Mapping the Diagnostic Journey of Patients With Pulmonary Hypertension Associated With Interstitial Lung Disease (PH-ILD): A Retrospective U.S. Claims Analysis

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Background

- PH-ILD is a severe, progressive, and life-limiting cardiopulmonary condition characterized by elevated pulmonary artery pressure in concert with interstitial inflammation and fibrosis[1]
- Timely diagnosis is difficult and often delayed owing to nonspecific and frequently overlapping symptoms between ILD and PH, a low index of suspicion among non-PH specialists, and lack of standardized diagnostic algorithms for WHO Group 3 PH[1,2]
- This study characterizes the diagnostic journey of PH-ILD patients using real-world claims data, focusing on the diagnosis of PH in patients with ILD

ILD, interstitial lung disease; PH, pulmonary hypertension; WHO, World Health Organization.

1. Nathan SD, et al. *J Manag Care Spec Pharm*. 2025;31(1-a suppl):S1-S18. 2 de Belen E, et al. *J Am Heart Assoc*. 2023;12(2):e026413.

Methods

- A retrospective cohort study was conducted using the NorstellaLinQ fully integrated claims and electronic health record dataset from January 2019 to December 2024

Inclusion Criteria

- Adults with ≥1 claim for ILD and ≥1 claim for PH
- PH-ILD was defined using ICD-10 codes indicative of both ILD and PH, excluding other PH groups

Exclusion Criteria

- COPD or emphysema
- CV conditions associated with WHO Group 2 PH
- Heart failure in medical record prior to PH diagnosis

- Time intervals were calculated from symptom presentation to PH diagnosis
- The diagnosing physician’s specialty, as well as diagnostic procedures, were examined

COPD, chronic obstructive pulmonary disease; CV, cardiovascular; ILD, interstitial lung disease; PH, pulmonary hypertension; WHO, World Health Organization.

Patient cohort selection

Level 1:
Ensures patient has pre-capillary PH

n=1,968,570

- Patients with ≥1 diagnosis claim of I27.0, I27.21, 127.23 AND ≥1 procedure code from list ***OR***
- Patients with ≥2 diagnosis claims of I27.20 (unspecified) AND ≥1 procedure code from list

Level 2:
Ensures patient does NOT have post-capillary PH

n=264,530

Excluded patients with diagnoses of:

- Diastolic dysfunction or valvular heart diseases
- Heart failure that precedes the diagnosis of PH

Level 3:
Ensures patient DOES have PH-ILD

n=34,441

Included ONLY patients with ≥1 ILD codes

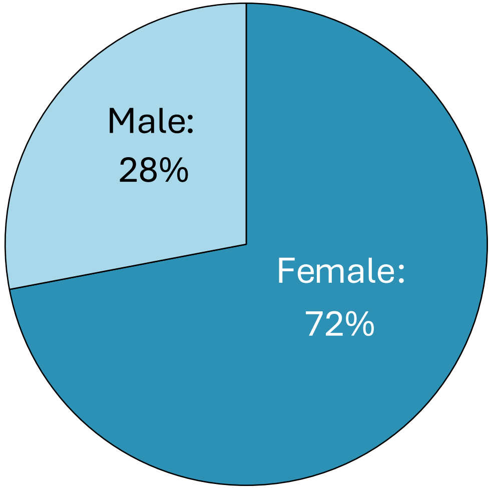
n=18,666
Final PH-ILD cohort

Excluded patients with COPD or emphysema codes

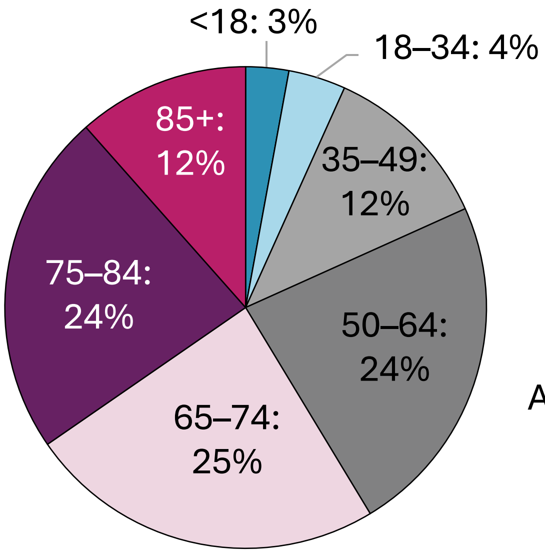
COPD, chronic obstructive pulmonary disease; ILD, interstitial lung disease; PH, pulmonary hypertension.

