Interstitial pneumonia with autoimmune features (IPAF)

Fact sheet

Pulmonary Fibrosis Foundation™

pulmonaryfibrosis.org

What is interstitial pneumonia with autoimmune features?

Interstitial pneumonia with autoimmune features (IPAF) is a research classification describing chronic lung disease in which inflammation and/or scar tissue ("fibrosis") builds up around the airways and in the walls of the air sacs of the lungs. People diagnosed with IPAF often have symptoms, findings on physical exam, or blood tests suggestive of, but not definitive for, an underlying autoimmune condition. Some examples of autoimmune conditions are rheumatoid arthritis, Sjögren's syndrome, scleroderma, and myositis. While the term "pneumonia" is referenced in the name, IPAF is not an infectious pneumonia typically referred to by the general public.

IPAF falls into a large category of diseases called "interstitial lung diseases" or "ILD." There are over 200 types of ILD. Most ILDs have either fibrosis and/or inflammation in the walls of the air sacs. IPAF is one type of ILD. "Pulmonary fibrosis" (PF) is used to refer to any type of ILD in which fibrosis (also known as scarring) is present.

What are the symptoms of interstitial pneumonia with autoimmune features?

People with IPAF can experience symptoms including breathlessness during activities or cough. People can also experience symptoms from other parts of the body including joint pain, stiffness or swelling, dry eyes or dry mouth, difficulty swallowing or heartburn, muscle weakness, fatigue, or rash. These symptoms may not all occur at the same time. These symptoms can also overlap with other disease conditions and are not specific to an IPAF diagnosis.

What causes interstitial pneumonia with autoimmune features?

The cause of IPAF is currently not known. There is ongoing research into the causes of IPAF.

How is interstitial pneumonia with autoimmune features diagnosed?

The diagnosis of IPAF is usually made by a lung doctor (pulmonologist) in conjunction with a specialist in autoimmune diseases that affect multiple organs (a rheumatologist). When a doctor or other healthcare provider suspects that a patient has ILD, they will perform tests that include pulmonary function tests (PFTs), blood work, and a high-resolution CT scan (also known as CAT scan). Pulmonary function tests provide an estimate of lung function.

Brief overview of findings on high-resolution CT scan

Normal lung tissue looks dark gray/black on a high-resolution CT scan (HRCT). Inflammation and/or scar tissue appears light gray or white. Your doctor can review the results of your HRCT scan to show you the areas that are abnormal.

How is interstitial pneumonia with autoimmune features managed?

Currently, there is no specific FDA-approved therapy for IPAF. Your doctor will likely recommend monitoring via regularly scheduled office visits and pulmonary function tests. Depending on your symptoms and PFT results, your doctor may monitor the disease without medications. In other situations, your doctor may use medications that affect the immune system to help stabilize your lung function or other medications to slow decline of the lungs.

Pulmonary rehabilitation, supplemental oxygen, smoking cessation, vaccinations (including COVID-19, influenza, and pneumonia vaccines) are vital to living with IPAF. You can learn more about supplemental oxygen at **pulmonaryfibrosis.org/oxygentherapy**.

You can learn more about pulmonary rehabilitation at **pulmonaryfibrosis.org/pulmonaryrehab**.

Lung transplantation may be an option for some people living with ILD. Early evaluation for lung transplant is important because the process involves a series of appointments and tests to determine if patients are appropriate candidates. You can learn more about lung transplantation at: **pulmonaryfibrosis.org/lungtransplant.**

What is my prognosis?

The prognosis in IPAF is highly variable. Some cases remain stable while others can experience lung function decline over time. The rate of progression can be slow or more rapid. Progression of the lung condition can lead to respiratory failure which is a life limiting condition. No one can predict exactly how long you will live with IPAF, and every individual is different. Your doctor may be able to give you more detailed information about your prognosis.

Are there experimental therapies available?

IPAF is an emerging area of research. Research studies can be found at PFF Clinical Trial Finder at **trials.pulmonaryfibrosis.org**.

Other resources

Fischer A, Antoniou KM, Brown KK, Cadranel J, Corte TJ, du Bois RM, et al. An official European Respiratory Society/American Thoracic Society research statement: interstitial pneumonia with autoimmune features. Eur Respir J. 2015;46(4):976-87.

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