

Supplemental Table 1. Body weight and body composition in male R6/2 mice

Genotype	WT				R6/2				
	Weeks	4	8	10	12	4	8	10	12
Body wt (g)		21.6±0.5	27.7±0.5	29.5±0.4	29.0±0.9	21.5±0.9	27.3±0.9	26.2±0.7***	25.6±0.6***
Fat wt (g)		1.0±0.1	1.6±0.1	2.0±0.1	3.0±0.2	1.4±0.1**	2.9±0.3***	3.1±0.3***	3.5±0.3
Fat %		4.3±0.3	5.8±0.4	6.6±0.4	10.2±0.7	6.3±0.5**	10.5±1.3***	11.8±1.3***	13.1±1.1*
Lean wt (g)		17.4±0.4	21.6±0.4	23.0±0.3	21.2±0.6	16.7±0.6	19.8±0.8*	18.6±0.7***	17.3±0.2***
Lean %		80.7±0.5	78.0±0.5	77.8±0.5	73.4±0.9	77.7±1.3*	72.1±1.7***	70.9±1.6***	65.1±3.3*
Fluid wt (g)		1.7±0.0	2.0±0.0	2.3±0.0	2.2±0.1	1.7±0.1	2.2±0.1	2.1±0.1*	2.0±0.0*

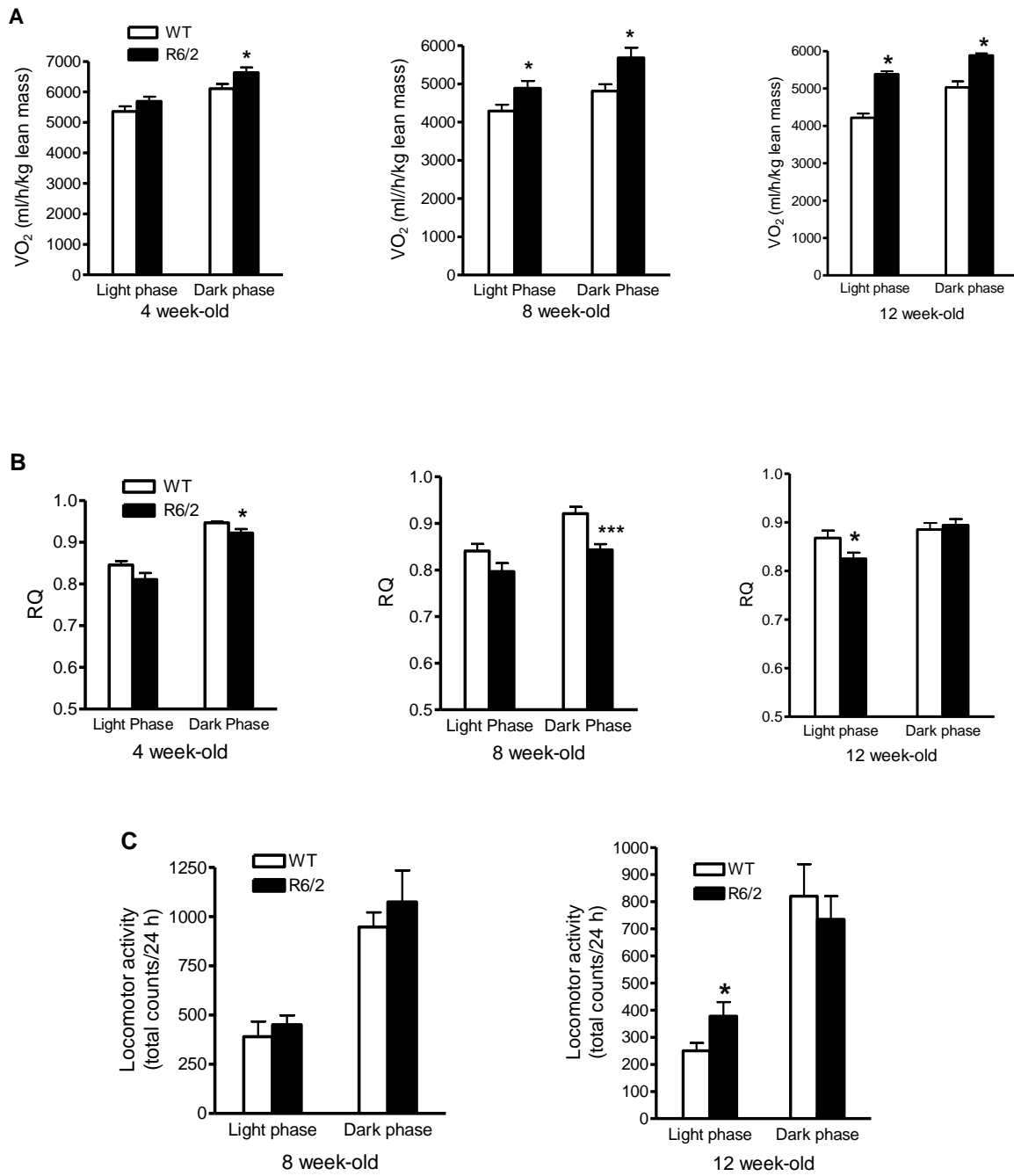
N=8 for R6/2 TG and 20 for WT, * $P < 0.05$, ** $P < 0.01$, and *** $P < 0.001$ vs. WT at the same age.

