Misdiagnosis of Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy

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ARVD/C Misdiagnosis. ***Introduction:*** **Diagnosis of arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) has major implications for the management of patients and their first-degree relatives. Diagnosis is based on a set of criteria proposed by the International Task Force for Cardiomyopathies. We report our experience in providing a re-evaluation for patients who previously have been diagnosed with ARVD/C.**

Methods and Results: **We studied 89 patients who requested a re-evaluation for diagnosis of ARVD/C at our center. Each of these patients had been diagnosed with ARVD/C at their initial evaluation. Each patient was re-evaluated with clinical history, physical examination, and noninvasive testing at our center. Invasive testing, which included electrophysiologic testing, right ventricular angiography, and endomyocardial biopsy, was performed when clinically indicated. Sixty (92%) of the 65 patients who had undergone magnetic resonance imaging (MRI) at an outside institution were reported to have an abnormal MRI consistent with ARVD/C. Among these patients, the only abnormality identified was the qualitative finding of intramyocardial fat/wall thinning in 46 patients. On re-evaluation, these qualitative findings were not confirmed. None of these 46 patients ultimately were diagnosed with ARVD/C. Among the entire patient group, only 24 (27%) of the 89 patients met the Task Force criteria for ARVD/C.**

Conclusion: **This study demonstrates that the high frequency of "misdiagnosis" of ARVD/C is due to over-reliance on the presence of intramyocardial fat/wall thinning on MRI, incomplete diagnostic testing, and lack of awareness of the Task Force criteria. Diagnosis of ARVD/C cannot rely solely upon qualitative features on MRI.** *(J Cardiovasc Electrophysiol, Vol. 15, pp. 300-306, March 2004)*

arrhythmogenic right ventricular dysplasia/cardiomyopathy, magnetic resonance imaging, signal-averaged electrocardiography

Introduction

Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) is characterized by structural and functional abnormalities of the right ventricle (RV) and the development of ventricular arrhythmias.[1-7] Presenting symptoms can vary from palpitations to sudden death.[1,8-10] Overt forms of the disease are straightforward to diagnose based on the major and minor criteria proposed by the International Task Force for Cardiomyopathy (Task Force) (Table 1).[11] The diagnosis of early and more localized forms of the disease often is difficult. One of the main differential diagnoses of ARVD/C is idiopathic RV tachycardia, which is a benign condition occurring in a structurally normal heart. Differentiating these two conditions is clinically important, because the former is an inherited cardiomyopathy with a risk of sudden death, whereas idiopathic ventricular tachycardia (VT) is a nongenetic condition associated with an excellent prognosis.[4,7,12-17] Therefore, it has been recommended that patients suspected of having ARVD/C undergo thorough initial evaluation with noninvasive testing. Standard noninvasive testing for ARVD/C includes an electrocardiography (ECG), echocardiography, signal-averaged ECG (SAECG), Holter monitoring, and magnetic resonance imaging (MRI).[6,11] If noninvasive testing reveals findings consistent with ARVD/C, then invasive testing including RV angiography and RV endomyocardial biopsy are recommended to confirm the diagnosis.[11] The Johns Hopkins ARVD/C program was established in 1999 to care for patients with ARVD/C and to systematically study the disease. In the process of evaluating patients previously diagnosed with ARVD/C who were referred for re-evaluation, we became increasingly aware that many patients had received the diagnosis without a complete evaluation and/or without meeting the Task Force criteria. The aim of this study is to describe the clinical features of several of these patients and report our experience in providing a second opinion for a large consecutive series of patients with ARVD/C.

