UZUN QT SENDROMLARINDA ICD ENDİKASYONLARI

DR. MURAT SUCU

GAZİANTEP ÜNİVERSİTESİ TIP FAKÜLTESİ

KARDİYOLOJİ AD.

Tanım

- QT intervali = QRS Dalgasının Başından T Dalgasının Sonuna Kadar Olan Süre.
- Ventrikül Miyokardının Aktivasyon ve İstirahat Süresinin Toplamıdır.
- Bazett formülü: QT/ $\sqrt{RR}$
- Ortalama Tanı Yaşı:14
- Tedavi Edilmemiş Hastalarda Yıllık AKÖ oranı:%0,9-0,33
- Senkop Geçiren Hastalarda AKÖ oranı %5

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STATE-OF-THE-ART PAPER

Long QT Syndrome

Ilan Goldenberg, MD, Arthur J. Moss, MD
Rochester, New York

Table 1 Suggested Bazett-Corrected QTc Values for Diagnosing QT Prolongation

Rating	1–15 yrs	Adult Male	Adult Female
Normal	<440	<430	<450
Borderline	440–460	430–450	450–470
Prolonged	>460	>450	>470

Tanı

- Tekrarlanan EKG'lerde QTc$\geq 480\ msn$
- Yada 3 den fazla Risk Faktörünün Varlığı
  - Kanıt Düzeyi Ic

Risk Skor(Schwartz Skor)

Findings			Points
ECG [1]	QTc [2]	≥480 ms	3
		=460-479 ms	2
		=450-459 ms (in males)	1
		≥480 ms during 4th minute of recovery from exercise stress test	1
	Torsade de pointes [3]		2
	T wave alternans		1
	Notched T wave in 3 leads		1
	Low heart rate for age [4]		0.5
Clinical history	Syncope [3]	With stress	2
		Without stress	1
Family history	Family member(s) with definite LQTS [5]		1
	Unexplained sudden cardiac death at age <30 years in immediate family [5]		0.5
Total score			

≤1.0 Puan = Düşük ihtimal LQTS
1.5-3.0 Puan = Muhtemel LQTS
≥3.5 Puan = Yüksek İhtimalle LQTS

Edinsel Nedenler

- Metabolik
  - Hipopotasemi
  - Hipomagnezemi
  - Hipokalsemi
  - Anoreksia
  - Hipotiroidi
- İlaçlar
  - Antiaritmikler (Sotalol, Amiodaron, vb.)
  - Antibiotikler (Makrolidler, Fluoroquinolonlar)
  - Psikotropik İlaçlar (Halodol, TCAs, Thioridazin)
  - SSRI (Risperidon, Methadon, Droperidol) Proteaz inhibitörleri

Edinsel Nedenler

- Miyokard İskemisi ve İnfarktüsü
- İntrakraniyal Kanamalar
- Hipotermi
- HIV

İyon Kanallarının Hastalığı

- 13 Gende Mutasyon, Potasyum, Sodyum, Kalsiyum Voltaj Bağımlı Kanallar.
  - Aksiyon Potansiyelinin uzaması
  - Erken After Depolarizasyon
- Artmış Sempatik Aktivite
  - Kalbin Sempatik İnervasyonundaki Dengesizlik.

LQTS GENLERİ

Gene	Syndrome	Frequency	Locus	Protein (Functional Effect)
KCNQ1 (LQT1)	RWS, JLNS	40—55	11p15.5	Kv7.1 (↓)
KCNH2 (LQT2)	RWS	30—45	7q35—36	Kv11.1 (↓)
SCN5A (LQT3)	RWS	5—10	3p21—p24	NaV1.5 (↑)
ANKB (LQT4)	RWS	<1%	4q25—q27	Ankyrin B (↓)
KCNE1 (LQT5)	RWS, JLNS	<1%	21q22.1	MinK (↓)
KCNE2 (LQT6)	RWS	<1%	21q22.1	MiRP1 (↓)
KCNJ2 (LQT7)	AS	<1%	17q23	Kir2.1 (↓)
CACNA1C (LQT8)	TS	<1%	12p13.3	L-type calcium channel (↑)
CAV3 (LQT9)	RWS	<1%	3p25	Caveolin 3 (↓)
SCN4B (LQT10)	RWS	<1%	11q23.3	Sodium channel-β4 (↓)
AKAP9 (LQT11)	RWS	<1%	7q21—q22	Yotiao (↓)
SNTA1 (LQT12)	RWS	<1%	20q11.2	Syntrophin α1 (↓)
KCNJ5 (LQT13)	RWS	<1%	11q24	Kir3.4 (↓)

LQTS indicates long-QT syndrome; *KCNQ1*, potassium voltage-gated channel, KQT-like subfamily, member 1; RWS, Romano-Ward syndrome; JLNS, Jervell and Lange-Nielsen syndrome; *KCNH2*, potassium voltage-gated channel, subfamily H, member 2; *SCN5A*, sodium voltage-gated channel, type V, α subunit; *ANKB*, ankyrin B; *KCNE1*, potassium voltage-gated channel, ISK-related subfamily, member 1; MinK, minimal K+ ion channel; *KCNE2*, potassium voltage-gated channel, ISK-related subfamily, member 2; *MiRP*, MinK-related peptide 1; *KCNJ2*, potassium channel, inwardly rectifying, subfamily J, member 2; AS, Andersen syndrome; *CACNA1C*, calcium voltage-dependent channel, L type, α-1C subunit; TS, Timothy syndrome; *CAV3*, caveolin 3; *SCN4B*, sodium voltage-gated channel, type IV, β subunit; *AKAP9*, A-kinase anchor protein 9; *SNTA1*, syntrophin α1; *KCNJ5*, potassium channel, inwardly rectifying, subfamily J, member 5.

Functional effect: (↓) loss-of-function or (↑) gain-of-function at the cellular in vitro level.

