

ONLINE FIRST

RESEARCH LETTER

Competitive Sports Participation in Athletes With Congenital Long QT Syndrome

To the Editor: Competitive sports participation for athletes with long QT syndrome (LQTS) is guided by the 36th Bethesda Conference, which recommends that patients with either (1) symptoms, (2) a corrected QT interval (QTc) greater than 470 milliseconds (males) or 480 milliseconds (females), or (3) an implantable cardioverter-defibrillator (ICD) not participate in most sports.¹ The European Society of Cardiology guidelines are more restrictive, disqualifying athletes from all sports based solely on a stringent QTc cutoff (>440 milliseconds in males, >460 milliseconds in females).² We sought to determine the outcomes of patients with LQTS who chose to remain athletes against guideline recommendations.

Methods. In this institutional review board–approved study (with waiver of consent), we reviewed records for patients with LQT1-3 genotypes, aged 6 to 40 years, who were first evaluated in the Mayo Clinic LQTS Clinic between July 2000 and November 2010. Records were reviewed for athletic participation after LQTS diagnosis and LQTS-related events during a mean (SD) follow-up of 5.1 (2.9) years. No patients were lost to follow-up. All were reevaluated or contacted by phone after July 1, 2011. A “competitive athlete” was defined as one participating in organized competitive sports at the little league, middle or high school, collegiate, or professional level.

The approach in the Mayo Clinic LQTS Clinic is to provide the athlete and their family with sufficient information to enable an informed decision regarding sports continuation. All patients received a comprehensive 2- to 3-day clinical and genetic evaluation, including a 1- to 2-hour consultation with an LQTS specialist (M.J.A.) and additional consultations as needed.³⁻⁵ Extensive counseling was provided to discuss individual prognosis and athletic participation guidelines. If a minor, the athlete and both parents had to agree to sports continuation. Tailored therapy included β -blockers, left cardiac sympathetic denervation, an ICD, or a combination. QT drug avoidance, electrolyte and hydration replenishment, and minimization of core body temperature elevations were advised. Each athlete obtained an automatic external defibrillator as part of the sports gear, and relevant school officials and coaches were informed.

Statistical analysis was performed using JMP version 8.0 (SAS Institute). A 2-tailed $P < .05$ was considered significant.

Results. Of 353 LQT1-3 patients (199 females; mean [SD] age, 17 [11] years; mean [SD] QTc, 472 [42] milliseconds), the majority (223, 63%) either were not

involved in sports (196, 88%) or chose to discontinue sports (27, 12%) following evaluation. Overall, 130 patients (37%, 60 females; age, 11 [7] years; QTc, 471 [46] milliseconds) remained in competitive athletics, including 20 with ICDs. There were no significant differences between the total cohort and athletes except that the nonathletes were older (TABLE). The 130 athletes competed in a variety of competitive sports (FIGURE),⁶ and 49 of 130 (38%) participated in more than 1 sport. There were 32 athletes (25%) competing in high school and 8 (6%) competing at the college, university, or professional level.

Seventy athletes (54%) were competing contrary to European guidelines but within Bethesda guidelines. None had a sport-related event. Of the 60 LQTS athletes (46%) continuing in sports contrary to both guidelines, only 1 experienced sporting-related events: a 9-year-old boy with LQT1, extreme QT prolongation (QTc >550 milliseconds), and a history of aborted cardiac arrest. He received 2 appropriate ventricular fibrillation-terminating ICD shocks, both while warming up before games. Each episode occurred in the setting of admitted β -blocker nonadherence.

The overall rate of events per athlete-year was 0.003 (1 event in 331 athlete-years; 95% CI, 1 in 92 to 1 in 2763 athlete-years).

Comment. Although many individuals with LQTS will choose to remain in competitive sports, athletes and their families are capable of self-disqualification. With more than 650 athlete-years of follow-up, we report a low rate of LQTS-triggered cardiac events during sports. Limitations include the small sample size, limited length of follow-up, and unknown generalizability.

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Study concept and design: Johnson, Ackerman.

Acquisition of data: Johnson.

Analysis and interpretation of data: Johnson, Ackerman.

Drafting of the manuscript: Johnson, Ackerman.

Critical revision of the manuscript for important intellectual content: Johnson, Ackerman.

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Study supervision: Ackerman.

