

**LES
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ALS
FOUNDATION**

Patient and Family Resource Guide to ALS

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Forward

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The Les Turner ALS Foundation is here for you.

Contact the Director of Patient Services to learn how
the Foundation has provided free support services
for families like yours since 1977.

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Welcome!

Welcome to the Les Turner ALS Foundation's Resource Guide.

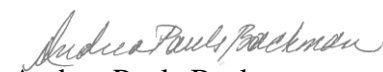
We are the family of experts you never expected to need. We understand it can be overwhelming to hear that you or a family member has been diagnosed with Amyotrophic Lateral Sclerosis (ALS). ALS is a difficult and complex disease. Know that we are here to help guide you through a myriad of confusing medical jargon and a multitude of online resources at a time when you most need clear direction and understanding.

While there is no cure yet for ALS, scientists and clinicians have made much headway in not only uncovering causes of ALS, but also in offering medical interventions to improve the quality of life and possibly slow the progression of the disease. Our goal, as it has been for over 40 years, is to be **with you every step of the journey**, with support provided by our team of highly experienced nurses, social workers and other ALS health professionals who work seamlessly with your doctors and clinical team at the **Lois Insolia ALS Clinic at the Les Turner ALS Research and Patient Center at Northwestern Medicine**. Over the decades, the Les Turner ALS Foundation team of Patient and Family Advocates has developed the most comprehensive ALS support program in the country.

This updated resource guide is based on that experience. It offers sections on understanding the disease, coping with the aftermath of diagnosis and countless practical tips to improve your everyday life with ALS. You will find links to outside resources for clinical trials, assistance with speech issues and explanations of insurance and legal issues to protect you and your family, all of which are based on our many years of experience with people with ALS and their families. This resource guide is designed to provide the information you need when you need it. It is intended to be used alongside visits with your neurologist and pulmonologist and the multidisciplinary team at the Lois Insolia ALS Clinic as well support from the Foundation's Patient and Family Advocates, who can help you determine what works best for you. Even if you live outside Chicagoland and are not affiliated with Lois Insolia ALS Clinic or the Foundation, this resource guide is a tremendous wealth of knowledge.

We hope this revised version of our resource guide will serve as a helpful reference for you and your loved ones throughout this journey. When you have questions or concerns, we are only a phone call or email away.

Together towards a cure,


Andrea Pauls Backman
Chief Executive Officer



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Patient and Family Resource Guide to ALS

Section 1 ALS Overview

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ALS Overview

Amyotrophic lateral sclerosis (ALS), formerly known as Lou Gehrig's Disease, is a progressive, degenerative disease of the nervous system. It is one of a group of diseases, called motor neuron diseases (MND), in which specialized nerve cells that control movement of the voluntary muscles gradually cease functioning and die. These nerve cells, called motor neurons, carry impulses from the brain to the brainstem and the spinal cord. The impulses are then carried to the muscles. The muscles respond to these messages by coordinated relaxation or contraction corresponding to willed movement. In ALS and other motor neuron diseases, motor neurons gradually deteriorate. Because the nerve cells that stimulate them have died, the muscle tissues waste away. This results in progressive muscle weakness, atrophy, and often spasticity, or excess muscle tone. Only the motor neurons are affected. Other nerve cells, such as sensory neurons that bring information from sense organs to the brain, remain healthy.

Symptoms of ALS

The early symptoms of ALS vary in different individuals, depending on which part of the central nervous system (the brain and spinal cord) the disease affects first. Upper motor neurons travel from the surface of the brain (the cortex) down to the brainstem and/or to the spinal cord. Within the brainstem or spinal cord, the upper motor neuron connects with a lower motor neuron. The lower motor neuron leaves the brainstem or spinal cord and travels to the muscle that it controls. ALS may involve damage to either upper or lower motor neurons, but it usually affects both kinds of neurons.

The symptoms and clinical features of the disease depend on the location of the affected neurons. Speech and swallowing impairments are called bulbar symptoms. They indicate that neurons in the brainstem are affected. Weakness of the respiratory muscles, muscle weakness, and loss of mobility in the arms and legs are called somatic symptoms. They indicate spinal cord involvement. In classical ALS, a mixture of upper and lower motor neurons is involved, with both bulbar and somatic symptoms. Early symptoms of ALS can include weakness, clumsiness, fatigue, stiffness, muscle twitching, cramping, and difficulty in chewing, swallowing, speaking, or breathing.

Lower Motor Neuron Symptoms

Weakness and muscle wasting are common when lower motor neuron involvement predominates. The patient or physician usually notices fasciculation, or muscle twitching. Fasciculation is a sign of muscle irritability, as the normal action of the lower motor neuron on the muscle is impaired. The sole involvement of lower motor neurons can be seen in a form of ALS called progressive muscular atrophy. Fasciculation is described as "benign" if there is no muscle weakness, atrophy, or impairment of motor function. Fasciculation is described as "pathologic" when it occurs in ALS with other symptoms.

Upper Motor Neuron Symptoms

Spasticity, or stiffness, in the lower limbs, face or jaw indicates upper motor neuron involvement. Spasticity in the legs often produces severe walking difficulties. An individual may complain of heaviness, fatigue, stiffness, or lack of coordination of any affected limb. Reflexes are very brisk, or exaggerated. Outbursts of laughter or crying with minimal provocation can occur. This is called emotional lability and is referred to as a pseudo-bulbar affect. Both brisk reflexes and emotional lability involve the inability to inhibit reflexes.

Primary lateral sclerosis (PLS) characteristically involves progressive spasticity, difficulty in walking, and pseudo-bulbar affect related to upper motor neuron involvement of bulbar and somatic muscles. In progressive bulbar palsy, an ALS variant, speech, swallowing, and behavioral symptoms predominate, related to upper and lower motor neuron involvement.

Clinical Course of ALS

Weakness of the bulbar and somatic muscles produces a decline in speech, swallowing, and limb strength and function. The person with ALS generally remains alert throughout the course of the illness and retains normal sensation, vision, and bowel function. Bladder function is impaired in a small percentage of affected individuals. Generally, ALS is not a physically painful condition. Discomfort can result from immobility and joint contractures, a shortening of muscles resulting in deformity. The problems are related to advancing muscle weakness and the inability to change positions easily. Proper positioning, exercise, physiotherapy and medications can help keep patients comfortable.

While most people with ALS do not have significant loss of intellectual function, some may have subtle changes in mood, behavior or personality. In a small minority of patients, more significant changes in behavior and judgment suggest a form of dementia called frontal/temporal dementia (FTD).

People with ALS with significant bulbar involvement may require help to improve communication or ensure safe and adequate nutrition. A gastrostomy (feeding) tube may be suggested in the following situations: recurrent pneumonia, high risk of aspiration (inhaling food or liquids into the lungs), inadequate nutrition, rapid weight loss, or extended feeding time. A wide range of devices and techniques can address problems with communication. Ultimately, ALS may result in respiratory decline, requiring consideration of respiratory support, including non-invasive ventilation such as a VPAP, or a tracheostomy and a ventilator, which are invasive procedures.

Each person with ALS is unique in regard to the rate and characteristics of the progression of the disease. The Lois Insolia ALS Clinic at the Les Turner ALS Research and Patient Care Center at Northwestern Medicine provides a multidisciplinary approach to patient care, including an individualized treatment plan guided by each patient's personal preferences and wishes. Although the clinical progression can vary greatly, statistically 50 percent of people with ALS will succumb to the illness within five years of the onset of symptoms.

Diagnosis of ALS

An individual who might have ALS is usually evaluated by a neurologist, a doctor who specializes in disorders of the nervous system. The diagnosis of ALS is "clinical" in nature. That is, it is determined by what the doctor hears and sees. A series of tests are usually ordered to exclude conditions that can mimic ALS. Testing for conditions that might mimic ALS is important because many of these conditions are treatable. Once the diagnosis of ALS is confirmed, the neurologist and the Lois Insolia ALS Clinic staff provide information and resources to the patient and family, and an individualized plan of care for the patient is established.

Three kinds of diagnostic tests are usually performed: imaging studies, electro diagnostic studies, and fluid and tissue analysis.

Imaging studies, such as X rays, CT (Computerized Tomography) and/or MRI (Magnetic Resonance Imaging) scans, and myelography, are performed to exclude any structural abnormality of the nervous system.

The EMG (electromyogram), which records the electrical activity of muscle, and the NCV (nerve conduction velocities), which quantifies a nerve's ability to transmit electrical impulses, are electro diagnostic studies. They evaluate the integrity of muscles and peripheral nerves.

Fluid analysis includes the evaluation of blood, urine, and occasionally cerebrospinal fluid. These fluids are screened for potential metabolic, endocrine, immunologic, infectious, and toxic abnormalities. Tissue analyses, including muscle and/or nerve biopsy, are occasionally indicated to establish the diagnosis or to exclude other neuromuscular conditions.

In addition, approximately 2–3percent of all ALS cases are due to mutations in the gene for Cu, Zn, Superoxide dismutase (SOD1). Genetic testing for SOD1 and other genes are available at the Lois Insolia ALS Clinic. In addition to the SOD1 gene mutation, approximately 30 other genes have been identified, the most common of which is C9orf72.

In general, patients with ALS will not have significant abnormalities on imaging studies and fluid analysis, but will show characteristic results on electro diagnostic studies and muscle biopsy.

Although the diagnostic work-up is extensive, the tests are generally not risky or painful. Once the diagnosis of ALS has been established, the short and long-term care can be planned. Professionals who care for people with ALS believe a multidisciplinary approach is most appropriate. A therapeutic team of professionals, trained in a variety of disciplines, ensures optimal patient and family care. This team includes practitioners in neurology, nursing, social work, speech, nutrition, genetic counseling, and occupational and physical therapies. Such care has been provided by the Lois Insolia ALS Clinic since 1986, a nationally-recognized multidisciplinary ALS clinic.

Causes of ALS

Each year, two new ALS cases per 100,000 people are diagnosed. At any given time, approximately 20-30,000 people afflicted with ALS are living in the United States. Most affected persons are between 50 and 60 years of age, although the disease can strike at any age in adulthood. Men are affected slightly more frequently than women. Some studies have identified areas that at certain times have had greater than expected numbers of cases. This has occurred in the past in the western Pacific islands and in parts of Japan and Australia. Other areas in the continental United States have been reported but have not stood up to careful epidemiological investigations.

Familial ALS accounts for approximately 10 percent of all ALS cases. Researchers investigate the genetic basis of ALS by studying families in which a number of members have the disease. Such research has identified several genes that cause ALS. One such gene is on chromosome 21 (the copper-zinc superoxide dismutase, or SOD1, gene). It accounts for about 20 percent of the familial ALS cases, or 3 percent of all ALS cases. Development of a mouse model of ALS, which has mutant SOD1 genes in its chromosomes, has enhanced our understanding of how ALS develops. The mouse model also serves as an important “testing ground” for newly developed therapies. Another gene, ALSIN, causes ALS and PLS in children. Molecular genetic investigation is likely to uncover additional genes and their products that will account for the majority of familial ALS cases. The identification of additional genes and gene products will significantly advance our understanding of sporadic (non-familial) ALS. In addition, the search for a genetic predisposition in sporadic ALS is being investigated.

For several years, scientists have investigated naturally-occurring toxic chemicals as a possible cause of ALS. The presence of abnormal amounts of glutamate, a chemical that occurs naturally in the brain, has been observed in ALS patients. Glutamate helps carry messages from one nerve cell to another. Normally, after glutamate does its work, it is removed by another chemical, called a transporter. If the transporter doesn't properly remove the glutamate, excess glutamate might remain in the nervous system. This is postulated to over-stimulate motor neurons and result in their death. The glutamate hypothesis is one basis for riluzole therapy. Riluzole is a chemical that, among other actions, reduces the amount of glutamate released into the nervous system, thus reducing damage to the motor neurons. The FDA approved Rilutek, which is the brand name for riluzole, in 1995.

Other Theories of Causation

Viruses and abnormalities of the immune system are other possible causes of ALS. A virus or foreign substance may trigger the immune system, which normally fights infection. The immune system may then attack the body's own nervous system (an autoimmune process). Immune system abnormalities and autoimmune diseases may be more frequent in those with ALS and their relatives. However, specific and consistent abnormalities are difficult to identify and related trials have not shown success in the treatment of ALS.

A large number of people with ALS may have abnormal antibodies. However, it is not clear which of the antibodies produce disease and which are associated with the disease process but do

not actually cause disease. As noted above, a viral "triggering" agent of motor neuron disease has been suggested. This hypothesis has been generated because certain viruses such as the polio virus or the West Nile virus can cause an acute infection of the motor neurons. This has led to the idea that ALS might be a persistent viral infection, resulting in progressive motor disability. However, no virus has been seen or otherwise identified in autopsy material from ALS patients. It is true that many polio survivors begin to deteriorate years after the acute phase of their illness. However, the cause of this progressive, post-polio muscular atrophy is unclear. Post-polio patients do not appear to have greater risk for ALS.

Neurotrophic factors, or chemicals that affect the growth of neurons, are found throughout the nervous system. One theory of ALS suggests that a deficiency of growth factors results in reduced survival and eventual degeneration of motor neurons. Nerve growth factor (NGF), ciliary neurotrophic factor (CNTF), brain-derived neurotrophic factor (BDNF), glial-derived neurotrophic factor (GDNF), and insulin-like growth factor (IGF-1) all promote motor neuron survival in tissue culture and in animal models of motor neuron disease. However, clinical trials using neurotrophic factors in ALS have had generally negative results. Overall, the neurotrophic factors do not appear to be effective in treating ALS.

Many other possible causes and associations have been suggested, including environmental toxins, premature aging, endocrine factors, heavy metal poisoning and various nerve abnormalities. No single cause has yet explained the variety of motor neuron disorders or ALS. Many interacting factors are probably involved in the clinical and pathological abnormalities in ALS.

More recently the subcellular organelle called a mitochondrion has been implicated in motor nerve cell death in some experiments. Mitochondria are the energy factories of the motor neuron. Not only could there be energy failure for some reason but in addition there are at least two potential toxic things that could come out of mitochondria. All mitochondria normally produce some free radicals sometimes called reactive oxygen species. In excess free radicals can kill cells. Nature has evolved several ways to handle free radicals including the SOD enzyme. It is possible these detoxifying systems could be overwhelmed. Another possible mitochondrial explanation goes by the name of apoptosis. This is a cell suicide pathway that is triggered by enzymes called caspases. One of these is produced by the mitochondria.

Another area of interest is the way the disease spreads. It seems to stick with the first limb for a while, progress there and then usually but not always spread to another limb. This has led some investigators to think that there is a problem with the supporting cells in the nervous system called glia or astrocytes (because they are star shaped). The thought there is that it may be the neighborhood that matters as much as the neuron itself.

Disclaimer: All care has been taken in preparing this document. This information is of a general nature and should be used as a guide only. Always consult your health care team before starting any treatments.

Patient and Family Resource Guide to ALS

Section 2 Medication and Drug Research

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ALS Medications and Drug Research

Medications

Currently, there are no drugs available to cure or reverse ALS, due to the complexity of the disease. However, there are several interventions and drug therapies that treat symptoms and can make living with ALS easier.

The three types of medications most commonly used in the treatment of ALS are:

- prescription drugs developed specifically to treat ALS,
- prescription drugs used to relieve the symptoms of ALS,
- over-the-counter medications.

A physician must be involved in the decision to take prescription drugs, but is not required for over-the-counter medications. However, over-the-counter medications can interact with each other and with prescription drugs. Therefore, before using over-the-counter medications, people with ALS (PALS) are advised to discuss the medications being considered and the dosage with their neurologist. The neurologist should also be aware of all the prescription drugs someone is taking.

Prescription Drugs Developed Specifically to Treat ALS

At present, there are two drugs approved by the U.S. Food and Drug Administration (FDA): Rilutek (riluzole) and Radicava (edaravone).

Rilutek: according to the clinical drug trial that led to approval of Rilutek by the FDA in 1995, taking 100 milligrams of Rilutek each day is modestly effective in prolonging survival for patients with ALS. However, no increase in muscle strength or in bulbar function was noted. Rilutek is available in generic form as riluzole.

Radicava: the clinical trial using Radicava was conducted in Japan and approved by the FDA in May 2017. The data showed a 33 percent slowing of loss of function in participants as rated on the ALS Functional Rating Scale; survival was not studied. Radicava is an intravenous drug - it is given daily over 60 minutes for a 60 mg dose (two 30 mg IV bags) for 14 consecutive days in month one, then 14 days off. Monthly follow up treatments are given 10 days over a two weeks period; then two weeks off and repeated each month thereafter. Length of treatment should be discussed with your clinician.

Prescription Drugs Used to Relieve Symptoms of ALS

Several prescription drugs are used to relieve symptoms of ALS. In some people, prescription drugs are helpful in controlling such symptoms as excess saliva, muscle stiffness, emotional lability (excessive response to a laughing or crying situation), insomnia, muscle cramps,

constipation, gastric reflux, depression, and pain. The following is a partial list of prescription drugs used to help relieve symptoms of ALS:

- Lioresal (baclofen) or Valium (diazepam) may be prescribed for muscle stiffness
- Elavil (amitriptyline) may be prescribed for excess saliva and for emotional lability.
- Nuedexta, approved by the FDA in 2011, is used to treat pseudobulbar affect, which is characterized by inappropriate laughing or crying.

All of the medications have side effects, which must be taken into account when deciding whether to use them.

Over-the-Counter Medications

Chemicals called free radicals have been shown to cause damage to cells throughout the body. Scientists think that the free radicals may be one cause of damage of the motor neurons of people with ALS. Certain other chemicals, called antioxidants, may help to counteract the cell damage caused by free radicals. A number of nutrients, including certain vitamins, minerals, and herbs are antioxidants. The most commonly used antioxidants are vitamin E, vitamin C and beta-carotene, which is the precursor of vitamin A. Coenzyme Q-10 and melatonin are also antioxidants. Be aware that taking excessive amounts of these vitamins may be counterproductive. As with any over the counter medication, ask the neurologist what dosage you should take and how frequently the vitamins should be taken.

There are few hard and fast rules for determining which over-the-counter medications to take and at what dosage. This decision depends upon a person's onset of disease, current symptoms, age, weight, and other health supplements. Many outrageous, unproven claims are made for nutritional supplements. This is done by people who take advantage of people who may be vulnerable and willing to try almost anything. Their methods can border on quackery. It is best to discuss these issues with the neurologist before taking such medications.

Another good resource for claims of ALS cure or treatments is ALS Untangled at <http://www.alsuntangled.com>

Clinical Drug and Therapy Trials

Everyone with ALS seek a cure for their disease. Short of a cure, an effective treatment to halt the progression of the disease would be acceptable. Pharmaceutical companies spend millions of dollars conducting clinical drug trials trying to find effective treatments. The Food and Drug Administration (FDA) approves only drugs that have been found to be safe, well-tolerated and significantly effective. The process by which drugs are approved in the US is outlined below. Drugs are tested by a series of clinical trials, also called drug studies.

How Can I Find Out About Drug Studies?

The best source of information about drug studies for which you might be qualified is your neurologist. Major ALS drug and research studies are regularly conducted at the **Lois Insofia ALS Clinic at the Les Turner ALS Research and Patient Center at Northwestern Medicine**. You may or may not qualify for any or all of the studies, but your doctor can assist in helping

you evaluate them. Other sources of applicable ALS drug studies are listed below, especially the National Institutes of Health (NIH) Clinical Trials.gov website, the Northeast ALS Consortium (NEALS) and the National ALS Registry conducted by the Centers for Disease Control (CDC). People with ALS who have enrolled in the National ALS Registry are eligible to receive notifications of applicable clinical trials from researchers or pharmaceutical companies.

Resources for Current Drug Trial Information

www.clinicaltrials.gov

<https://www.neals.org/lesturnerals.org>

<https://wwwn.cdc.gov/als/ALSResearchNotificationClinicalTrialsStudies.aspx>

Who Can Enroll in a Drug Study?

Strict guidelines for participation in clinical drug studies are an important factor in achieving acceptable results. To be eligible for an ALS drug study, a person with ALS must have a diagnosis of ALS made by a qualified neurologist who follows internationally accepted criteria. These criteria eliminate the chance of misdiagnosis.

Other considerations might also be required for clinical trial participation, such as:

- **Age**—a particular age range might be required for inclusion
- **Gender**—the study might include only males, only females, or both males and females
- **Past medical history**—certain health conditions might be disqualifying, such as other muscle and nerve diseases or heart, lung, endocrine or kidney diseases
- **Present condition**—might affect the study results or a person with ALS's ability to follow the study procedures. For example, alcoholism, mental disorder, or hypertension might be disqualifying factors
- **Current medications**—might interact with the study drug, cause harm or confuse the study results

The ability to travel to the testing center for regularly scheduled visits is also an important requirement.

Factors that might confuse the analysis of the information collected would also require exclusion from a clinical trial. These factors might not be related to the study drug, but could distort information about its effectiveness. For example, persons with ALS using permanent ventilation usually are not eligible for ALS drug studies because judging the drug's effectiveness is based on survival, improved or stabilized breathing capacity, muscle strength, functional abilities and quality of life.

Why Do ALS Drug Studies Use Placebos?

Placebo-controlled drug studies help prove the effectiveness of the study drug. Persons enrolled in any drug study may believe that they feel better because they are actively involved in the drug study process, rather than because of the effectiveness of the drug. Comparison with a group taking a placebo, or inactive substance, can determine how much better the study drug is working over and above the “placebo effect.” FDA approval requires that there be no doubt that the study drug is effective. The placebo group in the study is the standard against which the drug groups are measured.

