JACC: CASE REPORTS VOL. 22, 2023

CASE REPORT

ADVANCED

CLINICAL CASE

Congenital Long QT Syndrome and Cardiac Arrest in a Military Tactical Athlete

Theodore F. Chung, MD, Andrea N. Keithler, DO, Alexandra J. Smith, MD, Kelvin N.V. Bush, MD

ABSTRACT

We present a case of a U.S. marine who experienced cardiac arrest during military Special Forces underwater diving exercises whose evaluation revealed congenital long QT syndrome. This case highlights the expanding role for systematic electrocardiogram screening in target athletic and military populations given their stress and tactical exposures. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2023;22:101990) © 2023 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 22-year-old, previously healthy, active-duty Marine was participating in routine Special Forces daily swim training when she experienced sudden lightheadedness and collapse. At the poolside, she immediately received cardiopulmonary resuscitation for 2 minutes and a single automated external defibrillation (AED) with successful return of spontaneous circulation. AED interrogation (**Figure 1**) revealed polymorphic ventricular tachycardia (VT). Electrocardiogram (ECG) upon hospital presentation demonstrated sinus rhythm with a QTc of 502 ms in the absence of any acquired causes for QT prolongation (**Figure 2**) consistent with congenital long QT syndrome (LQTS).

LEARNING OBJECTIVES

- To discuss the role of electrocardiogram screening in tactical athletic and military populations.
- To review the epidemiology of sudden cardiac death conditions in military tactical populations.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis for polymorphic VT etiologies include coronary ischemia, Purkinje-related polymorphic VT, torsades de pointes secondary to congenital or acquired long QT syndrome, pseudotorsades de pointes, electrolyte abnormalities such as hypomagnesemia or hypokalemia, Brugada syndrome, short QT syndrome, and catecholaminergic polymorphic VT. Mimickers of polymorphic VT include pre-excited atrial fibrillation, other supraventricular tachycardias with aberrancy, and electrocardiogram motion artifacts.

INVESTIGATIONS

AED interrogation revealed polymorphic VT consistent with torsades de pointes in the setting of prolonged QT interval and exercise history. Family

From the Division of Cardiology, San Antonio Military Medical Center, Fort Sam Houston, Texas, USA.
The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received April 18, 2023; revised manuscript received May 30, 2023, accepted June 27, 2023.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2023.101990

ABBREVIATIONS AND ACRONYMS

AED = automated external defibrillation

LQTS = long QT syndrome

SCD = sudden cardiac death

VT = ventricular tachycardia

history was notable for a half-sister who experienced unexplained cardiac arrest in her 30s, early sudden deaths in 2 maternal relatives, and a paternal aunt with an LQTS diagnosis (Figure 3). There were no electrolyte abnormalities, and urine drug screening results were negative upon hospital presentation. Transthoracic echocardiogram and cardiac magnetic resonance demonstrated a structurally normal heart without evidence of late gadolinium enhancement. Treadmill exercise testing revealed a pathologic QTc increase to 560 ms during the fourth minute of recovery (Figure 4) without evidence of exercise-induced ischemia or ectopy. Genetic testing and counseling identified a potassium voltage-gated channel subfamily Q member 1 *(KCNQ1)* heterozygous pathogenic variant with autosomal dominant inheritance.

MANAGEMENT

The servicemember was diagnosed with congenital long QT syndrome. She was initiated on beta-blocker therapy and provided exercise restrictions, which immediately disqualified her from continued Special Forces activities and training. She received a secondary prevention implantable cardioverter-defibrillator during the index hospitalization.

