Amyotrophic Lateral Sclerosis

Overview

Amyotrophic lateral sclerosis (ALS) is commonly referred to as “Lou Gehrig’s Disease.” Lou Gehrig was a famous major league baseball player. He was forced to retire from baseball at 36 years of age after a diagnosis of ALS and died two years later. ALS is a progressive neurodegenerative disease affecting nerve cells in the brain and spinal cord. The only nerve cells affected by ALS are motor neurons. These neurons are located from the brain to the spinal cord and from the spinal cord to the muscles throughout the body. As the neurons degenerate the brain loses ability to initiate and control muscle movement. The degeneration of the motor neurons eventually cause cell death. As ALS progresses it leads to muscle weakness, paralysis, and respiratory failure. The cause of ALS is not fully understood. There have been studies regarding the physiology of the disease. Scientists have discovered that neurodegeneration is a complex action involving several mechanisms. There is currently no cure but there are treatments that can reverse or slow the progression of the disease.

Population

There are over 5,600 people in the United States that are diagnosed with ALS each year, and is estimated that 30,000 people currently have ALS in the U.S. ALS is more common in men than women. People usually develop ALS between ages 40 to 70 with the mean age being 55. It is rare for a person to be diagnosed in their twenties and thirties, but it does happen. Currently, the most frequent population of people to develop ALS are military veterans deployed during the Gulf War. There are researchers studying several other causes of ALS including genetics, chemical imbalances, disorganized immune responses, protein mishandling and environmental factors. Approximately 10 percent of people who have ALS also have a parent with the disease.

Symptoms

In the beginning symptoms are mild and frequently go undetected. Early signs are related to muscle weakness and usually include tripping, dropping things, abnormal fatigue of arms or legs, slurred speech, muscle cramps and uncontrollable periods of laughing or crying. As the disease progresses symptoms include:

- Muscle weakness in one of more of the following: hands, arms, legs or the muscles involved in speech, swallowing or breathing. Muscle weakness is the hallmark sign of ALS.
- Twitching (fasciculations) and cramping of muscles.
- Impairment of the use of the arms and legs.
- “Thick speech” and difficulty in projecting voice.
- In advanced stages, shortness of breath, difficulty breathing and swallowing. When breathing muscles are affected, patients will need ventilator support in order to survive.
Diagnosis/Prognosis

This is a difficult disease to diagnose. There is not a single diagnostic tool that can be used to identify ALS. A clinical examination and a series of diagnostic tests have to be performed to confirm a diagnosis. The following procedures are usually performed:

- Electromyography and nerve conduction velocity tests
- Blood and urine studies
- Spinal tap
- X-rays and MRI
- Myelogram of cervical spine
- Muscle and/or nerve biopsy
- Thorough neurological examination

Progression rates vary from person to person. The mean survival time after diagnosis is 3-5 years. About ten percent of people with ALS survive up to 10 years after diagnosis. Not all people with ALS experience the same symptoms or the same sequence of progression. However, progressive muscle weakness and paralysis are the universal symptoms among patients. As the disease progresses the following complications are common:

- Breathing problems – In later stages of ALS, some people require tracheostomy. The most common cause of death for people with ALS is respiratory failure.
- Eating problems – When the muscles involved in swallowing are affected, people can develop malnutrition and dehydration. A feeding tube can be inserted to reduce the risk of aspirating food.

- Dementia – Some people with ALS experience frontotemporal dementia (problems with memory and making decisions).
- Muscle wasting – Muscles become weak and then atrophy. People with ALS are confined to a wheelchair when their muscles become paralyzed in later stages.

Forms

There are three classifications of ALS that have been identified.

- Sporadic – This is the most common form of ALS in the United States. This type of ALS includes both upper motor neuron and lower motor neuron involvement.
- Familial – Linked to family lineage with genetic dominant inheritance. This only accounts for a small percentage of cases in the United States.
- Gaumanian – There was an extremely high incidence of ALS observed in Guam and the Trust Territories of the Pacific in the 1950’s.

Treatment

There is no cure or effective treatment for ALS. The drug riluzole (Rilutek) was the first and is currently the only medication approved by the FDA for slowing ALS. There are several other treatments that are in clinical trials. A doctor may also prescribe medication for some of the symptoms of ALS. Physical therapy recommends a low-impact range of exercise to maintain cardiovascular fitness, muscle strength, and range of motion as long as possible. PTs can help a patient get fitted for a brace, walker or wheelchair. Speech therapy can teach adaptive techniques to make speech more clearly understood and explore alternative methods of communication such as computer based equipment. There are several resources available for people with ALS and their families to find out more about the disease and how to cope with the progression of ALS.

References

- www.alshopefoundation.org
- www.alsa.org
- www.mayoclinic.com
- www.als.net

Other News:

**If you have any suggestions for newsletter topics, please contact Dean Susan Hanrahan at hanrahan@astate.edu.**

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The Arkansas State University Employee Wellness Newsletter is published monthly during the academic year by the College of Nursing and Health Professions. Health questions can be addressed to Dean Susan Hanrahan, Ph.D., ext. 3112 or hanrahan@astate.edu. Produced by Michelle Williams, graduate student in the College of Nursing and Health Professions, Physical Therapy Program.