control group, the suspected and the definite ARVC/D group.

agreed that fat was present in the RV myocardium in 1 of the patients. In another patient in the control group, only 1/13 readers (8%) identified the presence of fat. Since the maximum agreement rates between the control group and the patient group are very high and quite similar for fat signal in the myocardium (92% control group and suspected ARVC/D groups, 85% definite ARVC/D group), this suggests that (1) a fat signal was present in all the groups and (2) the fat signal did not discriminate between subgroups. The differences between the maximum and

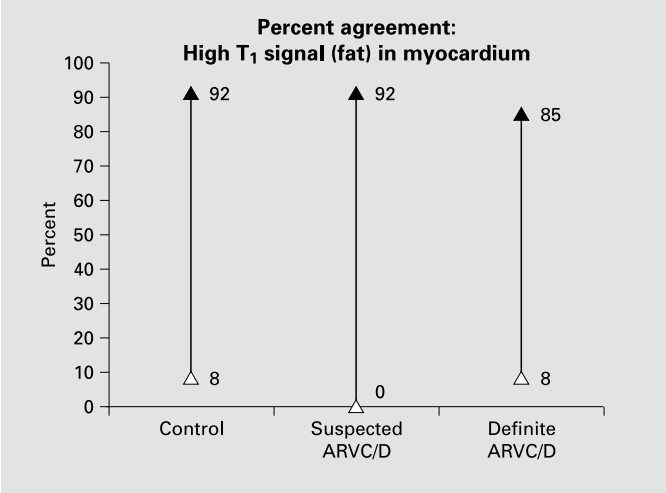

Fig. 6. Percentage reader agreement for identification of increased T_1-signal intensity (corresponding to fat signal) in theRV. The minimum (△) and maximum (▲) rates of agreement *on a per-patient basis* are shown for the control group, the suspected and the definite ARVC/D group. For example, for one patient in the control group, 12/13 (92%) readers identified increased T_1-signal intensity in the RV, while in another control patient, only 1/13 (8%) readers identified a fat signal.

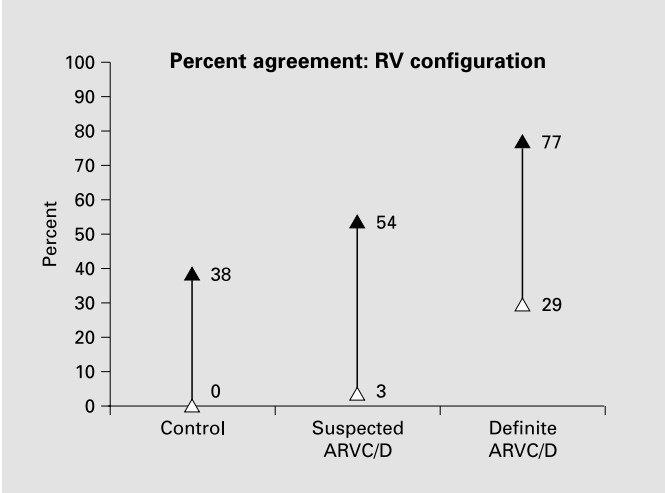

Fig. 8. Percentage reader agreement for abnormal morphology of the RV. The minimum (△) and maximum (▲) rates of agreement *on a per-patient basis* are shown for the control group, the suspected and the definite ARVC/D group. For example, for 1 patient in the control group, 5/13 (38%) readers identified abnormal right ventricular morphology, while in another control patient, 0/13 (0%) readers described abnormal right ventricular morphology.

