An electrocardiographic scoring system for distinguishing right ventricular outflow tract arrhythmias in patients with arrhythmogenic right ventricular cardiomyopathy from idiopathic ventricular tachycardia

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BACKGROUND Ventricular arrhythmias in patients with arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) and idiopathic ventricular tachycardia (VT) can share a left bundle branch block/inferior axis morphology. We previously reported electrocardiogram characteristics during outflow tract ventricular arrhythmias that helped distinguish VT related to ARVD/C from idiopathic VT.

OBJECTIVE To prospectively validate these criteria.

METHODS We created a risk score by using a derivation cohort. Two experienced electrophysiologists blinded to the diagnosis prospectively scored patients with VT/premature ventricular contractions (PVCs) with left bundle branch block/inferior axis pattern in a validation cohort of 37 ARVD/C tracings and 49 idiopathic VT tracings. All patients with ARVD/C had their diagnosis confirmed based on the revised task force criteria. Patients with idiopathic VT were selected based on structurally normal hearts with documented right ventricular outflow tract VT successfully treated with ablation. The scoring system provides 3 points for sinus rhythm anterior T-wave inversions in leads V_1–V_3 and during ventricular arrhythmia: 2 points for QRS duration in lead I $\geq$120 ms, 2 points for QRS notching, and 1 point for precordial transition at lead V_5 or later.

RESULTS A score of 5 or greater was able to correctly distinguish ARVD/C from idiopathic VT 93% of the time, with a sensitivity of 84%, specificity of 100%, positive predictive value of 100%, and negative predictive value of 91%.

CONCLUSIONS We describe a simple scoring algorithm that uses 12-lead electrocardiogram characteristics to effectively distinguish right ventricular outflow tract arrhythmias originating from patients with ARVD/C versus patients with idiopathic VT.

KEYWORDS ARVD/C; RVOT-VT; Idiopathic VT; Electrocardiogram; Risk score

ABBREVIATIONS **ARVD/C** = Arrhythmogenic right ventricular dysplasia/cardiomyopathy; **CI** = confidence interval; **ECG** = electrocardiogram; **LBBB** = left bundle branch block; **PVC** = premature ventricular contraction; **ROC** = receiver operator characteristic; **RV** = right ventricular; **RVOT** = right ventricular outflow tract; **VT** = ventricular tachycardia

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Introduction

Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) is an inherited disorder characterized by fibrofatty infiltration of the myocardium, with ventricular arrhythmias and sudden cardiac death being hallmarks of the disease.[1–4] In contrast, idiopathic ventricular tachycardia (VT) from the right ventricular outflow tract (RVOT) is a relatively benign condition and occurs in patients with structurally normal hearts.[5,6] Regardless of the underlying process, ventricular arrhythmias from the RVOT share a left bundle branch block (LBBB) QRS morphology with inferior axis pattern, but methods to distinguish the two entities have been limited. On the basis of a

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retrospective analysis, we recently described surface electrocardiogram (ECG) characteristics (during VT or premature ventricular contractions [PVCs]) that appeared to be useful in distinguishing patients with ARVD/C from patients with idiopathic VT with an LBBB and an inferior QRS axis pattern.[7]

The purpose of this study was to prospectively validate the scoring system derived from our original cohort.

Methods

Description of the new validation cohort

We prospectively scored ECGs from new patients with ARVD/C selected from tertiary care referral centers experienced in the treatment of patients with ARVD/C. All ECGs in this study were based on a new group of 12-lead ECGs and did not include any from either the original cohort from which the ECG score was derived or any prior study. All patients had confirmed ARVD/C based on the revised task force criteria.[8,9] Only patients presenting with a ventricular arrhythmia characterized by an LBBB QRS morphology with inferior axis pattern were included.

The idiopathic VT group was composed of patients with arrhythmias arising from the RVOT region that were successfully treated with ablation at a tertiary care referral center. All patients with idiopathic VT had structurally normal hearts as assessed by physical examination, echocardiography (and/or magnetic resonance imaging), or right ventricular (RV) voltage maps. Patients with a diagnosis of idiopathic VT were included if they had at least one 12-lead ECG tracing with spontaneous PVCs or VT that were targeted for ablation having an LBBB/inferior QRS axis morphology.

