Inherited cardiomyopathies and sports participation

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Abstract
Competitive sports activity is associated with an increased risk of sudden cardiovascular death (SCD) in adolescents and young adults with inherited cardiomyopathies. Many young subjects aspire to continue competitive sport after a diagnosis of cardiomyopathy and the clinician is frequently confronted with the problem of eligibility and the request of designing specific exercise programs. Since inherited cardiomyopathies are the leading cause of sudden cardiovascular death during sports performance, a conservative approach implying disqualification of affected athletes from most competitive athletic disciplines is recommended by all the available international guidelines. On the other hand, we know that the health benefits of practicing recreational sports activity can overcome the potential arrhythmic risk in these patients, provided that the type and level of exercise are tailored on the basis of the specific risk profile of the underlying cardiomyopathy. This article will review the available evidence on the sports-related risk of sudden cardiac death and the recommendations regarding eligibility of individuals affected by inherited cardiomyopathies for sports activities.

Keywords
Athletes · Cardiomyopathy · Exercise prescription · Sports cardiology · Sudden cardiac death

Introduction
Competitive sports activity is associated with an increased risk of sudden cardiovascular death (SCD) in adolescents and young adults with clinically silent cardiovascular disorders [2]. Inherited cardiomyopathies are recognised leading causes of sports-related cardiac arrest in young competitive athletes, with hypertrophic cardiomyopathy (HCM) accounting for one-third of fatal cases in the USA and arrhythmogenic right ventricular cardiomyopathy for approximately one-fourth in Italy [3–5]. Preparticipation evaluation may allow the identification of asymptomatic athletes who have potentially lethal cardiomyopathies and protect them from the risk of SCD through restriction from competitive sports (Fig. 1; [6, 7]). However, many affected athletes aspire to continue practicing leisure-time sports activities or at least maintain a physically active lifestyle despite their non-eligibility to engage in competitive sports.

This article will review the available data on the sports-related risk of SCD and the recommendations regarding the eligibility to engage in sports activity, either competitive or recreational, of individuals affected by inherited cardiomyopathies. We will focus on major inherited cardiomyopathies which have been implicated as leading causes of SCD during sport activity and/or require a specific clinical work-up for differential diagnosis with physiologic remodelling of the athlete’s heart. We refer for sports participation with inherited channelopathies to Panhuyzen et al. in this same issue [8].

Hypertrophic cardiomyopathy
Hypertrophic cardiomyopathy, the most common inherited heart muscle disease, is caused by mutations of genes encoding for sarcomeric proteins. The phenotype of HCM is characterised by left ventricular (LV) hypertrophy, either symmetric or asymmetric, which occurs in the absence of abnormal loading conditions. The diagnosis traditionally relies on the demonstration by echocardiography of otherwise unexplained diffuse or segmental LV wall and/or septal thickening ≥15 mm; a lower cut-off value (≥13 mm) is used in the context of family members of affected individuals, especially if they are mutation carriers [9, 10]. Cardiac magnetic resonance imaging may provide additional diagnostic and prognostic information given its ability to better detect segmental LV hypertrophy in some regions, such as...
Inherited cardiomyopathies are recognized leading causes of sports-related cardiac arrest in young competitive athletes and for this reason both European and American recommendations for sports eligibility agree that athletes with cardiomyopathy should be restricted from competitive sports activity.

Carriers of desmosomal gene mutation with no signs of arrhythmogenic right ventricular cardiomyopathy should also be disqualified from competitive sports activity because intense physical exercise promotes the development and progression of the disease phenotype.

There is growing evidence that patients with an implantable cardioverter defibrillator may practice sports safely. However, it must be emphasized that participation in competitive sports at high cardiovascular demands may promote disease progression.

 Patients with cardiomyopathy and mild phenotypic expression at low risk of sudden death may practice moderate-intensity recreational exercise. Patients should be educated to avoid peak exertion and recognize warning symptoms.

