New test could lead to personalized treatments for cystic fibrosis

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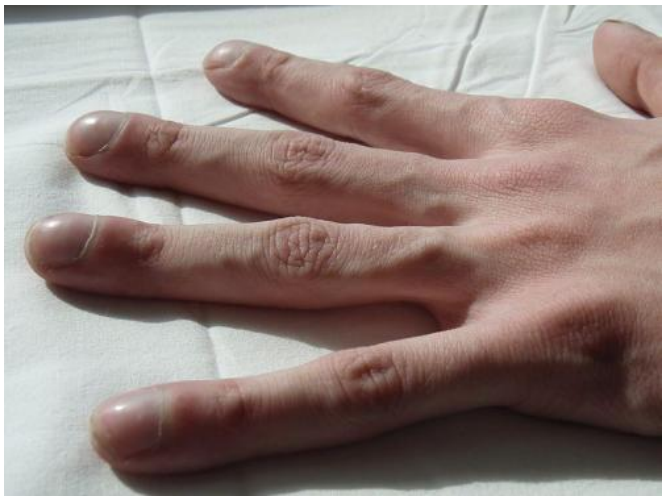

"Clubbing" of the fingers is a classic features of Cystic Fibrosis, although not present in many patients. Credit: Jerry Nick, M.D./ Wikipedia

Cystic fibrosis is a devastating disease caused by mutations in a specific gene, known as the CFTR gene. But not everyone with cystic fibrosis has the same symptoms or responds to drug treatments in the same way. In a new pilot study, researchers from the University of Cambridge and Yale University developed a novel, straightforward way to test multiple drugs on cells obtained from individual patients with cystic fibrosis, raising the possibility of highly personalized drug treatment.

The test combines high-speed video microscopy with a novel video analysis algorithm to measure the coordinated movement of cilia—hair-like structures covering airway cells that remove mucus from the lungs and upper airways. In people with cystic fibrosis, thick mucus accumulates in the airways causing chronic infections, which obstruct breathing and disrupt normal ciliary movement. The research team used their test to study the movement of cilia in cells derived from multiple patients with different cystic fibrosis mutations, comparing those samples to normal cells. They then measured the response of those cells to six different drug treatments, including ones not currently approved for cystic fibrosis.

The team found that the patients responded differently to the drugs, suggesting that what works for one person might not work for another, even if each carries the same mutation. Going forward, the researchers hope that their test could be used to recommend more personalized treatments for this life-threatening disease.

Maurizio Chioccioli, a postdoctoral researcher in the Section of Pulmonary, Critical Care, and Sleep Medicine at Yale, is first author of the paper, which is published in *Nature Communications*.

More information: M. Chioccioli et al. Phenotyping ciliary dynamics and coordination in response to CFTR-modulators in Cystic Fibrosis respiratory epithelial cells, *Nature Communications* (2019). DOI: 10.1038/s41467-019-09798-3

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