Methods

The study population consisted of 89 patients who had been diagnosed with ARVD/C by their local cardiologist and sought a second opinion at our institution. Thirty of these patients (34%) had undergone placement of an implantable

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TABLE 1

Task Force Criteria for Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy (ARVD/C)

1. Global and/or regional dysfunction and structural alterations
Major
Severe dilation and reduction of right ventricular ejection fraction with no (or only mild)
LV impairment
Localized right ventricular aneurysms (akinetic or dyskinetic areas with diastolic bulging
Severe segmental dilation of the right ventricle
Minor
Mild global right ventricular dilatation and/or ejection fraction reduction with normal LV
Mild segmental dilation of right ventricle
Regional right ventricular hypokinesis
2. Tissue characterization of walls
Major
Fibrofatty replacement of myocardium on endomyocardial biopsy
3. Repolarization abnormalities
Minor
Inverted T waves in right precordial leads ($V_2$and V_3) (age >12 years, in absence of right bundle branch block)
4. Depolarization conduction abnormalities
Major
Epsilon waves or localized prolongation (>110 ms) of the QRS complex in right precordial leads (V_1–V_3)
Minor
Late potential (signal-averaged ECG)
5. Arrhythmias
Minor
Left bundle branch block type ventricular tachycardia (sustained and nonsustained) on
ECG, Holter, or exercise testing
Frequent ventricular extrasystoles (>1,000/24 hours on Holter monitoring)
6. Family history
Major
Familial disease confirmed at necropsy or surgery
Minor
Familial history of premature sudden death (<35 years) due to suspected right ventricular dysplasia
Familial history (clinical diagnosis based on present criteria)
To have ARVD/C one should meet the following:
2 major criteria **OR** 1major AND 2 minor criteria **OR** 4 minor criteria

LV = left ventricle.

cardioverter defibrillator (ICD). The remaining 59 patients had been advised to undergo ICD placement. After obtaining informed consent according to the protocol approved by Human Studies Research Committee, detailed results of the patients' initial evaluation that led to the diagnosis of ARVD/C were obtained and reviewed. These patients were re-evaluated at our center between January 1999 and January 2003. Their evaluation included a detailed clinical history and physical examination. All patients then underwent noninvasive testing, which included 12-lead ECG, SAECG, exercise stress testing, Holter monitoring, two-dimensional echocardiography, and cardiac MRI. Cardiac computed tomography (CT) was performed in patients when MRI was contraindicated due to ICD hardware. Invasive testing, which included electrophysiologic (EP) study, RV angiography, and endomyocardial biopsies, was performed in 63, 22, and 24 patients, respectively, as clinically indicated.

Noninvasive Testing

Twelve-lead ECG and SAECG

Standard 12-lead ECGs and double-speed, double-amplitude ECGs were obtained. Findings considered to be consistent with ARVD/C include T wave inversions beyond V_2, QRS duration >110 ms in the right precordial leads, and/or presence of an epsilon wave.[18–21] SAECG was obtained using a MAC 15 system (Marquette Inc., Milwaukee, WI, USA) with high-gain amplification and bidirectional Butterworth filters (40–250 Hz).[21] Late potentials were considered present if ≥2 of the following criteria were fulfilled: (1) fQRS ≥114 ms, (2) LAS40 ≥38 ms, and (3) RMS40 <20 μV.[21] A symptom-limited exercise stress test using the Bruce protocol was performed. Frequent premature ventricular contractions (PVCs), nonsustained VT, and/or sustained VT with exercise were considered abnormal. Holter monitoring was considered abnormal when >1,000 PVCs were present in 24 hours.[6,11] Two-dimensional echocardiography was performed with specific emphasis on the RV. Abnormalities consistent with ARVD/C noted on echocardiography were global and/or regional RV dilation and/or dysfunction.[11]