**KEY MESSAGE**

1. Inherited cardiomyopathies are recognized leading causes of sports-related cardiac arrest in young competitive athletes and for this reason both European and American recommendations for sports eligibility agree that athletes with cardiomyopathy should be restricted from competitive sports activity.

2. Carriers of desmosomal gene mutation with no signs of arrhythmogenic right ventricular cardiomyopathy should also be disqualified from competitive sports activity because intense physical exercise promotes the development and progression of the disease phenotype.

3. There is growing evidence that patients with an implantable cardioverter defibrillator may practice sports safely. However, it must be emphasized that participation in competitive sports at high cardiovascular demands may promote disease progression.

4. Patients with cardiomyopathy and mild phenotypic expression at low risk of sudden death may practice moderate-intensity recreational exercise. Patients should be educated to avoid peak exertion and recognize warning symptoms.

Left ventricular hypertrophy may develop as a consequence of heart adaption to physical exercise (athlete’s heart) and is particularly prominent in athletes involved in endurance sports activities, such as rowing and cycling [12], but maximal wall thickness rarely exceeds 13 mm [13–15]. A minority of highly trained athletes, particularly males of African/Afro-Caribbean descent, may exhibit more pronounced LV hypertrophy (13–16 mm), which requires an accurate clinical work-up for differential diagnosis with HCM [13–15]. In addition, in African/Afro-Caribbean athletes the electrocardiogram (ECG) often demonstrates a variant of the anterior early repolarisation pattern characterised by J-point/ST-segment elevation and T-wave inversion in the anterior precordial leads (V1 to V4) which may raise the suspicion of an underlying cardiomyopathy (Fig. 2; [16, 17]). A LV wall thickness >16 mm should be considered diagnostic of HCM even in highly trained athletes, irrespective of ethnicity [18, 19]. In athletes with LV hypertrophy falling into the ‘grey zone’ (13–16 mm), differential diagnosis between HCM and athlete’s heart is based on several parameters including imaging features (cavity size, distribution of hypertrophy, outflow tract obstruction, mitral valve abnormalities, diastolic function, and late enhancement at cardiac magnetic resonance imaging with a characteristic pattern of patchy involvement [20]) and ECG findings (ST-segment depression, T-wave inversion in inferolateral leads, pathological Q waves and conduction disturbances) (Fig. 3; [18, 19]).

Hypertrophic cardiomyopathy is one of the leading causes of SCD among athletes in the USA where it has been reported to account for more than one third of fatal cases [3, 4]. By contrast, in European studies HCM is reported to cause less than 10% of SCDs ([5, 21]; Table 1). Besides ethnic and genetic differences between the populations, this discrepancy may be explained by the preventive identification of HCM through systematic preparticipation screening including ECG, which is abnormal in up to 90% of athletes with HCM (Fig. 4; [1, 22–24]). ECG screening is common practice in most European countries and is compulsory in Italy, but it is not routinely performed in the USA.

High intensity competitive sports may promote life-threatening ventricular arrhythmias even in the absence of traditional risk factors and there is no evidence that algorithms developed to predict the risk of SCD in the general population of patients with HCM can be extrapolated to athletes. Accordingly, both European and American recommendations for sports eligibility agree that athletes with HCM should be restricted from competitive sports activity.

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**Fig. 1** Trends of sudden death for cardiomyopathies among athletes in the Veneto region of Italy after implementation of a national preparticipation screening programme. Adapted with permission from Corrado et al. [1]
Instead, participation in disciplines at low cardiovascular demand (such as bowling, golf and brisk walking) may be allowed to asymptomatic patients with mild LV hypertrophy (wall thickness <20 mm), no ventricular arrhythmias at exercise testing or ambulatory ECG monitoring and negative family history for SCD ([29, 30]; Table 2).

