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Review article

Hypertrophic cardiomyopathy and sports



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ABSTRACT

This work summarizes current knowledge and recommendations regarding sports activities of patients with hypertrophic cardiomyopathy. Competitive sports, sports with high intensity level and burst exertion are not recommended. Also performing sports in extremely adverse environmental conditions as hot or cold weather, or high humidity is strongly discouraged; dehydration represents a risk factor as well. Recreational sports are permitted, particularly when individual risk factors and level of outflow tract obstruction are assessed in an individual patient.

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Although hypertrophic cardiomyopathy (HCM) is a disease, which is known almost 60 years, change of opinions on the condition has occurred many times in history, and sometimes there has been 180-degree turn around. The most fundamental change concerned the prognosis of HCM. Until recently, it has been generally accepted that the prognosis of HCM is utterly unfavorable, and patients are at any time in danger of sudden cardiac death, which affects most of them with annual mortality range from 2% to 4% in adults and 6% in children. Majority of deaths have been supposed to be sudden [1–5].

Contemporary view is different. Prognosis of HCM seems to be a quite good in low-risk patients with annual mortality not exceeding 1% in last 40 years [6–8]. Only half of deaths have been sudden, and remaining deaths have been caused by heart failure or stroke [6,7]. Lower mortality in these patients cannot be explained by the positive impact of treatment strategies (except for implantable cardioverter-defibrillator implantations in last years), but rather by more sensitive diagnostic

algorithms enabling to identify mild (or even borderline) forms of the disease, which has not been formerly detectable. Well known risk factors are associated with risk of sudden death, young people with early phenotype of HCM have also serious prognosis [7]. Severe left ventricular outflow tract obstruction ($V_{\max} > 4$ m/s) can deteriorate the prognosis of HCM, and is more often associated with development of heart failure [8].

Current opinion of pregnancy in patients with HCM has changed dramatically. Pregnancy in women with HCM has been strongly discouraged in past [1,2], but nowadays, it is considered safe and repeatable [9].

On the other hand, it would be incorrect to become excessively optimistic because HCM takes its death toll, particularly in young people. For example, in children aged 6–18 years, HCM is the main cause of sudden deaths together with arrhythmogenic right ventricular cardiomyopathy [10].

Initially, patients with HCM have been strongly advised against engaging in any sports activities [1–4]. Do our today's

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thinking and recommendations regarding sports in HCM reflect current opinions on the HCM prognosis? We would like to discuss this question in detail on following lines.

Even for non-professionals, it is evident while reading and listening mass media and other sources that HCM can cause death in the not negligible portion of young athletes. Recently, particularly two tragic deaths have been made public: the sudden death of a 19-year old Canadian hockey player of Windsor Spitfires junior hockey team (Ontario Hockey League) and also the death of another hockey player of same age – former Jagr's teammate from Avangard Omsk. Both men died from sudden death (the second one straight on the hockey rink) after an unsuccessful resuscitation for cardiac arrest. In both men, sudden death was caused by HCM. Naturally, such events occur more often, but we don't know accurate numbers of such cases.

Older works show unequivocally that HCM is the most common cause of sudden death in young athletes. HCM accounts for one third to one half of sudden death cases [11–13]. Also, the last work published on the topic of sudden death in young athletes clearly demonstrates [14] that HCM is the predominant cause of this condition (Table 1). Forty seven cases out of 182 cases of sudden death in young athletes reported in 10 years had been confirmed to be of cardiovascular cause. Out of them almost one half (21 students) died from HCM. The most frequent sports presented in this report were basketball (23 students) and soccer (16 students). Twenty nine students died while engaged in competitive sports, and 14 students were engaged in recreational sports.

Of note, other work has dealt with sudden death in marathon runners [15]. In total 10.9 million runners registered between years 2000–2010, and 59 of them (aged 42 ± 13 years, 51 men) experienced cardiac arrests. Vast majority of arrests was caused by HCM plus coronary artery disease. The strongest predictor of survival was except for early cardiopulmonary resuscitation also other etiology than HCM, which certainly suggested an important role of this disease in the genesis of cardiac arrest resulting in sudden death in marathon runners.

Thus, HCM is the most common cause of sudden deaths in young people. The patients who can be very old, and have almost no symptoms of the disease, and usually die from other

(mostly non-cardiac) causes, are on the other end of the age spectrum of patients with HCM. Such findings have been demonstrated by the data from Maron's database of 1297 patients with HCM [16] comprising 26 patients (2%) older than 90 years (the oldest patient was almost 97-years old) who were mostly women (69%), which is common in similar cases. In young patients with HCM, however, the situation is rather different; prognosis of the disease is significantly unfavorable in this population. Nevertheless, the older patients are not usually engaged in sports activities.

The view on sports activities in HCM has changed step by step. Any physical activity has been strongly discouraged in older works [1–4], but over the years, more detailed recommendations have been created and published by American College of Cardiology in years 1985, 2005, and 2008 [17–19]. In 2005, also the statement of European Society of Cardiology has been released [20]. It is understandable that the detailed guidelines could have not been created and based solely on evidence-based medicine because the appropriate data covering all aspects of this topic are not available. Thus, the recommendations have been based mostly on consensus of experts.

So what about sports activities and HCM? Competitive sports with high level of physical intensity represent high risk [8–21]. Also sports with burst exertion (running and sports associated with it as soccer, basketball and other sports) should be avoided. Aerobic (not isometric) exertion is preferred. Sports activities performed in extremely adverse environmental conditions (hot or cold weather, high humidity) are not advised. It is important to replenish fluids (dehydration presents very high risk in patients with HCM, and this is true not only during physical activity). The activities as using sauna or whirlpool are not suitable [22,23]. In recreational sports activities, recommendations should be assessed on an individual basis with respect to his/her risk factors, particularly ventricular arrhythmias [18] and left ventricular outflow tract obstruction [21]. On the other hand, it is important to emphasize that even patients with HCM without all “classical” risk factors are not spared of sudden death risk, especially while engaged in competitive sports with high level of physical intensity [19,24]. Undoubtedly, it is related to the fact that not all risk factors are known currently, and we are not able to identify reliably all endangered patients. One of such risk factor, in which we are not able to describe the level of risk and danger for their carriers, is the presence of extensive fibrosis, especially affecting papillary muscles, which could impact an arrhythmogenic substrate [25].

Table 2 presents the categorization of sports with regard to the intensity level. According to the guidelines of American professional societies [21], all sports are assigned grades indicating eligibility. The graded scale starts with sports generally not advised in HCM (0–1), and ends with sports admissible and probably permitted in HCM (4–5). There is continuous spectrum between those two poles (2–3). Table 2 shows that the sports with high intensity level usually cannot be advised, and on the contrary, the sports with low intensity level can be recommended except for diving.

The conference in Bethesda in 2008 [19] was also concerned with the problem of patients with the positive cardiomyopathy genotype (not only HCM, but also dilated cardiomyopathy,

Table 1 – Sudden death in young athletes with confirmed cardiovascular cause [14].

Cause	Number
HCM	21
Coronary artery anomaly	8
CAD	5
Aortic dissection	3
ARVC	3
Dilated cardiomyopathy	2
Myocarditis	2
Long QT syndrome	1
Mitral valve prolapse	1
Kawasaki syndrome	1

ARVC, arrhythmogenic right ventricular cardiomyopathy; CAD, coronary artery disease; HCM, hypertrophic cardiomyopathy.

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