Sport bei ACM: Ja, Nein, Vielleicht?

DGK 89. Jahrestagung

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Disclosure Statement of Financial Interest

I, J. Scharhag, DO NOT have a financial interest/arrangement or affiliation with one or more organizations that could be perceived as a real or apparent conflict of interest in the context of the subject of this presentation.

Plötzlicher Herztod Italien

TABLE 2. CAUSES OF SUDDEN DEATH IN ATHLETES AND NONATHLETES 35 YEARS OF AGE OR LESS IN THE VENETO REGION OF ITALY, 1979 TO 1996.

CAUSE	ATHLETES (N=49)	NONATHLETES (N=220)	TOTAL (N=269)
	number (percent)		
Arrhythmogenic right ventricular cardiomyopathy	11 (22.4)	18 (8.2)*	29 (10.8)
Atherosclerotic coronary artery disease	9 (18.4)	36 (16.4)	45 (16.7)
Anomalous origin of coronary artery	6 (12.2)	1 (0.5)†	7 (2.6)
Disease of conduction system	4 (8.2)	20 (9.1)	24 (8.9)
Mitral-valve prolapse	5 (10.2)	21 (9.5)	26 (9.7)
Hypertrophic cardiomyopathy	1 (2.0)	16 (7.3)	17 (6.3)
Myocarditis	3 (6.1)	19 (8.6)	22 (8.2)
Myocardial bridge	2 (4.1)	5 (2.3)	7 (2.6)
Pulmonary thromboembolism	1 (2.0)	3 (1.4)	4 (1.5)
Dissecting aortic aneurysm	1 (2.0)	11 (5.0)	12 (4.5)
Dilated cardiomyopathy	1 (2.0)	9 (4.1)	10 (3.7)
Other	5 (10.2)	61 (27.7)	66 (24.5)

*P=0.008 for the comparison with the athletes.

†P<0.001 for the comparison with the athletes.

Corrado D et a. N Engl J Med 1998

Ursache Plötztlicher Herztod Spanien

TABLE 1. **Sudden death during athletic activities (1995-2001)**

	No. of cases	Age (years)	Sex	Pathology	Sport	
Total	61	11-65 (31.9±14.2)	59 M 2 W		Cycling (21), soccer (13), gymnastics (5), jogging (4)	paddle/fronton (4), basketball (2), other (12),
CAD	25 (40.9%)	28-65 (44.4±9.4)	25 M	Atheroma (88%) Scars (56%) AMI (8%) Thrombosis (28%)	Cycling (11), Soccer (4), Gymnastics (2), Fronton (2)	jogging (2) paddle, mountain climbing marching, other (4)
ACM	10 (16.3%)	13-39 (25.5±8.3)	10 M	Biventricular (4) RV (2) LV (2)	Cycling, Fronton, tennis, Gymnastics, marathon (5)	soccer (3) other sport (2)
HCM	4 (6.5%)	11 30 44 45	M M M W	Heart: 252 g (symmetrical) Heart: 405 g (asymmetrical) Heart: 478 g (asymmetrical) Heart: 401 g (symmetrical)	Gymnastics Cycling Cycling Jogging	
LVH	3 (4.9%)	28 20 18	M M M	Heart: 512 g (?) Heart: 528 g (↑34%) Heart: 459 g (↑Y20%)	Other sport Basketball Soccer	
Myocardial fibrosis	2 (3.2%)	20 17	M M	LV subepicardial fibrosis LV/RV fibrosis	Basketball Cycling	
DCM	1 (1.6%)	14	M	DCM (Heart: 550 g)	Soccer	
Coronary anomalies	2 (3.2%)	22 16	M M	LC in right sinus RC between aorta and pulmonary Posterior LV fibrosis	Cycling Soccer	
Aortic valve disease	2 (3.2%)	12 15	M M	Supravalvular AS Complicated bicuspid valve	Gymnastics Other sport	
ASD	1 (1.6%)	17	M	ASD 9 × 10 mm RV hypertrophy	Cycling	
Flecainide	1 (1.6%)	51	M	Heart: 464 g	Cycling	
Indeterminate	10 (16.3%)	15-29 (20.2±5.3)	9 M 1 W	No significant changes	Cycling (3), soccer (3), jogging (1)	badminton (1), other sport (2),

Suarez-Mier
Rev Esp Cardiol 2002

Ursachen Plötzlicher Herztod Spanien

ACM	10 (16.3%)	13-39 (25.5±8.3)	10 M	Biventricular (4) RV (2) LV (2)	Cycling, Fronton, tennis, Gymnastics, marathon (5)	soccer (3) other sport (2)

TABLE 2. **Sudden death during athletic activities, by age (1995-2001)**

	No. of cases	CAD	HCM	ACM	LVH	Fibrosis/DCM	Coronary anomalies	Aortic valve disease	Others	Indeterminate
≤ 30 years	32	2 (6.2%)	2 (6.2%)	7 (21.8%)	3 (9.3%)	3 (9.3%)	2 (6.2%)	2 (6.2%)	1 (3.1%)	10 (31.2%)
> 30 years	29	23 (79.3%)	2 (6.8%)	3 (10.3%)	–	–	–	–	1 (3.4%)	–
P		<.00001	NS	NS	NS	NS	NS	NS	NS	<.001

CAD indicates coronary atheromatous disease; LVH, idiopathic left ventricular hypertrophy; ACM, arrhythmogenic cardiomyopathy; DCM, dilated cardiomyopathy; HCM, hypertrophic cardiomyopathy; NS, non-significant.

