

Poster



Lung function trajectories in patients with idiopathic pulmonary fibrosis: data from the IPF-PRO Registry

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Lung function trajectories in patients with idiopathic pulmonary fibrosis: data from the IPF-PRO Registry



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INTRODUCTION

- Idiopathic pulmonary fibrosis (IPF) is a progressive fibrosing interstitial lung disease characterized by decline in lung function.¹
- Decline in forced vital capacity (FVC) or diffusing capacity of the lungs for carbon monoxide (DLco) in patients with IPF has been shown to be predictive of mortality,^{2,3} but few data are available on trajectories of decline in pulmonary function tests (PFTs).
- The Idiopathic Pulmonary Fibrosis Prospective Outcomes (IPF-PRO) Registry is an observational US registry of patients with IPF.⁴

AIM

- To evaluate trajectories of lung function in patients with IPF.

METHODS

IPF-PRO Registry

- Patients with IPF that was diagnosed or confirmed at the enrolling center in the previous 6 months were enrolled at 46 sites between June 2014 and October 2018.
- Patients were followed prospectively, with lung function data collected as part of routine clinical care until death, lung transplant, or withdrawal.

Analyses

- We assessed the trajectories of FVC % predicted and DLco % predicted over 48 months in subgroups based on time from enrollment to a terminal event (no event, ≤ 1 year, > 1 to ≤ 2 years, > 2 to ≤ 3 years, > 3 years).
 - A terminal event was defined as death, lung transplant, entry into hospice care, or withdrawal from the registry due to worsening of IPF.
- We used a joint model that accounted for the irregular frequency of measurements and for potential differences in trajectories of lung function between patients who did and did not have terminal events.
 - The following covariates were included: age, sex, race/ethnicity, body mass index, family history of ILD, diagnostic criteria,⁵ diagnosis of IPF prior to referral to enrolling center, oxygen use with activity and at rest, oxygen use with activity only, prior/current use of antifibrotic therapy, ever smoked, obstructive sleep apnea.
- Analyses were conducted in patients who had ≥ 1 FVC or DLco measurement after or ≤ 30 days before enrollment.

CONCLUSIONS

- Patients in the IPF-PRO Registry who had shorter times to a terminal event (death, lung transplant, entry into hospice care, or withdrawal due to worsening of IPF) had lower FVC % predicted and DLco % predicted values at enrollment.
- Based on joint models that adjusted for factors such as demographics, disease severity, and visit patterns, the trajectories of FVC and DLco % predicted suggest that rates of decline were fairly constant over time.

