Entirely Subcutaneous Implantable Defibrillator
Safest Option in a Young Girl With Ventricular Tachycardia and Ebstein Anomaly

Laura Cipolletta, MD; Mario Luzi, MD; Luca Piangerelli, MD; Federico Guerra, MD; Alessandro Capucci, MD

Case Report
A 16-year-old female patient with Ebstein anomaly was admitted to the hospital for 2 episodes of hemodynamically unstable ventricular tachycardia (VT; Figure 1). Two months earlier, she had undergone surgical tricuspid valve replacement with a biological prosthetic valve and cavopulmonary anastomosis (Figure 2). Both VT episodes were treated with external direct current cardioversion, and the patient was then referred to our center to attempt VT ablation. Unfortunately, the fact that the right ventricle (RV) was papyraceous and the free ventricular wall was thin would have exposed the patient to a high risk of catheter perforation. Because transvenous implantable cardioverter-defibrillator (ICD) implantation had the same risk of cardiac perforation, and access to the right cardiac chambers had been made unfeasible by cavopulmonary anastomosis, subcutaneous (S)-ICD implantation was considered. In assessing the patient’s adequacy for S-ICD implantation, the morphology of the QRS was screened to avoid T wave oversensing (Figure 3). Because of the bizarre morphology of the QRS attributable to severe dilatation of the RV, QRS screening was performed with the left arm electrode placed to the right side of the xiphoid process (Figure 4). Under local anesthesia and conscious sedation, the catheter was inserted subcutaneously from the pocket in the left midaxillary region to the right side of the xiphoid process (Figure 4). The tip was then advanced up to the manubrio-sternal junction, 1 cm to the right of the midsternal line. At the end of the procedure, a defibrillation test was performed: clinical VT was induced, correctly sensed, and treated after 12.5 seconds with a 65 J shock (conventional polarity configuration). On 2-month follow-up examination, the patient did not have any complications, and no significant events were recorded.

Discussion
To our knowledge, this is the first case of S-ICD implantation reported in a patient with Ebstein anomaly and cavopulmonary anastomosis. In congenital heart defect (CHD), the impaired RV might create an arrhythmogenic substrate. Traditional transvenous ICD implantation is associated with infectious complications and difficulty in maintaining long-term lead integrity. Implanting an S-ICD in patients with CHD may help to overcome most of the acute and chronic complications of transvenous ICD implantation. The S-ICD is better able to discriminate non-ventricular arrhythmias than most transvenous systems, and its rate of inappropriate shocks is lower. In our case, despite the bizarre morphology of the QRS and its repolarization, correct sensing was achieved. The peculiarity of this case is that the defibrillation catheter was placed to the right of the sternum because positioning the vector to the left of the sternum, as usually suggested, would have created a shock vector that covered only a small portion of the heart because of the severe dilatation of the right chambers. Positioning the catheter on the right side of the sternum provided sufficient coverage of the heart and yielded correct and effective sensing and treatment of ventricular arrhythmias (Figure 4).

Conclusions
S-ICD can be safely implanted in patients with CHD not requiring pacing in whom it is impossible to reach the right chambers. In the event of severe dilatation of the right chambers, placement of the S-ICD catheter along the right parasternal line can enable ventricular arrhythmias to be treated more correctly.

Disclosures
A. Capucci received a fee from Boston Scientific in 2013. The other authors report no conflicts.

References

Key Words: death, sudden, cardiac, ventricular, tachycardia.
Figure 1. A 12-lead ECG of hemodynamically unstable ventricular tachycardia.

Figure 2. Cardiac magnetic resonance. A, Sagittal image of the heart. B, Transversal image of the heart. AR indicates anterior right; FR, foot right; HL, head left; LV, left ventricle; MRN, magnetic resonance number; PL, posterior left; and RV, right ventricle.

Figure 3. Screening ECG at 10 mm/mV in sitting (A) and in supine (B) positions. ECG at both positions was considered adequate to predict a good vector/posture combination.

Figure 4. Chest x-ray film. A, Posterior-anterior projection: blue arrows show correct shock vector with catheter placed at the right side of the sternum. Yellow dotted arrows show shock vector with the catheter in the standard position (left parasternal line). B, Lateral projection.
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_Circ Arrhythm Electrophysiol._ 2014;7:358-359
doi: 10.1161/CIRCEP.113.001320

_Circulation: Arrhythmia and Electrophysiology_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 1941-3149. Online ISSN: 1941-3084

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