# Functional Decline Over Time in Patients With Idiopathic Pulmonary Fibrosis Treated With Pirfenidone: The PROOF Registry

## Background

- Idiopathic pulmonary fibrosis (IPF) is a progressive, fatal, necro-inflammatory lung disease associated with a median survival time from diagnosis of 3-5 years.
- Progressive and debilitating symptoms, including cough and exercise intolerance, cause restrictions to daily activities and may depend on family members and caregivers.

## Methods

- The PROOF registry enrolled patients from eight centers in Belgium and Luxembourg between October 2013 and January 2016.
- Eligible patients were over 18 years of age, with a multidisciplinary diagnosis of idiopathic or probable IPF.
- Patients were treated in the absence of disease that could explain the symptoms and were appropriate for any medication used.
- Patients were excluded if they were on clinical trial that might influence the results of the registry or if they were unwilling to participate.
- Data collection was ongoing during follow-up in the PROOF registry for any duration of treatment.

## Results

### Population

- Between October 2013 and January 2014, 277 patients with IPF were entered into the PROOF registry.
- In total, 230 (83.1%) patients were treated with pirfenidone at any time during follow-up in the PROOF registry, for any duration of treatment.
- The percentage of patients that experienced a relative decline in percent predicted DLco from Month 0 was assessed at Months 3, 6, 12 and 24 post-inclusion using:
  - Mean (SD) percent predicted FVC was 81.2% (19.0) and DLco was 66.0% (26.2)

### Decline in Lung Function

- The percentage of patients experiencing categorical lung function decline (assessed with Month 0) was assessed at Months 5, 9, 13 and 17 and was using the following definitions:
  - A 20% decline in percent predicted FVC
  - A 10% decline in percent predicted DLco
  - A relative decline of 10% absolute decline from 100% and 18% and a relative decline of 5% absolute decline from 50% to 18% were considered complete.

### Kaplan-Meier Curves

- Kaplan-Meier curves were used to graphically display time-to-event analyses for:
  - 15% absolute or relative decline in percent predicted DLco from Month 0
  - 10% absolute or relative decline in percent predicted FVC from Month 0

## Conclusions

- The PROOF registry provides valuable real-world data on disease progression in patients with IPF.
- Over 12 months of follow-up in the PROOF registry, patients treated with pirfenidone demonstrated better outcomes in comparison with placebo over a minimum of 24 months in patients with IPF.
- The percentage of patients experiencing lung function decline compared with placebo over a minimum of 24 months in patients with IPF.

## Limitations

- Use partial substitution of this registry is the existence of a relatively small population of patients treated across a limited geographic area.
- Missing data due to patients being lost to follow-up, although patient numbers were lower.

## Acknowledgments

- The PROOF registry is an ongoing, observational study initiated in 2013 to monitor disease progression in a real-world population of patients with IPF.
- The PROOF registry was sponsored by F. Hoffmann-La Roche, Ltd.
- The PROOF registry is a part of the European Respiratory Society International Conference, 2017.

## References