Supplementary Figure 1.

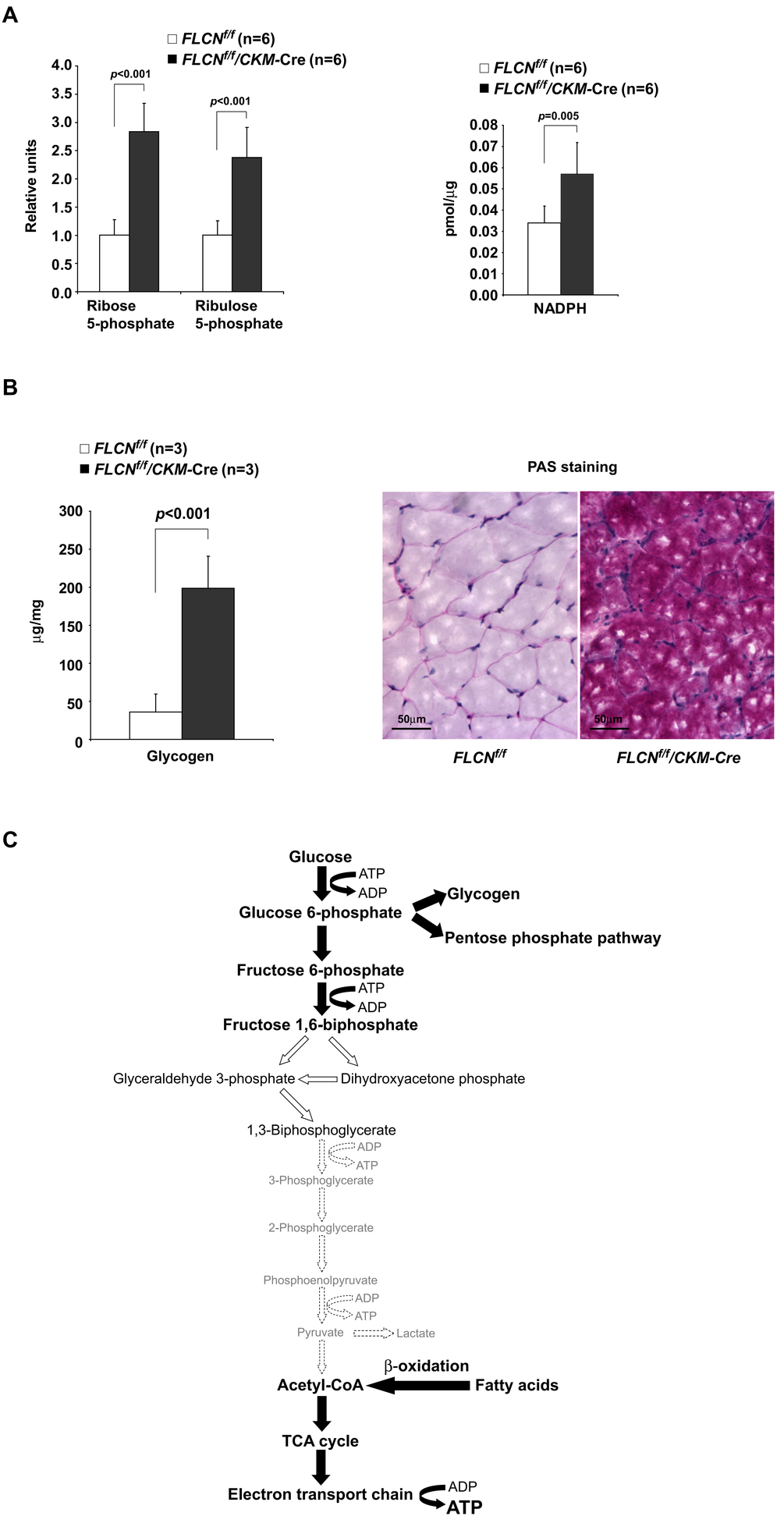

Figure S1. The glycolytic pathway, pentose phosphate pathway and glycogenesis in *FLCN*-deficient muscle tissues

(A) Metabolite profiling analyses using LC/MS and GC/MS were carried out with extracts from quadriceps muscle, and relative units of metabolites related to pentose phosphate pathway were obtained. Mean ± 95%CI. Two-sided student's *t*-test. Quadriceps, n=6. (Left panel). NADPH in quadriceps muscle was measured using a commercially available kit (Biovision, Milpitas, CA). Mean ± 95%CI. Two-sided student's *t*-test. Quadriceps, n=6. (Right panel).
(B) Glycogen in quadriceps muscle was measured using a commercially available kit (Sigma-Aldrich, St. Louis, MO). Mean ± 95%CI. Student's *t*-test. (Left panel). Quadriceps muscle was stained with PAS staining. Scale bar: 50μm.(Right panel)
(C) Scheme of metabolic pathway in *FLCN*-deficient muscle tissues.