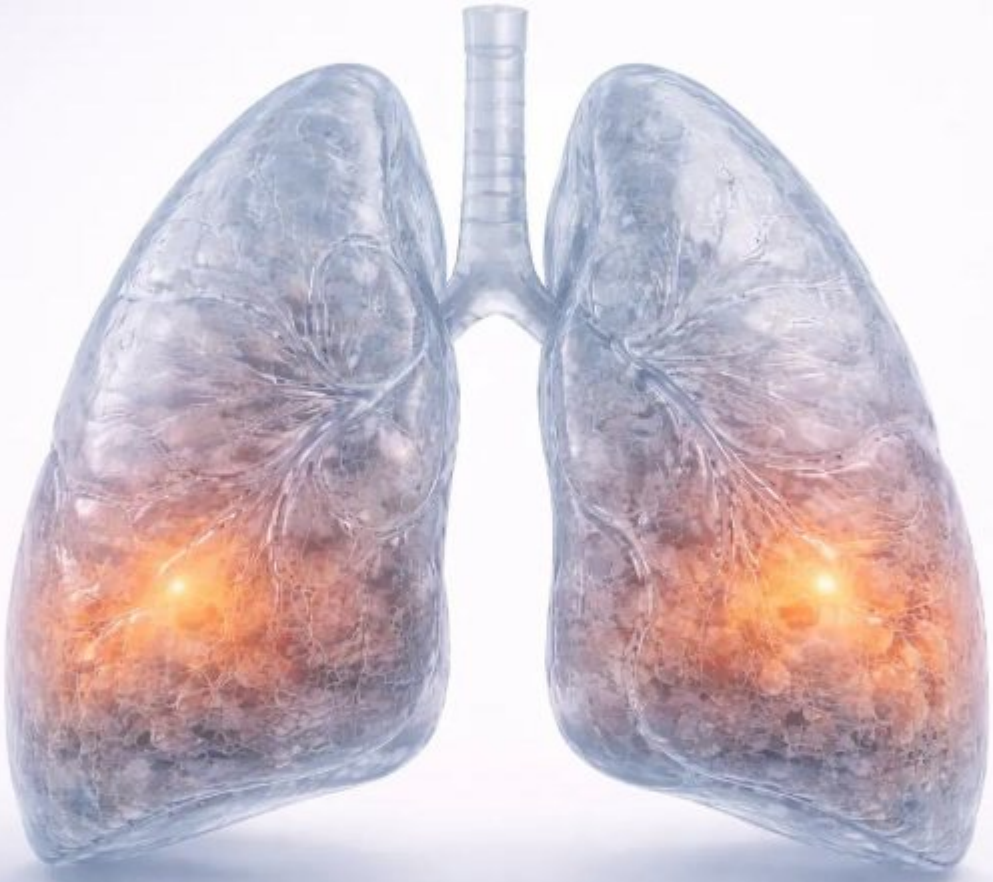
INTERSTITIAL LUNG DISEASE

National Standard Treatment Guideline



Ministry of Health
Republic of Maldives

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World Health Organization
Maldives

National Standard Treatment Guidelines

- Acid Peptic Disease
- Acute Anxiety
- Acute Pancreatitis
- Acute Psychosis
- Acute kidney Injury
- Arrhythmia
- Chronic Liver Disease
- Chronic Pancreatitis
- Chronic kidney disease
- Congenital Heart Diseases
- Dementia
- Depression
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- Interstitial Lung Disease
- Liver Failure
- Obesity
- Obstructive Sleep Apnoea
- Osteoarthritis
- Ovarian Cancer
- Pneumonia
- Stroke
- Upper Gastrointestinal bleed
- Unstable Angina

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GUIDELINES DEVELOPMENT METHODOLOGY

The development of the Maldives Standard Treatment Guidelines (STGs) followed a structured, evidence-informed, and consensus-driven methodology adapted from internationally accepted guideline-development standards and the Delhi Society for Promotion of Rational Use of Drugs (DSPRUD) model. The process combined systematic evidence retrieval, critical appraisal, contextual adaptation, and multidisciplinary expert review to ensure feasibility, clinical relevance, and national ownership.

1. Determining Scope and Priority Conditions

Priority clinical conditions were identified through consultation with national programme managers, specialty clinicians, and health-system stakeholders. Selection criteria included: (i) major causes of morbidity and mortality, (ii) observed variation in clinical practice or prescribing patterns, (iii) potential to improve patient outcomes, and (iv) the feasibility of implementation across health-facility levels in Maldives. The final list of diseases reflected national epidemiology, service-delivery capacity, and essential-medicine availability.

2. Identification of Existing Evidence and Source Guidelines

A targeted search strategy was used to identify high-quality existing clinical guidelines. Searches were conducted across international guideline repositories (e.g., WHO, NICE, SIGN and other intergovernmental bodies, international and national guideline repositories, specialty societies and professional associations.

3. Quality Appraisal of Source Guidelines

Retrieved guidelines were screened for transparency of development, methodological rigour, clarity of recommendations, applicability to health-system reality, editorial independence. Guidelines were included if they met the Institute of Medicine (IOM) definition of a clinical guideline and addressed treatment or management of priority conditions. Guidelines that did not meet minimum quality standards, review articles, diagnostic criteria, or technical standards were excluded.

4. Adoption, Adaptation, and Contextualization

The guideline-development team employed an adopt–adapt–contextualize model:

- **Adoption**: High-quality recommendations that aligned with Maldivian health-system realities were retained without modification.
- **Adaptation**: Recommendations were modified when local considerations such as diagnostic capacity, medicine availability, workforce skills, referral pathways, or cost constraints affected feasibility.

- **Contextualization**: Where evidence was absent or inconclusive, conditional recommendations were formulated based on expert consensus, with explicit consideration of pragmatism, safety, and local workflows.
  Medicines were selected in alignment with the Maldives National Essential Medicines List (NEML), based on suitability, efficacy, safety, and availability.

5. Expert Consensus and Multidisciplinary Input

Draft recommendations were initially prepared by experts from the DSPRUD, India, providing a strong methodological foundation for the process. Building on this, a collaborative and participatory process brought together clinicians from internal medicine, paediatrics, obstetrics-gynaecology, surgery, emergency medicine, endocrinology, cardiology, general practitioners, and public health representing different levels of healthcare. Consensus was achieved through moderated discussions, iterative revisions, and resolution of divergent views. For topics lacking strong evidence, recommendations were derived from expert clinical judgment grounded in extensive practice experience.

6. Drafting, Peer Review, and Validation

Each guideline section was organized in a standard format including key clinical features, essential investigations, non-pharmacological management, pharmacological therapy (with step-up/step-down options where relevant), referral criteria, paediatric considerations, and follow-up requirements. Drafts were peer-reviewed by senior clinicians and national experts. Reviewer comments were systematically integrated to strengthen clarity, accuracy, and applicability.

7. Addressing Conflicts of Interest

All contributors declared the absence of conflicts of interest. Individuals with potential or perceived conflicts were excluded from authorship or decision-making roles.

8. Updating and Future Revisions

The STGs were conceptualized as a living document. Future updates will incorporate new scientific evidence, changes in essential-medicine availability, national programme priorities, and user feedback from clinicians. Periodic review cycles will ensure the continued relevance and reliability of recommendations.

9. Distinctive Features of the Guidelines

Developed through a collaborative process involving a large group of multidisciplinary experts from different levels of healthcare, the guidelines incorporate the following distinctive features:

- **Diagnostic Assumption and Confirmation**: While assuming that an initial diagnosis has been established by the healthcare provider, the guidelines provide essential information for confirming diagnoses. This includes a comprehensive overview of major signs and symptoms, descriptions of confirmatory tests, and clear guidance on practices that are prohibited, discouraged, or unreliable—promoting evidence-based medicine supported by relevant references.
- **Comprehensive Treatment Approach**: The guidelines offer a systematic, up-to-date framework for managing medical conditions across the continuum of care. They begin at the primary care level and extend to secondary and tertiary care, incorporating protocols for treatment response assessment and referral criteria as integral components.
- **Diverse Treatment Modalities**: Recommendations encompass both non-pharmacological and pharmacological interventions and surgical intervention where applicable, providing flexibility for individualized treatment plans. Cautionary notes are included where necessary to ensure safe and effective use of therapies.
- **Assessment and Referral Criteria**: Clear criteria and goals for evaluating patient response to treatment are provided, along with guidance on when referral to higher levels of care is warranted ensuring continuity and comprehensiveness in patient management.

INTERSTITIAL LUNG DISEASE

ACKNOWLEDGEMENTS

The Government of the Republic of Maldives is committed to ensuring universal access to quality health services for all citizens. The Constitution of Maldives mandates the progressive realization of rights, including the right to good standards of health care for the population. In line with this national commitment, standardized quality health services are regarded as the foundation of a strong and equitable healthcare system.

This important work would not have been possible without the cooperation and support of many individuals and institutions. We express our sincere appreciation to the Honourable Minister of Health, Abdullah Nazim Ibrahim, for his leadership, commitment, and continuous guidance throughout the development process. We are grateful to WHO and ADB for their significant contribution, support, and technical assistance.

Our heartfelt gratitude is extended to the technical lead and editor, Dr. Sangeeta Sharma, Professor, Neuropsychopharmacology, IHBAS and President, Delhi Society for Promotion of Rational Use of Drugs (DSPRUD), and her team. We express our deepest appreciation to the Maldivian and DSPRUD experts and contributors who played a pivotal role in this process. Their technical expertise and dedication to adapt the standards to the Maldivian context have been instrumental in the development and finalization of these guidelines. The time, experience, generous sharing of knowledge and insights contributed by all parties have not only enriched the work but also have been invaluable in making these standards practical, locally acceptable, and aligned with the needs of the resident population.

