Subcutaneous implantable cardioverter defibrillator in patients with arrhythmogenic right ventricular cardiomyopathy: Results from an Italian multicenter registry☆

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ABSTRACT

Background: Despite expanding indication of the subcutaneous implantable cardioverter defibrillator (S-ICD) in clinical practice, limited data exists on safety and efficacy of S-ICD in arrhythmogenic right ventricular cardiomyopathy (ARVC) patients. The aim of this multicenter study was to evaluate the safety and efficacy of S-ICD in ARVC patients.
Methods: The study population included 44 consecutive patients with definite ARVC diagnosis according to the 2010 ITF criteria (57% male, mean age 37 ± 17 years [range 10–75 years]) who received an S-ICD. Eighteen (41%) patients were implanted for secondary prevention.
Results: At implant, all inducible patients (34/44) had conversion of ventricular fibrillation at 65 J. No early complications occurred. During a median follow-up of 12 months (7–19), 3 (6.8%) patients experienced complications requiring surgical revision. No local or systemic device-related infections were observed. Six patients (14%) received a total of 61 appropriate and successful shocks on ventricular arrhythmias. Six (14%) patients experienced 8 inappropriate shocks for oversensing of cardiac signal (4 cases) and non-cardiac signal (4 cases) with one patient requiring device explantation. No patients had the device explanted due to the need for antitachycardia pacing.
Conclusions: The study shows that S-ICD provides safe and effective therapy for termination of both induced and spontaneous malignant ventricular tachyarrhythmias with high energy shocks in ARVC patients, but the risk of inappropriate shocks and complications needing surgical revision should be considered.
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1. Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inheritable heart muscle disease that predominantly affects the right ventricle (RV) and predisposes to ventricular arrhythmias (VA) and sudden cardiac death (SCD) [1]. Although ICD therapy provides the most effective life-saving protection for patients with ARVC, by effectively terminating life-threatening VA [1–4], the transvenous ICD (TV-ICD) system is associated with sizable morbidity and device-related complications requiring surgical revision such as lead failure/fracture, infection and inappropriate ICD interventions, especially in younger population [1–6].

Subcutaneous ICD (S-ICD) has recently entered into the clinical practice and may represent a valid alternative to the TV-ICD, especially among patients with limited vascular access, increased risk of infection and structurally normal heart with no need for pacing [7–9]. However, the precise clinical role of S-ICD in ARVC patients remains to be defined because of the possibility of electrocardiographic depolarization/repolarization changes leading to double QRS counting and P- or T-wave oversensing and potential inappropriate shock delivery [10,11].

The aim of this multicenter study was to evaluate the efficacy and safety of S-ICD in ARVC patients.

2. Methods

The study population consisted of 44 patients diagnosed with definite ARVC according to the 2010 ITF criteria [12] who underwent S-ICD implantation for SCD prevention and were enrolled in the Italian "RHYTHM DETECT" registry from 12 Italian centers. Seven patients were included in a previous study reporting the preliminary experience of S-ICD in ARVC patients [13].

Baseline clinical characteristics, electrocardiographic abnormalities, indication for implantation, electrocardiogram (ECG) screening, and technical device characteristics were prospectively collected. The local Ethics Committee approved the study protocol and all patients provided written consent to be enrolled in the registry. The study protocol conforms to the ethical guidelines of the 1975 Declaration of Helsinki as reflected in a priori approval by the institution's human research committee.

2.1. S-ICD implantation technique, defibrillation testing and device programming

Before implantation, all patients were screened for eligibility for S-ICD using the Boston Scientific manual ECG screening tool or the automated screening tool based on the surface ECG limb lead recording over the left and/or right parasternal regions to simulate the three S-ICD sensing vectors. The implantation technique used was the conventional technique suggested by the manufacturer described in the S-ICD User's Manual, or the two-incision intermuscular technique. Briefly, the two-incision intermuscular technique abandons the superior parasternal incision and consists of creating an intermuscular pocket (between the anterior surface of the *serratus anterior muscle* and the posterior surface of the *latissimus dorsi muscle*) for the pulse generator instead of a subcutaneous pocket using anatomical landmarks (see Supplementary data for details).