RESULTS

Analysis cohort

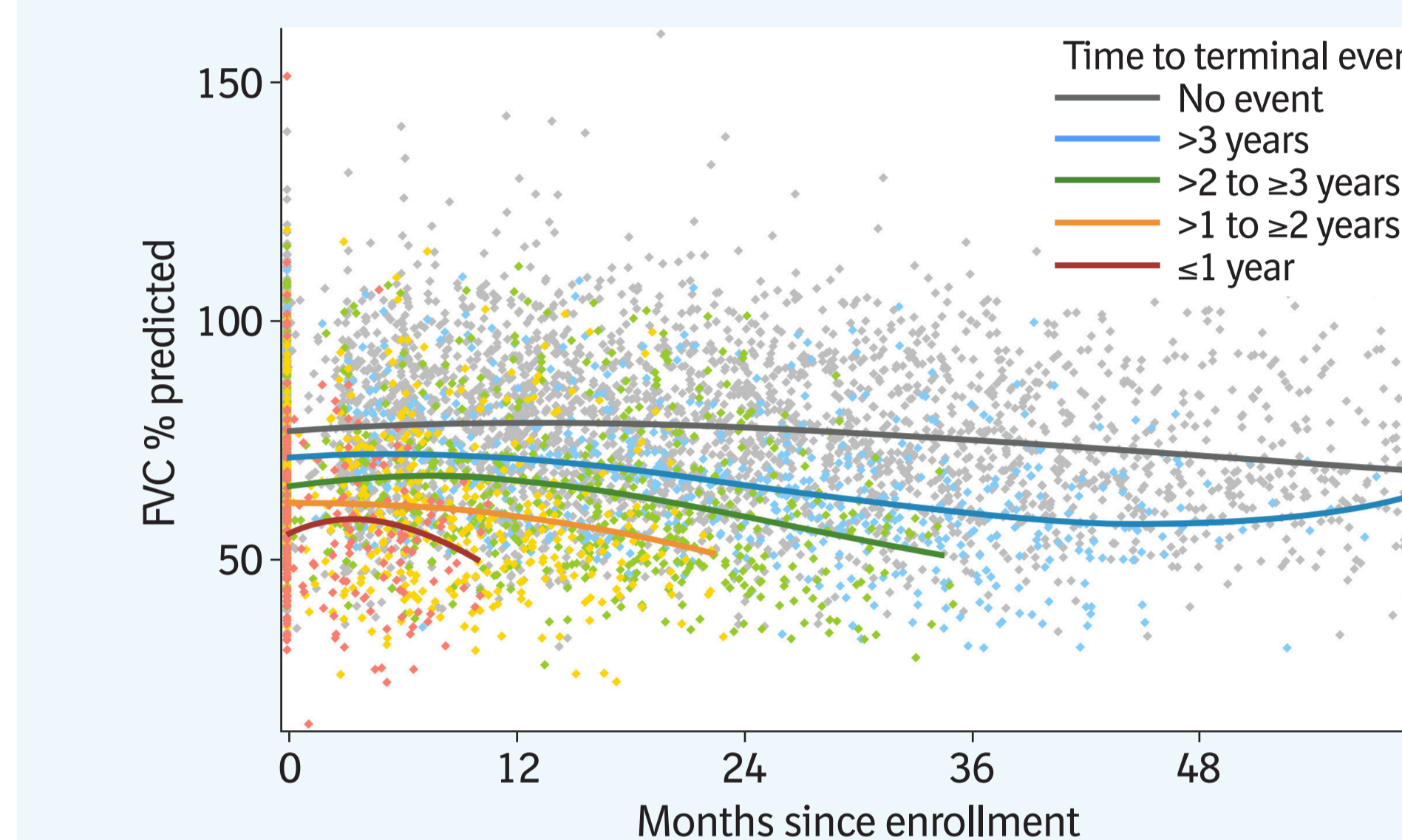
	FVC	DLco
Patients who had ≥ 1 measurement	940	901
Median (Q1, Q3) measurements per patient	4 (2, 8)	4 (2, 7)

Characteristics at enrollment (n=941)

Male	698 (74)
Age, years	70 (65, 75)
Body mass index, kg/m ²	28.9 (26.1, 32.3)
Definite IPF ⁵	613 (65)
Family history of ILD	175 (20)
FVC % predicted	72.9 (62.1, 84.2)
DLco % predicted	42.5 (33.0, 51.9)
Oxygen use at rest	178 (19)
Oxygen use with activity	315 (33)

