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LIVING WITH ALS

What's It All About?

ALS Association
Fighting Lou Gehrig's Disease

Cover: *Renaissance Scene* by Jean Watson.
The artwork on the covers of the *Living With ALS* manuals was created by individuals with ALS or their family members.

Message from the Artist

"I appreciate and do not take for granted the ability to feel renewed after a night's sleep, how ice-cold water can quench thirst and relax my throat, or the range of tastes and textures and smells of all the different foods. I enjoy the cycle of morning, afternoon, and evening light; the many variations of a setting sun. The blue sky, clouds, sounds of birds, rushing wind, and rain are pleasures for the senses. Colors of green, the infinite number of blues of our Lake Superior, of autumn, and the winter snow are all there for you to see. There is a world of delight in a child or a grandchild's smile, or in a friend's greeting.

We have so much life in us and around us. I believe you should think and look for the positive and you will feel and act positive. Through this you will be really alive each and every day."

Jean Watson
Artist
Person living with ALS

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Manual 1

LIVING WITH ALS

What's It All About?

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A note to the reader: The ALS Association has developed the *Living With ALS* manuals for informational and educational purposes only. The information contained in these manuals is not intended to replace personalized medical assessment and management of ALS. Your doctor and other qualified health care providers must be consulted before beginning any treatment.

LIVING WITH ALS
What's It All About?

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Introduction

You, a family member or a friend, have been told that **amyotrophic lateral sclerosis** (**ALS**) – a motor neuron disease (MND) – is now a part of your life. The purpose of this manual is to provide hope and help you recognize how much can be done to assist people with ALS to live their lives with high quality. The information will give you an overview of ALS, answer many of your questions, and let you know that there are people and organizations to help and support you. Think of this manual as your own personal resource guide which you can refer to for the following:

- basic information about the disease itself
- insurance information
- health care providers
- medical equipment and supplies
- employment and disability issues
- funding sources
- community resources
- numerous other specific and general resources and services

The first section of this manual covers the basic information about ALS and how it affects the body. The second section covers the range of resources available to help you cope with the disease and also explains how you can use them effectively.

Questions and Answers about ALS

WHAT ARE MND AND ALS?

In many areas of the world, ALS is known as **motor neuron diseases** (**MND**). Motor neuron diseases are characterized by progressive muscle weakness and atrophy (wasting away or loss of muscle). There are several types of motor neuron diseases. If only the upper motor neurons (originating in the brain) are damaged, it is called **primary lateral sclerosis**. When only the lower motor neurons (originating in the spinal column) are damaged, it is referred to as **spinal muscular atrophy** or **progressive muscular atrophy**.

If both the upper and lower motor neurons are damaged, it is known as **amyotrophic lateral sclerosis** (**ALS**), which is the most common form of MND in adults of any age. Once the motor neurons die, they can no longer send messages to the muscles. This breakdown in the sending of nerve impulses to the muscles is what causes the weakness in ALS.

WHEN WAS ALS FIRST RECOGNIZED?

ALS was identified as a specific disease by Jean Martin Charcot, a pioneering French neurologist working in Paris in the late 1800s, and thus is still sometimes called Charcot's disease in France. Relatively little progress was made in understanding ALS until the 1990s, when there were major research efforts with encouraging results.

WHY IS ALS ALSO REFERRED TO AS "LOU GEHRIG'S DISEASE"?

Lou Gehrig was a famous baseball player for the New York Yankees. He played in more consecutive baseball games than any other player, until his record was broken by Cal Ripken, Jr., in 1995. For his efforts Lou Gehrig was a symbol of indestructibility – the "iron man" of baseball. He was nicknamed the "Iron Horse." On May 2, 1939, he pulled himself out of the lineup of players "for the good of the team." He was not playing well and knew that something was physically wrong. Within a few months, Mr. Gehrig was diagnosed with ALS. He died two years later. In the United States, ALS is often referred to as "Lou Gehrig's Disease."

WHAT DO THE WORDS "AMYOTROPHIC LATERAL SCLEROSIS" MEAN?

A-myo-trophic comes from the Greek language. *"A"* means no or negative; *"myo"* refers to muscle, and *"trophic"* pertains to nutrient or nourishment – no muscle nourishment. In this case, however, nourishment does not refer to food, but to signals or messages sent by the motor neurons in the brain and spinal cord to the muscles – signals or messages that the voluntary muscles need to work properly. When these signals can no longer reach the muscles, the result is weakened muscles that *"atrophy"* (shrink) and later become paralyzed. Atrophy of muscle is called *"amyotrophy."*

The upper motor neuron axons travel in a bundle along each side (**lateral** edges) of the spinal cord. You can think of the upper motor neuron axons as if they were telephone wires that are bundled in cables. The cables run down the side (**lateral** area) of the spinal cord. When the upper motor neurons die, they leave behind a scarred area on the lateral portions of the spinal cord. **Sclerosis** is a general medical term that means hardening and scarring.

Therefore, **lateral sclerosis** refers to scarring along the sides of the spinal cord caused by the death of upper motor neurons.

The words "**amyotrophic lateral sclerosis**" are simply descriptive words used to illustrate what Dr. Charcot saw in people with ALS. He observed shrunken muscles (**amyotrophic**) and scarring in the lateral area of the spinal cord (**lateral sclerosis**).

WHAT ARE MOTOR NEURONS?

ALS is a disorder of a specific nerve – those that tell your muscles what to do. Because muscles make you move, the nerves that control movement are called **motor nerves** or **motor neurons**; the words **nerves** and **neurons** have the same meaning. Without the ability of motor nerves to send messages to muscles, the muscles do not work properly and you become weak.

It is important to know that ALS only affects your voluntary muscles – the muscles that **you** can voluntarily control. For example, you can voluntarily move your arm. ALS does **not** affect muscles that you can not voluntarily control. For example, you can not make your heart beat faster or slower. The heart is not a voluntary muscle and it is not affected by ALS.

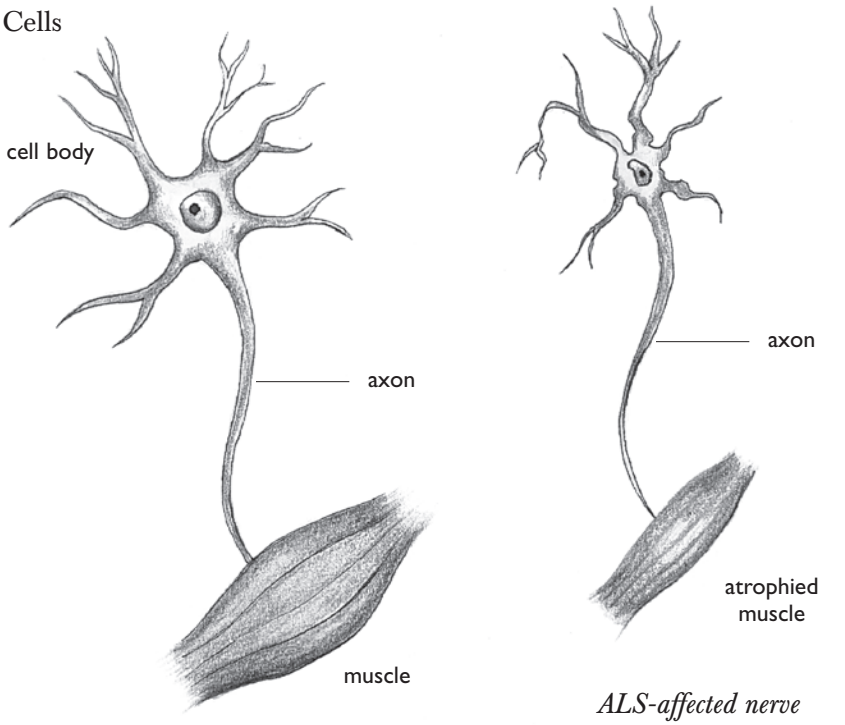
WHAT DOES A NEURON DO?

Neurons or nerves are the **electrical wires** of the human body. They accept a **chemical signal**, convert it into an **electrical impulse**, send that impulse along the **nerve fiber** (**axon**), and then transfer the impulse to another nerve or to a muscle.

