iterations were formed millions of years ago, the civilized world brings about changes in material and spiritual world often during one or several generations. Human biosystem cannot always manage to adapt to these facts. The alarm reaction represents in fact an unconditioned automatic reflex. It functions immediately unerroneously, stereotypically. However, it has a disadvantage in lacking plasticity and ability of modification, resulting thus in loosing its justification. The stressor evokes a reaction according to experience gained during millions of years. Today majority of stressors do not entail fight, but flight. The majority of stress reactions are not linked to muscular work. In spite of that human organism mobilizes catecholamines, glucocorticoids, and other stress hormones. The blood pressure elevates, glucose and fatty acids are mobilized, blood circulation is rebuilt, sodium is resorbed, potassium and magnesium decrease, blood clotting increases. What is mobilized is not subsequently utilized by muscular work.

These facts result in inadequately prolonged increased blood pressure, which gradually becomes permanent and primary hypertension develops. Hypertension and hyperlipemia may cause accumulation of lipids in vascular walls and development of atherosclerosis.

3.11 Congenital heart diseases

Congenital cardiovascular malformations result from an abnormal embryonal development of the normal structures or its absence. They develop due to genetic causes and the effect of the external environment, which is affecting the growing embryo between 3–7 week of justation. Those factors are (ionizing radiation, some chemical substances, pharmaceutics, viruses).

Cardiac and large vessels malformation have different clinical manifestations. Some disorders are well tolerated by the body. Yet during adulthood the haemodynamic situation may deteriorate. Some malformations are manifested late in the 4th or 5th decade of life. An example of this are shunts, in these cases problems appear only when the pulmonary hypertension is stabilized due to the structural changes in the lung field. Pulmonary hypertension accompanies more than one cardiovascular malformation. The state of pulmonary area or (the lungs) will decide the intensity of clinical manifestation of the disease, as well as the possibility of a surgical treatment. In the beginning the pressure in the pulmonary field depends on the pulmonary blood flow. Later it depends as well on the vascular resistance, finally there will be some structural changes in the lung field. That is why it is very important to measure the blood flow into the lungs as well as the pulmonary vascular resistance.

As an attempt to compensate for oxygen insufficiency and in chronic hypoxia here will be increment in the number of erythrocytes in the peripheral blood. That is why here will be an extreme increase in the haematocrit. As a result of this there will be a change in blood viscosity. Increase in the erythrocyte count (polycythaemia) is a cause of hypervolaemia. Polycythaemia, a high haematocrit, and hypervolaemia accompany congenital cyanotic heart diseases. On one hand by the effect of these changes it is easier for the blood to carry oxygen due to high blood capacity for oxygentransport, but on the other hand these changes give many side effect. There is the occurrence of thrombotic complications and hemorrhages. That is why drugs that potentiate vascular thrombosis are strictly contraindicated in patients with congenital cyanotic heart diseases. In cases of a very prominent polycythaemia we have to reduce the erythrocyte number by replacement with plasma or albumin to decrease the blood viscosity, increasing blood flow through the tissues, and hence provide more oxygen supply. Yet repeated venepunctions are not indicated because by this the organism is losing too much iron and to compensate the erythrocyte loss they are replaced by fast formation of small erythrocytes - microcytes. They have a lower defearbility, and hence are not good for oxygen transport.

The inborn incorrected cardiac diseases are a risk factor in females in time of gravidity and during birth. During gravidity the condition deteriorates in case there is already presence of pulmonary hypertension. Mortality rate is increasing in the group of females, in whom gravidity is terminated by cesarean section. In mothers with corrected cardiac
malformation or with artificial valves there is a tendency for the development of thrombotic complications. But the possible anticoagulative treatment is dangerous for the fetus. Most commonly it is the threatening of possible intra cranial hemorrhage during birth. In the first two trimesters of gravidity the anticoagulative treatment can lead to abortion.

**Surgically corrigated congenital malformations** of the heart are accompanied by the risk of developing infective endocarditis. In later stages post surgically, many examination methods and some surgical attempts may also lead to infective endocarditis. This is why in these patients antibiotics are given preventively during procedures like cystoscopy, proctosigmoidoscopy, in gastrointestinal, surgical and stomatological procedures.

The congenital cardiovascular diseases are classified by many ways. Commonly according to haemodynamic changes, anatomical changes, or according to clinical manifestations. The used classification is that one which divides the malformation into cyanotic and non cyanotic. The non cyanotic group is further classified into two types depending on the presence or absence of left to right shift.

The presence of cyanosis and the direction of the shunt are in dynamic development. Cyanosis should be absent from the beginning and shunt can occur later on due to morphological changes in the heart and vessels which are caused by the primary defect. That is why we will not consider the last type of classification so that we can concentrate on the effects of the malformation on the heart, vessels, and the circulation.