2.2. Follow-up

All patients were followed-up at 1 month, 3 months, and every 6 months thereafter. At these visits, patient's clinical conditions, S-ICD interrogation and complications were assessed. Acute complications were defined as complications that occurred during or within 24 h of S-ICD implantation and were classified as (1) procedure-related complications, including pneumothorax, pleural effusion, hematoma 2 cm, drop in hemoglobin >2 g/dL, bleeding requiring wound exploration or transfusion or generator/lead dislocation at the chest x-ray obtained 24 h after the procedure; and (2) technical complications, such as failure of the device to communicate with the programmer. Late complications were defined as those occurring >24 h after the procedure and included: pocket discomfort, incomplete wound healing, skin erosion of pulse generator or electrode, local and systemic device-related infections, and migration of pulse generator or electrode and technical complications such as failure of the device to communicate with the programmer or premature battery depletion. Captured S-ECG tracings from all shock episodes stored in the S-ICD were interpreted by two experienced electrophysiologists. In case of disagreement, a third cardiologist was consulted. Interventions were considered inappropriate when triggered by anything other than ventricular tachycardia (VT) or ventricular fibrillation (VF) above the programmed rate zone, including supraventricular arrhythmias (SVT) cardiac/non cardiac oversensing or device or lead malfunction. Cardiac oversensing, was defined as T-wave oversensing (TWOS), QRS oversensing, P-wave oversensing or oversensing due to a low-amplitude signal, and other/combined types of cardiac oversensing. Non-cardiac oversensing, was defined as any kind of oversensing due to non-cardiac causes (e.g., electromagnetic interference and myopotentials). Episodes of inappropriate therapy were reviewed and verified with Boston Scientific Technical support team.

2.3. Statistical analysis

Categorical differences between groups were evaluated by using the χ^2 test or the Fisher exact test as appropriate and presented with actual numbers and frequencies. Continuous variables were expressed as mean ± standard deviation (SD) or median with 25–75% for normally distributed and skewed variables, respectively, and compared with Student t-test or Wilcoxon rank sum test, as appropriate. Normal distribution of continuous variables was assessed by using the Kolmogorov-Smirnov test. Event-free survival curves were drawn with the Kaplan-Meier method. Patients were censored at the time of their first event or at the time of their last clinical follow-Up. A 2-tailed $p < 0.05$ was considered statistically significant. Data were analysed with SPSS, version 23 (IBM).

3. Results

3.1. Study population

The study population included 44 consecutive ARVC patients (57% male, mean age 37 ± 17 years [range 10–75 years]) who received an S-ICD implantation between April 2013 and March 2018 for SCD prevention. Baseline clinical characteristics are reported in Table 1. Eighteen (41%) patients were implanted for secondary prevention ($n = 17$ with sustained VT; $n = 1$ presented with SCD). All 26 (59%) patients who received an ICD for primary prevention had a IIa class recommendation for the device implantation according to the 2015 ITF consensus document [1]. Seven (16%) patients were ≤18 years of age. At the time of ICD implantation, 26 (59%) patients were being treated with a beta blocker while 13 (29.5%) were receiving an antiarrhythmic agent. Thirteen (30%) patients showed a significant LV involvement resulting into biventricular phenotype in 11 (25%) patients and left-dominant phenotype in 2 (4.5%). A positive electrophysiological study inducibility was reported in 3/13 (23%) patients and 14 (32%) subject presented non-sustained VT.

Table 1
Baseline clinical characteristics of the study population.

