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V. N. Karazin Kharkiv National University

CARDIOMIOPATHIES

Methodical recommendations
for the preparation of students of higher education in the 5th year of study
in the discipline «Internal Medicine. Module: Internal Medicine»

Electronic resource

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C 24 **Cardiomyopathies :** methodical recommendations for the preparation of students of higher education in the 5th year of study in the discipline «Internal Medicine. Module: Internal Medicine» [Electronic resource] / compilers O. V. Al-Trawneh, I. V. Shop, L. V. Bogun, N. V. Murovtseva, O. V. Kanishcheva, N. Ye. Tselik. – Kharkiv : V. N. Karazin KhNU, 2025. – (PDF 45 p.)

The methodological recommendations present the key aspects of the etiology, pathogenesis, classification, diagnosis, and treatment of cardiomyopathies.

For students of the 5th year to prepare for practical classes in the discipline «Internal Medicine. Module: Internal Medicine».

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CONTENT

LIST OF CONVENTIONAL ABBREVIATIONS

AAD – Antiarrhythmic drug

ACEI – Angiotensin-converting enzyme inhibitors

ACMP – Arrhythmogenic cardiomyopathy

AF – Atrial fibrillation

AFL – Atrial flutter

AH – Arterial hypertension

ALT – Alanine aminotransferase

ARB – Angiotensin II receptor blockers

AST – Aspartate aminotransferase

β-AB – Beta-blockers

BP – Blood pressure

CAD – Coronary artery disease

CBC – Complete blood count

CHF – Chronic heart failure

CMP – Cardiomyopathy

CV – Cardiovascular

CVS – Cardiovascular system

DCM – Dilated cardiomyopathy

EchoCG – Echocardiography

ECG – Electrocardiography

EF – Ejection fraction

FC – Functional class

HCM – Hypertrophic cardiomyopathy

HR – Heart rate

ICD – Implantable cardioverter-defibrillator

IHD – Ischemic heart disease

INR – International normalized ratio

IVS – Interventricular septum

LA – Left atrial

LBBB – Left bundle branch block

LVEF – Left ventricular ejection fraction

LV – Left ventricle

LVNC – Isolated noncompacted left ventricle

MRI – Magnetic resonance imaging

MV – Mitral valve

NCCM – Noncompaction cardiomyopathy

NYHA – New York Heart Association

RCMP – Restrictive cardiomyopathy

RBBB – Right bundle branch block

RV – Right ventricle

SCD – Sudden cardiac death

SGLT2 – Sodium-glucose cotransporter 2

TEE – Transesophageal echocardiography

WPW – Wolff-Parkinson-White

1. BASIC KNOWLEDGE, SKILLS, ATTAINMENTS NECESSARY FOR THE TOPIC STUDYING

Names of previous disciplines	Acquired skills
Foreign language	To be able to work with foreign literature to obtain data on modern methods of diagnosis and treatment of cardiological patients
Medical informatics	Apply modern computer programs and be able to work with them, have statistical methods for processing the results of clinical studies, analyze the results of studies, be able to evaluate and interpret the results of clinical research from information sources, have the ability to work with electronic databases
Human anatomy. Normal physiology. Histology, cytology and embryology	Know the normal structure, functions, and regulation of the cardiovascular system (CVS), and to understand and determine the relationship between its structure and function with other organs and systems of the human body.
Microbiology, virology and immunology	To be able to analyze the peculiarities of the microorganism's vital activity, to know the basics of antibiotic therapy, the conditions for the development of microorganism's resistance. To understand the pathogenesis of infectious diseases of the body and the cardiovascular system (CVS), and to assess the effectiveness and safety of antimicrobial agents.
Pathomorphology Pathophysiology	Know typical pathological processes in the human body: the mechanisms of development, changes in the human body, compensatory reactions, the impact of these changes on human organs and systems, the development of «cause-effect» relationships in the pathology of the whole organism. Describe and schematically represent the mechanism of development of typical pathological syndromes in cardiological diseases, justify pathogenetic approaches to drug therapy.

Pharmacology	Be able to navigate in the nomenclature of medicines. Know the mechanism of action of medicines, their pharmacodynamics, indications and contraindications for their use. Know the features of the clinical pharmacology of drugs used in the treatment of cardiological diseases, the features of the pharmacological action of these drugs in different categories of patients. Make a reasonable choice of individual drugs and therapy schemes in accordance with the principles of evidence-based medicine, optimization of treatment schemes, evaluate the effectiveness and safety of pharmacotherapy taking into account the individual characteristics of the patient, the presence of concomitant diseases.
Propaedeutics of internal medicine	Conduct a physical examination of patients according to the recommended scheme with a targeted examination of the CVD. To determine the leading syndromes and symptoms in cardiological patients. Analyze the results of basic laboratory and instrumental research methods. To be able to make a differential diagnosis based on the physical examination of cardiological patients and the data of additional research methods, justify and formulate the diagnosis.

1.1. The student should know:

- anatomy and functions of CVS;
- definition and classification of cardiomyopathies (CMP);
- definition, epidemiology and classification of hypertrophic cardiomyopathy (HCM), pathogenesis, and pathomorphology;
- clinical presentation of HCM, diagnosis, differential diagnosis, and treatment;
- definition, epidemiology, etiology, clinical presentation, diagnosis, differential diagnosis, and treatment of dilated cardiomyopathy (DCM);
- concept of restrictive cardiomyopathy (RCMP), course characteristics, diagnosis, and treatment;

- concept of arrhythmogenic right ventricular (RV) dysplasia, clinical presentation, modern diagnostic methods, and treatment;
- unclassified CMP.

1.2. The student must be able to:

- collect anamnesis rom patients with heart diseases;
- conduct a sequential examination to determine the nature of CVS damage;
- perform an objective patient examination and interpret the obtained data;
- develop an examination plan and analyze laboratory and instrumental findings in heart diseases;
- make a preliminary diagnosis and conduct differential diagnosis of CMP and other cardiovascular diseases;
- demonstrate proficiency in the principles of treatment, rehabilitation, and prevention of CMP patients in accordance with current Ministry of Health regulations;
- demonstrate adherence to the moral and deontological principles of the medical profession.

2. TOPIC CONTENT

2.1. Relevance of the topic

Cardiomyopathies (CMP) are a heterogeneous group of heart muscle diseases that significantly impact morbidity and mortality rates.

Currently, CMPs remain among the least studied cardiological pathologies. The growing interest in myocardial diseases is driven by the need for further research into their etiology and pathogenesis, the nonspecific and diverse clinical manifestations, as well as the significant diagnostic and therapeutic challenges they pose.

The increasing frequency of various CMP forms is attributed both to a rising number of affected patients and to advances in modern diagnostic technologies. Furthermore, over the past decade, a fundamentally new concept has emerged regarding the definition of «CMP» and their place within the structure of heart diseases. This shift is closely linked to advancements in medical genetics, morphology, immunology, and molecular endocrinology. The ongoing evolution of knowledge is reflected in the continuous revision, updating, and refinement of the terminology and classification of CMPs.

