ICD Implantation in Infants and Small Children: The Extracardiac Technique

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Background: There is no clear methodology for implantation of an internal cardioverter-defibrillator (ICD) in infants and small children. The aim of this study was to assess efficacy and safety of an extracardiac ICD implantation technique in pediatric patients.

Patients and Methods: An extracardiac ICD system was implanted in eight patients (age: 0.3–8 years; body weight: 4–29 kg). Under fluoroscopic guidance a defibrillator lead was tunneled subcutaneously starting from the anterior axillar line along the course of the 6th rib until almost reaching the vertebral column. After a partial inferior sternotomy, bipolar steroid-eluting sensing and pacing leads were sutured to the atrial wall (n = 2) and to the anterior wall of the right ventricle (n = 8). The ICD device was implanted as “active can” in the upper abdomen. Sensing, pacing, and defibrillation thresholds (DFTs) as well as impedances were verified intraoperatively and 3 months later, respectively.

Results: In seven of eight patients, intraoperative DFT between subcutaneous lead and device was <15 J. In the eighth patient ICD implantation was technically not feasible due to a DFT >20 J. During follow-up (mean 14.5 months) appropriate and effective ICD discharges were noted in two patients. DFT remained stable after 3 months in four of six patients retested. A revision was required in one patient due to lead migration and in another patient due to a lead break.

Conclusions: In infants and small children, extracardiac ICD implantation was technically feasible. Experience and follow-up are still limited. The course of the DFT is unknown, facing further growth of the patients. (PACE 2006; 29:1319–1325)

Introduction

Since Mirowski et al.1 reported the first use of an implantable cardioverter defibrillator (ICD) in humans in 1980, these devices have become the gold standard therapy for prevention of sudden arrhythmogenic cardiac death in adult patients. Children with various types of cardiomyopathy, primary electrical diseases, and after surgical repair of congenital heart defects are at risk for sudden arrhythmic death.2,3 However, up to now, there was no clear methodology for the implantation of an ICD in infants and small children. Particularly, infants pose a technical challenge due to body size, physical activity, and growth which may result in multiple surgical procedures in order to adjust electrode positions. Despite technological progress, implantation of transvenous ICDs is not recommended in infants due to the small vessel size and the diameter and length of the electrodes currently available. In addition, the ICD shocking coil may straddle the tricuspid valve and may result in significant tricuspid valve insufficiency. Epicardial patch electrodes, often used in the past, were associated with extensive trauma and unfavorable defibrillation thresholds (DFTs). Accordingly, several implantation techniques have been applied in infants and small children, but experience is limited due to small sample size.4,5,6 Just recently, a multicenter study7 reported the largest experience concerning different ICD implantation techniques without a transvenous shocking coil or
epicardial patches in pediatric patients thus far. In that study various types of configurations with a small number of patients per configuration were described.

The present prospective study was performed in order to evaluate efficacy and safety of an extracardiac ICD implantation technique using a defibrillation lead subcutaneously together with bipolar epicardial sensing and pacing electrodes and an abdominally placed ICD device in infants and small children. This study represents the largest series so far using this unique implantation technique.

Patients and Methods

Patients

From July 2004 to October 2005, eight consecutive pediatric patients (four boys and four girls) referred to the three institutions for ICD implantation were enrolled into the study. The criteria to implant an ICD using the extracardiac technique were referred to the three institutions for ICD implantation of pediatric patients (four boys and four girls). This study represents the largest series so far using this unique implantation technique.

Surgical Technique

Under general anesthesia a short left lateral subaxillary incision was performed at the level of the 6th rib. Under fluoroscopic guidance a metal mandrin covered by a plastic sheath was pre-shaped according to the chest of the individual patient and tunneled subcutaneously along the course of the 4th–6th rib posteriorly and superiorly in a subscapular fashion until almost reaching the vertebral column. After withdrawal of the metal mandrin a defibrillator lead (Medtronic Transvene 6937* SN, 35 or 52 cm, respectively, 6.9-Fr single Superior Vena Cava [SVC] defibrillation coil with a coil length of 8 cm, approved for use in the SVC without any fixation, currently not available in the United States; Medtronic Inc., Minneapolis, MN, USA) was introduced through the plastic sheath which was removed subsequently (Fig. 1).

After a partial median inferior sternotomy, bipolar steroid-eluting sensing and pacing leads (Capsure Epi 4968; 25 cm or 35 cm, respectively; Medtronic Inc.) were sutured to the anterior wall of the right ventricle in all patients (Fig. 1) and to the right atrial appendage in two patients (patients 1 and 2, Table I). Sensing and pacing thresholds were determined.

