Journal of Interventional Cardiac Electrophysiology (2019) 54:151–159
https://doi.org/10.1007/s10840-018-0451-y

Efficacy and safety of non-transvenous cardioverter defibrillators in infants and young children

Matthias J. Müller[1] · Jana K. Dieks[1] · David Backhoff[1] · Heike E. Schneider[1] · Wolfgang Ruschewski[2] · Theodor Tirilomis[2] · Thomas Paul[1] · Ulrich Krause[1]

Received: 25 June 2018 / Accepted: 7 September 2018 / Published online: 25 September 2018

Abstract

Background Implantable cardioverter defibrillators (ICD) protect from sudden cardiac death (SCD). In infants and young children, ICD implantation and programming is challenging due to small body size, elevated heart rates, and high physical activity.

Purpose We report our experience applying a non-transvenous ICD (NT-ICD) system to infants and children < 12 years of age and < 45-kg body weight.

Methods Between 07/2004 and 07/2016, NT-ICD had been implanted in 36 patients. Nine out of 36 patients (25%) had NT-ICD implantation for primary and 27/36 (75%) for secondary prevention. Underlying diseases included inherited primary electrical arrhythmogenic diseases ($n = 26$; 72%), cardiomyopathies ($n = 8$; 22%), and congenital heart defects ($n = 2$; 6%). The median (interquartile range) age at implantation was 6 (1.9–8.4) years, and the median body weight was 21.7 (11.2–26.8) kg. Three different NT-ICD implantation techniques had been applied over time: (1) abdominal device/subcutaneous shock coil, (2) abdominal device/pleural shock coil, and (3) subcardiac device/pleural shock coil.

Results During median follow-up of 5.2 (2.7–7.2) years, appropriate ICD discharges were documented in 12 (33.3%) and inappropriate shocks in 4 patients (11.1%). In 12/36 individuals (33.3%), a total of 25 surgical revisions were required due to NT-ICD malfunction. Eighteen out of 25 (72%) surgical revisions were necessary in patients with subcutaneous shock coil/abdominal device position. Surgical revisions (3/25, 12%) were significantly reduced ($p < 0.001$) after modifying the implantation technique to subcardiac device/pleural shock coil.

Conclusions NT-ICD was safe and effective in infants and young children. Appropriate ICD discharges occurred in a considerable number of patients. After modifying the implantation technique, the need for surgical revision could significantly be decreased.

Keywords Non-transvenous cardioverter defibrillator · Infants · Children · Defibrillation threshold · ICD discharge

✉ Matthias J. Müller
matthias.mueller@med.uni-goettingen.de

Jana K. Dieks
jana.dieks@med.uni-goettingen.de

David Backhoff
david.backhoff@med.uni-goettingen.de

Heike E. Schneider
heike.schneider@med.uni-goettingen.de

Wolfgang Ruschewski
wrusch@med.uni-goettingen.de

Theodor Tirilomis
theodor.tirilomis@med.uni-goettingen.de

Thomas Paul
tpaul@gwdg.de

Ulrich Krause
ukrause1@gwdg.de

[1] Department of Pediatric Cardiology and Intensive Care, University Medical Center, Georg August University Goettingen, Robert-Koch-Str. 40, 37075 Göttingen, Germany

[2] Department of Thoracic, Heart, and Cardiovascular Surgery, University Medical Center, Georg August University Goettingen, Robert-Koch-Str. 40, 37075 Göttingen, Germany

1 Introduction

Sudden cardiac death (SCD) is a rare event in infants and young children with an incidence of approximately 1–4 deaths/100,000 population/year. Underlying cardiovascular diseases responsible for SCD include electrical arrhythmogenic diseases, cardiomyopathies, myocarditis, coronary abnormalities, and structural congenital heart disease (CHD) [1]. Implantable cardioverter defibrillators (ICD) are life-saving in patients with increased risk for SCD due to life-threatening ventricular tachyarrhythmias [16]. In the recent past, the total number of ICD implants has increased 3-fold in patients < 18 years of age [6, 8]. ICD implantation in young patients and CHD patients requires special considerations: (1) body size, as leads of transvenous ICD systems may cause vessel obstruction and relevant tricuspid regurgitation [3, 18]. It is standard of care to refrain from implanting a subcutaneous ICD in children weighing less than 25–30 kg due to the size of the device [5]. (2) Congenital heart defects and vascular abnormalities may limit venous access to the heart as in patients with single ventricle physiology after hemi-Fontan and Fontan-type palliation. Residual intracardiac shunts carry an increased risk of thromboembolic events [19]. (3) Rapid heart rates in infants and children and/or atrial tachyarrhythmias require special attention as well as individual programming [12]. (4) ICD systems implanted in pediatric patients have to withstand high levels of physical activity [17]. Despite continuously improved ICD technology, an ICD system designed to meet the special requirements of small pediatric patients with or without CHD is unlikely to be developed. Facing the special requirements and limitations in pediatric patients, several alternative ICD implantation techniques have been developed [4, 5, 9, 13, 15, 18, 19, 21–23]. It is uncertain which of these ICD systems will be considered ideal. In order to address this gap in knowledge, we present more than one decade of experience using a completely non-transvenous ICD (NT-ICD) in pediatric patients with and without CHD.