Otozomal Dominant LQTS

- LQTS7 (Andersen–Tawil sendromu),
  - Uzamış QT interval
  - Belirgin U dalgası
  - Polimorfik yada Bidireksiyonel VT
  - Fasiyal dismorfizim
  - Hiper/Hipokalemik periyodik Paralizi
- LQTS8 (Timothy Sendromu),
  - Uzamış QT
  - Sindaktili
  - Kardiyak Malformasyonlar
  - Otizim
  - Dismorfizim
- Romano–Ward sendromu
  - Prevalans:1/2500
  - QT intervalinde uzama
  - LQT1–6 ve LQT9–13

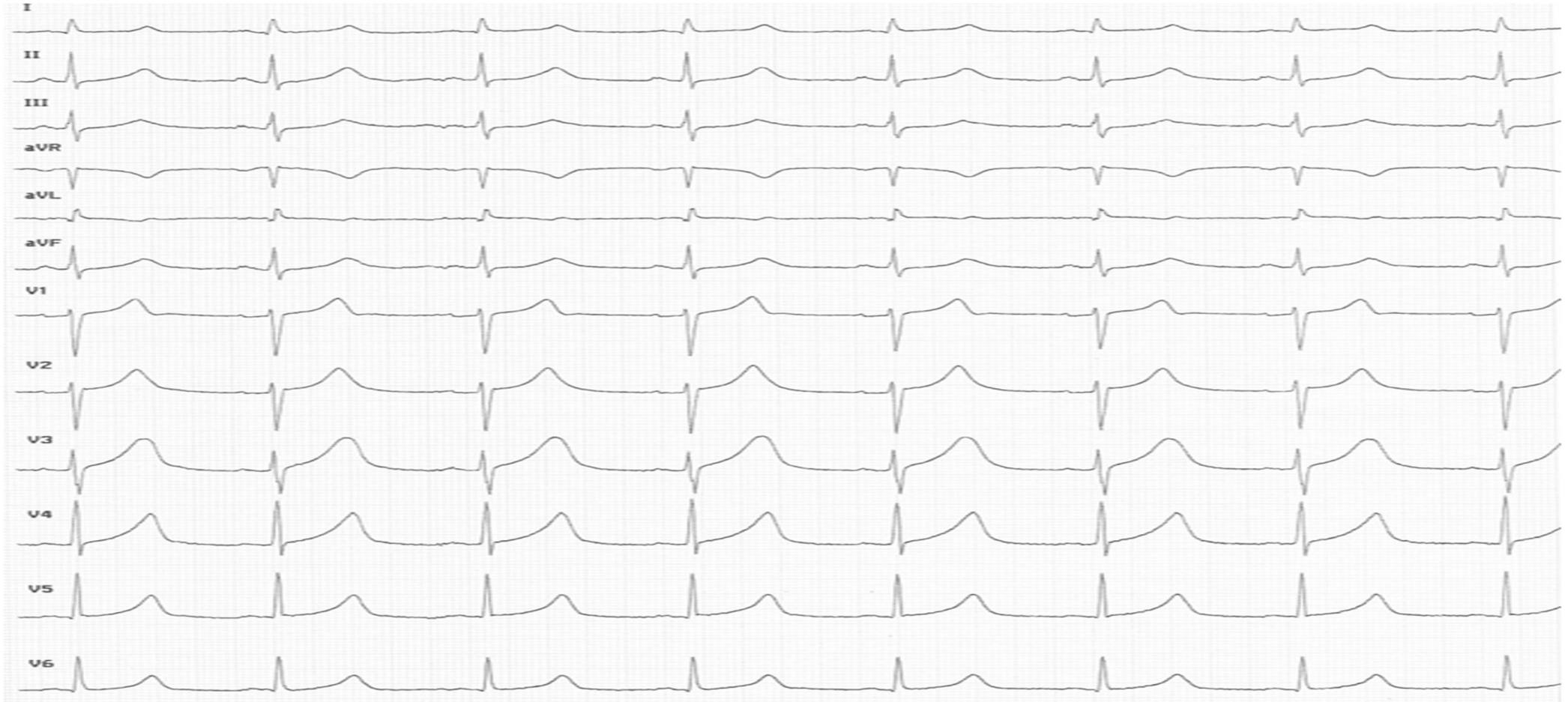
Otozomal Resesif LQTS

- Jervell and Lange–Nielsen Sendromu
  - Uzamış QT İntervali
  - Konjenital Sağırlık.

