Original research

Top 10 research priorities for people living with pulmonary fibrosis, their caregivers, healthcare professionals and researchers

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ABSTRACT

Introduction People with pulmonary fibrosis (PF) experience a high symptom burden, reduced quality of life and a shortened lifespan. Treatment options are limited and little is known about what patients, caregivers and healthcare professionals (HCPs)/researchers consider as the most important research priorities. This study aimed to identify the top 10 research priorities for PF across all stakeholders.

Methods Participants included people with PF, caregivers and HCPs/researchers involved with PF. The research priority setting exercise involved three stages: (1) identifying priorities using an open-ended questionnaire and thematic analysis, (2) development of specific research questions at a face-to-face workshop, and (3) online ranking of research questions to identify the top 10 research priorities using nominal group ranking method.

Results 196 participants completed stage 1 generating 560 questions and 14 research themes were identified. Stage 2 involved 32 participants and generated 53 indicative questions from which 39 were used for the final ranking. Stage 3 was completed by 270 participants. The top ranked priorities focussed on medications to reverse scarring in the lungs (ranked 1st), improving lung function (ranked 2nd, 6th and 8th), interventions aimed at alleviating symptoms (ranked 5th and 7th), prevention of PF (ranked 3rd and 4th) and the best exercise programme for PF (ranked 10th). There was good consensus among patients/carers and HCPs/researchers on the top 10 priorities, however, causes of acute exacerbations and early diagnosis for improving survival, was ranked higher by HCPs/researchers.

Conclusion Interventions for preserving lung health and alleviation of symptom burden were top research priorities for PF stakeholders.

Key messages

What is the key question?

► What are the most important research priorities for pulmonary fibrosis as determined by people with the disease, caregivers, healthcare professionals and researchers?

What is the bottom line?

► People with pulmonary fibrosis, caregivers and healthcare professionals/researchers collectively identified medications to reverse scarring in the lungs, medications to improve lung function and strategies to prevent pulmonary fibrosis as their top three most important priorities that should be addressed by research.

Why read on?

► This is the first research setting exercise for pulmonary fibrosis. The results will allow researchers, funders and policymakers to ensure that future research efforts are well aligned with stakeholder priorities.

INTRODUCTION

Pulmonary fibrosis (PF) is a group of life-limiting, interstitial lung diseases with no curative options and few effective treatments, despite decades of multidisciplinary research efforts. Idiopathic pulmonary fibrosis (IPF), PF of unknown aetiology, accounts for the majority of all cases, and is the form of PF that is most extensively researched.[1] Recent research has confirmed that all forms of PF are generally progressive, leading to increasingly debilitating symptoms, falling quality of life, declining lung function and ultimately shortened survival.[2] To date, research efforts have mainly focussed on effective therapeutic options to reduce IPF's symptom burden and improve survival. However, the extent to which this is a shared priority of patients, caregivers and healthcare professionals (HCPs)/researchers has not been assessed in detail, such that there may be a mismatch between current research outputs and the priorities of people living with the disease.[3]

PF is associated with deterioration in lung function, dyspnoea, chronic cough and impaired quality of life. Patients often experience a relentless increase in symptoms that become distressing for themselves and caregivers and presents an ongoing challenge in maintaining quality of life.[4] Proven treatments for PF are limited. Antifibrotic therapies (nintedanib and pirfenidone) slow the decline in lung function in people with IPF[5] and more recent findings suggest a similar role for nintedanib in people with other types of PF.[2] However, while antifibrotic therapies slow down (rather than prevent) the decline in lung function, neither therapies have shown a beneficial effect on the debilitating symptoms and impaired

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quality of life. Therefore, most people with PF continue to experience a high symptom burden, a progressive decline in lung function and a reduced physical capacity that negatively impacts their physical, psychological and social well-being.[6–8]

As a life-limiting disease, there are many aspects of the PF journey that contribute to the dismal prognosis and psychological impact that require focussed research efforts. Yet, despite the gravity of the disease and its devastating impact on patients and families, there is a paucity of information regarding what research matters most to those living with the disease as well as those involved with their treatment and management. As such, current research priorities and research investments may not necessarily align with stakeholder priorities.

The last decade has seen a concerted effort to include patients and caregivers in establishing research priorities. Initiatives such as the UK-based James Lind Alliance (JLA)[9] have developed frameworks for research priority setting projects that aim to raise the awareness of research questions that have direct relevance and potential benefits to patients, caregivers and HCPs. These initiatives bring together all stakeholders in a priority setting partnership with the objective of shaping research agendas by identifying and describing research priorities as determined by the stakeholders. Such partnerships have been developed for a range of conditions that include pulmonary hypertension, chronic kidney disease, cancer, diabetes and depression.[10] However, no priority setting exercise has been undertaken for PF.

The aim of this study was to identify the research questions that are most important to people living with PF, their caregivers, HCPs and researchers, so as to better align stakeholder priorities to research efforts.

METHODS

A research priority setting exercise was undertaken based on the JLA framework for research priority setting partnership[9] to produce a list of the 'top 10' research questions. Results were reported using the REporting guideline for PRIority SEtting of health research (REPRISE) guidelines.[11] (online supplemental file 1)

Eligibility

Eligibility criteria for participation included (1) person with PF: diagnosed by a physician as having PF; (2) caregiver: identified as being involved in the care of someone with PF; (3) HCPs: involved in the diagnosis, treatment and/or management of people with PF or involved in research focussed on PF; (4) researchers: individuals involved in clinical or academic research focussing on PF. There were no exclusion criteria for this study and eligibility for participation in each stage was independent of participation in any previous stage.

