Southern Caregiver Resource Center

Caring for those who care for others

Fact Sheet

Amyotrophic Lateral Sclerosis (ALS)

What is ALS?

Amyotrophic Lateral Sclerosis (ALS), also called Lou Gehrig's disease or motor neuron disease, is a progressive, neuromuscular disease that attacks nerve cells and pathways in the brain and spinal cord. Motor neurons, among the largest of all nerve cells, reach from the brain to the spinal cord and from the spinal cord to muscles throughout the body. When these motor neurons die, the brain can no longer start and control muscle movement. At this time, there is no cure for the disease; however, over the past decade, we have made amazing strides in our understanding of the brain, the nervous system, and genetics. Discoveries in each of these areas bring hope to people with ALS (PALS) and their families that someday a cure will be found.

Amyotrophic comes from the Greek language. "A" means no or negative. "Myo" refers to muscle, and "trophic" means nourishment, thus "no muscle nourishment." When a muscle receives no nourishment, it atrophies or wastes away. "Lateral" identifies the areas in a person's spinal cord where portions of the nerve cells that nourish the muscles are located. As this area degenerates, it leads to scarring or hardening ("sclerosis") in the region. As more and more nerves and muscles are affected, the person with ALS loses the ability to move and eventually suffering complete paralysis. The muscles used for breathing, speaking, and swallowing also become affected. ALS does not affect the person's senses (feeling, tasting, smelling, etc.), bladder and bowel function, or sexual drive and function.

Who Gets ALS?

There are over 20,000 people living with ALS in the United States, and approximately 6,000 Americans are newly diagnosed with ALS each year. Symptoms usually appear in individuals between the ages of 40-70, though, the disease occurs in both younger and older persons. Survival after the confirming diagnosis is, on average, three to five years. The progression of ALS varies with each individual. Approximately, ten percent of those diagnosed with ALS live for ten years or more. Family caregivers, physicians, nurses, physical therapists, speech therapists/pathologists, occupational therapists, and social workers all working together with the person with ALS can help ensure a high quality of life and as much independence as possible.

ALS occurs more frequently in men than in women. For 90-95% of all ALS cases, there is no clearly known cause. These cases, which appear to occur at random, are referred to as *sporadic ALS*. Between 5-10% of ALS cases are inherited. This genetic form of ALS is known as *familial ALS*. Sporadic and familial ALS both have similar symptoms and progression. ALS is not contagious.

891 Kuhn Drive, Ste. 200 • Chula Vista, CA 91914
(858) 268-4432 • (800) 827-1008 • (858) 268-7816 Fax • E-mail: scrc@caregivercenter.org
Web Site: www.caregivercenter.org

Symptoms

Some patients become aware of the onset of the disease when their hands become clumsy, causing difficulty in the performance of tasks like unlocking doors or writing. Others experience weakness in the legs and may trip or stumble. Other people notice they have problems speaking or difficulty swallowing. ALS may be present for some time before any symptoms are noticed. This lack of noticeable symptoms occurs because the remaining functioning nerve cells compensate for the lost or damaged nerve cells. One early symptom is generalized fatigue. As muscle cells deteriorate, patients may experience stiffness, occasional jerking of the arms or legs, or twitching (fasciculations). Often symptoms begin in the hands and feet, then travel inward toward the center of the body. One side is usually more affected than the other. Eventually paralysis may be complete except for the muscles of the eyes.

Some of those affected by ALS may exhibit uncontrollable laughter or crying. This is referred to as *pseudobulbar affect*. Depression can also be a symptom. Some PALS experience *sialorrhea*, drooling. Some PALS experience problems with thinking (which is referred to as *cognitive impairment*), and/or changes in behavior. Less commonly, some of the PALS may develop frontotemporal dementia (FTD), in which there is progressive worsening of behavior, personality and/or language that impairs daily function.

Diagnosis

There is no specific clinical test that can identify ALS. Diagnosis is made by a neurologist through a physical examination, a thorough patient medical history, and neurological testing. Diagnostic testing often includes the nerve conduction study (NCS) and electromyogram (EMG) to test nerves and muscles, CT Scan, or MRI (Magnetic Resonance Imaging) of the brain and/or spine, and extensive blood work. Sometimes muscle and/or nerve biopsies are performed. The diagnostic process involves ruling out other potential causes of the symptoms the person is exhibiting. Because there is no conclusive test, people may find themselves with a diagnosis of *probable* or *possible* ALS until further identifying symptoms appear.

