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34 Guidelines for Sports Practice in Athletes with Cardiovascular Disease

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It is a widely accepted clinical perception, occasionally substantiated by scientific evidence, that athletes with underlying cardiovascular disease (CVD) have an increased risk of sudden cardiac death (SCD) or clinical deterioration when compared with relatively sedentary individuals, by virtue of the demands placed on the CV system during regular intensive exercise regimes and sports participation [1]. Therefore, an expert consensus document is required to guide physicians and cardiologists in the evaluation of athletes with CV abnormalities and to recommend sports activities that can be safely performed. Two such documents currently exist, namely the American Heart Association/American College of Cardiology (AHA/ACC) guidelines, from the United States [2], and the European Society of Cardiology (ESC) Consensus Recommendations [1], from Europe. Both were originally published in 2005. Since then, the US guidelines have been updated and the European guidelines are in the process of being updated. Whilst most recommendations in the two documents are similar, it is important to appreciate that they draw from largely different cultural, social and legal backgrounds and, therefore, in some instances present different approaches to disqualification decisions and implications for clinical practice. It must be emphasised that both sets of recommendations are based on published scientific evidence where available, but given the scarcity and inconsistency of scientific investigations concerning the effect of regular sporting activities on the pathophysiology and clinical course of several CVDs [1], they are largely reliant on circumstantial evidence and the expert opinion of the respective consensus panels. Given that risk of clinical deterioration or SCD is difficult to predict, both guidelines are conservative in principle, in an attempt to encompass all preventable deaths, and acknowledge that many athletes who may never suffer such events will be unnecessarily restricted. Over the last 10 years, few studies have been conducted that would challenge the current recommendations, but efforts should be made wherever possible to tailor precise advice to the individual.

Competitive athletes are defined as individuals of young and adult age, either amateur or professional, who are engaged in exercise training on a regular basis and who participate in official sports competition [1].

This chapter will address each group of CVDs in turn, compare the US and European recommendations and explain the underlying rationale behind them. It includes relevant evidence that has come to light since their original publications.

Reference will be made to the Mitchell classification (Figure 34.1), which divides various sports activities into two main categories (dynamic and static) and according to intensity (low, moderate and high) [3]. The classification is intended to provide a schematic indication of the CV demand associated with different sports and to identify those disciplines associated with increased risk of bodily collision or syncope, which should be avoided in certain cardiac patients [1].

IOC Manual of Sports Cardiology, First Edition. Edited by Mathew G. Wilson, Jonathan A. Drezner and Sanjay Sharma.
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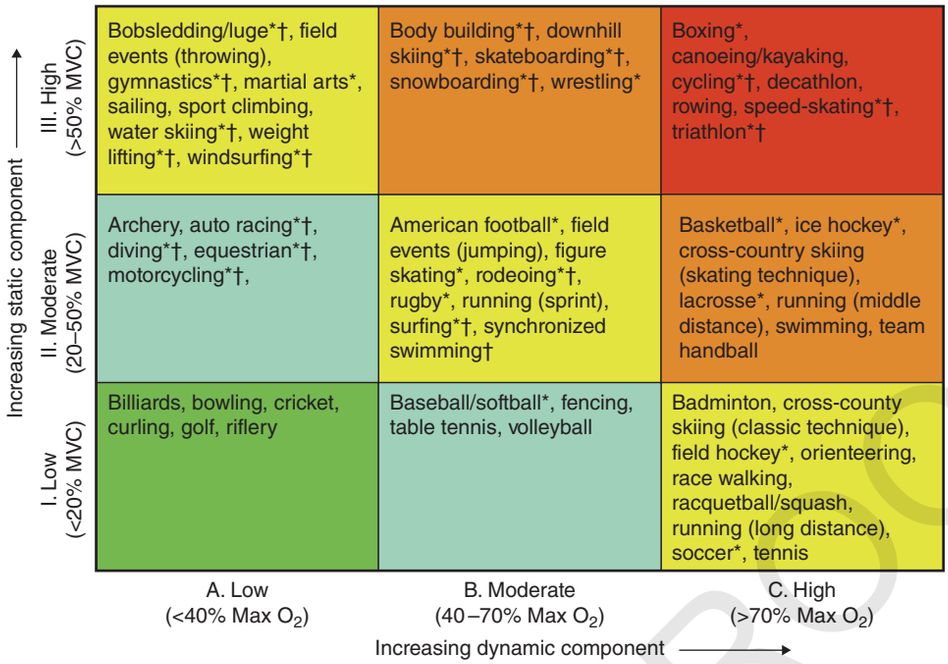

Figure 34.1 Sports classification based on peak static and dynamic components achieved during competition. The lowest total CV demands (cardiac output and blood pressure, BP) are shown in green, and the highest in red. Max O_2, maximal oxygen uptake; MVC, maximal voluntary contraction. *Danger of bodily collision. †Increased risk if syncope occurs (*Source*: Mitchell et al. [3]. Reproduced with permission of Elsevier)

Cardiomyopathies, Myocarditis and Pericarditis

The ESC and AHA/ACC recommendations for competitive athletes with cardiomyopathies, myocarditis or pericarditis are summarised in Table 34.1.

