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Hypertrophic Cardiomyopathy and Sudden Cardiac Death Due to Physical Exercise in Croatia in a 27-Year Period

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ABSTRACT

The paper deals with the sudden cardiac death during physical exercise in males in Croatia. The data are a part of a retrospective study dealing with 69 sudden death due to physical activity in men in Croatia during 27 years: from January 1, 1984 to December 31, 2010. Three of them suddenly died during training and two of them died during recreational physical exercise, probably because of malignant ventricular arrhythmia due to hypertrophic cardiomyopathy. One had an obstructive form of hypertrophic cardiomyopathy with i.v. septum of 40 mm and four had a non-obstructive forms of hypertrophic cardiomyopathy with left ventricular wall of 18–20–22–25 mm. First athlete was a short trails runner, aged 24, with no any previous physical discomforts, who suddenly collapsed and died during training. The second athlete was a soccer player aged 18, with no any previous physical discomfort, who suddenly collapsed and died during training. The third aged 15, was a school boy, basketball player, with no any previous physical discomfort, who collapsed and died during training. Two aged 25 and 34, were with no physical discomfort during exercise and died suddenly during recreational soccer games. A sudden cardiac death due to physical exercise in young athletes in Croatia suffered of hypertrophic cardiomyopathy reached 0.06/100 000 yearly ($p=0.00000$) in 27 years, in teenagers 0.26/100 000 ($p=0.00226$), in teenagers suffered of hypertrophic cardiomyopathy reached 0.10/100 000 ($p=0.00000$), in all young athletes suffered of other heart diseases reached 0.19/100 000 ($p=0.00005$), and in the total male population aged 15 or more, engaged in sports and recreational physical exercise: 0.71/100.0000 ($p=0.00001$).

Key words: athletes, training, recreational physical exercise, hypertrophic cardiomyopathy, sudden cardiac death

Introduction

In young persons who died suddenly due or immediately after physical exercise, the most common reasons are cardiovascular diseases^{1–8}. Hypertrophic cardiomyopathy (HCM) or asymmetrical septal hypertrophy or dynamic hypertrophic subaortic stenosis or hypertrophic obstructive cardiomyopathy or hypertrophic nonobstructive cardiomyopathy or idiopathic hypertrophic subaortic stenosis, is a most common cause of sudden cardiac death in young competitive athletes⁹. HCM means thickened (not dilated) the wall of the left ventricle, without any other disease leading to left ventricular hypertrophy (LVH): f.e. arterial hypertension, aortic stenosis (coarctation), aortic valvular stenosis etc. The etiology of HCM is unknown. For the first time it is described by Teare¹⁰

as asymmetrical hypertrophy of the heart. HCM is a primary myocardial diseases¹¹ with 450 different mutations identified in 13 genes, in which three genes are responsible for more than 50% of all genotype cases: cardiac troponin-T (chromosome 1), myosin-binding protein C (chromosome 11) and beta-myosin heavy chain (chromosome 14). In HCM, LVH is asymmetrical and could be concentric, localized in the basal anterior septum, posterior septum, anterolateral wall, posterior wall or in apex. The consequence of HCM could be sudden cardiac death due to decreases cardiac output leading to dyspnea during exercise, postural arterial hypotension, palpitations or syncope, myocardial ischemia with typical or atypical anginal chest pain during exercise and with a high risk of

ventricular instability and malignant ventricular arrhythmias. LVH increased the risk of sudden cardiac death by promoting subendocardial ischemia¹².

The aim of this study is to analyze the course of the illness in young men died suddenly because of HCM, during sport or recreational exercise in Croatia.

Sample and Methods

In a period of 27 years: from January 1, 1984 to December 31, 2010, a total number of 69 sudden and unexpected deaths due or immediately after sport or recreational exercise in persons of all ages in Croatia are registered. Six of them were athletes and 63 were engaged in physical exercise recreationally. The data are part of a retrospective study and are collected from the whole population consisted 4,500,000 persons. Three of athletes and two of persons engaged in recreational exercise suffered of HCM. The data were found from the registry of Services of Forensic Medicine, Public Health Registry and Sports clubs. The statistical difference was calculated using χ^2 -test and Poisson rates.

Results

Case 1

The first athlete, aged 29, was a former rugby player, and actually rugby referee. He had no symptoms of any heart disease. He had not slept at least 48 hours before the incident. At that very day in June 1998, he was swimming in the swimming pool (he was a good swimmer), he felt exhausted, stopped to swim for a few minutes, then swam again, felt increased tiredness, collapsed and died. All reanimation efforts were unsuccessful including a Mobile Emergency Medical Unit.

