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Cystic Fibrosis Research News

Title:

Rapid lung function decline in adults with early-stage cystic fibrosis lung disease

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What was your research question?

We wanted to see how many adults with cystic fibrosis are living with early-stage lung disease (defined as lung function >= 80% predicted). Then, amongst these people, investigate the potential signs of a future substantial decrease in lung function. by five percent or more per year.

Why is this important?

More people with CF are progressing to adulthood with lung function of 80% predicted or greater (i.e. the early stages of CF lung disease). In thd future, we expect even more individuals living with CF to have the early stage lung disease due to advancements in medications, increased screening for disease, and earlier treatment for infection. Because most research has focused on individuals with worse lung function, there is not much research available about adults with early-stage CF lung disease. It is important to learn more about early-stage disease so that we better understand risk factors for sudden disease progression and best ways to handle care for these individuals.

What did you do?

We looked at people in the Cystic Fibrosis Foundation Patient Registry (CFFPR) aged 18 and over between 2010-2013 with a predicted lung function of 80 percent and above. We excluded people with organ transplants or who had missing measurements for weight or lung capacity measurements at the time of first lung function value recorded. We split the study population into groups by age, and we developed a mathematical model to estimate lung

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function decline. We then evaluated the variability in lung function and airway obstruction as risk factors to see if they appeared to be associated with substantial lung function decline in the future.

What did you find?

Among adults with early-stage lung disease, about fifteen percent were shown to have a large decrease in lung function. Variability in predicted lung function and blockages in the airway were shown to indicate people at risk for large decreases in lung function.

What does this mean and reasons for caution?

These finding supports the idea that decreases in lung function can be delayed but not avoided entirely in this group of people. We determined that adults with early-stage lung disease should continue to be observed closely in clinic to watch for the beginning of decline in lung function.

What's next?

It is important that we continue to learn more about the community of people with cystic fibrosis who have early stage lung disease. One next step could include studies looking at the risk factors found in this study in another CF population outside the US to support these findings. A second step is to understand what can be done to keep patients with early stage lung disease and prevent these large drops in lung function from occurring in the first place.

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