The brain's powerhouses are damaged in Creutzfeldt-Jacob disease

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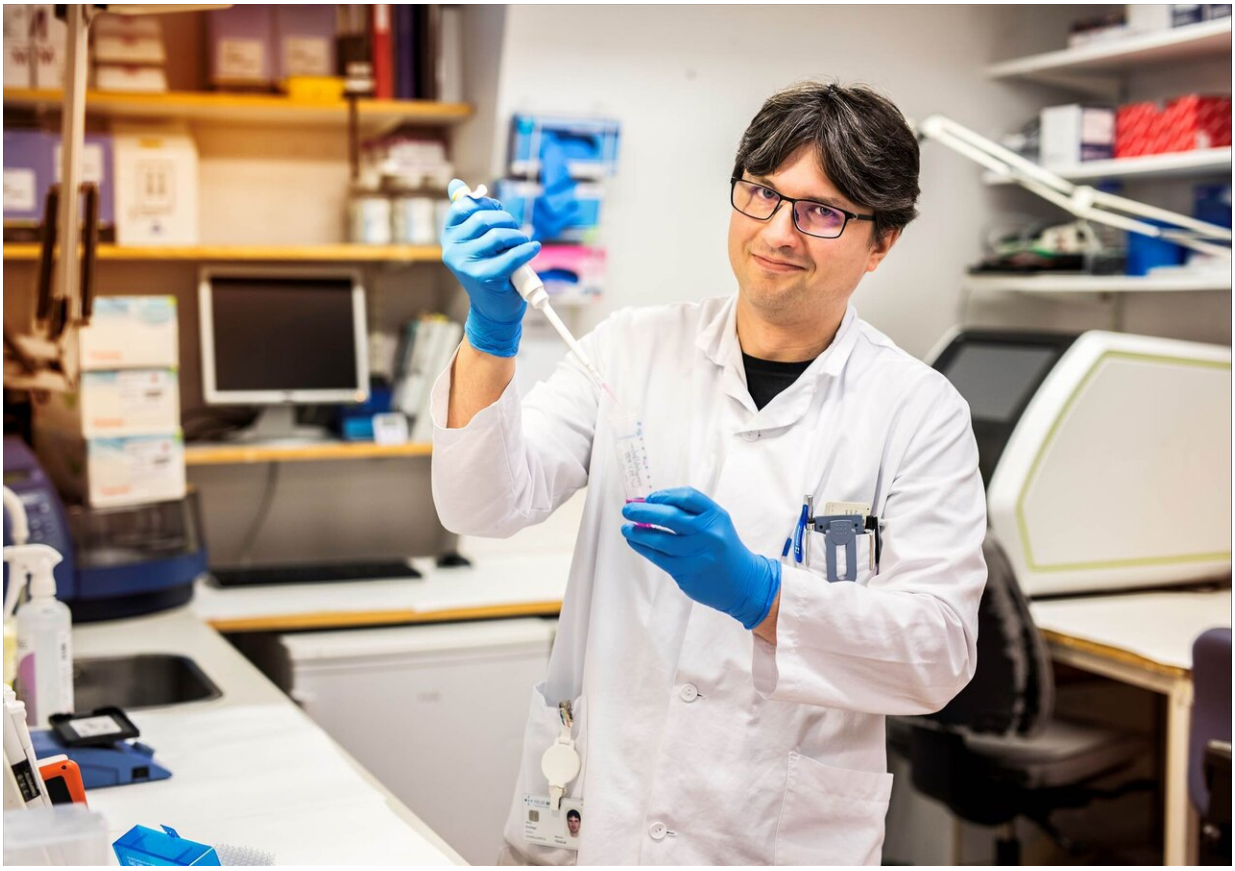

Charlampos "Haris" Tzoulis with groundbreaking research on Creutzfeldt-Jacob disease. Credit: Ingvild Festervoll Melien

A study performed by researchers from the University of Bergen, Norway, and the University of Vienna, Austria, shows damage of the

mitochondria—the cell's microscopic powerhouses—in the brains of people with Creutzfeldt-Jacob disease.

The researchers found that the mitochondrial power generator (known as the respiratory chain) is severely impaired in brain cells from people who died with Creutzfeldt-Jacob disease.

"These mitochondrial defects were widespread in the brain and correlated with the severity of disease," says Professor Charalampos Tzoulis at the University of Bergen and Haukeland University Hospital, Bergen.

Damaged mitochondria are no longer able to provide the energy required for neuronal maintenance and function. "These findings strongly suggest that mitochondrial failure contributes to the pathogenesis of Creutzfeldt-Jacob disease," says Tzoulis.

Hoping for new treatment

Tzoulis highlight the need to better understand how mitochondrial failure contributes to Creutzfeldt-Jacob disease.

"We hope that this study may be exploited towards developing treatments for this incurable and deadly condition," says Charalampos Tzoulis.

Facts: Creutzfeldt-Jacob disease

- Creutzfeldt-Jacob disease is a rare and rapid progressive and severe disease that causes loss of brain tissue.
- Creutzfeldt-Jacob disease is a lethal brain disease that annually affects one to two people per million, and causes death within

four to 24 months.

- The disease normally affects individuals above the age of 60, and causes a combination of invaliding symptoms, often dementia and movement disorders.
- In spite of years of research, there is currently no cure.

More information: Irene H. Fløtnes et al. Mitochondrial respiratory chain deficiency correlates with the severity of neuropathology in sporadic Creutzfeldt-Jakob disease, *Acta Neuropathologica Communications* (2020). DOI: 10.1186/s40478-020-00915-8

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