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Implantable Cardioverter Defibrillator Therapy for Life-Threatening Arrhythmias in Young Patients

Christopher B. Stefanelli, David J. Bradley, Sarah Leroy, Macdonald Dick II, Gerald A. Serwer, and Peter S. Fischbach

University of Michigan Congenital Heart Center, Women's Hospital, UMHS, L1242, Box 0204, Ann Arbor, MI 48109-0204

Abstract. ***Objectives:*** **This study examined the indications, efficacy and outcomes of implantable cardioverter defibrillator (ICD) use in the pediatric population.**

Background: **ICDs are first-line therapy for adults resuscitated from sudden cardiac death (SCD) or at high risk for life-threatening ventricular arrhythmias. Use of ICDs in children and young adults is infrequent and there are few data regarding this group.**

Methods: **We abstracted and analyzed data for all patients in whom ICDs were implanted.**

Results: **A total of 38 devices were implanted in 27 patients. Age ranged from 6 to 26 years (mean, 14) and weight ranged from 16 to 124 kg (mean, 47). Diagnoses included long QT syndrome (9), hypertrophic cardiomyopathy [6], repaired congenital heart disease [5], and idiopathic ventricular tachycardia/fibrillation [4]. Indications comprised resuscitated SCD [15], syncope [9], and life-threatening ventricular arrhythmia [3]. Initial device placement was infraclavicular in 13, abdominal in 13 and intrathoracic in 1. Epicardial leads were used with 5 systems. A single coil lead was used in 17. Seven patients, all previously resuscitated from SCD, experienced 88 appropriate successful discharges. There were 6 inappropriate discharges in 3 patients. Mean time to device replacement was 3.1 years (n = 11). Complications included 2 infected systems, 2 lead dislodgments, 2 lead fractures, 1 post-pericardiotomy syndrome, 1 adverse event with defibrillation threshold (DFT) testing, and 1 patient with psychiatric sequelae. No deaths occurred with implanted ICDs.**

Conclusions: **These data demonstrate that ICDs provide safe and effective therapy in young patients. The indications for ICDs as primary preventive therapy remain uncertain.**

Key Words. **children, implantable cardioverter defibrillator, sudden cardiac death, ventricular arrhythmia**

Introduction

Since Mirowski and associates [1] reported the first use of implantable cardioverter-defibrillators (ICDs) in 1980, these devices have become a widely accepted therapy for patients who have survived aborted sudden cardiac death (SCD). The MADIT [2] and MUSTT trials [3] demonstrated the usefulness of ICDs for primary prevention of SCD in certain high-risk populations. In 1989, Kral and colleagues [4] reported the first use of ICDs in young patients. Since this initial report, ICD use in pediatric patients has increased substantially, paralleling the increase in adults. The increased use has been due in part to technologic advancements, including smaller devices and electrodes, transvenous lead systems, improved detection algorithms, expanded diagnostic storage capabilities, single coil configurations using the generator as the second high voltage electrode, and devices with dual chamber pacing and sensing capabilities. Additionally, identification of children at high risk for SCD has expanded, resulting in implantation of ICDs as a primary preventive measure.

Pediatric patients, however, still represent less than 1% of all persons with ICDs [5] and the clinical experience in young patients remains limited. Previous reports have consisted of retrospective surveys of multiple institutions [6–9], small series [10–15], and case reports [16,17]. The purpose of this study is to report a single center's experience with this evolving technology and to provide data on the indications, efficacy and outcomes of ICD use in young patients.

Methods

The patient database was queried for all individuals in whom an ICD was implanted at the University of Michigan Congenital Heart Center. Medical records, electrocardiograms, Holter tracings, electrophysiologic studies, implantation data, telemetered data, and interrogation

Address for correspondence: Macdonald Dick, II MD, Women's Hospital, UMHS, L1242, Box 0204, Ann Arbor MI, 48109-0204.

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data were abstracted for these individuals. Indications for ICD implant and clinical histories were reviewed. Device type and configuration, pacing parameters, defibrillation thresholds (DFTs), device activity, and complications were recorded.

Mean, standard deviation and median values were calculated for patient demographic variables, follow-up intervals and defibrillation thresholds. Freedom from first appropriate defibrillation discharge, freedom from inappropriate defibrillation discharge and freedom from ICD-related complication were assessed by Kaplan-Meier analysis using Graph Pad Prism Version 3.00 (Graph Pad Software, Inc., San Diego, CA). Implant and post-implant follow-up DFTs were compared using unpaired Student t tests.

