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ALS Information

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Common Questions about ALS

What is ALS?

ALS is the abbreviation for "Amyotrophic Lateral Sclerosis." "A" means no or negative. "Myo" refers to muscle, and "Trophic" means nutrition or stimulation. When a muscle has no stimulation, it "atrophies" or wastes away. "Lateral" identifies the area of the spinal cord where the pathways for motor nerves, those that innervate the muscles, are located. As this area degenerates it leads to scarring or hardening, "sclerosis," in the region.

Amyotrophic Lateral Sclerosis, also known as Lou Gehrig's disease, and Motor Neuron Disease, is a progressive neuromuscular disease. ALS was first identified by French neurologist Jean-Martin Charcot in 1869. This disease affects the brain's motor neuron pathways causing progressive muscle weakness and can cause loss of function for speech, swallowing, and movement.

What are the symptoms of ALS?

Initial symptoms may include tripping, stumbling and falling, or loss of strength or muscle control in hands and arms, called **limb-onset ALS**. For some it may show up as difficulty speaking, swallowing or breathing, called **bulbar-onset ALS**. Symptoms may include: muscle stiffness (spasticity), twitching (fasciculations), muscle cramps, chronic fatigue or exaggerated reflexes. The voluntary muscles or skeletal muscles are affected by ALS, eventually leading to paralysis.

Involuntary movement, such as eye movement, bladder and bowel control, and sexual function are not usually affected, nor are the senses: touch, vision and hearing.

Pain is not specifically a symptom of ALS but can occur as a result of muscle cramps, loss of muscle strength and loss of mobility. Common pains as a result of ALS include pressure sores, muscle cramps, joint contractures, constipation, burning eyes, swelling feet, and muscle aches.

Some recent studies do show that a small percentage of ALS patients have experienced dementia or Frontotemporal Dementia (FTD). FTD symptoms include cognitive and behavioral changes.

Who gets ALS?

ALS occurs in all races and all around the world. It is not contagious. Approximately 2 in 100,000 people get ALS.

In the United States, approximately 30,000 people are living with ALS. There are over 5,000 new cases per year diagnosed within the United States. In Michigan, an estimated 1,000 people are living with ALS and 200 are newly diagnosed each year. Both men and women get ALS but statistics show that men are 20% more likely to get ALS. ALS can affect people at any age, and cases have been found in persons as young as 12 and as old as 98. Approximately 80% of ALS



cases begin between the ages of 40 and 70 with the average age of onset being age 55. There appears to be a trend of younger patients in their 20's and 30's being diagnosed with ALS.

ALS is classified as either “sporadic” ALS, meaning it occurs randomly, or “familial”, which means other family members have had ALS. Approximately 10 percent of the ALS population has familial ALS. In familial ALS the disease is autosomal dominant, meaning that if a parent has had ALS, their children have a 50% chance of inheriting the defective gene. While the risk of inheriting the defective gene is 50% for each child of an affected person, not all people with the defective gene will develop the disorder.

What causes ALS?

The cause of ALS is unknown, and researchers have been unable to identify why this disease strikes some and not others. In searching for a cause, researchers have investigated several environmental factors such as exposure to toxic or infectious agents. Other research has examined the possible role of an individual’s diet and or traumas they may have sustained at an earlier age. However, they have been unable to link these factors in causing ALS.

In 1993 scientists supported by the National Institute of Neurological Disorders and Stroke (NINDS) discovered that mutations in the gene that produces the SOD1 enzyme (superoxide dismutase) were associated with some cases of familial ALS. This enzyme is an antioxidant that protects the body from damage caused by free radicals. If not neutralized, free radicals can accumulate and cause damage to the DNA and proteins within cells. It is not yet clear how the SOD1 gene mutation leads to motor neuron degeneration, researchers have theorized that an accumulation of free radicals may result from the faulty functioning of this gene.

How is ALS diagnosed?

An experienced physician, usually a neurologist will complete a complete medical history and physical exam. Some of the diagnostics procedures might also include the following tests.

- ❖ Laboratory tests
- ❖ Muscle and/or nerve biopsy
- ❖ Cerebral spinal fluid analysis (spinal tap). This procedure is used to make an evaluation or diagnosis by examining the fluid withdrawn from the spinal column.
- ❖ Magnetic resonance imaging (MRI) - a way to image soft tissues that's noninvasive and that doesn't involve X-rays. MRI produces a sharp, two-dimensional view of the brain and spinal cord.



- ❖ Electromyography also known as EMG. This procedure is used to evaluate and diagnose disorders of the muscles and motor neurons. Electrodes are inserted into the muscle, or placed on the skin overlying a muscle. Electrical activity and muscle response is then evaluated.

The diagnosis is made by ruling out other diseases and by meeting specific criteria for ALS. There is no one test for ALS.

What is the treatment for ALS?

