

A Case Report of "Borderline" Long QT in Asymptomatic Athlete. What Should We Do?

Massimo Bolognesi*

Center for Sports Cardiology, Ausl della Romagna, District of Cesena, Cesena, Italy

Case Study

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*For Correspondence

Massimo Bolognesi, Center for Sports Cardiology, Ausl della Romagna, District of Cesena, Cesena, Italy, Tel: +390547645074.

E-mail: massbolo1@tin.it

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ABSTRACT

Long QT Syndrome (LQTS) is a potentially lethal cardiac channelopathy that can be suspected by the presence of palpitations, syncope and seizures. Because of increased public and physician awareness of warning signs suggestive of LQTS, there is a potential for the condition to be overdiagnosed. The 12-lead electrocardiogram (ECG) remains the universal initial diagnostic test in the evaluation of LQTS, however, it is subject to miscalculation and misinterpretation. Herewith the author describes an anecdotal case report of a 29 year old male athlete with borderline QTc interval who was disqualified from competitive sports.

INTRODUCTION

Sudden cardiac death in young athletes is a rare event, but it always raises serious concerns, especially because most victims don't have warning symptoms. Sports pre-participation screening aims to identify young athletes at risk, but there is not yet a general consensus regarding the best way to accomplish this ^[1]. The cardiac channelopathies are a collection of primary, genetically mediated, electrical disorders that are generally associated with a structurally normal heart and a propensity for syncope, seizures or sudden cardiac arrest precipitated by episodes of dangerous arrhythmias. The most common channelopathy is Long-QT Syndrome (LQTS), involving an estimated 1 in 2,000 people ^[2]. Restriction from virtually all competitive sports has been the guideline-based recommendation since 2005 for athletes with a cardiac channelopathy ^[3,4]. The lack of compelling evidence for restricting concealed LQTS patients induced the 26th Bethesda Conference recommendations, revised in 2005, to allow genotype positive-phenotype negative LQTS individuals to participate in competitive sports, except those in LQTS type 1 who remain prohibited from swimming participation. On the other hand, for European athletes with LQTS, sports participation is still impossible irrespective of their genotype/phenotype relationship or clinical history ^[5].

CASE DESCRIPTION

A 29 year old athlete of athletics (110 m hurdles) came to our center for a second opinion on sports preparticipation screening, although he has been disqualified in previous years by some sports physicians because of suspected Long QT Syndrome. He declared that for over 15 years he has been a competitive athlete (in athletics with at least 5-6 workouts per week over 2 h per day) and he has always been fit. Furthermore, he did not have a family history of sudden death. Also, he never had any symptoms during physical exertion, such as syncope or palpitations. Despite that, the evidence of a supposed abnormal electrocardiogram (ECG) with a QTc (calculated by Bazett's formula: $QTc=QT/RR^{1/2}$). Interval over 440 ms and below 460 ms (i.e., QT interval borderline) (**Figures 1 and 2**) has prompted doctors to require further clinical and genetic testing.



Figure 1. Resting ECG shows sinus rhythm at 56 bpm heart rate; normal repolarization. QT 450 ms - RR 990 ms - QTc 452 ms; U wave.