Results

Between October, 1992 and June, 2001, a total of 38 devices were implanted in 27 patients (Table 1). There were 14 (52%) females. The age at implantation ranged from 6.2 to 26.3 years (mean, 14.5 ± 4.4; median, 14.6 years). The weight of the patients ranged from 16 to 124 kg (mean, 49 ± 21; median, 47 kg). The mean follow up interval was 32 ± 29 months (range, 1–100; median, 24 months). The diagnoses of the 27 patients in whom ICDs were implanted are summarized in Table 1. The most frequent diagnoses were long QT syndrome ($n = 9$, 33%), hypertrophic cardiomyopathy ($n = 6$, 22%), repaired complex congenital heart disease ($n = 5$, 19%), and idiopathic ventricular tachycardia/ventricular fibrillation ($n = 4$, 15%). The remaining 3 devices were implanted in a patient with coronary arteriopathy and cardiomyopathy following an orthotopic heart transplantation, a patient with Marfan syndrome, and a patient with arrhythmogenic right ventricular dysplasia. Surgical repairs for patients with congenital heart disease included transannular outflow tract patch for tetralogy of Fallot ($n = 2$), Mustard procedure ($n = 1$) for transposition of the great arteries, intraventricular tunnel for double outlet right ventricle ($n = 1$), and arterial switch operation and ventricular septal defect closure for double outlet right ventricle with subpulmonary ventricular septal defect (Taussig-Bing anomaly; $n = 1$). Three of the 6 individuals with hypertrophic cardiomyopathy had undergone myectomy.

Indications for initial ICD placement in the 27 patients (Table 1) were grouped into three categories: (1) resuscitated sudden cardiac death (SCD; $n = 15$), (2) syncope ($n = 9$), and (3) life-threatening ventricular arrhythmia ($n = 3$), defined as ventricular tachycardia or fibrillation on Holter or during electrophysiologic testing. The patients in whom the initial indication was syncope or life-threatening ventricular arrhythmia were also considered the primary prevention group as they had not experienced resuscitated SCD ($n = 12$). Seven of the 9 individuals with long QT syndrome and all of the individuals with idiopathic ventricular tachycardia/fibrillation experienced out-of-hospital resuscitated SCD. Of the 9 individuals with syncope, 6 had documented ventricular tachycardia or fibrillation either on electrocardiographic monitoring or during electrophysiologic study. Among the remaining 3 individuals with syncope, 2 had long QT syndrome and a family history of SCD, and 1, in whom a dual chamber ICD was implanted, had hypertrophic cardiomyopathy, rapidly progressive left ventricular outflow tract obstruction and non-sustained ventricular tachycardia.

Initial ICD placement was infraclavicular in 13 patients (48%), abdominal in 13 (48%) and intrathoracic in 1 (4%). With two exceptions, replacement devices were placed in the pocket created for the original device. Among the 13 initial abdominal implants, 9 utilized transvenous leads tunneled from the subclavian area (Fig. 1). Six of the initial infraclavicular devices were placed beneath the pectoralis muscle. Infraclavicular ICD placement was used in patients as young as 7 years and as small as 30 kg.

Thirty-three lead systems were placed in the 27 patients, of which 28 were transvenous. Replacement leads were implanted for increased lead impedance in 3 patients and for exchanging epicardial for endocardial systems in 3 patients. Of the 28 transvenous lead systems, a single coil to active device ("hot can") configuration was used in 17 and a dual coil configuration in 11. Of the 17 generators serving as the second electrode, 7 were placed in an abdominal pocket and 10 in an infraclavicular pocket. The youngest and smallest patients to receive transvenous leads were 7 years and 22 kg, respectively. An active device configuration was used in patients as young as 7 years and as small as 24 kg. In 3 patients, lead extenders were utilized to tunnel endocardial leads from the infraclavicular region to abdominally located devices (Fig. 1). The only epicardial system placed in the last six years was in a 16 kg child.

DFTs were obtained using a basic stepdown protocol ($n = 33$) and at implant ranged from 5 J to 21 J (mean, 11 ± 5; median, 10 J). Follow-up DFTs, 6–8 weeks after implant ($n = 19$), ranged from 5 J to 20 J (mean, 13 ± 5 J; median 11; $p = 0.13$). There were no significant differences in the implant or follow-up mean DFTs related to the device position, lead configuration or use of a lead extender.

