Long-term follow-up of children and young adults treated with implantable cardioverter-defibrillator: the authors’ own experience with optimal implantable cardioverter-defibrillator programming

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Aims

Young implantable cardioverter-defibrillator (ICD) recipients present a high rate of inappropriate interventions. Some of them are caused by suboptimal pre-discharge programming of the device. There are conflicting data as regards antitachycardia pacing (ATP) effectiveness in children and young adults. We report our experience with ICD programming and a rate of complications during a 10 year follow-up.

Methods and results

We analysed the use and effectiveness of ATP and complications rate in 63 patients aged 6–21 years. Antitachycardia pacing (burst or ramp) was programmed ON in 14 patients (22%), 49 patients (78%) had only ventricular fibrillation (VF) therapy when discharged after implantation. The incidence of effective vs. ineffective or harmful ATP therapy: 5% of patients vs. 19% of patients differed significantly (P<0.05). Fourteen patients (22%) received ≥1 appropriate shock(s) for ventricular tachycardia/VF and 17 patients (27%) had one or multiple inappropriate therapy (IT). Inappropriate therapy resulted from T-wave over-sensing (nine patients), sinus tachycardia (three patients), fast atrial fibrillation (five patients), and lead insulation disruption (1%). Reprogramming of the device eliminated IT in all cases. There were 13 (21%) surgical complications. Serious psychological sequelae developed in 27 (43%) patients. There was one death during the follow-up period.

Conclusion

Antitachycardia pacing therapy is rarely effective and often harmful in young ICD recipients. In most patients, programming ICD for only VF therapy is sufficient. Routine pre-discharge programming against inappropriate interventions (especially T-wave over-sensing) helps to reduce the incidence of discharges during the follow-up. The incidence of complications and inappropriate therapies is high in young ICD recipients and affects 50% of patients.

Keywords

Implantable cardioverter-defibrillator • Antitachycardia pacing effectiveness • Complications • Long-term follow-up

Introduction

Ten-year experience with cardioverter-defibrillator implantations in the youngest patients in Poland allows us to resume the results of this therapy in our institution. A high percentage of inappropriate therapies, considerably higher than in adults, still remains an essential problem in the implantable cardioverter-defibrillator (ICD) patients.\(^1\) During the past few years there have been more implantations on the basis of primary prevention indications. At present, they constitute nearly half of all the
implantations. This approach is based on many clinical trials, among others ESC guidelines for the prevention of sudden cardiac death (SCD) published in 2006. However, in contrast with the group of adults there is lack of unambiguous indications in favour of ICD therapy in children. The latter group differs from that of adults in terms of aetiology but also of the extent of the heart damage.\(^1\)–\(^4\) Physical exercise and child’s growth often cause a lead failure, which often leads to a series of inappropriate discharges.

Psycho-social disturbances in this group of patients are another difficult problem, still underestimated. It has already been proved that the younger age of patients and more frequent ICD discharges are factors of the development of this type of complications which considerably diminish the quality of life (QoL). New implantation techniques in children have appeared recently,\(^5\)–\(^7\) and work on further miniaturization of the ICD and leads and on the development of the leadless ICD is in progress.

The present study is a continuation of the project undertaken 10 years ago in the form of a publication.\(^8\)

### Aims of the study

(i) Analysis of the occurrence of appropriate and inappropriate ICD therapies, frequency and time of elective device replacements in the study group.

(ii) Evaluation of the ATP effectiveness.

(iii) Analysis of the ICD therapy complications in the group of patients under 21 years of age.

(iv) Optimization of the device programming in order to minimize the number of unjustified interventions.

### Methods

Sixty-three patients aged 6–21 years (mean age at implant 13.5, SD 5.3) weighing 20–68 kg (mean 44, SD 13.6) were subjected to ICD therapy in the 2nd Department of the Coronary Artery Disease, The National Institute of Cardiology in Warsaw in 1996–06, among 598 implantations in the same period. Thirty-one patients (49%) were survivors of SCD, 32 patients (51%) implanted on the basis of primary prevention indications. The device placement was infraclavicular in 62 patients (transvenous leads), abdominal in 1 (epicardial lead). Forty-two (67%) were single chamber ICDs, 21 (33%) dual chamber. There were no biventricular devices in the analysed period. We implanted Belos VR, Phylax: 03, 03XM, Phylax: 06, 06XM, Microhylax Plus, Tachos devices from Biotronik, Photon: VR, DR, Epic and Atlas: VR, DR from St. Jude Medical and Microjewel II, Gem II: VR, DR from Medtronic. The mean follow-up was 64 months (range: 3–126). In the group analysed, the following three age categories can be distinguished: the youngest children (6–12 years, \(n = 30\)), adolescents (13–17 years, \(n = 22\)), and young adults (18–21 years, \(n = 11\)). Fifty patients (79%) had a normal or nearly regular ejection fraction (EF) (>50%), ventricular function was abnormal in 13 patients (21%). Nine patients (14%) had decreased EF (30–50) and only four children (6%) had low EF (<30%). The characteristics of the study group are shown in Table 1.