	$n = 44$
Male gender	25 (57)
Age, years	37 ± 17
Height, cm	174 ± 10
Weight, kg	73 ± 14
BMI	24 ± 4
LV ejection fraction	53 ± 11
LV ejection fraction ≤50%	13 (30)
New York Heart Association	
Class I	39 (89)
Class II	5 (11)
Previous transvenous ICD	7 (16)
Primary prevention	26 (59)
History of sustained VT	17 (39)
History of SCD	1 (2)
NSVT	14 (32)
Syncope	10 (23)
Inducibility at EPS	3/13 (23)
ECG characteristics	
Sinus rhythm	44 (100)
QRS duration, ms	94 ± 16
PQ interval	175 ± 4
QTc interval	429 ± 9
Negative T-wave in V1–V3 leads	26 (59)
Negative T-wave in lateral/inferior leads	14(32)
Epsilon wave	4 (9)
Medications at implant	
Beta-blockers	26 (59)
Amiodarone	2 (4)
Sotalol	11 (25)

Values are number of patients n (percentage, %) or mean ± standard deviation. BMI: body mass index; ICD: implantable cardioverter defibrillator; LV: left ventricular; VT: ventricular tachycardia; VF: ventricular fibrillation; NSVT: non-sustained ventricular tachycardia; EPS: electrophysiology study; SCD: sudden cardiac death.

3.2. ECG screening

Nine (20.4%) patients had all 3 suitable sensing vectors, 39 (89%) had at least 2 suitable sensing vectors. The secondary sensing vector was the most compatible (86%), followed by the primary vector (84%) and the alternate vector (39%). Three (6.8%) patients unsuitable became eligible on a single vector (the primary vector for 1 patient and the secondary vector for the other) after moving the sensing electrodes from the left to the right parasternal line. There were no cases with adjudication disagreement.

3.3. S-ICD implant characteristics

Nine (20.5%) patients were implanted with a first-generation S-ICD (Cameron Health model SQ-RX 1010) and 35 (79.5%) patients with the new S-ICD generation (Emblem Boston Scientific model A209, n = 7 and Emblem Boston Scientific model A219, n = 28). Baseline technical device characteristics are reported in the supplementary Table 1. In 33 (75%) patients the two-incision technique for the lead placement and in 25 (57%) patients the intermuscular technique was used for pulse generator placement. In 7 (16%) patients the S-ICD was implanted after removal of a previous TV-ICD system for complications. In the remaining patients, the choice of implanting a S-ICD instead of a TV-ICD was at the discretion of the physician based on clinical indications. The procedure was performed under general anesthesia in 17 (38.5%) patients, local anesthesia with sedation in 25 (57%) and with ultrasound-guided serratus anterior plane block in the remaining 2 (4.5%) patients. The average procedure time ("skin to skin") was 68 ± 24 min. Defibrillation testing (DT) was performed in 39 (88.6%) patients while VF could not be induced even after several attempts in 5/39 (12.8%) patients. Among 34 patients with inducible VF, all patients had conversion of the induced VF at an output of 65 J standard polarity at first attempt except in two (6%) who received a successful DT testing at 65 J with inverted polarity. Mean time from VF induction to shock delivery was 15 ± 5 s. No early complications occurred. A postoperative chest radiography confirmed stable device and lead location in all patients. Dual zone programming for tachyarrhythmia detection was chosen in all patients.

3.4. Follow-up

During a median follow-up of 12 months [7–19], 3(6.8%) patients experienced late complications in the absence of signs of infection, requiring surgical revision, including pulse generator pocket revision (no removal of the device), for incomplete superficial wound healing (n = 1), lead revision for skin erosion at the superior parasternal incision (n = 1) or lead dislodgement (n = 1) (Supplementary Fig. 2A). The 3 patients who required surgical revision belonged to the group of 11 who were implanted by using the traditional 3 incisions technique while no complications were observed among the 33 patients who were implanted with the 2 incisions technique (p = 0.01). No local or systemic device-related infections, and migration of pulse generator were observed. Six patients (14%) received a total of 61 appropriate and successful shock for VA (range 1–40) with a mean cycle length of 305.4 ± 45.51 milliseconds, consisting of 49 monomorphic VT (80%) and 12 polymorphic VT/VF (20%) (Supplementary Fig. 2B and Fig. 1). The time to therapy for spontaneous episodes was 16 ± 4 s. Two patients presented VT storms, one successfully treated with antiarrhythmic drug dose adjustment while the other underwent successful catheter ablation. Six (14%) patients experienced 8 inappropriate shocks (range 1–3) for oversensing of cardiac signal in 4 cases such as SVT (n = 1), TWOS (n = 2) and P-wave oversensing (n = 1) or noncardiac signal in 4 cases such as artifacts due to suspected transient air in the lead 24 h after the implant (n = 1), artifacts due to lead dislodgment, that resolved after surgical revision of the system (n = 1), and myopotential (n = 2) (Supplementary Fig. 2C, Figs. 2, 3). No

