Sudden Cardiac Death and the Use of Implantable Cardioverter-Defibrillators in Pediatric Patients

Michael J. Silka, MD; Jack Kron, MD; Ann Dunnigan, MD; and Macdonald Dick II, MD; for the Pediatric Electrophysiology Society

Background. During the past decade, the implantable cardioverter-defibrillator (ICD) has emerged as the primary therapeutic option for survivors of sudden cardiac death (SCD). Investigation of the clinical efficacy of these devices has primarily assessed outcome in adults with coronary artery disease. The purpose of this cooperative, international study was to evaluate the impact of ICDs on the pediatric population of SCD survivors, based on an analysis of the clinical characteristics and outcomes of young patients who underwent ICD implantation following an episode of life-threatening ventricular tachycardia or resuscitation from SCD.

Methods and Results. An initial data base, established by contacting the manufacturers of the various commercially and investigationally available devices, identified 177 patients who were less than 20 years of age at the time of initial implantation of an ICD. With this data base as a reference, detailed responses were subsequently obtained from physicians involved in the care of 125 (71%) of these patients. The patients ranged in age from 1.9 to 19.9 years (mean, 14.5±4 years) and weighed 9.7-117 kg (mean, 44.6±14 kg). Of the 125 patients, 76% were survivors of SCD, 10% had drug refractory ventricular tachycardia, and 10% had syncope with heart disease and inducible sustained ventricular tachyarrhythmias. The most common types of associated cardiovascular disease were hypertrophic and dilated cardiomyopathies (54%), primary electrical diseases (26%), and congenital heart defects (18%). Ventricular function was abnormal in 46% of the patients. During a mean follow-up of 31±23 months, at least one ICD discharge occurred in 85 of the 125 (68%) patients. Seventy-three patients (59%) received at least one appropriate ICD discharge, and 25 patients (20%) had one or more spurious or indeterminate discharges. Duration of follow-up >24 months (p=0.001) and inducibility of a sustained ventricular arrhythmia (p=0.05) were correlated with appropriate ICD discharges. There were nine deaths during the study period: five sudden, two due to recurrent ventricular arrhythmias, and two related to congestive heart failure. Abnormal ventricular function (p=0.002) and prior ICD discharge (p=0.01) were univariate correlates of patient mortality; by multivariate logistic regression, abnormal ventricular function was the only significant correlate of death (p=0.005). By actuarial analysis, the estimated overall post-ICD implant survival rates at 1, 2, and 5 years were 95%, 93%, and 85%, respectively. The corresponding sudden death-free survival rates were 97%, 95%, and 90%.

Conclusions. Pediatric patients resuscitated from SCD appear to remain at risk for recurrence of life-threatening tachyarrhythmias. During a mean follow-up of 31 months, the ICD provided an effective therapy for such arrhythmias in the majority of patients in this study. Following ICD implant, impaired ventricular function was the primary factor correlated with mortality. The patterns of ICD discharge observed in young patients and, thus, inferred risk of recurrent life threatening arrhythmias are similar to those of adult survivors of SCD. Thus, the use of ICDs in pediatric patients, with implant selection criteria similar to adults, appears valid. (Circulation 1993;87:800-807)

Key Words • ventricular arrhythmia • pediatric cardiology • congenital heart disease • cardiomyopathy • cardioverter-defibrillator • sudden cardiac death • children

During the past decade, advances in the development of implantable cardioverter-defibrillators (ICDs) have significantly altered both the approach to and prognosis for patients resuscitated from sudden cardiac death (SCD).1-4 However, because SCD represents a common final mode of expression for many diverse types of cardiovascular disease, evaluation of the use of ICDs requires reference to the differing substrates of SCD.5,6 This type of analysis may be most relevant in young patients, in whom SCD is an infrequent event (one to eight events per 100,000 patient-years).7,8 This study examines outcomes in pediatric patients who underwent ICD implantation in the past decade at six international centers. The purpose of this cooperative, international study was to evaluate the impact of ICDs on the pediatric population of SCD survivors, based on an analysis of the clinical characteristics and outcomes of young patients who underwent ICD implantation following an episode of life-threatening ventricular tachycardia or resuscitation from SCD.

