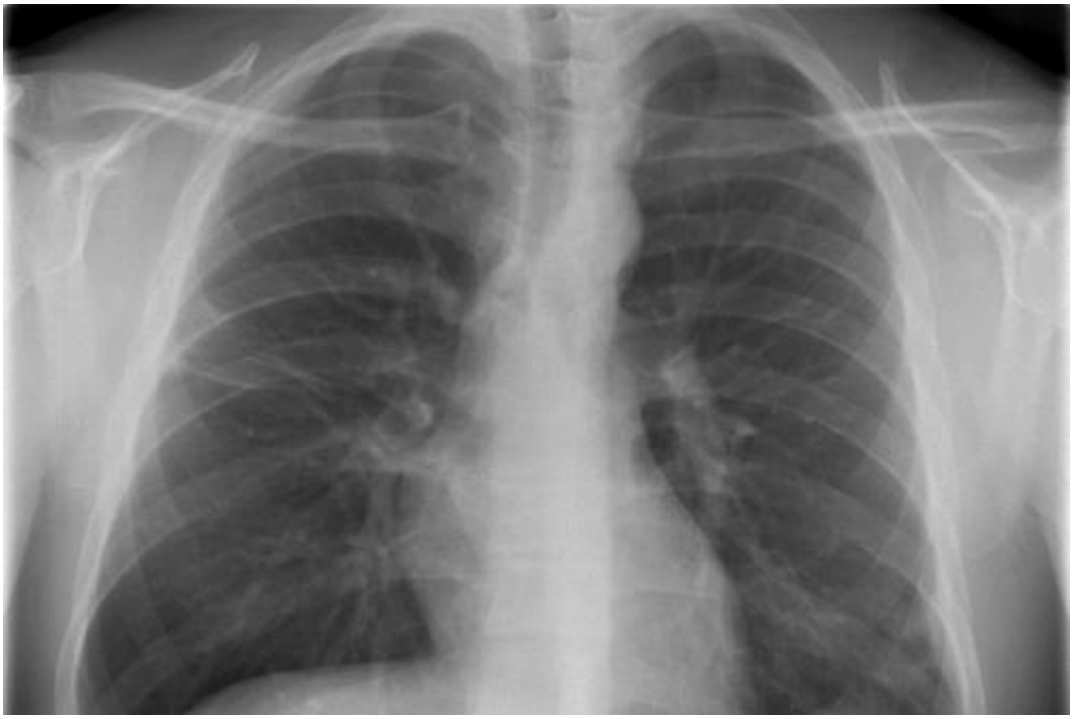
Research could be life-prolonging for cystic fibrosis patients

March 6 2015



Preventing lung infections in childhood could stop later life-threatening infections for people with cystic fibrosis (CF), according to the latest research carried out at Aston University.

Academics at the University are investigating how bacteria found in children with CF can disarm their natural defence mechanisms, making

it easier for more virulent bacteria to infect their airways.

By the time many CF patients reach adulthood, they have already contracted the chronic *Pseudomonas aeruginosa* bacterial strain, which can cause extensive and eventually fatal damage to the lungs. The infection is currently extremely difficult to treat effectively.

Dr Lindsay Marshall, who is leading the study, will use a replica model of a human airway with CF to treat early childhood infections with a range of antibiotics to calculate the extent to which *P. aeruginosa* can be stopped.

The model, made entirely of human cells, was developed at Aston University and can accurately duplicate the workings of a human airway and the progression of the disease.

Dr Marshall said: "We still do not fully understand the link between the cause of cystic fibrosis and the contraction of cystic fibrosis lung disease. There is a clear need to learn more about the disease process to allow us to develop new and better treatments to keep patients healthy for longer.

"This project will also allow us to demonstrate how well our layered human cell model replicates CF lung disease and in what ways it could be used to evaluate the effectiveness of future treatments. We hope to use different human cell models in future to examine how the body's natural defences are altered by a variety of other inflammatory and infectious conditions."

A further major aim of the study, funded by a £99,336 Human Research Trust grant, is to establish new methods to reduce the number of animals used in respiratory studies. Last year in the UK, more than 115,000 animals were involved in testing for conditions including smoke-related

lung diseases and asthma.

Despite many species of animal being used for CF research, they do not naturally have the disease. It is costly to genetically manipulate them to contract a form of CF, which still does not recreate typical human CF.

Dr Marshall added: "We simply cannot use animals to model the decline in lung function seen in people with CF, the infections typical of people with CF or the administration and dosage of drugs required to treat the condition. Our human CF model, which contains different layers of cells found naturally in the airways, is extremely representative of what happens in human airways and is both ethically and scientifically an improvement upon current animal models."

Cystic Fibrosis is the most common life-threatening inherited disease in the UK, affecting around 9,000 people. The condition causes the lungs to produce large quantities of thick, dehydrated mucus that block the airways, resulting in a decline in lung function with age and an increased susceptibility to infection. Thanks to the introduction of new treatments and therapies, the average life expectancy of people with cystic fibrosis has risen to 41 years of age.

Provided by Aston University