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HYPERTROPHIC CARDIOMYOPATHY: CAUSES, SYMPTOMS, AND DIAGNOSTICS

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ANNOTATION

Hypertrophiccardiomyopathy- The disease of the heart muscle, in which the massive thickening of one or more segments of the left (or less often - right) ventricle is observed. In patients with GKMP, the thickness of the interventricular partition or the free wall of the heart is 1.5 cm and more (normally no more than 1 cm).

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Hypertrophiccardiomyopathy- The disease of the heart muscle, in which the massive thickening of one or more segments of the left (or less often - right) ventricle is observed. In patients with GKMP, the thickness of the interventricular partition or the free wall of the heart is 1.5 cm and more (normally no more than 1 cm). One of the important conditions for the diagnosis of GPM is considered the absence of a patient a chronically increased blood pressure (i.e. arterial hypertension) or heart valves vices that could explain the expressed myocardial hypertrophy. In this disease, the thickening of the heart wall usually occurs on a limited area, and not evenly in all departments. Most often the interventricular partition is affected. Among the adult population, the GPM is detected at about 1 out of 500 people. Men are somewhat more likely to suffer from this disease compared to women (ratio of 2-3: 1).

Etiology

More than half (70%) of all cases of GKMP are family-friendly, with the main type of inheritance - autosomal dominant. The remaining cases of GKMP - sporadic, in which they cannot identify blood relatives with a pronounced thickening of the heart wall. The GPM is based on the generation of genes encoding the synthesis of the contractile proteins of myocardium. As a result, the arrangement of muscle fibers in the heart wall is disturbed, which leads to its sharp thickening. Mutations of the heavy chain of beta-myosin, myosin-binding protein with and heartтропонинаТ are considered the most frequent, meeting more thanHalf patients with GKMP. Occasionally, mutations simultaneously addly several genes encoding the synthesis of myocardial gearboxes; The forecast of such patients is considered the most

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unfavorable.

In 5-10% of persons with massive hypertrophy of the heart, hereditary metabolic and neuro-muscular diseases, chromosomal anomalies and genetic syndromes, which act as an independent reason for this hypertrophy may be revealed. Such cases are made to call GPM phenocopy, which can imitate changes in the heart and clinical picture of true GPM. This group includes Anderson Fabry , Danone disease, Leopard syndromes,Nuan , Kosmelo. With severe hypertrophy, myocardials may also flow and some non-mentioned diseases, such as amyloidosis of the heart.

What is the danger of GKMP? The aggressiveness of the CMP flow depends on the peculiarities of the genetic breakage underlying the disease. According to various estimates, 1-3% of adult patients and 4-6% of children and adolescents with GKMP die over the year. Most often, patients are dying suddenly due to the development of arrhythmias life-threatening (mainly ventricular tachycardia and ventricular fibrillation). It should be remembered that the sudden arrhythmic death among young patients may be the first and only manifestation of GKMP. An increasing heart failure, thromboembolism from the heart in the brain, infective endocarditis may be at the other reasons.

The factors increasing the risk of sudden death include the following:

- young or young age;
- Paroxysms of ventricular tachycardia according to the results of the 24-hour monitoringECG;
- Anamnesis instructions for registration of ventricular fibrillation, ventricular tachycardia or heart stop, implantation-defibrillator (ICD) due to the presence of ventricular tachycardia or ventricular fibrillation;
- instructions for sudden heart death in relatives, including implantation of the ICD due to ventricular violations of the heart rhythm;
- inexplicable episodes of loss of consciousness (fainting);
- drop in blood pressure in response to dosage physical exertion;
- Significant myocardial hypertrophy (the thickness of the interventricular partition and / or the back wall of the left ventricle is 30 mm or more)
- The presence of a pronounced obstruction of the left ventricular

Classification

Based on dataDoppler-Ehocardiography fundamentally allocate two forms of disease, depending on the presence or absence of a gradient of systolicPressure in the left ventricular cavity, respectively. This gradient is the pressure difference below and above the narrowing (up to the closure), which is reduced as a result of the reduction of the thickened part of the heart wall (more often - interventricular partition). Actually, obstruction (i.e., a sharp difficulty of blood current) occurs as a result of contact of the front sash of a mitral valve with a disproportionately

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thickened interventricular partition, while the pressure gradient in the left cavity increases sharply Stomach during systole.

