Original Article

Incidence of Shock and Quality of Life in Young Patients with Implantable Cardioverter-Defibrillator

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Summary

Objectives: To assess the incidence and causes of ICD shocks in children and adolescents and their impact on quality of life (QoL).

Methods: From March, 1997 to February, 2006 29 patients (15.7 ± 5.4 years of age) underwent ICD implantation. Resuscitated cardiac arrest (41.5%), sustained ventricular tachycardia (27.6%), and primary prophylaxis of sudden cardiac death (30.9%) were the indications for ICD implantation. The number of therapies was assessed by interview and using the ICD telemetry. The SF-36 questionnaire was used to assess QoL, which was compared to that of healthy individuals. The Kaplan-Meier method was used for the analysis of shock-free survival.

Results: After 2.6 ± 1.8 years of follow-up, eight (27.6%) patients received 141 appropriate shocks due to polymorphic ventricular tachycardia (VT) (6) or ventricular fibrillation (VF) (2), and 11 (37.9%) received 152 inappropriate shocks due to supraventricular tachyarrhythmias (8) or oversensing (3). Appropriate shock-free survival expectancy was 74.2% ± 9.0 and 66.7% ± 10.7 after one and three years, respectively. Decreased QL was observed as regards physical functioning (61.7 ± 28.7), vitality (64.7±19.1), mental health (65.9±22.7) and emotional aspects (66.7±38.5). All patients reported fear and concern related to the ICD.

Conclusion: Despite the great efficacy of this therapy, the high incidence of shocks interfered with QoL and adaptation to the device.

Key words: Defibrillators, implantable; quality of life; adolescent.

Introduction

The use of implantable cardioverter-defibrillators (ICD) is already well established in the prevention of sudden cardiac death in adults with malignant ventricular arrhythmias[1-6]. However, few reports of their use among children and adolescents are found in the literature, given its low frequency among this population. Available data show that less than 1% of the ICDs are implanted in patients younger than 21 years of age[7].

In the young population, indications for this device are different from those for the adult population because they are mostly restricted to the primary or secondary prevention of sudden death in genetic diseases: congenital long-QT syndrome, Brugada syndrome, hypertrophic cardiomyopathy, and arrithmogenic dysplasia of the right ventricle[6-11].

Although the impact of ICD therapies on children and adolescents is still infrequently described, the incidence of shocks, whether appropriate or not, is known to be higher among this population than among adults.

The objective of the present study was to analyze the incidence and causes of ICD shocks among children and adolescents, verifying the impact of this therapy on quality of life, as well as the social and psychological adaptation of these patients and their compliance to treatment.

Methods

Population characteristics - From March, 1997 to February, 2006 29 patients younger than 21 years of age underwent ICD implantation in the Heart Institute (InCor) of the Medical School of the University of São Paulo (FMUSP).

At the time of implantation, age ranged from 2 to 21 years, with a mean of 15.7 ± 5.4 years and a median of 17.4 years. Ten patients were female (34.5%) and 19 were male (65.5%).

Resuscitated cardiac arrest (CA) (41.5%), spontaneous or induced sustained ventricular tachycardia (SVT) (27.6%), and primary prevention of sudden cardiac death (SCD) in high-risk individuals (30.9%) were the indications for ICD implantation.

The main heart diseases identified were: congenital long-QT syndrome in seven (24.1%) patients, hypertrophic cardiomyopathy in seven (24.1%), dilated cardiomyopathy in five (17.2%), arrhythmogenic dysplasia of the right ventricle in three (10.3%), catecholaminergic polymorphic ventricular tachycardia (PVT) in two (6.9%), and Brugada syndrome in one (3.4%). The patients' clinical characteristics are summarized in Table 1.

