Competitive Athletic Participation for Patients with LQTS.

Who and How Much?

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<table>
<thead>
<tr>
<th>Congenital LQTS</th>
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<tbody>
<tr>
<td><strong>Epidemiology</strong></td>
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<tr>
<td>❖ Affects 1 in 2000-5000 people worldwide.</td>
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<tr>
<td>❖ Accounts for 4000 annual deaths in US</td>
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<tr>
<td>❖ In a population of 85 million, it is estimated that &gt; 40000 have LQTS</td>
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<tr>
<td>❖ Is underestimated due to concealed/missed cases or unexplained SCD.</td>
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<table>
<thead>
<tr>
<th>Type</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>LQT1</td>
<td>30-50%</td>
</tr>
<tr>
<td>LQT2</td>
<td>25-40%</td>
</tr>
<tr>
<td>LQT3</td>
<td>5-10%</td>
</tr>
<tr>
<td>LQT4</td>
<td>&lt;1%</td>
</tr>
<tr>
<td>LQT5</td>
<td>1%</td>
</tr>
<tr>
<td>LQT6</td>
<td>&lt;1%</td>
</tr>
<tr>
<td>LQT7</td>
<td>&lt;1%</td>
</tr>
<tr>
<td>LQT8</td>
<td>&lt;1%</td>
</tr>
<tr>
<td>LQT9</td>
<td>&lt;1%</td>
</tr>
<tr>
<td>LQT10</td>
<td>&lt;1%</td>
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</tbody>
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Pathophysiology

**LQT: 1,2**

Inactivating mutation affecting Ikr

**ECG**

Pathophysiology

**LQT: 3**

Activating mutation affecting Na current

**ECG**
\[ \text{T-peak to T-end} = \text{Transmural dispersion of repolarization} \]

Transmural functional substrate for reentry
LQT1

All LQTS patients, with either a personal or family history of a near drowning, have a defective KCNQ1 (LQT1)


LQT-2

SCD occurs with sudden auditory stimuli, typically alarm clocks
LQT-3

SCD occurs typically during night sleep

Methods of measuring QT interval

Threshold method

Tangent method
**Modified Schwartz score for the diagnosis of LQTS**

<table>
<thead>
<tr>
<th>Variable</th>
<th>Points</th>
</tr>
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<tbody>
<tr>
<td><strong>ECG findings</strong></td>
<td></td>
</tr>
<tr>
<td>QTc ms ≥ 480</td>
<td>3</td>
</tr>
<tr>
<td>460–470</td>
<td>2</td>
</tr>
<tr>
<td>450 (in males)</td>
<td>1</td>
</tr>
<tr>
<td>Torsade de pointes</td>
<td>2</td>
</tr>
<tr>
<td>T wave alternans</td>
<td>1</td>
</tr>
<tr>
<td>Notched T wave in three leads</td>
<td>1</td>
</tr>
<tr>
<td>Low heart rate for age</td>
<td>0.5</td>
</tr>
<tr>
<td><strong>Clinical history</strong></td>
<td></td>
</tr>
<tr>
<td>Syncopy</td>
<td></td>
</tr>
<tr>
<td>With stress</td>
<td>2</td>
</tr>
<tr>
<td>Without stress</td>
<td>1</td>
</tr>
<tr>
<td>Congenital deafness</td>
<td>0.5</td>
</tr>
<tr>
<td><strong>Family history</strong></td>
<td></td>
</tr>
<tr>
<td>Family members with definite LQTS</td>
<td>1</td>
</tr>
<tr>
<td>Unexplained sudden cardiac death &lt; age 30 years</td>
<td>0.5</td>
</tr>
<tr>
<td>among immediate family members</td>
<td></td>
</tr>
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**Score ≥3.5 : Definite LQTS**  
**Score 2-3: Intermediate probability**  
**Score ≤1: Low probability**


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

**Diagnostic clues**

**Holter monitoring**
- Identifies paradoxic behavior of QTc to HR (lack of normal shortening with high HR)
- Identifies dynamic morphology of T wave