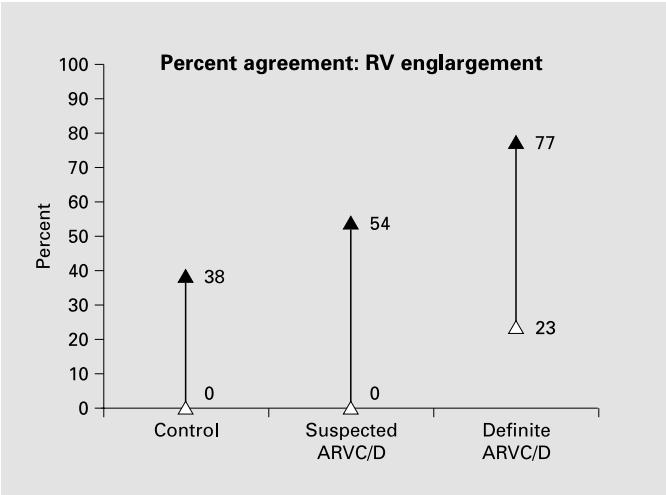

Fig. 7. Percentage reader agreement for identification of enlargement of the RV. The minimum (△) and maximum (▲) rates of agreement *on a per-patient basis* are shown for the control group, the suspected ARVC/D group and the definite ARVC/D group. For example, for 1 patient in the control group, 5/13 (38%) readers identified enlargement of the RV, while in another control patient, 0/13 (0%) readers described right ventricular enlargement.

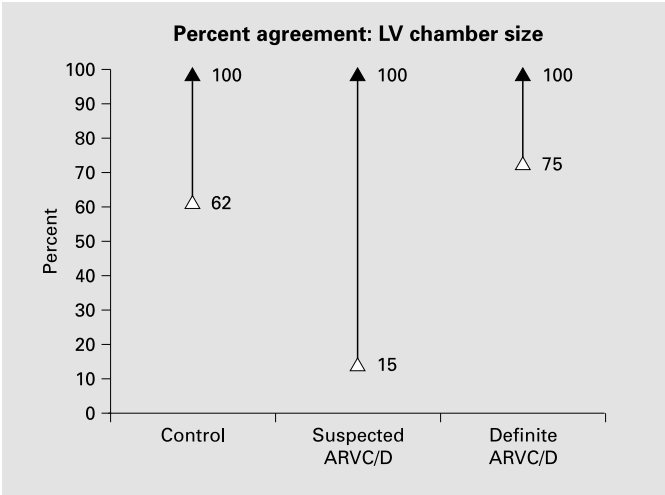

Fig. 9. Percentage reader agreement for identification of normal size of the LV. The minimum (△) and maximum (▲) rates of agreement *on a per-patient basis* are shown for the control group, the suspected and the definite ARVC/D group. For example, for 1 patient in the control group, 13/13 (100%) readers identified normal LV size, while in another control patient, 8/13 (62%) readers described a normal size LV.

minimum rates of agreement suggest that right ventricular chamber size and morphology show lower degrees of variability between readers compared to high T_1 signal (fat) in the myocardium for suspected ARVC/D and definite ARVC/D patients.

MR Image Quality

There was no relationship between image quality (χ^2 = 6.88, d.f. = 3, $p > 0.05$), the presence or severity of motion artifacts ($\chi^2 = 2.087$, d.f. = 3, $p > 0.05$), or the presence and severity of other artifacts ($\chi^2 = 1.62$, d.f. = 3, $p > 0.05$) and whether a case was reported as ARVC/D present or absent. The image quality, motion and other artifact scores are summarized in table 2.

Table 2. Percent rating responses for image quality, motion artifacts and other artifacts

Parameter	Rating	Definite ARVC/D n = 7	Suspected ARVC/D n = 32	No ARVC/D n = 6
Image Quality	Excellent	7	7	9
	Good	37	38	47
	Fair	44	42	35
	Poor	12	13	9
Motion Artifacts[1]	None	15	17	21
	Mild	32	42	57
	Moderate	45	32	19
	Severe	8	9	3
Other Artifacts	None	52	53	72
	Mild	13	17	21
	Moderate	30	22	7
	Severe	5	8	0

All figures are percentages.

[1] Mild = Artifact(s) do not interfere with interpretation; moderate = artifact(s) interfere somewhat with diagnosis; severe = artifact(s) interfere extensively with diagnosis, may render image uninterpretable.