Patient demographics

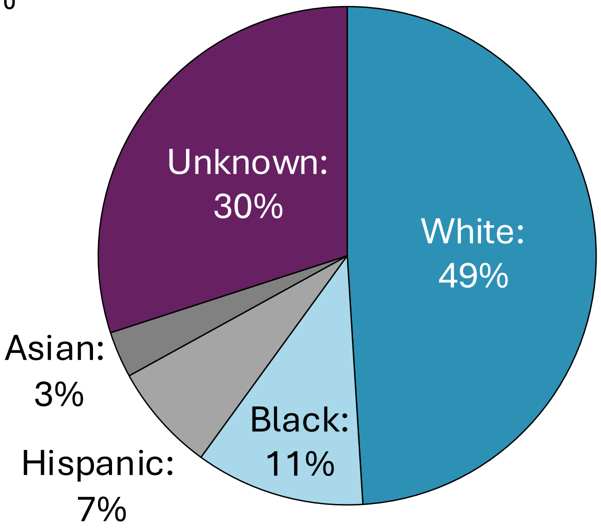
- Among 18,666 PH-ILD patients identified, the majority were female, White, and age ≥65 years



Gender



Age, years



Race

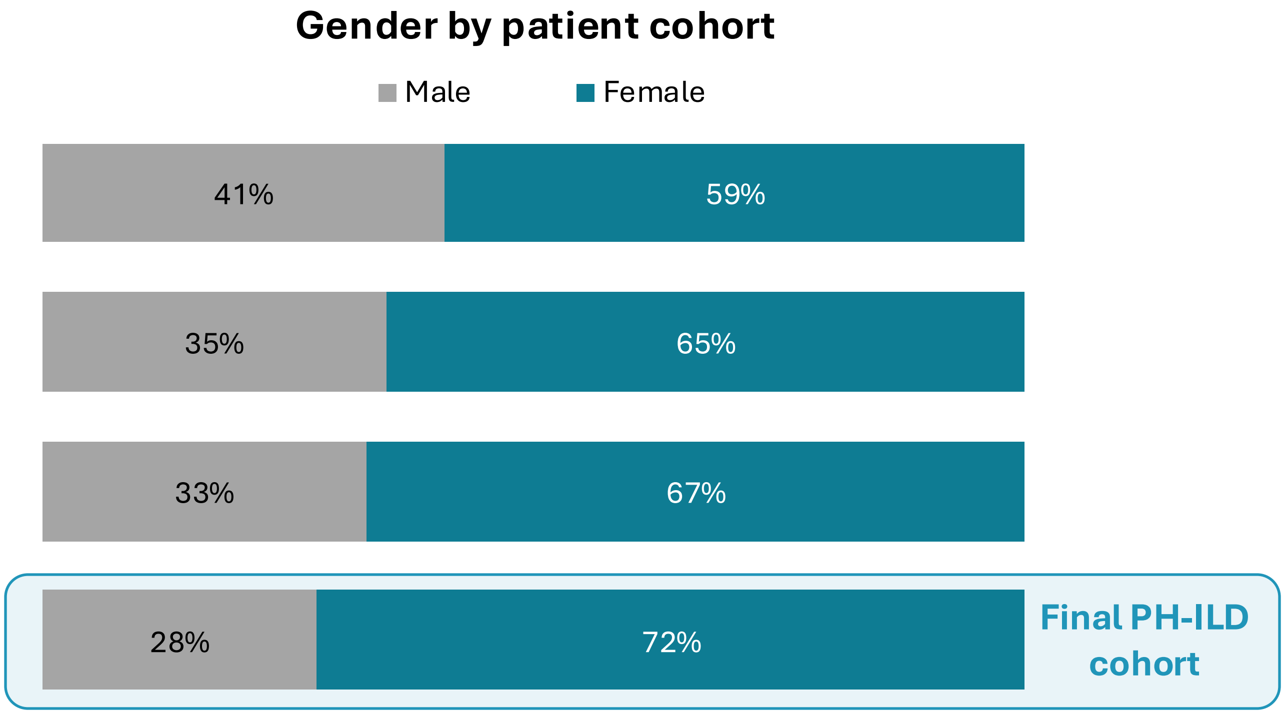
ILD, interstitial lung disease; PH, pulmonary hypertension.

