[Primary Care]

Evaluation and Management of Athletes With Long QT Syndrome: An Evolved Paradigm

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Context: The congenital long QT syndrome (LQTS) is an inherited channelopathy known for its electrocardiographic manifestations of QT prolongation and its hallmark arrhythmia, torsades de pointes (TdP). TdP can lead to syncope or sudden death and is often precipitated by triggers such as physical exertion or emotional stress. Given that athletes may be at particular risk for adverse outcomes, those suspected of having LQTS should be evaluated, risk stratified, treated, and receive appropriate counseling by providers with sufficient expertise according to the latest guidelines.

Evidence Acquisition: The following keywords were used to query MEDLINE and PubMed through 2016: *LQTS, LQT1, LQT2, LQT3, long QT, long QTc, prolonged QT, prolonged QTc, QT interval, QTc interval, channelopathy, channelopathies, athletes, torsades de pointes*, and *sudden cardiac death*. Selected articles within this primary search, in addition to relevant references from those articles, were reviewed for relevant information and data. Articles with pertinent information regarding pathophysiology, evaluation, diagnosis, genetic testing, treatment, and guidelines for athletes were included, particularly those published in the prior 2 decades.

Study Design: Clinical review.

Level of Evidence: Level 3.

Results: Diagnosis of LQTS involves eliciting the patient's family history, clinical history, and evaluation of electrocardiographic findings. Genetic testing for common mutations can confirm suspected cases. β-Blockers represent the mainstay of treatment, though interventions such as implantable cardioverter-defibrillator placement or left cardiac sympathetic denervation may be required. Properly evaluated and treated athletes with LQTS have a low risk of cardiac events.

Conclusion: Detection and management of LQTS in the athletic population is crucial given the possibility of adverse outcomes with the stress of athletic participation. Preparticipation screening examinations should include a thorough clinical and family history. Screening electrocardiograms may display key findings consistent with LQTS while genetic testing can confirm the diagnosis. Formerly considered a strict contraindication to athletic participation, LQTS is now an increasingly manageable entity with proper evaluation and treatment by qualified and experienced providers.

Keywords: long QT syndrome; athletic participation; arrhythmia; sudden cardiac death

Anton Jervell and Fred Lange-Nielsen[11] first described an association between congenital deafness, syncope, and sudden death in connection with a prolonged QT interval among siblings in 1957. Soon thereafter, Cesarino Romano[24] and Owen Conor Ward[40] independently described similar findings among patients without congenital deafness in 1963 and 1964. These hereditary forms of QT prolongation have been more clearly delineated since their initial identification, and mutations in at least 13 different genes are now known to be associated with long QT syndrome (LQTS). This review will

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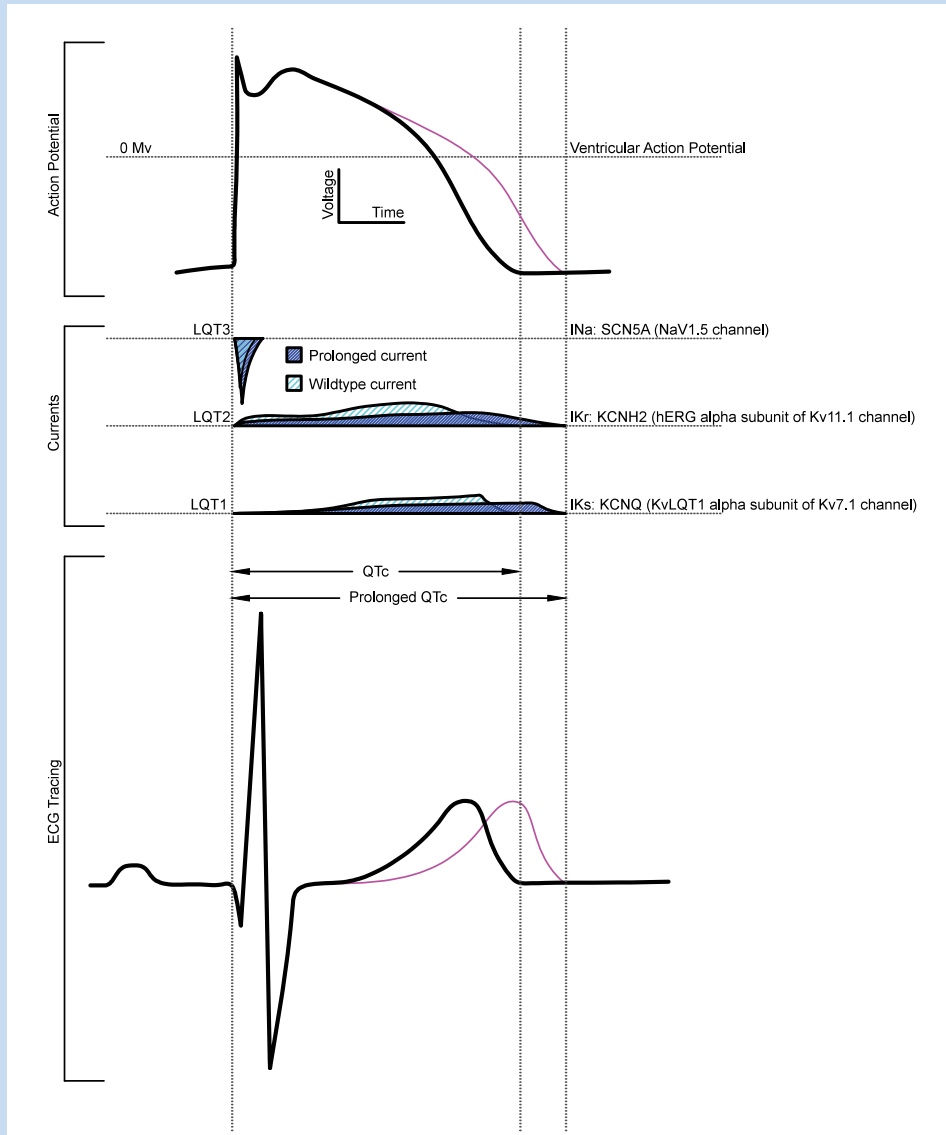The authors report no potential conflicts of interest in the development and publication of this article.