Magnetic resonance imaging

MRI was performed using a 1.5-T scanner (CV/i, General Electric Medical Systems, Waukesha, WI, USA) using ECG gating. The subject was positioned supine in the scanner, and a cardiac phased-array surface coil was used for radiofrequency signal detection. For the transaxial black blood images, double-inversion recovery (blood suppression) fast spin echo was acquired. Imaging parameters included the following: TR = 1–2 R-R intervals, TE = 5 ms, slice thickness = 5 mm, interslice gap = 5 mm, and field of view (FOV) = 24–28 cm. Anterior coil elements only were used, and the posterior coil was switched off to reduce "wraparound" artifact. Following this, the same sequence was repeated with the chemical shift fat suppression manually tuned to the fat peak, to generate fat-suppressed axial black blood images. Bright blood cine imaging in the axial and short axis planes was acquired by a steady-state free precession pulse sequence. The imaging parameters for

the bright blood sequence were as follows: TR = 3.5, TE = 1.2 ms, flip angle = 45°, slice thickness = 8 mm, inter-slice gap = 4 mm, and FOV = 36–40 cm, 10 views per segment. Images were obtained from the diaphragm to the level of the right coronary artery (i.e., to include the pulmonary outflow tract) for all the axial sequences. In the short-axis plane, images were acquired from the apex to the mitral valve plane (base of heart). The temporal resolution for cine images was ≤40 ms without view sharing. A phased-array cardiac coil was used for all studies. MRI findings consistent with ARVD/C were global and/or regional RV dilation and dysfunction. Intramyocardial fat and wall thinning noted on MRI were not included in the final diagnostic consideration because they are not a part of the Task Force criteria.

CT scan

In patients in whom MRI was contraindicated due to ICD placement, a prospective ECG-gated contrast-enhanced cardiac CT was performed using a multidetector (four-detector) helical scanner (Siemens), slice thickness 2.5 mm, matrix 512 × 512, and pitch <1. FOV was 24–28 cm. Images were acquired in the axial plane. Coverage included root of main pulmonary artery superiorly to the floor of RV inferiorly. Abnormalities on CT scan consistent with ARVD/C included an enlarged RV and an enlarged outflow tract.[22,23]

Invasive Testing

The invasive testing protocols used in our study are those recommended by the multicenter US ARVD study.[6] Following are the brief descriptions of the EP study, RV angiography, and RV endomyocardial biopsy.

EP study

Standard programmed electrical stimulation was performed at two sites, three drive train cycle lengths, and up to triple extrastimuli. If noninducible, burst pacing and repeat programmed electrical stimulation were performed during an infusion of 3 μg/mL isoproterenol.

RV angiography

With the patient under local anesthesia, right heart catheterization via jugular venous puncture and hemodynamics are performed prior to RV angiography. Using a pigtail (5 French or larger) catheter, 40–50 mL of low-toxicity dye is injected at 12–15 mL/s, followed by RV angiography in four standard projections: (1) 30° RAO, (2) 60° LAO, (3) posteroanterior, and (4) lateral. Cine angiography at 15 images/s is performed during deep inspiration (diaphragm) and breath-hold.

RV endomyocardial biopsy

Target sampling was performed using a medium-sized jaws bioptome, guided by abnormal areas (as seen by echocardiography or angiography) and likely areas (right ventricular outflow tract, apex, basal inferior walls). Samples were taken at the myocardial border of the RV free wall and septum (but not strictly septal) by directing halfway posteriorly in the 60° LAO view. Five samples of sufficient size (2 mm) are taken and placed in the five samples in 10% phosphate-buffered formalin. Biopsy samples were placed in formalin, embedded in a paraffin block, and sent for pathologic analysis.

Statistical Analysis

Data are expressed as mean ± SD where appropriate. For categorical characteristics such as presenting symptoms and individual tests, data are expressed as frequency (percentage) during initial and re-evaluation.