There was an increase in female predominance at each step of the patient cohort funnel

Level 1 — Patient has **precapillary PH** n=1,968,570

Level 2 — Patient does **NOT** have **post-capillary PH** n=264,530

Level 3 — Patient DOES have **PH-ILD** n=34,441; Patient does **NOT** have **COPD** or **emphysema** n=18,666



Within the claims analysis cohort, a large proportion were classified as having CTD-ILD (33%)

ILD Subtype	Patients, n (%)*
Connective tissue disease	6,218 (33)
IPF	1,881 (10)
Toxin	1,112 (6)
Hypersensitivity pneumonitis	304 (2)
Occupational / environmental	164 (1)
Radiation	76 (0.4)
Other ILDs[a]	10,118 (54)

*Patients may have had more than one type of ILD diagnosed; [a]Other IIP was defined as patients with ICD-10 code J84, not including J84.112 (IPF), including other interstitial pneumonias.
ILD, interstitial lung disease; IPF, idiopathic pulmonary fibrosis.

Expected patterns of ILD demographics

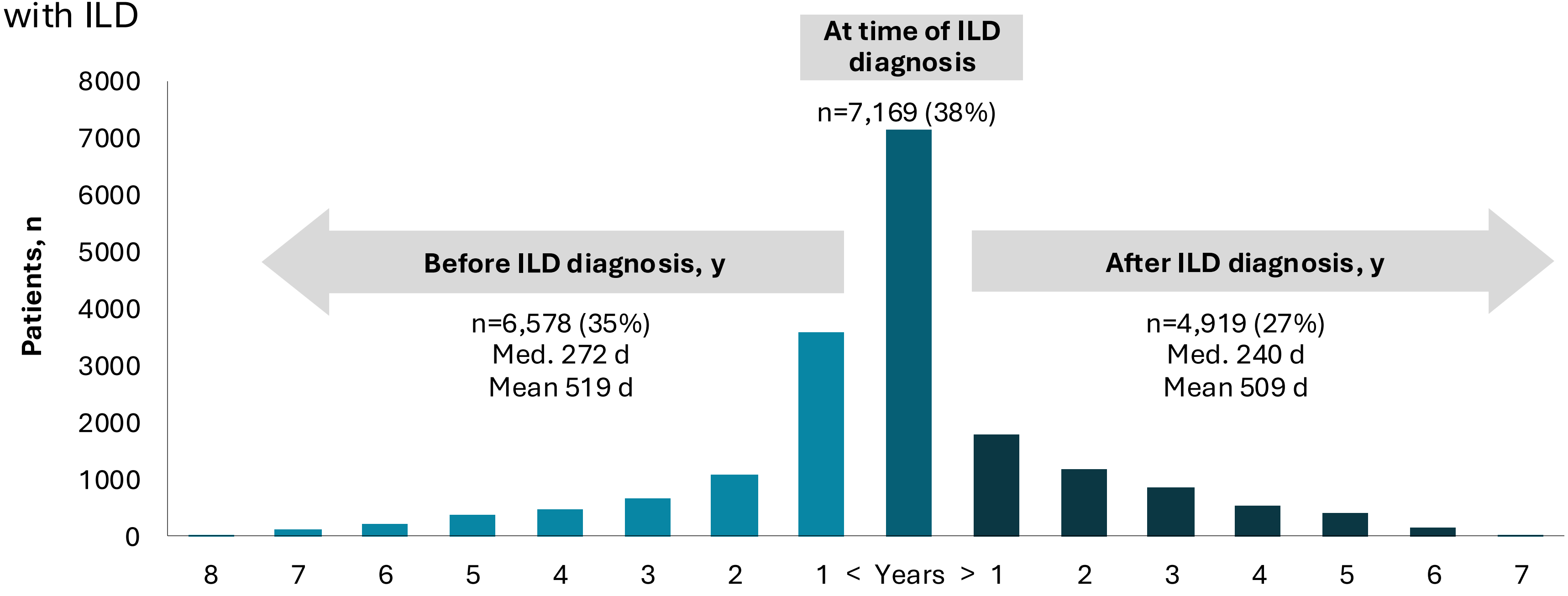
- IPF accounts for ~ 1/3 of ILD cases and is strongly male-predominant, which has historically shaped perceptions of ILD demographics[1]
- However, most other ILD subtypes—including CTD-ILD, IPAF, NSIP, and fibrotic hypersensitivity pneumonitis—skew female due to their autoimmune associations[2–8]
- Patients with emphysema, including CPFE, which exhibits a male predominance of 9:1,[9] were excluded from this cohort
- These patterns may explain why this PH-ILD cohort was predominantly female, driven by the high proportion of non-IPF ILDs

