Cystic Fibrosis Research News

Title:
Autoimmunity in People with Cystic Fibrosis Journal of Cystic Fibrosis

Lay Title:
Autoimmunity (a condition where the body's immune system mistakes its own healthy tissues as foreign and attacks them) in People with Cystic Fibrosis

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What was your research question?
To defend the body, the immune system must be able to distinguish “self” from “non-self”. When this system is not in balance it can lead to autoimmune diseases. We do not know whether some difficult-to-manage symptoms experienced by people with cystic fibrosis (pwCF) may be due to autoimmunity.

Why is this important?
Some symptoms experienced by pwCF may be difficult to manage, such as unusual gastrointestinal (GI) complaints, vasculitis (which may appear as rashes), or arthritis. These may or may not be due to autoimmunity. For a variety of reasons, care center teams may not recognize or may dismiss ongoing pain or physical distress that is inconsistent with typical CF. It can be frustrating to pwCF and family members when challenged by symptoms that do not fit the “normal box” of CF. We want to know whether autoimmunity could be a potential cause of some of these symptoms.

What did you do?
We reviewed the medical literature to explore what is known about autoimmunity in pwCF and to identify areas where there are gaps in knowledge.

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What did you find?

The immune response, inflammation, nutritional factors, and changes in the healthy bacteria of the airways and gut (gastro-intestinal (GI) tract) (the "microbiome") may be factors that lead to autoimmunity. Cystic fibrosis transmembrane regulator (CFTR) may also have a role in autoimmunity, even in people without CF. Although we don't know why, blood tests for autoimmune markers are increased in pwCF but these may or may not be connected to organ involvement, making diagnosis difficult. We know most about links between autoimmune involvement and the GI tract in pwCF, but our understanding of the role that autoimmunity might play in skin or joints is less clear.

What does this mean and reasons for caution?

We have proposed ways that the changes seen in pwCF might put them at higher risk for autoimmunity and be a possible explanation for symptoms that don't respond to regular CF treatments. However, much of the evidence about autoimmunity in pwCF is either old or of poor quality. Blood tests for autoimmune markers can be misleading in pwCF. We did not explore whether drugs used to treat autoimmunity in people without CF would be helpful to pwCF.

What's next?

We think that next steps to understand autoimmunity in CF should include an in-depth assessment of the community perspective on its impact. Autoimmunity is a complex topic. Bringing together specialists in fields such as pulmonology, gastroenterology, immunology, and rheumatology, would help define the path forward in basic science and clinical practice.

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