Cardiomyopathies are myocardial diseases in which there is structural and functional disorder of the heart muscle, in the absence of coronary artery disease, hypertension, valvular disease and congenital heart disease. Cardiomyopathies are grouped into specific morphological and functional phenotypes: dilated cardiomyopathy, hypertrophic cardiomyopathy, restrictive cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy and unclassified cardiomyopathies. Each phenotype is subclassified into familial and non familial forms, where familial refers to the occurrence, in more then one family member, of either the same disorder or a phenotype that is caused by the same genetic mutation.1, 2, 3

Most familial cardiomyopathies are monogenetic disorders. Monogenetic disorders can be sporadic, when the causative mutation is de novo, and those patients are assigned to the familial category as their disorder can be transmitted to their offspring. Non familial cardiomyopathies are clinically defined by the presence of the cardiomyopathy in the patient in the absence of disease in other family members. They are subdivided in idiopathic (no identifiable cause) and acquired cardiomyopathies in which ventricular dysfunction is complication of the disorder rather then feature of the disease.1

DILATED CARDIOMYOPATHY

Dilated cardiomyopathy is characterized by dilatation and systolic dysfunction of left ventricle in the absence of hemodynamic disorder (hypertension, valvular disease) or coronary artery disease. Right ventricular dilatation may be present but is not necessary. The prevalence of dilated cardiomyopathy in the general population is not clearly specified because it varies with age and geography. At least 25% of patients in western populations have familial form with autosomal dominant inheritance.1

Dilated cardiomyopathy can occur at a late stage after cardiac infection or inflammation and is characterized by the presence of chronic inflammatory cells. Therefore, in its diagnosis, in addition to echocardiographic findings of left ventricular dilatation and reduced systolic function, histological and immunocytochemical methods need to be done.
Special form of dilated cardiomyopathy is peripartum cardiomyopathy that presents with heart failure due to systolic left ventricular dysfunction during the last month of pregnancy or within first 5 months after delivery. Etiology of this cardiomyopathy is not cleared but it is assumed that abnormal adaptation to pregnancy induced hemodynamic disorders, inflammatory and autoimmune processes are possible reasons for its occurrence.¹

Dilated cardiomyopathy is characterized by, usually gradual, development of symptoms of left and right ventricular failure. In addition to dyspnea and fatigue, vague chest pain may be present, syncope due to arrhythmias and systemic embolism originating from ventricular thrombus. During preoperative evaluation of patients with dilated cardiomyopathy, physical examination may reveal distended neck veins, crackles in the lungs, the third and fourth heart sounds, murmurs of mitral and tricuspid regurgitation, hepatomegaly, ascites, peripheral edema, and in severe heart failure, weight loss and cachexia. Chest radiography shows heart silhouette enlargement due to left ventricular dilatation and signs of pulmonary congestion or pleural effusion. ECG shows sinus tachycardia or atrial fibrillation, ventricular arrhythmias, low voltage, diffuse nonspecific ST segment and T wave changes, as well as atrioventricular and intraventricular conduction disturbances. Echocardiography shows left ventricular dilatation, with normal, thickened or thinned walls and systolic dysfunction. Brain natriuretic peptide level in blood is elevated. Coronary angiography is often performed to rule out ischemic heart disease.⁴

In preoperative preparation continuation of standard therapy for heart failure is indicated. In patients with symptomatic heart failure and left ventricular ejection fraction less then 40% ACE inhibitors, ß-blockers and loop diuretics should be used, unless contraindicated. Angiotensin receptor blockers are used in patients with symptoms despite optimal treatment with ß-blockers and ACE inhibitors, unless also taking aldosterone antagonist. If ejection fraction is less then 35%, aldosterone antagonists should be used, in the absence of hyperkalemia and renal insufficiency. Digoxin and oral anticoagulants are indicated in patients with symptomatic heart failure and atrial fibrillation.⁵

Patients with dilated cardiomyopathy are prone to the development of congestive heart failure in the perioperative period, and it is associated with poorer outcome in non-cardiac surgery.⁶,⁷

