Primary prevention with the implantable cardioverter-defibrillator in high-risk long-QT syndrome patients

Yitschak Biton^{1,2*}, Spencer Rosero³, Arthur J. Moss¹, Ilan Goldenberg², Valentina Kutyifa¹, Scott McNitt¹, Bronislava Polonsky¹, Jayson R. Baman^{1,4}, and Wojciech Zareba¹

¹Department of Medicine, Division of Cardiology, Heart Research Follow-up Program, University of Rochester Medical Center, 265 Crittenden Blvd., PO Box 653, Rochester, NY 14642, USA; ²Department of Medicine, Division of Cardiology, University of Rochester Medical Center, Rochester, NY, USA; ³Heart Institute, Sheba Medical Center, Ramat Gan and Sackler School of Medicine, Tel Aviv University, Tel Aviv, Israel; and ⁴Department of Medicine, Feinberg School of Medicine, Northwestern University, Chicago, IL, USA

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Aims

Prospective data regarding the role of implantable cardioverter-defibrillator (ICD) for the primary prevention of sudden cardiac death in patients with long QT syndrome (LQTS) is scarce. Herein, we explore the prospective Rochester LQTS ICD registry to assess the risk for appropriate shock in primary prevention in a real-world setting.

Methods and results

We studied 212 LQTS patients that had ICD implantation for primary prevention. Best-subsets proportional-hazards regression analysis was used to identify clinical variables that were associated with the first appropriate shock. Conditional models of Prentice, Williams, and Peterson were utilized for the analysis of recurrent appropriate shocks. During a median follow-up of 9.2 ± 4.9 years, there were 42 patients who experienced at least one appropriate shock and the cumulative probability of appropriate shock at 8 years was 22%. $QTc \geq 550$ ms [hazard ratio (HR) 3.94, confidence interval (CI) 2.08–7.46; $P < 0.001$] and prior syncope on β -blockers (HR 1.92, CI 1.01–3.65; $P = 0.047$) were associated with increased risk of appropriate shock. History of syncope while on β -blocker treatment (HR 1.87, CI 1.28–2.72; $P = 0.001$), QTc 500–549 ms (HR 1.68, CI 1.10–2.81; $P = 0.048$), and $QTc \geq 550$ ms (HR 3.66, CI 2.34–5.72; $P < 0.001$) were associated with increased risk for recurrent appropriate shocks, while β -blockers were not protective (HR 1.03, CI 0.63–1.68, $P = 0.917$). LQT2 (HR 2.10, CI 1.22–3.61; $P = 0.008$) and multiple mutations (HR 2.87, CI 1.49–5.53; $P = 0.002$) were associated with higher risk for recurrent shocks as compared with LQT1.

Conclusion

In this prospective ICD registry, we identified clinical and genetic variables that were associated appropriate shock risk. These data can be used for risk stratification in high-risk patients evaluated for primary prevention with ICD.

Keywords

Long QT syndrome • Implantable cardioverter-defibrillator • Primary prevention • Shock

Introduction

Implantable cardioverter-defibrillators (ICD) are a well-established treatment modality for patients with long QT syndrome (LQTS) patients that survived aborted cardiac arrest (ACA).^{1,2} The role of ICD in patients that did not experience ACA is very limited and is

mostly based on expert opinion and prior secondary prevention studies.^{3–9} Currently, ICD is recommended for primary prevention in patients that had syncope or ventricular tachycardia (VT) while receiving β -blocker (Class IIa indication), and may be considered in patients with risk factors for sudden cardiac death (SCD).¹

* Corresponding author. Tel: +1 585 275 5391; fax: +1 585 273 5283. E-mail address: yitschak.biton@heart.rochester.edu; yitscb@gmail.com

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What's new?