Hypertrophic Cardiomyopathy

Hypertrophic cardiomyopathy (HCM) is a relatively common genetically transmitted primary myocardial disease, with a prevalence of 1 in 500 in the general population and is the most common cause of unexpected SCD in young people, including competitive athletes [4]. Mutations in the genes encoding sarcomeric contractile proteins are responsible in 50–60% of cases. The condition is characterised by unexplained left ventricular hypertrophy (LVH) and a predilection to fatal ventricular arrhythmias, but is highly heterogeneous with respect to cardiac morphology, clinical penetrance and natural history. Although most individuals with HCM are unable to augment stroke volumes sufficiently long to compete in sports at an elite level, due to a small LV cavity size, impaired diastolic function, dynamic LV outflow tract (LVOT) obstruction, microvascular ischaemia or a combination of these factors, some are completely asymptomatic and may be able to demonstrate excellence in sports disciplines with an explosive start–stop component, such as basketball, football and American football. Unfortunately, their index presentation may be SCD, during or immediately after exercise.

The ESC guidelines restrict any individual with HCM and cardiac symptoms or risk factors for SCD from all competitive sports. Risk factors for SCD include prior cardiac arrest, ventricular tachycardia (VT), a family history of SCD, syncope, an LV wall thickness ≥30 mm and an abnormal BP response to exercise. HCM individuals without symptoms or risk factors are restricted to low-dynamic, low-static (class IA) sports, with annual follow-up.

The ACC guidelines restrict all HCM individuals to low-intensity (class IA) sports.Given that most class IA sports are relatively sedentary, the authors believe that all patients with HCM should be permitted to participate in class I sport irrespective of risk factors.

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Condition	ESC	AHA/ACC
HCM with symptoms or high risk	No competitive sports	Low-dynamic, low-static sports (IA)
HCM low risk	Low-dynamic, low-static sports (IA)	Low-dynamic, low-static sports (IA)
HCM G+/P–	No competitive sports, only recreational sports[a]	All competitive sports
ARVC	No competitive sports	Possibly low-dynamic, low-static sports (IA)
ARVC G+/P–	No competitive sports	Low-dynamic, low-static sports (IA)
DCM with symptoms or high risk	No competitive sports	Possibly low-dynamic, low-static sports (IA)
DCM low risk	Low–moderate-dynamic and low-static sports (IA, IB)	Possibly low dynamic low static sports (IA)
Myocarditis and pericarditis	No competitive sports for 6 months, then all competitive sports upon satisfactory evaluation	No competitive sports for 6 months, then all competitive sports upon satisfactory evaluation

Table 34.1 Summary of selected differences between ESC and AHA/ACC recommendations for competitive athletes with cardiomyopathies, myocarditis or pericarditis

[a] Revised recommendations will permit such individuals from competing in all sports.

HCM, hypertrophic cardiomyopathy; ARVC, arrhythmogenic right ventricular cardiomyopathy; DCM, dilated cardiomyopathy.

Genotype-Positive/Phenotype-Negative Individuals

The natural history of genotype-positive/phenotype-negative (G+/P−) individuals is not fully understood. Previous reports suggest that such individuals exhibit a high concentration of serum biomarkers of collagen synthesis, late gadolinium enhancement on cardiac MRI [5], expanded extracellular volume detectable by T1 mapping [6] and subtle impairment of diastolic dysfunction [7]. The impact of exercise on the development of the overt phenotype or the arrhythmogenic substrate is unclear, so the ESC recommendations adopt a cautious view and advise that competition be confined to class IA sports in affected athletes. However, over the past few years emerging studies have shown that G+/P− individuals have a benign clinical course, with low penetrance and absence of symptoms or adverse events [8, 9]. Based on these considerations, it is recommended that all G+/P− individuals be assessed comprehensively with a cardiac MRI, exercise test and Holter monitor to exclude the broader phenotypic features of HCM. In the absence of abnormal investigations, the athlete may compete in all sports, but they should be kept under annual surveillance as follow-up studies in this cohort are relatively short.

According to the AHA/ACC guidelines, G+/P− individuals can participate in all competitive sports, provided that they are asymptomatic and do not have a family history of SCD [10]. In the light of new evidence, future ESC guidelines are likely to become more liberal and take the same view.