A diagnosis of ALS can be devastating to both the individual and the family. Anger, denial, fear, and a sense of loss are common reactions. For those who have been through a lengthy diagnosis process and the anguish of not having a diagnosis, finally knowing that they have ALS may actually bring a feeling of relief.

Treatment and Management of ALS

The Physician's Role

The American Academy of Neurology has established guidelines for physicians treating individuals with ALS. These are based upon the following four principles:

1. Patient self-determination and autonomy are priorities; however, the way in which care and information are delivered to the person with ALS and/or his or her family should take into account the social, psychological, and cultural background of the family. For example, in some instances the patient may prefer not to know the diagnosis and/or prognosis and the family may appoint someone else as medical decision-maker.
2. Physicians should provide the family with information well in advance of the time that decisions will have to be made regarding treatment options. Everyone involved should understand that decisions may change over time.
3. Caring for a person with ALS requires a coordinated effort among many people including family caregivers, therapists, nurses, and physicians.

The physician should be responsible for ensuring the coordination of care from the time of diagnosis to the last stages of the disease.

4. A Living Will (Advance Directives, Power of Attorney for Healthcare) defining the treatment preferences of the person with ALS can be an excellent tool for communication among the person with ALS, the family, and the medical care team. However, this document should be reviewed every six months to reflect possible changes in preferences. This can often be a difficult topic, and the physician should help by offering the family a chance to talk about it and to review it. In preparation, the physician should provide the family with information about the terminal phases of the disease.

Helping people with ALS and their families requires empathy and sensitivity on the part of everyone involved.

Drug Treatment

In 1995, the FDA approved riluzole (Rilutek), which was the first drug for the treatment of ALS. C Riluzole is a medication that is taken by mouth (i.e., a pill). Clinical trials of riluzole showed that the drug had a modest effect (2-3 months) on slowing the progression of ALS.

In 2017, the FDA approved a second drug for treatment of ALS: edaravone (Radicava). Edaravone is a medication that is given intravenously (i.e. infused into a vein). A small clinical trial showed that edaravone slowed the decline of daily function in a limited group of PALS who were in the early stages of ALS. However, in a clinical trial involving a larger group of PALS, the drug did not show benefit compared to placebo.

While there is no cure for ALS, researchers continue to conduct clinical trials of potential new treatments for ALS. People with ALS (PALS) may want to discuss with their physicians the option of participating in a clinical trial. Depression, uncontrollable laughing/crying, cramps, insomnia, spasticity, and drooling can be treated with medications and should be brought to the attention of the physician.

Living with ALS

While it is true that there is no cure for ALS, much can be done to help the person live with the disease. Treatment aimed at relieving symptoms can be very effective. Generally, people with ALS should continue their usual daily activities, stopping before they become fatigued. They should be encouraged to set their own limits of exertion and to plan how they will use their energy and strength. The physician will probably suggest exercises, including breathing exercises, to strengthen unaffected or less-affected muscles. These exercises are not vigorous or tiring, but are intended to help maintain mobility and prevent joint stiffness and muscle contracture.

There are a wide variety of assistive devices for enhancing the quality of life for the person with ALS:

Mobility

By working with physical and occupational therapists, PALS and their caregivers can identify appropriate devices to help maximize independence and reduce caregiver strain as changes in mobility occur. Braces, canes, and walkers can help PALS maintain independent mobility for as long as possible. As the disease progresses, wheelchairs, lifts, and other special equipment can enable PALS and their caregivers to work as a team to keep the person with ALS connected to the world around them and to ensure appropriate care.

Communication

A speech therapist can help PALS maintain their speaking ability for as long as possible; however, the loss of speech does not mean the loss of the ability to communicate. Assistive communication devices, from simple boards to sophisticated electronic devices and computer applications permit communication even into the latest stages of ALS. Rapid advances in technology have resulted in products that dramatically increase the independence of people with very limited mobility allowing them to "speak," operate lights and other controls, and remain contributing members of their families and communities. Medicare may cover some communication devices.

