

mous tension in the ventricular wall, namely not only during diastole, but also in consequence of decreased contractility during systole. As a result this type of maintenance of the minute volume is very uneconomical. The excessive tension in the ventricular wall replenishes energy of the heart, and its pumping function in spite of maximal utilization of Frank-Starling's mechanism definitely fails.

3.8 Valvular defects of the heart

The role of both atrioventricular and semilunar valves is to assure an interrupted movement of the blood in the direction from atrii toward the aorta or a. pulmonalis. In dependence on the degree and speed of the development the valvular disorder has a distinct impact on organism. A moderate degree of a valvular lesion does not entail any hemodynamic changes. The only symptom may be an auscultation finding, sometimes it is even entirely symptomless. On the other hand a severe anatomical valvular change can lead to a significant deterioration of hemodynamics and to alteration of the health state of a patient. An especially dramatic clinical picture develops when the severe valvular lesion supervenes rapidly. The resultant clinical picture is moreover determined also by the functional reserve of the cardiac muscle.

Two types of valvular lesions are distinguished:

- valvular stenosis – manifests itself in that phase of the cardiac cycle, when the valve is closed and the blood is thus propelled via a narrowed orifice.
- valvular insufficiency – manifests itself in the phase of the cardiac cycle when the valve is to be closed; in consequence of imperfect packing of the valvular orifice a part of the blood regurgitates into the precedent compartment of the heart.

Pure valvular stenosis or insufficiency are rare.

The majority of cases yields a simultaneous development of both types of valvular lesions.

Valvular lesions can be inborn or acquired. Acquired valvular lesions develop most frequently due to rheumatic fever. Less frequently they can develop in consequence of bacterial endocarditis, syphilis and at an older age degenerative changes as valvular fibrotization and calcification.

3.8.1 Mitral stenosis

Stenosis of the mitral valve develops predominantly in consequence of rheumatic endocarditis. It develops in the course of several years following the attack of the rheumatic fever. In consequence of immunoalterative inflammatory process the free margins of the mitral valve stick together. The mitral orifice which is in an adult person 4–6 cm² in surface diminishes. The clinical picture depends upon the degree of narrowing of this orifice.

The hemodynamic disturbance manifests itself during ventricular diastole when the blood flows from the left atrium into the left ventricle. Moderate stenosis of the left AV orifice does not manifest itself hemodynamically. It is the stenosis below 2,02 when the blood flow from the left atrium into the left ventricle deteriorates. The subsequent consequence is represented by accumulation of the blood in the left atrium with a subsequent elevation of atrial pressure. The character of the diastolic filling of the left ventricle substantially alters. A normal diastole has three phases:

1. The phase of rapid filling of ventricles when after the opening of AV valves the ventricles are dumped with 80% of the total amount of in-flowed blood.
2. The first phase is followed by the phase of diastasis. The elevated pressure in the left ventricle encloses the AV valve nearly completely, and the blood flow into the ventricle soon ceases.
3. The third phase is the period of atrial systole; during this time the remnant 20% of the filling volume of blood is forced into the ventricle

The stenosis of the mitral valve disturbs the three-phase character of the diastolic filling. The blood during diastole flows but gradually via the narrowed orifice into the left ventricle. In consequence of high

pressure in the left atrium during diastole there is a large pressure gradient between the left atrium and ventricle. The ventricle is being filled in a slow even manner during the entire diastole. **The ventricle is being filled for a longer period and under a higher pressure gradient.** Providing the stenosis is not too tight such a filling assures a normal diastolic volume of the left ventricle.

Tachycardia accompanying the physical exercise substantially deteriorates the hemodynamic situation in the heart afflicted with mitral stenosis. Increase in frequency hence takes place chiefly on the account of diastole which becomes significantly shorter. Thus the period of the left ventricular filling becomes shorter. Tachycardia deteriorates the left ventricular filling in a heart afflicted with mitral stenosis in a greater extent than in a heart with a normal mitral orifice. The substantial part of the blood in a normal heart (app. 80% of the total volume of inflowed blood) reaches the ventricle during the first phase of diastole. Therefore the shortening of the diastolic period in tachycardia decreases the diastolic filling of the ventricle in a healthy heart to a proportionally only a little. In contrary to the latter the stenosis of the mitral orifice entails a continuous filling of the ventricle during the entire diastole and therefore shortening of diastole significantly decreases the end-diastolic volume of the left ventricle.

