

ILD · Interstitial Lung Disease And Pulmonary Fibrosis

A plain-English guide for you and your family

WHAT IS HAPPENING

Your lungs are made of millions of tiny air sacs (alveoli) where oxygen jumps into your blood. Between each sac is a paper-thin space called the interstitium, just thin enough for oxygen to hop across. In interstitial lung disease (ILD), that thin space gets thick and scarred over time. The scarring can come from your immune system attacking by mistake (like in sarcoidosis or connective tissue disease such as rheumatoid arthritis or scleroderma), from breathing in mold or bird proteins over and over (hypersensitivity pneumonitis), from certain drugs (like the heart drug amiodarone or the chemotherapy drug bleomycin), or from an unknown cause (idiopathic pulmonary fibrosis, or IPF, where with no known cause (idiopathic) means we do not know why). When the interstitium thickens, oxygen cannot cross easily, so you feel short of breath and cough a dry cough with no mucus. Your body tries to help by breathing faster and making your heart pump harder, but the real problem is the scarring itself, which makes the lungs stiff like leather instead of soft like a sponge. Over months, low oxygen can cause your fingernails to curve and bulge (clubbing), and pressure builds in the lung blood vessels (pulmonary high blood pressure (hypertension)), which can tire out the right side of your heart. Each type needs a different fix: if it is IPF, anti-fibrotic drugs (pirfenidone or nintedanib) slow the scarring (they do not reverse it, but they buy time); if it is inflammation from your immune system or a trigger, immune-calming drugs like prednisone help; if it is from a drug or something you breathe in, stopping that trigger is the most important step. The goal is to catch it early with a high-resolution chest CT scan (HRCT, which shows the scarring pattern) and pulmonary function tests (PFTs, which show stiff lungs), name the type, and stop the damage before scarring becomes permanent. For advanced disease, lung transplant is the only cure.

What you might feel

- Shortness of breath that gets worse over weeks to months, especially during activity
- Dry cough that doesn't go away and doesn't bring up mucus

- Crackling sounds (fine crackles or 'Velcro crackles in the lungs (rales)') heard through a stethoscope at the bottom of both lungs
- Feeling very tired all the time (fatigue)
- Clubbing of the fingers—fingertips become round and the nails curve downward
- Unintended weight loss
- Bluish color of the lips or fingernails (cyanosis)
- Fast, shallow breathing even at rest

Call the doctor right away if...

- Oxygen level (SpO₂) below 88% at rest or dropping below 88% during walking—means the lungs can't keep blood oxygen high enough and the patient may need oxygen therapy or hospital care right away.
- Sudden, severe shortness of breath or chest pain—could signal a collapsed lung (pneumothorax), blood clot in the lung (pulmonary a traveling clot lodging in a vessel (embolism)), or heart failure from lung disease, all of which need emergency care.
- Confusion, sleepiness, or trouble staying awake—may mean carbon dioxide is building up in the blood because the lungs can't clear it, a dangerous sign that breathing is failing.
- Rapid worsening over days to weeks (acute flare-up (exacerbation) of IPF)—scarring suddenly speeds up, oxygen drops fast, and the person may need the ICU and a ventilator; this can be life-threatening.
- Swelling of the ankles and belly, along with a raised neck vein—suggests the right side of the heart is failing because high pressure in the scarred lungs is making the heart work too hard (cor pulmonale).

What the care team will do

- Check oxygen saturation with pulse oximeter at rest and after walking 30 feet; give supplemental oxygen by nasal cannula to keep SpO₂ above 88 percent.
- Listen to lung sounds in all fields and report new or louder Velcro-like crackles at the lung bases.
- Monitor respiratory rate, work of breathing, and use of neck or belly muscles to breathe every 4 hours.
- Teach energy-saving techniques like sitting while dressing, using a shower chair, and pacing activities with rest breaks.
- Weigh the patient daily at the same time and watch for sudden weight gain or new swollen ankles.

- Give pirfenidone or nintedanib with food at the same times every day and teach the patient to report nausea, diarrhea, or skin sensitivity to sunlight.
- Screen for depression and social isolation; connect the patient with pulmonary rehabilitation programs and ILD support groups.
- Teach infection prevention: hand washing, avoiding crowds during flu season, and getting pneumonia and yearly flu vaccines.

Medicines you may be given — and why

Pirfenidone

Pirfenidone slows down the scarring (fibrosis) in the lungs by blocking several chemical signals that tell scar-making cells (fibroblasts) to keep laying down thick, stiff tissue. It also dials down inflammation. Think of it like putting the brakes on a runaway scarring process that stiffens the lung's air sacs.

