

Clinical Experience With the Subcutaneous Implantable Cardioverter–Defibrillator in Adults With Congenital Heart Disease

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Background—Sudden cardiac death is a major contributor to mortality for adults with congenital heart disease. The subcutaneous implantable cardioverter–defibrillator (ICD) has emerged as a novel tool for prevention of sudden cardiac death, but clinical performance data for adults with congenital heart disease are limited.

Methods and Results—A retrospective study involving 7 centers over a 5-year period beginning in 2011 was performed. Twenty-one patients (median 33.9 years) were identified. The most common diagnosis was single ventricle physiology (52%), 9 palliated by Fontan operation and 2 by aortopulmonary shunts: *d*-transposition of the great arteries after Mustard/Senning (n=2), tetralogy of Fallot (n=2), aortic valve disease (n=2), and other biventricular surgery (n=4). A prior cardiac device had been implanted in 7 (33%). The ICD indication was primary prevention in 67% and secondary in 33% patients. The most common reason for subcutaneous ICD placement was limited transvenous access for ventricular lead placement (n=10) followed by intracardiac right-to-left shunt (n=5). Ventricular arrhythmia was induced in 17 (81%) and was converted with ≤80 Joules in all. There was one implant complication related to infection, not requiring device removal. Over a median follow-up of 14 months, 4 patients (21%) received inappropriate and 1 (5%) patient received appropriate shocks. There was one arrhythmic death related to asystole in a single ventricle patient.

Conclusions—Subcutaneous ICD implantation is feasible for adults with congenital heart disease patients. Most candidates have single ventricle heart disease and limited transvenous options for ICD placement. Despite variable anatomy, this study demonstrates successful conversion of induced ventricular arrhythmia and reasonable rhythm discrimination during follow-up. (*Circ Arrhythm Electrophysiol.* 2016;9:e004338. DOI: 10.1161/CIRCEP.116.004338.)

Key Words: congenital heart disease ■ Fontan procedure ■ subcutaneous implantable cardioverter–defibrillator ■ sudden cardiac death ■ ventricular arrhythmia

The subcutaneous implantable cardioverter–defibrillator (S-ICD) has emerged as a novel tool for the treatment of life-threatening ventricular arrhythmia in patients deemed to be at an elevated risk for sudden cardiac death (SCD). This technology has been shown to perform with comparable safety and efficacy to transvenous ICD systems in large-scale postmarket clinical studies.¹ Many adults with congenital heart disease (ACHD) are at increased risk for SCD based on a multitude of conventional and lesion-specific risk factors,^{2,3} prompting consensus-based recommendations regarding primary and secondary prevention indications.⁴ However, placement of traditional transvenous ICD systems in the ACHD population may be problematic because of various anatomic constraints, the presence of residual intracardiac shunting, lack of vascular access, and an elevated rate of lead malfunction.^{5–7}

The S-ICD has been advocated as a means of circumventing many of these limitations and, therefore, may be ideally suited to the ACHD population.^{8–11}

Although there are emerging data regarding eligibility for the S-ICD among patients with ACHD,^{9,12} there is little clinical experience with this technology. Knowledge of the forms of congenital heart disease most likely to benefit, reasons for device placement, and clinical performance are needed.⁹ The present study from the AARCC (Alliance for Adult Research in Congenital Cardiology) examines the preliminary clinical experience with the S-ICD in the ACHD population. It is hypothesized that the S-ICD would be most used for forms of congenital heart disease lacking transvenous access to the heart and that it would be associated with both reliable conversion of induced ventricular arrhythmia at implant and reasonable rhythm discrimination during follow-up.

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WHAT IS KNOWN

- The implantable cardioverter defibrillator is associated with a reduction in mortality for patients at elevated risk for sudden cardiac death (SCD).
- SCD risk for adults with congenital heart disease (ACHD) varies by congenital heart lesion and associated clinical risk factors.

