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Subcutaneous implantable cardioverter defibrillator in cardiomyopathies and channelopathies

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Cardiomyopathies and channelopathies are heterogeneous disorders that increase the risk of sudden cardiac death (SCD). Implantable cardioverter-defibrillator (ICD) therapy is safe and effective for preventing SCD in patients at risk for malignant ventricular arrhythmias. Because of the poor positive predictive value of current risk stratification tools, the majority of patients implanted with an ICD will never receive a life-saving therapy but will be exposed to the risk of complications such as device infection, lead failure and inappropriate therapy. Subcutaneous ICD (S-ICD) now constitutes a valuable alternative to conventional transvenous ICD in patients with cardiomyopathies and channelopathies as it provides protection from SCD while avoiding the risks of intravascular lead infection or failure. This may be particularly advantageous for young patients with a very long life expectancy. On the other hand, S-ICD cannot deliver antitachycardia pacing or antibradycardia pacing. The purpose of this article is to review the available

evidence and the future perspectives of S-ICD therapy in patients with cardiomyopathies or channelopathies.

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Introduction

Sudden cardiac arrest is the most common cause of death in patients with cardiomyopathies and channelopathies.¹ For this reason the implantable cardioverter defibrillator (ICD) has become the gold standard for prevention of sudden cardiac death (SCD).² The advent of the fully subcutaneous ICD (S-ICD) has provided a valuable alternative for patients with cardiomyopathies and channelopathies.³ The S-ICD allows patients who do not need pacing to benefit from arrhythmia protection without the risk of systemic infection and potentially reducing lead failure, which is an important complication of transvenous ICD (T-ICD).⁴ This may be particularly advantageous for young patients with a long life expectancy who might avoid the complications associated with transvenous leads as well as the procedural risks associated with their extraction.⁵

This review article focuses on the available scientific information on safety and efficacy of the use of S-ICD in patients with cardiomyopathies and channelopathies in

order to increase knowledge for the optimal use of this innovative technology in this specific group of patients.

Prevention of sudden cardiac death in cardiomyopathies and channelopathies

Risk stratification in order to identify patients at high risk of malignant ventricular arrhythmias at the time of initial evaluation and during follow-up is strongly recommended by current guidelines.⁷ However, weighting the risks and benefits of ICD therapy in the daily clinical practice remains a difficult task and the decision to implant such a device has to take into account the needs and desires of the individual patient.

Complications of conventional implantable cardioverter defibrillator

Transvenous ICDs are implanted worldwide in an increasing number of patients.² Unfortunately, complications or failure of a conventional ICD lead may result in adverse clinical events, including failure to pace and

defibrillate, inappropriate shocks, pocket infection, endocarditis, cardiac perforation and even death.⁵ Recent studies have reported an increased risk of re-intervention after implantation of high-voltage ICD leads compared with right ventricular pacing leads.⁶ This can be attributed to the more complex structure, larger caliber, and increased rigidity of high-voltage leads which, in addition, require to satisfy more stringent lead parameters at the time of implant and during follow-up.

The risk of complications associated with T-ICD implantation ranges between 3 and 8%. Kirkfeldt *et al.*⁸ reported that 9.5% of patients who received an ICD experienced at least one complication, whereas 0.6% of patients had multiple complications. Lead-related re-intervention was the single most common complication (2.4%). System upgrades or lead revisions are associated with a higher rate of complications than first implants, primarily because of the higher risk of infections that are infrequent but carry significant morbidity and mortality. Tarakji *et al.*⁹ aimed to assess risk factors for 1-year mortality among patients with ICD infection and to evaluate if the type of infection and the presence of vegetation affect survival. The authors reported a 20% mortality at 1-year of follow-up; ‘endovascular infection’ was associated with significantly higher 1-year mortality compared with ‘pocket infection’ independently of the presence of vegetations. Olde Nordkamp *et al.*¹⁰ performed a systematic review and meta-analysis of inappropriate shocks and/or other ICD-related complication rates in patients with inherited arrhythmia syndromes. In this population, inappropriate shocks occurred in 20% of patients with an annual rate of 4.7% per year. Moreover, 22% experienced ICD-related complications (4.4% per year) and ICD-related mortality was 0.5%. Lead malfunction was the primary complication in this study occurring in 10% of patients.