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years\textsuperscript{7,8}\right] and in whom SCD is rarely associated with myocardial ischemia or prior infarction.\textsuperscript{9}

In contrast to the proliferation of clinical studies analyzing the use of ICDs in adults resuscitated from or at risk for SCD,\textsuperscript{10} only a few, descriptive studies have been published regarding the use of ICDs in young patients.\textsuperscript{11-13} Thus, this multicenter, international survey was initiated under the auspices of the Pediatric Electrophysiology Society to analyze the patient characteristics, device utilization, and prognostic factors related to outcome in young patients who had undergone ICD implantation following an episode of life-threatening ventricular tachycardia or resuscitation from SCD.

Methods

Patient Identification

For the purposes of initial patient identification, a primary data base was established by reference data obtained from the manufacturers of the commercially and investigationally available ICDs. Admission criteria included patient age less than 20 years at the time of initial ICD implant and ICD implantation between October 1980 and October 1991. To maintain confidentiality, patients were referred to only by age; sex; and the date of physician involved in, and institution of implant. A total of 177 subjects were thus identified.

Detailed responses were subsequently obtained regarding 125 of the 177 (71\%) patients, who are the subjects of this analysis. A total of 102 physicians, representing seven countries, contributed responses to this study. Data were acquired as follow-up of a preliminary study of use of ICDs in 40 young patients,\textsuperscript{13} a survey of the experience of members of the Pediatric Electrophysiology Society in the use of ICDs, or a survey of physicians of patients identified by the manufacturers' data base who were not identified by the two prior methods. At least two attempts were made to contact the physician of each patient. A standardized, open-ended data entry format, designed to maximize participation, was used in each case. Specific information was requested regarding associated cardiovascular disease, ventricular function, electrophysiological testing, profile of ICD discharges, complications, and patient survival.

Definitions

Sudden cardiac death was considered death occurring within 1 hour of onset of symptoms in a previously stable patient, an unwitnessed death, or death during sleep.\textsuperscript{14} Tachyarrhythmic nonsudden cardiac death was death associated with sustained ventricular arrhythmias, persisting or recurring over a period of time exceeding 1 hour before death.\textsuperscript{15} Cardiac nonarrhythmic death was witnessed death of a cardiac nature, other than sudden or tachyarrhythmic nonsudden causes.

Appropriate discharge was an ICD shock, in the opinion of the responding physician, delivered in response to a ventricular arrhythmia. These shocks were either associated with resolution of the symptoms of presyncope or syncope, presumed to be associated with a spontaneous sustained ventricular arrhythmia, or documented by cardiac monitor to be associated with a sustained ventricular arrhythmia.\textsuperscript{16} Episodes of ventricular tachycardia terminated by overdrive pacing were excluded from analysis of ICD performance due to the limited availability and utilization of this option.

Spurious discharge was an ICD shock, in the opinion of the responding physician, delivered for reasons other than a sustained ventricular arrhythmia. These shocks were either not preceded by symptoms of presyncope or syncope or were documented during cardiac monitoring not to be associated with a sustained ventricular arrhythmia.\textsuperscript{16} Indeterminate discharge was an ICD shock that could not be classified as either appropriate or spurious by the above definitions.

In statistical analysis, data are presented as range and mean±SD. Categorical differences between groups were evaluated by $\chi^2$ or Fisher's exact test. Differences between group means were compared by the unpaired $t$ test. Actuarial survival analysis was estimated by Kaplan-Meier life table methods, with data reported as ±SEM. Patients referred for orthotopic heart transplantation or who underwent ICD removal were included among survivors until the time of transplant or device explant, at which time they were censored from continued analysis. The observed proportion of categorical patient characteristics in this pediatric cohort was compared with the established frequencies of such variables in the adult population of ICD patients using the binomial test for normal distribution (Table 1). The potential correlation between multiple variables and patient outcome was analyzed with stepwise multivariate logistic regression.

Results

Demographics

Implant data, acquired from manufacturer's records, identified 65 female and 112 male patients. Patient age at time of ICD implant ranged from 22 months to 19.9 years (mean, 15.2±3 years) and weight ranged from 9.7 to 115 kg (mean, 48±16.5 kg). The year of initial ICD implant for all 177 patients is illustrated in Figure 1. Compared with the total group of 177 patients, there were no statistically significant differences in the demographics of the study group of 125 patients, comprised of 43 female and 82 male patients, with a mean age of 14.5±4 years and a mean weight of 44.6±14 kg.

