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

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The cardiac channelopathies are a collection of primary, genetically mediated heart rhythm disorders (also referred to as the primary electrical disorders) that are generally associated with a structurally normal heart and a propensity for syncope, seizures, or sudden cardiac arrest precipitated by a channelopathy-mediated episode of nonsustained or sustained polymorphic ventricular tachycardia (torsade de pointes) or ventricular fibrillation. These cardiac channelopathies include long-QT syndrome (LQTS), catecholaminergic polymorphic ventricular tachycardia (CPVT), Brugada syndrome (BrS), early repolarization syndrome, short-QT syndrome, and potentially idiopathic ventricular fibrillation. Approximately 1 in 1000 people are affected by a cardiac channelopathy, with LQTS being most common, involving an estimated 1 in 2000 people.[1]

Presently, these channelopathies should be viewed as potentially lethal but highly treatable conditions. However, unlike the various bradyarrhythmias and tachyarrhythmias detailed in the Task Force 9 report,[2] there remains significant variability and heterogeneity among pediatric and adult heart rhythm specialists in terms of their ability to diagnose, risk stratify, and treat patients with these conditions. For example, in 1 study, 40% of the patients who received a second opinion evaluation at a LQTS specialty center for a previously rendered diagnosis of LQTS by a heart rhythm specialist were reclassified as otherwise normal, having insufficient evidence to merit that diagnostic consideration.[3] This is explained in part by the advanced knowledge and training required to evaluate and treat these less common channelopathies. Accordingly, any return-to-play decision for an athlete suspected of having a cardiac channelopathy necessitates that the athlete be evaluated, risk stratified, treated, and counseled by a heart rhythm specialist or genetic cardiologist with sufficient experience and expertise in these syndromes.[4]

For the most part, restriction from virtually all competitive sports has been the guideline-based recommendation since 2005 for athletes with a cardiac channelopathy, regardless of the underlying channelopathy.[5,6] This universal recommendation was given despite the observation that exercise or competitive

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The Preamble and other Task Force reports for these proceedings are available online at http://circ.ahajournals.org (*Circulation*. 2015;132:e256–e261; e262–e266; e267–e272; e273–e280; e281–e291; e292–e297; e298–e302; e303–e309; e310–e314; e315–e325; e330–e333; e334–e338; e339–e342; e343–e345; and e346–e349).

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athletics has only been established as a potentially proarrhythmic trigger for CPVT and LQTS (particularly LQT1).[7,8]

Since 2005, there have been 4 fundamental developments that inform these current recommendations. First, genetic testing is now a widely available clinical test used routinely in the evaluation of a patient with a suspected channelopathy. The first Heart Rhythm Society/European Heart Rhythm Association–sponsored guideline as to the clinical use of genetic testing for the cardiac channelopathies was published in 2011.[9]

Second, despite increased discovery of more family members (athletes and nonathletes alike) with genotype positive/phenotype-negative (ie, concealed disease) status secondary to the availability and use of genetic testing, there has been no report of athletes with concealed channelopathic substrates in the United States experiencing their sentinel event during sport. Thus, consistent with our expert opinion–based recommendations from a decade ago, there has been no observational evidence to support the European position to disqualify an athlete based solely on a positive genetic test.[5,6]

Nevertheless, it remains prudent for an athlete with a channelopathy, whether concealed or manifest, to exercise simple precautionary measures, including (1) avoidance of QT-prolonging drugs for athletes with LQTS (http://www.crediblemeds.org), (2) avoidance of drugs that exacerbate the BrS in affected athletes (http://www.brugadadrugs.org), (3) electrolyte/hydration replenishment and avoidance of dehydration for all, (4) avoiding/treating hyperthermia from febrile illnesses or training-related heat exhaustion/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) establishing an emergency action plan with the appropriate school/team officials.

Third, observational evidence, derived from a large series of athletes with either concealed, electrocardiographically manifest, or symptomatic LQTS who chose to remain competitive despite the 2005 guideline-based recommendations for their disqualification, now exists.[10,11] In this single-center study of LQTS athletes, only 1 of the 130 athletes with LQTS (LQT1 specifically) experienced 2 LQT1-triggered events that resulted in appropriate ventricular fibrillation–terminating implantable cardioverter-defibrillator (ICD) therapies while playing baseball on 1 occasion and soccer on another occasion in >650 athlete-years of observation. An important caveat is that every athlete underwent an extensive 2- to 3-day evaluation that included being diagnosed, risk stratified, treated, and counseled by a single LQTS specialist. This program's experience has been reproduced independently in a study involving sports participation in genotype-positive children at another center.[12]

At this point in time, no similar data exist for athletes with CPVT. Given that CPVT is likely the channelopathy most vulnerable to exercise as a proarrhythmic trigger, the likelihood of a CPVT-triggered breakthrough event despite β-blocker use is much higher than in LQTS,[7] and the potential for an arrhythmia/ICD storm is greatest in patients with CPVT,[13] competitive sports (beyond class IA sports) are not recommended for the athlete with CPVT and documented exercise-induced frequent premature ventricular contractions/nonsustained ventricular tachycardia. Whether or not such an athlete could be cleared in the setting of combination drug therapy (for example, β-blockers and flecainide) or after left cardiac sympathetic denervation would require consultation with a CPVT disease specialist.

