



Patient and caregiver shared experiences of pulmonary fibrosis (PF): A systematic literature review

Matthew Wells^{a,*}, Sam Harding^b, Giles Dixon^{a,c}, Kirsten Buckley^d, Anne-Marie Russell^{e,f}, Shaney L. Barratt^{a,g}

^a Bristol Interstitial Lung Disease Service, North Bristol NHS Trust, Bristol, UK

^b Research and Development, North Bristol NHS Trust, Bristol, UK

^c Department of Clinical and Biomedical Sciences, University of Exeter, Exeter, UK

^d Library and Knowledge Services, North Bristol NHS Trust, Bristol, UK

^e Birmingham Regional Interstitial Lung Disease Service, University Hospitals Birmingham NHS Trust, Birmingham, UK

^f Faculty of Life Sciences, University of Exeter, Exeter, UK

^g Academic Respiratory Unit, Department of Clinical Sciences, University of Bristol, Bristol, UK

ARTICLE INFO

Keywords:

Pulmonary fibrosis
Supportive care needs
Idiopathic pulmonary fibrosis
Palliative care

ABSTRACT

Pulmonary Fibrosis (PF) describes a group of lung diseases characterised by progressive scarring (fibrosis). Symptoms worsen over time and include breathlessness, tiredness, and cough, giving rise to psychological distress. Significant morbidity accompanies PF, so ensuring patients' care needs are well defined and provided for, represents an important treatment strategy.

The purpose of this systematic review was to synthesise what is currently known about the psychosocial morbidity, illness experience and needs of people with pulmonary fibrosis and their informal caregivers.

Eight databases (MEDLINE, EMBASE, PUBMED, Cochrane database of Systematic reviews (CDSR), Web of Science Social Sciences Citation Index, PsycINFO, PsycARTICLES and CINAHL) were used to identify studies exploring the supportive needs of adults with PF and/or their caregivers. Methodological quality was assessed using the Mixed Methods Appraisal Tool.

53 studies were included, the majority using qualitative methodology (79 %, 42/53), 6 as part of mixed methodological studies. Supportive care needs were mapped to eight domains using an *a priori* framework analysis.

Findings highlight a lack of psychological support throughout the course of the illness, misconceptions about and barriers to, the provision of palliative care despite its potential positive impacts. Patients and caregivers express a desire for greater disease specific education and information provision throughout the illness.

Trials of complex interventions are needed to address the unique set of challenges for patients and carers living with PF.

1. Introduction

Pulmonary Fibrosis (PF) describes a group of lung diseases where scar tissue progressively and irreversibly replaces normal lung. It is commonly a disease of senescence (mean age at diagnosis 70 years), often afflicting patients with multiple co-morbidities [1]. Median survival is 3–5 years from diagnosis [1,2]. Advanced disease is characterised by debilitating breathlessness and/or cough, alongside poor quality-of-life (QoL) and social isolation [3–7]. There is no curative

treatment, so the optimal approach to management is relieving symptoms and improving or maintaining QoL. Notably many patients living with PF report psychological distress including worry, fear, anxiety, hopelessness and helplessness [8–11].

When healthcare is inconsistent with patients and caregivers' needs unnecessary suffering, reduced patient satisfaction, increased healthcare expenditure and disengagement in health preventative behaviours may result in negative health outcomes for both patient and caregiver [12–14]. Given the significant morbidity that accompanies pulmonary fibrosis, an important treatment strategy is to ensure that patients care

* Corresponding author.

E-mail addresses: matthew.wells@nbt.nhs.uk (M. Wells), sharding.jb@gmail.com (S. Harding), giles.dixon@nbt.nhs.uk (G. Dixon), a.russell.1@bham.ac.uk (A.-M. Russell), shaney.barratt@nbt.nhs.uk (S.L. Barratt).

<https://doi.org/10.1016/j.rmed.2024.107659>

Received 26 February 2024; Received in revised form 30 April 2024; Accepted 3 May 2024

Available online 8 May 2024

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Abbreviations

ACP	advanced care planning
BTS	British thoracic society
DAS	dyadic adjustment scale
EOL	end of life
FG	focus groups
GAD-SI	generalized anxiety disorder single item questionnaire
HCP	healthcare professionals
IPF	idiopathic pulmonary fibrosis
MMAT	mixed methods appraisal tool
PC	palliative care
PF	pulmonary fibrosis
PRISMA	preferred reporting items for systematic reviews and meta-analyses
PwPF	people with pulmonary fibrosis
QoL	quality of life
SSc	systemic sclerosis
SSc-PF	systemic sclerosis associated pulmonary fibrosis

needs are well defined and provided for. Current evidence suggests there are significant unmet needs for patients with pulmonary disease [15, 16], with negative impact on the emotional health of the caregiver [17]. Further currently available disease modifying medications carry a significant side effect profile and are intolerable for many. Here, needs represent an important treatment target to improve the significant morbidity that accompanies pulmonary fibrosis.

The purpose of this systematic review is to synthesise what is currently known about the psychosocial morbidity, illness experience and needs of people with PF (PwPF) and their informal caregivers. Studies involving both PwPF and the caregiver as active research participants may provide a more holistic picture of the shared couple experience, and offer novel insight and interpretation to enhance our understanding.

2. Methods

This review was conducted in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement [18] and is registered in the International Prospective Register of Systematic Reviews (PROSPERO CRD42022368074).

Studies exploring the supportive needs of adult PwPF (>18 years) and/or their caregivers were included. The full search strategy is presented in Supplementary Data 1. There was no comparator group. Studies describing the supportive care needs of other lung diseases were excluded unless findings relating to PF were reported separately. Electronic searches of studies were performed in MEDLINE, EMBASE, PUBMED, Cochrane database of Systematic reviews (CDSR), Web of Science Social Sciences Citation Index, PsycINFO, PsycARTICLES and CINAHL from inception to October 31, 2023, without language restriction. Studies evaluating the psychometric properties of tools designed to measure supportive care needs or studies assessing the impact of an intervention on supportive care needs were excluded. Non-original research publications (e.g., editorials, reviews) were also excluded.

Covidence software (Veritas Health Innovation, Melbourne, Australia) was used as the formal systematic review management platform to combine multiple database results and facilitate deduplication. All identified titles and abstracts were screened independently by two review authors (SB and GD). Any discrepancies/disagreements were resolved by discussion between reviewers, involving a third party if necessary (SH). A grey search of references from included articles was performed. Abstracts published prior to 2021 were excluded following a manual google search to identify any subsequent full publication.

Data was extracted by one author (MW) and verified by another (SH, SB or AMR) using a standardised data extraction tool created on the Covidence platform. Data pertaining to study country of origin, date of publication, journal or publication source, study design, initial population of study, patient characteristics (age, gender, type of PF, severity and treatment) and caregiver characteristics (age, gender), inclusion and exclusion criteria of study participants were collected.

Each included article was critically appraised by two authors (MW and either SH or SB) to assess the methodological quality using the Mixed Methods Appraisal Tool (MMAT) (Supplementary data 2) [19,20]. Each criterion was graded as met (Y, green), unmet (N, red) or can't tell (CT, amber). Any disagreements were resolved by discussion.

A meta-analysis of quantitative studies was planned if appropriate and feasible. In the event of wide heterogeneity in study design, a narrative, qualitative data synthesis was planned. Supportive care needs were mapped to eight previously described domains using an *a priori* framework analysis methodology; physical, emotional, psychological, social/societal, informational/education, spiritual, practical and family related [21]. Framework analysis allows for both inductive and deductive coding, as such, an 'other' domain was added to ensure that any additional domains or codes not falling within the prespecified domains were captured. A free text section was included for additional notes on inchoate themes and verbatim quotes, to facilitate a rich and deep analysis of each included study. There was an *a priori plan* to perform subgroup analyses of supportive care needs according to disease severity and PF subtype.

3. Results

Literature searches identified 3358 studies and a further 9 studies from 'grey search' of articles. Following the removal of duplicates (n = 613), 2754 articles were screened. After full text review (194 articles), 53 studies (51 full text articles, 2 abstracts [22,23]) were included for data extraction (Fig. 1). Abstracts were included if published within 2 years of the search, as it was felt that this was the maximum period of time to allow for the abstract to be converted to a full peer review journal article.

3.1. Study characteristics

Characteristics of included studies are summarised in [Supplementary Table 1](#). Most were performed exclusively in the USA (11/53), UK (7/53) or in single European countries (13/53). 9/53 were multinational studies, of which only 1 was multicontinental [24]. 22 full text articles (42%) had been published since 2020, suggesting expanding research interest in this field.