Syncope was present in 23 (79.3%) patients, and was associated with tachycardic palpitations in 12 (41.4%);

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Table 1 – Clinical characteristics of the patients undergoing ICD implantation

N	Gen-der	Age (years)	Heart Disease	Indication for Implantation	HF FC (NYHA)	Beta-bloc.	Anti-arrhythm.	EPS
1	F	2	Long-QT S.	Resuscit. CA	I	yes	Yes	-
2	M	3	Long-QT S.	SVT	I	yes	-	-
3	M	4	Long-QT S.	SVT	II	yes	yes	-
4	M	5	Cong. heart def.	SVT	I	-	-	-
5	F	11	Long-QT S.	Resuscit. CA	I	yes	-	-
6	M	13	HCM	Resuscit. CA	I	yes	-	Not induced
7	F	13	Long-QT S.	SVT	I	yes	-	-
8	M	13	Long-QT S.	Resuscit. CA	I	-	-	Polym. VT
9	F	14	Catechol. PVT	Syncopes and SCD of 5 siblings	I	yes	-	-
10	M	14	ADRV	SVT	I	-	-	-
11	M	16	HCM	Resuscit. CA	I	-	-	Not induced
12	M	16	Idiopathic Aneurysm in LV	Resuscit. CA	I	yes	-	Polym. VT
13	F	17	DCM	Resuscit. CA	II	-	-	-
14	M	17	ADRV	SVT	I	yes	-	Polym. VT
15	M	17	HCM	Familial SCD (2 aunts and grandmother); IV septum = 28 mm	II	yes	yes	-
16	M	17	None	Resuscit. CA	I	-	-	-
17	M	18	Catechol. PVT	SCD of 5 siblings	I	yes	-	NSVT
18	M	19	HCM	Familial SCD (father and twin brother); IV septum = 38 mm	II	yes	-	-
19	F	19	DCM	Resuscit. CA	III	-	-	-
20	M	19	Long-QT S.	SVT	I	yes	-	-
21	M	19	DCM	Resuscit. CA	I	-	-	-
22	M	20	ADRV	NSVT	I	-	-	Polym. VT
23	F	20	HCM	Resuscit. CA	II	yes	-	Not induced
24	M	20	DCM	Resuscit. CA	II	yes	yes	-
25	M	20	DCM	SVT	II	-	-	Not induced
26	F	21	HCM	Familial SCD (5 siblings); IV septum = 24 mm	II	yes	yes	-
27	F	21	HCM	Familial SCD (2 siblings); IV septum = 22mm	I	-	-	VF
28	F	21	Brugada Syndrome	Familial SCD (2 siblings)	I	-	-	VF
29	M	21	Cong. heart def.	SVT	I	yes	yes	-

F= female; M= male; Long-QT S.= Congenital long-QT Syndrome; Cong. Heart def..= Congenital heart defect; HCM= hypertrophic cardiomyopathy; DCM= dilated cardiomyopathy; Catechol. PVT = catecholaminergic polymorphic ventricular tachycardia; ADRV = arrhythmogenic dysplasia of the right ventricle; Brugada S. = Brugada Syndrome; Resuscit. CA= Resuscitated cardiac arrest; SCD= sudden cardiac death; SVT= sustained ventricular tachycardia; NSVT = non sustained ventricular tachycardia; IV septum = interventricular septum width; FC= functional class; HF= heart failure; NYHA= New York Heart Association; Beta-bloc.= beta-blocker; EPS= electrophysiological study; VF= ventricular fibrillation; Polym. VT= polymorphic ventricular tachycardia.

Family history of SCD was reported by nine (31.0%) patients, mainly related to the presence of hypertrophic cardiomyopathy or congenital long-QT syndrome.

Prior to implantation, 58.6% and 18.5% of the patients were taking beta-blockers and amiodarone, respectively. Electrophysiological study was performed in 11 (37.9%)

patients, with induction of ventricular tachyarrhythmias in seven (24.1%).

ICD implantation procedures - Fifteen (51.7%) patients underwent ventricular device implantation and 14 (48.3%) received atrioventricular systems.

Transvenous approach was the most frequently used for lead implantation, via subclavian vein in 25 (86.2%) patients and via femoral vein in only three (10.3%) preschool children (Figure 1a). Implantation via transthoracic transatrial approach was performed in one infant due to difficulty of venous access (Figure 1b). The pulse generator was implanted in the abdominal wall in four (13.8%) patients and in the thoracic wall in 25 (86.2%), whether in the infraclavicular (65.5%) or in the submammary region (20.7%).

