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Efeitos das Intervenções de Autogestão em Pessoas com Doença Pulmonar Intersticial – Revisão Sistemática e Meta-Análise

Effects of Self-Management Interventions in People with Interstitial Lung Disease - Systematic Review and Meta-Analysis

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Dissertação apresentada à Universidade de Aveiro para cumprimento dos requisitos necessários à obtenção do grau de Mestre em Fisioterapia, ramo Respiratória, realizada sob a orientação científica da Doutora Ana Oliveira, Professora Adjunta da Escola Superior de Saúde da Universidade de Aveiro e coorientação científica da Doutora Alda Marques, Professora Coordenadora da Escola Superior de Saúde da Universidade de Aveiro.

Dedico este trabalho à minha querida mãe.

O júri

Presidente	Professor Doutor Rui Costa Professor Coordenador da Escola Superior de Saúde da Universidade de Aveiro
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Palavras-chave Cuidados de saúde, Doença respiratória crónica, Empoderamento, Fibrose pulmonar, Qualidade de vida relacionada com a saúde.

Resumo

Introdução: As intervenções de autogestão (IA) visam capacitar as pessoas com doença crónica para realizarem uma gestão mais eficaz da sua saúde. Em pessoas com doenças respiratórias crónicas, como por exemplo a asma, as IA resultam em aumentos significativos da qualidade de vida relacionada com a saúde (QVRS). Contudo, a sua eficácia ainda não foi sistematizada em pessoas com doença pulmonar intersticial (DPI), o que limita a sua implementação nos cuidados de saúde.

Objetivos: Rever e sumariar os efeitos das IA na QVRS (medida principal), capacidade e *performance* funcional, fatores psicológicos e sociais, sintomas, exacerbações, utilização dos serviços de saúde, e sobrevida, em pessoas com DPI.

Métodos: O protocolo desta revisão sistemática foi registado (PROSPERO ID: CRD42022329199). Realizou-se uma pesquisa por estudos randomizados e controlados em 6 bases de dados, a 31 de maio de 2022 com atualizações mensais até fevereiro de 2023. Foram incluídos estudos que implementaram IA em pessoas com qualquer tipo de DPI. Dois revisores independentes implementaram a avaliação do risco de viés da Cochrane (RoB2) e o sistema de classificação de recomendações, avaliação, desenvolvimento e apreciação (GRADE). As diferenças entre grupos, tabelas de direção do efeito e meta-analises foram utilizadas para sintetizar os resultados.

Resultados: Quatro estudos que examinaram 217 participantes (81% homens, 71 anos, 91% fibrose pulmonar idiopática) foram incluídos. Verificou-se grande heterogeneidade na duração, conteúdo e estrutura das IA e pouco detalhe no reporte das intervenções de controlo. Não se verificaram diferenças estatisticamente significativas entre grupos na QVRS (diferença média padronizada: 0.08; 95% intervalo de confiança: -0.21 a 0.37; *p-value*: 0.58), nem nas medidas secundárias. A qualidade da evidência variou entre baixa e muito baixa.

Conclusões: Existe evidência baixa a muito baixa de que as IA não alterem significativamente a QVRS, *performance* funcional, fatores psicológicos e sociais, sintomas, e a utilização dos serviços de saúde em pessoas com DPI. Não foi encontrada evidência para os efeitos da IA na capacidade funcional, exacerbações e sobrevida. É necessário encontrar uma definição universal e consensual de IA de forma a implementar intervenções comparáveis e fornecer resultados mais confiáveis.

Keywords Healthcare, Chronic respiratory disease, Empowerment, Pulmonary fibrosis, Health-related quality of life.

Abstract

Background: Self-management interventions (SMIs) aim to empower people with chronic diseases to manage their health more effectively. In people with chronic respiratory diseases, such as asthma, SMIs significantly improve health-related quality of life (HRQoL). However, their effectiveness has not yet been systematized in people with interstitial lung disease (ILD), which limits their implementation in healthcare.

Objectives: To review and summarize the effects of SMIs on HRQoL (primary outcome), functional capacity and performance, psychological and social factors, symptoms, exacerbations, healthcare utilization, and survival, in people with ILD.

Methods: The protocol of this systematic review has been registered (PROSPERO ID: CRD42022329199). A search was performed for randomized controlled studies in 6 databases, on May 31, 2022, with monthly updates until February 2023. Studies implementing SMIs in people with any type of ILD were included. Two independent reviewers implemented the Cochrane tool for risk of bias assessment (RoB2) and the grading of recommendations, assessment, development, and evaluations (GRADE) system. Between groups differences, effect direction plots, and meta-analysis were used to summarize the results.

Results: Four studies that examined 217 participants (81% men, 71 years old, 91% idiopathic pulmonary fibrosis) were included. There was great heterogeneity in the duration, content, and structure of SMIs, and little detail in the reporting of control interventions. There were no statistically significant between-groups differences in HRQoL (standardized mean difference: 0.08; 95% confidence interval: -0.21 to 0.37; p-value: 0.58) nor in the secondary measures. The quality of evidence ranged from low to very low.

Conclusions: There is low to very low evidence that SMIs do not significantly change HRQoL, functional performance, psychological and social factors, symptoms, and healthcare utilization, in people with ILD. No evidence for the effects of SMIs on functional capacity, exacerbations, and survival was found. It is necessary to find a universal and consensual definition of SMIs to implement comparable interventions and provide more reliable results.

Abbreviations and/or acronyms

ATAQ-IPF - A Tool to Assess Quality of Life in Idiopathic Pulmonary Fibrosis
CENTRAL - Cochrane Central Register of Controlled Trials
CI – Confidence Interval
COPD – Chronic Obstructive Pulmonary Disease
EMBASE - Excerpta Medica Database
EQ-5D-5L - 5-level EuroQol 5-Dimensional questionnaire
EQ-VAS - EuroQol-Visual Analog Scale
GRADE - Grading of Recommendations, Assessment, Development, and Evaluation
HADS - Hospital Anxiety and Depression Scale
HRQoL – Health-Related Quality of Life
ILD – Interstitial Lung Disease
K-BILD - King's Brief Interstitial Lung Disease health status questionnaire
MD – Mean Difference
MET – Metabolic Equivalent of Task
NR – Not Reported
P – P-value
PRISMA – Preferred Reporting Items for Systematic Reviews and Meta-Analyses
PROMIS-29 - Patient-Reported Outcome Measurement Information System-29
RCT – Randomized Controlled Trial
RoB – Risk of Bias
RR - Risk Ratio
SF-36 - 36-item Short Form
SMD – Standardized Mean Difference
SMI – Self-Management Intervention
SWiM - Synthesis Without Meta-analysis
UCSD-SoBQ - University of California at San Diego Shortness of Breath Questionnaire
VAS -Visual Analog Scale

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Introduction

Interstitial lung disease (ILD) encompasses a heterogeneous group of pulmonary disorders characterized by diffuse inflammation and/or fibrosis of the lung interstitium, which result in a restrictive pulmonary disorder with gas exchange impairment[1]. Consequently, people with ILD may experience significant symptom burden (e.g., dyspnea, fatigue, and anxiety), functional impairment (e.g., inability to complete basic daily tasks), decreased health-related quality of life (HRQoL), and financial and social difficulties (e.g., loss of income, social isolation, and social stigma)[2–4]. Nevertheless, several studies show that people with ILD want to actively manage their condition, highlighting their need for information on the disease (e.g., disease course) and how to manage (e.g., symptom management) their condition[3,5,6].

Self-management can be defined as "actions individuals and others take to mitigate the effects of a long-term condition and to maintain the best possible quality of life"[7]. It is a concept that involves empowerment of individuals to manage the bio-psycho-social aspects of their lives[8,9]. Over the years various stakeholders took interest in self-management, as it may improve population's health by balancing the demand and supply of health services while reducing healthcare costs[7,10,11]. This is especially important considering the current global economic and health services situation, in which health costs are rising, prevalence of chronic diseases is increasing, and health workforce is scarce[7,10,12,13].

Previous reviews on the effects of self-management interventions (SMIs) in people with chronic respiratory diseases, such as chronic obstructive pulmonary disease (COPD) and asthma, report significant improvements in HRQoL, while significantly reducing unplanned hospital admissions[7,14–20]. To our knowledge, there is currently no systematic review about the effects of SMIs in people diagnosed with ILD. Although people with ILD and people with other chronic respiratory diseases share some symptoms (e.g., dyspnea, fatigue, and cough), the lung physiology (i.e., restrictive pulmonary pattern), treatment (e.g., anti-fibrotic drugs), and disease course (i.e., in most cases the progression is fast and unpredictable), are unique in people with ILD[21,22]. Therefore, as the disease characteristics

are different, the content, structure, and effects of SMIs may also vary in this population[5,23,24].

This systematic review aims primarily to summarize the effects of SMIs on the HRQoL of people with ILD[25]. The secondary objectives are to explore SMIs' effects on functional capacity, functional performance, psychological and social outcomes, symptoms, exacerbations, healthcare utilization, and survival, and to summarize the content and structure of SMIs used with this population.

Methods

Registration and Protocol

This systematic review was conducted according to the Cochrane handbook for systematic reviews of interventions and reported following the preferred reporting items for systematic reviews and meta-analysis (PRISMA) guideline[26,27]. The filled PRISMA checklists can be found in appendix. The review protocol was registered in the international prospective register of systematic reviews network (no. CRD42022329199), and approved on the 10th of May 2022. An amendment to the protocol was conducted and registered (approved on the 12th of December 2022). Details can be found in appendix.

Eligibility Criteria

Population

Studies with adult participants (≥18 years of age) diagnosed with any type of ILD were included. Studies including participants with acute exacerbations of ILDs (i.e., an acute, clinically significant respiratory deterioration characterized by evidence of new widespread alveolar abnormality)[28] or up until 30 days of an exacerbation were excluded. Additionally, articles with people with various diseases were only included if more than 50%

of their sample was composed of people with ILD, or if data about participants with ILD was separately provided in the article or upon request by the reviewers.

Intervention

For the scope of this review, SMIs were included according to a previously published definition[29]:

> *"… self-management intervention is structured but personalised and often multi-component, with goals of motivating, engaging and supporting the patients to positively adapt their health behaviour(s) and develop skills to better manage their disease."*

Comparator

Studies comparing SMIs to usual care, no intervention (e.g., waitlist), or any other type of intervention were included. Studies adding SMIs to other types of interventions were only included if the other intervention was the same in both the experimental and control groups (e.g., exercise program plus SMI vs. the same exercise program). Studies comparing SMIs to other SMIs (e.g., virtual educational program for self-management vs. face-to-face educational program for self-management) were excluded.

Outcomes

Articles must have included at least one of the following outcomes:

- HRQoL (primary outcome), defined as "[…] patient reports of functioning and well-being in physical, mental, and social domains of life"[30].
- Functional capacity (secondary outcome), defined as "[…] one's maximum potential to perform those activities people do in the normal course of their lives to meet basic needs, fulfill usual roles, and maintain their health and well-being"[31].

- Functional performance (secondary outcome), defined as "[…] the physical, psychological, social, occupational, and spiritual activities that people actually do in the normal course of their lives to meet basic needs, fulfil usual roles, and maintain their health and well-being"[31].
- Psychological and social outcomes (secondary outcome), including self-efficacy, patient activation, coping, social interaction, or others assessing these domains.
- Symptoms burden (secondary outcome), defined as the prevalence, frequency, intensity, severity, or impact of symptoms on the individual[32].
- Exacerbations (secondary outcome), defined as "[…] an acute, clinically significant respiratory deterioration characterized by evidence of new widespread alveolar abnormality"[28].
- Healthcare utilization (secondary outcome), defined as "[…] the quantification or description of the use of services by persons for the purpose of preventing and curing health problems, promoting maintenance of health and well-being, or obtaining information about one's health status and prognosis"[33].
- Survival (secondary outcome), defined as the number of people who are still alive over a certain period of time[34].

Types of studies

This systematic review only included randomized controlled trials (RCTs) to provide a synthesis of the highest level of evidence currently published[35]. Qualitative studies, research protocols, thesis, dissertations, unpublished work, protocols, ongoing studies, conference papers, and abstracts were excluded. Only articles written in English, Portuguese, Spanish, Italian, or French were included.

Information Sources

Articles were searched from inception to May 31st, 2022, in the following databases: PubMed/MEDLINE, Excerpta Medica Database (EMBASE), Cochrane Central Register of Controlled Trials (CENTRAL), Scopus, Web of Science Core Collection, and PsycInfo

(Ovid). The search was complemented by monthly automatic updates active from the date of the initial search until February 2023. Additionally, the reference list of included studies and other relevant resources (e.g., literature and systematic reviews on the research topic) were manually screened to identify additional articles left out in the preliminary search[3–5,7,8,18,24,36–54].

Search Strategy

The search strategy (available in appendix) was developed using text words and controlled vocabulary words (e.g., medical subject headings terms) for the condition (i.e., ILD), and the intervention of interest (i.e., SMI). Synonyms, thesaurus, abbreviations, or terms related to both the condition and the intervention were also considered. Additionally, search strategies of systematic reviews that studied ILD or SMI were manually inspected to find relevant terms[3,18,55].

One reviewer (S.F.) drafted a search strategy model for PubMed/MEDLINE, which was examined, discussed, and approved by the whole team before adaptation to other databases. Furthermore, the search strategy for every database was validated by testing whether it could identify two relevant articles to be included, previously sought through google scholar[50,51]. No search restrictions or filters were applied for the reference search.

Selection Process

References were imported from the electronic databases to the Mendeley platform or directly extracted to research information systems files (i.e., .ris files). Then, the reference list files were uploaded to Rayyan (https://www.rayyan.ai/) for duplicate removal and screening[56]. Duplicate removal was first performed automatically on Rayyan, then one reviewer (S.F.) sought the remaining records for duplicates. Two reviewers (S.F. and A.B.) independently screened the titles and abstracts. Pilot testing was performed using the first 20 records to clarify any discrepancies. If articles did not meet the selection criteria, they were excluded. The same two reviewers (S.F. and A.B.) independently performed the full-text

screening process. If articles did not meet the selection criteria, they were excluded at this phase, and reasons for exclusion were recorded. Disagreements, emerging either during the 1st or 2nd screening phase were resolved by consensus or by consulting a third reviewer (A.O.).

Data Collection Process

One reviewer (S.F.) extracted data from the included articles to a standard table and two other reviewers (A.O. and A.M.) double-checked the extracted data. The extracted data included: (1) full title; (2) author(s) name(s); (3) year of publication, (4) country where the study has been carried out; (5) study design; (6) characteristics of participants with ILD (sample size, sex, age, pulmonary function data, and type of ILDs being studied), (7) characteristics of the experimental group and control group interventions (setting, content, frequency, duration of sessions and length of the intervention), (8) follow-up and data collection timepoints, (9) outcomes and outcome measures assessed, and (10) results for each outcome measure.

In case of missing or unclear data, the authors of the study were contacted for clarification. Additionally, if a reply was not gathered from the authors, WebPlotDigitizer (https://automeris.io/WebPlotDigitizer/) was used to extract data from figures and graphs.

Risk of Bias Assessment

Results of between-group differences related to the outcomes of interest (e.g., the between-group difference of the King's brief ILD health status questionnaire [K-BILD]) were assessed for risk of bias (RoB) using the 2nd version of the Cochrane risk-of-bias tool for randomized trials[57]. This tool classifies the result into low RoB, some concerns, or high RoB by assessing five domains where potential bias may arise, namely in (1) randomization, (2) deviations from the intended interventions, (3) missing outcome data, (4) measurement of the outcome, and (5) selection of the reported result[58].

Two reviewers (S.F. and P.R.) independently assessed the results for the HRQoL outcome. In case of disagreements, the reviewers discussed until a consensus was reached. A third reviewer was consulted (A.M.) in case of persistent disagreements. The remaining results were then assessed by S.F. In case of missing or unclear data, the authors of the study were contacted for clarification.

Publication Bias Assessment

For outcomes where at least 10 studies reported results in sufficient detail, publication bias was planned to be assessed through funnel plot visual inspection and complemented by Egger's test (for continuous outcomes), and Harbord's and Peters' tests (for dichotomous outcomes)[35,59–62]. For both statistical tests, a p-value (P) <0.05 indicates the presence of small-study effects[59].

Certainty Assessment

Two reviewers (S.F. and A.O.) independently assessed the quality of evidence using the grading of recommendations, assessment, development, and evaluations (GRADE) system to assess the quality of evidence[63]. This system considers several factors including, but not limited to, study design, RoB, inconsistency, indirectness, imprecision, and publication bias of the included studies to categorize the quality of evidence into high, moderate, low, or very low quality[63]. The assessment was performed for the primary outcome (i.e., HRQoL) and the other secondary outcomes where a meta-analysis was performed. Individual studies were not assessed as the GRADE does not give clear guidance for the assessment of the quality of evidence in these situations. In case of disagreements, the reviewers discussed until a consensus was reached. If a consensus could not be reached a third reviewer was consulted (A.M.). In appendix, a detailed description of the assessment criteria can be examined.

Data Analysis and Synthesis

The level of inter-reviewer agreement for the screening process was calculated using Cohen's kappa statistic[64] and interpreted as: no agreement (0-0.20), minimal agreement (0.21-0.39), weak agreement (0.40-0.59), moderate agreement (0.60-0.79), strong agreement (0.80-0.90), and almost perfect agreement (>0.90)[64].

Synthesis of studies' characteristics was performed using ranges of data (e.g., range of ages across studies), and descriptive summary statistics, calculated from the extracted data (e.g., the total number of participants, and percentage of males across studies). Effects of interventions were synthesized reporting between-group differences. Studies were grouped according to the outcome measures they used.

Whenever pooling of study outcomes was possible (i.e., at least two studies provided sufficient data for that outcome), a meta-analysis was performed using RevMan[65]. The mathematical model used for the meta-analysis was decided upon examining the heterogeneity of the studies, assessed with the Cochran Q (or Chi-square) and Higgins I^2 statistics[35]. If the P of the Cochran Q test was <0.1, and the I^2 statistic was ≥50%, statistically significant heterogeneity was assumed, and a random-effect model was used[35]. If only one of the two conditions was verified, a visual inspection of the forest plot, to assess the overlap of the 95% confidence intervals (CIs) of the different included studies, was performed to make the final decision on the model selection[35]. If none of the conditions was met (i.e., the P of Cochran Q test >0.1, and I^2 statistic <50%), homogeneity was assumed, and a fixed-effect model was used[35].

For the primary outcome, primary and secondary analyses were performed, while for secondary outcomes only primary analyses were carried out. Primary analysis included the baseline and post-intervention outcome scores. Secondary analyses included the baseline and follow-up outcome scores. The follow-up was divided into short-term (≤6 months), medium-term (>6 to ≤12 months), and long-term (>12 months), when available[27]. Results of the meta-analysis were presented with the effect estimate, 95% CI, heterogeneity (i.e., I^2 statistic), and P[66].

In case performing a meta-analysis was possible and a study provided multiple eligible outcome measures for the same outcome, the following criteria were used to select one outcome measure: 1st validation for people with ILD, 2nd validation for people with respiratory diseases, and 3rd outcome measure complete scores. A sensitivity analysis was also performed by testing different combinations of outcome measures to ascertain the robustness of the findindgs[27].

Differences in the direction of the scale of the outcome measures included in the meta-analysis (e.g., in the K-BILD a higher score indicates better HRQoL, while in the tool to assess quality of life in idiopathic pulmonary fibrosis [ATAQ-IPF] a higher score indicates worst HRQoL), were dealt by multiplying the mean difference scores of one outcome measure by -1 to ensure that all the scales appointed in the same direction[27].