2.2. Definition, historical background and classification of cardiomyopathies

The term «cardiomyopathy» originates from Greek (kardia – heart, myos – muscle, pathos – disease) and translates to «heart muscle disease». It was first introduced by W. Bridgen in 1957 to describe myocardial diseases of unknown etiology, characterized by cardiomegaly (heart enlargement), electrocardiographic (ECG) abnormalities, progressive symptoms leading to heart failure (HF), and a poor prognosis.

According to contemporary concepts, CMP refers to a disease of the myocardium associated with structural and functional abnormalities in the heart

muscle without the presence of other cardiovascular (CV) diseases such as coronary artery disease (CAD), arterial hypertension (AH), valvular pathology, or congenital heart defects that would otherwise explain these dysfunctions.

CMPs arise from various causes, though they are most commonly linked to genetic mutations.

According to the latest guidelines of the European Society of Cardiology (ESC, 2024), CMP classification is based on specific phenotypes, distinguished by structural and functional characteristics:

– hypertrophic cardiomyopathy (HCM);

– dilated cardiomyopathy (DCM);

– nondilated cardiomyopathy of the left ventricle (LV);

– arrhythmogenic cardiomyopathy (ACMP) of the RV;

– restrictive cardiomyopathy (RCMP);

– other CMPs, including noncompacted myocardium and stress-induced CMP (Takotsubo Syndrome).

2.3. Hypertrophic cardiomyopathy (HCM)

Definition, Epidemiology, Classification, Pathogenesis

Hypertrophic cardiomyopathy (HCM) is a genetically determined disease with a predominantly autosomal dominant inheritance pattern. It is characterized by marked hypertrophy of the LV, sometimes affecting the RV, normal or reduced LV chamber size, significant diastolic dysfunction, and frequent arrhythmias.

HCM is a familial disease caused by mutations in multiple genes encoding sarcomeric proteins, including myosin heavy chain, actin, tropomyosin, and titin. These mutations disrupt the organization of myocardial fibers, leading to hypertrophy. Approximately 25 % of first-degree relatives of HCM patients exhibit signs of myocardial hypertrophy. HCM is the leading cause of sudden cardiac death (SCD) among children and adolescents during physical exertion. The incidence in the United States is estimated at 3–5 cases per 1 million pediatric

population (S. Miyake, 2011). Among adults, recent studies indicate a higher prevalence than previously thought, equal across racial groups, at 0.2 %. In Ukraine, the population prevalence is 0.47 %, with annual mortality rates ranging from 3.1 % to 8 % among HCM patients.

Major morphological types of HCM:

I – Predominant hypertrophy of the basal interventricular septum (IVS) (55–90 %).

II – Asymmetric hypertrophy of the IVS along its entire length.

III – Concentric (symmetric) LV hypertrophy (5–30 %).

IV – Apical hypertrophy (apical HCM) (~1 %).

Etiology

Advances in modern genetics have established that genetic factors – hereditary anomalies or spontaneous mutations – are the foundation of HCM development. These mutations occur in several gene loci that regulate the structure and function of myocardial contractile proteins, including β-myosin heavy chain, troponin T, troponin I, α-tropomyosin, and myosin-binding protein C. These genes are located on chromosomes 1, 2, 7, 11, 14, and 15. The defect is linked to alterations in the amino acid sequence of proteins. Most known mutations result in the synthesis of abnormal β-myosin heavy chain, and less frequently, troponin T or α-tropomyosin. These abnormal proteins disrupt sarcomere organization, leading to structural and functional impairment.

Genetic transmission and familial characteristics:

– approximately 50 % of HCM cases have a familial nature;

– inheritance follows an autosomal dominant pattern;

– first-degree relatives (parents, siblings) often exhibit echocardiographic (echoCG) signs of IVS hypertrophy.

In other cases, the disease is associated with spontaneous mutations, possibly influenced by negative environmental factors (sporadic forms of HCM).

A significant role is attributed to the influence of neuroendocrine factors, including:

– catecholamines;

– insulin;

– growth hormone;

– dysfunction of the thyroid and parathyroid glands.

Hypertrophy as a diagnostic criterion

Hypertrophy is considered clinically significant when **LV wall thickness ≥ 15 mm** (in some cases ≥ 13 mm in first-degree relatives of confirmed HCM patients).

Classification by degree of hypertrophy (LV wall thickness):

1. Mild – 13–15 mm.
2. Moderate – 16–19 mm.
3. Severe – ≥ 20 mm.

Wall thickness ≥ 30 mm is associated with a higher risk of SCD, particularly in young athletes.

Morphological classification of HCM based on localization:

1. Asymmetric septal hypertrophy (most common form).
2. Symmetric (concentric) hypertrophy.
3. Apical hypertrophy.
4. Mid-ventricular form.
5. Lateral or free LV wall hypertrophy.

Pathogenesis

The following changes are observed in HCM:

– pronounced LV hypertrophy with reduced chamber size;

– diastolic dysfunction, increasing left atrial (LA) pressure and causing pulmonary congestion;

– LA dilation due to increased LV filling pressure and average LA pressure;

– reduced cardiac output, despite relatively normal or elevated myocardial contractility and LV systolic function;

– relative coronary insufficiency;

– electrical heterogeneity and instability, contributing to arrhythmias and SCD;

– dynamic LV outflow tract (LVOT) obstruction (in obstructive HCM cases).

Characteristics of dynamic LVOT obstruction.

Unlike fixed obstruction, such as aortic valve stenosis, LVOT obstruction in HCM is dynamic. It primarily occurs in asymmetric IVS hypertrophy.

Mechanism of Obstruction:

– LVOT is formed by the proximal IVS and the anterior mitral leaflet;

– hypertrophied IVS narrows the LVOT

– during LV ejection, blood velocity increases, reducing lateral pressure (Bernoulli principle);

– a low-pressure zone pulls the anterior mitral leaflet toward the IVS (Venturi effect);

– the anterior mitral leaflet may temporarily come into contact with the IVS, leading to significant obstruction of blood ejection.

Consequences of LVOT obstruction: **development of a significant IV pressure gradient** (can reach 80–100 mmHg in severe cases).

Factors that exacerbate obstruction:

– Increased LV contractility (exercise, sympathetic nervous system activation, tachycardia, inotropic agents).

– Reduced preload (upright posture, Valsalva maneuver, hypovolemia, nitrate use).

– Decreased afterload (lower total peripheral vascular resistance and arterial pressure).

Dynamic nature of obstruction explains why the intraventricular pressure gradient can fluctuate even within a single day in the same patient.

Obstruction may develop:

– Not only in IVS hypertrophy but also in lesions affecting other regions of the LV.

– In rare cases, within the RV outflow tract.

Depending on the range of the IV pressure gradient, obstruction stages are classified according to the appropriate classification system.