DISCUSSION

The history of combat diving started in World War II based on developing needs to obtain tactical access to strategic locations and conduct unconventional clandestine warfare. Rigorous selection and training programs were subsequently developed to produce mission-capable divers for amphibious reconnaissance and water-borne operations.[1]

There is an increasing role for using electrocardiogram screening to identify tactical athletes and military service members with potentially life-threatening channelopathies and cardiomyopathies before enrollment in strenuous training programs. In a cohort of 4,095 U.S. Naval Academy midshipmen, 16 of 19 cardiac abnormalities with potential sudden cardiac death (SCD) vulnerability were identified with electrocardiogram screening in addition to the standard American Heart Association-recommended approach (questionnaire and physical examination) to cardiovascular screening.[2] We propose that a screening electrocardiogram would have identified this patient's LQTS diagnosis and potentially led to exercise restrictions and occupational career guidance changes.

Universal electrocardiogram screening currently is not a component of the medical evaluation of military applicants.[3] Presently, there is a paucity of cost-effectiveness data and available resources needed to mandate continuous annual screening programs with the prevalence of sudden death in young athletes remaining low. Additionally, there is a shortage of military physicians and cardiologists available to read screening electrocardiograms. Currently, there are efforts to initiate artificial intelligence algorithms to screen electrocardiograms for the military population. The incidence of borderline QTc prolongation detected by military screening programs in unclear at this time, highlighting the role for development of further risk stratification tools.

According to the Defense Medical Epidemiology Database, the incidence of exertion-related SCD in U.S. military service members is approximately 1.63 per 100,000 person-years and at least 3.84 per 100,000 person-years for those aged 35 years and older.[4] Though the overall incidence is low, it remains the leading cause of nontraumatic death in the U.S. military population. The incidence of SCD in military recruits is comparable to or greater than the rates observed collectively in National Collegiate Athletic Association athletes.[5] The most common causes of sudden death in young athletes in U.S.

FIGURE 1 Automated External Defibrillation Interrogation Demonstrating Polymorphic Ventricular Tachycardia