Inappropriate shocks and device-related complications may be more prevalent in young patients because of an active lifestyle and because of a longer period of exposition to the risks of ICD therapy. Migliore *et al.*¹¹ specifically evaluated the risk–benefit ratio of ICD in 96 young patients with cardiomyopathies and channelopathies who were 18–35 years old at the time of ICD implantation. During a mean follow-up of 72.6 ± 53.3 months, 20 patients (21%) experienced at least one appropriate ICD intervention (4%/year), 9 (9%) at least one inappropriate shock and 20 (21%) suffered from device-related complications requiring system revision. In particular, the most common complication, which occurred in 9 patients, was lead failure/fracture requiring lead extraction.¹¹

Potential advantages of subcutaneous implantable cardioverter defibrillator

The S-ICD might overcome the limitations of conventional ICDs, as the absence of transvenous leads might reduce implantation and lead-related complications in patients without an indication for brady and

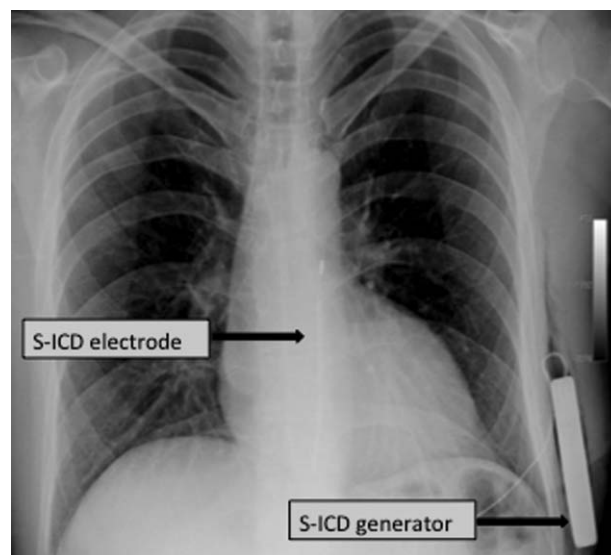
antitachycardia pacing (ATP). The EFFORTLESS registry¹² has provided data to evaluate the safety and efficacy of the S-ICD in a large population.

Patients were followed-up for 3.1 ± 1.5 years and 82 completed a 5-year follow-up period. The S-ICD system and procedure complication rate were 4.1% at 30 days and 8.4% at 360 days. The 1-year complication rate trended toward improvement from the first (11.3%) to the last (7.4%) quartile of enrollment. Few device extractions occurred because of need for antitachycardia ($n=5$), biventricular ($n=4$) or bradycardia pacing ($n=1$). Inappropriate shocks occurred in 8.1% of patients at 1 year and 11.7% after 3.1 years. The 1-year and 5-year rates of appropriate shock were 5.8 and 13.5%, respectively. Conversion success for discrete spontaneous episodes was 97.4% overall. Overall, these data suggest that S-ICD is associated with favorable outcomes.

Subcutaneous implantable cardioverter defibrillator: device and implantation technique

The S-ICD consists of a pulse generator and a single lead with a shock coil. The pulse generator is implanted in the left lateral position. A single lead for sensing is usually positioned 1–2 cm to the left of and parallel to the sternum, with the distal tip near the manubriosternal junction.⁴ The lead consists of sensing electrodes at the subxiphoid (proximal) and manubriosternal junction (distal) positions, separated by an 8-cm shocking coil (Fig. 1). The conventional implantation technique requires three incisions to implant the electrode and

Fig. 1



Chest X-ray in a patient with subcutaneous implantable cardioverter-defibrillator. The generator is positioned on the lateral side of the chest wall. The defibrillator lead is tunneled under the skin, and located along the chest wall in the left parasternal position.

the device that is positioned in a subcutaneous pocket at the level of the left axillary line.⁴

Recently, a two-incision technique, eliminating the superior parasternal incision, has been developed.¹³ This less-invasive and simplified technique is safe and may help reducing complications in S-ICD patients. An intermuscular pocket has also been proposed as an alternative to the standard subcutaneous pocket. This technique consists of creating a pocket for the device between the anterior surface of the serratus anterior muscle and the posterior surface of the latissimus dorsi muscle using anatomical landmarks.^{14–16} The intermuscular technique provides a wider pocket in a more posterior left axillary region, an extra layering, and a virtual space between device and chest, resulting in potential reduction of pocket-related complications, especially skin erosions and infections.^{16,17} As the whole system, including the electrode, is located outside the vascular system, S-ICD is by far less prone to infection, and the lead is less prone to fracture. Due to the lack of an electrode, however, the device is unable to deliver bradycardia pacing, ATP, or cardiac resynchronization therapy. On the other hand, the fact that the device is not in contact with the heart allows patients with S-ICD to safely undergo magnetic resonance imaging.⁵

Indications to subcutaneous implantable cardioverter defibrillator in patients with cardiomyopathies and channelopathies

