

Diagnosis of the Origin of Cardiomyopathies and the Advantages of Modern Clinical Diagnostic Methods

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Abstract: Cardiomyopathy is a primary lesion of the heart muscle, not associated with inflammation, tumor, ischemic genesis, its typical manifestations are cardiomegaly, progressive heart failure and arrhythmia. There are dilated, hypertrophic, restrictive and arrhythmogenic cardiomyopathies. As part of the diagnosis of cardiomyopathy, ECG, echocardiography, chest radiography, MRI and cardiac MSC are performed. For cardiomyopathies, a gentle regimen and drug therapy are prescribed (diuretics, cardiac glycosides, antiarrhythmic drugs, anticoagulants and antiplatelet agents); According to indications, heart surgery is performed.

Key points: Dilated (congestive) cardiomyopathy, Hypertrophic cardiomyopathy, Restrictive cardiomyopathy, Arrhythmogenic right ventricular cardiomyopathy, Cardiomyopathies.

Introduction

The definition of "cardiomyopathy" is general for a group of idiopathic (of unknown origin) diseases of the myocardium, the development of which is based on dystrophic and sclerotic processes in the heart cells - cardiomyocytes. With cardiomyopathies, the function of the heart ventricles always suffers.

Coronary artery disease, hypertension, vasculitis, symptomatic arterial hypertension, diffuse connective tissue diseases, myocarditis, myocardial dystrophy, and other pathological conditions (toxic, drug, alcoholic effects) are considered as specific secondary cardiomyopathies, the main cause of which is myocardial damage secondary to the underlying cause.

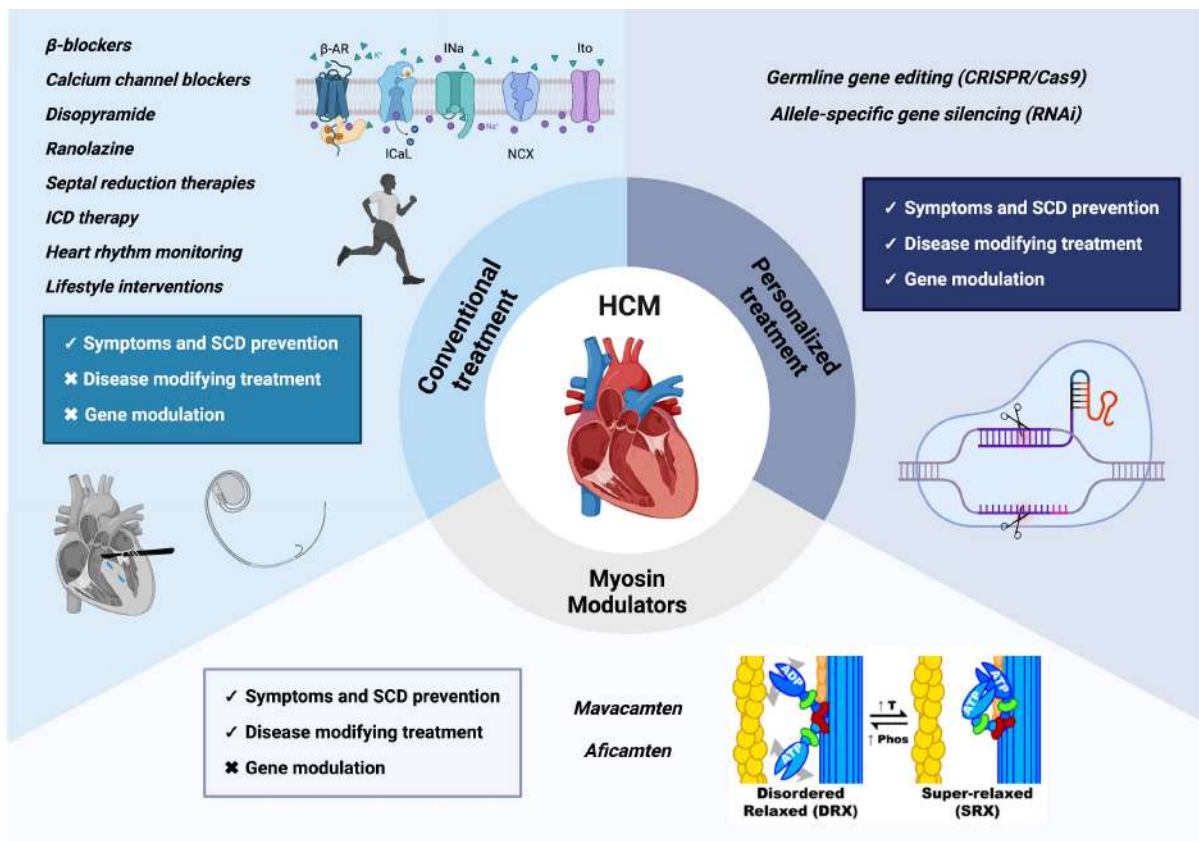
Cardiomyopathy is a group of heart diseases, often caused by genetic abnormalities and accompanied by mechanical and/or electrophysiological disturbances in its functioning.

