Time Dependence of Risks and Benefits in Pediatric Primary Prevention Implantable Cardioverter-Defibrillator Therapy

Elizabeth S. DeWitt, MD; John K. Triedman, MD; Frank Cecchin, MD; Doug Y. Mah, MD; Dominic J. Abrams, MD, MBBS; Edward P. Walsh, MD; Kimberlee Gauvreau, ScD; Mark E. Alexander, MD

Background—Implantable cardioverter defibrillators (ICDs) used to prevent sudden cardiac arrest in children not only provide appropriate therapy in 25% of patients but also result in a significant incidence of inappropriate shocks and other device complications. ICDs placed for secondary prevention have higher rates of appropriate therapy than those placed for primary prevention. Pediatric patients with primary prevention ICDs were studied to determine time-dependent incidence of appropriate use and adverse events.

Methods and Results—A total of 140 patients aged <21 years (median age, 15 years) at first ICD implantation at Boston Children’s Hospital (2000–2009) in whom devices were placed for primary prevention were retrospectively identified. Demographics and times to first appropriate shock; adverse events (including inappropriate shock, lead failure, reintervention, and complication); generator replacement and follow-up were noted. During mean follow-up of 4 years, appropriate shock occurred in 19% patients and first adverse event (excluding death/transplant) occurred in 36%. Risk of death or transplant was ≈1% per year and was not related to receiving appropriate therapy. Conditional survival analysis showed rates of appropriate therapy and adverse events decrease soon after implantation, but adverse events are more frequent than appropriate therapy throughout follow-up.

Conclusions—Primary prevention ICDs were associated with appropriate therapy in 19% and adverse event in 36% in this cohort. The incidence of both first appropriate therapy and device-related adverse events decreased during longer periods of follow-up after implantation. This suggests that indications for continued device therapy in pediatric primary prevention ICD patients might be reconsidered after a period of nonuse. *(Circ Arrhythm Electrophysiol. 2014;7:1057-1063.)*

Key Words: arrhythmias, cardiac ▪ death, sudden, cardiac ▪ defibrillators, implantable ▪ pediatrics

Implantable cardioverter defibrillator (ICD) therapy is increasingly being used in pediatrics for primary and secondary prevention of sudden cardiac arrest (SCA). When ICDs were initially introduced into pediatric practice, most implants were reserved for secondary prevention of SCA. More recent pediatric data show that as many as 80% of devices are currently implanted for primary prevention of SCA.2,3 These devices are implanted in this population for a heterogeneous group of diseases, including cardiomyopathies, cardiac channelopathies, and palliated congenital heart disease. Collectively the prevalence of these diseases is low, contributing <1% to the total volume of ICD implants in the United States.4-5

Clinical Perspective on p 1063

Approximately 25% of ICDs implanted in children have provided appropriate therapy4,4 with higher rates of appropriate therapy in devices placed for secondary prevention.2 Offsetting this benefit are several difficulties with ICD therapy, which have been explored in both pediatric and adult populations. Rates of inappropriate shocks and other device-related adverse events are significant, and particularly so in children. Factors that may amplify these problems in the pediatric population include size mismatch between devices and patients, somatic growth, the frequent occurrence of sinus tachycardia and supraventricular tachycardia, and the need for the use of nonstandard hybrid and epicardial approaches for device placement.4-6 Pediatric patients with ICDs also report decreased psychosocial and physical quality of life scores when compared with peers with other chronic illness, and such patients may avoid activities for fear of getting shocked.7-9 These problems constitute a significant burden of therapy, particularly in primary prevention patients who by definition have had no previous personal event that underlines the specific need for an ICD to the patient and family.

This study reviews this center’s recent institutional experience to define the temporal occurrence of favorable (appropriate shock) and unfavorable (death, inappropriate shock, other ICD-related adverse event, and reintervention) outcomes
among pediatric patients receiving a first ICD implant for primary prevention of SCA.