Athletes with Marked Repolarisation Changes but a Structurally Normal Heart

On occasion, and particularly among athletes of African or Afro-Caribbean origin, the presence of deep T-wave inversion on the electrocardiogram (ECG) may raise the possibility of HCM. In such circumstances, if the echocardiogram cannot confirm the diagnosis, comprehensive assessment with cardiac MRI, exercise stress test and a 24-hour Holter monitor is necessarily to exclude the broader phenotypic features of HCM. In the absence of a diagnosis, both sets of recommendations permit competition in all sports, but both suggest that the athlete should be evaluated annually whilst competing, since such repolarisation changes may reflect the onset of cardiomyopathy in the future [1, 10].

Arrhythomogenic Right Ventricular Cardiomyopathy

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a primary myocardial disease characterised histologically by fibro-fatty replacement of the RV myocardium and clinically by life-threatening ventricular tachyarrhythmias in young individuals. ARVC represents the commonest cause of SCD in young athletes in Italy [1]. The ESC guidelines restrict athletes with a definite diagnosis of ARVC from all competitive sports [1].

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The ACC guidelines state that all individuals with a definite or probable diagnosis of ARVC should be excluded from most competitive sports, with the possible exception of low-intensity (class IA) sports [10]. The authors believe that the American guidelines are more pragmatic and that there is no evidence to disqualify affected athletes from class IA sports.

G+/P– Individuals

Data from animal studies show that plakophilin-deficient mice develop the ARVC phenotype with intensive exercise [11]. Recently, a plethora of publications have shown that asymptomatic gene-positive family members of first-degree relatives with overt ARVC who exercise regularly are more likely to go on to develop overt disease, potentially fatal arrhythmias and heart failure, as compared to sedentary gene-positive family members [12-15]. The AHA/ACC guidelines permit low-intensity (class IA) sports [10]. By virtue of the recommendations for G+/P– HCM individuals, the authors assume that the ESC guidelines restrict G+/P– athletes with ARVC from all competitive sports [1].

Dilated Cardiomyopathy

Dilated cardiomyopathy (DCM) is a myocardial disease characterised by LV dilatation and impaired systolic function. DCM includes disorders that are familial/genetic in origin; that are secondary to infection or inflammation, exposure to toxic substances or metabolic disorders; or that are of idiopathic origin. Although not frequent, DCM can cause SCD in athletes [1].

The ESC guidelines restrict athletes with DCM and symptoms or risk factors for SCD from all competitive sports. Risk factors for SCD with DCM include an ejection fraction (EF) <40%, complex ventricular arrhythmias and an abnormal BP response to exercise. Athletes with DCM but no symptoms or risk factors are permitted to participate in low–moderate-dynamic and low-static (class IA and IB) sports, with annual follow-up [1].

AHA/ACC guidelines acknowledge that whilst little information is available regarding DCM and other myocardial diseases, it is prudent to exclude athletes with DCM from most competitive sports, with the possible exception of low-intensity (class IA) sports in selected cases [10]. The authors consider the AHA/ACC recommendations to be more pragmatic.

Left Ventricular Noncompaction

Left ventricular noncompaction (LVNC) is a relatively novel yet unclassified cardiomyopathy that is characterised by prominent trabeculation within the LV myocardium, separated by deep recesses. The disease is characterised by progressive systolic heart failure, and a predisposition to serious ventricular arrhythmias and systemic thromboembolism. The diagnosis of LVNC can be difficult in athletes because LV trabeculations are common in sports players (see Chapter 21). Based on the available literature, we would not consider an asymptomatic athlete with incidental features of LVNC to necessarily harbour a cardiomyopathy in the absence of symptoms, family history, abnormal ECG patterns and impaired LV function. In athletes with impaired LV function and T-wave inversion, we propose abstinence from competitive sports involving medium- or high-intensity exercise, as for all other cardiomyopathies.

Myocarditis

Myocarditis is defined as an inflammatory process of the myocardium, with histological evidence of myocyte degeneration and necrosis of nonischaemic origin, associated with inflammatory infiltration [1].

The ESC guidelines state that athletes with active myocarditis should be restricted from competitive sports for a convalescence period of 6 months following the onset of clinical manifestations. Athletes may then return to all competitive sports, provided there are no remaining symptoms, LV function has returned to normal and there are no arrhythmias as evaluated by ECG, echocardiography and exercise testing [1]. The AHA/ACC guidelines are almost identical in this respect, but they add that serum markers of inflammation and heart failure must normalise before the athlete returns to training and competitive sport [10].

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The convalescence period of 6 months is based purely on consensus advice and is not supported by any clinical evidence. It may be possible for athletes to return to competitive sport as early as 3 months after the onset of symptoms, if LV function is completely normal and there are no resting or exercise-induced arrhythmias. Both recommendations have also relied on the echocardiogram, whereas the gold standard imaging test for myocarditis is cardiac MRI, which has the advantage of detecting the extent of myocardial inflammation and fibrosis. In this regard, there are no data relating to the management of an athlete with preserved LV size and function who is free of arrhythmias but is rendered with permanent myocardial scarring. Further studies in this field, particularly relating to the quantity of scar burden, are prudent to inform future recommendations, but the authors would advise that all athletes with scarring should be assessed annually with an echocardiogram and an exercise stress test.