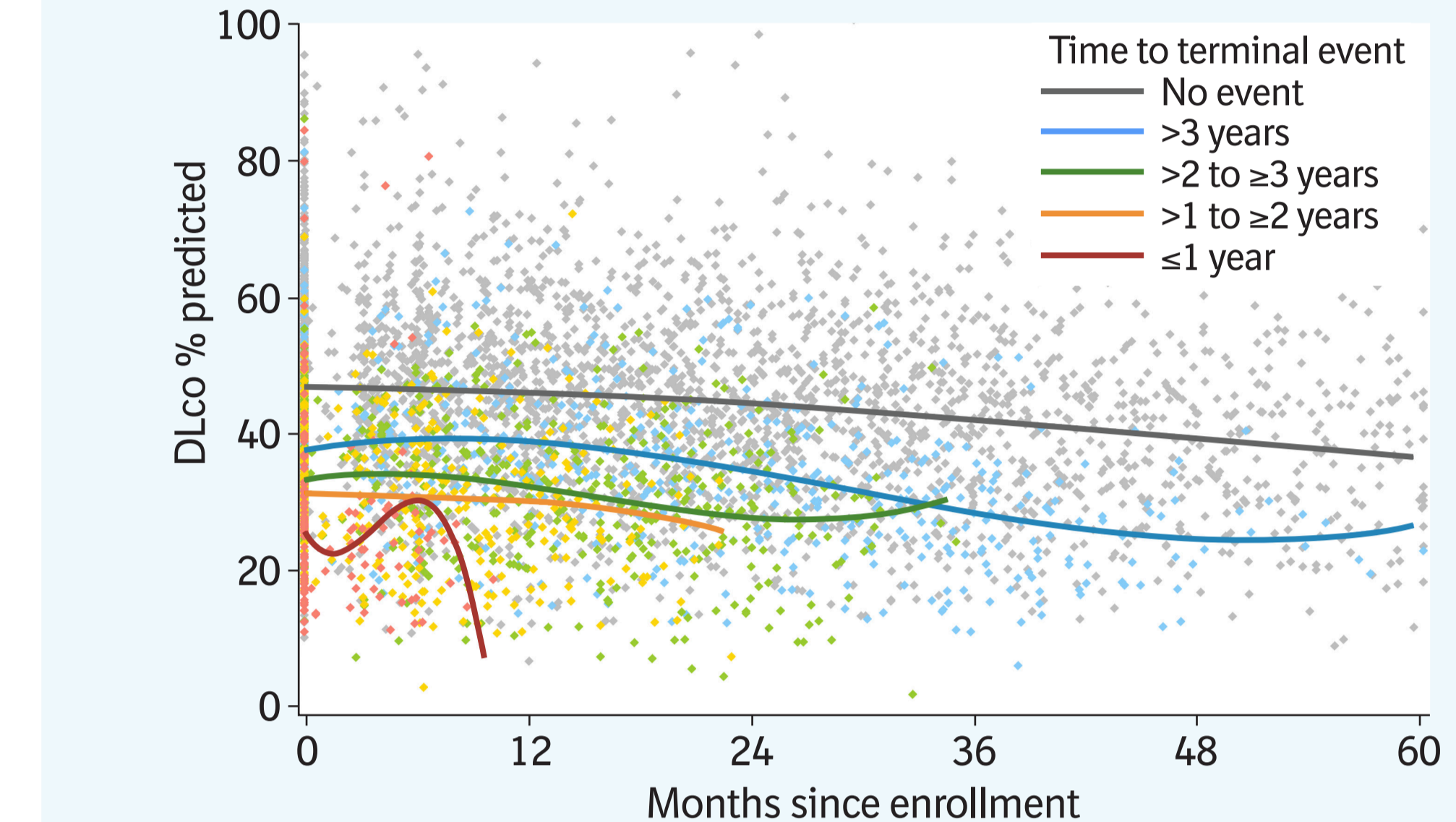
Data are median (Q1, Q3) or n (% of patients).

