

primary studies - published RCT

Efficacy and safety of ivacaftor in patients with cystic fibrosis and a non-G551D gating mutation

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

Randomised, controlled, crossover trial

Participants

14 adults with mild to severe CF lung disease (forced expiratory volume in 1 s % predicted 31-113%)

Interventions

20 min of resting breathing (control), treadmill exercise at 60% of the participant's peak oxygen consumption or PEP therapy (including huffing and coughing).

Outcome measures

Mucus clearance was measured using the radioaerosol technique and gamma camera imaging.

Main results

Treadmill exercise improved whole lung mucus clearance compared to resting breathing (mean difference 3%, 95% CI 2-4); however, exercise alone was less effective than PEP therapy (mean difference -7%, 95% CI -6- -8). When comparing treadmill exercise to PEP therapy, there were no significant differences in mucus clearance from the intermediate and peripheral lung regions, but significantly less clearance from the central lung region (likely reflecting the huffing and coughing that was only in PEP therapy).

Authors' conclusions

It is recommended that huffing and coughing are included to maximise mucus clearance with exercise.

<http://dx.doi.org/10.1016/j.jcf.2014.09.005>

See also

J Cyst Fibros. 2014 Dec;13(6):674-80.

Keywords

Aerobic training; Airway clearance technique; Chest physiotherapy; exercise; non pharmacological intervention - devices OR physiotherapy; Positive-Pressure Respiration- PEP- pep mask; Respiratory Tract Diseases; strength training; training;