Subcutaneous implantable cardioverter defibrillator placement in a 5-year-old patient: Modifications for the smallest patients

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Introduction

The subcutaneous implantable cardioverter defibrillator (S-ICD) has become a valid alternative to the traditional transvenous implantable cardioverter defibrillator (ICD) in patients who do not require antitachycardia/antibradycardia pacing or cardiac resynchronization therapy.[1] The absence of intravascular or intracardiac components removes several complications of transvenous systems that make it particularly favorable in younger patients. Limiting factors in children have been the relatively large size of the pulse generator, concerns of device erosion, and a mandatory distance between sensing electrodes.

Case report

The patient is a 5-year-old, 20.1 kg, 122 cm female child presenting with cardiac arrest following an emotional outburst with documented ventricular fibrillation. Successful cardioversion required several external defibrillations and the patient was initially maintained on amiodarone, followed by propranolol, at an outside institution. Electrocardiograms were unremarkable and showed no evidence of preexcitation, QT prolongation, Brugada syndrome, or epsilon wave of arrhythmogenic right ventricular cardiomyopathy. An echocardiogram and cardiac magnetic resonance imaging revealed normal cardiac/coronary anatomy and normal biventricular function. Provocative testing with isoproterenol and procainamide were negative. A limited long QT syndrome genetic panel had been sent several years prior following an atypical syncopal event and was negative. An expanded panel was sent following the most recent event and demonstrated a pathologic mutation in *CALM1* c.398G>A (p Glyl33Glu), though this information was not available during the current hospitalization secondary to family preference to delay genetic testing.

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S-ICD screening was performed while the patient was seated and standing and revealed appropriate screening vectors during rest and elevated heart rates in the left secondary and right primary, secondary, and alternate positions. Given the patient's age and motor delays in the early arrest period, screening during the administration of isoproterenol was substituted for an exercise stress test to assess the effect of elevated heart rates on eligibility.

Implantation

The S-ICD implant was performed with the patient intubated and under general anesthesia. The xyphoid process, costal margins, and suprasternal notch were marked prior to prepping the chest. For implant planning purposes, a model S-ICD was secured into the left axilla, approximately at the fourth intercostal space, and in the midaxillary line. A model lead was secured from the device to an area 1 cm superior and rightward to the xyphoid process and 1 cm inferior and leftward to the suprasternal notch. Given the patient's small size, this arrangement achieved sensing in a right primary vector (electrode rightward of the xyphoid process to device generator) and a left secondary vector (electrode leftward of the suprasternal notch and device generator). A completely right parasternal lead position was considered but the resultant lead curve, projecting either extreme rightward or leftward, did not position the heart shadow appropriately between the device generator and coil (Figure 1). The chosen position also allowed the necessary spacing for the electrodes. These locations were then marked on the skin with a marker.

The patient was prepped and draped in the typical manner and a 6 cm curvilinear incision was made in the left axilla and a subcutaneous pocket created inferiorly using blunt dissection. The anterior margin of the latissimus major was identified, and care was taken to ensure that the pocket did not extend below it. Additionally, the pocket did not extend anteriorly such that it would encroach on breast tissue. A 2 cm transverse incision was then made at the right xyphoid location and 2 0-silk sutures were placed in the fascial plane for use to later secure the lead. The manufacturer tunneling tool was used to create a subcutaneous tract extending from the

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KEY TEACHING POINTS

- The subcutaneous implantable cardioverter defibrillator can be successfully implanted in pediatric patients of small size.
- Modifications to the traditional implant procedure may be necessary and include screening in the right and left parasternal positions and creation of a "C-curve" in the shock coil to place the sensing electrodes.
- An axillary approach to pulse generator placement may prevent erosion in the smallest patients.

xyphoid pocket to the inferior aspect of the axillary pocket. A 48-cm-length 0-silk suture was used to tie the anchoring hole of the lead to the tunneling tool, creating a 15–16 cm loop. The lead was pulled through the tract and secured to the floor of the xyphoid pocket. Care was taken to ensure that the sensing electrode had a near vertical orientation and was positioned over a bony aspect, with minimal intervening adipose tissue. A 2 cm vertical incision was then made at the superior left sternal location and a small pocket to the fascial layer was created. With the ICD lead attached by suture, the tunneling tool was used to create a subcutaneous tract from the inferior to superior pocket and the lead pulled through and secured. This orientation created a "C-curve" with the redundant shocking coil projecting rightward (Figure 1).