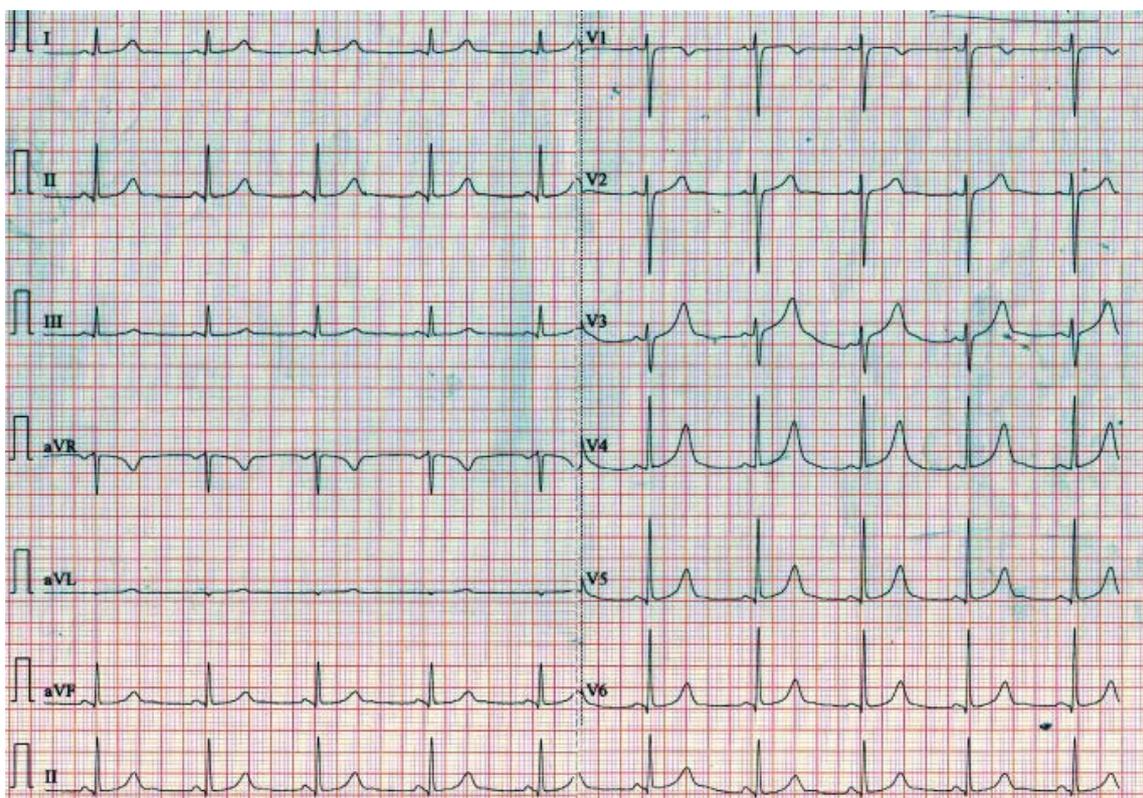


Figure 2. Resting ECG shows sinus rhythm at 60 bpm heart rate; normal repolarization. QT 445 ms - RR 990 ms - QTc 447 ms; slight U wave and some artifacts on leads V3-V4.

The genetic investigation was positive for the M1V variant of the KCNQ1 gene, which however was identified as a variant of unknown significance. Furthermore, an ECG-Holter reported an average prolongation of the QTc interval approximately to 469 ms, as reported by Center of Cardiovascular Genetics in Pavia. Therefore, interruption of physical activity was recommended and low dose Beta-blocker (Nadolol 20 mg/day) was prescribed. No alteration of blood tests was present and no drug was administered. At this time, the clinical examinations performed at our center showed normal ECG tracing patterns with QTc interval all within the normal range, either at rest or during the treadmill maximal test (**Figures 3 and 4**).