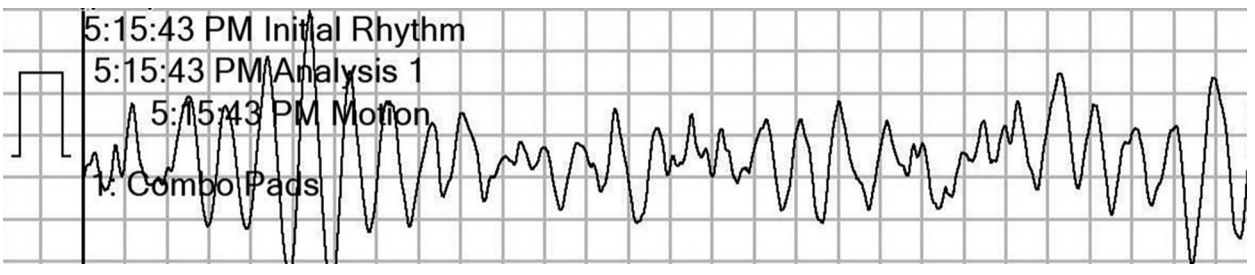
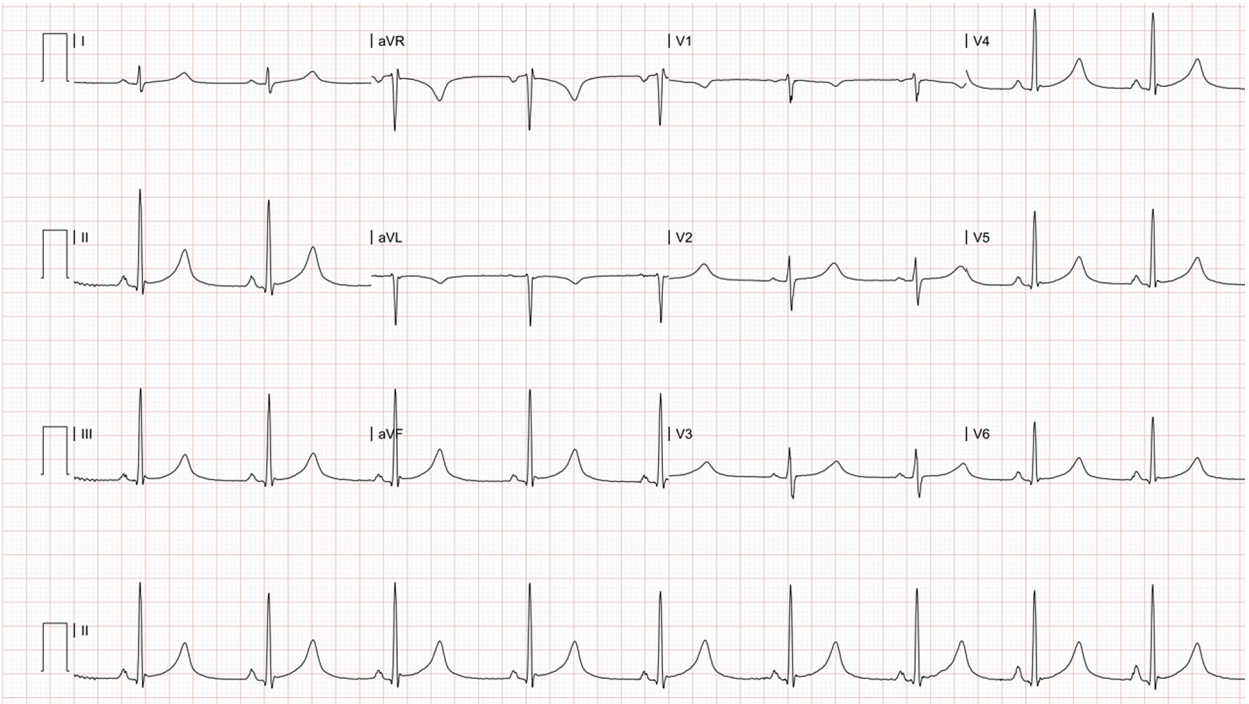
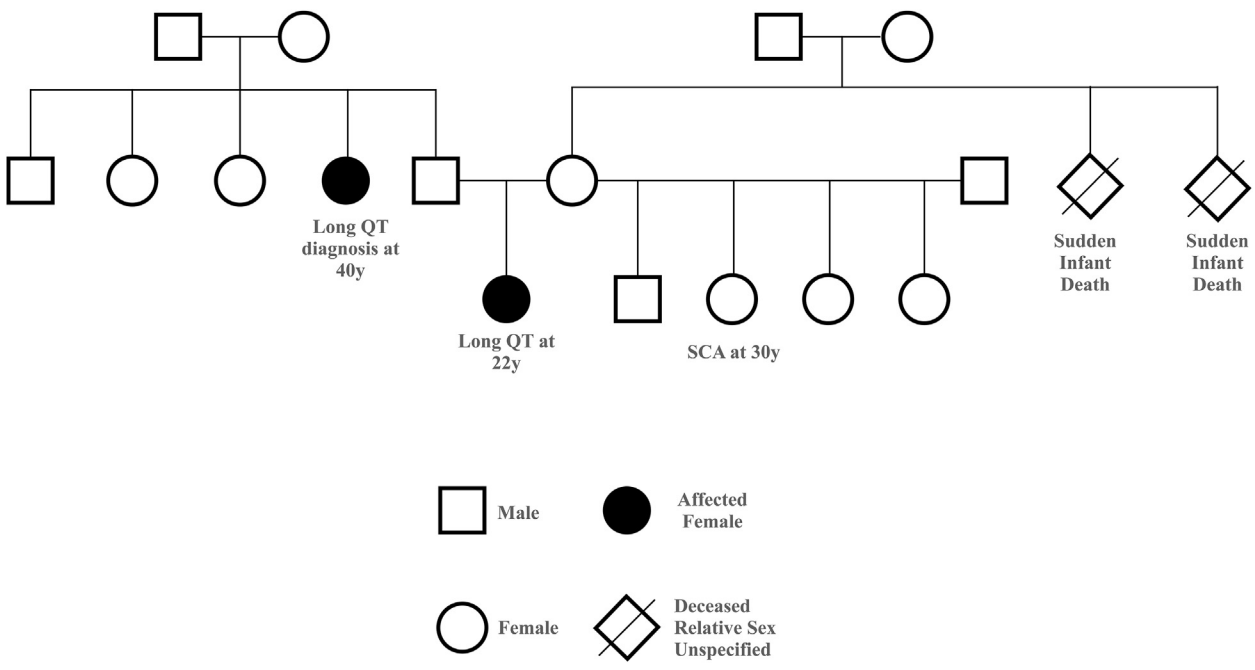