Fig. 1. S-ICD stored electrogram showing an appropriately detected and treated ventricular arrhythmia episode.
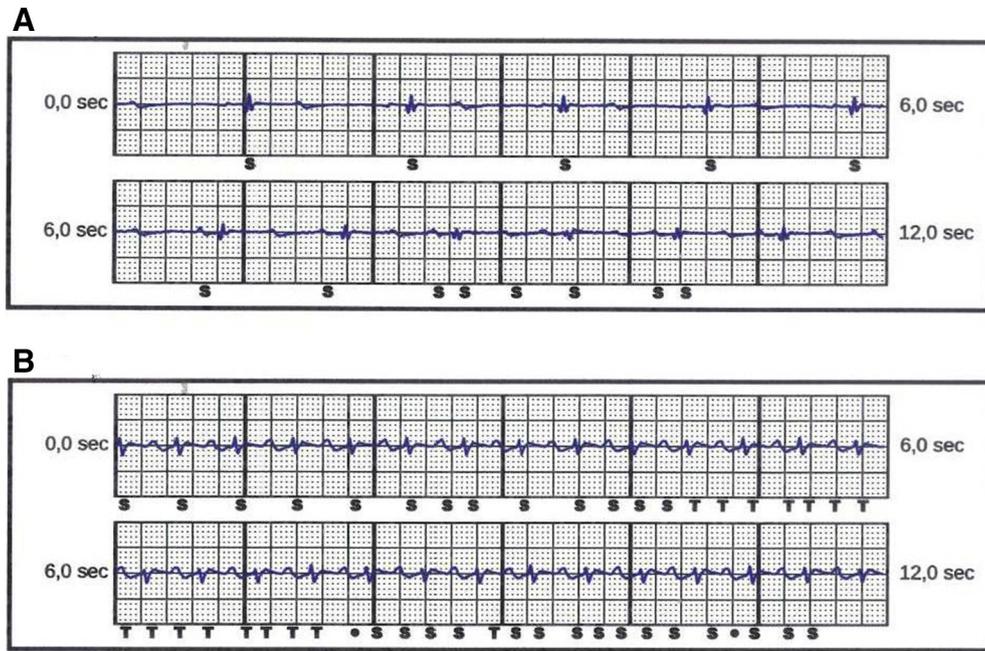
A

B

Fig. 2. S-ICD stored electrogram (alternate sensing vector) showing P-wave oversensing during rest and during effort (secondary sensing vector).

