Sudden Death and Defibrillators in Transposition of the Great Arteries With Intra-atrial Baffles
A Multicenter Study

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Background—Transposition of the great arteries with intra-atrial baffle repair is among the congenital heart defects at highest risk of sudden death. Little is known about mechanisms of sudden death and the role of implantable cardioverter defibrillators.

Methods and Results—We conducted a multicenter cohort study in patients with transposition of the great arteries to determine actuarial rates of implantable cardioverter defibrillator shocks, identify risk factors, assess underlying arrhythmias, and characterize complications. Overall, 37 patients (age, 28.0±7.6 years; 89.2% male) were enrolled from 7 sites. Implantable cardioverter defibrillators were implanted for primary prevention in 23 (62.1%) patients and secondary prevention in 14 patients (37.8%). Annual rates of appropriate shocks were 0.5% and 6.0% in primary and secondary prevention, respectively (P=0.0366). Independent predictors were a secondary prevention indication (hazard ratio, 18.0; P=0.0341) and lack of β-blockers (hazard ratio, 16.7; P=0.0301). In patients with appropriate shocks, intracardiac electrograms documented supraventricular tachycardia preceding or coexisting with ventricular tachycardia in 50%. No patient with inducible ventricular tachycardia received an appropriate shock in comparison with 37.5% of noninducible patients (P=0.0429). Inappropriate shocks occurred in 6.6% per year, more so in patients of lesser weight (hazard ratio, 0.91 per kg; P=0.0168). Additionally, 14 patients (37.8%) experienced complications: 5 (13.5%) acute, 1 (2.7%) late generator related, and 12 (32.4%) late lead related.

Conclusion—In patients with transposition of the great arteries, high rates of appropriate shocks are noted in secondary but not primary prevention. Supraventricular arrhythmias may be implicated in the etiology of ventricular tachyarrhythmias; β-blockers seem protective, and inducible ventricular tachycardia does not seem to predict future events. Inappropriate shocks and late lead-related complications are common. (Circ Arrhythmia Electrophysiol. 2008;1:250-257.)

Key Words: death, sudden • heart arrest • defibrillation • heart defects, congenital • tachyarrhythmias • transposition of great vessels

Complete transposition of the great arteries (D-TGA) with intra-atrial redirection surgery (Mustard or Senning baffles) is among the congenital heart defects at highest risk for sudden cardiac death. Indeed, sudden death is the most common cause of late mortality in this patient population.1–7 A population-based study reported an incidence of 4.9 per 1000 patient-years, second only to aortic stenosis and more than threefold greater than tetralogy of Fallot.1 Identification of risk factors and pathophysiological mechanisms for sudden death have been somewhat elusive. Although bradyarrhythmias were once thought to be primary triggers, this notion was later refuted, as pacemakers were not found to afford protection against sudden death. Identified risk factors are largely confined to arrhythmia symptoms and documented atrial tachyarrhythmias.4,8

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Experience with implantable cardioverter defibrillators (ICDs) in patients with D-TGA is limited, whether prompted by a sustained ventricular tachyarrhythmia or resuscitated cardiac arrest (secondary prevention indication), or a clinical profile deemed high risk in the absence of a near fatal event (primary prevention indication). We conducted a multicenter retrospective cohort study with the objectives of determining...
Data regarding inappropriate ICD shocks were likewise collected. All ICD events (ie, antichadrycardia pacing or shock) and tracings were requested, up to a maximum of 5 per “appropriate” and “inappropriate” category for each patient.

A blinded adjudicating committee reviewed and classified all ICD events, with each tracing analyzed by ≥2 independent electrophysiologists. Appropriate therapy was subclassified as monomorphic ventricular tachycardia (ie, electrogram with a uniform and constant morphology), polymorphic ventricular tachycardia (ie, electrogram with relatively constant amplitude but displaying a shift in morphology or axis), or ventricular fibrillation (ie, constant shift in axis and morphology of the electrogram accompanied by marked and variable changes in amplitude). Inappropriate ICD shocks were further categorized according to type of most likely underlying rhythm (ie, noise interference or oversensing, sinus tachycardia, atrial flutter, atrial fibrillation, and other form of supraventricular tachycardia).

ICD Complications

ICD complications were considered “periprocedural” if they occurred within 30 days of implantation and “late” thereafter. Late complications were subdivided into lead and generator-related events. Lead-related complications included lead dislodgement, lead failure, endocarditis, and undersensing or oversensing. Pain, erosion, pocket infection, migration, and device malfunction were considered generator-related complications.

