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Implantation of a completely right sided subcutaneous cardioverter-defibrillator in a patient with situs inversus dextrocardia

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ABSTRACT

Dextrocardia is a congenital anomaly where the heart is abnormally located in the right hemithorax. In these patients, the implementation of transvenous implantable cardioverter-defibrillator (TV-ICD) can be technically challenging and pose a higher risk of complications than the general population. We present the case of a male patient that was successfully submitted to right-sided implantation of subcutaneous ICD (S-ICD) as an alternative to transvenous ICD (TV-ICD) for primary prevention of sudden cardiac death. This option is not only feasible but may potentially be ideal for these patients, as it circumvents challenges and potential complications of TV-ICD insertion.

1. Introduction

Dextrocardia is a congenital anomaly where the heart is abnormally located in the right hemithorax with the apex positioned towards the caudal and rightward orientation. Although there is no association between dextrocardia and heart failure, some patients may develop left ventricular systolic dysfunction. Just as the general population, these patients are likely to benefit from implantable cardioverter-defibrillator (ICD) for primary or secondary prevention of sudden cardiac death [1]. When patients with dextrocardia qualify for device therapy, placement of transvenous leads can be technically challenging and may prove time-consuming. We present the case of a male patient that was successfully implanted with subcutaneous ICD (S-ICD) as an alternative to transvenous ICD (TV-ICD) for primary prevention of sudden cardiac death.

1.1. Case report

A 68-year-old man with dextrocardia with situs inversus, coronary artery disease, and ischemic dilated cardiomyopathy with a left ventricular ejection fraction of 25% presented for device-based therapy consideration on an outpatient basis after six months of guideline-directed medical therapy. Physical examination was remarkable for normal heart sounds over the right hemithorax, and the absence of heart sounds over the lateral aspect of the left hemithorax. A 12-lead electrocardiogram demonstrated sinus rhythm with negative P waves in the lead I, positive P wave in lead aVR, and an abnormal precordial progression; the QRS width was 114 msec. These findings were consistent with dextrocardia. Given the patient had no indication for atrial or ventricular pacing, nor cardiac resynchronization therapy, the options of TV-ICD vs. S-ICD were discussed with the patient. He opted for a subcutaneous system. An electrocardiographic screening was performed by placing the electrodes in the same manner as per the manufacturer's instructions but on the right hemithorax. The patient successfully passed supine sitting and standing screening position required for ICD placement. A subcutaneous defibrillator (Boston Scientific Emblem MRI S-ICD A219) was successfully implanted on the right hemithorax several weeks after the initial encounter. The device pocket was located at the level of the sixth intercostal space on the lateral aspect of the right hemithorax (Fig. 1). The

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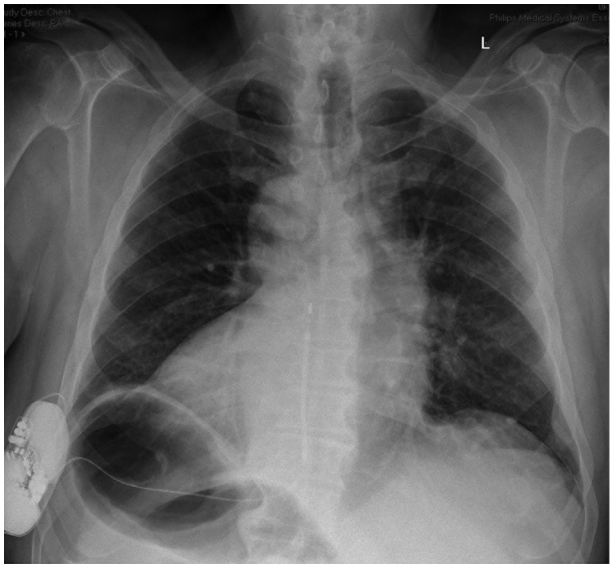

Fig. 1. Chest x-ray posteroanterior view showing dextrocardia situs inversus with S-ICD and electrodes in place.

defibrillation electrode was positioned 2-cm from the Xyphoid process and 1-cm to lateral and parallel to the right sternal border. Defibrillation threshold was performed by delivering a 65 J discharge after induction of ventricular fibrillation upon completion of device implantation. A single shock successfully terminated tachycardia and reestablished sinus rhythm. The patient has not experienced any device-related complications nor received any device therapy after twelve months of follow-up. Electrode impedance and sensing are normal, showing no significant changes from time of implantation.

