Implantable Cardioverter-Defibrillators in Patients With Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy

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OBJECTIVES
The aim of this study was to assess the outcome of arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) patients treated with an implantable cardioverter-defibrillator (ICD).

BACKGROUND
Arrhythmogenic right ventricular dysplasia/cardiomyopathy is associated with tachyarrhythmia and an increased risk of sudden death.

METHODS
This study included 42 ARVD/C patients with ICDs (52% male, age 6 to 69 years, median 37 years) followed at our center.

RESULTS
Mean follow-up was 42 ± 26 months (range 4 to 135 months). Complications associated with ICD implantation included need for lead repositioning (n = 3) and system infection (n = 2). During follow-up, one patient died of a brain malignancy and one had heart transplantation. Lead replacement was required in six patients as a result of lead fracture and insulation damage (n = 4) or change in thresholds (n = 2). During this period, 33 of 42 (78%) patients received a median of 4 (range 1 to 73) appropriate ICD interventions. The median period between ICD implantation and the first firing was 9 months (range 0.1 to 66 months). The ICD firing storms were observed in five patients. Inappropriate interventions were seen in 10 patients. Predictors of appropriate firing were induction of ventricular tachycardia (VT) during electrophysiologic study (EPS) (84% vs. 44%, p = 0.024), detection of spontaneous VT (70% vs. 15%, p = 0.001), male versus female gender (91% vs. 65%, p = 0.04), and severe right ventricular dilation (39% vs. 0%, p = 0.013). Using multivariate analysis, VT induction during EPS was associated with increased risk for firing in ARVD/C patients; odds ratio 11.2 (95% confidence interval 1.23 to 101.24, p = 0.031).

CONCLUSIONS
Patients with ARVD/C have a high arrhythmia rate requiring appropriate ICD interventions. The ICD therapy appears to be well tolerated and important in the management of patients with ARVD/C. (J Am Coll Cardiol 2004;43:1843–52) © 2004 by the American College of Cardiology Foundation

Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) is a myocardial disease characterized by ventricular arrhythmias in the setting of structural and functional abnormalities of the right ventricle (RV) due to replacement of myocardium by fatty and fibrous tissue (1–6). Diagnosis of ARVD/C is often difficult because there is no single test that can be used either to establish or exclude this diagnosis (7–9). However, the results of a history, physical examination, and a number of specific cardiac tests can be used to establish the diagnosis (2,10,11). Patients with ARVD/C generally come to medical attention because of symptoms that result from arrhythmias arising in the RV. The most common presenting symptoms are palpitations, chest discomfort, near syncope, or syncope. The arrhythmias range from isolated premature ventricular beats to sustained ventricular tachycardia (VT) or ventricular fibrillation (VF) that may lead to sudden death (3,4).

Once a diagnosis of ARVD/C is established, the most important management decision is whether to implant an implantable cardioverter-defibrillator (ICD) for treatment of sustained ventricular arrhythmias and prevention of sudden cardiac death (SCD) (12). This is a critically important decision because these are young patients with few or no symptoms who are expected to live many years with a device that is not complication-free. Furthermore, ICDs require replacement every five years, and the probability of lead failure increases with time. In addition, magnetic resonance imaging, which is valuable in the evaluation and follow-up of patients with ARVD/C, is presently contraindicated after ICD implantation.

Despite the importance of the decision to implant an ICD, remarkably few data exist concerning the safety and efficacy of ICD therapy in ARVD/C patients (13–25). Therefore, the purpose of this study is to present the clinical characteristics and follow-up of a large series of ARVD/C patients who received ICDs. Particular attention is focused
on defining the frequency and predictors of appropriate and inappropriate ICD therapies, the prognostic importance of experiencing VT storm, and the incidence of short-term and long-term complications in this patient population.

**METHODS**

**Patient population.** This study was approved by the institutional review board of the Johns Hopkins University. The Johns Hopkins ARVD/C Program was established in 1995 to provide clinical care for patients with ARVD/C and to study this disease. At the initiation of this program, a registry was established consisting of the medical records of patients from North America diagnosed with ARVD/C. All those in the registry who had implantation of an ICD, a follow-up period of at least three months after implantation of the device, and documentation of the clinical outcome as of September 2002 were screened and invited to participate in the study. The population in the present study is comprised of all patients who agreed and signed the consent form. Files were reviewed for clinical and ICD records. Patients were contacted by telephone in the period from October through December 2002. A detailed questionnaire was completed, and in cases in which data were missing, documents and/or interrogations were sent to the registry for further evaluation and investigation.

