



# Cystic Fibrosis Research News

#### Title:

INTERLEUKIN-1 IS ASSOCIATED WITH INFLAMMATION AND STRUCTURAL LUNG DISEASE IN YOUNG CHILDREN WITH CYSTIC FIBROSIS

### **Authors:**

Samuel T. Montgomery<sup>1</sup>, A. Susanne Dittrich<sup>2,3</sup>, Luke W. Garratt<sup>4</sup>, Lidija Turkovic<sup>4</sup>, Dario L. Frey<sup>2,3</sup>, Stephen M. Stick<sup>1,4,5,6</sup>, Marcus A. Mall<sup>2,7,8</sup>, Anthony Kicic<sup>1,4,5,6,9</sup>, & AREST CF<sup>1,5,10,11</sup>.

#### **Affiliations:**

<sup>1</sup>School of Paediatrics and Child Health, University of Western Australia, Nedlands, 6009, Western Australia, Australia.

<sup>2</sup>Department of Translational Pulmonology, Translational Lung Research Center Heidelberg (TLRC), German Center for Lung Research (DZL), University of Heidelberg, Heidelberg, Germany;

<sup>3</sup>Department of Pneumology and Critical Care Medicine, Thoraxklinik at the University Hospital Heidelberg, Heidelberg, Germany

<sup>4</sup>Telethon Kids Institute, University of Western Australia, Nedlands, 6009, Western Australia, Australia.

<sup>5</sup>Department of Respiratory Medicine, Princess Margaret Hospital for Children, Perth, 6001, Western Australia, Australia.

<sup>6</sup>Centre for Cell Therapy and Regenerative Medicine, School of Medicine and Pharmacology, University of Western Australia, Nedlands, 6009, Western Australia, Australia.

<sup>7</sup>Department of Pediatric Pulmonology and Immunology, Charité-Universitätsmedizin Berlin, Berlin, Germany

<sup>8</sup>Berlin Institute of Health (BIH), Berlin, Germany

<sup>9</sup>School of Public Health, Curtin University, Bentley, 6102, Western Australia, Australia.

<sup>10</sup>Murdoch Children's Research Institute, Parkville, 3052, Melbourne, Victoria, Australia

<sup>11</sup>Department of Paediatrics, University of Melbourne, Parkville, 3052, Melbourne, Victoria, Australia

## What was your research question?

Does lung inflammation (which is caused by mucus blockage in the airways) occur early in life in young children with Cystic Fibrosis (CF)? In addition, is it related to CF inflammatory markers we already know about and is there a relationship with lung damage.





# **Cystic Fibrosis Research News**

## Why is this important?

Early diagnosis and treatment in CF has improved patient outcomes, but lung disease which has been caused by inflammation still causes most CF-related complications. Often this inflammation is present without infection in the airways, however evidence from other diseases has shown that this happens when cells die due to lack of oxygen. Recent studies have suggested that thick mucus present in CF airways results in a lack of oxygen in airway cells, causing cell death and inflammation. However, these studies were performed in mouse models and there is a lack of data investigating this relationship in young children with CF.

## What did you do?

We took airway samples from young children with CF with and without lung infections and measured specific inflammatory markers including interleukin-1alpha, interleukin-8, neutrophils and neutrophil elastase in the samples. We then investigated how the inflammatory markers we had measured related to the extent of lung damage which we measured using lung imaging.

### What did you find?

In the airways of children both with and without infection, we found a connection between interleukin-1alpha and interleukin-8 and also between interleukin-1alpha and neutrophils. In the airways of children with respiratory infection we also found a connection between interleukin-1alpha and neutrophil elastase. In the airways of children without infection, we found connections between the amount of lung damage and interleukin-1alpha, interleukin-8, and neutrophil elastase. Finally, the relationship between lung damage and interleukin-1alpha was the strongest that we measured.

#### What does this mean and reasons for caution?

This study measured interleukin-1alpha early in life in young children with CF, and found a relationship between interleukin-1alpha and lung damage. This suggests interleukin-1alpha plays a role in lung damage in CF, and confirms the earlier findings from animal models. However, although our work found connections between the inflammatory markers and lung damage it doesn't mean interleukin-1alpha directly causes the lung damage, but it may contribute to the damage in some way. Further experiments are need to test this specifically.





# Cystic Fibrosis Research News

#### What's next?

Although we found connections between inflammatory markers and lung damage, we need to investigate exactly how cell death and inflammation cause this lung damage. We will do this using airway cells from young children with CF.

# Original manuscript citation in PubMed

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