2 Patients and methods

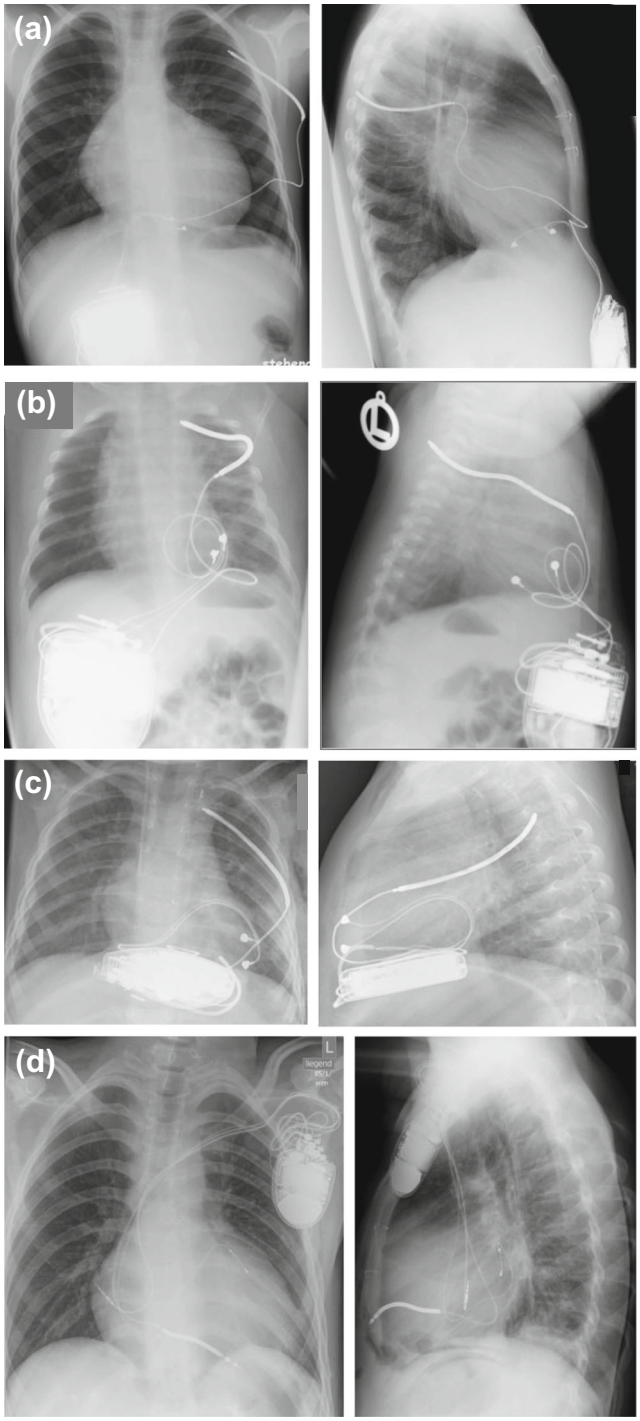
All patients < 12 years of age and < 45-kg body weight who had a NT-ICD implanted at our institution between 07/2004 and 07/2016 were included into the study. Demographic data were obtained as well as surgical technique, type of ICD generator, NT-ICD parameters including defibrillation threshold testing (DFT), pacing threshold of the pace/sense leads, and impedance of the shock coil as well as of the pace/sense electrodes. Appropriate and inappropriate ICD discharges were recorded. Inappropriate shocks were defined as shocks delivered for other reasons than VT or ventricular fibrillation (VF). Complications attributable to the NT-ICD and surgical revisions due to system malfunction during follow-up were also analyzed. The study was approved by the institutional review board and fully complies with the Declaration of Helsinki.

2.1 Surgical procedure

All surgical procedures were performed under general anesthesia and by experienced pediatric and congenital cardiovascular surgeons (WR, TT). From 07/2004 to 03/2006, NT-ICD was implanted in a subcutaneous coil/abdominal device configuration as previously described [13, 14]. In brief, bipolar ventricular (and atrial if indicated) steroid-eluting epimyocardial pace/sense electrodes were placed at the anterior right ventricular wall and at the right atrial appendage if appropriate. The shock coil lead was introduced and fixed subcutaneously along the course of the left third-sixth rib at the area close to the proximal part of the coil. The ICD device was placed in the upper abdomen behind the right rectus abdominis muscle and was fixed to the fascia of the muscle with non-absorbable suture (Fig. 1a). Since 04/2006, the shock coil was implanted and fixed in the left pleural space along the third-sixth rib as described by Bauersfeld et al. [4] (pleural shock coil/abdominal device, Fig. 1b). Since 09/2008, device implantation was further modified according to Bauersfeld et al. and Tomaske et al. [4, 21]. Now, the device was implanted in a subcardiac, extrapericardial position below the right ventricle and fixed to the pericardium (pleural shock coil/subcardiac device, Fig. 1c). The shock coil lead was placed in the left pleural space posteriorly along the third-sixth intercostal space and secured with a stitch close to the proximal end of the coil. Likewise, since 09/2008, bipolar steroid-eluting epimyocardial pace/sense electrodes were sutured onto the lateral left ventricular wall and onto the left atrial appendage in dual-chamber devices. Then, the leads were connected to the device. At the conclusion of surgery, defibrillation threshold testing (DFT) was performed in the operating room in all patients. VF was induced either by T wave shock or high-frequency stimulation (50 Hz). A DFT ≤ 15 Joule (J) was considered sufficient to obtain a double-safety margin for defibrillation. A DFT > 15 J at the conclusion of surgery required either device or shock coil reposition for improvement of the electrical field between coil and device.