Conflict of Interest Disclosures: Both authors have completed and submitted the ICMJE Form for Disclosure of Potential Conflicts of Interest. Dr Ackerman is a consultant for Biotronik, Boston Scientific, Medtronic, St Jude Medical, and Transgenomic. Intellectual property derived from Dr Ackerman's research program resulted in license agreements in 2004 between Mayo Clinic Health Solutions (formerly Mayo Medical Ventures) and PGxHealth (formerly Genaisance Pharmaceuticals and now Transgenomic). The Mayo Foundation for Medical Education and Research receives royalties from Transgenomic for the intellectual property used in its FAMILION LQTS genetic test. No other disclosures were reported.

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Table. Demographics of Competitive Athletes With Long QT Syndrome^a

	Total Cohort	Athletes	Within Bethesda but Contrary to ESC Guidelines	Contrary to Both Guidelines	P Value ^b
No. of patients	353	130	70	60	
Age at diagnosis, mean (SD), y	17 (11)	11 (7)	11 (7)	12 (6)	.91
Sex					.21
Male	154	70	41	29	
Female	199	60	29	31	
Mean (SD) QTc, ms	472 (42)	471 (46)	444 (23)	501 (46)	<.001
Symptoms, No. (%)	111 (31)	29 (22)	1 (1)	28 (47)	<.001
β-Blockers, No. (%)	280 (79)	112 (87)	55 (79)	57 (95)	<.008
ICD, No. (%)	78 (22)	20 (15)	0	20 (33)	<.001
Follow-up available, mean (SD), y	5.5 (3.4)	5.1 (2.9)	5.1 (2.9)	5.0 (3.0)	.98
Genotype, No. (%)					
LQT1	182 (52)	74 (57)	41 (59)	33 (55)	.65
LQT2	130 (37)	41 (32)	20 (29)	12 (35)	.47
LQT3	37 (10)	11 (8)	8 (11)	3 (5)	.20
Multiple ^c	4 (1)	4 (3)	1 (1)	3 (5)	.19

Abbreviations: ESC, European Society of Cardiology; ICD, implantable cardioverter-defibrillator; LQTS, long QT syndrome; QTc, corrected QT interval.

^aDemographics of the overall cohort of eligible LQT1-3 patients 6-40 years old (N=353), which includes the 130 LQTS athletes including those participating against ESC guidelines but within Bethesda guidelines (n=70) and those athletes participating against both guidelines (n=60).

^bComparing athletes who are participating within Bethesda Conference guidelines but against ESC guidelines vs those participating against both guidelines.

^cPatients with >1 LQTS-causing mutation.

Figure. Primary Sport Classification (According to the 2005 Bethesda Conference Guidelines⁶) for the Competitive Athletes With Long QT Syndrome

Static Component, Peak % Maximal Voluntary Left Ventricular Contraction	No. of Patients		
	A Low <40	B Moderate 40-70	C High >70
III High >50	25	4	0
II Moderate 20-50	0	14	34
I Low <20	3	22	28
	Dynamic Component, Peak % Maximal Oxygen Uptake		
	A Low <40	B Moderate 40-70	C High >70

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1. Zipes DP, Ackerman MJ, Estes NA III, Grant AO, Myerburg RJ, Van Hare G. Task Force 7: arrhythmias. *J Am Coll Cardiol.* 2005;45(8):1354-1363.
2. Pelliccia A, Fagard R, Bjørnstad HH, et al; Study Group of Sports Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology; Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology. Recommendations for competitive sports participation in athletes with cardiovascular disease: a consensus document from the Study Group of Sports Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology and the Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology. *Eur Heart J.* 2005;26(14):1422-1445.
3. Ackerman MJ. Cardiac channelopathies: it's in the genes. *Nat Med.* 2004;10(5):463-464.
4. Goldenberg I, Moss AJ, Peterson DR, et al. Risk factors for aborted cardiac arrest and sudden cardiac death in children with the congenital long-QT syndrome. *Circulation.* 2008;117(17):2184-2191.
5. Liu JF, Jons C, Moss AJ, et al; International Long QT Syndrome Registry. Risk factors for recurrent syncope and subsequent fatal or near-fatal events in children and adolescents with long QT syndrome. *J Am Coll Cardiol.* 2011;57(8):941-950.
6. Mitchell JH, Haskell W, Snell P, Van Camp SP. Task Force 8: classification of sports. *J Am Coll Cardiol.* 2005;45(8):1364-1367.

CORRECTION

Incorrect Wording in Patient Page: In the Patient Page entitled "Neonatal Hyperbilirubinemia," published in the May 16, 2012, issue of *JAMA* (2012;307[19]:2115), incorrect wording was used. The lighting in the Figure should be labeled "Special light source (blue-green spectrum)," and in the "Phototherapy" section under "Treatment," the words "ultraviolet (blue) lights" should read "a special light source (blue-green spectrum)." This article was corrected online.