WHAT THE STUDY ADDS

- In this series of at-risk ACHD patients, the most common reasons for choosing the subcutaneous implantable cardioverter-defibrillator (S-ICD) included single ventricle physiology or known intracardiac right-to-left shunt.
- The S-ICD was effective for acute detection and conversion of induced ventricular arrhythmias, and clinical sensing failure and inappropriate shocks during follow-up were comparable to those of the general population.
- Special considerations for S-ICD placement in the ACHD population may include abnormalities in ventricular repolarization, higher than usual prevalence of supraventricular tachycardia, and an increased potential for interdevice interactions.

Methods

Institutional Review Committee approval was obtained at the participating centers, and all patients provided informed consent for S-ICD placement. Patient data were deidentified and electronically transferred to the coordinating center through a secure, web-based server (REDCap). Patients were eligible for inclusion if ≥ 18 years of age at the time of implant and had a history congenital heart disease with or without surgical repair. Categories of data collection included the following: (1) patient demographics, (2) preimplant clinical characteristics (congenital diagnosis and details regarding surgical repair/palliation; results of the most recent catheterization and noninvasive imaging; a copy of the preimplant 12-lead ECG; results of S-ICD eligibility screening both at baseline in supine and standing positions, as well as with exercise testing at the discretion of the treating physician; existing cardiac implantable electronic device [CIED] details; antiarrhythmic drug therapy; ICD indication; and motivation for use of the S-ICD), and (3) implant characteristics (implant techniques, results of defibrillation testing, initial S-ICD programming, a copy of the postprocedural chest x ray, procedural complications, and postprocedural length of stay). Fifteen patients received an SQ-RX model 1010 pulse generator (Boston-Scientific, Marlborough, MA) beginning on January 12, 2011, during the early period of the study and a further 6 received the EMBLEM model A209 beginning May 20, 2015. Acute complications were defined as those occurring within 30 days of implant or before discharge from the hospital. Outcomes during clinical follow-up and device interrogation results for all therapies were submitted to the coordinating center and were independently adjudicated by 2 separate electrophysiologists. Therapies were classified as appropriate if delivered for ventricular tachyarrhythmia; otherwise, they were considered inappropriate.

Data are presented as mean \pm standard deviation or median (interquartile range) for continuous variables as appropriate and as frequencies and percentages for dichotomous variables. The study is descriptive, with no inferential statistics performed. Descriptive analyses were performed with JMP software (SAS Inc, Cary, NC).

Table 1. ACHD Population Characteristics

Characteristic	N=21
Age, y	34 (24–41)
Body mass index, kg/m ²	24.1 (20.8–26.7)
Congenital/surgical anatomy	
Single ventricle*	11
DTGA, atrial switch	2
TOF	2
Aortic valve disease	2
CCTGA, LV-to-PA conduit	1
Ebstein anomaly	1
Hypoplastic RV	1
Congenital PS	1
Male, n (%)	13 (62)
Systemic EF, %	
LV	41 (35–63)
RV	33 (20–45)
AV valve regurgitation, n (%)†	
None to mild	15 (71)
Moderate	4 (19)
Severe	1 (5)
Saturation $\leq 90\%$, n (%)	6 (29)
QRS duration	113 (99–136)
Prior CIED, n (%)	
Transvenous	1 (5)
Epicardial	6 (29)
Antiarrhythmic drug	
Amiodarone	5
Sotalol	2
Beta-blocker	11
Digoxin	4
Flecainide	1
Mexilitine	1

Numbers in parentheses represent percent (%) or interquartile range (IQR). ACHD indicates adults with congenital heart disease; CCTGA, congenitally corrected transposition of the great arteries; CIED, cardiac implantable electronic device; DTGA, *d*-transposition of the great arteries; EF, ejection fraction; ICD, implantable cardioverter-defibrillator; LV, left ventricle; PA, pulmonary artery; PS, pulmonic stenosis; RV, right ventricle; and TOF, tetralogy of Fallot.

*Palliated by Fontan operation in 9 patients (5 lateral tunnel, 2 extracardiac conduit, 1 intra-atrial conduit, 1 atriopulmonary connection) or aortopulmonary shunt in 2 patients.

