

AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 3: Hypertrophic Cardiomyopathy, Arrhythmogenic Right Ventricular Cardiomyopathy and Other Cardiomyopathies, and Myocarditis



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

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Hypertrophic cardiomyopathy (HCM) (1,2) is a major focus of this document given that it is the single most common cause of sudden death in young competitive athletes in the United States, responsible for at least one-third of these events (3).

HYPERTROPHIC CARDIOMYOPATHY

HCM is the most frequent nontraumatic cause of sudden death in the young (1,2) and a common genetic heart disease, occurring in at least 1 in 500

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people in the general population (4). HCM is a clinically and genetically heterogeneous disease, associated with >1,500 mutations in ≥ 11 major genes (and a variety of other susceptibility genes with lesser evidence for pathogenicity), encoding proteins of the cardiac sarcomere, adjacent Z disk, and calcium handling (5).

Although HCM is associated with substantial diversity in morphological expression (6), clinical diagnosis usually occurs with recognition of the characteristic disease phenotype, that is, left ventricular (LV) hypertrophy without chamber dilatation in the absence of another cardiac or systemic disease capable of producing the magnitude of hypertrophy evident (1,6). Neither systolic anterior motion of the mitral valve, hyperdynamic LV function, or identification of pathogenic sarcomere mutations is obligatory for the clinical diagnosis of HCM (2). Atrial fibrillation is a common cause of morbidity in HCM, occurring in $\approx 20\%$ of patients, although usually after 30 years of age (1,2). Notably, the clinical presentation and course are diverse, with unexpected sudden death in the young the most visible disease complication.

SUDDEN DEATH RISK

A major impetus in HCM has been the identification of those patients at increased risk for sudden death. Indeed, a risk-stratification algorithm has been largely effective in identifying those people at highest risk who are eligible for primary prevention of sudden death with an implantable cardioverter-defibrillator (ICD) (7-10), thereby markedly reducing HCM-related mortality to 0.5% per year (7). Sudden death events are attributable to potentially lethal ventricular tachyarrhythmias (ventricular tachycardia/ventricular fibrillation) and usually occur in the presence of ≥ 1 the major risk markers (appropriate ICD interventions of 4% per year in patients implanted for primary prevention) (7-10). Some HCM patients may nevertheless die suddenly in the absence of all conventional risk factors (0.6% per year in non-ICD populations) (7).

Indeed, in the presence of underlying (and often unsuspected) HCM, participation in high-intensity competitive sports may itself promote ventricular tachycardia/ventricular fibrillation and act as a potent (yet modifiable) independent risk factor, even in the absence of conventional risk markers intrinsic to the disease process (3,7,11,12). Notably, the underlying electrophysiological substrate in HCM is unpredictable (1,2,7-10) and potentially subject to instability by interaction with physiological stresses inherent in athletic training and competition, including alternations in hydration, blood volume, and electrolytes, as well as the catecholamine surge.

Given these principles, it is difficult to apply conventional risk-stratification strategies to make reliable eligibility decisions specifically for aspiring competitive athletes with

HCM. The estimation of risk level based on phenotypic expression (e.g., specific LV wall thickness or LV outflow tract gradient) or other aspects of the clinical profile is a highly problematic endeavor. Such considerations are influenced by the morphological diversity of HCM and the unpredictable instability of the myocardial substrate, as well as the additive risk created by intense training and competition in susceptible patients with HCM (3). Therefore, in HCM, the most common cause of sudden death in young athletes (1-3), engagement in intense competitive sports is itself an acknowledged modifiable risk factor (1-3).

These observations necessitate conservative and prudent recommendations regarding sports eligibility applied in a homogeneous fashion across the broad HCM disease spectrum. This may unavoidably result in recommendations for disqualification in some athletes with HCM probably at low risk and unlikely to ever experience sudden death, who could potentially compete and train safely. Notably, the present disqualification/eligibility guidelines for competitive athletes with HCM do not differ measurably from those previously stated in the 36th Bethesda Conference (11), because alternative new data or insights have not emerged sufficient to substantially alter the recommendations.

