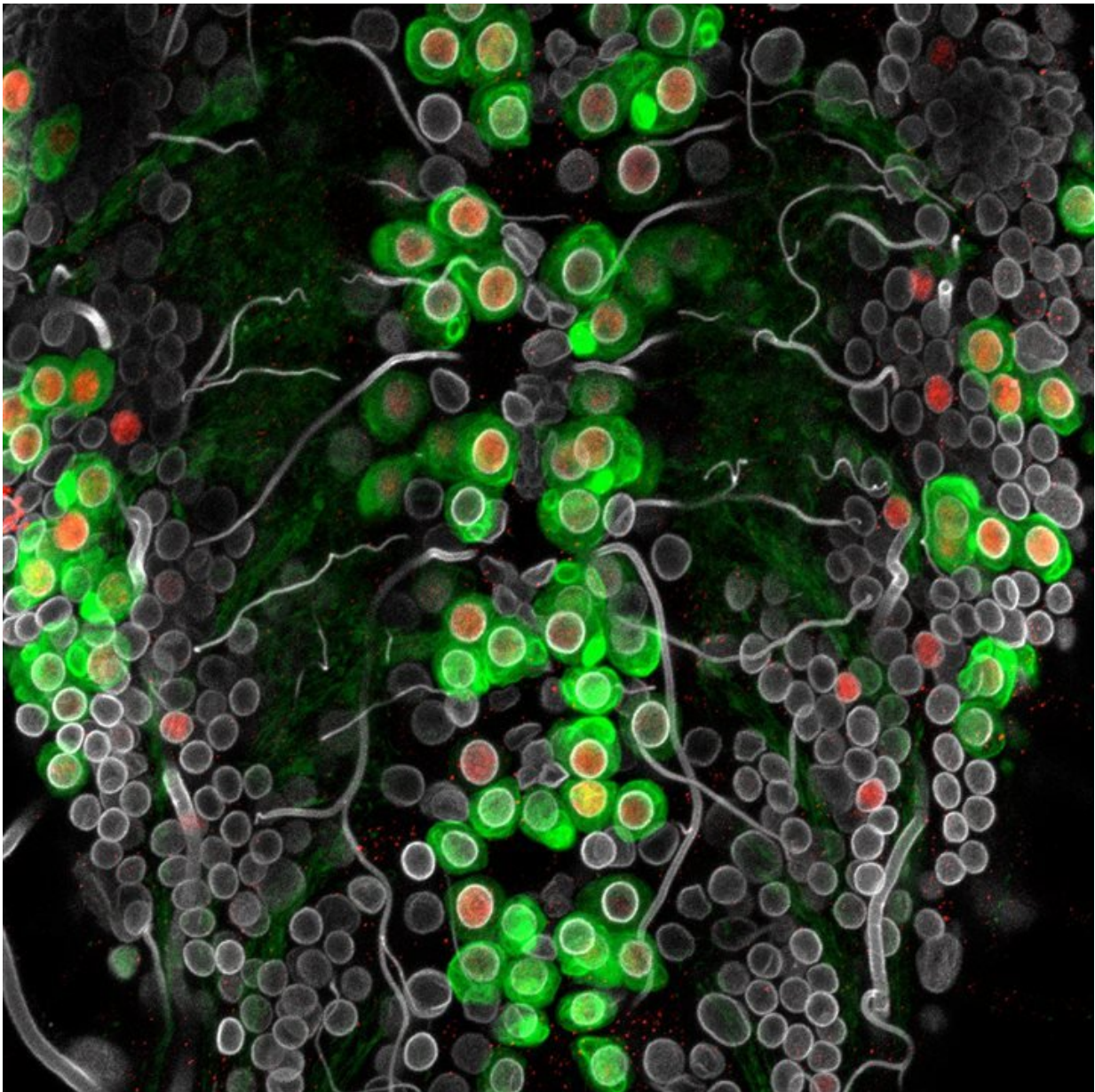


Defects in gene expression partially responsible for ALS and dementia

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Neurons in the 'spinal chord' of the fruit fly. The Xrp1 protein in red. Motor neurons in green. Credit: Radboud University

The Xrp1 protein appears to play an important role in the development of certain forms of ALS and dementia caused by a mutation in the FUS gene. This finding of Radboud University researchers was published on September 12th in the *Journal of Cell Biology*.

The FUS protein is known to be involved in the development of two incurable diseases: ALS ([amyotrophic lateral sclerosis](#)) and FTD ([frontotemporal dementia](#)). Mutations in the FUS gene that promotes the production of FUS protein cause a hereditary form of ALS, while protein accumulations containing the FUS protein are found in the brain of about 10 percent of FTD patients.

By studying a genetic model of the fruit fly (*Drosophila melanogaster*) for these diseases, molecular neurobiologists of Radboud University have shown that FUS proteins causing ALS substantially increase the amounts of another [protein](#), Xrp1. In turn, this causes defects in the expression of more than 3,000 other [genes](#), which probably contributes to the symptoms of the two disorders.

"This finding is an important step in the right direction, since it is now clear that defects in gene expression play an important role in the development of FUS-associated ALS and FTD," says Erik Storkebaum, the leader of the research group. "Future treatments of these incurable diseases should therefore focus on repairing or reducing these defects."

More information: Moushami Mallik et al. Xrp1 genetically interacts with the ALS-associated FUS orthologue caz and mediates its toxicity, *The Journal of Cell Biology* (2018). [DOI: 10.1083/jcb.201802151](https://doi.org/10.1083/jcb.201802151)

Provided by Radboud University

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