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CONSENSUS DOCUMENT

Consensus for the prevention of sudden cardiac death in athletes

Consenso para prevenir la muerte súbita cardíaca de los deportistas

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Introduction

Sports medicine is a specialty which emerged in parallel with the great importance sports acquired in society from the second half of the twentieth century. Its body of knowledge has always encompassed a unique interest in the effects of training on the cardiovascular system and its state in terms of its ability to cope with the different strains exerted by physical activity and sports, especially high-level. Therefore, specialists in sports medicine have always worked closely with cardiology specialists, which has led to a growing understanding of the effects of practising sports and, in turn, a better elucidation of the physiological changes that intense activity gives rise to in the hearts of those who practise it.

Due to all of the above, and taking into account the social impact of sudden death in athletes, from a range of institutions concerned, we decided to try and establish a minimum clinical agreement intended as a contribution to the criteria to be met from a cardiovascular standpoint in order to minimise the potential risks associated with practising sports.

This document is simply intended as a way to demonstrate that there are ways in which different medical specialties can collaborate and reach agreements to clarify their stances on issues as difficult in the world of sports as sudden death.

#1# Sudden death and sports

Sudden death is the kind that occurs abruptly and unexpectedly within one hour of the onset of symptoms. When it affects young patients, especially athletes, it is devastating and challenges the very concept of sport, which is typically regarded as beneficial for cardiovascular health. This causes major social concern and sparks a great deal of media attention; the strong interest for the implementation of measures which would contribute to the prevention of sudden death in athletes is therefore not surprising. This document aims to summarise the consensus reached by a group of Catalan experts operating in different medical disciplines in relation to the measures to be taken to prevent sudden death in the field of Catalan sports.

The incidence of sudden death in the context of athletic activities described in the different series is between 0.5 and 3 per 100,000 people/year[1]. Despite current limitations in the records compiled to date, such as the lack of centralisation and the sources based on news reports published in the media, these figures appear fairly consistent in the literature. For example, a recent follow-up study of the Boston Marathon over a 10-year period showed an incidence of sudden death of 0.54 per 100,000 participants/year[2], similar to that described in another equally recent French study conducted in athletes from different disciplines, where it was found to be between 0.5 and 1.7 per 100,000 participants/year[3]. Moreover, the incidence rate varies according to the gender, race and intensity of training assessed, and based on the division or category played by the athletes, the highest incidence being observed in black athletes (6 per 100,000/year) and first division players, who are theoretically subjected to more demanding and intensive training[4]. Lastly, studies based on post-mortem records in military populations have also described incidence rates of 4 per 100,000/year in subjects aged less than 35 years[5].

In the vast majority of cases, sudden death is caused by inherited or acquired underlying heart disease. In young people aged less than 35 years, the most common causes are hypertrophic cardiomyopathy (which affects 1 in 500 people), arrhythmogenic cardiomyopathy and other conditions, such as coronary artery disorders or channelopathies (with long QT syndrome, which affects one in every 2,500 inhabitants). In older subjects, atherosclerotic coronary disease is the leading cause; in addition, in Western countries its prevalence is on the rise in younger age groups[6].

The causal relationship between sporting activity and sudden death is still being debated. Based on the Italian experience[7], when dividing the population between athletes and non-athletes, the incidence of sudden cardiovascular death is almost three times higher in athletes than in the population that does not practice high level sports (2.3 *versus* 0.9 per 100,000/year). When analysing the cases of sudden death according to underlying heart disease, it was found to be much more common in athletes than in non-athletes, suggesting a causal rather than accidental relationship between the practice of sports and the occurrence of sudden death with a predisposing, underlying heart condition. Furthermore, in subjects aged over 35 years in whom the risk of sudden death is relative to the prevalence of ischemic heart disease, it has also been shown that physical exercise increases the risk, especially when practised on an occasional basis[8,9]. Other authors argue that the Italian study is the only evidence that the incidence of sudden death is higher in athletes, and that more recent records question the robustness of this data; in fact, there are no further data comparing rates of sudden death in athlete and non-athlete populations, which raises the question of whether it would be acceptable to selectively evaluate young athletes without performing a broader assessment of the entire population[10]. The potential role of banned substances as triggers of these malignant arrhythmias is also being debated, as is that of the drugs used in the treatment of attention deficit disorder, a common condition among child and adolescent athletes[11].

