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CYSTIC FIBROSIS FOUNDATION
ADDING TOMORROWS

Cystic Fibrosis Research News

Citation:

Taylor C, Commander CW, Collaco JM, Strug LJ, Li W, Wright FA, Webel AD, Pace RG, Stonebraker JR, Naughton K, Dorfman R, Sandford A, Blackman SM, Berthiaume Y, Paré P, Drumm ML, Zielenski J, Durie P, Cutting GR, Knowles MR, Corey M. A Novel Lung Disease Phenotype Adjusted for Mortality Attrition for Cystic Fibrosis Genetic Modifier Studies. Pediatr Pulmonol. 2011 September; 46(9): 857–869.

What was your research question? (50 words maximum)

Current genetic studies of lung disease in cystic fibrosis patients lack a severity measure that accounts for disease progression and loss of life. We wanted to develop a new measure that ranged from “mild” to “severe” and could be used in both pediatric patients and older adults.

Why is this important? (100 words maximum)

This new measure of lung disease severity will allow researchers to compare patients across a broad age-range and include those that have passed away.

What did you do? (100 words maximum)

We studied hundreds of CF patients from several large genetic studies in CF in the U.S. and Canada. Using this information, we developed a new framework for measuring lung disease severity that could for patients of any age.

What did you find? (100 words maximum)

We found that our new measure correlated well with previously developed methods. However, the new measure worked better for very young and very old patients.

What does this mean and reasons for caution? (100 words maximum)

This new measure of disease severity has increased our ability to make strong research comparisons without the need for studying long term data. There are some limitations to this measure as it is based on data collected between 1994 and 2001. It is known that lung function numbers have improved since that time.

What’s next? (50 words maximum)

This new measure of lung disease severity will allow better studies that are focused on developing new treatments for CF lung disease.