**Diagnosis of ARVD/C.** The diagnosis of ARVD/C is based on the presence of certain major and minor diagnostic criteria employed by the international task force criteria (7) (Table 1). Each major criterion is scored 2 points, and each minor criterion is scored 1 point. The diagnosis is established by the presence of two major criteria or one major criterion plus two minor criteria or four minor criteria. The patients were classified as having “definitive” ARVD/C when they met the full criteria (4 points or more).

**Defibrillators.** The patients in this study underwent placement of their initial ICD between July 1991 and February 2002. Implantation was performed through a thoracotomy with epicardial lead systems in three patients, all of whom were later implanted with transvenous systems. Most of the devices were third- and fourth-generation defibrillators with the capacity to provide antitachycardia and antibradycardia pacing; the devices had diagnostic memory and the ability to record and store electrocardiographic data, including intra-

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**Table 1. ARVD Diagnosis Criteria**

<table>
<thead>
<tr>
<th>Major Criteria</th>
<th>Minor Criteria</th>
</tr>
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<tbody>
<tr>
<td>Structural abnormalities in the right ventricle</td>
<td>Mild to moderate structural abnormalities of the right ventricle</td>
</tr>
<tr>
<td>Abnormal myocardial tissue in the right ventricle with fatty infiltration</td>
<td>Ventricular arrhythmias such as sustained or nonsustained ventricular tachycardia with a left bundle morphology or more than 1,000 PVCs per 24 h</td>
</tr>
<tr>
<td>Repolarization abnormalities on the electrocardiogram with T-wave inversion in leads V1 to V3 or beyond</td>
<td>ECG abnormalities such as T-wave inversion in V2 and V3 or late potentials on a signal-averaged ECG</td>
</tr>
<tr>
<td>Conduction abnormalities on the ECG such as a QRS duration ≥100 ms in V1, V2, or V3 or an epsilon wave</td>
<td>A family history or premature sudden cardiac death</td>
</tr>
<tr>
<td>Family history of ARVD confirmed by autopsy results</td>
<td></td>
</tr>
</tbody>
</table>

Each major criterion is scored 2 points and each minor, 1 point. The diagnosis is established by the presence of two major criteria or one major criterion plus two minor criteria or four minor criteria. The patients were classified as having definitive ARVD/C when they met the full criteria (4 points or more).

ARVD/C = arrhythmogenic right ventricular dysplasia/cardiomyopathy; ECG = electrocardiogram; PVCs = premature ventricular contractions.
cardiac electrograms, for subsequent review. Programmed antitachycardia pacing was activated at the discretion of the cardiologist. Stored data were reviewed after all discharges. Interrogations were performed routinely every three to six months.

Classification of discharges and interpretation of intra-cardiac electrograms. When available, stored data were analyzed to classify the arrhythmias responsible for precipitating defibrillator discharges, according to the following definitions (26). Ventricular fibrillation or flutter was defined as ventricular arrhythmia with a cycle length of 240 ms or less. Ventricular tachycardia was defined as a regular (monomorphic) or irregular (polymorphic) ventricular arrhythmia with a cycle length of more than 240 ms. Defibrillator shocks were considered appropriate versus inappropriate on the basis of standard criteria (24). When complete ICD interrogation information was not available on a given patient, we relied on the outside the electrophysiologist interpretation of the ICD tracings. Electrical storm was defined according to the definition of Credner et al. (27) as the occurrence of VT or VF resulting in device intervention (antitachycardia pacing and/or shock delivery) three or more times within a 24-h period.

Statistical analysis. Continuous data are expressed as means ± SD. The cumulative probability of survival was determined by the Kaplan-Meier method (28), and differences in survival between groups were evaluated with the log-rank test. Multivariate analysis was performed using variables that in univariate analysis were found statistically significant as well as conditional logistic regression analysis.
adjusting for variables that may influence ICD firing. A value of $p < 0.05$ was considered statistically significant.

