

Results Summary

Introduction: Research evaluating tailored pulmonary rehabilitation (PRP) for patients with interstitial lung diseases (ILD) are lacking, particularly in a sub group of patients with combined pulmonary fibrosis and emphysema (CPFE). This study aimed to better understand the outcomes of patients with CPFE and evaluate the feasibility of Inspiratory muscle training (IMT) combined with PRP in these patients.

Methods: A feasibility study with randomized controlled trial design was conducted. Patients with ILD were randomized into an intervention group receiving IMT + PRP and a control group PRP only. The PRP consisted of one session/week for 8 weeks. POWERBreathe[®] was used to perform IMT twice daily. Due to the exploratory nature of this study data was predominantly analysed descriptively

Results: This study was able to describe a rare group of patients with CPFE (n=203), managed by a regional centre for ILD in the UK.

Patients with CPFE were relatively old 72 years (SD=8.7), majority male 122 (82%), with history of smoking 144 (95%). Patients with CPFE had a poor prognosis with mortality of 75 (50%) at 3.3 years with a median survival time of 40.1 months. A regional clinic service of PRP for patients with ILD was established.

Thirty-four participants with ILD including 9 patients with CPFE participated in the PRP study. PRP had good attendance and completion rates. No side effects were reported during the study. The feasibility RCT showed improved maximum inspiratory pressure (MIP) in all participants. Improvement were also seen in six-minute walk test (6MWT). Patients gave positive feedback to PRP where they valued the exercise, meeting others with same condition, and the educational sessions. Potential Improvements in Matrix metalloproteinase 7 (MMP7) and sarcopenia after PRP were seen.

Conclusion: This project contributed to the establishment of a clinical PRP program tailored for patients with ILD in the North East of England. The PRP program was feasible and well received by patients, including those with CPFE. Data in this study shows the potential beneficial effects of PRP for these patients where other therapeutic options are limited. Larger sample studies are feasible, supported by patients and are needed in this important area of translational research.