It is important to acknowledge the immense efforts, involvement, timely coordination, collaboration, and dedication of the Quality Assurance and Regulation Division team who made it possible for these Clinical Treatment Guidelines to come into existence.

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QUICK REFERENCE GUIDE

Interstitial lung diseases (ILDs) are increasingly recognized worldwide, with prevalence rising especially among older adults. They predominantly affect people over 50 years, though autoimmune-related ILDs often occur in younger women. Morbidity is high due to progressive breathlessness, hypoxemia, and complications like pulmonary hypertension, while mortality varies by subtype - idiopathic pulmonary fibrosis has a median survival of 3-5 years if untreated. Outcomes depend on early diagnosis and phenotype-specific therapy; timely intervention can slow progression but rarely reverses established fibrosis.

ILD is a disorder causing inflammation and/or fibrosis of the lung interstitium leading to impaired gas exchange and progressive dyspnea. Major phenotypes are:

- Fibrosing ILD: idiopathic pulmonary fibrosis (IPF), progressive fibrosing ILD.
- Inflammatory/autoimmune ILD: connective-tissue-disease-associated ILD (CTD-ILD), hypersensitivity pneumonitis (HP), sarcoidosis.
- Exposure/drug-related ILD: occupational/environmental antigens, medications, radiation.

Causes, risk factors & triggers

- Causes/associations: autoimmune disease, chronic antigen exposure (birds, mold, dusts), drugs (amiodarone, nitrofurantoin, checkpoint inhibitors), radiation.
- Risk factors: age >50, smoking, family history, occupational exposures.
- Triggers of decline/exacerbation: respiratory infections, aspiration/gastroesophageal reflux disease (GERD), thromboembolism, drug toxicity, unchecked antigen exposure.

Evaluation for diagnosis

- Clinical features: exertional dyspnea, dry cough, bibasal crackles; clubbing (often fibrosing disease).
- Physical examination: oxygen saturation at rest and on exertion; signs of pulmonary hypertension (loud P2, edema).
- Laboratory investigations: autoimmune panel as indicated (antinuclear antibodies, rheumatoid factor, anti-cyclic citrullinated peptide, extractable nuclear antigen/myositis antibodies); exclude infection.
- Physiology: pulmonary function tests (PFTs) with forced vital capacity (FVC) and diffusing capacity for carbon monoxide (DLCO); six-minute walk test (6MWT) with oximetry.
- Imaging (key): high-resolution computed tomography (HRCT). Definite/probable usual interstitial pneumonia (UIP) pattern + right clinical context can establish working IPF.

- Confirmation: multidisciplinary team (MDT) review (pulmonology, radiology, rheumatology ± pathology). Bronchoalveolar lavage/biopsy (transbronchial cryobiopsy or video-assisted thoracoscopic surgery) only if needed to resolve uncertainty.

Classification / severity assessment

- By biology: fibrosing vs inflammatory.
- Progression flags: ≥10% relative FVC decline, or 5-9% with worsening symptoms/imaging; ≥15% DLCO fall; rising oxygen need; shorter 6MWT distance.
- Impact: dyspnea grade (modified Medical Research Council, mMRC), quality of life tools (King's Brief Interstitial Lung Disease, K-BILD).

Differential diagnosis

Asthma/COPD overlap, heart failure, pulmonary embolism, infections (including tuberculosis), eosinophilic lung disease, drug toxicity, malignancy/lymphangitic spread.

Management goals & principles

- Goals: slow progression, control inflammation (when present), reduce symptoms and exacerbations, maintain function/QoL, minimize harm, plan ahead.
- Principles: phenotype-driven therapy; remove triggers; shared decisions within an MDT; stepwise, risk-adjusted care; early pulmonary rehabilitation and oxygen when indicated.

Approach to management

1. All patients: stop smoking; vaccinations (influenza, pneumococcal, COVID-19 per local policy); exposure removal; GERD management; education and action plan for flares.
2. Inflammatory/autoimmune or HP with active inflammation: immunomodulation (see table).
3. Fibrosing disease/IPF or progressive pulmonary fibrosis: antifibrotic therapy (see table).
4. Supportive care throughout: pulmonary rehabilitation, breathlessness strategies, psychosocial support, palliative care integration; long-term oxygen therapy (LTOT) if criteria met.

Non-pharmacological interventions

- Lifestyle/trigger control: antigen avoidance (birds/mold/dust), mask use/ventilation at work, reflux precautions.
- Pulmonary rehabilitation: supervised then home-based exercise, pacing, energy conservation, breathing retraining.
- Oxygen: titrate to keep SpO_2 ≥90% at rest/exertion/sleep when hypoxemic.

Pharmacological therapy

Phenotype / purpose	Drug	Usual dose & route	Duration	Major cautions/notes
IPF / progressive fibrosing ILD - antifibrotic	Nintedanib	150 mg orally twice daily with food (consider 100 mg bid if intolerant)	Long-term if tolerated	Diarrhea, ↑ liver enzymes; avoid in severe hepatic impairment; bleed risk.
	Pirfenidone	Titrate to 801 mg orally three times daily (total 2403 mg/day)	Long-term if tolerated	Photosensitivity, GI upset, raised liver enzymes; avoid with severe hepatic/renal failure.
Inflammatory CTD-ILD / HP with active inflammation	Prednisolone (glucocorticoid)	0.5-1 mg/kg/day short course, then taper to lowest effective	Weeks-months, taper	Infection, hyperglycemia, osteoporosis; gastric protection if risk.
	Mycophenolate mofetil	500-1500 mg orally twice daily	Long-term steroid-sparing	Cytopenias, teratogenic; monitor CBC/LFTs.
	Azathioprine	1-2 mg/kg/day orally	Long-term steroid-sparing	TPMT deficiency risk, cytopenias, hepatotoxicity.
	Rituximab	1000 mg IV day 1 & 15 (or 375 mg/m² weekly ×4)	Re-dose per response	Infusion reactions, infections; screen hepatitis B.
	Cyclophosphamide (severe rapidly progressive CTD-ILD)	500-750 mg/m² IV every 2-4 weeks	Limited induction	Cytopenias, hemorrhagic cystitis, infertility; prophylaxis/monitoring needed.
Symptom control	Antitussives (trial)	e.g., low-dose opioids for refractory dyspnea; gabapentin for cough (off-label)	Short courses	Sedation, falls; use cautiously.
Comorbidity management	Proton-pump inhibitor for reflux; diuretics in right-heart failure; anticoagulation only if another clear indication	Standard dosing	As indicated	Align with overall goals of care and drug interactions.

Note: Use the lowest effective dose, reassess frequently, and adjust for renal/hepatic function. Prophylaxis for pneumocystis jirovecii pneumonia (PJP) may be needed with multi-agent immunosuppression.

Assessment of response, review, and adjustment

- Follow-up cadence: stable every 3-6 months; progressive/unstable monthly-quarterly.
- Track: symptoms (mMRC, cough/ fatigue scores), PFTs (FVC, DLCO) every 3-6 months, 6MWT, oxygen needs; HRCT only for significant unexplained change or complications.
- Step-up when: ≥10% FVC decline (or 5-9% plus symptoms/imaging), rising oxygen need, functional drop, or new fibrosis progression.
- Step-down when: stable 6-12 months with low burden and no progression; taper steroids slowly; continue antifibrotic if tolerated.
- Before changing therapy: confirm adherence, remove exposures, treat infection, review drug toxicity, and discuss goals in MDT.

Referral (tiered)

- Primary to Secondary: uncertain diagnosis; persistent/worsening symptoms; resting desaturation; abnormal X-ray suggestive of ILD.
- Secondary to Tertiary/ILD center: need for HRCT protocol, biopsy, or antifibrotics/immunosuppression starts; progressive disease; pulmonary hypertension; acute exacerbation; complex comorbidities; transplant consideration; palliative needs beyond local scope.

Complications

Acute exacerbation, secondary infections, pulmonary hypertension, right-ventricular failure, respiratory failure requiring LTOT or non-invasive ventilation (NIV) in selected overlap, treatment toxicities (hepatotoxicity, cytopenias), thromboembolism, osteoporosis from steroids.

Patient education: objectives & instructions

- Understand the illness: often slowable, not usually reversible; aim for function and symptom control.
- Do: take medicines as prescribed; attend all reviews; keep active with paced exercise; use oxygen safely; avoid triggers (mold/birds/dust); vaccinate; stop smoking.
- Red flags - seek urgent care: sudden dyspnea, new resting hypoxemia, chest pain, hemoptysis, fever with increased cough/sputum.
- Advance care planning: discuss ceilings of care, ventilation preferences, and complete Physician Orders for Life-Sustaining Treatment (POLST) or Medical Orders for Scope of Treatment (MOST) where available; revisit after any major change.

INTERSTITIAL LUNG DISEASE

INTRODUCTION

Interstitial lung diseases (ILD) are a diverse set of conditions that inflame or scar the lung interstitium, impair gas exchange, and cause progressive breathlessness with high morbidity and mortality if missed. The global burden is rising, especially in older adults; Asian data show the same trend with more fibrosing phenotypes such as idiopathic pulmonary fibrosis and connective-tissue-disease-associated disease. Prevalence estimates vary because definitions and diagnostic capacity differ, but studies from Asia show growing recognition and underdiagnosis outside tertiary centers. Many ILDs are chronic and not curable, yet early identification, trigger removal, and disease-modifying therapy can slow decline; some causes (for example, hypersensitivity pneumonitis or drug-induced ILD) improve if the trigger is removed early. A clear, standardized evaluation and treatment pathway enables timely diagnosis, consistent therapy, appropriate referral, and routine data capture to improve outcomes.