There are several types of cardiomyopathy:

- dilatation;
- hypertrophic;
- limiting;
- arrhythmogenic;

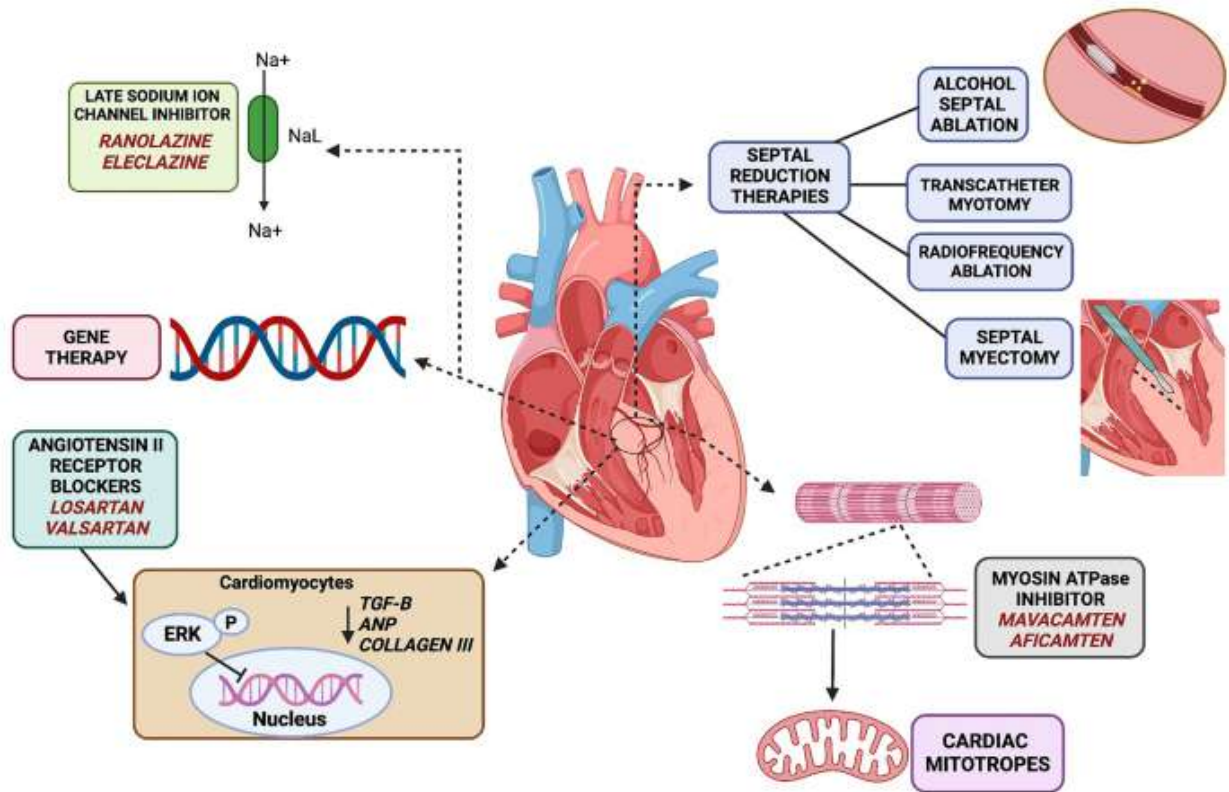
unclassified (not related to any of the above forms).

Their characteristics are reflected in their names. For example, dilatation is the expansion of the heart chambers, hypertrophic is characterized by thickening of the heart wall due to excessive development of the myocardium, restrictive is characterized by a decrease in the filling of the ventricles with blood during diastole.



Cardiomyopathies are differentiated according to:

- prevalence (expanded ones are the most common);
- gravity and flow rate. For example, untreated dilated cardiomyopathy can lead to death within a few years, while hypertrophic cardiomyopathy can lead to sudden cardiac death or may not affect life expectancy;
- symptoms - some do not manifest themselves for many years, while others are already at the beginning, accompanied by shortness of breath, swelling, chest pain, and a feeling of interruptions in the heart.
- But what all types of cardiomyopathies have in common is the progressive deterioration of the mechanical (contractile) function of the heart and the development of arrhythmias and conduction blocks, often life-threatening. As a result, serious complications develop:
- heart failure;
- sudden death;
- Pulmonary embolism resulting from atrial fibrillation.