Programming and assessment of ICD therapies - Programming of the devices was customized considering the underlying heart disease, characteristics of the arrhythmia, clinical conditions and pharmacologic therapy used (Table 2).

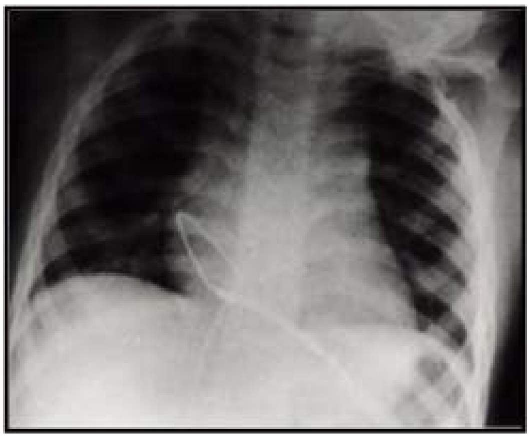

Fig. 1a – *Chest radiograph in posterior anterior view demonstrating lead positioning of the ICD implanted via femoral vein.*

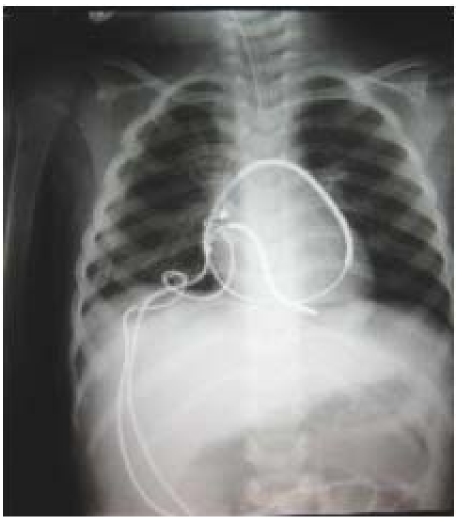

Fig. 1b – *Chest radiograph in posterior anterior view showing lead positioning of the ICD implanted via transatrial transthoracic approach.*

Table 2 – ICD customized programming

Heart disease	Bradycardia therapies	Programmed Tachycardia therapies* ATP	Programmed Tachycardia therapies* Shocks
Long-QT S.	70-80 bpm	No	200 bpm
HCM	50 bpm	No	180 bpm
DCM	40 bpm	170 bpm	200 bpm
Brugada S.	40 bpm	No	200 bpm
ADRV	40 bpm	170 bpm	200 bpm

*Long-QT S.- Congenital long-QT syndrome; HCM - hypertrophic cardiomyopathy; DCM - dilated cardiomyopathy; ADRV - arrhythmogenic dysplasia of the right ventricle; Brugada S. - Brugada syndrome; bpm - beats per minute; ATP - anti-tachycardia therapy with pacing * - the first therapeutic zone was always programmed as a monitor.*

Clinical follow-up started immediately after implantation and included clinical and electronic assessment of the device. ICD therapies were assessed by analyzing the electrograms, and correlating them with the patient's symptoms.

Assessment of quality of life and adaptation to the device - Quality of life was assessed after ICD implantation using the SF-36 instrument. The sample was comprised of 15 adolescents from our population.

The SF-36 is a multidimensional questionnaire consisting of 36 items encompassing 8 domains: functional capacity (10 items), physical functioning (4 items), bodily pain (2 items), general health (5 items), vitality (4 items), social functioning (2 items), emotional aspects (3 items), mental health (5 items), and one additional question of comparative assessment between the current health and that of one year earlier. After the questionnaire is applied, a score is attributed to each question. The scores are then transformed in a 0-to-100 scale, where zero corresponds to the worst health condition, and 100 to the best, and each domain is analyzed separately[12].

