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Midterm experience with implantable cardioverter-defibrillators in children and young adults†

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Aims This single-centre study was undertaken to review our experience with implantable cardioverter-defibrillator (ICD) implantation in children with relatively different aetiologies.

Methods and results We retrospectively reviewed the records of the paediatric patients who underwent ICD implantation between October 2001 and December 2008. The data of these patients were collected by reviewing the patients' medical records and computerized departmental pacemaker databases. A total of 28 patients who underwent ICD implantation during this period were included in this study. The median age was 12 years and median weight was 32 kg. Most of the patients had ion-channel diseases ($n = 13$) or cardiomyopathy ($n = 11$). Devices were implanted for either secondary ($n = 22$) or primary ($n = 6$) prevention. The selected ICD generator type was single chamber in 22 patients, dual chamber in 5 patients, and biventricular in 1. Nineteen patients received 122 shocks. Fifteen of 22 patients (68.2%) from the secondary prevention group and 2 of 6 patients (33.3%) from the primary prevention group experienced at least one appropriate shock during a median period of 11.3 months (range: 4 days–6.5 years). Forty-two inappropriate shocks were delivered in seven (31.8%) patients from the secondary prophylaxis group during a median period of 11.3 months. The most important reason for inappropriate shocks was T-wave oversensing. In six patients, lead-related acute or chronic complications occurred.

Conclusion The ICD was safe and effective in interrupting malignant arrhythmias in children and adolescents with a high risk of sudden cardiac death. However, the occurrence of lead complications is significant. The incidence of therapies delivered by the device, with appropriate and inappropriate shocks, was high and interfered with the quality of life. The most important reason for inappropriate shocks was T-wave oversensing. Careful programming is mandatory to reduce the inappropriate shocks.

Keywords Appropriate shock • Children • Implantable cardioverter-defibrillator • Secondary prophylaxis • Sudden cardiac death

Introduction

Indications for implantable cardioverter-defibrillators (ICDs) have evolved considerably from their initial implantation exclusively in patients who had survived one or more cardiac arrests and failed pharmacological therapy.[1] Multiple clinical trials have established that ICD use results in improved survival compared with antiarrhythmic agents for secondary prevention of sudden cardiac death (SCD).[2–5] Large prospective, randomized, multicentre studies have also established that ICD therapy is effective for primary prevention of sudden death and improves total survival in selected patient populations who have not previously had a

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cardiac arrest or sustained ventricular tachycardia (VT).[6,7] Similar indications are applied in the paediatric population. However, the experience in children is limited and accounts for less than 1% of all implanted devices. Children at risk for SCD have a wide variety of underlying cardiac diseases such as the broad spectrum of congenital heart disease, inherited arrhythmogenic diseases, and hypertrophic or dilated cardiomyopathy (HCMP or DCMP).[8] Indications for ICD therapy have gradually changed from secondary to primary prevention.[9,10] Current data on paediatric ICD therapy are derived from small single-centre studies and retrospective multicentre studies.[8,11–15] This single-centre study was undertaken to review our experience with ICD implantation in children with relatively different aetiologies.

Methods

We retrospectively reviewed the records of the paediatric patients who underwent ICD implantation between October 2001 and December 2008. A total of 28 patients who underwent ICD implantation during this period were included in this study. The data of these patients were collected by reviewing the patients' medical records and computerized departmental pacemaker databases.

Demographic information, indications for ICD implantation, previous medications, results of electrophysiological study (EPS), implant electrical parameters, appropriate and inappropriate shock data, complications, and survival were recorded for all implants. Demographics included patient age, weight, gender, anatomic diagnosis, and electrical diagnosis.

The implant data included type of ICD, type of lead, indication for implant, electrical parameters, and defibrillation threshold when available. The shock data included the receipt of any shocks (whether appropriate or inappropriate), time of shock since the ICD was implanted, reasons for shock (whether appropriate or inappropriate), and whether or not the shock was successful at converting rhythm. Defibrillation therapy for a ventricular arrhythmia faster than the programmed detection criteria, and accurately detected by the ICD, was categorized as an appropriate shock. Defibrillation therapy received for anything other than a ventricular arrhythmia faster than the programmed detection criteria was categorized as an inappropriate shock.

