Cystic fibrosis breakthrough reveals why females fare worse than males

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(Medical Xpress) -- Cystic Fibrosis (CF) is the most common life-threatening inherited disease in Ireland with the highest incidence of this disease globally seen on this island. Females with CF have a poorer outcome as a result of serious bacterial infections in their respiratory tract. Collaborative research conducted in Dublin between the Royal College of Surgeons, Beaumont Hospital and the School of Medicine, Trinity College has furthered our understanding of this phenomenon and was recently published in the prestigious *New England Journal of Medicine*.

“This is a fundamental breakthrough in understanding why females with cystic fibrosis have a poorer prognosis than males,” says Trinity’s Dr Stephen Smith, of the Department of Clinical Microbiology in the School of Medicine. It shows for the first time that the female hormone oestrogen promotes the presence of a particular mucoid form of the bacterium Pseudomonas aeruginosa in the lungs of women with CF. When in the mucoid form, bacteria are surrounded by a layer of gelatinous material and this is poorly penetrated by antibiotics. In the clinic, the mucoid type of Pseudomonas aeruginosa is more difficult to treat and causes significant lung inflammation, explaining, in part, why females with CF, have a worse outcome. This study opens the way to new understanding and potential therapeutic approaches in CF.

Professor Gerry McElvaney, Director of the Respiratory Research Laboratory RCSI/Beaumont Hospital and a senior author on the paper stated: “This research study is among the first examples which shows the

effects of gender hormones on infections and therefore has major implications for conditions beyond cystic fibrosis including other respiratory diseases such as asthma."

Dr Sanjay Chotirmall, a Molecular Medicine Ireland (MMI) Clinician-Scientist fellow is the first author on the paper and Dr. Catherine Greene is the joint senior author.

The clinical research was carried out in the Cystic Fibrosis Unit, Beaumont Hospital and also utilised data from the National Cystic Fibrosis Registry of Ireland.

This paper is an excellent example of the translational research presently underway in RCSI and illustrates interdepartmental and inter-institutional links as co-authors on the paper include colleagues from the Departments of Medicine (Dr Sonya Cosgrove), General Practice (Dr Borislav Dimitrov) and Molecular Medicine (Professor Brian Harvey), RCSI; Beaumont Hospital/RCSI (Dr Cedric Gunaratnam, Professor Shane O'Neill); and the School of Medicine (Dr Stephen Smith), Trinity College, Dublin.

The study's results are being presented by Dr Greene at the American Thoracic Society meeting in San Francisco this month and by Dr Chotirmall at the European Respiratory Society meeting in Vienna later this year.

More information: "Effect of Estrogen on Pseudomonas Mucoidy and Exacerbations in Cystic Fibrosis" *New England Journal of Medicine*.

Provided by Trinity College Dublin

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