Impairment of muscle glycogenolysis in McArdle's disease (myophosphorylase deficiency) leads to exercise intolerance and exercise-induced myalgia. The pathophysiology of these symptoms is not entirely clear. We used phosphorus magnetic resonance spectroscopy to measure muscle phosphate metabolite concentrations and intracellular pH during brief ischemic exercise and in the period of aerobic metabolic recovery after exercise, with special attention to cytoplasmic adenosine 5'-diphosphate (ADP). In 5 patients with McArdle's disease, calculated muscle intracellular ADP concentrations at the beginning of recovery were higher than in normal control subjects (70-425 mmol/L, control mean: 73 +/- 40 mmol/L, $P < 0.05$). The half-time for intracellular ADP recovery after exercise, an index of maximal mitochondrial oxidative phosphorylation, was 0.16 +/- 0.07 in normal controls and was independent of metabolic state or intracellular pH. ADP recoveries were abnormally slow in all patients with McArdle's disease (range: 0.32-0.83 min, mean = 0.2 min, $P < 0.0001$). These results are indicative of a limitation in the rate of oxidative phosphorylation in muscle of patients with McArdle's disease, most likely due to impaired substrate delivery to mitochondria. This impairment of mitochondrial function may contribute to the exercise-related symptoms in McArdle's disease [1)].

1)
De Stefano N, Argov Z, Matthews PM, Karpati G, Arnold DL. Impairment of muscle mitochondrial oxidative metabolism in McArdles's disease. Muscle Nerve. 1996 Jun;19(6):764-9. doi: 10.1002/(SICI)1097-4598(199606)19:6<764::AID-MUS12>3.0.CO;2-L. PMID: 8609928.