The majority of studies used qualitative methodology (42/53, 79%), 6 as part of combined qualitative and quantitative methods [17,25–29]. Interview strategies were used in 32 studies and focus groups (FG) in 11 studies, with 4 using a combination of both [22,23,30,31]. Other qualitative approaches included a World Cafe discussion [32], observation of conversations and mock consultations [24], analysis of a narrative diary [33], open ended survey questions [28], expert panel discussion or Delphi consensus [29,34] and analysis of online forums/blogs [35]. Some studies used mixed qualitative approaches (n = 8) [22–24,30–34].

Questionnaires, surveys and patient reported outcome measures were ubiquitously employed in studies with a quantitative component. These reported on QoL/activities of daily living [17,25,29,36,37], access to healthcare/resources [25,28,29,37,38], education [26,28,37,39], levels of anxiety/worry [25,26,40], depressive or cognitive symptoms [29,41,42] and impact upon intimacy and relationships [43]. Several addressed healthcare professionals (HCP) perspectives and practice with respect to end of life (EOL) and palliative care (PC) [44–46]. Meta-analysis was not possible due to the heterogeneity of quantitative studies reported.

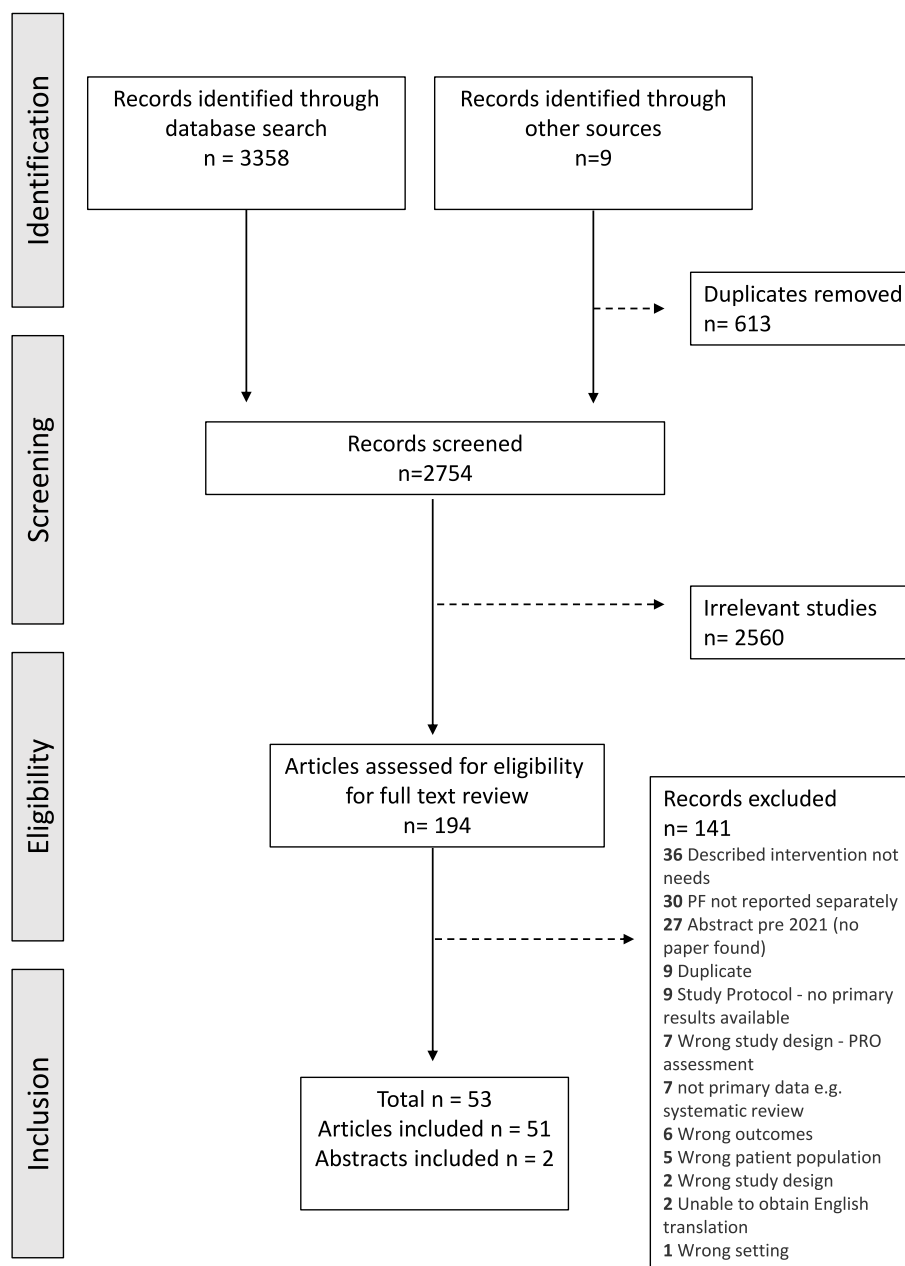


Fig. 1. Study selection flow diagram presented according to Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement. Abbreviations: PF, pulmonary fibrosis; n, number.

3.2. Participant populations

Included studies totalled at least 6718 research participants, comprising at least 3036 patients (45%), 1630 (24%) caregivers and 1745 HCP (26%). 253 'other' participants (4%) were accounted for, including relatives [26], patient advocacy group board members [47] and experts in industry and policy [32,48]. One study reported on responses from an online forum/blog on an interactive PF website, as such it was not possible to determine the role of individual participants [35]. On two occasions, 2 published references reported on a single participant cohort [4,49–51], whilst a further two studies pooled patient and caregiver numbers together [48,52]. The included abstracts did not provide details of the numbers of PwPF/caregivers and HCPs involved in their FG and semi-quantitative interviews [22,23].

Over half of all studies assessed both PwPF and caregiver disease experience (29/53, 55%) several of which reported the percentage of

patients interviewed with a caregiver present [9,27,50,51]. 2 studies reported dyadic interviewing [10,53]. A smaller number of studies focused on either the PwPF (n = 5) or caregiver (n = 8) individually. Where stated, 32% of patients required supplementary oxygen and 55% of PwPF and 40% of IPF patients were female.

19 studies examined HCP opinion on supportive care needs, either in isolation (n = 5) [29,44–46,54] or in combination with other research participants (n = 14). HCP comprised physicians, primary care providers, nurses, physiotherapists and occupational therapists.

Approximately half of all studies (24/53) solely recruited participants from clinics at least 18 of which were specialist clinics, and 13 solely from organisations such as support, association and advocacy groups. Six recruited from both clinics and organisations. Other recruitment settings included a national IPF registry [55], via postings on online PF forums [35], at mandatory government interviews [56] and attendees of information meetings [57].

23/53 studies recruited PwPF and caregivers exclusively affected by Idiopathic pulmonary fibrosis (IPF) and 2/5 studies examining HCP opinion enquired exclusively about the experiences of IPF patients [44, 46]. A small number of studies focused on other specific causes of PF, including systemic sclerosis (SSc) (n = 5) [24,34,36,43,52], silicosis (n = 1) [41] and occupational PF (n = 1) [56]. The remaining studies recruited heterogeneous PF populations, including a small number of those with hypersensitivity pneumonitis (n = 24 PwPF), idiopathic non-specific interstitial pneumonitis (n = 6 PwPF), fibrotic organising pneumonia (n = 1 PwPF), unclassifiable (n = 3 PwPF), other connective tissue disease associated PF (n = 58 - phenotype not further defined) and sarcoidosis (n = 1) (Supplementary Table 1 for further details). It was not possible to perform planned subgroup analyses based on PF subtype and disease severity due to these small numbers.

3.3. Quality assessment

Full details of the MMAT quality assessment are included in Table 1.

Quality assessment of qualitative studies was generally good, with strengths of presenting clear research questions and coherence between data sources, analysis and interpretation. Weaknesses mainly surrounded the sampling strategy (convenience (n = 13) and purposive sampling (n = 7) commonly reported) with risk of selection bias and potential lack of generalisability of findings. The analytical theoretical framework used was not reported in 4 studies [32,51,52,58]. Quantitative studies presented clear research questions and reported appropriate measurement methods but were often subject to risk of response bias (denominator of surveys/questionnaires infrequently known), selection bias (e.g., self-selection to participate or invitation to patient organisations where individuals may be more highly motivated with good disease insight). Furthermore, ethnic and cultural diversity was limited across all studies. Included abstracts [22,23] lacked detail compared to full text articles, and as such scored lower in terms of the quality assessment criteria used.

3.4. Supportive care needs

There was considerable overlap with identified issues frequently impacting upon several of the proposed domains. Therefore, some issues are discussed under multiple sections.

