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Significance of etiological diagnosis of hypertrophic cardiomyopathy phenocopies

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Introduction: Clarification of the etiology of phenocopies of hypertrophic cardiomyopathy (HCM) is of great importance due to the growing numbers of widely available therapeutic options.

Purpose: The aim of our study was to determine the diseases responsible for HCM phenocopies in the patient group managed at our Heart Failure Outpatient Clinic.

Methods: We examined 58 heart failure patients with HCM phenotype (left ventricular wall thickness = 15mm) at our Heart Failure Outpatient Clinic between 1 January 2016 and 31 July 2018. After an echocardiography was performed, every patient underwent cardiac magnetic resonance imaging (CMR) test as well.

Results: The final diagnosis after the CMR test was HCM in half of the patient group (29 patients), hypertension-related left ventricular hypertrophy (LVH) in 18 patients, cardiac amyloidosis in 8 patients, and respectively 1-1 case of Löffler endocarditis, non-compaction cardiomyopathy and Fabry disease. From the 8 patients, that the CMR suggested cardiac amyloidosis (CA), 3 proved to be transthyretin (ATTR) amyloidosis, and 2 AL amyloidosis. In one case, after the negative medical examinations (serum electrophoresis, free light chain measurement, 99mTcHDP scintigraphy, gingival biopsy), a myocardial biopsy has been performed, confirming a previously suffered myocarditis. In the last 2 cases, due to one patient’s non-compliance, and the other patient’s death, no further testing could be carried out. Currently, 1 of the 3 ATTR patients is already receiving tafamidis treatment, in the other two cases, treatment initiation is in progress. The 2 AAL patients are being treated in co-operation with haematologists. The patient suffering from Fabry’s disease is receiving enzyme replacement therapy and the patient with Löffler endocarditis is treated with steroid therapy.

Conclusions: The etiological diagnosis of HCM phenocopies suspected by echocardiography is of great importance, because this could allow an effective and often prognosis improving treatment in a large number of patients.