

Implementation of guideline recommendations and outcomes in patients with idiopathic pulmonary fibrosis: data from the IPF-PRO Registry

Joao de Andrade,¹ Tejaswini Kulkarni,² Megan L Neely,^{3,4} Anne S Hellkamp,^{3,4} Amy Hajari Case,⁵ Kalpalatha Guntupalli,⁶ Shaun Bender,⁷ Craig S Conoscenti,⁷ Laurie D Snyder^{3,4} on behalf of the IPF-PRO Registry investigators

¹Vanderbilt University School of Medicine, Nashville, Tennessee, USA; ²University of Alabama at Birmingham, Birmingham, Alabama, USA; ³Duke Clinical Research Institute, Durham, North Carolina, USA; ⁴Duke University Medical Center, Durham, North Carolina, USA; ⁵Piedmont Healthcare, Atlanta, Georgia, USA; ⁶Baylor College of Medicine, Houston, Texas, USA; ⁷Boehringer Ingelheim Pharmaceuticals, Inc., Ridgefield, Connecticut, USA.

INTRODUCTION

- International guidelines for the management of IPF were published in 2011 and updated in 2015.^{1,2}
- Few data are available on the extent to which the care provided to patients with IPF is in line with the recommendations provided in international guidelines, or whether alignment with these recommendations is associated with better outcomes.
- The Idiopathic Pulmonary Fibrosis Prospective Outcomes (IPF-PRO) Registry (NCT01915511) is a prospective observational US registry of patients with IPF.³

AIM

- To investigate the extent to which guidelines for the management of IPF have been implemented at centers in the IPF-PRO Registry and whether implementation of these recommendations was associated with differences in clinical outcomes.

METHODS

- We assessed the implementation of eight recommendations within the 6 months after enrollment:



- An implementation score was calculated as the number of recommendations achieved divided by the number for which the patient was eligible. Scores ranged from 0 (no recommendations implemented) to 1 (all recommendations implemented).
- Associations between implementation score and outcomes following measurement of the implementation score were analyzed using Cox proportional hazards models or Fine and Gray models.

CONCLUSIONS

- Data from the IPF-PRO Registry suggested that recommendations made in international guidelines for the management of IPF were more likely to be implemented in patients with greater disease severity.
- When adjusted for variables known to be associated with the outcomes, no association was found between the overall implementation of management guidelines and the risk of death, lung transplant, or hospitalization.