**Methods**

**Study Design**

Patients who underwent initial ICD placement at this institution from 2000 to 2009 were retrospectively identified through departmental databases. Approval for data review was obtained from the Committee for Clinical Investigation at Boston Children’s Hospital.

**Study Population**

Children followed by the Arrhythmia Service of the Department of Cardiology at Boston Children’s Hospital were considered for study. Included patients were aged <21 years at implantation and had no previous history of aborted SCA. Only patients without previous history of ICD therapy were included in the study population.

**Measurements**

Demographics, ICD indications, pacing indications, method of ICD placement (epicardial, hybrid, or transvenous), and device/lead characteristics (number and configuration of leads, manufacturer, cardiac resynchronization therapy) were collected from the patient’s medical record. Underlying disease was classified as primary electric disease (long-QT syndrome, catecholaminergic polymorphic ventricular tachycardia [VT], and Brugada syndrome), cardiomyopathy (hypertrophic, dilated, arrhythmogenic right ventricular cardiomyopathy, and other), congenital heart disease, no underlying diagnosis (which included patients with history of sustained, nonsustained, or inducible ventricular tachycardias), or “other.” The category “other” included patients who were postmortem heart transplant or had left ventricular aneurysm or rhabdomyosarcoma.

A patient was classified as having a primary prevention ICD if the patient had not previously experienced a sustained ventricular arrhythmia or aborted SCA. The indication for the ICD was derived from retrospective review of the patient’s chart and categorized by type and class of recommendation based on the best available published guidelines (class I, IIa, IIb, or unclassified). Time-to-first occurrence of each of the following was recorded: appropriate shock, inappropriate shock, atrial lead failure, ICD lead failure, reintervention (including lead extraction or revision, placement of a new array or addition of a new lead, and pocket revision or washout), implant complication (including infection, bleeding, thrombosis, and pneumothorax), death, transplant, additional interventions for ventricular arrhythmias (including ablation or sympathectomy), generator change, and time-to-last follow-up. Long-term complications were defined as those occurring >30 days after implantation. Appropriate shock included therapy for rapidly conducted atrial tachycardias in addition to ventricular arrhythmias. First inappropriate shock, atrial or ICD lead failure, reintervention, or other long-term complications were classified as adverse events. Generator changes and implant complications within 30 days of ICD placement were tabulated separately.

**Statistical Methods**

Outcomes evaluated were time from ICD implantation to first appropriate shock, and time from ICD implantation to first adverse event. Cumulative probabilities of each outcome over time because implantation were displayed graphically. Cox proportional hazards models were used to explore associations between patient risk factors and time-to-first appropriate shock and time-to-first adverse event, and the proportional hazard assumption was validated. Risk factors examined in univariate analyses included age at implant, diagnosis, and method of ICD implantation.

To gain insight into how occurrence of appropriate therapy and adverse events changed over time, analyses were repeated conditional on survival to a specified time point. First, time 0 for the survival analyses was chosen to be the date 1 year after ICD implantation. Patients with appropriate therapy occurring in the first year after implantation and those with <1 year of follow-up were excluded. Next, time 0 was set at the date 3 years after ICD implantation; again, subjects who had already experienced appropriate therapy and those with <3 years of follow-up were excluded. Cox regression was used to estimate hazard ratios (HRs) under these conditions.

**Results**

**Demographics**

A total of 140 patients (84 male) were included with a median age at implant of 15 years (range, 8 months to 21 years; Table 1). Mean follow-up interval was 4 years (1 day to 11 years); 69 patients had ≥4 years of follow-up and 53 patients had >5 years of follow-up after device placement. Patient classifications included 35 (25%) patients with primary electric disease, 57 (41%) with cardiomyopathy (hypertrophic, dilated, or other), and 31 (22%) with congenital heart disease. There were 17 (12%) patients who did not fall into any of these categories: 9 of these had structurally normal hearts with nonsustained or inducible VT and the remaining 8 had underlying diagnoses including left ventricular aneurysm, rhabdomyoma, and previous orthotopic heart transplantation. The majority (114; 84%) were traditional transvenous implants (Table 1). Device characteristics included 94 (67%) dual-chamber devices and 7 (5%) cardiac resynchronization ICDs (cardiac resynchronization therapy defibrillator). Seventy four (53%) of the ICD leads were dual coil.