The S-ICD is approved for the prevention of SCD among candidates for a conventional T-ICD but who do not have

an indication for antibradycardia pacing or cardiac resynchronization therapy, recurrent ventricular tachycardia (VT) treatable with ATP, or preexisting unipolar pacemaker leads. Although patients enrolled in early S-ICD prospective registries were relatively young, with less advanced heart disease, and sometimes with ‘niche’ indications (i.e. high risk of infections, congenital heart disease, poor vascular access), contemporary S-ICD patients are more severely diseased and have more comorbidities.¹⁸ Notwithstanding, these young patients with cardiomyopathies and primary electrical heart diseases are ideal candidates for ICD (Table 1). Indeed, cumulative lead-related and device-related complications of the T-ICD are well known and have been reported to be as high as 2–4% per year in patients with cardiomyopathies.^{10,11} The S-ICD is safe and effective in cardiomyopathy patients, and because of the negligible risk of systemic infection and increased durability of the lead system, it is an attractive option for most cardiomyopathy patients without a pacing indication.¹⁹

At present, S-ICDs is recommended only in a specific subset of patients. The 2015 guidelines for the prevention of SCD of the European Society of Cardiology have established a Class IIa-level C recommendation for the use of S-ICD as an alternative to transvenous ICDs when pacing therapy for bradycardia, cardiac resynchronization or ATP is not needed.²⁰ Also, S-ICD has a Class IIb-level C recommendation as an alternative to T-ICD in case of difficult venous access, young patients, and after the removal of a previous T-ICD system for infection.

Table 1 Number of patients with cardiomyopathies or channelopathies with subcutaneous implantable cardioverter defibrillator included on S-ICD registries

Author group	Reference	Number of patients
Hypertrophic cardiomyopathy		
Arias AM, et al.	<i>Rev Esp Cardiol</i> 2017 Dec 5. pii: S1885-5857	9
Özkartal T, et al.	<i>Swiss Med Wkly</i> 2017;147:w14518	9
Maurizi N, et al.	<i>Int J Cardiol</i> 2017; 231:115–119	50
Lambiase PD	<i>Heart Rhythm</i> 2016; 13:1066–1074	99
Frommeyer G, et al.	<i>Clin Res Cardiol</i> 2016; 105:89–93	18
Francia P, et al.	<i>J Cardiovasc Electrophysiol</i> 2015; 26:893–899	3
Galvao P, et al.	<i>Rev Port Cardiol</i> 2014; 33:511–517	4
Kobe J, et al.	<i>Heart Rhythm</i> 2013; 10:29–36	10
Dilated cardiomyopathy		
Kobe J, et al.	<i>Heart Rhythm</i> 2013; 10: 29–36	69
Brouwer TF, et al.	<i>J Am Coll Cardiol</i> 2016; 19: 2047–2055	30
Arrhythmogenic cardiomyopathy		
Migliore F, et al.	<i>JACC Clinical Electrophysiol</i> 2017; 3:785–786	1
Boersma I, et al.	<i>JACC</i> 2017; 70:830.841 [12]	32
Brugada syndrome		
Kamakura T, et al.	<i>Heart Vessels</i> 2017; 32:1277–1283	6
Ishibashi K, et al.	<i>J Arrhythm</i> 2017; 33:156–158	1
Morani G, et al.	<i>Pacing Clin Electrophysiol</i> 2017; 40:1180–1183	1
Sideris S, et al.	<i>Hellenic J Cardiol</i> 2017; 58:4–16	2
Hai JJ, et al.	<i>Europace</i> 2015; 17 Suppl 2:ii63-8	6
Yap SC, et al.	<i>Heart Rhythm Case Rep</i> 2015;1:156–158	1
Galvão P, et al.	<i>Rev Port Cardiol</i> 2014; 33:511–517	4
Köbe J, et al.	<i>Heart Rhythm</i> 2013;10:29–36	6
Olde Nordkamp LR, et al.	<i>J Am Coll Cardiol</i> 2012; 60:1933–1999	9
Jarman JW, et al.	<i>Eur Heart J</i> 2012; 33:1351–1359	3
De Maria E, et al.	<i>J Interv Card Electrophysiol</i> 2012;34:105–113	1
Dabiri Abkenari L, et al.	<i>Clin Res Cardiol</i> 2011; 100:737–744	2
Conte G, et al.	<i>Europace</i> 2018; 20:1188–1193	61

The 2017 American guidelines on prevention of SCD⁷ state that S-ICD is recommended in patients who meet criteria for an ICD who have inadequate vascular access or are at high risk for infection, and in whom pacing for bradycardia or VT termination or as part of cardiac resynchronization therapy is neither needed nor anticipated (Class I). Moreover, S-ICD is an option in patients who meet indication for an ICD if pacing for bradycardia or VT termination or as part of cardiac resynchronization therapy is neither needed nor anticipated (class II). Finally, S-ICD is contraindicated if bradycardia pacing, cardiac resynchronization therapy or ATP is required (class III).