Suarez-Mier
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AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive With Cardiovascular Abnormalities: Preamble, Principles, and General Considerations

A Scientific Statement From the American Heart Association and American College of Car

Barry J. Maron, MD, FACC, *Co-Chair**
Douglas P. Zipes, MD, FAHA, MACC, *Co-Chair**
Richard J. *Co-Chair**

This document addresses medical issues related to trained athletes with cardiovascular abnormalities. The objective is to present, in a readily useable format, consensus recommendations and guidelines principally addressing criteria for eligibility and disqualification from organized competitive sports for the purpose of ensuring the health and safety of young athletes. Recognizing certain medical risks imposed on athletes with cardiovascular disease, it is our aspiration that the recommendations that constitute this document will serve as a useful guide to the practicing communi making. The ultimate goal death in the young, although to unfairly or unnecessarily healthy athletic lifestyle or may be physiologically an twined with good quality o being) because of fear of liti the recommendations in t with sound clinical judgment safer playing field for young

*On behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology.

The American Heart Association and the American College of Cardiology make every effort to avoid any actual or potential conflicts of interest that may arise as a result of an outside relationship or a personal, professional, or business interest of a member of the writing panel. Specifically, all members of the writing group are required to complete and submit a Disclosure Questionnaire showing all such relationships that might be perceived as real or potential conflicts of interest. The Task Force reports for these proceedings are available online at www.onlinejacc.org. J Am Coll Cardiol 2015;XX:000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; and 000-000).

This statement was approved by the American Heart Association Science Advisory and Coordinating Committee on June 24, 2015, and the American Heart Association Executive Committee on July 22, 2015, and by the American College of Cardiology Board of Trustees and Executive Committee on June 3, 2015.

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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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ESC GUIDELINES

2020 ESC Guidelines on sports cardiology and exercise in patients with cardiovascular disease

The Task Force on sports cardiology and exercise in patients with cardiovascular disease of the European Society of Cardiology (ESC)

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† We would like to pay tribute to Professor Galderisi who passed away in March 2020.

ESC Committee for Practice Guidelines (CPG), National Cardiac Societies document reviewers and Author/Task Force Member affiliations: listed in the Appendix.

ESC entities having participated in the development of this document:

Associations: Association of Cardiovascular Nursing & Allied Professions (ACNAP), European Association of Cardiovascular Imaging (EACVI), European Association of Preventive Cardiology (EAPC), European Heart Rhythm Association (EHRA), Heart Failure Association (HFA).

Working Groups: Adult Congenital Heart Disease.

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ACC/AHA 2015

At present, it is unresolved whether resolution of myocarditis-related LGE should be required to permit return to competitive sports.

3. **Athletes with probable or definite myocarditis should not participate in competitive sports while active inflammation is present. This recommendation is independent of age, gender, and LV function *(Class III; Level of Evidence C).***

ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a cause of sudden death in young people and athletes, particularly in the northeastern (Veneto) region of Italy (54), but is seemingly less common in the United States (3). ARVC is characterized by a broad phenotypic spectrum and characteristically by loss of myocytes in the right ventricular myocardium, with fatty or fibrofatty replacement, which results in segmental or diffuse wall thinning, but there is also frequent involvement of the LV and an association with myocarditis (55). Genetics studies have demonstrated that ARVC is a desmosomal cardiomyopathy that results from genetically defective cell-adhesion proteins such as plakoglobin, plakophilin-2, desmoplakin, desmocollin-2, and desmoglein-2 (56,57).

Clinical diagnosis can be challenging but relies largely on familial occurrence, left bundle-branch pattern ventricular tachyarrhythmias, ECG findings of T-wave inversion in precordial leads V_1 through V_3, and epsilon waves, as well as right ventricular dilation or segmental wall motion abnormalities, aneurysm formation, or fatty deposition in the right ventricular wall identified with CMR imaging if substantial and unequivocal (or by biopsy tissue analysis). Diagnostic criteria for ARVC have been revised and updated and now include quantitative variables (58).

These criteria include global or regional structural dysfunction, as documented by echocardiography or CMR, biopsy abnormalities, ECG repolarization or depolarization abnormalities, arrhythmias, and family history. Each of these criteria is separated into major and minor criteria based on the severity of the finding. Patients meet an ARVC diagnosis if they possess 2 major, or 1 major and 2 minor, or 4 minor criteria. Borderline patients are those with 1 major and 1 minor criterion or 3 minor criteria. Patients with possible ARVC have 1 major criterion or 2 minor criteria. Athletes with borderline or possible ARVC, as well as those who are genotype positive-phenotype negative, should receive continued follow-up, because ARVC may progress phenotypically, and become more clinically apparent with time.

There is evidence in the experimental murine model that exercise increases the penetrance and arrhythmic risk in mutational carriers of ARVC (59). More recently, these data have been confirmed in genetically positive patients (60), which is particularly relevant to the athlete, raising concern not only with regard to competitive sports but also regarding participation in moderate to extreme recreational physical activities.