Management strategies on ICD programming were used to minimize inappropriate therapy throughout the cohort of patients. The majority of devices were manufactured by Guidant/Boston Scientific (108; 77%). Twenty-eight patients (20%) had pacing indications; 27 patients (19%) had Fidelis leads.

**Table 1. Patient Characteristics**

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median age, y (range)</td>
<td>15 (6-21)</td>
</tr>
<tr>
<td>Median height, cm (range)</td>
<td>163 (79-190)</td>
</tr>
<tr>
<td>Median weight at implant, kg (range)</td>
<td>60 (7-136)</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>84 (78%)</td>
</tr>
<tr>
<td>Diagnostic group</td>
<td></td>
</tr>
<tr>
<td>Primary electric disease</td>
<td>35 (25%)</td>
</tr>
<tr>
<td>Long QT</td>
<td>27 (19)</td>
</tr>
<tr>
<td>Brugada</td>
<td>1 (1)</td>
</tr>
<tr>
<td>CPVT</td>
<td>7 (5)</td>
</tr>
<tr>
<td>Cardiomyopathy</td>
<td>57 (41%)</td>
</tr>
<tr>
<td>HCM</td>
<td>39 (28%)</td>
</tr>
<tr>
<td>DCM</td>
<td>9 (6)</td>
</tr>
<tr>
<td>Other myopathy</td>
<td>9 (6)</td>
</tr>
<tr>
<td>Congenital heart disease</td>
<td>31 (22%)</td>
</tr>
<tr>
<td>Other</td>
<td>8 (6)</td>
</tr>
<tr>
<td>No underlying diagnosis</td>
<td>9 (6)</td>
</tr>
<tr>
<td>Placement method</td>
<td></td>
</tr>
<tr>
<td>Transvenous</td>
<td>114 (81%)</td>
</tr>
<tr>
<td>Epicardial/hybrid</td>
<td>26 (19)</td>
</tr>
<tr>
<td>Mean follow-up duration, y (range)</td>
<td>4 (1 to 11 y)</td>
</tr>
</tbody>
</table>

CPVT indicates catecholaminergic polymorphic ventricular tachycardia; DCM, dilated cardiomyopathy; and HCM, hypertrophic cardiomyopathy.
In total, 8 patients met class I indications (1 with long-QT syndromes, 4 with catecholaminergic polymorphic ventricular tachycardia, and 3 with arhythmogenic right ventricular cardiomyopathy), 66 met class IIa indications, 45 met class IIb indications, and 21 patients were unable to be classified because of lack of available published guidelines relevant to their underlying disease. More detailed indications are presented in the Data Supplement.

Outcomes

Outcomes are presented in Table 2. Twenty-seven patients (19%) experienced ≥1 appropriate shock during the follow-up period. Of these, 5 (19%) were for rapidly conducted atrial tachycardias (supraventricular tachycardia or atrial fibrillation) and the remainder were for ventricular fibrillation or VT that met device criteria for therapy. There were 79 adverse events (excluding death and transplant) observed in 51 (36%) patients. There were 59 patients (42%) at the end of the follow-up period that were free from any events, of which 19 patients had hypertrophic cardiomyopathy, 16 patients had long QT, and 12 had congenital heart disease (median follow-up, 2 years; range, 1 day to 10 years). Fifteen patients had both appropriate therapy and an adverse event.

Two (1%) patients died and 7 (5%) underwent heart transplant. One patient of 27 who received appropriate therapy was transplanted, and 8 of 113 patients who did not experience appropriate therapy died (n=2) or were transplanted (n=6); this difference was not significant (Fisher exact test, P=0.687). Two patients who were transplanted underwent revision of their ICD systems before transplantation.

