Cystic Fibrosis Research News

Title:

Minimal change in structural, functional and inflammatory markers of lung disease in newborn screened infants with cystic fibrosis at one year

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What was your research question?

We wanted to find out whether one year old infants with CF diagnosed by newborn screening had changes in lung function and on chest CT scans (a detailed type of x-ray) which reflected inflammation and infection in their lungs.

Cystic Fibrosis Research News

Why is this important?

With the widespread introduction of newborn screening for CF, it is important that there are ways of measuring how healthy the lungs are in early life, for two reasons: 1) in order to understand which factors are linked to more severe lung disease in later childhood and adult life, and 2) to work out how best to measure the impact of new treatments, including which children stand to gain most benefit.

What did you do?

The London Cystic Fibrosis Collaboration recruited babies with CF diagnosed by newborn screening for this study. At 1 year of age, a range of tests was performed when the infants were in a stable condition. These included lung function, chest CT scan, and a test where a flexible tube called a bronchoscope is placed into the airways while the baby is anaesthetised. This allowed a sample of airway secretions (called BAL) to be collected and sent to the laboratory to look for signs of infection and inflammation. We also looked for infection on routine cough swabs during the first year of life.

What did you find?

65 infants with CF had the tests. Overall, their lung function was only mildly less than we would expect from healthy infants, and for the majority lung function was normal. Similarly, we saw very mild changes on chest CT scan, and these were only weakly related to the lung function results. Despite the mild changes, we could see that infants had worse lung function (measured by a test called the lung clearance index or "LCI") if they had infection detected by 1 year of age. We found an important marker of airway inflammation ("neutrophil elastase") in a fifth of infants, but this did not relate to how severe any lung changes were on their CT scans.

What does this mean and reasons for caution?

In this UK newborn screened group of infants, lung and airway damage was much milder at one year of age than expected. In contrast to reports from other research groups, the relationships between inflammation, abnormal lung function and changes on chest CT were weak, which may have reflected the mild nature of the changes that we found. It is possible that different tests or ways of interpreting test results might be needed to better understand the most important measures of health in newborn screened infants with CF, but the current results are very encouraging.

Cystic Fibrosis Research News

What's next?

It is really important that we understand the long-term implications of the very mild changes that we saw at 1 year of age, and this will be achieved by ongoing follow up of this London Cystic Fibrosis Collaboration newborn screened cohort.

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