

itive sports that pose a danger of bodily collision because such trauma may damage the pacemaker system. Before allowing athletes to engage in these activities, an exercise test should be done at the level of activity demanded by the particular sport to be certain that the paced heart rate increases appropriately.

Congenital complete heart block. The clinical approach to evaluating the severity of the cardiovascular abnormality includes an echocardiogram, 12-lead ECG, 24-h ECG recording during exercise, and exercise stress test (exercise testing should be performed at the same exercise level as that during the sports activity).

Recommendations:

1. Athletes with a structurally normal heart and normal cardiac function, with no history of syncope or near syncope, a narrow QRS complex, ventricular rates at rest greater than 40 to 50 beats/min increasing appropriately with exertion, no or only occasional premature ventricular complexes, and no VT during exertion can participate in all competitive sports.
2. Athletes with ventricular arrhythmia, symptoms of fatigue, near-syncope, or syncope should have a pacemaker implanted before they participate in competitive sports. Athletes with pacemakers should not participate in competitive sports when the danger of bodily collision exists because such trauma may damage the pacemaker system. Before allowing athletes to engage in these activities, an exercise test should be conducted at the level of activity demanded by the particular sport so as to be certain that the paced heart rate increases appropriately.
3. Athletes with abnormal hemodynamic status, as those with an intracardiac shunt, cannot participate in any competitive sports without a pacemaker. Restrictions are the same as those in recommendation 2.

Acquired complete heart block.

Recommendations:

1. Patients with acquired complete heart block should be treated with pacing before any athletic activity.
2. Athletes with a pacemaker should not participate in competitive sports posing a danger of bodily collision because such trauma may damage the pacemaker system.

Complete right bundle-branch block. Evaluation includes a 12-lead ECG, exercise test, and echocardiogram. In some instances, a 24 h ECG recording may be indicated.

Recommendation:

1. Athletes without ventricular arrhythmias who do not develop AV block with exercise and who have no symptoms can participate in all competitive sports

consistent with their cardiac status. This also applies to athletes with associated left-axis deviation.

Complete left bundle-branch block. Evaluation includes a 12-lead ECG, exercise test, and echocardiogram. In some instances, a 24-h ECG recording may be indicated. Because of the rarity of acquired left bundle-branch block in children and its association with syncope from presumed paroxysmal AV block, an invasive EPS should be considered in young patients.

Recommendations:

1. Adult athletes with acquired left bundle-branch block should follow the recommendations under the section entitled Complete Right Bundle-Branch Block.
2. Athletes with a normal HV interval and a normal AV conduction response to pacing can participate in all competitive sports consistent with their cardiac status.
3. Athletes with abnormal AV conduction characterized by an HV interval greater than 90 ms or a His-Purkinje block should have pacemaker implantation. They should be restricted from competitive sports that hold a danger of bodily collision because such trauma may damage the pacemaker system.

Inherited arrhythmia syndromes. LONG QT SYNDROME. The definitive clinical diagnosis of congenital long QT syndrome (LQTS) can be complex (20). Debate continues as to what QTc constitutes the upper limit of normal. An increasing proportion of asymptomatic individuals with genetically proven LQTS are found to have a normal resting ECG with a heart rate corrected QT interval (QTc) by Bazett's formula of less than 460 ms (genotype positive/phenotype negative LQTS). In addition, a QTc of 440 ms, used in the past as an upper limit of normal, is present in far too many normal individuals (greater than 25%) to serve as a meaningful upper limit cut-off value. In general, a QTc of 470 ms or more in males and 480 ms or more in females requires further investigation as to the presence of congenital (or acquired) causes of QT prolongation. A patient with LQTS and a resting QTc of 500 ms or more is generally considered at increased clinical risk for a significant arrhythmia (21). One approach to the diagnosis of congenital LQTS is to utilize the "Priori-Schwartz" score that incorporates QTc, T-wave morphology, symptomatic presentation, and family history into the diagnostic algorithm (21). A "Priori-Schwartz" score of 4 or more suggests high clinical probability for LQTS. In addition, genetic testing for the five cardiac ion-channel genes responsible for 75% of LQTS (LQT1, LQT2, LQT3, LQT5, and LQT6) is now available as a commercial diagnostic test (22).