3.4.1. Physical

'Even brushing my teeth is an exertion' ... [31]

Shortness of breath and cough were the most prevalent symptoms reported in the included studies. These symptoms were distressing and had a significant impact on functional ability and influenced other domains as discussed below. Uncontrolled cough can lead to incontinence [59] and impaired sleep for both PwPF and caregiver [4,8]. This can frustrate family members, leading to feelings of guilt and difficulties at work [8,17], as well as driving hypervigilance and anxiety, particularly when prolonged bouts of coughing occur [60].

For PwPF fatigue and exhaustion were prominent symptoms [9,31], often contributed to by insomnia or interrupted sleep [4,31].

There was general acknowledgement that interventions were helpful for symptomatic relief, such as supplemental oxygen [51,55] and medications [27,54,58], although there are often significant consequences and barriers to accessing such treatments. For example, two Japanese quantitative studies demonstrated less use of opiates for IPF patients than lung cancer patients [44,45].

There were also suggestions that non-specialist HCPs may lack the experience and confidence to prescribe appropriate doses of palliative medication [4,54].

3.4.2. Emotional

'I can't pull myself together; this is what's most exasperating' ... [10]

Many PwPF and caregivers expressed frustration at misdiagnosis and

diagnostic delay [8–10,30,37,47,50,51,53,59,61]. They reported that community teams seem to lack awareness of PF and that early symptoms were either not adequately investigated or were put down to alternative problems [10,48,50,53,59]. Initial uncertainty appeared to promulgate the frustration and contribute to feelings of isolation and loneliness experienced throughout the illness [50].

Most PwPF and caregivers experienced significant negative emotions at diagnosis such as low mood, hopelessness and anxiety [53,56], alongside shock and fear when it was made clear that PF is a progressive and terminal diagnosis [31,61].

Throughout the illness, HCP recognise depressive symptoms as one of the most common comorbidities [29]. Quantitative studies highlight the prevalence of mental health symptoms in PF (Supplementary Table 1). One study demonstrated all 100 of included people with IPF suffered with depressive symptoms, with 39 % 'extreme' as assessed with the Beck Depression Inventory [42]. Two thirds of PwPF suffered with symptoms of anxiety as defined by the validated Generalized Anxiety Disorder single item questionnaire (GAD-SI) in another study [40]. The impact of PF can be far reaching with depressive symptoms as assessed by the Center for Epidemiologic Studies Depression Scale (CES-D) extending to involve 68 % of caregivers of patients with silicosis [41]. Further interpretation of these studies is hindered by the use of screening tools to determine mental health morbidity without follow up with diagnostic interviews or evaluation.

Conversely, five studies referenced initial relief at diagnosis [8,51, 62–64], favouring the diagnosis of PF over their own preconceived fears about lung cancer. This was invariably followed by 'confusion' upon realisation of the terminal nature of some forms of PF [51,61,62]. Such misunderstanding of PF is widespread throughout society, with PwPF reporting that friends, family and society misunderstood the diagnosis, leading to feelings of isolation [40]. One person with IPF commented that 'if I had cancer, people would empathise more' [51].

3.4.3. Psychological

I have never received any psychological support, I really need it' ... [9]

The lack of available psychological and emotional support for both PwPF [9,25,32,34,52,64] and caregivers [17,28,32] was prevalent throughout the included literature. In Russell et al.'s [9] multinational study of 45 IPF patients, only 1 in 5 had received professional psychological support. Included quantitative research indicated 80 % of Dutch and 79 % German patients felt psychological support was lacking in current care [40]. Lack of support was recognised as a barrier to the involvement of specialist PC services by HCP in a Japanese study [45].

PwPF and caregivers expressed desire for access to formal psychological support in numerous studies including psychologists or counselling social workers [30], peer support groups [50,51,65] and as part of pulmonary rehabilitation [47]. Recognition of the benefits of a caregiver only support group were described [17,53,66]. This may allow more frank discussions between caregivers about the challenges faced, providing a safe space to say things 'I could never say' otherwise [17].

Some participants suggested that there are downsides to peer support. Being in the presence of people less well than themselves may be perceived as a threat to a person's ability to cope [62] and whilst supportive relationships can be formed, the loss of attendees (or drop out from groups) can be 'horrendous', even if online [63,66]. Others may simply decline support group services as they may feel so negative - 'I drag everyone down' [56].

Only one study assessed the impact of the CoViD 19 pandemic upon PwPF and caregivers' QoL and wellbeing [25]. This reported higher levels of worry during the pandemic than the general public, with concerns about hospital attendance. Fear of infection was also described pre-pandemic [31,53].

The psychological and practical burdens of the introduction of supplementary oxygen were recurring themes. Oxygen therapy is described numerous times as a visual and audible cue to illness which can contribute to feelings of shame and stigmatisation [9,48,51,53,67] and

Table 1

Methodological quality as assessed using the Mixed Methods Appraisal Tool (MMAT). Each criterion was graded as met (Y, green), not met (N, red) or can't tell (CT, amber).

Study	Type	Is there a clear research question?	Does the collected data allow the research question to be addressed?	1.1 Is the qualitative approach appropriate to answer the research question?	1.2 Are the qualitative methods adequate to address the research question?	1.3 Are the findings adequately derived from the data?	1.4 Is the interpretation of results sufficiently substantiated by data	1.5 Is there coherence between data sources, collection, analysis and interpretation?
Ahmed 2023	Qualitative interview study	Y	Y	Y	Y	Y	Y	Y
Albright 2016	Qualitative study of blogs/forum	Y	Y	Y	CT	Y	Y	Y
Bajwah 2013 ¹	Qualitative interview study	Y	Y	Y	CT	Y	Y	Y
Bajwah 2013 ²	Qualitative interview study	Y	Y	Y	CT	Y	Y	Y
Belkin 2014	Qualitative analysis of FG	Y	Y	Y	Y	Y	Y	Y
Bonella 2016	Qualitative interview study	Y	Y	Y	Y	Y	Y	Y
Bridges 2015	Qualitative analysis of interviews	Y	Y	Y	Y	Y	Y	CT
Burnett 2019	Qualitative analysis of interviews	Y	Y	Y	Y	Y	Y	Y
Caminati 2021	Expert panel + qualitative interviews •	Y	Y	Y	Y	CT	CT	Y
Cassidy 2021 ¹	World café with interviews	Y	Y	Y	Y	Y	Y	Y
Cassidy 2021 ^{2*}	Qualitative interviews	Y	Y	Y	CT	CT	CT	CT
Dean 2021	Qualitative FG	Y	Y	Y	Y	CT	CT	CT
Delameillieure 2021	Qualitative interview	Y	Y	Y	Y	CT	CT	CT
Denton 2021	Qualitative interviews and observed conversations	Y	Y	Y	Y	Y	Y	Y
Duck 2015	Qualitative interviews	Y	Y	Y	Y	Y	CT	Y
Egerod 2019	Qualitative interview, field notes and diary	Y	Y	Y	Y	Y	Y	Y
Giot 2013	Qualitative interviews	Y	Y	Y	CT	CT	CT	CT
Graney 2017	Qualitative interviews	Y	Y	Y	Y	CT	CT	Y

Grant-Orser 2022*	Qualitative FG	Y	Y	Y	Y	Y	Y	Y
Hoffmann-Vold 2021	Qualitative interviews	Y	Y	Y	Y	CT	Y	Y
Holland 2015	Qualitative Interview	Y	Y	Y	Y	Y	Y	Y
Kalluri 2021*	Qualitative interviews	Y	Y	Y	Y	Y	Y	Y
Kalluri 2022	Qualitative interviews	Y	Y	Y	Y	CT	CT	Y
Lindell 2017	Qualitative FG	Y	Y	Y	Y	Y	Y	Y
Li 2021	Qualitative Interview	Y	Y	Y	Y	CT	CT	Y
Masefield 2019	Qualitative FG	Y	Y	Y	CT	CT	Y	Y
Morriset 2016	Qualitative FG and interview	Y	Y	Y	Y	Y	Y	Y
Overgaard 2016	Qualitative Interview	Y	Y	Y	Y	CT	CT	CT
Pooler 2018	Qualitative interviews	Y	Y	Y	Y	Y	Y	Y
Ramadurai 2018	Qualitative FG	Y	Y	Y	Y	CT	CT	CT
Ramadurai 2019*	Mixed Open ended questions	Y	Y	Y	N	Y	Y	Y
Russell 2016	Qualitative interviews	Y	Y	Y	Y	CT	Y	Y
Russo 2022	Qualitative interviews	Y	Y	Y	Y	Y	Y	Y
Sampson 2015	Qualitative interviews	Y	Y	Y	Y	Y	Y	Y
Scerri 2022	Qualitative interviews	Y	Y	Y	Y	Y	Y	Y
Schoenheit 2011	Qualitative interviews	Y	Y	Y	Y	Y	Y	Y
Senanayake 2018	Qualitative interviews	Y	Y	Y	Y	Y	Y	Y
Shah 2018*	Qualitative FG	Y	Y	Y	Y	Y	Y	Y
Swigris 2005	Qualitative FG and interview	Y	Y	Y	Y	CT	CT	Y
Trapman 2023	Qualitative FG and interview	CT	CT	Y	CT	CT	CT	CT
Trapman 2023	Qualitative FG and interview	CT	CT	Y	CT	CT	CT	CT
Wuyts 2020*	Delphi consensus	Y	Y	Y	Y	Y	Y	Y