Obviously, unless specific tests are performed, most of these pathologies can only be detected in the event of actual sudden death, since nearly all of them progress without any other symptoms until the occurrence of a potentially lethal arrhythmic event[12]. On the other hand, it is well known that there are heart conditions, such as hypertrophic cardiomyopathy or valvular heart disease, in which exertion can lead to cardiac collapse due to the onset of malignant arrhythmias, as these are associated with an increase in haemodynamic requirements and adrenergic discharge. Other medical conditions carry a risk of death directly related to exercise, e.g. adrenergic ventricular tachycardia, which occurs in connection with the same kind of adrenergic discharge as that which takes place during physical exertion. Different clinical practice guidelines advise against physical exercise in the presence of many of these pathologies. It therefore makes sense to

Table 1 The 12 points proposed by the American Heart Association

Medical history-taking

1. Chest pain or discomfort
2. Unexplained syncope or presyncope
3. Dyspnoea or unexplained fatigue, disproportionate to the level of exertion
4. History of heart murmur
5. High blood pressure
6. History of premature death (before age 50) in the family (either sudden or unexpected) from cardiac causes
7. Heart disease in a relative under 50 years
8. Known family history of heart disease: hypertrophic or dilated cardiomyopathy, Marfan syndrome, channelopathies or other arrhythmias

Physical examination

9. Heart murmur
10. Peripheral pulses to rule out aortic coarctation
11. Marfan syndrome stigmata
12. Measurement of blood pressure

think that exercise may at least act as a trigger for arrhythmic events in subjects with underlying heart disease.

Preventative measures

The non-negligible reported incidence of sudden death in athletes, its impact on the healthcare system and the fact that this underlying disease could potentially be diagnosed in the majority of cases if the appropriate measures were taken all raise the need for active prevention of sudden death in athletes.

The first measure proposed is having access to automated external defibrillators (AEDs) in stadiums and sports venues, creating cardioprotective spaces with on-site staff trained in basic cardiopulmonary resuscitation (CPR). Indeed, this measure has been largely facilitated by the simplification of AED devices, which these days require virtually no staff intervention and rely on automated algorithms for their effective operation.

A further measure which aims to prevent sudden death in athletes is the early diagnosis of underlying heart conditions with the potential to act as a proarrhythmic substrate, through the systematic cardiological evaluation of athletes. The prevalence of cardiovascular diseases likely to prove lethal due to the associated risk of inducing sudden death in young athletes is reported to range between 0.2 and 0.7%[13].

Since a preparticipation screening programme was introduced in the Veneto region in 1982, the annual rate of sudden death in athletes has decreased from 3.6 to 0.4 per 100,000 persons/year; this reduction is owed to the increased identification of patients with cardiomyopathies, including hypertrophic cardiomyopathy, arrhythmogenic right ventricular dysplasia and dilated cardiomyopathy, the incidence of which increased from 4.4% in 1979 to 9.4% in 2004. At the same time, untested athlete populations exhibited no change whatsoever in the incidence of sudden death, suggesting that the significant reduction in mortality was not due to population changes, but rather to a genuine reduction in deaths from cardiomyopathies[1]. This experience remains to be confirmed, but recent data from a study conducted in Israel[14] and the significant limitations associated with creating a register of sudden deaths based on the information given to the media rather than data from a centralised register call its reliability into question.

Therefore, there is somewhat of a consensus now whereby a cardiac evaluation should be performed; unfortunately, there is also intense debate regarding the components that a systematic evaluation program should ideally include. This has resulted in a failure to adopt a uniform evaluation strategy. The purpose of this Consensus is to establish uniform guidelines within the territory in order to be able to draw conclusions regarding what really happens in our environment.