Due to the poor prognosis of most cardiomyopathies, their development at a young age, and the lack of clear clinical manifestations at the onset of the disease, their diagnosis is of particular importance. Specialized cardiac centers in Germany perform the following:

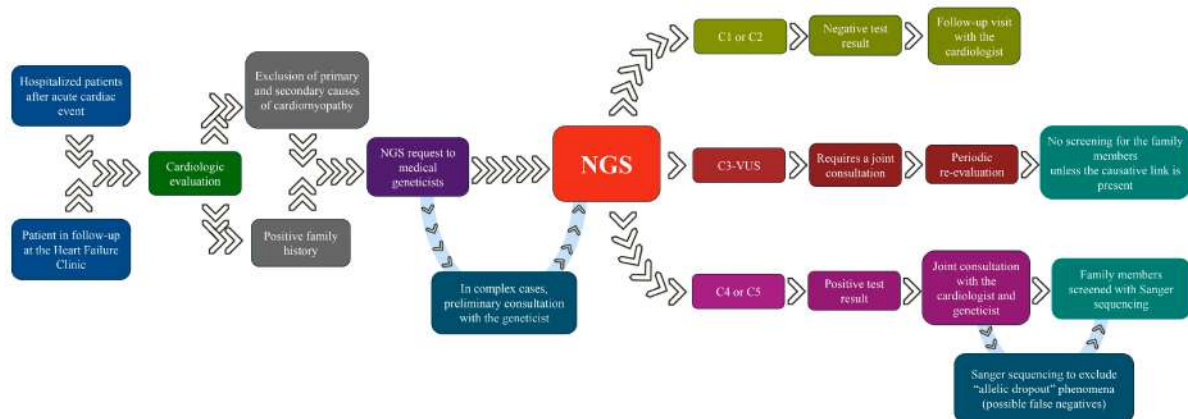
Genetic studies to confirm the hereditary nature of heart pathology;

X-ray examination of the chest organs to identify characteristic changes in the shape of the heart and changes in the lungs caused by hemodynamic disturbances;

ECG, including stress tests and Holter monitoring for differential diagnosis with ischemic heart disease, to identify and determine the origin and severity of heart failure, to determine the overload on various parts of the heart, to determine the fact of arrhythmia and to determine its nature;

Echocardiography, including echo-CG with stress tests and a transesophageal sensor to determine the size of the heart chambers, the characteristics of the movement of blood through them, the thickness of its walls, the condition of the valve apparatus, the presence of blood clots inside the heart;

Radioisotope ventriculography with the introduction of radioactive isotopes into the ventricles allows you to study not only their overall contractility, but also the activity of individual muscle groups;



cardiac catheterization - a minimally invasive X-ray surgical diagnostic intervention to measure intracardiac blood pressure, ultrasound scans inside the heart, and blood flow in the coronary arteries;

During its catheterization, a biopsy of the heart muscle is performed. Subsequent histological examination of the obtained sample reveals the characteristics of the myocardial tissue that are characteristic of different types of cardiomyopathy.

There is no etiologic, i.e., causal, treatment for cardiomyopathies. Therefore, it is very difficult and often requires a combination of conservative and surgical methods. Its goals are:

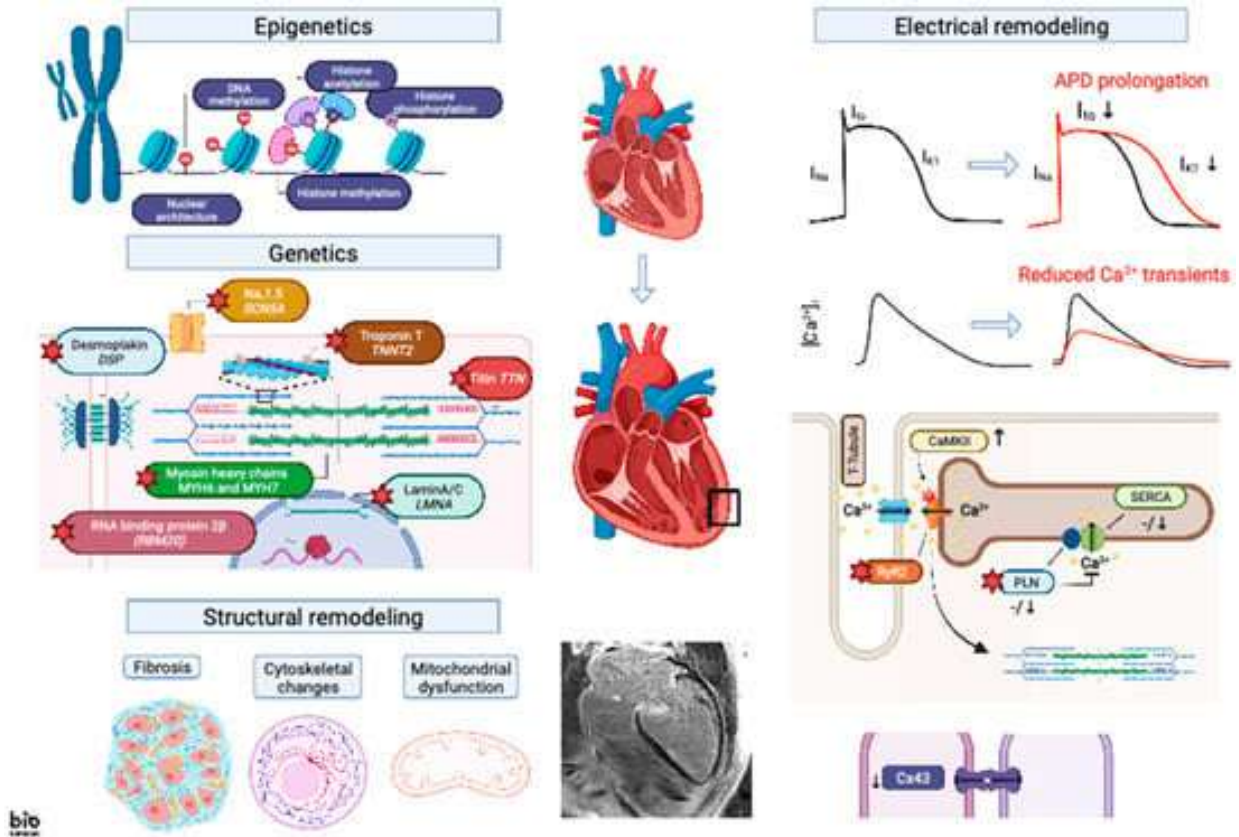
- a. reducing the degree of heart failure;
- b. restore the correct heart rhythm;
- c. prevention of sudden death, pulmonary embolism and other serious complications.
- d. Conservative therapy in German clinics is represented by several groups of drugs:
- e. ensuring normal blood pressure;
- f. has an antiarrhythmic effect;
- g. reduce the load on the myocardium;
- h. increase heart muscle contractility;
- i. improve blood supply to the heart;
- j. prevention of thrombosis;
- k. reducing congestion in the systemic and pulmonary circulation.