Figure 1. Action potential, currents, and electrocardiogram (ECG) findings. The figure outlines, conceptually, the connection between action potential prolongation, ECG findings, and the associated currents in LQT1-3.

focus on the 3 most prevalent variants of LQTS, namely the autosomal-dominant LQT1-3 syndromes, as they compose 75% to 80% of clinically definite LQTS.[2,33,37] LQT1, LQT2, and LQT3 compose approximately 30% to 35%, 25% to 40%, and 5% to 10% of disease caused by LQTS, respectively.[2] Schwartz et al[30] have estimated the prevalence of LQTS to be approximately 1:2500. Prevalence of a prolonged QT interval in elite athletes is 1:250 or 0.4%, though this does not necessarily imply these athletes have LQTS.[4]

Acquired causes of QT prolongation, often related to medications, electrolyte abnormalities, or other disease processes, have been described, and their characterization is beyond the focus of this review. Instead, this review describes the pathophysiology, clinical findings, electrocardiographic findings, diagnosis, genetic testing, management, and the latest athlete-specific guidelines related to LQTS.

PATHOPHYSIOLOGY

QT prolongation generally occurs when there is a delay between the onset of depolarization and the completion of repolarization, thus increasing action potential duration (Figure 1).[34] Ventricular depolarization occurs largely as a result of the I_{Na} current.[34] In contrast, ventricular repolarization occurs with the activation of the rapid and slow inward delayed rectifier potassium currents, I_{Kr} and I_{Ks}, respectively.[33,34]

The *KCNQ1* gene on chromosome 11 (locus 11p15.5) encodes the alpha subunits (KvLQT1) of the voltage-gated potassium
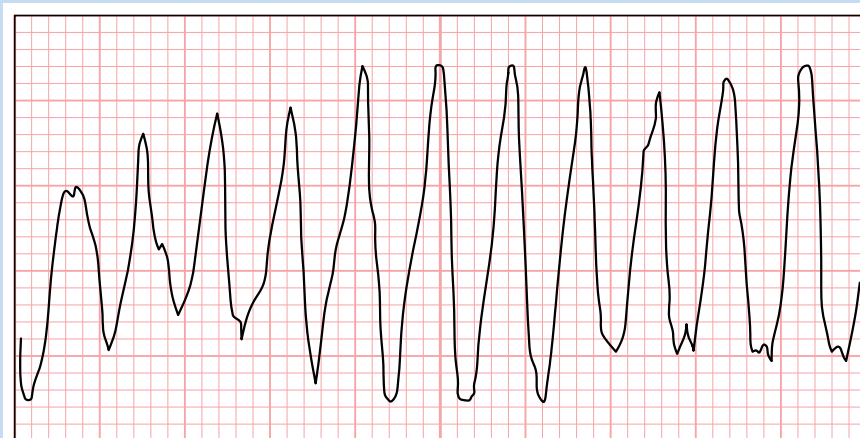
Figure 2. Torsades de pointes (TdP): A rapid polymorphic ventricular tachycardia characterized by varying amplitudes that seem to twist around a central axis. TdP may degenerate into ventricular fibrillation.

