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

(Heart Rhythm 2013;10:477–482)

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

The abstract of this article was presented at the Heart Rhythm Society Scientific Sessions 2012 in Boston, MA. Dr Marcus has received speaker fees from Biotronik and St Jude Medical and research support from St Jude Medical and Astellas. Dr Dixit has received research grant from Medtronic and honoraria from Biosense and St Jude Medical. Dr Calkins has received research support from Medtronic, Boston Scientific, and St Jude Medical. Dr Scheinman has received speaker fees for lectures from St Jude Medical, Boston Scientific, Medtronic, Biosense, and Biotronik and consultant fees from Jansen. **Address reprint requests and correspondence:** Dr Melvin M. Scheinman, University of California, 500 Parnassus Avenue, MUE 434, San Francisco, CA 94143-1354. E-mail address: scheinman@medicine.ucsf.edu.

1547-5271/$-see front matter http://dx.doi.org/10.1016/j.hrthm.2012.12.009

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.