Statistical Analysis

Time zero was defined as time of ICD implantation. Patient-years were accrued from time of entry until occurrence of an ICD shock or the study termination date. Censoring occurred in the event of cardiac transplantation, loss to follow-up, or death from other causes not involving ICD therapy. Continuous variables are summarized by mean±SD or median and interquartile range (25th, 75th percentile), depending on normality of distribution. Categorical variables are represented by frequencies and percentages. Baseline comparisons between patients with ICDs for primary versus secondary prevention were performed by Mann-Whitney rank sum, student t, or χ² tests where appropriate. Freedom from appropriate and inappropriate ICD shocks and overall survival was plotted using the Kaplan-Meier method, with comparisons by log-rank statistics.

To assess predictors of ICD shocks, univariate and stepwise multivariate Cox proportional hazard models were used after verifying proportional-hazards assumptions. Variables with probability values <0.1 in univariate analyses were considered in multivariate models. Two-tailed probability values <0.05 were considered statistically significant. Analyses were performed with SAS version 9.1 (SAS Institute, Cary, NC).

The authors had full access to and take full responsibility for the integrity of the data. All authors have read and agreed to the manuscript as written.

Results

Baseline Characteristic

A total of 37 patients (mean age, 28.0±7.6 years; 89.2% male), were enrolled from 7 sites. ICDs were implanted for primary prevention in 23 patients (62.1%) and secondary prevention in 14 patients (37.8%). Age at time of ICD implantation is graphically depicted in Figure 1. Baseline characteristics in all patients and by primary versus secondary prevention indications are summarized in Table 1. Patients with ICDs for primary versus secondary prevention were older at implantation, had a higher prevalence of documented nonsustained ventricular tachycardia, and exhibited a trend toward more moderate or severe right ventricular systolic dysfunction. ECGs were available in all patients, echocardiograms in 36 patients (97.3%), chest x-rays in 34 patients (91.9%), Holters in 29 patients (78.4%), cardiac catheteriza-

ICD Implantation

Indications prompting ICD implantation were recorded and included presyncope/dizziness, syncope, palpitations, clinical nonsustained ventricular tachycardia, QRS duration ≥180 ms, severe systemic ventricular systolic dysfunction, inducible ventricular tachycardia, clinical sustained ventricular tachycardia, ventricular fibrillation/resuscitated cardiac arrest, and other. Patients were stratified according to whether the ICD was indicated for primary or secondary prevention. “Secondary prevention” was defined by clinical sustained ventricular tachycardia, ventricular fibrillation, or resuscitated cardiac arrest. As per convention, barring such events, the ICD was considered indicated for “primary prevention.”

Procedural characteristics were logged and medical therapy at the time of discharge was noted, with particular attention to β-blockers, amiodarone, sotalol, dofetilide, and class IA or IC antiarrhythmic agents.

ICD Shocks

The main outcome consisted of appropriate ICD shocks. The ventricular tachycardia cycle length, programmed defibrillator zone (ie, slow ventricular tachycardia, fast ventricular tachycardia, or ventricular fibrillation), and success or failure of therapy was noted.

Baseline Characteristics

A similar protocol was previously described in patients with tetralogy of Fallot. Data collection was conducted in accordance with individual hospital institutional review board policies. Details regarding demographic variables, surgical history including type of intra-atrial baffle repair (ie, Mustard or Senning), associated anomalies, interventions before or concomitant with surgical repair, electrocardiography, chest radiography, 24-hour Holter monitoring, 2D and M-mode Doppler echocardiography, cardiac magnetic resonance (CMR) imaging, cardiac catheterization, and electrophysiology studies were collected. The most recent data preceding ICD implantation were requested, with a maximum acceptable time interval of 2 years.

Electrocardiographic data included heart rate, presence of an underlying ventricular-paced rhythm, longest dominant QRS duration, and QT intervals. Cardiothoracic ratios were derived from posteroanterior chest radiographs. The number of premature ventricular complexes in 24 hours and presence of nonsustained ventricular tachycardia (>3 beats, <30 seconds) were captured from Holter monitors. Echocardiographic and CMR data included biventricular size and function, degree of valvar regurgitation, and estimates of systolic pulmonary arterial pressure. When both echocardiographic and CMR studies were performed, CMR data were retained. Data extracted from programmed ventricular stimulation studies included inducibility of sustained monomorphic or polymorphic ventricular tachycardia.

