Yuldasheva S.L., assistant
department faculty therapy
Andijan State Medical Institute
Uzbekistan, Andijan.

THE HYPERTROPHIC CARDIOMYOPATHY

Summary. The article is devoted to the most pressing and important problems of clinical diagnosis and covers the genetic role in the development of hypertrophic cardiomyopathy with the position of evidence-based cardiology. The article includes tactics for diagnosing hypertrophic cardiomyopathy. The diagnosis of the disease is based on the data of echocardiography, revealing structural changes in the heart muscle according to the type of hypertrophy, while the genesis of these changes remains unclear

Key words*:* Cardiomyopathy, diagnosis, genetics, heart.

Юлдашева С.Л., ассистент
кафедра факультет терапия
Андижанский государственный медицинский институт
Узбекистан, г. Андижан.

ГИПЕРТРОФИЧЕСКАЯ КАРДИОМИОПАТИЯ

Резюме. Статья посвящена наиболее актуальным и важным проблемам клинической диагностики и охватывает генетическую роль в развитие гипертрофической кардиомиопатии с позицией доказательной кардиологии. Статья включает тактику диагностики гипертрофической кардиомиопатии. В основу диагностики заболе- вания положены данные эхокардиографии,

выявляющие структурные изменения в сердечной мышце по типу гипертрофии, при этом генез этих изменений остается невыясненным.

Ключевые слова: Кардиомиопатия, диагностика, генетика, сердца.

Cardiomyopathy remains one of the least studied cardiologic diseases, being the object of an actively developing field of modern cardiology. Increased interest in the problem of studying myocardial diseases is due to the need for further study of the etiology and pathogenesis, the diversity and nonspecific city oftheir clinical manifestations, significant diagnostic and therapeutic difficulties. Theconstant increase in the frequency of occurrence of various forms ofcardiomyopathies is apparently connected both with a true increase in the numberof such patients and with the progress of modern diagnostic technologies. In addition, over the past decade, a fundamentally new concept is being formed on the definition of the concept of "cardiomyopathy" and their place in the structureof heart diseases, which is associated with the achievements of medical genetics, morphology, immunology and molecular endocrinology. Reflection of the modernevolution of knowledge is the constant revision, updating and refinement o f the corresponding concept and classification.

Cardiomyopathy is defined as a heterogeneous group of myocardial diseases associated with mechanical and / or electrical dysfunction, usually accompanied by myocardial hypertrophy or dilated cardiac chambers and developing due tovarious causes, but more often of a genetic nature. The pathological process can belimited to heart damage - primary cardiomyopathies (genetic, mixed and acquired)- or part of a generalized, systemic disease (secondary cardiomyopathies), often lead to the development of heart failure, its complications and deaths Hypertrophic cardiomyopathy (HCMC) is one of the main and most common forms of primary cardiomyopathies. The half-century history of studying the problem of HCMC reflects a significant evolution of knowledge in the issues of etiology, pathogenesis, prognosis, clinical course variants and treatment of the disease. In-depth study of various aspects of

pathology was closely linked and ensured the emergence of progressive instrumental, morphological and, finally, molecular genetic methods of diagnosis. The introduction of modern imaging techniques: echocardiography (ECHO-KG), dopplerography, single-photon emission computer (OECT) and positron emission tomography (PET), magnetic resonance imaging (MRI), ECG monitoring, etc., has determined progress in understanding mechanisms pathogenesis, diagnosis of the disease and the rationale Clinical definition of HCM The American College of Cardiology Foundation (ACCF) and American Heart Association (AHA) Task Force on Practice Guidelines provide a widely cited clinicopathologic definition of HCM. The guidelines define HCM as a cardiac disorder marked by unexplained left ventricular (LV) hypertrophy (thickening) associated with non-dilated ventricular chambers in the absence of other cardiac or systemic disorders in sufficient amounts capable of producing LV hypertrophy.

