Improving our understanding of progressive fibrosing interstitial lung diseases (ILDs): design of the ILD-PRO™ Registry

IPF-PRO™ ILD-PRO™

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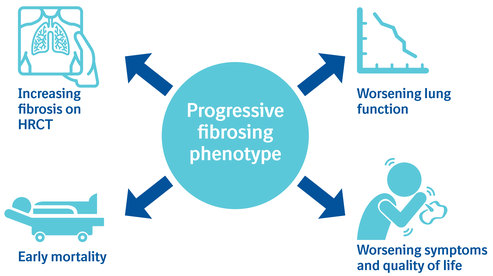
INTRODUCTION

- Idiopathic pulmonary fibrosis (IPF) is a chronic fibrosing ILD that is always progressive. A subset of patients with other chronic fibrosing ILDs also develop a progressive phenotype characterized by increasing fibrosis on HRCT; worsening lung function, symptoms and quality of life; and early mortality.[1,2]



- In 2018, the Idiopathic Pulmonary Fibrosis Prospective Outcomes (IPF-PRO) Registry,[3] an observational US registry involving 1002 patients with IPF, was expanded to include an additional arm comprising patients with non-IPF chronic progressive fibrosing ILDs to form the IPF-PRO/ILD-PRO Registry.

AIM

- To describe the objectives and design of the ILD-PRO Registry.

CONCLUSIONS

- The IPF-PRO/ILD-PRO Registry will provide a better understanding of the natural history of chronic progressive fibrosing ILDs, their impact on patients, and current practices in their diagnosis and management.
- Collection of biological samples will provide the opportunity to discover diagnostic, prognostic and theragnostic biomarkers.

STUDY DESIGN

Objectives of ILD-PRO Registry

- Describe current approaches to the diagnosis of chronic progressive fibrosing ILDs.
- Examine the natural history of chronic progressive fibrosing ILDs and their impact on patients, including on their quality of life.
- Describe interactions with the healthcare system and treatment practices among patients with chronic progressive fibrosing ILDs.
- Investigate disease biomarkers using biological samples linked to clinical data.

Design of ILD-PRO Registry

- The ILD-PRO Registry will enroll patients with chronic progressive fibrosing ILDs other than IPF at over 45 sites across the US.

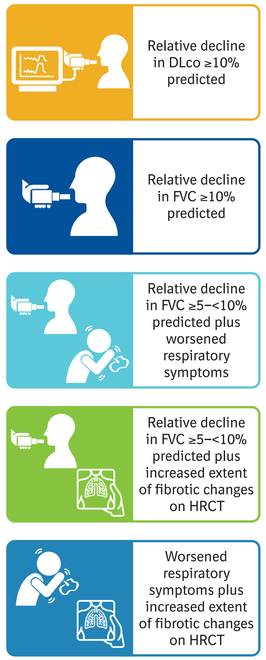
- Patients aged ≥30 years with a non-IPF ILD of any duration that was diagnosed or confirmed at the enrolling center, and reticular abnormality and traction bronchiectasis (with or without honeycombing) confirmed by HRCT scan and/or lung biopsy, are eligible to participate.

Non-IPF ILDs include (but are not limited to):

- Idiopathic non-specific interstitial pneumonia
- Unclassifiable idiopathic interstitial pneumonia
- Autoimmune ILDs (e.g. related to rheumatoid arthritis, systemic sclerosis, or Sjögren's syndrome)
- Chronic hypersensitivity pneumonitis
- Sarcoidosis-related ILD
- Exposure-related ILDs