2.2 NT-ICD programming

Initial pacemaker settings were VVI or DDD with a lower rate limit of 40 to 80 bpm depending on patient's age at implantation and individual pacing requirements. VT detection was inactivated in all patients and VF detection was set to 240–300 ms. The initial detection was programmed at 24–30 cycles whereas redetection was set at 9–12 cycles. Antitachycardia pacing (ATP) during charging was programmed in all patients. The first shock energy was set 10 J above individual DFT. Subsequent five shocks were set at maximum device output of 35 J.



◀ **Fig. 1** **a** Posterior-anterior and lateral chest X-rays after NT-ICD implantation (8-year-old boy with Taussig-Bing complex, weight 18.5 kg, height 119 cm) applying the subcutaneous coil/abdominal device technique. The shock coil was fixed subcutaneously along the posterior course of the left rib. The ICD generator was positioned in the upper abdomen behind the right musculus rectus abdominis. Bipolar epicardial pace/sense electrodes were sutured to the anterior right ventricular wall. **b** Posterior-anterior and lateral chest X-rays after NT-ICD implantation applying the pleural coil/abdominal device technique (2-month-old boy with hypertrophic cardiomyopathy, weight 5.6 kg, height 60 cm). The shock coil was placed in the posterior pleural space along the course of the ribs. **c** Posterior-anterior and lateral chest X-rays after NT-ICD implantation (9-month-old girl with long QT syndrome, weight 9.7 kg, height 83 cm) applying the pleural coil/subcardiac device. The ICD device has been positioned in a horizontal extra-pericardial position below the right ventricle with fixation of the device to the pericardium. **d** Posterior-anterior and lateral chest X-rays (15-year-old boy, with Taussig-Bing complex, weight 46 kg, height 167 cm) 11 years after NT-ICD implantation (the same patient as in Fig. 1a). The NT-ICD was completely removed and a transvenous ICD (CRT-D) was implanted

2.3 Postsurgical care

All patients were admitted to the PCICU postoperatively. Repeat DFT was performed prior to discharge. A safety margin ≥ 15 J to maximum device output was accepted to be sufficient. In addition, biplane chest X-ray, a 2D echocardiogram, and a 12-lead ECG were obtained prior to discharge.

2.4 Follow-up

After discharge from hospital, all patients were regularly followed at our outpatient clinic or by their referring pediatric cardiologist. After 3 and every 12 months thereafter, patients were scheduled for routine DFT as described above. In cases of DFT increase ≥ 10 J, a biplane chest X-ray was obtained to assess geometry of the NT-ICD and the DFT interval was shortened from 12 to 6 months. In case of a safety margin <15 J to maximal device output or lead and/or device dislodgement on chest X-ray, surgical revision of the generator position or the shock coil or both was performed. Indication for switch to a transvenous ICD system was body weight ≥ 45 kg and end of battery capacity and/or any technical NT-ICD problems requiring surgical revision.

2.5 Complications

Complications were defined as any incident requiring diagnostic and/or therapeutic intervention beyond standard of care. ICD generator exchange due to end of battery capacity was not considered as complication.

2.6 Statistics

Statistical analysis was performed using SPSS® 24.0 software (IBM, New York, USA). Numerical data are presented as median (interquartile range; IQR) unless otherwise stated. For comparison of categorical data, chi-square test was used. The Kaplan-Meier method was used to assess freedom from re-operation after NT-ICD implantation. A p value < 0.05 was considered significant.

