IMPLANTABLE DEFIBRILLATION THERAPY

COMPLEX CASE STUDY

The Subcutaneous Implantable Cardioverter-Defibrillator as Part of Dual Device Therapy in Complex Congenital Heart Disease

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ABSTRACT. We report the case of a 44-year-old with unrepaired complex congenital heart disease (CHD), a pre-existing permanent pacemaker, and a subcutaneous implantable cardioverter-defibrillator (SICD) implanted for secondary prevention. Right parasternal electrode positioning and step-down defibrillation testing were integrated. Application of the SICD for device therapy in CHD patients remains quite limited but can have an expanding role in this population.

KEYWORDS. adult congenital heart disease; pediatrics; subcutaneous implantable cardioverter-defibrillator.

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Introduction

Advances in cardiac implantable electronic device (CIED) technology have introduced new devices that may avoid known complications related to standard transvenous systems. They have also extended the benefits of specific CIED functions, such as defibrillation, to subgroups of patients who may have absolute or relative contraindications to devices that ordinarily require transvenous lead passage. We present a case highlighting both of these in a patient with unrepaired complex congenital heart disease (CHD) implanted with a subcutaneous implantable cardioverter-defibrillator (SICD). This is followed by an extended discussion and review of the current experience with the SICD and specific considerations when contemplating its application in patients with CHD.

Case report

A 44-year-old male with unrepaired complex CHD consisting of congenitally corrected transposition of the

great arteries (ccTGA), large unrestrictive ventricular septal defect (VSD), and pulmonary stenosis has been conservatively managed without prior surgical procedures or percutaneous interventions. Four years prior, he was treated at an outside hospital on several occasions for atrial fibrillation resulting in electrical cardioversions and initiation of amiodarone. Given his arrhythmia history, cardiac catheterization was arranged along with a diagnostic electrophysiologic study to evaluate for other conventional arrhythmia substrates that may have secondarily degenerated into atrial fibrillation. While no other SVT mechanism was identified, a markedly prolonged HV interval was measured and pacemaker implantation was recommended, particularly in light of the known progressive risk of heart block in ccTGA. A transthoracic echocardiogram performed at that time demonstrated normal systemic right ventricular and subpulmonary left ventricular systolic function. A transvenous device was implanted following thorough discussion with the patient regarding the risks of thromboembolism with transvenous leads and concomitant intracardiac shunts, which the patient was willing to accept in favor of a less invasive implant approach and chronic anticoagulation.

Four years later, in an effort to reduce the risks of amiodarone toxicity, the patient was transitioned to flecainide at a low dose without evidence of subsequent electrocardiographic abnormalities. Approximately 1 month

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Figure 1: Recorded pacemaker atrial and ventricular channel electrogram tracings revealing rapid ventricular tachycardia with cycle length of 240 ms.

after flecainide initiation, the patient experienced a syncope event. Cardiopulmonary resuscitation was performed with subsequent return of consciousness without neurologic sequelae. The patient was transported by ambulance to an outside hospital where diagnostic catheterization and echocardiography were performed, showing no significant hemodynamic or functional changes. Pacemaker interrogation revealed an episode of rapid ventricular tachycardia (VT) (**Figure 1**). Due to concerns over the possibility of flecainide-related proarrhythmia, flecainide was discontinued and amiodarone was restarted.

On subsequent outpatient follow-up, the patient reported a near-syncope event that coincided with a second spontaneous VT episode captured through his pacemaker. Given this second unprovoked event, this time occurring off flecainide, a recommendation was made to proceed with secondary prevention ICD implantation. A thorough discussion regarding options for ICD implantation was conducted with the patient and included transvenous implantation with or without concomitant lead extraction, surgical ICD implantation, and SICD implantation. The patient agreed most strongly with proceeding with the SICD.

Pre-implantation surface electrocardiogram (ECG) screening was performed. Screening was carried out during paced and native ventricular rhythms, in supine and standing positions, and with left and right parasternal electrode positions. Using a left parasternal electrode position, the patient failed screening in all vectors and positions during his native ventricular rhythm and

Figure 2: Final single-shot fluoroscopic image of implanted SCD and transvenous dual chamber pacemaker.

passed in only the alternate vector during ventricular pacing. Screening was successful during both ventricular rhythms while standing and lying supine with a right parasternal proximal electrode-to-left chest can configuration (primary vector). Device implantation proceeded without complication (**Figure 2**). A three-incision technique was employed and the procedure was completed with minimal fluoroscopy (< 1 minute). Excellent ECG signals were observed with the recommended electrode and device positions (**Figure 3**). The patient's pacemaker

Figure 3: Example of SICD sensed electrogram showing clear P and R wave deflections without significant artifact or over-sensing.

pulse generator was concomitantly exchanged for a generator that could be programmed to disable automatic switching from bipolar to unipolar pacing. Given the complex CHD substrate and less conventional SICD electrode positioning, the decision was made to perform complete defibrillation threshold (DFT) testing. Step-down testing was performed from 65 joules to 15 joules with failure to defibrillate at 15 joules, but confirmed success at 25 joules on two separate inductions. On follow-up, the patient has done well with normal SICD and transvenous pacemaker functions.

