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Epicardial cardioverter-defibrillator implantation in a 4-month-old infant bridged to heart transplantation

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Abstract

Implantable cardioverter-defibrillator (ICD) is the gold standard therapy for the prevention of sudden cardiac death. Nevertheless, ICD placement in the paediatric population is still limited because of several technical difficulties. Several implantation techniques have been proposed but experience in infants with very low weight and less than 6 months is very limited. We herein describe a case of a minimally invasive ICD epicardial implantation in a 4-month-old infant weighing 5 kg. A diagnosis of arrhythmic cardiomyopathy with left ventricular non-compaction disease with ventricular tachycardia storms, QT prolongation and Wolff-Parkinson-White pattern was made. Antiarrhythmic drugs, radiofrequency ablation and sympathetic denervation were not effective. ICD implantation was successful allowing the infant to survive and bridging to heart transplantation.

Keywords: Congenital heart disease • Implantable cardioverter-defibrillator (ICD) • Paediatric cardiomyopathy • Electrical storms • Heart transplantation

CASE REPORT

An infant was transferred to our institution because of electrocardiographic abnormalities and recurrent syncope. Electrocardiogram monitoring showed torsades de pointes-like ventricular tachycardia, QT prolongation and a Wolf-Parkinson-White pattern.

The subsequent echocardiography showed a moderately impaired left ventricular ejection function (40%), with evidence of a non-compaction cardiomyopathy.

Trials with antiarrhythmic medical therapy (metoprolol, amiodarone, mexiletine, lidocaine and flecainide) were administered with different dosages and routes without being effective. Ablation therapy and sympathetic denervation also failed.

Because of recurring ventricular malignant storms and despite maximum tolerated antiarrhythmic therapy, an implantable cardioverter-defibrillator (ICD) implantation was advised.

Occlusion of the femoral, subclavian and left jugular vein due to the indwelling catheters made the epicardial implantation the only choice left.

The child was 4-month-old weighing 5 kg at the time of the surgery.

A partial inferior median sternotomy with a short subxiphoid extension was made. Bipolar pace sensing electrode (Medtronic, Minneapolis, Minnesota) was sutured to the epicardium of the right ventricular anterior wall.

A defibrillation coil was subcutaneously tunnelled according to the patient's chest shape, reaching the vertebral column through multiple small cutaneous incisions.

The ICD device (Medtronic Protecta XT) was then inserted into an abdominal pocket in the right iliac fossa. Finally, the leads were connected to the ICD device (Fig. 1). Sensing and pacing thresholds were determined and a 20-J ICD shock was effective to interrupt an induced ventricular tachycardia.

In the early postoperative course, no lead-related complications were observed.

Defibrillation threshold was 20 J at 1 month (child weight 6 kg), 40 J at 6 months (child weight 7 kg) and no more threshold adjustments were needed due to the retarded growth of the severely ill infant.

In the following months, recurrent episodes of ventricular tachycardia were detected and ICD shocks were effective at terminating them. Generator change was required at 7 months after the surgery as a result of the depletion of the battery. The infant showed a good psychomotor development until cardiac transplantation was performed 1 year after ICD implantation. Both electrical device and coils were removed at the time of transplantation.

DISCUSSION

Experience of ICD implantation in paediatric age, particularly in small body sized patients, is limited. A multidisciplinary discussion with surgeons, paediatric cardiac electrophysiologist and heart transplant specialist is needed: device therapy in toddlers is a challenging field because the myriad of devices available are

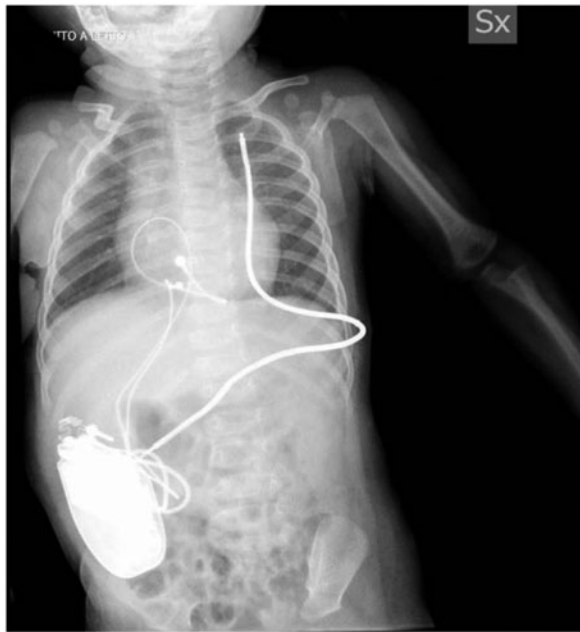


Figure 1: Chest X-ray (frontal projection). The implantable cardioverter-defibrillator was implanted in the abdominal pocket, the subcutaneous defibrillation coil was placed along the left chest wall and the epicardial leads were connected to the right ventricle.

not designed specifically for this kind of patients and there is a scarcity of prospective studies to guide best practice.

In the last years, technological developments have increased the use of cardiac defibrillator in infants and several implantation techniques have been proposed [1].

Initially, epicardial patches via thoracotomy were used, but they were associated with pericardial inflammation and high defibrillation threshold. Transvenous ICD implantation has always been difficult in small patients and not possible in case of anomalies of the venous system or single-ventricle physiology.

In 2001, Thogersen *et al.* [2] described for the first time an ICD implantation with an extracardiac system via a midline sternotomy in a 4-month-old infant using a transvenous subcutaneously lead.

Kriebel *et al.* [3] reported the largest series of ICD implantation using extracardiac technique by a partial sternotomy and a left

lateral subaxillary incision in 8 patients; among them, only 1 patient was 4-month-old (weight: 4.4 kg).

Silver *et al.* [4] described a case of ICD placement at 1 month (3.6 kg) via a median sternotomy; ICD was located in a pocket in the preperitoneal space with atrial and ventricular leads, while shocking coils were located in the left pleural space.

Briant *et al.* [5] described the case of a 4.9-kg, 5-week-old infant boy who presented with cardiopulmonary arrest and underwent an epicardial cardioverter-defibrillator implantation performed via a median sternotomy. We described a case of an extracardiac ICD placement in a 4-month-old baby weighing 5 kg with malignant ventricular arrhythmias and severe occlusions of the major veins. The implantation was realized via a partial inferior sternotomy and small cutaneous incisions. No lead-related complications were reported.

CONCLUSION

This report shows the feasibility and the safety of a minimally invasive ICD implantation technique in a paediatric patient with low weight and a complex arrhythmic cardiomyopathy.

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