Management of Atrial tachyarrhythmia

1. Rate control using ß-blockers and non-dihydropyridine calcium channel antagonists, alone or in combination, is recommended in patients with paroxysmal, persistent or permanent AF*. Digoxin and Class IC anti-arrhythmics should be avoided. Amiodarone should be considered for rhythm control and to maintain sinus rhythm after cardioversion.

2. In new onset AF*, elective DC* cardioversion should be considered after a minimum of 3 weeks of effective anticoagulation with a vitamin K antagonist.

3. Use of the CHA2DS2-VASc score to calculate stroke risk is NOT recommended in patients with HCM*.

4. Life long therapy with oral anticoagulants is recommended even when sinus rhythm is restored.

Sudden death prevention

1. Patients with HCM* should be advised not to participate in competitive sports and discouraged from intense physical activity, especially when they have risk factors for sudden cardiac death or left ventricular outflow tract obstruction.

2. ICD* implantation is recommended in patients who have survived a cardiac arrest due to ventricular fibrillation or experienced spontaneous sustained ventricular tachycardia causing haemodynamic compromise.

3. Risk assessment in all other patients should include clinical evaluation, family history, 48 hour ambulatory ECG*, TTE* (or CMR* in the case of poor echo windows) and a symptom limited exercise test. A predefined set of prognostic variables are then used to estimate the 5-year risk of SCD* using the HCM* Risk-SCD* model (see online calculator http://doc2do.com/hcm/webHCM.html) in order to provide advice on prophylactic ICD* therapy.

Routine follow-up

1. A clinical evaluation, including 12-lead ECG* and transthoracic echocardiogram is recommended every 12–24 months in clinically stable patients and whenever there is a change in symptoms.

2. 48-hour ambulatory ECG* is recommended every 12–24 months in clinically stable patients, every 6–12 months in patients in sinus rhythm with left atrial dimension ≥45 mm, and whenever patients complain of new palpitations.

**AF = atrial fibrillation; CMR = cardiac magnetic resonance; DC = direct current; ECG = electrocardiogram; HCM = hypertrophic cardiomyopathy ; ICD = implantable cardioverter defibrillator; SCD = sudden cardiac death; TTE = transthoracic echocardiogram.*

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General lifestyle considerations for patients with hypertrophic cardiomyopathy

Topic	General guidance
Exercise	• 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.
Diet, alcohol and weight	• Patients should be encouraged to maintain a healthy body mass index. • Large meals can precipitate chest pain, particularly in patients with LVOTO. Smaller, more frequent meals may be helpful. • Avoid dehydration and excess alcohol, particularly in patients with LVOTO. • Constipation is a frequent side-effect of verapamil/disopyramide and should be managed with diet and if necessary aperients.
Smoking	• There are no data that show an interaction between tobacco smoking and HCM, but patients should be provided with general advice on the health risks associated with smoking and, when available, information on smoking cessation.
Sexual activity	• Patients should be given the opportunity to discuss their concerns about sexual activity. Anxiety and depression following a diagnosis are frequent and some patients may express guilt or fear about their genetic diagnosis and the risk of transmission to offspring. • Patients should be counselled on the potential effect of their medication on sexual performance. • In general, patients should avoid PDE_5 inhibitors, particularly when they have LVOTO.
Medication	• Patients should be provided with information about their medication, including potential side-effects and interactions with prescribed medications, over-the-counter remedies and other complementary therapies. • Where possible, peripheral vasodilators should be avoided in patients, particularly when they have LVOTO.
Vaccination	• In the absence of contra-indications, symptomatic patients should be advised to have yearly influenza vaccination.
Driving	• Most patients should be eligible for an ordinary driving licence and can continue driving unless they experience distracting or disabling symptoms. • Advice on driving licences for heavy goods or passenger-carrying vehicles should be in line with local legislation. • For further advice on driving with ICD see EHRA guidance[a] and local rules.
Occupation	• Most people with HCM will be able to continue in their normal job. The implications of heavy manual jobs that involve strenuous activity should be discussed with the appropriate specialist. • For some occupations such as pilots, and military and emergency services, there are strict guidelines on eligibility. • The social and financial implications of a diagnosis of HCM should be included in the counselling of relatives before clinical or genetic screening.
Holidays and travel insurance	• Most asymptomatic or mildly symptomatic patients can fly safely. For further advice see Fitness to fly for passengers with cardiovascular disease[b]. • Insurance companies may charge more for travel insurance. In some countries, patient support organizations can provide further advice about obtaining reasonable insurance.
Life insurance	• The diagnosis of HCM will result in difficulty obtaining life insurance or mortgages. Advice on the rules that apply in different countries should be provided to patients at diagnosis.
Pregnancy and childbirth	• Counselling on safe and effective contraception and referral for specialist pre-pregnancy risk assessment is indicated in all women of fertile age.
Education/ schooling	• Teachers and other carers should be provided with advice and written information relevant to the care of children with HCM. • In the absence of symptoms and risk factors, children should be allowed to perform low to moderate level aerobic physical activity, in accordance with advice from their cardiologist. • Provision should be made for children with learning difficulties and other special needs.