An interview addressing psychosocial aspects related to the adaptation to the device was also carried out. Pain caused by the ICD shock was assessed using a numeric scale (ranging from zero to ten), and a verbal scale (very severe, severe, moderate, mild, and none).

The application of the SF-36 questionnaire and the interviews were standardized and performed by a doctor or nurse participating in the study.

Variables studied and statistical analysis - The expectation of shocks in relation to time was determined by the non-parametric Kaplan-Meier method, and the difference between the frequency of appropriate and inappropriate shocks in time was compared using the Log-Rank test.

The analysis of predictive factors (age, gender, heart disease, and indication for ICD implantation) for the occurrence of shocks was made using the Student's t test or the Fisher's exact test.

The influence of ICD on the patients' quality of life was analyzed according to the mean values of all domains of the SF-36 instrument in comparison with the values found among the general population. Reliability of the SF-36 instrument in the assessment of quality of life of the population studied was measured by the Cronbach's coefficient alpha, and alpha values ≥ 0.7 were considered satisfactory.

All data were analyzed using the Statistical Package for Social Sciences (SPSS) software program, and *p* values ≤ 0.05 were considered significant.

Results

Clinical follow-up - The follow-up period was of 2.6±1.8 years, ranging from five days to 5.7 years. Two patients were lost to follow-up and three were followed in the service they had been referred from. Four deaths (13.8%) occurred, and the causes were: untreatable arrhythmia (1), cardiogenic shock (1), bronchopneumonia (1), and unknown (1).

Reoperations were performed 16 times due to: pulse generator exhaustion (9), problems related to the lead (3), change in pacing mode (2), infection (1), and defibrillation patch implantation (1).

During the follow-up period, 293 shocks were delivered in 15 (51.7%) patients. Mean time elapsed between the implantation and the first ICD shock was 26.3 ± 28.8 months.

Shocks considered appropriate were delivered in eight (27.6%) patients, in a total of 141 therapies. The arrhythmias treated were polymorphic VT and VF in six and two patients, respectively. One patient with arrhythmogenic dysplasia of the right ventricle had an arrhythmic storm and received 63 shocks in one day.

Shocks considered inappropriate were delivered in 11 (37.9%) patients, in a total of 152 therapies. The causes for the shocks were tachycardic atrial rhythms in eight patients and oversensing of signals other than the QRS complex in three (10.4%). Among the supraventricular tachycardias, sinusal tachycardia occurred in three patients, and atrial fibrillation in one patient who received approximately 100 inappropriate shocks.

Analysis of event-free survival showed a 74.2% ± 9.0 expectancy of patients to be free of appropriate shocks after one year and 66.7% ± 10.7 after three years of follow-up. (Figure 2)

No significant differences were observed between the incidence of appropriate and inappropriate shocks (p=0.1) during the follow-up period. No variable (age, gender, heart disease, and indication for ICD implantation) was identified as a predictive factor both for the occurrence of appropriate and inappropriate shocks.

Three complications required surgical correction: 1) one case of bacterial endocarditis in a patient with long-term tracheostomy, so that system replacement was necessary; 2) one case of high defibrillation threshold which required epicardial patch lead implantation for defibrillation, and 3) one case of lead displacement which required repositioning.

Quality of life and adaptation to the device - The quality of life domains that suffered the major alterations, considering the scores ranging from zero to 100, were: physical functioning (61.7±28.7), vitality (64.7±19.1), mental health (65.9±22.7) and emotional aspects (66.7±38.5).

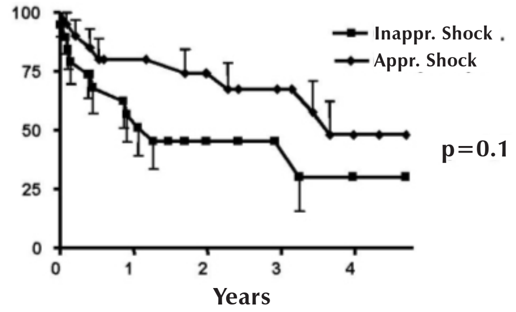

Fig. 2 *– Analysis of appropriate and inappropriate shock-free survival expectancy using the Kaplan-Meier method.*

Comparison of mean values of each domain of the SF-36 among the population of this study and the general population[13] is shown in Table 3.