Qualitative Studies

Study	Is there a clear research question?	Does the collected data allow the research question to be addressed?	2.1 Is randomisation appropriately performed?	2.2 Are the groups comparable at baseline?	2.3 Are there complete outcome data?	2.4 Are outcome assessors blinded to the intervention provided?	2.4 Did the participants adhere to the assigned intervention?
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Randomised controlled trials

Study	Is there a clear research question?	Does the collected data allow the research question to be addressed?	3.1 Are the participants representative of the target population?	3.2 Are measurements appropriate regarding both the outcome and intervention?	3.3 Are there complete outcome data?	3.4 Are the confounders accounted for in the design and analysis?	3.5 During the study period, is the intervention administered as intended?
Shaheen 2021	Y	Y	N	Y	Y	CT	Y

Quantitative non-randomised trials

Study	Is there a clear research question?	Does the collected data allow the research question to be addressed?	4.1 Is the sampling strategy relevant?	4.2 Is the sample relevant to target population?	4.3 Are the measurements appropriate?	4.4 Is the risk of non-response bias low?	4.5 Is the statistical analysis appropriate to answer the research questions?
Akiyama 2020	Y	Y	Y	CT	CT	Y	Y
Cassidy 2021 ² *	Y	Y	N	CT	Y	N	CT
Collard 2007	Y	Y	Y	CT	Y	CT	Y
Cottin 2021	Y	Y	Y	CT	Y	N	Y
Fujisawa 2023	Y	Y	Y	Y	Y	CT	Y
Galetti 2021	Y	Y	N	N	Y	N	CT
Grant-Orser 2022*	Y	Y	CT	CT	Y	N	Y
He 2019	Y	Y	CT	CT	Y	CT	Y
Kalluri 2021*	Y	Y	CT	CT	Y	CT	Y
Lindell 2023	Y	Y	Y	CT	Y	CT	Y
Ramadurai 2019*	Y	Y	N	CT	Y	N	Y
Rosato 2014	Y	Y	N	Y	Y	N	Y
Sato 2023	Y	Y	Y	CT	Y	CT	Y
Shah 2018*	Y	Y	CT	CT	Y	N	Y
Van Manen 2017	Y	Y	CT	CT	Y	CT	Y
Wuyts 2020*	Y	Y	Y	CT	Y	CT	Y

Quantitative descriptive trials

Study	Is there a clear research question?	Does the collected data allow the research question to be addressed?	5.1 Is there an adequate rationale for using a mixed methods design to address the research question?	5.2 Are the different components of the study effectively integrated to answer the research question?	5.3 Are the outputs of the integration of qualitative and quantitative components adequately interpreted?	5.4 Are divergences and inconsistencies between quantitative and qualitative results adequately addressed?	5.5 Do the different components of the study adhere to the quality criteria of each tradition of the methods involved?
Cassidy 2021 ² *	Y	Y	Y	Y	Y	Y	CT
Grant-Orser 2022*	Y	Y	Y	Y	Y	Y	Y
Kalluri 2021*	Y	Y	Y	Y	Y	Y	Y
Ramadurai 2019*	Y	Y	Y	Y	Y	Y	N
Shah 2018*	Y	Y	Y	Y	Y	Y	N
Wuyts 2020*	Y	Y	Y	Y	Y	Y	CT

Mixed Methods studies

is viewed forebodingly by PwPF not yet requiring it [65]. It impacts on both caregiver and PwPF perception of self, implying increasing vulnerability and can be a depressing reminder of deterioration as oxygen requirements increase [8,53,67]. The practical limitations of oxygen therapy are discussed below and contribute to social isolation with the associated impact on QoL. Cassidy et al. highlighted the commencement of home oxygen therapy as one of the significant touchstone moments with a lack of available psychological support at a distressing time for PwPF and their loved ones [32].

3.4.4. Social/societal

'I cannot have a social life. I am useless' ... [56]

A lack of societal awareness of PF was well reported by PwPF [30,40,47,48,50,63]. PwPF and caregivers volunteered that they felt PF was poorly understood by non-specialist HCP [8,48,52,68], likely contributing to the frequent delays in diagnosis or misdiagnosis and under-utilisation of PC.

Whilst the benefits of PC are frequently mentioned in the included qualitative work [8,26,32,55,58,59,62,63,68], access to these services appear limited. PwPF and societal preconceptions about opiates and PC present obstacles [32,54] with one patient stating 'I do not want to be a heroin addict' [54], and another referring to palliative care as 'that path to death' [62]. Only 2 PwPF out of 27 in Sampson's UK based study had received palliative care despite 90 % requiring oxygen, with both reporting positive experiences [62]. 25 % of 1023 HCP included in a Japanese study had referred patients to palliative care services, despite recognising the burden of symptoms and nearly half being aware of the availability of PC services [45]. A quantitative study in South Carolina suggests that only around 30 % of 32 responding pulmonologists strongly agreed they are comfortable discussing prognosis and PC referral for PwPF, with none reporting referral at diagnosis [46]. Patients value PC support; five out of 8 PwPF included in a Canadian study recommended PC referral at presentation with one citing the reasoning as it can 'help you live until you die' [58].

The inclusion of studies of SSc patients allows for a closer look at a younger cohort of PwPF for whom the condition may carry occupational consequences. Those with lung involvement are at higher risk of loss or change of job [36]. Half of caregivers in this study reported an economic impact of the PwPF illness, again more likely if there was lung involvement. Some people with SSc-PF made efforts to hide their illness from the workplace due to concerns over its impact on employability [34]. Scerri et al. looked at the burden of caregiving for a parent with IPF (daughters with mean age of 41.2) [60]. Half had their own young

families, and all were initially in employment. They described economic impacts with 2 out of 6 leaving their jobs and the 4 that kept working fulltime reported high levels of exhaustion and stress.

3.4.5. Information/education

'I don't understand it near as well as I feel I need to' ... [8]

Ramadurai conducted an online survey of 160 IPF patients, 29 caregivers and 182 HCP to identify what informational needs were deemed most important [28]. Both PwPF and caregivers listed 'disease progression/what to expect' as their single most important informational need. When presented with open ended suggestions, patients volunteered a need for information on 'emotional and psychosocial support' and the 'practicalities of portable oxygen use'. Caregivers wished for support in 'how to communicate with a frustrated, angry, depressed patient', amongst other suggestions. Similarly, Japanese quantitative work demonstrated PwPF and caregiver primary informational need was also for 'disease progression and what to expect' [38]. The top three informational needs of both PwPF and caregivers in this study were the same, differing only beyond this.

Consistent with these findings, Denton et al. showed that the 'unspoken topics' specifically around SSc-PF prognosis and mortality were of great importance to PwPF and caregivers [24]. This study was unique in that it analysed clinical communication between groups. Results suggested that PwPF feel their informational and educational needs are unmet. Observation of mock consultations concluded that physicians tend to lead consultations and during interview some admitted to trying to avoid questions about mortality and prognosis, for fear of damaging hope. This fear of negatively impacting upon hope was also described elsewhere [28,49,69]. PwPF may feel uncomfortable, intimidated or embarrassed to ask specific questions which may be of importance to them, and both HCP and PwPF reported limited consultation time as a barrier to effective communication [24]. Alongside Ramadurai's work [28], this highlights the primary informational needs of PwPF and throws light upon how these are unmet.

Time was identified as a barrier to the provision of verbal information and opportunity for questions [24,50,54,61,64]. It follows that supplementary educational materials need to be readily available and from a trustworthy source [61], although the preferred style, mode and timing of information delivery varies. The value of reliable written information as a future reference was acknowledged [55,61]. These can be '[taken] away and once the initial shock has worn off you can actually look at it' [62], especially when there may be difficulty remembering what was discussed in clinic [52]. Non-IPF patients can feel that much of

the educational material available is targeted at patients with IPF [30].

PwPF demonstrated awareness that the internet may be a misleading and overwhelming information source [24,30,53,62,69,70]. However, it can also provide a useful platform to share experiences and offer peer support, with the benefit of online anonymity allowing some to discuss more sensitive issues such as intimacy [35]. Signposting to trustworthy online sources was proposed by PwPF [61].