Ventricular tachyarrhythmias and sudden death in ARVC commonly occur during exertion, including competitive sports (55,60,61), and frequent endurance exercise increases the risk for ventricular tachycardia/ventricular fibrillation and heart failure (60). However, risk factors for sudden cardiac death in ARVC are not as well defined as in HCM (1,2,7,8). There is general agreement that a prior history of sudden cardiac death, sustained ventricular tachycardia, or syncope represent the most important prognostic factors and define many high-risk patients who are most appropriately treated with a primary prevention ICD (62-64).

Recommendations

1. **Athletes with a definite diagnosis of ARVC should not participate in most competitive sports, with the possible exception of low-intensity class 1A sports *(Class III; Level of Evidence C).***
2. **Athletes with a borderline diagnosis of ARVC should not participate in most competitive sports, with the possible exception of low-intensity class 1A sports *(Class III; Level of Evidence C).***
3. **Athletes with a possible diagnosis of ARVC should not participate in most competitive sports, with the possible exception of low-intensity class 1A sports *(Class III; Level of Evidence C).***
4. **Prophylactic ICD placement in athlete-patients with ARVC for the sole or primary purpose of permitting participation in high-intensity sports competition is not recommended because of the possibility of device-related complications *(Class III; Level of Evidence C).***

Other recommendations for sports participation in patients with ARVC and ICDs can be found in the Task Force 9 report on "Arrhythmias and Conduction Defects" (23).

PERICARDITIS

The causes of pericarditis/myopericarditis are varied and are either infectious or noninfectious. The natural history is incompletely resolved, although long-term prognosis is generally favorable. The diagnosis of acute pericarditis is typically based on clinical criteria: chest pain, pericardial rub, ST-segment elevation, or new/worsening pericardial effusion. This syndrome may be considered part of the clinical spectrum of myocarditis. Recurrences are a significant consideration, and follow-up surveillance with echocardiography or CMR is recommended to exclude pericardial thickening or restriction consistent with restrictive pericarditis (50).

- ARVC seltener in USA
- Sportler*innen mit "Borderline" oder möglicher ARVC oder Genotyp +/Phänotyp -

  → regelmäßige Folgeuntersuchungen

  Zeitraum ?
- Risiko der Penetranz nicht nur im Wettkampfsport relevant, sondern auch bei „moderate to extrem recreational physical activities"
- VT oder SCD üblicherweise bei Anstrengung einschl. Wettkampfsport und regelmäßigem Ausdauersport

ACC/AHA 2015

At present, it is unresolved whether resolution of myocarditis-related LGE should be required to permit return to competitive sports.

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Empfehlungen

1. Sportler mit eindeutiger Diagnose (2 major, 1/2 m+m, 4 minor)

   → kein Wettkampfsport, außer niedrig-intensiv

2. Sportler mit Borderline-Diagnose (1 major + 1 minor, 3 minor)

   → kein Wettkampfsport, außer niedrig-intensiv

3. Sportler mit möglicher Diagnose (1 major oder 2 minor)

   → kein Wettkampfsport, außer niedrig-intensiv

4. Keine primärprophylaktische ICD-Implantation zwecks Teilnahme an Wettkampfsport

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*).

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.	Class III/ Level C
These patients should be advised to limit their exercise programmes to leisure-time physical activities and remain under regular clinical surveillance.	

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 fibro-fatty 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

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.[75] 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'.[89–93] 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.[42,94]

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.

ESC 2019

- VT und SCD häufig durch Sport getriggert
- Diagnose auf Basis der ARVC-Kriterien
  - Ruhe-EKG
  - Echokardiographie und Kardio-MRT; Problem: physiologische Hypertrophie Ausdauersportler; ggf. Stress-Echokardiographie
  - Bel.-EKG/Spiroergo: Diagnose über Leistungsfähigkeit nicht möglich
  - LZ-EKG und Bel.-EKG: VES/VT mit LSB-Morphologie und super. Achse
  - Genetische Testung, aber: nur 50% Sensitivität, Problem “falsch-positive“

Risikostratifzierung

- mögliche Phenotyp-Triggerung der ACM durch regelmäßiges sportliches Training
- auch Ausdauersport ungünstig für die Penetranz
- RV Vergrößerung bei ACM-Sportlern im Vgl. zu ACM-Nicht-Sportlern
- Wettkampfsport assoziiert mit höherer Inzidenz von VT und Symptomen
- Wettkampfsport in jungen Jahren assoziiert mit früherer Symptomatik
- Sportreduktion führt zur Abnahme von VT oder SCD

→ Wettkampfsport insgesamt ungünstig

→ Freizeitsportprogramm mit geringerem Risiko wie bei inaktiven Patienten

Empfehlungen

- Eindeutige Aufklärung und umfassende Erklärung
- Sportler mit „exercise-induced RV cardiomyopathy“ ohne genetischen Nachweis unterliegen den gleichen Empfehlungen

