

ILD · Interstitial Lung Disease And Pulmonary Fibrosis

ILD is lung scarring between air sacs. Always name which type. Use pirfenidone or nintedanib (anti-fibrotic drugs that slow scarring) for IPF. Use prednisone or other immune-calming drugs for inflammatory types like NSIP, hypersensitivity pneumonitis, or connective tissue ILD. Remove the trigger in drug-caused or exposure-related disease. High-resolution chest CT (HRCT) is the key test to see the pattern. Pulmonary function tests show stiff lungs (restrictive pattern). Check oxygen with walking (6-minute walk test) to catch hidden drops. Lung transplant is the only cure for end-stage fibrosis.

THE ONE-LINE TRUTH

Interstitial lung disease is long-lasting swelling and scarring in the thin space between the air sacs, making it hard for oxygen to jump into the blood.

The chain — how it unfolds

1. The interstitium (the thin space between air sacs and blood vessels) gets swollen and scarred. — why: Depending on the type of ILD, the trigger is different: in IPF, the cause is unknown but the scarring is relentless; in hypersensitivity pneumonitis, repeated exposure to mold (often from hot tubs, humidifiers, or water-damaged buildings), bird proteins (from pet birds or feather pillows), or other antigens triggers an immune reaction; in connective tissue disease (like systemic sclerosis, rheumatoid arthritis, or polymyositis), the immune system attacks the lungs; in sarcoidosis, clumps of immune cells (noncaseating granulomas) form in the interstitium; in drug-induced ILD, medications like amiodarone, bleomycin, methotrexate, or nitrofurantoin damage lung tissue. No matter the cause, the result is the same: the thin interstitium thickens and scars, turning soft, stretchy lung tissue into stiff scar tissue that does not work.

2. Oxygen cannot cross easily from the air sac into the blood, so blood oxygen drops (hypoxemia). — why: Normally, oxygen hops across the paper-thin interstitium in a fraction of a second. When the interstitium is thick and scarred, oxygen has to travel through a thick wall, which takes longer and some oxygen never makes it across. Blood leaving the lungs has less oxygen than normal (often oxygen saturation below 88 percent with activity, sometimes below 90 percent at rest in advanced disease), and you feel short of breath (dyspnea), especially with activity like climbing stairs or walking fast.
3. The lungs become stiff and lose stretch (reduced lung compliance, restrictive pattern on pulmonary function tests). — why: Scar tissue is stiff and does not stretch like healthy lung tissue. As more interstitium scars, the lungs shrink and resist expansion. On pulmonary function tests (PFTs, breathing tests), total lung capacity (TLC, the total amount of air your lungs can hold) and forced vital capacity (FVC, the amount of air you can blow out after a deep breath) drop (restrictive pattern, usually TLC and FVC below 80 percent of predicted). You feel like you cannot take a deep breath, and breathing takes more effort.
4. You cough constantly, usually a dry cough with no mucus. — why: The scarred interstitium irritates nerve endings in the lungs (like the vagus nerve, which senses irritation and triggers coughing), triggering a cough reflex. The cough is the body trying to clear something that feels wrong, but there is no mucus or infection to clear — just the stiff, irritated tissue itself.
5. On exam, the doctor hears fine crackles (Velcro crackles in the lungs (rales)) at the bases (bottoms) of the lungs. — why: Scarred air sacs pop open during a deep breath, making a crackling sound like pulling apart Velcro. This classic finding is heard best at the bottom (bases) of the lungs, where scarring often starts, and is a hallmark of ILD on physical exam.
6. Over months to years, your fingernails curve and bulge (digital clubbing). — why: Long-lasting low oxygen (chronic low oxygen in the blood (hypoxemia)) triggers changes in the fingertips. The body grows extra tissue and blood vessels under the nail, making the nail curve and the fingertip bulge. Clubbing is a sign that oxygen has been low for a long time (usually months) and is seen in about 50 percent of people with IPF, less often in other ILD types.
7. Pressure in the lung blood vessels goes up (pulmonary high blood pressure (hypertension)), and the right side of the heart works harder and can eventually fail (cor pulmonale). — why: Scarring and low oxygen make blood vessels in the lungs tighten and narrow (pulmonary vasoconstriction). The right side of the heart (right ventricle) has to pump harder to push blood through these tight vessels. Over time, the right heart muscle gets tired, thick, and weak (right ventricular hypertrophy and failure), leading to right heart failure: legs and belly swell (peripheral swelling (edema) and fluid buildup in the belly (ascites)), liver gets congested (hepatomegaly), neck veins bulge (jugular venous distension), and breathing gets even worse.

8. In IPF, acute exacerbations (sudden, severe worsening) can happen, where breathing becomes extremely hard, oxygen drops fast, and the lungs look worse on CT scan with new ground-glass opacities (hazy patches that look like frosted glass). — why: We do not fully understand why, but in IPF the scarring can suddenly speed up (acute flare-up (exacerbation) of IPF, or AE-IPF), often triggered by a virus (like influenza or COVID-19), silent infection, aspiration (breathing in food or stomach acid), or for no clear reason. The already-damaged lungs cannot handle the extra hit, and the patient crashes. Mortality is high in acute exacerbations (about 50 percent die within a few months) even with ICU care, oxygen, and steroids.

