Sudden Death and Sport. Is There a Feasible Way to Prevent it in Athletes?

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INTRODUCTION

Physical activity is considered a faithful reflection of the state of health and quality of life of a society, which is why public opinion finds it hard to understand how an apparently healthy young person can die while showing great vitality in his or her usual athletic activities. Cases of sudden death fill the headlines of the sports press, especially when they occur in well-trained athletes with an excellent athletic performance. Unfortunately, these athletes did not know that they were silent carriers of potentially lethal cardiovascular diseases, in spite of undergoing numerous medical controls in the course of their athletic career.

In recent years, several of the cardiovascular diseases that most frequently are responsible for the sudden death of trained athletes or of apparently healthy young persons have been identified. The article of Suárez-Mier and Aguilera published in this issue of the REVISTA ESPAÑOLA DE CARDIOLOGÍA provides interesting information on the most frequent causes of sudden death associated with athletic activities in Spain.

PRESENT STATUS OF THE PROBLEM

Findings indicate that sports appreciably increase the risk of suffering sudden death during intense athletic activity. The seasonal distribution of sudden death and its greater frequency at certain times of the day support this fact. In competitive athletes, understanding as such athletes who participate in organized sports, sudden deaths occur most frequently in autumn and spring, which are the seasons in which most competitive sports events are celebrated, and in the early hours of the afternoon, coinciding with the times when most athletic events are held. In recreational athletes deaths occur in the early hours of the morning and late hours of the afternoon, coinciding with the times of day in which these activities take place.[1] In addition, people who practice intense athletic activities have a greater incidence of sudden death than non-athletes, 1.6 deaths per 100 000 versus 0.75 per 100 000.[2]

Cardiovascular disease is the most frequent cause of sudden death, as in persons who do not practice sports. Between 74% and 94% of non-traumatic deaths that occur during athletic activities are due to cardiovascular causes.[3] In the article of Suárez-Mier and Aguilera,[4] which examines 61 cases of sudden death that occurred during athletic activity, the predominant pathology was coronary atheromatous disease, with 25 cases (40.9%), and arrhythmogenic cardiomyopathy, with 10 cases (16.3%). Unlike other published series, hypertrophic cardiomyopathy, idiopathic left ventricular hypertrophy, myocardial fibrosis, anomalies of the coronary arteries, and aortic valve disease had a scant prevalence. Nevertheless, the number of deaths of indeterminate cause, 10 cases, all in athletes under the age of 30 years, is noteworthy, being 16.3% of the total sample and 31.2% of the sudden deaths in this age group, in which sports competitions are more frequent. The explanation could lie in the main mechanisms involved in sudden death, which are related to the hemodynamic and electrophysiological changes that take place during exercise and can differ with the type of exercise. During physical exercise, an increase in circulating cathecholamines takes place that is increased by the stress generated by competition. This enhances the responses of blood pressure, heart rate, and myocardial contractility, with the consequent increment in myocardial O_2 consumption. On the other hand, sympathetic stimulation can in itself favor the appearance of arrhythmias or aggravate underlying myocardial ischemia.

Causes of sudden death in athletes by age

Age conditions the prevalence of sudden death

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during athletic activity. In persons under the age of 35 years, the risk is exceptionally small, with an incidence of 1/200 000/year, whereas in persons over the age of 35 years the risk of suffering sudden death is 1/18 000/year.[5] The pathologies responsible for death are also related to age; consequently, the causes of death are generally congenital and almost never of ischemic origin in young athletes. In American series, the main causes of death in this age group are hypertrophic cardiomyopathy and congenital anomalies of the coronary arteries. Nevertheless, statistics for the European continent indicate appreciably different results, arrhythmogenic right ventricular cardiomyopathy and myocarditis being the most frequent pathologies.[6,7] Other, much rarer, causes of death in young athletes are arrhythmogenic syndromes, cerebral vascular malformations, bronchial asthma, and *commotio cordis* syndrome. This strange phenomenon deserves a special mention because it is associated with sudden death in children and very young athletes, less than 20 years old. Death occurs as a result of a non-penetrating direct impact to the chest over the cardiac region, with a forceful object that acts as a projectile, in susceptible persons and at a specific moment of the cardiac cycle. In these cases there is no baseline heart disease and no structural cause is found that predisposes to death. There are several possible mechanisms implicated in this syndrome: apnea, excessive vasovagal reflex, vasospasm, and primary ventricular arrhythmia.