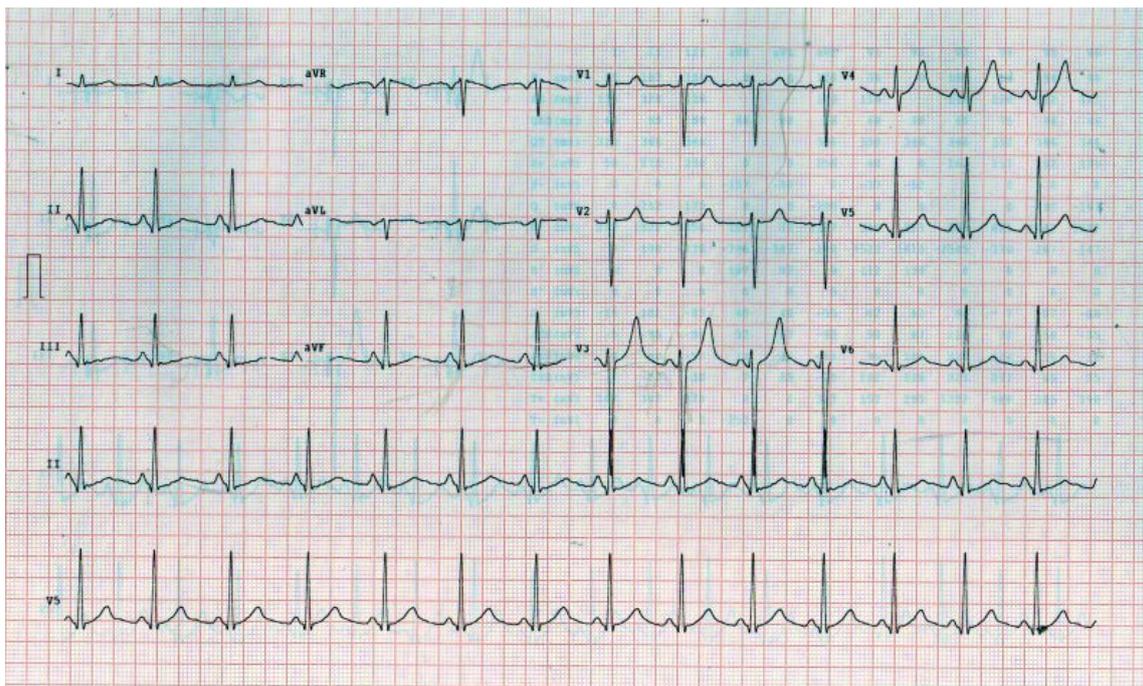


Figure 3. Resting ECG shows sinus rhythm at 82 bpm; normal repolarization; QT 360 ms - RR 680 ms - QTc 437 ms; U wave.

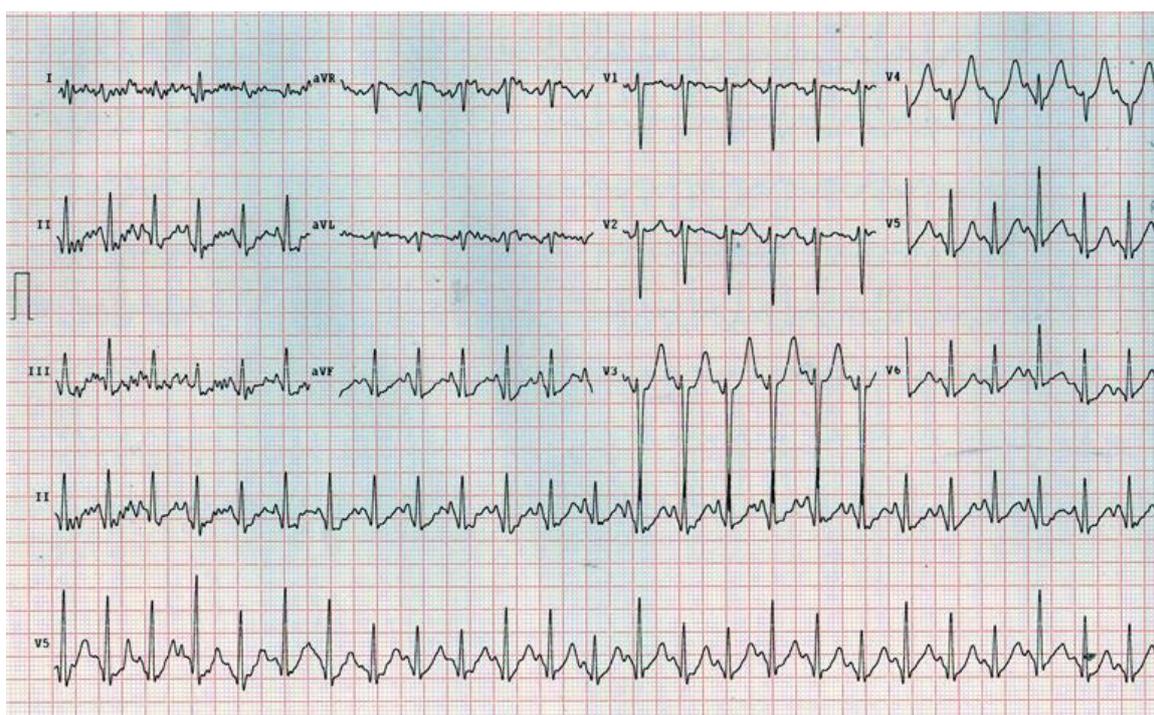


Figure 4. ECG at peak exercise shows sinus tachycardia; normal repolarization; QT 280 ms - RR 400 ms - QTc 443 ms.

Also an echocardiogram was normal. Therefore, considering the lack of evidence of a significant prolongation of QTc interval both at rest and during exercise stress testing, the absence of a family history of sudden death and especially the absence of any symptoms, it was considered that there is no real life-threatening risk of underlying arrhythmias during competitive sports activity. So, the athlete was considered healthy and fit for competitive sports, without any limitation.