Surgical treatment of cardiomyopathies in Germany requires special qualifications of cardiac surgeons, specially equipped operating rooms for interventional and hybrid operations, as well as cardiac intensive care units capable of providing all types of monitoring (including intracardiac) and emergency care. Therefore, they are performed only in the world's leading cardiac surgery centers. These are full-fledged interventions, from X-ray endovascular myectomy to heart transplantation. Preference is given to minimally invasive interventions. For various types of cardiomyopathy, the following are performed:

Transaortic septal myectomy for hypertrophic cardiomyopathy using a minimally invasive endovascular method. Improves survival to almost 90% at 10 years. There is a positive experience of monitoring patients after similar operations for 40 years. Various options are used to excise thickening of the interventricular septum, including in complex cases of impaired patency of the outflow tracts of both ventricles;

transcatheter septal ablation - a catheter is inserted endovascularly through a peripheral artery. Upon reaching the artery supplying the hypertrophied interventricular septum under X-ray control, several milliliters of concentrated ethyl alcohol are injected through it. This causes limited necrosis in the planned area, eliminating the hypertrophied zone;

Implantation of pacemakers and cardioverter-defibrillators to maintain a normal heart rhythm, stop dangerous ventricular arrhythmias, and restore cardiac function in the event of sudden death due to ventricular fibrillation;



