Trening for pasienter med arvelige kardiomyopatier

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Oversikt

- Center for Cardiological Innovation og
  Enhet for genetisk kardiologi og kardiomyopati
- Arvelige kardiomyopatier
- Hjertet, trening og sykdom
- Spesifikke anbefalinger og kunnskapshull

Oversikt

- **Center for Cardiological Innovation og Enhet for genetisk kardiologi og kardiomyopati**
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- Spesifikke anbefalinger og kunnskapshull

Center for Cardiological Innovation

- Center director: Professor Thor Edvardsen
- Center Director of Cardiology Research: Ass. Professor Kristina H Haugaa
- Hosted by Department of Cardiology, Oslo University Hospital
- Research partners: University of Oslo, Simula Research Center
- Industrial partners
- Next generation ultrasound, sudden cardiac death, heart failure

Enhet for genetisk kardiologi og kardiomyopati

- Leder: Kristina Hermann Haugaa, overlege og førsteamanuensis
- Kardiologisk poliklinikk, OUS Rikshospitalet
- Utredning, behandling og oppfølging av voksne pasienter med arvelige hjertesykdommer: ionekanalsykdommer og kardiomyopatier
- Team av leger og sykepleiere
- Tett samarbeid med Avdeling for medisinsk genetikk

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Kardiomyopati

1.
A myocardial disorder in which the heart muscle is structurally and functionally abnormal, in the absence of coronary artery disease, hypertension, valvular disease and congenital heart disease sufficient to cause the observed myocardial abnormality.

Elliott, Eur Heart J 2008

2.
A heterogeneous group of diseases of the myocardium associated with mechanical and/or electrical dysfunction that usually (but not invariably) exhibit inappropriate ventricular hypertrophy or dilatation and are due to a variety of causes that frequently are genetic.

Harrison`s principles of internal medicine 2012

Hypertrofisk kardiomyopati

Hypertrofisk kardiomyopati

European Heart Journal (2014) **35**, 2733–2779
doi:10.1093/eurheartj/ehu284

ESC GUIDELINES

2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy

The Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC)

Hypertrofisk kardiomyopati

Increased left ventricular wall thickness not explained by abnormal loading conditions.

Left ventricular hypertrophy in the absence of hypertension and valve disease occurs in approximately 1:500 of the general population.

Hypertrofisk kardiomyopati

- Left ventricular outflow obstruction
- Diastolic dysfunction
- Myocardial ischemia
- Mitral regurgitation

→

- heart failure
- chest pain
- arrhythmias

Dilatert kardiomyopati

Dilatert kardiomyopati

European Heart Journal
doi:10.1093/eurheartj/ehv727

ESC REPORT

Proposal for a revised definition of dilated cardiomyopathy, hypokinetic non-dilated cardiomyopathy, and its implications for clinical practice: a position statement of the ESC working group on myocardial and pericardial diseases

Dilatert kardiomyopati

Left ventricular dilatation and systolic dysfunction.
Right ventricular dilation and dysfunction may be present.

>25% of patients in Western populations have evidence for familial disease with predominantly autosomal dominant inheritance.

Dilatert kardiomyopati – Lamin A mutasjon

- Conduction defects and arrhythmias before left ventricular dysfunction and dilatation
- Poor prognosis compared with noncarriers of LMNA mutations
- High rate of major cardiac events such as sudden cardiac death, malignant ventricular tachycardia, extreme bradycardia due to high degree atrioventricular block, atrial standstill and end-stage heart failure.
- The progressive heart failure eventually becomes resistant to treatment
- No drugs are curative
- Heart transplantation is frequently necessary

Risk factors associated with major cardiac events:

- Ejection fraction <45%
- Nonsustained ventricular tachycardia
- Highly dynamic competitive sport activity
- Male sex
- Nonmissense LMNA mutations

Cattin, Curr Opin Cardiol 2013

Arytmogen høyre ventrikkeldysplasi

Arytmogen høyre ventrikkeldysplasi

Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia

Proposed Modification of the Task Force Criteria

Circulation. 2010;121:1533-1541.

Arytmogen høyre ventrikkeldysplasi

Right ventricular dysfunction (global or regional), with or without left ventricular disease, in the presence of histological evidence for the disease and/or electrocardiographic abnormalities

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- **Spesifikke anbefalinger og kunnskapshull**

Anbefalinger og retningslinjer

2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy

Anbefalinger og retningslinjer

AHA/ACC Scientific Statement

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 3: Hypertrophic Cardiomyopathy, Arrhythmogenic Right Ventricular Cardiomyopathy and Other Cardiomyopathies, and Myocarditis

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

Anbefalinger og retningslinjer

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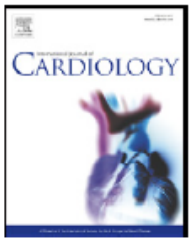

Exercise restrictions for patients with inherited cardiac conditions: Current guidelines, challenges and limitations

Matthew Hammond-Haley [a], Riyaz S. Patel [a,b], Rui Providência [b], Pier D. Lambiase [a,b,*]

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Hypertrofisk kardiomyopati

Anbefalinger og retningslinjer – Hypertrofisk kardiomyopati

1. Participation in competitive athletics for asymptomatic, genotype-positive HCM patients without evidence of LV hypertrophy by 2-dimensional echocardiography and CMR is reasonable, particularly in the absence of a family history of HCM-related sudden death (Class IIa; Level of Evidence C).