Classification according to the New York Heart Association (NYHA Stages):

– Stage I – pressure gradient ≤ 25 mmHg; patients experience no symptoms during normal physical activity.

– Stage II – pressure gradient 26–35 mmHg; symptoms appear during the ordinary exertion.

– Stage III – pressure gradient 36–44 mmHg; heart failure (HF) symptoms at rest, angina.

– Stage IV – pressure gradient > 45 mmHg; severe manifestations of HF.

Clinical presentation, diagnosis and treatment

The clinical manifestations of HCM are polymorphic and nonspecific, ranging from asymptomatic forms to severe functional impairment and SCD.

Key symptoms of HCM:

– Progressive dyspnea and HF symptoms.

– Cardialgia (typical and atypical anginal pain due to LV microcirculation disturbances and myocardial ischemia; may occur at rest or during exertion).

– Syncope, presyncope, dizziness (causes include hypovolemia, complete heart block, sinus node dysfunction, sustained ventricular tachycardia).

– Palpitations (can indicate supraventricular or ventricular tachycardia, atrial fibrillation (AF); 48-hour ambulatory electrocardiography (ECG) recommended for frequent palpitations).

– Systolic murmur (detected on auscultation in obstructive HCM).

– HCM can combinate with AH.

The presence and sequence of symptom onset in HCM are determined by which ventricle is affected and the form of the disease. LV hypertrophy may remain unnoticed for years or even decades, with symptoms appearing only when LV function begins to deteriorate. Isolated RV involvement is extremely rare. In the non-obstructive form of HCM, where blood outflow from the ventricle is unobstructed, patients may remain asymptomatic, although occasional dyspnea during intense physical exertion or irregular heartbeats may occur. The presence of obstruction significantly reduces cardiac output during exercise, manifesting as progressive dyspnea, angina, and syncope, which often prompt patients to seek medical attention. Symptoms progress gradually, a characteristic feature of HCM. In patients with the obstructive form, auscultation reveals a systolic murmur at the apex or along the left sternal border. This murmur may arise from two mechanisms: relative mitral valve insufficiency, which produces a decreasing murmur, or turbulent flow in the outflow tract obstruction zone, where murmur intensity is directly proportional to obstruction severity, resulting in an increasing murmur.

Differential diagnosis of HCM should include conditions such as RCMP, aortic stenosis, Fabry disease, and glycogen storage disease type II.

Diagnostic studies recommended for HCM confirmation:

– Patient history and family history evaluation.

– Physical examination.

– Blood pressure (BP) measurement.

– Chest X-ray.

– Laboratory tests (complete blood count (CBC), urinalysis, alanine aminotransferase (ALT), aspartate aminotransferase (AST), bilirubin, creatinine, cholesterol, blood glucose, potassium, sodium).

– HCM Risk-SCD Score (Assessing 5-year risk of sudden death in patients ≥ 16 years: high, intermediate, or low risk).

– EchoCG: assessment of the presence and degree of hypertrophy, as well as the pressure gradient in the LV outflow tract using provocative tests that modify loading conditions and contractility (Valsalva maneuver in various positions: sitting, semi-recumbent, supine, and standing). The main echoCG feature of HCM is a zone of asymmetric myocardial thickening with reduced contractility in the affected area.

In obstructive HCM, the following echoCG findings are observed: asymmetric IVS thickening, systolic anterior motion of the mitral leaflets toward the IVS, high-velocity turbulent systolic flow in the LVOT obstruction zone, LA regurgitation due to relative mitral insufficiency, mid-systolic aortic valve opening due to reduced cardiac output.

– Transesophageal echocardiography (TEE): surgical strategy assessment for myectomy, pre-ablation mitral valve evaluation, monitoring septal alcohol ablation.

– Magnetic resonance imaging (MRI) (for differential diagnosis, as it provides superior detection compared to EchoCG for apical and anterolateral hypertrophy, aneurysms, thrombi, and papillary muscle anomalies).

– Standard ECG (used for the diagnosis and differential diagnosis of HCM versus post-infarction cardiosclerosis), potential ECG changes in HCM include: LV hypertrophy, ST-segment and T-wave abnormalities (high R-waves in V5–V6, deep S-waves in V1, T-wave inversion), pathological Q-wave appearance (III, aVF, V1–V6)—unknown cause, AF and atrial flutter (AFL), ventricular extrasystoles, shortened P–R (P–Q) interval.

In patients with HCM, a shortened P–R (P–Q) interval is sometimes observed. This may be due to several mechanisms:

1. Possible concomitant conduction system anomaly (e.g., accessory pathways). Some HCM patients exhibit Wolff-Parkinson-White (WPW)

syndrome or other variants of ventricular preexcitation. This represents a hereditary phenotype, sometimes associated with sarcomeric protein gene mutations causing HCM. In such cases: P–R interval < 120 ms, presence of a delta wave (in WPW).

2. Enhanced conduction through the atrioventricular (AV) node. In some HCM patients, increased sympathetic nervous system activity may accelerate AV node conduction. This can also shorten the P–R interval, especially without structural conduction system abnormalities.

3. Phenotypic heterogeneity of HCM

Certain HCM-causing mutations simultaneously affect the cardiac conduction system. This is particularly relevant in multisystemic genetic forms, such as HCM in Danon disease or PRKAG2 syndrome, where a high prevalence of accessory pathways and shortened P–R intervals is observed.

– In apical HCM, «giant» negative T waves (> 10 mm) frequently appear in leads V1–V6.

– 48-hour ambulatory ECG monitoring (for the detection of atrial and ventricular arrhythmias). Recommended: every 12–24 months for clinically stable patients and every 6–12 months for patients with sinus rhythm and left atrial size > 45 mm.

– Stress EchoCG test (for symptomatic patients if the provocation peak ΔP ≥ 50 mmHg in the LV outflow tract is not reached).

– Coronary angiography. Indications: angina (NYHA functional class (FC) III–IV), sustained ventricular tachycardia, and survivors of cardiac arrest.

– Genetic testing (for atypical clinical presentation or suspected other genetic disorders; first-degree relative screening).

All patients with confirmed or suspected HCM should be referred to a cardiologist at an appropriate medical institution.

Prognosis and risk factor assessment

Myocardial wall thickening typically does not continue to progress after the ages of 20–30, and only a small percentage (< 5 %) of patients develop LV dilation and systolic dysfunction.

– Approximately one-fifth of HCM patients are at risk of SCD, which is usually caused by malignant ventricular arrhythmias (ventricular tachycardia or fibrillation).

– In the vast majority of cases, the disease course is mild, and patients achieve a life expectancy comparable to healthy individuals of the same age.

– Clinical risk assessment should be conducted for all HCM patients, considering factors that increase SCD risk.

– Implantable cardioverter-defibrillator (ICD) therapy is highly effective in preventing cardiac death in HCM patients. ICD implantation indicates that the patient has been resuscitated from ventricular fibrillation or has a persistent ventricular tachyarrhythmia.