Complications were compiled by time since implant and were divided into acute (within 30 days of ICD implant) and chronic complications.

Devices were implanted under general anaesthesia into subpectoral pockets, using transvenous lead systems, except in one patient. Defibrillation threshold testing and measurement of R-wave amplitude and pacing threshold of the rate sensing/pacing leads were performed intraoperatively in the standard fashion. Stored ICD data were obtained within 24 h of treatment, at the first and third months, and every 6 months thereafter.

Statistical analysis was performed with SPSS 11.0 (SPSS Inc., Chicago, USA). All parameters are expressed as medians and ranges. Shock frequencies were analysed using the Fisher exact test based on the sample size. A P-value of <0.05 was considered statistically significant.

Results

Patient data

During the study period, a total of 28 patients underwent ICD implantation. The median age of the subjects at the time of first ICD implant was 12 years (range: 2–25 years) and the median weight was 32 kg (range: 10.3–94 kg). Eighteen of the children (72%) were male. The mean follow-up period between implantation and study endpoints or the last follow-up was 2.6 ± 1.9 years (range: 1 month –7 years). Diagnostic categories were cardiac ion-channel diseases ($n = 13$), cardiomyopathy ($n = 11$), and congenital heart diseases ($n = 4$). Cardiac ion-channel diseases included catecholaminergic polymorphic VT (CPVT; $n = 7$), long QT syndrome (LQTS; $n = 5$), and the Brugada syndrome. All congenital heart disease patients had post-operative tetralogy of Fallot (TOF; *Table 1*).

Implantable cardioverter-defibrillator indications and previous treatment

The indications for ICDs that were implanted for secondary prevention were syncope ($n = 12$), aborted SCD ($n = 8$), and VT induced by programmed stimulation in EPS ($n = 1$) or detected in Holter ($n = 1$). In six (21%) patients, ICDs were implanted for primary prevention (*Table 2*). In these patients, the indications for primary prevention were: family history of SCD in a patient with LQTS with a very long QT interval ($n = 1$), family history of SCD in a CPVT patient ($n = 1$), very low ejection fraction ($\leq 30\%$) in patients with DCM ($n = 2$), and induced ventricular fibrillation (VF) and VT on EPS in post-operative TOF patients ($n = 2$).

Before implantation, in 89% of the patients, at least one anti-arrhythmic drug was used (*Table 2*). Multiple anti-arrhythmic drugs were used in six patients. The use of anti-arrhythmic medication remained virtually unchanged after ICD implantation. Three patients had pacemaker therapy before the actual ICD implantation. Nine of the 10 patients with clinical VT and VF were

Table 1 Indications for ICD and diagnosis of patients ($n = 28$)

Age at implant (years)	11.6
M/F	18/10
Indication for ICD	
Syncope	11
Sudden cardiac death survivor	8
EPS (VT/VF) or VT (Holter monitoring)	3
Primary prevention	5
Cardiac ion channel diseases	13
Catecholaminergic polymorphic ventricular tachycardia	7
Long QT syndrome	5
Brugada syndrome	1
Cardiomyopathy	11
Dilated	7
Hypertrophic obstructive cardiomyopathy	2
Arrhythmogenic right ventricular cardiomyopathy/ dysplasia	2
Congenital heart disease	4
Corrected Fallot patients	4

ICD, implantable cardioverter-defibrillator; EPS, electrophysiological study; VF, ventricular fibrillation; VT, ventricular tachycardia.