Nerve cells have three main sections: the **cell body**, the **long tail** (**axon**), and the **branches at the end** (**terminal branches**) (Fig. 1). The **cell body** serves as the "**brain**" of the nerve; it accepts the chemical message, processes it, converts it into an impulse, and sends the impulse down the axon. The axon is like the wire in an electrical cord, carrying the message from one area to another.

At the end of the axon, the nerve splits into hundreds of branches. These terminal branches reach out to the cell bodies of other neurons or to muscle fibers. Thus, one nerve impulse travels to another nerve and then to muscle fibers, until the message can finally do its job to produce movement. This information travels through the body at a relative speed of over 200 miles an hour!

FIGURE 1
Nerve Cells



Normal nerve cell and muscle

ALS-affected nerve cell and muscle

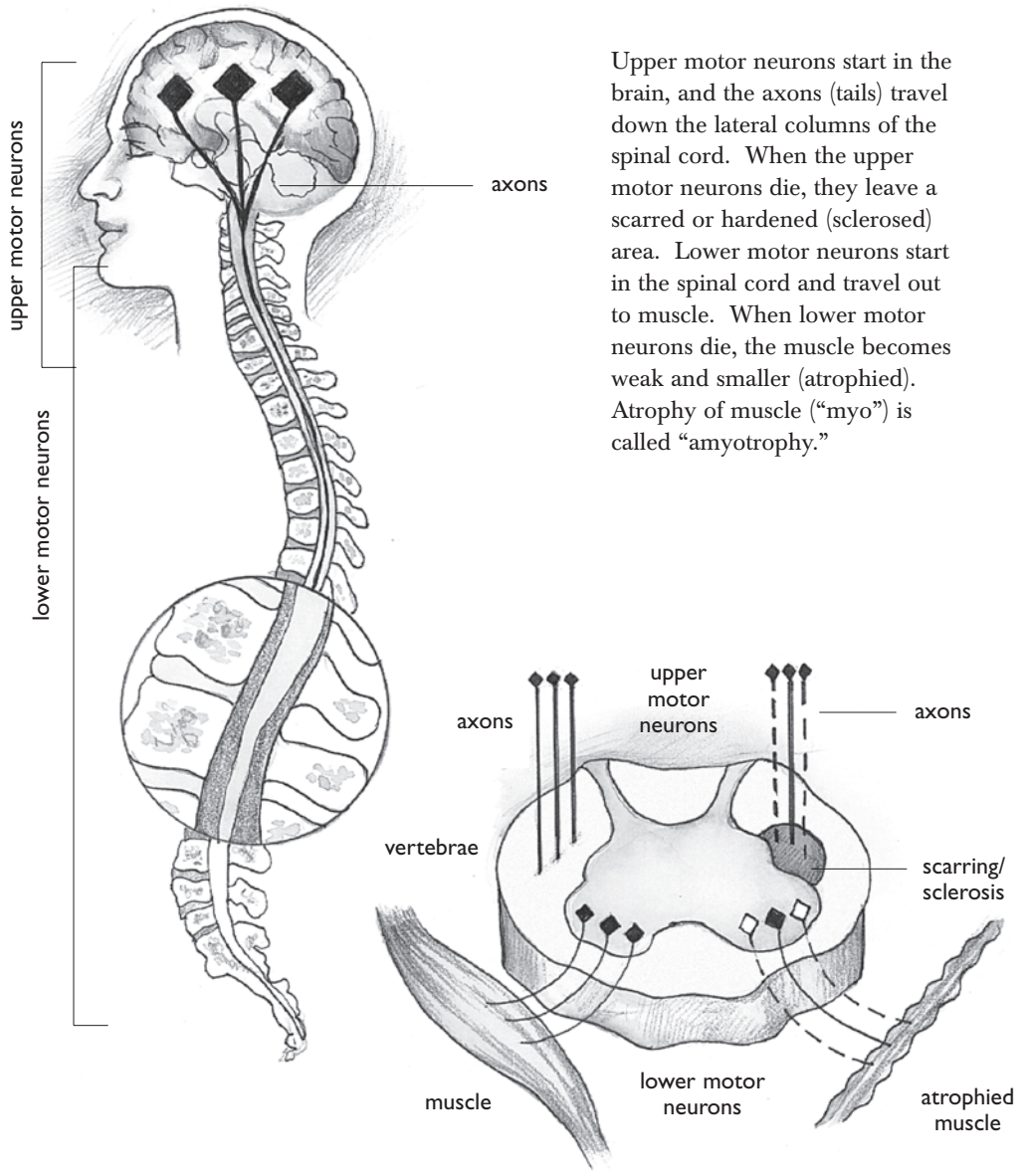
WHAT TYPES OF NERVES MAKE YOUR BODY WORK PROPERLY?

The body has many kinds of nerves. There are nerves involved in the process of thinking, memory, and of detecting sensations (such as hot/cold, sharp/dull), and others for vision, hearing, and other bodily functions. The nerves that are affected when you have ALS are the motor neurons that provide **voluntary movements** and **muscle power**. Examples of voluntary movements are your making the effort to reach for the phone or step off a curb; these actions are controlled by the muscles in the arms and legs.

The heart and the digestive system are also made of muscle but a different kind, and their movements are not under voluntary control. When your heart beats or a meal is digested, it all happens automatically. Therefore, the heart and digestive system are not involved in ALS. Breathing also may seem to be involuntary. Remember, though, while you cannot stop your heart, you can hold your breath – so be aware that ALS will eventually have an impact on breathing. (For more information about breathing, see Manual 6.)

FIGURE 2

Motor neurons



Upper motor neurons start in the brain, and the axons (tails) travel down the lateral columns of the spinal cord. When the upper motor neurons die, they leave a scarred or hardened (sclerosed) area. Lower motor neurons start in the spinal cord and travel out to muscle. When lower motor neurons die, the muscle becomes weak and smaller (atrophied). Atrophy of muscle ("myo") is called "amyotrophy."

Detailed slice of the spinal cord showing healthy lower motor neurons/muscle on left side and ALS-affected lower motor neurons/muscle on right side.

HOW DOES A VOLUNTARY MOVEMENT OCCUR?

It may be helpful for you to understand how normal movements occur, before discussing how ALS affects muscle control (Fig. 2). If you want to pick up a pencil, for example, your brain makes up a complex plan, which is sent to motor nerves in the brain called **upper motor neurons**. The cell bodies of these neurons are in a special part of the brain called the **motor cortex**, and their tails or axons travel down the spinal cord.

It is the job of the upper motor neurons to send messages from the brain to the **lower motor neurons** and their cell bodies in the spinal cord. The tails and branches of the lower motor neurons go out to muscles and send the message to individual muscle fibers, which must be activated before they contract and perform the desired task – i.e., picking up a pencil. And it all happens in an instant. To summarize, picking up a pencil can be simplified into four basic steps:

1. You think a thought (I want to pick up the pencil) and your brain makes a plan.
2. The upper motor neurons receive the plan and send it to the lower motor neurons.
3. The lower motor neurons receive the plan and send it to the muscle fibers.
4. The muscles do their job and you pick up the pencil!

WHAT CAUSES NERVES TO DIE?

To date, no one completely knows what causes the death of the motor neurons in ALS nor exactly how it occurs. It is thought that the cell bodies of the motor neurons are damaged and die. Once the cell body dies, the remaining parts of the nerve, such as axons and terminal branches, cannot operate and also die. The result is that muscles do not receive the necessary messages and therefore do not function properly. They become weak.

WHAT CAUSES ALS TO DEVELOP?