Safety is just as important as efficacy. Side effects related to a given treatment cannot be identified without using a control group that does not get the treatment under study. For example, if during a clinical trial the investigator asks about nausea or headaches or muscle cramps, these conditions could be falsely attributed to the drug being studied since many people will experience these over the course of a year. Without a placebo group, it cannot be determined if these conditions are treatment related.

Why Should I Participate in a Drug Study?

All persons enrolled in drug studies receive increased attention to their condition and have the best current medical care. Study participants are not only actively trying to help themselves; they also are participating for the benefit of current and future persons with ALS or various other medical conditions. Even studies that do not have positive results add important information about the underlying process of the disease.

What Happens in the Drug Approval Process?

A pharmaceutical company targets a need for treatment of a specific condition. Researchers develop a drug that they think will improve, slow the progress of, or cure the disease. A written protocol describing the methods to be used in the study is submitted to the FDA for approval. Once it has been approved, the company selects medical centers with specific patient populations to participate in the research. The protocol is submitted to the each university medical school’s Institutional Review Board (IRB) in the Office of Protection of Research Subjects. The IRB ensures that patient safety is properly adhered to.

Another possibility is that investigators will suspect that a treatment already approved for other diseases might work for ALS. They may apply to the NIH or non-governmental organizations for financial support and safety oversight to conduct experiments.

Pre-Clinical Studies are done in animals. Investigation of safety and effectiveness begins here.

Clinical Trial Phases

Phase I: Phase I studies are done with normal volunteers to evaluate whether the drug is safe for human consumption. Once the drug is known to be safe, the correct dose must be determined.

Phase II: Phase II studies measure safety, tolerability and “dose ranging.” Often people with ALS take doses of study drugs for only a short period of time to determine how much can be

taken before the amount is toxic. Then, persons with the disease are enrolled in the study to determine the correct doses, as well as to get some measures of the drug's effect. In Phase II studies, some subjects will get no drug, others will get a small amount of drug, while still others may get a larger dose. The goal is to find the one or two most effective dosages to use in a larger Phase III study. The safety of all doses is carefully monitored; large doses are not necessarily better. Phase II studies also allow researchers to select the tests or measurements that have the best chance of recognizing aspects of success in the Phase III study. Side effects are considered at all times. An accurate record of all side effects, as well as of other medications a person may be taking, is needed to determine the safety and effectiveness of the study drug.

Phase III: A Phase III study is a large trial, usually done at several medical centers. This study determines the effect of the drug on a large number of individuals over a longer period of time. ALS studies are typically done for 12 to 18 months to determine long-term safety and effects. Phase III studies include the greatest number of patients. The number depends on the length of the study and what size effect or “statistical significance” is expected. The selection of doses and tests is based on the Phase II study results. The duration of the study or length of time each subject takes the drug or placebo is no trivial matter. Some studies require 12-month participation, while others need 18 months. It takes a certain amount of time to enroll several hundred participants and answer about the effect of the drug can be available until all subjects have completed their 12 or 18 months. If many people will participate in a 12-month study, it might take more than 6 months to enroll everyone. Fewer subjects could be enrolled in an 18-month study in a shorter period of time and produce the same “significance” or effect. A prolonged enrollment period also limits the number of drugs that can be tested at many centers because the number of persons with ALS who meet all the study criteria is limited.

Compassionate Use/Open Label

Subjects who have participated in a Phase II or Phase III study are given the opportunity to take the study drug after they have completed their 12 or 18 months. People with ALS who were taking the “active” study drug may continue to take it. The control or placebo group can begin taking the study drug. These patients can continue to take the study drug while other subjects complete 12 or 18 months and while the study data is being analyzed and presented to the FDA. The patients may continue to receive the drug for another year or more while data is analyzed and a request for marketing is completed. This is often an incentive for people to participate in a Phase III study even if there is a high chance they will be receiving a placebo.

Expanded Access

When Phase III studies are complete, the pharmaceutical company may make a promising drug available, while awaiting FDA approval, to patients who did not participate in the studies. Some people with ALS who have not participated in the studies ask if they can take a drug before completion of a Phase III study. The answer is no for two reasons. Drug studies investigate the safety and efficacy of drugs. While the drugs are being investigated, there are no clear-cut efficacies or safety profiles. If a patient who has not been screened for medications or other medical conditions takes a drug and becomes ill, researchers will not know if it was the drug or other factors that caused the effect. The patient is at risk for serious side effects. Also, if a study

drug is made available before the study is completed, some participants, especially those who feel they are in the placebo group, might drop out and get on the drug being studied. Unfortunately, this could jeopardize the study conclusions and make it impossible to obtain FDA approval, which impacts the entire ALS community. Drug companies cannot provide a drug forever at no cost, and they cannot charge for it without FDA approval for marketing.

Will I Ever Know What I Was Taking?

The pharmaceutical company may decide to inform participants what dose of the drug or placebo they were taking. This information can only be released after all participants have completed the course of the trial, all data has been collected from all centers, and all the information has been verified. This is called “locking the database.” After this occurs, there is no risk that information might be changed or that researchers recording the data could be influenced by knowing whether a certain subject was taking the drug.

Stem Cell Treatment

As of this writing, there have been a small number of international and North American stem cell trials conducted in individuals with ALS. These studies are on-going and definitely showing some progress, but more work has to be done. Currently treatment strategies could be grouped along several methods. One method aims to replace damaged glia at the cellular level and hopefully halt the disease. Another method could possibly replace damaged motor neurons and repair damage that has already occurred. It might also be possible to use stem cells to coax the axons of damaged upper motor neurons to remake their connections with the lower motor neurons and increase coordination. While these avenues of research look hopeful, more work needs to be done in this very complex area of potential ALS treatment.

Laboratory-Based Research

Most research that aims to find effective treatments for ALS starts in the laboratory. This kind of research is known as basic research, or bench research.

Cultures

Some experiments take place in cultures, or small flat dishes that contain a substance in which different types of cells can grow outside the body. Various chemicals or drugs can be added to the dishes to see if any changes occur in the growth process or if there are any positive or negative effects on the cells. The information gained from this type of research is published for other scientists, who may take it into consideration as they do future investigations.

Gene Studies

The DNA in cells is usually studied by examining the genes in white blood cells found in blood taken from people with ALS and their relatives. The DNA from the cells is spread on a special gel sheet, so that the genes in different samples can be compared. Similarities and differences among the samples help trace family traits. Matching the characteristics of the samples against the family history of disease helps determine whether the gene contributes to the disease. The

family history can show a disease occurring in several ways. It may be sporadic, with only one individual in a family having the disease. It may be familial, occurring in more than one individual in the family. It may be genetic, indicating that a specific gene is passed from one family member to another.

Scientists need to study large numbers of families to learn about a specific gene and its mutations, or changes. Information from a single sample does not give a definite result, the way a test for blood sugar or cholesterol does. Only after a gene has been identified as one that contributes to a disease process can a specific test be developed that gives a “lab result.” However, even a definite result means little until scientists understand how the gene works. Each gene causes body cells to produce one or more proteins, complex chemicals that cause certain actions and reactions in the body. No test result is meaningful until all of these actions and reactions are known and understood for each gene.

Since the co-discovery of the first gene for ALS, SOD-1, by Teepu Siddique, MD, at the Les Turner ALS Research and Patient Center at Northwestern Medicine in 1993, over 30 genes for ALS have now been identified and the chromosomal addresses are now known. Having one of these genes does not predict whether a person will actually develop the disease, at what age it might begin, or what course it might take.

Animal Research

Using animals in research helps scientists understand the normal mechanisms and pathological processes in the body and enables them to test possible treatments. An investigator may test a certain substance in a laboratory animal to find out if a living animal can tolerate the substance and to observe any changes in the animal’s alertness, muscle strength, and coordination. Only animals that show the same disease symptoms as humans do are useful for testing these substances. By altering the DNA of mice and other animals, scientists can produce animal models that are useful for testing substances that might successfully treat diseases in humans. Animals with altered DNA are called transgenic animals.

The transgenic SOD-1 mouse was developed to show the same disease symptoms as people with ALS. Other transgenic mice have been developed for other conditions. Many important clinical research studies have been developed based on information gained from these animal studies. All animal projects done at the Les Turner ALS Research and Patient Center at Northwestern Medicine are reviewed by the Northwestern University Animal Care Committee. The Animal Care Committee of the IRB oversees animal research.

Resources for FDA Drug Approval Process

<https://www.fda.gov/Drugs/DevelopmentApprovalProcess>

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Patient and Family Resource Guide to ALS

Section 3 Coping with ALS

3

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Coping with ALS

Managing the emotional stress of ALS for patients and caregivers can be an incredibly difficult task for both patients and family members. The unpredictable progression of the patient's physical deterioration contributes to a myriad of feelings that can be very overwhelming.

Depending on the physical challenges, a patient may experience the loss of the ability to:

- Walk, which reduces mobility and the sense of independence
- Use of upper extremities, making dressing, bathing and toileting difficult, which can contribute to a loss of dignity, independence and self-image.
- Speak, resulting in communication and human interaction being more difficult.
- Eat, leading to the loss of independence and opportunities for social interaction.

Trying to identify and name the feelings associated with these issues, such as helplessness, fear, loneliness or anger may help a person to process those feelings. Naming or labeling helps a person with ALS or family member to feel less out of control. It may also help to improve communication between the patient and loved ones.

People with ALS have emotional and physical issues with which to cope. Sometimes when a person cannot move freely, muscles become sore and painful from remaining in the same position. People also have emotional responses to the changes they experience in their lives as a result of physical challenges. For example, some individuals can feel worthless when they can no longer manage responsibilities at home, such as mowing the lawn, taking out the garbage, cooking, or doing the laundry. Others experience intense feelings when they are no longer able to work outside the home or to take care of young children.

The family also has emotional and physical issues to deal with, including helplessness and frustration. Sometimes people with ALS and family members are experiencing different emotions and have different needs. Emotions change from day-to-day; some days they even change from hour to hour. Depression and/or anxiety are very common when trying to cope with ALS. The Les Turner ALS Foundation social workers are available to people with ALS and their family members for counseling in the home and they can help plan strategies to manage the day-to-day challenges brought about by ALS.

It is common to feel overwhelmed when the home environment becomes difficult to navigate. It is important to recognize this, as it is often the cause of frustration, anger and emotional angst. A home assessment by one of our ALS Patient and Family advocates can assist families in maximizing home accessibility and identify when upcoming changes may be needed. The individual's physical changes may also cause a change in roles within the extended family and circle of friends. Those who are used to being "helpers" or "givers" can find it especially difficult to accept the fact that they now need help themselves. This is the time to accept help, and to even ask for what is needed. Often family and friends want to do something, but they

don't know exactly how to help. There are many websites that allow an ALS family to identify their needs and request help. The following are some recommended websites:

- www.caringbridge.org
- www.takethemameal.com
- www.mealtrain.com
- www.lotsahelpinghands.com

Tips for Coping for Persons with ALS

- Break tasks into smaller pieces and do a little bit at a time
- Change your expectations of yourself; understand that you cannot do all that you were doing before AND manage this disease
- Talk to your friends and share the emotional burden
- Ask for help
- Accept help

Tips for Coping for Caregivers

- Don't ignore your own body, especially if you have medications to take and doctor's appointments to keep
- Arrange to get out of the house on a regular basis, even if just for a 15-minute walk.
- Although you may feel that it is much more important to focus on the person with ALS, you cannot take care of him/her if you become ill
- Talk to your friends and share the emotional burden
- Ask for help
- Accept help

Resources for Coping:

- **Counseling**
A Licensed Clinical Social Worker from the Les Turner ALS Foundation can provide counseling for the person with ALS and/or family members, including children in your own home. These visits are scheduled by appointment and are available at *no cost* to the patient and family.
- **General Caregiver information**
www.caregiver.org provides information from the Family Caregiver Alliance
www.nfcacares.org provides information from the National Family Caregivers' Association
- **Support groups**
Support groups are led by Les Turner ALS Foundation professionals. There are in-person monthly meetings in Barrington, Skokie, Wheaton, Chicago as well as virtual

support groups. Please check the Foundation website for current meeting dates (www.lesturnerals.org) or call 847-679-3311.

What is a support group?

A support group is an opportunity for persons with ALS and caregivers to give and receive emotional support and to share information and practical tips. This may include sharing ideas for how to improve function, receiving drug trial information or learning the latest in technological advances. Each group is led by two professional facilitators, which allows participants to divide into two groups: a PALS group and a caregiver group. While they are separated, members of each group have the opportunity to speak freely without worrying about hurting a loved one's feelings. Occasionally a guest speaker attends a meeting. All information and support provided is geared to help cope with the disease in order to maintain as high a quality of life as possible.

Why participate in an ALS support group?

Support groups are a resource to help patients and family members feel a sense of camaraderie with others who are living with ALS. No one can understand the feelings involved better than another patient or caregiver who is also struggling to live with ALS. At support group meetings, some things just do not have to be explained, as they often do in other social situations. The group provides a kind of emotional respite, with understanding and compassion.

Can I come to a group if I am afraid to share my feelings?

Some participants may decide to simply listen, and that is okay. However, most group members soon feel that the support group is a safe place to share feelings. It provides an opportunity to learn how other patients and families cope with similar problems. Most people find that talking with others whose lives have been altered by ALS is helpful.

Can I ask questions about practical matters such as drug trials or equipment?

You can ask about absolutely anything at a support group meeting. The meeting's agenda is to meet the needs of the patients and family members who attend the group.

Coping with the Stress of ALS in the Family

The issue of how children cope when ALS is in the family is extremely complicated for several reasons. The physiological capability of the brain is quite different at various ages. These differences are often referred to as the developmental stage of a child. The developmental stage tells us what we may presume a child has the capability to process and understand. However, within those stages there are always individual variations. Regardless of the child's age or developmental level, the most important thing you can tell her/him is the truth.

When someone in the family has ALS, there is a certain amount of pain from which children cannot be shielded. Therefore, it is most important that children know they can still go to the most trusted grown-ups in their lives to obtain information as they feel ready to ask questions. If children do not ask questions, it is equally as important for those grown-ups to regularly let them know that they are welcome to talk about any concerns at any time. It is a good idea to consult

with one of the Foundation's social workers regarding the extremely important task of supporting the children in the family at various points throughout the disease process.

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Patient and Family Resource Guide to ALS

Section 4 Resources for Daily Living

4

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Resources for Daily Living

Who can help you?

The Lois Insolia ALS Clinic at the Les Turner ALS Research and Patient Center at Northwestern Medicine utilizes a multi-disciplinary team approach where you will have access to an occupational therapist, orthotist, and wheelchair specialist; referrals may be made to meet with a physical therapist. This section will introduce what they do and how they can help you.

Occupational Therapist

An occupational therapist evaluates how a person with ALS performs daily tasks, including personal care, mobility, recreation, and school and work activities. The assessment is done through interviews and evaluation of function in the ALS Clinic or at home. The therapist recommends needed assistive devices such as hand-wrist supports or braces; home modifications; durable medical equipment such as wheelchairs, hospital beds and bath chairs; and home or outpatient therapies. The therapist can train patients and caregivers to use assistive devices and proper body mechanics, can give instruction in ways to conserve energy, and can provide written guidelines for exercises and range of motion activities. Usually, occupational and physical therapists work together to assess body mechanics, exercise programs and wheelchair needs. You need written prescriptions from your physician for evaluation and treatment from therapists. Check with your insurance company or case manager for information about coverage. Check with your Lois Insolia ALS Clinic therapist for information on Medicare coverage.

Physical Therapist

A physical therapist evaluates how a person with ALS manages general functional mobility. This includes assessing a person's neck, trunk, arm, and leg strength and the ability to make the motions needed to roll side to side; to assume sitting, kneeling, and standing positions; and for walking and propelling a wheelchair. Assessments of how the strength of muscles and the range of motion at joints affect mobility are usually done in an outpatient clinic or in the home by a therapist. The therapist evaluates the person's strength, balance, and coordination, and then makes recommendations for treatment. The therapist can help you learn to use the appropriate devices and techniques for safely walking and moving from one place to another. A written plan for an exercise program may be worked out that can maintain and possibly improve overall range of motion and muscle strength. Special treatments are available for "frozen shoulders," spasticity, or muscle stiffness and overall weakness.

Help for the Caregiver

The caregiver may need instructions on how to safely assist a person with ALS. Adequate information and instruction will help improve care, reduce anxiety, and decrease the risk of injury to both the PALS and the person helping. Written prescriptions from your physician are needed for physical therapy evaluations and treatments. Check with your insurance company or case manager for information on policy coverage. Check with your therapist for information on Medicare coverage.

Conserving Your Energy

You can help yourself by taking charge of how you perform the tasks you need to do every day. How you perform the tasks of daily living can affect how you feel for the rest of the day. The following steps will help you determine what you need to do and how you do it.

- **Consider what routines are necessary.** Decide what you can do, what someone else can do and what can be eliminated from the routine. Examples of routines are dressing, grooming and bathing.
- **Consider the timing or scheduling of your activities.** Your time and schedule and your caregiver's time and schedule are equally important. Plan activity periods and rest periods; pace yourselves.
- **Consider the best use of your energy.** When you climb a mountain, you must have enough energy left to return to base camp. Use assistive devices whenever possible to help reduce fatigue and frustration. Assistive devices include walkers with wheels, wheelchairs for use when shopping, and handicapped parking cards or special handicapped license plates.
- **Eat high-energy foods in small amounts every two to three hours.** Eating large meals is tiring, and takes more time and energy than eating small amounts more often. The use of dietary supplements such as Ensure may be helpful.
- **Place frequently used items in the most convenient place.** Place heavier items on the lowest level that you can reach. This is especially important in the bathroom, kitchen and office.
- **Purchase "gadgets" or other energy-saving devices** based on recommendations from people who are knowledgeable about their actual success rates. Otherwise, you will have wasted time and money. Good information can be found at support groups. However, please keep in mind that not all suggestions at support groups or online will fit your specific problem or situation. Check with your ALS clinic staff or Les Turner home team member since they usually have heard comments or complaints about items or resources. Your occupational therapist can help sort out the good advice and determine if specific recommendations or techniques meet your needs.

Useful Gadgets

The following sections include examples of equipment, devices, and "gadgets" that can help you with everyday tasks. Examples of sources and suppliers of the items are given, but the supplier lists are by no means exhaustive.

For Weakness in Arms and Hands

Wrist and hand muscles may become weak, affecting the strength of the grasp for eating, dressing, grooming, and work activities. Many useful, inexpensive devices and gadgets are

available to help. Assessment of specific needs will help in selecting appropriate items. Hand devices are usually not covered by insurance. Check with the occupational therapist to determine sources and prices for devices. Ask which devices will help.

Aids for Eating

- Built-up handles on lightweight eating utensils
- Foam tubing on heavy-duty plastic eating utensils
- Utensil holder
- Large handled cup for hot and cold liquids allows all four fingers to fit through the handle, so grasp does not need to be strong
- Offset spoon or fork that can be angled right or left
- Clear plastic, clip-on plate-guard
- "Octopus" suction device, a non-slip disc; or Dycem, a gel pad used to stabilize plates and cups
- Long rigid or flexible straws
- Sports drink container with a straw
- Flexi-Mug
- Food blender
- Food thickener (Thick It, Thick & Easy)

Aids for Dressing

- Velcro closures for clothing and shoes
- Knit shirts, pull-on pants, fewer closures
- Elastic thread for cuff buttons, elastic cufflinks
- Large-handled button hook
- Stretchy shoelaces
- Long-handled shoehorn
- Fleece socks for cold legs and feet
- Adaptive clothing

- www.adaptationsbyadrian.com
- www.buckandbuck.com
- www.silverts.com

Aids for Hygiene

- Baby wipes for toileting
- Toilet attachments for cleansing, such as Lubidet
- Chair urinals
- Foam tubing on toothbrush handle
- Disposable Dentips for mouth cleaning
- Electric toothbrush with rotary brush, plaque remover (Braun, Colgate, Sonicare)
- Electric flosser (Waterpik)
- Electric tongue cleaner
- Hand-held shower hose
- Plunger-type liquid soap dispenser
- Long-handled sponge
- Wall-mount soap dispenser

Aids for Grooming

- Wall-mounted hair dryer
- Shampoo tray
- Foam tubing on handles of comb and brush
- Large-handled, lightweight comb and brush
- Long-handled comb and brush
- Nailbrush with suction cups
- Nail clippers on stabilizing platform or board
- Nail file holder

- Foam tubing on razor
- Electric shaver

Aids for Recreation

- Adjustable-height tilt-top over-the-bed table
- Card holders
- Electronic games or gadgets, with hand controls
- Computer games
- Electronic books
- Page turners
- Book holders
- Rubber finger tips for help in turning pages in books, magazines, or newspapers

Telephone and Monitor Aids

- Telephone adaptations such as headset, receiver holder, speakerphone, memory keys for dialing frequently used phone numbers or a voice activated phone
- Emergency call device (some are worn around your neck, some on your wrist)
- Infant monitor, walkie-talkie, wireless pager, portable doorbell for in-home communication

Devices for Computer Use

- Computer arm supports
- Keyboard aid (pointer), Futuro wrist brace
- Foot-operated computer mouse
- Head- or eye-control electronic/computer device
- Voice activated programs such as Dragon Speak

Other Useful Devices

- Voice-activated house controls (Google home, Alexa, etc.)

- Wide pen or pen with grip
- Key holder
- Hand Keyper (key holder, tab lifter, letter opener, magnet, nail file)
- Door handle levers
- Touch light switches
- Offset hinges for doors that widen the doorway without reconstruction

Mobile Arm Supports

These supports can be attached to an adjustable-height table on casters for use anywhere in the home. The supports allow horizontal and vertical arm motion, for reaching the plate and the mouth. They also work well over the computer keyboard. Table-mount clamps attach the arm support to a table or computer desk. A supinator attachment allows some rocking motion of the forearm trough. A T-bar attachment can be added to support a weak wrist. A therapist must order these attachments and can demonstrate their use.

