The family library

About Amyotrophic Lateral Sclerosis (Lou Gehrig's disease)

What is Amyotrophic Lateral Sclerosis (ALS)?

Amyotrophic lateral sclerosis (a-mi-oh-TROH-fik LAT-ur-ul skluh-ROH-sis), (ALS), sometimes called Lou Gehrig's disease after the famous baseball player who was stricken with the disease in the early 1940s, is a rapidly progressive neurologic disorder that attacks the nerve cells (neurons) responsible for controlling voluntary muscles.

A-myo-trophic comes from the Greek language. "A" means no or negative. "Myo" refers to muscle, and "trophic" means nourishment—"no muscle nourishment." When a muscle has no nourishment, it "atrophies" or wastes away. "Lateral" identifies the areas in a person's spinal cord where portions of the nerve cells that signal and control the muscles are located. As this area degenerates, it leads to scarring or hardening (sclerosis [sklə rŏssiss]) in the region. The disease belongs to a group of disorders known as motor neuron diseases, which are characterized by the gradual degeneration and death of motor neurons.

What are the symptoms of ALS?

The onset of ALS may be so subtle that the symptoms are frequently overlooked. The earliest symptoms may include twitching, cramping, or stiffness of muscles; muscle weakness affecting an arm or a leg; slurred and nasal speech; or difficulty chewing or swallowing. These general complaints then develop into more obvious weakness or atrophy that may cause a physician to suspect ALS.

The parts of the body affected by early symptoms of ALS depend on which muscles in the body are damaged first. In some cases, symptoms initially affect one of the legs, and patients experience awkwardness when walking or running or they notice that they are tripping or stumbling more often.

About ALS (cont.)

Symptoms (cont.)

Some patients first see the effects of the disease on a hand or arm as they experience difficulty with simple tasks requiring manual dexterity such as buttoning a shirt, writing, or turning a key in a lock. Other patients notice speech problems. Regardless of the part of the body first affected by the disease, muscle weakness and atrophy spread to other parts of the body as the disease progresses.

Although the sequence of emerging symptoms and the rate of disease progression vary from person to person, eventually patients will not be able to stand or walk, get in or out of bed on their own, or use their hands and arms. Difficulty swallowing and chewing impair the patient's ability to eat normally and increase the risk of choking. Maintaining weight will then become a problem. Because the disease usually does not affect cognitive abilities, patients are aware of their progressive loss of function and may become anxious or depressed. A small percentage of patients may experience problems with memory or decision-making, and there is growing evidence that some may even develop a form of dementia.

Diagnosing ALS

No one test can provide a definitive diagnosis of ALS, although the presence of upper and lower motor neuron signs in a single limb is strongly suggestive. Instead, the diagnosis of ALS is primarily based on the symptoms and signs the physician observes in the patient and a series of tests to rule out other diseases. Physicians obtain the patient's full medical history and usually conduct a neurologic examination at regular intervals to assess whether symptoms are worsening.

Other tests conducted may include:

- Electromyography (EMG) to detect electrical activity in muscles;
- Nerve conduction velocity (NCV) to looks for specific abnormalities that may rule out a diagnosis of ALS;
- Magnetic resonance imaging (MRI) is often normal in ALS; however, it can be used to rule out other conditions, such as a spinal cord tumor or herniated disk in the neck.

About ALS (cont.)

Because of the prognosis carried by this diagnosis and the variety of diseases or disorders that can resemble ALS in the early stages of the disease, patients may wish to obtain a second neurological opinion.

Treating ALS

No cure has yet been found for ALS. However, the Food and Drug Administration (FDA) has approved the first drug treatment for the disease—riluzole (Rilutek). Riluzole is believed to reduce damage to motor neurons by decreasing the release of glutamate. Clinical trials with ALS patients showed that riluzole prolongs survival by several months, mainly in those with difficulty swallowing. The drug also extends the time before a patient needs ventilator support.

There are other supportive treatments available that are designed to relieve symptoms and improve the quality of life for patients with ALS. Physicians can prescribe medications to help reduce fatigue, ease muscle cramps, control spasticity, and reduce excess saliva and phlegm. Drugs also are available to help patients with pain, depression, sleep disturbances, and constipation. Pharmacists can give advice on the proper use of medications and monitor a patient's prescriptions to avoid risks of drug interactions. Physical therapy and special equipment can enhance patients' independence and safety throughout the course of ALS.