3 Results

3.1 Patients

NT-ICD was implanted in 36 pediatric patients ($n = 14$ female, 38.9%). Seven out of 36 patients (19.4%) received a subcutaneous shock coil/abdominal device position, 5/36 (13.9%) a pleural shock coil fixation/abdominal device position, and 24/36 (66.7%) a pleural shock coil/subcardiac device fixation. The median age at implantation was 6.0 (IQR 1.9–8.4; range 0.1–11.6) years, median body weight 21.7 (IQR 11.2–26.8; range 4.6–42) kg, and median body height 118.5 (IQR 88.8–130; range 51–155) cm. NT-ICD was implanted for primary prevention of SCD in 9 (25%) and for secondary prevention in the remaining 27 (75%) patients. Table 1 provides detailed patient characteristics at time of NT-ICD implantation. Electrical arrhythmogenic diseases (long QT syndrome $n = 16$; Brugada syndrome $n = 6$; catecholaminergic polymorphic ventricular tachycardia $n = 4$) were the most frequent underlying diagnoses ($n = 26$, 72%), followed by cardiomyopathies (total $n = 8$, 22%; hypertrophic cardiomyopathy, $n = 6$; dilated cardiomyopathy, $n = 1$; arrhythmogenic right ventricular cardiomyopathy, $n = 1$) and congenital heart defects in two patients (6%; Taussig-Bing complex, $n = 1$; interrupted aortic arch associated with atrial and ventricular septal defect, $n = 1$).

3.2 ICD systems and parameters

The following ICD generators were implanted: EnTrust™ $n = 2$, Evera™ $n = 6$, Marquis™ $n = 4$, Protecta™ $n = 13$, Secura™ $n = 7$, and Virtuoso™ $n = 4$ (all Medtronic Inc., Minneapolis, MN, USA). Used pace/sense electrodes were CapSure Epi® 4968; 25–35 cm and shock coils Transvene® SVC 6937; 35–58 cm (all Medtronic Inc., Minneapolis, MN, USA). During the study period, NT-ICD implantation was successful except in an 8-year-old boy (weight 32 kg) with CPVT whose intraoperative DFT was > 20 J despite repeated attempts to optimize the shock field. Therefore, an additional transvenous shock coil (Sprint Fidelis® 6449, 58 cm; Medtronic Inc., Minneapolis, MN, USA) was implanted resulting in a DFT ≤ 10 J.

Single-chamber systems were used in 26 (72.2%) and dual-chamber systems in 10 (27.8%) individuals. Median intrinsic P wave amplitude at implantation was 3.9 (IQR 2.8–4.0) mV; median intrinsic R wave amplitude, 11.5 (IQR 7.0–16.7) mV; median atrial pacing threshold, 1.1 (IQR 0.8–2.4) V @ 0.4 (0.4–0.4) ms; median ventricular pacing threshold, 1.9 (IQR 1.0–3.0) V @ 0.5 (IQR 0.4–0.8) ms; and median DFT 10 (IQR 10–10) J. Shock coil impedance differed significantly between patients with subcutaneous shock coil ($n = 7$, median 74; IQR, 70–80 Ω) and those with pleural shock coil (pleural shock coil/abdominal device, $n = 5$, median 58; IQR 58–69 Ω, $p < 0.001$, pleural shock coil/subcardiac device, $n = 24$, median 52; IQR 46–58 Ω, $p = 0.01$) position. Table 2 gives a

Table 1 Detailed patient characteristics ($n = 36$) at time of NT-ICD implantation

Patient at NT-ICD implantation		All patients ($n = 36$)	Subcutaneous shock coil/abdominal device position ($n = 7$)	Pleural shock coil/abdominal device position ($n = 5$)	Pleural shock coil/subcardiac device fixation (n = 24)	p value subcutaneous shock coil/abdominal device vs. Pleural shock coil/subcardiac device and pleural shock coil/abdominal device vs. pleural shock coil/subcardiac device
Female gender	n (%)	14 (38.9)	3 (42.9)	2 (40.0)	9 (37.5)	$p = 0.56/p = 0.644$
Age (years)	median	6.0	2.8	6.3	6.1	$p = 0.216$ / $p = 0.518$
	(IQR)	(1.9–8.4)	(2.2–6.0)	(0.6–8.1)	(2.0–9.0)	
	(range)	(0.1–11.6)	(0.5–8.8)	(0.1–8.5)	(0.5–11.6)	
Body weight (kilogram)	median	21.7	20.4	18.0	22.9	$p = 0.872$ / $p = 0.556$
	(IQR)	(11.2–26.8)	(11.9–27.2)	(7.9–27.5)	(11.5–27.5)	
	(range)	(4.6–42)	(5.5–32)	(5.4–33)	(4.6–42)	
Body height (cm)	median	118.5	115.0	117.0	122.0	$p = 0.444$ / $p = 0.594$
	(IQR)	(88.8–130.0)	(91–122)	(71.0–132.0)	(89.0–132.0)	
	(range)	(51–155)	(61–139)	(61–141)	(51–155)	
Follow-up interval (years)	median	5.2	9.8	7.3	4.1	$p = 0.005$ / $p = 0.007$
	(IQR)	(2.7–7.2)	(4.7–11.3)	(5.8–7.7)	(1.3–5.5)	
	(range)	(0.1–11.5)	(1.7–11.5)	(5.4–7.8)	(0.1–8.1)	
SCD prevention						
Primary	n (%)	9 (25.0)	0	1 (20)	8 (33.3)	$p = 0.193$ / $p = 0.193$
Secondary	n (%)	27 (75)	7 (100)	4 (80)	16 (66.7)	

detailed overview on baseline parameters at the time of NT-ICD implantation.

