sis. Dysphagia is due to the compression and infiltration of oesophagus. Paresis of the phrenic nerve with elevation of the diaphragm can lead to dyspnoea. Involvement of pericardium can be manifested by arrhythmias or heart failure.

Carcinomas, invading the pleura from the apical parts into the upper thoracic aperture are the cause of the **Pancoast's syndrome**. They infiltrate the brachial plexus, the thoracic and cervical parts of sympathetic nervous system, the ribs and vertebrae. A cruel pain in the shoulder is present, later paresis of the hand, with muscle atrophy, loss of sensibility and the Horner's syndrome (constriction of the pupil, ptosis, anhidrosis on the affected side of the head and neck) occur.

The compression of superior vena cava leads to **the vena cava superior syndrome** development. Cyanosis and oedema of the head and neck are present, accompanied with oedema of upper extremities and the trunk. The veins are congested and winding. The patient suffers from headache and disturbances of vision.

The extrapulmonary symptoms form the **paraneoplastic syndromes**. They are usually present in bronchogenic carcinoma, yet they occur also in other malignant tumours. Their cause is not well known. Sometimes they precede the primary tumour, or they disappear following its removal, and during the relapse they reappear. The **manifestation of the bone and joint involvement** includes the clubbing and hypertrohic osteopathy with pain in joints imitating the rheumatoid arthritis. Pains in long bones, profuse sweating and myastenic syndrome are also present. Peripheral neuropathies (manifested by paresis of the fibular nerve) are observed. Sometimes also **endocrine disturbances** are present. Cushing's syndrome develops due to extreme hyperproduction of a peptide similar to ACTH. Because of overproduction of a peptide imitating the effects of pituitary hormones which stimulate the ovaries, gynecomastia develops. Hyponatriaemia is due to the production of a peptide with effects of ADH. The cancer cells often produce calcitonin. The endocrine changes can be associated with the resemblance of the cancer cells with those of the APUD system.

Phlebitis migrans, anaemias, leucocytosis and thrombopenia are commonly occuring **extrapulmonary manifestations**. A nonbacterial thrombotic endocarditis can develop. The prognosis in patients with bronchogenic carcinom is in spite of application of the most proven therapeutic methods not satisfactory. Since the diagnosis has been established merely 10 per cent of patients survive the period of five years.

Sarcoma of the lungs. It is a rarely occuring malignant tumour. It occurs in older persons. The prognosis is bad.

1.11.3 Metastatic malignant tumours of the lungs

Metastatic malignant tumours of the lungs constitute 20–30 per cent of all malignant tumours. Direct penetration of tumours arising from the adjacent organs (breast carcinoma, tumours of the thymus, of the thyroid gland, oesophagus and tumours of the mediastinum) can be involved. Osteosarcoma, malignant tumours of testes, kidneys, suprarenal glands, stomach, large intestine, prostata and breast metastasize into the lungs by blood vessels.

Solitary metastases are not accompanied with clinical symptomatology. Clinical manifestations of **disseminated** metastases include an irritant cough and progressive dyspnoea. Solitary metastases are resected. The patients with disseminated metastases are treated by chemotherapy or radiotherapy. The prognosis is bad.

1.12 Disorders of the diaphragm, thorax, pleura and mediastinum

1.12.1 Disorders of the diaphragm

The diaphragm is the most important respiratory muscle. It separates *hermetically* the thoracic cavity from the abdominal cavity. The central part is tendinous and not contractile. The peripheral part consits of muscle fibres attached to the distal osseous part of thorax. Some formations and structures pass the diaphragmatic openings from the thoracic into the abdominal cavities. The muscles are governed

by the phrenic nerve, the motor neurons originate in the cervical part of the spinal cord from segments C3–C5. During the contraction of diaphragma the organs in the abdominal cavity are displaced downward. This creates a negative pressure in the thoracic cavity during the inspiration. The function of diaphragm has to be perfectly coordinated with the function of other respiratory muscles. In addition, the diaphragm has an extraordinary importance in non-respiratory functions (delivery, defecation, speech). The diaphragmatic muscles are very well supplied with blood.

