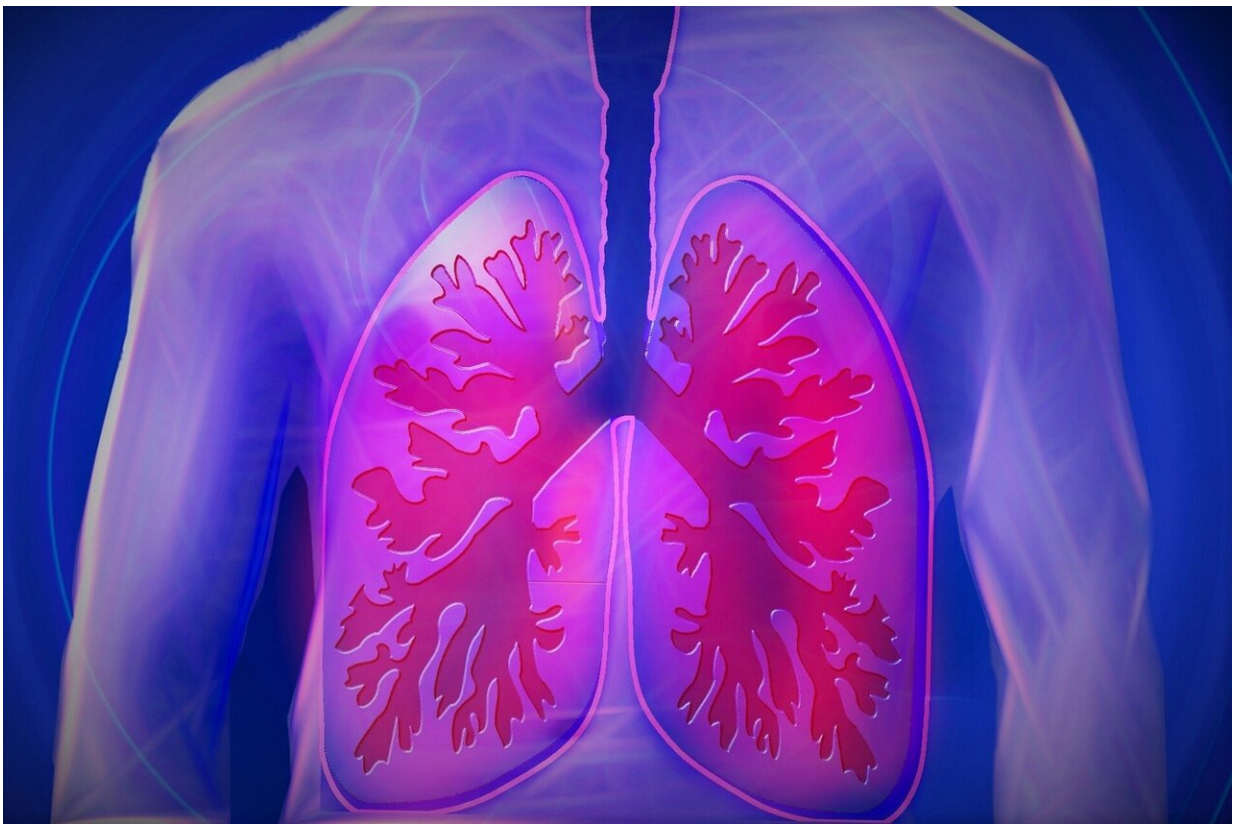
New therapy for acute exacerbations of idiopathic pulmonary fibrosis could potentially save lives

November 24 2021, by Adam Pope



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Researchers with the University of Alabama at Birmingham Marnix E. Heersink School of Medicine have published a new study in *PLOS ONE*

detailing a new therapy for acute exacerbations of idiopathic pulmonary fibrosis, or AE-IPF, that could potentially be lifesaving.

Recent research studies have suggested that proteins called antibodies that are produced by the immune system might be involved in the lung damage of AE-IPF. Antibodies produced by the immune system normally help to fight infections by attacking bacteria and viruses without harming our own tissues, but when this process is dysregulated, it can cause harm to various organs.

In many patients with AE-IPF, and especially those with acute exacerbations, there is evidence that certain antibodies—called autoantibodies—attack the lung and contribute to the injury and scarring that occurs in AE-IPF. According to the National Institutes of Health, about 100,000 people in the United States have AE-IPF, and approximately 30,000 to 40,000 new cases are found each year. This disease carries a high mortality rate with median survival time of 3–5 years after diagnosis; there are approximately 15,000 deaths attributable to IPF every year.

"Our recent studies have found that many AE-IPF patients appear to have excessive autoantibody levels in blood and lungs that might make their disease worse," said Steve Duncan, M.D., principal investigator and professor with the UAB Division of Pulmonary, Allergy and Critical Care Medicine.

Duncan and colleagues used a treatment protocol to reduce autoantibodies that included therapeutic plasma exchange, a medication approved by the Food and Drug Administration called rituximab for the treatment of autoantibody diseases such as rheumatoid arthritis, and intravenous immunoglobulin, which resulted in significant clinical improvement in about half the AE-IPF patients. The majority of AE-IPF episodes treated with conventional medications succumb from their

disease, often within days.

"In our study, patients who responded to this treatment protocol had higher pre-treatment levels of antibodies," said Tejaswini Kulkarni, M.D., assistant professor in the Division of Pulmonary, Allergy and Critical Care Medicine. "Understanding this further may help identify patients who are most likely to benefit from these treatments and optimize the utilization of this treatment protocol."

"Incremental trials could further refine and optimize autoantibody reduction regimens, validate measures to facilitate the personalized applications of these treatments, and potentially illuminate other important upstream disease mechanisms aside from or in addition to humoral autoimmunity," Duncan said.

More information: Tejaswini Kulkarni et al, Correlates of survival after autoantibody reduction therapy for acute IPF exacerbations, *PLOS ONE* (2021). DOI: 10.1371/journal.pone.0260345

Provided by University of Alabama at Birmingham