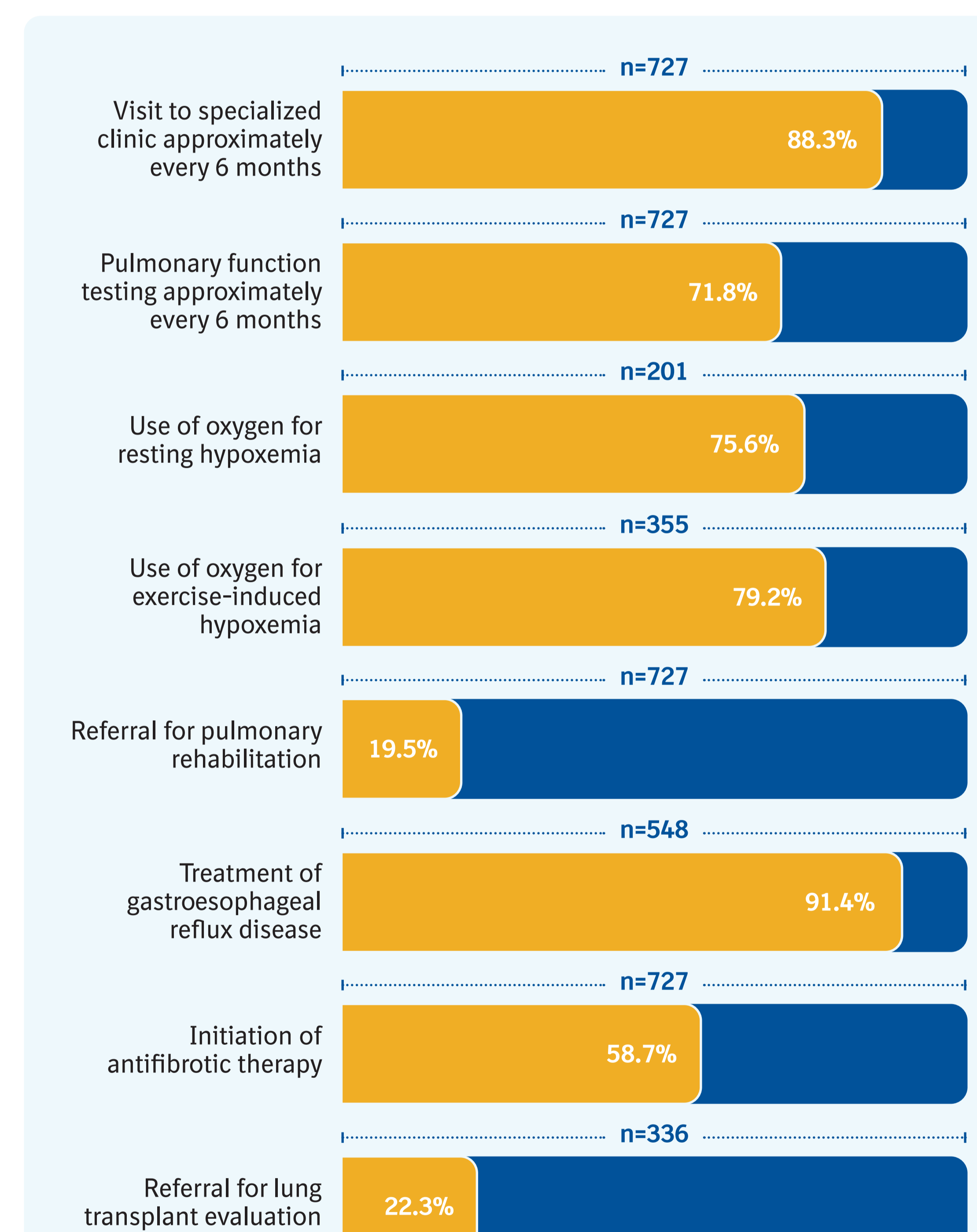
RESULTS

Implementation of recommendations

- Of 1002 patients enrolled in the registry, 79 were excluded as they were not alive and in the registry 6 months after enrollment and 196 had insufficient data for determining the implementation score. Thus, the analysis cohort comprised 727 patients.

6.3% of patients had all the recommendations that they were eligible for implemented.

Proportion of eligible patients in whom each recommendation was implemented



n, number of patients eligible for the recommendation.

Implementation score

- Median (Q1, Q3) implementation score was 0.6 (0.5, 0.8).
- Patients with an implementation score >0.6 had greater disease severity and worse quality of life than patients with an implementation score ≤0.6.

