Implanted pacemaker and cardioverter-defibrillator in a patient with ectopia cordis

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Introduction

Ectopia cordis (EC) is a rare congenital cardiac malformation defined as complete or partial displacement of the heart outside the thoracic cavity. Its estimated prevalence is 5.5–7.9 per million live births and it is present in 0.1% of patients with congenital heart disease. Although the first case of EC was identified during the early 1600s, fewer than 100 cases have been reported in the literature,[1–3] all of which are focused on childhood management. Commonly associated congenital heart defects in EC include conotruncal anomalies, atrial septal defects, and ventricular septal defects. Cardiac surgery can be achieved in patients with EC, though it is associated with high mortality and significant long-term morbidity.[1] There is limited literature on cardiac surgery in EC, and there are no reported cases of patients with a transvenous pacemaker or implantable cardioverter-defibrillator (ICD).

We report the implantation of and appropriate therapy from a dual-chamber transvenous defibrillator in a patient with EC and repaired tetralogy of Fallot (TOF).

KEY TEACHING POINTS

- Implantable cardioverter-defibrillator implantation in children and patients with congenital heart disease can be complicated and limited by patient size, lack of vascular access, and abnormal cardiac anatomy. Each patient requires surgical planning to determine appropriate location of leads and generator in relation to the cardiac mass.
- Defibrillation testing should still be considered in patients with congenital heart disease, especially those with right-sided device implantations and atypical lead/shock vector orientations.
- Dual-coil leads should be considered in right-sided implants or in patients in whom the distal right ventricular coil sits more anteriorly than usual, owing to significant ventricular dilation or hypertrophy.

Case report

The subject is a 26-year-old woman with EC, TOF, and pulmonary atresia. Although the heart was located outside the thoracic cavity, it remained within the pericardial sac, which was completely covered by skin. No attempt was made to internalize the heart, and she was initially palliated with a left Blalock-Taussig shunt at 3 days of age. At 2 years of age, she underwent a right ventricle–to–pulmonary artery conduit and ventricular septal defect closure. She subsequently underwent 3 surgical conduit revisions, followed by a transcatheter Melody valve placement within the conduit at 18 years of age. Her heart remained in an extrathoracic location throughout (Figure 1A and B). The Melody valve was revised at 20 years of age owing to a stent fracture, and then an additional surgical conduit revision was required at 22 years of age. Due to a history of presyncope and nonsustained, monomorphic ventricular tachycardia, a surgical cryoablation was performed from the tricuspid annulus to the proximal conduit insertion at the time of her most recent conduit revision. She was discharged home on Dilantin postoperatively, as she had an oral aversion to mexiletine. In the postoperative setting, she had isolated ventricular premature beats and short runs of slow atrial tachycardia.

In the year after her surgery, she developed worsening heart failure requiring inpatient admissions for aggressive diuresis and recurrent hospitalizations. In the setting of hypokalemia, she had 3 separate episodes of polymorphic ventricular tachycardia requiring defibrillation. She was