3.3 ICD discharges

During the median follow-up of 5.2 (IQR 2.7–7.2, range 0.1–11.5) years, a total of 121 ICD therapies including ATP and inappropriate ICD discharges were delivered. Appropriate ICD discharges were noted in 12/36 (33.3%) patients (primary prevention $n = 1$, secondary prevention $n = 11$) with a total of 77 shocks delivered. Four out of 36 individuals (11.1%, primary prevention $n = 1$, secondary prevention $n = 3$) experienced a total of 44 inappropriate shocks. Reasons for inappropriate ICD shocks were rapidly conducted atrial tachycardia ($n = 2$), far-field sensing of external alternating current ($n = 1$), and lead fracture ($n = 1$). Forty out of 44 inappropriate shocks were delivered due to lead fracture of a subcutaneous shock electrode in one patient. Three out of four patients with inappropriate shocks also received appropriate shock therapy (Table 2).

3.4 DFT

A total of 302 regular DFT were performed. A significant DFT increase necessitating surgical revision was detected in 13/302 (4.3%) tests in a total of 10 patients (27%). None of the patients with significant DFT increase had evidence for lead failure during NT-ICD interrogation prior to DFT. There were no complications related to DFT.

3.5 Complications and system survival

In 12/36 (33.3%) patients, a total of 25 surgical revisions were required due to NT-ICD malfunction (Table 3). Majority of surgical revisions (18/25, 72%) were needed in those seven patients with subcutaneous shock coil/abdominal device configuration. Reasons for revision included dislocation (7/25 revisions, 28%) and fracture (3/25 revisions, 12%) of the subcutaneous shock coil, dislocation of the abdominal device (5/25 revisions, 20%), and malfunction of the epimyocardial pace/sense leads (3/25 revisions, 12%), respectively. Due to

Table 2 Baseline NT-ICD parameters at the time of NT-ICD implantation ($n = 36$) and shock delivery during follow-up

NT-ICD characteristics at implantation		All NT-ICD ($n = 36$)	Subcutaneous shock coil/abdominal device position ($n = 7$)	Pleural shock coil/abdominal device position ($n = 5$)	Pleural shock coil/subcardiac device fixation ($n = 24$)	p value subcutaneous shock coil/abdominal device vs. pleural shock coil/subcardiac device and pleural shock coil/abdominal device vs. pleural shock coil/subcardiac device
Single-chamber system	n (%)	26 (72.2)	5 (71.4)	5 (100)	16 (66.7)	$p = 0.318$
Dual-chamber system	n (%)	10 (27.8)	2 (28.6)	0 (0)	8 (33.3)	
Atrial sensing	mV median (IQR)	3.9 (2.8–4.0)	3.1 (n/a)	n/a	3.8 (2.9–4.0)	$p = 1.0$ / n/a
Ventricular sensing	mV median (IQR)	11.5 (7.0–16.7)	7.7(4.0–16.0)	8.8 (5.9–13.0)	11.0 (7.0–16.9)	$p = 0.391$ / $p = 0.323$
Atrial pacing threshold	V median (IQR) ms median (IQR)	1.1 (0.8–2.4) 0.4 (0.4–0.4)	0.8 (n/a) 0.4 (n/a)	n/a n/a	1.8 (0.8–2.8) 0.4 (0.4–0.4)	0.267 / n/a
Ventricular pacing threshold	V median (IQR) ms median (IQR)	1.9 (1.0–3.0) 0.5 (0.4–0.8)	1.0 (1.0–1.8) 0.5 (0.4–0.6)	0.8 (0.7–3.1) 0.5 (0.4–0.7)	2.5 (1.5–3.0) 0.5 (0.4–1.0)	0.012 / 0.270
Impedance of shock coil	Ohm median (IQR)	57 (48–66)	74 (70–80)	58 (58–69)	52 (46–58)	$p < 0.001$ / $p = 0.011$
Defibrillation threshold test	Joule median (IQR)	10 (10–10)	10 (10–15)	10 (10–13)	10 (10–10)	$p = 0.274$ / $p = 0.889$
Patients with appropriate therapy (shock/ATP)	n (%)	12 (33.3)	4 (57.1)	3 (60)	5 (20.8)	$p = 0.15$ / $p = 0.112$
Patients with inappropriate shock	n (%)	4 (11.1)	1 (14.3)	1 (20)	2 (8.3)	$p = 0.55$ / 0.446

Table 3 Cause of NT-ICD failure and surgical revisions of the NT-ICD considered separately for the implantation technique