Results

Case Presentations

Case report

A 17-year-old male presented with symptoms of palpitation and presyncope. Twelve-lead ECG performed by the patient's physician revealed T wave inversion in precordial leads V_1 and V_2. Holter monitoring revealed 407 unifocal PVCs. Transient ventricular bigeminy with left bundle branch block morphology was observed during the recovery phase of an exercise stress test. He was referred to a cardiologist for further evaluation. An echocardiogram revealed normal cardiac structure and function. Cardiac MRI was performed and was interpreted as showing increased intramyocardial fat signal in the RV free wall with wall thinning. Results of baseline EP testing was within normal limits. Ventricular fibrillation was induced with closely coupled three extrastimuli during a combined infusion of norepinephrine and dopamine. Based on these results, he was diagnosed with ARVD/C and an ICD was placed. He subsequently received two inappropriate ICD shocks as a result of sinus tachycardia. Upon re-evaluation at Johns Hopkins, ECG revealed an incomplete right bundle branch block but the patient was otherwise normal. SAECG showed no evidence of late potentials. Cardiac CT showed normal RV chamber size and wall thickness with no evidence of intramyocardial fat. EP testing demonstrated no inducible ventricular arrhythmias in the baseline state or during isoproterenol infusion. RV angiography revealed normal RV structure and function, and RV endomyocardial biopsy was normal. Based on this evaluation, we concluded that the patient did not fulfill the diagnostic criteria for ARVD/C. At the request of the patient and his parents, the ICD was explanted. He has remained symptom-free during the subsequent 2 years.

Patient Population

The baseline characteristics of the study population are given in Table 2. Mean age was 37±13 years, and 51% of the

TABLE 2
Baseline Characteristics of the Study Population

No. of patients	89
Age (years)	37 ± 13
Male gender	46 (51%)
Presenting symptom	
Asymptomatic	11(12%)
Syncope	25 (28%)
Palpitation	52 (58%)
Other	5 (6%)
Presenting arrhythmia	
VT	30 (34%)
NSVT	54 (61%)
None	17 (19%)
Family history	5 (6%)

NSVT = nonsustained ventricular tachycardia; VT = ventricular tachycardia.

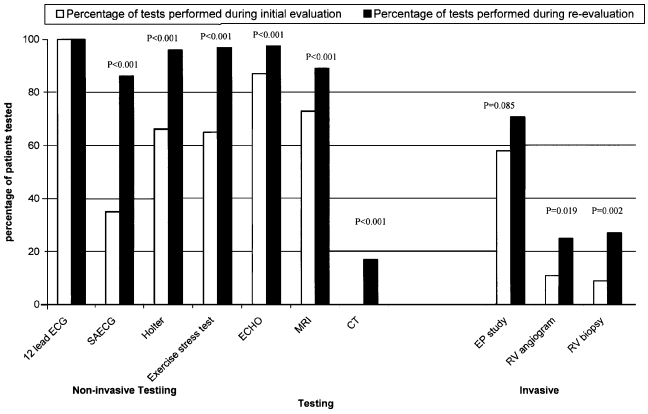

Figure 1. *Noninvasive and invasive tests in 89 suspected arrhythmogenic right ventricular dysplasia/cardiomyopathy patients. CT = computed tomography; ECHO = echocardiography; EP = electrophysiologic; MRI = magnetic resonance imaging; RV = right ventricular; SAECG = signal-averaged electrocardiography.*

patients were men. The most common presenting symptom was palpitations, followed by syncope. The most common presenting arrhythmia was nonsustained VT. Of 89 patients, 5 had a family history of ARVD/C.

Diagnostic Testing

Figure 1 shows the proportion of patients who underwent each of the recommended noninvasive and invasive diagnostic tests for ARVD/C at the time of initial evaluation and re-evaluation. Following re-evaluation, >90% of the patients had all of the noninvasive testing performed. This information, combined with the results of the diagnostic testing, is given in Table 3. The most commonly performed diagnostic tests at the time of initial evaluation were ECG, echocardiography, and MRI performed in 100%, 87%, and 73% of patients, respectively. SAECGs were obtained initially in only one third of patients. RV angiograms and endomyocardial biopsies were obtained in approximately 10% of patients during initial evaluation.