ICD Complications

ICD complications were classified by type and location. Lead-related complications included lead dislodgement, lead failure, endocarditis, and undersensing or oversensing. Pain, erosion, pocket infection, migration, and device malfunction were considered generator-related complications.

Statistical Analysis

Time zero was defined as time of ICD implantation. Patient-years were accrued from time of entry until occurrence of an ICD shock or the study termination date. Censoring occurred in the event of cardiac transplantation, loss to follow-up, or death from other causes not involving ICD therapy. Continuous variables are summarized by mean±SD or median and interquartile range (25th, 75th percentile), depending on normality of distribution. Categorical variables are represented by frequencies and percentages. Baseline comparisons between patients with ICDs for primary versus secondary prevention were performed by Mann-Whitney rank sum, student t, or χ² tests where appropriate. Freedom from appropriate and inappropriate ICD shocks and overall survival was plotted using the Kaplan-Meier method, with comparisons by log-rank statistics.

To assess predictors of ICD shocks, univariate and stepwise multivariate Cox proportional hazard models were used after verifying proportional-hazards assumptions. Variables with probability values <0.1 in univariate analyses were considered in multivariate models. Two-tailed probability values <0.05 were considered statistically significant. Analyses were performed with SAS version 9.1 (SAS Institute, Cary, NC).

The authors had full access to and take full responsibility for the integrity of the data. All authors have read and agreed to the manuscript as written.
tion in 25 patients (67.6%), electrophysiological studies in 17 patients (45.9%), and CMR in 11 patients (29.7%).

In patients with primary prevention indications, defibrillator implantation was prompted by the following nonexclusive reasons: presyncope in 3 patients (13.0%), syncope in 8 patients (34.8%), palpitations in 12 patients (52.2%), nonsustained ventricular tachycardia in 11 patients (47.8%), systemic right ventricular ejection fraction \( \leq 35\% \) in 8 patients (34.8%), QRS duration \( \geq 180 \) ms in 7 patients (30.4%), and inducible sustained ventricular tachycardia in 7 patients (30.4%). Indications for secondary prevention were ventricular fibrillation and/or resuscitated cardiac arrest in 10 patients (71.4%) and clinical sustained ventricular tachycardia without cardiac arrest in 4 patients (28.6%).

**Appropriate Antitachycardia Pacing or Shocks**

Overall, 139 ICD discharges occurred in 12 patients over a median follow-up of 3.6 (1.5, 5.5) years: 3.5 (1.4, 5.3) years and 3.9 (2.3, 6.6) years in patients with primary and secondary prevention indications, respectively. Of all first ICD discharges, 33.3% were appropriate. Five patients (13.5%) received a total of 47 appropriate shocks (1 in primary prevention; 46 in secondary prevention) clustered as follows: 1, 2, 4, 5, and 35 shocks. The median detected ventricular tachyarrhythmia rate at time of first appropriate shock was 222 (165, 261) bpm. Monomorphic ventricular tachycardia constituted 48.9% of appropriately treated arrhythmias, polymorphic ventricular tachycardia constituted 34.0%, and ventricular fibrillation constituted 17.0%. First ICD discharges consisted of antitachycardia pacing alone in 1, antitachycardia pacing followed by a shock in 1, and shocks alone in 3 patients. The patient with initial antitachycardia pacing alone later received an appropriate ICD shock.

**Appropriate ICD Shocks**

High rates of appropriate ICD shocks were noted for secondary (28.6%) but not primary (4.3%) prevention indications, with annualized actuarial event rates of 6.0% and 0.5%, respectively \( (P=0.0366) \). The only primary prevention ICD patient to receive an appropriate shock was a 33-year-old man with a Mustard baffle, severe systemic right ventricular dysfunction (ejection fraction 12%), moderate tricuspid regurgitation, and nonsustained ventricular tachycardia. Freedom from appropriate ICD shocks in primary and secondary prevention is plotted in Figure 2A. All first appropriate ICD shocks were successful. Univariate and multivariate predictors of appropriate ICD shocks are listed in Table 2. Figure 3 depicts freedom from appropriate ICD shocks according to whether or not patients received β-blockers. Class III antiarrhythmic agents did not significantly modulate risk of appropriate shocks.

Of the 18 adjudicated appropriate shocks, supraventricular tachyarrhythmias were recognized as preceding or coexisting with ventricular tachyarrhythmias in 9 patients (50.0%), ventricular tachyarrhythmias appeared primary in 4 patients (22.2%), and in the remaining 5 patients (27.8%), data provided from single-chamber ICDs precluded inferences regarding prior or concomitant supraventricular tachyarrhythmias.