channel (Kv7.1) responsible for the slow component of the repolarizing delayed rectifier current, I_{Ks}.[2] A mutated *KCNQ1* gene causing a reduction (loss of function) in the I_{Ks} current results in LQT1 by delaying repolarization, thereby prolonging the action potential.[18,25,31,33] The *KCNH2* gene on chromosome 7 (locus 7q35-q36) encodes the alpha subunits (hERG) of the voltage-gated potassium channel (Kv11.1) that mediates the rapidly activating component of the repolarizing delayed rectifier current, I_{Kr}.[2] Similar to LQT1, a mutated *KCNH2* gene causes a reduction (loss of function) in the I_{Kr} current, resulting in LQT2 by delaying repolarization and prolonging the action potential.[18,25,31,33] The *SCN5A* gene on chromosome 3 (locus 3p21) encodes the voltage-gated cardiac sodium channel (NaV1.5) responsible for the depolarizing sodium current, I_{Na}.[2] In contrast to LQT1 and LQT2, a gain of function mutation in the *SCN5A* gene causing increased I_{Na} current results in LQT3 by prolonging the plateau phase of the action potential.[18,25,33] Action potential prolongation and decreases in repolarizing currents cause cytosolic Ca^{2+} overload, which facilitates early after-depolarizations and triggering of torsades de pointes (TdP).[18,31,33,34]

CLINICAL ASPECTS OF LQTS

The cardinal manifestations of LQTS are its clinical findings (palpitations, presyncope, syncope, or cardiac arrest) and electrocardiographic abnormalities. The classic clinical history involving repeated episodes of loss of consciousness under emotional or physical stress in a subject with a prolonged QT interval should raise suspicion for nonsustained TdP (Figure 2). If TdP is sustained, it may lead to ventricular fibrillation and sudden death.[25] Among symptomatic index cases, the untreated 10-year mortality is approximately 50%.[2] Asymptomatic patients, or silent mutation carriers, are relatively common, varying between 10% and 36% in one cohort among LQT1-3 patients.[21]

Specific triggers cause symptoms in patients with LQTS.[28] LQT1 patients are susceptible to cardiac events with physical exertion (62%) and emotional stress (26%); those affected by LQT1 are especially prone to events while swimming.[7,28] LQT2 patients suffered events related to emotional triggers most often (43%), followed by events occurring during sleep without arousal (29%); auditory events, such as alarm clocks, are also a trigger in those with LQT2.[28] LQT3 patients were most susceptible to events that occurred during sleep without arousal (39%).[28]

ELECTROCARDIOGRAPHIC ASPECTS OF LQTS

Beyond a prolonged QT and TdP, other ECG findings may be recognized in patients with LQTS, including specific T-wave morphologies associated with each LQTS type and T-wave alternans. The majority of LQTS index cases present with QT prolongation on their resting 12-lead electrocardiogram (ECG), while approximately 10% to 40% have nondiagnostic QT intervals at rest.[2]

Diagnosis of LQTS requires proper identification of a prolonged QTc. The ECG machine or computer-generated QTc should be confirmed using Bazett's formula[10] (QTc = QT / $\sqrt{RR}$; QT in milliseconds, RR in seconds) as some ECG tracings may not provide a clear substrate for accurate calculation by the machine or computer (Figure 3).[5,25] Bazett's correction decreases in accuracy with extremes in heart rate, undercorrecting for slower heart rates and overcorrecting for higher heart rates.[8] Since many athletes may be bradycardic at rest, a repeat ECG should be obtained after mild aerobic activity to ensure the heart rate is >50 beats/min. For sinus arrhythmia, manually confirm the QTc by averaging the QT interval and RR interval across the 10-second rhythm strip, best assessed with lead II or V5; leads II and V5 often provide the clearest delineation of the T-wave. If a

