

Cystic Fibrosis Research News

Journal of

stic Fibrosis

The Official Journal of the European Cystic Fibrosis Society

Title:

ELEXACAFTOR-TEZACAFTOR-IVACAFTOR IMPROVES SINONASAL OUTCOMES IN CYSTIC FIBROSIS

Lay Title:

ELEXACAFTOR-TEZACAFTOR-IVACAFTOR IMPROVES SINUS PROBLEMS IN CYSTIC FIBROSIS

Authors:

Amanda L. Stapleton^a, Adam J. Kimple^b, Jennifer L. Goralski^c, S.M. Nouraie^d, Barton F Branstetter^{aef}, Amber D Shaffer^a, Joseph M Pilewski^d, Brent A Senior^b, Stella E Lee^{ag}, Anna C. Zemke^d.

Affiliations:

- a. Department of Otolaryngology Head and Neck Surgery, University of Pittsburgh
- b. Department of Otolaryngology Head & Neck Surgery, University of North Carolina
- c. Division of Pulmonary Diseases & Critical Care Medicine, University of North Carolina
- d. Department of Medicine, Division of Pulmonary, Allergy & Critical Care Medicine, University of Pittsburgh
- e. Department of Radiology, University of Pittsburgh
- f. Department of Biomedical Informatics, University of Pittsburgh
- g. Current Affiliation: Division of Otolaryngology—Head & Neck Surgery, Brigham and Women's Hospital, Harvard Medical School

What was your research question?

Does the new cystic fibrosis drug elexacaftor/texacaftor/ivacaftor [Trikafta] help sinus problems caused by CF?

Why is this important?

About two thirds of people with CF have symptoms of sinus disease that impact their quality of life, and nearly everyone with CF has sinus disease that is detectable on a CT scan. Sinus problems include a stuffed nose, nasal drainage, loss of the ability to smell, and headaches. Nasal polyps (extra tissue growing in the nose) may also be present and severe cases require surgery. Ongoing sinus problems may make lung function worse, and some people tend to have worsening of their sinus symptoms around the time of a pulmonary exacerbation. Finally, sinus problems continue after lung transplant, and bacteria from the sinuses may infect the transplanted lungs.

What did you do?

To answer the research question, we did a study with 34 people with CF and sinus problems at the University of Pittsburgh and University of North Carolina in the USA. The study participants were aged

Cystic Fibrosis Research News

cfresearchnews@gmail.com





Journal of

Cystic Fibrosis

The Official Journal of the European Cystic Fibrosis Society

between 12-60 years and one third of participants were teenagers. Study participants either had two F508 del mutations, or one F508 del mutation and one minimal function mutation. Before starting Trikafta, the study participants had an endoscopy (camera examination of their nose and sinuses) and a CT scan of their sinuses. A scratch and sniff type test was used to determine a participant's ability to smell and a symptom questionnaire was also completed. After 9 months, these measurements were repeated.

What did you find?

Over time about 90% of people had an improvement in their sinus symptoms, CT scans and endoscopies. The improvement in symptoms started within a week of starting Trikafta and lasted until at least 6 months. About half of the study participants had nasal polyps; however, the polyps shrunk in everyone and disappeared in most participants after starting on Trikafta. The only area where an improvement was not observed was the ability to smell (olfaction).

What does this mean and reasons for caution?

9 in 10 people who start Trikafta will have fewer sinus problems. About 10% of people in the study did not an have improvement in sinus symptoms or endoscopies and the reason for this finding was unknown. It might be that some people have sinus problems driven by allergies or some other cause in addition to CF. Everyone in our study had an improvement in lung function, weight, and sweat chloride measurements, therefore, a lack of improvement in sinus problems does not relate to how other CF symptoms will change with treatment. At this point, we do not know if the general lack of improvement in olfaction might take longer than 9 months, or if Trikafta would protect the ability to smell if started earlier in childhood.

What's next?

We are currently doing a similar study in a younger group (children aged 6-12 years) who have started Trikafta and we will determine if Trikafta their sinus problems improve. Another group of researchers is following adults for two years to determine if the improvement in sinus symptoms lasts for a longer period after starting Trikafta.

Cystic Fibrosis Research News

cfresearchnews@gmail.com



Cystic Fibrosis Research News

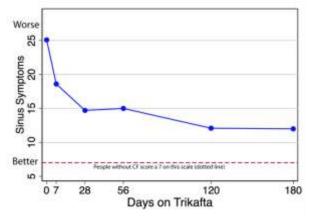
Journal of

Cystic Fibrosis

The Official Journal of the European Cystic Fibrosis Society

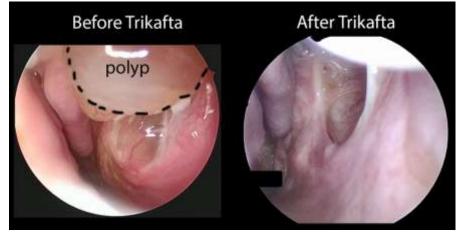
Optional Figure Caption 1:

People in the study reported their sinus symptoms after starting Trikafta. Higher numbers are worse symptoms. Within a week, people reported fewer symptoms.



Optional Figure 2

People in the study had pictures taken from inside their nose with a small camera. Before Trikafta, this person had a large polyp that went away after taking Trikafta. Before Trikafta, there was also more mucus and the inside of the nose was more swollen.



Original manuscript citation in PubMed https://pubmed.ncbi.nlm.nih.gov/35300931/

Cystic Fibrosis Research News

cfresearchnews@gmail.com