



THE UNIVERSITY *of* EDINBURGH

News Release

Issued: Day Month Year

Cystic fibrosis study offers new therapy hope for patients

Fresh insights into how cystic fibrosis affects immune cells could pave the way for new treatments for the condition.

Scientists have discovered that cells in patients with cystic fibrosis that normally defend against infection can also perpetuate damage to the lungs.

Drugs that target these cells could help to stem progression of the disease, the researchers say.

The team focused on immune cells called neutrophils, which are part of the body's first line of defence against infections.

Once an infection has been cleared, neutrophils are usually programmed to die off quietly, so that they do not mistakenly cause damage to healthy tissues.

In patients with cystic fibrosis, neutrophils survive longer than they are supposed to and are a key contributor to the lung damage associated with the condition.

Researchers from the University of Edinburgh discovered that neutrophils from cystic fibrosis patients are more resistant to the usual mechanism of cell death – a process called apoptosis.

Tests on human and pig cells showed that the ability of neutrophils to survive longer is directly related to the underlying genetic mutation that causes cystic fibrosis.

Instead, the cells die by a different process, which causes them to disintegrate and expel their damaging contents into the surrounding area of the lung.

This process – known as NETosis – promotes inflammation and may therefore promote damage to healthy tissues in the lung.

The researchers were able to block this process by treatment with a drug that encourages neutrophils to die by apoptosis, paving the way for new therapies for the disease.

Cystic Fibrosis causes thick, sticky mucus to build up in patient's lungs and digestive tracts. It causes persistent coughs and breathing difficulties, as well as leaving patients vulnerable to recurrent infections.

Ranked among the top universities in the world

Around 11,000 people in the UK are living with the disease. The current predicted life expectancy is 47 years, although babies born today may expect to live longer thanks to improvements in care and new therapies.

The research, published in the journal *Thorax*, was funded by the Medical Research Council and Wellcome. The international team included researchers in the US and Ireland.

Dr Robert Gray, of the Medical Research Council Centre for Inflammation Research at the University of Edinburgh, said: “Therapies targeting inflammation are not readily available but are needed for the treatment of cystic fibrosis. This work, although at an early stage, will help in the development of new anti-inflammatory treatments for this debilitating condition.”

Dr Donald Davidson, Senior Research Fellow at the MRC Centre for Inflammation Research, said: “How our immune cells die can be as important as how they function in life for protecting our bodies. This study shows that encouraging them to die quietly can help to prevent damaging inflammation.”

For more information please contact:

Jen Middleton, Press & PR Office, 0131 650 6514; 07795 640662; Jen.Middleton@ed.ac.uk

Ranked among the top universities in the world