**Inducible Ventricular Tachycardia and Appropriate ICD Shocks**

Of 17 patients with electrophysiological studies, 10 (58.8%) had primary and 7 (41.2%) had secondary prevention indications. Overall, 9 patients (52.9%; 6 primary prevention) had inducible sustained ventricular tachycardia, subclassified as monomorphic in 3 patients (2 primary prevention) and polymorphic in 6 patients (4 primary prevention). Appropriate ICD shocks were received by 0 of 9 patients with inducible sustained ventricular tachycardia and 3 of 8 (ie, 0 of 4 and 3 of 4 with primary and secondary prevention indications, respectively) noninducible patients (37.5%; \( P=0.0429 \)). Two of the 3 noninducible patients with appropriate ICD shocks received them for monomorphic ventricular tachycardia, whereas the remaining patient had recurrent polymorphic ventricular tachycardia.

**Inappropriate ICD Shocks**

Overall, 9 patients (24.3%) received 92 inappropriate ICD shocks, 67 of which were in 5 patients (21.7%) with primary prevention and 25 of which were in 4 patients (28.6%) with secondary prevention indications. Underlying causes were noted for 78 inappropriate shocks: lead fracture, failure, or oversensing in 48 patients (61.5%), supraventricular tachycardia in 21 patients (26.9%), and sinus tachycardia in 9 patients (11.5%). The 21 inappropriate shocks for supraventricular arrhythmias occurred in 7 patients and were all designated “atrial flutter.” The median detected ventricular tachycardia rate was 188 (165, 200) bpm, significantly lower than appropriately shocked arrhythmias \( (P=0.0078) \).

Freedom from inappropriate ICD shocks at 1, 2, and 5 years was 83.2%, 75.6%, and 71.1%, respectively, corresponding to an average actuarial rate of 6.6% per year. No difference was observed between primary and secondary prevention groups \( (P=0.9765) \). In univariate analyses, risk factors for inappropriate shocks were female sex (hazard ratio, 4.9; 95% CI, 1.2 to 19.6; \( P=0.0261 \)) and lighter weight (hazard ratio, 0.91 per kg; 95% CI, 0.85 to 0.98; \( P=0.0106 \)).
In multivariate analyses, the latter was the only independent predictor of inappropriate ICD shocks (hazard ratio, 0.91 per kg; 95% CI, 0.85 to 0.98; \( P = 0.0168 \)).

### Complications

Complications other than inappropriate shocks occurred in 14 patients (37.8%) overall: acute in 5 (13.5%), late generator
related in 1 (2.7%), and late lead related in 12 (32.4%). Table 3 summarizes complications according to subtype; the categories are not mutually exclusive. Three patients had both acute and late complications.

Overall Survival
Three patients (8.1%) died during follow-up: 2 (5.4%) from heart failure and 1 (2.7%) suddenly. The overall actuarial survival rate was 100%, 96.9%, and 91.0% at 1, 2, and 5 years, respectively, corresponding to an averaged actuarial mortality rate of 1.9% per year. As depicted in Figure 2B, the mortality rate was higher in ICD recipients with secondary versus primary prevention indications, with annualized rates of 3.4% versus 0% \((P=0.0206)\).

The only episode of sudden death occurred in a 22-year-old man with a Senning baffle, an ICD for secondary prevention, and a prior appropriate and effective shock. His fatal event was triggered by polymorphic ventricular tachycardia that prompted 4 appropriate ICD shocks. Although the final shock successfully interrupted ventricular fibrillation, electromechanical dissociation ensued with an agonal escape rhythm.

Discussion
D-TGA accounts for 5% to 7% of all congenital cardiac malformations. Although largely supplanted by arterial switch surgery, intra-atrial Senning and Mustard baffles permitted survival into adulthood, often with good quality of life.

