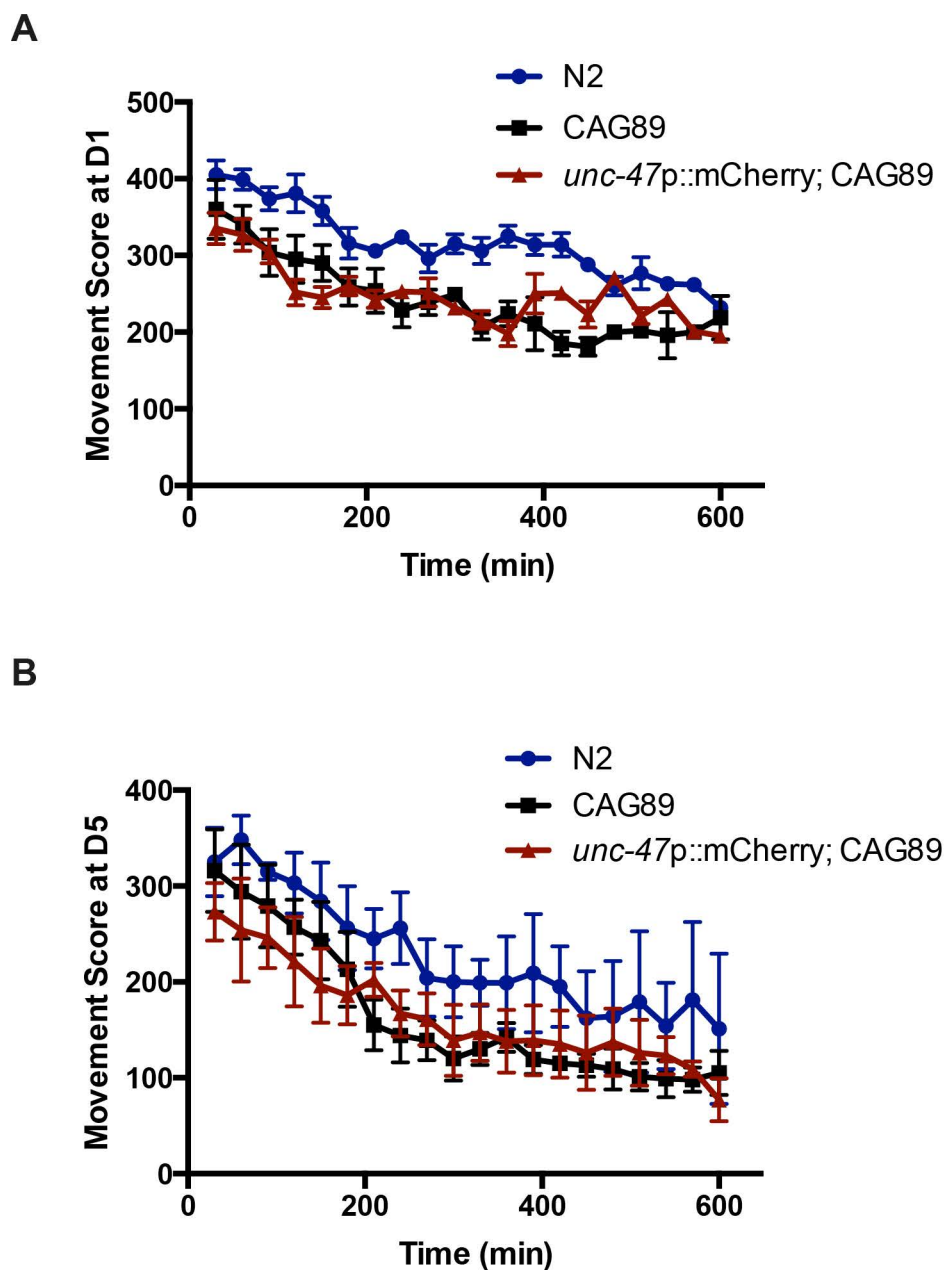


Supplementary Figure 1. *atx-3(tm1689)* do not affect the paralysis and neurodegeneration phenotypes observed in ATXN3-CAG89 transgenic worms

(A) No difference between *unc-47p::mCherry; CAG89* and *(unc-47p::mCherry; CAG89); atx-3(tm1689)* transgenic worms were identified for the paralysis phenotype. This experiment was replicated for 3 times.

(B) Quantification of neurodegeneration in transgenic worms at days one, five and nine of adulthood. At adult day one worms, there was none significant neurodegeneration phenotype for the transgenic worms when compared to *unc-47p::mCherry*. ATXN3-CAG89 transgenics showed a significant increase of neurodegeneration compared to *unc-47p::mCherry* and ATXN3-CAG10 controls (\*\*\*\* $P < 0.0001$  for day five of adulthood and \*\*\* $P < 0.001$  for adult day nine worms, by Student's t-test) (N=100 for each condition). No difference between *unc-47p::mCherry; CAG89* and *(unc-47p::mCherry; CAG89); atx-3(tm1689)* transgenic worms were identified for the neurodegeneration phenotype for none of the three days of the study (days one, five and nine of adulthood). These experiments were replicated for 3 times.



Supplementary Figure 2. *unc-47p::mCherry* do not affect the swimming activity of ATXN3-CAG89 mutant worms

The swimming activity of ATXN3-CAG89 mutants was compared to *unc-47p::mCherry*; CAG89 worms for a period of ten hours at (A) day one of adulthood and, (B) day five of adulthood. No differences for the motility was observed between ATXN3-CAG89 and *unc-47p::mCherry*; CAG89 worms.