SCOPE OF THE GUIDELINES

These guidelines apply to adult and adolescent patients with suspected or confirmed ILD across primary, secondary, and tertiary care in the Maldives. Primary care professionals are expected to recognize symptoms, red flag signs, and acute exacerbations, perform initial screening, and arrange early referral when indicated. The document covers case recognition, basic workup, referral decision rules, and first-line pharmacologic and non-pharmacologic management, distinguishing fibrosing from inflammatory phenotypes when feasible. It presents a stepwise management approach, defines criteria for multidisciplinary review or therapy escalation, and outlines (without detailed protocols) pulmonary rehabilitation, lung transplantation referral, and integration of palliative care.

Intended users

Medical officers, family physicians, internists/physicians, pulmonologists, radiologists, rheumatologists, thoracic surgeons, intensive care teams, respiratory therapists, radiology teams, nurses, community health workers, and the multidisciplinary team (MDT).

Primary care (island/atoll clinics)

- Spot symptoms/red flags and acute exacerbations. Do basics: oximetry, chest X-ray if available, exposure/autoimmune history, initial labs. Start first steps: trigger removal, vaccines, smoking cessation, oxygen if hypoxic. Refer early using clear criteria.

Secondary care (regional hospitals)

- Confirm suspicion; perform spirometry ± diffusion capacity when available. Arrange high-resolution CT (HRCT) or coordinate with tertiary hub. Distinguish likely fibrosing vs inflammatory patterns; begin guideline-based therapy and pulmonary rehab where feasible. Escalate to multidisciplinary team (MDT) for uncertain or progressive cases.

Tertiary care (central centers)

- Provide advanced diagnostics: HRCT protocols, autoimmune panels, bronchoscopy/ biopsy when indicated. MDT diagnosis and staging; initiate/monitor antifibrotics or immunosuppressants. Manage complex exacerbations and comorbidities; coordinate rehab, transplant referral, and palliative care.

Pulmonary function testing (including diffusion capacity) may not be uniformly available. Access to antifibrotics, certain immunosuppressants, lung biopsy, and transplantation is limited; choices about therapy and invasive diagnostics should be guided by local capacity and, when needed, escalated through appropriate referral.

DEFINITIONS

Interstitial lung disease (ILD) refers to a group of roughly 200 disorders that cause inflammation and scarring of the lung interstitium. Common features across ILDs include progressive dyspnea, dry cough, hypoxia, declining lung function, bilateral diffuse infiltrates on imaging, and reduced functional capacity and quality of life. Most cases have an identifiable driver: environmental or occupational exposures (mold, birds, silica, asbestos), medications, or underlying systemic autoimmune disease. Genetic susceptibility and chronic exposure to pollutants modulate individual risk and disease behavior.

Idiopathic pulmonary fibrosis (IPF) is the most aggressive and prototypical fibrosing ILD. It produces irreversible lung scarring, leads to chronic respiratory decline, and carries a poor prognosis: median survival is 3-5 years from diagnosis if untreated.

Classification follows the American Thoracic Society/European Respiratory Society framework:

Idiopathic Interstitial Pneumonias (IIPs)

- IPF, marked by a high risk of progressive fibrosis and variable tempo of decline.
- Chronic IIPs: desquamative interstitial pneumonia, pleuroparenchymal fibroelastosis, idiopathic nonspecific interstitial pneumonia, etc.
- Acute IIPs: acute interstitial pneumonia.

- Subacute IIPs: cryptogenic organizing pneumonia.
- Unclassifiable ILD: cases that defy clear categorization due to mixed or atypical features.

Autoimmune-related ILD

- **Acute presentations:** rapidly progressive forms like diffuse alveolar hemorrhage in ANCA-associated vasculitis, severe lung involvement in systemic lupus erythematosus, or anti-MDA5 antibody-associated amyopathic dermatomyositis.
- **Chronic forms:** connective tissue disease-associated ILD (CTD-ILD), including systemic sclerosis, rheumatoid arthritis, idiopathic inflammatory myopathies (dermatomyositis, polymyositis), Sjögren's syndrome, and others. These are immune-mediated, with T- and B-cell-driven inflammation; genetics, environmental exposures, and other host factors contribute. ILD occurs in an estimated 15% of patients with CTDs, with higher prevalence in systemic sclerosis and rheumatoid arthritis. Risk skews toward women under 50 and coexisting comorbidities.

Exposure-related ILDs

- **Chronic:** Chronic exposure includes hypersensitivity pneumonitis from repeated inhalation of organic antigens (birds, mold), pneumoconioses from inorganic dusts, respiratory bronchiolitis-associated ILD, and postinfectious ILD. Prolonged inhalational insults drive persistent inflammation and fibrotic remodeling. Occupational contact with silica and asbestos is a major contributor: crystalline silica inhalation causes silicosis, a progressive fibrosing disease that persists despite exposure cessation and raises the risk of tuberculosis and lung cancer. Asbestos fibers induce asbestosis through ongoing alveolar injury and fibrosis and are also linked to pleural disease, mesothelioma, and bronchogenic carcinoma. Both produce restrictive physiology, reduced diffusing capacity, and worsening dyspnea.
- **Acute:** drug-induced or radiation-induced lung injury from agents such as certain chemotherapies, antiarrhythmics, antibiotics, or radiation can cause direct toxicity, hypersensitivity, or autoimmune-like injury.

Cystic or airspace-filling ILDs

- Lymphangioleiomyomatosis (LAM): abnormal smooth muscle proliferation, primarily in women of childbearing age.
- Pulmonary Langerhans cell histiocytosis: accumulation of pathological dendritic cells in the lung.
- Pulmonary alveolar proteinosis and microlithiasis: abnormal intra-alveolar accumulation of surfactant or calcium phosphate material.

ILDs associated with distinct primary diseases

- **Sarcoidosis:** multisystem granulomatous disorder with predominant pulmonary involvement. It features non-caseating granulomas driven by dysregulated macrophage and T-cell activation. Etiology is unclear; genetic factors (e.g., HLA-DRB1), environmental exposures (insecticides, mineral dusts), and host immune interactions modulate susceptibility and severity. Clinical presentations include cough, fever, breathlessness, weight loss, and bilateral hilar lymphadenopathy. Diagnosis requires concordant clinical, radiologic, and histologic evidence. Differential diagnoses include infections, malignancies, and autoimmune conditions. Endobronchial ultrasound-guided transbronchial needle aspiration is preferred for tissue sampling.

Other ILDs

Examples include chronic eosinophilic pneumonia, acute eosinophilic pneumonia, and malignancy-associated interstitial involvement (e.g., lymphangitic carcinomatosis).

- **ILD:** Group of disorders causing diffuse parenchymal lung pathology with varying degrees of inflammation and fibrosis.
- **Progressive fibrosing ILD (PF-ILD):** Subset showing decline in lung function, worsening fibrosis on imaging, or clinical deterioration despite standard care.
- **Acute exacerbation:** Sudden worsening

RISK FACTORS & TRIGGERS

Category	Details
Autoimmune activation	■ Connective tissue diseases and vasculitides drive lung inflammation via dysregulated T- and B-cell responses. ■ Immune profiling shows aberrant monocyte and lymphocyte patterns, with regulatory T cell expansion and a lung-blood recruitment axis linked to progressive disease.
Genetic predisposition	■ Variants affecting telomere maintenance (shortened telomeres), surfactant biology, and polymorphisms (e.g., MUC5B promoter, TOLLIP) increase susceptibility and influence phenotype, including familial pulmonary fibrosis.
Age and sex	■ Risk increases with age, especially >65 years, due to telomere shortening and reduced repair capacity. ■ Male sex has higher IPF risk; certain CTD-ILDs and lymphangioleiomyomatosis are more common in women, reflecting hormonal and biological differences.
Tobacco smoking	■ Strongly linked to RB-ILD, desquamative interstitial pneumonia, and pulmonary Langerhans cell histiocytosis. ■ Its role in IPF and RA-ILD is complex; high prevalence but unclear direct causality.

Tobacco smoking	■ Strongly linked to RB-ILD, desquamative interstitial pneumonia, and pulmonary Langerhans cell histiocytosis. ■ Its role in IPF and RA-ILD is complex; high prevalence but unclear direct causality.
Environmental & occupational exposures	■ Chronic inhalation of organic antigens (hypersensitivity pneumonitis), inorganic dust (silica, asbestos), fumes, gases, and pollutants triggers inflammation and fibrosis. ■ Air pollution (PM2.5, PM10, NO_2) is associated with onset and faster decline.
Drug & radiation injury	■ Certain chemotherapy agents, antiarrhythmics, antibiotics, and thoracic radiation cause direct toxicity, hypersensitivity reactions, or autoimmune-like lung injury, leading to acute/subacute ILD.
Infections & postinfectious injury	■ Certain chemotherapy agents, antiarrhythmics, antibiotics, and thoracic radiation cause direct toxicity, hypersensitivity reactions, or autoimmune-like lung injury, leading to acute/subacute ILD.
Gastroesophageal reflux (GERD)	■ Micro-aspiration may injure epithelium and is associated with IPF progression in observational studies.
Socioeconomic & contextual modifiers	■ Limited care access, delayed diagnosis, and environmental living conditions may influence presentation and outcomes, though contribution is less well quantified.
Trigger-host interaction	■ Triggers act on a susceptible background; removing or mitigating exposures (smoking, antigen exposure, offending drugs) is key in management.