†Data unavailable for 1 patient.

Results

Baseline Demographics

A total of 21 patients (62% male; median age 34 years, interquartile range 24–41 years) met eligibility criteria and were included in the study. Baseline characteristics are summarized

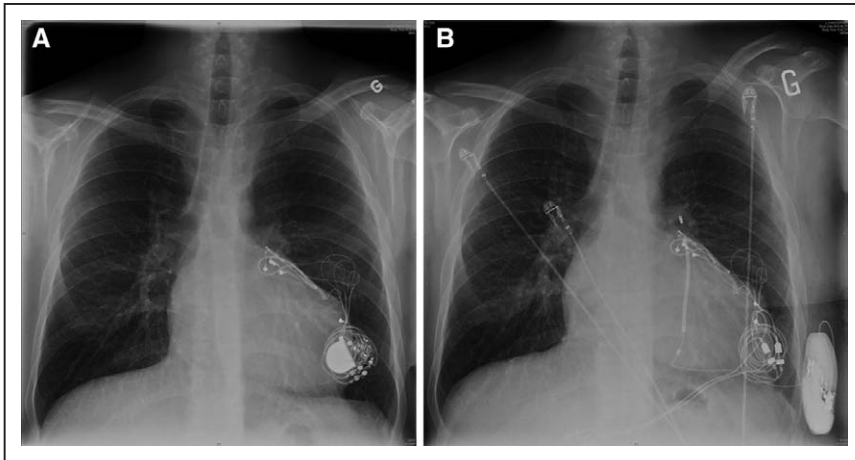


Figure 1. Preimplant strategy for avoidance of interdevice interaction with the subcutaneous implantable cardioverter defibrillator (S-ICD) and congenital heart disease. **A**, 26-year-old male after lateral tunnel Fontan had undergone epicardial pacemaker for symptomatic sinus node dysfunction. **B**, The patient underwent generator replacement with a dedicated bipolar device to avoid future interdevice interaction, as well as relocation to the right upper abdomen to exclude the device from the future S-ICD shock vector.

in Table 1. The most common congenital cardiac diagnosis was single ventricle physiology in 11 patients (52%), palliated by either Fontan surgery (n=9) or by aortopulmonary shunt (n=2). Seven patients (33%) had either an existing epicardial (pacemaker in 4 and ICD in 2) or transvenous CIED (ICD in 1). Postimplant pacing was continued in 6 of 7 patients: 5 for sinus node dysfunction (4 using an algorithm to minimize ventricular pacing and 1 programmed to AAIR) and 1 programmed to DDD for underlying atrioventricular block. Two patients (hypoplastic left heart syndrome palliated by an extracardiac Fontan operation and *d*-transposition of the great arteries after Mustard operation with intracardiac right-to-left shunt) had failure of a prior epicardial defibrillation system before S-ICD placement. There was one prior transvenous ICD system failure in a patient with hypoplastic right ventricular and surgical outflow tract reconstruction. One patient underwent surgical replacement/relocation of an existing pacemaker generator before S-ICD placement to avoid potential interdevice interactions (Figure 1). In addition to standard baseline eligibility screening in supine and standing positions, 1 patient also underwent exercise stress testing, and 2 patients were screened while pacing at heart rates compatible with peak exercise through an existing CIED.

S-ICD Indications

Reasons for ICD placement are listed in Table 2 and consisted of secondary prevention indications in 7 patients (33%; documented sustained ventricular tachycardia (VT) in 4 and aborted cardiac arrest in 3) and primary prevention indications in 14 patients (67%). The median number of primary prevention indications per patient was 3 (range 1–3). The main reason for selecting the S-ICD was absent or limited venous connection to the subpulmonary ventricle in 10 patients (because of Fontan palliation in 9 and tricuspid valve replacement in 1), presence of an intracardiac shunt in 5, lack of vascular access in 3, patient/physician preference in 2, and severe pulmonary hypertension in 1.