Table 3 - Comparison of mean values of SF-36 domains between patients with ICD and the general population

SF-36 Domains	Patients with ICD (mean value)	General population (mean value)
Functional Capacity	68.0	72.3
Physical Functioning	61.7	68.0
Bodily Pain	72.4	68.8
General Health	67.3	64.5
Vitality	64.7	58.8
Social Functioning	85.0	80.6
Emotional Aspects	66.7	78.5
Mental Health	65.9	75.8

The Cronbach's coefficient alpha, considering all SF-36 domains, was 0.74. The physical component, which encompasses the domains of functional capacity, physical functioning, bodily pain, and general health, showed a coefficient of 0.61. The mental component, which encompasses the domains of mental health, emotional aspects, social functioning and vitality, showed a coefficient of 0.72.

During the interviews, 50.0% of the patients reported learning difficulties, 58.3% professional difficulties, and 25.0% social relationship difficulties.

After implantation, 86.7% of the patients felt more confident and 53.4% reported improvement in health with reduction of symptoms. Fear and concern related to the use of the ICD, however, were reported by all patients interviewed.

As regards the sensation caused by the shocks delivered by the ICD, severe or very severe pain was reported by 60.0% of the patients, and 75% reported fear of receiving a further shock.

Discussion

Despite the vast experience already obtained with the clinical use of ICDs[1-5], studies involving children and adolescents with this type of implantable device are not frequently found in the literature, and are limited to case reports or case series[7,14-17].

Clinical and epidemiological aspects of the population of our study, number of patients involved, mean age, etiology of the cardiovascular disease, and indication for ICD implantation are similar to those of the three most recent studies[8,10,11]. Mortality in these studies ranged from 4.2% to 20% during the mean follow-up period of 3.6 years, and was also similar to our results, which showed a mortality rate of 13.8%.[8, 10,11]

Appropriate and inappropriate shocks - Another relevant aspect in the follow-up of this population is the incidence of inappropriate shocks, which is higher when compared with that of the adult population, and may be present in approximately 38 to 61% of the cases (and in 20 to 30% in adults)[9,18-20].

Consistent with these data, our study showed that the incidence of inappropriate shocks was 38%, and the predominant cause was supraventricular tachyarrhythmias which occurred in four patients for whom medication optimization, adjustment in the ICD detection zones, and supraventricular tachyarrhythmia discrimination algorithms were chosen. Catheter ablation associated with pharmacologic therapy was used as a therapeutic strategy in one patient with paroxysmal atrial fibrillation who received approximately 100 inappropriate shocks.

Korte et al[8] studied 20 young patients with a mean follow-up period of 51.0 ± 31.0 months, and reported 127 appropriate therapies in 15 (75%) patients and 112 inappropriate therapies in 10 (50.0%). In this study, supraventricular arrhythmias were the main cause of inappropriate therapies, occurring in 8 (40.0%) patients. Other studies reported that appropriate therapies occurred in 21.7% to 68.0% of the young patients, and inappropriate therapies in 30.4% to 40.0%[10,11]. In the present study, inappropriate shocks due to sinus tachycardia were also present, mainly in patients in a lower age range and were related to physical exercise in the majority of the cases. The other inappropriate shocks occurred in three patients: in one patient with Brugada Syndrome due to T-wave oversensing, for whom epicardial lead for left ventricular pacing and sensing was implanted via coronary sinus (Figure 3); in one patient with hypertrophic cardiomyopathy, for whom a specific function programming (decay delay) was performed, and one patient who presented noise in the ventricular channel due to lead displacement, which required repositioning.

