primary studies - published, non RCT

Real-World Impact of Elexacaftor/Tezacaftor/Ivacaftor on Health-Related Quality of Life in Adults with Cystic Fibrosis: A 12-Month Multicenter Cohort Study.

Code: PM42019640 **Year:** 2026 **Date:** **Author:** Kharraz R

Study design (if review, criteria of inclusion for studies)

Prospective cohort study

Participants

353 individuals with cystic fibrosis (CF).

Interventions

Elexacaftor/tezacaftor/ivacaftor (ETI)

Outcome measures

Health-related quality of life (HRQoL) was assessed using the Cystic Fibrosis Questionnaire-Revised (CFQ-R), Cystic Fibrosis Quality-of-Life Evaluative Self-Administered Test (CF-QUEST), and Sino-Nasal Outcome Test (SNOT-22) at baseline and at 3, 6, 9, and 12 months post-initiation. Lung function (percent predicted forced expiratory volume in one second [ppFEV1]) and BMI were also monitored. Individuals were classified as lung function responders (â‰¥5% increase in ppFEV1) or non-responders (

Main results

Among 353 individuals, significant improvements in CFQ-R, CF-QUEST, and SNOT-22 scores were observed within one month of ETI initiation and sustained over 12 months. Notably, both lung function responders and non-responders experienced significant HRQoL improvements, with no evidence of a difference between groups in terms of mean changes in CFQ-R respiratory scores (28.8 vs. 24.2, p=0.2), CF-QUEST global scores (14.8 vs. 13.0, p=0.48), or SNOT-22 scores (-13.8 vs. -13.7, p=0.97).

Authors' conclusions

Our real-world data involving a large cohort of PwCF demonstrate that HRQoL improvements are sustained across multiple domains regardless of lung function response. These findings underscore the importance of using a multimodal approach to evaluate the effectiveness of modulator therapy in people with CF.

http://dx.doi.org/10.1016/j.rmed.2026.108827

See also

Respir Med. 2026 Apr 20:108827. doi: 10.1016/j.rmed.2026.108827.

Keywords

Adult; Aged; CFTR Modulators; Genetic Predisposition to Disease; pharmacological_intervention; placebo; VX-770; VX-661; ivacaftor; Aminophenols; tezacaftor; VX-445; elexacaftor; Trikafta; kaftrio;