- Beta-blockers are the mainstay of treatment in long QT syndrome, but many patients still experience both fatal and non-life-threatening events while on beta-blocker therapy, thus raising concern that medical therapy alone may be not be sufficient for high-risk patients.
- We found that, in patients implanted with prophylactic implantable cardioverter-defibrillator (ICD), the presence of long QT syndrome type 2 and multiple long QT syndrome-related mutations were associated with higher risk for recurrent shocks.
- Of the beta blockers studied, metoprolol was associated with the highest rate of appropriate shock therapy.
- In a population of patients who received ICD placement for primary prevention in long QT syndrome, we were able to identify genetic and clinical criteria to identify high-risk patients for whom ICD provides appropriate and necessary shocks.

The prospective Rochester LQTS ICD registry provides a unique opportunity to assess the role of ICD for primary prevention in a real-world setting. In this study, we investigated clinical outcomes in LQTS patients who did not experience ACA and we aimed to: (i) evaluate the current indications for ICD implantation in these patients; (ii) evaluate whether high risk clinical factors can be used to predict the risk for first and recurrent appropriate ICD shocks; (iii) appreciate the incremental predictive value of genotype data; and (iv) explore the clinical factors that are associated with inappropriate ICD therapy.

Methods

Study population

The study population consisted 212 out of 334 patients from the prospective Rochester LQTS-ICD registry. The institutional review board approved the study protocol. Patients were included if they had ICD implantation but did not experience ACA prior to the implantation. Patients that had ICD implantation for a secondary prevention indication were not included in this study ($n=122$). Clinical information was obtained using pre-specified data forms, with follow-up information acquired annually. The baseline electrocardiogram was used for QTc and heart rate measurements. Indications for ICD therapy were categorized according to the history preceding ICD implantation. The follow-up time started at the day of the ICD implantation. Patients were censored at the day of device extraction if they were not re-implanted in the same day or at the age of 50 years in order to avoid ischaemic heart disease as a competing risk factor for ventricular arrhythmia.

Device interrogation

ICD interrogation data were collected periodically and adjudicated by two physicians (S.R., W.Z.) to determine appropriateness of ICD therapy.

Outcome measures

The primary outcome of the current study was the presence or absence of first appropriate ICD shock. Secondary outcomes were (i) recurrent

appropriate ICD shocks defined as a subsequent shocks occurring at least 48 h apart and (ii) the first inappropriate ICD shock.

Genetic testing

If a patient was a proband, he was tested for the following five genes: KCNQ1, KCNH2, SCN5a, KCNE1, and KCNE2. Patients who were family members of a proband with LQTS were first tested for the gene that their proband carried and some of these patients were additionally tested for all five genes. Not all patients in our registry (about 10%) were genetically tested since testing was not readily available at the time of ICD implant or may not have been performed due to patient's preference or high insurance copayment.

Statistical analysis

Baseline clinical characteristics were compared between patients from the four genotype groups (LQT1, LQT2, LQT3, and multiple mutations) using the χ^2 test or Fisher's exact test for categorical variables. For purposes of consistency, we used the Kruskal–Wallis test for all continuous variables. Categorical data are presented as frequencies and percentages and continuous variables as means and standard deviations.

The cumulative probability of appropriate shock and inappropriate shock was calculated according to the Kaplan–Meier method with comparisons of cumulative event rates by the log-rank test. Event rate per 100 patients years of appropriate ICD shocks were calculated by dividing the total number of all the appropriate shocks (including recurrent shocks) by patient years and multiplying the results by 100, with comparisons of the event rates by the negative binominal regression.

We pre-specified several risk factors that are known to be associated with the risk of SCD in LQTS patients in order to predict the outcome of appropriate ICD shocks including: age (categorized either at 15 years or 20 years), gender, QTc (using two cut-offs either 500 ms or 550 ms),^{7,10–13} genotype,¹⁴ mutation location,^{15–17} history of prior syncope on/off β -blockers,^{13,18–20} family history of ACA/SCD, triggers of prior syncope, and the M-FACT score [calculated as the sum of these variables: age of implantation ≤ 20 years, events on therapy and 500 < QTc ≤ 550 ms (one point for each variable and two points for QTc > 550 ms)].⁷

Multivariate Cox proportional hazards regression and the best subset regression modelling were used to assess the effect of the pre-specified risk factors on the outcomes of appropriate shock and inappropriate shock. β -Blocker therapy was assessed in the multivariate model in a time-dependent manner by incorporating data for each patient that identifies the effect of each follow-up time 'on' and 'off' β -blocker therapy during the follow-up. For the analysis of recurrent appropriate ICD shocks the conditional models of Prentice, Williams, and Peterson were utilized.^{21,22} If a subject had more than eight events they were censored at the eighth event.