Implant Characteristics

The pulse generator was placed in the left midaxillary line in all but one patient with dextrocardia, where it was placed at the right midaxillary line. The defibrillation coil was placed

in a parasternal location ipsilateral to the pulse generator in 20 (95%) and contralateral in 1 (5%) procedure. See Figure 2 for examples of postimplant chest x rays. Defibrillation testing was attempted in 18 patients at the time of implant (86%) and resulted in conversion of induced ventricular arrhythmia

Table 2. S-ICD Characteristics

Characteristic	N=21
ICD indication	
Primary prevention	14 (67)
NSVT	7
Inducible VT	8
Syncope	9
Depressed systemic ventricular EF	8
Prolonged QRS duration	2
Other	3
Secondary prevention	7 (33)
Aborted cardiac arrest	3
Sustained VT	4
General anesthesia	12
Conscious sedation	9
VT/VF induced at implant	17 (81)
Defibrillation successful	17 (81)
Implant conditional zone, ms	200 (190 - 210)
Implant shock zone, ms	225 (220 -238)
Hospital length of stay, days	1.5 (1 - 6)
Postimplant programmed vector	
Primary	9 (43)
Secondary	7 (33)
Alternate	5 (24)

Numbers in parentheses represent percent (%) or interquartile range (IQR). EF indicates ejection fraction; ICD, implantable cardioverter-defibrillator; NSVT, nonsustained ventricular tachycardia; S-ICD, subcutaneous implantable cardioverter defibrillator; VF, ventricular fibrillation; and VT, ventricular tachycardia.