Leg Supports

Many kinds of devices, braces, and durable medical equipment, or DME, are available to assist a patient with hip, knee or ankle weakness. The extent of weakness and of endurance should be properly evaluated before any recommendations for equipment are made. Check with the insurance company or Medicare about coverage and which preferred providers or vendors they use. Gait, or manner of walking, should be evaluated by a physical therapist in an outpatient clinic to test stability and safety when using the appropriate walking aid. A prescription and a letter of medical necessity are needed, that can be written by either the therapist or the physician, but must be signed by the physician.

Aids for Walking

- Early in the disease process, a straight cane such as the Hurry Cane, helps provide balance and stability. Even if it seems unnecessary, using a cane will help make other people more careful and less likely to bump into you.
- Rollator walkers are a good choice for persons with leg weakness, stiffness or poor walking endurance. This type of walker provides a stable base while ambulating and allows a person to sit and rest.
- A standard walker, the lightweight, folding type can be easily carried in a car. Swivel front wheels and back gliders can be added if needed.
- Considerations in making a selection for a walker include: overall stability for your height (the three-wheeled walkers are not as stable), adjustability of the height of the hand grips, the type of hand brake system, ease of operation and stability, ease of turning

the walker, folding feature for easy transport in a car, and seat and basket options. Standard folding walkers can be adapted with front casters either 5" non-swivel casters or 3" swivel casters and back gliders.

Aids for Transfers

- A gait belt is used around the person's waist so a caregiver can assist with standing or sitting. Some belts have buckle closures, some have Velcro and some also have handles. Check with your clinic occupational therapist or physical therapist for the appropriate style for you and for training in its use.
- Transfer boards are used to move between a wheelchair and a bed or the seat of a car.
- The BeasyTrans® is a sliding transfer board, which consists of a sliding disc on a transfer board.
- Hoyer lift – this is a hydraulic lift mechanism that utilizes a sling to safely transfer a patient in/out of bed, into a chair and/or onto the toilet. This requires a prescription and letter of medical necessity from a physician which is then submitted to insurance. Once you receive the equipment, training from a physical therapist is needed. Medicare will usually cover the cost of a manual lift but not an electronic lift.
- The Easy Pivot Lift is a mechanical lift. This lift tilts the person forward to make dressing and toileting easier when the person cannot stand. However, it can place pressure on a person's diaphragm which could affect breathing. In addition, a PALS must be able to hold on to the handles and have good head control.
- The Lyko Lift is a power lift that stands a person before transfer. This lift may possibly be purchased through insurance with a prescription and a letter of medical necessity.
- **Note:** Medicare and public aid will not cover the cost of the Easy Pivot or Lyko Lift but they may be covered by private insurance.
- For information on hydraulic and power lifters and ceiling track lifters, talk with your occupational therapist about what device would be most appropriate.
- Seat-lift recliner chairs assist the patient to a standing position, change position to make the patient more comfortable when sitting or reclining, raise the legs and feet to reduce or avoid swelling, or change position to support the head and neck in a reclined, comfortable position. Insurance should cover the cost of the electronic portion of the chair with a prescription and letter of necessity from your physician.

Aids for Swelling in Lower Extremities

- TED hose are used to reduce mild swelling in feet, ankles and legs. They also promote circulation to help reduce the risk of blood clots. These are available by prescription from

your physician and can be obtained at your pharmacy.

- Jobst garments are used to control or reduce mild to moderate swelling in feet, ankles and legs. They require a physician prescription and measurement for accurate fit.
- Sequential compression devices require a physician's prescription. Phlebopump, Ace /pp-1000 (1 888 4PHLEBO) is used for severe swelling of lower extremities.

For Weakness in Both Arms and Legs

A variety of devices, some of which are listed below, are available that can make doing everyday tasks easier when you have weakness in both arms and legs.

Aids for Toileting

- A raised toilet seat, made of molded plastic 4 to 5 inches in height, fits snugly inside the rim of the toilet on most toilets. It has all smooth surfaces and can be easily cleaned. It can be removed easily when other family members use the toilet. It can be carried in a zippered bag or shopping bag for use when visiting other homes.
- A standard commode can be placed over the toilet to provide a raised seat and armrests.
- A shower commode chair on wheels can be rolled over the toilet to provide a raised seat and armrests as well as being used in the shower.

Aids for Bathing

- A shower commode chair on wheels has a padded seat and back and floor brakes or wheel locks. Padded arm troughs can be ordered if needed. Some commode chairs have tilt seats with headrests, reclining backs or straight backs. They can be used at the bedside, over the toilet and in shower stalls.
- Bath benches are set up across the side of the tub; they can be plastic or padded. They cannot be used with tubs that have sliding glass doors unless the doors are removed and a shower curtain has been installed. The bench also cannot be used if a vanity is located next to the tub because the patient does not have enough legroom to turn while sitting. Benches are available through numerous resources such as www.performancehealth.com, Amazon, Walgreens, CVS, etc.
- A tub seat is a small seat that can be placed inside the tub. It can be used with tubs that have sliding glass doors. A tub seat can be used only if the patient can step into the tub. It must be placed in the tub after the patient steps over the side of the tub.
- High-back resin deck chairs with arms can be used in the shower stall if the stall is large enough. They are very lightweight.
- A simple wooden bar stool can be placed in the shower stall to provide a high seat, making it easier for the patient to stand up.

- A hand-held shower hose attached to a showerhead or faucet allows water to spray from an appropriate height.
- Soap dispensers can be suctioned to a tiled wall to hold shampoo, conditioner, or soap. There are no bottles or caps to turn or drop.
- Long-handled scrubbers can be used to clean feet.
- Long foam-handled razors provide better grip and length.
- Grab bars securely fastened in the shower wall at the appropriate height can provide a "shelf" for weak arms to rest on while washing your hair, shaving, washing your face.
- Tub rail clamps securely to the side of the tub for stability while climbing into and out of the bathtub, approx. 12 inches to 15 inches high.

Resources/Catalogs

- Performance Health (formerly Patterson Medical)
 - www.performancehealth.com
 - 1-800-323-5547
- Gold Violin
 - www.goldviolin.com
 - 1-877-648-8466
- Harriet Carter Catalog
 - www.harrietcarter.com
- QUEST Magazine: Muscular Dystrophy Association
 - www.mdausa.org
 - 520 529 2000
- AliMed
 - www.alidmed.com
 - 1 800 225 2610
- Amazon
 - www.amazon.com

Chicago Area Suppliers of Durable Medical Equipment (DME)

- Walgreens – various locations
 - www.walgreens.com
- Mark Drug Medical Supply – Wheeling
 - 847 537 8500
- Fitzsimmons Surgical Supply – Tinley Park
 - 708 532 1199

For those PALS seen at the Lois Insolia ALS Clinic or by a member of the home and community team, please contact your clinic nurse or Patient and Family Advocate for assistance before ordering equipment.

Orthotics

Orthotics are orthopedic appliances such as splints and braces that are used to support or straighten weak areas of the body. In ALS, orthotics may be used for weakness of the neck, trunk, arms, and legs. A certified orthotist evaluates a patient's need for a specific orthotic. Orthotics usually require a prescription from your physician. Your insurance company may require that you use only a particular supplier or suppliers, such as one in the insurance company's network. Contact your insurance company or case manager to check on suppliers before making an appointment. Insurance may not cover shoes, or shoe inserts depending on the specific diagnosis. Medicare covers most orthotics. Check with the orthotic company.

Orthotics to Support, Protect and Rest Weak Neck Muscles

- A buddy pillow is a buckwheat travel pillow with a fleece cover. It supports the neck in bed, in the recliner chair, in the car, or on the plane.
- A soft cervical collar is a simple, inexpensive orthotic that can be purchased at Walgreen's, CVS, Bed, Bath and Beyond, Osco or any medical supply house. The collar may restrict swallowing if too snug. It is not covered by insurance.
- A plastizote collar is lightweight, flesh-colored, firm foam, two-piece collar with a chin support. The chin support is not movable. The collar may cause pressure under the chin or on collarbones. It does not restrict swallowing or feel tight around the throat, but does not allow the patient to speak or eat. A prescription from your doctor and a fitting by an orthotist are required.
- Aspen, Miami J, or CervMax collars are light, gray, foam-lined, two-piece collars with washable liners. The structure is less firm than the Plastizote, but more comfortable. It does not restrict swallowing or feel tight around the throat. A prescription from your doctor and a fitting by an orthotist are required.

- A headmaster collar is a wire-foam collar with padded tubular frame, a chin support, and more open areas around the throat and neck.
- An oxford collar is made from wire and foam. It provides support for the chin and the back of the neck, and allows side-to-side motion of the head and bending and stretching of the neck. A prescription from your doctor and a fitting by an orthotist are required.
- Soma-occipital-mandibular-immobilizer provides firm chin and neck support and has a chest harness to support the weight of the head over the shoulders. It is more important for walking than sitting. A prescription from your doctor and a fitting by an orthotist are required.

Orthotics that Help Arm and Hand Weakness

Shoulder supports must be evaluated by your physician and therapist. Assessment of shoulder joint mobility, circulation, and range of motion is needed for proper selection of the correct orthosis.

- A wrist-hand orthotic (WHO), also called a forearm or resting hand splint, is used to support weak wrist and hand muscles during the day or at night. The splint should be as lightweight as possible. Most splints can be pre-formed, but some must be custom made. The occupational therapist will choose an appropriate splint depending on how much muscle weakness or stiffness is present. Wearing a splint on each hand while you sleep is usually not advisable, since one hand should be free. Alternate wearing splints on right or left hand each night or day. The Futuro wrist brace is available in Walgreens, Osco, and CVS. Sammons/Preston wrist braces; Neutral position WHO, and TheraPlus hand positioners are available from www.performancehealth.com.
- A thumb-wrist support, or wrap, is a functional hand orthotic (FHO), which supports the thumb and index finger to improve fine coordination. It does not place the thumb and index finger in pinch position, and it does not make the hand stronger. It is usually made from neoprene or very lightweight splinting material. Examples include the Neoprene thumb/wrist support or wrist/thumb wrap available from www.performancehealth.com.
- Slings for severely weak arms and hands will support shoulder joints and decrease the risk of shoulder subluxation or stretching of the shoulder joint with subsequent pain.

Trunk Supports

Trunk supports are used to support weak trunk muscles, improve posture, and relieve muscle pain from strained muscles while sitting or walking.

- Elastic abdominal supports provide mild to moderate support to abdomen and low back. Good hand strength or assistance from a caregiver is needed to put one on. Supports are available from local pharmacy such as Walgreens, Osco or CVS. No prescription is needed.

- Lightweight corsets give support to the trunk and low back but require more dexterity to put on. When wearing a corset, less trunk flexibility is possible when rising from a seated position. The corset must be ordered from an orthotic company and requires measurement and adjustment for a correct fit. A prescription from your doctor and fitting by an orthotist are required.

Leg Braces Help Foot, Ankle and Leg Weakness

- A knee-ankle-foot orthotic (KAFO) is a long leg brace. It is not recommended with persons with ALS because they are usually unable to take a standing position with the knee in a locked position.
- An ankle-foot orthotic (AFO), which used to be called a short leg brace, is used to stabilize weak ankle muscles, as in drop foot and in weak knee extension. It fits inside your shoe and usually should be custom molded to your leg. You might need an articulating ankle on this orthotic to allow movement at the ankle joint, or floor-reaction, which assists knee extension and helps lock the knee joint. This modification will help in climbing stairs.
- A supra-malleolar orthotic (SMO) is used to stabilize the ankle and forefoot.

Sources for Orthotics

- Ballert Orthopedics www.ballert.com
 - NW, 233 E. Erie, Ste 200, Chicago 312 787 4400
 - 2434 W. Peterson, Chicago 773 878 2445
 - 125 E. Lake Cook Road, Buffalo Grove 847 459 9006
 - 141 Front Street, Wood Dale 630 694 9305
- Scheck & Siress www.scheckandsiress.com
 - Rush, 1725 W. Harrison, #220, Chicago 312 942 2011
 - 1525 E. 55th St, #204, Chicago 312 757 5270
 - 2835 N. Sheffield, #301, Chicago 773 472 3663
 - University of Illinois, 1740 W. Taylor, Rm C100, Chicago 312 996 6450
 - 1551 Bond Street, Ste. 311, Naperville 630 637 4638
 - One S. 376 Summit Ave., Ct E, Oakbrook Terrace 630 424 0392

- 401 Harrison Street, Oak Park 708 383 2257
- 8641 W. 95th, Hickory Hills 708 599 8336
- 6629 W. Lincoln Hwy Rte. 30 Suite 1,
Scherverville, IN 219 864 9501
- Shirley Ryan Abilities Lab www.sralab.org 312 238 2810
(formerly the Rehabilitation Institute of Chicago)
345 E. Superior, Room 1764, Chicago
- Marionjoy Rehabilitation Center www.marionjoy.org 630 909 8000
26W171 Roosevelt Road, Wheaton

Hospital Beds, Mattresses and Lifts

Hospital beds and appropriate mattresses can help in positioning and in preventing such pressure-related problems as bedsores. Hospital beds require a prescription and a letter of medical necessity from your physician in order to receive insurance coverage.

Hospital Beds

- Manual frame: The mattress height can be set at low or high position; manual cranks are used to change the position of the head and foot sections.
- Semi-electric frame: The height can be set at a low or high position. A power switch raises the head and foot positions.
- Full electric frame: A power switch adjusts the bed frame height to make transferring possible either from the wheelchair or from standing at the bedside, as well as adjusting head and foot positions. Full electric frame does not mean a full-size bed.
- Side rails: Full-length or half-length side rails give you leverage to turn yourself from side to side if this is difficult. Half-length rails make it easier to transfer to and from the bed.

Pressure-Relief Pads or Mattresses

- Egg-crate foam is used under the bottom sheet. It does not provide enough pressure relief for long-term use.
- Artificial sheepskin can be used under the sheet. Use on top of the sheet allows more air circulation. It is washable and more buoyant than egg-crate foam.
- An alternating pressure mattress is used under the sheet. It works with an electric compressor to raise and lower pockets of air under the body area. A prescription and

letter of medical necessity is required.

- Thermorest air mattress is used under sleeping bags, provides insulation and pressure padding. A nylon cover allows easier movement in bed when the mattress is placed under the sheet. Various depths are available from sporting goods stores and from L.L. Bean.
- Roho mattress is available as a low-profile or high profile air mattress. It can be a sectional (three sections for a hospital bed) or a full-length bed mattress. A prescription and a letter of medical necessity are required.
- Temperfoam mattress, a gel-foam mattress or pad provides maximum pressure relief. It is heavy once in place, and needs a prescription and letter of medical necessity.
- A low air-loss mattress moves air from one side of the mattress to the other to reduce pressure under the shoulders, hips, knees and ankles. A prescription and a letter of medical necessity are needed for insurance coverage.

Patient Lifts

- Hydraulic patient lifts, such as Hoyer and Invacare, are used with a separating sling. This type of lift supports the person in a seated position. The separating sling can be placed under a patient who is sitting or lying down without physically lifting. It also can be removed without lifting the patient. The Easy Pivot® lift uses two straps behind the shoulder and knees and tilts the person forward for transfers.
- Power patient lifts (Hoyer and Invacare), are similar to the hydraulic lifts, but are used with a battery- powered source attached to the lift. The Lyka Sabena Ilee® support and stands the person before transferring to a wheelchair.
- Ceiling track patient lifts use electric power.

Wheelchairs

Determining what kind of wheelchair is appropriate depends on the person's short-term as well as long-term needs.

Insurance Coverage

Check your health insurance policy to find out if durable medical equipment (DME) is covered or ask your insurance case manager. Most insurance policies will cover only one wheelchair; therefore, it is recommended to use insurance for a power/electric wheelchair. A manual wheelchair, needed for transportation and safety, is usually priced between \$200 and \$2000 if custom made. A power wheelchair, needed for independence and weight-shifting, is usually priced at \$25,000 or more. Medicare will cover a manual or a power wheelchair but usually not both; once a manual wheelchair has been covered; it is unlikely that a power wheelchair will be covered. Check with your therapist before you need a wheelchair to plan the best strategy.

Factors to Consider When Buying a Wheelchair

- Your age and the age, health, and strength of your caregiver, who may have to place a manual wheelchair in the car
- The type of car and its storage space such as hatch back, minivan, full-size van, and van with wheelchair conversion; and the space available in the garage and driveway
- The entrance or exit to your home, including placement of outside steps, inside steps, railings, deck, outside porch, and enclosed porch; and the possible need for a ramp or a porch lift
- The widths of the front and back doors and of the interior doors, especially the bedroom and bathroom
- The widths of the hallways and the space available for turning into and in the bedroom and bathroom

Wheelchair Features

- Lightweight, manual wheelchairs are used for transportation to and from the car. They can also be useful if the patient has retained enough strength to move himself about or if a caregiver is available to push the chair. However, patients should not sit in this kind of wheelchair for a long period of time. The chair must have a pressure-relief seat-cushion. Other useful features include detachable armrests and swing-away footrests. Some manual wheelchairs have four small wheels, others have quick-release large back wheels; both of these kinds are easier to place in a car. Types of manual chairs include: transport type, lightweight manual, and ultra-lightweight manual wheelchair.
- Power wheelchairs are used for independent mobility and for independent weight-shifting to decrease risk of pressure sores. Various features are available for these chairs. These chairs should be custom made and require a prescription from your physician for insurance coverage.
- Tilt-in-Space seat in a power wheelchair can be tilted back to relieve pressure on the seat or lower back. It requires special electronics to move the chair forward and change the seat position. It usually needs standard footrests.
- A tilt and recline system enables the seatback to be fully reclined. Elevating leg rests may be needed in the reclined position.
- Adjustable seat height allows the seated position to raise and lower from wheelchair base.

Other seating features include molded back inserts, tall back inserts, custom contoured back inserts, lateral supports for weak trunk muscles, head rests, molded seat inserts, custom contoured seat inserts, various kinds of pressure-relief cushion fillers including air, gel, gel-foam, and foam; adjustable height armrests; desk-length armrests; full-length armrests; regular arm

pads; trough arm pads with hand supports; swing-away footrests; heel loops; elevating leg rests; and angle-adjustable footrests. Electronic environmental control devices can be used to turn on lights or to access TV, VCR, stereo/CD, computer, and telephone.

The joystick control can be modified for different degrees of hand weakness. The joystick control box can be placed to the right or to the left or in the center of the wheelchair base, depending on the person's ability to manage it. Varying drive speeds can be programmed as well as varying amounts of pressure needed to push or pull the joystick. Head or foot control can be used if hand control is not possible. An attendant control can be placed near the headrest. Breath control is used for some persons with ALS. All features must be assessed by both the therapist and the wheelchair specialty representative.

Motorized Scooters or Carts

Scooters and carts are known as power-operated vehicles for Medicare coverage. They are usually not recommended for persons with ALS because they do not provide adequate back support, head support, or arm support.

Van Conversions

When you are considering using a manual or power wheelchair, you must consider how you will transport it in your present car or vehicle. You may need to consider a mini-van or full-size van, depending on the person's ability to transfer from the car to the wheelchair, the strength and ability of the caregiver, and your lifestyle and resources. Get good advice before purchasing the wheelchair or van. Consider that your garage may need a ramp or other modification. If you do not use a garage and you park on the street, you may need a special parking zone sign from the city for your parking area.

- New Ability Inc. www.newabilityinc.com 708 345 3939
- Mobility Works www.mobilityworks.com 630 782 1900 or 847 673 4300
- Sherman Dodge www.shermandodgeillinois.com 888 430 3783

Home Modifications

Each person with ALS has a different course of disease progression, a different lifestyle, different resources and different family commitments. Decisions about modifying the home to ease care and mobility problems should be made with careful consideration of both short-term and long-term needs of the patient and the family. Trilevel, bilevel and two-story homes with turning stairways are the most difficult challenges. Solutions depend on family resources. Moving the person with ALS to the most accessible level of the home and to make modifications on that level to meet needs for toileting and bathing may be the most feasible solution in the long run. Check with companies that use Americans with Disabilities Act (ADA) guidelines and modify homes in your area.

- Extended Home Living Services, Inc.
 - www.ehls.com
 - 847 215 9490

- Given the Ability, Inc
 - www.giventheability.com
 - 866 568 9704

- Home Access Services, Inc
 - www.homeaccessservices.com
 - 877 491 5525

- 101 Mobility
 - www.101mobility.com
 - 847 906 8897

- Home for Life Advantage, Inc
 - www.homeforlifeadvantage.com
 - 630 466 2611

Easy access to the home for walking or using a wheelchair must be considered. Keep an open mind and look at all the options. A platform area for the wheelchair is needed for safety and stability inside and outside the entrance doorway. This platform must be at the same level as the doorsill.

Ramps

A platform at least 36 x 36 inches will allow the wheelchair to safely sit outside the door before going up or down the ramp. A handrail or wheel rail should be attached along the sides of the ramp. Ramps for outside the home can be constructed from deck wood; for inside the garage or home they can be made from plywood. The maximum recommended angle, or grade, for indoor ramps is 12 inches of ramp for every 1 inch of rise or a 1:12 ratio. A 12-foot ramp is recommended for a 1-foot rise. Sometimes a sharper, steeper rise is needed because there is not enough space for a longer ramp. Remember who is pushing the wheelchair up or down the ramp; more strength is needed to control the wheelchair on a steeper ramp. Ramps outside the house should use a 1:20 inch ratio (a 20-foot-long ramp for a 1 foot rise), which provides a generous, long ramp. Walkways along the side of the house may allow space for such a long ramp. A "Z"-shaped ramp is necessary where short front yards or backyards do not provide enough space for a long, safe straight incline. A 5-foot flat area at the bottom of the ramp is recommended for stopping and turning the wheelchair. Local building ordinances must also be taken into account in planning a ramp. Portable, folding, aluminum ramps are commercially

available. These can be taken in the car or van for use when you go to a place that has one or two steps and no ramp.