Electrocardiographic ARVD/C risk score

We previously described ECG characteristics that differentiated ARVD/C from idiopathic VT.[7] Multivariate logistic regression of ventricular arrhythmia characteristics revealed duration of QRS in lead I $\geq$120 ms, earliest onset QRS in lead V_1, QRS notching, and precordial transition at lead V_5 or later—all significantly increased the odds of ARVD/C.[7] On the basis of our previously published findings, we created an ECG-based ARVD/C risk score. The risk score was created for simplicity and for everyday use without the need for simultaneous or computerized digital measurement. To that end, the earliest onset QRS in lead V_1 was eliminated. The presence of sinus rhythm anterior T-wave inversions in precordial leads V_1–V_3 was found in 75% of our original patients, suggesting that both the electrocardiographic findings described for VT/PVCs as well as the surface ECG can be used for useful noninvasive screening, so this criterion was added as well.

The risk score was derived using the logistic regression coefficients from our multivariate model in our previous cohort with the addition of anterior T-wave inversions during sinus rhythm. The final score included T-wave inversions in leads V_1–V_3 during normal sinus rhythm (3 points), lead I QRS duration $\geq$120 ms (2 points), QRS notching in multiple leads (2 points), and precordial lead transition at lead V_5 or later (1 point). The scoring algorithm is given in Table 1.

Table 1 Electrocardiographic ARVD/C risk score

ECG characteristic	Points
Anterior T-wave inversions (V_1–V_3) in sinus rhythm	3
VT/PVC	
Lead I QRS duration $\geq$120 ms	2
QRS notching (multiple leads)	2
V_5 transition or later	1
Maximum total	8

ARVD/C = Arrhythmogenic right ventricular dysplasia/cardiomyopathy; ECG = electrocardiogram; PVC = premature ventricular contraction; VT = ventricular tachycardia.

Definitions

Anterior T-wave inversions were defined as T-wave negativity in at least leads V_1–V_3. Lead I QRS duration $\geq$120 ms was defined as the duration from the initial deflection of the QRS complex to the end of the QRS complex in lead I. QRS notching in multiple leads was defined as a QRS complex deflection on the upstroke or downstroke of $>$ 0.05/mV that did not cross baseline occurring in at least 2 leads (Figure 1). The precordial transition point was designated as the earliest precordial lead where the R-wave amplitude exceeded the S-wave amplitude.

ECG analysis and scoring

We used de-identified ECGs that recorded 12 simultaneous leads. The amplitudes were set at 1 mV/cm for all patients. They were read as is without magnification or alteration. All ECGs were edited to include only the 12-lead ECG tracing, with all identifiable labeling removed (patient name, institution, computerized interpretation, dates, etc). Each case included a 12-lead ECG with ventricular arrhythmia (VT or PVCs) and a baseline sinus rhythm (to evaluate for anterior T-wave inversions). The 12-lead ECGs were scored by 2 experienced electrophysiologists blinded to the clinical data (N.B. and E.P.G.). An example of the ECG scoring is shown in Figure 2. Randomization of the order of ECG tracings was performed by using a random sequence generator. Disagreements between reviewers were deemed minor if the score differed by 2 points or less and major if the disagreement was greater than 2 points or changed the predicted diagnosis. Disagreements were then adjudicated by a third electrophysiologist blinded to the clinical data (M.M.S.).

Data analysis

The electrocardiographic ARVD/C risk score was created on the basis of the multivariate logistic regression coefficients in the original cohort that has been described previously.[7] Regression coefficients were rounded to the nearest whole number for ease of clinical utility. A cut point of 5 points or greater for prediction of ARVD/C was determined on the basis of testing characteristics measured in the original cohort. Standard definitions were used for testing characteristics including sensitivity, specificity, positive predictive value, and negative predictive value.[10] We assessed interobserver agreement for the electrocardiographic ARVD/C risk score by using an unweighted kappa statistic.

