



# Cystic Fibrosis Research News

## **Citation:**

A semiparametric approach to estimate rapid lung function decline in cystic fibrosis. RD Szczesniak, GL McPhail, LL Duan, M Macaluso, RS Amin, JP Clancy. *Annals of epidemiology* 23 (12), 771-777

## **What was your research question? (50 words)**

We wanted to understand how calculating the timing and degree of lung function decline differs using different statistical methods. We applied traditional and new statistical methods to answer this question.

## **Why is this important? (100 words)**

Decline in lung function happens at an irregular pace for individuals with cystic fibrosis (CF), and more rapid decline is associated with increased mortality. Understanding progression of CF lung disease through lung function decline may lead to more timely treatments for patients and improve survival.

## **What did you do? (100 words)**

We performed advanced statistical analysis using data from the Cystic Fibrosis Foundation Patient Registry to determine factors associated with rapid lung function decline. We compared our results to the results found using traditional statistical methods. We considered key influences on patients' health that have been identified in other health research studies, including age, gender, CF-related diabetes, infections, and socioeconomic status.

## **What did you find? (100 words)**

We found that pace of lung function decline slows between ages 18-35. Lung infections, lower socioeconomic status, being female, and having CF-related



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diabetes were associated with more rapid lung function decline. Unlike traditional models, our statistical model found that lung function decline is not linear over time, regardless of age (that is, lung function does not decline at a steady pace). Both models show similar rate of lung function decline until adolescence. Our model depicts more rapid decline in lung function into early adulthood and average age of most rapid decline around 18 years. Rate of decline slowly continues until around 35 years old, when it continues at a weaker rate.

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

Accurately predicting periods of rapid lung function decline is key to providing early treatments that are important to patients' health and improving outcomes. Further studies are needed to understand additional influences, including smoking, nutritional status, genetic mutations, and effectiveness of treatments on lung function decline.

## **What's next? (50 words)**

We plan to implement our new statistical model of lung function decline within CF centers through an online app. The app will enable clinicians to use this information to enhance monitoring of patient-specific lung-function decline at their centers.