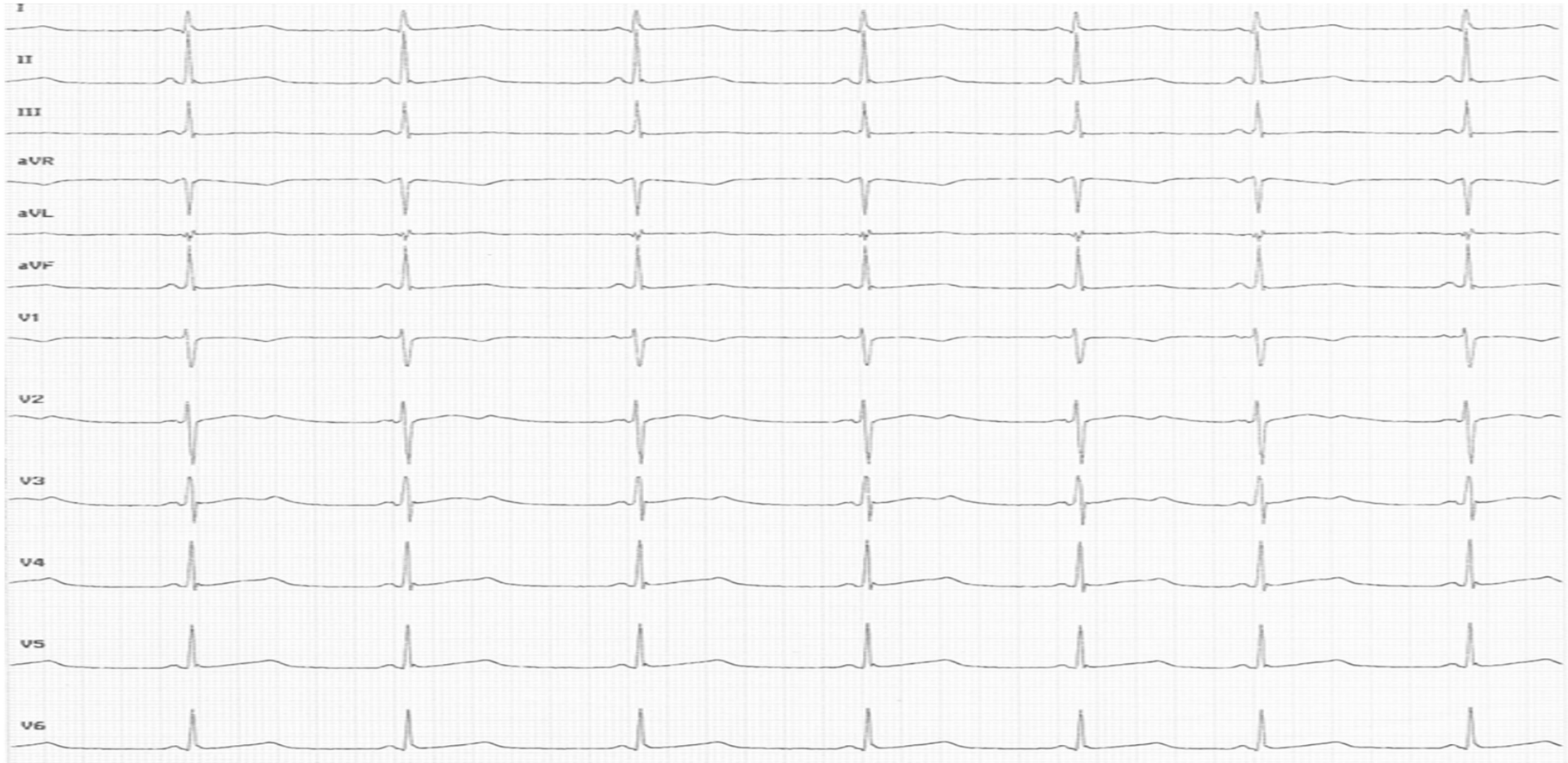
Fenotiplerin Genlerle Korelasyonu

Gen	Fenotip	Ortalama QTc	ST-T-Dalga Morfoloji	Kardiyak Olay İnsidansı	Kardiyak Olay Tetikleyici	Ani Ölüm Riski
KCNH2	LQTS tip 2	480 msn	Bifid T-dalgası	%46	Ses, Emosyonel Stres, Ekzersiz, Uyku	6%-8%
KCNQ1	LQTS tip 1		Geniş T-dalgası	%63	Eksersiz, Emosyonel Stres	6%-8%
SCN5A	LQTS tip 3	~490 msn	Uzun ST, Küçük T	%18	Uyku	6%-8%