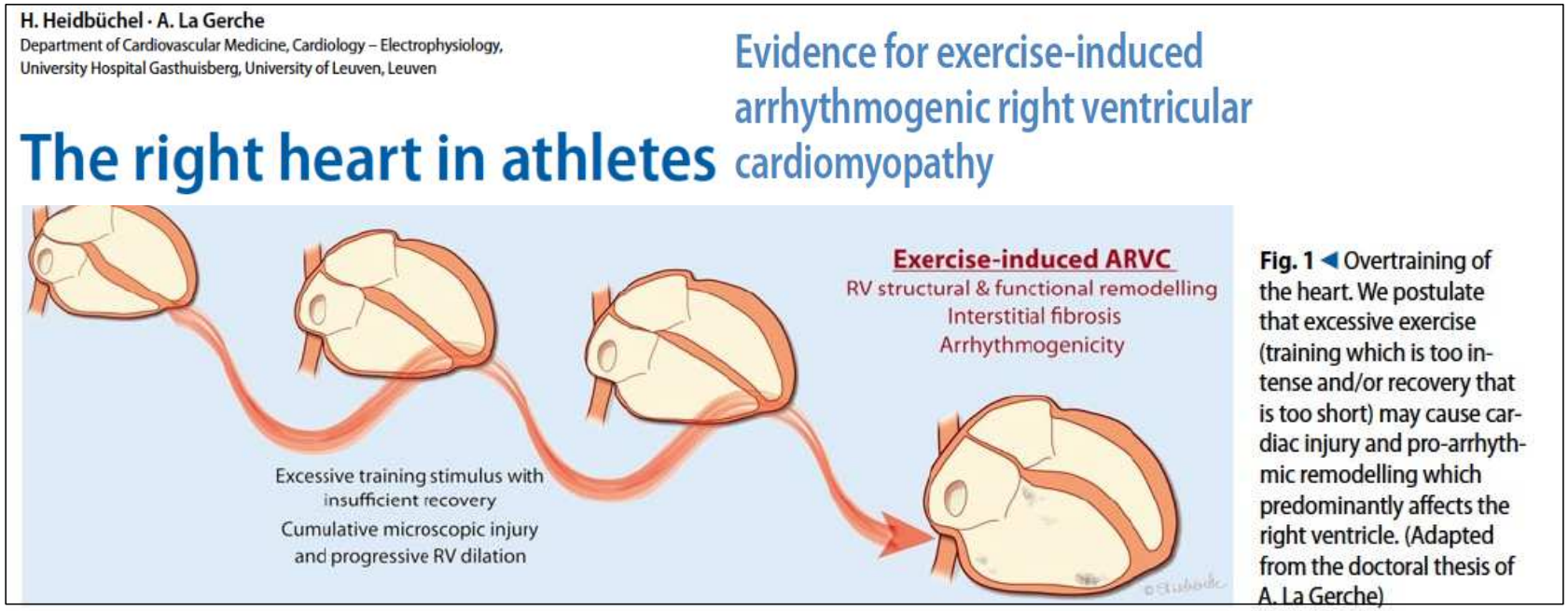


Fig. 1 ◀ Overtraining of the heart. We postulate that excessive exercise (training which is too intense and/or recovery that is too short) may cause cardiac injury and pro-arrhythmic remodelling which predominantly affects the right ventricle. (Adapted from the doctoral thesis of A. La Gerche)

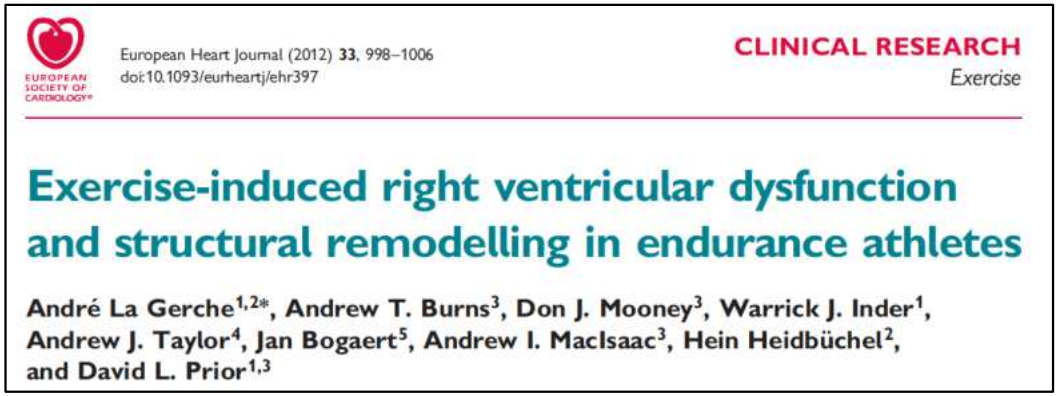
MAYO CLINIC

UNDER EMBARGO UNTIL JUNE 4, 2012, 12:00 AM ET

REVIEW

Potential Adverse Cardiovascular Effects From Excessive Endurance Exercise

James H. O'Keefe, MD; Harshal R. Patil, MD; Carl J. Lavie, MD; Anthony Magalski, MD; Robert A. Vogel, MD; and Peter A. McCullough, MD, MPH



European Heart Journal (2012) **33**, 998–1006
doi:10.1093/eurheartj/ehr397

EUROPEAN SOCIETY OF CARDIOLOGY®

CLINICAL RESEARCH
Exercise

Exercise-induced right ventricular dysfunction and structural remodelling in endurance athletes