Subcutaneous implantable cardioverter defibrillator in hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy (HCM) is a genetic sarcomeric disorder associated with myocyte disarray and scar deposition, which predisposes to malignant ventricular arrhythmias.¹ The disease is a leading cause of SCD in young adults²¹ and high-risk HCM patients benefit from ICD therapy, although they may suffer long-term device-related complications.²¹ The S-ICD is a promising option for HCM patients, who are usually young and rarely require pacing.^{22–24}

The S-ICD performance in HCM patients has been evaluated extensively.^{23–28} It proved effective in the detection and cardioversion of induced VT/ventricular fibrillation (VF) with equivalent frequency of inappropriate shocks between HCM and non-HCM patients. Specifically, reported rates of induced VT/VF conversion were 95.5–98% at 65 J and 98–100% at 80 J.^{26–28} Of note, defibrillation failures at 65 J were mostly attributed to severe obesity and its consequences on defibrillation threshold rather than on the HCM phenotype.^{26,27} In fact, patients with left ventricular maximal wall thickness at least 30 mm or a marked increase of left ventricular mass showed a similar rate of effective 65 J defibrillation of induced VT/VF to those with milder phenotype.^{26–29} New implantation techniques, including intermuscular placement, have been developed in order to avoid this problem.^{14–16}

Reduction of inappropriate shocks remains a significant challenge. Up to 12.5% of patients with HCM experienced inappropriate shocks during a median follow-up of 2 years, mainly due to T-wave oversensing.²⁸ However, recent improvements in the S-ICD discrimination algorithm as well as accurate patient selection are offering a partial mitigation to such an undesired event. Pocket infection occurred in up to 5% of patients, but they were all managed with standard antibiotic treatment.²⁸ Lead fracture was extremely rare as well as lead displacement.²⁸ In conclusion, in HCM patients who do not need pacing, S-ICD is a safe and effective prophylactic therapy for SCD prevention^{23–28} (Fig. 2).

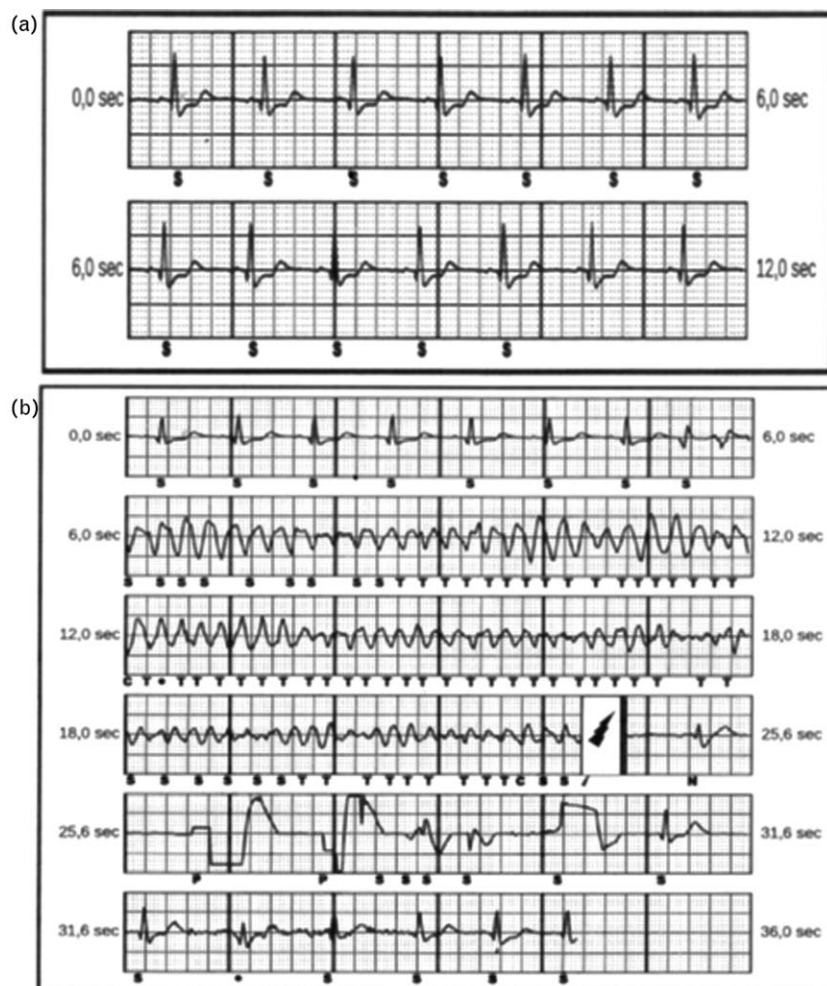
The risk of inappropriate shocks should be mitigated by accurate preimplant evaluation and dual zone programming. Specifically, because HCM patients often show markedly abnormal ECG, a careful preimplantation evaluation is required to avoid inappropriate sensing by the S-ICD, which uses a morphology-based rhythm discrimination algorithm.^{23–25} Recent studies examining ECG vector screening at rest have demonstrated that 7–16% of HCM patients fail preimplant screening, mainly because of massive hypertrophy, broad QRS, prior myectomy procedure, and T-wave inversions in more than two leads on standard 12-lead electrocardiogram.^{24,25} Moreover, in HCM, the S-ICD system may be challenged in the differentiation between supraventricular tachycardia and VT because of the relative R:T wave amplitude variation during tachycardia, potential ST-segment elevation, or development of rate-dependent aberrancy. The latter can be mitigated by storing a template of previous aberrancy (i.e. during exercise), altering the sensing vector or modifying the cut-off rate, but cannot be avoided if the changes are transient or unpredictable.²⁸ These dynamic QRS-T wave changes emphasize the importance of exercise testing these patients prior to implant to optimize template vector selection.^{23–28} Usually two (of the three available) vectors should pass the screening process in order to provide alternative vectors for sensing. However, a single passing vector that remains eligible both at rest and during exercise is acceptable when avoiding endovascular leads is warranted. Also, right parasternal chest electrodes should be assessed in the case of screening failure using conventionally positioned electrodes.²⁴