Initially, 25 cases of unexplained cardiac arrest were described, but in none of them was the blow apparently of sufficient magnitude to cause death. Recently, from the data of the U.S. Commotio Cordis Registry, 124 cases have been identified in which only 14% of the victims have survived *commotio cordis*, thanks to rapid cardiopulmonary resuscitation maneuvers.[8]

In the series of Suárez-Mier and Aguilera,[4] of persons under 30 years, although the cause of death could not be identified in more than 30% of cases, the predominant pathology was arrhythmogenic cardiomyopathy (21.8%). As in the Italian series, there was a statistically significant difference with respect to persons of the same age who did not practice sports. This finding corroborates the idea that sports are a risk factor in patients with this disease. Unlike series from northern Europe, myocarditis was present only in the group of non-athletes and anomalies of the coronary arteries (second cause of death in the American series and third cause in the Italian series) had a scant prevalence, only two cases. These differences are probably due to the fact that the organs were from judicial *post mortem* studies sent by forensic physicians. These pathologies were diagnosed previously and since it was an anatomopathological study in which the clinical history was unknown, pathologies like long QT syndrome, Wolff-Parkinson-White syndrome, idiopathic ventricular fibrillation, *commotio cordis* phenomenon, or anaphylaxis induced by exercise could not be diagnosed and are included in the group of indeterminate origin.

In the group of athletes over 35 years, ischemic heart disease is the first cause of death and more than 90% of the cases involve confirmed coronary heart disease. Although evidence exists that the type of physical exercise that develops resistance protects against the development of coronary arteriosclerosis and that the probability of suffering ischemic heart disease is lower than in persons who practice sports, the risk of suffering a cardiovascular accident is higher during or immediately after exercise.[5,6]

APPROACH TO THE PROBLEM

The prevention of sudden death associated with sports must be based on three pillars: the pre-athletic event cardiological examination (PECE), taking the necessary measures for cardiopulmonary resuscitation, prompt defibrillation, and, finally, the preparation of national registries in which all deaths are recorded.

The pre-athletic event cardiological examination

The main objective of the pre-event cardiological examination must be the early detection of cardiac pathologies that carry a risk of sudden death. Nevertheless, the PECE is a topic of debate regarding the diagnostic methods necessary, the capacity to identify subjects at risk, and the effectiveness of the examination in modifying the natural history of the diseases most frequently associated with sudden death during athletic activities. Nevertheless, economic reasons are among the most important for introducing this type of examinations, and in some countries like the U.S., they are not well accepted because they are considered a breach of privacy and personal freedom. Nonetheless, the PECE meets the necessary conditions to be considered of public utility and should be promoted by the institutions responsible for the welfare of athletes.

When designing a PECE, three main questions must be answered: what should we be looking for? What tests should be included? And, finally, is it effective? The first question is relatively easy to answer because, out of the large variety of heart diseases described as responsible for sudden death in young athletes, some occur more frequently. According to observations made in post mortem studies in the U.S., hypertrophic cardiomyopathy and coronary anomalies represent almost two-thirds of all causes of sudden death. In Italy and in Spain, according to the work of Suárez-Mier and Aguilera,[4] arrhythmogenic cardiomyopathy of the right ventricle constitutes about 25% of all causes. Among the many other pathologies that occasionally

are responsible for cardiac sudden death, the least rare are myocarditis, cardiac valve anomalies (prolapse of the mitral valve and aortic stenosis), Marfan syndrome, long QT and Wolff-Parkinson-White syndromes.

In older athletes, there is no doubt that the pathology that should be screened is ischemic heart disease.[5,8]

With regard to what tests should be included and their effectiveness, the examination is the object of much speculation. Many restrictive studies that are guided by purely economic criteria recommend assessing only the history and making a clinical examination, whereas at the other extreme is the Italian model of examination, which also includes a resting electrocardiogram (ECG), submaximal exercise stress test, and an echocardiogram.

The screening protocol should always be simple enough to be easily applied, but should always have the capacity to identify (or at least suspect) the presence of cardiac disease of risk. The American Heart Association and American Sports Medicine Association, in scientific statements about the pre-event cardiovascular examination for competitive athletes,[9] establish the need for screening for cardiovascular disease in participants in competition sports. Screening should include a careful study of personal and familial history and a physical examination designed to identify the cardiovascular injuries that can induce sudden death or disease progression.

A meticulous physical examination can alert us to the presence of certain heart diseases. Unfortunately, most young athletes with condition that put them at risk are asymptomatic, have normal examination results, and present an excellent athletic performance. Inclusion of a resting ECG increases the probability of identifying subjects at risk, since it is abnormal in 95% of cases of hypertrophic cardiomyopathy and abnormal in arrhythmogenic cardiomyopathy and in the long QT and Wolff-Parkinson-White syndromes.