Two patients died during follow-up; neither experienced appropriate therapy nor an adverse event from their device. Twenty patients (14%) experienced ≥1 inappropriate shock and 16 (11%) experienced ICD lead failure; only 2 patients experienced inappropriate shock attributed to the ICD lead failure. Thirty-six (26%) underwent an unanticipated reintervention during the follow-up period. Eleven (7.8%) patients experienced other complications (infection, bleeding, thrombosis, and pneumothorax). ICD indicates implantable cardioverter defibrillator.

Analyzing the outcomes on the basis of class of indication and disease subtype, the small number of devices placed for class I indications had a high prevalence of appropriate therapy and adverse events, when compared with devices placed for IIa/IIb or other indications, but these differences were not statistically significant (P=0.63 and 0.99 by χ², respectively; Table 3). Family history of SCA, nonsustained ventricular tachycardia, and inducible ventricular fibrillation/VT at electrophysiology study all had rates of appropriate therapy similar to the overall cohort of 25%.

Predictors of Appropriate Therapy and Adverse Events

Results of HRs estimated from the Cox models are presented in Table 4. Surgical/hybrid placement was significantly associated with time-to-first appropriate shock (HR, 2.42; P=0.03). Shorter time-to-first adverse event was associated with younger age at implant, with a HR of 2.86 for patients aged <12 years when compared with adolescents aged 17 to 21 years (P=0.01). The presence of a Fidelis lead also predicted a greater hazard of adverse events (HR, 2.06; P=0.03).

Conditional Survival Analysis

A conditional survival analysis was performed to determine whether patients who have already survived for 1 or 3 years after implant without an appropriate shock have changes in their likelihoods of future events. There were no significant predictors of time to appropriate therapy after 1 and 3 years of follow-up without previous appropriate therapy. Predictors of increased likelihood of adverse events after 1 year included the use of a Medtronic Fidelis lead (HR, 2.88; P=0.02). Examining the frequency of events by year (Figure 4), there is a higher frequency of appropriate therapy in the first years immediately after placement of a device. The incidence of appropriate therapy in patients who have not been censored for lack of follow-up and who have not had previous appropriate therapy also

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Total (Percentage of Patients Affected) N=140</th>
</tr>
</thead>
<tbody>
<tr>
<td>Appropriate shock</td>
<td>27 (19)</td>
</tr>
<tr>
<td>Death</td>
<td>2 (1)</td>
</tr>
<tr>
<td>Transplant</td>
<td>7 (5)</td>
</tr>
<tr>
<td>Adverse event</td>
<td>79 (56); adverse events occurred in 51 patients (36)</td>
</tr>
</tbody>
</table>

Percentage of patients affected reflects the average over the cohort except where noted (adverse events include inappropriate shock, ICD or atrial lead failure, reintervention, long-term complications). Complications includes infection, bleeding, thrombosis, and pneumothorax. ICD indicates implantable cardioverter defibrillator.
seems to decrease as time from device placement increases. The rate of first adverse events also decreases as the time from device placement increases, and there is a greater percentage of patients who experience first adverse events by year 5 after implantation than who experience first appropriate therapy.

**Discussion**

Appropriate ICD shocks were experienced by 19% of the patients in this study group, consistent with previous studies. However, as shown in Figure 3, if a patient did not experience appropriate therapy within the first year after ICD placement, the probability of subsequent appropriate therapy seems to decrease thereafter. The probability of an adverse event was also higher immediately after device placement and then decreased across follow-up period (although the incidence of adverse events was consistently greater than that of appropriate therapy). For patients who do not use their ICDs in the first years after implant, therefore, the risk:benefit ratio of subsequent ICD therapy increased.

