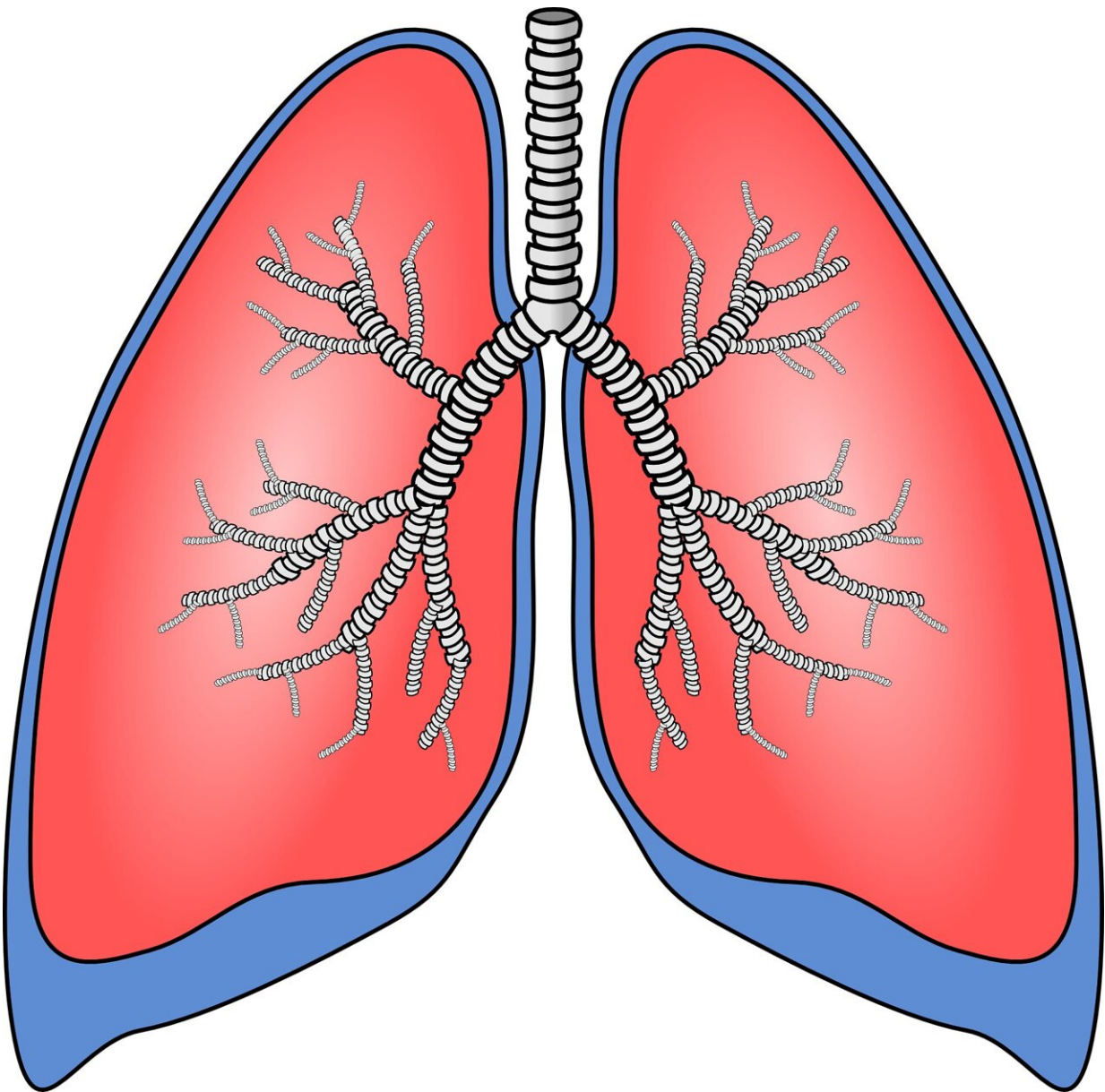


Researchers release results from clinical trial for treatment of primary ciliary dyskinesia

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Primary ciliary dyskinesia (PCD) is a rare genetic disease that causes the tiny hair-like filtration structures in the respiratory tract to stop working. As a result, the lungs accumulate thick mucus and become more susceptible to inflammation and recurring infections, both of which can result in long-term lung damage.

Thomas W. Ferkol, Jr, MD, a pediatric pulmonologist at UNC Health and chief of pulmonology in the UNC Department of Pediatrics, and researchers at Parion Sciences Inc. recently led a phase 2 study of idrevloride in hypertonic saline in people with primary ciliary dyskinesia (PCD).

Their results, which [were published](#) in *The Lancet Respiratory Medicine*, demonstrate that idrevloride in hypertonic saline is safe and associated with a significant improvement in lung function over a 28-day period in individuals with primary ciliary dyskinesia when compared to hypertonic saline alone.

"Before this manuscript, there were only two published, randomized controlled trials of any treatment for PCD, and much of our [current practice](#) has generally been extrapolated from other forms of bronchiectasis, like cystic fibrosis," said Ferkol.

CLEAN-PCD was a phase 2, randomized, double-blind, placebo-controlled crossover trial conducted at 32 adult and pediatric care centers and university hospitals around the world. A total of 123 adults and adolescents with primary ciliary dyskinesia aged 12 years or older were randomly assigned to receive idrevloride in hypertonic saline, hypertonic saline alone, idrevloride alone, or placebo.

Idrevloride is an inhaled investigational epithelial sodium channel (ENaC) inhibitor, that is formulated to hydrate the mucus in the lung and improve clearance of mucus by cough and lung function. The ENaC inhibitor has been well-tolerated in multiple clinical trials in healthy volunteers and patients with other [respiratory diseases](#) who accumulate excessively concentrated mucus in their lungs, including primary ciliary dyskinesia.

"There is a clear need for newer and better treatments for this rare lung disease, and hopefully the planned, more extensive [clinical trials](#) will further show the benefits of inhaled idrevloride in hypertonic saline," said Ferkol.

In addition to publication in *The Lancet Respiratory Medicine*, Ferkol presented the CLEAN-PCD study results at the European Respiratory Society International Congress 2023 during the Abstracts Leading to Evolution in Respiratory Medicine Trials (ALERT) session.

More information: Felix C Ringshausen et al, Safety and efficacy of the epithelial sodium channel blocker idrevloride in people with primary ciliary dyskinesia (CLEAN-PCD): a multinational, phase 2, randomised, double-blind, placebo-controlled crossover trial, *The Lancet Respiratory Medicine* (2023). [DOI: 10.1016/S2213-2600\(23\)00226-6](https://doi.org/10.1016/S2213-2600(23)00226-6)

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