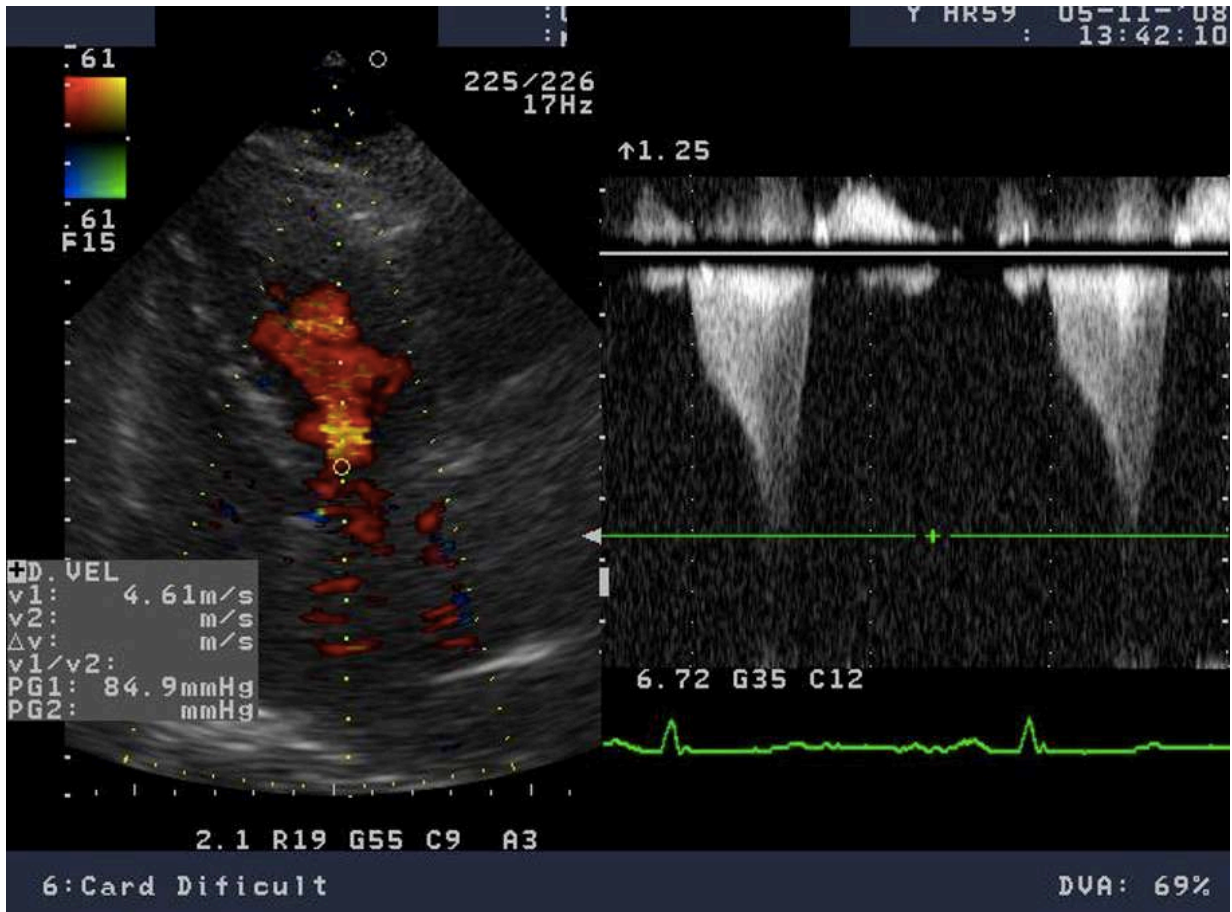
plastic surgery of heart valves and their transplantation for dilated and hypertrophic cardiomyopathy;

Artificial ventricle transplantation as a temporary measure to combat heart failure while preparing a patient for heart transplantation;

heart transplant;

The introduction of mesenchymal stem cells into the body, which develop into healthy cardiomyocytes. As a result, the progression of cardiomyopathy stops and the altered heart muscle tissue is replaced with normal tissue.

The prevalence of cardiomyopathies is a fraction of a percent, so the likelihood of their early diagnosis and adequate treatment by a general practitioner is very low. Currently, specialists from leading German cardiology and cardiac surgery centers have extensive experience in the conservative and surgical treatment of such patients. In addition, they are regularly given the opportunity to participate in clinical trials of promising experimental techniques. These centers closely cooperate with research institutes or are themselves engaged in the scientific development of new, more effective methods of combating cardiomyopathies and their complications.



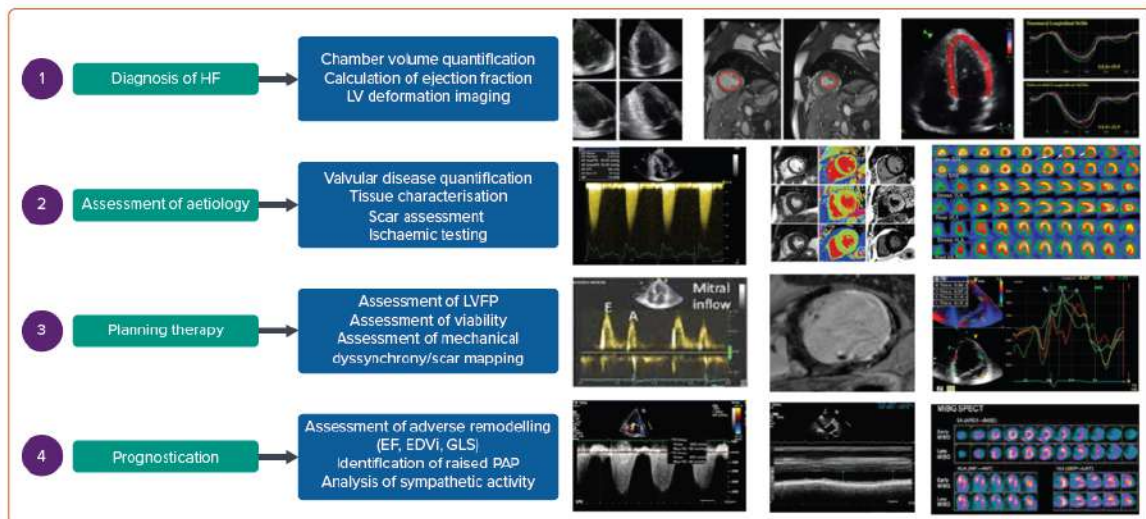
Reasons

- The etiology of primary cardiomyopathies is not fully understood to date. Among the possible causes of the development of cardiomyopathies are:
- Viral infections caused by Coxsackie viruses, herpes simplex, influenza, etc.;
- hereditary predisposition (a genetic defect that causes improper formation and functioning of muscle fibers in hypertrophic cardiomyopathy);
- anterior myocarditis;
- damage to cardiomyocytes by toxins and allergens;
- endocrine regulation disorders (damaging effects of somatotrophic hormone and catecholamines on cardiomyocytes);
- immune regulation disorders.