There is no cure for ALS but the physician or Neurologist; preferably one experienced with ALS will work with the patient and family to manage ALS symptoms. The US Food and Drug Administration (FDA) approved the drug Rilutek®, the first drug that has shown to prolong the survival of persons with ALS. There are medications to relieve muscle cramping, excessive saliva, depression, and anxiety. Physical Therapy and Range of Motion exercises can be done (see treatment section of this manual for these exercises) to ease any stiffness or cramps. Dieticians are able to show ways to promote good eating habits and what to do if swallowing becomes an issue. Meeting with a speech pathologist about techniques that will allow them to continue to speak or use devices to communicate are available. Durable medical equipment (DME) such as bath equipment, eating utensils, walkers, and wheelchairs are forms of DME and can be used when mobility becomes an issue.

What is the future of a person with ALS?

ALS progresses at different rates in each individual. Statistics show the average survival for someone affected by ALS is three to five years. A small percentage of people have a very slow progression and live 10-20 years. Again, each patient is different and it is hard to predict how the disease will impact each person. Each patient chooses different treatment plans and this could impact their future. Improved treatment is allowing ALS patients to live longer than ever before. Patients who stay involved in their treatment and treat their symptoms seem to do better than those who choose to not follow up with regular medical appointments. Patients with a positive attitude and good emotional health tend to do better than patients who are severely depressed. Being educated about the latest treatments and being open to what is available to ALS patients will benefit both the patient and family.

Who Was Lou Gehrig?



Lou Gehrig was a famous baseball player who played for the New York Yankees in the late 1920s through 1939. Lou Gehrig died from ALS in 1941. He wore the number 4 and was nicknamed the "Iron Horse" due to the record setting number of consecutive games he played during his career.

Lou Gehrig was the Yankees first baseman during the late 20's through 1939. He had over 200 hits in eight seasons and had a career average of .340. His most noted accomplishment was his playing streak, which stretched to 2,130 consecutive games, a Major League record which stood for 56 years. Lou Gehrig set an American League record with 184 runs batted in 1931. He holds the record for most career grand slams (23) and won a Triple Crown in 1934. He led the American League in home runs three times, led in RBI five times, and won the League MVP in 1936 when he hit 49 home runs, scored 167 runs, and batted .354.

On July 4th, 1939 the Yankee organization held "Lou Gehrig Appreciation Day." On this day, Lou Gehrig gave the famous speech quoted below. He was also presented with various awards and his number 4 was retired, the first player ever to have his number retired. The following December, he was unanimously elected into the Hall of Fame. The Baseball Writers Association waived the existing rule which required a player to be retired one year before he could be elected. On June 2nd, 1941, Lou Gehrig died in New York from ALS.



Lou Gehrig's July 4, 1939, Lou Gehrig Appreciation Day Speech at Yankee Stadium.

"Fans, for the past two weeks you have been reading about the bad break I got. Yet today I consider myself the luckiest man on the face of the earth. I have been in ballparks for 17 years and have never received anything but kindness and encouragement from you fans. Look at these grand men. Which of you wouldn't consider it the highlight of his career just to associate with them for even one day? Sure I'm lucky. Who wouldn't consider it an honor to have known Jacob Ruppert? Also, the builder of baseball's greatest empire, Ed Barrow? To have spent six years with that wonderful little fellow, Miller Huggins? Then to have spent the next nine years with the best manager in baseball today, Joe McCarthy?"

"Sure I'm lucky. When the New York Giants, a team you would give your right arm to beat, and vice versa, sends you a gift - that's something. When everybody down to the groundskeepers and those boys in white coats remember you with trophies - that's something. When you have a wonderful mother-in-law who takes sides with you in squabbles with her own daughter - that's something. When you have a father and a mother who work all their lives so you can have an education and build your body - it's a blessing."

"When you have a wife who has been a tower of strength and shown more courage than you dreamed existed - that's the finest I know."

"So, I close in saying that I may have had a tough break, but I have an awful lot to live for."

--Lou Gehrig



Recommended Steps after Diagnosis

- If you suspect you have ALS but are not diagnosed yet, look into the possibility of getting additional term life insurance and/or long-term disability insurance.
- ALS is difficult to diagnose. If you were not diagnosed by a neurologist who specializes in neuromuscular diseases, request a referral to one who specializes in ALS for a confirmation of the diagnosis.
- Register with ALS of Michigan, Inc. (800) 882-5764. Request information about available services and publications.
- Contact an attorney who specializes in elder care law and make an appointment to discuss legal and financial planning recommendations. Complete a Durable Power of Attorney or living will that lists your medical wishes.
- Consider attending a support group or if you are a family member attend a caregiver conference. Contact ALS of Michigan for a listing of local ALS support group meetings and upcoming conferences.
- If you served in the military, contact the Department of Veteran Affairs (800-827-1000) about eligibility for health, vocational rehabilitation and disability programs and services.
- If you are employed, contact Michigan Rehabilitation Services at (800) 605-6722 or www.michigan.gov/mdcd and request information about available services.
- When the time comes, be open to the medical equipment and treatment plans that are offered. These options will protect you and may give you a better quality of life.
- Don't be afraid to ask for help. There are many services as well as individuals who do want to help you, allow them to.
- Talk with your physician or social worker about social security disability and how to apply.