

Cystic Fibrosis Research News

Title:

Pulmonary exacerbations and acute declines in lung function in patients with cystic fibrosis

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What was your research question?

To better understand how and why clinicians may treat pulmonary exacerbations (PEx) in people with cystic fibrosis (CF), we asked two questions regarding their care – which patients are treated with antibiotics and how drops in lung function relate to diagnosis of PEx .

Why is this important?

Patients with CF endure repeated PEx and progressive loss of lung function. One reason why PEx are important is that frequently they involve an acute drop in lung function. Often this loss in lung function is not recovered following treatment. This lack of recovery helps explain the association of PEx with decreased survival in patients with CF. However, clinicians often diagnose and treat a PEx even when there is no acute drop in lung function, while sometimes patients experience an acute drop in lung function and are not treated. This lack of treatment for drops in lung function happens even though treatment, and especially IV antibiotic treatment, can lead to better recovery.

What did you do?

We performed two separate analyses using data from the Epidemiologic Study of CF. We analysed this very large dataset to explore outcomes of two clinical groups: 1) patients with acute drops in lung function who were treated with antibiotics compared to those who were not, and 2) patients diagnosed by their clinician as having a PEx with acute drop in lung function compared to those diagnosed as having a PEx without an acute drop in lung function. We considered decreases of at least 10% FEV1 as an acute drop in lung function.



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What did you find?

For our first question, we found that only 70.7% of acute drops in lung function were treated with antibiotics. Patients with at least 1 prior IV-treated PEx were more than twice as likely to be treated with antibiotics; those with 2 prior IV treated PEx were more than 4 times as likely to be treated. Other factors that meant a patient was more likely to be treated with antibiotics include: female sex, Medicaid insurance, hemoptysis (coughing up of blood or blood-stained mucus), sinusitis (runny nose), daily cough, clubbing (enlargement of the tips of the fingers or toes), lower baseline lung function, and airway sputum culture positive for *P.aeruginosa*, *S.maltophilia* or *B.cepacia*.

For our second question, we found that about one third (32.0%) of diagnosed PEx events, had a drop in FEV1 of less than 10%. These patients with a <10% drop tended to be non-Hispanic white, have non-F508del genotypes, have higher baseline lung function, be in better nutritional health, and not have a bacterial culture recorded from their airway sputum.

What does this mean and reasons for caution?

Although lung function has long been considered a standard for defining disease severity and survival probability, it appears that detecting an acute drop in lung function does not, in itself, drive the clinician to diagnose a PEx. In fact, data from our first question indicates that one-third of the time these large drops are not treated. Instead, a past history of recurrent PEx and clinical findings such as clubbing and daily cough seem to be major criteria driving the decision to treat. Furthermore, our second question reveals clinicians treat a large number of patients for a PEx (nearly one-third), even when there is relatively little change in lung function.

What's next?

How events producing a <10% change in FEV1 differ from ones producing a greater impact on lung function requires further investigation. Additionally, more information is needed to understand why clinicians do not treat some acute drops in FEV1, since we know that failure to treat is associated with worse long-term lung function.

Original manuscript citation in PubMed

<https://www.ncbi.nlm.nih.gov/pubmed/29685810>