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Cystic Fibrosis Research News

Title:

REDUCED SURVIVAL IN ADULT CYSTIC FIBROSIS DESPITE ATTENUATED LUNG FUNCTION DECLINE

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What was your research question?

We wanted to find out more information about lung function decline and survival outcomes for people diagnosed with cystic fibrosis (CF) as an adult. We believed that people with an adult diagnosis of CF will have slower progression of disease and will have longer life expectancies than those diagnosed with CF at a younger age.

Why is this important?

There is scant literature available on disease progression in people diagnosed with CF after age 18. Publications available are based upon small numbers of people or a limited subset of the CF population.

What did you do?

We utilized the Cystic Fibrosis Patient Registry, maintained by the CF Foundation, which is a database of CF patients followed at CF care centers across the U.S and has information on more than 11,000 CF diagnoses. In those diagnosed as children, adolescents, or adults, we examined lung function decline for the first 5 years after diagnosis. We calculated survival rates at 10 and 15 years after diagnosis for those people with CF diagnosed at age 18 and above. We also looked at clinical factors associated with these outcomes, such as bacteria in the sputum, lung function at diagnosis, age, and gender.

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What did you find?

People who were diagnosed with CF in adulthood had a slower rate of lung function decline as compared to those diagnosed with CF in childhood or adolescence. Survival for adult diagnosis is excellent for those with good lung function at time of diagnosis. For those diagnosed when >45 years old, or with low lung function at diagnosis, survival rates are lower. The risk factors for worse lung health or lower survival rates amongst adult diagnosis were similar to that seen in children with CF, such as having Pseudomonas infection in the lung.

What does this mean and reasons for caution?

Despite the perception that people diagnosed at an older age with CF are those with mild disease, there are certain adult diagnosis CF individuals that have a high risk of death or lung transplant at 10 and 15 years after diagnosis. These tend to be the people with lower lung function and an older age at time of diagnosis. Although this study is based upon a very large dataset, information obtained from the CF Patient Registry is limited, as it is based on manually entered data on characteristics and death from each CF care center and can vary in accuracy or completeness.

What's next?

For a more definitive knowledge on outcomes for people diagnosed with CF, a prospective study following individuals after their diagnosis over a long term would be most informative. However, the data from our study will help frame a discussion regarding disease course and management for adults newly diagnosed with CF.

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