All statistical tests were two-sided, a P -value of <0.05 was considered statistically significant. Analyses were carried out with SAS software (version 9.4, SAS institute, Cary, NC, USA).

Sensitivity analyses

We performed a sensitivity analysis with the follow-up starting at 11 days after the ICD implantation in order to make sure that arrhythmias that are caused by mechanical stimulation of the electrode in the first few days are not biasing the results. Also, we repeated all the analyses by either adjusting the models for Torsades de Pointes (TdP) or excluding patients who had TdP and had no syncope on β -blocker.

Risk score

The risk score built was based on variables that were significant predictors of the first and recurrent appropriate ICD shocks in the multivariate

Table 1 Patient baseline clinical characteristics and post-ICD implantation treatment and outcomes categorized by genotype

	All (n = 212)	LQT1 (n = 45)	LQT2 (n = 68)	LQT3 (n = 15)	Multiple (n = 10)	Other (n = 74)
Age at first cardiac event (years)	16 ± 11	16 ± 11	15 ± 10	18 ± 11	7 ± 8	17 ± 13
Age at ICD implantation (years)	26 ± 14	24 ± 13	28 ± 13	28 ± 14	14 ± 13 ^a	28 ± 13
Female	(70)	(71)	(68)	(67)	(80)	(72)
QTc (ms)	504 ± 61	514 ± 57	511 ± 67	515 ± 60	552 ± 73	481 ± 47
QTc ≥ 550 ms	(23)	(25)	(26)	(33)	(50)	(12)
Follow-up (years)	9.2 ± 4.9	9.0 ± 4.7	9.8 ± 5.3	8.1 ± 4.1	11.6 ± 3.2	8.6 ± 5.0
M-FACT score	1.2 ± 1.1	1.5 ± 1.3	1.2 ± 1.2	1.2 ± 1.2	2.2 ± 0.8	1 ± 0.9
Probands	(50)	(62)	(43)	(13)	(60) ^a	(54)
Congenital deafness	(3)	(2)	(0)	(0)	(20) ^a	(4)
Family history for probands						
LQTS	(48)	(50)	(48)	(100)	(83)	(44)
Syncope/aborted cardiac arrest	(46)	(39)	(45)	(100)	(50)	(49)
Sudden cardiac death	(41)	(36)	(38)	(50)	(17)	(50)
Treatment prior to ICD implantation						
β-Blockers	(79)	(82)	(81)	(87)	(90)	(73)
Sodium channel blockers	(4)	(0)	(0)	(27)	(0) ^a	(7)
Pacemaker	(14)	(9)	(15)	(0)	(0)	(22)
LCSD	(1)	(0)	(1)	(0)	(0)	(1)
Indications for ICD						
Syncope off β-blockers/sodium channel blockers	(49)	(53)	(51)	(27)	(60)	(46)
Syncope while on β-blockers	(30)	(31)	(28)	(13)	(30)	(34)
Syncope while on sodium channel blockers	(2)	(0)	(0)	(7)	(0)	(5)
Torsades de pointes	(11)	(2)	(16)	(0)	(0) ^a	(16)
Treatment after ICD implantation						
β-Blockers	(89)	(84)	(90)	(87)	(100)	(91)
Sodium channel blockers	(9)	(2)	(4)	(40)	(20) ^a	(9)

Values are presented as mean ± standard deviation and numbers in parentheses are percentages.

ICD, implantable cardioverter-defibrillator; LQT, Long QT; Multiple, multiple mutations; Other, non genotyped; LCSD, left cardiac sympathetic denervation.

