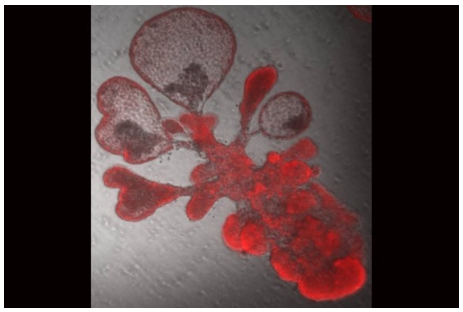
Cell and animal models of pulmonary fibrosis

Unmet Need

Pulmonary fibrosis is an irreversible and progressive disease characterized by the buildup of fibrotic material in the lungs, reducing lung function. There are over 200 different types of pulmonary fibrosis resulting from numerous causes such as exposure to medication, environmental factors, genetics and more. One of these is idiopathic pulmonary fibrosis (IPF), a complex disease with unknown etiology. IPF affects 1 in 200 adults over the age of 70– in the United States, IPF affects approximately 250,000 individuals, with 50,000 new cases and 40,000 deaths annually. Since the cause of disease is unknown, current treatments for IPF only slow disease progression, but ultimately a lung transplant is required. There is a need for a system to better understand the disease etiology of pulmonary fibrosis and identify potential treatments.

Technology

Duke inventors have developed cell and animal models of pulmonary fibrosis. This is intended to be used as a platform for drug discovery and toxicity assessment. Specifically, Duke inventors have developed an *ex vivo* lung fibrosis organoid model composed of co-cultured alveolar fibroblasts and pre-alveolar type-1 transitional state (PATs) cells, a distinct cellular population found between the alveolar type-1 and type-2 epithelial cells. PATs cells are enriched in human fibrotic lungs and have been implicated in the initiation and progression of the disease. Additionally, Duke inventors have developed an *in vivo* murine model by ablating the alveolar type-1 epithelial cells to initiate lung fibrosis and emphysema. Both models enable direct assessment of potential treatments *ex vivo* or assess the effects of a potential injurious agent given *in vivo*.



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Meet the Inventors

Tata, Purushothama "Tata"
Kobayashi, Yoshihiko "First name: □□ Family name: □□"
Konkimalla, Arvind "Arvind"
Tata, Aleksandra

Contact For More Info

Ferguson, Christy
919-681-7581
christy.ferguson@duke.edu

Department

Cell Biology

Publication(s)

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External Link(s)

• From the lab of Dr. Purushothama Rao Tata

This technology has been demonstrated in human tissues where PATs cells were associated with defective fibrotic foci. This technology demonstrated that PATs cells induce fibrosis by stimulating the surrounding cells into a pathogenic state. This was also demonstrated in both a murine model of injury by bleomycin and a murine model of genetic susceptibility that reflects human disease progression.

Other Applications

This technology can also be used in conjunction with another Duke technology for producing 3D cultures of alveolopheres (T-006618). Duke inventors have developed a method of producing *ex vivo* cellular cultures from both mouse and human pulmonary tissues that recapitulate the alveolar compartment, comprising a chemically defined media that allows for cells to be grown in a serum free and feeder free manner as well as an extracellular matrix to allow for the proper formation of 3D structure. Together, these technologies can be used as a platform for pulmonary drug discovery.

Advantages

- A novel disease modeling platform for pulmonary fibrosis.
- A novel mouse model that expands the PATs cell population to allow for an *ex vivo* lung fibrosis model that genetically recapitulates human disease progression.
- The platform is scalable.