Cystic Fibrosis Research News

Title:

Sensitive Structural and Functional Measurements and 1-Year Pulmonary Outcomes in Pediatric Cystic Fibrosis

[Sensitive measurements of structural changes in the lungs and lung function in children with cystic fibrosis – results after 1 year]

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What was your research question?

We aimed to provide additional information on newer/more-sensitive methods of measuring lung disease in comparison to the standard clinical practice in a group of children with cystic fibrosis (CF). For each method, we report the accuracy as well as links to structural abnormalities and results for clinical outcomes after one year.

Why is this important?

These newer methods of measuring lung structure and the decline in lung function have already been shown to be more sensitive to early lung disease. Also, several structural abnormalities can occur in CF lung disease where different treatments and courses of action might be better for limiting any future decline in lung function. However, we do not really know which lung function most accurately measure the different types of structural abnormalities and there may be some biases. So, if we have better information about the links between lung structure and lung function and how they affect clinical outcomes, clinicians can make more informed decisions on the best course of action/treatment.

What did you do?

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On the same day we collected different measures of lung function and structure from 27 children with CF. We then looked at how the structural measurements were linked to the lung function measurements. We then compared the 1-year outcomes from the children's electronic medical records to the structural and functional measures we took on the initial assessment. We looked at outcomes including changes in clinically measured lung function and the number of pulmonary exacerbations requiring admittance to hospital.

What did you find?

Three measurements (MBW, ^{129}Xe MRI, and UTE MRI) found significant levels of lung disease in a higher percentage of children (over 78%) than standard clinical measurements (spirometry, under 30%); this agrees with previously published results. MBW and ^{129}Xe MRI were best able to identify mucus plugging and airway changes while standard spirometry was best able to identify consolidations. Airway changes were also most strongly linked to future pulmonary exacerbations. Additionally, some of the children began CFTR modulator therapy during the following year which affected whether certain structural/functional measures were linked to outcomes.

What does this mean and reasons for caution?

The results show how using the more sensitive structural/functional measures of CF lung disease might better track disease progression. Additionally, the results suggest clinicians might be able to use the different functional measures possibly combined with structural measures to choose the best course of action/treatment. Therefore, using imaging to visualize both structural (UTE MRI or CT) and functional (^{129}Xe MRI) abnormalities will probably benefit future studies/clinical decisions. However, further work with a larger group of patients is necessary to confirm the results and improve sensitivity to smaller effects and biases.

What's next?

We plan to continue collecting these data in a larger number of patients to better gauge the potential structural links/biases for each functional measure. Additionally, we plan to continue assessing how these functional and structural measurements change with disease progression and treatments.

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