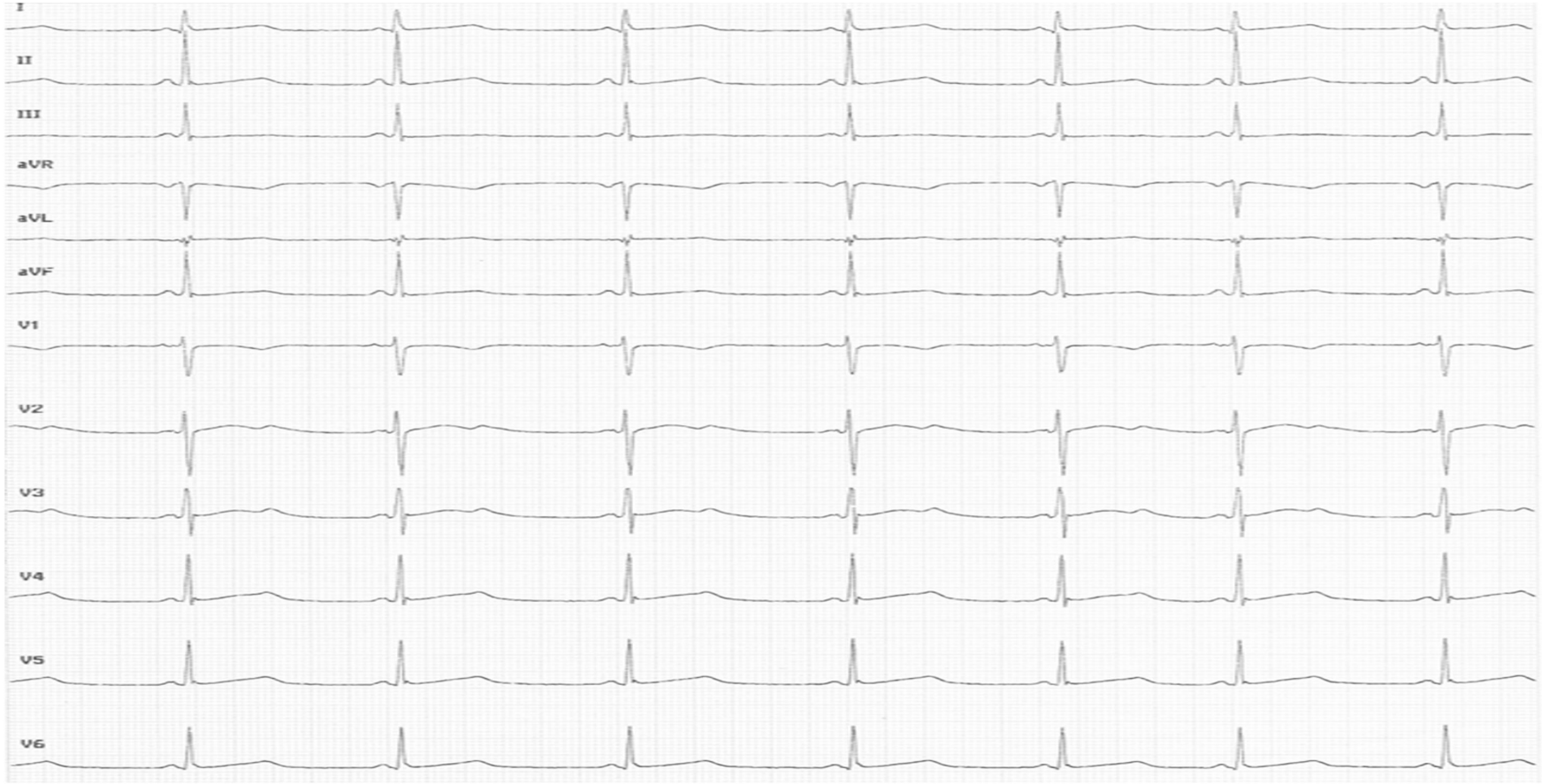
Uzun QT Tip 1



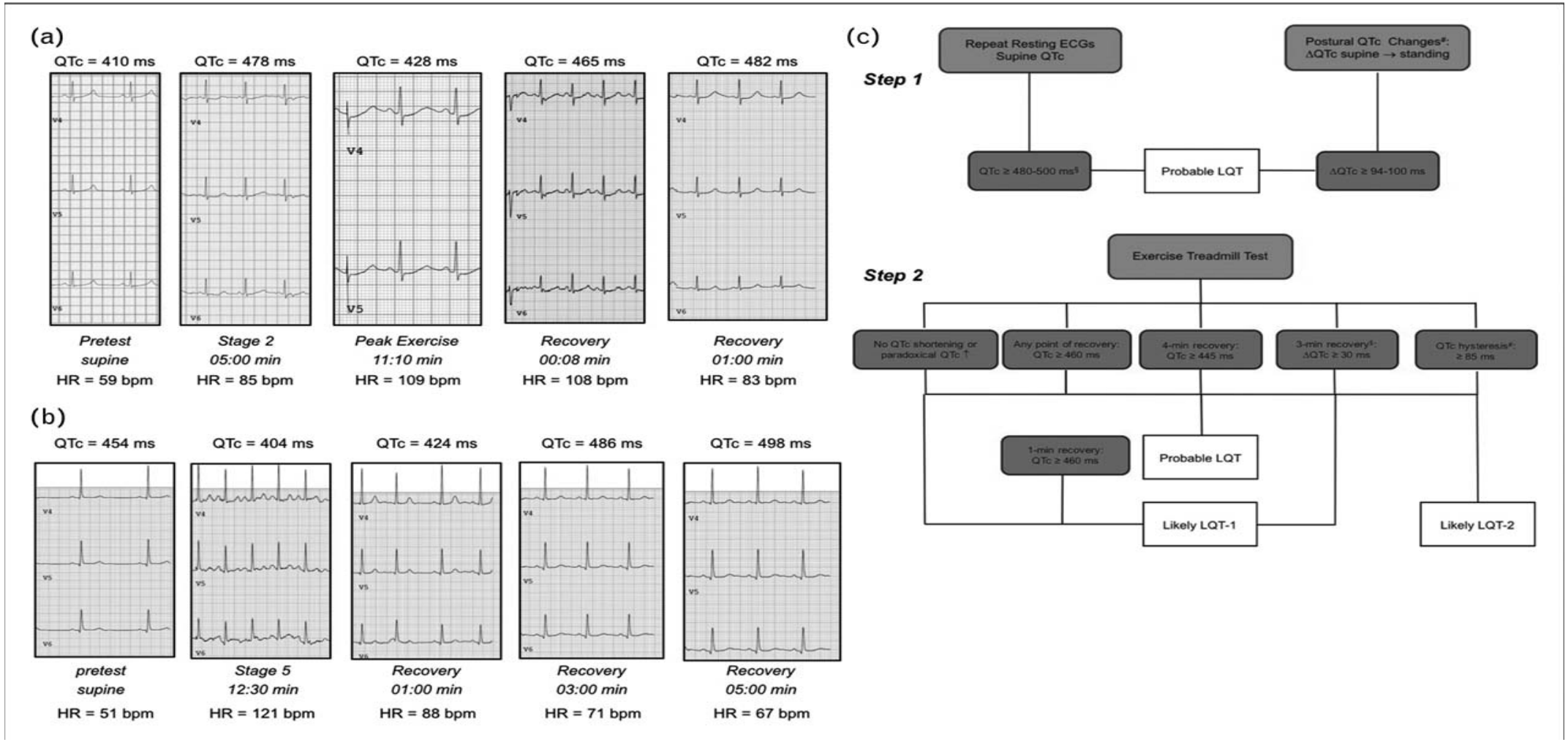
Uzun QT Tip 2



Uzun QT Tip 3



Egzersiz Testi



Epinefrin Testi

- 0.025 mg/(kg x dk) -0.2 mg/(kg x dk).
- QT interval en az 30 msn uzama 0.1 mg/(kg x dk).
- Yüksek Kalp Hızında Yalancı Pozitif test (%25).
- β Bloker Sonucu Etkiliyor.

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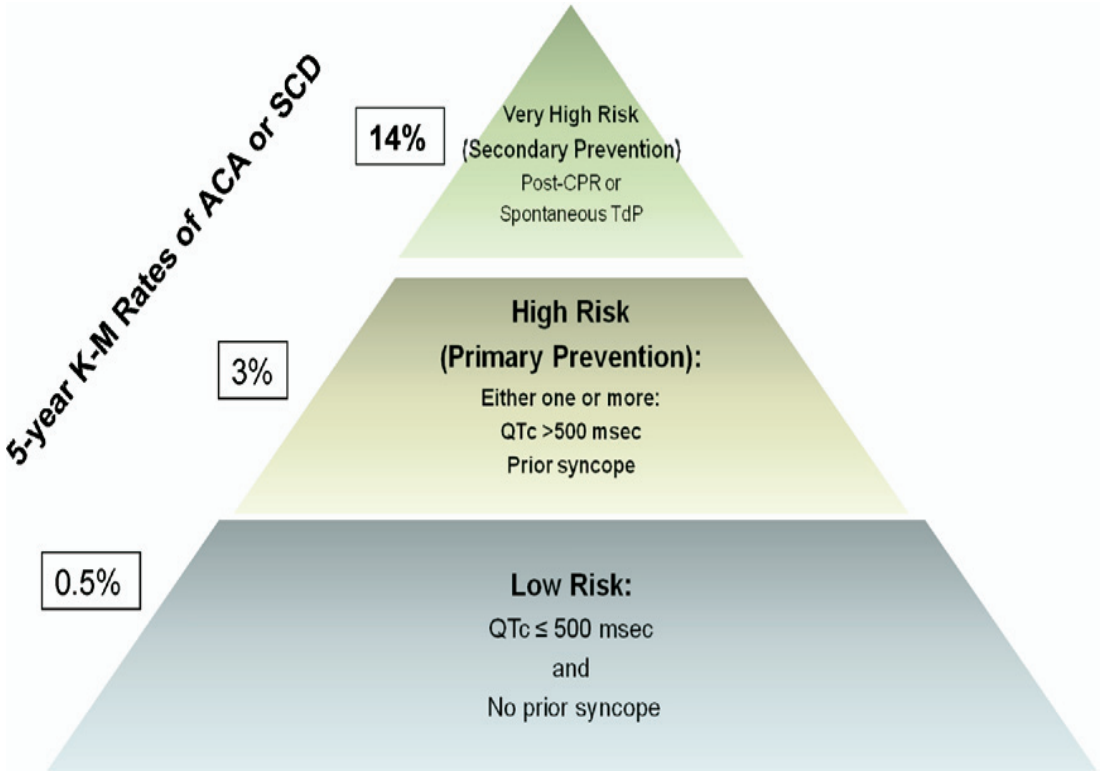

Suggested Risk-Stratification Scheme for ACA or SCD in LQTS Patients

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Ilan Goldenberg, MD, Arthur J. Moss, MD
Rochester, New York

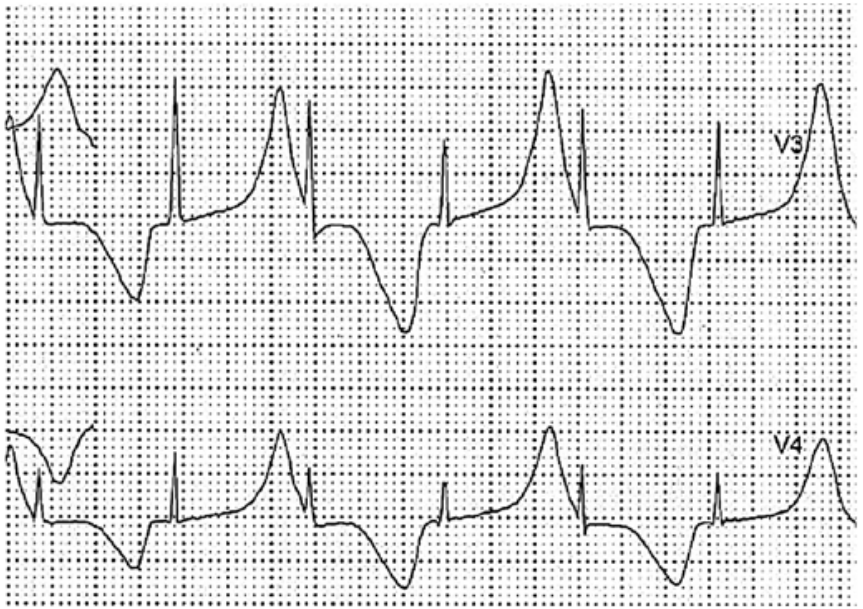
Table 3 Age-Specific Risk Factors for Life-Threatening Cardiac Events in LQTS Patients*

