



Cystic Fibrosis Research News

Citation: Adler FR, Liou TG. The Dynamics of Disease Progression in Cystic Fibrosis. PLOS ONE. 2016;11(6):e0156752. doi:10.1371/journal.pone.0156752.

What was your research question? (50 words maximum)

How do lung function (FEV₁%) and lung infections in CF interact and affect patient health and survival?

Why is this important? (100 words maximum)

CF is a progressive disease in which lung function declines over time. This decline is measured using FEV₁%. Declines are largely due to bacterial infections in the lungs and the body's inflammatory response to infections. Knowing how these infections affect lung function and ultimately survival provides valuable information to people with CF and their care teams.

What did you do? (100 words maximum)

Using data from the Cystic Fibrosis Foundation Patient Registry, we created different models to describe how lung function and three key infections in CF interact with each other. These common infections were *Pseudomonas aeruginosa* (*P. aeruginosa*), *Burkholderia* complex, and methicillin-sensitive *Staphylococcus aureus* (MSSA). We were able to use the models to predict patient survival.

What did you find? (100 words maximum)

We found that changes in lung function and the presence or absence of certain bacterial infections have a direct impact on survival. Specifically, infections of *P. aeruginosa* or *B. cepacia* can lead to rapid decline in lung function and having a lower lung function can make it harder to rid the lungs of this bacteria. In



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In addition, it was discovered that the presence of MSSA alone related to higher lung function levels.

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

Survival is affected by lung function, the presence or absence of *S. aureus*, *P. aeruginosa* and *B. cepacia*, and the interactions between these bacterial infections. Eradication of *P. aeruginosa* and *B. cepacia* is beneficial.

Though this information is promising for predicting survival for people with CF related to different bacteria, there are many factors that affect survival. These include: CF mutation, pancreatic sufficiency (whether the pancreas works or does not), CF-related diabetes, and weight. There are also errors in lung function measurement and sputum culture information. We will extend our methods to take these into account.

What's next? (50 words maximum)

Further studies of the interactions between different bacteria and their effect on lung function and survival are planned. In this study we excluded patients who had received lung transplants. We aim to extend the models to include the transplanted population as well.