Pericarditis

Pericarditis is an inflammatory process of the pericardium which may also affect the subepicardial layers of the myocardium.

The ESC guidelines for athletes with pericarditis are the same as those for myocarditis: restriction for 6 months, with return to all competitive sports upon normalisation of ECG, echocardiography and exercise testing [1]. The AHA/ACC guidelines add a requirement for normalisation of serum markers of inflammation before the athletes return to play. Also, the presence of chronic pericardial disease resulting in constriction disqualifies athletes from all competitive sports [10].

As with myocarditis, the authors believe that a convalescence period of 3 months may suffice in some athletes.

Primary Electrical Disease

The ESC and AHA/ACC recommendations for competitive athletes with primary electrical disease are summarised in Table 34.2.

Congenital Long QT Syndrome

Congenital long QT syndrome (LQTS) can be a difficult clinical diagnosis to secure definitively, and a diagnostic algorithm, such as the Priori–Schwarz score, is best employed for this purpose (see Chapter 26) [16]. An increasing proportion of asymptomatic individuals with genetically proven LQTS are found to

Condition	ESC	AHA/ACC
Long QT syndrome	No competitive sports	No competitive sports until asymptomatic on treatment for 3 months and precautionary measures taken Then all sports except swimming/diving for LQT1
Long QT syndrome G+/P–	No competitive sports	All sports, except swimming/diving
Brugada syndrome	No competitive sports	All sports with precautionary measures taken
CPVT	No competitive sports (as for malignant VT)	Possibly low-dynamic, low-static sports (IA)
WPW	All competitive sports 3 months after successful catheter ablation	Low-risk accessory pathway: all competitive High-risk pathway: all competitive sports 4 weeks after catheter ablation
ICD patients	No competitive sports	Possibly low-dynamic, low-static sports (IA)

Table 34.2 Summary of selected differences between ESC and AHA/ACC recommendations for competitive athletes with primary electrical disease

LQT3, type 3 long QT syndrome; LQT1, type 1 long QT syndrome; CPVT, catecholaminergic polymorphic ventricular tachycardia; WPW, Wolff–Parkinson–White syndrome; ICD, implantable cardioverter-defibrillator.

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have normal resting ECGs with a heart rate–corrected QT interval (QTc) by Bazette's formula of <460 ms (G+/P− LQTS). To add complexity, a QTc of 440 ms, used in the past as an upper limit of normal, is found far too frequently in normal individuals (>25%) [17]. In general, a QTc of >470 ms in males or >480 ms in females requires further investigation as to whether a congenital or acquired cause of QT prolongation is present. Exercise testing and 24-hour Holter monitoring can be useful in making a diagnosis. Gene testing for responsible cardiac ion channel mutations can identify an abnormality in 75% of LQTS sufferers [17]. All affected individuals should receive treatment with a nonselective beta blocker, and this in itself may result in disqualification from particular sports.

According to the ESC, competitive sports are contraindicated in all athletes with LQTS, including those who are G+/P− [1]. The AHA/ACC guidelines restrict only athletes with symptomatic or electrocardiographically manifest LQTS from competitive sports. However, once rendered asymptomatic on therapy for 3 months, competitive sports participation can be considered provided precautionary measures are taken, including avoidance of QT-prolonging drugs, electrolyte and hydration replenishment, avoidance of hyperthermia, acquisition of a personal automated external defibrillator as part of the athlete's personal safety gear and establishment of an emergency action plan [18]. Asymptomatic G+/P− athletes are allowed to participate in all competitive sports with the appropriate precautionary measures listed in the text. It is acknowledged that although the risk of SCD may be higher than baseline, there are insufficient compelling data to justify excluding these individuals from competitive sports [17]. However, due to a strong association between SCD in sports involving swimming/diving and type 1 LQTS (LQT1), athletes with G+/P− LQT1 should refrain from competitive sports with swimming/diving.

Brugada Syndrome

Brugada syndrome is a genetic condition characterised by a typical ECG pattern in anterior precordial leads V1–V3, with a 'coved-type' ST segment elevation >2 mm. It may be spontaneous or induced by pharmacological sodium channel blockade. It is associated with arrhythmia-related syncope or cardiac arrest. Up to 20% of affected individuals carry a gene mutation in the sodium channel gene, *SCN5A* [19]. Most events occur at rest, and often at night, when sympathetic activity is withdrawn and vagal tone increased. High temperature is known to increase ST segment elevation and is associated with event risk. However, no clear relationship between exercise and arrhythmia has been found (see Chapter 27).