EVALUATION FOR DIAGNOSIS

Category	Details
Symptoms	■ Progressive exertional dyspnea and persistent dry cough; may start subtly but worsen over weeks to months, limiting activity. ■ Fatigue and unintended weight loss are common.
Physical examination predisposition	■ Fine inspiratory crackles (“Velcro” sounds) at lung bases indicating early fibrosis. ■ Digital clubbing, especially in idiopathic pulmonary fibrosis (IPF), signals chronicity. ■ Look for systemic features (skin thickening, inflammatory arthritis, Raynaud’s) suggesting autoimmune-associated ILD.

History & risk assessment	■ Structured occupational, environmental, drug, and smoking history to identify reversible triggers.
Basic monitoring	■ Pulse oximetry at rest and exertion to detect oxygen desaturation and gauge functional limitation.
Laboratory tests	■ CBC and basic metabolic panel to check for comorbidities and alternative causes. ■ If CTD suspected, order targeted autoimmune serology (ANA, RF, anti-CCP, Scl-70, others as indicated).
Imaging	■ Chest X-ray may show reticular patterns or low lung volumes; ■ HRCT is key for phenotyping and subtype classification (UIP, NSIP, HP patterns, etc.).
Pulmonary function tests	■ Restrictive defect with reduced FVC and DLCO. ■ Six-minute walk test captures exertional desaturation and exercise tolerance.
Common comorbidities	■ GERD (up to 94% in IPF), pulmonary hypertension, cardiac disease, infections, thromboembolism, lung cancer, obstructive sleep apnea (60-90% in IPF), mood disorders. Screen and treat simultaneously.
Severity evaluation & specialist input	■ After diagnosis, assess severity, subtype, and impact. ■ Definite/probable IPF, NSIP, or CTD-ILD cases should undergo multidisciplinary team (MDT) review (pulmonology, radiology, pathology, rheumatology). ■ MDT guides therapy (antifibrotics, immunomodulators) and follow-up plans. Regular reassessment is essential.

CONFIRMATION OF DIAGNOSIS

Confirmation of an ILD diagnosis requires synthesis of clinical, radiologic, physiologic, and, when needed, pathologic data to arrive at a confident subtype and to exclude mimics.

Component	Purpose	Key elements / findings	When it may confirm (and avoid biopsy)	When to escalate / next step
Clinical history & exam	Frame likely subtype; exclude mimics	Symptom timeline; exposures (mold, birds, silica, drugs); autoimmune features; clubbing/crackles	Classic risk/exposure pattern aligning with imaging	Atypical course or mixed features then broaden labs, consider early multidisciplinary team (MDT) review
Pulmonary function tests (PFTs)	Quantify impairment	Restriction (↓ forced vital capacity, FVC); ↓ diffusing capacity (DLCO)	Supports ILD presence/severity alongside high-resolution computed tomography (HRCT)	Discordant with symptoms/ imaging get MDT review; repeat PFTs; assess for coexisting airway/cardiac disease
Basic labs	Exclude infection/ systemic illness	CBC, inflammatory markers; infection screen if febrile	Normal results with consistent HRCT can support non-infective ILD	Abnormal or septic picture - treat infection first; re-evaluate

Autoimmune serology	Detect connective-tissue disease-associated ILD	ANA, RF, anti-CCP, myositis panel, ENA as indicated	Positive serology + compatible HRCT + clinical features can establish subtype	Negative/indeterminate but suspicion persists then get rheumatology input; MDT
HRCT (first-line imaging)	Define pattern; stage disease	Usual patterns (e.g., basal/subpleural usual interstitial pneumonia, UIP; hypersensitivity pneumonitis signs)	Definite/probable UIP in right clinical context → working dx of idiopathic pulmonary fibrosis without biopsy	Indeterminate/atypical or discordant with clinic get MDT review; consider bronchoscopy/ biopsy
Exclude mimics	Prevent misdiagnosis	Heart failure, infection, drug toxicity, environmental/ occupational disease	Clear alternative cause found → manage that condition	Cause unclear after workup – MDT review, targeted tests
Bronchoalveolar lavage (BAL)	Support/ exclude etiologies	Infection rule-out; lymphocytosis (HP), eosinophilia (EP) patterns	When BAL clarifies infection/HP and aligns with HRCT	Non-diagnostic or conflicting - consider biopsy
Transbronchial lung cryobiopsy	Tissue with lower morbidity	Larger samples than forceps; good yield in experienced centers	Can secure diagnosis in selected patients, avoiding surgery	Contraindicated/low yield expected - proceed to surgical lung biopsy
Surgical lung biopsy (VATS/ open)	Gold standard tissue diagnosis	Multilobe targeted sampling	When required diagnostic certainty achieved	Use when cryobiopsy unsuitable or nondiagnostic; weigh risks carefully
MDT discussion	Integrate all data; assign confidence	Radiology, pathology, pulmonology, rheumatology	Radiology, pathology, pulmonology, rheumatology	Low confidence after MDT - plan further testing or close follow-up with re-assessment
Diagnostic confidence & follow-up	Decide therapy vs more tests	Definite/probable vs possible	Definite/probable → start phenotype-appropriate treatment and monitoring	Low confidence after MDT - plan further testing or close follow-up with rPossible/ uncertain and biopsy declined/ unsafe - adopt working diagnosis, remove triggers, close interval reviewe-assessment

DIFFERENTIAL DIAGNOSIS

Differential diagnosis must systematically exclude other conditions that can mimic or coexist with ILD, because management and prognosis differ substantially.

Condition	Key Clinical Clues	Diagnostic Differentiators
Heart failure (CHF)	Dyspnea, basilar crackles, history of cardiac disease, peripheral edema	Elevated JVP, elevated BNP, echocardiography showing reduced EF or diastolic dysfunction, pulmonary vascular redistribution on CXR
Chronic obstructive pulmonary disease (COPD)	Exertional breathlessness, cough, smoking history	Spirometry: reduced FEV1/FVC, hyperinflation, preserved/increased lung volumes; imaging shows emphysema, not fibrosis; assess bronchodilator responsiveness
Pulmonary infections (TB, atypical pneumonias)	Cough, fever, weight loss, imaging changes	Microbiology: sputum culture/microscopy, NAAT for TB, serology/PCR for atypicals; systemic signs (fever, leukocytosis); temporal evolution; postinfectious scarring patterns
Pulmonary vasculitis (e.g., GPA, MPA)	Cough, hemoptysis, systemic features (skin lesions, renal involvement)	Positive ANCA, abnormal renal function, urinalysis with active sediment, biopsy confirmation, multisystem involvement
Pulmonary edema (cardiac or renal origin)	Rapid onset breathlessness, orthopnea, signs of fluid overload	Elevated filling pressures, renal dysfunction, CXR with interstitial/alveolar edema, rapid improvement with diuresis, echo findings
Obstructive airway disease with atypical presentation (asthma with fixed obstruction, bronchiolitis)	Variable breathlessness, wheeze, cough	PFT: obstruction with preserved DLCO; bronchodilator challenge; imaging with air trapping (not reticulation)

MANAGEMENT GOALS

1. Slow Disease Progression - Preserve lung function via early phenotype identification and timely initiation of disease-modifying therapy.
2. Control Inflammation - Suppress active immune-mediated injury to prevent irreversible fibrosis in inflammatory phenotypes.
3. Improve Symptoms - Reduce dyspnea, cough, and fatigue; improve exercise tolerance through optimized treatment and rehabilitation.
4. Prevent Acute Exacerbations - Identify and address triggers (infection, aspiration, medication changes) early.
5. Enhance Quality of Life - Address physical, emotional, and social needs; minimize treatment burden; support shared decision-making.
6. Manage Comorbidities - Prevent their impact on disease trajectory and avoid confounding assessment.

MANAGEMENT PRINCIPLES

1. Precise Phenotyping - Determine if ILD is predominantly fibrosing (e.g., IPF, progressive fibrosing ILD) or inflammatory (e.g., CTD-ILD, hypersensitivity pneumonitis) to guide therapy.
2. Address Modifiable Factors - Remove or reduce exposures (smoking, offending drugs, occupational/environmental antigens) and treat systemic drivers such as autoimmune disease.
3. Targeted Disease-Modifying Therapy
    a. Fibrosing phenotypes: Antifibrotics to slow FVC decline.
    b. Inflammatory phenotypes: Immunosuppression (steroids ± steroid-sparing agents) tailored to activity and risk.
4. Multidisciplinary Approach - All key treatment decisions made with MDT input, considering comorbidities, frailty, and patient preferences.
5. Supportive Care - Pulmonary rehabilitation, oxygen therapy when indicated, vaccination, nutritional support, symptom control, psychosocial support, and advance care planning.
6. Comorbidity Management - Systematic screening and treatment of pulmonary hypertension, GERD, infections, sleep-disordered breathing, and cardiovascular disease.
7. Ongoing Monitoring - Serial PFTs, oxygen needs, imaging when indicated, and patient-reported outcomes to detect progression or adverse effects.
8. Escalation/De-escalation Pathways - Intensify therapy or re-refer to MDT if decline or diagnostic uncertainty; consider de-escalation in stable patients.
9. Integrated Care - Coordinate with cardiology, rheumatology, palliative care, and other relevant specialties.

NON-PHARMACOLOGICAL INTERVENTIONS

Non-pharmacological interventions are essential across all ILD etiologies and stages; they complement disease-modifying therapy, address complications, and preserve function and quality of life.