Subsequently, the ICD device (Marquis DR 7274 or Marquis VR 7230, Medtronic Inc.) was inserted into an abdominal pocket behind the right rectus abdominis muscle. Finally, the leads were connected to the ICD device. Ventricular fibrillation was induced by high-frequency bursts or T-wave shocks to determine the individual DFT using the defibrillation electrode as the cathode and the “active can” device as the anode. The general guideline in all institutions was to achieve a DFT <20 J. Therefore, the DFT was determined starting with 10 J. If ventricular fibrillation was sufficiently terminated by two successful defibrillations, no further testing was performed. Otherwise energy output was increased to 15 J. If 15 J was not successful in terminating ventricular fibrillation, the position of the electrode was modified appropriately starting with a new position 1–2 cm below or above the previous location. No further testing was performed when the DFT was ≤10 J. The first shock energy was programmed at the maximum output of the device.

Postoperative Management

After the surgical procedure, patients were transferred to the intensive care unit for at least 24 hours and monitored for at least 5 additional days in the hospital as standard of care in the participating institutions after sternotomy. All patients received three doses of cephazolin (100 mg/kg/d) as perioperative antibiotic prophylaxis. A surface electrocardiogram, a 24-hour Holter monitor, and a 2D echocardiographic study for exclusion of pericardial effusion were obtained before discharge. In addition, chest x-rays in the posterior-anterior and lateral projections were performed to delineate lead and device positions. PACe thresholds and ventricular sensing were again measured prior to discharge. Antiarrhythmic medication was continued as before.
### Table I.
Data of the ICD Testing

<table>
<thead>
<tr>
<th>Pt. no.</th>
<th>Age (Years)</th>
<th>BW (kg)</th>
<th>Diagnosis/Indication</th>
<th>Intraoperatively</th>
<th>3 Month Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>S (mV)</td>
<td>PT (V/ms)</td>
</tr>
<tr>
<td>1</td>
<td>0.3</td>
<td>4.4</td>
<td>AVSD, VT/VF, cAVB</td>
<td>A: 2.5</td>
<td>2.0 V/0.5 ms</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>V: 8.8</td>
<td>2.5 V/0.5 ms</td>
</tr>
<tr>
<td>2</td>
<td>0.5</td>
<td>5.5</td>
<td>LQTS, WPW</td>
<td>A: 8.6</td>
<td>0.4 V/0.5 ms</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>V: 5.8</td>
<td>0.5 V/0.5 ms</td>
</tr>
<tr>
<td>3*</td>
<td>2</td>
<td>15</td>
<td>LQTS</td>
<td>V: 6.7</td>
<td>0.8 V/0.5 ms</td>
</tr>
<tr>
<td>4</td>
<td>3</td>
<td>13</td>
<td>Taussig-Bing complex, VF after MI</td>
<td>V: 10</td>
<td>1.0 V/0.4 ms</td>
</tr>
<tr>
<td>5</td>
<td>3</td>
<td>13</td>
<td>LQTS</td>
<td>V: 6.3</td>
<td>1.0 V/0.8 ms</td>
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<tr>
<td>6*</td>
<td>5</td>
<td>20</td>
<td>JLNS</td>
<td>V: 11</td>
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<td>7</td>
<td>6</td>
<td>27</td>
<td>LQTS</td>
<td>V: 5.1</td>
<td>0.7 V/0.5 ms</td>
</tr>
<tr>
<td>8</td>
<td>8</td>
<td>29</td>
<td>Catecholaminergic VT</td>
<td>V: 4.5</td>
<td>2.0 V/0.4 ms</td>
</tr>
</tbody>
</table>

A = atrial; cAVB = complete atrioventricular block; AVSD = atrioventricular septal defect; DFT = defibrillation threshold; HVI = high voltage impedance of the defibrillation lead; LQTS = long QT syndrome; MI = myocardial infarction; ms = milliseconds; PT = pacing threshold; S = sensing; V = ventricular; VF = ventricular fibrillation; VT = ventricular tachycardia; WPW = Wolff-Parkinson-White syndrome.

*In patient 3 fracture and in patient 6 dislocation of the defibrillation lead were noted requiring surgical revision.
Figure 1. Schematic drawing of the extracardiac technique illustrating the ICD device (1) placed in the upper right abdomen, the subcutaneously introduced transvenous lead in the back (2), and bipolar steroid-eluting sensing and pacing leads sutured to the right ventricle (3). Energy is delivered between the defibrillation lead as cathode and the “active can” device as anode (see inserted transversal plane at the right lower corner).

