Cystic Fibrosis Research News

Title:
Effect of highly effective modulator treatment on sinonasal symptoms in cystic fibrosis

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What was your research question?
Does treatment with elexacaftor/tezacaftor/ivacaftor improve sinus symptoms and breathing related thus enhancing quality of life in adults with CF?

Why is this important?
Sinus disease is very common in people with CF and leads to significant complaints. To date, clinical trials of elexacaftor/tezacaftor ivacaftor did not include assessment of effect on sinus disease. CF modulator therapy may have a favourable impact on sinus symptoms.

What did you do?
We conducted a study of adults (18 years of age and above) with CF to determine the effect of elexacaftor-tezacaftor-ivacaftor on sinus symptoms. Participants were evaluated before starting treatment with elexacaftor-tezacaftor-ivacaftor and again three months after start of treatment using a series of questionnaires with proven value. Questionnaires were completed electronically via a Qualtrics link (Qualtrics, Provo, UT).

What did you find?
43 participants completed the study; mean age was 34.0 years and mean BMI was 21.8 kg/m2. Thirty-three percent of participants had two copies of the F508del mutation; 29 were female. 23 participants (53%) were being treated with other CF transmembrane conductance (CFTR) modulators at the time of study participation. Sixty-seven percent of patients reported sinus surgery in the past. All participants experienced a clear (significant) improvement in sinus symptoms and symptoms of the lungs. All domains of the sinus symptoms questionnaires

improved. Patients previously taking CFTR modulators experienced a greater benefit both in sinonasal and lung symptoms.

What does this mean and reasons for caution?

Adults with CF have significant sinonasal symptom burden at baseline which improves after start of elexacaftor-tezacaftor-ivacaftor therapy. These benefits are greater in patients already taking CFTR modulator therapy before the introduction of elexacaftor-tezacaftor-ivacaftor. One limitation of this study is that there is no control group without CFTR modulators for comparison.

What's next?

Further studies are required to investigate the changes through other outcomes, including nasal endoscopy and CAT scan scores.

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