HYPERTROPHIC CARDIOMYOPATHY

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ABSTRACT

The article deals with information about hypertrophic cardiomyopathy (HCMP) – the primary damage of the heart characterized by thickening of walls of left ventricle and development of heart insufficiency especially diastolic. Hypertrophy of left ventricle wall more than 15 mm of unclear genesis is considered to be the diagnostic criteria. The data of investigation confirm the genetic character of HCMP. The conduction of proper diagnostics will give the possibility to find out subclinical changes of myocardium and optimize the treatment, improving further prognosis.

Keywords: *hypertrophy, cardiomyopathy (CMP), genetic character, diagnostics.*

АННОТАЦИЯ

В статье представлена информация о гипертрофической кардиомиопатии (ГКМП) - первичном поражении сердца, характеризующемся утолщением стенок левого желудочка и развитием сердечной недостаточности, преимущественно диастолической. Гипертрофия стенки левого желудочка более 15 мм неясного генеза считается диагностическим критерием ГКМП. Данные исследования подтверждают наследственный характер ГКМП. Есть основания считать, что патологические изменения встречаются чаще, чем диагностируются. Это можно объяснить тем, что у значительной части больных невозможно детальное обследование с применением лабораторных и инструментальных методов диагностики (вентрикулография, коронарография, ЭФИ), вследствие этого больным

ошибочно ставится диагноз ИБС, гипертоническая болезнь. Назначается неадекватное лечение, что ведет к прогрессированию заболевания. Проведение своевременной диагностики позволит выявлять ранние субклинические изменения миокарда и оптимизировать лечение, все это значительно снизит вероятность развития осложнений, в том числе фатальных, и улучшит дальнейший прогноз пациентов с гипертрофической кардиомиопатией.

Ключевые слова: *гипертрофия, кардиомиопатия (КМП), генетически обусловленная, диагностика.*

Hypertrophic cardiomyopathy (HCM) is a primary lesion of the heart characterized by caused by thickening of the walls of the left ventricle and the development of heart failure, predominantly diastolic. Hypertrophy of the left ventricular wall more than 15 mm is unclear genesis is considered a diagnostic criterion for HCM. There are the following options: symmetrical HCM (increase with the involvement of all walls of the left ventricle), asymmetric HCM (hypertrophy involving one of the walls), apical HCM (hypertrophy covers only the apex of the heart in isolation), obstructural HCM (ventricular septum or idiopathic subaortic stenosis), HCM of the free wall of the left ventricle. Important common features of HCM (both with and without obstruction) are high frequency of cardiac arrhythmias, primarily ventricular extrasystole and paroxysmal tachycardia, and impaired diastolic filling of the left ventricle, which can lead to heart failure. Arrhythmias are associated with sudden death occurring in 50% of patients with HCM. The etiology of the disease in many patients is hereditary. HCMP in some cases, it occurs as a result of a mutation of genes encoding contractile proteins of myocard. Patients with the so-called sporadic form have no relatives with this patology.

Pathogenesis: as a result of gene mutation, left ventricular hypertrophy occurs and lines of disorganization of cardiomyocytes.

The symptoms of the disease are diverse and unspecific, associated with hemodynamic disorders (diastolic dysfunction, dynamic obstruction of the outflow

tract, mitral regurgitation), myocardial ischemia, pathology of the autonomic regulation of blood circulation and violation of electrophysiological processes in the heart. The range of clinical manifestations is extremely large: from asymptomatic to steadily progressive forms that are difficult to treat with medication, accompanied by severe symptoms. Increased diastolic pressure in the left ventricle due to diastolic dysfunction, dynamic obstruction of the outflow tract of the left ventricle are manifested by shortness of breath at rest and during exercise, fatigue, weakness. An increase in pressure in the pulmonary circulation is accompanied by the development of acute left ventricular failure (more often at night - cardiac asthma, alveolar pulmonary edema). A drop in the ejection fraction during exercise or heart rhythm disturbances is accompanied by a deterioration in the blood circulation of the brain. Transient ischemia of the brain structures is manifested by a short-term loss of consciousness (fainting) or pre-syncope states (sudden weakness, dizziness, darkening of the eyes, noise and "congestion" in the ears).

Research methods:

ECHO-KG is the main method for diagnosing HCM. The criteria for diagnosing HCM based on ECHO-CG data are:

- Asymmetric ventricular septal hypertrophy (> 13 mm)
- Anterior systolic movement of the mitral valve
- Small cavity of the left ventricle
- Hypokinesia of the interventricular septum
- Midsystolic aortic valve occlusion
- Intraventricular pressure gradient at rest more than 30 mm Hg. Art.
- Intraventricular pressure gradient at a load of more than 50 mm Hg. Art.
- Normo- or hyperkinesia of the posterior wall of the left ventricle
- Reducing the tilt of the diastolic occlusion of the anterior leaflet of the mitral valve
- Mitral valve prolapse with mitral regurgitation

- The wall thickness of the left ventricle (in diastole) is more than 15 mm

If there is no left ventricular outflow tract obstruction at rest, it can be provoked by medication (amyl nitrite inhalation, isoprenaline, dobutamine) or functional tests (Valsalva test, exercise), which reduce preload or increase left ventricular contractility. Cardiac catheterization and coronary angiography are performed to assess the coronary bed before myectomy or mitral valve surgery, and to determine the cause of myocardial ischemia.

According to modern concepts, the treatment strategy is determined in the process of dividing patients into categories, depending on the course and prognosis described above. All individuals with HCM, including carriers of pathological mutations without phenotypic manifestations of the disease and patients with an asymptomatic course of the disease, need dynamic observation, during which the nature and severity of morphological and hemodynamic disorders are assessed. Of particular importance is the identification of factors that determine an unfavorable prognosis and an increased risk of sudden death (in particular, latent, prognostically significant arrhythmias).

General events

Limiting significant physical exertion and prohibiting sports that can cause aggravation of myocardial hypertrophy, an increase in the intraventricular pressure gradient and the risk of sudden death.

Medical treatment

The basis of drug therapy for HCM is drugs with a negative inotropic effect: β-blockers and calcium channel blockers. Disopyramide (a class IA antiarrhythmic drug) and amiodarone are also used to treat heart rhythm disturbances that are very common in this disease.

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