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SPECIAL ARTICLE
Sports cardiology

Recommendations for participation in competitive and leisure time sport in athletes with cardiomyopathies, myocarditis, and pericarditis: position statement of the Sport Cardiology Section of the European Association of Preventive Cardiology (EAPC)

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Myocardial diseases are associated with an increased risk of potentially fatal cardiac arrhythmias and sudden cardiac death/cardiac arrest during exercise, including hypertrophic cardiomyopathy, dilated cardiomyopathy, left ventricular non-compaction, arrhythmogenic cardiomyopathy, and myo-pericarditis. Practicing cardiologists and sport physicians are required to identify high-risk individuals harbouring these cardiac diseases in a timely fashion in the setting of preparticipation screening or medical consultation and provide appropriate advice regarding the participation in competitive sport activities and/or regular exercise programmes. Many asymptomatic (or mildly symptomatic) patients with cardiomyopathies aspire to participate in leisure-time and amateur sport activities to take advantage of the multiple benefits of a physically active lifestyle. In 2005, The European Society of Cardiology (ESC) published recommendations for participation in competitive sport in athletes with cardiomyopathies and myo-pericarditis. One decade on, these recommendations are partly obsolete given the evolving knowledge of the diagnosis, management and treatment of cardiomyopathies and myo-pericarditis. The present document, therefore, aims to offer a comprehensive overview of the most updated recommendations for practicing cardiologists and sport physicians managing athletes with cardiomyopathies and myo-pericarditis and provides pragmatic advice for safe

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participation in competitive sport at professional and amateur level, as well as in a variety of recreational physical activities.

Keywords Myocardial diseases • Sudden cardiac death • Hypertrophic cardiomyopathy • Arrhythmogenic cardiomyopathy • Myo-pericarditis • Cardiomyopathies • Athletes

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Preamble

The benefits of exercise on the cardiovascular system are well established and regular physical activity is an important component of minimizing the risk of several diseases. Participation in intensive exercise, however, may increase the risk of fatal arrhythmias in predisposed individuals. Inherited cardiomyopathies are among the leading causes of sudden cardiac death/cardiac arrest (SCD/CA) in young exercising individuals, including elite athletes. The link between cardiomyopathy and exercise-related SCD/CA is largely based on circumstantial evidence, due to the absence of systematic national registries. Nevertheless, based on the widespread impact of such tragedies on our society, there are several incentives to identify affected individuals on a timely basis through various screening processes, endorsed by the European Society of Cardiology (ESC) and the American Heart Association (AHA).[1–3]

The detection of a cardiomyopathy in an asymptomatic individual has important implications with respect to on-going participation in intensive exercise and competitive sport. In such circumstances, it is essential to strike a balance between protecting the athlete from the adverse effects of exercise (including adrenergic surges and haemodynamic loading) on abnormal myocardium and depriving the individual of the multiple benefits of exercise and a legitimate aspiration to excel in sport.

In 2005, the ESC published the recommendations for sport participation in athletes with cardiovascular diseases, which aimed to provide a practical guide for physicians.[4] Over the past decade, there have been several developments pertaining to exercise and sport participation in individuals with the cardiomyopathies, such as (i) a better understanding of the clinical features of cardiomyopathies in athletes, (ii) clarification of the natural history of the disease process in patients with arrhythmogenic cardiomyopathy, (iii) novel methods of differentiating physiological adaptations to exercise and early phenotypic manifestations of cardiomyopathy, (iv) the role of cardiovascular magnetic resonance (CMR) imaging in diagnosis and risk stratification in these patients, (v) the emergence of genotype-phenotype relationships in cardiomyopathies, and (vi) the potential role of competition with an implantable cardioverter defibrillator (ICD).

Based on the aforementioned considerations, the sports cardiology nucleus was compelled to provide an update for physicians involved in the management of athletes with cardiomyopathies or myopericarditis. As previously, the document is intended to provide a practical guide to aid the physician in the management of the athletes with these disorders. In the absence of robust evidence, these guidelines largely reflect expert opinion and cannot be considered as legally binding. Therefore, they should not discourage individual physicians to practice outside the remit of this document, based on their scientific and professional experiences in sports cardiology. In contrast with the previous document, and in line with developments in good clinical practice, the present document encourages shared decision making with the athlete and respects the autonomy of the athlete after careful information about the potential risks of an adverse event.

Shared decision making process

One of the shortcomings of managing athletes with heart disease is the scarcity of established risk stratification protocols in this cohort. Current risk stratification algorithms for patients with cardiomyopathy are derived from a sedentary population and are difficult to extrapolate to competitive athletes, who are subject to greater physical and metabolic stresses during intense exercise, which may culminate in fatal arrhythmias. Most deaths in athletes with cardiomyopathy affect individuals who do not have any of the conventional risk factors for SCD/CA. Furthermore, deaths may occur several years after embarking an athletic career without prior symptoms.

The unpredictable nature of SCD/CA in sport has historically necessitated a cautious approach regarding advice for intense exercise and competitive sport in individuals with a cardiomyopathy.[4] Such practice has inevitably affected several athletes who would never have experienced an adverse event during their entire sporting career and has been disputed as an intrusion into the personal freedom of the patient, rather than a precautionary measure aimed to reduce the unforeseeable risk associated with sport competition.

Based on the evolving knowledge in this field, we advocate adoption of an individualized approach that considers symptomatic status, established risk factors for SCD/CA, natural history of the disease, age of the athlete, duration of competition prior to the diagnosis, and characteristics of the sport discipline, when advising participation or precautionary disqualification from competitive sport.

We are mindful that disqualification from competitive sport has extensive personal consequences for the athlete, even beyond the medical perspective, including loss or compromise of significant sums of financial remuneration and the athlete's personal visibility. We, therefore, believe that it is crucial to involve the athlete in the decision-making process, which should include detailed information of the disease and open discussion about the potential risks associated with on-going competitive sport and high-intensity exercise programmes.

Athletes with full judgement and complete understanding who wish to participate in competitive sport despite contrary medical advice should not prejudice the examining physician. Furthermore, permission should be sought to inform attending coaches and team physicians to facilitate on-going clinical surveillance, including potential occurrence of SCA during training and competition.

Finally, we acknowledge that at present this process may be received differently in Europe, according to the cultural and societal rules and the medical competencies existing in each country.[5] The weight of an athlete's autonomy is also largely influenced by different legal systems in place and, therefore, the shared decision-making should be viewed and adapted in the context of the existing medical and legal rules.

Below, we provide a practical guide for the management of athletes with cardiomyopathy or myopericarditis. Based on novel data from cohorts of athletes with cardiac disease, the document includes brief updates in the diagnosis of cardiomyopathy in athletes and the risk stratification relative to exercise and sport participation. Finally, guidance for leisure sporting activities is also included to promote the cardiac and general benefits of physical activity in patients with cardiomyopathy.

Diagnosis of cardiomyopathy in athletes

The evaluation of athletes with suspected cardiomyopathy includes a detailed personal and family history, 12-lead ECG, echocardiogram, CMR imaging, and additional investigations for risk stratification. The diagnosis of cardiomyopathy relies on cardiac imaging studies, although a large proportion of affected patients also demonstrate electrocardiographic abnormalities. The electrical, structural and functional changes encompassing the definitions of various cardiomyopathies are generally beyond the usual changes observed in response to athletic training, but a small proportion of athletes do exhibit electrical and/or structural changes that overlap with subtle forms of cardiomyopathy. Such diagnostic dilemmas more commonly affect black athletes (of West-African or Afro-Caribbean origin), and endurance athletes of all races. The differentiation between the physiological adaptation to exercise and cardiomyopathy is paramount, given the potential consequences of erroneous interpretation. The presence of cardiac symptoms and family history of cardiomyopathy are important red flags for pathology. Specific enquiry regarding epilepsy, drowning or unexplained fatal car accidents is relevant because arrhythmic events may present in such fashion. The current document outlines the pertinent electrical, structural and functional features, which facilitate the differentiation between physiological cardiac adaptation and cardiomyopathy in athletes.[6]

Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy (HCM) is a primary myocardial disease caused predominantly by mutations within genes encoding sarcomeric contractile proteins and is characterized by a hypertrophied left ventricle in the absence of cardiac or systemic disease capable of inducing the same magnitude of left ventricular (LV) hypertrophy.[7,8] The condition is of special interest because it is one of the most common causes of exercise-related SCD/CA in athletes.[9–11] The vast majority of the young/adult athletes with HCM are asymptomatic and are identified following the investigation of an abnormal ECG. However, angina, syncope, or palpitation are recognized presenting features of the disease.

