



## ***Lung Transplantation in Cystic Fibrosis Patients: Clinical Presentations and Surgical Considerations***

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### **LITERATURE REVIEW**

#### **RESUMO**

**Introdução:** O transplante pulmonar tem se consolidado como uma opção vital para pacientes com fibrose cística avançada, uma doença genética que provoca a produção de muco espesso e viscoso, levando a severas complicações respiratórias e comprometimento da função pulmonar. A fibrose cística resulta em infecções pulmonares recorrentes e danos estruturais significativos, tornando o transplante pulmonar uma intervenção necessária quando os tratamentos convencionais já não são suficientes para controlar a progressão da doença. A abordagem cirúrgica neste contexto envolve não apenas a substituição dos pulmões comprometidos, mas também a gestão de complicações específicas e a adaptação do paciente ao novo órgão. **Objetivo:** Avaliar as práticas clínicas e considerações cirúrgicas associadas ao transplante pulmonar em pacientes com fibrose cística, enfocando as apresentações clínicas e os desafios cirúrgicos envolvidos. **Metodologia:** A revisão sistemática foi conduzida seguindo o checklist PRISMA para assegurar a precisão e transparência. As buscas foram realizadas nas bases de dados PubMed, Scielo e Web of Science. Foram utilizados os descritores “fibrose cística”, “transplante pulmonar”, “apresentações clínicas”, “considerações cirúrgicas” e “resultados pós-operatórios”. Os critérios de inclusão foram artigos publicados nos últimos 10 anos, estudos focados em pacientes com fibrose cística e transplante pulmonar, e publicações que descreviam tanto as práticas clínicas quanto os resultados cirúrgicos. Foram excluídos estudos que não apresentavam dados relevantes sobre o transplante em fibrose cística, artigos que abordavam outras doenças pulmonares e revisões ou meta-análises sem dados originais. **Resultados:** Os estudos revisados mostraram que o transplante pulmonar para pacientes com fibrose cística é associado a melhorias significativas na função pulmonar e na qualidade de vida. No entanto, a complexidade do procedimento cirúrgico e a necessidade de manejo pós-operatório intensivo foram destacados como fatores críticos. A abordagem cirúrgica exige um planejamento meticuloso para evitar complicações como rejeição do enxerto



e infecções. A pré-operatória avaliação detalhada e o acompanhamento rigoroso foram identificados como essenciais para o sucesso a longo prazo do transplante. Conclusão: O transplante pulmonar representa uma opção eficaz para pacientes com fibrose cística que alcançaram a fase avançada da doença pulmonar. As práticas clínicas e cirúrgicas devem ser cuidadosamente adaptadas às necessidades específicas desses pacientes para otimizar os resultados e minimizar as complicações. A integração de estratégias robustas de avaliação e acompanhamento tem mostrado ser fundamental para o sucesso do transplante e para a melhoria substancial da qualidade de vida dos pacientes.

**Palavras-chaves:** “fibrose cística”, “transplante pulmonar”, “apresentações clínicas”, “considerações cirúrgicas” e “resultados pós-operatórios”.

#### **ABSTRACT**

**Introduction:** Lung transplantation has become a critical option for patients with advanced cystic fibrosis, a genetic disorder that causes the production of thick and sticky mucus, leading to severe respiratory complications and impaired lung function. Cystic fibrosis results in recurrent lung infections and significant structural damage, making lung transplantation necessary when conventional treatments are no longer sufficient to manage disease progression. In this context, the surgical approach involves not only the replacement of compromised lungs but also the management of specific complications and the adaptation of the patient to the new organ. **Objective:** To evaluate clinical practices and surgical considerations associated with lung transplantation in patients with cystic fibrosis, focusing on clinical presentations and surgical challenges. **Methodology:** The systematic review was conducted following the PRISMA checklist to ensure accuracy and transparency. Searches were performed in the PubMed, Scielo, and Web of Science databases. The descriptors used were "cystic fibrosis," "lung transplantation," "clinical presentations," "surgical considerations," and "postoperative outcomes." **Inclusion criteria** were articles published in the last 10 years, studies focused on patients with cystic fibrosis and lung transplantation, and publications describing both clinical practices and surgical outcomes. **Exclusion criteria** included studies without relevant data on cystic fibrosis lung transplantation, articles addressing other pulmonary diseases, and reviews or meta-analyses without original data. **Results:** The reviewed studies indicated that lung transplantation for cystic fibrosis patients is associated with significant improvements in lung function and quality of life. However, the complexity of the surgical procedure and the need for intensive postoperative management were highlighted as critical factors. The surgical approach requires meticulous planning to avoid complications such as graft rejection and infections. Detailed preoperative assessment and rigorous follow-up were identified as essential for long-term transplant success. **Conclusion:** Lung transplantation represents an effective option for patients with cystic fibrosis who have reached the advanced stage of pulmonary disease. Clinical and surgical practices must be carefully adapted to the specific needs of these patients to optimize outcomes and minimize complications. The integration of robust assessment and follow-up strategies has proven fundamental for transplant success and substantial improvement in patient quality of life.



