Implantation of the subcutaneous implantable cardioverter-defibrillator with retroperitoneal generator placement in a child with hypoplastic left heart syndrome

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Introduction
Implantable cardioverter-defibrillators (ICDs) for secondary prevention of sudden cardiac death are increasingly indicated in patients with congenital heart disease (CHD).1 Children with CHD present unique challenges for the implantation of traditional ICDs owing to both cardiac and venous anatomy and patient size. Novel ICD systems previously described for small children with CHD in whom a traditional ICD cannot be implanted include a generator inserted abdominally with a subcutaneous array electrode placed in the thorax.2 This technique required the placement of a transvenous or epicardial pacing and sensing lead. Others have used a transvenous design ICD lead placed on either the epicardium or subcutaneous tissue.1 In 2012, an entirely subcutaneous implantable cardioverter-defibrillator (S-ICD) was approved for use in the United States by the Food and Drug Administration after a decade of clinical testing.3 The S-ICD system (Cameron Health, Inc, San Clemente, CA) is composed of a pulse generator and a subcutaneous lead that functions as a sensing lead and a defibrillation coil. The pulse generator is typically inserted in a subcutaneous pocket over the left lateral chest wall, and the lead is positioned left of the sternal border. This system requires no chest entry and no venous access for sensing leads, making it ideally suited for patients with complex anatomy related to CHD, those with active infections, and young patients who will require the device for many years. To date, the S-ICD system has been implanted in the standard fashion in children as small as 34 kg.4 We describe the implantation of the S-ICD in a 3-year-old boy born with hypoplastic left heart syndrome (HLHS) for ventricular fibrillatory arrest.

Case report
This child was born with HLHS with intact atrial septum, requiring an atrial septostomy and atrial septal stent at birth. At 5 days of age, he underwent stage 1 palliation with the Norwood procedure and right ventricle to pulmonary artery (RV-PA) shunt. This was unfortunately complicated by a pulmonary artery pseudoaneurysm, requiring revision of the RV-PA shunt in his fifth week of life. Because of pulmonary artery hypoplasia with multilevel branch pulmonary stenosis, he underwent complex pulmonary arterioplasty before bidirectional cavopulmonary anastomosis. He has now undergone 4 sternotomies and is awaiting stage 3 palliation for HLHS.

A year after his bidirectional Glenn and while in his normal state of health, this patient complained of sudden onset of abdominal pain and became unresponsive. Cardiopulmonary resuscitation was initiated, and he was defibrillated for polymorphic ventricular tachycardia (VT) by first responders (Figure 1). He was wearing an event monitor at the time because of several unexplained syncopal episodes in the preceding 6 weeks. Rhythm strips from this event showed supraventricular tachycardia and sinus tachycardia with a premature ventricular contraction occurring on a T-wave–initiating polymorphic VT. An electrophysiology study showed no inducible supraventricular tachycardia or VT, and cardiac catheterization was nondiagnostic for a reversible cause of this event. He was, therefore, referred for an ICD.

Because of his altered venous anatomy after bidirectional Glenn and his size, implanting a transvenous ICD system was not possible. An epicardial ICD was considered, but we would have had to accept the risk of redo sternotomy or...
thoracotomy in the setting of 4 previous sternotomies. After considering the options, we elected to attempt the placement of an S-ICD.

Procedure

Careful consideration was given to appropriate placement of the generator and lead for this system. Because of the patient’s small size (13.5 kg), the recommended left thoracic subcutaneous placement of the generator was not feasible because it would have been less than the necessary 14 cm from the sensing electrodes on the lead (Figure 2). We determined that the least obtrusive location for the generator would likely be retroperitoneal in the left flank. To create adequate sensing and defibrillation vectors, the lead was best oriented vertically over the right chest just medial to the anterior axillary line. Before implantation the chest was mapped with surface leads and appropriate sensing was obtained at this location on the right chest.

A left subcostal incision was made 6 cm in length. After dividing the muscles of the abdominal wall, the peritoneum was reflected in a medial direction and a retroperitoneal pocket was created such that the device would be located in the iliac fossa and extend to the subcostal region. The disposable tunneling device was then used to create a subcutaneous tunnel for the lead from a right subcostal counter incision to the abdominal pocket. In a similar fashion, the lead was tunneled from the right subcostal incision to a third incision below the clavicle just medial to the anterior axillary line. The tip of the lead was fixed to the pectoral fascia at this infraclavicular location, and the midportion of the lead was fixed to the abdominal fascia in the right subcostal region. The lead was then attached to the generator, which was inserted into the retroperitoneal pocket. Each incision was then closed in multiple layers (Figure 3).