Subcutaneous implantable cardioverter defibrillator in dilated cardiomyopathy

In the context of dilated cardiomyopathy (DCM), ICD is recommended in patients with haemodynamically not tolerated VT/VF or in patients with symptomatic heart failure and an ejection fraction less than 35% despite at least 3 months of optimal medical therapy.⁷ Most DCM patients with ICD indication are suitable S-ICD recipients,³ but, in clinical practice, only a few centers propose S-ICD as a first-choice therapy.²⁹

The absence of ATP functionality is an important barrier to the widespread adoption of the S-ICD in patients with DCM. The device can obviously not be used in patients requiring bradycardia or resynchronization pacing. However, the need for pacing in DCM patients in historical ICD trials³⁰ and in recent S-ICD studies^{12,31} has been very low when patients are properly selected and there is no indication in the current guidelines for preventive pacemakers when conduction system disease is not expected. Similarly, the role of ATP in the era of therapy reduction programming is not clear. In the MADIT-RIT trial,³² very few patients received ATP in the high-rate and delayed-therapy arms. These findings raise the

Fig. 2



A 31-year-old man with hypertrophic cardiomyopathy (left ventricular maximal wall thickness 38 mm) who underwent subcutaneous implantable cardioverter defibrillator implantation for primary prevention (a). Two years after follow-up, the patient received appropriate ICD intervention on ventricular fibrillation (b). ICD, implantable cardioverter defibrillator.

question of whether the potential benefit of ATP is outweighed by the risk of transvenous lead complications.

Although no study has prospectively investigated the benefit of S-ICD in specific etiological subgroups of DCM, patients with this cardiomyopathy are well represented in the trials. Particularly, several clinical studies have demonstrated the safety and effectiveness of the S-ICD in these patients.^{12,31} In a propensity-matched cohort (20% of patients with DCM) of S-ICD, the authors demonstrated that the S-ICD has a significant benefit over ICDs with respect to lead-related complications.³³ More recently, the EFFORTLESS S-ICD Study Mid-term Results¹² showed that the rate of complications directly caused by the S-ICD system as well as the rate of all S-ICD system and procedure-related complications were low. Specifically, no significant higher risk of

complications was observed in DCM patients. Moreover, the EFFORTLESS/IDE pooled cohort confirmed that the favorable outcomes achieved with the S-ICD continue up to 3 years postimplantation.¹² In the coming years, the UNTOUCHED (Understanding Outcomes with the S-ICD in Primary Prevention Patients with Low Ejection Fraction Study), a prospective registry of primary prevention patients with reduced left ventricular ejection fraction³⁴ will provide valuable data on S-ICD in this cohort of patients.

Subcutaneous implantable cardioverter defibrillator in arrhythmogenic cardiomyopathy

According to the 'International Task Force Recommendations for arrhythmogenic right ventricular cardiomyopathy (ARVC) treatment,³⁵ the decision whether to implant a S-ICD in ARVC needs to be patient-specific,

balancing lead-related complications with the likelihood of recurrent VT that may be effectively pace-terminated. Antitachycardia pacing appears very effective in terminating VT in ARVC patients. Among 108 patients with an ICD (mean age 40 years, 48% secondary prevention) enrolled in the North American ARVC registry,³⁶ 48 experienced a total of 489 episodes of monomorphic VT during follow-up: of those, 450 were treated with ATP that terminated the arrhythmia in 92% of cases. The efficacy of ATP was consistent throughout different tachycardia cycle length suggesting that ATP can interrupt even fast VT. These data suggest that the inability of the S-ICD to deliver ATP is an important limitation in the setting of ARVC, especially considering that a sub-analysis of the MADIT trial showed that ICD shocks, but not ATP, were associated with adverse prognosis in ICD recipients.³² 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 $\approx 2/3$ of VTs that were interrupted by ATP in the North American study were slower than 200 beats per minute, that is, probably nonlife-threatening considering that the majority of ARVC patients have a normal or near-normal left-ventricular ejection fraction.³⁶ Moreover, the arrhythmic presentation (i.e. VF vs. VT) of ARVC is age-dependent: although 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 acute electrical instability of early disease, which progresses through so-called 'hot phases' (recurrent bouts of acute myocyte death with reactive inflammation).^{37,38} Finally, young patients are particularly prone to lead-related complications requiring device explant.¹¹ Taking all these factors into account, S-ICD may be a valuable alternative to T-ICD in young ARVC patients without a history of sustained monomorphic VT.

