CF is a genetic disorder that occurs due to mutations affecting the Cystic fibrosis transmembrane conductance regulator gene. This affects subsequent chloride transport. This may result in a pulmonary decline or complications. Certain modulators have arisen like that of elexacaftor-tezacaftor-ivacaftor (ETI), and immunosuppressive therapies – however, despite these advancements lung transplant remains the standard of care.

A literature review was conducted synthesizing findings from peer reviewed studies and clinical data evaluating both lung transplant and modulators. Key subsections identified include timing, survival, and quality of life. The impacts of modulators, timing, center expertise, and associated outcomes were compared. These findings establish lung transplant (LTx) as the most effective intervention.

Scientific studies indicate modulators can have a positive impact on outcomes. Center expertise has been found to impact clinical outcomes related to forced expiratory volume (FEV1) and mechanical ventilation (MV). Timing — particularly the point at which a patient is referred or listed for transplant — has been found to play a vital role in lung transplant outcomes as well. Although CFTR modulators may delay transplant, given irreversible damage, lung transplant remains the best option. CF modulators cannot reverse the advanced stages of lung disease. Given this, transplant is the only long-term living option. Major predictors of lung transplant outcomes are grouped into center expertise, timing of transplant, and evaluation of long-term survival — i.e., whether a patient with CF undergoes a lung transplant or is able to forego one over a defined number of years. Today, lung transplant remains the superior treatment option.