- Handi-Ramp, Inc. www.handiramp.com 800 876 7267
Constructs aluminum and galvanized steel ramps and concrete decks for homes and businesses.
- Performance Health Catalog www.performancehealth.com 800 323 5547
Has portable aluminum ramps
- American Ramp www.americanramp.com 800 649 5215
Provides affordable ramps for rent or purchase
- RampNow – sales and rentals www.RampNow.com 630-892-7267
- Access Living 312 253 7000
Assists Chicago residents with the cost of ramps based on need and available funds

Porch Lifts

Porch lifts can be placed at doorways inside or outside the home, depending on the placement of stairs and the space for the lift itself. Porch lifts can be placed inside bi-level and tri-level homes and allow use of two levels without major renovation to the home. The installer will assess construction requirements.

Stair Lifts

Stair lifts can be rented or purchased and can be fitted to straight or curved stairs. Costs depend on length and curve of the track. Sitting balance and neck weakness of the patient must be considered. Some stair lifts have a fold-up seat. A wheelchair or other mobile chair at the top and bottom of the stairs is needed if the person cannot stand.

- Acorn Stairlifts www.acornstairlifts.com 888 211 1245
- Bruno Independent Living Aids, Inc. www.bruno.com 800 882 8183
- Extended Home Living Services www.ehls.com 847 215 9490
- Home for Life Advantage, Inc www.homeforlifeadvantage.com 630 466 2611
- Stannah Stair lift www.stannah.com 800 877 8247
Installs new and used stair lifts, sale and rental, for homes and businesses.

Ceiling Patient Lifts

Ceiling lifts can be installed over the bed, in the bathroom, or at the top and bottom of stairs to meet individual needs.

- Sure Hands Lift Systems www.surehands.com 800 724 5305
- Barrier Free Lifts www.bfl-inc.com 800 582 8732
- Home for Life Advantage, Inc www.homeforlifeadvantage.com 630 466 2611
- Waverly Glen System, Ltd. www.waverlyglen.com 800 265 0677

Elevators

Elevators can be installed for two or three levels within a home but assessment for adequate space is necessary

- Extended Home Living www.ehls.com 847 215 9490

Door Width and Halls

A doorway must be least 32 inches wide with a door that swings inward. Offset door hinges can replace regular door hinges if there is enough room to set the door behind the door jamb. This will give you about 1 to 2 inches more clearance. Wheelchairs are too wide to go through most bathroom doors. If the bathroom doorway is at least 24 inches to 25 inches wide a rolling shower commode chair can be used. Most shower commode chairs are 21 inches to 22 inches wide, and can be used over the toilet or in the shower and go through most bathroom doors easily.

Bathrooms

Shower stalls are easier to negotiate than bathtubs. Remodeling is very expensive, but a tiled floor with a recessed drain allows a shower commode chair easy access for patient and caregiver. An oblong shower stall can be modified by adding a wood deck and removable ramp. Glass doors must be removed and replaced with an expandable curtain rod and a shower curtain. Place the curtain rod inside the shower area to prevent the water from dripping outside the shower stall.

- Best Bath System www.best-bath.com 800 727 9907
- Extended Home Living www.ehls.com 847 215 9490
- Home for Life Advantage, Inc www.homeforlifeadvantage.com 630 466 2611

Resources for other Assistive Devices:

<http://www.every90minutes.org/als-technology-guide/>

Disclaimer: All care has been taken in preparing this document. This information is of a general nature and should be used as a guide only. Always consult your health care team before starting any treatments.

Patient and Family Resource Guide to ALS

Section 5 Caring for Persons with ALS

5

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847 679 3311
lesturnerals.org
info@lesturnerals.org

Caring for Persons with ALS

Often ALS progresses to the point at which a person can no longer be independent in the activities of daily living (ADLs). It may even progress to the point at which the family can no longer provide the needed care. In these circumstances, the options are to hire a caregiver to supplement the family's care of the patient or to consider a nursing home. This section addresses both in-home care and nursing home care, how to choose between them, and the financial aspects of each. It also covers health insurance and additional resources that may be used to care for the patient.

Caring for Persons with ALS at Home

The term *home health* can be very confusing when discussing caring for a person with ALS (PALS) at home. A person can receive services in the home from various health care professionals from a home health organization, such as a nurse, a nurse's aide and a physical therapist. The organization is usually affiliated with an area hospital. A doctor's order is necessary to initiate the process of identifying a need for health care professionals and to bill the services to the health insurance company or to Medicare. In addition, these services can only be provided for a limited time. If family members cannot provide all the care needed, hiring an in-home caregiver could be considered if family finances allow for such an expense. Long-term assistance with the activities of daily living, which may be called maintenance, custodial or unskilled care is rarely covered by health insurance policies. It may, however, be covered by a long-term care policy. Some PALS may also be eligible for a state-funded assistance program that helps to provide care in the home.

Home Health Benefits

The five ADLs that are used as a measure of need are: dressing, bathing, feeding, toileting and transferring. Typically, the need for assistance with three of the five activities will satisfy the requirements for benefits. The following programs may provide assistance in obtaining and paying for in-home care.

Department on Aging: Eligibility for benefits checklist provides an evaluation of the services you may be entitled to at a low or no-fee rate.

Illinois Department on Aging (www.illinois.gov/aging) 312-814-2630

Chicago Department of Aging (www.illinois.gov) 312-744-0784

National Council on Aging: This website helps disabled people, as well as seniors, to identify programs that may improve the quality of their lives. www.benefitscheck.org

Community Care Program: This program is administered through the Department on Aging. It provides in-home personal care at sliding-scale rates. The patient must be at least 60 years old. There are savings limits for individuals and couples, not including one's home and car. Application must be made through Public Aid. www.cityofchicago.org

Respite Care: This program provides 5 to 7 days per calendar year in a nursing home in addition to an allotted weekly maximum number of hours of in-home care. Check with the Community Care Program case manager regarding contracted nursing homes. The Les Turner ALS Foundation has a grant program to assist families with the cost of respite care. Certain restrictions apply.

For more information, please contact the Les Turner ALS Foundation’s Director of Patient services at 847 679 3311.

Illinois Department of Human Services is also called DHS (formerly the DORS/Department of Rehabilitative Services) program. This program is similar to the Community Care Program; however, the person with ALS must be less than 60 years old. The Personal Assistant program provides in-home personal care at sliding-scale rates. The agency also may provide an electronic emergency response system, home delivered meals, assistive equipment or environmental modifications.

Call for an evaluation to be made in your home.

Website: www.dhs.state.il.us

General inquiries 800 843 6154

Client information services 800 641 3929

Long-Term Care Insurance: A privately-held long-term care insurance plan may cover all or part of the cost of in-home and/or nursing home care with contracted agencies or facilities. Call the insurance company and ask for a case manager to review benefits and requirements.

Veterans Administration: The Department of Veterans Affairs offers many services to veterans with ALS. ALS veterans with an honorable or general discharge from the military should contact the VA as well as a National Service Officer for the Paralyzed Veterans of America (PVA) to receive information concerning benefit options.

Veterans Affairs 877 222 VETS www.VA.gov

312 663 5510

708 865 6580

Paralyzed Veterans of America pva.org

800 424 8200 ext.12

Chicago Area Representative 708 202 5623

Hiring a Caregiver Privately: The cost for an in-home caregiver can range from \$18 to \$25 or more per hour; the differences in price may vary with the level of experience. Many agencies

provide excellent caregivers for a fee and the staff at the Les Turner ALS Foundation can provide names of agencies that have ALS care experience. Please call the Les Turner ALS Foundation Director of Patient Services, 847 679 3311 for a list of recommended agencies. A home-health care agency provides background checks and coverage for days off, as well as caregivers to interview and choose among. It also acts as a liaison between you and a caregiver you have employed. A caregiver can also be hired through word of mouth, referral from a friend or newspaper, or from a registry, which provides the names and phone numbers of caregivers.

Questions to Ask when Hiring a Caregiver

- Do you smoke?
- Do you mind pets in the home (if applicable)?
- How many days out of the week would you be available?
- What hours are you available; or if hiring a live-in caregiver, what days of the week would you want off?
- Would you be available 24 hours each day, or would you expect to have an 8, 12, or 16 hour block of time during which you are not responsible or available to help? If so, what block of time?
- How do you feel about being awakened during the night to assist the patient?
- Would you be willing to do the following?
 - Cooking, meal preparation, feeding the patient?
 - Shopping, laundry, housework?
 - Patient's hygiene: dressing, bathing, toileting, etc.?
 - Assisting patient with ambulation that may include lifting?
- Would you be willing to learn to prepare tube feedings?
- How would you expect to be paid, by the hour, day, or week?
- Do you know anything about ALS, and have you worked with any other people with ALS ?
- Have you worked with anybody who is disabled?
- Do you have references that I can contact?
- When you are off duty, would you be staying here?
- Are you able to do lifting? Do you have any physical limitations or restrictions on your activities?

- What kind of sleeping arrangements do you require? Would you share a room or do you want your own room and bathroom?

Questions to Ask of an Agency

- Do you have male and female caregivers from which to choose?
- How long has the agency been in business?
- Is the agency licensed and accredited by appropriate governmental agencies?
- What kind of background checks does the agency run on employees?
- Are these professionals bonded?
- Does the agency supply references for its professionals?
- What language(s) do the caregivers speak?
- Exactly what services can we expect from a caregiver?
- What are the hourly fees or daily rates?
- How is the billing and payment of services handled?
- Who is responsible for the caregiver's taxes and social security contributions?
- Does the agency cover the caregiver for worker's compensation?
- Will the services be provided by the same person each day (if you are not hiring a live-in caregiver)?
- What kind of backup system does the agency have for emergencies, holidays, or sick days?
- Is someone at the agency available to handle questions or problems 24 hours a day?
- Does the agency supervise the caregivers?

For a listing of care agencies, contact the Les Turner ALS Foundation's Director of Patient Services, 847 679 3311

Nursing Home Care

Deciding if a Person with ALS Needs Nursing Home Care

Determining whether a loved one needs nursing home care is a difficult and often painful decision. Considering the following questions may help in making the decision.

- How much physical assistance does the patient need?
- How many times each day does the patient need to be transferred?

- Is the caregiver physically able to do the transfers without being in physical danger?
- How many times each night is the caregiver awakened to help the patient?
- Can the caregiver rest during the day if necessary?
- Is anyone available to help the patient and the caregiver with hands-on care?
- Is anyone available to help the patient and the caregiver with other household tasks?
- Are there other people to call on for help?
- What are the financial resources?

Nursing Home Care

Nursing homes provide two kinds of care:

Skilled Care: A registered nurse is on duty 24/7. Services of specially trained professionals, such as physical, occupational, and respiratory therapists, may also be included.

Maintenance care: Services are provided that assists a PALS with activities of daily living. This includes personal care and hygiene, as well as ambulation (movement) and transfers from one place to another, e.g. from the bed to the wheelchair. Assisted living services are sometimes covered by Long Term Care Insurance. Check your policy.

Ways to Pay for Nursing Home Care

Medicaid: If you cannot afford to pay for a nursing home, you may be eligible to have Medicaid pay the costs. Application is through Public Aid, and there are financial restrictions. An individual may have no more than \$2,000 in assets, not including a home and car. A patient's well spouse may keep almost \$109,000 in assets, but the patient's name must be legally transferred off of the assets.

Private long-term care insurance: A private long-term care insurance policy is separate from Medicare or other types of health insurance. If there is a long-term care policy, read the policy carefully to determine its restrictions and benefits. Many policies require a statement of medical need from a doctor and a waiting period before benefits are paid.

Medicare: Nursing home benefits available through Medicare are very specific. Medicare pays only for skilled care in a facility with a Medicare license. Although we may feel that the PALS's needs require great skill, Medicare has specific guidelines that define "skilled" care and determine when it is needed. Under Medicare's definition, most ALS patients are considered to be in need of "maintenance care," and so are not eligible for nursing home benefits from Medicare.

Private pay: If none of the previous options are applicable, and the financial resources are available, you may choose almost any nursing home and pay privately.

What to Ask and Observe When Choosing a Nursing Home

- Who is the doctor who will provide regular checkups, write orders, and prescribe medication and devices
- Is the residence clean and odor-free and is the temperature appropriate?
- Can the room and bathrooms accommodate the necessary durable medical equipment, such as a wheelchair?
- Are the residents clean?
- Who will be the liaison for questions or concerns?
- Is a 24-hour emergency response system accessible for the patient? What kind of meals are served, and who will help the patient eat, if necessary?
- Have you ever had an person with ALS as a resident?
- Are private and semi-private rooms and baths available?

Hospice and Palliative Care

The World Health Organization (WHO) defines *palliative care* as the active total care of patients whose disease does not respond to curative treatment. The goal of palliative care is to achieve the best possible quality of life for patients and their families. Palliative care includes control of pain and other symptoms. It also addresses psychological, social, and spiritual issues. Until there is a cure for ALS, palliative care should be considered beginning at the time of diagnosis.

For the person with advanced ALS, palliative care in the home or in a nursing home can be provided by a hospice program with an interdisciplinary team of professionals. In order to obtain care from a hospice, a patient must have a doctor's referral indicating that the patient has a life expectancy of six months or less if the disease runs its "normal" course. The hospice may also require a signed statement from the patient and doctor which includes a DNR/DNV, i.e. Do Not Resuscitate/Do Not Ventilate order. This order means that the patient does not wish to be placed on a ventilator in case of an emergency, and that he or she does not want any heroic measures performed to maintain life. For example, this document would instruct medical personnel not to use cardiopulmonary resuscitation (CPR) if the patient's heart stops beating.

Hospice is covered by most private medical insurance policies and is part of the Medicare Part A and Medicaid plans. Some hospices are located in a hospital as a special unit, others may be located in a free-standing facility. However, all hospice services can be provided in the home. The hospice team includes a doctor, nurses, certified nurse's aides (CNAs), a social worker, and a chaplain. The team may also include a music therapist, a physical therapist, and volunteers who provide brief periods of respite for the caregiver. A nurse is on call to help answer questions 24 hours a day, 7 days a week. One of the goals of hospice care is to keep the patient comfortable and as pain-free as possible. The staff arranges for all the necessary medications and equipment pertaining to the terminal illness to be delivered to the home. The hospice team of professionals provides the patient and family with expertise in managing physical and emotional end of life issues.

The doctors at the Lois Insolita ALS Clinic at the Les Turner ALS Research and Patient Center at Northwestern Medicine will help determine when it is appropriate to involve hospice in the care of the ALS patient. The primary doctor remains involved in the case along with the hospice staff. In addition, the ALS Clinic staff and members of the Les Turner ALS Foundation's Home and Community team remain available for consultation and assistance while hospice is involved.

Resources

Medicare hotline for hospice benefits 800 MEDICARE

National Hospice and Palliative Care Organization for information on local hospices:

800 658 8898

nho.org

Illinois State Hospice Organization 888 844 7706

isho.org

EMERGENCIES

PALS may have or may develop other medical problems or conditions unrelated to ALS. The types of problems that could occur cannot be predicted by the ALS Clinic staff and many can be difficult to separate from end-stage ALS issues.

Emergency Response Systems

This small, waterproof call button can be worn around the neck or around the wrist. If a problem occurs, the person with ALS or caregiver simply presses the button and an electronic signal activates a special speaker that is wired through the phone line. The line is monitored 24 hours a day, seven days a week, and someone will immediately ask what kind of help is needed. The help could just be someone to help the individual up, such as a neighbor, or it could be the paramedics for a more urgent situation. Some systems will automatically send help for people with ALS who cannot speak.

Philips Lifeline www.lifeline.philips.com

1- 800- LIFELINE

5 Star Urgent Response www.greatcall.com

1-800-463-5412

Direct Link 708 755 8440

directlink1.net

When to Call and Who to Call

Conditions or symptoms that should be handled by calling 911 may include, but are not limited to, the following:

- Chest pain with or without shortness of breath, sweating or nausea may indicate a heart attack or other heart problem.
- Pain in the jaw or in the upper arm, with or without shortness of breath, sweating, or nausea may indicate a heart attack.
- Back pain between the shoulder blades or in the lower back that may be sudden and/or intense, with or without shortness of breath, sweating, or nausea may indicate a heart attack or other heart problems.
- Sudden changes in breathing may indicate a heart problem or a lung problem, such as a blood clot.
- Sudden changes in level of consciousness may indicate a stroke.
- Sudden changes in function, such as sudden inability to speak or to move an extremity may indicate a stroke.
- Sudden change in color or swelling or feeling in a leg or an arm may indicate a blood clot.

If the patient is in extreme distress and you are not certain what to do, **do not hesitate to call 911**. Do not spend valuable time attempting to reach the ALS Clinic doctor or nurse. **If the patient is in hospice care, call the hospice 24-hour number immediately**. The on-call nurse will suggest how to help the patient, and may also visit the home to evaluate and/or treat the patient's symptoms.

Resuscitation or DNR

Activating the local emergency response system by calling 911 requires certain actions by the Paramedics or Emergency Medical Technicians (EMTs) who respond. They are required by the State of Illinois to attempt resuscitation unless there is a valid Do Not Resuscitate (DNR) order or POLST on the premises. This means they must begin cardiopulmonary resuscitation (CPR) unless you can show them a DNR or POLST order signed by the patient. Paramedics may be required to defibrillate (shock) certain heart rhythms and to give various medications. State of Illinois guidelines also require them to transport the patient to the closest hospital, unless the designated hospital acting as medical control for that particular system gives them permission to transport to another location. These guidelines are mandatory and cannot be altered or changed by the paramedic or EMT once they are on the scene.

If the patient does not wish to be placed on a ventilator or to have heroic measures performed to maintain life, the family must have available a signed statement from the patient which includes a valid Do Not Resuscitate (DNR/POLST) order. For example, this document would instruct medical personnel not to use cardiopulmonary resuscitation (CPR) if the patient's heart stops beating. Everyone involved in the patient's care should know where the DNR/POLST order is located. Other advance directives, or legal forms can ensure that the patient's wishes to have or not have heroic measures performed to maintain life are followed. These forms include the 5 Wishes document or a Living Will, which spells out the patient's wishes and a power of attorney for health care which appoints another person to make health care decisions if the patient is unable to speak for her/himself. Consult with your physician, clinical nurse coordinator or Les Turner ALS Foundation Patient and Family Advocate or Social Worker.

Disclaimer: All care has been taken in preparing this document. This information is of a general nature and should be used as a guide only. Always consult your health care team before starting any treatments.

Patient and Family Resource Guide to ALS

Section 6 Nutritional Support

6

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5550 W. Touhy Avenue, Skokie IL, 60077
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lesturnerals.org
info@lesturnerals.org

Nutritional Support

Many factors can affect the nutritional status of a person with ALS. The goal of nutrition therapy in ALS is to maintain weight and muscle mass. Depending on individual symptoms, maintaining adequate nutrition may involve high calories and high protein foods, thickened liquids or pureed foods, or giving formula through a feeding tube. Your doctor and dietitian will be able to help recommend the best way for you to meet your nutritional needs.

Maintaining Weight

Severe weight loss equals muscle loss. Therefore, it is important that a person with ALS not become underweight. There are many reasons why people with ALS lose weight. Difficulty chewing and swallowing causes choking. Arm/hand weakness limits self-feeding. Other factors include decreased appetite, constipation, shortness of breath, fatigue due to the long and tiring process of eating, and increased metabolism with ALS.

Studies suggest survival significantly improved with early, aggressive nutritional management. While consuming adequate protein, vitamins and minerals is important for people with ALS, the most important dietary factor is the consumption of adequate calories. This prevents deterioration due to poor nutrition. The goal for people with ALS is to maintain weight and preserve muscle strength, endurance, and function.

Maintaining Calories

Studies have shown that ALS patients are hypermetabolic, meaning that they are burning more calories at rest, when compared to a healthy population¹. Contributors may include increased energy used in breathing, increased effort to move around, and muscle twitching.

This makes maintaining weight more difficult so ALS patients need to increase their calories to maintain weight. The challenge is to increase the amount of calories without significantly increasing the amount of food.

A few ways to increase calories would be to eat small, frequent meals during the day to provide more opportunities for calories (recommend to eat every 2-3 hours). Include high calorie or nutrient dense foods at meals/snacks (ie peanut butter, granola, dried fruit, muffins, pudding, nuts, and avocado). Avoid diet foods and choose the full-fat version for more calories. Add butter, honey, gravy, cream sauces, and mayonnaise to foods. Drizzle olive oil over foods such as vegetables, meat, and soup (1 tablespoon = 120 calories). Also, add high protein foods such as cheese, eggs, powdered milk to casseroles and soups to increase calories and protein.

Nutritional supplements are another way to increase calories and add extra vitamins, minerals, and protein. Commercial products include Boost Plus, Ensure Plus, Carnation Instant Breakfast, and Boost Breeze. There are also generic supplements available at most large retailers (Ex: Walgreens, Costco, Sam's Club and Walmart). Note that oral supplements with the word "plus" are better than the "high-protein" varieties as they contain more calories and protein.

Eating

Chewing and swallowing difficulties (dysphagia) make mealtimes exhausting for the person with ALS due to the need to concentrate and go slowly to keep from choking. Sometimes a person with ALS eats so slowly during a meal that their meal turns cold and everyone else is finished eating and the dishes are cleared up before the person is done – adding to a sense of isolation from the family.

Signs of swallowing difficulty include choking or coughing while eating, increased saliva or excessive drooling, sensation of food getting stuck in the throat or chest, needing more time to finish a meal, frustration during meals, avoiding certain foods, and loss of appetite.