Implant Indications

The most common reported indication for ICD implantation was aborted sudden cardiac death, due to ventricular fibrillation or hypotensive ventricular tachycardia, in 95 of 125 (76\%) of the patients. An ICD was implanted in 13 patients due to drug refractory ventricular tachycardia and in 12 patients due to unexplained syncope in association with both structural heart disease and inducible sustained ventricular arrhythmias during programmed stimulation. In five patients, an ICD was implanted due to the presence of structural heart disease and a history of SCD in immediate family members with the same form of heart disease (Table 2).

Associated Cardiovascular Disease

Three types of cardiovascular disease were present among the 125 patients. Cardiomyopathy was the most common (58\%)—hypertrophic cardiomyopathy in 44
TABLE 1. Characteristics of Pediatric Patients Versus Adult Patients

<table>
<thead>
<tr>
<th></th>
<th>Pediatric patients (n)</th>
<th>Adult series (n)</th>
<th>Reference</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>63% (177)</td>
<td>81% (9,807)</td>
<td>41</td>
<td>0.001</td>
</tr>
<tr>
<td>Coronary artery disease</td>
<td>1.6% (125)</td>
<td>72% (1,681)</td>
<td>22</td>
<td>0.001</td>
</tr>
<tr>
<td>VT/VF inducible</td>
<td>60% (92)</td>
<td>73% (1,227)</td>
<td>22</td>
<td>0.004</td>
</tr>
<tr>
<td>Abnormal ventricular function</td>
<td>46% (101)</td>
<td>78% (889)</td>
<td>22</td>
<td>0.001</td>
</tr>
<tr>
<td>Implant mortality</td>
<td>0.8% (125)</td>
<td>2.5% (889)</td>
<td>22</td>
<td>NS</td>
</tr>
<tr>
<td>Antiarrhythmic drugs</td>
<td>84% (125)</td>
<td>66% (889)</td>
<td>22</td>
<td>NS</td>
</tr>
<tr>
<td>Amiodarone</td>
<td>19% (125)</td>
<td>27% (889)</td>
<td>22</td>
<td>0.003</td>
</tr>
<tr>
<td>Mean follow-up (months)</td>
<td>31 (125)</td>
<td>20 (889)</td>
<td>22</td>
<td>NA</td>
</tr>
<tr>
<td>Appropriate ICD shock</td>
<td>59% (125)</td>
<td>56% (889)</td>
<td>22</td>
<td>NS</td>
</tr>
<tr>
<td>SCD free at 1 year</td>
<td>97% (99)</td>
<td>98% (5,392)</td>
<td>41</td>
<td>NS</td>
</tr>
<tr>
<td>SCD free at 5 years</td>
<td>90% (20)</td>
<td>95% (292)</td>
<td>41</td>
<td>NS</td>
</tr>
<tr>
<td>Total survival at 1 year</td>
<td>95% (99)</td>
<td>95% (5,392)</td>
<td>41</td>
<td>NS</td>
</tr>
<tr>
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<td>85% (20)</td>
<td>87% (292)</td>
<td>41</td>
<td>NS</td>
</tr>
</tbody>
</table>

VT/VF, ventricular tachycardia or fibrillation; NA, not applicable; NS, not statistically significant (p>>0.2 in all cases); SCD, sudden cardiac death.

patients and idiopathic dilated cardiomyopathy in 22 patients. In addition, two patients each had restrictive and ischemic cardiomyopathies. Primary electrical diseases (no structural heart disease) were reported in 26% of the patients, as either idiopathic ventricular fibrillation (19 patients) or the prolonged QT syndrome (14 patients). Congenital heart disease was present in 22 patients (18%), with postoperative Mustard repair of d-transposition of the great arteries, postoperative repair of tetralogy of Fallot, and aortic valvotomy or valve replacement for aortic stenosis accounting for 19 of these 22 patients.

Ventricular Function

Ventricular function was evaluated by angiographic, echocardiographic, or radionuclide techniques. Due to the multiple methods employed and lack of standardization, global ventricular function was categorized as normal, decreased, hyperdynamic obstructive, or restrictive associated with severe hypertrophy. Estimates of ventricular function were available for 101 of the 125 patients and were reported as normal in 54%, decreased in 40%, and hyperdynamic/restrictive in 6% of the patients.