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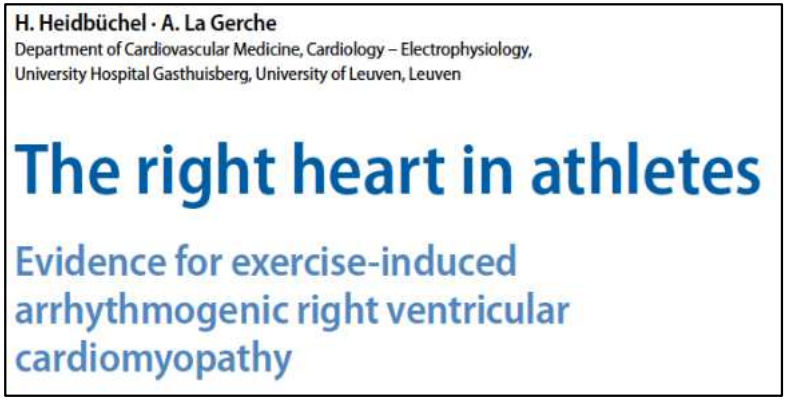

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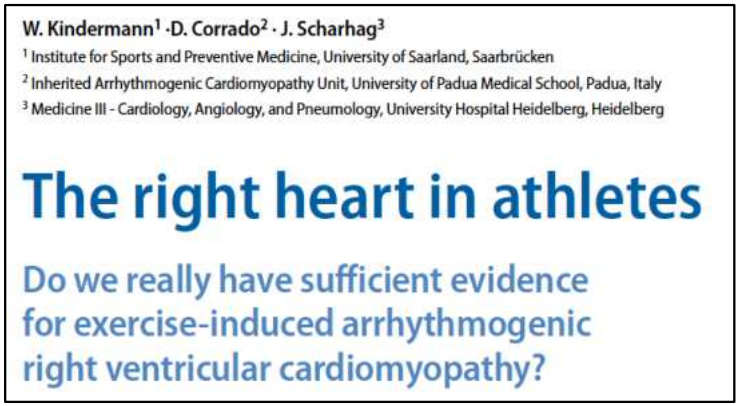
The right heart in athletes

Evidence for exercise-induced arrhythmogenic right ventricular cardiomyopathy



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The right heart in athletes

Do we really have sufficient evidence for exercise-induced arrhythmogenic right ventricular cardiomyopathy?

Empfehlungen

- Eindeutige Aufklärung und umfassende Erklärung
- Sportler mit „exercise-induced RV cardiomyopathy“ ohne genetischen Nachweis unterliegen den gleichen Empfehlungen

1. **Sportler mit eindeutiger oder wahrscheinlicher Diagnose**
   - → kein Wettkampfsport
   - → Sportprogramm reduzieren auf Freizeitaktivitäten unter klinischer Kontrolle
2. **Sportler mit ACM-assoziierten Desmosom-Mutationen ohne Phänotyp**
   - → kein Wettkampfsport
   - → Sportprogramm reduzieren auf Freizeitaktivitäten unter klinischer Kontrolle

Recommendations for exercise and sports participation in individuals with hypertrophic cardiomyopathy

Recommendations	Class[a]	Level[b]
Exercise recommendations		
Participation in high-intensity exercise/competitive sports, if desired (with the exception of those where occurrence of syncope may be associated with harm or death), may be considered for individuals who do not have any markers of increased risk[c] following expert assessment.	IIb	C
Participation in low- or moderate-intensity recreational exercise, if desired, may be considered for individuals who have any markers of increased risk[c] following expert assessment .	IIb	C
Participation in all competitive sports, if desired, may be considered for individuals who are gene positive for HCM but phenotype negative.	IIb	C
Participation in high-intensity exercise (including recreational and competitive sports) is not recommended for individuals who have ANY markers of increased risk[c].	III	C
Follow-up and further considerations relating to risk		
Annual follow-up is recommended for individuals who exercise on a regular basis.	I	C
Six-monthly follow-up should be considered in adolescent individuals and young adults who are more vulnerable to exercise-related SCD.	IIa	C
Annual assessment should be considered for genotype-positive/phenotype-negative individuals for phenotypic features and risk stratification purposes.	IIa	C

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BP = blood pressure; ESC = European Society of Cardiology; HCM = hypertrophic cardiomyopathy; LVOT = left ventricular outflow tract obstruction cardiomyopathy; SCD = sudden cardiac death.
[a]Class of recommendation.
[b]Level of evidence.
[c]Markers of increased risk include: (i) cardiac symptoms or history of cardiac arrest or unexplained syncope; (ii) moderate ESC risk score (≥4%) at 5 years; (iii) LVOT gradient at rest >30 mmHg; (iv) abnormal BP response to exercise; (v) exercise-induced arrhythmias.
Refer to *Table 4* for different indices of exercise intensity and training zones.

5.5.2 Arrhythmogenic cardiomyopathy

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is defined pathologically by the presence of fibro-fatty replacement of the right ventricle and clinically by life-threatening VAs. The condition was initially recognized as a predominantly RV disease, and diagnosis is currently based on probabilistic Task Force Criteria that encompass electrophysiological, anatomical, functional, and clinical features of the disease.[375] Since its first description, the concept of ARVC has evolved to include concealed or subclinical phenotypes and biventricular disease. It is now well established that both ventricles are affected in most cases.[376–378] This has led to the development of a new term, arrhythmogenic cardiomyopathy, that embraces an array of diagnostic terms for different (genetic and acquired) pathologies. Although the definition of 'arrhythmogenic cardiomyopathy' is yet to be agreed, it can be considered as an umbrella term for a family of diseases that are characterized by biventricular myocardial abnormalities, including fibro-fatty infiltration and scarring, identified by pathological examination and/or cardiac imaging and VA.