electromagnetic interferences were observed. Inappropriate shock due to SVT tachycardia was solved with catheter ablation and by increasing the threshold of conditioned intervention. One case of oversensing due to myopotential was resolved after activation of the SMART Pass filter by Boston Scientific technical services. At the end of follow-up, 1 patient had the device explanted and opted to TV-ICD because of the impossibility to solve the oversensing of both cardiac (T/P-wave oversensing) and non-cardiac signal (myopotential). No patients had the device removed because of a perceived need for antitachycardia pacing (ATP). In one patient an episode of non-syncopal sustained VT with a cycle
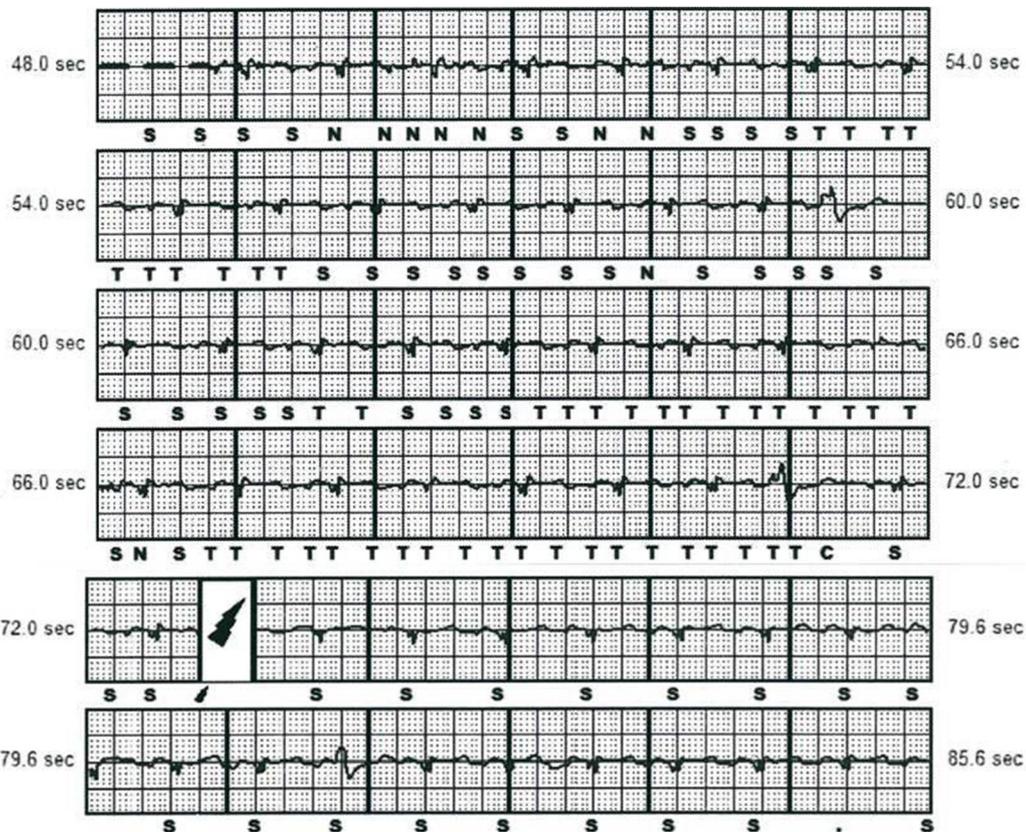

Fig. 3. S-ICD stored electrogram of inappropriate shock due to P/T-wave oversensing during effort in the same patient of Fig. 2.

length below the conditional zone (200 bpm) occurred. After modification of the antiarrhythmic drug therapy and catheter ablation no recurrences of sustained VT were observed. No death or need for transplant occurred in this cohort.

4. Discussion

The main results of the present multicenter study were that: 1) S-ICD system implantation is safe with both the traditional and two incision intermuscular technique; 2) delayed device-related complications were observed in 6.8% of patients during a median follow-up of 1 year with a higher prevalence (27% versus 0%) among those who were implanted with the three incisions technique than the two incisions technique; 3) S-ICD was effective in terminating both induced VF and clinical ventricular arrhythmias demonstrating that S-ICD is a potential valid alternative to TV-ICD in ARVC patients for the prevention of SCD; 4) no patients had the device removed because of a perceived need for ATP or pacing; 5) the main causes of inappropriate shocks were oversensing of cardiac (T/P-wave oversensing) and non-cardiac signals (myopotential, artifacts), highlighting the need for effective strategies to prevent inappropriate shocks, such as appropriate pre-implantation ECG screening and accurate implantation technique.

The current study extends previous preliminary observations [13] on S-ICD in ARVC patients based on a larger multicenter study population and provides a more balanced view on the device benefits versus disadvantages, including inappropriate shocks and device-related complications needing surgical revision.

4.1. Transvenous ICD therapy in ARVC

The available data, coming from observational studies/registries of large populations of ARVC patients, have established the efficacy and safety of ICD therapy [1–3]. These studies consistently document that the TV-ICD successfully interrupts lethal VA and improves long-term outcome of selected high-risk ARVC patients.