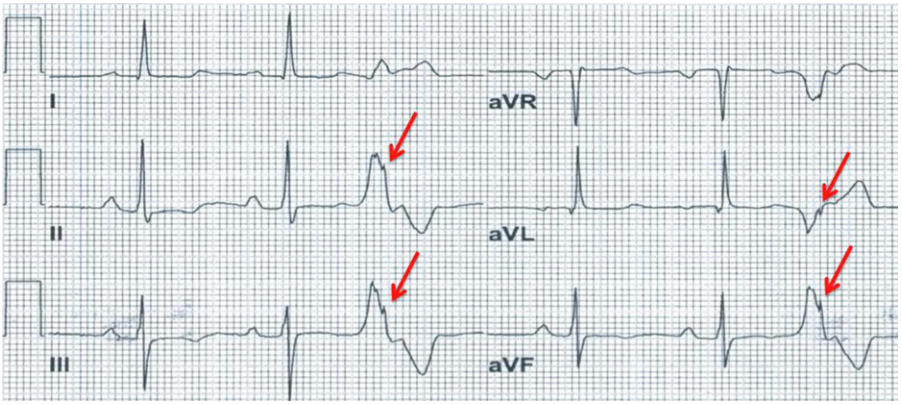

Figure 1 Example of QRS notching. Arrows show QRS notching in leads II, III, aVF, and aVL.

Stata version 10.0 (College Station, TX) was used for all statistical analyses.

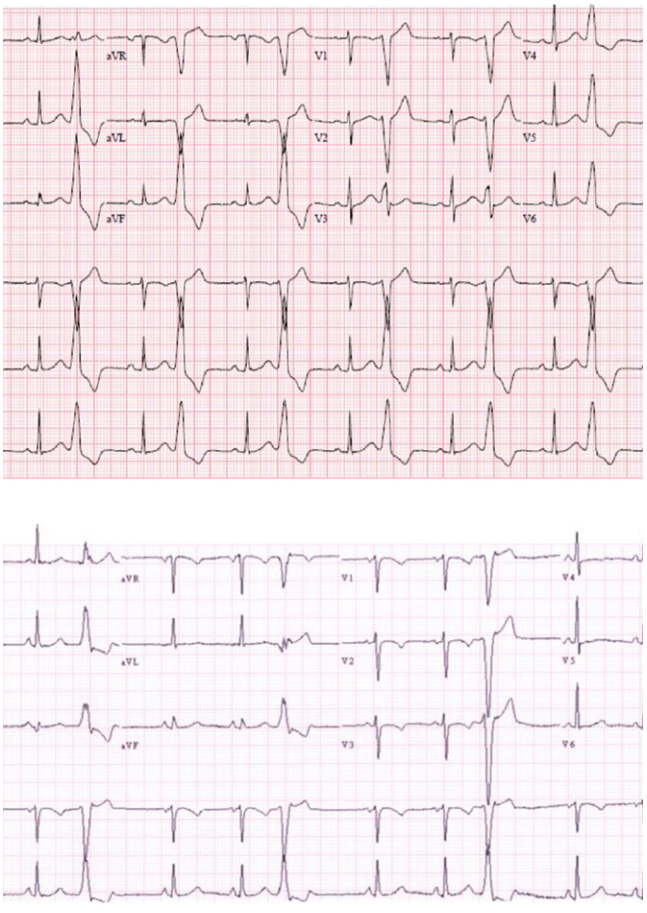

Figure 2 **A:** An electrocardiogram from a patient with idiopathic ventricular tachycardia. The tracing shows bigeminy with a right ventricular outflow tract PVC. This tracing would receive 0 points, as there are no anterior T-wave inversions (confirmed without bigeminy), lead I QRS duration <120 ms (110 ms), no notching, and the precordial transition is V_3/V_4. **B:** An electrocardiogram from a patient with arrhythmogenic right ventricular dysplasia/cardiomyopathy. The tracing shows bigeminy with a right ventricular outflow tract PVC. This tracing would receive 8 points, as there are anterior T-wave inversions (confirmed without bigeminy), lead I QRS duration ≥ 120 ms (120 ms), notching seen in leads I, III, aVL, V_5, and the precordial transition is V_5.