Table 2 ICD indications and previous treatment

Patient number	Age	Cardiac disease	Indication	Follow-up (months)	Appropriate shocks	Cause of appropriate shock	Inappropriate shocks	Cause of inappropriate shock	Medication	Complication
1	5	CPVT	PP	36	3	VT			At	
2	13	CPVT	Syncope	35	4	2 VF, 2 VT	21	Lead problem	At, V	Lead problem
3	17	CPVT	Syncope	45	5	VT			At, V	
4	7	CPVT	Syncope	19	7	2 VF, 5 VT			Prop	Failed shocks
5	14	CPVT	Syncope	28			5	T-wave sensing	Prop, V	
6	11.5	CPVT	RCA	18						
7	8.5	CPVT	Syncope	1	1	VT	2	T -wave sensing	At	
8	4	LQTS	Syncope	35	2	VF	2	T-wave sensing	Prop	
9	12	LQTS	Syncope	38	2	VT			Prop	
10	3	LQTS	PP	14	13	12 VF, 1 VT			At, M	Lead problem, failed shocks
11	11	LQTS	Syncope	18	10	VT			Prop	
12	2	LQTS	RCA	11						
13	3.5	BS	Syncope	33	4	VT	1	Sinus tachycardia	A, Q	
14	12	DCM (NC)	Syncope, VT	62					A, D	
15	10	DCM (NC)	EPS VT	82	4	3 VF, 1 VT	1	Sinus tachycardia	A, D	Failed shocks
16	6	DCM (NC)	RCA (EPS VF)	86	7	5 VF, 2 VT			A, M	Lead problem
17	9	DCM (NC)	PP	25					A, Pr	Lead problem
18	14	DCM	RCA	13	2	VF, VT			A	
19	14	DCM	PP	16						
20	17	DCM	VT (Holter)	11	8	3 VF, 5 VT			A, M, prop	
21	9	HCMP	RCA	61			10	Sinus tachycardia, T-wave sensing	A	
22	12	HCMP	RCA	40	4	VT			A, prop	
23	16	ARVD	Syncope	13					A	
24	12	ARVD	Syncope	2	2	VT			A, prop	
25	12	FT (TC)	RCA	51	2	VT			A	
26	25	FT (TC)	PP (EPS, VF)	43					S	
27	19	FT (TC)	RCA	13					D, A	
28	23	FT (TC)	PP	23					At	Infection

A, amiodarone; ARVD, arrhythmogenic right ventricular dysplasia; At, atenolol; BS, Brugada syndrome; CPVT, catecholaminergic polymorphic ventricular tachycardia; D, digoxin; DCM, dilated cardiomyopathy; EPS, electrophysiologic study; FT, Fallot tetralogy; HCMP, hypertrophic cardiomyopathy; LQTS, long QT syndrome; M, mexiletine; NC, non-compaction; PP, primary prophylaxis; Pr, propafenone; Prop, propranolol; Q, quinidine; RCA, resuscitated cardiac arrest; S, sotalol; TC, total correction; V, verapamil; VF, ventricular fibrillation; VT, ventricular tachycardia.

inducible at programmed ventricular stimulation. Accessory pathways were ruled out in all cases.

Implantation, leads, and devices

All of the defibrillators except one with epicardial were implanted the left pectoral region by percutaneous subclavian puncture. Contrast medium injection was performed from left arm to visualize subclavian and cephalic vein, and roadmap was used before subclavian or cephalic venipuncture. An extrathoracic subclavian vein approach was taken pain over. All of the transvenous defibrillator leads had the active-fixation mechanism and dual coils, and the right ventricular apical region was used for implantation. There were 6 atrial leads (all Medtronic), 1 left ventricular lead (Medtronic), 28 ventricular leads [25 Medtronic (3 Sprint Fidelis); 2 St Jude Medical-Riata 1580; 1 Guidant], and 1 Medtronic SQ lead implanted. The defibrillation threshold at implantation was 16 J (range: 10–35 J). A total of 28 patients underwent ICD implantations. The selected ICD generator type was single chamber in 22 patients, dual chamber in 5 patients, and biventricular in 1. Single-chamber devices were used mainly in young patients and/or patient with cardiac ion-channel diseases; however, dual-chamber devices were mainly used in patient with operated congenital heart disease and/or heart failure due to cardiomyopathy. There were 31 implantations of new devices (28 Medtronic, 1 Guidant, and 2 St Jude Medical) during the follow-up. Three of them were replacement procedure due to battery depletion ($n = 2$) and infection ($n = 1$).There were no implant procedure-related deaths.

Programming

The VF zone was programmed in all patients to >180–240 b.p.m. depending on the patient's age, heart failure, and physical activity, at a minimum of 10–20 b.p.m. above the maximal sinus rhythm seen at the exercise test. The VT zone was additionally programmed in six patients with proven VT to 150–220 b.p.m., which was 10–20 b.p.m. below the documented or induced VT rate. In patient with haemodynamically stable, it was programmed to antitacycardia pacing followed by shock treatment; otherwise all treatments were programmed to shock therapy. All shocks were programmed to 20 J or above depending on the threshold values.