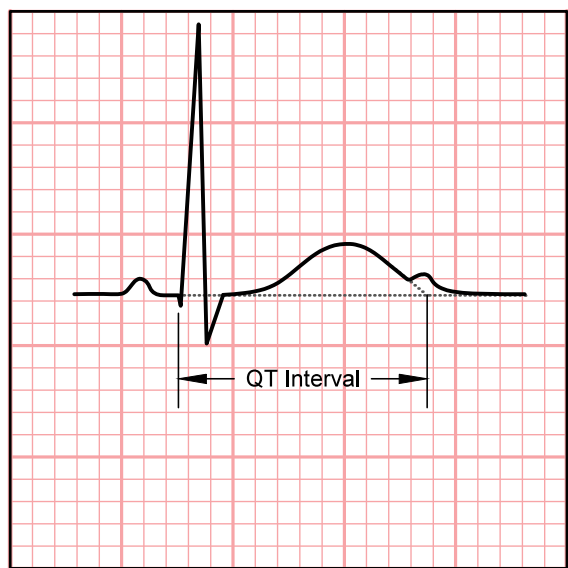Figure 3. Using Bazett's correction with the above QT (490 ms) and RR (0.91 s), the patient's QTc is 514 ms.

Figure 4. The "Teach-the-Tangent" method or "Avoid-the-Tail" method of measuring the QT interval in the presence of a U-wave.

Table 1. Formulas to calculate the QTc[a]

Name	Formula
Bazett	$QTc = QT / \sqrt{RR}$
Fredericia	$QTc = QT / \sqrt[3]{RR}$
Framingham	$QTc = 0.154 (1000 - RR)$
Hodges	$QTc = QT + 105 (1 / RR - 1)$

[a]Formulas published by Luo et al.[15]

U-wave is present, use the "Teach-the-Tangent" or "Avoid-the-Tail" method, which will provide a more accurate estimation of the QTc (Figure 4). A QTc of 470 ms in men or 480 ms in women should prompt further evaluation for the presence of LQTS as these are the often cited cut-offs for the 99th percentile.[10] High-risk patients are those with a QTc >500 ms.[22] Although Bazett's correction is commonly used, other formulas (Hodges, Framingham, Fredericia) to calculate the QTc exist that may provide equal, if not increased, accuracy (Table 1).[15]

A variety of T-wave morphologies are associated with the most common long QT syndromes (Figure 5). The T-wave should be inspected as its appearance may increase suspicion about the presence of LQTS.[2,10,12] LQT1 has been associated with smooth, broad-based T-waves while LQT2 is associated with notched or bifid T-waves. Notched T-waves occur more commonly in symptomatic patients and likely reflect the presence of subthreshold early after depolarizations.[25] LQT3 is often associated with a prolonged isoelectric ST segment with a late onset, asymmetrical T-wave, or biphasic T-waves.[25,42]

T-wave alternans (Figure 6), characterized by beat-to-beat variability in polarity or amplitude of the T-wave, may be present at rest for brief moments but usually appears during emotional or physical stress or with QT-prolonging medications.[41] T-wave alternans often precedes TdP and indicates electrical instability assisting in the identification of high-risk patients who may require review and reassessment of therapy.[22,25]