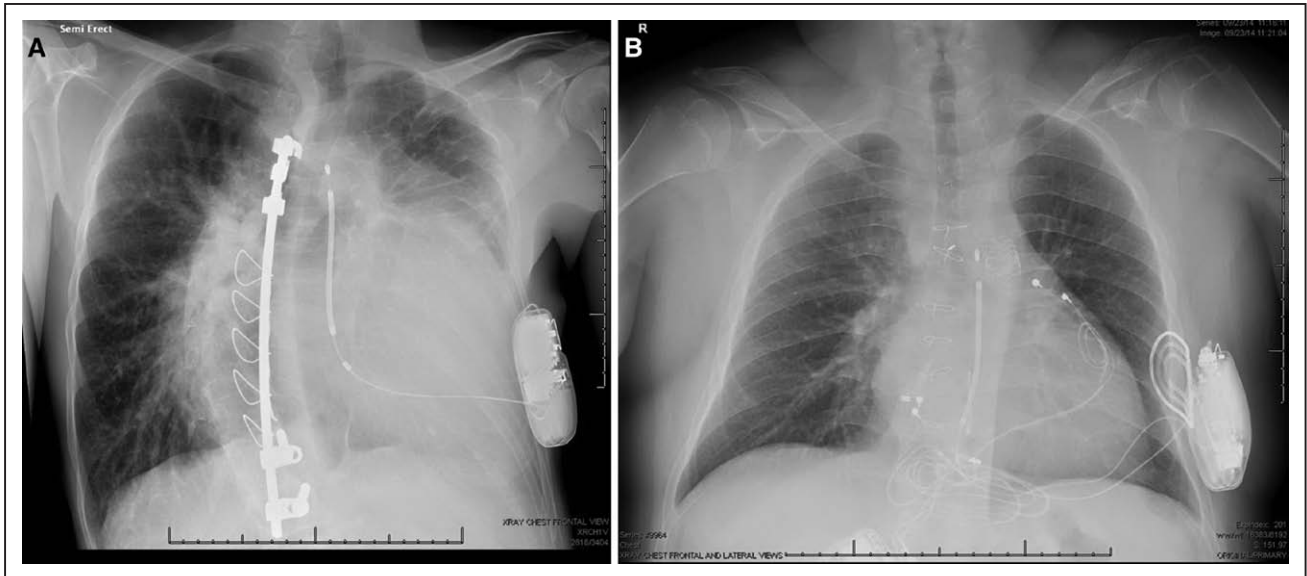


Figure 2. Examples of postimplant chest x rays. **A**, 41-year-old male with tricuspid atresia and pulmonary stenosis previously palliated by Waterston shunt in childhood who underwent subcutaneous implantable cardioverter defibrillator (S-ICD) placement for a left ventricular ejection fraction (LVEF) of 35% and **(B)** 22-year-old male with hypoplastic left heart syndrome palliated by extracardiac Fontan operation who had a previously failed epicardial implantable cardioverter defibrillator (ICD) system that had been placed for nonsustained ventricular tachycardia (VT) in the setting of unexplained syncope and systemic right ventricular ejection fraction (RVEF) of \approx 10%.

at an output of 65 Joules in 15 of 17 (88%) patients and \leq 80 Joules for all patients. One patient could not be converted with an output of 65 Joules, despite 2 attempts, but was converted to sinus rhythm with a third attempt using 80 Joules. The next patient at the same center was successfully converted with an initial output of 80 Joules. Defibrillation testing was not attempted in 3 patients because of operator discretion. For one additional patient, induction of ventricular arrhythmia was unsuccessful, resulting in deferral of further testing. Dual zones were programmed for tachyarrhythmia detection in all patients except one. There was one acute implant complication related to device infection that was treated medically and did not require device removal. See Table 2 for a more detailed description of the procedural characteristics.

Outcomes

Follow-up data were available for 20 patients over a median period of 14 months (interquartile range 3–19 months). During this time, 4 patients experienced inappropriate shocks (IAS; 20%), and 1 patient received appropriate shocks (5%). IAS were delivered for (1) supraventricular tachycardia with a ventricular rate greater than the conditional zone in a patient with unrepaired tetralogy of Fallot and pulmonary atresia, (2) T-wave oversensing in a patient with bicuspid aortic valve and aortic valve replacement, and (3) postprocedural subcutaneous air, resulting in a decrease in the sensed QRS amplitude and detection of low-amplitude artifact in a patient with prior Mustard operation. The final patient with IAS is described in greater detail below. See Figure 3 for examples of inappropriate device shocks. One patient with congenital pulmonic stenosis and previous right ventricle outflow tract muscle resection and later pulmonary valve replacement received a total of 15 appropriate shocks for sustained monomorphic VT over a period of 16 months after S-ICD implantation. The VT

was ultimately controlled with a combination of antiarrhythmic drugs and catheter ablation.

During follow-up, 2 patients died and 2 underwent cardiac transplantation. One death resulted from pulmonary hemorrhage in the postprocedural period unrelated to device implantation. The second death occurred in a patient who developed an asystolic arrest and later in the episode received multiple IAS because of autogain-induced oversensing of asystole and agonal rhythm (Figure 4). This patient had double-inlet left ventricle palliated by intra-atrial conduit Fontan surgery. The S-ICD had been placed after an episode of syncope while driving and the subsequent finding of inducible sustained VT at electrophysiology study. Acute defibrillation testing was successful at an output of 65 Joules. One month after S-ICD placement, the patient was found unconscious on the floor at home. By the time of arrival to the referring center, the patient had developed multiorgan dysfunction and severe neurological sequelae and expired on withdrawal of support.

Discussion

This is the largest multicenter study of S-ICD implantation for ACHD and provides a detailed description of the underlying anatomy, reason for S-ICD implant, procedural aspects, and clinical performance of the S-ICD in this population. Previous experience with this technology has generally been limited to case reports and small series that describe the feasibility of implantation with limited follow-up.^{8,13} Recently, a pooled analysis from the combined EFFORTLESS and IDE postmarket studies containing a modest number of adult patients with congenital heart disease ($n=16$) was published.¹⁴ The results of this registry suggested reasonable sensing discrimination, but the study was limited by a lack of detail regarding surgical palliation and an absence of tachyarrhythmia events during