This research priority setting exercise for PF involved three stages:

Stage 1: Questionnaire - *identifying important domains of research in PF*

Participants: People with PF and their caregivers were identified through the Australian Idiopathic Pulmonary Fibrosis Registry (AIPFR) and Lung Foundation Australia's (LFA) website and electronic direct mail (EDM) database. Healthcare professionals with experience in PF were identified through the Australian IPF Network (AUS IPF-net), a special interest group for HCPs also managed by LFA.

Methods: The initial questionnaire aimed to gather a wide range of experiences from people with PF, their caregivers, HCPs and researchers. The questionnaire included one question with an open text response: '*What are the two most important problems for people with pulmonary fibrosis that you feel should be addressed by research?*' Additional demographic information such as current age, gender, years living with PF and residential state/territory in Australia was also collected. The questionnaire was piloted by patients at an interstitial lung disease clinic to ensure clarity of items and developed as an online survey using Qualtrics XM software accessible through an anonymous link. Alternatively, a paper copy was posted if requested. The questionnaire remained open for 8 weeks from early July 2019 to end of August 2019. In addition, information at the end of the questionnaire provided respondents with an opportunity to register their interest in participating in stage 2 of the study, a face-to-face workshop.

Consent: Implied consent was obtained through the completion of the questionnaire.

Analysis: Each of the 560 questions submitted as part of the stage 1 questionnaire were independently reviewed by two researchers (a senior qualitative researcher (AEH) and a mid-level researcher with experience in qualitative research (GT) using thematic analysis. This involved identifying 'key words' or codes in each response to identify patterns or themes. Duplicate or similar questions were incorporated under themes and no question was considered 'out of scope'. A total of 52 initial themes were identified that encompassed all 560 questions. The themes were then compared and discussed to generate a list of 14 overarching themes to guide discussion at the stage 2 workshop. The number of questions related to each of the 14 themes were tallied based on the proportion of total respondents to determine the order of the themes. Online supplemental file 2 lists the initial 52 themes, the number of questions that were categorised into each theme and how these related to the 14 overarching themes.

Stage 2: Face-to-face workshop - *development of specific research questions*

The objective of the face-to-face workshop was to use the themes generated in stage 1 to develop more specific research questions within each theme.

Participants: People with PF, caregivers, HCPs and researchers were invited to participate in stage 2.

Methods: A half-day, face-to-face workshop was held in Melbourne, Australia, in August 2019. The workshop was promoted at the end of the stage 1 questionnaire as well as through LFA, inviting all stakeholders to be involved. The workshop involved small group discussions, each group led by a facilitator (AEH, JYTL, GT, and AJHC) with the overall process guided by an experienced facilitator (AT). During the workshop, the themes identified in stage 1 were presented to the collective group. The areas were discussed within each of the small groups with participants tasked with developing specific research questions based on the PICO model where possible: population (P), intervention/indicator (I), control (C - if applicable) and outcome (O).[12] For example in the question '*What treatments are most effective in improving mental well-being (mood, anxiety, depression, optimism) in people with PF?*' P=people with PF, I=treatments and O=mental well-being including mood, anxiety, depression and optimism. Questions were documented on paper by a spokesperson from each group. Discussions at each table were audio recorded (with consent) to

allow for the context of questions to be re-examined after the workshop.

Consent: Informed written consent was obtained in person at the workshop.

Analysis: In the last stage of the workshop, each participant within each group used coloured adhesive dots to indicate what for them, were the three most important questions from the group generated lists. The leading facilitator (AT) then collated the list of questions ranked by each group, to determine which questions were ranked as the top priorities and reported them back to the group.

Stage 3: Prioritisation questionnaire - *ranking of the top 10 research priorities*

Participants: The questionnaire was distributed to all stakeholders using a similar approach adopted in stage 1.

Consent: Implied consent was obtained through the completion of the questionnaire.

Methods: The specific research questions generated during stage 2 were collated and reviewed by three researchers (GT, AEH and AT) to ensure that the content of the questions did not overlap, the wording was unambiguous and where possible, the question followed the PICO model. The final list of questions was developed into an online questionnaire using Qualtrics XM software, accessible through an anonymous link or made available in paper form if requested. The questionnaire was piloted with several IPF patients. Participants were asked to read through the list of questions and then identify the 10 questions that were most important to them by numbering them from 1 (most important) to 10 (least important).

A concerted effort was made to achieve a representative number of respondents across all stakeholder groups in particular, HCPs and researchers, by promoting the questionnaire through the Thoracic Society of Australia and New Zealand (TSANZ) weekly e-e-newsletter and local branch meetings and a more direct distribution of the questionnaire to colleagues in departments of respiratory medicine across local hospitals. The questionnaire remained open for 5 months from November 2019 to March 2020.

Analysis: Demographic data was analysed descriptively as n (% of total). Identification of the final 10 research priorities was based on the Nominal Group Technique used for ranking healthcare priorities.[13] This involves computing an importance score for each question defined as the average of the reciprocal rankings. The reciprocal rank is defined as 1 over the rank assigned by the participant for that question. Questions that were not ranked by participants were given a 0 as the reciprocal ranking. The importance score ranges from 0 to 1, with a higher score indicating a higher priority. Values approaching 1 indicate a highly prioritised outcome based on higher ranks and more frequent nominations, whereas values approaching 0 indicate infrequently and/or poorly ranked outcomes. CIs (95%) were calculated for each importance score using bootstrapping. Importance scores were used to identify the top 10 research priorities for all respondents, people with PF, caregivers and HCPs/researchers, respectively.

All calculations were performed using Microsoft Excel 2013 or Stata/MP (V.15.1; StataCorp, Texas, USA).