Electrocardiography

The 12-lead ECG is abnormal in over 95% of adult athletes with HCM.[12–14] Large QRS voltages are common in all individuals with HCM irrespective of athletic status. Concomitant inferior and lateral T-wave inversion, ST-segment depression and pathological Q waves are the most common abnormalities. Of note, lateral T-wave inversion has a 14% diagnostic yield for HCM but is also detectable in 4% of normal black athletes.[15] Electrical anomalies may precede overt structural disease by several years.[16] A minority (5–10%) of young individuals with HCM may have a normal ECG, or increased R/S wave voltage in isolation.[14]

Echocardiography

The diagnosis of HCM is based on the presence of unexplained LV hypertrophy with maximum end-diastolic wall thickness >15 mm in any segment on 2D echocardiography or CMR imaging.[7,8] The diagnosis of HCM may be considered in individuals with a LV wall thickness of 13–15 mm in the context of another family member fulfilling conventional diagnostic criteria for HCM, or positive genetic testing, or the aforementioned repolarization anomalies.[6–8] Whereas the maximal LV wall thickness rarely exceeds 12–13 mm in healthy white athletes (2%), this scenario is relatively common (13–18%) in male black athletes, who may reveal a LV wall thickness measurement of up to 16 mm.[6,13,17–21] The morphological overlap between physiological LV hypertrophy and HCM is less common in adolescent athletes, and observed in <1% of white athletes, but up to 7%, in African athletes.[22,23] Notably, LV wall thickness does not exceed 11 mm in white female athletes and 13 mm in black female athletes.[18,24]

As with sedentary HCM individuals, most athletes with HCM show an asymmetric pattern of LV hypertrophy[6–8] and in one series, approximately one-third of athletes with HCM had the apical variant.[13] In contrast, athletes with physiological LV hypertrophy show a more homogeneous and symmetric distribution of wall thickness, with only mild differences between contiguous segments and a symmetric pattern of LV hypertrophy.[17]

Left ventricular hypertrophy usually becomes evident during adolescence or early adulthood,[7,8] but may not be manifest phenotypically until the 4th or 5th decades of life. Left ventricular end-diastolic cavity size in individuals with HCM is normal or reduced, whereas athletes with physiological LV hypertrophy usually reveal an enlarged cavity (i.e. LV end-diastolic diameter >54 mm).[19] A subset (about 15%) of athletes with the apical variant of HCM demonstrates an enlarged LV cavity size.[13] Other subtle structural anomalies supporting the diagnosis of HCM include elongation of mitral valve leaflets, anomalous insertion of papillary muscles, or myocardial crypts and recesses.[7,8] Although diastolic function is usually abnormal in sedentary patients with HCM patients, most athletes with HCM reveal normal indices of diastolic transmitral filling[13,19] and reliance on abnormal transmitral Doppler filling pattern to exclude HCM has poor sensitivity in athletes. Abnormalities of the relaxation pattern of Tissue Doppler Imaging (TDI) may, however, be present in the early stage of the disease.[6] According to a study of 1510 athletes and 58 young asymptomatic individuals with HCM (50% of whom had a LV wall thickness ≤15 mm), septal E' <12 cm/s and lateral E' <12 cm/s on TDI were the best diastolic markers for HCM.[25] Left ventricular systolic function is usually within the range of normality in both athletes and HCM patients. Only subtle differences in myocardial contraction may be detected by the LV strain assessment.[6]

Cardiovascular magnetic resonance

Cardiovascular magnetic resonance imaging is recommended in all athletes with equivocal findings, such as LV wall thickness of 13–16 mm, major electrical abnormalities (deep T-wave inversion in the inferior and/or lateral leads), or symptoms suggestive of cardiac disease in the context of a family history of HCM. Cardiovascular magnetic resonance has unrivalled value in excluding localized hypertrophy, in segments that cannot be visualized adequately with echocardiography, such as the LV apex or the antero-lateral segments. The presence of late gadolinium enhancement (LGE) with non-ischaemic pattern in the hypertrophied segments is suggestive of myocardial fibrosis, and consistent with the diagnosis of HCM.[26,27]

Exercise and cardiopulmonary test and 24–48 h Holter monitoring

The presence of symptoms, non-sustained ventricular tachycardia, or ST-segment depression/T wave inversion during exercise, or flat blood pressure response (<20 mmHg rise in systolic pressure from baseline to peak exercise) and a low peak oxygen consumption (<84% predicted) are in favour of pathological LV hypertrophy.[28] Normalization of basal T-wave inversion or ST-depression during exercise does not exclude HCM.

Similarly, the occurrence of non-sustained ventricular tachycardia or any complex ventricular arrhythmias in an athletic individual with a LV wall thickness >12 mm to ≤16 mm during 24-h (or 48-h) ECG monitoring also favours the diagnosis of HCM.

Genetic testing

Genetic testing in athletes with an equivocal diagnosis of HCM should only be performed after detailed clinical and family assessment by an experienced cardiologist, as the absence of a sarcomere mutation does not exclude familial HCM and variants of uncertain significance can be difficult to interpret.[15] For this reason, genetic testing is currently recommended only in patients clearly fulfilling the diagnostic criteria for HCM, to enable cascade genetic screening of their relatives.[7]

Detraining

Finally, when the differential diagnosis cannot be resolved with conventional tests, useful information may be gained from detraining. Serial echocardiographic studies demonstrating regression of LV hypertrophy after a 3-month period of complete deconditioning in an athlete without family history of HCM are consistent with diagnosis of physiological LV hypertrophy.[29]

Risk stratification

Risk stratification in athletes with HCM is challenging, due to lack of prospective studies reporting the outcome of the disease in patients engaging in regular, intensive exercise. The current ESC[7] and AHA[8] algorithms to assess the risk of SCD in the general HCM population cannot be extrapolated to individuals regularly exposed to the haemodynamic and metabolic stresses of an athletic lifestyle.