- Prevention begins with minimizing additional lung injury: Immunizations - annual influenza and appropriate pneumococcal vaccines reduce risk of superimposed respiratory infections that can precipitate exacerbations.
- Early mobilization in hospitalized or acutely unwell patients prevents muscle loss and preserves endurance.

- Nutritional assessment and support are critical, since chronic inflammation and increased breathing work predispose to weight loss and cachexia; optimizing caloric and protein intake helps maintain muscle mass and resilience.
- Smoking cessation must be actively supported through counseling and pharmacologic aids if needed; even in fibrosing disease, stopping tobacco reduces ongoing insult and may improve responsiveness to other therapies.
- Comorbidities and frailty amplify disability; systematic screening for conditions such as gastroesophageal reflux, sleep-disordered breathing, cardiovascular disease, and depression allows targeted management that indirectly benefits pulmonary status.

PHARMACOLOGICAL THERAPY

- **Phenotype-driven:** Treat fibrosing ILD with antifibrotics; treat inflammatory/autoimmune ILD with immunomodulators; use disease-specific agents where indicated.
- **Team-based decisions:** Choose therapy within a MDT; reassess response, tolerability, and evolving phenotype regularly.
- **Balance risks:** Match treatment to disease activity and progression rate, considering comorbidities and drug risks.
- **Overlap/uncertain cases:** Use combination or sequential strategies (e.g., add antifibrotic when fibrosis progresses despite immunosuppression) after MDT review.
- **Ongoing monitoring:** Track efficacy and adverse effects; adjust dose, switch, or add second-line agents as needed.

Indication / Phenotype	Drug / Class	Mechanism / Role	Usual Dose	Duration / Continuation	Key Monitoring / Precautions
Fibrosing ILD / IPF / Progressive fibrosing phenotype	Nintedanib	Tyrosine kinase inhibitor; slows fibroblast activation and FVC decline	150 mg orally twice daily (reduce to 100 mg twice daily if intolerance)	Continuous long-term until unacceptable toxicity or clear progression despite therapy	LFTs before start and periodically; manage diarrhea proactively; caution with bleeding (especially if on anticoagulants)
	Pirfenidone	Anti-fibrotic with anti-inflammatory effects	Titrated to 2403 mg/day in three divided doses (per label escalation)	Continuous long-term; interruptions ≥14 days require re-titration	LFTs prior and during therapy; photosensitivity (sun protection); GI side effects; drug interactions (CYP1A2)

Systemic sclerosis-associated ILD / other progressive fibrosing ILDs	Nintedanib	Same antifibrotic role; approved for SSc-ILD and other PFF	150 mg twice daily (adjust if needed)	Continuous as for IPF, guided by MDT and disease behavior	Same as above; assess liver function and GI tolerance
Autoimmune / inflammatory ILD (CTD-ILD, hypersensitivity pneumonitis, vasculitis)	Prednisolone (or equivalent)	Broad immunosuppression to control active inflammation	0.5-1 mg/kg/day initial, then gradual taper based on response	High-dose induction for weeks to months, then taper to lowest effective or discontinue if stable (often supplemented with steroid-sparing agent)	Blood glucose, blood pressure, bone protection, infection risk, adrenal suppression
	Mycophenolate mofetil	Inhibits lymphocyte proliferation; steroid-sparing	1,000-1,500 mg twice daily orally	Maintenance long-term; often started early to facilitate steroid taper	CBC, renal function, GI intolerance, infection surveillance
	Azathioprine	Purine synthesis inhibitor; steroid-sparing	1-2 mg/kg/day (after TPMT activity if available)	Maintenance: used to sustain remission and reduce steroid exposure	TPMT prior if feasible; CBC, liver enzymes, infection risk
	Cyclophosphamide (IV pulses)	Potent immunosuppression in rapidly progressive disease	500-1,000 mg/m² IV monthly (specialist-led)	Induction over 3-6 months, then transition to maintenance agent	Hematologic toxicity, hemorrhagic cystitis (MESNA prophylaxis), fertility impact, infection
	Rituximab	B-cell depletion for refractory or severe autoimmune ILD	Two doses of 1,000 mg 2 weeks apart; repeat per protocol (often every 6 months based on B-cell recovery)	Cyclical; guided by clinical response and B-cell monitoring	B-cell counts, infection risk (HBV reactivation), infusion reactions
ANCA-associated / vasculitis-related ILD	Cyclophosphamide or Rituximab	Induction of remission	Cyclophosphamide pulses or rituximab regimen as above	Induction typically 3-6 months, followed by maintenance (e.g., azathioprine, rituximab spacing)	See respective drug monitoring

Lymphangioleiomyomatosis (LAM)	mTOR inhibitor (e.g., Sirolimus/ Everolimus)	Inhibits abnormal smooth muscle proliferation	Sirolimus targeting trough 5-15 ng/mL	Continuous while disease is active/ progressive; may be re-evaluated if stable long-term	Lipid profile, renal function, mouth ulcers, infection risk
Pulmonary alveolar proteinosis (PAP)	GM-CSF (inhaled or subcutaneous)	Immune modulation to restore surfactant clearance	Variable protocols (daily or alternate dosing)	Course individualized; often weeks to months with reassessment	Monitor clinical/ physiologic response, injection-site reactions, infection
	Whole lung lavage (non-pharmacologic)	Mechanical clearance of accumulated surfactant	Procedural	As needed based on symptomatic recurrence and physiologic impairment	Periprocedural respiratory support; risk of transient hypoxemia
Sarcoidosis with significant pulmonary involvement	Prednisolone ± steroid-sparing (methotrexate/ azathioprine)	Anti-inflammatory / immunosuppression	Prednisolone 20-40 mg/day initial, then gradual taper; methotrexate weekly 7.5-25 mg	Induction (months) followed by taper to lowest effective dose; steroid-sparing agent often continued longer-term	Steroid adverse effects; methotrexate liver and hematologic monitoring

Systemic corticosteroids (e.g., Prednisolone): Initial: 0.5-1 mg/kg/day (usually up to 60 mg) with taper based on clinical response. **Cautions:** Hyperglycemia, hypertension, infection risk, bone protection required if prolonged use

Steroid-sparing immunosuppressants (depending on subtype)

- Mycophenolate mofetil: 1,000-1,500 mg twice daily orally (adjust for tolerance)
- Azathioprine: 1-2 mg/kg/day after TPMT activity check if available
- Cyclophosphamide: Reserved for severe/refractory disease (IV pulses, specialist-managed)
- Rituximab or tacrolimus in select autoimmune etiologies (specialist decision)

SYMPTOMATIC MANAGEMENT

Provide parallel supportive care to reduce disability and maintain quality of life while disease-modifying therapy is underway.

Dyspnea: Optimize underlying ILD therapy.

- Oxygen: titrate to keep SpO2 ≥90% (rest/exertion); trial ambulatory oxygen if exertional desaturation limits activity.
- Teach breathing/pacing techniques (pursed-lip, diaphragmatic breathing) via pulmonary rehabilitation.
- Address anxiety/panic with brief behavioral support; consider low-dose anxiolytics only if needed.

Cough: Exclude reversible causes (infection, reflux, upper airway).

- Use suppression techniques, ensure hydration, and treat reflux aggressively.
- For refractory cough, refer for specialist evaluation; consider neuromodulators or speech therapy.

Fatigue / exercise intolerance:

- Enroll in pulmonary rehabilitation (graded exercise, education).
- Encourage activity pacing and optimize sleep.
- Screen/treat contributors (anemia, depression).
- Provide nutritional support to prevent or reverse cachexia.

Comorbidity management:

- Screen for and treat gastroesophageal reflux, sleep-disordered breathing, pulmonary hypertension, and cardiac disease.
- Integrate their control into symptom relief.

Infection prevention:

- Vaccinate annually for influenza and per schedule for pneumococcus.
- Early evaluation and appropriate treatment of respiratory infections to avoid exacerbations.

Advanced / palliative care:

- Identify psychosocial distress, cachexia, and functional decline early.
- Introduce palliative principles: symptom prioritization, advance care planning, and supportive counseling.

Monitoring:

- Regularly reassess symptoms (dyspnea, cough, fatigue) with patient-reported tools and functional measures.

- Adjust supportive interventions based on change or progression.

