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Amyotrophic lateral sclerosis

Overview

Amyotrophic lateral sclerosis, or ALS, is a disease of the nerve cells in the brain and spinal cord that control voluntary muscle movement.

ALS is also known as Lou Gehrig's disease.

Symptoms

Symptoms usually do not develop until after age 50. Persons with ALS have a loss of muscle strength and coordination that eventually gets worse. This eventually makes one unable to do routine tasks such as going up steps, getting out of a chair, or swallowing.

Breathing or swallowing muscles may be the first muscles affected. As the disease gets worse, more muscle groups develop problems.

ALS does not affect the senses (sight, smell, taste, hearing, touch), bladder or bowel function, or a person's ability to think or reason.

Symptoms include:

- Difficulty breathing
- Difficulty swallowing
    - Gagging
    - Chokes easily
- Head drop due to weak spinal and neck muscles
- Muscle cramps
- Muscle weakness that slowly gets worse
    - Commonly involves one part of the body first, such as the arm or hand
    - Eventually leads to difficulty lifting, climbing stairs, and walking
- Paralysis
- Speech problems, such as a slow or abnormal speech pattern
- Voice changes, hoarseness

Additional symptoms that may be associated with this disease:

- Drooling
- Muscle contractions
- Muscle spasms
- Ankle, feet, and leg swelling
- Weight loss

Treatment

There is no known cure for ALS. The first drug treatment for the disease is a medicine called riluzole. Riluzole may prolong life, but does not reverse or stop the disease from getting worse.

The goal of treatment is to control symptoms. Baclofen or diazepam may be used to control spasticity that interferes with activities of daily living. Trihexyphenidyl or amitriptyline may be prescribed for people with problems swallowing their own saliva.

Physical therapy, rehabilitation, use of braces or a wheelchair, or other orthopedic measures may be needed to maximize muscle function and general health.

Choking is common. Patients may decide to have a tube placed into their stomach for feeding. This is called a gastrostomy.

A nutritionist is very important to help prevent weight loss. The illness itself appears to increase the need for food and there is usually limited ability to swallow.

The use of devices to assist in breathing includes machines that are only used at night as well as constant mechanical ventilation. Patients should discuss their wishes regarding artificial ventilation with their families and doctors.

Causes

In about 10% of cases, ALS is caused by a genetic defect. In other cases, the cause is unknown.

In ALS, nerve cells (neurons) waste away or die, and can no longer send messages to muscles. This eventually leads to muscle weakening, twitching, and an inability to move the arms, legs, and body. The condition slowly gets worse. When the muscles in the chest area stop working, it becomes hard or impossible to breathe on one's own.

ALS affects approximately 1 out of every 100,000 people.

Except for having a family member who has a hereditary form of the disease, there are no known risk factors.
Tests & diagnosis

An exam of the nerves and muscles shows weakness, often beginning in one area. There may be muscle tremors, spasms, twitching, or loss of muscle tissue (atrophy). Atrophy and twitching of the tongue are common.

The person's walk may be stiff or clumsy. Reflexes may be abnormal and may include loss of the gag reflex. Some patients have trouble controlling crying or laughing. This is sometimes called "emotional incontinence."

Tests that may be done include:

- Blood tests to rule out other conditions
- Breathing test to see if lung muscles are affected
- EMG to see which nerves do not work properly
- Genetic testing, if there is a family history of ALS
- Head CT or MRI of head to rule out other conditions
- Swallowing studies
- Spinal tap (lumbar puncture)

Prognosis

There is progressive loss of ability to function or care for oneself. Death often occurs within 3 to 5 years of diagnosis, about 20% of patients survive more than 5 years after diagnosis.

Prevention

Genetic counseling may be advised if there is a family history of ALS.

Complications

- Inhaling food or fluid
- Loss of ability to care for self
- Lung failure (See: Adult respiratory distress syndrome)
- Pneumonia
- Pressure sores
- Weight loss

When to contact a doctor

Call your health care provider if symptoms suggest ALS, particularly if there is a family history of the disorder.

Call your health care provider if ALS has been diagnosed and symptoms worsen or new symptoms develop. Increased difficulty swallowing, difficulty breathing, and episodes of apnea are symptoms that require immediate attention.