The lead was attached to the device, which was placed into the pocket and secured with 2 0-silk stay sutures. The 3 pockets were washed using bacitracin saline and closed with a single layer of 2-0 dissolvable interrupted sutures. Sensing revealed appropriate R:T ratios in the primary and secondary vectors with the secondary sensing vector selected by the device. The alternate sensing vector was not appropriate with low-amplitude R waves (Figure 2). Defibrillation testing was then conducted with 50 Hz burst, placing the patient into ventricular fibrillation. The S-ICD appropriately detected the ventricular fibrillation and successfully defibrillated with a single 40 J shock following 14.4 seconds for detection and charging (Figure 3). The device was programmed with a conditional ventricular tachycardia zone at 220 beats per minute and a ventricular fibrillation shock zone at 240 beats per minute.

The overlying skin was closed using 3-0 dissolvable interrupted and 4-0 dissolvable subcuticular sutures. Occlusive dressings were placed over the incisions and remained for 48 hours prior to removal.

Follow-up

The incisions were clean, dry, and intact at the 1-week postoperative wound check. During an 18-month follow-up period, the patient has done well, demonstrating excellent wound healing and no evidence of skin erosion at the generator site. She has been maintained on a beta blocker with no sensed ventricular arrhythmias or appropriate and inappropriate device discharges. Following her cardiac arrest, the patient has been followed by Physical Therapy, who notes no limitations to movement of her left arm and shoulder. Though palpable as it crosses the superior sternum, the lead is not visible under the skin and causes no discomfort.

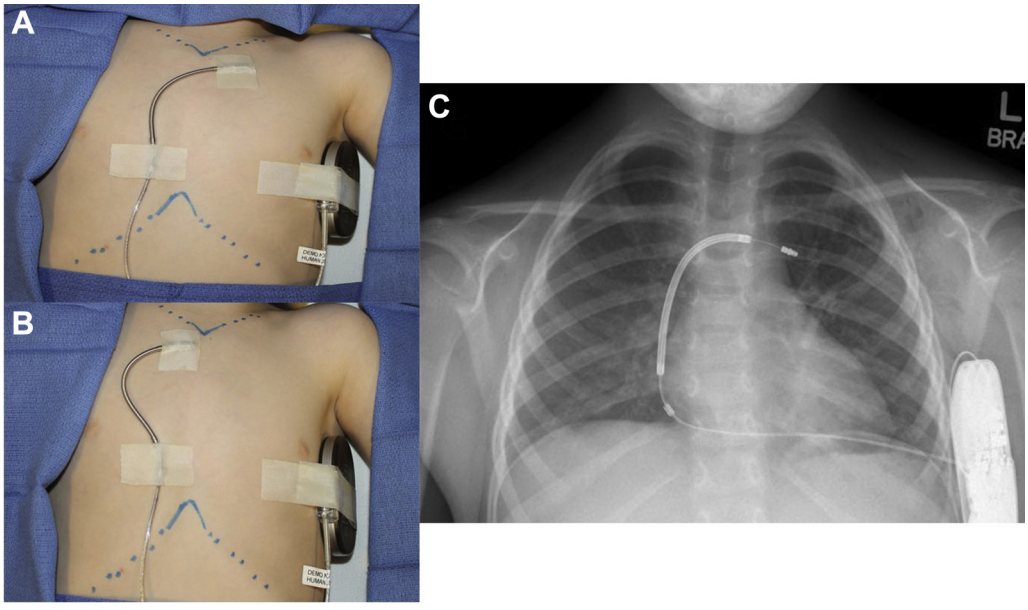

Figure 1 Model lead and generator secured **A:** with both inferior and superior sensing electrodes in a right parasternal position and **B:** with inferior sensing electrode in a right parasternal and superior electrode in a left parasternal position. **C:** Anteroposterior chest radiograph demonstrating final pulse generator and sensing lead positioning. The inferior sensing electrode is secured to the right of the xyphoid with the superior electrode secured in a left parasternal position. The redundant lead creates a "C-curve" projecting rightward.