Psychological problems and QoL in the study group were analysed in a detailed questionnaire addressing important issues associated with living with an ICD. These findings were already published as a separate study.\(^9\) Psychological care was taken pre- and post-implantation by a psychologist who was a member of our team and performed the above mentioned questionnaire in the follow-up. Fifty-five subjects (out of 63) fulfilled the criteria for participation in this part of the study, and full data were obtained and analysed from 45 patients. The transvenous technique with an impulse generator placement under the pectoral muscle used routinely in our centre was described in detail in the paper published in 2005.\(^9\) We used the VVI system in the youngest children, assuming the possibility of its conversion to DDD once the child’s growth is terminated and when the device has to be replaced. It proved to be an effective and appropriate technique in the experience of other pediatric centres.\(^10\)–\(^11\)

The informed consent was obtained after the nature and possible consequences of this study were explained to every patient and/or parents in the studied group. The research protocol was approved by the locally appointed ethics committee. The study complies with Declaration of Helsinki.

### Statistics

Statistical analysis was performed using the SAS (version 8e) statistical package. For descriptive purposes, the data are presented as mean ± SD (continuous variables) or absolute frequencies and percentages where indicated (categorical variables). The Pearson’s \(\chi^2\), or Fisher’s exact test were used to compare proportions. All test procedures were two-sided with a \(P\)-value of <0.05 indicating statistical significance.

### Results

In all patients qualified for an ICD therapy, the implantation of the device was effective and feasible transvenously, except for one patient who required an epicardial electrode technique primarily with abdominal impulse generator placement after the correction of Ebstein anomaly with a previous implantation of an artificial tricuspid valve. During the observation period, 13 patients (21%) needed an elective ICD replacement (ERI) because of battery depletion. The mean time from the implantation to the ERI was 74.5 months (37–112). In five patients (8%) it was necessary to use a device with increased energy of the defibrillating impulse due to high defibrillation threshold (DFT) > 25 J during the first implant; \(n = 3\), or subsequent operation: \(n = 2\), due to a massive

### Table 1  Study group characteristics

<table>
<thead>
<tr>
<th>Aetiology, ICD type and indications for implantation.</th>
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<td>Study group characteristics</td>
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<tr>
<td>Hypertrophic cardiomyopathy (HCM)</td>
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<tr>
<td>Long QT syndrome (LQTS)</td>
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<tr>
<td>Primary VF</td>
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<tr>
<td>Arrhythmogenic right ventricular cardiomyopathy</td>
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<td>Dilated cardiomyopathy (DCM)</td>
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<td>Catecholaminergic polymorphic VT (CPVT)</td>
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<td>Congenital heart disease: Tetralogy of Fallot, Ebstein anomaly of the tricuspid valve</td>
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<td>Brugada syndrome</td>
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<tr>
<td>Myocardial infarction (vasospasm)</td>
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<tr>
<td>Primary prevention</td>
</tr>
<tr>
<td>Secondary prevention</td>
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<tr>
<td>VVI (including 1 VVI epicardial)</td>
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<td>DDD</td>
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\(n = 45\)
myocardial hypertrophy in the course of hypertrophic cardiomyopathy (HCM) and predicted progression of the disease.