When the reason was sinus tachycardia for inappropriate shock, mainly β-blocker dosage was rechecked according to the patient body weight and increased to a dosage effective enough in limiting maximal heart rate at exercise test, and VT and/or VF detection zones were increased above sinus tachycardia values if possible.

Shocks and complications

Nineteen patients received 122 shocks. Eighty of these shocks were appropriate shock at a mean of four shocks per patient (median: 3, range: 1–12). Only two of the six patients with dual chamber or biventricular ICD experienced shock. All of them were appropriate. The time to first appropriate ICD therapy was 17 ± 20 months (range: 4 days–78.5 months). Five patients experienced more than five shocks. The reason for appropriate shocks were VT ($n = 50$) and VF ($n = 30$). In three patients who received a total of 24 appropriate shocks, the shocks failed to terminate VT/VF. Fifteen of 22 patients (68.2%) from the secondary prevention group and 2 of 6 patients (33.3%) from the primary prevention group experienced at least one appropriate shock during a median period of 11.3 months (range: 4 days–6.5 years).

Forty-two inappropriate shocks were delivered in seven (31.8%) patients from the secondary prophylaxis group during a median period of 11.3 months (range: 4 days–6.5 years). Three patients had sinus tachycardia, which led to inappropriate ICD discharges. In two patients, the reason was T-wave oversensing. One patient had sinus tachycardia and/or T-wave oversensing. Inappropriate shocks were delivered due to artefact after a fracture of a lead in another patient 15 months after implantation (*Figure 1*). Further inappropriate discharges were prevented by using negative chronotropic drugs ($n = 3$) or device reprogramming to prevent T-wave sensing ($n = 2$).

Three acute complications occurred in three patients during the first 30 days of implantation. One patient with an epicardial lead developed a significant decrease in defibrillation impedance 3 days after the procedure. The patient was taken to the electrophysiology laboratory again, and it was seen that the defibrillation threshold had increased from 15 to 35 J. Lead connection problem with battery developed in the second patient. The third patient developed lead infection and acute bacterial endocarditis (*Staphylococcus aureus*) 1 week after ICD implantation. The whole system including the lead and battery were removed. He made a complete recovery under medical treatment and received a new ICD 2 months later. Chronic complications related to lead issues ($n = 3$) included lead fractures ($n = 2$) 17 and 19 months after implantations and lead dislodgement ($n = 1$) 7 months after implantation.

Four batteries were replaced during the study period, due to endocarditis in one and end-of-life in three patients. Battery

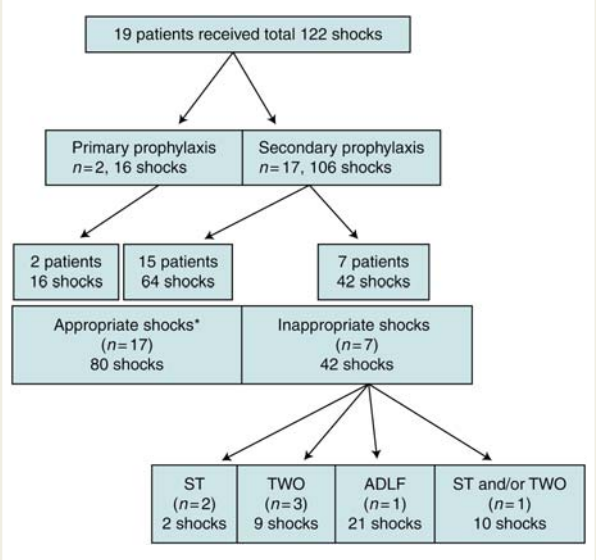

Figure 1 Distribution of type and number of shocks and reasons for inappropriate shocks. Asterisks denote that in some patients, both appropriate and inappropriate shocks were observed. ADLF, artefact due to lead fracture; ST, sinus tachycardia; TWO, T-wave oversensing.

end-of-life developed prematurely in one of them at the 19th month.