### 3.11.1 Atrial septal defect (ASD)

Corresponds for a certain type of communication between the atria (see fig. 3.14 on page 146).

It is often discovered during examination of adults. It is more common in females. The atrial septal defect can be in the upper or lower part of the septum. Here it can be associated with other defects. The atrial septal defect represents a state where foramen ovale is closed only functionally from the embryonal period. It is known as foramen ovale persistens. In some cases it is closed anatomically but in such a way that there will be an opening which remains and this opening is of different sizes. This case is known as foramen ovale apertum. The atrial septal defect are markedly variable. The foramen ovale apertum or persistens can combine with a defect of the neighboring part of the septum. The size of the defect and the pressure changes in the atria will determine the amount of blood flowing from the left to the right atrium. In physiological state the pressure in the left atrium is higher than the pressure in the right atrium. So the determining factors of the size of (left to right shunts) are the amount of the blood flow, the pulmonary resistance, and the state of both atria. The flow of blood through the defect in the atrial septum during diastole, leads to an increment of the filling of the right ventricle and hence an increment of the blood flow through the pulmonary area. More blood flows to the left atrium yet once again part of it is transferred to the right atrium via the defect and is recirculated through the lung field. In long lasting and markedly increased pulmonary blood flow there will be an increased arterial vascular tonus of the lung field. So the arterial blood pressure is gradually increasing in the pulmonary artery and as a consequence of this there will be right ventricular hypertrophy, then to an increment in the connective tissue and finally even the formation of atherosclerosis in the small circulation. These organic changes result in further increase of the arterial wall resistance in the lung field and hence progression of pulmonary hypertension and right ventricular hypertrophy. Those patients are not symptomatic from the
3.11. Congenital heart diseases (I. Hulín, L. Zlatoš)

beginning. Usually there are some respiratory infections. In older ages there are cardiorespiratory problems. Later there might be atrial arrhythmias and pulmonary hypertension. When there is hypoxia due to other causes than cardiac ones, pulmonary hypertension develops much earlier.

An increased diastolic filling of the right ventricle can be the cause of the accentuation of tricuspid closure sound. Second heart sound is usually doubled. Commonly there might be some rhythm disturbances which arise in the atria for example atrial fibrillation. Right ventricular hypertrophy can present in children.

The abnormality can be corrected in optimal situations between 3rd and 6th year of life. Good results are achieved before the age of 40, but there should not be pulmonary hypertension.

3.11.2 Ventricular septal defect

It usually occurs in combination with other abnormalities (see fig. 3.15 on page 147).

![Figure 3.15: Defect of ventricular septum](image)

It is usually localized in the membranous or muscular part. The resulting situation depends on the size of the defect and on the state of lung field. Very small defects can close spontaneously. Larger ones can be fatal due to cardiac decompensation in young age. In early neonatal period there is usually heart failure because neonatal myocardium cannot tolerate a haemodynamic load. It is the result of immature intracellular structures of cardiomyocytes (myofibrils, mitochondria, sarcoplasmic reticulum) and hence a low functional capacity of their myocardium. Another factor of the low capacity of their myocardium. Another factor of the low capacity of a newborn myocardium to tolerate high working demands is that the heart rate and the end diastolic volume are both markedly high even at rest. In a long lasting defect there might be a development of pulmonary hypertension. In cases of large defect with pulmonary hypertension there is a large risk of possible development of an anatomical obstruction of pulmonary vessels. This condition is irreversible. That is why the correction of this problem is limited by the reconstruction of the pulmonary vascular field. Pulmonary vascular obstruction is manifested by dyspnoe, retrosternal pain, syncope and hemoptysis. In normotensive pulmonary blood pressure the VSD (ventricular septal defect) carries very good prognosis even without correction.

3.11.3 Patent ductus arteriosus

During the fetal period ductus Arteriosus forms a normal junction between aorta and the pulmonary artery. (Ductus arteriosus in the fetal period is a junction which arises from the pulmonary artery bifurcation). During this period blood flows form the pulmonary artery to the aorta. This duct is closed immediately after birth. The closure occurs as a result of a change in the pressure relations and partly due to the active contraction of this duct which follows changes in oxygen saturation in the blood. In cases of a patent ductus arteriosus which persists even after birth the blood flows form aorta to the pulmonary artery. This flow is continuous during the whole cardiac cycle (see fig. 3.16 on page 148).

The blood flow is faster during systole. This is why we can usually hear a very typical systole-diastolic murmur which is marked as a machinery or continuous murmur. When the amount of blood flowing through the duct is large, pulmonary hypertension and shunt would be later irreversible (hypertension is fixed by fixed vascular obstruction) and more over becomes reversed. Blood then flows from the pulmonary artery back to the descending aorta. Cyanosis is noticed in the lower limb but not in the
upper limbs (hands). This case eventually leads to cardiac decompensation. A fatal end could be caused by rupture of the duct and its aneurysm, dilatation, or calcification.

