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Cystic Fibrosis Research News

Title:

A 3-YEAR PROGNOSTIC SCORE FOR ADULTS WITH CYSTIC FIBROSIS

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What was your research question?

Our goal was to re-assess factors in people with cystic fibrosis (CF) which predict the likely course of the disease and to build a prognostic (predictive) score that will help to estimate the risk of death or lung transplantation (LT) within a 3-year period.

Why is this important?

Life expectancy for people with CF has dramatically improved over past decades due to advances in integrated care provided by multidisciplinary teams in CF centers. As a result, prognostic factors have evolved over time and studies performed using data obtained previously may not be appropriate for any current evaluation of people with CF. Moreover, death in children with CF has almost disappeared in developed countries due to improvements in patient care by multidisciplinary teams.

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What did you do?

Using data from the French CF Registry, we developed a model to predict death or LT in adults with CF. Predictors were combined into a prognostic score that can be used in daily practice to estimate the individual risk of death or LT within 3 years.

What did you find?

We analyzed data from 2,096 adults of whom 268 died or received LT. The predictive model identified eight risk factors associated with death or LT: forced expiratory volume in one second (FEV_1), body mass index (BMI), *Burkholderia cepacia* complex colonization, oral corticosteroids, long-term oxygen therapy, non-invasive ventilation, the number of intravenous antibiotics courses per year and the number of days of hospitalization per year. The prognostic score was calculated from these eight predictors and allowed patients to be given a low, moderate or high risk of death or LT. Importantly, this prognostic score which was based on a range of indicators allowed a better prediction of the risk of death or LT than just using a value of FEV_1 below 30%.

What does this mean and reasons for caution?

The score we developed was able to distinguish individuals who experienced death or LT from those who did not in 91% of cases. The prognostic score is made up of eight factors including three directly related to the patient's clinical characteristics (FEV_1, BMI, *Burkholderia cepacia* complex colonization) and five therapeutic factors. This score could be a useful tool in identifying patients requiring an evaluation for LT.

What's next?

We plan to develop a model which provides dynamic predictions of death and LT separately, taking into account how FEV_1 changes over time. Moreover, this model would identify different profiles of how CF might progress.

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