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An Editorial on Pulmonary Cystic Fibrosis Exacerbation

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Editorial

The clinical spectrum of cystic fibrosis (CF) continues to expand as patient survival improves and extremely effective cystic fibrosis transmembrane conductance regulator medication becomes available. Acute pulmonary exacerbation is one of the most important clinical episodes in the course of CF for patients. Clinical and microbiological epidemiological investigations of CF pulmonary exacerbations continue to shed light on the disease's progression, prognosis, and consequences. This finding has sparked a slew of large-scale clinical trials aimed at altering the therapy paradigm for CF pulmonary exacerbation. The major purpose of this study is to provide a concise overview of CF pathogenesis, clinical epidemiology, microbiological epidemiology, outcome, and treatment.

Although better understanding of the natural course of cystic fibrosis (CF) has led to treatment techniques that have improved pulmonary health and extended the lives of people with the condition, lung disease continues to be the leading cause of morbidity and mortality in CF patients. Dehydrated mucus, poor mucus clearance, and mucus adherence to airway surfaces are thought to be caused by airway epithelial abnormalities in ions-water transport. The production of endobronchial mucus plaques and plugs, which become the principal sites of air flow restriction, infection, and inflammation, leading to early small airways illness and the development of bronchiectasis, suggests an increase in mucin secretion. Lung involvement is frequently gradual, with exacerbations every now and then. Although aggressive management and breakthroughs in treatment slow the progression of lung disease, they do not prevent it.

Respiratory failure develops as a result, and is the leading cause of mortality. For the majority of the disease's duration, the lung parenchyma remains relatively unaffected. In cystic fibrosis, a chronic pulmonary infection is a defining feature of lung disease. Infections caused by the *Burkholderia cepacia* complex, which consists of atleast 18 closely related gram-negative bacteria, are extremely difficult to cure. It's possible that these infections are linked to a fulminant necrotizing pneumonia. Many popular antibiotics are resistant to *Burkholderia cepacia* complex bacteria, and they can acquire resistance to many more. The more severe epidemic strains are not as common after patient segregation in cystic fibrosis medical care, and new infections are less common.

Although there are evidence-based guidelines for treating *Pseudomonas aeruginosa* respiratory exacerbations, these cannot be applied to *Burkholderia cepacia* complex infections. The natural course of cystic fibrosis lung illness is one of chronic progression punctuated by episodes of abrupt symptom worsening known as acute pulmonary exacerbations. These flare-ups almost always necessitate medical attention. Appropriate therapy must be recommended based on the best available evidence of efficacy and safety. As a result, the Cystic Fibrosis Foundation formed a committee to define critical questions about pulmonary exacerbations, examine clinical evidence using an evidence-based methodology, and make recommendations to doctors. It is intended that these guidelines would aid professionals in the care of cystic fibrosis patients.

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