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Keywords

Amyotrophic lateral sclerosis – whole genome unbaised screen - *Drosophila* model – FUS

Science

Amyotrophic lateral sclerosis (ALS) is a devastating neurodegenerative disease in which the motor neurons of the motor cortex, brainstem and spinal cord degenerate. The observation that mutations in FUS cause ALS emphasizes the involvement of FUS in the disease pathogenesis. In order to gain better insights into the molecular mechanisms of FUS in neurodegenerative disorders, we generated four transgenic fly lines, allowing expression of wild-type and three disease-associated mutant human FUS proteins.

Selective expression of hFUS transgenes in the motor neurons resulted in a partial pupal lethality with adult escapers showing a wing inflation phenotype. Furthermore the adult escapers show motor performace defects and a reduced lifespan. Restriction of hFUS expression to CCAP neurons, which regulate the wing inflation after eclosion, shows the accelerated cell death of CCAP neurons as the underlying cause. Because the pupal phenotype observed when hFUS transgenes were expressed in motor neurons correlated well with the adult phenotypes and because it is a easy screening phenotype, we decided to perform a whole genome unbaised screen. With this screen we will try to identify new disease-modifying genes to unravel the pathobiology of FUS toxicity.