SAECG

Thirty-one (35%) of the 89 patients had an SAECG performed as part of their initial evaluation (Table 3). The SAECG was abnormal in 12. Among the 12 patients with an abnormal SAECG, 4 had a right bundle branch block on resting 12-lead ECG, which makes SAECG uninterpretable. After re-evaluation at our institution, 81 (91%) patients had SAECG performed. Of these 81 SAECGs, 19 (21%) were abnormal. The remaining 8 patients did not have a SAECG performed due to the presence of either right bundle branch block or frequent PVCs.

MRI/CT

Sixty-five (73%) patients underwent MRI as a part of the initial evaluation (Table 3). Of these patients, 60 (92%) were reported to have abnormal findings consistent with a diagnosis of ARVD/C. The most common abnormality reported was RV free-wall thinning and/or focal intramyocardial fat in 46 of the 60 patients who had abnormalities noted on MRI. Additionally, RV regional and/or global dysfunction was reported in 14 (23%) patients.

The majority of patients (75 [84%]) underwent MRI examination during re-evaluation at our hospital. In none of the patients who initially were diagnosed with ARVD/C based on wall thinning and intramyocardial fat alone were the findings confirmed on repeat MRI. None of these patients ultimately were diagnosed with ARVD/C. In contrast, of the 14 patients in whom RV functional abnormalities were noted on MRI performed at the time of initial evaluation, the abnormalities were confirmed during re-evaluation in all of patients. All of these patients met the Task Force criteria for ARVD/C during the re-evaluation. Ten (71%) of these 14 patients had a high intramyocardial signal consistent with fat on repeat evaluation. In 15 patients (17%) in whom MRI was contraindicated due to ICD placement, cardiac CT was performed to evaluate

TABLE 3

Noninvasive and Invasive Testing Performed During Primary Evaluation and Re-Evaluation

	Initial Evaluation		Re-Evaluation	
	N (%)	**N (% Abnormal)**	**N (%)**	**N (% Abnormal)**
N	89		89	
Twelve-lead ECG	89 (100)	39 (44)	89 (100)	30 (34)
SAECG	31 (35)	12 (13)	81 (91)	19 (21)
Holter	59 (66)	29 (33)	85 (96)	31 (35)
Exercise stress test	58 (65)	15 (17)	86 (97)	16 (18)
ECHO	77 (87)	36 (40)	89 (100)	27 (30)
MRI	65 (73)	60 (67)	82 (92)	27 (30)
Global dysfunction		10 (11)		15 (17)
Regional dysfunction		9 (10)		11 (12)
Wall thinning		51 (57)		18 (20)
Fat infiltration		52 (58)		27 (30)
CT	0 (0)		15 (17)*	10 (11)
EP study	52 (58)	29 (33)	63 (71)	30 (31)
RV angiogram	10 (11)	6 (7)	22 (25)	9 (10)
RV biopsy	8 (9)	2 (2)	24 (27)	6 (7)

*Eight patients had both MRI and CT.

CT = computer-assisted tomography; ECHO = echocardiography; EP study = electrophysiologic study; MRI = magnetic resonance imaging; RV = right ventricle; SAECG = signal-averaged ECG.

TABLE 4
Criteria Met During Initial and After Second Evaluation

	Initial Evaluation	Re-Evaluation
No criteria met	28	32
One minor criterion	36	26
Two minor criteria	10	5
Three minor criteria	1	2
Four criteria*	14	24

*For the purposes of this table, 1 major criterion is considered 2 minor criteria.

the RV. Ten of these CTs (11%) were abnormal. The diagnosis of ARVD/C was confirmed based on the Task Force criteria in all of these patients.