Results

A total of 86 ECGs with ventricular arrhythmias were analyzed from 85 patients (1 patient with ARVD/C had 2 different LBBB/inferior axis VT morphologies). Thirty-seven were from the ARVD/C cohort, in which 22 of 37 (59%) of the ECGs demonstrated VT and 15 of 37 (41%) showed PVCs. Forty-nine ECGs were from idiopathic VT patients; of these 4 of 49 (8%) ECGs of the idiopathic VT group demonstrated VT, while 45 of 49 (92%) had PVCs.

The mean age of patients with ARVD/C was 38.3 ± 2.4 years compared to 46.6 ± 2.2 years for controls with idiopathic VT ($P = .014$). There was no significant difference in sex (ARVD/C man 51.2% vs 48.8% idiopathic VT controls; $P = .11$). The majority of patients with ARVD/C were on antiarrhythmic agents (27 of 36, 75%), with sotalol being most common (19), followed by mexilitine (3), amiodarone (2), propafenone (1), flecainide (1), and disopyramide (1).

Scoring system threshold and testing characteristics

A score of 5 or greater occurred in 31 of 37 (84%) ARVD/C tracings vs 0 of 49 (0%) in idiopathic VT controls ($P < .0001$). A score of 5 or greater therefore had a sensitivity of 84%, specificity of 100%, positive predictive value of 100%, and negative predictive value of 91% in identifying a patient with ARVD/C; this score correctly classified 93% of the tracings. A dot plot is shown in Figure 3. The area under the receiver operator characteristic (ROC) curve was 0.93 ± 0.03 (95% confidence interval [CI] 0.86–0.99), and the ROC table for all the possible testing scores is given as Table 2.

The QRS duration in lead I ≥ 120 ms was found in 73% of ARVD/C ECGs vs 43% in idiopathic VT ECGs ($P = .005$). QRS notching in at least 2 leads was more often observed in ARVD/C ECGs compared with idiopathic VT ECGs (68% vs 43%; $P = .005$) (Fig. 3). Precordial transition at lead V_5 or later was also observed more commonly in ARVD/C ECGs compared to idiopathic VT ECGs (35% vs 8% [4 of 49]; $P = .002$).

Among the individual testing characteristics of the risk score, T-wave inversion had the best testing characteristics (Table 3). This was expected, as this characteristic carried the highest point assignment in our score, signifying the best prediction.

Table 2 Receiver operator characteristic (ROC) table

Cut point	Sensitivity	Specificity	Correctly classified	LR+	LR−
≥0	100	0	43.0	1	
≥1	97.3	36.7	62.8	1.5	0.07
≥2	97.3	40.8	65.1	1.6	0.67
≥3	86.5	69.4	76.7	2.8	0.20
≥4	86.5	75.5	80.2	3.5	0.18
≥5	83.8	100	93.0		0.16
≥6	51.4	100	79.1		0.49
≥7	35.1	100	72.1		0.65
≥8	18.9	100	65.1		0.81
>8	0	100	57.0		1.0

Area under the ROC curve = 0.926 (95% confidence interval 0.863–0.989).

Eliminating anterior T-wave inversions from the score, ARVD/C was suggestive if you satisfied 2 of the remaining 3 criteria, with a sensitivity of 70%, specificity of 73%, and correct classification of 72% (ROC area 0.79; 95% CI 0.70–0.88).