^aIndicates P-value <0.05 for comparison between LQT1, LQT2, LQT3, and multiple mutations.

models. Each risk variable was assigned for a score roughly reflecting the relative parameter estimate in the model (two points for QTc ≥ 550 ms and one point for each of the other variables: QTc 500–550 ms, syncope while on β-blockers treatment, LQT2 genotype, and multiple mutations). The score was calculated for each patient according to the total sum of his risk variables.

Results

The baseline clinical characteristics, the medical treatment, and the outcomes after ICD implantation are shown in Table 1. Females comprised 70% of the entire cohort. The mean age of ICD implantation was 26 ± 14 years. The mean QTc was 504 ± 61 ms. Prior to ICD implantation, the majority of the patients were treated with β-blockers, 4% were treated with sodium channel blockers (SCB), 14% had pacemaker, and 1% had left cardiac sympathetic denervation (LCSD). The genotype was identified in about two-thirds of the patients, among which third were LQT1, about half were LQT2, 11% were LQT3,

and 7% had multiple mutations. The most common indication for ICD implantation was syncope off therapy.

In total, 42 subjects experienced at least one appropriate shock. Device programming data was only available for 18 patients with appropriate shocks (n = 18; 43%). All of these patients were programmed to high rate cut-off (cycle length 291 ms ± SD of 40 ms) with programmed delay (11 s ± SD of 2.6 s).

Overall, 89% of the patients were treated with β-blockers and 9% were treated with SCB after ICD implantation. During a mean follow-up time of 9.2 ± 4.9 years, the cumulative probabilities of appropriate shock and inappropriate shock were 23% and 34%, respectively.

First appropriate shock

In a univariate Kaplan–Meier analysis (Figure 1A) only baseline QTc duration ≥ 550 ms was significantly associated with risk for appropriate shock. Notably, age (categorized at either 15 or 20 years), gender,

Figure 1 (A) The Kaplan–Meier curves of appropriate shocks by QTC duration categorized at 550 ms. (B) The Kaplan–Meier curves of appropriate shocks in patients with known genotype are categorized by genotype. ICD, implantable cardioverter-defibrillator; LQT, Long QT; Multiple, multiple mutations.

and the combination of these two variables were not significantly predictive of appropriate shock in the univariate (all *P*-values > 0.10).

In a multivariate analysis adjusted for age and stratified by gender (Table 2), QTC duration ≥550 ms and syncope while on β-blocker treatment were significantly associated with appropriate shocks, and β-blockers trended towards being protective (*P* = 0.075). The results remained similar after adjusting multivariate model for family history of SCD and ACA, which were not significant predictors [hazard ratio (HR) 0.76, confidence interval (CI) 0.35–1.66; *P* = 0.490 and HR 0.86, CI 0.40–1.86; *P* = 0.702 accordingly].

We conducted a second analysis restricted to patients with known genotype. In the univariate Kaplan–Meier analysis (Figure 1B), LQT2 and multiple mutations were associated with higher risk for appropriate shock. We further explored the location of the mutations as a predictor of appropriate ICD shock. In LQT1 the missense C-Loop mutations were not predictive, while in LQT2 the pore mutations trended to be at higher risk in the univariate Kaplan–Meier analysis

Table 2 Multivariable model for the risk of first appropriate ICD shock

Variables	Hazard ratio ^a	95% CI	P-value
QTC ≥550 ms	3.94	2.08–7.46	<0.001
Prior syncope while on β-blockers	1.92	1.01–3.65	0.047
Time dependent β-blockers treatment	0.52	0.26–1.07	0.075
LQT2 vs. LQT1 ^b	2.47	0.93–6.52	0.059
LQT3 vs. LQT1 ^b	0.39	0.05–3.34	0.392
Multiple mutations vs. LQT1 ^b	2.87	0.88–9.42	0.081

CI, confidence interval; ICD, implantable cardioverter-defibrillator; LQT, Long QT.