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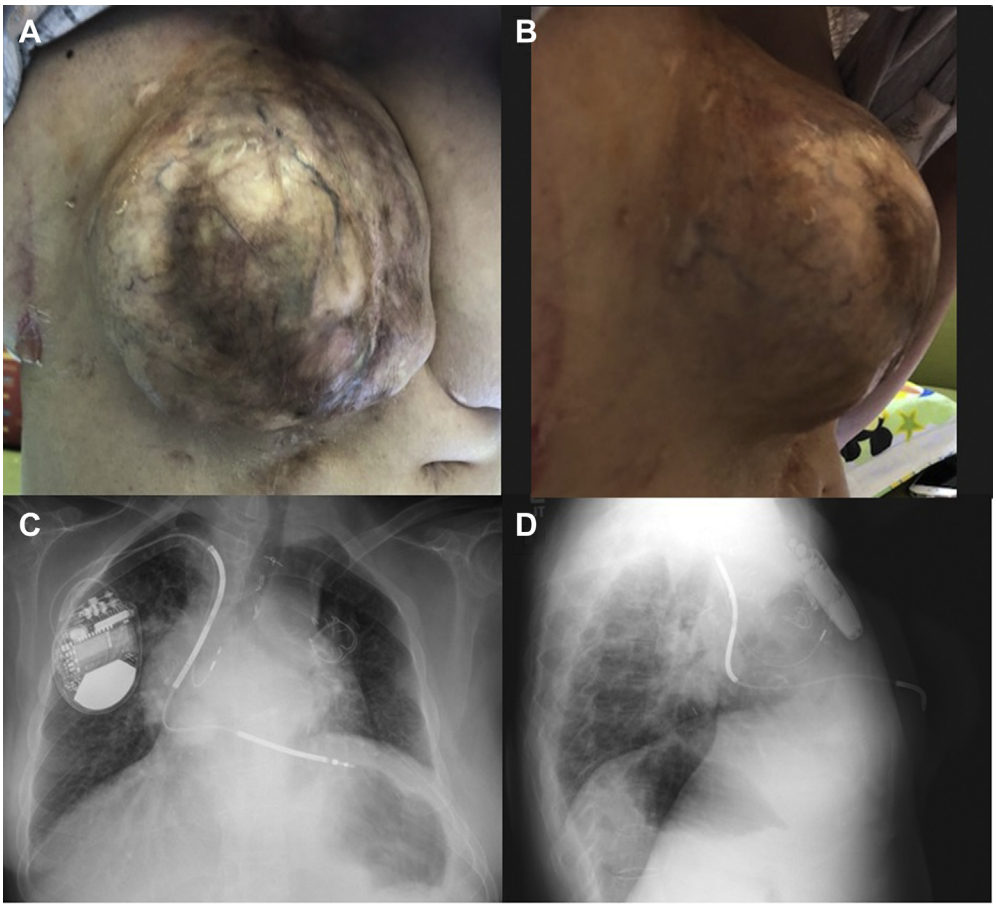

Figure 1 **A, B:** Anterior (**A**) and lateral (**B**) view of a woman with ectopia cordis and tetralogy of Fallot, requiring placement a right-sided transvenous dual-chamber implantable cardioverter-defibrillator (**C, D**).

subsequently transitioned to amiodarone, with improvement in her malignant arrhythmias. However, owing to worsening of her baseline sinus node dysfunction, an atrial pacemaker (Medtronic Advisa A2DR01, Medtronic 3830 49-cm Select Secure lead; Medtronic, Minneapolis, MN) was placed at 25 years of age. Because of the presence of bilateral superior vena cavae, the pacemaker was implanted on the right side.

The patient's overall health deteriorated in the subsequent year, with a new diagnosis of type II diabetes mellitus. In the setting of significant metabolic derangement and hypokalemia owing to inadequate diabetes control, she had a cardiac arrest requiring bystander cardiopulmonary resuscitation, defibrillation for ventricular fibrillation (VF), and mechanical support using extracorporeal membrane oxygenation. Given her tenuous status, and after an extensive multidisciplinary discussion, the decision was made to upgrade her pacemaker to a dual-chamber ICD.

At the time of implant, an angiogram was performed to delineate the right ventricular anatomy (Figure 2). The ICD lead (Medtronic 6947 Sprint Quattro Secure MRI SureScan 55-cm dual-coil lead; Medtronic) was placed in the midseptal right ventricle. Attempts to place the lead in a more apical location resulted in frequent lead dislodgement. After the lead was secured, the lead was attached to the ICD generator (Medtronic Evera DDMB1D4) and placed in a prepectoral pocket. Defibrillation testing was then performed. Despite the right-sided implant and distal coil located in an extracardiac location (as noted on the lateral radiograph, Figure 1D), the induced VF was successfully terminated with a 25 J shock