NT-ICD implantation technique	Patients with surgical revisions	Cause for malfunction	Surgical revision (overall $n = 25$)
Subcutaneous shock coil/abdominal device position ($n = 7$)	$n = 7$ (100%)	Shock coil dislocation	$n = 7/25$ (28%)
		shock coil fracture	$n = 3/25$ (12%)
		device dislocation	$n = 5/25$ (20%)
		p/s lead failure	$n = 3/25$ (12%)
Pleural shock coil fixation/abdominal device position ($n = 5$)	$n = 3$ (60%)	Shock coil fracture	$n = 1/25$ (4%)
		device dislocation	$n = 2/25$ (8%)
		p/s lead insulation breakdown	$n = 1/25$ (4%)
Pleural shock coil fixation/subcardiac device fixation ($n = 24$)	$n = 2$ (8.3%)	Shock coil dislocation/fracture	$n = 0/25$ (0%)
		device dislocation	$n = 2/25$ (8%)
		p/s lead failure	$n = 1/25$ (4%)

the high failure rate, all subcutaneous shock coils were subsequently replaced by pleural shock electrodes. Four out of 25 (16%) surgical revisions were necessary in 3/5 patients with pleural shock coil/abdominal device position: lead fracture of the pleural shock coil (1/25, 4%), insulation failure (1/25, 4%) of the p/s leads, and dislocation of the abdominally positioned device (2/25, 8%). In 2/24 patients with pleural shock coil/subcardiac device, 3/25 (12%) surgical revisions had to be performed. Indications for surgical revisions were device dislocation (2/25, 8%) and failure of the pacing/sensing leads (1/25, 4%). Kaplan-Meier analysis revealed a significantly improved performance of the pleural shock coil/subcardiac device configuration compared to subcutaneous shock coil/abdominal device combination ($p \leq 0.001$, Fig. 2) with respect to surgical revisions. There was no significant difference between pleural shock coil/abdominal device and pleural coil/subcardiac device ($p = 0.107$, Fig. 2) position.

One patient (0.5 years of age, 4.7-kg body weight, diagnosed long QT syndrome) died 47 days after NT-ICD implantation due to fulminant urosepsis. Urosepsis

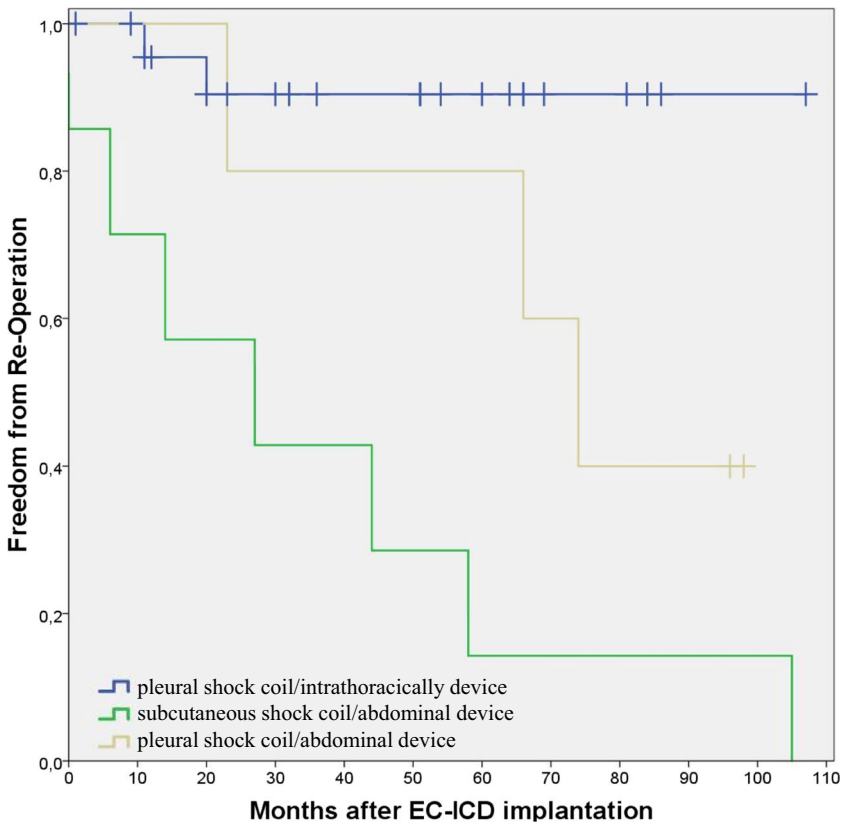


Fig. 2 Kaplan-Meier curve of freedom from re-operation in patients with subcutaneous coil/abdominal device (green line), patients with pleural coil/abdominal ICD (yellow line), and patients with pleural coil/subcardiac device (blue line). Vertical bars denote censored data. Time from NT-ICD implantation to first surgical revision was significantly shorter in patients with subcutaneous coil/abdominal device in contrast to patients with pleural coil/subcardiac device ($p < 0.001$ by log rank test). There was no statistically significant difference between patients with pleural coil/abdominal device and patients with subcutaneous coil/abdominal device ($p = 0.122$ by log rank test) or pleural coil/subcardiac device ($p = 0.107$ by log rank test)

was not considered as an operation-related complication. All other patients were still alive at the end of the study period.

3.6 Transition to endocardial ICD system

During follow-up with ongoing growth of the patients, NT-ICD was completely removed and changed to transvenous ICD system in 7/36 (19.4%) patients without any problems (Fig. 1d). It is of note, that no surgical complications related to removal of the non-transvenously positioned leads were observed.