FVC % predicted values over time



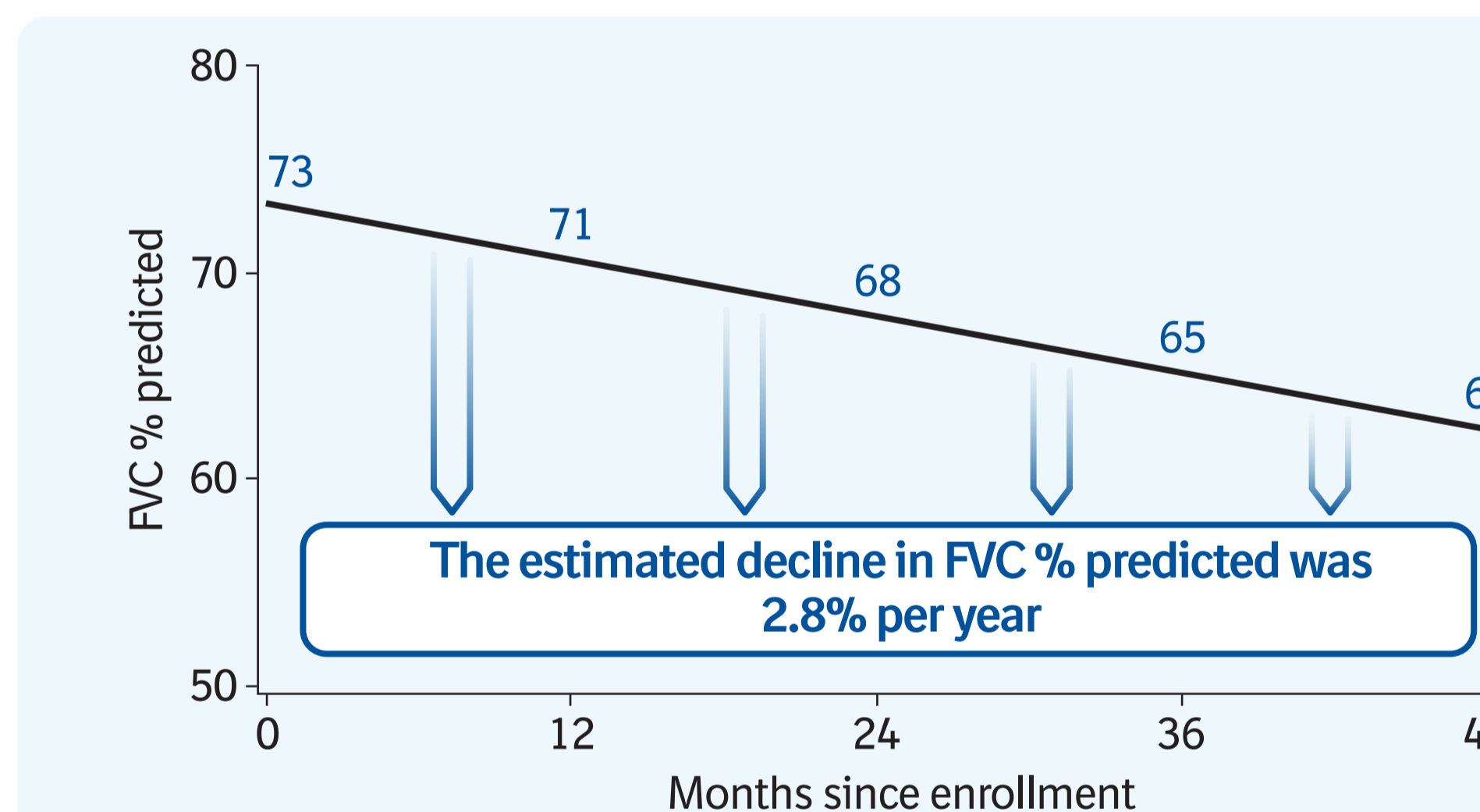
Median (Q1, Q3) follow-up in the registry was 35.1 (18.9, 47.2) months. Smoothed lines are cubic functions fit through daily means.

DLco % predicted values over time



Median (Q1, Q3) follow-up in the registry was 35.1 (18.9, 47.2) months. Smoothed lines are cubic functions fit through daily means.