Risk factors for SCD that may indicate ICD need:

– family history of SCD;

– history of syncope;

– ventricular wall thickness > 3 cm;

– short episodes of ventricular tachycardia detected via Holter monitoring;

– blunted or hypotensive BP response to exercise testing;

– LV dilation and reduced ejection fraction (EF);

– AF;

– enlarged left atrium;

– elevated plasma levels of natriuretic peptide and/or troponin T (TnT);

– significant myocardial fibrosis on MRI (> 15 % of LV mass).

A risk calculator is available online to assist in assessing SCD risk and determining ICD necessity.

Treatment

General recommendations

1. Avoid intense physical exertion and competitive sports (an increased pressure gradient between the LV and aorta may trigger arrhythmias and syncope).

2. Prevent dehydration and excessive alcohol consumption; weight reduction is recommended.

3. Strenuous work conditions, such as night shifts or rotating schedules, are generally unsuitable.

4. Patients should be screened for arterial disease risk factors and receive appropriate treatment, as coexisting CAD significantly increases mortality.

Pharmacological Treatment

1. First-line drugs: β-blockers (β-AB) (e. g., propranolol, metoprolol at heart rate (HR) < 60 bpm) without vasodilatory effects, titrated to the maximum tolerable dose.

– Alternative to β-AB if contraindicated or poorly tolerated: verapamil (starting dose 40 mg 3× daily, increasing to 320–480 mg/day max).

– Adjunct to β-AB or verapamil: disopyramide (class IA antiarrhythmic) at a maximum tolerated dose (typically 400–600 mg/day), provided QT < 480 ms and no prostatitis or glaucoma. Disopyramide may also be considered as monotherapy.

– Diltiazem (starting at 60 mg 3× daily, up to 360 mg/day max) if β-AB or verapamil are contraindicated or intolerable.

– Oral or IV β-AB and vasoconstrictors (e. g., epinephrine) may be considered in severe LV obstruction with hypotension and pulmonary edema (vasodilators like nitrates and inotropes like dopamine are contraindicated).

Surgical Treatment

– Ventricular septal myectomy (Morrow procedure) (complications: AV block, defect of IVS, and aortic regurgitation). Concurrent mitral valve surgery is required in 11–20 % of cases.

– Septal alcohol ablation, inducing scar formation in the IVS (complication: AV block in 7–20 % of patients).

– Dual-chamber permanent pacing (for mild hypertrophy < 16 mm).

Anginal pain management

In HCM, anginal pain is not always due to coronary stenosis but often results from microvascular dysfunction and increased myocardial wall stress.

– β-AB (first-line therapy).

– Verapamil, diltiazem (alternative to β-AB if contraindicated or poorly tolerated).

– If no signs of obstruction, oral nitrates may be cautiously considered.

Management of HF II–IV NYHA without obstruction

– Preserved LV EF (EF >50 %): β-AB/verapamil or diltiazem, plus low-dose loop or thiazide diuretics (25–50 mg/day).

– Reduced LV EF (< 50 %): ACE inhibitors (ACE-I)/Angiotensin receptor blockers (ARB), β-AB, low-dose loop/thiazide diuretics, aldosterone receptor antagonists (eplerenone, spironolactone), and possibly digoxin (0.125–0.5 mg/day for rate control in AF), heart transplantation (for refractory HF III–IV NYHA and persistent life-threatening ventricular arrhythmias).

Management of arrhythmia (AF).

1. Electrical cardioversion, amiodarone, β-AB/verapamil, catheter ablation.

2. Oral anticoagulants (vitamin K antagonists—warfarin with a target level of international normalisation ratio (INR) of 2.0–3.0—or direct inhibitors).

2.4. Dilated cardiomyopathy (DCM)

DCM is a disease characterized by LV dilation with systolic dysfunction, occurring without hemodynamic overload (such as hypertension, valvular pathology) or CAD that could explain the impairment of LV systolic function. RV dilation and dysfunction may be present, but are not required for diagnosis.

Definition

DCM refers to primary myocardial impairment, marked by significant cavity dilation and ventricular contractile dysfunction. The term DCM applies only when chamber enlargement **is not due to**:

– coronary circulation disorders (ischemic heart disease – IHD);

– congenital heart defects;

– valvular heart diseases;

– systemic or pulmonary arterial hypertension;

– pericardial diseases.

Epidemiology

DCM is the most common CMP in children, observed worldwide and at all ages. The incidence of DCM in children is 0.57 cases per 100,000 per year, which is ten times lower than in adults. Most affected individuals are male, with a prevalence ranging from 62 % to 88 %.

Etiology

In most cases, DCM etiology remains unknown (idiopathic form). Current research focuses on several areas:

– *Genetic predisposition:* Approximately 30 % of cases are familial. Mitochondrial forms result from mitochondrial anomalies; X-linked forms arise due to mutations in genes encoding dystrophin and emerin, leading to muscular dystrophy, DCM, and joint contractures.

– *Role of chronic viral infection:* About 20 % of patients have a history of infectious myocarditis. Persistent viral agents (especially enteroviruses) damage myofibrils via viral RNA integration or autoimmune mechanisms.

– *Role of autoimmune processes:* Some patients exhibit high titers of autoantibodies against cardiac structures (myosin, tropomyosin, actin) and elevated cytokine levels.

– *Alcohol toxicity:* Alcohol-induced myocardial damage disrupts contractile protein synthesis, mitochondrial function, and energy metabolism, leading to chamber dilation.

Thus, the following factors play a significant role in the development of DCM: genetic predisposition, exogenous influences (viruses, alcohol), autoimmune disorders.

At early disease stages, the underlying cause may sometimes be identified, but in advanced stages, the clinical picture is largely independent of the initial trigger. So, the majority of DCM cases are classified as idiopathic dilated cardiomyopathy.

Pathogenesis

Several mechanisms contribute to the development of DCM:

– *Primary cardiomyocyte damage* due to reduced energy metabolism leads to fewer functional myofibrils, impairing contractile function.

– *Chamber dilation:* per the Frank-Starling mechanism, dilation initially maintains stroke volume and ejection fraction.

– *Compensatory tachycardia:* sympathetic activation helps preserve cardiac output but is metabolically inefficient.

– *Increased afterload:* per Laplace's law, maintaining pressure in a dilated ventricle requires greater wall tension.

Consequences of these processes:

– Ventricular myocardial hypertrophy.

– Increased oxygen consumption → myocardial ischemia.

– Progressive cardiomyocyte damage.

– Formation of diffuse or focal myocardial fibrosis.

– Declining pump function.

– Relative mitral and tricuspid insufficiency.

– Venous congestion in pulmonary and systemic circulation.

– Impaired peripheral organ perfusion.

– Despite myocardial hypertrophy, ventricular walls remain thin due to excessive dilation.

Significance of neuroendocrine changes:

– Activation of the sympathetic nervous system, renin-angiotensin-aldosterone system, and antidiuretic hormone.