Effect sizes of risk ratios, risk differences, and number needed to treat were interpreted according to Hancock and Kent[67]. Standardized mean difference (SMD) was categorized into trivial (0.0-0.19), small (0.20-0.49), moderate (0.50-0.79), or large effect (≥0.80)[68]. The minimal clinically important difference (MCID) of each outcome measure was used to interpret clinical significance. The MCID used for each outcome were: (1) K-BILD total score (3.9 [range 2.9 to 4.9] points)[69], (2) K-BILD chest symptoms domain (9.8 [range 8.4 to 11.2] points)[69], (3) K-BILD breathlessness and activities domain (4.4 [range 4.0 to 5.1] points)[69], (4) K-BILD psychological domain (5.4 [range 4.1 to 6.1] points)[69], (5) visual analog scale (VAS) fatigue (14.5 [95% CI = 8 to 20] mm)[70], (6) VAS dyspnea (22.0 [95% CI = 12 to 35] mm)[70], (7) dyspnea-12 (2.97 [95% CI = 1.94 to 4.00] points)[71], (8) university of California at San Diego shortness of breath questionnaire (UCSD-SOBQ; 4.6 [range 1.1 to 8.4] points)[72], (9) 36-item short form (SF-36) mental component (7 [range 7 to 14] points)[73], (10) SF-36 physical component (5 [range 7 to 14] points)[73], (11) 5-level EuroQol 5-dimensional questionnaire (EQ-5D-5L; 0.095 points)[74], (12) EuroQol-VAS (EQ-VAS; 9.7 mm)[74], (13) hospital anxiety and depression scale (HADS) anxiety subscale (1.32 points)[75], (14) HADS depression subscale (1.40 points)[75], (15) steps per day (750 steps)[76], and (16) sedentary time per day (-25 min)[76]. There is/are currently no known MCID for ATAQ-IPF, life-space mobility, VAS cough, VAS general well-being, Manchester respiratory activities of daily living questionnaire, beck anxiety inventory, perceived stress

scale, patient-reported outcome measurement information system (PROMIS-29), beck depression inventory-II, global rating of change questionnaire, self-efficacy for managing chronic disease 6-item scale, duration of time above 3 metabolic equivalents of tasks (METs) per day, hospitalizations, and unscheduled healthcare visits in people with ILD or other chronic respiratory diseases. Additionally, statistical significance was determined with $P < 0.05$[68].

In case meta-analysis was not possible, the synthesis without meta-analysis (SWiM) guideline was used to resume the effect of the interventions[77]. An effect direction plot was used. This plot uses the direction of the effect, RoB, and sample size to display the effect of an intervention in studies that use different measures within an outcome[78]. A conclusion about the effect was made by counting the effect direction of individual studies and using the proportion of the effects[27]. If more than 50% of the studies reported a positive or a negative result, a positive or a negative effect was assumed, respectively. If 50% of the results reported a positive or a negative result; or if all studies reported mixed/conflicting findings, no assumption on the effect was made. A sign test was used to complement the assumptions[78].

Results

From the database search, 8'870 records were retrieved. After the removal of 3'288 duplicates, 5'547 records were removed in the first screening phase and further 31 records in the second screening phase. Therefore, 4 studies were included[50–53]. The reviewers' agreement was weak ($k = 0.47$) in the first screening phase and moderate in the second screening phase ($k = 0.77$). Reasons for records exclusion are reported in figure 1 and appendix. No extra articles fulfilling the inclusion criteria were found in other sources, nor in the updates from the databases.

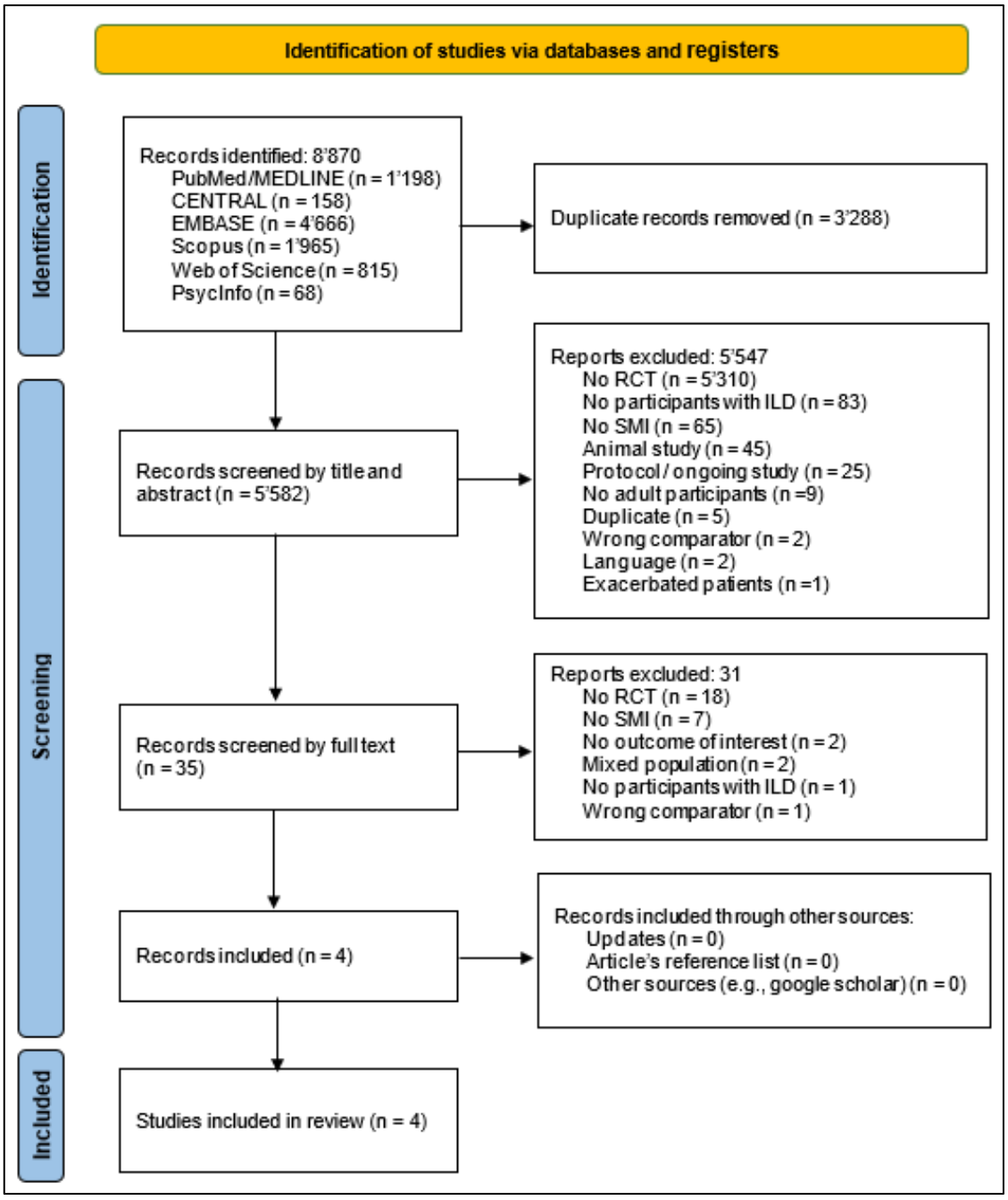


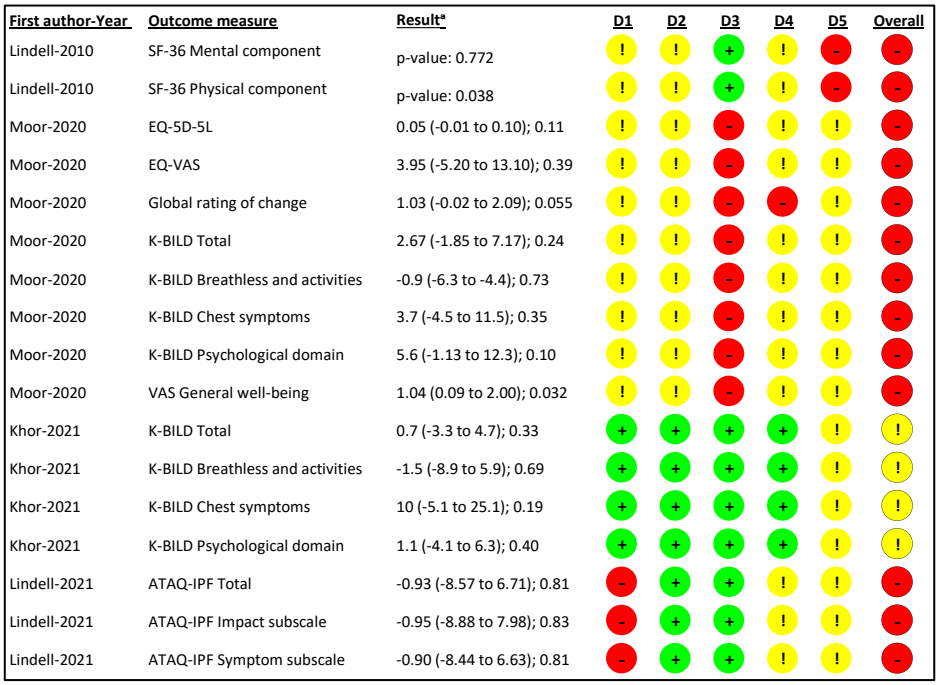
Figure 1 – Flow diagram of studies selected on self-management interventions in people with interstitial lung disease according to the preferred reporting items for systematic reviews and meta-analysis (PRISMA).

Abbreviations - **CENTRAL**: Cochrane Central Register of Controlled Trials; **EMBASE**: Excerpta Medica database; **ILD**: Interstitial Lung Disease; **SMI**: Self-Management Intervention; **RCT**: Randomized controlled trial.

RoB Assessment

The overall RoB for the HRQoL results was mostly high (3 out of 4 studies), with only one study [53] presenting some concerns. Primary reasons for high RoB were related to

the randomization process[51], missing outcome data[50], and selection of the reported result[52]. A summary of the assessments is displayed in figure 2. Detailed assessments are available in appendix.



First author-Year	Outcome measure	Result[a]	D1	D2	D3	D4	D5	Overall
Lindell-2010	SF-36 Mental component	p-value: 0.772	!	!	+	!	-	-
Lindell-2010	SF-36 Physical component	p-value: 0.038	!	!	+	!	-	-
Moor-2020	EQ-5D-5L	0.05 (-0.01 to 0.10); 0.11	!	!	-	!	!	-
Moor-2020	EQ-VAS	3.95 (-5.20 to 13.10); 0.39	!	!	-	!	!	-
Moor-2020	Global rating of change	1.03 (-0.02 to 2.09); 0.055	!	!	-	-	!	-
Moor-2020	K-BILD Total	2.67 (-1.85 to 7.17); 0.24	!	!	-	!	!	-
Moor-2020	K-BILD Breathless and activities	-0.9 (-6.3 to -4.4); 0.73	!	!	-	!	!	-
Moor-2020	K-BILD Chest symptoms	3.7 (-4.5 to 11.5); 0.35	!	!	-	!	!	-
Moor-2020	K-BILD Psychological domain	5.6 (-1.13 to 12.3); 0.10	!	!	-	!	!	-
Moor-2020	VAS General well-being	1.04 (0.09 to 2.00); 0.032	!	!	-	!	!	-
Khor-2021	K-BILD Total	0.7 (-3.3 to 4.7); 0.33	+	+	+	+	!	!
Khor-2021	K-BILD Breathless and activities	-1.5 (-8.9 to 5.9); 0.69	+	+	+	+	!	!
Khor-2021	K-BILD Chest symptoms	10 (-5.1 to 25.1); 0.19	+	+	+	+	!	!
Khor-2021	K-BILD Psychological domain	1.1 (-4.1 to 6.3); 0.40	+	+	+	+	!	!
Lindell-2021	ATAQ-IPF Total	-0.93 (-8.57 to 6.71); 0.81	-	+	+	!	!	-
Lindell-2021	ATAQ-IPF Impact subscale	-0.95 (-8.88 to 7.98); 0.83	-	+	+	!	!	-
Lindell-2021	ATAQ-IPF Symptom subscale	-0.90 (-8.44 to 6.63); 0.81	-	+	+	!	!	-

Figure 2 – Risk of bias of between-group differences regarding the effect of self-management interventions versus usual or standard care on health-related quality of life of people with interstitial lung disease according to the Cochrane risk-of-bias tool for randomized trials.

[a] Data is presented as mean (95% confidence interval); p-value unless otherwise stated.
Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **ATAQ-IPF**: A Tool to Assess Quality of Life in Idiopathic Pulmonary Fibrosis; **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **EQ-5D-5L**: 5-level EuroQol 5-Dimensional questionnaire; **EQ-VAS**: EuroQol-Visual Analog Scale; **K-BILD**: King's Brief Interstitial Lung Disease health status questionnaire; **SF-36**: 36-item short form; **VAS**: Visual Analog Scale**.**

RoB assessment for the secondary outcomes showed high RoB[50–52] and some concerns[50,53]. Summaries of these assessments are displayed in appendix.

Characteristics of Included Studies

The four included studies were published between 2010 and 2021 and were conducted in the Netherlands (n =1)[50], United States of America (n = 2)[51,52], and Australia (n = 1)[53]. All of them used a parallel group trial design.

Overall, 217 people with ILD were examined (with sample sizes varying from 21 to 90), most were men (n = 176; 81%) and presented mean ages from 66 to 72 years old. Participants had mean forced vital capacity from 69 to 80% of predicted and mean diffusing capacity for carbon monoxide from 42 to 50% of predicted. Idiopathic pulmonary fibrosis was the most predominant type of ILD (n = 197; 91%). Characteristics of the participants included in each study can be examined in table 1.

Table 1 – Study and sample characteristics of studies on self-management interventions in people with interstitial lung disease (n = 4).

First author Year Country	Study Design	Total Sample Characteristics			EG Sample Characteristics			CG Sample Characteristics		
		Number (% male) Age (years)[a]	Lung function (FVC % predicted and DLCO % predicted)[a]	ILD subtype (n, %)	Number (% male) Age (years)[a]	Lung function (FVC % predicted and DLCO % predicted)[a]	ILD subtype (n, %)	Number (% male) Age (years)[a]	Lung function (FVC % predicted and DLCO % predicted)[a]	ILD subtype (n, %)
Lindell, O. K. 2010 U.S.A.	Parallel RCT	n = 21 (76.2% ♂) 66.2 (10.9) years	FVC % predicted NR DLCO % predicted NR	IPF (21, 100%)	n = 10 (33.3% ♂) 65.2 (10.3) years	FVC % predicted NR DLCO % predicted NR	IPF (10, 100%)	n = 11 (42.9% ♂) 67.1 (11.9) years	FVC % predicted NR DLCO % predicted NR	IPF (11, 100%)
Moor, C. C. 2020 Netherlands	Parallel RCT	n = 90 (91% ♂) 71 (6.9) years	FVC % predicted 80.1 (17) DLCO % predicted 48.2 (13.5)	IPF (90, 100%)	n = 46 (85% ♂) 70 [53–83] years	FVC % predicted 82 (17.7) DLCO % predicted 48 (13.8)	IPF (46, 100%)	n = 44 (98% ♂) 72 [58–84] years	FVC % predicted 78 (16.0) DLCO % predicted 49 (13.0)	IPF (44, 100%)
Khor, Y. H. 2021 Australia	Parallel RCT	n = 30 (53% ♂) 72.7 years	FVC % predicted mean: 72.9 DLCO % predicted mean: 41.9	IPF (10, 33%) CTD- ILD (9, 30%) FHP (4, 13%) Unclassifiable ILD (3, 10%) NSIP (2, 7%) Asbestosis (1, 3%) Drug-induced ILD (1, 3%)	n = 15 (47% ♂) 73.7 (10.5) years	FVC % predicted 77.6 (18.0) DLCO % predicted 42.1 (11.5)	CTD-ILD (6, 40%) IPF (5, 33%) Drug-induced ILD (1, 7%) FHP (1, 7%) NSIP (1, 7%) Unclassifiable ILD (1, 7%)	n = 15 (60% ♂) 71.7 (7.3) years	FVC % predicted 68.2 (15.3) DLCO % predicted 41.7 (12.2)	IPF (5, 33%) CTD-ILD (3, 20%) FHP (3, 20%) Unclassifiable ILD (2, 13%) Asbestosis (1, 7%) NSIP (1, 7%)
Lindell, O. K. 2021 U.S.A.	Parallel RCT	n = 76 (81.6% ♂) 71.01 years	FVC % predicted 69.5 (16.5) DLCO % predicted 49.7 (18.2)	IPF (76, 100%)	n = 50 (80% ♂) 70.3 (5.3) years	FVC % predicted 68.7 (19.1) DLCO % predicted 46.3 (19.1)	IPF (50, 100%)	n = 26 (85% ♂) 72.3 (6.3) years	FVC % predicted 71.3 (19.6) DLCO % predicted 51.7 (20.4)	IPF (26, 100%)

[a] Data is presented as mean (standard deviation) or median [interquartile range] unless otherwise stated.

Abbreviations - ♂: male; **CTD-ILD**: Connective Tissues Disease-related Interstitial Lung Disease; **DLCO**: Diffusing Capacity of Carbon Monoxide; **FHP**: Fibrotic hypersensitivity pneumonitis; **FVC**: Forced Vital Capacity; **ILD**: Interstitial Lung Disease; **IPF**: Idiopathic Pulmonary Fibrosis; **NR**: Not Reported; **NSIP**: Non-Specific Interstitial Pneumonia; **RCT**: Randomized Controlled Trial; **U.S.A.**: United States of America.

Experimental interventions included a home monitoring program[50], two educational programs[51,52], and a handheld fan intervention for symptom control[53]. Control interventions included standard or usual care (n = 4)[50–53]. Interventions were carried out in the outpatient (n = 3)[50–52], home (n = 2)[50,51], and daily life context (n = 1)[53] settings. The length of interventions varied from 2 to 32 weeks, and the frequency of sessions ranged from daily to 3 visits in a 6 to 8-month period. The session's length was reported in a single study, approximately 120 minutes[52]. Characteristics of the interventions of each study are displayed in table 2.

Table 2 – Interventions and outcomes characteristics of studies on self-management interventions in people with interstitial lung disease (n = 4).

First author Year Country	EG Intervention		CG Intervention		Follow-up	Data Collection Timepoint	Outcome (outcome measure)
Lindell, O. K. 2010 U.S.A.	**Setting**	Outpatient (clinic)	**Setting**	Outpatient (clinic)	No follow-up	Baseline; 6 weeks	• HRQoL (SF-36) • Psychological and social outcomes (BAI; BDI-II; PSS) • Symptoms (UCSD-SoBQ)
	Content	PRISIM program (education group sessions to inform about IPF, symptom management, energy conservation, oxygen therapy, and exercise; discuss CBT, depression, and stress; plan for terminal illness, communicate with clinicians, cope, and plan to one's affairs) + book "Feeling Good: The New Mood Therapy".	**Content**	Usual care (visits by clinical care team members; available phone support and monthly support group; psychologic counseling was available) + book "Feeling Good: The New Mood Therapy".			
	Frequency	Weekly	**Frequency**	Monthly			
	Duration of sessions (minutes)	120	**Duration of sessions (minutes)**	NR			
	Length of intervention (weeks)	6	**Length of interventio n (weeks)**	6			
Moor, C. C. 2020 Netherlands	**Setting**	Home and outpatient (clinic)	**Setting**	Home and outpatient (clinic)	No follow-up	Baseline; 12 weeks; 24 weeks	• HRQoL (K-BILD; EQ-5D-5L; EQ-VAS; GRC; VAS General well-being) • Psychological and social outcomes (HADS) • Symptoms (VAS Dyspnea; VAS Fatigue; VAS Cough) • Healthcare utilization (Number of hospitalizations; Number of extra healthcare visits)
	Content	Home monitoring program (assessment of lung function, PROMs, symptoms and side effects with medication, information about IPF, medication coach, and eConsultations on a tablet) + standard care (clinic visits with pulmonary function testing, PROMs)	**Content**	Standard care (clinic visits with pulmonary function testing, PROMs).			
	Frequency	Daily	**Frequency**	Monthly			
	Duration of sessions (minutes)	NA	**Duration of sessions (minutes)**	NA			

	Length of intervention (weeks)	24	**Length of intervention (weeks)**	NA			
Khor, Y. H. 2021 Australia	**Setting**	Participant's daily life	**Setting**	NA	No follow-up	Baseline; 2 weeks	• HRQoL (K-BILD) • Functional performance (MRADLQ; steps per day; total energy expenditure; total METs; duration of sedentary time/day; duration of time >3 METs/day) • Psychological and social outcomes (SEMCD6) • Symptoms (Dyspnea-12)
	Content	Handheld fan (instructions on how to use a handheld fan for symptom control) + usual care	**Content**	Usual care			
	Frequency	Whenever needed	**Frequency**	NA			
	Duration of sessions (minutes)	NA	**Duration of sessions (minutes)**	NA			
	Length of intervention (weeks)	2	**Length of intervention (weeks)**	NA			
Lindell, O. K. 2021 U.S.A.	**Setting**	Home and outpatient (Clinic)	**Setting**	Home and outpatient (clinic)	No follow-up	Baseline; 24-32 weeks	• HRQoL (ATAQ-IPF) • Psychological and social outcomes (PSS; PROMIS-29) • Symptoms (PROMIS-29) • Healthcare utilization (Number of healthcare visits)
	Content	SUPPORT program (education about IPF, self-management, and introduction to advanced care planning in face-to-face, printed, and digital formats)	**Content**	Standard care plus printed patient education about IPF			
	Frequency	3 visits	**Frequency**	NR			
	Duration of sessions (minutes)	NR	**Duration of sessions (minutes)**	NR			
	Length of intervention (weeks)	24-32	**Length of intervention (weeks)**	NA			

Abbreviations - **ATAQ-IPF**: A Tool to Assess Quality of Life in Idiopathic Pulmonary Fibrosis; **BAI**: Beck Anxiety Inventory; **BDI-II**: Beck Depression Inventory-II; **CG**: Control Group**; CTB**: cognitive behavioral techniques; **EG**: Experimental Group; **EQ-5D-5L**: 5-level EuroQol 5-Dimensional questionnaire; **EQ-VAS**: EuroQol-Visual Analogue Scale; **GRC**: Global Rating of Change; **HADS**: Hospital Anxiety and Depression Scale; **HRQoL**: Health-related Quality of Life; **IPF**: Idiopathic Pulmonary Fibrosis; **K-BILD**: King's Brief Interstitial Lung Disease health status questionnaire; **MET**: Metabolic Equivalent of Task; **MRADLQ**: Manchester Respiratory Activities of Daily Living Questionnaire; **NA**: Not Applicable; **NR**: Not Reported; **PRISIM**: Program to Reduce Idiopathic Pulmonary Fibrosis Symptoms and Improve Management; **PROM**: Patient-reported Outcome Measure; **PROMIS-29**: Patient Reported Outcome Measurement Information System; **PSS**: Perceived Stress Scale; **SEMCD6**: Self-efficacy for Managing Chronic Disease 6-item Scale; **SF-36**: 36-item short form; **UCSD-SoBQ**: University of California at San Diego Shortness of Breath Questionnaire; **SUPPORT**: Symptom management, Understanding the disease, Pulmonary rehabilitation, Palliative care, Oxygen therapy, Research participation, and Transplantation; **U.S.A.**: United States of America; **VAS**: Visual Analog Scale.