History is crucial in detecting cardiac diseases that may represent a high risk for surgical intervention, including dilated cardiomyopathy. In taking history special attention should be paid to symptoms of heart failure.⁸

Physical examination should include at least assessment of vital signs (including blood pressure measurements on both arms), carotid pulse, jugular venous pressure, auscultation of heart and lungs, abdominal palpation and extremity inspection, in order to find signs of heart failure.⁹

Routine diagnostic evaluation should include ECG, chest radiography, complete blood count (hemoglobin, leukocytes, platelets), electrolytes, creatinine, glomerular filtration rate, glucose, liver enzymes, urin analysis and possibly BNP.⁵,⁸

Classification of status according to New York Heart Association (NYHA) is useful. In patients with NYHA class III or IV cardiology consultation needs to be done before general anesthesia or any medium and high risk procedure. Minor procedures in stable patients can be done.⁸

In all patients with dyspnea preoperative evaluation of left ventricular function is desirable. Also, in patients with previously established heart failure and worsening of symptoms preoperative evaluation of left ventricular function needs to be done, if not performed within 12 months.⁶ Studies have shown that left ventricular ejection fraction less then 35% is predictor of postoperative adverse cardiac events. However, patients with preserved left ventricular ejection fraction may also have increased cardiovascular risk and for them, similar preoperative preparation is recommended, as in patients with a reduced ejection fraction.⁹

Systolic function can be determined noninvasively by echocardiography or radionuclide techniques. Echocardiography provides additional information on diastolic function as well as valvular function, which can also contribute to congestive heart failure.⁷,⁹

Studies have shown that preoperative use of ACE inhibitors, ß-blockers, statins and aspirin is associated with reduced mortality in patients with left ventricular dysfunction in non-cardiac surgery.⁹,¹⁰

Drug therapy should be optimized and continued preoperatively. This particularly refers to ß-blockers, which are recommended in the perioperative period in all high-risk patients. ß-blockers are also taken in the morning, on the day of surgery. Attention should be paid to fluid balance because fluid overload can lead to decompensation. Surgery should be delayed (unless urgent) in patients with decompensated or untreated cardiomyopathy.⁸,⁹

**HYPERTROPHIC CARDIOMYOPATHY**

Hypertrophic cardiomyopathy is characterized by the presence of increased ventricular wall thickness or mass in the absence of hemodynamic load (hypertension, valvular disease). It occurs in approximately 1:500 of the general population. Many patients have familial form with autosomal dominant inheritance.¹

Unfortunately, first clinical manifestation of hypertrophic cardiomyopathy can be sudden death, often in children or young adults during or after exercise. In symptomatic patients the most common complaint is dyspnea followed by syncope, angina pectoris and fatigue. The most important sign is harsh, late systolic murmur along left sternal border and at the apex. ECG shows left ventricular hypertrophy, diffuse, deep, broad Q waves and atrial and ventricular arrhythmias in many patients. Chest radiography may be normal but mild to moderate enlargement of heart silhouette is common. Primary diagnostic method is echocardiography which shows concentric or asymmetric hypertrophy of the left ventricle, small left ventricular cavity, systolic anterior movement of mitral valve, left
ventricular outflow tract obstruction, often mitral regurgitation.4

Therapy of hypertrophic cardiomyopathy includes β-blockers, which relieve angina pectoris and syncope, amiodarone for prevention of arrhythmias, nondihydropyridine calcium channel blockers which reduce left ventricular outflow tract obstruction. This therapy is continued in the perioperative period. Diuretics, dihydropyridine calcium channel blockers, vasodilators and β-adrenergic agonists should be avoided. Surgical myotomy/myectomy of the hypertrophic septum should be considered in patients who do not respond to medical therapy.4,11

Patients with hypertrophic cardiomyopathy are prone to the development of congestive heart failure as well as supraventricular and ventricular arrhythmias in the perioperative period, and they require special attention.6,7,11

