Activation of autophagy attenuates motor deficits and extends lifespan in a *C. elegans* model of ALS

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Supplementary Materials:

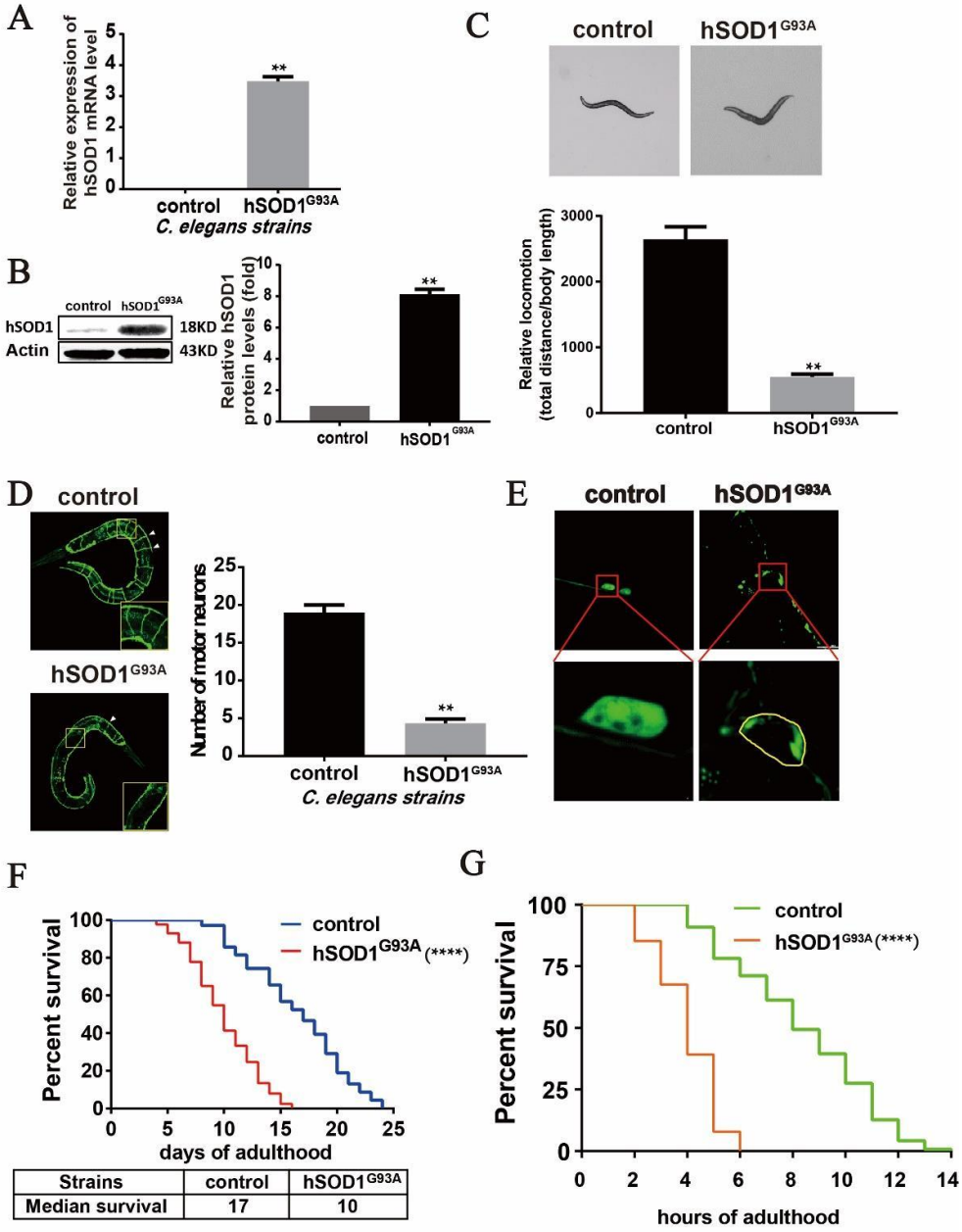
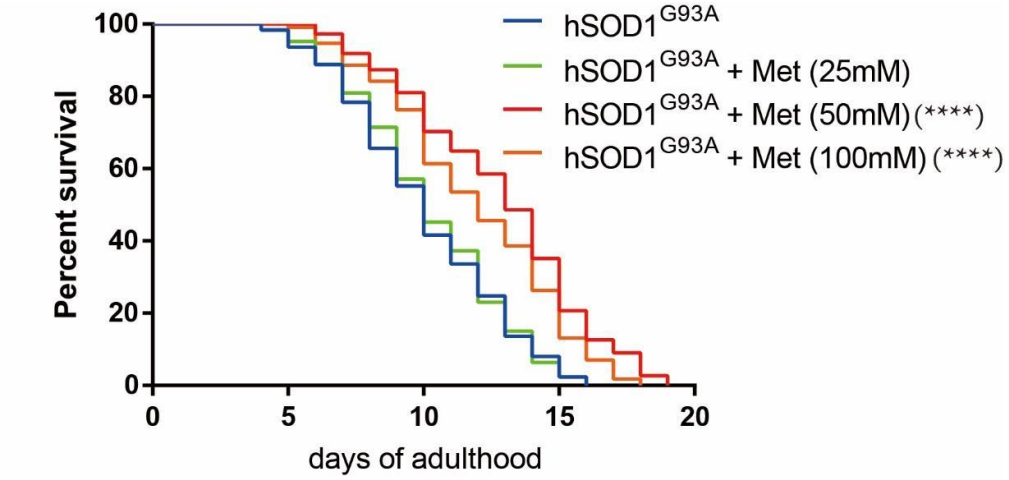


Figure S1. hSOD1 mutation shortens lifespan, causes motor neuron loss, and diminishes the stress response in a *C. elegans* model of ALS. (**A**) Relative expression of hSOD1 mRNA in hSOD1^{G93A} worms. (**B**) Western blot analysis of hSOD1 protein level in the hSOD1 transgenic strains. hSOD1 expression was higher in hSOD1^{G93A} worms than in controls. (**C**) Locomotor behavior in transgenic worms. The movement of hSOD1^{G93A} worms was uncoordinated and relative locomotion was significantly reduced. (**D, E**) Survival and morphology of motor neurons in hSOD1 mutant worms. Motor neurons were almost completely broken down or absent in hSOD1^{G93A} worms, indicating that hSOD1 mutation caused motor neuron degeneration. (**F**) Survival analysis. The median survival of hSOD1^{G93A} worms was 10 days compared to 17 days

for control worms. (**G**) Oxidative stress response in transgenic worms. hSOD1 mutation shortened the lifespan of worms exposed to paraquat. **P<0.001, ****P<0.0001 (N≈100 worms per condition from 3 independent experiments).



Concentration	control	25mM	50mM	100mM
Median survival	10	10	13	12

Figure S2. Determination of optimal metformin concentration. Survival curves for hSOD1^{G93A} worms treated with metformin at different concentrations of 25, 50, and 100 mM. Metformin at the concentrations of 50 mM significantly prolong the lifespan of hSOD1^{G93A} worms. ****P<0.0001, (N≈100 worms per condition from 3 independent experiments).