Despite this, the ESC guidelines restrict all athletes affected with Brugada syndrome from competitive sports [1]. The AHA/ACC guidelines acknowledge the lack of association between exercise and SCD and permit competitive sports participation provided appropriate precautionary measures and disease-specific treatments are in place and that the athlete has been asymptomatic on treatment for at least 3 months [18].

It is difficult to justify restrictions based on currently available evidence. In the future, ESC recommendations may be liberalised, allowing more competitive sports (taking into account dehydration and sporting environments that might precipitate hyperthermia). Individuals who develop a type 1 Brugada ECG pattern on exercise testing may be an exception. At present, it is difficult to justify any restriction on individuals who harbour an SCN5A mutation but do not have any evidence of the Brugada ECG pattern.

Catecholaminergic Polymorphic Ventricular Tachycardia

Catecholaminergic polymorphic VT (CPVT) is characterised by exercise-induced PVT (often with a 'bidirectional pattern'), which can degenerate into ventricular fibrillation. This condition has been linked to mutations of the ryanodine receptor and calsequestrin genes, which result in abnormal calcium release from the sarcoplasmic reticulum (SR). CPVT does not manifest any abnormalities on the resting ECG, but it requires exercise testing or epinephrine provocation to establish the diagnosis. All affected individuals require treatment with a nonselective beta blocker and consideration of an implantable cardioverter-defibrillator (ICD).

The ESC guidelines restrict all athletes with CPVT from all competitive sports, on the basis that they have a permanent substrate for malignant VT [1]. The AHA/ACC guidelines restrict all athletes from competitive sports, with the possible exception of minimal contact, class IA activities. It is also advised that affected individuals are restricted from competitive sports involving swimming. Asymptomatic sufferers, identified

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on family screening, should be treated the same way if exercise-induced or pharmacological provocation-induced VT is present. A less restrictive approach may be suitable for the G+/P− (asymptomatic, no inducible VT) athletes [18].

Wolff–Parkinson–White Syndrome

Wolff–Parkinson–White (WPW) syndrome is defined as the presence of paroxysmal arrhythmias in a patient with overt ventricular pre-excitation. The tachyarrhythmias related to WPW syndrome include atrioventricular (AV) re-entry tachycardia (either orthodromic or antidromic), atrial fibrillation and, rarely, ventricular fibrillation.

Both ESC and AHA/ACC guidelines are in agreement that symptomatic athletes with ventricular pre-excitation and atrial fibrillation or flutter should undergo catheter ablation [1, 20]. For those who are asymptomatic, a low but definite risk of SCD exists, and for this reason the ESC guidelines recommend that catheter ablation is the first-choice treatment, due to its high success rate and low risk of complications [1]. For athletes who refuse or in whom the procedure is associated with higher risks, competitive sport participation is allowed if invasive electrophysiological (EP) study demonstrates a low-risk accessory pathway with a long refractory period [1]. The AHA/ACC guidelines advocate exercise stress testing assessment of the EP properties of the accessory pathway. Specifically, abrupt and complete loss of pre-excitation during exercise denotes a low-risk pathway with a long refractory period. Those with a low-risk accessory pathway can participate in all competitive sports. If an athlete cannot be ascertained as being at low risk by stress testing, then an EP study is advocated. Those with high-risk features of the accessory pathway, with an effective refractory period ≤250 ms, should undergo catheter ablation and are then permitted to participate after 4 weeks, if follow-up EP study is satisfactory [20].

Athletes with Implantable Cardioverter-Defibrillators

Athletes with ICDs are generally considered to have cardiac diseases that are life-threatening and, therefore, represent a contraindication for competitive sports. Furthermore, rapid lunging movements of the upper limbs and bodily collision risk lead to fracture and damage to the generator box, respectively. Both ESC and AHA/ACC guidelines recommend restriction from competitive sports, although the American guidelines allow class IA sports if free from arrhythmia requiring device therapy for 3 months [1, 20]. The AHA/ACC guidelines also allow scope for participation in sports with higher peak static and dynamic components based on an individualised approach in stable athletes. Similar concerns exist in athletes with pacemakers, due to the risks of damage to a device or lead; sports with physical contact are contraindicated in ESC guidelines and cautioned in AHA/ACC guidelines [1, 20].

A new registry tracking athletes with normal LV EFs and transvenous ICDs who choose to continue participation in sports of higher classifications than IA shows low rates of cardiac events and no increased risk of device/lead malfunction [21]. These observations may provide justification in the future for more liberal recommendations for some athletes with ICDs and pacemakers. Ultimately, the underlying cardiac condition will remain the most significant factor influencing the appropriateness of sports participation.

Aortopathies, Coronary Artery Disease and Hypertension

The ESC and AHA/ACC recommendations for competitive athletes with aortopathies, coronary artery disease or hypertension are summarised in Table 34.3.