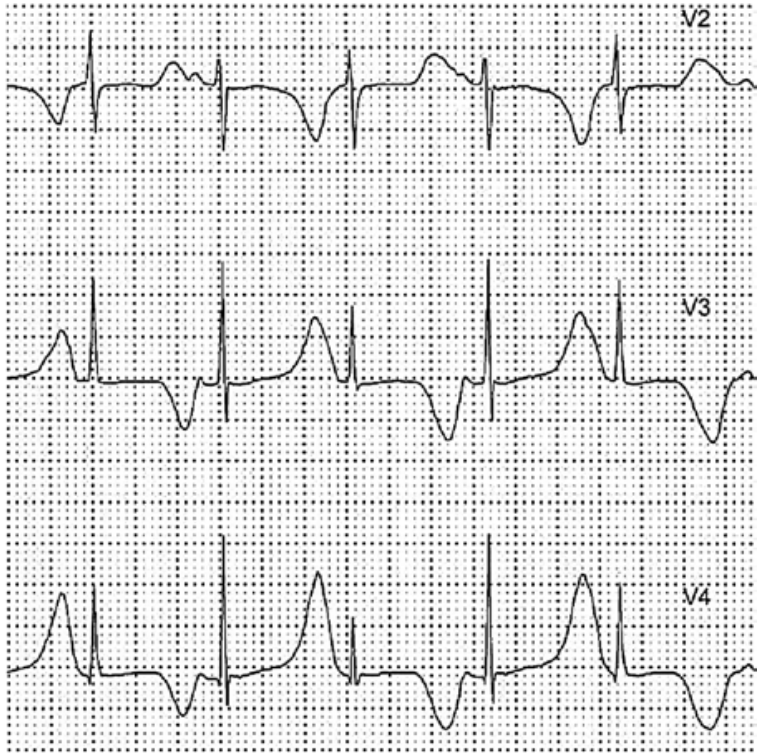
Age Group (Ref. #)	Risk Factor	Hazard Ratio (p Value)	Beta-Blocker Efficacy, % Reduction (p Value)
Childhood (1–12 yrs) (33)	Male gender	3.96 (<0.001)	73% (0.002)
	QTc >500 ms	2.12 (0.02)	
	Prior syncope		
	Recent (<2 yrs)	14.34 (<0.001)	
	Remote (≥2 yrs)	6.45 (<0.001)	
Adolescence (10–20 yrs) (28)	QTc >530 ms	2.3 (<0.001)	64% (0.01)
	Syncope		
	≥2 syncopal events in past 2 yrs	18.1 (<0.001)	
	1 syncopal event in past 2 yrs	11.7 (<0.001)	
	≥2 syncopal events in past 2–10 yrs	5.8 (<0.001)	
	1 syncopal events in past 2–10 yrs	2.7 (<0.001)	
Adulthood (18–40 yrs) (29)	Female gender	2.68 (<0.05)	60% (<0.01)
	QTc duration		
	QTc ≥500 ms	6.35 (<0.01)	
	QTc 500–549 ms	3.34 (<0.01)	
	Prior syncope	5.10 (<0.01)	
Adulthood (41–60 yrs) (53)†	Recent syncope (<2 yrs)	9.92 (<0.001)	42% (0.40)‡
	QTc >530 ms	1.68 (0.06)	
	LQT3 genotype	4.76 (0.02)	

Fenotip–Genotip İle İlişkili Risk Sınıflaması

Clinical risk factors[a]	Combined risk factors[a]	Genetic risk factors[a]
Extremely high risk (≥80%)		
QTc ≥ 600 ms		Timothy syndrome (LQT-8)
≥10 cardiac events < age 18 years		Jervell–Lange-Nielsen syndrome
High risk (≥50%)		
QTc ≥ 550 ms	LQT-1 + male 0–14 years old	Compound or digenic heterozygosity
≥2 but <10 cardiac events before age 18 years	LQT-2 + female 15–40 years old	Certain LQT-1 mutations
Cardiac event <age 7 years		C-loop mutations
Cardiac event on appropriate beta-blocker treatment		A341V
		Certain LQT-2 mutations
		Pore mutations
		CALM1 or CALM2 mutations
Intermediate risk (30–49%)		
QTc = 500–549 ms		LQT-3
<2 cardiac events < age 18 years		
Female sex		
Low risk (<30%)		
QTc < 500 ms	LQT-1 + female 0–14 years old	LQT-1 minor genotypes
No cardiac event < age 18 years	LQT-2 + male 0–40 years old	

T Dalga Alternans





Tedavi

- Primer Korunma
  - β Bloker
  - Yoğun ve Ağır Spor Aktivitelerinden Kaçınmak
  - Left Kardiak Sempatik Denervasyon
- Sekonder Korunma
  - β Bloker
  - QT Uzatan Ajanlardan Kaçınmak
  - ICD

Proflatik ICD

- ICD Tedavisi Bireyselleştirilmelidir
- Yüksek Riskli Hastalar
  - Kadınlarda LQT2 ve QTc 500 msn,
  - QTc 500 ms ve elektriksel Düzensizlik Bulguları (T dalga Alternans) ve Yüksek Riskli Genetik Profil (İki Mutasyonla beraber , Jervell ve Lange–Nielsen Sendromu veya Timothy Sendromu).

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CLINICAL RESEARCH

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

Yitschak Biton[1,2]*, Spencer Rosero[3], Arthur J. Moss[1], Ilan Goldenberg[2], Valentina Kutyifa[1], Scott McNitt[1], Bronislava Polonsky[1], Jayson R. Baman[1,4], and Wojciech Zareba[1]

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 ≥ 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 ≥ 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.