FIGURE 2 **Electrocardiogram**



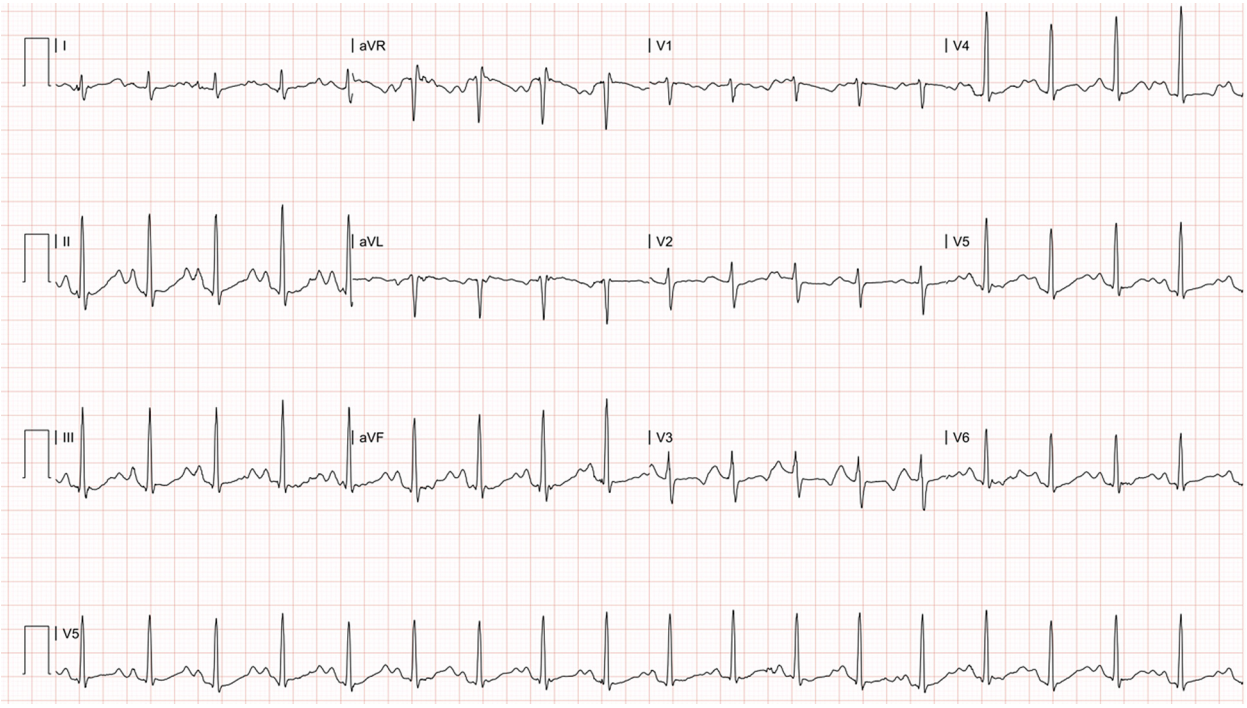
Baseline electrocardiogram demonstrating a QTc of 502 ms with broad and symmetric T waves.

FIGURE 3 **Family Pedigree**



The patient's family history was notable for a maternal half-sister who experienced unexplained cardiac arrest in her 30s, early sudden deaths in 2 maternal relatives, and a paternal aunt with long QT syndrome. SCA = sudden cardiac arrest.