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 voltages of QRS complexes 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 (Fig. 3).^{39–41}

Subcutaneous implantable cardioverter defibrillator in Brugada syndrome

Brugada syndrome (BrS) is an inherited arrhythmogenic disorder that exposes young men to polymorphic VT or VF and SCD.^{42–48} To date, the literature focusing on S-ICD in BrS is limited. The main limitation is represented by lack of pacing capability. Patients with BrS are

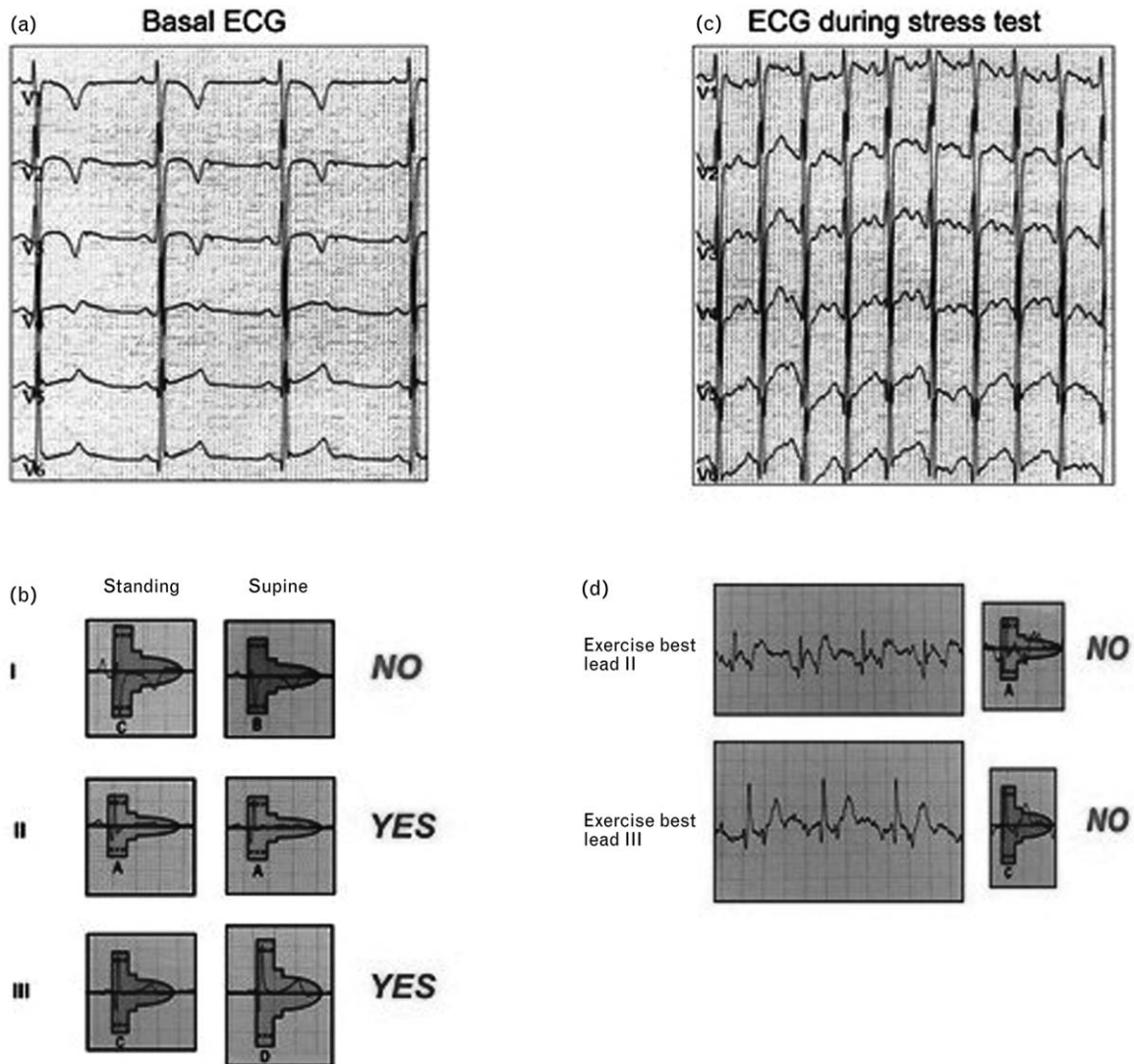
potentially at risk of progressive conduction disorders because of genetically defective sodium channels similar to the Lenègre disease phenotype.⁴⁷ However, recent reports underlined that advanced atrioventricular block only develops at advanced age, whereas ATP is not considered essential, as VF and polymorphic VT are the most common arrhythmias occurring in BrS⁴⁴ while monomorphic VT is rare.⁴⁹

