PROJECTS SUPPORTED BY JPND



CureALS

Stress granules and proteostasis in motor neurons: towards a mechanistic understanding of ALS

Amyotrophic lateral sclerosis (ALS) and related motor neuron diseases are among the most devastating age-related disorders, and therapies to treat these diseases are still unavailable. In recent years, evidence has been accumulating that ALS is caused or exacerbated by aggregates of proteins and RNAs and a failure of the machinery that controls the behavior of these proteins and RNAs.

In this project, we will study the molecular causes of motor neuron degeneration, focusing on the role of these RNA/protein aggregates as key mediators of cellular decline. We hypothesize that the formation of aberrant RNA/protein aggregates initiates a vicious cycle, which eventually triggers the degeneration of motor neurons and the onset of the first clinical symptoms. As an experimental system, we will use motor neurons and glial cells derived from immortalized patient cells, which will offer an unprecedented window into the disease. Our approach will offer definitive insight into the complex causes of motor neuron disorders and will provide a solid basis for diagnostics and therapy development.



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