If you notice any of these signs, try to determine what types of foods and liquids are the easiest to tolerate. If necessary, change the consistency of the foods taken in during the course of the day. A few strategies to make meals easier include cutting up food into tiny bites before eating, avoiding dry and crumbly foods, and using sauces and gravies to moisten foods and ease swallowing.

Drinking

Adequate fluid intake is essential for keeping saliva and mucus thin and avoiding constipation. Because drinking thin liquids can be difficult, and because drinking leads to urination, which can be time-consuming and require help, people with ALS sometimes don't drink enough fluids.

Ensure proper fluid intake with a goal of at least eight to ten 8-ounce cups a day. This will help prevent dehydration, which can lead to physical deterioration, constipation, weakness, headache, and thickened mucus that can cause choking. Also, avoid alcohol and caffeinated beverages, which can be dehydrating (although can be consumed with adequate hydration). Consider the use of a sports drink with electrolytes or other flavored beverages to increase fluid intake. Certain foods such as soup, Jello, sherbet, and fruit will also help to increase fluid intake.

Monitor for signs of dehydration:

- Dark colored urine
- Dry itchy skin
- Headache
- Confusion
- Dizziness/Lightheadedness
- Flushing/Fever
- Increased fatigue
- Decreased urine output

If thin liquids like water cause choking, serve thicker liquids such as milkshakes, smoothies, nectars, tomato juice, and pureed soups. Powders like Thick-It and Resource ThickenUp Clear add thickness to fluids without changing the taste. Other thickening options, which also increases calorie intake, include baby rice cereal, mashed potato flakes, and pureed baby foods.

Choking

As throat muscles weaken, in addition to weakness of the tongue and lips, the risk of food or liquids “going down the wrong way” increases and poses a real danger.

“The wrong way” means breathing food or liquid into the lungs (aspiration) instead of swallowing down the esophagus into the stomach. Aspiration can cause respiratory infections or a frightening choking spell, and is a leading cause of pneumonia, a life-threatening event in ALS.

You can reduce choking episodes in several ways:

- Do not try to talk while eating, as this and other distractions make choking more likely.
- Serve smaller but more frequent meals, avoiding dry, crumbly or large chunky foods.
- Take smaller bites and sips of liquids and swallow several times with each bite.
- Keep mucus and saliva thin by ensuring adequate fluid intake. You may also try papaya, pineapple, or lemon juice in water.

Feeding Tubes

If the person with ALS can't maintain his or her weight by eating or swallowing becomes too exhausting, time-consuming, or dangerous, a feeding tube should be considered. It is a much easier route to maintaining nutrition and hydration than trying to eat or drink everything by mouth. Feeding tubes can lessen the stress a person feels when they are unable to eat enough to maintain their weight.

A feeding tube allows the person to eat what they are able, then supplement with adequate calories through the tube. Vitamins and medications can also be easily taken through the tube. It is important to emphasize that having a feeding tube does not prevent oral intake, but offers a convenient method of nutrition, hydration, and medications.

Early studies suggest a correlation between feeding tube use and longer survival and better quality of life. People who use feeding tubes can save time and energy, which can allow time for more interesting things. It also gives control back to the person with ALS in terms of when to eat and how much. The feeding tube will severely lessen the likelihood of inhaling food or liquids into the lungs, which will prevent chances of aspiration pneumonia.

When to Get a Feeding Tube

As breathing becomes weaker, having a tube placed is more difficult because the person with ALS is at greater risk for complications. Ideally the tube should be placed before the FVC (forced vital capacity, a respiratory measurement) falls below 50 percent of normal, as the procedure is safer and recovery is easier. And having the tube placed while the person can still eat by mouth allows for a gradual transition to tube feedings. Even if the tube is placed and isn't used right away, it will be in place for when it is needed.

If the FVC is adequate, the tube is inserted directly into the stomach via a PEG (Percutaneous Endoscopic Gastrostomy) procedure. This is done under light anesthesia and will require hospitalization.

If the FVC has advanced beyond the safe level for a PEG, doctors may choose to perform a RIG (Radiologically Inserted Gastrostomy). Air is pumped into the stomach and the tube is inserted with guidance of a fluoroscope (an x-ray that projects images onto a screen) rather than an endoscope (a camera attached to a tube). The procedure has been shown effective and safe for those with moderate or severe respiratory impairment, although recovery may be more uncomfortable than with a PEG – another reason to get the tube early!

Maintenance of the Feeding Tube

For a few weeks after the insertion, the feeding tube requires special attention to prevent infection (as with any post-surgical wound). After the site is healed, daily cleansing with soap and water is all that is necessary.

Feedings through the tube are generally pre-mixed liquid formulas. Commercial formulas are often recommended over homemade formulas because they are nutritionally complete, sterile and much easier and less time-consuming for caregivers than homemade formulas.

The feedings themselves are generally put into a special feeding bag and allowed to drip into the tube by gravity. Following each feeding, the tube is flushed with water to clear it. Most often, several feedings are given through the course of the day, just as with regular meals. Some patients receive tube feedings overnight using a special pump and take only liquids and medications during the day. The dietitian, physician, and patient will determine the proper feeding schedule after discussion of needs and preferences.

Tips:

- Flush with room temperature water before and after putting anything through the tube. Gently squeeze the tube as the water is running through to dislodge anything sticking to the inside.
- Flush the tube with water prior to giving medication and follow by flushing again.
- Tube feedings sometimes cause heartburn or nausea because the stomach is being filled too full, especially after weeks of under eating. Solutions include having the feeding material at room temperature, sitting upright for a period of time after the tube feeding, feeding more slowly, giving smaller more frequent feedings, and checking with the doctor about heartburn medications.

Disclaimer: All care has been taken in preparing this document. This information is of a general nature and should be used as a guide only. Always consult your physician before starting any treatments.

References:

1. Bouteloup C, Desport JC, Clavelou P, et al. Hypermetabolism in ALS patients: an early and persistent phenomenon. *J Neurol.* 2009;256(8):1236-1242.

Adapted from the MDA ALS Caregiver's Guide, Chapter 5, Nutrition Issues. Published on <http://mda.org/print/book/export/html/37397>

Patient and Family Resource Guide to ALS

Section 7 Speech

7

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Speech/Communication

ALS often affects the muscles used in speaking and swallowing. These muscles include the lips, tongue, soft palate, larynx (voice box, throat), and the muscles used in breathing. Weakness and/or poor coordination of some of all of these muscles may lead to difficulty in pronouncing words clearly. In some cases, language and thinking skills may also be affected.

Problems in Communication May Include:

- Hoarse or strained voice
- Soft voice
- Unclear or slurred speech
- Nasal or muffled-sounding speech

In some persons with ALS (PALS), speech difficulties remain stable or relatively mild. In others, communication problems progress from mild to severe. At times, the PALS will perceive his or her speech problem differently than the listener does. Both the speaker and the listener should identify the factors that affect communication and work together to manage them. Speech-Language Pathologists (SLPs) at the Lois Insolia ALS Clinic at the Les Turner ALS Research and Patient Center at Northwestern Medicine can help people with ALS manage their changing communication skills for as long as possible. SLPs also can assist clients in adapting to alternate forms of communication, if necessary.

Tips for Maximizing the Communication Environment

- **Get the listener's attention before speaking.** Agree on a special signal the individual can use when ready to speak.
- **Speak face-to-face.** Most listeners use lip reading to help them understand speech. This is especially important when the speaker's mouth and face muscles do not move well.
- **Identify the topic of conversation (or interest).** Identify this at the beginning of the conversation, so the words chosen may be more familiar or expected.
- **Speak in a quiet environment.** At home, turn down extraneous noise such as the radio, TV or music. In a restaurant, find a quiet place for conversation.
- **Rephrase if you are not understood.** It may be necessary to change the words that you use if you are not understood.

Techniques to Maintain Communication

- Speak slowly and distinctly.
- Pause between phrases or thoughts, and even between words, if needed.

- Over-articulate speech by exaggerating consonants, especially the first consonant in the word. Learn whether the lips or tongue or both are involved.
- Clearly pronounce each syllable in longer words.
- Use energy saving ideas. Rest your voice if you know you will need to talk later in the day. Techniques that work in the morning may be less effective later in the day.
- Project your voice. Think of listeners as being farther away than they are.
- Use non-verbal strategies to add to your speech.

ALS communication specialists generally advise against rigorous, traditional exercises designed to strengthen weak or uncoordinated muscles, particularly when there are detectable speech disturbances. Exercises designed to learn and utilize speech strategies may help a PALS' changing communication needs. Some people will need just a few lessons to learn them. Others will be able to incorporate new strategies on their own. Ask your Speech-Language Pathologist for specific advice for your concerns.

Assistive Communication Devices

Assistive communication devices are any devices that may enhance a person's ability to communicate effectively. Communication difficulties in ALS vary and can change over time. Some individuals may need only one type of device; others may move from one device or access method to another as symptoms change and progress. SLPs assist in the selection of specialized equipment for a specific communication need. The types of devices available may be discussed at your Clinic visit.

Types of Assistive Communication Devices

- **Amplification:** A personal voice amplifier is used to make speech louder in people with soft or whispered voice. It is usually portable and works with a microphone placed near the mouth. Sometimes, increased volume is all that is needed for better understandability, but it cannot make less clear speech more intelligible.
- **Low technology devices:** Low technology devices include alphabet (letter) boards, word boards, picture boards or notebooks that can be used by pointing to the desired letter, word, picture or phrase. Anything goes! Some families create their own boards; others purchase a commercially produced product. Including a place for identification of a topic also helps to narrow down the ideas. These boards can be used with partner-assisted communication techniques, as explained below.
- **Moderate technology devices:** Moderate technology devices include smartphones and tablets (e.g. iPad, Android-based tablet, Kindle, etc.) that can serve as a communication device through various applications (apps) which are downloaded to the device. Many personal computers (desktops, laptops) can be modified to be used as a communication device.

- **Communication apps:** Communication apps can be purchased and/or downloaded for personal devices. Apps can be text-based or picture-based. Most provide text-to-speech features where the PALS types a message or part of a message that is then “spoken” when the item is selected. Offerings change regularly and are too numerous to list here.
- **High technology devices:** High technology devices include “dedicated” computers with voice synthesizers, usually called “speech generating devices” (SGD). These specialized products can be used through a variety of access methods as described in the next section. When a specialized SGD is being considered, an SGD evaluation by an SLP familiar with these systems is required before purchase. The SGD market changes regularly regarding products and funding so it is best to consult with an SLP. Many SGDs and apps are available in languages other than English.
- **Palatal lift:** A palatal lift is a dental apparatus, similar to a retainer, which is worn to keep air from escaping out of the nose during speech. It is an intervention for select speech problems, though recommended infrequently. It works by lifting the soft palate. Several visits to a specialized dentist, called a prosthodontist, are required for proper fabrication and fitting. This device is most appropriate for PALS whose speech problems progress slowly and who have primarily excess nasality during speech. It is not appropriate for those with rapid speech decline or weakness of the speech muscles, such as the tongue, which affects the majority of PALS. Insurance may cover a portion of the palatal lift but it is best to check your individual coverage.

Input Selection Methods for Communication Devices

Direct Selection

Direct selection is the most efficient selection method. It requires accuracy of the upper extremities, head or eyes.

- **By touch:** The user makes direct contact with the device, such as typing or touching a computer/device screen. A stylus also can be used.
- **By mouse:** The user moves the mouse around the screen and uses a “click” to select the desired target. There are adapted mouse devices for those with less dexterity.
- **By head mouse pointer:** This method requires the use to wear a specialized “dot” and move the head to the desired target letter or phrase on the SGD or computer.
- **By eye access selection or eye tracking:** This method involves directing one’s “gaze” to an on-screen keyboard or display and “dwelling” on the desired target letter or phrase. The “dwell” feature then selects the target to the text box or for the message to be spoken. This method is needed when other body movements are not possible.

- **Other:** Creative solutions may be developed by families to help with access. A laser pointer rigged to glasses or a baseball cap brim can also serve as a head pointer on a communication board or poster. If a laser pointer is used, though, care would be needed to avoid direct contact with someone's eyes.

Partner Assisted Communication

Partner-assisted communication is a technique most often used with a letter or word board between a person who is unable to use his or her hands and a communication partner or the "listener." The "partner" shows the board and/or reads the row name to the "communicator." When the desired row is mentioned, the communicator selects it by a mutually-agreed upon signal, such as a head nod or eye blink. Then the partner shows/reads the individual letters or words within that row until the communicator makes a selection. That selection becomes the first letter (or word) in the user's message. These actions are repeated until the message is complete.

Some letter boards are home-made. Others are available on the Internet or from an SLP. The **AEIOU** set-up is the most common and is organized by the vowels down the left column:

ABCD

EFGH

IJKLMN

OPQRST

UVWXYZ

Other boards can be arranged by quadrant or by color or by topic.

Scanning

Scanning is a method by which individuals who no longer are able to use a keyboard by typing can operate a computer or alternative communication device. This method takes longer for creating a message than direct selection. Displays, such as rows of letters or pictures (also known as icons) or sections, are "scanned," that is highlighted in a sequence, until the user activates a switch to make a selection. A section can be a row, several rows or even a block of icons. Once the general section is highlighted, then each individual icon within that section is highlighted until the user makes his or her choice. The number of icons displayed on the main screen can vary from only a few to more than 50 items.

There are many switches available and depend of the user's dexterity. Size, shape and access for the switch vary. A switch may be round, plate-like or a joy-stick. It can be activated by touch or motion. The particular switch may be activated by any body part or motion, such as hand, finger,

foot, eyebrow or eye blink. The SLP or possibly Occupational Therapist may suggest a particular switch depending on the PALS' skill.

Voice Banking

Voice banking is a process to preserve the PALS's voice and use it when needed on an SGD. The process needs to begin the PALS's voice remains relatively unaffected, that is early in the process. Alternatively, a family member can also complete the process if their voices sound similar.

The ModelTalker System is a revolutionary speech synthesis software package developed by the Nemours Speech Research Laboratory and designed to benefit people who are losing or who have already lost their ability to speak. It allows people who use a Speech Generating Device (SGD) to communicate with a unique personal synthetic voice that is representative of their own voice. Visit the website www.modeltalker.org for details. It requires recording phrases and sentences that subsequently will be synthesized into his or her voice.

As of July 1, 2017, there is a \$100 charge for users to enable download of completed voices.

BeSpoke™ Voices

BeSpoke™ Voices are uniquely created (customized) voices for an SGD that searches the VocaliD database for a voice match. It results in a digital voice that is understandable and personalized. A few seconds of vocalization of the voice recipient's voice is needed. The TobiiDynavox company is partnering with VocaliD in this project. Information can be found on www.tobiidynavox.com/vocalid. As of July 2017, the cost for this service is approximately \$1,200.

Specialized Features

Most SGDs and moderate technology devices, like smartphones and tablets, have specialized features that further shorten the task, reducing the energy used in creating messages. Even with a homemade word or letter board, these anticipation techniques can be helpful in improving the communication between user and listener.

- **Word completion:** The device anticipates how a word is spelled, based on the first few letters, and finishes it for you. Some programs will offer selections among which the user can select.
- **Word prediction:** The device anticipates words that come after one another in context. For example: If the phrase: "I want to" is formulated, the next word is likely to be "go." Sophisticated computer programs learn how phrases are used by a specific user or what it likely to be used based on English grammar rules.

Commonly Recommended Communication Devices

The ever-changing nature of the speech generating device market and funding regulations makes specific recommendations challenging. However, several companies have products which may be appropriate for some PALS.

Tobii-Dynavox: www.tobiidynavox.com

Asyst Communication Company www.asyst.us

Prentke-Romich: www.prentkeromich.com

Medicare Coverage

Medicare recognizes assistive communication devices as durable medical equipment (DME) which is a covered benefit under the Social Security Act. The Steve Gleason Act of 2015 (S.984) was signed July 30, 2015 and was supposed to end October 1, 2018. It amended the title XVIII of the Social Security Act to provide Medicare beneficiary access to eye tracking accessories for speech generating devices and to remove the rental cap for durable medical equipment under the Medicare Program with respect to speech generating devices (“S. 984 — 114th Congress: Steve Gleason Act of 2015.” www.GovTrack.us. 2015. June 7, 2017

<<https://www.govtrack.us/congress/bills/114/s984>). ALS advocacy groups were working to make this important legislation permanent and as of February 2018, the Bipartisan Budget Act of 2018 (H.R.1892) was signed. By signing this legislation, which included the Steve Gleason Act, there are no longer policy limits for speech generating devices for our PALS.

Eligibility for obtaining an SGD remains based on medical necessity, as determined by a PALS’ physician and Speech-Language Pathologist. A communication device evaluation by a licensed SLP is required.

Medicare’s policy covers 80% of the cost of the device, up to predetermined limits. The remaining portion can be covered by a secondary policy, outside funding source or self-pay.

Unfortunately, Medicare does not fund devices for users living in a skilled nursing facility or for those enrolled in hospice care. Benefits are allowable for those in palliative treatment, those living in an independent living facility or in assisted living. Medicare, at this time, funds SGD peripheral equipment, such as a wheelchair or rolling mount, or an eye access module, with proper evaluation and documentation. Medicare currently funds one device every five years, based on the date of shipment of the device.

It is best to inquire with an SLP for current Medicare policies regarding SGDs.

Resources for Evaluating Communication Devices

Centers in the Chicago region for demonstration and/or evaluation of high technology devices include the following locations:

- **The Technology Center at the Shirley Ryan Ability Lab (formerly the Rehabilitation Institute of Chicago (RIC)):** This Center is staffed by augmentative communication specialists, usually SLPs, and Occupational Therapists. In addition to evaluation for a dedicated SGD, RIC will evaluate for specialized hardware or software for full computer access, keyboard modifications and/or alternative mouse devices, as well as for environmental controls to help with daily living issues. A physician's referral is needed. Medicare and private insurance typically pay for an evaluation and follow-up visit (though as with any medical coverage, check with your insurance carrier for details). A wide array of communication devices is available for demonstration. www.sral.org. Technology Center contact: 312-238-2988. Main RIC phone number: 312-238-1000.
- **Marianjoy Rehabilitation Center, Wheaton, IL:** www.marianjoy.org. General outpatient services phone 630-909-7150.
- **VA Hospital System:** The VA Hospital System provides SLP services and evaluation to qualifying veterans. The PALS needs to be registered at his or her local VA to attend. The VA provides SGDs and access equipment as appropriate.
- Some SLPs affiliated with hospitals or free-standing centers can provide SGD evaluations with the assistance of an SGD vendor.

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Patient and Family Resource Guide to ALS

Section 8 Respiratory Care

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Respiratory Care

At some time in the course of the disease, ALS may affect the bulbar nerves. These nerves control the movement of the muscles related to swallowing, speaking, coughing and keeping the airway open for breathing. As the nerve cells of the bulbar region of the brain degenerate, these muscles waste away. In some people with ALS, respiratory muscles may be affected late in the course of the disease; in others, these muscles are affected early in the disease process. The nerves that control movement of the diaphragm, the major muscle of breathing, are also affected.

Because of weakened respiratory muscles, people with ALS have problems inhaling enough air and exhaling enough carbon dioxide. The carbon dioxide level builds up in the blood, and the oxygen level drops. This problem may not be obvious, but a variety of symptoms can indicate that breathing is affected. Treatment for breathing problems depends upon the cause of the problem. Additional testing may be needed to determine the underlying cause in a particular person.

Problems

Early in the disease, sleep may be affected causing inefficient breathing during REM sleep. Signs of breathing problems include:

- morning headaches
- daytime sleepiness
- sleeping poorly
- waking frequently
- snoring
- noisy breathing
- poor appetite
- shortness of breath when moving or when lying flat
- nausea (without such other gastrointestinal symptoms as vomiting or diarrhea)

Other symptoms of poor breathing at night can include increased irritability, forgetfulness, and apathy. If any of these symptoms appear, notify your clinic nurse at the Lois Insolia ALS Clinic to discuss possible solutions.

How Is Breathing Monitored?

Breathing problems will be monitored during regular visits to your ALS Clinic by tests that measure the strength of respiratory muscles and how well the lungs are functioning. In the clinic, the patient can exhale into a spirometer, a device that indicates how the ability to exhale, or Forced Vital Capacity (FVC), compares with that of most people of the same sex, height, and

age. A Maximum Inspiratory Pressure (MIP) will measure the ability to inhale forcefully. Other measures of breathing issues include a pulse oximeter, which is a device that noninvasively measures arterial blood oxygen saturation noninvasively using a comfortable finger sensor. An overnight pulse oximetry can detect changes in breathing during sleep. The measurement of arterial blood gases (ABG) is another test that can determine the severity of breathing problems. A small amount of blood is taken from an artery, and the amount of oxygen and of carbon dioxide in the blood is compared to the normal ranges.

For severe symptoms of breathing difficulty, especially in nighttime breathing, a sleep study might be needed to help find out the specific cause of the problem. In a sleep study, breathing, oxygen levels, leg movement, and sleep cycles are measured while the patient is asleep. This information can help determine the best course of treatment, whether it is medication or the use of a noninvasive assistive breathing device.