Table 1. *Clinical data on 38 ICDs in 27 patients*

Patient number	Implant date	Age at implant	Sex	Weight(kg)	Diagnosis	Indication	Family history	Concurrent medication	Device location	Lead config.	S/D chamber	Initial DFT (J)	Succ. disch.	Unsucc. disch.	Inappr. disch.	Follow-up (months)	Complications
1	Nov-97	13	M	60	LQTS	RSCD	pos, scd	BB	A	TVS	S	10	2	0	0	43	
2	Jan-01	7	M	27	LQTS	RSCD	pos, scd	BB	A	TVS	D	15	0	0	0	5	
3	Feb-93	14	F	32	LQTS	RSCD	neg	BB	A	EPI	S	n/a	52	0	0	100	post-pericardiotomy
	Nov-94	14		39					A	TVD	S	5	3	2	0		PTSD
	Jun-97	17		44					A	TVD	D	n/a	2	0	0		DFT event
	Sep-00	20		42					A	TVD	D	6	0	0	0		
4	Jun-99	12	M	41	IVT/VF	RSCD	pos, scd	BB	I	TVD	S	10	0	0	0	24	lead dislodgment
													0	0	0		
5	Aug-98	15	M	50	LQTS	RSCD	neg	BB	I	TVS	S	15	3	12		34	
	Apr-01	18		61					I	TVS	D	10	0	0	0		
6	Jan-01	21	F	124	HCM	LTVA	pos	Amio	I	TVS	S	15	0	0	0	5	
7	Aug-99	14	M	54	LQTS	syncope	pos, scd	BB	I	TVD	S	10	0	0	0	22	
8	Mar-99	12	F	49	IVT/VF	RSCD	neg	BB	I	TVS	S	15	0	0	0	27	
9	Oct-97	10	M	24	HCM	RSCD	neg	CCB, Amio	A	TVS	S	10	2	0	0	44	
	May-00	13		35					I	TVS	S	5	0	0	0		
10	Nov-97	15	M	44	CHD	syncope	neg	BB	A	TVS	S	5	0	0	0	43	
11	Sep-97	8	M	22	IVT/VF	RSCD	neg	BB	A	TVD	S	5	7	2	0	45	lead fracture
12	Jul-95	7	M	30	IVT/VF	RSCD	neg	CCB	IT	EPI	S	10	0	0	0	71	
	Aug-00	13		56					I	TVS	S	10	0	0	0		
13	Oct-92	11	F	37	LQTS	RSCD	pos	BB	A	TVD	S	n/a	0	0	0	1	infection
14	Jun-97	6	M	16	HCM	RSCD	neg	BB	A	EPI	S	5	0	0	0	1	
15	Mar-99	10	F	33	HCM	syncope	neg	CCB	A	TVS	D	15	0	0	0	27	
	Feb-00	11		37					A	TVS	D	10	0	0	0		
16	Nov-99	9	M	37	CHD	syncope	neg	BB	I	TVS	D	6	0	0	0	19	
17	Feb-93	16	M	50	CHD	RSCD	neg	Amio	A	EPI	S	n/a	4	0	0	100	lead dislodgment
	Apr-96	20		55					A	EPI	S	n/a	7	0	3		
	Sep-99	23		60					A	EPI/TVA	D	21	5	0	0		
18	Jan-96	14	F	63	LQTS	RSCD	pos, scd	BB	I	TVD	S	10	0	0	1	65	lead fracture
	Jun-99	17		67					I	TVD	S	15	0	0	0		
19	Nov-99	18	F	45	Marfan	LTVA	pos	CCB	I	TVD	D	15	0	0	2	19	
20	Jul-93	17	F	50	CHD	RSCD	neg	BB	A	EPI	S	18	0	0	0	1	infection
21	Sep-00	10	F	27	CHD	syncope	neg	Amio	A	TVS	S	10	0	0	0	10	
22	Apr-01	26	F	50	ARVD	syncope	pos, scd	BB	I	TVS	D	10	0	0	0	2	
23	Mar-99	12	F	70	HCM	syncope	neg	CCB	I	TVD	D	11	0	0	0	27	
24	Jul-98	23	M	75	OHT	syncope	pos	CCB	I	TVS	S	10	0	0	0	14	
25	Jun-94	14	F	40	LQTS	RSCD	neg	Amio	A	TVD	S	20	1	0	0	84	
	Mar-00	20		56					A	TVD	S	15	0	0	0		
26	Jun-99	15	F	106	HCM	LTVA	neg	BB	I	TVD	D	10	0	0	0	24	
27	Apr-00	14	F	67	LQTS	syncope	neg	BB	I	TVS	D	5	0	0	0	14	

A = abdominal; ARVD = arrhythmogenic right ventricular dysplasia; BB = beta blocker; CCB = calcium channel blocker; CHD = congenital heart disease; D = dual; DFT = defibrillation threshold; EPI = epicardial; HCM = hypertrophic cardiomyopathy; I = infraclavicular; Inappr. Disch. = inappropriate discharge; IT = intrathoracic; IVT/VF = idiopathic ventricular tachycardia/ventricular fibrillation; LQTS = long QT syndrome; LTVA = life-threatening ventricular arrhythmia; n/a = not available; neg = negative; OHT = orthotopic heart transplant; pos = positive; RSCD = resuscitated sudden cardiac death; S = single; SCD = sudden cardiac death; Succ. Disch. = successful appropriate discharge; TVA = transvenous atrial; TVD = transvenous dual coil; TVS = transvenous single coil; Unsucc. Disch. = unsuccessful appropriate discharge.