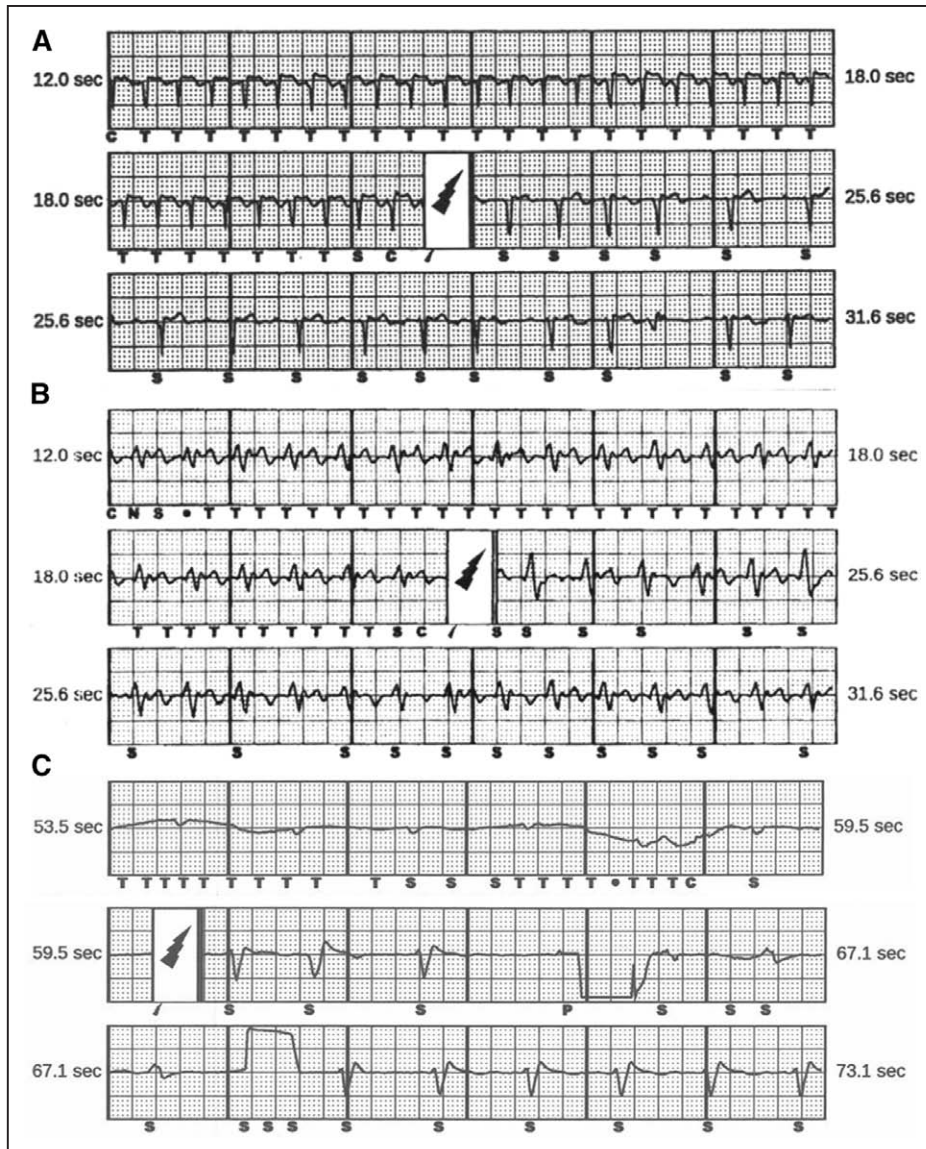


Figure 3. Examples of inappropriate subcutaneous implantable cardioverter-defibrillator (S-ICD) therapies. **A**, Inappropriate detection of supraventricular tachycardia in a 34-year-old male with unrepaired tetralogy of Fallot and pulmonary atresia, **B**) T-wave oversensing in a 26-year-old male with previous aortic valve replacement, and **C**) transient oversensing of low-amplitude artifact related to subcutaneous air the night of the procedure in a 35-year-old male with *d*-transposition of the great arteries and prior Mustard operation.