1. Maher TM. *JAMA*. 2024;331(19):1655-1665. 2. Gezgin T, et al. *Eur Research J*. 2023;9(6):1360-1367. 3. Kaul B, et al. *Front Med (Lausanne)*. 2021;8:751181. 4. Fernandes L, et al. *Front Med (Lausanne)*. 2019;6:209. 5. Sebastiani M, et al. *Biomedicines*. 2020;9(1):17. 6. Belloli EA, et al. *Respirology*. 2016;21(2):259-268. 7. Creamer AW, Barratt SL. *Eur Respir Rev*. 2020;29(156):190167. 8. Torres-Rojas MB, et al. *Pulm Circ*. 2025;15(1):e70035. 9. Jankowich MD, Rounds SIS. *Chest*. 2012;141(1):222-231.

CPFE, combined pulmonary fibrosis and emphysema; CTD, connective tissue disease; ILD, interstitial lung disease; IPAF, interstitial pneumonia with autoimmune features; IPF, idiopathic pulmonary fibrosis; NSIP, non-specific interstitial pneumonia.

PH diagnosis occurrence relative to ILD diagnosis

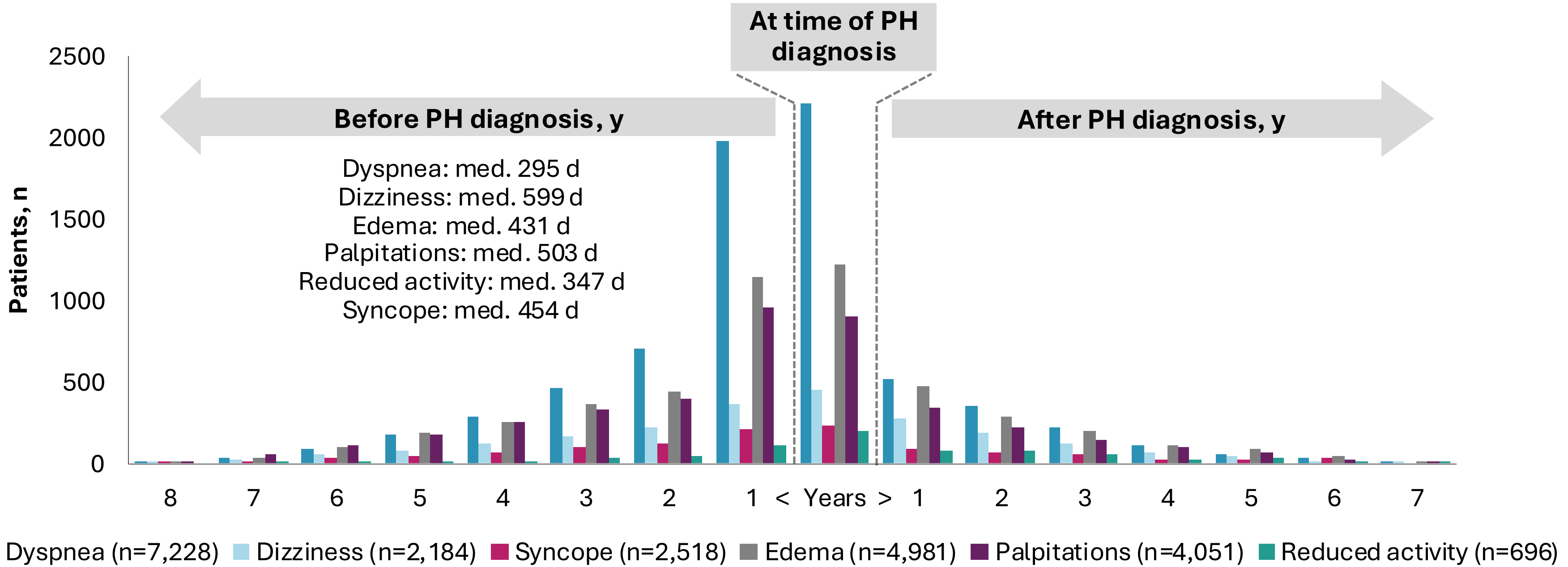
Most patients (65%) received a PH diagnosis at the same time as, or after, being diagnosed with ILD



d, day; ILD, interstitial lung disease; med, median; n, number; PH, pulmonary hypertension; y, year.

Symptom occurrence in the years prior to and after PH diagnosis

The most common symptoms preceding PH diagnosis were dyspnea (20%), dizziness (14%), edema (14%), and palpitations (12%)



d, day; med; median; n, number; PH, pulmonary hypertension; y, year.