Discussion

The SICD has become a reasonable device to consider in individuals meeting an accepted indication for ICD implantation, who lack a need for permanent pacing, and who do not have documented ventricular arrhythmias successfully terminated with antitachycardia pacing. Experience in using the SICD in CHD patients is quite limited. In the EFFORTLESS SICD registry, only 7% of recipients had CHD.¹ The number of pediatric versus adult SICD recipients with CHD was also not explicitly reported in the registry publication. While the SICD theoretically could have several advantages in the CHD population, patient eligibility, selection, and post-implantation experience remain low. This may, in part, be attributable to the concomitant pacing needs that these patients frequently have, but may also relate to a relatively unproven platform compared to standard transvenous ICDs in this population. Additionally, the comparatively large footprint of the first-generation device compared to transvenous generators discouraged implantation in smaller-sized patients.

The presented case highlights three important considerations when integrating the SICD in the implantation of defibrillators in CHD patients: consideration of a right parasternal electrode course, DFT testing, and concomitant SICD use with pacemakers.

Pre-procedural ECG screening remains a critically important step in determining candidacy for SICD implantation. Observation of desired R and T wave signal amplitudes in at least 1 of 3 sensing vectors is recommended in order to proceed with device placement. The challenge that arises in CHD patients is the issue of the varying cardiac

positions and orientations that can exist and may affect sensing in the traditional vectors with the SICD. Alternate vectors have been introduced, including a right parasternal electrode course. Left and right parasternal electrode sensing in CHD and non-CHD patients was evaluated by Wilson et al.² Among 37 patients screened in the study, 27 patients had adult CHD, including TGA (10), tetralogy of Fallot (10), and single ventricle physiology (7), while 10 patients served as controls with normal hearts. Among the adult CHD group as a whole, there was no significant difference in the R and T wave amplitudes and the R:T ratio in either left or right parasternal positions. Significant differences were noted in the control group, primarily accounted for by differences in the alternate sensing vector. In our patient's case, the right parasternal electrode position was demonstrated to be ideal during pre-procedural screening in the context of complex CHD with ventricular inversion and levocardia. Post-implantation sensing with the right parasternal electrode and standard left lateral chest wall generator placement afforded excellent results during both native and paced ventricular rhythms.

Defibrillation threshold assessment has been studied in limited fashion with the SICD. Systematic testing is not required and has not been consistently applied in pre- or post-approval studies. Formal DFT values from pooled data were not reported in the EFFORTLESS registry either. While functional defibrillation efficacy appears excellent, true threshold ranges are lacking. Bardy et al reported DFTs of 36.6 ± 19.8 joules based on early clinical testing of functional systems.³ We recently reported our experience with very high DFTs and failed defibrillation at maximum output in morbidly obese patients receiving SICDs with conventional electrode positioning.⁴ An off-label approach to address defibrillation failure with conventional left parasternal electrode position was reported by Guenther et al, where the electrode was tunneled beneath the sternum from a subxyphoid incision, resulting in successful defibrillation.⁵ Formal step-down testing was not performed in that case. Even less is known regarding DFT ranges in pediatric and adult CHD patients. A step-down approach to DFT testing was performed in our patient, demonstrating defibrillation failure at 15 joules, but repeated successful defibrillation at 25 joules. While the

programmed therapy cannot be configured to anything besides maximum shock output (80 joules) for real-life events, demonstration of this wide safety margin was reassuring. A right parasternal electrode position could possibly afford improved defibrillation efficacy owing to a broader area of coverage around the heart and better opposing position of the electrode relative to the can, though this would need to be systematically evaluated. When these results are coupled with those from Wilson et al regarding parasternal electrode position for sensing, a reasonable question to ask is whether a right parasternal electrode position, in general, may be more favorable in CHD patients if sensing remains consistent and more reasonable DFTs are achieved in a cohort undergoing formal step-down testing.

