

Amyotrophic lateral sclerosis may involve a form of sudden, rapid aging of the immune system

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Premature aging of the immune system appears to play a role in the development of amyotrophic lateral sclerosis (ALS), or Lou Gehrig's disease, according to research scientists from the Maxine Dunitz Neurosurgical Institute at Cedars-Sinai Medical Center, the Weizmann Institute of Science in Israel, and Sheba Medical Center in Israel.

A study published in the *Journal of Cellular and Molecular Medicine* shows that CD4+ T cells, which grow and mature in the thymus before entering the bloodstream, are reduced in number in patients who have ALS as the thymus shrinks and malfunctions. Theoretically, devising therapies to support or replace these cells could be a strategy in treating the disease.

The research was led by Michal Schwartz, Ph.D., a visiting professor at the Center of [Neuroimmunology](#) and Neurogenesis in the Department of Neurosurgery at Cedars-Sinai and professor of neuroimmunology at the Weizmann Institute in Rehovot, Israel.

The findings are consistent with evidence collected over a decade by Schwartz's group suggesting that a well-functioning immune system plays a pivotal role in maintaining, protecting and repairing cells of the [central nervous system](#). Studies conducted in animals have shown that boosting immune T-cell levels may reduce symptoms and slow progression of certain [neurodegenerative diseases](#).

Results from the current study suggest that premature aging of the immune system and thus a decrease in protection from immune T cells could contribute to the aggressive and rapid progression of amyotrophic lateral sclerosis, which attacks motor neurons - [nerve cells](#) responsible for muscle strength and voluntary movements. The researchers found that thymic malfunction occurs simultaneously with motor neuron dysfunction, both in laboratory mice bred to mimic amyotrophic lateral sclerosis and in humans suffering from the disease.

Motor neurons extend from the brain to the spinal cord and from the spinal cord to the muscles of the body. Amyotrophic lateral sclerosis damages [motor neurons](#) in the spinal cord, leading to their death, the inability to control muscle action, and the wasting away of muscle tissue. About 5,600 people are diagnosed with amyotrophic lateral sclerosis each year. Up to 10 percent of cases are inherited because of certain gene mutations but the majority occur in the general population with no known cause.

Life expectancy varies greatly but generally ranges from two to five years after diagnosis. More than half of patients survive more than three years, and about 5 percent live 20 years, according to the ALS Association. The disease has been known to spontaneously stop progressing, and in rare cases, the symptoms have actually reversed. Amyotrophic lateral sclerosis is often referred to as Lou Gehrig disease in recognition of the baseball great whose career with the New York Yankees was cut short by the disease in 1939. He died two years later.

The thymus gland, where immune cells called T lymphocytes mature before entering the bloodstream, normally reaches its peak in size and production in childhood. It then slowly shrinks, becoming virtually nonexistent in the elderly, but the lifespan of newly produced T cells ranges from three to 30 years.

This study found that the thymus glands of mice and patients with the disease undergo accelerated degeneration. In addition to using laboratory tests that provide a noninvasive measure of thymic function, the researchers performed imaging scans on three relatively young patients and found no evidence of thymic remnants. Additional studies showed that patients with the disease had dramatically reduced numbers of five genes that are known to support immune responses. Patients also were found to have a significant deficiency of another gene that may make T cells susceptible to a process that causes cell death.

"If T-cell malfunction is confirmed to be a contributing factor to ALS, as we propose, therapeutic strategies may be aimed at overcoming this deficiency through rebuilding, restoring or transplanting the thymus," said Schwartz, the journal article's senior author.

More information: *The Journal of Cellular and Molecular Medicine*, "Thymic Involution in [Amyotrophic Lateral Sclerosis](#)," published online July 24, 2009.

Source: Cedars-Sinai Medical Center

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