Circumstantial evidence has suggested that intensive exercise and sport participation itself may play a role in triggering SCD/CA. Among young athletes in US, HCM is the most common cause of exercise related SCD/CA.[9] Participation in high-intensity competitive sports itself has been considered as an independent risk factor for SCD/CA, even in the absence of the conventional risk markers, due to accompanying alterations in hydration, electrolyte, and acid base status and surges in catecholamine levels.[30] The incidence of SCD is significantly higher in highly-dynamic sport, such as basketball, football, swimming, particularly in athletes of Division I compared with Division II and III, suggesting that intensity of training and level of achievement might have a causative role.[31] The age of an athlete may also have an impact on risk. The mean age of athlete in the largest series of SCD from the US was 17.1 years old.[9] In a recent report of outcomes in over 11 000 screened soccer players aged 16–17 years old who were followed up for just over 10 years, two of the eight cardiac deaths occurred in individuals diagnosed with HCM who chose to continue playing despite medical advice.[32]

On the other hand, retrospective observations indicate that SCD/CA is relatively rare in HCM even among affected individuals who exercise regularly.[33] Lampert *et al.*[34] reported that individuals with HCM who were implanted with an ICD had similar rates of appropriate or inappropriate shocks during exercise or at rest. A case series of HCM athletes, previously reported by Maron *et al.*[35] suggested that within the large spectrum of the disease, there are individuals who had been engaged in intense athletic training and competitions for many years, without incurring symptoms or clinical worsening or dying suddenly. Recently, Pelliccia *et al.* reported that in a small cohort of 35 athletes with HCM, who were followed up for a 9-year period, there were no differences in the incidence of symptoms or major events between athletes who had become sedentary ($n = 20$) after diagnosis and athletes who continued to engage in competitive sport ($n = 15$). In this cohort, there was one CA during the surveillance period, which was unrelated to exercise.[36]

Overall, the current literature indicates that not all individuals with HCM are vulnerable to fatal arrhythmias despite engaging in exercise

Table 1 Recommendations for athletes with HCM

	Class/level of evidence
1. Participation in intensive exercise programmes and competitive sport should be considered on an individual basis, after full evaluation of the disease characteristics and risk determinants. Specifically, conditions that reasonably represent absolute contraindications for sport participation include: (1) History of aborted SCD/CA; (2) Symptoms, particularly unheralded syncope; (3) Exercise-induced ventricular tachycardia; (4) High ESC 5-year risk score[7], (5) Significant increase in LV outflow gradient (>50 mmHg) (6) Abnormal blood pressure response to exercise.	Class IIb/Level C
2. Following comprehensive evaluation and explanation of the disease characteristics, risk factors and potential outcomes and assuring that a reasonable understanding and agreement has been reached between the athlete and the physician, it seems reasonable that adult athletes with: (1) Mild clinical expressions of HCM (2) Low ESC risk score[7] (3) Adult age may selectively be allowed to participate in all competitive sports, with exception of those where occurrence of syncope may be associated with harm or death (see *Figure 1*). Such athletes should be reviewed annually to assess symptoms and changes in risk profile.	Class IIb/Level C

programmes. In this regard, systematic restriction from competitive sport in all affected individuals is probably unjustified and a more liberal approach to sport participation may be reasonable after considering the age of the athlete, duration in competitive sport prior to diagnosis and the presence of conventional risk factors for SCD. It is indisputable, however, that absence of all major risk factors does not convey immunity to SCD, and even patients with low-risk may die suddenly.[37] Therefore, when advising an individual with HCM regarding participation to intensive exercise programmes and competitive sport this consideration should be an integral part of discussion during the shared decision-making process.

Recommendations

This recommendation should be viewed in the context of the cultural and customary medical care in place and should not override the existing medical and legal boundaries existing in the different countries (*Table 1*).

Genotype-positive, phenotype-negative hypertrophic cardiomyopathy athletes

Advances in the molecular genetics of HCM coupled with improvement in methods for gene sequencing has resulted in an increasing number of asymptomatic relatives of HCM patients being detected as gene-carriers in the absence of morphological or clinical features of HCM. Several reports have suggested that such individuals (G+P-) may exhibit a number of subtle abnormalities, such as high concentration of serum biomarkers of collagen synthesis, expanded extracellular volume at T1 mapping on CMR, LV wall thickness at upper normal values and subtle impairment of myocardial relaxation.[38–40]

Over the past few years, a limited number of studies have suggested that G+P- individuals have a benign clinical course, with absence of symptoms or adverse events and a small number develop the phenotype in the third decade, suggesting incomplete penetrance during most of a competitive athlete's career.[41–43]

Although the number of individuals studied in these reports, and their follow-up was limited, it is important to emphasize that there were no clinical events in mutation carriers before the development of an overt HCM phenotype (i.e. LV hypertrophy). In conclusion, available evidence from relatively small studies suggests that gene carriers for HCM do not incur risk until the phenotypic spectrum of the disease is developed, and therefore, the G+P- status should not equate to having the disease.

Recommendations

Adolescent athletes G+P-, should undergo periodical surveillance (at least, yearly), to monitor the potential progression to the overt HCM phenotype. A more liberal attitude is justified in adult (>25 years old) athletes, with recommended follow-up every 2 years (*Table 2*).

Dilated cardiomyopathy

Dilated cardiomyopathy (DCM) is a myocardial disease characterized by a dilated and hypokinetic LV or both ventricles [with or without associated right ventricular (RV) dysfunction].[45] Dilated cardiomyopathy may be genetic in origin, or secondary to infection or inflammation, toxic agents, ischaemia or idiopathic. Although infrequent, DCM is a recognized cause of SCD in athletes.

Athletes with an enlarged LV cavity with mildly reduced systolic function and/or associated ventricular arrhythmias fall into a grey zone between physiological LV enlargement and DCM.

Table 2 Recommendations for athletes genotype positive-phenotype negative for HCM

	Class/level of evidence
1. G+P- individuals should be assessed to exclude the broader phenotypic and clinical features of HCM (with ECG, CMR, exercise test, and 24-h ECG monitoring). In the absence of phenotypic features of HCM, these athletes may be allowed to engage in all competitive sports.	Class IIa/Level C
2. There is limited data in individuals with positive genotype, negative phenotype, and an abnormal ECG in isolation; therefore, until further data is available it is recommended that such individuals are managed similarly to those without LV hypertrophy.[44]	Class III/Level C)

12-lead ECG

The ECG may be normal or show similar changes to those of athletic training, such as voltages for atrial enlargement, axis deviation, or large QRS voltages in the lateral leads. Recognized electrical manifestations of DCM include T-wave inversion, intraventricular conduction defect, or left bundle branch block (LBBB). Of note, some ECG alterations, such as 1st degree atrioventricular (AV) block or pathological Q waves in the inferior and/or-lateral leads may be expressions of specific genotypes (i.e. Lamin A/C or dystrophin) mutations. Occurrence of ventricular arrhythmias, particularly during exercise, may be common.[45]

Echocardiography

In DCM, the LV cavity is usually enlarged and LV shape becomes more spherical with time, when the mitral annulus enlarges with distortion of leaflets and resultant valvular regurgitation. Most importantly, LV systolic function is reduced and the ejection fraction is usually below 50%. Regional wall motion abnormalities are occasionally present.[46,47]

It is recognized that male athletes engaged in mixed and endurance sport frequently demonstrate an enlarged LV cavity size. Approximately 15% of elite male athletes show a LV end-diastolic diameter >60 mm.[48] Left ventricular cavity enlargement in athletes should be interpreted in the context of the sporting discipline and body size (it is advised to relate LV cavity to body surface area) and is usually associated with increased wall thickness.[6,48,49]

The presence of concomitant RV remodelling, with normal RV function is consistent with a physiological LV remodelling.[6]

Occasionally, athletes with an enlarged LV cavity may demonstrate a mildly reduced LV ejection fraction (i.e. >45%, <55%) and bradycardia.[50] In this scenario, the differentiation between physiological LV remodelling and DCM may be challenging. In such instances, regional wall motion abnormalities and abnormal diastolic function may support DCM. Exercise echocardiography (or CMR) are both effective methods for assessing myocardial reserve and an increase in LV ejection fraction of >10–15%, during exercise, supports the diagnosis of physiological LV dilatation.[51,52]

Cardiac magnetic resonance

Cardiac magnetic resonance is the gold standard for the assessment of biventricular dimensions and function in equivocal cases. In DCM, LGE identifies focal replacement fibrosis and it is detectable in approximately one-third of patients with a distinctive mid-wall distribution. The presence of mid-wall LGE and its extent has emerged as an important tool in risk stratification of DCM.[53–55] Exercise CMR is useful for assessing myocardial reserve.