The situation deteriorates also due to the advancement of the mitral stenosis. As a result, the filling of the left ventricle is reduced diminishing thus the stroke volume. (forward failure). Simultaneously the pressure in the left atrium, pulmonary veins and pulmonary capillaries elevates. It manifests itself as dyspnea of various degree up to pulmonary edema (backward failure). The symptoms resemble those of the left side heart failure. Conversely to the left ventricular heart failure in which the left ventricle is dilated the left ventricle in mitral stenosis is of normal size or even atrophic as it works with smaller amount of blood. The minute volume is decreased in severe mitral stenosis. Arterial hypotension reflexively stimulates the sympathetic system. The latter increases the contractility of the left ventricle and evokes peripheral vasoconstriction in order to assure perfusion of vital organs – the cardiac muscle and brain.

Prolonged duration of a severe form of mitral stenosis evokes pulmonary changes which are con-

sidered to **protect the lungs against the pulmonary edema:**

1. Significant thickening of alveolocapillary membrane, which acts as a barrier against the entrance of fluid into alveoli.
2. The enhanced resistance in pulmonary arterioles which is both functionally and later also anatomically determined. **Functional vasoconstriction** originates due to hypoxaemia in consequence of the deteriorated ventilation of the lungs which are overfilled with blood. Aside from this, pressure elevation in pulmonary veins and capillaries alone which is brought about by stimulation of vascular receptors, reflexively evoke spasms of arterioles. Prolonged duration of pressure elevation in a.pulmonalis brings about **morphologic changes** as well. They manifest themselves by the thickening of intima and media.

Increase in resistance in pulmonary arterioles has both positive and negative impacts on the heart. The positive implication resides in a decreased supply of blood into the lungs which inhibits the origin of pulmonary edema. On the other hand the increased resistance in the pulmonary arterioles elevates the pressure in a.pulmonalis and in the right ventricle. As a result the right ventricle fails, the blood supply into the lungs is decreased and the dyspnea withdraws. The life of a patient is prolonged on the account of the reduced minute volume of the left side of the heart and right ventricular failure. The quality of a patients life with a severe mitral stenosis is therefore significantly decreased.

The chief clinical symptom of mitral stenosis is **dyspnea**. The degree and character of dyspnea have similar character as in left ventricular failure. The degree of dyspnea correlates with the size of the mitral orifice surface and pressure in the left atrium. Reduction of the orifice to 0,5 cm² results in very high pressure in the left atrium and pulmonary edema threats to develop.

The picture of a severe mitral stenosis includes the **classical triade of symptoms:** embolization into arterial system, hemoptysis and cyanosis.

- **Embolization** is caused by turbulent blood flow and blood stagnation in the left atrium. The predilection site of thrombi formation is the

atrial auricle. This is located aside from the main stream of blood flowing into the left atrium and thus the blood in the auricle almost does not flow. Therefore thrombi originate and may tear off causing embolization. Embolization occurs most frequently in the brain. It can however occur in any part of the arterial system (Limbs, kidneys, mesenterial arteries). Embolization often occurs repeatedly.

- **Hemoptysis** occurs either in the form of minute hemorrhage or as a massive apoplexy. It is brought about predominantly by rupture of the anastomosis between the pulmonary and bronchial veins. Under normal conditions the blood does not flow through these two veins. In consequence of retrograde pressure elevation in the pulmonary veins the anastomoses open wide and as their walls are thin they are likely to rupture.
- **Cyanosis** is of cold, acral type. As a result of compensatory peripheral vasoconstriction the blood flow via capillaries is decreased. On the basis of tissue acidosis the extraction of oxygen intensifies and thus the amount of reduced hemoglobin at the venous end of capillaries increases. The typical facies mitralis is determined by means of combination of cyanosis of the lips and erythema of the cheeks.

Cases of severe mitral stenosis are frequently linked to atrial fibrillation. The reason of atrial fibrillation origin is the dilatation of the left atrium and the high intraatrial pressure. According to the Laplace's law the stress in the atrial wall is thus significantly increased and the energy consumption as well. The $\text{Na}^+\text{-K}^+$ ATP-ase fails and numerous ectopic foci originate in atri. These represent the basis for the origin of extrasystoles, atrial tachycardia, or if the ectopic foci are numerous, atrial fibrillation may occur. Atrial fibrillation has three basic consequences:

- deterioration of the left ventricular filling
- disadvantageous tachyarrhythmia
- stagnation of the blood in the dilated left atrium and thus formation of thrombi

Fatigue and decreased physical performance.