Effects of Interventions

None of the included studies measured the effect of SMIs on functional capacity, exacerbations, or survival outcomes. Additionally, none of the studies performed follow-up assessments.

HRQoL

HRQoL was assessed in all four studies[50–53] using the EQ-5D-5L[50], ATAQ-IPF[53], EQ-VAS[50], global rating of change questionnaire[50], K-BILD[50,53], SF-36[52], and the VAS[50].

Significant between-group differences in favor of the control group for the SF-36 physical component (MD = not reported [NR]; 95% CI = NR; P = 0.038)[52] and in favor of the experimental group for the VAS general well-being (MD = 1.0; 95% CI = 0.09 to 2.0; P = 0.032)[50] were reported.

Non-significant but clinically important changes were found in the K-BILD chest symptom domain (MD = 10 [95% CI = -5.1 to 25.1]; P = 0.19)[53] and in the K-BILD psychological domain (MD = 5.6 [95% CI = -1.1 to 12.3]; P = 0.10)[50].

Non-significant and not clinically important between-group differences were found in the 5Q-5D-5L (MD = 0.05; 95% CI = -0.01 to 0.10; P = 0.11)[50], EQ-VAS (MD = 3.95; 95% CI = -5.2 to 13.1; P = 0.39)[50], K-BILD total score (MD = 2.7; 95% CI = -1.9 to 7.8; P = 0.24 and MD = 0.7; 95% CI = -3.3 to 4.7; P = 0.33)[50,53], K-BILD chest symptoms domain (MD = 3.7 [95% CI = -4.5 to 11.5]; P = 0.35)[50], K-BILD breathlessness and activities domain (MD = -0.9 [95% CI = -6.3 to -4.4]; P = 0.73 and MD = -1.5 [95% CI = -8.9 to 5.9]; P = 0.69)[50,53], and in the K-BILD psychological domain (MD = 1.1 [95% CI = -4.1 to 6.3]; P = 0.40)[53]. Non-significant between-group differences in the global rating of change questionnaire (MD = 1.0; 95% CI = -0.02 to 2.1; P = 0.055)[50], ATAQ-IPF total score (MD = -0.93; 95% CI = -8.6 to 6.7; P = 0.81)[51], ATAQ-IPF subscales[51], and in the SF-36 mental component (MD = NR; 95% CI = NR; P = 0.77)[52] were found. Detailed results of each study for the HRQOL outcome are displayed in table 3.

Table 3 – Results of self-management interventions versus usual or standard care on health-related quality of life of people with interstitial lung disease (n = 4).

First author Year Country	Outcome measure	Baseline data[a]	Post intervention data[a]	Within-group difference[a]	Between-group difference[b]
Lindell, O. K. 2010 U.S.A.	SF-36 Mental component↑	NR	EG: 55.98 (2.7)	NR	p-value: 0.77
		NR	CG: 55.6 (2.7)	NR	
	SF-36 Physical component↑	EG mean: 40.1	EG: 31.0 (4.6)	NR	p-value: 0.038+
		CG mean: 34.3	CG: 36.0 (4.6)	NR	
Moor, C. C. 2020 Netherlands	EQ-5D-5L ↑	EG: 0.77 (0.17)	NR	EG: 0.02 (0.02)	0.05 (-0.01 to 0.10); 0.11
		CG: 0.77 (0.17)	NR	CG: -0.03 (0.17)	
	EQ-VAS↑	EG: 63.1 (24.9)	NR	EG: -0.89 (3.6)	3.95 (-5.2 to 13.1); 0.39
		CG: 64.4 (21.9)	NR	CG: -4.8 (2.8)	
	Global rating of change↑	NR	NR	EG: 0.34 (0.4)	1.0 (-0.02 to 2.1); 0.055
		NR	NR	CG: -0.70 (0.4)	
	K-BILD Total↑	EG: 57.2 (10.9)	NR	EG: 2.7 (9.5)	2.7 (-1.9 to 7.8); 0.24
		CG: 56.2 (7.7)	NR	CG: 0.03 (10.4)	
	K-BILD Breathless and activities↑	EG: 48.8 (19.3)	NR	EG: -1.8 (10.7)	-0.9 (-6.3 to -4.4); 0.73
		CG: 41.3 (15)	NR	CG: -0.93 (12.8)	
	K-BILD Chest symptoms↑	EG: 74.3 (18.8)	NR	EG: 1.58 (13.3)	3.7 (-4.5 to 11.5); 0.35
		CG: 73 (18.9)	NR	CG: -2.1 (20.1)	
	K-BILD Psychological domain↑	EG: 54.4 (13.9)	NR	EG: 5.1 (15.8)	5.6 (-1.1 to 12.3); 0.10
		CG: 56.2 (11)	NR	CG: -0.48 (13.3)	
	VAS General well-being↑	EG: 5.6 (0.36)	NR	EG: 0.65 (0.36)	1.0 (0.09 to 2.0); 0.032+
		CG: 5.5 (0.31)	NR	CG: -0.39 (0.31)	
Khor, Y. H. 2021 Australia	K-BILD Total↑	EG: 48.0 (2.5)	EG: 50.3 (2.5)	EG: 2.3 (1.9)	0.7 (-3.3 to 4.7); 0.33
		CG: 52.3 (2.4)	CG: 54.0 (2.4)	CG: 1.7 (1.9)	
	K-BILD Breathlessness and Activities↑	EG: 29.2 (3.9)	EG: 32.2 (3.9)	EG: 3.0 (3.0)	-1.5 (-8.9 to 5.9); 0.69
		CG: 32.5 (3.7)	CG: 37.0 (3.7)	CG: 4.5 (2.9)	
	K-BILD Chest Symptoms↑	EG: 47.0 (5.2)	EG: 56.1 (5.4)	EG: 9.1 (4.1)	10 (-5.1 to 25.1); 0.19
		CG: 59.6 (5.0)	CG: 58.8 (5.2)	CG: -0.8 (3.95)	

	K-BILD Psychological domain↑	EG: 51.7 (4.2)	EG: 53.4 (4.2)	EG: 1.7 (3.25)	1.1 (-4.1 to 6.3); 0.40
		CG: 56.9 (4.1)	CG: 57.5 (4.1)	CG: 0.6 (3.18)	
Lindell, O. K. 2021 U.S.A.	ATAQ-IPF Total↓	EG: 47.1 (16.8)	EG: 46.9 (16.5)	EG: -0.2 (12.9)	-0.93 (-8.6 to 6.7); 0.81
		CG: 45.7 (17.5)	CG: 42.1 (16.4)	CG: -3.6 (13.17)	
	ATAQ-IPF Impact subscale↓	EG: 47.5 (18.2)	EG: 47.1 (18.0)	EG: -0.4 (14.02)	-0.95 (-8.9 to 7.98); 0.83
		CG: 46.7 (18.9)	CG: 43.2 (18.4)	CG: -3.5 (14.45)	
	ATAQ-IPF Symptom subscale↓	EG: 46.7 (16.2)	EG: 46.8 (15.6)	EG: 0.1 (12.33)	-0.90 (-8.4 to 6.6); 0.81
		CG: 44.7 (16.7)	CG: 40.9 (15.9)	CG: -3.8 (12.6)	

[a] Data is presented as mean (standard deviation) or median [interquartile range] unless otherwise stated.
[b] Data is presented as mean (95% confidence interval); p-value unless otherwise stated.
[+] p-value < 0.05
↑ A higher score indicates better HRQoL, or well-being.
↓ A lower score indicates better HRQoL.
Abbreviations - **ATAQ-IPF**: A Tool to Assess Quality of Life in Idiopathic Pulmonary Fibrosis; **CG**: Control Group; **EG**: Experimental Group; **EQ-5D-5L**: 5-level EuroQol 5-Dimensional questionnaire; **EQ-VAS**: EuroQol-Visual Analog Scale; **K-BILD**: King's Brief Interstitial Lung Disease health status questionnaire; **NR**: Not Reported**; SF-36**: 36-item short form; **U.S.A.**: United States of America; **VAS**: Visual Analog Scale**.**

In the direction plot (figure 3), for the HRQoL outcome, two studies reported a positive effect towards the EG[50,53], one study reported negative[51] and one study conflicting/mixed effects[52] (P of sign test = 1.0).

First author, Year, Country	Study Design	HRQoL	Psychological and social outcome						Symptom					Healthcare utilization
			Functional performance	Anxiety	Depression	Stress	Self-efficacy	Social outcome	Dyspneia	Cough	Fatigue	Pain	Sleep disturbance	
Lindell, O. K. 2010 U.S.A.	Parallel RCT	◄►		▼										
Moor, C. C. 2020 Netherlands	Parallel RCT	▲		▲	▲				▼	▼	▼			◄►
Khor, Y. H. 2021 Australia	Parallel RCT	▲	▲				▲		▲					
Lindell, O. K. 2021 U.S.A.	Parallel RCT	▼		▼	▼	▼		▲			▼	▲	▼	◄►

Figure 3 – Effect direction plot of self-management interventions versus usual or standard care in people with interstitial lung disease.

Effect direction: upward arrow ▲= positive health impact, downward arrow ▼= negative health impact, sideways arrow ◄►= no change/mixed effects/conflicting findings.
Sample size: Final sample size (individuals) in the intervention group: large arrow ▲ >300; medium arrow ▲ 50-300; small arrow ▲ <50.
Study quality: denoted by cell color: green = low risk of bias; orange = some concerns; red = high risk of bias.
Abbreviations – **HRQoL**: Health-Related Quality of Life; **RCT**: Randomized Controlled Trial.

Meta-analysis included three studies[50,51,53] that used the K-BILD and ATAQ-IPF questionnaires. A trivial, non-significant effect (SMD = 0.08; 95% CI = -0.21 to 0.37; I^2 = 36%; P = 0.58) was found (figure 4). Sensitivity analysis also showed non-significant results (P from 0.30 to 0.39), with trivial to large (SMD from 0.15 to 1.03) effects. Details of the sensitivity analysis with the forest plots can be found in appendix.

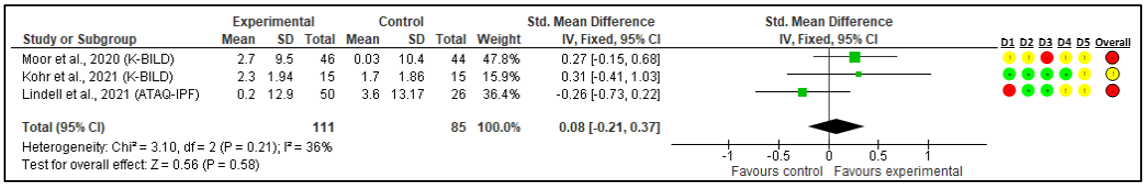


Figure 4 – Meta-analysis on the effect of self-management interventions versus usual or standard care on health-related quality of life of people with interstitial lung disease (n = 3).

Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **ATAQ-IPF**: A Tool to Assess Quality of Life in Idiopathic Pulmonary Fibrosis; **CI**: Confidence Interval; **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **IV**: Inverse-Variance; **K-BILD**: King's Brief Interstitial Lung Disease health status questionnaire; **P**: P-value; **SD**: Standard Deviation.

Functional Performance

Functional performance was assessed in one study[53] using the life-space mobility, the Manchester respiratory activities of daily living questionnaire, and an activity monitor.

Non-significant but clinically important between-group difference was found in the duration of sedentary time per day (MD = 66; 95% CI = -30 to 162; P = 0.18)[53]. Non-significant and not clinically important between-group difference was found in the steps per day (MD = 74; 95% CI = -807 to 956; P = 0.87)[53]. Additionally, non-significant between-group differences in the life-space mobility (MD = 2.4; 95% CI = -10.4 to 15.2; P = 0.72), Manchester respiratory activities of daily living questionnaire (MD = -2.5; 95% CI = -4.8 to 0.3; P = 0.08), duration of time above 3 METs per day (MD = 26; 95% CI = -16.1 to 68.3; P = 0.23), total energy expenditure (MD = 104; 95% CI = -197 to 404; P = 0.50) and total METs (MD = 0.003; 95% CI = -0.04 to 0.44; P = 0.88)[53] were found. Detailed results of each study can be found in table 4.

Table 4 – Results of self-management interventions versus usual or standard care on functional performance of people with interstitial lung disease (n = 1).

First author Year Country	Outcome measure	Baseline data[a]	Post intervention data[a]	Within-group difference[a]	Between-group difference[b]
Khor, Y. H. 2021 Australia	Duration of sedentary time per day (mins)↓	EG: 1129 (38)	EG: 1139 (36)	EG: 10.0 (28.7)	66 (-30 to 162); 0.18
		CG: 1205 (32)	CG: 1149 (31)	CG: -56.0 (24.4)	
	Duration of time above 3 METs per day (mins)↑	EG: 221 (27)	EG: 243 (27)	EG: 22.0 (20.9)	26 (-16.1 to 68.3); 0.23
		CG: 187 (23)	CG: 183 (23)	CG: -4.0 (17.8)	
	Life-space mobility↑	EG: 58.6 (6.3)	EG: 58.0 (6.4)	EG: -0.6 (4.9)	2.4 (-10.4 to 15.2); 0.72
		CG: 66.6 (6.1)	CG: 63.7 (6.1)	CG: -2.9 (4.7)	
	MRADLQ↑	EG: 14.4 (0.9)	EG: 14.6 (1.0)	EG: 0.2 (0.7)	-2.5 (-4.8 to 0.3); 0.08
		CG: 15.1 (0.9)	CG: 16.9 (0.9)	CG: 1.8 (0.69)	
	Steps per day↑	EG: 3423 (540)	EG: 3620 (527)	EG: 197.0 (413.4)	74 (-807 to 956); 0.87
		CG: 3082 (453)	CG: 3206 (488)	CG: 124.0 (365.9)	
	Total energy expenditure (kCal/day)↑	EG: 8269 (343)	EG: 8383 (340)	EG: 114.0 (264.5)	104 (-197 to 404); 0.50
		CG: 8470 (288)	CG: 8481 (287)	CG: 11.0 (222.7)	
	Total METs↑	EG: 1.1 (0.04)	EG: 1.12 (0.04)	EG: 0.02 (0.03)	0.003 (-0.04 to 0.44); 0.88
		CG: 1.1 (0.03)	CG: 1.1 (0.03)	CG: 0.01 (0.02)	

[a] Data is presented as mean (standard deviation) or median [interquartile range] unless otherwise stated.

[b] Data is presented as mean (95% confidence interval); p-value unless otherwise stated.

↑ A higher score indicates better performance.

↓ A lower score indicates better performance.

Abbreviations - **CG**: Control Group; **EG**: Experimental Group; **MET**: Metabolic Equivalent of Task; **MRADLQ**: Manchester Respiratory Activities of Daily Living Questionnaire.

In the direction plot (figure 3), a positive effect towards the EG[53] was found. Meta-analysis was not possible to be conducted for this outcome.

Psychological and Social Outcomes

Psychological outcomes explored in the included studies were anxiety[50–52], depression[50–52], stress[51,52], and self-efficacy[53], while satisfaction with social roles[51] was explored as a social outcome. Results related to the psychological and social outcomes are displayed in table 5.

Table 5 – Results of self-management interventions versus usual or standard care on psychological and social outcomes of people with interstitial lung disease (n = 4).

First author Year Country	Outcome measure	Baseline data[a]	Post intervention data[a]	Within-group difference[a]	Between-group difference[b]
Lindell, O. K. 2010 U.S.A.	Beck anxiety inventory↓	Mean EG: 7.9	EG: 15.13 (6.9)	NR	p-value: 0.077
		Mean CG: 17.1	CG: 8.5 (6.95)	NR	
	Beck depression inventory-II↓	NR	EG: 9.7 (4.3)	NR	p-value: 0.89
		NR	CG: 9.4 (4.4)	NR	
	Perceived stress scale↓	NR	EG: 19.3 (3.6)	NR	p-value: 0.53
		NR	CG: 18.2 (3.7)	NR	
Moor, C. C. 2020 Netherlands	HADS Anxiety↓	EG: 4.7 (2.5)	NR	EG: 0.13 (0.35)	-0.05 (-1.1 to 0.99); 0.93
		CG: 4.6 (2.2)	NR	CG: 0.18 (0.38)	
	HADS Depression↓	EG: 3.4 (3.2)	NR	EG: 0.34 (0.43)	-0.40 (-1.6 to 0.81); 0.51
		CG: 3.6 (3.6)	NR	CG: 0.74 (0.43)	
Khor, Y. H. 2021 Australia	SEMCD6↑	EG: 5.4 (0.6)	EG: 5.7 (0.7)	EG: 0.3 (0.51)	0.07 (-1.9 to 2.0); 0.94
		CG: 5.5 (0.6)	CG: 5.7 (0.6)	CG: 0.2 (0.46)	
Lindell, O. K. 2021 U.S.A	PROMIS-29 Anxiety/fear↓	EG: 52.9 (9.9)	EG: 51.5 (10.9)	EG: -1.4 (8.1)	0.01 (-5.6 to 5.6); 0.99
		CG: 52.8 (9.3)	CG: 50.5 (9.2)	CG: -2.3 (7.8)	
	PROMIS-29 Depression/sadness↓	EG: 50.9 (9.6)	EG: 50.6 (10.1)	EG: -0.3 (7.6)	-0.12 (-6.1 to 5.9); 0.97
		CG: 49.6 (11.3)	CG: 48.8 (8.7)	CG: -0.8 (8.1)	
	PROMIS-29 Satisfaction with Social Roles↑	EG: 45.3 (9.5)	EG: 46.2 (8.4)	EG: 0.9 (7.0)	3.7 (1.0 to 8.3); 0.12
		CG: 43.3 (8.5)	CG: 42.2 (8.9)	CG: -1.1 (6.8)	
	Perceived Stress Scale ↓	EG: 14.8 (6.1)	EG: 14.7 (7.1)	EG: -0.1 (5.2)	1.2 (-1.6 to 4.0); 0.39
		CG: 15.2 (5.8)	CG: 14.4 (3.9)	CG: -0.8 (4.1)	

[a] Data is presented as mean (standard deviation) or median [interquartile range] unless otherwise stated.

[b] Data is presented as mean (95% confidence interval); p-value unless otherwise stated.

↑ A higher score indicates better performance.

↓ A lower score indicates better performance.

Abbreviations - **CG**: Control Group; **EG**: Experimental Group; **HADS**: hospital anxiety and depression scale; **NR**: Not Reported; **PROMIS-29**: Patient Reported Outcome Measurement Information System-29; **SEMCD6**: Self-efficacy for Managing Chronic Disease 6-item Scale; **U.S.A.**: United States of America.

Anxiety was assessed in three studies[50–52], using the beck anxiety inventory[52], the HADS[50], and the PROMIS-29[51]. A non-significant and non-clinically important difference in the HADS anxiety (MD = -0.05; 95% CI = -1.1 to 0.99; P = 0.93)[50] was found. Non-significant differences in the beck anxiety inventory (MD = NR; 95% CI = NR; P = 0.077)[52] and in the PROMIS-29 anxiety/fear (MD = 0.01; 95% CI = -5.6 to 5.6; P = 0.99)[51] were also found.