Studies reported positive patient and caregiver experiences with support groups as information sources [26,53] and other studies looked at the utility of pulmonary rehabilitation courses as a platform for patient education [30,55,70]. HCP recognise that poor disease education and information provision can significantly contribute towards barriers to accessing PC [45].

3.4.6. Spiritual

'I try to do everything that has a meaning' ... [27]

Findings were limited with respect to the spiritual needs of both PwPF and their caregivers. Nonetheless, 'love' was proposed as a coping strategy for the illness and challenges of oxygen therapy [51,67]. An Irish 2021 world café analysis of PwPF reported on a failure to meet individuals' spiritual beliefs near the end of life, with many unaware of the various support options potentially available [32]. Effective symptom control can help convey meaning into PwPF's lives [69] and one study reported on the positive effects of people with IPF on sense of spiritual self and appreciation of family togetherness [31]. The same study however also commented that IPF could increase strain on family relationships, suggesting several complex internal and external factors at play.

3.4.7. Practical

'Our life revolves around him constantly' ... [65]

The introduction of home oxygen therapy brings with it considerable practical issues, for example impact on ability to travel and loss of spontaneity [9,31,50,53,55,59,65]. Caregivers often need to assume responsibility for forwards planning, packing and maintaining oxygen delivery devices [53,67]. Words such as 'tethered', 'tied', 'caged' and 'on a leash' were used to describe the experience [31,50,67] with impact on a variety of other domains.

Appropriate advanced care planning (ACP) and EOL planning are dependent upon efficient and open communication with PwPF and caregivers to ensure their understanding of the underlying disease process and prognosis is solid. There was no unifying agreement on when and how to commence discussions and preferences vary from individual to individual, with this heterogeneity further complicated by the unpredictable nature of PF. HCP therefore may feel these conversations are held later than would be ideal [45].

Qualitative findings vary, with PwPF participants within the same publications expressing differing preferences around ACP and EOL planning. Clearly such discussions are emotive and challenging with 2 of the included studies describing PwPF perception of confrontation when discussing the disease and ACP [64,68]. On the other hand, other studies suggest PwPF desire early discussions [4,22,49,69], with participants suggesting that the onus for initiating discussions should reside with the HCP [22,58]. Lindell's study assessed PwPF and caregiver perceptions of ACP and familiarity with PC [8]. Participants expressed hesitancy in discussing the practicalities of ACP and misconceptions about the goals of PC. One caregiver became distressed when discussing PC - 'I didn't know this had anything to do with PC, and I do have a problem focusing on the fatality' [8].

Egerods' work indicates the benefits of early ACP and EOL care [33]. Caregiver participants were distressed if PwPF were taken to unfamiliar or unplanned places to die. Home deaths can be overwhelming to caregivers but eased by the involvement of homecare nurses. Dean et al. examined hospice provided caregiver support groups and all participants reported positive experiences [66]. Another publication identified the benefits to early consideration of financial and legal matters as well

as funeral planning, extending from beyond practical to emotional - '[she] made her final wishes known, and that helped my sister and I' [58].

3.4.8. Family related

'It changes the dynamics of your marriage' ... [17]

Whilst we have reported on the benefits of the supportive family network above, PwPF and caregivers commented on numerous potential sources of conflict. Caregivers can struggle with evolving roles particularly in the early days before patients experience functional deterioration; 'I'm confused, because I become this caregiver that isn't giving any care, but more of a nag' [61]. Overbearing and protective behaviour from family members can contribute to physical deconditioning and friction with PwPF being critical of what they can view as usurping their role [4,29,63]. This is well illustrated by a remark made in an interview by an IPF patient 'my family checks everything I do, I don't feel free' [9].

As symptoms progress their influence on financial, social and relationship dynamics grows and patients can vent frustrations at family members [51,56]. Additionally, the ability to socialise and enjoy previous hobbies is compromised and can further generate feelings of loss [62]. This introduces new strains into marital relationships; one caregiver remarked, 'we don't really talk about interesting things anymore. It changes the dynamics of your marriage' [17]. Graney described the phenomenon of 'shrinking world syndrome' affecting both patient and caregiver, mirrored by a caregiver in another study remarking 'my world is getting smaller and smaller' [56,67]. Family members who harbour any negative emotions towards the patients' needs may feel remorseful and guilty for doing so [8,60,65].

As the illness progresses, caregivers sacrifice more and take on greater responsibility - 'things we used to split. Now I had to do them all' [17]. Whilst some patients reported the experience drew them closer to their family members [31], findings were more frequently illustrative of the strain the illness places on family dynamics. This is a sensitive area, Overgaard et al. reported that when a change in family roles was discussed in dyadic interviews, it could feel at times like confrontation particularly if this hadn't been discussed previously [10]. Sampson reported that male caregivers may feel that gender roles are being reversed, further challenging a caregiver's sense of self [62].

A loss of libido was acknowledged in 4 studies [9,31,35] and female SSc-PF patients scored poorly in the Dyadic Adjustment Scale (DAS); a self-reported measure designed to assess relationship satisfaction [43]. It is likely such sensitive issues are under reported in populations where there is a reticence to discuss 'intimate' issues.

4. Discussion

This systematic literature review identified 53 studies assessing the needs of PwPF, caregivers and the perceptions of HCP with respect to PF, predominantly IPF, across 4 continents. Findings highlight a lack of psychological support throughout the illness, misconceptions and barriers to the provision of PC and a desire amongst PwPF and caregivers for greater disease specific education and information provision throughout the illness.

The introduction of oxygen is a significant event for PwPF and caregivers. Our findings align with those of a recent systematic review analysing barriers and facilitators to oxygen therapy use in PwPF [71]. This highlighted the psychological and domestic impact of oxygen therapy and concluded that PwPF need more information and support, including psychological support, at initiation, as well as calling for technological innovations to reduce the practical burdens of oxygen therapy.

Lack of informational resources and education was a major recurrent theme. The unpredictable and variable nature of PF, compounded with varying individual backgrounds and preferences, presents challenges in the provision of disease specific information. This is further complicated by the likely under-representation of certain groups within the

literature. Whilst a nurse led palliative care intervention has been shown to be an effective means of improving PwPF and caregiver knowledge and preparedness [72], all participants were Caucasian and 24 % of those eligible and interested declined due to fatigue levels. The authors suggest future consideration of alternative delivery methods, for example telemedicine which may widen access.

Symptom control leads to improved QoL with positive impacts on all supportive care need domains described above, yet PwPF receiving PC support appeared underrepresented in the literature. Recent British Thoracic Society (BTS) registry data suggests 83 % of IPF patients have their PC needs assessed in the UK [73], however this data relates to IPF patients and is incomplete.

This review highlights several obstacles to the involvement of PC in PF, both PwPF and HCP dependent. Stigma associated with PC has been described in the cancer literature [74,75], and our findings show that participants strongly associated PC with cancer and death. The benefits of PC were well described in the included qualitative literature and a recent study has demonstrated improved survival in people with IPF receiving integrated PC [76]. Interestingly, people with cancer who received PC in Zimmerman's work felt strongly that a specialty name change was necessary [75]. Future work should look to draw upon these findings to explore means of 'destigmatizing' PC, removing associations with cancer, death and addiction. Cassidy et al. proposed a 'training needs analysis' for EOL planning for HCP involved in the care of PwPF [32] and elsewhere a call for more research into symptomatic treatment and the introduction of evidence based prescribing guidelines specifically for symptomatic progressive PwPF was suggested [4,45].

It should be noted however that studies assessing interventions incorporating palliative and supportive care are not always favourable. A 2020 randomized, controlled trial, comparing usual care to usual care with concomitant PC in IPF patients did not find improvement in the primary outcomes of QoL, anxiety and depression at 6 months [77]. Older work demonstrated a deterioration in health-related QoL following a support group intervention [78]. These findings should be treated with caution; the authors suggested there may be selection bias due to the preconceptions surrounding PC. Benefits reported during a qualitative arm in the latter work may suggest that patient reported outcome measures are less sensitive to benefits better uncovered by in depth qualitative research. Benefits reported in this systematic review would reflect this.

The emotional burden and psychological impact of PF was clearly documented in this review. Moreover, the desire for both PwPF and carers to access formal psychological support was a dominant theme. Despite this, beyond breathlessness focussed work [79] there are no systematically trialled complex interventions to address PwPF and carer psychosocial needs in PF. These should be considered a priority area for research and development. PwPF reported positive experiences with peer support groups, with added value given to those held in a private setting [66], whilst Shah's work [10] suggested that separated sessions for PwPF and caregivers may allow more open dialogue.

