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Review

The Safety of Exercise in Individuals With Cardiomyopathy

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ABSTRACT

The cardiomyopathies are a heterogeneous group of primary myocardial diseases characterized by a propensity to fatal arrhythmias and are the leading cause of sudden cardiac death in young athletes. Apart from the underlying pathologic substrate, a combination of neurohormonal, mechanical, and oxidative stressors; dehydration; electrolyte abnormalities; and acid-base disturbances may trigger fatal arrhythmias during intensive exercise. Current consensus–based documents recommend that affected athletes abstain from most competitive sports, with the exception of those involving minimal dynamic or static components, to minimize the risk of sudden cardiac death. This article aims to describe the rationale underlying current recommendations and provides guidance for recreational exercise in many asymptomatic individuals. The article concludes with pragmatic recommendations for symptomatic patients with cardiomyopathy in whom physical activity is associated with beneficial effects on the quality and, possibly, the quantity of life.

RÉSUMÉ

Les cardiomyopathies sont un groupe hétérogène de troubles myocardiques primaires qui se caractérisent par une propension aux arythmies mortelles. Elles constituent la principale cause de mort subite d'origine cardiaque chez les jeunes athlètes. Mis à part le substrat cardiaque pathologique sous-jacent, la combinaison de divers facteurs de stress neurohormonaux, mécaniques et oxydatifs, la déshydratation, les anomalies électrolytiques et les perturbations de l'équilibre acide-base peuvent déclencher une arythmie mortelle pendant l'exercice intensif. Afin de réduire au minimum le risque de mort subite d'origine cardiaque, les recommandations consensuelles actuelles indiquent que les athlètes touchés doivent éviter la plupart des sports de compétition, à l'exception de ceux qui comportent des composantes dynamiques ou statiques minimes. Cet article explique les raisons motivant ces recommandations et fournit des directives en vue de la pratique d'activités sportives récréatives chez les nombreuses personnes asymptomatiques. L'article se termine par une série de recommandations pragmatiques à l'intention des personnes atteintes d'une cardiomyopathie symptomatique pour qui l'activité physique est susceptible d'améliorer la qualité et, possiblement, la durée de vie.

The cardiovascular benefits of moderate exercise are well established, and individuals who exercise regularly for 10-20 metabolic equivalent hours per week have a longer life span than do sedentary individuals. This equates to jogging at a pace of 10-15 minutes per mile for a cumulative period of 2 hours per week.[1,2] However, intense exercise may occasionally trigger arrhythmogenic sudden cardiac death (SCD) in athletes with cardiac disease. Indeed, 70%-80% of all non-traumatic SCDs in young athletes are attributable to inherited or congenital abnormalities of the heart, and sudden death is the sentinel event in up to 80% of cases.[3]

The primary cardiomyopathies are a heterogeneous group of largely inherited diseases of the myocardium that are associated with mechanical or electrical dysfunction of the heart, or both, and are most commonly implicated in SCD in young athletes.[4] Apart from the underlying pathologic substrate, a combination of neurohormonal and oxidative stressors, increased wall stress, dehydration, electrolyte abnormalities, and acid-base disturbances may trigger fatal arrhythmias during sports activities in such athletes.

Both the American Heart Association (AHA) and the European Society of Cardiology (ESC) advocate cardiac screening for the detection of young athletes with potentially serious diseases on the premise that the natural history of these diseases can be modified using various strategies, ranging from life style modification to the implantation of a cardioverter defibrillator.[5,6] Differences in screening methods and the concerns associated with such practice are beyond the scope of this article; however, most asymptomatic athletes are detected by preparticipation screening or through familial evaluation after a diagnosis of cardiomyopathy in a first-degree relative. The diagnosis of cardiomyopathy has major implications for an athlete given the association with SCD, particularly among adolescents and young adults, who are generally the most vulnerable group. Consensus-based recommendations have been developed to clarify important

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issues relating to sport eligibility of athletes with cardiac conditions and to aid the decision-making process for physicians in often challenging circumstances. The AHA/ACC (replacing the former 36th Bethesda recommendations) and the 2005 ESC recommendations advocate that athletes with a definitive diagnosis of cardiomyopathy should abstain from most competitive sports (Table 1), with the exception of a limited number of sports involving a low dynamic and static component of exertion (Fig. 1).[7-10] These recommendations are controversial and open to ethical questions, because there is insufficient scientific evidence regarding the consequences of regular exercise training and sports activities on the pathophysiology and clinical course of cardiomyopathies. The link between SCD and sports is based on circumstantial evidence, and many individuals with cardiomyopathy, particularly hypertrophic cardiomyopathy (HCM), have a good prognosis and normal life span.[11] Data from screening studies demonstrate that 1 in 300 asymptomatic athletes have cardiac conditions with the potential of triggering SCD.[12] The low incidence of SCD in athletes suggests that exercise rarely has a negative impact on the overall prognosis in most individuals who exercise. Furthermore, in our experience, many athletes diagnosed with cardiomyopathy continue to compete against medical advice and remain free of adverse cardiac events. Nevertheless, the unpredictability of sudden death from cardiomyopathy during exercise and the visibility afforded by these tragedies commands conservative recommendations. The ultimate aim is to encompass all preventable deaths.