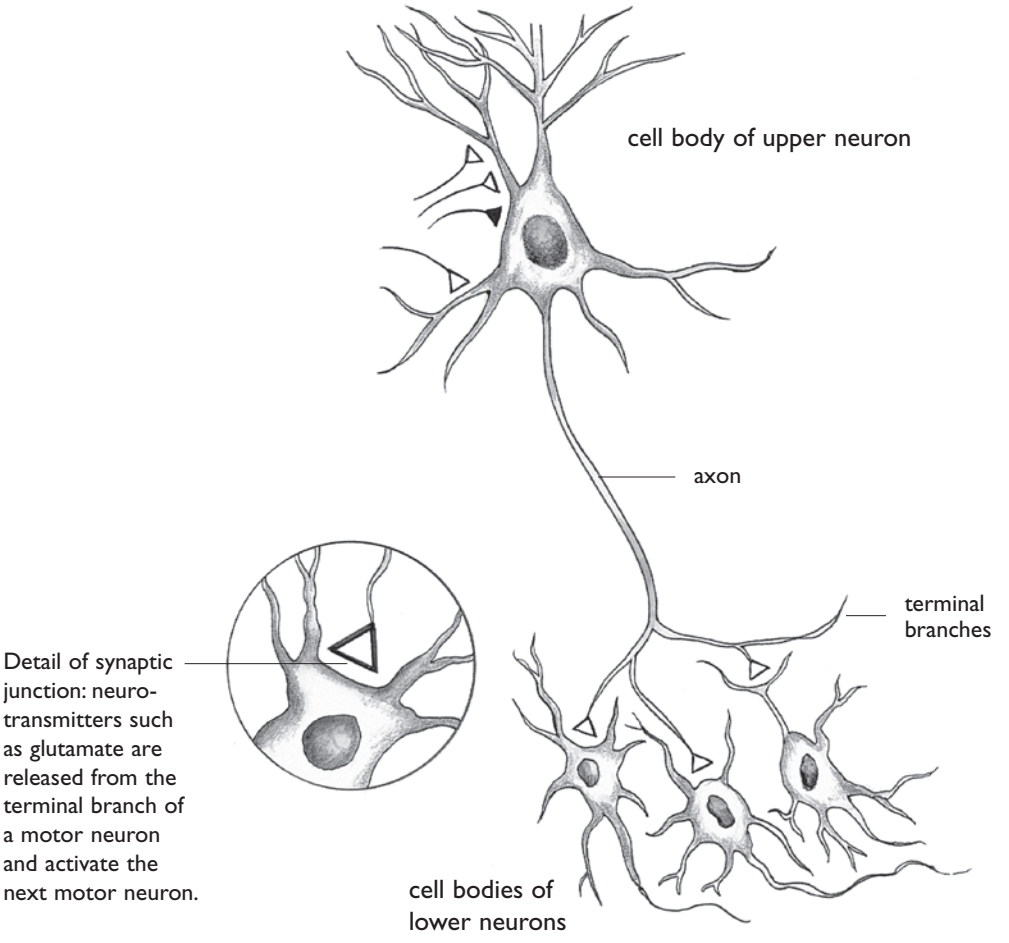
Many research studies have focused on finding the cause(s) of ALS. Although recent research has led to promising new clues, and advances in the tools needed to expand research, the answers have not been found. It is easier to begin with what is not a causative factor. The disease is nothing that anyone did to you, nor anything that you did to yourself. You cannot "catch" ALS. ALS is not contagious.

A small percentage of ALS is inherited (familial ALS) meaning that it occurs more than once in a family lineage. One of the major breakthroughs in the 1990s in ALS research was the identification of the mutant SOD1 gene associated with about 20% of familial ALS cases. Genetic research has been accelerated to identify other mutant genes associated with the remaining 80 % of inherited ALS that will contribute to our understanding of both familial and sporadic ALS. (See page 22.)

The field of ALS research and the scientific understanding of the disease is rapidly expanding. Several compounds are in clinical trials, and there are plans to begin new trials in the near future. While ALS is certainly a serious disease, it is far from hopeless.

FIGURE 3

Synaptic junction



WHAT ARE SOME THEORIES ABOUT MOTOR NEURON DEGENERATION AND THE DEVELOPMENT OF ALS?

Excess Glutamate

Messages must be sent from one neuron to another in order for your body to function. Neurons do not actually touch one another; there is a tiny space between them called the **synapse** or **synaptic junction** (Fig. 3). A special chemical, called a **neurotransmitter**, is released from one neuron to help the message cross the synaptic junction so it can be received by the next neuron. This neurotransmitter is called **glutamate,** an amino acid (organic compound necessary for metabolism or growth).

You need a certain amount of glutamate for motor nerves to communicate, but too much can be toxic to nerve cell bodies. People with ALS have too much glutamate in their spine and the excess glutamate damages the motor neurons. A major area of ALS research has been aimed at finding drugs that will reduce the amount of glutamate in the synaptic area between neurons.

Viral Mechanism

A viral mechanism has long been hypothesized. In past years publications describing the presence of certain viruses in post-mortem spinal cord tissue of ALS patients, but not in control tissue, rekindled an interest in understanding the possible link between viruses and ALS. More recent studies failed to confirm earlier research. Viruses can trigger inflammation and that in itself is an area of keen interest among researchers studying neurodegenerative diseases.

Inflammation/Inflammatory Response

There is growing interest in the role of inflammation in neurodegenerative diseases. Several investigators have shown evidence for the possible role of inflammatory responses in ALS – perhaps as one mechanism involved in the chain of events in the cell death process. Whether inflammatory mechanisms are one of the primary causes of ALS or secondary to other pathological mechanisms is unclear. Inflammation can be triggered by invading microbes such as viruses or bacteria; injurious chemicals, or by physical injury. The trigger may come from within the organism such as a disease affecting the immune system or nervous system.

In neurodegenerative diseases such as ALS, inflammation might result from any one of a number of causes: abnormal protein accumulation, molecules released from or associated with injured cells, or even imbalances between pro- and anti-inflammatory processes. Inflammatory factors have multiple roles. Inflammatory responses also recruit immune mechanisms. Microglia and astrocytes, cells that support the neurons, are the main cells that initiate inborn immune response in the central nervous system (CNS).

Environment

No conclusive evidence exists that proves environmental factors influence development of ALS, although there are suggestive findings. Thus, more research is needed to understand the role environment may play. This would include epidemiology studies of what appear to be "clusters" of ALS cases in a given geographic area; and more studies of groups such as Gulf War and other military veterans who, according to early research data, may have a higher incidence of ALS than is considered normal for a population of that size and age.

Axonal Transport Abnormalities

Abnormal transport of important molecules from nerve endings to the cell body is seen early in disease progression in the ALS mouse model. Alterations in the transport mechanism are thought to play a key role in motor neuron degeneration. Recent research has identified mutations in two proteins (one in transgenic ALS mice, the other in humans) essential to normal axonal transport function.

WHAT ARE SOME OF THE SIGNS AND SYMPTOMS OF ALS?

Signs are what the physician can see, and **symptoms** are what you experience. The major sign as well as symptom of ALS is that the muscles become smaller and steadily get weaker over months and years. The muscles that can be affected are those that you can voluntarily control. This progressive weakness causes difficulty with your daily activities such as moving about, lifting or carrying objects, caring for yourself, speaking, chewing and swallowing, and, eventually, breathing.

You also might have "charleyhorse"-type muscle cramps, twitches under the skin called **fasciculations**, weight loss, stiffness, or poor coordination. Some people may experience exaggerated laughing or crying, or feel like their yawning is forced. You also may experience **urinary urgency**, which means that you feel like having to go to the bathroom right away, but only because the signal is too late.

A number of signs and symptoms of ALS have been listed above, but *not everyone with ALS develops all of them*; however, almost every person with ALS has at least some muscles become markedly weak and smaller. Eventually, there is progressive weakness of the **diaphragm** (the main muscle for breathing, located above the abdominal cavity) which, when healthy, keeps the lungs moving properly. (Manual 3 offers a discussion of these symptoms.) The diaphragm can be thought of as the motor that runs the lungs. The motor neurons can be thought of as the wires that supply electricity (power) to the motor. Without the power supply (motor neurons) the diaphragm can not make the lungs work.

HOW DOES ALS PROGRESS?