Spot it — what you see, and why

- Shortness of breath that gets worse over weeks to months, especially during activity: Scarring (fibrosis) thickens the walls of the air sacs (alveoli) in the lungs, making it harder for oxygen to pass from the air into the bloodstream. When the body needs more oxygen during exercise, the damaged lungs can't keep up, so you feel breathless.
- Dry cough that doesn't go away and doesn't bring up mucus: Scarring and inflammation irritate the airways and the interstitium (the thin tissue between the air sacs). The body tries to clear the irritation by coughing, but because this isn't caused by mucus or infection, the cough stays dry.
- Crackling sounds (fine crackles or 'Velcro crackles in the lungs (rales)') heard through a stethoscope at the bottom of both lungs: When scarred air sacs pop open during a deep breath, they make a crackling sound—like pulling apart Velcro—that the doctor can hear with a stethoscope. This tells the doctor that the tiny air sacs are stiff and fibrotic.
- Feeling very tired all the time (fatigue): The scarred lungs can't pull in enough oxygen to meet the body's needs, so cells don't get the fuel they need to make energy. This leaves you feeling drained and worn out even after rest.
- Clubbing of the fingers—fingertips become round and the nails curve downward: Long-term low oxygen in the blood signals the body to grow extra tissue at the fingertips, making them bulge and the nails curve. Clubbing is a sign that the lungs have been struggling for a while.
- Unintended weight loss: Breathing with stiff, scarred lungs takes extra energy, and chronic inflammation can speed up the body's metabolism. Together, these burn more calories than you take in, leading to weight loss.

Red flags — act now

- 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).

Treat it — in this order

1. Identify the type of interstitial lung disease — with no known cause (idiopathic) pulmonary fibrosis (IPF) versus inflammatory ILD (NSIP, connective tissue disease-ILD, sarcoidosis, hypersensitivity pneumonitis) versus drug-induced — using high-resolution CT, clinical history, autoimmune labs, and sometimes lung biopsy → Knowing which type of scarring the patient has tells you whether to use antifibrotic drugs (for IPF, which is scarring without much inflammation), anti-inflammatory drugs like corticosteroids and immunosuppressants (for inflammatory ILDs where immune cells are attacking the interstitium), or simply stop the trigger (for drug-induced or hypersensitivity pneumonitis). The type is the map that chooses which medicine will work.
2. For with no known cause (idiopathic) pulmonary fibrosis (IPF): start pirfenidone (an anti-inflammatory and antifibrotic drug) or nintedanib (a tyrosine kinase inhibitor that blocks scar-building signals) to slow the rate of lung stiffening and decline in lung function → These antifibrotic drugs tell fibroblasts (the cells that lay down scar tissue) to slow down. They cut the rate of lung-function decline by about half and reduce the risk of hospitalization and death. Patients still get worse over time, but much more slowly.

3. For inflammatory ILDs (NSIP, connective tissue disease-ILD, sarcoidosis, chronic hypersensitivity pneumonitis): use corticosteroids (prednisone starting at 0.5–1 mg/kg/day, tapered over months) plus a steroid-sparing immunosuppressant such as mycophenolate mofetil, azathioprine, or rituximab (a monoclonal antibody that wipes out B cells) to calm the immune attack on the interstitium → Immunosuppressants calm T cells, B cells, and macrophages (immune cells) that are inflaming and damaging the interstitium (the tissue web between air sacs). Less inflammation means less new scarring, better oxygen crossing into blood, and lung function that can stabilize or even improve.
4. Give supplemental oxygen (by nasal cannula or portable concentrator) when oxygen saturation drops below 88% at rest or with activity → Extra oxygen in the air the patient breathes pushes more oxygen across the stiff, scarred interstitium (tissue web between air sacs) into the blood, so the body and brain get what they need. It reduces shortness of breath, lets patients move more, and improves survival in patients with chronic low oxygen (< 88% at rest or with activity).
5. Prescribe pulmonary rehabilitation — a 6- to 12-week program of breathing exercises, walking training, strength training, and disease education — supervised by a respiratory therapist or physical therapist → Rehab trains the muscles and breathing patterns so the patient uses less oxygen during daily tasks and feels less short of breath with the lungs they have. Patients walk farther, feel less tired, and report better quality of life.

Your top priorities at the bedside

1. 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. — why: Scarred lung tissue cannot exchange oxygen well, and low oxygen (hypoxemia, which means not enough oxygen in the blood) damages the heart and brain; keeping SpO₂ above 88 percent protects organs while doctors work on the underlying disease.
2. Listen to lung sounds in all fields and report new or louder Velcro-like crackles at the lung bases. — why: Velcro crackles (fine crackling sounds like pulling apart Velcro) mean stiff, scarred air sacs (alveoli, which are tiny lung balloons where oxygen moves into blood); worsening crackles signal disease progression needing treatment adjustment.
3. Monitor respiratory rate, work of breathing, and use of neck or belly muscles to breathe every 4 hours. — why: Using accessory muscles (extra breathing muscles in the neck and abdomen) or breathing faster than 20 times per minute at rest means the lungs are failing to move enough air, signaling respiratory failure that may need ICU care or mechanical ventilation (breathing machine).