The recommendations accepted in the US and what is commonly done in this country involves simply administering a clinical questionnaire and conducting a physical examination to rule out potential heart disease (12 questions from the American Heart Association, table 1)[15]. Alternatively, the European consensus document proposes, based on the Italian experience, the incorporation of ECG as a tool to improve the detection of specific diseases, such as certain cardiomyopathies or channelopathies (Brugada syndrome, for example)[16]. Lastly, the International Olympic Committee also recommends systematic cardiovascular evaluations, including the performance of an ECG.

The American approach has been defended on the basis of the large number of athletes, the lack of dedicated or specialised doctors and available resources, and most of all, the potential induction of false positives when interpreting ECGs.

At least four studies have demonstrated improved sensitivity and even cost-effectiveness when an ECG was added to the medical records of athletes and their physical examinations. In the studies by Wilson et al.[17] and Hevia et al.[18], all cases of cardiovascular disease were detected by ECG and could not have been detected based on clinical history and physical examination alone. In the studies by Bessem et al.[19] and Baggish et al.[20], the sensitivity of detection of potentially lethal cardiovascular disease increased from 33 to 67% with the addition of ECGs. Overall, these studies evaluated more than 4,800 athletes from different countries (UK, US, Netherlands and Spain), detecting 17 cases of potentially fatal cardiopathy; the diagnostic utility of history-taking plus physical examination was 12%, increasing to 88% simply by adding in ECG.

One of the main criticisms of the implementation of ECGs is that the high rate of false positives leads to further and unnecessary examinations, sometimes even to discontinuation of the sports practice. Updated criteria for interpreting athletes' ECGs were recently proposed to enable a better distinction of what may be an actual physiological adaptation to changes suggestive of underlying heart disease[21]. The most significant change in these new guidelines was the elimination of isolated QRS voltage criteria to definite left ventricular hypertrophy, which is not a very sensitive marker, being found in up to 40% of athletes and less than 2% of patients with hypertrophic cardiomyopathy[22]. The application of these modern ECG interpretation criteria in athletes has led to a significant decrease in the rate of false positives, which now lies between 2 and 4% in Europe, and around 4 to 9% in the US.

Regarding the cost-effectiveness of adding ECG to history-taking and physical examination, several studies have shown that ECG screening is effective to improve diagnostic sensitivity and for the detection of index cases involving the diagnosis of other family members. Further, Wheeler et al.[23] showed that much of the cost of cardiovascular assessments in athletes was already established based on the medical history and physical examination, and that the addition of ECG optimises the cost-effectiveness of these cardiovascular evaluations.

Despite improvements in diagnostic ECG criteria, it is known that ECG readings do not detect all heart diseases likely to predispose to sudden death: an ECG cannot diagnose coronary arteriosclerotic disease or anomalies of the origins of the coronary arteries; neither can it identify patients with ascending aortic disease. In addition, it may fail to diagnose up to 5% of patients with hypertrophic cardiomyopathy[24]. In fact, the limited predictive and diagnostic role of ECG has been invoked to introduce the application of exercise stress testing and non-invasive cardiac imaging techniques, especially echocardiography, due to its safety and wide availability, in athlete screening programmes[25]. The usefulness and possibility of performing a limited echocardiogram integrated in athlete cardiovascular assessment programmes was also recently described[26]. In an Italian study, Rizzo et al.[27] found heart disease on the echocardiograms of 2% of the 3,100 athletes studied and concluded that some cardiac abnormalities, although minor but requiring cardiac monitoring, are not detected through history-taking, physical examination and ECG, and that therefore, the introduction of echocardiography, at least in the initial cardiovascular assessment of athletes, could help increase the effectiveness of screening programmes and the prevention of sudden death in athletes. However, the maximum exercise stress test is also recommended in the cardiovascular assessment of middle aged or older subjects (> 35 years) who play sports for arteriosclerotic heart disease screening purposes[28].

