What Is Amyotrophic Lateral Sclerosis?

Amyotrophic lateral sclerosis (ALS) is a neurological disorder that results in damage to nerve cells in the brain and spinal cord.

Patients with ALS (also known as “Lou Gehrig disease”) have progressive muscle weakness and muscle atrophy that leads to accelerated physical decline. Death typically occurs within 2 to 5 years after onset of ALS symptoms.

Common Symptoms of ALS

The most common initial symptom of ALS is limb weakness in an arm or a leg, which occurs in 70% of patients. Approximately 25% of patients initially have weakness of muscles used to speak, chew, and swallow, and less than 5% have respiratory muscle weakness. As ALS progresses, common symptoms include increasing weakness in the arms and legs, muscle twitching, limb cramping or stiffness, speech that is slow, slurred, or halting, difficulty chewing or swallowing, weak cough, and shortness of breath. Approximately 5% to 15% of patients with ALS develop dementia, and many experience mild personality changes or decreased ability to control their thoughts, emotions, and behavior.

Who Is Affected by and at Risk of ALS?

ALS affects 2 to 3 people per 100,000 worldwide each year. The disease typically develops in middle-aged and older individuals but can affect younger adults, and is slightly more common in men than in women. Exposure to some chemicals and pesticides may slightly increase the likelihood of developing ALS. Approximately 10% of patients have a family history of ALS; this type of ALS is caused by a specific genetic variant.

How Is the Diagnosis of ALS Made?

Diagnosis of ALS is made by a neurologist based on a patient’s symptoms, physical examination findings, and electromyography and nerve conduction studies, which are tests that evaluate muscle and nerve function. For patients with suspected ALS, brain and spine magnetic resonance imaging (MRI) is usually performed, and some patients may undergo a lumbar puncture, specialized blood testing, or muscle biopsy to rule out other diseases. Individuals with a family history of ALS may be tested for genetic variants.

Treatment of ALS

Although ALS is not curable, several drugs are currently available that may affect ALS disease progression. Riluzole is an oral medication that prolongs survival by an average of 3 months. Edaravone, which may affect ALS disease progression. Riluzole is an oral medication that may modestly slow the progression of ALS, although some recent studies have not shown a clear benefit of this drug. A new medication, which is a combination of taurursodiol and sodium phenylbutyrate, received US Food and Drug Administration approval in September 2022, although the effectiveness of this drug. For more information, visit the National Institute of Neurological Disorders and Stroke website.

FOR MORE INFORMATION

National Institute of Neurological Disorders and Stroke
www.ninds.nih.gov/amyotrophic-lateral-sclerosis-als-fact-sheet

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