Patients who require permanent pacing or who have ventricular arrhythmias that are amenable to antitachycardia pacing are not traditionally eligible for use of the S-ICD alone for treatment of their arrhythmia conditions. Of special note is the application of the S-ICD in patients with previously implanted pacemakers, as in our patient's case. In such a scenario, an S-ICD can be considered in a patient who has their pacing support addressed with a separate system, while utilizing the S-ICD for ventricular tachyarrhythmia detection and treatment only. Patients with concomitant S-ICDs and pacemakers comprised only 2.5% of combined registry patients.^{1,6} Currently, it is recommended that automatic switching from bipolar to unipolar pacing configurations be disabled in pacemaker generators, as unipolar pacing may lead to over-sensing of a large unipolar pacing artifact and inappropriate under-sensing of ventricular arrhythmias by the S-ICD. In cases where pacemaker generators cannot be programmed in this manner, elective replacement has been recommended with a pulse generator that supports this programming option. Implantation of a leadless pacemaker and S-ICD was reported by Mondésert et al.⁷ The patient initially underwent primary prevention S-ICD implantation in the setting of bilateral subclavian vein stenosis and presence of a hemodialysis catheter. Complete heart block developed months after S-ICD implantation, prompting leadless pacemaker implantation, again due to the vascular access limitations. Single-chamber pacing sufficed, as the patient also had permanent atrial fibrillation. The authors reported that maximum bipolar pacing through the leadless device did not result in over-sensing by the S-ICD, and maximum S-ICD shock delivery did not negatively impact leadless pacemaker function or position. Such a combination of implants could be considered in individuals only requiring single-chamber pacing and defibrillation protection in favor of preservation of the vasculature and elimination of issues related to transvenous leads. Interestingly, a right parasternal electrode position was also selected in that patient due to large T wave amplitude in the left parasternal configuration during screening.

Early S-ICD implant experience and outcomes data have generally been encouraging.^{1,6} The EFFORTLESS post-market registry has reported excellent defibrillation efficacy and progressive declines in complication rates

and inappropriate shock incidence, mostly attributable to increased implant experience and changes in implantation technique and device programming in response to observed complications in the past. However, the overall number of implants, duration of follow-up, and the number of actual shock events remain proportionately small in comparison to the experience with transvenous ICDs. Two-year follow-up shows predominant uncomplicated recovery following S-ICD implantation with normal device function and excellent defibrillation success for appropriately sensed ventricular tachyarrhythmias (90.1% for first-shock conversion efficacy and 98.2% for overall shock conversion efficacy). More specific to CHD patients, the S-ICD offers a very reproducible implantation approach and is not dependent on venous or intracardiac anatomy. Inappropriate shock incidence has been of concern, but appears to be declining with improvements in device programming and utilization of discriminators within a programmable conditional shock zone. However, the estimated 3-year inappropriate shock rate was still 13.1% overall among registry patients. The large size of the generator has been of some concern when considering implantation in smaller-sized patients. Among combined registry data, approximately 20% of all complications could be attributable to the pocket itself. In a published single-center experience with the S-ICD in pediatric and adult patients, a 19% incision site complication was reported (all cases involving pediatric patients), though non-device factors following implantation influenced the development of this complication in the majority of cases.⁸

Conclusion

Early experience with the S-ICD in young patients and those with CHD is growing but remains limited. This technology can provide life-saving treatment for prevention of sudden death in some patients with CHD, as well as in young patients where preservation of venous patency is desired. The S-ICD with its features, potential benefits, and limitations should be considered in these unique patient groups. Careful patient selection, implantation technique, and device programming are imperative to ensure appropriate device function and patient satisfaction. This reported case highlights the importance of considering an alternate right parasternal electrode position, consideration of formal DFT testing at the time of implantation to ensure adequate functionality and a safety margin, and appropriate programming of pacemaker pulse generators in patients with concomitant S-ICD/permanent pacemaker use.

References

1. Lambiase PD, Barr C, Theuns DA, et al. Worldwide experience with a totally subcutaneous implantable defibrillator: early results from the EFFORTLESS S-ICD registry. *Eur Heart J*. 2014;35(25):1657–1665.
2. Wilson DG, Zeb M, Veldtman G, Dimitrov BD, Morgan JM. Left and right parasternal sensing for the S-ICD in adult congenital heart disease patients and normal controls. *Pacing Clin Electrophysiol*. 2016;39(3):282–290.

3. Bardy GH, Smith WM, Hood MA, et al. An entirely subcutaneous implantable cardioverter-defibrillator. *N Engl J Med*. 2010;363(1):36–44.
4. Do K, Carlson S, Konecny T, Chang PM, Doshi RN. High defibrillation threshold in obese patients receiving the subcutaneous implantable cardioverter-defibrillator. Poster presented at: 37th Annual Scientific Sessions Heart Rhythm Society (HRS); May 4–7, 2016; San Francisco, CA.
5. Guenther M, Kolschmann S, Knaut M. Substernal lead implantation: a novel option to manage DFT failure in S-ICD patients. *Clin Res Cardiol*. 2015;104(2):189–191.
6. Burke MC, Gold MR, Knight BP, et al. Safety and efficacy of the totally subcutaneous implantable defibrillator. *J Am Coll Cardiol*. 2015;65(16):1605–1615.
7. Mondésert B, Dubuc M, Khairy P, Guerra PG, Gosselin G, Thibault B. Combination of a leadless pacemaker and subcutaneous defibrillator: first in-human report. *Heart Rhythm Case Rep*. 2015;1(6):469–471.
8. Jarman JW, Lascelles K, Wong T, Markides V, Clague JR, Till J. Clinical experience of entirely subcutaneous implantable cardioverter-defibrillators in children and adults: cause for caution. *Eur Heart J*. 2012;33(11):1351–1359.