On the other hand, the present American Heart Association/American College of Cardiology recommendations do not strictly exclude in absolute terms fully informed athletes from participating in competitive athletic programs as long as such a decision is ultimately made in concert with their physician and third-party interests (e.g., high schools and colleges). Although this expert consensus report serves as a prudent guideline regarding sports eligibility or disqualification, there will always be tolerance in the system for some degree of flexibility, individual responsibility, and choice in making these decisions for individual student athlete-patients.

Genotype Positive-Phenotype Negative

An increasing number of HCM family members are recognized with documented pathogenic (disease-causing) sarcomere mutations, but in the absence of a clinical HCM phenotype (i.e., LV hypertrophy) (5,13). Such patients have been identified at a broad range of ages, although they are most commonly adolescents and young adults, and some wish to engage in competitive sports.

Spontaneous conversion to LV hypertrophy in this subset appears to occur most often in adolescence between 12 and 20 years of age (1,13) but has also been observed in midlife and beyond (14,15). Nevertheless, such changes are unpredictable, and some genetically affected people will probably never develop the HCM phenotype. Spontaneous morphological conversions are not usually accompanied by cardiac symptoms, disease progression, or events (1,2,13-15). However, once LV hypertrophy

evolves, that person may theoretically be subject to an unstable HCM electrophysiological substrate.

With negative or ambiguous genetic test results, potentially affected relatives can nevertheless be suspected clinically by the presence of several echocardiographic or cardiovascular magnetic resonance (CMR) findings in the nonhypertrophied myocardium, that is, blood-filled crypts, elongated mitral valve leaflets, diastolic dysfunction, and myocardial scarring (5,16-19). At present, the risk for sudden death in gene-positive-phenotype-negative family members appears to be extremely low and likely no different from the risk in the general population of the same age without heart disease (5,20). CMR imaging is also an important consideration in family members who are gene positive and judged to be phenotype negative based on echocardiography, because areas of segmental LV hypertrophy may be detected only by CMR, particularly in the anterolateral free wall and apex (6,21).

Recommendations

- 1. Participation in competitive athletics for asymptomatic, genotype-positive HCM patients without evidence of LV hypertrophy by 2-dimensional echocardiography and CMR is reasonable, particularly in the absence of a family history of HCM-related sudden death (Class IIa; Level of Evidence C).**
- 2. Athletes with a probable or unequivocal clinical expression and diagnosis of HCM (ie, with the disease phenotype of LV hypertrophy) should not participate in most competitive sports, with the exception of those of low intensity (class IA sports) (see “Classification of Sport” [22]). This recommendation is independent of age, sex, magnitude of LV hypertrophy, particular sarcomere mutation, presence or absence of LV outflow obstruction (at rest or with physiological exercise), absence of prior cardiac symptoms, presence or absence of late gadolinium enhancement (fibrosis) on CMR, and whether major interventions such as surgical myectomy or alcohol ablation have been performed previously (Class III; Level of Evidence C).**
- 3. Pharmacological agents (e.g., β -blockers) to control cardiac-related symptoms or ventricular tachyarrhythmias should not be administered for the sole purpose of permitting participation in high-intensity sports. Notably, such drugs may also be inconsistent with maximal physical performance in most sports (Class III; Level of Evidence C).**
- 4. Prophylactic ICDs should not be placed in athlete-patients with HCM for the sole or primary purpose of permitting participation in high-intensity sports competition because of the possibility of device-related complications. ICD indications for competitive athletes with HCM should not differ from those in nonathlete patients with HCM (Class III; Level of Evidence B).**

Other recommendations for sports participation in patients with HCM and ICDs can be found in the Task Force 9 report on “Arrhythmias and Conduction Defects” (23).

