

Supplemental Data

Animals had *ad lib* access to food and water throughout the study period. Nine or 10 mice of each gender and genotype were enrolled in each group. During the course of the study period, some animals were used for non-survival procedures (e.g *in vitro* contraction test) and therefore 3 to 4 animals remained in each group at the end of the study period. Body mass was measured weekly, starting at 4 weeks of age and continuing through 52 weeks. Grip strength (Chatillon meter, Columbus Instruments; Columbus, OH) was determined monthly as the average of five trials for each muscle group, in each animal.

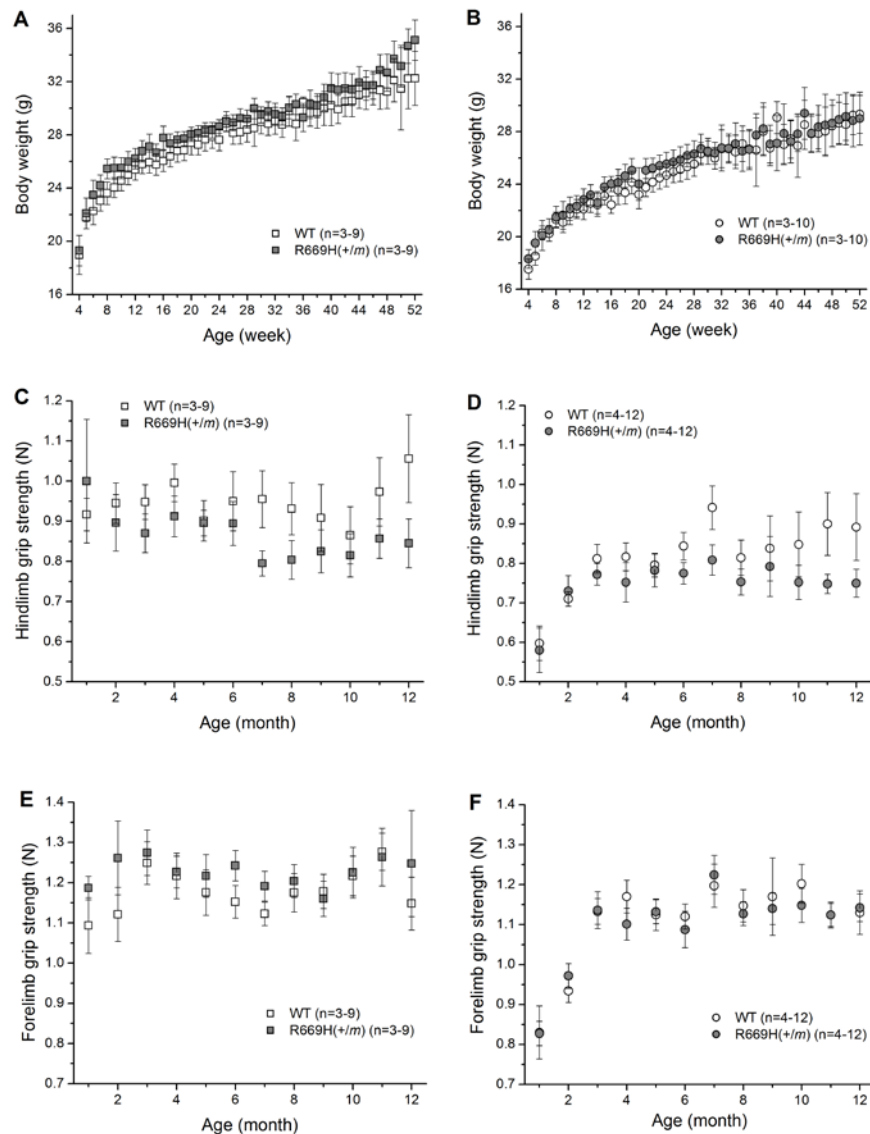


Figure S1. Body mass and grip strength. Initial body mass and weight gain for male (A) and female (B) mice was comparable for WT (+/+) and heterozygous (+/m) R669H mice. Grip strength of the hindlimb was initially comparable for (+/+) and (+/m) mice, but then showed a mild reduction of ~10% for (+/m) animals, beginning at 7 months of age. This difference occurred for male (C) and female (D) mice, but did not achieve statistical significance, $p = 0.1$. (E, F) Grip strength of the forelimbs was indistinguishable between (+/+) and (+/m) mice throughout the observation period. Open symbols for (+/+) and filled for (+/m).