Invasive Testing

During re-evaluation, invasive testing, which consisted of RV angiography and endomyocardial biopsy, was performed in 22 patients (25%). All of these patients were found to have abnormalities on noninvasive testing. Of these patients, 11 showed fibrofatty tissue interspersed with islands of normal myocardial cells consistent with ARVD/C. Of the remaining 11 patients, endomyocardial biopsy revealed nongranulomatous myocarditis in 2, sarcoidosis in 1, cardiomyocyte disarray and interstitial fibrosis consistent with idiopathic dilated cardiomyopathy (no intervening normal myocytes along with interstitial fibrosis) in 2, and normal biopsies in 6.

Final Diagnosis

Of the 89 study patients who were diagnosed with ARVD/C by their local cardiologist, 24 (27%) ultimately were determined to meet the Task Force criteria for the diagnosis of ARVD/C. Table 4 summarizes the number of diagnostic criteria for ARVD/C that were present at the time initial evaluation and re-evaluation. In the patients who did not meet the criteria for ARVD/C, idiopathic VT was the most common final diagnosis 52 (58%). None of these patients showed any abnormality consistent with ARVD/C on noninvasive and invasive testing. Myocarditis, idiopathic cardiomyopathy, sarcoidosis, and occasional palpitations in normal individuals were the final diagnosis in the remaining 13 (15%) patients. The majority of patients either did not meet any criteria (n = 32) or met one criterion for ARVD/C (n = 26). Only 2 patients met 3 minor criteria and were diagnosed to have probable ARVD/C on re-evaluation. Twenty-four patients either had 1 major and 2 minor criteria or had 4 minor criteria, which confirms the diagnosis of ARVD/C.

Discussion

The results of our study demonstrate that many patients are prematurely diagnosed with ARVD/C. Among the 89 patients diagnosed with ARVD/C by their local cardiologist, the diagnosis was confirmed after re-evaluation in only 24 patients (27%). This high frequency of "misdiagnosis" of ARVD/C reflects the lack of complete diagnostic testing, over-reliance on the presence of intramyocardial fat and wall thinning on MRI, and lack of awareness of the Task Force criteria for ARVD/C.

MRI has been shown to be an important diagnostic tool for the diagnosis of ARVD/C because of its ability to demonstrate both morphologic and structural abnormalities of the heart. It is considered by some to be the noninvasive gold standard for evaluation of the RV in patients suspected of having ARVD/C.[24-30] This likely resulted in the high frequency with which MRI was performed at the time of initial evaluation, as well as over-reliance on the results of MRI in establishing the diagnosis of ARVD/C. Of 60 patients who underwent MRI during initial evaluation, 46 (77%) patients were reported to have RV free-wall thinning and/or focal intramyocardial fat as the only abnormal MRI finding. None of these patients, whose initial diagnosis of ARVD/C was based on RV wall thinning and intramyocardial fat, showed any structural and functional abnormalities consistent with ARVD/C on MRI performed at the time of the initial evaluation and after re-evaluation at our center. These patients also did not meet the Task Force criteria for ARVD/C. RV regional and/or global dysfunction were reported in 14 (23%) patients during initial evaluation. These findings were confirmed by MRI and/or CT in all of these patients, who ultimately met the Task force criteria after re-evaluation. In close agreement with Tandri et al.,[31] the finding of intramyocardial fat was observed in 10 (71%) of the 14 patients who were diagnosed with ARVD/C based on the Task Force criteria on repeat evaluation. Even though MRI is a useful tool in ARVD/C diagnosis, it is operator dependent, and most MRI centers lack experience with ARVD/C evaluation. The results of this study also are consistent with a recent study, which showed that there is poor agreement among MRI physicians in interpretation of qualitative findings such as intramyocardial fat and wall thinning, and currently there is no standardized MRI protocol for ARVD/C.[32] The study emphasizes that it is important to recognize that findings of free-wall thinning and/or increased intramyocardial fat signal on MRI are not part of the Task Force criteria, and that experts in the field do not recommend equating intramyocardial fat signal on MRI to fat infiltration observed on biopsy.