At the forensic autopsy he had hypertrophic cardiomyopathy: by the criteria of gross pathological changes of cardiomegaly, left ventricular hypertrophy: 25 mm, with normal cavities of the whole heart, normal coronary arteries, and pulmonary and cerebral edema.

Case 2

The second athlete, aged 15, was a school boy, basketball player, and referred no cardiovascular or other symptoms. One day in May 2000 during a game he felt exhausted, and was standing at the site, watching the others playing basketball. Then he resumed training, jumped suddenly, put the hand on his chest, collapsed and died. All reanimation attempts were unsuccessful including attempts at the Regional University Hospital.

The forensic autopsy finding showed hypertrophic cardiomyopathy with interventricular septum of 40 mm, left ventricular hypertrophy of 17 mm, right ventricular hypertrophy of 7 mm. The space of the left ventricle was narrowed because of the septal hypertrophy. He had an acute pulmonary and cerebral edema.

Case 3

The third athlete aged 18 was a school boy and professional soccer player. He did complaint of any discomfort during physical exercise. During a soccer game in May 2006 one player kicked his head with a ball. He felt general weakness with a short breath and he felt down. He was transferred to the nearest University Hospital where he have been cured because of bilateral bacterial pneumonia, and died two days after admittance in a state of a septic shock, in spite of all therapeutic efforts.

The forensic autopsy showed left ventricular wall reached 20 mm, and normal cavities of the whole heart, a large bilateral bacterial pneumonia, fibrinous pericarditis, cerebral contusions with edema and pointed bleeding.

Case 4

The fourth was a physician/cardiovascular pathologist aged 34, who played soccer twice a week recreationally. He was without any physical discomfort during physical exercise. One day in August 1984 in the evening during a soccer game, he did not feel well, went to a car and drove few hundred meters, than stopped, went from a car, collapsed and died. All reanimation efforts were unsuccessful, including reanimation attempts in a nearest hospital – Regional Army Hospital.

The forensic autopsy finding showed left ventricular hypertrophy of 18 mm, with no dilatation of the left ventricular cavity. Left descending coronary artery was narrowed. He had an acute pulmonary and cerebral edema.

Case 5

The fifth aged 25, played soccer recreationally. He was without any physical discomfort during physical exercise. One day in January 2005 in the evening during a game in a sport hall of a primary school, he suddenly collapsed and died. All reanimation attempts at the field were unsuccessful including a Mobile Emergency Medical Unit.

The forensic autopsy showed left ventricular hypertrophy of 22 mm and right ventricular hypertrophy of 7 mm, with normal cavities of the whole heart. The left and the right coronary arteries were narrowed at the ostial places to 1 mm (hypoplasia ostii arteriarum coronarium cordis). He had an broulosus emphysema of the left pulmonary wing. He had an acute pulmonary and cerebral edema.

The statistical difference of those data was calculated using Chi-square test and Poisson rates. A sudden cardiac death due to physical exercise in young athletes in Croatia suffered of hypertrophic cardiomyopathy reached 0.06/100 000 ($p=0.00000$) in 27 years, in teenagers 0.26/100 000 ($p=0.00226$), in teenagers suffered of hypertrophic cardiomyopathy reached 0.10/100 000 ($p=0.00000$), in all young athletes suffered of other heart diseases reached 0.19/100 000 ($p=0.00005$), and in the total male population aged 15 or more, engaged in sports and recreational physical exercise: 0.71/100.0000 ($p=0.00001$).

TABLE 1
CHARACTERISTICS OF FIVE MALES WHO DIED SUDDENLY DUE TO PHYSICAL EXERCISE BECAUSE OF HYPERTHROPIC
CARDIOMYOPATHY

Case	Age	Profession/physical exercise	Symptoms	Physical finding/ ECG	Lethal event	Resuscitation	Forensic autopsy
1	29	Student swimming/rugby referee	No	No data	June 1998 during swimming	Yes	LVH 25 mm normal cavities of the whole heart, normal coronary arteries, pulmonary and cerebral edema
2	15	High-school student/basketball	No	No data	May 2000 during a game	Yes	LH 17 mm, RVH 7 mm, IV septum 40 mm, narrowed cavities of the left ventricle, normal coronaries, myocardial fibrosis and scars up to 3 mm in diameters, pulmonary and cerebral edema
3	18	High-school student/professional soccer player	No	No data	May 2006 kicked in a chest during a game, died in a regional hospital 2 days after admittance	Yes	LVH 20 mm during normal cavities of the whole heart, normal coronary arteries, fibrinous pericarditis, bilateral broncho-pneumonia
4	34	Physician-pathologist, soccer recreationally	No	No data	August 1984 during a game	Yes	LVH 18 mm, normal cavities of the whole heart, LADCA narrowed, pulmonary and cerebral edema
5	25	Soccer recreationally	No	No data	June 2002 during a game	Yes	LVH 22 mm, ostium of the RCA narrowed to 1 mm, malposition of the ostium of the RCA, ostium of the LCA narrowed to 1 mm, normal coronary arteries, pulmonary edema