RESULTS

Stage 1

One hundred and ninety-six questionnaires were completed in stage 1. Characteristics of respondents are described in table 1

Table 1 Characteristics of respondents of the identifying priorities and prioritisation questionnaires

Characteristic	Stage 1 Identifying priorities questionnaire (n=196)	Stage 3 Prioritisation questionnaire (n=261)
Category of respondent		
Person with PF	152 (78)	134 (51)
Caregiver of person with PF	32 (16)	38 (15)
Healthcare professionals/ researchers	12 (6)	89 (34)
Gender		
Male	91 (46)	124 (48)
Female	98 (50)	110 (42)
Not reported	7 (4)	27 (10)
Current age, years (people with PF and caregivers)	**(n=184)**	**(n=172)**
≤54	9 (5)	10 (6)
55–64	19 (10)	12 (7)
65–74	82 (45)	74 (43)
75–84	61 (33)	55 (32)
≥85	10 (5)	12 (7)
Not reported	3 (2)	9 (5)
Residence (people with PF and caregivers)	**(n=184)**	**(n=172)**
ACT	3 (2)	3 (2)
NSW	60 (33)	71 (41)
NT	3 (2)	2 (1)
QLD	17 (9)	13 (8)
SA	19 (10)	14 (8)
TAS	11 (6)	7 (4)
VIC	56 (30)	45 (26)
WA	12 (7)	8 (4)
Not reported	0 (0)	9 (5)
Years since diagnosed (people with PF)	**(n=152)**	**(n=134)**
0–1	12 (8)	16 (12)
2–4	71 (47)	115 (86)
≥5	65 (43)	59 (44)
Don't know	1 (1)	3 (2)
Relation of caregiver to person with PF (caregivers)	**(n=32)**	**(n=38)**
Husband	12 (37)	11 (29)
Wife	14 (44)	15 (40)
Partner	0 (0)	1 (2)
Child	5 (16)	5 (13)
Other	1 (3)	0 (0)
Not reported	0 (0)	6 (16)
Health professionalspeciality(>1option allowed)	**(n=12)**	**(n=89)**
Allied health professional	3 (25)	15 (17)
Nurse	1 (8)	6 (7)

Continued

Table 1 Continued

Characteristic	Stage 1 Identifying priorities questionnaire (n=196)	Stage 3 Prioritisation questionnaire (n=261)
Researcher	1 (8)	10 (11)
Respiratory physician	0 (0)	38 (43)
Specialist ILD physician	7 (58)	12 (13)
Physicianandresearcher	0 (0)	2 (2)
Other	0 (0)	12 (13)*
Years of experience working in PF (HCPs)		**(n=89)**
0–5	NA	35 (39)
6–10	NA	15 (17)
11–15	NA	15 (17)
16–20	NA	9 (10)
21+	NA	15 (17)

Results are expressed in terms of numbers (% of n)
NA= not asked
*Others include: exercise physiologist (n=1), respiratory attendant/trainee (n=5), registrar (n=3), physician trainee (n=2), clinical psychologist (n=1)
ACT, Australian Capital Territory; HCPs, healthcare professionals; ILD, interstitial lung disease; NA, not asked; NSW, New South Wales; NT, Northern Territory; PF, pulmonary fibrosis; QLD, Queensland; SA, South Australia; TAS, Tasmania; VIC, Victoria; WA, Western Australia.

(column 2). In summary, 78% of all respondents were people with PF and 50% were women. Of respondents who were people with PF or caregivers, 45% were aged between 65 and 74 years with 44% of caregivers being wives of people with PF. Of the 152 with PF, 43% had been diagnosed five or more years ago. We had representation from each state and territory in Australia, with the distribution of participants consistent with that in the AIPFR and the general Australian population.[14]

Of the 560 individual questions extracted from the questionnaires, thematic analysis identified 52 initial themes based on 'key words' that encompassed all 560 responses. For example, '*Quicker/better/more certain diagnosis – early detection, better prognosis*' was based on the inclusion of key words such as '*early detection*' and '*better prognosis*'. In total, there were four questions that included such words for which 87 of the 196 respondents mentioned these words in the research questions they submitted. The themes were then compared and discussed to generate 14 overarching themes to discuss at the stage 2 workshop. (table 2) (online supplemental file 2).

Stage 2

The stage 2 face-to-face workshop involved a total of 32 participants who were randomly allocated to one of five small groups. Of the 32 participants, 63% (n=20) were people with PF, 15% (n=5) were caregivers and 22% (n=7) were HCPs and researchers. A total of 53 specific research questions were generated from small group discussions. A review of questions by three researchers (GT, AEH and AT) identified 14 overlapping questions. For example, two questions read as '*What breathing techniques improve life participation?*' and '*Breathing retraining/strategies (trumpet playing!)*'. These were incorporated into the question '*Can breathing exercises improve symptoms and life participation in people with PF?*' For the majority of questions, the wording was not rephrased by the researchers but rather they are reported verbatim as expressed by participants during the workshop. This resulted in a final list of 39 research questions for prioritisation. (online supplemental file 3)

Stage 3

A total of 270 individuals completed the prioritisation questionnaire. However, 9 (3%) were incorrectly completed leaving a total of 261 (97%) valid questionnaires that form the basis of these analyses. Table 1 (column 3) describes the characteristics of the 261 respondents. Of these, 51% were people with PF,

Table 2 Stage 1 – overarching themes identified from the responses to the stage 1 questionnaire (n=560)

Overarching themes	Number of questions incorporated into final theme	% of respondents for final themes (n=196)
(1) Quicker/better/more certain **diagnosis** – *early detection, better prognosis*	4	44%
(2) Better **treatments** – *fewer side effects, reduce disease progression, prolong life, lung transplant eligibility and post-transplant medications*	6	40%
(3) Understand the **cause** of PF – *family history, occupation, exposures, mechanism*	4	39%
(4) Improve **education / information / awareness** – *reduce stigma, understand experience of living with PF*	5	35%
(5) Relieve **symptoms** – *breathlessness, cough, fatigue*	5	29%
(6) Find a **cure** for PF – *provide hope for those with disease*	1	24%
(7) Better **supportive care** – *optimal exercise and rehab programmes, oxygen therapy, diet, end of life planning and care*	6	19%
(8) **Psychosocial support** for patients and carers– *awareness of support groups and where to seek assistance*	6	16%
(9) Improve **self-management** – *empower individual to deal with disease*	6	12%
(10) Improve **quality of life** for patients and carers – *ability to perform basic tasks, dependency on carers*	2	8%
(11) Access to **research and clinical trials** – *access to trials, information on research*	3	8%
(12) **Better access to care**, especially in rural/regional areas – *need to travel long distances for care, impact on patient and carers*	2	6%
(13) Understand my **prognosis** – *stages of progression, life expectancy, what to expect*	2	3%
(14) Decrease **costs** of care – *long-term use of oxygen expensive*	1	1%

PF, pulmonary fibrosis.