**RESULTS**

**Patient population.** Data from 87 patients with ICDs was screened for this study. Thirteen patients who did not meet the criteria for ARVD/C and 22 patients who were classified as “probable” ARVD/C (score = 3) (Table 1) were excluded from the study population. Ten additional patients were contacted but did not return a signed consent form and therefore were also excluded. Thus, the final patient population analyzed in this study consisted of 42 patients all classified as definitive ARVD/C, 22 (52%) of whom were male. Shown in Figure 1 is the distribution in patients’ age at the time of ICD implantation. The clinical characteristics of the patients in this study are summarized in Table 2. The age range was 6 to 69 years (mean age, 36 ± 13 years, median 37 years); 28 patients (66%) were <41 years old. Seven patients were engaged in intense sport activity at the time of presentation, and 20 were involved in mild activity.

**Implantation of ICD systems.** All patients received multifunctional third- or fourth-generation ICDs. The implanted device was a single-chamber device in 27 (64%) patients and a dual-chamber device in 15 (36%) patients. The mean R-wave amplitude at the time of ICD implantation was 9.3 ± 4.6 mV. The R-wave amplitude in four patients was ≤5 mV (3.4 mV, 3.9 mV, 4.0 mV, and 4.3 mV). Appropriate sensing was achieved in all patients at the time of device implantation. The mean pacing threshold achieved at the time of device implantation was 0.85 ± 0.65 V. The pacing threshold was >1 V in five patients (1.3 V, 1.5 V, 2.0 V, 3.0 V, and 3.0 V). The mean defibrillation threshold was 13.5 ± 4.4 V. No patient had a defibrillation threshold ≥20 J.

In 4 (10%) patients, ICDs were implanted after either resuscitation from cardiac arrest or after experiencing an episode of sustained, spontaneous VT, and in 21 (50%) the reason was a syncopal episode. Of the remaining 17 patients, 15 had a history of one or more family sudden deaths (n = 7, of whom 5 had inducible VT on electrophysiologic testing), VT induction in electrophysiologic study (EPS) (n = 13), and left ventricular (LV) involvement (n = 2). Two patients had none of these high-risk markers. The major implantation reason and EPS findings in the study patients are summarized in Table 3.

Among the remaining 17 (40%) patients, 7 had a history of one or more family sudden deaths, 3 had a family history of ARVD/C, 13 had VT induction in EPS, 2 had also LV involvement, and 1 had fat in biopsy. Two of these patients had none of these high-risk markers.

In this study, all but one patient had an EPS before device

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**Table 3. Main Indications for ICD Implantation in the Study Patients**

<table>
<thead>
<tr>
<th>Main Reason for ICD Implantation</th>
<th>Number of Patients</th>
<th>Inducible VT in EPS*</th>
<th>Monomorphic</th>
<th>Polymorphic</th>
<th>Need for Beta-Agonists to Induce VT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sudden death*</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Sustained VT†</td>
<td>2</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Syncope</td>
<td>21</td>
<td>18</td>
<td>11</td>
<td>7</td>
<td>5</td>
</tr>
<tr>
<td>Familial sudden death</td>
<td>7</td>
<td>5</td>
<td>4</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Familial ARVD/C</td>
<td>3</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Near syncpe/palpitation that led to the diagnosis of definitive ARVD/C</td>
<td>10</td>
<td>4</td>
<td>3</td>
<td>1</td>
<td>3</td>
</tr>
</tbody>
</table>

*EPS was performed in 41 patients in this study; one patient of the sudden death group did not have an EPS. †All monomorphic VTs in this group had left bundle-branch block pattern.

ARVD/C = arrhythmogenic right ventricular dysplasia/cardiomyopathy; EPS = electrophysiologic study; ICD = implantable cardioverter-defibrillator; VT = ventricular tachycardia.

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**Figure 2.** (A) Interval between implantation of the defibrillator and the first appropriate discharge in 33 patients. (B) Number of appropriate implantable cardioverter-defibrillator (ICD) discharges in 33 patients.
implantation. The EPS revealed induced sustained ventricular arrhythmia in 31 patients (74%) (polymorphic in 12 and monomorphic in 19). Eight patients underwent catheter ablation before or at the time of ICD implantation. Three patients had an ablation performed after the ICD implantation (0.1 to 5 months).

A complication at the time of implantation occurred in five (8%) patients.

Three patients required lead repositioning within several days after the initial implant procedure owing to inadequate R-wave amplitude. None had myocardial perforation and/or tamponade. One patient with an acute infection underwent system removal and replacement, whereas a second patient with subacute infection was treated successfully with antibiotics.

