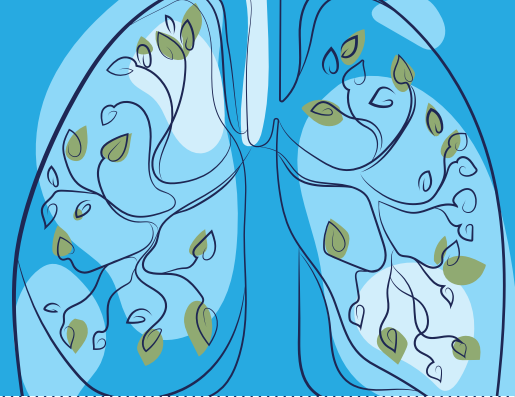


Addressing the Burden of PH-ILD

Early Recognition, Evidence-Based Management, and Care Coordination



RAPID RECAP

The Increased Clinical Burden of Pulmonary Hypertension and Interstitial Lung Disease (PH-ILD)¹⁻⁵

- The estimated prevalence of PH in ILD ranges widely depending upon the underlying cause of ILD, disease severity, and diagnostic modality used
- In patients with IPF, the prevalence of PH varies depending on disease severity
 - 8%-15% at initial diagnosis
 - 29%-46% at evaluation for lung transplant
 - 86% at lung transplant
- ILD patients with PH have significantly reduced 6MWT distance, DLCO, and oxygen saturation nadir (SpO₂) compared to patients without PH
- Patients with PH-ILD have significantly increased utilization of health care resources, including inpatient admissions and costs, following an initial diagnosis of PH
- Relative to other forms of PH, patients with PH-ILD have increased mortality

Early Diagnosis of PH-ILD and the Importance of Proactive Screening^{6,7}

Risk Factors

- History of pulmonary edema or heart failure (HF)
- Dizziness, palpitations, and syncope

Signs

- Altered heart sound (loud P2 or S2)
- Ankle swelling/peripheral edema
- Jugular venous distension
- Signs of right HF
- Hepatomegaly/ascites

Predictive Index

- The FORD (FVC/DLco, Oxygen saturation, Race, Distance) calculator and FORD index are simple, validated tools incorporating noninvasive parameters that can be applied to identify patients with IPF at risk of PH who may benefit from invasive testing
- Parameters include race, 6MWT distance, FVC/DLCO percent predicted ratio, and SpO₂
- Echocardiography does not always accurately predict PH in patients with IPF
- Consensus approach to screening