Interreader variability

Kappa statistics were calculated for the overall score as well as each individual criterion between the 2 blinded ECG scorers. Overall score agreement was 96%, with a kappa of 0.8. Most disagreements were minor in nature, and there were only 2 major disagreements (>2 points discrepancy); none of the disagreements changed the diagnosis. For example, the most common disagreement was an ECG of a patient with idiopathic VT scored as a "0" by one reader and "2" as the other, with QRS duration ≥120 ms being the discrepant characteristic. Anterior T-wave inversions were by far the best agreement between the 2 readers (kappa of 0.98). There was a single discrepancy in a tracing with T-wave inversions in leads V_1 and V_2 but not V_3. The criterion with the worst agreement was lead I QRS duration with a kappa of 0.5.

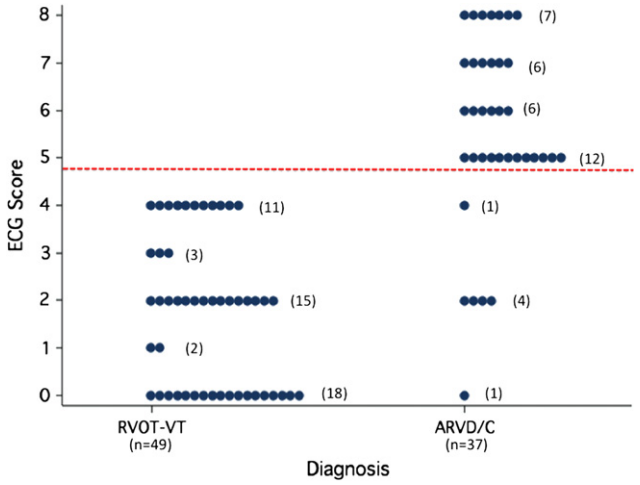

Figure 3 A dot plot showing the electrocardiogram (ECG) score of all patients stratified by diagnosis, with idiopathic ventricular tachycardia (VT) on the left and arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) on the right. Each "circle" represents a scored ECG. The number of patients with the individual score is noted in parentheses. All patients with idiopathic VT scored 4 or less, while the majority of patients with ARVD/C scored a 5 or greater. In fact, 7 patients with ARVD/C scored the maximum of 8 points. A score of 5 or greater was seen only in the ARVD/C group (red line).

Sensitivity analysis with patients with PVC only vs patients with VT only

In a subgroup analysis just including patients with PVCs (n = 60 total; ARVD/C = 15 and idiopathic VT = 45), the results remained the same. A score of 5 or greater had a sensitivity of 80%, specificity of 100%, positive predictive value of 100%, and negative predictive value of 94% in identifying a patient with ARVD/C; this score correctly classified 95% of the tracings. The area under the ROC curve was 0.93 ± 0.04 (95% CI 0.85–0.99).

Alternatively, in a subgroup analysis just including patients with VT (n = 26; ARVD/C = 22 and idiopathic VT = 4), the results were very similar. A score of 5 or greater had a sensitivity of 82%, specificity of 100%, positive predictive value of 100%, and negative predictive value of 50% (only 4 patients with idiopathic VT included) in identifying a patient with ARVD/C; this score correctly classified 85% of the tracings. The area under the ROC curve was 0.87 ± 0.06 (95% CI 0.74–0.99).

Epsilon waves

Epsilon waves, the pathognomonic reproducible low-amplitude signals between the end of the QRS complex and the onset of the T wave in the right precordial leads V_1–V_3, were seen only in 2 of 36 (5.6%) patients with ARVD/C. This finding continues to be very helpful if seen; however, in most series, like ours, it is seen in the minority of patients (10%–37%).[11]

Site of successful VT/PVC origin during ablation

Sixteen of thirty-six patients underwent ablation in the ARVD/C cohort. The most common locations were the epicardial RVOT (n = 7), freewall (n = 5), and anteroseptal RVOT (n = 4). The idiopathic VT cohort ablation locations were almost entirely septal, with anteroseptal (24 of 49, 49%) and posteroseptal (16 of 49, 33%) being most common with the remaining locations including mid-septal (8 of 49, 16%) and nonseptal/freewall (1 of 49, 2%).