**Patient follow-up.** After a mean follow-up of 42 ± 26 months (range 4 to 135 months), one patient died of a brain malignancy, and in one patient who had end-stage disease with severe biventricular dysfunction and cavity dilation, heart transplantation was performed. There were no cases of late infections, myocardial perforation, or the development of a thromboembolism. During follow-up, ICD leads were replaced in another six patients because of lead fracture or insulation damage (n = 4) or because of a decrease in R-wave amplitude (n = 2). These patients did not have more extensive disease. The ICD replacement was performed in 10 patients because of battery depletion.

**ICD discharges.** Thirty-three (78%) patients received a median of 4 (range 1 to 75) appropriate ICD interventions. Thirty patients were treated with defibrillation shocks and three with antitachycardia pacing. Ventricular tachycardia was the arrhythmia treated by the ICD in 92% of events. The patients were involved at different levels of exercise in 62% of ICD firing events. Shown in Figure 2A is the interval between ICD implantation and the first appropriate ICD intervention (median 9 months, range 1 week to 66 months). The interval was two or more years in eight (19%) patients. Conversely, 15 (35%) patients had an initial appropriate discharge <6 months after implantation; in 12 of these 15 patients, there was at least one additional discharge (Fig. 2B). With an average follow-up of 3.5 years after implantation, the rate of appropriate discharges for the study group was 22% per year.

The ICD firing storms were observed in five patients, who experienced 6, 7, 9, 30, and 42 shocks within 24 h, respectively. Four of these patients had advanced ARVD/C with RV dysfunction. The interval between the ICD implant and storm was 1 to 45 months. The VT storm episode was the first ICD discharge in four of these patients. In two patients, no further ICD firing occurred with further follow-up; in the other three patients, one, four, and five single appropriate firings, respectively, occurred during follow-up. After the electrical storm episode, none of the patients subsequently presented with a second storm episode and none required either catheter ablation or cardiac transplantation for management of their ventricular arrhythmias.

Fifty-nine percent of all patients who experienced an ICD therapy reported that it occurred in association with exercise or other type of physical exertion. The ICD discharge occurred at rest in only one patient. The remainder of patients who experienced an ICD discharge (38%) were engaged in mild physical activity at the time of the device therapy.

**Table 4** summarizes the antiarrhythmic drug therapy use among the 33 patients who received an appropriate ICD therapy and those who did not. After an appropriate ICD therapy, the use of antiarrhythmic drugs increased. This was particularly observed among the five patients who experienced VT storm. Each of these patients was receiving combination antiarrhythmic therapy at last follow-up. At the time of most recent follow-up, 22 patients (52%) were receiving beta-blockers, 8 patients (19%) were receiving amiodarone, and 11 patients (26%) were receiving sotalol.

Ten (24%) patients experienced inappropriate interventions. In 9 of the 10 patients, there was also at least one appropriate discharge. The inappropriate discharges (all defibrillation shocks) were due to sinus tachycardia (in six patients), another type of supraventricular arrhythmia in two patients, and electrical noise resulting from lead fracture in two patients. Eight patients had only one inappropriate discharge. Seven patients had a single-lead device, whereas...
three had a dual-lead device. In the latter group the underlying rhythm was supraventricular tachycardia (in two patients) and lead disruption (in one patient).

During the follow-up period, nine (21%) patients did not experience an ICD firing. Two (22%) of these patients had a history of sustained symptomatic VT. Two (22%) had mild or moderate RV dysfunction, and none had severe RV dysfunction. Electrophysiology testing was performed in all nine patients and was positive in three (33%).

**Predictors of appropriate ICD firings.** Shown in Table 2 are the variables that were analyzed as potential predictors of ICD firing. Among them, four variables were identified as predictors: 1) induction of VT during EPS; 2) detection of spontaneous non-sustained VT on electrocardiogram, Holter monitoring, or exercise treadmill test; 3) RV moderate to severe dilation as compared with no dilation; and 4) male gender. The cumulative rates of first appropriate ICD discharge according to these four variables are shown in

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**Figure 3.** Cumulative rates of first appropriate discharges according to (A) gender and (B) detection of spontaneous ventricular tachycardia (VT) in electrocardiogram, Holter monitoring, or exercise treadmill test. Continued on next page.
Figure 3. Predictors of appropriate ICD firing were induction of VT during EPS (81% vs. 44%, respectively, p = 0.024), detection of spontaneous VT (70% vs. 15%, respectively, p = 0.001), male versus female gender (91% vs. 65%, respectively, p = 0.04), and severe RV dilation (39% vs. 0%, respectively, p = 0.013).

