**ILD – interstitial lung disease - information for a patient**

***What do I suffer from?***

You have just found out that you have interstitial lung disease (ILD), or your doctor suspects it. ILDs constitute a large and diverse group of disorders that are characterized by a number of similarities in terms of clinical presentation, results of pulmonary function tests and the presence of abnormal, diffuse parenchymal infiltrations in lung imaging, and are therefore classified together.

ILD may have known causes or clinical context:

• diseases resulting from environmental exposure to inhaled agents that may lead

to ILD (hypersensitivity pneumonia - HP),

• related to occupational exposure (pneumocioses, e.g. silicosis, asbestosis),

• related to systemic connective tissue diseases (e.g. scleroderma-related

interstitial lung disease - SSc-ILD, rheumatoid arthritis-related interstitial lung

disease - RA-ILD)

• resulting from lung damage resulting from the use of various drugs or

radiotherapy as a form of cancer treatment

or may have no known cause:

• idiopathic interstitial pneumonia (IIP),

• sarcoidosis

• other ILDs

Idiopathic pulmonary fibrosis (IPF) is a specific form of chronic progressive and fibrosing interstitial pneumonia of unknown cause that occurs mainly in the elderly. It is a common form of ILD in clinical practice and accounts for approximately 20% of all diagnoses in this group. The disease is limited exclusively to the lungs and is associated with a radiological and/or histopathological pattern of usual interstitial pneumonia (UIP).

It is worth knowing that ILDs are considered rare diseases. The incidence of specific disease entities within the ILD group is less than 5 per 10,000 people. For comparison, diabetes occurs in about 6 out of 100 people, which is about 600 times more common. ILD can appear at any age, but it most often affects older people.

***Why do I suffer?***

The causes of ILDs can vary. Some of them can be identified during the diagnostic process, and in some cases, despite careful diagnostics, the direct cause cannot be identified (see above). However, the predisposition to the disease largely depends on the genetic material that we inherit from our parents. In several to a dozen or so percent of patients with ILD, a detailed history collected from the patient may indicate that other family members have or are suffering from the same disease.

***What are the symptoms of ILD?***

ILD usually leads to very non-specific disease symptoms, which is why its proper diagnosis is often delayed. ILD should be suspected in patients with the following symptoms and abnormalities in additional tests:

- chronic, gradually developing dyspnea on exertion of unknown cause accompanied by cough,

- bilateral basal crackles on auscultation of the lungs,

- clubbed fingers

- restrictive pattern in lung function tests,

- decrease in blood oxygenation during physical exercise,

- diffuse parenchymal infiltrations in lung imaging

***How ILD is diagnosed?***

The differential diagnosis of ILD requires clinical, radiological and pathological assessment (if a lung biopsy is needed to make the diagnosis). Optimally, such an assessment takes place through a multidisciplinary discussion, when medical specialists experienced in diagnosing ILD - a pulmonologist, rheumatologist, radiologist and possibly a pathologist - can discuss the collected medical data about a specific patient during a joint meeting.

Clinical assessment includes:

• medical interviews regarding possible exposure to factors causing ILD (including drugs used), respiratory symptoms and possibly other extrapulmonary symptoms that may accompany selected forms of ILD (e.g. joint and muscle symptoms in the case of ILD accompanying systemic connective tissue diseases, or possible multi-organ symptoms of sarcoidosis)

• physical examination

• pulmonary function tests to assess the severity of the disease, such as:

- spirometry, allowing the assessment of flows and selected lung volumes and

capacities,

- assessment of the efficiency of gas exchange in the lungs (TLco - assessment of the transfer factor of the lung for carbon monoxide),

- plethysmography, allowing the assessment of total lung capacity,

- 6-minute walk test, a simple exercise test assessing the ability to tolerate physical exercise and its impact on blood oxygenation, measured indirectly using pulse oximetry

• arterial blood gasometry, directly assessing the content of respiratory gases in the blood (oxygen and carbon dioxide) and the need for possible home oxygen treatment (to perform it, the radial artery located near the wrist of the hand should be punctured)

• tests assessing the presence of autoantibodies in blood serum collected from a peripheral vein (this test may be helpful in establishing the diagnosis of systemic connective tissue disease or systemic vasculitis, diseases in the course of which lung involvement may occur in the form of ILD)

• peripheral blood morphology tests and blood biochemical tests assessing the functioning of important organs, such as: such as liver or kidneys (to perform them, blood must be taken from a peripheral vein)

• endoscopic examinations of the respiratory system - bronchofiberoscopy with/or without bronchoalveolar lavage (BAL) and biopsy of the bronchial mucosa, allowing in a specific clinical situation, among others, exclusion of infection or cancer in the differential diagnosis of ILD (these tests are not always necessary in the diagnosis of ILD, their need will be discussed with you by your doctor). Bronchofiberoscopy examination involves inserting a thin endoscope into the lumen of the lower respiratory tract, which allows viewing the lumen of the bronchi and collecting material for additional tests. This examination is performed under local anesthesia, pharmacological premedication or short-term general intravenous anesthesia. The doctor conducting or performing the bronchofiberoscopy examination will provide you with comprehensive information on this subject.

