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New drug target found for cystic fibrosis lung disease

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Vancouver researchers have discovered the cellular pathway that causes lung-damaging inflammation in cystic fibrosis (CF), and that reducing the pathway's activity also decreases inflammation. The finding offers a potential new drug target for treating CF lung disease, which is a major cause of illness and death for people with CF.

"Developing new drugs that target lung inflammation would be a big step forward," says Dr. Stuart Turvey, who led the research. Dr. Turvey is the director of clinical research and senior clinician scientist at the Child & Family Research Institute and a pediatric immunologist at BC Children's Hospital. He is an associate professor in the Department of Pediatrics at the University of British Columbia.

The research was published online last week in the *Journal of Immunology*.

For the study, researchers compared the immune response of normal lung cells with that of CF lung cells after exposing both types of cells to bacteria in the lab. In healthy cells, exposure to bacteria triggers the cell to secrete special molecules that attract immune cells to fight the infection.

In CF lung cells, the researchers discovered that a series of molecular events called the unfolded protein response is more highly activated. It causes the CF lung cells to secrete more molecules that attract an excessive amount of immune cells, which leads to increased

inflammation.

They also found that treating the CF cells with a special chemical normalized the unfolded protein response and stabilized the cells' immune response.

CF is the most common genetic disease affecting young Canadians. One in every 3600 children born in Canada has CF. There is no cure. A build-up of mucus in the lungs causes people with CF to be susceptible to bacterial lung infections, which trigger inflammation and swelling. Over time, the recurring cycle of infections and inflammation damages the lungs and can lead to the need for lung transplantation. The only treatments for lung inflammation are steroids and anti-inflammatory medications, which can have significant side effects.

The researchers are planning further study to validate these findings in a larger number of lung cell samples from people with CF.

Provided by Child & Family Research Institute

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