During the follow-up period, which ranged from 3 to 126 months (mean 64), at least one appropriate ICD intervention caused by tachyarrhythmias [ventricular tachycardia (VT) or ventricular fibrillation (VF)] verified by the ICD Holter memory occurred in 14/63 (22%) patients in the study group. In 17/63 (27%) patients single or multiple inappropriate interventions (IT) occurred. Incidence of appropriate interventions in groups with a single chamber and dual chamber ICD was eight patients (19%) vs. six patients (29%), respectively, $P = 0.522$, ns. IT resulted from T-wave over-sensing (nine patients), sinus tachycardia (three patients), fast atrial fibrillation (FAF) (five patients), and lead insulation disruption (1%). Reprogramming of the device, i.e. VF detection zone for 220 b.p.m. in the case of FAF with fast ventricular response or sinus tachycardia with simultaneous inclusion or increasing of beta blocking agents caused effective control of unjustified shocks in all the above-mentioned cases. The implementation of the enhanced T-wave suppression programme (in the Biotronik devices) or decay delay: at least 60 ms (empirical increase in the range up to 125 ms) and/or threshold start-min: 62.5–75% (St. Jude Medical devices) eliminated IT in all patients with a T-wave over-sensing phenomenon. In one patient there occurred a series of IT in the mechanism of the lead insulation disruption. Reoperation made it possible to reconstruct the lead insulation damage within the ICD pocket. No IT were observed after the repair and the new lead was implanted when ERI time was reached. The assessment of antitachycardia therapy (ATP) efficacy and necessary changes of the ICD programme in the course of the follow-up are presented in Figure 1. In 6 (10%) patients it became necessary to reprogramme ATP to the OFF function, programmed primarily after implantation as a burst or ramp at the discretion of the implanting physician. Only in three patients (5% of the group) ATP was successful during four episodes of VT registered in the ICD memory. The ATP after reprogramming the device remained on in 8 (13%) patients. The comparison of incidence of inappropriate, appropriate therapies and a complication rate in the HCM group, non-HCM and in age groups is presented in Table 2. There was no significant difference between the incidence of IT in patients with single chamber and dual chamber devices: 8 (38%) vs. 9 (21%), respectively, $P = 0.160$.

In the follow-up, there was one death in the group. An 18-year-old woman with HCM died during an attempted laser extraction of the lead despite an immediate cardiosurgical intervention. Extensive laceration of the right atrium and superior vena cava was found during the operation. An indication for the removal of the lead was an increased stimulation threshold and ineffective internal defibrillation ($DFT > 30 \text{ J} = \text{maximum energy of this ICD}$) caused by the massive fibrous growth on the lead.

Early perioperative complications and necessary redo procedures (up to 30th day) appeared in five patients (8%). In one female patient an active fixation lead penetration of the right ventricle with symptomatic tamponade was found (an urgent cardiosurgical intervention with a good outcome.) In another case (massive hypertrophy in HCM), a high $DFT > 20 \text{ J}$ had to be re-operated with the reposition of the lead and the use of a high energy device. The other three patients required redo procedures due to the lead dislocation: one atrial and two ventricular with the

![Figure 1](https://academic.oup.com/eurheartj/article-abstract/124/9/1245/507101) Antitachycardia pacing effectiveness in the follow-up. FAF, fast atrial fibrillation; ST, sinus tachycardia.
loss of capture and an unacceptable increase of the stimulation threshold.

In two boys with catecholaminergic polymorphic ventricular tachycardia (CPVT) (3.2% of the group) 2 weeks after the implantation, a dramatic electrical storm developed suddenly. ~50 shocks appeared during several consecutive days in the mechanism of the polymorphic catecholaminergic tachycardia. The arrhythmia was terminated by high doses of metoprolol iv. In seven patients (11%), late complications appeared, i.e. infectious endocarditis on the ICD electrode requiring removal of the entire system by thoracotomy (n = 1), infection of the device pocket with the subsequent removal of the whole system and then after complete cure another implantation of a new system on the counter-lateral side (n = 2). In one patient, it came to a late asymptomatic (2 years after implantation) screw-in electrode penetration through the heart wall to pericardium (elective cardiosurgical procedure with the removal of the lead with a new system implantation on the opposite side). In one boy with DCM the lead displacement within the ventricle between control visits a few months after the implantation occurred with unsuccessful therapy and cardiac arrest. Cardio-pulmonary resuscitation (CPR) carried out by family members and then by paramedics with external defibrillation was successful without neurological deficits and subsequent implantation of a new active fixation lead. In one case high DFT 3 years after the implantation caused unsuccessful shocks with successful CPR requiring implantation of a new system with a high-energy device. One patient required a re-operation 2 years after the first implant due to a series of IT in the mechanism of the lead insulation disruption. Seven patients (11%) developed keloid formation.