![Figure 2](image)

**Figure 2.** Cumulative prevalence of first recorded outcomes after implantable cardioverter-defibrillators placement. Rate of cumulative events per patient at years 1 to 5 of follow-up. By 1 year after implant, 13 patients (9%) had experienced ≥1 appropriate shock and 23 patients (16%) had experienced an adverse event. By year 5, 27 (19%) had received appropriate therapy and 42 (29%) had experienced an adverse event.

![Figure 1](image)

**Figure 1.** Timing of first events by patient during follow-up period. Lifeline presentation of first events by patient in the 3 largest patient groups represented in this study, totaling 123 of total 143 patients (86% of all subjects); those with cardiomyopathy, congenital heart disease (CHD), and primary electric disease. In this graphic, individual patients are ordered from left to right by earliest first event after implant; the height of the blue lines represents total follow-up for that patient in days. Specific first events (appropriate shock [green diamond], inappropriate shock [yellow triangle], lead failure [orange triangle], complex reintervention [red square], death/transplant [black X], and simple generator change [gray square]) are represented at the time of their occurrence. In 2 cases in cardiomyopathy, events are recorded beyond the date of last follow-up (1 death/transplant and 1 appropriate shock); these represent events reported from other institutions without a local encounter. Although the 3 groups show subtle differences in the temporal pattern of first events, all 3 demonstrate a clustering of both appropriate shocks and adverse events in the first year after implantation.
implantation and throughout the life of the device. The data here confirm that those younger or more complicated patients with hybrid approaches are at higher risk of adverse events because of technical issues and physiological risks, both of which are increased by smaller somatic size and increased rates of growth.\textsuperscript{4,6} Although these adverse events are typically individually of lesser consequence than SCA, their cumulative burdens often come to dominate the clinical experiences of individual patients.

Neither risk of adverse event nor potential for life-saving benefit should be considered in isolation in determining indications for ICD implant or removal. However, the decision to implant has generally been considered to be a lifetime commitment because the underlying diseases that drive ICD therapy (and therefore, the imputed likely benefits) are thought unlikely to change. This is a particularly burdensome philosophy in pediatric patients, whose long anticipated lifespans predict the need for many future interventions to maintain ICD function. Recent studies in the adult literature have identified high mortality rates after elective generator replacement and have raised questions about eligibility for ICD replacement.\textsuperscript{19,20} The findings presented here raise the question of whether ICD therapy in pediatrics could similarly be considered a medium-term measure intended to protect against SCA during a sustained period of clinical observation and

![Figure 3](image-url) Percentage of patients not lost to follow-up experiencing first appropriate therapy and first adverse event in the first 5 years after device placement. The frequency of first appropriate therapy and first adverse event both decrease after implantable cardioverter defibrillators implantation. The incidence of adverse events remains greater than the incidence of appropriate therapy throughout follow-up.

![Figure 4](image-url) Time-stratified cumulative probability of first appropriate shock stratified by time from implantable cardioverter defibrillators (ICD) implantation. The rate of appropriate therapy in all patients from the time of placement of the ICD seems to decline gradually as the time increases from device placement. Patients were only included at 1 and 3 years after device placement if that had not yet had an appropriate shock.

**Table 3. Outcomes After Device Placement Stratified by Indication and Class of Indication**

<table>
<thead>
<tr>
<th>Indication</th>
<th>Total</th>
<th>Appropriate Therapy (%)</th>
<th>Adverse Event (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Syncope</td>
<td>32</td>
<td>4 (12.5)</td>
<td>13 (41)</td>
</tr>
<tr>
<td>NSVT</td>
<td>24</td>
<td>6 (25)</td>
<td>4 (17)</td>
</tr>
<tr>
<td>Family history of SCA</td>
<td>16</td>
<td>4 (25)</td>
<td>4 (44)</td>
</tr>
<tr>
<td>Inducible VT/VF at EPS</td>
<td>28</td>
<td>6 (21)</td>
<td>14 (50)</td>
</tr>
<tr>
<td>LV wall thickness &gt;30 mm</td>
<td>16</td>
<td>1 (6)</td>
<td>5 (31)</td>
</tr>
<tr>
<td>Other</td>
<td>24</td>
<td>6 (25)</td>
<td>8 (33)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Class of indication</th>
<th>Total</th>
<th>Appropriate Therapy (%)</th>
<th>Adverse Event (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>8</td>
<td>5 (63)</td>
<td>5 (63)</td>
</tr>
<tr>
<td>Ila</td>
<td>66</td>
<td>12 (18)</td>
<td>27 (41)</td>
</tr>
<tr>
<td>Ilb</td>
<td>45</td>
<td>7 (16)</td>
<td>15 (33)</td>
</tr>
<tr>
<td>Unclassified</td>
<td>21</td>
<td>3 (14)</td>
<td>4 (19)</td>
</tr>
</tbody>
</table>

