primary studies - published, non RCT

Persistence of lung hyperinflation and small airway dysfunction in school-aged children with cystic fibrosis treated with elexacaftor-tezacaftor-ivacaftor: results from the real-world MODUL-CF study.

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

Data from a national, real-world cohort of school-aged chCF

Participants

Children with CF (chCF) treated with elexacaftor-tezacaftor-ivacaftor (ETI). A total of 192 chCF aged from 6 to 18â€…years at ETI initiation

Interventions

Elexacaftor-tezacaftor-ivacaftor (ETI)

Outcome measures

Data from multiple breath washout (MBW) and body plethysmography (pleth) from a national, real-world cohort of school-aged chCF. Data were collected from seven centres prior to ETI initiation (month 0 (M0)), after a year of treatment (M12) and at M6 (when available). Analyses were conducted to assess the evolution of per cent predicted (pp) residual volume (ppRV), total lung capacity (ppTLC) and the ppRV/TLC ratio, as well as the difference between pp functional residual capacity (ppFRC)(pleth) and ppFRC(MBW) as markers of TG. Global and regional VI indices were compared to those of age- and sex-matched healthy controls.

Main results

Significant reductions were observed in ppRV, ppTLC, ppRV/TLC and TG. However, 28% of the chCF cohort had ppRV/TLC values >120% at M12. MBW-derived outcomes improved significantly, but 51.9% of the cohort had lung clearance index (LCI(2.5)) values at M12 that had not returned to healthy control levels.

Authors' conclusions

ETI significantly improved SAF. However, not all outcome measures returned to normal, indicating residual lung disease in some chCF.

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See also

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Keywords

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