Cystic Fibrosis Research News

Title:

Variation in lung function and nutritional decline in cystic fibrosis by genotype: An analysis of the Canadian Cystic Fibrosis Registry

Authors:

Garvin J. Leung[1], Ting J. Cho[2], Thomas Kovesi[1,2,3], Jemila S. Hamid[1,2], Dhenuka Radhakrishnan*[1,2,3]

Affiliations:

1 Faculty of Medicine, University of Ottawa, Ottawa, ON, K1H 8M5, Canada
2 Children's Hospital of Eastern Ontario Research Institute, Ottawa, ON, K1H 5B2, Canada
3 Children's Hospital of Eastern Ontario, Ottawa, ON, K1H 8L1, Canada

What was your research question?

1. How do lung function and nutritional status change over time in Canadians with cystic fibrosis (CF)?
2. Do people with various genetic types of CF have different rates of change in their lung function and nutritional status over time?

Why is this important?

Lung function and nutritional status are important outcome measures of health status that cystic fibrosis (CF) health care providers use in caring for patients. Investigating how these outcomes change over time in different genetic types of CF provides a better understanding of this disease especially as new gene – specific CF therapies are being developed. We were particularly interested to see whether there are genetic types of CF that have comparable changes in lung function and nutritional status over time to the most common genetic profile, called ΔF508/ΔF508. This information would allow researchers to more fully evaluate the effectiveness of new gene-specific CF treatments.

What did you do?

The Canadian Cystic Fibrosis Registry, which is a national database of all Canadians with CF, was used to investigate changes in lung function and body mass index (a marker of nutritional status) between patients with different genetic types of CF. Four genetic groups containing a total of 2812 people were created and each of these were compared to the most common genetic group, ΔF508/ΔF508. Statistical testing was used to calculate rates of change in lung

function and body mass index over a 9 year time period while accounting for factors such as age and sex that can also affect lung function and body mass index.

What did you find?

We were successful in identifying certain genetic types of CF that have comparable lung function changes over time as people with the most common genetic type, ΔF508/ΔF508. These genetic types had similar functionality to the ΔF508/ΔF508 group in their cystic fibrosis protein receptor.

We did not identify any important differences in nutritional status (measured using standardized BMI score) change over time between all genetic types of CF.

What does this mean and reasons for caution?

This study shows that people with CF with different genetic profiles can have comparable natural disease trajectories. Knowing this information supports the use of another method to investigate how well new gene-specific CF medications perform in improving or stopping decline in lung function and nutritional status over time. Reasons for caution include that we were not able to account for certain patient and disease characteristics (such as tobacco exposure and microbiology data) in the analysis; however, we believe this provides a more real world comparison between groups.

What's next?

This study provides an overview of how genes can affect lung function and nutritional status in CF over time. Next steps include to consider including patients with different genetic types of CF as comparison groups when assessing the benefits and risks of recently developed gene-specific medications for CF.

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