ATS

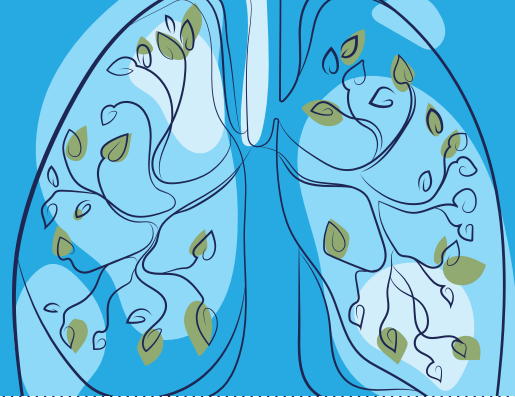
American
Thoracic
Society



The
France
Foundation

Addressing the Burden of PH-ILD

Early Recognition, Evidence-Based Management, and Care Coordination

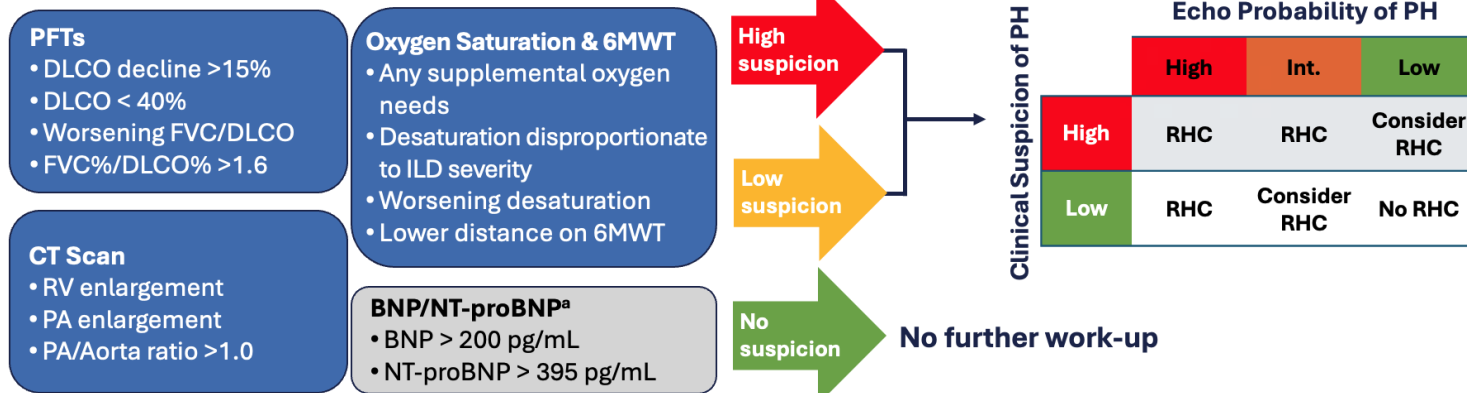


RAPID RECAP

Early Diagnosis of PH-ILD and the Importance of Proactive Screening^{6,7}

Tests

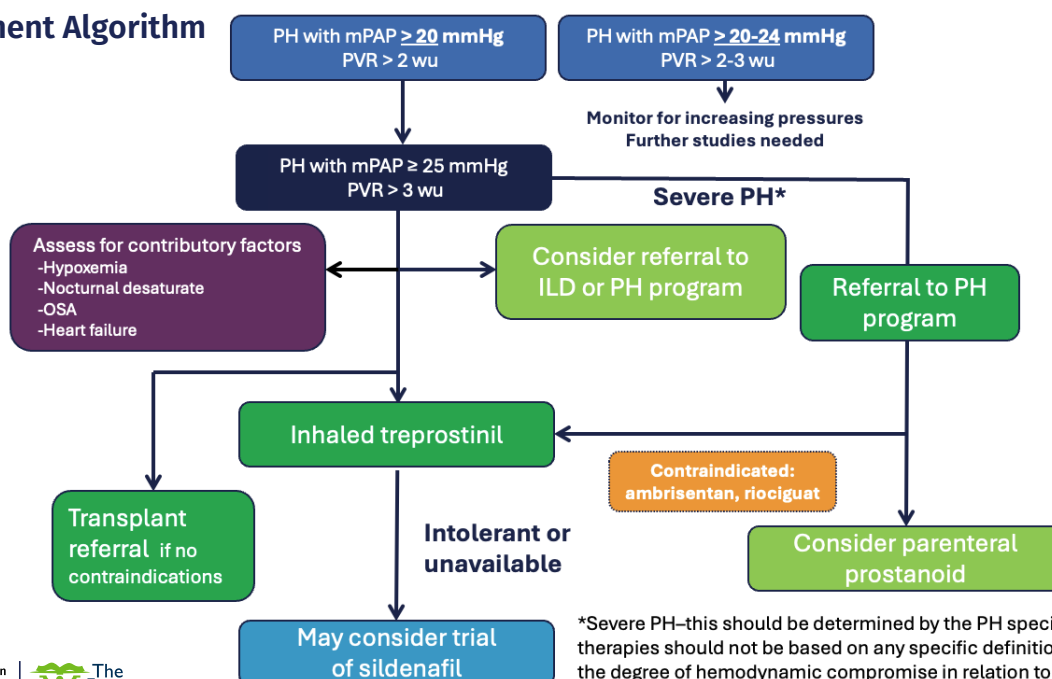
(used in concert to risk stratify for PH)



- Tests that are routinely obtained in patients with ILD
- Not routinely obtained, but considered if there is suspicion for underlying HF or PH in patients with ILD

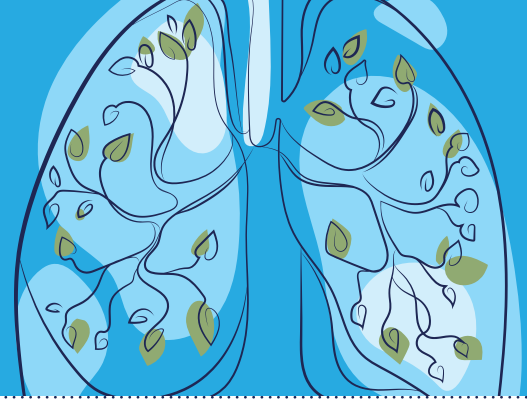
Assessing the Clinical Data and Practice Guidelines⁸

Treatment Algorithm



Addressing the Burden of PH-ILD

Early Recognition, Evidence-Based Management, and Care Coordination



RAPID RECAP

Key Takeaways

- PH-ILD is underappreciated and underdiagnosed
- PH-ILD has a dismal prognosis
- Screening involves looking at routine ILD testing through a different lens
- Clinicians should have a low threshold for RHC to identify PH in patients with ILD
- Inhaled treprostinil is the one approved therapy for PH-ILD
- Multimodality therapy for PH is a critical component of management of IPF and other ILDs

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