# Finding might hold answer to preventing lung disease in cystic fibrosis

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(Medical Xpress)—Researchers at the Institute, Queensland Children's Medical Research Institute (QCMRI), and the Telethon institute for Child Health Research, Perth; are one step closer to preventing serious lung disease which is the main cause of suffering in cystic fibrosis.

Cystic fibrosis is an inherited condition that affects one in 2800 children, with one in 25 people a carrier of the gene that causes the condition. There is no current cure however researchers have been working to prevent the onset of lung disease.

The study, which is part of the Australian Respiratory Early Surveillance Team for Cystic Fibrosis (AREST CF) program, sought to determine risk factors for the onset of bronchiectasis. Bronchiectasis is a progressive and persistent form of lung disease.

Researchers studied 127 infants diagnosed with cystic fibrosis and found that they could determine which children were at most risk of developing bronchiectasis by three years of age.

The study showed children with cystic fibrosis who have neutrophil elastase in their lungs at three months are seven times more likely to develop bronchiectasis by 12 months, and four times more likely by three years.

Neutrophil elastase is one of the body's mechanisms for destroying bacteria in the body.

The levels of neutrophil elastase in the body is normally controlled, however in cystic fibrosis excess neutrophil elastase can cause breakdown of the lung structure. This leads to irreversible lung damage like bronchiectasis.

Lead researcher, Professor Sly from QCMIR said, "If we have a mechanism for mopping up neutrophil elastase then we can potentially stop or delay the start of damage to the lungs. We are currently undertaking a clinical trial using azithromycin which has anti-inflammatory effects to prevent bronchiectasis in infants with cystic fibrosis".

The trial, COMBAT CF, is a clinical trial undertaken in Australia and New Zealand which is the first in the world to try and prevent irreversible lung damage early in life.

"We are seeing lung damage in children with cystic fibrosis as young as three months old. Clearly lung disease starts in the first months of life so early intervention is paramount to improving health and quality of life."

This study was a joint collaboration between the Queensland Children's Medical Research Institute under the direction of Professor Peter Sly, The Telethon Institute for Child Health Research under the direction of Professor Stephen Stick and Murdoch Childrens under the direction of Professor Sarath Ranganathan.

The study was published in the *New England Journal of Medicine*.

Provided by Murdoch Childrens Research Institute