ALS begins in one region of the nervous system and causes the upper and lower motor neurons to die in that area; then the muscles they control become weaker and smaller. The strength of any voluntary muscle group can be affected in ALS, including those muscles that control facial expressions, chewing, swallowing, speaking, the neck, arms, trunk, legs, and breathing. ALS can start in any muscle group and then move to any other; however, there is no clear prediction of where the weakness will spread next. The only exception is that when one arm or leg is involved first, then the opposite arm or leg is likely to weaken next.

WHAT KINDS OF TREATMENT ARE AVAILABLE FOR ALS?

Although to date there is no medication available to stop or cure ALS, there is a great deal that can be offered to manage the symptons. One drug, Rilutek®, is approved by the FDA and has been shown to modestly increase survival. A great deal of dedicated time and money is being invested in researching drugs that would slow down the progression of weakness and/or increase survival. Ask your ALS doctor for details about what these drugs can do, potential side effects, and cost. There are medicines that can help with the symptoms of cramps, stiffness, exaggerated laughing and crying, forced yawning, and urinary urgency. You, along with your doctor, must weigh many factors when deciding if a particular medication is right for you. (See Manual 3.)
There are procedures and equipment that can help you with speaking, swallowing and breathing problems.

WHEN AND HOW LIKELY IS IT FOR ALS TO OCCUR?

The annual incidence (number of new cases per year) of ALS is two per 100,000 population. According to past epidemiologic studies, the prevalence of ALS in the U.S. (number of cases at any given time) is said to be between six and eight per 100,000 population. Based on the 2000 census, this would mean there are as many as 22,500 Americans with ALS at any given time using the prevalence of eight per 100,000. However, as many as 30,000 Americans may have ALS at any given time, given the difficulty in diagnosing ALS and the lack of required case reporting. The annual mortality rate attributed to ALS is two deaths per 100,000 population in the U.S. The age of onset is most common between the ages 40 and 70, with the peak age about 55.

WHAT ARE THE TYPES OF ALS?

Sporadic ALS

In 90% to 95% of the people with ALS, the disease just seems to happen. No other family members have ever had the disease. This is called "sporadic" ALS. No one knows why or how sporadic ALS starts although there are numerous theories being investigated.

Familial ALS

About five to ten percent of ALS cases are called "familial" ALS. People with familial ALS (FALS) have other family members who have had ALS. FALS is caused by a mutation or abnormality in a gene. The majority of FALS is inherited in an autosomal dominant manner, which means that each time a FALS patient conceives a child there is a 50% chance the child will inherit the abnormal gene. Most, but not all, who inherit the abnormal gene will go on to develop the disease later in life. Why this is so is not yet understood. It is possible that someone who has the abnormal gene could pass the gene on to a child without showing any sign of the disease himself/herself.

It should be noted that there is no discernible difference in how sporadic and familial ALS affect the person with the disease. The symptoms and progression of the disease, whether sporadic or familial, are the same. It is the presence of a family history of the disease that is the one known distinction.

Thus far one gene has been identified with about 20% of FALS cases. This is the gene that codes for a protein called superoxide dismutase 1 or SOD1. The main function of normal SOD1 is the "clean up" or detoxification of free radicals that are byproducts of cellular metabolism (waste products of the work of the cell). When abnormal SOD1 was identified in 1993 as a causative gene for FALS, it was assumed that it led to a loss or decrease of function of the protein that would result in a buildup of toxic free rad-

icals. However, research studies suggest the problem is not a loss of function, but rather the gain of a new and toxic function -- one that is not yet understood. This continues to be an area of intense laboratory study. There are now more than 100 mutations in SOD1 associated with ALS. It is hoped that understanding how SOD1 causes FALS will help us understand the mechanisms involved in the development of sporadic ALS cases as well.

Extensive efforts are also underway to identify the genes that cause the remaining FALS cases. Thus far there are several locations that have been associated (linked) with the disease, but no specific genes have been identified to date. Chromosome 16 and the X chromosome have been associated with dominantly inherited FALS.

Also, there are locations on chromosomes 2 and 15 for rare, recessive forms of FALS, in which the faulty gene must be inherited from both parents for the disease to occur. In these rare cases of inherited ALS (sometimes referred to as juvenile ALS), there is an early onset of ALS with the disease having a long duration. The abnormal gene on chromosome 2 has been identified and its protein *alsin* is the subject of several research studies. Again, it is hoped that information from this research will yield answers about sporadic ALS.

Also, recently a location on chromosome 9 has been identified for FALS that is associated with frontotemporal dementia. Although most people with ALS do not develop any mental impairment, new research reveals that a small percentage of patients, 10-15%, may have symptoms of frontotemporal (FTD) dementia. Symptoms of FTD may include personality changes and difficulty with work generation, memory or other executive functions. The relationship between ALS and FTD is not completely understood, but remains under investigation by researchers.

FALS AND GENETIC TESTING

People often ask if they should have their blood tested for familial ALS. Since SOD1 is the only known abnormal gene that causes FALS, the only test currently available is one for an abnormal SOD1 gene. Remember that SOD1 FALS accounts for only 20% of all FALS cases and FALS represents about 10% of all ALS. This means that only some two to three percent of all ALS is caused by mutations in SOD1.

If there is no history of ALS in the family, SOD1 testing will most likely not give you any useful information. If there is a family history of the disease, it is by definition familial, but it may not be caused by a mutation in SOD1. In fact, it is more likely that it is not.

If a person with ALS had the SOD1 blood test and a mutation was detected, that would raise the possibility of testing unaffected family members. Anyone in whom a mutation was identified would likely develop ALS if he or she lived long enough, and would have a 50% chance of passing the mutation to each of his or her children. Now, if a person from a family in which a mutation in SOD1 had been detected (in another family member with ALS) was tested, and no mutation was detected, that person would not get ALS and could not pass it on to his/her children.

Let's consider another situation in which there is a family history of ALS. A person with ALS in that family takes the SOD1 test and it comes back negative (no SOD1 mutation found). The ALS is still familial because of the family history, but it is not caused by the SOD1 mutation. Instead, it is caused by an, as yet, unidentified gene. For that reason, no further testing can be done. Also, testing for the mutant SOD1 gene in unaffected members of their family would be meaningless because one would not be looking for the gene that causes the disease in *this* family. There is no current blood test that would give any information to these family members about whether or not each person will later get ALS. All we can could say is that each child of an affected person has a 50:50 chance of inheriting the abnormal gene, and if that gene is inherited, a high likelihood, but not absolute certainty, of developing ALS.

A special note: Testing unaffected persons in SOD1-FALS families should be thought about very carefully. The result of the test, whether positive or negative, can have a large impact on a person's life and emotional health. It is strongly recommended that a person without symptoms talk with their neurologist and a genetic counselor before having SOD1 testing. Testing of minor children is highly controversial.

WHAT DOES THE WORD "BULBAR" MEAN?

The **bulb** area of the central nervous system is located between the brain and the spinal cord. Also known as the **medulla**, the bulb was named by physicians who studied anatomy over a century ago and who were looking inside of human beings for the first time. They did not know its purpose, but since it looked like a tulip bulb, they named it the "bulb."

The term **bulbar** refers to the motor neurons located in the bulb region of the **brain stem** which control the muscles of chewing, swallowing, and speaking. When these muscles become weak and uncoordinated, the diagnosis of **bulbar**, **pseudobulbar**, or **progressive bulbar palsy** is made; eventually this condition progresses to ALS and causes the weakening of arm, leg, and respiratory muscles as well. Remember that not everyone with ALS becomes weak in all of his or her muscles. For example, some people may never need a wheelchair, but about 25% of people with ALS start with weakness in their legs. Likewise, some people will never have a weakness of the bulbar muscles, but about one third of people with ALS start with bulbar weakness.

WHAT IS NOT AFFECTED BY ALS?

Muscles such as those that move the eyes and the **external sphincters** that hold stool in the bowel and urine in the bladder are very rarely involved in ALS. Other areas not affected are: internal organs (such as the heart, liver, and kidneys), sexual functions, and the five senses (sight, hearing, smell, taste, and touch/pressure). The mind and mental abilities are affected infrequently.