**Arrhythmogenic cardiomyopathy**

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inherited cardiomyopathy characterised by progressive loss of myocytes and replacement by fibro-fatty tissue, which creates the substrate for life-threatening ventricular arrhythmias. In its classical form the phenotypic manifestations predominantly involve the right ventricle, but biventricular and left-dominant variants exist. Molecular genetic studies showed that ARVC is a genetic disorder resulting from defective desmosomal proteins, most often with an autosomal dominant pattern of inheritance [31, 32].

The clinical manifestations of the disease usually occur between the second and forth decade of life and consist of ECG depolarisation and repolarisation changes, typically localised in the right precordial leads (V1–V3/V4), structural abnormalities, such as global or regional dilatation/dysfunction of the right ventricle and, most importantly, ventricular arrhythmias with a left bundle branch block morphology, ranging from isolated premature ventricular beats to sustained ventricular tachycardia, which can degenerate into ventricular fibrillation [33, 34].

The diagnosis of ARVC requires a combination of different criteria from various categories such as 1) histopathological abnormalities at endomyocardial biopsy; 2) morphofunctional abnormalities consisting of regional right ventricular wall motion abnormalities (regional akinesia, dyskinesia or bulging) plus right ventricular dilatation or global...
Fig. 3 Differential diagnosis between athlete’s heart and hypertrophic cardiomyopathy. A subset of athletes exhibits an increase in left ventricular wall thickness that falls in the ‘grey zone’ of overlap between physiological adaptation to exercise and pathological hypertrophy. In these cases, the presence of positive family history of sudden cardiac death, cardiac arrest or inherited cardiac disease, electrocardiographic changes, echocardiographic abnormalities or late enhancement at cardiac magnetic resonance imaging suggests an underlying hypertrophic cardiomyopathy. On the other hand, symmetrical left ventricular hypertrophy with concomitant left ventricular dilatation compatible with gender (male > female), ethnicity (African/Afro-Caribbean > Caucasian) and intensity of training with no other abnormal features is suggestive of athlete’s heart.

Arrhythmogenic cardiomyopathy has been demonstrated to be the leading cause of SCD in athletes of the Veneto region of Italy (14% of cases), whereas in the USA it accounts for only 6% of cases ([3–5]; Table 1). This finding may depend on the experience of the pathologist or coroner who performs post-mortem investigation as ARVC is rarely associated with cardiomegaly and may spare the left ventricle, so that the affected heart may be erroneously diagnosed as normal. Of note, in a British study on SCD in athletes, whose hearts were referred for post-mortem to a tertiary centre, the prevalence of ARVC was similar (13%) to that found in the Italian series [21].

The ‘left dominant’ variant of ARVC is characterised by an early and predominant LV involvement, as a result of a specific genetic background [36]. At variance with the classic ‘right-dominant’ variant (Fig. 5), the power of traditional investigations including routine ECG and standard echocardiography for the diagnosis of ‘left-dominant’ ARVC is limited because repolarisation abnormalities and left ventricular systolic dysfunction, either regional or global, are observed in a minority of affected patients. The reason is that the fibro-fatty scarring process predominantly involves the sub-epicardial and mid-mural myocardial layers, which contribute less to ventricular contraction, and the disease lesion can be only identified in the form of late gadolinium enhancement using contrast-enhanced cardiac magnetic resonance imaging (Fig. 6; [37]). Not surprisingly, the incidence of SCD in patients with the classic ARVC variant has markedly decreased since the introduction of ECG preparticipation screening, while the difficult-to-diagnose ‘left dominant’ variant is now an increasingly emergent substrate at post-mortem [38, 39].