The term arrhythmogenic cardiomyopathy (ACM) is used throughout these recommendations; however, it is important to recognize that most of the literature on the influence of exercise on disease progression and risk of SCD is derived from cohorts with classical ARVC. This is reflected in the recommendations provided in these Guidelines. It is possible therefore that the recommendations may not accurately reflect predominantly LV disease, which constitutes a small proportion of the disease spectrum where the impact of exercise on disease phenotype and risk is less clarified than the RV variant. Where appropriate, guidance is provided for other conditions that can be reasonably considered under the umbrella of ACM [including subtypes of dilated cardiomyopathy (DCM)].

5.5.2.1 Risk stratification in arrhythmogenic cardiomyopathy

ACM accounts for a significant proportion of SCDs in young and athletic individuals.[28] Established risk factors for SCD that should prompt consideration for an ICD include aborted SCD, unheralded syncope, ventricular tachycardia, and impaired RV and/or LV systolic function.[379] A novel risk prediction model for VAs has recently been proposed but is yet to be validated.[380] Regular and high-intensity exercise programmes are associated with acceleration of the disease process and worse outcomes.[381–389]

In an experimental model of heterozygous plakoglobin-deficient mice, exercise training accelerated RV dysfunction and arrhythmias.[382] Similar results have been confirmed in human desmosomal mutation carriers participating in vigorous (>70% VO_{2max}) endurance sports.[384] Similar findings were reported in patients with ACM and asymptomatic gene-positive family members, despite a more conservative definition of athletic status (exercise with intensity ≥6 METs for ≥4 h/week for ≥6 years).[386] Recently, the results from the North American multidisciplinary study reported that patients engaging in competitive sports were at two-fold increased risk of ventricular tachyarrhythmias or death and earlier presentation of symptoms, compared with patients who participated in recreational sports and sedentary individuals.[385] Among patients engaging in competitive sports, early age of sports 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.[385] Finally, in a multinational registry of 393 competitive athletes implanted with an ICD who continued to participate in regular competitions, 20% of athletes with ACM received a shock during exertion compared to 10% at rest, during a median follow-up of 44 months. The diagnosis of ACM was the only variable associated with receiving appropriate shocks during competition.[359,389]

5.5.2.2 Baseline assessment of patients with arrhythmogenic cardiomyopathy

A systematic approach is required when assessing individuals with ACM who request exercise advice. The baseline evaluation should include a comprehensive history of symptoms and family history of ACM or SCD, assessment of the severity of the ACM phenotype, and the presence of any conventional risk factors for SCD/SCA.

5.5.2.3 History

Syncope due to presumed arrhythmia is an important risk marker for SCD/SCA and a predictor of future appropriate ICD therapies.[390–394] The presence of symptoms attributed to ACM should reinforce the conservative exercise recommendations. Individuals with a history of cardiac arrest or unheralded syncope and individuals with exercise-induced symptoms should be advised to engage only in low-intensity recreational exercise programmes.

5.5.2.4 Resting and ambulatory ECG

Apart from its diagnostic utility, the 12-lead ECG may provide useful information relating to risk stratification in ACM. The presence of extensive T-wave inversion affecting ≥3 precordial leads or T-wave inversion in two of the three inferior leads confers some additional risk for SCD/SCA.[395,396]

Ambulatory ECG monitoring is important for detecting VAs. Every effort should be made for the monitoring period to include the proposed exercise session. The presence of NSVT or significant burden of ventricular ectopy (≥1000/24 h), even in asymptomatic individuals, confers an increased risk of fatal arrhythmias.[392,393,397]

5.5.2.5 Echocardiography and cardiac magnetic resonance imaging

In relation to risk stratification for SCD, the clinician should assess the severity of RV and LV involvement in terms of ventricular dilatation and systolic dysfunction. CMR imaging is more useful than echocardiography for assessing RV wall motion abnormalities and can also quantify the degree of myocardial fat infiltration and/or scar. The more extensive the disease the higher the arrhythmic risk.[398,399]

5.5.2.6 Exercise testing

Exercise testing should be part of the routine assessment of every individual with ACM who wishes to exercise, as it can provide information regarding functional capacity and risk stratification. Exercise testing in patients with ACM should not be performed during 'hot phases'. The presence of exercise-induced symptoms or arrhythmias should result in more conservative recommendations.

5.5.2.7 Genetic testing

Genotype may also be of prognostic value. In the ARVC variant, a number of studies have reported that carriers of multiple pathogenic variants in the same desmosomal gene or mutations in ≥2 genes may have an almost four-fold higher arrhythmic risk than those with a single mutation.[400] Particular genotypes such as DSP and TMEM43, but also LMNA and FLNC, associated with other ACM phenotypes (see section *5.5.4*) have a propensity for high arrhythmic burden that can pre-date the structural phenotype.[401,402]

5.5.2.8 Exercise recommendations

The overall scientific evidence supports the concept that in patients with ACM participation in high-intensity sports should be discouraged, because it is associated with accelerated disease progression, greater risk of VAs and major events. This recommendation is also applicable to genetic carriers of pathogenic variants for ACM even in the absence of overt disease phenotype.

