Pulmonary arterial hypertension

Pulmonary arterial hypertension is increase in pulmonary pressure above norm.

- Systolic pressure > 35 mmHg,
- **Mean arterial pressure > 25 mmHg**,
- Diastolic pressure > 12 mmHg.

The value of **the mean pulmonary pressure** are significant in the diagnostic.

Pathological classification

náhled|upright=1.6|Video v angličtině, definice, patogeneze, příznaky, komplikace, léčba According to the value of mean pulmonary pressure, we distinguish between mild, moderate and severe pulmonary hypertension.

	mean arterial pressure (mmHg)	systolic pressure (mmHg)
norm	**< 25**	< 35
mild	**26–35**	36–45
moderate	**36–45**	**46–60**
severe	**> 45**	> 60

The values of the systolic pressure are not crucial for the diagnosis of pulmonary hypertension, nevertheless they can be useful in determining the diagnosis.

In terms of **etiology** we distinguish:

- primary pulmonary hypertension,
- secondary pulmonary hypertension.

Primary pulmonary hypertension

Primary pulmonary hypertension develops idiopathically (especially in younger women) with the insufficiency in the small arteries (typical plexiform lesions), the cause is apparently primary damage to the pulmonary endothelium.

- Survival usually doesn't exceed 3 years, dying of right heart failure.
- Among the symptoms dominates **progressive dyspnea** with peripheral cyanosis, chest pain, fatigue.

Secondary pulmonary hypertension

Secondary pulmonary hypertension develops on ground of another disease.

In terms of **hemodynamics and pathophysiology** we distinguish:

- precapillary pulmonary hypertension,
- postcapillary pulmonary hypertension,
- hyperkinetic pulmonary hypertension.

Precapillary pulmonary hypertension is characterized by normal values of **pressure in the wedge** (this is equal to the pressure in the left atrium). It typically occurs in lung diseases (COPD, pulmonary fibrosis, sarcoidosis, pneumoconiosis), chronic pulmonary embolism of primary pulmonary artery disease and after lung resection, or also in lung hypoventilation. This also includes vasculitis in systemic connective tissue diseases.

In **postcapillary pulmonary hypertension**, wedge pressure is always increased. The cause is in left heart disease (left heart failure, mitral stenosis, hypertrophic cardiomyopathy), constrictive pericarditis.

Hyperkinetic pulmonary hypertension is based on left-right heart shunts such as persistent *ductus arteriosus*, atrial or ventricular septal defects, or conditions with high cardiac output (e.g., hyperthyroidism).

Diagnostic classification

In terms of causes, prognosis and therapy of the disease (the so-called Venetian classification) we distinguish:

- pulmonary arterial hypertension,
- pulmonary hypertension in left heart disease,
- pulmonary hypertension in respiratory diseases or in hypoxemia,
- pulmonary hypertension in chronic thrombotic or embolic disease,
- pulmonary hypertension from other causes.

Pulmonary arterial hypertension

The cause lies in **the reconstruction of the walls of the pulmonary arteries**, especially hypertrophy of the smooth muscle of the tunica media, which leads to an increase in the resistance of the pulmonary arteries and thus to an increase in pressure in the pulmonary artery. The wedge pressure has normal values (up to 12 mmHg). In pulmonary arterial hypertension, **high values of mean pulmonary pressure** around 60 mmHg are usually, they can be close to the values in the systemic circulation. This is also related to the poor prognosis of the disease. Fortunately, the incidence of this disease is small (about 10 per 1 million inhabitants).

These include in particular:

- primary (idiopathic) pulmonary arterial hypertension,
- hereditary pulmonary arterial hypertension.

The choice of treatment is **pharmacotherapy** (see below).

Pulmonary hypertension in left heart disease

The most common type of pulmonary hypertension. In left heart failure, increased blood pressure propagates from the left atrium to the pulmonary circulation. Thus, both the mean pressure in the lungs and the pressure in the wedge (pressure in the left atrium) are increased. Typically in mitral stenosis, left heart failure. The development of pulmonary hypertension increases the mortality of patients with heart failure.

The treatment is **heart failure therapy**.

Pulmonary hypertension in respiratory diseases

The second most common type of pulmonary hypertension. The underlying cause of the disease is in the lung parenchyma and not in the lung vasculature. The wedge pressure is normal. Typically caused by COPD, interstitial pulmonary fibrosis, sarcoidosis. Pulmonary hypertension developing due to the aforementioned lung diseases is indicative of a really bad prognosis - increases mortality.

There is no special treatment, **the primary lung disease is treated**.