Discussion

MR imaging is frequently performed when the diagnosis of ARVC/D is considered. MR imaging readily evaluates the morphology and size of the RV using spin-echo images. Additionally, MR has the potential to identify fat signal within the myocardium [5, 7–12]. While these findings have been reported to be key MR features of patients with ARVC/D, their reproducibility has previously not been determined in a multi-center study. The results of this study indicate that identification of a fat signal is less reliable than identification of right ventricular enlargement or morphologic changes (fig. 1–3). Further, the identification of the fat signal by the readers and the final diagnosis of ARVC/D were highly associated ($p < 0.0001$), suggesting readers relied on this finding for their final diagnosis of ARVC/D. We conclude that for MR imaging performed on conventional MR scanners (without dedicated cardiovascular MR software), identification of fat signal using conventional MR scanners is not a reliable predictor of ARVC/D, and is not reproducibly identified by expert MR readers.

There are several reasons that may account for these results. The RV has a complex geometry, is asymmetric and highly trabeculated [5]. In normal individuals, the mean right ventricular free-wall thickness is only 2.7 ± 0.4 mm, and only 1.9 ± 1.1 mm at the anterior right ventricular apex [6]. Epicardial fat is usually present, especially in association with the right coronary artery and left anterior descending artery. Tongues of epicardial fat may extend into the myocardium in normal individuals [14, 17]. Pathologically, these individuals have been found to have fatty infiltration but no fibrosis of the right ventricular myocardium. This is in contrast to patients with ARVC/D, who usually do have fibrosis in addition to fat in the RV [14, 17]. MR identification of a right ventricular fat signal requires high resolution images without significant artifacts. Unfortunately, patients with ARVC/D are likely to have frequent premature ventricular beats that can cause motion artifacts and diminish image quality. Image quality for patients in this study was considered excellent in less than 10% of cases (table 2). We note that morphologic findings and overall assessment of right ventricular size by MR imaging are less dependent on very high resolution detail and correspondingly had better interobserver agreement as well as better agreement with the final patient diagnosis.

An additional reason for reader variability may be the lack of a standardized imaging protocol for the MR examination of patients with suspected ARVC/D. Thus some of the views obtained may not be optimal. In addition, other aspects of the test were not standardized. These included whether MR scanning was performed in the prone or supine position, the use of a body coil or a surface coil (i.e., phased-array thoracic coil) and the orientation of phase encoding and read-out gradients. Thus, in contrast to our results, a single-center study showed greater than 80% agreement between three independent ob-

Table 3. Proposed optimal acquisition and analysis protocol for cardiovascular MR imaging in ARVC/D

(I) Pharmacologic control of arrhythmia (e.g., beta-blocker).
(II) 1.5 Tesla MR scanner.
(III) Thoracic torso or cardiac coil (phased array receiver coil).
(IV) Sequences (field of view preferred 24-26cm, but less than 32 cm). Anterior surface coil may be used to reduce 'wrap around' artifact.
1. Ventricular anatomy, wall thinning, fatty infiltration.
a. ECG-gated spin echo, transaxial plane, to cover from above the pulmonary valve to the diaphragm with suppression of blood pool signal (such as preparatory double inversion pulse) with either 3-mm multi-slice, 4 NEX, 5-mm gap, respiratory compensation or 5-mm single slices, 1 NEX, 3-mm gap with multiple acquisitions during breath holds. Fast spin-echo technique recommended only for breath-hold images. In-plane pixel resolution should be $\leq 1.5 \times 1.5$ mm. Motion/artifact control techniques such as the use of a saturation band over the anterior chest wall is encouraged.
b. Same spin-echo sequence in the short axis plane from base to apex of both ventricles.
2. Ventricular function (regional and global): Breath-hold cine gradient-echo sequences (steady-state free precession) sequences preferred such as TrueFISP, balanced fast-field echo, Fiesta) encompassing the entire right and LVs in both transaxial and short axis planes (planes to correspond with the spin-echo acquisitions above to allow direct comparison). Ten-millimeter gap between center of slices and optimal thickness according to local practice, but not less than 5 mm.
3. Other planes and sequences optional; for example fat suppression double inversion recovery fast spin echo images
(V) Analysis to include:
1. Quantification of global RV and LV volumes, function and mass.
2. Examination for regional wall-motion abnormalities on dynamic cines.
3. Examination for RV anatomical abnormalities such as thinning, bulging, abnormal trabeculation.
4. Examination for possible fatty infiltration, excluding known sources of fat such as around the right coronary artery and left anterior descending coronary artery, atrial lipomatous hypertrophy and obesity.
5. Direct comparison of possible fatty infiltration sites with regional wall motion in corresponding areas on dynamic cines.
6. Examination for LV involvement (clinically apparent in up to 15% of cases).

servers [12] for MR imaging of ARVC/D. The authors of this study suggest an MR imaging protocol for ARVC/D (table 3) that includes imaging in at least 2 planes, insuring adequate coverage of the RV for known areas of predominance of ARVC/D (RV inflow and outflow tracts and apex). Other alternative protocols have been used [5, 7, 12] and optimal protocol selection will require additional studies.