Important for the preoperative preparation of patients with hypertrophic cardiomyopathy is the fact that hypertrophic cardiomyopathy may affect hemodynamics by dynamic left ventricular outflow tract obstruction or may precipitate congestive heart failure due to diastolic dysfunction. Reduction of blood volume, decreased systemic vascular resistance and increased capacity of venous system may cause a reduction of left ventricular volume and thus increase tendency to outflow tract obstruction, with potentially adverse results. Then, reduced filling pressures may lead to significant decrease of stroke volume due to reduced compliance of the hypertrophic ventricle. Reduced compliance of the left ventricle can make patients with hypertrophic cardiomyopathy very sensitive to small amounts of excess intravascular volume, while underfilled left ventricle may increase the dynamic left ventricular outflow tract obstruction, with a resulting decrease in stroke volume and systemic hypotension. Therefore, preoperative preparation should be directed to maintaining intravascular volume within narrow range and controlling heart rate. These procedures reduce the likelihood of congestive heart failure as well as the degree of the left ventricular outflow tract obstruction. β-adrenergic agonists should be avoided because they can increase the degree of left ventricular outflow tract obstruction and reduce diastolic filling.6,7

Atrial fibrillation is the most common arrhythmia in patients with hypertrophic cardiomyopathy. It is usually well tolerated, but can lead to heart failure, syncope or embolic complications, and in some patients to serious ventricular arrhythmias. Electrical or pharmacologic cardioversion is indicated in patients in the first 48 hours from onset, while amiodarone is used to prevent recurrence of arrhythmia. Therefore, amiodarone should be always available in the operating room for treating rhythm disturbances in these patients, if these disturbances occur intraoperatively. The recommended dose for intravenous use is 150mg in rapid infusion, then 1mg/min in the next 6 hours and then 0,5mg/kg in the next 18 hours. Maintenance dose is 200mg per day orally. Aggressive strategy is required for maintaining sinus rhythm in patients with hypertrophic cardiomyopathy. For heart rate control β-blockers, verapamil and digoxin are used, and antiarrhythmic therapy for prevention of embolic complications.11

Patients with hypertrophic cardiomyopathy are at risk of sudden death from ventricular arrhythmias. Preoperative preparation should include holter ECG for detecting ventricular arrhythmias, their prevention with amiodarone and ICD implantation when indicated. In the case of ventricular tachycardia or fibrillation with cardiac arrest in the perioperative period, it should be treated according to current guidelines on cardiac arrest.6,11

RESTRICTIVE CARDIOMYOPATHY

Restrictive cardiomyopathy is characterized by ventricular filling in which increased myocardial stiffness causes large increase in ventricular pressure, with little increase of volume in the presence of normal or reduced diastolic ventricular volumes, normal or reduced systolic volumes and normal ventricular wall thickness. The exact prevalence of restrictive cardiomyopathy is not known, but it is probably the rarest of all cardiomyopathies. It can be idiopathic, familial (usually autosomal dominant) or result from various systemic disorders, in particular amyloidosis, sarcoidosis, carcinoïd, scleroderma and anthracycline toxicity.1

The inability of ventricles to fill limits cardiac output with resultant increase in filling pressures leading to dyspnea and exercise intolerance. These patients have distended neck veins, hepatomegaly, ascites and edema. Jugular venous pressure is elevated and does not fall normally with inspiration (Kussmaul’s sign). Heart sounds are faint and third and fourth heart sounds are common. ECG shows low voltage, nonspecific ST segment and T wave abnormalities and various arrhythmias. Echocardiography reveals symmetrically thickened left ventricular walls, normal or slightly reduced ventricular volumes and systolic function as well as dilated atria.4

In the preoperative period diuretics should be given carefully as well as beta blocker and ACE inhibitors. Atrial fibrillation is common in these patients, and as it worsens ventricular filling, it is necessary to maintain sinus rhythm.6,7

ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY

Arrhythogenic right ventricular cardiomyopathy is histologically characterized by replacement of right ventricular myocardium with adipose and fibrous tissue. Clinically it is characterized by right ventricular dysfunction in the presence of histological evidence of disease. It is rare disease (1:5000) and is usually inherited autosomal dominant.1