HOW WILL ALS AFFECT YOU?

ALS is a progressive disorder, which means that you will become weaker over time, gradually but steadily. The weakness will occur in new areas as well. The rate of progression varies from person to person and can be slow, moderate, or rapid. For some people, there may be a markedly slowed-down rate in the later stages of the disease. Between 15% and 25% of people with ALS live 10 years after they first noticed symptoms; moderate or average progression is over two-to-five years; rapid progression is respiratory failure within a year, which is very unusual. Thus, there is a great deal of variation in the progression of this disease.

It is important to distinguish the difference between the rate of progression of the disease and the length of survival of a person with ALS. There are many things that a person can choose that will lengthen their life beyond the natural course of the disease.

WHAT IS THE BEST WAY FOR YOU TO MANAGE ALS?

A team effort will benefit the management of your ALS. The team may be large or small, and members will vary according to your needs and the resources available in your geographic area. The health care professionals on your team should have experience and knowledge about ALS. Their goal is to present the most appropriate options to you and offer support as you make decisions about your care.

The medical team can include your neurologist, primary care doctor, and nurse. Other members, such as social workers and therapists, can assist with insurance, social issues, and with your emotional state. Depending on your needs, speech therapists, physical therapists (PTs), and occupational therapists (OTs) are very helpful. Some medical centers have comprehensive ALS clinics; for the one closest to you, please contact the Patient Services Department at the National Office of The ALS Association.

Additionally, family and friends are important members of your team. For information on additional resources contact your ALS Association chapter or call The ALS Association National Office Patient Services Department at 1-800-782-4747.

The Last Word

Now that you have an overview of what ALS is all about, remember that you are not alone. There are people available to help you through this journey with ALS. Those who know a lot about ALS can give you advice and support so that you can be independent as long as possible.

Your entire health care team views your *quality of life* as its most important commitment. The members of your team will work together with you and help you access all the support available to address the needs of both your body and spirit.

In the next section of this manual you will learn how your doctor, nurse, and other ALS team members can help provide direct care, and how national and regional organizations are ready to offer information and support.

Resources and Resourcefulness

Life is full of joys, sorrows, and uncertainties, and how people respond is as distinct and different as the stars in the sky. Being diagnosed with ALS is an event that will challenge your emotional resilience, and the way you cope with uncertainty will be a factor in your overall adjustment. This section will help you to become resourceful in your fight against this disease.

Your financial security may be closely tied to the degree of coverage that your insurance provides for the items and services you need. This coverage frames each stage of your illness, and knowing the details of your insurance will assist you in planning for the future.

Many people with ALS are worried about whether or not they have adequate insurance for the evaluations, services, and necessary supplies. Finding out what your insurance plan does and does not cover will aid in maximizing the benefits that you receive, while minimizing your out-of-pocket expenses. Learning how to access community resources, obtain medical equipment, and purchase adaptive devices will empower you for an easier transition to each stage of the illness. Your preparation will strengthen your ability to cope with the realities of everyday life with ALS.

THE ART OF BEING RESOURCEFUL

Being resourceful is an attitude. The following three suggestions that will aid you in becoming your own best advocate while you investigate your insurance policy, as well as while you communicate with health care professionals.

1) Ask Questions.

Insurance terms and medical jargon can be like a foreign language. Do not be intimidated by how unfamiliar you are with the health care system or in dealing with insurance matters. The more you ask, the more informed you will be.

2) Assume Nothing.

Review your understanding of your insurance policy with the customer service representatives (phone numbers are usually listed on insurance cards). Review how to use your benefits: How do you obtain medical equipment – through your doctor or on your own? Can you take the order to any supplier, or are you directed to use a preferred provider chosen by your insurance plan? Do you need a referral or prescription?

3) Don't Take "NO" for an Answer.

If you are told a piece of equipment or anything you may need, which is ordered by your doctor, is not covered by you insurance, first determine whether the denial is based upon lack of coverage or the insurance company's belief that the service is not medically necessary. Then ask what the appeal process is and follow it exactly. Many people are surprised when a denial is reversed. Some people do not have coverage for **durable medical equipment** (DME – medically necessary reusable equipment), but may have coverage for respiratory equipment. Be as specific as possible when talking with your insurer, and also ask for the assistance of the doctor, his/her staff, or a social worker in proceeding with an appeal. Be diligent and persistent.

Sometimes an insurance decision maker is not knowledgeable about ALS; you need to educate them about how the service, equipment, or prescription is medically appropriate for this disease. He/she may then authorize the request. Of course, it is not always possible to receive benefits for items not outlined in your plan, but you will never know unless you try.

Remember, insurance policies vary from person to person. If you and your neighbor both have Blue Cross through your employers, he or she may have a very different plan from yours and may include different benefits. Do not assume that specific insurers offer the same coverage. Your plan may cover medical equipment at 100%, while your neighbor's is at 80%.

Refer to the outline of questions in the next section for guidance on what to ask your insurance representative. The areas where you should focus are: DME, home health care, private duty nursing (particularly necessary if considering ventilator support), and prescription benefits**.**

It is helpful to maintain a written "log" or record of the names of the people with whom you speak and correspond. Include their contact information and the agency or organization along with the date of your communication, issues discussed and the outcome of the conversation or written communication.

Understanding Your Insurance Coverage

In order to maximize the benefits of your insurance plan, it is important for you to keep a record of the name, phone number, identification, and group number of your insurance plan. You also should note who the subscriber is (you or your spouse), his/her date of birth, and Social Security number. Identify any other insurance benefits you may have, such as a separate prescription benefit card or a long-term-care policy.

If you have more than one insurance plan, determine which policy is primary and which is secondary; confusion with this matter can result in billing errors. Your insurance company customer service representative can be helpful in identifying which plan is to be billed first. Also keep up to date on your insurance; if your coverage changes in any way, notify all your doctors, medical suppliers, and pharmacists immediately, so that the next time you access their services, you will have helped prevent billing errors.

Contact your insurance company directly, and ask specific questions about your benefits. Keep a notebook and always note the date and the person who provided the information. Remember, having your benefits described over the phone does not guarantee coverage. Your physician may be asked to write a letter of medical necessity and complete specific forms to verify your medical condition and eligibility for whatever item or service is being requested.

Ask if your policy offers case management. Case managers either work for the insurance company, or are contracted by them to monitor and advocate for patients whose costs are high or who have complicated needs. They are often helpful in cutting internal "red tape" (problems), gaining access to little-known insurance benefits, and reducing your overall out-of-pocket expenses. Case management can be activated at various stages of your illness, depending on how your insurance company has defined the benefit. Having direct communication with your insurance carrier will help you gain an overview of your policy and remove the mystery of how your plan works.

Use the following guide when contacting your insurance company. Be sure to have all of the specific information about your insurance plan in front of you before calling. Do not hesitate to ask what certain words or terms mean.

WHAT YOU NEED TO KNOW ABOUT YOUR HEALTH INSURANCE POLICY

General Questions

1. Is there an annual deductible?
2. Is there an annual out-of-pocket expense limit or maximum? If I meet my limit, does my coverage increase and to what extent?
3. Do I have a major medical plan? Is there an annual or lifetime maximum?
4. Do I need to complete any claim forms?
5. Am I subject to pre-existing condition regulations?
6. For what services do I need pre-authorization?

Durable Medical Equipment (DME) Questions

7. Does my plan cover DME? What about ventilator and non-invasive ventilator coverage (i.e. BiPAP); are they under respiratory equipment or DME?
8. What is the percentage of my coverage?
9. Is there a preferred provider I must see?
10. Is preauthorization or a medical review required?

Prescription Questions

11. Does my plan cover prescription drugs? What are the terms of this coverage, and is coverage different based on using brand-name versus generic drugs?
12. Is there a specific pharmacy/supplier network I must use?
13. Is there a limit on the amount of prescription drugs I can get through this plan?

14. Is there coverage for *all* FDA-approved drugs, or is coverage provided only for those listed on your **formulary** (a list of drugs that an insurance policy covers)?