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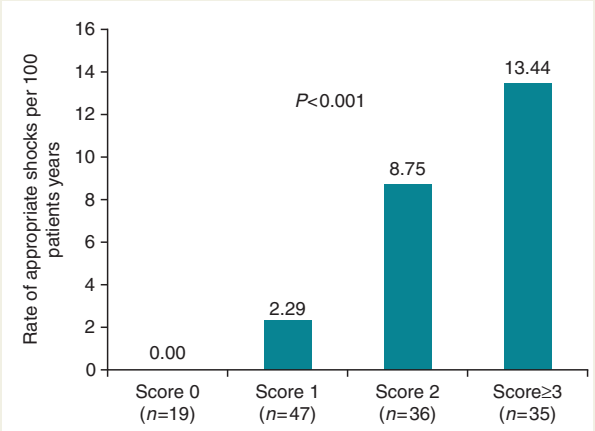

Figure 3 Event rate of appropriate ICD rendered shocks (including recurrent) by the new risk score. Event rate of all the appropriate shocks (including recurrent) per 100 patients-years were calculated by dividing the total number of appropriate shocks during the period of follow up by person-years and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

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.

[a]The model is stratified by gender and adjusted for the age of implantation.

[b]The hazard ratios for genotype were acquired when the same model was applied for patients with known genotype.

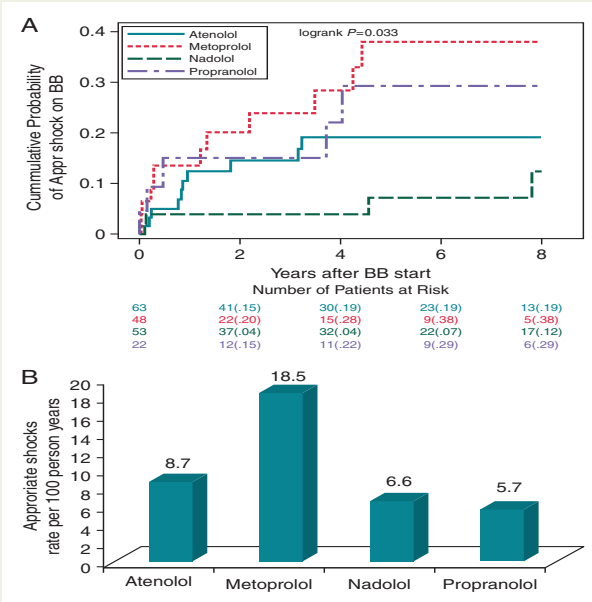

Figure 4 The Kaplan–Meier curves of first appropriate shocks in patients that were treated with different types of β-blockers (*A*) and event rate of recurrent appropriate ICD rendered shocks by different types of β-blockers (*B*). Event rate of all the appropriate shocks (either first or recurrent) 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.

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Table 3. M-FACT Risk Score

	−1 Point	0 Point	1 Point	2 Points
Event free on therapy for >10 y	Yes			
QTc, ms		≤500	>500 to ≤550	>550
Prior ACA		No	Yes	
Events on therapy		No	Yes	
Age at implant, y		>20	≤20	

M-FACT indicates M for Minus 1 point for being free of cardiac events while on therapy for >10 y; F for Five hundred and Five hundred and Fifty millisecond QTc; A for Age ≤20 y at implant; C for Cardiac arrest; T for events on Therapy; ACA, aborted cardiac arrest. Modified from Ref 72.

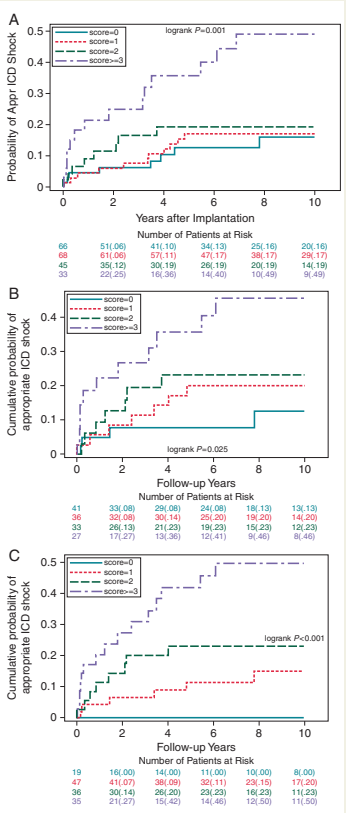

Figure 2 The Kaplan–Meier curves of appropriate shocks by the M-FACT risk score all study patients (*A*) M-FACT score only patients with known genotype (*B*) and the new risk score (*C*). ICD, implantable cardioverter-defibrillator.

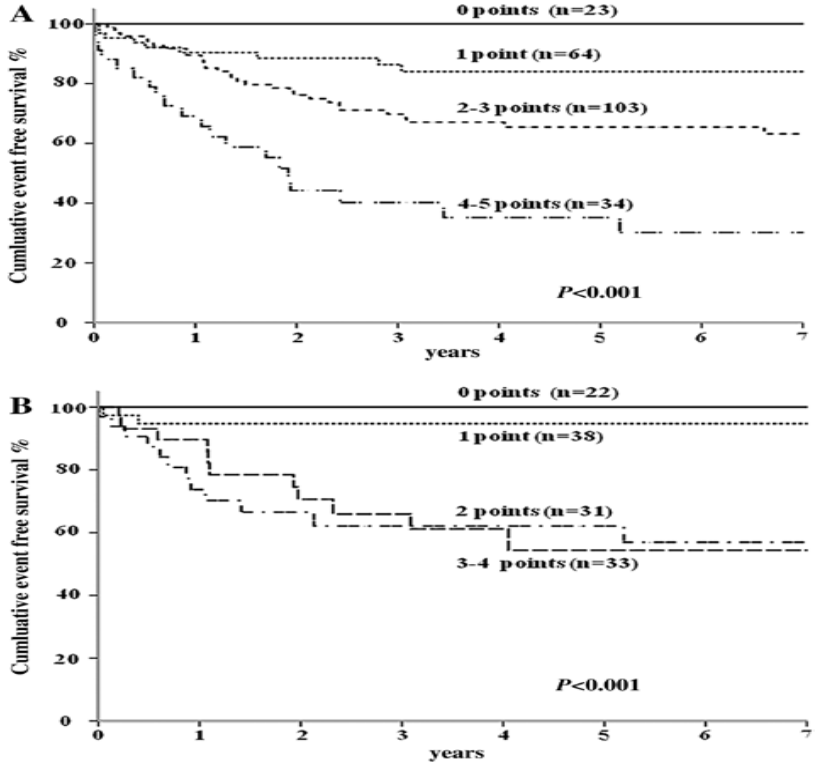

Figure 5. Cumulative event-free survival for a first appropriate implantable cardioverter-defibrillator shock according to an increasing risk score (M-FACT) in (**A**) all patients and (**B**) in patients with no prior aborted cardiac arrest (modified from Ref 72). M-FACT indicates M for Minus 1 point for being free of cardiac events while on therapy for >10 years; F for Five hundred and Five hundred and fifty millisecond QTc; A for Age ≤20 years at implant; C for Cardiac arrest; T for events on Therapy.