In some patients with GKMM, there is no such gradient in peace, but when conducting certain provocative tests - appears. These tests include a sample Valsave (exhale with closed nose and mouth), the transition from the horizontal position of the body into the sedentary or vertical, dosed physical activity on the cyergometer or the treadmill, the use of some drugs. In such cases, it is customary to talk about GKMM with latent obstruction.

Symptoms

There are two main age periods when the clinical manifestations of the disease are needed for the first time - adolescent age and range from 30 to 40 years. Symptoms GKMP are diverse and . Actively for medical care is treated less than half of patients with GKMM. Their main complaints are shortness of breath during exercise (in 30-50% of cases), pain in the chest (in 40-60% of cases), heartbeat and interruptions in the heart of the heart (in 30-40% of cases), dizziness and States (fainting) (in 15-25% of cases). Medical errors in the first formulation of the diagnosis of GKMM are found in each second case. Often the disease is found randomly during dispensarization , medical and military or medical examination in the implementation of such studies as electrocardiography or echocardiography. GKMP can be diagnosed at any age independently of gender and races, but predominantly the disease is detected in people of young people who are in the physically active and creative period of life.

Several clinical current scenarios were described:

- stable benign oligosymtomatic during;
- progressive course (the appearance of shortness of shortness of shortness of breath, pain in the chest, dizziness, fainting);
- Sudden death;
- Atrial fibrillation attachment (flickering arrhythmia) with its complications, including thromboembolism from the heart in the brain;
- The development of the so-called. The ultimate stage of the disease with the development of symptoms of heart failure (shortness of shortness, edema) caused by the expansion of the left ventricle cavity and the decrease in its ability to be reduced normally.

Diagnosis

Inspection of the patient usually does not give any significant information, but some patients have an experienced doctor to listen to systolic noise to the left of the sternum, which is intensified when rising from the position of sitting in squatting, Taking Nitroglycerin.

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For GKMP, specific deviations of blood and urine indicators obtained during routine laboratory studies are not peculiar. At the same time, genetic violations confirmed in specialized laboratories are characterized.

Electrocardiography (ECG). Under GKMM, a variety of changes on the ECG, reflecting the heart rate disorders (atrial and ventricular extrasystolia, atrial fibrillation, ventricular tachycardia, Wolf-syndrome Parkinson- Like and others), signs of increasing the left atrium, thickening the wall of the left ventricle, signs that imitate disorders of the coronary blood supply or a myocardium infarction.

24-hourmonitoringECG Designed to diagnose transient heart rate and conductivity disorders. This method acquires a special value in patients with high risk of sudden death, primarily fainting or having cases of sudden death in blood relatives under the age of 40 years. It is also advisable to be carried out to control the effectiveness of antiarrhythmic therapy.

Doppler. The main ultrasound signs of GKMM are considered the following.

Asymmetric left ventricular hypertrophy.In adults, the GPPs are diagnosed with an increase in the thickness of the left ventricle wall ≥1.5 cm in one or more segments of this heart chamber. Typical myocardial hypertrophy, affecting only the interventricular partition.

Obstruction of the left ventricular ending path.To confirm it, a systolic gradient of pressure in the left ventricle cavity is determined using the Doppler scanning.DiagnosticallyThe gradient is more significant. art. And in such cases they say aboutobstructiveThe form of GKMP. If the gradient is less than 30 mm Hg. art. At rest in the position of Lözh, it is recommended to measure it against the background of provocative tests - sample, transition to the patient's position sitting and standing. Under the increase in the gradient to the values exceeding 30 mm Hg. art. Owing to provocative tests, state GKMP with latent obstruction of the left ventricular ending path.

The movement of the front sash of the mitral valve,Which also creates conditions for the narrowing of the left ventricle's ending path.

With ultrasound examination of the heart in patients with GPM, such non-specific changes are often identified as an increase in the size of the left atrium, a decrease in the left ventricular cavity, non-physiological increase in its contractility (emission fraction is more than 70 And even 80%), a violation of the ability to adequate to relax the left ventricle during diastole.

