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Coronary arteries anomalies (CAA) as a cause of a sudden cardiac death (SCD) among young athletes – review

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ABSTRACT

Introduction and objective

Congenital coronary arteries anomalies (CAA) are one of the most common causes of a sudden cardiac death (SCD) among young athletes. CAA are serious diagnostic and clinical issue because in many patients there are no characteristic clinical symptoms and commonly used tests are not enough to reveal many types of anatomical abnormalities. Authors of this review would

like to present a current state of knowledge of CAA and their diagnostics, as well as recommendations for proceeding in patients with diagnosed CAA to prevent SCD.

Abbreviated description of the state of knowledge

According to statistics CAA are rare congenital heart defects however they are associated with a more frequent occurrence of SCD. The mechanism of this phenomenon has not been fully explained but there are several hypotheses. The most important, of which concerns the dimension of the angle of branch of the coronary artery from the sinus of Valsalva. Numerous clinical studies on SCD pay attention to the occurrence of asymptomatic CAA as the main cause of SCD right after hypertrophic cardiomyopathy. Among strategies of proceeding recommended after the diagnosis of CAA, there are pharmacological and surgical treatment, however it has not been established which of them is the best for young athletes.

Summary

One of the most important things to prevent SCD in the course of CAA seems to be creating universal guidelines for early diagnosis and treatment of this congenital defect. Widespread learning first aid and increasing availability of automatic external defibrillators (AED) are also important. SCD of a young athlete is a tragedy not only for the family but also for the whole sporting environment, that is why it is in the public interest to find an answer to the question how we can prevent that cases in the future.

INTRODUCTION

The term “sudden cardiac death” (SCD) is already well known. It was first introduced by Kuller in 1966 but his definition has been modified over the years. SCD is recently known as acute coronary syndrome and stands for sudden death caused by various heart disorders. They can be preceded by the loss of consciousness of an apparently healthy person which took place within an hour before first visible symptoms [2,3].

Majority of the studies, which are available in the literature clearly show that physical activity significantly reduces the risk of cardiovascular diseases, diabetes, cancer as well as prolongs human life [4-8]. It is commonly believed that "sport is healthy” and sportspeople are considered to be “the healthiest group of people”. That is why the sudden death of a sportsman or sportswoman seems to be a tragedy not only for the relatives but also for friends, trainers, sport clubs and finally for fans because dies a young and athletic person who is considered to

be a symbol of health. Sudden death in this particular group always evokes strong emotions and leads to stormy discussions about harmful consequences of intensive sport.

Sudden deaths are more frequent in a group of sportspeople than in any other group. This frequency depends on the register and the kind of population which is studied. In 2003, Italian researchers published their results of the research which has been conducted for many years showing that the risk of sudden death of a young sportsman is 3,5 times higher than of a person who does not do any sport [9]. Moreover, mortality among sportspeople estimated on the basis of the American studies amounts to 1-2/100 000 sportspeople every year [10,11]. Sudden cardiac death is almost 10 times more frequent in a group of young men than in women engaged in sports such as basketball, football or running[12].

THE AIM OF THE STUDY

In this paper authors decided to highlight the most common causes of sudden death among young athletes caused by coronary artery anomalies. They would like to present the current state of knowledge concerning proper diagnostics and proper procedures in case of coronary anomalies as well as point out prescriptions after diagnosis.

CURRENT STATE OF KNOWLEDGE

The main causes of sudden cardiac death among athletes are presented in Table 1. These causes vary according to age. The age limit of the youngest group of athletes is no more than 35 years old. In this particular group, the preponderant cause of the SCD is genetically conditioned musculoskeletal disease, for instance, hypertrophic cardiomyopathy (HCM) as well as coronary artery anomalies which are not visible in people who do not do any sport. [13,14]

THE MAIN CAUSES OF SUDDEN CARDIAC DEATH AMONG ATHLETES	
CONGENITAL AND GENETIC	**ACQUIRED**
Hypertrophiccardiomyopathy(HCM)	Coronaryatherosclerosis
Arrhythmogenic right ventricular cardiomyopathy (ARVC)	Myocarditis
Othercardiomyopathies	Kawasaki disease
Congenital anomalies of the course of coronary arteries	Acquired long QT syndrome (e.g. drug-induced)
Aortic defects (e.g. aneurysm of the ascending aorta in Marfan syndrome)	Heartconcussion
Valvular defects (e.g. congenital aortic valve stenosis, mitral valve prolapse syndrome)	

Table 1.

One of the causes of sudden cardiac death among young athletes is coronary artery anomaly. In order to understand its effect on cardiac hemodynamics of heart, it is necessary to focus on some basic information concerning heart vascularization, which is described in normal anatomy.

