Non-pharmacological therapy

Pulmonary rehabilitation

Pulmonary rehabilitation (PR) programmes involve aerobic conditioning, strength and flexibility training, education, and nutritional and psychosocial support.[1] PR is recommended for the majority of patients with idiopathic pulmonary fibrosis (IPF) but may not be a reasonable choice in a minority of patients with IPF.[1]

Efficacy in IPF: studies have shown that PR improves dyspnoea, quality of life and 6-minute walk distance.[2-5] Studies have demonstrated that PR has more long-lasting effects in patients with less-advanced disease, and, therefore, it is recommended to commence IPF patients on a PR programme at time of diagnosis or before the disease becomes advanced.[6] Additionally, PR is beneficial in lung transplant candidates, particularly for improving the pre- and post-operative outcomes.[7,8]

Efficacy on comorbid conditions: PR has been associated with an improvement in anxiety and depression symptoms.[7]

Oxygen therapy

Long-term oxygen therapy (LTOT) is recommended for patients with IPF who have resting hypoxaemia (resting SpO_2 of <88%).[1]

Efficacy in IPF: currently, there are no trials to demonstrate the efficacy of LTOT in IPF; however, it has been demonstrated previously that resting hypoxaemia is common in IPF patients.[1,7] LTOT may improve quality of life and provide symptomatic relief for patients with IPF.[7] Therefore, LTOT is commonly utilised but should be titrated carefully to prevent potentially harmful hyperoxia.[9]

Conversely, clinical studies investigating ambulatory oxygen therapy have shown a significant benefit of this in patients with IPF versus patients without oxygen therapy.[10,11] These studies demonstrated administration of ambulatory oxygen therapy provided a significant improvement in 6-minute walk distance and dyspnoea.

Lung transplantation

Lung transplantation is recommended for patients with advanced IPF, for whom no effective medical therapy is available.[1] The International Society for Heart and Lung Transplantation[12] defined the criteria for lung transplantation in patients with interstitial lung disease as follows:

Timing of referral	***Timing of listing***
Confirmed usual interstitial pneumonia (UIP) or nonspecific interstitial pneumonia (NSIP), regardless of lung function	FVC decline ≥10% during 6 months of follow-up (note: ≥5% may also warrant listing)
Abnormal lung function: • FVC <80% predicted or DLco <40% predicted	DLco decline ≥15% during 6 months of follow-up
Any dyspnoea or functional limitation attributable to lung disease	Desaturation to <88% or 6MWD <250 m or 6MWD decline >50 m over a 6-month period
Any oxygen requirement, even if only during exertion	Pulmonary hypertension on right heart catheterisation or 2D echocardiography
	Hospitalisation because of respiratory decline, pneumothorax or acute exacerbation

Efficacy in IPF: the primary goal of lung transplantation is to improve survival and previous studies have demonstrated that 5-year survival rates after lung transplantation in IPF are estimated at 45–56% and 22%, respectively.[1,13] These survival rates though have been reported to be significantly poorer than survival associated with other causes of end-stage lung disease.[13] Currently, there is no recommendation concerning single versus bilateral lung transplantation in patients with IPF.[1,14] However, in terms of survival, previous studies have indicated no difference in survival in relation to single versus bilateral lung transplantation.[14]

Summary of the non-pharmacological therapies as recommended in the 2011 guidelines[1]

Treatment	*Recommendation*	*Level of evidence*
Long-term oxygen therapy	Strongly recommended in patients with IPF and clinically significant resting hypoxaemia	Very low-quality evidence; there are no data that directly inform the use of long-term oxygen therapy
Lung transplantation	Strongly recommended in appropriate patients	Low-quality evidence; patient population in retrospective studies include those with other forms of fibrotic lung disease
Pulmonary rehabilitation	Suitable for the majority of patients. May not be reasonable in a minority of patients	Low-quality evidence
Mechanical ventilation	Not recommended in the majority of patients. May be a reasonable choice in a minority of patients	Low-quality evidence

References

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