– Sodium and water retention exacerbates congestion.

– Elevated catecholamines, angiotensin II, and cytokines further damage the myocardium and disrupt coagulation.

Clinical manifestations

DCM presents with three primary syndromes:

1. Chronic HF (CHF) (LV or biventricular) with congestive signs in pulmonary and systemic circulation.

2. Rhythm and conduction disturbances (ventricular arrhythmias, AF, AV blocks, bundle branch blocks (BBB)).

3. Thromboembolic complications (pulmonary embolism (PE), systemic arterial embolism, intracardiac thrombi).

Main mechanisms of thrombus formation in DCM:

1. Blood stagnation due to LV dilation:

– Enlarged chambers → slower blood flow, especially at LV apex, predisposing to apical thrombosis.

2. Reduced LV EF (EF < 35–40 %):

– Systolic dysfunction → blood pooling in chambers.

– Hypokinesis disrupts laminar flow, increasing thrombosis risk.

3. Endothelial dysfunction: chronic myocardial stretching damages the endocardium, triggering coagulation cascade activation.

4. AF: frequently associated with DCM → LA stasis and thrombus formation in the left atrial appendage.

5. Inflammatory/autoimmune mechanisms: in myocarditis-related or post-viral DCM, systemic inflammation induces prothrombotic states.

Thus, DCM symptoms and diagnostic findings are nonspecific, complicating differential diagnosis. The diagnosis of DCM is established by excluding other conditions causing ventricular systolic dysfunction (CAD, hypertension, myocarditis, pulmonary heart disease, etc.).

DCM can remain asymptomatic for a long time, despite echoCG evidence of ventricular dilation and dysfunction. Usually, initial clinical symptoms arise due to cardiac decompensation: pulmonary congestion, followed later by systemic congestion, and reduced cardiac output.

Main complaints of patients:

– dyspnea with exertion and at rest;

– worsened dyspnea in supine position (orthopnea);

– nocturnal dyspnea attacks (cardiac asthma) and pulmonary edema in severe cases;

– fatigue;

– muscle weakness;

– heavy legs during exertion.

Signs of right ventricle (RV) failure (occurring later in disease progression):

– lower limb edema;

– right upper quadrant heaviness;

– hepatomegaly;

– ascites;

– jugular vein distension (sign of increased central venous pressure, indicating RV failure).

Mechanisms in DCM:

1. Systemic circulation congestion:

– DCM → LV systolic dysfunction → ↑ pressure in LA → ↑ pressure in pulmonary veins → pulmonary hypertension → RV overload.

– RV gradually fails to manage pulmonary pressure → RV failure.

2. RV dilation:

– Late-stage DCM leads to remodeling of right heart chambers.

– RV stretches → reduced pump function → systemic venous congestion, including superior vena cava dilation.

3 Increased central venous pressure:

– Jugular vein distension reflects elevated pressure in the superior vena cava.

– Best observed in a supine position with the torso elevated 30–45°.

Clinical course of DCM

The most critical clinical feature of DCM is the rapid and relentless progression of HF symptoms, often refractory to standard CHF treatment. Some patients develop rhythm and conduction disorders, leading to palpitations and irregular heartbeats.

Severe complications include thrombosis and thromboembolism, occurring in ~20 % of patients. Thromboembolic events are most common in patients with AF.

Chest pain in DCM differs from classic angina attacks in most cases:

– has atypical localization;

– is usually unrelated to physical exertion.

Differential diagnosis

DCM should be differentiated from congenital heart defects, chronic myocarditis, arrhythmogenic myocardial dysfunction, arrhythmogenic RV dysplasia, rheumatic heart disease, exudative pericarditis, and specific CMPs.

Physical examination

During patient assessment, clinical features of CHF may be observed, including orthopnea, acrocyanosis, jugular vein distension, and lower limb

edema. Auscultation findings include congestive moist rales over the lower lung fields and displacement of the apical impulse (leftward and downward).

Cardiac auscultation:

– The I heart sound at the apex is diminished.

– With the development of pulmonary hypertension, an accentuated and splitting II heart sound is detected.

– A protodiastolic gallop rhythm (due to the presence of a pathological III heart sound) is often auscultated at the apex, associated with significant ventricular volume overload.

– Characteristic systolic murmurs at the apex and at the tricuspid valve auscultation point indicate the development of relative mitral and tricuspid insufficiency.

Diagnostics

ECG: Findings may remain normal or only show nonspecific repolarization disturbances. Conduction abnormalities are observed in almost 80 % of patients, including first-degree AV block, left BBB (LBBB), and nonspecific IV conduction disorders. Right BBB (RBBB) is less common. Conduction abnormalities are more frequent in patients with long-standing disease, reflecting progressive interstitial fibrosis and myocardial hypertrophy. Signs of LV hypertrophy are present. AF develops in approximately 20 % of patients with DCM.

24-hour ECG monitoring: Arrhythmias and latent myocardial ischemia.

EchoCG:

– dilation of all cardiac chambers;

– hypokinesis with areas of akinesis, marked reduction in LV EF;

– Regurgitation observed at the mitral and tricuspid valves;

– Presence of thrombi in cardiac chambers;

– Lack of ventricular wall thickening.

Chest X-ray (frontal and lateral views): cardiomegaly, increased vascular markings.

Cardiac catheterization and angiocardiography: measurement of pressure and oxygen content in cardiac chambers and pulmonary vessels. Angiocardiography allows imaging of not only central but also peripheral sections of major vessels.

Myocardial biopsy: A surgical technique enabling microscopic examination of myocardial tissue samples from different chambers, necessary for determining the nature of the disease and planning further treatment. Myocardial biopsy is often performed simultaneously with coronary angiography, helping to rule out IHD as a condition mimicking DCM. Both studies are conducted under X-ray guidance.

MRI: anatomical, geometrical, and biochemical characterization of the myocardium; useful when EchoCG cannot provide adequate visualization, and for assessing the relationships between the heart, vessels, and other organs.

Radionuclide imaging and positron emission tomography: assessment of myocardial viability, identification of areas of damage or insufficient perfusion.

Exercise testing: bicycle ergometry or treadmill stress test until stopping criteria are met. A six-minute walk test is also performed.

Laboratory diagnostics:

– CBC;

– urinalysis and urine output assessment;

– blood biochemistry;

– immunological and serological tests;

– coagulation profile;

– blood gas analysis and acid-base balance assessment;

– determination of natriuretic peptide levels;

– measurement of myocardial injury markers (troponin I or T).

Treatment

Therapy is based on the correction and prevention of the main clinical manifestations of the disease and its complications, including CHF, arrhythmias, and thromboembolism.

List and scope of essential medical services.

Patients should receive comprehensive therapy, including:

1. Etiotropic therapy for secondary CMPs:

– Surgical treatment methods (ischemic and endocrine CMPs).

– Etiological treatment for inflammatory CMP (chronic diffuse myocarditis).