They originate in consequence of the decreased cardiac output and of decreased minute volume. They are not caused by left ventricular failure. The function of the left ventricle is on the contrary excellent and the ejection fraction is even increased. The low minute volume is determined by insufficient left ventricular filling during diastole. If mitral stenosis develops in children it may result in retardation of physical and psychical development due to chronic hypoperfusion of the peripheral tissues.

Less characteristic symptoms of mitral stenosis are: dysphagia and hoarse voice (in consequence of compression of oesophagus and nervus recurrens owing to left atrial dilatation).

Heart failure and pulmonary oedema occurs most often in coincidence with tachycardia and tachyarrhythmia of atri which deteriorate the ventricular filling. The cause may reside also in intercurrent infection which increases the demands of the periphery to oxygen, or a progression of the mitral valve stenosis in consequence of a prolonged course or repeated attacks of rheumatic fever.

3.8.2 Pathogenesis of clinical symptoms

Auscultation reveals **an accentuated first heart sound, a clap sound due to the mitral valve opening and holodiastolic murmur with a presystolic accentuation.**

- Accentuation of the first heart sound is determined by several factors. Under normal circumstances at the end of diastole the ventricle is filled with blood and the mitral valve encloses. If the mitral orifice is narrowed, the end-diastolic content is not large enough to enclose the mitral valve. Mitral valve is enclosed as late as during the rapid ventricular pressure elevation in the phase of isometric contraction. This sudden enclosure of mitral valve brings about the accentuation of the first heart sound which is also delayed. Loud enclosure of the mitral valve is supported also by strong contraction of the left ventricle. The latter takes place in consequence of activation of the sympathetic nerve due to insufficient left ventricular stroke volume and peripheral hypotension.

- The opening mitral tone is determined by an abrupt opening of the mitral valve and also by fast blood flow into the ventricles in consequence of the increased atrioventricular gradient. The opening mitral tone is not audible when the valves are rigid due to their calcification.
- The diastolic murmur at the apex is of low-frequency and lasts throughout the entire diastole. The presystolic accentuation is caused by accelerated blood flow via the narrowed orifice in consequence of atrial contraction in the final phase of diastole. The presystolic accentuation disappears when atrial fibrillation occurs. The murmur is best audible on the left side position. The pulse in mitral stenosis is small (pulsus parvus) (In consequence of pulmonary hypertension the pulmonal proportion of the second heart sound is usually accentuated).

The methods of imaging detect dilatation of the left atrium and the small left ventricle. In late phases both the right atrium and right ventricle dilate as well. In consequence of the right ventricular dilatation the anulus of the tricuspidal valve extends and a functional tricuspidal insufficiency develops. This state is referred to as **tricuspidal** mitral defect. Pulmonary hypertension can determine the functional insufficiency of the pulmonal valve. The course of the disease depends upon the fact as to whether the stenosis is deteriorating, as well as upon possible concomitant complications. The states which deteriorate the course of mitral stenosis include gravidity (augments the circulating volume), virus infections (increase the demands of tissues in oxygen), great physical exertion. These factors should be avoided by patients.

3.8.3 Mitral insufficiency

Represents the most frequently occurring acquired valvular defect. It can occur alone or in combination with mitral stenosis. In dependence on etiology various parts of the valve may be afflicted:

- The front or back cusps of the mitral valves - may be deformed in consequence of rheumatic carditis. The cusps themselves are usually afflicted also in the so-called prolapse of mitral valve, and in an inborn splitting of one of the

cusps. At an older age calcification is often present with consequently restricted movement of the cusps.

- Fibrous tendons which attach the valves to the papillary muscle - are often calcified or coalescent after rheumatic carditis
- Papillary muscle - is often hypofunctional in ischemic heart disease, and infarction may bring about its rupture accompanied with huge regurgitation.
- Mitral anulus - is dilated in left ventricular failure. Intact valves with preserved movability do not manage to enclose the enlarged mitral orifice and the so-called relative mitral insufficiency develops.

The basic functional defect in mitral insufficiency is the backward flowing of the blood from the left ventricle into the left atrium during ventricular systole. The left atrium is filled with the blood from the pulmonary veins and, surplus, it is filled also with the blood from the left ventricle. An increased volume overload of the left atrium is in effect.

During the ventricular diastole the left ventricle receives blood, the amount of which is increased by the volume which had escaped into the left atrium during the systole. The left ventricle is thus volume overloaded.