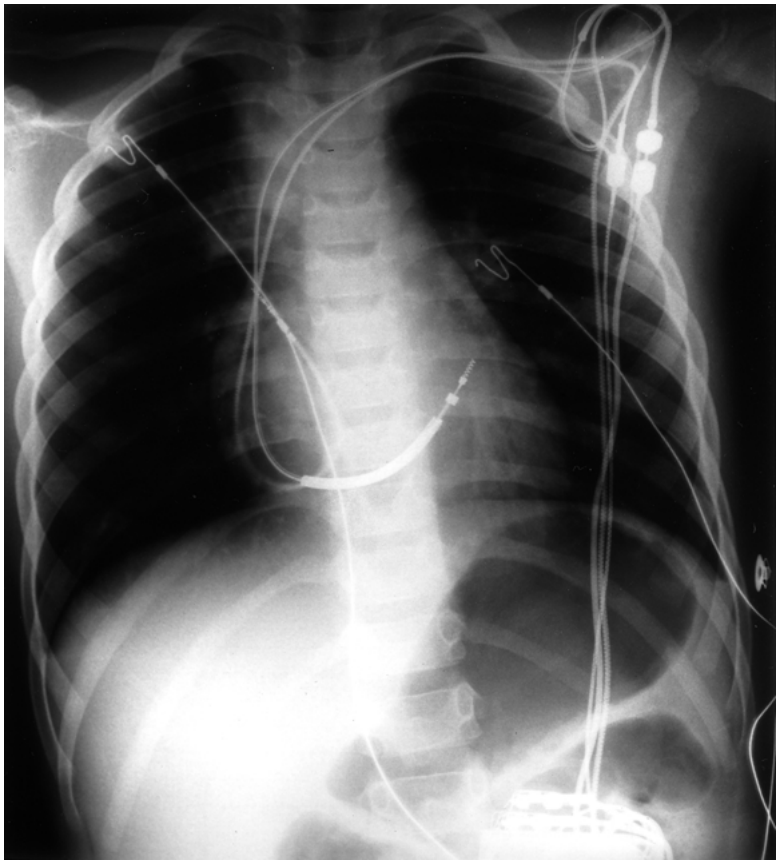


Fig. 1. *Radiograph of patient 2 demonstrating transvenous leads tunneled from the infraclavicular region to an ICD placed in the left upper quadrant. The use of lead extenders may permit future placement of an infraclavicular device without removal of the endocardial leads. This is an active device configuration between the abdominally placed device and the coil placed along the right ventricular diaphragmatic surface.*

Seven of the 27 (26%) patients experienced a total of 88 appropriate successful ICD discharges (Fig. 2A). Appropriate discharges were confirmed by ICD telemetry and defined as ventricular tachycardia or fibrillation accurately detected according to the programmed recognition cycle lengths and number of cycles. Among these 7 patients, 4 had long QT syndrome (63 discharges) and 1 each had hypertrophic cardiomyopathy (2 discharges), congenital heart disease (16 discharges) and idiopathic ventricular tachycardia/fibrillation (7 discharges). A single patient with long QT syndrome was defibrillated 45 times over a 48 hour time period during a "torsades storm." Overdrive ventricular pacing controlled her arrhythmia. All 7 of these patients underwent initial device placement for secondary prevention of SCD; their initial clinical indication was resuscitated SCD. No patient in whom the initial clinical indication was syncope or life-threatening ventricular arrhythmia (primary prevention of SCD) experienced an appropriate discharge during the follow-up period.

Three (11%) patients experienced a total of 6 inappropriate discharges (Fig. 2B). One patient whose device discharged inappropriately was found to have a fractured sensing lead. Rapid motion of her left arm generated electrical noise that was misinterpreted as ventricular fibrillation by the device and resulted in an inappropriate device discharge. The faulty lead was replaced and the patient has experienced no further discharges. One patient received three inappropriate discharges due to atrial tachycardia misinterpreted as ventricular tachycardia. The patient subsequently had a dual chamber device implanted with resolution of the inappropriate arrhythmia detection. The third patient was shocked twice during fast sinus tachycardia with frequent ventricular premature beats. Reprogramming of the device to a shorter sensed ventricular fibrillation cycle length corrected the problem.

Two (7%) of the patients received unsuccessful initial ICD discharges and were successfully defibrillated by subsequent programmed therapies.