Classification

- Based on anatomical and functional changes in the myocardium, several types of cardiomyopathies are distinguished:
- dilatational (or stagnant);
- hypertrophic: asymmetric and symmetrical; obstructive and non-obstructive;
- limiting: destructive and dissipative;
- arrhythmogenic right ventricle;
- Takotsubo cardiomyopathy.
- Dilated (congestive) cardiomyopathy

Figure 3: The Role of Imaging in Heart Failure with Reduced Ejection Fraction



Symptoms

Dilated cardiomyopathy (DCM) is characterized by significant dilation of all chambers of the heart, hypertrophic phenomena and a decrease in the contractile ability of the myocardium. Symptoms of dilated cardiomyopathy appear already at a young age - 30-35 years. The etiology of DCM includes infectious and toxic influences, metabolic, hormonal and autoimmune diseases, probably in 10-20% of cases, cardiomyopathy is familial;

The severity of hemodynamic disorders in dilated cardiomyopathy is determined by the degree of reduction in myocardial contractility and pumping function. This leads to an increase in pressure first in the left ventricle, and then in the right ventricle. Clinically, dilated cardiomyopathy is manifested by symptoms of left ventricular failure (shortness of breath, cyanosis, cardiac asthma and pulmonary edema), right ventricular failure (acrocyanosis, pain and enlargement of the liver, ascites, edema, swelling of the jugular veins), heart pain that is not relieved by nitroglycerin, palpitations.

Diagnostics

Objectively, there is a deformation of the chest (heart tails); cardiomegaly with expansion of the borders to the left, right and up; muffled heart sounds at the apex, systolic murmur (with relative insufficiency of the mitral or tricuspid valve), gallop rhythm are heard. With dilated cardiomyopathy, hypotension and severe forms of arrhythmias are detected (paroxysmal tachycardia, extrasystole, atrial fibrillation, blockades).

Electrocardiographic examination mainly reveals left ventricular hypertrophy, cardiac conduction and rhythm disturbances. EchoCG shows diffuse myocardial damage, a sharp expansion of the heart chambers and its predominance over hypertrophy, intact heart valves and left ventricular diastolic dysfunction. Radiologically, with dilated cardiomyopathy, an expansion of the cardiac borders is detected.

Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy (HCM) is characterized by localized or diffuse thickening (hypertrophy) of the myocardium and narrowing of the ventricular chambers (mainly on the left). HCM is a hereditary pathology with autosomal dominant inheritance and most often develops in men of different ages.

Hypertrophic cardiomyopathy is characterized by symmetrical or asymmetrical hypertrophy of the ventricular muscle layer. Asymmetric hypertrophy is characterized by a predominant thickening of

the interventricular septum, while symmetric HCM is characterized by uniform thickening of the ventricular walls.

Depending on the presence of ventricular obstruction, two forms of hypertrophic cardiomyopathy are distinguished - obstructive and non-obstructive. With obstructive cardiomyopathy (subaortic stenosis), the outflow of blood from the left ventricular cavity is impaired, with non-obstructive HCM, there is no stenosis of the outflow tract;

Symptoms

The characteristic manifestations of hypertrophic cardiomyopathy are signs of aortic stenosis: cardialgia, dizziness, weakness, fainting, palpitations, shortness of breath, pallor. Later, congestive heart failure occurs.

Diagnostics

Percussion reveals an enlarged heart (more to the left), auscultation reveals muffled heart sounds, systolic murmurs in the III-IV intercostal spaces and at the apex, arrhythmias. A shift of the heart impulse down and to the left, a small and slow pulse in the periphery are detected. With hypertrophic cardiomyopathy, ECG changes are mainly manifested in myocardial hypertrophy in the left parts of the heart, inversion of the T wave, and the recording of a pathological Q wave.

Of the non-invasive diagnostic methods for HCM, the most informative is echocardiography, which reveals a decrease in the volume of the heart chambers, thickening and poor mobility of the interventricular septum (with obstructive cardiomyopathy), decreased myocardial contractility, and abnormal systolic prolapse of the mitral valve leaflet.

Restrictive cardiomyopathy

Restrictive cardiomyopathy (RCMP) is a rare myocardial lesion, usually characterized by endocardial damage (fibrosis), impaired diastolic relaxation of the ventricles, and impaired cardiac hemodynamics, with impaired myocardial contractility and the absence of pronounced hypertrophy.