Table 3. Acute and Late Complications

<table>
<thead>
<tr>
<th>Variable</th>
<th>N (%)</th>
</tr>
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<tbody>
<tr>
<td>Acute complications</td>
<td></td>
</tr>
<tr>
<td>Hematoma</td>
<td>1 (2.7)</td>
</tr>
<tr>
<td>Pneumothorax</td>
<td>1 (2.7)</td>
</tr>
<tr>
<td>Reintervention</td>
<td>4 (10.5)</td>
</tr>
<tr>
<td>Lead dislodgement</td>
<td>3 (8.1)</td>
</tr>
<tr>
<td>Diaphragmatic stimulation</td>
<td>2 (5.4)</td>
</tr>
<tr>
<td>Late complications</td>
<td></td>
</tr>
<tr>
<td>Generator related</td>
<td>1 (2.7)</td>
</tr>
<tr>
<td>Pocket infection</td>
<td>1 (2.7)</td>
</tr>
<tr>
<td>Lead related</td>
<td>12 (32.4)</td>
</tr>
<tr>
<td>Dislodgement</td>
<td>3 (8.2)</td>
</tr>
<tr>
<td>Failure and/or fracture</td>
<td>6 (16.2)</td>
</tr>
<tr>
<td>Endocarditis</td>
<td>1 (2.7)</td>
</tr>
<tr>
<td>Under and/or over sensing</td>
<td>4 (10.8)</td>
</tr>
<tr>
<td>Superior vena cava thrombosis</td>
<td>1 (2.7)</td>
</tr>
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life. Sudden death is the most common cause of mortality. Indeed, D-TGA with an intra-atrial baffle ranks among the highest risk congenital heart defects, far exceeding tetralogy of Fallot. Unlike some congenital heart defects where hazard is appreciable only years after surgery, propensity for sudden cardiac death in Mustard and Senning recipients with D-TGA appears early. Risk seems relatively constant over time, with estimated sudden cardiac death-free survival rates of 96% at 10 years and 91% at 20 years.

Little is known about ICDs in this patient population, with the literature largely limited to case reports and small case series. One series of 5 patients reported no appropriate shocks at a median of 20 months. The second series included 7 patients with D-TGA as part of a larger cohort of adults with congenital heart disease. Subgroup analyses specific to D-TGA were not reported. To our knowledge, our multicenter study is the first to explicitly address the role of ICDs in high-risk patients with D-TGA. Importantly, a high rate of appropriate shocks was observed in patients with secondary prevention indications. Ventricular tachyarrhythmias were reliably sensed and effectively interrupted. In contrast, the rate of appropriate ICD shocks in patients with primary prevention indications was exceedingly low, highlighting our limitations in reliably identifying appropriate high-risk candidates. Consequently, prophylactic ICDs in D-TGA for indications such as systemic right ventricular dysfunction, nonsustained ventricular tachycardia, syncope, and inducible ventricular tachycardia remain unsubstantiated.

Little is known about the pathophysiology of sudden death in patients with D-TGA and intra-atrial baffles. To this regard, our study design provided a unique opportunity to clarify mechanistic triggers. Intracardiac ICD tracings were requested on all patients with shocks and subject to systematic blinded adjudication. Interestingly, supraventricular tachyarrhythmias were often found to precede or coexist with ventricular tachycardia. It can be conjectured, therefore, that supraventricular tachyarrhythmias may be an important contributor to sudden death in this patient population and that benefit may be derived from the suppression or elimination of these substrates. This notion is consistent with reported risk factors for sudden death. In a case-control study, Kammeraad et al found that arrhythmia symptoms and documented atrial fibrillation or flutter were most predictive of sudden death. Several cohort studies likewise reported an increased incidence of sudden death in patients with supraventricular arrhythmias, although it remained unclear whether these were causative or surrogate markers for structural or hemodynamic impairment.

Factors that may enhance susceptibility to degeneration of supraventricular arrhythmias into fatal events include impaired atrioventricular transport with failure to augment right ventricular filling rates during tachycardia, atrioventricular nodal conduction capable of rapid ventricular responses, systemic right ventricular dysfunction, and subendocardial ischemia resulting from a right coronary circulation irrigating a systemic ventricle. In addition to potentially suppressing primary ventricular arrhythmias, \( \beta \)-blockers may have salutary effects on a combination of these factors, which may explain, in part, our finding that \( \beta \)-blockers afford protection against appropriate shocks. \( \beta \)-blockers may suppress supraventricular arrhythmias, increase diastolic filling time, slow ventricular responses by increasing the atrioventricular nodal effective refractory period, and reduce myocardial ischemia. Unlike the lack of benefit with angiotensin receptor blockade, preliminary data suggests that carvedilol may be associated with positive right ventricular remodeling in patients with systemic right ventricles.