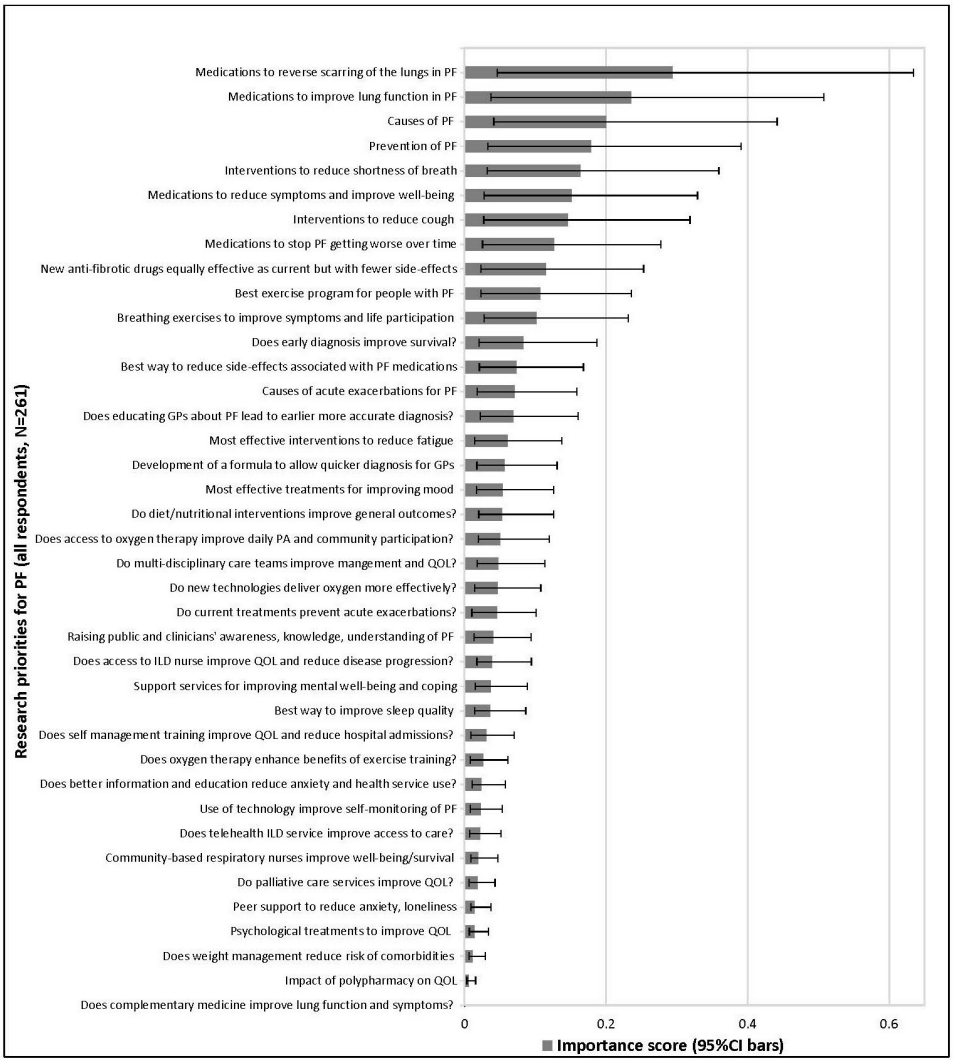

Figure 1 Overall importance scores based on all stakeholders (95% CI bars). GPs, general practitioners; ILD, interstitial lung disease; PA, physical activity; PF, pulmonary fibrosis; QOL, qualityof life.

34% HCPs/researchers and 15% were caregivers (15%) with 48% of respondents being men. Of the respondents who were people with PF or caregivers, 43% were aged between 65 and 74 years, with 40% of caregivers being wives of people with PF, and 44% of people with PF having been diagnosed five or more years ago. For HCPs/ researchers, 41% of the 89 respondents reported their speciality as being respiratory physician and within this group, 39% had between 0 to 5 years of experience working in PF.

Top 10 research priorities for PF

Figure 1 shows the ranking and importance scores for each of the 39 research questions developed for prioritisation based on all participants. Table 3 (overall rank P1-P10) lists the final top 10 research priorities for PF. Four of the 10 priorities related to the development of medications (P1, P2, P6 and P8), two related to interventions aimed at alleviating symptoms (P5 and P8), two related to understanding the cause and prevention of PF (P3 and P4) and one on the best exercise programme for people with PF (P10).

The rankings for each of the stakeholder groups respectively are provided in online supplemental file 4 (4A – people with PF, 4B – caregivers, 4C – HCPs/researchers) while table 3 provides the respective top 10 priorities. Comparing across stakeholder groups, there was generally good consensus between the groups on the top 10 priorities, however, the actual rank varied. Caregivers and HCPs/researchers identified some research priorities that were not included in the overall top 10 list. For example, caregivers ranked '*Does educating GPs about PF lead to an earlier or more accurate diagnosis of PF?*' as their ninth most important priority, while HCPs/researchers prioritised '*What are the causes of acute exacerbations?*' as their eighth top priority. (table 3).

