Case Report

Epicardial implantation of a transvenous implantable cardioverter-defibrillator system in Ebstein's malformation with tricuspid valve repair

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ABSTRACT

In this case report, we describe the implant of a transvenous implantable cardioverter-defibrillator lead on the left ventricular epicardial surface of a young patient with Ebstein's anomaly, facilitating effective defibrillation and anti-tachycardia pacing. The difficulties of conventional placement in Ebstein's malformation as well as the technique and outcome of epicardial lead implantation of a transvenous defibrillator-pacer system are discussed.

<Learning objective: This case report highlights the possible placement of a conventional transvenous defibrillator lead on alternative site such as epicardial surface as a solution in patients with Ebstein's anomaly undergoing open heart surgery. This case report also discusses technical challenges, which precluded the transvenous implantation route in a post-operative patient with Ebstein's anomaly. The advantages of epicardial versus subcutaneous system are also discussed.>

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Introduction

Sudden cardiac death is a recognized cause of late mortality in patients with congenital heart disease despite successful corrective surgery early in life. The role of implantable cardioverter-defibrillators (ICD) has become increasingly important as a component of care in patients with congenital heart disease to prevent sudden cardiac death [1]. However, implanting ICDs in congenital heart patients may require creative solutions due to various technical challenges [1–4]. In Ebstein's malformation, the morphological abnormalities limit the placement of endocardial lead in the functional right ventricle. The difficulties of conventional lead placement in Ebstein's malformation and the effectiveness of a transvenous ICD implanted on an alternative site such as the epicardial surface is still not clear.

Case report

A 17-year-old (58 kg) college student presented following an out-of-hospital cardiac arrest and successful resuscitation with an

automatic external defibrillator. Prior to this, he complained of tiredness and shortness of breath on minimal exercise. He was taking no medications, including over the counter and recreational drugs. On examination he was acyanotic with saturations of 95% in room air. The first and second heart sounds were accompanied by a grade 2/6 systolic murmur at the left sternal edge. There were no signs of cardiac failure. Electrocardiogram showed sinus rhythm with right-axis deviation and wide QRS right bundle branch block. The PR interval and QT interval were normal and there was no obvious pre-excitation seen.

An echocardiogram was consistent with severe Ebstein's malformation; there was displacement of the septal leaflet of the tricuspid valve by approximately 5 cm in the right ventricular cavity. In addition, there was mild to moderate low-velocity tricuspid regurgitation. There was no accompanying right ventricular outflow obstruction. Left ventricular systolic function was well preserved.

The patient was known to the adult congenital service, and had been waiting for elective tricuspid valve repair and cavopulmonary shunt prior to his cardiac arrest. Following his cardiac arrest, his body temperature was cooled for 24 h and the patient was subsequently extubated with intact neurology.

An electro-physiological (EP) study was performed, which ruled out pre-excitation; however, monomorphic ventricular tachycardia (VT) was easily inducible from programmed stimulation in the

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right ventricle. The morphology of the VT suggested a left ventricular exit site, which was easily pace-terminated during the EP study.

His presentation merited an ICD therapy as a prevention for sudden cardiac death, but several issues were raised. Firstly, in view of his need for a tricuspid valve repair in the future, a conventional transvenous system which crossed the tricuspid valve would be best avoided. Secondly, he may need a 1.5-ventricle by creation of a bidirectional cavopulmonary anastomosis to offload the right ventricle (RV) after tricuspid valve repair, in which case, a transvenous system would not be possible. A subcutaneous ICD system would have been a viable alternative, but this would have sacrificed the ability to treat any future VTs via anti-tachycardia pacing, and it is worth recalling that the induced VT during his EP study was readily pace-terminated.

We hypothesized that fixing a transvenous lead on the epicardial surface at the time of his surgery might offer the best compromise. However, we were unsure whether the shock-coils of the ICD lead implanted on the epicardial surface would be as effective at defibrillating the heart.

A review of the literature did not reveal any report of the effectiveness of this approach, so we decided to attempt the implant with on-table defibrillation testing.

