How should patients with progressive pulmonary fibrosis be identified? Consensus findings from a modified Delphi study

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INTRODUCTION

- The term progressive pulmonary fibrosis (PPF) is generally used to describe progressive lung fibrosis in a patient with a fibrosing interstitial lung disease (ILD) other than IPF.
- A clinical practice guideline published by ATS/ERS/JRS/ALAT in 2022[1] proposed that PPF be defined based on at least two of the following occurring in the past year:
  - Absolute decline in FVC % predicted >5% and/or DLco % predicted >10%
  - Worsening symptoms
  - Radiological progression.
- In clinical trials, PPF has been defined in a variety of ways.[2-4]

AIM

- We sought consensus among practising respiratory physicians on the identification of progression of fibrosing ILD using a modified Delphi process.

METHODS

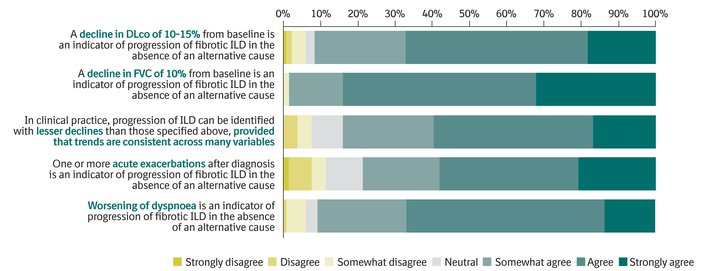
- Following a literature review, statements on the prediction, identification and monitoring of progression of ILD were developed by a panel of physicians with specialist expertise.
- Practising respiratory physicians, identified via a country-by-country internet search, were sent a survey asking them to indicate their level of agreement with the proposed statements on a binary scale or a 7-point Likert scale (-3 [strongly disagree/not at all important] to 3 [strongly agree/very important]), or to select answers from a list.
- Consensus was considered to be achieved if ≥70% of respondents agreed with a statement on a binary scale or selected the same answer from a list, or, for responses on a Likert scale, if the median score was ≤ −2 (disagree/not important) or ≥ 2 (agree/important) with an interquartile range ≤1.
- There were three rounds of the survey. Statements that were regarded as close to consensus were adapted and surveyed in the next round.
- Survey 1 was sent to 405 physicians in 32 countries. Surveys 1, 2 and 3 were completed by 207, 131 and 94 physicians, respectively.
- Surveys were completed between March 2022 and July 2023. The ATS/ERS/JRS/ALAT guideline on the definition of PPF[1] was published between surveys 1 and 2.

CONCLUSIONS

- This modified Delphi process provided consensus statements on the identification of ILD progression that were supported by a broad group of clinicians and may help to inform clinical practice until robust evidence-based guidelines are available.

RESULTS

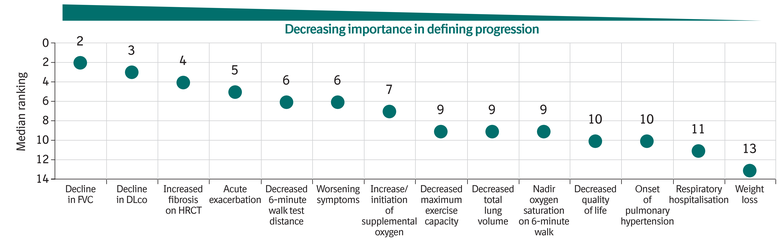
Consensus on variables that are indicators of progression of ILD



n=131 (survey 2).

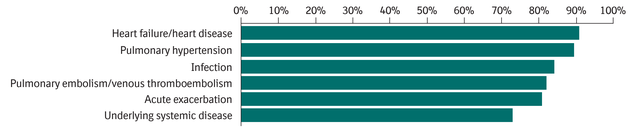
Consensus on determining progression of ILD

Most important endpoints for defining progression of ILD

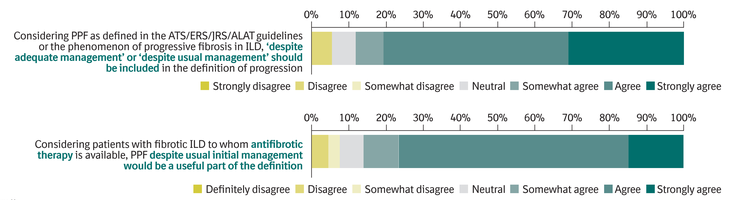


Respondents were asked to rank the endpoints from 1 (most important) to 14 (least important).

Consensus on alternative explanations for disease progression in patients with fibrosing ILD



Consensus on whether "despite usual management" should be included in definition of PPF



n=94 (survey 3).

Consensus on time periods over which progression should be defined

- Consensus was reached that progression of ILD is progression irrespective of time since diagnosis
- Consensus was *not* reached on whether there is a minimum or maximum period for assessing progression of ILD
- Consensus was *not* reached on specific periods over which changes in DLco, FVC, 6-minute walk distance, exercise capacity, quality of life, total lung volume, fibrosis on HRCT, oxygen use, weight, or symptoms can define progression

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REFERENCES
1. Raghu G et al. Am J Respir Crit Care Med 2022;205:e18–e47.
2. Flaherty KR et al. N Engl J Med 2019;381:1718–1727.
3. Maher TM et al. Lancet Respir Med 2020;8:147–157.
4. Behr J et al. Lancet Respir Med 2021;9:476–486.

ACKNOWLEDGEMENTS AND DISCLOSURES
This study was supported by Boehringer Ingelheim. The authors meet criteria for authorship as recommended by the International Committee of Medical Journal Editors (ICMJE). The authors did not receive payment for the development of this poster. Elizabeth Ng of Fleishman-Hillard, London, UK, provided editorial assistance, which was contracted and funded by Boehringer Ingelheim. Boehringer Ingelheim was given the opportunity to review the poster for medical and scientific accuracy as well as intellectual property considerations. Athol Wells was a member of the expert committee for this study and has received consulting and speaker fees from Boehringer Ingelheim, Roche, and Veracyte.

Poster developed for the American Thoracic Society International Conference, 2024.