^aThe model is stratified by gender and adjusted for the age of implantation.

^bThe hazard ratios for genotype were acquired when the same model was applied for patients with known genotype.

(0.052). In the multivariate model (Table 2) the genotype was not a significant predictor of the first appropriate shock.

We analysed the effect of TdP, by including this parameter into the model and then by excluding patients who had TdP and no history of syncope on beta-blocker treatment, and the effect size was similar.

Recurrent appropriate shocks

There were 117 ventricular arrhythmia episodes of which 63 were VT and 54 were ventricular fibrillation. Consistent with the first appropriate shock analysis the predictors of recurrent appropriate shocks were QTC duration (500–549 ms and ≥550 ms) and syncope while on β-blockers. For patients with known genotype, LQT2, and multiple mutations were significantly associated with increased risk for appropriate shocks.

Risk score for the first and recurrent appropriate shocks

We attempted to validate the M-FACT score⁷ that was previously shown to identify patients at low risk for appropriate shocks. The M-FACT risk score includes the following parameters: absence of syncope or ACA while on therapy for >10years, QTC, history of prior ACA, presence of syncope or ACA while on therapy, and age at ICD implantation. As the M-FACT score does not utilize genetic data, we first analysed the predictive capacity in the entire patient population (Figure 2A). The M-FACT score was very useful for stratifying high-risk patients, however, it failed to identify low-risk patients and even patients with a score of 0 had a considerable rate of appropriate shocks: 13% after 6 years and 16% after 10 years. This was consistent even after excluding one patient that had appropriate shock in the first 10 days after the implantation. We, therefore, created a new risk score incorporating clinical variables and genetic data that were significant in the multivariate models (Tables 2 and 3) the score calculation is shown in Table 4. In order to allow a comparison between the new score and the M-FACT score, we only analysed patients with available genetic data (Figure 2B and C). There were 19 patients with score of 0, 47 patients with score of 1, 36 patients with score of

Figure 2 The Kaplan–Meier curves of appropriate shocks by the M-FACT risk score all study patients (A) M-FACT score only patients with known genotype (B) and the new risk score (C). ICD, implantable cardioverter-defibrillator.

2, and 35 patients with score of ≥ 3 , the cumulative probabilities of the first appropriate shock were 0%, 15%, 23%, and 50%, respectively after 10 years, P -value < 0.001 . Consistently, the risk score was highly predictive of the rate of appropriate shocks per 100 patient years, $P < 0.001$ (Figure 3).

Figure 3 Event rate of appropriate ICD rendered shocks (including recurrent) by the new risk score. Event rate of all the appropriate shocks (including recurrent) per 100 patients-years were calculated by dividing the total number of appropriate shocks during the period of follow up by person-years and multiplying the results by 100. The negative binomial regression was used to calculate the P -value. ICD, implantable cardioverter-defibrillator.

β -Blocker treatment

Overall β -blockers trended to be protective for the first appropriate shock. As expected patients with cardiac events on β -blockers before the ICD placement had appropriate shocks while on β -blockers. Importantly, even patients without a history of events on β -blockers still had appropriate shocks while on treatment. β -Blockers were not protective for recurrent ICD appropriate shocks. Thus, if a patient had an appropriate shock on β -blocker, he had a high risk for subsequent shocks on β -blocker.

We further analysed the role of specific subtypes of β -blockers. There were 63 patients that were treated with atenolol (median dose and interquartile range at start of follow-up, respectively: 50 mg, 25 mg; median dose and interquartile range at end of follow-up, respectively: 50 mg, 50 mg). Forty-eight patients were treated with metoprolol (50 mg, 25 mg; 100 mg; 100 mg), 53 with nadolol (40 mg, 20 mg; 40 mg; 60 mg), and 22 with propranolol (60 mg, 50 mg; 120 mg, 110 mg). Patients that were treated with nadolol had the lowest cumulative probability of first appropriate shock and those who were treated with metoprolol had the highest probability (Figure 4A). The rate of recurrent shocks was highest with metoprolol and similar among other β -blockers (Figure 4B).