The arterial vascularisation of heart comes from the coronary arteries that are combined with the ascending coronary aorta – sinus of Valsalva. In most cases, there are two main coronary arteries: right coronary artery (RCA) and left coronary artery (LCA). The left coronary artery typically comes from the left sinus of Valsalva, giving two branches of the artery on its course: circumflexartery (LCx) and left ascending artery (LAD). The LCA range includes 2/3 of the anterior ventricular septum, left atrial wall, left ventricle wall (excluding posterior wall) and anterior wall of right ventricles. The right coronary artery comes from sinus of Valsalva respectively, the main artery going from it is the posterior descending artery (PDA). The RCA is responsible for arterial vascularisation of 1/3 of the posterior septum, right atrial wall, right ventricular anterior wall and left ventricle posterior wall. It is clinically significant that the major part of the cardiac conduction system (excluding the anterior branch of the left atrioventricular fasciculus) is supplied with arterial blood by the RCA.

According to literature coronary arteries anomalies (CAA) are a rare congenital heart defect, which applies to 0,64-1,4% of population [19]. Nonetheless, it is thought that these statistics are not precise. It is because of the fact, that numerous patients with this anomaly has not been diagnosed with this defect due to none specific clinical symptoms.

There are many classifications of anomalies of coronary arteries. The most commonly used is a classification elaborated in 2003 by Rigatelli and his associates. A detailed division of congenital anomalies of coronary arteries based on this classification is shown in Table 2.

ANOMALOUS AORTIC ORIGIN OF A CORONARY ARTERY (AAOCA)
Hypoplasia/atresia
Single coronaryartery
Congenitalcoronaryarteriovenousfistula
The beginning of coronary artery starting in other coronary artery
Ectopic beginning of left or right artery from opposite coronary sinus (AAOCA)
Anomalous division of coronary arteries (individual beginnings for LAD, LCx and doubled LAD)
Intramuscularcourse

Table 2.

Basing on pathophysiology and symptomatology of all anomalies, CAA is classified as: anomalies with permanent ischaemia of myocardium which are observed mostly in case of anomalous left coronary artery from the pulmonary artery (ALCAPA) known as Bland-White-Garland syndrome; anomalies without ischaemia which represent the most of CAA cases - they have no clinical symptoms and intermittent anomalies which lead to ischaemia of myocardium with rare dangerous effects [15].

As literature sources convey, abnormalities connected with beginning of right coronary artery are 6 times more often than anomalies in left coronary artery. Nonetheless, left coronary artery anomalies cause up to 85% of cases of sudden cardiac death associated with CAA [15].

Although there is an evident connection between occurrence of CAA and SCD episodes, previous researches did not give an explanation on a mechanism leading to this dependence. There are few hypothesis, which can explain more frequent occurrence of sudden cardiac deaths among patients with CAA. In case when coronary artery begins in the opposite sinus of Valsalva, it often goes between aorta and pulmonary trunk. Intense physical exercises may lead to constriction of the coronary artery. Additionally, coronary arteries of patients with CAA often have a diverse intramural part inside of ascending aorta. Presumably, increase of blood pressure during physical exercises can cause a compression of intramural part of coronary artery. The angle of branch of coronary artery from sinus of Valsalva has also a significant impact on SCD frequency among patients with CAA. In case of proper anatomical structure of heart, coronary vessels and aortic bulb, the angle of coronary vessel branch is nearly 90°. In case of coexistence of AAOCA, the anomalous coronary artery begins with an acute angle. It is assumed that the more acute angle, the more abnormalities in blood flow occurs. Furthermore,

among patients with AAOCA the beginnings of coronary arteries have aperture form instead of circular/ oval. Therefore in case of strong adrenergic stimulation during physical exercises coronary arteries might get occluded.

The common feature of all foregoing abnormalities, is a decrease of a blood flow in coronary vessels evoked by intense workout. Depending on the artery, in which the flow was disturbed, various types of nonspecific symptoms are observed e.g. tachycardia, ventricular fibrillation or ischaemia of the myocardium, which is sometimes extensive enough to cause cardiogenic shock and subsequently death [18].

Although sudden cardiac death is not the main cause of death among young athletes (accidents remain to be the primary cause) [20, 21], it is a significant problem in sport. Anomalies usually emerge during physical activities [22]. All cases of sudden cardiac deaths evoke agitation among a lot of communities and activate people to attempt to prevent such cases in the future. That is why a lot of studies were performed to evaluate the etiology and the frequency of occurrence of sudden cardiac death among young athletes. Scientists are searching for efficacious diagnostic methods that would allow to define a risk group and prevent these people from sudden cardiac death.

Most of the scientific researches that evaluate the cause of sudden cardiac death are retrospective analyses which use data from electronic registers [22]. An example of such analysis is a demographic and epidemiological research performed by Maron and associates [23]. They used the data from US National Registry of Sudden Death in Athletes and distinguished a group of 842 people, at the age from 13 to 25. The causes of death among these people were cardiovascular system anomalies. Among 158 people anomalies in coronary arteries structure were detected. They represented 19% of the examined cases and were the second most common cause of death from cardiological reasons after hypertrophic cardiomyopathy. Interestingly, anomalies in coronary arteries occurred more often amidst females.

Peterson and associates [22] performed a retrospective analysis to evaluate the etiology of cardiac arrest and sudden cardiac death among competitive athletes in the age from 11 to 29 (mean average was 16,6). 16 out of 179 examined people had anomalies in the structure of coronary vessels, which lead to sudden cardiac death among13 people and cardiac arrest among 3 people. Anomalies of coronary arteries represented 13,7% of examined cases and they were also the second most common cause of sudden cardiac death after hypertrophic cardiomyopathy.