DISCUSSION

A recent report sheds light on this controversial topic by describing a new screening protocol that offers advantages over American Heart Association (AHA) recommendations and shows that the ECG is the best single screening method [6]. The ECG is more sensitive in detecting heart muscle problems (cardiomyopathies) and potentially life-threatening electrical disorders such as Wolff-Parkinson-White and Long QT Syndrome [7]. The prevalence of QT interval prolongation in asymptomatic athletes was

recently reported as 0.4%, as well as 1 in 286 patients^[8]. Only four patients in this study with QTc values between 460 and 500 ms were studied further, but not disqualified from their sport because of lack family histories and negative provocative testing. An interesting publication on the Italian pre-participation screening program reported disqualification of 2% of all athletes on the basis of screening, with only five athletes out of 42,386 ultimately diagnosed with LQTS^[9]. The majority of disqualifications in this valuable Italian work were for cardiomyopathy, hypertension, valvular disease and not-LQTS rhythm abnormalities^[9].

In each of these studies, the European guidelines cut-off values for diagnosing prolonged QT interval were used: 440 ms for males and 460 ms for females^[10]. The 36th Bethesda Conference guidelines were first published in 2005 and established recommendations for athletic activity restrictions in patients with LQTS. As reported, a competitive sports disqualification was adopted for any patient with a history of out-of-hospital cardiac arrest or LQTS-related syncopal episode, regardless of underlying genotype or QTc. For asymptomatic patients with a QTc of >470 ms in males or >480 ms in females the recommended limitation was to only class IA sports, with further liberalization possible if the asymptomatic patient had the LQT3 genotype. A good revisitation of the competitive sports disqualifications was deemed reasonable for asymptomatic genotype-positive LQTS patients with QTc interval values in the overlapping/borderline range, except for competitive swimming in athletes with concealed LQT1. The ability to differentiate findings suggestive of a potentially lethal cardiovascular disorder from benign physiological adaptations occurring as the result of regular, intense training (i.e., athlete's heart), is mandatory for sports cardiologists. Nowadays, several reports have outlined ECG criteria finalized to distinguish normal ECG findings in athletes from ECG abnormalities requiring additional evaluation^[11]. As evident in the growing literature on the psychological consequences of physical inactivity and its huge impact on the patient's quality of life, it appears that restricting LQTS patients unnecessarily from sports participation is not without a price^[12]. New research suggests that up to 40% of patients diagnosed with long-QT syndrome (LQTS) don't actually have it and are receiving treatment unnecessarily^[13]. In this case, the author note that the decision to disqualify the athlete based on the QTc being "borderline" and for the presence of a genetic variant of unknown significance is wrong. First of all because it is contrary to the International recommendations and shared guidelines, but above all because there is clearly no phenotype that could identify this athlete as being at risk of LQTS1^[14].

KEY POINTS

I) Firstly, careful reading of the ECG is crucial in the settings of sports medicine and sports cardiology. Most of the errors stemmed from miscalculation of the QTc interval, misunderstanding of the normal distribution of QTc values, and/or misreading of symptoms. For example, the majority of the mistakes were due to overestimation of the QTc interval, which can result from the erroneous inclusion of the U-wave in the calculation of the interval.

II) Secondly, LQTS can be diagnosed based on the specific shape of T waves produced during a quick standing test. The T-wave shape was most likely to help identify patients with LQT2 and least likely to help identify patients with LQT3.

III) The third point is that genetic analysis of unclear significance shouldn't have any relevance in making decision for sports eligibility. So, all QTc measurements should be interpreted in the context of the patient's personal and family history.

IV) The fourth point discusses the Italian law on eligibility for competitive sports that is certainly too restrictive. Anyway, more clear indication must be done in QTc calculation and more clinical recommendations should be given, because the risk to penalize athletes is too high.

V) Finally, the first problem of pre-participation screening consists in the unacceptably high false-positive rate and the costs associated with screening large numbers of athletes.

CONCLUSION

Sports physicians should know that false positive results raise concerns about the unnecessary investigations, erroneous disqualification and psychological harm to the athlete. Conversely, Sudden Cardiac Deaths in sport are highly visible, claiming young lives. Clinicians should recognize that medical eligibility versus disqualification decisions have become increasingly complex. Indeed, these decisions may be fraught with potential legal liability risks. It is unwise to be influenced by the desires of athletes (with a high potential cardiovascular risk) but it is equally important to not penalize athletes with a poor diagnosis. In conclusion, sports physicians must be able to make a correct ECG interpretation.

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