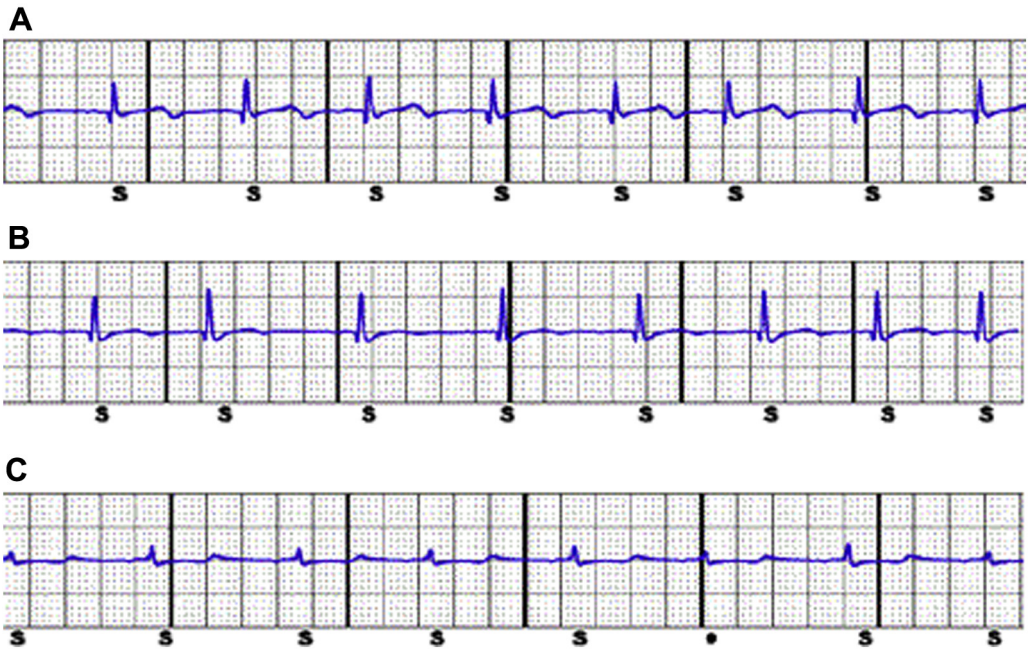

Figure 2 Sensing revealed appropriate R:T ratios in the primary (**A**) and secondary (**B**) vectors, with the secondary sensing vector selected by the device. **C:** The alternate sensing vector was not appropriate, with low amplitude R-waves.

Discussion

This report describes the smallest known patient successfully implanted with an S-ICD via the subcutaneous approach. Our center previously described successful placement in a 13.5 kg patient, though this was via retroperitoneal generator placement.[2] The S-ICD has been demonstrated to be a viable alternative to the traditional transvenous ICD for patients that do not require therapeutic pacing. It avoids the potential

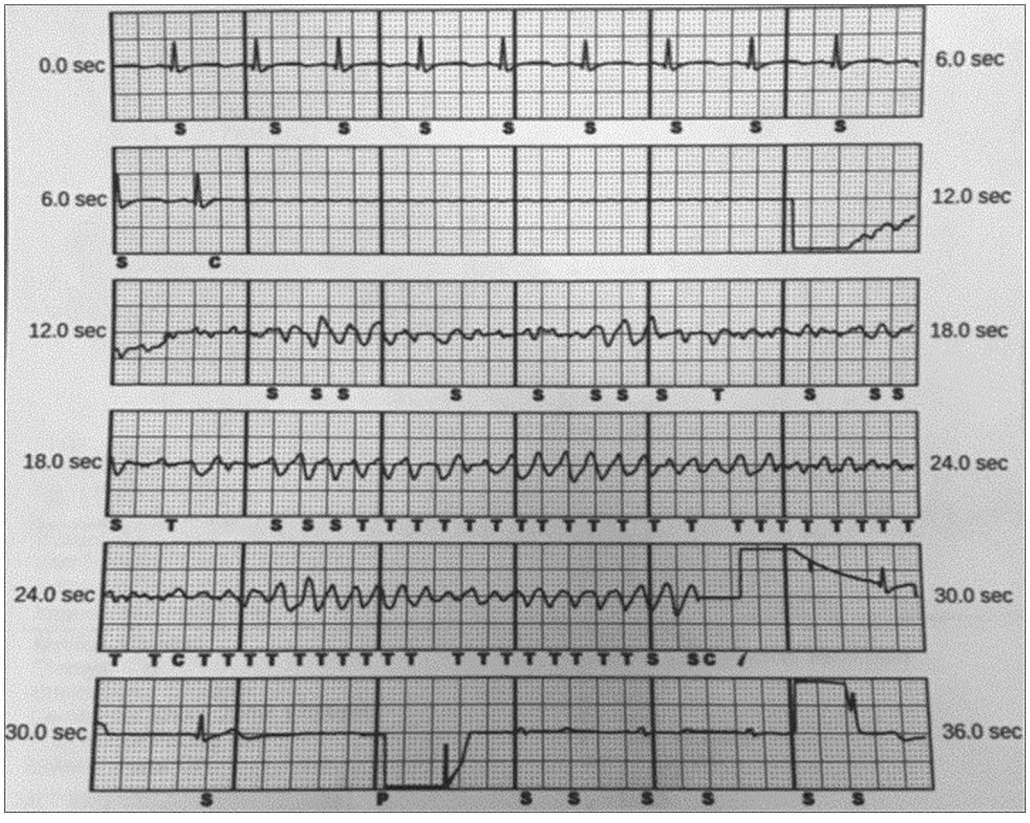

Figure 3 Ventricular fibrillation was induced with a 50 Hz burst, appropriately detected by the device, and successfully defibrillated with a single 40 J shock following 14.4 seconds.

complications of pneumothorax, venous stenosis, endocarditis, and future intracardiac lead extraction, as well as allowing nonepicardial options in those with limited vascular access from congenital heart disease. Young patients facing multiple system changes over their lifespan, with an inherently higher risk of lead fracture, are ideal candidates for an S-ICD.