4 Discussion

NT-ICD systems offer the opportunity to protect children and those with limited vascular access from SCD due to ventricular tachyarrhythmia. A variety of NT-ICD implantation techniques have been described before [4, 5, 9, 13, 15, 18, 19, 21–23]. Data on mid- to long-term performance of such ICD systems are sparse [2, 23]. Compared to transvenous ICD, higher failure rates of NT-ICD have been described. Shock coil failures and high defibrillation thresholds were the most frequent reasons for NT-ICD failure [17]. In the present study, we were able to provide follow-up data over a median observation period of 5.2 years in a notable number of patients after implantation of a NT-ICD. Main findings of the present study include (1) NT-ICD implantation was feasible even in small infants without any major morbidity or mortality related to the surgical procedure. (2) Pleural shock coil/subcardiac device configuration of the NT-ICD had a significantly lower revision rate compared to subcutaneous shock coil/abdominal device position. (3) NT-ICD was life-saving in a considerable number of our young patients.

4.1 Indications for ICD implantation

In general, NT-ICD implantation for primary prevention of SCD in children and young adults has increased over the last years [2]. In pediatric patients with CHD, ICD implantation for primary prevention of SCD was more common than for secondary prevention, whereas, in pediatric patients with electrical arrhythmogenic diseases, secondary prevention was the most common indication for ICD implantation [10]. These data are confirmed by the present study as a secondary prevention indication was given in 75% of the subjects with electrical arrhythmogenic disease.

4.2 Implantation technique and system survival

NT-ICD implantation at our institution commenced in 2004 with subcutaneous shock coil/abdominal device configuration followed by pleural shock coil/abdominal ICD position in 2006. Both techniques required a remarkable number of surgical revisions. After modification of the NT-ICD implantation technique to pleural shock coil/subcardiac device position in 2008, need for surgical NT-ICD revision decreased significantly. Favorable results are most likely due to firm pleural fixation of the shock coil as well as tight positioning of the subcardiac generator. These modifications prevented dislocation of either component, prevented damage from external forces, and ensured a stable electrical field which is the prerequisite for reliable defibrillation. It is of note, that NT-ICD with pleural shock coil/subcardiac device position in our study showed better performance with respect to surgical revisions compared to transvenous or subcutaneous ICD systems in pediatric patients [9, 17]. It is well known that extraction of transvenous leads potentially causes major and minor complications [7]. In contrast, we observed no complications due to extraction of the subcutaneously or pleurally positioned shock coils.

For the whole study group, shock coil impedance as well as pacing threshold was in a range as described before [21, 23], but shock coil impedance was significantly higher in subcutaneous compared to pleural shock coil position. This difference is most likely due to a higher mass of tissue between the electrode and the device in patients with subcutaneous shock coil.

4.3 Appropriate and inappropriate shocks

ICD therapies are often painful and may result in increased psychiatric morbidity in children [11]. Incidence of appropriate (33.3%) and inappropriate (11.1%) shock delivery in our study group was within the range as reported previously in pediatric patients with a comparable NT-ICD implantation technique [23]. In contrast to this finding, adults and children with subcutaneous ICD experienced more than twice the inappropriate shocks as our study population [9]. Sinus tachycardia, fast conducted supraventricular/atrial tachycardia, T wave oversensing, and malfunctioning ICD leads were common causes for inappropriate shocks [9, 23]. In order to avoid inappropriate shock delivery caused by rapidly conducted supraventricular/atrial tachycardia, it is therefore important to program higher detection rates compared to adults and to treat rigorously atrial/supraventricular tachycardia by catheter ablation or medication. Inappropriate ICD discharge due to lead fracture was observed only in one of our patients with subcutaneous shock coil. Results highlight increased safety of pleural shock coil implantation in pediatric NT-ICD. Individualized ICD programming and innovative lead monitoring algorithms including remote monitoring will likely increase safety of ICD therapy.

4.4 DFT

The need for regular DFT in young patients with NT-ICD is still under debate and data are limited. Stephenson et al. reported 5.1% DFT resulting in ICD system revision in pediatric patients and patients with congenital heart disease [20]. Although we did not find evidence for lead failure at ICD interrogation, we identified failing NT-ICD systems with indication to surgical revision in 4.3% of routine DFT. As we prioritize reliable detection and termination of life-threatening arrhythmias, we strongly advocate regular DFT in growing patients with NT-ICD.

5 Conclusions

NT-ICD was effective and safe for prevention of SCD in infants and young children. The evolution of implant techniques at our center has demonstrated that the pleural shock coil/subcardiac device approach is superior with respect to the need for surgical revision, when compared with the subcutaneous shock coil/abdominal device approach, during long-term follow-up. The pleural shock coil/subcardiac device configuration was non-inferior when compared to complications reported after transvenous or subcutaneous ICD implantation in the pediatric population. NT-ICD allowed safe growth of small patients until body size allowed implantation of a transvenous ICD.