2. Treatment of systolic CHF in idiopathic DCM and secondary CMPs, using:

– ACEI (recommended for all patients for an indefinite duration).

– β-AB (metoprolol, carvedilol, bisoprolol), indicated for all hemodynamically stable patients without contraindications.

– Sodium-glucose cotransporter 2 (SGLT2) inhibitors (gliflozins): dapagliflozin, empagliflozin.

– Saluretics, used when fluid retention is present.

– Cardiac glycosides, primarily digoxin, especially for tachycardic and normosystolic variants of AF.

– ARBs (for patient's intolerant to ACEI).

– Aldosterone antagonists (spironolactone, used as a temporary diuretic and to modulate neurohumoral mechanisms in CHF, improving survival prognosis at a dose of 25 mg/day).

Additional therapy.

1. Amiodarone for symptomatic or severe ventricular arrhythmias, and AF with rapid ventricular response cases resistant to other medications.

2. Intravenously sympathomimetic agents (dopamine and/or dobutamine)—used for CHF management in hospitalized patients, preferably with special infusion devices.

3. Nitrates, administered intravenously or sublingually (e.g., nitroglycerin or isosorbide dinitrate) for LV failure, discontinued after hemodynamic stabilization. Long-term use is appropriate for ischemic CMP or secondary CMPs with concurrent angina.

4. Anticoagulants (indirect) for patients with permanent AF, thromboembolic complications in anamnesis, or intracardiac thrombi. Mandatory INR monitoring. If INR measurement is unavailable, prothrombin index determination is required.

5. Implantable cardioverter-defibrillator (ICD) for recurrent ventricular fibrillation or persistent ventricular tachycardia resistant to antiarrhythmic drugs.

6. Implantation of a triple-chamber pacemaker (DDDR mode) in refractory CHF with significant IV conduction disturbances and desynchronization.

7. Heart transplantation.

Expected outcomes.

Absence of CHF progression, increased life expectancy.

Treatment duration.

Patients require daily medication for an indefinite period. Hospitalization duration depends on CHF severity and treatment efficacy.

Treatment quality criteria:

– reduction or elimination of CHF symptoms (dyspnea, palpitations, fatigue);

– increased LV EF;

– resolution of clinical signs of fluid retention;

– improved quality of life;

– prolonged intervals between hospitalizations.

Prognosis

DCM carries a poor prognosis, with up to 50 % of patients dying within 5 years, mostly due to sudden ventricular fibrillation.

Other causes of mortality include:

– progressive HF;

– thromboembolic complications.

Indicators of poor prognosis in DCM:

– high FC of CHF (NYHA class IV);

– end-diastolic dimension > 7.6 cm (by one-dimensional echoCG);

– LV EF < 35 %;

– Development of LBBB;

– Presence of high-grade ventricular arrhythmias on 24-hour ECG monitoring;

– Persistent AF.

2.5. Restrictive cardiomyopathy (RCMP)

Definition

According to the World Health Organization, restrictive RCMP is a myocardial disease characterized by ventricular wall rigidity, reduced ventricular filling, and decreased diastolic volume of one or both ventricles, while maintaining normal systolic function and unaltered wall thickness.

RCMPs form a heterogeneous group of primary (idiopathic) and secondary cardiac diseases, affecting the endocardium and/or myocardium. This pathology leads to:

– severe fixed restriction of ventricular filling in diastole;

– development of diastolic dysfunction of one or both ventricles;

– progressive diastolic CHF.

Classification

– Primary (idiopathic) RCMP—the exact etiology remains unknown.

– Secondary restrictive cardiac disorders—develop as a consequence of specific, known diseases.

Common features of RCMP:

– Severe restriction of ventricular filling during diastole, gradual reduction in ventricular chamber size up to obliteration.

– Diastolic filling occurs only at the start of diastole (during a short rapid filling phase), after which it almost ceases.

– Further ventricular expansion is impossible, leading to sharp increases in end-diastolic pressure in the ventricles.

– Elevated pressure in the atria and veins of both the pulmonary and systemic circulations.

Development of diastolic CHF – LV, RV or biventricular, depending on the predominant affected cardiac region.

Hemodynamic and morphological changes:

– Marked thickening, stiffening, and reduced elasticity of the endocardium and/or myocardium.

– Restricted ventricular filling, occurring only in the early diastolic phase.

– Decreased ventricular chamber size, with significant atrial enlargement due to volume overload, leading to mitral and tricuspid regurgitation.

– Congestion in the pulmonary and/or systemic venous circulation.

– Formation of intracardiac thrombi and increased risk of thromboembolic complications.

Treatment

Medical therapy is generally ineffective in most cases. For hypereosinophilic syndrome, prescribed treatments include: corticosteroids, immunosuppressants.

In cases of CHF, management involves: diuretics (hydrochlorothiazide, furosemide), vasodilators (nitrates), indirect anticoagulants.

However:

– The effectiveness of medical therapy is limited.

– Diuretics and nitrates may reduce venous return to the heart, further restricting ventricular filling and lowering cardiac output.

Cardiac glycosides are not recommended in RCM because:

– Myocardial contractility and ventricular systolic function remain normal in most cases.

– Diastolic filling impairment is the primary limiting factor, rather than systolic dysfunction.

Prognosis

Once CHF symptoms manifest, prognosis is poor – most patients die within 1.5–2 years. Prognosis worsens with the development of arrhythmias and thromboembolic complications.

In Löffler's endocarditis, surgical treatment may significantly improve outcomes.

2.6. Arrhythmogenic right ventricular cardiomyopathy (ACMP)

Definition

ACMP of RV is an inherited myocardial disease that predominantly affects the RV, leading to ventricular arrhythmias and an increased risk of SCD.

The exact prevalence is unknown, as ACMP RV is a relatively newly recognized condition, often asymptomatic in its early stages. Population estimates range between 6:10,000 and 44:10,000, varying across ethnic groups. 80% of ACMP RV cases are diagnosed before the age of 40, rarely in older individuals. Men are three times more likely to be affected than women.

The underlying cause of ACMP remains unclear, though Marcus F. I. (1982) first noted its hereditary nature.

Classification

According to microscopic studies, two histological variants of ACMP RV are distinguished:

– lipomatous variant (40%) – characterized by primary RV outflow tract dilation or total RV dilation;

– fibro-lipomatous variant (60%) – associated with RV aneurysms, particularly affecting the posterior leaflet of the tricuspid valve, the apex, and inflow tract.

In the first variant, a burdened family history, arrhythmias manifesting at an early age, and a malignant course of arrhythmias are observed. Histological examination of the RV myocardium reveals a predominance of adipocytes.

In the second variant, the disease onset occurs after the age of 25, with no evident hereditary burden in the family history.