15. Does my plan offer a mail-order pharmacy option? Describe this benefit.

Home Health Questions

16. Does my plan have home health coverage? Describe this benefit.

17. Do I have coverage for a home health aide (for **skilled** or **custodial care**)?

18. Is there a preferred home health care agency I must use? Is there private duty nursing coverage at home? Describe this benefit.

20. Does my plan offer case management? At what point does case management get involved and for how long?

Hospice Questions

21. Does my plan have hospice coverage? Describe this benefit.

22. Is there a preferred Hospice agency that I must use?

Questions for Health Maintenance Organization/Preferred Provider Organization (HMO/PPO) Subscribers

23. Is my ALS neurologist (or other ALS specialist) a member of the network or a participating provider?

24. Explain the referral process. Do I need a referral from my primary care physician every time I go to the neurologist or other specialist; is there a limit to the number and frequency of referrals?

A NOTE ABOUT PRESCRIPTION PLANS

Find out the following details about your prescription benefit: if there is a limit, if injectable medications are covered, what the terms are, and if there is a mail-order option. If you do not have prescription coverage, or if there is a limit on coverage, explore other ways to fund your medications. Some states have pharmaceutical assistance programs or specific programs for the elderly and disabled; however, you may have to meet certain income criteria. There are some drug manufacturers that provide medications free of charge to physicians whose patients may have limited finances. Your doctor or social worker must make the initial contact with the pharmaceutical company. To find out more about these types of programs, please have your health care provider review the Pharmaceutical Manufacturers Association manual, available by calling 1(800) 762-4636, or by going to the Web site at www.phrma.org.

If you are paying for your medications privately, you may consider national pharmaceutical mail-order houses which order in high volume and pass their savings on to you. Contact your local ALS Association chapter for an updated list.

THE ECONOMIC IMPACT OF LIMB WEAKNESS

If you experience weakness in your arms and/or legs, you will want to find ways to enhance your independence and minimize safety hazards. In addition, it is important for you to know what is covered under your insurance policy, so that the economic impact of muscle weakness is kept to a minimum.

Medical Equipment

The type of medical equipment you might need to assist with daily living is different for each person and coverage for medical equipment varies from one insurance plan to another. Currently, Medicare covers, in part, most of the items listed below. A prescription is required along with a certificate of medical necessity from your doctor. Coverage issues can be complicated by the following: the *order* in which the equipment is obtained; if you reside in a nursing home; if hospice care is involved; or if you have insurance through a working spouse. You should find out from the medical supplier (store) if any coverage problems are anticipated.

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Equipment Usually Covered by Insurance

- Hospital Bed
- Walker
- Wheelchair (one)
- Commode
- Cane
- MAFO (molded ankle-foot orthosis)
- Patient Lift
- Certain communication devices

Some vendors accept Medicare payment as payment in full and others charge the portion Medicare does not cover. Ask whether your vendor is Medicare certified or not.

Adaptive Devices/Supports

To promote independence and safety at home, take advantage of grab bars, stair glides, emergency call devices, and other items listed below. These innovative products are typically not covered by insurance plans and vary greatly in their cost. Social workers or your case manager may be able to help identify ways to borrow or obtain funding for these items through county or state-funded programs, local chapters of The ALS Association, local volunteer organizations, Veteran's Affairs, or community agencies.

Equipment Usually NOT Covered by Insurance

- Grab Bars
- Shower Chairs
- Ramps
- Hand Splints
- Seat-lift Chairs
- Speaker Phones
- Emergency Call Devices
- Stair Glides
- Built-up Utensils
- Car/Van Adaptations

Help at Home

Acknowledging your limitations and needs may be awkward and difficult. Once you decide you need help at home, take the time to learn what funding is available and how to go about obtaining it. This process may be troublesome if you do not know where to turn.

Skilled Care versus Custodial (Basic) Care: Insurance companies have defined the kind of care that is provided at home as either skilled or custodial (basic).

- **Skilled care** is usually covered by insurance, but check for specific eligibility guidelines and limitations. This type of care is provided by a trained professional such as a nurse, physical therapist (PT), occupational therapist (OT), or speech therapist. These health care providers bring their professional skills and services to your home because of your difficulty getting to a medical facility. Some insurance companies that offer a home care benefit also may include a home health aide if you are also receiving care from a skilled professional.

- **Custodial care** is primarily for helping you with personal needs such as bathing, dressing, and meal preparation. This type of care can be provided safely by a home health aide or nursing assistant. Insurance companies rarely pay solely for custodial care; however, some states or counties offer this service at no cost or reduced cost if you meet certain income guidelines and/or specific medical criteria. Never assume your income is over the maximum for these programs – eligibility varies.

First, discuss your eligibility, then contact your doctor who will determine if you have a need that meets the requirements for skilled home care. If there is no skilled need present and you require help with personal care you may have to pay for a home health aide. You also can contact your local ALS Association chapter regarding resources for financial assistance or recommendations for appropriate providers.

Social workers employed by an ALS Certified Center℠, hospital or home care agency are great resources, and they are familiar with how to access well-known community programs or those in your area that may be well-kept secrets. If you exceed the income requirement for such services or if no subsidized programs exist locally, ask the social worker for recommendations for reputable and affordable home health agencies whose services you would pay for privately. You also can contact your local ALS Association chapter to inquire about financial assistance or recommendations for appropriate providers.

Nursing Home

Most ALS patients never need a nursing home but it may be necessary when all other options for care have been exhausted. Such facilities are funded by a variety of sources: Medicare, Medicaid, private insurance, and the very unpopular spending of one's life savings. If insurance helps at all with reimbursement, the criteria are very strict and may require you to be hospitalized for a few days before coverage can start. Some states have a **Spousal Impoverishment Program** which allows for marital assets to be divided more equitably, preventing the healthy spouse from becoming poverty stricken by placing their spouse in a care facility.

Speaking Difficulties

If your speech becomes difficult to understand (**dysarthria**), writing your message down or using an alphabet board may offer an easier way to communicate. When you use the phone, a **TTY** (teletypewriter) also known as a **TDD** (telecommunications device for the deaf) may offer an alternative way for you to have a conversation. It is a portable device with a typewriter-like keyboard that allows you to transmit text over telephone lines. A TTY allows someone with speaking difficulties to type a message directly to the person on the other end who has a TTY. If the receiving person does not have a TTY, message relay centers (offered by the telephone company) can read your typed message to a standard-phone user. This service is free and is described in your telephone book.

For anyone who needs to communicate other than by writing, an **AAC** (augmentative alternative communication) device is often recommended by speech and language pathologists. These systems can be sophisticated and expensive. Funding assistance for these items does exist in some communities and with certain insurance plans such as Medicare.

Swallowing Difficulties

Swallowing difficulties (**dysphagia**) can lead to weight loss; therefore, you may need over-the-counter nutritional supplements. Ensure® and similar products vary in their expense and are usually not covered by insurance unless such products are the sole source of nutrition. Carnation Instant Breakfast is tasty, less expensive, and an excellent source of nutrition when compared to Ensure®. Medicare currently covers 80% of nutritional or "enteral" formulas (such as Jevity®, Ultracal®, or Pro Balance®), when they are administered through a feeding tube and are documented as the **sole source** of nutrition. Contact your insurance plan regarding coverage for the enteral formula if a feeding tube has been put in place. (For more information on how to cope with bulbar symptoms, please read Manual 5.)

Respiratory Problems

There are two types of machines used to help you when breathing is more difficult. Non-invasive positive pressure ventilation (i.e. BiPAP ®) can **assist** your own breathing. Invasive (tracheostomy) ventilation **takes the place** of your breathing when you can no longer breathe on your own.

Non-invasive breathing machines are commonly used while you sleep. They **assist** you by adding a flow of air so that you take a bigger breath than you would take on your own. It is ordered by your physician when you have certain symptoms. It will help to increase your oxygen and decrease your carbon dioxide while you sleep. This is a common treatment and does not take the place of your own breathing.