Adverse events: inappropriate therapy, atrial or implantable cardioverter defibrillator lead failure, long-term device complications, reintervention. Other includes abnormal blood pressure response to exercise, abnormal findings on cardiac MR, left ventricular ejection fraction <35% with signs of heart failure, episodes of sustained ventricular tachycardia, and other indications that occurred at low frequency or were unclassifiable. Unclassified refers to devices placed in circumstances without clear published guidelines for therapy. EPS indicates electrophysiology study; LV, left ventricle; NSVT, nonsustained ventricular tachycardia; SCA, sudden cardiac arrest; and VT/VF, ventricular tachycardia/ventricular fibrillation.
accumulation of patient-specific data. At the time of the first (or subsequent) generator change—or at the time of reinter-
vention to address an adverse event—the original indications for device therapy may be carefully reconsidered in the
context of years of information.

In addition to refining indications for ICD placement and
improving ICD technology to mitigate its risk to pediatric
patients, the data presented here support the notion that addi-
tional research is warranted to determine under what condi-
tions ICD therapy could be withdrawn. Additional analyses
such as this one performed on patient populations followed up
at multiple centers will be necessary to determine how many
years of monitoring without need for appropriate therapy is
sufficient to consider the discontinuation of an ICD.

Limitations
This retrospective, single-center study, while representing a
large study population for pediatrics, is small in comparison
with large-scale adult ICD studies. Because of the heterogene-
ous patient population and multiple diagnostic categories, this study
lacks the statistical power to identify differences of outcome
in subgroup analysis. Refining those indications further would
require more robust data and continued analysis of the marginal
benefits of ICD therapy. These data account for only the first of
any type of event (be that an appropriate shock or any adverse
event), and so these results underestimate both repeated episodes
of appropriate therapy from ICDs and the repeated complica-
tions of therapy. In addition, it is evident that all adverse events
defined here do not constitute similarly poor outcomes for patients:
a hemothorax during a lead extraction or a psychologically
disabling ICD storm from a lead failure are different from
an uncomplicated extraction for a prospectively identified lead
deterioration. This analysis is unable to balance these risks of
therapy with the potential benefits fully. Finally, this experience
is limited to those patients undergoing their first ICD implant
and practically speaking, limiting statistical analysis to 
4 years of follow-up. Thus, these findings may not apply to patients who
have had multiple ICDs during longer periods of time.

Conclusions
Determining the need for primary prevention ICD therapy
in pediatric patients involves careful balancing of risks and
benefits. These data suggest that the annual risk of adverse
events decreases over the initial lifespan of an ICD system but
remains greater than the anticipated further benefits of ICD
therapy in patients who have not had appropriate therapy during
the first several years after placement.

Sources of Funding
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Boston Children’s Heart Foundation.

Disclosures
None.

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CLINICAL PERSPECTIVE

Implantable cardioverter defibrillators are being used with increasing frequency in pediatrics. The majority of these devices are currently being implanted for primary prevention of sudden cardiac death. In this study of 140 patients aged <21 years, we review our recent experience with primary prevention implantable cardioverter defibrillators with a particular focus on the time dependence of both appropriate therapy and adverse events. We found that throughout the follow-up period, there was a higher rate of adverse events (inappropriate therapy, lead failure, and need for unanticipated reintervention) than there was appropriate therapy, and that the incidence of both new appropriate therapy and adverse events decreased in the first few years after placement. We conclude that the use of primary prevention implantable cardioverter defibrillators in pediatrics deserves careful review of the potential for risk and benefit, both at the time of initial implantation and at subsequent device interventions.
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### SUPPLEMENTAL MATERIAL.

