

Amyotrophic Lateral Sclerosis

DEFINITION

Amyotrophic lateral sclerosis (a-mi-oh-TROH-fik LAT-ur-ul skluh-ROH-sis), or ALS, is a serious neurological disease that causes muscle weakness, disability and eventually death. ALS is often called Lou Gehrig's disease, after the famous baseball player who was diagnosed with it in 1939. In the U.S., ALS and motor neuron disease (MND) are sometimes used interchangeably.

Worldwide, ALS occurs in 1 to 3 people per 100,000. In the vast majority of cases — 90 to 95 percent — doctors don't yet know why ALS occurs. About 5 to 10 percent of ALS cases are inherited.

ALS often begins with muscle twitching and weakness in an arm or leg, or with slurring of speech. Eventually, ALS affects your ability to control the muscles needed to move, speak, eat and breathe.

(Source: Reprinted from the MayoClinic.com article "Amyotrophic Lateral Sclerosis: Definition"
<http://www.mayoclinic.com/health/amyotrophic-lateral-sclerosis/DS00359>)

SYMPTOMS

Early signs and symptoms of ALS include:

- Difficulty lifting the front part of your foot and toes (footdrop)
- Weakness in your leg, feet or ankles
- Hand weakness or clumsiness
- Slurring of speech or trouble swallowing
- Muscle cramps and twitching in your arms, shoulders and tongue

The disease frequently begins in your hands, feet or limbs, and then spreads to other parts of your body. As the disease advances, your muscles become progressively weaker until they're paralyzed. It eventually affects chewing, swallowing, speaking and breathing.

(Source: Reprinted from the MayoClinic.com article "Amyotrophic Lateral Sclerosis: Symptoms"
<http://www.mayoclinic.com/health/amyotrophic-lateral-sclerosis/DS00359/DSECTION=symptoms>)

CAUSES

In ALS, the nerve cells that control the movement of your muscles gradually die, so your muscles progressively weaken and begin to waste away. Up to 1 in 10 cases of ALS is inherited. But the remainders appear to occur randomly.

Researchers are studying several possible causes of ALS, including:

- **Gene mutation.** Various genetic mutations can lead to inherited forms of ALS, which appear nearly identical to the non-inherited forms.
- **Chemical imbalance.** People who have ALS typically have higher than normal levels of glutamate, a chemical messenger in the brain, around the nerve cells in their spinal fluid. Too much glutamate is known to be toxic to some nerve cells.

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- **Disorganized immune response.** Sometimes a person's immune system begins attacking some of his or her body's own normal cells, and scientists have speculated that this may trigger the process that results in ALS.
- **Protein mishandling.** There's evidence that mishandled proteins within the nerve cells can lead to a gradual accumulation of abnormal forms of these proteins in the cells, eventually causing the nerve cells to die.

(Source: Reprinted from the MayoClinic.com article " Amyotrophic Lateral Sclerosis: Causes"
<http://www.mayoclinic.com/health/amyotrophic-lateral-sclerosis/DS00359/DSECTION=causes>)

RISK FACTORS

Established risk factors for ALS include:

- **Heredity.** Up to 10 percent of the people who have ALS inherited it from their parents. If you have this type of ALS, your children have a 50-50 chance of developing the disease.
- **Age.** ALS most commonly occurs in people between the ages of 40 and 60.
- **Sex.** Before the age of 65, slightly more men than women develop ALS. This sex difference disappears after age 70.

It may be that ALS, similar to other diseases, is triggered by certain environmental factors in people who already carry a genetic predisposition to the disease. For example, some studies examining the entire human genome (genome-wide association studies) found numerous genetic variations that people with ALS held in common, and that might make a person more susceptible to ALS.

Environmental factors under study that may modify a person's individual risk of ALS include:

- **Smoking.** Smoking cigarettes appears to increase a person's risk of ALS to almost twice the risk of nonsmokers. The more years spent smoking, the greater the risk. On the other hand, quitting smoking can eventually lower this increased risk to that of a nonsmoker.
- **Lead exposure.** Some evidence suggests that exposure to lead in the workplace may be associated with the development of ALS.
- **Military service.** Recent studies indicate that people who have served in the military are at higher risk of ALS. Exactly what about military service may trigger the development of ALS is uncertain, but it may include exposure to certain metals or chemicals, traumatic injuries, viral infections and intense exertion.

(Source: Reprinted from the MayoClinic.com article " Amyotrophic Lateral Sclerosis: Risk Factors"
<http://www.mayoclinic.com/health/amyotrophic-lateral-sclerosis/DS00359/DSECTION=risk-factors>)

COMPLICATIONS

As the disease progresses, people with ALS experience one or more of the following complications:

Breathing problems

- ALS eventually paralyzes the muscles needed to breathe. Some devices to assist your breathing are worn only at night and are similar to devices used by people who have sleep apnea (for

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example, continuous positive airway pressure, or CPAP, masks). In the latter stages of ALS, some people choose to have a tracheostomy — a surgically-created hole at the front of the neck leading to the windpipe (trachea) — to enable the full-time use of a respirator that inflates and deflates their lungs.

- The most common cause of death for people with ALS is respiratory failure, usually within three to five years after symptoms begin.

Eating problems

- When the muscles that control swallowing are affected, people with ALS can develop malnutrition and dehydration. They are also at higher risk of aspirating food, liquids or secretions into the lungs, which can cause pneumonia. A feeding tube can reduce these risks.

Dementia

- Some people with ALS experience problems with memory and making decisions, and some are eventually diagnosed with a form of dementia called front temporal dementia.

(Source: Reprinted from the MayoClinic.com article " Amyotrophic Lateral Sclerosis: Complications"
<http://www.mayoclinic.com/health/amyotrophic-lateral-sclerosis/DS00359/DSECTION=complications>)