Discussion

When a patient presents with an LBBB/inferior axis VT/PVC, the question frequently arises whether a more extensive workup is necessary to exclude ARVD/C. Echocardiographic findings suggestive of RV dilatation or aneurysms typically result in further evaluation, but echocardiographic examination of the right ventricle is often limited and many patients with ARVC may have normal screening ECGs.[9] Distinguishing LBBB/inferior axis ventricular arrhythmias between patients with ARVD/C and idiopathic VT is of great clinical importance, as those with ARVD/C and VT/PVCs typically warrant prophylactic drug and/or treatment with an implantable defibrillator to prevent sudden death while those with idiopathic VT have a benign course.[12–14] We describe a simple scoring

Table 3 Prospective cohort testing characteristics

Test algorithm	ARVD/C (n = 37)	Idiopathic VT (n = 49)	*P*	Sensitivity	Specificity	Positive predictive value	Negative predictive value	Correctly classified
ARVD/C risk score ≥5	31 (84%)	0 (0%)	<.0001	84%	100%	100%	91%	93%
Anterior T-wave inversions	29 (78%)	2 (4%)	<.0001	78%	96%	94%	85%	88%
Lead I QRS duration ≥120 ms	27 (73%)	21 (43%)	.005	73%	58%	56%	74%	64%
QRS notching (multiple leads)	25 (69%)	19 (40%)	.014	68%	59%	56%	74%	63%
V_5 transition or later	13 (35%)	4 (8%)	.002	35%	92%	76%	65%	67%

ARVD/C = Arrhythmogenic right ventricular dysplasia/cardiomyopathy; VT = ventricular tachycardia.

algorithm using 12-lead ECG characteristics that proved highly effective in distinguishing the etiology of RVOT arrhythmias in a cohort of patients with ARVD/C with a sensitivity of 84% and specificity of 100%. This means an ARVD/C risk score of 5 or greater clearly identifies those patients who require further evaluation to exclude ARVD/C. To our knowledge, this is the largest series of patients with ARVD/C with LBBB/inferior axis ventricular arrhythmias that have been described.

A score of 5 or greater was able to correctly distinguish the diagnosis of ARVD/C and idiopathic VT 94% of the time, with a sensitivity of 84%, specificity of 100%, positive predictive value of 100%, and negative predictive value of 91%. The clinical utility of our ARVD/C ECG score lies particularly in the 100% specificity and positive predictive in tracings that score 5 or greater, ruling out idiopathic VT as the diagnosis.

The differences observed in the ECG characteristics between the two groups may be due to several factors. Lead I QRS duration, QRS notching, and precordial QRS transition were all statistically significant predictors that favored ARVD/C. Among the 3 characteristics in this validation cohort, late precordial transition was the most specific and lead I QRS duration was the most sensitive, as in the original cohort. The latter is in agreement with Ainsworth et al,[15] who were the first to describe lead I QRS duration ≥120 ms as more common in ARVD/C. The fibrofatty replacement of the myocardium of the right ventricle delays cell-to-cell conduction, which can facilitate the development of reentrant arrhythmias and cause prolongation of the QRS complex and irregularities of conduction manifesting as notching of the QRS complex.[4] Abnormalities of repolarization in sinus rhythm lead to the right precordial T-wave inversions. Origin of the arrhythmias from nonseptal sites in ARVD/C may account for a later precordial transition and QRS notching.[7,16,17]

T-wave inversions were found to be the most significant predictor of ARVD/C. The overall score, however, performed better in identifying patients with ARVD/C than T-wave inversions alone. The 100% specificity and positive predictive value of the ECG score improves on anterior T-wave inversions alone. While recent data have shown that these T-wave inversions may be present in only 32% of patients with ARVD/C as well as 1%–3% of normal young patients,[9,18–20] our prospective cohort exhibited T-wave inversions in 29 of 37 (78%) of ARVD/C tracings and 2 of 49 (4%) idiopathic VT tracings. Among the 8 cases of ARVD/C that did not have anterior T-wave inversions, our scoring system made the diagnosis of ARVD/C in 2 (25%). Conversely, of the 2 patients with idiopathic VT with T-wave inversions in sinus rhythm, neither case met any further criteria, with a final score of 3 placing them correctly in the idiopathic VT category in each case. The reason why our ARVD/C tracings had such a high percentage of anterior T-wave inversions is unknown, conceivably there may be a link between the findings of early precordial T-wave inversions and PVCs from the outflow tract in ARVD/C.

