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MRI-based screening for metabolic insufficiency of leg muscle during aerobic exercise in Cystic Fibrosis

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There is evidence for mitochondrial dysfunction in various tissues in Cystic Fibrosis (CF) including muscle. Among others, a slow rate of high-energy phosphate resynthesis following exercise involving single limb muscle activity was found in human CF using in vivo ³¹P magnetic resonance spectroscopy (MRS). This raises the question whether this outcome would be ameliorated versus exacerbated if instead an exercise regime is used that puts a significant cardiopulmonary load on the body as in running or bicycling. This is of interest because exercise therapy is commonly prescribed in CF. To investigate this matter, ten pediatric CF patients (age 12–16 years) and healthy peers performed two ramp exercise tests to volitional exhaustion using a bicycle ergometer fit for use inside a MR scanner. Endurance, VO_{2max} and heart rate were determined in the exercise laboratory. Quadriceps muscle energy- and acid/base balance during exercise and recovery were measured on a separate day using MR imaging-based ³¹P MRS. This study brings together for the first time this powerful biomedical imaging platform and whole body exercise testing in the clinical setting of human CF.