Fourth, the observational experience from the North American ICD Sports Registry currently comprising >340 athletes with an ICD suggests that these athletes with an ICD can continue to participate with negligible mortality (0 deaths with 31 months' average follow-up to date) and no discernible excess in damage to the implanted device or inappropriate shocks to the patient.[13] The most common heart disease represented among these athletes with an ICD was LQTS, followed by hypertrophic cardiomyopathy and arrhythmogenic right ventricular cardiomyopathy.

Despite these 4 new developments over the past decade, there remains an overall lack of data or evidence regarding the true risk that an athlete with a channelopathy faces by remaining in competitive sports. As such, these recommendations are buttressed by only Level of Evidence C.

For the purposes of this document, a previously symptomatic athlete describes one who has experienced at least 1 channelopathy-triggered/suspected syncope, seizure, or aborted/resuscitated cardiac arrest. On the other hand, an athlete with a concealed channelopathy describes an asymptomatic athlete with a positive genetic test who lacks electrocardiographic evidence on a 12-lead ECG at rest (ie, corrected QT interval <460 ms for LQTS, no spontaneous type 1 Brugada electrocardiographic pattern in the right precordial leads for BrS, no horizontal or downsloping early repolarization pattern in the inferolateral leads for early repolarization syndrome, or corrected QT interval >380 ms for short-QT syndrome) or during exercise stress testing for CPVT (ie, no exercise-induced premature ventricular contractions in bigeminy, couplets, or worse). An athlete with a concealed channelopathy is also referred to as genotype positive/phenotype negative.

In addition, for the purposes of this document, disease-specific treatments may include either drug therapy, denervation therapy (ie, left cardiac sympathetic denervation for LQTS and CPVT), device therapy (generally an ICD rather than a pacemaker if device therapy is indicated), or a combination thereof. The athlete's treatment program should be based primarily on the severity of the disease phenotype and should not be unduly influenced by the patient's athlete status. In other words, an ICD should not be implanted just because the patient happens to be an athlete in order for the patient to remain an athlete. This individualized treatment program should be sought from a center or program dedicated to patients with cardiac channelopathies and implemented by a heart rhythm specialist or genetic cardiologist with sufficient experience and expertise with these disorders.[4] Finally, it may be prudent to temporarily restrict an athlete who experiences a cardiac event and is suspected of having a channelopathy or the athlete with a known channelopathy who experiences a breakthrough cardiac event for 3 months to ensure adequate time for evaluation, counseling, and initiation or modification of the athlete's treatment program.

Recommendations

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; Level of Evidence C)*.**
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; Level of Evidence C)*.**
3. **It is reasonable for an asymptomatic athlete with genotype-positive/phenotype-negative (ie, concealed channelopathy) LQTS, CPVT, BrS, early repolarization syndrome, idiopathic ventricular fibrillation, or short-QT syndrome to participate in all competitive sports with appropriate precautionary measures, including (1) avoidance of QT-prolonging drugs for athletes with LQTS (http://www.crediblemeds.org), (2) avoidance of drugs that exacerbate the BrS in affected athletes (http://www.brugadadrugs.org), (3) electrolyte/hydration replenishment and avoidance of dehydration for all, (4) avoidance or treatment of hyperthermia from febrile illnesses or training-related 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)*.**
4. **Competitive sports participation may be considered for an athlete with either previously symptomatic or electrocardiographically evident BrS, early repolarization syndrome, or short-QT syndrome assuming appropriate precautionary measures and disease-specific treatments are in place and that the athlete has been asymptomatic on treatment for at least 3 months *(Class IIb; Level of Evidence C)*. If therapy includes an ICD, refer to the Task Force 9 report.[2]**
5. **For an athlete with either symptomatic LQTS or electrocardiographically manifest LQTS (ie, 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; Level of Evidence C)*. If treatment includes an ICD, refer to the Task Force 9 report[2] for recommendations regarding restrictions after the procedure, lead replacements, and so forth.**
6. **For an athlete with previously symptomatic CPVT or an asymptomatic CPVT athlete with exercise-induced premature ventricular contractions in bigeminy, couplets, or nonsustained ventricular tachycardia, participation in competitive sports is not recommended except for class IA sports *(Class III; Level of Evidence C)*. Exceptions to this limitation should be made only after consultation with a CPVT specialist.**

Disclosures

Writing Group Disclosures

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Michael J. Ackerman	Mayo Clinic	NIH (R01 grants)†	None	None	None	None	Boston Scientific*; Gilead Sciences*; Medtronic*; St. Jude Medical*	Transgenomic†
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*Modest.

†Significant.

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