Cystic Fibrosis Research News

Title:
Structural Lung Disease in Preschool Children with Cystic Fibrosis: An 18 Month Natural History Study

Lay Title:
Looking at lung disease over time in young children with CF

Authors:
Alan S. Brody[1], Rui Huang[2], Bin Zhang[2], Frederick R. Long[4], Scott W. Powers[3]

Affiliations:
From the Department of Radiology[1], the Division of Biostatistics and Epidemiology[2], and the Division of Behavioural Medicine and Clinical Psychology[3] at the Cincinnati Children's Hospital and the University of Cincinnati College of Medicine, Cincinnati, OH, USA, and from the Department of Radiology, Nationwide Children's Hospital and the College of Medicine, Ohio State University, Columbus, OH, USA[4].

What was your research question?
How much lung disease is there in 2–6-year-old children with CF? How does their lung disease change over time? This group of children had not been treated with CFTR modulators, so these results can give us a "baseline", something to compare to lung disease in children treated with modulators.

Why is this important?
We know that CF lung disease starts in infancy. Studies have shown that lung disease increases over the first few years, but there is less information available in older preschool children. This information helps us in designing research studies to show which new treatments are effective, and which are not. Knowing how much disease progresses, and how much that progression varies, can help us decide how often we should evaluate lung disease in preschool children.

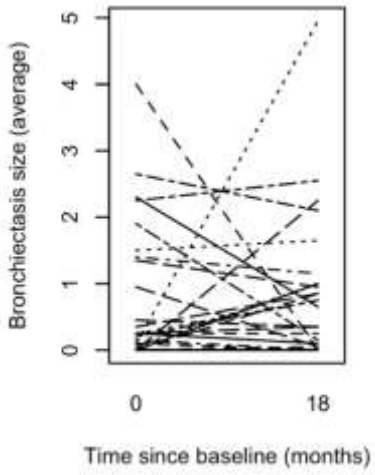
What did you do?
We did CT scans at the beginning and end of an 18-month nutrition study in 42 children with CF. CT scans were used because they give us the most detailed look at the appearance of the lungs. Two radiologists scored each CT scan for the amount and severity of bronchiectasis

(abnormally enlarged airways), the amount of mucous in the airways, and the amount of air trapping, which is a way to look at how hard it is to get air in and out of the lungs.

What did you find?

We found lung disease in 2/3 of the children at the beginning, 3/4 at the end. All abnormalities increased, but almost as many children showed an improvement in enlarged airways as showed worsening. In two children with specific mutations, one child showed greater improvement and the other greater worsening than children with the most common mutation.

The graph below shows differences among the children. In many there is little change, the line is close to horizontal. In some there is a large increase (the line angles up), while others show a large decrease (the line angles down).



What does this mean and reasons for caution?

Over an 18 month period young children vary widely in how their lung disease changes. In some their lung disease gets worse, and in some lung disease gets better. It has been widely accepted that bronchiectasis (enlarged airways) can get worse but cannot go back to normal. Our results suggest that, at least for young children, this is not true. Our finding that ling disease is different in children with different mutations is potentially important, but there were only 2 children with these mutations, so it is uncertain whether studies with more children would find the same thing.

Cystic Fibrosis Research News

What's next?

It will be important to see if larger studies find the same results. Our study suggests that changes in lung disease are more variable and more complicated than we thought. This may affect both research studies and recommendations for imaging in preschool children.

Original manuscript citation in PubMed

https://pubmed.ncbi.nlm.nih.gov/34961706/

cfresearchnews@gmail.com