LV NONCOMPACTION

LV noncompaction (LVNC) is an uncommon and recently recognized cardiac disease with sporadic or familial occurrence (24). Its true incidence and prevalence are not known, in part because of difficulty in making the diagnosis and lack of agreement on criteria, as well as its heterogeneous clinical spectrum and usual requirement of CMR for reliable diagnosis. Furthermore, its clinical presentation and implications differ with respect to genetic pathogenesis, race/ethnic origin, presence in isolation or in association with other diseases, or depending on the presence or absence of right ventricular involvement (25).

The natural history of LVNC remains incompletely resolved because of its relatively recent recognition with a short available follow-up period (26-34). The clinical expression of LVNC is variable, even within families: with or without symptoms, heart failure, atrial and ventricular arrhythmias or preexcitatory pathways, thromboembolic events, or sudden death (35). While LVNC patients with heart failure and systolic dysfunction, thromboembolic events, and sudden cardiac death have been reported (26,31,33,34), many uncomplicated cases are less likely to be recognized or appear in the literature (34). Risk for adverse consequences, including mortality, presently appear to be largely associated with LV systolic dysfunction or ventricular tachyarrhythmias (34).

Few competitive athletes with LVNC have been reported clinically, and therefore, the consequences of LVNC in this specific population are unknown. Furthermore, to date, forensic registries of sudden deaths in young athletes do not include LVNC as a cause (3), although the diagnosis may still be widely underappreciated in the routine medical examiner autopsy setting. Therefore, given the lack of long-term follow-up studies and other obstacles, it is not yet possible to reliably apply risk-stratification strategies to new patients (or athletes) with LVNC. This is not unlike the situation with other uncommon myocardial diseases for which few data concerning sudden death risk during competitive sports are available (e.g., dilated cardiomyopathy [DCM] or infiltrative diseases). Therefore, the complete natural history of noncompacted ventricular myocardium remains unresolved.

A variety of inheritance patterns have been reported (ie, autosomal dominant, autosomal recessive, and X-linked) (24,27). Mutations in genes encoding sarcomeric proteins, which previously have been implicated in the

pathogenesis of HCM and DCM, have also been identified in patients with isolated LVNC (24,27). These observations suggest that LVNC shares genetic overlap with other cardiomyopathies, and indeed, some individual patients have been reported with morphological features consistent with both HCM and LVNC (32).

LVNC is thought to be caused by the intrauterine arrest of the compaction process of the primordial embryonic myocardium. Diagnosis is considered in the presence of a 2-layered LV chamber that consists of noncompacted trabeculations with intertrabecular recesses layered on top of the typical compacted myocardium, with or without systolic dysfunction. The trabeculated layer is predominantly confined to the distal and mid portions of the LV chamber, sparing the base. Currently, there are no universally accepted criteria or guidelines for the morphological diagnosis of LVNC, although a ratio of noncompacted to compacted myocardium $>2.1:1$ at end systole (echocardiography) or $>2.3:1$ in end diastole (CMR) have been proposed (30,32,36). It is uncertain how athletic training may alter those definitions (28,29) or the frequency of LVNC-appearing morphology in a normal athlete population. CMR is generally superior to echocardiography for identification of regions of non-compacted myocardium and for more definitive diagnosis of LVNC.

Recommendations

- 1. Until more clinical information is available, participation in competitive sports may be considered for asymptomatic patients with a diagnosis of LVNC and normal systolic function, without important ventricular tachyarrhythmias on ambulatory monitoring or exercise testing, and specifically with no prior history of unexplained syncope (Class IIb; Level of Evidence C).**
- 2. Athletes with an unequivocal diagnosis of LVNC and impaired systolic function or important atrial or ventricular tachyarrhythmias on ambulatory monitoring or exercise testing (or with a history of syncope) should not participate in competitive sports, with the possible exception of low-intensity class 1A sports, at least until more clinical information is available (Class III; Level of Evidence C).**

OTHER MYOCARDIAL DISEASES

A number of other uncommon diseases of the myocardium deserve consideration as potential causes of sudden death in athletes. These include DCM (attributable to a variety of causes, including genetic), primary non-hypertrophied restrictive cardiomyopathy, and systemic infiltrative diseases with secondary cardiac involvement, such as sarcoidosis. Few data are available at present

regarding the relative risks of athletic training and competition in athletes with these myocardial diseases.

