European Cystic Fibrosis Society

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Cystic Fibrosis Research News

Title:

Magnetic Resonance Imaging Detects Improvements of Pulmonary and Paranasal Sinus Abnormalities in Response to Elexacaftor/Tezacaftor/Ivacaftor Therapy in Adults with Cystic Fibrosis

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Magnetic Resonance Imaging Detects Improvements of Pulmonary and Paranasal Sinus Abnormalities in Response to Elexacaftor/Tezacaftor/Ivacaftor Therapy in Adults with Cystic Fibrosis

Authors:

Lena Wucherpfennig[1,2,3], Simon M. F. Triphan[1,2,3], Sabine Wege[4], Hans-Ulrich Kauczor[1,2,3], Claus P. Heussel[1,2,3], Niclas Schmitt[5], Felix Wuennemann[1,2,3], Victoria L. Mayer[1,2], Olaf Sommerburg[2,6], Marcus A. Mall[2,6,7,8,9], Monika Eichinger[1,2,3*], Mark O. Wielpütz[1,2,3*]

* These authors contributed equally as last authors.

Affiliations:

1. Department of Diagnostic and Interventional Radiology, Subdivision of Pulmonary Imaging, University Hospital Heidelberg, Im Neuenheimer Feld 420, 69120 Heidelberg, Germany
2. Translational Lung Research Center Heidelberg (TLRC), German Center for Lung Research (DZL), Im Neuenheimer Feld 156, 69120 Heidelberg, Germany
3. Department of Diagnostic and Interventional Radiology with Nuclear Medicine, Thoraxklinik at University Hospital Heidelberg, Röntgenstr. 1, 69126 Heidelberg, Germany
4. Department of Pulmonology and Respiratory Medicine, Cystic Fibrosis Center, Thoraxklinik at University Hospital Heidelberg, Röntgenstr. 1, 69126 Heidelberg, Germany
5. Department of Neuroradiology, University Hospital Heidelberg, Im Neuenheimer Feld 400, 69120 Heidelberg, Germany
6. Department of Translational Pulmonology and Division of Pediatric Pulmonology and Allergy and Cystic Fibrosis Center, University of Heidelberg, Heidelberg, Germany
7. Department of Pediatric Respiratory Medicine, Immunology and Critical Care Medicine, Charité - Universitätsmedizin Berlin, corporate member of Freie Universität Berlin and Humboldt-Universität zu Berlin, Berlin, Germany
8. German Center for Lung Research (DZL), associated partner site, Berlin, Germany
9. Berlin Institute of Health at Charité – Universitätsmedizin Berlin, Berlin, Germany

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What was your research question?

The novel triple combination therapy of Elexacaftor/Tezacaftor/Ivacaftor was recently approved for treatment of adult cystic fibrosis (CF) patients. This therapy was shown to improve lung function and symptoms of chronic rhinosinusitis. The aim of our study was to assess the effects of the therapy by magnetic resonance imaging (MRI).

Why is this important?

MRI is a non-radiative, non-invasive imaging technique which can identify potentially reversible and irreversible lung abnormalities and characteristics changes in the paranasal sinuses in patients with CF. MRI of the chest and paranasal sinuses could show which areas and to what extent these CF-related abnormalities improve under novel therapies. MRI could therefore be a useful imaging tool for monitoring disease progression during therapy and for endpoint data in further clinical trials.

What did you do?

19 adults with CF (19-55 years) underwent standardized chest MRI, and nine additional sinonasal MRIs during the same session, before therapy and then after commencing Elexacaftor/Tezacaftor/Ivacaftor therapy for at least month. Additionally, 24 CF control patients (20-44 years) not taking Elexacaftor/Tezacaftor/Ivacaftor underwent longitudinal chest MRIs, and of them eleven also completed two sinonasal MRIs. Each MRI was assessed using a validated chest MRI score and chronic rhinosinusitis-MRI score. Moreover, lung function was measured by spirometry.

What did you find?

In patients receiving treatment with Elexacaftor/Tezacaftor/Ivacaftor, the chest MRI scores improved. In contrast, the chest MRI scores were unchanged in patients not taking Elexacaftor/Tezacaftor/Ivacaftor. The improvement with the novel triple combination therapy was mainly seen through a reduction of bronchiectasis/wall thickening and mucus plugging and correlated with an improvement in lung function. The chronic rhinosinusitis-MRI score also improved in patients receiving treatment with Elexacaftor/Tezacaftor/Ivacaftor, mainly seen in a reduction of mucopyoceles in the maxillary and ethmoid sinus, while the score was unchanged in patients not taking Elexacaftor/Tezacaftor/Ivacaftor.

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What does this mean and reasons for caution?

These results show that MRI of the chest and paranasal sinuses can depict improvements in adults under Elexacaftor/Tezacaftor/Ivacaftor therapy. Thus, MRI could be used for further studies as an endpoint assessing novel therapies as a combined examination of the upper and lower airways.
Nevertheless, the number of patients in our study treated with the novel triple combination therapy was only moderate and the number of patients who underwent paranasal sinus MRI was low.

What's next?

Further studies with larger numbers of patients should be carried out to confirm our results.

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