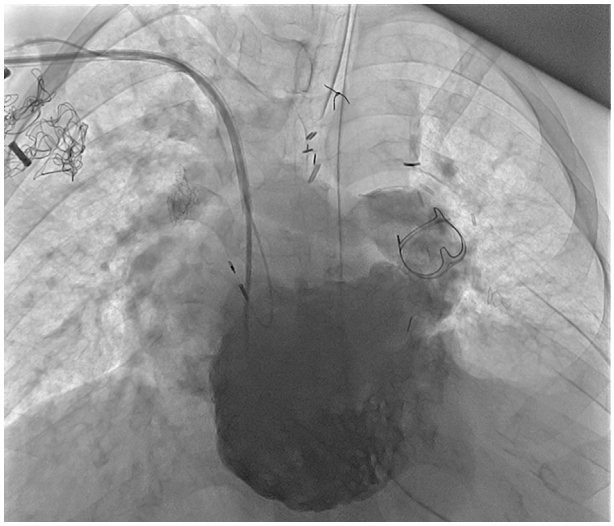

Figure 2 Angiogram of the right ventricle at the time of the ventricular lead placement.

from the device (programmed right ventricular coil to superior vena cava [SVC] coil/can).

Approximately 8 months later, she suffered an out-of-hospital arrest with a successful shock delivered from her device. Interrogation of the ICD following the episode revealed an appropriate 35 J shock for VF, followed by successful conversion to her baseline atrially paced rhythm. She was again noted to be hypokalemic at the time of her arrest, with a potassium of 2.8 mmol/L. She had a prolonged hospitalization with optimization of her medical therapy and has not had further malignant arrhythmias since that time.

Discussion

We report a case of a transvenous ICD in a patient with EC, who had successful conversion of VF both intraoperatively and clinically after implantation. To our knowledge, this is the first reported case in the literature of an ICD placed in a patient with EC. Defibrillation testing was performed owing to the novel extrathoracic location of the distal coil and right-sided implant, which ultimately reflected the ICD's ability to terminate a spontaneous episode of VF within a year of implant.

The use of ICDs in the pediatric and congenital population continues to grow.[4] Though lead-related complications and inappropriate ICD discharges are not uncommon, the cumulative beneficial effects of ICDs may be higher in young adults with congenital heart disease compared to those with acquired heart disease, owing to a younger age at implantation.[5] Patients with TOF constitute the largest subgroup of ICD recipients and comprise approximately half of all ICD implantations in congenital heart disease. In these patients, high rates of appropriate ICD therapies have been reported for both primary- and secondary-prevention indications, suggesting that ICDs may play an important role in the prevention of sudden death.[6]

ICD implantation in children and patients with congenital heart disease is complicated and often limited by patient size, lack of vascular access, anatomical obstruction to lead placement, or contraindication to transvenous implantation owing to residual intracardiac lesions. Furthermore, abnormal cardiac anatomy and positioning may require nonstandard sites for generator placement and vectors for ICD electrode positioning.[7] Each patient requires presurgical planning to determine the appropriate location of implanted coils in relation to the ICD generator. Given this, it is not surprising that electrophysiologists treating children and young adults with congenital heart disease are still more likely to perform defibrillation testing at the time of device implantation.[8]

This is in contrast to adults with structurally normal hearts, where randomized controlled trials have noted that, although safe, defibrillation testing may be unnecessary in the majority of ICD implants.[9,10] However, right-sided and subcutaneous ICDs were not included in these studies, with subsequent studies noting the importance of defibrillation testing in subcutaneous ICDs.[11] Studies on right-sided implants are lacking. Interactive simulations of defibrillation models[12] have noted that right-sided ICDs have higher defibrillation thresholds. Given this, defibrillation testing is routinely performed in all right-sided implants at our institution.