DISCUSSION

This is the first research priority setting exercise undertaken specifically for PF that involved people with PF, caregivers, HCPs and researchers. We have identified the top 10 list of research questions for PF determined by stakeholders as a collective. These covered three broad areas: the development of new pharmacological medications aimed at preserving lung function, interventions to help manage chronic symptoms and identifying the causes of PF.

Overall, there was good consensus across stakeholders in terms of what questions were included in their top 10 research priorities. That is, people with PF nominated the same eight questions in their top 10 list as caregivers and HCPs/researchers.

Table 3 Top 10 research priorities for PF as based on all stakeholders (overall rank), people with PF, caregivers and healthcare professionals/researchers, respectively

Overall rank (n=261)	Research question (importance score)	People with PF (n=134) Rank (IS)	Caregivers (n=38) Rank (IS)	HCPs/researchers (n=89) Rank (IS)
P1	What medications can reverse scarring in the lungs of people with PF? (0.294)	1 (0.300)	1 (0.359)	1 (0.256)
P2	What medications can improve lung function in people with PF? (0.235)	2 (0.252)	2 (0.271)	5 (0.195)
P3	What are the causes of PF? (0.200)	3 (0.188)	3 (0.198)	3 (0.219)
P4	How can we prevent PF? (0.179)	6 (0.151)	5 (0.162)	2 (0.229)
P5	What are the most effective interventions to reduce shortness of breath in people with PF? (0.164)	4 (0.161)	10 (0.108)	6 (0.190)
P6	What medications can reduce symptoms and improve well-being in PF? (0.151)	8 (0.131)	>10	4 (0.202)
P7	What are the most effective interventions to reduce cough in people with PF? (0.146)	5 (0.153)	8 (0.123)	7 (0.144)
P8	What medications can stop PF getting worse over time? (0.126)	9 (0.125)	6 (0.161)	9 (0.113)
P9	Can we develop anti-fibrotic drugs for PF that are equally effective as current options but with fewer side effects? (0.115)	7 (0.137)	7 (0.128)	>10
P10	What is the best exercise programme for people with PF? (0.106)	10 (0.117)	>10	>10
>10	*Can breathing exercises improve symptoms and life participation in people with PF?*	>10	4 (0.166)	>10
>10	*Does educating GPs about PF lead to an earlier or more accurate diagnosis of PF?*	>10	9 (0.116)	>10
>10	*What are the causes of acute exacerbations of PF?*	>10	>10	8 (0.117)
>10	*Does early diagnosis improve survival for PF?*	>10	>10	10 (0.109)

>10 indicates question was not ranked in the top 10.
GPs, general practitioners; HCPs, healthcare professionals; IS, important score; P, priority; PF, pulmonary fibrosis.

For people with PF and caregivers, their top three priorities focussed on finding medications for improving lung function and understanding the aetiology of PF. Given breathlessness and cough are the most debilitating and prevalent of unmanaged PF symptoms, it is not surprising that these areas ranked as the most important.[15] However, there was some discordance between patients/caregivers and HCPs/researchers in terms of the top 3 and top 10 priorities with HCPs/researchers ranking prevention of PF as their second priority. In addition, HCPs/researchers included the importance of early diagnosis on survival and causes of acute exacerbations (AE) as top 10 priorities. This may be attributed to the different perspectives and experiences between people with PF and HCPs who deal with the management of the disease. Given an AE-IPF is associated with a mortality rate as high as 85% and mean survival periods of between 3 to 13 days, it is understandable that this challenging area would rate highly in the priority list for HCPs.[16]

To date, there has been little previous work of this kind undertaken for PF. Our results showed that top priorities for stakeholders reflect many unmet supportive care needs of people with PF and caregivers as previously reported in qualitative studies and in a recent systematic review. Identified needs include effective treatments to address debilitating symptoms such as dyspnoea and cough, and medications that can reverse or halt the disease process.[6] Of interest was the finding that research into the causes and prevention of PF were ranked as top 10 research priorities for people with PF and HCPs/researchers. Loss of independence, becoming a burden on the family and fears of passing on the disease to other family members are reported in the literature to be prominent concerns for people with PF and caregivers, providing possible insight into why cause and prevention research questions were considered such high priorities.[17] This finding also demonstrates how the impact of living with the disease and the challenges associated with its treatment and management are critical areas requiring research attention and highlights the role that discovery research will play in addressing such priorities.

Growing evidence indicates that non-pharmacological therapies such as pulmonary rehabilitation have important effects on symptoms, functional capacity and well-being in people with PF.[18] The inclusion of an optimal exercise programme for PF as a top 10 priority suggests that such interventions continue to play an important role in the management of PF and warrant further consideration.

Several prevalent themes from stage 1, focussed on rapid and accurate diagnosis, improving education/awareness of the disease, better psychosocial support for patients and carers and self-management, were not ranked in the final top 10 in stage 3. This may stem from the proportion of stakeholders who participated in stages 1 and 3. In stage 1, the ratio of HCPs/researchers to people with PF was 1:13 however, this ratio was more balanced at 1:1.5 in the prioritisation stage. Therefore, we feel that a more indicative representation of stakeholders' priorities emerged from the final prioritisation.

The list of research questions ranked as the top 10 priorities are not prescriptive, rather they will allow researchers to develop more specific hypotheses to test in these priority areas. To achieve this end will require experts, researchers and stakeholders to work collaboratively to identify and develop research proposals based on specific, answerable research questions. Consensus on proposals should consider: *relevance* (burden of disease, equity and evidence gaps), *appropriateness* (scientific rigour and suitability of methodology to answer research question), *significance of research outcomes* (impact, innovation and capacity building), *feasibility* (team quality and research environment) and *value for money* (potentially cost effective).[19] In order to inform future work, an evaluation of the process should also be undertaken, to understand the impact of the research priority setting process. This would require outcome measures of performance that can be monitored and evaluated over time,

evaluating process indicators such as stakeholder satisfaction, transparency of process and successful submission of research proposals.

