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Cystic Fibrosis Research News

Title:

Risk factors for progression of structural lung disease in school-age children with cystic fibrosis

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What was your research question?

The aim was to better understand if regular surveillance of school age children with cystic fibrosis (CF) using computed tomography of the thorax (chest-CT) can help us to identify risk factors for lung damage and faster progression of lung disease.

Why is this important?

Chest-CT is a good method to detect lung damage but to date, there is no agreement about how often or at what age chest-CT should be performed in children with CF. It is also unknown how intermittent and chronic bacterial airway infections impact the extent of lung damage and influence the yearly progression rate of lung disease seen on chest-CT. The finding from this study may enhance our understanding about how and when to use chest-CT in order to prevent CF lung disease progression.

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What did you do?

The study included 75 school-age children with CF attending the Gothenburg CF Centre who were born between 1990 and 2009. Starting at 6 years of age, all children underwent chest-CT every three years at their annual assessment. Lung function tests were performed regularly between the ages 6 to 18 years. Demographic data and information about sputum cultures for each child were retrieved from the Swedish CF Registry.

What did you find?

A. A single infection with *Pseudomonas aeruginosa* was linked to irreversible lung damage

B. The yearly progression rate of lung damage immensely increased with chronic infection with *Pseudomonas aeruginosa*

C. At the age of 7 years, greater lung damage was a risk factor for a faster yearly progression rate of CF lung disease.

D. Children diagnosed with CF after one year of age hade twice as much lung damage compared to children diagnosed with CF before the age of one.

E. Lung function tests did not provided enough information about the ongoing progression rate of CF lung disease

What does this mean and reasons for caution?

A. This study further emphasizes the importance of early detection and eradication of *Pseudomonas aeruginosa.*

B. Relying on spirometry as a sole objective method to evaluate the effect of treatment of *Pseudomonas aeruginosa* will probably underestimate the progression rate of CF lung disease.

C. Chest-CT or tests other than spirometry are needed in children under the age of 7 to identify those at risk of a faster progression rate for CF lung disease

D. Chest-CT is an important method of assessing CF lung disease and should be combined with other methods to track the progression rate of CF lung disease.

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What's next?

We plan to investigate whether repeated measurements of lung clearance index (LCI) measured with multiple breath washout can be used to identify children at risk of a faster progression rate of lung damage before the age of seven years.

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