In the direction plot (figure 3), two studies reported a negative[51,52], and one study a positive effect towards the EG[50] (P of sign test = 1.0). Meta-analysis included two studies[50,51] that used the HADS and PROMIS-29 questionnaires. It revealed a trivial, and non-significant effect (SMD = 0.03; 95% CI = -0.28 to 0.34; I^2 = 0%; P = 0.86) (figure 5).

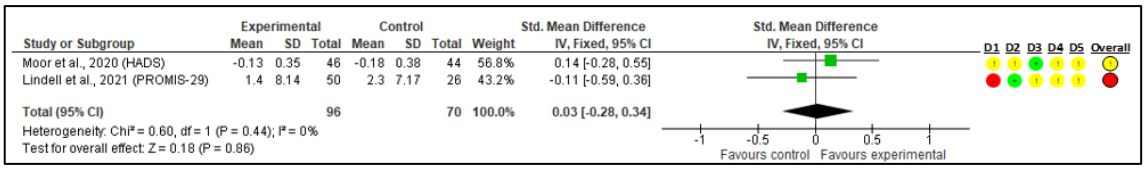


Figure 5 – Meta-analysis on the effect of self-management interventions versus usual or standard care on anxiety of people with interstitial lung disease (n = 2).

Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **CI**: Confidence Interval; **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **HADS**: Hospital Anxiety and Depression Scale; **IV**: Inverse-Variance; **P**: P-value; **PROMIS-29**: Patient Reported Outcome Measurement Information System-29; **SD**: Standard Deviation.

Depression was assessed in three studies[50–52], using the beck depression inventory-II[52], the HADS[50], and the PROMIS-29[51]. A non-significant and non-clinically important difference in the HADS depression (MD = -0.40; 95% CI = -1.6 to 0.81; P = 0.51)[50] was found. Additionally, a non-significant difference in the beck depression inventory-II (P = 0.89)[52], and in the PROMIS-29 depression/sadness (MD = -0.12; 95% CI = -6.1 to 5.9; P = 0.97)[51] was also found.

In the direction plot (figure 3), one study reported a negative[51] and another study reported a positive effect towards the EG[50] (P of sign test = 1.0). Meta-analysis included two studies[50,51], that used the HADS and PROMIS-29 questionnaires, and revealed a small, non-significant effect (SMD = 0.43; 95% CI = -0.53 to 1.4; I^2 = 89%; P = 0.38) (figure 6).

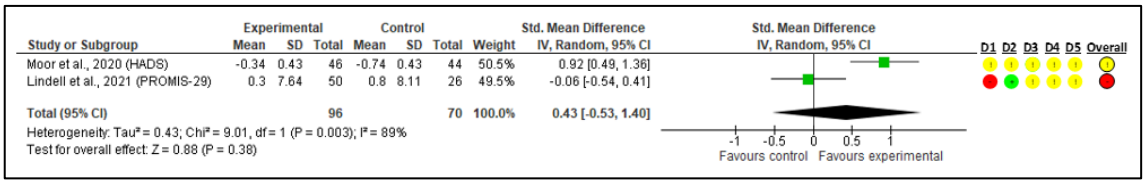


Figure 6 – Meta-analysis on the effect of self-management interventions versus usual or standard care on depression of people with interstitial lung disease (n = 2).

Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **CI**: Confidence Interval; **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **HADS**: Hospital Anxiety and Depression Scale; **IV**: Inverse-Variance; **P**: P-value; **PROMIS-29**: Patient Reported Outcome Measurement Information System-29; **SD**: Standard Deviation.

Stress was assessed in two studies using the perceived stress scale[51,52]. Non-significant between-group differences (MD = 1.2; 95% CI = -1.6 to 4.1; P = 0.39 and MD = NR; 95% CI = NR; P = 0.53)[51,52] were reported. In the direction plot (figure 3), one study reported a negative effect towards the EG[51]. Meta-analysis was not possible to be conducted for this outcome.

Self-efficacy was assessed in one study using the self-efficacy for managing chronic disease 6-item scale[53]. A non-significant difference (MD = 0.07; 95% CI = -1.9 to 2.0; P = 0.94) was reported with a positive effect being reported in the direction plot (figure 3). Meta-analysis was not possible to be conducted for this outcome.

Satisfaction with social roles was measured in one study, using the PROMIS-29[51]. A non-significant difference (MD = 3.7; 95% CI = 1.0 to 8.3; P = 0.12) was reported with a positive effect being observed in the direction plot (figure 3). Meta-analysis was not possible to be conducted for this outcome.

Symptoms

Dyspnea[50,52,53], fatigue[50,51], cough[50], pain[51], and sleep disturbance[51] were the symptoms explored in the included studies. Results related to the symptoms are displayed in table 6.

Table 6 – Results of self-management interventions versus usual or standard care on symptoms of people with interstitial lung disease (n = 4).

First author Year Country	Outcome measure	Baseline data[a]	Post intervention data[a]	Within-group difference[a]	Between-group difference[b]
Lindell, O. K. 2010 U.S.A.	UCSD-SoBQ↓	NR	EG: 49.5 (22.6)	NR	p-value: 0.97
		NR	CG: 49.9 (22.6)	NR	
Moor, C. C. 2020 Netherlands	VAS Cough↓	EG: 4.6 (0.45)	NR	EG: 0.51 (0.45)	0.82 (-0.52 to 2.8); 0.23
		CG: 4.7 (0.33)	NR	CG: -0.31 (0.50)	
	VAS Dyspnea↓	EG: 4.9 (0.38)	NR	EG: 0.41 (0.32)	0.63 (-0.23 to 1.5); 0.15
		CG: 5.8 (0.34)	NR	CG: -0.23 (0.30)	
	VAS Fatigue↓	EG: 4.8 (0.43)	NR	EG: 0.46 (0.40)	0.18 (-0.88 to 1.2); 0.74
		CG: 5.3 (0.38)	NR	CG: 0.28 (0.35)	
Khor, Y. H. 2021 Australia	Dyspnea-12↓	EG: 16.1 (2.2)	EG: 13.4 (2.3)	EG: -2.7 (1.8)	-2.2 (-6.4 to 1.9); 0.29
		CG: 13.3 (2.2)	CG: 12.8 (2.2)	CG: -0.5 (1.7)	
Lindell, O. K. 2021 U.S.A.	PROMIS-29 Fatigue↓	EG: 52.9 (7.9)	EG: 52.9 (8.2)	EG: 0.03 (6.2)	1.4 (-5.4 to 2.6); 0.49
		CG: 51.7 (6.6)	CG: 51.5 (8.8)	CG: -0.2 (6.3)	
	PROMIS-29 Pain interference↓	EG: 50.8 (9.8)	EG: 49.8 (9.9)	EG: -1.0 (7.6)	-1.4 (-5.8 to 3.1); 0.54
		CG: 50.1 (9.8)	CG: 49.9 (9.2)	CG: -0.2 (7.4)	
	PROMIS-29 Sleep disturbance↓	EG: 53.0 (4.7)	EG: 52.9 (3.8)	EG: -0.1 (3.4)	2.6 (-0.53 to 5.8); 0.10
		CG: 53.0 (2.6)	CG: 50.2 (5.9)	CG: -2.8 (4.5)	

[a] Data is presented as mean (standard deviation) or median [interquartile range] unless otherwise stated.

[b] Data is presented as mean (95% confidence interval); p-value unless otherwise stated.

↓ A lower score indicates less symptom burden.

Abbreviations - **CG**: Control Group; **EG**: Experimental Group; **NR**: Not Reported; **PROMIS-29**: Patient Reported Outcome Measurement Information System-29; **UCSD-SoBQ**: University of California at San Diego Shortness of Breath Questionnaire; **U.S.A.**: United States of America; **VAS**: Visual Analog Scale.

Dyspnea was assessed in three studies[50,52,53], using the UCSD-SoBQ[52], the VAS[50], and the Dyspnea-12 questionnaire[53]. A non-significant and non-clinically important between-group difference was found in the dyspnea-12 (MD = -2.2; 95% CI = -6.4 to 1.9; P = 0.29)[53] and in the VAS dyspnea (MD = 0.63; 95% CI = -0.23 to 1.5; P = 0.15)[50]. Additionally, a non-significant difference in the UCSD-SoBQ (MD = NR; 95% CI = NR; P = 0.97)[52] was found.

In the direction plot (figure 3), one study reported a negative effect[50], and another study reported a positive effect towards the EG[53] (P of sign test = 1.0). Meta-analysis included two studies[50,53], that used the VAS and the Dypnea-12. The result revealed a small non-significant effect (SMD = -0.43; 95% CI = -3.62 to 2.76; I^2 = 98%; P = 0.79) (figure 7).

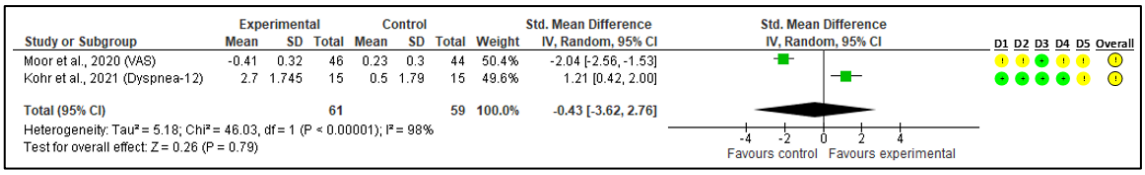


Figure 7 – Meta-analysis on the effect of self-management interventions versus usual or standard care on dyspnea of people with interstitial lung disease (n = 2).

Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **CI**: Confidence Interval; **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **IV**: Inverse-Variance; **P**: P-value; **SD**: Standard Deviation; **VAS**: Visual Analog Scale.

Fatigue was assessed in two studies[50,51], using the VAS[50] and the PROMIS-29[51]. A non-significant and non-clinically important difference was found in the VAS fatigue (MD = 0.18; 95% CI = -0.88 to 1.2; P = 0.74)[50] and a non-significant result was found in the PROMIS-29 fatigue (MD = 1.4; 95% CI = -5.4 to 2.6; P = 0.49)[51].

In the direction plot (figure 3), the two studies reported a negative effect towards the EG[50,51] (P of sign test = 0.5). Meta-analysis included two studies[50,51], that used the VAS and the PROMIS-29 questionnaire and revealed a small non-significant effect (SMD = -0.28; 95% CI = -0.60 to 0.03; I^2 = 46%; P = 0.08) (figure 8).

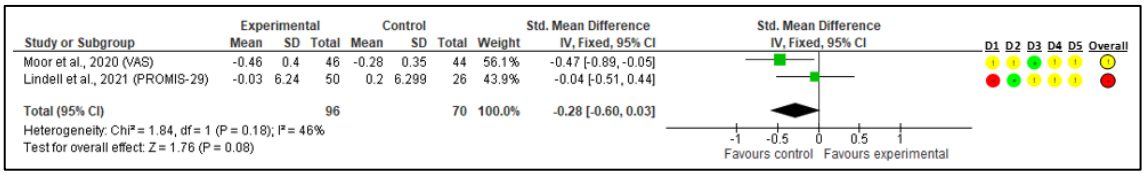


Figure 8 – Meta-analysis on the effect of self-management interventions versus usual or standard care on fatigue of people with interstitial lung disease (n = 2).

Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **CI**: Confidence Interval; **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **IV**: Inverse-Variance; **P**: P-value; **PROMIS-29**: Patient Reported Outcome Measurement Information System-29; **SD**: Standard Deviation; **VAS**: Visual Analog Scale.

Cough was assessed in one study using the VAS scale[50]. A non-significant result (MD = 0.82; 95% CI = -0.52 to 2.8; P = 0.23)[50] was reported with a negative effect being observed in the direction plot (figure 3). Meta-analysis was not possible to be conducted for this outcome.

Pain was assessed in one study using the PROMIS-29[51]. A non-significant result (MD = -1.4; 95% CI = -5.8 to 3.1; P = 0.54)[51] was reported with a positive effect being observed in the direction plot (figure 3). Meta-analysis was not possible to be conducted for this outcome.

Sleep disturbance was assessed in one study using the PROMIS-29[51]. A non-significant result (MD = 2.6; 95% CI = -0.53 to 5.8; P = 0.10)[51] was reported, with a negative effect being observed in the direction plot (figure 3). Meta-analysis was not possible to be conducted for this outcome.

Healthcare Utilization

Healthcare utilization was assessed in two studies[50,51], where hospitalizations[50] and healthcare visits[50,51] were measured. Results related to the healthcare utilization outcome are displayed in table 7.

Table 7 – Results of self-management interventions versus usual or standard care on healthcare utilization of people with interstitial lung disease (n = 2).

First author Year Country	Outcome measure	Patients that experienced the event	Number of events experienced[a]	Risk ratio[b]	Risk difference[c]	NNT (harm or benefit)
Moor, C. C. 2020 Netherlands	Extra visits	EG: 13	NR	1.2 (0.61 to 2.5); 0.55	5.5% (-12.4% to 23.5%)	18.0 (harm)
		CG: 10	NR			
	Hospitalizations	NR	EG: 6 in total	p-value: 0.27	NR	NR
		NR	CG: 4 in total			
Lindell, O. K. 2021 U.S.A.	Emergency visits	EG: 8	EG: 0.16 (0.37)	1.4 (0.40 to 4.8); 0.61	4.5% (-11.4% to 20.4%)	22.4 (harm)
		CG: 3	CG: 0.12 (0.33)			
	Inpatient visits	EG: 13	EG: 0.26 (0.69)	1.7 (0.61 to 4.7); 0.31	10.6% (-7.8% to 29.1%)	9.4 (harm)
		CG: 4	CG: 0.19 (0.49)			
	Outpatient visits	EG: 43	EG: 5 [1-9]	0.9 (0.796 to 1.1); 0.38	-6.3% (-7.7% to 20.4%)	15.9 (benefit)
		CG: 24	CG: 5 [3-7]			

[a] Data is presented as mean (standard deviation) or median [interquartile range] unless otherwise stated.
[b] Data is presented as risk ratio (95% confidence interval); p-value unless otherwise stated.
[c] Data is presented as risk (95% confidence interval) unless otherwise stated.
Abbreviations - **CG**: Control Group; **EG**: Experimental Group; **NNT**: Number Needed to Treat; **NR**: Not Reported; **U.S.A.**: United States of America.

Non-significant results in the risk for having hospitalizations (MD = NR; 95% CI = NR; P = 0.27)[50], extra healthcare visits (risk ratio [RR] = 1.2; 95% CI = 0.61 to 2.5; P = 0.55)[50], outpatient visits (RR = 0.93; 95% CI = 0.796 to 1.1; P = 0.38)[51], inpatient visits (RR = 1.7; 95% CI = 0.61 to 4.7; P = 0.31)[51], or emergency visits (RR = 1.4; 95% CI = 0.40 to 4.8; P = 0.61)[51] were observed.

In the direction plot, the two studies reported mixed/conflicting findings[50,51] (figure 3). Meta-analysis was performed for unplanned healthcare visits which were defined as extra or emergency visits. A non-significant result favoring the control group was observed (RR = 1.28; 95% CI = 0.69 to 2.39; I^2 = 0%; P = 0.43) (figure 9).

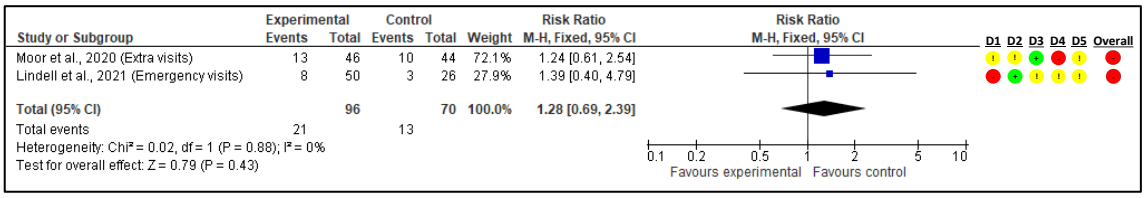


Figure 9 – Meta-analysis on the effect of self-management interventions versus usual or standard care on unplanned healthcare visits of people with interstitial lung disease (n = 2).

Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **CI**: Confidence Interval; **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **IV**: Inverse-Variance; **P**: P-value; **SD**: Standard Deviation.

Certainty Assessment (GRADE Assessment)

Quality of evidence was rated as low for HRQoL, anxiety, and fatigue and very low for depression, dyspnea, and unplanned healthcare visits. Primary reasons for lowering the evidence were related to RoB, inconsistency, and imprecision. A summary of the GRADE findings is in tables 8 and 9.

Table 8 – Grading of recommendations, assessment, development, and evaluation (GRADE) evidence profile regarding the effects of self-management interventions versus usual or standard care on health-related quality of life, anxiety, depression, dyspnea, and fatigue of people with interstitial lung disease.

Outcome	Quality assessment						Summary of findings			
Number of studies (Design)	Risk of bias	Inconsistency	Indirectness	Imprecision	Publication bias	Other considerations	Participants in the EG (N)	Participants in the CG (N)	Effect estimate*	Quality
Effect of SMI on HRQoL										
3 (parallel RCT)	Serious[a]	Not inconsistency	Not serious	Serious[e]	Likely[g]	None	111	85	0.08 (-0.21 to 0.37); 36%; 0.58	Low ⊕⊕□□[i]
Effect of SMI on psychological and social outcomes (anxiety)										
2 (parallel RCT)	Serious[b]	Not inconsistency	Not serious	Serious[e]	Likely[h]	None	96	70	0.03 (-0.28 to 0.34); 0%; 0.86	Low ⊕⊕□□[i]
Effect of SMI on psychological and social outcomes (depression)										
2 (parallel RCT)	Serious[b]	Serious[c]	Not serious	Very serious[f]	Likely[h]	None	96	70	0.43 (-0.53 to 1.40); 89%; 0.38	Very low ⊕□□□[j]
Effect of SMI on symptoms (dyspnea)										
2 (parallel RCT)	Not serious	Serious[d]	Not serious	Very serious[f]	Likely[h]	None	61	59	0.43 (-2.76 to 3.62); 98%; 0.79	Very low ⊕□□□[j]
Effect of SMI on symptoms (fatigue)										
2 (parallel RCT)	Serious[b]	Not inconsistency	Not serious	Serious[e]	Likely[h]	None	96	70	-0.28 (-0.60 to 0.03); 46%; 0.08	Low ⊕⊕□□[i]

Abbreviations - **EG**: Experimental Group; **CG**: Control Group; **RCT**: Randomized Controlled Trial.

* Data presented as standardized mean difference (95% confidence interval); I^2 statistic; p-value unless otherwise noted.

[a] Serious risk of bias because 67% of studies have high risk of bias (rate down one level).

[b] Serious risk of bias because 50% of studies have high risk of bias (rate down one level).

[c] Serious inconsistency because p-value of Q test < 0.00001 and I^2 = 98% (rate down one level).

[d] Serious inconsistency because p-value of Q test = 0.003 and I^2 = 89% (rate down one level).

[e] Serious imprecision due to not sufficiently large sample size (n < 400) (rate down one level).

[f] Very serious imprecision due to not sufficiently large sample size (n < 400) and very large confidence interval (i.e., interval limits are ≥ 0.5 of the effect estimate) (rate down two levels).

[g] Publication bias is likely to be present as 67% of the studies do not attain the calculated sample size for an adequately powered trial (rate down one level).

[h] Publication bias is likely to be present as 50% of the studies do not attain the calculated sample size for an adequately powered trial (rate down one level).

[i] Quality of evidence was rated down due to risk of bias and imprecision. Although publication bias is likely present, it was not accounted for the final assessment as it could not be rigorously assessed due to few included studies being included.

[j] Quality of evidence was rated down due to risk of bias, inconsistency, and imprecision. Although publication bias is likely present, it was not accounted for in the final assessment as it could not be rigorously assessed due to few included studies being included.

Table 9 – Grading of recommendations, assessment, development, and evaluation (GRADE) evidence profile regarding the effects of self-management interventions versus usual or standard care on unplanned healthcare visits of people with interstitial lung disease.

Outcome	Quality assessment						Summary of findings				
Number of studies (Design)	Risk of bias	Inconsistency	Indirectness	Imprecision	Publication bias	Other considerations	Participants in the EG (N)	Participants in the CG (N)	Relative risk*	Absolute risk**	Quality
Healthcare utilization (unplanned healthcare visits)											
2 (parallel RCT)	Very serious[a]	Not serious	Not serious	Very serious[b]	Likely[c]	None	96	70	1.40 (0.78 to 2.50); 0%; 0.26	0.08 (-0.05 to 0.21)	Very low ⊕□□□[d]

Abbreviations - **EG**: Experimental Group; **CG**: Control Group; **RCT**: Randomized Controlled Trial.