Long-term invasive ventilation support takes the place of your breathing when you can no longer breathe on your own. Although this type of life-support is available, not everyone with ALS chooses to use it. It presents unique challenges for the patient and family. The financial impact is tremendous. The availability of caregivers, coverage for medical equipment, private duty nursing and other required resources influence this decision. (For more information on how to deal with respiratory problems, please read Manual 6.) If there are no available caregivers to help with invasive ventilation, you may find yourself residing indefinitely in hospitals or nursing homes. The latter often do not offer care for ventilator-dependent patients, so once you locate one, you may end up living farther away from home than intended. If you are considering this option, gather complete information about your private duty nursing coverage and care offered in the community, as well as local nursing home availability.

End of Life

Hospice care is an option when it is evident that you are at the later stages of your disease, and you have decided against long-term ventilator support. This type of care can be a welcome alternative because the patient's *level of comfort* takes priority over everything else. "Hospice" is becoming an insurance term as well as a description of a home care philosophy. Depending on your insurance coverage, choosing hospice may be very much like joining an HMO, because in many cases it receives **capitated payments**; this term means that the hospice decides how many home health aide hours to approve, what equipment it *will* and *will not* rent, what doctor(s) will be seen, what treatments it will allow, and more. You must consider what, if anything, you will have to give up, versus what you will gain from this arrangement. Your ALS care team and your local hospice can work together to give you the information necessary for you to arrive at a decision that is right for you.

Your Health Care Team at a Glance

It is important for you to keep track of who is coming to your home, the purpose of the visit, and what person or agency is represented. The following information sheet can be used as a quick reference, and it can help you become more organized in managing your health and care.

Also keep several copies of the following: the medications you are taking, known allergies, and **Advance Directives** (a written document you prepare, stating how you want medical decisions made if you lose the ability to make them yourself).

Health Care Team	*Name/Contact*	*Phone Number*
ALS ASSN. Chapter		
Family Doctor		
Neurologist		
Registered Nurse		
Physical Therapist		
Occupational Therapist		
Speech Therapist		
Dietitian/Nutritionist		
Social Worker		
Mental Health Specialist/Therapist		
Gastroenterologist		
Pulmonary Specialist		
Pharmacy		
Community Agency		
Home Health Care Agency		
Medical Equipment Company		
Ambulance Company		
Hospital		
Insurance Company (Case Manager)		
Family Member/Friends		
Feeding Supplement Vendor		
Respiratory Therapist/ Vendor		
Pastoral Care		

Employment Issues

THE AMERICANS WITH DISABILITIES ACT (ADA)

Years ago, people with ALS were reluctant to disclose information about the diagnosis to their employers, fearing it would have a negative effect on their continued employment. Today, the ADA guarantees equal opportunity for disabled individuals in employment, public accommodations, transportation, state and local government services, and telecommunications. The ADA requires that employers modify jobs, the work environment, or the manner in which jobs are customarily performed to enable qualified individuals to continue employment. Three examples of reasonable accommodations may include:

- making the building and work site accessible (e.g., installing ramps for wheelchairs);
- modifying set work hours to permit disabled persons to drive or take public transportation during non-peak hours;
- acquiring special equipment or devices (e.g., a headset telephone or speaker phone for an employee with hand or arm weakness).

An accommodation is not required if the change would impose an undue hardship on the employer, such as if it were unreasonably costly, intrusive, or would fundamentally change the nature of the position or business. If discrimination is believed to have occurred, a complaint may be registered in writing to the Coordination and Review Section, Civil Rights Division, U.S. Department of Justice, 950 Pennsylvania Avenue, NW, Washington, DC 20530-0001; or by calling 1-800-514-0301 (voice) or 1-800-514-0383 (TDD).

FAMILY AND MEDICAL LEAVE ACT

Under the 1994 Family and Medical Leave Act, employees must be offered at least 12 weeks (within a 12 month period) of unpaid leave to care for an ill family member – a parent, spouse, or child – or when the employee suffers from an incapacitating health condition. The Act applies to all businesses with 50 or more employees (at sites within 75 miles of one another). An employee must have worked at least 1,250 hours in the previous 12 months (about 24 hours a week) to be eligible for leave. The Act mandates that:

- If possible, employees must give 30-days' notice that they are taking a leave.
- Employees (except for 10% of those in highest-paid positions) are entitled to get their previous jobs back, or a job with equal duties, benefits, and pay.
- Employees are entitled to their full health benefits while on leave; however, an employer can demand to be paid back for insurance premiums, if the employee quits the job at the end of the leave.
- Leave can be taken intermittently, *if* the employer and employee both agree on the arrangement.
- An employer may require certification of the health condition by a health care provider.
- An employer can require that vacation or sick days be used at the beginning of the leave.

The Family and Medical Leave Act has enabled many family members to take an extended period of time off from work to attend to their loved ones with ALS, without putting their jobs at risk.

Income Issues

SHORT OR LONG-TERM DISABILITY POLICIES

Having a diagnosis of ALS does not automatically mean you should stop working. You may choose to work as long as you are able. If you find you are no longer able to work because of your physical limitations, you may be eligible for short or long-term disability benefits from your employer. A handful of people have private disability insurance. To activate benefits you will need a doctor to verify your diagnosis and limitations.

SOCIAL SECURITY DISABILITY

To be eligible for **Social Security Disability**, you must be "covered" under the Social Security program. The number of quarters an individual needs to have paid FICA taxes, as an employee or self-employed individual, depends on the type of benefit an individual is applying for and their age when making the initial application. Under the Social Security Disability Program, there is usually a five-month waiting period for disability payment after the established onset of your disability. You cannot apply in advance to offset the waiting period, even if you had physical limitations prior to leaving your job. Disability is awarded when a person is unable to work because of his/her symptoms. You are not considered disabled by the Social Security Administration until the day you stop working. You may apply for your disability benefits at this point. Advocacy efforts led by The ALS Association guarantee that having a diagnosis of ALS is a legal presumption of disability which speeds up the application process.

The amount of your monthly disability benefit is based on your lifetime average earnings covered under Social Security. That information is available to you at any time. Just call Social Security and ask the representative to send you the **Personal Earnings and Benefit Estimate Statement** form; provide the information requested and send it back. Subsequently, the details about your personal Social Security benefits will be sent to you. By requesting this information, you will eliminate any uncertainty and be able to plan your financial future.

When you apply, you will be asked for the following information: the names, addresses, and phone numbers of all the doctors, hospitals, clinics, and institutions that cared or are currently caring for you; the dates of treatment; a summary of where you worked in the past 15 years and the kind of job you did; a copy of your W-2 Form or, if you are self-employed, your federal tax return for the past year. You must include the Social Security number and proof of age for each person applying for payment, including your spouse and children, because certain members of your family may qualify for Social Security benefits under your coverage.

Be sure to submit your medical records along with a list of the medications you are currently taking. In addition, clearly explain the type of work you were doing prior to becoming disabled and why you can no longer do it. All of this information is needed to establish a valid claim.

Eligibility criteria for Social Security Disability may be different for those with private or employer group-sponsored disability plans. The fact that you qualify for disability elsewhere does not mean you will be eligible for Social Security Disability. For more information about disability benefits or to schedule an appointment at your local Social Security office, call the nationwide toll-free number, 1(800) 772-1213. A helpful resource for free information and suggestions regarding Social Security Disability, contact the A.C.C.E.S.S. Program at 1-888-700-7010.

A.C.C.E.S.S. is a free program to help people with ALS (and certain other diseases) to apply for and obtain Social Security Disability and Medicare insurance benefits. Effective July 1, 2001, people with ALS who are under 65 years and who have been approved for Social Security Disability, will automatically become eligible for Medicare benefits after the five-month waiting period. The 24-month waiting period for Medicare eligibility no longer applies to people with ALS.

SUPPLEMENTAL SECURITY INCOME (SSI)

SSI is a needs-based program for those who are disabled (or 65 or older or blind) **and** who have low income. It is a federal program which is supplemented differently in each state. SSI adds to other income you may have in order to raise your earnings to what is considered the subsistence level in your state. Individuals who apply for Social Security Disability are automatically screened for SSI.