The overall agreement for the ECG scoring system was 96% between the 2 expert electrophysiologists blinded to the clinical data (kappa of 0.82). A kappa greater than 0.8 is considered "almost perfect."[10] The characteristic that showed the lowest agreement was lead I QRS duration ≥120 ms with a kappa of 0.5, which by most statistical authorities is still considered a substantial agreement.[10] This fits with recently published results from Jain et al[21] that showed anterior T-wave inversions and right bundle branch block had a higher consistency among ECG parameters with more variability in the measurement of QRS duration.

The presence of more than one form of VT should increase the pretest probability for ARVD/C, as this likely reflects more diffuse pathology affecting multiple sites of the right ventricle.[22,23] Multiple VT forms including LBBB/superior axis essentially excludes idiopathic VT. In our study, at least 6 patients with ARVD/C had different VT morphologies; however, we included arrhythmias only from the RVOT and not from non-RVOT areas (inflow, RV apex, etc). Multiple PVC/VT morphologies seen were not used in our scoring system, because we did not specifically look for this parameter in our original study cohort.

Strengths and limitations

As with our original cohort, a limitation is our relatively small sample size. ARVD/C is a rare disease, and we limited our analysis to LBBB/inferior axis ventricular arrhythmias. However, to our knowledge, this is the largest collection of LBBB/inferior axis ventricular arrhythmias published in patients with ARVD/C.

Similarly, we increased our sample size by including patients with VT and PVCs. The score was developed in our original cohort that included both VT and PVCs, and there was no difference in the use of the criteria for each group; consequently, we included both groups for the present study.

This agrees with the study of Ainsworth et al,[15] who previously analyzed patients with PVCs and found identical findings compared with those of VT. In fact, our sensitivity analyses looking at PVCs only or VT only was remarkably similar.

In contrast to prior reports, we found a very high percentage of patients with ARVD/C with anterior T-wave inversions, which proved to be a major factor for skewing the scoring system toward the diagnosis of ARVD/C. Nevertheless, in 25% of those that did not have this finding, the scoring system correctly diagnosed ARVD/C. In addition, 2 patients with idiopathic VT who had anterior T-wave inversions were correctly diagnosed as idiopathic VT, since none of the other criteria were met.

A strength of our study is that we included patients with only definite ARVD/C by using the modified task force criteria from tertiary centers experienced in the care of patients with ARVD/C.

In order to maximize clinical utility and ease of use, we kept our individual scoring components limited to easily identifiable 12-lead ECG characteristics without the need for more sophisticated computerized measurements. We see the simplicity of the scoring system as a strength, as it can be universally used clinically without the need of more advanced computerized measurements.

We did not attempt to correlate the ARVD/C ECG score with severity of disease. We have no a priori indication of more severe diseased group, as consecutive patients were enrolled from participating centers.

The majority of patients with ARVD/C were on antiarrhythmic drug therapy. Class 1C agents may play a role in QRS prolongation and notching, as these drugs have important effects on cell-to-cell conduction. However, most patients were on sotalol and only 2 on class 1C agents.

This test should not be used as a substitute for the revised task force criteria. The ARVD/C inclusion criteria included only patients with revised task force criteria and did not prospectively enroll patients with PVCs and then ascertain their diagnosis. We did not compare the accuracy of our scoring system with the revised task force criteria.

Conclusions

We describe an easy-to-use ECG scoring algorithm that is very effective in distinguishing RVOT arrhythmias originating from patients with ARVD/C vs patients with idiopathic VT.

Appendix A. Characteristic ECG Features and Score

Supplementary data

Supplementary data associated with this article can be found in the online version at http://dx.doi.org/10.1016/j.hrthm.2012.12.009.

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