During the ventricular systole the blood flows in two directions. The insufficient mitral valve lets the excessive amount of blood to regurgitate into the left atrium and the periphery receives a normal amount of blood. The volume which in consequence of mitral insufficiency returns into the left atrium moves in a pendulous manner between the left atrium and ventricle. Volume overload inflicts both the left atrium and left ventricle.

The above mentioned changes result in an increased amount of blood in the left ventricle at the end of diastole. It determines left ventricular hypertrophy development of volume type which compensates the presented volume overload:

1. The extent of regurgitation is determined foremostly by anatomical changes in the mitral valve. Aside from this the backward blood flow depends upon the pressure in the left ventricle. High ventricular pressure is accompanied

by a large ventriculo-atrial gradient. Therefore, all the states which elevate the intraventricular pressure (hypertension, stenosis or aortic coarctation) augment the reflux of blood into the atrium even in moderate degree of mitral insufficiency.

2. The state of atrial musculature. Providing the atrial musculature is of optimal elasticity and contractile state, the left atrium "resists" to the excessive delivery of blood by increasing its tonus. The pressure in atri and pulmonary veins thus elevates. On the contrary the atri afflicted with degenerative processes and thus having their musculature partially substituted with afunctional connective tissue dilate in consequence of overload in the blood volume, but the atrial pressure does not elevate significantly. Under these conditions regurgitation of blood manifests itself by an enlarged left atrium and but a small increase in pressure in pulmonary veins and capillaries.
3. The velocity of mitral insufficiency development. Gradual enlargement of mitral regurgitation exposes the left atrium to an increased delivery of blood. The left atrium accomodates to this situation by hypertrophy and dilatation, by virtue of which the pressure in pulmonary beds elevates but moderately. Sudden infliction with mitral regurgitation (rupture of the papillary muscle due to infarction, perforation of a valve due to bacterial endocarditis) abruptly elevates pressure in an unprepared left atrium. The pressure elevation is conveyed into the pulmonary capillary bed and pulmonary edema develops.

3.8.3.1 Pathogenesis of clinical symptoms

It is generally known that the individual heart parts can cope easier with volume than with pressure overload. This can be applied also as far as mitral insufficiency is concerned. The symptoms due to mitral insufficiency are therefore similar but less serious than symptoms due to mitral stenosis.

Small mitral insufficiency manifests itself solely by an auscultatory finding. However, danger impends in the origin of bacterial endocarditis on an anatomically altered valve. Palpitations frequently represent an initial symptom. They develop in consequence

of large volume which the left ventricle has to process and also of dysrhythmias present. **Hemoptoe, embolization, cyanosis and fibrillation** occur less frequently than in mitral stenosis. Severe regurgitation entails congestion in the pulmonary circuit and pulmonary edema and later even dilatation of the right compartments of the heart with triculsoidal and pulmonary insufficiencies.

The auscultatory finding is characterized by the first heart sound being weak and a blowy holosystolic murmur.

- The attenuation of the first heart sound develops as a result of imperfect enclosure of a deformed mitral valve during ventricular systole.
- The murmur is soft, blowy and of low frequency. It develops as soon as during isometric contraction of ventricles when the pressure in the left ventricle exceeds the pressure in the left atrium. It lasts throughout the entire systole and overlaps the beginning of diastole. Its intensity is maintained on the same level throughout the whole systole. It is best audible at the apex and propagates into the direction of axilla. The intensity of the murmur is well correlated with the degree of regurgitation. The second heart sound is in case of pulmonary hypertension split with an accentuation of its second component.

The prognosis depends upon the degree of hemodynamic change, the progression of the disease and upon the complications. The disease is deteriorated by intercurrent infections (the demands of the periphery put on the heart work increases), tachyarrhythmia of atri (diastole shortens and the overfilled left ventricle insufficiently evacuates) and repeated attacks of rheumatic carditis (incompetence of valvular enclosure deteriorates).

3.8.4 Aortic stenosis

Aortic stenosis develops most frequently as a result of rheumatic fever. Less frequently represents a consequence of sclerotic changes in valves, or bacterial endocarditis. Stenosis of the aortic valve often occurs together with aortic insufficiency or mitral stenosis. The surface of the aortic mouth which is in an adult app. 3,5 cm² diminishes. The clinical picture depends upon the degree of the aortic valve narrowing.