2. Athletes with a probable or unequivocal clinical expression and diagnosis of HCM (ie, with the disease phenotype of LV hypertrophy) should not participate in most competitive sports, with the exception of those of low intensity (class IA sports).

This recommendation is independent of age, sex, magnitude of LV hypertrophy, particular sarcomere mutation, presence or absence of LV outflow obstruction (at rest or with physiological exercise), absence of prior cardiac symptoms, presence or absence of late gadolinium enhancement (fibrosis) on CMR, and whether major interventions such as surgical myectomy or alcohol ablation have been performed previously (Class III; Level of Evidence C).

Anbefalinger og retningslinjer – Hypertrofisk kardiomyopati

2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy

- Patients with HCM should avoid competitive sports activities, but should maintain a healthy lifestyle

- Advice on recreational activities should be tailored to symptoms and the risk of disease-related complications including sudden cardiac death

Anbefalinger og retningslinjer – Hypertrofisk kardiomyopati

Disorder	Summary of the major AHA and ESC exercise recommendations		
	AHA/ACC (2005) [67,78]	AHA/ACC (2015) [58,63]	ESC [62,70]
HCM	High intensity recreational sports are not advised or strongly discouraged and moderate intensity sports should be assessed clinically on an individual basis. Low intensity non-competitive sports participation is probably permitted Athletes should be limited to low intensity competitive sports only There is insufficient evidence to restrict G + P − patients from most modest recreational sports activities	Athletes with probable or unequivocal clinical expression of HCM should not participate in competitive sports, with the exception of those of low intensity. (*level of evidence C*) G + P − patients without evidence of LV hypertrophy by echocardiography may reasonably participate in competitive sports, particularly if there is no family history of HCM-related SCD (*level of evidence C*)	No competitive sports for athletes with a definitive diagnosis of HCM, with the exception of low dynamic, low static sports, if considered low risk G + P − patients may partake in non-competitive recreational sporting activities

Pasienter med hypertrofisk kardiomyopati skal ikke delta i konkurranseidrett.

Personer med mutasjon kan trene, men konkurranseidrett?

Anbefalinger og retningslinjer – LV non-compaction

1. Participation in competitive sports may be considered for asymptomatic patients with a diagnosis of LVNC and normal systolic function, without important ventricular tachyarrhythmias on ambulatory monitoring or exercise testing, and specifically with no prior history of unexplained syncope.
   (Class IIb; Level of Evidence C)

2. Athletes with an unequivocal diagnosis of LVNC and impaired systolic function or important atrial or ventricular tachyarrhythmias on ambulatory monitoring or exercise testing (or with a history of syncope) should not participate in competitive sports, with the possible exception of low-intensity class 1A sports, at least until more clinical information is available.
   (Class III; Level of Evidence C)

Dilatert kardiomyopati

Anbefalinger og retningslinjer – DCM

Symptomatic athletes with DCM, primary nonhypertrophied restrictive cardiomyopathy, and infiltrative cardiac myopathies should not participate in most competitive sports, with the possible exception of low-intensity (class 1A sports) in selected cases, at least until more information is available *(Class III; Level of Evidence C)*.

Arytmogen høyre ventrikkeldysplasi

Exercise Increases Age-Related Penetrance and Arrhythmic Risk in Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy–Associated Desmosomal Mutation Carriers

James, Journal of the American College of Cardiology, 2013

Anbefalinger og retningslinjer – ARVC

EUROPEAN SOCIETY OF CARDIOLOGY®

European Journal of Heart Failure (2014) **16**, 1337–1344
doi:10.1002/ejhf.181

Vigorous physical activity impairs myocardial function in patients with arrhythmogenic right ventricular cardiomyopathy and in mutation positive family members

Jørg Saberniak[1,2], Nina E. Hasselberg[1,2], Rasmus Borgquist[3], Pyotr G Platonov[3], Sebastian I. Sarvari[1,2], Hans-Jørgen Smith[4], Margareth Ribe[1,2], Anders G. Holst[5], Thor Edvardsen[1,2], and Kristina H. Haugaa[1,2]*

Anbefalinger og retningslinjer – ARVC

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).

Anbefalinger og retningslinjer – ARVC

Disorder	Summary of the major AHA and ESC exercise recommendations		
	AHA/ACC (2005) [67,78]	AHA/ACC (2015) [58,63]	ESC [62,70]
ARVC	Athletes with probable or definite diagnosis of ARVC should be excluded from most competitive sports, with the possible exception of those of low intensity. High intensity recreational sports are generally not advised or strongly discouraged. Most low intensity recreational sports are probably permitted and moderate activities should be assessed on an individual basis.	Athletes with a definite, borderline, or possible diagnosis of ARVC should not participate in most competitive sports, with the possible exception of low-intensity sports (*level of evidence C*)	Patients with a definite diagnosis of ARVC should not participate in competitive and/or endurance sports and should be restricted from participation in all athletic activities, with the possible exception of recreational low-intensity sports. Restriction from competitive sports activity may be considered in ARVC family members with a negative phenotype, both G+P− or unknown genotype [79].

Pasienter med ARVC skal ikke trene med høy intensitet

- hvor går grensen?

Personer med uklar fenotype eller kjent mutasjon bør vurdere treningsrestriksjon.

Sammendrag

- Center for Cardiological Innovation (Forskning)
- Enhet for genetisk kardiologi og kardiomyopati (henvisninger)
- Kardiomyopati er ikke bare «syke hjerter»
- Trening er medisin – kan ha bivirkninger, må doseres!
- Trening må rekvireres ut fra diagnose og stadium
- Konkurranseidrett må vurderes av spesialistsenter