The dysfunction of diaphragm occurs during hyperinflation of the lungs. The hyperinflation can be of acute character e.g. in the asthmatic attack, or permanent in chronic obstructive ventilatory disorders. Hyperinflation inhibits the necessary movements of the diaphragma. In this situation the diaphragm is not able to fulfill the reqiurements of increased ventilation of the lungs. During forced breathing, paradoxical movements of anterior abdominal wall can be observed (the abdominal paradox).

Sometimes **unilateral diaphragmatic paralysis** can develop. It usually occurs in phrenic nerve affliction, e.g. in bronchogenic cancer, poliomyelitis, encephalitis and herpes zoster. Paralysis of the phrenic nerve can develop also after a cardio-surgical intervention. Paralysis of the diaphragm can be caused by aortal aneurism, substernal goitre, trauma of the thorax and spine (bilateral paralysis). Muscle dystrophies and myelopathies affect also the muscles of diaphragm. During diaphragmatic paralysis dyspnoea or its deterioration are observed.

A rapid, sudden contraction of the diaphragm associated with a reflexive closure of glottis and accompanied with a sound phenomenon is called **singultus** (hiccup). It is a reflex; the afferent impulse of which originates in the sensitive fibres of the phrenic or vagus nerves. It leads to the respiratory centre in the brain stem. The efferent pathway consists of the phrenic nerve motor fibres leading to the diaphragm. The peripheral hiccup arises due to irritation of the phrenic nerve, in disorders of mediastinum, lungs, heart, oesophagus, diaphragm, or diseases localized in the abdominal cavity, and after surgical interventions. Central hicccup occurs in encephalitis, meningitis, stroke, brain tumours, uraemia and alcoholic intoxication.

The hiccup can be elicited also by psychogenic stimuli. A transitory hiccup can be caused by a cold drink, intake of greater volume of food, concentrated alcohol, but also by a cold shower and other non – specific stimuli.

Traumatic rupture of diaphragm results in displacement of abdominal organs into the thoracic cavity. Also the inborn diaphragmatic defects may be involved.

1.12.2 Disorders of the thorax configuration

Disorders of the thorax configuration lead to very similar consequences, concerning the respiratory functions. They are especially:

1. alveolar hypoventilation
2. changes in the compliance of thorax
3. various degrees of lung compression
4. irregular ventilation-perfusion ratio
5. pulmonary hypertension and cor pulmonale

Clinical symptoms include dyspnoea without cough, sputum and chest pain.

Kyphoscoliosis. It is a deformity of the spine in two directions. **Kyphosis** is a pathologic backward deformity of the spine in the sagittal plane. **Scoliosis** is a lateral curvature of the spine in the frontal plane. It is always associated with rotation of vertebrae around the long axis. Kyphosis is important regarding the respiration, if the deformity of the spine exceeds the 70°C angle. Scolioses are classified as being either functional or structural. Functional scolioses are due to the shortening of the lower extremity, can be postural and hysterical. The structural scolioses include the idiopathic, neuromuscular, congenital forms and scolioses due to other diseases (neurofibromatosis, rheumatic diseases, disorders of the mesenchyma, posttraumatic scolioses). **Idiopathic scolioses** constitute more than 80 per cent of all scolioses. The children's scoliosis is found in children below the age of 3 years, the juvenile scoliosis below the age of 9 years and the adolescent scoliosis before the end of growth. The main problem regarding the respiratory functions arise from the rigidity of the thorax. During infection hypoventilation and

hypoxaemia can impair intensively. The progression of the disease leads to the cardiopulmonary failure.

Pectus excavatum. It is a congenital deformity of the distal part of the sternum. Sternum is impressed into the thorax, especially in its distal part. This disorder (a tunnel-shaped thorax) is often associated with a number of other abnormalities and with high mortality. It is often combined with the mitral valve prolapse. Deformations of a lesser degree do not cause troubles and have rather the character of cosmetic defects.

There are still other deformations of the thorax, disturbing to different degree the function of respiratory organs. In ankylosing spondylitis the mobility in the costovertebral joints is reduced. Deformities may occur after traumatic injuries and after disorders of the pleural cavity.