DIAGNOSIS

Clinical suspicion of LQTS is based on electrocardiographic findings, clinical history, and family history. The 1993 to 2011 LQTS diagnostic criteria, or the "Schwartz criteria," provide a
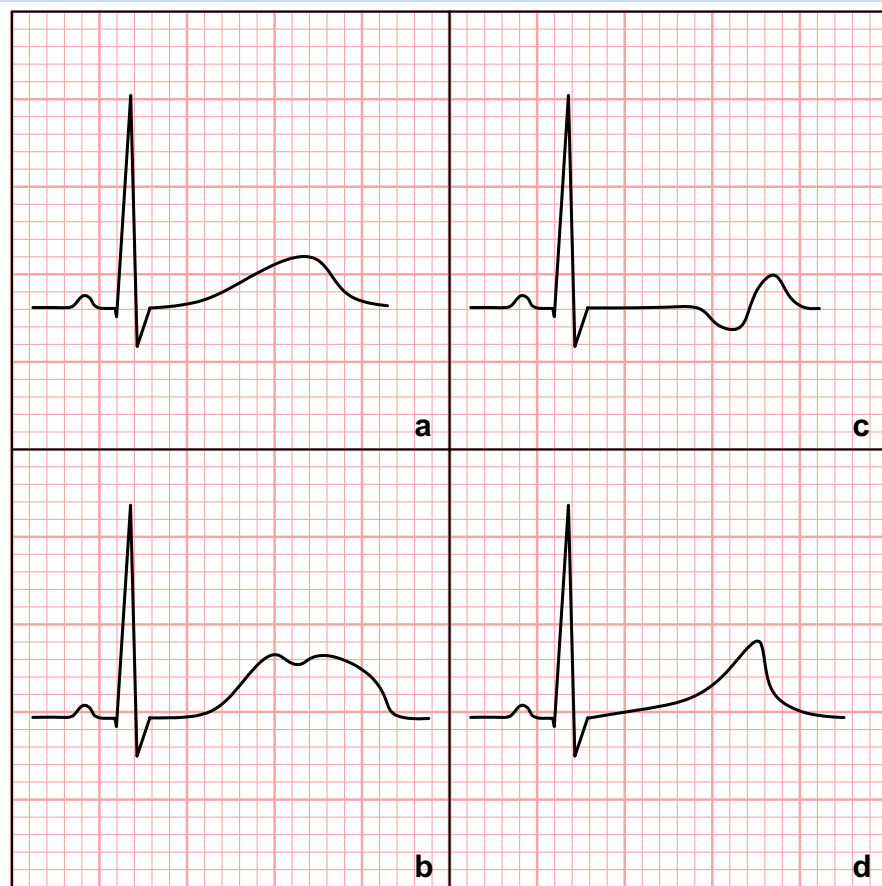
Figure 5. Various T-wave configurations in the most common long QT syndromes (LQTSs). (a) LQT1 is associated with broad-based T-waves. (b) LQT2 is often associated with bifid or notched T-waves. LQT3 is associated with (c) biphasic or (d) asymmetrical and peaked T-waves.
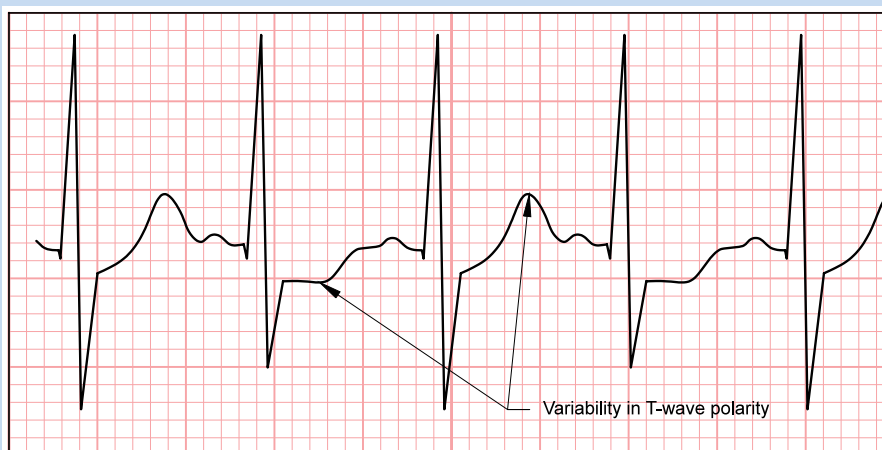

Figure 6. T-wave alternans, a finding associated with electrical instability often preceding torsades de pointes.

Table 2. 1993-2011 LQTS diagnostic criteria[a]

	Points
Electrocardiographic findings	
QTc[b] ≥480 ms 460-479 ms 450-459 ms (in males)	3 2 1
QTc[b] 4th minute of recovery from exercise stress test ≥480 ms	1
Torsades de pointes[c]	2
T-wave alternans	1
Notched T-wave in 3 leads	1
Low heart rate for age[d]	0.5
Clinical history	
Syncope[c] With stress Without stress	2 1
Congenital deafness	0.5
Family history	
Family members with definite LQTS[e]	1
Unexplained sudden cardiac death below age 30 years among immediate family members[e]	0.5

LQTS, long QT syndrome.
[a]Table adapted from Schwartz and Crotti.[26] Score: ≤1 point, low probability of LQTS; 1.5 to 3 points, intermediate probability of LQTS; ≥3 points, high probability of LQTS.
[b]QTc calculated using Bazett's formula.
[c]Mutually exclusive.
[d]Resting heart rate below the 2nd percentile for age.
[e]The same family member cannot be counted in either row 1 or 2 of this category.

tool to risk stratify patients into low (≤1 point), intermediate (1-3 points), or high probability (≥3 points) categories (Table 2).[26] The Heart Rhythm Society, European Heart Rhythm Association, and the Asia Pacific Heart Rhythm Society (HRS/EHRA/APHRS) indicate that the diagnosis of LQTS can be made if the patient has 1 of the following: Schwartz criteria score of ≥3.5, unequivocal pathogenic mutation, QTc ≥500 (using Bazett's correction), or QTc between 480 and 499 ms with symptoms (unexplained syncope in the absence of secondary causes and pathogenic mutation).[22]

