Kilpatrick

Insights: Publications

A Functional Anatomic Defect of the Cystic Fibrosis Airway

190 AM. J. RESPIRATORY & CRITICAL CARE MED. 421

August 15, 2014

Written by

The mechanisms underlying cystic fibrosis (CF) airway pathophysiology are unknown and remain a topic of debate. Current hypotheses include depleted airway surface liquid and altered mucus biosynthesis secondary to absent CF transmembrane conductance regulator (CFTR)-mediate bicarbonate transport. Using innovative technology enabling concurrent quantification of multiple functional parameters of airway epithelium in a colocalized fashion, we have shown that the interrelationship between airway hydration and mucus transport is altered in CF. This disruption appears to be mediated by absence of bicarbonate transport and may constitute a fundamental mechanism underlying the pathogenesis of CF lung disease.