Cardiopulmonary exercise test and 24-h Holter monitoring

In DCM patients, exercise performance may be only mildly impaired in the early stages of the disease and may not be sensitive for differentiating physiological from pathological LV remodelling. However, the presence of a low peak oxygen consumption in an athlete with an enlarged LV and borderline/low LV ejection fraction supports DCM.

Arrhythmias are reported in approximately 30% of DCM patients and may be present even in the presence of only mildly dilated LV cavity.[55,56] In this regard, 24-h ECG monitoring (including a training session) is of particular importance in the risk stratification of DCM patients.

Risk stratification

The clinical course of DCM is variable and cardiac events (including SCD/CA) are related to LV function and the presence of atrial and ventricular tachyarrhythmias. The risk of adverse consequences in patients with DCM participating in regular exercise programmes is largely unknown, due to the lack of long-term observational studies. However, scientific evidence from rehabilitation studies in heart failure supports the concept that high-intensive exercise programmes are feasible and beneficial in patients with idiopathic DCM, and are not associated with an increased risk of SCD/CA.[57,58]

Recommendations

Participation in intensive exercise programmes and competitive sport should be considered on an individual basis, after evaluation of the disease characteristics and risk determinants. Advising an athlete with DCM regarding participation in competitive sport requires a comprehensive and clear explanation, and assurance of understanding of the associated risk on behalf of the athlete (*Table 3*).

Genotype-positive, phenotype-negative dilated cardiomyopathy athletes

Currently, there are over 50 genes implicated in DCM, which encode sarcomeric, cytoskeletal, nuclear envelope and sarcolemmal proteins, ion channels, and intercellular junctions.[59]

The impact of regular exercise training on the natural history of G+P- individuals is not yet fully understood, although individuals with an isolated pathogenic variant, in the absence of overt phenotypic features of DCM, should be not considered to have the disease.

Table 3 Recommendations for athletes with DCM

	Class/level of evidence
1. The presence of LV cavity dilatation with preserved LV function, in the absence of a family history of DCM, abnormal ECG patterns, and atrial/ventricular tachyarrhythmias should be considered to represent expression of physiological cardiac remodelling rather than DCM. Therefore, no restriction to competitive sports is applicable to this cohort of athletes.	Class IIa/Level B
2. Athletes with an unequivocal diagnosis of DCM, but mildly reduced LV systolic function (EF ≥40%) may selectively engage in all competitive sport, with the exception of those where occurrence of syncope may be associated with serious harm or death (*Figure 1*), if they are: (1) Asymptomatic, (2) Without prior history of unexplained syncope, and (3) Without frequent/complex ventricular tachyarrhythmias on ambulatory ECG monitoring and exercise testing. Exceptions include patients with a family history of SCD/CA and/or those previously identified during cascade screening for familial DCM, with mutations that are associated with an increased risk of life-threatening arrhythmias (such as Lamin A/C or Filamin C mutation), irrespective of the severity of LV dysfunction and dilatation. These patients should be advised not to engage in competitive sports.	Class IIb/Level C
3. Athletes with a diagnosis of DCM who are: (1) symptomatic, or have (2) LV ejection fraction <40%, or (3) extensive LGE (i.e. >20%) on CMR and/or (4) frequent/complex ventricular tachyarrhythmias on ambulatory ECG monitoring and exercise testing, or (5) history of unexplained syncope. should be advised not to engage in competitive sports. These patients should be advised to limit their exercise programmes to leisure-time activities and undergo regular clinical surveillance, consistent with current recommendations for the management of DCM.	Class III/Level C

Table 4 Recommendations for athletes genotype positive-phenotype negative for DCM

	Class/level of evidence
The G+P- athletes should be assessed to exclude the phenotypic and clinical features of DCM (including CMR, exercise test, and 24-h ECG). In the absence of evidence for DCM, these individuals may be allowed to engage in all competitive sports, with the recommendation to undergo a periodical evaluation, at least annually, in order to early detect the phenotypic expression of the disease.	Class IIa/Level C

Recommendations

See (*Table 4*).

Left ventricular non-compaction cardiomyopathy

Left ventricular non-compaction (LVNC) is a relatively novel cardiomyopathy characterized by prominent trabeculations within the LV myocardium, separated by deep recesses, usually associated with LV dysfunction.[60] The disease has a genetic basis in some instances and a variety of inheritance patterns have been reported. Clinical presentation of LVNC includes varying degrees of systolic heart failure, ventricular tachyarrhythmias and/or systemic thromboembolism.[60] In athletes, however, a suspicion of LVNC usually emerges in the absence of symptoms, based on the identification of a prominent trabecular pattern by cardiac imaging.[6]

12-lead ECG

Common ECG abnormalities in LVNC include T-wave inversion, ST-segment depression or ventricular conduction delay, mostly LBBB.[61]

Echocardiography/cardiac magnetic resonance

The morphological diagnosis of LVNC relies on the presence of a two-layered structure of the myocardial LV wall on imaging testing. Current diagnostic criteria are based on the demonstration of a high ratio of non-compacted to compacted layer of the LV myocardium, either at echocardiography (i.e. ratio >2) or at CMR (>2.3), although the validation of these criteria has been questioned.[6,62,63] Additional

supportive features include LV systolic dysfunction, with reduced (<50%) ejection fraction, but also a very thin compacted epicardial layer (i.e. <8 mm in systole on echocardiography)[64] and abnormal myocardial relaxation (e' <9 cm/s at TDI.[65]

Athletes frequently show increased trabeculations in the LV cavity (i.e. so-called hypertrabeculation pattern), and up to 8% may fulfil the morphological criteria for LVNC.[65] It has been postulated that an increased cardiac preload may simply unmask pre-existing trabeculations and make them more prominent. This hypothesis is supported by a longitudinal study, using the pregnancy model, which showed that almost 25% of primigravida women developed prominent trabeculation and 8% fulfilled criteria consistent with LVNC as pregnancy progressed to the third trimester.[66]

Only a small proportion (0.9%) of athletes with hypertrabeculation exhibit other clinical abnormalities supportive for diagnosis of a cardiomyopathy; these athletes need to be thoroughly investigated.[65] Specifically, athletes with LV hypertrabeculation and an abnormal ECG and/or mildly reduced LV function, or positive family history should undergo a complete evaluation including CMR and exercise echocardiography to assess the LV response to effort, and ECG Holter monitoring to ascertain the presence of arrhythmias, all findings that will support the diagnosis of LVNC.[6,65,66–70]

Risk stratification

The clinical outcome of LVNC is variable, even within families, and governed by the magnitude of LV dysfunction and prevalence of atrial and ventricular arrhythmias or thromboembolic events. Adverse consequences are largely associated with LV systolic dysfunction or major ventricular tachyarrhythmias. It is noteworthy that no major cardiac events have been reported in the absence of LV dysfunction, regardless the severity of hypertrabeculation.[65,67–70]

Recommendations

As specified above, advising an individual with LVNC regarding participation to competitive sport requires a comprehensive and clear explanation, and assurance of understanding of the associated risks on behalf of the candidate (*Table 5*).