Deaths

During the follow-up, two patients died. Both of them were from the cardiomyopathy group and had left ventricle idiopathic non-compaction. One patient who received his ICD at the age of 9 years and died after 25 months after implantation due to intractable congestive heart failure. The other patients received her ICD at the age of 6 years and experienced appropriate shocks five times, but died 7 years after implantation of the ICD due to VF sensing problem. It was probably due to an acute lead problem. The lead was Medtronic Sprint Fidelis and patient's medical record about ICD and the lead controls till the last control before death event were normal. Another reason could be arrhythmia storm at the home which may induce electromechanical dissociation after time.

Discussion

Implantable cardioverter-defibrillator therapy is increasingly indicated in the paediatric population for cardiac ion-channel diseases and arrhythmias associated with cardiomyopathies or congenital heart disease.[14–18] Sudden cardiac death is unfortunately common in untreated channelopathies and malignant myocarditis/cardiomyopathies, which are associated with a 25–100-fold increased risk for SCD.[8] This study shows that ICD therapy in children and young adolescents for primary and secondary prevention of a life-threatening ventricular arrhythmia is feasible and effective.

Clinical and epidemiological aspects of the population of our study, number of patients involved, mean age, aetiology of the cardiovascular disease, and indication for ICD implantation are similar to those of the five most recent studies.[11,13,14,16–18] Mortality in these studies ranged from 0 to 20% during the mean follow-up period of 3.4 years and was also similar to our results, which showed a mortality rate of 7.4%.

In the present series, 13 of 28 ICD implantations were performed in patients with cardiac ion-channel diseases (CPVT, LQTS, and Brugada). The most appropriate shocks were received in ion-channel disease patients in our study population (10 of 13). In this group, CPVT was the most frequent diagnosis (7 of 13). Catecholaminergic polymorphic VT may cause syncope, convulsions, and sudden death during physical activity or emotional distress. Syncope was the main complaint in five (71%) patients. The mortality rate was given as 10–25%.[19–21] Medical treatment with propranolol and verapamil may decrease the incidence of arrhythmia. Implantation of intracardiac defibrillators should be considered in those resistant to drug therapy. Delay in diagnosis and inadequate treatment can result in SCD.[21] In the present series, we also found a relatively large number of appropriate shocks in CPVT patients (five of seven).

The most frequent incidence of an appropriate shock was also observed in LQTS patients in the ion-channel diseases group. In four of five LQTS patients, 27 appropriate and 2 inappropriate shocks were observed. Goel *et al.*[22] reported their experience with ICD therapy for children with LQTS and observed appropriate therapies in 5 of 12 patients, inappropriate shocks in 4 of 12, two complications, and one death after the electrical storm.

In our series, 9 of 11 patients with cardiomyopathy had an ICD implant because of DCMP or HCMP; the 10th and 11th patients had arrhythmogenic right ventricular dysplasia. In patients with DCMP, the reason was non-compaction in the majority of the patients. The ICD was implanted for primary prophylaxis, as well as after syncope and resuscitated cardiac arrest (RCA). In patients with idiopathic DCMP, an ejection fraction below 30% and non-sustained VT on a Holter monitoring, both prophylactic implantation of an ICD as well as implantation after syncope or sustained VT or VF seems appropriate.[23] In patients with DCMP, such appropriate shocks are not equivalent to life-saving shocks[24]; therefore, the reported higher shock rates for patients with DCMP compared with those with HCMP or ischaemic heart disease should not be taken as equivalent to a higher rate of SCD prevention. In the present series, four of seven patients with DCMP received appropriate shocks. Two patients were HCMP. In both conditions, ICD implantation was due to RCA. Although one patient received appropriate shocks, the other received inappropriate shocks because of sinus tachycardia. Patients with HCMP who survive cardiac arrest or sustained VT remain at risk for a recurrent event.[25] In these patients, ICD implantation should be considered.[26] In patients with HCMP without sustained ventricular arrhythmias or syncope, prophylactic ICD implantation may be considered when there is a family history of SCD at a young age.[26]

In our series, 4 of 28 patients had an ICD implantation because of improved surgical outcome for patients with congenital heart disease; the most common cause of death among these patients is SCD.[27] The use of pacemaker technology and radiofrequency catheter-ablation may control some of these arrhythmias, whereas VTs may especially lead to SCD.[14] In our series, we had a relatively small number of patients with congenital heart disease (all of them operated TOF). In this group, one patient received two appropriate shocks.