Patient characteristics at enrollment by implementation score

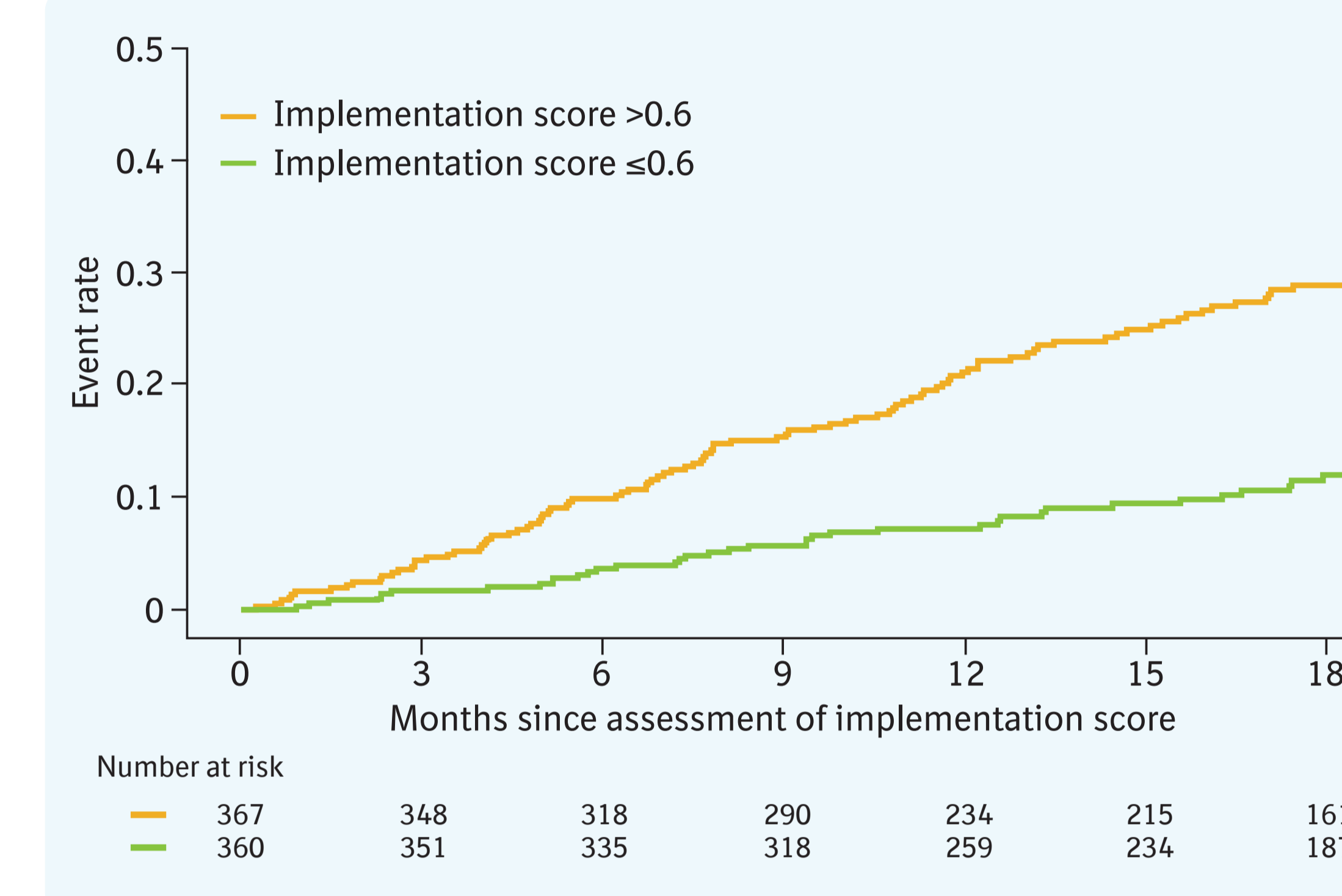
| Implementation score ≤0.6 (n=360) | | Implementation score >0.6 (n=367) |
|-----------------------------------|------------------------------------|-----------------------------------|
| 70 (64, 76) | Age, years | 71 (66, 75) |
| 71.9 | Male | 75.2 |
| 94.9 | White | 94.1 |
| 61.7 | Current/former smoker | 68.9 |
| 24.0 | Prior hospitalization* | 31.9 |
| 11.5 | Prior respiratory hospitalization* | 21.3 |
| 71.9 (62.7, 82.1) | FVC % predicted* | 68.2 (57.2, 79.9) |
| 79.8 (68.8, 90.6) | FEV ₁ % predicted* | 76.1 (64.4, 89.0) |
| 47.2 (39.1, 55.4) | DLco % predicted* | 39.6 (31.2, 48.1) |
| 38.1, 53.4, 8.5 | GAP stage I, II, III* | 25.1, 52.4, 22.5 |
| 48.8 (42.4, 55.5) | CPI* | 54.6 (47.8, 61.3) |
| 4.3 | Oxygen use at rest* | 26.0 |
| 12.8 | Oxygen use with activity* | 46.5 |
| 33.2 (19.4, 47.8) | SGRQ total score* | 40.6 (29.4, 53.3) |
| 47.7 (29.6, 66.2) | SGRQ activity score* | 59.5 (47.7, 72.8) |
| 20.5 (10.6, 36.0) | SGRQ impact score* | 27.7 (16.9, 40.3) |
| 40.6 (26.4, 56.7) | SGRQ symptoms score | 42.9 (29.4, 59.2) |
| 81.3 (59.4, 96.9) | CASA-Q cough impact domain | 81.3 (59.4, 96.9) |
| 58.3 (41.7, 83.3) | CASA-Q cough symptoms domain | 62.5 (41.7, 75.0) |
| 0.8 (0.7, 1.0) | EQ-5D index score | 0.8 (0.7, 1.0) |
| 80.0 (70.0, 90.0) | EQ-5D VAS score* | 75.0 (60.0, 85.0) |
| 54.3 (45.9, 59.7) | SF-12 MCS | 54.1 (46.0, 59.9) |
| 41.8 (34.0, 50.0) | SF-12 PCS* | 37.9 (32.1, 44.4) |

Data are % of patients without missing data or median (Q1, Q3).

*p<0.05 for comparison between groups. Continuous variables were compared using Wilcoxon rank sum tests. Categorical variables were compared using Pearson's chi-squared tests.

CASA-Q, cough and sputum assessment questionnaire; CPI, composite physiologic index; GAP, gender, age and lung physiology; MCS, mental component summary; PCS, physical component summary; SF-12, 12-item short form survey; SGRQ, St. George's Respiratory Questionnaire; VAS, visual analog scale.