Competitive sports activity increases the risk of SCD by 5-fold in adolescent and young adults with ARVC [5]. Sports has also been implicated as the most important environmental factor promoting ARVC progression and worsening of the disease arrhythmic substrate (Fig. 7). Actually, genetically determined impairment of cell-to-cell adhesion may predispose to myocyte detachment and death, mostly under condition of increased mechanical wall stress such as that occurring during competitive sports activity [41]. Kirchhof et al. [42] demonstrated that in heterozygous plakoglobin-deficient mice, endurance training accelerated the development of right ventricular abnormalities and ventricular arrhythmias. James et al. [43] and Saberniak et al. [44] confirmed in clinical studies that endurance sports and intense physical exercise increase age-related penetrance, risk of ventricular tachyarrhythmias and occurrence of heart failure in ARVC desmosomal gene carriers. Ruwald et al. [45] reported that the absolute risk of malignant arrhythmic events in ARVC patients who participated in competitive sports reached 61% at 40 years of age. On the other hand, early identification of affected patients by preparticipation screening and disqualification from competitive sports activity offers the potential to prevent SCD [1]. Accordingly, both European and American recommendations for sports eligibility in patients with heart diseases and the International Task Force consensus document on ARVC treatment agree that restriction from competitive sports activity of patients with ARVC should be regarded as a therapeutic measure aimed to reduce the risk of SCD (Table 2; [29, 30, 46]).

Dilated cardiomyopathy and left ventricular noncompaction

Dilated cardiomyopathy (DCM) is characterised by dilatation and systolic dysfunction of the left ventricle which...
| Ref | Study period | Region | Population | Results (in %) |
|-----|--------------|--------|------------|---------------|
| Corrado, JACC 2003 [5] | 1979–1999 | Veneto, Italy | SD among athletes 12–35 years old | Cardiovascular SD: 86 |
| | 1979–1999 | Veneto, Italy | Cardiovascular SD: 86 |
| | 1979–1999 | Veneto, Italy | CAD: 22 |
| | 1979–1999 | Veneto, Italy | ARVC: 14 |
| | 1979–1999 | Veneto, Italy | Myocarditis: 12 |
| | 1979–1999 | Veneto, Italy | Mitral valve prolapse: 10 |
| | 1979–1999 | Veneto, Italy | Conduction system disease: 10 |
| | 1979–1999 | Veneto, Italy | HCM: 9 |
| | 1979–1999 | Veneto, Italy | Aortic rupture: 5 |
| | 1979–1999 | Veneto, Italy | Non cardiovascular SD: 14 |
| | 1979–1999 | Veneto, Italy | Unexplained SD: 8 |
| | 1979–2004 | Veneto, Italia | SD for structural heart disease among athletes: |
| | 1979–2004 | Veneto, Italia | Cardiomyopathy: 25 |
| | 1979–2004 | Veneto, Italia | CAD: 20 |
| | 1979–2004 | Veneto, Italia | Coronary anomalies: 13 |
| | 1979–2004 | Veneto, Italia | Myocarditis: 13 |
| | 1979–2004 | Veneto, Italia | Mitral valve prolapse: 11 |
| | 1979–2004 | Veneto, Italia | Conduction system disease: 7 |
| | 1979–2004 | Veneto, Italia | SD for structural heart disease among non-athletes: |
| | 1979–2004 | Veneto, Italia | Cardiomyopathy: 31 |
| | 1979–2004 | Veneto, Italia | CAD: 20 |
| | 1979–2004 | Veneto, Italia | Myocarditis: 15 |
| | 1979–2004 | Veneto, Italia | Conduction system disease: 9 |
| | 1979–2004 | Veneto, Italia | Mitral valve prolapse: 7 |
| | 1979–2004 | Veneto, Italia | Coronary anomalies: 5 |
| Maron, Circulation, 2009 [3] | 2001–2006 | USA | SD among 10,700,000 athletes 13–25 years old | Unexplained: 34 |
| | 2001–2006 | USA | CAD: 20 |
| | 2001–2006 | USA | Myocarditis: 11 |
| | 2001–2006 | USA | Coronary anomalies: 5 |
| De Noronha, Heart, 2009 [25] | 1996–2008 | United Kingdom | Athletes with SD referred to the National Heart and Lung Institute and Royal Brompton Hospital pathology dpt. | Structurally normal heart: 23 |
| | 1996–2008 | United Kingdom | Idiopathic LV hypertrophy: 31 |
| | 1996–2008 | United Kingdom | ARVC: 14 |
| | 1996–2008 | United Kingdom | HCM: 11 |
| | 1996–2008 | United Kingdom | Idiopathic LV fibrosis: 6 |
| | 1996–2008 | United Kingdom | Coronary anomalies: 5 |
| Harmon, Circulation Arrhythm Electrophysiol, 2014 [26] | 2004–2008 | USA | SD among National Collegiate Athletic Association athletes 17–24 years old | Unexplained: 31 |
| | 2004–2008 | USA | Coronary anomalies: 14 |
| | 2004–2008 | USA | Idiopathic LV hypertrophy: 8 |
| | 2004–2008 | USA | Aortic dissection: 8 |
| | 2004–2008 | USA | Myocarditis: 8 |
| | 2004–2008 | USA | Dilated cardiomyopathy: 8 |
| | 2004–2008 | USA | CAD: 5 |
| | 2004–2008 | USA | Unexplained: 42 |
| | 2004–2008 | USA | Idiopathic LV hypertrophy/fibrosis: 16 |
| | 2004–2008 | USA | ARVC: 13 |
| Finocchiaro, J Am Coll Cardiol, 2016 [21] | 1994–2014 | United Kingdom | Athletes with SD referred to St. George’s University of London Cardiac Pathology dpt | Unexplained: 42 |
| | 1994–2014 | United Kingdom | CAD: 5 |
| | 1994–2014 | United Kingdom | Myocarditis: 5 |
Table 1  Main studies reporting the causes of cardiac arrest/sudden death in the athletes (Continued)