follow up. Given the significant heterogeneity of the congenital heart population in terms of cardiac morphology, distribution of ventricular mass, and known variable response to defibrillation,^{15,16} further detailed analysis of this new technology is needed. Important findings of this study include the following: (1) the 2 most frequent indications for S-ICD placement in this ACHD population were absence of a systemic venous connection to the heart because of prior surgery and the presence of residual intracardiac shunt, (2) reliable detection and conversion of induced ventricular arrhythmia with the S-ICD can be achieved despite heterogeneous anatomy, and (3) rhythm discrimination seems to be acceptable up to 1-year postimplant.

The most common reason for implantation of an S-ICD in the present study was lack of a venous connection to the subpulmonary ventricle after Fontan palliation. ICD placement may be particularly problematic in this group of patients because transvenous options are limited-to-nonexistent, and surgical

approaches are typically required. A variety of techniques have been proposed to implant defibrillation systems in this patient group, all involving either a sternotomy for epicardial placement or hybrid transvenous/epicardial approach.^{7,15} Limitations of these strategies include the need for frequent system revisions, inadequate or evolving defibrillation thresholds over time, and system failure, particularly, with epicardial patches. Given that the single ventricle population contributes significantly to the burden of SCD in the ACHD community² yet represents only a small fraction of total ICD implants,¹⁰ this group seems to be underrepresented with respect to SCD prevention. The S-ICD may, therefore, represent a more straightforward and uniform approach for this particularly vulnerable population.

Clinical rhythm discrimination for the S-ICD in the ACHD population has not been extensively described.¹⁴ Despite significant variations in congenital anatomy, surgical repair, and resultant abnormalities in electrocardiographic

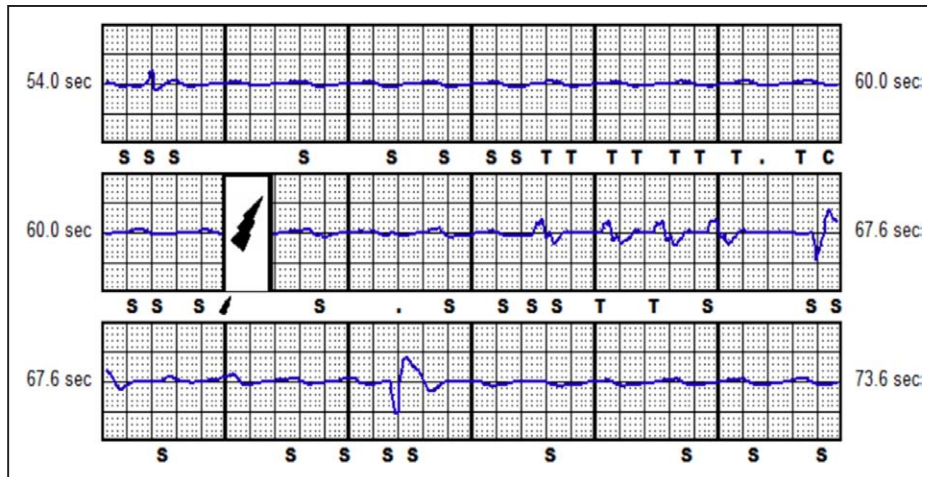


Figure 4. Detection of low-amplitude artifact during a period of asystole in a 30-year-old male with double-inlet left ventricle palliated by intra-atrial conduit Fontan operation.

