Management of Atrial tachyarrhythmia

1. Rate control using β-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 intensive 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 quality) and 48 hour Holter recording in patients with HCM*.

4. Life long therapy with oral anticoagulants is recommended even when sinus rhythm is restored. Digoxin and Class IC anti-arrhythmics should be avoided. Amiodarone should be considered for rhythm control and to maintain sinus rhythm after cardioversion.

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.

Summary Card for General Practice

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SUMMARY CARD FOR GUIDELINES FOR THE DIAGNOSIS AND MANAGEMENT OF HYPERTROPHIC CARDIOMYOPATHY

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

HEADLINE:
GUIDELINES FOR THE DIAGNOSIS AND MANAGEMENT OF HYPERTROPHIC CARDIOMYOPATHY

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HCM = hypertrophic cardiomyopathy; LVOTO = left ventricular outflow tract obstruction; PDE = phosphodiesterase 5.
**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.

**Recommended tests in patients with definite or suspected HCM**

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 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.

**Schematic summarising the general approach to the diagnosis of hypertrophic cardiomyopathy**