MedicalXpress

New study discovers immune system protein can fix cystic fibrosis cells

September 2 2015

Scientific experiments examining what happens to the faulty channel protein that causes cystic fibrosis during inflammation have yielded unexpected and exciting results. The study, conducted by Sara Bitam and her colleagues at INSERM in France, has just passed peer review on open science publishing platform *F1000Research*.

Cystic fibrosis is a life-limiting auto¬somal recessive monogenic disorder that affects 1 in every 2000 - 3500 newborns in the EU and US per year. It is caused by mutations in the gene that encodes the CFTR protein, an epithelial ion channel involved in salt and fluid transport in multiple organs including those in the respiratory system. F508del, the most common mutation, produces a faulty CFTR protein that degrades shortly after creation. The lack of a functional CFTR protein makes individuals more prone to respiratory infections with excessive inflammation, which in turn leads to deterioration of lung function, the main cause of death in patients.

In their article, titled 'An unexpected effect of TNF-α on F508del-CFTR maturation and function', the authors report that a 10 minute exposure of cells expressing the faulty CFTR channel to TNF-α, an inflammatory cytokine produced by white blood cells, can 'fix' these cells, at least temporarily.

They first found a transient form of this effect in HeLa cells (derived from a cervical cancer cell line). This was surprising, and when they repeated the experiments in human bronchial epithelial cells, which have

more clinical relevance to the condition, they found an even better effect with the cells working as they should for at least 24 hours.

Further experiments elucidated that TNF-α achieves this by preventing the faulty CFTR protein from being degraded before it is successfully transported to the cell membrane.

Aleksander Edelman, the Principal Investigator of the study, outlined his group's next steps: "We hope to investigate whether TNF-α is linked to the severity of cystic fibrosis; if so then TNF-alpha levels might be one of a panel of molecules that could be used as a prognostic marker in the disease."

Rebecca Lawrence, Managing Director of *F1000Research*, said: "These findings are an exciting development in the bid to find a cure for cystic fibrosis - a devastating disease that affects many thousands of people worldwide.

"It is really important that results of this nature are published in the public domain as quickly as possible for peer review and we are pleased to be able to facilitate this through our open science publishing platform."

More information: Bitam S, Pranke I, Hollenhorst M et al. An unexpected effect of TNF-α on F508del-CFTR maturation and function [v2; ref status: indexed, f1000r.es/5tv] F1000Research 2015, 4:218. DOI: 10.12688/f1000research.6683.2

Provided by Faculty of 1000

Citation: New study discovers immune system protein can fix cystic fibrosis cells (2015,

September 2) retrieved 10 April 2024 from https://medicalxpress.com/news/2015-09-immune-protein-cystic-fibrosis-cells.html

This document is subject to copyright. Apart from any fair dealing for the purpose of private study or research, no part may be reproduced without the written permission. The content is provided for information purposes only.