Legend: LVH=left ventricular hypertrophy; RVH=right ventricular hypertrophy; IV=interventricular; LCA=left coronary artery, RCA=right coronary artery, LADCA=left anterior descending coronary artery

Discussion

Health incidents due to sports or recreational activities are rare. In athletes who died suddenly because of malignant heart arrhythmias due to training, the most common reasons for such events are cardiomyopathies, coronary anomalies and myocarditis^{13,14}. Among cardiomyopathies, HCM reached 0.2% of population¹⁵. HCM has variable thickening of the septum and the left ventricular wall and different clinical presentations. It could be obstructive forms of HCM (in about 30%) and non-obstructive forms (in about 60%), and in additional 10%, obstruction could be presented due to vigorous physical exercise¹⁶. There is no strong correlation between the degree of HCM and subjective symptoms¹⁷. There are five risk factors for sudden death in persons suffering of HCM: family history of sudden death, syncope, an extreme LVH (>30 mm), non-sustained ventricular tachycardia in 24-hours ECG and abnormal blood pressure response to exercise^{18,19}.

It is known that a magnitude of left ventricular or septal hypertrophy leads to high risk for sudden cardiac death and is a strong independent predictor to prognosis. The risk of sudden cardiac death rised progressively with left ventricular wall thickness: if LVH is lesser than 19 mm, a risk for sudden death is low, but LVH 30 mm or more doubled the risk of sudden cardiac death¹⁷. In our

five presented cases of HCM, a heavy obstructive form of HCM was seen in one athlete, school boy aged 15, with no previous symptoms, who died suddenly: i.v. septum reached 40 mm. It confirms the rule that extreme hypertrophy are seen in the youngest with mild or no symptoms¹⁷. In other two athletes died suddenly, LVH reached 20 and 25 mm. In two of persons engaged in physical activities recreationally and died suddenly, LVH reached 18–22 mm, in spite of the opinions that in persons with mild or moderate LVH the risk is low.

In preparticipation screening for HCM, a familial history data have to be included especially to sudden cardiac death (it could be negative), personal symptoms including recurrent exertional syncope, physical examination by physician (it could be normal finding in most of these patients), an ECG (it could be non-specific), LVH by echocardiography (<19 mm = low risk, >30 mm = high risk), and 24 h ECG-Holter (malignant ventricular arrhythmias).

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HIPERTROFIJSKA KARDIOMIOPATIJA I NAGLA KARDIJALNA SMRT ZA VRIJEME TJELOVJEŽBE U HRVATSKOJ U 27-GODIŠNJEM RAZDOBLJU

SAŽETAK

Izneseni su podaci retrospektivnog istraživanja 69 naglih kardijalnih smrti za vrijeme tjelovježbe u muškaraca Hrvatskoj u 27-godišnjem razdoblju: od 1. siječnja 1984 do 31. prosinca 2010 g. Od toga su petorica preminuli vjerojatno zbog zloćudne aritmije klijetki tijekom hipertrofijske kardiomiopatije: tri sportaša za vrijeme treninga i dva za vrijeme rekreacijske tjelovježbe. Jedan je imao opstruktivni oblik hipertrofijske kardiomiopatije s debljinom i.v. septuma od 40 mm, ostali su imali neopstruktivni oblik s debljinom lijeve klijetke 18–20–22–25 mm. Prvi sportaš bio je trkač kratkih pruga, dobi 24 g., bez prethodnih zdravstvenih teškoća, koji je naglo kolabirao i preminuo za vrijeme treninga trčanja. Drugi i treći sportaš bili su košarkaši, dobi 18 i 15 g., oba bez prethodnih zdravstvenih teškoća, koji su kolabirali i preminuli za vrijeme treninga. Dvojica dobi 25 i 34 g. bavili su se nogometom rekreacijski, oba su bili bez zdravstvenih tegoba, oba su kolabirali i umrli za vrijeme nogometne igre. Nagla kardijalna smrt zbog hipertrofijske kardiomiopatije za vrijeme tjelovježbe u mladih sportaša u Hrvatskoj iznosi 0,06/100 000 godišnje ($p=0,00000$), u ostalih mladih sportaša koji su bolovali od drugih bolesti srca iznosi 0,19/100 000 ($p=0,00005$), a u svih muškaraca dobi 15–40 g. koji se bave sportskom ili rekreacijskom tjelovježbom iznosi 0,71/100 000 ($p=0,00001$).