Patients and caregivers can learn from speech therapists and nutritionists how to plan and prepare numerous small meals throughout the day that provide enough calories, fiber and fluid, and how to avoid foods that are difficult to swallow. Patients may begin using suction devices to remove excess fluids or saliva, and prevent choking. When patients can no longer get enough nourishment from eating, doctors may advise inserting a feeding tube into the stomach. The use of a feeding tube also reduces the risk of choking and pneumonia that can result from inhaling liquids into the lungs. The tube is not painful and does not prevent patients from eating food orally if they wish.

About ALS (cont.)

Social workers and home care and hospice nurses help patients, families, and caregivers with the medical, emotional, and financial challenges of coping with ALS, particularly during the final stages of the disease. Social workers provide support such as assistance in obtaining financial aid, arranging durable power of attorney, preparing a living will, and finding support groups for patients and caregivers. Respiratory therapists can help caregivers with tasks such as operating and maintaining respirators, and home care nurses are available not only to provide medical care but also to teach caregivers about giving tube feedings and moving patients to avoid painful skin problems and contractures. Home hospice nurses work in consultation with physicians to ensure proper medication, pain control, and other care affecting the quality of life of patients who wish to remain at home. The home hospice team can also counsel patients and caregivers about end-of-life issues.

Hospice Care

Because of the progressive and terminal nature of ALS, most, if not all ALS patients qualify for Hospice care. Two physicians must specify that the patient has approximately 6 months or less to live.

The goal of hospice is to provide comfort and maintain the highest quality of life for as long as life remains. The focus is not on death, but on compassionate, specialized care for the living. Hospice is not intended for people who are still seeking a cure for their illness, but is completely focused on comfort and relief of symptoms. How comfort is defined is up to the patient or, if the patient is incapacitated, the patient's family. This can mean freedom from physical, emotional, spiritual and/or social pain.

Care may be provided in a patient's home or in a designated facility, such as a nursing home, hospital unit or freestanding hospice, with level of care and sometimes location based upon frequent evaluation of the patient's needs. The four primary levels of care provided by hospice are routine home care, continuous care, general inpatient, and respite care.

About ALS (cont.)

Routine home care

Routine home care is the most common level of care provided. In spite of its title, routine home care does not indicate a location of care, but a level (or intensity) of care provided. Routine care may be provided at a nursing home or assisted living facility, although the majority of hospice patients are treated at home. Interdisciplinary team members supply a variety of services during routine home care, including offering necessary supplies, such as durable medical equipment, medications related to the hospice diagnosis and incidentals like diapers, bed pads, gloves, and skin protectants. Twenty-four hour on-call services must be available as needed. Typically this is provided after normal business hours by a registered nurse prepared to address urgent patient concerns.

Continuous care

Continuous care is a service provided in the patient's home. It is for patients who are experiencing severe symptoms and need temporary extra support. Once a patient is on continuous care, the hospice provides services in the home a minimum of eight hours a day. Because the criteria for continuous care is similar to inpatient care, and due to the challenges a hospice can face with staffing extended care in the home, continuous care is intended to be used for short periods of time.

General inpatient care

General inpatient care is an intensive level of care which may be provided in a nursing home, a specially contracted hospice bed or unit in a hospital, or in a free-standing hospice unit. General inpatient criteria pertains to patients who are experiencing severe symptoms which require daily interventions from the hospice team to manage. Often, patients on this level of care have begun the "active phase" of dying—when their prognosis is measured in days as opposed to weeks or months. Although there is a limit to how long Medicare will cover this level of care, it is usually provided for brief periods of time, with five to seven days being the average.

About ALS (cont.)

Respite

Respite care (sometimes referred to as respite inpatient) is a brief and periodic level of care a patient may receive. Respite is a unique benefit in that the care is provided for the needs of the family, not the patient. Should a family member need a "break" from caregiver or even if they have a vacation planned then this level of care may be provided. During respite, the patient is transferred from the home to an institutional setting; this can be a nursing, assisted living, hospital or an inpatient hospice unit. Should a patient be transferred to an assisted living facility, nursing home or hospital the hospice would continue to provide care to the patient which is on par with the services provided under the routine home care benefit. In this way, the only difference between respite and routine care is that the hospice pays the room and board charges of the facility. Should a patient receive respite in an inpatient hospice unit the care would be similar to what other patients of the hospice unit receive. Respite is provided for a maximum of five days every benefit period.

Hospice services are often used by the ALS patient and his/her family due to the progressive and terminal nature of the disease. The hospice philosophy of end-of-life care which focuses on the physical, emotional, spiritual and/or social needs of the dying patient and his/her family provides comfort during a time of enormous strain for the individual and family.

For more information on ALS, click HERE.