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HCM

GUIDELINES FOR THE DIAGNOSIS AND MANAGEMENT OF HYPERTROPHIC CARDIOMYOPATHY

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ESC ESSENTIAL MESSAGES

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)

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ESSENTIAL MESSAGES FROM FROM THE 2014 ESC GUIDELINES ON DIAGNOSIS AND MANAGEMENT OF HYPERTROPHIC CARDIOMYOPATHY

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Take home messages

1. Definition

- Hypertrophic cardiomyopathy (HCM) is defined by the presence of increased left ventricular (LV) wall thickness that is not solely explained by abnormal loading conditions.
- In an adult, this represents a wall thickness ≥15 mm in one or more LV myocardial segments (or ≥13 mm in a first degree relative of someone with HCM) measured by any imaging technique.

2. Prevalence & Aetiology

- Hypertrophic cardiomyopathy (HCM) occurs in 0.02–0.23%of adults, with much lower rates in patients diagnosed under the age of 25 years.
- Hypertrophic cardiomyopathy can be caused by many genetic and non-genetic disorders.
- In up to 60% of patients with HCM, the disease is an autosomal dominant trait caused by mutations in cardiac sarcomere protein genes.
- 5-10% of adult cases are caused by other genetic disorders including inherited metabolic and neuromuscular diseases, chromosome abnormalities and genetic syndromes.

3. Genetic Counselling & Testing

- Genetic counselling is recommended in all patients with unequivocal HCM when it cannot be explained solely by a non-genetic cause.
- When a definite causative genetic mutation is identified in a patient, his or her first degree relatives should first be genetically tested. They should undergo clinical evaluation if they are found to carry the same mutation.

4. Left ventricular outflow tract obstruction

- Two-thirds of patients with HCM have dynamic obstruction of the left ventricular outflow tract (LVOTO) at rest or during exercise caused by contact between the mitral valve and the interventricular septum during systole.
- In patients with a resting LVOT gradient <50 mmHg bedside physiological provocation with Valsalva manoeuvre and standing should be routinely performed during echocardiography to determine if LV outflow obstruction can be provoked.
- Exercise stress echocardiography is recommended in symptomatic patients with an LVOT gradient <50 mmHg at rest or during physiological provocation.
- When a gradient is detected in the LV cavity, the presence of sub-aortic membranes, structural mitral valve leaflet abnormalities and mid-cavity obstruction should be systematically excluded.

5. Assessment of symptoms

- Most people with HCM are asymptomatic and have a normal life expectancy but some develop symptoms, often many years after the first manifestation of ECG or echocardiographic abnormalities.
- Systematic 2-D and Doppler echocardiography, exercise testing and ambulatory ECG monitoring are usually sufficient to determine the most likely cause of symptoms.

Take home messages

6. Prevention of Sudden Cardiac Death

- While the risk of sudden death is low for most patients with HCM, a small number are prone to life-threatening ventricular arrhythmias.
- The use of a new risk calculator (HCMRisk-SCD) (http://doc2do.com/hcm/webHCM.html) is recommended to guide the use of implantable cardioverter defibrillators (ICD).
- In all patients, clinical efficacy of ICD implantation should be balanced against its potential risk.

7. Symptomatic treatment

- In symptomatic patients with LVOTO, the aim is to reduce dyspnoea and chest pain by using drugs, surgical myectomy, alcohol ablation or pacing.
- Therapy in symptomatic patients without LVOTO focuses on management of arrhythmias, reduction of LV filling pressures, and treatment of angina.
- Patients with symptomatic progressive LV systolic or diastolic dysfunction refractory to medical therapy may be candidates for cardiac transplantation.

8. Atrial Arrhythmias

- Patients with HCM and paroxysmal, persistent or permanent AF should receive treatment with vitamin K antagonists.
- Lifelong therapy with oral anticoagulants is recommended, even when sinus rhythm is restored.
- Patients in sinus rhythm with LA diameter ≥45 mm should undergo 6–12 monthly 48-hour ambulatory ECG monitoring to detect AF.

9. Management of Pregnancy

- Most women with HCM tolerate pregnancy well but require expert advice and monitoring throughout pregnancy.
- All women with HCM should receive advice on contraception, sterilisation and termination when appropriate.

10. Multidisciplinary Care

- Clinicians should consider referral of patients to multidisciplinary teams with expertise in the diagnosis, genetics, risk stratification and management of myocardial disease.