The definition also recognizes that genotype-positive patients could be phenotypically negative without overt hypertrophy [5]. The European Society of Cardiology (ESC) working group on myocardial and pericardial diseases also provides another widely cited definition of HCM. The ESC defines HCM as "*the presence of increased left ventricular (LV) wall thickness that is not solely explained by flow-limiting coronary artery disease (CAD) or abnormal loading conditions*" [6]. The definitions by ACCF/AHA and ESC establish the hallmark of HCM is LV hypertrophy (wall thickness) in the interventricular septum. Other characteristic pathological features are myocardial fiber hypertrophy, disorganized myocardial cells, interstitial fibrosis and thickened intra-myocardial coronary vessels [3].

The main method of diagnosis remains echocardiographic study (ECHOKG). Currently, a special place is occupied by modern imaging techniques, in particular magnetic resonance imaging (MRI), which has a higher resolving power and allows obtaining more accurate information about structural disorders in the heart.

HCM is a disease in which, in addition to shortness of breath, a frequent symptom is a variety of pains in the heart that are characterized by cardialgia and /or typical angina. The efforts of many scientists are aimed at the origin of this clinical phenomenon, on the detection of ischemia as a potential cause, as well as on the recognition of the genesis of myocardial blood supply deficiency in patients with HCM.

Among the possible causes of myocardial ischemia in patients with HCMC, the following can be identified: relative coronary insufficiency, caused by a decrease in the density of the capillary bed in hypertrophied myocardium; microvascular disease; disturbance of the left ventricular dysfunction, leading to a decrease in the effective pressure of coronary perfusion in the diastole and pathological changes in the microcirculation of the myocardium; a decrease in the coronary reserve; the presence of "muscle bridges" over the epicardial coronary arteries; compression of the septal branches of the coronary arteries; obstruction of LV LV; spasm of large coronary arteries; primary metabolic disorders of glucose, oxygen and fatty acids in cardiomyocytes; and also joined atherosclerosis of the coronary arteries, especially in older age groups.

In patients with HCMC, severe ischemia can occur until the development of myocardial infarction. Pathoanatomical studies confirmed the presence in patients with HCMC with intact coronary arteries of focal or diffuse changes of the myocardium from initial small-focal fibrosis to large areas of large-scleral transmural cardiosclerosis, which are the result of a long -term ischemic process. As the disease progresses, a number of patients are registered with a decrease in the pump function of the LV and / or PJ, associated with the de velopment of myocardial fibrosis. Fibrosis in this case can be the result of transformation of myocardial cells that have lost intercellular contacts, which is often observed in places of chaotic arrangement of muscle fibers, or consequence of ischemia and myocardial infarction against the background of microvascular pathology, and also joined coronary atherosclerosis. Large-focal fibrosis of the myocardium can lead to thinning of ventricular walls, reduction

of ventricular ventricular obstruction, disruption of systolic function, increase in volume of heart cavities, with dilatation less pronounced than with DCM.

The growth of the collagen network from the compensation factor at the initial stages of the disease is becoming an important factor in pathogenes is and gradually increasing chronic HF. In the process of remodeling, myocardial ischemia leads to damage and death of a part of cardiomyocytes, sclerosis of dead cells occurs, the location of the remaining cardiomyocytes changes, the geometry and sizes of the heart cavities change and, as a consequence, its function is disrupted. These structural and functional changes are the basis for the progression of the disease.

Atherosclerosis of the coronary arteries may be associated with HCM, especially in older age groups. Among patients with HCMC older than 45 years, atherosclerosis of the coronary arteries occurs in about 25% of cases, and in some cases is the cause of the manifestation of a disease that previously occurred latent. It should be noted that coronary atherosclerosis can be one of the causes of syncope and sudden death of patients with HCM of middle and old age. Given the impossibility of completely eliminating atherosclerotic changes in the coronary arteries when assessing complaints and conducting noninvasive studies, coronaroangiography is mandatory in patients with HCM with pain in the chest.

In HCM, the presence of significant LVEF obstruction, severity of myocardial hypertrophy, DD, and myocardial ischemia are the most important and closely related determinants of the severity of clinical manifestations, functional disorders and the "quality of life" of patients.

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