# Cardiopulmonary exercise testing in young people with cystic fibrosis – methods, limitations, and clinical practice

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Cystic fibrosis (CF) is a genetically inherited disease that currently affects ~11,000 people in the UK. The disease presents with an accumulation of thick mucus that blocks the airways and digestive systems, and can reduce exercise tolerance and increase breathlessness. As there is currently no cure for CF, exercise forms an important treatment and regular exercise testing is recommended, with cardiopulmonary exercise testing (CPET) acknowledged as the 'gold standard' by international clinical organisations (Hebestreit et al., 2015). Aerobic fitness (as defined using peak oxygen uptake, $\dot{V}O_{2peak}$), a primary outcome of CPET, is significantly associated with mortality in CF (Vendrusculo et al., 2018), highlighting the need to accurately identify this value and understand pathophysiological causes behind any changes. However, there are several aspects surrounding its application that have warranted further investigation.

By its nature, maximal exercise testing requires maximal effort from patients, which is not always possible due to clinical status, breathlessness or motivation. As such, suitable submaximal parameters have warranted investigation. This programme of research investigated the oxygen uptake efficiency slope; firstly utilising allometric scaling to remove residual effects of body size (Tomlinson et al., 2017b), then subsequently determining its invalidity as a submaximal alternative to $VO_{2peak}$, as it is unable to discriminate fitness in the same way as $VO_{2peak}$ (Williams et al., 2018). However, we identified that the oxygen uptake efficiency plateau instead holds interesting potential, as it does not require scaling for body size, and is associated with disease status and severity in CF (Tomlinson et al., 2018a). Interestingly, these results contrast similar studies in heart failure, highlighting that clinical groups cannot be simply grouped together, but that each warrants its own dedicated avenue of research.

Furthermore, this programme of research has further utilised CPET, alongside magnetic resonance imaging, to investigate the musculoskeletal basis of exercise

intolerance in CF. This research identified that once thigh muscle volume has been fully quantified (not estimated) and mathematically scaled for, there remains a difference in $\dot{V}O_{2peak}$ between children with CF, and healthy matched controls (Tomlinson et al., 2017a). This therefore provides evidence towards a qualitative defect in skeletal muscle in CF, which has implications for management of the disease, in particular the selection of exercise training regimens to improve musculoskeletal health.

Finally, a clinical application of research findings is essential for the benefit of the end user – people with CF themselves. Integrating CPET into standard clinical care is a priority, and to fulfil this requirement, a working group of clinically-based exercise professionals has been established in the UK. This 'Cystic Fibrosis and Exercise Network' has an emphasis on continued professional development, meeting annually to exchange best practice. Surveys from this group reveals that exercise technicians (individuals typically trained in sport and exercise sciences) play an increasingly important role in clinics, responsible for exercise testing and training within CF centres. We have also been identified that one-third of clinical teams are not confident in discussing exercise with patients (Tomlinson et al., 2018b), thus highlighting an urgent need to improve, increase, and standardise exercise education for clinicians in both CF and beyond.

There is a promising future for applied exercise science in clinical environments, as well as an anticipated need for increased numbers of exercise scientists in clinical roles. As the importance of exercise in the management of CF continues to grow, as does the demand on both health care professionals and the exercise science community to collaborate and further integrate exercise into health care for chronic disease.

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