It is important to differentiate physiological LV enlargement caused by systematic training from pathological DCM. Long-term aerobic athletic training can lead to cardiac morphological changes, including increased LV cavity dimension and calculated mass. Increased cavity size can produce a higher stroke volume, and thus, the ejection fraction at rest may be in the low-normal to mildly reduced range. Up to 15% of trained athletes will have substantial enlargement of the LV cavity, with end-diastolic dimensions up to 70 mm in men and 66 mm in women (37,38). Ejection fraction in trained athletes has been shown to be as low as 45% (37). Whether newer imaging techniques such as myocardial Doppler tissue imaging, strain imaging, or contrast-CMR scanning can differentiate patients with borderline LV enlargement and low-normal or mildly reduced ejection fraction from DCM is unresolved.

It is unclear whether asymptomatic patients with DCM are at risk for sudden death during competitive athletics, because ventricular tachyarrhythmias are most common in patients with more advanced disease, that is, with cardiac symptoms and lower ejection fraction.

Recommendations

- 1. Symptomatic athletes with DCM, primary non-hypertrophied restrictive cardiomyopathy, and infiltrative cardiac myopathies should not participate in most competitive sports, with the possible exception of low-intensity (class 1A sports) in selected cases, at least until more information is available (Class III; Level of Evidence C).**

MYOCARDITIS

General Considerations

Myocarditis commonly presents with disproportionate dyspnea on exertion, chest pain, and arrhythmias. It can also present as an acute myocardial infarction-like syndrome with sudden death in the presence of normal epicardial coronary arteries (39-44). The contribution of myocarditis to cardiovascular sudden death varies significantly with age, causing cardiovascular sudden death in $\approx 2\%$ of infants, 5% of children, and 4% to 7.5% of athletes (3,40). Higher rates of myocarditis are occasionally reported in postmortem studies from general populations younger than 35 to 40 years of age (41). Most cardiovascular sudden deaths attributable to myocarditis occur in males (42), and in some cases, myocarditis results in sudden death without antecedent symptoms or macroscopic cardiac abnormalities (40,42,43).

The data linking myocarditis to sudden death are strong and include autopsy studies and experimental myocarditis

models. For example, strenuous physical exertion was associated with sudden death in a cohort of U.S. military recruits, with the most frequent underlying cause being myocarditis (44). Case series of sudden death in athletes have established myocarditis as a significant risk in this specific group (3). In a murine model of coxsackie B3 myocarditis, 60 minutes of swimming daily increased viral titers, worsened cardiomyopathy, and increased the likelihood of death (45). In a chronic autoimmune myocarditis model, humeral and cellular immunity directed against heart tissues increased with treadmill exercise (46). Unlike heart failure, the risk of sudden death caused by myocarditis does not appear to correlate with the severity of myocardial inflammation (40). Sudden death has been observed occasionally after myopericarditis in association with normal LV function (47-49).

The pathogenesis of myocarditis consists of 3 overlapping phases: acute injury, often caused by a virus; the host innate and acquired immunologic response; and finally, recovery or a transition to scar and DCM. There is rarely a clear distinction between these phases clinically. The initial injury may cause an acute DCM with contractile impairment mediated by cytokines generated by the local inflammatory process. Several months later, the same dilated ventricle may have poor contractility caused by diffuse scar, with little or no inflammation. The transition from acute myocarditis to chronic DCM probably occurs over months, with substantial individual variability (50).