**Keywords:** "cystic fibrosis," "lung transplantation," "clinical presentations," "surgical considerations," and "postoperative outcomes."

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## **INTRODUCTION:**

Lung transplantation is established as a crucial intervention for patients with advanced cystic fibrosis, a genetic condition that causes the production of thick and sticky mucus. This mucus accumulates in the lungs, leading to severe respiratory complications and progressive deterioration of lung function. Cystic fibrosis results in chronic pulmonary infections, inflammation, and structural damage to lung tissues, making conventional treatment often insufficient when the disease reaches advanced stages. In this context, lung transplantation emerges as the definitive solution, offering a new opportunity for respiratory function recovery and quality of life improvement.

The surgical approach in lung transplantation for cystic fibrosis patients is complex and requires careful preparation. The procedure is not limited to the replacement of compromised lungs but also involves a series of critical considerations to ensure the success of the transplant. The complexity of the surgery is largely related to managing the patient's specific conditions, which may include chronic pulmonary infections and anatomical changes due to cystic fibrosis. Additionally, transplantation requires precise coordination among different medical teams and detailed planning to avoid complications such as graft rejection and postoperative infections. Meticulous preparation and precise execution of the procedure are crucial for achieving the best possible outcomes and minimizing the risks associated with surgery.

Lung transplantation for cystic fibrosis patients faces significant challenges related to adaptation and postoperative management. Post-transplant care is a critical aspect of the process, requiring constant vigilance to ensure the integrity of the new organ and the patient's complete recovery. After the transplant, patients need rigorous follow-up to monitor potential complications such as graft rejection and infections, which can occur due to the immune system being suppressed to prevent rejection. Additionally, monitoring lung function levels and managing potential side effects of immunosuppressive medication are essential for the long-term success of the transplant.

The challenges related to patient adaptation to the new organ are also substantial. The integration of the transplant involves complex adjustments in



lifestyle and medication therapies. Patients often need to modify their routines to accommodate immunosuppressive treatment and undergo a series of regular tests and consultations. The psychological and emotional impact of the transplant is considerable, and psychological support is frequently necessary to help patients cope with changes in health and the new dynamics of life.

Moreover, long-term outcome evaluation is crucial to measure the effectiveness of the transplant and its impact on patients' quality of life. Studies show that, despite significant improvements in lung function and overall quality of life, patients still face ongoing risks of complications and require prolonged follow-up. Detailed analysis of postoperative results and implementation of strategies for continuous management are essential to maximize the benefits of the transplant and provide a healthy and productive life for patients.

Thus, the success of lung transplantation in cystic fibrosis patients depends on a comprehensive approach that includes intensive postoperative care, patient adaptation, and ongoing evaluation of outcomes. Effective management of these aspects is crucial to optimize results and ensure the quality of life for patients after the transplant.

## **METHODOLOGY**

The methodology for the systematic review was conducted following the PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) checklist to ensure rigor and transparency in the process. The databases consulted were PubMed, Scielo, and Web of Science, chosen for their comprehensiveness and relevance in the fields of health and biomedical sciences. The search descriptors used were “cystic fibrosis,” “lung transplantation,” “clinical presentations,” “surgical considerations,” and “postoperative outcomes.”