Arrhythmogenic (right ventricular) cardiomyopathy

Arrhythmogenic right ventricular cardiomyopathy (ARVC), or simply arrhythmogenic cardiomyopathy (AC) is an inherited myocardial disease caused predominantly by mutations in genes encoding desmosomal proteins. The disease is characterized histologically by fibrofatty replacement of the right ventricle and/or LV myocardium, and clinically by life-threatening ventricular tachyarrhythmias.[71] Sudden death usually occurs in young AC individuals and is often triggered by exercise. Arrhythmogenic cardiomyopathy represents a common cause of SCD in prospective studies of young athletes in Italy[72] and in an unselected population of young adults from Australia.[73]

The diagnosis of AC is based on the criteria proposed by an expert consensus panel that recognize electrophysiological, anatomical, and clinical features of the disease.[74]

12-lead ECG

The ECG is of particular value in raising suspicion for AC and is abnormal in the majority (>60%) of individuals.[75] The most common abnormalities in the right-dominant variant include inverted T-waves in the right precordial leads (V1–V3), prolonged QRS duration >110 ms with right bundle branch block (RBBB) pattern and a delayed upstroke (>55 ms) of the S wave in V1–V2. Rare is the presence of an epsilon wave in V1 or V2. In the left-dominant variant low voltages of R/S wave in the limb leads are increasingly recognized, as well as the presence of diffuse

Table 5 Recommendations for athletes with LVNC

	Class/level of evidence
1. Athletes with incidental discovery of LV hypertrabeculation should not be diagnosed as LVNC in the absence of symptoms, positive family history, abnormal ECG patterns and, most importantly, impaired LV function. In such cases, no restriction for all competitive sports apply.	Class IIa/Level B
2. Athletes with unequivocal/reasonable diagnosis of LVNC but near-normal LV systolic function may participate in all competitive sports, with the exception of those where occurrence of syncope may cause serious harm or death (*Figure 1*), if they are: (1) asymptomatic, (2) without frequent and/or complex ventricular arrhythmias, or non-sustained VT on ambulatory monitoring and exercise ECG testing, and (3) no prior history of unexplained syncope	Class IIb/ Level C
3. Athletes with an unequivocal diagnosis of LVNC and (1) impaired LV systolic function and/or (2) frequent and/or complex ventricular arrhythmias, or non-sustained VT on ambulatory monitoring or exercise testing should be advised to abstain from participation in competitive sports. These patients should be advised to limit their exercise programmes to leisure-time physical activities and remain under regular clinical surveillance.	Class III/ Level C

T-wave inversion in the antero-lateral and inferior leads.[71,75] Not rarely, isolated premature ventricular beats (PVBs) are present, typically with LBBB pattern and vertical/horizontal axis (in the right-variant), or RBBB and superior axis (in the left-variant). Electrical changes may precede morphological abnormalities by several years.[71]

Echocardiography and cardiac magnetic resonance

Echocardiography and CMR may show an enlarged RV cavity in the right-dominant variant, with morphological abnormalities (i.e. thinning, bulging, and aneurysms of the RV wall), associated with wall motion abnormalities, which are evident only in advanced stage of the disease.[6] In the right-variant, the outflow tract is commonly more enlarged respect to the inflow tract.[76] In the early stage of the disease, however, morphological RV changes may be only mild or not so evident.

Although 2D echocardiography provides a readily available imaging tool, it has important limitations for visualizing the complex geometry of the right ventricle. Therefore, modern imaging relies more on CMR, which has superior diagnostic value in identifying segmental morphological RV abnormalities, including regional wall motion anomalies.[6,77]

In the left-dominant AC, the morphological abnormalities of the left ventricle may be mild or even undetectable at echocardiography, and CMR is the only imaging test to identify altered signal intensity, consistent with fibro-fatty replacement in the sub-epicardial region or mid-wall of the left ventricle.[71,77]

It is well known that endurance athletes develop an enlarged RV cavity in association with enlarged LV, both with preserved shape, as consequence of the physiological adaptation of both ventricles to chronic exercise training.[78,79] The physiological RV remodelling in athletes is characterized by a proportionate increase in the inflow and the outflow tract and the absence of segmental morphological thinning or wall motion abnormalities.[6,78,79] The RV dimensions by themselves may be insufficient to distinguish physiological from pathological RV remodelling and need to be associated with wall motion abnormalities to suggest AC.[74] Finally, care should be taken to avoid misinterpretation of certain CMR findings in athletes as pathological (such as RV apex dilatation, or localized apical bulging of the RV wall at the level of the moderator band).[77]

Similar to DCM, exercise imaging (by echocardiography or CMR) may be useful for discriminating between physiological RV enlargement with preserved systolic function in healthy athletes from pathological RV myocardial remodelling in AC.[51]

Cardiopulmonary exercise test and 24-h Holter monitoring

In young AC patients, exercise performance may be preserved. However, ventricular arrhythmias (PVBs and/or VT with LBBB morphology in the right-dominant, or RBBB in the left-dominant), are usually present at an early stage of the disease, and are usually triggered by exercise.

Genetic testing

In patients with ARVC, the most commonly affected genes encode desmosomal proteins.[71] The overall rate of successful genotyping among patients meeting the diagnostic criteria for ARVC is not more than 50%.[80] Moreover, the interpretation of an apparently positive genetic test is made challenging by the difficulty in differentiating pathogenic variants for ARVC, especially missense mutations, from non-pathogenetic variants and polymorphisms present in a minority of normal population.[81]

Clinically, genotyping is indicated to identify a pathogenic variant mutation in a proband who already fulfils the phenotypic diagnostic criteria in order to facilitate cascade screening of first degree relatives. Genotyping should not be used to confirm the diagnosis in an isolated patient with a borderline or questionable phenotype.

Risk stratification

In predisposed individuals, with abnormal cell-to-cell binding of the myocytes, the dilation of the right ventricle associated with regular exercise training may lead to myocardial damage and subsequent fibro-fatty replacement, thereby triggering the morphological features of the disease. Ventricular tachyarrhythmias and sudden death in AC commonly occur in association with exertion and AC accounts for a substantial proportion of deaths in young athletes.[71,82]

Prior aborted SCD, unheralded syncope, ventricular tachycardia and impaired right and/or left ventricular function are established risk factors for arrhythmogenic CA in AC. Exercise also appears to be an independent risk factor for expediting the disease phenotype and promoting fatal arrhythmias.[71,82,83]

In an experimental murine model of cardiac desmoplakin mutations, exercise training has been shown to increase the penetrance and the arrhythmic presentation of the disease.[84,85] Similar results have been confirmed in AC genetically positive human patients. Specifically, James *et al.*[86] investigated the penetrance of AC in 87 desmosomal mutation carriers, and found that endurance exercise training was associated with higher penetrance of the disease, earlier onset of symptoms, and increased risk of ventricular tachyarrhythmias and heart failure. Saberniak *et al.*[87] investigated myocardial function in AC patients, and found reduced RV function in athletes AC when compared with non-athletes AC. Recently, the results from the North American multidisciplinary study of ARVC[88] found that patients engaged in competitive sport were incurring a larger incidence of ventricular tachyarrhythmias/death and earlier presentation of symptoms, compared with patients who participated in only recreational physical activity and those who were sedentary. Among patients engaged in competitive sports, early age of sport initiation was associated with premature presentation of symptoms and adverse clinical profile. Reducing exercise intensity was associated with a substantial decrease in the risk of ventricular tachyarrhythmias or death, to the same level as inactive patients.[88] In summary, the overall scientific evidence supports the concept that participation in competitive sport is associated with earlier onset of symptoms and greater risk of ventricular arrhythmias and major events in AC patients.

These considerations are clinically relevant and support a restrictive advice regarding the participation in intensive exercise programmes and competitive sports in affected AC patients. Conversely, recreational exercise programme conveys a reduced risk, such as that of patients physically inactive.

Recommendations

Advising an athlete with AC regarding participation to exercise programmes or sport requires a comprehensive and clear explanation, and assurance of an understanding of the whole spectrum of exercise-related risks on behalf of the candidate (*Table 6*).