HCM = hypertrophic cardiomyopathy; LVOTO = left ventricular outflow tract obstruction; PDE_5 = phosphodiesterase 5.
[a]Vijgen J et al. Eur J Cardiovasc Nurs.
[b]Smith D, et al. Heart 2010;96 Suppl 2:ii 1–16.

For more information
www.escardio.org/guidelines

SUMMARY CARD FOR GENERAL PRACTICE

Committee for Practice Guidelines
To improve the quality of clinical practice and patient care in Europe

HCM

GUIDELINES FOR THE DIAGNOSIS AND MANAGEMENT OF HYPERTROPHIC CARDIOMYOPATHY

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Definition

Hypertrophic cardiomyopathy (HCM) is defined by the presence of increased left ventricular (LV) wall thickness not solely explained by abnormal loading conditions. In adults the diagnostic criterion is an LV wall thickness ≥15 mm in one or more LV myocardial segments assessed by any imaging technique and in children, an LV wall thickness more than two standard deviations above the predicted mean. Defined in this way, HCM affects around 0.2% of adults.

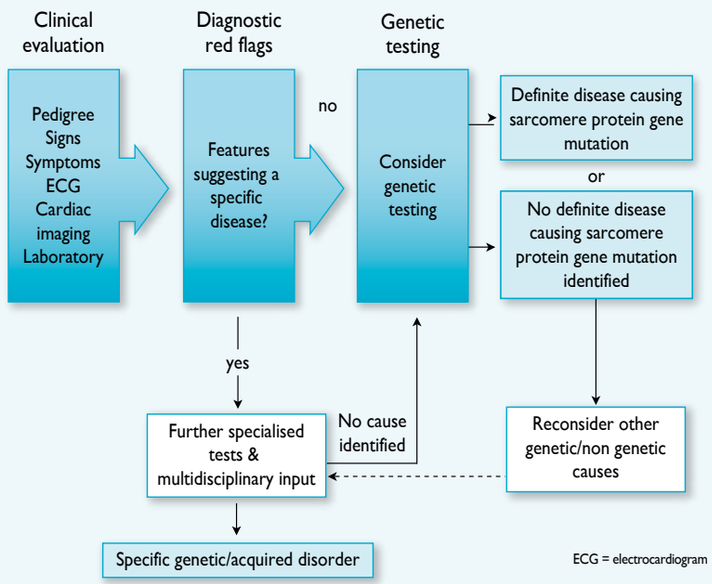
Aetiology

In up to 60% of adolescents and adults with HCM, the disease is an autosomal dominant trait caused by mutations in cardiac sarcomere protein genes. Five to ten per cent of adult cases are caused by other genetic disorders including inherited metabolic and neuromuscular diseases, chromosome abnormalities and genetic syndromes. Some patients have non-genetic disorders that mimic genetic forms of the disease.

Clinical Presentation

Many individuals with HCM complain of few, if any, symptoms. In such cases the diagnosis can be incidental or the result of screening. Some patients experience angina, dyspnoea, palpitations and syncope caused by dynamic left ventricular outflow tract obstruction, systolic and diastolic LV dysfunction and arrhythmia.

Schematic summarising the general approach to the diagnosis of hypertrophic cardiomyopathy



From the ESC Guidelines on the diagnosis and management of hypertrophic cardiomyopathy (European Heart Journal (2014) 35, 2733–2779 - doi 10.1093/eurheartj/ehu284) - Chairperson: Perry M.Elliott (UK). Cardiology Department - The Heart Hospital - 16-18 Westmoreland Street - London W1G 8PH, UK Email: perry.elliott@ucl.ac.uk .