1.12.3 Disorders of the pleura

Pleura is a semitransparent membrane which consist of a network of fibroelastic tissue, lymphatic and blood vessels. It contains a layer of mesothelial cells of different size. The mesothelial cells have their surface fine cilia-microvilli. The surface of mesothelial cells is lined with fine cilia-microvilli. The mesothelial cells produce glycoprotein rich in hyaluronic acid. This facilitates the sliding of pleural surfaces over each other. **Parietal pleura** covers the surface of the chest wall, diaphragm and mediastinum. It is supplied with blood from the systemic circulation. It contains sensitive nerves and cells with a dense ciliary layer. **The visceral pleura** covers the surface of lungs, including the interlobar fissures. The visceral pleura is supplied with blood from the pulmonary circulation. It does not contain sensitive nerves and the cells have few cilia. Boths pleural surfaces closely adhere to each other. The pleural space contains only 5–10 ml of fluid enabling the lungs *to follow* the movements of the thorax during inspiration and expiration. **The pleural fluid** contains, under physiological conditions, few proteins (less than 2 g/100 ml). The level of glucose and pH are identical with those in the plasma. The hydrostatic pressure in the parietal pleura is identical with the pressure in the systemic circulation and that in the visceral pleura is identical with the pressure in the pulmonary circulation. The parietal lymphatic system serves as a draining system for the fluid of pleural space (see the figure 1.4).

The pain arising from pleura is always unilateral. It spreads into the arm and neck, or into the abdomen. Dyspnoea can be present because of the lung compression, due to accumulation of fluid in the pleural space. The fluid in pleural space does not have to be detected by the roentgenographic imaging in the back to front projection in a recumbent patient (e.g. at an intensive care station).

The pleural fluid examination has a great diagnostic importance The thoracocentesis has only few contraindications (haemorhagic diatheses, anticoagulant therapy and mechanical ventilation). The examination of the pleural effusion detects the total content of proteins, lactate dehydrogenase, the leucocytes count and their differential count, the level of glucose and pH. The analysis of the pleural effusion attests these facts :

1. ratio of the proteins in the pleural fluid and in serum is above 0.5

2. ratio of LDH in pleural liquid and in serum is above 0.6

The pleural fluid is examined for the presence of malignant cells, pus, microorganisms, erythrocytes and also other substances according to the assumed cause of the effusion. So e.g. the ratio of the creatinine level in the pleural fluid and in serum is in urinothorax above 2. The level of glucose is below 3.33 mmol/l in infections, rheumatoid pleuritis and tuberculosis. A decrease in pH below 7.2 is observed usually in emphysema, in malignant processes, tuberculosis and pleuritis. Polymorphonuclear leucocytes signify very probably a bacterial infection. Lymphocytes are present in tuberculosis, yet also in lymphomas and in leukaemic effusion.

Transudation into the pleural space. Transudation, in general, is a state, when the leakage of fluid into the serous cavities is evident. **The cause of transudation is the change in osmotic and hydrostatic pressures**. In heart failure the volume of fluid in the pleural space increases owing to the venous pressure elevation, especially in the failure of both heart ventricles. Changes concerning filtration and reabsorption of fluid in the capillaries are involved. The process of filtration continues normally, but the reabsorption is retarded due to the increased pressure in the venous end of capillaries. The increase is not caused by heart failure, i.e. by the inability of heart