Similarly to other cardiomyopathies, reduction of inappropriate shocks is pivotal in patients with BrS. In this regard, it is important to accurately assess the sensing vectors at preimplant screening, as dynamic ST-segment elevation in right precordial leads is observed in various daily life situations such as during sleep, following a meal, and after exercise⁵⁰ (Fig. 4). Screening tests conducted only at rest may be inadequate for the prediction of the T-wave oversensing in S-ICD candidates with BrS, and drug challenge test may be useful in appropriately qualifying BrS patients for S-ICD.⁵¹ To increase the precision of arrhythmia detection by S-ICD, an updated software applies high-pass filter that diminishes the amplitude of lower frequency signals such as T waves, whereas higher frequency signals such as R waves, polymorphic VT and VF amplitudes remain largely unchanged.⁵² Finally, supraventricular tachycardias, including atrial fibrillation, are synergistically discriminated by two algorithms named ventricular scatter analysis, which monitors atrial fibrillation vs. nonatrial fibrillation episodes based on R-R intervals, and heart rate density index algorithm, which controls heart rate distribution.⁵³

Potential limitation of the subcutaneous implantable cardioverter defibrillator

Selection of eligible patients is crucial for S-ICD and is based on a template ECG morphology screening, which evaluates QRS complex and T-wave morphology in order to detect QRS/T-wave oversensing.⁵⁴ Inability to find an ECG lead suitable for the screening template implies a potential high risk of inappropriate detections by S-ICD owing to oversensing of cardiac signals, mostly T wave, and advises against S-ICD implant.⁵⁴ Predictors of ECG-screening failure, especially in younger patients,⁵⁵ are longer QRS duration, longer QTc interval, and a lower R to T amplitudes (R:T ratio),^{56–58} which are depolarization/repolarization ECG abnormalities typically observed in patients with cardiomyopathies and channelopathies.^{21,37,44} These peculiarities can lead to a higher rate of ECG-screening failure in this subgroup of patients, decreasing the number of those who could benefit from the attractive therapeutic option of S-ICD implantation. In this regard, Olde Nordkamp *et al.*^{56,57} showed that HCM was an independent predictor of screening failure, but this was not reported in other studies.^{23,24,55} Conte *et al.*⁵⁹ addressed rate and specific aspects of S-ICD eligibility in patients with inherited primary arrhythmia syndromes. They found that 13% of patients potentially

Fig. 3



Representative example of a young patient affected by arrhythmogenic cardiomyopathy with right precordial negative T waves on basal ECG (a) that normalize during exercise (c). Morphology analysis of QRS and T wave showed that although two of three sense vectors were appropriate at baseline (b), they became inappropriate during exercise (d), precluding a subcutaneous defibrillator implantation. Reproduced from Migliore *et al.*⁴⁰

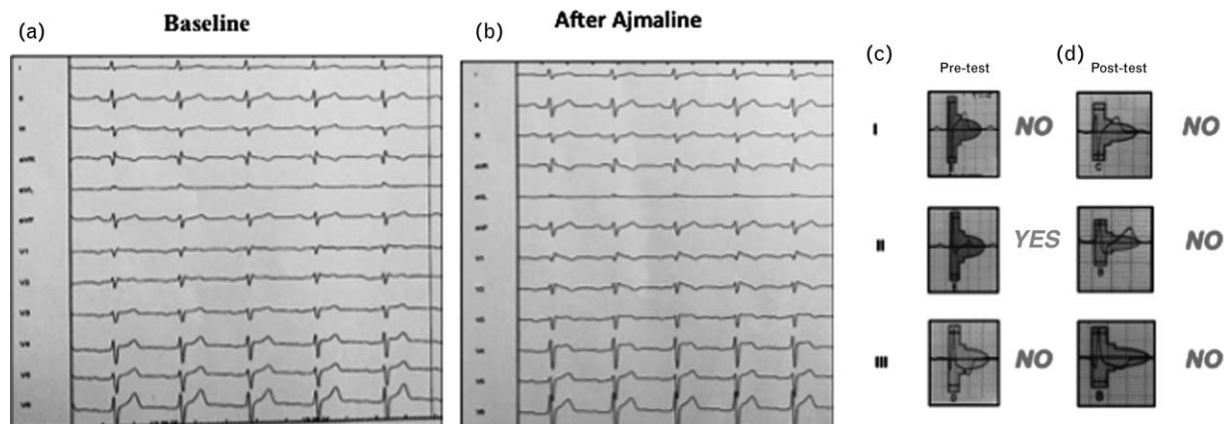
candidates for S-ICD implantation, failed the screening morphology analysis. In particular, patients with BrS showed the highest rate of screening failure and the lowest number of appropriate S-ICD vectors compared with patients with other channelopathies.⁵⁹ However, in another study, Rudic *et al.*⁶⁰ did not observe any screening failure in a cohort of patients with inherited primary arrhythmia syndrome. The different sample size and the ECG screening performed or not after ajmaline challenge may explain the discrepancy in eligibility rates.