Our experience has also resulted in the use of dual-coil leads in some patients with right-sided implants, not only because of their historically lower defibrillation threshold, but also for the added benefit of the SVC coil to draw the shock vector posteriorly. Single-coil leads, especially in patients with significant dilated or hypertrophied ventricles, may pull the defibrillation vector anteriorly, away from the bulk of the myocardium, leading to concerns about effective defibrillation in certain anatomic configurations. However, the literature has shown no significant difference in first-shock efficacy and all-cause mortality between single-coil and dual-coil leads.[13] The presence of an SVC coil also results in a higher difficulty extracting the lead in the future.[14] Nonetheless, in select cases, as in this patient, the presence of a posteriorly located SVC coil may be desirable, as the lateral chest radiograph shows that it is most likely important in drawing the voltage gradient posteriorly in order to encompass the left ventricle.

In conclusion, we report, to our knowledge, the first implantation of an ICD in a patient with EC and repaired TOF. Despite the patient's unusual anatomy and subsequent atypical ICD lead position, with the distal coil located in an intracardiac but extrathoracic location, the ICD was successful in defibrillating the patient from VF induced at implant and clinically a year after implant. Defibrillation testing remains important in right-sided implants, congenital heart disease, and novel lead configurations.

References

1. Escobar-Diaz MC, Sunderji S, Tworetzky W, Moon-Grady AJ. The fetus with ectopia cordis: experience and expectations from two centers. Pediatr Cardiol 2017; 38:531–538.
2. Gabriel A, Donnelly J, Kuc A, et al. Ectopia cordis: a rare congenital anomaly. Clin Anat 2014;27:1193–1199.
3. Shad J, Budhwani K, Biswas R. Thoracic ectopia cordis. BMJ Case Rep 2012; 2012:bcr1120115241.
4. Czosek RJ, Meganathan K, Anderson JB, Knilans TK, Marino BS, Heaton PC. Cardiac rhythm devices in the pediatric population: utilization and complications. Heart Rhythm 2012;9:199–208.
5. Vehmeijer JT, Brouwer TF, Limpens J, et al. Implantable cardioverter-defibrillators in adults with congenital heart disease: a systematic review and meta-analysis. Eur Heart J 2016;37:1439–1448.
6. Khairy P, Harris L, Landzberg MJ, et al. Implantable cardioverter-defibrillators in tetralogy of Fallot. Circulation 2008;117:363–370.
7. Stephenson EA, Batra AS, Knilans TK, et al. A multicenter experience with novel implantable cardioverter defibrillator configurations in the pediatric and congenital heart disease population. J Cardiovasc Electrophysiol 2006;17:41–46.
8. Baysa SJ, Olen M, Kanter RJ, Fishberger SB. Defibrillation testing strategies of pediatric and adult congenital electrophysiologists during ICD implantation. Pacing Clin Electrophysiol 2016;39:843–847.
9. Bansch D, Bonnemeier H, Brandt J, et al. Intra-operative defibrillation testing and clinical shock efficacy in patients with implantable cardioverter-defibrillators: the NORDIC ICD randomized clinical trial. Eur Heart J 2015; 36:2500–2507.
10. Healey JS, Hohnloser SH, Glikson M, et al. Cardioverter defibrillator implantation without induction of ventricular fibrillation: a single-blind, non-inferiority, randomised controlled trial (SIMPLE). Lancet 2015; 385:785–791.

11. le Polain de Waroux JB, Ploux S, Mondoly P, et al. Defibrillation testing is mandatory in patients with subcutaneous implantable cardioverter-defibrillator to confirm appropriate ventricular fibrillation detection. Heart Rhythm 2018; 15:642–650.
12. Jolley M, Stinstra J, Pieper S, et al. A computer modeling tool for comparing novel ICD electrode orientations in children and adults. Heart Rhythm 2008;5:565–572.
13. Sunderland N, Kaura A, Murgatroyd F, Dhillon P, Scott PA. Outcomes with single-coil versus dual-coil implantable cardioverter defibrillators: a meta-analysis. Europace 2018;20:e21–e29.
14. Epstein LM, Love CJ, Wilkoff BL, et al. Superior vena cava defibrillator coils make transvenous lead extraction more challenging and riskier. J Am Coll Cardiol 2013;61:987–989.