The ECG must be completed in the absence of QT-prolonging medications (http://crediblemeds.org), supplements, or other acquired causes, such as hypocalcemia, hypokalemia, or hypothyroidism.[22] In addition, the presence of a bundle branch block or left ventricular hypertrophy can increase the QRS duration, which will also prolong the QT interval. Normal athlete-specific findings should be kept in mind when assessing the ECG (eg, Seattle criteria).[9] Further considerations include additional confounders, including diurnal variation and seasonal dependence of the QTc.[12] Epinephrine infusion testing has been used to assess for increases in the QT interval and is effective, particularly in LQT1.[32,38] However, given the limited availability and differing protocols for epinephrine infusion, treadmill exercise testing is more frequently used to assess LQTS.[26,35]

Family history must be properly evaluated. A family history of sudden death, especially in those younger than 30 years, should be further characterized; unexplained drowning, automobile accidents, seizures, or sudden infant death in family members should raise suspicion, as these may be the result of syncope or cardiac arrest from LQTS.[26]

One study indicated that approximately 2 of every 5 patients suspected of having LQTS referred to a LQTS specialty center were reclassified as normal, having insufficient evidence to merit the diagnosis. The majority of those misdiagnosed were the result of a miscalculated QTc, misinterpretation of the normal distribution of QTc values, and misinterpretation of associated symptoms, thus highlighting the importance of careful ECG analysis, awareness of diagnostic criteria, and an understanding of the clinical symptoms.[36]

GENETIC TESTING

The HRS and EHRA recommend comprehensive or targeted LQT1-3 (*KCNQ1, KCNH2, SCN5A*) genetic testing in those:

1. Whom a cardiologist has established a strong clinical index of suspicion (eg, Schwartz criteria) for LQTS based on examination of the patient's clinical history, family history, and ECG phenotype (resting 12-lead and/or provocative stress testing with exercise or catecholamine infusion) (class I)
2. With QT prolongation in the absence of other clinical conditions that might prolong the QT interval (ie, electrolyte abnormalities, hypertrophy, bundle branch block) on serial 12-lead ECGs defined as QTc >480 ms (prepuberty) or >500 ms (adults) (class I)
3. Any asymptomatic patient with otherwise idiopathic QTc values of >460 ms (prepuberty) or >480 ms (adults) on serial 12-lead ECGs (class IIB)

The HRS/EHRA guidelines, though widely cited, are not specific to athletes, so ECG interpretation should be assessed with common athlete-specific findings in mind (eg, Seattle criteria).[9] The HRS/EHRA further recommend mutation-specific genetic testing for family members and other appropriate relatives following the identification of the LQTS-causative mutation in an index case.[2] For clarity, an index case is the first identified individual in a group of related cases among those with a possible heritable or transmissible disease. It is important

to note that LQTS penetrance is variable, meaning genetically positive patients may have a normal or borderline QTc.[20,26,33] These silent mutation carriers accounted for 36% of LQT1 patients, 19% of LQT2 patients, and 10% of LQT3 patients in 1 cohort.[21] Genetic counseling prior to and after genetic testing should be administered so that the patient and family understand the uncertainties of testing interpretation and implications of the results.

MEDICAL TREATMENT

β-Blocker therapy should be the first-line therapy for LQT1-3 barring contraindications, such as active asthma.[2,22,25] β-Blockers are recommended in patients with a diagnosis of LQTS who are asymptomatic with QTc ≥470 ms and/or symptomatic with syncope or documented ventricular tachycardia/ventricular fibrillation (class I).[22] LQTS patients who are asymptomatic with QTc ≤470 ms may also benefit from β-blocker therapy (class IIa).[22] β-Blockers should therefore be used in most patients, though one reasonable exception includes men older than 40 years with LQT1 and QTc <500 ms, as patients in this group rarely have cardiac events.[25] Medication compliance is essential, as most events occur because of incomplete compliance.[25] Recurrent cardiac events while on therapy are a strong predictor of further recurrence.[2]

Propranolol is widely used (2-3 mg/kg/d and sometimes increased to 4 mg/kg/d or more). Nadolol is also frequently used given its longer half-life permits twice-daily administration (1-1.5 mg/kg/d). Atenolol and metoprolol have been associated with clinical failures more often than propranolol or nadolol.[1,25] Specifically, metoprolol has been associated with a 4-fold greater risk of event recurrences.[6] Abrupt discontinuation of β-blockers should be avoided as this may increase the risk of exacerbation.[22]

β-Blockers are prohibited in certain sports as they may confer a competitive advantage. According to the World Anti-Doping Agency (WADA), β-blockers are prohibited while competing in archery, automobile sports, billiards, darts, golf, shooting, certain ski/snowboard events, and certain underwater sports.[39]

Sodium channel blockers, such as mexiletine, can serve as adjunctive therapy for LQT3 patients who shorten their QTc following an acute oral drug test with 1 of these compounds (class IIa).[22] Targeting the pathologic LQT3-associated late sodium current with a β-blocker and the addition of mexiletine, flecainide, or ranolazine may be the best option for patients with LQT3.[2]

Patients with LQTS should avoid medications that prolong the QT interval (http://crediblemeds.org), such as certain antiarrhythmics, antifungals, antibiotics, antipsychotic agents, and nutritional supplements, among others. In addition, diuretics that lower potassium and magnesium should be used with caution.