Marfan Syndrome

Marfan syndrome is an autosomal-dominant connective-tissue disorder caused by fibrillin 1 (*FBN1*) gene defects and by mutation of transforming growth factor beta receptor 2 (TGFBR2). Penetrance is complete, but there is large phenotypic heterogeneity, with variable involvement of different organs/tissues, including osteoskeletal, CV, ocular, skin, pulmonary and nervous system. The primary cause of mortality in athletes is aortic root dilatation, dissection and rupture.

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Condition	ESC	AHA/ACC
Marfan syndrome	Full phenotype or positive genetic test: no competitive sports	Aortic root dilatation, moderate-to-severe mitral regurgitation, left ventricular (LV) systolic dysfunction (EF < 40%) or family history of aortic dissection: no competitive sports
	Incomplete phenotype, no family history, no gene mutation or family history alone: all competitive sports with annual follow-up	None of the above: low dynamic, low–moderate-static sports (IA, IIA)
Ischaemic heart disease	High probability of events: no competitive sports	Unstable IHD: no competitive sports
	Low probability of events: low–moderate-dynamic and low-static sports (IA, IB)	Stable, clinically manifest IHD: low-dynamic and low–moderate-static sports (IA, IIA)
		Clinically concealed IHD with low probability of events: all competitive sports
Anomalous coronary artery origins	–	No competitive sports with possible exception of low-dynamic, low-static class IA sports.
		Postsurgical correction: all competitive sports, upon satisfactory evaluation after 3 months
Hypertension	Low risk: all competitive sports with annual follow-up	Mild hypertension with no target organ damage: all competitive sports
	Moderate risk: all sports except high-static, high-dynamic sports (IIIC), with annual follow-up	Moderate hypertension without target organ damage: all sports except high-static sports (IIIA–C), until hypertension controlled
	High risk: all sports with exclusion from high-static sports (IIIA–C) with annual follow-up	
	Very high risk: only low–moderate-dynamic, low-static sports (IA, IB), with 6-monthly follow-up	With any associated clinical condition: dependent on type and severity of condition
	With any associated clinical condition: no competitive sports	

Table 34.3 Summary of selected differences between ESC and AHA/ACC recommendations for competitive athletes with aortopathies, coronary artery disease or hypertension

The ESC guidelines recommend that athletes with the full Marfan syndrome phenotype, as well as those with an incomplete phenotype but positive family history, should refrain from all competitive sports. Athletes with an incomplete phenotype, no FBN1 mutation and no family history are allowed to continue sports participation with annual follow-up. The same recommendations apply to athletes with a family history of Marfan syndrome, negative phenotype and no FBN1 mutation [1].

The AHA/ACC recommendations are based largely on aortic root dimensions, with 6–12 monthly monitoring by echocardiography or magnetic resonance angiography. Athletes with Marfan syndrome who do not have ≥1 of aortic root dilatation (transverse dimension ≥40 mm in men or ≥36 mm in women, or >2 standard deviations from the mean for body surface area in children); moderate-to-severe mitral regurgitation; LV systolic dysfunction (EF < 40%); or a family history of aortic dissection at an aortic diameter <50 mm are restricted to low–moderate-static, low-dynamic (class IA, IIA) competitive sports [22]. Otherwise, athletes with Marfan syndrome at higher risk of aortic dissection are restricted from competitive sports. The authors concur with the AHA/ACC recommendations.

Ischaemic Heart Disease

Ischaemic heart disease (IHD) is the leading cause of SCD in athletes over the age of 35 years. Younger athletes with familial hypercholesterolaemia are prone to premature atherosclerosis and are at risk of SCD

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and acute coronary syndromes. These events may occur in asymptomatic individuals without any prior symptoms of ischaemia.

The ESC guidelines recommend assessing all athletes who have a definite diagnosis of IHD (which includes angina, acute coronary syndromes and previous revascularisation) for the probability of future cardiac events. The assessment includes exercise-induced ischaemia, symptoms of complex ventricular arrhythmias significant coronary stenosis (70% of major coronary artery or >50% of left main stem) and reduced LV EF (<50%). Any of these signs indicates a higher probability of future cardiac events and results in disqualification from all competitive sports. In their absence, the probability of cardiac events is deemed low, and low–moderate-dynamic, low-static (class IA, IB) sports are permitted, with annual follow-up [1].

The AHA/ACC guidelines recommend a similar assessment of risk for future cardiac events. Athletes deemed at higher risk (exercise-induced ischaemia, impaired LV function or haemodynamically significant coronary artery stenosis) are restricted from competitive sports participation, though once stabilised may return to sports with low-dynamic and low-to-moderate static demands, classes IA and IIA. Athletes with clinically manifest IHD who are asymptomatic and athletes with clinically concealed IHD can participate in all competitive sports provided they have no inducible ischaemia or electrical instability and a resting LV EF >50%. Aggressive risk factor modification with high-intensity statin therapy is also advocated to reduce the chance of plaque disruption [23].