* Data expressed as risk ratio (95% confidence interval); I^2 statistic; p-value unless otherwise noted.

** Data expressed as risk reduction (95% confidence interval) unless otherwise noted.

[a] Very serious risk of bias because 100% of studies have high risk of bias (rate down two levels).

[b] Very serious imprecision due to few events (n = 34), and large confidence interval (i.e., interval limits are > 0.25 of the effect estimate) (rate down two levels).

[c] Publication bias is likely to be present as 50% of the studies do not attain the calculated sample size for an adequately powered trial (rate down one level).

[d] Quality of evidence was rated down due to risk of bias, and imprecision. Although publication bias is likely present, it was not accounted for in the final assessment as it could not be rigorously assessed due to few included studies being included.

Discussion

This systematic review and meta-analysis showed low to very low-quality evidence and non-significant effects of SMIs when compared to usual or standard care in the HRQoL, functional performance, psychological and social outcomes, symptoms, and healthcare utilization in people with ILD.

Our findings contrast with the significant improvements in HRQoL, anxiety, depression, and healthcare utilization previously reported in other chronic respiratory diseases (e.g., COPD and asthma)[14–17,20,23,79]. In this review, two to three studies were included in the meta-analysis, whilst systematic reviews examining the effect of SMIs vs. usual care included at least three times that number of RCTs in their meta-analysis[14–17,20,79], resulting in more consistent findings[80,81]. Furthermore, when reported, the intervention duration of the studies in those reviews was generally longer (i.e., most trials had interventions that lasted at least three months)[16,17,20,79], while half of our studies[52,53] included interventions of six or fewer weeks.

In a recent consensus study involving healthcare professionals and people living with the disease, the key contents of SMIs for people with pulmonary fibrosis were reported[6]. The interventions in this review included at least one of the essential components stated by the authors, yet they failed to show significant improvements. The consensus also highlighted that SMIs should include individualization, goal setting, and feedback[6]. In this review, the interventions were designed to meet the needs of people with ILD, however, goal setting and/or feedback were missing in all of them[51–53]. Additionally, although the control interventions in included studies were not considered as SMIs, they may have incorporated strategies that promoted participants' self-management, such as education on medication management and how to recognize an exacerbation, which may have decreased the effects of SMIs compared to usual or standard care[17,50]. This is important to consider particularly in two studies where we do not have a clear description of the content of the usual care[51,53].

The quality of evidence was low to very low in our findings, meaning that there is limited to little confidence in the effect estimate observed in the meta-analysis. Two Cochrane reviews on the effects of SMIs in people with COPD reported moderate to very

low quality of evidence in their findings, primarily due, as in this review, to high RoB, imprecision, and inconsistency[17,19]. Heterogeneity in the content, structure, and duration of interventions may have contributed to the inconsistency, especially considering that SMIs represent a concept, without a standardized, and consensual definition, therefore interventions will vary across studies, producing different results[9,17,23].

Implications for the Future

Previous studies[50,52,53] have shown that people with ILD perceived SMIs as beneficial for their health, nevertheless, implementation of these interventions into clinical practice cannot be suggested based on the quantitative results of the included studies. Given, the overall low quality of the available evidence, high-quality RCTs are recommended to strengthen our conclusions on the effects of SMIs in people with ILD. Additionally, future studies should assess the self-management abilities of participants at baseline (i.e., ascertain if the control and experimental group are comparable in terms of self-management ability), include their self-management abilities as an outcome (i.e., ascertain if SMIs are effective in modifying the self-management ability of participants), and confirm whether participants are performing SMIs outside the trial (i.e., ascertain if participants adopt self-management strategies outside the trial protocol which might affect results). Outcome measures such as the patient activation measure and the self-management ability scores could be used to assess the self-management abilities of participants[82,83].

Furthermore, the development of a clear, universal, and criteria-based definition of SMI and the pertaining minimal components is advised. The concept of SMI has been operationalized in several manners across studies which may lead authors to report different results about the effects SMIs and lead experimental trials to implement SMIs that are not effectively focused on promoting self-management behaviours[9,24,84]. For the scope of this review, a definition of SMI, provided in a consensus statement, for people with COPD, was used[29]. However, this definition was developed considering the characteristics and needs of people with COPD, which may not be valid for people with ILD.

Finally, future studies should implement longer interventions, perform follow-up assessments to investigate the short-, medium- and long-term effects of SMIs, should clearly

describe the control interventions, and explore the effects of these interventions on core patient-centered outcomes of people with ILD (e.g., functional status, and survival)[1,85].

Limitations

Results obtained from the included studies have limitations that need to be acknowledged. Firstly, our conclusions are based on a small number of studies, with heterogeneous interventions, high RoB, and low to very low-quality evidence, which limits the confidence and consistency of the findings. Secondly, results are based mostly on older men with idiopathic pulmonary fibrosis which limits the generalization of the results for all people with ILD. Although epidemiological studies show that people with ILD are predominantly men, there is evidence that sex/gender are potentially significant modifiers of disease course and response to treatment[86,87].

The review process has also some limitations. Firstly, only peer-reviewed publications included in databases were searched, while additional interventional studies may exist in the unpublished grey literature. Secondly, only RCTs were included. Although randomized trials provide the highest level of information, good-quality non-randomized studies might provide high-quality evidence, complementing the results obtained from the RCT[63]. Thirdly, the criteria used for the certainty assessment was based on reviewers' adaptation of the GRADE guidelines, as the guidelines do not provide clear guidance about the quality assessment of individual studies, the assessment of publication bias with few studies, or about the rating up of RCT. Fourthly, RoB assessment for the secondary outcome measures and data extraction was performed by one reviewer, due to limited time and resources.

Conclusions

There is low to very low-quality evidence that SMIs have no significant effect on the HRQoL, functional performance, psychological and social outcomes, symptoms, and healthcare utilization of people with ILD, when compared to usual or standard care. No evidence was found regarding the effects of SMIs on the functional capacity, exacerbations, and survival of people with ILD. Issues regarding to SMIs definition (e.g., necessary criteria

to be considered a SMI) and implementation (e.g., measure the self-management abilities of the participants) should be addressed in future studies to give more reliable results.

Funding

No funding was received for this project.

Conflict of Interest

The reviewers have no conflicts of interest to declare.

Appendices

In appendix, a Word file containing the PRISMA checklists, the amendment to the initial study protocol, the search strategies for each database sought in the selection process, the GRADE assessment criteria used in the certainty assessment, the table with the reasons for exclusion of each record in the full-text screening phase, the RoB assessment results for the secondary outcomes, and the forest plots regarding the sensitivity analysis for the HRQoL outcome, can be found. Additionally, an Excel file containing the detailed results of the RoB assessment can be examined.

Additional information

This work has been submitted for presentation to the European Respiratory Society International Congress 2023 (abstract number: 37353). A copy of the submitted abstract can be found in appendix.

References

1. Aronson KI, Swigris JJ, Bajwah S, et al. Patient-centered outcomes research in interstitial lung disease: An official american thoracic society research statement. *Am J Respir Crit Care Med*. 2021;204(2):E3-E23. doi:10.1164/rccm.202105-1193ST

2. Diamantopoulos A, Wright E, Vlahopoulou K, Cornic L, Schoof N, Maher TM. The Burden of Illness of Idiopathic Pulmonary Fibrosis: A Comprehensive Evidence Review. *Pharmacoeconomics*. 2018;36(7):779-807. doi:10.1007/s40273-018-0631-8

3. Lee JYT, Tikellis G, Corte TJ, et al. The supportive care needs of people living with pulmonary fibrosis and their caregivers: A systematic review. *European Respiratory Review*. 2020;29(190125):1-12. doi:10.1183/16000617.0125-2019

4. Kreuter M, Swigris J, Pittrow D, et al. Health related quality of life in patients with idiopathic pulmonary fibrosis in clinical practice: Insights-IPF registry. *Respir Res*. 2017;18(139):1-10. doi:10.1186/s12931-017-0621-y

5. Lee JYT, Tikellis G, Glaspole I, Khor YH, Symons K, Holland AE. Self-management for pulmonary fibrosis: Insights from people living with the disease and healthcare professionals. *Patient Educ Couns*. 2022;105(4):956-964. doi:10.1016/j.pec.2021.07.005

6. Lee JYT, Tikellis G, Khor YH, Holland AE. Developing a self-management package for pulmonary fibrosis: An international Delphi study. *ERJ Open Res*. Published online October 13, 2022:00349-02022. doi:10.1183/23120541.00349-2022

7. Taylor JS, Pinnock H. Supported self-management for respiratory conditions in primary care. *Primary Care Respiratory*. 2017;4(3):11-15.

8. Kelly C, Heslop-Marshall K, Jones S, Roberts NJ. Self-management in chronic lung disease: what is missing? *Breathe*. 2022;18(210179):1-4. doi:10.1183/20734735.0179-2021

9. van de Velde D, de Zutter F, Satink T, et al. Delineating the concept of self-management in chronic conditions: A concept analysis. *BMJ Open*. 2019;9(7). doi:10.1136/bmjopen-2018-027775

10. World Health Organization. *Health Workforce Requirements for Universal Health Coverage and the Sustainable Development Goals*. Vol 17. (Human Resources for Health Observer, ed.). World Health Organization; 2016. http://www.who.int/about/licensing/

11. Wijsenbeek MS, Moor CC, Johannson KA, et al. Home monitoring in interstitial lung diseases. *Lancet Respir Med*. Published online October 2022. doi:10.1016/s2213-2600(22)00228-4

12. Meghji J, Mortimer K, Agusti A, et al. Improving lung health in low-income and middle-income countries: from challenges to solutions. *The Lancet*. 2021;397(10277):928-940. doi:10.1016/S0140-6736(21)00458-X

13. Soriano JB, Kendrick PJ, Paulson KR, et al. Prevalence and attributable health burden of chronic respiratory diseases, 1990–2017: a systematic analysis for the Global Burden of Disease Study 2017. Published online 2020. doi:10.1016/S2213-2600(20)30105-3

14. Newham JJ, Presseau J, Heslop-Marshall K, et al. Features of self-management interventions for people with COPD associated with improved health-related quality of life and reduced emergency department visits: A systematic review and meta-analysis. *International Journal of COPD*. 2017;12:1705-1720. doi:10.2147/COPD.S133317

15. Panagioti M, Richardson G, Small N, et al. Self-management support interventions to reduce health care utilisation without compromising outcomes: a systematic review and meta-analysis. Published online 2014. doi:10.1186/1472-6963-14-356

16. Pinnock H, Parke HL, Panagioti M, et al. Systematic meta-review of supported self-management for asthma: A healthcare perspective. *BMC Med*. 2017;15(1). doi:10.1186/s12916-017-0823-7

17. Schrijver J, Lenferink A, Brusse-Keizer M, et al. Self-management for people with chronic obstructive pulmonary disease. *Cochrane Database of Systematic Reviews*. 2022;(1):1-189. doi:10.1002/14651858.CD002990.pub4

18. Song X, Hallensleben C, Zhang W, et al. Blended Self-management interventions to reduce disease burden in patients with chronic obstructive pulmonary disease and asthma: Systematic review and meta-analysis. *J Med Internet Res*. 2021;23(3):1-19. doi:10.2196/24602

19. Zwerink M, Brusse-Keizer M, van der Valk PDLPM, et al. Self management for patients with chronic obstructive pulmonary disease. *Cochrane Database of Systematic Reviews*. 2014;2014(3). doi:10.1002/14651858.CD002990.pub3

20. Hodkinson A, Bower P, Grigoroglou C, et al. Self-management interventions to reduce healthcare use and improve quality of life among patients with asthma: systematic review and network meta-analysis. *BMJ*. 2020;370(m2521):1-12. doi:10.1136/BMJ.M2521

21. Huppmann P, Sczepanski B, Boensch M, et al. Effects of inpatient pulmonary rehabilitation in patients with interstitial lung disease. *European Respiratory Journal*. 2013;42(2):444-453. doi:10.1183/09031936.00081512

22. Zibrak JD, Price D. Interstitial lung disease: Raising the index of suspicion in primary care. *NPJ Prim Care Respir Med*. 2014;24. doi:10.1038/npjpcrm.2014.54

23. Trappenburg J, Jonkman N, Jaarsma T, et al. Self-management: One size does not fit all. *Patient Educ Couns*. 2013;92(1):134-137. doi:10.1016/j.pec.2013.02.009

24. Coster S, Norman I. Cochrane reviews of educational and self-management interventions to guide nursing practice: A review. *Int J Nurs Stud*. 2009;46(4):508-528. doi:10.1016/j.ijnurstu.2008.09.009

25. Aronson KI, Suzuki A. Health Related Quality of Life in Interstitial Lung Disease: Can We Use the Same Concepts Around the World? *Front Med (Lausanne)*. 2021;8(October):1-12. doi:10.3389/fmed.2021.745908

26. Page MJ, Moher D, Bossuyt PM, et al. PRISMA 2020 explanation and elaboration: Updated guidance and exemplars for reporting systematic reviews. *The BMJ*. 2021;372. doi:10.1136/bmj.n160

27. Higgins JPT, Thomas J, Chandler J, et al. *Cochrane Handbook for Systematic Reviews of Interventions. 2nd Edition.*; 2019. doi:10.1002/9781119536604

28. Collard HR, Ryerson CJ, Corte TJ, et al. Acute Exacerbation of Idiopathic Pulmonary Fibrosis An International Working Group Report. Published online 2016. doi:10.1164/rccm.201604-0801CI

29. Effing TW, Vercoulen JH, Bourbeau J, et al. Definition of a COPD self-management intervention: International expert group consensus. *European Respiratory Journal*. 2016;48(1):46-54. doi:10.1183/13993003.00025-2016

30. Kaplan RM, Hays RD. Health-Related Quality of Life Measurement in Public Health Keywords. *Annu Rev Public Health*. 2022;43:355-373. doi:10.1146/annurev-publhealth

31. Kline Leidy N. Functional status and the forward progress of merry-go-rounds: Toward a coherent analytical framework. *Nurs Res*. 1994;43(4):196-202.

32. Shi D, Li Z, Yang J, Liu BZ, Xia H. Symptom experience and symptom burden of patients following first-ever stroke within 1 year: A cross-sectional study. *Neural Regen Res*. 2018;13(11):1907-1912. doi:10.4103/1673-5374.239440

33. Carrasquillo O. Health Care Utilization BT - Encyclopedia of Behavioral Medicine. In: Gellman MD, Turner JR, eds. Springer New York; 2013:909-910. doi:10.1007/978-1-4419-1005-9_885

34. NIH. Definition of overall survival - NCI Dictionary of Cancer Terms - National Cancer Institute. National Cancer Institute. Published 2021. Accessed June 17, 2022. https://www.cancer.gov/publications/dictionaries/cancer-terms/def/overall-survival-rate

35. Ahn E, Kang H. Introduction to systematic review and meta-analysis. *Korean J Anesthesiol*. 2018;71(2):103-112. doi:10.4097/kjae.2018.71.2.103

36. Newman S, Steed L, Mulligan K. Self-management interventions for chronic illness. *Lancet*. 2004;364:1523-1537.

37. Cottin V. Interstitial lung disease. *European Respiratory Review*. 2013;22(127):26-32. doi:10.1183/09059180.00006812

38. Antoniou KM, Margaritopoulos GA, Tomassetti S, Bonella F, Costabel U, Poletti V. Interstitial lung disease. *European Respiratory Review*. 2014;23(131):40-54. doi:10.1183/09059180.00009113

39. Barlow J, Wright C, Sheasby J, Turner A, Hainsworth J. *Self-Management Approaches for People with Chronic Conditions: A Review*.

40. Delameillieure A, Dobbels F, Vandekerkhof S, Wuyts WA. Patients' and healthcare professionals' perspectives on the idiopathic pulmonary fibrosis care journey: a qualitative study. *BMC Pulm Med*. 2021;21(1). doi:10.1186/s12890-021-01431-8

41. Cuthbert CA, Farragher JF, Hemmelgarn BR, Ding Q, McKinnon GP, Cheung WY. Self-management interventions for cancer survivors: A systematic review and evaluation of intervention content and theories. *Psychooncology*. 2019;28(11):2119-2140. doi:10.1002/pon.5215

42. Guler SA, Corte TJ. Interstitial Lung Disease in 2020: A History of Progress. *Clin Chest Med*. 2021;42(2):229-239. doi:10.1016/j.ccm.2021.03.001

43. Haase KR, Puts M, Sattar S, et al. Protocol for a systematic review of self-management interventions for older adults living with cancer. *Syst Rev*. 2020;9(1). doi:10.1186/s13643-020-01346-1

44. Gobeil-Lavoie AP, Chouinard MC, Danish A, Hudon C. Characteristics of self-management among patients with complex health needs: A thematic analysis review. *BMJ Open*. 2019;9(5). doi:10.1136/bmjopen-2018-028344

45. Delameillieure A, Vandekerkhof S, van Grootven B, Wuyts WA, Dobbels F. Care programs and their components for patients with idiopathic pulmonary fibrosis: a systematic review. *Respir Res*. 2021;22(1). doi:10.1186/s12931-021-01815-8

46. Swigris JJ, Brown KK, Abdulqawi R, et al. Patients' perceptions and patient-reported outcomes in progressive-fibrosing interstitial lung diseases. *European Respiratory Review*. 2018;27(150). doi:10.1183/16000617.0075-2018

47. Sunjaya AP, Sengupta A, Martin A, di Tanna GL, Jenkins C. Efficacy of self-management mobile applications for patients with breathlessness: Systematic review and quality assessment of publicly available applications. *Respir Med*. 2022;201. doi:10.1016/j.rmed.2022.106947

48. Cottin V, Wollin L, Fischer A, Quaresma M, Stowasser S, Harari S. Fibrosing interstitial lung diseases: Knowns and unknowns. *European Respiratory Review*. 2019;28(151). doi:10.1183/16000617.0100-2018

49. Wuyts WA, Peccatori FA, Russell AM. Patient-centred management in idiopathic pulmonary fibrosis: Similar themes in three communication models. *European Respiratory Review*. 2014;23(132):231-238. doi:10.1183/09059180.00001614

50. Moor CC, Mostard RLM, Grutters JC, et al. Home monitoring in patients with idiopathic pulmonary fibrosis: A randomized controlled trial. *Am J Respir Crit Care Med*. 2020;202(3):393-401. doi:10.1164/rccm.202002-0328OC

51. Lindell KO, Klein SJ, Veatch MS, et al. Nurse-led palliative care clinical trial improves knowledge and preparedness in caregivers of patients with idiopathic pulmonary fibrosis. *Ann Am Thorac Soc*. 2021;18(11):1811-1821. doi:10.1513/AnnalsATS.202012-1494OC

52. Lindell KO, Olshansky E, Song MK, 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*. 2010;39(4):304–313. doi:doi:10.1016/j.hrtlng.2009.08.005

53. Khor YH, Saravanan K, Holland AE, et al. A mixed-methods pilot study of handheld fan for breathlessness in interstitial lung disease. *Sci Rep*. 2021;11(6874):1-8. doi:10.1038/s41598-021-86326-8

54. Wagner EH. Chronic disease management: what will it take to improve care for chronic illness? *Eff Clin Pract*. 1998;1(1):2-4. http://www.ncbi.nlm.nih.gov/pubmed/10345255

55. Jolly K, Sidhu MS, Bates E, et al. Systematic review of the effectiveness of community-based self-management interventions among primary care COPD patients. *NPJ Prim Care Respir Med*. 2018;28(1). doi:10.1038/s41533-018-0111-9

56. Ouzzani M, Hammady H, Fedorowicz Z, Elmagarmid A. Rayyan—a web and mobile app for systematic reviews. *Syst Rev*. 2016;5(1):1-10. doi:10.1186/S13643-016-0384-4

57. Sterne JAC, Savović J, Page MJ, et al. RoB 2: A revised tool for assessing risk of bias in randomised trials. *The BMJ*. 2019;366. doi:10.1136/bmj.l4898

58. Higgins JP, Savović J, Page MJ, Sterne JAC. RoB 2 Guidance: Parallel Trial. *The Cochrane Collaboration*. 2019;(July):1-24.

59. Harbord RM, Harris RJ, Sterne JAC. Updated tests for small-study effects in meta-analyses. *Stata J*. 2009;9(2):197-210.