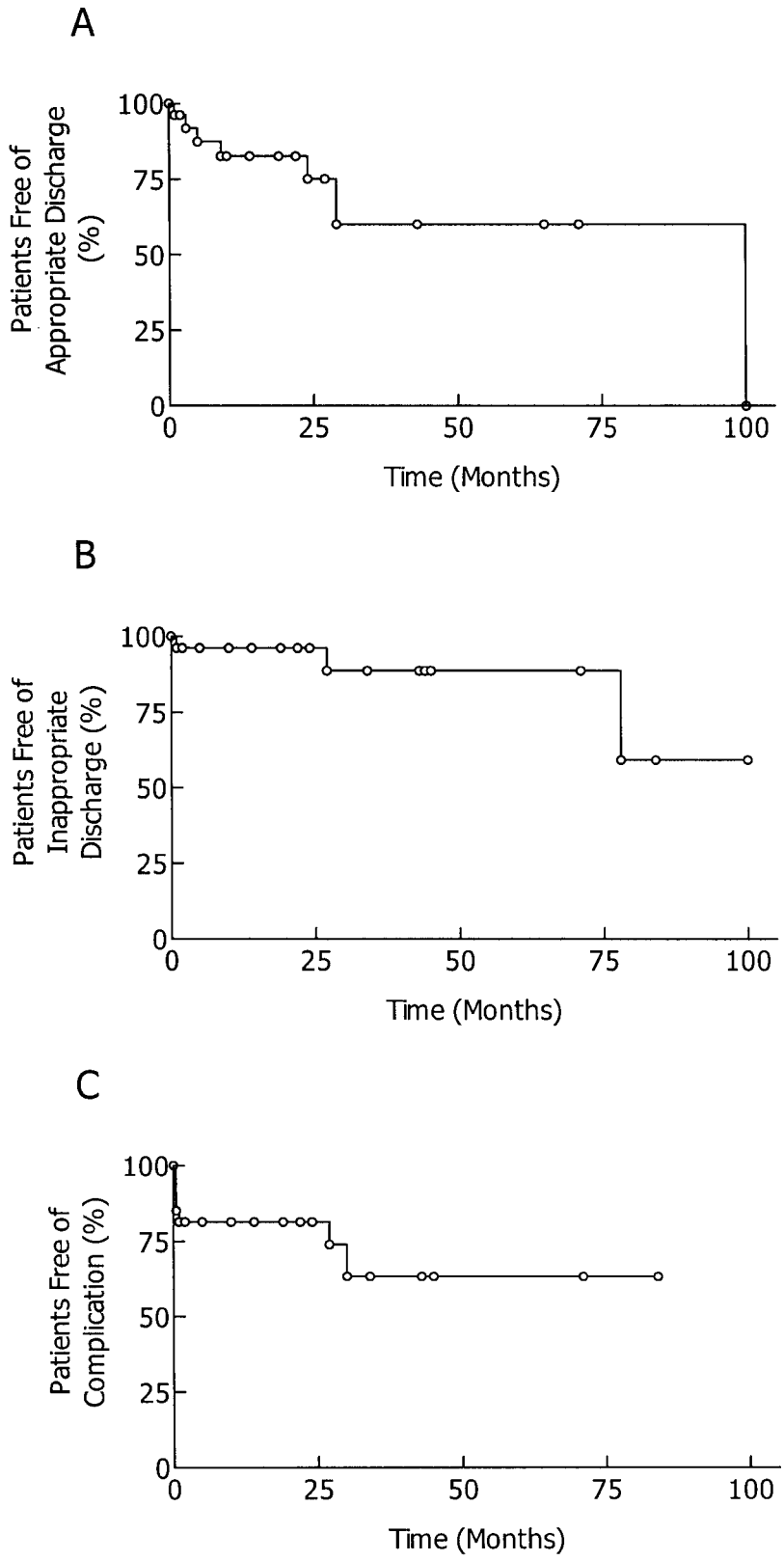


Fig. 2. *Kaplan-Meier analysis for freedom from appropriate device discharge (A), freedom from inappropriate device discharge (B), and freedom from ICD-related complication (C).*

A patient with idiopathic ventricular tachycardia/fibrillation was defibrillated 7 times; on 2 of those occasions, the first 20 J shock was unsuccessful in restoring sinus rhythm. Subsequent 30 J discharges were successful and the device has since been reprogrammed to deliver a 30 J initial shock. A patient with LQTS was successfully defibrillated 3 times and experienced a total of 12 unsuccessful discharges. Following the last event, the device was removed and a new ICD capable of delivering a biphasic shock waveform was implanted. Both of these patients had low initial and follow-up DFTs at testing, 5 J and 10 J, and 15 J and 15 J, respectively.

Eleven devices were replaced during the study time period, with a range of time to replacement of 11 months to 5.7 years (mean, 3.2 ± 1.4; median, 3.2 years). Indications for replacement included generator elective replacement time ($n = 4$), updating device technology ($n = 5$), lead change and approaching elective replacement time ($n = 1$), and recall of a device ($n = 1$). Among the 5 ICDs replaced to update technology, 4 were to add an atrial lead for dual chamber sensing and pacing and 1 was to deliver a biphasic shock waveform.