The presence of late potentials on SAECG is considered one of the minor criteria for establishing the diagnosis of ARVD/C by the Task Force committee.[11] It is striking that in our series only 35% had SAECG performed as a part of the initial evaluation. In at least one fourth of these patients, the presence of right bundle branch block made SAECG results uninterpretable. After re-evaluation, SAECG was abnormal in only 19 of 81 patients. Of these 19 patients, 17 had met the criteria for ARVD/C. These findings are consistent with a prior study that reported the presence of SAECG abnormalities in 47% to 100% of ARVD/C patients and that the abnormalities also were dependent on the extent of disease.[20,21]

In our study, 5 (6%) patients were being evaluated because of a family history of ARVD/C. Of these 5 patients, 1 (20%) ultimately was diagnosed with ARVD/C based on the Task Force criteria. As pointed out in a recent article by Hamid et al.,[33] it is important to have a high degree of clinical suspicion of ARVD/C when there is a family history of ARVD/C. These prior authors propose that the current Task Force criteria be made less stringent for patients with a family history of disease in recognition of the fact that nondiagnostic ECG, echocardiography, or Holter abnormalities may reflect early or mild disease expression.

It is notable that in our study, one third (34%) of the patients had undergone placement of an ICD before being seen for re-evaluation; the remaining 59 patients were advised to undergo ICD placement. Six (20%) of the patients who

had received an ICD did not meet the criteria for ARVD/C. The remarkably low threshold for ICD placement observed in our study is striking and reflects in part the low threshold for ICD placement seen in the United States. Additional research and/or guidelines are needed to help clinicians determine when to implant an ICD in this patient population. Previously identified risk factors for sudden death among ARVD/C patients, as shown by Turrini et al.,[34] include history of syncope, physical exercise, spontaneous VT or ventricular fibrillation, RV dysfunction, left ventricular involvement, right precordial negative T wave, right bundle branch block, QT-QRS dispersion, right precordial ST-segment elevation, and late potentials. Turrini et al. concluded that only QRS dispersion, history of syncope, and right and/or left ventricular abnormalities at radionuclide angiography proved to be independent noninvasive predictors of sudden death.

A limitation of this study, as with all studies of ARVD/C, is that there is currently no gold standard that we can use to definitely diagnose or exclude the diagnosis of ARVD/C. The Task Force criteria that we rely upon in this study have never been validated. Therefore, it is possible that some patients with subtle evidence of ARVD/C but who do not meet the complete Task force criteria actually have early or mild disease expression. It is for this reason that these patients should continue to be followed closely by a cardiologist at annual or biannual repeat evaluations.

Conclusion

The results of this study demonstrate that incomplete diagnostic testing, over-reliance on the presence of intramyocardial fat and wall thinning on MRI, and lack of widespread awareness of the Task Force criteria for ARVD/C have led to a high rate of "misdiagnosis" of ARVD/C. The findings of this study have several implications for physicians involved in the care of patients suspected of having ARVD/C. First, the results of the study emphasize the importance of complete testing in order to use the Task Force criteria. At present it is recommended that 12-lead ECG, SAECG, Holter monitoring, exercise stress testing, echocardiography, and cardiac MRI are the minimum required noninvasive tests for the evaluation of patients suspected of having ARVD. Invasive testing, including EP testing, RV angiography, and RV endomyocardial biopsy, should be considered based on the results of the noninvasive test and performed when clinically indicated to confirm ARVD/C. The results of this study also caution physicians not to diagnose ARVD/C when the abnormalities are detected solely on MRI. Although MRI is one of the emerging tools aiding in the diagnosis of ARVD/C, it is reiterated that MRI findings of intramyocardial fat and/or thinning of RV free wall are not criteria set forth by the Task Force for diagnosis of ARVD/C. It is anticipated that the currently enrolling Multidisciplinary Study of Right Ventricular Dysplasia/Cardiomyopathy will provide important new information on the diagnosis and genetic basis of ARVD/C.[6]

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