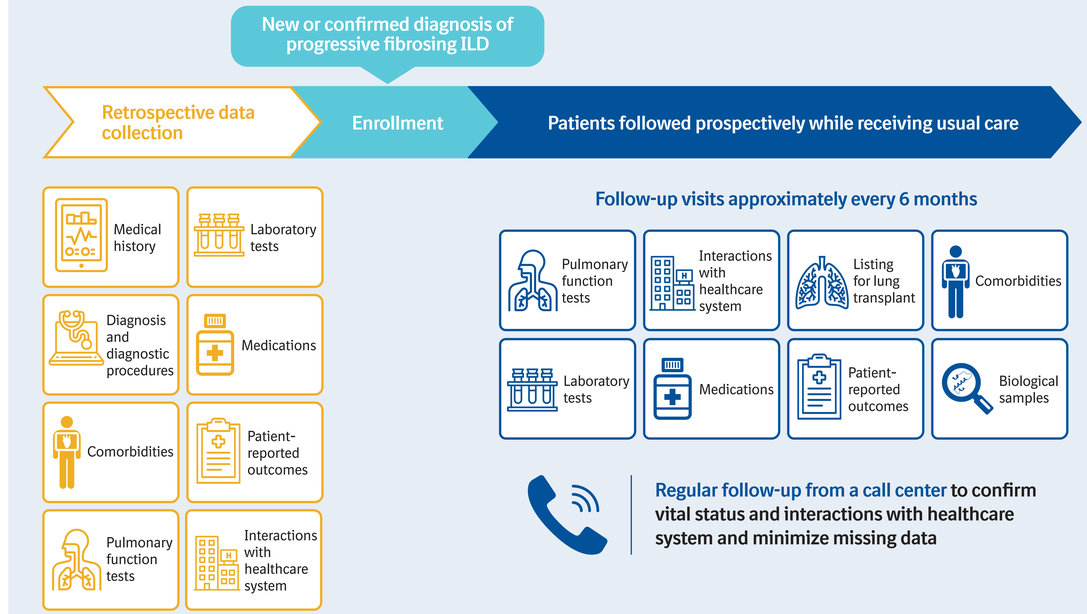
(e.g. asbestosis, coal workers' pneumoconiosis)

- Patients must meet ≥1 of the following criteria for ILD progression in the past 24 months:



Data collection

- Retrospective data from the prior 24 months will be collected at enrollment. Patients will then be followed prospectively while receiving usual care. Regular follow-up from a call center will confirm vital status. Biological samples, including DNA, RNA and plasma, will also be collected.



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3. O'Brien EC, et al. BMJ Open Respir Res 2016;3(1):e000108.

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ILD-PRO Registry enrolling centers: Albany Medical Center, Albany, NY; Baylor College of Medicine, Houston, TX; Baylor University Medical Center at Dallas, Dallas, TX; Cleveland Clinic, Cleveland, OH; Columbia University Medical Center/New York Presbyterian Hospital, New York, NY; Duke University Medical Center, Durham, NC; Froedtert & The Medical College of Wisconsin Community Physicians, Milwaukee, WI; Houston Methodist Lung Center, Houston, TX; Lahey Clinic, Burlington, MA; Loyola University Health System, Maywood, IL; Lynchburg Pulmonary Associates, Lynchburg, VA; Medical University of South Carolina, Charleston, SC; National Jewish Health, Denver, CO; NYU Medical Center, New York, NY; Piedmont Healthcare, Austell, GA; Ponce Research Institute, Ponce, Puerto Rico; Pulmonary Associates of Stamford, Stamford, CT; Pulmonlx LLC, Greensboro, NC; Renovatio Clinical, The Woodlands, TX; Salem Chest and Southeastern Clinical Research Center, Winston Salem, NC; South Miami Hospital, South Miami, FL; St. Joseph's Hospital, Phoenix, AZ; Stanford University, Stanford, CA; Temple University, Philadelphia, PA; The Oregon Clinic, Portland, OR; Thomas Jefferson University, Philadelphia, PA; Tulane University, New Orleans, LA; UNC Chapel Hill, Chapel Hill, NC; University of Alabama at Birmingham, Birmingham, AL; University of California, Davis, Sacramento, CA; University of California Los Angeles, Los Angeles, CA; University of Chicago, Chicago, IL; University of Cincinnati Medical Center, Cincinnati, OH; University of Louisville, Louisville, KY; University of Miami, Miami, FL; University of Michigan, Ann Arbor, MI; University of Minnesota, Minneapolis, MN; University of Pennsylvania, Philadelphia, PA; University of Pittsburgh, Pittsburgh, PA; University of Virginia, Charlottesville, VA; UT Southwestern Medical Center, Dallas, TX; Vanderbilt University Medical Center, Nashville, TN; Vermont Lung Center, Colchester, VT; Wake Forest University, Winston Salem, NC; Washington University, St. Louis, MO; Weill Cornell Medical College, New York, NY; Wilmington Health and PMG Research, Wilmington, NC; Yale School of Medicine, New Haven, CT.

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