Ivacaftor improves smooth muscle function in cystic fibrosis patients

April 7 2016

Cystic fibrosis (CF) is caused by mutations in the chloride channel CFTR, which disrupts fluid transport in the lungs. CF patients have a variety of complications, including airway obstruction, infection, and pathological tissue remodeling. Alterations in airway smooth muscle have been observed in CF patients but it is not clear if these abnormalities are directly due to loss of CFTR in airway smooth muscle cells.

In this issue of *JCI Insight*, David Stoltz of the University of Iowa and colleagues provide evidence that CTFR dysfunction directly alters the elasticity and blood supply of the airway. The authors evaluated smooth muscle function in a cohort of CF patients before and immediately after treatment with ivacaftor, which restores CFTR function in this set of patients. Treatment rapidly restored CFTR function, improved airflow, and increased the capacity and flexibility of small airways.

The results of this study indicate that loss of CFTR in airway smooth muscle is responsible for some CF-associated symptoms.

More information: Ryan J. Adam et al. Acute administration of ivacaftor to people with cystic fibrosis and a G551D-CFTR mutation reveals smooth muscle abnormalities, *JCI Insight* (2016). DOI: 10.1172/jci.insight.86183