| Ref | Study period | Region        | Population                  | Results (in %) |
|-----|--------------|---------------|----------------------------|----------------|
| Grani, Eur J Prev Cardiol, 2016 [27] | 1999–2010     | Switzerland    | Exercise-related SD         | CAD: 28        |
|     |              |               | 10–39 years old            | HCM: 14        |
|     |              |               |                             | Unexplained: 13|
|     |              |               |                             | Valvular: 8    |
|     |              |               |                             | Coronary anomalies: 7 |
|     |              |               |                             | ARVC: 7        |
|     |              |               |                             | Idiopathic myocardial fibrosis: 7 |
|     |              |               |                             | Aortic dissection: 7 |
|     |              |               |                             | Dilated cardiomyopathy: 6 |

ARVC arrhythmogenic right ventricular cardiomyopathy, CAD coronary artery disease, HCM hypertrophic cardiomyopathy, LV left ventricular, SD sudden death, WPW Wolff-Parkinson-White syndrome

Fig. 4 Electrocardiographic and echocardiographic findings in a 15-year-old male soccer player with hypertrophic cardiomyopathy. The electrocardiogram shows T-wave inversion in lateral leads (I and aVL) and pathological Q-wave (duration >25% of the height of the ensuing R-wave) in inferior leads (III and aVF) (a). The echocardiogram shows an asymmetric left ventricular hypertrophy with maximal septal thickness of 31 mm (b). (VS = ventricular septal, LV = left ventricle, LA = left atrium, AO = aorta). Reproduced with permission from Migliore et al. [28]

is genetically determined in approximately one third of cases. Up to 40 genes have been identified, which affect proteins of a wide variety of cellular structures such as the sarcomere, the nuclear envelope, the cytoskeleton, the sarcolemma and the intercellular junction [48]. Differential diagnosis between athlete’s heart and DCM may be challenging as a sizeable proportion of athletes, mostly those engaged in endurance sports activities, exhibit an enlarged left ventricle and a subset of them show a mild reduction in ejection fraction, i.e. between 45 and 55% [49, 50]. The diagnosis of DCM is suggested by the presence of the following features: depressed systolic function with ejection fraction below 45%, associated regional wall motion abnormalities, concomitant right ventricular dysfunction, positive family history of SCD, cardiac arrest or inherited cardiac disease, and evidence of ECG abnormalities, such as T-wave inversion or intraventricular conduction defects, complex ventricular arrhythmias at 24-hour Holter monitoring or stress testing and late gadolinium enhancement at contrast-enhanced cardiac magnetic resonance. In borderline cases, demonstration of a significant (>10%) increase of the ejection fraction during exercise echocardiography indicates a preserved contractile reserve and may support the diagnosis of athlete’s heart versus DCM [51].