2. Discussion

Subcutaneous implantable cardioverter-defibrillator devices have seen an increase in clinical use since its FDA approval in 2012. The advantages of S-ICD over TV-ICD have been well described and include no risk of pneumothorax or cardiac perforation, no radiation exposure, lower incidence of systemic infections, lower risk of lead-related complications such as lead fracture, and lower morbidity associated with lead extraction. Another advantage is that although infection rates are higher in patients with S-ICD, these tend to be localized to subcutaneous tissue, and usually respond to antibiotic therapy, rarely ever requiring device extraction. The S-ICD shares similar indications with transvenous device counterparts, but it is not the best option for patients who indicate anti-bradycardia pacing, cardiac resynchronization therapy, or documented monomorphic ventricular tachycardia, which may respond to anti-tachycardia pacing (ATP). S-ICDs have been shown to be effective in terminating ventricular tachycardia or fibrillation with first shock success rates similar to TV-ICD systems [2].

S-ICDs were initially reserved for patients' inadequate venous access and those who are prone to bloodstream infections. However, as operators gained experience with this system, S-ICDs began to be used in other patient populations. Some investigators contend that S-ICDs may be the ideal choice for patients with complex congenital heart disease [1].

In this case, we present a patient with dextrocardia and situs inversus who qualified for cardioverter-defibrillator therapy for primary prevention of sudden cardiac death. Although implantation of a TV-ICD in such a patient is feasible, insertion of transvenous leads in dextrocardia is associated with some technical challenges due to the distorted anatomy [3]. Fluoroscopic images may be difficult to interpret. Previous experience with pacemaker implantation in patients with dextrocardia suggests the use of inverted fluoroscopic images to simulate normal anatomy and overcome some of these challenges. Some authors argue in favor of employing intraoperative angiography, computed tomography, or magnetic resonance imaging to delineate anatomy and relationship of cardiac chambers, anticipating other possible coexisting anatomic anomalies, and assisting the operator in placing the lead. Dextrocardia is commonly associated with variations in venous circulation, for which angiogram may be necessary before transvenous device implantation. These factors would require additional testing, use of contrast, and could potentially result in longer procedure times. Also, transvenous leads are exposed to added stress due to the angle generated by the abnormal course of the superior vena cava relative to the reversed position of the right atrium and ventricle in patients with dextrocardia [4].

Ceresnak et al. [1] described the use of S-ICD in an adult patient with dextrocardia and tetralogy of Fallot. Our case adds to the still scarce body of evidence regarding the feasibility of S-ICD as an alternative to TV-ICD in patients with dextrocardia, as few reports describe the use of S-ICD in unusual positions such as we described. To ensure appropriate device function, it is vital that the system can accurately sense intracardiac signals. Appropriate electrocardiographic (ECG) screening needs to be performed prior S-ICD placement to determine ideal device and coil location, and thus achieve optimal sensing. In this case, we used a mirror image of the conventional ECG screening method. Chan et al. [5] described a successful right-sided screening in a patient who failed initial conventional screening. However, there is scant data regarding right-sided ECG screening for S-ICD in patients with dextrocardia. At the time of implantation, we confirmed appropriate device sensing in its primary vector, which further supports the use of right-sided ECG screening in such patients.

Finally, for a defibrillator shock to be effective, a critical mass of myocardium must be captured by it. For this reason, S-ICD lead position affects the defibrillation threshold. Defibrillation threshold testing was successful in our patient with a single shock at the usual 65 J setting, as recommended by the manufacturer. This confirms a right-sided subcutaneous lead position results in a suitable electrical vector for effective defibrillation.

Current practice guidelines establish the S-ICD as a reasonable option to TV-ICD in patients who meet indication of an ICD for primary or secondary prevention of sudden cardiac death. Given the potential challenges and complications of implantation of transvenous systems in patients with dextrocardia, along with the demonstrated feasibility of S-ICD in this population, we maintain that S-ICD should be considered the device of choice in these patients. However, studies with larger dextrocardia patient populations and long-term follow-up may be needed before the efficacy of S-ICD can be established in patients with this condition.

3. Conclusion

In patients with dextrocardia, right-sided implantation of an S-ICD is a feasible alternative to TV-ICD. This option circumvents technical challenges of TV-ICD insertion, carries a lower risk of procedural complications, and appears to be effective in this population. Although further studies and experience with this technique are needed, S-ICD may potentially be the ideal option for patients with dextrocardia who qualify for cardioverter-defibrillator therapy for primary or secondary prevention of sudden cardiac death.

Conflict of interest declaration

The author declares that the views expressed in the submitted article are his own. There is no private funding or conflict of interest to disclose.

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