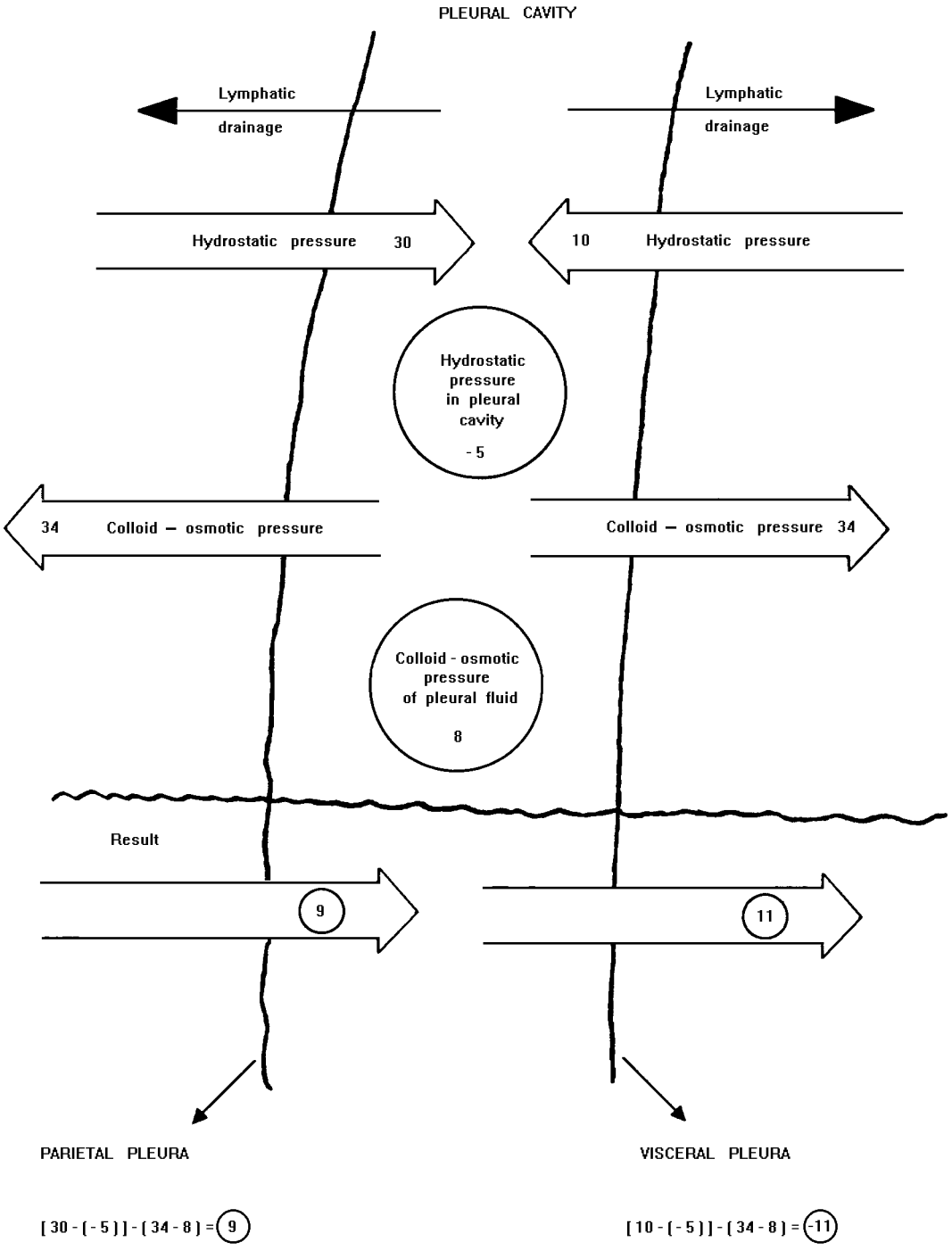

Figure 1.4: Pressure relations in pleural cavity

to pump the necessary "*offered*" volume of the venous blood from the venous part of circulation into the arterial part. Heart failure leads within several months to an increase in the content of proteins in the pleural fluid above 3 g/100 ml. Transudation into the pleural space is observed also in hepatic cirrhosis. The ascitic fluid can get through the diaphragmatic defects into the pleural space or it can be removed by the lymphatic system and "trasferred" from the abdominal to the thoracic cavity. Transudation in nephrotic syndrome is due to very low oncotic pressure. Peritoneal dialysis can also be the cause of fluid transudation into the pleural space.