60. Lin L, Chu H, Murad MH, et al. Empirical Comparison of Publication Bias Tests in Meta-Analysis. *J Gen Intern Med*. 2018;33(8):1260-1267. doi:10.1007/s11606-018-4425-7

61. Page MJ, Higgins JP, Sterne JA. Chapter 13: Assessing risk of bias due to missing results in a synthesis | Cochrane Training. In: *Cochrane Handbook for Systematic Reviews of Interventions*. ; 2019.

62. Ahmed I, Sutton AJ, Riley RD. Assessment of publication bias, selection bias, and unavailable data in meta-analyses using individual participant data: A database survey. *BMJ (Online)*. 2012;344(7838):1-10. doi:10.1136/bmj.d7762

63. Schunemann H, Bro J, Guyatt G, et al. *GRADE Handbook*.; 2015. Accessed June 18, 2022. https://gdt.gradepro.org/app/handbook/handbook.html#h.9rdbelsnu4iy

64. McHugh M L. Interrater reliability: the kappa statistic. *Biochem Med (Zagreb)*. 2012;22(3):276-282.

65. Review Manager (RevMan).

66. du Prel JB, Hommel G, Röhrig B, Blettner M. Confidence Interval or P-Value?Part 4 of a Series on Evaluation of Scientific Publications. *Dtsch Arztebl*. 2009;106(19):335-339. doi:10.3238/arztebl.2009.0335

67. Hancock M, Kent P. Interpretation of dichotomous outcomes: Risk, odds, risk ratios, odds ratios and number needed to treat. *J Physiother*. 2016;62(3):172-174. doi:10.1016/j.jphys.2016.02.016

68. Cohen J. A power primer. *Psychol Bull*. 1992;112(1):155-159. doi:10.1037/0033-2909.112.1.155

69. Nolan CM, Birring SS, Maddocks M, et al. King's Brief Interstitial Lung Disease questionnaire: responsiveness and minimum clinically important difference. *Eur Respir J*. 2019;54(3). doi:10.1183/13993003.00281-2019

70. Yates H, Adamali HI, Maskell N, Barratt S, Sharp C. Visual analogue scales for interstitial lung disease: a prospective validation study. *QJM*. 2018;111(8):531-539. doi:10.1093/QJMED/HCY102

71. Ekström MP, Bornefalk H, Sköld CM, et al. Minimal Clinically Important Differences and Feasibility of Dyspnea-12 and the Multidimensional Dyspnea Profile in Cardiorespiratory Disease. *J Pain Symptom Manage*. 2020;60(5):968-975.e1. doi:10.1016/J.JPAINSYMMAN.2020.05.028

72. Chen T, Tsai APY, Hur SA, et al. Validation and minimum important difference of the UCSD Shortness of Breath Questionnaire in fibrotic interstitial lung disease. *Respir Res*. 2021;22(1). doi:10.1186/s12931-021-01790-0

73. Kalluri M, Luppi F, Vancheri A, et al. Patient-reported outcomes and patient-reported outcome measures in interstitial lung disease: Where to go from here? *European Respiratory Review*. 2021;30(160). doi:10.1183/16000617.0026-2021

74. Tsai APY, Hur SA, Wong A, et al. Minimum important difference of the EQ-5D-5L and EQ-VAS in fibrotic interstitial lung disease. *Thorax*. 2021;76(1):37-43. doi:10.1136/THORAXJNL-2020-214944

75. Puhan MA, Frey M, Büchi S, Schünemann HJ. The minimal important difference of the hospital anxiety and depression scale in patients with chronic obstructive pulmonary disease. *Health Qual Life Outcomes*. 2008;6. doi:10.1186/1477-7525-6-46

76. Driver CN, Novotny PJ, Benzo RP. Differences in Sedentary Time, Light Physical Activity, and Steps Associated with Better COPD Quality of Life. *Chronic Obstructive Pulmonary Diseases: Journal of the COPD Foundation*. 2022;9(1):34. doi:10.15326/JCOPDF.2021.0230

77. Campbell M, McKenzie JE, Sowden A, et al. Synthesis without meta-analysis (SWiM) in systematic reviews: Reporting guideline. *The BMJ*. 2020;368. doi:10.1136/bmj.l6890

78. Boon MH, Thomson H. The effect direction plot revisited: Application of the 2019 Cochrane Handbook guidance on alternative synthesis methods. *Res Synth Methods*. 2021;12(1):29-33. doi:10.1002/JRSM.1458

79. Jonkman NH, Schuurmans MJ, Groenwold RHH, Hoes AW, Trappenburg JCA. Identifying components of self-management interventions that improve health-related quality of life in chronically ill patients: Systematic review and meta-regression analysis. *Patient Educ Couns*. 2016;99(7):1087-1098. doi:10.1016/J.PEC.2016.01.022

80. Flather MD, Farkouh ME, Pogue JM, Yusuf S. Strengths and limitations of meta-analysis: Larger studies may be more reliable. *Control Clin Trials*. 1997;18(6):568-579. doi:10.1016/S0197-2456(97)00024-X

81. Davey J, Turner RM, Clarke MJ, Higgins JP. Characteristics of meta-analyses and their component studies in the Cochrane Database of Systematic Reviews: A cross-sectional, descriptive analysis. *BMC Med Res Methodol*. 2011;11. doi:10.1186/1471-2288-11-160

82. Schuurmans H, Steverink N, Frieswijk N, Buunk BP, Slaets JPJ, Lindenberg S. How to measure self-management abilities in older people by self-report. The development of the SMAS-30. *Quality of Life Research*. 2005;14(10):2215-2228. doi:10.1007/s11136-005-8166-9

83. Roberts NJ, Kidd L, Dougall N, Patel IS, McNarry S, Nixon C. Measuring patient activation: The utility of the Patient Activation Measure within a UK context—Results from four exemplar studies and potential future applications. *Patient Educ Couns*. 2016;99(10):1739-1746. doi:10.1016/J.PEC.2016.05.006

84. Jonkman NH, Schuurmans MJ, Jaarsma T, Shortridge-Baggett LM, Hoes AW, Trappenburg JCA. Self-management interventions: Proposal and validation of a new operational definition. *J Clin Epidemiol*. 2016;80:34-42. doi:10.1016/j.jclinepi.2016.08.001

85. Ann Saketkoo L, Mittoo S, Huscher D, et al. Connective tissue disease related interstitial lung diseases and idiopathic pulmonary fibrosis: provisional core sets of domains and instruments for use in clinical trials. *thorax* . 2014;69:428-436. doi:10.1136/thoraxjnl-2013-204202

86. Kawano-Dourado L, Glassberg MK, Assayag D, et al. Sex and gender in interstitial lung diseases. *European Respiratory Review*. 2021;30(162). doi:10.1183/16000617.0105-2021

87. Kaul B, Cottin V, Collard HR, Valenzuela C. Variability in Global Prevalence of Interstitial Lung Disease. *Front Med (Lausanne)*. 2021;8(November):1-10. doi:10.3389/fmed.2021.751181

Appendix

Index

Appendix 1 – Preferred reporting items for systematic reviews and meta-analysis (PRISMA) 2020 checklist

Section and Topic	Item #	Checklist item	Location where item is reported
TITLE			
Title	1	Identify the report as a systematic review.	1
ABSTRACT			
Abstract	2	See the PRISMA 2020 for Abstracts checklist.	-
INTRODUCTION			
Rationale	3	Describe the rationale for the review in the context of existing knowledge.	13, 14
Objectives	4	Provide an explicit statement of the objective(s) or question(s) the review addresses.	14
METHODS			
Eligibility criteria	5	Specify the inclusion and exclusion criteria for the review and how studies were grouped for the syntheses.	14-16
Information sources	6	Specify all databases, registers, websites, organisations, reference lists and other sources searched or consulted to identify studies. Specify the date when each source was last searched or consulted.	16, 17
Search strategy	7	Present the full search strategies for all databases, registers and websites, including any filters and limits used.	17 and appendix
Selection process	8	Specify the methods used to decide whether a study met the inclusion criteria of the review, including how many reviewers screened each record and each report retrieved, whether they worked independently, and if applicable, details of automation tools used in the process.	17, 18
Data collection process	9	Specify the methods used to collect data from reports, including how many reviewers collected data from each report, whether they worked independently, any processes for obtaining or confirming data from study investigators, and if applicable, details of automation tools used in the process.	18
Data items	10a	List and define all outcomes for which data were sought. Specify whether all results that were compatible with each outcome domain in each study were sought (e.g., for all measures, time points, analyses), and if not, the methods used to decide which results to collect.	15, 16, 18
	10b	List and define all other variables for which data were sought (e.g. participant and intervention characteristics, funding sources). Describe any assumptions made about any missing or unclear information.	18
Study risk of bias assessment	11	Specify the methods used to assess risk of bias in the included studies, including details of the tool(s) used, how many reviewers assessed each study and whether they worked independently, and if applicable, details of automation tools used in the process.	18, 19
Effect measures	12	Specify for each outcome the effect measure(s) (e.g. risk ratio, mean difference) used in the synthesis or presentation of results.	20-22
Synthesis methods	13a	Describe the processes used to decide which studies were eligible for each synthesis (e.g. tabulating the study intervention characteristics and comparing against the planned groups for each synthesis (item #5)).	20-22
	13b	Describe any methods required to prepare the data for presentation or synthesis, such as handling of missing summary statistics, or data conversions.	20-22
	13c	Describe any methods used to tabulate or visually display results of individual studies and syntheses.	20-22
	13d	Describe any methods used to synthesize results and provide a rationale for the choice(s). If meta-analysis was performed, describe the model(s), method(s) to identify the presence and extent of statistical heterogeneity, and software package(s) used.	20-22
	13e	Describe any methods used to explore possible causes of heterogeneity among study results (e.g. subgroup analysis, meta-regression).	Not done
	13f	Describe any sensitivity analyses conducted to assess robustness of the synthesized results.	21
Reporting bias	14	Describe any methods used to assess risk of bias due to missing results in a synthesis (arising from reporting biases).	19

assessment			
Certainty assessment	15	Describe any methods used to assess certainty (or confidence) in the body of evidence for an outcome.	19
RESULTS			
Study selection	16a	Describe the results of the search and selection process, from the number of records identified in the search to the number of studies included in the review, ideally using a flow diagram.	22, 23
	16b	Cite studies that might appear to meet the inclusion criteria, but which were excluded, and explain why they were excluded.	Appendix
Study characteristics	17	Cite each included study and present its characteristics.	24-29
Risk of bias in studies	18	Present assessments of risk of bias for each included study.	23, 24 and appendix
Results of individual studies	19	For all outcomes, present, for each study: (a) summary statistics for each group (where appropriate) and (b) an effect estimate and its precision (e.g. confidence/credible interval), ideally using structured tables or plots.	30-43
Results of syntheses	20a	For each synthesis, briefly summarise the characteristics and risk of bias among contributing studies.	30-43 and appendix
	20b	Present results of all statistical syntheses conducted. If meta-analysis was done, present for each the summary estimate and its precision (e.g. confidence/credible interval) and measures of statistical heterogeneity. If comparing groups, describe the direction of the effect.	30-43
	20c	Present results of all investigations of possible causes of heterogeneity among study results.	Not done
	20d	Present results of all sensitivity analyses conducted to assess the robustness of the synthesized results.	33 and appendix
Reporting biases	21	Present assessments of risk of bias due to missing results (arising from reporting biases) for each synthesis assessed.	Not reported
Certainty of evidence	22	Present assessments of certainty (or confidence) in the body of evidence for each outcome assessed.	43-45
DISCUSSION			
Discussion	23a	Provide a general interpretation of the results in the context of other evidence.	46, 47
	23b	Discuss any limitations of the evidence included in the review.	48
	23c	Discuss any limitations of the review processes used.	48
	23d	Discuss implications of the results for practice, policy, and future research.	47
OTHER INFORMATION			
Registration and protocol	24a	Provide registration information for the review, including register name and registration number, or state that the review was not registered.	14
	24b	Indicate where the review protocol can be accessed, or state that a protocol was not prepared.	14
	24c	Describe and explain any amendments to information provided at registration or in the protocol.	14 and SM
Support	25	Describe sources of financial or non-financial support for the review, and the role of the funders or sponsors in the review.	49
Competing interests	26	Declare any competing interests of review authors.	49
Availability of data, code and other materials	27	Report which of the following are publicly available and where they can be found: template data collection forms; data extracted from included studies; data used for all analyses; analytic code; any other materials used in the review.	49

Appendix 2 – Preferred reporting items for systematic reviews and meta-analysis (PRISMA) 2020 abstract checklist

Section and Topic	Item #	Checklist item	Reported (Yes/No)
TITLE			
Title	1	Identify the report as a systematic review.	Yes
BACKGROUND			
Objectives	2	Provide an explicit statement of the main objective(s) or question(s) the review addresses.	Yes
METHODS			
Eligibility criteria	3	Specify the inclusion and exclusion criteria for the review.	Yes
Information sources	4	Specify the information sources (e.g., databases, registers) used to identify studies and the date when each was last searched.	Yes
Risk of bias	5	Specify the methods used to assess risk of bias in the included studies.	Yes
Synthesis of results	6	Specify the methods used to present and synthesise results.	Yes
RESULTS			
Included studies	7	Give the total number of included studies and participants and summarise relevant characteristics of studies.	Yes
Synthesis of results	8	Present results for main outcomes, preferably indicating the number of included studies and participants for each. If meta-analysis was done, report the summary estimate and confidence/credible interval. If comparing groups, indicate the direction of the effect (i.e. which group is favoured).	Yes
DISCUSSION			
Limitations of evidence	9	Provide a brief summary of the limitations of the evidence included in the review (e.g., study risk of bias, inconsistency and imprecision).	Yes
Interpretation	10	Provide a general interpretation of the results and important implications.	Yes
OTHER			
Funding	11	Specify the primary source of funding for the review.	No
Registration	12	Provide the register name and registration number.	Yes

Appendix 3 – Amendment to the review protocol on the definition of self-management intervention

In the initial protocol, self-management intervention was operationalized as:

> "[…] any structured and individualized multicomponent or isolated intervention aimed at empowering the person in the day-to-day management of his / her condition will be included. Additionally, these interventions must also include at least one of the following components: (1) Self-monitoring, self-recording (optional), and self-intervention; (2) Behavior change component; (3) Development of active relationships between patient, healthcare professional, family, or other community members; and (4) Development of action plans or skills to live with the biological, psychological, or social consequences of the disease."

However, due to few studies being included at the end of the second screening phase (n = 3), the definition of self-management intervention was changed to a more inclusive definition that was previously published in a consensus statement[1]. With that definition, the records that were excluded in the first and second screening phases for not complying with the previous definition of self-management intervention were re-screened. This resulted in the inclusion of 1 additional study[2]. This amendment was registered in the review protocol and approved on the 12th of December 2022.

Appendix 4 – Search strategy of each database

PubMed/MEDLINE

#1 ("anti-glomerular basement membrane disease" [tw] OR "coal workers pneumoconiosis" [tw] OR "diffuse parenchymal lung disease*" [tw] OR "extrinsic allergic alveolitis" [tw] OR "granulomatosis with polyangiitis" [tw] OR "hypersensitivity pneumonitis" [tw] OR "idiopathic interstitial pneumonia*" [tw] OR "idiopathic pulmonary fibrosis" [tw] OR "interstitial lung disease*" [tw] OR "interstitial pneumonia*" [tw] OR "interstitial pneumonitis" [tw] OR "interstitial pulmonary disease*" [tw] OR "pulmonary fibrosis" [tw] OR "pulmonary langerhans-cell histiocytosis" [tw] OR "pulmonary sarcoidosis" [tw] OR "pulmonary siderosis" [tw] OR "radiation pneumonitis" [tw] OR DPLD [tw] OR ILD [tw] OR IPD [tw] OR IPF [tw] OR pneumoconiosis [tw] OR (anthracosis [tw] OR asbestosis [tw] OR berylliosis [tw] OR byssinosis [tw] OR "caplan syndrome" [tw] OR siderosis [tw] OR silicosis [tw]))

#2 ("alveolitis, extrinsic allergic" [mh] OR "anti-glomerular basement membrane disease" [mh] OR "histiocytosis, langerhans-cell" [mh] OR "idiopathic interstitial pneumonias" [mh] OR "idiopathic pulmonary fibrosis" [mh] OR "lung diseases, interstitial" [mh] OR "pulmonary fibrosis" [mh] OR "radiation pneumonitis" [mh] OR "sarcoidosis, pulmonary" [mh] OR pneumoconiosis [mh] OR (anthracosis [mh] OR asbestosis [mh] OR berylliosis [mh] OR byssinosis [mh] OR "caplan syndrome" [mh] OR siderosis [mh] OR silicosis [mh]))

#3 (autonom* [tw] OR empower* [tw] OR self-administ* [tw] OR self-assess* [tw] OR self-car* [tw] OR self-control* [tw] OR self-efficac* [tw]OR self-examination [tw] OR self-govern* [tw] OR self-help* [tw] OR self-inspection [tw] OR self-manag* [tw] OR self-medication [tw] OR self-monitor* [tw] OR "monitoring program" [tw] OR self-policing [tw] OR self-regulat* [tw] OR self-rule [tw] OR self-supervision [tw] OR self-testing [tw] OR self-treat* [tw] OR self-support* [tw] OR "management plan*" [tw] OR "management program*" [tw] OR "decision making" [tw] OR "behav* chang*" [tw] OR "action plan*" [tw] OR "disease management" [tw]**)**

#4 ("self administration" [mh] OR self-assessment [mh] OR "self care" [mh] OR self-efficacy [mh] OR empowerment [mh] OR self-examination [mh] OR self-control [mh] OR self-management [mh] OR self-medication [mh] OR "decision making" [mh] OR "behavior therapy" [mh] OR self-testing [mh] OR disease management [mh])

#5 #1 OR #2

#6 #3 OR #4

#7 #5 AND #6

Cochrane Central Register of Controlled Trials (CENTRAL)

#1	("anti-glomerular basement membrane disease" OR "coal workers pneumoconiosis" OR "diffuse parenchymal lung disease*" OR "extrinsic allergic alveolitis" OR "granulomatosis with polyangiitis" OR "hypersensitivity pneumonitis" OR "idiopathic interstitial pneumonia*" OR "idiopathic pulmonary fibrosis" OR "interstitial lung disease*" OR "interstitial pneumonia*" OR "interstitial pneumonitis" OR "interstitial pulmonary disease*" OR "pulmonary fibrosis" OR "pulmonary langerhans-cell histiocytosis" OR "pulmonary sarcoidosis" OR "pulmonary siderosis" OR "radiation pneumonitis" OR DPLD OR ILD OR IPD OR IPF OR pneumoconiosis OR anthracosis OR asbestosis OR berylliosis OR byssinosis OR "caplan syndrome" OR siderosis OR silicosis):ti,ab,kw
#2	MeSH descriptor: [Lung Diseases, Interstitial] explode all trees
#3	(autonom* OR empower* OR self-administ* OR self-assess* OR self-car* OR self-control* OR self-efficac* OR self-examination OR self-govern* OR self-help* OR self-inspection OR self-manag* OR self-medication OR self-monitor* OR "monitoring program" OR self-policing OR self-regulat* OR self-rule OR self-supervision OR self-testing OR self-treat* OR self-support* OR "management plan*" OR "decision making" OR "behav* chang*" OR "action plan*" OR "management program*" OR "disease management"):ti,ab,kw
#4	MeSH descriptor: [empowerment] explode all trees
#5	MeSH descriptor: [self-examination] explode all trees
#6	MeSH descriptor: [self-control] explode all trees
#7	MeSH descriptor: [self-assessment] explode all trees
#8	MeSH descriptor: [self efficacy] explode all trees
#9	MeSH descriptor: [self care] explode all trees
#10	MeSH descriptor: [self-management] explode all trees
#11	MeSH descriptor: [decision making] explode all trees
#12	MeSH descriptor: [behavior therapy] explode all trees
#13	MeSH descriptor: [disease management] explode all trees
#14	#1 OR #2
#15	#3 OR #4 OR #5 OR #6 OR #7 OR #8 OR #9 OR #10 OR #11 OR #12 OR #13
#16	#14 AND #15

Scopus

#1 TITLE-ABS-KEY("anti-glomerular basement membrane disease" OR "coal workers pneumoconiosis" OR "diffuse parenchymal lung disease*" OR "extrinsic allergic alveolitis" OR "granulomatosis with polyangiitis" OR "hypersensitivity pneumonitis" OR "idiopathic interstitial pneumonia*" OR "idiopathic pulmonary fibrosis" OR "interstitial lung disease*" OR "interstitial pneumonia*" OR "interstitial pneumonitis" OR "interstitial pulmonary disease*" OR "pulmonary fibrosis" OR "pulmonary langerhans-cell histiocytosis" OR "pulmonary sarcoidosis" OR "pulmonary siderosis" OR "radiation pneumonitis" OR DPLD OR ILD OR IPD OR IPF OR pneumoconiosis OR (anthracosis OR asbestosis OR berylliosis OR byssinosis OR "caplan syndrome" OR siderosis OR silicosis))

