Pulmonary Fibrosis
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NIH Awards Major Grant In Pulmonary Fibrosis Research

New Study To Pave Way For Precision Medicine

CHICAGO, Nov. 4, 2019 – The Pulmonary Fibrosis Foundation (PFF) today announced the launch of PRECISIONS, a study that aims to transform the diagnosis and treatment of idiopathic pulmonary fibrosis (IPF), by moving it into the era of precision medicine. The study is supported by a $22 million grant from the National Institutes of Health (NIH) and Three Lakes Partners, a philanthropic family organization.

More than 200,000 Americans are living with pulmonary fibrosis, a progressive and deadly lung disease. IPF, the most common form of pulmonary fibrosis, has no known cause. The symptoms of pulmonary fibrosis, which include persistent, dry cough, shortness of breath and fatigue, are similar to indications of other diseases, making it difficult to diagnose and treat.

New technologies, however, that allow scientists to isolate and analyze patients' precise genetic and molecular differences, offer new hope for IPF patients. Led by co-principal investigators Dr. Fernando Martinez and Dr. Imre Noth, the PRECISIONS (**P**rospective t**R**eatment **E**ffi**C**acy in **I**PF u**S**Ing gen**O**type for **N**ac **S**election) study will use this meticulous, high-tech approach to achieve three primary objectives –

1. Determine whether well tolerated N-Acetyl-cysteine (NAC) is an effective treatment in a subset of patients with IPF who carry a particular gene variant, TOLLIP rs3750920 T/T — about 25 percent of IPF patients have this variant;
2. Distinguish IPF from non-IPF interstitial lung diseases using unbiased combinations of blood transcriptomics, large-scale molecular analysis of blood samples, and proteomics, an extensive analysis of protein expressions in the body; and
3. Identify genetic variants that play a role in IPF risk.

Dr. Martinez, the overall principal investigator, said, "This innovative study highlights the value of a partnership between a broad range of investigators, the PFF, a philanthropic organization (Three Lakes Partners), and the National Heart, Lung, and Blood Institute (NHLBI). Most importantly, it seeks to provide patients with interstitial lung disease (ILD) access to personalized medicine in which the right medication is used for the right patient."

Dr. Noth, co-principal investigator, added, "PRECISIONS has the potential to really change the scientific landscape over how we view IPF and ILDs by providing molecular classifications while determining if a pharmacogenetically driven treatment can change outcomes."

This push toward precision medicine would not be possible without the PFF Patient Registry and Biorepository, which has enrolled more than 2,000 patients with a diverse range of PF disorders at 42 Care Center Network sites nationwide. The Registry samples will provide baseline phenotypic data for the study—disease symptoms and demographic and social characteristics and longitudinal

data—showing changes over time. More than 1,500 patients in the Registry have consented to be contacted for future research.

"The PFF Patient Registry will serve as an invaluable tool to facilitate more efficient enrollment into the NAC pragmatic trial and to further define the genetic risk factors influencing the development and potential progression of the disease," said Dr. Gregory Cosgrove, Chief Medical Officer of the PFF. "It will hopefully allow for the identification of important biomarkers to assist in the diagnosis and care of patients with PF."

PRECISIONS will set the stage for a wide range of important future research. Investigators will characterize gene expression and protein biomarkers for the PFF Patient Registry cohort and define these genetic and molecular "signatures" for distinguishing IPF from non-IPF interstitial lung diseases. This approach will help distinguish individual diseases and predict disease course and response to therapy.

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About the Pulmonary Fibrosis Foundation
The Pulmonary Fibrosis Foundation mobilizes people and resources to provide access to high quality care and leads research for a cure so people with pulmonary fibrosis will live longer, healthier lives. The PFF collaborates with physicians, organizations, patients, and caregivers worldwide. The Pulmonary Fibrosis Foundation has a three-star rating from Charity Navigator and is a Better Business Bureau accredited charity. For more information, visit pulmonaryfibrosis.org or call 844.TalkPFF (844.825.5733) or 312.587.9272 from outside of the U.S.