The introduction of complex psychosocial interventions to support patients and carers with progressive and life limiting diseases has been studied in other diseases [80,81]. Published reviews of psychosocial interventions for patients and carers affected by cancer, identify a broad range of approaches [81,82]. Psychoeducation intervention (with opportunity for skill development e.g. coping skills training) is the dominant approach, with positive impact on patient and carer QoL [81,82]. Psychologist-led therapeutic counselling sessions for patient/carer pairs, underpinned by PC input, led to improvements in coping and communication between pairs [82]. Other strategies reported in the literature [81,82] include mindfulness, cognitive behavioural therapy and arts-based therapy. Unlike trajectories in dementia or cancer, the PF disease course is uniquely characterised by periods of stability, sporadically dispersed with acute episodes of unanticipated 'worsening' (exacerbations) that are life threatening and may result in costly hospitalisation episodes [83]. This poses a unique set of challenges for PwPF

and carers. It is hoped that the synthesis of this data can be used to inform the development of bespoke evidence based-field tested programmes to address these needs that can be tested and then scaled to reach global populations.

A recently published scoping review examining the needs of patients with IPF [16] compliments the findings of this review, which includes an expanded patient cohort. This particularly highlighted the psychological burden associated with a diagnosis of IPF, underutilisation of PC resources and the gap between informational needs and provision.

Whilst the strengths of the present study include its robust search strategy and thus broad genesis of data across different settings, it does have some limitations. There is a lack of data addressing those with non-IPF diagnoses, who may have distinct unmet needs due to differences in demographics and prognosis. Furthermore, the decision to include global studies allows for a rich data extraction but is complicated by different healthcare provision and culture which can threaten generalisability.

5. Conclusions

This systematic review with narrative synthesis of current research, presents the current landscape of PwPF and caregiver needs. It highlights persistent unmet needs, despite a rapid growth in research interest in recent years. This suggests that interventions to address these supportive care needs may not have kept pace with developments elsewhere in the disease e.g., development of medical therapies. The volume and intensity of these needs can be expected to increase in the era of widening access to antifibrotic and other novel medications which may also present new challenges such as the management of side effects [9, 48,53,55,70]. Systematically trialled complex interventions are needed to address the unique set of challenges for PwPF and carers living with PF.

Funding

No external funding was received for this study.

CRediT authorship contribution statement

Matthew Wells: Writing – original draft, Validation, Formal analysis, Data curation. **Sam Harding:** Writing – review & editing, Supervision, Methodology, Data curation. **Giles Dixon:** Writing – review & editing, Software, Data curation. **Kirsten Buckley:** Investigation. **Anne-Marie Russell:** Writing – review & editing. **Shaney L. Barratt:** Writing – review & editing, Writing – original draft, Supervision, Project administration, Methodology, Formal analysis, Conceptualization.

Declaration of competing interest

S. L. Barratt reports personal fees for an advisory board and financial support to attend an educational conference from Boehringer Ingelheim, outside the submitted work.

The authors have no further declarations.

Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.rmed.2024.107659>.

References

- [1] G. Raghu, D. Weycker, J. Edelsberg, W.Z. Bradford, G. Oster, Incidence and prevalence of idiopathic pulmonary fibrosis, *Am. J. Respir. Crit. Care Med.* 174 (7) (2006 Oct 1) 810–816.
- [2] C. Vancheri, M. Failla, N. Crimi, G. Raghu, Idiopathic pulmonary fibrosis: a disease with similarities and links to cancer biology, *Eur. Respir. J.* 35 (3) (2010 Mar) 496–504.