5.5.2.9 Special considerations

Young age of presentation and male sex are associated with increased risk of malignant arrhythmias in ACM.[379] Although young age should not exclude an individual from moderate-intensity exercise in the absence of high-risk features, age should be considered in the discussion with the patient and the parents. In addition, one should consider that specific highly dynamic, start–stop sports, such as basketball and football, may pose a higher risk of SCD particularly in athletes who compete at the highest level.[37,365]

5.5.2.10 Follow-up

An annual follow-up is recommended for most individuals with ACM who exercise on a regular basis. More frequent (6-monthly) follow-up should be considered for adolescent and young adults whose ACM phenotype, and therefore risk of SCD, may still be evolving, particularly if they engage in moderate- to high-intensity exercise. More frequent follow-up should also be considered in individuals with high arrhythmic risk genotypes such as DSP, TMEM43, and carriers of multiple pathogenic variants. New symptoms should prompt interruption of exercise and re-evaluation.

Recommendations for exercise and sports participation in individuals with arrhythmogenic cardiomyopathy

Recommendations	Class[a]	Level[b]
Exercise recommendations		
Participation in 150 min of low-intensity exercise per week should be considered for all individuals.	IIa	C
Participation in low- to moderate-intensity recreational exercise/sports, if desired, may be considered for individuals with no history of cardiac arrest/VA, unexplained syncope, minimal structural cardiac abnormalities, <500 PVCs/24 h and no evidence of exercise-induced complex VAs.	IIb	C
Participation in high-intensity recreational exercise/sports or any competitive sports is not recommended in individuals with ACM, including those who are gene positive but phenotype negative.[384,386]	III	B
Follow-up and further considerations relating to risk		
Annual follow-up is recommended for individuals who exercise on a regular basis.	I	C
Six-monthly follow-up should be considered in adolescent individuals and young adults who are more vulnerable to exercise-related SCD.	IIa	C
Annual assessment should be considered for genotype-positive/phenotype-negative individuals for phenotypic features and risk stratification purposes.	IIa	C
Six-monthly follow-up should also be considered in individuals with high arrhythmic risk genotypes such as DSP, TMEM43, and carriers of multiple pathogenic variants.	IIa	C

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ACM = arrhythmogenic cardiomyopathy; PVC = premature ventricular contraction; SCD = sudden cardiac death; VA = ventricular arrhythmia.
[a]Class of recommendation.
[b]Level of evidence.
Refer to *Table 4* for different indices of exercise intensity and training zones.

Risikostratifzierung

- s.v.
- 2 x so häufig ICD-Schocks bei 393 Wettkampfsportlern während Belastung wie in Ruhe (20% vs. 10%) während 44 Monaten

Diagnostik

- Anamnese: Synkope → nur niedrig intensive Belastungen
- EKG, LZ-EKG (NSVT , >1.000 VES/24h)
- Echokardiographie und Kardio-MRT
- CPX: bei allen sportlich aktiven Patienten, HRST?
- Gentests
- Sportempfehlungen, Abraten von hoch-intensivem oder hoch-dynam. Sport
- Verlaufskontrollen jährlich bis dreimonatlich

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Recommendations	Class[a]	Level[b]
Exercise recommendations		
Participation in 150 min of low-intensity exercise per week should be considered for all individuals.	IIa	C
Participation in low- to moderate-intensity recreational exercise/sports, if desired, may be considered for individuals with no history of cardiac arrest/VA, unexplained syncope, minimal structural cardiac abnormalities, <500 PVCs/24 h and no evidence of exercise-induced complex VAs.	IIb	C
Participation in high-intensity recreational exercise/sports or any competitive sports is not recommended in individuals with ACM, including those who are gene positive but phenotype negative.[384,386]	III	B
Follow-up and further considerations relating to risk		
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Six-monthly follow-up should be considered in adolescent individuals and young adults who are more vulnerable to exercise-related SCD.	IIa	C
Annual assessment should be considered for genotype-positive/phenotype-negative individuals for phenotypic features and risk stratification purposes.	IIa	C
Six-monthly follow-up should also be considered in individuals with high arrhythmic risk genotypes such as DSP, TMEM43, and carriers of multiple pathogenic variants.	IIa	C

ESC 2020

150 min niedrig-intensive Aktivität pro Woche

Sport mit niedriger bis mittlerer Intensität möglich

< 500 VES/24h

keine bel.-induzierten komplexen VA

Keine hoch-intensiven Bel. oder Wettkampfsport

Jährliche Kontrollen

6 Mo-Kontrollen Jugendliche und junge Erwachsene

Jährliche Kontrollen G+/P-

6 Mo-Kontrollen bei Risiko-Genotypen

Take home

Nein!