Nintedanib

Nintedanib blocks several growth-factor receptors (docking stations on cells) that normally tell fibroblasts to multiply and make scar tissue. By jamming these receptors — especially for PDGF (platelet-derived growth factor), FGF (fibroblast growth factor), and VEGF (vascular endothelial growth factor) — it slows the lung's scarring process.

Prednisone (or other corticosteroids)

Prednisone is a man-made version of cortisol (the body's natural stress hormone). It powerfully turns down the immune system's attack and inflammation. In inflammatory ILDs (like hypersensitivity pneumonitis, nonspecific interstitial pneumonia NSIP, organizing pneumonia, or ILD linked to autoimmune disease), prednisone tells immune cells to stop releasing inflammatory chemicals and to quiet down, which reduces lung damage and scarring.

Mycophenolate mofetil

Mycophenolate blocks a key enzyme (IMPDH, inosine monophosphate dehydrogenase) that lymphocytes (a type of white blood cell) need to multiply. By stopping lymphocytes from dividing, it turns down the immune attack in inflammatory ILDs (like NSIP, connective-tissue ILD, or sarcoidosis) and is often used as a steroid-sparing agent to lower the prednisone dose.

Azathioprine

Azathioprine is converted in the body into chemicals that interfere with DNA and RNA production in dividing cells, especially lymphocytes (immune cells). By slowing lymphocyte multiplication, it turns down the immune attack in inflammatory ILDs. It is sometimes used as

an alternative or add-on to mycophenolate, though newer evidence favors mycophenolate in many ILDs.

Rituximab

Rituximab is a laboratory-made antibody that targets CD20, a protein on the surface of B cells (a type of white blood cell that makes antibodies). By binding to CD20, rituximab marks B cells for destruction. This is used in ILDs driven by autoimmune disease (especially connective-tissue ILD like rheumatoid arthritis-ILD or systemic sclerosis-ILD) where B cells fuel the immune attack.

Supplemental oxygen

Supplemental oxygen is breathed in through a nasal tube (cannula) or mask to raise the amount of oxygen in your blood when your scarred or inflamed lungs cannot pull in enough on their own. It does not treat the disease itself, but it keeps your organs (especially your heart and brain) fed with the oxygen they need, prevents strain on the heart, and helps you feel less short of breath and more able to move.

Pulmonary rehabilitation

Pulmonary rehabilitation is a supervised exercise and education program that trains your breathing muscles, builds stamina in your legs and arms, and teaches you breathing techniques and energy-saving strategies. It does not change the lung disease itself, but it helps you use the lung function you have more efficiently, reduces shortness of breath during activity, and improves your quality of life and ability to do daily tasks.

Vaccinations (influenza, pneumococcal, COVID-19, RSV)

Vaccines train your immune system to recognize and fight specific viruses and bacteria before you get infected. In ILD, your lungs are already damaged and cannot handle the extra injury from infections like flu, pneumonia, COVID-19, or RSV (respiratory syncytial virus). Vaccines significantly lower your risk of these infections and of severe illness if you do catch them.

Proton pump inhibitor (e.g. omeprazole, esomeprazole) for gastroesophageal reflux disease (GERD)

Proton pump inhibitors (PPIs) block the proton pumps in the stomach lining that produce stomach acid. By lowering acid production, PPIs reduce acid reflux (when stomach contents wash back up into the throat and can be breathed into the lungs — called aspiration). In ILD, repeated micro-aspiration of acid can worsen lung inflammation and fibrosis. Treating reflux may slow disease progression, especially in IPF.

N-acetylcysteine (NAC)

N-acetylcysteine is an antioxidant that can neutralize harmful oxygen radicals (reactive molecules that damage cells) and may replenish glutathione (the body's own antioxidant). It was once thought to slow fibrosis in IPF by reducing oxidative stress, but large trials (PANTHER-IPF) showed NO benefit in IPF and possible harm when combined with prednisone and azathioprine. It is now NOT recommended for routine use in IPF. It may still be used in some other ILDs on a case-by-case basis, and it is used as a mucolytic (mucus thinner) in some lung diseases, though evidence in ILD is weak.

Where this information comes from

- ATS/ERS/JRS/ALAT Clinical Practice Guideline: Idiopathic Pulmonary Fibrosis (2018)
- American Thoracic Society (ATS)
- Fleischner Society
- CHEST

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