In clinical practice, myocarditis is often suspected but infrequently confirmed by endomyocardial biopsy, which creates a need for noninvasive diagnostic criteria to guide recommendations for athletic participation. For the purposes of this document, probable acute myocarditis is diagnosed when both of the following criteria are met:

1. A clinical syndrome that includes acute heart failure, angina-type chest pain, or myopericarditis of <3 months' duration.
2. An otherwise unexplained elevation in serum troponin; electrocardiographic features of cardiac ischemia; otherwise unexplained high-degree AV block or arrhythmias; wall motion abnormalities; pericardial effusion on echocardiography or CMR imaging. Additional CMR findings that suggest myocarditis in the acute clinical setting include characteristic alterations in tissue signal on T2- or T1-weighted images and the presence of late gadolinium enhancement (LGE).

CMR features that may be used to diagnose probable myocarditis include a regional increase in water content visible on T2-weighted images, an increase in regional contrast-enhanced T1-weighted epicardial or mid-myocardial signal obtained within a few minutes of the gadolinium bolus ("hyperemia" or "early-enhancement"

sequences), and epicardial or midmyocardial LGE (51). A regional and reversible increase in wall thickness that indicates myocardial edema is a supportive finding of acute myocarditis. Myocardial fibrosis, the late sequelae of myocarditis characteristic of DCM, may be indistinguishable from active myocarditis on LGE sequences. The sensitivity of CMR for myocarditis also decreases a few weeks after the initial illness (51).

Although acute myocarditis is associated with the characteristic findings of myocardial injury described in the diagnostic criteria above, there is no sensitive or specific test that can determine when the inflammatory process ends. DCM associated with acute myocarditis often resolves over 6 to 12 months. Athletes in whom the findings of acute inflammation have resolved may still have a risk of arrhythmias related to the resultant myocardial scar. The presence of LGE may convey a heightened risk for arrhythmias (52). The interval between initial assessment and retesting before resumption of sports will vary depending on the severity of the initial illness. A reasonable minimum interval for retesting based on experimental models is 3 to 6 months. The recommendations presented here recognize these gaps in knowledge and the need for additional clinical research to refine risk stratification for sudden death after acute myocarditis.

A diagnosis of myocarditis by biopsy is usually not required to guide clinical management, but a biopsy may be considered in select cases according to current professional society recommendations from the American Heart Association, American College of Cardiology, and European Society of Cardiology (53). Confirmation of myocarditis by endomyocardial biopsy creates a definitive diagnosis.

Recommendations

1. **Before returning to competitive sports, athletes who initially present with an acute clinical syndrome consistent with myocarditis should undergo a resting echocardiogram, 24-hour Holter monitoring, and an exercise ECG no less than 3 to 6 months after the initial illness (Class I; Level of Evidence C).**
2. **It is reasonable that athletes resume training and competition if all of the following criteria are met (Class IIa; Level of Evidence C):**
 - a. **Ventricular systolic function has returned to the normal range.**
 - b. **Serum markers of myocardial injury, inflammation, and heart failure have normalized.**
 - c. **Clinically relevant arrhythmias such as frequent or complex repetitive forms of ventricular or supraventricular ectopic activity are absent on Holter monitor and graded exercise ECGs.**

At present, it is unresolved whether resolution of myocarditis-related LGE should be required to permit return to competitive sports.

- 3. Athletes with probable or definite myocarditis should not participate in competitive sports while active inflammation is present. This recommendation is independent of age, gender, and LV function (Class III; Level of Evidence C).**

ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a cause of sudden death in young people and athletes, particularly in the northeastern (Veneto) region of Italy (54), but is seemingly less common in the United States (3). ARVC is characterized by a broad phenotypic spectrum and characteristically by loss of myocytes in the right ventricular myocardium, with fatty or fibrofatty replacement, which results in segmental or diffuse wall thinning, but there is also frequent involvement of the LV and an association with myocarditis (55). Genetics studies have demonstrated that ARVC is a desmosomal cardiomyopathy that results from genetically defective cell-adhesion proteins such as plakoglobin, plakophilin-2, desmoplakin, desmocollin-2, and desmoglein-2 (56,57).