The initial search in the databases was performed using combinations of the mentioned descriptors, employing Boolean operators to refine the results. The selection of articles was conducted based on a series of inclusion and exclusion criteria to ensure the relevance and quality of the studies included in the review.

The inclusion criteria were as follows: Articles published in the last 10 years, ensuring that the information was current and relevant to contemporary



clinical practice. Studies focused exclusively on patients with cystic fibrosis who underwent lung transplantation, to ensure the specificity of the topic. Works providing detailed data on clinical practices, surgical considerations, or postoperative outcomes, allowing for a comprehensive analysis of approaches and results. Studies presenting original data and not reviews or meta-analyses without new data, ensuring the inclusion of primary evidence. Publications in English, Spanish, and Portuguese to maximize the inclusion of relevant and accessible studies.

The exclusion criteria were established to filter out non-relevant or low-quality information: Articles that did not specifically address lung transplantation in patients with cystic fibrosis, to ensure content relevance. Studies with inadequate or inaccurate data on postoperative management or transplantation outcomes, to maintain information integrity. Publications that did not provide complete information on treatment protocols or postoperative results, limiting the capacity for analysis. Works addressing other pulmonary conditions not related to cystic fibrosis, to maintain focus on the central topic of the review. Studies published before the last 10 years or not meeting the defined quality criteria, to ensure the timeliness and relevance of the evidence.

After applying these criteria, the selected articles were reviewed in detail, and data were extracted for analysis. The methodology ensured a rigorous and systematic approach to the review, ensuring that the results were based on current and pertinent evidence.

## **RESULTS**

Lung transplantation is a crucial solution for patients with cystic fibrosis in advanced stages of the disease, when conventional treatments become ineffective. This genetic condition results in the production of thick and viscous mucus, leading to severe and progressive respiratory complications. When conservative therapeutic methods, such as medications and respiratory therapies, fail to control the deterioration of lung function, transplantation becomes a necessary alternative to ensure survival and improve quality of life. The decision to perform a transplant is generally based on a detailed evaluation of the patient's clinical condition, considering the severity of respiratory



symptoms, the frequency of pulmonary infections, and the overall progression of the disease.

The evaluation for lung transplantation involves analyzing various clinical parameters. Lung function is assessed through spirometry, which measures residual respiratory capacity and the degree of impairment. The presence of comorbidities, such as diabetes or hypertension, is also examined, as it can affect both the success of the procedure and the postoperative prognosis. The medical team uses this information to determine if the patient meets the eligibility criteria and if the benefits of transplantation outweigh the risks associated with the surgery and postoperative period. Thus, the decision is based on a comprehensive analysis, ensuring that the treatment is suited to the specific needs of each patient.

The selection process for lung transplantation is meticulous and requires consideration of multiple factors. Initially, the severity of cystic fibrosis is assessed through clinical and functional examinations, including lung function tests and imaging studies. Criteria include the presence of chronic and irreversible respiratory failure, identified by a significant reduction in vital capacity and forced expiratory volume in one second. These parameters are crucial to ensure that the surgery is the appropriate solution and that the patient has a real chance of substantially improving lung function after the transplant.

Additionally, selection considers the presence of other medical conditions that may impact the outcome of the transplant. Comorbidities, such as cardiovascular or renal diseases, can complicate both the surgery and recovery, making the screening process complex. The transplant team performs a multidisciplinary evaluation to ensure that all aspects of the patient's health are considered and that the patient is prepared to face postoperative challenges. This comprehensive process aims to maximize the effectiveness of the transplant and minimize risks, ensuring that the patient benefits from the procedure in the long term.

The challenges associated with postoperative management of lung transplantation in cystic fibrosis patients are substantial and require rigorous monitoring. After the surgery, constant vigilance is crucial to detect and manage potential complications. The risk of graft rejection is a major concern, as the





recipient's immune system is intentionally suppressed to prevent rejection. This immunosuppression, while essential, makes the patient more susceptible to infections and other immune-related complications. Therefore, the administration of immunosuppressive medications must be meticulously monitored and adjusted to balance rejection prevention with minimizing adverse effects.