- keinen Wettkampfsport
- keinen intensiven oder hoch-dynamischen Freizeitsport
- keinen “erschöpfenden“ Ausdauersport

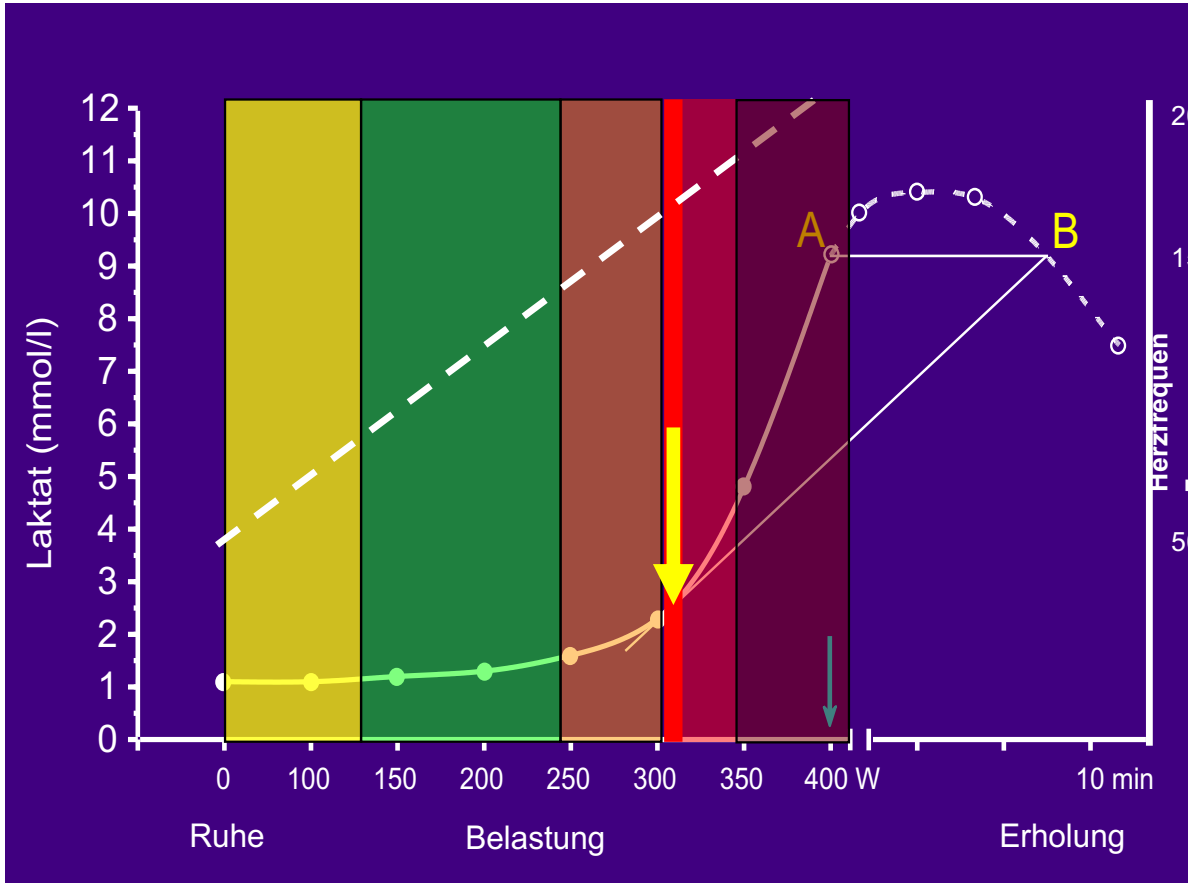
Vielleicht

- Sport mit mittlerer Intensität bei unauffälligem Bel.-Tests inkl. LZ-EKG

Ja

- Gesundheitssport im regenerativ-extensiven Bereich

Trainingsbereiche



Laktat und Spiroergometie

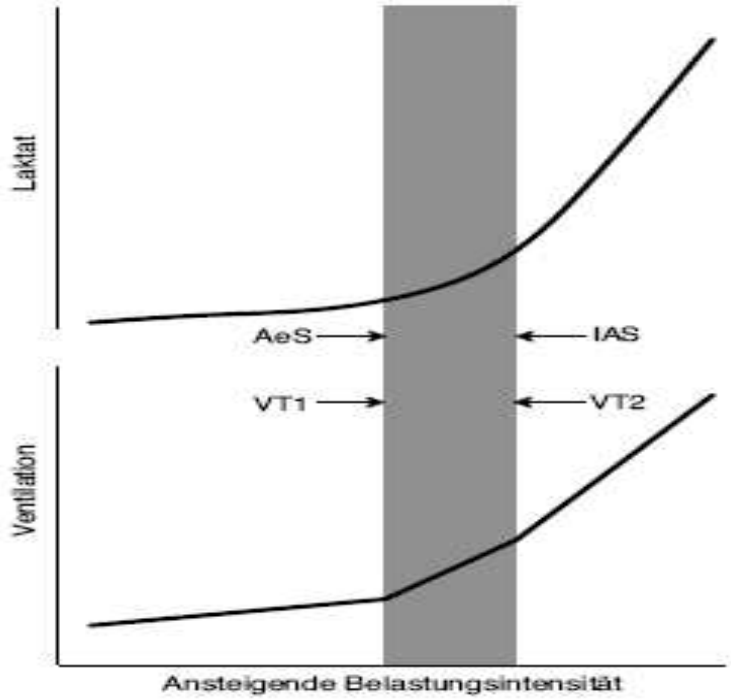


Abbildung 1: Schematische Darstellung des aerob-anaeroben Übergangs (grauer Bereich). Laktat-Leistungskurve (oben) und Ventilation (unten) bei ansteigender Belastungsintensität. AeS: aerobe Schwelle; IAS: individuelle anaerobe Schwelle; VT1: ventilatorische Schwelle 1; VT2: ventilatorische Schwelle 2 (respiratorischer Kompensationspunkt)

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