Immunodeficient patients with secondary lung disease benefit from combined chemotherapy

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A team of researchers at the Medical College of Wisconsin and Children's Hospital of Wisconsin Research Institute defined a new treatment for a potentially fatal lung disease in patients with a primary immunodeficiency known as common variable immunodeficiency (CVID).The findings are published in the *Journal of Clinical Immunology*.

Common variable immunodeficiency (CVID) is the most common primary immunodeficiency that requires regular treatment with medication, specifically immunoglobulin (antibodies) replacement therapy. With immunoglobulin therapy, deaths from infection in patients with CVID have decreased and deaths due to non-infectious complications have increased. One of the most frequent causes of death now in CVID is a lung disease known as granulomatous-lymphocytic interstitial lung disease (GLILD). In GLILD, white blood cells abnormally accumulate in the lung, which leads to a progressive decline in lung function and, eventually, respiratory failure. The researchers evaluated patients with GLILD to see if rituximab and azathioprine (combination chemotherapy) would improve pulmonary function and/or radiographic abnormalities by killing these white blood cells in the lung.

"The most common medication used for GLILD are corticosteroids but in our hands this type of medication did not clear the disease. Therefore, we designed therapy aimed at killing the specific type of lymphocytes, B

cells and T cells, we found in abnormal numbers in the lung biopsies we examined. We reasoned if we killed the lymphocytes in the lung, lung function and radiographic abnormalities would both improve, which is what we found." said John M. Routes, M.D., professor and chief of asthma, allergy and immunology at the Medical College of Wisconsin (MCW), medical director of allergy/clinical immunology at Children's Hospital and researcher at the Research Institute.

Provided by Medical College of Wisconsin