Escalate to specialist/MDT if symptoms worsen despite optimized symptomatic care or if new red flags appear as below:

Pharmacologic symptom control in the palliative care of ILD

Symptom / Indication	Drug	Starting Dose	Duration / Titration	Key Cautions / Monitoring
Refractory dyspnea	Morphine (oral immediate release)	2.5-5 mg every 4 hours as needed	Start low; titrate slowly based on relief and side effects; maintenance or sustained-release formulations individualized	Monitor for sedation, respiratory depression, constipation (prophylaxis), renal/hepatic impairment; adjust in frail or opioid-naive
Chronic cough (if refractory and noninfectious)	Low-dose morphine	2.5 mg every 4-6 hours	Short to intermediate term; titrate to symptom control	Same as for dyspnea; watch for accumulation in renal dysfunction
Chronic cough / neuropathic component	Gabapentin	100 mg at night; increase by 100 mg every 2-3 days to 300 mg two-three times daily	Maintenance as needed; dose reduction in renal impairment	Dizziness, somnolence, peripheral edema; adjust for renal function
Anxiety / panic (acute)	Lorazepam	0.5 mg once or twice daily	Short course; avoid long-term dependence	Risk of respiratory depression in severe ILD, cognitive impairment, falls in elderly; avoid in sleep apnea without caution
Anxiety / depression (chronic)	Sertraline	25-50 mg once daily	Start low; assess response over 4-6 weeks; continue long term if benefit	GI upset, hyponatremia (elderly), drug interactions (other serotonergic agents), initial activation
Excess airway secretions	Glycopyrrolate	1-2 mg orally two-three times daily or 0.2-0.4 mg subcutaneously	As needed or scheduled	Dry mouth, urinary retention, constipation, confusion in elderly/frail
Nausea / vomiting	Ondansetron	4 mg orally or IV every 8 hours	Short-term; adjust for hepatic function	QT prolongation (caution with other QT-prolonging drugs), constipation
Nausea (prokinetic)	Metoclopramide	10 mg orally or IV every 6-8 hours	Short course (usually <4-6 weeks)	Extrapyramidal symptoms, especially in young or elderly; avoid in Parkinsonism, tardive dyskinesia risk with prolonged use
Acute inflammatory symptom flare / appetite support	Prednisone (low dose)	10-20 mg once daily	Short-term trial; taper based on response	Hyperglycemia, immunosuppression, muscle weakness, osteoporosis with longer use; monitor blood pressure/glucose

Red flag signs in ILD (warrant urgent evaluation)

- Sudden or rapid worsening of dyspnea over days to weeks (possible acute exacerbation)
- New or high-grade fever, especially with increased cough or sputum (infection)
- Acute onset chest pain or pleuritic pain (consider pulmonary embolism, pneumothorax, cardiac ischemia)
- Hemoptysis (could signal diffuse alveolar hemorrhage, infection, malignancy, vasculitis)
- Significant desaturation at rest or with minimal exertion (drop in SpO2 ≥4% or to <88%)
- New-onset or worsening cyanosis, confusion, or altered mental status (severe hypoxemia)
- Syncope or presyncope (suggests pulmonary hypertension or arrhythmia)
- Signs of right heart strain/failure: peripheral edema, elevated jugular venous pressure, hepatojugular reflux
- Rapid decline in pulmonary function (e.g., sharp fall in FVC or DLCO) not explained by adherence or obvious trigger
- Acute pleuritic findings with asymmetric breath sounds (possible pneumothorax)
- New arrhythmia or palpitations in context of ILD (may reflect cardiac involvement or pulmonary hypertension)
- Weight loss or cachexia accelerating despite stable therapy (could indicate malignancy, uncontrolled systemic disease, or severe deconditioning)

Action: Any of these should prompt expedited clinical review, pulse oximetry/ABG, imaging (chest X-ray/HRCT), and specialist input. Early differentiation between infection, exacerbation, thromboembolism, cardiac causes, and other complications is critical.

ACUTE EXACERBATION OF ILD (AE-ILD)

An acute exacerbation is a rapid, clinically significant respiratory deterioration over ≤1 month, with new bilateral ground-glass opacities or consolidation on imaging, not fully explained by cardiac failure, fluid overload, or an alternative cause. The presentation includes sudden worsening dyspnea, increased oxygen requirement, and hypoxemia.

Initial assessment and diagnostic steps:

Promptly evaluate:

- History/exam to exclude extrapulmonary causes.
- Pulse oximetry/arterial blood gas.
- Chest HRCT to confirm new diffuse alveolar changes and rule out pneumothorax, pulmonary embolism, or other mimics.
- Basic labs, infection screen (blood/sputum cultures, viral panels), and cardiac evaluation to exclude heart failure.
- Identify possible triggers (infection, recent surgery/biopsy, drug exposure); classify as triggered versus idiopathic exacerbation.

Immediate management:

- Admit to appropriate care level; continuous monitoring of oxygenation and vital signs.
- Provide supplemental oxygen to maintain target saturation (typically ≥90%) while avoiding hypercapnia in susceptible patients.
- Empiric broad-spectrum antibiotics if infection cannot be excluded; de-escalate based on results.
- Consider high-dose corticosteroids (institutional protocol varies, e.g., methylprednisolone 0.5-1 g daily for 3 days or prednisone equivalent), recognizing evidence is limited and decision is individualized via MDT.
- Evaluate for reversible contributors (volume overload, thromboembolism) and correct.
- Avoid routine mechanical ventilation unless goals of care justify escalation; if used, discuss prognosis and consider lung-protective strategies. Non-invasive support (e.g., high-flow oxygen or CPAP) may be trialed carefully.

Adjunct and escalation:

- Consider advanced immunomodulatory strategies (e.g., tacrolimus, cyclosporine, rituximab with plasma exchange/IVIG) only in select cases after specialist/MDT review.
- Continue or reassess background antifibrotic therapy; do not abruptly stop unless contraindicated.
- Early involvement of palliative care for goal alignment given high mortality.

Monitoring and follow-up:

- Frequent reassessment of respiratory status, oxygen requirements, and infection parameters.
- Repeat imaging only if clinical trajectory is unclear or complication suspected.
- Once stabilized, re-evaluate baseline therapy, optimize, and plan closer follow-up to detect recurrence.

AE-ILD carries high short-term mortality. Decisions about invasive support, ICU admission, or escalation should be made in the context of baseline function, trajectory, patient values, and advance care planning. Early recognition and supportive management offer the best chance of recovery; prevention strategies include prompt treatment of infections and close monitoring (including remote/home spirometry where feasible) to detect early decline.

HYPERSENSITIVITY PNEUMONITIS

- Trigger avoidance (primary)
- Corticosteroids for moderate/severe inflammation

LONG-TERM OXYGEN THERAPY

Long-term oxygen therapy is indicated for patients with resting hypoxemia (SpO2 ≤88%) or significant desaturation on exertion; it alleviates dyspnea, improves exercise tolerance, and may reduce pulmonary hypertension progression. Criteria for home LTOT see below.

Home long-term oxygen therapy (LTOT):

- Indicated (room air, stable state):
  - Resting SpO_2 ≤88% or PaO_2 ≤55 mmHg.
  - PaO_2 56-59 mmHg (or SpO_2 89%) with cor pulmonale/right-heart failure, pulmonary hypertension, or hematocrit >55%.
  - Exertional desaturation to ≤88% (e.g., on 6-minute walk) that improves with oxygen.
  - Nocturnal desaturation to ≤88% for ≥5 min with symptoms or pulmonary hypertension.
- **Prescription:** use ≥15 h/day (include sleep); titrate to keep SpO_2 ≥90% at rest, with exertion, and during sleep.
- **Recheck:** confirm stability before starting (usually 2-3 weeks after an acute event); reassess at 1-3 months and after any clinical change.
- **Safety/notes:** strict no-smoking (fire risk); educate on device use and maintenance; monitor for hypercapnia in at-risk patients.

PULMONARY REHABILITATION

Pulmonary rehabilitation is a structured, multidisciplinary program for people with ILD aimed at improving exercise capacity, symptom control, and quality of life in patients with exertional dyspnea, deconditioning, and reduced functional tolerance despite optimized medical therapy.

- It combines supervised exercise training (endurance and strength), breathing retraining, education on self-management for disease, and psychosocial support.
- Key components: personalized exercise prescription, teaching techniques like pacing and diaphragmatic or pursed-lip breathing, nutrition counseling to counter cachexia, and coping strategies for anxiety/fatigue.
  Benefits include improved six-minute walk distance, reduced breathlessness perception, better mood, and increased ability to perform daily activities.
- Referrals for pulmonary rehabilitation should be early at diagnosis or on first signs of functional decline and prioritized for those with moderate limitations (e.g., mMRC grade ≥2 or reduced six-minute walk).
- Contraindications or precautions include unstable cardiac disease, recent acute exacerbation, or severe hypoxemia without proper oxygen support; programs should monitor oxygen saturation and adjust supplemental oxygen during sessions.
- Regular reassessment ensures the regimen stays aligned with evolving capacity.

LUNG TRANSPLANTATION

Lung transplantation is the definitive treatment for selected patients with advanced, progressive ILD who continue to worsen despite optimized therapy.

- Indications include significant physiological decline (≥10% relative drop in FVC or ≥15% in DLCO over 6 months), severe resting or exertional hypoxemia (SpO2 <88%), reduced exercise capacity (6-minute walk distance <250 m or a fall of ≥50 m over 6 months), pulmonary hypertension, or frequent exacerbations.
- Lung transplantation is generally considered for patients under 65 years of age, though there is no absolute age cutoff. Candidates between 65 and 70 may be accepted depending on physiological fitness, comorbidities, and psychosocial support, while those over 70 are rarely considered due to increased surgical risks and reduced long-term survival. The International Society for Heart and Lung Transplantation (ISHLT) guidelines emphasize individualized assessment over chronological age, focusing on functional status and overall health (Leard et al., J Heart Lung Transplant, 2021).

- Referral should happen early before irreversible decline, so candidacy can be evaluated and modifiable factors optimized. Evaluation and listing often take months to years, creating emotional and logistical strain for patients and families.
- Eligibility requires adequate overall health, absence of prohibitive comorbidities (active malignancy, uncontrolled infection, severe frailty, major extrapulmonary organ dysfunction, or extreme obesity in some centers), and psychosocial readiness; relative contraindications include ongoing smoking and poor adherence potential.
- Pre-transplant workup includes cardiopulmonary assessment, infection screening, nutritional optimization, and rehabilitation. Post-transplant care involves lifelong immunosuppression, surveillance for rejection and infection, and ongoing rehabilitation.
- Outcomes can be durable: roughly two-thirds of IPF recipients survive beyond three years and about half beyond five. Benefits include marked improvement in dyspnea, exercise tolerance, and quality of life. Risks include chronic lung allograft dysfunction, infections, and complications from immunosuppression.
- Management from candidacy through postoperative care should be coordinated by a multidisciplinary transplant center within a shared-care framework.
- Patients who are not transplant candidates because of age, severe obesity, malnutrition, or other contraindications should have the focus shifted to aggressive symptom control, advance care planning, and comfort measures (dyspnea pacing, optimized oxygen therapy, and psychosocial support).