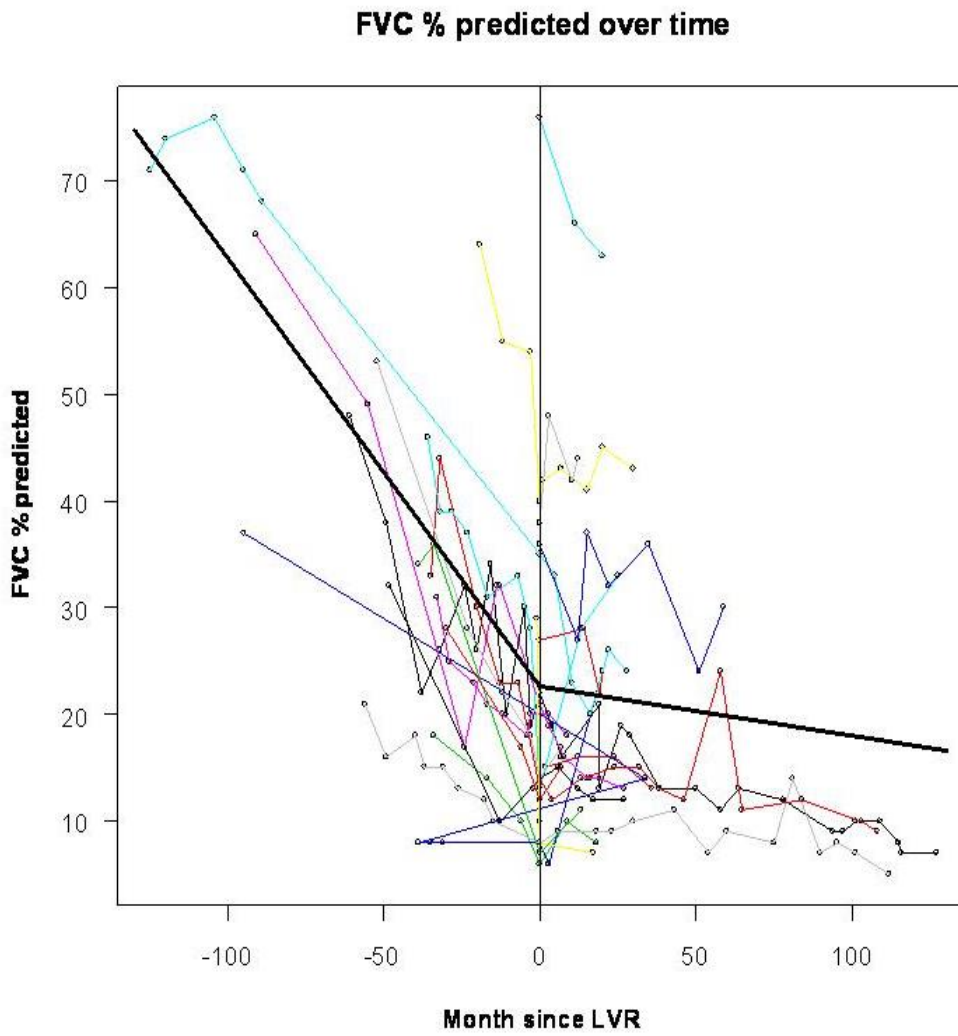
Noninvasive Solutions

Difficulty in breathing while lying flat may be relieved with the use of a wedge pillow or a hospital bed set so that the person with ALS lies at a 30–45 degree angle. At this angle, the main muscle of breathing, the diaphragm, does not have to work against gravity, as it does when lying flat. People who have a reclining chair may find relief by sleeping in that, as it allows you to lie at the appropriate angle for maximum breathing comfort.

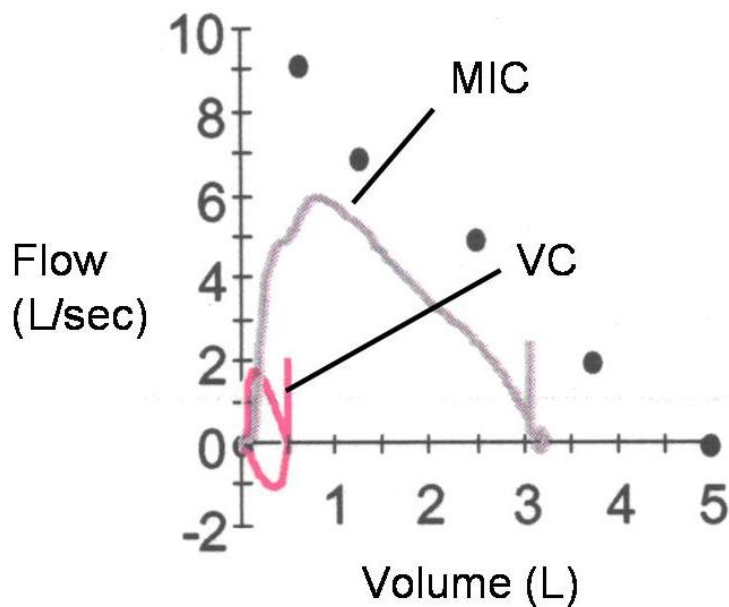
Lung Volume Recruitment

Lung volume recruitment or breathstacking can help to keep lungs open and chest muscles flexible. A daily routine of sitting tall and taking 5-10 slow, deep breaths twice a day will also help open those airways. This technique should be assisted with the use of an Ambu™ bag and mask if deep breathing becomes difficult. It is easy to do at home or on the go. This can be taught to PALS and caregivers during a clinic visit.





McKim, DA. (2011). *American Thoracic Society*, Presentation.



Lung volume recruitment is helpful because it assists the PALS with taking a bigger breath than they can take on their own. The example above shows a red line where the person with ALS is on their own and the gray line shows where they should be. The assisted breaths with the Ambu™ bag helps you achieve a much fuller breath. Data presented to the American Thoracic Society shows that patients were able to decrease the progression of their forced vital capacity by regularly performing lung volume recruitment.

Positive Air Pressure (PAP) Devices

The most commonly used device to assist nighttime breathing in PALS is a PAP, which stands for Positive Airway Pressure. There are specific versions of these devices now designed for ALS patients.

- VPAP ST
- iVAPS
- Astral
- AVAPS
- Trilogy

PAP is not a ventilator or “respirator,” and it is not invasive. It may stimulate a breath, but it does not “breathe” for the person with ALS as a ventilator does. PAP is a supportive device that can help ease breathing difficulty and its symptoms. Use of a PAP can improve nighttime sleep, resulting in decreased daytime fatigue and sleepiness, and increased energy levels. It is not limited to nighttime use; many people use this device during the day, particularly while napping.

PAP is a machine that works by providing pressure through a mask worn over the nose and mouth or through nasal pillows. The mask is similar to the kind one might use to receive oxygen in the hospital. The machine provides pressure when the person inhales and less pressure when the person exhales. This helps the person draw in oxygen and expels carbon dioxide. The machine also has a *back-up rate*. If it does not count a certain number of breaths taken by the user in one minute, it will stimulate the user to take “extra” breaths. Some PAP devices are set by pressure and some are set to guarantee a certain volume. Your ALS team will work to help determine the best device and settings for you.

Does Oxygen Help in ALS?

Oxygen is used to treat conditions where oxygen levels are low. People with ALS do not usually have significantly low oxygen levels in the blood, unless they have a lung or heart problem in addition to ALS. However, as the muscles that assist with breathing weaken, it is more difficult to push carbon dioxide out than it is to take the oxygen in. Therefore, people with ALS do develop higher than normal levels of carbon dioxide in the blood because of poor gas exchange caused by the disease. Over time, this causes the respiratory center, the part of the brain that controls involuntary breathing, to function improperly.

The use of high levels of oxygen in ALS can actually cause breathing to slow and even stop. When the level of carbon dioxide in the blood is high, control of breathing shifts from the brain to alternative chemical receptors, which are cells that are sensitive to the presence of certain chemicals. These cells are located in the carotid artery in the neck and in the aorta, a blood vessel in the chest. The alternative receptors become accustomed to telling the body to breathe under the condition of high levels of carbon dioxide. If high levels of oxygen are then introduced, the alternative receptors stop working. Then neither the brain nor the alternative receptors tell the body to breathe. ALS patients don't usually get extra oxygen because it isn't needed unless there is a heart or lung problem and because it can knock out the body's involuntary drive to breathe.

People with ALS need the simulation of inhaling a proper breath provided by a PAP device. Oxygen levels should be measured while the person with ALS is on PAP. If the levels are low with PAP, supplemental oxygen may be required.

What Can Help with Thick Secretions, or Phlegm?

Many persons with ALS develop a problem with thick secretions, or phlegm, in the back of the throat. This can cause some increase in swallowing and breathing problems. The problems can develop for several reasons.

Dehydration: Hardly anyone actually takes in enough fluids during the day to maintain adequate hydration of the body. The basic recommendation is 8 cups (64 ounces), or 2 liters (2000 cc) of

fluid a day. It is usually difficult for anyone to drink that much fluid, and it is especially difficult when there are problems swallowing or getting to the bathroom. Many PALS take in only 400-500 cc of fluid a day. Increasing the daily fluid intake by even a glass or two can help make secretions thinner and easier to handle. Water is the preferred fluid, although anything that does not contain alcohol or caffeine may also help. Use fluids of substance like Gatorade (G2 is low in sugar), popsicles, broths or fruit juices.

Humidity: Home heating and air conditioning rob the air of moisture. Even if the furnace has a built-in humidifier, it is not usually adequate for the needs of someone with ALS. To help reduce secretions and relieve dry mouth, use a room humidifier, either hot or cold, to add extra moisture to the air. Place the humidifier in the room where the person with ALS spends the majority of his or her time, and run it 24 hours a day.

Dairy products: The protein in dairy products causes an increase in thick secretions in the throat. If someone with ALS is regularly ingesting large quantities of dairy-based products, decreasing such intake may help reduce the thickness of mucus in the back of the throat.

Medications: Some medications can help to thin the secretions if the suggestions listed above are not effective.

- **Nebulized saline:** A Nebulizer is a device that turns a liquid into small particles that can be inhaled through a mouthpiece or face mask. Nebulized saline, or salt water, can thin secretions enough for them to be coughed or suctioned out. This can be done every four hours or before meals or bedtime. The pharmacy will require a prescription for the saline and for the Nebulizer.
- **Nebulized medication:** Saline is tried first because it does not cause any side effects, but sometimes it becomes ineffective. Then a medication must be used to thin the secretions. The medication that is prescribed is called Mucomyst. It is used in the same way as the Nebulized saline.

What about thin secretions or drooling?

Nasal Secretions: Some people may find it hard to simply blow their nose. This impairment creates the need for an agent to dry up those nasal passages such as antihistamines. Other ways to keep the sinuses open are to use sinus rinse products or a WaterPik™ System for the nasal passages.

Oral Care: Oral hygiene is a very simple task to prevent buildup of bacteria in the mouth. Diluted mouthwash, especially the varieties with baking soda, is very effective to swish and spit. For those who cannot coordinate those motions, using mouth swabs to wipe out the oral mucosa will do the trick. This care should be done at least twice a day, in addition to normal brushing of the teeth and the tongue. Waterpik™ has systems for ultrasonic cleaners and suctioning.

Excess Saliva: Sialorrhea or drooling occurs when muscles such as the tongue and those controlling swallow begin to fail. It is necessary to remove as much excess saliva to prevent choking, drooling and aspirating saliva into the airways. To decrease the saliva, people with ALS can use red grape juice to swab out their mouth or papaya enzymes to thin out the saliva to make it easier to clear. Use of an oral suction wand may also be necessary. These devices come in small and portable packages. Some people with ALS can be prescribed medications such as amitriptyline (Elavil), glycopyrrolate (Robinal) or scopolamine patches that have side effects of dry mouth when used in small doses. Botox™ or Myobloc™ is also an option for consideration to block out the saliva glands. However, it is important to maintain a certain amount of moisture and clearance of the mucosa to prevent infection.

Airway Clearance Devices

Chest Physiotherapy is a technique used by respiratory therapists to mobilize secretions from the outer airways by cupping their hands and lightly pounding over the lung fields. This technique can be very exhausting for both the person with ALS and the caregiver. Similar motions can be created with vibrating massage wands available online and in department stores.

High Frequency Chest Wall Oscillator: This very important airway clearance device has recently been described as effective for airway clearance in people with ALS when therapy is started with the Forced Vital Capacity (FVC) at 70% to 40%. This device creates tiny bursts of air movement that helps mobilize the mucous in the outer airways to the central, bigger airways. This mobility is lacking in people with ALS when physical mobility decreases and it becomes harder to take deep breaths on their own.

Cough Devices: Moving mucous into the airways allows persons with ALS to cough up those secretions. However, if the diaphragm and other respiratory muscles are too weak, they may need assistance to cough effectively. A CoughAssist™ or VitalCough™ device is available to simulate the cough mechanism. It creates a negative pressure, then a positive pressure in a cycle simulating a cough. A suction device may also aid in the final clearance of those secretions from the mouth.

Suction: A suction machine can be used in the home. A battery-operated portable suction machine can be obtained for use when traveling. Suction machines use a catheter, a tube similar to those used at the dentist's office, to remove secretions from the mouth and the back of the throat. If a person with ALS cannot cough effectively or spit out secretions, a suction machine should be ordered to help clear the secretions.

What if an ALS patient catches a cold?

The ALS patient is not any more susceptible to colds and viruses than anyone else. However, when the person with ALS has an increase in nasal, oral or chest secretions, dealing with those secretions can be a lot of work. It is important to contact the ALS physicians at the Lois Insolia ALS Clinic for assistance in handling a cold before it becomes a more serious respiratory infection.

Power Considerations for Respiratory Devices

People living with ALS may require the use of devices throughout their daily activities. There are mobile power devices that will help provide access to these devices. This is a good idea in case of severe weather or other electrical failure.

Battery Geek, Cpapman, Cpap.com and Battery Power Solutions have support people available by phone. We suggest calling and providing the specific model PAP you use to get direction on which batteries would work best with your specific PAP device.

It may be helpful to have a car inverter should you like to travel using your devices in an automobile.

Here are some contacts/ websites you might find useful:

www.cpap.com	800-356-5221
www.cpapman.com	855-235-7126
www.thebatterygeeks.com/default.asp	512-692-6837
www.batterypowersolutions.net	877-445-5228

Local power companies also allow people requiring breathing devices to list with their emergency services in case of power failure.

Air Travel

People with ALS traveling with medical devices are encouraged to contact their airline before making a reservation. Devices may be carried on the aircraft and are not considered a carry-on item. The airlines recommend having enough battery power for devices to consist of one and one-half the flight time. This will ensure a safe amount of time for arrival.

The following websites are helpful for traveling:

http://www.faa.gov/passengers/prepare_fly/

<http://www.tsa.gov>

VENTILATORS

A ventilator, or respirator, is a mechanical device that provides artificial breathing for a person who can no longer breathe effectively on his or her own. In ALS, the muscles necessary for breathing, that is for adequate inhalation of oxygen and exhalation of carbon dioxide, eventually become severely impaired. Without intervention, respiratory failure can result. People with ALS are faced with the decision of whether or not to use a ventilator. This is a difficult decision to make. The person with ALS and family must be fully aware of all the consequences of the decision, whether it is for or against the ventilator. Using a ventilator will significantly alter the manner in which a person with ALS lives and the way he or she is cared for in the home or a nursing home.

Tracheostomy

Anyone who needs a ventilator for respiratory system support for longer than about two weeks will require a surgical opening of the airway called a tracheostomy. A tracheostomy is a surgical incision through which a short tube is passed into the throat. The tube keeps the airway open and connects an external ventilator to the lungs. Air from the lungs flows out through the tracheostomy opening in the throat, instead of through the nose or mouth.

The tracheostomy is located below the vocal cords. Because air movement across the vocal cords is required for speech, a tracheostomy alters the ability to communicate in a “normal” fashion. A person who could speak before the surgery, will find his or her ability to communicate greatly altered. Speech is not impossible with a tracheostomy, but it may be difficult if the ventilator is required constantly. Speech requires a special type of tracheostomy tube adapter, called a “speaking valve,” that may not be appropriate for all patients. Alphabet boards, alternative speech devices or adaptive methods of communication may be required in order for a patient using a ventilator to make his or her needs and wishes known. These devices and methods are described in detail in Section G, Speech, of this Resource Guide.

Daily care is required to keep the tracheostomy site clean and to prevent infection. To keep the airway open, secretions from the lungs must be regularly cleared from the tube with a suction catheter. Equipment in contact with the tracheostomy must be kept clean to prevent lung infections.

The Ventilator and Other Equipment

The ventilator is an external piece of equipment that provides respiratory support and/or additional oxygen. Ventilators are available in different types and sizes, including portable ventilators. The type of ventilator required depends on the individual patient’s needs and is

determined by the surgeon and/or the pulmonary or respiratory specialist involved in the patient's care. The family and other caregivers must be instructed in how to operate the ventilator and what to do if it malfunctions. Tubing that runs from the machine to the tracheostomy site carries air to and from the lungs. Humidifiers are usually required to provide moisture to the lungs and to aid in thinning secretions. Drainage systems to collect excess moisture are usually present in at least one place along the tubing. They need to be emptied periodically throughout the day. A suction machine and disposable catheters are needed to clear the tracheostomy of secretions. The frequency of suctioning depends on the patient's needs and the amount of secretions that the lungs produce.

A generator, or emergency power source, is required as a backup in case of a power failure in the home's electrical system. A portable oxygen tank and a special device called an ambu-bag can be used to provide oxygen and artificial breathing for the patient in case of a power outage or ventilator failure. The ambu-bag is connected to the oxygen tank and then squeezed to provide a breath. The family and caregivers in the home will need to be trained in the use of this device.

A person on a ventilator requires 24-hour care because of the need for suctioning and for monitoring for alarm warnings. Occasionally, an insurance plan may provide care by an outside agency for some portion of these hours. Medicare does not pay for such services. If a person with ALS is insured and is considering ventilator support, it is prudent as part of the decision-making process to check with the insurance company regarding the extent of coverage for home care. Some people may not have a caregiver or a home environment in which care with a ventilator is possible. If desired, these people can go to an extended care facility that is capable of caring for them and the ventilator.

Persons with ALS usually cannot be removed from dependence on the ventilator for breathing. The muscles necessary for breathing become so severely impaired that they eventually do not function at all. A person who is dependent on a ventilator and later has it removed will not be able to breathe adequately enough to sustain life. However, one may decide to be removed from the ventilator, even though he or she probably cannot survive without it. If that decision is made, an evaluation is normally done by a physician to determine the appropriateness of the request and to discuss the implications of the decision. Removal from the ventilator may occur in the hospital or at an in-patient hospice unit. The patient would be sedated during that time to ease the discomfort associated with the difficulty breathing, and to make the final moments as comfortable as possible. It may be minutes to many hours or even a few days before breathing ceases completely after the ventilator is discontinued.

Ventilator support alters the manner in which a patient is cared for, communicates, and lives. This issue may be brought up by the physician during one of your clinic visits at the Lois Insolia ALS Clinic. The ventilator impacts both the person with ALS and family in many ways, including financially and emotionally. The patient and family can also view a video that

demonstrates the impact of ventilator support. Although the Clinic's staff members feel it is best to view the video with a staff member present for support and additional information, the video is available on the Les Turner ALS Foundation's www.lesturnerals.org

Disclaimer: All care has been taken in preparing this document. This information is of a general nature and should be used as a guide only. Always consult your health care team before starting any treatments.

Patient and Family Resource Guide to ALS

Section 9 Insurance, Financial & Legal Issues

9

LES
TURNER
ALS
FOUNDATION

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Insurance, Financial and Legal Issues

Health Insurance

It is very important for every person with ALS to maintain health insurance coverage. ALS typically requires a variety of medical treatments and equipment. Many of these items can quickly diminish a family's assets if insurance is not available to fund a significant portion of the costs. Even medications can present an overwhelming expense.

Group Health Insurance Policies

Most people obtain health coverage through their employer or their spouse's employer at group rates. While some persons with ALS are fortunate enough to continue to be eligible for benefits through an employer, most lose their group health insurance when their employment ceases. If a person with ALS has a short-term disability plan through an employer, then the person is on the payroll until that short-term disability plan ends. At that time, the person is no longer considered to be actively working. Under a federal law, the Consolidated Omnibus Budget Reconciliation Act of 1985, or COBRA, an employee who worked for a business with 20 or more employees is entitled to continue existing health insurance coverage for 18 months after leaving work. COBRA policies can be extended to 29 months after leaving work if the coverage is being taken due to disability. Under a law that went into effect as of July 2001, people with a confirmed diagnosis of ALS can receive coverage within six months of the date of disability. For information on obtaining COBRA coverage, contact the human resources department at your place of employment.

COBRA for a person with ALS may be necessary until Medicare coverage goes into effect. However, since the employer is not required to fund COBRA coverage, premiums under COBRA can rise significantly. As a result, family members covered under the group coverage may choose to select separate health insurance at the time the person with ALS exercises COBRA rights.

Individual Health Insurance Policies

Under the Affordable Care Act, passed into law in 2010, pre-existing medical conditions cannot be used to deny coverage. Due to ongoing changes in this law, you are encouraged to find out the current status of the availability of coverage. In Illinois, find out more at: <https://www.illinois.gov/hfs/MedicalClients/AffordableCareAct/Pages/default.aspx>

Government Health Coverage

Medicare

If you are 65 or older, you are automatically eligible for Medicare. However, people with ALS who are 64 and younger, and have a confirmed diagnosis of ALS, qualify for Medicare health insurance. In order to obtain Medicare, the person with ALS must apply for long-term disability through Social Security. As soon as the first disability check is received, five to six months from the official last date of active work, the person with ALS is also covered by Medicare.

Medicare Part A covers 80 percent of most inpatient charges in a hospital or skilled nursing setting.

Medicare Part B covers 80 percent of most outpatient charges, as well as lab work and X-rays. However, a fluctuating charge for Part B coverage will be automatically deducted from the Social Security or disability check each month.

For more information, go to <https://www.medicare.gov/> or call Medicare 1-800-MEDICARE (1-800-633-4227). If you are 65 or over, go to <https://www.ssa.gov/disabilityssi/> or call Social Security at 800-772-1213.

Medicare Supplemental Coverage

Some private health insurance companies supply Medicare supplement policies, often called Medigap policies. These provide coverage for the 20% of medical charges not covered by Medicare, as well as the Part A hospitalization deductible. In Illinois, these companies are required to accept any resident over 65 with Medicare Parts A and B, regardless of the present state of health. However, companies are not required to accept those who are under age 65 and disabled.