It is important to recognize that the survival benefit of TV-ICD treatment is obtained at the expense of significant complications during follow-up, with estimated rates of lead/device related complications and inappropriate ICD therapies of 4.4%/year and 3.7%/year, respectively, especially in young patients [1]. This high rate of lead-related adverse events may be explained by the peculiar ARVC pathobiology which leads to progressive loss of myocardium with fibrofatty replacement, also affecting the site of RV lead implantation [1]. Inappropriate TV-ICD interventions occur in 10% to 25% of patients with ARVC, mostly at young age, and are usually caused by sinus tachycardia or atrial tachyarrhythmia [1–6,14], are painful and may have a profound clinical and psychological impact on patients. Although the use of a dual-chamber detection algorithm offers the potential to reduce the number of inappropriate interventions by improving discrimination of VA from SVT, an additional lead in the atrium predisposes to a higher incidence of early and late postoperative complications.

Experience with ICD therapy consistently highlights the beneficial effect of ATP which is highly effective in terminating VT episodes in ARVC patients [3] and this is particularly relevant considering that a sub-analysis of the MADIT-RIT trial showed that ICD shocks, but not ATP, were associated with adverse prognosis in ICD recipients [15].

4.2. Subcutaneous ICD in ARVC patients

Despite expanding indication of the S-ICD in clinical practice, limited data exists on the safety and efficacy of S-ICD in ARVC patients. Previous studies enrolled only very small number of ARVC patients embedded as part of larger cohorts with different diseases [7,8,10,11,16]. We reported, our clinical experience of S-ICD therapy in a cohort of ARVC patients at risk of SCD. Our study results showed that the S-ICD is effective in terminating both induced VF and clinical VA in ARVC patients.

Comparable to prior experience in unselected cohorts of patients with other cardiomyopathies and channellopathies, all induced tachyarrhythmias during DFT were successfully converted [7,8,16]. Moreover, all episodes of VT/VF during a 1-year follow-up were appropriately detected and successfully treated with no episodes of syncope or SCD. This demonstrates that the S-ICD is an effective long-term option for this high arrhythmic risk population. The longer time to shock for clinical VA (16 ± 4 s) of S-ICD compared to the time of shock during test in the lab (15 ± 5 s) may be explained by the different type of treated arrhythmic events (VT vs VF) and different shock energy, i.e., 80 J and 65 J respectively. Delayed device-related complications were observed in 6.8% of patients during a median follow-up of 1 year with a higher prevalence (27% versus 0%) among those who were implanted with the three incisions technique than the two incisions technique, highlighting the importance of technical issues in S-ICD implantation.

4.3. Pros and cons of transvenous and subcutaneous ICD system in ARVC

The main potential limitation of the S-ICD is the inability to deliver ATP which may be an effective "pain-free" therapy in patients with structural heart diseases including non-ischemic cardiomyopathies [3,15,17].

To this regard, according to the current "International Task Force Recommendations for ARVC treatment" [1], the decision whether to implant a S-ICD in ARVC needs to be patient specific, balancing lead-related complications, typically observed in TV-ICD carriers, with the likelihood of recurrent VT that may be effectively pace-terminated. However, to judge whether this should be regarded as an absolute contraindication to S-ICD in ARVC, other factors should be taken into account. First of all, it has to be noted that ≈2/3 of VTs that were interrupted by ATP in the North American ARVC study [3] were slower than 200 beats per minute, i.e. probably non-life-threatening and potentially self-limited, considering that the majority of ARVC patients have a normal or near-normal LV ejection fraction. It is noteworthy, that multicenter studies on ARVC patients with TV-ICD implanted for either secondary or primary prevention reported a rate of effective ATP intervention in 9.8% and 2.8% of patients, respectively [2,4]. On the other hand, although episodes of slow VT are not life-threatening one should be consider that they may be sustained and potentially amenable to ATP therapy which offer the potential to reduce the shock burden with important implications on the quality of life. In this regard, in our study, six patients received a total of 61 appropriate and successful shock for VA including 80% monomorphic VT (occurring as VT storm in two patients) that could be interrupted by ATP.

