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Cystic Fibrosis Research News

Title:

Use of Cystic Fibrosis inhaled medication before and after Elexacaftor/Tezacaftor/Ivacaftor initiation

Lay Title:

Use of Cystic Fibrosis inhaled medication after Elexacaftor/Tezacaftor/Ivacaftor in comparison to before

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What was your research question?

In the present study we aimed to compare use of inhaled medications before and after Elexacaftor/Tezacaftor/Ivacaftor (ETI) initiation in three Cystic Fibrosis (CF) centers in Greece, where over 95% of the adult CF population of the country is followed.

Why is this important?

ETI has significantly improved various aspects of CF, including pulmonary function, quality of life and glucose tolerance. Although ETI is designed and approved to be used on top of standard treatment, evidence suggests reduction in use of inhaled medication after its initiation. People with CF (PwCF) have reported significant reductions in use of supportive therapies, in particular inhaled antibiotics. Furthermore the SIMPLIFY study showed that discontinuation of hypertonic saline or dornase alpha for six weeks did not lead to clinically meaningful differences in pulmonary function in PwCF under ETI with an very good FEV_1 (above 95%)

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What did you do?

Adults with CF who had completed one year on ETI were included and their data were compared to a year before, without ETI. Use of medication was assessed by the Medication Possession Ratio (MPR) for dornase alpha and inhaled antibiotics. The MPR was calculated as the ratio of the number of doses of a drug which were received from the pharmacy in one year by a participant, to the number of doses the participant was advised to receive the during the same year. The MPR of each medication during the year on ETI were compared to the year without ETI.

What did you find?

We evaluated 71 adults, 38 women and 33 men, with mean age of 31.8 years and moderately to severely impaired pulmonary function. After ETI initiation, a statistically significant improvement was observed in weight, pulmonary function parameters, *P. aeruginosa* colonization, days of hospitalization and exacerbations. Use of all inhaled medications together dropped from 63% to 43% of what participants were advised to receive. Use of inhaled antibiotics separately and all together as well as use of dornase-alpha also dropped significantly. Only 21% of participants had an MPR above 80% the year on ETI vs 38% in the year before.

What does this mean and reasons for caution?

Our results indicate a significant reduction in use of inhaled medication after ETI initiation in PwCF with moderately to severely impaired lung function, despite the strong encouragement of the CF teams for the continuation of pre-advised nebulized treatments. On the other hand improvement in various parameters of CF was observed. Our results are limited mainly by the retrospective nature of the study, the rather small sample size, and the use of MPR since supply of a medication from the pharmacy does not guarantee its use.

What's next?

The longitudinal clinical significance of inhaled treatments of the pre-ETI era after the initiation of CFTR modulators according to the pulmonary function and the microbiological status of PwCF remains to be seen in large prospective trials.

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