### 3.11.4 Anomalies of coronary artery emergence

The left coronary artery emerges from the pulmonary artery. Here the myocardial perfusion is supplied by the right coronary artery. Collaterals develop between the two arteries. Never the less there will be myocardial fibrosis. Myocardial infarction is not unusual. Only 20% of the affected children reach the age of adolescence. In cases of ischemia or repeated myocardial infarction there will be papillary muscle dysfunction. Papillary muscle dysfunction eventually leads to mitral regurgitation. Surgical correction is possible. The prognosis are dependent upon the extent of the myocardium which is already injured by hypoxia.

Sometimes there might be an abnormal communication between the coronary artery and one of the ventricles or the right atrium. This is called arterial-venous coronary fistula. The shunt is usually tiny. There might be a potential thrombus formation or there might be a development of aneurysm. In some cases pulmonary hypertension or cardiac decompensation might develop.

### 3.11.5 Congenital aortic stenosis

We can divide aortic stenosis into valvular aortic stenosis supravalvular aortic stenosis and discrete sub aortic stenosis.

**Valvular stenosis.** In this congenital deformity the aortic valve is actually formed of two crescentic valves of irregular shape. Only later the valve is narrowed, stenozed. Sometimes it is very hard to distinguish this case from the rheumatic or degenerative stenosis. Stenosis leads to concentric hypertrophy of the left ventricle and a post stenotic dilatation of the ascending aorta. Stenosis result in a high pressure gradient between the left ventricle and the aorta. Patients may remain asymptomatic for a long time. If stenosis develop suddenly the patient complains of weakness, syncope, and myocardial ischemia. On auscultation we find a systolic murmur which propagates to the carotids. The bicuspid aortic valve is a very favored place for the colonization of bacterial endocarditis. That is why stenosis can be later on combined with aortic regurgitation of blood.

**Supravalvular stenosis.** Is narrowing of aorta at the upper edge of the semilunar valves. In usual cases the site of coronary arteries emergence is not affected.

**Subaortic stenosis.** Is already present at birth. There is a fibrous ring situated immediately below the semilunar valves. The ring can have a membranous character.

### 3.11.6 Coarctation of aorta

Presents a narrowing or asphyxiation of the aortic lumen. That might occur anywhere along the aorta. The narrowing is usually distal to the branching of the left subclavian artery near the insertion of ligamentum arteriosus. It is more common in boys, in whom we quite often find gonadal disgenesis. The haemodynamic state depends on the extent of the narrowing and on the presence of other anomalies. In small coarctation the defect might be symptomless. More prominent coarctation can result in frequent headaches, epistaxis, cold extremities, and claudication on exertion, hypertension in the upper limbs.
and a weak pulsation of the femoral artery. Upper limbs and chest is more developed in comparison with the lower limbs. It is the result of the haemodynamic state. We can hear a systolic murmur on the lateral thoracic wall. There are many collaterals which are compensatory dilated. The left ventricle hypertrophies. Aorta dilated above and below the site of coarctation. A non corrected aortic coarctation is the cause of a marked hypertension, which might lead to cerebral aneurysms, hemorrhages, aortic ruptures, left ventricular failure and infectious endocarditis. Surgical correction in form of resection of the affected site (site of coarctation) and end to end anastomosis, will relief the haemodynamic exertion on the heart but not necessary remove the hypertension. The reason of hypertension can be due to a haemodynamic obstacles that were present before the correction, as well as the participation of renin angiotensin system which resulted from the low blood flow through the kidneys. Prior to surgical correction the hypertension presents only in the upper part of the body. Collaterals are palpable between the scapulae, and on the sides of chest. They are more prominent on bending. Coarctation without a prominent hypertension turns to be normotensive after correction. In 80% of the corrected cases there will be development of hypertension.

3.11.7 Pulmonary artery stenosis

When it occurs individually without other malformations it is usually localized in supravalvular, valvular, and subvalvular position. The most common position is the valvular localization. In case of normal right ventricular function there will be a high pressure gradient between the ventricle and pulmonary artery. It reaches 50–80 mm Hg. The progress of this disorder and patient’s symptoms are dependent upon the extent of the stenosis. Usually there is syncope, weakness, dyspnoe and right ventricular failure. In cases of severe stenosis the pressure in the right ventricle might become equal to the level of pressure in the left ventricle. The ejection time of the right ventricle is prolonged. The right ventricle is then markedly hypertrophied. Yet, this hypertrophy decreases the compliance of the right ventricle, and consequently worsens the right ventricular filling. The stenosis is the cause of a systolic murmur. Tricuspid regurgitation might result from a progressed stage of stenosis the main cause is the pulmonary stenosis with hypertrophy and affected fibrous ring of the heart. The right ventricular failure and the right atrial dilatation are both outcomes of a haemodynamic overload on the right ventricle.