ACMP RV – a progressive myocardial pathology with the following ***clinical-morphological variants:***

– «silent» myocardial anomaly—found in asymptomatic victims of SCD;

– «manifesting» disease—with segmental or global RV structural changes, often associated with ventricular arrhythmias;

– «end-stage» of biventricular involvement, representing DCM, leading to incurable progressive HF and requiring heart transplantation.

In 1994, McKenna W.J. et al. proposed diagnostic criteria for ACMP RV, adopted by the ESC. The diagnosis of ACMP is considered definitive when two major criteria, one major and two minor criteria, or four minor criteria are met. The McKenna W.J. et al. criteria are highly specific but have low sensitivity, particularly in the early stages of the disease. Currently, the medical community is actively discussing the criteria proposed by F. Marcus et al.

In the diagnosis of ACMP RV, key findings include results from ECG, radionuclide angiography, and MRI.

A standard 12-lead ECG holds significant diagnostic value: T-wave inversion in the right precordial leads is one of the most frequent ECG markers of ACMP, occurring in 70–75 % of patients who have experienced at least one episode of ventricular tachycardia. The «Epsilon» wave in lead V1, reflecting delayed RV depolarization and appearing as a notching on the ST segment (Figure

1), is an important ECG criterion. Additionally, an increase in QRS duration to 110 ms is regarded as a highly sensitive marker.

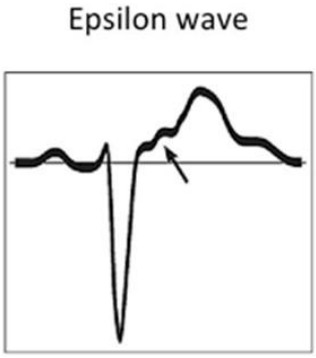

Fig. 1. Schematic representation of the «Epsilon» wave in ACMP

During ventriculography, dilation of the right ventricular outflow tract and aneurysmal bulging of the RV walls are detected, most commonly localized in the posterior subvalvular and anterior infundibular regions.

Diagnostic criteria for ACMP RV:

1. Structural/functional RV abnormalities (by EchoCG, MRI, Angiography)

Major:

– Akinesia/dyskinesia of the RV;

– Increased RV end-diastolic volume (MRI: ≥ 110 mL/m² in men; ≥ 100 mL/m² in women);

– Reduced RV EF < 40 %.

*Minor (l*ess pronounced structural changes):

– RV EF = 40–45 %.

2. Tissue changes (myocardial biopsy):

Major: – fibrofatty infiltration with loss of cardiomyocytes > 60 %.

Minor:– less pronounced fibrofatty infiltration with loss of cardiomyocytes 40–60 %.

3. Repolarization abnormalities (ECG):

Major: – T-wave inversion in V1–V3 in patients >14 years old (without RBBB).

Minor: – T-wave inversion only in V1 or V2.

4. Depolarization abnormalities (electrical criteria):

Major: – Epsilon wave in V1–V3 (small positive deflection after QRS).

Minor: – prolonged S-wave upstroke ≥ 55 ms in V1–V3;

– QRS duration > 110 ms in V1–V3 without RBBB.

5. Arrhythmias:

Major: – Ventricular tachycardia with LBBB morphology.

Minor: – frequent ventricular ectopic beats (>500/24 hours).

6. Family history/genetics:

Major: – confirmed ARVC in a first-degree relative;

– identified pathogenic genetic variant associated with ACMP RV.

Minor: – Suspected ACMP RV in a relative + SCD < 35 years old.

The ***diagnosis*** of ACMP is considered confirmed if:

– 2 major criteria from different categories, or

– 1 major + 2 minor criteria from different categories, or

– ≥4 minor criteria from different categories.

Key treatment objectives for ACMP RV:

1. reducing mortality;
2. preventing disease progression;
3. alleviating symptoms and improving quality of life through prevention/treatment of sinus tachycardia;
4. managing HF symptoms and increasing exercise tolerance.

Therapeutic approaches include lifestyle modifications, pharmacological treatment, catheter ablation, ICD placement, and heart transplantation**.**

Lifestyle modifications: recommended to limit physical exertion, with possible rehabilitative low-intensity physical activities.

In patients with ACMP RV, therapy includes the use of AADs, β-AB, and HF medications. The goal of AADs in the treatment of such patients is to improve their quality of life by preventing symptomatic ventricular arrhythmias. β-AB are

recommended for ACMP RV patients in cases of recurrent ventricular tachycardia, inappropriate ICD function, or false ICD activation caused by sinus/supraventricular tachycardia, AF/AFL with a rapid ventricular response.

Prognosis of life in ACMP patients depends on the extent of myocardial involvement and the development of life-threatening arrhythmias.

2.7. Other types of cardiomyopathies

Other forms of CMPs include:

– fibroelastosis;

– noncompaction CMP (NCCM);

– DCM with minimal dilation;

– mitochondrial CMP;

– Takotsubo CMP.

NCCM, according to the International Classification of Diseases, is designated as BA44.Z – other specified chronic IHD – and further specified as BA44.0 (or separately noted in clinical explanations depending on the local adaptation of the International Classification of Diseases-11 in Ukraine). This condition is genetically heterogeneous, with autosomal dominant and X-linked inheritance patterns identified in isolated forms of noncompaction LV. Isolated forms of noncompaction is a rare congenital variant of CMP. The disease originates during embryonic myocardial trabecular development, linked to disordered arrangement and compaction of the myocardial trabeculae. Histology does not reveal a specific pattern. Most pathologists report increased fibrosis in trabecular structures. The clinical presentation is similar to DCM, with a mortality rate of up to 80 % within 3–6 years. Patients frequently develop diastolic and systolic LV dysfunction, systemic embolization, ventricular and atrial tachycardia, and a predisposition to SCD.

ECG changes are non-specific and polymorphic. Reported cases include left-axis deviation, AV block (varying degrees), LBBB (21–44 %), AF (7–26 %), and other ventricular and supraventricular rhythm disturbances.

EchoCG is the most essential diagnostic tool for isolated forms of noncompaction LV. In short-axis echoCG, two myocardial layers are observed: a more compact subepicardial layer and a noncompact subendocardial layer due to trabeculation. An important diagnostic marker is the ratio of noncompact to compact myocardium at end-systole. For NCCM, the ratio is 2; for hypertensive LV hypertrophy, the ratio is 1.1; for DCM, the ratio is 0.8. Hypertrabeculated segments are typically hypokinetic.

No specific therapy exists.

The primary treatment strategies focus on heart failure management, prevention of embolic complications, and arrhythmia treatment.

Due to the poor prognosis and limited therapeutic efficacy, patients with NYHA Class III heart failure and reduced systolic function (EF < 40 %) are recommended for heart transplantation.

2.8. Examples of formulation of diagnoses

1. Hypertrophic Cardiomyopathy (HCM)

Diagnosis:

Hypertrophic cardiomyopathy, asymmetric hypertrophy of the interventricular septum, without left ventricular outflow tract obstruction, sinus rhythm, moderate risk of sudden cardiac death, CHF NYHA Class II.