The strength of this study is that all stakeholder groups across Australia were involved in identifying the top 10 research priorities for PF and the process was conducted using a systematic method. Limitations include stakeholders being from Australia, English speaking and not having data on ethnicity. In addition, our recruitment of HCPs did not specifically target any specific group such as palliative care providers who may have made an important contribution to our findings. Limited resources prevented a comprehensive, systematic review of the literature prior to commencing the priority setting process, however the limited number of current treatments for PF makes it less likely that this would have limited the scope of the priority setting exercise. We modified the JLA process to meet the needs of our patient group as reported in previous studies by holding a smaller face-to-face meeting in stage 2, followed by an online survey for stage 3.[20] This decision was made due to the challenges of travelling to a face-to-face meeting for many people with PF, particularly in Australia where travel distances are considerable, and our desire to maximise the engagement of the PF community in stage 3 using online methods. While this enabled a broader participation in the choice of the final priorities, it could have affected the outcome.

CONCLUSION

This research priority setting exercise for PF has involved all stakeholders in identifying the top 10 research priorities for PF. Research aimed at PF prevention, development of interventions to improve lung function and alleviating symptom burden were identified as being most important to stakeholders. These findings will help guide future research directions.

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Supplementary File 1: REPRISE REPORTING GUIDELINES

No	Item	Descriptor and/or *examples*
A	**Context and scope**	
1	Define geographical scope	National – Australian states and territories
2	Define health area, field, focus	Pulmonary fibrosis, identifying research priorities
3	Define end-users of research	People diagnosed and living with an interstitial lung disease particularly, pulmonary fibrosis, carers of people with pulmonary fibrosis, healthcare professionals and researchers involved in pulmonary fibrosis.
4	Define the target audience of the priorities	Policy makers, funders, researchers, industry
5	Identify the broad research area	Public health, health services research, clinical research, basic science
6	Identify the type of research question	Etiology, diagnosis, prevention, treatment (interventions), prognosis, health services, psychosocial, behavioral and social science,
7	Define the time frame	Long-term priorities
B	**Governance and team**	
8	Describe selection of the leadership and management team	Lung Foundation Australia, NHMRC- CRE
9	Describe the characteristics of the team, and the networks they represent	People with pulmonary fibrosis, Lung Foundation Australia, Thoracic Society of Australia and New Zealand, Australian IPF Registry
10	Describe any training or experience in priority setting	Collaborated and sought advice with Consultants experienced in priority setting
C	**Framework for priority setting**	
11	State the framework used (if any)	James Lind Alliance
D	**Stakeholders or participants**	
12	Define the inclusion criteria for stakeholders involved in priority-setting	Patients, caregivers, health professionals, researchers, non-governmental organisations.
13	State the strategy or method for identifying and engaging stakeholders	Partnership and recruitment through with organizations
14	Indicate the number of participants and/or organisations involved	Number of individuals and organisations, include number by stakeholder group
15	Describe the characteristics of stakeholders	Stakeholder group, demographic characteristics, areas of interest and expertise, discipline, affiliations
16	State if reimbursement for participation was provided	No reimbursement or financial incentive for participation
E	**Identification and collection of research priorities**	
17	Describe methods for collecting priorities from stakeholders	Questionnaires (online and paper format), workshop, voting of priorities nominal group technique
18	Describe methods for collating and categorizing priorities	Thematic analysis
19	Describe methods and reasons for removing priorities	Based on scope, clarity, duplication
20	Describe methods for refining or translating priorities into research topics or questions	Reviewed by project team leaders
21	Describe methods for checking whether research questions or topics have been answered	To be undertaken
22	Describe number of research questions or topics	Final prioritization based on total of 39 questions.

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F	**Prioritisation of research topics/questions**	
23	Describe methods and criteria for prioritising research topics or questions	Nominal group technique, ranking; online or paper copy
24	Provide reasons for excluding research topics/questions	None
G	**Output**	
25	Specificity of research priorities are clear	Area, topic, questions, PICO (population, intervention, comparator, outcome)
H	**Evaluation and feedback**	
26	Describe how the process of prioritization was evaluated	To be completed
27	Describe the approach for feeding back priorities to stakeholders and/or to the public; and how feedback was addressed and integrated	To be completed
I	**Implementation**	
28	Outline the strategy or action plans for implementing priorities	To be completed
29	Describe evaluation of impact	To be completed
J	**Funding and conflict of interest**	
30	State sources of funding	NHMRC Centre of Research Excellence in Pulmonary Fibrosis
31	Outline the budget and/or cost	n/a
32	Provide declaration of conflict of interest	No conflict of interest

Supplementary file 2: DEVELOPMENT OF 14 ORVERARCHING RESEARCH THEMES

Initial themes identified as part of Stage 1 (N=196) [listed alphabetically]	**Number of respondents**	**% of total**	**Corresponding overarching theme number**
Access to services (e.g. pulmonary rehabilitation, lung transplant)	5	3%	11
Adjustment to life with PF	10	5%	6
Better access to diagnosis and care in regional areas and interstate	6	3%	11
Better informed GPs	15	8%	3
Better treatments to prolong life/reduce disease progression	51	26%	12
Blood markers	3	2%	1
Cause	65	33%	2
Complementary and alternative therapies	3	2%	7
Cost of treatment	2	1%	14
Cure	47	24%	5
Drug treatments with fewer side effects	18	9%	12
Early detection	19	10%	1
Education of HCP	9	5%	3
Effects of repeated testing	1	1%	8
End of life care	6	3%	7
HCP Informing patients of what to expect with PF	2	1%	13
Holistic care	2	1%	8
Impact of specialist visits on patient and carers	2	1%	6
improve prognosis after transplant	3	2%	12
Improve QOL for carers	1	1%	9
Improve QOL for patients	14	7%	9
Information /education for patients	18	9%	3
Information on clinical trials/research developments	6	3%	10
Isolation due to PF	3	2%	6
Lung transplant eligibility	2	1%	12

