

PULMONARY FIBROSIS

DISEASE / PATHOLOGY

- Progressive scarring/thickening of lung interstitium causing stiff lungs and impaired oxygen diffusion.
- Restrictive disease: lungs cannot expand normally; work of breathing increases.
- Gas exchange worsens over months-years; hypoxemia is often exertional first, then at rest.
- Idiopathic pulmonary fibrosis is common in older adults and has unpredictable progression.
- Acute exacerbation = sudden worsening dyspnea over days-weeks; can be life-threatening.

RISK FACTORS

- Age >50-60, male sex, former/current smoking, family history/genetic predisposition.
- Occupational/environmental exposure: metal/wood dust, silica, asbestos, farming/bird/mold exposure.
- Autoimmune/connective tissue disease and chronic hypersensitivity pneumonitis.
- Medication/radiation causes: amiodarone, nitrofurantoin, bleomycin, methotrexate, chest radiation.
- GERD and recurrent microaspiration may worsen disease.

SIGNS AND SYMPTOMS

- Classic: progressive exertional dyspnea, dry nonproductive cough, fatigue, decreased exercise tolerance.
- Assessment: fine inspiratory crackles described as Velcro-like; tachypnea; shallow breathing.
- Late: digital clubbing, cyanosis, weight loss, pulmonary hypertension/cor pulmonale signs.
- Red flags: sudden worsening dyspnea, new fever, chest pain, hemoptysis, syncope, or rapidly falling SpO₂.
- Expected: oxygen needs often increase with exertion before resting hypoxemia appears.

DIAGNOSTIC TESTING

- High-resolution CT is key; usual interstitial pneumonia pattern supports IPF diagnosis.
- PFTs: restrictive pattern with reduced FVC/TLC and decreased DLCO.
- 6-minute walk test assesses exertional desaturation and functional capacity.
- Pulse ox/ABG evaluate hypoxemia; autoimmune labs help identify secondary causes.
- Lung biopsy may be considered if HRCT/clinical picture is not diagnostic.

TREATMENT

- Assess first: SpO₂ at rest/ambulation, work of breathing, cough, fatigue, and activity tolerance.
- Antifibrotics for IPF: nintedanib or pirfenidone may slow decline; monitor liver enzymes and GI effects.
- Oxygen therapy for hypoxemia; titrate with activity and sleep per prescription.
- Pulmonary rehab, vaccines, nutrition support, and early palliative symptom support improve function/quality.
- Report acute worsening immediately; evaluate for infection, PE, pneumothorax, heart failure, or exacerbation.
- Need-to-know: bronchodilators may not help unless there is coexisting airway disease.

PATIENT EDUCATION

- Pace activity; use oxygen exactly as prescribed, including during exertion if ordered.
- Stop smoking/vaping; avoid dust, fumes, birds/mold exposures if sensitive.
- Take antifibrotics consistently; report severe diarrhea, nausea, photosensitivity, or signs of liver injury.
- Keep vaccines current; avoid sick contacts; report respiratory infections early.
- Call provider for increased O₂ needs, worsening cough, fever, chest pain, syncope, or new leg swelling.
- Discuss advance care planning, transplant referral when eligible, and pulmonary rehab early.

ADDITIONAL NOTES:

- Restrictive = difficulty expanding lungs; obstructive = difficulty getting air out.
- High-yield sign: dry cough + exertional dyspnea + Velcro crackles + clubbing.
- Priority: new sudden dyspnea in pulmonary fibrosis is urgent, not "expected progression."