

A rare lung cell type unveiled

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Recent studies of different cells in the epithelia of lung airways, led by two independent teams of scientists have revealed a new rare cell type. Using single-cell RNA sequencing, gene expression of the whole set of cells isolated from the epithelia of the mouse airways has been analysed. A catalogue of different cell types was created by them to know their abundance and distribution. This study revealed the rare cell type that expressed high levels of cystic fibrosis transmembrane conductance regulator (CFTR), the mutation of which causes cystic fibrosis. The scientists named these cells pulmonary ionocytes owing to their resemblance with the ionocytes found in freshwater fishes and frogs, which regulate salt balance. Pulmonary ionocytes make up 1-2% of the airway cells. They move ions at the interface between tissues and the air with help of CFTR protein in their membrane. The function of CFTR protein is to regulate the passage of chloride ions through the membrane of secretory epithelia of the lungs, which produces mucous and sweat. The mutation of the CFTR gene causes cystic fibrosis, which is characterised by excessive loss of sweat electrolytes, bronchiectasis, etc. This dysfunction of CFTR causes abnormal ion transport, i.e. altered salt and water movement across the epithelia which obstructs the hydration and ionic composition of mucous secretion in the lungs. This leads to the inflammation and destruction of the lungs. To understand better, the research team conducted further studies by disrupting a critical molecular process in the pulmonary ionocytes in mice. As a result, the cells started showing some features of cystic fibrosis. The experiment resulted in the formation of dense mucous in the airways of the mice. The discovery of pulmonary ionocytes has opened the way for scientists to study more about the treatment and therapeutic strategies against cystic fibrosis and gene therapy to correct CFTR gene mutation. One of the strategies that scientists came up with to treat cystic fibrosis was to increase the number of pulmonary ionocytes in order to increase the amount of CFTR activity. The studies hence revealed the importance of the ionocytes in airway-surface regulation.

Keywords: CFTR, Cystic fibrosis transmembrane conductance regulator, Pulmonary ionocytes, Cystic fibrosis, Single-cell RNA sequencing

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