14 Overarching research themes

1. Quicker/better/more certain **diagnosis** – *early detection, better prognosis*

2. Understand the **cause** of PF – *family history, occupation, exposures, mechanisms*

3. Improve **education / information / awareness** – *reduce stigma, understand experience of living with PF*

4. Relieve **symptoms** – *breathlessness, cough, fatigue*

5. Find a **cure** for PF – *provide hope for those with disease*

6. **Psychosocial support** for patients & carers– *awareness of support groups and where to seek assistance*

7. Better **supportive care** – *optimal exercise and rehab programs, oxygen therapy, diet, end of life planning and care*

8. Improve **self-management** – *empower individual to deal with disease*

9. Improve **quality of life** for patients & carers – *ability to perform basic tasks, dependency on carers*

10. Access to **research and clinical trials** – *access to trials, information on research*

11. Better **access to care**, especially in rural/regional areas – *need to travel long distances for care, impact on patient and carers*

12. Better treatments – *fewer side effects, reduce disease progression, prolong life, lung transplant eligibility and post-transplant medications*

13. Understand my **prognosis** – *stages of progression, life expectancy, what to expect*

14. Decrease **costs** of care – *long-term use of oxygen expensive*

Manage side effects of meds	10	5%	8
Managing acute exacerbations	1	1%	8
More research	5	3%	5
More support groups for people with PF	1	1%	6
Non-lung effects of PF	5	3%	13
Optimal exercise/ pulmonary rehab program	11	6%	7
Optimising lung transplant	4	2%	12
Oxygen therapy	7	4%	7
Post lung transplant meds with fewer side effects	1	1%	12
Prevention	11	6%	5
Psychosocial support and counselling for patients	8	4%	6
Public awareness of condition	11	6%	3
Quicker/better/ more definite diagnosis	37	19%	1
Recovery from acute exacerbations	1	1%	12
Relieve breathlessness	24	12%	4
Relieve cough	21	11%	4
Relieve discomfort	2	1%	4
Relieve fatigue	8	4%	4
Relieve phlegm	1	1%	4
Role of diet	3	2%	7
Self-management /optimise living with PF	10	5%	8
Support/education for caregivers and families	7	4%	6
Treatments to improve ex capacity	8	4%	7
Understand disease mechanisms	2	1%	2
Understand prognosis	28	14%	1
Understand what research is happening	4	2%	10
Understanding experience of living with PF	16	8%	3
TOTAL	**560**		

14 Overarching research themes

1. Quicker/better/more certain **diagnosis** – *early detection, better prognosis*
2. Understand the **cause** of PF – *family history, occupation, exposures, mechanisms*
3. Improve **education / information / awareness** – *reduce stigma, understand experience of living with PF*
4. Relieve **symptoms** – *breathlessness, cough, fatigue*
5. Find a **cure** for PF – *provide hope for those with disease*
6. **Psychosocial support** for patients & carers– *awareness of support groups and where to seek assistance*
7. Better **supportive care** – *optimal exercise and rehab programs, oxygen therapy, diet, end of life planning and care*
8. Improve **self-management** – *empower individual to deal with disease*
9. Improve **quality of life** for patients & carers – *ability to perform basic tasks, dependency on carers*
10. Access to **research and clinical trials** – *access to trials, information on research*
11. Better **access to care**, especially in rural/regional areas – *need to travel long distances for care, impact on patient and carers*
12. **Better treatments** – *fewer side effects, reduce disease progression, prolong life, lung transplant eligibility and post-transplant medications*
13. Understand my **prognosis** – *stages of progression, life expectancy, what to expect*
14. Decrease **costs** of care – *long-term use of oxygen expensive*

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SUPPLEMENTARY FILE 3: STAGE 3 – Final list of research questions for prioritisation

1. What diet or nutritional intervention improves general outcomes (e.g. prevention, progression, symptoms etc.) for people with PF?

2. Can breathing exercises improve symptoms and life participation in people with PF?

3. Can new technologies deliver oxygen therapy more effectively to people with PF?

4. Does improving access to oxygen therapy improve daily physical activity and community participation in people with PF?

5. What treatments are most effective for improving mood (anxiety, depression, optimism) in people with PF?

6. What support services can improve mental wellbeing and coping in people with PF and their caregivers?

7. What medications can improve lung function in people with PF?

8. What medications can reverse scarring in the lungs in people with PF?

9. What medications can reduce symptoms and improve wellbeing in PF?

10. What are the causes of PF?

11. How can we prevent PF?

12. Can weight management reduce risk of heart disease and diabetes in people with PF?

13. Does educating GPs about PF lead to an earlier or more accurate diagnosis of PF?

14. Does PF self-management training (improving skills to manage PF) improve quality of life and reduce hospital admissions for people with PF?

15. What is the best exercise program for people with PF?

16. Do community-based respiratory nurses improve the wellbeing and survival of people with PF?

17. What is the best way to reduce the side effects associated with medications used for PF?

18. Which complementary therapies (e.g. herbal medicines, acupuncture, salt inhalation) can improve lung function and symptoms in PF?

19. What are the causes of acute exacerbations of PF?

20. Can any of our current treatments (medications, exercise) prevent acute exacerbations of PF?

21.How can technology be used to improve people's self-monitoring of PF?