Nine complications related to the ICD systems occurred in 7 patients (26%; Fig. 2C). There were 2 infected systems requiring removal within 1 month of placement. Two transvenous leads became dislodged; one lead was repositioned in the right ventricle, and 1, in 1993, was replaced with an epicardial lead. Two transvenous leads have fractured. One, a fractured sensing lead, resulted in an inappropriate discharge and was replaced. The second sustained a fracture in the proximal coil of a dual coil transvenous lead (Fig. 3). The device was reprogrammed to use the generator as the second high voltage electrode and the patient has received successful appropriate discharges using this configuration. One patient had post-pericardotomy syndrome following implantation of an epicardial lead system. Following implantation of a replacement device several years later, the same patient experienced severe bradycardia and hypotension during anesthesia induction for post-implant, follow-up DFT testing,

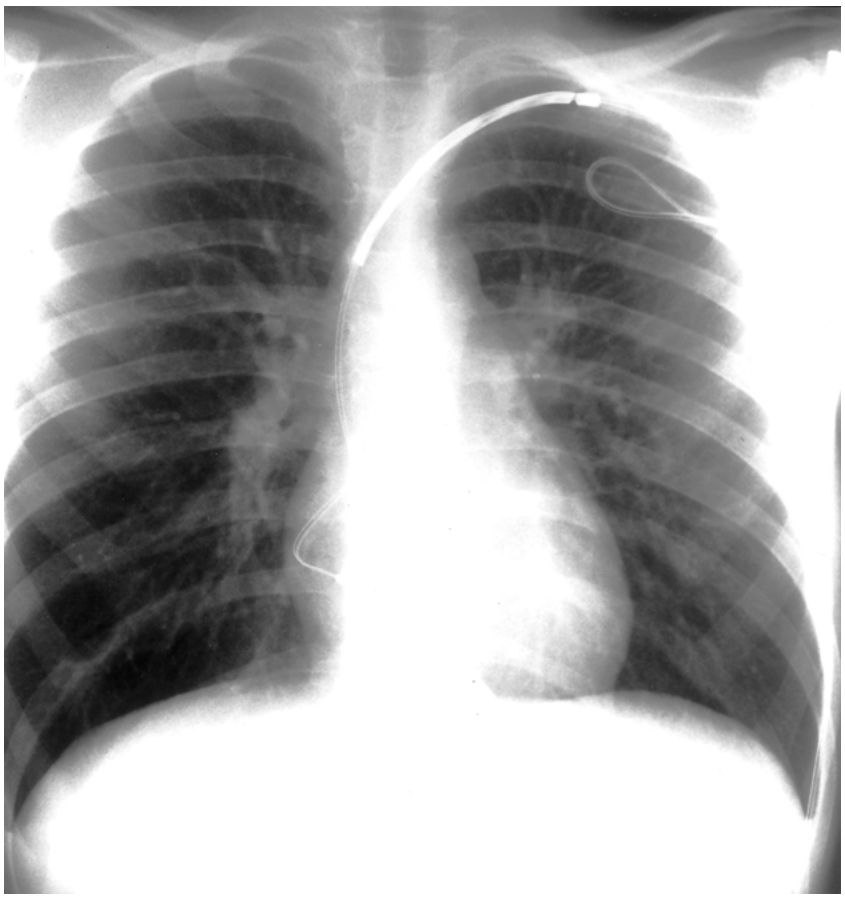


Fig. 3. *Radiograph of patient 11. The proximal coil of the dual coil transvenous lead is fractured at the junction of the clavicle and 1st rib (see text).*

requiring extensive resuscitative efforts and an intensive care unit admission. One patient with long QT syndrome and severe neurologic impairment following resuscitated SCD died suddenly 2 years after explant of an infected device; her parents had refused implantation of a second ICD. No patient with an implanted device died. Finally, one patient has suffered from severe post-traumatic stress disorder.

Discussion

The need for ICDs in children and young adults is infrequent. Secondary prevention of resuscitated SCD is a widely accepted indication. In contrast, device implantation for primary prevention of SCD remains controversial. Advances in the recognition of genetic conduction system disorders associated with SCD (e.g., long QT syndrome, Brugada syndrome and arrhythmogenic right ventricular dysplasia) and improving stratification of patients at risk for SCD (e.g., hypertrophic cardiomyopathy and repaired congenital heart disease) have contributed to the increase in use of ICDs in children for primary prevention of SCD. Despite these developments, as demonstrated by our experience and that of others, prospective identification of children and young adults at risk of SCD remains difficult and confounds the use of ICDs for primary prevention of SCD in this population [18].

Secondary prevention of SCD, in patients with long QT syndrome, hypertrophic cardiomyopathy, repaired congenital heart disease, and idiopathic ventricular tachycardia/fibrillation, was the most common indication for ICD implantation in our patients. Twelve of 27 (44%) and 10 of the last 13 initial implants, however, have been in individuals who have not experienced resuscitated SCD, and in whom the indication was considered primary prevention of SCD.

Certain technologic advancements have proven particularly germane to pediatric patients. The most obvious is the reduction of device size from 300 cc to < 35 cc over the last 10 years. Smaller device size has obviated the need for abdominal device placement in all but the smallest patients. Smaller diameter transvenous leads and shorter single coil leads have also permitted implantation of ICD systems in smaller patients without thoracotomy. Innovative configurations, such as a single coil transvenous lead placed along the right ventricular diaphragmatic surface linked to an abdominally placed device as the second high voltage electrode, allow deployment in smaller patients (Fig. 1). This arrangement, which has been utilized in 6 of our patients, has been associated with low DFTs (mean, 11 J), 4 appropriate successful discharges and no unsuccessful discharges [14]. Due to the increased morbidity associated with epicardial lead systems, our current practice is to implant transvenous systems in all but the smallest patients and in those in whom there is no venous access. To date, we have used a transvenous lead system in a patient as small as 22 kg.

