

primary studies - published, non RCT

Respiratory training with a specific device in cystic fibrosis: a prospective study.

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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

Individuals were classified as lung function responders ($\geq 5\%$ 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.jcf.2007.12.003>

See also

J Cyst Fibros. 2008 Jul;7(4):313-9. doi: 10.1016/j.jcf.2007.12.003. Epub 2008 Feb 1.

Keywords

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