Serious psychological and/or psychiatric problems (lack of the implanted device acceptance, fear of its functioning or more frequently fear of numerous discharges, depression, problems with memory, sleep or concentration) were found in 27 patients (43%). Six of them (10%) developed a serious anxiety neurosis. It required psychiatric pharmacological treatment in three patients. Anxiety associated with an ICD discharge was reported by 53 patients (84%) who experienced at least one shock. Phantom shocks appeared in 14 patients (22%).

We consecutively analysed patients’ serious complications and the combined rate (taking into account IT or ‘surgical complications’, i.e. thoracotomy, lead repositions, reimplantations, repairs, infections, or psychological and social problems) is 51% (32 patients). Some patients had several problems.

Discussion

The material presented above covers the largest number of patients with an ICD implanted at a young age from a single centre in Poland.

Complications

The high number of surgical complications in the study group (21%) exceeds the data from the biggest available meta-analysis (13%). The literature reports the number of complications in the group of children and young adults to be considerably larger than that in the group of adults (12.5 vs. 0.3%). Our experience is similar: in 2007, we performed the analysis of ICD therapy complications in our overall data base (598 patients implanted between 1995 and 2006). Infection was observed in five patients, lead-related problems in 19 patients and pocket revision in six patients so the combined complication rate is 30 patients (5%). The complication rate in young ICD recipients is 21 vs. 5% in adults in our centre during the same period (P < 0.05). The complication rate is high,although we think it is comparable to the data from other high volume centres. The history of ICD implantations in Poland started in 1995, so some of the complications were more frequent in the beginning. We are aware of the learning curve, especially in this pioneer field of ICD in pediatric patients. A smaller percentage of appropriate interventions (22%) in comparison with the world reports (33–60%) obviously results from a high percentage of prophylactic implantations when compared with the other groups described. The number of IT (27%) did not differ from the data published. It concerns around 30% of patients in most studies. Serious psychological problems in the study group, as they were found in 43% of patients, were similar when compared with other centres (38%). Some studies of QoL report assorted psychiatric disturbances affecting up to 87% of recipients. In our study, we found anxiety in 84% of patients, so the results are similar. Although frequent, these data seem to be underestimated, as patients treated in our centre come from the whole of the country and they often lack proper psychological care in their dwelling places. The psychological study was performed 10 years after the implantation in some patients when specific problems had gone with time. We currently pay more attention to more rigorous psychological care and proper psychological counselling in our centre because it might result in a reduction of anxiety, fear and depression after ICD.

Table 2 Comparison of incidence of inappropriate, appropriate therapies and complications rate in the HCM group, non-HCM and in age groups

<table>
<thead>
<tr>
<th></th>
<th>All (n = 63)</th>
<th>HCM (n = 33)</th>
<th>Others (n = 30)</th>
<th>P-value</th>
<th>Age 6–12 (n = 30)</th>
<th>Age 13–17 (n = 22)</th>
<th>Age 18–21 (n = 11)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inappropriate therapies</td>
<td>17 (27%)</td>
<td>10 (30%)</td>
<td>7 (23%)</td>
<td>0.5337</td>
<td>7 (23%)</td>
<td>7 (32%)</td>
<td>3 (27%)</td>
<td>0.7928</td>
</tr>
<tr>
<td>Appropriate therapies</td>
<td>14 (22%)</td>
<td>8 (24%)</td>
<td>6 (20%)</td>
<td>0.6858</td>
<td>5 (17%)</td>
<td>5 (23%)</td>
<td>4 (36%)</td>
<td>0.4074</td>
</tr>
<tr>
<td>Complications rate</td>
<td>13 (21%)</td>
<td>8 (24%)</td>
<td>5 (17%)</td>
<td>0.4580</td>
<td>5 (17%)</td>
<td>4 (18%)</td>
<td>4 (36%)</td>
<td>0.4115</td>
</tr>
</tbody>
</table>

HCM – hypertrophic cardiomyopathy group; others: non-HCM patients, all-study group; Complications rate combined: reinterventions, thoracotomy, lead dysfunction, i.e. lead repositions, repair, or reimplantations and infections (endocarditis or pocket).
implantation in young recipients. The most common cause of IT in the material under discussion was the phenomenon of T wave over-sensing. In our group there were a large number of patients with HCM (over a half of implants) mainly implanted for primary prevention which accounts for the fact of high percentage of IT. In the statistical analysis of complications, two cases of dramatic electrical storm shortly after implantations were not taken into consideration. It is hard to prove it but the lead mechanical effect (early depolarization?) after implantation cannot be excluded.