Medicaid

You may obtain Medicaid at any age, but there are financial restrictions. Assets not including the home or car that an individual may have are constantly changing, however, a patient's well spouse may keep ~\$109,000 in assets, but the patient's name must be legally transferred off of those assets.

In Illinois, for information, go to <https://www.medicaid.gov/> or call 800 252 8635 or 312 793 2608.

Veterans Affairs (VA)

ALS has been deemed a service-related disease, so veterans with ALS automatically can receive VA benefits. For medical care in VA facilities, the highest priority is given to veterans with service-connected disabilities. Veterans with an ALS diagnosis who did not serve overseas may be entitled to receive health care benefits from the VA, including medical and respite care, as well as equipment such as wheelchairs, walkers, canes, etc. Other possible benefits include prescriptions and medical supplies, and a home improvement and structural alteration grant (HISA). Eligibility will depend upon the period of service, type of discharge, and income level.

For information go to: <https://www.va.gov/> or call 800 827 1000 (0466) or 312 663 5510.

The Paralyzed Veterans of America (PVA) is a congressionally chartered veteran's organization that has been serving veterans with spinal cord injury and disease since 1946. You may call the PVA to receive information regarding how to obtain your maximum VA benefits at 800 424 8200, ext. 12, or in Chicagoland area, 708-202-5623 or go to www.pva.org.

Illinois Comprehensive Health Insurance Plan (ICHIP)

This is a state-administered program that provides medical insurance to persons whose medical conditions make them otherwise uninsurable by conventional standards. It is funded and supervised by the State of Illinois, but operated through a private insurance carrier.

For information, contact the Illinois Dept. of Insurance at: <http://www.chip.state.il.us/default.htm> or call 312 814 2427

All Kids/The Children's Health Insurance Program

This is an insurance program for Illinois children from birth through age eighteen. The household income may be larger than allowed by the financial restrictions for public aid. For information, call 1 866 255 5437 or www.allkids.com.

Handling Health Insurance Claims

The terms and benefit levels of commercial health insurance plans vary widely, and few generalizations can be made. All policies and plans have definitions of what constitutes covered medical treatments. None of these definitions were written with ALS and its medical ramifications in mind, and the policy language will not specifically address many treatments, devices and equipment. As a result, there may be no immediate answer for coverage on a particular claim item and dealing with insurance claims handlers is an art.

What Do You Need to Know about the Policy?

Gathering information on a policy will reveal at least some of the benefits provided as well as information on:

- Policy limits: no health insurance policy is limitless, and maximum lifetime benefit amounts can become significant issues.
- Deductibles and co-payments: these are the amounts the patient will pay on covered claims.
- Claim requirements: these include what permission or notice is required before certain medical treatment is deemed “covered.”
- Provision for large case management: this provision recognizes that certain catastrophic illnesses do not fit the mold of the policy and allows for expanded coverage based on an agreement by the doctor and the insurer to an overall treatment plan. ALS should qualify if the plan has large case management.

Dealing with Claims Personnel

ALS treatments are varied and extensive and may not be specifically addressed in a policy. Many claims examiner may not have heard of ALS, or handled an ALS claim. Dealing effectively with claims personnel can go a long way toward maximizing benefits and speeding up the payment of claims.

Establish a single person as a case manager. Having a single person on the insurer’s staff as the contact point avoids needing to explain ALS every time a claim is submitted. It also allows personal relationships to develop that will help as “gray area” claims arise. The contact person should be at least at a supervisory level, with some medical knowledge and the authority to exercise some discretion. Many insurers employ such ideal contacts as nurses and other medically trained individuals.

Establish a single family representative. To aid in communication, one person should become the insurance contact representing the family.

Establish a contact at the neurologist’s office. Personnel at the neurologist’s office may be skilled at dealing with insurance claims and can run interference or otherwise expedite the process.

Educate the insurance contact. Many claims personnel have no experience with ALS; inform them of the nature of the disease, and outline what might be expected in the future.

Keep the contact advised. For policy provisions that require notice before expenditure, inform the contact early about an upcoming purchase or service. This gives the insurer time to fully consider the claim before it is a “rush” decision, and allows for input. For example, many plans have cost containment provisions, and obtaining some items at a discounted price or purchasing a cheaper but comparable item may turn a questionable claim into a covered claim.

Ask the contact’s advice. While perhaps not knowledgeable about ALS, claims personnel often know a great deal about medical needs such as wheelchair models, equipment costs, and the best suppliers. This kind of advice, in addition to the advice of Center personnel, can be invaluable whether or not the claim is covered. Insurers also often can get some drugs and medical equipment at a lower cost. Even though a claim may be denied, a friendly claims supervisor might allow the patient to buy at the insurer’s preferred provider discounted price.

Get a physician’s prescription. Regardless of the nature of a purchase, from a shower stool to a communication system, a physician’s prescription or letter of medical necessity for the item always helps support a claim of medical benefit.

Remember that *no* does not always mean *no*. Many ALS-related claims are in a “gray area” of coverage, requiring discretion on the part of the insurer. A “no” may be simply a knee-jerk reaction that the square peg of the claim did not fit into the round hole of the coverage. Further discussion regarding medical benefit may result in either a reversal, or more likely, an offer by the insurer to pay some amount toward the claim. If the claims personnel persist in denying a claim the patient feels is clearly covered, avenues of recourse exist short of filing a lawsuit. Always ask for a supervisor and then the medical director. The person with ALS may enlist the support of the former employer’s employee benefits manager to help plead the case. Clearly, a large employer may have more leverage in getting claims paid due to their payment of higher premiums to the insurer.

Built-in grievance procedures may also allow for an appeal of a claims denial. This option can be particularly helpful in employer self-insured plans where the appeals board may include former work associates of the person with ALS. Finally, a complaint to a state insurance department may be helpful. State insurance departments control the licenses of health insurers, and if a state insurance department raises the issue, it will be at least addressed promptly and thoroughly.

Choose claims battles carefully. Insurance will not pay for everything the person with ALS wants covered. Some claims will be denied. It makes little sense to battle over reimbursement for a \$150 walker if that approval may affect the insurer’s prompt decision to pay for a \$30,000 electric wheelchair.

Financial Assistance

Social Security Disability Payments

ALS may prevent a person from working, at any age. Social Security Disability payments can relieve at least part of the financial burden. The following paragraphs are extracted from the Social Security Administration Office of Disability web site, <http://www.ssa.gov/dibplan>.

How to qualify: “To qualify for benefits, you must have worked in jobs covered by Social Security. Then you must have a medical condition that meets Social Security’s definition of disability. . . If you are receiving Social Security Disability benefits when you reach age 65, your disability benefits automatically convert to retirement benefits, but the amount remains the same. . . .”

“We consider you disabled if you cannot do the work you did before and we decide that you cannot adjust to other work because of your medical condition(s). . . . Your disability must last or be expected to last for a year or more. . .

How to apply: “You should apply at any Social Security office as soon as you become disabled. You may file online, by phone, mail or by visiting the nearest office. You can find out the name and address of the closest Social Security office here. To apply by phone, call our toll-free number, 800 772 1213, and we will set up a time for your local Social Security office to contact you.” Ask for a Disability Starter Kit that can help you get ready for your disability interview and will guide you through the application process. Each kit contains: (1) A fact sheet that answers most frequently asked questions, (2) a checklist of documents and information that will be requested, (3) a worksheet to help you gather and organize information, and (4) information on how you can file your claim online.

Information needed: “Claims for disability benefits take more time to process than other types of Social Security claims—from 60 to 90 days. You can help shorten the process by bringing certain documents with you when you apply, and by helping get any other medical evidence needed to show that you are disabled. Here is what you should bring us:

- Your Social Security number and proof of your age;
- Names, addresses and phone numbers of doctors, hospitals, clinics and institutions that treated you and the dates of treatment;
- Names of all medications you are taking;
- Medical records from your doctors, therapists, hospitals, clinics and caseworkers
- Laboratory and test results;
- A summary of where you worked and the kind of work you did;
- Your most recent W-2 form, or your tax return if you’re self-employed.

IMPORTANT: You will need to submit original documents or copies certified by the issuing office. You can mail or bring them to Social Security. We will make photocopies and return your original documents. If you don't have all the documents you need, don't delay filing for benefits. We will help you get the information you need."

When benefits start: "If your application is approved, your first Social Security benefits will be paid for the sixth full month after the date we find that your disability began. For example, if we find that your disability began on June 15, 2017, your first benefit would be paid for the month of December 2017, the sixth full month of disability. Social Security benefits are paid in the month following the month for which they're due. This means that the benefits due for December 2017 would be paid to you in January 2018, and so on."

Disability Insurance

In addition to Social Security benefits, a person with ALS may be entitled to disability insurance benefits as a result of the termination of the person's employment. While a few states have state-operated worker disability funds, many people with ALS are covered by private employer or union disability insurance policies and plans with proof of being permanently disabled. Each policy will specifically define the term "permanently disabled," but most require that the person with ALS be unable to perform any type of work for which he or she has any skills or can be trained. The definition and criteria generally parallel the criteria of Social Security. A patient with a confirmed diagnosis of ALS will qualify for disability payments when his progression no longer allows her/him to work.

Disability benefits usually are not payable until some stated time period has expired. This time period is usually six months of disability, or inability to work—the same waiting period as for Social Security Disability benefits. During this "qualifying period," the person with ALS may be entitled to some form of wage continuation or short-term benefit from an employer. Benefit amounts for short- or long-term disability depend on the specific coverage or plan in effect.

Long-Term Disability Benefits

Long-term disability payments are typically based on a stated percentage of the salary of the person with ALS, up to a maximum dollar amount. Almost all plans will deduct Social Security and similar payments received by the person with ALS from the benefit amount. Payments are often made monthly, and if the payment is made under a plan paid for by the covered person (either through direct premiums or employee contributions), it should not be taxable. Payments under employer-funded plans are taxable as income. However, there is no withholding requirement.

Like Social Security, long-term or permanent disability insurers will require detailed applications and medical documentation. Most neurologists should know how to complete these forms, and most disability insurers recognize ALS as likely to be qualifying. However, it is important that physicians list in detail the specific symptoms that prevent the person with ALS from working. A complete application and a thoughtfully prepared physician certification should result in a prompt adjudication of disability and avoid insurer-requested “second opinions.”

People with ALS and their families should find out well before stopping work what the exact terms and conditions of the short-term and long-term disability plans are. There may be added requirements or benefits, such as payment by the insurer of health and life insurance premiums. Plans may also allow the person with ALS to work part-time. If so, you will need to ascertain whether the wages may offset the plan benefits. Find out how and when the person with ALS can formally apply for benefits. The plan may not allow benefits until a six-month waiting period expires. However the insurer may recognize ALS as a permanently disabling illness. This may allow for early filing, early acceptance, and prompt check receipt when the waiting period concludes.

Additional Resources

Illinois Department of Insurance/Grievance Hotline 312 814 2420 or 866 445 5364. They will investigate consumer complaints regarding health insurance companies or health care providers when they are made aware of problems in how insured persons are managed.

Illinois Benefit Access Program (formerly Circuit Breaker) 800 252 8966 or revenue.state.il.us. This is a state-funded program for seniors and the disabled that provides grants to help reduce the impact of taxes and certain medications as well as license plate discounts and ride free transit cards.

Medicare Savings Programs

You can get help from your state paying your Medicare premiums. In some instances, the programs may also pay for Medicare Parts A and B, deductibles, coinsurance and copayments if you meet certain conditions. The 4 programs are:

Qualified Medicare Beneficiary Program(QMB)

Specified Low-income Medicare Beneficiary (SLMB) Program

Qualifying Individual Program (QI)

Qualified Disabled and Working Individuals (QDWI) Program

Please go to www.medicare.gov details regarding coverage and income eligibility

The Medicine Program 866 694 3893 or Themedicineprogram.com. This program seeks to aid those who have exhausted all other sources for help with medication. It helps people apply for enrollment in one or more of the many assistance programs now available through drug manufacturers. These programs provide free medicine to qualified individuals who cannot afford to purchase expensive prescriptions. Decisions concerning which medications are provided free of charge and which individuals are accepted into the program are made solely by the various pharmaceutical sponsors. Each manufacturer has established specific criteria to determine an applicant's eligibility.

Legal Considerations

People with ALS and their families will face numerous legal issues as the disease progresses. Because ALS is a terminal illness, the issue of living wills and medical powers of attorney must be addressed. Items such as preparing various powers of attorney and protecting rights under private and public disability and health insurance plans should be addressed before the disease affects the patient's ability to act and communicate. Many of these actions can be handled by the person with ALS and their family, but some will require consultation with a lawyer. The key, however, is advance planning on the part of both person with ALS and family to avoid unnecessary legal problems in the future.

Powers of Attorney and Other Documents

Since ALS may also eventually affect a person's ability to communicate, either verbally or in writing, the person with ALS and the family should decide whether or not to use one or more "powers of attorney." A power of attorney (POA) is a document that grants another person the authority to act on an individual's behalf and to execute documents on behalf of the individual. This may range from endorsing checks to signing titles and deeds. An attorney should prepare any POA to guarantee that it will be legally enforceable and recognized, particularly if real estate transactions are involved.

The person to whom the POA is granted may be anyone who is at least eighteen years old and is legally competent. Obviously, a person with ALS should grant this power only to someone he or she feels is completely trustworthy. The person holding the power of attorney has the authority to act fully on an individual's behalf until the POA is revoked in writing. This authority is subject only to the restrictions, if any, contained in the power of attorney. The need for a power of attorney may seem diminished with banking transactions, one of the major uses for a power of attorney, now increasingly done electronically and not requiring signatures. Having such a document, however, may prove to be essential if the person with ALS needs to execute unforeseen legal documents and cannot sign his name or otherwise act.

Medical Powers of Attorney

Persons with ALS will face a number of significant life or death decisions regarding their care. These decisions include whether to use a ventilator or to refuse “heroic” measures such as resuscitation. Since ALS may affect an individual’s intelligence or ability to make sound judgments, it is strongly suggested that these decisions be made early in the disease process. This allows for full discussion with loved ones, provides assurance that the person’s desires are clearly known, and permits such desire to be recorded in one or more effective documents.

Health care professionals will require effective and clear documents before they will refrain from taking all medically available means to prolong life. These documents may be called durable powers of attorney for health care, living wills, advance directives or some other name, and the form of such documents depends on state law. In Illinois and many other states, a person may also use Five Wishes (agingwithdignity.org or 888 594 7437) to express their end-of-life wishes. People with ALS may need an attorney’s assistance to guarantee that their wishes will be carried out. Hospitals may also require specific “Do Not Resuscitate” (DNR)/Practitioner Orders for Life-Sustaining Treatment (POLST) forms. If a person decides to forego any form of treatment or assistance, inform the individual’s attorney and physician to ensure that all needed documentation is in place.

An in-depth discussion with the neurologist will help the person with ALS to better understand all the possibilities that may occur, and to address each in the written document. The more specific the written document, the more likely the person’s desires will be carried out when situations arise. Like wills and other powers of attorney, documents dealing with heroic medical action and invasive life-extending surgery are not irrevocable. Should a prior decision be reconsidered, the person with ALS should retrieve and destroy any documents previously given to health care providers or family. Another way to facilitate changes of decisions is to have the documents prepared and executed but retained by a trusted family member or friend for delivery “when needed.”

The goal is to communicate, as specifically as possible, each person’s specific desires. Communication is just as important for the person who wants all of medical science’s resources used to prolong life as it is for the person who wants less than “full” medical treatment. The family will feel much more comfortable knowing that actions are truly determined by the person’s wishes, best evidenced by a signed and unambiguous document.

Wills and Estate Planning

Since ALS is a fatal disease, people with ALS patients should consider preparing a will and begin estate planning. The “plan” may range from utilizing existing life insurance and making specific tangible bequests to handling the custody of minor children. Whether a person needs a

will, which is a written directive on how the person wants his or her affairs handled after death, is as personal a matter as are the contents of a will itself. It should be considered carefully in consultation with the family's attorney and any financial advisors. If someone dies without a will, that is dies "intestate," state laws set forth how the estate will be distributed. These laws often provide for specified shares of the estate to a surviving spouse and to the children. However, such a simplistic formula may not be appropriate in the light of divorces, extended families and other special situations.

A will allows the individual to divide property appropriate to their circumstances and wishes. A will should be considered soon after an ALS diagnosis. If circumstances change as the disease progresses, the will can be changed or a new will can be drawn. A will is not irrevocable until death. However, a person with ALS should not procrastinate. Since ALS often affects a person's ability to communicate, a person who waits too long may not be able to clearly express his or her wishes. In addition, a fully debilitated and non-communicative patient may be perceived as not being of "sound mind" to execute a will.

The cost of a will varies, based on the complexity of the estate. A simple will should cost no more than a few hundred dollars. Will kits exist that allow for do-it-yourself wills and can be obtained online. However, we recommend the specialized skills of attorneys and accountants be used to prepare estate documents.

Life Insurance

Life insurance proceeds are often the major asset of an estate. A will is not necessary to specify how life insurance proceeds are divided. The person with ALS should specify a beneficiary in the policy itself. Shortly after diagnosis, it is important to gather up all life insurance policies—individual, group, mortgage and credit—to make certain the beneficiary designations are correct. Be sure premium payments are made promptly. **DO NOT LET A POLICY LAPSE.** ALS is a condition that makes obtaining further life insurance virtually impossible. A lapse in payment may result in cancellation of the policy with no right of reinstatement. Group policies, purchased through an employer, should be continued. Even when the person with ALS can no longer work, conversion rights exist. Check with your employee benefits manager to be sure that all conversion rights are exercised. Some companies have disability plans (discussed in detail under Disability Insurance) that make premium payments on the person's behalf once disabled.

Life insurance can also be a benefit before death. In addition to traditional policy provisions that allow for borrowing against the policy, "living" benefits may also exist. Some insurers and others have plans which allow a terminally ill individual to receive the policy proceeds prior to death, with the actual payment amount discounted by some percentage. Although this results in a reduced payment, the funds can be used for current and future needs. This is called a viatical settlement.

Family and Medical Leave Act

The Family and Medical Leave Act, or FMLA, is a federal law that requires employers to allow an employee up to 12 weeks leave per year for certain family and medical-related situations. As a serious health condition, ALS is an eligible illness under FMLA, provided the employee demonstrates that he or she is needed to care for a spouse, parent, or child who has ALS. FMLA time off is unpaid after the employee's vacation and sick time is used; however, two important rights are granted. The employee is entitled to continued employee benefits during the period of the leave, as well as to job security. This means that the employer must allow the employee to return to his or her job or a comparable position at the end of the leave period.

The FMLA does not apply to workers in small companies (as defined under FMLA) where worker absence would present an undue burden on the employer, or if the employee's position is within a 10 percent "key personnel" category, as specified by the employer. In addition, some seniority requirements exist.

An employee who is considering leave under FMLA should contact their human resources (HR) manager to ensure that all substantive and procedural requirements are met. Some states have enacted additional laws that offer greater leave than FMLA provides, and an HR manager should also be aware of any additional benefits such state laws provide.

Legal and Insurance Resources

The following offer a variety of services. Some groups have income or geographic restrictions:

Center for Disability and Elder Law **312 376 1880**
www.cdelaw.org

Service Cook County and senior citizens in Chicago.

Illinois Attorney General Office **312 814 3276**

www.illinoisattorneygeneral.gov Assists those who are disabled with a variety of services, including accessibility issues.

Disability Rights Bureau **312 814 5684**

A division of the Attorney General's Office

National Academy of Elder Law Attorneys naela.com

Site includes a listing by state of attorneys who specialize in elder law.

Public Benefits Hotline **888 893 5327**

Offer legal advice and assistance to low-income Cook County residents on matters such as Medicaid and childcare.

Disclaimer: All care has been taken in preparing this document. This information is of a general nature and should be used as a guide only. Always consult your health care team before starting any treatments.

Patient and Family Resource Guide to ALS

Section 10 Genetics of ALS

10

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Genetics of ALS

About 90 percent of people with ALS have no known family history of the disease – they are the only affected person in their family. For these individuals, the disease is called sporadic ALS, or SALS. Although genetic risk factors may yet be identified in SALS, the disease is not directly inherited in a family. The remaining 10 percent of people with ALS do have one or more affected family members and these individuals have an inherited form of the disease called familial ALS, or FALS.

Familial ALS

Genes and Chromosomes

Genes are very small units of inherited information that are found inside each of our cells. The genes provide instructions that direct the cell to make proteins, which are important in the individual's development and physical characteristics. Scientists now think that there are about 20,000 human genes that provide our cells with instructions on how to grow and function. Most genes are present in pairs, and a child gets one copy of the gene of each pair from the mother and one from the father. Genes are our cells' 'recipes' for proteins. Therefore, if a gene contains a change, also known as a mutation, the instructions of that gene are changed and this can result in an abnormal or absent protein. Genetic mutations can lead to genetic disorders, since the gene no longer provides the instructions that the cells need to function.

Everyone has "FALS genes" – when working properly, they provide instructions for our brain and nerve cells to make proteins necessary for normal functions. However, if one of these genes has a mutation that causes it to create an abnormal protein, that can lead to the symptoms of FALS.

If we think of genes as books, we can think of chromosomes like bookshelves – they contain the genes and keep them organized. Humans have 23 pairs of chromosomes in each cell, for a total of 46. The first 22 numbered pairs are the same in men and women, and are called autosomes. The 23rd pair of chromosomes are the sex chromosomes, which help determine if a person is male or female. Males typically have one X and one Y chromosome, and females typically have two X chromosomes. Egg and sperm cells each contain 23 chromosomes – one of each pair. When these two cells combine, the resulting cell contains a full 23 pairs of chromosomes.