Risk stratification and management in Long QT Syndrome		
Recommendations	**Class[a]**	**Level[b]**
The following lifestyle changes are recommended in all patients with a diagnosis of LQTS: a. Avoidance of QT-prolonging drugs (www.crediblemeds.org). b. Correction of electrolyte abnormalities (hypokalaemia, hypomagnesaemia, hypocalcaemia) that may occur during diarrhoea, vomiting, or metabolic conditions. c. Avoidance of genotype-specific triggers for arrhythmias (strenuous swimming especially in LQT1 and exposure to loud noises in LQT2 patients).	I	B
Beta-blockers are recommended in patients with a clinical diagnosis of LQTS.	I	B
ICD implantation with the use of beta-blockers is recommended in LQTS patients with previous cardiac arrest.	I	B

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ESC GUIDELINES

2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

Risk stratification and management in Long QT Syndrome (continued)		
Recommendations	**Class[a]**	**Level[b]**
Beta-blockers should be considered in carriers of a causative LQTS mutation and normal QT interval.	IIa	B
ICD implantation in addition to beta-blockers should be considered in LQTS patients who experienced syncope and/or VT while receiving an adequate dose of beta-blockers.	IIa	B
Left cardiac sympathetic denervation should be considered in patients with symptomatic LQTS when: a. Beta-blockers are either not effective, not tolerated, or contra-indicated; b. ICD therapy is contra-indicated or refused; c. Patients on beta-blockers with an ICD experience multiple shocks.	IIa	C

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Risk stratification and management in Long QT Syndrome (continued)

Recommendations	Class[a]	Level[b]
Sodium channel blockers (mexiletine, flecainide, or ranolazine) may be considered as add-on therapy to shorten QT interval in LQT3 patients with a QTc >500 ms.	IIb	C
Implantation of an ICD may be considered in addition to beta-blocker therapy in asymptomatic carriers of a pathogenic mutation in *KCNH2* or *SCN5A* when QTc is >500 ms.	IIb	C
Invasive EPS with PVS is not recommended for sudden cardiac death risk stratification.	III	C

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Role of implantable cardioverter defibrillator therapy in patients with acquired long QT syndrome: A long-term follow-up

Gerold Mönnig[1*†], Julia Köbe[1†], Andreas Löher[2], Kristina Wasmer[1], Peter Milberg[1], Stephan Zellerhoff[1], Christian Pott[1], Sven Zumhagen[3], Razvan Radu[1], Hans H. Scheld[2], Wilhelm Haverkamp[4], Eric Schulze-Bahr[3], and Lars Eckardt[1]

[1]Department of Cardiology and Angiology, Division of Experimental and Clinical Electrophysiology, University Hospital Münster, Münster, Germany; [2]Department of Thoracic and Cardiovascular Surgery, University Hospital Münster, Münster, Germany; [3]Department of Cardiology and Angiology, Institute for Genetics of Heart Diseases (IfGH), University Hospital Münster, Germany; and [4]Department of Cardiology, Campus Virchow Clinic, Charité – University Medicine Berlin, Berlin, Germany

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Aims The use of implantable cardioverter defibrillators (ICD) in patients with torsade de pointes (TdP) and ventricular fibrillation in the presence of acquired long QT syndrome (aLQTS) is under debate, partly due to the fact that aLQTS is potentially reversible and currently no long-term follow-up data are available. We aimed to evaluate the long-term follow-up of patients with acquired long QT syndrome (aLQTS) who had received an implantable cardioverter defibrillator (ICD) for secondary prevention of sudden cardiac arrest (SCA).

Method and results Over a 10 year period, 43 patients with an ICD after survived cardiac arrest (SCA) due to an aLQTS were included [female $n = 27$ (63%); mean age 61 $\pm$ 16 years]. There was no clinical evidence for congenital LQTS (Schwartz score 1.25 $\pm$ 0.8). Structural heart disease was present in 29 patients (47%; ischaemic $n = 13$; dilated cardiomyopathy $n = 9$; mean EF 41% $\pm$ 12). The most common proarrhythmic trigger happened to be antiarrhythmic drugs ($n = 34$; 79%). Other triggers included contrast agent ($n = 1$), haloperidol ($n = 2$), severe hypokalaemia ($n = 2$), drug abuse/alcohol ($n = 2$), and mere severe bradycardia ($n = 2$). Under trigger QTc interval measured 536 $\pm$ 58 vs. 438 $\pm$ 33 ms without trigger ($P < 0.001$). During a mean follow-up of 84 $\pm$ 55 months, appropriate shocks occurred in 19 patients (44%); inappropriate shocks in 13 patients (30%; only inappropriate $n = 3$). Appropriate shocks were almost as common in patients without as in those with structural heart disease (35 vs. 48%; $P = 0.32$). None of the patients were re-exposed to the initial trigger during the follow-up period. Beta-blocker medication did not prevent ICD shocks (12 of 19 vs. 11 of 24 on medication).

Conclusion Appropriate ICD shocks are a common finding in patients with aLQTS and SCA irrespective of the underlying cause or structural heart disease. Thus, even in the presence of relevant acquired proarrhythmia ICD may be beneficial.

Wearable cardioverter defibrillators for patients with long QT syndrome

Heidi J. Owen [a,b,1], J. Martijn Bos [b,c,d,1], Michael J. Ackerman [b,c,d,*]

[a] Department of Nursing, Mayo Clinic, Rochester, MN, USA
[b] Department of Pediatric and Adolescent Medicine, Division of Pediatric Cardiology, Mayo Clinic, Rochester, MN, USA
[c] Department of Molecular Pharmacology & Experimental Therapeutics, Windland Smith Rice Sudden Death Genomics Laboratory, Mayo Clinic, Rochester, MN, USA
[d] Department of Cardiovascular Medicine, Division of Heart Rhythm Services, Mayo Clinic, Rochester, MN, USA

