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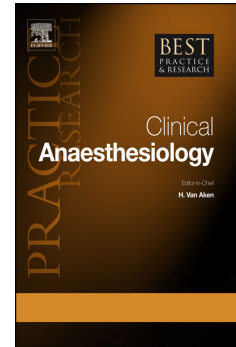
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LONG TERM MANAGEMENT OF PATIENTS WITH END-STAGE LUNG DISEASES

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Abstract

Long term management of end stage lung disease differs from interstitial lung disease to chronic obstructive pulmonary disease to cystic fibrosis to pulmonary vascular disease. The management includes pharmacological therapy that are disease specific such as antibiotic therapy for cystic fibrosis, antifibrotic drugs in idiopathic pulmonary fibrosis, long acting beta-agonists, long acting muscarinic antagonist and inhaled corticosteroids in chronic obstructive pulmonary disease and vasodilators in pulmonary arterial hypertension. Furthermore, non pharmacological therapy is essential in the treatment of these diseases; in particular: rehabilitation and supportive therapy which are necessary in all end stage lung diseases and specific intervention like non invasive ventilation in chronic obstructive pulmonary disease and cystic fibrosis, surgical therapy in chronic obstructive pulmonary disease, airway clearance in cystic fibrosis. The goal is not only to prolong survival but it is fundamental to keep patients in good general conditions for transplantation. Transplantation, indeed, remains the only therapeutic option that could prolong survival in patients with terminal lung disease when the medical or the surgical therapies are not available or not anymore effective.

Idiopathic Pulmonary Fibrosis (IPF)

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