FIGURE 4 Exercise Electrocardiogram



Treadmill exercise testing demonstrating a pathologic QTc increase to 560 ms during the fourth minute of recovery without evidence of ventricular arrhythmias or ischemia.

athletes under the age of 35 years are hypertrophic cardiomyopathy, followed by anomalous coronary arteries. Inherited channelopathies constitute 3% of SCDs.[5]

Congenital long QT syndrome is a heritable group of channelopathies characterized by cardiac repolarization abnormalities that persistently or intermittently prolong the QTc on electrocardiogram and increase the susceptibility for cardiac events, including arrhythmogenic syncope, torsades de pointes, and SCD. The 3 major autosomal dominant LQTS genotypes are LQT1, LQT2, and LQT3, with LQT1 resulting from a loss-of-function allelic variant in *KCNQ1*, which encodes the adrenergic-sensitive I_{ks} channel.[6] The prevalence has been estimated to be 1:2,000, or 0.05%, with a slight female predominance.[7] The symmetric and broad-based T-wave, long QTc, and clinical history of this patient were consistent with LQT1.

In this cohort of channelopathies, swimming is an absolute contraindication given the heightened drowning risk. In a study of 28 victims of swimming-related drowning, 8 (28.6%) had allelic variants, including 2 with *KCNQ1* sequence alterations and 6 with *RYR2* mutations. Nearly 30% of unexplained drowning deaths referred for postmortem genetic analysis carried a cardiac ion channel mutation. Of the 28 individuals who drowned while swimming, women were more likely to be mutation positive.[8]

FOLLOW-UP

Beta-blocker medical therapy was titrated to nadolol 80 mg daily. Cascade screening was recommended on all first-degree relatives given that the proband has an identified gene variant. There have been no recurrent ventricular tachyarrhythmias or syncope.

CONCLUSIONS

This case of congenital LQTS in an active-duty service member who experienced cardiac arrest during an intensive swimming military training exercise highlights unique stressors and risks to the tactical athletic and military populations. Screening electrocardiograms are warranted for military personnel and other tactical athletes to identify

those at risk for SCD who will engage in high-risk activities.

FUNDING SUPPORT AND AUTHOR DISCLOSURE

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Kelvin N. V. Bush, Division of Cardiology, San Antonio Military Medical Center, 3551 Roger Brook Drive, Fort Sam Houston, Texas 78234, USA. E-mail: Kelvin.n.bush.mil@health.mil.

REFERENCES

1. Grollimund T. Military diving—a history of innovation. Divers Alert Network. Published 2014. Accessed March 29, 2023. https://dan.org/alert-diver/article/military-diving/

2. Murphy C, Saperstein A, Klusewith S, et al. Effective cardiovascular screening of military recruits requires an electrocardiogram. *J Am Coll Cardiol*. 2022;79(Suppl 9):1638.

3. Magee C, Haigney MC. Cardiovascular screening in the U.S. military: time to reconsider the electrocardiogram. *Mil Med*. 2020;185:7-8.

4. Smallman DP, Webber BJ, Mazuchowski EL, Scher AI, Jones SO, Cantrell JA. Sudden cardiac death associated with physical exertion in the US military. *Br J Sports Med*. 2016;50:118-123.

5. Maron BJ, Thompson PD, Ackerman MJ, et al. Recommendations and considerations related to preparticipation screening for cardiovascular abnormalities in competitive athletes: 2007 update. *Circulation*. 2007;115:1643-1655.

6. Krahn AD, Laksman Z, Sy R, et al. Congenital long QT syndrome. *J Am Coll Cardiol EP*. 2022;8:687-706.

7. Schwartz PJ, Stramba-Badiale M, Crotti L, et al. Prevalence of the congenital long-QT syndrome. *Circulation*. 2009;120(18):1761-1767.

8. Tester DJ, Medeiros-Domingo A, Will ML, Ackerman MJ. Unexplained drownings and the cardiac channelopathies: a molecular autopsy series. *Mayo Clin Proc*. 2011;86(10):941-947.

KEY WORDS **cardiac arrest, congenital long QT syndrome, electrocardiogram screening, sudden cardiac death, tactical athlete**