22.Does having access to an ILD nurse or case manager improve quality of life and reduce disease progression in people with PF?

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23. Can a telehealth PF service (remote consultation with a physician, nurse, rehab, multi-disciplinary team) improve access to care and decrease health care utilisation for people with PF?

24. What is the impact of polypharmacy on quality of life for people with PF? What is the best way to make tablet regimens simpler for people with PF?

25. Can we develop anti-fibrotic drugs for PF that are equally as effective as current options but with fewer side effects?

26. What is the best way to improve sleep quality in PF?

27. Can oxygen therapy enhance the benefits of exercise training in people with PF?

28. Does early diagnosis improve survival in PF?

29. What medications can stop PF getting worse over time?

30. What psychological treatments can improve quality of life and social participation for people with PF?

31. Do multi-disciplined care teams improve management and quality of life for people with pulmonary fibrosis? (e.g. access to health services, dietitian, counselling, non-medical support).

32. Can we develop a formula that would allow GPs to diagnose PF more quickly?

33. What are the most effective interventions for reducing cough in people with PF?

34. What are the most effective interventions for reducing shortness of breath in people with PF?

35. What are the most effective interventions for reducing fatigue in people with PF?

36. Do palliative care services improve quality of life for people with PF and their carers?

37. Does providing better information and educational resources for people with PF improve disease understanding, reduce anxiety and reduce health service use?

38. How do we raise the public and clinicians' awareness, knowledge and understanding of pulmonary fibrosis?

39. Can peer support reduce anxiety, depression and loneliness in people with PF and their carers?

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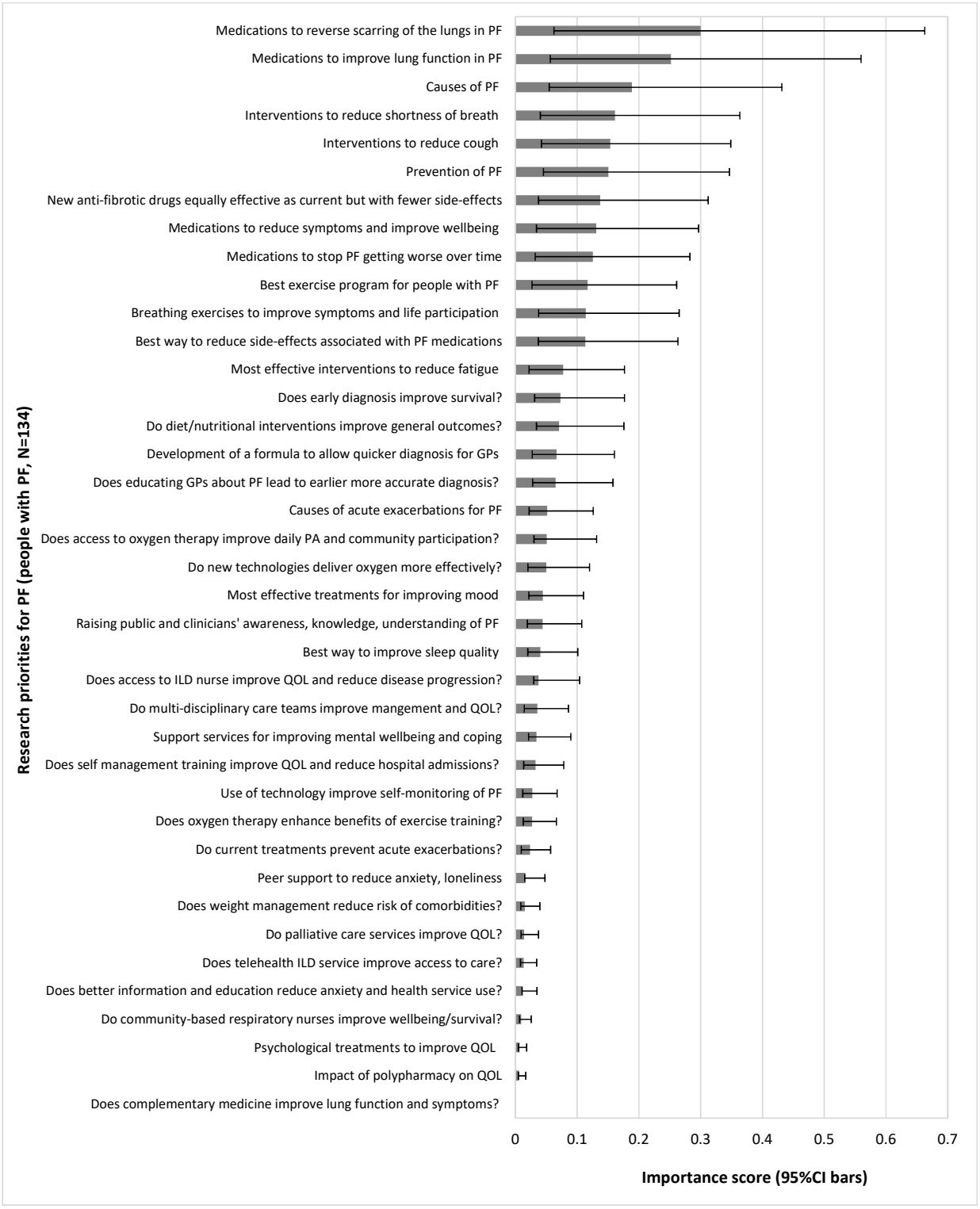
SUPPLEMENTARY FILE 4A: IMPORTANCE SCORES FOR PEOPLE WITH PULMONARY FIBROSIS



SUPPLEMENTARY FILE 4B: IMPORTANCE SCORES FOR CAREGIVERS

SUPPLEMENTARY FILE 4C: IMPORTANCE SCORES FOR HEALTHCARE PROFESSIONALS /RESEARCHERS