Magnetic resonance imaging (MRI) hearts. It is recommended to clarify the diagnosis of GKMP and the exception of other similar diseases in the following cases: when it is impossible to obtain an ultrasonic image of high quality; In patients with alleged isolated hypertrophy or aneurysm in the top of the left ventricle; If the amyloidosis of the heart or phenocopy of GKMP is suspected. MRI Allows you to

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effectively estimate the size of the cavities of the heart, the features of its structure and the state of systolic and diastolic functions of the ventricles.

Coronarografia. Invasive coronary angiography is recommended for adults with GKMP, survived a sudden stopping of the heart, as well as persons with pain in the chest, similar to angina. Coronary arteries in patients with GKMP are usually wide, well developed. During systole, a sharp narrowing of the coronary arteries may be observed.

Tests with exercise Treadmill- Test (running track) is recommended for refining the risk of sudden cardiac death, determining the functional ability of the patient and the effectiveness of the treatment.

Patients with GKMP, in which during echocardiography in the position of lying along or during provocative tests (sample Valsalva et al.) Systolic pressure gradient in the left ventricular path is more than 30 mm Hg. Art., But does not exceed 50 mm Hg. Art., It is recommended to conduct echocardiography under conditions of physical activity (stress-echocardiography) to clarify the degree of change in the obstruction of the left ventricle's ending path.

Myocardial biopsy. In the interests of the diagnosis of GPMM, this method can only be considered in some cases, for example, when there are good suspicions of other similar diseases flowing with the thickening of the heart walls (in particular, amyloidosis,lymphoma,sarcoidosis, myocarditis).

Genetic examination and family screening. The probability of detecting a mutation that led to the disease is highest in patients with a family form of the disease, and minimal - in senior patients and persons with non-classical manifestations. Medico-genetic consulting should be carried out by all sickness of GKMM, in which non-mental diseases are actually excluded (for example, hypertensive disease, amyloidosis, sports heart, heart valves vices). The identification of pathogenic mutation in the patient simplifies the diagnosis of GKMP in his relatives.

Literature

[1] European Resuscitation Council Guidelines for Resuscitation 2005 // resuscitation. - 2005.-67. - SUPL. 1.C. 1-86.

[2] Gurvich N.L. Basic principles defibrilation heart. - M.: Medicine, 1975. - 231 p.

[3]WiggersC.J., Bell J.R., Pain M. Studies of Ventricular Fibrillation CauseD by Electric Shock. Ii.Cinematographic and Electrocardiographic Observation of the Natural Process In the Dog's Heart.Its Inhibition by Potassium and The Revival of Coordinated Beats by Calcium // Am Heart J. -

[4] Emergency cardiology / Ed. - M.: Medical information-agency, 2004. - 520with.

ISSN 2181-3187

[5]SalamaG., KANAI A.,Efimov I. r. Subthreshold Stimulation of Purkinje Fibers Interrupts Ventricular Tachicardia In In Tact Hearts // Circulation Research. - Vol. 74. - N4. - P. 604-619.

[6]Treshkur TV, Parmon Ye.v.,Kapanadze idiopathic fibrillationinduced activity // Bulletin. - 2000. -№ 20. - P. 76-79.

[7] Ivanov G.G., Vostrikov V.A., Ovcharov R.S., Minin S.O. Spectral Analysis of the ECG atfast dying during the clinical death and early NE . therapy. - 1989. - № 2. - C. 42-45.

[8] Chen P-s., Wu T-s., Ting C-T.etal. A TALE OF TWO FIBRILLATIONS // CIRCULATION. - 2003. -108. - P. 2298.

[9] Weiss J.N., Q Z., Chen P-S.etal. The Dynamics of Cardiac Fibrillating // Circulation. -2005. - 112 (8). - P. 1232-1240.

[10] Nash M., Bradley Ch., Sutton P. et al. Whole Heart Action Potential Duration Restitution Properties in Cardiac Patients: A Combined Clinical and Modeling Study // Experimental Physiology. -2006. - 91.2 - P. 339-354.