Mutations involving the structural protein ankyrin-B underlie the rare LQT4 form (23). Mutations involving the *KCNJ2*-encoded IK1 potassium channel account for approximately one-half of Andersen-Tawil syndrome (ATS1)

characterized by abnormal U waves, a prolonged QU interval, periodic paralysis, and facial and skeletal dysmorphisms. The ATS1 has been annotated in the past as LQT7.

Physical exertion (particularly swimming) appears to be a common trigger for ventricular arrhythmias in LQT1, whereas individuals with LQT2 seem more at-risk to auditory/emotional triggers, and patients with LQT3 may be at greater risk during rest and inactivity (24,25). However, exceptions to these genotype-phenotype correlations hinder genotype-specific tailoring of competitive sports recommendations. The entire personal and family phenotype must be incorporated before any eligibility or disqualification decision is rendered.

Recommendations:

1. Regardless of QTc or underlying genotype, all competitive sports, except those in class IA category should be restricted in a patient who has previously experienced either: 1) an out-of-hospital cardiac arrest, or 2) a suspected LQTS-precipitated syncopal episode.
2. Asymptomatic patients with baseline QT prolongation (QTc of 470 ms or more in males, 480 ms or more in females) should be restricted to class IA sports. The restriction limiting participation to class IA activities may be liberalized for the asymptomatic patient with genetically proven type 3 LQTS (LQT3).
3. Patients with genotype-positive/phenotype-negative LQTS (i.e., identification of a LQTS-associated mutation in an asymptomatic individual with a nondiagnostic QTc) may be allowed to participate in competitive sports. Although the risk of sudden cardiac death is not zero in such individuals, there is no compelling data available to justify precluding these individuals (who are being identified with increasing frequency) from competitive activities. Because of the strong association between swimming and LQT1, persons with genotype-positive/phenotype-negative LQT1 should refrain from competitive swimming.
4. LQTS patients with an ICD/pacemaker should not engage in sports with a danger of bodily collision because such trauma may damage the pacemaker system. The presence of an ICD should restrict individuals to class IA activities.

SHORT QT SYNDROME. Individuals with the short QT syndrome (SQTS) (QTc less than 300 ms) have a short QT interval and ventricular refractory period, and at least some of them have “gain-of-function” abnormalities in either I_{Kr} (*KCNH2*) or I_{Ks} (*KCNQ1*) (26).

Recommendation:

1. Until the phenotype of SQTS is better understood, a universal restriction from competitive sports with the

possible exception of class IA activities seems to represent the most prudent recommendation (27).

CATECHOLAMINERGIC POLYMORPHIC VENTRICULAR TACHYCARDIA (CPVT). Approximately one-half of patients with CPVT is pursuant to mutations involving the *RyR2*-encoded ryanodine receptor (sarcoplasmic reticulum calcium release channel). Such individuals are vulnerable to exercise-induced VT/ventricular fibrillation.

Recommendations:

1. Symptomatic patients have a poor prognosis unless treated with an implantable cardioverter-defibrillator (ICD) (28) and all such patients are restricted from competitive sports with the possible exception of minimal contact, class IA activities. As with LQT1, patients with CPVT should be restricted from competitive swimming. Asymptomatic patients detected as part of familial screening with documented exercise- or isoproterenol-induced VT should refrain from all competitive sports except possible class IA activities. A less restrictive approach may be possible for the genotype-positive/phenotype-negative (asymptomatic, no inducible VT) athlete.

BRUGADA SYNDROME (BrS). Brugada syndrome (29), characterized by an accentuated J-wave primarily in leads V_1 through V_3 , with ST-segment elevation, often followed by a negative T-wave and an R prime, may be the cause of sudden unexplained death syndrome, typically during sleep. Only 15% to 20% of BrS is established as a channelopathy due to mutations involving the *SCN5A*-encoded alpha subunit of the cardiac sodium channel (30). Individuals with BrS and no previous cardiac arrests may have a high risk of sudden death if they have inducibility of ventricular arrhythmias and a previous history of syncope (28). Hyperthermia can potentially unmask the Brugada ECG pattern in patients with BrS, who can then display fever-induced polymorphic VT. Death often occurs with mild activity or during sleep.

Recommendations:

1. Although a clear association between exercise and sudden death has not been established, and because of the potential impact of hyperthermia, restriction to participation in class IA sports seems advisable.
2. The presence of an ICD device warrants the same restrictions to class IA sports as previously outlined.

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TASK FORCE 7 REFERENCES

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