Cardiomyopathy

Cardiomyopathies are a group of cardiac conditions of various aetiologies, whose main initial cause is the affection of the myocardium (heart muscle). They can subsequently evolve into cardiac insufficiency.

They can be:

- Primary – or idiopathic, i.e. without a known cause
- Secondary to other conditions, such as:
    - Metabolic conditions: nutritional, endocrine etc.
    - Systemic conditions: collagen diseases, neoplasias etc.
    - Neurological, neuromuscular, muscular conditions: dystrophies, ataxias, myopathies etc.
    - Toxic conditions: medication-related, biological
    - Conditions caused by physical agents: radiation

Cardiomyopathies are of three types:

- Obstructive and non-obstructive hypertrophic cardiomyopathy
- Restrictive cardiomyopathy
- Dilated cardiomyopathy

A. Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy is a myocardial condition of unknown cause (usually genetic) that is described as the thickening (hypertrophy) of the ventricular myocardium and the significant disorganisation of the myocardial (heart muscle) architecture. Hypertrophy can be symmetric, i.e. homogenously distributed or asymmetric. The latter mainly affects the interventricular septum (the wall that separates the two ventricles) and causes the obstruction of the left ventricular ejection fraction, thus leading to obstructive hypertrophic cardiomyopathy.

The symptoms vary from non-existent to fatigue caused by progressively smaller efforts, dyspnoea (shortness of breath), angina pectoris, dizziness, syncope (loss of consciousness) and palpitations.

Suspicion arises based on the physical exam: the presence of a systolic murmur during the ECG exam (that changes in approx. 80-90% of the cases). The diagnosis is determined through the echocardiography, when the physician determines the degree of thickening of the ventricular myocardium and the degree of obstruction in the left ventricular ejection fraction (elements that are extremely important for choosing the method of treatment).

When patients are diagnosed with hypertrophic cardiomyopathy, it is absolutely necessary for the members of their family (parents/children/brothers) to get examined, since, usually, this condition is genetically transmitted.

The evolution of the disease is variable and unpredictable, with a mortality rate of 3-4% per year. In general, death is sudden and it is caused by rhythm disorders (ventricular fibrillation).

Factors that predispose to sudden death:

- Major factors:
    - Antecedents of cardiac arrest

    - Non-sustained/sustained ventricular tachycardia proven through ECG, Holter ECG etc.
    - Sudden death in the family history
    - Unexplained syncope (loss of consciousness)
    - Ventricular wall thickness exceeding 30 mm (determined through echography)
    - Abnormal decrease of the arterial pressure while sustaining effort
- Potential factors:
    - Arterial fibrillation
    - Myocardial ischemia
    - Degree of obstruction of the left ventricular ejection fraction – measured through echography or cardiac catheterisation
    - Excessive effort (competition)

The methods of treatment are:

- Medication: beta blockers, antiarrhythmic drugs
- Interventional treatment: alcohol septal ablation
- Surgery: septal myomectomy, mitral valve replacement
- Dual chamber cardiac pacemaker implantation: in certain situations

All patients over 40 years that are diagnosed with hypertrophic cardiomyopathy, who present with retrosternal pain episodes or with risk factors for ischaemic heart disease need to undergo a coronary catheterisation.

B. Dilated cardiomyopathy

Dilated cardiomyopathy is described through an increase in the size of the left and/or right ventricle and it can be caused by the progression of other cardiac diseases: ischaemic heart disease with or without myocardial infarction, valvular disease, congenital heart diseases, arterial hypertension, other non-cardiac diseases: neurological, inflammatory (collagen diseases, autoimmune diseases etc.), infectious (viral, bacterial, fungal, parasitic etc.), metabolic (diabetes mellitus, endocrine, nutritional deficiencies), toxic (alcohol, medication, drug abuse etc.) or idiopathic (of unknown cause).

The signs and symptoms vary from non-existent to cardiac insufficiency due to the affection of the function of the left side of the heart: left ventricle and atrium (fatigue during progressively smaller efforts and during rest, dyspnoea – shortness of breath that occurs, at first, while sustaining effort. However, as the disease progresses, shortness of breath occurs during day to day maintenance activities or while resting, up to acute pulmonary oedema) In time other symptoms appear due to the affection of the right side of the heart, i.e. the affection of the right ventricle and atrium (accumulation of fluid in the legs, lungs, heart, abdomen, the increased size of the liver).

As the disease progresses, certain complication can occur:

- Systemic/pulmonary embolism: the formation of blood clots that reach the blood circulation and cause obstruction in the cerebral arteries (with cerebral vascular accidents), in the legs/arms (acute ischemia of the legs/arms) or in other vascular area;
- Arrhythmias: especially atrial fibrillation, supraventricular and ventricular extrasystoles, ventricular tachycardia, ventricular fibrillation;
- Sudden death;

Investigations

- ECG

- Echocardiography
- Coronary catheterisation
- Holter ECG

The treatment depends on the stage of the disease and consists of a combination of the following:

- Medication: beta blockers, diuretics, conversion enzyme inhibitors, digitalis dugs, antiaggregants, statin, nitrates, anticoagulants (depending on the cause, on the presence of complications and on the associated diseases);
- Cardiac defibrillator implantation: to prevent sudden death;
- Ventricle assisting devices;
- Heart transplant.