Hemodynamic impairment manifests itself during systole of ventricles when the blood is ejected from the left ventricle into the aorta. Moderate narrowing of the aortic orifice has not any hemodynamic manifestation. Yet, aortic narrowing down to 50 % or even 25 % invalidates the blood flow from the left ventricle into the aorta. The resistance against blood ejection increases. According to Laplace's law the systolic tension in the wall of the left ventricle, i.e. afterload, increases. Such gradually developing increased pressure overload is compensated by left ventricular hypertrophy. The left ventricular wall thickens, but its internal volume does not change. Hence hypertrophy originates being of pressure type and referred to as concentric hypertrophy where the ratio of the wall width to ventricular volume changes in favour of wall width. Hypertrophy has two basic impacts:

1. The wall stress returns to original values.
2. At the same time the total left ventricular contractile ability improves. Left ventricle is thus able to force the blood via constricted orifice and secure adequate pumping function of the heart against increased resistance.

Despite the secured adequate perfusion of periphery, the situation in the heart is substantially altered. Both systolic and diastolic pressures increase in the left ventricle. The systolic pressure reaches enormous values – 200 to 300 torr. Intraventricular pressure during systole mounts more steeply and reaches the peak sooner than under physiological conditions. There is a great pressure gradient between the left and right ventricles. The systolic pressure in aorta achieves lower values and increases less rapidly than under physiological conditions.

Also the diastolic pressure in the left ventricle is increased. This is a consequence of the loss of elasticity of the left ventricle due to its increased thickness. Due to this situation the normal end-diastolic pressure does not evoke adequate expansion of walls, it causes though excessive growth of end-diastolic pressure.

Owing to compensatory hypertrophy the pumping activity of the heart as a whole is maintained and patients need not suffer from any difficulties for a rather long period of the disease.

Pathogenesis of clinical picture. Clinical picture in cases of severe aortic stenosis is characterized by

gradual onset of the triade symptoms: angina pectoris, syncope, left heart failure. In addition there is a significant risk of sudden death in these patients.

1. **Angina pectoris** originates in consequence of oxygen insufficiency. A combination of several factors participates in the latter: decreased aortic diastolic pressure and thus decreased coronary perfusion pressure, increased pressure work of the heart, and quite often concomitant atherosclerosis of coronary arteries in older patients.
2. **Syncope** originate mostly due to physical work. They are the consequence of the left ventricular inability to increase the heart ejection in order to fulfil the increased oxygen demands of periphery. Muscular strain causes that the ejected blood is preferentially moved into the dilated muscular vascular bed with consequent hypotension and cerebral hypoxia. Cerebral hypoperfusion results in vertigo and collapse.
3. Aortic stenosis can reach a stage in which despite compensatory hypertrophy of the left ventricle the stroke volume decreases and the blood begins to stagnate in the left ventricle. Gradually a complex picture of **left ventricular failure** develops. Left ventricular failure is included among late symptoms and signalizes unfavourable prognosis.
4. The late stage of aortic stenosis with developed stenocardia yields a high risk of sudden death. It originates mostly due to physical strain. As the blood flows preferentially into the working muscles, it may even deepen the myocardial hypoxia. Numerous ventricular ectopic foci originate with high risk of ventricular fibrillation. Ventricular fibrillation or a pronounced cerebral hypoxemia represent the direct cause of sudden death.

The auscultation finding in aortic stenosis is characterized by a **systolic "clapp", rasp loud ejection murmur with its maximum above aorta (second intercostal space parasternally on the right) and weakened second heart sound.**

- The systolic "clapp" has an analogic reason as the mitral opening tone. It originates by abrupt opening of the aortic valve in consequence of

a great gradient between the left ventricle and aorta.

- The typical rasp murmur is propagated into carotids, to the apex and into interscapular space. It is of crescendo decrescendo (spindle) character. Chronologically it is localized into the meso or even telesystole. It is more pronounced in a sitting leaning forward posture, and with the breath held at the end of expiration.

The typical spindle shape of this murmur is caused by the fact that at the beginning and at the end of ejection period of systole the pressures in the left ventricle and in aorta are nearly the same with a relatively small gradient through the aortic orifice. The blood flow is minimally turbulent and both at the beginning and end of ejection the murmur is weak. During the ejection the pressure in the left ventricle elevates to the maximum. The pressure gradient grows, turbulent flow originates and the murmur reaches its maximum. During the second part of ejection the gradient is of declining character, the turbulence decreases and thus also the intensity of the murmur.