Sensitivity analyses

As some of the post-implantation shocks may be theoretically associated with the mechanical stimulation of the newly implanted lead, we performed a sensitivity analysis with the follow-up starting at 11 days after the implantation. The results were similar (see [Supplementary material online, Tables S1 and S2, Figures S1 and S2](#)).

To test our risk score in a subpopulation without a history of TdP, we excluded this sub population and the results were similar (see [Supplementary material online, Figure S3](#)).

Table 3 Multivariable model for the risk of recurrent appropriate ICD shocks

Variables	Hazard ratio ^a	95% CI	P-value
QTc 500–549 ms vs. QTc <500 ms	1.68	1.01–2.81	0.048
QTc ≥550 ms vs. QTc <500 ms	3.66	2.34–5.72	<0.001
Prior syncope while on β-blockers	1.87	1.28–2.72	0.001
Time dependent β-blockers treatment	1.03	0.63–1.68	0.917
LQT2 vs. LQT1 ^b	2.10	1.22–3.61	0.008
LQT3 vs. LQT1 ^b	0.37	0.08–1.60	0.183
Multiple mutations vs. LQT1 ^b	2.87	1.49–5.53	0.002

CI, confidence interval; ICD, implantable cardioverter-defibrillator; LQT, Long QT.
^aThe model is stratified by gender and adjusted for the age of implantation.
^bThe hazard ratios for genotype were acquired when the same model was applied for patients with known genotype.

Table 4 Risk score calculation

Risk variables	0 points	1 point	2 points
QTc	≤499 ms	500–549 ms	≥550 ms
Prior syncope while on β-blockers	No	Yes	
LQT2	No	Yes	
Multiple mutations	No	Yes	

The calculated score is the sum of the points of all four risk variables categories.

Inappropriate implantable cardioverter-defibrillator shocks and consequences

There were 88 inappropriate ICD shocks, 41% were caused due to T-wave oversensing, 14% due to trial flutter/fibrillation, 22% due to supraventricular tachyarrhythmias, and 20% were caused by electromagnetic interference. The event rates were not different between ICD manufacturers. The only predictor of inappropriate shock was age ≤20 years (log rank *P*-value = 0.029); other variables including gender, baseline heart rate, QTc and genetic data were insignificant (*P* > 0.05). Most of the inappropriate shocks ended without further consequences. Two patients (2%) had appropriate shocks preceded by inappropriate shocks.

Discussion

This study provides several important clinical implications for the use of ICD in high risk LQTS patients who did not experience ACA. First, QTc duration is the most important predictor for appropriate shock. Second, syncope while on β-blockers treatment was significantly predictive of both the first and recurrent appropriate shocks in the multivariate models and this provides further strength to the current knowledge. Third, β-blocker therapy remains an important treatment for the prevention of the first shock even in this very high-risk

Figure 4 The Kaplan–Meier curves of first appropriate shocks in patients that were treated with different types of β-blockers (A) and event rate of recurrent appropriate ICD rendered shocks by different types of β-blockers (B). Event rate of all the appropriate shocks (either first or recurrent) per 100 patients-years were calculated by dividing the total number of appropriate shocks during the period of follow up by person-years and multiplying the results by 100. ICD, implantable cardioverter-defibrillator.

population, however, in patients that had appropriate shock on β-blocker therapy, it is not protective for subsequent shocks. Lastly, inappropriate shocks are very prevalent in this group of patients, mostly due to T-wave oversensing, however, we showed most of which are terminated without further sequelae.