6 Study limitations

The present study is limited by sample size and single-center design. Comparison of findings with results of other studies is impaired due to varying ICD implantation techniques and different patient management strategies including individual ICD programming.

Compliance with ethical standards

The study was approved by the institutional review board and fully complies with the Declaration of Helsinki.

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Ackerman M, Atkins DL, Triedman JK. Sudden cardiac death in the young. Circulation. 2016;133:1006–26.
2. Aykan HH, Karagoz T, Gulgun M, et al. Midterm results of implantable cardioverter defibrillators in children and young adults from a single center in Turkey. Pacing Clin Electrophysiol. 2016;39:1225–39.
3. Bar-Cohen Y, Berul CI, Alexander ME, et al. Age, size, and lead factors alone do not predict venous obstruction in children and young adults with transvenous lead systems. J Cardiovasc Electrophysiol. 2006;17:754–9.
4. Bauersfeld U, Tomaske M, Dodge-Khatami A, Rahn M, Kellenberger CJ, Prêtre R. Initial experience with implantable cardioverter defibrillator systems using epicardial and pleural electrodes in pediatric patients. Ann Thorac Surg. 2007;84:303–5.
5. Bordachar P, Marquie C, Pospiech T, et al. Subcutaneous implantable cardioverter defibrillators in children, young adults and patients with congenital heart disease. Int J Cardiol. 2016;203:251–8.
6. Burns KM, Evans F, Kaltman JR. Pediatric ICD utilization in the United States from 1997 to 2006. Heart Rhythm. 2011;8:23–8.
7. Cecchin F, Atallah J, Walsh EP, Triedman JK, Alexander ME, Berul CI. Lead extraction in pediatric and congenital heart disease patients. Circ Arrhythm Electrophysiol. 2010;3:437–44.
8. 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.
9. Jarman JW, Lascelles K, Wong T, et al. Clinical experience of entirely subcutaneous implantable cardioverter-defibrillators in children and adults: cause for caution. Eur Heart J. 2012;33:1351–9.
10. Jordan CP, Freedenberg V, Wang Y, Curtis JP, Gleva MJ, Berul CI. Implant and clinical characteristics for pediatric and congenital heart patients in the national cardiovascular data registry implantable cardioverter defibrillator registry. Circ Arrhythm Electrophysiol. 2014;7:1092–100.
11. Koopman HM, Vrijmoet-Wiersma CM, Langius JN, et al. Psychological functioning and disease-related quality of life in pediatric patients with an implantable cardioverter defibrillator. Pediatr Cardiol. 2012;33:569–75.
12. Korte T, Koditz H, Niehaus M, et al. High incidence of appropriate and inappropriate ICD therapies in children and adolescents with implantable cardioverter defibrillator. Pacing Clin Electrophysiol. 2004;27:924–32.
13. Kriebel T, Ruschewski W, Gonzalez y Gonzalez M, et al. ICD implantation in infants and small children: the extracardiac technique. Pacing Clin Electrophysiol. 2006;29:1319–25.
14. Kriebel T, Ruschewski W, Paul T. Implantation of an "extracardiac" internal cardioverter defibrillator in a 6-month-old infant. Z Kardiol. 2005;94:415–8.
15. Pospiech T, Roubertie F, Jalal Z, Bordachar P, Thambo JB. New technique for surgical epicardial implantation of a cardioverter-defibrillator in children and adults with congenital heart disease. Ann Thorac Surg. 2016;102:615–9.
16. Priori SG, Blomstrom-Lundqvist C, Mazzanti A, et al. 2015 ESC guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: the Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC)endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC). Europace. 2015;17:1601–87.
17. Radbill AE, Triedman JK, Berul CI, Fynn-Thompson F, Atallah J, Alexander ME, et al. System survival of nontransvenous implantable cardioverter-defibrillators compared to transvenous implantable cardioverter-defibrillators in pediatric and congenital heart disease patients. Heart Rhythm. 2010;7:193–8.
18. Schneider AE, Burkhart HM, Ackerman MJ, Dearani JA, Wackel P, Cannon BC. Minimally invasive epicardial implantable cardioverter-defibrillator placement for infants and children: an effective alternative to the transvenous approach. Heart Rhythm. 2016;13:1905–12.
19. 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;17:41–6.
20. Stephenson EA, Cecchin F, Walsh EP, et al. Utility of routine follow-up defibrillator threshold testing in congenital heart disease and pediatric populations. J Cardiovasc Electrophysiol. 2005;16:69–73.
21. Tomaske M, Pretre R, Rahn M, Bauersfeld U. Epicardial and pleural lead ICD systems in children and adolescents maintain functionality over 5 years. Europace. 2008;10:1152–6.
22. Wilson WR, Greer GE, Grubb BP. Implantable cardioverter-defibrillators in children: a single-institutional experience. Ann Thorac Surg. 1998;65:775–8.
23. Winkler F, Dave H, Weber R, et al. Long-term outcome of epicardial implantable cardioverter-defibrillator systems in children: results justify its preference in paediatric patients. Europace. 2018;20:1484–90.