

# Cystic Fibrosis Research News

**Title:**

Ventilation and perfusion assessed by functional MRI in children with CF: reproducibility in comparison to lung function

**Authors:**

Sylvia Nyilas<sup>1,2</sup>, Grzegorz Bauman<sup>3,4</sup>, Orso Pusterla<sup>3,4</sup>, Kathryn Ramsey<sup>1</sup>, Florian Singer<sup>1</sup>, Enno Stranzinger<sup>2</sup>, Sophie Yammine<sup>1</sup>, Carmen Casaulta<sup>1</sup>, Oliver Bieri<sup>3,4</sup>, Philipp Latzin<sup>1</sup>

**Affiliations:**

<sup>1</sup>Pediatric Respiratory Medicine, Department of Pediatrics, Inselspital, Bern University Hospital, University of Bern, Switzerland

<sup>2</sup>Department of Diagnostic, Interventional and Pediatric Radiology, Inselspital, Bern University Hospital, University of Bern, Switzerland

<sup>3</sup>Department of Radiology, Division of Radiological Physics, University of Basel Hospital, Basel, Switzerland

<sup>4</sup>Department of Biomedical Engineering, University of Basel, Basel, Switzerland

**What was your research question?**

Chronic lung diseases can be monitored by imaging and lung function tests. Magnetic resonance imaging (MRI) techniques allow clinicians to estimate how much lung tissue is impaired in ventilation or perfusion in specific areas of the lungs. In this study, we examined short-term variability of this impairment using MRI scans and compared the results to lung function measurements in children with cystic fibrosis (CF).

**Why is this important?**

Children diagnosed with CF usually do not show any respiratory symptoms in the first few years of life. Radiological diagnostic tests like the CT scan can detect early signs of lung disease. However, a disadvantage with this test is a high radiation dose, particularly when used annually. Therefore, there is an urgent need for imaging which doesn't use radiation (such as MRI) to detect lung abnormalities. Furthermore, since newborn screening for CF is now commonly used in most countries, CF will be diagnosed in more people at a very young age; this further highlights the importance for tests to identify early lung changes using a radiation-free and non-invasive test.

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## What did you do?

We carried out an observational study at the University Children's Hospital of Bern, Switzerland between February 2016 and September 2017. We enrolled 23 children with CF aged from 6 to 18 years, irrespective of bacterial colonisation or antibiotic use to ensure a broad range of disease severity. Eligibility criteria included a confirmed diagnosis of CF. 12 children without CF who had no history of chronic lung disease or acute respiratory infection prior to the investigations were also included. Participants underwent lung function measurements and MRI scans, in that order, on the same day. The assessment was repeated 24 hours later.

## What did you find?

This study comprehensively assessed short-term repeatability of functional MRI indices, indicating the amount of lung tissue that is less good ventilated or perfused. We found that in the short-term the consistency of functional MRI indices is very good in children with CF and also good in children without CF. Provision of data on the reproducibility and the clinically meaningful difference in ventilation and perfusion impairment are necessary so that patients' response to therapeutic interventions can be monitored and novel therapeutic mechanisms can be identified.

We also confirm previously reported excellent associations between commonly used lung function test results as established markers of lung disease severity in CF and MRI indices. Furthermore, functional imaging using MRI is feasible in school-aged children and the results are available in an average of only seven minutes.

## What does this mean and reasons for caution?

The functional MRI technique represents an attractive non-invasive method to clinically monitor children with CF. In this study we did not examine the variability of MRI indices over a longer time period, thus we cannot comment on this. Further research is needed to identify how the indices change in the presence of exacerbations and/or after an interventional treatment.

## What's next?

Our results indicate that the combination of MRI and lung function tests can be used to non-invasively monitor the development and progression of lung impairment. Future studies and replication in a larger population with a wider range of disease severity will reveal if MRI indices can be used as trial end points for interventional clinical trials.



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