Table 6 Recommendations for athletes with AC

	Class/level of evidence
Athletes with unequivocal or probable diagnosis of AC should not participate in competitive sports. These patients should be advised to limit their exercise programmes to leisure-time activities, and remain under clinical surveillance.	Class IIa/Level C

Table 7 Recommendations for athletes genotype positive-phenotype negative for AC

	Class/level of evidence
Athletes who are genetic carriers of pathogenic AC-associated desmosomal mutations (even in the absence of phenotypic expression of the disease) should not participate in competitive sports. These athletes should be advised to limit their exercise programmes to leisure-time activities and remain under clinical surveillance.	Class IIa/Level C

Of note, life-long endurance athletes presenting with clinical features indistinguishable from AC, but without desmosomal mutations, are often referred to as 'gene-elusive AC' or 'exercise-induced RV cardiomyopathy'.[89–92] The work-up and recommendations in these athletes are identical as in inherited AC, as outlined above.

Genotype-positive, phenotype-negative arrhythmogenic cardiomyopathy athletes

A number of studies involving carriers of pathogenic desmosomal mutations, predominantly plakophilin-2 (PKP2) have shown that asymptomatic G+P- family members who exercise regularly are more likely to fulfil the criteria for the diagnosis, and develop potentially fatal arrhythmias and heart failure compared with sedentary G+P- counterparts.[86,87] Based on these reports, exercise recommendations in athletes who are G+P- with pathogenic desmosomal variants, are identical to those assigned in athletes with overt AC.

Recommendations

See (*Table 7*).

Athletes with isolated ECG abnormalities

Asymptomatic athletes with isolated ECG abnormalities suggestive of cardiac pathology (such as ST-segment depression, T-wave inversion, and pathological Q waves) in the absence of positive family history of SCD/CA or structural features of a cardiomyopathy on imaging tests deserve special attention. Several observations in athletes suggest that these ECG abnormalities, particularly T-wave inversion in inferior and lateral leads, are harbingers for the development of overt cardiomyopathies over the medium to long-term follow-up.[15,16,93] These athletes should be comprehensively evaluated with CMR, exercise stress test and 24-h Holter ECG monitoring and clinical evaluation of first-degree relatives if possible, to exclude the possibility of cardiomyopathy.[43,94]

Recommendations

See (*Table 8*).

Athletes with cardiomyopathy and implanted cardioverter defibrillator

The efficacy of the ICD in aborting SCD/CA in high-risk individuals with cardiomyopathy has led to several young active being implanted for primary and secondary prevention. A significant proportion of such individuals aspire to continue engaging in team and individual sport at competitive and recreational level and the issue of safe sport participation in ICD recipients has become highly relevant.

The risks associated with sports participation in athletes with ICDs was assessed in the multinational, prospective ICD Sports Safety Registry[34,95] which enrolled 440 participants, including a substantial proportion of patients with HCM ($n = 75$, 17%), and ARVC ($n = 55$, 13%). After a mean follow-up period of 4 years, there were no arrhythmic deaths, externally resuscitated tachyarrhythmias during sports participation, or injury resulting from arrhythmia-related syncope or shock during sports. These results suggest that exercise and sport participation are feasible and safe in cardiomyopathy patients with ICD. Medium-term data of this registry suggest that among clinical and demographic variables associated with receiving appropriate shocks during competition/practice, the most relevant was the presence of ARVC.[95]

A measure of caution regarding sport participation in patients with cardiomyopathies is, however, justified considering that more participants received shocks during competition/practice or physical activity than at rest (20% vs. 10%; $P < 0.001$) and specifically, the proportion of appropriate shocks was greater during competition or other physical activity than during rest (11% vs. 6%; $P = 0.005$). Indeed, of 51 subjects who received shocks during sports, 20 decided to quit their sport practice. Finally, there were 31 definite and 13 possible lead malfunctions (10% of the overall cohort).[95]

In conclusion, athletes with cardiomyopathies and ICDs may participate in competitive sports without adverse events in the medium term; however, one in five will receive both appropriate and inappropriate shocks.[95]

Evaluation of individuals with cardiomyopathy and ICD who are willing to participate in competitive sport should be preferentially performed in experienced centres.

Table 8 Recommendations for athletes with isolated ECG abnormalities

	Class/level of evidence
Athletes with a markedly abnormal ECG who do not reveal any other features of cardiomyopathy should not be considered as affected by cardiomyopathy and may participate in all competitive sport. However, given the potential for developing a patent cardiomyopathy later on life, it is important that these individuals are assessed periodically (annually during adolescence and young adulthood) and educated with respect to the potential development of cardiac symptoms.	Class IIa/Level C

Table 9 Recommendations for athletes with cardiomyopathy and ICD

	Class/level of evidence
1. The indications for an ICD in competitive athletes should not differ from the general population with a diagnosis of cardiomyopathy. Specifically, the desire of the athlete with cardiomyopathy to compete should not constitute a primary (or unique) indication for ICD implantation.	Class IIa/Level C
2. Recommendation for sports participations in an individual with an ICD should be based on the fact that the arrhythmogenic substrate of the cardiomyopathy remains unaltered and the ICD does not prevent insurgence of a malignant arrhythmia, especially during intensive exercise, although it will prevent SCD/CA.	Class IIa/Level C
3. Participation in competitive sports may be considered in the individual patient with cardiomyopathy and ICD, after careful consideration of the type of underlying cardiomyopathy (for instance, AC represents reason for contraindication). The decision to participate in competitive sport should be made after full disclosure of the risks of sport participation with ICD, including the likelihood of appropriate/inappropriate shocks, leads failure, and the device-related trauma.	Class IIb/Level C

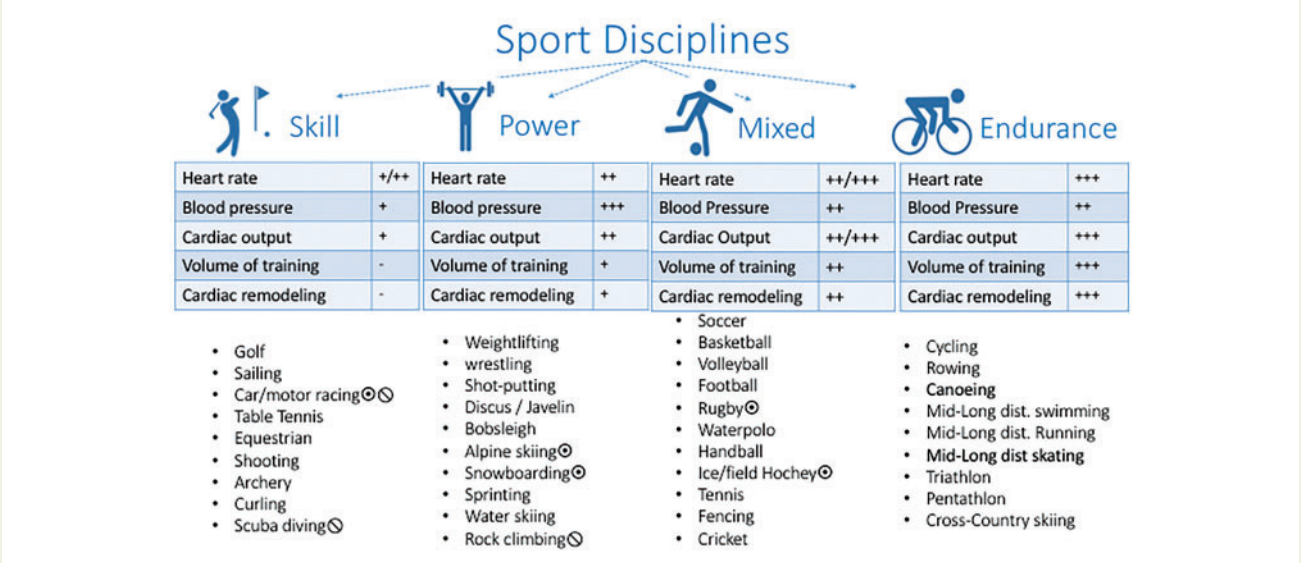

Figure 1 A schematic representation of the four different types of sport disciplines are shown. The common haemodynamic changes and cardiac remodelling occurring as a consequence of long-term training are indicated for each type of sport. Symbols: ⊙ indicates sport with increased risk of bodily collision. Ø indicates sport with intrinsic risk of serious harm or death for athlete and/or spectators in the event of syncope. Adapted and modified from Pelliccia *et al.*[6]

Recommendations

Advising participation in exercise programmes and sport in patients with ICD necessitates a comprehensive and clear explanation, and assurance of an understanding on of the whole spectrum of exercise-related risks behalf of the candidate (*Table 9*).