The American Heart Association consensus statement on definition and classification of cardiomyopathies considers
Table 2  Eligibility to competitive sports participation in athletes with cardiomyopathies based on the 2005 European Society of Cardiology and the 2017 American Heart Association/American College of Cardiology recommendations

| Cardiomyopathy | Phenotype | Characteristics | Recommendations |
|----------------|-----------|-----------------|----------------|
| HCM            | High risk | Definitive diagnosis of HCM with symptoms, moderate to severe hypertrophy, ventricular arrhythmias or family history of sudden death | No competitive sports |
|                | Low risk  | Asymptomatic athletes with mild left ventricular hypertrophy, no ventricular arrhythmias and no sudden death in 1st degree relatives | Only sports at low cardiovascular demands |
|                | Healthy gene carrier | Carrier of pathogenetic mutation with no signs of disease | All competitive sports |
| ARVC           | All phenotypes | Definitive diagnosis of ARVC | No competitive sports |
|                | Healthy gene carrier | Carrier of pathogenetic mutation with no signs of disease | Only sports at low cardiovascular demand |
| DCM            | High risk | Definitive diagnosis of DCM with symptoms, moderate to severe left ventricular dysfunction, ventricular arrhythmias or family history of sudden death | No competitive sports |
|                | Low risk  | Asymptomatic athletes with mild left ventricular dysfunction, no ventricular arrhythmias and no sudden death in 1st degree relatives | Only sports at low cardiovascular demand |
|                | Healthy gene carrier | Carrier of pathogenetic mutation with no signs of disease | All competitive sports |
| LVNC           | High risk | Athletes with LVNC and symptoms, systolic dysfunction or ventricular arrhythmias | No competitive sports |
|                | Low risk  | Asymptomatic athletes with LVNC, normal systolic function and no ventricular arrhythmias | Competitive sport may be considereda |
|                | Healthy gene carrier | Carrier of pathogenetic mutation with no signs of disease | All competitive sports |

aAccording to the 2017 American Heart Association/American College of Cardiology guidelines

the left ventricular noncompaction (LVNC) as a genetic heart muscle disorder [52], while a European Society of Cardiology consensus document reports it as ‘unclassified cardiomyopathy’ [53]. This inconsistency in the classification of LVNC can be explained by the uncertain nature of the disease, either a separate condition or a phenotypic variant of other cardiomyopathies. Pathogenic mutations have been identified in up to 41% of the affected subjects, in a significant proportion of cases (29%) as sarcomere gene mutations [54, 55]. Although current imaging criteria for the diagnosis of LVNC are based on the ratio between the compact and non-compact LV layers, the thickness of the non-compacted myocardium or the number of trabeculations, these morphologic features are non-specific and can be observed in up to 8% of highly trained athletes (particularly of African/Afro-Caribbean ethnicity). These morphologic changes have been interpreted as a result of the chronic increase in the LV preload occurring during intense training and exercise. In this regard, hypertrabeculation has also been observed in healthy pregnant women (with complete resolution or marked reduction of trabeculation after delivery) or in patients with chronic anaemia [56–58]. Hypertrabeculation is rarely associated with LV systolic dysfunction, ECG repolarisation abnormalities, or late gadolinium enhancement, suggesting a ‘LVNC cardiomyopathy’ [58]. Hence, hypertrabeculation in isolation can be part of the adaptive heart changes to exercise and should not be considered a cardiac disease [59].

