Cystic Fibrosis Research News

Title:

Prospective longitudinal association between repeated multiple breath washout measurements and computed tomography scores in children with cystic fibrosis

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What was your research question?

As patients with cystic fibrosis develop severe structural damage in their lungs over time, it is important to know how well lung function tests tell us the amount of structural damage present. We wished to explore the extent of structural lung damage in a Copenhagen cohort of children age 6-18 years and measure how much it worsened over 2 years. Our research question was how well do lung function tests match with the structural damage seen on CT?

Why is this important?

The progression of structural lung disease is a sign of worsening CF lung disease and eventually leads to increased sickness. However, imaging through CT scans exposes children to some radiation, so we need to minimize the numbers of scans. Lung function tests do not pose harm and can be done much regularly, so it is crucial to know which lung function tests are most informative.

What did you do?

In Copenhagen, Denmark, we performed a study where 57 children 6-18 years old with CF participated. The study was conducted before treatment with CFTR-modulators were available. A CT scan of the lungs, as well as the spirometry and multiple breath washout (MBW) lung function tests, were conducted and then repeated 2 years later. The CT scans and lung function test were performed when the children had no symptoms of acute respiratory tract infections. During the study period all children followed the normal routine monitoring and treatment of disease by monthly visits to the CF Center in Copenhagen, including spirometry and sputum samples for bacteria.

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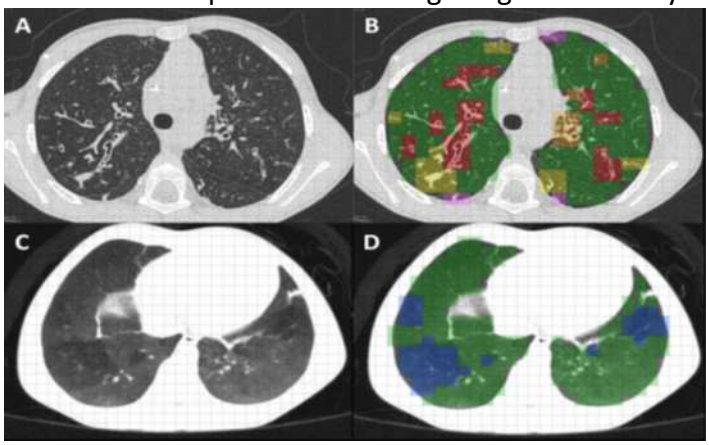
The CT scans were analyzed by a method called PRAGMA-CF, where a computer program calculates the proportion of the lung with structural lung disease after the observer has marked small squares on the lung images as healthy or diseased. (picture 1)



***Picture 1**:*
Green squares = healthy lung.
Yellow, red, purple and blue = different structural lung damages

What did you find?

On average we found no worsening of structural lung disease on the CT scans over the 2 years and there was no worsening in lung function according to the spirometry lung function test. However, the lung function data from MBW test worsened over the 2 years.

We found a statistically significant correlation between the MBW test results and structural lung disease at both baseline and follow-up, and also between the baseline MBW and follow-up CT. Children with improving or stable MBW results almost all had stable or improving structural lung disease seen on the CT scan. Finally, the MBW test predicted stable structural lung disease with a high probability.

On the other hand, we also found that MBW cannot predict worsening of structural lung disease, since many children with worsening MBW test results actually had stable or improved structural lung disease.

What does this mean and reasons for caution?

In conclusion we found that a 2-year interval for CT scans is too short as a standard for children with CF in the Copenhagen cohort. It should be noted that the MBW test and CT scans assess two different aspects of CF lung disease, but it seems that MBW might be used as a predictor of stable structural lung disease. More studies on the prediction of structural lung disease are needed for a more personalized timing of CT scans.

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Findings from studies should only be used for comparison between groups with similar treatments, as other studies from different countries have found progression of structural disease over both one and two years.

What's next?

In many CF centers around the world MBW is increasingly used in the clinic to monitor CF lung disease.

Studies exploring the use of MBW changes as a predictor of progressive structural lung disease are warranted.

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