Recommended tests in patients with definite or suspected HCM

1. Standard 12-lead electrocardiography.

2. Transthoracic 2-D and Doppler echocardiography (including assessment of left ventricular outflow tract obstruction at rest and during Valsalva manoeuvre in the sitting and semi-supine positions).

3. Upright exercise testing

4. 48 hour ambulatory ECG monitoring.

5. Cardiac magnetic resonance imaging should be considered if local resources and expertise permit.

Recommendations for genetic counselling and testing

1. Genetic counselling by trained professionals working within a multidisciplinary specialist team is recommended for all patients with HCM when their disease cannot be explained solely by a non-genetic cause.

2. Genetic testing is recommended in patients fulfilling diagnostic criteria for HCM when it enables cascade genetic screening of their relatives.

3. First degree relatives should be provided with information about the consequences of a diagnosis for life insurance, pension, occupation, sporting activities, and eligibility for fostering and adoption before they undergo genetic testing or clinical evaluation.

4. When a definite causative genetic mutation is identified, relatives should be first genetically tested and then clinically evaluated if they are found to carry the same mutation.

5. When genetic testing cannot be performed or fails to identify a definite mutation, first degree relatives should be offered clinical screening with an ECG and echocardiogram which is then repeated every 1-2 years between 10 and 20 years of age and then every 2-5 years thereafter.

6. Clinical and genetic testing of children should be guided by the best interests of the child and consider potential benefits and harms such as compromised life insurance prospects.

Management and prevention of important complications of HCM: heart failure, atrial fibrillation, and sudden cardiac death

Individuals who have severe symptoms or markers of an increased risk for disease-related complications should be referred to specialist teams for further investigation and management.

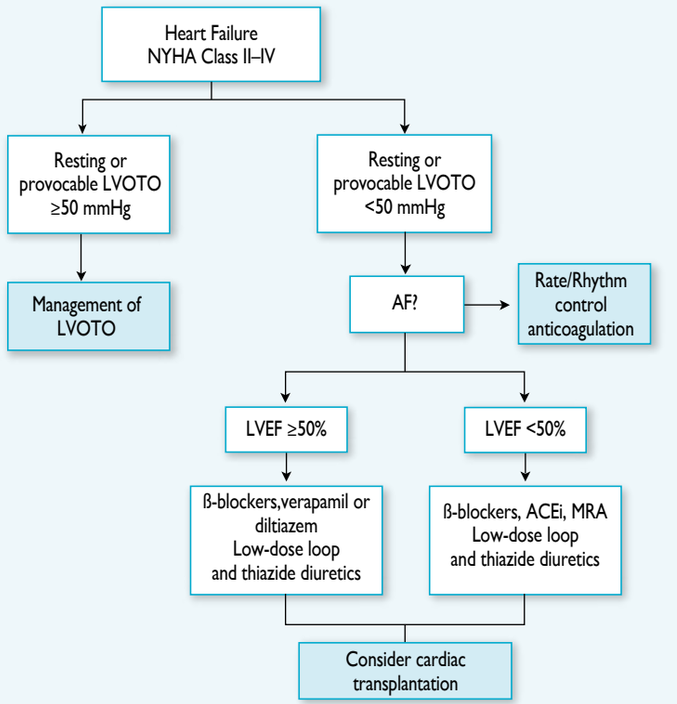
Management of left ventricular outflow tract obstruction

1. Patients with left ventricular outflow tract obstruction should avoid dehydration and excess alcohol consumption, and weight loss should be encouraged.

2. Non-vasodilating ß-blockers such as bisoprolol are recommended as first line therapy. If ineffective, additional therapy with disopyramide or alternatives such as verapamil or diltiazem should be considered after specialist evaluation.

3. Invasive treatment (surgery or alcohol septal ablation) to reduce left ventricular outflow tract obstruction should be considered in patients with a left ventricular outflow tract gradient ≥50 mmHg, moderate to severe symptoms (New York Heart Association (NYHA) functional class III-IV) and/or exertional or recurrent syncope resistant to maximum tolerated drug therapy.

Management of heart failure symptoms



ACEi = angiotensin converting enzyme inhibitor; AF = atrial fibrillation; LVEF = left ventricular ejection fraction; LVOTO = left ventricular outflow tract obstruction; MRA = mineralocorticoid receptor antagonist; NYHA = New York Heart Association.