Dilated cardiomyopathy is an uncommon cause of SCD in young competitive athletes. In the Veneto region of Italy, it accounted for 4% of fatalities with cardiovascular origin [5], while in the USA, Maron et al. found it in 2.5% [3] and Harmon et al. in 8% [26] of SCD victims. Finocchiaro et al. reported a 1% prevalence of fatal DCM in British athletes [21]. It is noteworthy that none of the above-mentioned studies reported isolated LVNC as a cause of SCD in athletes (Table 1). Athletes affected by either DCM or LVNC are considered at significant risk of SCD during exercise, and both European and American recommendations agree that affected athletes are not eligible to engage in competitive sports activity. Conversely, asymptomatic athletes with isolated LVNC, i.e. with a normal LV systolic function, no ECG abnormalities and/or ventricular arrhythmias, and no evidence of late enhancement at cardiac magnetic resonance imaging can participate in competitive sports ([29, 30]; Table 2).
at 10 years from ICD implant. The authors concluded that
lead malfunction was 97% at 5 years and 90%
or arrhythmia related injuries during sports activity. Free-
ertion). There were no deaths, resuscitated cardiac arrests
in either competitive (N = 44) or dangerous (N = 44)
sports. During a median follow-up of 31 months, 46 ath-
letic individuals. However, ARVC associated with desmo-
some gene mutation constitutes an important exception: in
this condition, physical exercise and sports activity have
been implicated as a key environmental factor for promot-
ing the development and progression of the disease pheno-
type, both in animal models [42] and in clinical studies on
gene mutation carriers [43–45].

Healthy gene carriers

At present, there is no scientific evidence demonstrating
that competitive sports activity increases the risk of disease
development or sudden death in athletes who carry a patho-
genetic mutation but who do not show any sign of pheno-
typic manifestation (i.e., genotype-positive/phenotype-neg-
ative individuals). However, ARVC associated with desmo-
some gene mutation constitutes an important exception: in
this condition, physical exercise and sports activity have
been implicated as a key environmental factor for promot-
ing the development and progression of the disease pheno-
type, both in animal models [42] and in clinical studies on
gene mutation carriers [43–45].

Implantable cardioverter defibrillator (ICD)
and sports activity

Patients with an ICD are traditionally considered not eligi-
ble to engage in competitive sports activity, except for disci-
plines characterised by a low cardiovascular demand which
do not expose to the risk of traumatic damage of the device
[60]. However, there is growing evidence that ICD patients
may practice sports safely. Lampert et al. [61] reported on
372 US athletes with ICDs (age 10–60 years) participating
in either competitive (N = 328) or dangerous (N = 44)
sports. During a median follow-up of 31 months, 46 ath-
letes had appropriate therapies (25 during physical exertion)
and 29 inappropriate interventions (25 during physical ex-
ertion). There were no deaths, resuscitated cardiac arrests
or arrhythmia related injuries during sports activity. Free-
dom from lead malfunction was 97% at 5 years and 90%
at 10 years from ICD implant. The authors concluded that
their results on efficacy and safety of ICD therapy in the
athletic population did not differ significantly from avail-
able data in non-athletic ICD patients and, thus, do not
support restriction of sports activity for ICD patients. In
line with these new data, the most recent recommendations
of the American Heart Association regarding the eligibility
disqualification of athletes with cardiovascular abnor-
malities to engage in competitive sports stated that compet-
itive sports may be allowed in selected athletes with an ICD
[62]. The new subcutaneous ICD system appears a valuable
therapeutic option for young patients with cardiomyopathy
to avoid the risk of transvenous lead damage. However, we
should emphasise that the reasons for competitive sports re-
striction in young patients with cardiomyopathy who have
received an ICD for the prevention of sudden cardiac death
be beyond the increased risk of unsuccessful shock, inap-
propriate interventions, injury to the patient, or damage of
the device. In fact, as discussed above for patients with
ARVC, sports participation may play a key role in the heart
muscle disease progression, worsening of the arrhythmic
substrate and adverse outcome.

