Medical Xpress

Protein opens hope of treatment for cystic fibrosis patients

September 11 2008

Scientists have finally identified a direct role for the missing protein that leaves cystic fibrosis patients open to attack from lung-damaging bacteria, the main reason most of them die before their 35th birthday, scientists heard today at the Society for General Microbiology's Autumn meeting being held this week at Trinity College, Dublin.

"Chronic lung infections are by far and away the biggest problem with the genetic disease cystic fibrosis, affecting almost 90% of patients. The majority of these bad infections are caused by one particular type of bacteria called Pseudomonas aeruginosa," said Professor Gerald Pier from Harvard Medical School in Boston, USA. "Once a chronic infection starts in a cystic fibrosis patient it is almost never cleared and will go on getting worse and causing lung damage over many years before killing the patient by the age of 35 years."

The US scientists trying to work out why cystic fibrosis patients get this infection have discovered that the protein called CFTR that is either missing or not working properly in their lungs is needed by our bodies to recognise when Pseudomonas aeruginosa bacteria are inhaled. People whose CFTR protein is working correctly can rapidly clear the infectious bacteria out of their lungs.

"In cystic fibrosis patients the recognition system is deficient or absent, and the patient does not rapidly expel the bacteria, allowing the microbes to settle into the lungs and cause a chronic infection," said Professor Pier.

By understanding how our lung cells use CFTR to recognize and properly respond to Pseudomonas aeruginosa infections the Harvard team hopes that they will be able to identify points of intervention that could be activated in cystic fibrosis lungs to increase their resistance to infection.

"At the moment our research is more geared to identifying the molecules that lung cells use to resist infection, although our longer term goal is to find ways to restore the resistance of the lung to infection," said Professor Pier. "Our major path right now is to further understand the general process of resistance to infection, using cystic fibrosis as a model as well as a real disease. But the next step will be to try some drug interventions to improve the resistance of cystic fibrosis patients to infection which will undoubtedly prolong their lives and enhance the quality of their life as well".

Source: Society for General Microbiology

Citation: Protein opens hope of treatment for cystic fibrosis patients (2008, September 11) retrieved 25 April 2024 from
https://medicalxpress.com/news/2008-09-protein-treatment-cystic-fibrosis-patients.html