PALLIATIVE CARE

Clinicians should introduce palliative care as a standard part of ILD management beginning at diagnosis and run alongside disease-modifying and supportive treatments. It is a holistic, patient- and family-centered approach focused on identifying and alleviating physical, psychological, social, and spiritual distress to improve quality of life.

- Structured tools (see below) should be used to assess symptom burden and goals, coordinate across specialties to avoid fragmentation, and actively communicate the purpose and benefits to reduce stigma. The intended outcomes are reduced symptom severity, care aligned with patient values, fewer unnecessary interventions or hospitalizations, and better support for patients and families throughout the illness trajectory.
- Early integration ensures proactive symptom control, dyspnea pacing, opioid use for refractory breathlessness, cough management, fatigue mitigation, and addresses anxiety, depression, and existential concerns through counseling and psychosocial support.

- Advance care planning, including clear goals of care, documentation of patient preferences, and contingency plans for escalation or de-escalation, should be initiated early and revisited as the disease evolves.
- Caregivers should receive support, including preparation for bereavement, and the entire process operates within a shared-care model coordinated among pulmonology, palliative specialists, nursing, and allied providers.
- Refer to palliative services n case of uncontrolled symptoms, frequent exacerbations or hospitalizations, progressive functional decline or frailty, complex decision points (such as transplant candidacy or treatment withdrawal), or explicit patient/family requests for clarity on prognosis.
- Hospice-level care should be considered when life expectancy is estimated at six months or less, particularly in rapidly progressive phenotypes like advanced IPF or when disease-modifying options are exhausted.
- Barriers such as limited awareness among clinicians and patients, discomfort discussing prognosis, and scarcity of trained palliative providers must be anticipated and addressed through education and normalization of palliative involvement.

Structured tools should be used to assess symptom burden and goals

Priority	Tool	Primary use	When to use
1	SPICT - Supportive and Palliative Indicators Tool	Flag needs for palliative input	At first assessment, repeat if condition changes
1	NECPAL - Necesidades Paliativas tool	Identify patients with unmet palliative needs	As above (alternative to SPICT)
1	"Surprise Question" ("Would I be surprised if this patient died in the next 12 months?")	Early trigger for palliative review	At intake and major reviews
2	IPOS - Integrated Palliative care Outcome Scale	Core symptom/concern tracker (incl. breathlessness)	Every visit or monthly
3	ESAS-r - Edmonton Symptom Assessment System, revised	Symptom severity profiling	Every visit or monthly
4	mMRC - modified Medical Research Council Dyspnea Scale	Quick dyspnea grade	Every visit
5	Serious Illness Conversation Guide	Goals/values and preferences	Early and at key inflection points
6	ACP - Advance Care Planning forms; POLST/MOST - Physician/Medical Orders for Life-Sustaining Treatment	Document decisions; reduce unwanted care	After goals talk; review periodically
7	PHQ-9 - Patient Health Questionnaire-9; GAD-7 - Generalized Anxiety Disorder-7; HADS - Hospital Anxiety and Depression Scale	Screen mood/anxiety	Baseline, then every 3-6 months or if symptoms

8	K-BILD - King's Brief Interstitial Lung Disease questionnaire; SGRQ - St George's Respiratory Questionnaire; EQ-5D - EuroQol-5 Dimensions	Quality of life and function	Baseline, then 3-6 monthly
9	6MWT - Six-Minute Walk Test (with oximetry)	Functional capacity and exertional desaturation	Baseline and every 3-6 months
10	Dyspnea-12; UCSD SOBQ - University of California, San Diego Shortness of Breath Questionnaire	Detailed dyspnea characterization	When mMRC/IPOS suggests high burden
11	Shared care plan templates; MDT checklists; crisis ('just-in-case') breathlessness plans	Coordination, avoid fragmentation, support caregivers	After baseline set-up; update at each review

Goals of care in ILD palliative care (for Advance Care Planning)

- **Purpose:** align all treatment with the patient's values as how they balance longevity, function, and comfort across stable disease, exacerbations, and end-of-life.
- **Core decisions to document:**
  - Symptom priorities (breathlessness, cough, anxiety, sleep).
  - Ceiling of care: home vs hospital/ICU; non-invasive ventilation acceptable or not; views on intubation/ventilation and cardiopulmonary resuscitation.
  - Use/continuation of antifibrotics or immunosuppression if disease advances.
  - Oxygen targets, rescue plans for acute breathlessness (fan, opioids, benzodiazepines), and steroids/antibiotics during suspected acute exacerbation of ILD.
  - Preferred place of care and dying; who to contact first.
  - Surrogate decision-maker/medical power of attorney and how to reach them.
- **Contingency plans:** clear triggers for escalation (e.g., severe hypoxemia, new chest pain, infection) and for de-escalation (refractory symptoms, patient preference for comfort-focused care), including just-in-case medications at home.
- **Review points:** at diagnosis, after any hospitalization or major change in function/ oxygen need, at least annually, and whenever preferences change.
- **Outputs:** signed Advance Care Planning form where available POLST (Physician Orders for Life-Sustaining Treatment)/MOST (Medical Orders for Scope of Treatment); shared care plan in the record and with family, primary care, pulmonology, and palliative teams.

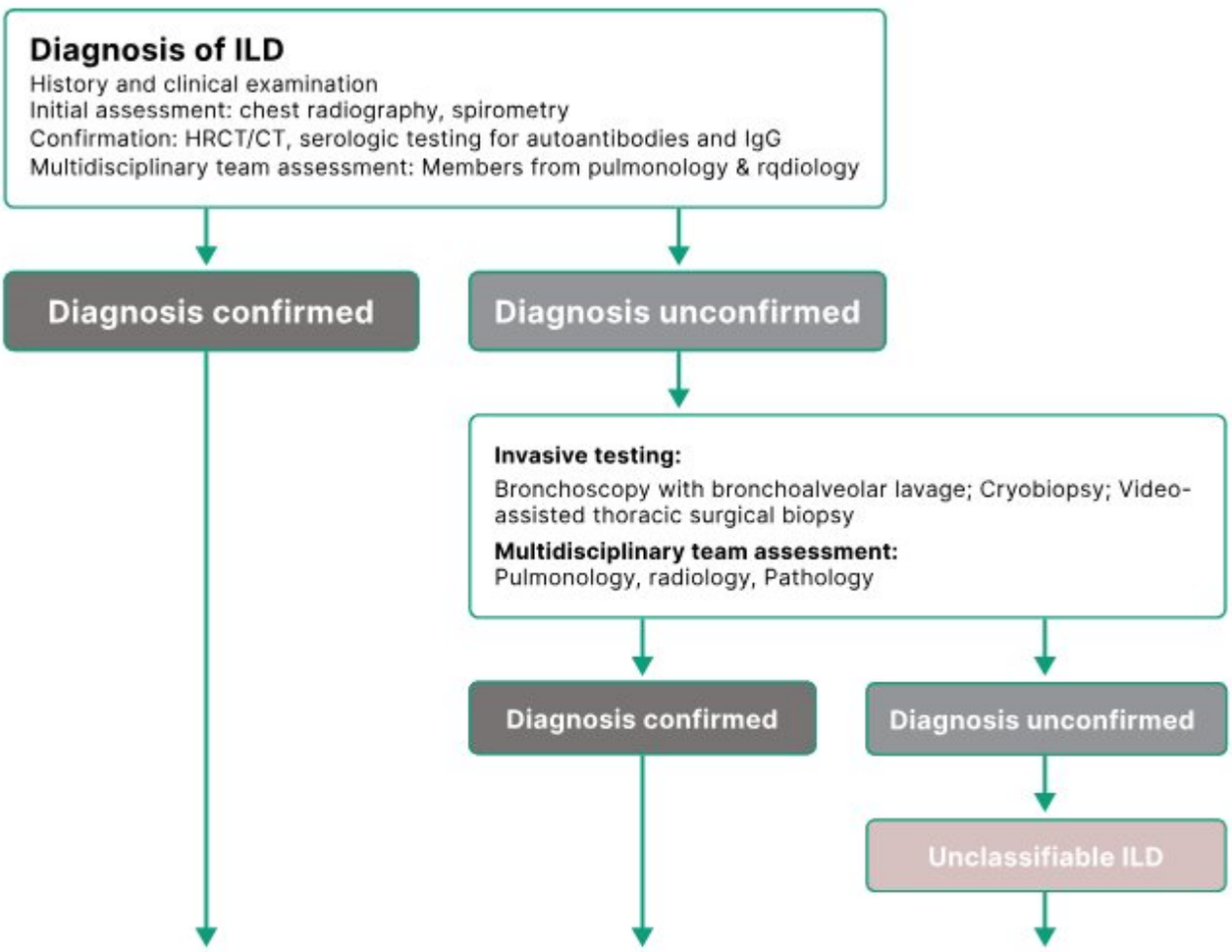
ASSESSMENT OF RESPONSE

Assessment of response in ILD combines objective measures and symptom monitoring to detect progression, treatment effect, or complications early. Integration of the data in multidisciplinary review ensures timely adjustment of therapy or escalation of care. Review and follow-up in ILD are risk-based.

