European Cystic Fibrosis Society

Journal of Cystic Fibrosis
The Official Journal of the European Cystic Fibrosis Society

Cystic Fibrosis Research News

Title:

Respiratory symptoms do not reflect functional impairment in early CF lung disease

Lay Title:

Respiratory symptoms are not more frequent or severe in infants with CF compared to healthy infants.

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What was your research question?

Lung disease can develop within the first year of life in infants with cystic fibrosis (CF). However, the frequency and severity of respiratory symptoms in infancy are not known. We aimed to investigate if infants with CF have more frequent or more severe respiratory symptoms compared to healthy controls.

Why is this important?

It is known that lung impairment in CF infants is already present at birth. Former studies could show an impaired lung function or elevated (impaired) respiratory rate measurements in infants with CF. However, it is not known if these changes can be captured by the clinical presentation of the infants. In older children with CF, respiratory symptoms are more frequent compared to healthy children, however this has not been investigated in infancy.

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This is important to know, to optimize treatment of CF patients in the beginning of life and improve the outcome of the disease later in life.

What did you do?

We assessed respiratory symptoms in 50 infants with CF and 50 healthy controls from the age of 5 weeks to 12 months. Respiratory symptoms and respiratory rate measurements were documented by standardized weekly telephone interviews throughout the first year of life. Furthermore, infants performed lung function measurements in the first weeks of life.

What did you find?

Respiratory symptoms were not more frequent or more severe in infants with CF compared to healthy controls. CF infants with impaired lung function measurements in the first weeks of life did not present with more frequent or more severe throughout the first year of life. In addition, more frequent or more severe respiratory symptoms could not be detected in CF infants with elevated respiratory rate measurements.

What does this mean and reasons for caution?

The fact that CF infants do not present with more frequent or more severe symptoms in the first year of life indicates that lung impairment might not be captured by the clinical presentation of patients. This highlights that a) additional parameters like lung function and/or respiratory rate measurements should be obtained to be able to assess disease state and b) treatment decisions should not only be based on the clinical presentation of the patient only.

What's next?

Future studies with a higher number of subjects might be able to detect small differences in symptom presentation, additional differences and/or risk factors. Larger studies could help to even better understand early lung disease in CF.

Original manuscript citation in PubMed

https://pubmed.ncbi.nlm.nih.gov/34088612/

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