In the development of RCM, a pronounced eosinophilia, which has a toxic effect on cardiomyocytes, plays a major role. With restrictive cardiomyopathy, endocardial thickening and infiltrative, necrotic, fibrous changes in the myocardium occur. The development of RCM goes through 3 stages:

- a. Stage I - necrotic - is characterized by pronounced eosinophilic infiltration of the myocardium and the development of coronary and myocarditis;
- b. Stage II - thrombotic - manifested by endocardial hypertrophy, parietal fibrinous deposits in the heart cavities, myocardial vascular thrombosis;
- c. Stage III - fibrotic - is characterized by widespread intramural fibrosis of the myocardium and nonspecific obliterating endarteritis of the coronary arteries.

Symptoms

Restrictive cardiomyopathy can be of two types: obliterating (with fibrosis and obliteration of the ventricular cavity) and diffuse (without obliteration). With restrictive cardiomyopathy, severe, rapidly developing signs of congestive circulatory failure are observed: severe shortness of breath, weakness with minor physical exertion, edema, ascites, hepatomegaly, swelling of the jugular veins.

Diagnostics

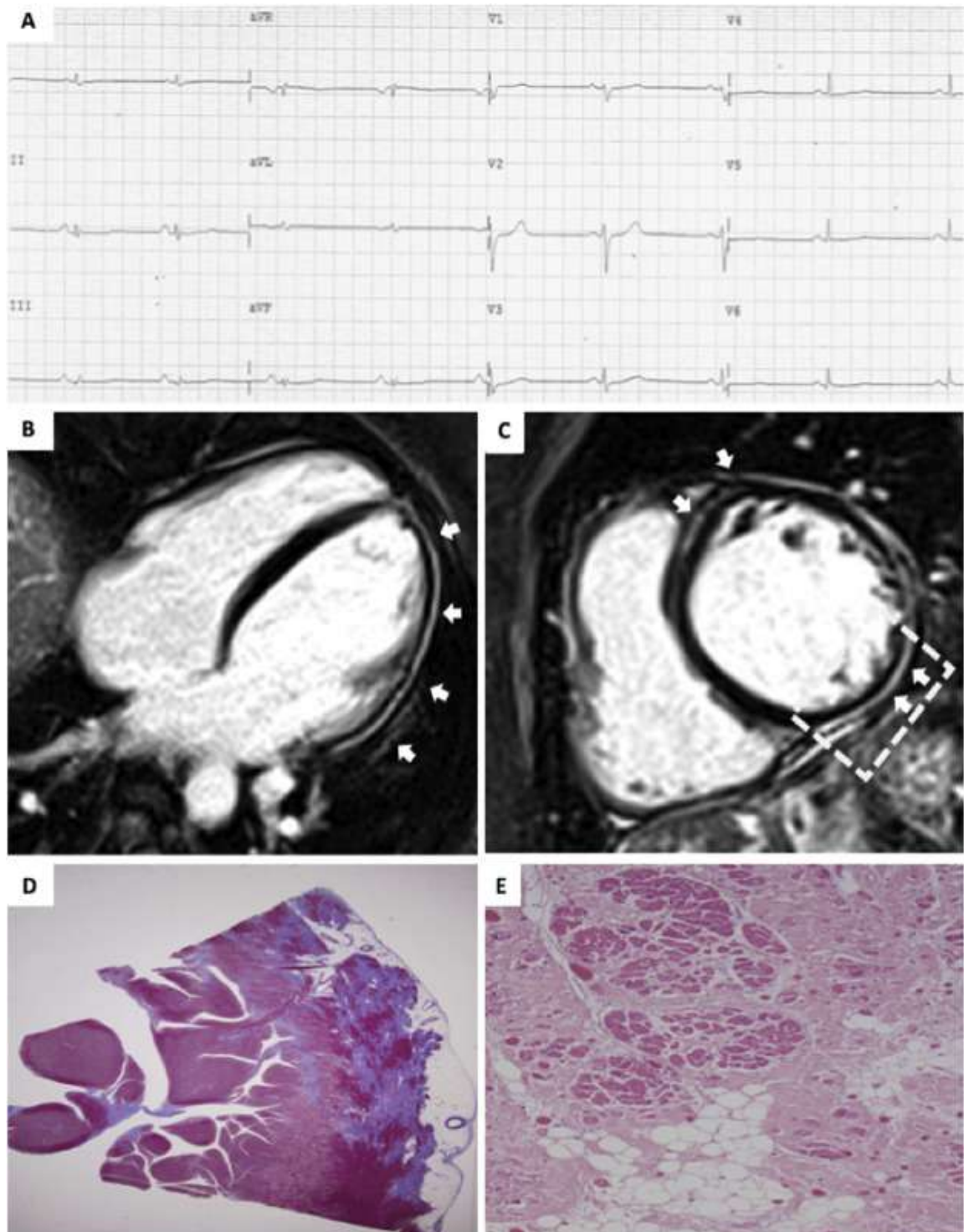
On auscultation, the heart is usually not enlarged; the ECG shows atrial fibrillation, ventricular arrhythmias, ST segment depression with T wave inversion, signs of pulmonary venous congestion, slightly enlarged or unchanged heart size. The echoscopic picture reflects insufficiency of the tricuspid and mitral valves, a decrease in the volume of the obliterated ventricular cavity, and impaired pumping and diastolic function of the heart. Eosinophilia is observed in the blood.