MR images obtained and interpreted at a single center also have the advantage of evaluating cine MR images [11] that aid in the diagnosis of ARVC/D and improve the reliability of MR imaging. For example, myocardial thinning on static MR images together with a wall motion abnormality in the same region of the RV on the cine loop has a high specificity for ARVC/D [18]. Cine images display right ventricular global function and regional wall motion abnormalities. A major deficiency of many MR scanners is the inability to distribute, e.g. on CD-ROM, cine loops of the acquired images. This deficiency is being addressed on newer MR scanners, and CD-ROM or videotape capability is more likely to be present on those MR scanners that have dedicated software and hardware for cardiovascular MR imaging. PACS technology is increasingly available and may further aid in the distribution of cine MR images. Thus, our results are only applicable to the use of static (generally spin-echo) images for ARVC/D diagnosis as performed at general MR imaging centers in the United States and Canada. Since cine loops of the cardiac cycle were not available in this study, we cannot fully assess the accuracy of cine MR imaging for ARVC/D diagnosis.

Except for the control patients, all other patients evaluated in this study had idiopathic ventricular tachycardia, and some of these patients likely had right ventricular outflow tract (RVOT) tachycardia. This could confound our results, because there are reports that patients with RVOT tachycardia have abnormalities on MR imaging. White et al. [18] reported right ventricular abnormalities on MR

imaging in 76% of 46 patients with RVOT tachycardia, including fixed thinning, reduced wall thickening or reduced wall motion. The most frequent location of these abnormalities was in the right ventricular anterior wall, but there was often caudal extension into the subparietal region in the anterior wall of the RV. Fatty replacement was observed in 8 (25%) patients, including those with an indeterminate diagnosis. Similar findings were reported by others [19–22]. In contrast, Grimm et al. [23] reported no cardiac MR imaging abnormalities in 14 patients with RVOT tachycardia. This wide range of results may be partly attributed to differences in patient selection, diagnostic criteria for RVOT tachycardia and definition of abnormal hyperintense signals in MR images as well as incomplete diagnostic evaluation, particularly by the lack of angiographic confirmation of the abnormalities [24]. Therefore the specificity of hypokinesis, akinesis, or even fatty infiltration by MR imaging to differentiate patients with RVOT tachycardia from those with ARVC/D remains unclear, particularly when these changes are limited to the body of the RV and are not present in the posterior subtricuspid region.

In this study, we did not quantitate ventricular function or size. ARVC/D is readily diagnosed when advanced structural abnormalities of the RV are present. The disease, however, is more difficult to diagnose with certainty when only minimal structural changes of the RV are present. Quantitative analysis of the RV is likely more reliable than visual inspection of chamber diameter or area and can be compared with published normal values using cardiovascular MR imaging [25]. Minor increases in RV volumes in the absence of other findings should be treated with caution as a possible early sign of ARVC/D which, in the correct clinical context, should lead to periodic reassessment. Normal volumes in the absence of other signs of ARVC/D is highly reassuring.

In conclusion, radiologists and cardiologists should recognize that the diagnosis of ARVC/D is presently based on the fulfillment of ARVC/D Task Force criteria. In this regard, MR imaging is useful for delineating the morphology of the RV. Using conventional MR scanners that are most widely available in the community setting, identification of right ventricular enlargement and abnormal morphology are more reliable than identification of a fat signal in the right ventricular myocardium. Dedicated cardiovascular MR scanners are now available at specialized centers, and evaluation of interobserver variability using this technology needs to be undertaken using a standardized imaging protocol and analysis method with experienced MR readers.

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