Given the concerns about lead problems in the longer term, there is currently a large interest in the applicability of subcutaneous-ICD devices in athletic patients. Initial experience with subcutaneous-ICD shows a high efficacy, although the current lack of data limits our understanding on long-term benefits and pitfalls.[96,97]

With regard to type of sport, selected patients with cardiomyopathy and ICD may participate in all sport, with exception of sports with bodily contact, such as rugby, American football and martial arts in order to avoid damage to the generator box and the cardiac leads (see *Figure 1*). For other sports with a smaller collision risk, such as soccer, basketball, and hockey, some have advocated padding of the ICD implantation site, although effectiveness of these protection systems has not been proven.[97]

Patients with cardiomyopathy and ICD should avoid sports where transient syncope from an arrhythmia or the jolt of a shock may cause serious injury or death, such as mountain climbing, surfing, motor racing, or deep-sea diving (*Figure 1*).[97,98]

Prescription for leisure-time physical activity in patients with cardiomyopathy

The clinician is frequently confronted with the request of designing leisure-time exercise programmes for young individuals with cardiomyopathies, who aspire to maintain a physically active lifestyle. The precise risk of habitual non-competitive sport activities in young and adult patients with cardiomyopathy is poorly defined. It has been argued that a certain degree of risk associated with participation in amateur and leisure-time sport activities will likely persist. However, given the plethora of benefits of moderate exercise on the cardiovascular system and longevity of life, it would seem appropriate to prescribe exercise programmes in all individuals with cardiomyopathy.[98–100]

In recent years, there has been accumulating scientific evidence to support the benefits of regular exercise programmes in patients with cardiomyopathy. A recent randomized clinical trial demonstrated that patients with HCM may safely engage in moderate-intensity, regular exercise programmes.[101] Just after 16 weeks of an active lifestyle, a mild (+6%), statistically significant increase in VO_2 max and exercise capacity was observed. Of relevance, there were no episodes of sustained ventricular arrhythmia, CA, or appropriate defibrillator shocks even in patients with a moderate or high-risk profile.[101] Klempfner *et al.*[102] reported similar results in symptomatic HCM patients enrolled in a supervised cardiac exercise programme. After a short-term period (average, 40 h exercise) the vast majority of patients (80%) reported subjective improvement in their clinical condition and the New York Heart Association class improved from baseline by >1 grade. Functional capacity, assessed by the change in maximally attained METs, improved by 46% and during the 12 month follow-up period, none of the patients experienced clinical deterioration, significant adverse events or ICD discharges.[102]

It is reasonable that the actual risk associated with mild to moderate exercise programmes is less than previously anticipated and, therefore, the majority of patients with HCM should not be deprived of the many cardiovascular, metabolic and psychological benefits afforded by regular exercise. Regular exercise improves functional capacity and may reduce the impact of excessive body weight on symptom progression in HCM patients, in whom obesity promotes worsening of the disease.[103]

Although it is well recognized that intensive exercise programmes aggravate the clinical course in patients with AC, there also is evidence that recreational and low-intensity exercise may not have an adverse impact on the natural history of the disease. In the North American multidisciplinary study of ARVC,[88] there were no differences in RV volumes between sedentary patients and those participating in recreational sports and there were no differences in age at clinical presentation or incidence of ventricular tachyarrhythmias or SCD/CA between inactive patients and patients who participated in mild recreational physical activities. Therefore, recreational exercise might not be as deleterious as competitive sport and current evidence justifies a less restrictive approach to leisure exercise in these patients.

Modality of leisure-time exercise programmes

Patients with cardiomyopathy should not participate in exercise that requires pursuit of excellence or pressure to excel against others and involves explosive bouts of intensive exercise, apart from the exceptions as previously stated. It is necessary for clinicians to individualize exercise prescription, balancing the clinical status of the patient with the type, intensity and frequency of the physical and/or sport activity being considered. Exercise prescription should preferentially be guided by exercise testing, aiming for a submaximal, well-tolerated level of exercise intensity.

There are specific recommendations that may reduce the arrhythmogenic risk in patients with cardiomyopathy.[98–100] Patients should be fully informed of the clinical peculiarities of the disease, including the modalities of clinical presentation, and warned about the incidence of symptoms that may occur in association with exercise. Patients should be advised to start exercise sessions with a warm-up period and at the end of the session an appropriate cool-down period is also recommended.

Exercise programmes characterized by a progressive increase in intensity should be closely monitored by the physician (ideally in contact with the coach), in order to adapt the intensity, duration and frequency of training to the individual cardiac capabilities. Patient should avoid exercising in adverse environmental conditions without prior acclimatization.

In the choice of indoor activities, patients with cardiomyopathies should be encouraged to exercise in environments equipped with an automatic defibrillator and personnel trained in its use. Finally, patients with cardiomyopathy should preferentially avoid high-intensity free weight lifting, to reduce the risk of injury in the event of syncope.

Myocarditis

Myocarditis is defined as an inflammatory process of the myocardium, with histological evidence of myocyte degeneration and necrosis of

non-ischaemic origin, associated with inflammatory infiltration.[104,105] Case series in athletes have established myocarditis as a potential cause of SCD/CA, in 2–8% of cases.[9–11] Myocarditis may present with arrhythmia, heart failure, or simulate myocardial infarction. The illness may be preceded by coryzal symptoms or diarrhea.[104,105] Rarely, athletes may present non-specific symptoms, such as fatigue which may elude the diagnosis.

Serum cardiac biomarkers for inflammation are elevated and should be measured in suspected cases.[104,105]

12-lead ECG and 24-h ECG monitoring

The ECG abnormalities include frequent and/or complex ventricular and/or supraventricular arrhythmia, ST-segment alterations, T-wave inversion and occasionally, LBBB or atrioventricular block.[106] Individuals presenting with heart failure may show low QRS voltages particularly in the presence of a pericardial effusion.

Echocardiography

The LV may be mildly dilated with thin myocardial walls, resembling DCM, or there may be a non-dilated cavity with increased myocardial wall thickness due to myocardial oedema.[107] Regional wall motion abnormalities are recognized and LV global systolic function may range from almost normal to severely depressed. The presence of severe LV dysfunction may predict severe clinical outcome.[108] Concomitant pericardial effusion suggests pericardial involvement.

Cardiovascular magnetic resonance

Cardiovascular magnetic resonance has excellent sensitivity for detecting myocarditis and can identify hyperaemia, inflammation oedema, and/or focal scar.[109,110] Furthermore, the identification of LGE may confer prognostic information, since it is a strong independent predictor of events during follow-up.[109–113]

Histology

Endomyocardial biopsy (EMB) is considered the gold-standard for the diagnosis of myocarditis. It allows a definitive diagnosis, based on immune-histochemical testing, and the possible identification of viral genome in the cardiomyocytes through the polymerase chain reaction analysis.[114] However, EMB is not usually performed for clinical diagnosis, but in selected cases, when therapeutic strategies are debated and the result is likely to influence the treatment or prognosis of patients, such as in the case of suspected giant cell myocarditis of cardiac sarcoidosis.[115]

Risk stratification

There are robust data linking the presence and persistence of myocarditis to SCD/CA in young individuals. A murine model of coxsackie B3 myocarditis showed that daily exercise training increased viral titers, worsened cardiomyopathy, and increased the likelihood of death.[116] In one of the first pathological surveys of SCD in a cohort of U.S. military recruits, myocarditis was reported as the most frequent disorder associated with death during strenuous physical exertion.[117] More recent case series in athletes have established myocarditis as a definite risk factor for SCD.[9–11,105,118]

The risk of sudden death does not always correlate with the severity of myocardial inflammation and serum concentrations of cardiac troponin.[119] Left ventricular dysfunction is the major prognostic determinant of an adverse event,[11,120] however, SCD/CA may occur despite normal LV function and is mostly related to tachyarrhythmias.