Exudation into the pleural space. Exudation is a state, with an evident production and accumulation of fluid in the serous cavities. **The cause of exudation resides in the serous membrane**. The exudation is induced by neoplasma, infection, inflammation or trauma. In the process of exudation the mesothelial cells *play an important role*. Exudation is often observed in pneumonias and pulmonary abscesses. Uncomplicated exudation can be effectively treated by antibiotics. The count of leucocytes in the pleural fluid is not too high, pH usually does not exceed 7.3 and LDH is below 0.8 μkat/l. In complicated cases, the number of polymorphonuclear leucocytes is higher than 100 000/μl and pH is below 7.2, the level of glucose does not reach 2.2 mmol/l, LDH is above 16 μkat/l. The exudation contains bacteria. The pleural sepsis is a very severe complication. Exudation into the pleural space occurs very often in pulmonary tuberculosis, yet it occurs also during other infections.

Haemothorax is usually the complication of a traumatic injury of the thorax. The pleural space contains bloody effusion with haematocrit exceeding 20 per cent. It can occur in disorders of blood coagulation, in pleural malignant processes and due to rupture of aortal aneurysm. The blood in the pleural space does not coagulate. It is relatively quickly eliminated by the lymphatic system.

Chylothorax is a state, when chyle accumulates in the pleural space. It is usually associated with malignant processes in the thoracic cavity, e.g. lymphomas, and with disorders of lymphatic vessels.

Pleural exudation can develop also **in other diseases**. In about a half of patients with rheumatoid arthritis in various stages pleural exudate is observed. Sometimes it precedes the manifestation of rheumatoid arthritis. In such cases the exsudate contains a low level of glucose (less than 1.66 mmol/l), high level of LDH and low pH. The clinical manifestation is not very remarkable. Increased body temperature does not have to be present. The condition may be complicated by formation of fibrotic tissue connecting both pleural surfaces.

Pleura is *affected* also in other diseases. Fluid can appear in the pleural space especially in consequence of lung diseases. In asbestosis an unilateral serous-sanguinolent exudate can be present. The number of polymorphonuclear or mononuclear leucocytes is increased. Uraemia leads to development of polyserositis. The pleural lesion in uraemia displays a picture called urinothorax. This condition must be distinguished from the hydrothorax, which develops during the nephrotic syndrome without the renal failure.

Frequent causes of the pleural exudation forming are the malignant processes. The breast carcinoma and carcinoma of the lungs, more rarely those of the ovary and stomach penetrate the pleural space or metastasize in it. Patients can be asymptomatic, or they suffer from cough, pain in the chest and dyspnoea. The exudate usually contains erythrocytes and mononuclear leucocytes (lymphocytes more than 50 per cent). Glucose is lower than 3.33 mmol/l. The pH is lower than 7.3. The exudate contains malignant cells. Pleural exudate if present in malignities has a very unfavourable prognosis. It occurs also in non-Hodgkin's lymphomas. Moreover, if a mediastinal invasion is present, lymphatic blockade can develop. In 80–90 per cent of patients with manifested asbestosis malignant mesothelioma of pleura develops. Mesothelioma is accompanied with dyspnoea, cough, weight loss and chest pain. The tumour spreads diffuselly from the pleura into the lungs. The pleural exudate is abundant. The cytological identification of mesothelioma is not unambigous and so is the differential diagnosis between the benign and malignant forms of mesothelioma. Benign mesothelioma has the character of fibrosis, reaches great dimensions and is usually combined with a hypertrophic pulmonary osteoarthropathy.

Pneumothorax.Pneumothorax is characterized as air accumulation in the pleural space. It can occur due to:

1. perforation of visceral pleura and the leakage of air from the lungs

2. penetration of thoracic wall, diaphragm, mediastinum and oesophagus

3. production of gas by microorganisms in empyema

The perforation of visceral pleura can occur also in healthy young people without any disorder of the respiratory organs. Most commonly the rupture occurs in the apical parts of the lungs. Acute chest pain, dyspnoea and cough are the main symptoms of pneumothorax. In 7–14 days the air can be spontaneously reabsorbed.