Electrophysiological Testing

The results of electrophysiological testing prior to ICD implantation were reported for 92 of the 125 patients and idiopathic dilated cardiomyopathy in 22 patients. In addition, two patients each had restrictive and ischemic cardiomyopathies. Primary electrical diseases (no structural heart disease) were reported in 26% of the patients, as either idiopathic ventricular fibrillation (19 patients) or the prolonged QT syndrome (14 patients). Congenital heart disease was present in 22 patients (18%), with postoperative Mustard repair of d-transposition of the great arteries, postoperative repair of tetralogy of Fallot, and aortic valvotomy or valve replacement for aortic stenosis accounting for 19 of these 22 patients.

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TABLE 2. Clinical Characteristics

<table>
<thead>
<tr>
<th></th>
<th>Patients</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Indication for ICD</td>
<td></td>
<td></td>
</tr>
<tr>
<td>SCD survivor</td>
<td>95</td>
<td>76</td>
</tr>
<tr>
<td>Drug refractory VT</td>
<td>13</td>
<td>10</td>
</tr>
<tr>
<td>Syncope, +EP test</td>
<td>12</td>
<td>10</td>
</tr>
<tr>
<td>Familial SCD</td>
<td>5</td>
<td>4</td>
</tr>
<tr>
<td>Cardiovascular disease</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cardiomyopathy</td>
<td></td>
<td>58</td>
</tr>
<tr>
<td>Hypertrophic</td>
<td>44</td>
<td></td>
</tr>
<tr>
<td>Dilated</td>
<td>22</td>
<td></td>
</tr>
<tr>
<td>Restrictive</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Ischemic</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Primary electrical</td>
<td></td>
<td>26</td>
</tr>
<tr>
<td>Idiopathic VF</td>
<td>19</td>
<td></td>
</tr>
<tr>
<td>Long QT</td>
<td>14</td>
<td></td>
</tr>
<tr>
<td>Congenital heart</td>
<td></td>
<td>18</td>
</tr>
<tr>
<td>D-TGA</td>
<td>9</td>
<td></td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>L-TGA</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Anomalous PV return</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>VSD-PHT</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Ventricular function</td>
<td></td>
<td>54</td>
</tr>
<tr>
<td>Normal</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hyperdynamic</td>
<td></td>
<td>6</td>
</tr>
<tr>
<td>Decreased</td>
<td></td>
<td>40</td>
</tr>
</tbody>
</table>

VT, ventricular tachycardia; +EP test, inducible sustained ventricular arrhythmia; VF, ventricular fibrillation; TGA, transposition of the great arteries, where D=deoro and L=levo; PV, pulmonary venous; VSD-PHT, ventricular septal defect with pulmonary hypertension.
patients. Programmed ventricular stimulation was performed according to each institution's protocol and varied between two and four extrastimuli and one to three drive cycles. Sustained ventricular tachyarrhythmias were induced in 56 patients (60%); ventricular fibrillation was the most frequent arrhythmia, induced in 35 patients. Sustained monomorphic ventricular tachycardia was induced in 15 patients, and sustained polymorphic ventricular tachycardia was induced in six patients.

Operative Complications
Operative or perioperative complications were reported in 16 of 125 (13%) patients. The most common complication, in five patients, was a defibrillation threshold exceeding 20 J during predischarge testing, which required surgical reconfiguration of the defibrillation patches or electrodes. Four patients had ICD pocket wound infections requiring device removal, and two had device erosion (one cutaneous, one peritoneal). Three patients had pericardial effusions requiring pericardiocentesis, and there was one pneumothorax.

There was one intraoperative fatality reported in the 125 patients. This occurred in a 10-year-old boy with a restrictive cardiomyopathy and recurrent SCD. Ventricular fibrillation, which was induced intraoperatively at the time of defibrillation threshold testing, was intractable and could not be terminated. There were three other fatalities within 30 days of ICD implantation,17 of which one was sudden, one due to recurrent ventricular fibrillation, and one due to severe congestive heart failure.

Concurrent Antiarrhythmic Therapy
Antiarrhythmic drug therapy was prescribed in 84% of the patients following ICD implantation. The most common therapy was β-blockade (47%), either alone (41%) or in combination with a class 1 drug (6%). A class I drug was utilized in 19%, a class III drug in 20%, and a class IV drug in 4%. Concomitant antiarrhythmic medications were either not initiated or discontinued in 16% of the 125 patients following ICD implant.