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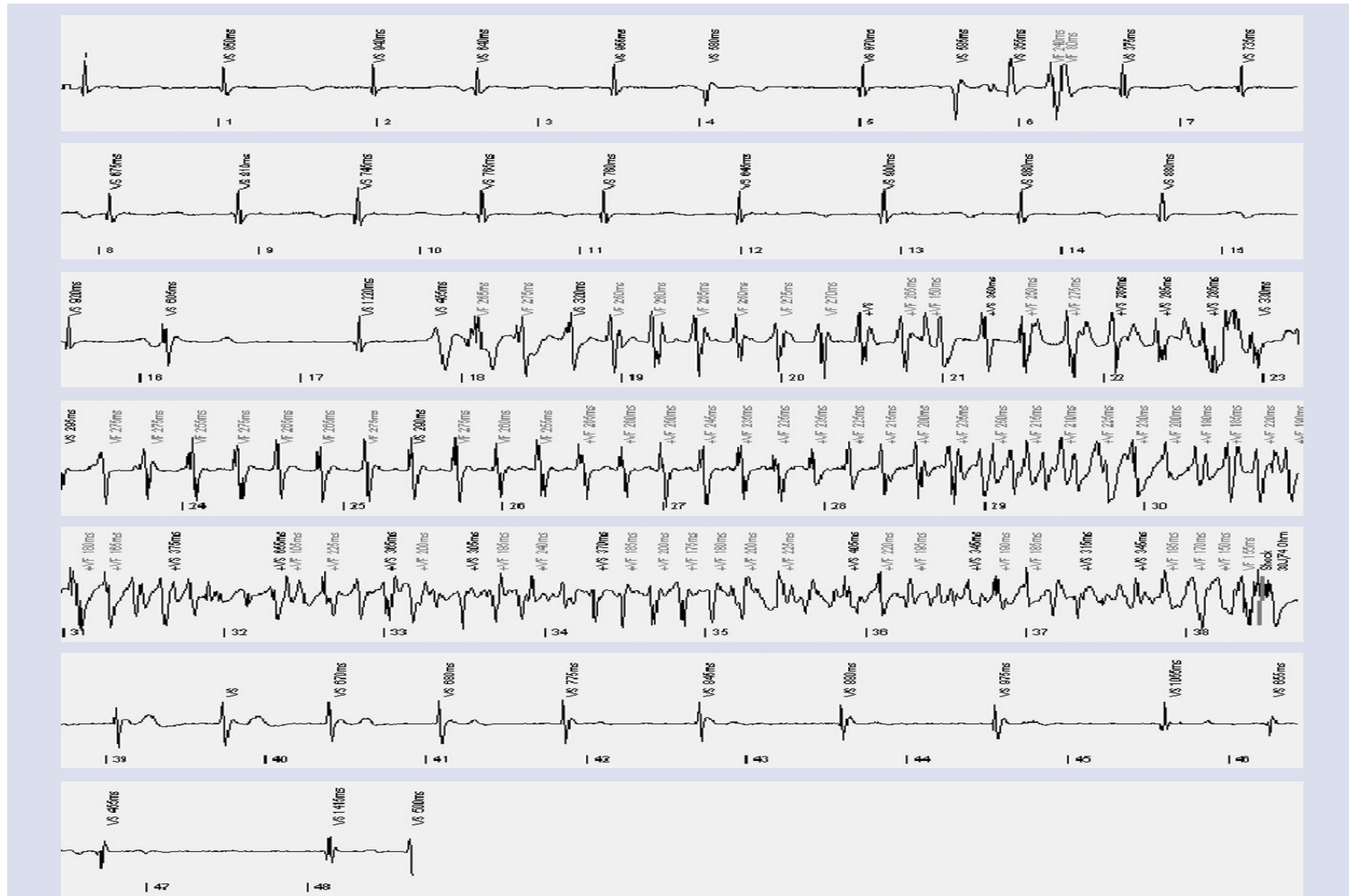
ABSTRACT

Background: Long QT syndrome (LQTS) is a potentially lethal cardiac channelopathy, but with the appropriate treatment strategy, such as beta-blockers, left cardiac sympathetic denervation (LCSD), and/or an implantable cardioverter defibrillator (ICD), most LQTS-triggered tragedies can be avoided. Since 2001, wearable cardioverter defibrillators (WCD:LifeVest™) have been available clinically.
Objective: Herein, we evaluated the use and outcome of WCDs in patients with LQTS.
Methods: We performed a retrospective review of 1027 patients with LQTS to identify patients who received a WCD, and collected pertinent clinical information regarding their LQTS diagnosis as well as indication and experience regarding use of the WCD.
Results: Overall, 10 LQTS patients (1%, 8 females, age at diagnosis 29 ± 18 years, mean QTc 488 ± 34 ms) were prescribed a WCD. Most common indication for WCD was as bridge to treatment during (temporary) situation of assessed high risk of sudden cardiac arrest (SCA; $n = 6$). The mean time of WCD use was 24 days (range 0 to 114 days). One patient (female, age 42, LQT2) received an appropriate VF-terminating shock 2 days after receiving her WCD. No inappropriate treatments or adverse events from wearing the WCD have occurred.
Conclusions: A WCD can be considered in patients with LQTS deemed to be at high risk for SCA while up-titrating beta blockers, considering ICD therapy, or when navigating short term periods of increased SCA-risk, like the post-partum period in LQT2 women, ICD revision or temporary inactivation, or during short term administration of known QT prolonging medications.


VS 950ms
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+VF 200ms
+VS 306ms
+VF 186ms
+VF 240ms
+VS 370ms
+VF 186ms
+VF 200ms
+VF 176ms
+VF 186ms
+VF 200ms
+VF 224ms
+VS 405ms
+VF 220ms
+VF 196ms
+VS 345ms
+VF 186ms
+VF 186ms
+VS 316ms
+VS 345ms
+VF 186ms
+VF 170ms
+VF 150ms
+VF 135ms
Shock
30J/74.0ohm
VS
VS 670ms
VS 888ms
VS 773ms
VS 846ms
VS 888ms
VS 976ms
VS 1065ms
VS 865ms
VS 462ms
VS 1141ms
VS 500ms

Bireysel Risk Değerlendirilmesi

- Kardiyak Arrest Sonrası Yaşayanlar– Yüksek Rekürrens Riski, β Bloker Kullansa Bile (5 yıl içinde rekürrens %14): ICD Tedavisini Destekler.
- Multiple Mutasyonlarda Risk Yüksek
- Senkop - Kardiyak Arrest Riski Artıyor.
- Sessiz Mutasyon Taşıyıcı – Orta Derecede Kardiyak Risk Taşırlar (10%) Doğum ile 40 yaş arası.
- β Bloker Kullanımı Önerilmektedir.

Proflaktif ICD Tedavisi Yüksek Riskli Hastalar

- Kadın Cinsiyet LQT2 + QTc ＞500 ms,
  - QTc 500 ms + Elektriksel İnstabilite
- Postpartum 9 ayda AKÖ riski artmıştır.
- Yüksek Riskli Genetik Panel (2 Mutasyon, Jervell ve Lange–Nielsen sendromu veya Timothy Sendromu)

- β Bloker Rekürren Uygun ICD Şoklarının Önlenmesinde Koruyucu Rolü Yoktur.
- β Bloker Tedavisi Altında Hastalarda Ardışık Şok uygulaması Açısından Yüksek Risk Altındadırlar.
- JLN veya TS Hastalarda Antiandranerjik Tedavi Kısmen Etkilidir. Bu hastalara Üçlü Tedavi Uygulanmalıdır (β Bloker +LCSD+ICD).
- Dual ICD (30/40 veya 2.5 sn tanıma zamanı, VF zone $\geq 220\ msn$ Ö)neriliyor.

Sabrınız için Teşekkür Ederim