primary studies - published, non RCT

The long-term effect of elexacaftor/tezacaftor/ivacaftor on cardiorespiratory fitness in adolescent patients with cystic fibrosis: a pilot observational study.

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

Single group prospective observational study

Participants

10 adolescent patients with cystic fibrosis who completed two CPET measurements between January 2019 and February 2023. During this period, elexacaftor/tezacaftor/ivacaftor treatment was initiated in all of them.

Interventions

Elexacaftor/tezacaftor/ivacaftor medication. Between January 2019 and February 2023 elexacaftor/tezacaftor/ivacaftor treatment was initiated.

Outcome measures

Cardiorespiratory fitness: peak workload, VO(2 peak), VO(2VT1), VO(2VT2), V(E)/VCO(2) slope, V(E), V(T), RQ, VO(2)/HR peak and RR peak.

Main results

We observed significant improvement in peak workload, VO(2 peak), VO(2VT1), VO(2VT2), V(E)/VCO(2) slope, V(E), V(T), RQ, VO(2)/HRÂ peak and RRÂ peak. The mean change in VO(2Â)peakÂ was 5.7Â mL/kg/min, or 15.9% of the reference value (SDâ€‰Â±â€‰16.6;Â p=â€‰0.014). VO(2VT1) improved by 15% of the reference value (SDâ€‰Â±â€‰0.1;Â p=â€‰0.014), VO(2VT2Â)improved by 0.5 (SDâ€‰Â±â€‰0.4;Â p=â€‰0.01). There were no differences in other parameters.

Authors' conclusions

Exercise tolerance improved after elexacaftor/tezacaftor/ivacaftor treatment initiation. We suggest that the CFTR modulator alone is not enough for recovering physical decondition, but should be supplemented with physical activity and respiratory physiotherapy. Further studies are needed to examine the effect of CFTR modulators and physical therapy on cardiopulmonary exercise tolerance.

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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; kaftrio;