Despite the official and commonly agreed position of the European Society of Cardiology, which recommends the cardiovascular assessment of athletes based on clinical history, physical examination and ECG[16], the practices adopted by the various sports associations and federations of European countries are inconsistent, even within one same country. Thus, although each one of them includes these three points, many also require the performance of an exercise stress test and echocardiogram[13].

Cardiovascular assessment of athletes in Catalonia

Although the debate regarding the optimal assessment method remains unresolved, it stands to reason to think that athletes should at the very least be entitled to receive information on the possibility of these screening programs. These would be applied especially to elite athletes, who are those with the highest visibility, but also to those with the greatest potential in terms of financial resources.

The clinical implications derived from the screening programme essentially lie in the disqualification of certain athletes; despite this, increasing efforts are being made towards understanding the cardiovascular pathophysiology of exercise and higher therapeutic potential, including the use of automatic defibrillators in certain cases.

We propose the adoption of a common practice throughout Catalonia for the cardiovascular evaluation of athletes. There are 600,000 federated athletes in Catalonia, 2,500 of who are included in the ARC [Alt Rendiment Català (High Performance Sports Training)] programme. In addition, based on data obtained from the Survey of Physical Activity Habits in Catalonia, an estimated 1,400,000 people practice some kind of sport in this region. The population targeted by the Consensus is therefore wide.

The objectives of this consensus are as follows:

- Early diagnosis of underlying heart disease.
- Monitoring of the effects of high intensity training on the cardiovascular system.
- Early treatment of sudden death in sports.
- Laying the foundations for starting a sudden death register in Catalonia.

Early diagnosis

We propose a common and standardised basic cardiovascular assessment programme and an advanced cardiovascular assessment programme.

Basic cardiovascular assessment programme

This is a minimum requirement programme to be extended to all of the large athlete population, whose main objective and priority consist in its deployment throughout Catalonia, according to a standard format. This programme would include clinical history-taking and physical examinations based on the 12 points proposed by the American Heart Association (table 1), plus the performance of a 12-lead ECG. Extending the inclusion of an ECG interpreted by trained and skilled staff ought to be the first objective of this consensus and the first measure to be implemented in our territory to assess the condition of any person wishing to practice sports. However, the routine performance of exercise stress tests is being ruled out to facilitate the realistic implementation of this programme.

The interpretation of ECGs and the issuance of fitness reports would be the responsibility of sports physicians operating in certified sports medicine centres; these doctors would also be in charge of referring athletes to cardiology units or a cardiologist, at their discretion To ensure the proper interpretation of ECGs, it is necessary to implement an adequate infrastructure across the network of accredited medical centres, with a reference cardiology centre which should be easily accessible to discuss any concerns that may arise or expand diagnostic studies, if necessary. However, it is essential for all health personnel assigned to these centres to receive proper and continuing training, and for these personnel to be responsible for the interpretation of these ECGs and performing physical examinations. Lastly, it is necessary to optimise and adapt existing resources to ensure the proper execution of the programme.

The programme would be mandatorily applicable to all federated athletes.

Advanced cardiovascular assessment programme

Second, we intend to propose an advanced cardiovascular assessment scheme which, in addition to medical history-taking and physical examinations based on the 12 items proposed by the American Heart Association (table 1) and the 12-lead ECG, should include echocardiography and a maximum voluntary effort test with heart rate and blood pressure monitoring, plus a 12-lead ECG. The scope of this program would be professional athletes, those who practice risky or extreme sports and athletes over 35 years, given that a higher prevalence of coronary atherosclerotic disease has been demonstrated in this age-segment of the population[8,9].

These assessments would be carried out in advanced or level II accredited sports medicine centres and/or cardiology clinics subspecialised in sports cardiology.

Athlete monitoring

In this regard, a realistic approach is required to strike a balance with available resources and maintain efficiency. We therefore propose the following medical monitoring schedule, to be guided by the programmes detailed hereafter:

Basic cardiovascular assessment programme (fig. 1)

- Regular, biennial assessment of most athletes.
- Annual assessment of:
  - Subjects with ECG abnormalities.
  - Athletes with a normal advanced cardiovascular assessment (combined biennially with the advanced cardiovascular assessment).