Analysis of ICD Performance
The median duration of patient follow-up, from the time of initial ICD implantation until the closure date of this study, was 31±23 months (range, 0.1–102 months). During this study interval, at least one ICD discharge occurred in 85 of the 125 patients (68%). In 73 patients, at least one ostensible appropriate ICD discharge was reported. In addition, one or more spurious or indeterminate discharges were reported in 25 patients (20%). The number of appropriate ICD discharges for the 73 patients ranged from one to 42, with an average of 7.8±4 discharges among patients receiving appropriate ICD discharges. The mean interval from time of ICD implant to first appropriate ICD discharge was 12.2±9 months, with a range of 1 day to 47 months. The majority of spurious or indeterminate discharges occurred early in the postimplant period, at a mean interval of 3.9±2.2 months.

Analysis of ICD Discharges
Four variables potentially related to appropriate ICD discharges were analyzed: duration of follow-up, type of associated cardiovascular disease, ventricular function, and responses to programmed ventricular stimulation.

By actuarial analysis, the incidence of at least one appropriate ICD shock at 1- and 2-year intervals following ICD implant was estimated to be 30±7% and 50±11%, respectively. The incidence of appropriate ICD shocks increased to 59±13% at 36 months after implant. Appropriate ICD discharges were reported in 50 of the 66 patients followed for intervals greater than 24 months compared with 23 of 59 patients followed for 24 months or less (p=0.001).

The prevalence of an appropriate ICD discharge was next evaluated for the three major substrates of SCD: cardiomyopathy, primary electrical disease, and congenital heart disease—and their major subtypes. No statistically significant difference in the prevalence of ICD discharges was identified among the major types of cardiovascular disease (Figure 2). The only subtype with less than a 50% incidence of appropriate ICD discharge was idiopathic ventricular fibrillation.

As noted, due to the differing methods used in the assessment of ventricular function, patients were grouped by estimates of ventricular function as normal, decreased, or hyperdynamic/restrictive. Appropriate ICD discharges were reported in 27 of 40 patients with decreased function, four of six patients with restrictive or hyperdynamic function, and 28 of 55 patients with normal ventricular function (p=NS).

Three different categories of responses to programmed ventricular stimulation were evaluated in relation to appropriate ICD discharges. Appropriate ICD discharges were reported in 17 of 35 patients with inducible ventricular fibrillation, 15 of 21 patients with sustained ventricular tachycardia, and 12 of 36 patients with no inducible sustained ventricular arrhythmia. The inducibility of a sustained ventricular arrhythmia was correlated with appropriate ICD discharge (p=0.05).
Patient Survival Following ICD Implantation

During the mean follow-up of 31 months, there were nine deaths among the 125 patients. Five deaths were sudden and thus presumably arrhythmic in origin. This includes one patient with documented ICD battery depletion, who refused generator replacement. Two deaths were documented to be due to recurrent ventricular arrhythmias, with multiple appropriate ICD discharges (tachyarrhythmic nonsudden cardiac death). Two patients died due to progression of congestive heart failure associated with their underlying cardiovascular disease. In addition, five patients underwent orthotopic heart transplantation, with ICD therapy used as a "bridge" to transplant. One patient, censored from analysis at the time of transplant, subsequently died due to allograft rejection.

Two to 17 appropriate ICD discharges were reported in each of the patients who died. Four deaths were in the hospital and documented to be either due to recurrent or refractory ventricular fibrillation in three patients. The fourth death was due to low cardiac output and progressive bradycardia following a wide QRS complex tachycardia that required multiple ICD discharges for termination. The forms of cardiovascular disease in the nine patients who died were hypertrophic cardiomyopathy (n=4), dilated cardiomyopathy (n=3), restrictive cardiomyopathy (n=1), and congenital heart disease (n=1). Excluding the one patient with documented ICD battery depletion, there were no reported device failures among the eight other patients who died.

Decreased global systolic function was reported in seven of the nine patients who died, whereas two had severe hypertrophic or restrictive cardiomyopathies. There were no deaths among the 55 patients reported to have "normal" ventricular function in contrast to nine deaths among the 46 patients with abnormal ventricular function (p=0.002). Also, no deaths occurred among the 52 patients who had not received appropriate shocks in contrast to nine deaths among the 73 patients who had received such shocks (p=0.01).

