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Review

Cystic Fibrosis and Lung Transplantation: A Review of Contraindications and Clinical Indications

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ABSTRACT

Cystic fibrosis is a severe genetic disease primarily affecting the lungs and the digestive system. As the disease progresses, many patients experience severe respiratory failure, making lung transplantation a vital treatment option. This article reviews the main contraindications and clinical indications for lung transplantation in cystic fibrosis patients, addressing clinical criteria, preoperative evaluations and post-transplantation challenges.

Keywords: Cystic fibrosis; Transplantation; Genetics; Pulmonary

Introduction

Cystic fibrosis (CF) is an autosomal recessive hereditary disease caused by mutations in the CFTR gene (cystic fibrosis transmembrane conductance regulator), which encodes an essential protein for chloride ion transport in epithelial cells[1-3]. Dysfunction of this protein leads to the production of thick and viscous secretions, impairing various organs, especially the lungs and gastrointestinal tract[4]. In the lungs, these secretions promote chronic infections and inflammation, resulting in bronchiectasis, progressive decline in lung function and eventually respiratory failure[5]. Despite therapeutic advances such as CFTR modulators, respiratory physiotherapy and antibiotics, many patients progress to critical stages where lung transplantation becomes the only therapeutic option. Lung transplantation is a complex intervention involving the replacement of compromised lungs with healthy donor organs[6]. However, the procedure carries significant risks, including acute or chronic rejection of the transplanted organ, infections and complications related to immunosuppression[7]. The selection of transplant candidates is a highly rigorous process, considering factors such as age, nutritional status, comorbidities and psychological condition. Moreover, proper management of contraindications is essential to ensure transplant success[8,9].

Objective

This study aims to review the main clinical indications and contraindications for lung transplantation in cystic fibrosis patients, as well as discuss the challenges and advances in the field.

Materials and Methods

A bibliographic search was conducted in databases such as PubMed, Scopus and SciELO, including articles published in the last 10 years, prioritizing systematic reviews, clinical studies and international guidelines.

Discussion

Lung transplantation is considered the gold standard for patients with advanced-stage cystic fibrosis. The main indications include accelerated decline in lung function (FEV1 < 30%), increased frequency of pulmonary exacerbations, pulmonary hypertension and persistent hypoxemia[10]. Additionally, patients unresponsive to conventional treatments or with significantly compromised quality of life are considered candidates. On the other hand, contraindications include active infections by resistant pathogens such as Burkholderia cepacia, which are associated with unfavorable post-transplant outcomes[11]. Other contraindications involve severe comorbidities such as renal or hepatic failure, active cancer, recent oncological diseases and non-adherence to treatment[12,13].

A critical aspect in managing these patients is the multidisciplinary pre-transplant evaluation, which includes pulmonary function analysis, imaging studies, microbiological tests and psychological assessment. Furthermore, post-operative follow-up is essential to prevent complications and improve quality of life[14].

Despite advances in surgical techniques and immunological management, long-term survival after lung transplantation remains limited, primarily due to chronic rejection and obliterative bronchiolitis. Future studies should focus on new biomarkers and immunomodulatory therapies to improve clinical outcomes[15].

Conclusion

Lung transplantation is an essential intervention for patients with advanced cystic fibrosis, providing significant life extension and improved quality of life. However, careful candidate selection and rigorous management of contraindications and complications are indispensable for the procedure's success.

Considerable challenges remain, including chronic rejection and limited organ availability. Collaborative efforts among healthcare professionals, patients and researchers are necessary to advance the understanding and management of cystic fibrosis and lung transplantation. Investments in new therapies organ preservation techniques and personalized immunosuppression hold promise to transform the treatment landscape in the coming years.

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