Advanced cardiovascular assessment programme (fig. 2)

- Regular biennial assessment of professional athletes, subjects who practise risky or extreme sports and athletes over 35 years.
- Annual assessment for patients with no disqualifying disorders.
- When directed by the cardiologist and sports physician.

Early treatment of sudden death in sports

The early treatment of sudden death in athletes requires the wide availability of automated external defibrillators (AEDs) in all sports facilities and in all sports medicine centres. The recent approval of a Decree Law empowering and authorising any individual to use an AED (without needing to be specifically trained or certified), and the technological advances which have simplified their operation constitute key facts to support the extension of their use not only to major sports venues (which tend to have more funds) but to smaller ones too. For the same reasons and in view of increasing their availability and use, it is necessary to simplify DAE training and maintenance rules. Lastly, in addition to expanding access to AEDs, it is necessary to ensure that all staff working in sports facilities and sports medicine centres receives immediate CPR training (table 2).

Laying the foundations for starting a sudden death register in Catalonia

In order to assess our situation and review the efficiency of our programmes, it is essential to keep a record of all cases of sudden death occurring in our territory. A sports cardiology reference group will be created which will consist of cardiologists and sports physicians who will be in charge of designing and filling in a centralised register. Within this principle, a database comprising clinical cases linked to heart disease and sports will be created to set up a clinical practice fund.

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Conflicts of interest

The authors declare that there are no conflicts of interest.

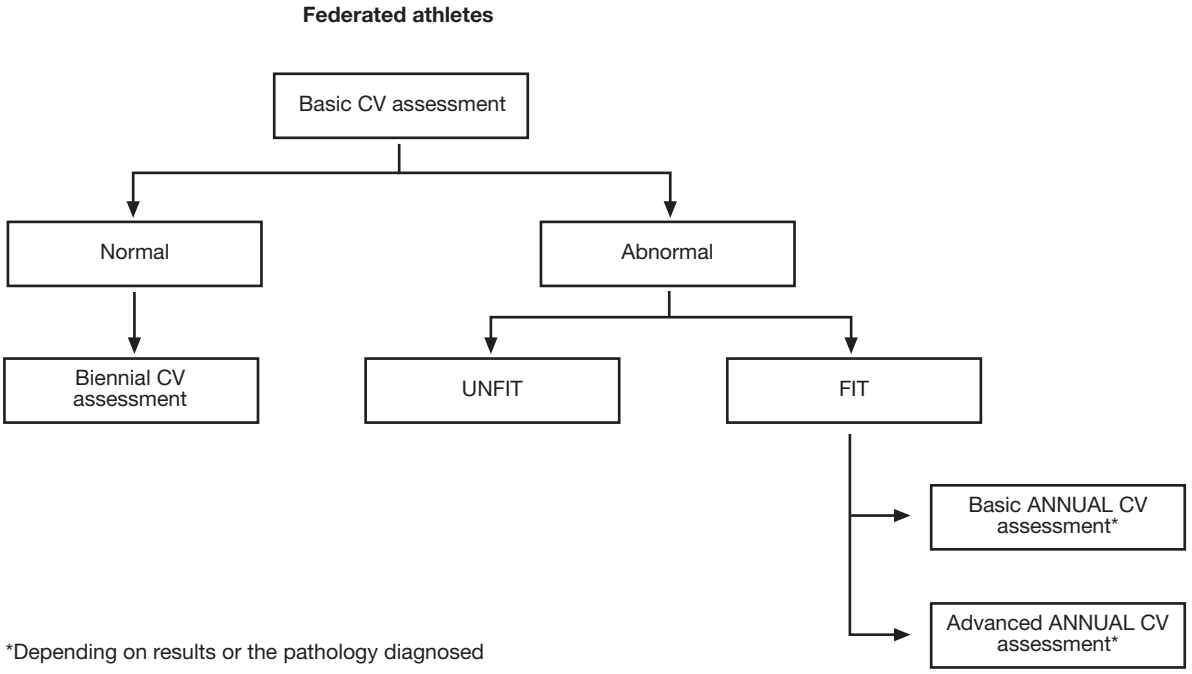

Figura 1 Tracking algorithm for federated athletes undergoing a basic cardiovascular assessment; the reviews to be carried out or implications in relation to fitness to practise sports are based on findings.