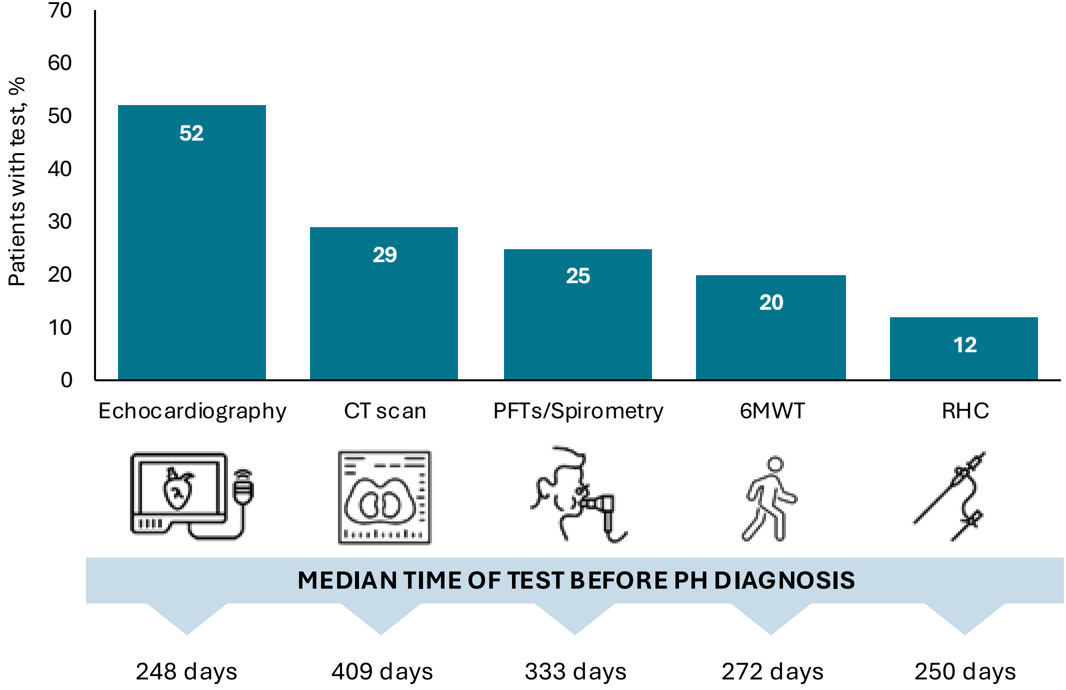
Specialties of HCPs who diagnosed PH

A substantial percentage of non-experts are diagnosing PH.

HCPs diagnosing PH-ILD	Patients, n (%)
Diagnosed by PH-focused specialties	**10,189 (55)**
Cardiology	5,371 (29)
Pulmonology	3,610 (19)
Rheumatology	1,070 (6)
Other	53 (<1)
Diagnosed by non-PH-focused specialties[a]	**8,477 (45)**

[a]Family/internal medicine (21%), nurse practitioner-other (4%), hospitalist (3%), radiology-diagnostic (3%), critical care (3%), pediatrics (2%).
HCP, healthcare professional; ILD, interstitial lung disease; n, number; PH, pulmonary hypertension.

Most common diagnostic tests performed before or in the same year as PH diagnosis



- 22% of patients used supplemental oxygen before or in the same year as PH diagnosis

6MWT, six-minute walk test; CT, computed tomography; DLCO, diffusing capacity of the lungs for carbon monoxide; PFT, pulmonary function test; PH, pulmonary hypertension; RHC, right heart catheterization.

Pulmonologists most frequently ordered diagnostic tests or supplemental O_2 (except for echocardiography)

Diagnostic test or supportive measure administered by specialist type

Echocardiography
- Cardiologists 23%
- Pulmonologists 18%

CT scan
- Pulmonologists 21%
- Cardiologists 21%

Right heart catheterization
- Pulmonologists 29%
- Cardiologists 15%

Spirometry
- Pulmonologists 29%
- Cardiologists 21%

Supplemental oxygen use
- Pulmonologists 26%
- Cardiologists 18%

6-minute walk test
- Pulmonologists 34%
- Cardiologists 19%

CT, computed tomography; PH, pulmonary hypertension.

Limitations

- Patients had ≥1 claim for PH and ≥1 claim for ILD but there was no requirement for confirmatory diagnosis claims for each condition