Moreover, post-transplant follow-up includes regular monitoring of lung function. Frequent exams, such as spirometry and imaging studies, are essential to assess the integrity of the new organ and to detect early signs of dysfunction or rejection. Managing the side effects of immunosuppressive therapies, which may include hypertension, diabetes, and renal problems, is also crucial for maintaining the patient's overall health. Effective coordination between healthcare professionals and the patient is essential to tailor the treatment to individual needs and ensure the best possible recovery.

Adapting to the new organ represents a significant challenge for patients. The process of integrating a lung transplant requires substantial changes to daily routines and medication regimens. Patients need to adjust their lifestyle habits to accommodate the new treatment and monitoring requirements, including changes in diet, increased medical visits, and a rigorous medication regimen to prevent complications. The psychological impact of these changes can be significant, with patients facing stress and anxiety related to new health responsibilities and the fear of graft rejection.

Additionally, psychological support is often needed to help patients cope with changes in their health and daily life. Adapting to a new lifestyle requires not only physical but also emotional adjustments. Psychological support programs and support groups play a crucial role, providing patients with the necessary help to face the emotional and psychological challenges resulting from the transplant. This support can ensure a smoother transition and improve overall quality of life after the procedure.

Long-term outcome assessment is essential to determine the effectiveness of lung transplantation and its impact on patients' quality of life. Ongoing analysis of transplant success goes beyond simply monitoring lung function, also encompassing the evaluation of the patient's overall health conditions and quality of life. Studies show that despite significant improvements





in respiratory function and reduction in cystic fibrosis symptoms, patients still face ongoing risks and challenges related to the transplant. Meticulous monitoring of postoperative outcomes is crucial for early identification of any complications and for adjusting interventions as needed.

Long-term success indicators include the stability and function of the graft, control of associated comorbidities, and the overall functional capacity of the patient. Regular evaluation of lung function parameters, such as forced expiratory volume and vital capacity, provides a clear view of the new organ's health. The frequency of respiratory infections and graft rejection should be closely monitored, as these factors can influence patient survival and quality of life. Continuous follow-up is essential for early detection of problems and implementation of effective management strategies, ensuring the longevity and health of the transplant.

The impact of lung transplantation on the quality of life of cystic fibrosis patients is well-documented. After the surgery, many patients report a significant improvement in respiratory capacity and a substantial reduction in cystic fibrosis symptoms, such as frequent pulmonary infections and breathing difficulties. Post-transplant recovery often results in increased functional capacity and autonomy, allowing patients to resume daily activities and improve their quality of life. However, this positive transformation is not universal, and some patients continue to face challenges that may impact their overall sense of well-being.

Furthermore, psychological and social adaptation after the transplant plays a crucial role in patients' quality of life. The transition to a new routine, which includes continuous monitoring of lung function and administration of immunosuppressive medications, can be challenging. Therefore, the need for psychological and social support becomes evident. Rehabilitation programs and psychological support are often recommended to help patients manage changes in their health and cope with transplant-related stress. Continuous assistance in these areas can significantly contribute to patients' satisfaction and overall well-being after the surgery.

In the long term, managing complications and ongoing monitoring of patients' health status are fundamental to ensuring transplant effectiveness and maintaining quality of life. Regular follow-up helps detect and treat potential



complications, such as graft rejection and infections, which can compromise transplant function and overall patient health. Thus, integrating rigorous monitoring strategies and continuous support tailored to each patient's individual needs is essential to maximize the benefits of lung transplantation.

Additionally, analyzing the economic impact of the transplant is relevant. The cost of lung transplantation and postoperative treatment can be high, and analyzing costs and benefits is essential for resource allocation and clinical and administrative decision-making. Studies addressing the economic impact of transplantation provide a comprehensive view of the resources required and the financial implications for patients and the healthcare system. Understanding these aspects can help optimize treatment management and improve access to transplantation for those in greatest need.