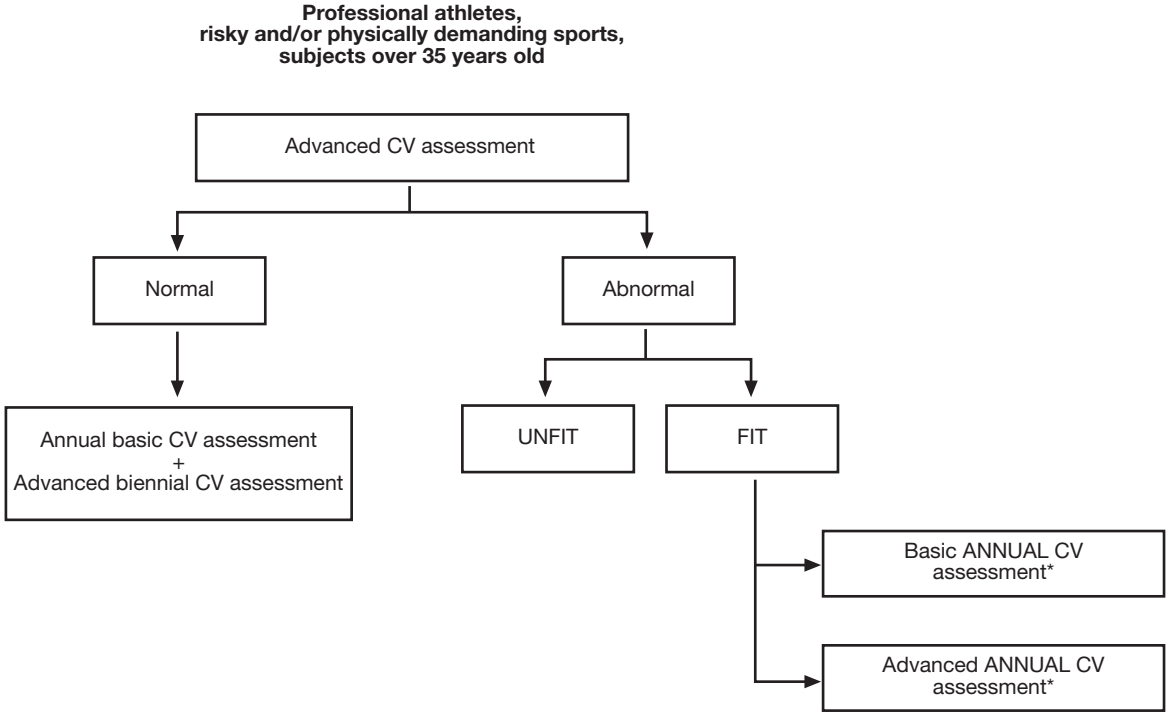

Figura 2 Tracking algorithm for professional athletes, those practising risky or extremely demanding sports or older than 35 years, in whom an advanced cardiovascular assessment is being initiated. The reviews to be carried out or implications in relation to fitness to practise sports are based on findings.

Table 2 Summary of the measures proposed for the prevention of sudden death in Catalan athletes

Early treatment
Access to an automated external defibrillator
CPR training
Early diagnosis
Basic cardiovascular evaluation (all athletes). Yearly
Medical history-taking
Physical examination
12-Lead ECG
Advanced cardiovascular assessment (competitive and professional athletes)
Medical history-taking
Physical examination
12-Lead ECG
Echocardiogram (at baseline and as per schedule)
Maximal exercise stress test (baseline and as per schedule)
Centralised register of clinical cases of sudden death

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