

# Saving the brain's white matter with mutated mice

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Vanishing White Matter (VWM) disease is a devastating condition that involves the destruction of brain myelin due to a mutation in a central factor. To understand the disease and test potential treatments that could apply to other disorders, such as multiple sclerosis, Prof. Orna Elroy-Stein of Tel Aviv University's Department of Cell Research and Immunology is leading a scientific breakthrough by developing laboratory mice which carry the VWM mutation -- an important new tool.

The mice harbor a mutation of the eIF2B enzyme, which regulates protein synthesis in every cell throughout the body. The key to the new development, says Prof. Elroy-Stein, was the use of genetically-engineered [embryonic stem cells](#) to introduce the mutation.

The [brain](#) is made up of two components: grey matter, or nerves, and [white matter](#), or glial cells which support the nerves and produce myelin, which wraps around and protects nerve extensions. Recently described in the prestigious journal *Brain*, the creation of these mutated mice allows for new research on VWM diseases, which trigger loss of myelin in the brain, leading to paralysis and possible death.

## A new breed of mouse and man

Until now, VWM researchers did not have a comparable animal model with which to study the disease. Now, for the first time, researchers can

use a living organism to follow the exact process of myelin destruction, providing valuable information about the molecular mechanisms of the disease, which are currently unknown.

The big surprise in this discovery, she says, is that the gene mutation impacts only the [white matter](#) of the brain, rather than causing disease throughout the body. This will allow researchers to learn about myelin formation and maintenance.

"For the first time ever, we can follow the regulated expression of the protein components of myelin," explains Prof. Elroy-Stein, whose techniques include [magnetic resonance imaging](#) to investigate what's happening in the brains of the mice.

Not only will this discovery lead to a greater understanding of diseases that affect the brain's white matter, the [mice](#) are also an invaluable testing ground for new treatments. But according to Prof. Elroy-Stein, understanding the disease is the first step. "In order to develop effective therapies, one has to understand the mechanisms," she says.

Provided by Tel Aviv University

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