3.11.8 Transposition of the great vessels (arteries) TGA

Occurs in combination with other abnormalities. The aorta arises from the right ventricle and the pulmonary artery from the left ventricle. In such cases there is usually a patent ductus arteriosus (PDA) or a ventricular septal defect (VSD). Otherwise this case (Transposition of the great arteries) is incompatible with life. The whole condition depends upon the ventricular function and the state of pulmonary (lung) field. It also depends on whether there is enough oxygen in the blood of the coronary arteries.

3.11.9 Ebstein anomaly

The tricuspid valve is absent, dysplastic, and pushed into the right ventricle, it is partially adherent to the right ventricular wall. This is why actually the right ventricle is small compared to the large right atrium. In this case there is a considerable tricuspid regurgitation. There might be a presence of tachyarrhythmia.

3.11.10 Tetralogy of Fallot

The classical picture of tetralogy of Fallot represents the simultaneous presence of major and basic changes which are: pulmonary stenosis, ventricular septal defect, transposition of the aorta, and a right ventricular hypertrophy (see fig. 3.17 on page 150). The pulmonary stenosis can be valvular or infundibular type. The dextroposition of the aorta can be partial or complete. The increasing right ventricular pressure during systole leads to right ventricular hypertrophy. The increasing pressure is the consequence of the communication between both ventricles and pulmonary artery stenosis. The pulmonary stenosis can occur together with stenosis of the pulmonary artery branches. The blood flows to the pulmonary vasculature instead of flowing to the aorta. The right to left shunt is the cause of hypoxemia and cyanosis, finger clubbing, and polycythaemia. Cyanosis doesn’t manifest itself from the very beginning of this anomaly. If cyanosis doesn’t occur
shortly after birth, it usually appears before the first year of life. The explanation of this phenomena is that ductus arteriosus remains patent for few months after birth (its closure is opposed by the pressure gradient). The limiting factors of the proper haemodynamics are the pulmonary stenosis in the first place and the size of ventricular septal defect in the second place. The blood flow across the pulmonary artery can represent only $\frac{1}{3}$ the blood flow in the aorta.

When Fallot tetralogy is associated with an atrial septal defect we are dealing with a case of pentalogy. The child becomes cyanosed during feeding and when crying. The child will be become bluish, dyspnoic, spasms may occur, and there might be loss of consciousness. Later in the child development we notice a marked loss of performance and frequent resting. There is a retardation of growth. There will be what is know as cardiac nazism with infantilism. An increased blood viscosity and hypoxia are helping factors in the occurrence of thrombi, especially in the infective endocarditis.

In those children the precordium is usually prominent and we can palpate a right ventricular heave. We can hear a harsh systolic murmur in the left parasternal area in the second and forth intercostal spaces. The second heart sound above the pulmonary artery can not be heard.

### 3.12 Infective endocarditis

When the endocardium is colonized by microbes the resulting disease is known as infective endocarditis. The most significant pathological changes are taking place on the edges of valves. There is some distraction which is mainly shown as changes in the shape of the valves and formation of vegetation. Infective endocarditis is the new term of what is previously known as bacterial endocarditis. the main reason of the new nomenclature is that this disease can be caused by fungi or chlamidia. From the clinical point of view we are usually talking about acute and sub acute bacterial endocarditis, valvular endocarditis, and valvular endocarditis occurring after valve transplantation. Non bacterial thrombotic endocarditis is an individual clinical unit.

Infective endocarditis is most commonly (80%) caused by streptococci, staphylococci. *Streptococcus a-haemoliticus viridians* is the dominant bacteria causing infective endocarditis. It forms 65% of cases. Infective endocarditis can occur following many various infective diseases. The can even form complexes of diseases such as pneumococcus pneumonia, meningitis, and endocarditis, this triad is called Austrian syndrom.

Valvular defects were the previously dominating problem in the developed countries, those defects were the results of a chronic rheumatic process. Meanwhile these causes are in regression. The problem now is infective endocarditis which result from some predisposing factors like some congenital diseases and mainly some discrete changes being for example mitral valve prolapse. Another predisposing factors are some cardiosurgical procedures. Of course every overcomed or treated case of infective endocarditis carries a potential danger of recurrence. Many factors predispose to the occurrence of infective endocarditis. Bacteriemia as such being (the presence of pathological microbial agents in the blood) is not enough to cause infective endocarditis. The first condition for the occurrence of this