DFTs have been consistently low in our patients irrespective of lead configuration. No patient has required the placement of an extra superior vena cava coil or a subcutaneous array in order to lower DFTs. Controversy exists concerning the necessity of post-implant DFT testing in pediatric patients. Post-implant DFT testing involves patient inconvenience, discomfort, and expense, in addition to the inherent risks of anesthesia, ventricular arrhythmia induction and defibrillation. Of the 27 patients in this series, one with long QT syndrome developed a profound bradyarrhythmia with hemodynamic collapse requiring extensive resuscitation prior to the induction of ventricular tachycardia/fibrillation for DFT testing. In addition, no patient in our cohort required reprogramming of their device based on the results of DFT testing [19]. Based on this experience and reports suggesting that follow-up DFT testing is most appropriately reserved for individuals in whom device dysfunction is suspected [20], we no longer perform routine follow-up DFT testing.

Only three (11%) of our patients experienced inappropriate discharges, a proportion lower than previously reported in young patients [4,6–8,11]. Expanded sensing and programming capabilities and the use of dual chamber devices has resulted in improved discrimination between sinus tachycardia or atrial arrhythmias and ventricular arrhythmias. This rate of inappropriate discharges compares with a reported rate of approximately 5% in large adult series [21]. Differentiation of sinus tachycardia from ventricular tachyarrhythmias is particularly important in pediatric patients, as they may achieve sinus rates approaching or exceeding 200 beats per minute. Treadmill exercise testing is useful in determining the appropriate high rate zone for individual patients.

While only 7 of the 27 patients (26%) in this series received appropriate device discharges, among the 16 patients with at least 2 years follow up, 7 (44%) were appropriately and successfully defibrillated. This finding is consistent with other reports, in which 40–60% of young patients with ICDs received appropriate device discharges within 1.5–2 years of implantation [6–9,11]. Because younger patients have the potential for considerable longevity and thus a longer period of risk, lower appropriate shock rates are acceptable.

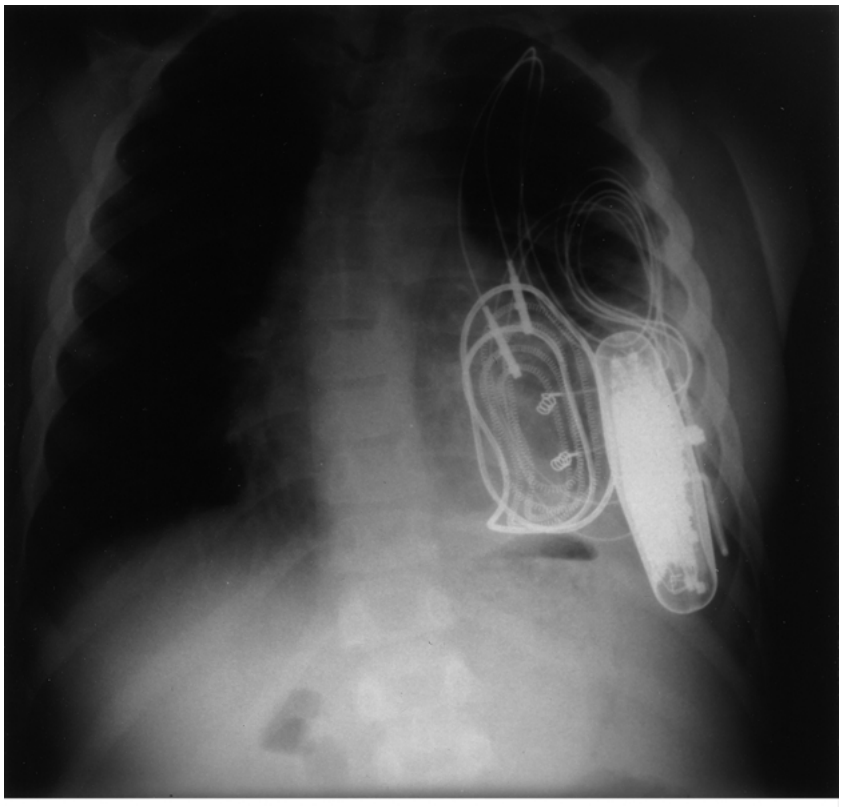
Complex cardiac and systemic disorders in some pediatric patients requiring ICDs may necessitate creative device configurations [15]. One

patient in our series has Eagle-Barrett (prune belly) syndrome, renal failure requiring a kidney transplant and idiopathic ventricular tachycardia/fibrillation. Due to the absence of abdominal musculature and his small size, the ICD was implanted intrathoracically utilizing an epicardial lead system (Fig. 4). His device was subsequently replaced, at which time he had grown sufficiently to allow the placement of an infraclavicular device and transvenous leads.