appearance, IAS were observed in only 4 patients (20%) during the study period. After exclusion of the asystolic arrest in which therapy was delivered during a fatal arrhythmic event, the proportion of IAS (15%) is only marginally higher than that of the initial Food and Drug Administration trial for the S-ICD (13.1%) and somewhat higher than in the EFFORTLESS registry (7%). Although IAS represented the most significant morbidity after S-ICD placement, the causes of events were not unique to the ACHD population, and many were potentially preventable.^{11,17,18} Previous data suggest that the IAS rate may decrease with increased operator experience and improved device programming.¹ Moreover, specific solutions for the most common causes of IAS exist. For example, device therapy for sustained VT at a rate above the conditional zone, as occurred with the first patient in this series, may arguably be appropriate, but could be prevented by a variety of methods, including increasing the rate of the conditional zone, administration of AV nodal blocking agents, and preemptive catheter ablation.¹¹ T-wave oversensing after S-ICD placement can also be largely avoided by postimplant template acquisition during exercise testing.¹⁷ Finally, IAS related to postprocedural subcutaneous air entrapment can be avoided by careful review of the postimplant chest x ray. When present, it can be dealt with by disabling of detection for the first 24 hours after implant or reprogramming of the sensing vector.¹⁸ Given that the present population was a particularly high-risk group with limited alternatives for ICD placement, strategies that aggressively preempt these types of inappropriate therapies may be preferable to the implantation of alternative technologies with their associated limitations.

Perhaps, most importantly, interdevice interactions were not found to be a source of sensing abnormalities in this ACHD cohort. This issue deserves greater consideration in the ACHD population because of the concomitant need for bradycardia pacing in many patients at risk for SCD. As opposed to prior clinical studies of the S-ICD where a pacing system was only present at the time of placement in 1% to 3% of patients,^{1,19,20} a coexisting CIED was present in approximately one third of patients in the present study. Unipolar pacing should be avoided after S-ICD placement because this may

result in both inappropriate detection and classification of paced events as ventricular arrhythmia, as well as impede the detection of true events. The latter phenomenon is of great importance because of the potential for untreated ventricular arrhythmia. Pacemaker generators that automatically revert to unipolar pacing should, therefore, be avoided in association with S-ICD placement, a consideration that led to surgical replacement of an existing epicardial device for one patient in the present study (Figure 1). When it is a programmable feature, adaptive lead monitoring should also be disabled for these devices. Although left to the discretion of the operator, defibrillation testing with asynchronous pacing through the existing CIED was felt to be beneficial in many instances to exclude the possibility of future life-threatening interdevice interactions.²¹

Known drawbacks of the S-ICD as compared with conventional ICDs include (1) absence of antibradycardia pacing, (2) inability to pace-terminate monomorphic VT, and (3) no opportunity for resynchronization therapy.²² The first of these is highly relevant to the ACHD population where sinus node dysfunction and high-grade AV block may be prevalent depending on the underlying congenital lesion and subsequent cardiac surgery. The single arrhythmic death in the present study was attributable to severe bradycardia manifesting as asystole in a single ventricle patient without the possibility for bradycardia pacing. This tragic outcome highlights one of the critical limitations of the S-ICD in the ACHD population. In addition, anti-tachycardia pacing would likely have been of benefit to another patient in this study who experienced recurrent, albeit effective, appropriate shocks for monomorphic VT. These issues should be carefully considered before recommending this device in clinical practice.¹¹

Limitations

This study was retrospective and, therefore, subject to associated limitations. Relevant information, such as the number of screening failures for potential S-ICD candidates over the study period at each institution could not be obtained, nor were the results of the preprocedure screening available for most of the patients included in this study. The study was also

limited by relatively small sample size; despite aggregate data from 7 ACHD centers, clinical experience with S-ICD performance in terms of clinical VT/ventricular fibrillation detection and conversion efficacy remains rudimentary. To circumvent these issues, a prospective multicenter study involving several major ACHD programs is recommended.

Conclusions

S-ICD implantation is feasible in the ACHD population. Most candidates have single ventricle heart disease and limited transvenous options for ICD implantation or intracardiac shunts. The present findings suggest that the S-ICD can be implanted safely, with reliable conversion of induced ventricular arrhythmia and with acceptable long-term sensing in the ACHD population. Further prospective evaluation of this treatment modality is needed.

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Disclosures

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Clinical Experience With the Subcutaneous Implantable Cardioverter–Defibrillator in Adults With Congenital Heart Disease

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