NeurologyNow®

AMERICAN ACADEMY OF NEUROLOGY®

AMERICAN BRAIN FOUNDATION®

ALS: The Basics

WHAT IS ALS?

Amyotrophic lateral sclerosis (ALS), sometimes called Lou Gehrig's disease after the New York Yankees baseball player whose career ended in 1939 because of the illness, is a progressive neurodegenerative condition that affects the nerve cells (motor neurons) in the brain and spinal cord that control voluntary movement. ALS most commonly occurs in people between the ages of 40 and 70, but younger and older individuals also can develop the disease.

WHAT ARE THE SYMPTOMS?

Weakness and loss of muscle mass (atrophy), which occur as a result of the degeneration of motor neurons, are the primary symptoms. In later stages, people may be unable to stand or walk, use their hands and arms, chew and swallow food, or speak. They ultimately may be unable to breathe without the support of a machine (mechanical ventilation).

The disease is almost always fatal, usually because the muscles necessary for breathing become too weak. On average, people survive for about three to five years, but up to 10 percent will live for 10 years or more.

WHAT CAUSES IT?

ALS is caused by degeneration of the motor neurons, but scientists have not yet discovered why the disease occurs in some people but not others. Ninety to 95 percent of ALS cases are "sporadic," meaning they occur randomly, with no clear risk factors or causes. Individuals with sporadic ALS have no family history of the disease, nor are their family members at increased risk of developing it. In the hereditary form of ALS (only 5 to 10 percent of cases), researchers have identified mutations in more than a dozen genes.

HOW IS IT DIAGNOSED?

There is no specific diagnostic test for ALS. Instead, clinicians rely on a thorough medical history and neurologic examination, supplemented by electromyography (an electrical test of nerve and muscle function). Before making the diagnosis and to rule out other possible causes, doctors may perform a series of other tests, including blood and urine tests, a lumbar puncture (spinal tap), magnetic resonance imaging (MRI), or, in rare cases, a muscle biopsy.

HOW IS IT TREATED?

There is no cure, but the US Food and Drug Administration has approved riluzole (Rilutek), a drug that has been found to slow the disease's progression and prolong survival by several months. Medications are also available to relieve symptoms such as fatigue, muscle cramps, spasticity, constipation, pain, and excess saliva. Depression and sleep disturbances, which are common in ALS, can be treated effectively with medication and therapy. As the disease progresses, patients may require feeding tubes and ventilator assistance.

WHAT RESEARCH IS BEING DONE?

Current research projects funded by the American Brain Foundation (**americanbrainfoundation.org**), the National Institute of Neurological Disorders and Stroke (**ninds.nih.gov**), the ALS Association, the Muscular Dystrophy Association, and several other advocacy organizations seek to understand the mechanisms that selectively trigger motor neurons to degenerate in ALS and to identify ways to halt the processes leading to cell death. Scientists are also working to develop better biomarkers and tools to diagnose and assess disease progression and the efficacy of treatments.

For more *Neurology Now* articles on ALS, go to **bit.ly/NN-ALS**.

For more resources and support, contact:

- ALS Association: **alsa.org**; 800-782-4747
- ALS Therapy Development Institute: **als.net**; 617-441-7200
- Augie's Quest: **augiesquest.org**; 617-441-7268
- Les Turner ALS Foundation: **lesturnerals.org**; 847-679-3311
- Muscular Dystrophy Association: **mda.org**; 800-572-1717
- Prize4Life: **prize4life.org**; 617-545-4882
- Project ALS: **projectals.org**; 212-420-7382

SOURCES: NATIONAL INSTITUTE OF NEUROLOGICAL DISORDERS AND STROKE; ALS ASSOCIATION

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