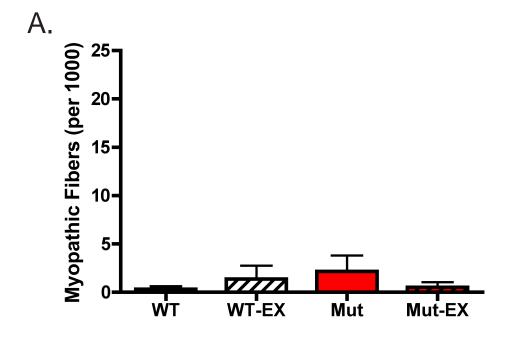
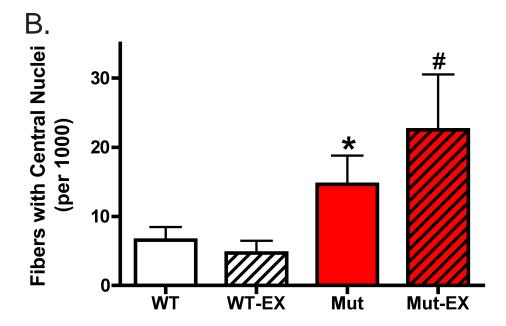
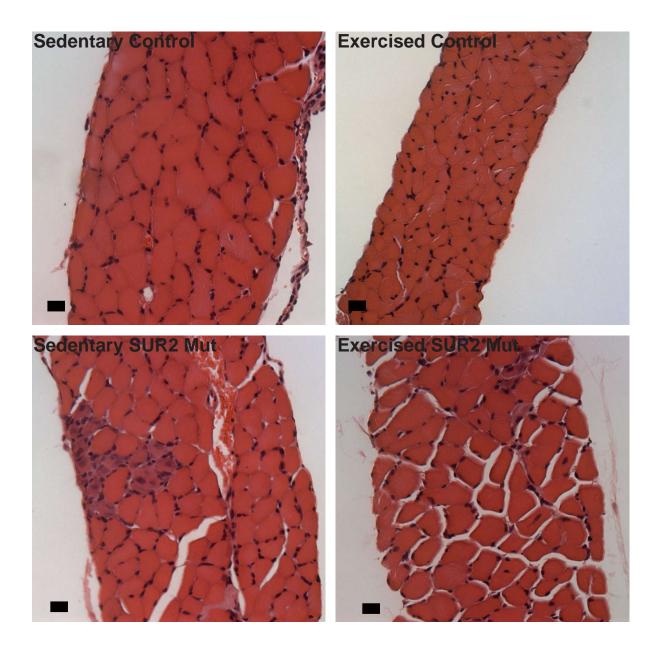


Supp Fig 1- TA





Supp Fig 2 - EDL



Supp Fig 3 - Diaphragm

Supplemental Figure 1. Evidence of myopathic in the tibialis anterior muscle of SUR2 mutant mice. Tibialis anterior muscle was isolated from sedentary and exercised cohorts of control (n=5 and 5, respectively) and SUR2 mutant mice (n=5 and 4, respectively) after the four week exercise period. A. Representative photomicrographs of paraffin-imbedded tibialis anterior muscle from sedentary and exercised cohorts of control and SUR2 mutant mice. Sedentary SUR2 mutant mice exhibit skeletal muscle myopathy that does not worsen with exercise. Myopathic fibers were typically present in foci throughout the muscle body. (Scale bar = $10 \mu m$). B. Quantification of myopathic muscle fibers. C. Quantification of fibers with central nuclei. * p<0.05 vs sedentary control, # p<0.05 vs exercised control by t-test. D. Quantification of total fiber number. E. Quantification of Fiber Size. * p<0.05 vs sedentary control by t-test.

Supplemental Figure 2. Evidence of myopathic in the extensor digitorum longus (EDL) muscle of SUR2 mutant mice. EDL muscle was isolated from sedentary and exercised cohorts of control (n=5 and 5, respectively) and SUR2 mutant mice (n=5 and 4, respectively) after the four week exercise period. Unlike Kir6.2 null mice, EDL muscles were not significantly more affected than other muscle groups, and actually displayed qualitatively less damage than quadriceps and tibialis anterior muscles. A. Quantification of myopathic muscle fibers. B. Quantification of fibers with central nuclei. * p<0.05 vs sedentary control, # p<0.05 vs exercised control both by t-test.

Supplemental Figure 3. Evidence of myopathic in the diaphragm muscle of SUR2 mutant mice. Diaphragm muscles from SUR2 mutant mice of both sedentary and exercised cohorts contained myopathic fibers and fibers of central nuclei of similar number. Myopathic fibers were most often present in small foci, while fibers with central nuclei were present usually along the periphery. No significant fibrosis or inflammatory infiltrate was observed in any sample. Because of limited tissue in some animals, quantitative analysis was not feasible as completed

in quadriceps, tibialis anterior, and EDL muscles. Representative photomicrographs of paraffinimbedded diaphragm from sedentary and exercised cohorts of control and SUR2 mutant mice are shown (Scale bar = $10 \mu m$).