There is no specific test that can establish complete resolution of the inflammatory process in myocarditis. Athletes in whom the

Table 10 Recommendations for athletes with myocarditis

	Class/level of evidence
1. General consensus exists that athletes with diagnosis of myocarditis should be restricted from exercise programmes for a period of 3–6 months, according to the clinical severity and duration of the illness, LV function at onset, and extent of inflammation on the CMR. This time period is considered appropriate to ensure clinical and biological resolution of the disease.[3,4,105,118–120]	Class IIb/Level C
2. Individuals with previous myocarditis have an increased risk for recurrence and silent clinical progression of the disease. Therefore, athletes with previous myocarditis should undergo a periodical re-assessment, particularly within the first 2 years.	Class IIa/ Level C
3. It is reasonable for athletes to resume training and competition after a myocarditis if all of the following criteria are met: (1) LV systolic function has returned to the normal range. (2) Serum biomarkers of myocardial injury have normalized. (3) Clinically relevant arrhythmias, such as frequent or complex repetitive forms of ventricular or supraventricular arrhythmias are absent on 24-h ECG monitoring and exercise test.	Class IIa/ Level C
4. The clinical significance of persistent LGE in an asymptomatic athlete with clinically healed myocarditis is unknown, however, myocardial scar is a potential source of ventricular tachyarrhythmias.[111–113] At present, it seems reasonable for these athletes to resume training and participate in competitive sport if LV function is preserved and in the absence of frequent or complex repetitive forms of ventricular or supraventricular arrhythmias during maximal exercise and on 24-h ECG monitoring (including session of training/competition). Asymptomatic athletes with LGE, however, should remain under annual clinical surveillance.	Class III/ Level C

findings of acute inflammation have resolved may still harbour the risk for arrhythmias related to the resultant myocardial scar. Therefore, the interval between initial assessment and retesting before resumption of sports will vary on individual basis depending on the severity of the initial illness and morphological sequelae.

Recommendations

See (*Table 10*).

Pericarditis

Pericarditis is defined as an inflammatory process of the pericardium, which may also affect the sub-epicardial layers of the myocardium.[105,121]

Clinical presentation

Pericarditis is usually preceded by upper respiratory or gastrointestinal symptoms, and clinical presentation may include chest pain, increased fatiguability or exertional dyspnoea. In developed countries, viruses are the most common aetiological agents for pericarditis, whereas tuberculosis is a frequent cause of pericardial disease in developing countries.[121,122] Pericarditis and myocarditis may coexist in 20–30% of patients due to overlapping aetiologies. The presence of concomitant myocarditis is indicated by elevation of serum cardiac troponins and myocardial oedema on CMR. Patients with pericarditis concomitant with myocarditis (usually referred as myo-pericarditis) have potential for a higher rate of complications including LV dysfunction.

Serum cardiac biomarkers for cardiac inflammation and necrosis should be measured to exclude the possibility of myocardial involvement.[122]

12-lead ECG

A spectrum of ECG abnormalities may be present typically including a new widespread ST-segment elevation, or PR interval depression in the acute phase.

Echocardiography/cardiac magnetic resonance

Often a pericardial effusion is present at the onset of the disease, with increased reflectivity and separation of the thickened pericardial layers. Athletes with pericarditis and raised biomarkers for cardiac damage should be investigated with CMR to assess the extent of myocardial involvement.

Risk stratification

Individuals with pericarditis usually have an excellent prognosis with complete resolution of the pathological process.[105,121,122] However, patients with idiopathic acute pericarditis and certain features at presentation (temperature >38°C), subacute course, large pericardial effusion, and resistance to non-steroidal anti-inflammatory drugs comprise a subset of patients with a more guarded prognosis and a greater risk for recurrence and progressing to pericardial constriction.[105,120,122]

Recommendations

See (*Table 11*).

Cardiac classification of sport activities

Sports activities are here classified according to the cardiovascular changes associated with the exercise training and the long-term impact on cardiac morphology. In this regard, sport disciplines may be schematically divided in four major groups, i.e. skill, power, mixed, and endurance.[6]

Specifically, *skill sports* are disciplines with prominent technical characteristics, where the achievement is mostly based on the athlete's neuro-muscular coordination and skill (i.e. low dynamic, low static). The cardiovascular response to regular exercise is characterized by an increase in heart rate, which may be substantial, with only modest changes in blood pressure and cardiac output. The long-term cardiac adaptation to these disciplines is characterized by *minimal or no morphologic cardiac remodelling*.

Table 11 Recommendations for athletes with pericarditis

	Class/level of evidence
1. Athletes with pericarditis should not participate in competitive sports during the acute phase. Athletes can return to sport activity only after complete resolution of the active disease. The time period of 3 months is considered appropriate to ensure a complete clinical and biological resolution of the disease, but shorter period (at least 1 month) may be considered in selected cases with only mild clinical picture and prompt resolution.	Class III/Level C
2. It is reasonable to return to play if the serum biomarkers have normalized, LV function is normal and there are no resting, or exercise-induced frequent/complex ventricular arrhythmias detectable on 24-h ECG monitoring or exercise ECG.	Class IIa/Level C
3. Athletes with concomitant myocardial involvement should be treated in accordance with the recommendations for myocarditis.	Class IIa/ Level C
4. Asymptomatic athletes with small pericardial effusion, detected incidentally by imaging testing, but without clinical, biochemical and CMR evidence of myocardial inflammation, should not be considered as affected by myopericarditis and should not be restricted from sport participation. A periodical surveillance is however advisable.	Class IIa/ Level C

Power sports are disciplines with a prominent muscle strength component, where achievement is due to the generation of explosive muscle power (i.e. high-static exercise). The cardiovascular response to power exercise is characterized by substantial increases in blood pressure and heart rate for several, short repetitive bursts. The long-term cardiac adaptation to power disciplines is characterized by an *increase in LV wall thickness and modest change in LV cavity size*.

Mixed sports present alternate phases of work (either dynamic and/or static) and recovery periods. Typical examples are ball games and the team disciplines. The duration and intensity of work varies greatly according to the type of discipline, the role of the athlete and the trend of the game. The cardiovascular response to mixed exercise includes phasic increases in heart rate and blood pressure, which may reach near-maximum values, alternating with recovery phases, when the heart rate, blood pressure and cardiac output decrease. The long-term cardiac adaptation to mixed disciplines is characterized by an *increase in LV cavity size and modest change in LV wall thickness*.

Endurance sports are disciplines characterized by prolonged and intensive dynamic exercise (i.e. high-dynamic, often associated with high-static exercise), where the achievement is related to the athlete's capability to attain and maintain very high cardiac output, through persistent increase in heart rate and blood pressure. Duration of the haemodynamic load is usually prolonged for 1–2 h, with habitually 6–12 session a week in elite athletes. Cardiac remodelling with *increased LV cavity size and wall thickness* characterizes the cardiac adaptation to endurance disciplines. The degree of cardiac remodelling may even be marked, depending on the type of discipline, the gender, ethnic origin and body size, and composition of the endurance athletes.

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