During follow-up, patients were seen on a regular basis by their referring pediatric cardiologist.

Results

Data of atrial/ventricular sensing, pacing and DFTs, and impedance of the defibrillation leads intraoperatively and at 3-month follow-up in the individual patients are listed in Table I.

ICD Implantation

In seven of the eight patients, the extracardiac ICD implantation was successful with an intraoperative DFT between subcutaneous lead and device of <15 J (Fig. 2). In the eighth patient (patient 8, Table I) no DFT <20 J despite multiple positions of the subcutaneous defibrillation lead and the device could be achieved intraoperatively. Subsequently, a transvenous system was implanted using a thin 6-Fr endocardial lead (Sprint fidelis 6949, 58 cm, Medtronic Inc.). The ICD was repositioned in a left pectoral pocket. In the configuration right ventricular coil versus device the DFT was >15 J. Therefore to increase the safety margin, an additional subscapular subcutaneous defibrillation electrode was implanted. Finally, using the configuration right ventricular coil versus subcutaneous lead, the DFT was <10 J intraoperatively and 7 days later at repeat testing.

In two patients a dual-chamber ICD system was implanted. In patient 1 (Table I) a dual-chamber ICD system was implanted due to postoperative complete atrioventricular (AV) block after surgical repair of an atrioventricular septal defect. In patient 2 (Table I) additional sensing and pacing leads were attached to the right atrium due to recurrent episodes of supraventricular tachycardia based on Wolf-Parkinson-White syndrome. During preoperative electrophysiological study, multiple accessory pathways were ablated. In this patient, long QT syndrome diagnosis was established according to recurrent episodes of Torsades de pointes tachycardia. In this patient, the early postoperative course was complicated by inadequate feeding which was probably caused by the size of the ICD device in the right upper abdomen. After adjusting the feeding to slow volumes and higher feeding rates this problem finally resolved. In addition, Mexiletine needed to be added to propranolol in this patient postoperatively due to transient electrical storm related to the long QT syndrome with repeated appropriate ICD shocks. Subsequently, ventricular ectopy stopped and no further sustained ventricular tachycardia or ICD discharges were noted as verified by the ICD.

No other acute complications related to the ICD implantation were noted in any of the patients. Particularly, no major bleeding or penetration of the pleural space was observed during the subcutaneous tunneling of the metal mandrin.

Follow-Up

After 3 months the DFT was determined in six of the seven patients with an initial successful extracardiac ICD implantation. Data on follow-up DFT are lacking in patient 1 (Table I), as the parents refused retesting. In four of the six patients studied, the DFT remained stable. In patient 6 initial DFT was <15 J. After 3 months DFT had increased to <20 J (Table I) due to a shift of the subcutaneous defibrillation lead to the lateral chest wall (Fig. 3). DFT testing was repeated after another 3 months. At this time, a further movement of the defibrillation lead was noted and the DFT had increased to >20 J. Therefore, revision of the defibrillation lead was performed resulting in a DFT <10 J. Due to the lead migration in this patient, additional sutures were placed at the tip of the defibrillation lead at the back and around the lead at the lateral incision during the revision in this patient and in all following procedures. No further movement of the
defibrillation lead was noted during the 12-month follow-up after revision in patient 6 or in any of the other patients.

In patient 3 (Table I) alarm of the device occurred after physical exercise. On admission, electrode impedance of the shocking coil had increased to >200 Ω and chest x-ray showed a fracture of the subcutaneous defibrillation lead (Fig. 4). This lead was revised the next day using the same electrode position resulting in a DFT <15 J. This patient is scheduled for retesting after another 3 months. In both procedures lead extraction was performed without any difficulties. No further lead fracture was noted in any of the patients during routinely performed chest x-rays.

After a mean follow-up of 14.5 (3–20) months, appropriate and successful ICD discharges were noted in two patients (Table I) requiring modification of the antiarrhythmic treatment. No inappropriate discharges were observed.

Figure 2. Chest x-ray (left panel: posterior-anterior projection; right panel: lateral projection) of a 6-year-old girl (patient 7; Table I) demonstrating the extracardiac technique. The ICD was placed abdominally, the subcutaneous defibrillation lead is positioned posteriorly along the left chest wall in a subscapular fashion, and the epicardial bipolar sensing and pacing leads are connected on the right ventricle.