During a mean follow-up of 9 years, the cumulative probability of the first appropriate shock was 23% and none of the study patients died from LQTS related causes. We recognize the possibility that not all shocks are necessarily lifesaving. To put this finding in perspective we explored the mortality rate in patients from the Rochester LQTS registry that had LCSD but did not have ICD placement. Among 41 patients that we identified, the mortality rate was 17%, and 7% had ACA (from birth to the age of 50). This finding demonstrates the effectiveness of ICD to prevent SCD, as we demonstrated in a previous study.³

As expected, the duration of the QTc was shown to be highly associated with the risk for appropriate shock. This is in agreement with previous studies in which the QTc duration was associated with increased risk for cardiac events in LQTS patients in general and particularly in those implanted with ICD.^{7,23–25}

We showed that β -blocker treatment was associated with lower risk for the first appropriate shock. Specifically, nadolol was associated with the lowest risk and metoprolol with the highest risk. The risk with metoprolol was consistently higher even for recurrent appropriate shocks. This finding was demonstrated in previous studies, and thus metoprolol utilization should probably be avoided in LQTS patients.²⁶ Our finding that syncope while on β -blockers treatment, but not syncope off treatment, was associated with the risk for appropriate shock provides further support for the current guidelines, and it stresses the importance of β -blocker treatment in the management of LQTS.^{27–30} With that said, one should always keep in mind that β -blockers treatment failures do occur, more commonly in patients with LQT2 and LQT3.^{14,31} Importantly, we showed that patients who had an appropriate shock while on β -blockers were more likely to experience subsequent appropriate shocks despite β -blockers treatment.

Multiple mutations have been previously shown to be at higher risk for appropriate shocks in a univariate analysis by Schwartz *et al.*⁷; however, a multivariate analysis was not available. Herein, we show that multiple mutations and LQT2 genotype exhibited higher risk when compared with LQT1 among those who received a prophylactic ICD. This could be explained by the higher efficacy of β -blockers in LQT1 as compared with LQT2 and LQT3,¹⁴ and the higher risk nature of multiple mutations.^{32,33} We were not able to show increased risk related to LQT3 and in specific mutation locations in LQT1¹⁵ and LQT2,¹⁶ most probably due to a lack of power in the current study.

Inappropriate ICD therapies were very frequent in our study, even more so than appropriate therapies. Most of which were attributable to oversensing, and the second common cause was supraventricular arrhythmias, which is well correlated with previous studies.⁷

Limitations

Our study has several limitations. First, this is a retrospective analysis of a prospective registry and no control group was available. Second, due to population size limitations we had no validation cohort. Third, we had very few patients with LQT3 genotype, and therefore, the risk of this group of patients may be underestimated. Lastly, due to the long period of the follow-up time, some time-dependent differences in the medical and device treatment between the patients may exist.

Conclusions

In conclusion, in high risk LQTS patients implanted with ICD for primary prevention, QTc duration, history of syncope while on β -blockers treatment, LQT2 genotype, and multiple LQTS-associated mutations were associated with the risk for appropriate ICD shocks. These data can be used for risk stratification in high-risk patients who are candidates for ICD implantation for primary prevention. Inappropriate shocks are very prevalent in this group of patients and dedicated programming to avoid T-wave oversensing is required.

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IMAGES IN ELECTROPHYSIOLOGY

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Electroanatomical delimitation of recipient atrium in heart transplantation

Adolfo Fontenla*, Maria Lopez-Gil, and Fernando Arribas

Cardiology Department, University Hospital 12 de Octubre, Avenida de Cordoba SN, Madrid 28041, Spain

* Corresponding author. Tel: +34 69 901 2607; fax: +34 91 390 8071. E-mail address: drfontenla@gmail.com

A 42-year-old patient with previous history of biatrial orthotopic heart transplantation (a technique prone to develop atrial arrhythmias) was submitted for electrophysiological study due to persistent atrial tachycardia.

Activation mapping with *Ensite Precision*TM showed a counter-clockwise activation in the donor atrium around the tricuspid annulus and a dissociated sinus rhythm in the recipient atrium. The isolated recipient atrium was displayed as an area of random coloured spots (we named this ‘patchwork sign’) showing an evident boundary dividing the recipient and the donor atria (arrows). Cavotricuspid isthmus ablation in donor atrium interrupted the arrhythmia without change in the activation of the recipient (*).

Supplementary material is available at *Europace* online.