The chronologic localization of this murmur into the mid and late phase of systole originates in consequence of stenosis of the aortic orifice. The smaller the orifice, the later the sufficient pressure elevation in the left ventricle is achieved in order to force the blood through the stenotic orifice.

The weakening of the second heart sound is determined by slow pressure elevation in aorta. In consequence of this the closure of aortic valve is not rapid, but gradual, which fact deprives the intensity of the second heart sound.

Palpation above aorta reveals a rasp scratching whirl. It has the same localization and propagates itself into the same areas as the murmur. It is determined by the turbulent blood flow.

The peripheral pulse is small, with a slow growth and decline (similarly as the intensity of murmur) – the so-called *pulsus parvus and tardus*. It is delayed after the apex beat, which coincides with the delayed increase in aortic pressure.

During the period of many years the patients with aortic stenosis do not suffer from any difficulties. The left ventricle, being the strongest compartment of the heart has a great ability of compensation. When the

symptoms (stenocardia, collapse, or left ventricular failure) occur, it means that the compensatory abilities of the heart are exhausted. The prognosis is hence restricted to a period of three or four years. In such a stage the only solution which prolongs a patient's life significantly and improves its quality is an operation.

3.8.5 Aortic insufficiency

Aortic insufficiency occurs either alone, or more frequently in combination with aortic stenosis or mitral defect. The most frequent cause is rheumatic fever, less frequently atherosclerosis of the valvular cusps, bacterial endocarditis and syphilitic aortitis. The aneurysm of ascending aorta brings about dilatation of the aortic annulus and results in insufficient enclosure of the aortic orifice by intact valvular cusps.

The basic functional impairment in insufficiency of aortic valve is represented by reverse blood flow from aorta back into the left ventricle during the ventricular diastole. The left ventricle is filled simultaneously by both the blood from the left atrium and regurgitated blood from aorta. The latter fact thus increases the volume load of the left ventricle.

During the ventricular systole the aorta receives an amount of blood increased by the volume which regurgitated into the left ventricle during systole. In this way also the aorta is overloaded and its dilatation may arise.

The blood in aorta thus flows in two directions during diastole. Via the incompletely closed aortic valve the "excessive" volume of blood returns into the left ventricle and the periphery receives a normal amount of blood. The volume which in consequence of aortic insufficiency returns into the left ventricle moves therefore **pendulously** between the left ventricle and aorta.

The presented alterations result in increased amount of blood in the left ventricle at the end of diastole. In consequence of volume overload the internal volume of the ventricle increases and the ventricular wall less markedly thickens. The ratio of ventricular volume to the wall width shifts in favour of the volume. Hypertrophy of the volume type – eccentric hypertrophy develops which compensates the volume overload for a long period (10–30 years).

The regurgitated amount of blood can represent 30–70% of the stroke volume. The hypertrophic left ventricle reaches enormous size and weight of

700–1000 grammes. The working capacity of the heart markedly increases. The stroke volume of the left ventricle inclusive of the regurgitated component can reach 300 ml, which corresponds with a minute volume of 30 l. This cardiac output resembles the maximal output of a trained sportsman. It suggests that the maximal cardiac working capacity is approximately equal under physiological and pathological circumstances.

The pressure conditions in the left ventricle depend on the size of regurgitation, state of ventricular musculature and frequency of contractions.

1. The size of regurgitated volume depends on the severity of the valvular defect. In addition to that the regurgitation increases parallelly with the increase of arterial pressure.
2. The left ventricular wall hypertrophy itself deteriorates the elastic properties and at the given end-diastolic volume the ventricular pressure thus increases.
3. The increased frequency shortens the whole cardiac cycle, and especially the diastole. Tachycardia thus restricts regurgitation and decreases the diastolic pressure. In this disease tachycardia may act as a certain compensatory mechanism.

3.8.5.1 Pathogenesis of clinical symptoms

Clinical symptoms are similar but less pronounced than in aortic stenosis and have also a similar pathogenesis. **Angina pectoris, syncope, left ventricular failure, sudden death** occur relatively late and manifest a severe stage of the disease. Earlier symptoms include:

1. **Palpitations**, which coincide with a large systolic stroke volume, strong pulsation of the left ventricle and ventricular extrasystoles.
2. **The consequences of peripheral vasodilatation.** In consequence of high systolic pressure the peripheral vessels reflexively dilate. The patients suffer from vertigo, increased perspiration and do not tolerate heat.

