HYPERTROPHIC CARDIOMYOPATHY MANAGEMENT IN A TYPICAL UK SECONDARY CARE SETTING

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Introduction Hypertrophic cardiomyopathy (HCM) is a common inherited cardiomyopathy characterised by thickening of the ventricular wall. HCM has a variable spectrum of phenotypic expression, and a subset of patients are at risk of sudden cardiac death. Close follow-up and risk assessment of HCM is mandated; the European Society of Cardiology (ESC) has developed a risk-stratification calculator, as well as clear guidelines for management of HCM patients.

Methods We aimed to evaluate contemporary outpatient care of HCM patients in a typical UK secondary care setting. Using hospital diagnosis coding data, we retrieved medical records for 111 patients with a label of hypertrophic cardiomyopathy over the last 15 years. The medical records and cardiac investigations of these patients were then analysed in detail, in particular with reference to current ESC guidelines. Specifically, we examined: 1. Cause of HCM phenotype, 2. Family screening, 3. Risk-stratification on initial assessment, 4. Relevant echocardiographic measurements and reporting, 5. Follow-up schedules and subsequent risk stratification.

Results The average age was 66 years (59.3% males). Among the 111 patients, 47 (42.7%) were diagnosed with genetic/sarcomeric HCM, 40 (36.4%) were thought due to elevated afterload or infiltrative disease, and 24 (21.6%) were unclear. 49 cases (45.4%) were offered genetic testing and 37 of them (33.3%) underwent family screening, with only 8 (7.2%) having a family history of sudden cardiac death. The mean LA diameter recorded 4.42 (SD: 0.945), the mean maximum wall thickness was 2.08 (SD: 0.444), the mean LVOT max gradient was 34.98 (SD: 46.82). 44 (39.6%) patients underwent cardiac MRI scanning and 23 (20.7%) of the group had an ICD inserted during the follow-up period. Only 55 (49.5%) of patients were formally risk-stratified on initial evaluation. Echo reported a maximum left ventricular outflow tract (LVOT) gradient in 48 (43.2%) patients. 12 (10.8%) had an LVOT gradient of more than 50mmhg. Only 5 (4.5%) had a Valsalva manoeuvre documented, and 25 (22.5%) had a maximum wall thickness specifically quoted. During follow-up, 62 (55.9%) had an echocardiogram, and only 40 (36.0%) had Holter monitoring within 2 years, significantly limiting the ability to repeat risk stratification.

The average age and the youngest age of death were 76 years and 29 years respectively. However, 35 patients died during follow-up.

Conclusions In this study, we have demonstrated that the care of HCM patients in a typical UK secondary care setting is variable, with clear scope for improvement. In particular we identified deficiencies of risk stratification, reporting of key echo parameters and referral for family screening.

A dedicated clinic with pathways for new and follow-up patients would likely improve care for HCM patients. A ‘hub and spoke’ arrangement with inherited cardiac conditions specialists in tertiary care would be one potential approach.

Conflict of Interest

THE EFFECT OF SURGICAL SCAR ON SPIROMETRY AT CARDIOPULMONARY EXERCISE TESTING FOR ADULT CONGENITAL HEART DISEASE PATIENTS

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Introduction One of the commonest symptoms in Adult Congenital Heart Disease (ACHD) is breathlessness which is often