At surgery, the tricuspid valve was repaired using the cone technique. The annulus and atrialized portion of the RV were reduced taking care not to ensnare the coronary arteries. The atrial septal defect was closed. The superior vena cava was divided and joined to the right pulmonary artery to create a bidirectional cavopulmonary shunt. A single-chamber Medtronic Protecta XT VR (ICD-VVI; Medtronic, Minneapolis, MN, USA) defibrillator-pacer unit was implanted as follows: (i) the defibrillator coil lead (Medtronic Transvene® 6937 single SVC coil) was sutured on the epicardial surface of the left ventricular free wall adjacent to the obtuse marginal branch (Fig. 1). Therefore, the ICD coil lead lied in the pericardial cavity posteriorly; (ii) the V pace/sense lead (Medtronic CapSure Epi® 4968-bipolar 35 cm) was placed on the apical position of the LV epicardial surface; (iii) a sub-pectoral pocket was then created in the left infra-clavicular area and the leads were tunneled under the pectoral muscles to connect to the defibrillator-pacer unit. The cardiopulmonary bypass was then weaned off uneventfully. The defibrillator was tested and functioned satisfactorily. Sensed R wave was 22.5 mV, with a pacing threshold of 2.5 V at 0.5 ms. Epicardial pacing lead impedance was measured at 949 Ohms and shock impedance at 49 Ohms. Ventricular fibrillation was induced using

Fig. 2. Post-operative chest X-ray showed the position of the defibrillator coil lead on the left ventricular free wall as well as the pacing leads at the apical position of the left ventricle.

50 Hz frequency, and the device successfully defibrillated the rhythm back to sinus at 35 J, after a first shock of 25 J failed to terminate ventricular fibrillation.

Post-operatively, his recovery and discharge were uneventful (Fig. 2). The post-operative transthoracic echocardiography demonstrated an excellent tricuspid valve repair. At one-month follow-up, an ICD check showed an R-wave of 16 mV, ventricular threshold of 0.75 V at 0.4 ms, ventricular pacing impedance of 893 Ohms, and shock impedance of 54 Ohms, and no events had been detected or treated. He remained clinically well at follow-up and echocardiography showed an excellent result from cone reconstruction with a satisfactory ICD-pacemaker check at approximately 18 months post-surgery.

Discussion

The implantation of an ICD on epicardial surface can be considered when there is a venous access problem or in small children, where conventional approaches would be difficult [4]. This case highlighted the challenges in implanting a defibrillator unit in a conventional way in Ebstein's anomaly. In this malformation, the true RV cavity is small due to atrialized portion. Additionally, the septal and posterior leaflets are adherent to the RV endocardium. These morphological abnormalities limit the placement of endocardial lead in the functional RV. These limitations will not be overcome by simply plicating the atrialized ventricle or reduction annuloplasty. The adherent leaflets have to be extensively mobilized as in cone reconstruction technique to increase the functional RV area. However, this involved extensive surgery on the tricuspid valve and placement of a defibrillator lead across the reconstructed valve is not ideal. The placement of lead across this newly reconstructed valve could also induce or aggravate tricuspid regurgitation, which would be detrimental for RV function. Furthermore, the decision to create a superior cavopulmonary shunt would preclude any transvenous lead placement post-operatively.

The morphology of the VT suggested a left ventricular exit site, highlighting the fact that ventricular architecture may be abnormal

Fig. 1. The defibrillator coil lead was sutured on the epicardial surface of the left ventricle free wall adjacent to the obtuse marginal branch.

throughout the heart in Ebstein's anomaly, despite the condition being considered as an "RV problem". The positioning of the shock-coil lead was decided as a way of including a significant part of the LV in the shock vector; we chose this, as the clinical VT was originating from the LV. In hindsight, perhaps other orientations and positions of the shock-coil lead should have been attempted in an attempt to optimize the defibrillation threshold and aim for values under 25 J; however, we were also keen to achieve a good pacing threshold as we want the device to be able to retain its overdrive pacing function. When the defibrillation thresholds were discussed with the electrophysiologist, it was felt that the device offered safety, even though the lack of the 10 J safety margin in the threshold is a compromise; furthermore, threshold may improve after chest closure. In future, we would recommend that perhaps several positions across both the left ventricle and RV should be tried to ensure the best option.

As previously discussed, a subcutaneous ICD system would be an alternative strategy, but this system does not allow the delivery of anti-tachycardia pacing, meaning that all arrhythmias would be treated with shocks as a first-line therapy [1]. Furthermore, discriminating a true VT from sinus tachycardia would have been problematic in the presence of wide right-bundle branch morphology in our patient's resting electrocardiogram. Consequently, exercise-induced sinus tachycardia may falsely trigger inappropriate shocks in a young and active student.

This case report highlights the possible placement of a conventional transvenous defibrillator lead on an alternative site such as epicardial surface as an elegant solution to these specific issues with a good functional result.

Conflict of interests

None declared.

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