Clinically there are manifestations of right heart failure with distension of jugular veins, hepatomegaly and edema, as well as ventricular tachyarrhythmias. ECG shows QRS prolongation and left bundle branch block-type ventricular tachycardia. Antiarrhythmic therapy with β-blockers and amiodarone is useful, as is prevention of sudden death by implantation of ICD.4,12 In the perioperative pe-
period there is the risk of malignant ventricular arrhythmias, therefore antiarrhythmic therapy should be continued.\(^9,12\)

**UNCLASSIFIED CARDIOMYOPATHIES**

Takotsubo cardiomyopathy represents transient regional systolic dysfunction of apex and the middle of left ventricle in absence of coronary artery disease. Patients present by sudden onset of angina-like chest pain, and have diffuse T wave inversions, sometimes with ST segment elevation and mild increase of cardiac enzymes. It is most common in post-menopausal women, and symptoms are often preceded by emotional or physical stress.\(^1\)

In the Center for endocrine surgery of Clinical center of Serbia, patient, who was previously hospitalized due to clinical and ECG picture of acute anteroseptal myocardial infarction in the Coronary care unit of the Emergency center, underwent surgery for pheochromocytoma.\(^2\)

Echocardiography, in addition to left ventricular apex "ballooning", has shown outflow tract obstruction. Selective coronary angiography excluded the existence of coronary artery disease. CT findings of right adrenal tumor and elevated serum and urine catecholamines confirmed the diagnosis of pheochromocytoma. After preparation with phenoxybenzamine patient was operated. Intraoperatively there were expected hypertensive crisis and postextirpation hypotension but changes were not significantly different compared to other patients which underwent surgery for pheochromocytoma, without cardiomyopathy as coexisting disease. This, catecholamine induced cardiomyopathy was reversible. After surgery, echocardiographic findings completely normalized. This case shows that existence of Takotsubo cardiomyopathy does not increase perioperative risk significantly.

**CONCLUSION**

Preoperative evaluation of patients with cardiomyopathies as comorbidity in non-cardiac surgery requires integrated multidisciplinary approach of anesthesiologists, cardiologists and surgeons. Preoperative guidelines are based on knowledge of the pathophysiology of myopathic process. Knowledge of the primary causes of heart muscle disease prior to surgery is of great importance in terms of perioperative management of intravenous fluids. In patients with history or signs of heart failure, echocardiographic evaluation of left ventricular function is recommended to determine the degree of systolic and diastolic dysfunction.

**SUMMARY**

**PREOPERATIVNA PRIPREMA BOLESNIKA SA KARDIOMIOPATIJAMA U NEKARDIJALNOJ HIRURGIJI**

Kardiomiopatije su oboljenja miokarda kod kojih postoji strukturni i funkcionalni poremećaj srčanog mišića, u odsustvu oboljenja koronarnih arterija, hipertenzije, valvularne bolesti i urođene srčane bolesti.

Kardiomiopatije su grupisane u specifične morfološke i funkcionalne fenotipove: dilatativna kardiomiopatija, hipertrofnična kardiomiopatija, restrijktivna kardiomiopatija, aritmogena kardiomiopatija desne komore i neklasifikovane kardiomiopatije.

Pacijenti sa dilatativnom i hipertrofničnom kardiomiopatijom su skloni nastanku kongestivne srčane insuficijencije u perioperativnom periodu. Takodje, pacijenti sa hipertrofničnom i aritmogennom kardiomiopatijom desne komore su skloni poremećajima ritma u perioperativnom periodu.

Preoperativna evaluacija uključuje anamnezu, fizikalni pregled, EKG, radiografiju grudnog koša, krvnu sliku, elektrolite, kreatinin, brzinu glomerularne filtracije, glikemiju, hepatogram, analizu urina, BNP i echokardiografsku evaluaciju funkcije leve komore.

Medikamentoznou terapiju bi trebalo optimizovati i nastaviti preoperativno. Operaciju treba odložiti (osim ako je hitna) kod pacijenata sa dekompensiranoj ili nelećenoj kardiomiopatijom.

Preoperativna evaluacija zahteva integrirani multidisciplinarni pristup anesteziologa, kardiologa i hirurga.

Ključne reči: kardiomiopatije, preoperativna priprema, nekardijalna hirurgija, anesteziologija

**REFERENCES**


