

**Supplementary Table 2.** Metabolites in glycolytic pathway of *FLCN*-deficient quadriceps muscle\*

Metabolite	KO (n=6) / WT (n=6)	P †
Glucose	1.260	0.0004
Glucose 6-phosphate	4.581	0.0007
Fructose 6-phosphate	4.687	0.0006
Fructose 1,6-biphosphate	6.666	0.0001
3-Phosphoglycerate	0.066	0.0032
2-Phosphoglycerate	0.059	0.0028
Phosphoenolpyruvate	0.033	0.0049
Pyruvate	0.625	0.0133
Lactate	0.920	0.0159

\*Metabolite profiling analyses using LC/MS and GC/MS were carried out with extracts from quadriceps muscle, and KO (*FLCN<sup>ff</sup>/CKM-Cre*) / WT (*FLCN<sup>ff</sup>*) ratios of metabolites related to glycolytic pathway were obtained. Glucose, glucose 6-phosphate, fructose 6-phosphate and fructose 1,6-biphosphate are metabolites in the early glycolysis pathway, and 3-phosphoglycerate, 2-phosphoglycerate, phosphoenolpyruvate, pyruvate and lactate are metabolites in the late, three-carbon portion of the glycolysis pathway. KO = knockout. WT = wild-type.

† Student's *t*-test (two-sided).