2. Dilated Cardiomyopathy (DCM)

Diagnosis:

Dilated cardiomyopathy, reduced left ventricular ejection fraction to 28 %, CHF NYHA Class III, permanent atrial fibrillation.

3. Restrictive Cardiomyopathy (RCMP)

Diagnosis:

Restrictive cardiomyopathy, preserved left ventricular ejection fraction (EF 55 %), severe diastolic dysfunction, CHF NYHA Class II, sinus rhythm.

4. Arrhythmogenic Cardiomyopathy (ACMP)

Diagnosis:

Arrhythmogenic right ventricular cardiomyopathy, frequent ventricular extrasystoles, paroxysmal ventricular tachycardia, implanted cardioverter-defibrillator, CHF NYHA Class II.

TEST TASKS FOR SELF-CONTROL

1. Reduction of LV cavity is characteristic of:

– Ventricular septal defect.
– Hypertrophic cardiomyopathy.
– Aortic insufficiency.
– Dilated cardiomyopathy.
– Mitral insufficiency.

2. Diagnostic criteria for dilated cardiomyopathy include all except:

– Cardiomegaly – significant increase in cardiothoracic index (> 0.55).
– Reduced LVEF to 45 %.
– Fractional shortening < 25 % based on echocardiography.
– End-diastolic LV size exceeding 117 % of the expected value relative to body surface area.
– Pulmonary heart disease.

3. Left ventricular ejection fraction in dilated cardiomyopathy patients:

– Slightly increases.
– Slightly decreases.
– Significantly decreases.
– Significantly increases.
– Does not change.

4. The most characteristic feature of restrictive cardiomyopathy is:

– Ventricular cavity enlargement with systolic dysfunction.
– Pronounced concentric hypertrophy of the interventricular septum.
– Restricted diastolic filling with preserved systolic function.
– Reduced LVEF < 35 %.
– Aneurysmal enlargement of the right ventricle.

5. Medications not used in the treatment of dilated cardiomyopathy:

– Cardiac glycosides.
– β-AB.
– ACE inhibitors.
– Calcium channel blockers.
– Diuretics.

6. Restrictive cardiomyopathy is characterized by:

– Ventricular myocardial hypertrophy.
– Diastolic myocardial dysfunction.
– Reduced systolic ventricular function.
– Decreased systemic and pulmonary venous pressure.

7. A patient is diagnosed with hypertrophic cardiomyopathy. Which ECG finding is most typical for this condition?

– Prolonged QT interval.
– Epsilon wave in V1.
– Tall R waves in V5–V6, deep S waves in V1, T-wave inversion.
– Appearance of Q waves in II, III, aVF.
– Right bundle branch block.

8. The main drugs used in the treatment of hypertrophic cardiomyopathy are:

– Cardiac glycosides.
– Beta-blockers and calcium channel blockers.
– Peripheral vasodilators.
– Cardiac glycosides and nitrates.
– Cardiac glycosides and diuretics.

9. The most characteristic ECG changes in hypertrophic cardiomyopathy:

– Pathological Q waves in V4–V6.
– Deep negative T waves (> 10 mm) in chest leads (usually V4–V6)
– Increased QRS amplitude, often in V5–V6 with tall R waves (possible amplitude up to 30 mm).
– Arrhythmia and conduction disturbances.
– All of the above.

10. Medications not used in the treatment of dilated cardiomyopathy:

– Cardiac glycosides.
– Hypertrophic cardiomyopathy.
– ACE inhibitors.
– Calcium channel blockers.
– Diuretics.

Answer key:

1 – B, 2 – E, 3 – C, 4 – C, 5 – D, 6 – B, 7 – C, 8 – B, 9 – E, 10 – D.

SITUATION TASKS

1. A 36-year-old patient complains of dyspnea, right upper quadrant pain, dry cough, and leg edema. He has been ill for 4 months, previously treated for rheumatism without success. Diffuse cyanosis, leg edema, RR – 28/min, temperature – 36.8 ºC. Fine crackles in the lower lung regions. Heart borders enlarged in all directions, weakened heart sounds, systolic murmur at the 5th point. Ps – 90/min, BP – 100/80 mmHg. Liver palpable 4 cm below the costal margin. Which examination will be the most informative for diagnosis?

A. ECG.
B. EchoСП.
C. MRI.
D. Exercise stress test.
E. Scintigraphy.

2. A 34-year-old patient reports resting dyspnea, nighttime dyspnea, palpitations, and persistent cough for four months. No prior health conditions. Orthopnea posture, acrocyanosis, leg edema. Diminished vesicular breath sounds, fine wet crackles over the lungs. Heart borders enlarged bilaterally, weakened heart sounds, gallop rhythm, hepatomegaly. Chest X-ray shows a globular heart shape. What is the most likely diagnosis?

A. Dilated cardiomyopathy
B. Exudative pericarditis
C. Hypertensive disease
D. Hypertrophic cardiomyopathy
E. Restrictive cardiomyopathy

3. A 21-year-old male complains of episodes of syncope. In the past few months, he has experienced fainting during football, basketball, and sprinting. His family history includes multiple cases of sudden cardiac death, including his father at 52 years old. Echocardiography shows severe hypertrophy and abnormal systolic movement of the mitral valve leaflet. What is your preliminary diagnosis?

A. Hypertrophic cardiomyopathy
B. Dilated cardiomyopathy
C. Arrhythmogenic right ventricular cardiomyopathy
D. Restrictive cardiomyopathy

4. A 42-year-old male complains of dyspnea, right upper quadrant heaviness, and leg edema. Symptoms have been present for two weeks. Afebrile (36.2°°C), globally enlarged cardiac borders, severely weakened heart sounds. Echocardiography reveals an enlarged left ventricular cavity and reduced ejection fraction (EF 30 %). What is the preliminary diagnosis?

A. Dilated cardiomyopathy

B. Rheumatism
C. Ischemic heart disease
D. Hypertrophic cardiomyopathy
E. Myocarditis

5. A 39-year-old male complains of palpitations, dyspnea, irregular heart rhythm, dizziness, and rapid fatigue. Examination reveals pronounced cardiomegaly, muffled heart sounds, systolic murmur at the apex, signs of cardiac decompensation (IIA stage). ECG shows atrial fibrillation, left bundle branch block, and inverted T waves in multiple leads. Echocardiography reveals diffuse dilation of cardiac chambers with reduced myocardial contractility. Likely diagnosis?

A. Constrictive cardiomyopathy
B. Hypertrophic cardiomyopathy
C. Exudative pericarditis
D. Myocarditis
E. Dilated cardiomyopathy

ANSWER KEY: 1 – B, 2 – A, 3 – A, 4 – A, 5 – E.

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Електронне навчальне видання комбінованого використання
Можна використовувати в локальному та мережному режимі

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КАРДІОМІОПАТІЇ

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з дисципліни «Внутрішня медицина. Модуль внутрішня медицина»

(Англ. мовою)

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