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Title:

TREATMENT AND DEMOGRAPHIC FACTORS AFFECTING TIME TO NEXT PULMONARY EXACERBATION IN CYSTIC FIBROSIS

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What was your research question?

We wanted to understand what clinical characteristics are related to an increased risk of pulmonary exacerbation (PEX) for a person with CF.

Why is this important?

During PEX, symptoms of lung infection increase while quality of life decreases. PEX have been linked to lung function loss and earlier death. For these reasons, an important measure of whether a CF treatment improves health is if it reduces future PEX risk. However, other characteristics beyond daily medications may affect PEX risk, such as type of CF gene mutations, presence of bacteria in the lungs, living environment, and many more. If we learn which other factors predict future PEX risk, we can take them into account when we study how a CF treatment reduces PEX risk.

What did you do?

Using CF Foundation Patient Registry data from 193 children and adults who were treated for a PEX after Jan 1, 2010 at the Cleveland Ohio CF Care Center, we studied 65 different characteristics (things like CF mutation, presence of diabetes, bacterial infection, use of different chronic therapies such as Pulmozyme® or TOBI®, and how long and where they were treated with antibiotics for their PEX) to see if any measures could help us predict how

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soon each patient would have another PEx. Most of these patients (155) were treated again for a second PEx by the end of September, 2014.

What did you find?

One characteristic was by far the most likely to predict increased future PEx risk: the number of times a person had been treated for PEx in the previous year. Compared to persons without PEx in the previous year, those treated once or twice in the previous year were at 4-times greater risk of PEx and those treated 3 or more times were at 25-times greater PEx risk. No other factors, including some that we thought would be important such as type of bacterial lung infection or stage of lung disease, were nearly as informative.

What does this mean and reasons for caution?

Our results suggest that persons with CF can be divided into different categories that we call 'phenotypes' based upon the number of times they have had a PEx in the prior year, and that other measures commonly used to assess the health of persons with CF are not very helpful in identifying a person's PEx phenotype. Because our study was limited to a single CF Care Center, some of our observations may not apply to individuals living in other geographic areas. However, our results suggest that PEx phenotype may be important when studying treatments designed to reduce risk of PEx.

What's next?

We will repeat our study with all US CF Patient Registry patients treated for a PEx after Jan 1, 2010. If we see a similar result, then we will include prior-year PEx phenotype as a factor when studying therapies to reduce PEx risk in future CF clinical trials.

Original manuscript citation in PubMed

http://ac.els-cdn.com/S1569199315000478/1-s2.0-S1569199315000478-main.pdf?_tid=ee9852de-ea7b-11e4-aeac-00000aab0f6b&acdnt=1429878053_2e52cd9684905cfd70bd38501874fbb2

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