- Generic ILD codes may contribute to the lower-than-expected percentage of IPF patients in this cohort
- No physician chart review was performed to differentiate WHO Group 3 PH from WHO Group 1 PH (for example, in patients with CTD)
- Sarcoidosis was not explicitly excluded, and therefore some Group 5 PH patients may have been included in this cohort

CTD, connective tissue disease; ILD, interstitial lung disease; PH, pulmonary hypertension; WHO, World Health Organization.

Conclusions

- This analysis offers insights into symptom presentation, physician specialties, and modalities used for PH-ILD diagnosis
- The female predominance in this cohort may be explained by the high representation of CTD and other non-IPF ILD patients and the exclusion of CPFE patients in the analysis
- There was a delay of up to 1.6 years between initial symptom presentation and PH diagnosis, likely attributed to the overlapping symptomatology of PH and ILD
- A high percentage of diagnoses was made by non-PH specialists in acute and primary care settings, contributing to potential additional delays in referral to appropriate specialty care centers
- Key diagnostic modalities—including RHC, PFTs, and 6MWT—remain underutilized and may represent missed opportunities for comprehensive evaluation and timely intervention in this high-risk population
- Opportunities exist to accelerate disease awareness, recognition, and referral to reduce diagnostic delays and optimize outcomes for this high-risk population

6MWT, 6-minute walk test; CPFE, combined pulmonary fibrosis and emphysema; ILD, interstitial lung disease; IPF, idiopathic pulmonary fibrosis; PFT, pulmonary function test; PH, pulmonary hypertension; RHC, right heart catheterization.