Overall, the strategies used to minimize the incidence of inappropriate ICD therapies in young patients aim at: optimizing the use of medications such as beta-blockers and antiarrhythmic drugs; interrupting atrioventricular conduction or ablating atrial fibrillation using radiofrequency; and adjusting detection zones of tachycardias. Korte et al[8] used a single discrimination zone, high rate levels for the detection of VT and medication optimization in 80% of the patients. Catheter ablation was used when necessary. Ten Harkel et al[11] recommend the use of longer periods of detection and confirmation of tachycardia associated with high heart rates. The maximum heart rate achieved during exercise was calculated prior to hospital discharge. In order to prevent inappropriate T-wave sensing, alteration in the device sensing programming is made at the moment of implantation. Love et al[21] also recommend an elevation in the cut-off rate and in detection time, routine prescription of beta-blockers, and specific guidance to optimize compliance to medications. These authors question, however, the safety and validity of the use of supraventricular arrhythmia discrimination algorithms in the pediatric population.

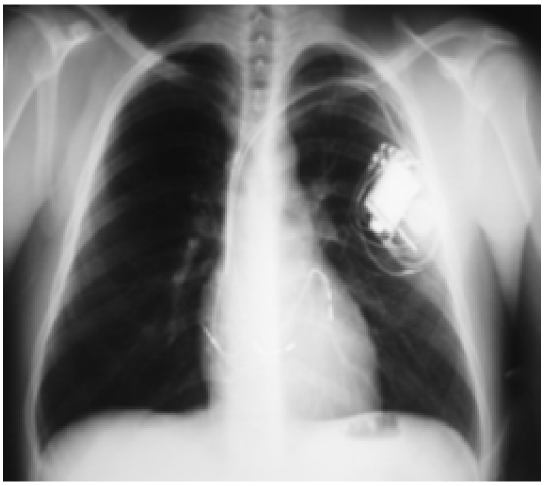

Fig. 3 *– Chest radiograph in posterior anterior view showing implantation of the epicardial lead in the left ventricle via the coronary sinus.*

Quality of life - Studies conducted in adults with ICD show a reduction in quality of life and alterations in the emotional status and in social relationships. In the pediatric and adolescent population, however, this issue remains not well defined[22,23].

In the present study, we sought to assess the quality of life and adaptation to the device, both in the emotional and social aspects. Quality of life was assessed after implantation of the device, using the SF-36 which is one of the most frequently used instruments to assess quality of life of individuals with cardiovascular diseases, and which may also be applied to the adolescent population[12].

Reduction in quality of life of patients with ICD has been mainly related to the incidence of shocks[22,24,25]. In the population studied, quality of life could not be compared between the patients who received and those who did not receive shocks, related to the small number of cases and the high incidence of shock therapies.

Mean values of the SF-36 domains found in the present population were compared to those found among healthy American individuals[13]. This comparison showed that our patients presented lower means in the Physical Functioning, Functional Capacity, Emotional Aspects, and Mental Health domains, whereas the means of the Social Functioning, Vitality, Bodily Painand General Health domains were higher. Although the ICD had interfered with quality of life, these data suggest that our population does not show a very significant reduction in quality of life when compared with healthy individuals.

Measurement of internal consistency is the most frequently used method to estimate reliability between the items of a scale. Within this category, the Cronbach's coefficient alpha is, undoubtedly, the most frequently used test. The values of this coefficient found in the present study showed that the SF-36 has a reasonably adequate reliability to measure quality of life of our population[26,27].

Some studies[22,28-30] defined that age under 50 years, occurrence of multiple shocks, and poor knowledge of the disease and the device are predictive factors of reduction in quality of life and of occurrence of psychosocial disorders in patients with ICD. Other factors pointed out also include professional difficulties or restrictions, as well as socioeconomic difficulties. These authors reinforce the need to establish educational strategies and multi-professional approach as an important intervention for an improved adaptation to the device.

Conclusions

The use of ICD was safe and effective in interrupting malignant arrhythmias in children and adolescents with a high risk of sudden cardiac death. The incidence of therapies delivered by the device, with appropriate and inappropriate shocks, was high and interfered in quality of life. The results of the present study reinforce the need for a specific multi-professional approach for this specific population.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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