No correlation was observed between mortality and responses to programmed stimulation. Of the nine patients who died, three had inducible ventricular fibrillation, two had inducible sustained ventricular tachycardia, and four had no inducible sustained arrhythmia.

Following univariate analysis, multiple logistic regression was performed to evaluate the potentially confounding factors related to mortality. By such analysis, abnormal ventricular function was the only significant risk factor for death following ICD implantation (p=0.005).

Estimates of Patient Survival

Actuarial survival was estimated for both total and sudden cardiac death-free intervals. The estimated total patient survival rates at 1, 2, and 5 year intervals were 95%, 93%, and 85%, respectively. The corresponding sudden death-free survival rates were 97%, 95%, and 90% (Figure 3). In an attempt to estimate the risk of recurrent sudden death, and thus potential impact of the ICD on survival, projected sudden death-free survival in the absence of ICD therapy was also estimated, with the first appropriate ICD discharge as the defined end point. Using this paradigm for analysis, sudden death-free survival rates (freedom from appropriate ICD shocks) of 70% at 12 months and 50% at 24 months were projected (Figure 4).

Discussion

The infrequency of SCD in pediatric patients contrasts with the absolute magnitude of this problem in adults. Given the large population base of adult SCD survivors, multiple cooperative studies have documented progress in the evaluation and treatment of these patients.18-20 However, the relevance of these

![Figure 3](http://circ.ahajournals.org/)

**FIGURE 3.** Plot of life-table analyses of both total and sudden cardiac death (SCD)-free survival, estimated from the time of initial implantable cardioverter-defibrillator implant. The divergence between the lines reflects nonsudden cardiac mortality. The numbers in parentheses below the lines refer to the number of patients with the corresponding duration of follow-up.

![Figure 4](http://circ.ahajournals.org/)

**FIGURE 4.** Plot of life-table analysis of survival free of appropriate implantable cardioverter-defibrillator shocks compared with total patient survival. By this analysis, the estimated "sudden death-free survival" rates (freedom from tachyarrhythmia termination by the implantable cardioverter-defibrillator) were 70% at 1 year, 50% at 2 years, and 40% at 3 years of follow-up from the time of initial implantation of the implantable cardioverter-defibrillator. By comparison, the actual total patient survival rates at these corresponding intervals were 95%, 93%, and 88%.
clinical studies to specific subsets of young patients is uncertain.

This multicenter study of pediatric patients demonstrates that over a mean follow-up period of 31 months, the ICD provided an effective therapy for recurrent ventricular arrhythmias in the majority of young patients resuscitated from an initial episode of SCD. Furthermore, despite the marked differences in patient characteristics, many similarities were observed regarding both the use of ICDs and prognosis for young patients resuscitated from SCD and data that have been established for adult survivors of SCD (Table 1). However, in a group of patients who would otherwise survive for many decades, this experience cannot be considered definitive.

Three central issues were examined regarding the role of ICD therapy for young survivors of SCD: first, the risk of a recurrent life-threatening ventricular arrhythmia; second, the related issue of appropriate ICD discharges, and thus, the potential impact on survival; and third, overall patient survival, and factors related to outcome.

Regarding the risk of recurrent ventricular arrhythmias, this study may allow certain inferences regarding the frequency of such life-threatening events in young survivors of SCD. Excluding patients who underwent ICD implantation due to a family history of SCD, 80% of patients with greater than 24 months' follow-up after device implantation were reported to have experienced at least one appropriate ICD discharge. Because these ICD discharges were responses to apparent life-threatening arrhythmias, this suggests a high rate of recurrence of such arrhythmias in young patients. However, until devices routinely allow storage and telemetry of the electrograms resulting in ICD responses, conclusions regarding the appropriateness of a discharge, and thus clinical occurrences of ventricular tachycardias, will be based on inferences from the clinical circumstances of the ICD shock.21

Further confounding the evaluation of these apparent life-threatening events was the limited ability to reliably identify these arrhythmias during electrophysiological testing. The lower frequency of tachycardia induction in this study (60%) compared with inducibility rates of 70–80% in adult survivors of SCD may be related to the prevalence of nonischemic forms of cardiovascular disease in young patients.24–26 Noninducibility of sustained ventricular arrhythmias during programmed ventricular stimulation would appear to be of limited prognostic significance in young SCD survivors. Conversely, ventricular fibrillation, the most common induced sustained ventricular arrhythmia in this study, may represent a nonspecific response to an aggressive ventricular stimulation protocol.27,28 Comprehensive electrophysiological testing remains indicated in potential ICD recipients to evaluate the supraventricular as well as ventricular arrhythmias29,30 and to guide selection of the most appropriate therapeutic modality. However, when electrophysiological testing cannot be used to accurately predict efficacy of other therapies, ICD implantation would appear to be indicated in pediatric survivors of SCD, as in adult survivors.31,32