Area	What to measure / tool	Thresholds that trigger action	Next step
Symptoms & function	mMRC dyspnea scale, cough scores, patient-reported fatigue; 6MWT	Worsening 6MWT distance or higher mMRC grade	Reassess diagnosis, triggers, and treatment plan
Pulmonary function	FVC and DLCO every 3-6 months	≥10% relative fall in FVC or ≥15% fall in DLCO	Review phenotype; consider therapy escalation or change
Oxygen needs	Resting and exertional SpO_2; home oxygen flow rates	New resting desaturation, higher exertional need, or rising flow requirement	Re-titrate oxygen; evaluate for progression/exacerbation
Imaging	HRCT only for significant change	Unexpected decline, atypical symptoms, or suspected complication	HRCT to clarify cause; update management
Acute exacerbations	Sudden dyspnea, new/ worse hypoxemia, rapid decline	Any acute change	Urgent evaluation; exclude infection, pulmonary embolism, cardiac causes; treat promptly
Follow-up cadence	Clinic review frequency	Stable: every 3-6 months; progressive/unstable: monthly-quarterly	Adjust interval to disease behavior and access
Before escalating therapy	Check adherence, comorbidities, infections, exposure removal	Progression confirmed by symptoms, PFTs, oxygen need, or imaging	Escalate within Multidisciplinary Team (MDT) framework
Before reducing immunosuppression	Ensure stability window	Stable 6-12 months	Gradual taper; close monitoring for relapse
MDT oversight	Multidisciplinary Team review (pulmonology, radiology, rheumatology, pathology as available)	Any major treatment change or diagnostic uncertainty	Decide on escalation/de-escalation, additional tests, and follow-up plan

SCHEMATIC FLOWCHART OF THE DIAGNOSIS, ASSESSMENT, AND TREATMENT PATHWAY FOR ILD.

Diagnosis. Assessment & Treatment Of Interstitial Lung Disease (ILI))



REFERRAL CRITERIA AND TIERED APPROACH

Referral follows a tiered approach based on suspicion, severity, and complexity.

Primary care / island clinic:

- Suspected ILD (chronic unexplained dyspnea with crackles) → initial evaluation, chest X-ray, basic labs, pulse oximetry → early referral to secondary center or central hub if available.
- Acute exacerbation or significant desaturation → urgent referral.

Secondary care (regional hospital):

- Arrange HRCT if feasible
- Begin initial supportive therapy (oxygen, symptom management)
- Initiate corticosteroids for inflammatory phenotypes
- Coordinate with central pulmonology/tertiary center for antifibrotic decisions

Tertiary care / specialist center (likely in Malé or via Tele consult):

- Multidisciplinary review (radiology, pulmonology, rheumatology)
- Confirm diagnosis (consider biopsy if indeterminate)
- Start disease-modifying agents (antifibrotics, complex immunosuppression)
- Manage complications, consider advanced care planning.

PROGNOSIS AND PROGRESSION

Prognosis in ILD varies by subtype, patient factors, comorbidities, and tempo of decline. Poorer outcomes are linked to older age, male sex (especially in IPF), rapid loss of lung function (≥10% relative decline in FVC or ≥15% in DLCO over 6 months), reduced exercise capacity, pulmonary hypertension, and untreated comorbidities. In IPF, survival is roughly 88% at 1-2 years but drops to about 30% beyond five years.

Progression, especially in progressive fibrosing ILD (PF-ILD), is defined by a combination of worsening respiratory symptoms, physiological decline (absolute FVC drop >5% or DLCO >10% in 12 months), and radiologic evidence of increasing fibrosis (new or worsening traction bronchiectasis, reticulation, honeycombing, or ground-glass with fibrotic features). Trial-based frameworks (e.g., INBUILD, RELIEF, uILD) use these criteria, sometimes in composite form (e.g., FVC decline, 6MWD drop, or death),

to identify clinically meaningful progression.

Since individual trajectories remain unpredictable and validated biomarkers are lacking, serial monitoring of symptoms, lung function, and targeted imaging is essential for timely risk stratification and management adjustments.

COMPLICATIONS

ILD carries several serious complications. Early recognition, regular surveillance, and proactive management of these complications are essential to limit downstream morbidity and guide timely escalation of care.

Complication	Why it happens / impact	Red flags & screening	First steps in management	Notes
Acute exacerbation of ILD	Sudden diffuse alveolar injury on background fibrosis → high mortality	Rapid ↑ dyspnea, new/worse hypoxemia; CXR/HRCT with new bilateral opacities	Urgent workup for infection/PE/MI; bloods, cultures, viral panel, CT pulmonary angiography if indicated; start oxygen, treat trigger; consider corticosteroids per local protocol	Admit if moderate-severe; early MDT input
Pulmonary hypertension (PH)	Chronic hypoxic vasoconstriction and vascular remodeling → ↓ exercise capacity, worse prognosis	Unexplained desaturation on exertion, loud P2, edema; screen with echocardiography; confirm with right-heart cath in selected cases	Optimize oxygen (SpO_2 ≥90%), diuretics for right-heart failure; refer to PH/ILD specialist for targeted therapy decisions	Manage comorbid sleep apnea and thromboembolism
Progressive parenchymal loss → respiratory failure	Ongoing fibrosis reduces gas exchange	Rising oxygen needs, resting desaturation, declining 6MWT/FVC/DLCO	Titrate LTOT; pulmonary rehab; discuss escalation limits and advance care planning	Consider non-invasive ventilation for selected overlap conditions; early palliative involvement
Secondary infections (bacterial/viral/fungal)	Impaired lung defense ± immunosuppression; can mimic or trigger exacerbation	Fever, purulent sputum, focal changes; raised inflammatory markers; new lobar changes on imaging or positive microbiology	Prompt cultures and targeted antimicrobials; hold or reduce immunosuppression case-by-case; supportive care	Vaccinate (influenza, pneumococcal, COVID-19); review exposure risks
Right-ventricular (RV) dysfunction/failure	Chronic hypoxia/PH overloads RV → edema, hepatomegaly, ascites	↑ JVP, peripheral edema, hepatojugular reflux; echo shows RV strain	Oxygen, cautious diuretics, salt/fluid review; manage PH; consider cardiology input	Watch renal function and blood pressure with diuresis

Treatment-related toxicity	Antifibrotics/ immunosuppressants → hepatotoxicity, cytopenias, infections	LFT derangements, low counts, infection signs; drug-specific adverse effects	Routine labs (LFTs, CBC), dose adjust/ hold drug; manage adverse events; prophylaxis where indicated (e.g., PJP)	Educate on early symptom reporting; reconcile meds each visit

PREVENTION AND PROMOTION

Prevention and promotion in ILD aim to reduce new cases and mitigate progression can shift care upstream, catching disease earlier and lowering preventable morbidity.

- Public awareness campaigns should highlight avoidance of known inhalational antigens, especially for hypersensitivity pneumonitis, and promote smoking cessation.
- Clinicians must routinely review and rationalize prescriptions of pneumotoxic drugs, stopping or substituting when risks outweigh benefits.
- Frontline providers need training to recognize early symptoms such as persistent unexplained dyspnea, dry cough, crackles so that referrals happen before advanced damage.
- Vaccination drives (influenza, pneumococcus) and infection control reduce triggers and exacerbations. Public awareness about avoidance of known inhalational antigens (especially in hypersensitivity pneumonitis)

PATIENT EDUCATION

Objectives of patient education in ILD include:

- Help patients understand the disease: ILD is chronic; progression can often be slowed but not always reversed, and treatment goals focus on preservation of function and symptom control.
- Emphasize consistent adherence to prescribed medications and keeping scheduled follow-ups to monitor disease behavior and adjust therapy.
- Train patients to recognize red flags such as sudden worsening of breathlessness, new fevers, increased cough or fatigue so they seek timely care.
- Support and motivate smoking cessation as a modifiable driver of lung injury.
- Teach safe use of supplemental oxygen: proper delivery, flow settings, handling, and precautions (fire safety, equipment maintenance).

- Identify and avoid known environmental or occupational triggers (dust, mold, bird exposure, irritants) that can exacerbate inflammation or fibrosis.
- Reinforce the importance of vaccinations (influenza, pneumococcal) to prevent respiratory infections that can destabilize ILD.

Instructions to Patient/Caregiver

Do	Don't
■ Take all medicines on schedule; ask before changing doses.	■ Don't stop steroids or antifibrotic therapy abruptly.
■ Continue antifibrotic or steroid plans unless your clinician advises otherwise.	■ Don't ignore new or worsening symptoms.
■ Attend every follow-up, even when you feel well.	■ Don't smoke or allow smoking near oxygen equipment.
■ Maintain regular, clinician-approved activity, pace and rest.	■ Don't overexert to the point of severe breathlessness or prolonged recovery.
■ Use supplemental oxygen exactly as prescribed, check flow, tubing, and device function.	■ Don't self-medicate with over-the-counter anti-inflammatories or antibiotics without guidance.
■ Keep strict fire safety with oxygen: no flames, no smoking, keep devices well-ventilated.	■ Don't delay care after an acute change because a visit was "already scheduled."
■ Avoid known triggers at home/work; improve ventilation; use masks if exposure is unavoidable.	
■ Get annual influenza vaccine and pneumococcal vaccines per schedule.	
■ Bring a list of all medicines to visits and report side effects early.	

Red flags - seek urgent care

- Sudden increase in breathlessness, new resting hypoxemia, or rapid drop in oximetry.
- New fever, purulent sputum, chest pain, hemoptysis, or marked fatigue.

Caregiver tips

- Learn oxygen setup and emergency steps; check supplies.
- Help track symptoms (simple diary or phone notes).
- Know the preferred hospital/contacts and bring advance-care documents to visits.

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