How Is FALS Inherited?

The most common inheritance pattern for FALS is called *autosomal dominant*. Autosomal means the gene that carries the mutation that causes the disease is located on one of the autosomes – the chromosomes that are the same in men and women. If a parent is affected, then

female and male offspring are equally likely to inherit the gene copy with the mutation or gene copy without the mutation. Dominant means that only one copy of the gene pair needs to have a mutation to cause symptoms of ALS.

Each parent randomly passes on one copy of each gene pair to the offspring. If a child has one parent who has FALS and one parent who does not, that child has a 50% chance of inheriting the FALS gene mutation and a 50% chance of inheriting their affected parent's non-mutated, working copy of the gene. The chance is 1 out of 2, or 50%, because the parent who has FALS will pass on either the mutated copy of the FALS gene (leading to an increased risk for FALS for that child) or the normal, working copy of the FALS gene (leading to no increased risk for FALS). FALS gene mutations are rare, so the unaffected parent will always pass on a working copy of the FALS gene in question.

A person who inherits a mutation for FALS can have up to a 90% chance of developing symptoms by age 70; however, each gene and mutation is different so this percentage can vary widely. Therefore, inheriting the gene for FALS does not guarantee that the person will develop ALS, and the severity and scope of the onset of the disease cannot be predicted. The cause of the variability of the disease progression is unknown.

Genetic Tests

Can a genetic test diagnose ALS?

No, a diagnosis of ALS cannot be made by a genetic test. A neurologist familiar with ALS makes the diagnosis after reviewing a person's symptoms, the results of a neurologic examination and the results of nerve and muscle function tests. Clinically, SALS and FALS are identical.

Is there a genetic test for FALS?

Yes, but only for specific genetic mutations that have been identified to cause FALS. Currently, about 60% of all FALS cases can be connected with an identified FALS gene. The remainder of FALS cases have genetic causes that have not yet been identified by researchers.

In 1993, Dr. Teepu Siddique, at the Les Turner ALS Research Laboratory, now part of the Les Turner ALS Research and Patient Center at Northwestern Medicine, working with collaborators from Massachusetts General Hospital and Duke University, identified the first gene that causes FALS. Changes in this gene have been identified in about 20% of the families with FALS. This gene, located on chromosome number 21, is called copper-zinc superoxide dismutase, or *SOD1*. *SOD1*'s normal job is to interact with certain substances in the body, called free radicals, which can harm cells. Normally, *SOD1* changes the free radicals so they are no longer harmful. Researchers think that mutations in the *SOD1* gene cause the gene to function in a new way that

somehow damages or injures motor neurons instead of preventing injury to the cell/motor neuron.

Another gene called *C9orf72* has been found to cause FALS in about 40% of affected families. The protein that this gene codes for is important for sending and receiving signals between neurons. The *C9orf72* protein likely plays a role in many processes involving the chemical cousin of DNA, known as RNA. This protein is thought to influence the production of RNA from genes, the production of proteins from RNA, and the transport of RNA within the cell. The *C9orf72* gene contains a section of repeated letters – like a word in a paragraph that has been repeated over and over. This is a normal part of this gene, and up to a certain threshold the gene functions normally. However, if the number of repeats is beyond that threshold, the protein that the gene encodes is abnormal and this puts a person at increased risk to develop symptoms. Testing for this gene ‘counts’ the number of repeats present in each of the two copies of this gene that everyone has. Mutations in this gene have also been found to cause other symptoms, including frontotemporal dementia (FTD) and Parkinson-like features.

There are approximately 20 identified genes that cause FALS, but they are much rarer than *SOD1* and *C9orf72*. Together, all other known genes account for about 60% of all FALS.

Forty percent of families with FALS do not have a mutation in the *SOD1* or *C9orf72* gene or other identified genes. Therefore, FALS in these families is caused by a mutation in other genes that have not yet been identified. Because we don’t know where these genes are located, we cannot test for them in either patients or their family members. Researchers are searching for other genes that might cause FALS, but at this time we do not know the genetic cause of FALS in all FALS families. The determination that an individual has FALS is typically based on family history (more than one family member with ALS) rather than on a genetic test.

How is the genetic test done?

A blood sample is taken and sent to a specialized lab. Our blood cells contain DNA, and this is separated and removed for testing. DNA is the language of genes – the substance that makes up the chromosomes and controls a cell’s activities. The gene or genes being tested can then be amplified, or copied. Using a variety of methods, the sequence of the gene can be ‘proofread’ and compared with the sequence of a normal, working version of the gene.

Who should have a genetic test?

Testing is appropriate for anyone who has symptoms of ALS and a family history of ALS, such as a parent, grandparent, aunt, uncle, brother, or sister who has or had the disease. Additionally, if the family history is unknown or if a parent passed away at a young age, testing may be appropriate.

A positive genetic test means that the genetic cause of a patient's FALS has been identified. However, not all people with ALS will have a mutation – about 60 percent of all FALS patients have a mutation in a FALS gene for which we can currently test. A negative test means that the genetic cause of the ALS has not been identified in the family and does not change the diagnosis or the chance that a family member might develop ALS. Other currently unidentified genes cause FALS in about 40% of FALS families. Researchers might ask for samples from additional family members in order to help identify these other genes.

A person with a family history of FALS might have a genetic test even if he or she doesn't have symptoms. This is called pre-symptomatic testing. Testing is available only if a mutation in a known FALS gene has been found in a family member who has ALS. A negative or a positive result of a pre-symptomatic test in a FALS family can have a great psychological impact. Therefore, genetic counseling and a neurological evaluation are required before such testing. Part of the discussion with a genetic counselor will involve the benefits and limitations of getting pre-symptomatic testing for FALS. Currently, there are no preventative options, the age at which a person with the gene will get ALS is unpredictable and it is not even certain that a person with the gene mutation will definitely get ALS. However, individuals may be interested in learning their status for financial or insurance planning purposes. Additionally, there are reproductive technologies available that can greatly reduce the risk of passing on a FALS gene mutation, therefore ensuring that a person's children will not be affected. Everyone has different reasons for choosing to test or not test, and it is a very individual decision – even siblings within the same family may feel differently about whether or not they want to be tested. Pre-symptomatic testing is a big decision, and should not be taken lightly. The Genetic Counselor at the Lois Insolia ALS Clinic at the Les Turner ALS Center at Northwestern Medicine can discuss in detail the issues involved in pre-symptomatic testing.

Laboratory-Based Genetic Research Studies

Genetic research studies, unlike drug studies, do not provide a potential direct therapeutic benefit to the patient. However, by investigating the genetics of ALS, researchers hope to aid in the development of new treatments and prevention. One of the Les Turner ALS Research Laboratories, headed by Dr. Teepu Siddique, approaches this task from several aspects:

- Gene studies in familial, or hereditary, ALS (FALS)
- Animal studies
- Genetic studies of risk factors in sporadic, or nonhereditary, ALS (SALS)

Genetic Studies in FALS

For large FALS families with no identifiable genetic cause, several strategies may be used to find potential genes of interest. One strategy is a linkage study, which involves collecting blood samples from both healthy and affected family members and then studying genetic markers to try to pinpoint an area of shared inheritance amongst affected individuals on a chromosome where an ALS gene may lie. Once such an area is found, additional families are included in the study to help narrow the region until a single affected gene can be identified.

A second gene mutation that causes an inherited form of ALS was also identified at the Les Turner ALS Research and Patient Center at Northwestern Medicine. The gene is responsible for a rare, slowly progressive, early-onset form of the disease, called juvenile inherited ALS (ALS2). The gene mutations in *ALS2* may cause a loss of function of the alsin protein. Identification of this and other FALS genes assist researchers in determining cellular pathways that may intersect with those of mutant *SOD1*, and other FALS genes.

Genetic studies in SALS

Because the causes of sporadic ALS are unknown, Dr. Siddique's research team at the Les Turner ALS Research and Patient Center at Northwestern Medicine is trying to determine what genetic factors may "predispose" an individual to developing sporadic ALS by conducting genetic risk factor studies. Genetic markers in SALS patients are compared to immediate family members, either parents or siblings (preferably a brother or sister who is older than the age of the patient at the onset of symptoms). Participating in a genetic research study is voluntary and confidential. Typically, participation only requires having a blood sample drawn at a physician's office or hospital clinic and sent to the research laboratory, as well as answering a few voluntary questionnaires regarding family history and environmental exposures. The research program covers all costs associated with the study.

Animal Studies

In 1994, researchers developed a strain of mice that have the *SOD1* mutation. This animal model helps the researchers understand how a change in the *SOD1* gene can lead to the symptoms of ALS and how the disease develops. Mouse models exist for other FALS genes as well. These mouse models also allow researchers to test the effectiveness of possible drug treatments on the disease. New therapies are being tried on this animal model to slow or halt the progression of ALS. Although results are still in the future, gene therapy to correct mutations is also being studied.

Disclaimer: All care has been taken in preparing this document. This information is of a general nature and should be used as a guide only. Always consult your health care team before starting any treatments.

Patient and Family Resource Guide to ALS

Section 11 Useful Resources

11

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lesturnerals.org
info@lesturnerals.org

Useful Resources

Personal Computers

“The fact is that my computer has been as vital to my well-being as my wheelchair or any medicine.” —Ben, a Chicago-area ALS patient

Most people with ALS have found that personal computers add significantly to their ability to cope with the disease. As the disease affects each person in different ways, so, too, the personal computer can help each one in different ways. The personal computer can be thought of as an extension of yourself. It can help do many things you can no longer easily do. The computer can provide information about ALS, support from other patients, and techniques for dealing with limitations imposed by the disease. It can provide a way to communicate with relatives and friends, or serve as the “voice” for a person who can no longer speak. It can help you shop online when your ability to physically shop is limited or help you read a book if you can no longer turn pages. There is a good reason for calling it a “personal” computer—it can be adapted to your personal needs. All you need is an open mind, some patience, and a positive outlook.

A recently diagnosed person with ALS (PALS) will probably be able to use a computer, tablet or smart phone in a conventional manner. The many adaptive devices available allow each patient to continue to use and enjoy the computer regardless of the progression of their ALS. If using a standard keyboard becomes difficult, an on-screen keyboard can be used. This keyboard can be controlled by many different devices ranging from a thumb switch, such as the one used by English physicist Dr. Stephen Hawking, to a device activated by blinking an eye.

Some Uses for Personal Computers (with appropriate software)

Environmental control: A computer can be adapted to help control the world around you. For example, it can turn lights on and off, turn the TV on and off, change TV channels, or signal for help to someone in another room or to a neighbor. With devices like Google Home or Amazon Echo, you can play music, ask about the weather, sports scores or the stock market or even order your groceries.

Household administrative chores: Classical uses for a PC include budgeting, taxes, tracking investments, and tracking income and expenses. Banking and investing can be done online. Even if your handwriting is illegible, you can still pay bills or perform other personal business transactions. Managing medical expenses is especially useful for PALS. On a simple spreadsheet you can record physician, laboratory, hospital, equipment, and prescription charges; insurance payments; and out of pocket costs. The spreadsheet can be very useful at tax time and for negotiating with insurers.

Recording disease progress: A computer can help to track your medical condition. You can record symptoms as they appear or change, the effects of therapies (such as vitamin and mineral supplements, medications, activities, exercises, and alternative medicine techniques), and the results of medical tests. Keeping a record of data provides a picture of the disease progression. It can act as a base for discussions with your physician and may even help the physician make recommendations for therapies. There are apps being developed to track ALS disease progression that will be more widely available in the near future.

Communication assistance: For many PALS, losing speech is the most frustrating aspect of the disease. If this happens to you, a computer can help restore your ability to communicate. It can take a phrase you have keyed, convert it to speech and play it through a speaker. Commonly used phrases can be stored in the computer and played when they are needed. With a speakerphone, you can even make telephone calls. Refer to [Section 7, Speech, for information on communication devices.](#)

Correspondence: At some point you may find that your handwriting is illegible and that it may be easier to type and communicate online.

Games: The computer makes a superb game platform. A number of games are included as part of the computer's basic programming support or can be easily purchased online.

How Is the Internet Useful to PALS?

Many websites offer information about ALS, such as current and contemplated therapies, advocacy efforts and equipment designed to assist PALS in their daily lives. Information about legislation passing through Congress that relates to ALS can also be found at www.lesturnerals.org or other medical advocacy groups. You can follow its progress and express your opinions to your Senators and your Representatives. As with any collection of information, care needs to be taken to realize that some “quacks” also frequent the internet. Chat rooms and bulletin boards are helpful in exposing dubious products and therapies. Of course, you can always contact a member of the Foundation's Home and Community Team or our physician to confirm the truthfulness and appropriateness of information that you read online.

Support groups: ALS support groups are available online. PALS and caregivers share ideas and experiences, offer help and emotional support. The interchange is not immediate in an online support group, but PALS and caregivers are involved from next door to literally around the world. “Chat rooms” are a way of communicating with others in a conversational dialog. Chat rooms occur on a scheduled basis. The Les Turner ALS Foundation also offers virtual, real-time support groups, allowing those who cannot attend in person to join a support group. Check the Foundation's website at www.lesturnerals.org to learn more.

Shopping: Online shopping can replace nearly all physical shopping and can save time and energy for persons with ALS.

Reading: If you have difficulty holding a book, newspaper, or magazine and turning pages, you can easily find the information online. Books, including access to your local library, many of which are free, can be downloaded and read on a computer. Most newspapers and many magazines have online versions and you can have access to publications of every locality as well as national, international, and specialty publications.

Electronic Books: For many people with ALS, the pleasure of reading books becomes a casualty of the disease. Books become too heavy to hold. Pages are too hard to turn. The struggle outweighs the pleasure of reading. Electronic books, commonly referred to as e-books, are readily available.

There is no dearth of formats for e-books or of devices for reading them. A little bit of research is needed to decide on the environment best for you.

The basic formats for electronic books are Microsoft Reader and Adobe Acrobat e-Reader. All have the capability of setting multiple bookmarks, a search function, selection of text size for clarity, highlighting, and annotating. You can read an e-book on a computer, either desktop or laptop, Windows PC or Macintosh; a mobile phone, tablet/iPad.

Microsoft Reader is free computer software for electronic books. It is available for computers running Windows, but not for Macintosh computers. The books are downloaded to and read on the computer, tablet or smart phone.

Adobe Acrobat eReader is also free computer software. Books in this format are read on a computer, either a desktop or laptop, Windows or Macintosh.

Acquiring an e-book is done on the Internet. Some suppliers of e-books include:

Amazon.com	amazon.com
Barnes and Noble	bn.com
Book Bub	www.bookbub.com

Purchasing a separate device, as is the case with an iPad, Kindle, etc, adds to the cost, but significantly enhances the convenience and the prices are reasonable.

Hobbies

Hobbies that can no longer be actively pursued due to physical limitations can still be followed “virtually” through the myriad of internet sites devoted to almost any endeavor.

Transportation and Travel

The Foundation is not affiliated with any of the transportation companies listed below. You will need to contact the company for rates, service areas, etc.

Open Taxis – Fully accessible vehicles equipped for manual or power wheelchairs. For additional information and/or scheduling call 773-657-3006

Special Needs Chicago: Fully accessible vans and service cars available with disability trained and experienced drivers. Transportation provided on a same-day basis with a 2-3 hour advance call. For additional information contact Michelle Dacy, General Manager, at 630-668-9999.

Pace: Special bus service is available for wheelchair-bound patients in various suburbs and the city.

RTA ADA Paratransit program – program available to those whose disability and/or health condition prevents them from using CTA and/or standard PACE service. These vehicles are equipped to transport patients on a ventilator. Patients must apply and be approved to use this service. Call 312-663- 4357 for an application.

First Transit, Inc.– program through the Illinois Department of Healthcare and Family Services to assist patients on Medicaid with non-emergent transportation needs. 877-725-0569

Handicapped Parking Privileges: For information about cards or license plates: 217 782 2434 or 312-793-1010.

Travel Information: Smarter Travel; <https://www.smartertravel.com>

Van Conversion

When you are considering using a manual or power wheelchair, you must consider how you will transport it in your present car or vehicle. You may need to consider a mini-van or full-size van, depending on the person's ability to transfer from the car to the wheelchair, the strength and ability of the caregiver, and your lifestyle and resources. Get good advice before purchasing the wheelchair or van. Consider that your garage may need a ramp or other modification. If you do not use a garage and you park on the street, you may need a special parking zone sign from your city or town for your parking area.

Accessible Van-Rental and/or Sales

- **Mobility Works**, 888 378 9166 or mobilityworks.com is a nationwide supplier of wheelchair accessible vans and driving aids. They stock new and pre-owned mobility vans, rental vans and a variety of adaptive driving equipment. Locations in Illinois include Niles, Villa Park and Plainfield.

- **Sherman Dodge**, Skokie, IL 855 828 5268, www.shermanmobility.com
- **New Ability Inc.**, Melrose Park, IL, 708 345 3939. Provides adaptive vehicle equipment ie: hand controls, ramps, etc. www.newabilityinc.com
- **Wheelchair Getaways** – van rentals, www.wheelchairgetaways.com 800 637 2597 or 847 967 2083.

Regional Transportation Authority (RTA)

The RTA offers a Reduced Fare Permit for senior citizens and qualified persons with disabilities to ride **ALL RTA** services at a reduced rate. These services include:

- CTA Buses
- Rapid Transit
- Pace Buses
- Metra Trains in Cook, DuPage, Lake, Kane McHenry and Will counties

Benefits of using the reduced permit are fares that are approximately half the full fare and you save on gas.

Who is eligible for the permits?

- All seniors 65 years or older
- Persons with disabilities who have been prequalified by Social Security, the Veterans Administration or their doctor.

How do persons with disabilities apply?

- You will need the following pieces of identification: Driver's License, State ID card or Passport and a Veterans Validation of 100% disability or valid Medicare card as proof of disability. This can be obtained by calling the Social Security Administration at 800 772 1213, visiting any Social Security Office or going online at www.ssa.gov

How long does it take to receive my permit?

- It takes approximately three to four weeks to receive your permit. If you have questions call 312 913 3110

Alternative Treatments

PALS have always been interested in alternative treatments. With the rise in the use of social media and the ease of mass communications, the promotion and marketing of such treatments has never been easier. As a result, the International Alliance of ALS/MND Associations, of

which the Les Turner ALS Foundation is a founding member, has developed a “Statement on Alternative Treatments, which follows:

International Alliance of ALS/MND Associations

Statement on Alternative Treatments

The International Alliance of ALS/MND Associations recognizes the interest that people affected with ALS/MND can have in seeking alternative forms of treatments.

The International Alliance supports the individual’s right to choose what treatment they wish to undertake but would strongly encourage anyone considering any treatment to fully discuss the issues around such treatment with their doctor, health care professional and family before making a final decision.

The International Alliance believes that treatments for and research into ALS/MND should be legal, have a sound scientific rationale and have the potential to bring us closer to the cause, treatment or cure for the condition.

The International Alliance only recommends treatments that have been proven through thorough scientific testing and clinical trials to be safe and effective.

An excellent resource in identifying tested alternative treatments is ALS Untangled, through the Duke University ALS Clinic, at www.alsuntangled.com

The International Alliance recommends all providers of non-proven and/or alternative treatments for those affected by ALS/MND to conduct scientific research and submit papers to the appropriately recognized journals so that peer review can be undertaken and the information can be shared amongst the whole ALS/MND community.

Guiding Principles

When looking at alternative treatments, the International Alliance would recommend that you give careful consideration to the following questions to help you think through the issues and to make an informed decision:

What claims are being made for the treatment?

Often there will be claims of stopping the progression of the disease or a reversal/improvement in symptoms. Check who is making these claims and what evidence there is to back them up. If the claims are genuine then they will have been published in recognized scientific journals and there will be published results of clinical trials. Often there will be testimonials from people with ALS/MND of the improvements they have experienced. It is important to find out how

long the improvements lasted for as there is a recognized phenomenon called the “placebo” effect which occurs when individuals experience beneficial effects only because they believe that they’re receiving beneficial treatment. Does their doctor agree that there has been a benefit in undergoing the treatment?

How are people finding out about the treatment?

Is it mainly being promoted through the mass media, i.e., newspapers, magazines, the Internet, etc.? Any genuinely safe and effective treatment will be promoted and recommended by your doctor and the ALS/MND associations.

Who is offering the treatment?

Is the treatment being offered by an appropriately recognized institution? Is it being offered by a number of different institutions or just one? If it’s just one then why are others not following and doing the same? Do you have to travel to another country to receive the treatment and if so why is it not available in your country?

What are the risks involved?

Is it clearly stated what risks are involved in undergoing the treatment? Are there any side effects and how long may they last? Has the treatment been proved to be safe and effective and if so how was this done? Don’t forget that there can be financial risks associated with treatment particularly if it is expensive and involves overseas travel.

What follow-up monitoring is carried out after the treatment?

Follow-up monitoring is extremely important not just for you but for all those with ALS/MND. For you it is important to know that you will be monitored so that many adverse effects can be picked up as soon as they occur. For all those with ALS/MND they need to know if the treatment is successful and that they can rely on the claims being made.

Web Sites Related to ALS

There are numerous web sites related to ALS. We recommend the following:

The Les Turner ALS Foundation www.lesturnerals.org

ALS Untangled www.ALSuntangled.com

Northeast Amyotrophic Lateral Sclerosis Consortium (NEALS)
www.alsconsortium.org

Centers for Disease Control (CDC) www.cdc.gov/als

ALS Association

www.alsa.org

Muscular Dystrophy Association

www.mdaua.org

International Alliance for ALS/MND Associations

www.alsmndalliance.org

Disclaimer: All care has been taken in preparing this document. This information is of a general nature and should be used as a guide only. Always consult your health care team before starting any treatments.