- [3] C. Bausewein, S. Booth, M. Gysels, R. Kühnrich, B. Haberland, I.J. Higginson, Understanding breathlessness: cross-sectional comparison of symptom burden and palliative care needs in chronic obstructive pulmonary disease and cancer, *J. Palliat. Med.* 13 (9) (2010 Sep) 1109–1118.
- [4] S. Bajwah, I.J. Higginson, J.R. Ross, A.U. Wells, S.S. Birring, J. Riley, et al., The palliative care needs for fibrotic interstitial lung disease: a qualitative study of patients, informal caregivers and health professionals, *Palliat. Med.* 27 (9) (2013 Oct) 869–876.
- [5] P. Edmonds, S. Karlsen, S. Khan, J. Addington-Hall, A comparison of the palliative care needs of patients dying from chronic respiratory diseases and lung cancer, *Palliat. Med.* 15 (4) (2001 Jul) 287–295.
- [6] J.M. Gore, C.J. Brophy, M.A. Greenstone, How well do we care for patients with end stage chronic obstructive pulmonary disease (COPD)? A comparison of palliative care and quality of life in COPD and lung cancer, *Thorax* 55 (12) (2000 Dec) 1000–1006.
- [7] V. Weingaertner, C. Scheve, V. Gerdes, M. Schwarz-Eywill, R. Prenzel, C. Bausewein, et al., Breathlessness, functional status, distress, and palliative care needs over time in patients with advanced chronic obstructive pulmonary disease or lung cancer: a cohort study, *J. Pain Symptom Manag.* 48 (4) (2014 Oct) 569, 81. e1.
- [8] K.O. Lindell, D. Kavalieratos, K.F. Gibson, L. Tycon, M. Rosenzweig, The palliative care needs of patients with idiopathic pulmonary fibrosis: a qualitative study of patients and family caregivers, *Heart Lung* 46 (1) (2017) 24–29.
- [9] A.M. Russell, E. Ripamonti, C. Vancheri, Qualitative European survey of patients with idiopathic pulmonary fibrosis: patients' perspectives of the disease and treatment, *BMC Pulm. Med.* 16 (2016 Jan 14) 10.
- [10] D. Overgaard, G. Kaldan, K. Marsaa, T.L. Nielsen, S.B. Shaker, I. Egerod, The lived experience with idiopathic pulmonary fibrosis: a qualitative study, *Eur. Respir. J.* 47 (5) (2016 May) 1472–1480.
- [11] M. Kalluri, F. Luppi, G. Ferrara, What patients with idiopathic pulmonary fibrosis and caregivers want: filling the gaps with patient reported outcomes and experience measures, *Am. J. Med.* 133 (3) (2020 Mar) 281–289.
- [12] K.Y. Wen, D.H. Gustafson, Needs assessment for cancer patients and their families, *Health Qual. Life Outcome* 2 (2004 Feb 26) 11.
- [13] R. Schulz, S.R. Beach, Caregiving as a risk factor for mortality: the caregiver health effects study, *JAMA* 282 (23) (1999 Dec 15) 2215–2219.
- [14] R. Schulz, J. Newsom, M. Mittelmark, L. Burton, C. Hirsch, S. Jackson, Health effects of caregiving: the caregiver health effects study: an ancillary study of the Cardiovascular Health Study, *Ann. Behav. Med.* 19 (2) (1997) 110–116.
- [15] C.J. Schroedel, S.E. Yount, E. Szmuliowicz, P.J. Hutchison, S.R. Rosenberg, R. Kalhan, A qualitative study of unmet healthcare needs in chronic obstructive pulmonary disease. A potential role for specialist palliative care? *Ann Am Thorac Soc* 11 (9) (2014 Nov) 1433–1438.
- [16] C. Bramhill, D. Langan, H. Mulryan, J. Eustace-Cook, A.M. Russell, A.M. Brady, A scoping review of the unmet needs of patients diagnosed with idiopathic pulmonary fibrosis (IPF), *PLoS One* 19 (2) (2024 Feb 14) e0297832.
- [17] R.J. Shah, H.R. Collard, J. Morisset, Burden, resilience and coping in caregivers of patients with interstitial lung disease, *Heart Lung* 47 (3) (2018) 264–268.
- [18] M.J. Page, J.E. McKenzie, P.M. Bossuyt, I. Boutron, T.C. Hoffmann, C.D. Mulrow, et al., The PRISMA 2020 statement: an updated guideline for reporting systematic reviews, *BMJ* 372 (2021 Mar 29) n71.
- [19] R. Pace, P. Pluye, G. Bartlett, A.C. Macaulay, J. Salsberg, J. Jagosh, et al., Testing the reliability and efficiency of the pilot Mixed Methods Appraisal Tool (MMAT) for systematic mixed studies review, *Int. J. Nurs. Stud.* 49 (1) (2012 Jan) 47–53.
- [20] Q.N. Hong, S. Fàbregues, G. Bartlett, F. Boardman, M. Cargo, P. Dagenais, et al., The Mixed Methods Appraisal Tool (MMAT) version 2018 for information professionals and researchers, *Educ. Inf.* 34 (4) (2018 Dec 18) 285–291.
- [21] M.I. Fitch, Supportive care framework, *Can. Oncol. Nurs. J.* 18 (1) (2008) 6–14.
- [22] L. Trapman, L.M. Dijkman, J.C. Grutters, E. De Graaf, S.C.C.M. Teunissen, Barriers and facilitators when talking about personal values, wishes and needs near the end-of-life. A qualitative exploration of a patient and partner perspective, in: *Idiopathic Interstitial Pneumonias*, European Respiratory Society, 2023, p. PA1718.
- [23] L. Trapman, E. de Graaf, L. Dijkman, J. Grutters, S.C. Teunissen, Barriers and facilitators in exploring patients' values, wishes and needs in end of life, perspective from health care professionals, in: *EAPC Abstracts*, 2023, pp. 250–251.
- [24] C.P. Denton, B. Laird, L. Moros, J.L. Luna Flores, Things left unsaid: important topics that are not discussed between patients with systemic sclerosis, their carers and their healthcare professionals—a discourse analysis, *Clin. Rheumatol.* 40 (4) (2021 Apr) 1399–1407.
- [25] N. Cassidy, D. Sheahan, L. Fox, L. Brown, L. Galvin, E. Cassidy, et al., Perspectives of interstitial lung disease patients and carers during COVID-19, *Ir. Med. J.* 114 (7) (2021 Aug 19) 410.
- [26] A. Grant-Orser, N. Avitzur, J. Morisset, C.D. Fell, K.A. Johansson, Perceptions of genetic testing: a mixed-methods study of patients with pulmonary fibrosis and their first-degree relatives, *Ann Am Thorac Soc* 19 (8) (2022 Aug) 1305–1312.
- [27] M. Kalluri, S. Younus, N. Archibald, J. Richman-Eisenstat, C. Pooler, Action plans in idiopathic pulmonary fibrosis: a qualitative study—'I do what I can do', *BMJ Support. Palliat. Care* 8 (2021 Apr).
- [28] D. Ramadurai, S. Corder, T. Churney, B. Graney, A. Harshman, S. Meadows, et al., Idiopathic pulmonary fibrosis: educational needs of health-care providers, patients, and caregivers, *Chron. Respir. Dis.* 16 (2019) 1479973119858961.
- [29] W.A. Wuyts, S. Papiris, E. Manali, M. Kilpeläinen, J.R. Davidsen, J. Miedema, et al., The burden of progressive fibrosing interstitial lung disease: a delphi approach, *Adv. Ther.* 37 (7) (2020 Jul) 3246–3264.
- [30] J. Morisset, B.P. Dubé, C. Garvey, J. Bourbeau, H.R. Collard, J.J. Swigris, et al., The unmet educational needs of patients with interstitial lung disease. Setting the stage for tailored pulmonary rehabilitation, *Ann Am Thorac Soc* 13 (7) (2016 Jul) 1026–1033.
- [31] J.J. Swigris, A.L. Stewart, M.K. Gould, S.R. Wilson, Patients' perspectives on how idiopathic pulmonary fibrosis affects the quality of their lives, *Health Qual. Life Outcome* 3 (2005 Oct 7) 61.
- [32] N. Cassidy, L. Fox, M. Love, I. Byrne, A.M. Doyle, B. Korn, et al., Fibrotic interstitial lung disease - palliative care needs: a World-Café qualitative study, *BMJ Support. Palliat. Care* (2021 Oct 11).
- [33] I. Egerod, G. Kaldan, S.B. Shaker, M.B. Guldin, A. Browatski, K. Marsaa, et al., Spousal bereavement after fibrotic interstitial lung disease: a qualitative study, *Respir. Med.* 146 (2019 Jan) 129–136.
- [34] A. Caminati, B. Vigone, S. Cazzaglio, P. De Nigris, I. Galetti, S di Nunzio, et al., Expert opinion and patients' in-depth interviews on the impact of pulmonary complications in systemic sclerosis, *Curr. Med. Res. Opin.* 37 (sup2) (2021 Nov) 17–26.
- [35] K. Albright, T. Walker, S. Baird, L. Eres, T. Farnsworth, K. Fier, et al., Seeking and sharing: why the pulmonary fibrosis community engages the web 2.0 environment, *BMC Pulm. Med.* 16 (2016 Jan 12) 4.
- [36] I. Galetti, S di Nunzio, L. Brogelli, V. Mirisola, C. Garbagnati, How do systemic sclerosis manifestations influence patients' lives? Results from a survey on patients and caregivers, *Curr. Med. Res. Opin.* 37 (sup2) (2021 Nov) 5–15.
- [37] H.R. Collard, G. Tino, P.W. Noble, M.A. Shreve, M. Michaels, B. Carlson, et al., Patient experiences with pulmonary fibrosis, *Respir. Med.* 101 (6) (2007 Jun) 1350–1354.
- [38] R. Sato, T. Handa, K. Tanizawa, T. Hirai, Variation in information needs of patients with interstitial lung disease and their family caregivers according to long-term oxygen therapy: a descriptive study, *BMC Pulm. Med.* 23 (1) (2023 Dec 5) 486.
- [39] V. Cottin, E. Bergot, A. Bourdin, H. Nunes, G. Prévot, B. Wallaert, et al., Expectations about treatment of idiopathic pulmonary fibrosis: comparative survey of patients, carers and physicians (the RESPIR French survey), *Respir. Med. Res.* 79 (2021 May) 100811.
- [40] M.J.G. van Manen, J.J.M. Geelhoed, N.C. Tak, M.S. Wijsenbeek, Optimizing quality of life in patients with idiopathic pulmonary fibrosis, *Ther. Adv. Respir. Dis.* 11 (3) (2017 Mar) 157–169.
- [41] J. He, W. Dai, Y. Li, L. He, R. Huang, Frequency of depression-related symptoms in caregivers of patients with silicosis, *J. Healthc. Eng.* 2019 (2019) 6035920.
- [42] H.A. Shaheen, A.F. Elessawy, W. Abdel-Wahed, A.N. Mohamed, R.A. Elhefny, Cognitive impairment and depression in patients with idiopathic pulmonary fibrosis, *Minerva Psychiatr.* 62 (2) (2021 May).
- [43] E. Rosato, C. Rossi, I. Molinaro, M.A. Di Giulio, A.C. Trombetta, A.M. Marra, et al., Sexual distress, sexual dysfunction and relationship quality in women with systemic sclerosis: correlation with clinical variables, *Int. J. Immunopathol. Pharmacol.* 27 (2) (2014) 279–285.
- [44] N. Akiyama, T. Fujisawa, T. Morita, K. Mori, H. Yasui, H. Hozumi, et al., Palliative care for idiopathic pulmonary fibrosis patients: pulmonary physicians' view, *J. Pain Symptom Manag.* 60 (5) (2020 Nov) 933–940.
- [45] T. Fujisawa, N. Akiyama, T. Morita, T. Koyachi, Y. Matsuda, M. Mori, et al., Palliative care for interstitial lung disease: a nationwide survey of pulmonary specialists, *Respirology* 28 (7) (2023 Jul) 659–668.
- [46] K.O. Lindell, M. Madisetti, T. Fasolino, M. Pittman, P. Coyne, T.P.M. Whelan, et al., Pulmonologists' perspectives on and access to palliative care for patients with idiopathic pulmonary fibrosis in South Carolina, *Palliat. Med. Rep.* 4 (1) (2023 Oct 1) 292–299.
- [47] F. Bonella, M. Wijsenbeek, M. Molina-Molina, A. Duck, R. Mele, K. Geissler, et al., European IPF Patient Charter: unmet needs and a call to action for healthcare policymakers, *Eur. Respir. J.* 47 (2) (2016 Feb) 597–606.
- [48] J.F.P. Bridges, V.F. Paly, E. Barker, D. Kervitsky, Identifying the benefits and risks of emerging treatments for idiopathic pulmonary fibrosis: a qualitative study, *Patient* 8 (1) (2015 Feb) 85–92.
- [49] S. Bajwah, J. Koffman, I.J. Higginson, J.R. Ross, A.U. Wells, S.S. Birring, et al., "I wish I knew more ..." the end-of-life planning and information needs for end-stage fibrotic interstitial lung disease: views of patients, carers and health professionals, *BMJ Support. Palliat. Care* 3 (1) (2013 Mar) 84–90.
- [50] G. Schoenheit, I. Becattelli, A.H. Cohen, Living with idiopathic pulmonary fibrosis: an in-depth qualitative survey of European patients, *Chron. Respir. Dis.* 8 (4) (2011) 225–231.
- [51] C. Giot, M. Maronati, I. Becattelli, G. Schoenheit, Idiopathic pulmonary fibrosis: an EU patient perspective survey, *Curr. Respir. Med. Rev.* 9 (2) (2013 Jul 31) 112–119.
- [52] A.M. Hoffmann-Vold, E. Bendstrup, T. Dimitroulas, R. Hesselstrand, A. Morais, R. Peltomaa, et al., Identifying unmet needs in SSC-ILD by semi-qualitative in-depth interviews, *Rheumatology* 60 (12) (2021 Dec 1) 5601–5609.
- [53] L. Ahmed, E. Bergot, G. Prévot, V. Cottin, Ethnographic survey of patients and caregiver's life journey in idiopathic pulmonary fibrosis, *Respir. Med. Res.* 83 (2023 Jun) 100955.
- [54] L. Russo, K. Willis, N. Smallwood, Assisting people with their living, not their dying: health professionals' perspectives of palliative care and opioids in ILD, *Am. J. Hosp. Palliat. Care* 39 (2) (2022 Feb) 211–219.
- [55] K. Burnett, I. Glaspole, A.E. Holland, Understanding the patient's experience of care in idiopathic pulmonary fibrosis, *Respirology* 24 (3) (2019 Mar) 270–277.
- [56] P.W.C. Li, D.S.F. Yu, S.Y.S. Tam, The lived experience of patients and family caregivers in managing pneumoconiosis, *Qual. Health Res.* 31 (9) (2021 Jul) 1645–1656.