Major gaps in evidence

1. Genotype-Phenotype Studies.
2. Frequency of screening in mutation carriers and the offspring of affected individuals.
3. Prevention of disease development in asymptomatic mutation carriers without a phenotype.
4. Randomized, controlled, clinical trials of drug therapies for symptom relief.
5. Prevention of left ventricular remodelling and the development of progressive heart failure.
6. Optimal management of asymptomatic left ventricular outflow tract obstruction.
7. Risk stratification and prevention of SCD in the young.

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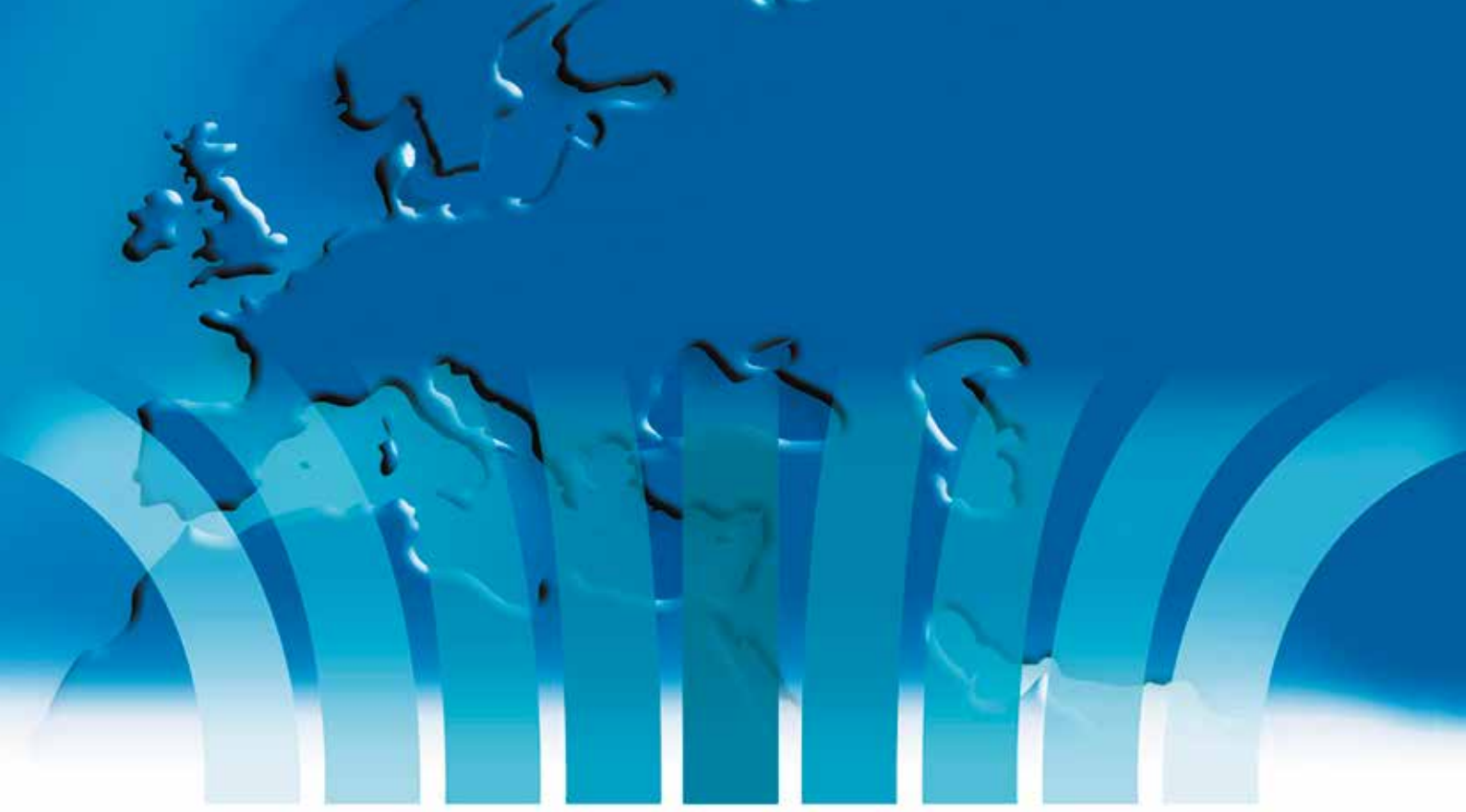

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