Nevertheless, the ECG cannot detect anomalies of the coronary arteries, which is why it has been proposed that echocardiography be included in the screening protocols of cardiovascular diseases for large population groups or at least in athletes planning to carry out programmed athletic activity. Some authors defend the use of an echocardiography limited to the longitudinal parasternal axis views in bidimensional mode as very useful.[10] Conclusions vary and are conditioned by criteria of economics and the prevalence of cardiovascular disease. In contrast, the Italian model without doubt has the most experience in the extensive use of echocardiography as a screening method in competitive athletes. So far there have been no conclusions about this model because no national record is kept in Italy of the persons excluded from practicing sports due to the presence of risk pathologies, and there is no national sudden death register.

The fact that the most frequent cause of sudden death in relation to exercise in persons over 35 years is ischemic heart disease and the high prevalence of cardiovascular disease in this population indicates that pre-event examinations can be effective in this age group. Nevertheless, the low sensitivity and specificity of the ECG at rest and the exercise ECG mean that there is no unanimity of criteria with respect to this group.

The results of the Italian experience allow some conclusions to be made regarding the effectiveness of screening to identify athletes at risk. Hypertrophic cardiomyopathy appears in a surprising low proportion of athletes who die suddenly in Italy in comparison with statistics for the U.S.A., and this discrepancy is not due to a lower prevalence of the disease. With respect to right ventricular arrhythmogenic cardiomyopathy, screening seems to be less effective. The incidence of this pathology is greater among athletes who die suddenly than among non-athletes, a finding that is confirmed by the results of Suárez-Mier and Aguilera.[4] Nevertheless, most athletes who die suddenly present relevant anomalies in the interview, clinical examination, or ECG, but the physician carrying out the examination fails to reach a diagnosis, probably because the presence of this cardiomyopathy is not even suspected or investigated. It seems reasonable to think that the availability of more information on the characteristics of this pathology, which until now has been little known, could improve the effectiveness of the examination.

With regard to congenital anomalies of the coronary arteries, the assessment of screening results is certainly discouraging. The family history is negative, symptoms are rarely found, and only a small proportion has ECG disturbances. Finding such anomalies in life is truly exceptional, which is why it seems reasonable that the diagnosis can elude the examining physician. In addition, in these subjects the effort ECG usually does not demonstrate signs of ischemia, but ventricular arrhythmias instead. On the other hand, it should be remembered that the echocardiogram makes it possible to visualize the ostium and proximal segment of the epicardial coronary arteries, which is why it can be a simple and decisive diagnostic method.

Considering the results of the study of Suárez-Mier and Aguilera,[4] the medical examinations currently made in Spain are insufficient to detect the most frequent pathologies responsible for sudden death associated with sports, because in 16 cases (26.2%) the pathological history was known, but the disease was identified in only three. Therefore, considering the recommendations of the Guidelines for Clinical Practice of the Spanish Society of Cardiology of 2000, the PECE should be specific for the age group and

level of athletic activity, and should always include a health questionnaire, medical history with detailed interview, exhaustive cardiovascular examination, and an ECG. In intensive organized and recreational sports, an echocardiogram should be included for young athletes, and a maximum stress test for older athletes.[11]

Cardiopulmonary resuscitation and sudden death register

Identification of the athletes at risk makes it possible to separate them from athletic activities in order to reduce risk and possibly prevent sudden death. Nonetheless, sudden death is a phenomenon that unfortunately occurs, generally in close chronological relation with the athletic activity (most events occurs during or immediately after training or competition). Therefore, although the cultural and legislative approach is very different in each country, responsible institutions should establish the necessary regulations to make possible fast and effective cardiopulmonary resuscitation. This means training members of the athletic community to perform basic cardiopulmonary resuscitation maneuvers and installing semiautomatic defibrillators everywhere athletic activities take place (sports centers, gymnasiums, university campuses, etc).

In addition, all deaths should be included in the National Registry of Accidental and Sudden Death during Sports (ASDS), and a post mortem study should be made of all athletes who die suddenly. Forensic pathologists should be aware of the need to submit sufficient clinical and anatomopathological information, both gross and histological, to eliminate bias and obtain data of statistical and epidemiological value.

In conclusion, although physical activity is beneficial for health, it carries an increased risk of sudden death. The study of Suárez-Mier and Aguilera, although lacking in epidemiological or statistical value, contributes very interesting information on the most frequent causes of sudden death associated with sports activities in Spain. Their results corroborate the similarity between the Spanish and Italian populations and the difference with the American and Scandinavian populations, confirming the need to modify medical examinations of athletes.

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