Anomalous Coronary Artery Origins

Anomalous coronary artery origins deserve special mention as an important cause of SCD in athletes in a large US registry [4]. The AHA/ACC guidelines recommend exclusion from all competitive sports with the possible exception of class IA sports. In order to be eligible to compete, an athlete with this condition must undergo surgical correction, with re-evaluation for ischaemia, arrhythmia and dysfunction on maximal exercise testing after 3 months. Any athletes presenting with a myocardial infarction (MI) should be risk-assessed in the same way as someone with IHD [24].

Hypertension

Hypertension is the most common CV condition observed in competitive athletes [25]. It is defined as a systolic BP ≥140 mmHg and/or diastolic BP ≥90 mmHg. Although hypertension may be associated with an increased level of risk for complex ventricular arrhythmias and sudden death, it has not directly been implicated as a cause of SCD in young competitive athletes. Athletes with hypertension should be treated according to the general guidelines for the management of hypertension. However, in endurance athletes, diuretics and beta blockers may impair exercise performance or cause electrolyte and fluid disturbances, and they may be prohibited by certain athletic training bodies. Therefore, calcium channel blockers and blockers of the renin–angiotensin system are the medications of choice for hypertensive endurance athletes [1].

The ESC guidelines recommend that athletes are risk-stratified according to the presence of risk factors, 10-year CV risk scores, target organ damage and/or associated clinical conditions. Athletes with well-controlled, mild (grade 1) hypertension, with no risk factors, a 10-year CV risk of <20% and no end organ damage, can participate in all competitive sports, with annual follow-up. Athletes with well-controlled mild-to-moderate hypertension (grade 1–2) with one or two risk factors but no end organ damage should not participate in high-dynamic, high-static (class IIIC) sports and be followed up yearly. Athletes with well-controlled mild-to-moderate (grade 1–2) hypertension with three or more risk factors, diabetes or target-organ damage should be excluded from all high-static (class IIIA–IIIC) sports, with annual follow-up. The same recommendation applies to athletes with well-controlled severe (grade 3) hypertension and no other risk factors. Athletes with treated severe (grade 3) hypertension and any risk factors, including those with target organ damage, should be restricted to low–moderate-dynamic, low-static (class IA, IB) sports, with 6-monthly follow-up. Any athlete with well-controlled hypertension who has an associated clinical condition of cerebrovascular disease, IHD, peripheral vascular disease (PVD), established nephropathy or retinopathy should be restricted from all competitive sports [1].

The AHA/ACC guidelines are simpler: athletes with mild (stage 1) hypertension without target organ damage or concomitant heart disease are eligible for all sports. Athletes with moderate (stage 2) hypertension

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Condition	ESC	AHA/ACC
Mitral stenosis (MS)	Mild MS, sinus rhythm: all competitive sports, excluding high-dynamic, high-static (IIIC)	Mild MS, sinus rhythm, normal pulmonary artery pressure: all competitive sports
	Mild MS and atrial fibrillation: non-contact, low–moderate-dynamic, low–moderate-static (IA, IB, IIA, IIB)	Severe MS or elevated pulmonary artery pressures: low-intensity sports (IA)
	Moderate and severe MS: non-contact, low-dynamic, low-static (IA)	Anticoagulation for atrial fibrillation: no contact sports
Mitral regurgitation (MR)	Mild-to-moderate MR, sinus rhythm, normal LV size/function and normal exercise testing: all sports	Mild-to-moderate MR, sinus rhythm, normal LV size/function and normal pulmonary artery pressures: all competitive sports
	On anticoagulation for atrial fibrillation: no contact sports	Mild-to-moderate MR and only mild LV dilatation, compatible with athletic training (EDV < 60 mm): all competitive sports
	Mild-to-moderate MR, mild LV dilatation (ESV < 55 ml.m^{-2}): Low–moderate-dynamic, low–moderate-static sports (IA, IB, IIA, IIB)	
	Severe MR or mild-to-moderate MR with marked LV dilatation (ESV > 55 ml.m^{-2}) or LV dysfunction (EF < 50%): no competitive sports	Severe MR, sinus rhythm and only mild LV dilatation: low-intensity and some moderate-intensity sports (IA, IIA, IB)
		Severe MR, definite LV enlargement (EDV ≥ 60 ml.m^{-2}), pulmonary hypertension or any degree of LV systolic dysfunction: low-intensity sports (IA)
		On anticoagulation for atrial fibrillation: no contact sports
Aortic stenosis (AS)	Asymptomatic mild AS, normal LV size/function, no significant arrhythmia: low–moderate-dynamic, low–moderate-static sports (IA, IB, IIA, IIB)	Mild AS: all competitive sports if normal maximal exercise response, with annual re-evaluation
		Moderate AS and satisfactory exercise tolerance test: low–moderate-static, low–moderate dynamic sports (IA, IIA, IB)
	Moderate AS or frequent/complex arrhythmias: low-dynamic, low-static sports (IA)	Asymptomatic severe AS: low-intensity competitive sports (IA)
	Severe AS or moderate AS with symptoms or LV dysfunction: no competitive sports	Symptomatic severe AS: no competitive sports
Aortic regurgitation (AR)	Mild-to-moderate AR, normal LV size/function, normal exercise testing and no arrhythmia: all competitive sports, with annual follow-up	Mild-to-moderate AR
		+ only mild LV enlargement and satisfactory exercise test: all competitive sports
	Mild-to-moderate AR with progressive LV dilatation: low-dynamic and low-static sports only (IA)	+ moderate LV enlargement (ESV < 50 mm in men and <40 mm in women) and satisfactory exercise test: all competitive sports
	Severe AR, or mild-to-moderate AR with dilatation of the ascending aorta or ventricular arrhythmias: no competitive sports	Severe AR, moderate enlargement and EF >50%: all competitive sports if satisfactory exercise test
		Symptomatic severe AR, EF <50% and more than moderate LV enlargement: no competitive sports
Prosthetic heart valves	Valve function and LV function normal: low–moderate-dynamic, low–moderate-static sports (IA, IB, IIA, IIB)	Valve function and LV function normal: low–moderate-dynamic, low–moderate-static sports (IA, IB, IC, IIA)
	Anticoagulation required: no contact sports	Anticoagulation required: sports with low likelihood of bodily contact