Auscultation finding is characterized by **the diastolic decrescendo blowing murmur, best audible above aorta (second intercostal space parasternally**

on the right). It propagates into Erb's point – third to fourth intercostal space parasternally on the left approximately 3 cm from the sternum. It has a high frequency decrescendo character. Chronologically it is localized into the proto- and mesodiastole. It is more pronounced at a leaning forward sitting posture with a breath held at the end of expiration. **The first heart sound is attenuated.** It is caused by a premature enclosure of the mitral valve as a consequent of high intraventricular pressure, owing to which the mitral component does not participate in the origin of the first heart sound. **Also the second heart sound can be weakened.** In this case the weakening is caused by incomplete enclosure of the deformed aortic valve.

Severe aortic insufficiency produces a **diastolic Flint-Austin's murmur** audible over the apex. It resembles the murmur audible in mitral stenosis. On the contrary to mitral stenosis the accentuated first sound is not present, neither the mitral opening tone. The origin of this murmur can be caused by several factors:

- Relative mitral stenosis is caused by enclosure of one of the mitral valve cusps by the regurgitating flow.
- Premature enclosure of mitral valve by high intraventricular pressure.
- Collision of the mitral and regurgitation streams.

The systolic pressure is in consequent of large stroke volume high. The diastolic pressure is on the contrary very low and in severe insufficiency drops to zero. The decrease in diastolic pressure is caused by regurgitation of blood into the left ventricle on one hand and peripheral vasodilatation on the other.

Pressure changes correspond with the character of pulse. The pulse quickly reaches high values and rapidly declines – **pulsus celer et altus**.

Enormous progressive movement of the blood during systole and retrograde movement of the blood during diastole manifest themselves as a Musset's symptom (rhythmic movement of the head simultaneously with each ventricular systole) and Quinck's capillary pulsation (cyclic reddening and paling of the nail's bed and skin of fingers). The development of the disease and prognose are similar to those in aortic stenosis. The hypertrophic left ventricle

compensates the volume overload for decades. **The patients even manage physical strain.** This situation implies that tachycardia which is concomitant to physical exertion, shortens the diastole and thus also regurgitation and to a certain extent has a compensatory impact. Occurrence of symptoms shortens the survival to 3-4 years. Deterioration of clinical state is entailed especially by progression of the primary disease and thus increased regurgitation and by potential setting of bacterial endocarditis. Experimental and clinical studies suggest that also unchanging extent of regurgitation may result in left ventricular failure after prolonged duration of the disease.

3.8.6 Valvular defects of the right heart

3.8.6.1 Tricuspidal insufficiency

Tricuspidal insufficiency is in most cases a relative valvular disorder. It may be seen in the connection with right cardiac failure with consequent dilatation of the right ventricle. This situation causes dilatation of the annulus of tricuspidal valve and thus a complete enclosure of the valve becomes impossible.

The right atrium is thin and has a small compensatory ability. It relatively quickly fails and signes of blood gession before the right heart develop.

The clinical manifestation of the disease results from stagnation of the blood in the systemic circuit: **increased content of jugular veins, hepatomegaly, hepatojugular reflux, oedema, ascites.** Palpably revealed manifestations include **systolic pulsation of the liver** which originates by transfer of the regurgitation wave into hepatic veins. Stagnation of the blood in the liver deteriorates the hepatic function. **Hepatocellular icterus arises and symptoms of haemorrhage originates** in consequence of decreased production of blood clotting factors. **Dyspepsia** originates as a result of stagnation of the blood in the digesting tract. A symptom of severe right heart failure is considered a **rapid withdrawal of dyspnea** in patients with left ventricular failure. This situation occurs in consequence of deteriorated pumping function of the right ventricle. This results in decreased blood supply into the lungs and decreased capillary pulmonary pressure.

Auscultation finding resembles that of mitral insufficiency but is localized at the inferior left border of the sternum. It is a holosystolic, soft, blowing mur-

mour with a weakened first heart sound. It is typical of this murmur that it intensifies in inspirium, when the blood supply from the capacity bed into the right heart increases.

3.8.6.2 Tricuspidal stenosis

Tricuspidal stenosis occurs as a consequence of rheumatic fever together with mitral or aortic valvular defects. Stenotic valve inhibits the inflow of the blood into the right ventricle during diastole. The blood accumulates in the right atrium which fails. The clinical picture resembles that of tricuspidal insufficiency.