The development of new adjunctive therapies in the context of lung transplantation for cystic fibrosis patients is an expanding research field aimed at enhancing clinical outcomes and patients' quality of life. Emerging therapeutic approaches often focus on improving the effectiveness of immunosuppressive treatments and reducing associated side effects. Adjunctive therapies may include new pharmacological agents, advanced monitoring techniques, and integrative strategies that address not only graft rejection but also secondary complications, such as infections and organ dysfunctions. Such innovations seek to provide a more effective balance between rejection prevention and preservation of the patient's immune function, resulting in more precise and personalized management.

Additionally, advances in cystic fibrosis research and the development of new targeted treatments are fundamental to improving post-transplant outcomes. Identifying specific biomarkers and implementing genetic therapies are promising areas with the potential to transform disease management. Incorporating new scientific discoveries into treatment protocols can optimize transplant responses and minimize complications. Ongoing research advancements offer promising prospects for personalized treatment and promoting a more effective and sustainable recovery for patients.

The role of psychosocial factors in post-transplant recovery also deserves detailed attention. Adapting to life after lung transplantation involves significant



changes not only in physical health but also in the emotional and psychological state of patients. Studies demonstrate that psychological support, including therapy and counseling, is crucial for helping patients cope with the stress and anxiety associated with the recovery process and lifestyle changes. Adequate psychological follow-up contributes to treatment adherence and the patient's ability to handle daily demands, positively impacting their quality of life.

Moreover, integrating social and family support strategies plays an important role in patients' recovery and well-being. Having a solid support network can assist in adapting to the new treatment regimen and managing the complexities associated with transplantation. Active involvement from family and friends is often associated with quicker recovery and better psychological adjustment, highlighting the importance of comprehensive support that extends beyond direct medical care. Collaboration between healthcare professionals, patients, and their families is essential to ensure a successful and sustainable recovery process.

## **CONCLUSION**

The analysis of the impact of lung transplantation in cystic fibrosis patients revealed significant conclusions regarding the effectiveness of the procedure and its implications for patients' quality of life. Scientific studies demonstrated that lung transplantation had become a crucial solution for patients with advanced cystic fibrosis, especially when conventional treatments proved ineffective. The genetic condition led to excessive production of thick mucus, resulting in severe and progressive respiratory complications. The transplantation emerged as a necessary alternative to ensure survival and improve quality of life, particularly when other therapeutic options had failed.

According to the scientific literature, the decision to proceed with transplantation was based on a detailed assessment of the patients' clinical status, taking into account the severity of respiratory symptoms, the frequency of pulmonary infections, and disease progression. The decision to perform the surgery was supported by evidence that deteriorated lung function was no longer manageable by conventional methods, such as medications and respiratory therapies.



Postoperative results indicated a significant improvement in respiratory capacity and a reduction in cystic fibrosis symptoms, including the frequency of pulmonary infections and breathing difficulties. Patients frequently reported increased functional capacity and autonomy after the surgery, allowing them to resume daily activities and enhance their quality of life. However, challenges persisted, with some patients encountering issues related to the new organ and the need for continuous adaptation to the treatment.

Rigorous follow-up and careful management of immunosuppressive medications were crucial for the long-term success of the transplantation. The literature highlighted that the risk of graft rejection and immunological complications, such as infections, were predominant concerns requiring constant monitoring. Managing side effects like hypertension, diabetes, and renal issues was also essential for maintaining patients' overall health.

Moreover, psychological and social adaptation played a significant role in post-transplant recovery. Patients faced considerable emotional and psychological challenges as they adjusted to new daily routines and health responsibilities. Psychological and social support proved essential in helping patients cope with these changes and facilitating a smoother transition.

Economically, the analysis indicated that the cost of transplantation and postoperative care was high, but the benefits in terms of quality of life often outweighed the costs. Economic studies suggested that resource allocation should consider not only financial costs but also health and quality of life benefits.

In summary, the studies concluded that despite the complexities and challenges associated with lung transplantation, the procedure brought substantial improvements for most patients with advanced cystic fibrosis. The success of transplantation depended on an integrated approach involving careful medication management, ongoing follow-up, and psychological support. Ongoing advancements in therapies and research on cystic fibrosis promise to further optimize future outcomes for these patients.

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