**Exercise testing**
- Pts with concealed LQT1,2 have diminished chronotropic response and exaggerated prolongation of the QT interval during recovery

**Epinephrine test**
- Sensitivity and specificity up to 91% and 100% respectively
Epinephrine test for concealed LQTS

Treatment of congenital LQTS

- Avoiding the triggering activities and medications that prolong QT.
- BBs (LQT1,2).
- Left stellectomy in poorly responders to BBs.
- Pacing (LQT3)
- Mexiletine (LQT3): (no data proving effects on SCD prevention)
  - Adjuvant to ICD.
  - In neonates and small infants.
- K supplements (LQT2)?.
- ICD for pts with syncope (IIa), or as 1ry prevention (IIb) in pts with QTc >500 ms, LQT2,3.
- Family screening-pt education.
Activity should be restricted to low-intensity sports (e.g., billiards, golf, curling, riflery) for athletes with LQTS who have had cardiac arrest or an episode of syncope.

2. Asymptomatic athletes with prolongation of the QT interval on the ECG should be restricted to low-intensity sports.

3. Asymptomatic athletes, who are known gene carriers, may participate fully in their sports.

4. Athletes with an ICD should participate only in low-intensity sports and should avoid all situations where bodily injury might occur.
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**ESC guidelines in 2005**

- Congenital long QT syndrome is a contraindication for any type of sports, even without documented major arrhythmic events.

- Recommended using QTc values of >440 ms (males) or >460 ms (females) as a trigger for further evaluation.

<table>
<thead>
<tr>
<th>Long QT syndrome</th>
<th>History, ECG, (24 h Holter, genetic testing)</th>
<th>Positive long QT syndrome</th>
<th>No competitive sports</th>
</tr>
</thead>
</table>

There are well known cases of athletes who have decided to continue to play at a highly competitive level. One famous case is of Olympic swimmer Dana Vollmer who competed at the London Olympic Games in 2012. At the age of 15, Dana was diagnosed with LQTS after experiencing a dizzy spell while training.

Vollmer and her family decided to implant a defibrillator, and closely monitor her training regimen so she could continue to swim. Dana is an inspiration to all athletes living with LQTS as her perseverance and bravery earned her a gold medal at the 2012 London Olympic Games.

Although Dana now appears to have outgrown her LQTS symptoms, her decision to continue to swim “illustrates that some athletes can still participate in competitive sports despite cardiac defect”

If he has LQTS, shall you disqualify him ?!

Three major interval changes have occurred since the prior 2005 guidelines

1- Widely available genetic testing permits confirmation of suspected channelopathies

2- There have been no reports of athletes with concealed LQTS in the United States experiencing their sentinel event during sport.

3- New data from recent studies and observational experience
Recent studies have changed the paradigm

In a cohort of 130 athletes with long QT syndrome (LQTS) who continued to participate in competitive athletics following their diagnosis, only one had a cardiac event in over 650 athlete-years of follow-up.

One patient with an event was a known high-risk 9-year-old LQT1 male with a history of cardiac arrest and implantable cardioverter-defibrillator (ICD) placement, who received two appropriate ICD therapies, both while warming up prior to practices, and both during admitted non-compliance with β-blocker medication.


Recent studies have changed the paradigm

Another study included a cohort of both competitive and recreational sports

Competitive

Recreational
Recent studies have changed the paradigm

- No patients participating in competitive sports had syncope, documented arrhythmia, or aborted cardiac arrest during sports-related activity.

- No patients competing in recreational sports had syncope, documented arrhythmia, or aborted cardiac arrest during sports-related activity.


The ICD Sports Safety Registry

- Included 372 competitive athletes with an ICD, 44 of whom competed in high-risk sports (eg, skiing).

- LQTS affected 73 athletes, and this comprised the most common diagnosis in the cohort.

- 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.

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 genotypepositive/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; (class IIa)
AHA/ACC Scientific Statement in 2015 outlines the following participation guidelines, relevant to LQTS:

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)


**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 condition not as a contraindication to sports participation, even if an ICD is present.
Thank You

Osama Diab