The last but not the least, the arrhythmic presentation (i.e. VF vs. VT) of ARVC is age-dependent: while older patients with advanced disease more often experience re-entrant VT around a stable fibro-fatty myocardial scar, in young patients it is common to observe the abrupt onset of VF reflecting the acute electrical instability of early phases of the disease, which progresses through "hot phases" (i.e., recurrent bouts of acute myocyte death with reactive inflammation). An alternative arrhythmogenic mechanism of VF in early disease has been identified at molecular and cellular level as the result of the cross-talk between altered desmosomes and both voltage gated sodium-channel and gap junction proteins. Accordingly, loss of expression of desmosomal proteins may induce electrical ventricular instability by a concomitant sodium channel dysfunction with current reduction, before the disease structural abnormalities occur [1,18]. Young patients are particularly prone to lead-related complications requiring device explant [5,6].

Finally, catheter ablation of VT in ARVC should be considered a potentially effective strategy for eliminating frequent VT. To this regard, in our study one patient experienced an episode of non-syncopal sustained VT with a cycle length below the conditional zone which was effectively treated with catheter ablation. In our study, no patients had the device removed because of a perceived need for ATP, probably

due to the high S-ICD programmed rate cutoff and the discriminative sensing algorithm that may minimize therapy for self-terminated VT episodes.

Taking all these factors into account, S-ICD may be a valuable alternative to TV-ICD, particularly in young patients, and the potential risk of S-ICD shocks on future monomorphic VT must be weighed against the high incidence of long-term lead-related complications requiring surgical revision in ARVC patients which young patients are particularly prone [1,5,6]. Future data from ongoing PRAETORIAN (A PRospective, rAndomizEd Comparison of subcuTaneOus and tRansvenous ImplANtable Cardioverter Defibrillator Therapy) and ATLAS (Avoid Transvenous Leads in Appropriate Subjects) trials, directly comparing safety and efficacy of TV-ICD versus S-ICD, will provide more scientific evidences assisting implanters to make an individual patient-based choice of the type of defibrillator [19,20].

4.4. Inappropriate therapies of the S-ICD in ARVC patients

In comparison to previous S-ICD studies on large and heterogeneous cohorts of patients, we observed a higher incidence of inappropriate shocks (14%) at 1 year of follow-up. This discrepancy can be explained by the different characteristics of the study populations, including younger patients, and to the high prevalence of basal ECG abnormalities typically observed in ARVC patients which is an important predictor of cardiac oversensing leading to inappropriate shocks [10,15,21,22]. Possible strategies that may reduce inappropriate shocks are proper pre-implantation ECG screening [10], device programming (single- vs dual-zone programing) [21], new implantation techniques (Supplementary data, reference 3) and software upgrade including the "SMART Pass" which is a recently introduced filter that has been reported to reduce oversensing [23]. In the assessment of S-ICD eligibility of ARVC patients, some peculiar aspects of the disease should be taken into account. First, patients typically exhibit reduced QRS voltages amplitude and large, negative T-waves and/or right atrial enlargement (peaked P waves) that may cause double-counting and inappropriate interventions. Second, repolarization abnormalities may significantly modify with increasing heart rate so that electrocardiographic screening should be performed both at baseline and during exercise testing [10]. Moreover, ARVC is a progressive disease characterized by R-wave amplitude decline during follow-up [1,2,4] predisposing this population to possible cardiac and/or non-cardiac oversensing and subsequent inappropriate therapy. Consequently, it may be desirable to have at least 2/3 vectors suitable in S-ICD.

4.5. Limitations

The present study has limitations predominantly related to the small study sample, the low event rate, and the relatively limited follow-up period (median 1 year). ARVC is a progressive disease characterized by ECG changes during follow-up, potentially predisposing to cardiac and/or non-cardiac oversensing, with subsequent inappropriate therapy or S-ICD shock failure. Therefore, in ARVC patients it is desiderable to have at least 2 of 3 suitable vectors in the S-ICD screening template to better manage potential device-related oversensing. Further larger prospective studies with predefined comparable TV-ICD cohort are needed to verify our findings and the clinical benefit from device choice.

5. Conclusions

The study shows that the S-ICD provides safe and effective therapy for termination of both induced and spontaneous malignant ventricular tachyarrhythmias with high energy shocks in ARVC patients, but the risk of inappropriate shocks and complications needing surgical revision should be considered.

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ijcard.2019.01.041.

Conflict of interest

None.

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