Table 1. Recommendations for sport eligibility

Variable	2005 ESC recommendations[9]	AHA/ACC[7]
HCM	No competitive sports	No competitive sports, with the possible exception of those of low intensity
G^+P^- HCM	No competitive sports	All sports permitted
ARVC	No competitive sports	No competitive sports, with the possible exception of those of low intensity
G^+P^- ARVC	No competitive sports	No data
DCM	No competitive sports	No competitive sports, with the possible exception of those of low intensity
LVNC	No data	No competitive sports, with the possible exception of those of low intensity
Restrictive cardiomyopathy	No data	No competitive sports, with the possible exception of those of low intensity
Athletes with ICDs	Low-moderate dynamic and low static sports if asymptomatic and no discharges. Avoid collision sports.	No competitive sports, with the possible exception of those of low intensity. Avoid collision sports.

ARVC, arrhythmogenic right ventricular cardiomyopathy; DCM, dilated cardiomyopathy; G^+P^-, genotype positive/phenotype negative; HCM, hypertrophic cardiomyopathy; ICDs, implantable cardioverter defibrillators; LVNC, left ventricular noncompaction.

Although risk stratification is possible in patients with cardiomyopathy, these methods are far from perfect, and it is generally difficult to extrapolate assessment in a clinical laboratory to the athletic arena. Similarly, although several life style measures, pharmacologic treatment, and an implantable cardiac defibrillator may modify the clinical course of the cardiomyopathies in relatively sedentary patients, their effect on individuals who push physical boundaries on a day-to-day basis is unclear.

This article provides an overview on the safety of sports and exercise in athletes with cardiomyopathies, with particular focus on the risk of SCD. Because of their common implication in SCD, the authors will reference HCM and arrhythmogenic right ventricular cardiomyopathy (ARVC) to illustrate current recommendations for athletes affected with cardiomyopathies. Although there are very limited data on other cardiomyopathies in the context of sports, these recommendations also intuitively apply to athletes with a definitive diagnosis of dilated cardiomyopathy (DCM), restrictive cardiomyopathy, and other unclassified cardiomyopathies such as left ventricular noncompaction.

Exercise-Related SCD From Cardiomyopathy

The cardiomyopathies are the commonest cause of SCD in young athletes.[13] The risk of SCD from cardiomyopathy among young competitive athletes is almost 3-fold greater than in their relatively sedentary counterparts.[14] HCM is considered the commonest cause of SCD in athletes worldwide. The US National Registry of Sudden Death in Athletes, which is a database derived from a review of public records, reports that HCM accounts for almost 40% of all SCDs in young athletes.[3] In contrast, data from the Veneto region (Italy) pathology registry suggest that ARVC is the commonest cause of death and, HCM accounts for only 2% of deaths in athletes.[14] The discrepancy between the 2 countries may reflect the fact that the Italians have a nationally sponsored electrocardiographic (ECG) screening program for athletes that is effective for detecting most athletes with HCM through an abnormal electrocardiogram, whereas the diagnosis of ARVC is more elusive. Recently, Harmon et al.[15] reported that HCM accounted for only 8% of all deaths in athletes affiliated with the National Collegiate Athletic Association, suggesting that the risk of SCD during sports from HCM may have been overestimated. Most deaths occur in male athletes who compete in sporting disciplines associated with intermittent explosive bursts of exercise such as basketball and soccer. Data from the United States suggests that athletes of African/Afro-Caribbean origin may be at higher risk of SCD from HCM compared with white athletes.[3] Data from Italy and our own institution indicates that there is a particularly strong association between SCD during competitive sport and ARVC. Thiene et al.[16] observed that among 60 consecutive patients presenting with SCD, ARVC was diagnosed at postmortem examination in 12 patients, the majority of whom (10 [83%]) died during exertion. Recent findings from > 300 cases of SCD among athletes showed a 6-fold risk of dying suddenly from ARVC during intensive exercise compared with during rest or sleep.[17]