Modelling of FVC % predicted over time



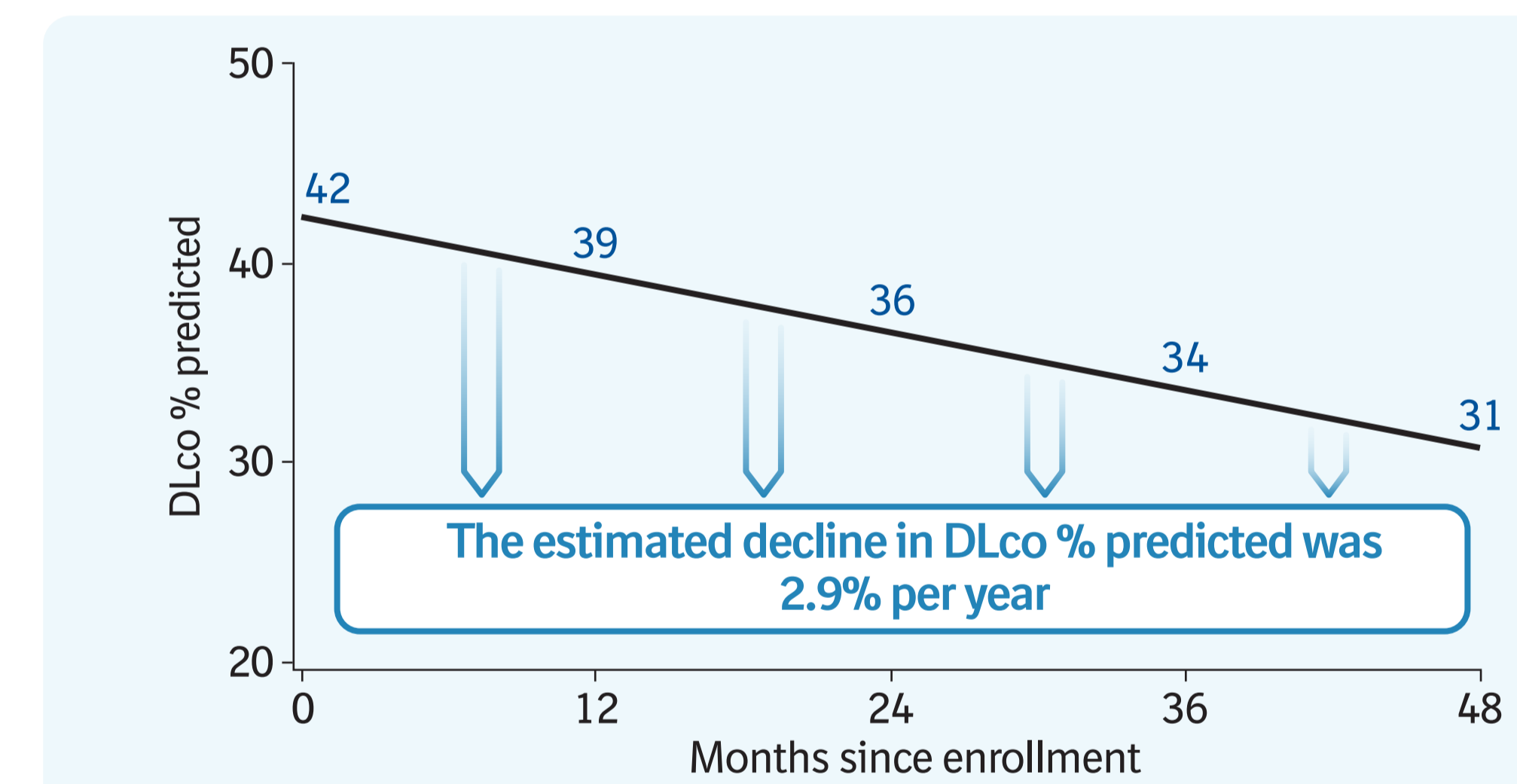
Covariates associated with a greater rate of decline in FVC % predicted

- Male
- White
- Oxygen use
- Family history of ILD
- Antifibrotic drug use

Covariates associated with a higher baseline value but not with the rate of decline in FVC % predicted

- Older age
- Higher BMI
- Current or former smoker

Modelling of DLco % predicted over time



Covariates associated with a greater rate of decline in DLco % predicted

- Male
- White

Covariates associated with a higher baseline value but not with the rate of decline in DLco % predicted

- Older age
- Higher BMI
- Never smoker
- No oxygen use
- Possible IPF

REFERENCES

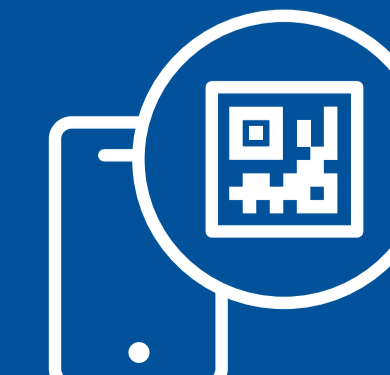
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IPF-PRO Registry enrolling centers: Albany Medical Center, Albany, NY; Baylor College of Medicine, Houston, TX; Baylor University Medical Center at Dallas, Dallas, TX; Cleveland Clinic, Cleveland, OH; Columbia University Medical Center/New York Presbyterian 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; Pulmonary Associates of Stamford, Stamford, CT; Pulmonix 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; 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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