- [57] M.J.G. van Manen, M. Kreuter, B. van den Blink, U. Oltmanns, K. Palmowski, E. Brunnemer, et al., What patients with pulmonary fibrosis and their partners think: a live, educative survey in The Netherlands and Germany, *ERJ Open Res.* 3 (1) (2017 Jan).
- [58] C. Pooler, J. Richman-Eisenstat, M. Kalluri, Early integrated palliative approach for idiopathic pulmonary fibrosis: a narrative study of bereaved caregivers' experiences, *Palliat. Med.* 32 (9) (2018 Oct) 1455–1464.
- [59] A. Duck, L.G. Spencer, S. Bailey, C. Leonard, J. Ormes, A.L. Caress, Perceptions, experiences and needs of patients with idiopathic pulmonary fibrosis, *J. Adv. Nurs.* 71 (5) (2015 May) 1055–1065.
- [60] D. Scerri, C. Borg Xuereb, J. Scerri, Caring for a parent with pulmonary fibrosis in Malta: the lived experiences of daughters in mid-adulthood, *Health Soc. Care Community* 30 (1) (2022 Jan) e195–e203.
- [61] D. Ramadurai, S. Corder, T. Churney, B. Graney, A. Harshman, S. Meadows, et al., Understanding the informational needs of patients with IPF and their caregivers: "You get diagnosed, and you ask this question right away, what does this mean?", *BMJ Open Qual.* 7 (1) (2018) e000207.
- [62] C. Sampson, B.H. Gill, N.K. Harrison, A. Nelson, A. Byrne, The care needs of patients with idiopathic pulmonary fibrosis and their carers (CaNoPy): results of a qualitative study, *BMC Pulm. Med.* 15 (2015 Dec 4) 155.
- [63] S. Senanayake, K. Harrison, M. Lewis, M. McNarry, J. Hudson, Patients' experiences of coping with Idiopathic Pulmonary Fibrosis and their recommendations for its clinical management, *PLoS One* 13 (5) (2018) e0197660.
- [64] A. Delameillieure, F. Dobbels, S. Vandekerckhof, W.A. Wuyts, Patients' and healthcare professionals' perspectives on the idiopathic pulmonary fibrosis care journey: a qualitative study, *BMC Pulm. Med.* 21 (1) (2021 Mar 18) 93.
- [65] A. Belkin, K. Albright, J.J. Swigris, A qualitative study of informal caregivers' perspectives on the effects of idiopathic pulmonary fibrosis, *BMJ Open Respir. Res.* 1 (1) (2014 Jan) e000007.
- [66] A. Dean, B. Miller, C. Hewitt, M. Cook, C. Nicell, K. Spicer, Caring for carers: exploring the benefits of group support for carers of people with pulmonary fibrosis, *Prog. Palliat. Care* 29 (1) (2021 Jan 2) 5–10.
- [67] B.A. Graney, F.S. Wamboldt, S. Baird, T. Churney, K. Fier, M. Korn, et al., Informal caregivers experience of supplemental oxygen in pulmonary fibrosis, *Health Qual. Life Outcome* 15 (1) (2017 Jul 1) 133.
- [68] S. Masefield, N. Cassidy, D. Ross, P. Powell, A. Wells, Communication difficulties reported by patients diagnosed with idiopathic pulmonary fibrosis and their carers: a European focus group study, *ERJ Open Res.* 5 (2) (2019 Apr).
- [69] M. Kalluri, S. Orenstein, N. Archibald, C. Pooler, Advance care planning needs in idiopathic pulmonary fibrosis: a qualitative study, *Am. J. Hosp. Palliat. Care* 39 (6) (2022 Jun) 641–651.
- [70] A.E. Holland, J.F. Fiore, N. Goh, K. Symons, L. Dowman, G. Westall, et al., Be honest and help me prepare for the future: what people with interstitial lung disease want from education in pulmonary rehabilitation, *Chron. Respir. Dis.* 12 (2) (2015 May) 93–101.
- [71] G. Tikellis, M. Hoffman, C. Mellerick, A.T. Burge, A.E. Holland, Barriers to and facilitators of the use of oxygen therapy in people living with an interstitial lung disease: a systematic review of qualitative evidence, *Eur. Respir. Rev.* 32 (169) (2023 Sep 30) 230066.
- [72] K.O. Lindell, S.J. Klein, M.S. Veatch, K.F. Gibson, D.J. Kass, M. Nouraie, et al., Nurse-led palliative care clinical trial improves knowledge and preparedness in caregivers of patients with idiopathic pulmonary fibrosis, *Ann Am Thorac Soc* 18 (11) (2021 Nov) 1811–1821.
- [73] British Thoracic Society, BTS ILD registry annual report 2021 12 (5) (2021 Dec).
- [74] M.J. Shen, J.D. Wellman, Evidence of palliative care stigma: the role of negative stereotypes in preventing willingness to use palliative care, *Palliat. Support Care* 17 (4) (2019 Aug) 374–380.
- [75] C. Zimmermann, N. Swami, M. Krzyzanowska, N. Leighl, A. Rydall, G. Rodin, et al., Perceptions of palliative care among patients with advanced cancer and their caregivers, *Can. Med. Assoc. J.* 188 (10) (2016 Jul 12) E217–E227.
- [76] J. Lu-Song, J.A. Bakal, S. Younus, O. Moran-Mendoza, I. Harle, M. Morales, et al., The impact of integrated palliative care on survival in idiopathic pulmonary fibrosis: a retrospective multicenter comparison, *Am. J. Hospice Palliat. Med.* (2023 Aug 8).
- [77] K. Janssen, D. Rosielle, Q. Wang, H.J. Kim, The impact of palliative care on quality of life, anxiety, and depression in idiopathic pulmonary fibrosis: a randomized controlled pilot study, *Respir. Res.* 21 (1) (2020 Jan 3) 2.
- [78] K.O. Lindell, E. Olshansky, M.K. Song, T.G. Zullo, K.F. Gibson, N. Kaminski, et al., Impact of a disease-management program on symptom burden and health-related quality of life in patients with idiopathic pulmonary fibrosis and their care partners, *Heart Lung* 39 (4) (2010) 304–313.
- [79] S. Booth, R. Ryan, A. Spathis, Service delivery of complex interventions for refractory breathlessness, *Curr. Opin. Support. Palliat. Care* 10 (3) (2016 Sep) 228–235.
- [80] L.S. Evangelista, A. Strömberg, J.N. Dionne-Odom, An integrated review of interventions to improve psychological outcomes in caregivers of patients with heart failure, *Curr. Opin. Support. Palliat. Care* 10 (1) (2016 Mar) 24–31.
- [81] C.J. Treanor, Psychosocial support interventions for cancer caregivers: reducing caregiver burden, *Curr. Opin. Support. Palliat. Care* 14 (3) (2020 Sep) 247–262.
- [82] W. Xiao, K.M. Chow, W.K.W. So, D.Y.P. Leung, C.W.H. Chan, The effectiveness of psychoeducational intervention on managing symptom clusters in patients with cancer: a systematic review of randomized controlled trials, *Cancer Nurs.* 39 (4) (2016) 279–291.
- [83] V. Cottin, R. Teague, L. Nicholson, S. Langham, M. Baldwin, The burden of progressive-fibrosing interstitial lung diseases, *Front. Med.* 9 (2022) 799912.