Breathing

As muscles weaken, breathing becomes more difficult and less effective in the person with ALS. Individuals may find that they have problems sleeping, wake up feeling tired, or wake up with a headache. Several options exist for individuals whose breathing becomes inadequate.

- Several noninvasive options to help breathing exist. One of the most frequently used is called a BiPAP (biphasic positive airway pressure). The person with ALS wears a mask over his or her nose at night connected to the BiPAP, which helps increase the flow of air into and out of the lungs.
- When the person requires more help breathing than can be offered with any of the noninvasive methods, a permanent ventilator can be used. The ventilator then does the person's breathing for him or her. This option can prolong the life of a person with ALS, but it increases the need for care and the cost of care. The willingness and availability of caregivers can impact this decision.

Deciding whether or not to use the various types of assistive breathing devices will be one of the hardest decisions PALS and their families need to make. Preferences regarding breathing support should be discussed with the physician and included in the Advanced Directives.

Nutrition

It is important for people with ALS to receive proper nutrition and to maintain a normal weight. As problems with swallowing develop, food can be processed to make it easier to swallow. A nutritionist or registered dietitian and speech therapist or speech pathologist can help develop strategies to ensure that the person is receiving enough nutrients, calories, and fluids. If the person is not getting enough food or if choking and food inhalation become a problem, he or she may want to consider a feeding tube. The tube, referred to as a PEG, is surgically inserted into the person's stomach. Food goes through the tube and enters the stomach directly. This can be another very difficult decision for PALS and their families. Preferences regarding the placement of a PEG should be discussed before the need arises and included in the Advanced Directives. As with all treatment decisions, they should be reviewed periodically.

Gathering the appropriate resources to meet the challenges of ALS can be financially difficult for many families. Both the Amyotrophic Lateral Sclerosis Association (ALSA) and the Muscular Dystrophy Association (MDA) have programs through which people with ALS can borrow assistive devices and equipment that they might otherwise not be able to afford.

Reaching Out

It is important that the person with ALS and his or her primary caregiver get the information and the support they need. Financial, emotional, and caregiving challenges can take their toll on PALS and their caregivers. Preparing for upcoming

changes, establishing good communication with the physician and other medical team members, and pulling together a supportive community of family, friends, and social service professionals will help ensure the highest possible quality of life. While the care needs of PALS can be intense, caregivers must also be aware of their own needs for respite, rest, and emotional support. A diagnosis of ALS does not mean an end to enjoyment. PALS and their caregivers will find that they continue to laugh, to find joy in their lives, and to gain strength from the love of their families and friends.

Resources

Southern Caregiver Resource Center
891 Kuhn Drive, Ste. 200
Chula Vista, CA 91914
(858) 268-4432 | (800) 827-1008
E-mail: scrc@caregivercenter.org
Website: www.caregivercenter.org

The Southern Caregiver Resource Center offers services to family caregivers of adults with chronic or disabling conditions in San Diego and Imperial counties. Services include information and referral, needs assessments, care planning, family consultation, case management, individual counseling, legal and financial consultation, respite care, education and training, and support groups.

Family Caregiver Alliance
(415) 434-3388 | (800) 445-8106
Website: www.caregiver.org
E- mail: info@caregiver.org

Family Caregiver Alliance (FCA) seeks to improve the quality of life for caregivers through education, services, research and advocacy. FCA's National Center on Caregiving offers information on current social, public policy and caregiving issues and provides assistance in the development of public and private programs for caregivers.

ALS Association (ALSA)
www.alsa.org

The ALS Motor Neuron Treatment & Research Center at UCSD
www.health.ucsd.edu

The Muscular Dystrophy Association (MDA)
www.mdausa.org

International Alliance of ALS/MND Associations on the Internet
www.alsmndalliance.org

National Institute of Neurological Disorders and Stroke
www.ninds.nih.gov/Disorders/All-Disorders/Amyotrophic-Lateral-Sclerosis-...

Paralyzed Veterans of America
www.pva.org/als-stories/outreach-to-veterans-with-als

National ALS Registry
www.cdc.gov/als

This fact sheet was prepared by Family Caregiver Alliance in cooperation with California's Caregiver Resource Centers and reviewed by Richard K. Olney, M.D. in 2001. Updated by Janice C. Wong, M.D., and reviewed by Catherine Lomen- Hoerth, M.D., The ALS Center at UCSF in 2018.

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