Despite a predominance of supraventricular arrhythmias, we also observed primary ventricular tachyarrhythmias. Importantly, these occurred exclusively in patients with secondary prevention indications. In a manner similar to dilated and hypertrophic cardiomyopathy, programmed ventricular stimulation was of limited prognostic value. Fewer appropriate shocks occurred in inducible patients. Although this likely reflects a chance finding, it is improbable that a larger sample size would detect a statistically significant association in the opposite direction. This is unlike tetralogy of Fallot where inducible sustained ventricular tachycardia is of value in predicting clinical ventricular tachyarrhythmia, sudden death, and appropriate shocks.

As for complications, in patients with congenital heart disease, high rates of inappropriate ICD shocks are consistently reported. This likely reflects the relatively young age of the study population, propensity for coexisting supraventricular tachyarrhythmias, and higher susceptibility for lead failure. Indeed, nearly one third of our patients experienced lead-related complications. Prior studies have likewise noted such high rates of complications in patients with congenital heart disease with pacemakers and ICDs. Although we found lighter weight to independently predict inappropriate shocks, younger age was previously identified as a risk factor for lead fracture.

Although some overlap was noted, supraventricular arrhythmias that provoked inappropriate shocks were generally slower than ventricular tachyarrhythmias. Post hoc analysis suggests that inappropriate shocks could have largely been prevented by programming the ventricular tachycardia detection rate >200 bpm. The desirability of such an approach remains speculative, as it cannot be conclusively determined whether inappropriate shocks for supraventricular tachyarrhythmias may have circumvented otherwise fatal events (i.e., appropriate “inappropriate” shocks).

Limitations

Our study was retrospective, detection rates for ICD shocks were not uniformly programmed, and medical decisions were not randomly allocated. Primary prevention indications were recorded and assessed, but standardized selection criteria were not applied. Despite being the largest study of its kind, the ability to identify relevant predictors is limited by the overall low event rate. As such, a conservative probability value cutoff (<0.1) was selected to limit potential variables considered in multivariate models. Potential for human error remains despite efforts to minimize misclassification of appropriate versus inappropriate ICD shocks through systematic appraisal by a blinded expert adjudicating committee. In 28% of cases, appropriate ICD shocks were confirmed but provided data were insufficient to comment on preceding
supraventricular tachyarrhythmias. Importantly, ICD recipients were not compared to patients with similarly high-risk features but without defibrillators, precluding quantification of survival benefits.

**Conclusion**
High rates of appropriate and effective ICD shocks were observed in patients with D-TGA, Mustard or Senning baffles, and clinical sustained ventricular tachyarrhythmias or resuscitated cardiac arrest. This suggests an important role for ICDs in secondary prevention against sudden death. In contrast, protective effects from primary prevention ICDs could not be surmised from our findings. Although primary ventricular arrhythmias are observed in this population of patients, supraventricular tachyarrhythmias seem to be a common trigger. β-blockers afford protection against appropriate shocks and programmed ventricular stimulation does not seem to be of prognostic value. Frequent adverse events include inappropriate shocks and lead-related complications.

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**Disclosures**
None.

**References**


CLINICAL PERSPECTIVE

Transposition of the great arteries with Mustard or Senning baffles is among the congenital heart defects at highest risk for sudden cardiac death. We conducted a multicenter cohort study to determine rates of appropriate and inappropriate ICD shocks, identify risk factors, characterize complications, and elucidate potential mechanistic triggers for sudden deaths. Overall, 37 patients (age, 28.0 ± 7.6 years) were enrolled from 7 sites with ICDs for primary prevention in 23 and secondary prevention in 14. A high rate of appropriate shocks was noted in secondary (6.0% per year) but not primary (0.5% per year) prevention. Independent risk factors were limited to a secondary prevention indication (hazard ratio, 18.0) and lack of β-blockers (hazard ratio, 16.7). Inducible ventricular tachycardia did not seem to predict appropriate shocks. In 50% of cases, intracardiac electrogram tracings recorded supraventricular tachycardia preceding or coexisting with ventricular tachycardia. Inappropriate shocks occurred in 6.6% per year, and 37.8% of patients experienced complications, predominantly late lead related. Although these results generally support the use of ICDs for secondary prevention, they likewise highlight our limitations in reliably identifying appropriate candidates for prophylactic ICDs. Indications such as systemic right ventricular dysfunction, nonsustained ventricular tachycardia, syncope, and inducible ventricular tachycardia remain unsubstantiated. Importantly, it is unknown whether inappropriate shocks for supraventricular tachyarrhythmias may have prevented fatal events (ie, appropriate “inappropriate” shocks). As supraventricular tachyarrhythmias seem to be important contributors to sudden death in this patient population, it may be hypothesized that benefit may be derived from suppressing or eliminating these substrates.
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