Should sports participation be restricted in LQTS?

L. Környei Hungarian Pediatric Heart Center







5th February 2017 14:00 – 15:15 Inherited Arrhythmia Syndromes VI: Long QT Syndrome: Complicated Cases



- 1. Early data
- 2. Recent data
- 3. Recent recommendation
- 4. The "real" question to be answered



- increased frequency of cardiac events during exercise in patients with LQT1
 - (swimming 62% vs sleeping 3%)

Schwartz, Circulation 2001

 In untreated patients, LQTS-related cardiac events occurred in 55%

Shimizu, JACC 2001

• Thus, guidelines have historically advocated for an aggressive restriction from competitive sports, with the exception of low-intensity sports

Bethesda 36, ESC 2005

"historical decision making"

- beta-blockade
- avoidance of medications that prolong the QT interval
- "you're not to play sports"

Physiologic benefits of exercise

sports participation brings to children and adolescents

- enhanced self-confidence,
- sense of psychological, physical and social wellbeing
- improved overall quality of life

...but the pendulum is now swinging away...

It started with ..

ONLINE FIRST

RESEARCH LETTER

Competitive Sports Participation in Athletes With Congenital Long QT Syndrome

Johnson, Ackerman, JAMA, August 22/29, 2012-Vol 308, No. 8

Research letter

- 105 mutations
- most common mutation was KCNQ1 (58%)
 - KCNH2 (35%),SCN5A (6%),KCNE2 (2%),KCNE1 (1%)
- All patients were treated with **beta-blockade**
 - (1 noncompliant, 1 intolerant)
- 26 pts competitive sports (15 girls; mean follow-up 6.9 years; mean QTc, 461 ms
- 77 pts recreational sports (35 girls; mean follow-up 7.3 years; mean QTc, 470 ms
- During 755 patient-years of follow-up, no patients had long QT syndrome symptoms during sports
- 5 appropriate ICD shocks in 2 pts, but non were related to sports

...but the pendulum is now swinging away...

It is accelerated by ..

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Sports Participation in Genotype Positive Children With Long QT Syndrome



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Pediatric data

- Children's Hospital of Philadelphia
- from 1998 to 2013
- 212 patients with LQTS
- 103 participated in sports (53 girls)
- all were treated with BB—(nadolol for 97 pts)
- mean follow-up, 7.1 years
- mean QTc, 468 ms
- there were no cardiac events and no deaths

Recent data

Sports participation appears safer today for children with long QT syndrome ?

Explanation ?

Different patient population

- Ten years ago before genotyping was mainstream, most patients were diagnosed as a result of an event
- With the introduction of genetic testing and cascade familial testing, there are a lot more patients being identified with LQTS, without any event
- the chance of a patient without symptoms presenting for the first time with sudden cardiac death/cardiac arrest is approximately 5%

"shared decision"

- if the restriction is devastating, and in some cases it might be, harming the child's psychological, physical, or social well-being
- sports participation might be safe in certain circumstances

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AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 10: The Cardiac Channelopathies

A Scientific Statement From the American Heart Association and American College of Cardiology

Michael J. Ackerman, MD, PHD, FACC, Chair*

Douglas P. Zipes, MD, FAHA, MACC* Richard J. Kovacs, MD, FAHA, FACC* Barry J. Maron, MD, FACC* Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 10: The Cardiac Channelopathies

Recommendations

 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; Level of Evidence C). Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 10: The Cardiac Channelopathies

Recommendations

 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

Eligibility and Disqualification



Recommendations for Competitive Athletes

It is reasonable for an **asymptomatic** athlete with genotype-positive/ phenotype-negative LQTS

1) avoidance of QT-prolonging drugs for athletes with LQTS (<u>http://www.crediblemeds.org</u>)

3) electrolyte/ hydration replenishment and avoidance of dehydration

4) avoidance or treatment of hyperthermia from febrile illnesses or trainingrelated heat exhaustion or heat stroke for athletes with either LQTS or BrS

5) acquisition of a personal automatic external defibrillator as part of the athlete's personal sports safety gear, and

6) establishment of an emergency action plan with the appropriate school or team officials (**Class IIa**; Level of Evidence C).

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 10: The Cardiac Channelopathies

Recommendations

5. For an athlete with either symptomatic LQTS or electrocardiographically manifest LQTS (i.e., corrected QT interval >470 ms in males or >480 ms in females), 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)

Practical recommendations

- BB 15%-20% reduction in peak HR during exercise
- avoid routinely exceeding 65% of their target HR
- emphasize the importance of
 - compliance with medication
 - maintaining hydration
 - resting when experiencing symptoms
 - rescue systems are in place
- good relationship with the family
 - surveillance monitoring for efficacy of medical therapy (Holter, exercise testing)
 - access to advice and feedback regarding exercise

Cheung et al, Canadian Journal of Cardiology 32 (2016) 452e458

Making recommendations from evaluation to participation



Cheung et al, Canadian Journal of Cardiology 32 (2016) 452e458

Fact and question

Fact:

aerobic activity is a "potential risk-taking behavior" for patients with LQTS

Question:

Whether the increased risk can be "circumvented, neutralized, or minimized" rather than disqualifying the patient from sports entirely. *M. Ackerman* Cartoon to encourage safe and supervised physical activity in children with LQTS



Cheung et al, Canadian Journal of Cardiology 32 (2016) 452e458