SURGICAL TREATMENT

In higher risk patients, surgical treatment may be the most effective intervention. The primary options include implantable cardioverter-defibrillator (ICD) placement and left cardiac sympathetic denervation (LCSD).

ICD implantation is recommended in patients with a diagnosis of LQTS who are survivors of a cardiac arrest (class I). ICD implantation can also be useful in patients with a diagnosis who experience recurrent syncopal events while on β-blocker therapy (class IIa). Except under special circumstances, ICD implantation is not indicated in asymptomatic patients who have not been tried on β-blocker therapy (class III).[22] The M-FACT score is a tool that may identify patients who may benefit from an ICD implant.[29] Pacemaker implantation without an ICD can be used in a very select group of patients to prevent pause-dependent TdP, especially in those with LQT3, where bradycardia is a risk factor. Since an ICD also has pacemaker capabilities, an ICD is a more reasonable option in most patients if device implantation is planned.[25]

LCSD involves the removal of the first 4 thoracic ganglia and is very effective for reduction in cardiac events in high-risk candidates, including those with syncopal episodes despite full-dose β-blocker therapy.[25,27] LCSD is recommended in high-risk patients in whom ICD therapy is contraindicated or refused and/or β-blockers are not effective in preventing syncope/arrhythmias, not tolerated, not accepted, or contraindicated (class I). LCSD can be useful in patients with a diagnosis of LQTS who experience breakthrough events while on therapy with β-blockers/ICD (class IIa).[22]

GUIDELINES FOR ATHLETES

Given the association of cardiac events with exertion or other triggers, prior guidelines for athletes with LQTS were conservative and restrictive. The 2005 Bethesda Conference guidelines recommended that all previously symptomatic LQTS patients or those with ECG manifestations be restricted to class IA sports, such as billiards, bowling, cricket, curling, golf, or riflery.[43] Class IA sports are those with low static (I) and low dynamic (A) stresses according to the classification system outlined by Mitchell et al.[17] Static exercise involves large intramuscular force with minimal change in muscle length or joint movement and dynamic exercise involves significant change in muscle length and joint movement with relatively small intramuscular force.[17] Even more restrictive than the Bethesda Conference guidelines, the European Society of Cardiology (ESC), also published in 2005, recommended that all patients with LQTS not participate in any sport, even for those without documented major arrhythmic events.[19]

The prior rigid recommendations have evolved with the rise of shared decision making and strong supporting evidence to support the liberalization of the former guidelines.[1] The HRS, EHRA, and APHRS guidelines in 2013 facilitated this change, stipulating that an athlete with LQTS who desires to continue to participate in his or her sport be evaluated by an LQTS expert to determine whether he or she may participate.[22] Three major interval changes have occurred since the prior 2005 guidelines. First, widely available genetic testing permits confirmation of suspected channelopathies.[2,16] Second, even with increased discovery of genotype-positive/phenotype-negative or

concealed disease, resulting from wider availability of genetic testing, there have been no reports of athletes with concealed LQTS in the United States experiencing their sentinel event during sport.[16] Third, new data from the prospective ICD Sports Safety Registry and observational experience from 2 retrospective cohorts support liberalizing the 2005 recommendations; given the importance of the findings from these studies, they are further outlined below.[3,13,14,16]

The ICD Sports Safety Registry included 372 competitive athletes with an ICD, 44 of whom competed in high-risk sports (those in which an athlete could temporarily lose control, eg, skiing, surfing).[14] LQTS affected 73 athletes, and this comprised the most common diagnosis in the cohort. More than 20 different sports were included across all levels, from pre–high school to postgraduate. There were no occurrences of the primary endpoint, including tachyarrhythmic death, externally resuscitated tachyarrhythmia during or after sports participation, or severe injury resulting from arrhythmia-related syncope or shock during sport. Appropriate and inappropriate shocks occurred in 10% of subjects during competition or practice, 8% during other physical activity, and 6% at rest. Among the group with LQTS, only 2 had an ICD shock during competition or practice, both of which terminated with 1 shock. The ICD lead survival rate was 97% at 5 years and 90% at 10 years, which is similar to previously described rates in nonathlete ICD populations.[14]