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<th>Indication</th>
<th>Number (Percentage)</th>
</tr>
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<tbody>
<tr>
<td><strong>Hypertrophic Cardiomyopathy</strong></td>
<td>39 (28% of total patients)</td>
</tr>
<tr>
<td>Septal Thickness &gt;30 mm</td>
<td>16 (41%)</td>
</tr>
<tr>
<td>Family History of SCA</td>
<td>7 (18%)</td>
</tr>
<tr>
<td>Syncope</td>
<td>5 (13%)</td>
</tr>
<tr>
<td>Abnormal BP response to exercise</td>
<td>2 (5%)</td>
</tr>
<tr>
<td>NSVT</td>
<td>2 (5%)</td>
</tr>
<tr>
<td>Abnormal CMR findings</td>
<td>1 (3%)</td>
</tr>
<tr>
<td>Other</td>
<td>6 (15%)</td>
</tr>
<tr>
<td><strong>Dilated Cardiomyopathy</strong></td>
<td>9 (6% of total patients)</td>
</tr>
<tr>
<td>LVEF &lt;35% or NYHA Class II or III</td>
<td>2 (22%)</td>
</tr>
<tr>
<td>Inducible VT</td>
<td>1 (11%)</td>
</tr>
<tr>
<td>Syncope</td>
<td>1 (11%)</td>
</tr>
<tr>
<td>NSVT</td>
<td>4 (44%)</td>
</tr>
<tr>
<td>Family History of SCA</td>
<td>1 (11%)</td>
</tr>
<tr>
<td><strong>Arrhythmogenic Cardiomyopathy</strong></td>
<td>5 (4% of total patients)</td>
</tr>
<tr>
<td>Syncope</td>
<td>1 (20%)</td>
</tr>
<tr>
<td>NSVT</td>
<td>2 (40%)</td>
</tr>
<tr>
<td>Inducible VT at EPS</td>
<td>1 (20%)</td>
</tr>
<tr>
<td>Other</td>
<td>1 (20%)</td>
</tr>
<tr>
<td><strong>Congenital Heart Disease</strong></td>
<td></td>
</tr>
<tr>
<td>Syncope</td>
<td>4 (13%)</td>
</tr>
<tr>
<td>Inducible VT/VF at EPS</td>
<td>14 (45%)</td>
</tr>
<tr>
<td>LVEF &lt;35% or NYHA Class II/ III</td>
<td>1 (3.2%)</td>
</tr>
<tr>
<td>NSVT</td>
<td>12 (39%)</td>
</tr>
<tr>
<td><strong>Long QT</strong></td>
<td>27 (19% of total patients)</td>
</tr>
<tr>
<td>Syncope despite beta blockade</td>
<td>14 (52%)</td>
</tr>
<tr>
<td>Family History of SCA</td>
<td>5 (19%)</td>
</tr>
<tr>
<td>Recurrent Torsades</td>
<td>1 (4%)</td>
</tr>
<tr>
<td>Other</td>
<td>7 (26%)</td>
</tr>
<tr>
<td><strong>CPVT / Brugada</strong></td>
<td>8 (6%)</td>
</tr>
<tr>
<td>Syncope</td>
<td>3 (37.5%)</td>
</tr>
<tr>
<td>Family History of SCA</td>
<td>3 (37.5%)</td>
</tr>
<tr>
<td>NSVT</td>
<td>1 (12.5%)</td>
</tr>
<tr>
<td>Inducible VT/VF at EPS</td>
<td>1 (12.5%)</td>
</tr>
</tbody>
</table>