Conditional logistic regression analysis was performed using variables that in univariate analysis were statistically significant (Table 2) and adjusting for several variables that may influence ICD firing (signal-averaged electrocardiogram, age, beta-blockers usage, syncope, LV function, RV function). After adjustment for all of these variables, the induction of VT in EPS remained statistically significant (odds ratio [OR] 11.2, 95% confidence interval [CI] 1.23 to 101.24, p = 0.031). There was a trend toward RV dilation OR 3.41 (95% CI 0.88 to 14.21, p = 0.07) and, with male gender, OR 2.64 (95% CI 0.80 to 8.71, p = 0.11).

DISCUSSION

This study reports the outcome of 42 patients with ARVD/C who were treated with ICD therapy. The results of our study establish an important role for ICD therapy in the treatment of ventricular arrhythmias and possibly in the prevention of sudden death in patients with ARVD/C. Three findings of this study are of particular importance.
First, more than three-quarters of ARVD/C patients received an appropriate ICD therapy during an average of 3.5 years of follow-up. The ICD proved reliable in sensing and terminating sustained ventricular arrhythmias, with no patients experiencing SCD. Second, this study identifies predictors of an appropriate ICD therapy; induction of VT during electrophysiologic testing, detection of nonsustained VT on noninvasive monitoring, male gender, and severe RV dilation. A third finding of this study is that ICD therapy is feasible and safe in patients with ARVD/C, with a low incidence of short-term and long-term complications.

Arrhythmogenic right ventricular dysplasia/cardiomyopathy is an uncommon but important cause of SCD, as most of these individuals are young. It is estimated that it accounts for up to one-fifth of all episodes of SCD that occur in patients below the age of 35 years (3). Although the role of ICD therapy for primary prevention of sudden death in patients with ischemic heart disease and for secondary prevention of SCD is well established (29–34), few studies have examined the role of ICD therapy in patients with ARVD/C (13–16). Those studies have enrolled relatively small numbers of patients. It is therefore not surprising that in the recently published jointrecommendations of the American College of Cardiology and the American Heart Association, ARVD/C is not listed as a standard indication for placement of an ICD (12). Calkins and Marcus (3) have proposed that an ICD should be implanted in ARVD/C patients with an increased risk for SCD, based on the presence of a previous cardiac arrest, syncope due to VT, evidence for extensive RV disease, LV involvement, and presentation with polymorphic VT and RV apical aneurysm (which is associated with a genetic locus on chromosome 1q42-43). In our population, only five had severe LV dysfunction, three of whom had ICD firing.

In many patients in the present study, ICDs were implanted prophylactically for primary prevention of sudden death. This differs from clinical data and recommendations on ARVD/C derived from European experience (20,24,25,31). It seems that the clinical approach and decision making regarding ICD implantation may be different in the U.S. Still, this is the first long-term study which implies that prophylactic ICD implantation for primary prevention of sudden death in ARVD/C is feasible.

Several previous studies have examined the safety and efficacy of ICD therapy in patients with ARVD/C. Tavernier et al. (16) reported that six of nine ARVD/C patients received appropriate firings after a follow-up of 32 ± 24 months. Inappropriate interventions were seen in five patients. Link et al. (14) reported that 8 of 12 patients with ARVD/C received appropriate therapy after a follow-up of 22 ± 13 months (range 1 to 45 months). They reported no short-term or long-term complications of RV lead placement and one sudden death at one month of follow-up. Breithardt et al. (13), in the largest series published so far, reported that 9 of 18 patients received appropriate therapy after a follow-up of 17 ± 11 months (range 1 to 40 months).

The results of the present study confirm and extend the findings of these previous reports of ICD therapy in ARVD/C patients. The 78% appropriate ICD discharge rate during a mean follow-up of 3.5 years in this study is remarkably similar to these previous reports. This rate is similar to the rate of appropriate ICD discharges reported for the long QT syndrome (33) but greater than has been reported for patients with coronary artery disease (31), hypertrophic cardiomyopathy (26), or idiopathic VF (13,32). The high ICD firing rate observed in this trial likely reflects a number of factors, including the clinical characteristics of the patient population, the prophylactic use of beta-blocker therapy in less than one-third of patients, and the small proportion of the patient population that were receiving class 1 and 3 antiarrhythmic agents after device implantation.

