My Diagnosis: Amyotrophic Lateral Sclerosis (ALS)/ Lou Gehrig's Disease

Diagnosis Overview

Amyotrophic lateral sclerosis (ALS) is a disease of the nerve cells in the brain, brain stem and spinal cord that control voluntary muscle movement. ALS is also known as Lou Gehrig's disease after the baseball player who was diagnosed with it.

Causes

One in 10 cases of ALS is due to a genetic defect. The cause is unknown in most other cases. In ALS, motor nerve cells (neurons) waste away or die and can no longer send messages to muscles. This eventually leads to muscle weakness, twitching, and an inability to move the arms, legs and body. The condition slowly gets worse and can affect other muscles in the face and chest, making it hard to speak, swallow and breathe.

ALS affects approximately five out of every 100,000 people worldwide.

Having a family member who has a hereditary form of the disease is a risk factor for ALS. Other risks include military service. The reasons for this are unclear, but it may have to do with environmental exposure to toxins.

Symptoms

Symptoms usually do not develop until after age 50, but they can start in younger people. People with ALS have a loss of muscle strength and coordination that eventually gets worse and makes it impossible for them to do routine tasks such as going up steps, getting out of a chair or swallowing.

Weakness can first affect the arms or legs, or the ability to breathe or swallow. As the disease gets worse, more muscle groups develop problems.

ALS does not affect the senses (sight, smell, taste, hearing, touch). Most people are able to think normally, although a small number develop dementia, causing problems with memory.

Muscle weakness starts in one body part, such as the arm or hand, and slowly gets worse until it leads to the following:

- Difficulty lifting, climbing stairs, and walking
- Difficulty breathing
- Difficulty swallowing — choking easily, drooling, or gagging
- Head drop due to weakness of the neck muscles
- Speech problems, such as a slow or abnormal speech pattern (slurring of words)
- Voice changes, hoarseness

Other findings include:

- Depression
- Muscle cramps
- Muscle stiffness, called spasticity
- Small muscle twitches, called fasciculations
- Weight loss

What to Expect

Tests

Your neurologist will examine you and ask about your medical history and symptoms. The physical exam may show:

- Weakness, often beginning in one area
- Muscle tremors, spasms, twitching, or loss of muscle tissue
- Twitching of the tongue
- Abnormal reflexes
- Stiff or clumsy walk
- Difficulty controlling crying or laughing (sometimes called pseudobulbar affect)
- Loss of gag reflex

Tests that may be done include:

- Blood tests to rule out other conditions
- Breathing test to see if lung muscles are affected
- Computerized tomography (CT) scan or magnetic resonance imaging (MRI) scan to be sure there is no disease or injury to the brain or spine, which can mimic ALS
- Electromyography to assess the function of the motor nerves and to better characterize the nature of the problem
- Genetic testing
- Swallowing studies
- Spinal tap (lumbar puncture)

Atlantic Health System

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Treatment

There is currently no cure for ALS, but there are medications to help slow the progression. There are three FDA-approved medications for the treatment of ALS:

- Riluzole (Rilutek®): A pill or liquid taken twice per day. The main side effects include nausea and elevated liver blood tests.
- Edaravone (Radicava®): A liquid or IV taken once daily for 14 days followed by a 14-day drug-free period for the initial cycle. Monthly thereafter, it is taken once daily for a 10-day period, followed by a 14-day drug-free period. The main side effects related to the IV formulation are headache and bruising.
- Sodium phenylbutyrate and taurursodiol (Relyvrio®): One packet dissolved in liquid taken once daily for three weeks, then increased to one packet twice a day ongoing. The main side effects include abdominal pain, diarrhea and nausea.

Treatments to control other symptoms include:

- Baclofen or diazepam for spasticity that interferes with daily activities
- Dextromethorphan HBr and Qunidine Sulfate (Nuedexta®) can be used for inappropriate laughing or crying (pseudobulbar affect)
- Physical therapy, rehabilitation, use of braces or a wheelchair, or other measures may be needed to help with mobility, fall prevention and general health.

People with ALS tend to lose weight. The illness itself increases the need for food and calories. At the same time, problems with choking and swallowing make it hard to eat enough. To help with feeding, a tube may be placed into the stomach. A dietitian who specializes in ALS can give advice on healthy eating.

Breathing devices include machines that are used only at night, and constant mechanical ventilation.

Medicine for depression may be needed if a person with ALS is feeling sad. They also should discuss their wishes regarding artificial ventilation with their families and providers.

Care Team

ALS is a diagnosis which affects not only the patient, but the whole family. At Atlantic Health System, we are with you every step of the way. We employ a multidisciplinary care team to support you along the journey. These include: neurologists, nurses, physical, occupational and speech and swallow therapists, pulmonologists, gastroenterologists and social work.

Frequently Asked Questions

Is Atlantic Health System involved in any clinical trials?

We are not currently running clinical trials for neuromuscular disease. There are several clinical trials in the NY and NJ area that are active and recruiting participants. Please speak with your neurologist about your eligibility. You can find more information at clinicaltrials.gov.

Are there any dietary restrictions?

ALS can cause weight loss and loss of muscle mass. It is important to maintain your weight with calorie-dense, high-protein foods. Consultation with a nutritionist is often recommended. As the disease progresses, it may become more difficult to swallow. Your doctor will talk to you about the benefits of supplemental nutrition, including a feeding tube.

Where can I find additional information about ALS, including support groups?

ALS Association, visit alsa.org.

There is a local chapter which works to serve the families in the Greater NY/NJ area.

Find additional information at als-ny.org.

Where can I find more information about help at home, financial assistance, transportation and other social work needs?

Atlantic Neuroscience Institute offers a resource guide which can be provided at your request or downloaded online.

SCAN ME

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MyChart

MyChart is your personal, patient-centered, fully secure connection to Atlantic Health System. With MyChart, patients who receive care at any of our locations — including Atlantic Medical Group physician practices, Chilton, Hackettstown, Morristown, Newton and Overlook medical centers — can take an active role in managing their own health.

- eCheck-In
- Access health records, lab results and proof of vaccinations
- Request and manage appointments
- Request prescriptions refills
- Bill pay

Scan to learn more at MyChart.atlantichealth.org

Notes

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