#2 TITLE-ABS-KEY(autonom* OR empower* OR self-administ* OR self-assess* OR self-car* OR self-control* OR self-efficac* OR self-examination OR self-govern* OR self-help* OR self-inspection OR self-manag* OR self-medication OR self-monitor* OR "monitoring program" OR self-policing OR self-regulat* OR self-rule OR self-supervision OR self-testing OR self-treat* OR self-support* OR "management plan*" OR "decision making" OR "behav* chang*" OR "action plan*" OR "management program*" OR "disease management")

#3 #1 AND #2

Web of Science Core Collection

#1 TS=("anti-glomerular basement membrane disease" OR "coal workers pneumoconiosis" OR "diffuse parenchymal lung disease*" OR "extrinsic allergic alveolitis" OR "granulomatosis with polyangiitis" OR "hypersensitivity pneumonitis" OR "idiopathic interstitial pneumonia*" OR "idiopathic pulmonary fibrosis" OR "interstitial lung disease*" OR "interstitial pneumonia*" OR "interstitial pneumonitis" OR "interstitial pulmonary disease*" OR "pulmonary fibrosis" OR "pulmonary langerhans-cell histiocytosis" OR "pulmonary sarcoidosis" OR "pulmonary siderosis" OR "radiation pneumonitis" OR DPLD OR ILD OR IPD OR IPF OR pneumoconiosis OR (anthracosis OR asbestosis OR berylliosis OR byssinosis OR "caplan syndrome" OR siderosis OR silicosis))

#2 TS=(autonom* OR empower* OR self-administ* OR self-assess* OR self-car* OR self-control* OR self-efficac* OR self-examination OR self-govern* OR self-help* OR self-inspection OR self-manag* OR self-medication OR self-monitor* OR "monitoring program" OR self-policing OR self-regulat* OR self-rule OR self-supervision OR self-testing OR self-treat* OR self-support* OR "management plan*" OR "decision making" OR "behav* chang*" OR "action plan*" OR "management program*" OR "disease management")

#3 #1 AND #2

PsycInfo (OVID)

#1 (anti-glomerular basement membrane disease or coal workers pneumoconiosis or diffuse parenchymal lung disease* or extrinsic allergic alveolitis or granulomatosis with polyangiitis or hypersensitivity pneumonitis or idiopathic interstitial pneumonia* or idiopathic pulmonary fibrosis or interstitial lung disease* or interstitial pneumonia* or interstitial pneumonitis or interstitial pulmonary disease* or pulmonary fibrosis or pulmonary langerhans-cell histiocytosis or pulmonary sarcoidosis or pulmonary siderosis or radiation pneumonitis or DPLD or ILD or IPD or IPF or pneumoconiosis or anthracosis or asbestosis or berylliosis or byssinosis or caplan syndrome or siderosis or silicosis).mp. [mp=title, abstract, heading word, table of contents, key concepts, original title, tests & measures, mesh word]

#2 (management program* or disease management or autonom* or empower* or self-administ* or self-assess* or self-control* or self-efficac* or self-examination or self-govern* or self-help* or self-inspection or self-manag* or self-medication or self-monitor* or monitoring program or self-policing or self-regulat* or self-rule or self-supervision or self-testing or self-treat* or self-support* or management plan* or decision making or behav* chang* or action plan*).mp. [mp=title, abstract, heading word, table of contents, key concepts, original title, tests & measures, mesh word]

#3 exp autonomy/

#4 Self-Evaluation/

#5 exp self-efficacy/ or exp self-management/ or exp self-monitoring/

#6 self-care/

#7 self-help techniques/ or behavior modification/

#8 self-regulation/

#9 self-medication/

#10 behavior change/ or behavior modification/

#11 disease management/

#12 2 or 3 or 4 or 5 or 6 or 7 or 8 or 9 or 10 or 11

#13 1 and 12

Excerpta Medica Database (EMBASE)

#1 (anti-glomerular basement membrane disease or coal workers pneumoconiosis or diffuse parenchymal lung disease* or extrinsic allergic alveolitis or granulomatosis with polyangiitis or hypersensitivity pneumonitis or idiopathic interstitial pneumonia* or idiopathic pulmonary fibrosis or interstitial lung disease* or interstitial pneumonia* or interstitial pneumonitis or interstitial pulmonary disease* or pulmonary fibrosis or pulmonary langerhans-cell histiocytosis or pulmonary sarcoidosis or pulmonary siderosis or radiation pneumonitis or DPLD or ILD or IPD or IPF or pneumoconiosis or anthracosis or asbestosis or berylliosis or byssinosis or caplan syndrome or siderosis or silicosis).mp. [mp=title, abstract, heading word, drug trade name, original title, device manufacturer, drug manufacturer, device trade name, keyword heading word, floating subheading word, candidate term word]
#2 exp interstitial lung disease/
#3 exp occupational lung disease/
#4 exp pneumoconiosis/
#5 exp lung fibrosis/
#6 exp lung sarcoidosis/
#7 exp lung alveolitis/
#8 exp fibrosing alveolitis/
#9 exp allergic pneumonitis/
#10 exp lung hemosiderosis/
#11 exp asbestosis/
#12 exp silicosis/
#13 (management program* or disease management or autonom* or empower* or self-administ* or self-assess* or self-control* or self-efficac* or self-examination or self-govern* or self-help* or self-inspection or self-manag* or self-medication or self-monitor* or monitoring program or self-policing or self-regulat* or self-rule or self-supervision or self-testing or self-treat* or self-support* or management plan* or decision making or behav* chang* or action plan*).mp. [mp=title, abstract, heading word, drug trade name, original title, device manufacturer, drug manufacturer, device trade name, keyword heading word, floating subheading word, candidate term word]
#14 exp patient autonomy/
#15 exp self evaluation/
#16 exp self care/
#17 exp empowerment/
#18 exp self control/
#19 exp self medication/
#20 exp behavior therapy/
#21 exp self-testing/
#22 exp decision making/
#23 #1 OR #2 OR #3 OR #4 OR #5 OR #6 OR #7 OR #8 OR #9 OR #10 OR #11 OR #12
#24 #13 OR #14 OR #15 OR #16 OR #17 OR #18 OR #19 OR #20 OR #21 OR #22
#25 #23 AND #24

Appendix 5 – Grading of recommendations, assessment, development, and evaluations (GRADE) assessment criteria used in the certainty assessment

1. **Risk of Bias**
   - If 50-74% of included studies have an overall high risk of bias – Rate down one level.
   - If 75% or more of the included studies have an overall high risk of bias – Rate down two levels.

Note: The assessment of this criteria was based on just the percentage of studies with high RoB because in studies that tested self-management interventions the risk of bias is generally higher due to the content of the interventions and not because of poor study methods/execution (e.g., lack of blinding in included studies).

2. **Inconsistency**
   - If the p-value of the Cochran Q test is <0.1, and the Higgins I^2 statistic is ≥50%, statistically significant heterogeneity is assumed[3] – Rate down one level.
   - If only one of the criteria is present, examine whether the effect sizes of the studies are very different from each other and whether the 95% confidence intervals show minimal or no overlap, with a visual inspection of the forest plot, to confirm the presence/absence of significant heterogeneity (i.e., there is a significant inconsistency) – Rate down one level if detected.
3. **Indirectness**
   - If participants differ from those of interest – Rate down one level.
   - If the tested intervention in the included studies differs from the intervention of interest – Rate down one level.
   - If the outcomes (and outcome measures) differ from those of primary interest – Rate down one level.
   - If direct comparisons between two or more interventions of interest are not available (e.g., being interested in evaluating drug A vs drug B, but we only have studies that evaluate drug A vs placebo and drug B vs placebo) – Rate down one level.

Note: Maximum rate down two levels.

4. Imprecision

- Assess whether there is enough information to calculate an accurate effect estimate[4]. For dichotomous results, information is likely to be insufficient if the total number of events is less than 300, or if the total (cumulative) sample size is less than the number of participants necessary for a study with adequate power (i.e., sample calculation)[4]. For continuous results, information is likely to be insufficient if the total number of participants is less than 400[4].
- Assess the accuracy of the effect estimate (i.e., confidence interval)[4]. For dichotomous results, a confidence interval that includes a reduction in relative risk or an increase in relative risk greater than 25% in the limits of the confidence interval, suggests a decrease in the precision of the result[4]. For continuous results, if the limits of the confidence interval cross the effect size (i.e., standardized mean difference) of 0.5 in either direction suggests a decrease in the precision of the result[4].
- Based on the above criteria, define whether there is any inaccuracy (rate down one level) or if there is a serious inaccuracy (rate down two levels). If only one of the criteria is met, rate down one level; if the two criteria are met, rate down two levels.

5. Publication bias

- If 50% of the studies made the sample calculation and if 50% of the studies reached the number of participants defined by the sample calculation – Do not rate down.
- If 50% of the studies did not calculate the sample size needed for the trial or if 50% of the studies did not reach the number of participants defined by the sample calculation – Rate down one level.
- If 75% of the studies did not carry out the sample calculation or if 75% of the studies did not reach the number of participants defined by the sample calculation – Rate down two levels.

6. Rating up the quality of evidence

We did not raise the level of evidence, as the grading of recommendations, assessment, development, and evaluations (GRADE) guidelines do not give clear guidance for rating up randomized controlled trials.

Appendix 6 – Reasons for record exclusion in the full-text screening phase

First author Year	Title	Reason for exclusion
Sinclair, C. 2017	Advance care planning uptake among patients with severe lung disease: a randomised patient preference trial of a nurse-led, facilitated advance care planning intervention [5]	No outcome of interest
Hoffman, B. 2015	Development and psychometric properties of the Pulmonary-specific Quality-of-Life Scale in lung transplant patients [6]	No RCT
Timms, K. 2014	A dynamical systems approach to understanding self-regulation in smoking cessation behavior change [7]	No participants with ILD
Moor, C. 2020	A randomized controlled trial of a home monitoring program in newly treated patients with idiopathic pulmonary fibrosis [8]	No RCT
Rodrigue, J. 2005	A randomized evaluation of quality-of-life therapy with patients awaiting lung transplantation [9]	Wrong comparator
Pumar, M. 2019	Cognitive behavioural therapy (CBT) for patients with chronic lung disease and psychological comorbidities undergoing pulmonary rehabilitation [10]	Mixed population
Kalluri, M. 2016	Early integrated palliative care in a multidisciplinary interstitial lung disease (ILD) collaborative reduces hospitalizations for idiopathic pulmonary fibrosis (IPF) [11]	No RCT
De Las Heras, J. 2022	Effect of a Telerehabilitation program in sarcoidosis [12]	No SMI
Prajapat, B. 2011	Effect of mid-thigh cross sectional area on CT as a marker of muscle mass in interstitial lung diseases after pulmonary rehabilitation [13]	No RCT
Thombs, B. 2020	Evaluation of the Scleroderma Patient-centered Intervention Network COVID-19 Home-isolation Activities Together Program [14]	No RCT
Lindell, K. 2018	Feasibility and acceptability of an early palliative care intervention in patients with idiopathic pulmonary fibrosis and their caregivers [15]	No RCT
Reilly, C. 2012	Feasibility of a new out-patient breathlessness support service [16]	No RCT
Kohr, Y.H. 2018	Handheld fan for breathlessness in interstitial lung disease [17]	No RCT
Frith, P. 2013	Health outcomes in carer-patient dyads of a randomized control trial of carer training for patients receiving long term domiciliary oxygen therapy [18]	No RCT
Yuen, H. 2019	Home-Based Pulmonary Rehabilitation for Patients With Idiopathic Pulmonary Fibrosis: A pilot study [19]	No SMI
Johannson, K. 2017	Home monitoring improves endpoint efficiency in idiopathic pulmonary fibrosis [20]	No RCT
Wijsenbeek, M. 2018	Home Monitoring in Idiopathic Pulmonary Fibrosis; Improving Use of Anti-fibrotic Medication and Quality of Life [21]	No RCT
De Las Heras, J. 2020	Is Virtual Autonomous Physiotherapist Tele-rehabilitation Program feasible in Idiopathic Pulmonary Fibrosis? [22]	No RCT

Sinclair, C. 2020	Impact of a Nurse-Led Advance Care Planning Intervention on Satisfaction, Health-Related Quality of Life, and Health Care Utilization Among Patients With Severe Respiratory Disease: A Randomized Patient-Preference Trial [23]	Mixed population
Mazzoleni, S. 2014	Interactive videogame as rehabilitation tool of patients with chronic respiratory diseases: Preliminary results of a feasibility study [24]	No SMI
Wallaert, B. 2020	Long-term effects of pulmonary rehabilitation on daily life physical activity of patients with stage IV sarcoidosis: A randomized controlled trial [25]	No SMI
Early, F. 2015	Patient agenda setting in respiratory outpatients [26]	No outcome of interest
Van Manen, M. J. G. 2017	Patient and partner empowerment programme for idiopathic pulmonary fibrosis [27]	No RCT
Van Manen, M. J. G. 2016	Patient and partner "empowerment" program in idiopathic pulmonary fibrosis (ppepp): Improving quality of life in patients and their partners [28]	No RCT
Edwards, C. 2017	Patient-reported Monitoring of Symptoms and Spirometry Via the patientMpower Platform in Idiopathic Pulmonary Fibrosis [29]	No RCT
Mueller, K. 2017	Physical activity of patients with occupational lung diseases [30]	No RCT
Frith, P. 2020	Pragmatic randomised controlled trial of a personalised intervention for carers of people requiring home oxygen therapy [31]	No SMI
Holland, H. 2008	Short term improvement in exercise capacity and symptoms following exercise training in interstitial lung disease [32]	No SMI
Moretta, P. 2021	Subject preferences and psychological implications of portable oxygen concentrator versus compressed oxygen cylinder in chronic lung disease [33]	No SMI
Wong, C. 2012	Tele-monitoring of home oxygen user (THOU): A program to ensure maximal therapeutic benefit in patients commencing on long-term oxygen therapy (LTOT) [34]	No RCT
De Las Heras, J. 2019	Tele-rehabilitation program in idiopathic pulmonary fibrosis [35]	No RCT

Abbreviations - **ILD**: Interstitial Lung Disease; **RCT**: Randomized Controlled Trial; **SMI**: Self-Management Intervention.

Appendix 7 – Results of risk of bias assessment of between-group differences regarding the effects of self-management interventions in people with interstitial lung disease on secondary outcomes

First author-Year	Outcome measure	Result[a]	D1	D2	D3	D4	D5	Overall
Khor-2021	Lifespace mobility	2.4 (-10.4 to 15.2); 0.72	+	+	+	+	!	!
Khor-2021	MRADLQ	-2.5 (-4.8 to 0.3); 0.08	+	+	+	!	!	!
Khor-2021	Duration of sedentary time per day (mins)	66 (-30 to 162); 0.18	+	+	+	+	!	!
Khor-2021	Duration of time above 3 METs per day (mins)	26 (-16.1 to 68.3); 0.23	+	+	+	+	!	!
Khor-2021	Steps per day	74 (-807 to 956); 0.87	+	+	+	+	!	!
Khor-2021	Total energy expenditure (kCal/day)	104 (-197 to 404); 0.50	+	+	+	+	!	!
Khor-2021	Total METs	0.003 (-0.04 to 0.44); 0.88	+	+	+	+	!	!

Figure 1 – Risk of bias of between-group differences regarding the effect of self-management interventions versus usual or standard care on functional performance of people with interstitial lung disease according to the Cochrane risk-of-bias tool for randomized trials.

[a] Data is presented as mean (95% confidence interval); p-value unless otherwise stated.
Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations - **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **MET**: Metabolic Equivalent of Task; **MRADLQ**: Manchester Respiratory Activities of Daily Living Questionnaire.

First author-Year	Outcome measure	Result[a]	D1	D2	D3	D4	D5	Overall
Lindell-2010	Beck Anxiety Inventory	p-value: 0.077	!	!	+	!	-	-
Lindell-2010	Beck Depression Inventory-II	p-value: 0.894	!	!	+	!	-	-
Lindell-2010	Perceived Stress Scale	p-value: 0.531	!	!	+	!	-	-
Moor-2020	HADS Anxiety	-0.05 (-1.08 to 0.99); 0.93	!	!	+	!	!	!
Moor-2020	HADS Depression	-0.40 (-1.61 to 0.81); 0.51	!	!	!	!	!	!
Khor-2021	SEMCD6	0.07 (-1.9 to 2.0); 0.94	+	+	+	+	!	!
Lindell-2021	PROMIS-29 Anxiety/fear	0.01 (-5.55 to 5.57); 0.99	-	+	!	!	!	-
Lindell-2021	PROMIS-29 Depression/sadness	-0.12 (-6.10 to 5.86); 0.97	-	+	!	!	!	-
Lindell-2021	PROMIS-29 Satisfaction with social roles	3.66 (1.02 to 8.34); 0.12	-	+	+	!	!	-
Lindell-2021	Perceived Stress Scale	1.23 (-1.59 to 4.05); 0.39	-	+	+	!	!	-

Figure 2 – Risk of bias of between-group differences regarding the effect of self-management interventions versus usual or standard care on psychological and social outcomes of people with interstitial lung disease according to the Cochrane risk-of-bias tool for randomized trials.

[a] Data is presented as mean (95% confidence interval); p-value unless otherwise stated.
Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **HADS**: Hospital Anxiety and Depression Scale; **PROMIS-29**: Patient Reported Outcome Measurement Information System-29; **SEMCD6**: Self-efficacy for Managing Chronic Disease 6-item Scale.

First author-Year	Outcome measure	Result[a]	D1	D2	D3	D4	D5	Overall
Lindell-2010	UCSDSoBQ	p-value: 0.972	!	!	+	!	-	-
Moor-2020	VAS Cough	0.82 (-0.52 to 2.17); 0.23	!	!	!	!	!	!
Moor-2020	VAS Dyspnea	0.63 (-0.23 to 1.50); 0.15	!	!	+	!	!	!
Moor-2020	VAS Fatigue	0.18 (-0.88 to 1.23); 0.74	!	!	+	!	!	!
Khor-2021	Dyspnea-12	-2.2 (-6.4 to 1.9); 0.29	+	+	+	+	!	!
Lindell-2021	PROMIS-29 Pain interference	1.38 (-5.37 to 2.62); 0.49	-	+	+	!	!	-
Lindell-2021	PROMIS-29 Fatigue	-1.35 (-5.78 to 3.07); 0.54	-	+	!	!	!	-
Lindell-2021	PROMIS29 Sleep disturbance	2.63 (-0.53 to 5.79); 0.10	-	+	+	!	!	-

Figure 3 – Risk of bias of between-group differences regarding the effect of self-management interventions versus usual or standard care on symptoms of people with interstitial lung disease according to the Cochrane risk-of-bias tool for randomized trials.

[a] Data is presented as mean (95% confidence interval); p-value unless otherwise stated.
Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **PROMIS-29**: Patient Reported Outcome Measurement Information System-29; **UCSDSoBQ**: University of California at San Diego Shortness of Breath Questionnaire; **VAS**: Visual Analog Scale.

First author-Year	Outcome measure	Result[a]	D1	D2	D3	D4	D5	Overall
Moor-2020	Extra visits	1.25 (0.61 to 2.54); 0.55	!	!	+	-	!	-
Moor-2020	Hospitalizations	p-value: 0.27	!	!	+	-	!	-
Lindell-2021	Emergency visits	1.39 (0.40 to 4.79); 0.61	-	+	!	!	!	-
Lindell-2021	Inpatient visits	1.69 (0.61 to 4.67); 0.31	-	+	!	!	!	-
Lindell-2021	Outpatient visits	0.93 (0.796 to 1.09); 0.38	-	+	!	!	!	-

Figure 4 – Risk of bias of between-group differences regarding the effect of self-management interventions versus usual or standard care on unplanned healthcare visits of people with interstitial lung disease according to the Cochrane risk-of-bias tool for randomized trials.

[a] Data is presented as risk ratio (95% confidence interval); p-value unless otherwise stated.
Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result.

