primary studies - published, non RCT

Recovery of lung function following a pulmonary exacerbation in patients with cystic fibrosis and the G551D-CFTR mutation treated with ivacaftor.

Code: PM28651844 **Year:** 2017 **Date:** 2017 **Author:** Flume PA

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

Data summarized from a placebo-controlled trial

Participants

161 cystic fibrosis patients>/=12years old with the G551D-CFTR mutation

Interventions

Ivacaftor vs placebo.

Outcome measures

Pulmonary exacerbations (PEx). Short-term recovery was measured 2 to 8weeks after treatment, and long-term recovery was determined at the end-of-study, both compared with baseline measured just prior to the PEx.

Main results

Fewer patients receiving ivacaftor experienced a PEx than patients receiving placebo (33.7% vs. 56.4%; P=0.004) and had a lower adjusted incidence rate of PEx (0.589 vs. 1.382; P

Authors' conclusions

Ivacaftor treatment reduces the frequency of PEx but does not improve on the rate of complete lung function recovery after PEx when compared with placebo.

http://dx.doi.org/10.1016/j.jcf.2017.06.002

See also

J Cyst Fibros. 2017 Jun 24. pii: S1569-1993(17)30770-1. doi: 10.1016/j.jcf.2017.06.002.

Keywords

Aminophenols; CFTR Modulators; Genetic Predisposition to Disease; pharmacological_intervention; VX-770; ivacaftor; G551D-CFTR;