

Amyotrophic Lateral Sclerosis

Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig disease, involves progressive loss of **motor neurons** (a type of nerve cell controlling muscle movements) in the brain and spinal cord. ALS is a progressive, disabling, and ultimately fatal disease of unknown cause. Walking, speaking, swallowing, breathing, and other basic functions become impaired with time. About 30000 Americans currently have ALS. The yearly incidence rate is 1 to 2 new cases per 100000 individuals. The disease is commonly discovered during middle age and affects more men than women. The July 11, 2007, issue of *JAMA* includes an article discussing the diagnosis of ALS and recommendations for **palliative** (supportive) care.

SYMPTOMS

It is estimated that more than 50% of motor neurons are lost before symptoms such as muscle weakness become apparent.

- Gradual muscle weakness and wasting in arms and legs
- Muscle **fasciculations** (twitches visible in muscles)
- Difficulty with swallowing, speaking, and breathing
- Muscle stiffness, bodily pains, and cramps, especially at night

Respiratory failure is the usual cause of death in ALS. Other causes include pneumonia. In most cases, death occurs within 3 to 6 years after symptoms begin, although some individuals with ALS live for many years, even decades.

DIAGNOSIS

Diagnosis is based on a careful medical history, physical examination, and laboratory tests. **Electromyograms** (EMGs)—nerve conduction studies that evaluate nerve and muscle function—are the key tests. Other tests may include blood tests and neuroimaging studies such as CT or MRI scans of the brain and spinal cord. Molecular testing, cerebrospinal fluid tests, or muscle biopsy may be necessary.

TREATMENT

- Physical, occupational, and speech therapies can assist in daily functioning.
- Riluzole is the only medication approved for the treatment of ALS. It may prolong survival by a few months.
- Other medications may relieve symptoms such as muscle pain, cramping, drooling, spasms, and fatigue.

SUPPORTIVE CARE

- Appropriate exercises to help maintain mobility, strength, and energy
- Changes in diet to minimize episodes of choking and ensure adequate nutrition
- Effective use of assistive devices and braces such as neck collar, foot brace, cane, walker, or wheelchair
- Ramps, handrails, raised toilet seat, shower seat
- Erasable writing tablets or voice amplifiers and computers to help communication
- A noninvasive **ventilator** (breathing machine) may be important to support breathing

The progressive, disabling nature of ALS and the fact that there is no cure make it a difficult disease to manage. In addition to medical care, patients need emotional support from family, friends, doctors, and caregivers.

Sources: Muscular Dystrophy Association, Amyotrophic Lateral Sclerosis Association

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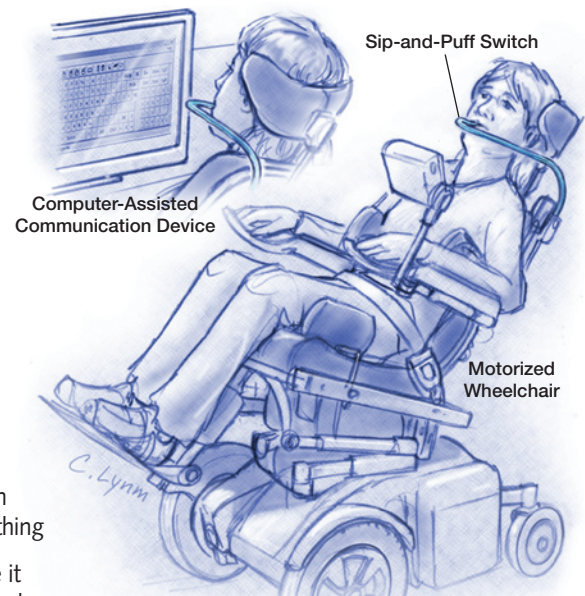
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FOR MORE INFORMATION

- Muscular Dystrophy Association ALS Division (MDA)
www.als.mdausa.org
- Amyotrophic Lateral Sclerosis Association (ALSA)
www.alsa.org

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With the aid of an assistive device, such as a "sip-and-puff" switch, individuals can control motorized wheelchairs, communication devices, and computer programs.

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