The early experience of S-ICDs in pediatric patients were marked with concerns for pulse generator erosion and need for reoperation.[3,4] Jarman and colleagues[3] described a series of 16 patients in which 3 required reoperation for threatened device erosion (2 patients) and wound dehiscence (1 patient). All 3 underwent successful generator repositioning to a more superior location with the ability to retain the original lead and generator. It was determined that a more superior location, towards the axilla, afforded greater tissue thickness, comfort, and protection in the smaller patients. Additionally, there has been a significant reduction in the thickness of the generator, currently comparable to a transvenous device.

In the current patient, screening was performed using the manufacturer-supplied morphology template and comparing primary, secondary, and alternate vectors in the left and right parasternal positions. Though an Automated Screening Template has been released by Boston Scientific (Marlborough, MA), it was not available at the time this patient presented. It is our practice to include the right parasternal position in all screenings, as its use has been demonstrated to result in improved R:T ratios in the primary vector.[5] Larger ratios have been associated with a lower incidence of inappropriate shocks. Our patient passed screening in the left secondary and right primary, secondary, and alternate vectors. By implanting utilizing the right primary and left secondary vector (Figure 1), we were able to achieve appropriate electrode separation and position the heart shadow between the coil and device generator. The alternate vector was no longer eligible, presumably because of the leftward position of the superior electrode.

The significant heart rate variability in pediatric patients and concern for QRS and T-wave changes at higher rates necessitates screening during exercise or at elevated heart rates. T-wave oversensing results in a significant percentage of inappropriate shocks, and exercise screening allows for appropriate vector selection or consideration of a transvenous device.[6] Our patient had initial motor delays as a result of her cardiac arrest and so an isoproterenol infusion was utilized to simulate exercise.

Once appropriate sensing vectors have been selected, only minor modifications to the standard implantation procedure are required, even for the smallest patients. As has been previously published for the implantation of transvenous cardiac devices, there is a trend among pediatric implanters to place the generator via an axillary incision.[7] This allows for a more cosmetic result and does not affect pacemaker or ICD performance. For the S-ICD, the typical generator incision is in the inframammary crease and extends from the fifth to sixth intercostal spaces in the midaxillary line, with the pocket made posteriorly from the incision. With an axillary S-ICD approach, care is taken beforehand to confirm that the planned position places the cardiac silhouette between the lead and generator, with no more than 50% of the cardiac silhouette being below the generator during deep inspiration.

For the current patient, the most notable modification was positioning the sensing lead into a “C-curve” in order to position the electrodes over the bony prominences of the inferior and superior sternum. The inferior electrode was placed to the right of the xyphoid and correlated with a primary sensing vector (electrode to generator) and the superior electrode was placed in a left parasternal position, correlating with a secondary sensing vector. With this atypical spacing, the alternate sensing vector (inferior to superior electrode) was not appropriate following implantation. Care should be taken to minimize the amount of soft tissue or adipose tissue between the electrode and bony structure, as this has been associated with variable sensing and inappropriate shocks (Boston Scientific internal communication).

Defibrillation testing is performed as routine following S-ICD implantation in our institution, though true threshold testing is not. The nominal shock strength is 80 J for permanent programming, though this can be adjusted during implant testing to perform defibrillation threshold testing with available outputs including 10 J, 20 J, 40 J, and 80 J. It is our practice to perform an initial defibrillation utilizing 40 J, but not to perform additional testing if this is successful. In a follow-up period of 18 months, our patient demonstrated appropriate sensing and device parameters, with no change in the device-selected sensing vectors. She was followed by Physical Therapy for postarrest delays but made a complete recovery with no residual deficits. Importantly, she has full range of motion in her left arm and shoulder, with no sensation of impingement or discomfort. The superior aspect of the lead is palpable as it crosses the sternum, though it is not visible and does not cause discomfort. Patient care has been transferred to another institution after the family moved with a plan to continue remote transmissions every 3 months and annual in-person interrogations. A change in sensing, either vector or morphology, may necessitate repeat defibrillation testing.

Conclusion

The S-ICD is a viable alternative to traditional transvenous devices, and our report suggests that it can be successfully implanted in pediatric patients of small size. Though modifications to the standard implantation procedure may be necessary, this technology could potentially be considered for the majority of eligible pediatric patients.

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