

Mutation identified that contributes to Lou Gehrig's disease and possibly Alzheimer's

A team led by scientists at St. Jude Children's Research Hospital and Mayo Clinic has identified a basic biological mechanism that kills neurons in amyotrophic lateral sclerosis (ALS) and in a related genetic disorder, frontotemporal dementia (FTD), found in some ALS patients. ALS is popularly known as Lou Gehrig's disease.

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Phase separation is a mechanism by which proteins assemble into organized assemblies, called membrane-less organelles, necessary for orderly cell functions. The researchers found that the ALS/FTD mutation produces an abnormal version of a protein called TIA1 that is a building block of such organelles. As a result, in ALS, the proteins within the organelles accumulate and kill neurons that control muscles. In FTD, the accumulation kills neurons in the brain. The researchers noted that abnormal phase separation may also underlie Alzheimer's disease.

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[Researcher J. Paul Taylor, chair of the St. Jude Cell and Molecular Biology Department] said, "These findings are part of an emerging theme that there is a whole spectrum of diseases that includes ALS, and some forms of dementia and myopathy, that are caused by disturbance in the behavior of these structures that perturbs cellular organization."

The GLP aggregated and excerpted this blog/article to reflect the diversity of news, opinion, and analysis. Read full, original post: [Scientists identify basic biological mechanism that kills neurons in ALS](#)