Appendix 8 – Sensitivity analysis regarding the effects of self-management interventions versus usual or standard care on health-related quality of life of people with interstitial lung disease

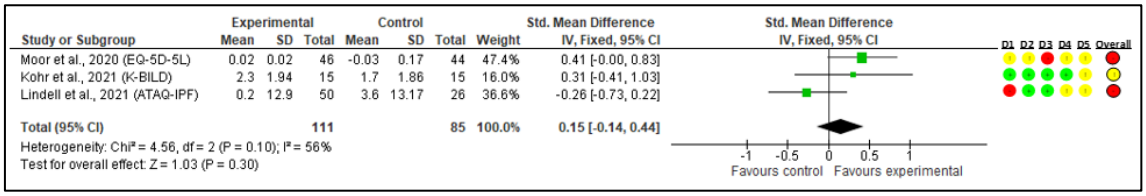


Figure 5 – Meta-analysis on the effect of self-management interventions versus usual or standard care on health-related quality of life of people with interstitial lung disease measured with 5-level euroqol 5-dimensional questionnaire, king's brief interstitial lung disease health status questionnaire, and the tool to assess quality of life in idiopathic pulmonary fibrosis (n = 3).

Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **ATAQ-IPF**: A Tool to Assess Quality of Life in Idiopathic Pulmonary Fibrosis; **CI**: Confidence Interval; **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **EQ-5D-5L**: 5-level EuroQol 5-Dimensional questionnaire; **IV**: Inverse-Variance; **K-BILD**: King's Brief Interstitial Lung Disease health status questionnaire; **P**: P-value; **SD**: Standard Deviation.

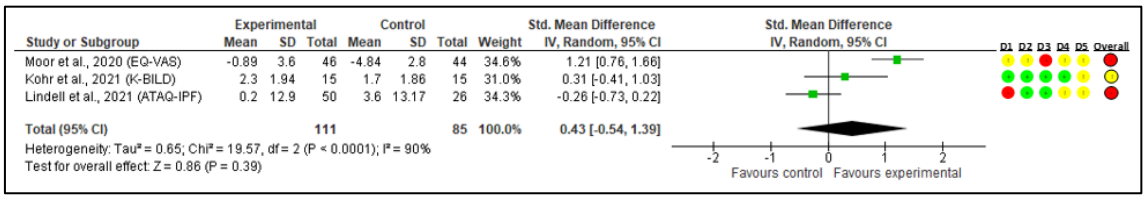


Figure 6 – Meta-analysis on the effect of self-management interventions versus usual or standard care on health-related quality of life of people with interstitial lung disease measured with EuroQol-visual analog scale, king's brief interstitial lung disease health status questionnaire, and the tool to assess quality of life in idiopathic pulmonary fibrosis (n = 3).

Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **ATAQ-IPF**: A Tool to Assess Quality of Life in Idiopathic Pulmonary Fibrosis; **CI**: Confidence Interval; **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **EQ-VAS**: EuroQol-visual analog scale; **IV**: Inverse-Variance; **K-BILD**: King's Brief Interstitial Lung Disease health status questionnaire; **P**: P-value; **SD**: Standard Deviation.

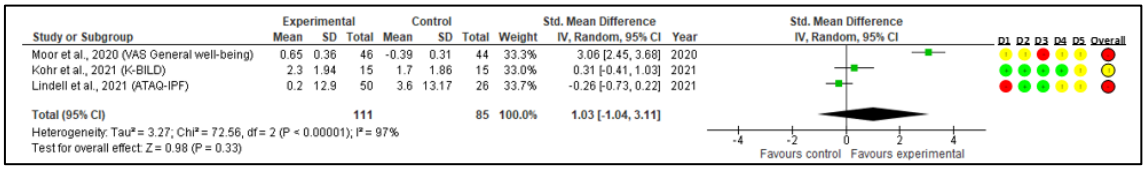


Figure 7 - Meta-analysis on the effect of self-management interventions versus usual or standard care on health-related quality of life of people with interstitial lung disease measured with visual analog scale, king's brief interstitial lung disease health status questionnaire, and the tool to assess quality of life in idiopathic pulmonary fibrosis (n = 3).

Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **ATAQ-IPF**: A Tool to Assess Quality of Life in Idiopathic Pulmonary Fibrosis; **CI**: Confidence Interval; **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **IV**: Inverse-Variance; **K-BILD**: King's Brief Interstitial Lung Disease health status questionnaire; **P**: P-value; **SD**: Standard Deviation; **VAS**: Visual Analog Scale.

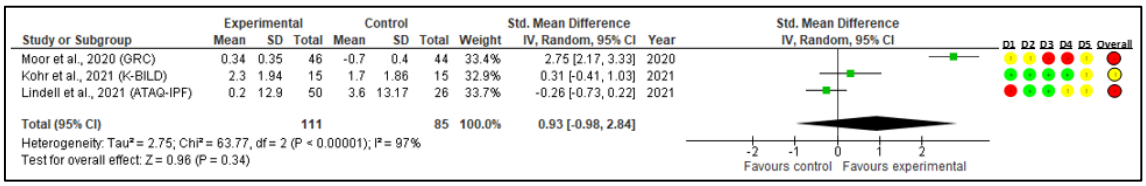


Figure 8 - Meta-analysis on the effect of self-management interventions versus usual or standard care on health-related quality of life of people with interstitial lung disease measured with global rating of change, king's brief interstitial lung disease health status questionnaire, and the tool to assess quality of life in idiopathic pulmonary fibrosis (n = 3).

Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **ATAQ-IPF**: A Tool to Assess Quality of Life in Idiopathic Pulmonary Fibrosis; **CI**: Confidence Interval; **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **GRC**: Global Rating of Change; **IV**: Inverse-Variance; **K-BILD**: King's Brief Interstitial Lung Disease health status questionnaire; **P**: P-value; **SD**: Standard Deviation.

References

1. Effing TW, Vercoulen JH, Bourbeau J, et al. Definition of a COPD self-management intervention: International expert group consensus. *European Respiratory Journal*. 2016;48(1):46-54. doi:10.1183/13993003.00025-2016

2. Khor YH, Saravanan K, Holland AE, et al. A mixed-methods pilot study of handheld fan for breathlessness in interstitial lung disease. *Sci Rep*. 2021;11(1). doi:10.1038/s41598-021-86326-8

3. Ahn E, Kang H. Introduction to systematic review and meta-analysis. *Korean J Anesthesiol*. 2018;71(2):103-112. doi:10.4097/kjae.2018.71.2.103

4. Ryan R. *How to GRADE the Quality of the Evidence*. https://gradepro.org/

5. Sinclair C, Auret KA, Evans SF, et al. Advance care planning uptake among patients with severe lung disease: a randomised patient preference trial of a nurse-led, facilitated advance care planning intervention. *BMJ Open*. 2017;7(2).

6. Hoffman BM, Stonerock GL, Smith PJ, et al. Development and psychometric properties of the Pulmonary-specific Quality-of-Life Scale in lung transplant patients. *Heart Lung Transplant*. 2015;34(8):1058-1065. doi:10.1016/j.healun.2015.03.005

7. Timms KP, Rivera DE, Collins LM, Piper ME. A dynamical systems approach to understanding self-regulation in smoking cessation behavior change. *Nicotine Tob Res*. 2014;16 Suppl 2(Suppl 2). doi:10.1093/NTR/NTT149

8. Moor C, Mostard RLM, Grutters JC, et al. A randomized controlled trial of a home monitoring program in newly treated patients with idiopathic pulmonary fibrosis. *Am J Respir Crit Care Med*. 2020;201(1). https://www.atsjournals.org/doi/abs/10.1164/ajrccm-conference.2020.201.1_MeetingAbstracts.A4553

9. Rodrigue JR, Baz MA, Widows MR, Ehlers SL. A randomized evaluation of quality-of-life therapy with patients awaiting lung transplantation. *American Journal of Transplantation*. 2005;5(10):2425-2432.

10. Pumar MI, Roll M, Fung P, et al. Cognitive behavioural therapy (CBT) for patients with chronic lung disease and psychological comorbidities undergoing pulmonary rehabilitation. *J Thorac Dis*. 2019;11:S2238-S2253. http://www.jthoracdis.com/

11. Kalluri M, Claveria F, Haggag M, Richman-Eisenstat J. Early integrated palliative care in a multidisciplinary interstitial lung disease (ILD) collaborative reduces hospitalizations for idiopathic pulmonary fibrosis (IPF). *Chest*. 2016;150(4):472A.

12. de las Heras JC, Balbino F, Catalan-Matamoros D, Lokke A, Hilberg O, Bendstrup E. EFFECT OF A TELEREHABILITATION PROGRAM IN SARCOIDOSIS. *SARCOIDOSIS VASCULITIS AND DIFFUSE LUNG DISEASES*. 2022;39(1).

13. Prajapat B, Menon B, Bansal V, Vijayan V. Effect of mid thigh cross sectional area on CT as a marker of muscle mass in interstitial lung diseases after pulmonary rehabilitation. *Respirology*. 2011;16:325.

14. Thombs BD. Evaluation of the Scleroderma Patient-centered Intervention Network COVID-19 Home-isolation Activities Together Program. Published online 2020. Accessed October 9, 2022. https://clinicaltrials.gov/ct2/show/NCT04335279

15. Lindell KO, Nouraie S, Curtis J, et al. Feasibility and acceptability of an early palliative care intervention in patients with idiopathic pulmonary fibrosis and their caregivers. *Am J Respir Crit Care Med*. 2018;197. http://www.atsjournals.org/doi/abs/10.1164/ajrccm-conference.2018.197.1_MeetingAbstracts.A2646

16. Reilly CC, Bausewein C, Jolley C, et al. Feasibility of a new out-patient breathlessness support service. *Thorax*. 2012;67:A76. http://thorax.bmj.com/content/67/Suppl_2/A76.2.abstract?sid=7c828d23-77e7-42ba-a6bd-fa2a14c22257

17. Khor YH. Handheld fan for breathlessness in interstitial lung disease. Published online 2018. Accessed October 9, 2022. https://trialsearch.who.int/Trial2.aspx?TrialID=ACTRN12618001949279

18. Frith P, Sladek R, Woodman R, et al. Health outcomes in carer-patient dyads of a randomized control trial of carer training for patients receiving long term domiciliary oxygen therapy. *Am J Respir Crit Care Med*. 2013;187. http://www.atsjournals.org/doi/pdf/10.1164/ajrccm-conference.2013.187.1_MeetingAbstracts.A5030

19. Yuen HK, Lowman JD, Oster RA, de Andrade JA. Home-Based Pulmonary Rehabilitation for Patients With Idiopathic Pulmonary Fibrosis: A PILOT STUDY. *J Cardiopulm Rehabil Prev*. 2019;39(4):281-284. http://journals.lww.com/jcrjournal

20. Johannson KA, Vittinghoff E, Morisset J, Lee JS, Balmes JR, Collard HR. Home monitoring improves endpoint efficiency in idiopathic pulmonary fibrosis. *EUROPEAN RESPIRATORY JOURNAL*. 2017;50(1).

21. Wijsenbeek M. Home Monitoring in Idiopathic Pulmonary Fibrosis; Improving Use of Anti-fibrotic Medication and Quality of Life. Published online 2018. Accessed October 9, 2022. https://clinicaltrials.gov/ct2/show/NCT03420235

22. de las Heras JC, Balbino F, Hilberg O, Lokke A, Bendstrup E. Is Virtual Autonomous Physiotherapist Tele-rehabilitation Program feasible in Idiopathic Pulmonary Fibrosis? *EUROPEAN RESPIRATORY JOURNAL*. 2020;56.

23. Sinclair C, Auret KA, Evans SF, et al. Impact of a Nurse-Led Advance Care Planning Intervention on Satisfaction, Health-Related Quality of Life, and Health Care Utilization Among Patients With Severe Respiratory Disease: A Randomized Patient-Preference Trial. *J Pain Symptom Manage*. 2020;59(4):848-855. doi:10.1016/J.JPAINSYMMAN.2019.11.018

24. Mazzoleni S, Montagnani G, Vagheggini G, et al. Interactive videogame as rehabilitation tool of patients with chronic respiratory diseases: Preliminary results of a feasibility study. *Respir Med*. 2014;108(10):1516-1524.

25. Wallaert B, Kyheng M, Labreuche J, Stelianides S, Wemeau L, Grosbois JM. Long-term effects of pulmonary rehabilitation on daily life physical activity of patients with stage IV sarcoidosis: A randomized controlled trial. *Respir Med Res*. 2020;77:1-7. doi:10.1016/j.resmer.2019.10.003

26. Early F, Everden AJT, OBrien CM, Fagan PL, Fuld JP. Patient agenda setting in respiratory outpatients. *Chron Respir Dis*. 2015;12(4):347-356. https://www.scopus.com/inward/record.uri?eid=2-s2.0-84944057171&doi=10.1177%2f1479972315598696&partnerID=40&md5=587e10adae23a21788b1cf28a468c1a8

27. van Manen MJG, van't Spijker A, Tak NC, et al. Patient and partner empowerment programme for idiopathic pulmonary fibrosis. *European Respiratory Journal*. 2017;49(4). doi:10.1183/13993003.01596-2016

28. van Manen M, Van't Spijker A, Tak N, et al. Patient and partner "empowerment" program in idiopathic pulmonary fibrosis (ppepp): Improving quality of life in patients and their partners. *QJM*. 2016;109:S46.

29. Edwards C, Costello E, O'Regan A. Patient-reported Monitoring of Symptoms and Spirometry Via the patientMpower Platform in Idiopathic Pulmonary Fibrosis. Published online 2017. Accessed October 9, 2022. https://clinicaltrials.gov/ct2/show/NCT03104322

30. Mueller K, Kotschy-Lang N, Konig S, Wagner P. Physical activity of patients with occupational lung diseases. *European Respiratory Journal*. 2017;50. http://erj.ersjournals.com/content/50/suppl_61/PA3453

31. Frith P, Sladek R, Woodman R, et al. Pragmatic randomised controlled trial of a personalised intervention for carers of people requiring home oxygen therapy. *Chron Respir Dis*. 2020;17.

32. Holland AE, Hill CJ, Conron M, Munro P, McDonald CF. Short term improvement in exercise capacity and symptoms following exercise training in interstitial lung disease. *Thorax*. 2008;63(6):549-554.

33. Moretta P, Molino A, Martucci M, et al. Subject preferences and psychological implications of portable oxygen concentrator versus compressed oxygen cylinder in chronic lung disease. *Respir Care*. 2021;66(1):33-40. http://rc.rcjournal.com/content/respcare/66/1/33.full.pdf

34. Wong CK, Wong KY, Shum YW, et al. Tele-monitoring of home oxygen user (THOU): A program to ensure maximal therapeutic benefit in patients commencing on long-term oxygen therapy (LTOT). *Respirology*. 2012;17:36.

35. de Las Heras JC, Hilberg O, Lokke A, Bendstrup E. Tele-rehabilitation program in idiopathic pulmonary fibrosis. *European Respiratory Journal*. 2019;54. https://erj.ersjournals.com/content/54/suppl_63/PA2232

09.02 - Physiotherapists

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Effects of self-management interventions in people with interstitial lung disease - systematic review and meta-analysis

Idiopathic pulmonary fibrosis, Quality of life, Chronic diseases

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People with interstitial lung disease (ILD) want to actively manage their condition, however, the effects of self-management interventions (SMIs) in this population have not been synthesized. This review summarizes the effects of SMIs on health-related quality of life (HRQoL), functional status, psychological and social factors, symptoms, exacerbations, healthcare utilization, and survival, in people with ILD.

Six digital databases were searched in May 2022 with monthly updates until February 2023. We included randomized trials implementing SMIs, defined according to Effing et al. [Eur Respir J 2016; 48(1): 46–54], in adults with ILD. Risk of bias and quality of evidence were assessed with the Cochrane RoB-II and the GRADE. Meta-analysis was used to summarize results.

Four studies examining 217 participants (81% men, 71 years old, 91% idiopathic pulmonary fibrosis) with highly heterogenous SMIs were included. No statistically significant differences were observed for HRQoL or any of the secondary measures (figure 1). The quality of evidence ranged from low to very low.

Current studies show that SMIs have no significant effect on people with ILD. This conclusion is limited by high methodological heterogeneity. Studies optimizing SMIs to target the individual needs of people with ILD, and a universal, and consensual definition of SMI are required.

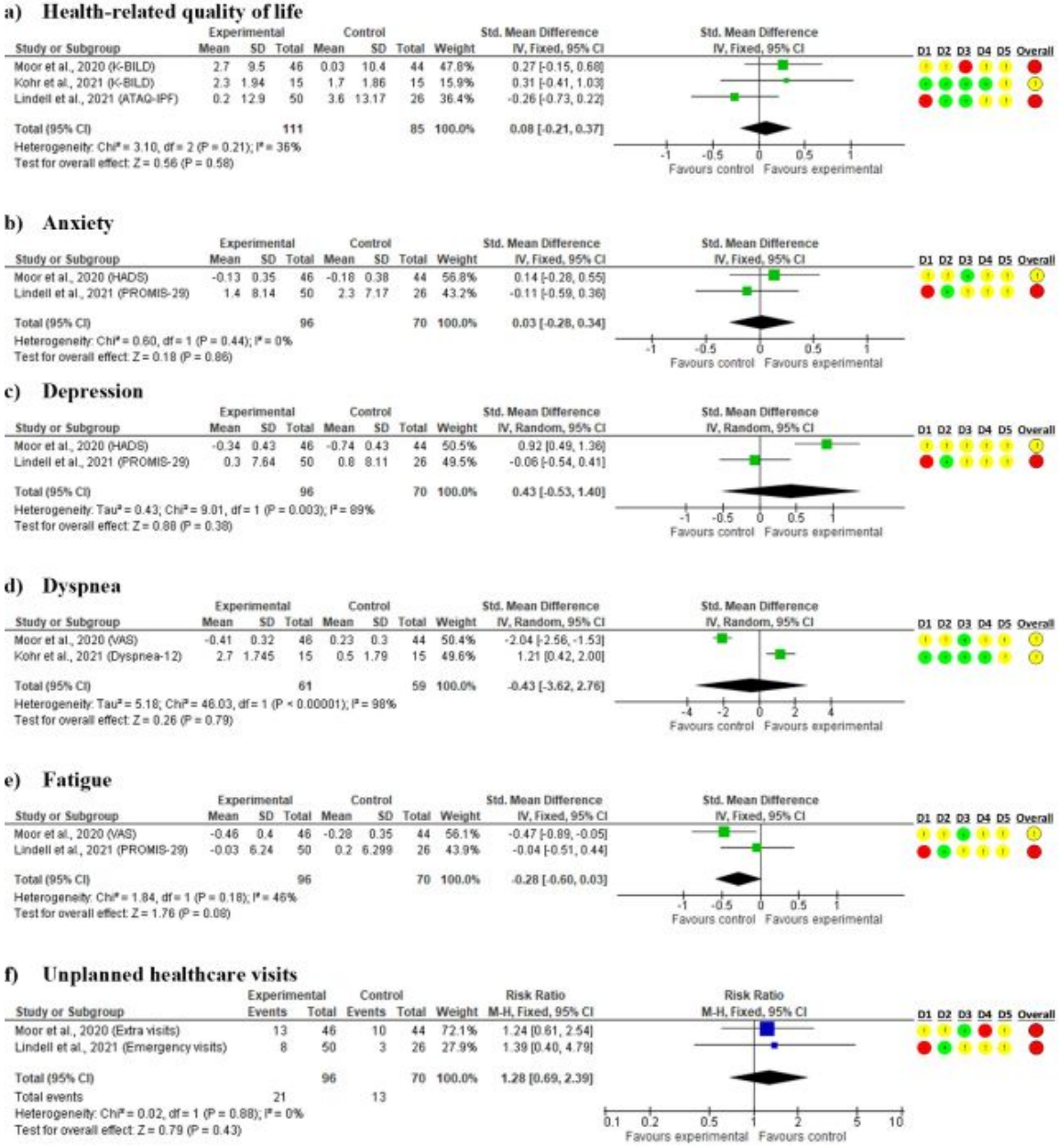

Figure 1 - Meta-analysis on the effects of self-management interventions versus usual or standard care on a) health-related quality of life, b) anxiety c) depression, d) dyspnea, e) fatigue and d) unplanned healthcare visits of people with interstitial lung disease.

Green circle: low risk of bias; **Yellow circle**: some concerns; **Red circle**: high risk of bias.
Abbreviations – **ATAQ-IPF**: A Tool to Assess Quality of Life in Idiopathic Pulmonary Fibrosis; **CI**: Confidence Interval; **D1:** Randomization process; **D2:** Deviations from the intended interventions; **D3:** Missing outcome data; **D4:** Measurement of the outcome; **D5:** Selection of the reported result; **HADS**: Hospital Anxiety and Depression Scale; **IV**: Inverse-Variance; **K-BILD**: King's Brief Interstitial Lung Disease health status questionnaire; **P**: P-value; **PROMIS-29**: Patient Reported Outcome Measurement Information System; **SD**: Standard Deviation; **VAS**: Visual Analog Scale.