In further support of this paradigm shift, Johnson and Ackerman[13] studied a cohort of 130 competitive and recreational athletes not limited to class IA sports. The majority of athletes were treated with β-blockers (86%); 16% had an LCSD procedure and 15% had an ICD. All 130 patients participated contrary to the ESC guidelines, while 60 athletes participated against the recommendations of both the ESC and 2005 Bethesda Conference guidelines. There were no deaths in this cohort. One high-risk 9-year-old male participant (history of VF-resuscitated, aborted cardiac arrest) received 2 appropriate ICD shocks in the setting of admitted noncompliance with β-blocker therapy. In total, the event rate was 1 in 650 athlete-years at publication in 2013.[13] A recent editorial by Ackerman[1] indicates that the event rate continues to remain low within this cohort, with a current rate of 1 in more than 1000 athlete-years.

Aziz et al[3] also provide support to the latest recommendations with similar findings. In a cohort of 103 LQTS genotype-positive patients aged >4 and <21 years and followed for 15 years, there were no occurrences of the primary endpoint (tachyarrhythmic death or externally resuscitated tachyarrhythmia, syncope, or severe injury resulting from arrhythmia-related syncope during or after sports participation) or secondary endpoints (appropriate shock, inappropriate shock, automated external defibrillator [AED] shock during or after sports, or sports-related ICD system damage). All patients in this group were taking a β-blocker, and 3 patients were on mexiletine (2 with SCN5A mutations). One appropriate ICD discharge occurred for TdP following mild exertion in the setting of β-blocker therapy

noncompliance. No cardiac events and no deaths were observed in treatment-compliant LQTS patients of this cohort during sports participation in 755 patient-years of follow-up.[3]

Based on the strength of these studies and a growing willingness to permit participation, the American Heart Association and American College of Cardiology (AHA/ACC) Scientific Statement in 2015 outlines the following participation guidelines, relevant to LQTS:

1. For athletes with a suspected/diagnosed cardiac channelopathy, a comprehensive evaluation by a heart rhythm specialist or genetic cardiologist with sufficient experience and expertise with these disorders is recommended (class I).
2. It is recommended that symptomatic athletes with any suspected or diagnosed cardiac channelopathy be restricted from all competitive sports until a comprehensive evaluation has been completed, the athlete and his or her family are well informed, a treatment program has been implemented, and the athlete has been asymptomatic on therapy for 3 months (class I).
3. It is reasonable for an asymptomatic athlete with genotype-positive/phenotype-negative (concealed channelopathy) LQTS to participate in all competitive sports with appropriate precautionary measures, including: avoidance of QT-prolonging drugs; electrolyte/hydration replenishment and avoidance of dehydration; avoidance or treatment of hyperthermia from febrile illnesses, training-related heat exhaustion, or heat stroke; acquisition of a personal AED as part of the athlete's personal sports safety gear; and establishment of an emergency action plan with the appropriate school or team officials (class IIa).
4. For an athlete with either symptomatic LQTS or ECG manifest LQTS (QTc >470 ms men and >480 ms in women), competitive sports participation (except competitive swimming in a previously symptomatic LQT1 host) may be considered after institution of treatment and appropriate precautionary measures, assuming the athlete has been asymptomatic on treatment for at least 3 months (class IIb).[16]

The 2015 AHA/ACC Scientific Statement also concludes that indications for competitive athletes should not differ from those applicable to the general population with appropriate diagnoses and clinical profiles (class I).[16] Although data are limited with regard to athletes with ICDs, it suggests they may play sports safely. According to the 2015 AHA/ACC guidelines, an athlete with an ICD may be permitted to participate in sports if there have been no shocks for 3 months. Permitting athletes to return to their preferred sport, including those with ICDs, remains highly dependent on the values of the patient and their family. Clearly the patient's autonomy and their decision should be the most important factor in returning to play, assuming adequate counseling and discussions with providers with sufficient expertise.[16,23]

CONCLUSION

The recommendations surrounding athletes with LQTS have evolved over the prior decade, empowering patients and their families to determine their preferred level of sports. The most recent 2015 ACC/AHA guidelines provide support and clarification regarding recommendations that have evolved from recent research. Despite its potential adverse consequences, LQTS should be perceived as a manageable and treatable condition not as a contraindication to sports participation, even if an ICD is present.

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