Seventeen of 28 patients (60.7%) received 80 appropriate shocks, and 7 of 28 (25%) patients received inappropriate shocks mainly because of sinus tachycardia, T-wave oversensing, or lead problems. In this study, 9 of 12 patients (75%) with syncope history and 4 of 8 with RCA history received appropriate shocks. Our data suggest appropriate ICD discharges in this young patient population and high efficacy in the prevention of SCD. All of the 56 spontaneous VT and VF episodes were terminated by ICD therapy. In particular, ICD therapy is demonstrated to be effective in appropriately selected high-risk patients presenting with syncope or RCA. Korte *et al.*[13] reported that 127 spontaneous VT and VF episodes were terminated with ICD therapy. Same authors[13] categorized shocks in paediatric ICD recipients as appropriate in 15 of 20 patients (75%) and inappropriate in 10 of 20 patients (50%), attributable mainly to supra-VT, T-wave oversensing, or lead failure. In our study, only seven patients (25%) received 42 inappropriate shocks, and all of these patients were in the secondary prophylaxis group. Hamilton *et al.*[28] reported only 39% of young ICD recipients without inappropriate ICD discharges after 3 years of follow-up, and inappropriate therapies were mainly caused by sinus tachycardia. Korte *et al.*[13]

confirmed a particularly high incidence of inappropriate ICD therapies in children and adolescents. The main causes in our series were sinus tachycardia and T-wave oversensing followed by lead complications. As in other studies,[13,14] most inappropriate therapies occurred during early follow-up, i.e. within the first 18 months after ICD implantation. Alexander *et al.*[29] reported a 38% complication rate over a 2-year follow-up in 76 paediatric and young adult ICD patients, including infection, lead failure, and potential for electrical storm. Eicken *et al.*[17] reviewed 16 patients who received an ICD and found 7 of the 16 (44%) received appropriate therapies, whereas 4 (25%) received inappropriate shocks. In a recent study including 22 children with HCMP who received an ICD,[30] only four (18%) received appropriate shocks and four also received one or more inappropriate shocks; complications were reported for 3 of 22 patients. Increased recognition of the risks of ICD lead fractures and inappropriate shocks have led to the development of novel implantation techniques and lead-less ICD systems for small children and patients with complex congenital heart disease.[31]

Implantation of an ICD has potential complications, including infections of the pocket, pleural or pericardial effusion, and lead malfunction. The incidence of infections and lead malfunctions may be higher in paediatric patients.[32] In our study, lead-related complications were frequent. Five of 28 patients (18%) developed lead problems. The numbers in this study are too small to show whether skeletal growth or the lead system used directly influenced the occurrence of inappropriate therapies due to the lead failure. However, in this centre, a previous study indicated that none of the patient-related risk factors was correlated with pacemaker lead fracture. Among lead-related risk factors, only the fixation mechanism was found to be correlated with lead fracture.[33] The rate of lead complications was high in other groups of patients (44%) and reflects the difficulties in the paediatric age group.[13,17]

In 74% of our paediatric patients, additional anti-arrhythmic drug therapy was considered appropriate to either reduce or prevent the occurrence of ventricular tachyarrhythmias. Ten of 28 of our patients did not experience any shocks. We suggest that careful attention to optimizing device programming, medical management, and encouraging compliance with prescribed therapies and recommendations may diminish inappropriate shock frequency in this patient population.

In the two patients who died in the present series, irreversible cardiomyopathy contributed to the congestive heart failure in one of them. The cause in the other was probably untreatable arrhythmia. This mortality rate was similar to that in the literature.[11,13,14,16–18]

Conclusion

The ICD was safe and effective in interrupting malignant arrhythmias in children and adolescents with a high risk of SCD. However, the occurrence of lead complications is significant. The incidence of therapies delivered by the device, with appropriate and inappropriate shocks, was high and interfered with the quality of life. The most important reason for inappropriate shocks was T-wave oversensing. Careful ICD programming of the individual child immediately after ICD implantation and during the follow-up period is mandatory to reduce the inappropriate shocks.

Conflict of interest: none declared.

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