Arrhythmogenic right ventricular cardiomyopathy

The development of arrhythmogenic right ventricular cardiomyopathy (ARVC) is characterized by the gradual replacement of right ventricular cardiomyocytes with fibrous or fatty tissue, which is accompanied by various ventricular rhythm disturbances, including ventricular fibrillation. The disease is rare and poorly understood, with heredity, apoptosis, viral and chemical agents cited as possible etiological factors;

Symptoms

Arrhythmogenic cardiomyopathy can develop in adolescence or young adulthood and is manifested by palpitations, paroxysmal tachycardia, dizziness, or fainting. In the future, there is a risk of developing life-threatening types of arrhythmias: ventricular extrasystoles or tachycardia, episodes of ventricular fibrillation, atrial tachyarrhythmia, atrial fibrillation or flutter.



Diagnosics

In arrhythmogenic cardiomyopathy, the morphometric parameters of the heart do not change. Echocardiography shows moderate enlargement of the right ventricle, dyskinesia, and local protrusion of the apex or lower wall of the heart. MRI reveals structural changes in the myocardium: local thinning of the myocardial wall, aneurysms.

With all types of cardiomyopathies, heart failure develops, arterial and pulmonary thromboembolism develops, cardiac conduction disorders, severe arrhythmias (atrial fibrillation, ventricular extrasystole, paroxysmal tachycardia), and sudden cardiac death syndrome is possible.

Diagnosics

When diagnosing cardiomyopathies, the clinical picture of the disease and data obtained from additional instrumental methods are taken into account. The ECG usually shows signs of myocardial hypertrophy, various forms of rhythm and conduction disturbances, and changes in the ST segment of the ventricular complex. A chest X-ray can reveal pulmonary dilatation, myocardial hypertrophy, and congestion.

EchoCG data, which determine myocardial dysfunction and hypertrophy, its severity and the leading pathophysiological mechanism (diastolic or systolic failure), are especially informative for cardiomyopathies. According to indications, an invasive examination can be performed - ventriculography. Modern methods for visualizing all parts of the heart are cardiac MRI and MSCT. Examination of the heart chambers allows you to collect cardiobiopsy samples from the heart chambers for morphological examination.

Chest CT scan. Significant cardiac enlargement (mainly on the left side) in a patient with cardiomyopathy.

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Treatment of cardiomyopathies

There is no specific therapy for cardiomyopathies, therefore all therapeutic measures are aimed at preventing life-threatening complications. Treatment of cardiomyopathies in the stable stage is carried out on an outpatient basis, with the participation of a cardiologist; Periodic planned hospitalization in the cardiology department is indicated for patients with severe heart failure, in emergency situations - in cases of the development of paroxysms not associated with tachycardia, ventricular extrasystole, atrial fibrillation, thromboembolism, pulmonary edema.

Lifestyle changes are necessary for patients with cardiomyopathy:

- a. decreased physical activity
- b. following a diet with limited consumption of animal fats and salt
- c. exclusion of harmful environmental factors and habits.
- d. These measures significantly reduce the load on the heart muscle and slow the progression of heart failure.

It is recommended to prescribe drug therapy for cardiomyopathies:

- Diuretics to reduce pulmonary and systemic venous congestion
- cardiac glycosides for impaired myocardial contractility and pumping function
- antiarrhythmic drugs to correct heart rhythm
- anticoagulants and antiplatelet agents to prevent thromboembolic complications.

In extremely severe cases, surgical treatment of cardiomyopathies is performed: septal myotomy (resection of the hypertrophied area of the interventricular septum) with mitral valve replacement or heart transplantation.

Forecast

As for the prognosis, the course of cardiomyopathy is very unfavorable: heart failure is steadily developing, the likelihood of arrhythmic, thromboembolic complications and sudden death is high. Once dilated cardiomyopathy is diagnosed, the 5-year survival rate is 30%. With systemic treatment, the condition can be stabilized indefinitely. There have been cases of patients surviving for more than 10 years after heart transplantation.

Although surgical treatment of subaortic stenosis in hypertrophic cardiomyopathy undoubtedly gives a positive result, it is associated with a high risk of patient mortality during or shortly after the operation (every 6th operated person dies). Women with cardiomyopathy should avoid pregnancy due to the high risk of maternal mortality. Specific preventive measures for cardiomyopathies have not been developed.

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