The second issue, related to recurrence of ventricular arrhythmias, is the percentage of patients experiencing appropriate ICD shocks. Based on actuarial analysis, it is projected that given current indications for ICD implantation, approximately 60% of young SCD survivors who receive an ICD will experience an “appropriate” ICD discharge within 36 months of follow-up (Figure 4). This analysis may underestimate the impact of ICD therapy, as episodes of ventricular tachycardia terminated by overdrive pacing methods were not included in this analysis.33 The mean interval from time of ICD implant to first appropriate shock in patients receiving such shocks was 12.2±6 months.

Estimates regarding the prevalence of spurious or indeterminate ICD shocks are less certain due to the uncertainty regarding the clinical significance of many such discharges, resulting in incomplete documentation and responses to this survey-based study. Based on the available data, the majority of inappropriate or spurious shocks were reported within a 4-month interval following ICD implant (mean, 3.9±2.2 months). Duration of follow-up was the primary variable correlated with appropriate ICD discharge. It is notable that appropriate ICD discharges were reported in each of the nine patients who died during follow-up and in four of five patients prior to referral for orthotopic heart transplantation. This finding is concordant with adult studies suggesting a better prognosis for patients who do not receive ICD shocks after implant compared with those who do receive appropriate shocks.34,35

Regarding survival following ICD implantation, impaired ventricular function was the principal correlate of mortality in the young patients in this study. There were no deaths among the 55 patients reported to have normal ventricular function in this study in contrast to nine deaths among the 46 patients with abnormal ventricular function. Similar data implicating the role of impaired ventricular function as the primary determinant of long-term survival are established for adult ICD recipients.36,37 It should be noted that the prevalence of appropriate ICD shocks was similar between the two groups, occurring in 51% of patients with normal and 67% of patients with abnormal ventricular function. Quantification of the degree of ICD benefit in patients with impaired ventricular function remains controversial.38 Analyses of the shock incidence data in this study suggests a significant short-term (<12 month) improvement in the survival of patients with abnormal ventricular function. However, impaired ventricular function remains a long-term risk factor for an adverse outcome following ICD implantation.

This study is limited by multiple methodological aspects that require emphasis. The fundamental design of this study is that of a retrospective survey of over 100 physicians, each with inherently different methods of patient evaluation, criteria for ICD implantation, and protocols of follow-up. Given the diversity of practices, incomplete data were often rendered regarding patient parameters, ventricular function, or electrophysiological testing. Such imprecision clearly invokes the need for prospective study of the use of ICDs in young patients. However, as an 11-year interval was required to generate the data on these 125 patients, many years will be required to complete a prospective evaluation in the use of ICDs in young patients.

The second major limitation is that of selection of end points for analysis. The limited ability to reliably classify ICD discharges as appropriate, spurious, or indetermi-
nate has been previously discussed and continues to result in controversy regarding the methods of analysis of ICD function and impact on patient survival. Given these uncertainties and limitations, the analyses in this study were performed with appropriate shock-free survival, sudden death–free survival, and total patient survival as end points for analysis.

Conclusions

The ICD clearly represents an effective form of therapy for recurrent ventricular tachyarrhythmias in young survivors of SCD. The main determinant of long-term outcome following ICD implantation appears to be ventricular function. In young patients with ventricular dysfunction and lethal ventricular arrhythmias, the ICD may represent a form of palliation, with potential for use of the ICD as a bridge to transplantation. The ICD would appear to improve the short-term prognosis (<24 months) in these patients, although long-term survival (>60 months) may not be significantly improved. Conversely, in young SCD survivors with relatively normal ventricular function, the ICD would appear effective on both a short- and a long-term basis. Prospective study with long-term follow-up will be required to ascertain the prognosis for young survivors of SCD and the role of ICD therapy. Until these studies have been completed, this analysis would support the use of ICDs in young patients resuscitated from SCD, with implantation criteria similar to those currently established for adult survivors of SCD.

Appendix

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