After implantation, 1 induction of ventricular fibrillation was performed with successful defibrillation using 80 J. Shock impedance was 81 Ω, and time to therapy was 19 seconds.

Discussion

To our knowledge, this is the first documented ventricular fibrillatory arrest in a patient with HLHS and history of an RV-PA shunt with stage I Norwood repair between stages II and III of palliation. Ventricular arrhythmias and sudden death as a consequence of ventriculotomy has been a critique of the Sano modification of the Norwood repair despite the 1-year survival benefit of the RV-PA shunt shown by the randomized multicenter single ventricle reconstruction trial. The arrhythmogenic focus in the case of an RV-PA shunt is thought to be the fibrosis and scar related to the right ventriculotomy, with a substrate similar to patients with tetralogy of Fallot, in whom ventricular arrhythmias and sudden death are clearly documented. Only 1 study has shown increased postoperative arrhythmias with the RV-PA shunt compared with the modified Blalock-Taussig shunt, but ventricular arrhythmias were rare in that study and there

Figure 1  Rhythm strip showing polymorphic ventricular tachycardia with return of normal sinus rhythm after defibrillation.

Figure 2  The patient’s chest demonstrating a sternal length shorter than the 14-cm proprietary mapping ruler for the subcutaneous implantable cardioverter-defibrillator.

Figure 3  Photograph of the device and abdominal pocket placement.
was no difference in rates of VT between the 2 shunt techniques. Similarly, documented instances of VT or ventricular fibrillation are low in the few studies that have reported arrhythmias in this patient population, making comparisons of these 2 shunt types and the association of VT/ventricular fibrillation difficult.9,10 Although the rate of interstage sudden cardiac death at home for HLHS has been shown to be as high as 42%, the effect of the RV-PA shunt was not studied in that series.11

As the RV-PA shunt is increasingly used in HLHS for stage I shunt, interstage and late ventricular arrhythmias in these patients may become more frequently observed. Thus, the need for ICD therapy would also be likely to increase. This case illustrates the complexity of delivering ICD therapy in patients with HLHS and other CHD palliated by the 3-stage approach, which ultimately reroutes the systemic venous return away from the heart and directly to the pulmonary arteries. Therefore, the transvenous approach to the heart is not an option. All the previously described approaches require an epicardial sensing electrode placed directly on a ventricle.7,2

Because our patient had undergone 4 previous sternotomies with the need for a fifth sternotomy for stage 3 palliation, the possibility of sternotomy or even a limited thoracotomy was considered high risk and undesirable for long-term procedural success. Knowing that epicardial systems carry the increased risk of failure,12,13 an epicardial approach, even with a subcutaneous array, could potentially lead to further sternotomies or thoracotomies. In addition to the advantage of extrathoracic implantation offered by this system, a single case series of S-ICD implantation in children and adolescents observed a lower rate of reoperation and inappropriate therapy with the S-ICD compared with transvenous ICD systems.14 For these reasons, the S-ICD was the best option for our patient.

However, the patient’s small size posed technical challenges that required deviation from the manufacturer’s recommended approach. The youngest patient previously described receiving the S-ICD was 10 years old and the lowest weight was 34 kg.14 Our patient was 13.5 kg with minimal subcutaneous fat. The risk of generator erosion was judged to be high, especially considering the dimensions of the generator, 69.9 cm$^3$ and 145 g, compared with the dimensions of a standard single-lead ICD system, 30.5 cm$^3$ and 72 g (Incepta E160, Boston Scientific, Dublin, Ireland). Ultimately, retroperitoneal generator placement, which is the standard approach of our team in small infants with epicardial pacemakers,15 was considered the lowest risk of generator erosion.

With the R-wave sensing based entirely on the vector of the 2 cutaneous electrodes to the generator, the potential for T-wave oversensing and inappropriate shocks with this atypical configuration was a significant concern. Therefore, before implantation, the potential vectors were re-created with skin electrodes in different location on the patient’s skin and with different heart rates and postures to mimic the vectors of implant. This was further limited by the 14-cm length of the coil, which ultimately required a position more lateral to the sternum than recommended, to provide a shocking vector from the coil to the generator and to avoid the sternum for future sternotomies.

**Conclusion**

We report a life-threatening ventricular arrhythmia in a 3-year-old boy weighing 13.5 kg with HLHS after RV-PA shunt creation, in whom the implantation of the S-ICD was successful with the placement of a retroperitoneal generator. The S-ICD is a promising option for patients with HLHS in whom transvenous endocardial systems are complicated by systemic venous discontinuity with the ventricle and in whom epicardial systems are complicated by multiple previous sternotomies and small patient size, although innovative configurations may be necessary.

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