Recreational sports activity

Recreational sports include a wide range of physical activi-
ties, from modest to vigorous in intensity, performed either
in a regular or inconsistent basis not requiring systematic
training or pursuit of excellence, nor involving the same
psychological pressure to surpass other participants, which
is characteristic of competitive sports.

Although there is a paucity of scientific data regarding
the risk of SCD in patients with cardiomyopathies engage-
in recreational sports activity, it is likely that a certain
Fig. 5  Electrocardiographic and echocardiographic findings in a 14-year-old male soccer player with right ventricular dominant (classic phenotype) arrhythmogenic cardiomyopathy. The electrocardiogram shows T-wave inversion in right precordial leads (V1–V2) (a). The echocardiogram reveals right ventricular dilatation (right ventricular outflow tract diameter of 39 mm on end-diastolic parasternal short-axis view) (b) and right ventricular dysfunction (akinesia of right ventricular outflow tract and posterobasal, subtricuspidal regions) (not shown). Reproduced with permission from Migliore et al. [28]

Fig. 6  Representative example of a left dominant arrhythmogenic cardiomyopathy. Post-contrast cardiac magnetic resonance findings of an asymptomatic 23-year-old female carrying a desmoplakin gene mutation. Four-chamber (a) and short-axis (b) views showing late gadolinium enhancement mostly involving the subepicardial layer of the posterolateral left ventricular wall at mid-basal level (white arrows), in the absence of other morpho-functional ventricular abnormalities. The electrocardiogram and echocardiogram of this patient were normal. Reproduced with permission from Zorzi et al. [40]
Fig. 7 Schematic representation of ARVC course from desmosomal gene mutation to phenotypic expression and cardiac arrest due to ventricular fibrillation. Sports activity may promote development of phenotypic expression, accelerate disease progression and trigger life-threatening ventricular arrhythmias. Reproduced with permission from Corrado et al. [47]. (ARVC arrhythmogenic right ventricular cardiomyopathy)

degree of risk will remain. However, it seems reasonable to state that this potential risk of exercise should not entirely deprive cardiomyopathy patients of the many benefits offered by regular exercise [63]. A recent study in a small cohort of HCM patients with mild phenotype suggested that moderate-intensity physical activity may provide benefits in terms of fitness with no significant arrhythmic risk [64]. Ruwald et al. [45] reported that the absolute risk of ventricular tachyarrhythmias/death at 40 years after birth was high (33%) in ARVC patients practicing recreational sports, but did not significantly differ from that of physically inactive ARVC patients (22%).

With regard to the prescription of recreational exercise programmes in patients with a diagnosis of cardiomyopathy, the following criteria which characterise a ‘low risk’ of SCD should be taken into account: 1) no symptoms; 2) negative family history for SCD; 3) ‘mild’ structural abnormalities; 4) normal or near-normal response to exercise (i.e. no ST-segment depression and normal increase of blood pressure during stress testing); 5) absence of clinically relevant arrhythmias at exercise testing or during 24-hour ambulatory ECG monitoring. When all these conditions are fulfilled, the physician can reassure the patient with respect to his ability and safety to sustain a regular exercise programme; if one or more risk criteria are present, only low intensity physical activities should be prescribed (Table 3).

Patients should be educated to start an exercise session with a warm-up period and specifically avoid peak exertion, characterised by high-intensity interval exercise. At the end of the session, an appropriate cool-down period is also recommended. The clinician should teach patients to take control over the level of exertion by assessing heart rate with commercially available devices. Exercise in extremely adverse environmental conditions, including very hot, humid, or extremely cold weather should be avoided. Patients should be carefully informed on the specific risk profile of their disease, the potentially dangerous sports activities, mostly in the presence of a history of syncope, and the warning symptoms that may occur in association with exercise. Finally, the availability of an automated external defibrillator in the athletic field should be considered as an additional ‘back-up’ measure to prevent SCD.

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