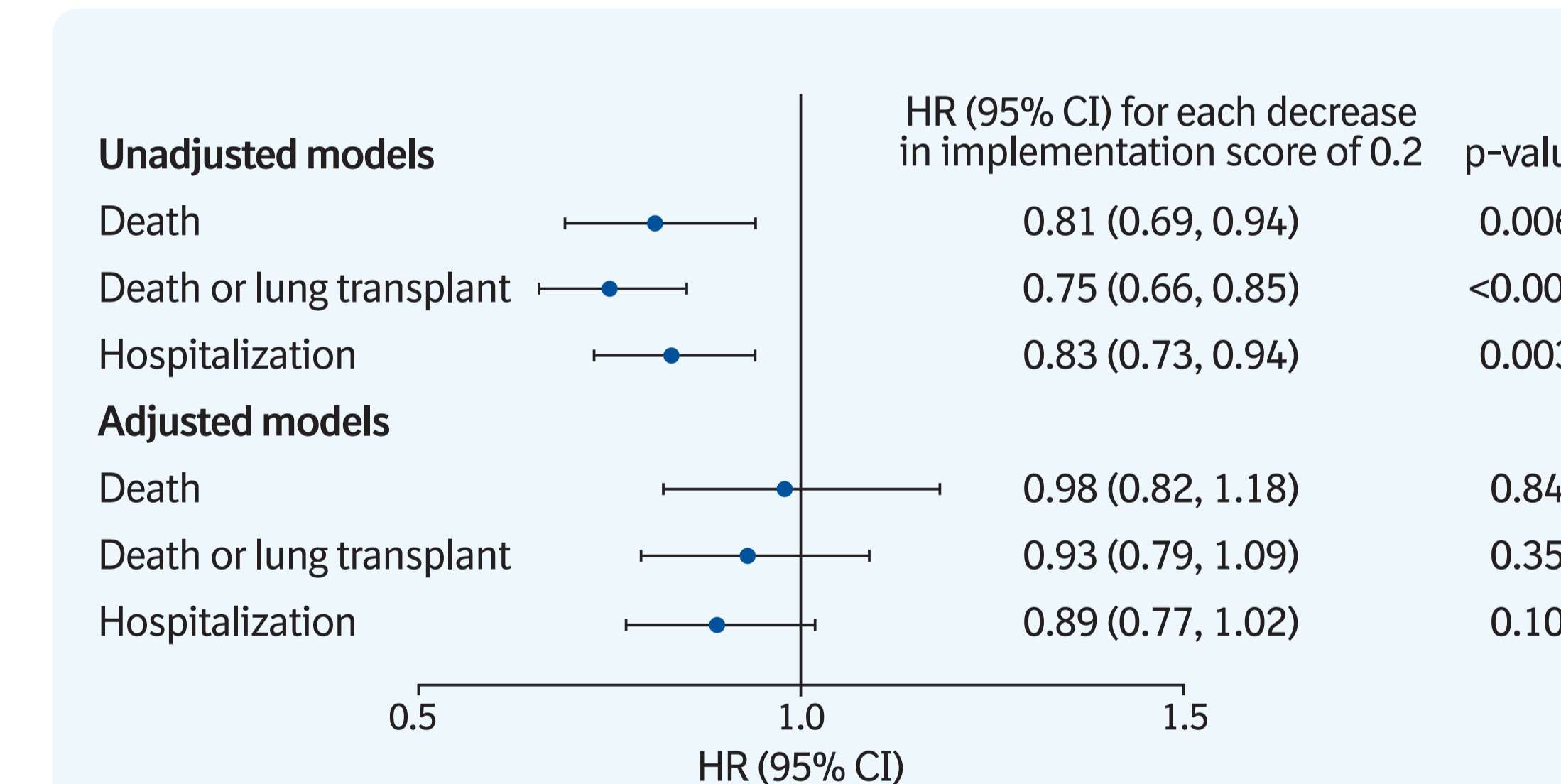
Time to death or lung transplant following assessment of implementation score



Associations between implementation score and outcomes

- In unadjusted models, patients with higher implementation scores had greater risks of death, death or lung transplant, and hospitalization. No significant associations were observed in models adjusted for other variables associated with the outcome.

Association between implementation score and death, death or lung transplant and hospitalization



Adjusted models included age, BMI, FEV₁ % predicted, FVC % predicted, DLco % predicted, oxygen use with activity, oxygen use at rest, coronary artery disease or heart failure at enrollment and diagnosis of IPF prior to referral to the enrolling center for death or lung transplant; age, BMI, FVC % predicted, DLco % predicted, oxygen use at rest and coronary artery disease or heart failure at enrollment for death; BMI, FEV₁ % predicted and oxygen use at rest at enrollment for hospitalization.

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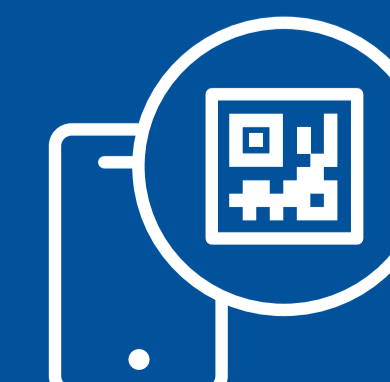
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