Figure 3. Chest x-rays of a 5-year-old boy (patient 6, Table 1) at time of ICD implantation (left panel) and 3 months postoperatively (right panel). A shift of the subcutaneous defibrillation lead to the lateral chest wall is evident which resulted in a significant increase of the DFT.
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Discussion

Our data support the feasibility of using a transvenous defibrillation lead in a subcutaneous position in infants and small children. Due to low incidence of sudden cardiac death in patients under the age of 20 years only a minor portion of ICD implantations is performed in this population.2–4 In infants and small children, experience is restricted to a few case reports10–12 and one multicenter study8 including 15 patients <8 years. Initially, ICD implantation in small children was performed using epicardial patch electrodes and an abdominally placed ICD device. The approach was associated with a high incidence of complications resulting in significant early and late morbidity and often unfavorable DFTs.4,7,13,14 Within the last years implantation of transvenous ICD systems became favorable even for pediatric patients as small as 12 kg.15 However, in smaller patients this technique has often been shown to be associated with venous occlusion, lead malfunction, and difficult lead removal.16 In addition, in pediatric patients with univentricular circulation this technique is not applicable due to lack of a transvenous access to the subpulmonary ventricular myocardium. Therefore, no clear methodology for the implantation of ICDs especially in small children and infants has been established yet.

A minimally invasive approach using subcutaneous finger electrodes has been reported resulting in acceptable DFTs and a low complication rate. Cannon et al.6 described the surgical placement of an ICD coil directly into the pericardial sac in six patients with limited venous access to the heart. Thogersen et al.20 reported for the first time ICD implantation of an extracardiac system using a transvenous lead subcutaneously in the back in a 9-week-old infant. A multicenter study by Stephenson et al.8 reported the experience concerning different ICD implantation techniques without a transvenous shocking coil or epicardial patches in pediatric patients including the extracardiac system in two patients.

In the present study we used a comparable technique in our pediatric patients representing the largest series of ICD implantation using this extracardiac technique. In seven of our eight patients ICD implantation using a defibrillation lead subcutaneously was successful with adequate DFTs intraoperatively. Further, clinical efficacy was demonstrated in the present study in two patients who received several appropriate and successful discharges during a limited follow-up.

It may be speculated that the shorter defibrillation coil of the transvenous electrode when compared to the longer electrodes of the subcutaneous arrays allows to establish a sufficient electrical field for defibrillation after individual positioning especially in the tiny chest of infants and small children. However, in the multicenter study by Stephenson et al.8 the mean DFT at implant was comparable with 15.5 J using various types of ICD implantation configurations. The DFTs of the present study and of others8 using a subcutaneous defibrillation lead are slightly higher compared to the DFTs achieved using endocardial systems (mean 11.5 J) in older children.5 However, the DFTs in the present approach are not completely comparable to other studies.

In one patient (patient 8, Table I) the extracardiac technique failed due to a high DFT. Of the 14 patients reported by Stephenson et al.8 two patients had a high DFT using a subcutaneous configuration. One patient with a structurally normal heart was changed to a high-energy device, the second patient with congenital heart disease remained in the hospital and was transplanted 2 months following ICD implantation. The reason for failure of the extracardiac technique in our patient may be related to the patient’s weight as this patient was the heaviest patient in the present series. Berul et al.18 have shown in an animal model that the DFT increases with weight when using subcutaneous arrays. However, in this patient a standard transvenous system also failed to achieve...
an acceptable DFT and therefore the reason for the high DFT remains unclear at the present time. The additional use of a subcutaneous defibrillation lead finally resulted in an acceptable DFT.21,22 Due to the initial high DFT using the extracardiac technique a different safety margin of <15 J was defined. Using two transvenous electrodes subcutaneously might have been an alternative approach in this patient.

In general, lead fracture and migration after ICD implantation remains a serious problem in pediatric patients. In 27 older children with a total of 38 endocardial ICD implantations two dislodgments and two lead fractures were noted.5 Stephenson et al.8 reported on the migration of the subcutaneous defibrillation lead in one patient. In the present study, using the extracardiac technique technical complications including lead fracture and lead dislodgment resulting in an increase of the DFT with the need of surgical revision was noted in two patients. This problem highlights the need of a timely follow-up of these patients. From our learning curve during the present study, based on a small series, we recommend the additional fixation of the defibrillation electrode to prevent lead migration.

Limitations
Number of patients and follow-up of the present study are still limited. Further experience is needed to assess the role of the extracardiac technique in our pediatric patient population. Furthermore, the course of the DFT is unknown facing further growth of the patients.

Conclusions and Future Prospects
This series of patients demonstrates that this technique with an implantation of an extracardiac ICD system using a transvenous lead subcutaneously is feasible in infants and small children. An extended thoracotomy and the insertion of leads in the vascular system could be completely avoided. This technique may be particularly useful in children with complex congenital heart defects and no vascular access to the subpulmonary ventricular myocardium.

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