Although there was no statistically significant difference in beta-blockers usage between those who had an ICD firing and those who did not, there is a small trend towards a protective effect of beta-blockers confirming clinical experience. We believe that beta-blocker therapy should be considered prophylactically for all ARVD/C patients who undergo placement of an ICD.

The present study of ICD therapy in ARVD/C patients is unique because the larger patient population allowed us to examine predictors of receiving an appropriate ICD discharge and also to examine the incidence of experiencing VT storm. Multivariate analysis revealed that induction of VT during EPS was the most significant predictor for ICD firing in patients with definitive diagnosis of ARVD/C. It is notable that the interval between ICD implantation and a first appropriate ICD intervention exceeded two years in 19% of patients with appropriate discharges.

Five patients experienced an ICD firing storm between 1 and 45 months after implantation. Four of these patients had advanced ARVD/C with RV dysfunction. It is notable that these VT storms occurred on only a single occasion in these patients and were successfully controlled with alterations in antiarrhythmic drug therapy. None of these patients died during follow-up or required cardiac transplantation.

There are several potential complications of ICD therapy, which may be more likely to occur in ARVD/C patients as a result of the replacement of RV myocardium with fat and fibrotic tissue. These include an increased risk of: 1) perforation caused by thinning of the RV wall; 2) difficulty in lead placement owing to inadequate R-wave amplitudes or high pacing thresholds; 3) inadequate sensing or pacing during follow-up resulting from disease progression and deterioration of R-wave amplitudes and rising pacing thresholds; and 4) failure to terminate ventricular arrhythmias owing to rising defibrillation thresholds over time resulting from disease progression. Despite these concerns, it is somewhat reassuring that in this cohort of ARVD/C
patients, the short-term and long-term risks of ICD therapy were similar to those in previous reports in non-ARVD/C patient populations (35,36).

Currently, there is no firm evidence to determine whether a single chamber or a dual chamber ICD is preferable for patients with ARVD/C. For patients with concomitant sinus node dysfunction, a dual chamber ICD is clearly preferred. However, for patients with appropriate sinus node function and excellent AV nodal conduction, the potential benefits of better discrimination of atrial arrhythmias must be balanced against the long-term complications associated with increased hardware in patients. This is particularly relevant because most of these patients are young and will require more than 30 years of device therapy. Dual chamber devices allow for better discrimination between supraventricular arrhythmias and ventricular arrhythmias. This is especially important because exercise was identified as a common precipitant of arrhythmias in this and previous studies of ARVD/C (37).

Study limitations. There are several limitations to consider when interpreting the results of this study. First, the average follow-up in this patient series was 3.5 years. Because ARVD/C is a progressive disease, it is possible that additional complications of ICD systems may become apparent with longer follow-up. Secondly, sustained VT are not always sine qua non to sudden death, because they may be tolerated by patients with ARVD/C. We did not have complete information on the cycle lengths of all treated arrhythmias or on the detailed symptoms that patients experienced at the time of all device therapies. When intracardiac tracings were unavailable for review, we relied on the outside the electrophysiologist interpretation of the tracing for classification of the device therapy as appropriate or inappropriate and also as to whether the arrhythmias were VT or VF. Although some may interpret the high rate of appropriate ICD interventions as a clear recommendation to implant ICDs in all ARVD/C patients, the results of this study do not allow us to draw this conclusion. It is important to note that a far more conservative approach to classification of the device therapy as appropriate or inappropriate: ICD therapy as appropriate or inappropriate classification of the device therapy as appropriate or inappropriate.

The results of this study confirm that ICD therapy is safe and effective in patients with ARVD/C. The ICD provides lifesaving protection by effectively terminating ventricular tachyarrhythmias in patients with ARVD/C, many of whom are young. These results also represent an initial step toward developing better criteria for ICD implantation in patients with ARVD/C. The results of this study provide support for the high-risk markers proposed by Marcus (3,37) but also identify several new risk factors for experiencing an appropriate ICD intervention.

REFERENCES