Background: Patients with fibrotic interstitial lung diseases (ILD) have a high symptom burden. Improvements in symptom management may decrease patient hospital admissions and improve quality of life (Qol).

Aim: This systematic review aimed to examine the interventions to improve symptom control and quality of life in patients with ILD.

Methods: Synthesis of relevant studies from 3 previous reviews, with updated systematic review and meta-analyses of data from 2011 to November 2017 (nine databases searched).

- Population - Adults with a confirmed diagnosis of a fibrotic ILD (including idiopathic pulmonary fibrosis and non-specific interstitial pneumonia).
- Intervention - Any symptom control or palliative intervention aimed at managing symptoms and improving Qol in ILD.
- Comparator - All comparators considered.
- Outcomes - All symptom control outcomes including cough, breathlessness, Qol and measures of function such as 6MWD.
- Designs - Prospective studies of any design included.

Eligibility and quality were independently assessed by two authors. Risk of bias was assessed using tools from the National Institute for Health, National Heart, Lung and Blood Institute and the Cochrane risk of bias tool.

Results: 52 studies on pharmacological and non-pharmacological interventions published in 57 articles were included of which 7 and 4 were included in the two meta-analyses (figures 1 and 2 respectively). 2,531 participants and 36 different outcome measures were used.

Interventions: 20 of the included studies assessed other non-pharmacological interventions; pulmonary rehabilitation (PR) or exercise training was evaluated by 27 studies. The remaining 5 studies assessed other non-pharmacological interventions.

Study quality: Many studies were uncontrolled, with small sample sizes and unclear quality aspects. Follow-up was often short and few interventions provided evidence that could be meta-analysed.

Pulmonary Rehabilitation (PR): PR was evaluated by 27 studies (11 RCTs, 1 controlled clinical trial, 3 quasi-experimental studies, 1 controlled cohort study, and 11 before-and-after studies). A statistically significant difference between PR and control was found in 6MWD immediately following the intervention (MD 45.93 m, 95% CI 22.59, 68.29, I² 67%, Random effects) Figure 1. At longer-term follow-up (3 or 6 months), there was no significant difference in change in 6MWD between groups (MD 5.26 m, 95% CI -12.88, 23.40, I² 66%, Fixed effects) Figure 2. Data on immediate effect on dyspnoea was contradictory with no effect on long-term follow up. Overall, effect on Qol was inconsistent with most RCTs finding no statistically significant effect on both short and long-term follow up.

FIGURE 1: Meta-analysis of PR vs control for 6MWD immediately following intervention

FIGURE 2: Meta-analysis of PR vs control for 6MWD at longest follow-up


Discussion:
Data on interventions to improve symptoms and Qol in ILD is slowly growing. Our data shows PR achieved short-term improvements in 6MWD without clear evidence of improvements in the severity of exertional dyspnoea or improvement in quality of life. This is in contrast to the Cochrane review* which found improvements in functional exercise capacity, dyspnoea and Qol immediately following PR.
Notably, studies assessing PR used nine different dyspnoea measures. Future use of validated consistent measures may permit meta-regression to identify service and patient characteristics related to better outcome. Future research of the effectiveness of this complex intervention should be supplemented with embedded qualitative methods.

Strengths & limitations: Strengths included a registered protocol, comprehensive searches, and input from multiple stakeholders (researchers, clinicians, service user representatives).

Conclusion:• The body of evidence around interventions to improve symptoms and quality of life in ILD is slowly growing.
• Further research is needed to assess the efficacy of complex interventions like pulmonary rehabilitation in both the short and long-term.
• This body of work strongly highlights the need to build consensus around outcome measures in palliative care ILD research.

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