• lung biopsy techniques such as transbronchial lung biopsy or cryobiopsy (performed during a bronchofiberoscopy examination) and surgical lung biopsy, a more invasive procedure involving an incision in the chest wall, performed in the operating room. Biopsy examinations are performed under local anesthesia, pharmacological premedication or full general anesthesia, depending on the procedure used. A lung biopsy is not always necessary to diagnose a specific ILD - its need and the benefits and risks associated with using a specific biopsy technique will be discussed with you by your treating doctor. Lung biopsy may be necessary when clinical and imaging assessments do not allow a diagnosis of specific ILD to be made with a high probability.

Radiological assessment includes:

• initial imaging examination, which is a chest x-ray

• a key examination in imaging assessment, called the "gold

standard" of procedure, namely high resolution computed tomography

of the lungs (HRCT - high resolution computed tomography) allowing

the determination of radiological patterns observed in various ILDs

Pathological assessment includes:

• performed when a lung biopsy was performed (surgical lung biopsy, cryobiopsy

or transbronchial lung biopsy) or biopsy of the bronchial mucosa

***What happens if I don’t give a consent for a lung biopsy?***

In a patient in whom biopsy is omitted when the clinical and radiological evaluation does not allow for a correct diagnosis, there is a greater risk of incorrect diagnosis. This may have serious consequences because suspected ILD requires differentiation from diseases such as cancer and infections, including tuberculosis. Delay in the diagnosis of these diseases, and therefore in their treatment, may have serious consequences, including the risk of death. However, it should be remembered that not every clinical situation of suspected ILD requires a biopsy.

***Will I be treated while a diagnosis is established?***

Correct diagnosis is the basis for implementing ILD therapy. Pharmacological treatment of ILD, depending on its type, involves the administration of immunomodulatory drugs (changing/reducing the natural immunity), including glucocorticosteroids and immunosuppressive drugs, as well as antifibrotic drugs (inhibiting pulmonary fibrosis). Treatment of ILDs is usually long-term, and is most often continued for many years or until the end of life. Indications for starting therapy, its form, available pharmacological options and possible side effects related to the implementation of a specific form of therapy will be discussed with you by your doctor after establishing the proper diagnosis of ILD. In addition to pharmacotherapy, an important element of the ILD treatment process are non-pharmacological interventions, including quitting smoking, vaccinations, respiratory rehabilitation, chronic oxygen treatment and palliative care. If treatment options are exhausted, the disease progresses and respiratory failure develops, and the patient meets the qualification criteria, lung transplantation may be considered.

***Will pharmacotherapy of ILD be well-tolerated?***

Side effects associated with long-term use of glucocorticosteroids include increased appetite and weight gain, gastric and duodenal ulcers, diabetes, osteoporosis, mood disorders, sleep disorders, and increased susceptibility to infections. Treatment with immunosuppressive drugs is associated with the risk of infection, liver dysfunction and bone marrow damage. Therapy with antifibrotic drugs may be associated with decreased appetite, nausea, vomiting, diarrhea, weight loss or hypersensitivity to sunlight.

For this reason, treatment must be systematically monitored for tolerability and safety.

***What will happen if I don’t start ILD therapy?***

Failure to treat ILD when there are indications for its implementation may result in disease progression in the lungs, and in the case of multi-organ diseases (e.g. sarcoidosis) also in other affected organs, which may ultimately lead to damage or permanent organ failure. If ILD progresses in the lungs, respiratory failure may develop, which may result in premature death.

***What symptoms are related to disease progression?***

In the respiratory system, symptoms of advanced changes in the lungs or their intensification over time include shortness of breath during physical exercise, deterioration of exercise tolerance, and worsening cough. Objective assessment involves a comparative analysis of the results of lung imaging and lung function tests (spirometry, TLco), which will demonstrate the deterioration of the measured parameters. The progression of the disease is also evidenced by a shortening of the walking distance in the 6-minute walk test.

***How often should I visit a doctor?***

Typically, follow-up visits take place slightly more frequent when starting ILD therapy (even every 1 month), and then usually every 3 to 6 months, if the treatment is tolerated well and the disease is stable. During follow-up visits, selected tests are repeated, most often laboratory tests, functional and imaging tests of the lungs.