

Cystic fibrosis (CF) is a rare genetic disease that causes lung damage and early death. Lung transplantation is a treatment option that prolongs life for people with end-stage CF. Due to the scarcity of donor lungs, transplant candidates need to be carefully selected so that their long-term survival is maximized. The timing of transplantation is also crucial for survival. Accurate prediction of who has an increased risk of death can be an aid in making decisions about who should be prioritized in getting the transplant and what the optimal timing is. I conducted a comprehensive literature search to identify 16 studies that have developed mortality prediction models for CF. These studies developed different prediction models with varying sets of variables and employed a variety of variable selection methods. A seminal paper by Kerem, et al., published in 1992, showed that FEV1 (i.e., forced expiratory volume in one second, a measure of lung function) predicts mortality in individuals with CF. This result has been used for establishing consensus guidelines for selecting lung transplant candidates, which are still in use today. The studies that followed Kerem, et al., have yet to be applied clinically. To evaluate the studies, I compared them against the Transparent Reporting of a multivariable prediction model for Individual Prognosis Or Diagnosis (TRIPOD) Statement checklist. Overall, reporting transparency was poor across the 16 studies, especially for model performance metrics. These 16 studies also have varying outcomes, sample sizes, age, population, and data sources. These factors make direct comparison between the studies challenging and preclude the possibility of identifying an optimal prediction model. Therefore, future analyses will be necessary for comparing the models and for finding an optimal model. By fitting the models on the Canadian CF registry dataset and clearly assessing them using TRIPOD, I will provide a systematic summary of the models and statistical tools available to develop prediction models for people with CF. This work will inform decision-makers to help identify individuals who would benefit from transplant referrals.