**Antitachycardia pacing effectiveness and the final programming of the device**

Results concerning the ICD programming method in young ICD recipients constitute a separate problem for discussion. In the majority of centres the programming of the VF detection zone exclusively (220/b.p.m.) is practised routinely except for situations when the documented VT is effectively interrupted by ATP. In young adults VT in the vast majority of cases is polymorphic in character. Therefore, it cannot be interrupted effectively by ATP. In our group the decision of ATP reprogramming to ‘off’ was caused by the lack of effectiveness of this form of therapy or what was even worse, ventricular rhythm acceleration during FAF or sinus tachycardia (harmful action of the device). In all these cases, ATP provoked a series of unnecessary ICD discharges. As regards its value in the period of distant observation, there are contradictory reports. In some of them high efficacy of this form of therapy was presented, but they covered groups consisting of a considerably smaller number of patients and different aetiology. Our results proved ATP to be rarely effective and what is even worse to be frequently ineffective or harmful in action. Therefore, we perform the simplest ICD programming (VF therapy only) with the application of ATP just in a few selected cases. The long-term follow-up of patients who experienced the ATP reprogramming to ‘off’ with no arrhythmic death over a period of 10 years proves that such a decision was correct. We did not try to programme extended delay as in the Pain Free Study in the observation period because results of this very important trial for the reduction of shocks were published in 2004.

After closing the data base for this publication, in two patients with multiple shocks, we programmed the fast ventricular tachycardia (FVT) detection zone defined within the VF zone (FVT via VF) for CL of 240–320 ms (250–188 b.p.m.). The first therapy in the FVT zone was a single ATP sequence (8-pulse burst pacing train at 88% of the VT CL) which terminated FVT successfully without a shock.

After the review of our own results, we changed the approach to ICD pediatric patients with important clinical implications. The current ICD pre-discharge programme in this population, except for a few cases with documented VT in the history, is as follows: we implant only programmable sensing devices in order to eliminate T-wave over-sensing when it appears. The VF zone is set as high as 222 b.p.m., with shock therapy only. Pacing is programmed to VVI 40’, the VT zone only to monitor. An exercise test is routinely done to assess sinus response in young patients and reveal circumstances for T-wave over-sensing. Currently, we routinely start therapy or increase the dose of beta-blocking agents for controlling conduction in atrio-ventricular junction whenever possible.

Creating a lead loop within the right atrium and vena cava proved to be effective. There was no dislocation of the lead due to the child’s growth in any case. Recommendations concerning this procedure in patients with HCM suggest the use of the DDD system in patients with paroxysmal atrial fibrillation and substantial intraventricular gradient exclusively. In all other cases of primary prevention the VVI system should be used, with a lower risk for lead related complications. No pacemaker syndrome was observed in the follow-up. We stopped to use proximal shocking coil in the pediatric and young adult patients.

**Conclusions**

(i) Endocardial ICD implantation in children and young adults is a safe and feasible technique in a vast majority of patients during a 10-year follow-up.

(ii) Incidence of surgical complications (as defined above)-21%; serious psychiatric sequelae (anxiety, fear or neurosis)-43%; and the rate of IT-27% (resulting in most cases from T-wave over sensing) is high in young ICD recipients.

(iii) Routine pre-discharge programming of ICD against IT (especially T-wave over-sensing) helps to reduce its incidence during the follow-up.

(iv) ATP therapy is rarely effective and is often reprogrammed to ‘off’ in young ICD recipients. The programming of ICD for VF only therapy is sufficient in the majority of patients.

(v) The above data should be taken into consideration in prophylactic ICD implantation when counselling patients, as the combined complications rate exceeds 50% of the study group.

**Study limitations**

This is a retrospective, observational study but obviously randomized studies concerning ICD implantations in the pediatric population are unacceptable. The study population is heterogeneous concerning the underlying heart disease and age.

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Medtronic, Vitatron, and Sorin, travelling grants from Medtronic, Biotronik, and Sorin, and lecturer’s fees from Medtronic. C.L.: no conflict. A.P.: Consultant for Biotronik, Principal Investigator in Medtronic’s sponsored trial, scientific grant from Biotronik, travelling grants from Medtronic, Biotronik, and SJM, and lecturer’s fees from Medtronic and SJM.

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