The main problem in prevention of cardiovascular system accidents is an asymptomatology in the course of disorders which are the main cause of sudden cardiac death. According to Maron and associates [24] 80% of patients have no symptoms until the accident occurs. Some patients with anomalies in the structure of coronary arteries have non-specific symptoms e.g. pain in the chest, tachycardia, dyspnea or syncope [18]. Standard medical examinations performed among young athletes which tests capacity and correctness of cardiovascular system often do not show abnormalities. It leads to a fact, that defining a risk group of sudden cardiac death is a serious diagnostic problem. Sometimes electrocardiography (ECG) can show alterations which signaliseischaemia or arrhythmia which are indications to extended diagnostics. It is recommended to perform transthoracic echocardiography which is a non-invasive, low-cost and secure type of examination [18,25]. To affirm the diagnosis and to define the exact course of coronary vessels it is suggested to perform a CTA (computed tomography angiography) or MRA (magnetic resonance angiography) [18,26]. If performed examinations cannot make a diagnosis based on 36th Bethesda Conference recommendations, coronarography should be done [27].

Currently there are no unambiguous recommendations that pertain to therapeutic proceeding among patients with diagnosed anomalies in coronary arteries structure. It is ascertained that surgical procedure should be offered to selected group of patients which fulfil criteria such as ischaemia symptoms, age below 30 years, ventricular arrhythmia or syncope [18]. There are no recommendations for patients that do not fulfil those requirements. There are performed a lot of researches on the application of beta blockers to prevent sudden cardiac death [18, 20]. A contentious issue is a limitation of physical activity. Recommendations suggest resigning from competitive sports in favour of less dynamic and low-intensity activities [20]. Importantly, regular physical activity brings lot of health benefits and is genuinely crucial in correct development and growth [18].

Cardiopulmonary resuscitation (CPR) skills among people and accessibility to an automated external defibrillator (AED) have also a significant impact in sudden cardiac death prevention [20, 28]. They can allow giving a first aid to a person with cardiac arrest until medical assistance arrives. Immediately proceeded actions should reduce death rate during cardiovascular accidents. To accomplish that first aid training courses should be performed in schools and work places and the awareness about first aid should be raised [20].

SUMMARY

Sudden cardiac death in group of young athletes is and probably will remain an important problem in medicine, especially in sport cardiology. The death of a young athlete, viewed as a model of physical strength and health, is always sensational. Despite the rapid progress of cardiological diagnostics and treatment options in recent years, some diseases still have too low detection rates and, consequently, lead to life-threatening effects.

Congenital coronary artery anomalies belong to rarely diagnosed cardiovascular defects. The majority of CAA is mild and asymptomatic, but selected entities could be the cause of life-threatening conditions, including sudden cardiac death. Clinical suspicion of aforementioned anomalies should be considered, especially in the group of young patients complaining of cardiac pain, dyspnea or syncopes repeated during or immediately after intense physical exercise. There are many scientific reports clearly indicating the pathogenetic association of SCD with coronary artery abnormalities. Although, there has been no significant decrease in mortality for that cause in recent years.

There is a need to elaborate guidelines and algorithms for identifying potential causes of SCD among young athletes. This type of supervision should be conducted by a multidisciplinary team including specialists in various branches of medicine, because the diagnostics, treatment and adjudication about the ability to do sports are problems from the borderline of many medical specialties usually. Therefore, it seems necessary to set up a diagnostic and treatment groups which should include sports medicine physician, specialist in medical rehabilitation and physiotherapy, sports psychologist or specialist in diagnostic imaging. The core of such a team should be a cardiologist with experience in working with sportspeople.

Prevention of SCD in athletes should be focused on not only regular medical examination, but also the education about worrying symptoms and negative effects of pharmacological support. Early diagnosis and detection of abnormalities allow to select a group of people at risk and taking appropriate action in a particular clinical situation. Awareness of the scale of the problem and knowledge about first aid is also of great importance in the prevention of sudden cardiac death. Therefore, the positive trend to provide the access to an automated external defibrillator (AED) in every sport facility is observed. As demonstrated by numerous studies, defibrillation performed in the shortest possible time (optimally up to 5 minutes) in the cause of cardiac arrest has nearly 60% effectiveness in athletes.

The analysis of data provokes several questions, of which the most important seems to be “Is sudden cardiac death in athletes a fatal complication of intense physical exercise, or

perhaps a previously undiagnosed anomaly of the cardiovascular system, manifested for the first time as sudden cardiac death?" The solution to this problem remains open and, at the same time, very complicated issue, as research suggests that SCD in athletes is a process that combines certain genetic predispositions and the induction of hidden pathology resulting from physical exertion.

Nowadays, as the blurring of lines between hobby and professional sport is observed, and athletes are taking extreme effort to get the highest result, medicine – especially sport cardiology - faces a huge challenge to early diagnose and treat discussed conditions effectively for prevention of dramatic incidents of the death of young people in sports arenas.

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