Supplemental Table 2. Body weight and body composition in female R6/2 mice

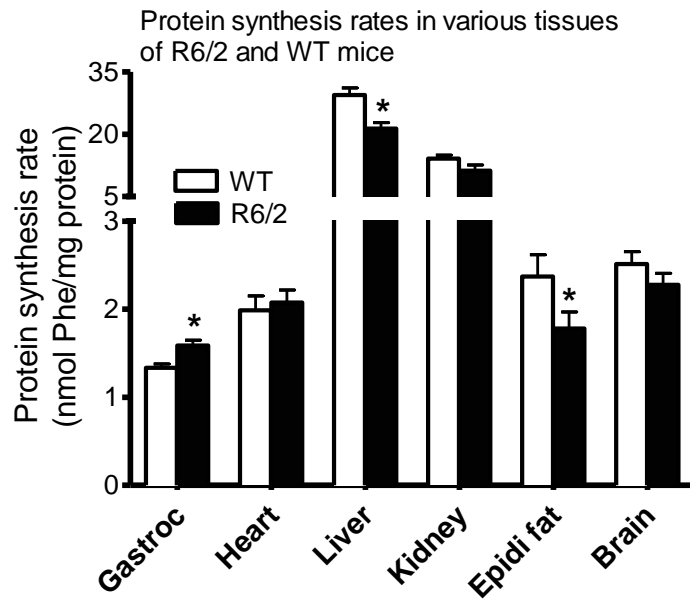
Genotype	WT				R6/2			
	4	8	10	12	4	8	10	12
Body wt (g)	18.1±0.4	21.4±0.6	23.8±0.7	26.3±1.0	18.0±0.7	24.0±0.8*	23.3±0.8	22.2±0.6***
Fat wt (g)	1.1±0.2	1.6±0.2	2.8±0.3	3.7±0.4	1.6±0.2	4.2±0.3***	3.5±0.3	2.5±0.3*
Fat %	5.4±1.1	7.4±0.8	11.6±1.0	13.2±1.2	8.4±0.7*	16.9±0.9***	14.7±0.8*	11.0±0.9
Lean wt (g)	14.3±0.3	16.1±0.4	17.0±0.3	18.0±0.4	13.8±0.4	15.6±0.3	15.9±0.4*	15.8±0.3***
Lean %	79.2±1.0	75.2±0.8	71.7±0.9	69.0±1.5	77.1±1.0	65.5±1.1***	68.9±0.9*	71.6±0.9
Fluid wt (g)	1.6±0.0	1.8±0.1	2.0±0.1	2.3±0.1	1.6±0.1	2.1±0.1	2.1±0.1	1.9±0.1***

N=16 for R6/2 TG and 12 for WT, * $P < 0.05$, ** $P < 0.01$, and *** $P < 0.001$ vs. WT at the same age.