Table 34.4 Summary of selected differences between ESC and AHA/ACC recommendations for competitive athletes with valvular heart disease (VHD)
ESV, end systolic volume; EDV, end diastolic volume.

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without evidence of target organ damage should be restricted from high-static (class IIIA–IIIC) sports, until hypertension is controlled. Eligibility for competitive sport participation in athletes with coexistent CVD is usually based on the type and severity of the associated condition [25].

Valvular Heart Disease

Valvular heart disease (VHD) encompasses any stenotic or regurgitant lesion of any or multiple cardiac valves, as well as patients with prosthetic heart valves. In general, valvular regurgitation is better tolerated than valvular stenosis and exercise recommendations for these individuals are more liberal. The general recommendations are summarised in Table 34.4, and a detailed account can be found in Chapter 23.

Congenital Heart Disease

Congenital heart disease (CHD) encompasses a broad range of different cardiac abnormalities, varying from simple to highly complex lesions. The available literature regarding exercise and sports participation with CHD is sparse, and some conditions are not compatible with the haemodynamic changes required in exercise due to the morphological severity/complexity or the tendency to compromising arrhythmias. However, the haemodynamic balance in patients with CHD varies considerably, even amongst patients with the same lesions. This makes it impossible to state recommendations that are valid in all cases, and highlights the importance of cardiologists with expertise in CHD tailoring recommendation to the individual patient. As a general principle, only those patients with CHD who are likely to deteriorate as a consequence of regular physical exercise should be restricted from sports participation [1].

Otherwise, summarised simply, the ESC guidelines allow sports competition for athletes with atrial septal defects, small ventricular septal defects, AV septal defects with competent valvular function, anomalous pulmonary venous connection and patent ductus arteriosus, provided that they are asymptomatic and have normal ventricular function, normal pulmonary artery pressures, a normal BP response to exercise and no significant arrhythmias [1]. More complex lesions are covered in greater detail in both the ESC and AHA/ACC guidelines [1, 24]; this falls beyond the scope of this chapter.

This chapter comprehensively summarises the current guidelines pertaining to CV conditions associated with SCD in athletes or which one might commonly expect to face in an athletic population. In general, it is prudent to advise any individual with a CV condition to avoid sudden explosive exertions, such as sprinting. Graded increases in workload are better tolerated by the heart. They should also avoid exercising in extreme adverse environmental conditions (hot, humid or very cold), as this may precipitate cardiac events in predisposed individuals. Finally, it may be considered safe and conservative practice to advise athletes with certain CV conditions to maintain their heart rate at ≤80% of maximum or at the level for the anaerobic threshold, if this is known.

In summary, whilst it is appreciated that exercise can trigger significant cardiac events in individuals with underlying CV conditions, it is difficult to predict the risk of a future cardiac event and what impact regular physical activity and competitive sports participation might have on this risk. Current guidelines are predominantly based on the interpretation of circumstantial evidence by a consensus group and are conservative in nature in an attempt to encompass all preventable deaths. Nevertheless, some athletes may choose not to follow the recommendations of their physicians and although they place themselves at perceived risk, they should be followed up and included in registries to add to the currently limited evidence base.

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