Clinical diagnosis can be challenging but relies largely on familial occurrence, left bundle-branch pattern ventricular tachyarrhythmias, ECG findings of T-wave inversion in precordial leads V₁ through V₃, and epsilon waves, as well as right ventricular dilation or segmental wall motion abnormalities, aneurysm formation, or fatty deposition in the right ventricular wall identified with CMR imaging if substantial and unequivocal (or by biopsy tissue analysis). Diagnostic criteria for ARVC have been revised and updated and now include quantitative variables (58).

These criteria include global or regional structural dysfunction, as documented by echocardiography or CMR, biopsy abnormalities, ECG repolarization or depolarization abnormalities, arrhythmias, and family history. Each of these criteria is separated into major and minor criteria based on the severity of the finding. Patients meet an ARVC diagnosis if they possess 2 major, or 1 major and 2 minor, or 4 minor criteria. Borderline patients are those with 1 major and 1 minor criterion or 3 minor criteria. Patients with possible ARVC have 1 major criterion or 2 minor criteria. Athletes with borderline or possible ARVC, as well as those who are genotype positive-phenotype negative, should receive continued follow-up, because ARVC may progress phenotypically, and become more clinically apparent with time.

There is evidence in the experimental murine model that exercise increases the penetrance and arrhythmic risk in mutational carriers of ARVC (59). More recently,

these data have been confirmed in genetically positive patients (60), which is particularly relevant to the athlete, raising concern not only with regard to competitive sports but also regarding participation in moderate to extreme recreational physical activities.

Ventricular tachyarrhythmias and sudden death in ARVC commonly occur during exertion, including competitive sports (55,60,61), and frequent endurance exercise increases the risk for ventricular tachycardia/ventricular fibrillation and heart failure (60). However, risk factors for sudden cardiac death in ARVC are not as well defined as in HCM (1,2,7,8). There is general agreement that a prior history of sudden cardiac death, sustained ventricular tachycardia, or syncope represent the most important prognostic factors and define many high-risk patients who are most appropriately treated with a primary prevention ICD (62-64).

Recommendations

- 1. Athletes with a definite diagnosis of ARVC should not participate in most competitive sports, with the possible exception of low-intensity class 1A sports (Class III; Level of Evidence C).**
- 2. Athletes with a borderline diagnosis of ARVC should not participate in most competitive sports, with the possible exception of low-intensity class 1A sports (Class III; Level of Evidence C).**
- 3. Athletes with a possible diagnosis of ARVC should not participate in most competitive sports, with the possible exception of low-intensity class 1A sports (Class III; Level of Evidence C).**
- 4. Prophylactic ICD placement in athlete-patients with ARVC for the sole or primary purpose of permitting participation in high-intensity sports competition is not recommended because of the possibility of device-related complications (Class III; Level of Evidence C).**

Other recommendations for sports participation in patients with ARVC and ICDs can be found in the Task Force 9 report on "Arrhythmias and Conduction Defects" (23).

PERICARDITIS

The causes of pericarditis/myopericarditis are varied and are either infectious or noninfectious. The natural history is incompletely resolved, although long-term prognosis is generally favorable. The diagnosis of acute pericarditis is typically based on clinical criteria: chest pain, pericardial rub, ST-segment elevation, or new/worsening pericardial effusion. This syndrome may be considered part of the clinical spectrum of myocarditis. Recurrences are a significant consideration, and follow-up surveillance with echocardiography or CMR is recommended to exclude pericardial thickening or restriction consistent with restrictive pericarditis (50).

Recommendations

1. Athletes with pericarditis, regardless of its pathogenesis, should not participate in competitive sports during the acute phase. Such athletes can return to full activity when there is complete absence of evidence for active disease, including effusion by echocardiography, and when serum

markers of inflammation have normalized. For pericarditis associated with evidence of myocardial involvement, eligibility should also be based on the course of myocarditis. Chronic pericardial disease that results in constriction disqualifies the person from all competitive sports (Class III; Level of Evidence C).

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*Modest.

†Significant.

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