Pneumothorax can arise due to traumatic injury of the thorax or pulmonary diseases. The injury of the thorax enables in some cases the air to enter the pleural space during the inspiration. This condition is termed the valvular pneumothorax. Pneumothorax can be partial or complete. In emphysema the rupture of a subpleural bulla can easily occur. Also the rupture of pulmonary abscess can occur. In such cases pyopneumothorax develops. Pneumothorax can arise also during an asthmatic attack, in interstitial diseases of the lungs and in neoplasmas. At intensive care stations iatrogenic pneumothorax may occur caused by barotrauma during mechanical ventilation or during introduction of the caval catheter. A severe complication is the pneumothorax associated with bronchopleural fistula.

1.12.4 Disorders of the mediastinum

Mediastinum is an anatomical space in the centre of the thorax, which separates both pleural spaces. It is limited by the diaphragm and the upper thoracic aperture. In the small mediastinal space some important vital organs are localized. That is why the changes in mediastinum are manifested by marked symptoms. **The anterior compartment** of mediastinum is limited by the pericardium, ascendent aorta, brachiocephalic trunk and sternum. Herein are situated: the thymus, substernal part of the thyroid gland, and the parathyroid glands, vessels, pericardium and lymph-nodes. **The midle compartment** is limited by the anterior compartment, and at the back by the pericardial line. It contains the heart, large vessels, trachea, the main bronchi, lymph nodes, and the phrenic and vagus nerves. **The posterior compartment** reaches from the posterior line to the back wall of the thorax. Here are situated the vertebrae, descending aorta, oesophagus, thoracic duct, the azygos and hemiazygos veins, the lower portion of the vagus nerve, the sympathetic ganglia, and mediastinal lymph-nodes (see fig. 1.5).

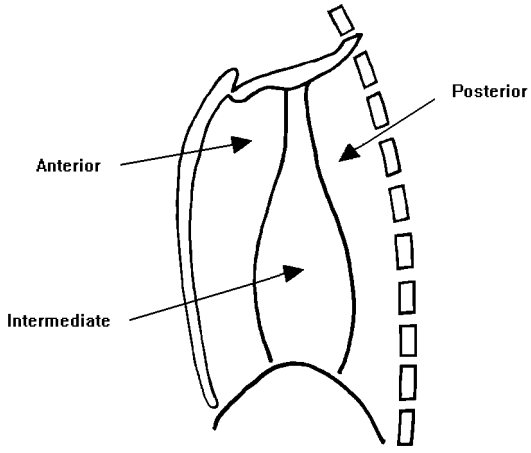

Figure 1.5: Mediastinal compartments

The symptoms of expansive processes in the mediastinum include pain in the chest, cough, hoarseness and dyspnoea. Stridor, dysphagia and the Horner's syndrome occur rarely. About a half of the patients with myastenia gravis suffer from thymomas. Hypoglycaemia may occur in patients with mesothelioma, fibrosarcoma or teratoma.

Neurogenic tumours in the mediastinum lead to neurological symptoms. The tumours are represented mainly by benign tumours (neurofibroma, ganglioneuroma). They can produce hormones. Hypertension is usually present. Neuroblastoma (a malignant tumour consisting of sympathetic ganglia cells) has a little better prognosis than the neuroblastoma originating in suprarenal gland medulla. Neurogenic tumours manifest themselves by pain in the chest, unproductive cough and the compression of trachea.

Thymomas represent about 20 per cent of all mediastinal tumours. Thymomas are considered to be malignant tumours. Among the lymphatic tumours most frequently occur the Hodgkin's lymphomas.

The non-Hodgkin's lymphomas, plasmacytomas and hamartomas have the same clinical manifestation as other mediastinal tumours. Their prognosis is unfavourable. The intrathoracic goitre is accompanied with stridor, cough and dyspnoea. Pericardial cysts are usually situated in the anterior compartment of the mediastinum. The vascular tumours are usually represented by vascular hamartomas, lymphangiomas, haemangiomas. They are benign. Less frequent are the malignant liposarcomas, mesotheliomas, rhabdomyosarcomas and mesenchymomas.

Diaphragmatic hernias can be presented as expansion of the mediastinum. The clinical manifestation of the pneumomediastinum may be similar. It occurs usually in consequence of a rupture of the bronchi or oesophagus. An alveolar rupture and the subsequent pneumomediastinum can arise due to the mechanical ventilation.