Obstruction of the tricuspidal orifice evokes regurgitation of the blood from the right atrium into the jugular veins causing thus their pulsation. This state is referred to as **atrial venous pulse.**

Auscultation reveals diastolic murmur which is of a similar character as that audible in mitral stenosis. It is though localized at the inferior border of the sternum.

3.8.6.3 Insufficiency of the pulmonary valve

Pulmonary insufficiency rarely originates on the basis of rheumatic or bacterial endocarditis. Predominantly it occurs as a relative-functional incomplete enclosure in states with pulmonary hypertension and dilatation of pulmonary arteria which bring about distension of the pulmonary annulus.

The auscultation finding is similar to that in aortic insufficiency. It is characterized by a blowing, **proto-** and **mesodiastolic** murmur being of high frequency, (Graham-Steel's murmur). It is best audible above the pulmonary artery, i.e. above the second intercostal space parasternally on the left. The second sound is weakened in consequence of the deformed pulmonary valve.

3.8.6.4 Stenosis of pulmonic valve

It is rare and originates in consequent of rheumatic endocarditis. The right ventricle becomes hypertrophic. Its compensatory abilities are though significantly smaller than in case of the left ventricle. Right heart failure takes place relatively fast. The auscultation finding resembles that of aortic stenosis – it is a systolic, loud, scratchy murmur which is of crescendo-decrescendo character. It is best audible

above the pulmonary artery. The second sound is weakened in consequence of deformation of the pulmonary valve.

3.9 Cardiomyopathies

Cardiomyopathies represent a group of heterogeneous diseases of the heart which involve a chronic, idiopathic pathologic process inflicting especially the cardiac muscle. The group of cardiomyopathies includes neither ischemic nor inflammatory impairment of myocardium.

The term cardiomyopathies (respectively myocardopathies) has currently a significantly different meaning in comparison with its original content. It was a tradition to distinguish coronary and noncoronary cardiomyopathies. Coronary cardiomyopathies included diffuse ischemic impairment of the cardiac muscle. Noncoronary cardiomyopathies included nonischemic impairments of myocardium, namely those with unknown etiology (primary cardiomyopathies), or secondary affliction of the cardiac muscle in the frame of a particular extracardiac disease (secondary cardiomyopathies).

Nowadays cardiomyopathies are comprehended as noncoronary diseases of the cardiac muscle with unclear etiology. If the myocardium is afflicted in the frame of extracardiac primary disease we speak about specific diseases of the cardiac muscle (these are not included into cardiomyopathies).

Currently the classification of cardiomyopathies is accepted including three basic types:

- dilating cardiomyopathy
- hypertonic cardiomyopathy
- restricting cardiomyopathy

3.9.1 Dilating cardiomyopathy

Dilating cardiomyopathy is characterized by several typical properties which are however not specific and can occur in different types of heart impairments.

Let simultaneous occurrence of more of them, unknown etiology of the particular state of the heart and an echographic finding of dilated heart enable to state the diagnosis of dilating cardiomyopathy. The most important features of this disease include:

- dilatation of the left ventricle, alone, or in connection with dilatation of the left atrium and right compartments of the heart.
- diffuse impairment of the kinetics of the left ventricular walls (sometimes the same can be applied by the right ventricle)
- significant reduction of systolic cardiac function
- formation of thrombi in hypokinetic compartments of the heart with subsequent embolization
- impairments of cardiac rhythm which may be of malign character (ventricular tachycardia, ventricular fibrillation)
- heart failure

The given set of signs originates in consequence of primary impairment of cardiac contractility. In consequence of impaired contractility the pumping function of the heart decreases. The ejection fraction decreases from normal 60–70% to extremely low values – often just 15–20% and first symptoms of left ventricular failure appear. The right ventricle can fail in consequence of primary failure of the left ventricle by propagation of pressure into the pulmonary artery, or it can fail at the beginning of the disease simultaneously with the left ventricle. In sporadic cases the left ventricle begins to fail as first.

Marked dilatation of ventricles yields mitral or tricuspidal insufficiency in consequence of dilatation of the left atrioventricular valve annulus. These valvular defects lead to further volume overload of the heart and enhance heart failure.

Thrombi originate in any of dilated heart compartments in consequence of stagnation of the blood. They may tear off and cause embolism in the arterial bed (in case of their left heart origin) or pulmonary bed (if they come from the right heart compartments). Consumption of ATP increases in the dilated ventricles with increased wall stress, and ectopic foci of electric activity originate. In this way extrasystoles, tachycardia, resp. atrial or ventricular fibrillations occur. In consequence of fibrotic changes