Prior reports of ICDs in the young have described complication rates requiring system revisions in 29% [8] and 45% [22] of patients. In our series, 7 of the 27 (26%) of the patients have experienced a total of 9 device-related complications. Five of the complications (19%) necessitated system revisions. Two of the 27 (7%) patients developed a pocket infection requiring device explant. Link et al. [22] previously reported an infection rate of 18% in pediatric patients undergoing ICD implant. The infection rate in their experience was higher than that observed in adult patients who underwent device implantation by the same physicians, during the same time period and in the same laboratory. The cause of the increased incidence of infection among their pediatric patients was not identified. Finally, one of the patients in our cohort experienced a lead dislodgment and underwent successful reimplantation of the indwelling lead. To avoid this particular complication, we use active fixation defibrillation leads in the ventricle.

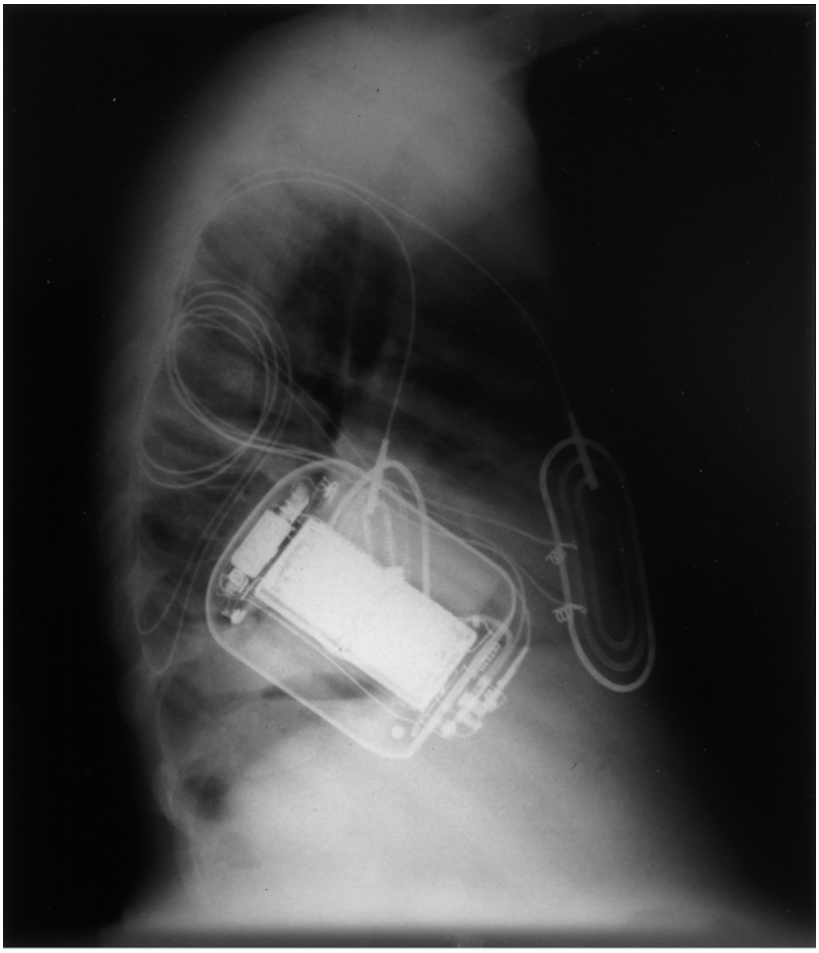
The potential for life-threatening arrhythmia and receiving or anticipating ICD discharges are sources for considerable anxiety, particularly for children. Three patients who were repeatedly shocked experienced anxiety that persisted for more than one month. Two patients developed school phobias. One patient developed post-traumatic stress disorder, requiring antidepressant and anxiolytic medication with ongoing psychiatric therapy. All of our patients participate in support groups and most attend a national annual retreat for young patients and children with ICDs [23].

In summary, the indications and use of ICDs in the pediatric population are expanding. Improved device and lead technology combined with earlier identification of patients at risk for SCD has contributed to this increase. This report, though smaller than adult series, represents the largest



(A)

Fig. 4. *Posteroanterior (A) and lateral (B) chest radiographs of patient 12, in whom intrathoracic device placement was necessitated by the absence of abdominal musculature (Eagle-Barrett syndrome) and small size.*



(B)

Fig. 4. *(Continued)*

single center pediatric series to date and demonstrates the applicability of ICD systems in this population. While primarily designed for use in adult patients, these devices can be configured in a variety of ways to suit a diversity of young patients. With careful programming of the arrhythmia detection parameters, ICDs can be used safely and effectively in children and young adults resuscitated from SCD. However, for the use of ICDs as primary prevention in children, the indications remain unclear and the efficacy uncertain.

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