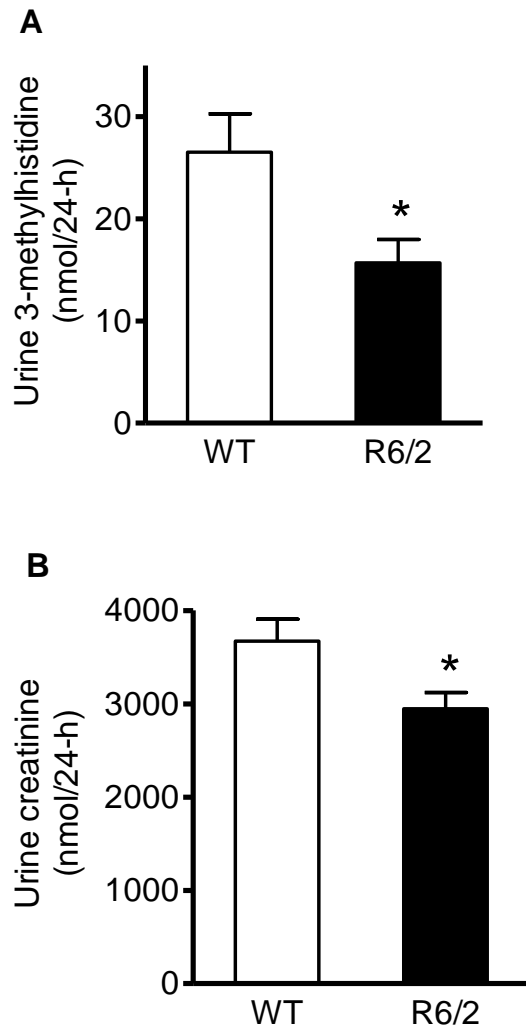
Supplemental material Figure 1



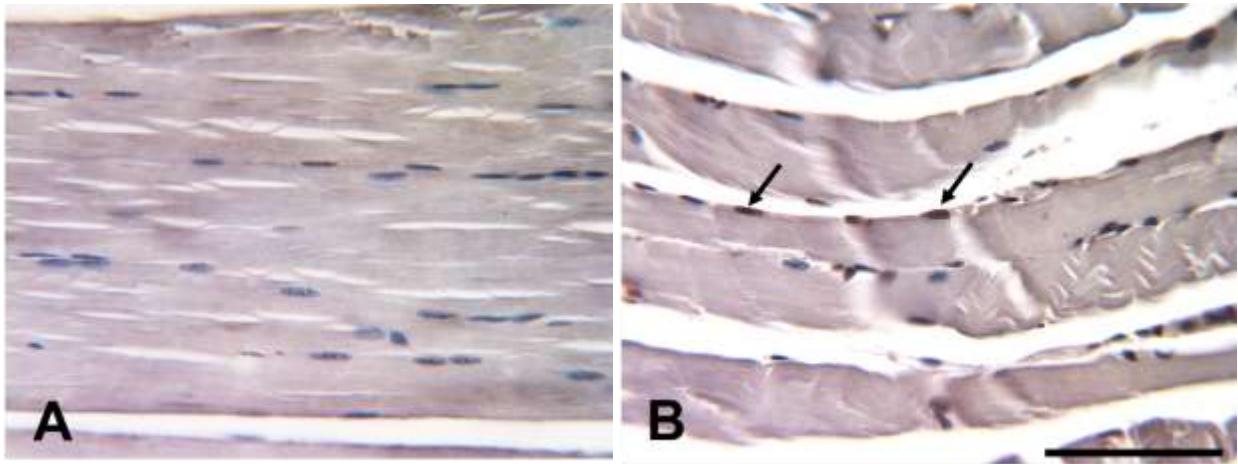
Supplemental material Figure 2



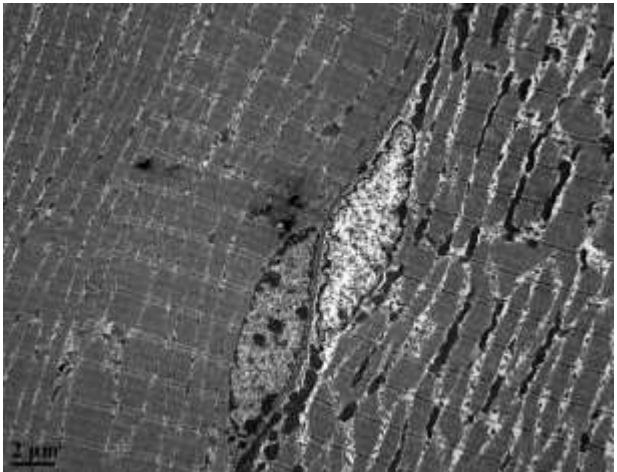
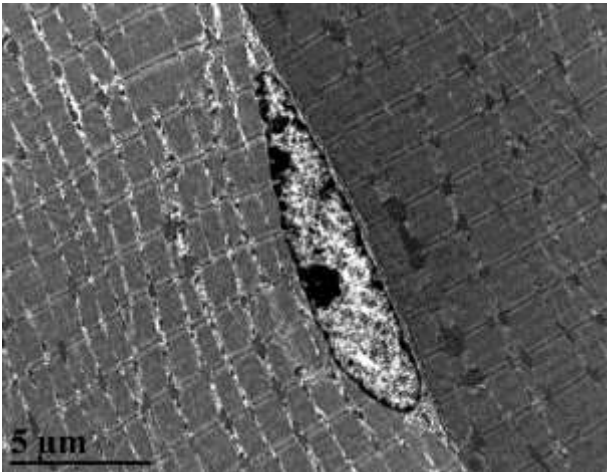
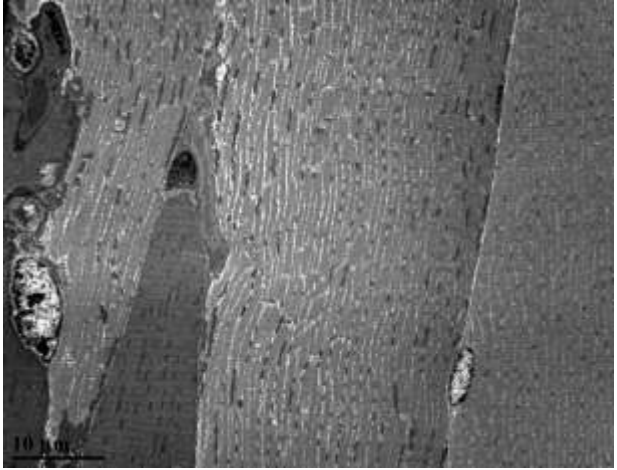
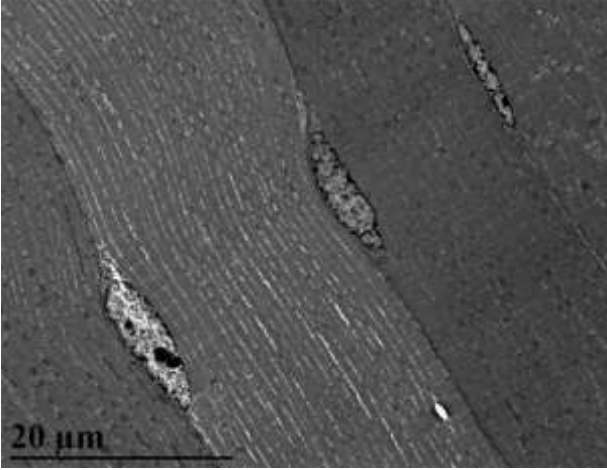
Supplemental material Figure 3



Supplemental material Figure 4



C



Supplemental material figure legends

Fig. 1 Oxygen consumption (A), RQ (B), and locomotor activity (C) in R6/2 and WT mice. * $P < 0.05$ vs. WT control, n=8-10.

Fig. 2. Protein synthesis rates in various tissues of R6/2 and WT mice. * $P < 0.05$ vs. WT control, n=10/group.

Fig. 3. Twenty hour-hr urinary secretion of 3-methylhistidine and creatinine in n R6/2 and WT mice. * $P < 0.05$ vs. WT control, n=10 for WT and 9 for R6/2 mice.

Fig. 4. TUNEL assay and electron micrographs of longitudinal or oblique sections of gastrocnemius muscle fibers of R6/2 mice. Representative fields of TUNEL stained gastrocnemius muscle sections. A. Control mouse. B. Knockout mouse. Note the prevalence of TUNEL+ skeletal muscle nuclei (brown; arrows) in B. Nuclei are counterstained blue. Scale bar represents 50 μ m. Representative electron micrographs of gastrocnemius sections of 12 weeks of age R6/2 (C). EM pictures with different magnifications and abnormal myocytes flanked by relatively normal myocytes in R6/2 mice were shown. n=2.