In the assessment of S-ICD eligibility of patients with cardiomyopathy and channelopathies, some peculiar aspects of the diseases should be taken into account. Important dynamic changes in surface ECG may occur

with increasing heart rate, so that ECG screening should be considered both at baseline and during exercise stress testing.^{24,40,61} Moreover, cardiomyopathies are progressive diseases predisposing this population to worsening of sensing reliability leading to possible cardiac and/or non-cardiac oversensing and subsequent inappropriate therapy, increasing defibrillator energy or pacing requirement.^{37,62} Consequently, it may be desirable to have at least two of three vectors suitable in S-ICD screening template in order to facilitate future management of potential device-related oversensing during follow-up.

Possible strategies that may increase S-ICD eligibility and reduce inappropriate shocks are proper

Fig. 4



A representative example of preimplant screening test in a patient with Brugada syndrome. Normal QRS, ST-segment and T wave on basal ECG (a) and 'coved-type' ST-segment elevation (type 1 Brugada ECG) after ajmaline test (b). Morphology analysis of QRS and T wave at baseline showed that only one (II) of the three sense vectors was appropriate because of the high amplitude of the T wave of the sense vectors I and III (c). Morphology analysis repeated after ajmaline administration showed that also the previously appropriate sense vector was no longer acceptable because of the occurrence of the 'coved-type' ST-T waveform (d). Owing to the lack of consistency of appropriate sense vectors both at baseline and after ajmaline challenge, the patient received a transvenous single-chamber ICD. ICD, implantable cardioverter defibrillator.

preimplantation ECG screening,^{24,40,61} device programming (single-zone vs. dual-zone programming),¹² new implantation techniques,^{13–16} and software upgrading including the 'SMART Pass', which is a recently introduced filter that has been reported to reduce over sensing.⁶³

The major limitation for S-ICD comes from the lack of permanent pacing capability and inability to deliver ATP, which may be an effective 'pain-free' therapy in patients with structural heart diseases including nonischemic cardiomyopathies.^{7,32,36} Patients who undergo ICD implantation because of clinical episodes of monomorphic VT should receive a device able to deliver ATP in order to avoid shocks; hence, for these patients S-ICD should not be the first choice. Notably, in the long-term follow-up of the entire EFFORTLESS registry cohort, only 0.5% of patients had the device removed for conceived need for ATP.¹² These data suggest that in patients without a history of sustained VT implantation of a S-ICD should not be excluded based only on the inability of the device to deliver ATP. Instead, S-ICD may be a valuable alternative to T-ICD in patients with cardiomyopathies and channelopathies and the potential risk of S-ICD shocks rather than ATP on future monomorphic VT must be weighed against the high incidence of long-term lead-related complications requiring surgical revision.

Another potential limitation of the first S-ICD system was the battery longevity of about 5 years, which is shorter than T-ICDs, meaning more replacements and potential complications.⁶⁴ However, the new S-ICD generation (EMBLEM MRI model A219, Boston Scientific, USA) is an advanced MRI conditional technology with a prolonged battery life of up to 7 years

(<http://www.bostonscientific.com/en-US/products/defibrillators/emblem-s-icd-system/device-overview/specifications.html>). Finally, the higher cost of the S-ICD compared with T-ICD could be a limitation in clinical practice. However, despite the existing significant difference in unit cost of the S-ICD, overall S-ICD costs may be mitigated vs. T-ICDs over a longer follow-up period.⁶⁵

Conclusion

The S-ICD represents an important advance in clinical electrophysiology, offering life-saving defibrillation while limiting lead-related complications. Technical refinements including size reduction, increased battery longevity, and improved T-wave detection will enhance its clinical utility. The availability of a defibrillator system associated with fewer complications than traditional T-ICD may expand the proportion of patients at risk of SCD to whom ICD therapy confers a net clinical benefit. Specifically, it may lower the threshold for ICD implantation in patients with cardiomyopathies who have a long quality-adjusted life expectancy but are also particularly prone to the risk of complications of traditional ICD (and dangerous transvenous lead-extraction procedures).

Conflicts of interest

There are no conflicts of interest.

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