



# Cystic Fibrosis Research News

## **Title:**

**USE OF HYPERPOLARIZED HELIUM-3 MRI TO ASSESS RESPONSE TO IVACAFTOR TREATMENT IN PATIENTS WITH CYSTIC FIBROSIS**

## **Authors:**

Talissa A. Altes<sup>a</sup>, Mac Johnson<sup>b</sup>, Meredith Fidler<sup>b</sup>, Martyn Botfield<sup>b</sup>, Nicholas J. Tustison<sup>c</sup>, Carlos Leiva-Salinas<sup>d</sup>, Eduard E. de Lange<sup>d</sup>, Deborah Froh<sup>e</sup>, John P. Mugler III<sup>f</sup>

## **Affiliations:**

<sup>a</sup>Department of Radiology, One Hospital Drive, University of Missouri, Columbia, MO 65212, USA;

<sup>b</sup>Vertex Pharmaceuticals Incorporated, 50 Northern Ave Boston, MA 02210, USA;

<sup>c</sup>University of Virginia, 480 Ray C. Hunt Drive, Snyder Building, Office 124 Charlottesville, VA 22903, USA;

<sup>d</sup>University of Virginia, University Hospital, First Floor 1215 Lee St., Charlottesville, VA 22908, USA;

<sup>e</sup>University of Virginia, 100 Hospital Dr #5408, Charlottesville, VA 22903, USA;

<sup>f</sup>University of Virginia, Snyder Translational Research Building – Room 154, 480 Ray C. Hunt Drive, Charlottesville, VA 22903, USA

## **What was your research question?**

Can a new imaging technique called helium-3 magnetic resonance imaging (MRI) be used to detect changes in the lungs of people with cystic fibrosis (CF) aged 12 years and older following treatment with a CF drug called ivacaftor?

## **Why is this important?**

Helium-3 MRI shows where air goes in the lungs during breathing. By taking pictures of the lungs of someone with CF before starting ivacaftor and comparing those pictures with pictures taken after ivacaftor treatment, one can see how the lung responds to the medication. The ability to take and compare these pictures is useful for testing new treatments for CF including ivacaftor. In the future, helium-3 MRI may also be able to help individuals manage their disease.

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[cfresearchnews@gmail.com](mailto:cfresearchnews@gmail.com)



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## What did you do?

We included people with CF aged 12 years and older who had a specific gene mutation (*G551D-CFTR*) that causes CF in the study. Everyone underwent medical tests before and after taking ivacaftor, including a sweat test (people with CF have higher levels of salt in their sweat), spirometry (which measures how well lungs are working), laboratory tests, and helium-3 MRI. Two groups of people were studied: Group A took ivacaftor for 4 weeks and Group B took ivacaftor for 48 weeks. The helium-3 MRI pictures from each person were looked at by a physician and analyzed by computer.

## What did you find?

The forced expiratory volume in 1 second ( $FEV_1$ ), a standard measure of lung function, improved after ivacaftor treatment in Group A (8 participants) by an average of 12.8 percentage points and in Group B (9 participants) by an average of 5.2 percentage points. Helium-3 MRI showed a tendency to improve in Group A and significantly improved in Group B when analyzed by computer programs. As demonstrated in an example below, the pictures of the lung from helium-3 MRI in one person from Group A show air flow before taking ivacaftor, after 4 weeks of treatment, and then 2 weeks after the person was off treatment. Areas of the lung with good air flow appear bright white, and those areas with poor air flow appear dark. Helium-3 MRI of this person showed improved air flow following ivacaftor treatment.

## What does this mean and reasons for caution?

This study confirmed the findings of earlier studies, showing that in people with CF and the *G551D-CFTR* mutation,  $FEV_1$  improved following treatment with ivacaftor. Helium-3 MRI clearly showed improved air flow in the lungs of people treated with ivacaftor. This finding suggests that helium-3 MRI has the potential to be useful in assessing lung disease in people with CF.

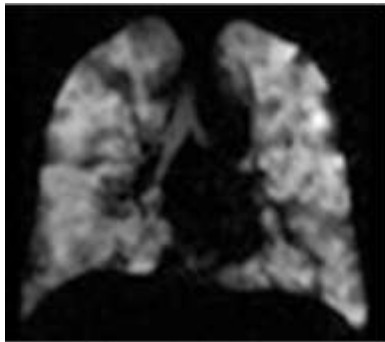
## What's next?

Although helium-3 MRI is in the early stages of development, this study has shown that it has the potential to be useful in looking at the effects of drug treatments on the lungs. Further studies are needed to determine whether helium-3 MRI could be used in individual people with CF to see how well different treatments are working for them.

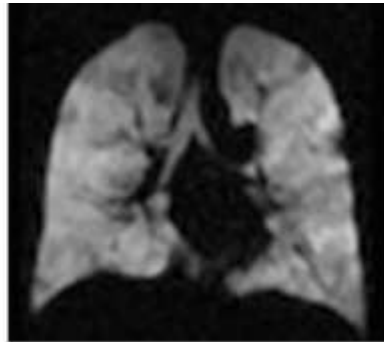
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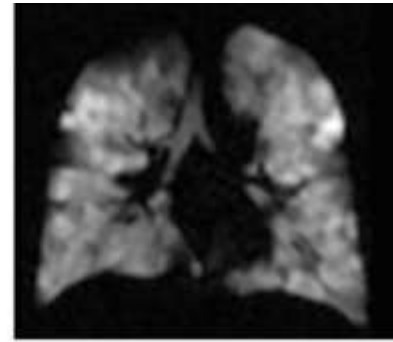
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Before ivacaftor



After 4 weeks on  
ivacaftor



After 2 weeks off  
ivacaftor

Helium-3 MR images of the lungs in a person immediately before starting ivacaftor, after 4 weeks on ivacaftor, and 2 weeks after discontinuing ivacaftor. A lung with good air flow looks bright, and a lung with poor air flow looks dark on these images. After 4 week on ivacaftor, the lungs appear much whiter, suggesting a much better air flow.

### Original manuscript citation in PubMed

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