In the event that Social Security decides that the applicant is eligible for SSI, you should ask about the possibility of a "**Presumptive Disability Decision**." This action potentially could get SSI benefits for you more quickly. During the interview at the Social Security office or by phone, ask for a presumptive decision under Section G or E, since it is not always considered routinely and your requesting it may expedite your claim. People who get SSI also may be eligible for food stamps and health insurance called **Medicaid** or **Medical Assistance**. For more information about SSI, contact your local welfare office, board of assistance, or Social Security office.

Benefit Issues and Options

EMPLOYER GROUP HEALTH PLAN

If you are still employed, you will want to check with your employer's benefits office to get information about your group health plan. If you stop working because of your disability, you will want to review your COBRA coverage (see next page), and find out how much it will cost you to continue the benefit. It is usually best to say "yes" to COBRA. Some employees may be eligible for a retirement or disability package that enables coverage to continue indefinitely at minimal cost (not to be confused with COBRA).

Larger companies may offer several insurance plans, and each year you may have the option to switch without penalty. During these open-enrollment periods, you may investigate other plans and determine if another policy would offer more comprehensive coverage for the items you need – paying particular attention to DME, home health care, and prescriptions.

The Health Insurance Portability and Accountability Act (HIPAA) of 1996 offers protection for many people with ALS who are still working. This law covers portability and continuity of health care insurance and includes consideration for pre-existing conditions, special enrollment rights and discrimination protection. For more specific information, visit the HIPAA web site at www.dol.gov/dol/topic/health-plans/portability.htm.

CONSOLIDATED OMNIBUS BUDGET RECONCILIATION ACT (COBRA) OF 1985

Depending on the company's size and your state's insurance regulations, you may be able to maintain your group coverage for a time after you leave your job as provided by COBRA (the Consolidated Omnibus Budget Reconciliation Act of 1985). If you worked for a business with 20 or more employees, COBRA entitles you and your dependents to continued coverage for at least 18 months under your former employer's plan.

If you are insured through your spouse's plan at work and your spouse dies, you become divorced or separated, or your spouse becomes eligible for Medicare, COBRA provides coverage up to 36 months. You can lose this benefit if you do not pay the premiums, you become eligible for Medicare, your employer discontinues health insurance for all employees, or you join another plan.

If you are not eligible for COBRA because your former employer has fewer than 20 workers or is a church organization, you still may have some protection under state laws. If your state provides for continuation of benefits, you may be able to stay on your employer's group policy for as little as three months in some states or as long as 18 months in others. Review the COBRA guideline with the representative at your employer's staff benefits office. The A.C.C.E.S.S. program can also advise you about COBRA. For additional information visit the web site at www.dol.gov/dol/topic/health-plans/cobra.htm.

MEDICARE

Medicare is a federal health insurance program for people 65 or older and for **certain disabled persons under 65**. It is run by the Centers for Medicare and Medical Services (CMS). If you are under 65, you may be eligible to receive Medicare benefits. Check with Social Security to see if you have worked long enough to be eligible under these programs. People with ALS who are under 65 may become eligible for Medicare if they are disabled and are approved for Social Security Disability (SSD) benefits. A five-month waiting period for disability payments and Medicare eligibility is required once SSD is approved.

Medicare is made up of Part A – hospital insurance, and Part B – medical insurance. Part A has deductibles and coinsurances and pays for 1) inpatient hospital care; 2) inpatient care in a skilled nursing facility, following a three-day hospital stay; 3) home health care; and 4) hospice. There is a limit on how many days of hospital or skilled nursing care Medicare will pay for in each benefit period; it does not pay for nursing home care that is considered primarily custodial. If you become eligible for Medicare, you cannot waive the Part A coverage. It is not optional. Medicare eligibility may create an impact on other health insurance plans you may have. For guidance regarding how your eligibility for Medicare impacts your current insurance, contact your State Health Insurance Counseling and Assistance Program (SHIP), which provides free insurance counseling to individuals eligible for Medicare. For the SHIP nearest you, call 1-800-677-1116 or go to www.aafp.org./x19934.xml.

Part B is optional, and a monthly premium can be deducted automatically from Social Security payments. An annual deductible must be met before payments begin, and a copayment of 20% is required in most cases. Medicare Part B helps pay for 1) a doctor's services; 2) outpatient hospital care; 3) diagnostic tests; 4) DME; 5) ambulance services (strict criteria for coverage); and 6) many other health services and supplies that are not covered by Medicare Part A.

MEDICARE HMOS

Many Medicare beneficiaries have joined managed care plans – prepaid, coordinated care programs, most of which are HMOs (health maintenance organizations). Managed care plans focus on the relationship between the patient and the primary care physician, whereby the doctor authorizes, arranges for, and coordinates all services. Such plans may offer benefits not covered by Medicare, at little or no additional cost, including preventive care, prescription drugs, dental care, hearing aids, and eyeglasses.

The criteria for joining a Medicare HMO are enrollment in Medicare Part B and continued payment of that premium and residence in the plan's service area. Individuals in hospice care or who have end-stage renal disease are not eligible. If you enroll in a managed care plan, usually you will be required to get all services through that plan. In most cases, care not authorized by the HMO will not be covered through the plan or Medicare. When you join an HMO, be sure to read your membership materials thoroughly to learn your rights and coverage. Also verify that your ALS care team participate in the HMO.

MEDICARE AND OTHER INSURANCE

If you are considering buying a private insurance policy to supplement Medicare, shop carefully. This information is available at your state insurance department website http://www.naic.org/state_web_map.htm

MEDICAID

SSI recipients may be eligible for Medicaid. Medicaid helps pay for doctors, hospital bills, medical equipment, medical supplies, prescriptions, home health care and nursing facility services. You can get more information about Medicaid at your local welfare or board of assistance office.

OTHER FUNDING SOURCES

Civic and religious organizations may offer some options of financial support, coordination of free or volunteer services, or limited financial grants. People with ALS who have ties to these organizations should consider using them.

For additional information contact your ALS Association chapter or call The ALS Association National Office Patient Services Department at 1-800-782-4747.

The Last Word

Help *is* out there. You may need to be persistent and explore various government programs and community resources. Remember, make the following your overall motto:

Ask Questions.
Assume Nothing.
Don't Take "No" for an Answer.

Your advocacy efforts will help you identify community, state, and national resources, while at the same time minimize your out-of-pocket expenses associated with ALS.

The following is a list of the topics covered in the *Living With ALS* manuals:

Manual 1

What's It All About?

This manual provides an overview of ALS, what it is, and how it affects your body. It also provides information on what kinds of resources are available to help you deal with ALS more effectively.

Manual 2

Coping with Change

This manual addresses the psychological, emotional, and social issues that you must face when your life is affected by ALS. It provides information on how to cope with the many lifestyle changes and adjustments that occur when you live with ALS.

Manual 3

Managing Your Symptoms and Treatment

This manual discusses the symptoms that affect you when you have ALS and how to treat them. It also covers the most recent breakthroughs in medications and how these treatments can improve the quality and duration of your life.

Manual 4

Functioning When Your Mobility Is Affected

This manual covers the full range of mobility issues that occur with ALS. It specifically discusses exercises to maximize your mobility, as well as how to adapt your home and activities of daily living to help you function more effectively.

Manual 5

Adjusting to Swallowing and Speaking Difficulties

This manual addresses swallowing difficulties and how to maintain a balanced diet with ALS. It also covers how speech can be affected by ALS and the specific techniques and devices available for improving communication.

Manual 6

Adapting to Breathing Changes

This manual explains how normal breathing is affected by ALS. Specifically, it explains how to determine if you have breathing problems and what options are available to assist you as your breathing capacity changes.

The information contained in this manual can be very valuable to people living and dealing with ALS. Please donate this manual to your local library if you no longer need it.

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Information and Referral: 800/782-4747
www.alsa.org

The ALS Association is the only national not-for-profit voluntary health organization dedicated solely to the fight against amyotrophic lateral sclerosis (often called Lou Gehrig's Disease) through research, patient and community services, advocacy, professional education and public awareness.

Member of the National Health Council and Community Health Charities (CHC)