Chronic thromboembolic pulmonary hypertension (CTEPH)

A condition in which, despite adequate therapy, thrombi do not recanalize sufficiently after pulmonary embolism; on the contrary, they reorganize, fibrotize, permanently adhere to the artery wall, and possibly subsequently grow (this occurs in 2-4% of patients after pulmonary embolism). The pressure in the wedge (if we are able to catheterize it despite the presence of thrombi) is normal.

Therapy is surgical - removal of reorganized thrombus by pulmonary endarterectomy (PEA).

Pulmonary hypertension from other causes

These include all unclassifiable causes of pulmonary hypertension such as sarcoidosis, histiocytosis X, lymphadenopathy and cancer.

Therapy is causal.

Clinical symptoms

náhled|250px|Stick fingers as a symptom of pulmonary hypertension in a patient with Eisenmenger syndromeme There are no symptoms that are typical of pure pulmonary hypertension. All symptoms are **non-specific**, which is unfortunately the cause of the often late detection of the disease.

- Exertional dyspnea,
- quick fatigue,
- chest pain,
- syncope,
- hemoptysis,
- swelling of the lower limbs (and other symptoms of right heart failure).

Diagnostics and examination

- **ECG** – reveals signs of right ventricular hypertrophy (pointed P waves in leads II and III, vertical inclination of the electrical axis of the heart, high oscillation R in lead V_1, inverted T in leadsV_1 – V_3, prolongation of activation time of the right ventricle).
- **Chest X-ray** – may reveal enlargement of the right heart compartment, enlargement of the pulmonary trunk.
- **ECHO** – detects hypertrophy of the right ventricle, based on Doppler examination of the pressure gradient on the tricuspid valve, we find out the oriental systolic pressure in the pulmonary trunk, pulmonary or tricuspid

valve insufficiency.
- **HRCT of the chest** – a more detailed view of the cardiac compartments, typically enlargement of the right ventricle, enlargement of its muscle compared to the left ventricle (it is oppressed and has the shape of the letter D).
- **Functional lungs examination**:
  1. spirometry – reveals lung dysfunction,
  2. whole body pletysmography
  3. ventilatory perfusion scintigraphy - reveals a mismatch between ventilation and lung perfusion,
- **Right-sided cardiac catheterization will definitely determine the diagnosis!** – measured:
  1. **mean pulmonary pressure (> 25 mmHg)**,
  2. **wedge pressure (> 12 mmHg)**,
- six-minute walk test - a very simple method, we measure the distance the patient walks in 6 minutes (norm approx. 650 m, patient with pulmonary hypertension about 250 m), the result of the examination correlates with the severity and prognosis of the disease,
- lung biopsy,
- **laboratory examination** – markers of heart failure (natriuretic peptides, cardiac troponins).

PAH Therapy

Determining the right therapy

Usually according to the degree of dyspnea:

- NYHA I–II – conventional therapy,
- NYHA II – oral treatment (endothelin receptor antagonists or PDE5 inhibitors), preventing progression to NYHA III or IV,
- NYHA III – endothelin receptor antagonists (bosentan, ambrisentan), PDE5 inhibitors (sildenafil), or prostanoids (epoprostenol), we prevent progression and strive for improvement,
- NYHA IV – prostanoids administered continuously by infusion, at the same time included in the transplant program, we are trying to improve or bridge to transplantation.

Pharmacotherapy

- **Conventional treatment of PAH** – this includes:
  1. anticoagulant therapy – warfarin,
  2. calcium channels blockers (BCC) – vasodilation (effective in about 5% of patients).

- **Specific treatment for PAH** – therapy that seeks to prevent vasoconstriction or promote vasodilation in the vasculature
  1. endothelin receptor antagonists - prevention of vasoconstriction,
  2. phosphodiesterase 5 (PDE5I) inhibitors - increase the vasodilatory effect of NO by inhibiting the enzyme,
  3. prostacyclin analogues - vasodilatory and antiproliferative effect, due to the short biological half-life the patient must have an infusion pump.

Non-pharmacological treatment

- **lung transplant** – patients with NYHA IV are indicated,
- **percutaneous balloon atrial septostomy (PBAS)** – artificial opening of the atrial septum (creation of a right-left short circuit), part of the blood does not flow through the lungs, the result is an increase in cardiac output of the left ventricle but also blood desaturation.

Odkazy

Související články

- Srdeční selhání (rozcestník)
- Plicní embolie
- Cor pulmonale
- Plicní hypertenze/Repetitorium

Použitá literatura

-
-

Externí odkazy

-
- Plicní hypertenze – video (https://www.youtube.com/watch?v=Dx4QgdN_hI4)

Kategorie:Kardiologie Kategorie:Pneumologie Kategorie:Vnitřní lékařství Kategorie:Interní propedeutika Kategorie:Články s videem